Edited by
Peter Cameron
Gary Browne
Biswadev Mitra
Stuart Dalziel
Simon Craig

Textbook of PAEDIATRIC EMERGENCY MEDICINE

Third Edition

ELSEVIER

Textbook of Paediatric Emergency Medicine

THIRD EDITION

Editors

Peter Cameron, MBBS MD FACEM

Professor of Emergency Medicine, The Alfred Hospital and Monash University, Melbourne, Australia

Gary Browne, MD MBBS MSpMed FRACP FACEM

Professor of Emergency Medicine, The Children's Hospital at Westmead
Chair of Discipline of Emergency Medicine, The University of Sydney
Head of Academic Emergency Medicine, The Children's Hospital at Westmead, Sydney, Australia

Biswadev Mitra, MBBS MHSM PhD FACEM

Professor of Emergency Medicine, The Alfred Hospital and Monash University, Melbourne, Australia

Stuart Dalziel, MBChB FRACP PhD

Director of Emergency Medicine Research, The University of Auckland and Starship Children's Hospital, Auckland, New Zealand

Simon Craig, MBBS MHPE MPH FACEM

Adjunct Clinical Associate Professor, Monash University; Emergency Physician, Monash Medical Centre, Monash Health, Melbourne, Australia

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Content Strategist: Laurence Hunter
Content Development Specialist: Alexandra Mortimer
Content Coordinator: Kirsty Guest
Senior Project Manager: Manchu Mohan
Design: Amy Buxton
Illustration Manager: Amy Naylor

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Preface to third edition

It is now more than 10 years since the publication of the first edition and interest in paediatric emergency medicine continues to grow at a local and international level. For generalist clinicians, who see both adult and paediatric patients, children account for a minority of the daily workload, however the anxiety associated with providing safe and effective care to this population ensures a keen interest in updating skills and knowledge. The principles in managing paediatric patients are the same as adults; however, there are significant differences in patterns of illness and responses to illness. In addition, the therapies available vary widely between adult and paediatric practice. Not all of the differences have been evidence based; however, the last couple of decades have seen a major improvement in research supporting guidelines for the management of paediatric emergencies.

The formation of research groups such as PREDICT (Paediatric Research in Emergency Departments International Collaborative) and PERN (Pediatric Emergency Research Network) has generated solid evidence to underpin some of the new guidelines for assessment and management of paediatric patients. The assessment of head injuries, oxygen therapy, fluid management, sedation and analgesia are just some of the areas that have had major changes in approach over the last few years due to excellent research by the paediatric research networks.

New technologies and better application of older techniques have also led to changes in practice. For example, ultrasound is now being used more routinely in clinical practice for placement of lines, incision and drainage, resuscitation of critically ill patients and assessment of minor injuries. More research needs to be done into what place bedside imaging takes in specific circumstances, such as FAST in trauma patients. It is likely that management algorithms in children are significantly different to adults, because of different patterns of injury and response to injury.

This new third edition attempts to capture the major changes in guidelines across the specialty, whilst refining established approaches to practice in most subject areas. The authors have significantly updated the resuscitation and trauma sections, clinical applications of bedside ultrasound, analgesia and sedation. There is also a new focus on the teaching and research sections.

This revision has involved the focused input of more than 100 clinicians and academics for over 1 year. The dedication to purpose and attention to detail is reflected in the quality of the book. I would also like to thank the publication staff at Elsevier, in particular Ms Alexandra Mortimer, and my Personal Assistant, Ms Angela Hodges, for coordinating the many people involved and keeping focus.

2018

P. C.

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Preface to second edition

Following the successful launch of the first edition as a companion to the Adult Textbook of Emergency Medicine, we have had considerable feedback on the content and layout from doctors, nurses and paramedics working in paediatric emergency practice from around the world. The feedback has been positive, particularly regarding the importance of a text with a standardised, easily accessible format. Despite major advances in computerisation, most clinicians studying detailed clinical material, still prefer a well-presented book with carefully edited text to on-line material. It is likely that further advances in technology will enable electronic versions of this text shortly and it is intended for this to occur after the printed version has been released.

In this edition we have reviewed each chapter and updated guidelines and management protocols where appropriate. Material has been reviewed by chapter authors and editors to ensure that it is consistent with best practice internationally. The structure has remained the same to enable easy access for readers.

Since the first edition, there has been consolidation of paediatric emergency medicine as a specialised domain of clinical expertise. Standards for paediatric patient care in emergency departments have been published in the United Kingdom and elsewhere and training programmes have been developed in many countries. There is a high degree of cooperation within the international emergency paediatric community and international networks for research (e.g. PERN – Paediatric Emergency Research Network) and other activities are being considered. Hopefully texts such as this can further consolidate the convergence of clinical knowledge and practice internationally.

This edition was developed over approximately 18 months with contributions from authors around the world including Australia, New Zealand, United Kingdom, Hong Kong, and the United States. The commitment and effort required to coordinate and cajole the many people involved, required dedication

from all involved but we would particularly like to thank Helen Leng from Elsevier who remained focused throughout.

2012

P. C.

G. J.

I. E.

G. B.

J. R.

Contributors

Richard Aickin, MBChB FRACP FACEM, Paediatric Emergency Specialist, Starship Children's Hospital, Auckland, New Zealand

David Armstrong, MBChB FRACP MD
Adjunct Associate Professor, Paediatrics, Monash University
Head, Department of Respiratory Medicine, Monash Children's Hospital, Clayton, Australia

Franz E. Babl, MD MPH DMedSc FRACP FAAP FACEP, Associate Professor of Paediatrics, Royal Children's Hospital, Murdoch Children's Research Institute and University of Melbourne, Melbourne, Australia

Bindu Bali, BMedSci BMBS MRCPCH FRACP MMEd
Paediatrician, Emergency Department, Sunshine Hospital
Paediatrician, Royal Children's Hospital, Melbourne, Australia

Peter L.J. Barnett, MBBS FRACP MSc FACEM MSpMed
Consultant, Emergency Department, Royal Children's Hospital
Associate Professor, Department of Paediatrics, University of Melbourne
Honorary Research Fellow, Emergency Services, Murdoch Children's Research Institute, Parkville, Australia

Katherine Barton, MBBS (Hons) FRACP, Paediatric Emergency Physician, Princess Margaret Hospital, Perth, Australia

Tom Beattie, MB BAO BCh MSc FRCPE FRCSE FFSEM RCS&PI, Co-director MSc Paediatric Emergency Medicine, Child Life & Health, University of Edinburgh, Edinburgh, UK

Nader Beshay, MMed MBBS, Registrar, Emergency Medicine, Royal North Shore Hospital, Sydney, Australia

Robyn Brady, FRACP FACEM LCCH CCPU
Paediatric Emergency Physician and Ultrasound Lead

Lady Cilento Children's Hospital, Brisbane, Australia

Adrian Mark Bonsall, MA MBBS, Paediatric Emergency Hospitalist, Emergency Department, Children's Hospital at Westmead, Sydney, Australia

Christine Brabyn, BSc MBChB FRACP FACEM
Emergency Physician, Paediatric Emergency Physician, Waikato Emergency Department, Hamilton, New Zealand

Drago Bratkovic, MBBS FRACP, Unit Head, Metabolic Clinic, Women's and Children's Hospital, North Adelaide, Australia

Simon G.A. Brown, PhD FACEM
Director, Aeromedical and Retrieval (AMR), Ambulance Tasmania
Emergency Physician, Royal Hobart Hospital, Hobart, Tasmania
Clinical Professor, Emergency Medicine, University of Western Australia, Hobart, Australia

David H.F. Buckley, BSc MBChB FANZCA FCICM, Paediatric Anaesthesia and Intensive Care, Starship Children's Health, Auckland, New Zealand

Jim P. Buttery, MBBS FRACP MSc MD
Head, Infection and Immunity, Monash Children's Hospital
Professor of Paediatric Epidemiology, Epidemiology and Preventive Medicine & Paediatrics, Monash University, Melbourne, Australia

Adam Bystrzycki, MBBS FACEM PGDipEcho
Emergency Physician, Emergency & Trauma Centre, Alfred Hospital
Senior Lecturer, Department of Medicine, Monash University, Sandringham, Australia

Gar Ming Chan, MD FACEM, Specialist, Emergency Medicine Calvary Hospital, Lenah Valley, Tasmania, Australia

Annette Chang, MBBS MMed Surgery, Registrar, Paediatric Surgery Department, Monash Children's Hospital, Melbourne, Australia

John A. Cheek, MBBS FACEM, Deputy Director, Performance and Improvement, Emergency Department, The Royal Children's Hospital, Melbourne, Australia

Nicholas Cheng, MBBS BSc(Med) DCH FRACP

Paediatric Emergency Consultant, Emergency Department, Children's Hospital at Westmead
Conjoint Lecturer, Faculty of Medicine, University of NSW, Conjoint Lecturer, Paediatrics, Western Sydney University, Westmead, Australia

Robin Choong, MBBS FRACP FCICM, Senior Staff Specialist, Paediatric Intensive Care, The Children's Hospital at Westmead, Sydney, Australia

Jane Cocks, MBBS DCH FRACP FACEM FCICM, PGCert Aeromedical Retrieval, Paediatric Emergency Physician, Paediatric Emergency Department, The Women's and Children's Hospital, North Adelaide, Australia

Elizabeth Cotterell, MBBS MPH FRACP, Associate Professor of Paediatrics, School of Rural Medicine, University of New England, Armidale and Armidale Rural Referral Hospital, Armidale, Australia

John Craven, BSc (Hons) BMBS FRACP FACEM
Head of Unit, SAAS MedSTAR Kids Retrieval Service
Paediatric Emergency Physician, Flinders Medical Centre & Women's and Children's Hospital, Adelaide, Australia

Nigel W. Crawford, MBBS MPH PhD FRACP, General Paediatrician, The Royal Children's Hospital and the Murdoch Children's Research Institute, Melbourne, Australia

John Cronin, MB BAO BCh AFRCSI DCH FRCEM, Consultant in Emergency Medicine, Emergency Department, St Vincent's University Hospital, Dublin, Ireland

Sarah Davidson, MBBS FACEM PEM, Paediatric Emergency Medicine Physician, The Sunshine Coast University Hospital and Health District, Queensland, Australia

Tessa Davis, MBChB BSc(Hons) MA MRPCH FRACP, Co-Founder, Clinical Fellow in Paediatric Emergency Medicine, Royal London Hospital, Don't Forget The Bubbles, London, UK

Anthony Delaney, MBBS MSc PhD FACEM FCICM
Senior Staff Specialist, Malcolm Fisher Department of Intensive Care Medicine, Royal North Shore Hospital
Associate Professor, Sydney Medical School, University of Sydney
Adjunct Senior Research Fellow, School of Public Health and Preventative

Medicine, Monash University, St. Leonards, Australia

Peter Downie, MD FRACP
Head, Paediatric Haematology-Oncology, Monash Children's Hospital
Director, Centre for Cancer and Blood Diseases, Monash Children's Hospital
Senior Lecturer, Department of Paediatrics, Monash University, Clayton, Australia

Evelyn Doyle, MB Bch BAO MRCP FRACP, Consultant Paediatric Emergency Physician, Paediatric Emergency Department, Women's and Children's Hospital, Adelaide, Australia

Linas Dziukas, MBBS MD FRACP FACEM, Emergency Physician, Emergency and Trauma Centre, Alfred Hospital, Melbourne, Australia

Daryl Efron, MBBS FRACP MD
Consultant Paediatrician, General Medicine, Royal Children's Hospital
Senior Research Fellow, Murdoch Children's Research Institute
Senior Lecturer, Paediatrics, University of Melbourne, Parkville, Australia

Tom Everitt, MBBS FRACGP, General Practitioner, Cranbourne, Victoria, Australia

Michael Fairley, MBBS FRANZCP
Consultant Child Psychiatrist, Child Protection Unit, Sydney Children's Hospital
Child and Adolescent Psychiatrist, Youth and Family Mental Health, Lismore Base Hospital, Strathfield, Australia

Bruce Fasher, MBBS DCH (RCP) DRCOG (Lond) FRCP (Lond) FRACP, Physician, Emergency Department, Royal Alexandra Hospital for Children, Sydney, Australia

Dominic Fleischer, MBChB FACEM, Specialist Emergency Physician, Emergency Department, Christchurch Hospital, Christchurch, New Zealand

Jeremy Furyk, MBBS FACEM MSc MPH&TM
Emergency Physician, University Hospital Geelong, Victoria, Australia
Associate Professor (Adjunct), James Cook University, Townsville, Australia

Lalith Gamage, MMBS DCH DCH MD(PAEDS) FRACP
Staff Specialist, Emergency Department, Women's and Children's Hospital
Staff Specialist, Paediatric Department, Port Augusta Hospital
Lecturer, Rural School of Medicine, University of Adelaide, Auldana, Australia

Gary Geelhoed, MBBS FRACP FACEM MD, Clinical Professor, Chief Medical Officer Western Australia, University of Western Australia, Perth Australia

Ed Giles, MBBS FRACP MRCPCH PhD
Consultant Paediatric Gastroenterology, Paediatrics, Monash Children's Hospital
Research Fellow, Paediatrics, Monash University, Clayton, Australia

Laura Graley, BMBS MRCPCH, Melbourne, Australia

Joanne Grindlay, MBBS FACEM FRACGP FARGP DA(UK) Grad Dip(Rural GP) EMDM HOSM
Deputy Director, Emergency Medicine, The Royal Children's Hospital
Clinical Associate Professor, University of Melbourne, Department of Paediatrics
Research Associate, Murdoch Children's Research, Melbourne, Australia

Sonia R. Grover, MBBS FRANZCOG MD FFPMANZCA
Director, Gynaecology Department, Royal Children's Hospital
Clinical Professor, Dept Paediatrics, University of Melbourne
Head of Family Planning Unit, Austin Health
Head of Gynaecology Unit, Mercy Hospital for Women, Austin Health, Clifton Hill, Australia

Naren Gunja, MBBS MSc FACEM FACMT
Clinical & Forensic Toxicologist, Westmead Hospital
Clinical Toxicologist, NSW Poisons Information Centre
Clinical Associate Professor, Discipline of Emergency Medicine, Sydney Medical School, Australia

Wayne Hazell, MBBS DipObs GCCT MCLINED FACEM, Emergency Physician, The Prince Charles Hospital Emergency Department, Deputy Head of the Prince Charles Clinical School, University of Queensland, Brisbane, Australia

Malcolm Higgins, BM BS FRACP, Medical Unit Head, Paediatric Emergency Department, Women's and Children's Hospital, Adelaide, Australia

Rupert Hinds, MBBS MRCP MRCPCH FRACP, Consultant Gastroenterologist and Head of Department of Gastroenterology at Monash Children's Hospital and Senior Lecturer in Paediatrics at Monash University, Clayton, Australia

Andrew John Anderson Holland, BSc (Hons) MBBS PhD Grad Cert Ed Studies FRCS (Eng) FRACS (Paed) FACS
Professor of Paediatric Surgery, Discipline of Paediatrics and Child Health, The University of Sydney
Senior Clinical Academic, Douglas Cohen Department of Paediatric Surgery, The Children's Hospital at Westmead, Westmead, Australia

Jason Hort, MBBS MRCP FRACP
Staff Specialist, Emergency, The Children's Hospital at Westmead
Clinical Senior Lecturer, Emergency, Sydney Medical School, Westmead, Australia

Sarah Jamison, BHB MBChB FRACP, Paediatric Emergency Specialist, Children's Emergency Department, Starship Children's Hospital, Auckland, New Zealand

Shefali Jani, FRACP(PEM) MD(Paed) MClinEpid MBBS
Paediatric Emergency Physician, Children's Hospital at Westmead
Clinical Associate lecturer, University of Sydney, Sydney, Australia

Sheena Kaul, MBBS, Paediatric Registrar, Monash Children's Hospital, Clayton, Australia

Colin S. Kikiros, MBBS FRACS, Senior Consultant Surgeon, Paediatric Surgery, Perth Children's Hospital, Perth, Australia

Erica Kreismann, MD FACEM, Director of Emergency Medicine, Calvary Hospital, Lenah Valley, Tasmania, Australia

David M. Krieser, MBBS FRACP
Paediatric Emergency Physician, Emergency Department, Sunshine Hospital
Clinical Associate Professor, Department of Paediatrics, University of Melbourne
Honorary Fellow, Murdoch Children's Research Institute
Honorary Fellow, Joseph Epstein Centre for Emergency Medicine Research, St Albans, Australia

Ben Lawton, MBChB BSc(Hons) FRACP(PEM), Paediatric Emergency Physician, Logan Hospital, Lady Cilento Children's Hospital and the University of Queensland, Brisbane, Australia

Richard Lennon, MBBS FRACP(Paed) FACEM MBioeth, Cherrybrook,

Australia

Damir Ljuhar, BBioMed MBBS MPHTM, Paediatric Surgery Registrar, Monash Children's Hospital, Melbourne, Australia

Elliot Long, FRACP
Consultant, Emergency Medicine, The Royal Children's Hospital
Research Fellow, Critical Care, Murdoch Children's Research Institute
PhD Candidate, Paediatrics, University of Melbourne, Parkville, Australia

Niki Talić, MBBS FACEM-PEM, Dr, Paediatric Emergency Consultant, Royal Children's Hospital, Parkville Australia and Monash Children's Hospital, Clayton, Australia

Jeanette Marchant, MBChB DCH MRCPCH(UK) FRACP, Specialist, Paediatric Emergency and Retrieval, The Children's Hospital, Westmead and NSW Newborn and Paediatric Transport Service, Sydney, Australia

Susan Marks, MD M Forensic Medicine FRACP FRCPA, Staff Specialist, Child Protection Unit, Children's Hospital at Westmead, Westmead, Australia

Jennie Martin, MBBS (Hons) DipCH FACEM, Emergency Physician, Emergency, Royal North Shore Hospital, St Leonards NSW, Australia

Sarah Martin, MBBS MSc DTM&H FRACP FACEM, Paediatric Emergency Physician, Emergency Department, Lady Cilento Children's Hospital, Brisbane, Australia

Mary Elinor McCaskill, MBBS BSc(med) DipPaeds MBA FACEM, Paediatric Emergency Physician, Emergency Department, The Children's Hospital at Westmead, Sydney, Australia

Dermot Thomas McDowell, MB BAO BCh MRCS, Dr, Department of Paediatric Surgery, Children's Hospital at Westmead, Westmead, Australia

Shona McIntyre, MbChB FACEM, Emergency Physician, Emergency Department, Sunshine Hospital, Melbourne, Australia

Damien McKay, MBBS BAppSc (Physio) MSpMed FRACP
Staff Specialist Paediatrician, Children's Hospital at Westmead
Paediatric Rheumatologist, Liverpool Hospital, Sydney, Australia

Ben McKenzie, MBBS FACEM AFRACMA
Emergency Physician, Bendigo Health, Victoria, Australia

Critical Care Coordinator and Retrieval Physician, ARV, Ambulance Victoria
Adjunct Senior Lecturer, Monash University, Victoria, Australia

Christopher McKinlay, MBChB PhD DipProfEthics FRACP CCPU
Senior Lecturer, Liggins Institute and Department of Paediatrics, Child and Youth Health, University of Auckland
Neonatologist, Kidz First Neonatal Care, Counties Manukau Health, Auckland, New Zealand

Robert Melvin, FACEM DTMH MBBS MA(Cantab) BA Hons, Deputy Director of Emergency Medicine, Clinical Lead for Paediatric Emergency Medicine, Emergency Physician, Alfred Health, Melbourne, Australia

Alastair D. McR. Meyer, BSc(Hons) BMedSci MBBS FACEM FRCEM(Lond) FRCP(Edin) FRACGP
Director of Emergency Medicine, Emergency Department, Casey Hospital
Adjunct Associate Professor, Medicine, Nursing and Health Sciences, Monash University, Melbourne, Australia

Erin Mills, MBBS (Hons) BMedSci FRACP PEM, Paediatric Emergency Physician, Paediatric Emergency Department, Monash Medical Centre, Clayton, Australia

Yuresh Naidoo, MbChb FACEM GradCert HPE(UWA), Emergency Medicine Specialist, Director of Clinical Training, Clinical Lead Service Improvement Unit, Joondalup Health Campus, Western Australia

Murali Narayanan, FRACP DNB (Ped) DCH MBBS, Consultant Paediatrician, Emergency Department, Fiona Stanley Hospital, Murdoch, Australia

Ramesh Nataraja, BSc(Hons) MBBS FRCSEd(PaedsSurg) GCCS (Hons) FFSTEd FRACS(Paeds)
Consultant Paediatric Surgeon, Director of Simulation, Monash Children's Hospital
Senior Lecturer, Paediatric, Monash University, Melbourne, Australia

Jocelyn Neutze, MBChB FRACP FACEM, Paediatric Emergency Specialist, Kidz First, Middlemore Hospital, Auckland, New Zealand

Kenneth Patrick Nunn, MBBS (Hons) PhD FRACPsych FRANZCP, Senior Paediatric Neuropsychiatrist, Psychological Medicine,

Children's Hospital Westmead, Westmead, Australia

Felix Oberender, MRCPCH PhD FCICM
Director, Paediatric Intensive Care Unit, Monash Children's Hospital
Honorary Consultant, Paediatric Intensive Care Unit, The Royal Children's Hospital
Adjunct Senior Lecturer, Monash University, Melbourne, Australia

Adam O'Brien, MBBS DDU FACEM, Emergency Physician, The Royal Children's Hospital, Melbourne, Australia

Fenton O'Leary, MBBS FACEM, Clinical Associate Professor Emergency Medicine, The Children's Hospital at Westmead and University of Sydney, Sydney, Australia

Kim Lian Ong, MBBS FRCS (Edin) FHKCEM FHKAM (Emergency Medicine) FAMS
Consultant, Accident and Emergency Department, Pok Oi Hospital
Honorary Clinical Associate Professor, Accident and Emergency Medicine Academic Unit, Chinese University of Hong Kong, Hong Kong SAR, China

David Orchard, MBBS FACD, Director of Dermatology and Associate Professor, Royal Children's Hospital and Melbourne University, Melbourne, Australia

Michael Osborn, MBBS FRACP FRCPA, Haematologist/Paediatric, Adolescent and Young Adult Oncologist, Women's and Children's Hospital and Royal Adelaide Hospital, Adelaide, Australia

Colin Parker, MBChB DCH(Lon) MRCPCH FACEM
Deputy Director, Emergency Medicine, Joondalup Health Campus
Emergency Physician, Perth Children's Hospital
Clinical Senior Lecturer, Emergency Medicine, University of Western Australia, Perth, Australia

Jacqueline E.L. Parkinson, BPharm (Hons) MPharm GradCert HlthServManagement BCGP MSHP, Senior Pharmacist, Pharmacy Department, Monash Health, Melbourne, Australia

Sameer A. Pathan, MBBS CABEM MRCEM
EM Consultant, Emergency Department, Hamad General Hospital, Hamad Medical Corporation

Department of Epidemiology & Preventive Medicine, Monash University, Mesaieed, Qatar

Scott Pearson, MB ChB FACEM, Emergency Physician, Emergency, Christchurch Hospital, Christchurch, New Zealand

Ioannis Pegiazoglou, Dr. med.
Deputy Director, Interdisciplinary Pediatric Emergency Department, University Children's Hospital Basel (UKBB) Basel, Switzerland
Clinical Associate Lecturer, Medical School, University of Sydney, Sydney, Australia

Roderic Phillips, MBBS FRACP PhD, Clinical Associate Professor, Paediatric Dermatology, Royal Children's Hospital and Monash University, Melbourne, Australia

Susan Phin, MBBS FRACP, Paediatric Emergency Physician, Emergency Department, The Children's Hospital at Westmead, Westmead, Australia

Colin Victor Eric Powell, MB ChB FRACP FRCPCH MRCP(UK) MD, Consultant Paediatrician Noah's Ark Children's Hospital for Wales Cardiff and Reader in Paediatrics and Deputy Dean for Research, Division of Population Medicine, School of Medicine, Cardiff University, Wales, United Kingdom

Harsh Priyadarshi, MS, Complex Spine Fellow, Macquarie Neurosurgery, Associate Lecturer, Macquarie University, Sydney, Australia

Diana Purvis, MBChB FRACP, Paediatric Dermatologist, Paediatric Dermatology, Starship Children's Hospital, Auckland, New Zealand

Karen Quay, BHB MBChB FACEM, Paediatric Emergency Specialist, Childrens Emergency Department, Starship Childrens Hospital, Auckland, New Zealand

Kottayam Radhakrishnan, MBBS FRACP FRCPA, Consultant Paediatric Haematologist, Children's Cancer Centre, Monash Children's Hospital, Melbourne, Australia

Damayanthi Rasanathan, FRACP, Paediatric Emergency Specialist, Children's Emergency Department, Starship Hospital, Auckland, New Zealand

Meenakshi Rattan, MBBS MD (Paediatrics) DCH(UniSyd) FRACP MPH

MHM, Consultant Paediatrician, Wyong and Campbelltown Hospital; Associate Clinical Lecturer, University of Sydney, Wahroonga, Australia

Felix Regenfelder, MD
Dr, Orthopaedics, The Children's Hospital at Westmead, Sydney, Australia
Dr, Orthopaedie und Unfallchirurgie, Buergerspital Solothurn, Solothurn, Switzerland

Sasha Rossaye, MBBCh BSc FRACP, Paediatric Emergency Specialist, Paediatric Emergency Department, Women's and Children's Hospital, Adelaide, Australia

John M. Ryan, FCEM FRCSEd(A&E) FFSEM DipSportsMed DCH, Professor, Emergency Department, St Vincent's University Hospital, Dublin, Ireland

Vered Schildkraut, BSc MD FRACP
Consultant Paediatric Gastroenterologist, Paediatric Gastroenterology Children's Program, Monash Children's Hospital
Consultant Paediatric Gastroenterologist, Gastroenterology and Clinical Nutrition, The Royal Children's Hospital, Melbourne, Australia

Scott Schofield, MBBS FRACP, Paediatric Emergency Physician, Sunshine Coast Hospital and Health Service, Queensland, Australia

Robert Seith, MBChB MRCPCH FRACP, Paediatric Emergency Consultant, Director of Emergency Medicine Training, Monash Children's Hospital/Monash Medical Centre, Melbourne, Australia

Michael Shepherd, MBChB FRACP MPH, Paediatric Emergency Specialist, Children's ED, Starship Hospital, Auckland, New Zealand

James Tibballs, BMedSci (Hons) MBBS MEd MBA MHlth&MedLaw MD FCICM FACLM DALF
Associate Professor, Departments of Paediatrics and Pharmacology, The University of Melbourne
Senior Intensivist, Paediatric Intensive Care Unit, Royal Children's Hospital Melbourne, Melbourne, Australia

Loren Sher, MBBCh DCH FACEM
Paediatric Emergency Physician, Head of Paediatric Emergency, The Northern Hospital

Honorary Clinical Lecturer, The University of Melbourne, Melbourne, Australia

Kam King Sinn, MBBS FRACP, Emergency Paediatrician, Senior Specialist, Emergency Department, The Canberra Hospital, Canberra, Australia

Holly Smith, MDCM FRACP, Paediatric Emergency Specialist, Emergency Department, Royal North Shore Hospital, St Leonards, Sydney, Australia

Soundappan S.V. Soundappan, MBBS MS(Gen Surg) MCh(Paed Surg) FRACS(Paeds)
Head of Trauma, The Children's Hospital at Westmead
Senior Lecturer, University of Sydney, Sydney, Australia

Mike Starr, MBBS FRACP, Paediatrician, Infectious Diseases Physician, Consultant in Emergency Medicine, Director of Paediatric Education, Honorary Clinical Associate Professor, University of Melbourne, The Royal Children's Hospital Melbourne, Melbourne, Australia

Greg Stevens, BHB MBChB FACEM, Specialist, Emergency Medicine, Waikato Hospital, Hamilton, New Zealand

Emma Tavender, BSc (Hons) MA PhD
Knowledge Translation Co-ordinator, Emergency Research Group, Murdoch Children's Research Institute
Honorary Fellow, Department of Paediatrics, University of Melbourne, Melbourne, Australia

James Tilleard, MBBS FACEM MEd, Senior Staff Specialist, Emergency Department, Sunshine Coast Hospital and Health Service, QLD, Australia

Joseph Ting, MBBS BMedSc MSc (Lond) PGDipEpi DipLSTHM FACEM
Senior Staff Specialist, Emergency Medicine, Mater Hospital Brisbane
Senior Staff Specialist, Emergency Medicine, Ipswich General Hospital
Adj Associate Prof, School of Public Health & Social Work, Queensland University of Technology
Clinical Senior Lecturer, School of Anaesthesiology & Critical Care, University of Queensland, Australia

Henry Patrick John Walsh, MBchB MCNorth FRCS FRACS, Associate Professor, Department of Paediatric Orthopaedics, Lady Cilento Hospital and University of Queensland, Brisbane, Australia

Donald Payne, MD MBBChir, Adolescent Medicine, Princess Margaret

Hospital, Subiaco, Western Australia

Rachel H. Webb, MBChB MPH&TM FRACP
Senior Lecturer in Paediatrics, Paediatrics, University of Auckland
Paediatrician, Kidz First, Counties Manukau Health
Paediatric Infectious Diseases Specialist, Starship Children's Hospital, Auckland, New Zealand

Christopher Webber, MBBS Dip RACOG FRACP
Head of Department & Paediatric Emergency Physician, Emergency Department, Sydney Children's Hospital
Consultant, Neoanatal & Paediatric Retrieval, NETS New South Wales, Randwick, Australia

Adam West, MBBS DRANZCOG FACEM
Director Paediatric Emergency Medicine, Monash Emergency, Monash Health
Adjunct Lecturer, Department of Medicine, Monash University, Clayton, Australia

Julian White, MBBS MD FACTM, Professor & Department Head, Toxinology Department, Women's & Children's Hospital, North Adelaide, Australia

Gary David Williams, MBBS FRACP, Paediatric Intensivist, Paediatric ICU, Sydney Children's Hospital, Randwick, Australia

Simon Vincent Wood, MBBS(Syd) DipPaed(NSW) FACEM, Emergency Physician, Joondalup Health Campus, Joondalup, Australia

Kim Yates, MBChB MMedSci PGDipClinEd FACEM, Emergency Medicine Specialist, Emergency Department, North Shore & Waitakere Hospitals, Auckland, New Zealand

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SECTION 1

Approach to the Paediatric Patient

OUTLINE

1.1

Approach to the paediatric patient

Adam West, and Tom Everitt

ESSENTIALS

1 Gaining rapport with the child and the confidence of the parents is the key to assessing children. Never underestimate the power of distraction and entertainment.

2 A child needs to be approached according to chronological and developmental age.

3 Observation is a vital diagnostic tool, which is vastly more important in children than in adult patients.

4 The need for investigations is a balance between an invasive stress on a child and the potential gain of information to aid decision making.

5 Always back up discharge with a concrete action plan and definitive follow-up.

6 It is often more important to exclude serious illnesses than make a definitive diagnosis. This may be more easily achieved with timely review.

7 Addressing parental concerns is an important part of the therapeutic process.

8 Emergency physicians should stay within their comfort zone and when in doubt, consult.

9 A febrile child should be considered as potentially sick until one can confidently conclude that he/she is well following a thorough assessment and period of observation.

10 Always reflect on the potential fears of the child and parents.

Introduction

Who sees paediatric emergencies?

It is essential that all doctors are familiar with the recognition and management of the seriously ill child. The majority of children presenting to emergency departments (EDs) are taken to mixed departments that see both adults and children, while a number present to tertiary paediatric centres. Occasionally, children will arrive, due to close proximity in an emergency, at adult departments where staff may be less familiar with their management. Likewise, paediatric emergencies occur remote from EDs and may require stabilisation by general practitioners, paramedical staff or laypersons prior to subsequent referral. It is an important role of EDs to be an available resource to support the community in the management of paediatric emergencies. This function may occur through liaison, education, and the provision of advice.

Some critically ill children will arrive in a more predictable fashion via ambulance, and some preparation can occur to plan for their initial treatment. On the other hand, a child in extremis may arrive unannounced, rushed in from a family car. Systems of preparedness for these situations are critical for the immediate assessment and optimal early management of children by ED staff (see Section 2).

Identifying the potentially sick child

Of the vast number of children attending EDs, approximately 2–5% are classified as immediate emergencies (Australasian Triage Scale (ATS) 1 and 2) that require urgent assessment and management.[1] Importantly, while children can present with less urgent triage categories they may rapidly deteriorate from evolving sepsis or airway compromise. The majority of paediatric presentations consist of less emergent problems involving a wide spectrum of injuries and illness. Of this group, there is a subset where the diagnosis is not immediately apparent.

Thus, paediatric patients can generally be divided into three broad groups: the obviously well, the obviously sick, or the potentially sick child. One of the major

tasks for the emergency physician is to identify the ‘sick child’ from a large, undifferentiated group of children who may present as potentially sick. It is by a ‘filtering process’ via history, observation, examination, appropriate investigation and consultation that one identifies the *potentially* sick child (Fig. 1.1.1).

This group of patients includes: those children who have progressed to a severe form of a usually benign illness; those with early, subtle signs of a serious disease; or those who on initial assessment appear unwell but require observation or investigation to help rule out serious disease. It is often through *observation* of a child that one is able to more accurately assess each of these possibilities.[2] With experience, the ability to appreciate a ‘sick child’ improves. Serial observation and considered investigation can be of use in identifying children with life-threatening conditions. Ongoing uncertainty of the underlying problem may provide justification for admission for a more prolonged period of observation. If a life-threatening problem like sepsis is considered, treatment should be initiated after collection of appropriate pathology samples where this can be done in a suitable time frame.

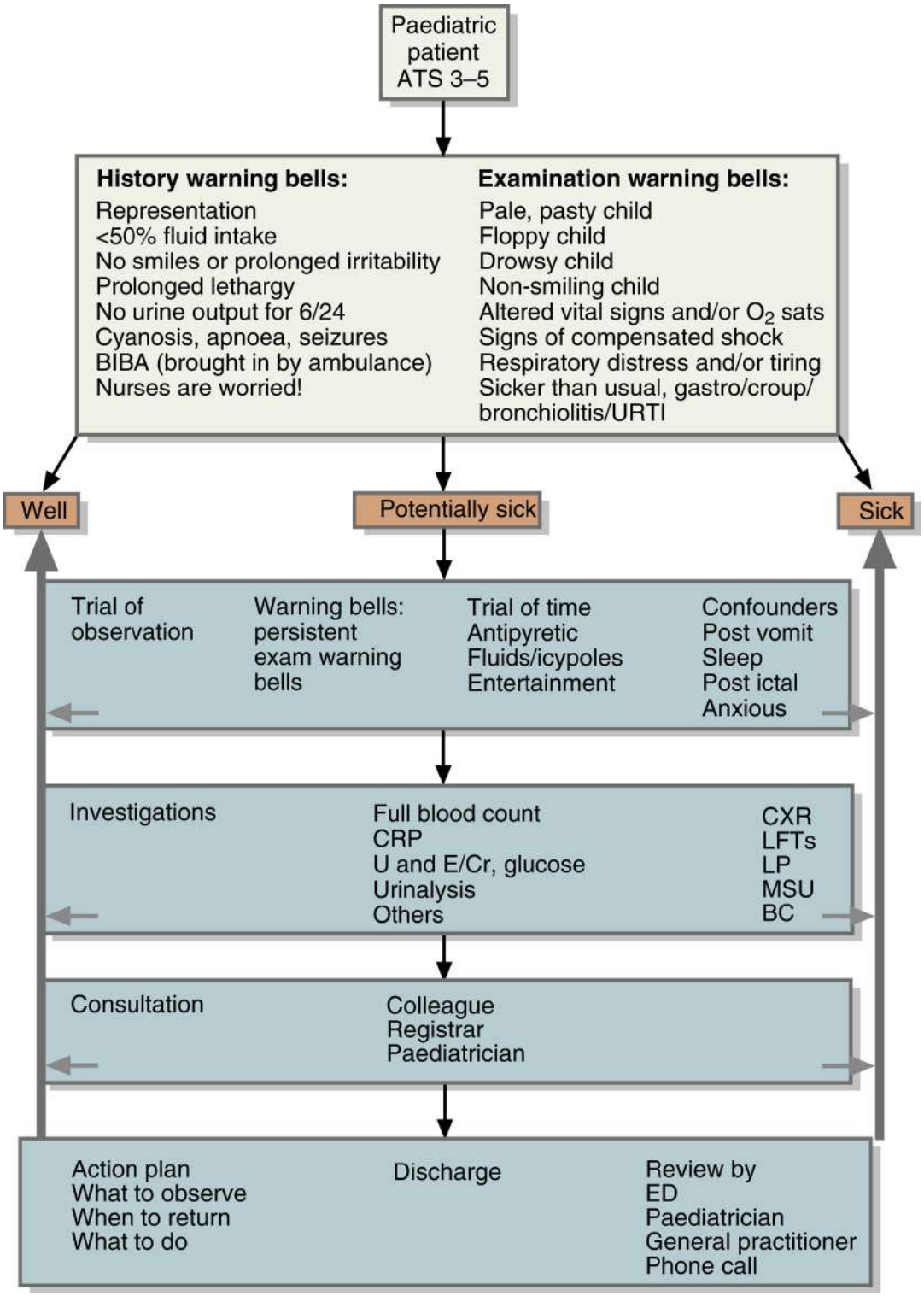

FIG. 1.1.1 Algorithm of paediatric decision making.

Children with fever

The concept of ‘occult bacteraemia’ (OB) highlights the difficulties in detecting significant illness in febrile young children. With the introduction of widespread vaccination to the common agents of OB (*HiB*, *Pneumococcus*) the prevalence of paediatric sepsis has diminished significantly, and the clinical experience of managing septic children has been diluted in developed countries. Hence, one needs to have a planned approach to the assessment of febrile children at various ages.

Bacteraemia in its most obvious form presents as a febrile, pale, pasty, mottled child, centrally warm but with cool peripheries. Some young children with bacteraemia, however, can appear completely well apart from fever. Investigations *may* demonstrate a high white cell count or elevated C-reactive protein (CRP), but these inflammatory markers are unfortunately often non-discriminatory between benign and serious causes.

The problem is not so much that children with OB are sick at the time of initial assessment, but the possibility of the later development of serious bacterial sequelae necessitates timely treatment. However, many bacteraemic children will spontaneously clear the organism without therapy. Therefore, these children remain in the potentially sick category of patients and should have either admission for observation or discharge with frequent planned reviews for sequelae and a definitive action plan for their parents should the condition change.

Evolving illness in children

Due to differences in anatomy, physiology, development and psychology, children’s diseases are age-specific, with serious illness often taking time to evolve.[3] Many children present to an ED in the early stage of an illness, and making a definitive diagnosis may require time and repeated review. The clinical status of paediatric patients may also change rapidly as they can compensate remarkably well during serious illness. Deterioration can occur in response to prior trauma, evolving sepsis, toxin absorption or a seizure and necessitate a change in the initial priority to receive treatment.

The younger the child, the greater the potential for rapid deterioration as the early manifestations of a serious illness may be subtle and non-specific. For these reasons clinicians must be vigilant for the early signs of compensated shock such as tachycardia, decreased capillary refill, mottled skin, cool peripheries, decreased urine output, or drowsiness. Early detection and fluid

resuscitation at this point may prevent hypotension in a child with evolving sepsis. Children with severe and deteriorating respiratory illness will manifest fatigue. It is the early recognition of children with serious illness or the potential to deteriorate that is critical to the timely initiation of effective treatment.[2]

An important principle in emergency paediatrics is to be proactive. One must be aware of the importance of regularly reviewing a child's response to a given therapy, escalate treatment if required and be vigilant for subtle signs of deterioration.

The environment

The physical environment of the ED needs to be child and family friendly with appropriately equipped cubicles for the reception of children accompanied by their carers. Despite the noise inherent in a busy department of sick children, the environment should be as calm and relaxed as possible. Ideally, the care of children is managed in an area separate to adult patients thus shielding children from what can at times be a confronting environment in the emergency department.

Wall or ceiling posters, mobiles, and a selection of toys and books are useful to distract younger children from the distress and threat of an unfamiliar hospital environment. Familiar characters from current movies and television shows can provide distraction as well as facilitating central nervous system (CNS) assessment through recognition.

A few initial moments gaining a child's confidence with a toy or bubbles will usually reward the clinician with a more rapid and thorough assessment of the reluctant child. Stickers or bravery certificates are excellent rewards to have on hand for young frightened children who have undergone imaging or blood tests.

Even in a mixed department children should be completely separated from adult patients. A separate waiting area set up for children and families is also highly recommended. Adult patients who are behaviourally disturbed, severely ill, or covered in blood will be distressing for a child and family to see or hear in a nearby cubicle. Departments should be designed such that this is a very unlikely scenario.

Likewise, if a child is to undergo a procedure during which he/she may become distressed, such as intravenous insertion or laceration repair, it is best performed in a closed dedicated procedure room. This will avoid visual or auditory distress to other children and parents. A mounted television/video

monitor in this setting can be an excellent distraction during procedures, as an adjunct to analgesia and sedation. If available, play therapists provide an invaluable asset to assist explaining procedures in an age-appropriate manner to children and parents as well as assisting in distraction during the procedure. Comfort positioning with a parent is preferable to the former practice of wrapping children in a sheet for the procedure. For neonates and small infants, a radiant heater over the examination bed will aid temperature stability, examination and often the discovery of veins for cannulation.

The paediatric resuscitation area should include wall charts, which refer to emergency algorithms and drug dose guidelines, which can be rapidly referred to during the resuscitation of critically ill children. Although white boards may be useful for pre-sizing and dose calculations for the imminent arrival of a sick child, this is prone to error, particularly during a stressful paediatric resuscitation. Where possible, resources which have pre-calculated doses and equipment sizes should be available to clinicians. These include the Broselow tape, medication books, software programs or smartphone apps. Hospitals should have agreed and readily accessible weight-based protocols on preparation and administration of emergency medications and infusions for children.

Updated electronic and hard-copy clinical guidelines for the management of common paediatric emergency conditions are useful. State or national guidelines and pathways can ensure consistency of management from all levels of clinical staff, as well as improving continuity of care in children who require admission to an inpatient unit.

Patient/parent information resources such as handouts, leaflets and information videos should also be available. They should cover a range of common paediatric conditions and ideally be translated into languages which reflect the demographics of the hospital’s local community.

Triage

Paediatric patients arriving in the ED undergo triage according to the standardised Australasian Triage Scale (ATS 1–5) or equivalent so that they are seen in a prioritised fashion according to urgency. In mixed EDs where triage nurses may have had less paediatric experience, there has been a tendency to up-triage paediatric patients.[1] The use of scoring systems for specific conditions or a Triage Observation Tool may be helpful in improving the reliability of triage in young children, who may present with non-specific symptomatology.[3]

Children with life-threatening conditions may not be easily recognised during brief triage assessments. A secondary nursing assessment should occur as early as possible, with further observations performed at the bedside, so that any change in condition can be detected early and acted on promptly. The senior doctor in the department should immediately be informed of children triaged as ATS 1 or 2 to direct timely management. The senior doctor and nurse should also be informed of any child with vital signs that are in the range to otherwise trigger a medical emergency team (MET) call. In times of high workload, children with an ATS 3 may not be definitively assessed within 30 minutes and should have a senior doctor or nurse rapidly assess status and initiate therapy, if required. It may be necessary to modify normal triage systems when ED numbers are affected by surges in demand such as during significant influenza outbreaks or similar events.

Front loading care

Some initiation of treatment is appropriate during the triage process or soon after. This can include the provision of analgesia for pain or an antipyretic in a miserable febrile child. It is important that all patients with pain are given early and appropriate analgesia or have injuries splinted when required. This will facilitate a more comfortable, reliable and expeditious assessment. The use of opiates, when required, will only enhance, rather than detract from, the subsequent physician's physical examination.[4] The use of visual analogue scales such as the Wong–Baker faces[5] may assist the assessment of a child's response to analgesia as may asking the parents if their child is in pain. A process of fast tracking appropriate children with isolated limb injuries for an X-ray prior to definitive medical review may improve efficiency through the department.

Other interventions which may be instituted early in a child's ED visit include: topical anaesthesia for open wounds, topical anaesthetic creams for those likely to require intravenous access or venipuncture, oral ondansetron for those who are actively vomiting, oral rehydration fluids or icypoles for gastroenteritis, corticosteroids for croup, commencement of bronchodilators for asthma, and provision of relevant information leaflets.

Febrile children who present with a rash, not clearly due to a viral exanthema or benign phenomena, should be fast tracked to be seen by a senior doctor to consider the possibility of meningococcaemia.

It is useful to have documented management plans for children who may

repeatedly present to the department. This includes children with complex healthcare needs, brittle asthma, cyclical vomiting, metabolic conditions or recalcitrant seizures where a clear plan of management can expedite care by ED staff. These management plans are often developed as a result of cooperative efforts between the ED and the child's usual specialists.

The paediatric approach

The evaluation process of a child in the ED involves history, observation, examination and sometimes investigations.

Each of these components needs to be considered in the formulation of a diagnosis and disposition plan. A child needs to be considered in the context of the family. The assessment of children in the ED setting can be both challenging and very rewarding. It is a challenge to modify the clinical approach according to the chronological and developmental level of the individual child. Likewise, treating paediatric patients is a rewarding area of emergency medicine, as children will often respond rapidly to management within the time frame of the ED attendance.

Gaining rapport

Efforts to gain initial rapport with a child and the confidence of the parents are the key to assessing children in the ED setting. An unrushed, gentle and caring manner will rapidly settle the fears and anxieties of most children and their parents. Playing simple games with a child can often provide useful information as well as building trust and rapport. This usually allows the examination to proceed in a non-threatening fashion and improves the reliability of clinical signs. It will take time, experience and the observation of colleagues' techniques for every emergency physician to develop his/her own individual approach to children. A thorough examination without causing distress to a child is very reassuring to a parent. Many children arrive at an ED miserable, in pain, fearful, or with some trepidation of what lies ahead. With a child-friendly approach by all staff, most will leave feeling much better and, hopefully, even having enjoyed the experience.

Age appropriate

The approach to any child in the ED is dictated by the child's age and developmental level. It is useful to have a modified approach to suit newborns, infants, toddlers, preschoolers, school-children and adolescents.

An understanding of the range of normal neonatal behaviours, which will often precipitate ED visits, can assist in reassuring new parents that their baby is in fact well (see Chapter 3.1). This understanding will also assist in recognising when behaviour falls outside of the expected normal range and may indicate an underlying problem requiring further evaluation.

A preverbal or developmentally delayed child won't tell you of pain which has shifted to the right iliac fossa. An unwell 14-month-old clinging to his/her mother may actively resist a stranger's initial attempts to examine him/her. The absence of familiarity with a family or child that his/her usual doctor may have may further impede the assessment of anxious children.

When explaining procedures to children it is important to be age appropriate and above all honest. Explain in age-appropriate terms what it may feel like, but also create an environment to suitably reassure and distract the child.[6]

Maintaining a child's trust at all times is crucial and will positively influence any subsequent medical contacts the child may have. The demonstration of a procedure on a doll may decrease the anticipatory trepidation in a child. Additional resources include online videos and procedure-specific handouts. When available, play therapists and experienced nursing staff are excellent sources of information for parents and children prior to procedures.

The assessment of a child should always be carried out in the presence of the parent or carer, unless the child arrives by ambulance or other means without the parent/carer present *and* the child's medical needs warrant immediate attention. Otherwise, it is prudent in the non-urgent situation to provide a staff member to support the child and defer the assessment until carers are present. Non-urgent procedures in younger children are best carried out with the child in a comfortable position (usually the parent's lap), with suitable distraction and good anaesthesia/analgesia.

Developmentally appropriate

Infants particularly benefit from the constant presence of their parent in their visual field in order to avoid stranger distress. It is often easier to examine children in their parent's arms. Neonates can be examined on the ED bed as long as they are kept warm. It is a useful sign of illness or other pathology to note

when young children do not exhibit these normal stranger anxieties. The preschooler who enjoys a sense of play and imagination can usually be relaxed during an examination or procedure by storytelling or engaging in play with a toy. An anxious early school-aged child may respond to participation in the examination or being asked about school or other favoured activities. Adolescents, on the other hand, need to be approached in a more adult fashion and should be offered confidentiality and the opportunity to choose whether their parents are present (see Chapter 30.1).

In the event of an uncooperative child resisting any examination, one may have to modify the approach to gain essential clinical findings in a gentle, sensitive manner. It is unusual, however, for a child to remain 'unexaminable' if appropriate analgesia is given and the child is left undisturbed for a period of time. Often observation will provide as much or more useful information than a traditional examination.

Parental involvement

In order to provide emotional support, parents should be encouraged to remain close to their child during any procedures. Ideally, children can sit on their parent's laps on the bed or trolley during the procedure. Parents who appear at risk of vagal syncope need to be safely positioned in the procedure room. This is another advantage of having them cuddle their children on the trolley where possible. Children's behaviour often mirrors that of their parents, so gaining the confidence of the parent will often make an anxious child relax prior to procedures. The use of a confident, calm, caring approach will be rewarded by a child who will allow a more reliable examination. It is very reassuring to the parents to see that the doctor is experienced and comfortable in dealing with children and anticipates the expected anxieties and reluctance to examination that a child may have when unwell.

History

The initial contact with the family must include an introduction of who you are and your role. The parents should be addressed and the child greeted by name, in an age-appropriate manner. It is important to consider one's approach in terms of the needs of both the child's illness and the parental concerns. The history is generally elicited from the parent or caregiver, but it is appropriate, in a verbal

child, to augment this information by directly questioning the child.

Referral letters and/or ambulance documentation should be reviewed to ensure a complete understanding of the child's visit to the ED. Other useful sources of information include hospital medical records, specialist letters and the child's baby record book which contains details of the child's immunisation, growth and development.

Critically ill child

Sometimes the normal routine of history, followed by examination, will need to be altered in a child who arrives critically ill. The management will need to be expeditious and occur simultaneously with the gathering of pertinent information from the parents. Parents must be given the opportunity to remain at the bedside of their critically ill child undergoing resuscitation, with a capable support person.

Parental issues

The clarity of the history given by parents can be affected by parental distress, anxiety or sleep deprivation. One should begin the history in a focused manner according to the presenting complaint. Later it may be useful to explore individual parental anxieties. One of the most important questions to a parent is 'What is your biggest worry or fear?' Some parents may have specific concerns such as a fear their febrile child has meningococcal disease when community alertness to this condition is heightened. Addressing this concern may occupy most of the doctor's time. The parent of a child who has sustained an accidental scald or injury may be feeling distressed or guilty, and sensitivity to this is required. Again, addressing guilt will involve much of the doctor's time.

Child-specific issues

In younger children, certain symptoms are less specific. The report of vomiting in an infant may be due to meningitis, pneumonia, tonsillitis or urinary sepsis rather than gastroenteritis. The assessment of wellness or otherwise in infants can be more challenging due to their limited psychomotor activities. Indeed, their spectrum of normal behaviours involves sleeping and waking to cry or demand a feed, followed by a return to sleep. Hence, it is important to enquire

into their feeding status and sleep/activity pattern as an indicator of compromise due to illness. One needs to carefully clarify what their current intake is compared to their normal breast- or bottle-feeding. An infant who is feeding less than 50% of normal may have significant compromise. Urine production and the frequency of nappy changes can be used as a rough guide to the adequacy of intake.

Box 1.1.1 History warning bells

- Child taking less than 50% of normal fluids
- The child with prolonged lethargy
- No urine output for 6 hours
- Prolonged irritability or inconsolability
- Report of cyanosis, pallor, seizures or significant apnoea
- The child who has not smiled over a period of hours
- Nursing staff feel the child is 'just not right'
- Unplanned re-presentations
- Parental concerns out of proportion to child's illness
- Brought in by ambulance
- History not compatible with injury/non-accidental injury

It is important to note the report of a young febrile child who remains lethargic and fails to smile or interact with parents. In the otherwise well-looking infant, who appears mottled, clarify with parents whether this may be usual for their child (i.e. physiological cutis marmoratum versus sepsis). In assessing young children with trauma, a thorough history of the timing and mechanism of injury, noting the child's developmental capabilities, is paramount to detecting possible non-accidental injuries (see Chapter 18.2 on Child at Risk). Non-accidental injuries must always be actively considered when assessing a child who has presented with trauma.

Other useful information to cover in the paediatric patient history is shown in Boxes 1.1.1 and 1.1.2.

Examination

Age appropriate

The examination technique used in paediatric patients depends on the age and developmental level of the child. The key is to gain the confidence and then the cooperation of the child. Older children are generally examined in a systematic fashion similar to adult patients. However, younger children usually need to be examined in a less formalised and opportunistic manner, whilst maintaining a high degree of vigilance. The order of the examination frequently needs to be adapted according to the individual child's responses and presenting problem. In a reluctant child, clinical findings may be achieved by surreptitiously examining through play as the opportunity arises. This is often an enjoyable and informative process for the clinician. Much can be ascertained in this situation by careful observation rather than hurrying to complete a clinical examination. Entertaining a young child in a professional manner during examination will generally allow the confounding influence of anxiety to diminish. Potentially significant examination findings are outlined in Box 1.1.3.

Box 1.1.2 Important elements of the paediatric history

Presenting complaint
Pregnancy
Delivery – gestational age, prolonged rupture of membranes, delivery type, APGARS, birth weight, need for resuscitation or special care nursery admission
Development – in a CNS problem, compatibility with injury mechanism
Immunisation status – need to clarify carefully
Previous illnesses/surgery/admissions/medications
Allergies
Infectious contacts/recent travel
Family history
Social history – family circumstances may influence a child's disposition significantly
Fasting status if relevant
Feeds – normal bottle or breast feeds for comparison
Urine output – number of wet nappies

Box 1.1.3 Examination warning bells

The pale, pasty child
The floppy child
The child who appears drowsy
Alteration in vital signs, SpO_2
Early signs of compensated shock
The tiring child with respiratory distress
The child who never smiles despite appropriate prompting
The child who looks sicker than the usual child with gastroenteritis/croup/bronchiolitis/URTI
Other specific signs
Non-blanching rash – petechiae/purpura-sepsis
Widespread blanching rash – toxin-mediated illness, including toxic shock
Bulging or full fontanelle – raised intracranial pressure
Bilious (green) vomiting – bowel obstruction
High-pitched cry – meningitis
Grunting – respiratory distress

Gentle, distraction, painful last

Children are usually reluctant to have any painful area disturbed. Confirming tenderness needs to be gentle and unhurried to minimise any distress, with appropriate prior analgesia. Many young children will respond to age-appropriate verbal banter during the examination, which distracts from the perceived threat of the examining hand. Alternatively, one may need to gently palpate a tender right iliac fossa, whilst using distraction such as the counting of the child's fingers. Sometimes a child may prefer their tender abdomen to be palpated with the examiner's hand 'through their own hand'.

The examination needs to be adapted to the child's responses, deferring distressing phases until the final moment of examination. Time used initially to gain a child's confidence will make subsequent assessment more rewarding and the clinical signs more reliable. It can be difficult and sometimes impossible to accurately make an assessment in an upset and distressed child.

Often the most reliable method of excluding peritonism or other serious problems in a child does not involve any palpation of the abdomen or specific examination. Asking a child to cough, walk, run, jump or climb the trolley is a useful manoeuvre to help exclude peritoneal irritation and will give a lot of useful information in other scenarios too.

The examination of ears and throat, a tender abdomen or a painful injury is best left until last in order to minimise upsetting a child and make the remaining routine examination difficult. If one detects that a child has an unfortunate fearful memory of a stethoscope or the like, a preliminary auscultation of a child's soft toy and warming the diaphragm will often allow this to subside.

Improvise

The examination of infants and young children is best done in the least threatening position. This is usually with the child being held comfortably in the parent's arms or on the knee. If a young child is sleeping, the opportunity should be taken to perform auscultation and palpate a fontanelle/abdomen, which will be altered in the crying state, prior to disturbing the child to wakefulness. A neonate examined supine needs to be kept warm with a blanket or a radiant heater. Hands should be warm. The crying fractious baby may be settled by offering a feed or a pacifier before the examination. The symmetry of normal infant reflexes is a useful screen for any focal motor problem or as a localiser of a painful limb that will modify normal symmetry of response. If possible, it is best to avoid waking a sleeping child. Instead and with parental agreement, return later and complete your assessment if this is clinically appropriate.

Observation

Your examination through observation begins prior to introducing oneself to the family and continues after the traditional examination, whilst writing up notes or between seeing another patient. It also includes reading and acknowledging nursing remarks and vital signs recorded in order to obtain additional information about the child's presenting problem. The trends of vital signs over time are useful indicators to detect evidence of disease progression or response to therapy. This 'ongoing triage', in effect, is particularly important to detect 'evolving illness' that may otherwise remain undetected. Subtle features that may be missed on examination include persistent tachycardia or tachypnoea that

is not clearly related to fever. Highly abnormal vital signs that fail to improve should not be attributed to fear, pain or distress and should raise concern of serious underlying pathology. One needs to be alert to the spectrum of stigmata of non-accidental injuries that may present to the ED (Chapter 18.2).

Observational variables

The general appearance of a child should include noting his/her level of alertness, eye contact, activity, quality of cry, posture, interaction with the environment, irritability, colour, hydration, perfusion, general growth and nutrition, respiratory distress and presence of any unusual smell (e.g. ketones). The lack of normal resistance to examination or a procedure expected of a child is an important observation to note. The sick child may make none of the resistance expected to examination or venipuncture.

Observational variables have been shown to be more predictive of serious disease than historical information in young children.[7] Likewise, clinical examination, considered alone, is a poor predictor of serious illness. Observation of a child needs to be performed as a separate process from the examination and may require a period of time for re-evaluation to detect disease progress. Researchers have used formalised scales such as the McCarthy Observation Scales to aid this assessment in febrile children.[8] In the ED setting, discussion of a child with a colleague can be a rewarding aid to decision making.

A child's posture, undisturbed, can be a useful clue to systemic illness, abnormal neuromuscular function or a painful limb or joint. Children with sepsis or meningitis may be floppy or flaccid. In other cases, the only sign of meningeal irritation may be a child who is holding his/her neck in a slightly extended position.

Observing breathing

When observing tachypnoea in a child, it is useful to determine whether a child has 'quiet tachypnoea' (breathing fast and quietly) with no evidence of increased work of breathing, such as may occur in conditions of fever, acidosis or cyanotic heart disease. This should serve as a trigger to look more broadly for the source of illness. Children with 'noisy tachypnoea' (breathing fast and hard) demonstrate increased work of breathing, due to conditions such as airway obstruction, pneumonia or heart failure.

Confounders

There are several observational confounders that can influence initial decision making, when children can appear transiently sicker than they really are. Frequently, an initially sick-appearing child can pick up and appear well again over a short period of time. This may be due to a response from analgesics or antipyretics and generally occurs within an hour or so of administration. The vomiting child will often look pale and ‘pasty’ for up to 20 minutes post vomiting. Children initially emerging from a simple febrile convulsion can look well again in 20 to 30 minutes. A young child’s physiological sleep can mimic septic drowsiness or somnolence resulting from head trauma. A fearful child, during examination, can escalate the examiner’s perception of his illness.

Re-evaluate

This reinforces the power of a period of observation (Box 1.1.4). It allows time for a trial of fluids, reducing fever with an antipyretic, or seeing if a child responds to distraction. Subsequent re-evaluation of the child often allows one to differentiate whether a child is sick or well. The use of observation really allows one to identify the persistence of the initial abnormal examination findings. A child with intussusception may intermittently appear well, and observation may be required to observe the episode to prompt the appropriate diagnostic investigation.

Box 1.1.4 Observation warning bells

Decreased level of alertness, activity, eye contact
Drowsiness or decreased interaction with the environment/parents
Abnormal posture
Abnormal quality of cry
Prolonged irritability or inconsolability
Ongoing pallor
Decreased peripheral perfusion or hydration appearance
Persistence of abnormal recorded vital signs
Respiratory distress/tachypnoea (‘quiet’ or ‘noisy’)
Persistence of examination warning bells
Confounders – post vomit/seizure/head injury, high fever, normal sleep,

anxiety

Respiratory examination

Noisy breathing in children can sometimes be difficult to determine whether it is due to airway obstruction of intra- (lower airway) or extra-thoracic (upper airway) origin. The localisation of airway obstruction to a particular segment of airway can often be aided by successive auscultation over the nares, mouth, larynx and peripheral airways. Remember, young children may manifest both upper and lower airway involvement (‘asthma/croup/bronchiolitis’) with inflammatory involvement of both segments of the respiratory tract.

Younger children are often easier to auscultate by listening through clothes (avoiding the ‘stethoscope–cry reflex’) from behind whilst being held by the parent. Modern stethoscopes tend not to be cold when placed on skin. Detection of ‘occult’ asthma in a child with suggestive symptoms but no wheeze may be aided by comparing the diminished volume and rate of airflow in expiration compared to inspiration or alternatively re-auscultation after exercising the child in the ED. Wheeze may also be unmasked through a therapeutic trial of salbutamol.

Young children with throat discomfort will be reluctant to volunteer a cough, but a gentle tickle of the axilla or palpating the anterior larynx will usually produce a bark to clarify suspicion of croup. Recognising the pattern of respiratory distress in a child from the end of the bed will often differentiate upper and lower airway obstruction, prior to any auscultation. Children with upper airway obstruction have slower inspiration, whereas gas-trapped wheezers will have diminished flow and speed of expiration on observation.

Abdominal examination

The abdominal examination needs to always conclude with the nappy area for otherwise occult torsions, hernias, skin problems and for stool examination, if present. Rectal examination in children is not routine and should only be performed with clear indication and almost always by a paediatric surgeon. Privacy and dignity must be maintained, particularly when examining older children and adolescents. The examination of a child with possible sexual abuse is outlined in Chapter 18.1.

ENT last

In preschool age and younger children examination of the ears and throat is best deferred to last. A gentle but rapid approach is necessary to achieve an accurate assessment of the oropharynx, followed by a cuddle from the parent. Despite the potential difficulty, the source of fever will often be overlooked if the throat is inadequately visualised in children. In infants, the throat is best examined with the child sitting in his/her parent's lap with both arms cuddled by one of the parent's arms whilst his/her other arm secures the child's head. A young child who is fearful of throat examination needs to be held as still as possible for a rapid 'one gag, one look' approach. Be careful though as this is a great technique for creating an infective aerosol that you may come to regret in several days' time. 'Let's count your teeth' is a less threatening signal to most children to open the mouth, rather than mentioning 'the tonsil or throat' words, particularly if parents warn you that 'Nobody has been able to get a look at his throat'. As children get older, this examination becomes easier to perform.

Following any distressing procedure, it is important to acknowledge bravery in a frightened child. Likewise, giving a child an honest, developmentally appropriate explanation of what to expect prior to any procedure, such as an IV insertion, must always occur. This is best done immediately prior to the procedure so that an anxious child's fears don't escalate in the intervening period.[6]

When to investigate

Investigations in children in the ED should be judicious and considered. Investigations serve more than one purpose. They might help confirm or refute clinical suspicions. Occasionally, parents may appear to initially want more reassurance than simple clinical assessment and explanation. In this situation, the utility of investigations (along with the associated distress and discomfort obtaining them) needs to be placed into context for the parents. Serial review over time may often prove a more effective and less distressing technique to evaluate a child for a serious illness. Investigation for parental reassurance should not be a routine practice unless also clinically appropriate.

The parents

Parents who accompany their child to an ED are often anxious and fearful

regarding the safety of their child. It is important to consider that the parents are entrusting the doctor with the wellbeing of their most cherished and precious possession.

The management of their fears and the identification of their needs and expectations are important roles of the doctor attending to their child. Listening to and addressing the parents' concerns in a sympathetic and unhurried fashion is often the main therapeutic strategy to reassure an anxious parent that a child with a relatively minor illness is safe.

Many parents may be sleep deprived due to attending to their sick child, and this will influence their ability to convey a lucid history or to receive new information. The time spent at triage or in the waiting area in a busy ED can frustrate the most patient parent. This needs to be anticipated and acknowledged at the start of the consultation. Sensitivity to potential cultural issues is important in all interactions with carers.

Managing the parents

The ED visit may follow previous medical consultation(s) where their concerns may not have been addressed or unfairly amplified, and it is important to explore these. Always acknowledge the parents' fears and anxieties; however, medical judgement should allow an objective decision about whether a child is sick or not. An exception to this is the parents of children with a chronic illness or special needs (Chapter 1.2). They are usually correct when they judge that their child is sick.

Aggressive and unreasonable carers will usually respond to a professional, polite and courteous senior doctor. There is usually a reason behind their behaviour that needs to be explored. Even the most anxious parent will usually respond to a thorough assessment of his/her child followed by an explanation of diagnosis and management. In unplanned second presentations where parents demand admission, it is usually best to admit.

Communication issues

Explanations to parents as a general rule should be appropriate to their level of understanding and education. Provision of appropriate handouts – in the parents' preferred language – early in the ED attendance allows parents to improve their knowledge while waiting to be seen, for investigations to occur, or for a period

of observation. This early information provision saves time at the end of the visit.

Once a plan is determined, a verbal explanation reinforced by written instructions is useful to ensure optimal understanding. Reviewing a parent's understanding of instructions prior to discharge will allow clarification and avoid communication problems. Parents may have fears related to anecdotal advice from family/friends, misinterpretation of media reporting, the internet and social media, or other sources, which need to be explored.Gaining the confidence of parents before they leave the department is an essential part of the therapeutic process and has a positive effect on compliance to therapy. It is useful to explain to parents the likely natural history of their child's illness and encourage review should significant deviations from this occur.

Where an ED assessment differs significantly from that of the referring clinician, great care must be taken to address differences in opinion and management plans. In addition, timely communication with the referring clinician should occur, in order to clarify any differences in clinical assessment and to provide an update on the child's ED course. Parents will return to primary care doctors for ongoing treatment, and it is important for emergency physicians to maintain and build confidence in the community healthcare providers.

Management of febrile children

The management of febrile, young children is a large part of emergency paediatric practice. Children less than 1 month old require a full septic evaluation if rectal temperature is greater than 38.0^{o}C. This should include FBE and cultures of blood, urine and cerebrospinal fluid (CSF), followed by empirical antibiotics. A chest X-ray (CXR) may be required if symptoms and examination findings suggest chest infection. Febrile children between 1 and 3 months require a graded approach with FBE, blood culture and urine culture assisting to risk stratify the child. Collection of CSF for culture and administration of empirical antibiotics should occur if the child looks unwell.

Children older than 3 months can be managed to a greater extent based upon clinical findings. A well, febrile child with a clear focus of infection can be managed as clinically indicated. Unwell children with a clear focus require further evaluation and admission for treatment. Children older than 3 months without a clear focus of infection who look well should have a clean urine sample collected for microscopy and culture. This well group should have

review arranged for the following day to assess progress and to check on laboratory results. Unwell-looking children in this age group without a clear focus should have a septic workup including FBE and blood and urine culture. A CXR and lumbar puncture (LP) may be indicated based upon clinical symptoms or signs. Admission for observation and potentially antibiotics can be arranged after discussion with the admitting paediatric service.

Parents whose child is discharged home should be clearly instructed to return to the department if their child deteriorates. The discharge action plan should give clear and understandable instructions on when to return. For example, in the febrile child, this should include if the child becomes more unwell, with a decrease in oral intake to less than 50% normal, with no urine output for 6 hours, or the child becomes drowsy beyond his/her normal sleeping.

Parents should be alerted to potential complications such as becoming limp, fitting or appearance of a rash, which warrant urgent review. Parents should always leave the ED feeling empowered to return for further medical care in the ED or with the GP if they feel their child is not improving or worsening. Never minimise or trivialise parental concern, as this may lead to reluctance to seek further medical advice.

The presence of fever itself provokes considerable parental anxiety, often more pronounced in parents (and other family members) from specific cultural groups. This is important to address with explanation and provision of culturally appropriate written information which can be taken home and shared with the extended family. Additionally, it is important to avoid routine administration of antipyretics to all febrile children at triage, a practice which reinforces caregiver ‘fever phobia’.

Reasonable expectations

Managing children is often about understanding the natural history of the illness and predicting when the child will improve. Presenting parents with a reasonable expectation of when they might expect their child to improve is one way of ensuring the child is safely managed and parents are reassured. This approach will not avoid the need for serial review in either the ED or by the GP, but it may provide some reassurance. A recent systematic review found that the expected duration of symptoms for some common childhood respiratory illnesses is quite prolonged: ear ache – up to 8 days; sore throat – up to 7 days; common cold – up to 15 days; acute cough – up to 25 days.[9]

Decision making

Making decisions in paediatric emergency medicine is a balance of history, examination, intuition, knowing when to trust the parents and maintaining objectivity. If you feel uneasy with your diagnosis regarding a child, respect that feeling, and gain support until you do feel comfortable with your decision. There are many strategies to do this:

1. Consider early consultation with an emergency or paediatric consultant colleague.
2. Organise early follow-up in the ED or with a paediatrician or general practitioner.
3. Have a colleague on the floor listen to the story or examine the child.
4. Phone review the patient's family yourself later that day or the next morning.
5. Provide a concrete action discharge plan.
6. Admit the patient for observation to either the ED observation ward or to the paediatric ward.
7. Empower the parents to return to the ED if they are concerned after discharge.

When to admit

The decision to admit or discharge a child from the ED is easily made when the child requires medical care that is only available in the hospital setting. The receiving ward will need to have appropriate resources for the ongoing management of the child, which should be clarified by discussion with the receiving inpatient unit/paediatrician. Some children may need to be discussed with and transferred to a tertiary paediatric environment when they require, or have the potential to require, paediatric intensive care facilities or paediatric subspecialty management.

Factors influencing disposition

However, many other factors need to be considered in the disposition decision (Box 1.1.5). The threshold to admit a child is influenced by the child's age, availability of appropriate follow-up, assessment of parents' ability to provide care and ongoing monitoring, the natural history of the illness and likelihood to

deteriorate, social factors, comorbidity, distance from hospital, time of day, parental anxiety levels, availability of an early paediatric opinion, and the possibility that a child may be at risk. One needs to assess in a non-judgmental fashion the ability of the parents to carry out any ongoing treatment and consider admission if there appears to be a need for ongoing support. When in doubt regarding whether or not to discharge a child, err on the side of caution. It may be prudent to consult, consider a period of observation in the ED, or admit the child to hospital.

Continuity of care

It is important for ED staff to liaise closely with the admitting paediatrician to provide continuity of care and to ensure ongoing care is expedited in the ED. Ongoing management and monitoring of the patient are important roles of medical and nursing staff after this decision has been made, particularly if there is a delay in the transfer process. Any significant change in a child's previous status or treatment needs to be communicated to the appropriate receiving team. The development of unstable vital signs or other evidence of severe illness requires an appropriate escalation in treatment, including activation of a medical emergency team or arranging inter-hospital transfer.

Box 1.1.5 Factors influencing admission threshold

Age of child
Availability of appropriate follow-up/review
Parental ability to provide care and monitoring, social factors including the care of other children
Comorbidity
Distance from hospital and ease of returning
Time of presentation
Parental anxiety levels
To enable further observation or obtain a paediatrician opinion
Possible child at risk outside hospital

Observation ward

Significant compromise from many childhood illnesses is often transient and will often respond rapidly to interventions commenced in the ED followed by a period of observation. Parents can often be reassured during this period of observation in hospital that their child has remained well and will respond to management strategies that subsequently can be continued at home.

Studies have shown that many children admitted to hospital only require a limited period of in-patient therapy and are discharged in less than 24 hours.[10] In a tertiary paediatric environment an effective way to manage these children is by admission to a short-stay observation ward. The ED needs to be appropriately resourced with staff to provide ongoing care and regular review of patients to expedite timely discharge.

Conditions suitable for consideration of an observation ward admission will vary with local resources and may include asthma, croup, gastroenteritis, febrile convulsion, presumptive viral illnesses, non-surgical abdominal pain, minor head injuries and other trauma, post-sedation recovery or ingestions.[11] In mixed departments, without the facility of a short-stay ward, it is often appropriate to use the paediatric ward to admit patients who would benefit from a period of observation.

Making a diagnosis

Not every child leaving the ED will do so with a specific diagnosis. The unwell febrile child needs to have serious diagnoses such as meningitis considered, before making a diagnosis of viral illness. Many children without a clear diagnosis can be managed expectantly and safely discharged home with organised review by a local doctor or paediatrician or return to the department. Giving the parents clear instructions to return should the state of their child not follow an expected course is essential. Children are often seen early in the natural history of their illness, and a diagnosis will only become clear with time.

It is important to communicate clearly, verbally or in writing, with the doctor who will be following up the child. Close liaison with a local doctor who has referred a patient to the ED is essential. One should always respect the concerns raised by a referring local doctor who usually has the advantage of familiarity with the child and family.

The role of the GP in paediatric emergency

management

Introduction

GPs are the cornerstone of the Australasian health system and often the child's first point of contact when unwell or injured. They are uniquely placed to have an intimate working knowledge of the biological, psychological and social dynamics that impact on a child's illness.

GPs are involved in the long-term care of family members, often for many years and in some cases several generations. It is this continuity of care and ongoing relationship with a family that are invaluable in assessing and triaging presenting medical conditions in children and their subsequent management. This is particularly important when caring for the health of children, who are often seen in the early prodromal phase of serious illnesses. It is the skill of the GP to differentiate the possibility of a serious illness in a child, particularly during the seasonal peaks of febrile presentations. This child may be non-specifically 'different' to similarly febrile children, but the experienced GP may just have a 'gut feeling' that a second opinion may be warranted and refer to the ED.

There are two distinct areas where the GP plays a vital role in paediatric emergency management:

1. Assessment, initial stabilisation and transfer of the child to the paediatric ED of the clearly 'unwell' or 'potentially unwell' child
2. Ongoing management and follow-up of the child after discharge from a hospital encounter, particularly when a diagnosis is yet to be made.

Management prior to hospital care

The GP is more often than not the first point of contact for the potentially unwell child. The fundamental clinical medical tools of history taking and examination are used to make an initial assessment of whether the child can be treated in the community or requires referral to an ED for further opinion and management. This can be a challenging task as the GP is not afforded the luxury of observation over time, readily available ancillary testing such as pathology and imaging, or an immediate further opinion from a specialist colleague.

Particularly in the case of early or undifferentiated illness, the GP will need to make a judgement call on whether or not a child can be safely managed at home.

Experienced GPs will not only use traditional methods of history and examination but will also listen to their ‘gut feeling’ when assessing children.

This may involve attaching importance to red flag symptoms or signs or heeding the warning signs reported by an anxious yet appropriately worried parent. This may depend on not only the medical status of the child but also the assessment of the social circumstances, education and competence of the parents/carers to detect their child is failing to ‘turn the corner’ or deteriorating. Often there is significant parental anxiety with an unwell child which cannot always be allayed by sound advice from an experienced GP when a child clearly has a self-limiting viral illness.

There may be parental demands for pathology testing to ensure ‘nothing is missed’ even though these may be deemed inappropriate by the family doctor. Parents may also report significant symptoms such as fever, an infant not feeding normally, cough or stridor which may no longer be present at the time of presentation to the GP. Some auscultatory chest findings are dynamic and therefore have a fluctuating presence, such as wheezing in bronchiolitis, so may vary greatly between the time of the GP and ED visit.

It is this complex interaction of medical and environmental factors which must be processed by the GP, often in the context of a 15-minute appointment. The outcome of this assessment may be the subsequent referral to hospital-level care. Remember that the GP’s decision is carefully considered with all the aforementioned factors coming into play.

Some of the more common reasons for referral to the ED may include the following:

- A serious time-critical illness which requires ambulance transfer, such as severe asthma, sepsis or meningitis
- Non-time critical illness which may not be responding to community-based treatment and requires further investigation or consideration of parenteral antibiotics, such as evolving pneumonia
- Illness or injuries which are beyond the level of facilities available to the GP to manage, for example unstable limb fractures or those that require a period of supervised acute treatment and/or prolonged observation
- Parental concern and anxiety which cannot be sufficiently allayed by the GP
- Social factors whereby the child cannot be adequately cared for or progress if monitored in the home setting due to lack of family

resources.

It is imperative once the decision has been made to refer the child on to the ED, that the clinical assessment and concerns of the GP are adequately communicated to the physician who will be the next link in the management chain. This is best done with a phone call to the ED outlining the reasons for referral. In potentially serious illnesses the ED clinician can provide initial phone advice.

A referral letter which contains the child's past medical history, allergies, immunisation status, list of current medications and any relevant investigations should accompany the child to the ED. This gives the ED doctor a head start in managing the child and avoids wasted time, effort and cost in repeating already established findings.

Management after hospital care

Once the child has been managed and discharged from the ED the circle of communication should include verbal and written feedback to the referring GP. This timely discharge communication has several benefits.

First and foremost, it ensures continuity of care for the child. If a treatment plan has been commenced by the hospital staff, the GP is then responsible for its implementation through continuing clinical assessment and adjustment of management according to progress. The natural history of illness and convalescence are dynamic processes which will vary from patient to patient and may require vigilant monitoring. This is most likely to be successful if the discharge plan is well communicated to the family doctor. It is vital that communication is not mislaid compromising patient care. The GP should receive information directly (fax or electronic) as well as via the patient or family as a backup if the usual communication systems fail.

Second, medical practitioners continue to accumulate knowledge and expertise throughout their careers so that reflective and sensitive feedback concerning outcomes of their referred patients is useful. This helps the GP to analyse and reflect upon their decision-making processes and contribute to their evolving clinical acumen, which is a career-long journey for all doctors. This is particularly so in the case of paediatrics as recognising the potentially unwell child can sometimes be as much art as science.

GPs can arrange further monitoring of the recovering child and are well

placed to arrange further tests (for example chest X-ray following complicated pneumonia) or specialist follow-up if needed. Often the busy ED is not the easiest place, especially after hours, to arrange such important steps in the child's follow-up care. The GP is also able to assess any psychological impact of the child's illness and offer ongoing support to the child and the family. These potential issues may not be evident at the time of the ED visit.

Integral to the communication process is a mutual respect between the GP and the ED physician with both having an understanding and appreciation of the environment and challenges that each is working under. GPs have strong attachments to their patient and families and will appreciate a follow-up phone call and/or letter advising of the status of a referred child. The letter should be timely, with appropriate information including diagnosis, medication and results of investigations with an access phone number for any results pending. It should be presented in a clear concise form with a structured plan of management. Computer-generated letters are often more legible than hand-written ones and reduce the chance of miscommunication in the discharge process. Some GP clinics now have secure email availability and may prefer to receive information this way.

If these strategies are implemented within a spirit of cooperation between GPs and EDs, this will ensure improved continuity of care and therefore better patient outcomes in the care of sick children.

Developmental milestones

It is important to have an understanding of the major developmental milestones throughout childhood for the provision of care to paediatric patients. These can be rapidly confirmed by examination or parental enquiry. This allows the use of an appropriate age-modified approach in the child's evaluation. Some specific behaviours, such as stranger anxiety in a 12-month-old, may challenge assessment, so it is important to adapt an approach to these expected behaviours. Significant deviations from normal warrant consideration of paediatric referral. Useful early milestones are shown in Table 1.1.1.

Growth

It is essential to measure a child's current weight on every ED visit in order to accurately dose any therapeutic drug and to quantify recent weight loss. Where a

child is critically unwell and unable to be weighed, estimation can be made via Broselow or other charts. Between the ages of 1 and 10 years an estimation of weight is 2 × (age + 4) kg.

Standardised percentile growth charts are useful to confirm suspicion of failure to thrive or discrepancy in linear or cranial growth. The trend of growth plotted on a growth chart over time is more important than a single measurement. As a general rule, birth weight doubles by 5 months and triples by 1 year. Newborns are often discharged from hospital in the first few days of life and may present in the first week to an ED. Following the expected initial weight loss, term babies should normally regain their birth weight by the end of the first week. Appropriate neonatal weight gain is an important index of wellness and can be tracked on the growth or centile charts that all babies receive after birth. Head circumference increases by 2 cm in the first 3 months, 1 cm in the next 3 months, followed by 0.5 cm per month thereafter (Table 1.1.2).

Table 1.1.1

Normal milestones in first 2 years of life

Neonate	Lift head, visually fix for period
6 weeks	Smile, follow past midline
4 months	Roll over
6 months	Sit, transfer toy between hands
9 months	Stand holding on, crawl, stranger anxiety
12 months	Walking, single words
18 months	Explorer (trauma/poisons), tantrums, several words
2 years	Combine words, run, jump

Table 1.1.2

Estimated normal growth

Age	Weight (kg)	Height (cm)	Head circumference (cm)
Birth	3.5	50	35
1 year	10	75	47
2 years	13	88	49
>2 years	+2 kg yr^{-1}	+6 cm yr^{-1}	N/A

Adapted from Gary R. Fleisher, Stephen Ludwig, Textbook of Pediatric Emergency Medicine, 6E, Lippincott Williams & Wilkins, 2010.

Immunisation

It is not the role of the ED to provide routine immunisations to children. It is useful, however, to clarify a child's immunisation status with regard to the possibility of a particular infection such as epiglottitis, whooping cough or measles. Despite immunisation, children may still acquire these diseases but usually manifest a modified form of these infections.

In children who are found to be incompletely or non-immunised, it is opportunistic to respectfully provide information regarding the normal vaccination schedule and refer to the local doctor or appropriate community facility for follow-up (Table 1.1.3). Being disrespectful or rude about parental choice to not vaccinate their child risks the loss of any therapeutic relationship with the family.

Vital signs

It is necessary to interpret the vital signs according to the age of a particular child. A wall chart in the paediatric resuscitation area is a useful reference as a guide to these parameters. A good rule to remember is any child with a persistent respiratory rate >60 or heart rate >160 is definitely abnormal (Table 1.1.4).

An important aspect of the care of children in the hospital setting is the increasing use and availability of 'early warning tools'. These identify and flag abnormal age-appropriate vital signs, in order to escalate the level of care for children with potentially critical illness.

Reflection on the Practice of Paediatric Emergency

The practice of paediatric emergency can be immensely rewarding. The opportunity to play games in the context of assessing patients is a wonderful way to spend a working day. However, great care and attention are required to identify those children with serious or life-threatening illness from those who have a viral illness. An unhurried, calm approach with serial review and close, careful follow-up can minimise the likelihood of missing a serious diagnosis. Involvement of the child's carers is vital to ensure they are comfortable with the assessment made in the ED, have had an opportunity to ask questions and have been provided with clear follow-up instructions.

Table 1.1.3

Australian standard vaccination schedule (0–5 years)

Age	Vaccine
Birth	HepB
2 months	HepB-DTPa-Hib-IPV; 13vPCV; Rotavirus
4 months	HepB-DTPa-Hib-IPV; 13vPCV; Rotavirus
6 months	HepB-DTPa-Hib-IPV; 13vPCV; Rotavirus
12 months	Hib-MenC; MMR
18 months	DTPa; MMRV
4 years	DTPa-IPV

Adapted from www.immunise.health.gov.au, Accessed December 2016. The most up to date schedule is available from the Immunise Australia website at www.immunise.health.gov.au., The footnote is as follows and is included in the online NIP schedule.∗ Rotavirus vaccine: third dose of vaccine is dependent on vaccine brand used. Contact your State or Territory Health Department for details.

Table 1.1.4

Normal vital signs

Age	Weight (kg)	RR (per min)	HR (per min)	sBP (mmHg)
Birth	3.5	40–60	100–170	50
3 months	6	30–50	100–170	50
6 months	8	30–50	100–170	60
1 year	10	30–40	100–170	65
2 years	13	20–30	100–160	65
4 years	15	20	80–130	70
6 years	20	16	70–115	75
8 years	25	16	70–110	80
10 years	30	16	60–105	85
12 years	40	16	60–100	90
14 years	50	16	60–100	90

Adapted from Royal Children's Hospital. Clinical practice guidelines resuscitation: emergency drug and fluid calculator. Melbourne, Australia, 2003.

It is a humbling situation to encounter a child with a life-threatening illness. In the event of an adverse outcome it is important to not only review the event but to care for the family and staff involved. The impact and distress caused by the death of a child are significant.

By reading this textbook you are on a journey to be well positioned to recognise and manage the sick child and hopefully change the outcome for the better. For that, the child's parents will be forever grateful.

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1.2

Common chronic paediatric conditions

Erin Mills

ESSENTIALS

1 Children with chronic medical conditions make up 10–20% of paediatric emergency department (ED) presentations.

2 These patients have a longer ED stay and have increased rates of hospital and intensive care unit admission.

3 Trust the parents/caregivers: if they are worried, you should be worried.

4 Minimise pain and anxiety associated with procedures.

Introduction

Paediatric patients with chronic medical conditions (Table 1.2.1) make up 10–20% of presentations to tertiary paediatric emergency departments (EDs).[1,2] Compared to children without chronic medical conditions, these patients have a longer ED stay and are more likely to be admitted to both the ward and the paediatric intensive care unit (PICU).[1–3] Therefore it is essential that the emergency physician is equipped to deal with this potentially complex group of patents.

Table 1.2.1

Common chronic childhood conditions

Neurological	Epilepsy Cerebral palsy Ventriculoperitoneal shunt Spina bifida
Cardiac	Congenital heart disease Arrhythmias
Respiratory	Asthma Cystic fibrosis Bronchopulmonary dysplasia
Renal/urological	Vesico-ureteric reflux Chronic renal insufficiency Nephrotic syndrome
Haemato-oncological	Haemoglobinopathies Coagulation disorders Cancer Immunodeficiency
Endocrine	Diabetes mellitus Obesity
Developmental	Autism Attention-deficit/hyperactivity disorder
Other	Psychiatric disorders Gastroenterological problems Ear, nose and throat problems

The emergency physician may be involved in the initial diagnosis but is mostly required to recognise and manage disease complications as well as acute illness unrelated to the chronic condition.

This group of patients can be very complex, with numerous medical and psychosocial issues and a baseline abnormal examination. It can be extremely difficult for the emergency physician, in a time-pressured ED scenario, to gain a thorough understanding of the patient and the level of deterioration. It is very important to listen to the parent or caregiver’s concerns, as well as involve the patient’s primary care team if possible. The parents are the best advocate for their child, and they know their child better than anybody else. If they are worried, the clinician should take this seriously.

Chronic illness brings with it a range of stressors for the child and family, including those surrounding painful procedures such as venous access. As with any child, but especially in this population who are likely to be subjected to multiple procedures over time, it is critical to minimise the trauma surrounding potentially painful procedures. Tools used may include distraction, parental presence, play therapy, positioning on a parent, topical anaesthesia, pharmaceutical analgesia, or procedural sedation. In the appropriate clinical setting, oral midazolam 0.5 mg kg up to 20 mg can be mixed with sucrose and

administered 20–30 minutes prior to attempts at intravenous access. Inadequate pain management is known to have long-term negative effects on children, including diminishing effects of adequate analgesia for subsequent procedures or needle phobia, which make future procedures more traumatic for the child and more difficult for the clinician.[4,5]

Table 1.2.2

Cerebral palsy motor syndromes

Spastic	Most common type Hyper-reflexia, weakness, hypertonia May be a: hemiplegia: unilateral arm and leg involved diplegia: both legs involved quadriplegia: all limbs and often trunk and facial muscles involved
Dyskinetic	Athetotic: involuntary, slow, writhing movements
	Dystonic: trunk movements are more affected than limb muscles, resulting in a twisted position
Ataxic	Voluntary movements are not well coordinated
Hypotonic	Muscle tone is decreased
Mixed	A combination of the above

This chapter will provide an overview of some of the more commonly encountered chronic paediatric conditions, including cerebral palsy (CP), spina bifida, cystic fibrosis (CF), the ex-premature infant and autism spectrum disorder.

Cerebral palsy

Introduction

CP is not a single entity but rather a heterogenous collection of clinical syndromes characterised by abnormal motor patterns and postures (Table 1.2.2). It is the most common chronic motor disability of children, with a prevalence of approximately 1/500 live births.[6] CP is caused by an insult to the developing brain, such as infection, preterm birth, intrauterine growth restriction, ischaemia, congenital malformations or head trauma. The degree of motor function may vary widely and can be communicated quickly using the GMFCS (Table 1.2.3). Children may have associated impairments including epilepsy or intellectual, speech, visual or hearing impairment. However, it is important to remember that many children have normal cognition.

Complications seen in the emergency department

Children with CP are susceptible to frequent illness because of their increased risk of epilepsy, respiratory complications and feeding difficulties, as well as complications associated with increased technology dependence. Increasing CP severity and complexity are associated with increased frequency of presentations, higher urgency of presentation and higher rates of hospital admission. The majority of presentations to the ED are due to respiratory or neurological causes, followed by gastrointestinal and musculoskeletal complications.[7,8]

Respiratory complications

Respiratory illness is the leading cause of mortality in individuals with CP, and chest infections are the most common reason for children with CP to require hospital admission.[9]

History

Predisposing factors include the following:

- Oropharyngeal motor incoordination (leading to aspiration of food and secretions)
- Gastro-oesophageal reflux
- Poor cough and airway clearance
- Immobility
- Kyphoscoliosis
- Sleep apnoea
- Malnutrition, leading to atrophy and weakness of respiratory muscles.

Causative organisms of pneumonia are similar to other children, though anaerobes may also be involved in the setting of possible aspiration. Risk factors for serious illness include previous ICU admission and severe scoliosis.

Examination and investigation

Assessment is similar to other children with respiratory illness:

- Respiratory distress (respiratory rate, intercostal/subcostal recession, grunting)
- Oxygen requirement
- Respiratory failure (conscious state, pH and CO_2 on venous blood gas)

• Wheeze and bronchodilator responsiveness.

Note that chest X-rays will be difficult to interpret in those with severe scoliosis.

Table 1.2.3

Gross motor function classification scale (GMFCS)

I	Can run, jump and climb stairs, but speed, balance or coordination is impaired
II	Can walk, run and climb in most settings but may need mobility equipment for safety or travelling long distances
III	Can walk using assistance and can self-propel manual or powered wheelchairs
IV	Capable of walking with a walking frame but mostly relies on wheeled mobility
V	Little or no voluntary control of movement and requires extensive support in all situations

Management

Oxygen and respiratory support

- Give oxygen if the saturations are <90% in room air
- If the child has chronic respiratory failure, aim for saturations 90–95%
- Consider respiratory and airway support as necessary.

Antibiotics

- Follow local guidelines for community-acquired pneumonia
- Have a lower threshold to start antibiotics in children with severe CP – these children are more likely to have complications of pneumonia and worse outcomes
- Penicillin is thought to provide adequate anaerobic cover for aspiration pneumonia
- If very unwell consider adding Gram-negative cover.

Salbutamol

- If there is evidence of bronchospasm.

Secretion management

- Nebulised saline may help to mobilise secretions (5 mL normal saline via nebuliser)
- Oro-pharyngeal suctioning if the child is unable to swallow secretions
- Chest physiotherapy may be helpful in those with poor cough.

Feeding

- If moderate or severe respiratory distress give intravenous (IV) fluids at 2/3 maintenance
- If respiratory distress is less severe and the child is percutaneous endoscopic gastrostomy (PEG) fed, continuous feeds at 2/3 maintenance may be appropriate.

Consider ICU referral if severely unwell, as per local guidelines. Remember to take account of the child's prognosis, level of understanding, quality of life and advanced care directives if available when formulating a management plan.

Neurological complications

Seizures

Up to 50% of children with CP will have epilepsy, and increased seizures or status epilepticus are common ED presentations. It is important to consider possible precipitants such as intercurrent illness, CNS infection, change in medication, poor compliance or ventriculo-peritoneal shunt malfunction if present. Management of afebrile seizures has been discussed in Chapter 8.3.

Cerebrospinal fluid shunt complications

Children with CP are more likely to have cerebrospinal fluid (CSF) shunts in situ. The assessment and management of shunt complications is discussed in Chapter 8.1.

Gastrointestinal complications

Percutaneous endoscopic gastrostomy tube complications

Many children with moderate or severe CP will require PEG placement due to malnutrition or recurrent aspiration secondary to oromotor or oesophageal dysphagia. Gastro-oesophageal reflux disease (GORD) is also common in this

patient population and if severe may require fundoplication in addition to PEG placement. Common PEG complications seen in the ED include infection, leakage and inadvertent tube removal. An approach to these complications is discussed in Chapter 24.16.

Constipation

Chronic constipation is a common complication of CP. Contributing factors include reduced fibre and fluid intake, reduced mobility and difficulty achieving optimal toileting posture, and prescription of anticholinergic or opiate drugs which increase transit time. Management of constipation has been detailed in Chapter 7.13.

Musculoskeletal complications

Musculoskeletal pathologies tend to be progressive in children with CP. Spasticity is common, affecting approximately 85% of children with CP to varying degrees.[10] Long-term management goals are to reduce contractures, therefore preserving function and reducing pain. Management of spasticity includes physiotherapy and splinting, as well as medical management, with surgery reserved for severe cases. It is important that emergency physicians are aware that hip dislocations and pathological fractures are more common in this population and must be considered in the patient who presents with irritability with no other cause found.

Common medications used in cerebral palsy

The medications used to manage spasticity may be unfamiliar to emergency practitioners. Oral medications include baclofen and dantrolene. Baclofen has poor oral bioavailability, so intrathecal baclofen is increasingly used for severe generalised spasticity or dystonia. Injected neurolytic medications such as botulinum toxin are also commonly used for focal spasticity.[11]

Baclofen

- Gamma-aminobutyric acid agonist that works by reducing release of excitatory neurotransmitters
- Administered orally or intrathecally
- Side effects: weakness, sedation, ataxia, nausea, dizziness and occasionally depression

- Overdose may cause severe flaccidity and coma, requiring intensive care support until the baclofen is metabolised
- A withdrawal syndrome may be caused by sudden discontinuation of oral or intrathecal baclofen and is characterised by seizures, hallucinations, hyperthermia, pruritis and rebound spasticity. Treatment of this potentially fatal syndrome is administration of oral or nasogastric baclofen, high-dose benzodiazepines and/or propofol (with appropriate airway and circulatory support) and, if intrathecal, prompt repair or replacement of the pump in conjunction with the patient's usual physician.

Intrathecal baclofen

- Allows for higher cerebrospinal fluid (CSF) concentration while reducing the likelihood or generalised side effects associated with oral administration
- The pump is planted subcutaneously in the abdomen, and the catheter is positioned intrathecally
- Side effects include weakness, fatigue, confusion and hypotonia.
- Sudden discontinuation such as pump malfunction can lead to the withdrawal syndrome described above.
- Pump complications include CSF leakage, catheter infections and meningitis.
- CSF leakage may be suspected due to postural headache, swelling around the pump, or clear discharge from the surgical wound site. It is important to notify both the neurosurgical team and the patient's usual management team, who will advise on appropriate imaging or surgical investigation.
- If infection is suspected the physician should commence a septic workup looking for alternate sources of infection. Lumbar puncture should NOT be performed before consulting the neurosurgeon and the patient's usual physician.

Dantrolene sodium

- Acts on skeletal muscle to reduce spasticity by inhibiting calcium release from the sarcoplasmic reticulum

- Administered orally
- The most significant side effect is weakness, though it may also cause drowsiness, diarrhoea, or rarely hepatotoxicity.

Botulinum toxin

- An exotoxin produced by the bacterium *Clostridium botulinum*
- Inhibits presynaptic acetylcholine release at the neuromuscular junction causing reversible partial flaccid paralysis of the muscle in which it is injected
- Administered by intramuscular injection
- Effects last 3–6 months
- Side effects include excessive muscle weakness, constipation, fever and incontinence
- Serious side effects such as dysphagia and aspiration pneumonia are rare.

Spina bifida

Introduction

Spina bifida is the incomplete formation of the spine and spinal cord and is the most frequent permanently disabling birth defect. It occurs due to a defect in the closure of the neural tube during the first month of pregnancy, leaving the spinal cord exposed and able to protrude through the open part of the spine. Patients with spina bifida have varying degrees of disability, including paralysis or weakness in the legs, bowel and bladder incontinence, hydrocephalus and specific learning difficulties. Symptoms vary depending on the position of the neural tube opening along the spine and on how much of the spinal cord or meninges protrude through the opening.

Types of spina bifida

Occulta

- Commonest and mildest form (estimated 5–10% of the population)
- Midline defect in vertebral bodies without protrusion of the spinal cord or meninges
- May be a dimple or tuft of hair over site

- Majority of people asymptomatic
- May present with urinary incontinence, toe walking gait, upper motor neuron signs in lower limbs.

Meningocoele

- Meninges protrude through a defect in the posterior vertebral arches
- Spinal cord usually undamaged, though there may be tethering or syringomyelia
- Often associated with hydrocephalus
- May be associated with constipation and bladder dysfunction.

Myelomeningocoele

- Most severe form
- Meninges and spinal cord protrude to form a sac that also contains CSF
- Abnormal development of spinal cord and nerves
- Disability depends on site of the lesion and extent of nerve damage
- Sacral lesions cause bladder and bowel incontinence with perineal anaesthesia
- Lumbar lesions cause flaccid paralysis of lower limbs, absent lower limb reflexes and reduced/absent sensation
- Upper thoracic and cervical lesions tend to have less severe neurological deficits
- All may be associated with hydrocephalus and Chairi II malformation.

Implications for emergency department

The most common reasons patients with spina bifida present to EDs are urinary tract infections and neurological complications.[12] Common neurological diagnoses are seizures, benign headaches and shunt malfunctions.

Latex allergy is common in spina bifida patients, and the risk of latex allergy increases with repeated exposure. Therefore exposure to latex products should be avoided for all spina bifida patients.

Cystic fibrosis

General

CF is the most common genetically inherited disease in white populations with an incidence of 1 in 2500 newborns.[13] It is a complex, multisystem disease that now has a life expectancy of more than 50 years. CF is usually managed in specialist centres, but they may present to any ED in times of crisis, and all emergency physicians should be aware of this disease and its complications.

Aetiology

CF is an autosomal recessive disease, with a carrier rate of 1 in 25. It is caused by mutations in a protein called the CFTR (CF transmembrane regulator), which is expressed in epithelial cells. The primary function of the CFTR protein is as an ion channel that regulates liquid volume on epithelial surfaces through chloride secretion and inhibition of sodium absorption.

Different mutations can have varying effects on CFTR function and thus cause differing phenotypes of disease, though the relationship between genotype and phenotype is highly complex and unpredictable.

The reduced volume of airway surface liquid in CF causes failure of mucociliary clearance, the lungs' innate defence mechanism. Secretions are retained and obstruct airways. Patients with CF also have an excessive respiratory response to pathogens and will develop significantly more inflammation from a respiratory tract infection than somebody without the disease – the reason for this is unknown. The end result is irreversible airway damage with bronchiectasis and respiratory failure in most patients.

Ion and water abnormalities may also cause disease in other epithelia-lined organs that also express the CFTR such as the gastrointestinal tract (together with the pancreas and biliary system), sweat glands of skin, and reproductive organs.

Diagnosis

The diagnosis should be suspected in:

- neonates presenting with meconium ileus
- infants with recurrent respiratory symptoms and failure to thrive
- older children with recurrent respiratory symptoms and finger clubbing.

The optimal diagnostic test for CF is the measurement of sweat electrolyte levels. Patients with the disease will have raised concentrations of sodium and chloride to >60 mmol L. Sweat testing is only performed in specialised secondary and tertiary centres.

Newborn screening programmes using the Guthrie blood spot test screen all newborns for CF. The initial screen is for raised concentrations of immunoreactive trypsinogen, with further testing as indicated. This screening is only about 95% sensitive, which means some cases will be missed. Antenatal screening is also offered. Early diagnosis and aggressive nutritional support improve growth and allow genetic counselling for the family; however, it may not improve pulmonary outcomes.

Management

Most patients have care coordinated by a tertiary CF centre, which has been shown to improve outcomes.

Management principles

- Segregation to prevent cross-infection:
    - Acquisition of some organisms, such as *B. cepacia* and *P. aeruginosa* are known to cause respiratory morbidity and accelerate respiratory decline.
    - Some of these organisms may be transferred from one patient with CF to another. Therefore children with CF should not share bays or waiting areas with other children with CF or with immunocompromised or other at-risk patients.
- Airway clearance techniques:
    - Physiotherapy, mucolytics, hypertonic saline, bronchodilators.
- Prevent respiratory infection:
    - Consider use of prophylactic antibiotics
    - Influenza vaccination.
- Treat infective exacerbations:
    - Oral or IV antibiotics appropriate for culture.
- Treat allergic bronchopulmonary aspergillosis (ABPA):
    - Prednisolone.
- Reduce airway inflammation:

- Macrolide antibiotics.

- Consider insertion of an indwelling IV access device:
  - Many patients requiring multiple courses of IV antibiotics will have poor venous access.
- Manage exocrine insufficiency (malabsorption, steatorrhoea, poor growth):
  - Supplement with pancreatic enzymes and fat-soluble vitamins.
- Manage constipation:
  - Dietary advice, laxatives.
- Manage insulin deficiency or diabetes:
  - Insulin, high-fat diet, occasionally oral hypoglycaemic agents
- Prevent osteopenia and pathological fractures:
  - Weight-bearing exercise, high dairy intake, vitamin D, consider bisphosphonates.
- Manage infertility:
  - Assisted fertilisation techniques.

Social and psychological support for patient and family:

Complications managed in the emergency department

Respiratory

Prompt and aggressive treatment of infective exacerbations is crucial to maintaining lung function, improving quality of life, and prolonging survival.[14]

History and examination

- Suspect an infective exacerbation in the patient presenting with:
  - change in sputum volume or colour
  - increased cough
  - increased lethargy
  - anorexia or weight loss
  - increased dyspnoea
  - chest pain (consider pneumothorax).
- Know results of recent sputum cultures: the presence or absence of particular colonising organisms (particularly *Pseudomonas*) will alter management.

- Consider aspergillus if there is new onset wheeze, pleuritic chest pain or new chest X-ray infiltrates.
- Ask about associated viral symptoms.

Investigations

- Chest X-ray looking for new infiltrates, pneumothorax or effusion
- Nasopharyngeal aspirate for viral aetiologies
- Sputum culture for bacterial aetiologies
- Pathology: FBE, UEC, CRP, BSL as baseline. LFT and IgE if indicated
- Lung function tests if available (particularly VC and FEV_1 to compare to baseline).

Management

- Oxygen and respiratory support as required
- The decisions surrounding antibiotic treatment (route, number of agents, choice of agent, duration of treatment) are complex and should be made in conjunction with the usual treating physician.
- Nebulised hypertonic saline and mucolytics may be beneficial
- Chest physiotherapy
- Nutritional support.

Gastrointestinal

Intestinal obstruction syndromes

Etiology

- Meconium ileus, distal intestinal obstruction syndrome (DIOS) and constipation are all due to increased viscosity of intestinal mucus and increased transit time in CF.[15]

Clinical

- Meconium ileus, unique to CF, is complete intestinal obstruction in the neonatal period due to accumulation of meconium. This presents with a clinical picture of bowel obstruction and no history of passing

meconium.

- DIOS is partial or complete intestinal obstruction due to faecal accumulation in the terminal ileum and proximal colon. Patients usually present with abdominal pain, distension, vomiting and a right lower quadrant faecal mass which is palpable or visible on plain X-rays. If obstruction is complete they may have bilious vomiting and small intestinal air-fluid levels on abdominal X-rays.
- Constipation may present in a similar way to DIOS, but the symptoms are usually milder and longer standing.

Management

- Meconium ileus requires referral to a paediatric surgeon who may treat conservatively with an enema or surgically.
- Most DIOS episodes are treated conservatively with intensive inpatient laxative treatment, with surgical intervention rarely required.
- Constipation can be managed with laxatives as an outpatient.

Endocrine

Cystic fibrosis–related diabetes

- The etiology of cystic fibrosis–related diabetes (CFRD) is complex and likely due to a combination of insulin deficiency and insulin resistance
- Patients with CFRD have an accelerated decline in clinical status, including lung function, and have a higher mortality
- Diagnosis is usually made via routine screening, usually with an oral glucose tolerance test
- It is usually treated aggressively with insulin, though sulphonylureas may be used to delay insulin therapy in some cases
- Diabetic ketoacidosis (DKA) is rare in CFRD, and if a CF patient presents to the ED with DKA he/she should be screened for type 1 diabetes mellitus (T1DM)

Table 1.2.4

Nomenclature

Gestational age	The elapsed time between the last menstrual period and the date of delivery
Chronological age	The time elapsed since birth
Corrected age	Chronological age minus the number of weeks premature

Table 1.2.5

Complications of prematurity

Neurological	Intraventricular haemorrhage Periventricular leukomalacia
Respiratory	Respiratory distress syndrome Chronic neonatal lung disease
Cardiac	Patent ductus arteriosus
Gastrointestinal	Necrotising enterocolitis Gastrointestinal reflux disease
Other	Developmental delay Growth reduction Hearing impairment Nosocomial infection Retinopathy of prematurity Iron deficiency anaemia Osteopenia

- Acute hypoglycaemia should be managed in the same way as in T1DM, with oral glucose if tolerated, or intramuscular glucagon or intravenous dextrose.

The ex-premature infant

Introduction

Advances in neonatal care have improved survival at the extreme of prematurity, and it is common to see ex-premature infants in the ED. Prematurity is defined as birth before 37 weeks' gestation, with extreme prematurity describing those born before 28 weeks' gestation. In Australia approximately 8% of babies are born prematurely, with extreme prematurity accounting for 1%.

Complications of prematurity

There are many complications of prematurity (Table 1.2.5). This chapter will focus on longer-term complications of prematurity relevant to ED care.

Chronic neonatal lung disease

Chronic lung disease (CLD) is defined as supplemental oxygen requirement past 36 weeks of gestational age and affects up to 40% those born at <28 weeks' gestation.[16]

Etiology

The etiology is usually multifactorial, including:

- Antenatal factors: arrested pulmonary development due to preterm delivery, ventilator-induced barotrauma and volutrauma, oxygen toxicity
- Complications, such as patent ductus arteriosus and lower respiratory tract infections.

These lead to small airways disease that may be also complicated by parenchymal and interstitial lung damage.

Clinical

Infants with CLD have tachypnoea, wheeze, cough, chest wall retraction and paradoxical respirations when they are well. These infants are more likely to develop lower respiratory tract infections and then are at increased risk for rapid deterioration and the need for extra oxygen and ventilatory assistance.

Management

The management of respiratory infections such as bronchiolitis and pneumonia in children with CLD is the same as for other children but with the appreciation that these children have the potential to rapidly become more unwell. Most will require admission to hospital for this reason. Liaison with the usual treating physician is recommended.

Nutrition

Low iron stores, decreased erythropoietin production, decreased red blood cell survival, infections and frequent venipuncture commonly lead to anaemia in this population; therefore, breast-fed preterm infants usually receive 2 mg kg day of supplemental iron between 6 weeks and 6 months of age.[17] Supplements cease

when formula or complementary foods supply the iron requirements.

Premature infants are also prescribed vitamin D, usually dosed at 400 IU daily, to prevent hypocalcemia and rickets. Adequate amounts of vitamin D are not provided by formula or breast milk.

Immunisations

- Premature infants are immunised based on chronological age according to routine schedules.
- To prevent complications from the respiratory syncitial virus (RSV) infection, palivizumab prophylaxis is recommended in the first year of life for all infants born before 29 weeks' gestation and for those born between 29 and 32 weeks' gestation with CLD.
- The influenza vaccine is also recommended for this high-risk patient group.

Development

Developmental and growth milestones are corrected for gestational age for the first 2 years of life.

Autism spectrum disorder

Overview

Autism is a complex developmental disability that affects a person's ability to communicate, understand language, play and socially interact with others. It has a prevalence of approximately 1% in the population. Autism is a spectrum disorder that affects every patient to a differing degree, though there are some common characteristics that may occur.

Characteristics of autism spectrum disorder

- Difficulties with verbal and nonverbal communication
- Deficits in social interactions
- Hyper- or hypo-reactivity to sensory input
- Stereotyped or repetitive motor movements, such as spinning or lining

up objects
- Difficulty with changes to surroundings or routines
- Challenging behaviours, such as aggression or self-harm.

Autism spectrum disorder in the emergency department

Individuals with autism spectrum disorder (ASD) have complex care needs with high rates of co-occurring physical and mental-health problems.[18,19] The ED can be an extremely stressful environment for these patients and their caregivers.

Children with ASD often find the experience of attending an ED frightening because of their inability to cope with change in routine or problems comprehending what is happening to them. Exacerbating this anxiety are the nature of the sudden event precipitating the ED visit, unpredictable waiting times, and sensory overload from the noise, lights and crowd in busy waiting rooms, plus the need to undergo investigations and treatment. Anxiety may become overwhelming and lead to withdrawal or behavioural outbursts, which makes examination and investigation particularly challenging.

A lack of understanding of ASD may mean that staff feel uncomfortable in treating these patients or even avoid them. Limited communication and anxiety make them difficult to assess, and common presenting complaints such as physical aggression or disruptive behaviour are sometimes perceived as ‘difficult’ or ‘unsatisfying’.

Management pearls

The goal is to minimise the anxiety for the child with ASD as much as possible, which will make assessment and management more timely and efficient. This may be achieved by:

- minimising waiting times. Waiting and the surrounding anxiety can provoke a behavioural disturbance, so give consideration to seeing these patients first regardless of triage category. If waiting is inevitable, aim to find a quiet area with minimal sensory stimulation where they can wait with a caregiver.
- recognising that proper assessment of individuals with ASD may require more time

- limiting the number of staff involved in the management
- letting the parent or caregiver help you. Obtain as much information from him/her as possible: functional level, specific sensitivities, and what may help to make the child feel safe and calm.
- using a quiet, calm voice, allowing the child extra personal space, and minimising words and touch
- preparing the child for procedures, breaking down actions required into small steps
- visually presenting information, such as demonstrating the use of objects and equipment, which may be better understood than verbal information and lead to increased chances of cooperation.

Summary

Increasing awareness of the special needs of children with ASD among health professionals can minimise the anxiety these children experience, resulting in a more positive encounter for everyone.

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SECTION 2

Resuscitation

OUTLINE

2.1

Paediatric cardiopulmonary arrest

Ben McKenzie, and Richard Aicken

ESSENTIALS

1 Cardiopulmonary arrest (CPA) can occur either as in-hospital cardiac arrest (IHCA) or as an out-of-hospital cardiac arrest (OHCA), but prevention of both is the key to decreasing paediatric deaths.

2 CPA in children usually results from the development of progressive hypoxia and/or shock, which may be due to a myriad of causes.

3 Paediatric patients who sustain OHCA have survival to hospital discharge rates similar to adults who sustain OHCA.

4 The goal is to identify evolving hypoxia and shock early in seriously ill children and proactively instigate appropriate therapy to prevent progression to cardiac arrest.

5 In contrast to adults, the most common arrest rhythm in children is non-ventricular fibrillation (VF)/ventricular tachycardia (VT) and is usually either asystole, pulseless electrical activity (PEA) or electromechanical dissociation (EMD).

6 Allowing parents to be present and supported during the resuscitation of their child may be associated with a better long-term psychological outcome.

7 Unless CPA is associated with drug toxicity or some situations with profound hypothermia, neurologically intact survival is unlikely after 30 minutes of CPR and several doses of adrenaline.

Epidemiology

The incidence of out-of-hospital cardiorespiratory arrest (OHCA) is reported as ranging from 2.6 to 19.7 per year per 100,000 paediatric population (age <18 years). Approximately 30% achieve return of spontaneous circulation (ROSC), 24% surviving to hospital admission, and 12% surviving to discharge.[1] Incidence of CPA appears unchanged over time.[2]

In Australia and New Zealand, 2% of paediatric intensive care unit (PICU) admissions are for cardiac arrest, of which 50% are for OHCAs and 50% for in-hospital events.[3] Approximately 10% of paediatric in-hospital cardiac arrests occur in the emergency department (ED).[4]

While survival from critical illness in paediatrics has improved significantly in recent years, improvements in survival rates from cardiorespiratory arrest have been more modest. Survival from paediatric cardiac arrest does, however, continue to improve in many (but not all) parts of the world, particularly for in-hospital events.

Previous studies of paediatric OHCA have reported poorer survival rates with severe neurological sequelae compared with adults;[5,6] however, more recent publications have challenged this, reporting similar rates of survival to hospital.[7,8]

Aetiology

The causes of cardiorespiratory arrest in children are diverse, but the final pathophysiological process is most commonly progressive tissue hypoxia and acidosis due to the development of respiratory failure or circulatory shock or both. The ECG finding in an arrested child is therefore usually bradycardia or asystole, reflecting the myocardial response to this final pathway of poor coronary perfusion and myocardial oxygenation. Children rarely have coronary artery disease and consequently have much lower incidences of ventricular tachycardia (VT) or ventricular fibrillation (VF) compared to adults.

Examples of conditions where circulatory shock is the primary pathology causing cardiorespiratory arrest are trauma, septicaemia, anaphylaxis and congenital heart disease causing heart failure or pulmonary hypertension. Examples of conditions causing primary respiratory failure and hypoxia leading to arrest are airway obstruction, asthma and other lung diseases. Not infrequently there is coexisting hypotension and hypoxia with both systems being affected in

the course of the pathological insult. Respiratory arrest may occur alone but if treated promptly may not progress to cardiac arrest.

With most aetiologies, there are preceding symptoms and a degree of physiological compensation such as tachycardia and tachypnea before the physiological limit of the child is reached. Once this threshold is passed, there is decompensation with bradycardia and/or respiratory arrest with subsequent loss of cardiac output. There has been considerable clinician focus in recent years on recognising and responding to deterioration before decompensation and preventing cardiac arrest in healthcare settings (see below).

Approximately 10% of paediatric OHCA victims have a shockable rhythm on arrival of the pre-hospital care providers.[7,8] The highest incidence of VT and VF is in the adolescent age group. Examples of conditions causing primary arrhythmia are ion channelopathies, congenital heart disease and poisoning.

Preventing cardiac arrest

Paediatric cardiac arrest occurs in the ED and other hospital settings where there is usually preceding physiological deterioration before the arrest event. Being able to recognise and respond to clinical deterioration is one of the core skills in emergency medicine.

Many healthcare organisations have instituted mechanisms to systematise the recognition (afferent limb of system) and response (efferent limb) to clinical deterioration with the intention of preventing cardiac arrest. Medical emergency teams (MET) and rapid response teams (RRT) are examples where there is a systematised efferent limb of escalating clinical care in response to the afferent input of detecting physiological deterioration. Formal MET and RRT systems require significant resources and can be expensive to implement.

The impact of MET and RRT on rates of cardiac arrest have been difficult to assess due to the observational nature of studies, low frequency of cardiac arrests, and confounding variables, such as concomitant clinical system improvements.

The International Liaison Committee on Resuscitation (ILCOR) guidelines recommend the institution of MET/RRT with a very weak supporting level of evidence. The guideline places significant and greater emphasis on the ability to recognise and respond to deteriorating illness rather than mandating a formal MET/RRT structure.

Systems which focus on the recognition of early clinical deterioration have

been adopted by some institutions. One such system is the Paediatric Early Warning System (PEWS), which assigns numeric scores to specific abnormal observations in several clinical domains that then trigger a response. The impact of this model is still being evaluated. Other strategies of systematically detecting early clinical deterioration include colour-coded, age-specific, vital-sign-observation charts that visually identify physiological parameters in a predefined range of clinical concern.

EDs in many jurisdictions have implemented paediatric clinical deterioration recognition and response systems either internally or as part of a whole hospital response; however, this is an area that is as yet little researched.

Outcome

Generally, the survival from respiratory arrest alone is much better than from CPA. Survival to discharge for children with respiratory arrest (pulse present) is around 75%, and of these up to 88% have a good neurological outcome. Cardiac arrest in children is a catastrophic event with a low survival rate that varies substantially between in- and out-of-hospital cardiac arrests as well as precipitating pathology. Survival to discharge from hospital for paediatric OHCA is 7%[7,8] and 36% for in-hospital cardiac arrests.[9] 'Survival to discharge' is a very crude marker of 'success' as it does not include a measure of neurological function. Limited data for neurological outcome exist but for in-hospital cardiac arrest, favourable neurological outcome rates are in the vicinity of 20% (lowest for trauma, highest for cardiac surgery patients).[4]

Unfortunately, the perception of the public, and even doctors and nurses, is that the expected survival rate is higher. Lay rescuers, physicians and nurses estimate the survival rate for CPA in children as being 63%, 45% and 41%, respectively (compared to 53%, 30% and 24% for adult CPA).[10] Undoubtedly, fictional medical television programmes contribute to this bias, and even non-fictional medical programmes rarely show death as an outcome.

Differences compared to adults

When comparing children to adults in relation to CPA, there are several important differences. The aetiology of the event is usually different. Adults who collapse are more likely to have VF or pulseless VT; hence the time to defibrillation is the single greatest determinant of survival. Thus the emphasis on

defibrillation and chest compressions is not applicable to most infants and children. Instead, there is an emphasis on airway and breathing, and this is reflected in breath-to-compression ratios (see Chapter 2.2 on Basic life support).

There are several anatomical and physical differences between children and adults. It is important to consider these differences in relation to the primary event leading to arrest and to the resuscitation techniques subsequently required (Table 2.1.1).

Development of resuscitation guidelines

The ILCOR has become the accepted peak body that informs international resuscitation guidelines. It attempts to systematically answer clinical questions and make treatment recommendations using available scientific data, regularly publishing their consensus findings.

The timeline of formation of ILCOR started in 1992 when the American Heart Association (AHA) guidelines for resuscitation were published. Subsequently representatives of seven resuscitation councils throughout the world, including the Australian Resuscitation Council, formed ILCOR, and advisory statements were produced. A subcommittee on paediatric resuscitation with representation from the AHA and other paediatric representatives from ILCOR further developed guidelines for paediatric patients. Ultimately the *International Guidelines 2000* were published and most recently in October 2015 as the ILCOR 2015 Consensus of Science with Treatment Recommendations (CoSTR).[11] The Australian Resuscitation Council (ARC) and New Zealand Resuscitation Council (NZRC) released updated guidelines in January 2016 to incorporate the recommendations. Subsequently resuscitation courses, like the Advanced Paediatric Life Support (APLS) course, have been amended to be consistent with the international recommendations. APLS courses in Australia and New Zealand are consistent with ARC and NZRC recommendations.

Ethics of paediatric resuscitation

Presence of family

Over recent years, the benefit of allowing the family, mainly parents, into the resuscitation room during active resuscitation has become clearer. This practice has required a cultural change and mandates appropriate and professional behaviour and language during resuscitation. Such practice should occur

regardless of the presence of any ‘outside’ witnesses. It is a professional standard that all medical, nursing and other health professionals should aspire to and maintain. It is probably even more important that parents are offered the opportunity to witness the resuscitation, when the outcome is the death of their child.

Table 2.1.1

Important differences between children and adults

Difference in children	Implication
Airway	
Prominent occiput tends to cause neck flexion	Neck extension, into a neutral or sniffing position (slight extension), is required to optimise the airway for an infant or child respectively
Mandible is relatively smaller	More difficult intubation
Tongue is relatively larger	Tends to obstruct airway More difficult intubation
Larynx is more cephalad (located almost at base of tongue)	More difficult intubation – tendency for inexperienced operator to insert laryngoscope blade into oesophagus
Epiglottis is proportionally larger and more ‘floppy’	Intubation may require straight-bladed laryngoscope to lift epiglottis forward to allow visualisation of vocal cords
Upper airways are more compliant (i.e. distensible)	Tend to collapse during increased work of breathing
Breathing	
Chest wall more compliant (particularly the newborn infant and more so the preterm infant)	Less efficient ventilation, when increased work of breathing Earlier fatigue
Greater dependence on diaphragm to generate tidal volume	Distended stomach impairs ventilation Importance of venting stomach with gastric tube
Circulation	
Maintains cardiac output and blood pressure by tachycardia initially	Diagnose and treat shock before hypotension develops Hypotension usually indicates decompensation
General	
Head has proportionally greater component of body surface area	Loss of body heat during primary event or resuscitation Greater chance of head injury
Compliant chest wall allows transmission of energy to underlying organs, resulting in traumatic damage/rupture, rather than dissipation of energy	Pulmonary, hepatic and splenic injury may occur without overlying rib fractures
Development Language Motor development (fine and gross) Social and cognitive development (including abstract thinking)	Must be considered when interacting with the child and understanding injuries (accidental versus non-accidental)
Parental and staff considerations	Psychosocial issues Presence of family during resuscitation Staff pressure to continue resuscitation Impact on staff from death of child

In hindsight, parents who have witnessed the resuscitation of their child have valued this opportunity and despite the occasionally chaotic environment, common positive perceptions relate to the efforts made by staff. To achieve a positive (psychosocial) outcome, in relation to the presence of parents during resuscitation, the following issues should be considered:

- Parents being present during the resuscitation of their child should be regarded more as the norm than the exception.
- Parents should not be coerced into being present, but gentle encouragement and explanation by a senior member of staff are usually helpful.
- Senior members of staff engaged in resuscitation should not feel undue pressure as a result of the attendance of parents. Occasionally, however, it may be preferable for the parents to wait outside the immediate

resuscitation area during challenging or visually disturbing procedures.

- Parents should have a dedicated support person, being a health professional, to remain with them throughout the resuscitation. The support person should obtain information from medical staff to keep the parents informed of their child's progress and explain procedures to the parents.
- Touching and talking to their child is often important for the parents where this is feasible.
- Parents need to have a quiet room to retreat to, if necessary. Discussion with family should not occur in a busy ED corridor as it will negatively affect the family's retention of information.
- Family requests and religious beliefs need to be respected.
- Obligations mandated by Coronial Law need to be explained with sensitivity.
- The emotional impact on all staff members when a child dies is considerable, and this may be compounded by the distress of parents during and following the resuscitation.
- With any resuscitation, particularly those resulting in death, debriefing of staff is essential, as is organising the appropriate support and medical follow-up of the parents.

Termination of resuscitative efforts

The decision to cease resuscitation efforts in a child in CPA is influenced by many factors, and there is no absolute consensus about when resuscitation should stop.

Considerations include the total arrest time, clinical response to therapy, premorbid state of the child, potential for any reversible factors, likely neurological outcome, information from colleagues who care for a child with long-term medical problems and the parental wishes. In the initial stage of the arrested child arriving in the ED, these details should be rapidly established whilst resuscitation is continued in order to help guide subsequent management.

Termination of resuscitation in a newly born baby is likely to be appropriate if the baby remains in cardiorespiratory arrest at 15 minutes. Even after 10 minutes of documented asystole, survival without severe disability is unlikely.

For children in established cardiac arrest the overall outcome is poor. If the child requires adrenaline (epinephrine) and fails to respond to two doses, then

survival is unlikely. Generally no longer than 30 minutes of advanced life support resuscitation is required to determine whether discontinuation of resuscitation is appropriate. Recurring/refractory VF or VT, toxic drug exposure or the presence of significant hypothermia in the setting of ice-cold immersion is a situation that may require more prolonged resuscitation efforts (see Chapter 22.2 on Drowning). Many children in cardiorespiratory arrest in Australia and New Zealand who are hypothermic, however, have lost body heat due to exposure, without spontaneous circulation after the arrest, and therefore this is unlikely to be neuroprotective.

The decision to stop resuscitative efforts in the ED setting due to a lack of return of spontaneous circulation is a medical decision. It is not a choice that is offered to the parents.

Non-initiation of resuscitative efforts

Resuscitation should not be initiated if there are signs of prolonged death, like rigor mortis or post mortem lividity. Children who are in a palliative phase of care may have an 'end of life care plan' that includes non-initiation of resuscitation should this have been previously determined by family and the child's coordinating specialist. Children with complex and disabling conditions will occasionally have life-threatening events occur. If prior discussions between parents (and child if appropriate) and physicians have not included limitations of interventions or medical support, then it is difficult to avoid commencing resuscitation. Early and urgent consultation with the child's primary physician is prudent in these situations. Physicians should not be coerced into undertaking care that they believe is morally wrong or fruitless.

There are specific situations in the newly born baby that may lead to the non-initiation of resuscitation, like extreme prematurity and congenital/chromosomal abnormalities not consistent with long-term survival. This issue is covered in Chapter 2.6 on neonatal resuscitation.

Non-accidental injury

It is important that all healthcare providers involved with children should be aware of their statutory obligations under state or federal law regarding notification of suspicion of non-accidental injury or neglect. Awareness of development of the child, feasibility or compatibility of the history with clinical

signs and any delay in presentation should be considered when assessing this possibility. Regardless of the outcome of the resuscitation, if there is a suspicion of abuse or neglect, appropriate notification should be made.

Organ donation

Organ donation is usually a consideration for staff when a child is in an intensive care unit and generally not in the ED. The only situation where it may be contemplated in the ED is when post mortem organs (like corneas, heart valves) may be retrieved for this purpose. In cases mandating notification of the coroner, then permission is required prior to any organ harvesting. Liaison with the appropriate local donor coordinator is essential. A senior member of staff should have this discussion with the parents, and occasionally the parents themselves raise the issue.

If resuscitation has resulted in the return of spontaneous circulation but with likely brain death, subsequent organ donation may be feasible. The determination of brain death would usually occur in an intensive care unit, as a repeat examination for brain death, after a period of time, is required. Consultation with and consideration of transfer to a paediatric intensive care unit are required. The focus in the ED at this point is the ongoing care of the child and support of the family.

Death certificates, notification to the coroner and other legal issues

Death in the ED is, by itself, not an indication to notify the death to the coroner. Coroners' Acts vary from state to state, and staff must be aware of their statutory obligations. Remember that if a death is to be notified to the coroner the body then becomes evidence and should be left intact at the termination of resuscitation. Common practices of taking hand/footprints, locks of hair, and removing catheters and tubes must not occur or staff risk being held in contempt of court. Such mementos can be collected following the post mortem examination. This usually requires liaison with staff in the forensic mortuary.

Some states have special processes in place for the management of sudden unexplained death in infancy (SUDI). Any sudden and unexplained death under 12 months of age warrants a detailed history as well as specific samples collected post mortem to identify metabolic and genetic conditions, and this

should be clarified with the local forensic or coronial service.

In the situation where a death does not require notification to the coroner, it must be clarified who can and will complete the death certificate, which includes the cause of death. Consultation with the primary physician involved with the long-term care of a child with chronic illness is obligatory. Medical staff need to be aware of other legal obligations, like the collection of blood alcohol specimens in pedestrian and vehicular accidents, but these would only be collected ante-mortem by hospital staff. These legal requirements vary between states.

Child death – follow-up of family

It is essential that family members are managed by a senior member of staff in a compassionate and sensitive manner. Parents should be given an opportunity to spend time comprehending what has happened to their child and to say goodbye. They should be able to leave the ED understanding what has occurred and what the next steps will be. Ability to retain information may be limited by grief, and clear communication is required. Written information including contact numbers should be given where appropriate.

Parents require a clear understanding of what will happen to their child in either a coronial or non-coronial process. They also need to know who will contact them next and who they can contact to find out further information. Potential sources of support should be identified if possible, such as family, friends, pastoral care, hospital or other community services available locally.

Their GP or usual healthcare provider must be informed immediately and be identified as a potential support person for the family. The GP should also be aware of the potential need for medical screening of other family members is raised (e.g. ECG) at the appropriate time in the future so this opportunity is not forgotten.

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2.2

Paediatric basic life support

Jane Cocks

ESSENTIALS

1 Paediatric resuscitation is different to adult resuscitation in many aspects, but the basic principles remain the same:
- Don’t panic
- Ensure both the patient and rescuer are safe
- Get help
- Initiate basic life support at the earliest possible moment: Airway, Breathing, Circulation.

2 **DRSABC** is a mnemonic to aid the sequence of events that should be followed when met with any collapsed patient. Check for **D**anger to self and patient, check for patient **R**esponsiveness, **S**end for help, open the **A**irway, check for **B**reathing, and if patient is not breathing normally give two rescue breaths; assess **C**irculation by checking for signs of life and if absent commence Cardiopulmonary resuscitation (**C**PR).

3 Quality chest compressions involve compressing the lower half of the sternum to at least one-third of the anteroposterior chest diameter at a rate of 100–120 compressions per minute with subsequent complete release to allow recoil of the chest wall.

4 Basic life support rescuers and lone rescuers should use a 30:2 compressions to ventilation ratio, while advanced life support rescuers performing two-rescuer CPR should use a 15:2 compressions to ventilation ratio for all children, except in the newly born infant where it is 3:1.

Introduction

Basic life support (BLS) or basic cardiopulmonary resuscitation (CPR) is a process by which basic cardiac and respiratory functions can be restored and maintained through a combination of rescue breaths and chest compressions. The provision of BLS sustains the vital functions of the collapsed individual without the need for specialised equipment, minimising the potential hypoxic damage that may occur while advanced life support is being activated.

Paediatric versus adult basic life support

The aim of basic life support in all age groups is the same, and the techniques utilised follow the same general principles, but paediatric BLS differs significantly from adult BLS in some very fundamental ways.

Aetiology of arrests

In children, the most common causes of cardiopulmonary arrest are hypoxic, and it is essential that the initial focus of BLS in children and infants is in establishing effective ventilation. Most children with hypoxia undergo a period of deterioration with worsening bradycardia preceding the final arrested state. It is important that paediatric BLS is commenced as soon as this bradycardic deterioration is noted and is not withheld until the patient becomes fully arrested and pulseless.

The currently accepted international recommendation for the commencement of chest compressions in children of all age groups who are unresponsive with no spontaneous breathing is a heart rate below 60 beats per minute (bpm).

Anatomy and physiology

The anatomy and physiology of the human body change markedly with age, progressing from the immature neonatal form to the more adult form of the older child, and these changes are relevant to the provision of BLS at any particular age. The following differences from the adult form are of direct importance in the provision of BLS in children:

- *Airway*: Large head (particularly the occiput) and short neck leading to neck flexion; large tongue which is predominantly intra-oral; easily

compressible oropharyngeal soft tissues; larynx lies more anteriorly and higher (C2–3); cricoid ring is the narrowest part of the airway; soft and short trachea.
- *Breathing*: Increased respiratory rate; more compliant chest wall; dependence on diaphragmatic breathing; reduced end-expiratory lung volume.
- *Circulation*: Increased heart rate; more dependence on heart rate for delivery of adequate circulation; small absolute volume of circulating blood (70–80 ml kg).
- *Metabolism*: Increased metabolic rate; increased body surface area (BSA).

Basic life support techniques and age

BLS techniques are modified depending on the child's age and size to ensure that they will be maximally effective. In view of this, children are arbitrarily divided into the following internationally accepted age groups:

- Newborn/neonate: birth to 1 month old
- Infant: between 1 month and 1 year of age
- Small child: age between 1 and 8 years
- Large child: age older than 8 years.

Preparation and equipment

Basic CPR requires no extra equipment other than personnel trained in its administration. It is advantageous, however, to have available certain basic equipment to assist in the resuscitation process:

- Portable suction and suction catheters, for clearing secretions, improve the ability to achieve and maintain a patent airway.
- An oropharyngeal airway (OPA) may assist in achieving a patent airway.
- A self-inflating bag and mask, suitable for the size of the patient, improves artificial ventilation and reduces the risk of cross-infection.
- Oxygen for ventilation will further reduce the risk of hypoxic injury.

Basic life support sequence

A ‘DRSABC’ approach

It is extremely important to ensure maximum rescuer and patient safety as a priority on first approaching the collapsed child. **DRSABC** is a mnemonic to aid the sequence of events that should be followed when met with any collapsed patient:

Danger: check for dangers or hazards that may affect you or the patient.
Responsiveness: check if the patient is responsive.
Send: send for help.
Airway: open the airway.
Breathing: check for breathing; if patient is not breathing normally give two rescue breaths.
Circulation: assess circulation by checking for a pulse and/or for signs of life; if the absent or inadequate pulse or no signs of life, commence CPR.

Check for Danger (D)

Ensure both the patient and rescuers are safe. This may involve removing obvious hazards from the direct environment or even moving the patient to a safer location prior to commencement of BLS. Endangering your own life will not benefit your patient if you are hurt or injured, so always ensure safety is a high priority.

Assess responsiveness (R)

Assess the responsiveness of the patient by gently, but firmly, stimulating the patient and asking if he/she is OK. It is important to remember the possibility of cervical spinal injury and, if this is likely, stabilise the cervical spine by placing a hand on the patient’s forehead prior to gently shaking his/her arm.

Send for help (S)

The activation of the emergency medical services (EMS) should be performed as early as possible in the sequence of resuscitation, provided there are adequate numbers of bystanders to allow this to occur without delaying the commencement of BLS. Most paediatric arrests will have an underlying hypoxic cause, and therefore any delay of BLS will significantly reduce its effectiveness. Commencement of BLS should therefore precede activation of EMS for a lone

rescuer in an unwitnessed collapse.

Note that in a witnessed sudden collapse of a child, the possibility of a sudden cardiac event is higher, and therefore activation of the EMS should occur immediately prior to commencement of BLS by a lone rescuer. These cases require rapid access to advanced cardiac life support measures, as is the case with most adult arrests.

Airway (A)

Open and maintain the airway

Position yourself at the patient's head, open the mouth and remove any obvious debris. Do not use a blind finger sweep in children as this may damage the delicate palatal tissues or move a foreign body further into the airway. Suction using a large-bore Yankeur catheter is useful for removing vomitus and secretions, preferably under direct vision.

Three manoeuvres will assist in opening and maintaining the airway (Figs 2.2.1, 2.2.2 and 2.2.3), which is most commonly obstructed by the child's tongue.

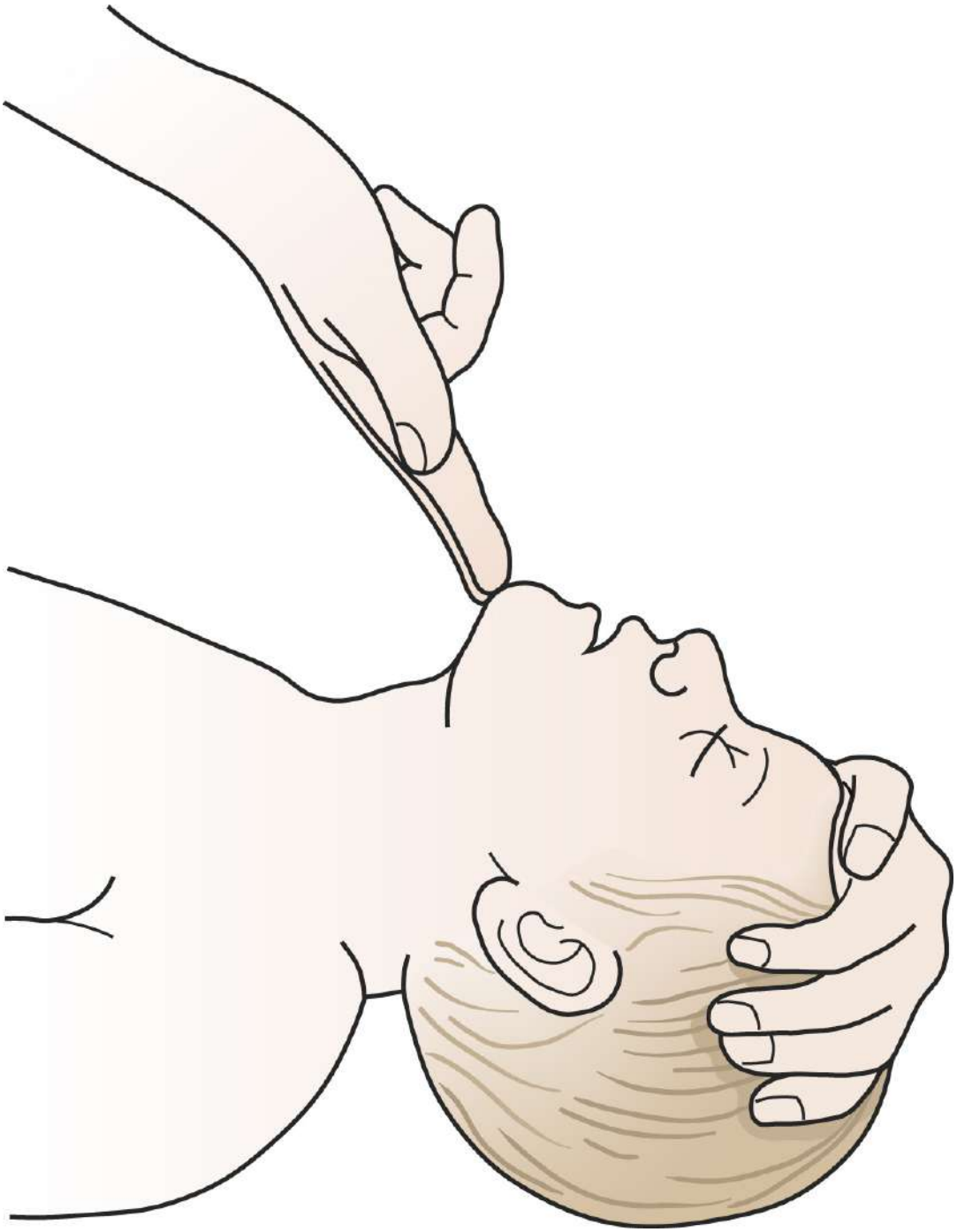

FIG. 2.2.1 Head tilt and chin lift in children.

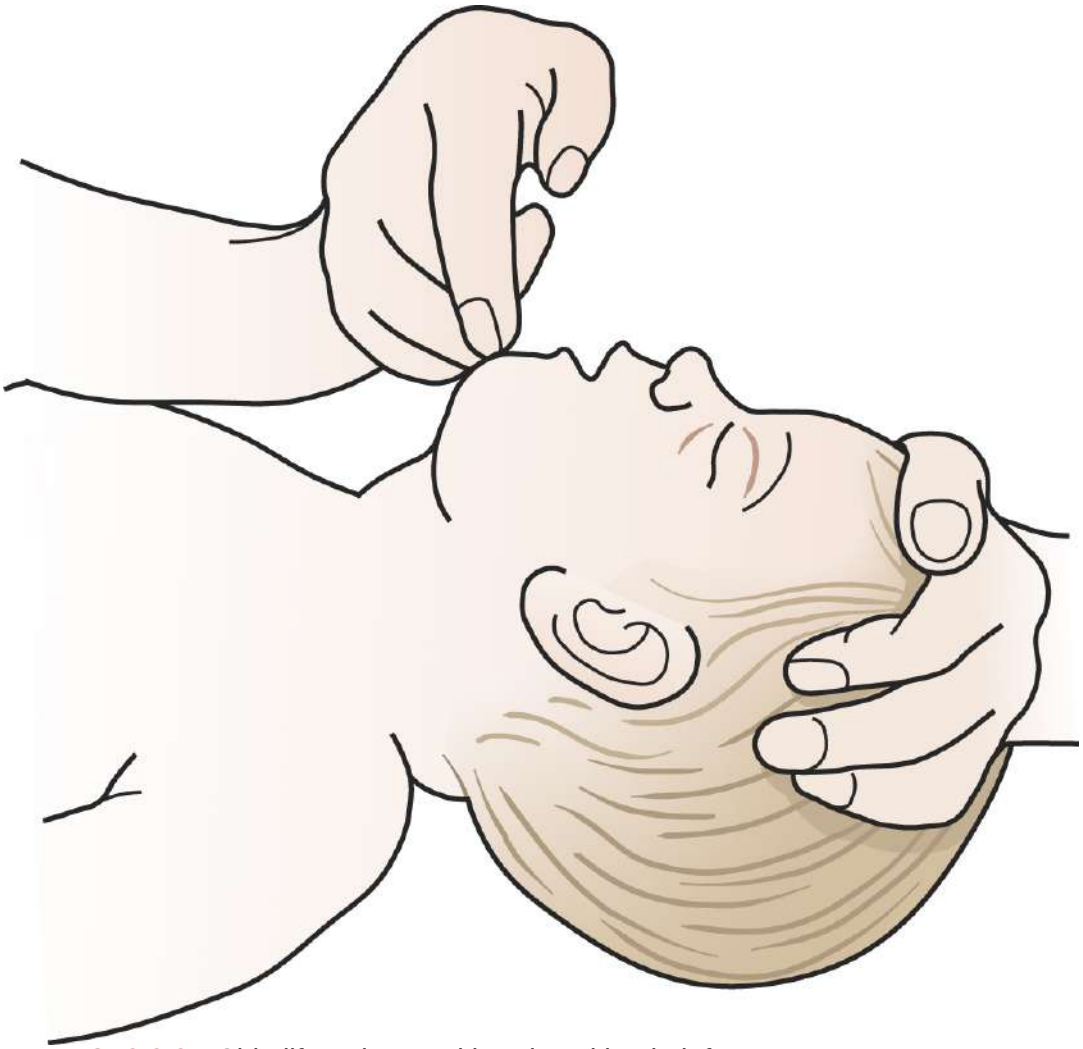

FIG. 2.2.2 Chin lift and neutral head position in infants.

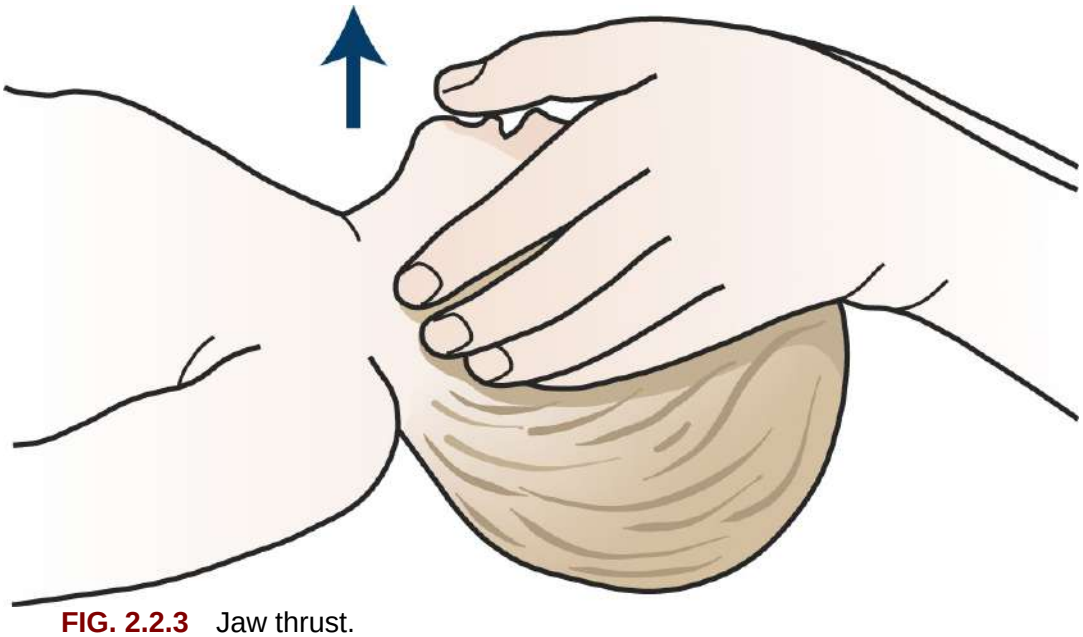

FIG. 2.2.3 Jaw thrust.

Head tilt

Place one hand on the patient's forehead and gently tilt the head back. In children the 'sniffing' position is desired, as in adult resuscitation. In an infant or neonate, a neutral position is required as they have a relatively large occiput and are naturally positioned in a sniffing position. A small towel placed under the infant's shoulders will eliminate the excessive neck flexion caused by the prominent occiput. If there is any possibility of cervical spine injury, the head tilt manoeuvre should be avoided.

Chin lift

Placing the fingers of the other hand on the jawbone, lift the chin, taking care not to compress the soft tissues of the floor of the mouth under the jaw. This manoeuvre is used in conjunction with head tilt.

Jaw thrust

Place two or three fingers under the angle of the mandible bilaterally, and lift the jaw upwards. This technique can be performed as an alternative to head tilt and jaw thrust and is indicated particularly if the possibility of cervical injury exists.

The patency of the airway can then be assessed by *looking* for chest movement, *listening* for breath sounds and *feeling* for exhaled breath. This assessment is best achieved from the patient's side, by placing an ear over the

patient’s mouth and nose, whilst watching the chest.

Airway adjuncts

Insertion of an oropharyngeal airway (OPA) may assist in maintaining patency of the airway if the above manoeuvres are inadequate. An appropriately sized oropharyngeal airway should reach from the central incisors to the angle of the jaw. In infants and small children, the oropharyngeal airway should be inserted under direct vision ‘the right way up’; that is, it should be inserted concave down in the position in which it will sit in the oropharynx. Gentle depression of the child’s tongue with a spatula will help facilitate airway placement. In children older than 8 years, the usual adult oropharyngeal airway insertion technique may be used. This involves inserting the airway concave up and then rotating it through 180 degrees to sit snugly in the oropharynx.

Oropharyngeal airways will not be tolerated in conscious or semi-conscious patients because of gagging. If this occurs, the airway should be removed. In this situation a nasopharyngeal airway (NPA) may be a useful alternative in the patient who is not completely obtunded. An appropriately sized nasopharyngeal airway should reach from the tip of the nose to the tragus of the ear.

Cervical spine

Be aware of the risk of cervical spine injury if the collapse is following a motor vehicle accident (MVA) or other form of significant trauma. If this is likely, stabilise the cervical spine by placing one hand on either side of the head and maintaining the head in line with the body at all times. CPR in this case generally requires two operators to perform successfully. If a second operator is not available, attempt to immobilise the cervical spine between sandbags or in a hard cervical collar before proceeding. Note that opening and maintaining an airway and providing effective CPR takes precedence over cervical spine immobilisation.

Breathing (*B*)

Once the airway is opened and patent, if the patient is not spontaneously breathing, deliver two to five rescue breaths. Each breath is delivered slowly over 1 to 1.5 seconds’ duration (inspiratory phase), and up to five may be required to ensure that two effective breaths are delivered.

Rescue breaths are most commonly performed as ‘mouth-to-mouth’ but may also be delivered using ‘mouth-to-mouth-and-nose’ in the smaller child. In the

‘mouth-to-mouth’ technique, the rescuer seals his or her mouth over the mouth of the patient, pinching off the nose with the free hand, whilst maintaining the patency of the airway with head tilt and chin lift. The ‘mouth-to-mouth-and-nose’ technique may be necessary for the infant or small child, and in that case the rescuer’s mouth should seal around the infant’s mouth and nose. In the hospital setting the rescue breaths will be delivered utilising bag and mask ventilation (see Chapter 2.3, Paediatric advanced life support).

Ensure that the degree of chest excursion is frequently reassessed during the rescue breaths. The chest must be seen to rise as if the child were taking a deep breath. Excessive tidal volumes or force may cause gastric dilatation and regurgitation. If there is no chest movement, the most likely cause is an obstructed airway due to poor positioning of the child’s head. Reposition the patient using the above manoeuvres and retry. If there is still no chest movement, there may be a foreign body obstructing the airway, which can be removed with suction or forceps under direct vision (see below).

Circulation (*C*)

Following the initial two rescue breaths, assess the circulation. Although the pulse check has always been considered the gold standard of circulation assessment, for non-healthcare professionals, the assessment of pulse has both poor sensitivity and specificity and often delays the decision to commence chest compressions. Current recommendations, therefore, suggest that lay rescuers assess for ‘signs of circulation’, specifically the presence of normal breathing, coughing or movement in response to rescue breaths.

Healthcare professionals may check for a pulse as well as assessing for signs of circulation. The pulses that are easiest to feel are the carotid, brachial or femoral pulses, and they should be palpated for no longer than a period of 10 seconds. The carotid pulse is difficult to feel in small children who have relatively short necks. If there is no pulse or severe bradycardia (heart rate <60 bpm) with signs of poor perfusion, then chest compressions on the lower half of the sternum should be commenced.

Chest compressions

The technique of providing chest compressions varies with the patient’s age:

- In infants and neonates, compressions are best performed with the rescuer encircling the infant’s chest with both hands and providing

compressions with the thumbs over the lower half of the sternum, avoiding the xiphisternum. An alternative but less effective technique involves the rescuer using two fingers to provide compressions at the same landmark site (Figs 2.2.4 and 2.2.5).

- In all other children, compressions are performed with the heel of one or two hands over the lower half of the sternum, avoiding the xiphisternum (Figs 2.2.6 and 2.2.7).
- The emphasis in cardiac compressions is to minimise interruptions. The depth of compression is relative and should be one-third to one-half of the anteroposterior diameter of the chest in all age groups. This will be approximately 4 cm in an infant and 5 cm in a small child. The chest should be seen to fully recoil before the next compression is commenced. Chest compressions should be performed on a firm flat surface.

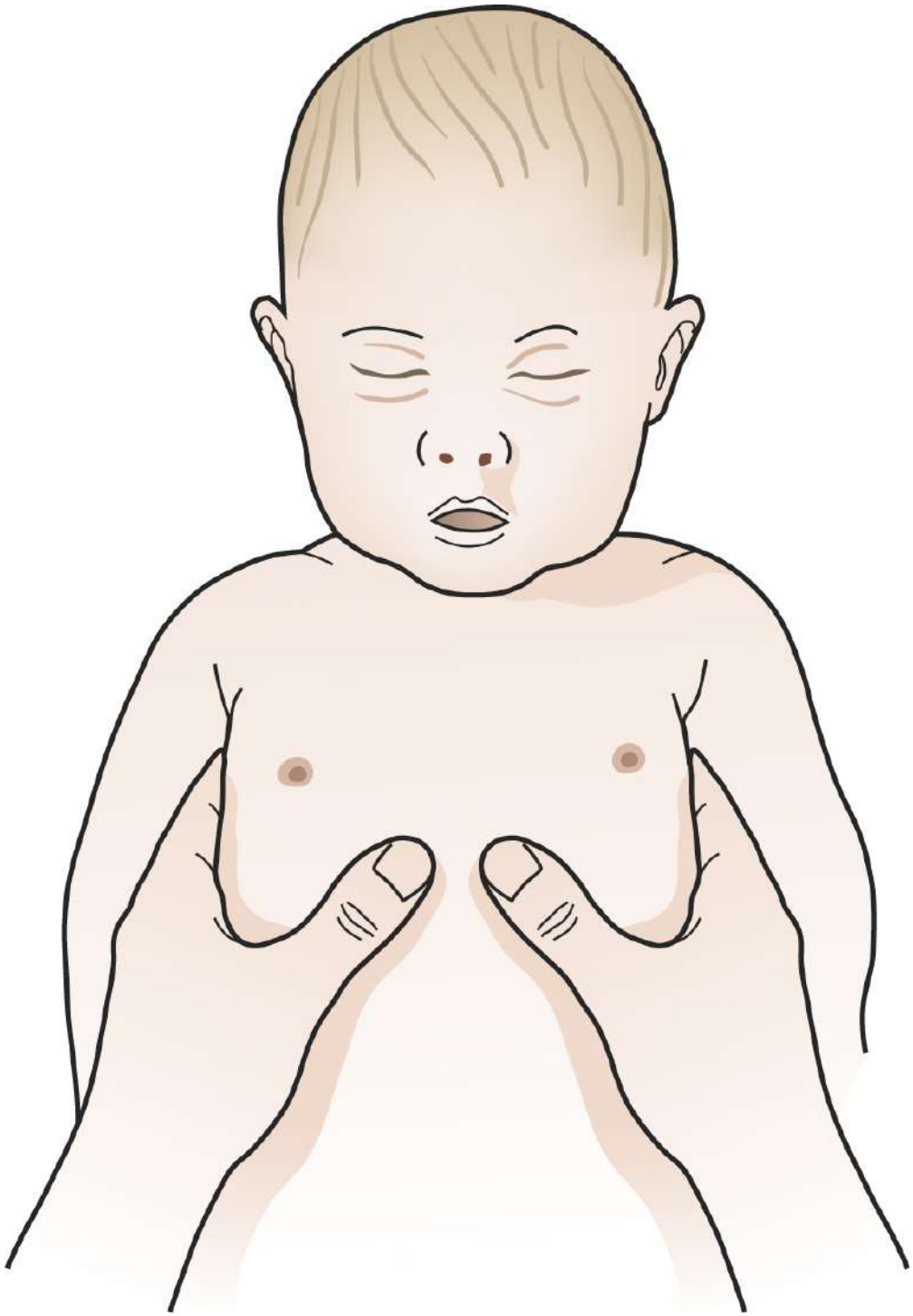

FIG. 2.2.4 Infant with two-thumb technique of chest compression.

- The rate of compressions is 100–120 per minute for all age groups, except in neonates, where it is 120 per minute. Note that this is the rate or speed of compressions, not the actual number delivered per minute. This rate will deliver approximately 75–90 compressions and 10 breaths

per minute.

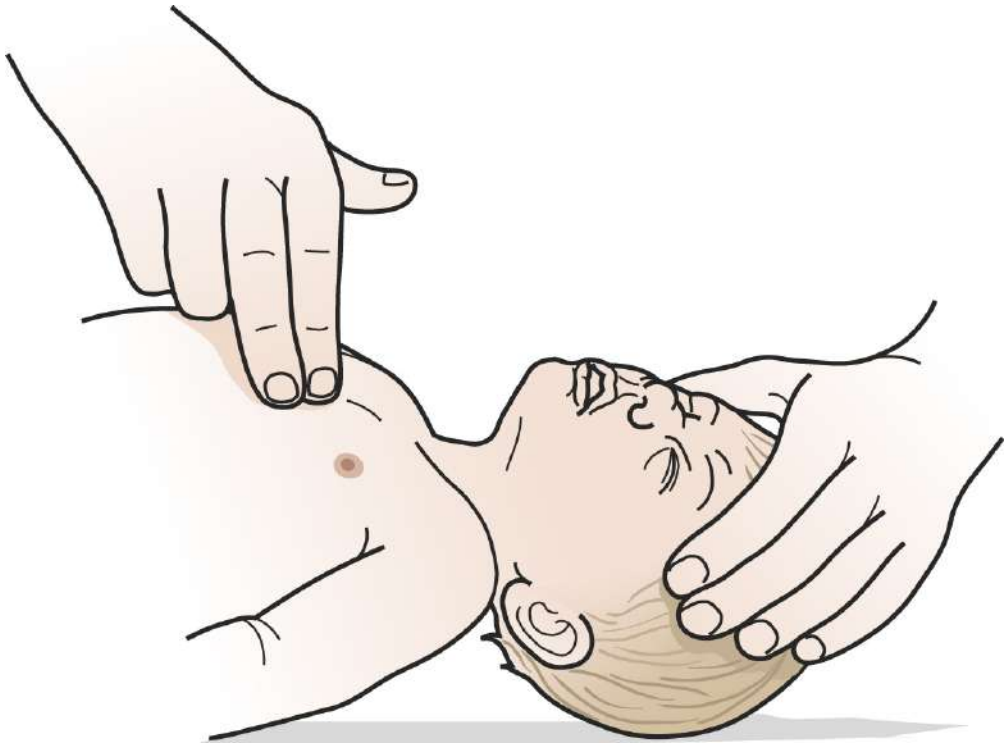

FIG. 2.2.5 Infant with two-finger technique of chest compression.

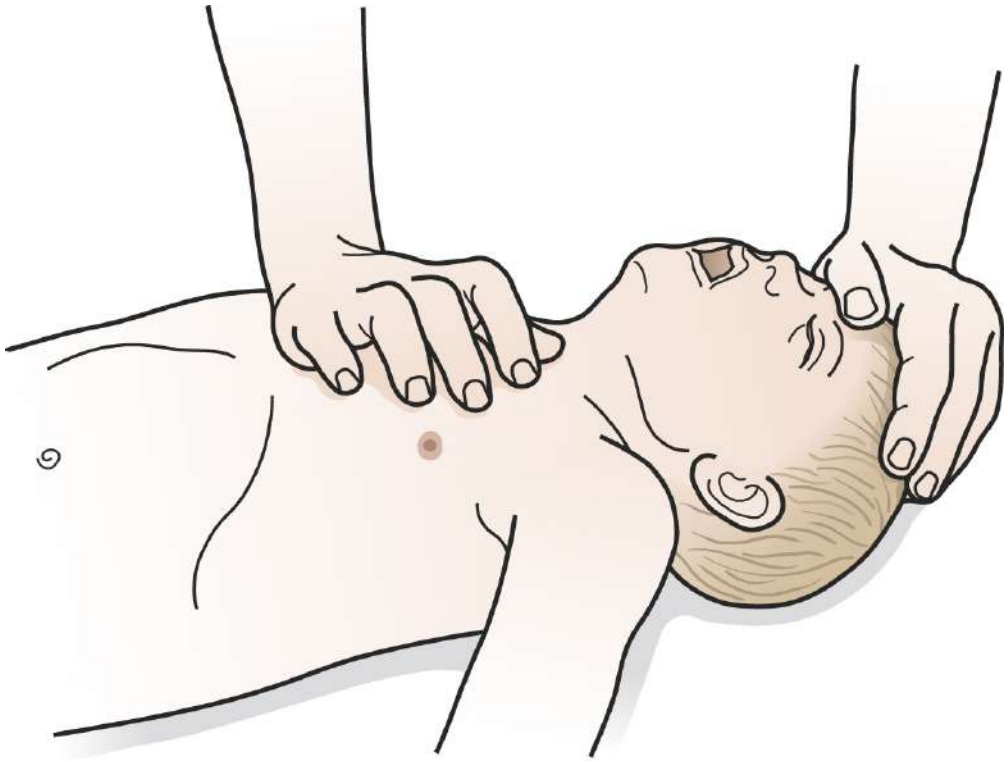

FIG. 2.2.6 Child with one-hand technique of chest compression.

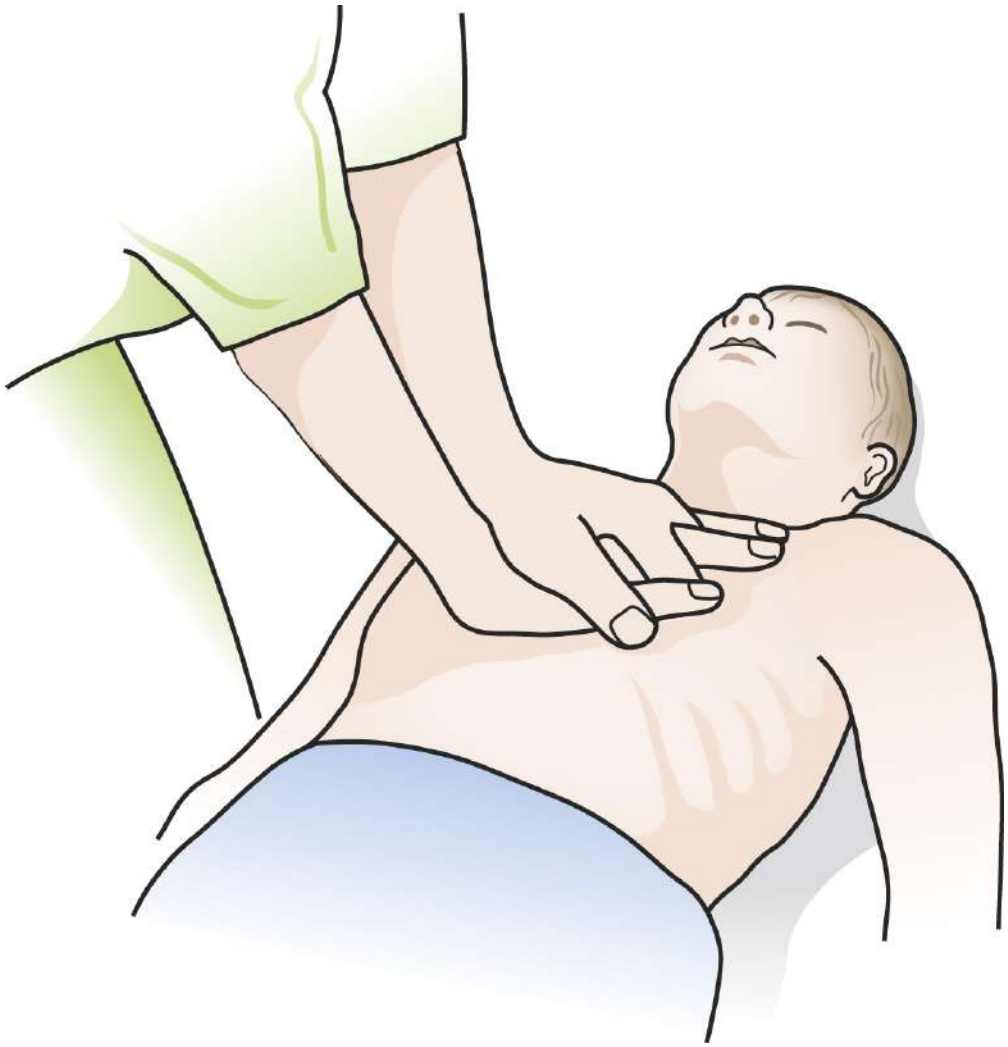

FIG. 2.2.7 Child with two-hand technique of chest compression.

Compression to ventilation ratio

For BLS rescuers or a lone rescuer, external cardiac compressions should be combined with rescue breaths in a ratio of 30 compressions to 2 ventilations for all age groups (except neonates – see below). Advanced life support rescuers performing two-rescuer BLS should use a ratio of 15 compressions to 2 ventilations for all infants and children (see Table 2.2.1). Note that the ratio of 30:2 is utilised in adults regardless of the number of rescuers.

Neonates require a combination of chest compressions to ventilation at a ratio of three compressions to every one ventilation and a rate of 120 ‘events’ per minute. This rate aims to achieve approximately 90 compressions and 30 breaths per minute.

It is important to check that the chest rises normally when the ventilation is

provided to ensure that an effective breath is being delivered. When two rescuers are delivering BLS, the rescuer providing chest compressions should pause compressions for the delivery of the breath. Once the airway is secured with intubation, this pause in chest compressions is no longer required.

Any interruption to BLS measures should last no longer than 10 seconds, after which BLS should be promptly recommenced to optimise outcome.

Mechanical devices to provide chest compressions during CPR have been designed and tested only in adults and are not recommended for use in children.

Duration of basic life support in the field

Continue BLS for a period of five cycles, and then reassess the patient. If there is no return of spontaneous breathing or circulation and the rescuer is alone, he/she should seek help by activating EMS. If the patient is small enough, the rescuer should take the patient with him/her and attempt to continue BLS whilst seeking help. If the patient is too large to be moved, the rescuer should position the patient on his/her side and leave to seek help. Once EMS has been activated, BLS should continue until EMS arrives or signs of life return.

Precautions and complications

Severe iatrogenic injuries are extremely uncommon in children who have undergone CPR, with an incidence of about 3%. The risk of infection to the rescuer from CPR is minimal, with the greatest risk being meningococcus from infected patients' airway secretions. Standard antibiotic prophylaxis is recommended for any rescuer involved in the resuscitation of the airway of a patient with known or suspected meningococcal infection. There have been no reported cases of transmission of hepatitis B or HIV from mouth-to-mouth ventilation to date. Airway secretions, tears, sweat and vomitus are low-risk fluids, but extra precautions should be taken when contact with blood or other bodily fluids is likely.

Relief of foreign body airway obstruction

Presentation

Foreign body airway obstruction (FBAO) in both adults and children is commonly caused by the aspiration of food, but in children it may also occur

during play with small objects. In both of these situations, it is likely that a parent or guardian will be present, and the event may have been witnessed. Clinical signs will include the sudden onset of respiratory distress, associated with coughing, gagging, inspiratory stridor, cyanosis and extreme anxiety. Many of these signs may also be caused by upper airway infections; however, the onset is slower and usually associated with other signs of infection, such as fever, lethargy, or coryza.

Foreign body airway obstruction management: conscious patient with effective cough

In a child or infant who is conscious with good respiratory effort and effective coughing, do not interfere with the child's spontaneous efforts. Any attempt to relieve the obstruction in this situation may dislodge the foreign body and worsen the situation by turning partial to complete airway obstruction. Encourage and support the child, call for help and get the child or infant urgently to an emergency facility.

Attempts to relieve FBAO should only be commenced when the cough becomes ineffective with increasing respiratory distress or the child becomes unconscious or apnoeic.

Foreign body airway obstruction: conscious patient with ineffective cough

A combination of back blows and chest thrusts is utilised to relieve the obstruction. Abdominal thrusts are not recommended for any age group because of the risk of trauma to abdominal structures.

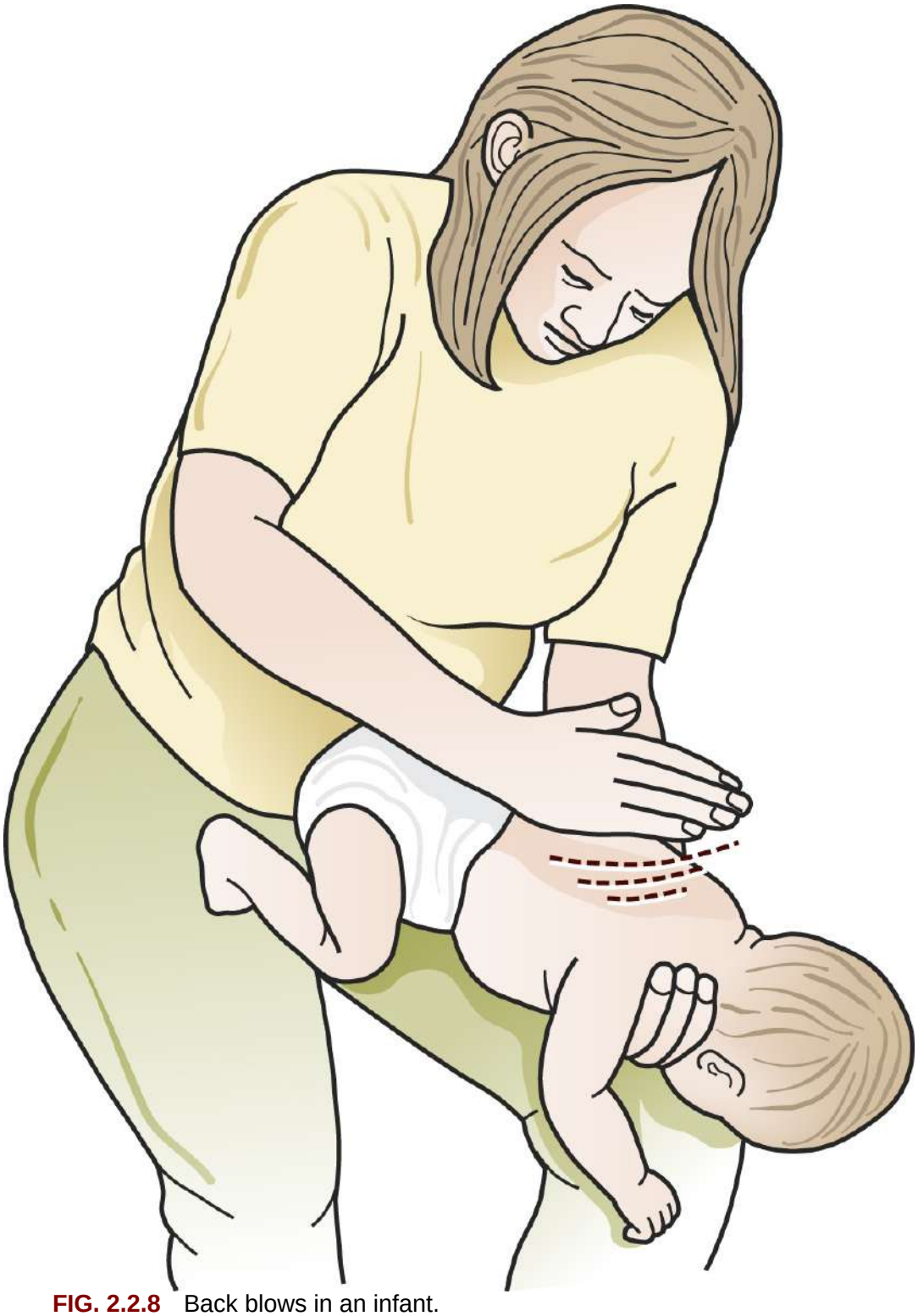

FIG. 2.2.8 Back blows in an infant.

For the child or infant who is conscious but has an ineffective cough:

- Deliver up to five back blows using the heel of the one hand in the middle of the back between the shoulder blades. Use sufficient force to dislodge a foreign body. Check after each back blow to see if the FB has been dislodged. Infants can be placed in a prone, head-down position lying along the rescuer's forearm with the jaw supported by the rescuer's hand to deliver back blows (Fig. 2.2.8).
- If the obstruction is not relieved perform up to five chest thrusts utilising the same position and technique as for CPR but at a rate of one compression per second with sufficient force to expel the foreign object. Check after each chest thrust to see if the FB has been dislodged. Infants should be positioned supine lying along the rescuer's thigh, in a head-down position (Fig. 2.2.9).

Continue the above sequence while the patient remains conscious until the obstruction is relieved, whilst seeking help.

Foreign body airway obstruction management: unconscious patient

The sequence of responses for an unconscious, apnoeic patient with FBAO is as follows:

- Open the airway using chin lift or jaw thrust. Look inside the mouth and remove any visible foreign body under direct visions using suction or Magills forceps. Do not perform a blind finger sweep because of the risk of damaging palatal tissues or pushing the FB further into the airway.

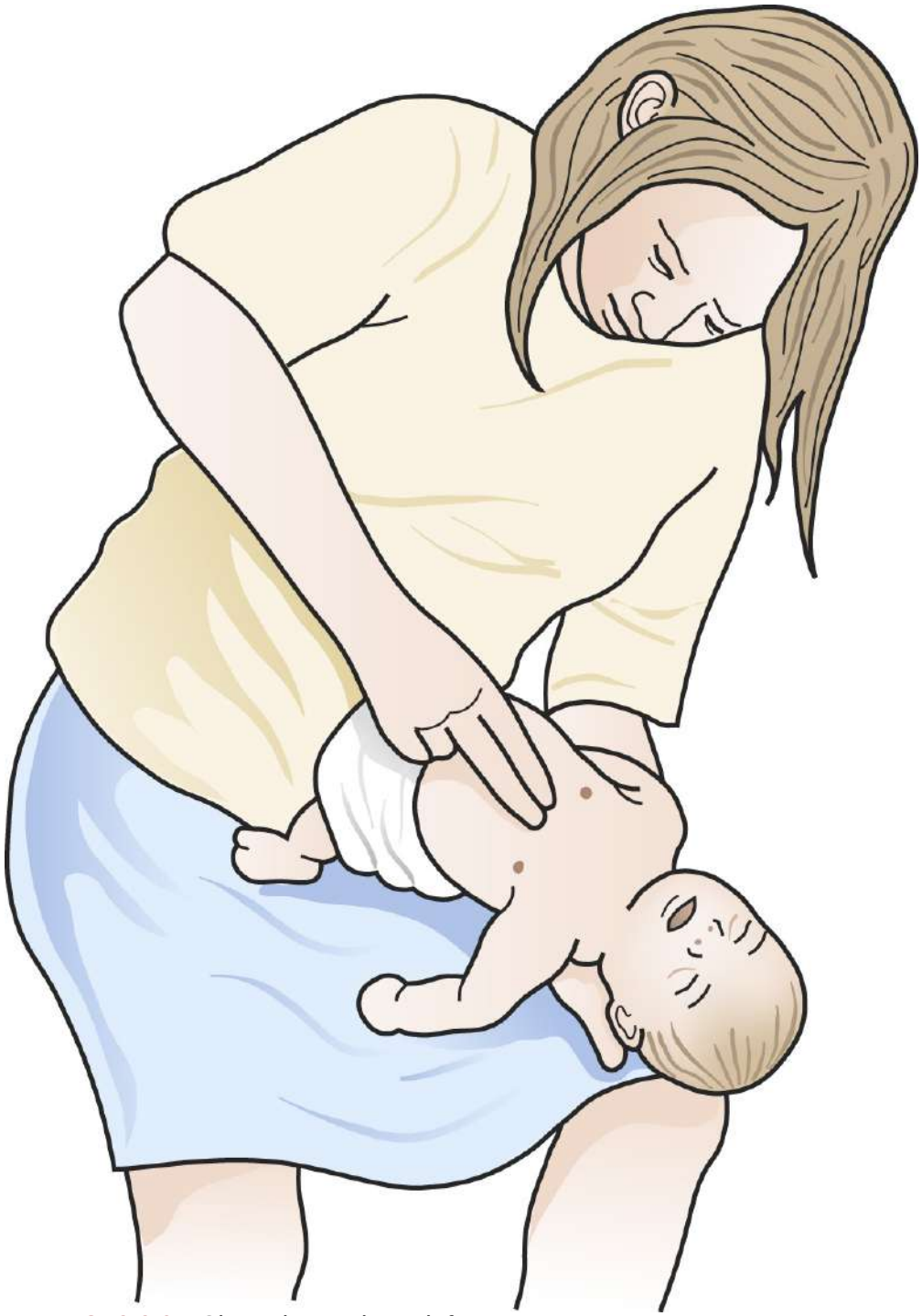

FIG. 2.2.9 Chest thrusts in an infant.

- Commence CPR and send for help.

For management in the emergency department setting see inhaled FB (Chapter 6.2).

Further reading

Australian Resuscitation Council Guidelines, ANZCOR Guidelines 4–6, 8, 9.1.6, 12.2, 13.6. http://www.resus.org.au/

European Resuscitation Council Guidelines for Resuscitation 2015. Section 6: paediatric life support. *Resuscitation*. 2015;95:223–248.

Maconochie I.K, de Caen A.R, Aickin R, et al. Part 6: pediatric basic life support and pediatric advanced life support: 2015 International Consensus on Cardiopulmonary Resuscitation and Emergency Cardiovascular Care Science With Treatment Recommendations. *Resuscitation*. 2015;95:e147–e168.

2.3

Paediatric advanced life support

Ben McKenzie, and James Tibballs

ESSENTIALS

1 Diagnosis of cardiac arrest by pulse palpation alone is unreliable among healthcare personnel. If the patient appears to have no circulation, commence advanced life support (ALS).

2 The major causes of cardiopulmonary arrest in infants and children include any cause of hypoxaemia or hypotension or both.

3 Aspects of cardiopulmonary resuscitation (CPR) are different for ‘the newly born’, infant, child and large (older) child.

4 Avoid hypoxaemia during attempts at intubation which should be limited to 30 seconds.

5 The approximate *uncuffed* endotracheal tube (ETT) size may be chosen for children over 1 year by the formula: size (mm) = age (years)/4 + 4. (*Cuffed* ETT: size (mm) = age (years)/4 + 3.5).

6 Confirm correct location of ETT in the trachea immediately after intubation.

7 ETT, face mask and laryngeal mask are all valid methods of administering artificial ventilation.

8 Have a plan to cope with a ‘can’t intubate, can’t ventilate’ situation.

9 Obtain intraosseous (IO) access if you cannot cannulate a vein rapidly.

10 Administer lipid-soluble drugs (adrenaline [epinephrine], atropine, lignocaine [lidocaine] and naloxone) via ETT if intravenous and IO access impossible.

11 Restore intravascular volume with a crystalloid solution (0.9% normal saline or Hartmann's solution) or a colloid in aliquots of 20 mL kg.

12 Base decisions to cease CPR on a number of factors including the duration of resuscitation, response to treatment, pre-arrest status of the patient, remediable factors, likely outcome if ultimately successful, opinions of personnel familiar with the patient and, whenever appropriate, the wishes of the parents.

Introduction

Definition of advanced life support

Advanced life support (ALS) is cardiopulmonary resuscitation (CPR) with trained personnel using specific skills and the equipment available in a hospital or ambulance setting. It includes the management of critically ill infants and children in pre-cardiorespiratory arrest (CPA), during arrest and post arrest.

The recommendations for advanced CPR given here are based on publications of the Australian Resuscitation Council,[1] the European Resuscitation Council,[2] the American Heart Association[3] and the International Liaison Committee on Resuscitation (ILCOR).[4] They are intended for use by medical and nursing personnel in hospital and by ambulance personnel in the field.

To add ability to knowledge, it is advisable to undertake a specialised paediatric cardiopulmonary resuscitation course such as the Advanced Paediatric Life Support (APLS) or Paediatric Advanced Life Support (PALS) courses.

Diagnosing cardiac arrest

Healthcare personnel (doctors and nurses) have difficulty diagnosing cardiac arrest in infants and children if they rely on pulse palpation alone. Their accuracy is approximately 80% with a sensitivity of 0.85 and specificity of 0.65,[5] which means that in 15% of circumstances they would not give CPR when needed and would give it in 35% when not needed. While application of CPR is not harmful when there is a circulation, the withholding of CPR when there is none dooms the patient to die. The time taken to diagnose cardiac arrest is longer than hitherto realised[6] – as a group, healthcare personnel take an average of 15

seconds to exclude cardiac arrest by finding a pulse but 30 seconds to diagnose real cardiac arrest by the absence of a pulse. However, the accuracy and expediency of diagnosis are related to experience and training. Only experienced personnel who palpate pulses on a daily basis are able to detect a real pulse within 10 seconds, but they, like inexperienced personnel, are unable to quickly diagnose cardiac arrest by the lack of a pulse and need on average about 25 seconds to confirm it. Clinical guidelines advise spending no more than 10 seconds on pulse palpation and to combine whatever information is gained with observable signs of circulation such as responsiveness, movement and presence or absence of normal respiration. In short, if the patient is unresponsive and not breathing normally there is no point wasting time on pulse palpation (it is inaccurate and time consuming). Instead give CPR immediately.

Epidemiology

The causes of cardiopulmonary arrest in infants and children are many and include any cause of hypoxaemia or hypotension or both. Common causes are trauma (motor vehicle accidents, near drowning, falls, burns, gunshots), drug overdose and poisoning, respiratory illness (asthma, upper airway obstruction, parenchymal diseases), post-operative (especially cardiac), septicaemia and sudden unexplained death in infancy.

Oxygen, ventilation and advanced airway support

Oxygen

Oxygen should be administered whenever hypoxaemia occurs, but evidence from animal and newborn infant studies suggests that as soon as oxygenation is achieved the inspired oxygen (FiO_2) should be regulated to yield arterial oxygen partial pressure in the normal range in order to limit oxygen-mediated cell damage.

The only exception to administration of oxygen is when it may cause pulmonary vasodilatation and thereby shunt blood to the lungs away from the systemic circulation, as may occur in an infant with a single ventricle which pumps blood to both circulations.

Numerous devices may be used to supply supplemental oxygen. The choice is

dictated by the required inspired oxygen concentration (FiO_2), cost, avoidance of CO_2 rebreathing, imposed airway resistance and tolerance by the patient.

Nasal prongs/nasal catheters

These are easy to use, cheap and are well tolerated, especially in the preschool age group. They do not cause gastric distension and do not need humidification. They deliver an unreliable oxygen concentration depending on whether a child is breathing by nose or by mouth (crying, nasal obstruction) and the size of the nasopharynx which acts as a reservoir. They are limited to 4 L minute and the unhumidified oxygen may cause dessication of mucosal membranes, particularly in younger children. They may become blocked with nasal secretions.

An oxygen catheter placed in the nasopharynx at a distance equivalent to that from ala nasi to tragus provides a small amount of positive end-expiratory pressure (PEEP) and indeed may be used for that purpose. Oxygen concentrations of 30%, 40% and 50% approximately are provided by flows of 45, 80 and 150 mL kg^{-1} min^{-1}, respectively. This technique to provide PEEP may be useful to temporarily abort central apnoea in the young infant with respiratory syncytial virus (RSV), whilst other treatment is established. Single oxygen catheters do not cause rebreathing and are well tolerated (permitting eating and drinking). Excessive flow may desiccate mucosal membranes and cause gastric distension, which can embarrass respiration.

High flow nasal cannula

The use of ‘high flow’ humidified oxygen as a means to provide continuous positive air pressure (CPAP) and thereby provide ventilatory support is being increasingly used for bronchiolitis and traditional indications for CPAP (RSV apnoea, apnoea of prematurity, congestive heart failure, children with neuromuscular disease, etc.). They can be quick to apply and may be useful in infants and children in extremis while they are being evaluated and further support prepared. Flow rates above 8 L min potentially generate PEEP of 5 cm water. A special circuit must be used, and the high flow rates mandate humidification. Flow rates of 2 L min kg have been studied in infants with bronchiolitis and appear safe.[17] For older children an example protocol is 2 L min kg for the first 10 kg body weight plus 0.5 L min kg for each kg body weight after to a max of 50 L min.

Inhaled bronchodilators cannot be given on top of high flow nasal cannula as the majority of the patient's inspiratory gas comes from the nasal cannula displacing any aerosolised medications.

Oxygen masks

Semi-rigid face masks of the Hudson type can supply approximately 35–70% O_2 at flow rates of 4–15 L min^{-1}. However, they may not be well tolerated by the infant or small child, and the distress they cause may consume energy in the tiring child. They may cause rebreathing or fail to deliver the desired FiO_2, especially when peak inspiratory flow rate (PIFR) is high, thus entraining excessive room air. Masks that incorporate a reservoir bag or a Venturi may deliver up to 80% O_2 but, likewise, may cause rebreathing if the flow rate does not match PIFR during respiratory distress.

Head boxes and incubators

A clear Perspex head box can administer a high concentration of oxygen to the unintubated infant. It allows a non-distressing delivery of oxygen and allows clear observation of the child. Precise oxygen therapy is possible but may be expensive, and rebreathing, heat loss and desiccation are potential problems. To avoid rebreathing, a large flow rate (10–12 L min^{-1}) of fresh gas with predetermined oxygen content should be introduced. The practice of introducing a low flow rate of 100% oxygen into a head box (to gain a lesser concentration) may cause hypercarbia. If 100% oxygen is the only compressed gas available, lesser concentrations of oxygen may be attained without rebreathing by using a flow of 100% oxygen and a Venturi device. The relatively large capacity of an incubator precludes the attainment of a high concentration of oxygen, but limited oxygen therapy, up to a maximum of 60%, can be achieved at the cost of very high flow rates. The concentration of oxygen in the head box can be monitored with an analyser and help monitor the progress of the lung disease and oxygen requirements.

Ventilation

Ventilation is emphasised in paediatric ALS and should be applied as soon as practicable. External ventilation can be applied to the child using three standard

pieces of equipment: a face mask, endotracheal tube (ETT) or laryngeal mask airway (LMA). Methods of ventilation to interface with these three pieces of equipment are the self-inflating bag, T-piece devices and mechanical ventilators.

Mask ventilation (bag-valve-mask)

This is usually the first technique employed to artificially ventilate a child using a self-inflating bag or T-piece device. Initial effective bag–mask ventilation is a necessary prerequisite for successful paediatric CPR, but it is a relatively difficult technique to learn and to perform well in emergency circumstances. Practice on mannequins or in an anaesthetic setting is invaluable in learning to perform this well for the infrequent paediatric resuscitation.

Face masks are of two types – either circular or conformed to the child's face. They may be air filled or made of soft plastic and are clear to be able to observe the face for cyanosis and detect vomiting.

Insertion of an oropharyngeal (Guedel) airway along with chin lift or jaw thrust may be necessary to facilitate bag–mask ventilation.

Self-inflating bags

These bags are portable, light weight, do not require a gas source to operate, and come in three sizes: 250, 500 and 1500 mL. The Laerdal series typifies these devices and is available in single-use disposable versions. The 250 mL bag is only suitable for use in small infants. Rebreathing is prevented by a one-way duck-bill valve, spring disk/ball valve or diaphragm/leaf valve. Pressure is simply and easily generated by squeezing the bag but can be difficult to regulate, potentially causing stomach distension or pneumothorax. A pressure-relief valve is often present and opens at 35 cm H_2O (3.5 kPa) which may be required to be overridden for high-resistance/low-compliance lungs. Supplemental oxygen is connected to the resuscitation bag and a reservoir bag may or may not be present. There is no positive end-expiratory pressure provided by self-inflating bags, but most have a provision to attach a separate PEEP valve for this purpose. While it is possible to spontaneously breathe through the bag, the resistance is significant and must be considered if this device is being used for pre-oxygenation or prior to rapid sequence intubation while the child is still breathing spontaneously.

These bags should not be used to provide supplemental oxygen to a

spontaneously breathing patient with a mask placed near or loosely over the face. With Laerdal and Partner bags, negligible amounts of oxygen (0.1–0.3 L min^{-1}) issue from the patient valve when 5–15 L min^{-1} of oxygen is introduced into bags unconnected to patients.[7] The patient valve is unlikely to open unless the mask is sealed well on the face. Although not recommended, if they are used in this way, it is vital to ensure that the patient valve opens or the reservoir bags deflates in unison with the chest movement.

The delivered oxygen concentration is dependent on the flow rate of oxygen, use of the reservoir bag, and the state of the pressure relief valve (whether open or closed). In the Laerdal series, with use of the reservoir bag and oxygen flow greater than the minute ventilation, 100% oxygen is delivered. Without the reservoir bag the delivered gas is only 50% oxygen, despite oxygen flow rate at twice minute ventilation. At an oxygen flow rate of 10 L min^{-1} to the infant resuscitator bag, the delivered gas is 85–100% oxygen without the use of the reservoir bag.

T-piece devices

These are small machine devices that generate a positive inspiratory and expiratory pressure that can be adjusted and set according to an inbuilt pressure gauge. They are typified by Neopuff and NeoPIP devices. They are dependent on a gas source and use a specific circuit that attaches to the mask or ETT. The thumb is used to press a valve and deliver the inspiratory pressure for as long it is held and then released to allow expiration. They are commonly used in neonatal resuscitation and are appropriate for use in small infants.

They have the benefit of being able to deliver a consistent pressure during inspiration but can still deliver large volumes of gas potentially causing gas distension of the stomach or worsening of any pneumothorax.

Flow-inflating bags

These are designed to give either positive pressure ventilation or to allow a patient to breathe spontaneously. They are exemplified by Jackson-Rees modified Ayre's T-piece and are used mostly in the anaesthetic environment. They are difficult to use in resuscitation settings and require specific training.

Rates and ratios of external cardiac

compression and ventilation in advanced life support

The techniques of external cardiac compression and expired air resuscitation (mouth-to-mouth) or rescue breathing are described in Chapter 2.2. The recommended ratio of external cardiac compression to ventilation in basic life support by a single rescuer is 30:2[1–4] which if able to be repeated five times in 2 minutes, with pauses for ventilations, would yield approximately 75 compressions and five breaths per minute.

In ALS the recommended ratio of compressions to ventilation by two healthcare rescuers is 15:2[1–4] which, if repeated five times in 1 minute, with pauses for ventilation by bag–mask, would yield approximately 75 compressions and 10 ventilations per minute. When the patient is intubated, it is undesirable and not necessary to pause cardiac compressions to give ventilations because they can be delivered effectively during cardiac compressions, preferably each breath being timed just after a compression. If an LMA is being used there are no data to indicate if ventilation is effective using this device during continuous CPR, and it is recommended to pause chest compressions to deliver breaths as per mouth-to-mouth and bag-valve-mask techniques.[1]

Interruptions to chest compressions should be minimised. Every time cardiac compression is interrupted, cardiac stroke volume obviously falls to zero and then several successive compressions are required to re-establish the stroke volume achieved before interruption. Necessary interruptions to cardiac compressions should be minimised by coordinated planning–for example, to analyse the cardiac rhythm or to give DC shock. Effort must be directed towards minimising ‘hands-off time’ during requirement for cardiac compression.

Advanced airway support

Tracheal intubation

The trachea should be intubated as soon as practicable, but it can be deferred if successful bag–mask ventilation can be given. It can be achieved during CPR or planned for a pause when the pulse is being checked. It should not be undertaken by inexperienced personnel out-of-hospital[8] because of complications and poorer outcomes compared with the use of bag–mask ventilation. Nonetheless, intubation has numerous advantages, which include establishment and

maintenance of the airway, facilitation of mechanical ventilation, titration of oxygen therapy, minimisation of the risk of pulmonary aspiration, enablement of tracheal suction, provision of a route for the administration of selected drugs and preferred for transport and long-term ventilation. Regurgitation of gastric contents is common during cardiac arrest.

Hypoxaemia should be avoided during attempts at intubation – which should be limited to 30 seconds. If difficulty is experienced, oxygenation should be re-established with bag–mask ventilation before a reattempt at intubation. Initial intubation should be via the oral route, not via the nasal route. The oral route is invariably quicker and is less likely to cause trauma and haemorrhage, and the ETT is more easily exchanged if the first choice is inappropriate.

A nasogastric tube should be inserted after intubation to relieve possible gaseous distension of the stomach sustained during bag–mask ventilation.

Correct placement of the ETT in the trachea must be confirmed immediately. In the hurried conditions of emergency intubation at cardiopulmonary arrest, it is not difficult to mistakenly intubate the oesophagus or to intubate a bronchus. There is no substitute for visualising the passage of the tip of the ETT through the vocal cords, confirmation of bilateral pulmonary air entry by auscultation in the axillae, continuous observation of rise and fall of the chest on ventilation, and maintenance of oxygen saturations. In addition, it is recommended that correct placement of the ETT be confirmed by capnography or CO_2 detection, with the realisation that CO_2 excretion can only occur with effective pulmonary blood flow. This implies that CO_2 detection cannot be expected unless spontaneous cardiac output returns or external cardiac compression is effective. Absent CO_2 detection mandates re-intubation or at least inspection that the tube is indeed passing through the vocal cords. High CO_2 indicates poor ventilation. Oxygenation should be confirmed with use of a pulse oximeter or measurement of arterial gas tension.

Table 2.3.1

Endotracheal tube sizes (internal diameter) and orotracheal and nasotracheal depths for infants and children

Age/body weight (kg)	Size (mm)	Oral depth (cm)	Nasal depth (cm)
Newborn (3.5)	3.0	9.0	11.01–1.5
1–6 months	3.5	9.5–11	12–13
6–12 months	4.0	11.5–12	13–14
2–3 years	4.5	13–13.5	15–16
4–5 years	5.0	14–14.5	17–18
6–7 years	5.5	15–15.5	19
8–9 years	6.0	16–16.5	20
10–11 years	6.5	17–17.5	21
12–13 years	7.0	18–18.5	22
14–16 years	7.5	19	23

Endotracheal tube size (Table 2.3.1)

Uncuffed sizes are 2.5 mm for a premature newborn <1 kg, 3.0 mm for infants 1–3.5 kg, 3.5 mm for infants >3.5 kg and up to age of 6 months, size 4 mm for infants 7 months to 1 year (see Table 2.3.1). The approximate size may be chosen for children over 1 year by the formula: size (mm) = age (years)/4 + 4. Tubes one size larger and smaller should be readily available. The correct size should allow a small leak on application of moderate pressure but also enable adequate pulmonary inflation. If the lungs are non-compliant, however, it may be necessary to insert a tube without a leak or insert a cuffed tube. Appropriate-sized **cuffed** tubes are a half size smaller than uncuffed tubes and may be estimated by the formula: size (mm) = age (years)/4 + 3.5.

Depth of tube insertion (see Table 2.3.1)

The tube is inserted to a specific numerical depth to avoid accidental extubation or endobronchial intubation. Most manufacturers of ETTs have a black indicator stripe on the tip of the tube to show the appropriate depth to insert beyond the vocal cords. Assessment of the depth of insertion during laryngoscopy by noting passage through the vocal cords is not completely reliable since alteration of head position affects depth of tube insertion. The tube depth increases with neck flexion (goes in) and decreases (comes out) during extension. Since intubation is usually performed with the head extended, the tube depth increases when the laryngoscope is removed and the head assumes a position of neutrality or flexion.

In a neutral head position, an appropriate depth of insertion measured from the centre of the lips for an **oral tube** is 9.5 cm for a term newborn, 11.5 cm for a 6-month-old infant and 12 cm for a 1-year-old. After 1 year the depth is given by the formula: depth (cm) = age (years)/2 + 12. An alternative formula for oral

tube depth of insertion is: depth (cm) = size (mm) × 3 when an appropriate tube size for age is used. The appropriate depth of insertion for a **nasal tube** in this age group is: depth (cm) = age (years)/2 + 15. On a chest X-ray taken with the head in neutral position, the tip of the tube should be at the interclavicular line.

Laryngeal mask airway

These have been used for resuscitation by medical, nursing and ambulance personnel trained in their selection and insertion. They may be used to maintain an airway and are a suitable alternative to the use of airway opening manoeuvres and use of oropharyngeal and nasopharyngeal airways. They are useful to establish an airway in the setting of airway obstruction or failed intubation.[9] An intubating LMA serves as a conduit for intubation.

Table 2.3.2

Laryngeal mask airways

Sizes are available to suit body weight (kg) of newborns, infants and children	
Size	**Weight (kg)**
1	<5
1 1/2	5–10
2	10–20
2 1/2	20–30
3	30–50
4	50–70
5	70–100
6	>100

However, the role of LMA in provision of mechanical ventilation remains uncertain and should be considered a rescue option. Like bag–mask ventilation, they do not protect the airway from aspiration, which occurs commonly during cardiopulmonary resuscitation. They are a suitable alternative to a face mask as a means to give ventilation before endotracheal intubation and when intubation is difficult. This is a better technique when the operator is unskilled in the use of LMA and intubation. Although insertion of an LMA is easier to learn than endotracheal intubation, training should not replace mastery of bag–mask ventilation. They should not be used in semi-conscious patients or when the gag reflex is present and are not suitable for long-term use or use during transport when endotracheal intubation is far preferable. They are subject to dislodgment during movement and transport. Appropriate sizes according to body weight are given in Table 2.3.2.

Management of the difficult airway

Most difficult paediatric airways are predicted in advance. Emergency medicine clinicians should have a method for assessing the difficulty of a patient's airway before embarking on sedation or rapid sequence induction (RSI) of anaesthesia.

The unanticipated difficulty is less common in paediatrics, but clinicians should articulate a plan for dealing with airway difficulties before embarking on the procedure. Pre-intubation checklists can be a useful aid in ensuring there is preparation for a difficult airway, even if the airway appears routine. Unfortunately morbidity and mortality still occur from complications arising from unanticipated difficult airways.

When the airway is predicted to be difficult and there are extensive preparations with skilled operators, cardiorespiratory arrest still occurs in 2% of patients, and hypoxaemia is very common.[18]

Causes of a difficult airway

The airway, or maintenance of airway, can be difficult because of unusual anatomy, injury or illnesses or because of loss of natural maintenance after administration of drugs which depress consciousness, muscle tone or activity:

Trauma – Direct laryngeal or tracheal injury may cause swelling and distortion of the airway. Facial injury may make mask ventilation impossible, and blood may obscure the view of the airway during intubation. Extensive subcutaneous emphysema in the neck may similarly cause distortion and difficulty. Cervical spine immobilisation and maintaining inline stabilisation limit head extension and ability to bring the glottic visual axis into line with the oropharyngeal axis. Every effort should be made to maintain neck stability, but successful ventilation/oxygenation takes precedent over spine immobilisation, and the forces potentially applied to the neck during intubation are small compared to the original injury.

Infection – Swelling causing distortion and impaired view of the airway will arise from epiglottitis and neck abscesses.

Angioedema – Tongue swelling or peri-glottic swelling may arise from angioedema from conditions such as IgE-mediated anaphylaxis or C1 esterase deficiency.

Syndromic children – Common anatomical variants which will disallow a

direct line of sight to the larynx are an ‘anterior’ larynx, prominent upper incisors, a large tongue or a small hypoplastic mandible. Treacher Collins syndrome and Pierre–Robin syndrome (sequence) with mandibular hypoplasia and Beckwith–Wiedemann syndrome with macroglossia are typical conditions in which difficult intubation should be anticipated.

Predicting a difficult airway

Anything that obscures the line of sight to the glottis or prevents the oropharyngeal visual plane being brought into line with the visual plane of the glottis will create a difficult airway.

Predictors include the relative tongue/pharyngeal size (‘Mallampati’ test): if the faucial pillars, soft palate and uvula are obscured by the extended tongue on maximum mouth opening, intubation will be difficult. Another predictor is the extent of possible atlanto-occipital extension. If this is less than 35 degrees intubation will be difficult. This corresponds to the angle between the occlusal surface of the upper teeth and horizontal plane when the head is maximally extended. The third predictor is the amount of mandibular space into which the tongue must be compressed to allow a line of sight to the glottis. This space can be judged by the distance from the thyroid cartilage or hyoid bone to the point of the mandible (thyromental, submental distances) or by the horizontal length of the mandible. In adult-sized patients a thyromental distance of greater than 6 cm and a mandibular length of greater than 9 cm predict easy intubation. Unfortunately, no such distances are known for children and infants.

Thus, the evaluation of intubation difficulty should include pharyngeal examination with extended tongue, inspection of mandibular size or sub-mental space and examination of atlanto-occipital angle. These special measures should be in addition to routine history of airway management, as would have occurred in previous anaesthesia, inspection of nostrils and history and examination of cardiorespiratory function.

Difficult intubation

With good technique and preparation, intubation can normally be achieved via direct laryngoscopy with a direct line of sight to the glottis. A plan to ventilate and oxygenate the patient with bag-valve-mask, ETT or LMA should exist, and

there should be willingness to employ/switch between these approaches to achieve ventilation should there be difficulty managing the airway with a particular technique.

Resuscitators must be well equipped and trained to optomise each approach. If all three approaches fail to achieve ventilation (‘can’t intubate, can’t ventilate’) then a plan to commit to a surgical airway intervention should exist and be executed in a timely fashion before hypoxaemic brain damage and death occur.

When the airway is predicted to be difficult, success of first pass ETT ranges from 2% to 50% in one registry and the likelihood of failure dramatically increased after two attempts at intubation regardless of technique.[18]

The gold standard for management of a likely difficult airway is a skilled operator, with a fibre-optic assisted intubation, with a surgeon experienced in surgical airways present to be able to perform an emergency surgical airway.

Endotracheal intubation technique optimisation

Please refer to Chapter 24.3 for details on endotracheal intubation.

Video laryngoscopes are increasingly available and usually improve the intubation grade (Cormack and Lehane) by at least 1 grade. Some video laryngoscopes use traditional Mackintosh or Miller blades and are easy for clinicians to adapt to. Others have different designs and require practice to manoeuver and guide the ETT while looking at the video screen.

The bevel of an ETT is angled so that it easily enters the trachea when introduced from the right side of the mouth. When introduced from the left side, the tip of the bevel may stick at the laryngeal inlet – but this can be easily remedied by rotating the tube slightly. If the tip of the upward curving tube sticks in the anterior larynx, flexion of the neck may direct the tube posteriorly and encourage it to enter the larynx.

If the larynx is anterior and difficult or impossible to see, use of an introducing stylet to create a more acute curve at the tube tip may achieve intubation, and/or application of posterior cricoid pressure may bring the anterior larynx into view. A tube may be placed ‘blindly’ into the trachea with or without the use of a stylet or bougie over which a tube can be railroaded. Skilled anticipatory assistance and good suction by a dedicated airway nurse who anticipates the needs of the intubator are essential. Occasionally, a gentle finger of an assistant retracting the right side of the mouth may enlarge the oral opening to afford improved vision and field to place the ETT.

A two-person intubating technique may achieve intubation in difficult circumstances. In this, one person holds the laryngoscope with his/her left hand and applies cricoid pressure with his/her right to bring the larynx into view, while a second person applies suction, holds the lower lip out of the way and attempts to pass the ETT.

Many other items of equipment are available to assist when intubation proves to be difficult. These include laryngoscopes that have a tip independent of the shaft of the blade, which is used to elevate the epiglottis (e.g. McCoy laryngoscope, Penlon) or which incorporate a prism to give a view of an anterior larynx (e.g. Belscope), illuminating stylets and intubating bronchoscopes. Illuminating stylets are malleable and give a ‘Jack-o'-lantern’ effect when correctly located in the trachea. Ultrasound has also been employed to confirm ETT placement. All items of equipment are useless unless they are ready-to-hand and are familiar to the operator.

Failed intubation

Attempts at intubation should not be prolonged and hypoxaemia avoided. It should be the responsibility of all of the resuscitation team to vocalise oxygen desaturations if they are caused by prolonged ETT attempts. Oxygenation should be restored with intermittent bag–mask ventilation. Similarly, intubation attempts should not be repetitive because the larynx will be traumatised, and this may render bag–mask or LMA ventilation, previously possible, now impossible for the more-skilled operator when they arrive.

If endotracheal intubation is impossible when needed, the resuscitator must be able to oxygenate the patient while preparations are made for tracheostomy or until intubation can be somehow achieved. Help should be called for already.

If the airway is totally obstructed and none of intubation, bag-mask-valve, or LMA is successful, the situation is one of ‘can’t intubate, can’t ventilate’, and a surgical approach must be achieved before hypoxaemic brain damage occurs.

Emergency surgical airway techniques

Whether needle cricothyroidotomy with jet insufflation or semipermanent open surgical cricothyroidotomy is used in the first instance, the decision must be made and implemented rapidly.

The cricothyroid membrane lies inferior to the thyroid cartilage and is the

point of access for emergency surgical airways. It is more difficult to identify and access in small children. An open surgical approach with a scalpel becomes more difficult under the age of 8, but there is no absolute cutoff if the procedure is achievable and life-saving. Smaller children have a higher complication rate of laryngeal injury and subglottic stenosis with surgical airways. In very small children, a needle approach is usually recommended while an airway competent surgeon, if available, performs an emergency tracheostomy.

Needle cricothyroidotomy

Adequate oxygenation (but not normal ventilation) can be obtained by jet insufflation oxygen via transcutaneous cannula into the trachea. This is a temporising measure that may oxygenate a child for 15 minutes or longer in a small child. There are proprietary kits for this method or a simple 14G IV cannula can be used. With either equipment, the cannula is inserted percutaneously into the trachea caudad via the cricothyroid membrane (which lies immediately inferior to the thyroid cartilage). To do this, the patient should be lying straight, with the cannula in the midline and angled towards the feet.

Some proprietary kits have larger seldinger technique catheters that can be connected to a standard 22 mm ETT connector and a bagging circuit. If an intravenous (IV) cannula is used, it can be connected by various ways to a source of oxygen. One of these is direct connection to a resuscitator or a bagging circuit using a connector from a 3.0 mm ETT. Alternatively, the cannula can be connected to continuous oxygen supply via a valveless three-way IV tap (to allow expiration) and a length of plastic tubing. Another option is use of plastic tubing alone that has a side hole cut or a Y-piece inserted, which is intermittently occluded to cause inspiration (1 second) and unoccluded (3 seconds) to allow expiration. With all these techniques care should be taken to allow expiration to avoid barotrauma. Expiration may need to be assisted by lateral chest compression as the arrested patient may not spontaneously expire much air.

A semipermanent solution for a totally obstructed airway is cricothyro(s)tomy. To do this the larynx is stabilised throughout the procedure with fingers of one hand while skin over the thyroid–cricoid membrane (between the thyroid and cricoid cartilages) is incised with a scalpel held in the other. Then, bluntly dissect into the trachea with forceps in the midline or incise vertically with the scalpel. Insert a small ETT, over a bougie if necessary. A formal tracheostomy should be organised while these measures are undertaken. An important part of a contingency plan to cope with unexpected difficulties is to have easily

contactable more experienced operators.

Monitoring

Vital signs

Routine monitoring of heart rate, respiratory rate and blood pressure is essential for infants and children with critical illness. It is prudent to have ready access or to have displayed the normal age-related values visually available in the paediatric resuscitation area of the emergency department (ED) as an *aide-memoire*.

Blood pressure is commonly forgotten and should not be omitted in the critically unwell child requiring resuscitation. Remember that the two common causes of arrest in children are respiratory failure and shock.

Oximetry

Transcutaneous oximetry (SpO_2) is essential monitoring in critically ill patients. It equates well to arterial haemoglobin oxygen saturation (SaO_2) but not when the SaO_2 is below 70%. It becomes less reliable when the waveform is poor and the patient is shocked.

Expired CO_2 detection

Confirmation of endotracheal intubation

End-tidal CO_2 ($P_{et}CO_2$) is recommended after every tracheal intubation and during mechanical ventilation to guard against inadvertent oesophageal intubation and inadvertent extubation, particularly when the intubated patient undergoes transport to, within or between hospitals. Small movements of head and neck, as may occur for example on transfer from one trolley to another or to a bed, may dislodge an ETT.

No CO_2 is excreted unless there is pulmonary blood flow. Using end-tidal CO_2 is therefore difficult in the child with cardiac arrest, and clinicians must use all cues (direct vision, chest auscultation, chest rise and fall) to be confident of correct ETT placement. If there is doubt about endotracheal placement during a cardiac arrest then mask ventilation may be more reliable.

To achieve optimum CO_2 excretion, cardiac output and pulmonary ventilation

must be matched. A common error in advanced CPR is giving ventilation in excess of the limited cardiac output achievable by external cardiac compression, which is likely to be no more than a third of normal cardiac output. Ventilation can be safely reduced proportionately. Moreover, excessive ventilation not only interferes with performance of external cardiac compression but also increases intrathoracic pressure inhibiting venous return and cardiac output and may cause hypocarbia which causes cerebral ischaemia by vasoconstriction. On the other hand, inadequate ventilation contributes to hypercarbia, acidosis and cerebral vasodilation. After tracheal intubation, end-tidal CO_2 should be monitored by capnography. If end-tidal CO_2 is low, excessive ventilation and inadequate external cardiac compression (rate, depth of compression) should be excluded as causes.

Electrocardiograph

The ECG should be displayed with either leads or pads. Drug therapy or immediate direct current shock is administered according to the existing rhythm. Electrolyte status, especially that of potassium and calcium, may be indicated by ECG patterns. There is increasing emphasis on ECG monitoring in neonatal resuscitation as palpation or auscultation of heart beat are less unreliable.

Vascular access

Peripheral venous cannulation

Access to the circulation via a peripheral vein should be attempted immediately on CPA. Any site is acceptable. Visible or palpable peripheral veins are to be found on the dorsum of the hand, wrist, forearm, cubital fossa, chest wall, foot and ankle. In infants, scalp veins are accessible, and the umbilical vein can be used up to about a week after birth.

The external jugular veins are an under utlised site for emergency vascular access. They have no valves, are usually distended during CPR, straining, crying, or if the patient is supine, and are easy to cannulate.

Intraosseous access

If peripheral IV access cannot be rapidly achieved, say within 60 seconds, intraosseous (IO) access should be obtained. This route has been used for

patients of all ages. It provides rapid, safe and reliable access to the circulation and serves as an adequate route for any parenteral drug and fluid administration. Syringing via a three-way tap is usually needed due to the increased resistance.

The use of purpose-made IO bone injection needles with a trocar or bone marrow drill (EZ-IO, Vidacare) is generally required. Care should be taken to not traverse both cortices with the bone marrow drill, and this requires the operator to either advance carefully or be cognisant and responsive when loss of resistance after passing the first cortex is achieved. Manual needles exist, and the handle of the needle device is held in the palm of the hand while the fingers grip the shaft about a centimetre from the tip. It is inserted perpendicular to the bone surface, and a rotary action is used to traverse the cortex. Sudden loss of resistance signifies entry to bone marrow, and the needle should stand unsupported. Correct positioning of the needle is confirmed by aspiration of bone marrow (which may be used for biochemical and haematological purposes), but that is not always possible.

Although many sites have been used for bone marrow injection, the easiest to identify is the anteromedial surface of the upper or lower tibia. The site of the latter is a few centimetres below the anterior tuberosity and the former a few centimetres above the medial malleolus. Care should be exercised to avoid complications, particularly cutaneous extravasation, compartment syndrome of the leg. Contra-indications include local trauma and infection.

Central venous cannulation

Cannulation of femoral, subclavian, or internal jugulars is an option. However, central cannulation is difficult in the setting of cardiorespiratory arrest and fraught with potential serious complications such as pneumothorax unless the operator is well practised. This technique is not recommended in the setting of an arrested child as the IO route is more timely.

Endotracheal route

Drugs are absorbed into the circulation from the airways. Lipid-soluble drugs (adrenaline [epinephrine], atropine and lignocaine [lidocaine]) may be administered via the ETT if either IV or IO access is non-existent. Although the optimal doses of these drugs by this route are not known, work in animal models suggests doses should be ten times the IV doses. The drugs should be diluted in

normal saline up to 2 mL for infants, 5 mL for small children and 10 mL for large children. It is acceptable and simplest to squirt the drugs from the syringe directly into the ETT and disperse them throughout the respiratory tree with bagging. Neither sodium bicarbonate nor calcium salts should be administered via the tracheal route because they injure the airways.

Other techniques

Surgical cutdown onto a long saphenous, saphenofemoral junction or basilic is a valuable skill sometimes required in traumatic exsanguination. Very occasionally, injection into the superior sagittal sinus of an infant[1] may be the only vascular access available.

Any pre-existing functioning IV line can be used provided it does not contain any drug or electrolyte, which may have caused the CPA.

Fluid therapy

Circulatory hypovolaemia is common in trauma, sepsis, dehydration and anaphylactic states. Restoration of intravascular volume should be with isotonic crystalloid solutions (0.9% normal saline or Hartmann's solution) or 4% albumin. There is insufficient evidence to choose between these. Aliquots of 20 mL kg^{-1} IV or IO are reasonable volumes to administer in shock states with titration against indices of vascular volume. It is reasonable to administer blood in haemorrhage if 20–40 mL kg^{-1} has not restored normal blood volume (70–80 mL kg^{-1}). The role of hypertonic solutions is not yet defined for children with hypovolaemic shock, but these solutions are in regular use for patients with severe head injury. Dextrose-containing solutions are inadvisable in acute resuscitation unless hypoglycaemia is proven since they may cause osmotic diuresis. Drugs should be flushed into the circulation with boluses of isotonic crystalloid solution.

Resuscitation drugs

Adenosine

This endogenous nucleoside is the drug of choice for treatment of supraventricular tachycardia (SVT) if circulation is adequate, otherwise DC shock is the preferred first treatment. It blocks atrioventricular (AV) node

conduction and thus re-entry circuits, which are the usual causes of SVT in infants and children. It has a very short half-life (about 10 seconds) in the blood because it is deaminated and inactivated by adenosine deaminase on the surface of red blood cells. Hence, it must be delivered as a rapid bolus. The dose is 0.1–0.3 mg kg^{-1}, and the end point is achieving evidence of AV block. The first dose should be 0.1 mg kg^{-1} to a maximum of 6 mg. If ineffective and no AV block is achieved, the second dose should be increased to 0.2 mg kg^{-1} maximum 12 mg. If AV block is achieved but SVT recurs, a second-line agent is often required.

Adrenaline

Adrenaline is the most frequently used drug in paediatric ALS. Its α-adrenergic vasoconstrictive actions are considered the most important by increasing aortic diastolic pressure and coronary perfusion pressure. Its β-adrenergic actions enhance contractility and spontaneous contraction. It has never been studied vs. placebo in children in cardiac arrest, but adult studies show improved short-term outcomes (return of spontaneous circulation). The dose is 10 mcg kg^{-1} given IV or IO. It should be administered IV/IO every 3–5 minutes in cardiac arrest after every second pulse check. In patients who require support of blood pressure it can be run as an infusion of 0.05–0.3 mcg kg^{-1} min^{-1} (0.3 mg kg^{-1} adrenaline in 50 mL at 1 mL hr^{-1} = 0.1 mcg kg^{-1} min^{-1}). Higher bolus doses (up to 200 mcg kg^{-1}) or infusions may be administered in specific scenarios such as arrest from β-blocking drug overdose. Side effects of adrenaline include risk of severe vasoconstriction, ischaemia, hypertension and onset of ectopy and tachyarrhythmias. If immediate access to the circulation is not available, it can be administered via the endotracheal route at a dosage of 100 mcg kg^{-1}, but absorption is variable.

Amiodarone

This drug is used to treat a wide variety of atrial and ventricular dysrrhythmias including atrial tachycardias, junctional tachycardia, ventricular tachycardia (VT) and DC-shock resistant ventricular fibrillation (VF). It inhibits α- and β-receptors, slows AV nodal conduction, and prolongs the QT interval and QRS duration. It thus may cause *torsade de pointes* VT. The loading dose is 5 mg kg^{-1} infused over several minutes to an hour depending on the dysrrhythmia being treated. Repeated doses to a maximum of 15 mg kg^{-1} may be given. Acute side effects are vasodilatation and hypotension, and chronic effects are thyroid

function disturbance, interstitial pneumonitis, corneal deposits and blue–grey skin discolouration.

Atropine

Atropine has no role in cardiac arrest. This parasympatholytic drug is used to treat bradycardia (<60 min^{-1}) caused by excessive vagal activity or a consequence of atrioventricular block (AV). If inadequate circulation or hypotension is present, severe bradycardia should be treated with adrenaline. Bradycardia caused by hypoxaemia should be treated initially with ventilation and oxygen. The dose is 20 mcg kg^{-1}, which may be repeated after 5 minutes. Unresponsive bradycardia should be treated with adrenaline. Atropine is commonly given prior to RSI in young children to prevent the vagal stimulation of intubation that may cause bradycardia; however, the 2015 ILCOR review identified conflicting evidence regarding benefit of this practice and made no recommendation in this domain, - it is not mandatory.

Calcium

Calcium should not be administered during acute resuscitation unless the cause of collapse is due to hypocalcaemia, hyperkalaemia, calcium channel blocker overdose or hypermagnesaemia. Although intimately involved in myocardial excitation–contraction coupling, it is not useful and possibly harmful, by causing cell death, in the regular treatment of asystole, electromechanical dissociation and ventricular fibrillation. Its use is associated with poor outcomes,[10] and it should not be used without a definite indication. If it is indicated, the dose is 0.2 mL kg^{-1} of 10% calcium chloride or 0.7 mL kg^{-1} of 10% calcium gluconate, and it may be repeated after 10 minutes according to the serum level if possible.

Glucose

Disturbance of glucose metabolism occurs during critical illness. Infants are particularly at risk, as they have limited glycogen stores and may sustain hypoglycaemia, whereas the stress response may cause hyperglycaemia. Blood levels should be monitored regularly. Hypoglycaemia (<2.5 mmol kg^{-1}) should be treated with a dextrose infusion of approximately 6–8 mg kg^{-1} min or equivalent. Bolus injection may be necessary (in situations such as seizure or coma due to hypoglycaemia) in which case a bolus dose of 2 ml kg^{-1} of 10% glucose is indicated. Ten per cent glucose is less likely to cause local tissue

injury or electrolyte shifts compared with more concentrated solutions. Hypoglycaemia and hyperglycaemia after brain injury should be avoided.

Lignocaine (Lidocaine)

Lignocaine is a sodium channel blocker that decreases automaticity and suppresses ventricular arrhythmias. It has been used for shock refractory VF and pulseless ventricular tachycardia (pVT). Weak evidence suggests that it may be as effective as amiodarone for this indication and is an area of ongoing controversy.[19] If used, the recommended dose is 1 mg kg^{-1} (0.1 mL kg^{-1} of 1%) by rapid injection followed by an infusion of 20–50 mcg kg^{-1} min^{-1}. A low infusion dose is recommended if renal or hepatic dysfunction exists. Adverse side effects include myocardial depression with hypotension and central nervous system depression with depression of conscious state and convulsions.

Magnesium

Magnesium inhibits calcium channels and causes a reduction in intracellular calcium, thereby causing muscle relaxation and increasing depolarisation thresholds. In paediatric resuscitation it is used as a bronchodilator in severe asthma and to treat *torsade de pointes* (polymorphic VT, see earlier) – a dysrhythmia associated with prolonged QT interval.

The dose of magnesium sulphate is 25–50 mg kg^{-1} (0.1–0.2 mmol kg^{-1}) by IV or IO infusion over several minutes.

Sodium bicarbonate

Sodium bicarbonate is not indicated in cardiac arrest and has not been shown to alter outcome for this indication. Although it can neutralise hydrogen ions in the blood it may worsen intracellular acidosis. The product of bicarbonate and hydrogen ions is carbonic acid, which freely dissociates to form water and carbon dioxide (CO_2) and, unless hyperventilation is given, the CO_2 may cross into cells, where it reforms carbonic acid and liberates hydrogen ions. It is, however, indicated in specific cases such as overdose from tricyclic antidepressants or other drug with sodium channel blocker properties where it is considered an antidote. It is sometimes used in severe ongoing acidosis, but it should be remembered that tissue and arterial pH do not correlate well, and alternative therapies may be more helpful. The essential treatment of metabolic acidosis is treatment of the cause.

If indicated, an appropriate dose is 1 mmol kg^{-1}. Adverse effects include hypernatraemia and hyperosmolality, hypokalaemia, hypocalcaemia and metabolic alkalosis (which limits oxygen dissociation from haemoglobin). It should not be allowed to mix in IV lines with catecholamines, which it inactivates, or with calcium salts, which it precipitates.

Vasopressin

The alternative to adrenaline as a vasopressor, vasopressin, has no better survival advantage for adult victims of in-hospital cardiac arrest[11,19] and has not been adequately investigated for use during CPR for children. Adrenaline remains the preferred vasopressor in cardiac arrest and resuscitation after the 2015 ILCOR review of this subject.

Direct current shock

Unsynchronised DC shock (defibrillation) is required as first treatment for VF and pulseless VT with 4 J kg^{-1} rounded up to the nearest number setting on the manual defibrillator.

Either monophasic or biphasic waveforms may be used. The optimum dose of external DC shock in terms of achieving first shock success with minimal damage to the myocardium is unknown. While some guidelines recommend a dose of 2–4 J kg^{-1},[3,4] the dose of 2 J kg^{-1} is considered too little by other guidelines[1,2] which advise 4 J kg^{-1}. This is supported by a recent study which showed that 2 J kg^{-1} converted only about 50% of patients to a perfusing rhythm.[12]

Many national resuscitation councils, including Australia and New Zealand, recommend all shocks to be at the same upper limit (4 J kg^{-1}) to ensure resuscitation algorithms are simple and easy to follow.

Synchronised DC shock may also be required for pulsatile VT and haemodynamically unstable SVT.

Defibrillators should have paediatric pads of cross-sectional area 12–20 cm^2 for use in children <10 kg. For others, adult-sized paddles (50–80 cm^2) are satisfactory provided the paddles do not contact each other. For use in the anterolateral positions, one pad/paddle is placed over the mid-axilla opposite the xyphoid or nipple, the other to the right of the upper sternum below the clavicle. An anteroposterior position of pads is the preferred positioning (one over cardiac apex or anterior chest, one over left scapula). Dextrocardia may be present with

congenital heart disease, and the position of the pads/paddles should be altered accordingly.

In the absence of a manual dose regulated defibrillator, semi-automated automatic external defibrillation (sAED) may be used for children but preferably should have an ‘attenuated’ adult dose. A dose of 50 J is appropriate for most infants and children. However, if a machine with such attenuated dosing is not available, the use of an adult sAED delivering 150–200 J is acceptable for infants and children[1–4] rather than leave a shockable rhythm untreated. The use of an sAED with adult doses of DC shock for children in hospital should not be considered unless a manual dose regulated defibrillator is not available or cannot be used or a body-weight-specific DC shock cannot be delivered within 3 minutes when indicated.

Operators of defibrillators should be constantly alert to the possibility of inadvertent electrocution and cardiac arrest of themselves and others by misuse.

Management of pulseless arrhythmias (Fig. 2.3.1)

The following discussion assumes that mechanical ventilation with oxygen and external cardiac compression (ECC) have been commenced and continued if an adequate pulse rate is not detectable. The treatment of pulseless arrhythmias (ventricular fibrillation, ventricular tachycardia, asystole, electromechanical dissociation and pulseless electrical activity) is summarised in Fig. 2.3.1

Specific causes of arrhythmias should be treated. The 4 Hs and 4 Ts serve as a reminder to seek and treat correctable pathology as the child is resuscitated: Hypoxia, Hypovolaemia, Hyper/Hypokalaemia/Metabolic causes, Hypo/Hyperthermia, Tension pneumothorax, Tamponade (cardiac), Thrombosis (pulmonary, coronary), Toxins.

For example, calcium channel blockade toxicity is treated with calcium IV or IO (chloride 10% 0.2 mL kg^{-1}, gluconate 10% 0.7 mL kg^{-1}) and high-dose insulin euglycaemic therapy. Hyperkalaemia is treated with calcium salt, sodium bicarbonate, hyperventilation, insulin and dextrose. All drugs should be flushed into the circulation with a small bolus of isotonic fluid. To prevent inactivation, drugs should not be mixed in the syringe or in infusion lines.

Asystole/severe pulseless bradycardia

Asystole and severe pulseless bradycardia (HR <60) that are unresponsive to oxygen and ventilation should be treated with adrenaline 10 mcg kg^{-1} IV or IO. If these routes are not available, adrenaline 100 µg kg^{-1} should be administered via ETT. Unresponsive asystole should be treated with similar doses (10 µg kg^{-1} IV, IO; 100 µg kg^{-1} ETT) every 3–5 minutes. Recurrent bradyarrhythmia or asystole may require an infusion of adrenaline at 0.05–3 mcg kg^{-1} min^{-1} infused into a secure large vein. Pacing (transcutaneous or other) if available may be effective but should not interfere with CPR.

Pulseless electrical activity and electromechanical dissociation

A relatively normal ECG complex without pulse is called pulseless electrical activity (PEA), although it is sometimes referred to as electromechanical dissociation (EMD). The blood pressure at which a pulse is palpable varies but the absence of consciousness should trigger the assumption that there is no pulse. If untreated, the ECG deteriorates to a broader abnormal QRS that is progressively more bradycardic usually until asystole ensues. PEA should be treated as for asystole and the cause ascertained and treated.

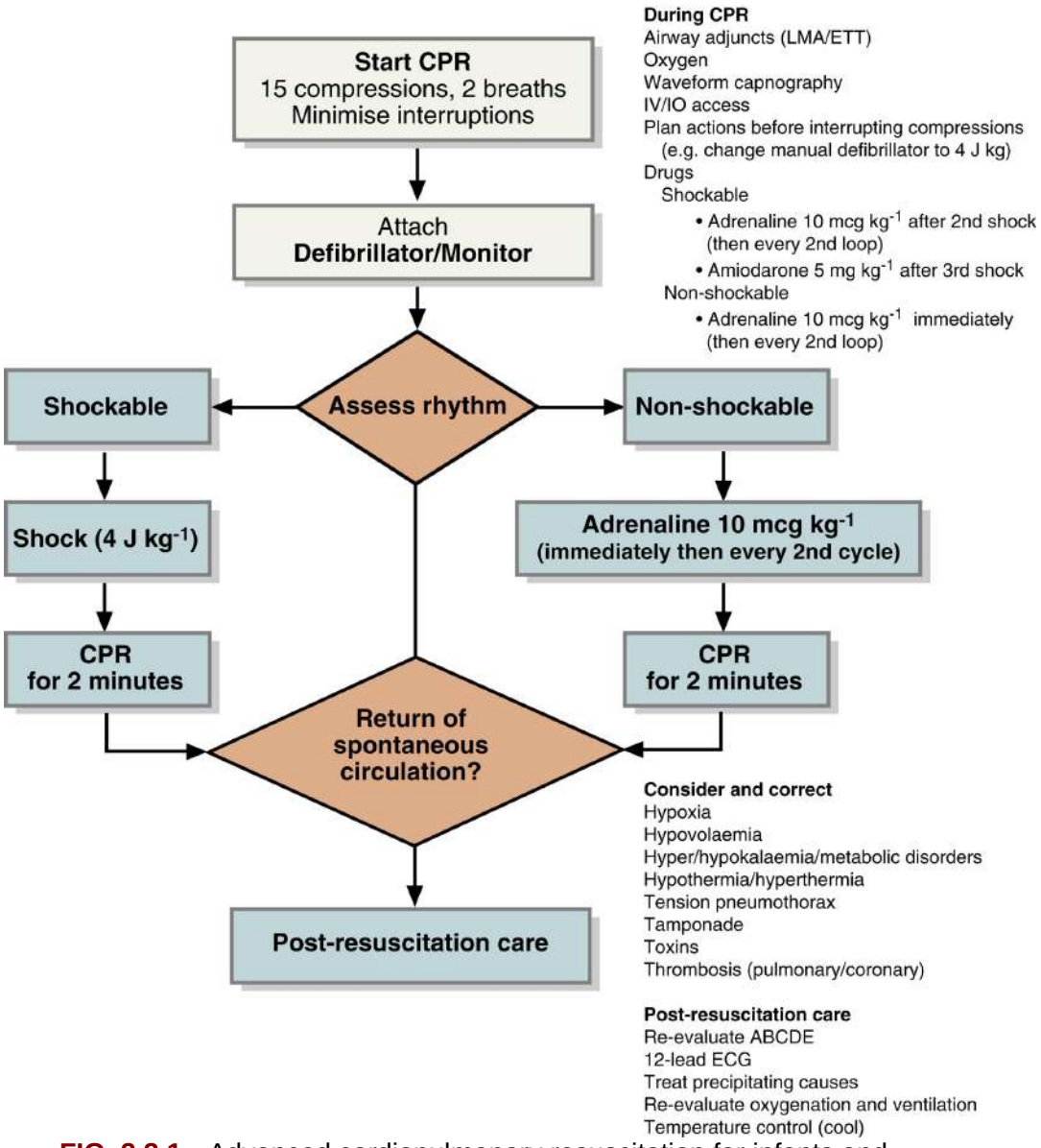

FIG. 2.3.1 Advanced cardiopulmonary resuscitation for infants and children. Adapted from resuscitation guidelines of the International Liaison Committee on Resuscitation, Australian Resuscitation Council, European Resuscitation Council and of the American Heart Association, December 2010. Australian Resuscitation Council.
Key: CPR, cardiopulmonary resuscitation; ECG, electrocardiograph; IO, intraosseous; IV, intravenous; J, joules; kg, kilogram; mg, milligram; mcg, microgram.

Ventricular fibrillation and pulseless ventricular tachycardia

In approximately 10% of paediatric cardiac arrests the initial identified rhythm is VF or pulseless VT. As soon as recognised, VF or pulseless VT should be treated with unsynchronised DC shock. If onset is witnessed in a monitored environment, a precordial thump may be given, but its efficacy has not been proven.

In contrast to previous recommendations, it is now recommended to give one shock followed immediately by uninterrupted CPR for 2 minutes without pausing to determine if another shock is required. Only in monitored witnessed onset of VF and immediate availability of defibrillation (first dose within 30 seconds) is a stack of up to three shocks (each 4 J kg^{-1})[1,2] without intervening CPR recommended. If ROSC has not occurred within 10 seconds of any of the three shocks, CPR should be commenced. DC shock may be more successful if front and back placement of pads is used.

Failure of VF or pulseless VT to revert immediately to a perfusing rhythm with DC shock (4 J kg^{-1}) should be treated with another single DC shock (4 J kg^{-1}) after 2 minutes of CPR. Persistent VF or pulseless VT should be treated with adrenaline 10 mcg kg^{-1} IV or IO or 100 mcg kg^{-1} ETT followed by another single shock if necessary. Persistent (refractory) or recurrent VF or VT may be also treated with antiarrhythmics (amiodarone, magnesium) interspersed with single DC shocks followed by 2 minutes of CPR. Irrespective of other drug therapy, adrenaline should be administered every 3–5 minutes.

Amiodarone is more efficacious in achieving ROSC than lignocaine for DC shock-resistant VF in adults, but paediatric registry data show no difference between the two drugs.[19] Amiodarone is the preferred drug, but if unavailable, lignocaine is a valid substitute. The dose of amiodarone is 5 mg kg^{-1} IV or IO over several to 60 minutes. It may be repeated to maximum of 15 mg kg^{-1}. The dose of lignocaine 1 mg kg^{-1} IV or IO bolus followed by an infusion if successful at 20–50 mcg kg^{-1} min^{-1}. Lignocaine can be given by ETT at a dose 2–3 mg kg^{-1}. Magnesium, 25–50 mg kg^{-1} (0.10–0.20 mmol kg^{-1}) is indicated for polymorphic VT (*torsades de pointes*).

Management of pulsatile dysrhythmias

Bradysrhythmias

In all age groups, bradycardia is defined as a heart rate <60 min^{-1} or rapidly declining rate with poor perfusion. Sinus bradycardia, sinus arrest with slow

junctional or idioventricular rhythm and atrioventricular block are the most common preterminal arrhythmias in paediatric practice. Untreated, bradycardia will potentially progress to asystole. The treatment is reversal of the cause (hypoxaemia, hypotension, acidosis, hypothermia, intracranial hypertension) and, if unresponsive, adrenaline 10 mcg kg^{-1} IV or IO or 100 mcg kg^{-1} ETT. Bradycardia caused by vagal stimulation should be managed with cessation of the stimulus (e.g. oropharyngeal suctioning, laryngoscopy) and/or atropine 20 mcg kg^{-1} IV or IO (minimum dose 100 µg). Persistent vagal-mediated bradycardia should be treated with adrenaline 10 mcg kg^{-1} IV or IO. If facilities are available, pacing (oesophageal, transcutaneous, transvenous, epicardial) may be effective if sinus node dysfunction or heart block exists.

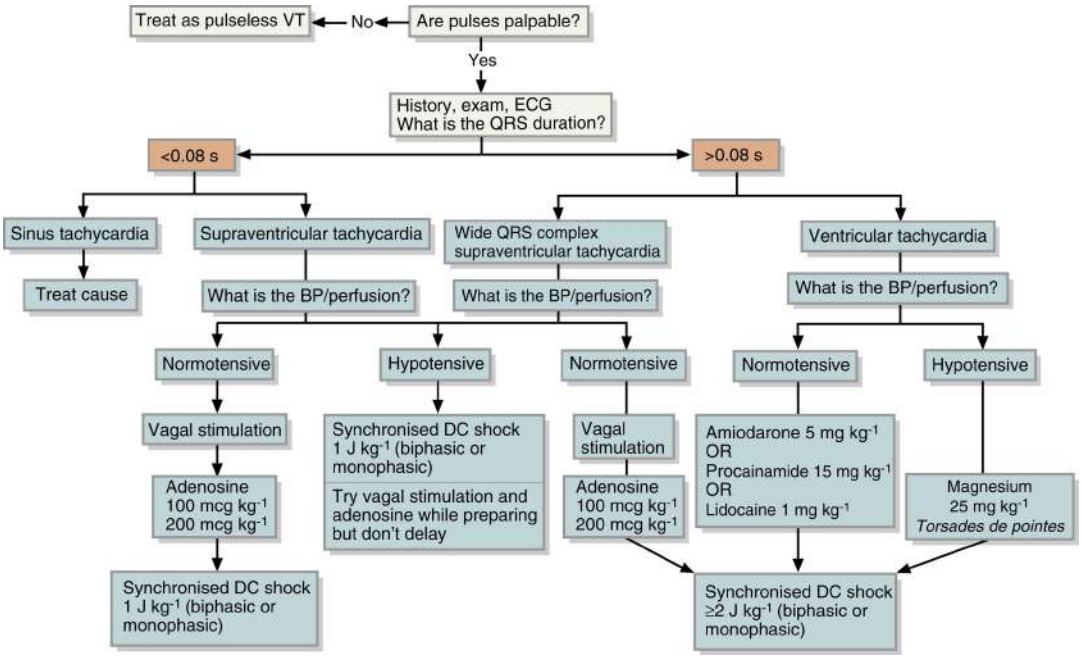

FIG. 2.3.2 Management of pulsatile tachydysrhythmias.

Tachydysrhythmias (Fig. 2.3.2)

Any heart rate above normal for age should be considered a tachydysrhythmia, particularly if associated with poor circulation and hypotension and if the patient has a history of cardiac disease, has had cardiac surgery or could have been poisoned with cardioactive drugs. Of course, such tachycardia may be the result rather than the cause of poor circulation, i.e. sinus tachycardia (ST). It is

important to determine the type and aetiology of the tachycardia, lest drug or other treatment exacerbate the situation. A history related to the tachycardia and a 12-lead ECG should be analysed carefully. If the diagnosis is not obvious, the rate and duration of the QRS are starting points to differentiate sinus tachycardia (ST), ventricular tachycardia (VT), supraventricular tachycardia (SVT) and wide QRS-complex SVT.

Pulsatile ventricular tachycardia

Haemodynamically stable VT may be treated with an antiarrhythmic agent such as amiodarone (5 mg kg^{-1} IV over 20–60 minutes) or procainamide (15 mg kg^{-1} IV over 30–60 minutes) or lignocaine (1 mg kg^{-1} IV over 2–4 minutes). Note that both amiodarone and procainamide prolong QT interval and should not be given together. If *torsade de pointes* (twisting of the peaks) is present, magnesium (25–50 mg kg^{-1}, 0.1–0.2 mmol kg^{-1} IV) may be used. If pulses are present but accompanied by hypotension and poor circulation, cardioversion is needed, in which case it should be synchronised at 2–4 J kg^{-1} under sedation/anaesthesia.

Supraventricular tachycardia

SVT is the most common spontaneous arrhythmia in childhood and infancy. Some infants may tolerate this rhythm for long periods; however, it may cause life-threatening hypotension. It is usually re-entrant with a rate of 220–300 min^{-1} in infants, usually less in children (approximately 180 min^{-1}). The QRS complex is usually narrow (<0.08 seconds), making it difficult sometimes to discern from sinus tachycardia. However, whereas ST is a part of other features of illness, SVT is a singular entity, and whereas the rate in ST is variable with activity or stimulation, it is uniform in SVT and often of sudden onset and offset. In both rhythms, a P wave may be discernible.

If haemodynamically stable (adequate perfusion and blood pressure), initial treatment of SVT should be vagal stimulation. For infants and young children, application to the face of a plastic bag filled with iced-water is often effective or alternatively submersion of the face into a slurry of ice and water in a bowl. Older children may be treated with carotid sinus massage or asking them to perform a Valsalva manoeuvre – such as blowing through a narrow straw. If unsuccessful, give adenosine initially at 0.1 mg kg^{-1} IV by rapid bolus (max dose

6 mg), increasing to 0.2 mg kg^{-1} or 0.3 mg kg^{-1}. In older children, it is important to describe the transient feeling of ‘chest heaviness or breathing difficulty or fearfulness’ that may accompany adenosine administration and/or precede the adenosine with a small amnestic dose of midazolam. If unsuccessful, give synchronised DC shock (cardioversion)[13] initially at 0.5–1.0 J kg^{-1} but subsequently up to 2 J kg^{-1}. If at the outset SVT is accompanied by haemodynamic instability, proceed to cardioversion (synchronised 1.0 J kg^{-1}) immediately, although vagal stimulation or adenosine (IV or IO) may be used, provided they do not delay cardioversion. Verapamil should not be used to treat SVT in infants and should be avoided in children because it induces hypotension by vasodilation and negative inotropic effect. Beta-blockers are the drug of choice for maintaining reversion if there is frequent recurrence and may be required acutely if the SVT is recurrent during a presentation.

Wide QRS complex supraventricular tachycardia

SVT with aberrant conduction may cause a wide-complex QRS (>0.08 seconds) and thus may resemble VT. If the blood pressure is low or circulation deemed inadequate, the rhythm should be regarded as VT and treated with synchronised DC shock at 2-4 J kg^{-1}. If pulses are absent, the rhythm should be regarded as pulseless VT and treated accordingly with DC shock.

Post-resuscitation management (Box 2.3.1)

Supportive care should be provided until there is recovery of function of vital organs. This may require provision of oxygen therapy, mechanical ventilation, inotropic/vasopressor infusion, renal support, parenteral nutrition and other therapy for several days or longer. Recovery of infants and children is usually slow because cardiorespiratory arrest is often secondary to prolonged global ischaemia or hypoxaemia, which implies that other organs sustain damage before cardiorespiratory arrest. The cause of the CPA should be investigated and treated appropriately, e.g. sepsis or drug overdose. Particular care should be taken to ensure adequate cerebral perfusion with well-oxygenated blood. Overoxygenation should be avoided. Hyperventilation to hypocarbic levels is contraindicated because of potential harmful cerebral vasoconstriction.

Peripheral circulatory failure (shock) is common after ROSC. At least the 5th centile of blood pressure appropriate for age should be maintained with the use

of parenteral fluids and inotropic-vasopressor support.[19] Invasive blood pressure measurement is usually required to accurately achieve this recommendation.

Hyperthermia post arrest is associated with a poor prognosis. Survival and neurological outcome are better when targeted temperature management (TTM) is conducted for at least 24 hours.[19] The optimal temperature to target is uncertain, and a target range of either 36–37.5°C or 32–34°C is acceptable for out-of-hospital cardiac arrests. For in-hospital arrests recommendations are less certain, but a TTM strategy avoiding hyperthermia is reasonable. Seizures should be actively sought and treated with anticonvulsant.

Box 2.3.1 Post-resuscitation checklist

Oxygenation and ventilation

Measure oxygenation and target normoxemia
Avoid hypoxia
Measure PaCO2, and target a clinically appropriate value
Avoid hypocapnia

Haemodynamic monitoring

Monitor blood pressure
Set haemodynamic goals during post-resuscitation care
Use parenteral fluids and/or inotropes or vasopressors to maintain a systolic blood pressure greater than the fifth percentile

Targeted temperature management

Measure and monitor core temperature; prevent and treat fever
In children, apply TTM (32–34°C or 36–37.5°C) for at least 24 h if unresponsive after ROSC. Prevent fever if rewarming

Neuromonitoring

Treat clinical seizures
Do not routinely use pharmacologic prophylaxis for seizures

Glucose control

Measure glucose
Avoid hypoglycaemia

Adapted from Part 6: Pediatric basic life support and pediatric life support: 2015 International Consensus on Cardiopulmonary Resuscitation and Emergency Cardiovascular Care Science with Treatment Recommendations. Resuscitation, Volume 95, October 2015, Pages e147–e168.

Blood sugar should be measured and controlled. In adults high blood sugar is associated with worse outcomes, and in the neonatal period, low blood sugar is associated with worse outcomes after hypoxic brain injury.

Complications of CPR should be sought especially if secondary deterioration occurs. A chest X-ray should be obtained to check the position of the ETT, to exclude pneumothorax, lung collapse, contusion or aspiration and to check the cardiac silhouette.

Cessation of cardiopulmonary resuscitation

Long-term outcome from paediatric CPR is poor, with approximately 5–10% of patients surviving out-of-hospital arrest[14] and 25–50% in-hospital cardiac arrest.[15,16] The decision to cease CPR should be based on a number of factors including the duration of resuscitation, its quality, response to treatment, pre-arrest status of the patient, remediable factors, likely outcome if ultimately successful, opinions of personnel familiar with the patient and, whenever appropriate, the wishes of informed parents. In general, unless hypothermia or drug toxicity exists, survival to normality is most unlikely if there has been a failure to respond to full CPR after 30 minutes and several doses of adrenaline, unless environmental hypothermia was an important aetiological or consequential factor. In the newborn infant, discontinuation of treatment is appropriate if CPR does not establish a spontaneous circulation within 15 minutes.[1] Family members should be kept informed, allowed to be present or asked if they want to be present during resuscitation (see Chapter 2.1).

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2.4

Paediatric resuscitation in specific circumstances

Ben McKenzie, and James Tibballs

Avoiding cardiac arrest during critical care management

When embarking on a critical procedure such as intubation in the critically unwell child, it is imperative to attempt to avoid further clinical deterioration and cardiac arrest.

Making sure the right healthcare professionals are present, that equipment is working and ready, that drug needs are anticipated and that there is a plan for potential difficulties is important. It is important that there is a shared mental model of resuscitation within the assembled team. Checklists, closed loop communication and fostering a shared culture of safety in organisations are likely to help in this regard.

In shocked or hypotensive children who are at the limit of their cardiovascular compensation, modification of rapid sequence induction (RSI) is required. Deterioration during RSI should be anticipated due to the direct effect of vasodilating induction drugs, loss of endogenous catecholamine effects, and loss of spontaneous breathing that aids venous return. RSI drugs with vasodilating or negative inotropic effects such as propofol and thiopentone should be avoided or the dose substantially reduced. The upper dose of muscle relaxant should be used to ensure potency in a compromised circulation. The need for vasopressor/inotropic infusions that is precipitated by RSI should be anticipated and either prepared or commenced before the procedure.

The child with myocarditis/severe fulminant dilated cardiomyopathy is at particular risk of deterioration during intubation. The 2015 International Liaison Committee on Resuscitation (ILCOR) review was unable to recommend a

particular approach, but an approach using the above principles is reasonable. In addition extra-corporal membrane oxygenation (ECMO), if available, may be required for these patients and its need anticipated. ECMO is an invasive form of cardiovascular support that should be considered for infants and children who suffer cardiac arrest.[1]

Anaphylaxis

Adrenaline is the life-saving drug to manage angioedema, bronchospasm and shock from anaphylaxis (see Chapter 22.5). It requires either intramuscular or, if severe, intravenous doses, but advanced life support (ALS) resuscitation algorithms remain the same. More than one dose of 20 ml kg^{-1} of crystalloid may be required to address intravascular volume state and vasodilation. The airway may be difficult if angioedema obscures the view and is the cause of the arrest.

Persons receiving β-blocking drugs have a potentiated risk of anaphylaxis. In such patients, it is more resistant to therapy and lasts longer – higher adrenaline doses should be anticipated.

Asthma

Cardiac arrest in asthma (see Chapter 6.5) is usually the terminal event after progressive worsening of symptoms and clinical deterioration. Arrest has been linked to failure of ventilation and decreased venous return from the following mechanisms: gas trapping hyperinflation (auto positive end-expiratory pressure [PEEP]) causing high intrathoracic pressure, asphyxia from severe bronchospasm, and tension pneumothorax. Less commonly cardiac arrest is from arrhythmias related to hypoxia, electrolyte abnormalities and drug therapy.[2]

Standard resuscitation algorithms should be applied with the following considerations. Endotracheal intubation should be considered early in the arrest as airway pressures are high and ventilation difficult. If gas trapping is suspected, consideration should also be given to disconnection of any bag or ventilator circuit to allow expiration, with or without chest wall compression to assist expiration. Cardiopulmonary resuscitation (CPR) should be continued and will act in assisting expiration. Evaluation for tension pneumothorax is essential.

Continuing bronchodilator administration is important but difficult once ventilatory support is initiated. Administration of drugs either nebulised and through T-pieces in the ventilator circuit or intravenously is required.

Drowning

Victims of submersion incidents suffer global hypoxaemia and, if arrested, global ischaemia. Associated injuries are aspiration pneumonitis and hypothermia. Aspiration of water and gastric contents is common (see Chapter 22.2). In addition, hypothermia (see Chapter 22.4) may be present, but unless the victim was subject to severe environmental hypothermia such as being submersed in ice-cold water (<5°C) this reflects lack of perfusion and is a bad prognostic sign. CPR should not be continued for a prolonged period in an asystolic child solely to warm the patient unless severe environmental hypothermia is the cause.

The outcome is often determined by the extent of neurological injury. Bad prognostic indicators are prolonged duration of submersion, lack of bystander CPR, prolonged pre-hospital resuscitation, pulseless arrhythmia on arrival at hospital, fixed dilated pupils, severe acidosis and apnoea. Nonetheless, vigorous resuscitation should be instituted on arrival in hospital of the pulseless victim, if not already commenced by ambulance personnel, in order to clarify the clinical details.

Intubation and mechanical ventilation with 100% oxygen should be instituted immediately. Regurgitation of stomach contents should be anticipated, and an RSI technique should be used. Sedative drugs with cardiovascular depressive actions should not be used, or in minimally required doses only. The lung compliance is likely to be poor, and it may be necessary to insert a larger-than-usual uncuffed or a cuffed endotracheal tube (preferred) to prevent a leak around the tube, to obtain adequate lung inflation in the setting of acute respiratory distress syndrome (ARDS) in order to achieve oxygenation. After restoration of cardiac rhythm myocardial contractility should be measured with echocardiography and optomised with inotropic agents.

During resuscitation, the goal is to provide maximum opportunity for cerebral recovery, and this is achieved by restoring cerebral perfusion with adequately-oxygenated blood and the avoidance of factors that decrease cerebral perfusion pressure. It is thus vital to restore cardiac output and blood pressure, to oxygenate blood and to avoid factors that would increase intracranial pressure, such as venous obstruction. Hypocapnia, hypercapnia, hypoglycaemia and hyperglycaemia should be avoided and convulsions treated.

Any pulseless arrhythmia may be encountered and should be managed along standard lines.

There are no important clinical differences between fresh and salt-water immersion. Altered levels of serum electrolytes, especially sodium and potassium, may be detected but are uncommon and in any case do not influence acute resuscitation.

Traumatic cardiac arrest

Traumatic cardiac arrest can arise through a limited number of scenarios that should be actively sought and treated (see Section 4). Almost all causes of reversible shock in traumatic cardiac arrest can be addressed by one or a combination of five interventions (although they will not always be successful). They are securing a patent airway, adequate ventilation, restoration of circulating blood volume (while controlling haemorrhage), decompression of any pneumothorax and decompression of pericardial tamponade. Within the framework of a skilled multidisciplinary team managing trauma reception and resuscitation, these are the five interventions that should receive priority in cardiac arrest secondary to trauma in addition to standard resuscitation algorithms.

Comotio cordis is an uncommon cause of cardiac arrest in trauma. Sudden death, usually due to ventricular fibrillation, occurs after a nonpenetrating discrete blow to the precordium (peak age 10–18 years), often during sport such as from a baseball or other sporting object.[3] The standard resuscitation algorithm with defibrillation is indicated.

Craniocervical dislocation in high-energy blunt trauma will also cause cardiac arrest in trauma from high injury to the spinal cord and sometimes brainstem. It may present as a profound bradycardia, hypotension and respiratory arrest from very high level quadriplegia and has a poor prognosis. Early imaging will detect this injury.

Toxicological emergencies

Standard resuscitation procedures apply to all children who have been poisoned and who are critically unwell or in cardiopulmonary arrest (CPA). In addition to this, consideration should be given to specific therapies that may reverse or ameliorate the effect of the poison (antidotes) and decontamination.

Decontamination may be an early consideration if potential exposure to a life-threatening poison can be reduced – for example, removing excess drug from an

intravenous line in iatrogenic poisoning. Decontamination may be also achieved by activated charcoal and whole bowel irrigation, but both of these procedures require either a patient that is alert and compliant or intubated with a secure airway to avoid complications from aspiration of therapy. Decontamination may also have a high priority during resuscitation if a toxic chemical, biological or radioactive (CBR) poison is the cause and poses a potential risk to healthcare professionals.

Standard antidote therapy and supportive care of organ systems should be administered in the critically ill child who is poisoned. It is reasonable to add antidotes to resuscitation algorithms in poisoned patients (one of the 4 Hs and 4 Ts, see last chapter)—for example, hydroxocobalamin for cyanide poisoning in a child exposed from a house fire, or calcium and high-dose insulin euglycaemic therapy in verapamil poisoning, or digoxin antibodies in digoxin poisoning.

Intravenous lipid emulsion

Local anaesthetics block cell membrane sodium channels and impair nerve conduction. Toxicity manifests as perioral tingling, seizures and cardiovascular collapse (cardiac arrest, hypotension, malignant ventricular arrhythmias). Intravenous lipid emulsion (ILE) is a relatively new antidote recognised in the ILCOR 2015 review,[1] particularly for the treatment of local anaesthetic systemic toxicity (LAST). The predominant theory for its mechanism of action is that by creating an expanded, intravascular lipid phase, equilibria are established that drive the offending drug from target tissues into the newly formed ‘lipid sink’. Lipid emulsion could also theoretically increase intracellular fatty acid content and therefore overcome the reduced adenosine triphosphate (ATP) production, which results from LA block of fatty acid transport and oxidation. ILE for systemic toxicity has shown consistent benefit in animal studies involving bupivacaine toxicity[4], and case reports/registry data in humans contain successful outcomes with LAST. Lipid emulsion is usually available as a 20% emulsion of long-chain triglycerides. The dose is 1.5 mL kg^{-1} followed by an infusion of 0.25 mL kg^{-1} $minutes^{-1}$ for 30 to 60 minutes. Its adverse effects include interference with laboratory testing, pancreatitis and pulmonary changes similar to adult respiratory distress syndrome. ILE may change the effectiveness of adrenaline (epinephrine) during cardiac arrest, but there is no certainty to be able to recommend changes to standard resuscitation algorithms regarding adrenaline dosing.

An area of further research is which other poisonings may potentially be treatable with ILE.

Drug-induced asystole

Infusions of KCl are hazardous. Molar solutions of KCl are in common use in intensive care units especially after cardiac surgery. They should not be available for regular ward use. An inadvertent IV bolus of potassium can cause asystole. Note that a 1 mL bolus of molar KCl (1 mmol) in a 10 kg child will theoretically raise the serum concentration by 2.5 mmol L^{-1} and cause immediate asystole.

Immediately acting treatment (within seconds) for hyperkalaemia is either 10% calcium chloride IV 0.2 mL kg^{-1} (or equivalent) or IV sodium bicarbonate 1 mmol kg^{-1} or both. Calcium antagonises the cardiac effects of potassium on the heart, while the bicarbonate lowers the serum concentration of potassium by a small amount. Better treatments of rapid onset (within minutes) are glucose 0.5 g kg^{-1} IV (e.g. 5 mL kg^{-1} of 10%) plus insulin 0.05 units kg^{-1} or salbutamol 0.25 mg kg^{-1} by aerosol or both.

Envenomation

Snake envenomation (see Chapter 22.1, Envenomation)

Australasian elapid snakes variably cause a triad of consumptive defibrinating coagulopathy, descending neurological paralysis and rhabdomyolysis depending on the snake involved. In addition to complications arising from the above clinical manifestations, early collapse and cardiac arrest occurs, particularly with brown snakes (*Pseudonaja* spp.) and rarely with taipan envenomation. The median time to onset of hypotension or collapse was 30 minutes (range 2–90 minutes) in 37 of 136 confirmed brown snake envenomations.[5] Five of these 37 patients died from pre-hospital cardiac arrest. There were no ventricular arrhythmias documented in the group with hypotension. Animal studies suggest that these early cardiovascular effects are caused by hypotension secondary to release of endogenous mediators and are unlikely to be due to direct cardiac effects. The cardiovascular effects are likely to respond to antivenom.[6]

Haemorrhage (particulary intracranial) from coagulopathy and respiratory failure from neurological paralysis from neurotoxins are uncommon potential

ways of a patient presenting in extremis post snake-bite envenomation.

Any patient with cardiac arrest secondary to suspected snake envenomation should receive one vial of the relevant snake antivenom for each snake species suspected. Antivenom is diluted in a 1:10 volume of normal saline and administered over 5 minutes/slow push during cardiac arrest. The incidence of anaphylaxis to antivenom is approximately 20% (10% severe)[7] and should be anticipated.

Marine envenomation

Box jellyfish (*Chironex Fleckeri*)

Most box jellyfish cause severe local pain requiring first aid, transport to hospital and analgesia. However, severe envenoming may result in cardiovascular collapse and death within 30 minutes, occurring more commonly in children compared with adults. In these rare extreme envenomations, survival depends on early CPR and adherence to standard resuscitation algorithms.

Treatment should consist of decontamination by removing the tentacles by hand or by washing off with sea water, applying vinegar liberally to inactivate remaining nematocysts, and applying ice packs and administering analgesia.

In the arrested patient not responding to conventional resuscitation, one vial of box jellyfish antivenom should be administered intravenously over 5 minutes in 1:10 dilution of normal saline (this may be repeated up to six vials). Magnesium sulfate 50 mg kg^{-1} as an IV bolus over 5 to 15 minutes may also play a role in refractory cases.[8]

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2.5

Shock

Robert Melvin

ESSENTIALS

1 Shock is a syndrome that arises because of acute failure of the circulation resulting in inadequate tissue perfusion. It may result from hypovolaemic, distributive, cardiogenic, obstructive or dissociative causes.

2 The normotensive child may have profound compensated shock.

3 As tachycardia may be a non-specific sign and hypotension occurs late, it is crucial to recognise the early features of shock in a child by assessing indices of peripheral perfusion and alterations of end-organ function.

4 The initial management of shock should be volume expansion with a bolus of 20 mL kg^{-1} crystalloid. Further boluses are given according to clinical response. Occasionally, in the exsanguinating child, universal donor blood is indicated.

5 The ongoing management of the shocked child will depend on the specific cause and may include interventions such as oxygenation, ventilation, glucose administration, cardioversion, broad-spectrum antibiotics, inotropic support, ductus arteriosus manipulation, adrenaline (epinephrine), atropine or surgical intervention.

Introduction

Shock is a syndrome that arises because of acute failure of the circulation. This

acute circulatory failure results in inadequate tissue oxygenation and inability to remove the waste products of metabolism. Shock is complex, having many causes and many manifestations. The clinical diagnosis and management of shock are complicated by the fact that many organ systems become involved and because many of the ‘signs of shock’ actually arise because of the body’s attempts at homeostasis rather than because of the underlying process.

The adequacy of circulation, and thus tissue perfusion, requires proper functioning of the heart, vessels and blood. The heart must pump enough blood to meet peripheral oxygen demand; the vessels that deliver the blood to both the lungs and the other organs must be patent (that is not obstructed), be regulated appropriately at a macro level to ensure delivery of blood at an appropriate pressure to the organs requiring oxygen, and must function appropriately in the periphery so as to allow oxygen diffusion without fluid loss. The blood must have sufficient oxygen-carrying capacity and must maintain the ability to allow oxygen exchange. Failure of any aspect of this complicated system will result in inadequate peripheral tissue perfusion and therefore shock.

Conventionally, shock is divided into the following five types (in order of frequency):

- Hypovolaemic
- Distributive
- Cardiogenic
- Obstructive
- Dissociative.

Hypovolaemic shock arises when there is circulatory volume inadequacy. Most commonly this occurs following gastrointestinal fluid losses such as with diarrhoea and vomiting or third space losses such as in intussusception or volvulus. Hypovolaemia in trauma occurs due to haemorrhage or burns. In distributive shock, peripheral vascular abnormalities result in failure to appropriately distribute pumped blood. This can be because of infection (as in the septic child), allergic reaction, spinal cord injury or drugs. Cardiogenic shock occurs when the heart fails to pump enough fluid. This can occur because the heart itself is failing secondary to infection, because the heart muscle has been injured (such as in cardiac contusion) or because of a problem with rhythm where the heart is beating too slow or too fast to achieve an adequate cardiac output. Obstructive shock is less common and arises when there are

abnormalities of flow. This can either be because particular vessels themselves are obstructed (such as in pulmonary embolus) or because of extra vascular abnormalities which obstruct the flow of blood (such as tension pneumothorax or cardiac tamponade). Finally, dissociative shock arises when the oxygen-carrying capacity of the blood is too low (profound anaemia) or has been reduced as in carbon monoxide poisoning.

Diagnosis and assessment

Early consideration and recognition of shock are key in all ill children. This requires a systematic approach to assessment that involves looking at the airway, breathing, circulation, and neurological status and exposing the child. Knowledge of age-related haemodynamic parameters is very important.

Airway

Generally, shock affects the airway by reducing conscious level. The presence of adequate breathing (as assessed by looking at chest wall movement, listening and feeling for exhaled air) also indicates an adequate airway.

In acute allergic reaction, airway obstruction (by swelling in the upper airway) and shock due to changes in vascular tone can both arise as part of the same process.

Breathing

Tachypnoea (increased respiratory rate) is often the first sign of shock. The effort of breathing, efficacy of breathing, oxygen saturations and effects of inadequate respiration should be examined.

Circulation

Circulation is the crucial component of shock assessment since homeostatic mechanisms function particularly well in children and compensation for inadequate circulatory function is good. Consequently, in a child it can be difficult to detect circulatory failure until a late stage; hence, one needs to be vigilant in detecting the earlier features of compensating shock.

Circulatory assessment should look at both circulatory status and also at the effects of inadequate circulation on other organ systems.

In paediatrics, capillary refill time, skin pallor or skin temperature can be as useful in detecting early signs of compensated shock as well as the more obvious measures of heart rate and blood pressure.

Heart rate

Tachycardia (relative to age norm) is a key sign of shock (see Chapter 1.1). This tachycardia is a homeostatic response to maintaining cardiac output. In overwhelming shock, bradycardia may occur and untreated will progress to asystole. The peripheral pulses may be weak, thready or absent.

Blood pressure

While it is true that a child with hypotension is shocked it is not true to say that a child that is normotensive is not shocked. This is because maintenance of blood pressure is the key end point of the homeostatic response, and decompensation (and thereby hypotension) occurs late and may be precipitous. The accurate measurement of blood pressure in a distressed child can be a challenge in itself; cuff size and placement are important, and a judgement as to whether a particular blood pressure is normal or abnormal in a particular age group in any particular circumstance is also challenging. Many widely quoted normal ranges by age are broad – furthermore many of these were derived in the resting outpatient child rather than in the seriously ill or injured child.

Capillary refill

The capillary refill time (CRT) is measured by applying pressure to the nail bed or other area with visible circulation for some 5 seconds and then measuring the length of time it takes for the blanching to disappear. Central capillary refill such as over the sternum is more reliable than the peripheries which can be influenced by vasoconstriction due to the ambient temperature effects:

- CRT <2 seconds is normal
- CRT 2–4 seconds should prompt consideration of shock
- CRT >4 seconds is definitely abnormal.

Changes in peripheral circulation (which are brought about by an increase in catecholamines) may also show as an increased core to periphery temperature

difference (more than 2°C is a sign of poor perfusion) or as a noticeable centripetal temperature gradient. Reduced skin perfusion may result in a mottled, cyanosed, pale appearance and coolness on palpation. Some infants have physiological mottling (cutis marmoratum) in the first months of life due to ‘immaturity’ of the autonomic control of skin vessels, which commonly occurs with exposure. One must clarify with parents whether this mottled appearance is ‘more than usual’ in their child, as infants with cutis marmoratum will become more mottled if they have an accompanying illness causing shock.

Effects of circulatory inadequacy including neurological status

Increasing respiratory rate is an early sign of inadequate circulation. Latterly, as acidosis ensues, deep sighing respirations occur due to ‘air hunger’.

Agitation and altered conscious level (that is response to voice or less) are important signs of circulatory inadequacy and the resultant cerebral hypoperfusion. In infants, the manifestations may be more subtle and increased ‘irritability’ or ‘floppiness’ or failure to recognise and make eye contact with parents may be the only obvious manifestation.

Consider hypoglycaemia as a cause of decreased conscious level in shock.

Urine output will decrease with inadequate circulation. If a child has a urine collection device in place then a flow of some 2 mL kg^{-1} hr^{-1} in the child under 1 year and 1 mL kg^{-1} hr^{-1} in older children indicates adequate renal perfusion. There is often no urinary collection device initially, but the weight of wet nappies may be helpful.

Initial management

The child with shock should be managed in the resuscitation area with monitoring of heart rate, blood pressure, respiratory rate, temperature and oxygen saturation. Urine output should also be monitored as an indicator of response to therapy. The airway and breathing should be managed as in any other case of the seriously ill child, with high-flow supplemental oxygen delivered. If ventilation is inadequate then this should be supported in the first instance with a bag-valve-mask device. Consideration should then be given to intubation and ventilation through an endotracheal tube if there is no improvement.

Circulation

Once airway and breathing have been managed, the next priority is to gain intravascular access. This should be obtained rapidly. An initial assessment to see whether there is a vein available to allow the placement of a short, relatively large, peripheral venous catheter is made. Up to two attempts can be allowed taking no longer than 5 minutes to perform. If these attempts are unsuccessful (or if there is no possibility of placing a venous catheter) then intraosseous access should be gained. In most cases this is done over the medial aspect of the tibia just distal to the knee (see Chapter 24.8).

If neither peripheral venous access nor intraosseous access is possible or desirable, then a Seldinger (guide wire) approach to the femoral vein is the next route of choice.

As soon as intravascular access is obtained then blood should be drawn to be tested for haemoglobin, white cell count and platelets together with urea, electrolytes, acid–base and lactate level. A blood culture should be taken and a glucose stick test performed to exclude hypoglycaemia.

An initial bolus of 20 mL kg^{-1} of fluid should be given. In most cases the initial fluid will be crystalloid, but occasionally universal donor blood may be indicated. A second fluid bolus of 20 mL kg^{-1} can then be used and titrated to clinical and biochemical response. The FEAST study showed that in certain areas of the world maintenance fluids alone, rather than a bolus, improved survival at 48 hours for certain causes of shock (not diarrhoea, trauma or burns patients).

If no other cause of shock can be found then it is reasonable to give a broad-spectrum antibiotic as part of the initial treatment, as sepsis is the most common precipitant of shock in children. A third-generation cephalosporin should be given immediately after blood cultures are taken, e.g. cefotaxime or ceftriaxone 50 mg kg^{-1}.

If a glucose stick test reveals significant hypoglycaemia (<3 mmol/L) then glucose should be administered (5 mL kg^{-1} of 10% dextrose). Hypoglycaemia may be the primary problem, but it may also coexist with other causes of serious illness, and resuscitation must therefore be continued if immediate recovery does not ensue with correction of the blood glucose.

If a tachydysrhythmia is identified as a cause of established shock then cardioversion is indicated. This should be undertaken without delay. If the child is alert or otherwise responsive then sedation is usually indicated. If the

tachydysrhythmia is supraventricular tachycardia then it may be quicker to give a single bolus of adenosine while preparing for cardioversion (see Chapter 5.10).

Further management

If the child is not responding to initial management, then consider early referral/transfer to a paediatric centre. Usually, however, once the initial assessment and stabilisation are complete, it is then possible to take a more detailed history and undertake a comprehensive examination to try and establish the underlying condition.

The following specific conditions are dealt with in more detail below:

- Hypovolaemia
- Sepsis
- Acute severe allergic reaction (anaphylaxis)
- Duct-dependent congenital heart disease
- Heart failure
- Neurogenic shock.

Hypovolaemia

This diagnosis is usually made because of the presence of trauma, fluid loss from vomiting or diarrhoea, or a surgical abdomen.

Further treatment depends on the response to initial fluid bolus. If there are still signs of shock then a second 20 mL kg^{-1} bolus should be given. At this stage a surgical opinion should be sought if an underlying surgical cause is suspected. In trauma patients, early consideration of blood products should be made. Packed red blood cells (10 mL kg^{-1}) should be given in conjunction with fresh frozen plasma and platelets, and one proposed ratio is 1:1:1 as the initial set of red cells, fresh frozen plasma and platelets.

It would be most unusual for a child with gastroenteritis to require more than two boluses of crystalloid and, if this is the case, then an alternative diagnosis (such as an underlying intra-abdominal surgical problem or adrenal insufficiency) should be considered.

If the child shows no further signs of shock after two fluid boluses and the underlying diagnosis is gastroenteritis, then it will still be necessary to correct any underlying dehydration, and this should be done in the normal manner (see

Chapter 7.12).

Septic shock

Septic shock arises because of a complex combination of hypovolaemia (relative and absolute), cardiogenic shock (due to myocardial depression) and distributive shock. The underlying cause is, of course, the infection, and this should be treated as a matter of urgency. As previously stated, any shocked child in whom there is not an obvious diagnosis should receive broad-spectrum antibiotics as part of the initial management. If a specific diagnosis of septicaemic shock is made and antibiotics have not been given then these broad-spectrum antibiotics (third-generation cephalosporins) should be given immediately. Consideration should also be given for anti-staphylococcal antibiotics such as flucloxacillin and vancomycin if there is evidence of cellulitis or a foreign body or if the clinical picture is of toxic shock syndrome (high fever, confusion, scarlatina-form rash with desquamation and subcutaneous oedema).

In all cases a further bolus of 20 mL kg^{-1} of fluid should be given if there is not rapid restoration of normal circulation following the first bolus. Serial measurement of lactate may be useful in titrating fluid responsiveness. More than 4 mmol dL is suggestive of severe shock, and <2 mmol/dL should be aimed for after treatment. With severe septic shock, urgent consideration of rapid sequence induction and elective intubation should be given. Many children will develop a degree of pulmonary oedema if a third fluid bolus is given, and oxygenation can only be maintained by positive pressure ventilation (often with the addition of positive end-expiratory pressure).

Cardiogenic shock is also a feature of sepsis, and this will require specific treatment. Adrenaline (epinephrine) at a rate of 0.05–0.2 mcg kg^{-1} min^{-1} should be commenced and adjusted according to the response. Noradrenaline (norepinephrine) may also be used instead. Both are suitable to be commenced peripherally in the emergency department.

Features of raised intracranial pressure should prompt consideration of sepsis due to meningitis. This is usually presaged by a decrease in conscious level together with abnormal posturing or focal neurology. Early appropriate treatment should be commenced, and this will include steroids, diuresis, intubation and ventilation and appropriate positioning of the patient. In this situation, a lumbar puncture is contra-indicated.

Acute severe allergic reaction (anaphylaxis)

A diagnosis of acute severe allergic reaction (anaphylaxis) is likely with an acute onset of hypotension or bronchospasm or upper airway obstruction, even if the typical urticarial rash is not present. It is more likely if there is a history of previous reactions.

Anaphylactic circulatory shock will usually respond to adrenaline. The recommended dose is 10 mcg kg^{-1} given intramuscularly into the lateral thigh. This can be repeated every 5 minutes as needed. Nebulised adrenaline can also be administered for upper airway obstruction.

If circulatory shock is resistant to the initial boluses of adrenaline then an intravenous bolus of 20 mL kg^{-1} of fluid should be given. At this stage, if the diagnosis is clear, then an infusion of adrenaline at 0.1 mcg kg^{-1} min^{-1} (and titrated to effect) should be commenced.

There is less urgency to prescribe antihistamines or steroids as neither has been proven to be effective in anaphylaxis.

Duct-dependent congenital heart disease

There are a number of congenital heart defects in which the presence of a patent ductus arteriosus is essential to the maintenance of pulmonary or systemic circulation. These conditions include pulmonary atresia, hypoplastic left heart syndrome, coarctation and critical aortic stenosis. The ductus usually closes functionally within 24 hours of birth – but may remain patent in the presence of cardiac abnormality.

Babies with critical pulmonary lesions will present within a few days of birth with tachypnoea and apparent breathlessness together with cyanosis, while those with systemic blood flow reduction will present with failure to feed, apparent breathlessness and collapse with poor peripheral circulation. These presentations are very similar to severe sepsis, and empirical antibiotics should be given until sepsis is excluded.

On examination, the babies are in heart failure (usually without a characteristic heart murmur), often with an enlarged liver and a gallop rhythm.

Immediate treatment is to maintain or increase ductus size with an infusion of alprostodil (0.05 mcg kg^{-1} min^{-1}). Immediate transfer to a cardiological centre is indicated. It will usually be necessary to intubate and ventilate these children before transfer.

Heart failure

This is uncommon in paediatrics. In babies, heart failure is usually due to structural heart disease, while in older children myocarditis and cardiomyopathy are the commonest diagnoses.

Babies present with breathlessness, feeding problems and failure to thrive and restlessness. In older children more general complaints such as fatigue, anorexia, exercise intolerance and cough are common.

There are signs of increased effort of breathing with increased respiratory rate, recession and positioning. There is an increased heart rate with cool, pale peripheries together with hepatomegaly. On auscultation there may be a gallop rhythm and basal crepitations.

Oxygen should be given in all cases and broad-spectrum antibiotics administered if there is any suspicion of sepsis. As discussed earlier, alprostadil should be given if the lesion is potentially duct dependent. In cases of cardiomyopathy, diuretics (frusemide 0.5–1 mg $kg^{-1)}$ should be given to offload the heart. A dobutamine infusion may be indicated to support the failing heart while urgent cardiological advice is sought.

Neurogenic shock

This is a diagnosis of exclusion, and hypovolaemia due to trauma must always be investigated first, as neurogenic shock is invariably caused by trauma in children. Bleeding sources should be actively sought and managed in all children in whom hypotension persists, with early surgical consultation.

A child with a spinal injury above T6 will have impaired sympathetic tone below this level once the initial catecholamine release that occurs at the time of injury has ceased to have an effect. The systemic vascular resistance will fall, and the reflex tachycardia usually seen as a response to hypovolaemia will not occur. The overall outcome is generalised vasodilatation, bradycardia and loss of temperature control. Systolic blood pressure will fall below 90 mmHg but the skin will appear paradoxically warm and pink.

Fluid management with a single bolus of 20 mL kg^{-1} will usually achieve an acceptable blood pressure. Atropine (20 mcg kg^{-1}) can be given if heart rate falls below 50. Very careful handling is necessary as these children may suffer from postural hypotension if tipped or lifted suddenly. In the early stages fluid management is very important, and this may require the insertion of arterial and

pulmonary lines. Urinary catheterisation will be necessary as bladder control will be lost.

Further reading

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2.6

Sepsis recognition and initial management

Dr Elliot Long, and Dr Anthony Delaney

ESSENTIALS

1 Paediatric sepsis is the final common pathway for many decompensated infections.

2 Clinician judgement is the best performing tool for early recognition of paediatric sepsis; vital signs are dynamic and prone to confounding, and screening blood tests have not been validated.

3 Initial assessment and management include intravenous access, sampling for blood culture and venous blood gas, and early administration of empiric intravenous antibiotics.

4 Fluid resuscitation should be titrated carefully to avoid the harms associated with inadequate and excessive administration.

5 Inopressors may be safely administered via peripheral intravenous cannula in children.

Introduction

Paediatric sepsis is a clinical emergency. Early recognition, prompt treatment with antibiotics, and carefully titrated fluid resuscitation are associated with improved outcomes. While the value of protocolisation in sepsis has been questioned, quality improvement initiatives are consistently associated with improved outcomes. Future directions include the use of ultrasound for assessment of the heart and lungs for haemodynamic resuscitation and the use of

novel methods for assessing tissue perfusion as a therapeutic target.

Background

Sepsis is a major public health concern. It accounts for over 5% of all paediatric hospital admissions, over 10% of paediatric intensive care unit admissions, and over $4.8 billion of total United States hospital costs in 2013.[1] Mortality from paediatric sepsis is 5–17% in industrialised countries and 20–30% in low and middle income countries.[2] Worldwide, sepsis causes over 6 million childhood deaths per year, primarily from pneumonia, severe diarrhoeal illness, and malaria. Paediatric sepsis has a bimodal age distribution, being most common in infancy, and a decreasing incidence until late adolescence.

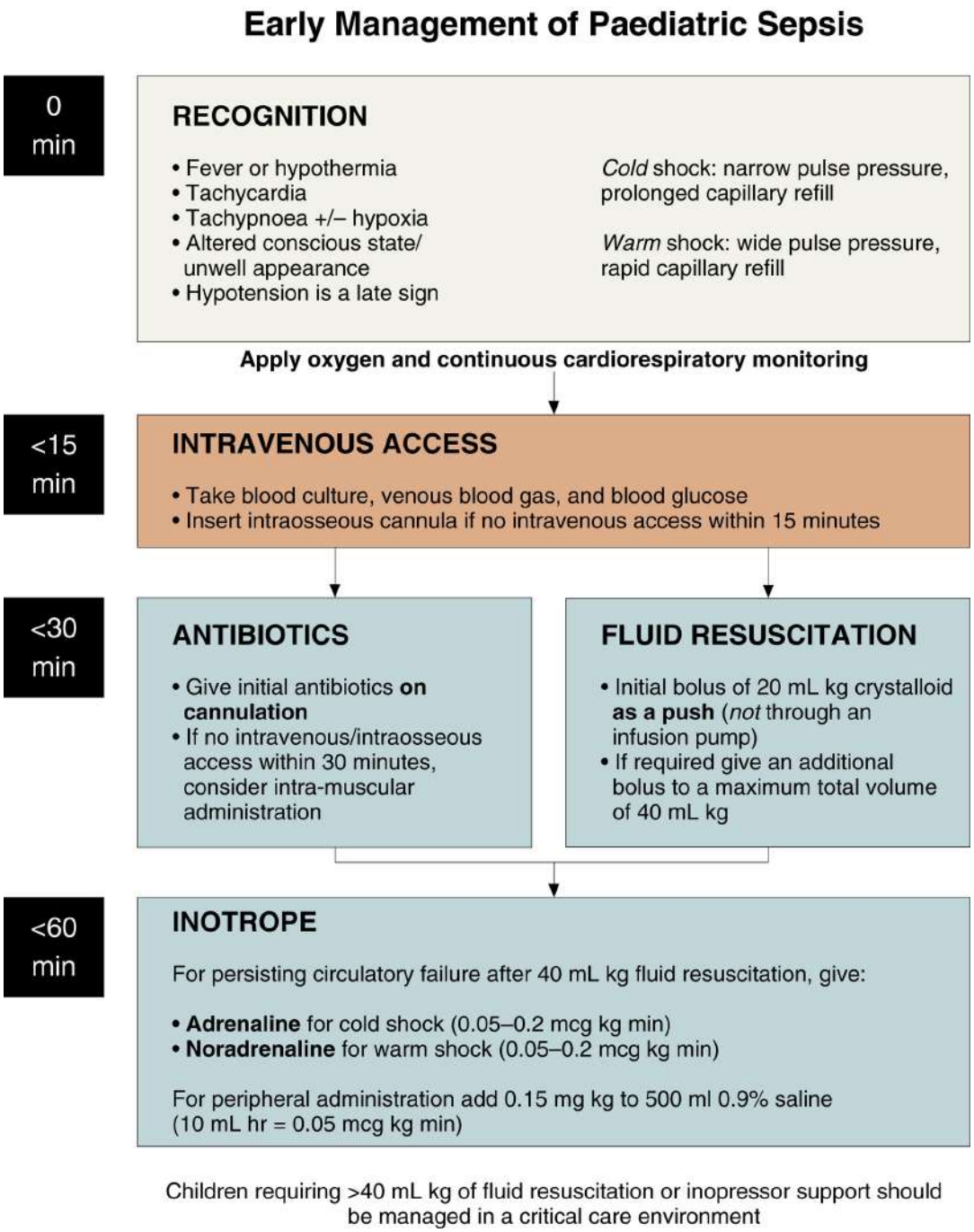

FIG. 2.6.1 Flow diagram for the initial management of paediatric sepsis.

Definition

Sepsis in broad terms can be described as a dysregulated host response to life-threatening infection. The international consensus definition for paediatric sepsis was last revised in 2005 and includes systemic inflammatory response syndrome

(SIRS) due to suspected infection (Fig. 2.6.1).[3] SIRS is defined in terms of vital signs breaching age-based threshold values. The consensus definition for paediatric severe sepsis and septic shock includes cardiovascular organ dysfunction criteria. Importantly, hypotension is not used in defining paediatric sepsis, and biochemical tests such as venous lactate have not been validated as screening tests for diagnosing occult paediatric sepsis.

Aetiology

The most common pathogens causing paediatric sepsis in Australia are *Staphylococcus* and *Streptococcus* species. For infants <3 months of age, the most common pathogens are *E. coli* and group B streptococcus. *Listeria* is very uncommon. Geographic location, immunisation status, immunodeficiency (primary or secondary), and presence of indwelling vascular catheters all influence the microbiological causes of paediatric sepsis.

Pathophysiology

Sepsis occurs when the host response to infection becomes dysregulated. As such, both pro- and anti-inflammatory pathways become activated, contributing to cell injury and the progression to multiorgan dysfunction and death. Traditionally, the haemodynamic effects of sepsis have been described as a combination of relative hypovolaemia, distributive, and cardiogenic shock. While endothelial dysfunction and capillary leak are well described in sepsis, the contribution of aggressive fluid resuscitation to this process through damage to the endothelial glycocalyx and release of atrial natriuretic peptide is becoming clearer. Cardiac dysfunction in paediatric sepsis is well described, with a high incidence of systolic and diastolic dysfunction particularly in younger children and infants.

Diagnosis

Heart rate, temperature, capillary refill, and conscious state are usually abnormal in sepsis but are all dynamic parameters and may change over time. There is also considerable overlap between perfusion abnormalities observed both in early sepsis and during the viraemic phase of self-limited viral infections. Therefore vital signs alone or in combination have poor test characteristics for early

recognition of sepsis. A high index of suspicion for sepsis is required in infants and neonates due to the non-specific nature of presenting symptoms. Triage tools for early sepsis recognition have a high false-positive rate and a low positive-predictive value. The correlation between clinically diagnosed sepsis and sepsis diagnosed using research criteria or coding criteria is poor.

Paediatric sepsis may be subdivided into cold and warm shock, with impaired cardiac function characterising the former, and low systemic vascular resistance characterising the latter. Clinically this is reflected by a narrow pulse pressure and prolonged capillary refill time in cold shock and a wide pulse pressure with 'flash' capillary refill in warm shock. Cold shock is more common in infants and neonates, while warm shock is more common in older children and adolescents. Importantly, infants and neonates with sepsis may present with hypothermia or bradycardia rather than fever or tachycardia. In addition, neonates with cardiovascular collapse should have alternative diagnoses considered, including congenital cardiac disease, endocrine disease, metabolic disease, trauma, or gastrointestinal catastrophe.

Initial emergency management (Fig. 2.6.1)

If sepsis is suspected, children should be managed in a resuscitation room environment. If a local sepsis guideline exists, it should be followed.

Monitoring

Attach continuous electrocardiography, respiratory rate, and pulse oximetry and frequently cycled non-invasive blood pressure.

General supportive measures

Oxygen should be applied for hypoxia. Non-invasive respiratory support may be used for hypoxic respiratory failure not responding to supplemental oxygen. High flow nasal cannula, continuous positive airway pressure, or bi-level positive airway pressure may be used depending on patient age and interface tolerance. Intubation of children with sepsis carries a high risk of precipitating complete cardiovascular collapse. This risk can be mitigated through adequate haemodynamic resuscitation prior to intubation, careful selection and dose titration of induction agent.

Intravenous access

Peripheral intravenous access should be obtained. Ultrasound guidance or trans-illumination may be helpful for difficult intravenous access. For severely unwell children, the most experienced operator should perform this procedure. If peripheral intravenous access has not been secured within 15 minutes, intraosseous access should be obtained.

Initial blood tests

A blood culture and venous blood gas should be taken for all cases of suspected sepsis. Additional blood tests, such as full blood count, electrolytes, liver function, and serology may be obtained if they do not delay resuscitation. Inflammatory markers such as C-reactive protein and procalcitonin have no role in sepsis resuscitation.

Initial treatment

Empiric intravenous antibiotics should be administered on cannulation. If intravenous or intraosseous access is not obtained within 30 minutes of sepsis recognition, most antibiotics can be administered via the intramuscular route. Empiric antibiotic choice should be based on local prevalence and resistance patterns.

Fluid resuscitation for haemodynamic compromise should be with a crystalloid solution (0.9% saline, Hartmann's solution, Ringers lactate, or Plasmalyte®) at a dose of 20 mL kg. The use of albumin is limited by cost, and semi-synthetic colloids have been associated with increased rates of renal replacement therapy and death. Excessive fluid administration is associated with end-organ oedema and dysfunction and in some populations with increased mortality;[4] therefore, fluid resuscitation should be administered carefully with attention to both beneficial and harmful effects.

The administration of 40 mL kg of resuscitation fluid should be a stop-point for re-evaluation of the patient diagnosis, consideration for vasopressor/inotrope infusion, and involvement of local critical care teams. If 0.9% saline has been used as an initial resuscitation fluid, the development of hyperchloraemic metabolic acidosis should prompt the use of a balanced crystalloid.

Inopressor support should start with adrenaline first line for cold shock and noradrenaline for warm shock.[5] Both may be safely administered via a peripheral

intravenous cannula, using a 10-fold dilution. Dopamine and dobutamine as initial inopressors have been associated with higher mortality in paediatric septic shock.

Therapeutic targets

Improvement in vital signs, perfusion, and conscious state are the current standard for monitoring response to treatment. There is limited evidence, however, that vital signs accurately reflect illness severity or response to treatment. Clearance of lactate has not been validated as a marker of adequate resuscitation in children. Harm from excessive fluid resuscitation is also monitored using clinical examination findings, though it remains unclear at what stage during fluid resuscitation these signs develop.

Disposition

Patients requiring >40 mL kg of intravenous fluids or inopressor support should be managed in a critical care environment. This may be an indication to transfer a patient to a paediatric referral centre; local pathways for consultation and transportation should be followed.

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SECTION 3

Neonatal Emergencies

OUTLINE

3.1

The normal neonate

Richard Aickin, and Sasha Rossaye

Definition and introduction

A neonate is an infant aged from birth to 28 days of life. This is a period of rapid change and vulnerability for the infant and his/her parents. Neonates are at higher risk than older infants to serious bacterial infections and may also present with previously unsuspected congenital abnormalities. Many parents have limited family support and little experience to guide them through this time. Stress and anxiety can be at a peak during the middle of the night when the emergency department (ED) is often the only service available. It is important for emergency clinicians to be familiar with the wide range of normal appearance, behaviour and development in neonates to avoid unnecessary and unhelpful over-investigation and treatment. Familiarity is best gained through the experience of evaluating large numbers of young infants while maintaining an open and curious attitude to learning – a challenge at 2 am!

Immediately after birth

Babies are sometimes born unexpectedly in the ED or the immediate vicinity (such as the back seat of the car in the ambulance bay or car park). A midwife or paediatrician may not be immediately available, and the ED staff will be responsible for the initial management of both mother and child.

Birth at term is usually an uncomplicated process producing a healthy infant; however, up to 1% of infants do require some type of resuscitation at birth. Preterm delivery is associated with higher risks and the need for a different approach and specialist assistance.

Emergency clinicians should be familiar with newborn resuscitation (see Chapter 3.5, Neonatal resuscitation) and attend skills training with regular refreshers to maintain competence. It is unlikely that emergency staff will be

required to perform newborn resuscitation sufficiently often to maintain skills without focused training sessions.

A newborn with good tone (flexed limbs, moving) and who responds to gentle stimulation (drying with a soft warm towel) by breathing/crying does not need resuscitation. Most newborn babies simply require drying and stimulating with a warm towel before being placed on the mother's chest for skin-to-skin contact. Babies are blue immediately after birth and can take several minutes to pink up. They do not require bag/mask ventilation nor oxygen provided that they have a satisfactory heart rate (>100/min) and establish regular breathing after their initial gasps. Excessive oxygenation may be harmful to newborn infants. Guidance on normal oximetry measurements and other details are available in ANZCOR newborn resuscitation guidelines (http://www.nzrc.org.nz/assets/Guidelines/Neonatal-Resus).

There is no rush to divide the umbilical cord unless there are urgent reasons to move and resuscitate mother or baby. It is possible that delaying cord clamping for at least 30 to 60 seconds may be an advantage to the infant.

Newborn babies are vulnerable to heat loss. Mother and baby should be looked after in a warm area and shielded from radiant, conductive, evaporative and convective heat loss.

The neonatal history

A neonatal history should include a strong focus on maternal health, history of the pregnancy and delivery, and antenatal screening. A number of maternal problems during pregnancy can place the infant at risk such as diabetes, infections, certain medications or illicit drug use. Prolonged rupture of membranes prior to delivery places the infant at greater risk of sepsis from organisms such as group B streptococcus. Postnatal depression is common and can present with feeding difficulties, poor weight gain and general anxiety about the baby's health. Birth weight should always be recorded and compared with current weight. All infants lose weight after birth, commonly up to 10% of birth weight. Most infants regain their birth weight by 10 to 12 days of age. Feeding can be a source of difficulty even in infants who are otherwise well, and this is a key sign of illness and illness severity. Jaundice is frequent and usually normal or physiological (see Chapter 3.4, Acute neonatal emergencies). Physiological jaundice appears on the first 1–2 days of life and usually peaks on days 4–5 of life. It may persist for several weeks, especially in breast-fed infants.

Physiological jaundice is unconjugated, with a conjugated component of <10% of the total. Jaundice which is unusually prolonged or continuing to increase after the first week of life should be evaluated to rule out biliary atresia and other liver disease. It is important to diagnose biliary atresia early, before six weeks of age, to achieve good treatment outcomes. Fever, on history or examination, is not always present in neonatal sepsis. Serious infection can present with poor feeding, sleepiness, pallor, hypotonia and normal or low temperature. Fever when present is an important sign which should normally trigger investigations for significant infections.

The neonatal examination

A standard newborn examination involves careful observation and a systematic head-to-toe survey for potential problems. The same approach can be used as a general screening examination for a neonate presenting to the ED.

The baby should be fully undressed in a warm environment. The infant's spontaneous posture and movements should be noted. A healthy term infant holds his/her limbs in a slightly flexed posture and will make active movements of all four limbs to a roughly equal extent. Primitive reflexes such as the Moro and grasp reflexes are a way to elicit active movement and compare right with left. Asymmetrical movement may be due to central nervous system (CNS) problems or birth injury such as Erb's palsy or a fractured clavicle.

Examining the head and scalp will reveal open fontanelles and sutures. Extensive moulding of the skull can occur during birth resulting in overlapping sutures and temporary changes to head shape. A caput or scalp swelling is also frequently present. The fontanelle is usually soft and pulsatile and may normally be raised, flat or sunken. A soft raised fontanelle does not indicate a problem in an otherwise well infant.

The infant's eyes should be inspected for symmetrical round pupils and a normal red reflex. A 'key hole' shaped pupil or coloboma may be associated with CNS abnormalities, and the absence of a red reflex can indicate an obstruction to light passing to/from the retina or an abnormality of the retina itself.

Sticky eyes do not necessarily indicate conjunctivitis. Obstructed tear ducts can take several months after birth to open up, and the infant may have persistent dry mucus accumulation in the corner of the eye. Redness, eyelid swelling and the presence of pus suggest infection.

The oropharynx should be inspected to exclude a cleft of the hard and/or soft palate, including palpation.

Nasal obstruction is common for infants with upper respiratory infections and may make it difficult for the infant to breathe and feed at the same time. Normal saline nose drops before feeding can clear the nose temporarily to improve feeding. Choanal atresia, where the back of the nose is not open to the pharynx, is suspected when there is severe nasal obstruction with inability to pass a soft suction catheter through the nose. It is a rare but important cause of nasal obstruction. Bilateral choanal atresia is a severe problem presenting shortly after birth, although unilateral obstruction may present later.

Stridor can be due to congenital laryngotracheomalacia, which is expected to improve spontaneously with growth. However if the stridor is associated with feeding difficulty, poor weight gain, or significant increased work of the breathing then the infant should be referred for specialist paediatric and otorhinolaryngology assessment.

Normal neonatal respiratory patterns are quite variable. Infants can cycle from short periods of slow deep breathing through periods of more rapid very shallow breathing or even short pauses of a few seconds. This is normal if there are no associated behaviour or colour changes and the pause is less than 10 seconds in duration.

Examination of respiratory effort and symmetry of chest wall movement is important. In addition to lower respiratory infections, congenital problems such as lung malformations and congenital heart disease may present at this age with tachypnoea and poor feeding.

Examination of the chest wall may reveal benign breast tissue enlargement as a response to maternal hormones. These occur in both infant boys and girls and occasionally a few drops of milk may be produced. No treatment is required, but you should distinguish this from a breast abscess–usually unilateral, inflamed, and painful.

Auscultation of the heart can be difficult with a resting heart rate of 140 to 160 beats/minute and an infant who may be crying and difficult to settle. Heart murmurs are common, with the most common being innocent or systolic flow murmurs. Any heart murmur should be evaluated by an experienced clinician regarding whether further investigation or referral is necessary. Further clinical examination of the cardiovascular system includes palpation of femoral pulses and of the size of the liver which is usually enlarged in heart failure. Any infant with a heart murmur should have four limb blood pressures recorded to screen

for coarctation of the aorta. Pulse oximetry should be performed in any infant where there is a concern regarding a cardiac or respiratory problem.

Examination of the abdomen should follow a standard approach of inspection and palpation of the major organs. Inguinal hernias are common and usually easily reducible. If obstructed they require urgent surgical consultation. If reducible they are usually planned for elective repair within the shortest time frame practicable. Umbilical hernias are also common and rarely require intervention. If the base of the hernia is less than 5 cm it can be expected to close spontaneously with time. In boys ensure that the testes are descended in the scrotum, which will require gently compressing the inguinal canal to prevent the cremasteric reflex from withdrawing the testes out of range of detection. A hydrocele is a simple fluid collection in the scrotum and, provided that there is no associated hernia, does not require surgery in the first months of life; they usually resolve without treatment.

The umbilical stump normally dries and separates naturally without any special care after a week or two. Occasionally the umbilical stump can become wet and smelly, but this rarely requires anything more than gentle washing and careful drying. Rarely a small granuloma may form after the cord has separated if there has been inflammation at the site. These can be left alone but sometimes require silver nitrate stick cautery if continuing to be troublesome after several weeks.

Examination of the hips for congenital dysplasia is an essential component of examination at this age. The posture and skin creases should be examined for symmetry. In a normal neonate the hips should be able to be easily abducted to close to 180 degrees. Each hip is then examined for stability using Ortolani's manoeuvre, feeling for clicks and/or clunks.

Finally examine the infant's back for asymmetry, bony discontinuities, or sinuses in the lumbosacral region. These are easily missed if the infant is never turned over during the examination.

Examining the skin will occur throughout the general examination of all areas. There are multiple newborn and neonatal rashes, both benign and more significant (see Chapter 3.3, Neonatal dermatology).

Common reasons for healthy neonates to present to the emergency department

Feeding problems

Breast-feeding is promoted strongly for good reasons; it is very much the best way to feed an infant. Unfortunately, breast-feeding doesn't always go smoothly for new mothers, and there can be considerable guilt in resorting to bottle-feeding. Infants can present to hospital with poor weight gain, persistent jaundice and sleepiness when initial feeding is not going well. These problems are difficult to fully resolve in the ED, and access to either hospital or community-based lactation consultants can be very helpful. Tongue tie is often raised as a concern when infants are struggling with latching and sucking. Frenotomy, or surgical release of the tongue tie, is usually not warranted. To date, the five randomised controlled trials evaluating frenotomy have failed to show that these procedures consistently improve infant feeding, although they are likely to improve maternal nipple pain. Assessment of the contribution of the tongue to the feeding difficulties of a particular infant may be helpful provided that it is performed by an appropriately trained person (usually a paediatrician, otorhinolaryngologist or lactation consultant).

Crying (see **Chapter 3.2**, The crying infant)

Intractable crying is a common reason for neonatal ED presentations. All babies cry; some cry much more than others, and this can be very difficult for tired parents to cope with. The approach to crying involves taking a careful history of the duration and pattern of crying. A sudden change in the infant's behaviour may indicate a new painful condition such as a corneal abrasion, a hair tourniquet around a digit, or an obstructed hernia. More persistent long-term crying may not yield any obvious cause after careful history and examination. It is not especially helpful to label these babies as having 'colic' since this focuses on the belief that the infant is experiencing abdominal pain when we have no knowledge of that. It is more helpful to carefully explore what helps and what doesn't that the parents have tried. Baths, massage, slings and infant packs to carry the infant, swaddling and propping the head of the bassinet mattress up by 20 degrees may all help some infants and not others. Occasionally after the parents have checked that the infant is not wet, dirty, hungry or unwell then all that they can do is leave the infant to cry for periods of time. It can be helpful for them to be reassured that they are not being poor parents or harming their infant by doing this, and it is certainly better than them becoming so desperate that they

shake the infant. It is important to know what local support services are available to support parents.

Vomiting and spilling

Most infants spill milk to some degree after feeding. This is not usually a problem. If the infant is gaining weight well and happy then it is mostly a laundry problem. Spilling large quantities can be upsetting to the infant and require smaller volume feeds more frequently. Keeping the infant upright after feeding on the parents shoulder for 15 minutes may help. Winding or burping infants doesn’t make any difference to spilling. A number of medications are used to manage gastro-oesophageal reflux in infants; in most cases these are unnecessary. Gaviscon infant powder can be used as a thickening agent and antacid but may cause constipation. Omeprazole use should be restricted to more severe cases where oesophagitis is limiting feeding.

Preventative advice

The attendance of a normal neonate to the ED is an opportunity for preventative health advice for the family.

Smoke exposure is particularly harmful for young infants. Short focused smoking cessation advice has been shown to be effective, especially if backed up by providing contact numbers for support and other resources.

Sudden unexplained death in infancy is to some extent preventable through safe sleeping advice. All parents should be encouraged to place their infant to sleep lying on his or her back, on a firm mattress with only bedding in the cot. Co-sleeping is a hazard especially if there is parental obesity, smoking or alcohol use. There are strong cultural preferences for co-sleeping in some communities. If the parent choice is to have the infant in the same bed then the risk can be mitigated by providing the infant with his or her own safe space in a Moses basket, Pepi-pod or similar device.

Inflicted injury continues to occur with distressing frequency to infants in our communities. National education campaigns regarding the severe brain injuries caused by shaking infants in frustration and anger have been implemented in some countries. Reinforcement of this information may be helpful.

Timeliness of immunisation is an important issue in order to achieve maximal protection of young at-risk infants. Thus an ED visit is an opportunity to review

the immunisation history, discuss any concerns, and offer catch-up immunisations if the department has the systems to support this and link back to primary care.

Any ED consultation should conclude with advice regarding when to seek further assessment or treatment. For the normal neonate this should include advising parents to register their infant with a GP, if they haven't already done so, and to attend for their six week well child check and immunisations. The emergency clinician should copy their discharge information to the domiciliary midwife, well child provider, as well as the GP.

Summary

Parents frequently bring young infants to the ED for assessment. Many neonatal presenting problems are aspects of normal behaviour and development or transient problems which are expected to improve with age. Infants at this age may also present with previously undiagnosed congenital abnormalities or serious infections. It can be challenging to screen for serious conditions while avoiding unnecessary investigations and increasing anxiety. A high level of comfort and familiarity with normal neonatal behaviour and appearance are invaluable to the emergency clinician.

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3.2

The crying infant

Jeanette Marchant

ESSENTIALS

1 Define if this presentation is part of a recurrent stereotypical pattern in an otherwise well infant or a single acute episode.
2 A careful history and examination will often lead to an appropriate diagnosis.
3 Screening tests, with the exception of urine culture, have little utility.
4 Review carefully carer's coping and supports.
5 Organise appropriate follow-up.

Introduction

Crying is normal physiological behavior in young infants and an important method of communication.[1] Carers are usually able to identify and manage the cause (e.g. hunger, tiredness, discomfort) and console the infant. Infants cry more during the first 4 months of life than at any other time. Brazelton, in his 1962 study of 80 infants of American families with minimal psychological stresses, defined 'normal crying' as 1 hour and 45 minutes per day at age 2 weeks, a peak of 2 hours and 45 minutes per day by 6 weeks, decreasing to less than 1 hour per day at age 12 weeks.[1] Medical advice may be sought if the crying is felt to be unusually intense or persistent, or the infant is unable to be consoled by the usual methods.[2] The classification of crying as normal or excessive is highly subjective and will vary according to infant, carer and situational factors. All three areas need to be assessed in this type of

presentation. Either a single episode of crying, or recurrence of a pattern of excessive crying, may precipitate emergency department (ED) presentation.

Recurrent crying

Colic

Most infants with excessive crying do not have an underlying medical cause. Recurrent excessive crying in an otherwise healthy infant is often termed colic. Colic was originally described in 1954. It is a diagnosis of exclusion which can only be made if the pattern is recurrent and stereotypical and careful history, examination and a period of follow-up have ruled out other important causes.[3] The definition of colic varies but is frequently arbitrarily defined as a total of more than 3 hours per day of irritability, fussing and crying on at least 3 days a week for at least 3 weeks.[4] The infant is otherwise healthy, active and thriving, with no other features suggestive of underlying disease. This pattern typically occurs in the afternoon or evening, ceasing by 3 to 4 months of age.[3] Crying may be associated with the infant going red in the face, flexing his/her legs or passing wind.[5] The aetiology of infantile colic is poorly understood and likely to be multifactorial.

Management

There is no definitive management of infantile colic with caregiver support being the foundation of management (see ‘General advice’ below). The self-limiting nature of the condition should be explained to caregivers. It is important to ascertain that caregivers have appropriate support and also to inquire if the mother of the infant has symptoms of postnatal depression (if so she may require follow-up for her own health with primary care). To date, randomised controlled trials of interventions for infantile colic have been disappointing. Studies suffer from methodological issues, and currently no universal recommendation regarding treatment can be made.[6] Mothers who are breast-feeding should be encouraged to continue to do so. Some breast-feeding mothers have found exclusion of dairy products from the diet for 2–4 weeks to be beneficial. Small randomised trials with methodological limitations of hydrolysed infant formula have shown some clinical improvement following cow’s milk protein elimination.[7,8] However, it is generally not appropriate to start such formulas in the ED without appropriate follow-up and plan for discontinuation. Follow-up

should be organised with well child providers and primary care within the next week, to monitor growth and confirm diagnosis. In cases of severe symptoms follow-up should be with GPs.

Gastro-oesophageal reflux

Gastro-oesophageal reflux (GOR) is the physiological passage of stomach contents back into the oesophagus due to the relaxation of the oesophageal sphincter. It is not usually a cause of crying in infants. Gastro-oesophageal reflux disease (GORD) is GOR associated with large volume regurgitation, feeding difficulties, poor weight gain and haematemesis. This may cause problematic crying. Initial treatment should be conservative management, including positioning, smaller more frequent feeds or thickened feeds. Medications including antacids, H_2-receptor antagonists and proton-pump inhibitors may have a role in infants with significant symptoms.

Anticolic medications should be avoided, as they have been shown to at best have no effect (simeticone) or risk serious adverse effects (anticholinergics).[4]

General advice

Behavioural interventions, including advice to reduce stimulation in combination with permission to leave the infant when the crying is no longer tolerable, are effective.[9] Reduction of stimulation advice includes avoiding excessive patting, winding, lifting, vigorous jiggling and loud noises or toys. Carers were advised not to intervene in the early part of sleep when the infant may appear restless and also given an assurance that a certain amount of crying is normal.[4] It is important to remember that even if behavioural interventions do not change the infant's temperament, they may well alter the impact colic has on the carer and on carer–infant interactions.[10]

Carers very reasonably assume that there must be something wrong either with the child or their parenting for an infant to cry frequently and excessively. Much reassurance that the child is healthy can be gained from witnessing the conduct of a complete history and thorough examination. Similarly, an explanation that this is a common problem that does not reflect on their parenting, that some babies may be assisted by some simple behavioural techniques and that the carers will be supported by appropriate referral will also reduce anxiety considerably.

Acute crying

The causes of a single episode of excessive crying in an infant are vast. In an afebrile infant without a cause apparent to the carer, a careful history has been shown to provide clues to the final diagnosis in 20% of cases. Physical examination was revealing in more than 50%, and a period of follow-up was often useful in patients where the diagnosis was still in question.[2]

Assessment

History includes the timing and amount of crying, duration of behaviour, measures taken to resolve the situation, specific carer concerns, the carer's response to crying, expectations and experience, specific social difficulties (including substance abuse), contact with child health nurse or other medical supports. A thoughtful review of the carer's supports and coping is essential.[9]

Also important are details of pregnancy, labour and neonatal difficulties, feeding activities including volumes of feed (considerations include under- and overfeeding), dietary changes, past medical history, vomiting (GOR, gastroenteritis, sepsis, meningitis). Consider changes in stools (constipation, gastroenteritis, bleeding anal fissure, intussusception), type of feeding (breast or bottle), type of milk, drug exposure, recent immunisation, fever, respiratory symptoms, rash, contacts with infectious illness and growth history.

Examination

A complete set of vital signs is essential, including oxygen saturation (tachypnoea with pneumonia, sepsis or metabolic acidosis, tachycardia with supraventricular tachycardia, sepsis or dehydration, desaturation with pneumonia or bronchiolitis). Examination should include tone and activity, alertness/conscious state (meningitis, encephalitis, sepsis, metabolic crisis, hypoglycaemia, electrolyte disturbance), perfusion, hydration, fontanelle (bulging with infection or trauma, sunken in poor feeding and dehydration), chest for respiratory distress (pneumonia, bronchiolitis, metabolic acidosis, cardiac failure) abdomen (herniae, testicular torsion, evidence of surgical abdomen), cardiovascular system (murmurs, femoral pulses, cardiac ischaemia due to aberrant coronary vessels is a reported but extremely rare cause), ears (otitis media) and oropharynx (herpes stomatitis, tonsillitis, upper respiratory tract infection).

Several additional manoeuvres may also assist:[2]

- Fluorescein staining of cornea (corneal abrasion)
- Palpation of long bones (fractures, osteomyelitis)
- Inspection of skin beneath clothing (bruising)
- Retinal examination (shaken baby syndrome)
- Close attention to digits and genitalia (hair tourniquet).

Investigations

Investigations should be guided by initial clinical assessment. Screening investigations, with the exception of urine analysis, microscopy and culture, have little utility.[2]

Disposition

The diagnosis of serious medical conditions during the initial clinical assessment will clearly indicate admission. Admission should also be considered in those cases where the clinical assessment is normal but the child continues to cry excessively in the ED beyond the time of the initial assessment. Persistent crying in these circumstances may be an indicator of serious illness.[2]

Occasionally it may be necessary to admit an infant with colic or minor medical problem to allow recovery of a sleep-deprived or poorly supported carer.[10] The involvement of social work services is appropriate under circumstances where the family or child is considered at risk. Serious injury to children by non-accidental shaking injury is preceded by other episodes of abuse or neglect in over 70% of cases.[11] Admission and social work assessment are always warranted if non-accidental injury or neglect is suspected.

Ensure timely and appropriate follow-up is organised for infants discharged from the ED – particularly where the diagnosis is not yet clear or where excessive crying is likely to be recurrent.

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3.3

Neonatal Dermatology

Diana Purvis, Roderic Phillips, David Orchard, and Mike Starr

Skin in the Neonatal Period

The skin has many important functions. It provides a physical, chemical and immunological barrier and has a major role in thermoregulation, electrolyte balance, metabolism, sensation and physical appearance. Defects or changes in the skin can impact on these functions.

At birth, the barrier function of the skin is reduced, particularly in premature infants. This makes newborns vulnerable to increased transepidermal water loss and increased absorption of topically applied agents. This barrier function improves rapidly after birth and even in premature infants, will have matured by 3–4 weeks of age. Neonates are also susceptible to percutaneous toxicity due to the increased ratio of surface area to volume and the frequent presence of occlusive agents (e.g. nappies). Skin-barrier function can be further reduced by the presence of inflammatory dermatoses such as eczema. Caution is needed with use of any topical product in neonates due to the risk of absorption, including topical antiseptics (iodine, chlorhexidine, alcohol) and corticosteroids. It is recommended that agents such as neomycin, boric acid, urea and salicylic acid are avoided in premature infants and neonates.

Table 3.3.1

Differential diagnosis of neonatal dermatological presentations

Red and scaly rashes

- Eczema/seborrhoeic dermatitis
- Psoriasis
- Neonatal lupus erythematosus
- Syphilis
- Ichthyosis – autosomal recessive congenital ichthyosis (ARCI), Netherton, epidermolytic ichthyosis

- Immunodeficiency – severe combined immunodeficiency, Omenn, immunodysregulation polyendocrinopathy enteropathy X-linked (IPEX) syndrome
- Langerhans cell histiocytosis
- Metabolic disorders

Pustular

- Infections – *Staphylococcus aureus*, group A streptococcus, *Candida*
- Benign inflammatory causes – erythema toxicum, neonatal cephalic pustulosis, transient neonatal pustular melanosis, acropustulosis of infancy, eosinophilic pustular folliculitis, milia

Vesicles and bullae

- Infections – neonatal varicella, herpes simplex, *S. aureus* (impetigo, pustules, staphylococcal scalded skin syndrome)
- Genetic – epidermolysis bullosa, incontinentia pigmenti
- Inflammatory – miliaria, Langerhans cell histiocytosis, mastocytosis, autoimmune blistering disorders
- Arthropod - scabies, bed bugs, fleas, other biting insects
- Traumatic – burns and scalds

Birthmarks

- Epidermal/sebaceous naevi
- Incontinentia pigmenti/focal dermal hypoplasia (Goltz syndrome)
- Congenital melanocytic naevi (CMN)
- Pigmentary mosaicism
- Cutis aplasia

Vascular lesions

- Transient – cutis marmorata, harlequin colour change
- Macular – salmon patch, capillary malformation, early haemangioma
- Palpable – haemangioma of infancy, other vascular tumours, vascular malformations

Brown/black

- Mongolian spots
- Pigmentary mosaicism
- Incontinentia pigmenti
- Epidermal naevi
- Café au lait macules (CALM)
- Mastocytosis
- Congenital melanocytic naevi, epidermal naevi

Blue/purple lesions

- Mongolian spots
- Congenital infection – rubella, toxoplasmosis, cytomegalovirus, herpes simplex, etc.
- Tumours – congenital leukemia cutis, neuroblastoma, Langerhans cell histiocytosis
- Purpura – bruising (trauma, bleeding disorder, e.g. haemolytic disease of the newborn), sepsis, vasculitis
- Vascular tumours/malformations
- Subcutaneous fat necrosis of the newborn

Skin care of neonates should in general be kept simple and minimise exposure to unnecessary products (e.g. no soaps, bubble baths, perfumes, herbal products or fragrances). It is usually recommended that infants are bathed every few days using either plain water or a gentle soap substitute. Baby wipes, although practical, can be a cause of napkin dermatitis, and in general plain water and a soft cloth is preferable. Barrier creams such as petroleum jelly or zinc and castor oil can be used to prevent irritant napkin dermatitis.

Neonatal erythroderma (Fig. 3.3.1)

Erythroderma in the neonatal period (defined as erythema affecting >90% of the body surface area) has many causes, some of which may be life threatening. Admission is required for investigation, diagnosis and management. Erythrodermic infants are at risk of temperature instability, fluid loss and infection due to abnormal skin barrier function.

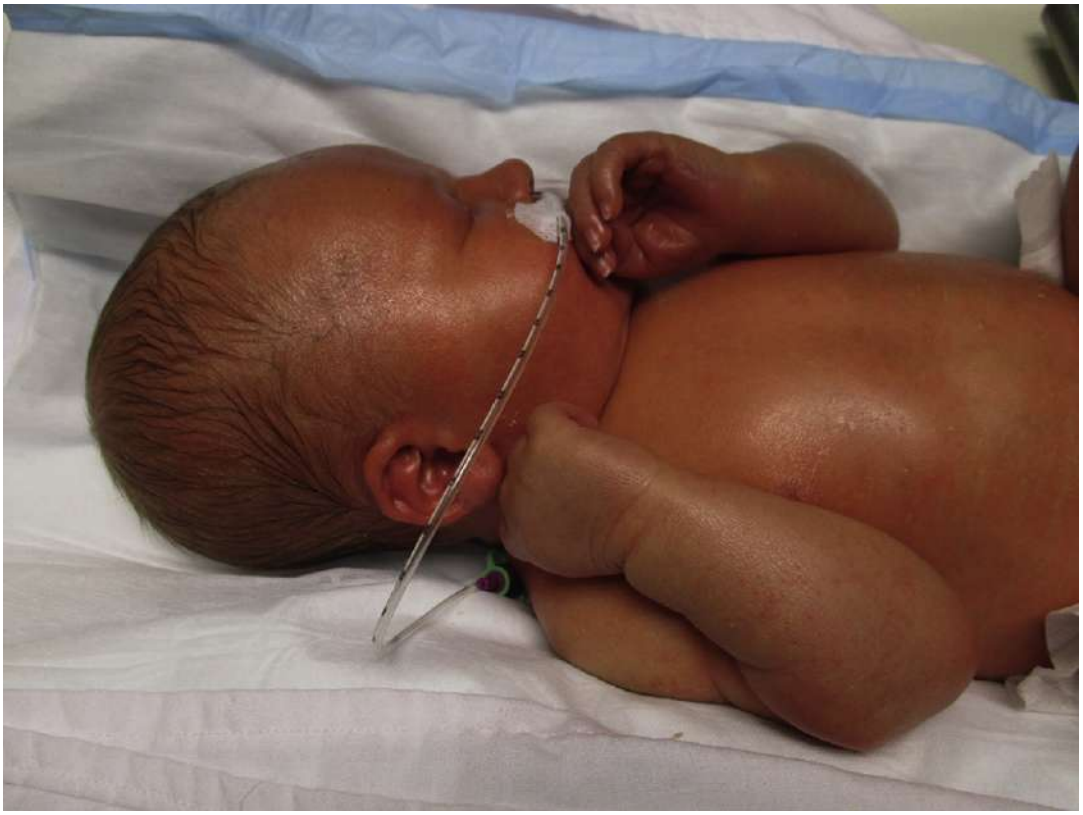

FIG. 3.3.1 Neonatal erythroderma.

Potential causes of erythroderma

- Inflammatory dermatoses:
    - Psoriasis, eczema, pityriasis rubra pilaris, diffuse cutaneous mastocytosis.
- Ichthyoses
- Immunodeficiency:
    - Severe combined immunodeficiency, Omenn syndrome,

immunodysregulation polyendocrinopathy enteropathy X-linked (IPEX) syndrome, DiGeorge syndrome, hyper IgE syndrome, graft versus host disease

- Drug reactions
- Metabolic diseases:
    - Acrodermatitis enteropathica, organic acidemias.
- Infections:
    - Staphylococcal scalded skin syndrome, staphylococcal toxic shock syndrome, congenital cutaneous candidiasis, syphilis, perinatal herpes simplex.

Investigations in erythroderma

Investigations should be determined by history and examination findings and are best determined in conjunction with inpatient management teams to minimise unnecessary tests and duplications but may include:

- blood count, renal and liver function, calcium, albumin, zinc
- arterial blood gas
- immunoglobulins and lymphocyte subsets
- plasma amino and urine amino and organic acids
- skin swabs for bacterial, fungal, viral studies
- skin biopsy
- hair microscopy
- genetic studies.

Management of the erythrodermic neonate

These infants should be admitted for investigation and management:

- Fluids and nutrition – close monitoring of hydration, electrolytes and nutrition is important. Nasogastric feeds or intravenous fluid may be needed in some infants.
- Thermoregulation – heat loss through inflamed skin may result in hypothermia. Keep the infant wrapped when possible or under heat lamps.
- Prevent secondary infection – erythrodermic infants are at risk of

staphylococcal, streptococcal or Gram-negative infections. Fever may be absent, even if infection is present. Good infection control measures are imperative. Have a low threshold for initiating antibiotic treatment. Where there is concern about possible severe immunodeficiency, protective isolation and positive pressure rooms should be used.

- Support skin-barrier function – application of a bland petrolatum-based emollient (e.g. 50:50 liquid:white soft paraffin) every 3 to 4 hours and regular bathing in plain water or oily baths are recommended. Caution should be taken with application of products to the skin due to the increased risk of percutaneous absorption or skin irritation.

Red scaly rashes

Infantile seborrheic dermatitis/eczema

Eczema usually presents later in infancy, although it can present in the neonatal period. This presentation has been described as infantile seborrheic dermatitis in the past, but international opinion is divided as to whether seborrheic dermatitis is a subset of eczema or a separate entity. It may commence on the scalp and face as confluent erythema and vesicles with adherent yellow-brown scale or within the nappy area where it results in moist erythema in the inguinal folds. The scale on the scalp can become quite thick and adherent, so-called cradle cap. In the flexures of the neck, axilla, antecubital and popliteal fossae there may be a confluent moist erythema. On the trunk there may be oval erythematous scaly lesions which may coalesce.

Treat with soap-free emollient washes and regular application of moisturiser. Topical antifungals (e.g. miconazole), and 1% hydrocortisone, can be effective if needed. Scale can be lifted from the scalp by massaging with a non-irritating oil or moisturiser and use of 2% ketoconazole shampoo. No feeding changes are needed.

Any neonate presenting with significant and widespread eczema should be assessed for conditions including ichthyoses, Netherton syndrome and other causes of erythroderma (see Chapter 12.1, Dermatology).

Psoriasis

Psoriasis is uncommon in the neonatal period and usually presents as anogenital erythema. Pustular psoriasis can cause sheets of exfoliative erythema with sterile

pustules (see Chapter 12.1, Dermatology).

Ichthyosis

Neonates with more severe forms of ichthyosis may present to emergency departments (EDs) with complications of dehydration (due to increased transepidermal water loss), infection or failure to thrive (see Chapter 12.1, Dermatology).

Neonatal lupus erythematosus (Fig. 3.3.2)

Neonatal lupus erythematosus occurs in infants whose mothers are positive for anti-Ro(SSA) and/or anti-La(SSB) antibodies. Most of these mothers are asymptomatic and unaware of their antibody status. About 5% of infants of antibody-positive mothers develop neonatal lupus.

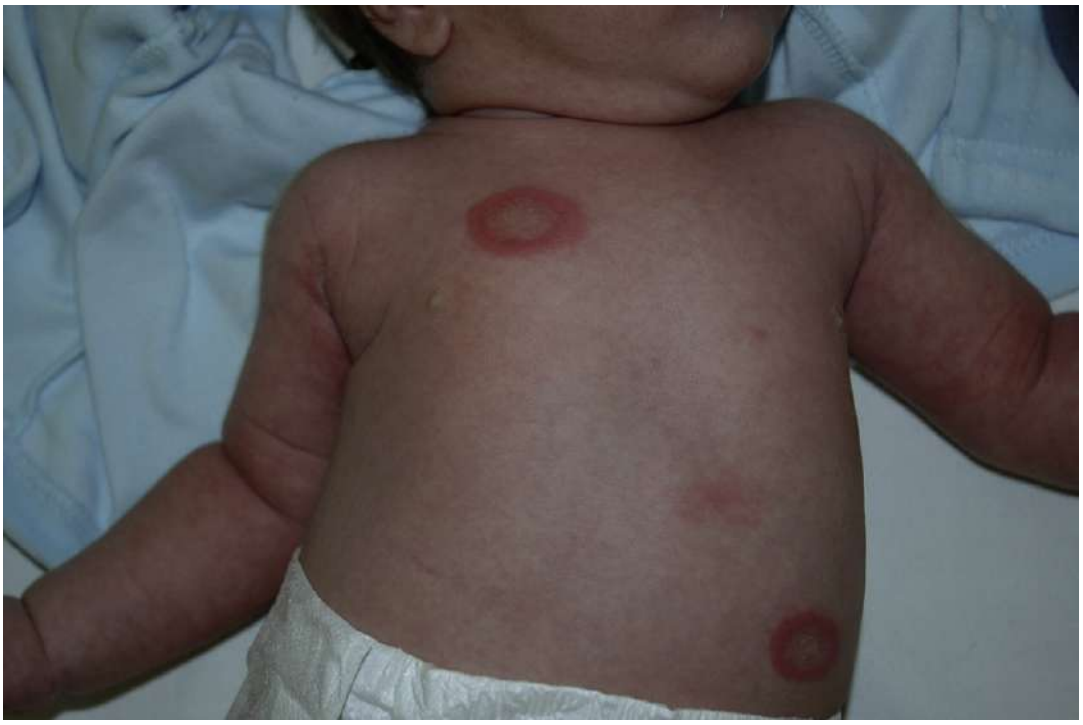

FIG. 3.3.2 Neonatal lupus. Erythematous annular lesions.

Skin lesions in neonatal lupus can first appear from a few days to a few months after birth. Widespread, erythematous, often annular and growing lesions develop on the face, scalp, trunk, extremities and neck. These can be

misdiagnosed as tinea or eczema. They persist for months before clearing as the maternal antibodies clear. Moderate potency topical steroid assists clearing. Sunlight and ultraviolet light can precipitate or worsen the rash.

The other major manifestation of neonatal lupus is complete heart block (from the third trimester). This is permanent and can be fatal; a pacemaker is required. About half of all babies with neonatal lupus have complete heart block, and about half have the characteristic skin lesions, but only a small proportion have both.

All infants with neonatal lupus presenting with skin lesions require ongoing follow-up as they appear to have an increased risk of developing autoimmune thyroid or rheumatological disorders during childhood. Approximately 25% of the mother's subsequent pregnancies develop neonatal lupus and so antenatal care should include cardiac monitoring of the fetus. Any older siblings should be assessed to exclude heart block. Most mothers eventually develop rheumatological disease and also require referral for follow-up.

Zinc deficiency/acrodermatitis enteropathica

Zinc deficiency, either inherited or acquired, can cause a progressive red scaly and crusted rash on the face, distal limbs and perianal region. Onset is usually later in infancy, although it can rarely be in the neonatal period (see Chapter 12.1, Dermatology).

Langerhans cell histiocytosis (see Chapter 12.1, Dermatology)

Tinea corporis

This is extremely uncommon in the neonatal period. Red annular rashes in neonates are more likely to be eczema, urticaria or neonatal lupus.

Congenital syphilis

Congenital syphilis is due to the transmission of *Treponema pallidum* from an infected pregnant woman to her fetus. The severity of disease is variable – as many as 90% of affected newborns have no signs at birth. Clinical findings include hepatomegaly, jaundice, nasal discharge, lymphadenopathy, pseudoparalysis and ophthalmological/central nervous system (CNS) and

skeletal changes. The rash usually appears 1–2 weeks after the rhinitis and consists of red-pink spots most prominently on the back, buttocks, palms and soles. It progressively desquamates and fades to a dull red/copper colour. Occasionally, congenital syphilis can present with bullae, periorificial fissuring or condylomata lata (moist perineal papules).

The diagnosis can be made by assessment of maternal and infant serology, darkfield examination of infected body fluids or histology of the placenta/umbilical cord or infected tissue with immunofluorescence. X-rays may show evidence of bony changes in longstanding disease.

Referral to a paediatric infectious disease specialist for management is appropriate. Child-protection issues need to be considered if acquired postnatally.

Vesicles and blisters

A neonate with vesicles (Fig. 3.3.3) or blisters requires urgent assessment, as potential causes include serious infections.

Collect epithelial cells from the base of a vesicle for viral culture and polymerase chain reaction assay (PCR). Care needs to be taken to collect sufficient cells for analysis.

Herpes simplex

Neonates who acquire herpes simplex virus (HSV) at birth usually present at a few days of age with grouped vesicles, often on the scalp. Lesions may rapidly spread and coalesce. Herpes infection may be associated with fever, pneumonitis, transaminitis and conjunctivitis. Approximately 30% of neonates with apparently localised skin disease also have CNS involvement.

In addition to viral testing of the vesicle as above, blood should be taken for blood count and liver function, and blood and cerebrospinal fluid (CSF) HSV PCR should be performed. Admission and empiric treatment with intravenous aciclovir are essential until HSV has been excluded.

Varicella

Although at least 90% of adults are immune to varicella, exposure during pregnancy is common. Fetal outcomes following maternal infection early in

pregnancy include:

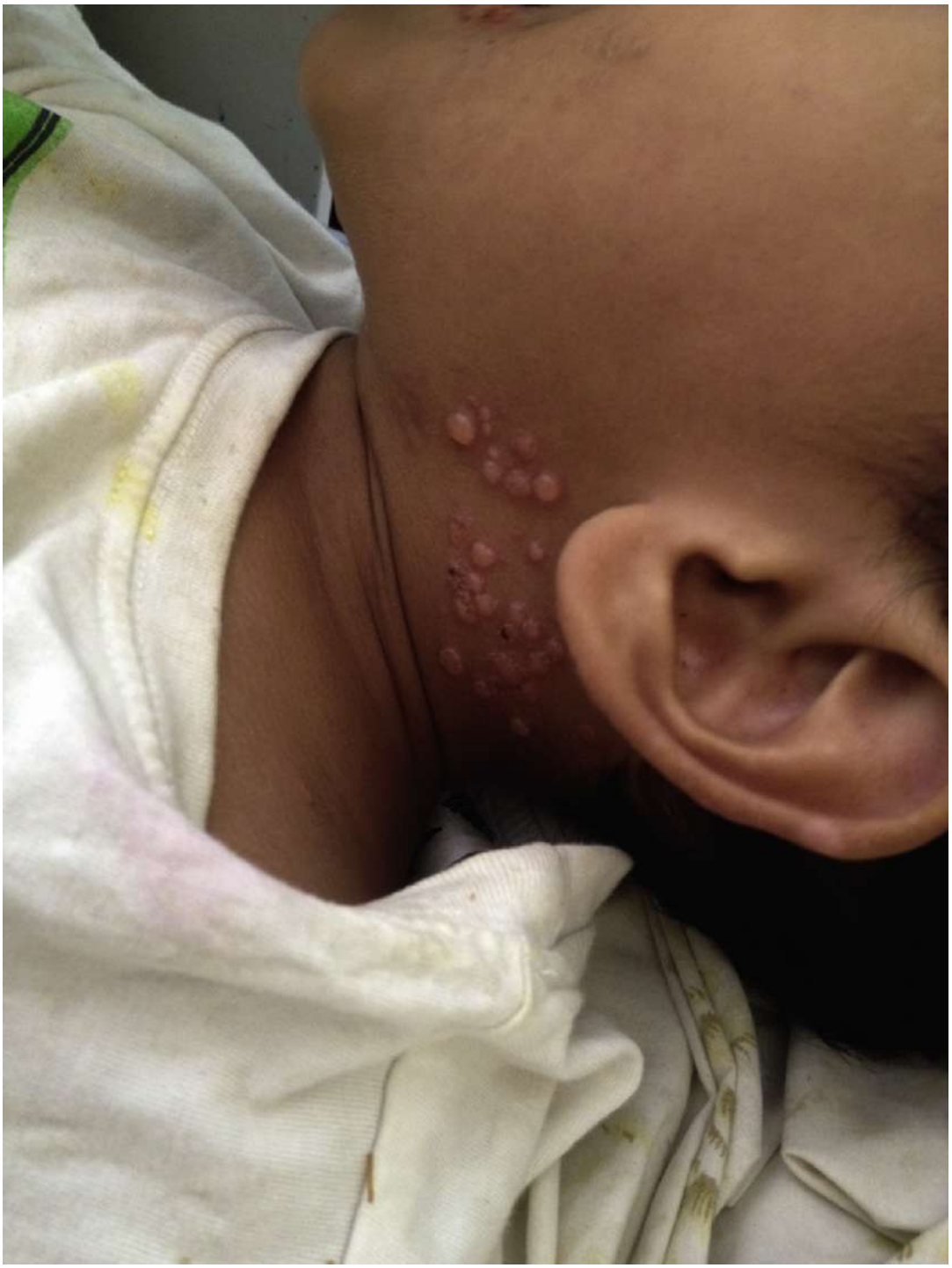

FIG. 3.3.3 Neonatal vesicles.

- uncomplicated self-limiting infection (10%)
- herpes zoster (shingles) in the first year of life (2–3%)
- fetal varicella syndrome (2–3% of cases), manifestations of which include growth restriction, skin scarring over a dermatomal distribution, ipsilateral limb or other skeletal hypoplasia, encephalopathy and abnormalities of various organs.

Varicella zoster virus (VZV) infection of the newborn results from transmission from a mother with chickenpox to her infant around the time of delivery, where the infant lacks the protection of maternal antibodies. The likelihood of infection depends on the timing of delivery in relation to when the mother develops the chickenpox rash. If the rash develops more than 7 days before delivery, this generally allows time for the development and transfer of protective maternal antibodies. However, as transfer of antibodies from the mother to the infant is limited before 26–28 weeks of gestation, maternal immunity to VZV does not usually protect preterm infants delivered before 28 weeks' gestational age.

The worst outcome is if maternal VZV infection occurs during the period from 5 days before delivery until 2 days afterwards, when infection of the infant may be complicated by pneumonia, hepatitis or encephalitis, with mortality up to 30%.

When maternal infection occurs more than 5 days before delivery, infection in the infant is usually mild. Infants exposed to varicella after the first few days of life also usually have mild disease, although this is variable and depends, amongst other factors, on the mother's immune status.

Diagnosis is usually clinical but can be confirmed by VZV DNA detection by PCR.

Preterm neonates with VZV infection or term neonates with severe disease should be isolated and treated with intravenous aciclovir.

Post-exposure prophylaxis with varicella zoster immune globulin (ZIG) should be offered to infants with significant varicella exposure where:

- maternal chickenpox occurs from 5 days before to 2 days after delivery.
- they are a hospitalised premature infant born <28 weeks, weighing <1000 g at birth, or whose mother is not immune.

If in doubt discuss with local paediatric infectious disease specialists.

Staphylococcus aureus

Staphylococcus aureus is inevitably acquired postnatally and often on the umbilical stump, and infection commonly presents between 5 and 10 days of life. Management depends on the clinical status of the neonate and the extent of the disease. Swabs should be taken before commencing therapy. Choice of antibiotic will depend on local guidelines:

- Localised disease – pustules or bullous impetigo:
  In a well full-term neonate, this may be treated with either topical or oral antibiotics for 7 days in an outpatient setting. Oral antibiotics should be used if lesions are large or multiple. In a preterm or low birth weight infant, or in an infant with fever or other features of systemic illness, blood cultures should be taken, and admission for intravenous antibiotics is recommended.
- Widespread disease or staphylococcal scalded skin syndrome:
  Admission for intravenous (IV) antibiotics is recommended.

Miliaria

Miliaria is caused by obstruction of the epidermal portion of the sweat duct. Miliaria crystallina results in small superficial clear vesicles often in the first few weeks of life and resolves with desquamation. No treatment is needed.

Miliaria rubra (prickly heat) occurs when the obstruction occurs more deeply in the epidermis and results in an irritating erythematous papular eruption typically in areas of occlusion, e.g. flexures or after lying on a waterproof sheet. Topical steroids may help with symptoms, but it takes some weeks to resolve.

Epidermolysis bullosa

Epidermolysis bullosa (EB) refers to a group of inherited blistering diseases which result in skin fragility. Subtypes range widely in severity with some also affecting internal organs and greatly reducing life expectancy. Presentation may be with non-inflamed blisters on the skin and oral mucosa, loss of fingernails or skin shearing with handling. Areas of absent skin (cutis aplasia) at birth, particularly on the lower legs, are also seen. Hoarseness, or vomiting, may represent involvement of the respiratory, or gastrointestinal tracts. Diagnosis

requires urgent expert dermatological assessment for skin biopsy and genetic testing.

Management in the emergency department

Patients with EB require careful handling to reduce skin damage from what would normally be minor friction. Specialised non-adhesive silicon dressings and tapes are recommended. In an emergency situation, normal adhesives may have to be used (e.g. monitoring leads, IV fixation). These can result in significant areas of skin loss when they are removed. If in any doubt leave these in place on the patient pending advice from an a specialist EB nurse:

- Ultrasound scans can be performed on patients with EB – use plenty of lubrication, or place a layer of cling film between the patient and the probe.
- Lacerations can be repaired using sutures or glue. Steristrips and tapes are not advised due to traction on the skin.
- Acute blistering of the conjunctivae can result in severe pain and photophobia. Antibiotic eye ointment can be applied, and urgent ophthalmology assessment is needed. Avoid trauma to eyelid skin.
- If the neonate is hoarse, there may be blistering in the airway. This can lead to airway obstruction and requires urgent assessment. Stridor and respiratory distress should be managed with adrenaline and dexamethasone acutely.
- Vomiting in a neonate with EB may be due to pyloric stenosis.
- Advice and specialist nursing support are available from the patient support group Debra (www.debra-international.org).

Autoimmune blistering disease

These are all rare in the neonatal period.

Pemphigoid gestationis causes an itchy blistering rash in mid to late pregnancy. Transplacental passage of antibodies can rarely result in disease in the neonate (see Chapter 12.1, Dermatology).

Other causes of vesicles and blisters (see Chapter 12.1, Dermatology)

- Burns and scalds
- Insect bites, bed bugs, scabies
- Severe acute contact dermatitis
- Vesicular phase of incontinentia pigmenti
- Mastocytosis
- Langerhans cell histiocytosis.

Pustular lesions

In the neonatal period, pustules may be part of several transient benign conditions. Pustules or vesicles may also be a marker of serious underlying illness, even in the absence of fever and lethargy. Any cause of neonatal vesicles can present with pustules as individual vesicles will appear pustular after 2–3 days. Also consider the following.

Infection

Either congenital or acquired, e.g. *Staphylococcus* species, group B streptococci, *Haemophilus influenzae*, *Candida* species.

Neutropenia

Superficial pustules may be the only sign of congenital neutropenia.

Neonatal pityrosporum folliculitis (Neonatal cephalic pustulosis) (Fig. 3.3.4)

This common eruption is often called ‘milk spots’ or ‘neonatal acne’; however, it is not true acne, and no comedones are present. It is a papulopustular follicular eruption on the face and torso appearing at 2–3 weeks of age, which is thought to be caused by an inflammatory reaction to a yeast known as *Malassezia*. Application of a topical azole (e.g. 2% ketoconazole cream) with or without 1% hydrocortisone may speed clearance.

Erythema toxicum neonatorum (toxic erythema of the newborn)

Despite the name, this is a common benign transient dermatosis seen during the first week of life. There is a migratory blotchy macular erythema, associated with papules or small sterile pustules filled with eosinophils. Neonates remain well, and the rash resolves within a few days. Microscopy of pustule contents can be helpful if there is doubt regarding diagnosis.

Transient neonatal pustular dermatosis

Loose pustules are present from birth and disappear within 2 days. This condition is more common in dark-skinned babies. In contrast to infective dermatoses, there is minimal erythema associated with the pustules, and they spontaneously heal with minimal scale.

Acropustulosis of infancy

Itchy vesicles and pustules appear on the hands and feet, usually commencing in the first couple of months of life and continuing for some months. New crops appear every 2–4 weeks. Sleep disturbance is common. The cause is unknown. In some infants it appears to follow previously treated scabies infection. Pustules contain neutrophils and are sterile. Multiple scrapings may be necessary to exclude active scabies infection. Potent topical steroids assist control of flares, and the condition remits over 1–2 years.

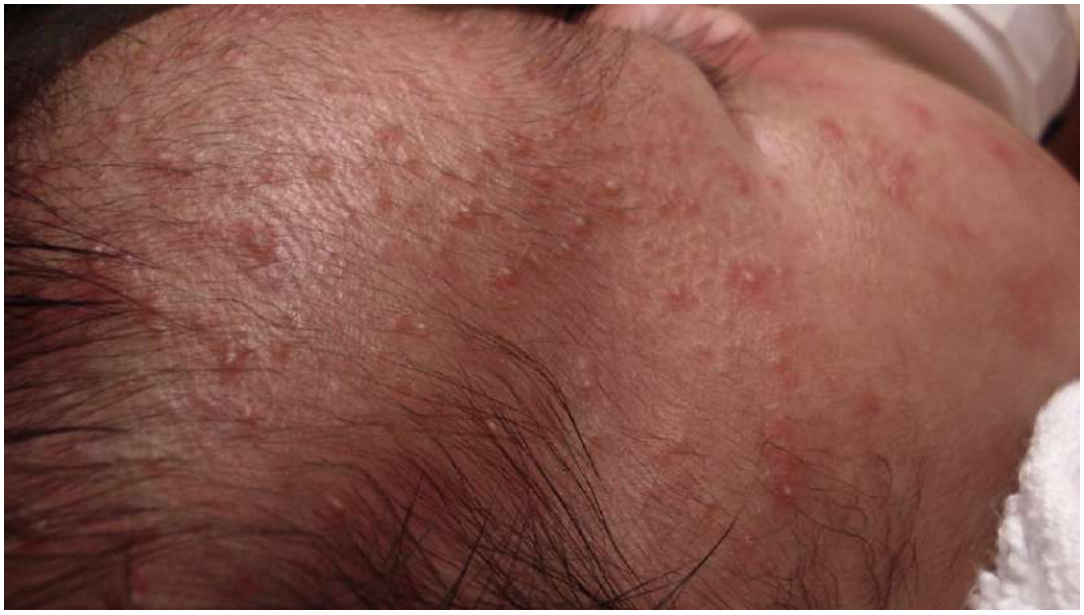

FIG. 3.3.4 Neonatal cephalic pustulosis.

Eosinophilic pustular folliculitis (scalp)

Transient sterile eosinophil-filled pustules appear over the face and scalp in early life and can be recurrent. The cause is unknown. Topical steroids can be used for itch.

Milia and sebaceous gland hyperplasia

Milia are small white pearly follicular epidermal cysts seen over the nose and forehead, often associated with sebaceous hyperplasia which causes multiple small yellow papules. Both resolve within weeks.

Birthmarks

Despite their name, some birthmarks are not evident at birth or if evident do not result in problems or complications until after the neonatal period (see Chapter 12.1, Dermatology). Birthmarks are due to abnormal proliferation of normal cells in the skin. Many are due to somatic or post-zygotic mutation, and the extent of the lesions will depend on the timing of that event. Lesions involving epidermal structures frequently follow Blaschko's lines, the developmental lines of the skin, resulting in a linear or whorled pattern and a clear midline demarcation.

Congenital melanocytic naevi

Congenital melanocytic naevi (CMN) are thickened, sharply defined, tan, dark brown or black birthmarks. The sizes below are as measured on the neonate.

Small (<0.5 cm) and medium (0.5–7 cm) congenital melanocytic naevi

Issues relate mainly to the appearance. Unlike larger lesions, there is no clear association with increased risk of melanoma. Surgical removal can be considered if warranted, but laser treatment is not generally recommended.

Large (7–15 cm) and giant (>15 cm) congenital melanocytic naevi

Lesions cover large segments of the body (e.g. bathing trunk naevus) and often have irregular colour, hair growth and proliferative nodules. Satellite lesions refer to smaller secondary lesions, which may continue to appear after birth.

Melanoma occurs in about 1% of patients with large and giant CMNs, often during childhood and in extra-cutaneous sites. Clinical and histological diagnosis can be challenging, and children should be under regular dermatologist review.

Neuromelanosis refers to proliferation of melanocytes within the CNS. This can be asymptomatic or associated with developmental delay, hydrocephalus and seizures. Neuromelanosis occurs in up to 25% of neonates with high-risk CMN (>15 cm, post-axial location, multiple satellites) and is symptomatic in 3–10%. Paediatric neurologist follow-up is recommended for those with evidence of neuromelanosis.

Cosmetic issues can result in significant psychosocial impact. Dermabrasion, serial excisions, tissue expansion with excision and laser are all at times used to treat large to giant CMN, and referral to an appropriate paediatric plastic surgeon with experience in this field is worthwhile.

Epidermal naevi

Epidermal naevi are due to post-zygotic mutations resulting in proliferation of keratinocytes and other epidermal structures following the lines of Blaschko. Most (though not all) are present at birth. The vast majority are small localised lesions with no medical complications. However, more extensive lesions can be associated with neurological and ophthalmological abnormalities.

Excision or laser may be useful to improve the appearance.

Inflammatory verrucous epidermal naevus

These are a rare variant and develop during childhood (often not noted at birth). Inflammatory verrucous epidermal naevus (ILVEN) are intensely pruritic hyperkeratotic papules and plaques following the lines of Blaschko. The itch may necessitate surgical removal for management.

Sebaceous naevus

Sebaceous naevi present as a yellow/pink velvety area on the scalp, head or neck

without hair growth. These often become thickened and verrucous during puberty, and there is a tiny risk of developing secondary basal cell carcinoma in later life and so non-urgent removal is often undertaken during childhood/teenage years.

Incontinentia pigmenti

This is an X-linked dominant disorder (usually lethal in males) due to defects in the NEMO gene resulting in apoptosis of affected cells. It has four phases:

- Vesicular – prenatal, neonatal or up to six months of age. Erythema and vesicles develop along the lines of Blaschko.
- Verrucous – from a few months to a few years of age. Warty lesions in the same distribution as blisters. Seen in one-third of cases.
- Hyperpigmented – 6 months to 16 years of age. Streaky tan-brown pigmentation.
- Atrophic – adults. Pale streaks with absent hair follicles. Often seen on the back of the legs.

Incontinentia pigmenti (IP) may be associated with ocular, neurological and dental abnormalities.

The mother and other close female relatives should be checked for signs of IP; many are only mildly affected and may be unaware that they are carriers.

Pigmentary mosaicism

This encompasses historical terms, hypomelanosis of Ito and linear and whorled hyperpigmentation. This is a sporadic disorder due to a mosaic pattern of pigmentation. The distribution of pigmentation may be along Blaschko's lines, phylloid or chequerboard.

Some children may have chromosomal mosaicism and associated neurological problems, but most are otherwise unaffected. Investigation and management should be based on clinical assessment.

Achromic naevus (also known as naevus depigmentosa) is probably a more localised variant. Lesions may not be evident at birth but become noticeable after sun exposure.

Café au lait macules

Café au lait macules (CALM) are flat areas of pigmentation slightly darker than the surrounding skin. They can be present at birth. As many as 30% of children have one or two CALM.

Children with more than six CALM (smooth bordered, associated with axillary freckling) are likely to have the diagnosis of neurofibromatosis type 1 and should be assessed for other clinical features.

McCune Albright syndrome typically has large CALM with irregular borders and is associated with precocious puberty and polyostotic fibrous dysplasia (see Chapter 12.1, Dermatology).

Cutis aplasia

Cutis aplasia refers to an area of congenital absence of the skin. Most commonly this is an isolated lesion affecting the vertex of the scalp. It heals with scarring but will leave an area of alopecia. Occasionally there can be associated bony defects (e.g. in Adams-Oliver syndrome; alopecia and limb malformations) or associations with other congenital anomalies.

The hair-collar sign refers to a ring of long dark hair around the area of cutis aplasia or other scalp lesions and can be a marker for an underlying cranial neural tube defect. Neuroimaging should be considered prior to any surgical procedures.

Blue/purple lesions

Blueberry muffin syndrome (Fig. 3.3.5)

This refers to multiple blue/purple cutaneous marks or nodules seen on a neonate. These can be due to extramedullary haematopoiesis (clusters of blood producing cells in the skin), bleeding or cutaneous metastases. Consideration needs to be given to the following diagnoses:

- Tumours:
    - Congenital leukaemia cutis
    - Langerhans cell histiocytosis
    - Neuroblastoma
    - Congenital rhabdomyosarcoma

- Disseminated juvenile xanthogranulomata
- Multiple vascular anomalies – haemangiomatosis, multiple venous or glomuvenous malformations.

- Blood disorders:
  - Haemolytic disease of the newborn
  - Rhesus or ABO incompatibility
  - Hereditary spherocytosis
  - Twin-to-twin transfusion syndrome.
- Congenital infections:
  - Rubella
  - Toxoplasmosis
  - Cytomegalovirus
  - Herpes simplex virus
  - Coxsackie virus, parvovirus, Epstein–Barr virus
  - Syphilis
  - Zika virus.

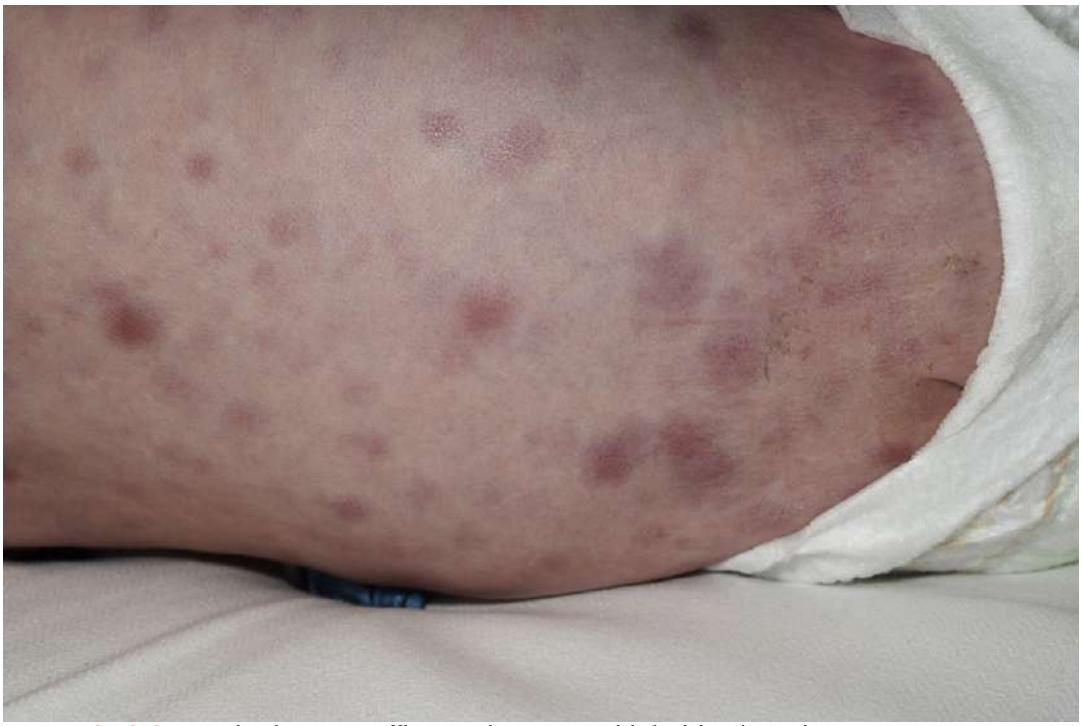

FIG. 3.3.5 Blueberry muffin syndrome. Multiple blue/purple cutaneous marks or nodules seen on a neonate.

Clinical presentation, including maternal history, and presence of growth

failure, jaundice, abnormal neurodevelopment or organomegaly may help to narrow down the diagnosis. Typically the infant is systemically well.

Investigations may include a full blood count and film, liver function tests, screening tests for congenital infection, skin biopsy, bone marrow biopsy, ultrasound of the abdomen, and ophthalmology examination. Urgent referral to general paediatrics/subspecialists to assess and determine the investigations is appropriate.

Other important causes

- Bruising from trauma – If this is thought to be present in a neonate, it should prompt consideration of bleeding disorders and non-accidental injury.
- Septicaemia causing purpura/petechiae – Assess for signs of infection.
- Mongolian spots – These are benign patches of dermal melanosis typically seen over the lower back and buttocks but occasionally also on the shoulders and lower legs. More common in Asian and darker-skinned infants. Typically they resolve during early childhood.
- Subcutaneous haemangiomas, vascular malformations (especially venous or lymphatic malformations), and cysts may present as bluish masses. Ultrasound scan of the lesion is often a useful initial examination.
- Cutaneous vasculitis is rare in the neonatal period – This would present as palpable purpura.
- Subcutaneous fat necrosis of the newborn – This is an inflammatory panniculitis seen in infants. It is thought to be related to hypoxia or hypothermia, but the precise causes are not understood. Infants present within the first 6 weeks of life with induration over the buttocks, thighs, shoulders or cheeks. The overlying skin may be bluish red in colour. Diagnosis is by skin biopsy. Infants with subcutaneous fat necrosis can develop marked hypercalcaemia at presentation or even a couple of months later. Hypercalcaemia may respond to treatment with diuretics, bisphosphonates or oral corticosteroids.

Vascular lesions in the neonatal period

Cutis marmorata

Physiological mottling or marbling of the skin resulting in a reticulate blue/purple vascular pattern is commonly seen in neonates in response to cooling. It typically improves or resolves on warming. This differentiates it from cutis marmorata telangiectatica congenita, a vascular anomaly causing a persistent reticulated vascular pattern associated with dermal atrophy and, at times, ulceration.

Harlequin colour change

This is a physiological vascular change which may be seen during the first few weeks of life. The infant develops a deep red colour on one half of the body and pallor on the other with a sharp line of demarcation along the midline. It tends to last for less than 30 minutes, and the infant remains well.

Salmon patch (naevus simplex, stork mark, angel's kiss)

This is an area of vascular ectasia typically seen symmetrically in the midline of the forehead, on the upper eyelids and nape of neck. It can also occur around the alar nasae and even on the limbs. These are usually pink in colour and become florid especially with crying. The facial lesions generally fade over the first 1–2 years of life (those on the nape of the neck may persist), and no treatment is needed. Darker and larger forehead lesions do not necessarily fade completely, and consideration should be given to referral to a paediatric laser service if still present after toddlerhood.

Vascular anomalies

This includes infantile haemangiomas, capillary malformations (port wine stains) and other vascular tumors and malformations (see Chapter 12.1, Dermatology).

3.4

Acute neonatal emergencies

Jeanette Marchant, and Christopher McKinlay

ESSENTIALS

1 Sepsis is the most common cause for the collapsed neonate in the emergency department (ED) in the first 4 weeks of life.

2 In the collapsed neonate consider bacterial sepsis first then look for signs of other rarer underlying disorders.

3 Left-sided obstructive heart lesions can present with shock, particularly at the time of ductal closure. Prostaglandin E1 maintains ductal patency.

4 Endocrine emergencies are rare but need consideration. Congenital adrenal hyperplasia is the commonest.

5 Persisting vomiting requires full evaluation, with bilious vomiting in a neonate being a surgical emergency until proven otherwise.

6 Neonatal seizures are relatively common and generally secondary to an underlying cause; thus they usually require full investigation. Neonatal seizures are often subtle, and a high index of suspicion is required.

7 Inspiratory stridor is most commonly due to laryngomalacia, but more serious pathology should be suspected if there is stridor from birth, biphasic stridor, abnormal cry, feeding difficulty, failure to thrive or history of choking or apnoea.

8 Jaundice is a common presentation to the ED. Extreme hyperbilirubinaemia is a medical emergency.

The Neonatal Period

The neonatal period is one of profound physiological change and, although cardiorespiratory transition is largely completed shortly after birth, organ function and homeostasis continue to mature over the first month of life.[1] The neonate is thus uniquely vulnerable and may deteriorate rapidly with illness and physiological stress. In addition to post-natal acquired disease, neonates are at risk of vertical infection and may present with a wide range of congenital, genetic and metabolic disorders. With early discharge of mothers from perinatal centres, presentation of neonates to emergency departments (EDs) in the first postnatal week is not uncommon. While this is mostly due to minor problems,[2] emergency physicians must be alert to a range of more serious conditions specific to the neonate. This chapter focuses on common medical and surgical emergencies in term neonates after the immediate birth transition.

Neonatal resuscitation (see Chapter 3.5)

Newborn life support focuses on the anatomic and physiologic adjustments needed to achieve the conversion from placental gas exchange to pulmonary respiration.[3] The two key steps in this transition are initiation of air breathing and change from the placental to the pulmonary circulation as the source of cardiac preload, both of which require lung aeration.[1] Thus guidelines for resuscitation of the newborn emphasise airway and breathing, with a compression to breath ratio of 3:1 for cardiopulmonary resuscitation.[3] However, evidence suggests that higher compression rates may be equally effective,[4,5] and once the neonate has been discharged from a perinatal centre, use of paediatric resuscitation guidelines is appropriate. Two important differences in paediatric guidelines are early assessment of cardiac rhythm and use of 15:2 compression ratio.[6] Nevertheless, attention to temperature control and blood glucose concentration remains important, and use of a T-piece device with oxygen blender in pulmonary resuscitation is recommended as this delivers controlled continuous end-expiratory positive pressure. The umbilical vein is useful for central access in the first few days after birth and often until the end of the first week.

Assessment of the neonate

Assessment of the unwell neonate should include review of the perinatal history, as this may contain clues as to possible diagnosis (Table 3.4.1). Although antenatal ultrasound detects many major congenital malformations there are several notable exceptions, such as transposition of the great arteries and coarctation. Feeding history is important, and it is useful to compare current weight with peak postnatal weight in assessing hydration. Clinical signs that should not be missed include cardiac murmurs, reduced or absent femoral pulses, imperforate anus and genital abnormalities, wide or full antenatal fontanelle and palate defects. Mild cyanosis is difficult to detect clinically and may only be detected by pulse oximetry. Radiographs should be screened for skeletal abnormalities, especially rib and vertebral anomalies.

Table 3.4.1

Perinatal history

	Salient features	Significance for the neonate
Maternal	Previous fetal or neonatal death	Genetic and metabolic disorders
	Diabetes	Malformations, hypoglycaemia
	Drugs, e.g. opioids, selective serotonin uptake inhibitors, amphetamines	Neonatal withdrawal syndrome, seizures, abnormal movements
	Major medical problems	Fetal effects
	Consanguinity	Recessive genetic disorders
Pregnancy	First trimester screen	Aneuploidy
	Mid-pregnancy anatomy scan	Intracranial, cardiac, thoracic, renal, gastrointestinal defects
	Fetal growth restriction	Chromosomal disorders, hypoglycaemia, thrombocytopenia
	Liquor volume	Polyhydramnios – upper gastrointestinal obstruction, aneuploidy Oligohydramnios – renal tract malformation, chromosomal disorders, fetal growth restriction, rupture of membranes
	Urinary tract infections, group B streptococcus on recto-vaginal swab, sexually transmitted diseases	Sepsis risk
	Maternal immunisation	Maternal immunisation during pregnancy reduces risk of influenza and pertussis infection
Birth	Late preterm and early term	Increased risk of early neonatal complications
	Resuscitation at birth	
	Instrumental delivery	Intracranial haemorrhage
	Vitamin K prophylaxis	
	Prolonged rupture of membranes (>18 hours), meconium stained liquor, maternal fever and leucocytosis	Sepsis risk

The collapsed neonate

The commonest cause of neonatal collapse is infection. There are, however, several less common non-infectious diagnoses that need to be considered because they are potentially life threatening yet treatable (Table 3.4.2).

Diagnoses such as duct-dependent congenital heart disease and inborn errors of metabolism can be difficult to differentiate from infection at the time of initial presentation. All these conditions can present with non-specific symptoms, such as lethargy, poor feeding or breathing difficulties. On examination the neonate may appear grey, pale or cyanosed. He/she may be irritable or lethargic. He/she may have tachypnoea, tachycardia and poor perfusion. Hypotension is a very late sign. Early recognition, stabilisation and management of the critically ill neonate may be life-saving.

The structured approach to the resuscitation of the collapsed infant involves support of airway, breathing and circulation and is presented in Fig. 3.4.1.

Sepsis

Infections represent an important cause of morbidity and mortality in the first month of life. A neonate who presents critically ill should initially be presumed to have bacterial sepsis and empiric antibiotics commenced. There are several reasons why neonates are at increased risk of infection, including the immaturity of the neonatal immune system and a lack of immunisation.

The signs and symptoms of sepsis may be quite subtle, and the duration of illness is variable, with some infants presenting after being unwell for several

days and others deteriorating rapidly. Any one or combination of the symptoms, such as lethargy, irritability, respiratory distress, poor feeding, vomiting, diarrhoea or fever, may be a manifestation of sepsis. A fever in a neonate is defined as a temperature of ≥38°C. Fever is a very unreliable finding, as many septic neonates will be hypothermic. It is important in the history to ask about perinatal risk factors for infection and infectious contacts (see Table 3.4.1).

On examination, the infant with severe sepsis is often pale, grey or cyanotic. The skin is usually cool and mottled owing to poor perfusion. The infant may seem lethargic, obtunded or irritable. The vital signs including temperature, respiratory rate, oximetry, heart rate, capillary refill time and blood pressure should be obtained. Disseminated intravascular coagulopathy may develop in severe sepsis with petechiae or purpura and is usually associated with poor outcome. A complete physical examination is required to look for focal signs of infection. If meningitis is present, there may be a bulging or tense fontanelle with a high-pitched cry, although more commonly neonatal meningitis presents with non-specific findings. If the infant has a respiratory infection there may be focal chest findings. It is important to examine for joint swelling and tenderness and to examine the umbilical stump for signs of omphalitis.

Table 3.4.2

Common aetiology of collapsed neonate

System	Aetiology
Infective	Septicaemia
	Meningitis
	Urosepsis
	Pneumonia
	Osteomyelitis
Cardiac	Congenital heart disease
	Arrhythmias
	Myocarditis
	Cardiomyopathy
Endocrine/metabolic	Hypoglycaemia
	Congenital adrenal hyperplasia
	Inborn errors of metabolism
Neurological	Seizures
	Intracranial haemorrhage
	Raised intracranial pressure
Gastrointestinal	Small bowel atresia
	Malrotation and midgut volvulus
	Hirschsprung enterocolitis

The goal of treatment in the ED is to support and stabilise the infant and to cover with antibiotics for serious bacterial infection such as bacteraemia, urinary tract infection, meningitis and osteomyelitis. In the collapsed neonate immediate resuscitation with support of airway, breathing and circulation must be initiated. If the infant develops signs of shock, stabilisation includes fluid resuscitation

and commencement of inotropic support.

A full septic screen should be initiated in any neonate with a fever or with clinical suspicion of sepsis. This includes full blood count, blood culture, urinalysis and urine culture (catheterised or suprapubic specimen), lumbar puncture and chest X-ray. A venous blood gas is important as it may show evidence of a metabolic acidosis and may reveal hypoglycaemia (blood glucose concentration <2.6 mmol L^{-1}). Serum biomarkers such as C-reactive protein and procalcitonin can be measured to aid in the diagnosis of serious bacterial infections in neonates. A coagulation profile should be performed if disseminated intravascular coagulation is suspected. Lumbar puncture should be postponed if the infant is too unwell to tolerate the procedure. When cerebrospinal fluid is obtained, it should be sent for cell count, Gram stain and culture, and molecular testing for herpes simplex virus and enterovirus.

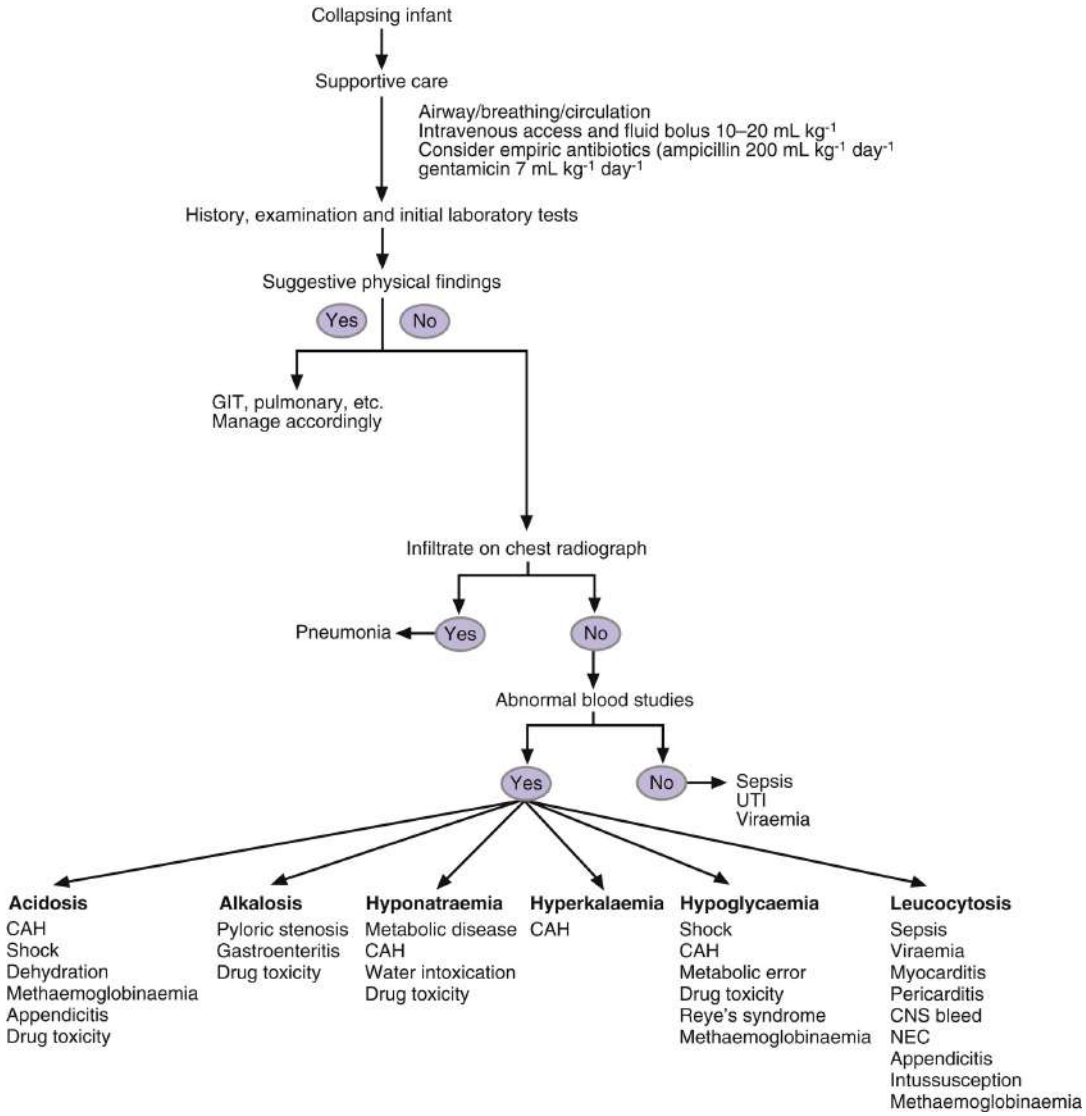

FIG. 3.4.1 Approach to the collapsing neonate.

Antibiotics

The goal is to commence antibiotics within the first hour of the recognition of the risk of sepsis and ideally after cultures are obtained. Intravenous antibiotics should cover both Gram-positive and Gram-negative bacteria. The most likely causative organisms in this age group are group B streptococci, *Escherichia coli* and *Listeria monocytogenes*. The choice of antibiotics depends on whether the source of the infection is suspected or known. Refer to state or local guidelines for prescribing (cefotaxime and amoxycillin are commonly used as broad-spectrum cover for unknown neonatal sepsis). Aciclovir should be commenced if infection with herpes simplex and varicella viruses is suspected. If intravenous

access is difficult, consider the intramuscular or intraosseous routes.

Viruses

Viral infections are more common than bacterial infections. Most are benign, but some may result in serious illness. Herpes simplex virus should be considered and, if suspected, treated. Neonatal herpes simplex virus is rare but carries significant risks of morbidity and mortality that can be reduced with antiviral therapy. Three distinct clinical presentations exist: skin, eye, and mouth infections; central nervous system infection; and disseminated infection. There may be some overlap with these presentations. Most mothers do not report a history of genital herpes.

Enterovirus infection in neonates usually presents as a sepsis-like illness. Respiratory distress is common, and haemorrhagic manifestations, including gastrointestinal bleeding or bleeding into the skin, may be seen. Seizures often occur as well as icterus, splenomegaly, congestive cardiac failure and abdominal distension. Mortality rates for enterovirus infections in neonates are quite high.

Bronchiolitis due to respiratory syncytial virus and other common respiratory viruses occur in winter, and infants may present with respiratory distress or apnoea. Those born prematurely, or with previous respiratory disorders, are especially susceptible to apnoea, and these infants may appear septic. Examination may reveal crackles and wheezes on auscultation. Nasopharyngeal aspirate for rapid viral testing may confirm the diagnosis. Chest X-ray may show diffuse patchy infiltrates and possibly lobar atelectasis or consolidation. Babies have a preference for nasal breathing,[8] and nasal secretions may cause significant upper airway obstruction and respiratory distress. Nasal suction may help considerably in overcoming this. Patients with severe respiratory distress may require respiratory support with humidified high flow nasal cannula oxygen therapy or continuous positive end-expiratory pressure. Infants with severe respiratory distress or apnoea may require intubation and ventilation.

Cardiac emergencies

Congenital heart disease (see Chapter 5.6) is one of the commonest congenital malformations, with an incidence of 0.6%. Only 30% to 60% of cases are diagnosed antenatally.[9] Cyanotic lesions generally present early, but neonates with ductal-dependent systemic circulations are often well in the early neonatal period and collapse around day 4 to 7 of life with closure of the ductus

arteriosus. Oxygen saturation screening of all newborn infants has been shown to increase the early identification of congenital heart disease.[10]

The commonest duct-dependent left-sided obstructive heart lesions are coarctation of the aorta, critical aortic stenosis, hypoplastic left heart syndrome and interrupted aortic arch. Presentation varies from mild heart failure with a history of poor feeding and poor growth to profound shock. Clinical signs may include tachypnoea, cardiac murmur, gallop rhythm, weak or absent femoral pulses or symmetrically reduced pulses, hepatomegaly and poor urine output.

Investigations to help confirm the diagnosis prior to echocardiogram include a blood gas looking for metabolic acidosis with an elevated lactate and chest X-ray to look for cardiomegaly (cardiothoracic ratio of >0.6) and plethoric lung fields.

The infant should be resuscitated with attention to airway, breathing and circulation. Early consultation with a paediatric cardiologist and intensivist should occur. The moribund and severely acidotic baby should be intubated and ventilated. This will reduce cardiac workload and help manage pulmonary oedema. Oxygen should be given to maintain saturations which are most reliably measured in the right upper limb. When concerned about cardiogenic shock, resuscitate with 10 mL kg^{-1} of crystalloid. Larger fluid boluses may exacerbate cardiac failure.

The mainstay of treatment in a neonate thought to have a duct-dependent lesion is an infusion of prostaglandin E1 (PGE1) to reopen and maintain patency of the ductus arteriosus. Side effects of PGE1 include apnoea, hypotension, seizures, and hyperpyrexia. Dosing should be in consultation with paediatric cardiologists and intensivists. Elective intubation and ventilation may be necessary for infants on infusion rates with a significant risk of apnoea.

Other therapies to consider after discussion with specialists include inotropes, sodium bicarbonate and diuretics. If the baby requires transfer to a paediatric cardiac facility, this will need involvement of a specialised retrieval team.

Sepsis still needs to be considered in any shocked neonate. Therefore antibiotics should be given, even if cardiac disease is thought the most likely diagnosis.

Arrhythmia (see Chapter 5.10)

An arrhythmia may cause an infant to present quite unwell. The most common dysrhythmia in an infant and child is supraventricular tachycardia (SVT). Infants with SVT often present with poor feeding, tachypnoea, and irritability which may go unrecognised at home for days. If untreated, the infant will develop

congestive heart failure and may present in a collapsed state, with a heart rate in excess of 300 beats per minute. The electrocardiogram (ECG), as in the older child, will show a narrow complex tachycardia with little rate variability and regular atrial and ventricular beats with 1:1 conduction, although p waves may be hidden. The chest X-ray may show cardiomegaly and pulmonary congestion. Stable SVT can often be managed initially with a simple vagal manoeuvre such as eliciting the diving reflex by dunking the infants face in a cold water or an ice bath. If this is ineffective, intravenous adenosine is recommended (initial starting dose is 100 micrograms kg^{-1}). Adenosine has a very short half-life. It should be administered through an intravenous cannula, ideally in the antecubital fossa attached to a three-way tap and followed by a flush. Adenosine works by blocking adenosine receptors in the atrioventricular node and slowing conduction. Unstable SVT is treated with synchronised cardioversion.

Endocrine disorders

Neonatal endocrine emergencies are rare. The commonest is congenital adrenal hyperplasia (CAH), which occurs in 1:16,000 to 1:20,000 births. CAH is an autosomal recessive condition resulting in the deficiency of cortisol and aldosterone, most commonly due to gene deletions or conversions of the enzyme 21α-hydroxylase. The history may reveal symptoms of poor feeding, vomiting, weight loss, lethargy, and irritability. Females may present with ambiguous genitalia since cortisol deficiency leads to adrenocorticotropic hormone hypersecretion, adrenal stimulation and androgen excess. Males may present with an enlarged penis and hyperpigmentation. Most cases are detected with newborn metabolic screening, but some babies present with a salt-losing crisis, usually during the first or second week of life. They are clinically shocked, with tachycardia, hypotension and hypothermia. Since these signs are also found in the septic infant, sepsis should always be considered in the differential diagnosis and antibiotics commenced.

Investigations should include a venous blood gas and formal electrolytes looking for hyponatraemia, hyperkalaemia, hypoglycaemia and acidosis. Ideally blood should be obtained for serum 17-hydroxyprogesterone, aldosterone, cortisol, and plasma renin prior to treatment. Management includes attention to airway, breathing and circulation. Fluid resuscitation is with 0.9% sodium chloride (10–20 mL kg^{-1}). Hypoglycaemia is treated with intravenous 10% glucose (2 mL kg^{-1}). Glucocorticoid deficiency is treated with intravenous

hydrocortisone (5 mg m^{-2} every 8 hours). If the infant has marked hyperkalaemia, specific treatment for this is required. Long-term follow-up will involve a paediatric endocrinologist.

The neonate with vomiting

Vomiting is a serious symptom in neonates that requires full assessment and exclusion of intestinal obstruction and major diseases outside the gastrointestinal tract, notably infection (meningitis, pyelonephritis, hepatitis), raised intracranial pressure (hydrocephalus, tumours) and metabolic disorders (Table 3.4.3). Vomiting or forceful emesis is distinguished from effortless regurgitation or gastro-oesophageal reflux. In the first week, vomiting is due to intestinal obstruction or malrotation until proven otherwise, whereas vomiting in the later neonatal period is less specific to obstruction. Newborns with infectious gastroenteritis typically present with diarrhoea rather than vomiting, and vomiting should not be attributed to gastroenteritis without further investigation. Bile-stained vomiting at any stage in the neonatal period is a surgical emergency and warrants urgent assessment.

Table 3.4.3

Causes of neonatal vomiting

Surgical	Upper intestinal obstruction: duodenal atresia, stenosis or web, jejunal atresia, hypertrophic pyloric stenosis Lower intestinal obstruction: ileal atresia, luminal obstruction (meconium ileus, meconium plug, milk inspissation), duplication cysts, Hirschsprung disease, anorectal malformation (imperforate anus) Malrotation and midgut volvulus Incarcerated inguinal hernia Necrotising enterocolitis†
Non-gastrointestinal	Infection: meningitis, viral hepatitis (CMV, HSV, enteroviruses, adenovirus, varicella),* pyelonephritis
	Raised intracranial pressure: hydrocephalus, brain tumour
	Endocrine: congenital adrenal hyperplasia, adrenal insufficiency, hypercalcaemia
	Chronic renal failure: bilateral renal dysplasia, severe obstructive uropathy or gross vesicoureteric reflux
	Inborn errors of metabolism: especially galactosaemia, organic acidurias, urea cycle defects, aminoacidopathies

† Usually confined to neonates born preterm or with cyanotic heart disease.

* Most common infections. HSV, herpes simplex virus; CMV, cytomegalovirus.

Relevant history includes the volume, frequency and content of the vomitus, including presence of blood or bile; stool pattern; feeding history; and presence of systemic symptoms, such as fever and lethargy. It is also important to enquire about the fetal anatomy scan (abdominal masses and cysts, genitourinary malformations), liquor volume (polyhydramnios in upper intestinal obstruction; oligohydramnios in obstructive uropathy), passage of meconium (normally <48 hours in term infants), completion of the newborn metabolic screen (Guthrie card) and perinatal risk factors for infection (see above). In addition to careful examination of the abdomen (distension, masses, organomegaly, bowel sounds),

neonates should also be assessed for jaundice, signs of raised intracranial pressure (setting sun sign, widened cranial sutures or an abnormal increase in head circumference), inguinal herniae, genital malformations and imperforate anus. Passage of meconium does not exclude the latter as meconium may be passed via a perineal fistula, especially in females.

First-line investigations include blood gas, glucose, electrolytes and renal function, liver functions, full blood count and abdominal X-ray (anteroposterior and lateral decubitus). Infants with bilious vomiting require surgical consultation and an upper gastrointestinal contrast study to assess for malrotation. Bowel loops are considered dilated when their diameter is greater than the width of the lumbar vertebral bodies. Multiple dilated loops suggest distal obstruction (ileocolic), but this may also be seen with ileus, although bowel loops in ileus are more uniform in appearance. Air fluid levels are seen to increase the more distal the obstruction but are often less prominent than in older children. The abdominal X-ray should be reviewed specifically for free air (central translucency or translucent rim under the diaphragm), calcification (meconium peritonitis), intramural or portal gas (necrotising enterocolitis), inguinal herniae and absence of rectal gas (in term infants gas reaches the rectum by the end of first day). A gasless abdomen can indicate ileus or ascites.

In the absence of an anatomical cause or if there are features suggestive of infection, infants should undergo septic screen (see above). Neuroimagining and further metabolic testing (blood for lactate, ketones, ammonia, amino acids, Guthrie card [if not already done] and galactosaemia screen, and urine for organic acids) may be indicated if vomiting persists.

Infants with abdominal distension and suspected intestinal obstruction require prompt stabilisation with gastric decompression by large-bore gastric tube (10–12 French), establishment of intravenous access and fluid resuscitation (0.9% saline 10 to 20 mL kg^{-1}), followed by cessation of feeds and maintenance fluid (0.9% saline with 10% dextrose at 100 to 120 mL kg^{-1} day^{-1}). Gross abdominal distension can compromise respiratory function, and supplementary oxygen may be required. Broad-spectrum intravenous antibiotics should be commenced, after obtaining blood culture, if there is perforation or distal obstruction (amoxycillin, aminoglycoside and metronidazole). Further investigation and management will be determined in consultation with surgeons.

Intestinal obstruction

Small bowel atresia

Intestinal atresia is the most common cause of neonatal intestinal obstruction.[11] Duodenal atresia classically has a double bubble appearance on X-ray, and approximately one-third of cases are associated with trisomy 21 (Down syndrome). Duodenal obstruction may be incomplete (stenosis or web) due to incomplete recanalisation of the lumen during the 10th week of gestation, whereas jejunoileal lesions usually result from mesenteric vascular accidents in utero leading to an atretic segment. Vomiting begins within a day or two of birth – the higher the atresia, the earlier the vomiting – and infants fail to pass meconium. Bilious vomiting is most common in jejunoileal obstruction but can occur in duodenal atresia as lesions are often distal to the ampulla of Vater. In jejunal atresia the X-ray typically has a few large dilated loops, whereas in ileal atresia there are usually multiple dilated loops and prominent abdominal distension.

Malrotation and midgut volvulus

In classical malrotation the midgut fails to complete its normal rotational development such that the duodenojejunal flexure lies to the right of the midline, the caecum is free-floating and the small bowel mesentery is attached to a narrow pedicle. This predisposes the midgut to volvulus around the superior mesenteric vessels. Most cases (75%) present in the neonatal period with sudden onset of pain, irritability and bilious vomiting. As strangulation ensues, there is progressive abdominal distension, rectal bleeding and hypovolaemic shock, although symptoms may be intermittent initially. Early diagnosis and surgery are essential to preserve gut viability; thus, it is imperative that neonates with unexplained bilious vomiting have an upper gastrointestinal contrast study to exclude malrotation.[12,13]

Meconium ileus

Meconium ileus is an obstruction of the small bowel, usually distal ileum, caused by highly viscoid meconium. Most cases (80% to 90%) are due to cystic fibrosis, and between 10% and 15% of infants with cystic fibrosis present in this way. There is marked distension early in the neonatal period, progressive bile-stained vomiting and failure to pass meconium, though small pale rectal plugs may be passed. Fluid levels are uncommon on X-ray because of the viscid meconium, which may give the intestine a bubbly appearance. Intraperitoneal

calcifications are indicative of fetal perforation. Surgery is required in complicated meconium ileus, but uncomplicated cases may be managed with an isotonic contrast enema.

Hirschsprung disease

Hirschsprung disease is caused by failed migration of colonic ganglion cells, leading to tonic intestinal contraction and functional obstruction. Most cases involve the rectosigmoid colon, but aganglionosis of the entire colon and rarely small bowel can occur. Hirschsprung disease is more common in males (4:1) and typically presents in the neonatal period with delayed passage of meconium, reluctance to feed, abdominal distension and vomiting, which may be bilious. Contrast enema may show a transition zone, and diagnosis is confirmed by rectal biopsy.[14] Initial management is by rectal lavage with warmed 0.9% saline, which also helps to distinguish Hirschsprung disease from meconium plug obstruction, in which a pale plug is passed. Infants with marked distension, explosive diarrhoea or fever require close monitoring for Hirschsprung entercolitis, a potentially fatal complication; treatment is with broad-spectrum antibiotics and repeated rectal lavage.

Hypertrophic pyloric stenosis

Hypertrophic pyloric stenosis is an acquired obstruction of the gastric outlet due to progressive thickening and elongation of the circular muscle of the pylorus. Risk factors include being male (4:1), Caucasian, formula fed, maternal smoking, and family history (up to 20-fold increased risk if a sibling or parent has been affected).[15] Infants develop non-bilious vomiting after feeds, typically from 2 to 6 weeks of age, which becomes progressively more frequent and forceful leading to dehydration and weight loss. They often appear hungry after emesis. Pyloric stenosis classically results in hypochloraemic, hypokalaemia metabolic alkalosis, which although present in only half of cases has high positive predictive value (88% if pH >7.45, chloride <98 mmol L^{-1} and base excess >3).[16] However, plasma electrolyte concentrations may significantly underestimate total body deficits. Diagnosis is confirmed by ultrasound.

Pyloromyotomy is delayed until correction of alkalosis ($HC0_3$ <30 mmol L^{-1}) to reduce the risk of post-operative apnoea. Once feeds are stopped, most infants can be managed without a gastric tube, and this helps to minimise electrolyte losses. Infants with moderate to severe dehydration (≥10%) should receive a

normal saline bolus of 20 mL kg^{-1}. The estimated fluid deficit is then replaced with 0.9% saline and 10 mmol KCL per 500 mL over 48 hours if ≥10% dehydration or 24 hours if <10%, in addition to intravenous maintenance fluids. Glucose, electrolytes and gas should be checked within 6 hours and then 6 to 12 hourly. Replacement of chloride is a key factor in reversal of alkalosis (Cl 10 mmol kg^{-1} reduces $HC0_3$ by approximately 3 mmol L^{-1}).[17]

The neonate with seizures

Seizures are more common in the first week of life than at any other time, partly because the neonatal brain has increased susceptibility to seizures. Most neonatal seizures occur as acute, reactive events, but a wide range of congenital disorders may also be implicated, including structural brain abnormalities, inborn errors of metabolism and epilepsy syndromes (Table 3.4.4). Early identification and treatment of seizures are important as there is increasing evidence that they can damage the brain or exacerbate pre-existing injury.[18] However, clinical diagnosis is challenging and electroclinical dissociation common, requiring a high index of suspicion.[19]

Neonatal seizures are divided into four main types – clonic, myoclonic, subtle and tonic – and further classified as focal, multifocal or generalised. Classic tonic-clonic seizures are very rare in neonates. Clonic seizures consist of rhythmic jerking of one limb or one side of the face or body and may be multifocal and migratory. Infants usually remain conscious during episodes. Myoclonus is sudden jerking movements, usually of flexor muscles groups, that are irregular and arrhythmic. It can be focal or involve generalised spasms. Subtle seizures include repeated, stereotypical movements of the mouth, face or limbs (e.g. blinking, lip smacking, cycling), apnoea and autonomic disturbances (change in blood pressure or heart rate). Many subtle phenomena do not represent seizures, but distinguishing subtle seizures from other behaviour is difficult on clinical grounds alone. Tonic seizures are the least common type and involve sustained posturing of the limbs or trunk or deviation of the head or eyes.

Several types of benign neonatal hyperkinetic movements are readily discriminated from seizures. Jitteriness resembles a tremor, is triggered by stimulation and is suppressed by gentle restraint. Jitteriness is common in preterm infants, can be exacerbated by metabolic disturbances such as hypoglycaemia, and is frequent during neonatal drug withdrawal, particularly

with selective serotonin reuptake inhibitors.[20] Hiccups are common in the neonatal period and are generally benign, though if excessive may indicate non-ketotic hyperglycinaemia. Benign neonatal sleep myoclonus involves myoclonic jerks during non-rapid eye movement sleep, either focally or bilaterally, but stops abruptly on arousal. It may be stimulated by rocking the baby to sleep. No treatment is needed, and resolution occurs by 6 months of age in most cases (95%).[21]

In term infants, over half of all seizures occur with hypoxic-ischaemic encephalopathy, typically at 18 to 20 hours of age with peripartum insults or in the first 12 hours with prelabour insults.[22] Among neonates presenting after the immediate postnatal period (>24 to 48 hours), seizures are most commonly due to biochemical disturbances, intracranial haemorrhage, arterial ischaemic stroke and meningoencephalitis.[23] A small amount of subarachnoid or subdural bleeding is not uncommon in healthy term newborns,[24] but more extensive extra-axial or cerebellar or cortical haemorrhage can cause seizures. Intraventricular haemorrhage, while common in preterm infants, is rare at term and should prompt further investigation for sinovenous thrombosis.[25] Intracranial haemorrhage and sinovenous thrombosis may result from non-accidental injury.

Infants with unilateral clonic seizures who are well appearing and have normal Apgar scores are likely to have middle cerebral artery stroke, which occurs more commonly on the left (right-sided symptoms). Peripartum stroke is thought to arise from placental emboli that enter the arterial circulation via the foramen ovale. Thus, many risk factors for neonatal stroke are related to placental disease, such as fetal growth restriction and pre-eclampsia.

Table 3.4.4

Causes of late neonatal seizures (>48 hours)

Cerebrovascular	Middle cerebral artery stroke Extra-axial haemorrhage Periventricular haemorrhage Sinovenous thrombosis
Biochemical	Hypoglycaemia Hypocalcaemia, hypomagnesaemia Hypernatraemia
Meningoencephalitis	Bacterial: predominantly group B streptococcus, *Escherichia coli*, *Listeria monocytogenes*, *Streptococcus pneumoniae*; rarely *Haemophilus influenzae*, other enterobacteriaceae Viral: HSV, enteroviruses
Miscellaneous	Neonatal drug withdrawal: opioids, selective serotonin reuptake inhibitors Non-accidental injury

Rare causes	Cerebral malformations and tumours Inborn errors of metabolism: pyridoxine dependency, biotinidase deficiency, non-ketotic hyperglycinaemia (glycine encephalopathy), other metabolic encephalopathies Benign seizure syndromes: benign familial neonatal seizures, benign non-familial neonatal convulsion (fifth-day fits) Malignant seizure syndromes: early myoclonic epilepsy, early infantile epileptic (Ohtahara) encephalopathy

HSV, herpes simplex virus.

Intracranial infections usually present after the first week, and group B streptococcus and *Escherichia coli* are the most common bacterial causes. Herpes simplex virus can mimic bacterial meningitis as approximately one-third of cases occur without cutaneous lesions, and mothers frequently do not have active genital herpes at the time of birth.

First-line investigations for suspected seizures include blood gas, lactate, glucose, electrolytes (sodium, calcium and magnesium), full blood count and, if not already completed, newborn metabolic screen (Guthrie card). Blood culture and lumbar puncture are usually performed but may be omitted if the baby appears extremely well and a non-infectious aetiology is likely. Cerebrospinal fluid, if sent, should be tested for herpes simplex virus and enteroviruses. Brain MRI is indicated in most cases and can usually be performed without sedation. If intracranial haemorrhage is detected, screening for coagulopathy and for haemophilia in males is warranted.

Hypoglycaemia (<2.6 mmol L^{-1}), hypocalcaemia (ionised calcium <1 mmol L^{-1}) and hypomagnesaemia (<0.8 mmol L^{-1}) should be corrected acutely (Table 3.4.5). If there is cerebrospinal fluid leucocytosis (>20 x 10E6 L^{-1} in neonates <7 days of age, >10 x 10E6 L^{-1} in neonates ≥7 days of age) antibiotics are commenced (cefotaxime, amoxycillin, gentamicin), with addition of aciclovir if no bacteria are seen on the Gram stain.

Antiepileptics are generally administered if clinical seizures last more than 3 minutes and phenobarbitone is used most commonly in neonates. Levetiracetam is increasingly being used either as a first-line agent or as an adjunct to phenobarbitone.[26] Other second-line agents include phenytoin and midazolam. Infants requiring antiepileptic drugs should be admitted for continuous (electroencephalogram) EEG monitoring as up to one-third fail to respond to treatment and electroclinical dissociation is common, especially after administration of phenobarbitone. Neurology consultation is recommended if more than two agents are required. Infants with possible seizures should be admitted for observation and diagnostic EEG.

The neonate with breathing difficulty

Signs of respiratory distress include tachypnoea (>60 breaths per minute), increased effort (intercostal and subcostal recession, grunt and tracheal tug), apnoea (a pause in breathing >20 seconds or pause of <20 seconds associated with bradycardia) and stridor. In neonates with breathing difficulty the oropharynx, chest and cardiovascular system should be carefully examined and a chest X-ray performed. Blood work will generally include blood gas, full blood count, inflammatory markers and blood culture, with further investigation according to likely aetiology. Neonates with respiratory distress should be observed and monitored closely. The causes of respiratory distress can be broadly divided into pathology of lung parenchyma, airway obstruction and non-pulmonary disease.

Lung parenchymal pathology

After the immediate newborn period, respiratory distress from parenchymal lung disease is mainly caused by pneumonia and rarely congenital lung malformations. Congenital bacterial pneumonia is transmitted via the amniotic fluid and presents with respiratory distress in the first 12 to 24 hours. Group B streptococcus is the most common aetiology followed by *Escherichia coli* and occasionally other bacteria such as *Haemophilus influenzae*, *Streptococcus pneumoniae* and Listeria. Late onset group B streptococcus infection (>48 hours) can occur either from vertical and horizontal transmission, but most cases of late disease present with septicaemia or meningitis rather than isolated pneumonia.[27] Intrapartum antibiotic prophylaxis may delay the presentation of group B streptococcal disease. In the later neonatal period acquired viral pneumonias predominate, especially respiratory syncytial virus, with adenovirus and parainfluenza also common. A history of respiratory infection in family members supports a viral cause.

Several atypical lung infections can also present in the later neonatal period, including pertussis, chlamydia and cytomegalovirus. Pertussis causes cough, vomiting, apnoea and choking spells, but whooping is rare in neonates. Maternal pertussis immunisation during pregnancy is up to 95% effective in reducing neonatal pertussis infection. Chlamydia pneumonia should be considered if there is a history of late onset conjunctivitis, which may be due to *chlamydia trachomatis*. Cytomegalovirus may be transmitted horizontally in breast milk

and cause a sepsis-like illness, including pneumonia. Recurrent pneumonia, stridor, choking or feeding difficulty should prompt further assessment for aspiration pneumonia, including consideration of barium swallow.

Chest X-ray appearance of neonatal pneumonia is highly variable, including lobar or diffuse opacities and coarse or nodular patterns. Effusions can occur with both group B streptococcal infection and congestive heart failure, though cardiomegaly and more prominent radiating hilar densities distinguish the latter. Nasopharyngeal aspirate for respiratory viruses and blood culture should be obtained. Neonates with pneumonia should be admitted and closely monitored, with supplementary feeding by nasogastric tube, as required. Low flow nasal oxygen is given to maintain oxygen saturations >92%, and if respiratory effort is marked, continuous positive end-expiratory pressure or heated humidified high flow may be required. Neonates with respiratory syncytial virus pneumonia are particularly prone to apnoea and bradycardia, though apnoea can occur with any respiratory infections and sepsis. Apnoea will often improve on positive pressure respiratory support, but caffeine is also used, usually starting with an oral loading dose (caffeine citrate 20 mg kg^{-1}), followed by maintenance if required (5 mg kg^{-1} daily). Broad-spectrum antibiotics (e.g. amoxycillin and gentamicin) are usually given initially because of the difficulty in distinguishing bacterial and viral pneumonia and risk of secondary infection.

Major congenital lung malformations cause respiratory distress at birth, but smaller lesions may present after the birth transition or later in the neonatal period. These include congenital diaphragmatic hernia (e.g. small posterolateral or anterior defects) and cystic lung malformations (e.g. congenital cystic adenomatoid malformation, congenital lobar emphysema). Gas trapping in cystic malformations can cause compression of underlying lung, and lesions may also become infected. Lung hyperinflation can also result from extrinsic bronchial compression from a bronchogenic cyst or vascular mass. Gas trapping should not be confused with pneumothorax, and lateral X-ray can be helpful in identifying the location of an air-filled lesion or air leak.

Table 3.4.5

Treatment of neonatal seizures

First line	Phenobaritone 20 mg kg^{-1} IV (up to 40 mg kg^{-1}) over 20 min Levetiracetam 20 mg kg^{-1} IV (up to 40 mg kg^{-1}) over 20 min
Second line	Phenytoin 20 mg kg^{-1} IV over 20 min Midazolam IV infusion 0.5–2 ug kg^{-1} min^{-1}

Hypocalcaemia	Calcium gluconate 10% 0.5 mL kg^{-1} (0.11 mmol kg^{-1} Ca^{2+}) over 20 min
Hypomagnesaemia	MgS04 50% 0.2 mL kg^{-1} IMI (0.4 mmol kg^{-1} Mg^{2+})
Hypoglycaemia	Dextrose 10% 2 mL kg^{-1} IV (200 mg kg^{-1}) bolus

Table 3.4.6

Causes of neonatal stridor

Luminal obstruction	Cysts, webs, haemangiomas
Airway wall	Laryngomalacia Tracheobronchomalacia Tracheal stenosis Laryngeal cleft
Vocal fold immobility	Birth trauma, Arnold–Chiari malformation, neurological disease
Extrinsic	Vascular rings Neck and mediastinal masses (e.g. haemangiomas)

Airway obstruction

Strider, or high-pitched noisy breathing, is an important sign of obstruction within the larynx, trachea or bronchi. The pattern of stridor can help to identify the level of obstruction: supraglottic is inspiratory, subglottic is biphasic, and lower tracheal or bronchial obstruction is expiratory and associated with prolonged expiration. Unilateral vocal cord immobility results in a husky cry and may cause inspiratory stridor. Bilateral vocal cord immobility can cause inspiratory or biphasic stridor and is associated with a vocal quality to the inspiratory noise. Tracheomalacia produces a barking cough. It is important to note that in the early neonatal period stridor may be soft due to small tidal volumes and low airflow velocity.

The most common cause of stridor in neonates is laryngomalacia, but there are many other congenital lesions to be considered (Table 3.4.6). Viral croup does not occur in neonates, and bacterial infections of the airway are rare. The following signs suggest more significant pathology, and further workup with blood gas, lateral neck X-ray and referral for diagnostic endoscopy is warranted: stridor from birth, biphasic stridor, abnormal cry, aspiration pneumonia, feeding difficulty, choking, apnoea or failure to thrive. Superficial haemangiomas of face and neck can be associated with subglottic haemangiomas. CT or MRI may be needed to exclude extrinsic lesions.

Neonates with acute respiratory distress and stridor can usually be stabilised with continuous positive end-expiratory pressure and oxygen. An adrenaline nebuliser may be tried (0.5 mL kg^{-1} of 1:1000 adrenaline mixed with 2–3 mL of normal saline). If there is respiratory failure despite these measures, ventilation will be required.

Laryngomalacia

Laryngomalacia results from laxity of the ~~subglottic~~ supraglottic structures which collapse into the laryngeal inlet on inspiration. Stridor is absent at birth and usually develops from the second or third week. It is exacerbated by feeding, crying and lying supine and is diminished in the prone position. If there is no respiratory distress and the infant is thriving, treatment is expectant, including monitoring of growth. Stridor may increase in the first few months of life but usually resolves spontaneously by around 18 months of age.[28] Infants with respiratory distress, failure to thrive, feeding difficulty or atypical signs should be referred to an otolaryngologist as secondary lesions are not uncommon, and surgical correction by supraglottoplasty may be required.[29] Infants with moderate to severe laryngomalacia often have gastro-oesophageal reflux and typically require anti-acid therapy.[29]

Non-pulmonary disease

The main non-pulmonary cause of breathing difficulty in neonates is congestive heart failure. Clinical features include tachycardia, tachypnoea, recession, hepatomegaly, cardiomegaly (cardiothoracic ratio >0.6), pulmonary plethora and often a systolic murmur. Left-to-right shunts (e.g. ventricular septal defect, patent ductus arteriosus) are the most common cause of neonatal heart failure. Differential diagnoses include left-heart obstructive lesions (e.g. coarctation), cardiomyopathy (e.g. hypertrophic, viral, sustained tachyarrhythmia), high output failure (e.g. severe anaemia, arteriovenous malformations) and other congenital lesions associated with increased pulmonary flow (e.g. truncus arteriosus, anomalous pulmonary venous drainage). Left-to-right shunts manifest as the pulmonary vascular resistance falls, with the postnatal decline in haematocrit also play a role.[30] Large shunts, especially with combined lesions, may present from two to three weeks of age, but moderate shunts present later, and the neonatal signs may be subtle, including poor feeding, slow weight gain and mild tachypnoea. Left-heart obstructive lesions, tachyarrhythmias, large

arteriovenous malformations and other complex congenital heart lesions generally present in the early neonatal period.

In the neonate with effortless tachypnoea and normal to low pCO_2 metabolic acidosis and hyperammonaemia should also be considered in the differential, especially in the absence of obvious cardiopulmonary disease.

The neonate with prolonged jaundice

Neonatal jaundice is a common presentation to the ED. While most cases represent simple physiological jaundice, it is important that conjugated or prolonged jaundice is identified and promptly investigated as this can be a sign of serious underlying pathology (Table 3.4.7). Extreme hyperbilirubinaemia is a medical emergency and must be treated aggressively as it can cause irreversible brain damage (kernicterus) and chronic bilirubin encephalopathy.

Most babies develop elevated bilirubin concentrations in the first week of life, and physiological jaundice occurs in more than 50%. This is due to increased production (accelerated red blood cell breakdown), decreased removal (transient liver enzyme insufficiency) and increased reabsorption (enterohepatic circulation) of bilirubin. The jaundice first appears on day two to three, and the peak serum bilirubin concentration is generally below 300 μmol L^{-1}. It usually disappears by one to two weeks of age, and the levels of bilirubin do not cause harm to the neonate. It is a diagnosis of exclusion.

Pathological jaundice is suspected when jaundice appears in the first 24 hours after birth, if the bilirubin concentration is increasing rapidly (>8.5 μmol L^{-1} hr^{-1}) or is significantly elevated (>50 μmol L^{-1} above the phototherapy threshold), is prolonged (>14 days in term or >21 days in preterm neonate), includes a significant conjugated component (>25 μmol L^{-1} or >20%) or stool is pale, or if the baby is unwell.

Jaundiced infants will need measurement of unconjugated and conjugated bilirubin concentrations, full blood count and film, blood group and direct Coombs test. If a mother has received antenatal anti-D immunoglobulin prophylaxis, the direct Coombs test may be weakly positive. Glucose-6-phosphate dehydrogenase deficiency screen should be considered in severe unconjugated hyperbilirubinaemia. Babies with prolonged unconjugated jaundice also need thyroid and liver function tests, urine for microscopy and culture, and reducing substances (or specific galactosaemia test). Haemoglobinopathy and red cell membrane and enzyme screen are indicated in

suspected non-immune haemolysis (anaemia, spherocytes or red cell fragments on film, reticulocytosis >6%).

Prolonged unconjugated hyperbilirubinaemia in a breast-fed infant may be due to breast-milk jaundice. This is a diagnosis of exclusion and should only be made if the infant is thriving, there is no evidence of haemolysis and screening investigations are normal. It is thought to be the presence of β-glucuronidase in breast milk which unconjugates bilirubin in the gut, resulting in increased enterohepatic circulation. Parents should be advised to seek medical advice if the nature of the jaundice changes, if there is failure to thrive or stool becomes pale.

Conjugated hyperbilirubinaemia is due to biliary atresia (extrahepatic biliary obstruction) until proven otherwise, as early corrective surgery must be performed if the infant is to avoid progression to liver failure. Biliary atresia causes acholic (white or pale) stool, although this may be absent in the first weeks after birth. An ultrasound is performed to identify a gall bladder, although presence of a gall bladder does not fully exclude the diagnosis. The differential diagnosis of conjugated hyperbilirubinaemia is extensive and workup should be performed in consultation with a gastroenterologist.

Phototherapy is commenced if the bilirubin concentration is significantly elevated (Table 3.4.8). If the bilirubin concentration is extreme, an exchange transfusion may be required in addition to treatment of the underlying cause. Intravenous immunoglobulin (0.5 g kg^{-1} over 4 hours) may reduce the need for exchange transfusion, though evidence of benefit is not conclusive.[31]

Table 3.4.7

Causes of prolonged and conjugated neonatal jaundice

Prolonged unconjugated	Haemolysis:*
	Isoimmune, e.g. ABO haemolytic disease
	Hereditary spherocytosis (25% sporadic, >10–15% spherocytes)
	Red cell enzyme defects, e.g. pyruvate kinase deficiency
	Alpha and gamma globulin chain structural abnormalities
	Hypothyroidism Glucose-6-phosphate dehydrogenase deficiency (X linked, usually not haemolytic in newborns) UGT1A1 defects (Crigler Najjar or Gilbert with ABO haemolytic disease) Urinary tract infection

	Galactosaemia Breast milk-associated jaundice
Conjugated	Hepatocellular dysfunction: Infection: syphilis, toxoplasmosis, rubella, cytomegalovirus, Epstein–Barr virus, herpes simplex virus, enteroviruses Metabolic: galactosaemia, tyrosinaemia Alpha-1 antitrypsin deficiency Neonatal haemochromatosis Idiopathic neonatal hepatitis Biliary obstruction: Intrahepatic: intravenous nutrition, severe Rhesus haemolytic disease, inspissated bile syndrome, cystic fibrosis, Alagille syndrome, Caroli disease, Byler disease, inborn errors of bile acid biosynthesis Extrahepatic: biliary atresia, choledochal cyst Inherited defects of bilirubin excretion, e.g. Dubin-Johnson

* Features of haemolysis include anaemia, reticulocytosis (>6% by day 3), spherocytes or red cell fragments.

Adapted from Management of hyperbilirubinemia in the newborn infant 35 or more weeks of gestation. *Pediatrics*. 2004;114(1):297–316.

Table 3.4.8

Guidelines for phototherapy in term and near-term newborns[32]

Postnatal age (hours)	Bilirubin concentration (μmol L^{-1})			
	≥38 weeks' gestation and well		35–37 weeks' gestation or risk factors*	
	Phototherapy	Consider exchange if intensive phototherapy fails	Phototherapy	Consider exchange if intensive phototherapy fails
24	205	325	170	280
36	235	350	200	300
48	265	375	225	325
60	290	390	240	340
72	310	410	260	360
96	340	430	290	385
≥120	360	435	290	385

* Risk factors include haemolysis, sepsis, asphyxia, hypoalbuminaemia. Immediate exchange should be considered with bilirubin concentration >430 μmol L^{-1} <48 hours or >510 μmol L^{-1} ≥48 hours or if the rate of rise is >8.5 μmol L^{-1} hr^{-1} despite intensive phototherapy (two overhead units and a BiliBed).

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3.5

Neonatal resuscitation

Gary David Williams

ESSENTIALS

1 Occasionally deliveries occur in the emergency department (ED) when labour cannot be delayed to enable transfer of the patient to a labour ward setting.

2 The ED should have available appropriate equipment and guidelines for the resuscitation of the newborn.

3 The unique physiological events at birth affect the resuscitative interventions in the newborn.

4 Ventilation is the main priority in the resuscitation of the newborn.

5 The heart rate is a reliable indicator of the newborn baby's degree of compromise and response to resuscitation. The easiest way to monitor newborn pulses is by palpation of the umbilicus.

6 Chest compressions are indicated if: (1) absent pulse; or (2) heart rate <60 min^{-1} despite adequate assisted ventilations for 30 seconds.

7 Compressions and ventilations should be cycled at a ratio of 3 compressions to 1 ventilation to achieve approximately 120 events per minute.

8 The preferred site of the vascular access during neonatal resuscitation is the umbilical vein.

9 Drugs are rarely required in the resuscitation of the newborn.

10 Volume expansion should be with crystalloid 10 mL kg^{-1} boluses by slow push.

Introduction

Epidemiology

Between 5% and 10% of newborns require some assistance to begin breathing at birth, and, in developed countries, approximately 1% need intensive resuscitative measures to restore cardiorespiratory function. It has been estimated that birth asphyxia significantly contributes to approximately 20% of the 5 million neonatal deaths that occur worldwide each year; outcome might therefore be improved for more than 1 million newborns per year through effective resuscitation at birth.

Neonatal resuscitation is unique in that it is required at a time when the newborn is undergoing a predetermined process of transition from a liquid-filled intrauterine environment to spontaneous breathing of room air. There is an accompanying sequence of dramatic alterations in physiology, each of which may be altered and require correction.

There are two important caveats in this process. First, the achievement of lung expansion with an appropriate oxygen-containing gas leading to establishment of a functional residual capacity and adequate spontaneous ventilation is of primary importance. Second, the significance of a vital sign abnormality depends greatly on the time since birth and the time during which effective resuscitation measures have been administered. For instance, bradycardia immediately after birth prior to any resuscitative manoeuvres likely indicates an intrapartum stress. The same heart rate after 1 to 2 minutes of adequate ventilation suggests a different range of aetiologies and requires a different resuscitative response.

The majority of circumstances where newborn resuscitation is needed can be predicted, allowing opportunity for preparation of appropriate equipment and personnel. Factors placing the newborn at high risk for neonatal resuscitation include those listed in Table 3.5.1, due to maternal, fetal and intrapartum circumstances.

Aetiology and pathophysiology

The sequence of physiological changes in the newborn around birth includes the following:

- Cessation of alveolar fluid production and clearance of this fluid from the gas-exchanging part of the airway

- Spontaneous respirations and establishment of functional residual capacity (FRC)
- Fall in pulmonary vascular resistance (PVR) (lung expansion, oxygen)
- Rise in systemic vascular resistance (SVR) (umbilical artery constriction)
- Reversal of flow from left to right across foramen ovale and ductus arteriosus
- Closure of the foramen ovale and ductus arteriosus.

This normal sequence may be interrupted at any point by the following:

- Inadequate clearance of endogenous lung liquid (precipitate, emergency or operative delivery) or excessive abnormal airway material (blood, mucus, meconium)
- Inadequate respiratory effort (maternal analgesics, central nervous system injury, sepsis) or excursion (congenital thoracic anomalies)
- Pulmonary disease preventing achievement and maintenance of adequate FRC (parenchymal disease, prematurity, space-occupying lesion) with secondary failure to lower PVR normally and possible intrapulmonary shunting
- Compromised myocardial function (structural or functional cardiac abnormalities, hypoxia secondary to pulmonary dysfunction).

Interventions should occur in a defined sequence recognising the primary and crucial role of adequate ventilation and be guided by frequent reassessment of other vital signs.

Preparation

1. Summon assistance: if it is anticipated that the infant is at high risk of requiring advanced life support resuscitative intervention, more than one experienced person should be mobilised.
2. Communicate with colleagues to seek available antenatal and intrapartum data.
3. In the case of extreme prematurity (≤24 weeks' gestation) or known congenital anomalies, if time allows, the most senior care provider should seek to discuss with the family their beliefs and desires regarding

the extent of resuscitation.[1]

4. Prepare the environment: this includes a warm, draught-free area with adequate light, radiant heater, pre-warmed blankets and a clock.
5. Prepare required equipment (Table 3.5.2).

Assessment at birth

If the infant is born at term, has good tone and is breathing or crying the infant can stay with the mother in a warm environment. The infant will need to be dried and wrapped to maintain normal temperature and be closely observed for any deterioration. Otherwise at birth the infant should be collected in a warm towel, dried and the umbilical cord clamped, and taken to the neonatal resucitaire. Gentle stimulation may be provided (rubbing the back, flicking soles of feet if required), and an assessment of initial cry, respiratory effort, heart rate, colour and tone should be made virtually simultaneously (Apgar score, Table 3.5.3).

Ventilation

The initial assessment is an evaluation of the presence and quality of respirations:

- If respirations are adequate, the heart rate is evaluated.
- If respirations are shallow or slow, a brief period of stimulation may be provided.

Table 3.5.1

Risk factors for needing neonatal resuscitation

Maternal	Fetal	Intrapartum
Premature or prolonged rupture of membranes	Multiple gestation	Fetal distress
Antepartum haemorrhage	Preterm (<35 weeks) or post-term (>42 weeks) gestation	Abnormal presentation
Hypertension	IUGR	Prolonged or precipitate labour
Diabetes mellitus	Polyhydramnios or oligohydramnios	Meconium staining of amniotic fluid
Substance abuse	Congenital abnormalities	Instrumental delivery or emergency caesarean section
Maternal infection or chronic illness		

Absence of antenatal care		

IUGR, intrauterine growth restriction.

Table 3.5.2

Equipment and drugs recommended for newborn resuscitation

Equipment	Drugs
Stethoscope	Adrenaline (epinephrine) 1 in 1:10000 solution
Suction catheters (6–12 French) and suction	Naloxone hydrochloride 1.0 or 0.4 mg mL^{-1}
8 French feeding tube and 20 mL syringe for gastric decompression	Dextrose 5% or 10%
Face masks	$NaHCO_3$ 4.2% solution
Oropharyngeal (Guedel) airways	Volume expander (0.9% saline, Ringer's lactate or 4% HSA)
Resuscitation system for PPV	
Laryngoscope with straight blade	
ET tubes 2.5 to 4 mm internal diameter	
ET stylets	
Tape for ETT and IV fixation	
Cannulae, syringes and UV catheterisation equipment	

HSA, human serum albumin; PPV, positive pressure ventilation.

- If the infant has not established adequate breathing by 30 seconds, face mask rescue breaths should be administered, and if this is not successful with breathing established by 2 minutes, endotracheal (ET) intubation should be performed. Correct position of the ET tube should be confirmed using colourimetric exhaled carbon dioxide detector.
- A laryngeal mask may be considered an alternative to tracheal intubation (size 1 LM recommended for all infants <5 kg) if face mask ventilation is unsuccessful or not feasible.
- There should be no delay in commencing rescue breaths if the infant is born with, or develops evidence of, asphyxia with signs of flaccidity, pallor and/or bradycardia (heart rate less than 60 beats min).

Artificial ventilation

Various bag and mask systems are available for neonatal resuscitation. T-piece

mechanical devices designed to regulate pressure, self-inflating bag or flow-inflating bag are all recognised as acceptable devices for ventilating newborn infants either via a face mask or ET tube. Target inflation pressures, continuous positive airway pressure (CPAP) and long inspiratory times are achieved more consistently using T-piece devices than when using bags, but the ability to achieve an increased inspiratory pressure when required in response to altered compliance (even for a few breaths) is greatest with the self-inflating bag.[2] It is suggested in fact that the invariable success of rescue breathing at birth is because the FRC is established by an induced inspiratory effort via Head's paradoxical reflex (inspiratory effort induced by any lung inflation). The corollary is that face mask rescue breathing is unlikely to be effective in the severely asphyxiated infant.

Table 3.5.3

Apgar score

Score	0	1	2
Heart rate	Absent	Less than 100	Greater than 100
Respiratory effort	Absent	Irregular	Crying
Muscle tone	Limp	Some flexion	Active motion
Reflex irritability	Absent	Grimace	Grimace and cough
Colour	Blue, pale	Acrocyanosis	Pink all over

Regardless of these issues, it is generally accepted that higher inflation pressures (>30 cmH_2O) and longer inflation times (>1.5 seconds) may be required for the first several (≈5) breaths. Initial peak inflating pressures required are variable and unpredictable. In general, the minimum pressure required to achieve an increase in the heart rate should be used. Visible chest wall movement and an increase in the heart rate are the best indicators of adequate ventilation. Ventilations should be administered at a rate of 40 to 60 breaths min, and after 30 seconds of effective ventilation, the heart rate should be evaluated.

Heart rate

Assessment of the heart rate can be done by palpating the umbilical stump, brachial or femoral pulse or auscultation of the apical heart sounds. Clinical assessment of heart rate by these methods can be intermittent and inaccurate, and prompt commencement of oximetry or application of three chest ECG leads can provide a continuous and more accurate number:

- The heart rate is a reliable indicator of the degree of compromise and

response to resuscitation. It should be assessed at least every 30 seconds in the first 2 minutes if necessary, until the baby's required level of support is established.

- If the heart rate is less than 60 beats min despite adequate assisted ventilation, compressions are required. When the heart rate is greater than 100 beats min, only ventilation is continued. If the heart rate is between these two points, the level of intervention should be increased or decreased depending on the serial change in the heart rate.
- If compressions are initiated it is recommended that inspired oxygen concentration be increased to 100%.
- If the heart rate is not rising after 30 seconds of effective ventilation with 100% oxygen (preferably through an ET tube) combined with chest compressions, then adrenaline (epinephrine) should be administered.
- Once a slow heart rate has increased above 60 beats min and is rising, cardiac compressions may be discontinued.

Compression technique

If required, compressions should use the two-thumbs-encircling-hands technique. The lower third of the sternum (just below an imaginary intermammary line) should be depressed one-third of the depth of the chest. These should be coordinated with ventilations (to avoid simultaneous delivery) in a ratio of 3:1 with about 90 compressions and 30 breaths each minute. The xiphoid portion of the sternum should not be compressed because such compression may damage the neonate's liver.

Colour

Once the heart rate has been evaluated, the infant's colour should be assessed by examining the trunk and mucosae. It is noteworthy that clinical assessment of colour in isolation is an unreliable indicator of oxygenation or the effectiveness of resuscitation:[3]

- Peripheral cyanosis (acrocyanosis) is common in the first few minutes after birth and is not pathological.
- Central cyanosis reflects inadequate oxygenation and may be pulmonary or cardiac in origin. If present despite adequate ventilation and a heart

rate greater than 100 beats per minute, pulse oximetry should be commenced.

- Oximetry should guide oxygen administration to limit hyperoxia, especially in the premature. The aim should be to target SpO_2 levels achieved in healthy term infants during the first minutes of life remembering that an SpO_2 of 70% is at the lower end of the normal range at 5 minutes of age in a healthy term baby (Table 3.5.4).
- Pallor is suggestive of a decreased cardiac output and may be due to myocardial dysfunction, severe anaemia, hypovolaemia, hypothermia or acidosis.

Table 3.5.4

Preductal SpO_2 targets after birth

Age	SpO_2 target
1 min	60–65%
2 min	65–70%
3 min	70–75%
4 min	75–80%
5 min	80–85%
10 min	85–95%

Muscle tone and reflex irritability

These physical signs are valuable composite reflections of the adequacy of cerebral perfusion and oxygenation. As such, they constitute two of the five components of the Apgar score (see Table 3.5.3) used to assess a newborn's condition after birth.

Medications

Medications are rarely required during neonatal resuscitation. One study suggested medications were required in only 0.12% of all births, for severe fetal acidosis or ventilatory problems. This reaffirms the primary and critical importance of achieving optimal ventilation before resorting to medications in neonatal resuscitation.

Vascular access

Adrenaline may be administered by intravenous (IV), intraosseous (IO) or ET routes. If there is respiratory depression once heart rate and colour have been restored by adequate rescue ventilation, naloxone may be given via the IV or intramuscular (IM) route. The other drugs and volume expansion, detailed below, require emergency vascular access. Once vascular access is achieved, if the child remains in arrest, an adrenaline dose should be immediately administered via the IV route.

The preferred site of the vascular access during neonatal resuscitation is the umbilical vein, the larger thin-walled single vessel (in comparison to the paired thicker-walled arteries), which appears when the umbilical cord is trimmed 1 cm above the skin. A 3.5 or 5 French catheter flushed with saline (to remove any air in the tubing) should be inserted only until a good blood return is achieved (usually to a depth of 1 to 4 cm below the skin). Peripheral veins on extremities or scalp may be attempted, or alternatively an IO cannula may be placed on the medial aspect of the tibia just below the tibial tuberosity, if umbilical or other direct venous access is not readily obtainable.

Adrenaline

Adrenaline is administered with the aim of producing α-adrenergic-mediated vasoconstriction, an increase in coronary perfusion pressure and myocardial blood flow. Adrenaline is indicated if the heart rate remains less than 60 beats per minute after a minimum of 30 seconds of adequate ventilation and 30 seconds of combined ventilation and chest compressions. The recommended IV dose is 0.1–0.3 mL kg of a 1:10,000 solution (10–30 mcg kg) repeated every 3 to 5 minutes as indicated. ET delivery, though of unproven efficacy, can be considered while IV access is being established and requires a higher dose (up to 100 mcg kg); it should be followed by 1 mL of normal saline and several good inflations to achieve optimal delivery to the pulmonary vascular bed. Most infant animal experimental dosing data supporting adrenaline's efficacy have been obtained in VF models and as such their value may not be directly applicable to the apparent preterminal bradyarrhythmia in an asphyxiated newborn with markedly elevated pCO_2.

Through the early 1990s some experimental and human data showed that higher intravenous doses of adrenaline (100 mcg kg^{-1}) were capable of achieving

higher plasma adrenaline levels as well as greater myocardial and cerebral blood flow. However, several subsequent adult and paediatric studies showed no ultimate clinical benefit in survival or neurological outcome, with a significant risk of adverse effects from the higher dose (myocardial dysfunction or necrosis, hyperadrenergic states, reduced cerebral cortical blood flow). Specifically, there is an increase in potential risk of intraventricular haemorrhages (IVH) in preterm infants. For these reasons, the currently recommended initial IV dose remains 10 mcg kg in neonates.

There is a paucity of both experimental and human data regarding dosage and efficacy of ET adrenaline in neonates. There are data to suggest a slower onset with a more prolonged and variable effect at higher dosages. For these reasons the dose recommended is 50–100 mcg kg.

Naloxone

Naloxone is a narcotic antagonist, recommended for the neonate with respiratory depression secondary to narcotics given to the mother within 4 hours of delivery. Prompt institution of adequate ventilation is the first priority in such a situation, and naloxone is not recommended for newborns whose mothers are suspected narcotic abusers as abrupt withdrawal may be precipitated. Following IV administration, onset of action occurs in 1–2 minutes, although it is variable in duration. The recommended IV dose is 0.1 mg kg. Since the duration of action of narcotics may exceed that of naloxone, continued surveillance and repeat administration are often required. Naloxone may be administered IM, with some adult data suggesting slower onset and more prolonged duration of action via this route. There are no studies examining the ET route of administration in the neonate.

Dextrose

Hypoglycaemia is a potential problem for all stressed and asphyxiated babies and should be treated by using a slow bolus of 2 mL kg of 10% dextrose IV, avoiding hyperglycaemia.

Volume expansion

If hypovolaemia is present because of known or suspected blood loss or loss of

vascular tone following asphyxia, volume expansion may be appropriate. Isotonic non-dextrose containing crystalloid (normal saline or Ringer's lactate) 10 mL kg IV over 5 to 10 minutes is recommended. Group O negative packed red cells may be indicated for replacement of large volume blood loss. Albumin-containing solutions are used less frequently because of limited availability, risk of infectious disease and an observed association with increased mortality. A recent randomised controlled comparison of albumin versus normal saline for hypotension in premature newborns showed that those who received albumin required significantly more volume expander to maintain normal blood pressure and had a higher mean percentage weight gain in the first 48 hours after birth.

Bicarbonate

Although correcting acidosis during cardiac arrest to improve both myocardial function and adrenaline's effectiveness makes theoretic sense, there are few supportive experimental data. Most adult studies have found either no difference or that bicarbonate had deleterious effects on myocardial performance. There are no neonatal animal or human studies specifically examining this question. Bicarbonate administration has multiple possible adverse effects (metabolic alkalosis compromising peripheral tissue oxygen delivery, paradoxical intracellular hypercarbic acidosis). Pertinent to neonates are studies demonstrating large increases in plasma osmolality and reductions in cerebral blood flow, both of which may increase the risk of IVH in newborns. Therefore bicarbonate administration is only recommended during prolonged arrests unresponsive to other therapy after establishment of adequate ventilation and perfusion. The dose is 1–2 mEq kg of 4.2% solution by slow IV push over at least 2 minutes.

Specific resuscitation situations

Premature neonate

Preterm newborns have an increased likelihood of respiratory depression requiring assisted ventilation at birth. This occurs because of diminished lung compliance, weak respiratory muscles and immature respiratory drive and may make it difficult to establish and maintain an adequate FRC. A recent study has shown that CPAP applied promptly to premature babies who breathe at birth may be effective in reducing need for ventilation. Therefore infants born at or before

32 weeks' gestation should receive prompt CPAP via face mask rather than routine intubation for positive pressure ventilation (PPV). Resuscitation of preterms less than 35 weeks' gestation should be initiated with low oxygen (21–30%)[4] and the oxygen concentration titrated using oximetry. Also, because preterm infants often have low body fat and a high body surface area to mass ratio, they are more difficult to keep warm and therefore at increased risk of cold stress. For this reason babies <1500 g should be covered in food-grade heat-resistant plastic wrapping or placed in a polyethylene bag with the head excluded and a hat placed on the head. Rapid boluses of volume expander and use of hyperosmolar solutions may produce large fluctuations in blood pressure (increased risk of IVH) or osmolarity and therefore are not recommended.

Meconium aspiration

Approximately one in 20 infants born through meconium-stained amniotic fluid (MSAF) develops meconium aspiration syndrome (MAS) due to aspiration into the distal airways either in utero or with the initial breaths following birth. However, of these, 25–50% require mechanical ventilation, and 5% die. Although a long established practice, it is no longer recommended that infants delivered through MSAF undergo intrapartum suctioning of the oro- and nasopharynx once the head is delivered, prior to delivery of chest or shoulders. If the baby born through MSAF is not vigorous after delivery (depressed respirations, muscle tone or heart rate less than 100 beats min) emphasis should be given to initiating ventilation within the first minute of life in the non-breathing or ineffectively breathing infant. This may include intubation and suctioning if the airway is obstructed.

Congenital heart disease

Central cyanosis at birth apparently unresponsive to 100% oxygen particularly in a vigorous baby with adequate spontaneous respiratory effort and minimal respiratory distress may indicate duct-dependent cyanotic congenital heart disease (primarily right heart obstructive lesions, transposition of the great vessels and anomalous pulmonary venous return with complete atrial admixture). The major differential diagnoses are primary pulmonary hypertension or major pulmonary structural abnormalities (e.g. congenital diaphragmatic hernia). In such a circumstance ventilatory support requirement

should be dictated by degree of respiratory distress with a target $PaCO_2$ of 35–40 mmHg.

Detailed cardiac auscultation should be attempted though there may be no abnormal murmurs audible; simultaneous pre- and postductal oximetry measurements should be performed. If a cyanotic cardiac abnormality is strongly suspected, documentation of preductal PaO_2 after breathing 100% oxygen for several minutes (hyperoxia test) will give the best indication of the presence and size of a significant intracardiac right-to-left shunt (see Chapter 5.1). An urgent bedside echocardiogram, if available, is indicated to delineate the cardiac anatomy. Where this is not available, one should consult with the local tertiary neonatal unit. If a diagnosis of a duct-dependent cyanotic cardiac lesion is confirmed on echocardiogram, IV alprostadil (PGE_1) maintains ductal patency until definitive decisions regarding surgical correction can be made. The main side effects of this medication are flushing, fever and possibly apnoea.

Alternatively, babies with duct-dependent systemic circulation usually due to some structural problem with left ventricular outflow (e.g. critical aortic stenosis, severe coarctation, hypoplastic left heart syndrome) may present in the first few days of life with heart failure and poor peripheral perfusion triggered by ductal closure. The most reliable physical signs of heart failure are tachycardia, tachypnoea and hepatomegaly. If shock is present (poor pulse volume, pallor, altered conscious state) respiratory support and fluid volume expansion may be required to restore the circulation. Subsequently, alprostadil or inotrope infusion and continuing judicious fluid administration may be required. The most important steps in restoring systemic circulation in this situation are artificial ventilation to normocapnia, together with re-establishing and maintaining ductal patency (see Chapter 5.5).

Post-resuscitation stabilisation

Post-resuscitation stabilisation should be directed towards preventing any ongoing or repeated primary insults (primarily to the brain) as well as limiting any secondary injury and organising a stable transfer to an appropriate neonatal unit:

- Artificial ventilation should be continued if required to maintain normocapnia (pCO_2 35–40 mmHg).
- Oxygenation should be optimised (aiming for pO_2 60–90 mmHg,

although this target may be set higher in the face of documented pulmonary hypertension).
- ET tube migration or obstruction should be vigorously avoided.
- With regard to control of body temperature while maintenance of normothermia remains the primary goal in infants less than 35 weeks' gestation, cooling of normally formed asphyxiated term and near-term infants is recommended. Previous studies support cooling to 33.5°C for 72 hours infants who either have ongoing need for respiratory support at 10 mins, Apgar score ≤5 at 10 mins or cord pH <7 and clinical signs consistent with moderate or severe encephalopathy.[5]
- Blood pressure should be monitored meticulously and hypotension treated promptly with fluid resuscitation or inotropes as required.
- Close monitoring of body weight, fluid balance, electrolytes, calcium and magnesium is indicated.
- Evidence of hypoxic insult to major organs other than the brain should be sought (urinalysis, serum creatinine, liver function tests, serum troponin level, coagulation parameters).
- Both hyper- and hypoglycaemia may aggravate injury following hypoxic ischaemic insult, and therefore normoglycaemia should be targeted.
- There is no evidence in neonatal post-resuscitation cerebral care for hypertonic or osmotic agents to treat or minimise cerebral oedema. The same applies to glucocorticoids and prophylactic anticonvulsant treatment.
- Liaison with and transfer to a neonatal intensive care unit (NICU) should be organised as soon as possible with clear documentation of the resuscitation interventions required and responses achieved.
- The parents should be kept well informed of the infant's condition and efforts made for them to have contact with the baby.

Controversies

- ***Respiratory function monitoring during resuscitation.*** As stated above assisted breathing can be delivered effectively at birth with several possible devices, each of which has advantages and disadvantages: maintaining CPAP is difficult with a self-inflating bag and inadvertently high peak pressures and tidal volumes frequently occur whereas safe use of a flow-inflating bag technically requires more expertise. T-piece

devices consistently target peak inflation pressures and longer inspiratory times; however, inadvertent excess pressure and air trapping can occur. Data clearly indicate that excessive inflation pressure is injurious to lungs, and accordingly some have suggested that rather than pressure, flow and delivered tidal volume should be continuously monitored and targeted. Such easily applicable bedside flow and volume monitoring devices are now available for use during resuscitation. Further research is required to determine the best interface and ventilation device to use in each situation and the role of bedside monitoring in improving response to and outcome of neonatal resuscitation.

- ***Hypothermia in resource-limited settings.*** There is no question that neonatal mortality rates are higher and the need for effective resuscitation and ongoing care are greater in developing countries. Studies to date confirming the effectiveness of therapeutic hypothermia in term and near-term newborns following asphyxia have generally been done in developed countries where available resources and systems have permitted the therapy to be administered under a strict protocol. It remains unclear if these findings and resulting recommendation can be extrapolated to resource-limited environments. Further adequately randomised controlled trials of simple methods of cooling in such settings are required to answer this question.
- ***Ethics of resuscitation.*** The decision to withdraw or withhold resuscitation from extremely premature newborns or those with severe congenital abnormalities requires clear communication with the family and accurate clinical data. Overall there is agreement that overly aggressive treatment is to be discouraged.[1]

Prognosis

Predicting outcome at an early stage may be difficult, but the most reliable early predictors of adverse outcome are abnormalities in the clinical examination (i.e. degree of encephalopathy) and electroencephalographic assessment. A sustained low-voltage EEG or discontinuous activity on EEG within 6 hours of birth is strongly predictive of death or significant adverse neurological sequelae.

Recent studies demonstrate extremely poor survival rates for infants less than 23 weeks' gestation (<1%) with a better but still poor rate for infants admitted to

the NICU (5%). Mortality is less at 23, 24 and 25 completed weeks (11%, 26% and 44% survival in one large recent study). However, severe disability in childhood is present in 20–30% of survivors at these gestations, and some disability can be expected in 50%.[6] Prognosis is best predicted by gestational age alone (if accurately known) rather than weight.

Non-initiation of resuscitation in the delivery room is appropriate for infants with confirmed gestation less than 23 weeks or birth weight less than 400 g, anencephaly or confirmed trisomy 13 or 18. Options include a trial of resuscitation, non-initiation or discontinuation after further assessment. Initiation of resuscitation does not mandate continued support.

International guidelines state that discontinuation of resuscitative efforts may be appropriate if resuscitation of a newborn infant with cardiorespiratory arrest continues to have an undetectable heart rate (Apgar score 0) at 10 minutes.

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SECTION 4

Trauma in Children

OUTLINE

4.1

Introduction to paediatric trauma

Dermot Thomas McDowell, and Soundappan S.V. Soundappan

ESSENTIALS

1 Trauma is the prime cause of death and serious injury throughout childhood, accounting for 40% of deaths in developed countries.

2 Motor vehicle accidents account for almost 50% of unintentional injury-related deaths.

3 Almost two-thirds of injury-related deaths in children are male.

4 Prevention strategies have resulted in most of the improvement in mortality during childhood.

5 The advent of trauma teams has led to great improvement in paediatric injury care.

6 Delayed management of airway obstruction and inadequate fluid management are the two most common contributors to preventable paediatric deaths in trauma.

7 The initial assessment of the seriously injured child should follow a structured approach so that life-threatening problems are rapidly identified and managed in rapid sequence.

8 The team leader needs to coordinate the resuscitation of the child with multisystem trauma so that patient care is at all times expeditious and tailored to the specific and prioritised needs of the child.

9 It is important to provide early psychological support to other family members who arrive with the child with major injuries.

10 One in ten children aged 19 years or younger will be seen in an

emergency department for a non-fatal unintentional injury.

Prevalence

Overall, trauma is the number three cause of mortality (6%) and serious morbidity throughout life. However, trauma is the leading cause of death and disability between the ages of 1 and 44 years. Therefore it is the prime cause of death and serious injury throughout childhood – rendering it the most important health issue among children and adolescents. In most Western societies, road trauma contributes about half (50%) of all serious injuries and deaths, with drowning incidents contributing up to 25%. The third major cause is burns, and the remainder includes a range of miscellaneous causes. In most series, child abuse contributes less than 10% of all paediatric deaths after injury.

This chapter concentrates on those aspects of trauma management that are different in children. Overall, paediatric surgeons and paediatricians have taken a holistic view towards assessment and management of injured children and have been heavily involved in aspects of prevention, immediate treatment and rehabilitation.

Prevention

Over the past decades, the death rate among children in Australia has reduced from approximately 22.3 deaths per hundred thousand in the 1970s to about 9.5 in 1990s and 5 in 2012.[1,2] However, while the death rate has reduced significantly, it is still higher than that reported in some of the best Organisation for Economic Co-operation and Development (OECD) countries. Unfortunately such reductions have not been observed in developing countries (unintentional injury death is 10 times higher in the continent of Africa than in high-income countries).[3] While developed countries have achieved much through prevention strategies, there is still much to be done throughout the world.

Prevention has involved the work of legislators (seat belts, baby capsules, bicycle helmets) through to educators and implementation groups such as ‘Kidsafe’ and ‘Safe kids worldwide’.[4,5] Community involvement has been vital in the success of these prevention initiatives in spite of the minor inconveniences that accompany improvements in child safety.

Table 4.1.1

Major trauma predictors at high risk of life-threatening injury

Mechanism	Injury
Motorcyclist, bicyclist, or pedestrian impact at >30 kph	Serious or suspected serious blunt or penetrating injuries to head, neck or torso
Crash speed >60 kph	Significant injury to two or more body areas
Ejection from vehicle	Burns ≥20%
Vehicle rollover	Two or more long bone fractures
Fall greater than 3 m or more than twice the child's height	Evisceration, severe crush injury, amputation, suspected spinal injury, pelvic fracture
Fatality in same vehicle	***Treatment***
Explosion and suspected inhalational burns	Intubation
Extrication >30 minutes	Any airway manoeuvre at any time
Signs	Assisted ventilation
Respiratory rate<10 or >30 per minute	Chest decompression
Systolic blood pressure <75 mmHg	Failure to control external bleeding
GCS <15	All inter-hospital trauma transfers
Oxygen saturation <90%	Significant comorbidity

Adapted from Cameron P. *Textbook of adult emergency medicine*, 4th edn. London: Elsevier; 2014.

Future progress will depend on campaigns refreshing important preventive messages, every few years for new generations of young parents, and it cannot be assumed that a good campaign 3 to 5 years previously will sustain its effectiveness. Concerns about risk should be balanced against adverse effects of sedentary recreational activities such as television viewing or computer gaming.

Succinct treatment (salvage)

The advent of trauma teams and trauma systems in hospitals that receive paediatric patients has led to great improvements in paediatric injury care. In one study the improvement in preventable paediatric trauma deaths from 21% to 7% after the implementation of trauma system changes in Ontario, Canada.[6]

A trauma team refers to a multidisciplinary group of health professionals who can provide rapid, expert assessment, resuscitation and initial treatment of a patient with multiple injuries. Hospitals now have trauma teams ready to receive the child. This may occur by forward notification from the emergency

management services. Trauma team activation should occur on notification when a child is at high risk of life-threatening injury according to prediction by predetermined clinical and mechanism parameters (Table 4.1.1).

The trauma team should include at minimum a team leader, airway doctor, procedure doctor, and nursing staff from within the emergency department (ED). Activation may involve alerting appropriate colleagues from radiology, anaesthetics, intensive care and surgical specialties according to local resources and protocols, in order to expedite emergent care. The activation process needs to be adapted to the varying local resources of the individual department, which vary between institutions.

Clinicians often worry about managing children with major trauma and wonder how much they should treat them as adults and how much they should take note of their differences. In adults it is well established that the *A, B, C, D, E primary survey* approach is the correct paradigm. The approach should be similar to adults, but there are additional nuances in children to optimise their care. Similar to adults, delayed management of airway control/chest trauma and inadequate fluid management are the two most common contributors to preventable paediatric deaths in trauma.[7–10] Chapters 2.2 and 2.3 provide a detailed discussion of basic life support and advanced life support (ALS) techniques in children applicable in trauma.

Regular trauma meetings to review cases or videotape evaluation of resuscitation can provide education, with lessons learnt on ‘how to do things better’. As major paediatric trauma may be relatively uncommon in some centres, mock paediatric resuscitation scenarios can provide the ED staff with an opportunity to improve preparedness.

Primary survey

The initial assessment of the seriously injured child should follow a structured approach so that life-threatening problems are rapidly identified and managed in the appropriate rapid sequence. This approach is based on the prioritised principles as outlined in the teaching dictums of courses such as the Advanced Paediatric Life Support (APLS) and Advanced Trauma Life Support (ATLS) groups, which are invaluable for practitioners who deal with paediatric trauma.

The approach includes initially the **primary survey** with opening and securing of (A) the airway, (B) optimising breathing and (C) circulation and immediate management of threats to life. The stabilisation of the cervical spine

occurs concurrently with airway management. Following this rapid initial stabilisation, a brief neurological assessment of the child should occur (D) with complete exposure for otherwise occult signs of injury and a prompt to consider environmental issues such as hypothermia (E).

The algorithm for the primary survey has recently been altered by the APLS group (not ATLS). The A, B, C, D, E approach is now preceded by (control of catastrophic external haemorrhage). This is done with judicious use of tourniquets and, where available, direct pressure haemostatic dressings following military experience.[11]

In the team approach, the management should 'occur horizontally', with simultaneous attention to these priorities by designated members of the team, overseen by the team leader. The role of the primary survey is therefore to detect and treat abnormal physiology immediately in order to prevent potential secondary insults due to hypoxia or hypovolaemia. A secondary survey follows with a head to toe, front and back examination of the child.

Paediatric differences

Catastrophic external haemorrhage control

Control of external exsanguinating haemorrhage becomes the immediate priority. A child's circulating blood volume is significantly less than that of an adult. Thus external haemorrhage can become catastrophic earlier than in adults. Haemorrhage is responsible for 30–40% of trauma mortality.[1] The use of tourniquets to an extremity can be effective in controlling external bleeding and should be used where application of direct pressure fails to be effective. It must be remembered that the use of tourniquets does carry a risk of ischaemic injury, and this should be considered during prolonged use.

A. Airway and cervical spine control

In the setting of trauma, 'A' refers to securing of the child's airway with concurrent attention to the possibility of an unstable cervical injury. The cervical spine should be immobilised using an appropriate-sized hard collar or sandbags, tape and a spinal board where appropriate. It may be helpful in an infant to place a small towel under the space in the shoulder region caused by 'the elevation off the bed' by the prominent occiput at this age. As discussed in the ALS section in Chapter 2.3, the characteristics of the paediatric airway make it more vulnerable

to obstruction. This can be exacerbated by the necessary supine positioning of the child on the resuscitation trolley. Due to comparatively higher oxygen demands and less reserve, a child manifests hypoxic decompensation earlier than adults. This reinforces the increased importance of airway management in achieving adequate ventilation. The initial airway-opening manoeuvre in trauma patients should be by the jaw thrust technique while maintaining cervical spine immobilisation. Suctioning of any oropharyngeal soiling from blood, vomit, teeth or other foreign bodies may be necessary. Adjuncts such as the placement of an oropharyngeal airway may be required. In children, these should be placed into the oral cavity directly, using gentle depression of the tongue with a spatula to allow for atraumatic positioning. Nasopharyngeal airways are not recommended due to the potential presence of cribriform plate fractures, and trauma to the turbinates may cause bleeding that may further complicate airway patency.

Definitive airway intervention should occur in the child who is apnoeic (usually coma related), has persistent airway obstruction despite the above manoeuvres, is requiring bag–mask ventilation to achieve oxygenation, has respiratory insufficiency from major chest injury, has significant ongoing bleeding due to facial trauma compromising the airway or has high risk of subsequent compromise such as from an airway burn.

The technique of intubation should be rapid sequence with cricoid pressure. The control of the cervical spine during intubation should occur by manual immobilisation by a dedicated assistant. The patient should have continuous monitoring, preoxygenation and difficult airway adjuncts available, if required. The choice and dosage of the sedation agent (thiopentone, midazolam, propofol, or fentanyl, for example) may be individualised according to factors such as the level of coma, cardiovascular status of the child, presence of other injuries and operator experience. A rapidly and short-acting muscle relaxant such as suxamethonium usually provides the most optimal intubation conditions, despite the potential for transient increase in intracranial and intraocular pressures through fasciculations. Children less than 2 years of age are prone to significant bradycardia on laryngoscopy, which may be blunted by the administration of atropine, prior to intubation. Confirmation of correct ETT placement should occur according to the methods described in Chapter 2.3. One needs to be prepared for the possibility of the difficult airway or failed intubation and have a planned algorithm to deal with this possibility. It is not the failed intubation that causes the problem but the lack of planned action to alternatively oxygenate the

patient in this situation.

B. Breathing and high flow oxygen

All children with severe trauma should receive high flow oxygen (10–15 L min) by a reservoir face mask, independent of the need determined by saturation monitoring. Inadequate spontaneous ventilation is supported by bag and mask assistance. If this need is ongoing, intubation is required. A rapid screen for life threats, such as a tension pneumothorax, should occur. This may require immediate decompression by needle aspiration for the child in extremis and a subsequent formal chest drain, as described in the section on chest trauma (see Chapters 24.8 and 24.9). The possibility of developing tension due to a pneumothorax should be borne in mind as one of the potential precipitants of rapid deterioration following the initiation of positive pressure ventilation.

C. Circulation and stop haemorrhage

It is important to be aware that a child may be profoundly shocked from blood loss resulting from trauma well before the occurrence of hypotension. A child responds to hypovolaemia with tachycardia and increasing peripheral vascular resistance. Therefore the assessment of circulation status needs to be focused on the heart rate, pulse volume and the parameters of skin perfusion such as capillary refill time, colour and temperature. Perfusion inadequacy should be proactively corrected with volume resuscitation, rather than waiting for hypotension as an indicator. Hypotension occurs late due to cardiac decompensation and indicates that a child is nearing collapse. Likewise, bradycardia is often a prelude to imminent arrest. The persistence of tachycardia in the child with trauma should prompt concern and evaluation for occult ongoing blood loss.

The child with major trauma requires urgent intravenous access with the largest practical cannulae into visible or palpable peripheral veins. In the shocked child, cephalic, femoral or great saphenous veins are usually the most accessible. Where vascular cannulation is unsuccessful after 60–90 seconds, the child who requires immediate fluids or drugs to facilitate intubation should have a rapid intraosseous needle placed, as described in Chapter 24.12. This is the second-line method that should be used in children without hesitation and is increasingly applicable in adults. In cases with clinical signs of obvious major

abdominal, pelvic or lower-limb trauma, it may be prudent to use additional alternative access into a vein that drains into the superior vena cava (e.g. external jugular or subclavian). In such rare situations, lower-limb intraosseous-infused volume may not access the circulation effectively due to disruption of the normal continuity of the intraosseous and intravascular spaces.

Fluid resuscitation should initially be with crystalloid or colloid titrated to clinical response. Boluses of 20 mL kg should be given using pressure infusion. Bleeding in an uncontrolled trauma setting predisposes the child to the lethal triad from rapid blood loss, hypothermia, and acidosis, resulting in coagulopathy bleeding. All centres expected to receive severely injured patients should have massive transfusion protocols in place. Massive transfusion in the paediatric population is commonly defined as the transfusion of blood components equaling one or more blood volumes within a 24-hour time frame or half of a blood volume in 12 hours. Improved outcomes have been associated with higher ratios of fresh frozen plasma (FFP) to transfused packed red blood cells (PRBCs) and platelets than previously recommended. It is the intent of MTPs to transfuse to a FFP/PRBC ratio of 1:1; however, the actual transfusion ratios can be considerably less and often delayed. Adjuncts to massive transfusion should be considered including cryoprecipitate/fibrinogen, when fibrinogen levels are low, and calcium.

Part of the circulation phase of management includes therapy to limit ongoing blood losses from external or fracture sites by direct pressure or splinting. This is of vital importance as a child's blood volume is only 80 mL kg^{-1}, and small ongoing losses contribute to haemodynamic instability.

The safety and efficiency of tranexamic acid in the injured paediatric population remain questionable. Limited evidence on the topic exists and in the military setting, tranexamic acid has been reported to be used in about 10% of cases, most with severe abdominal and extremity trauma. The use of tranexamic acid was independently associated with decreased mortality and no adverse safety or medication-related complications (i.e. cardiovascular or other thromboembolic phenomena) were observed.[12]

Although in major trauma the most likely cause of shock is blood loss, one needs to consider other possible contributors such as myocardial injury, pericardial tamponade, spinal shock or tension pneumothorax.

The benefits of the MTP may extend beyond the hemorrhaging child with the MTP improving viability of organ donation.

D. Disability

The most rapid way to assess a child's conscious level in the primary survey is by using the AVPU scale (A – alert, V – responds to voice, P – responds to pain only, U – unresponsive).

A child who scores a non-purposeful P or U correlates with a GCS of 8 or less and requires intubation and ventilation. The brief neurological assessment during the primary survey is completed by checking equality of pupil size and reactivity to light.

E. Exposure and environment

The child should be undressed fully to allow examination of the entire body in order to detect all external stigmata of injury. Once this is completed, the child should be covered by warm blankets. The potential for hypothermia can also be decreased by the use of radiant heaters and warm resuscitation fluids.

Other issues during initial stabilisation

Early analgesia

Children who have significant pain or distress need to have prompt provision of analgesia as soon as intravenous access is obtained. This is usually best achieved by titrating intravenous morphine in doses of 0.1–0.2 mg kg^{-1} to effect. Adequate analgesia allows a more rapid and reliable clinical assessment.

Continuous monitoring

This should include heart rate, blood pressure, respiratory rate, oxygen saturation and clinical assessment of perfusion.

Psychological support of child and family members

It is important to provide early psychological support to the child to help with assessment and relieve his/her anxiety. Presence of parents or family members is not uncommon in paediatric resus bays to achieve this. Play therapists are increasingly used in resus bays to help calm distressed children.

It is also important to support other family members who arrive with the child with major injuries. This should occur via a senior member of the team assisted by a nurse or social worker who is dedicated to this role. Non-comatose children are often fearful and distressed, and it is useful to have the parents available to provide comfort to them.

Secondary survey

The secondary survey follows when initial resuscitation has stabilised the child from immediate life threats. Continuous monitoring of vital signs and neurological status is paramount, as any deterioration should prompt immediate discontinuing of this phase of the assessment and return to the primary survey. The secondary survey should include the following:

- **History** – from ambulance staff, parents and child where possible:
  A: Allergies
  M: Medications
  P: Past history
  L: Last ate
  E: Event – in order to clarify mechanism of trauma and potential for injury. The more detail in the history the more accurate the assessment of potential injuries.
- **Head to toe, front and back examination** – the child should have a methodical and thorough clinical examination to identify all injuries. The head, face, neck, chest, abdomen, pelvis, spine and extremities (see orthopaedic trauma below) need to be examined. A more formal neurological assessment should use a paediatric Glasgow Coma Score (GCS). The spinal immobilisation needs to be maintained until clinical or radiological clearance, in cases where this is possible. During this phase, placement of an orogastric tube may occur if gastric decompression is indicated. A urinary catheter (with the standard precautions for a potential urethral injury) is indicated in the unconscious child or where the accurate measurement of urine flow is required.
- **Investigations** – appropriate bloods should be sent on initial intravenous access, along with a bedside glucose measurement. In the multiply injured child, initial radiological films should potentially include a

resuscitation room chest X-ray, lateral cervical spine and pelvis. A 12-lead ECG should be done where a child has sustained significant trauma to the chest.

Definitive care and disposition

After the secondary survey and reassessment of the child's physiological response to resuscitation, the next step is to prioritise injuries to determine the need and timing of further imaging or any surgical intervention. In the non-paediatric tertiary environment, early liaison with appropriate colleagues from a trauma centre with the capacity for definitive care of a child with major injuries will help determine the most appropriate means for the individual child optimally to reach definitive care. Transfer of a child to definitive care is discussed in Section 27.

Orthopaedic trauma

A discussion of specific injuries and their management is dealt with in detail in Section 25.

Bony injuries are common in children who have multiple trauma. Once the child is stable, the secondary survey must include assessment of all limbs and clavicles, as well as ribs, pelvis and spine. The splinting of long bone fractures before moving the patient from the resuscitation room is important, even in the face of other serious injuries. This avoids ongoing pain and blood loss as well as further soft tissue injury to the limb.

A careful tertiary survey the following day may reveal minor bony injuries, which were not detected during the initial resuscitation phase. These injuries need to be immobilised and, if necessary, a plan made for reduction as soon as the general condition of the child allows. Once the life-threatening injuries heal, a missed displaced bony injury is a major problem for the child in the longer term.

Isolated orthopaedic trauma is one of the most common types of trauma presenting to an ED. Review of history and mechanism of injury and an overall examination are needed to exclude concurrent injury. The possibility of a non-accidental injury must always be considered. The time of last oral intake should be documented, and the child should receive nothing orally until the management plan is established.

In order to avoid causing further pain to a child, much of the examination of the fracture area can be done by observation:

- Lack of spontaneous movement gives a clue to the location of a painful injury.
- Deformity or angulation, swelling and bruising, or breach of the overlying skin suggests an open fracture.
- Gently eliciting localised areas of tenderness helps direct the most efficient use of radiology to identify the site of injury. It is best to exclude non-tender regions first. A cooperative child can point to the area of maximal tenderness, whereas in a younger child the parent may be able to indicate which areas cause the child to be apprehensive.
- The function and range of motion of surrounding joints should always be checked.
- Vascular complications can be assessed by colour, perfusion and pulses of the distal limb. As pain often limits motor function, neurological assessment to test sensory components is important.
- Compartment syndrome should be looked for when a displaced fracture has significant associated swelling. In children the volar surface of the forearm is frequently the site involved. Pain on passive stretch of involved muscles is the hallmark. Distal pulses may be preserved. Neurological dysfunction often develops. Some fractures are at high risk for compartment syndrome, and the treating team should be aware of and monitor for the same.

Pain relief involves first immobilising the limb with a sling or a temporary backslab or seating the child in a wheelchair or bed. The need for medication for pain relief can then be assessed. Analgesia is required before radiological investigation so that any limb movement necessary to take the appropriate films is less distressing for the child.

Plaster immobilisation in the ED should generally involve using a backslab technique, rather than a circumferential plaster because of the potential for early limb swelling. The slab should be of sufficient strength to keep the fracture stable. Plaster of Paris strips usually require 6–8 layers for the upper limb and 10–12 layers for the lower limb. For displaced fractures an initial temporary plaster slab can reduce movement and therefore pain at the fracture site during the X-ray. The soft padding can be used to line the plaster slab, rather than

winding it around the limb to reduce movement on application.

The reduction of any displaced fractures should be done with appropriate analgesia, anaesthesia and sedation, which should be administered by a second doctor. Often this is best achieved with a general anaesthetic where there is significant need for manipulation or fracture instability. The reduction of any fracture in children should be done by a doctor with the appropriate training and experience so that multiple attempts and further injury are avoided. Complicated fractures and those in which the reduction will be hard to maintain require additional expertise.

Appropriate orthopaedic follow-up of even minor fractures in children is vital. Within 5–7 days the swelling subsides, and the backslab does not hold the fracture, which may lose position. At that stage a circumferential plaster can usually be used. Because the child frequently falls, a change in fracture position can easily occur. A repeat X-ray at review is necessary to determine if the fracture has maintained position.

Rehabilitation

Rehabilitation should start from the time of admission, and there should be early involvement of the appropriate teams. There have been major improvements in the diagnosis and care of cervical spine injuries and management of severe head injuries. There needs to be continuing work on the outcome of minor head injuries and the debilitating long-term consequences of serious long bone fractures.

Finally, the child's social situation and propensity to partake in risky behaviour need to be assessed, as there is some evidence that children with injuries tend to have further future injuries. Careful assessments of the child's social history may allow appropriate interventions. A full 'tertiary history' should include environmental factors (type of car/airbags/seat belts/helmets/details of window falls/pool fence gates, etc.) so as to allow future prevention programmes to be appropriately targeted.

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4.2

Paediatric neurotrauma

Soundappan S.V. Soundappan, and Dermot Thomas McDowell

ESSENTIALS

1 Neurotrauma is the leading cause of morbidity and mortality following injury.

2 A careful history and examination are important to assess the potential significance of the injury and determine the need for further investigation or admission for observation.

3 Clinicians should not attribute the cause of shock to head injury until other causes have been excluded. Infants may become hypovolaemic secondary to blood loss from a scalp laceration or intracranial bleed.

4 Any child with a Glasgow Coma Scale (GCS) ≤14, clinical evidence of a skull fracture or penetrating skull injury should have an emergent head CT scan.

5 The prime role of trauma resuscitation is to rapidly identify and correct hypoxia and hypotension in children with severe traumatic brain injury (TBI), as they are significant contributors to secondary brain injury.

6 Rapid sequence intubation, with in-line stabilisation of the cervical spine, is the preferred technique for securing the airway in the head-injured child.

7 Children sent home from the emergency department (ED) following a head injury must be discharged to the care of a responsible adult who is given clear discharge instructions and a written head-injury sheet containing advice on when to seek review.

8 The family of the child with a significant head injury requires appropriate support and explanation as an important facet of care whilst in the ED.

Introduction

Paediatric neurotrauma is a common presenting problem in emergency medicine practice. This chapter will deal principally with paediatric traumatic brain injury (TBI) and spinal cord injury. Spinal bony and ligamentous injuries are also covered in Chapter 24.4.

Epidemiology

TBI covers a spectrum of injury from trivial to lethal. It is the leading cause of morbidity and mortality in paediatric trauma.[1] In 2004–2005, of the 21,800 TBI admissions in Australia, 3700 were children under 14 years old. Males accounted for two-thirds of the admissions.[2] In Australia the most common cause of TBI is falls, followed by motor vehicle-related accidents, intentional injury by others and unintentional injuries.[2,3] It is important to remember that these figures only reflect hospital admissions, and the true incidence of TBI in the community is much higher. The overall incidence of spinal cord injury in Australia for children aged 0 to 14 years is unknown but significantly less than the adult population.[4,5] The age standardised incidence in the 15+-year-old population is 14 per million of population per year, with a male to female ratio averaging 4:1 but peaking at 9:1 in the 15–24-year age group.[3]

Preventive strategies have made the most impact on the improved outcomes of paediatric trauma over the past two decades.

Pathophysiology

TBI, like spinal injury, can be divided into primary and secondary trauma. Primary trauma occurs during the initial impact to the head, and only preventive measures such as using protective equipment (e.g. helmets and seat belts), better engineering (e.g. safer roads), education and legislation can alter the extent of this primary injury. There is necrosis of neurons with release of intracellular contents leading to inflammation. The impact causes fractures and bleeds in

various levels. Secondary trauma or insult occurs when post-traumatic acute phase response and mediators, or subsequent physiological insults, such as hypoxia, hypotension and increased intracranial pressure, occur and cause further damage to the already traumatised tissues or structures.[6] There is disturbance of the balance between vasoconstricting and dilating factors leading to oedema and ischaemia. The prevention of secondary injury, particularly due to cerebral hypoxia and reduced cerebral perfusion, is the primary focus of acute medical intervention, which begins pre-hospital and continues in the hospital setting.[6]

Children have unique anatomical, physiological and developmental differences when compared to adults. They have a large head-to-body ratio, leading to a high centre of gravity (falls) and to the head being the primary 'target' for trauma. The skull is thinner and more plastic and thus transmits rather than attenuates impact.[7,8] Skull fractures are therefore more common in children, and, importantly, serious brain injury can occur without an associated skull fracture.[9–13]

In children, the dura is more closely adherent to the skull when compared to adults, making extradural haematomas less common in children, particularly in infants.[8,14] Unfused sutures and an open fontanelle can expand to accommodate intracranial haemorrhage or cerebral oedema.[8] Some authors have thought children to be more prone to 'malignant cerebral swelling' that can cause rapid and sometimes fatal deterioration even after minor TBI.[9,15,16] However, this view has recently been questioned, and swelling may be no more common in children than in adults.[17]

High brain water content with varying degrees of myelination in different areas predispose to acceleration-deceleration injuries and shearing forces. Physiologically, children have a lower systolic and mean arterial blood pressure, which implies a lower cerebral perfusion pressure (CPP). This, in turn, may cause problems with maintaining adequate cerebral perfusion if they have raised intracranial pressure. Infants and small children may become hypovolaemic with large intracranial bleeds. This is not seen in larger children or adults.[7]

Cerebral blood flow (CBF) is often very low and may approach ischaemic levels following more severe TBI.[18] This may be related to a low brain metabolic rate in comatose patients, increased intracranial pressure (ICP) and vasospasm. Autoregulation of CBF may be lost following TBI, and in this setting CPP largely determines CBF. This underscores the importance of maintaining an adequate CPP in the head-injured patient, especially those with more severe

injuries. When the ICP exceeds 20 mmHg due to increase in volume of blood, cerebrospinal fluid (CSF) or brain matter, herniation occurs. This can occur at supratentorial, tentorium cerebelli, falx cerebri or formaen magnum sites with different consequences.[6]

Head injury, as opposed to TBI, may be described as extra-axial or intra-axial. Extra-axial injury refers to pathology outside the brain parenchyma.[8,19] Extra-axial structures include the skull, structures between the skull and brain and the ventricular spaces within the brain. Common extra-axial lesions include skull fractures and extradural, subdural, subarachnoid and intraventricular haemorrhages. Extradural haemorrhage occurs, as its name implies, outside the dura. Medical literature often refers to this as 'epidural haemorrhage', but in Australasia the term 'epidural' is generally used only to describe lesions outside the dura of the spinal cord. Intra-axial injuries are true TBIs and include contusion, laceration, haemorrhage and diffuse axonal injury (DAI). DAI may result in considerable disability with little to see on radiological investigation.[8]

Classification

The generally accepted method of classifying severity of TBI is by using the Glasgow Coma Score (GCS), although other measures such as duration of unconsciousness or amnesia are sometimes used. The GCS was first described for adults in 1974 and scores three variables: eye opening, verbal response and motor response (see 'Examination' and Table 4.2.1).[20] It has proved to be a very useful tool in rating severity of TBI and prognosis.[8]

The problem with using the GCS on young children, particularly those aged less than 2 years, is that the best verbal response is limited by their language development. In an attempt to overcome this difficulty, modified GCSs have been proposed, including the so-called Child Coma Scale (CCS).[9,21] It is important to note that, unlike the GCS, the CCS has never been adequately validated, and many studies of head injury in children deliberately exclude those aged less than 2 years.

TBI is usually divided into three categories: mild, moderate and severe.

Mild traumatic brain injury (GCS 14 to 15)

Mild TBI was originally defined as head trauma patients with a GCS from 13 to 15 (and/or varying periods of loss of consciousness [LOC] and amnesia).[22] The

problem with this definition is that patients with GCS 13 have a significantly higher risk of intracranial injury, with subsequent risks for deterioration and neurosurgery, than patients with a GCS of 14 or 15. They more properly belong in the moderate head injury group.[15,23]

Table 4.2.1

Adult and child Glasgow Coma Scores (GCS)

Score	Adult	Child
Eye opening		
4	Spontaneous	
3	To speech only	Same
2	To pain only	
1	No response	
Verbal		
5	Orientated in person, place and time	Happy/smiles/interacts normally
4	Confused	Crying but consolable
3	Inappropriate but intelligible speech	Inconsistently consolable
2	Incomprehensible sounds	Inconsolable and/or irritable
1	No response	No response
Motor		
6	Obeys commands	
5	Localises painful stimulus	
4	Withdraws to painful stimulus	Same
3	Flexor posturing to painful stimulus	
2	Extensor posturing to painful stimulus	
1	No response	

The original definition of GCS 13 to 15 continues to be used in international and Australasian literature, but the recognised definition of mild TBI in Australasia is GCS of 14 or 15.[16] Some authors believe that even this is too liberal, and the definition of mild TBI should be restricted to patients with a GCS of 15.[23,24]

Approximately 80% or more of children with TBI will fall into this category.[25] The reported incidence of intracranial haemorrhage (ICH) varies between 4% and 7% in children with GCS 15 and increases to approximately 10% in children with GCS 14.[11,15,26,27] The overall mortality in this group is reported to be as high as 2%.[15] These figures may be subject to significant selection bias.

The terms **‘minimal’** or **‘trivial’ TBI** are sometimes used to describe a subgroup of mild TBI who meet the following criteria: GCS 15, normal neurological examination and no signs of a skull fracture.[24,26,28] Transient LOC or amnesia does not exclude patients from this subgroup.

Moderate traumatic brain injury (GCS 9 to 13)

Approximately 18% of children with TBI fall into this category, although this is likely an over-estimate.[25] The incidence of ICH and overall mortality in this

group of children is uncertain (most research focuses on either mild or severe TBI).

Severe traumatic brain injury (GCS 8 or less)

Approximately 2% of children with TBI fall into this category.[25] Patients in this category are, by definition, comatose. Overall mortality in this group is between 30 and 40%.[29]

Assessment

History

Careful history and examination are important when assessing the significance of the injury and determining the need for further investigation or admission.

Initial assessment and treatment may need to occur simultaneously. Ensuring an adequate airway, breathing and circulation with resuscitation as required is the first priority. Following the early management of severe trauma (EMST) principles of primary and secondary survey similar to any other trauma patient is the best approach. Spinal precautions must be maintained until clinical and/or radiological clearance of the spine has been completed.

Assessment of the child may be extremely difficult, if not impossible, when the child is distressed with pain or is cerebrally irritated. The early use of a very small dose of parenteral narcotic such as morphine 0.05 mg kg^{-1} intravenously may make assessment and management much easier. Concern about masking changes in neurological function should not prevent the use of adequate analgesia for children who are distressed by pain.

The following **historical information** is relevant and should be ascertained if possible:

- Time and mechanism of the injury
- Any loss of consciousness or period of altered level of consciousness and its duration
- Any post-trauma seizure-type activity
- Any post-injury vomiting
- Any headache or other neurological symptoms such as diplopia, weakness or altered sensation
- Any retro- or anterograde amnesia

- Progression of any symptoms and signs from the time of injury
- Details of pre-hospital care assessment and therapies.

The possibility of non-accidental injury (NAI) must always be considered in children with skull fractures or intracranial injuries. Be particularly vigilant with children aged <2 years, when a parent or caretaker has delayed seeking medical care or the stated mechanism is not in keeping with degree of injury observed.[13]

Past history that is particularly relevant in the context of neurotrauma is:

- Previous head or spinal injury
- Congenital central nervous system (CNS) problems
- CNS surgery (such as the insertion of a ventriculo-peritoneal [VP] shunt)
- Bleeding diatheses such as haemophilia or thrombocytopenia
- Psychomotor or developmental problems, such as autism, can make assessment particularly difficult.

It is important to remember:

- Medications and other ingestants that may influence assessment of a head injury including possible illicit drugs or medications and chemicals to which a young child may have gained access at home
- Allergies
- Immunisation status
- Last food or fluid intake.

Examination

Check vital signs, including bedside blood sugar level, to immediately exclude the following:

- Hypoxia (SaO_2 ≤90% or PaO_2 ≤60 mmHg)
- Hypotension (systolic blood pressure [SBP] ≤5th percentile for age or SBP <90 mmHg)
- Hypoglycaemia (blood sugar level <3.0 mmol L^{-1}).

These reversible factors may be contributing to the child's altered level of consciousness and also cause secondary brain injury. Ascertain the child's

weight, early, either by measurement or inference from the child's age, as it is necessary for calculating drug and fluid requirements.

Table 4.2.2

AVPU scale

A	**Alert**	
V	Responds to voice	
P	Responds to pain	Purposeful Non-purposeful
U	Unresponsive	

Glasgow Coma Scale

This should be used to define the child's level of consciousness and to monitor any change over time. Serial neurological observations should be performed regularly to monitor for deterioration or improvement in the child's conscious state. Once a child's GCS is normal and stable, the frequency of neurological observations can be decreased. It is important to note that the GCS is intended to score global function not focal deficit. When using the GCS to classify the severity of TBI, the best post-resuscitation score should be used.

If a painful stimulus is needed to test motor function, apply pressure to the supra-orbital margin to test for localisation of pain. This is superior to a 'sternal rub'. To test for withdrawal or abnormal flexion/extension, use a pen or pencil to apply pressure to fingernail or toenail beds. Abnormal flexion is usually termed 'decorticate posturing' and abnormal extension termed 'decerebrate posturing', although this was discouraged in the original description of the GCS as it implied a specific physio-anatomical correlation.[20] The best score should be recorded after testing all four limbs, and any discrepancy between limbs should be recorded separately.[4]

It should also be noted that it might not be possible to score verbal or motor function in some patients, for example those who are intubated (verbal score) or have a high-level spinal cord injury or limb injuries (motor score).

Table 4.2.1 allows comparison of the GCS and the CCS used by Hahn et al. and others.[9,30] The CCS has never been properly validated, and assessment of verbal response is somewhat subjective, particularly in children aged less than 6 months.[21]

A rapid assessment of a child's neurological disturbance can also be made using the **AVPU scale**, which denotes the child's response to stimuli (Table 4.2.2). The child who demonstrates a non-purposeful response to pain (withdrawal, flexor or extensor responses) has a level of consciousness consistent with a GCS of <9 and the unresponsive child, a GCS of 3.

Assess the child carefully for signs of trauma to the head, neck and thoracolumbar spine. A skull vault fracture may be suggested by scalp haematoma, crepitus or palpable depression. A basilar skull fracture should be suspected in the presence of 'racoon eyes', Battle's sign (bruising around the mastoid process), haemotympanum or CSF rhinorrhoea/otorrhoea. Any sign of trauma above the clavicles increases the likelihood of intracranial pathology being detected on CT scanning.[31]

A careful examination of the cranial nerves and peripheral nervous system should follow to assess for any localising signs. If spinal cord injury is suspected the sensory level of injury (dermatome) should be defined, not forgetting to check for intact perianal sensation and normal anal tone. Hypotensive bradycardia, altered perspiration below the level of the lesion, priapism in boys and urinary retention all signify autonomic dysfunction, which may occur with spinal injury. Any sensory or motor function below the level of injury implies that the cord lesion is incomplete and denotes a better prognosis.

Pupil size, equality and reactivity, particularly in the unconscious child, should be checked and recorded. A dilated pupil is defined as >4 mm diameter, asymmetry as >1 mm difference between pupil size and non-reactivity as <1 mm movement when a bright light is used to test the reflex. The pupillary size and reaction may be also altered by direct trauma to the globe, so one needs to consider traumatic mydriasis when the overall features are not in keeping with an intracranial cause of unilateral pupillary dilatation. Whenever possible, neurological examination should include an assessment of gait.

Investigations

Bedside

In children with mild TBI tests are generally not indicated. Blood sugar, heart rate, blood pressure, respiratory rate and SaO_2 can be performed. A 12-lead ECG may be needed in children with associated chest trauma or in a child with moderate or severe head injury as catecholamine release from TBI may have myocardial effects.

Laboratory

In patients with mild TBI, laboratory investigations are not indicated unless there is known or suspected coexistent pathology such as haemophilia or thrombocytopenia.

In patients with moderate or severe TBI the following should be checked:

- Haemoglobin level and platelet count
- Coagulation profile including fibrin degradation products (FDPs). Tissue thromboplastin released from the brain can cause coagulopathy and disseminated intravascular coagulation, the presence of which denotes a poor prognosis.[32,33]
- Electrolytes – these serve as baseline during fluid resuscitation and subsequent monitoring for hyponatraemia. Transient hypokalaemia is a well-recognised consequence of TBI and may occur even in mild cases. It usually resolves spontaneously after several hours, and no treatment is required.
- Arterial blood gas (ABG) should be considered, depending upon clinical situation. In ventilated patients, ABGs should be used to confirm that the end-tidal CO_2 ($ETCO_2$) accurately reflects $PaCO_2$.

Radiological

Skull X-rays

Before the widespread availability of CT scanning, skull X-rays were often used to screen or help risk stratify which children should be observed in hospital or have a brain CT scan.[8,28] While it is acknowledged that children with a skull X-ray demonstrating a fracture have a higher incidence of ICH compared to those without a fracture, many children without fractures also have an ICH.[9,11,12,17,30] To compound this problem, the reported sensitivity and specificity of skull X-rays in detecting a fracture is reported to be as low as 21% and 53%, respectively.[34] Emergency department (ED) staff need to be familiar with the interpretation of paediatric skull X-rays in order to minimise the misinterpretation of sutures or vascular markings as fractures and vice versa.

The role of plain films is very limited in seriously injured patients but may still have a role in children suspected of child abuse and the complications of skull fractures such as growing fractures (leptomeningeal cyst).[35,36] Skull X-rays

may also be useful in screening for depressed skull fractures or penetrating skull injury, particularly in environments where CT scanning is not readily available.

Cervical and thoracolumbar spine X-rays

These should be obtained for any child with suspected cervical or thoracolumbar trauma or evidence of spinal cord injury and those in whom the spine cannot be cleared clinically. Young children (age <10 years) are more prone to high cervical spine injuries whereas older children, like adults, are more prone to lower cervical spine injuries.[37] Spinal cord injury without radiological abnormality (SCIWORA) was thought to be principally a paediatric phenomenon but is in fact seen much more often in adults.[38]

Box 4.2.1 Indications for CT scan

- GCS ≤14
- Focal neurological deficit
- Signs of skull fracture or penetrating skull injury

Strongly consider if:

- unequal pupils
- abnormal behaviour
- depressed skull fracture >1 cm
- penetrating injury
- persistent vomiting
- history of LOC*
- post-traumatic seizure (delayed)
- post-traumatic amnesia
- moderate to severe headache
- underlying bleeding risk

* The duration loss of consciousness (LOC) does not correlate well with the risk of intracranial pathology, and anything greater than transient LOC (i.e. >1 min) should be considered significant.[12,23]

A low threshold should be held for CT scanning the vertebral column in any child with high clinical suspicion of spinal injury or possible radiological abnormality on plain film.

Computed tomography scan

In cases of TBI, computed tomography (CT) scan is the modality of choice for imaging the skull for fractures and brain for acute haemorrhage, oedema, mass effect, pneumocephalus and hydrocephalus.[8,39] It is rapid, inexpensive and can accommodate a wide range of life support and monitoring equipment unlike MRI. Any child with a persistent GCS ≤14, clinical evidence of a skull fracture or penetrating skull injury should have an emergent cerebral CT scan (Box 4.2.1; see also 'mild TBI'). The child needs to be accompanied by appropriate staff and monitoring equipment to the CT scan environment to continue optimal monitoring and rapidly manage any potential complications. Limitations of CT include limited ability to detect non-haemorrhagic lesions, small haemorrhages in posterior cranial fossa and brainstem.[40]

The CT is also used as the initial investigation of choice to further evaluate suspected spinal injury, although it cannot exclude ligamentous injury, and it provides limited information on injury to the spinal cord itself.[41,42]

Magnetic resonance imaging

MRI is superior to CT scanning for detecting cerebral oedema, contusion and diffuse axonal injury in cases of TBI.[8] It is also superior to CT scanning for visualising the posterior fossa and brainstem regions. It is the investigation of choice for spinal cord injury, and the actual patterns of haemorrhage and oedema within the cord carry prognostic significance.[42,43] MRI imaging of ligamentous tissues can be used to investigate possible spine instability. However, clinical correlation with MRI findings in 'mild' cases is still lacking.[41] Principal disadvantages of MRI in emergency setting include presence of metallic foreign bodies, high costs, long acquisition times, limited access to patients, patient cooperation and poor sensitivity for fractures.

Management

Mild traumatic brain injury (GCS 14, 15)

The management of mild TBI is controversial. The debate primarily focuses on the question of whether or not children in this group can be risk stratified and

managed clinically without further investigation or whether they need to have a cerebral CT scan regardless of clinical findings. Concern stems from the observation that even children with a GCS of 15 and a normal neurological examination can harbour clinically significant intracranial pathology with attendant risks for subsequent deterioration and death. One study of 429 children with mild TBI found that 16% of the children with GCS 15 and no LOC had significant IC injury.[44] Of these, 1.4% required neurosurgical intervention and 2% died.

On the other hand, CT scanning involves a significant radiation dose and subsequent life-time risk of radiation-induced malignancies which may be as high as 1 per 1500 scans in very young children.[45] Also, if very young, the child may require sedation or a general anaesthetic, with his/her attendant risks of apnoea, hypoxia, aspiration and prolonged sedation.[12] In addition to the radiation risks, the cost of scanning all children with head trauma would be considerable.

The observation has been made that the greatest benefit in treating patients with TBI is not with aggressive management of the severely injured but in preventing deterioration and complications in those with mild or moderate injuries who appear to be at low risk.[13]

Risk factors for IC injury or deterioration, such as LOC, amnesia, headache, vomiting, seizures and focal neurological deficit, have been studied in an attempt to identify those children with mild TBI who do not require a CT scan.[12,15,19,27,31,36,44,46–48] Two prospective studies evaluated guidelines for imaging in children with mild head injury. The PECARN study has been validated by many studies and decreases CT use. The risk of missing a clinically significant intracranial injury is 1 in 4000, and there is a separate algorithm for those under 2.[49]

The role of newer imaging modalities, such as functional MRI, diffusion tensor imaging and single positron emission, is currently being researched.[50,51]

The risk of developing clinically significant intracranial pathology following the initial trauma decreases over time. Although current Australasian guidelines for children suffering mild TBI suggest discharge after 4 hours of observation if the child has a GCS of 15 and is asymptomatic,[16] a large study of over 28,000 children admitted to hospital following head injury demonstrated that 6 hours of post-injury observation was required to identify all children who were likely to deteriorate.[46]

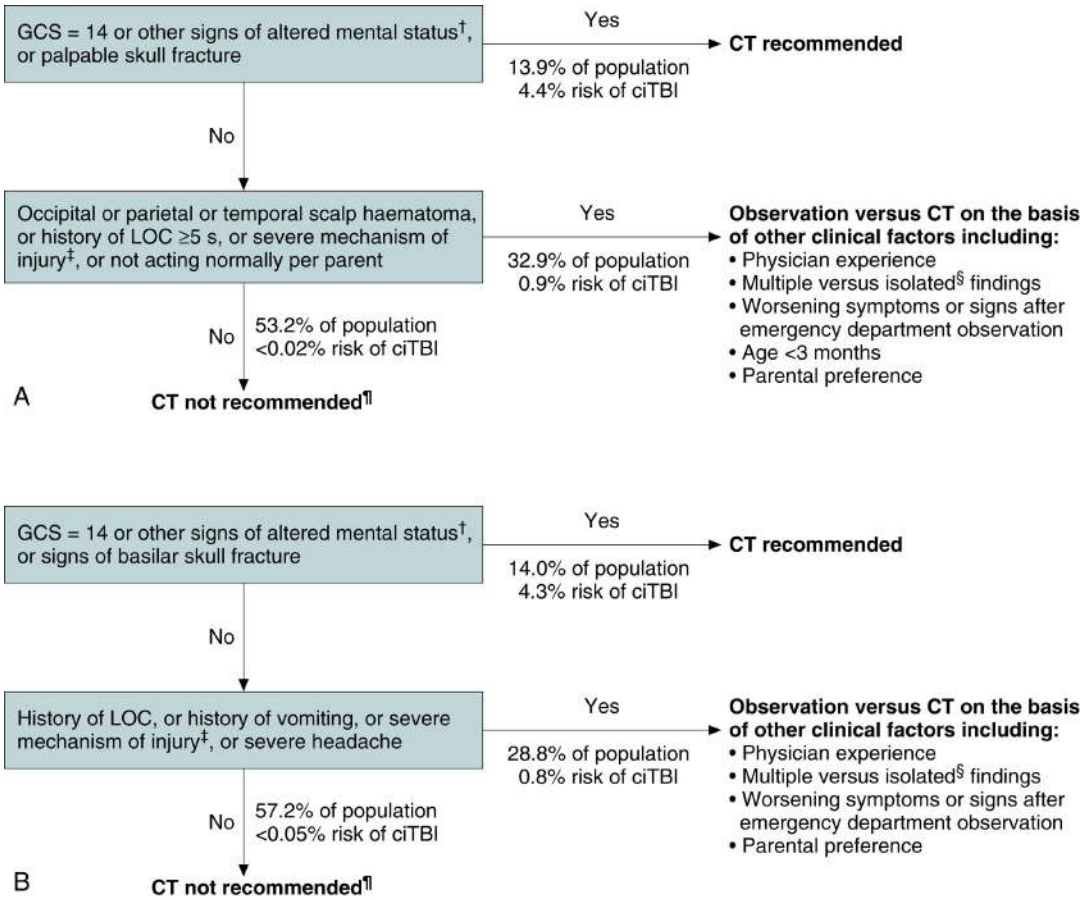

FIG. 4.2.1 Suggested management algorithm for mild traumatic brain injury in children younger than 2 years (A) and those aged >2 years (B). Kuppermann N, Holmes JF, Dayan PS, et al. Identification of children at very low risk of clinically-important brain injuries after head trauma: a prospective cohort study. *Lancet* 2009;**374**(9696):1160–70.

Based upon current knowledge, the management and disposition algorithm shown in Fig. 4.2.1 is suggested.

Moderate and severe traumatic brain injury

These groups will be considered together. There are no guidelines for the management of moderate head injury, but principles of brain protective management for severe TBI are applicable. It should be noted that the best GCS after resuscitation is used for classification of the TBI and that a child with a GCS ≤8 is, by definition, comatose. The priorities of management include maintenance of the airway, oxygenation and cerebral perfusion.

Airway control, oxygenation and ventilation

It is important to rapidly identify and correct hypoxia, as this is a significant

contributor to secondary brain injury.[44,50,52] Supplemental oxygen should be applied in order to maximise oxygen delivery to any ischaemic tissue, and the child's SaO_2 should be monitored.

Children who are not protecting their airway or maintaining adequate ventilation should be intubated and ventilated. In general, this would include any child with a GCS ≤8. In-line stabilisation of the cervical spine during rapid sequence oral intubation should occur to prevent potential secondary spinal injury. Rarely, the agitated or combative child with a higher GCS may require intubation in order to facilitate imaging or to stabilise a potentially at-risk cervical spine until it can be cleared.

Pharmacological adjuncts to intubation should be used as required. However, care is required to avoid transient hypotension as this worsens neurological outcome. While fentanyl is most commonly used in hemodynamically stable patients, sodium thiopental may be preferable because of its neuroprotective properties. The endotracheal tube should be securely fastened to the child's face with tape rather than using the standard 'tie around the neck'. This avoids the risk of impairing cerebral venous drainage and causing a consequent rise in ICP. Hard collars may also constrict the neck and therefore interfere with venous drainage, so they need to be fitted correctly.

An orogastric tube should be passed to decompress the stomach. The nasogastric route should be avoided as inadvertent intracranial placement can occur if the child has a basal skull fracture.

SaO_2 and PaO_2 are not good indicators of adequate ventilation, and continuous $ETCO_2$ monitoring should occur if the child is intubated. The child should be ventilated to maintain an $ETCO_2$ in the low–normal range (i.e. aiming for an $ETCO_2$ of 35 mmHg). Hypercarbia secondary to inadequate ventilation may result in cerebral vasodilation and secondary increase in ICP. Hypocarbia causes cerebral vasoconstriction and therefore decreases ICP but increases the risk of causing or exacerbating cerebral hypoperfusion with secondary ischaemia.[53] Routine 'prophylactic hyperventilation' in adults has been shown to worsen outcome and presumably does so in children. Therefore hyperventilation to a $PaCO_2$ of 25 mmHg should be reserved for the child who is rapidly deteriorating with signs of increased ICP or cerebral herniation, such as new onset pupil asymmetry or rapidly decreasing GCS.

Circulation

Hypotension is the single most significant factor contributing to secondary brain insult.[44,52] Hypotension is defined as an SBP <90 mmHg in adults and a SBP <5th percentile for age in children.[44,50–52,54] One or more episodes of hypotension from time of injury through resuscitation at least doubles mortality and significantly increases morbidity.[44,52,54–56]

Despite studies focusing upon SBP, the true objective in the patient with TBI is to maintain an adequate CPP. This is the difference between mean arterial pressure (MAP) and ICP (i.e. CPP = MAP – ICP). The normal ICP is 0–10 mmHg, and an ICP of 20 mmHg is generally regarded as the threshold for initiating specific therapy to reduce it. The optimal CPP is uncertain, but consensus opinion recommends not less than 50 mmHg and not greater than 70 mmHg, as aggressive attempts to maintain CPP >70 mmHg may also worsen outcomes.[18] In the pre-hospital and ED settings, the ICP is unknown so treatment is purely empirical. It is reasonable to aim for a MAP of between 70 and 90 mmHg that would maintain a CPP between 50 and 70 mmHg (i.e. allowing for ICPs up to 20 mmHg). Current recommendation is to maintain CPP between 40 and 60 mmHg.[6] There are no paediatric studies demonstrating survival advantage with a CPP above target threshold.[57]

Initial resuscitation should be with dextrose-free isotonic solutions such as 0.9% saline or compound sodium lactate (Hartmann's) solution, although the use of hypertonic saline may be considered (see below). In the hypotensive patient, give a 20 mL kg^{-1} bolus (if using an isotonic crystalloid), and monitor the response. Repeat if the patient remains hypotensive. After a total of 40 mL kg^{-1} volume resuscitation, consideration should be given to the use of vasopressor support to maintain adequate MAP for cerebral perfusion. In the setting of hypovolaemia due to acute blood loss, ongoing resuscitation fluid will require blood product replacement.

Hypertonic saline

Hypertonic saline solutions have been used for initial resuscitation, as maintenance fluid and as specific treatment for raised ICP. In addition to rapidly restoring circulating volume, increasing blood pressure and reducing ICP, these solutions appear to have important and beneficial immuno-modulatory and neuro-chemical effects that may reduce secondary brain injury.[58,59]

A suggested regimen for the use of 3% saline for either initial resuscitation or to rapidly decrease ICP is to give a 5 mL kg^{-1} bolus, repeated if necessary according to patient response. There is class II evidence to support use of small

boluses and class III evidence for continuous infusion of saline aimed at maintaining ICP <20. Serum osmolality should be maintained less than 360 mOsm L.[60] It would appear that rapid changes in serum sodium concentration do not cause complications such as central pontine myelinolysis in humans, and most studies do not place an upper limit on serum sodium concentration.[58,61]

However, there have been no randomised controlled trials demonstrating improved outcomes with hypertonic saline.

Mannitol

Mannitol has been used as both a resuscitation fluid (plasma expander) and as therapy for acute deterioration secondary to increasing ICP. Boluses of 0.25 g kg^{-1} to 1 g kg^{-1} body weight have been used successfully for short-term reduction of ICP.[62] Mannitol acts as an osmotic diuretic and can lead to subsequent problems with hypovolaemia and acute renal failure via acute tubular necrosis. A loop diuretic such as furosemide is sometimes used in addition to mannitol for treatment of acute rises in ICP. A urinary catheter is essential in any patient receiving mannitol or diuretics. No study has been performed in children to support this treatment strategy.[60]

Positioning

The child's head should be kept in the midline to avoid jugular venous compression. The neck collar should be snug but not tight. Venous drainage is improved if the head of the bed is elevated 15–30 degrees, with resultant decrease in ICP. However, CPP is also reduced by this manoeuvre, and there may be no net benefit unless ICP monitoring is in place and CCP adjusted as required.

Sedation and prophylactic anti-seizure therapy

Avoiding all noxious stimulus helps keep the patient calm. Sedatives and analgesics decrease cerebral metabolism and may decrease long-term psychological effects. Neuromuscular blockade has the benefits of decreasing metabolism, but there are no good studies to support their prophylactic use.[63]

Seizures that occur within 7 days of a TBI are termed early post-traumatic seizures (EPTS), and those thereafter are late post-traumatic seizures (LPTS).[57] The overall incidence of seizures in children with TBI is probably between 5 and 15% but rises with increasing severity of TBI, occurring up to 40% of the time in those with a GCS ≤8.[52,57,64,65] Greater than 95% of post-traumatic seizures are

early, and approximately 80% of these occur within the first 24 hours.

Seizures may cause or exacerbate secondary brain injury by increasing cerebral metabolic demands, increasing ICP and by causing or exacerbating cerebral hypoxia.[64,65]

The incidence of EPTS can be reduced by the use of prophylactic anticonvulsants such as phenytoin.[66] Their use should be considered[65] in those with:

- GCS <10
- seizure within 24 hours of injury
- depressed skull fracture or penetrating head wound
- subdural, extradural or intracerebral haematomas
- cortical contusion.

It should be noted, however, that reduced EPTS does not translate into reduced mortality[66,67], and it remains to be seen whether or not an overall improved level of functioning occurs in those survivors given prophylactic anticonvulsant therapy. It should also be noted that prophylactic anticonvulsants do not alter the incidence of LPTS, and their routine use after 7 days is not recommended.[66]

The management of active seizures should be with benzodiazepines and should follow the guidelines discussed in Chapter 8.3.

Intracranial pressure monitoring

Routine measurement of ICP is not indicated in all children with moderate or severe head injury. Intracranial hypertension is difficult to diagnose, can be present in children with normal fontanelles and is associated with poor neurological outcomes. While ICP measurement is indicated in children with severe head injury and abnormal CT, it may occasionally be indicated in conscious children with findings predictive of raised ICP.[68] ICP monitors also have the advantage of draining CSF to relieve pressure. Some systems allow measurement of tissue oxygenation; however, thresholds are based on adult data.[69]

Near infrared spectroscopy

Near infrared spectroscopy (NIRS) technology is used to monitor global blood flow. Normal values for children have been defined. Combination of NIRS with

CT has been used as surrogate marker of blood flow in adults. Its utility in paediatric TBI is yet to be defined.[69]

Antibiotics

Intravenous antibiotic prophylaxis with flucloxacillin 25–50 mg kg^{-1} 6-hourly should be given for open skull fractures or fractures in communication with sinuses.

Steroids

Various steroids have been used in an attempt to improve outcome from TBI. To date no overall improvements have been identified, and they may actually worsen outcome. Routine use is no longer recommended.[59,63,67]

Thermoregulation: prophylactic hypothermia and prevention of hyperthermia

Mild hypothermia (32–35°C) is known to decrease ICP and, in animal models, has been shown to be neuroprotective.[56,58,70] Outcomes in humans have not been consistently better, and mild hypothermia may increase complications such as sepsis, pneumonia, bleeding and mortality in the TBI child.[58,71] Three major trials in children have not demonstrated survival advantage so hypothermia should not be used prophylactically, and if used it should be for a short time.[69] Hyperthermia is associated with a worse outcome in children with severe TBI.[71] It is not known if actively cooling the patient alters outcome, and further research needs to be conducted in this area. In the meantime it would seem reasonable to attempt to cool a febrile brain-injured child.

Decompressive craniectomy

Decompressive craniectomy where a portion of the skull and dura is removed to relieve pressure is sometimes required to improve CBF. It is indicated in children showing early signs of deterioration or herniation. Research on the utility of this procedure has been difficult because of the variations in the procedure and indications. The DECRA study demonstrated poor outcome at 6 months, but the procedure is still performed when other methods to relieve ICP have failed.[72]

Spinal cord injury

There is a great paucity of research into the optimal management of acute spinal

cord injury in children. Therefore there are insufficient data to support diagnostic or treatment standards.[39] However, the principles of management of acute spinal cord injury are considered to be no different from TBI. The focus of therapy is to prevent secondary injury. Attention should be paid to the maintenance of strict spinal immobilisation, adequate oxygenation, ventilation, blood pressure and good supportive care as per moderate to severe TBI.

The use of high-dose steroids in acute spinal injury is controversial.[73,74] The issue is further complicated in the paediatric population by the fact that children <13 years were excluded from the major trials of steroids for spinal cord injury.[41] Routine use of high-dose steroids for spinal cord injury is no longer recommended by the Neurosurgical Society of Australasia.[16] If steroids are used, in consultation with local paediatric neurosurgical practice, they should be administered within 8 hours of injury. Practice currently varies between institutions. The recommended dosing schedule is: methylprednisolone 30 mg kg^{-1} bolus over 15 minutes followed by a 45-minute break, then 5.4 mg kg^{-1} hr^{-1} continuous infusion for 23 hours.[16]

Supportive care

Immobilisation of children at risk of spinal injury is difficult when they are non-cooperative and needs to include the body as well as the head. Involvement of parents or carers and calming of the child are important adjuncts. Pressure area care should occur for all patients who are immobilised. Children who arrive in the ED on ‘spinal boards’ should be moved off them as soon as possible as pressure areas may develop rapidly.

Hard collars should be checked for correct fit and potential pressure areas. They should be changed to padded collars such as Aspen or Philadelphia collars if immobilisation is needed for an extended period.

A urinary catheter should be placed in children who require volume resuscitation, mannitol or diuretics, who are unable to communicate their need to micturate because of TBI, or who have a spinal cord injury.

The child’s temperature should be monitored and inadvertent cooling avoided (see comments on thermoregulation above). The child’s analgesic requirements should be regularly reviewed and analgesia titrated as required. Attention should be given to eye protection in children who are sedated or ventilated.

Family considerations

There are considerable immediate stresses during the initial stabilisation phases on the parents and family of the injured child. The family requires appropriate support and explanation whilst in the ED. If possible, it is useful to provide a dedicated staff member to be with them. The family should be kept well informed of the child's status and his or her proposed management by a designated senior medical member of the resuscitation team, and consideration should be given to allowing parents into the resuscitation room to be with their child (see Chapter 2.1). Premature or vague conclusions of prognosis should be avoided until all relevant assessments and investigations have been made.

Disposition

The disposition for children with mild TBI has been outlined above. Children sent home from the ED following a head injury must be discharged to the care of a responsible adult who is given clear discharge instructions and a written head-injury sheet containing advice on when to seek review. Children who have had a significant concussive injury need to rest cognitively for the next 24–48 hours – avoiding television, computers, music inputs, to minimise excessive visual-auditory stimulation. Returning to these activities too soon will often cause a recrudescence of their symptoms. Simple analgesics such as paracetamol can be used for mild headache, which should be expected to be short-lived. Any child with a suspected NAI should be admitted to hospital for safety and further evaluation.

All children with moderate or severe TBI or spinal cord injury should be transferred to a paediatric high-dependency or intensive care unit for admission under the care of a neurosurgeon and rehabilitation service.

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4.3

Spinal injury

Harsh Priyadarshi, and Soundappan S.V. Soundappan

ESSENTIALS

1 Spine injuries are rare in paediatric patients, but high morbidity and mortality are associated with concomitant and associated injuries.

2 Upper cervical spine injuries are more common in children. Thoracic and lumbar spine injury is strongly associated with spinal cord injury along with gastrointestinal and orthopaedic injuries.

3 Adequate stabilisation of the entire spine is needed. Rigid spinal immobilisation may cause complications, especially in the patient with spinal cord injury.

4 Clinical evaluation of the cervical spine can only occur in a child who is conscious, able to communicate and free from distracting injuries. The cervical spine can be cleared on clinical findings alone in specific circumstances.

5 A good-quality three-view series of the cervical spine (lateral, AP and odontoid view) is adequate to assess the bony structures. Clinical findings should be considered when clearing the cervical spine radiographically.

6 Patients with spinal injury have a high incidence of multisystem involvement/fractures at another level.

7 Spinal cord injury may cause hypotension and relative bradycardia.

8 Spinal cord injury may occur without roentgenographic abnormality.

Introduction

Injuries to the spine and spinal cord are less common in children than adults. The injuries (varies in different age group of children depending on developmental, anatomical, biomechanical and physiological differences). Children account for up to 10% of all spinal injuries, but the mortality among spine-injured children is higher than in adults, with estimates ranging from 25% to 30%, with death most often due to associated injuries to other organs, especially the brain.[1]

The incidence of spinal cord injury amongst spine-injured children is estimated to be about 1%.[1,2] In neurologically impaired survivors, the injuries are most commonly at the upper cervical (C0–C4) level or at the cervico-thoracic junctional spine.[1] The common causes of spine and spinal cord injuries in children are motor vehicle crashes, falls, diving accidents, sports injuries and, occasionally, non-accidental injury.[1,3,5]

The majority of injuries in children occur in the cervical spine. In children under 8 years of age, most (about 80%) occur in the C1–C3 region, whereas after 8 years of age the incidence is similar to that in adults, with the majority in the lower three cervical vertebrae.[2,6,7] In children older than 9 years of age, spinal column injuries tend to occur in the thoracic, lumbar and sacral regions of the spine, rather than in the cervical spine as with very young children. The thoracolumbar junction is the most commonly injured area outside the cervical spine, with the thoracic and lumbar spines having roughly equal incidence of about 25%. There is an increased incidence of neurologic injury in fractures of the thoracolumbar junction. The relatively high incidence of injuries in this region is due to the increased range of motion and the changing orientation of the facet joints.[1,8] Approximately 30% of patients with spinal cord injury have fractures at more than one spinal level. A majority of these are in contiguous vertebral segments, but 5–15% may be in different regions.[8]

Developmental anatomy and physiology

There are a number of patterns of injury to the spine, especially the cervical spine, which are unique to children. In order to understand the differences between adult and paediatric spine injuries, knowledge of the developmental anatomy of the spine is essential.

Radiographic evaluation in cervical spine is rendered difficult due to cartilaginous ossifying centres which may mimic fractures. This is especially

true in the first few years of life and more so in the upper cervical vertebrae.

The *atlas* ossifies from three ossification centres: two ossification centres of the lateral masses and one ossification centre for the body which does not ossify until about 1 year of age. The posterior arches fuse by 3 or 4 years of age, while the synchondrosis between the lateral masses and the body fuses at approximately 7 years of age.[3,6]

The *axis* ossifies from seven ossification centres. The five primary ossification centres are two for the lateral masses, two for the odontoid (which are usually fused at birth but occasionally persist as a dens bicornis) and one for the body. The odontoid is separated from the body by a synchondrosis, which fuses between 3 and 6 years old. The two secondary ossification centres are the tip of the odontoid process (which appears at about 3 years of age and is usually fused by 12 years of age) and the inferior ring apophysis (which, like other ring apophyses, generally ossifies after 8 years of age and fuses in the early 20s).[3,7]

The remaining cervical vertebrae have three primary ossification centres, one for the body and one each for the two neural arches, and two secondary ossification centres, the ring apophyses. The neural arches fuse posteriorly by the age of 3 and three ossification centres anteriorly which fuse between 3 and 6 years of age. Importantly, the vertebral bodies are wedge shaped until the age of 7 when they begin to square off.[3,4]

The thoracic and lumbar spines develop in a similar way, with additional secondary ossification centres for the spinous process and the transverse processes. By the time the child is 8–10 years of age the spine has reached near-adult size.[3,4]

There are a few biomechanical and physiological considerations which make the paediatric spine more vulnerable. The fulcrum of movement at the neck is located at C2–C3 in the infants and at C3–C4 by the age of 6, and by the age of 8 the fulcrum is at C5–C6, comparable to the adult. There is a relatively large head and weak neck muscles; laxity of the ligaments and joint capsule; and relatively horizontal positioning of the facet joints with underdeveloped uncinate processes.[1,3,4] The thoracolumbar vertebrae which are still cartilaginous most often have multilevel vertebral compression resulting in plastic fracture of the spine.[5] All these features increase the risk of injury to the child's spine.

Initial assessment

All patients with significant injury should be assumed to have a spine or spinal

cord injury, and appropriate precautions must be taken to prevent further exacerbating any possible injury. The initial assessment of patients with potential spine or spinal cord injury should be directed at the airway, breathing and circulation, in line with trauma resuscitation guidelines (see Section 4). A thorough secondary survey is performed. At this point all possible spine and spinal cord injuries should be identified. The lateral cervical spine X-ray would be sometimes performed at this stage in patients with major trauma, unless computed tomography (CT) imaging is indicated. Thorough radiological assessment of the injuries should be completed once resuscitation and the secondary survey have been accomplished.

A thorough history of mechanism of injury, previous spinal injury, other illnesses (particularly respiratory and cardiac illness [acute or chronic]), problems, bone disorders, medication and allergies is needed to determine the patient's premorbid physiological status.

Spinal immobilisation

Spinal immobilisation is currently a controversial issue, especially in young children. A balance must be found that will diminish the risk of further injury to the child's spine but not interfere with the assessment, or the normal physiological functions, of the child. Traditionally the spine has been immobilised in a rigid cervical collar, on a spine board with a head immobiliser and straps, or with sandbags and tapes, thus providing adequate control of the entire spine (Box 4.3.1).[3,9,10]

Box 4.3.1 Indications for initial immobilisation of the spine

- Level of consciousness
- Inability to give history of pain
- Neck or back pain
- Neurological signs or symptoms
- Multiple system trauma
- History of significant trauma:
    - Fall from height >3 metres
    - Pedestrian or cyclist hit by car

- Unrestrained passenger in motor vehicle
- Crash diving accident

- History of spinal abnormality

There are a number of potential problems with this immobilisation (Box 4.3.2). Cervical collars are not made to fit infants, and alternative immobilisation is needed. An ill-fitting collar may cause the chin to become trapped under the chin support and may cause airway obstruction. The young child may become distressed from being rigidly immobilised, making further assessment difficult and potentially raising intracranial pressure. Rigid immobilisation in a collar and head immobiliser on a spinal board has also been shown to decrease tidal volume and respiratory excursion.[11] In addition, because the young child's head is disproportionately large, the neck is flexed when immobilised on a standard spinal board. This causes flexion of the cervical spine, which may cause movement at the site of injury. To prevent this, a spinal board with a recess for the head or padding that elevates the torso is needed for children less than 8 years old (Fig. 4.3.1).[12–14]

Box 4.3.2 Problems associated with spinal immobilisation in children

- Incorrectly fitted cervical spine collar causing distraction of the spine or allowing excessive movement
- Flexion of the spine in children under 8 years of age
- Reduction in tidal volume and limitation of respiratory effort
- Airway obstruction
- Increased intracranial pressure
- Discomfort
- Distress and anxiety
- Pressure sores

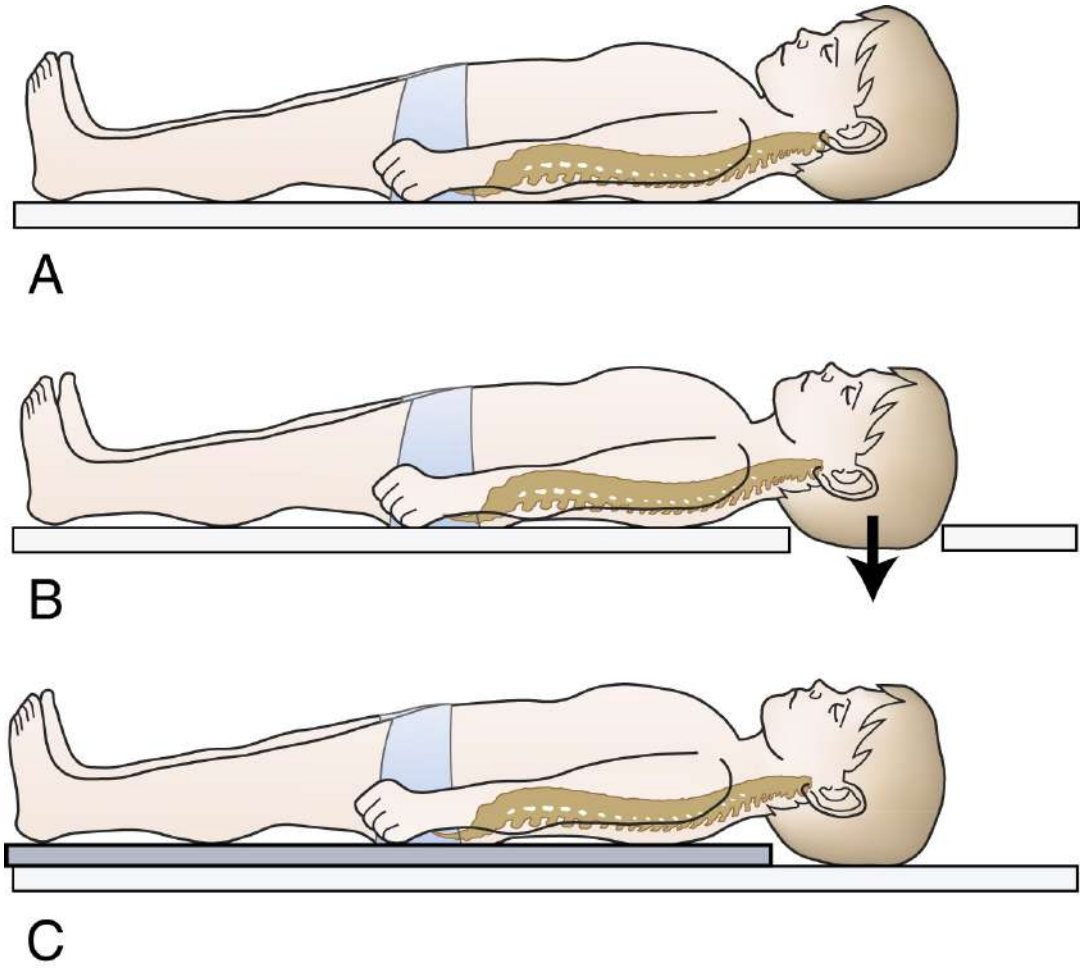


FIG. 4.3.1 Effects of spine board on cervical spine position in children. Child immobilised on a standard backboard (A), and on backboards modified with an occipital recess (B) and a mattress pad (C). Modified from Herzenberg JE, Hensinger RN, Dedrick DK, Phillips WA. Emergency transport and positioning of young children who may have an injury of the cervical spine: the standard backboard may be hazardous. *J Bone J Surg Am* 1989;**71**(1):15–22.

A number of different cervical collars are available, and the clinician must be familiar with the method of sizing and applying the collar available. An incorrect fit will allow undue movement of the neck if the collar is too small in width or will produce a distraction of any existing injury if too large.

For children who have less severe trauma but are potentially at risk of spinal injury, an appropriate level of immobilisation may be left to the discretion of the clinician. However, in all patients with multiple or significant distracting injuries spinal immobilisation is still recommended. If the child is not on a spine board, immobilisation solely in a cervical collar, maintaining alignment of the neck and the entire spine, is appropriate. Using sandbags and tape or other means to immobilise the head by fixing it to the bed risks further complications, including

airway compromise and pressure sores, and the risks are likely to outweigh the benefits.[15,16]

The back is examined by log rolling the child which requires at least three people in the smallest of children and up to five in larger patients. The child is freed from the head immobiliser and any strapping to the spinal board removed. One person must stabilise the cervical spine by supporting the head and neck in the neutral position throughout movement. No traction should be applied to the neck. The other team members control the shoulders, hips and legs, and one person must be free to inspect and palpate the back. The back is inspected for bruises, abrasions, wounds or deformity of the spine. The spinous processes are palpated for tenderness. The back of the neck and head and the buttocks and anus should be inspected. A rectal examination is not mandated in all children.

To limit discomfort and the likelihood of pressure sores a child should be kept on the spinal board for the shortest possible time. Once resuscitation and urgent procedures and investigations are completed the board should be removed. This is especially important in patients with suspected spinal cord injury.[17]

Cervical spine injuries

As the cervical spine is the most commonly injured region accounting for the majority of spinal cord injuries, a thorough knowledge of injuries in this region and the appropriate assessment of the spine both clinically and radiographically are mandatory. In children younger than 8 years of age the majority (but not all) injuries occur above the fourth cervical vertebra. After 8 years, the pattern of injury is similar to that seen in adults (the majority of injuries below C4).[2,18]

Mechanisms of injury

The mechanism of injury is an important historical factor, as it will determine the type and possible instability of the underlying injury (Table 4.3.1).[16]

Flexion

The most common mechanism of injury seen is hyperflexion. This type of injury produces a compressive force on the anterior segment of the vertebra – leading to a compression fracture or a teardrop fracture. The posterior elements of the spine are distracted with ligamentous injury, dislocations, or avulsion fractures

of the spinous processes. These injuries can be stable (such as the anterior wedge compression fracture or the clay shoveller's fracture) or unstable (such as a bilateral facet joint dislocation).

Extension

Hyperextension injuries produce distracting forces anteriorly, while compressing the posterior vertebral structures. Most extension injuries are unstable, and buckling of the ligamentum flavum into the posterior of the spinal canal can cause a central or posterior spinal cord syndrome.

Rotation

In the cervical spine isolated rotary injuries are uncommon. Subluxation can be spontaneous or follow minor or major trauma. It is generally a stable injury. However, if there is dislocation of the facet joints of C1/C2 the lesion is unstable.

Vertical compression

These injuries are due to axial compression of the cervical spine. This can produce a burst fracture of any vertebra, with a lesion at C1 being most unstable. Spinal cord involvement is from a retropulsed fragment of bone or intervertebral disc.

Table 4.3.1

A classification of spine injuries

Mechanism of injury	Stability
Flexion	
Flexion teardrop fracture	Very unstable
Bilateral facet joint dislocation	Unstable
Atlanto-occipital dislocation	Unstable
Displaced odontoid fracture	Unstable
Anterior subluxation	Unstable
Anterior wedge fracture	Stable
Clay shoveller's fracture	Very stable
Extension	
Atlantoaxial dislocation	Very unstable
Hangman's fracture C2	Unstable
Extension teardrop fracture	Unstable (in extension)
Rotation	
Rotary atlantoaxial dislocation	Unstable
Rotary atlantoaxial subluxation	Stable
Unilateral facet dislocation	Stable
Vertical compression	
Jefferson fracture (burst # C1)	Very unstable
Burst fracture vertebral body	Stable

Modified from Rosen P, Barkin R (eds). *Emergency medicine*, 4th ed. St. Louis: Mosby; 1997.

In many instances, not one but a combination of these mechanisms of injury is involved, producing more than one type of injury.

Clinical assessment

After stabilisation and resuscitation of the injured child the neck should be examined as part of the secondary survey. Neck abrasions and other telltale signs of injuries are looked for followed by a palpation of the soft tissue and bony contour of the cervical spine with a conscious effort not to hurt the patient. The decision to evaluate the cervical spine for injury should only be made in children who are conscious and alert, not influenced with drug or alcohol, and who do not have other injuries that are painful or distracting enough to make assessment of neck pain difficult.[2,18–20]

The cervical collar should be removed while another person holds the head inline to the body without any traction to the neck. The cervical spine is palpated for tenderness over the spinous processes. If there is tenderness at a specific region the collar should be reapplied and the spine evaluated with X-ray. If there is no tenderness (or only soft tissue tenderness) the child should be allowed to gently move the head from side to side. If this produces pain posteriorly in the neck the collar should be reapplied and the spine X-rayed. If there is no pain on movement the collar and cervical spine protection can be removed.[2,18–20]

Young children represent a difficult subgroup. They are preverbal and cannot follow commands or communicate easily. If palpation of the posterior cervical spine does not cause distress the neck should be let free; if the child spontaneously moves the neck without discomfort the neck can be cleared. It is the authors' experience that no child with a neck fracture will spontaneously move his/her neck without discomfort.

Radiographic images

Radiological evaluation is required for all children who do not meet all the criteria for clinical clearance of the cervical spine. As part of the secondary survey in major trauma patients the cross-table lateral cervical spine X-ray would have been performed. This is a guide to the presence of serious cervical spine trauma only and cannot be used to exclude cervical spine injury.[2,18] All patients who require radiological evaluation require a full cervical spine series.

The cervical spine series consists of a lateral film, an anterior-posterior film and an odontoid view. Using these three views nearly all bony abnormalities in the cervical spines would be detected, allowing further investigation to fully delineate the individual injuries.[2,6] All seven vertebrae must be included in the lateral view, along with the cervicothoracic junction. If this is not visible gentle traction should be applied to the arms and the film repeated, or a swimmer's view (transaxillary) should be obtained. Oblique views of the cervical spine may also be of assistance, especially if the cervicothoracic junction is difficult to visualise. Oblique views, however, do not provide much more information regarding the likelihood of injury compared to the standard three views.[2,6,18]

In young children, cooperation to obtain the odontoid (open mouth) view is difficult. It has been shown that this view can be excluded in children under 5 years of age with little likelihood of missing a fracture.[21,22]

Other specialised radiological investigations have been used to further evaluate cervical spine injuries.

Flexion and extension

Lateral cervical spine radiographs have been used to assess the spine for ligamentous injury. The suggested indications have been symptomatic patients with normal plain X-rays usually on a later date for a follow-up or the unconscious patient with normal X-rays and/or CT scans. These images have become popular in assessing adult patients, but investigations to date have consistently failed to show a benefit over other imaging, with a significant number of studies limited by inadequate motion in the acute setting.[23] Most of the paediatric cervical spine guidelines do not call for the routine use of these views.[24]

Computerised tomography

There have been a number of indications for routine CT suggested, with the most widely accepted being for further evaluation and elucidation of fractures identified or to view areas not seen adequately on the initial cervical spine series.[18] Other suggested indications are for the assessment of the unconscious patient with normal initial radiographs and for patients having a CT of the brain. Proponents of CT suggest scanning the entire spine in both of these instances and clearing the spine if scans are normal[20] (or progressing to flexion-extension views).[18,25] Another group suggests just scanning the upper cervical vertebrae in

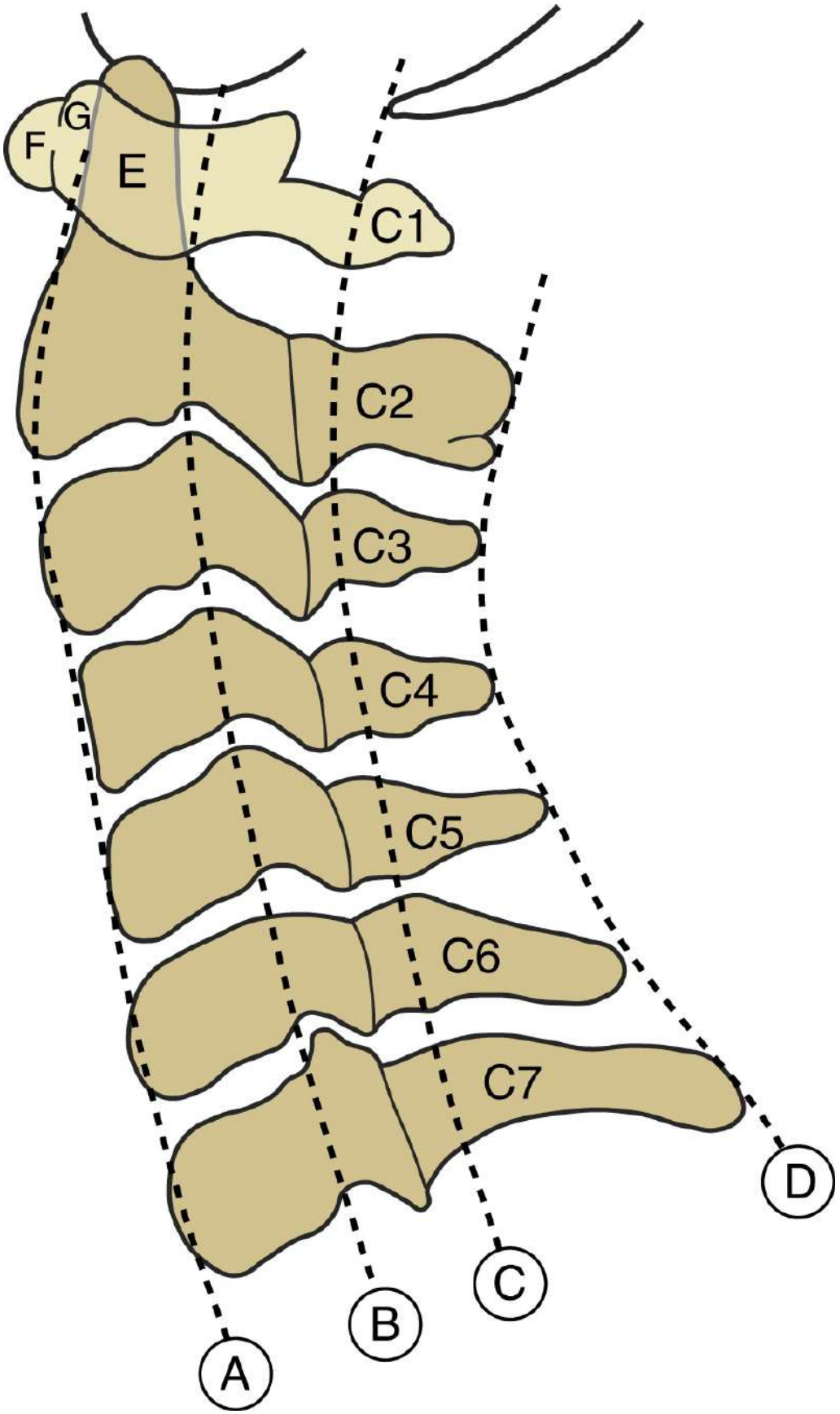
children under 8 years as most injuries occur in this region.[26,27] There are no studies that have systematically evaluated the role of CT in the evaluation of paediatric cervical spine injuries. CT appears to be as good as any other modality in identifying injuries, but the radiation exposure of young children needs to be considered.[24]

Magnetic resonance imaging

MRI is the imaging method of choice for assessing ligamentous injuries and for spinal cord injuries. MRI will visualise most ligamentous, discal, capsular, soft tissue injuries and all spinal cord or neural injuries. In many instances MRI will alter the specifics of surgical management in those who require surgical stabilisation. MRI may aid in prognosticating information in children with spinal cord injury.[18,28]

Radiographic evaluation

Once the X-rays have been obtained, care needs to be taken to interpret the images accurately and correlate the findings with the history and physical examination. Due to the physiological variations described previously, a number of normal radiological findings in children are significantly different from those in adults. The common findings that cause concern are pseudosubluxation of C2 on C3 (seen in up to 25% of children); exaggerated atlanto-dens distance (seen in 20% of children under 8 years of age); and radiolucent synchondrosis between the odontoid and C2 (seen in all children under 4 and in 50% of those under 10 years of age). Other normal findings that can be misinterpreted include a variable anterior soft tissue width – altering with head positioning and crying – the anterior ring apophyses of the vertebral bodies, and the anterior wedging of the vertebral bodies (especially C3).[18,29,30] All of these normal findings may be mistaken for acute traumatic injuries in children following trauma.


G
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C4
C5
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C7
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D

FIG. 4.3.2 Cervical spine lines.
Lateral cervical spine. (A) Anterior vertebral bodies; (B) posterior vertebral bodies and anterior spinal canal; (C) spinolamial line and posterior spinal canal; (D) spinous process tips C2–C7; (E) odontoid process of dens of C2; (F) anterior arch of C1; (G) predental space between posterior surface of anterior arch of C1 and anterior surface of odontoid process. Modified from Barkin RM, Rosen P (eds). *Emergency pediatrics,* 4th ed. St. Louis: Mosby; 1994.

Evaluation of the lateral cervical spine radiograph begins with assessment of the four lines, aligning to the anterior vertebral bodies, the posterior vertebral bodies, the spino-laminar line, and the tips of the spinous processes. All four of these lines should follow a smooth, even contour (Fig. 4.3.2). The articular facets should be parallel, the intervertebral disc spaces, at the posterior margin of the vertebral bodies, should be equidistant, and the two adjacent spinous processes should not elicit significant widening (fanning). Review of the soft tissue shadow should show a retropharyngeal space of not more than one-half the AP diameter of the vertebral body at C2 and no wider than the full width of the vertebral body at C6. As mentioned earlier, this may be difficult to interpret in the crying child.[1,3,18]

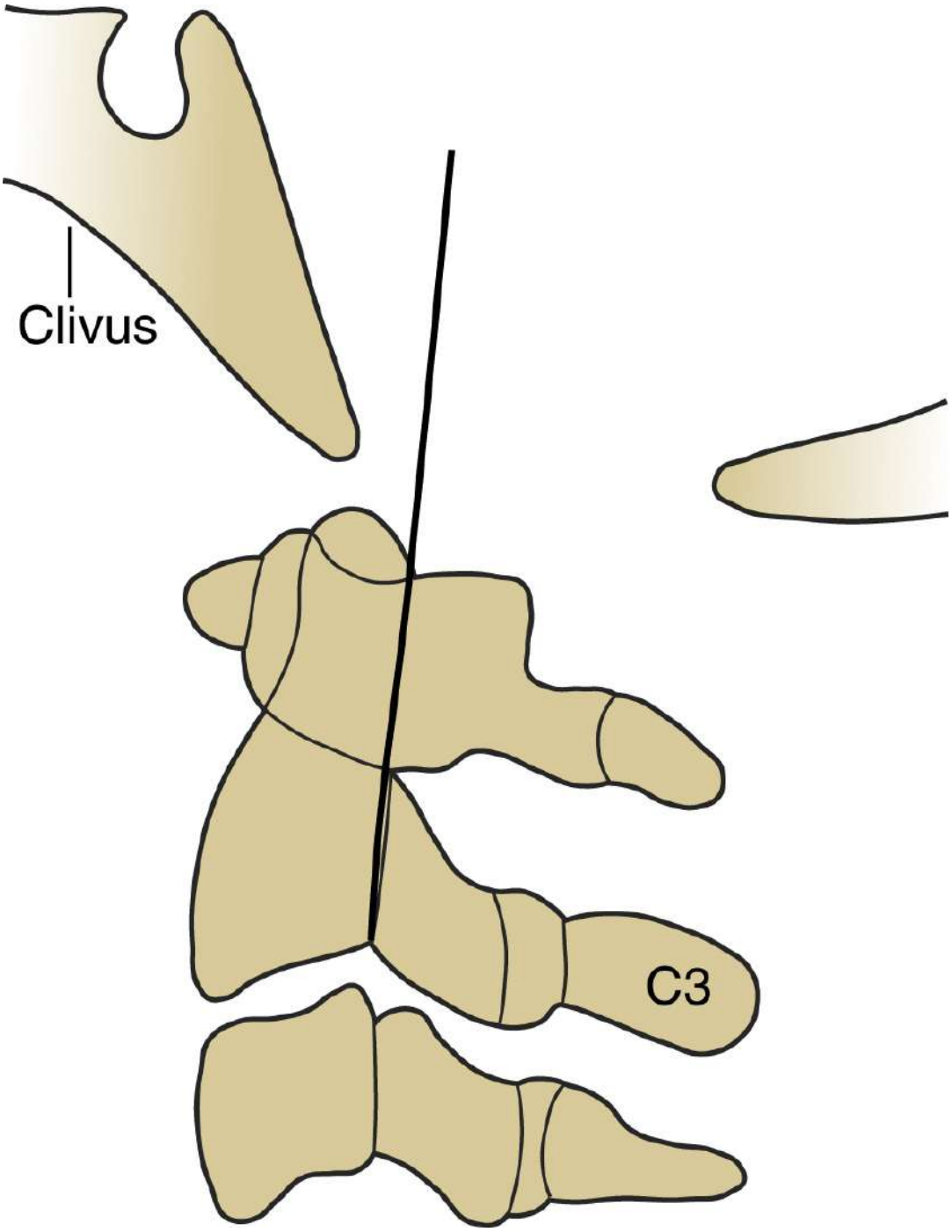

FIG. 4.3.3 Posterior axial line for identification of occipito-axial dislocation.

Assessment of these areas of possible abnormality has been made easier by the formulation of a series of normal range of measurements. For the atlantoaxial distance (AAD) at C1–C2 on a lateral film is a measurement from the posterior border of the anterior arch of C1 to the anterior margin of the odontoid which should be less than 5 mm in children under 8 years of age and ≤3 mm in older

children and adults. To assess the relationship of the basion of the skull to the atlas, the most reliable measurement in children is Harris's posterior axial line (Fig. 4.3.3), which should lie within 12 mm from the basion of the skull.

The subluxation of C2 on C3 and C3 on C4 consistently causes difficulty in interpretation of paediatric cervical spine films. Swischuk's line, joining the spino-laminar line of C1 to that of C3, substantiates the likelihood of the subluxation being physiological (Fig. 4.3.4). The spino-laminar junction of C2 should lie within 2 mm of this line. If the distance is greater than 2 mm a fracture or pathological subluxation is likely.

Cervical spine clearance guidelines

With the knowledge of mechanism of injury, immobilisation needed, clinical examination, and radiographic interpretation, it is possible to devise guidelines (Fig. 4.3.5) that allow safe and effective management of the potentially injured cervical spine while minimising the investigations needed. There is as yet no guideline that is well established for young children.[31,32] The guideline should include the following points:

- All patients with a significant mechanism of injury, altered conscious state, or neurological symptoms or who cannot be assessed should be immobilised.

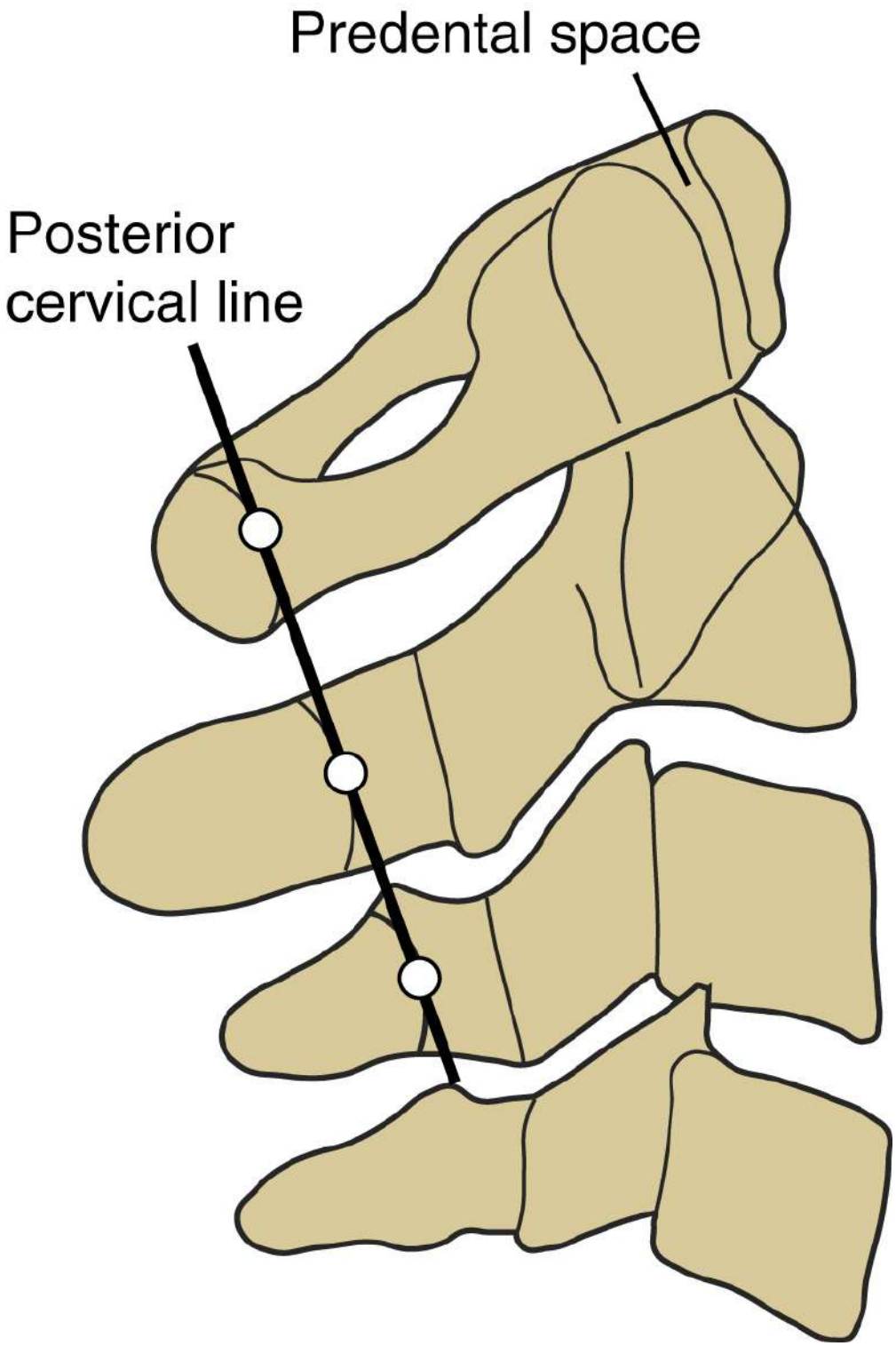

FIG. 4.3.4 Posterior cervical (Swischuk) line.

• If on assessment the patient is alert, not affected by drugs and has no other major injuries, with no posterior cervical tenderness or no pain on

movement, and having no neurological signs on clinical examination, then the immobilisation can safely be removed, and no further investigations are needed. If these criteria are not all satisfied or assessment is incomplete, immobilisation should be maintained and a three-view plain X-ray series may be performed.[19,20]

- If the X-rays are normal then the patient should be re-examined. If the patient has a normal examination, the immobilisation can be removed. But if there is still significant pain or tenderness, a CT scan of the focal region may be performed. However, correlation between clinical estimation of spinal level and radiology can be inaccurate. If this is normal and the patient is alert and cooperative, immobilisation should be kept in place and flexion and extension views performed 3–7 days later, once the muscle spasm has resolved. If the patient is significantly unwell, or has multiple trauma, CT or MRI scanning should be considered. Consultation with a paediatric orthopaedic surgeon or neurosurgeon is recommended.[19] Immobilisation should be maintained in the emergency department. Regular reassessment by the treating surgical team is required (within 12 hours) and MRI ordered if the spine cannot be cleared clinically.
- A normal CT scan and MRI scan provide definitive evidence on absence of unstable injuries. Cervical spinal precautions may be ceased with a normal MRI.

Atlantoaxial rotary subluxation

Fixed rotary subluxation at the atlantoaxial joint is more common during childhood than adulthood. It can present after minor trauma, in conjunction with an upper respiratory tract infection, and often no inciting cause is found. The clinical picture is that of the head turned to one side and held in the ‘cock-robin’ position. The child is unable to turn the head past midline, and any attempt to move it may often cause pain. The spasm of the sternocleidomastoid (SCM) muscle is on the side to which the head is turned (ipsilateral side), as the muscle is trying to right the head. In contrast, in a wry neck the SCM spasm is on the side opposite to which the head is turned (contralateral side), with the spasm causing the head turning.[1,16,18]

Plain radiographs may be diagnostic, revealing the lateral mass of C1 rotated anterior to the odontoid on the lateral view or rotation of the spinous processes to

the ipsilateral side on the AP view. If clinical examination and plain radiographs cannot confirm the diagnosis, CT imaging should be considered.[18]

Rotary subluxation of short duration will often spontaneously reduce; those that do not or that have been present for a longer duration (days) may need traction or manipulation to reduce. Post-reduction immobilisation is needed to maintain reduction, and the duration of immobilisation should vary according to the duration of subluxation.[1,18,33]

Thoracic and lumbar spine injuries

Fractures of the thoracic spine account for 25–30% of spine injury in children, while lumbar fractures account for 20–25%. Injuries to the thoracic spine and the thoracolumbar junction have a higher incidence of spinal cord injury, with neurological deficit seen in up to 40% of cases.[1,8] The high incidence of cord injury is related to the relatively large size of the spinal cord in the thoracic region, the inherent stability of the thoracic spine requiring larger forces to cause bony injury. Multiple level injuries are seen in 30–40% of children with thoracic or lumbar spine fractures.[8] Fractures of the lower thoracic and upper lumbar spine have associated small bowel and visceral injury in up to 50% of cases.[1] Road traffic accidents and falls account for most of the injuries, but non-accidental injuries in these regions do occur. Several authors have reported two frequency peaks for thoracolumbar fractures in children: between 0 and 5 years of age and then after 10 years.[34,35]

Mechanism of injury

Injuries to the thoracic and lumbar spine are flexion/extension or vertical compression injuries; often a combination of these is present, and also a degree of distraction is not uncommon.

A number of bony injuries are caused by these mechanisms. Compression of the vertebral body with anterior wedging is the most common. However, a degree of anterior wedging is normal in children. Burst fractures occur where there is disruption of the endplates and herniation of the disc into the vertebral body. Retropulsed fragments can cause spinal cord injury. The ‘seat-belt’ fracture, associated with the lap-only seat belt, is a hyperflexion and distraction injury most commonly seen at the thoracolumbar junction and is frequently associated with visceral or mesenteric injury. The posterior bony or ligamentous

elements are disrupted, and the fracture line extends horizontally into the vertebral body or through the intervertebral disc.[1,4,8,36]

Clinical assessment

As with all trauma patients, assessment of the spine should take place in the secondary survey after the airway, breathing and circulation have been assessed and stabilised. A history of mechanism of injury is important in identifying the risk of thoracic or lumbar spine injury. The search for a 'breath arrest' sensation at the moment of injury improves early detection of thoracolumbar spine fractures in children.[37] The presence of pain in the back makes injury more likely, but absence of pain does not exclude injury. Clinical examination should focus on tenderness and signs of bruising or deformity over the spine. This is assessed by log rolling the patient while spinal immobilisation is still in place. A search for signs of spinal cord or cauda equina lesions should also be made.

Any patient who has pain or tenderness over the spine should have the spine evaluated by radiography. Patients without pain or tenderness who have altered conscious state or other significant injuries are at risk of having thoracic or lumbar spine injuries missed (as they are for cervical spine injuries).[38] As multilevel injuries are common, any child with a proven cervical spine fracture or spinal cord injury should have the entire spine imaged with plain radiographs (see Fig. 4.3.5).

Radiographic evaluation

After clinical evaluation, radiographs should be taken on any child who is at risk of having a thoracic or lumbar spine injury (Box 4.3.3). The standard views for both areas are the anterior-posterior radiograph and the lateral film. In the thoracic region the radiographs need to be overexposed compared to a normal chest X-ray to allow adequate views of the spine. The shoulders often obscure the upper thoracic spine, and a swimmer's view may be needed to visualise the first two thoracic vertebrae.

The films should be evaluated by following the anterior and posterior vertebral body lines and the spinolaminar line on the lateral view. These three lines should have a parallel course. The height of each vertebral body should be assessed anteriorly and posteriorly and a difference of more than about 3 mm treated as pathological. On the AP view the paraspinal lines should be closely

inspected to detect evidence of paraspinal haematoma. The posterior elements should be visible through the vertebral body and should be in alignment. Each vertebra should be inspected; an apparently empty or invisible vertebral body indicates a fracture dislocation with distraction.[1,36]

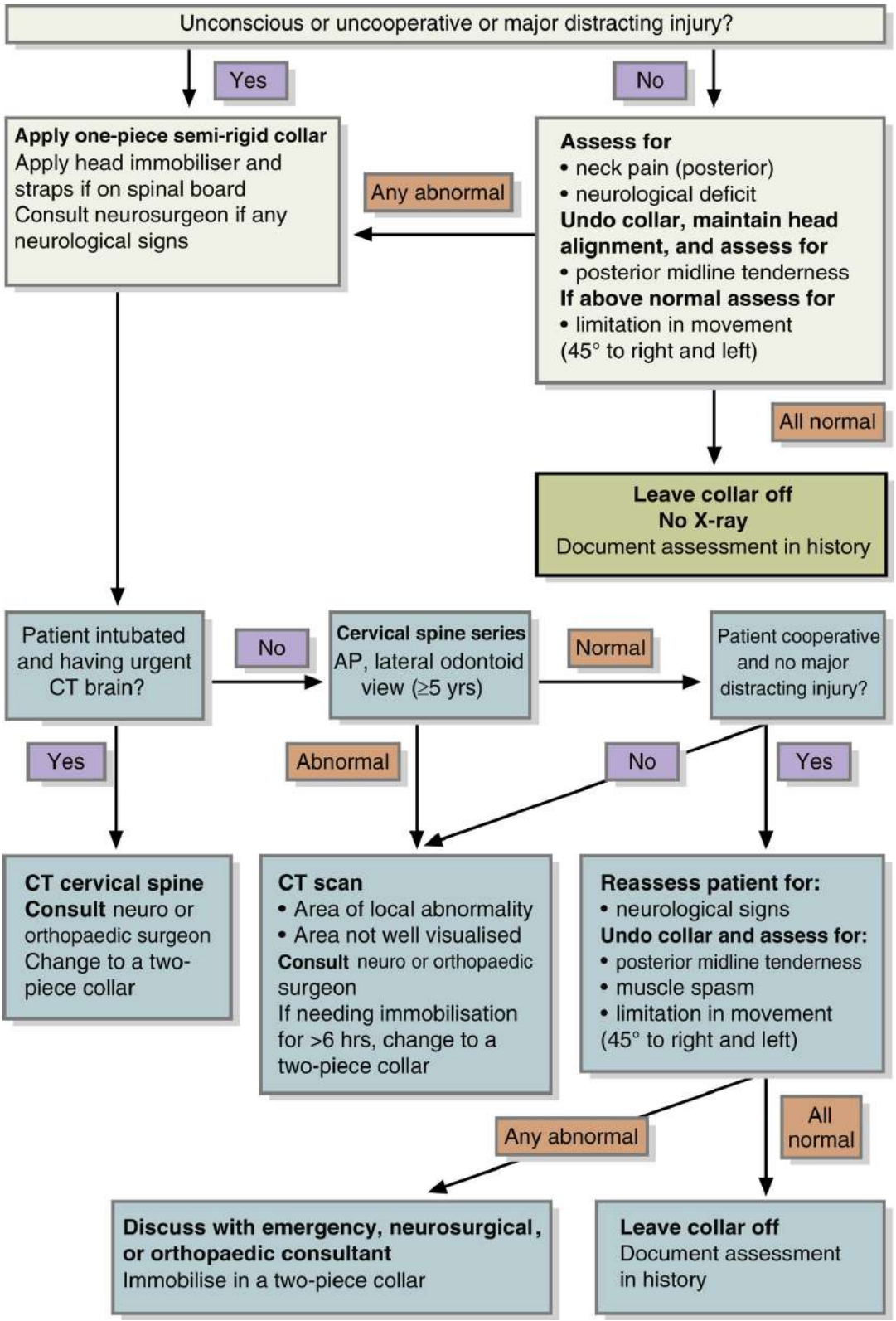

FIG. 4.3.5 Guide to management of the potentially injured cervical spine.

Box 4.3.3 Indications for thoracolumbar spine X-rays

- Pain in the thoracic or lumbar region
- Tenderness of the spine in the thoracic or lumbar region
- Significant bruising or deformity of the spine
- Altered conscious state
- Proven fracture in another region of the spine

There are a number of features on the radiographs that indicate an unstable fracture: vertebral body collapse with widening of the pedicles; greater than 33% compromise of the spinal canal by retropulsed fragments of the lamina, pedicles or body; translocation of more than 2.5 mm between vertebral bodies in any direction; bilateral facet joint dislocation; or greater than 50% anterior compression of the vertebral body associated with widening of the interspinous space.[36]

While most thoracic and lumbar spine fractures are diagnosed on the initial plain radiographs, these films often do not provide enough information on the extent of the injury, and a CT scan of the region is usually obtained to elucidate the full extent of the injury. MRI will be needed to visualise all ligamentous and spinal cord involvement.

Some fractures in the thoracic region can be difficult to see on plain films, especially if there were technical difficulties in obtaining the films in multiply injured patients. A number of secondary signs of thoracic spine injury exist and can help make the diagnosis. A paravertebral haematoma can usually be seen at the site of injury, blood in the pleural space may be seen as a pleural cap, and widening of the mediastinum may be present. Unfortunately these signs cannot differentiate a thoracic spine injury from an aortic arch injury.[8]

Management

Management must start with care of the airway, breathing and circulation. Only once these areas have been stabilised should management of the spine proceed. However, while the patient is being stabilised the spine should be maintained in alignment and the patient moved by log rolling. A thorough assessment and

investigation of the abdomen and chest are mandatory for all patients with significant thoracic and upper lumbar spine injuries, and injuries to the pelvis must not be forgotten with lumbar spine injuries. As many of these injuries are associated with intraabdominal injuries an ileus is common and nasogastric or orogastric tube should be inserted. Consultation with a paediatric orthopaedic surgeon or neurosurgeon should be sought for the definitive care of the injury. Surgical stabilisation is usually required for unstable fractures and those fractures associated with neurological injury.[4,8,36]

Spinal cord injury

The goal of management of injury to the spinal cord or cauda equina is to minimise the resulting neurological deficit. This commences with adequate immobilisation of the spine and complete and thorough investigation to detail the anatomy of the injury.[39]

Spinal cord injury should be suspected in any child who has multisystem trauma, minor trauma associated with spinal pain, or sensory or motor symptoms and any patient with altered consciousness. The patient must be adequately immobilised, such as on a spinal board with a cervical collar and head immobiliser (see section on spinal immobilisation) and any assessment of the back or patient movement accomplished by log rolling. Up to 50% of patients with spinal cord injury will have at least moderate head injury.[39]

Spinal cord injury syndromes

A number of patterns of neurological deficit are seen in patients with spinal cord injury, with the deficit dependent on the portion of the spinal cord damaged. Neurogenic shock – the manifestations of loss of sympathetic output to the cardiovascular system – is seen immediately after complete cord injury at the level of T6 or above. This should not be confused with spinal shock, which is the reversible dysfunction of the spinal cord associated with injury. It is likened to a concussion of the cord without permanent damage. It may exist alone or in combination with permanent cord injury. Its resolution is responsible for the improvement in neurological function seen in the first few days post-injury.[16,39]

The distinction between a complete injury and a partial injury – with preservation of some motor or sensory function below the lesion – is vital for prognosis. Many patients with partial injury will regain much or all of the

neurological function. A partial cord injury may occur at any region and with any mechanism of injury.

Complete cord injury is usually seen in injuries of the thoracic spine and thoracolumbar junction. The spinal cord is large in relation to the size of the spinal canal at this level. Complete cord injuries that remain at 24 hours rarely regain any significant function. Cauda equina lesions – injuries at or below L2 – involve the peripheral nerves rather than the spinal cord and can show significant recovery of lower limb and sphincter function even weeks after the injury.[16,39]

Central cord syndrome is usually seen in hyperextension injury resulting in herniation of the intervertebral disc into the spinal cord. The resulting injury causes a motor deficit that is greater in the arms than legs and most extensive in the small muscles of the hand. The sensory deficit is variable. Brown–Sequard syndrome – hemisection of the cord – causes a contralateral loss of pain and temperature sensation and an ipsilateral motor paralysis and loss of proprioception below the level of injury. Approximately two-thirds of those with central cord syndrome and one-third with Brown–Sequard syndrome will recover.[16,39]

Clinical assessment

Initial steps in the management of a patient with a suspected spinal cord lesion are the assessment and resuscitation of the airway, breathing and circulation. The most immediate threats to life and spinal cord function of patients with spinal cord injury remain hypoxia and hypotension. Spinal cord lesions in the upper cervical spine may impair respiratory function and require early intubation and mechanical ventilation. The unstable cervical spine must be maintained in alignment without traction during treatment of the airway. The loss of sympathetic vasomotor tone after cervical spinal cord injury will result in vasodilatation, venodilatation, and reduced venous return to the heart causing hypotension. There should be an associated relative bradycardia for age and existing blood pressure, which will help distinguish this response from haemorrhagic shock.[39]

Initial fluid resuscitation with 10–20 mL kg^{-1} should adequately replace the relative hypovolaemia. However, if hypotension persists, measurement of central venous pressure (CVP) may be needed to guide fluid replacement. Excessive fluid replacement that pushes central venous and pulmonary artery pressures above the normal range will result in pulmonary oedema. Treatment with a

vasoconstrictor, such as metaraminol, may be useful for the patient who has adequate CVP and remains hypotensive. In patients who are also significantly bradycardic, inotropic agents, such as dopamine or adrenaline (epinephrine), may be useful. Patients requiring more than 40 mL kg^{-1} of fluid replacement and having a low CVP must be assumed to have other injuries causing blood loss.

Examination of the neurological impairment is done as part of the secondary survey. A thorough examination of the motor function of the limbs and assessment of reflexes should be performed and a level of sensory deficit sought. Knowledge of the dermatomes and myotomes will allow determination of the level of neurological injury.

Radiographic evaluation

In all patients with suspected spinal cord injury the spine should be X-rayed. The radiographs should include the entire spine, as multiple levels of injury are common. Once the patient is stabilised, further investigation of the lesion should follow. The bony injuries should be investigated as discussed above. Investigation of the cord itself will require MRI. MRI should be performed as soon as possible after identification of a spinal cord injury, as it will allow identification of remedial intraspinal problems in patients with a partial neurological deficit. The appearance of the spinal cord on MRI also allows prediction of neurological outcome. Cord transection and major haemorrhage have a poor outcome, minor haemorrhage and oedema have a moderate to good outcome, and a normal MRI is associated with complete recovery.[1]

Treatment

Most of the treatments available for spinal cord injuries are supportive. Breathing and circulation must be supported as needed. As there will be a neurogenic bladder, catheterisation is necessary, and a nasogastric tube is needed to treat the gastric and bowel stasis that ensues. For transport, antiemetic is useful to prevent vomiting and spine movement or airway compromise. Subcutaneous low-molecular-weight heparin should be instigated once the patient is stable to prevent deep venous thrombosis.[40]

Specific treatment of the spinal cord lesion is controversial. Four substances have been studied in prospective, randomised trials – methylprednisolone, tirilazad, naloxone and GM-1 ganglioside. All studies to date have excluded

children under 13 years of age. Tirilazad and naloxone have failed to show any benefit in trials to date. There is conflicting evidence regarding the benefits of methylprednisolone and documented evidence that its use increases the risk of bacterial infection. There is limited evidence to support use of steroids in spinal cord injury. It can be considered in patients with isolated non-penetrating spinal cord injury in consultation with local spinal unit.[16,40] GM-1 ganglioside has yet to be shown to offer significant benefit in spinal cord injury and is not recommended for routine use.

Once the patient has been stabilised and investigated, transfer to a spinal cord injury unit should be expedited. These units and associated intensive care units are geared to manage the cardiorespiratory compromise that may occur in the ensuing weeks, the psychosexual issues that accompany spinal cord injury, the urological problems, and the potential for skin breakdown that are exaggerated in these patients.

Spinal cord injury without radiographic abnormality

Spinal cord injury without radiographic abnormality (SCIWORA) is defined as objective signs of myelopathy as a result of trauma with no evidence of fracture or ligamentous instability on plain X-rays or tomography.[41] SCIWORA is most frequently seen in younger children (especially <8 years of age) and in injuries of the cervical spine. Postulated causes include ligamentous laxity and bony immaturity allowing excessive, transient movement during trauma, causing distraction or compression of the spinal cord, or cord ischaemia due to vascular injury or hypoperfusion. The incidence reported in children is 1–10% of all spinal cord injuries.[1,16,42,43]

Younger children tend to have more profound neurological injury and hence less long-term improvement.[42,43] A number of children will present with minor neurological injury and progress to complete or partial spinal cord injury. The incidence of this delayed presentation of the serious symptoms is 5–50%. The delay to presentation of full symptoms has been as long as 4 days.[1,3] Because of these presentations, all children with history of neurological symptoms or any neurological deficit should be treated as patients with potential spinal cord injuries.

After the primary survey, resuscitation and secondary survey and radiographic evaluation, any patient with any neurological deficit should remain immobilised

until all bony, ligamentous and spinal cord injury is excluded or treated. Further investigation with a CT scan focused at the level of symptoms and MRI to view the cord should be performed. MRI provides the same prognostic information in SCIWORA injuries as in other spinal cord injuries.[16]

Controversies

- The clinical assessment of potential spinal injuries in the preverbal child can be a challenge.
- There is an ongoing debate about how to adequately immobilise the cervical spine of a child less than 2 years of age.
- The use of flexion and extension films in the acute situation is controversial.
- The role of routine CT of the cervical spine in major trauma has been suggested but needs to be balanced against the radiation exposure to children especially involving whole spine screening.
- The timing of removal of spinal immobilisation in the unconscious child remains controversial.
- The role of methylprednisolone in spinal cord injury remains unproven despite one large study suggesting time-bound benefit.

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4.4

Thoracic injuries in childhood

Soundappan S.V. Soundappan, and Dermot Thomas McDowell

ESSENTIALS

1 Thoracic injuries occur in 4–6%[1] of children admitted with trauma, usually due to blunt mechanism, and are often associated with other injuries.

2 Thoracic injuries are the second commonest cause of death after head injuries.[2]

3 The pathophysiological differences in children depend principally on their age and size. These must be appreciated in order to optimise outcome.

4 The most common injuries are pulmonary contusions, rib fractures, pneumothoraces and haemothoraces. These can usually be managed by a combination of adequate oxygenation, IV fluids, thoracostomy tube drainage and analgesia.

5 Injuries to the aorta and diaphragm may be clinically occult, and chest X-ray findings are often non-specifically abnormal. Further diagnostic evaluation is always indicated in suspect cases.

6 Surgical intervention is uncommonly required but is indicated in cases of massive haemothorax, cardiac tamponade, and major airway and oesophageal injury, as well as in aortic and diaphragmatic injuries.

7 Emergency department thoracotomy is principally indicated in cases of penetrating chest trauma and an arrest or pre-arrest presentation.

Introduction[3–6]

Traumatic injury is the most common cause of morbidity and mortality in childhood, and thoracic injuries are second to head injuries as a cause of mortality. Isolated chest injuries have a mortality of around 5%, but this increases substantially when combined with head and/or abdominal injury to as high as 20%. Injuries to the great vessels and bronchi, lung lacerations and cardiac tamponade are the chest injuries most likely to cause early death. Small children provide a small target in blunt trauma, so multiple injuries should be expected.

A recent review of an Australian trauma registry database found multiple body region injury to be almost universal (99%) in cases of severe blunt chest trauma. The most frequent associated serious injuries were to the head (46%), lower extremity (32%) and abdominopelvic injury (30%). By far and away the commonest chest injuries were pulmonary contusion, haemopneumothorax and rib fractures.

The mechanisms of injury vary with the age of the child. Overall, the majority (60–80%) are due to blunt trauma and involve a motor vehicle. In infants and toddlers, common mechanisms include being injured as passengers in motor vehicle crashes (MVC) or as pedestrians struck or run over by a vehicle (commonly in the driveway of the family home). Falls (from stairs, balconies, etc.) occur mainly in this age group. Child abuse also tends to predominate in this age group and should always be considered. In school age children, motor vehicle and bicycle-related traumas are common, and sporting (including extreme sports) injuries increase in frequency with age. With adolescence the occurrence of penetrating trauma emerges with an associated increased mortality risk; also, inexperienced teenage drivers have an increased incidence of MVC. Drug and alcohol intoxication is often associated with personal/interpersonal violence in this older age group.

There are a number of anatomical and physiological features of small children that must be appreciated when managing paediatric chest trauma. These are summarised in Box 4.4.1.

Initial approach in the emergency department[1,7,8]

Initial management follows the usual priorities. After ensuring airway patency, breathing should be assessed. High flow oxygen should be applied. Signs of

respiratory compromise and tension pneumothorax should be managed by needle or finger thoracostomy prior to chest X-ray (CXR) followed by intercostal catheter (ICC) insertion. A large haemothorax may compromise ventilation as well as circulation, requiring early ICC placement and fluid resuscitation, whilst an orogastric tube (OGT) should be placed early to decompress the stomach, as gastric distension may compromise ventilation. Mechanical ventilation should be instituted for signs of ongoing respiratory distress/respiratory failure not relieved by optimisation of oxygen delivery, chest tube insertion, closure of open chest wounds and OGT placement. Ongoing signs of circulatory compromise without evidence of blood loss should raise the possibility of cardiac tamponade and myocardial contusion in a child with chest injuries. A portable CXR should be the first radiological test ordered. CXR shows abnormalities in 60–95% of children with significant thoracic injuries. FAST (Focused Assessment with Sonography in Trauma) should occur early during resuscitation of a child, where available, and imaging of the pericardium should always be included to detect haemopericardium. Controversies around FAST in the paediatric population are discussed in Chapter 4.5 (Abdominal and pelvic trauma). Computerised tomography (CT) can demonstrate lung contusion, rib fractures and unsuspected injuries in 15% of children with normal CXR. CT imaging of the chest should be used selectively in children to reduce risk from radiation. It is indicated in high-impact trauma, when multiple injuries are present or there are suspected positive findings on clinical examination or CXR. The vast majority of chest injuries in childhood can be managed non-operatively. Drainage of pericardial blood may occasionally be performed in the emergency department (ED) in an unstable patient if operative intervention is not immediately available. Other indications for operative intervention are listed in Box 4.4.2.

Box 4.4.1 Important pathophysiological differences between children and adults in chest trauma

- Small target – multiple injuries common
- Narrow airway – prone to obstruction
- Anterior larynx – difficult intubation
- Increased surface area – increased risk of hypothermia
- Increased oxygen consumption, diaphragmatic breathing, low functional residual capacity – prone to hypoxia

- Increased pliability of ribs – decreased incidence of rib fractures
- Response to blood loss – hypotension is a late sign of shock
- Developmental considerations – paediatric coma scoring

Chest wall injury[9,10]

Rib fractures

The elasticity and flexibility of the younger child's chest wall lead to a lower incidence of rib fractures. Significant underlying intrathoracic injury can occur in the absence of rib fractures. In the 0- to 3-year age group rib fractures should raise the concern of abuse: in one study 2/3 of 0–3-year-olds with rib fractures were victims of abuse, and a careful assessment of all aspects of the clinical presentation is mandatory. Radiological findings suggestive of abuse include multiple fractures, fractures of varying ages and bilateral fractures. A bone scan may be a more sensitive test in the setting of potential abuse.

Rib fractures in children are a marker of potential severe associated injuries. Multiple rib fractures (>1) increase the risk of severe intrathoracic injury, multiple injuries and mortality. Fracture of the first rib requires significant force, mandating a high degree of suspicion of associated injuries to the great vessels and the trachea, though the incidence of such injuries is very uncommon. Lower rib fractures are often associated with liver, spleen and diaphragmatic ruptures. The risk of mortality increases linearly with rib fractures, from associated injuries that are often severe.

Flail chest injuries are rare in children and clearly indicate serious injury, with reduced ventilatory effectiveness and associated lung contusions contributing to the significant potential for respiratory failure. Assisted ventilation is indicated for those with respiratory failure despite optimal non-invasive ventilation and analgesia or associated injuries, particularly to the head.

Management of rib fractures involves analgesia, treatment of associated injuries, ongoing assessment of the child's respiratory status and close observation for complications that may arise. Analgesic options in the ED include oral paracetamol (also available intravenously [IV]) and anti-inflammatories, intranasal fentanyl and titrated IV narcotics. Prevention of atelectasis and pneumonia is a priority.

Box 4.4.2 Indications for operative intervention in chest trauma

- Great vessel injury
- Pericardial tamponade
- Large haemothoraces
- Tracheobronchial injuries
- Oesophageal injury
- Diaphragmatic lacerations
- Open pneumothorax with major chest wall defect
- Penetrating chest trauma that crosses the mediastinum

Pulmonary injury[11,12]

Contusion

In children it is important to appreciate that significant lung injury can occur in the absence of rib fractures or other external signs of chest wall injury. Pulmonary contusion most commonly results from blunt injury and requires significant force. Pathological findings are intra-alveolar haemorrhage, consolidation and oedema causing ventilation-perfusion mismatch, reduced lung compliance and subsequent hypoxaemia. They are usually asymptomatic but can cause respiratory compromise when extensive. Initial CXR could be normal in 60–90% and changes may appear over the next 4–6 hours. CXR findings of increased pulmonary opacity may be present in the ED but could evolve over time. CT scanning of the thorax may reveal contusion that is not evident on the initial plain films. Approximately half of the CXRs showing pulmonary contusion show other abnormalities, most commonly fractured ribs and pneumo/haemothoraces.

Management of pulmonary contusion involves the following:

- Maintenance of oxygenation. Supplemental O_2 is initially delivered by high flow mask (10–15 L min^{-1}). Persisting hypoxaemia requires non-invasive or invasive ventilatory support depending on severity and

associated injuries.
- Pain relief should be provided as indicated.
- Excessive intravenous fluids should be avoided.
- Respiratory physiotherapy is useful.

Both acute respiratory distress syndrome and pneumonia may complicate pulmonary contusion.

Pneumothorax[6,13,14]

Traumatic pneumothoraces vary in their size and clinical significance. They occur in about one-third of children with significant thoracic trauma, and associated injuries are common. All pneumothoraces should be considered as having the potential to cause cardiorespiratory compromise. The clinical signs of pneumothorax (PTX) vary from nothing to decreased air entry, hyperresonance and subcutaneous emphysema.

Small to medium-sized pneumothoraces may not be visible on a portable supine CXR. Ultrasound of the chest may be incorporated into the FAST scan protocol to detect pneumothoraces and haemothoraces. The sensitivity of ultrasound in detecting these complications of chest trauma is superior to supine CXR, but CT scanning remains the gold standard. Subtle signs on CXR include increased radiolucency on the ipsilateral side and a deep sulcus sign. Small pneumothoraces are commonly revealed on CT scan of the chest and/or abdomen. The significance and hence management of these small pneumothoraces are debated. A small uncomplicated PTX in a stable patient with isolated chest trauma, who is not likely to require positive pressure ventilation or prolonged transport (particularly aeromedical), can be considered for observation, high flow O_2 and analgesia in a high-dependency unit setting. Most other traumatic pneumothoraces require the insertion of a formal chest drain.

Tension pneumothorax

This is a clinical condition resulting from increasing intrathoracic pressure, lung collapse and mediastinal shift with subsequent impaired gas exchange, decreased venous return and cardiovascular collapse. The diagnosis is clinical, and treatment should precede radiology in clear-cut cases. Signs are of reduced air entry, hyperresonance and hyperexpansion plus reduced movement of the

affected side. The signs of tracheal deviation and elevation of the jugular venous pressure (JVP) may be difficult to detect in children who have short necks. JVP elevation may also be absent if there is associated hypovolaemia. Patients are always tachypnoeic with respiratory distress and tachycardia, but hypotension is a late sign if solely due to a tension PTX. Immediate needle decompression with a 16G cannula inserted into the 2nd intercostal space in the mid-clavicular line should occur if finger thoracostomy and formal intercostal tube insertion via a lateral approach are likely to be delayed. In rare situations when a chest drain is not available needle drainage or a glove finger as a unidirectional valve can be life-saving. It is important to note that post mortem CT frequently identifies misplacement of needle thoracostomies outside the pleural cavity.

However, hypotension and/or hypoxaemia may have other causes in the traumatised child. The differential diagnosis of a tension PTX includes haemorrhage, pericardial tamponade, haemothorax (which may cause tension), pulmonary contusion and air embolism. Common, easily preventable/treatable causes that may occasionally be confused with a tension PTX are intubation of the right main stem bronchus and gastric distension. Endotracheal tubes (ETT) must be inserted the appropriate distance (age/2 + 12 cm) and movement of the child's neck minimised. This is particularly so in small children where neck flexion (tube pulled out) or extension (tube down the [R] main bronchus) may result in the malposition of the ETT. An OGT should be placed early.

Open pneumothoraces may occur with penetrating chest trauma. Respiratory compromise relates to the effects of the PTX, underlying lung injury and a 'sucking chest wound' if the defect is large enough. If the diameter of the chest wound is approximately two-thirds or greater than that of the trachea, air will preferentially be sucked into the chest on inspiration, leading to acute severe respiratory compromise. Management requires urgent wound coverage on three sides only with an occlusive dressing, to prevent the development of an iatrogenic tension PTX, and chest tube placement away from the wound. Once the chest drain is in place the wound can be sealed and arrangements made for definitive surgical care. Significant ongoing respiratory distress is an indication for mechanical ventilation.

Pulmonary lacerations [7]

Pulmonary lacerations are principally caused by penetrating injuries but can occur with blunt mechanisms especially associated with rib fractures. They

usually result in a haemothorax (sometimes massive) or PTX and rarely can be complicated by air embolism. Air embolism typically occurs after the initiation of positive pressure ventilation and causes sudden haemodynamic deterioration with or without focal neurological signs. Development of a tension PTX, pericardial tamponade or massive haemothorax is a main other possibility in the chest for such a sudden deterioration. Lung hyperinflation due to overly aggressive positive pressure ventilation may also cause cardiorespiratory compromise and risk barotrauma.

In the setting of deterioration after initiation of ventilation, by far the most likely cause is the development of tension PTX, which should be managed as above. In the rare event of air embolism, if suspected, management includes 100% oxygen and reduction of the ventilation pressures. Emergency thoracotomy with clamping of the hilum on the affected side and aspiration of ventricular air has occasionally been life-saving. Small lacerations usually heal spontaneously while large lacerations with persistent air leak or bleeding require surgical repair.

Haemothorax[1,13,15]

Clinically relevant haemothoraces occur in about 15% of cases of blunt chest trauma but are more common if the injury is penetrating. The bleeding most commonly originates from lacerations to the lung, intercostal or internal mammary vessels or occasionally from mediastinal vessel injury (often fatal). Each hemithorax can hold up to 40% of a child's blood volume.

The clinical presentation is of varying degrees of hypovolaemia and respiratory compromise depending on the amount of blood lost into the chest, associated PTX and the development of increased intrathoracic pressure. Chest examination reveals reduced air entry and dullness to percussion with or without signs of tension. Management is with an ICC. Drainage of massive haemothoraces may precipitate further bleeding as the tamponade effect is removed.

Blood loss from an ICC, haemodynamic response to resuscitation, mechanism of injury (blunt vs. penetrating) and associated injuries (especially head) are used by cardiothoracic surgeons in determining the need for thoracotomy.

Indications for thoracotomy include:

- initial drainage >15 mL kg^{-1} of estimated blood volume

- continued bleeding >1–2 mL kg^{-1} hr^{-1}
- increasing bleeding
- significant residual haemothorax post-tube drainage.

Management involves oxygenation and ventilatory support if indicated, urgent chest tube placement of the appropriate size via the lateral approach directed posteriorly and volume resuscitation. The concept of hypovolaemic resuscitation in uncontrolled traumatic haemorrhage has not been evaluated in children. However, it is important to consider early surgical intervention in any patient who is haemodynamically compromised due to haemorrhage or showing signs of ongoing bleeding.

Longer-term complications of haemothoraces include haematoma organisation with secondary lung entrapment and empyema formation. Retained blood should be evacuated in a week by thoracotomy or thoracoscopy to avoid entrapment of lung and infection. Prophylactic antibiotics are indicated when chest tubes are placed for penetrating trauma. The adult trauma literature suggests a reduced infection rate even in previously closed traumatic haemothoraces requiring drainage.

Tracheobronchial injuries[1,14,15]

These uncommon injuries may occur with penetrating or severe blunt trauma and have a high mortality if not recognised and treated rapidly. In blunt trauma intrathoracic airway injuries usually occur near the origin of the main stem bronchi. Typically they present with respiratory distress and signs of subcutaneous (SC) emphysema, pneumomediastinum and a tension PTX. Haemoptysis also occurs. In the case of PTX urgent chest tube placement is required, and typically a large air leak will continue, and there will be failure of lung expansion on CXR. At this point an airway injury is usually considered. A second chest tube should be placed and urgent cardiothoracic surgical consultation obtained. Once stabilised, a CT scan may be helpful in further injury delineation and assessing lung inflation, as massive SC emphysema can make CXR interpretation very difficult. Bronchoscopy is useful as a diagnostic modality in suspected tracheobronchial injury, and operative intervention is often required. Mortality rate is very high with most occurring within the first hour.

Mediastinal injury[1,7,13,15]

Aortic transection

Aortic rupture is a rare event in young children, but the incidence increases with adolescents involved in high-speed MVCs. About 80% occur at the aortic isthmus just distal to the origin of the left subclavian artery. Most are rapidly fatal at the scene, and 30% of those who make it to hospital die within 6 hours. Diagnosis in hospital depends on clinical suspicion based on mechanism of injury, physical signs (often absent) and CXR findings. Physical signs indicative of aortic rupture include first rib fractures, sternal fractures, paraplegia, upper limb hypertension or lower extremity pulse deficit. CXR in cases of aortic rupture is usually abnormal, but findings are non-specific (Box 4.4.3), and the suspicion of aortic injury on clinical or radiological grounds requires further diagnostic imaging.

In young children the normal thymic contour may give the impression of a widened mediastinum. Further imaging usually involves CT angiography, aortogram or transoesophageal echocardiography depending on availability, expertise and local practices. The absence of signs of dissection and mediastinal haematoma on CT angiography is used to exclude aortic injury. Occasionally, formal aortography or transoesophageal echocardiography will be performed to exclude possible aortic injury.

Box 4.4.3 Chest X-ray signs of aortic injury

- Widened mediastinum (mediastinum to chest ratio >0.25)
- Loss or abnormal contour of aortic knob
- Depression of left main stem bronchus
- Deviation of the trachea to the right
- Deviation of the oesophagus (nasogastric tube or orogastric tube) to the right
- Left pleural cap
- Left haemothorax
- Upper rib fractures

Management in confirmed cases is surgical. β-blockers may be commenced preoperatively in haemodynamically stable patients to reduce vessel wall stress.

Non-aortic injuries are rare and experience is often limited to case reports in literature.

Cardiac injuries[16]

As with aortic injury, clinically significant cardiac injury from blunt trauma in children is uncommon and is usually associated with other intrathoracic injuries. Pericardial tamponade can certainly occur with blunt trauma, though it is more common in penetrating injuries (see below). Myocardial contusion may manifest as an arrhythmia or otherwise unexplained tachycardia and/or hypotension. Valvular injury and septal defects have also been reported. Diagnosis suffers from the lack of a gold standard and the questionable clinical relevance of test results. Most of the evidence comes from adult trauma patients. A normal ECG has a high negative predictive value for the occurrence of clinically significant complications in suspected myocardial contusion. Evidence for the value of cardiac markers is lacking. Echocardiography is a very useful modality in assessing suspected clinically significant myocardial contusion such as the presence of unexplained hypotension, tachycardia or new murmurs.

In the absence of ECG abnormalities, hypotension or new murmurs, ongoing ECG monitoring is usually not required. Treatment is supportive with inotropes as needed until spontaneous recovery.

Commotio cordis[17,18]

The phenomenon of sudden cardiac arrest following a localised blow to the chest is well documented in children. In these cases autopsy fails to identify myocardial contusion, structural cardiac abnormality, conduction system or coronary artery pathology. The proposed theory is that a blow to the chest during the vulnerable phase of the electrical cycle induces ventricular fibrillation/ventricular tachycardia. Protective chest guards are recommended in at-risk sports. Prompt defibrillation may be life-saving.

Penetrating cardiac trauma[19]

In children this occurs predominantly in the adolescent age group. Cardiac

lacerations may lead to rapid exsanguination or pericardial tamponade. Any penetrating chest or upper abdominal wound has the potential to injure the heart. Clinical signs of tamponade include tachycardia and elevation of the JVP (in the absence of hypovolaemia), with subsequent hypotension and cardiac arrest with pulseless electrical activity. The CXR is typically normal in the absence of associated mediastinal or lung injury. In trained hands and with satisfactory imaging conditions, FAST/echocardiography has excellent accuracy in the detection of pericardial blood and can be done rapidly in the ED. Management requires urgent cardiothoracic surgical involvement. A conscious patient with a perfusing blood pressure requires urgent surgery. A rapidly deteriorating patient in the ED requires needle pericardiocentesis if there is any surgical delay. Cardiac arrest with vital signs present at the scene, and a short transit time to hospital or arrest in the ED is an indication for ED thoracotomy or pericardiocentesis depending on the skills of personnel available. Pericardiocentesis is performed using a long 16 or 18G over-the-needle cannula via the subxiphoid approach at a 35 degree angle to the skin and aiming at the left shoulder with ECG monitoring. Ultrasound control may assist where available. Aspiration of 10–20 mL may result in significant clinical improvements. The needle should be removed, but the catheter should remain in place for repeat aspirations. Failure to aspirate blood does not exclude tamponade as the cannula may miss the pericardium, or the pericardial blood may have clotted.

Diaphragmatic injury[13,15,20]

Diaphragmatic injury is an uncommon paediatric injury, but it is important nonetheless, as undiagnosed, complications eventually will arise, though this may take years. Left-sided injury is more common than right-sided in blunt trauma, and associated intraabdominal injury is common.

Upper abdominal penetrating trauma that injures intrathoracic structures (and vice versa) must also have caused diaphragmatic injury and requires repair. Diagnosis is difficult unless CXR reveals clear signs of herniated stomach or bowel or the nasogastric tube (NGT) curling up into the thorax. More commonly the CXR is non-specifically abnormal with findings of an abnormal diaphragmatic contour with or without lower zone opacity. Often the diagnosis is made at laparotomy or laparoscopy for associated injuries. Barium studies are normal if bowel contents are not herniated. CT scanning may miss small tears.

Magnetic resonance imaging may have a role in diagnosing these injuries. Suspected occult diaphragmatic lacerations in penetrating trauma can be investigated by laparoscopy/thoracoscopy or open operation.

Oesophageal injury[7]

This is essentially only seen in penetrating trauma, and in these cases a high index of suspicion is required, as missed injuries cause inevitable serious morbidity and mortality. On initial CXR a finding of mediastinal air is an early clue. Over subsequent hours an evolving sepsis with pleural effusions and mediastinitis ensues. Conscious patients are able to verbalise complaints of chest and epigastric pain, but this is difficult to interpret in the setting of other chest injuries. Suspicion of an oesophageal injury must be followed by a water-soluble contrast study, oesophagoscopy or both. If positive, broad-spectrum antibiotics and urgent surgery are required.

Emergency department thoracotomy[19,21]

The only definite indication for thoracotomy in the ED is in the scenario of penetrating chest trauma with loss of vital signs shortly before arrival in the ED or during ED resuscitation, with the purpose of pericardial drainage, repairing penetrating injury to the heart or controlling bleeding from the hilum or lung. Open cardiac massage can also be performed as well as cross-clamping of the aorta. The universally poor outcome of blunt trauma patients who arrive at the ED with no vital signs/signs of life argues strongly against performing ED thoractomy in these patients. Blunt thoracic trauma patients who deteriorate in the ED despite full resuscitation may occasionally survive following ED thoracotomy, but in general these very unstable patients should go to the operating theatre if possible. A recent systematic review arrived at following conclusions:

1. Children with blunt trauma and no signs of life (SOL) are non-salvageable
2. Penetrating injury with SOL or without SOL a short time prior to arrival may be salvageable, and there is role for ED thoracotomy
3. No survivors under 14 following blunt trauma.

The role and benefit of this procedure in low volume paediatric trauma centres is questionable. However, trained resuscitationists operating with ultrasound findings of pericardial tamponade, even in blunt trauma, may be justified in attempting ED thoracotomy.

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4.5

Abdominal and pelvic trauma

Dominic Fleischer, and Scott Pearson

ESSENTIALS

1 The abdomen is the third most common region injured in paediatric trauma after limb and head trauma. It is the second most common region injured resulting in death, after head trauma.

2 Children may have injury to abdominal viscera with little initial clinical evidence of trauma.

3 Meticulous assessment and reassessment will detect subtle abdominal injuries through changes in physiological observations and serial examination findings.

4 The well child may be safely assessed by careful serial abdominal examinations without the need for routine CT scanning.

5 Non-operative management of solid organ injuries is the standard of care but requires institutional support and may necessitate transfer of the child to a hospital specialised in paediatric trauma care.

6 Pelvic fractures are a marker of severe injury, and there is a strong association with head, chest trauma and abdominal injury.

Introduction

Abdominal trauma is the second most common injury resulting in death after head trauma.[1] Paediatric abdominal trauma involves particular challenges to the emergency specialist. The history may be limited, the trauma seemingly innocuous, examination may initially be unremarkable or difficult to perform,

physiological observations may remain normal until a late deterioration, and there is a reluctance to use ionising radiation for imaging.

Abdominal injuries in children are almost exclusively the result of blunt trauma. While penetrating injuries increase in the adolescent population, this remains an unusual phenomenon in most Australasian communities. Abdominal injuries resulting from blunt trauma most commonly affect the solid organs, particularly the liver and spleen. Overall mortality is less than 5% for children with abdominal trauma, but obviously this depends on the specific injuries and systems involved.[2] Approximately one-third of paediatric trauma patients with an injury severity score (ISS) greater than 15 will have an intraabdominal injury.[2] In children with multi-trauma, the subtle early clinical findings of intra-abdominal injury may be masked by changes in conscious state and the presence of injuries in other body regions.

There are morphological characteristics of children that predispose them to intraabdominal injuries. The rib cage does not extend as far distally as in the adult, the ribs are more compliant, and the abdominal wall musculature is thinner. The organs are closely packed together, and there is less ‘padding’ soft tissue to absorb the kinetic energy transmitted by the impact.[5] This results in the liver and spleen being more exposed and less protected. However, there is a lower associated mortality for abdominal solid organ injuries in children within each abbreviated injury score (AIS) grade relative to adults. The most commonly injured organs are identical in both adults and children and in decreasing order of frequency are spleen, liver, kidneys, bowel, mesentery and the pancreas.

The very compliant body of the child is capable of absorbing considerable amounts of kinetic energy without revealing external signs yet be associated with significant internal derangement. Children are generally healthy, with few comorbidities and on few, if any, medications. In physiological terms, they are able to compensate extremely well for blood loss until precipitously decompensating.

The child with multiple injuries requires senior experienced clinicians involved in decision making during the assessment and resuscitation in the emergency department (ED). Such a patient invariably requires the involvement of several specialties and a trauma team approach, with clear leadership of the resuscitation. Despite less surgical intervention, surgical involvement in managing children with abdominal injury is important for ongoing observations, serial examinations and decisions involving imaging and intervention.

History

Obtaining details of the exact mechanism of injury cannot be over-emphasised. This often gives a clue to the potential injury pattern. Information can be obtained from witnesses, ambulance officers, family, friends, or caregivers. One member of the trauma team should be delegated to obtain this information so that the primary survey can occur simultaneously.

Particularly concerning mechanisms of injury in children include pedestrians or occupants involved in motor vehicle collisions, falls from a height, and direct blows to the abdomen (e.g. bicycle handlebar, seat belt and inflicted injuries). Information such as the positioning of the child when struck and likely speed of vehicles is useful in predicting injury patterns. Likewise, factors such as the use of restraint devices, type used, and wearing of a bicycle helmet, where appropriate, are helpful in defining the resulting injuries. Lap belts can be associated with rib and lumbar spine fractures and upper abdominal organ injuries. Handlebar injuries may cause serious blunt intraabdominal injuries. The resultant injuries to pancreas and duodenum can be subtle and delay the diagnosis. Hence the threshold for observation or imaging may need to be varied accordingly in children presenting with this mechanism of injury. Small children are particularly at risk of being unsighted and backed over in driveways by reversing vehicles and may sustain major internal injuries. The events leading to the fall, fall height and landing surface are all pertinent information that can usually be obtained rapidly.

The complaint of abdominal pain even without tenderness warrants a period of serial examinations.

The recognition of abuse as a causal mechanism in younger children and infants is important in patients with abdominal trauma. There may be minimal signs of external injury, and the reported history may suggest a minor incompatible mechanism or no history of injury at all. The emergency physician needs to maintain an index of suspicion in the infant who presents in shock or with an altered level of consciousness. Other information, such as medications, allergies, and significant past history, should be obtained.

Examination

Primary survey focuses on the ABCs and may result in early interventions such as intubation or treatment of shock. The examination of the abdomen is usually

delayed until the secondary survey. Where endotracheal intubation has occurred the information obtained by palpation of the abdomen is somewhat limited, and these children often require abdominal CT scanning, provided their vital signs are satisfactory and not deteriorating. Vital signs are essential. Signs of haemorrhage may be subtle initially. Particular attention should be given to the respiratory rate, pulse rate, non-invasive blood pressure, capillary refill time, urine output and mental status. Deterioration of these physiological markers may identify early hypovolaemia prior to the development of haemorrhagic shock. The blood pressure needs to be measured with an appropriately sized cuff for the child's habitus. Automated blood pressure machines, while useful in freeing staff to attend to other aspects of care, can be unreliable when hypotension exists and can result in delays in obtaining these recordings. Single vital sign recordings are of limited use, but it is the progression of recordings and the monitoring of perfusion that more accurately reflect volume status. In the critically ill child, pulse and blood pressure should be measured at 3–5-minute intervals.

The use of the terms *unstable* and *stable* is discouraged when conveying information regarding the child's status to colleagues. They are non-specific and are defined differently by individual practitioners. It is more useful, when relaying the circulatory status of a child, to convey the actual vital signs, progression over time, and response to fluid to indicate volume state. In children with less severe trauma, the technique of abdominal examination is important to reliably exclude significant intraabdominal injury clinically. Where physical examination is to be relied on as the major indicator of abdominal injury, it should ideally be performed regularly by the same observer. It is well recognised that significant injuries may not be clinically apparent on the initial physical assessment. With serial examination and vigilance to vital signs, changes are detected early and appropriate management implemented. The aim of the abdominal examination is to view and elicit physical signs. Tenderness, even minor, may warrant admission for ongoing serial observations and examinations. Signs of peritoneal irritation such as rebound, guarding, or rigidity may require evaluation by CT scanning and should be reviewed with a paediatric surgeon. Auscultation of the abdomen has limited usefulness. Signs of peritoneal irritation and bowel sounds unfortunately have a low interrater reliability. Altered level of consciousness and pain from injuries add to the difficulty of abdominal assessment.

There is an association between a lowered level of consciousness and intraabdominal injury. The PECARN study group demonstrated that of 12,044

children with blunt abdominal trauma, the frequency of intraabdominal trauma was 2% in a child with a GCS of 15, 4% with a GCS of 14 and 9% with a GCS of 13.[1] Judicious and early use of parenteral opiates is safe, decreases a child's distress, and allows a more accurate clinical assessment. Abdominal examination must be performed by gentle palpation with warm hands. Lower chest wall and rib tenderness should raise concern for liver and spleen injury. There should be a brief but careful visual assessment of the abdominal wall for the distribution of any penetrating wounds, bruising, or marks (e.g. seat belt sign, handlebar imprints or tire marks). The presence of these warrants a prolonged observation period and detailed inpatient tertiary survey, even for the child with no other positive findings. Information from clinical examination can usually be obtained by gentle palpation with occasional use of percussion tenderness. This technique is acceptable in children with minor trauma who require careful re-examination. The majority of children with hollow viscus injury will not have peritonitis on their initial assessment, though tenderness is a consistent finding. Rectal and vaginal examinations are not indicated routinely and are rarely indicated in the child with minor abdominal trauma. In the child with only minor injuries, the way the child moves around the emergency trolley or walks can be a useful screening tool as to whether intraabdominal injury exists.

Seat belts are the single most effective means of reducing injury to a motor vehicle occupant in a collision. They reduce collision-related mortality by approximately 50%. However, especially if poorly fitting, they are associated with a specific pattern of injury. A seat belt sign is shown by erythema, bruising and abrasions in a pattern consistent with a seat belt in a restrained child following a motor vehicle collision. It is associated with solid organ, hollow viscus and mesenteric injuries. With an identified seat belt sign, 14.4% of children will have an intraabdominal injury. There is an approximately 1% association with pancreatic injury. While there is no confirmed increased mortality rate with a seat belt sign, there is an associated 6.5% surgical intervention rate.[4] Seat belt-related injuries have a particular association with hollow viscus and mesenteric injuries which frequently require surgical intervention. Both hollow viscus and pancreatic injuries are challenging to identify early, and a normal initial CT does not preclude their ultimate confirmation.

Bicycle-related trauma is one of the leading causes of paediatric abdominal trauma. Handlebar imprints are a unique identifier of a potential significant injury to a child. Though this may occur with a seemingly low-impact event, a

significant amount of force is focused on a small area and may result in sinister intraabdominal trauma that is easily overlooked on the initial assessment. Pre-adolescent males are the most common group to present with this injury. About 20% of small bowel injury is bicycle related with half having a benign initial examination and half not demonstrating free gas on plain films. Handlebar impacts are also associated with solid organ injuries, injuries to the mesentery, mesocolon and abdominal wall hernias.[5]

The PECARN group has published a clinical prediction rule for children at very low risk of intraabdominal injury.[6] A child with no evidence of abdominal wall trauma or seat belt sign, GCS greater than 13, no abdominal tenderness, no evidence of thoracic wall trauma, no complaints of abdominal pain, no decreased breath sounds, and no vomiting is considered very low risk for an intraabdominal injury. The rule had a negative predictive value of 99.9% and a sensitivity of 97% for intraabdominal injury.

Investigations

Laboratory

Blood should be taken for group and hold, full blood examination (FBE), liver function tests, amylase, and blood glucose. Some studies have demonstrated that elevated transaminases in combination with an abnormal physical examination are associated with intraabdominal injury (although not a specific organ injury).[7,8] However, at present there are no laboratory studies that can be recommended as a screening tool for intraabdominal injury. In the context of pancreatic injury, serum amylase often rises, but the initial amylase may be normal, with increasing values over 3 days.[9] In general, neither amylase nor lipase is considered a sensitive or specific marker for the initial assessment of pancreatic injury.

Urine should be obtained for examination, although haematuria is an inconsistent finding in children with renal tract injury. The absence of haematuria does not exclude injury to the renal tract.

Focused assessment by sonography for trauma

Focused assessment by sonography for trauma (FAST) ultrasound is operator dependent and should be performed only by clinicians with appropriate training and credentialing.

FAST is controversial in children. There is much supportive literature for the use of FAST.[4–6] While FAST has become a standard part of the primary survey for the shocked injured child, its use as a part of the abdominal examination in the secondary survey warrants scrutiny. This is further discussed in Chapter 23.1 (Ultrasound).

Plain films

Plain abdominal radiographs are not useful in the ED assessment of abdominal trauma in a child. There is a move, albeit contentious, from routine to clinically indicated pelvic radiography in paediatric trauma. Unnecessary pelvic radiography exposes a child's developing skeleton and gonads to a significant ionising radiation dose. In the unstable child with concerns of a pelvic fracture, a plain film of the pelvis is indicated. A child with concerning features of pelvic or proximal lower limb bony injury should have imaging. Plain radiographs of the pelvis in children do not identify all pelvic fractures or dislocations, although a normal radiograph substantially reduces the risk of a clinically significant bony injury being present. CT is the gold-standard imaging medium for identifying injuries to the pelvis. If a child with clinical suspicion for a pelvic fracture is having a CT abdomen as part of his/her assessment then the plain pelvic film should be forgone in lieu of the CT abdomen and pelvis.

Computed tomography scan

CT is the best imaging modality in terms of sensitivity and specificity for assessing intraabdominal injury. CT is invaluable in assessing the haemodynamically stable child. There have been increased concerns from the unnecessary exposure to radiation from CT imaging. The judgement between the risk of a missed intraabdominal injury and the risk of radiation-induced malignancy remains a challenge. A child who has normal observations and no concerning features of intraabdominal trauma should not have a routine CT but rather should be assessed with serial examinations. For a stable child with concerns for intraabdominal trauma, CT is the investigation of choice. Issues of transport and sedation will need to be considered. With the acceptance of non-operative management of blunt abdominal injuries, diagnostic imaging is an essential component of the assessment process of the injured child to identify significant injury, plan admission length of stay and aid in guiding decisions for

interventional radiology or surgical intervention. In an institution without the availability of paediatric surgical services a child with concerning features of intraabdominal or pelvic trauma should have a CT as part of his/her assessment.[10] An unremarkable CT scan has a 99.6% negative predictive value for intra-abdominal injury and 99.8% negative predictive value for surgical intervention.[11] It identifies free intraabdominal fluid, solid visceral injury and the injury configuration, and loops of bowel and demonstrates free peritoneal gas. The retroperitoneum is well visualised. The injuries at risk for failed detection in the initial CT being pancreatic and mesenteric or hollow viscus injury. The sensitivity of the initial CT for a pancreatic injury may be as low as 60%. CT examination should be performed after the administration of intravenous contrast. This enables better visualisation, although in situations of renal hypoperfusion renal failure can be precipitated. Allergy to contrast is a rare but potentially serious problem. In general CT scanning should be performed for a child that is stable with clinical concerns for intraabdominal injury. Concerns include abdominal tenderness, peritonism, a lowered level of consciousness and risk of intraabdominal injury, a child with external evidence of abdominal trauma (seat belt sign, handlebar imprint, large abdominal wall haematoma or abrasions), children with a positive FAST scan, children with a falling haematocrit or elevated liver transaminases and a child with macroscopic haematuria. CT scanning should be reserved for those patients in whom there is a high index of suspicion of intraabdominal injury. The decision to CT should be a multidisciplinary decision involving at least the emergency physician and paediatric surgeon. Note that a child admitted to an adult trauma service appears more likely to undergo CT imaging than a child admitted to a specialised paediatric trauma service with no improvement in outcomes from the increased imaging rate.[12]

The addition of oral contrast prior to a CT of the abdomen used to be routine for the ED assessment of the injured child. But improvements in CT scanning technology have rendered the use of oral contrast for the initial assessment of the injured child obsolete. Oral contrast delays time to scanning, increases the rate of vomiting, reduces image quality for assessing the spleen and liver and does not significantly improve the early detection of a hollow viscus injury.

Formal ultrasound

A detailed upper and lower abdominal ultrasound examination takes longer to

perform and is more observer-dependent than CT examination. Abdominal distension caused by ileus and luminal distension, and the presence of abdominal tenderness in the child, can make ultrasound examination more difficult. Hence there is little role for ultrasound in the definitive diagnosis of abdominal trauma, except when CT scan is unavailable.

General management

The assessment begins with primary survey and any life-saving interventions, while historical details are obtained simultaneously. Oxygen should be administered and vital signs monitored regularly. Vascular access is obtained early and appropriate blood samples for blood cross-matching, haematology, and biochemistry. Children are more likely than adults to arrive without the pre-hospital emergency services obtaining intravenous access. Where there is delay in obtaining venous access, intraosseous access remains an effective method of resuscitation. Trauma series films of chest, pelvis, and lateral cervical spine should be obtained, when indicated, during the resuscitation. Views of thoracic and lumbar spine may also be required if indicated on mechanism or clinical findings. In the severely injured child, CT scanning may provide much of this information.[3]

The abdominal examination is usually reserved until the secondary survey. Attention should be exercised to ensure that the child is warm to prevent the development of hypothermia during resuscitation.

Fluid therapy should begin with 20 mL kg of warmed crystalloid (normal saline) and repeated if required. If further fluid therapy is required after two crystalloid boluses, blood should be used in volumes of 10 mL kg.[13] Fluid resuscitation should be guided by physiological markers. Both over and under resuscitation are potentially harmful. Permissive resuscitation, while common in adults, has not gained evidential support for its use in paediatric volume resuscitation.[14] The rapidity of blood loss may rarely dictate the use of O-negative or group-specific blood rather than waiting for full cross-matched blood. Massive transfusion protocols are now a part of most paediatric trauma resuscitation algorithms.

Early consideration of gastric decompression with a nasogastric tube assists abdominal assessment and aids ventilation in the intubated child. The insertion of a urinary catheter may be necessary, depending on the requirement to aid haemodynamic monitoring of fluid resuscitation and to detect haematuria.

Perineal haematoma and blood at the external urethral meatus are contra-indications to routine catheter insertion and mandate discussion with a paediatric surgeon. Ongoing management is usually dictated by the haemodynamic response of the child to fluid resuscitation. CT examination is ideal but may not be possible in a very small number of exsanguinating children with deteriorating vital signs despite fluid resuscitation. In this situation, early surgical consultation regarding urgent laparotomy is required.

Surgical issues

Non-surgical management of solid organ injuries has been advocated for adult trauma since the 1960s. Nearly all spleen and liver injuries, including high-grade injuries, will stop bleeding with supportive measures (Table 4.5). Bleeding from an injured spleen, liver, or kidney is generally self-limiting. In the clinically stable child with solid organ injury non-operative management has become the standard of care. The failure rate of non-operative management in children with liver, spleen or renal injury is less than 5% in specialised paediatric trauma centres. In children with equivalent spleen or liver grade of injury, those managed non-operatively will have lower rates of mortality, morbidity, transfusion requirements and length of stays.[15] As rates of surgical intervention have reduced so has the mortality of spleen and liver injury-related mortality. Surgical intervention generally occurs in the setting of continued cardiovascular instability despite resuscitation or peritonism or with diaphragmatic, pancreatic duct or hollow viscus injuries. A non-operative approach will be most successful in an institution with a paediatric surgeon with a commitment to the injured child and dedicated paediatric intensive care or high-dependency facility.[14] This may necessitate the transfer of the child to a major centre.

Table 4.5.1

American Association of Surgery for Trauma Liver injury scale

Grade	Injury description
1	Subcapsular hematoma, <10% surface area; capsule tear, <1 cm parenchymal depth.
2	Subcapsular hematoma, 10%-15% surface area; intraparenchymal, <10 cm in diameter; laceration 1–3 cm parenchymal depth, <10 cm in length.
3	Subcapsular hematoma, >50% surface area or expanding; ruptured subcapsular or parenchymal hematoma; intraparenchymal hematoma >10 cm or expanding; laceration >3 cm parenchymal depth.
4	Parenchymal disruption involving 25% to 75% of hepatic lobe or 1–3 Couinaud’s segments within a single

	lobe.
5	Parenchymal disruption involving >75% of hepatic lobe or >3 Couinaud's segments within a single lobe; vascular juxtahepatic venous injuries; i.e., retrohepatic vena cava/central major hepatic veins.
6	Vascular hepatic avulsion.

From Moore EE, Cogbill TH, Jurkovich GJ, et al. Organ injury scaling V: spleen and liver (1994 revision). *J Trauma* 1995;**38**:323.

Table 4.5.2

American Association of Surgery for Trauma Spleen injury scale

Grade	Injury description
1	Subcapsular hematoma, <10% surface area; capsule tear, <1 cm parenchymal depth.
2	Subcapsular hematoma, 10%–50% surface area; intraparenchymal, <5 cm in diameter; lacertion 1–3 cm parenchymal depth, <10 cm in length which does not involve a trabecular vessel.
3	Subcapsular hematoma, >50% surface area or expanding; ruptured subcapsular or parenchymal hematoma; intraparenchymal hematoma >5 cm or expanding; laceration >3 cm parenchymal depth or involving trabecular vessels.
4	Laceration involving segmental or hilar vessels producing major devascularization (>25% of spleen).
5	Completely shattered spleen; hilar vascular injury which devascularizes spleen.

From Moore EE, Cogbill TH, Jurkovich GJ, et al. Organ injury scaling V: spleen and liver (1994 revision). *J Trauma* 1995;**38**:323.

As more than 95% of spleen and liver injuries are managed non-operatively, it is useful to be aware of the advised length of the inpatient observation. The American Pediatric Surgical Association in 2000 recommended a period of bed rest equal to the grade of spleen or liver injury plus 1 day prior to discharge (e.g. a grade IV splenic laceration would mandate 5 days of hospital bed rest). Since then the abbreviated bed rest protocol (ABRP) for blunt liver and spleen injury recommends 1 day of bed rest for grade I–II injuries and 2 days' bed rest for grade III or higher injuries.[16]

Hollow viscus injuries

Operative management is the rule for hollow viscus injuries. Diagnosis is frequently delayed due to the challenge with the both clinical and radiological identification of a hollow viscus injury at the acute phase of presentation. There is no reliable examination finding for hollow viscus injury. The potential for minimal signs on presentation and the potential for paucity of radiological evidence emphasises the importance of observation and serial examinations.

Changes in serial examination findings remain the most sensitive tool for raising the concern for a hollow viscus injury. Fortunately hollow viscus injuries are uncommon. The jejenum is the most common site of injury followed by the duodenum, colon and stomach. Most injuries result from a direct blow: the handlebar of a bicycle, seat belt or as an inflicted injury. Some injuries, such as duodenal haematoma without evidence of perforation, may be managed without surgery. The decision about operative versus non-operative management is made by the surgeon who will have ongoing care of the child. This decision is strongly influenced by clear details regarding progression of vital signs, response to fluid therapy, and associated injuries. The diagnosis of a hollow viscus injury in a child less than 2 years of age should raise concerns for non-accidental injury if the mechanism is not in keeping with the presentation.

Pancreatic and renal injuries

Pancreatic injuries are rare, making up less than 1% of intraabdominal injuries. Pancreatic injuries are associated with a relatively high (5%) mortality rate.[17] Amylase is neither sensitive nor specific for pancreatic injury. CT also is insensitive in the initial stages. There is a role for serial CTs, MRI or endoscopic retrograde cholangiopancreatography (ERCP) for a definitive diagnosis if diagnostic concern is raised.[18] The management of pancreatic injuries remains controversial. There is a paucity of quality evidence to guide a clinician in management. The majority are managed non-operatively, the remainder with either ERCP or stenting or with surgery. The major complication rate following pancreatic injury is 25%. The rate of surgical intervention increases with the increasing grade of injury, higher ISS and if a high-grade duct or pancreatic body injury is evident. The majority are managed conservatively, although up to 25% will undergo surgical intervention.

Kidney injuries are uncommon despite a seeming predisposition for renal injury in children as the kidneys are less protected by the lower ribs and generally less cushioned by perinephric fat. The most common injuries are contusions. The large majority are managed non-surgically, and for those with ongoing haemodynamic instability despite resuscitation there is an emerging use of interventional radiology methods.

Interventional radiology

Endovascular interventional radiological procedures are well established for adult abdominal solid organ and pelvic trauma. In paediatric trauma interventional radiology is less common due to the relative lower numbers, reluctance of interventionalists to perform paediatric procedures and lack of equipment able to access the smaller vascular structures of children. With the development of both experience and equipment able to manage smaller vessels, in some centres interventional radiological techniques are becoming more common.[19]

Penetrating trauma

Penetrating abdominal injury in a child usually requires exploration by laparoscopy or laparotomy. Because the abdominal wall is often thinner than in adults, penetration into the peritoneal cavity occurs more readily. A careful assessment, including a log roll for back examination, to exclude other injuries, is necessary. Gunshot wounds should be explored in theatre. The approach to initial resuscitation is identical to that for blunt trauma. An erect chest X-ray or lateral decubitus film helps to identify the presence of free air. All but the most trivial wound should undergo formal surgical exploration as blind probing is unreliable.

Pelvic fractures

A child who sustains a fractured pelvis has been exposed to severe trauma. Children are more resistant to sustaining pelvic fractures relative to adults. These are uncommon injuries in children, occurring at half the frequency of adults. Adults sustain pelvic fractures at a higher rate than children for an equivalent impact.[20] There are several major differences in the bony pelvis between the child and adolescent or the adult. There is greater elasticity in the sacroiliac joints and pubic symphysis and plasticity of the bone in the paediatric pelvis; therefore, greater amounts of kinetic energy must be involved to cause fracture. Avulsion fractures occur in children and adolescents because cartilage is weaker than bone. This occurs at the physis. Greater laxity of the joints in the paediatric pelvis means that single fractures occur more commonly, as opposed to the adult pelvis, where there is the double-break concept. Fractures occurring through epiphyseal and apophyseal growth centres may result in growth arrest, leg-length discrepancy, and deformity. Children also have increased capacity for

remodelling.[21] There are several classification systems for pelvic fractures. None is ideal. Torode and Zeig described four groups of pelvic fracture but failed to include isolated acetabular fractures.[22] This has been modified by Silber et al., [21] whose classification by mechanism of injury and description is useful (Table 4.5.3). Associated injuries increase in frequency with the increasing severity of fracture type. Therefore, when a pelvic fracture is identified on initial X-ray, a thorough search for other injuries must be undertaken.

The management of those other injuries usually takes priority over the pelvic fracture management. Bladder injury, while more common than in the adult, is an infrequent association. In a review of 166 children with pelvic fractures, there was one urethral disruption and two bladder contusions. There is a strong association of these injuries with straddle type mechanism. Children commonly receive 'fall astride' injuries related to playground equipment or while riding bicycles. In Silber's series, 97% of children were treated non-operatively. The majority of these injuries (63%) were type 3 fractures, and the remainder were type 2 (17%) and type 4 fractures (17%).[23] In a study comparing paediatric and adult fractures, the mortality rate for children was 5.7% compared with 17.5% in the adult group. Haemorrhagic death due to a pelvic fracture is rare in children. This is thought to be due to the greater skeletal flexibility, relatively thicker and more adherent periosteum and the greater ability of paediatric arteries to vasoconstrict after injury. In those children in whom blood loss is significant, early involvement of an orthopaedic surgeon and interventional radiologist is essential to optimise management. External fixation and angiography have both been used successfully, particularly in adolescent children.

Table 4.5.3

Classification of paediatric pelvic fractures

Type	Mechanism	Description
1	Avulsion	Separation through or adjacent to an apophysis
2	Lateral compression	Iliac wing fractures
3	Anteroposterior compression (usually)	Simple ring fractures: • isolated pubic rami fractures • disruption of the pubis symphysis without disruption of the sacroiliac joint • isolated acetabular fractures
4	Anteroposterior compression (usually)	Ring disruption features: • fracture (or diastasis) of both anterior and posterior structures • pelvic fracture with an acetabular fracture • straddle fracture: bilateral superior and inferior pubic rami fractures

Originally described by Torode and Zeig and modified by Silber JS, Flynn JM, Koffler KM, et al. Analysis of the cause, classification and associated injuries of 166 consecutive pediatric pelvic fractures. *J Pediatr Surg* 2001;**21**:446–50.

Disposition

Almost all children with significant mechanism or signs of abdominal or pelvic injury require admission from the ED. The nature and severity of the injuries and intended management determine the most appropriate location for this to occur. Younger children who have experienced a significant mechanism of injury (e.g. fall from great height, high-velocity motor vehicle crashes, pedestrian hit, or run over by motor vehicle) but who are apparently injury-free or have only minor injuries should be admitted for observation (for 12–24 hours). Abdominal injuries in young children may initially have minimal or subtle signs, which become more apparent after observation and serial examination. Because of the plasticity of the paediatric skeleton, significant internal derangement can occur without obvious external evidence of trauma. In the older child, however, it may be appropriate to discharge the patient who is injury-free or has only minor injuries. This should occur after several assessments while in the ED and with arrangements for follow-up with a medical practitioner within 24 hours. Parents should be given clear instructions to return should a child's symptoms change. In general, children with ongoing abdominal pain after trauma should not be discharged, regardless of negative imaging results.

Controversies

- The use of FAST scanning. FAST has a role for the shocked child with concerns for intraabdominal trauma. There is a high false-negative and false-positive rate that merits caution in synthesising a FAST scan result with the child's assessment.
- Defining the haemodynamically unstable child. When in doubt, discuss the child with an emergency or paediatric surgical colleague. Children can be profoundly hypovolaemic with normal vital signs or only tachycardic. Ongoing fluid requirements and indices of peripheral perfusion are important indicators of volume status.
- There is debate about which subset of blunt abdominal trauma patients can be safely managed with serial examination and without CT scanning.

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4.6

Burns

Peter L.J. Barnett

ESSENTIALS

1 Burns are one of the leading causes of injury in children in Australia.
2 The definitive assessment of the ‘depth’ of the burn may be difficult early on, as the appearance can evolve during the first 24–48 hours.
3 The calculated amount of fluid (burn-deficient plus maintenance) to be replaced in 24 hours is only a guide and should be adjusted according to the child’s haemodynamic response.
4 Children with major or complicated burns should be treated in a paediatric burns unit.
5 Consider non-accidental injury if the presentation is delayed or if the history given is inconsistent with the burn sustained.
6 Electrical injuries are more commonly seen in two age groups: toddlers within the home setting and male adolescents involved in risk-taking behaviour. High-voltage exposures have a more serious outcome, as they are more likely to be associated with injury to internal structures.

Introduction

Burns sustained by children are a common presentation to emergency departments (EDs) and often cause significant distress to both the child and the parents. The mortality is increased in younger children. Deaths are generally related to flame burns, which may be complicated by inhalation of smoke and

other toxic gases (e.g. in house fires). Early fatalities are related to respiratory complications, whereas late deaths are usually related to infection. The use of early debridement and skin grafting has led to an increased survival rate in patients who would have previously died because of infection.

Most paediatric burns are fortunately less serious, resulting mainly from scalds. This commonly occurs in preschool-aged children due to their inquisitive nature precipitating accidents in the home. Flame burns occur in older children often experimenting with flammables. Chemical and electrical burns are uncommon. One must be alert to the possibility of burns presenting as a manifestation of non-accidental injury in a young child.

Several preventive strategies can help decrease the risk and degree of burns sustained, especially with thermal burns. Lowering the temperature of hot water heaters to a maximum of 50°C significantly increases the contact time needed to produce deep or full-thickness burns. Flame-resistant clothing and smoke detectors in homes have saved many lives. Spill-proof mugs, guards around wood fire stoves, and child-resistant taps have all been shown to prevent burns. Further prevention strategies will have a far bigger impact on burns than advances in burn management.

Pathophysiology

The skin is the largest organ in the body, and its functions include:

- preventing heat loss to the environment, thus regulating the body's temperature
- preserving body fluids by preventing water loss from the body
- acting as a barrier to infective organisms.

Therefore children with extensive burns have difficulty retaining fluid and regulating temperature, and are at risk of infection.

The skin is composed of two main layers:

1. *Epidermis*: composed of stratified squamous epithelium, which is largely non-viable. It acts as the barrier to infectious agents as well as preventing fluid loss from the body.
2. *Dermis*: contains the epithelial adnexal structures, e.g. hair follicles, sweat glands, and neural receptors for pain and pressure. It also contains

blood vessels, which contribute to temperature regulation of the body via radiant heat loss.

After a burn, injury to the deeper specialised epithelial cells prompts a change into stratified squamous epithelium. These cells proliferate, gradually covering the burn with a non-epithelial barrier. Therefore, if the dermal structures are damaged, skin grafting is the only means to cover the defect. When these deeper layers are involved, scarring results and contractures may occur.

The depth of the burn will depend on the temperature of the substance in contact with the skin, the length of time the substance is in contact with the skin, and the extent of subsequent cooling of the burned skin area. Hypothermia due to cooling occurs quickly in children due to their higher surface area to weight ratio, compared to adults. Also, children have thinner skin, which leads to deeper burns for a given contact temperature and duration.

Classification

Burns are generally classified into superficial, partial thickness or full thickness. Previous nomenclature (first, second, and third degree) has been replaced to give a more accurate description of the burn. In the ED setting, the definitive assessment of the ‘depth’ of the burn may be difficult, as the appearance can evolve during the first 24–48 hours. Likewise, the burn is not generally uniform in depth, and it may take time to delineate between superficial and deeper areas. Superficial and partial-thickness burns are the most common burns seen in children.

Superficial

Superficial burns generally involve only the epidermal layer. These are commonly seen resulting from sunburn or minor scalds. Blistering does not occur immediately but may over the next few days. The pain and swelling generally last only a few days. The skin is erythematous and blanches normally. The epidermis will often peel within 3–7 days and is completely healed by 7–10 days, without scarring.

Partial thickness

Partial-thickness burns occur when the whole epidermis is involved and part of

the dermis. The more of the dermis involved, the more the scarring potential.

Superficial partial thickness

Superficial partial-thickness burns involve the papillary layer of the dermis and are characterised by erythema with blistering. The blisters may remain intact and later burst due to an external pressure. The skin underlying the blister has a pink or red colour and moist appearance. These burns are extremely painful, as the nerve endings are exposed. The deeper the burn, the slower the healing process. Generally, superficial partial-thickness burns heal in 2–3 weeks.

Deep partial thickness

Deep partial-thickness burns involve the reticular layer of the dermis. They may be less painful than their more superficial counterpart, due to oedema lessening the exposure of the nerve endings. They are a paler colour with a speckled appearance due to thrombosed superficial vessels. The skin is non-blanching. It may initially be difficult to distinguish between full- and deep partial-thickness burns. Deep partial-thickness burns generally heal after 3–6 weeks. Scarring is common, and skin grafting is sometimes necessary.

Full thickness

Full-thickness burns generally occur after flame burns or after prolonged contact with a hot surface. Other causes include hot oil, prolonged immersion, or chemical burns. They involve the epidermis and all the dermis, including epidermal appendages. They have either a dry, hard, white, leathery appearance or may be black in colour. They usually have no sensation because the nerve endings have been destroyed, and pain is due to more superficial burns on the edge of the full-thickness burn. Full-thickness burns are able to heal only from skin regrowth from the edge of the burn, which causes scarring. Therefore most will require skin grafting.

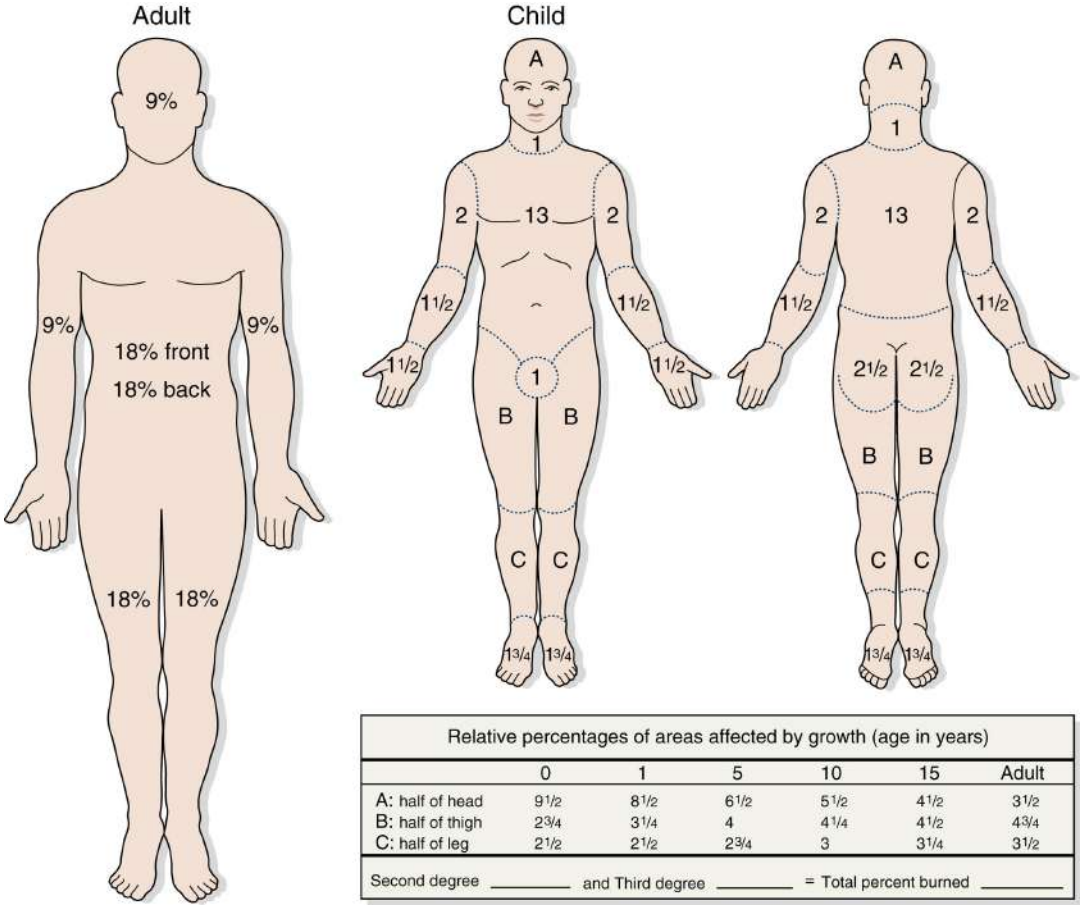
All significant burns will become colonised with bacteria. Heat causes coagulation of tissue, which leads to oedema and non-viable skin. This will potentially become a rich source of nourishment for bacteria. Adequate debridement is required to reduce the risk of infection. Infection will increase the depth of skin damage and thus the degree of scarring.

The location of the burn is also important regarding potential scarring, contracture, and caring for the acute burn. Facial, hand, foot, and perineal burns

may be difficult to dress. Hence the part of the body involved by the burn will influence whether inpatient or specialist care is required.

History

The history is generally obtained from the parent to clarify events. Ambulance officers may have important additional information. It is important to know the substance and estimated temperature of the substance that caused the burn (e.g. hot cup of tea, cooking oil, flame) and the duration of contact with skin (e.g. was the patient clothed at the time, in what, and for how long?).



Relative percentages of areas affected by growth (age in years)						
	0	1	5	10	15	Adult
A: half of head	9½	8½	6½	5½	4½	3½
B: half of thigh	2¾	3¼	4	4¼	4½	4¾
C: half of leg	2½	2½	2¾	3	3¼	3½

Second degree _____ and Third degree _____ = Total percent burned _____

FIG. 4.6.1 Lund Browder chart for estimation of percentage size of burn.

With flame burns, was the patient trapped in an enclosed space, therefore at risk of inhalation problems, or was there any loss of consciousness? These features may suggest inhalation issues, such as carbon monoxide poisoning. The historical information may determine the potential for associated injuries

resulting from falls or explosions.

Non-accidental injury should be considered if the presentation is delayed or where the history given is inconsistent with the burn sustained or if the burn has distinctive distribution (e.g. glove and stocking). Past medical history and immunisation status, particularly for tetanus, should also be obtained (see Chapter 1.1).

Examination

Note that children who are distressed may require the provision of immediate appropriate analgesia at the outset to aid examination.

Primary survey

The initial assessment of the child should be directed to the presence of any features that suggest potential airway involvement. These include singed nasal hairs or eyebrow hairs, oral or facial burns, coughing up carbonaceous sputum, barking cough, altered voice, stridor, wheeze, or respiratory distress. Airway compromise (upper or lower) may have an insidious onset, so frequent re-examination of the child is vital during the first 12–24 hours. If stridor or hoarse voice is present, this indicates upper airway involvement, and early intubation is required prior to evolving oedema causing total airway obstruction. Delayed intubation in this setting can be increasingly hazardous due to distortion of the normal laryngeal anatomy. Scalds to the face/neck rarely cause airway compromise, unless the child has ingested hot liquids.

The circulation status should be assessed next. Hypovolaemia resulting from third-space fluid loss will not occur for a few hours after a severe burn. Therefore, if early onset cardiovascular instability is present, then an alternative explanation, such as bleeding, should be sought. The peripheral perfusion of limbs should be assessed where circumferential burns are apparent. Comparing the pulse wave forms of various digits on the limb using a saturation oximeter may assist this in questionable instances.

Evaluation of burn area

The extent and depth of the burn should be assessed after the patient has been stabilised. This is usually done from a body chart (Fig. 4.6.1), which can be

useful to aid documentation of the burn. This chart is used because the surface area involved will alter depending on the age of the child and the parts of the body where the burn is located. A simple method, using the palmar surface of the child's hand and fingers, can also be used to estimate the area of burn. This correlates to approximately 1% of the child's total surface area. The adult formula using the 'rule of nines' can be used in adolescents older than 15 years.

Investigations

Patients who require intravenous resuscitation should have a baseline full blood count, electrolytes, creatinine and urea, and blood group and hold performed. In severe burns, replacement with blood, protein, and electrolyte may be necessary. Carboxyhaemoglobin levels should be obtained in patients with inhalation burns and extrapolated to the time of injury. A level of >15% on arrival in emergency suggests significant smoke inhalation. Urine output should be monitored in severe burns, both to guide adequacy of resuscitation and also to screen for presence of myoglobin.

Oxygen saturation monitoring and arterial blood gases (when indicated) aid clinical assessment of respiratory involvement. A chest X-ray should also be obtained if inhalation burns are suspected. Inhalation burns will evolve over time, and the initial film may be normal. The early appearance of X-ray changes generally indicates a more severe pulmonary injury.

Management

Pre-hospital

The main aims of pre-hospital care are stabilising ABCs, preventing ongoing burn injury, provision of analgesia, covering the area involved, and rapid transfer to an ED.

The first priority in any burn is to assess and stabilise the airway and breathing. Oxygen should be administered where there is suspected carbon monoxide poisoning with inhalation burns. Circulation is generally not a problem in the first hour after a major burn, and rapid transport to hospital should occur. If transfer time is greater than 1 hour, and the burn is greater than 20%, then intravenous fluid replacement should begin en route where possible.

The burns should be covered with water-soaked sterile cloth or tea tree oil-soaked pads. Excessive cooling of major burns causes hypothermia and worsens

the patient's outcome. Recently sustained minor burns should be cooled under running water for at least 20 minutes. This acts to minimise the extent of the burn and also affords some pain relief.

Analgesia is generally required early, and a single dose of a narcotic (e.g. fentanyl 1.5–2 mcg kg^{-1} intranasally) is a good choice in burns less than 20%. Greater than 20%, then intravenous access should be obtained and analgesia titrated to responses as well as administering intravenous fluids.

If electrical injury has been sustained, then cardiac monitoring should occur during transfer. Patients with chemical burns should undergo extensive washing of the affected area before transport.

Emergency department

The initial priority should focus on stabilisation of airway, breathing, and circulation, with concurrent provision of analgesia. If airway burns are suspected, then early intubation should be considered. Patients with obvious stridor due to upper airway compromise require urgent intubation. Supplemental oxygen should be instituted and oxygen saturation monitored.

Other potential indications for ventilation in major burns include:

- extensive burns (>60–70%), to decrease the patient's work of breathing
- full-thickness circumferential chest burns, which may compromise chest expansion
- severe inhalational lung injury causing pulmonary oedema and hypoxaemia. Inhalation burns generally get worse in the first 12–24 hours, and ventilation may be needed during this time. Early consultation with paediatric intensive care colleagues in these instances is appropriate.

The fluid losses due to the burn itself do not cause early circulatory failure, and other contributing injuries should be sought in the patient with early shock. It is also important to note the time the patient is evaluated, relative to the time of the burn. Children who have delay (e.g. a few hours) in presentation may arrive with circulatory compromise from skin fluid losses.

Intravenous access should be placed in all children with a burns body surface area (BSA) of >20%. Fluid resuscitation rates should be calculated using the time of the burn, not the time of presenting to the ED (see Fluid resuscitation,

below). Peripheral venous access, preferably through non-burnt skin, is preferred over central venous access for the initial resuscitation. Monitoring of urinary output (via urinary catheter, weighing nappies) is important in determining the adequacy of fluid replacement. A nasogastric tube should also be inserted in children with severe burns, as gastric dilatation can occur, leading to respiratory compromise.

Analgesia should be given early, during the stabilisation of the A, B and C. Intranasal fentanyl (1.5–2 mcg kg^{-1}) is a good first up treatment. In severe burns, a morphine infusion should be started after adequate initial intravenous or intranasal analgesia has been given. Large doses of narcotics are sometimes needed in severe burns to control the pain. Intramuscular morphine should never be used given the use of intranasal fentanyl.

A careful secondary survey should then be undertaken, looking at the extent, depth and anatomical relevance of the burns. It is important to determine if there are any circumferential burns to the limbs and chest. In superficial or partial-thickness burns, careful monitoring of circulation or ventilatory compromise is required. In full-thickness circumferential burns, an urgent escharotomy may be required to restore circulation to the limb or allow for adequate ventilation. A burn specialist should be consulted in this situation.

The secondary survey should also include careful examination for any other injuries requiring attention (e.g. head, neck, chest, limbs, pelvis, intraabdominal). Burns to the face should also include fluorescein staining of the eyes to check for corneal involvement.

Fluid resuscitation

Fluid resuscitation should be calculated based on the weight of the child and the total surface area of the burn. Several formulas are used to calculate the resuscitation fluid requirement in the first 24 hours. The **Parkland formula** (BSA affected % × weight (kg) × 4) gives the number of millilitres of *resuscitation fluid* to be given over the first 24 hours. Half the fluid is given in the first 8 hours and the remainder in the subsequent 16 hours. The 24-hour period should begin from the time of the injury. Thus if a patient has received very little fluid in transfer, and it is 4 hours since initial burn, then the fluid calculated should be given over the next 20 hours and half the calculated fluid given in the first 4 hours.

In addition, *maintenance fluid* for the 24-hour period should also be given.

This is calculated as 100 mL kg^{-1} for 0–10 kg, 50 mL kg^{-1} for 11–20 kg, and 25 mL kg^{-1} for >20 kg. Thus a 30 kg child's maintenance = 1000 + 500 + 250 = 1750 mL over 24 hours. It is also important to monitor ongoing losses (urinary output, respiratory loss, etc.).

The calculated amount of fluid (burn deficient plus maintenance) to be replaced in 24 hours is only a guide and should be adjusted according to the haemodynamic response. Patients need to be maintained in a positive fluid balance for the first 24–48 hours. The adequacy of fluid replacement is monitored by urine output and clinical parameters of perfusion. In children, 0.5–1 mL kg^{-1} per hour is the recommended urine output and should be monitored by a urinary catheter in severe burns. A central venous catheter is generally not required in the ED phase of management.

The type of fluid used varies between burn specialists, and it is best to be familiar with the preference of the local paediatric burns unit. In the shocked child, a 20 mL kg^{-1} bolus of normal saline should be given. This can be repeated if necessary to restore peripheral circulation. Ongoing replacement is generally with crystalloid in the first 24 hours, as colloid may leak through the burnt capillaries, causing worsening oedema. After 24 hours, colloid is used as part of the replacement fluids in the intensive care setting. When calculating the initial fluid requirements, it is important to subtract the bolus fluids given from this amount.

Additional fluids may be required in severe electrical burns causing muscle damage, as myoglobin may cause renal failure secondary to renal tubular deposition, and therefore maintaining adequate glomerular filtration is very important.

Management of burns (Box 4.6.1)

Major burns

These patients should be treated in a specialised burns unit. Covering the burn with a sterile dressing is required prior to transfer. Specific dressing type is best decided after discussion with the receiving unit. Within the burn centre, patients are generally dressed with topical silver sulfadiazine (SSD) cream that should be changed each day. At each change, the wound should be cleaned with warm water and debrided to remove any avascular tissue (which may lead to infection). The burns are covered with a non-stick dressing over the SSD cream

(e.g. Melolin) and then wrapped in crepe bandages to prevent contamination of the burn. The face and perineum are generally left open and covered with a water-based gel. SSD should not be used on the face, as the patient may spread it into the eyes. SSD is currently only recommended for inpatient care of patients with significant burns.

Box 4.6.1 Admission criteria for paediatric burns unit

- Require admission:
    - Partial-thickness burns >20% BSA
    - Full-thickness burns >5–10% BSA
    - Smoke inhalation or airway burn is suspected
    - Child abuse suspected
- Consider admission:
    - Burn to hands, feet, face, perineum, or joints
    - Burns <20% BSA and other concerns, e.g. age <12 months, parents not coping, etc.
    - Comorbidity
    - Other significant injuries

Minor burns

Patients with partial-thickness burns <20% or full-thickness burns <5–10% can often be managed on an outpatient basis and reviewed in a burns clinic.

There are several ways to dress wounds in the ED. Burns with intact blisters, unless over a joint surface or extremely large, should be left intact initially. The intact skin acts as a barrier to bacteria, and the skin underneath can heal. After 7–10 days, the intact blisters should be deroofed and the need for grafting assessed. If unclear at this stage, then they should be redressed and assessed in a further 7 days.

Superficial burns

Small superficial burns

Burns which consist of just erythema or some simple small (<1 cm) blisters should be left open and dressed with either Vaseline or skin moisturizer, i.e. they should be treated like a sunburn. Areas of superficial burns larger than this but less than 5 cm should be covered with Mepitel, Melolin and crepe bandage securing this with Hyperfix. Mepitel is a low-adherent wound-contact dressing made of silicone gel bound to a flexible polyamide net. The dressing is left intact until the patient is reviewed after 5–7 days. Repeat dressing may be required or the healing burn can be covered with Tegaderm or Hyperfix and be left on until it falls off.

More extensive superficial burns which have a large blistered area should be treated like a partial-thickness burn (see below).

Partial-thickness/small full-thickness burns

Use Acticoat®, a silver impregnated barrier dressing, moistened with sterile water, covered with IntraSite® Conformable, 'Gladwrap', ± crepe bandage. Secure with Hyperfix®. The exudate from the wound determines the number of dressing changes required. IntraSite® Conformable is a soft hydrogel dressing that combines the advantage of IntraSite® gel with a non-woven dressing. It creates a moist wound environment for the continued release of silver from the Acticoat®. Patients should be reviewed between 3 and 7 days for a change of dressing and to assess if skin grafting is needed.

Superficial burns on the face should be managed non-dressed and cleaned two or three times a day with warm water. Vaseline should be applied to the face a few times a day or when it has been rubbed off by the child. As a burn on the face starts to dry up, application of mild lanolin ointment/cream can be used to aid in healing by softening the skin. The burn should also be protected from the sun, as sunburn of an already burnt area causes increased pigmentation to the skin.

Superficial burns rarely become infected, but infection should be suspected if the patient has unexplained fever or has evolving pain, redness and tenderness. Foul discharge from the burn does not always indicate that infection is present. In this case, the dressing should be changed earlier and the burn inspected, as antibiotics may not be indicated. As the skin heals under a dressing, it becomes pruritic, which may require an antihistamine or cooling of the dressing (particularly in hot weather).

Tetanus prophylaxis is important in major burns or minor burns (which are contaminated). Antibiotics should be used only when a definite infection is

present and not simply for a smelly discharge. Prophylactic use of antibiotics is not recommended.

Pain management for minor burns is generally achieved by the dressing itself. Covering the burn decreases the pain substantially. Generally, paracetamol with or without ibuprofen should be sufficient during the first 24 hours.

Patients with burns involving hands, feet, or face should be referred to a burn specialist for ongoing management; they do not necessarily need urgent referral – dressing the burn appropriately (as above) and follow-up in a few days are usually adequate.

Electrical burns

Introduction

These are infrequent presentations to EDs but have unique problems. Electrical injuries are more commonly seen in two age groups: toddlers within the home setting and male adolescents involved in risk-taking behaviour.

Young children generally sustain electrical burns from low-voltage (<1000 V) or household currents (240 V). These may be due to frayed electrical cords or children inserting metal objects into power sockets. Mouth burns may occur when small children chew on power cords. In most states in Australia, safety switches are installed in all new houses, cutting the current when overloaded, thus preventing many severe electrical burns. These low-voltage exposures rarely result in significant internal injury, and most children are asymptomatic apart from distress from the cutaneous burn.

High-voltage (>1000 V) injuries are seen most often in adolescent males as a consequence of risk-taking behaviour (e.g. climbing electrical poles, train surfing). These high-voltage exposures have a more serious outcome, as they are more likely to be associated with injury to internal structures (e.g. muscles or internal organs).

Clinical effects

Electrical currents preferentially flow along low-resistance tissues such as blood vessels, nerves, and muscles, rather than the skin, causing internal injury particularly if extremities are involved. There is generally an entrance and exit burn in non-water-related current injuries, with increased tissue damage at these sites. Wet or moist skin increases current flow dramatically by decreasing the

tissue resistance.

Clinical manifestations vary according to the voltage exposure and may range from trivial to cardiac arrest.

Skin

- Entry and exit burns: well-demarcated pale areas with charred centre
- Arc burns: heat produced can cause extensive tissue injury
- Flame burns: due to ignition of clothing.

Cardiac

Arrhythmias:

- Low voltage: sinus tachycardia, atrial fibrillation, ventricular fibrillation. Myocardial injury is uncommon
- High voltage: asystole.

Muscular

- Prolonged tetany of musculature: apnoea, fractures and dislocations
- Muscle necrosis and rhabdomyolysis.

Neurological

- Acute: altered mental state; seizures; headache; speech, motor, or sensory disturbances
- Delayed: spinal cord injury, memory and mood disturbance.

Renal

- Renal failure secondary to myoglobinuria.

Other

- Eyes: cataracts
- Gastrointestinal tract: ulceration, perforation
- Trauma following associated falls.

Management

Assessment of the child's A, B and C is the initial priority with electrical exposure, followed by examining for skin burns and potential internal injuries. A baseline 12-lead ECG should be done (unless a trivial exposure) and cardiac monitoring only continued if the initial ECG is abnormal or the child is symptomatic (e.g. chest pain or impaired conscious state). A search for an entrance and exit wound should occur to determine the potential for deeper burns. Analgesia should be provided for the burn or muscle pain. Potential complications and associated injuries are treated on their merit.

Specific issues

Burns

All significant electrical burns warrant review by a burn specialist – particularly if there is ongoing pain or an entry and exit wound with high voltage involved.

Fluids

Fluid requirements for significant electrical burns are underestimated using the Parkland formula, as most of the damage is internal. Therefore fluid requirements are significantly more than estimated, and one should aim to maintain a urine output of 2 mL kg^{-1} per hour.

Myoglobinuria

Muscle involvement leads to myoglobinuria, which may cause renal failure. Adequate fluid resuscitation and alkalinisation may help prevent renal failure.

Compartment injury

Compartment syndrome may also occur due to oedema and burn of the affected muscles. Fasciotomy may be necessary, with debridement of non-viable muscle.

Disposition

The following are general guidelines for admission and discharge after burns.

Admission

- Any high-voltage injury
- Any child with evidence of cardiac or neurological abnormality
- Any child with significantly ongoing pain or myoglobinuria.

Discharge

- Asymptomatic child with low-voltage injury with normal ECG.

Chemical burns

Many chemical agents can cause burns through accidental exposure. They are generally either acid or alkali. Acid burns cause coagulation of the skin, which seems to limit the depth of penetration. Alkali burns cause liquefaction and thus result in a deeper injury. Caustic chemicals tend to give deeper burns than thermal injury, as there is generally a long duration of contact. Oedema tends to occur more quickly in chemical burns, which may cause a deeper burn to appear more superficial.

Treatment

Copious irrigation of the burn is the mainstay of treatment. Sufficient analgesia and topical anaesthesia are required to achieve this in a child. This can be done simply with water. Flushing of the burn should continue for at least 10–15 minutes and sometimes longer. Determination of wound pH via litmus paper may guide the duration of irrigation. If chemicals are introduced into the eye, then irrigation should continue until pH is neutral. This usually requires topical anaesthetic to the eye prior to flushing with saline. A Morgan lens is a good method of flushing eyes, as this requires less cooperation from the patient. Fluorescein staining of the eye should then determine the extent of the burn. Chemical burns to the eye require urgent ophthalmological referral. The treatment of a dermal chemical burn after decontamination should be the same as for any other burn.

Some chemicals may cause systemic toxicity from absorption through the burned skin, and these should be managed accordingly.

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4.7

Children in a disaster response

Mary Elinor McCaskill

ESSENTIALS

1 Consider children's vulnerabilities in a disaster response.
2 Children's care is delivered in a secure location with careful identification.
3 Children have reduced reserve to injury and illness and may have hidden injuries.
4 Normalise the environment for the child with human interaction and play.
5 Collection of data during a disaster response enables identification of developing threats.
6 Recovery of the community includes children's normal environment.

Introduction

A disaster occurs when the needs from a hazardous event exceed the coping capacity of a community. Hazards come in multiple forms – physical, such as earthquakes and transport accidents; infections, such as influenza; public health hazards, such as heat waves, toxins, and loss of infrastructure like electricity within a health system and so on. The cause may be natural or result from human action which in some cases is intentional. The response by health professionals aims to mitigate the impact of a disaster by caring for casualties and providing health care to the community. The priority is to bring 'the greatest good to the greatest number of people'. Looking at disasters in a generic manner enables

research and learning from past experience and planning for future unidentified events to mitigate the impact.[1]

Children in disaster situations

Children are particularly vulnerable in a multiple casualty incident as they are smaller and weaker than adults and unable to care for and protect themselves. At a disaster scene, children are less able to fend for themselves, and therefore care is needed even for those who are not injured. Incapacitation of carers of children has a multiplier effect on the number of casualties.

Phases of disaster response

Planning for a disaster response should be flexible enough to cover all hazards, including unanticipated ones, and also identify specific roles and concrete actions for staff to take. The plan can be escalated to include the whole of the health facility, the community and the region as needed. Hence each plan must fit into the plan of the larger setting. Regular practice ensures a plan is practical and staff are familiar with it.

Children's needs are a component of this planning, keeping in mind their vulnerabilities. Because children cannot care for themselves, a large number of unaccompanied children with minor injuries may be transported to the emergency department (ED). Planning for a disaster includes a sizeable area to accommodate these patients. This needs to be secure to contain frightened children and also to restrict access by the general public for the children's safety. Thought should be given to including equipment to facilitate play and entertainment for children in this space. Play is an important part of a child's response to the world and provides some sense of normalcy and familiarity which is especially important in the disruption following a disaster event.

Command and control are the basis of communication in a disaster response. Clear structures, which have been established in the planning phase, are brought into effect. The aim is to maximise the effectiveness of the response and coordinate the response within a facility, with pre-hospital services and between health facilities. This is assisted by a central operations centre with communication capacity and a small team of experienced staff providing leadership and oversight of operations, logistics, communications and planning for the ongoing response.

Mobilise human resources to assist with the care of patients including those currently at work and those on call. If significant numbers of children are involved, staff with paediatric experience can be very helpful. Physical resources include space for triage, treatment and observation of patients. Links with local community health and GPs are important to develop in preparation for a disaster response to events such as an infectious disease outbreak.

Staff protection against blood-borne pathogens, an infectious disease or a toxic substance is considered at the start of preparations in a disaster response. Children are significant vectors of infectious illness because of respiratory secretions and reduced personal hygiene capabilities.

Patient care in the ED focuses on initial stabilisation and rapid identification of patients needing urgent interventions such as surgery. Triage using set physiological parameters tends to give children a higher priority which is often accepted because of their increased vulnerabilities. The overarching aim is to deliver a high standard of care with careful planning and documentation, which is of utmost importance in a disaster response. Early surgical involvement in the care of children with trauma is important even though cases of splenic and liver injury are less likely to be treated operatively than for adults. Infectious disease agents causing mass casualties such as gastroenteritis or influenza can affect children more significantly through dehydration, sepsis or respiratory failure than in adults. Of particular importance in a mass casualty event is the communication between the ED, operating rooms, surgical teams, intensive care and ward teams to ensure patients receive appropriate care. Operating theatres have the potential to become a bottle neck, so surgical treatment focuses on careful prioritisation of life-saving surgery, damage control and quick turn-around operations. Children need to be carefully considered in this prioritisation because their injuries may be less evident and their haemodynamic reserve is more limited than in adult patients. Patients, including children, continue to present with more routine emergency problems which need treatment.

Psychological first aid is considered in the care of all patients from a disaster event. This involves encouraging a feeling of safety. For a child in the ED, aim for a single person to talk with the child quietly and calmly, introducing him/herself and using the child’s name. Normalising the child’s environment with play, stories or favourite TV shows can put the disaster event in perspective. Singing nursery rhymes can be very effective for infants. Listen to older children who want to talk about what they have seen, without asking for details beyond what they want to discuss. Children can be very concrete and clear about seeing

injury and death, but the underlying distress persists. Avoid TV images showing updates of the disaster event as this can recurrently revisit the child's memories and add to his/her distress. Approach a child's questions with honesty and straightforward answers. Children can be reassured that the team is trying to get in touch with their family members, but avoid false promises about when they will come. If there are several members of a family aim to treat them in the same area so they can support each other. This can be considered in the planning phase with a flexible resuscitation area. Parents and carers provide significant comfort to a child and so are kept close by them during all treatment.

Many young children cannot identify themselves and have no documents to assist this process unlike an adult's driver's license. Reunification with parents and carers may require photographic identification of both child and adult. The details of the adult collecting the child are carefully recorded to reduce distress when other family members arrive and reduce the risk of an unauthorised person posing as a carer.

Ongoing treatment of patients from a mass casualty event may impact on regular health services for an extended period. Early consideration of staffing shifts in the following days enables rest and recovery of staff who were involved in the initial response. Increased staffing may be necessary both during the response to the incident and for some time afterwards as regular activities of the health service are caught up.

Public health is important in a disaster situation to identify developing threats to health. Planning includes careful collection of data on patient numbers, demographics, injury and illness, especially emerging infectious illnesses. Reporting this information to a central unit enables a more complete picture of the disaster event and the health response.[2]

Recovery from a disaster event involves the whole community and includes the health service and staff. The return of structure such as school and day care and the opportunity to play provide normalisation of the child's environment. In turn this focuses the community on the recovery and the future. Ongoing psychological support for the community is part of this recovery.

Health staff also need psychological support in relation to the care given to patients during the disaster response. This includes team meetings, peer support, commemorations and celebrations as well as individual psychological support. In addition health staff who are part of the community may need special consideration for personal and community losses.

International disaster responses require trained and well-equipped staff with

international accreditation so a high standard of care is delivered as requested by the affected country. Integration with local health services is considered during the disaster response and for appropriate follow-up of patients. The team aims to support the local community without adding to the problems they face. Importantly the team is well prepared so reducing the chance of the team members requiring assistance themselves.[3]

A disaster response is a complex multifaceted process in health. Children have specific vulnerabilities in this environment which are included in planning.

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4.8

Wound management

Gar Ming Chan, and Erica Kreismann

ESSENTIALS

1 The goals of wound management are to avoid infection, minimise discomfort, facilitate healing and minimise scar formation. The care of the patient as a whole should be the first management priority.

2 In children, management often requires sedation, adequate local anaesthesia and analgesia. Sedation should only be undertaken by personnel experienced in its use and able to manage the complications of airway compromise, oxygen desaturation and respiratory depression.

3 The comprehension level and the cooperation gained from the child influence wound examination and the information gained. Distraction techniques, adequate topical anaesthesia and appropriate use of sedation can all aid in wound assessment in children. In the child less than 5 years old where examination participation is difficult, observation of posture, symmetry and general movement are surrogates.

4 Examination of function, sensation and circulation distal to the wound is best performed prior to exploration of the wound and prior to regional anaesthesia.

5 If presence of a foreign body is suspected, radiological investigation is advised.

6 Surgical debridement of non-viable tissue is vital to prevent wound infection or delayed wound healing.

7 Tissue adhesives are for external use only and should not be placed

within wounds or used on mucous membranes.

8 In general, sutures are removed earlier in children than in adults.

9 Non-accidental injury should always be considered, especially when the history and injury are inconsistent.

Introduction

Open wounds account for up to one-third of paediatric emergency presentations; two-thirds of open wounds occur in boys, and 40% involve a fall. The scalp and face account for more than 50% of all open wounds, and about 30% occur on the hands.[1–4] The management goals of these wounds are to avoid infection, minimise discomfort, facilitate healing and minimise scar formation. Meticulous attention to wound care and repair should ensure the best possible outcome and functional result. In children this unfortunately requires sedation on occasion. Hand hygiene and universal precautions should be followed when assessing or managing any wound.

Anatomy of the skin

The skin is composed of two layers: dermis and epidermis. The epidermis acts as a protective layer for the dermis, preventing infection and desiccation. It is avascular and relies on diffusion of nutrients from the dermis. The dermis is rich in collagen and thus provides most of the tensile strength of the skin. It has a rich network of nutrient vessels and capillaries. The subcutaneous fat is composed of loose connective and adipose tissues.

Pathophysiology of wound healing

The stages of wound healing are coagulation, inflammation, proliferation and maturation. Wound healing is a sequential process that begins immediately after tissue injury. Coagulation is initiated by platelet aggregation then by fibrin clot formation. This supplies haemostasis and allows accumulation of neutrophils and monocytes, which herald the inflammatory phase. The inflammatory phase provides phagocytosis of bacteria, other foreign matter, and dead tissue in the wound. The macrophages release factors that stimulate proliferation of local fibroblasts in the dermis. These provide a collagen network and stimulate new

vessel growth. This phase is characterised by pink granulation tissue and wound contraction. A warm moist environment that is supplied either by dressings or scab formation aids this process. Collagen synthesis reaches its peak towards the end of the first week of healing. Remodelling continues to occur for up to 12 months; thus the scar will usually fade and contract over the first 2 to 3 months, and the final appearance may not be obvious for up to 6 months post injury.

A number of factors affect the healing of a wound. Adequate nutrition (including vitamins C and A, which are required for collagen formation) is essential. Corticosteroids and immunosuppressive drugs interfere with cellular proliferation and immunity, and anticoagulants inhibit clot formation and initial wound stabilisation. Infection interferes with collagen synthesis and will delay wound healing and cause an increase in scar tissue formation.

Tensile forces of the surrounding skin affect the healing and scar formation of a wound. The most cosmetically pleasing outcome occurs when the wound is parallel to the direction of maximum skin tension – along Langer's lines of skin tension. Wounds that intersect perpendicularly to the lines of skin tension will heal with greater scarring; however, there is significant inter-child variability. Dynamic skin tension caused by joint movement may impair wound healing and increase scar formation, and immobilisation of joints while the laceration heals will minimise this effect.

Wound infection

Wound infection is relatively uncommon, occurring in about 5% of wounds presenting to emergency departments (EDs). In general, a wound in a child is less likely to become infected than a similar wound in an adult. Identified risk factors for infection include severe wound contamination, inadequate wound cleansing, inadequate debridement of dead tissue (especially in crush injuries), use of subcutaneous sutures, larger laceration (>5 cm) and site of injury. Specific sites identified as infection risks include axillae, perineum or groin, and feet. In general, limb wounds are at increased risk compared to head and neck wounds.[3,5,6]

Classification of wounds

There are three common classifications of wounds: lacerations, incised wounds and abrasions. As a general rule, if the wound penetrates into the dermal

capillaries it will bleed, and if it extends into the subcutaneous tissue it will gape. The first classification of wound, laceration, is the most common type of wound seen in the paediatric age group.[6] Lacerations can be caused by tension on the skin (usually seen in areas with significant subcutaneous tissue) or by compression of the skin between an object and bone resulting in wound edges which are ragged. There is always damage done to surrounding tissues, and healing is therefore delayed. Compression injuries usually have more surrounding tissue damage and thus tend to heal more slowly.

In contrast, a sharp object such as a knife blade or glass shard makes an incised wound. The wound margins of an incised wound are clearly defined and there is little or no surrounding tissue damage. Incised wounds heal faster than lacerations and, in general, have a lower incidence of infection.

Finally, abrasions are wounds caused by sheering forces on the surface of the skin. The upper layers of the epidermis and sometimes dermis are scraped away. The depth of injury usually varies throughout the wound. If the epidermis alone is involved there is no bleeding but a transudation of fluid. If the dermis is involved the wound will bleed, and there is said to be an increased incidence of infection and foreign body retention.

Evaluation of the patient with a laceration

The care of the patient should follow standard trauma protocols. The airway, breathing and circulation should be assessed and treated as appropriate and a thorough secondary survey undertaken in most patients to exclude or allow management of serious injuries as well as detecting other minor injuries.

History

The mechanism of trauma (cut, crush, fall, bite, burn) and the time of injury are important as they may alter the management of the wound. For example, crush and bite injuries characteristically cause significantly more surrounding tissue damage and thus are more likely to have delayed healing or infection. When possible, determine the cleanliness of the inflicting object, the amount of blood loss, the presence of a foreign body sensation, and the motor function and sensation distal to the affected area. The location of the wound should be noted and the possibility of injury to other structures considered.

Obtaining a thorough medical history may reveal chronic illnesses that may

impact wound healing – such as diabetes mellitus, obesity, malnutrition, chronic renal impairment, cyanotic congenital heart disease, chronic respiratory illness, tumours, and bleeding disorders.[3] Immunisation history should be obtained and further tetanus vaccination guided by the recommendations of the National Health and Medical Research Council (Table 4.8.1).[7] Awareness of medications for potential drug interactions with prescribed antibiotics and of medications that may interfere with wound healing – such as immunosuppressive drugs and corticosteroids – is important. A history of allergies must be determined prior to use of cleansing agents, dressings and tapes and prescription of medication. A history of latex allergy should be specifically sought. In wounds that require management under general anaesthesia or sedation a history of when the child last ate or drank is important. Non-accidental injury should always be considered in the vulnerable population.

Table 4.8.1

Tetanus prophylaxis in wound management

History of tetanus vaccination	Time since last dose	Type of wound	DTPa, DTPa-combinations, dT, dTpa, as appropriate	Tetanus immunoglobulin (TIG)*
≥3 doses	<5 years	Clean minor wounds	No	No
≥3 doses	<5 years	All other wounds†	No	No‡
≥3 doses	5–10 years	Clean, minor wounds	No	No
≥3 doses	5–10 years	All other wounds†	Yes	No‡
≥3 doses	>10 years	Clean minor wounds	Yes	No
≥3 doses	>10 years	All other wounds†	Yes	No‡
<3 doses or uncertain§	>10 years	Clean, minor wounds	Yes	No
<3 doses or uncertain§	>10 years	All other wounds†	Yes	Yes

* The recommended dose for tetanus immunoglobulin (TIG) is 250 IU, given by intramuscular injection, as soon as practicable after the injury. If more than 24 hours have elapsed, 500 IU should be given. Because of its viscosity, TIG should be given to adults using a 21-gauge needle. For children, it can be given slowly using a 23-gauge needle.

† All wounds, other than clean minor wounds, should be considered ‘tetanus-prone’.

‡ Individuals with a humoral immune deficiency (including HIV-infected persons who have immunodeficiency) should be given TIG if they have received a tetanus-prone injury, regardless of the time since their last dose of tetanus-containing vaccine.

§ Persons who have no documented history of a primary vaccination course (three doses) with a tetanus toxoid-containing vaccine should receive all missing doses and must receive TIG.

Examination

Once assessment and management of more serious injuries have occurred, the patient should be assessed for the current severity of any chronic illness and appropriate management initiated.

The cooperation able to be gained and comprehension level of the child

influence wound examination and the information gained. Distraction techniques, adequate topical anaesthesia and appropriate use of sedation can all aid in wound assessment. A calm, unhurried, friendly approach, involving the parents, will maximise the chances of cooperation. Adjuncts like audio-visual equipment and Child Life therapists should be employed if practicable. Examination of the wound should be done with optimal lighting and with bleeding minimised. Examination of function, sensation and circulation distal to the wound is best performed prior to exploration of the wound and prior to regional anaesthesia.[8–10]

Functional assessment requires the movement of all joints distal to the wound. In an older child each joint is examined individually on command and the strength documented. In the child less than 5 years old or when the ability to partake in the physical exam is limited, observation of posture, symmetry and general movement is a surrogate to a traditional exam. In wounds in which the flexor tendons of the hand are concerning, close attention should be paid to the resting position of the fingers (partial flexion). The finding of extension of one finger at rest and the failure of the finger to flex at play or after application of a noxious stimulus strongly suggest a tendon injury.

Injury to nerves is classically assessed with two-point discrimination, and this should be possible in older children. Using a paperclip bent so that its ends are separated 4–8 mm is useful in this process. In upper limb injuries formal assessment of the median, ulna and radial nerves is required. In children less than 5 years old this approach needs to be modified. A noxious stimulus applied to the fingers will illicit sensation but risks losing patient confidence. Although impractical, a method of determining intact innervation is to look for sweating of the fingers. Since autonomic response includes sweating, denervated fingers do not sweat. An ophthalmoscope can be used to examine for sweat beads, or the cleaned body of a pen can be run over the fingers, with less resistance in the denervated, thus dry, segment. Arterial circulation can be assessed by palpation of peripheral pulses, capillary return distal to the injury, and skin colour and temperature.

Assessment of the wound should include site, size, depth, nature of the edges, cleanliness, and presence of foreign bodies. The wound should be explored to determine the depth and involvement of any underlying tissues including vessels, nerves, tendons, ligaments, muscles, joints, bones and specialised tissues (especially ducts and glands). Bones adjacent to the wound should be palpated for deformity or crepitus and the wound searched for foreign bodies (including

the sound of glass on the metal forceps). This assessment and exploration should take place after appropriate anaesthesia of the wound and any required sedation.

Investigation

If presence of a soft tissue foreign body is suspected and not visualised on examination/exploration, radiological investigation can be performed. In wounds caused by glass, all but superficial wounds should be investigated with plain soft tissue X-ray of the region to exclude a glass foreign body. Most glass foreign bodies more than 2–3 mm in size should be visible. If a radiolucent foreign body is suspected, ultrasound can be useful to both confirm the presence of the foreign body and provide a guide to its depth and location in the wound.[9–11] Other investigations should be determined by the findings of possible injuries to adjacent structures, such as bony X-rays for fractures.

Treatment of wounds

Wound anaesthesia

Analgesia and sedation are discussed in more detail in Section 20. Anaesthesia is required to adequately examine and then treat most wounds. Often, in children, procedural sedation will be necessary – depending on the location of the wound, the involvement of underlying structures, and the age and anxiety of the child.

The options for anaesthesia include topical, regional, dissociative, general, or local infiltration. An alternative and adjunct to the above options for anaesthesia is *procedural sedation.* Procedural sedation is the practice in which the clinician administers sedatives and analgesics to achieve a depressed conscious state to allow tolerance of a painful procedure without impairing cardiopulmonary function.

There are a myriad of commercially available topical anaesthetics, some of which include a vasoconstrictor adrenaline (epinephrine) or, less often, cocaine. The vasoconstricting properties of adrenaline-containing anaesthetics resulted in dogma that its use in areas of end arteries (finger tips, nose, lips, ears, genitalia) was not permissible. However, it is now thought that the resulting vasoconstriction is not as detrimental as previously thought, and it can be used with caution. Some anaesthetics can be applied in the wound either as a liquid dripped onto a pledget of cotton wool placed into the wound or as a methylcellulose gel. The wound is then covered with an occlusive impermeable

dressing, and adequate anaesthesia is usually obtained within 30 minutes.[12–17]

Local infiltration is the classic method of anaesthetising a wound. The anaesthetic is injected into the wound margins. Pain of injection can be minimised by using warmed anaesthetic, buffering the drug with sodium bicarbonate (mix 10 mL of 1% lignocaine [lidocaine] with 1 mL of 8.4% sodium bicarbonate), infiltrating slowly, using the lowest concentration possible, infiltrating into injured, rather than intact skin and using needles sized 25 gauge or smaller. The most commonly used local anaesthetic is lignocaine 1% or 2% with or without adrenaline 1:100,000. The onset of action is rapid, with duration of action of 30 minutes to 1 hour. Addition of adrenaline is useful to prolong the duration of action and help minimise bleeding; however, adrenaline should be used with caution in regions of end arteries (fingers, nose, lips, ears, genitalia), and its use may increase the risk of infection. The safe dose of lignocaine is 3 mg kg^{-1} or 6 mg kg^{-1} for lignocaine mixed with adrenaline.[3]

Regional anaesthesia is useful for facial, hand and foot lacerations, where nerves are readily accessible near bony landmarks. A regional nerve block involves anaesthetising the nerve or nerves that supply a specific anatomic region. Regional anaesthesia is especially useful for large lacerations and lacerations where local infiltration causes distortion of tissue anatomy. Regional anaesthesia is especially useful for anaesthetising digits. Lignocaine or bupivacaine hydrochloride 0.5%, which has duration of action of 3 to 6 hours, is most commonly used. The safe dose of bupivacaine is 2 mg kg^{-1}.

Procedural sedation is occasionally required when treating lacerations in children. Options for sedation include benzodiazepines – such as midazolam or diazepam, fentanyl, nitrous oxide, ketamine, or propofol. Sedation should only be undertaken by personnel experienced in its use and able to manage the complications of airway compromise, oxygen desaturation, hypercapnia and respiratory depression. Adequate equipment to deal with these complications should also be available. Some form of physical restraint may also be necessary to prevent excessive movement during repair; however, the aim must be to provide adequate analgesia and anxiolysis.[13,18]

Wound preparation and cleansing

Hair near the wound should only be removed if it interferes with the meticulous closure of the wound. If hair removal is desired the hair should be clipped, not shaved, as shaving disrupts hair follicles and increases the incidence of wound

infection.[19] Eyebrow hair should not be removed because this may lead to abnormal or delayed regrowth.

The surrounding skin and wound edges should be thoroughly cleaned. This should be undertaken in a manner and with an agent that provides adequate antisepsis without tissue injury or impairing wound defense mechanisms. A solution such as aqueous povidone-iodine or aqueous chlorhexidene applied with gauze or cotton wool should be used. Care should be taken to minimise the amount of cleanser to penetrate the wound to minimise damage to wound defenses, increasing the risk of infection.

Surgical debridement of crushed or non-viable tissue is vital to prevent wound infection or delayed wound healing. However, as little tissue should be debrided as possible. Manual removal with forceps of large particles of foreign material should also be meticulously undertaken. When a heavily contaminated wound contains specialised tissues such as tendons or nerves, consultation is recommended.

Once the wound is adequately anaesthetised it should be thoroughly cleaned. Irrigation is the method of choice for removing dirt and bacteria from wounds. In hospital, saline (0.9%) is the irrigation solution of choice, as it causes no tissue damage, but tap water can be used.[20] The ability of irrigation to decontaminate a wound is directly related to pressure of the irrigating stream, the size of the particles to be removed, and the volume of irrigant. At least 100–200 mL per 2 cm of laceration is required. The fluid should be injected from a 30–60 mL syringe via an 18- to 20-gauge cannula. Higher pressures should be avoided as they may cause tissue damage and increase the incidence of wound infection.[21,22] The volume and pressure of irrigation should be modified as necessary according to the location and cause of the wound. High-pressure irrigation does not enhance the dissemination of bacteria into soft tissue wounds, but excessive use can cause local tissue oedema increasing risk of infection. Use of a device to minimise splashing of the irrigant is desirable and wearing of gloves, goggles and gown prudent.[21,22]

Antibiotic prophylaxis

The use of prophylactic antibiotics in wound care is controversial. Decontamination with appropriate irrigation techniques is more effective than the use of prophylactic antibiotics.[2,9,23,24] When indicated (Box 4.8.1), antibiotics should be given as soon as possible. The initial dose should be given

intravenously and dosed on the higher limit of weight-based dosing to provide rapid reliable high tissue concentrations. The first dose should be given before wound closure to ensure an effective concentration of antibiotic in the wound tissue fluid at the time of wound closure. When choosing an antibiotic the likely causative organisms should be considered: the organisms contaminating the wound and the concomitant organisms found in that region of the body. In general, bites and wounds in regions with high bacterial counts (hands, feet, groin) should be treated with antibiotics to cover *Staphylococcus epidermidis, S. aureus* and *Streptococcus* sp. The likelihood of anaerobic bacteria should be considered. Specific circumstances also need to be considered. Patients at risk of endocarditis should have all wounds treated with antibiotics to cover *S. aureus* and *S. epidermidis*. Ampicillin/amoxicillin is the currently recommended drug in Australia. However, in communities where the incidence of penicillin resistance is high a cephalosporin and an aminoglycoside are recommended.

Wounds associated with fractures, tendon or muscle involvement should be considered for prophylaxis, as should large wounds, wounds with significant devitalised tissue such as crush injuries and stellate lacerations. Wounds contaminated with faeces should be treated with coverage of coliforms and anaerobic bacteria. Wounds in children with a compromised immune system should all be considered for prophylactic antibiotics. Wounds with closure delayed more than 12 hours should also be considered high risk for infection. Treatment should be for 3 to 5 days with a penicillinase-resistant antibiotic such as a first-generation cephalosporin or amoxicillin-clavulanic acid, with or without metronidazole.[25]

Box 4.8.1 Indications for antibiotic prophylaxis in wounds

Wound characteristics

High risk anatomical site (hands, forefoot, groin, axilla)
Devitalised tissue
Extensive surrounding soft-tissue injury
Stellate lacerations
Contaminated with body fluids or organic matter or dirt
Large lacerations (>5 cm)
Closure delayed (>12 hours)

High risk for endocarditis

Prosthetic heart valves
Patent ductus arteriosis
Structural heart disease: tetralogy of Fallot, ventricular septal defects, coarctation of the aorta, damaged heart valves
Immunocompromised children
Prior history of endocarditis
Intravenous drug use

Wound closure

The aim of wound closure is to reduce discomfort, aid healing and produce the best cosmesis.[26] The technique chosen for wound closure depends on the type of wound. Most wounds in children can be managed with primary closure, as the risk of infection is relatively low. Infected, heavily contaminated wounds and wounds resulting from high-energy projectiles are best managed by delayed primary closure, with initial cleansing and packing then closure 3 to 5 days later, once the risk of infection has decreased. Wounds with delayed presentation (>24 hours) or those contaminated with saliva or faeces should also be considered for delayed closure. Some wounds, such as puncture wounds or contaminated wounds in areas of poor perfusion, should not be closed but allowed to heal by secondary intention. Once it is decided to close the wound, a technique that allows apposition of the wound edges that is secure and accurate and holds the wound edges in apposition until the strength of the wound is sufficient should be chosen. With improved technology the options for wound closure are growing. Those presently available include sutures, staples, tissue adhesives and tapes.

Sutures

Suturing is the traditional method of wound closure. Sutures are divided into two classes on the basis of their degradation properties. Absorbable sutures degrade rapidly in vivo and lose their tensile strength within 60 days. Sutures that degrade more slowly are classified as non-absorbable (Table 4.8.2 for individual suture material characteristics).

Absorbable sutures are made from either collagen or synthetic polymers. Gut

sutures are manufactured from the submucosa of ovine or bovine intestines. The collagen is then treated to strengthen the material and increase resistance to tissue degradation (plain gut). Coating with chromium trioxide provides more resistance to absorption (chromic gut). These suture materials have a somewhat unpredictable absorption. Synthetic absorbable sutures have improved strength with delayed and more reliable absorption characteristics. Absorbable sutures are used for closing deep layers of a laceration and can be used for skin closure – especially where removing sutures in a young child may be difficult.

Non-absorbable sutures are made from either natural (silk, cotton, linen) or synthetic (nylon, Dacron®) fibres. They can also be classified according to their physical characteristics. Monofilament sutures are made from a single filament (nylon, Prolene®), and sutures containing multiple fibres are called multifilament (silk, cotton, nylon). Of these sutures, only nylon is available in both types of filament. Non-absorbable sutures are used to close fascial layers (where healing is slow) and for skin closure.[27,28]

Table 4.8.2

Characteristics of common suture materials

Suture material	Ease of handling	Tensile strength	Degradation (d)*	Tissue reactivity	Infection potential
Non-absorbable					
Nylon (Ethilon®, Dafilon®)	Average	Good	–	Low	Very low
Polypropylene (Prolene®)	Poor	Very good	–	Very low	Low
Silk	Good	Poor	–	High	High
Absorbable					
Surgical gut (fast absorbing)	Poor	Average	4–7	High	High
Polyglactin (Vicryl Rapide®)	Average	Good	7–10	Low	Low
Chromic gut	Average	Average	10–14	High	High
Polyglactin (Vicryl®)	Average	Good	10–15	Low	Low
Polyglycolic acid (Dexon®)	Good	Good	25–30	Low	Low
Polydioxanone (PDS)	Average	Very good	25–30	Very low	Low

* Time to loss of 50% of tensile strength.

Sutures come in varying sizes. Suture size selection depends on the wound location and the tensile strength of the tissue to be sutured. Heavy sutures such as 4–0 should be used in the limbs and trunk and should also be used on mucous membranes and subcutaneous tissue. Heaviest sutures such as 3–0 should be used on thick skin (such as the sole of the foot) or over large joints. Small sutures such as 6–0 should be used on tissues with light tensions, such as facial skin and subcutaneous tissue.[27,28]

Needles

Needles come in varying sizes and shapes also. Needles are described by the arc

of curvature the needle possesses and the shape of the needle itself. The most commonly used for skin closure is the ⅜ circle (135°) needle or the ½ circle (180°) needle (Fig. 4.8.1). For closure of fascial layers ½ circumference needles are usually used. Needles that have two circumferences of curvature (compound needles) are able to be passed through the tissue with less rotation of the operator's forearm. Needles come with different shapes as well as curvatures (see Fig. 4.8.1). A reverse cutting needle is the most common type used for skin closure. The needle cuts an inverted triangle, and the cutting edge is on the underbelly of the needle, decreasing the likelihood of 'cutting out' contrasting with that of the conventional cutting needle which has a cutting edge pointed upwards towards the tissue surface and thus increasing the likelihood of 'cutting out' of the tissue. For fascia a taper point needle is used. The cross-section of these needles is a circle that is tapered to a point. It does not cut but pushes the tissues aside, causing less tissue damage and reducing the likelihood of the needle cutting out. For deep tissues that are stronger (such as tendon) a tapercut, or combination needle, is used – it has a tapered body, but the point is a reverse cutting edge.[28]

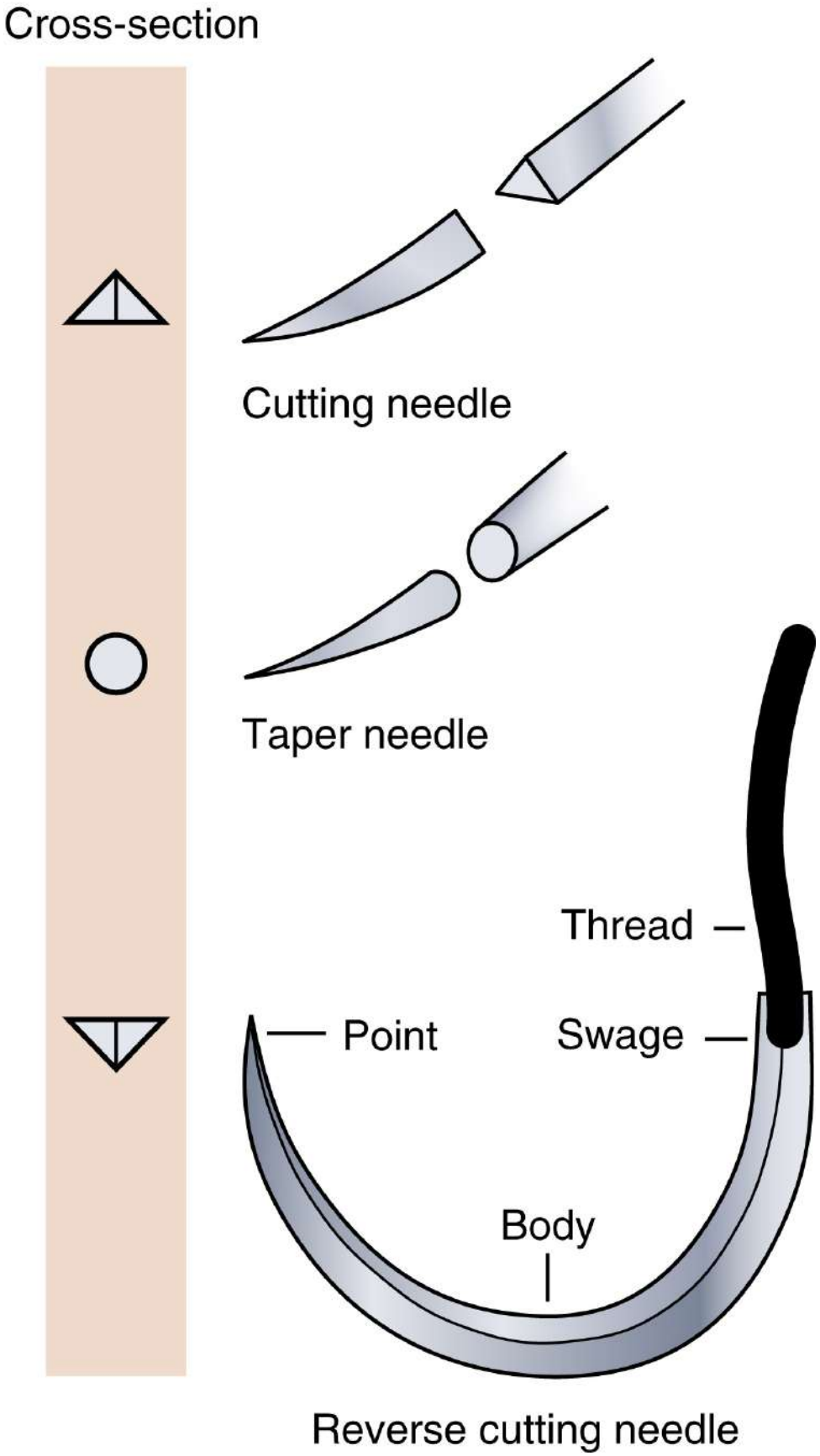

Cross-section
Cutting needle
Taper needle
Thread
Point
Swage
Body
Reverse cutting needle

FIG. 4.8.1 Surgical needle characteristics and types. From an original drawing by Elaine Wheildon.

Needles are grasped with a needle holder. The swage of the needle – the region where the needle is hollowed out to join with the suture – is the weakest point, and grasping the needle in this region should be avoided. The needle should be grasped in the body one-half or two-thirds of the distance from the tip of the needle.

Suturing techniques

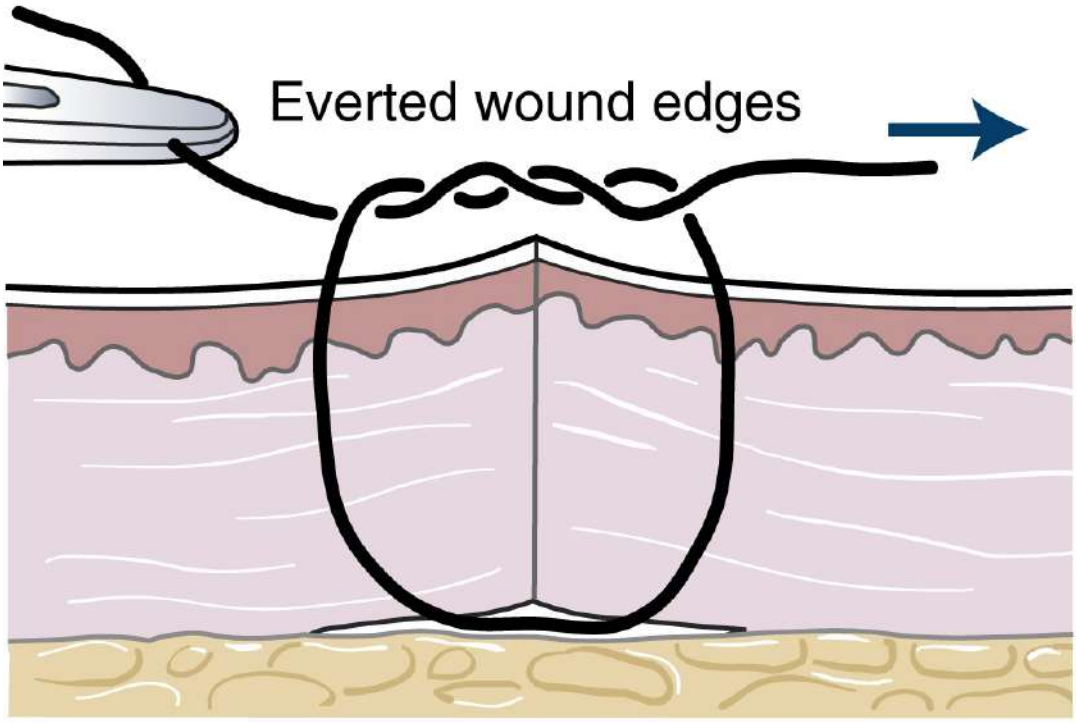
For closure of a wound with sutures, a number of instruments are needed to maintain a sterile field and to allow manipulation of the tissues and needle (Box 4.8.2). Finer instruments should be available for facial laceration repair.

Sutures should be placed to allow apposition of all injured layers of the skin. Proper suture placement should result in slight eversion of the wound edges, avoiding a depression of the scar when contraction takes place during wound healing. To ensure eversion of skin edges the skin suture must be placed so that an equal amount of tissue is included on each side of the wound and so that the needle bite includes a broad base (Figs. 4.8.2 and 4.8.3). This is accomplished by lifting the wound edge as the needle is passed through the skin on each side, maximising the deep tissues included in the suture.[28,29]

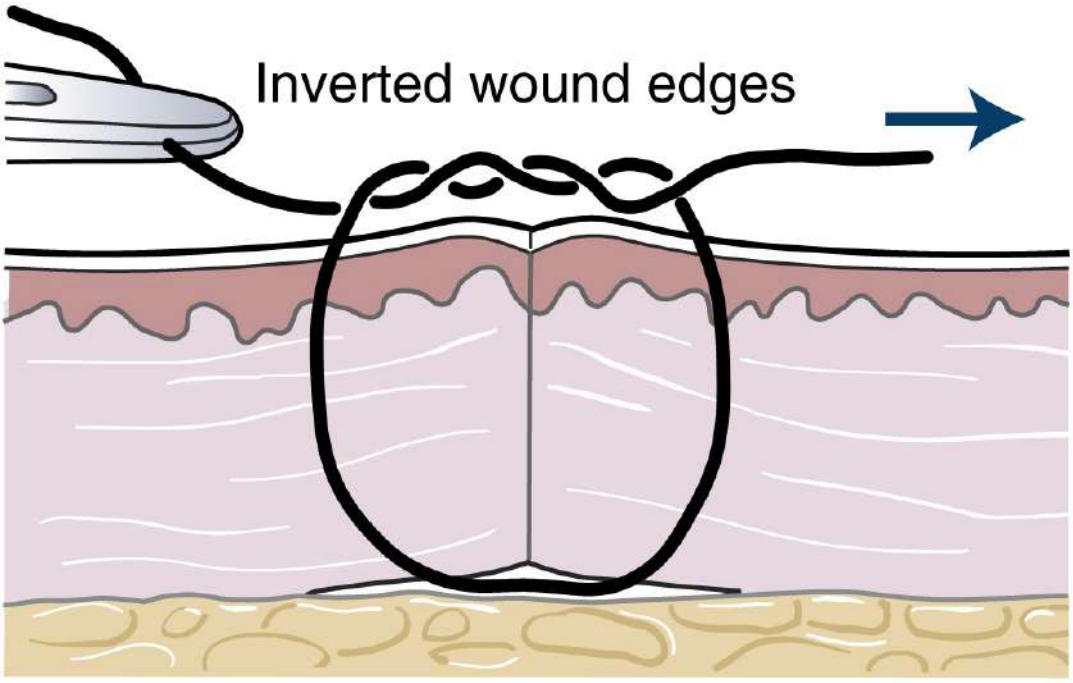
Most wounds sutured in the ED are closed with interrupted skin sutures. Synthetic non-absorbable sutures are most commonly used. However, rapidly absorbable sutures can be used to close the skin in children, avoiding the discomfort of suture removal. To place a simple interrupted suture the needle is held so the tip enters the skin at a right angle, and the hand is rotated to ensure the needle remains at right angles to the skin throughout its passage, which aids in maximising the deep tissues captured in the bite. The stitch should be secured with an instrument tie and the knots secured to one side of the wound to minimise inflammation to the healing tissue. The initial throw should include two wraps of the suture material around the needle holder; subsequent throws should be wrapped once. The knot should be tied just tight enough to oppose the skin edges. Tying the knot too tightly will cause a reduction in the blood supply to the wound edges and increase the risk of infection and poor cosmetic outcome. Synthetic sutures with poor handling should have four or five throws per knot.[28–30]

Box 4.8.2 Example of instruments required for a simple suture tray

1 × Needle holder (Halsey or Hegar)
1 × Toothed dissecting forceps
1 × Curved artery/mosquito forceps
1 × Straight artery/mosquito forceps
1 × Suture scissors



Correct



Incorrect

FIG. 4.8.2 Normal suture. Suturing technique for wound edge eversion.

The more sutures placed per centimetre, the finer the control over the wound edge. For facial lacerations, the skin sutures should be placed approximately 3 mm apart and enter the skin about 3 mm from the wound edge. For other areas of the body, sutures should be placed 4 mm to 5 mm apart and should pierce the skin about 5 mm from the wound edge. The number of sutures used to close a wound should be the minimal number that allows a desired cosmetic outcome. In general, the better the blood supply, the closer together sutures can be placed.

There are generally two methods for closing a laceration: either suturing from one end to the other or placing sutures that serially bisect the wound. A small linear wound is easily sutured from end to end, and long wounds without good landmarks on either side are most easily closed by placing the first stitch in the middle and then serially subdividing the wound. In wounds with definite landmarks, such as palmar skin creases or the vermillion border of the lip, the first suture should be placed to align these landmarks.[28,30]

Deep sutures should be placed where there are multiple layers of tissue involved and the skin sutures would be under tension. They are placed to reapproximate the dermal layers of the skin and remove skin tension, thus improving cosmetic outcome. Placing deep sutures inserts a foreign body into the wound and increases the risk of wound infection, so they should only be placed when necessary and the minimum number necessary used. For this reason, deep sutures should be avoided in the hands and feet. Deep sutures placed close to the skin are sometimes extruded through the wound. To place a deep suture, the needle is placed at the depth of the wound and removed at a more superficial level. The needle is then placed at the same superficial level on the opposite side of the wound and exits deeply so the knot is tied deeply in the wound (see Fig. 4.8.3).[28,30]

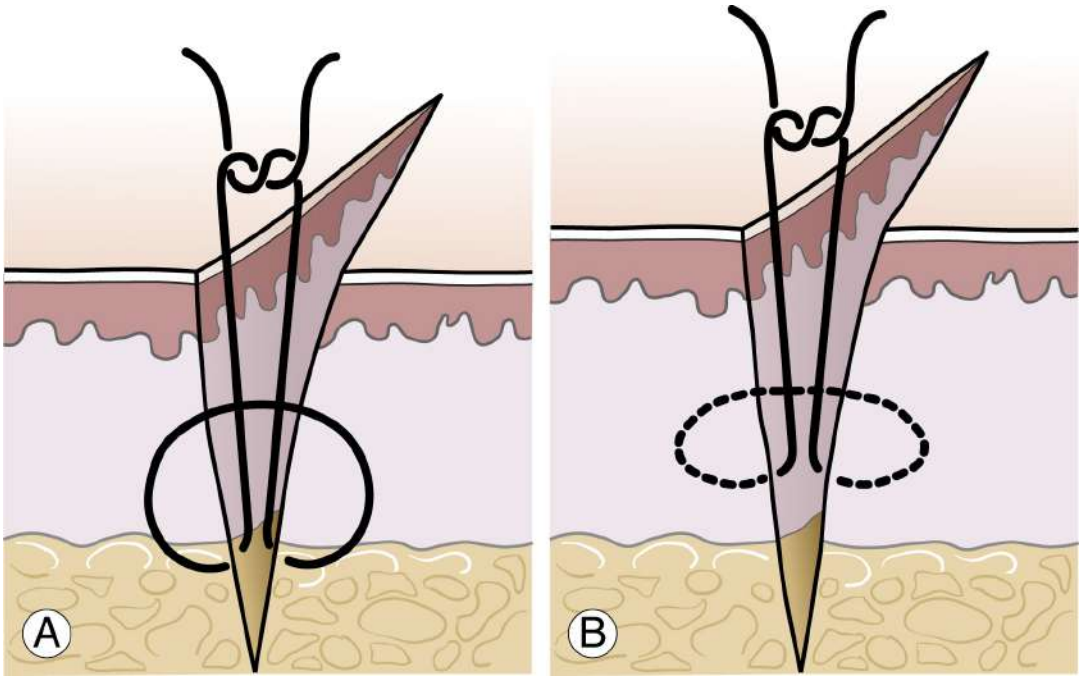


FIG. 4.8.3 Deep sutures. (A) The buried subcutaneous suture; (B) the horizontal dermal stitch.

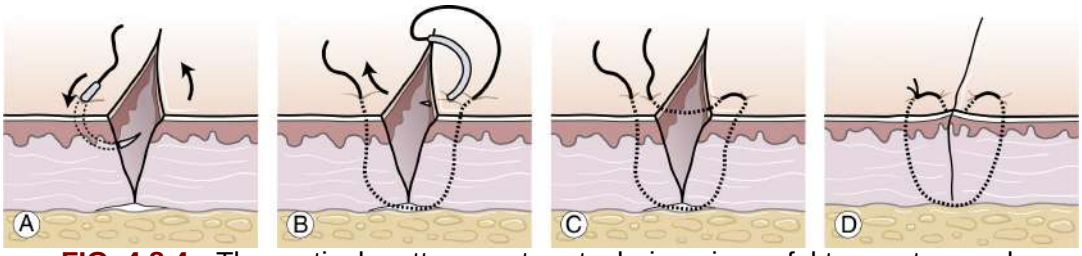


FIG. 4.8.4 The vertical mattress suture technique is useful to evert wound edges with a natural tendency to roll inwards despite correctly placed simple sutures. From an original drawing by Elaine Wheildon.

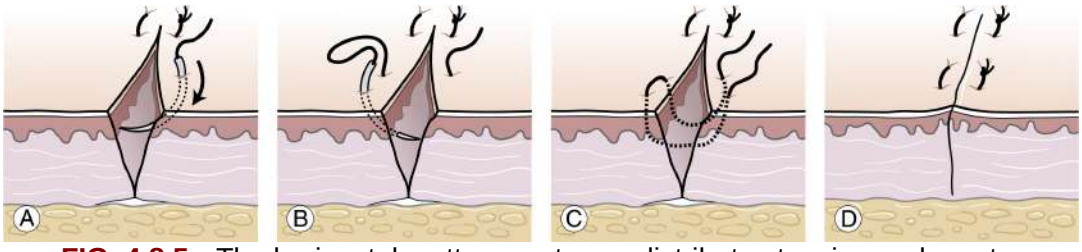


FIG. 4.8.5 The horizontal mattress suture redistributes tension and everts wound edges. From an original drawing by Elaine Wheildon.

Special suturing techniques

A variety of special suturing techniques are available with the sole purpose of aiding the provision of skin apposition with everted wound edges. The vertical mattress suture is useful in regions with minimal subcutaneous tissue where the edges are difficult to maintain in eversion. The technique is begun the same way as a simple skin suture, but after the suture loop is made, the skin is re-entered 1 mm to 2 mm from the wound edge and then tied (Fig. 4.8.4). The horizontal mattress suture reinforces the subcutaneous tissue and relieves skin tension but does not provide wound edge approximation as well as the vertical mattress suture (Fig. 4.8.5). The modified or half-buried horizontal mattress suture (or corner stitch) is the method of choice for closing a flap. It relieves tissue tension and avoids vascular compromise when approximating the tip of the flap (Fig. 4.8.6).[28,30]

A continuous suture can be used to close the laceration. It is faster to place than interrupted sutures, removal of sutures is easier and faster, and the tension is spread evenly along the wound. The continuous suture can be percutaneous or subcutaneous and made with absorbable or non-absorbable suture material. The disadvantages are that if one part of the suture breaks, the integrity of the whole wound is lost, and if the wound becomes infected, the whole wound needs to be opened to drain the pus. To place a percutaneous running stitch an interrupted suture is placed at one end of the wound, and only the free end of the suture is cut. Suturing is continued along the wound in a coil pattern ensuring that the needle passes perpendicularly across the wound with each pass. The loop is tightened after each pass and the last stitch placed beyond the end of the laceration. The stitch is tied using the last loop as the tail.[28,30]

Correction of dog ears

A dog ear is the term given to a conical pucker of redundant skin that may develop at the ends of the wound during suturing. It is avoided by suturing the wound from the middle by sequentially bisecting the wound. Dog ears can be removed in many ways, the simplest of which is the overlap excision technique. The redundant skin is pulled across the wound from one side and excised along the line of the wound. Redundant skin from the opposite side is pulled across the wound and also excised along the line of the wound. The wound is then closed (Fig. 4.8.7).[28]

Staples

Stainless steel staples can be applied more rapidly than sutures, and they are associated with a lower rate of foreign body reaction and infection. Staples are generally considered especially useful for lacerations of the scalp, trunk, and limbs. However, they do not allow such meticulous wound apposition as sutures and are slightly more painful to remove. They should not be used on the face or in any other wound where cosmetic outcome is a high priority.[31]

Tissue adhesives

Tissue adhesives have now been in use for several decades. The basis of the adhesive is a cyanoacrylate polymer. The cyanoacrylate polymerises in the presence of hydroxyl ions – found in water or blood – allowing it to bind to the skin. Tissue adhesives are for external use only and should not be placed within wounds or used on mucous membranes.

It is in the repair of lacerations to young children that tissue adhesives have become most popular. They are easy and relatively painless to apply and provide a cosmetic result that is as good as suturing, with no risk of causing suture marks. No removal is required, as they slough off in 7 to 10 days. They are, however, not suitable for use in all wounds. If the laceration cannot be approximated and the wound edges brought together with minimal tension, then tissue adhesive is not appropriate. Care should be taken not to apply too much tissue glue and to avoid placement over currently bleeding wounds, as the polymerisation is exothermic, and the patient may notice a heat sensation. Also, contact with excess blood causes polymerisation above the skin, limiting the tensile strength of wound edge closure.

The tissue glue is applied over the surface of the wound once its edges have been approximated by digital pressure. A number of thin layers of glue are applied across the wound and the wound held approximated for about 30 seconds. Care is taken not to allow glue to spill into eyes or orifices and to avoid fixing forceps or gloves to the patient. The cyanoacrylates also act as their own dressing providing moist wound healing conditions under the glue and have a degree of intrinsic antimicrobial activity. Careful attention to wound cleansing is still needed to avoid wound infections. In general, they are less expensive than sutures or staples and are strongly preferred by patients and families. A comparison of the various methods of wound closure is found in Table 4.8.3.[32–34]

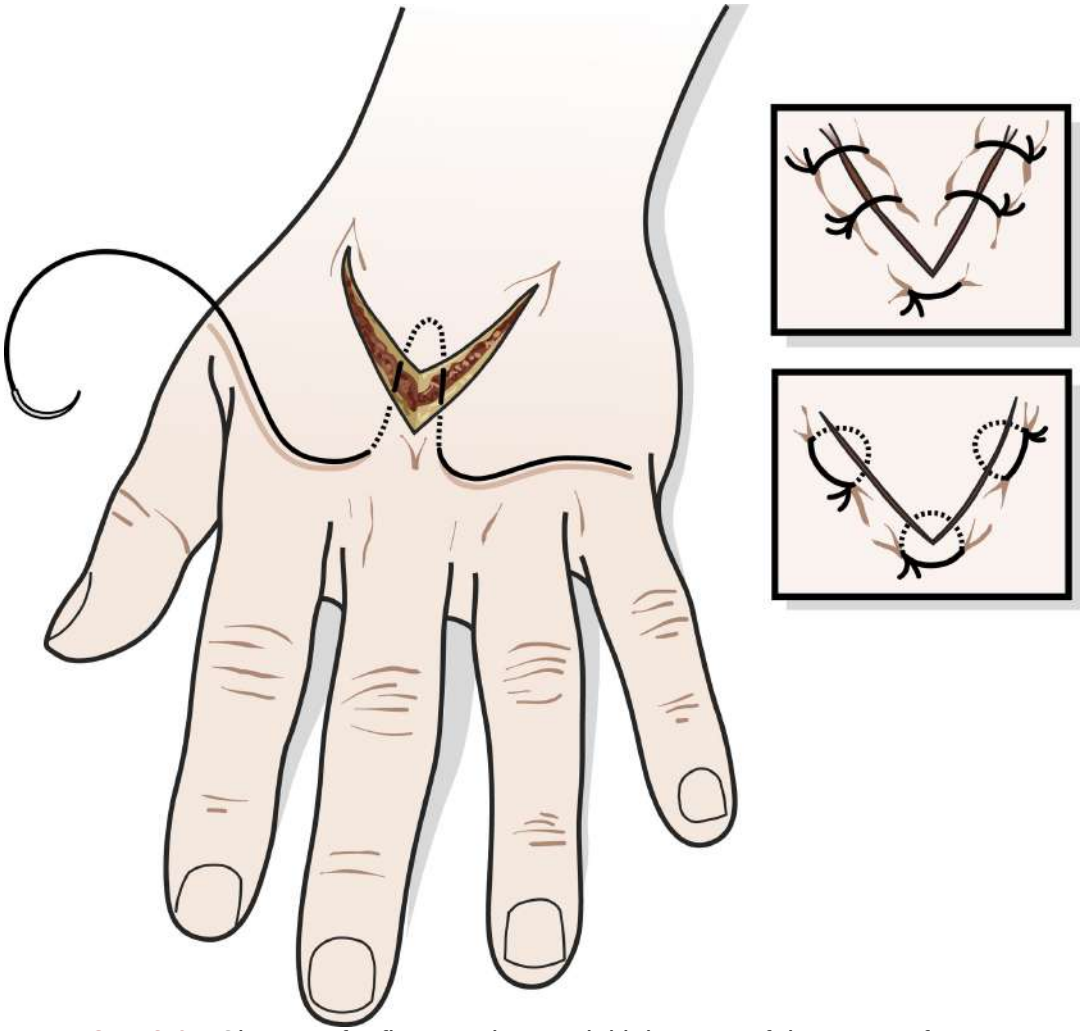


FIG. 4.8.6 Closure of a flap requires an initial suture of the apex, after which either simple or horizontal mattress sutures may be used. From an original drawing by Elaine Wheildon.

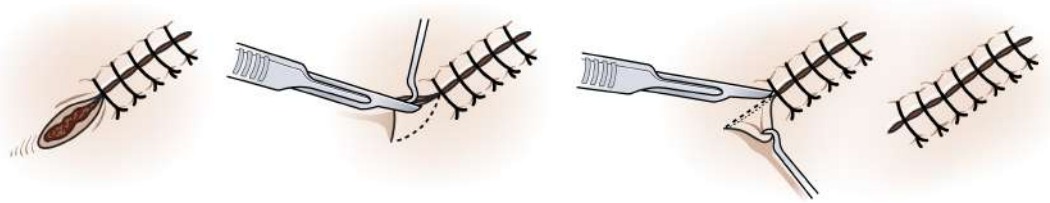


FIG. 4.8.7 Dog ear. Direct overlap excision technique. From an original drawing by Elaine Wheildon.

Skin tapes

Skin tapes can be used to close small wounds with low tensile forces. They cannot be used over areas of motion such as over joints. Wound haemostasis is vital as the tapes will not stick to wet skin. Application of an adhesive agent to the skin adjacent to the wound is necessary to provide adequate adhesion of the tapes. These adhesive agents (e.g. Tinc Benz®) are toxic to tissues and cause pain, so great care should be taken not to spill them into the wound. Tapes are not suitable for use in small children as they frequently pull them off. Tapes are a useful adjunct to wound closure – for example after suture removal or tissue adhesive application to decrease tension on the wound.

Post-wound-closure care

All patients should be provided with written information on care of their wound. Parents and children must understand the importance of ongoing wound care and be provided with instructions about follow-up.

Dressing and suture removal

After the wound has been sutured it should be covered with a non-adherent occlusive or semi-occlusive dressing to protect the wound from bacterial invasion and provide a moist healing environment and speed wound healing. Ideally, the dressing is left intact until suture removal. The dressing should only be removed if it becomes saturated or there is a risk of infection and inspection is warranted. If the wound is not covered with a waterproof dressing the dressing can be removed every few days for showering. Non-absorbable sutures should be removed at the appropriate time, depending on the location of the injury. Removal of the sutures too early risks dehiscence; leaving sutures too long increases tissue reaction and the risk of cross-hatching and wound infection (Table 4.8.4). In general, sutures are removed earlier in children than in adults. Wounds closed with tissue adhesive should not be covered with an occlusive dressing, as the extra moisture will more rapidly decrease the ability of the glue to maintain wound edge apposition. The wound should be kept dry for 2–3 days, after which the patients may shower but should avoid bathing and swimming. Wounds closed with skin tapes should be kept dry to prevent premature removal.

Immobilisation and drains

Wounds that cross joints or are in areas of highly mobile skin should be immobilised. The joint should be splinted in the position of function for 7–10 days. Plaster of Paris can be used to make a cheap and easily applied splint, or a bulky dressing can be used to limit motion and prevent the child from tampering with the wound.

In general, drains should not be used in wounds that have been closed as they promote wound infection. If a wound is considered at high risk for infection, delayed primary closure should be undertaken rather than closure and drainage.[9]

Table 4.8.3

Characteristics of wound closure techniques

Technique	Advantages	Disadvantages
Suture	Greatest tensile strength Meticulous closure	May require removal Painful/requires local anaesthesia ± sedation Slow application Costly Slow to apply Increased tissue reaction
Staples	Rapid application Low cost Low tissue reactivity	Less meticulous closure Painful/requires local anaesthesia ± sedation Requires removal
Tissue adhesives	Painless Rapid application No removal needed Low cost No risk of needle stick	Lower tensile strength Not for use over joints Slightly higher incidence of dehiscence
Surgical tapes	Least reactive Rapid application Patient comfort No risk of needle stick Low cost	Low tensile strength Difficulty maintaining adhesion May require use of toxic adjuncts to aid adhesion

Table 4.8.4

Timing of suture removal

Wound location	Time of removal (days)
Face	4
Scalp	5
Upper limbs, trunk	7–10
Lower limbs	8–10

Over joints	10–14

Treatment of selected injuries

Abrasions

In abrasions, the underlying tissues are relatively uninjured, providing a degree of protection against infection. Cleansing the abrasion is important to flush away bacteria and remove particulate matter, which should be removed to prevent infection or tattooing. Large abrasions or those heavily contaminated may need cleaning under procedural sedation and, rarely, general sedation. After adequate cleansing and debridement the wound should be covered with a non-adherent occlusive or semiocclusive dressing. A moist environment enhances healing, and the environment under a scab is ideal. However, wounds without extensive scab formation are more comfortable, so a dressing provides the moist environment. Children with large or deep abrasions should be reviewed in 2 to 3 days to check the wound. Ongoing review should be once or twice weekly.

Eyelid lacerations

A thorough eye examination needs to be performed for all eyelid lacerations. Attention should be given to the possibility of a ruptured globe (eccentric pupil) or trauma to the globe (hyphaema, dislocated lens or retinal detachment). Visual acuity should be measured and documented. Any wound that penetrates the tarsal plate or the inner canthus requires specialist attention, as do wounds involving the lid margins. Any wound that cannot be adequately assessed (e.g. in a young child) should be referred for evaluation under anaesthesia.[24]

Superficial wounds of the eyelid are relatively easily repaired with 6–0 fast absorbing gut, with sutures placed close to the wound edge. Care must be taken not to suture into the tarsal plate or other deep structures.[24]

Lip lacerations

Inspection of the teeth and oral mucosa is mandatory in all lip lacerations. Tooth injuries should be documented and referred for management where necessary; missing teeth warrant investigation to ensure they have not been aspirated or imbedded in the soft tissues of the mouth. Ideal anaesthesia is via nerve block of the mental or alveolar nerves for the lower lip or the infraorbital nerve for the

upper lip. Alternatives include sedation and direct infiltration, with or without application of methylene blue to the margins of the vermillion border.

Wounds that involve the vermillion border (the junction of the dry oral mucosa and the facial skin) must be exactly realigned to achieve acceptable cosmetic results. A 6–0 suture should be used, with the first suture placed to precisely reappose the vermillion border.[24] Further sutures should be used to close the skin and the dry mucosal surface of the lip, with the wet mucosal surface only closed if it is gaping significantly.

Deep or through and through lacerations of the lip require deep sutures to repair the orbicularis oris muscle: if this is the case, an absorbable 6–0 suture should be used. The deep sutures should be placed after the initial suture is placed at the vermillion border and before tying that stitch.[24]

Tongue lacerations

Most tongue lacerations can be left to heal without intervention with good results. Large lacerations involving the free edge of the tongue should be repaired to avoid healing with a notch, interfering with the function of the tongue. Large flaps (that gap when the tongue is in the resting position) and lacerations that continue to bleed should also be repaired. Care should be taken when repairing these injuries because of the risk of airway compromise, especially considering moderate to deep sedation is likely to be necessary. General anaesthesia and repair in the operating theatre should be considered if there is doubt.

The tongue should be maintained in position by a gentle pull using a towel clip or 4–0 suture placed through the tip. Interrupted 4–0 absorbable sutures should be placed using full-thickness bites to include both mucosal surfaces and the lingual muscle in each stitch. Multiple knots should be used to secure the sutures and the parents warned that while the tongue is anaesthetised the child may bite through the stitch.[24]

Fingertip amputation

Young children frequently injure their fingertips in doors and windows. Most of the injuries in young children are contused lacerations or partial amputations, with complete amputation being less common. With similar injuries, the etiologies of older children’s injuries are more often injuries with knives or tools.

Fractures are less common in the older age group. These wounds should be evaluated for tissue loss and with radiography for bony injury.

If the amputated fragment has been retained and involves any of the nailbed, some surgeons reimplant as a graft, with approximately 50% chance of the graft taking. If the tissue is not retained or is small, and there is no bone on view, it is most appropriate to allow the wound to heal by secondary intention. Fingertips allowed to heal naturally have greater length and better sensory outcome than those treated with grafts. These wounds should be covered with a non-adherent occlusive or semiocclusive dressing to allow moist wound healing after thorough cleaning and debridement as needed. Follow-up should be maintained until the wound is healed.

Injuries involving just the fingertip but not the nail heal very well. Injuries involving the nailbed or nail, but sparing bone, heal well. Those that involve the nailbed, nail and distal phalanx heal less well. Any injury with bone on view should be referred for specialist care.[23,24,35]

Nailbed lacerations

Trauma to the distal fingers is often associated with nailbed injury. An underlying fracture of the distal phalanx should be assessed with radiographs. Unrepaired nailbed lacerations can permanently disfigure the growth of the new nail from the matrix.

If the nail is lacerated, completely avulsed, or only loosely attached, the nailbed must be explored. This can be done under local anaesthesia with a ring block of the digital nerves or under general anaesthesia. The nail must be removed and the nailbed repaired with 5–0 or 6–0 absorbable suture material. The space between the nailbed and nailfold must be packed with paraffin gauze or the nail replaced to prevent adhesions. If a fracture is present, antibiotics should be given. If the nail is partially avulsed only and is tightly adherent to the nailbed, it is reasonable to leave this intact as it will adequately splint and maintain apposition of any nailbed injury.[24]

Subungual haematoma

A subungual haematoma is a collection of blood between the nail and nailbed. It is most commonly seen with blunt fingertip injuries and may be associated with a fracture of the distal phalanx. Drainage of the haematoma usually provides

symptomatic relief and should be undertaken whenever the haematoma is causing pain. Generally no local anaesthesia is required to drain the haematoma with cautery or needle burring using a 19-gauge needle. If there is an underlying fracture antibiotics should be administered. Nail removal for inspection of the nailbed should not be undertaken, regardless of the size of haematoma.[24]

Puncture wounds to the foot

Puncture wounds to the foot carry a high risk of infection and retained foreign body. All puncture wounds should be assessed for retained foreign body with radiography and ultrasound for radiolucent foreign bodies. Anaesthesia with local infiltration or a posterior tibial nerve block is needed. The wound should be soaked to remove any scab on the surface, debrided and irrigated. The wound should be left open once any foreign material has been removed. The wound should be cleaned with an antibacterial solution (such as Betadine®) and dressed with a non-adherent dressing. Close review is important to detect infection early. Prophylactic antibiotics have not been shown to prevent infection and may predispose to *Pseudomonas* infection.

Bites

Animal bites

Animal bites are a common presenting problem for the ED. Dog and cat bites account for virtually all bites seen in the ED, with dog bites being about six times more common. Rodents and other animals account for less than 1% to 2% of bites.

Dog-bite injuries tend to be relatively large, relatively superficial crush injuries, which are seen most commonly on the face, neck and scalp in children. The overall infection rate for dog bites is about 10%, with facial wound infection rates of about 5%. Dog-bite wounds are infected with multiple organisms on all occasions, with both aerobic and anaerobic bacteria. It is reasonable to cleanse and close most dog bites, with antibiotic prophylaxis provided only to wounds that are high risk for infection (Tables 4.8.5 and 4.8.6). Amoxicillin with clavulanate is the most useful drug.

Cat bites, on the other hand, are typically puncture wounds with less surrounding tissue injury. They have bacteria inoculated deep into the wound, which is difficult to explore, irrigate or debride. The risk of infection is

significantly higher than in dog bite – at least twice as likely – because of the puncture-type wound, the most common location of the bite being the hand, and the high incidence (about 80%) of *Pasteurella multocida* found in cats' mouths. *P. multocida* is a facultative, anaerobic Gram-negative rod that often results in rapidly progressive cellulitis. It is sensitive to the penicillins and variably sensitive to macrolides and first-generation cephalosporins. All these drugs have been documented as adequately treating infections of *P. multocida*, but treatment failures have been documented for erythromycin and first-generation cephalosporins. It is recommended that all cat bites receive prophylactic antibiotics (see Table 4.8.5).[36,37]

Human bites

Most human bites are considered high risk of infection; however, they are not considered to carry a high risk of human immunodeficiency virus transmission. Prophylactic antibiotics have been shown to reduce the risk of infection. Appropriate prophylactic antimicrobial choices for human bite injuries include amoxicillin with clavulanate.[36] However, the clenched-fist injury (or fight bite), which commonly causes a ragged laceration over the fourth or fifth metacarpophalangeal joint, is at high risk of infection. These latter wounds should all receive prophylactic antibiotics, as should human bites (including self-inflicted bites) that have high-risk properties (see Tables 4.8.5 and 4.8.6).[36]

Table 4.8.5

Wound infection risk factors

	High risk	Low risk
Biting species	Cat Human	Dog Rodent
Location of wound	Hand Over a joint Below knee Through and through oral	Face Scalp Mucosa
Wound type	Puncture Extensive crush Old	Large Superficial Recent

Table 4.8.6

Management of bite wounds

Species	Suturing	Antibiotics
Dog	Yes	High-risk wound type only
Cat	Face only	All
Rodent	Yes	No
Human – hand bites	No	Yes
Human – other bites	Yes	Large wounds

Controversies

- Subcutaneous sutures close deep wound dead space reducing fluid collection and infection, but deep sutures of themselves can be a nidus of infection.
- Dressing practice for all wounds is changing to promote moist wound healing; this speeds the rate of healing and improves wound comfort.
- A number of topical anaesthetic creams are being used in extremity wounds despite not being licensed for this use.
- Wound drains are not indicated in the management of wounds in the emergency department, and delayed primary closure is a preferred technique.
- Debridement should remove as little tissue as possible to maximise cosmetic outcome.
- Prophylactic antibiotics, while intended to prevent infection, have been shown in some wounds to increase the risk of infection with unusual organisms.

Acknowledgement

Dr Ed Oakley contributed to this chapter in previous editions.

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SECTION 5

Cardiovascular

OUTLINE

5.1

Cardiovascular assessment and murmurs

Damayanthi Rasanathan, and Karen Quay

ESSENTIALS

1 Cardiac problems are uncommon in children but should be considered in an infant or child with respiratory distress, cyanosis, palpitations, syncope, or shock.

2 Key features of the cardiac examination are palpation of peripheral pulses, blood pressure measurement, auscultation for a murmur and characteristics of the second heart sound and palpation of the liver.

3 Murmurs are commonly found on routine examination of children.

4 Innocent murmurs share common characteristics.

5 Children with murmurs should be referred if they have:

- symptoms which may indicate cardiac disease (e.g. breathlessness, cyanosis, chest pain)
- abnormalities of the heart sounds (e.g. fixed splitting of the second heart sound)
- a murmur that cannot be confidently identified as innocent
- a murmur with a thrill, grade 4 intensity or greater.

6 Chest X-ray and ECG may help in suspected structural disease but are unlikely to be helpful in an asymptomatic child with a murmur.

Introduction

Approximately 1% of infants and children in developed countries have congenital cardiac problems. Acquired diseases include rheumatic heart disease, myocarditis, pericarditis, cardiomyopathies, and coronary vascular disease such as Kawasaki disease (see Chapters 5.6–5.8).

History

Presenting features of cardiac disease can vary depending on age of presentation and whether the lesion is congenital or acquired.

Congenital heart disease may present neonatally with cyanosis, at any age with symptoms related to cardiac failure, or with an incidental murmur heard during routine examination.

Most neonates with congenital heart disease are asymptomatic at birth. Infants with *duct-dependent left-sided obstructive lesions* usually present in the first 2 weeks of life as the ductus arteriosus closes, causing shock. Infants with *left-to-right shunting lesions* usually present after 4 weeks of age when pulmonary resistance has decreased and heart failure develops.

A child with an acquired cardiac problem may present at any age.

Key factors to consider in the history are the timing and onset of symptoms, maternal factors, perinatal factors and family history.

Important symptoms to consider in **infancy** are growth, feeding, presence or absence of cyanosis or respiratory distress. If cyanosis is described, it is important to determine whether it is persistent or intermittent and its relationship to crying, feeding and activity. Normal infants may appear peripherally cyanosed when cold or febrile. Babies with cardiac failure are often dyspnoeic and diaphoretic with feeds. Poor weight gain and difficulty completing feeds are often noted. Recurrent respiratory infections may be a manifestation of cardiac aetiology.

Common symptoms in **older children** are poor exercise tolerance, fatigue, dyspnoea, orthopnoea and palpitations. Most presentations of chest pain in children are not caused by cardiac disease.

Details about the pregnancy are relevant for infants with cardiac problems. Maternal diabetes is associated with increased risk of structural heart disease and transient cardiomyopathy. Congenital heart block occurs in infants of mothers with lupus. Teratogenic drugs during pregnancy may cause heart disease. Infections during pregnancy may also carry an increased risk of congenital heart disease. As an example, rubella is associated with patent ductus arteriosus

(PDA) and peripheral pulmonary artery stenosis. Perinatal factors such as foetal distress and asphyxia may cause an ischaemic insult and cardiomyopathy.

Family history of congenital heart disease confers an increased risk to subsequent children. Most congenital cardiac defects are multifactorial, and the risk of another sibling being affected is around 1–3%.

When assessing an older child, a family history of sudden death, epilepsy or arrhythmia should be elicited. In most families with congenital prolonged QT, there is a family history of sudden or premature death or recurrent syncope.

Several diseases have an autosomal dominant pattern of inheritance, including hypertrophic obstructive cardiomyopathy (HOCM), supravalvular aortic stenosis, Marfan's syndrome, idiopathic mitral valve prolapse and some cases of atrial septal defect (ASD) and prolonged QT.

Physical examination

Key features in examining a child's cardiovascular system are careful assessment of general status, palpation of pulses, precordium and abdomen, and auscultation of the heart and lungs.

A general assessment of the child comes first. Note whether the child appears well and has any dysmorphic features and assess whether growth is appropriate for age. Assess for *central cyanosis* by looking at the tongue and mucous membranes. *Peripheral cyanosis* occurs when there is poor tissue perfusion. *Acrocyanosis* occurs in neonates shortly after birth due to cold and decreased tissue perfusion. Clubbing is a sign of chronic cyanosis and may be seen in undiagnosed cardiac disease.

Chest examination starts with looking at the rate and work of breathing and comparing the respiratory rate to normal ranges. Evidence of previous surgery includes a sternotomy scar or less visible thoracotomy scar. The pattern of breathing may provide clues to the diagnosis. Increased work of breathing and grunting suggest left-sided obstructive lesions or respiratory illness. Effortless tachypnoea may be found with cyanotic heart disease (see Chapter 1.1).

Pay particular attention to palpation of the peripheral pulses. Assess the rate, rhythm and character of the pulse. Compare the resting pulse rate to normal ranges for age (see Chapter 1.1). Variation of the heart rate with respiration (sinus arrhythmia) in children is more marked than in adults. Bounding pulses are often found in febrile children without heart disease but may be associated with PDA or a systemic-pulmonary shunt. Reduced volume or delay of the

femoral pulses compared with the right brachial pulse suggests coarctation of the aorta. Diffusely small pulses are associated with low-output cardiac failure or shock.

Blood pressure is a routine part of the cardiovascular examination in children. Blood pressure should be checked in the arm and at least one lower limb in neonates or if coarctation is suspected in the older child.

Locate and palpate the apex beat. Examine the precordium for parasternal heaves and thrills.

When auscultating the heart, listen carefully to the second heart sound. In children, splitting is usually only audible during inspiration at the upper left border of the sternum. Fixed splitting (the absence of variation between inspiration and expiration) occurs in ASDs. Third heart sounds are heard in 20% of normal children. Listen for abnormal clicks in early systole (indicating aortic and pulmonary stenosis).

Assess murmurs with regard to their:

- timing (systolic, diastolic or continuous)
- location – the point of maximum intensity
- loudness – increasing loudness from grades 1 to 6
- character – ejection, pansystolic, early/mid/late diastolic
- radiation – audible in areas away from precordium.

Complete examination with auscultation of the lungs noting air entry and presence or absence of crackles.

Palpate the abdomen noting liver span, edge and presence of pulsation.

At the completion of the cardiovascular examination, one should determine whether the child is cyanosed or not and whether the child has cardiac failure. The pulses, blood pressure and precordial findings assist with the assessment of the type of cardiac problem, even if a specific diagnosis is not possible. A chest X-ray and electrocardiograph (ECG) may assist.

Chest X-ray

The chest X-ray can provide information about cardiac size and shape, pulmonary blood flow and pulmonary oedema. Key features to assess on the chest X-ray are:

- cardiac position (dextrocardia, situs inversus)
- cardiothoracic ratio (usually less than 50% but up to 60% in AP films in neonates)
- cardiac contour. In right ventricular enlargement, the apex is upturned. The apex points towards the diaphragm in left ventricular enlargement. A prominent pulmonary artery occurs in conditions with *left-to-right shunting.*
- mediastinal contour. A narrow mediastinum is associated with transposition of the great arteries.
- lung fields. The distribution and prominence of the pulmonary vasculature aid in diagnosis. Obstructed lesions cause oligaemic lung fields, e.g. pulmonary atresia, tricuspid atresia, Ebstein's anomaly, tetralogy of Fallot and critical pulmonary stenosis. In acyanotic heart disease with left-to-right shunting there is increased pulmonary vascularity.

Electrocardiography

A systematic approach is vital in interpreting the paediatric ECG.

ECG criteria are age dependent. Most age-related changes are related to changes of right-to-left ventricular muscle mass. The right ventricle is larger at birth, but by age 6 months the ratio is 2:1 (left:right). ECG changes with age include decreasing rate, increased interval durations, changes in R/S ratio in precordial leads, and changes in T wave axis.

The rate, rhythm, and axis should be assessed. Normal QRS axis varies with age. Systematically look at the P waves, PR interval, QRS complexes, ST interval and T waves.

Atrial enlargement

- Right atrial enlargement – peaked P wave with height of >2.5 mm (Fig. 5.1.1).
- Left atrial enlargement – P wave >0.08 s, may also be plateau or notched (Fig. 5.1.2).

Ventricular enlargement

Right ventricular hypertrophy (Fig. 5.1.3):

- R greater than S in V1 after 1 year
- T wave upright in V1 after 1 week
- SV6 greater than 15 mm at 1 week, 10 mm at 6 months, 5 mm at 1 year
- Right axis deviation for the patient's age
- Abnormal R/S ratio in favour of the right ventricle.

Left ventricular hypertrophy (Fig. 5.1.4).

- SV1 + rV6 greater than 30 mm to 1 year
- SV1 + rV6 greater than 40 mm after 1 year
- Left axis deviation for the patient's age
- Abnormal R/S ratio in favour of the left ventricle.

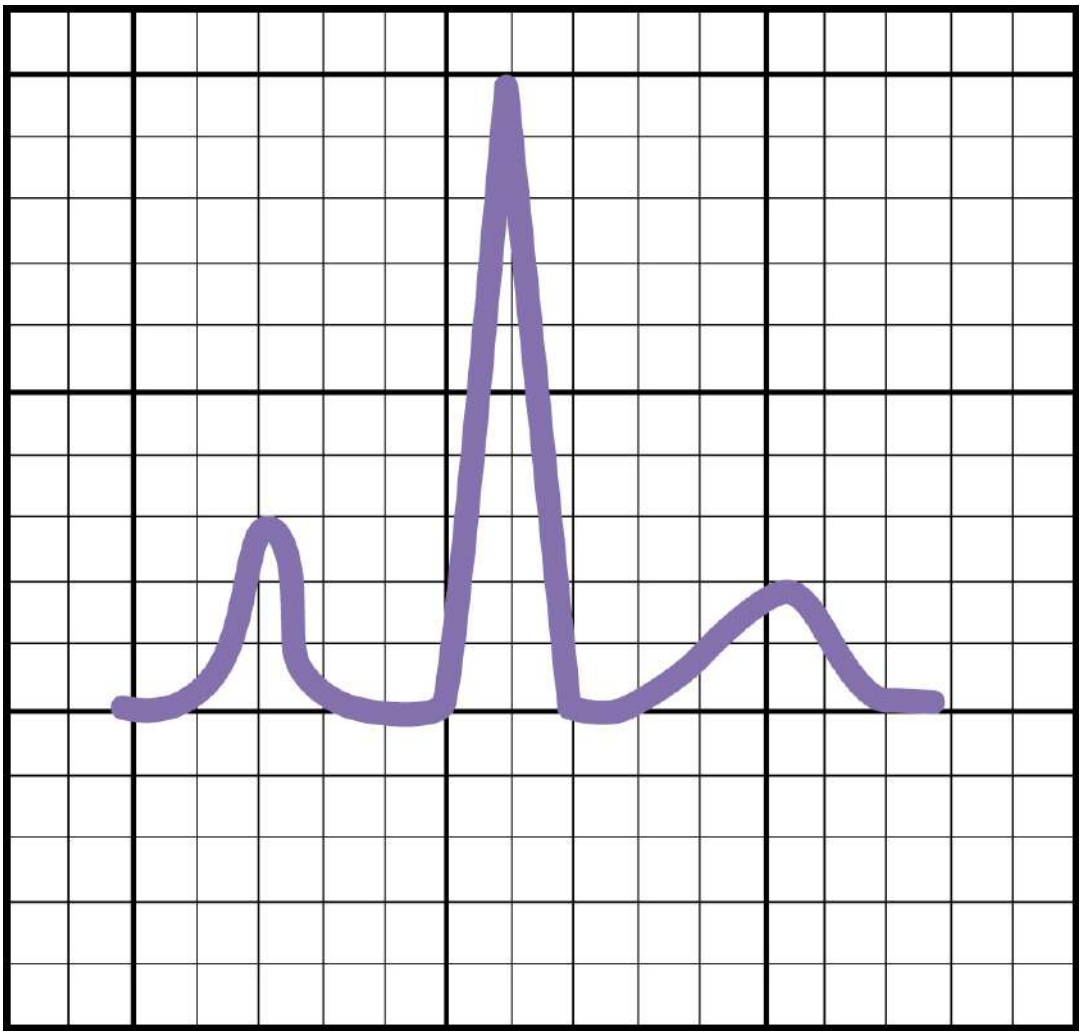
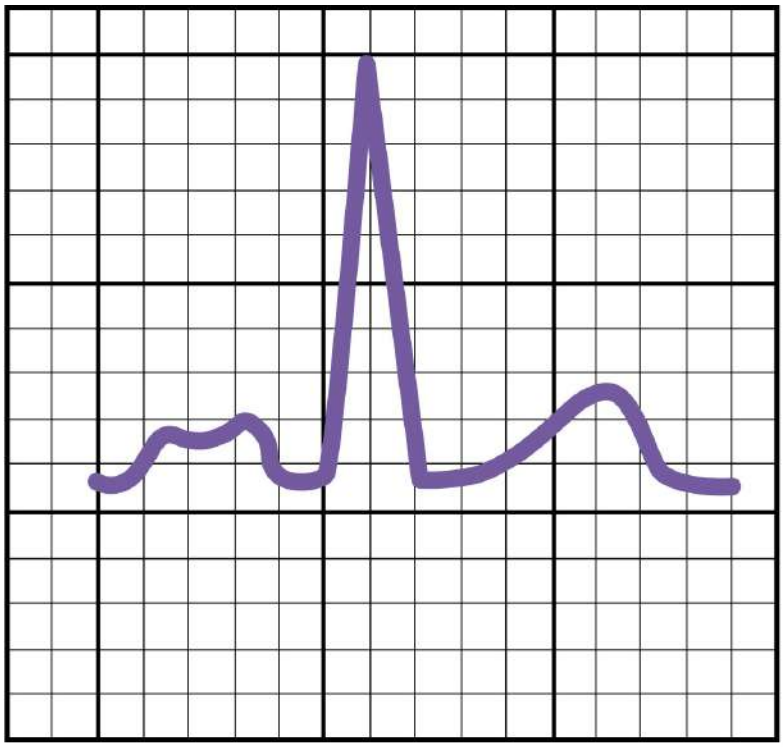


FIG. 5.1.1 Right atrial abnormality. Tall narrow P waves may indicate right atrial abnormality or overload (formerly referred to as *P pulmonale* pattern). From Goldberger: Clinical Electrocardiography: A Simplified Approach, 7th ed. 2006. Copyright © Mosby.



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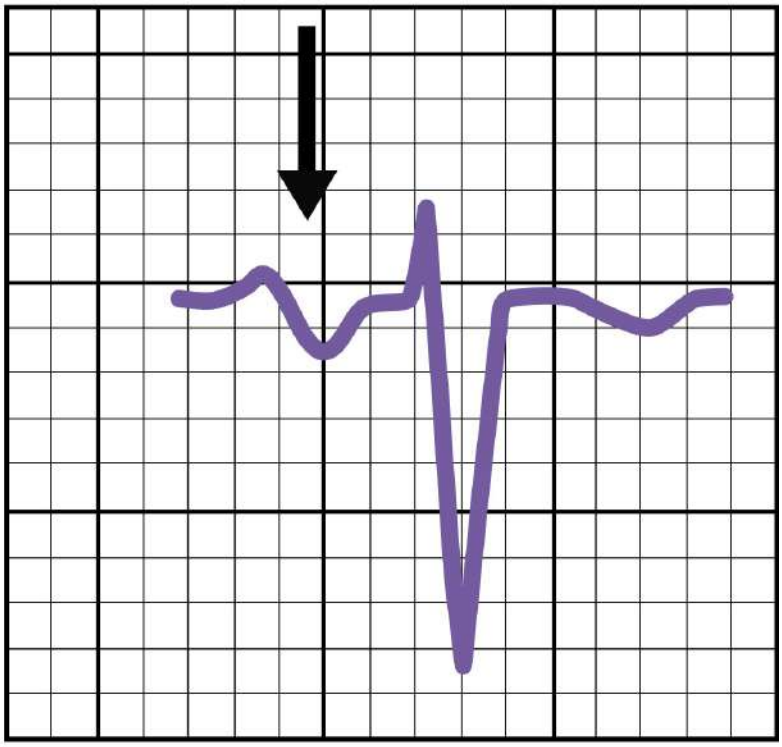

B

FIG. 5.1.2 Left atrial abnormality. Left atrial abnormality/enlargement may produce the following: (A) wide, sometimes notched P waves in one or more limb leads (formerly referred to as *P mitrale* pattern); and/or (B) wide biphasic P waves in lead V_1. From Goldberger: Clinical Electrocardiography: A Simplified Approach, 7th ed. 2006. Copyright © Mosby.

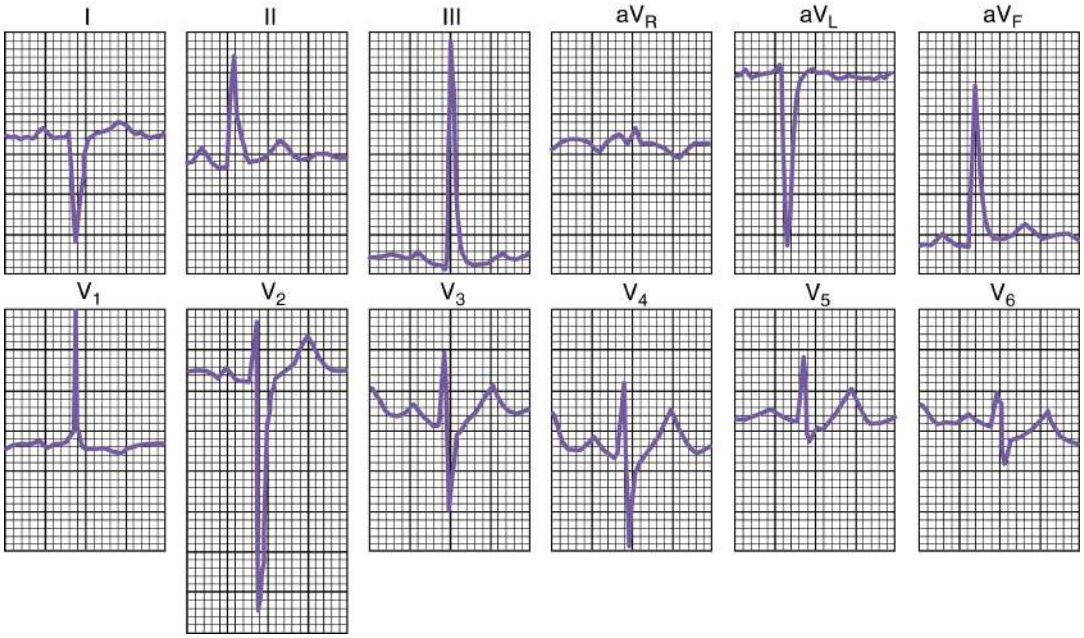

FIG. 5.1.3 Right ventricular hypertrophy. A tall R wave with an inverted T wave caused by right ventricular overload is seen in lead V_1 from a patient with tetralogy of Fallot. Marked right axis deviation is also present. (The R wave in lead III is taller than the R wave in lead II.) From Goldberger: Clinical Electrocardiography: A Simplified Approach, 7th ed. 2006. Copyright © Mosby.

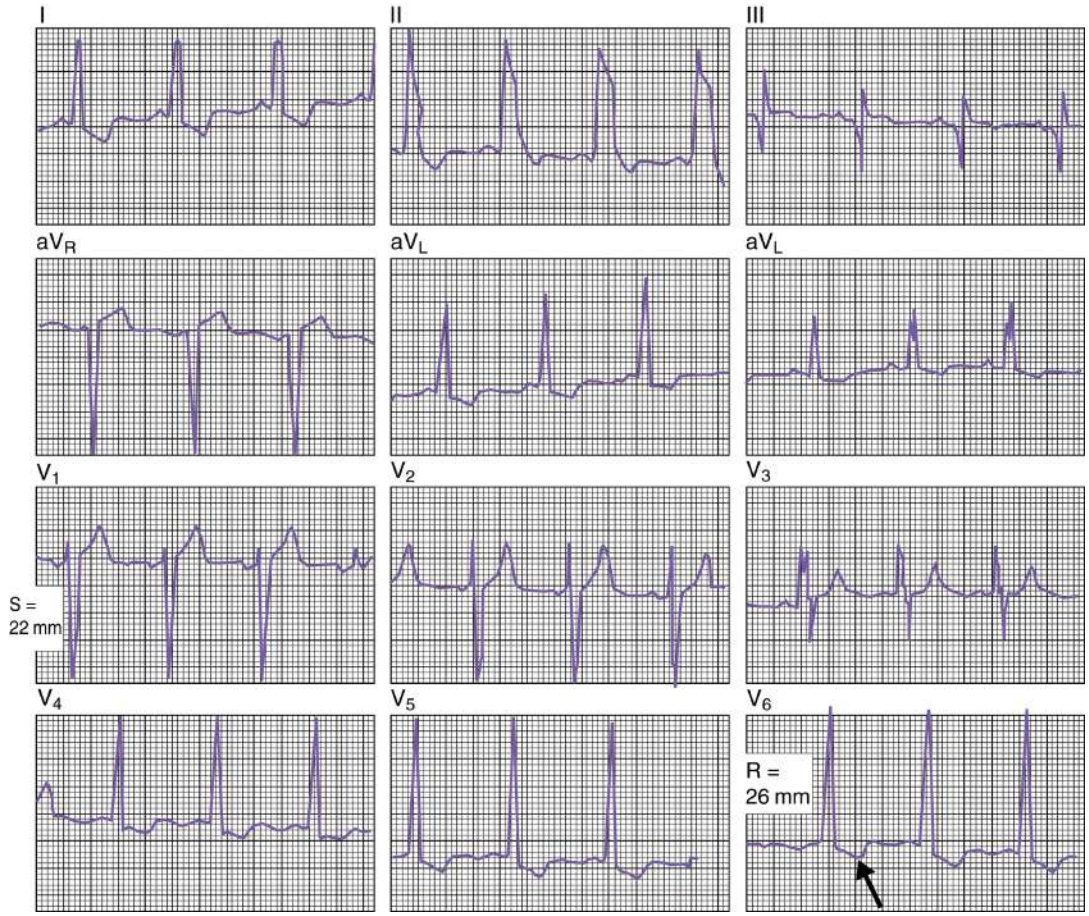

FIG. 5.1.4 Left ventricular hypertrophy. Tall voltages are seen in the chest leads and lead aV_L (R = 17 mm). A repolarisation (ST-T) abnormality (*arrow*), formerly referred to as a 'strain' pattern, is also present in these leads. In addition, enlargement of the left atrium is indicated by a biphasic P wave in lead V_1 and a broad, notched P wave in lead II. From Goldberger: Clinical Electrocardiography: A Simplified Approach, 7th ed. 2006. Copyright © Mosby.

Table 5.1.1

Characteristics of innocent paediatric cardiac murmurs

Murmur	Location	Intensity	Quality	Accentuated with	Diminishes with
Vibratory (still)	Midway between the apex and the lower left sternal border	Grade 1?2 systolic ejection	Vibratory, musical, medium frequency	Exercise, fever, supine position, agitation	Upright position, inspiration, neck extension
Pulmonary flow murmur	Second intercostal space, left sternal edge	Grade 1–2 ejection, systolic	Soft, blowing, often similar in quality to an atrial septal defect murmur but has a **normal** 2nd heart sound	Supine position, exercise, fever, agitation	Disappears with valsalva
Carotid bruit	Above the clavicle, radiates to the neck	Grade 1–2 ejection, systolic	Medium frequency, much softer below the clavicle	Exercise	Hyperextension of the shoulders
Venous hum	Over the neck or at the sternoclavicular junction	Grade 1–2 continous murmur	High pitched, continuous murmur	Sitting position, turning patient's head away from the side of the murmur	Disappears in supine position or with compression of the neck veins

Table 5.1.2

Timing of pathological cardiac murmurs

Systolic	Diastolic	Continuous
VSD	Aortic regurgitation	PDA
ASD	Pulmonary regurgitation	Fistula e.g. coronary
Aortic stenosis	Mitral stenosis	AP window
Pulmonary stenosis		AP collaterals
Valvular regurgitation		

VSD, ventricular septal defect; ASD, atrial septal defect; PDA, patent ductus arteriosus; AP, aortopulmonary.

The child with an asymptomatic murmur

Asymptomatic murmurs are very common in children. They may be heard at some time in 30% to 50% of normal school-aged children. Innocent murmurs occur when there is normal or increased blood flow through a normal heart and vessels. Innocent murmurs are more obvious during febrile episodes. Recognising common innocent murmurs enables exclusion of organic heart disease and the need for unnecessary investigation and referral. Innocent murmurs with the exception of the venous hum share the characteristics of being systolic, short, soft and usually well localized. The most common innocent murmurs are:

1. vibratory murmur (Still's murmur)
2. pulmonary flow murmur
3. carotid bruit
4. venous hum (Table 5.1.1).

ECG and chest X-ray are poor screening tests for children with asymptomatic murmurs.

Clinical assessment by a paediatrician or cardiologist correctly identifies almost all murmurs as innocent or needing further investigation. Echocardiography is not necessary in children identified by a specialist as having an innocent murmur.

Pathological murmurs

Pathological murmurs tend to be louder, harsher and longer than innocent

murmurs. They do not vary with respiration. Most diastolic or continuous murmurs are pathological with the exception of the venous hum (Table 5.1.2).

Disposition

Referral for specialist consultation is indicated for a child with a murmur who has:

- symptoms that may indicate cardiac disease (e.g. breathlessness, cyanosis, chest pain)
- abnormalities of the heart sounds (e.g. fixed splitting of the second heart sound)
- a murmur that cannot be confidently identified as innocent
- a murmur with an associated thrill (grade 4 intensity or greater).

Further reading

Park M.K. *Pediatric Cardiology for Practitioners*. 5th ed. Philadelphia: Mosby; 2008.

5.2

Chest pain

John A. Cheek

ESSENTIALS

1 Chest pain is a common reason for children to present to the emergency department.

2 Serious underlying pathology is rare.

3 Most children can be discharged after a history and examination, sometimes supplemented by an ECG or chest X-ray. Other investigations are rarely required.

Introduction

Chest pain is a frequent reason for children to present to emergency departments (EDs), although unlike adult patients they rarely have serious underlying organic pathology.[1,2] Retrospective studies have demonstrated a cardiac cause for chest pain of between 0% and 5% in children in a variety of settings including EDs.[2] Awareness in the community for chest pain being a sinister symptom is well established; parents and children will often seek reassurance. Even in the absence of a sinister cause it can have a significant impact on children's lives; one-third of children are woken from sleep, one-third miss school, and up to 45% complain of pain for more than 6 months.[3]

There are, however, a small number of infants, children and adolescents who do present either in extremis or with serious pathology, and as such a systematic approach for all those presenting with chest pain is required. Age is a significant factor in the etiology of chest pain; younger children are more likely to have an organic, cardiorespiratory cause for pain, whereas older children and adolescents

are more likely to have a psychological cause. These should be regarded as a diagnosis of exclusion, and consideration of a much broader differential (Box 5.2.1) should be routine.

Immediate approach

Children will rarely present to the ED with undifferentiated cardiovascular collapse or impending arrest. When this occurs, other causes (such as sepsis) are more common; however, a preceding history of chest pain should prompt consideration of differentials such as pneumothorax, arrhythmia, cardiomyopathy or myocarditis.

Box 5.2.1 Differential diagnoses of chest pain in children

Cardiac causes

- Coronary artery related:
    - Ischaemia/infarction
    - Coronary arteritis (Kawasaki disease)
    - Anomalous origin of coronary arteries
    - Coronary artery vasospasm
- Arrhythmias:
    - Supraventricular tachycardia
    - Ventricular tachycardia
- Structural abnormalities:
    - Hypertrophic cardiomyopathy
    - Dilated cardiomyopathy
    - Arrythmogenic right ventricular dysplasia
    - Left ventricular outflow tract obstruction
    - Pulmonary stenosis
- Infection/inflammation:
    - Pericarditis
    - Myocarditis

Non-cardiac causes

- Musculoskeletal disorders:
    - Costochondritis*
    - Trauma, muscle sprain*
    - Scoliosis
    - Precordial catch*
    - Idiopathic*
- Respiratory disorders:
    - Cough*
    - Asthma*
    - Pneumonia/respiratory infection with or without cough*
    - Pneumothorax, pneumomediastinum
    - Pulmonary embolism
    - Pleural effusion
- Psychological causes:
    - Anxiety*
- Other:
    - Gastro-oesophageal reflux, gastritis*
    - Pancreatitis, biliary disease
    - Oesophageal foreign body
    - Shingles
    - Dissecting aortic aneurysm (e.g. Marfans)
    - Sickle cell crisis
    - Lymphoma

* Common causes.

General approach (Table 5.2.1)

Most children do not need an urgent approach. A thorough history and examination, sometimes supplemented by some simple tests (most often an electrocardiogram or chest X-ray) will enable sinister causes to be excluded. There are some aspects of the history and examination which can be useful to indicate causes more likely in paediatrics.

History

Several aspects of history can prompt consideration of a more serious diagnosis (Table 5.2.1). Children with a recent, sudden onset of pain are more likely to have a pathological cause: in an older child, a pneumothorax, pulmonary embolism or arrhythmia; in a younger child also consider ingested oesophageal foreign body – beware button battery ingestion. Precipitation of pain during activity, particularly exercise, is concerning for an arrhythmia or structural lesion that obstructs left ventricular outflow during an attempted increase in cardiac output. Hypertrophic cardiomyopathy is more common in children with a family history of sudden unexpected death. Duration and radiation of pain should be elicited. Association with syncope, near syncope or palpitations are concerning for arrhythmias, structural lesions and myocarditis. Patients with myocarditis or pericarditis often have prodromal viral symptoms, but this is not universal. Unfortunately, concerning historical features do occur far more often than the significant diagnoses they are associated with – in one recent study of cardiology outpatients (none of whom died over the 10 years of the study because of a cardiac condition), 33% of children complained of exertional chest pain and 22% of palpitations.[4]

In adolescents, a history of recent stressors and social issues can be a pointer to a psychological cause of pain, and younger children can present with cardiac-sounding symptoms after close family members have had dramatic cardiac events. Sudden short pains (often left sided) experienced in healthy adolescents are typical of precordial catch syndrome.

Physical examination

A structured physical exam focused on the cardiorespiratory systems is essential; several specific pointers to etiology can be found for paediatric conditions. Don’t forget that chest pain can be caused by gastrointestinal disorders and systemic disease, so do not neglect these in your examination.

Fever is an important sign; its presence suggests an infective process, such as pneumonia, myocarditis or pericarditis. A cardiac and a respiratory exam should be undertaken. Abnormal cardiac findings – a murmur, rub or muffled heart sounds – may point to specific causes. A murmur that becomes louder during a Valsalva maneuver is a classic sign of hypertrophic cardiomyopathy, although this is not a sensitive finding. Chest palpation can elicit a musculoskeletal cause;

tenderness at the costochondral junction suggests costochondritis. Subcutaneous emphysema at the shoulders and neck can occur in both pneumomediastinum and pneumothorax. Evidence of dyspnea (either resting or with exertion) associated with chest pain is indicative of a more serious cause, particularly pneumonia, myocarditis and cardiomyopathy.

Table 5.2.1

Risk factors for serious underlying pathology presenting as chest pain[5]

Risk factor	Pathology to consider
Prior cardiac disease	Myocardial ischaemia, arrhythmia, pericarditis, pericardial effusion
Major chest trauma	Pneumothorax, haemothorax, cardiac or pulmonary contusion, mediastinal disruption
Sickle cell disease	Acute chest syndrome
Chronic respiratory disease	Pneumothorax
Chronic renal disease	Myocardial ischaemia
Kawasaki disease	Coronary artery aneurysm, myocardial ischaemia, arrhythmia
Family history of sudden death	Prolonged QT syndrome, hypertrophic cardiomyopathy
Hypercoagulable states (clotting disorders, cancer, connective tissue diseases, oral contraceptive use, prolonged immobility, central venous catheters, strong family history of thromboembolic disease)	Pulmonary embolism
Familial hyperlipidaemia syndromes	Myocardial ischaemia
Connective tissue disorders	Pericarditis, pericardial effusion, aortic dissection

Chest pain [Internet]. http://www.rch.org.au/clinicalguide/guideline_index/Chest_pain/.

Investigations

Most children require no investigations for chest pain after a history and examination. If a cardiac cause is being considered, an ECG can help reveal arrhythmias, pre-excitation, ischaemia and some secondary characteristics of structural disease (such as hypertrophy with some congenital lesions or small voltages in myocarditis or with cardiac effusions). A chest X-ray can reveal lung

pathology and assist in assessing for gross cardiomegaly.

Cardiac markers such as troponin are rarely useful in most children presenting with chest pain. However, these tests should be considered if pathology affecting the myocardium is being contemplated, such as myocarditis or ischaemia (e.g. anomalous left coronary artery arising from the pulmonary artery in neonates/infants, ischaemic heart disease in older teenagers with risk factors).

Other investigations, such as an echocardiogram for structural lesions, a holter monitor for arrhythmias, or an exercise ECG for QT prolongation can occasionally be useful. This is usually done after consultation with a paediatric cardiologist dependent on local referral patterns.

Summary

Most children in the ED with chest pain have a benign cause and after consideration of more sinister differentials can be discharged home with GP follow-up if required. A small number of children will need the involvement of a paediatric cardiologist.

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5.3

Syncope

Kim Yates, and Sarah Jamison

ESSENTIALS

1 The most common cause of syncope in children is vasovagal.
2 A careful and detailed history will usually enable vasovagal syncope to be established with confidence.
3 The main differential diagnoses of syncope in childhood include cardiovascular causes, seizures, migraines, drugs, hypoglycaemia, and psychogenic events.
4 A 12-lead ECG should be done for all children at the initial presentation with syncope.
5 Any child in whom a cardiac cause of syncope is either suspected or diagnosed should be referred to a cardiologist.

Introduction

Syncope is defined as an abrupt and transient loss of consciousness and postural tone due to cerebral hypoperfusion, followed by rapid complete recovery.

In the first two decades of life 15% to 35% of children experience an episode of syncope, with peak incidence amongst toddlers and adolescents. Syncope accounts for 1% to 3% of emergency department (ED) visits by children. Most causes of paediatric syncope are benign; however, it is important to identify children at risk of the rarer life-threatening causes.

Aetiology

Box 5.3.1 shows causes of syncope in children ranked by incidence. The differential diagnosis of pediatric syncope is wide; however, in childhood and adolescence the major cause of syncope is transient autonomic dysfunction.

In toddlers such episodes usually manifest as either blue breath-holding spells or reflex anoxic seizures (pale breathing-holding episode). The mechanism for the cyanosis in blue breath-holding spells is poorly understood but often follows a prolonged period of crying. The precipitant for reflex anoxic seizures may be a noxious stimulus such as a fright or a painful stimulus causing reflex asystole, which leads to an anoxic seizure.

In older children and in adolescents such episodes most commonly present as episodes of vasovagal syncope. A combination of hypotension and profound bradycardia or either bradycardia or hypotension alone leads to cerebral hypoxia, but the mechanisms are not completely understood. Other terms used to describe these episodes include neurocardiogenic syncope, vasodepressor syncope or neurally mediated syncope.

It should be noted that situational syncope (syncope that occurs during micturition, swallowing cold liquids, defecation or coughing) and carotid sinus sensitivity are rare in the paediatric population.

Typical presentations

Vasovagal syncope

In vasovagal syncope, the episode typically occurs whilst standing or sitting upright. There may or may not be a stressful antecedent event (this occurs less commonly in frequent recurrent vasovagal syncope). There is a prodrome of nausea, dizziness, visual disturbance and a sensation of warmth, followed by a period of loss of tone and consciousness. Witnesses will describe marked pallor. Seizure activity is unusual, but brief tonic–clonic activity or stiffening is possible, particularly if the patient fails to fall to a recumbent position. Urinary incontinence may also occur. Recovery to a normal level of consciousness is usually prompt once in the supine position. The child may have a headache or be fatigued for minutes to hours after the event.

Breath-holding spells and reflex anoxic seizures

Blue breath-holding spells are usually associated with a tantrum or prolonged episode of crying after which the child has a prolonged forced expiration and

apnoea and becomes cyanosed. This may be followed by a brief period of loss of consciousness, with a rapid recovery to full normal activity. They occur in children between the ages of 1 and 5 years, with a peak incidence at the age of 2 years. Reflex anoxic seizure or pale breath-holding episodes occur when an infant is suddenly startled or has a painful injury. The infant may give one or two cries, quiets and becomes pale, then abruptly loses consciousness. Tonic–clonic movements may occur. Episodes usually last less than 1 minute and are immediately followed by normal consciousness and posture. There is an association between iron deficiency and breath-holding spells, so when risk factors such as dietary restrictions or cow's milk intake of 400–500 mL per day are present, further investigation or empiric treatment can be considered in children with recurrent episodes. Although alarming, these episodes have no long-term sequelae and cease for most children by 6 years of age, although 10% to 20% may have vasovagal syncope in later life.

Box 5.3.1 Common causes of childhood syncope

Cause (incidence)
Vasovagal syncope (64–73%)
Breath-holding spell (6.4%)
- Reflex anoxic seizures

Cardiac (2.9–4.8%)
- Primary electrical disturbances: long or short QT syndromes, Wolff–Parkinson–White, ventricular tachycardia, Brugada syndrome, drug induced, sinus node dysfunction, atrioventricular blocks
- Structural heart disease: cardiomyopathies, coronary artery anomalies, aortic stenosis or other valve lesions, pulmonary hypertension, myocarditis, congenital heart disease, arrhythmogenic right ventricular dysplasia, cardiac masses, aortic dissection, pulmonary embolus, other outflow obstructions

Neurologic (2.1–4.6%)
- Seizures
- Vascular events: subclavian steal, vertebrobasilar insufficiency
- Disrupted cerebrospinal fluid circulation: colloid cyst of 3rd ventricle, posterior fossa tumour

- Vertiginous drop attack
- Basilar migraine
- Narcolepsy/cataplexy

Metabolic (0.8%)

- Bleeding, dehydration, hypoglycaemia, electrolyte disturbances, endocrine diseases, carbon monoxide poisoning

Psychiatric (2.2–2.3%)

- Conversion disorder, somatisation, anxiety, hyperventilation, Munchausen, malingering

Unknown or other (8.2–18.9%)

- Volume depletion, orthostatic hypotension, pregnancy related, unknown

Cardiac syncope

Cardiac syncope should be suspected in a patient with a history of congenital heart disease or with a family history of sudden unexplained death. Cardiac causes should be suspected if episodes occur with no warning, with associated chest pain, during exercise, whilst sitting or supine, or in association with palpitations (though palpitations are frequently described by individuals with vasovagal syncope and with hyperventilation).

Long QT syndrome is an ECG diagnosis that is associated with episodes of syncope or seizures caused by episodes of paroxysmal ventricular tachycardia (often *torsades de pointes*). It may result in sudden death. Syncopal episodes in patients with this diagnosis may be precipitated by exercise or a startle or may be spontaneous. The condition may be congenital or acquired. The ECG in sinus rhythm reveals a prolonged QT interval. The QT prolongation may be minimal, and a high index of suspicion is needed to make the diagnosis.

Hypovolaemic states

There will usually be a history suggestive of fluid or blood loss, and obvious signs of shock may be present. Orthostatic hypotension and tachycardia may be the only positive clinical signs. These tend to occur immediately, as distinct from the changes seen in vasovagal syncope, which occur after more prolonged orthostatic stress.

Seizures

It may be difficult to differentiate seizures from vasovagal episodes, as they may both be associated with brief convulsions as well as a loss of consciousness, although in seizures motor activity usually occurs before or with the collapse. A history of significant post-event disorientation is helpful in differentiating seizures from other causes of syncope. Seizures are also more likely to be associated with cyanosis, tongue biting, and a more prolonged period of loss of consciousness.

Hyperventilation and conversion syncope

Hyperventilation and conversion syncope are rare in children younger than 10 years. Hyperventilation often has a prodrome of apprehension and sighing respiration, then dyspnea, air hunger and chest tightness prior to loss of consciousness. Conversion syncope is a diagnosis to be made once all other possible causes have been excluded but often occurs in the presence of an audience, is not posture dependent, and associated injury is rare. The child may remember surrounding events, and there will be no neurologic, autonomic or cardiovascular changes.

Clinical

History

A careful and detailed history will usually enable the correct diagnosis of the most common cause of childhood syncope, vasovagal syncope, to be established with confidence. Any unusual features of the history should raise suspicion of an alternate diagnosis:

- The prodrome is most important. Nausea, sweating, light-headedness or visual changes (e.g. seeing spots, etc.) strongly suggest a vasovagal cause, although some children with cardiac syncope have also reported preceding light-headedness. An absence of a prodrome suggests a possible cardiac cause. Also enquire about perioral paraesthesia, carpopedal spasms, aura, palpitations, dyspnoea or chest pain.
- Circumstances of the event. Recent change of position, poor hydration or eating, a hot environment, phlebotomy, pain or emotional upset suggests

a vasovagal cause. Mid-exertional syncope suggests a cardiac cause. Post-exertional syncope or pre-syncope is typically benign but has been noted in children with cardiac pathology.

- Collateral history of event by a witness, if possible. Ask specifically about duration of loss of consciousness, timing of seizure activity, incontinence of urine, pallor or cyanosis, and post-ictal drowsiness or confusion.
- Relevant medical history including previous episodes of syncope, cardiac disease, epilepsy, diabetes, medication and drug use, dietary history and past sexual activity.
- Family history of sudden collapse or death (including sudden infant death syndrome, single vehicle car crashes or drowning in a competent swimmer) or cardiac disease may suggest cardiac syncope as a cause. Family history of epilepsy, syncope or metabolic disease also gives clues.

Examination

The physical examination, although rarely helpful, aims to seek potential diagnostic clues for the cause and identify any secondary trauma related to the syncopal event:

- Vital signs are helpful. The peripheral pulse rate, rhythm and character must be noted and the orthostatic blood pressure recorded (this is abnormal if there is a decrease in systolic blood pressure of more than 20 mmHg between measurements taken in the supine and sitting or standing position).

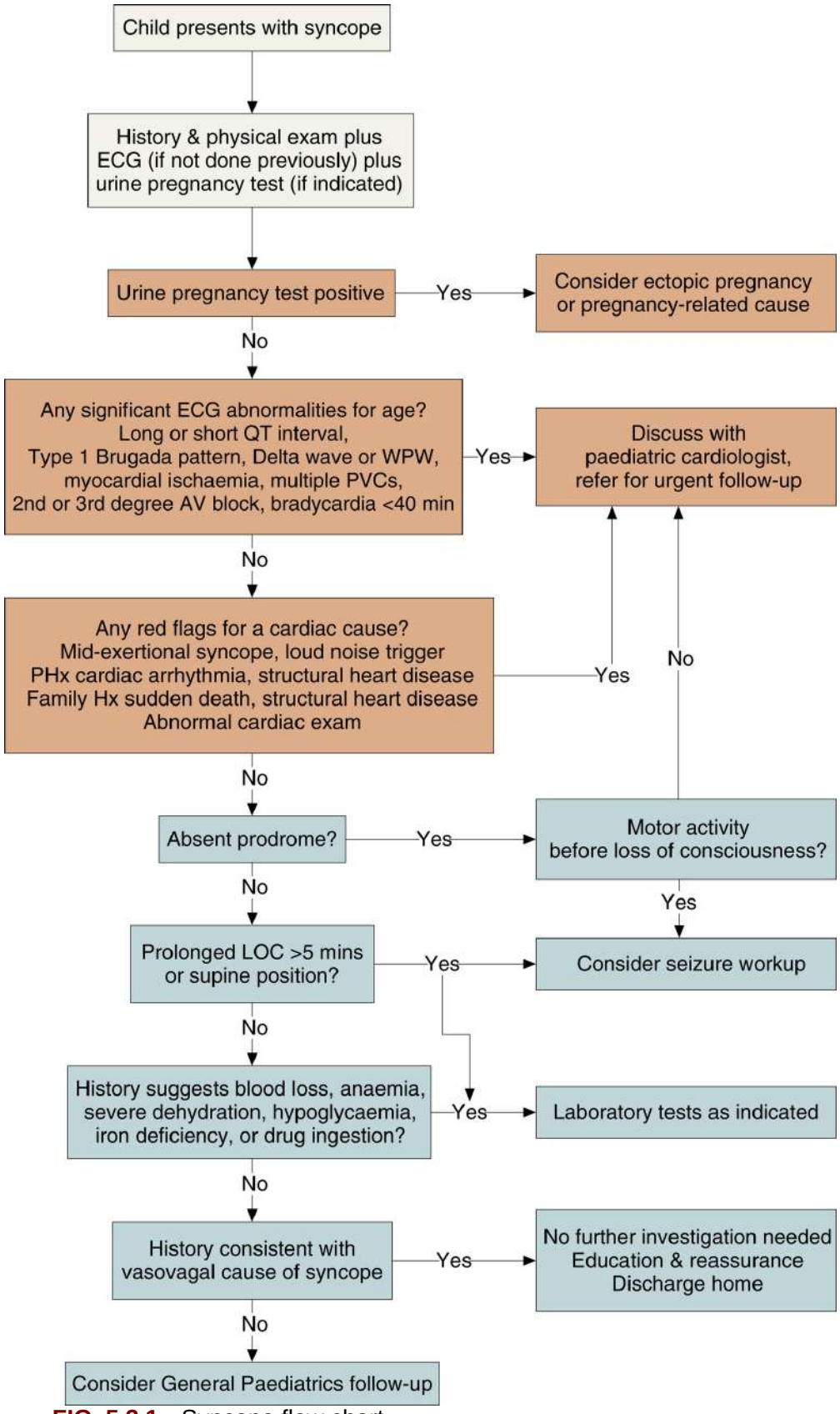

FIG. 5.3.1 Syncope flow chart.

ECG, electrocardiograph; WPW, Wolff–Parkinson–White; PVCs, premature ventricular contractions; AV, atrioventricular; PHx, past history; Hx, history; LOC, loss of consciousness.

- Cardiac and neurological exams aim to identify the rare occurrence of structural cardiac or neurological disease.
- Injuries that require treatment or further investigation should be identified.
- The history and the initial assessment should direct further examination of the child.

Investigations within the emergency department

Figure 5.3.1 shows an approach to working up a child who presents to the ED with syncope.

A 12-lead ECG is routinely done for all children at the initial presentation with syncope; however, abnormalities are rare. Paediatric ECGs change with age, so you may need to consult tables to determine whether an ECG is within normal limits for that age group. Long QT syndrome is the most common ion channelopathy so the QT interval should be calculated with Bazett's formula:

$$QTc = \frac{QT}{\sqrt{RR}}$$

where QTc is the corrected QT interval (normal <0.44 seconds), QT is the QT interval in seconds, and RR is the RR interval in seconds using lead II or V5.

A βHCG is indicated in fertile and sexually active adolescent females.

Other investigations are usually unnecessary and if done should be appropriate to the clinical history and examination findings for the child.

Further investigations performed in the ED may include:

- cardiac monitoring and pulse oximetry for any child in whom the history is suggestive of an arrhythmia or who remains unwell
- blood sugar level for a child with a history of diabetes, fasting, hunger, sweating, weakness, or drug ingestion or with a family history of metabolic disease

- blood haemoglobin concentration with a history suggestive of significant blood loss or anaemia
- iron studies in those children with recurrent breath-holding spells and a history suggesting iron deficiency
- serum electrolytes, urea and creatinine, and urinary specific gravity for severe dehydration
- a chest X-ray where a structural cardiac abnormality is suspected
- urine drug screen for patients with a history or examination that suggests intoxication with a specific substance; this is not a useful investigation when done routinely.

Any abnormality found in these investigations should then direct further investigation and referral.

Further investigations of syncope

Consider referral and further investigation for any child with an atypical history or abnormal examination or with severe or frequent vasovagal syncope (Box 5.3.2).

Any child in whom a cardiac cause of syncope is either suspected or diagnosed must be referred urgently to a cardiologist. After discussion with a paediatric cardiologist, further investigations may include an echocardiogram or cardiac monitoring such as a Holter prior to discharge from the ED. Some children may go on to have electrophysiological studies, exercise stress tests, cardiac imaging or an implantable loop device (e.g. Reveal).

Box 5.3.2 Indications for referral and further investigation of a child with syncope

- Atypical history
- Abnormal cardiovascular or central nervous system examination
- Suspect cardiovascular cause
- Suspect seizure
- Recurrent and problematic vasovagal syncope

Where a history obtained suggests that the child has had a seizure, an EEG should be arranged in consultation with the neurologist or paediatrician to whom he/she is referred.

Head-upright tilt-table testing may be done in children with frequent, recurrent syncope and in those children in whom a cause for syncope is not certain. Protocols for the test vary, but the requirements are that the child has a period of supine rest before tilting and is then tilted at a defined angle for a period of time. The most common positive response seen is a combination of hypotension and bradycardia prior to syncope or near syncope. Other positive responses seen are either isolated hypotension or asystole prior to syncope.

Management of syncope within the emergency department

For most children the diagnosis of vasovagal syncope will be made, and the majority of these will be able to be discharged from the ED. These patients must be given advice regarding the precipitants and management of vasovagal syncope. Avoidance of usual precipitating events is important. Children can also be taught to recognise the typical prodrome of the event and to then attempt to prevent any loss of consciousness by sitting or lying with their feet elevated. Increasing dietary salt and fluid intake has been shown to reduce vasovagal syncope recurrences.

Carers of children with a diagnosis of either reflex anoxic seizures or blue breath-holding spells should be educated and reassured prior to discharge.

Children seen with hyperventilation syncope may require treatment by rebreathing. They should be encouraged to breathe slowly and regularly, with a paper bag held over their mouth and nose. Once their breathing is regular and their symptoms improved, they should be encouraged to be calm and to sit or lie down for a time. Both the children and their carers should be taught the technique prior to discharge.

Summary

The most common cause of childhood syncope is vasovagal syncope. Careful history and examination are essential in making the correct diagnosis. All children who present with their first episode of syncope should have a 12-lead ECG.

If there is an atypical history or if there are any abnormalities in the clinical examination the child must be investigated and referred appropriately.

For children in whom a diagnosis of vasovagal syncope is made, education and reassurance of both the child and his/her carers must be done prior to discharge from the ED.

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5.4

Cyanotic heart disease and tetralogy of Fallot spells

Robin Choong

ESSENTIALS

1 Cyanosis is dependent on haematocrit and oxygen saturation.

2 Cyanosis is caused by arterial oxygen desaturation or increased capillary oxygen extraction.

3 Pulse oximetry screening is used for early detection of cyanotic congenital heart disease in the newborn.

4 If a cardiac cause is suspected check the pulses, blood pressure, preductal and post ductal pulse oximetry, ECG, and chest X-ray. Consult with a neonatologist, paediatrician or cardiologist *early.*

5 Presentation with cyanosis early in life may be due to a duct-dependent lesion. A prostaglandin infusion should be considered.

Tetralogy spells

1 Cyanosis is variable and depends on the amount of pulmonary blood flow and right-to-left shunting.

2 Hypercyanotic spells treatment:

- ‘Knee–chest position’ or over parent’s shoulder with knees bent
- Supplemental oxygen
- Sedation: intravenous or subcutaneous morphine, 0.1 mg kg
- Intravascular volume expansion.

3 In prolonged cyanosis consider a vasoconstrictor.

4 For prevention of spells, propranolol (0.5–1 mg kg po qid).

5 A Blalock–Taussig shunt may be used for palliation before corrective surgery.

Introduction

Cyanosis is a bluish discolouration of skin and mucous membranes due to excessive concentration of reduced haemoglobin in the blood.[1] Cyanosis is evident when deoxygenated haemoglobin is greater than 3 g dL in the artery and/or exceeds 5 g dL in cutaneous vein or capillary.[2] Deoxygenated haemoglobin may occur either from arterial blood desaturation or increased oxygen extraction by peripheral tissue. Central cyanosis is produced as a result of arterial desaturation, i.e. aortic blood carrying deoxygenated haemoglobin. Isolated peripheral cyanosis may result from excessive deoxy-haemoglobin caused by extensive oxygen extraction.[1] Haemoglobin level affects the presence of cyanosis. Cyanosis is detected at a higher oxygen saturation in children with polycythaemia and is more difficult to detect in children with severe anaemia. Causes of cyanosis are listed in Box 5.4.1.

Cyanotic congenital heart disease

Cyanotic congenital heart disease (CHD) generally presents in the neonatal period (Table 5.4.1). In the newborn, mild cyanosis (mild hypoxia SaO_2 >85%) may be difficult to detect clinically. Other confounding factors may be acrocyanosis (a normal finding, which may last 72 hours), polycythaemia (giving the appearance of cyanosis) and dark skin (cyanosis is more difficult to detect). Cyanosis is better appreciated in natural light than in artificial light.

Arterial oxygen saturation should always be assessed with pulse oximetry when considering cyanosis in the newborn. It is an accurate, reliable, and non-invasive method for monitoring oxygen saturation in infants. In severe cyanosis with respiratory distress, both preductal and post ductal oxygen saturations should be monitored to detect the gradient across the ductus arteriosus by placing the pulse oximeter probes over the right hand and a lower extremity (never the left hand).[2]

The hyperoxia test was historically used to differentiate between cardiac and pulmonary aetiologies. It is contra-indicated in preterm infants and does not differentiate persistent pulmonary hypertension from cyanotic congenital heart

disease (CHD). With the wide availability of echocardiography, the hyperoxia test is rarely required and should be considered only after discussion with a paediatric cardiologist.[2]

Box 5.4.1 Causes of cyanosis[2–6]

Differential diagnosis

Arterial oxygen desaturation (central cyanosis [pO_2 <50 mmHg]):

1. Airway
    - Airway obstruction
2. Breathing
    - Lung disease
    - Central nervous system depression (e.g. coma)
    - Respiratory muscle weakness
    - Reduced respiratory drive
3. Circulation
    - Intracardiac right-to-left shunt (e.g. cyanotic congenital heart disease, pulmonary hypertension)
    - Intrapulmonary shunt (e.g. pulmonary atrioventricular fistula)

Increased capillary oxygen extraction (peripheral cyanosis)

1. Circulatory shock (e.g. sepsis)
2. Congestive heart failure
3. Environmental (e.g. cold)
4. Acrocyanosis of the newborn (autonomic – may last 72 hours)

Abnormal haemoglobin (not related to level of oxygenation)

1. Methaemoglobinaemia (pO_2 >50 mmHg but low O_2 saturation)

Clinical features[2,3,5,6]

A comprehensive cardiac and respiratory examination is essential in any

cyanosed child. The key features are:

- pulses – rate, rhythm, volume
- blood pressure – all four limbs if the pulse volume is abnormal
- precordial impulse – heaves, thrills
- auscultation – abnormalities of P_2, murmurs
- pre- and postductal oximetry (right arm versus left arm/legs) if there is differential cyanosis.

Investigations

Investigations are directed at likely causes, after history and clinical examination. They may include arterial blood gas, serum electrolytes, glucose levels, full blood count, haematocrit and cultures.

Table 5.4.1

Congenital cyanotic heart disease in the newborn (the 5-Ts mnemonic)

Lesion	Features
T-Transposition of the great arteries (TGA)	Switch of the two outflow tracts (two parallel circulations) Most common is D-TGA (aorta anterior and to the right) No. 1 early presenting cyanotic lesion Intracardiac shunt → blood mixes at the PFO ± VSD Life-threatening if no intracardiac shunt → urgent balloon atrial septostomy.
T-Tetrology of Fallot (TOF)	Four components: VSD Overriding aorta Pulmonary stenosis Right ventricular hypertrophy Main cause of cyanosis is pulmonary stenosis or RVOTO (right ventricular outflow tract obstruction).
T-Total anomalous pulmonary venous return (TAPVR)	Pulmonary veins drain into right atrium (instead of left atrium) via superior vena cava, inferior vena cava, or hepatic veins → cyanosis + pulmonary congestion TAPVR can be partial or total, depending on whether *all* four pulmonary veins drain into right side or not Generally associated with an ASD/PFO for intracardiac shunting.
T-Truncus arteriosus	A single semilunar valve with *single* outflow tract originating from both ventricles, instead of two vessels (aorta + pulmonary artery) + VSD → cyanosis + increased pulmonary blood flow.
T-Tricuspid atresia	All RA blood is shunted R → L to left atrium via ASD/PFO Most have a VSD Stenosis of the tricuspid, and Ebstein's anomaly = inferiorly displaced tricuspid valve causing atrialisation of part of the right ventricle, a *small* functional right ventricle and RVOTO.

Others	Pulmonary atresia ± VSD (No VSD → duct-dependent) L → R shunt with pulmonary oedema (consider large VSD or PDA) = less severe cyanosis than in conditions where R → L shunt but more respiratory distress Single ventricle physiology (hypoplastic left or right heart) Low cardiac output states.

PFO, patent foramen ovale; VSD, ventricular septal defect; ASD, Atrial septal defect; R, Right; L, Left.

A chest X-ray (CXR) can be very useful and should be examined carefully for heart size (normal cardiothoracic ratio in AP film is <0.6 in neonates), oligaemic lung field with decreased pulmonary blood flow, e.g. tetralogy of Fallot, or pulmonary venous congestion with an obstructive lesion, e.g. total anomalous pulmonary venous drainage (TAPVD).

Transthoracic echocardiography can provide detailed and accurate information on both cardiac anatomy and function. Cardiac catheterisation has an important selected role especially when the cyanotic lesion requires urgent balloon atrial septostomy.[7,8]

Management[2–6]

The presentation of cyanosis in an infant, or child with suspicion of cardiac disease, mandates urgent review by a neonatologist, paediatrician or paediatric cardiologist. Echocardiography may be required urgently. Early discussion with a neonatologist or cardiologist can help clarify possible diagnoses and initial management in the emergency department (ED) setting.

An important issue in the management of cyanosis in the newborn period is recognising a duct-dependent lesion. A duct-dependent lesion is one that results from the ductus arteriosus remaining patent so that blood flow is delivered to both the pulmonary and systemic circuits:

- Pulmonary atresia/critical pulmonary stenosis. The ductus shunts right to left to ensure adequate pulmonary blood flow.
- Coarctation of the aorta/critical aortic stenosis/interrupted aortic arch. The ductus shunts right to left to ensure adequate systemic blood flow.
- Transposition of the great arteries. Mixing is required usually at atrial, ductus and ventricular levels.

If a duct-dependent lesion is considered, consult a paediatric cardiologist or neonatologist before starting an intravenous infusion of prostaglandin E_1 (PGE_1

5–60 ng kg min). Important side effects are respiratory depression and fever (usually transient). Avoid PGE_1 when a small heart accompanies cyanosis and pulmonary oedema, because these findings suggest TAPVD with obstructed veins (PGE_1 will worsen pulmonary oedema).

Chronic cyanosis stimulates a reactive polycythaemia that increases the oxygen-carrying capacity. When haematocrit reaches 65% or more, a large increase in blood viscosity occurs.

Hyperviscosity and coagulopathy often occur and in patients with a right-to-left intracardiac shunt and may result in stroke and brain abscess.

Disposition

Hospital admission and paediatric cardiology/neonatalogy consultation.

Tetralogy Spells

Introduction[3–6, 9–12]

Hypercyanotic, hypoxic or cyanotic spells occur in young infants with tetralogy of Fallot. These occur most commonly under 6 months of age and may be precipitated by agitation from any cause. The cause is likely to be multifactorial, including a component of dynamic infundibular obstruction. The net result is an imbalance of pulmonary to systemic blood flow.

Tetralogy of Fallot consists of anterior deviation of the outlet septum leading to the characteristic morphological features of:

- subpulmonary infundibular stenosis ± pulmonary valve stenosis ± hypoplastic pulmonary arteries
- subaortic perimembranous ventricular septal defect (VSD)
- aortic root overriding the crest of septum and VSD
- secondary right ventricular hypertrophy.

Clinical features include:

- intense cyanosis
- hyperpnoea and acidosis
- quieter and shorter ejection murmur (compared to before the spell)
- irritability, lethargy, loss of consciousness, seizures and death.

Triggers include dehydration, exertion and emotional distress associated with crying (increases pulmonary vascular resistance and hence decreases pulmonary blood flow). The child may have a history of previous squatting. Spells are usually self-limiting though sequelae and include hypoxic-ischaemic encephalopathy and death. Spells are less common since the advent of early systemic-pulmonary shunts or early corrective surgery.

Investigations[3–6]

Investigations include laboratory testing and imaging as follows.

Laboratory

Blood gas analysis – usually shows acidosis with hypoxia.

ECG

The ECG in tetralogy of Fallot usually shows:

- right ventricular hypertrophy in the unipolar and standard leads
- right axis deviation (+120° to +150°)
- dominant R wave in the right and a dominant S in the left precordial leads
- right atrial hypertrophy
- normal PR interval and QRS duration
- tall, peaked T waves
- reversal of the RS ratio.

Chest X-ray

The classic *coeur en sabot* or boot-shaped cardiac silhouette is caused by the elevation of the apex due to right ventricular hypertrophy, combined with a concavity in the area of the main pulmonary artery. A right-sided aorta is present in 25% of tetralogy patients.

Echocardiography

An echocardiogram will accurately confirm tetralogy of Fallot but is not useful

in acute management of spells.

Treatment[3–6, 9–12]

1. If possible, the child should be in a quiet, calm environment. Being held in parents' arms often diminishes crying and helps pulmonary blood flow.
2. The knee–chest or squatting position is preferred as it increases after load, thus decreasing right-to-left shunting.
3. Supplemental high-flow oxygen (10–15 L min of 100% oxygen via mask) should be provided.
4. Continuous monitoring of ECG, pulse oximetry, and non-invasive BP.
5. Morphine (0.1–0.2 mg kg intravenously [IV] or intramuscularly [IM]) should be used to treat hyperpnoea and decrease systemic catecholamines and often aborts crying, which perpetuates the spell. Alternatives include midazolam, fentanyl, and ketamine.
6. Consider a fluid volume challenge (5–10 mL kg) to increase preload and reduce dynamic outflow obstruction.
7. After discussion with a cardiologist or intensivist consider propranolol or esmolol to block β-receptors in the infundibulum, thereby lessening right ventricle outflow obstruction or metaraminol or phenylephrine to increase the afterload, thereby decreasing right-to-left shunt.
8. Consider sodium bicarbonate (1–2 mmol kg IV) for correction of acidosis.
9. Check temperature and blood glucose. Correct hypoglycaemia and hypothermia.

Disposition

Hospital admission and paediatric cardiology consultation.

Failure to respond to the above treatment mandates intubation, ventilation and deep sedation or general anaesthesia and intensive care admission. Emergency surgery is rarely indicated.

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5.5

Heart failure

Robin Choong

ESSENTIALS

1 Congestive heart failure (CHF) is a clinical syndrome and may result from:
- excessive workload caused by increased pressure or volume, usually increased pulmonary blood flow, and/or
- normal workload faced by a damaged myocardium.

2 Clinical signs may include tachycardia, tachypnoea, increased work of breathing, sweatiness, cardiomegaly and hepatomegaly. In addition, infants may have failure to thrive, recurrent lower respiratory tract infections and respiratory distress.

3 In older children, new onset heart failure may be less overtly symptomatic. Malaise, decrease in the level of daily activity, abdominal pain, nausea, anorexia and weight loss may be present.

4 Current medications used in heart failure include loop diuretics, angiotensin-converting enzyme (ACE) inhibitors, β-blocker and aldosterone antagonist.

Definition

Heart failure in children (0–18 years of age) is the failure of the heart to supply blood to either systemic or pulmonary circulation at an appropriate flow rate or to receive venous return at an appropriate filling pressure, causing detrimental effects on the heart and circulation.[1] This chapter focuses on chronic heart failure

rather than acute heart failure/cardiogenic shock.

Causes of congestive heart failure

Most cases of congestive heart failure in childhood result from congenital heart defects:[1–3]

1. Left-to-right shunts with increased pulmonary blood flow, e.g. ventricular septal defect (VSD) – most common, atrioventricular septal defect (AVSD), patent ductus arteriosus (PDA).
2. Acute left heart obstruction, e.g. aortic stenosis, coarctation of the aorta, interrupted aortic arch, hypoplastic left heart syndrome.

A smaller number of cases may be from:

1. Primary myocardial dysfunction, e.g. myocarditis, cardiomyopathy, anomalous left coronary artery.
2. Other causes, e.g. inflammatory/infective (e.g. viral myocarditis, Kawasaki disease, pleuropericarditis, rheumatic fever); anaemia; metabolic/genetic; toxic; dysrhythmia (e.g. sustained supraventricular tachycardia [SVT], VT).

Clinical manifestations and investigations

Presentation[1–3]

Infants may present with problems related to feeding, such as sweating, tachypnoea, reduced volume of feeds leading to failure to thrive. Respiratory problems, such as cough, recurrent respiratory infections, tachypnoea and increased work of breathing, are prominent. Lower respiratory tract infection may have features in common with CHF. Cardiac failure should be considered in atypical, persistent or recurrent cases of lower respiratory tract infection, particularly in infants.

General features[1–3]

- Tachycardia – due to fixed stroke volume

- Tachypnoea – due to decreased compliance with increased lung water
- Gallop rhythm, weak pulse
- Failure to thrive – from decreased energy intake, increased energy expenditure
- Cardiomegaly – may be due to left heart obstruction with left-to-right shunt or left ventricular dysfunction
- Sweaty (cold sweat) – prominent with feeding
- Fatigue/lethargy
- Hepatomegaly – not specific to right heart failure unlike in adults, may occur with any aetiology.

Staging[4–9]

Part of defining heart failure is staging and defining a spectrum of severity. The New York Heart Association (NYHA) Heart Failure Classification is not applicable to most infants and children. As a consequence, various classifications have been developed for the paediatric population. These include the following:

- The Ross Heart Failure Classification (developed to provide a global assessment of heart failure severity in infants and then modified for use in older children)
- The modified Ross Classification (incorporates feeding difficulties, growth problems, and symptoms of exercise intolerance into a numeric score comparable with the adult NYHA classification)
- The New York University Pediatric Heart Failure Index for children and adolescents (Connolly et al.) which yields a weighted score based on physiological indicators and medical therapy
- Pediatric Disease Staging for Heart Failure.

Assessment

Investigations[1–3]

Routine

- Chest X-ray:

Cardiomegaly (in an AP film the normal cardiothoracic ratio is <0.6 in neonates). Pulmonary plethora is associated with left-to-right shunt. Pulmonary venous congestion is associated with left heart obstruction/left ventricular dysfunction. Radiologic features include reticulogranular pattern, fluid in lung fissures, pleural effusions and pulmonary oedema.

- ECG (12-lead):

  Assess rhythm, myocarditis (voltages), ischaemia (ST-T wave).

- Biochemical and laboratory investigations:

  Assess electrolytes, glucose, acid–base status, liver function tests, thyroid function, full blood count at initial presentation of heart failure and repeated as needed to assess ongoing clinical status.

  Biomarkers may have use in monitoring the effects of therapy. Natriuretic peptides (BNP or NT-proBNP) are helpful in distinguishing heart failure from other non-cardiac disease and recommended as a confirmatory test in the acute assessment of paediatric heart failure.

Specialised

- Echocardiography
- Cardiac catheterisation
- Others: angiography (CT and MRI), viral studies (blood, throat swab, faeces), chromosomal analysis, urine metabolic screen, myocardial tissue biopsy.

Referral

The diagnosis of cardiac failure in an infant or child mandates urgent review by a paediatrician or paediatric cardiologist.

Table 5.5.1

Haemodynamic assessment matrix for acute decompensated heart failure

Perfusion at rest	No congestion at rest (dry)	Congestion at rest (wet)*

Normal perfusion (warm)	Warm and dry (goal physiology) Compensated Therapy: optimise oral therapy	Warm and wet Congested Therapy: diuretics
Low perfusion (cold)**	Cold and dry Low flow state Therapy: inotropes, vasodilators	Cold and wet Decompensated Therapy: diuretics, vasodilators, inotropes

* Signs of congestion: hepatomegaly, tachypnoea, retractions, grunting, facial/dependent oedema (older children), orthopnoea (older children), ascites.

** Signs of low perfusion: hypotension or tachycardia with narrow pulse pressure, cool extremities, irritable or depressed consciousness, diaphoresis.

Adapted from: Hoffman TM. Chronic heart failure. *Pediatr Crit Care Med* 2016;**17**(Supplement 8):S119–23. In: Nohria A, Lewis E, Stevenson LW. Medical management of advanced heart failure. *JAMA* 2002;**287**(5):628–40.

Management[1–3, 10–14]

Current medications used in heart failure include loop diuretics, angiotensin-converting enzyme inhibitors, β-blockers and aldosterone antagonists. Most data regarding paediatric chronic heart failure medications are extrapolated from adult studies.

Treatment goals include the following:

1. Preload optimisation by reducing fluid overload (with diuretics) but avoiding intravascular, especially intra-arterial, volume depletion
2. Reducing afterload (with vasodilators) without jeopardising the coronary perfusion pressure
3. Optimising myocardial oxygen consumption and re-establishing myocardial synchrony (pacing)
4. Down-regulating the neurohormonal responses (ACE inhibitors, β-blockade, aldosterone antagonists)
5. Providing inotropic support (with enteral digoxin)
6. Providing time to establish endogenous and exogenous repair mechanisms.

Acute management

1. Resuscitate with ventilatory support if required
2. If desaturated consider supplemental oxygen. Caveat: large left-to-right shunt lesions (e.g. truncus arteriosus) may worsen with high FiO_2.

Consult early in these cases
3. Fluid restriction and diuretic therapy
4. Infants with left heart obstruction may present with cardiorespiratory failure when the ductus arteriosus closes. Prostaglandin E1 infusion may reopen the ductus and assist systemic perfusion
5. If low cardiac output syndrome with end-organ dysfunction, consider inotropic therapy as a rescue strategy. Phosphodiesterase inhibitors (milrinone) and catecholaminergic drugs (dobutamine, dopamine, and low-dose adrenaline have all demonstrated efficacy in children). Mechanical extracorporeal support device may be considered in extreme cases.

Diuretics

Furosemide is the most commonly used diuretic for acute and chronic heart failure in children. Diuretics are recommended as safe (despite a lack of evidence-based studies) and effective in children when administered acutely as a parenteral medication and chronically by the oral route:

1. Acutely: furosemide 1 mg kg dose intramuscular (IM) or intravenous (IV) (anticipate hypokalaemia)
2. Chronically: furosemide 1–3 mg kg day in divided doses (q 6–12 h), with spironolactone, 1 mg kg po q 12-hourly. Beware potassium-sparing effect of ACE inhibitors.

Angiotensin-converting enzyme inhibitors

Though limited paediatric data are available regarding the use of angiotensin-converting enzyme (ACE) inhibitors, they are the primary therapy in the management for heart failure particularly in heart failure due to large left-to-right shunts and heart failure caused by ventricular failure. Oral captopril (0.1–1 mg kg^{-1} q 8-hourly) or lisinopril (0.1 mg kg day, increasing to 0.2–0.4 mg kg day over 4–6 weeks) may be used.

β-blockers

β-blockers have a role in the treatment of moderate-to-severe left ventricular systolic dysfunction. Doses are extrapolated from adult data. Carvedilol

(nonselective β-adrenoreceptor antagonist with α-adrenergic blocking activity and antioxidative properties) is commonly used; others used include the cardioselective agents metoprolol and bisoprolol.

Digoxin

Though traditionally added to diuretic therapy, there is little paediatric evidence supporting digoxin use in children with anatomically normal hearts and systolic dysfunction. Therefore routine digoxin use in paediatric heart failure is no longer recommended.

Levosimendan

Levosimendan is an IV calcium sensitiser used in acutely decompensated severe congestive cardiac failure. It has inotropic, vasodilatory, lusitropic and cardioprotective properties. In adults with severe cardiac failure, improvement in 31-day survival compared with dobutamine has been shown.

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5.6

Congenital heart disease

David H.F. Buckley, and Michael Shepherd

ESSENTIALS

1 Congenital cardiac disease should be considered as a possible diagnosis in an infant or child with respiratory distress, cyanosis or shock.

2 The common congenital cardiac problems have characteristic clinical features that assist with clinical diagnosis and initial management.

3 Immediate management of congenital cardiac disease follows basic principles of resuscitation, BUT all these patients require consultation with paediatric cardiology and/or paediatric intensive care unit (PICU)/transport service.

4 Complications following surgery for congenital cardiac disease are a frequent cause of presentation and require some specific knowledge and skills to manage.

Introduction

Incidence

Approximately 1% of infants and children in developed countries are born with congenital heart disease (CHD); however, severity varies greatly. Approximately 50% of these present in the first year of life. Eight lesions account for 80% of all cases of congenital cardiac abnormalities: ventricular septal defect (VSD); patent ductus arteriosus (PDA); atrial septal defect (ASD); tetralogy of Fallot; pulmonary stenosis; coarctation of the aorta; aortic stenosis (AS); and

transposition of the great arteries.

Patients may present with:

- undiagnosed CHD
- a complication of CHD or its treatment
- known CHD with the development of another illness.

With increased survival there are now more older children and adults in the community with ‘repaired’ CHD. While these older patients seldom present a diagnostic challenge, management of their cardiac condition can be challenging, or the CHD may affect their presentation with an intercurrent illness.

Pathophysiology

Fetal circulation transports oxygenated blood from the placenta to the fetal circulation with minimal flow to the fetal lungs. This right-to-left shunt is achieved at several levels – via the foramen ovale (right atrium to left atrium) and via the ductus arteriosus (pulmonary artery to aortic arch).

At birth, pulmonary vascular resistance drops as the lungs inflate, and systemic vascular resistance rises with the cessation of placental flow. This causes functional closure of the foramen ovale (left atrial pressure greater than right) and blood flow from left to right through the ductus arteriosus. Oxygenated blood flow through the ductus arteriosus normally causes functional closure over first 24 hours, with complete anatomical closure over the first 2 to 3 weeks of life.

Undiagnosed congenital heart disease

Most severe CHD is now diagnosed antenatally, and babies are delivered in a tertiary centre where there is the expertise to deal with complex CHD. However, a significant minority are not identified antenatally, and a number of less complex conditions (e.g. VSD) are often not diagnosed antenatally. These babies may be healthy at birth and present after discharge with a variety of problems. There are some findings on examination that should make the clinician suspicious of CHD.

Findings that may indicate congenital heart

disease

Is the child cyanosed?

- This is not always easy to detect clinically, and oxygen saturation should always be measured with pulse oximetry.
- Be aware that preductal saturations (right upper limb) may be normal or significantly higher than other limbs if there is a patent ductus arteriosus (measure saturations in both the right upper limb and a lower limb).
- Central cyanosis with normal breathing or only mild respiratory distress suggests a cardiac problem rather than a primary respiratory problem.
- Cyanosis that persists despite supplemental oxygen suggests a cardiac cause. Supplemental oxygen will, however, usually cause a small increase in saturation in these children.

Does the heart sound normal?

- Is there a continuous murmur? Loud, widely radiating, pansystolic or continuous are features more suggestive of CHD.
- Is there an additional heart sound?

Are the femoral pulses palpable?

Is the chest radiograph normal?

- Is the heart shape abnormal?
- Is the pulmonary vascularity increased or decreased?

Is the electrocardiograph normal?

- Is the axis normal?
- Is there atrial or ventricular hypertrophy?

If there is any suspicion of CHD, the child should be discussed with a paediatric cardiologist and ideally undergo cardiac echocardiography.

Diagnostic approach

Fig. 5.6.1

Common presentations and initial management

Neonate/infant

A significant proportion of patients with severe CHD rely on the ductus arteriosus for communication between the systemic and pulmonary circulations. As the duct closes (usually in the first week of life) these children will present with severe cyanosis or shock. The duct may not shut in neonates who are premature or who are cyanotic from birth. Some non-obstructive lesions present later in the neonatal period with congestive heart failure:

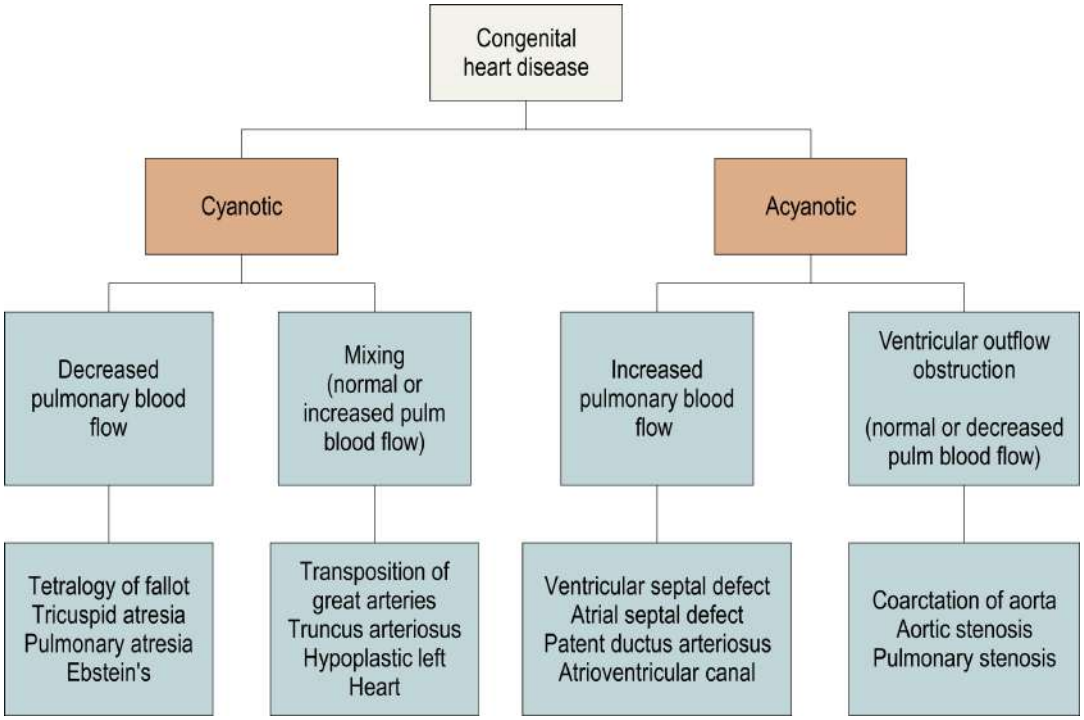

FIG 5.6.1 Schematic approach to common causes of common congenital heart diseases

Cyanosis:

- These patients either have decreased pulmonary blood flow or abnormal great vessel connections.
- They may have had undiagnosed cyanosis from birth or have become cyanotic as the ductus arteriosus closes over the first week of life.
- Commoner lesions are tetralogy of Fallot, transposition of the great arteries (TGA), pulmonary atresia, severe pulmonary stenosis, tricuspid atresia and Ebstein’s anomaly.

Treatment:

1. Oxygen usually has a small benefit but is not contra-indicated.
2. Prostaglandin E1 to reopen the ductus and maintain ductal patency (5–50 nanogm kg min). In some cases, this will bypass obstruction to pulmonary blood flow and in others it will allow for mixing of systemic and pulmonary blood. It may take 30 minutes or more before benefit is seen. Beware of apnoea and hypotension which are commoner in preterm neonates and if using higher doses.
3. Maximise end-organ oxygen delivery – fluid bolus to improve cardiac output, may need inotropes (adrenaline [epinephrine]), optimise haemoglobin (low threshold for transfusion)
4. Minimise oxygen demand – control fever, minimise work of breathing, may require ventilation
5. Always obtain cardiology advice if an arrhythmia is suspected especially in infants and small children.
6. Correction of acidosis if present
7. Ensure blood sugar adequate
8. Consider antibiotics – particularly if diagnostic uncertainty
9. Calcium if inonised calcium is <1.0
10. Urgent cardiology input for diagnosis and possibly to improve mixing via creation/enlargement of an ASD. Occasionally urgent cardiac surgery is required to create a shunt between the circulations or to correct TGA.

Shock:

- These patients usually have obstruction to systemic blood flow and a ductus dependent systemic circulation. They present when the ductus shuts, and they lose systemic blood flow beyond the obstruction, i.e. they retain circulation to the heart and brain but lose perfusion of the abdominal organs and lower limbs.
- Occasionally shock can be caused by a severe congenital cardiomyopathy or a severe arrhythmia.
- CHD is a commoner cause of shock in neonates than infection in the first week of life. Common lesions are coarctation of

the aorta, critical aortic stenosis, interrupted aortic arch, hypoplastic left heart syndrome, congenital cardiomyopathy, and congenital arrhythmia.

- Major aortic arch lesions can usually be excluded by palpating femoral pulses and performing four limb non-invasive blood pressures.
- If there is a VSD as well as obstructed systemic flow patients may have congestive heart failure as well as shock.

Treatment:

- Treatment centres upon restoring systemic blood flow by reopening the ductus arteriosus and improving cardiac output which may involve correction of acidosis, fluid bolus(es) and inotropes:

1. Oxygen
2. Positive intrathoracic pressure will help unload the left ventricle and can be provided noninvasively with continuous positive airway pressure (CPAP). A small amount of CPAP may be provided non-invasively with high flow nasal oxygenation.
3. Confirm sinus rhythm (rule out congenital arrhythmia).
4. Prostaglandin E1 – as above
5. Support of circulation with fluid bolus
6. If ongoing shock then start inotropes – adrenaline is recommended as the first line
7. Give broad-spectrum antibiotics if any uncertainty about the cause of the shock.
8. Early involvement of the paediatric cardiac service is essential to make a definitive diagnosis and to coordinate ongoing treatment.
9. These patients are at major risk of cardiac arrest if given drugs for intubation so obtain expert assistance/advice if intubation is required.
10. In older infants the duct may not reopen, and urgent surgery is required.

Congestive heart failure:

- Common symptoms are poor feeding, sweating, and increased work of breathing.
- Heart size is enlarged on chest X-ray and lung fields look plethoric. A large left atrium may be apparent in mitral stenosis/regurgitation. Heart sounds and murmurs may be difficult to detect at high hear rates but there may be a murmur or a 3rd heart sound audible.
- An enlarged liver is a very common and important finding.
- Common lesions are VSD, PDA, cardiomyopathy, congenital arrhythmia, congenital valvular disease (always consider a VSD or atrioventricular canal lesion in patients with trisomy 21):
    - Patients with a large VSD usually present at 4 to 12 weeks of age with congestive heart failure.
    - Patients with a PDA may present in a similar way though this is an uncommon condition beyond the neonatal period. PDA is common in preterm neonates so is actively looked for in these babies and treated but occasionally goes undiagnosed and presents later with congestive heart failure.
    - Severe cardiomyopathy often presents between 3 and 6 months of age. This may be post viral, although often no cause is identified. It carries a very poor prognosis.
    - Occasionally patients with congenital lesions causing high pulmonary venous pressures (occlusion of the pulmonary veins or mitral valve stenosis) present with congestive heart failure.
- Many of these children will already have had a diagnosis made and present with worsening symptoms due to a change in the underlying cardiac condition, or they develop an intercurrent illness.

Treatment:

1. Oxygen
2. Support ventilation as required
3. Diuresis with frusemide
4. Inotropes as required (commonly needed in cardiomyopathy

patients). An inodilator such as dobutamine or milrinone is a good choice, but it will not increase the blood pressure. Adrenaline should be used if blood pressure needs raising.

5. Vasodilators/beta-blockers – these are used chronically to improve long-term outcomes. Exercise extreme caution if considering giving these drugs to a patient with decompensated heart failure.
6. Never give a calcium channel antagonist for supraventricular tachycardia (SVT) in this setting.

Older child

Cyanosis:

- This is a less common presenting finding in older children but still occasionally occurs.
- The commonest cases are children with known CHD who have some progression of their disease or a complication of treatment, or they develop another problem which exacerbates their cardiac condition (see below).
- Patients may present with cyanosis that was missed at birth (d-transposition of the great arteries is the most common) or cyanosis that has developed since birth (missed PDA or VSD that has resulted in raised pulmonary artery pressures and shunting right to left) or cyanosis that is a complication of treatment (blocked systemic to pulmonary shunt).

Treatment:

1. ALWAYS give oxygen to a cyanosed child.
2. Check with parents/old notes re: diagnosis and previous treatment.
3. Tetralogy of Fallot not yet surgically remediated (Tet spells).
    a. Need to decrease pulmonary vascular resistance – calm, oxygen, morphine
    b. Increase preload – fluid bolus (20 mL kg initially)
    c. Increase systemic vascular resistance (SVR) – knee to chest/squatting, vasopressor may be

required (metaraminol or phenylephrine)

d. Beta-blockers can be effective; however, a small initial dose should be used in order to maintain SVR. Esmolol is a good choice as it is short acting and will often slow heart rate without dropping the SVR.

e. Obtain cardiology input to assist with managing these patients.

Shock:

- The commonest cause is cardiomyopathy/myocarditis.
- Congenital causes include myocardial ischaemia (abnormal coronary arteries), arrhythmia (primary or secondary), ventricular failure due to pressure overload. These patients often present with severe shock.
- Primary cardiac disease can be confused with sepsis on occasions and vice versa.
- A 12-lead elctrocradiogram (ECG) is very important to identify arrhythmia and ischaemia.

Treatment:

1. Diagnosis and treatment of tachyarrhythmias can be difficult and dangerous, and the opinion of a cardiologist should always be sought if considering a tachyarrhythmia as the cause of shock.
2. Acute treatment of low cardiac output involves the use of inotropes +/- vasodilators, and often these patients need intubating and ventilating.
3. Always obtain skilled assistance when intubating and ventilating, and be prepared for severe decompensation of the patient at induction.

Congestive heart failure:

- Most of these patients have a left-to-right shunt with excessive pulmonary blood flow. Some have poor myocardial function with right ventricle (RV) failure predominating. They often present with increased work of breathing and resulting poor feeding and failure to thrive.
- The commonest cause up until around 1 year of age is a large VSD, but beyond this cardiomyopathy/myocarditis is more

common.

- A number of these children are misdiagnosed as having a respiratory disease as they can present in a similar way. Often there is cardiomegaly that has been missed on the chest X-ray, and the liver is enlarged. Heart sounds and murmurs may be difficult to hear in an upset child with a rapid heart rate.

Treatment:

1. Oxygen
2. Support ventilation as required
3. Frusemide
4. Morphine may be used.
5. Inotropes if needed – adrenaline is a good choice at low dose (up to 0.05 mcg kg min), but above this it may cause unwanted tachycardia +/- arrhythmia.
6. Vasodilators – these are used more in the long term to lessen cardiac work. They should only be used acutely in a well-monitored area under the guidance of a paediatric cardiologist.

Complications/residua of congenital heart disease and its treatment

Congenital cardiac disease repair is often staged in more complex lesions; some of the more common repairs are the following:

- **Arterial switch** – Definitive repair for transposition (TGA), with aorta and coronary arteries switched to the left ventricle (LV) and pulmonary artery switched to the RV.
- **Modified Blalock–Taussig Shunt (MBTS)** – A modified BT shunt places a GorTex conduit from the right innominate artery to the right pulmonary artery. There are various other shunts that can be used to shunt blood from the systemic to the pulmonary circulation which are named depending upon where they connect to (e.g. Waterson, Potts, or Melbourne shunts). These patients will have an arterial saturation between 70% and 85% if the shunt is patent and the correct size. These shunts can block acutely (patient becomes acutely hypoxic), or the patient may outgrow them over time and become increasingly cyanotic.

These shunts are being used less frequently in the current era with surgeons opting for earlier complete repairs or cardiologists stenting lesions open in the catheter laboratory. Patients who undergo an MBTS today often will have complex underlying heart disease.

- **Bidirectional Glenn (BDG)** – The superior vena cava (SVC) is transected above the right atrium and anastomosed to the right pulmonary artery. Blood flows from the SVC through the lung and back to the left atrium (LA) down a pressure gradient from the SVC to the LA. This may be stage two of a three stage surgical approach to single ventricle anomalies. Patients continue to be cyanotic as inferior vena cava (IVC) blood returns to systemic circulation bypassing the lungs (arterial saturations are in the 75% and 90% range).
- **Fontan** – The modern Fontan procedure consists of anastomosing a GorTex conduit from the IVC to the pulmonary artery as well as the SVC to the pulmonary artery. Pulmonary blood flow is driven by the gradient from the venous circulation to the common atrium. There may be deliberate connection between this conduit and the common atrium which allows some blood to bypass the lungs. The result is some arterial desaturation, but atrial filling will be maintained despite increases in pulmonary vascular resistance (PVR).

Presentations with complications can be divided into early and late, although the two overlap. Early consultation with both cardiology and cardiothoracic teams is recommended.

Early

- Mediastinitis:
    - Patients may present with skin redness, wound discharge, and fever.
    - Most require admission for systemic antibiotics and possible debridement.
    - Cardiac surgical team should review early.
- Cardiac failure:
    - This may be the result of changes in patient medication or the development of a surgical complication (worsening function after valve repair can occur early).

    - Pericardial effusions can cause cardiac failure and tamponade.
    - Manage as per described for primary cardiac failure above.
- Respiratory failure:
    - Often secondary to cardiac failure
    - Cardiology review including echocardiogram should be sought.
- Arrhythmia:
    - Either brady or tacharrhythmias may occur and result in decreased cardiac output. This may be poorly tolerated by patients with reduced cardiac reserve.
    - Always obtain a 12-lead ECG if an arrhythmia is suspected.
    - Treatment likely to be complex, and early cardiology advice should be sought.
- Cyanosis:
    - Shunt thrombosis will reduce pulmonary blood flow resulting in worsening of cyanosis.

Late

- Endocarditis:
    - Patients with shunts, conduits, prosthetic valves are at highest risk. They typically present with low-grade fever and general deterioration. Cardiac failure may develop later.
    - Diagnosis requires a high index of suspicion.
    - Management requires prolonged antibiotic therapy.
- Cardiac failure:
    - Management as above.
- Cerebrovascular accident (CVA):
    - May be embolic or haemorrhagic.
- Arrhythmia:
    - Management as above.
- Cyanosis:
    - The commonest cases are children with known CHD who have some progression of their disease or develop another problem which exacerbates their cardiac condition, e.g. shunt thrombosis.
    - Tetralogy of Fallot with worsening RV outflow tract

obstruction, progressive pulmonary stenosis, stenosis of abnormal aortopulmonary collateral vessels.

- Management of ‘Tet spells’ described above.

Congenital heart disease and intercurrent illness

These patients should be seen by a senior emergency department (ED) doctor and discussed with a paediatric cardiologist before being discharged home from ED for any condition other than something very minor. It is recommended that ED clinicians have a low threshold for both admission to hospital and for seeking help in managing patients with CHD and intercurrent illness. Apart from problems with complex anatomy, they may have limited sites for intravenous access and may have challenging behaviours due to previous hospitalisations.

Dehydration – including gastroenteritis

- Patients with congenital heart disease often cope poorly with dehydration. This is particularly true of patients relying on relatively high right-sided pressures and those with single ventricle systems.
- Patients with shunts are also at high risk of shunt thrombosis with dehydration.
- Lower threshold for admission and more intensive management of gastroenteritis and/or dehydration are required.
- Patients on warfarin need careful management of their medication during intercurrent illnesses to avoid both thrombosis and/or haemorrhage.

Respiratory disease

- Respiratory syncytial virus (RSV) infection, which is the commonest cause of bronchiolitis, results in much higher mortality and morbidity in children with congenital cardiac disease.
- Mild respiratory disease can result in significantly worse cyanosis in some patients due to a rise in PVR.
- The combination of reduced pulmonary blood flow and respiratory disease (e.g. bronchiolitis) can result in very poor oxygenation.
- Lower threshold for admission and supplementary oxygen are

recommended.

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5.7

Acute rheumatic fever

Rachel H. Webb, and Jocelyn Neutze

ESSENTIALS

1 Acute rheumatic fever (ARF) is a systemic inflammatory response to group A streptococcal throat infection.

2 The major complication is progressive damage of cardiac valves leading to chronic rheumatic heart disease (RHD).

3 Although uncommon in developed countries, very high rates occur amongst Indigenous Australians, New Zealand Māori and Pacific Islanders.

4 Diagnosis is based on the presence of clinical features (carditis, arthritis, chorea, and rarely skin rashes and nodules) with evidence of systemic inflammation and recent group A streptococcal infection.

5 Treatment of ARF is supportive and involves controlling joint pain and managing carditis, heart failure and chorea.

6 Long-term antibiotic prophylaxis with intramuscular benzathine penicillin prevents further attacks of ARF and improves cardiac outcomes.

7 Antibiotics are recommended prior to dental procedures to prevent infective endocarditis.

Introduction

Acute rheumatic fever (ARF) is an inflammatory disease that may follow group A β-haemolytic streptococcal pharyngitis. It primarily affects connective tissue,

causing carditis, arthritis and chorea. Approximately 50% of individuals are left with chronic rheumatic heart disease (RHD), which is associated with significant morbidity and mortality.

Epidemiology

ARF is most common in children aged between 5 and 15 years; however, cases occasionally occur in preschool-aged children and young adults. Indigenous Australians, New Zealand Māori and Pacific Islanders have some of the highest reported rates of ARF and RHD in the world.[1,2] In highly developed countries, ARF and RHD are extremely uncommon.

The risk ARF is strongly linked to household crowding and material deprivation.[3] A family history of ARF is often observed; however, the genetic susceptibility is still incompletely understood.[4,5]

Pathophysiology

The exact pathogenesis of ARF remains unclear. It is postulated that in a susceptible individual, Gram-positive group A β-haemolytic streptococci (*Streptococcus pyogenes*) pharyngitis leads to an antibody response and that anti-streptococcal antibodies cross-react with host connective tissue in the cardiac valves, joint synovium and basal ganglia. This process is described as molecular mimicry.[6] Long-term immunity does not occur, and unless further group A streptococcal infections can be prevented, the person remains susceptible to recurrent attacks of rheumatic fever and progressive heart valve damage.

Diagnosis of acute rheumatic fever

There is no single diagnostic test for ARF. The diagnosis relies upon the identification of clinical and laboratory features. The National Heart Foundations of Australia and New Zealand and the American Heart Association have developed local diagnostic criteria.[7,8] These guidelines are adapted from the American Heart Association Jones Criteria[9] and take into account evidence applicable to high-risk populations in the Oceania region (Table 5.7.1).

An initial episode of ARF may be diagnosed where two major or one major and two minor manifestations are present, along with evidence of preceding group A streptococcal (GAS) infection. Persons at high risk who do not fulfil all

criteria may be labelled as ‘probable or possible ARF’.

Clinical manifestations (history and examination)

Some individuals may report pharyngitis several weeks prior, but this may not always be the case, especially in younger children.

Arthritis is the most common presenting manifestation. The arthritis of ARF is classically described as a migratory polyarthritis of the large joints; however, a much wider spectrum of joint manifestations can occur including monoarthritis and arthralgia. Arthritis in ARF usually responds very quickly to non-steroidal anti-inflammatory medication; an early administration of these can make the diagnosis more difficult. It is important to have a high index of suspicion in children from high-risk demographic groups who present with joint pain or limping.

Carditis manifested as valvulitis, myocarditis or pericarditis occurs in 80% of cases of ARF.[10] A new or changing murmur may be present; however, echocardiography can detect clinically inaudible valvular regurgitation and has been shown to be more sensitive and specific than auscultation for the diagnosis of carditis.[11] The mitral valve is most commonly affected, followed by the aortic valve. Carditis is usually asymptomatic unless severe and there is associated congestive heart failure and/or pericardial effusion. Cardiac arrhythmias can also occur in ARF. First-degree heart block is usually asymptomatic. Less commonly, junctional arrhythmias can cause palpitations, breathlessness or collapse.

Table 5.7.1

Diagnostic criteria for acute rheumatic fever (New Zealand and Australian High Risk individuals)

Major manifestations	Carditis (including subclinical carditis detected on echocardiography) Joint manifestations: Aus: polyarthritis, aseptic monoarthritis, polyarthralgia NZ: polyarthritis, aseptic monoarthritis Erythema marginatum Subcutaneous nodules Chorea
Minor manifestations	Fever (documented >38°C) ESR: Aus: ≥30 mm hr or CRP ≥30 mg L NZ: ≥50 mm hr or CRP ≥30 mg L

	Prolonged PR interval on ECG Joints: Aus: monoarthralgia NZ: polyarthralgia
	Plus evidence of recent group A streptococcal infection Positive throat swab culture for group A streptococcus OR elevated streptococcal antibody titres

Aus, Australia; NZ, New Zealand; ESR, erythrocyte sedimentation rate; CRP, C-reactive protein; ECG, electrocardiograph.

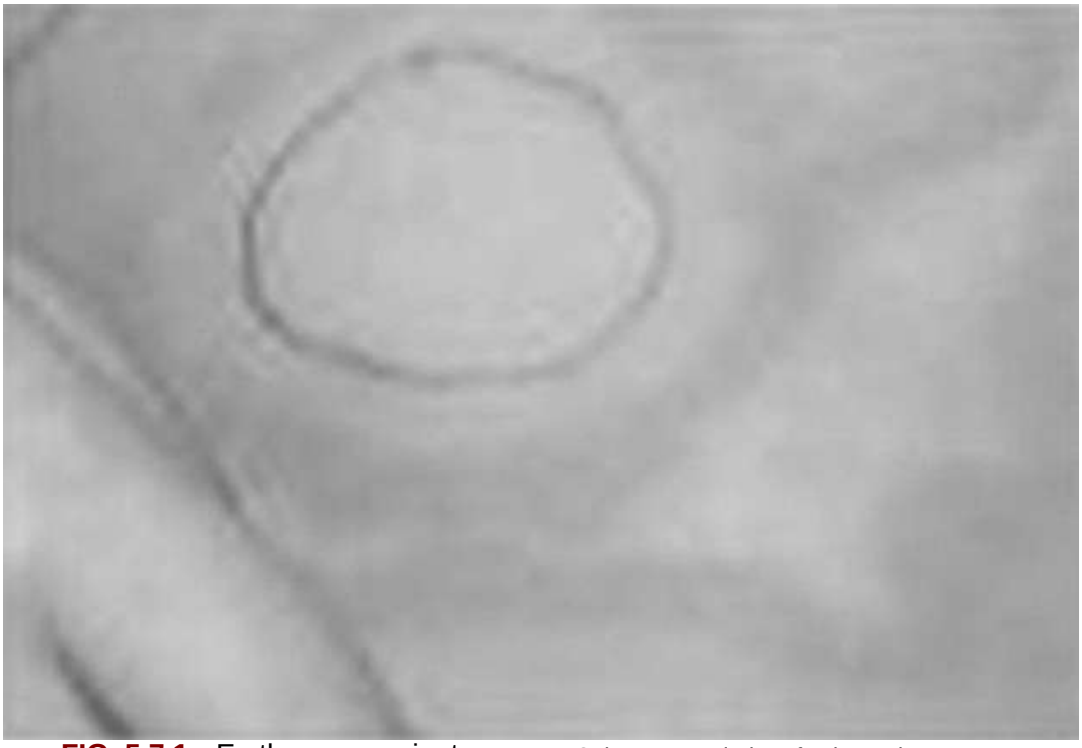

FIG. 5.7.1 Erythema marginatum. From Cohen & Powderly, Infectious Diseases, 2nd ed., Copyright © 2004 Mosby, An Imprint of Elsevier with permission.

Chorea may present with abrupt onset of involuntary movements of the face and limbs, poor attention span, unsteady gait or difficulty with handwriting. It occurs in up to 30% of patients with ARF. It presents with jerky, purposeless movement of limbs, speech impairment, involuntary grimacing or emotional lability and may appear several months after the original group A β-haemolytic streptococcal infection. Movements may be asymmetric and usually disappear during sleep. Symptoms may fluctuate and worsen during times of stress or intercurrent illness and may take several months to settle. The presence of chorea is sufficient for the diagnosis of ARF without other manifestations, providing differential diagnoses have been excluded.

Erythema marginatum is a distinctive feature of rheumatic fever but is rare and occurs in <5% of patients. Lesions usually begin as small, pink, non-pruritic macules or papules on the trunk or limbs. The lesions gradually spread to develop a raised pink serpiginous edge with central clearing and remain late into the course of illness. This rash may be difficult to see in dark-skinned individuals (Fig. 5.7.1).

Subcutaneous nodules are also very rare but are a highly specific manifestation of ARF. They are firm, non-tender nodules found over the extensor surface of the elbow, metacarpophalangeal joints, knees, and ankles. Up to three or four can appear in the first weeks of illness and may remain for up to a month.

Other symptoms are less specific symptoms, such as weight loss, fatigue, pallor, headache, abdominal pain and epistaxis, and can also occur. These do not form part of the formal diagnostic criteria.

Table 5.7.2

Streptococcal antibody titres (ref Australia and New Zealand rheumatic fever guidelines)

Age group	Anti streptolysin titre (ASOT) (IU mL)	Anti-deoxyribonuclease B titre (Anti-DNAase B) (IU mL)
Australia		
1–4 years	170	366
5–14 years	276	499
15–24 years	238	473
New Zealand – all ages		
All ages	480	680

Table 5.7.3

Differential diagnosis of acute rheumatic fever

Arthritis	Post-infectious reactive arthritis – Hepatitis B, Epstein–Barr virus, cytomegalovirus, rubella, influenza, parvovirus B19, mycoplasma, yersiniosis, Lyme disease Juvenile idiopathic arthritis and other connective tissue disorders Gout Leukaemia/lymphoma Sickle cell arthropathy Septic arthritis Gonococcal arthritis
Carditis	Infective endocarditis Viral myocarditis Pericarditis Congenital heart disease including mitral valve prolapse Innocent murmur Kawasaki disease
Chorea	Wilson’s disease Huntington’s chorea

	Ataxia telangiectasia Lesch–Nyhan syndrome Tic disorder Drugs (anticonvulsants, antidepressants, lithium, methylphenidate) Choreo-athetoid cerebral palsy Pregnancy Hyperthyroidism
Erythema marginatum	Urticaria Other causes of annular erythema
Subcutaneous nodules	Rheumatoid arthritis Erythema nodosum

Investigations

Laboratory evidence of group A β-haemolytic streptococcal infection

Preceding group A β-haemolytic streptococcal infection should be demonstrated by a positive throat culture or elevated or rising anti-streptococcal antibody titres (anti-streptolysin and anti-DNAase B). Antibodies rise in the first month post-infection, plateau at 3–6 months, and normalise after 6–12 months. Streptococcal antibody titres should be interpreted according to local reference ranges[7,8] (Table 5.7.2).

Acute phase reactants

The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) should be measured and interpreted according to local guidelines. The ESR is usually above 80 mm hr and typically remains elevated for several weeks.

ECG

An ECG should be performed to identify pericarditis and first-degree heart block. PR interval should be interpreted according to age and heart rate.

Imaging

Echocardiography is recommended in all cases of suspected ARF. Criteria for interpretation of echocardiography in suspected ARF can be found in the Australia and New Zealand guidelines.[7,8] As carditis may evolve over several weeks, if the first echocardiogram is normal, a follow-up echocardiogram is recommended in 2–4 weeks.

Differential diagnosis

Each clinical manifestation of ARF is associated with multiple alternative diagnoses, and these should be carefully considered whenever a person presents with suspected ARF (Table 5.7.3).

Joint manifestations can mimic post-infectious reactive arthritis due to a variety of infections (parvovirus, hepatitis B, cytomegalovirus, Epstein–Barr virus, mycoplasma, yersinosis, campylobacter and rubella) or autoimmune conditions such as juvenile idiopathic arthritis. Septic arthritis should be considered if a child presents with monoarthritis and fever. Rarely, blood disorders such as sickle cell disease and leukaemia may present with arthritis. In older and overweight individuals, gout should also be considered.

Table 5.7.4

Drug doses in acute rheumatic fever and rheumatic heart disease

Drug	Indication	Dose
Benzathine benzylpenicillin	Long-term secondary prophylaxis of acute rheumatic fever	900 mg (1.2 mega units) intramuscular (IM) every 28 days Small children <20 kg (Aus) <30 kg (NZ) 450 mg (0.6 mega units) IM every 28 days
Phenoxymethylpenicillin (VK)	Group A streptococcal pharyngitis	<20 kg 250 mg bd for 10 days >20 kg 500 mg bd for 10 days
Erythromycin	Penicillin allergy – Group A strep pharyngitis or long-term prophylaxis	20 mg kg dose bd
Naproxen	Arthritis	10–20 mg kg day divided into two doses (max 500 mg dose)
Ibuprofen	Arthritis	10 mg kg dose 8-hourly (max 400 mg dose)
Carbamazepine	Severe chorea	10 mg kg day divided into two doses
Sodium valproate	Severe chorea	15–20 mg kg day divided into three doses

IM, intramauscular; Aus, Australia; NZ, New Zealand.

Sydenham's chorea is a diagnosis of exclusion after eliminating other causes of movement disorder such as drug toxicity, systemic lupus erythematosus (SLE), intracranial tumour and Wilson's disease. Tic disorders may also be difficult to distinguish from rheumatic chorea. Careful consideration of the differential diagnosis is particularly important if the chorea is atypical, the child comes from a low-risk demographic group for ARF, or if there are no other

features to support a diagnosis of ARF. Assessment by an experienced RF clinician or paediatric neurologist may be required.

Treatment

Acute management

Children who present with suspected ARF should be admitted to hospital for further evaluation and management.

A throat swab should always be obtained and then penicillin given to eradicate group A streptococcus. Phenoxymethylpenicillin 250 mg or 500 mg po q 12 h is usually given initially before commencing long-term secondary antibiotic prophylaxis with intramuscular (IM) benzathine penicillin. Erythromycin may be substituted in cases of penicillin sensitivity. Penicillin is the only treatment shown to improve long-term cardiac outcomes.

Symptomatic treatment of arthritis

Rheumatic arthritis can be very painful, and children may require bed rest initially, along with regular analgesia. Paracetamol is preferred until the diagnosis of ARF is confirmed, as non-steroidals may mask the evolution of the joint features. Once the diagnosis is secure, naproxen twice daily is the preferred treatment. This is less likely to cause liver and metabolic derangement than high-dose aspirin.[12] Most individuals with ARF arthritis respond to non-steroidals within several days and require 1–2 weeks of regular treatment.

Treatment of carditis and control of heart failure

Bed rest is recommended for individuals with severe carditis, usually for several weeks followed by a period of gentle ambulation.

Angiotensin-converting enzyme inhibitors and diuretics may be indicated in severe carditis. Prednisone is sometimes used in selected cases with severe carditis and pericardial effusions, but its use is controversial. A 2015 Cochrane review found that corticosteroids did not improve cardiac outcomes at 12 months.[13] Heart valve surgery may be required for severe valvular heart disease. Surgery is usually deferred until the inflammatory phase of ARF has settled. Valve repair is preferred over mechanical replacement wherever technically possible, due to the high morbidity and mortality of mechanical valves and

anticoagulation in young people.[14,15]

Management of chorea

The treatment of chorea is supportive. Education for caregivers is important as chorea may take a fluctuating course, sometimes for a number of months. The child should be carefully assessed regarding safety of gait and capacity to perform activities of daily living. Medication is reserved for cases where there is substantial functional impairment (for example unable to walk unaided, unable to feed self). Carbamazepine and valproic acid are most commonly used.

Prevention and prophylaxis

Continuous anti-streptococcal prophylaxis is recommended in all patients with ARF/RHD to prevent recurrences. Australia and New Zealand guidelines recommend benzathine penicillin 900 mg (1.2 mega units) IM every 4 weeks (or 3 weekly following a recurrence). Oral penicillin use is discouraged as it is associated with higher risk of recurrence and poorer cardiac outcomes. In cases of penicillin allergy, specialist consultation should be sought. Oral erythromycin is used for severe penicillin allergy where desensitisation is not possible. Prophylaxis should continue for at least 10 years after the most recent episode of ARF or until age 21 years. Prolonged prophylaxis is until age 30, or 40 years is recommended for severe RHD. Antibiotic prophylaxis for endocarditis is also recommended_ENREF_16 (see Chapter 5.8 on Infective endocarditis), and annual influenza vaccination should be offered to those with significant RHD.

Prognosis

The inflammatory phase of ARF usually resolves within 2–3 months, occasionally lasting longer in severe carditis. Following a first episode of ARF, improvement in valvulitis occurs in most people with mild to moderate carditis; however, about 20% are left with severe RHD. Recurrences of ARF are extremely uncommon when there is excellent adherence with continuous 4-weekly injections. Recurrences lead to worsening RHD and increased risk of stroke, endocarditis, pregnancy complications for women and shortened life expectancy.

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5.8

Infective endocarditis

Rachel H. Webb, and Jocelyn Neutze

ESSENTIALS

1 Infective endocarditis is the microbial infection of cardiac endothelium.

2 Risk factors include congenital heart disease, central venous lines, implantable cardiac devices, intravenous drug use and immunodeficiency.

3 Diagnosis is based on the presence of infection with some or all of the following: valve dysfunction or intra-cardiac infection, peripheral embolisation and immunological phenomena.

4 Acute management includes elimination of causative organisms via antibiotics and surgery and treatment of associated complications, such as congestive heart failure.

5 In selected high-risk patients, antibiotic chemoprophylaxis is recommended prior to procedures likely to result in bacteraemia.

Introduction

Infective endocarditis (IE) is the microbial infection of the endothelial lining of the heart. Infection may be bacterial or fungal and usually occurs in those with structurally abnormal heart and/or risk factors, although it may occasionally occur in individuals with structurally normal hearts. Presentation is usually acute, although it can be subacute, and the long-term morbidity and mortality are high.

Epidemiology

In recent years, the global epidemiology of IE has changed, with increasing incidence driven by increasing rates of healthcare-associated IE and an increased burden of *Staphylococcus aureus*.[1] In children, incidence of IE is also increasing, due to the improved survival of children with complex congenital heart disease and increased numbers of medically fragile and immune-compromised children. In developed countries, approximately 70–80% of children with bacterial endocarditis have pre-existing structural heart disease.[2] Other risk factors include rheumatic heart disease, intravenous drug use, presence of indwelling central venous lines or implantable cardiac devices, immunodeficiency and previous bacterial endocarditis.[3]

Pathophysiology

Turbulent cardiac blood flow through a structurally abnormal heart can damage endothelium and lead to overlying thrombus formation. Transient bacteraemia may infect the thrombus, and a vegetation is formed; vegetations can then cause valvular insufficiency. Perivalvular extension of vegetations can lead to the formation of intracardiac abscess, fistulae and occasionally conduction system abnormalities. Extracardiac manifestations result from peripheral embolisation of thrombus material with subsequent infarction and/or infection of involved tissue. Bacterial embolisation can also lead to mycotic aneurysm formation, characterised by infection and distension of the arterial wall. Mycotic aneurysms are most common in intracranial arteries.

Microbiology

Staphylococcus aureus and viridans streptococci are the most common organisms in children with IE. Less common aetiological agents include enterococci, *Staphylococcus epidermidis* and Gram-negative HACEK (*Haemophilus parainfluenzae*, *Aggregatibacter actinomycetemcomitans* and *Aggregatibacter aphrophilus*, *Cardiobacterium hominis*, *Eikenella corrodens* and *Kingella kingae*) bacilli. Fungi such as *Candida* and *Aspergillus* species may cause IE in neonates or immune-compromised patients. Other agents such as *Coxiella*, *Streptococcus pneumoniae*, *Streptococcus pyogenes*, *non-toxigenic Corynebacterium diphtheriae*, Bartonella and Listeria are rare causes of endocarditis in childhood.

History

Patients with acute IE are often readily identified by systemic toxicity and bacteraemia. The less common presentation of subacute endocarditis is more non-specific and may include fever, anorexia, malaise, headache, abdominal pain and arthralgia. A high degree of suspicion is required to identify these patients. Any child presenting with unexplained prolonged fever, persistent bacteraemia, new neurological deficit or other embolic phenomena should be evaluated for endocarditis. Systemic embolisation occurs in up to 50% of patients with IE and is most frequently seen in the first month post-diagnosis. Embolic events may cause infarction or abscess formation in the brain, lungs, kidneys, spleen, bone and extremities. Endocarditis may also present with cardiac failure secondary to acute valvular dysfunction, intracardiac abscess, fistulous tract formation or destruction of indwelling prosthetic material.

Examination

The physical signs of endocarditis may be subtle. A new or changing murmur can be difficult to distinguish from an innocent murmur or pre-existing cardiac abnormality. Other findings include congestive heart failure. Evidence of peripheral embolisation such as Osler nodes (tender, red nodules of finger pulps), Janeway lesions (non-painful, haemorrhagic areas of palms or soles) and splinter and subungual haemorrhages are very unusual in children. Roth spots (retinal haemorrhages with a pale centre) are occasionally seen on fundoscopy. Acute neurological deficit may arise from embolic infarcts, abscesses or intracerebral haemorrhage. Splenomegaly is present in up to 30% of patients, and new onset clubbing may occur. Immune-mediated glomerulonephritis may result in haematuria, proteinuria and renal impairment.

Diagnosis is based on the modified Duke Criteria for Diagnosis of Infective Endocarditis[4] (see below).

Modified Duke criteria

Major criteria

Positive blood cultures for infective endocarditis

- Typical microorganisms from at least two separate blood cultures

(viridans streptococci, *Streptococcus bovis*, HACEK organisms, *Staphylococcus aureus*) or community-acquired enterococcus) or a single positive blood culture for *Coxiella burnetii*

- At least two positive cultures drawn >12 hours apart for organisms that are typical causes of IE or for organisms that are more commonly skin contaminants three, or a majority of four or more, cultures
- Evidence of endocardial involvement
- Echocardiogram positive for IE (mobile echo-dense intracardiac mass, periannular abscess, new partial dehiscence of prosthetic valve depicted on echocardiogram)
- New valvular regurgitation depicted on echocardiogram.

Minor criteria

- Predisposing factor, history of intravenous (IV) drug use or congenital heart disease
- Fever >38°C
- Vascular phenomena – arterial emboli, pulmonary infarcts, mycotic aneurysm, intracranial haemorrhage, conjunctival haemorrhage, Janeway lesions
- Immunological phenomena – glomerulonephritis, Osler nodes, Roth spots, positive rheumatoid factor
- Positive blood cultures or serological evidence of infection not meeting above criteria.

There are three diagnostic categories within the Modified Duke criteria: definite endocarditis, possible endocarditis and rejected cases. Definite IE requires the presence of two major, one major with three minor, or five minor criteria. Possible endocarditis is defined by one major with one minor or three minor criteria. Rejected cases are those where manifestations are explained by a clear alternate diagnosis or resolve after antibiotic therapy for 4 days or less.

Investigations

Laboratory

The most useful laboratory investigation for IE is the identification of a

causative organism from blood cultures. Wherever feasible, three sets of blood cultures should be collected prior to antibiotic administration. Up to 10% of all cases of IE remain culture negative, usually where organisms are highly fastidious or the patient has received prior antibiotic treatment. Inflammatory markers such as white cell count, C-reactive protein, erythrocyte sedimentation rate and rheumatoid factor may also be abnormal.

Imaging

The investigation of suspected endocarditis should include an electrocardiograph (ECG) and chest X-ray (CXR), but findings are non-specific. CXR may reveal evidence of cardiac or pulmonary complications but is not diagnostic of IE. Transthoracic echocardiography is essential, and image quality is usually adequate in most paediatric patients.[5]

Transoesophageal echocardiography (TOE) may be required in selected situations when transthoracic views are inadequate, particularly in right-sided IE or if para-aortic abscess is suspected.

Differential diagnosis

The presentation of IE is similar to many systemic inflammatory conditions. Positive blood cultures often confirm the diagnosis, but in presentations where blood cultures remain negative, other diagnoses must be considered. Rheumatic fever, Kawasaki disease, systemic lupus erythematosus and leukaemia can all have similar presenting features, hence the need for rigorous diagnostic criteria.

Treatment

Medical

Principles of managing IE in children follow those in adults.[6] As bacteria in vegetations are embedded in a dense fibrin matrix, eradication of infection requires prolonged high-dose parenteral therapy, preferably with bactericidal rather than bacteriostatic agent/s. The causative organism is usually unknown at presentation, and the choice of empiric treatment should take into consideration the patient's clinical presentation (acute vs. subacute) and background history (whether immune compromised, underlying heart disease, native valve or prosthetic material, colonisation with methicillin-resistant *Staphylococcus*

aureus [MRSA] or other multi resistant organisms). An anti-staphylococcal agent should be included in the initial combination.

For native valve IE in children, Australasian antibiotic guidelines recommend high-dose benzylpenicillin and flucloxacillin intravenously every 4 hours in combination with intravensous gentamicin. Vancomycin should be added if there is a prosthetic valve, penicillin allergy, healthcare-associated infection or risk of MRSA. The duration of parenteral therapy is usually 4–6 weeks, but longer courses may be required in prosthetic valve IE, recurrent IE, or complicated multifocal infection. Early consultation with infectious diseases/microbiology should be undertaken to optimise therapy and ensure appropriate laboratory investigations have been performed.

Therapeutic drug monitoring should be undertaken for patients receiving aminoglycosides or vancomycin, along with careful monitoring for nephrotoxicity and ototoxicity.

Surgical

The need for surgical management of IE depends on the severity of complications and response to medical management. Perivalvular abscesses, obstructive vegetations, cardiac fistulae and prosthetic dehiscence often require surgery. Other indications include persistent bacteraemia, recurrent embolisation, and fungal endocarditis.

In adults, earlier surgery is associated with improved outcomes.[7]

Prognosis

The overall mortality of IE remains high at approximately 5–10%,[2,3] and morbidity is considerable. Poor prognosis is associated with *Staphylococcus aureus* infection, fungal IE, IE where there is indwelling prosthetic material, significant embolic complications, and intra-cardiac abscess formation.

Prevention

Recommendations regarding antibiotic prophylaxis for IE vary across the world.[9,10] Australasian guidelines adopt recommendations from the American Heart Association but also acknowledge that rheumatic heart disease (RHD) is a significant risk factor for IE. Conditions where prophylaxis is recommended

include:

- prosthetic cardiac valves
- cardiac transplant with subsequent valvulopathy
- rheumatic heart disease
- previous infective endocarditis
- congenital heart disease if:
    - unrepaired cyanotic defect
    - completely repaired defect with prosthetic material up to 6 months post procedure
    - repaired defects with residual defect at prosthetic part.

Chemoprophylaxis depends on the procedure being performed. Australasian guidelines suggest dental and upper respiratory tract procedures (e.g. re-implantation of avulsed tooth) receive amoxicillin (50 mg kg orally) as a single dose 1 hour before the procedure. Cephalexin, clarithromycin or clindamycin is an alternative for penicillin sensitivity and for individuals with RHD on benzathine penicillin prophylaxis. Genitourinary and gastrointestinal procedures require ampicillin just before the procedure or vancomycin if penicillin allergic. Endotracheal intubation and urinary catheterisation do not require antibiotic prophylaxis. Current therapeutic guidelines should be consulted to optimise management in each case.[11]

Controversies

There is no published evidence to show that antibiotic prophylaxis prevents IE. Endocarditis is thought more likely to result from random bacteraemia than from bacteraemia associated with dental, GI or GU procedures. The American Heart Association has recently revised guidelines to reduce the categories of patients for whom prophylaxis is recommended, and in the UK the National Institute for Clinical Excellence (NICE) does not recommend routine antibiotic prophylaxis.[12]

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5.9

Kawasaki disease

Daryl Efron

ESSENTIALS

1 Kawasaki disease (KD) is an uncommon emergency department (ED) presentation but should be considered in any infant or young child with an unexplained prolonged fever, particularly in the presence of a rash and red eyes.

2 The diagnosis is *clinical* with the presence of fever and four out of five other criteria.

3 Diagnostic criteria can appear sequentially and may not all be present at the same time, which can make early diagnosis difficult.

4 Patients with 'incomplete (atypical) KD' are being increasingly recognised. These are often infants with fever ≥5 days with fewer than four of the other typical signs but with a clinical presentation consistent with KD.

5 Any child with suspected KD should be treated immediately regardless of the duration of symptoms to reduce the risk of cardiovascular complications.

6 Intravenous immunoglobulin (single dose of 2 g kg^{-1}) significantly reduces the risk of coronary artery abnormalities, especially if given in the first 10 days of the illness.

Introduction

Kawasaki disease (KD) is an acute, self-limiting vasculitic illness predominantly

affecting infants and young children. It is now a leading cause of acquired heart disease in children in Western countries. The diagnosis is made clinically, and effective treatment is available to reduce the likelihood of potentially fatal coronary vasculitis. KD was first described in 1967 as ‘mucocutaneous lymph node syndrome’ in a series of 50 Japanese children.[1] Although it is most common in Japanese and Korean children (annual incidence 90–215/100,000 children younger than 5 years), it occurs in all ethnic groups, with an annual incidence in the United States of approximately 10–20/100,000 children younger than 5 years old. The majority of cases (85%) occur in children aged less than 5 years of age. It is 1.5 times more common in boys than girls.[2]

Many features of KD suggest an infectious aetiology. These include seasonal variation (peak in winter/spring); occasional outbreaks; 10-fold higher risk in siblings than in the general population; rarity in young infants (suggesting protection from maternally acquired antibody); and low recurrence rate (4%, which suggests acquired immunity), as well as the resemblance of the clinical presentation to other self-limiting infectious diseases, such as measles, adenovirus infection and staphylococcal toxic shock syndrome. However, despite much investigation of a variety of viral and bacterial pathogens, there is as yet no good evidence to implicate any known organism in KD. The ethnic variation suggests a genetic predisposition.

There is debate as to whether the inflammatory response in KD is initiated by a conventional antigen or a superantigen, and there are some immunological features to support both hypotheses. Unreplicated reports of increased expression of specific T-cell receptor V β-regions suggest toxin activation, whereas infiltration of paratracheal and vascular tissue with reactive clonal IgA plasma cells suggests entry of a conventional antigen via the respiratory route.[3]

Pathophysiology

The pathophysiology of KD involves vasculitis of medium-sized vessels including coronary, renal, hepatic and splanchnic arteries, beginning in both adventitial and intimal surfaces and proceeding toward the media. Coronary changes occur in approximately 20% of untreated patients.[4] Immune activation involving cytokines and growth factors leads to inflammation and aneurysm formation, with the risk of thrombosis. The process evolves for a long period after the acute illness. In the majority (50–75%) of patients with echocardiographically demonstrable coronary artery lesions, the vessels remodel

and have a normal appearance within a year or so. However, there is evidence of subtle long-term changes in coronary artery function, the clinical significance of which remains unclear.[5] The risk of early adult coronary artery disease in these patients is unknown.

A diffuse inflammatory process of a variety of tissues has been found in autopsy specimens including lymph nodes, liver and gallbladder.[2] Endothelial changes are prominent, with hyperplasia, necrosis and thrombosis. Myocardial abnormalities include hypertrophy of myocytes and fibrosis.

Clinical features

KD should be considered in the differential diagnosis of all infants and young children with a fever, rash and red eyes, as well as those with a prolonged fever without an alternative explanation. The diagnostic criteria are outlined in Box 5.9.1.

The diagnosis can be made earlier than day 5 if other features are present. This is important, as there is evidence that earlier administration of intravenous immunoglobulin (IVIG) is associated with a shorter illness and reduced risk of coronary disease.[6] Individual clinical signs are often transient and appear in a variable sequence through the course of the illness, so the history often provides important clues together with repeated physical examination.

The fever in KD is typically high (>38.5°C) and poorly responsive to antipyretics. The fever lasts an average of 12 days without treatment. Most children with KD have marked irritability.

Box 5.9.1 Diagnostic criteria for Kawasaki disease

Fever (at least 5 days) without any other explanation:

- Plus four of the following five features:
    - Bilateral bulbar conjunctival injection
    - Enanthem – dry/cracked/fissured/injected lips, oropharyngeal injection/erythema, strawberry tongue
    - Exanthem – polymorphous rash
    - Peripheral changes – erythema (palms and soles), oedema (hands and feet), desquamation (2nd to 3rd week)
    - Cervical adenopathy (at least one node >1.5 cm in diameter).

In 70–90% of patients a rash is present. This may appear on the first few days of the illness as perineal erythema and desquamation, followed by a maculopapular, urticarial, scarlatiniform, morbilliform rash of the trunk and limbs. Redness or crusting of a BCG scar is commonly seen in KD.

The bright red eyes seen in KD demonstrate characteristic sparing of the avascular perilimbic region, giving the appearance of a thin white halo around the cornea. There is not usually an exudate. Many children with KD are photophobic. Anterior uveitis may also be present.

Mucositis is typical, with cracked red lips and a strawberry tongue.

Unilateral cervical lymphadenopathy may be present, usually in the anterior chain.

Extremity changes include erythema of the palms and soles and oedema of the dorsum of the hands and feet. The subacute phase occurs from 2 to 4 weeks, with resolution of fever. Periungual desquamation of fingers and toes may occur during this time.

KD is a multisystem disease with many and varied clinical manifestations. In addition to those in the diagnostic criteria, common features include marked irritability, diarrhoea, cough, arthralgia/arthritis (often seen in the convalescent phase), urethritis with sterile pyuria, mild hepatic dysfunction, hydrops of the gallbladder and aseptic meningitis. Diagnosis is more likely to be delayed in older children, in whom less classical manifestations such as gastrointestinal and joint symptoms often predominate[7] and also in children <6 months of age.

Incomplete Kawasaki disease

Cases of incomplete or ‘atypical KD’ are being increasingly recognised, in which full criteria are not met, but the patient has coronary artery abnormalities. This is more common in young infants.[8] The incidence of coronary artery aneurysms (CAA) is at least as high in incomplete KD as in classical cases. Given that infants under 6 months appear to be at increased risk of developing coronary abnormalities;[8] a lower threshold for treatment is probably indicated in this group. KD should be considered in all children with unexplained fever for 5 days and at least two major features of KD and any infant with unexplained fever for over a week.[4]

Differential diagnosis

Depending on which features are present, a broad differential diagnosis needs to be considered in cases of possible KD. These include viral infections (e.g. measles, adenovirus, enterovirus), streptococcal and staphylococcal toxic-mediated illness (scarlet fever, toxic shock syndrome), drug reactions (including serum sickness and Stevens–Johnson syndrome), inflammatory bowel disease, systemic juvenile idiopathic arthritis, and malignancy.

Complications

The major concern with KD is coronary artery disease. Younger children, especially under 12 months of age, are at highest risk. Changes seen include dilatation (ectasia) and discrete aneurysms. Aneurysms are classified according to the internal diameter into small (<3 mm), medium (3–6 mm), large (6–8 mm) and giant (>8 mm). Although aneurysms rarely form in the first 10 days of KD, early echocardiographic signs of coronary arteritis may be seen, including perivascular brightness, ectasia and lack of tapering.[4] Other findings may include decreased left ventricular contractility, mitral regurgitation, and pericardial effusion.

Coronary artery occlusion is most likely to occur in giant aneurysms, through a combination of sluggish blood flow and fibrotic stenosis at proximal and/or distal ends of the aneurysm. Thrombosis of coronary aneurysms can cause myocardial infarction and sudden death. Mortality is increased over the background rate within the first 2 months. Rarely, myocarditis can occur, causing congestive cardiac failure or arrhythmias. Fusiform aneurysms may develop in the brachial arteries (axillae), and peripheral perfusion may be compromised resulting in cool or discoloured digits. Thrombosis of peripheral arteries can lead to ischaemia and gangrene.

Macrophage activation syndrome is a rare but serious complication of KD. This should be considered if the patient has an extremely high serum ferritin level (>5000 ng mL).

Investigations

Any child who presents with possible features of KD should be discussed with a paediatrician.

There is no definitive diagnostic test for KD. Investigation is directed toward excluding alternative diagnoses, as well as gathering supporting evidence for the

diagnosis of KD. At least one set of blood cultures should be taken, and cultures of urine, cerebrospinal fluid and other sites may be indicated. Serological testing for group A *Streptococcus* and for specific viruses such as measles may be helpful. In KD, the peripheral white cell count and inflammatory markers are usually significantly raised, and there is often a normocytic, normochromic anaemia. The erythrocyte sedimentation rate (ESR) may continue to rise as the child improves. Note that intravenous immunoglobulin (IVIG) causes a raised ESR, so this test is not useful after treatment. The C-reactive protein (CRP) is a good marker of disease activity/recovery. A marked thrombocytosis is commonly seen in the 2nd to 3rd week of the illness. Pyuria, due to sterile urethritis, is often found on a voided sample (not on suprapubic aspirate or catheter). Liver function tests are often abnormal, particularly gamma glutamyl transferase and transaminases. Hyponatraemia can be seen and is associated with an increased risk of CAA. Exclusion of markers for connective tissue disorders may be useful.

Echocardiography should be considered as soon as the diagnosis of KD is suspected. The American Heart Association guidelines suggest echocardiograms at diagnosis, 2 weeks (time of maximal CAA formation), 6 weeks and 12 months (looking for late sequelae). However, it may be reasonable to avoid the initial two echocardiograms in those who present early (<10 days), have no ischaemia on their ECG and respond to IVIG.[9]

If abnormalities are demonstrated on echocardiography, closer cardiology follow-up may be required, depending on the lesion. A 12-lead ECG is most often normal but may show dysrhythmias, change in PR or QT intervals or non-specific ST changes.

Treatment

Treatment of KD is directed towards reducing the inflammation as rapidly as possible. The clinical markers of response to therapy are the temperature and patient's general well being, supported by reduction in the white cell count and CRP.

IVIG therapy has been demonstrated to induce resolution of fever as well as significantly reduce the risk of coronary artery abnormalities (from around 20% to around 3–5%) if given within the first 10 days of the illness.[10] The precise mechanism of action of IVIG is unknown. Theories include saturation blockade of Fc receptors, direct antibody activity against bacterial superantigen, an

unidentified causative pathogen or toxin, modulation of cytokine production or down-regulation of antibody synthesis. The recommended dose is 2 g kg^{-1} as a single infusion over 10–12 hours. There is some evidence that IVIG is also effective if given beyond 10 days;[11] however, treatment as early as possible is optimal.[6] IVIG should be given to patients with KD after the 10th day if they have persistent fever or other evidence of ongoing systemic inflammation.[4] The effect of IVIG in patients who have already developed CAAs is unknown, though there may be some benefit.[10] Different brands of IVIG may vary in their clinical effects, due to variation in sterilisation and other manufacturing procedures. However there is no evidence to support one brand over another. Haemolysis can occur in the days following IVIG administration. As passive antibody acquisition may interfere with immunogenicity, live vaccine administration (e.g. measles, varicella) should be postponed for 11 months in children who have been given IVIG.

In the 1980s, high-dose aspirin was shown to decrease the incidence of coronary artery involvement in KD.[12] The current role of aspirin in KD is difficult to determine as it has been used in combination with IVIG in the major trials. Many centres have used high-dose aspirin (30–50 mg kg^{-1} day^{-1} in 3–4 divided doses, for anti-inflammatory effect) initially and then switched to low-dose (3–5 mg kg^{-1} day^{-1}, antiplatelet effect) after the patient's fever resolves. However, in patients treated with IVIG, concomitant use of high-dose aspirin initially does not appear to result in shorter duration of fever or hospitalisation than low-dose.[13] Furthermore, the incidence of coronary artery aneurysm appears to be independent of aspirin dose.[14] Therefore low-dose aspirin seems to be sufficient for initial treatment. This is continued for 6–8 weeks and then stopped if there is no coronary involvement on echocardiogram. Ibuprofen is effective for KD-associated arthritis.

The role of corticosteroids in KD is a subject of continuing research. Studies of the addition of a single pulsed dose of steroids to IVIG as initial therapy have yielded conflicting results.[15,16] A large, multi-centre Japanese study found that the addition of corticosteroids (methylprednisolone 2 mg kg day in three divided doses for 5 days, followed by oral prednisolone 2 mg kg per day tapered over 15 days once the CRP had normalised) to IVIG was associated with reduced risk of coronary artery abnormalities in patients with severe KD from 23% to 3%.[17] This finding needs to be replicated in other populations as the incidence of coronary artery abnormalities is particularly high in Japanese KD patients. It is possible that a subset of patients at highest risk of developing CAAs may benefit;

however, risk scores for stratification of patients have not been validated.

Refractory Kawasaki disease

Up to 15% of patients with KD treated with IVIG and aspirin have persistence or early recrudescence of fever. In the absence of a clear alternative explanation (e.g. intercurrent infection) this is indicative of an ongoing vasculitic process[18] and a strong risk factor for the development of CAAs.[19] Children with ongoing or recurring fever beyond 24 hours after completion of IVIG infusion should be given a second dose of IVIG.[4] This is usually effective, although approximately 5% of patients remain febrile after two doses of IVIG.

Pulsed methylprednisolone (30 mg kg^{-1} daily for 1–3 days) has been shown to reduce fever in KD unresponsive to two doses of IVIG;[20] however, the effect on CAAs is uncertain.[4] A number of other treatments including plasmapheresis, cyclophosphamide, and tumour necrosis factor-α antagonists (e.g. infliximab, etanercept) have been reported in children with refractory KD. The role of these therapies remains unclear.[4]

Prognosis

Mortality in KD is less than 1%, mostly from myocardial infarction or arrhythmias. Recurrence is uncommon and is most likely to occur in children aged less than 3 years who had cardiac involvement initially and usually within 12 months of the initial episode.[21] Patients with recurrent KD appear to be at increased risk for cardiac sequelae, and so a lower threshold for treatment with IVIG in uncertain cases is appropriate.

Children without demonstrable cardiac disease appear to have an excellent prognosis, with long-term follow-up studies demonstrating absence of clinical sequelae for up to 21 years.[22] It is possible, however, that individuals who have had KD are at risk of early atherosclerotic heart disease.

The prognosis for children who have had coronary artery aneurysms is less clear. Most small- to medium-sized aneurysms resolve echocardiographically,[23] but healing involves fibrosis and calcification, with associated loss of vascular distensibility and reactivity. A proportion of CAAs progress to stenosis over time. Therefore children with KD who have had CAAs should have indefinite cardiology follow-up. These patients will generally be treated with long-term antithrombotic therapy to prevent myocardial ischaemia. Children receiving

long-term aspirin therapy should receive the influenza vaccine annually, as well as varicella vaccine, to prevent Reye syndrome. New antiplatelet agents are under investigation.[24] There have been reports of the use of thrombolytic agents in patients with KD with thrombus seen within coronary aneurysms.[25] Anticoagulation with warfarin is required for patients with 'giant' (≥8 mm) or multiple aneurysms, and surgical intervention (e.g. angioplasty, bypass grafting) is occasionally necessary.

Controversies

1 The immunological mechanism of the inflammatory response in KD is the subject of continuing research – there is some evidence to support a conventional antigen, but other evidence suggests a superantigen.
2 The optimal dose of aspirin in the acute febrile phase is uncertain.
3 The role of steroids in KD is debated.
4 The clinical significance of subtle long-term changes in coronary artery function in some patients who have had KD remains unknown.

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5.10

Cardiac arrhythmias

Gary David Williams

ESSENTIALS

1 Focus initially on airway, breathing and circulation, and then recognise and treat arrhythmias compromising cardiac output using the safest possible means.

2 If an arrhythmia is fast and regular try to assess the width of the QRS complex against age-based normal values.

3 Supraventricular tachycardia (SVT) accounts for 90% of all significant arrhythmias in children.

4 Any previous 12-lead electrocardiograph (ECG) may be of enormous value in diagnosing an arrhythmia.

5 Ventricular tachycardia (VT) has a wide range of potential aetiologies, and treatment is based on an assessment of degree of compromise and morphology of the tachycardia.

6 Continuous recording of a single-lead ECG during acute resuscitation manoeuvres (adenosine administration, vagal manoeuvres, cardioversion) may yield very valuable information.

7 As suggested by *Multi-Societal Database for Pediatric and Congenital Heart Disease* attempt to use a four-tiered approach to classify arrhythmias based on (1) location, (2) mechanism, (3) etiology and (4) duration.

8 Any child with a significant arrhythmia should be discussed with a paediatric cardiologist at the earliest possible time.

Introduction

Identification and management of the child with a cardiac arrhythmia in the emergency department (ED) require an initial focus on and, if required, attention to the patient's haemodynamic stability followed by a team approach to diagnose and treat the arrhythmia if necessary. Although arrhythmias occur less frequently in acutely ill infants and children compared to adults, vigilance is required, as a correct acute assessment and management of the arrhythmia will have significant long-term consequences.

The primary aim of this initial assessment and resuscitation phase in an unstable patient is to recognise and treat arrhythmias that are compromising cardiac output using the safest means possible so that longer-term management decisions can be made in consultation with a paediatric cardiologist.

Normal conduction system

The normal heartbeat is initiated by an impulse which originates from the sinoatrial (SA) node located in the wall of the right atrium near the superior vena cava junction. The impulse is then propagated via conducting cells that form a specialised system throughout the heart. Initially the impulse travels across the atria and via transatrial internodal pathways that converge on the atrioventricular (AV) node. The impulse proceeds down the bundle of His to the right and left bundle branches; the impulse then finally spreads throughout the Purkinje fibres to depolarise the ventricular muscle. The conducting cells of this specialised system have a rapid conduction velocity to rapidly propagate the electrical impulse throughout the heart. The various parts of this conduction system are also capable of spontaneous depolarisation and impulse generation under abnormal conditions.

The SA node is normally the dominant (fastest) cardiac pacemaker, but this can change if sinus node dysfunction occurs or if other parts of the conduction system develop increased automaticity. As stated above, the SA node is located near the junction of the superior vena cava (SVC) and the right atrium. The SA node is innervated by both sympathetic and parasympathetic nerve endings. Parasympathetic tone predominates during rest. Children are known to have relatively greater parasympathetic tone than adults and are also known to have developmental and age-dependent differences in action potential amplitude and conduction speed. Accordingly, there are age-dependent normal values for

resting heart rate as well as PR interval and QRS duration, in addition to many other electrophysiology parameters (Table 5.10.1).

Depolarisation initiated in the SA node spreads rapidly through the internodal pathways to converge on the AV node. Usually, the atria and ventricles are electrically separated from one another by a ring of fibrous tissue at the atrioventricular junction (the annulus fibrosis). Accordingly, in usual circumstances, the impulse must pass to the bundle of His via the AV node. The AV node is located in the inferomedial wall of the right atrium near the insertion of the septal leaflet of the tricuspid valve. Conduction through the AV node is slow to allow completion of atrial systole (and ventricular diastole). This delay (and therefore potentially the completeness of ventricular filling) can be reduced during sympathetic stimulation.

After leaving the AV node the impulse travels along the bundle of His for 1–2 cm along the posteroinferior margin of the membranous portion of the interventricular septum, before dividing into the right and left bundle branches. These extensions of the conducting system spread subendocardially across the ventricular chambers to the base of the papillary muscles. The left bundle separates into two distinct fascicles (anterior and posterior). From these specialised fibres the impulse spreads from endocardium to epicardium throughout the right and left ventricles.

Abnormally situated embryonic remnants of conducting tissue can persist as accessory tracts. These are most commonly found around the AV node and are capable of conducting electrical activity between the atria and ventricles thus bypassing the AV node. These remnants are the anatomical substrate for re-entrant tachyarrhythmias.

The cardiac action potential

The cardiac action potential is divided into five phases: 0, 1, 2, 3, 4:

Table 5.10.1

Normal resting heart rates, PR intervals and QRS durations in children

Age	Heart rate mean (beats min^{-1})	Range	PR interval (ms)	QRS duration (ms)
<1 day	119	94–145	70–120	50–84
1–7 days	133	100–175	70–120	40–79
7–30 days	163	115–190	70–120	40–73
1–3 months	154	124–190	70–130	50–80
3–6 months	140	111–179	70–130	60–80
6–12 months	140	112–177	80–130	50–80
1–3 years	126	98–163	80–150	50–80
3–5 years	98	65–132	90–150	60–84
5–8 years	96	70–115	100–160	50–80
8–12 years	79	55–107	100–170	50–84
12–16 years	75	55–102	110–160	40–80

From Liebman 1982.

Adapted from Liebman J. Pediatric electrocardiography. 1982. Williams & Wilkins

Phase 0 is depolarisation due to voltage-gated opening of sodium channels with sodium rushing into the cell. There is an intense entry of sodium for a brief period resulting in depolarisation of the entire cell as well as cell-to-cell propagation.

Phase 1 is a phase of partial repolarisation due to several factors but including chloride entry into and possibly potassium egress from the cell.

Phase 2 is the plateau phase thought to be important in coordinated and sustained ventricular contraction. During this phase there is slow inward calcium current and slow inward sodium current balanced by a gradually increasing outward potassium movement.

Phase 3 is repolarisation, and it occurs by inactivation or closure of the slow calcium and sodium channels and then a voltage-gated progressive opening of potassium channels leading to outward potassium ion movement and a progressively more negative membrane potential. Also during this phase, the fast sodium channels are reactivated, and the cell is primed for further depolarisation.

During *phase 4*, an energy-dependent membrane Na–K ATPase removes the sodium and restores the potassium to the cell. Most myocardial cells maintain a constant level of depolarisation during this phase, but Purkinje or conducting cells, by way of reduced outward potassium flow and some inward sodium flow, achieve slow depolarisation until the AP threshold is reached. These conducting cells have a shorter duration action potential of lower amplitude, thought to be mediated more predominantly by slow calcium flux.

Vaughan Williams antiarrhythmia drug classification

Antiarrhythmic drugs are classified into five classes in the Vaughan Williams classification by the mechanism or channel that they most reliably affect during in vitro studies. There is, however, considerable overlap.

Class 1 drugs block the fast inward sodium channels and thereby increase the refractory period. This class is further subdivided into:

- 1a – those that prolong action potential duration (e.g. quinidine)
- 1b – those that do not change or shorten action potential duration (e.g. lignocaine [lidocaine]) or
- 1c – those that produce some mild action potential prolongation (e.g. flecainide).

These agents are infrequently used in paediatrics, though lignocaine is an option in the ventricular fibrillation protocol, and flecainide may sometimes be used in supraventricular tachyarrhythmias.

Class 2 agents are β-blockers, which act by a combination of β-antagonism and a quinidine-like membrane stabilising effect. There is a slowing of conduction velocity, prolonging of action potential duration and a reduction of automaticity. Their main clinical use is in treating supraventricular tachyarrhythmias (SVT, atrial fibrillation, atrial flutter) by increasing the refractory period of the AV node. Propranolol or atenolol may be used, but in acute situations many clinicians favour esmolol, a very short-acting parenteral β-1-antagonist (half-life 9 minutes), which can be useful in SVT and possibly ventricular tachycardia (VT).

Class 3 drugs act primarily as potassium channel blockers and basically prolong the phase 3 repolarisation phase, thereby causing a prolongation of the action potential and increase in the effective refractory period. This class of drug is particularly useful in ventricular tachyarrhythmias and includes amiodarone and sotalol.

Amiodarone has a plethora of effects; it is primarily class 3 (i.e. potassium channel blocker) but also has sodium channel blocking, β-antagonism and calcium channel blocking effects. Oral amiodarone has unusual pharmacokinetics, with clinical effect only apparent after several days of treatment and a half-life of 3–15 weeks. Side effects are frequent and serious

(corneal photosensitivity, hyper- or hypothyroidism, pulmonary fibrosis and pro-arrhythmia). Intravenous (IV) amiodarone is used in the acute management of post-operative tachyarrhythmias (usually junctional ectopic tachycardia [JET]). In the adult ventricular fibrillation protocol, a randomised comparison between amiodarone and lignocaine found a greater chance of successful resuscitation (but not survival) with amiodarone. Therefore some authorities have recommended amiodarone as the agent of choice to help effect defibrillation after adrenaline (epinephrine). In haemodynamically stable VT the agent of choice is probably sotalol unless there is impaired ventricular function (ejection fraction <40%, signs of congestive heart failure) when amiodarone is recommended. In unstable or polymorphic VT the recommended agent depends on the initial trace. If it looks like *torsades de pointes* with the trace oscillating around the baseline (or in polymorphic VT with a normal QT interval), the clear recommendation is IV magnesium. Magnesium decreases the frequency of early after depolarisations and shortens the QT interval. If it is not *torsades-like* and the QT interval can be determined and is not prolonged, amiodarone is a second-line agent to β-blockers. If the QT interval is prolonged, then IV magnesium, followed if necessary by lignocaine, would be recommended.

Sotalol is classified as a class 3 drug, although it is a non-selective β-blocker with additional class 3 properties. It therefore combines the β-blocking effect on the SA and AV nodes with prolongation of AP duration and lengthening of refractory period elsewhere in the heart. It is valuable acutely and long term in preventing SVT of all sorts and is the agent of choice in monomorphic stable VT with apparent normal ventricular function. Like amiodarone, it does increase the QT interval and is therefore pro-arrhythmic as well as having the other risks of hypotension and bradycardia, presumably resulting from its β-blocking effects.

Class 4 agents are calcium channel blockers, and the prototype remains verapamil, which has wide-ranging effects on shortening the plateau phase and reducing contractility. Verapamil is primarily used in the adult setting because of its reduction of conduction velocity in the AV node, thereby controlling the ventricular response rate to SVT, atrial flutter or atrial fibrillation. Even in that situation it is still a second-line agent to adenosine for SVT termination in adults, provided there is good left ventricular (LV) function. However, because of serious bradycardia, hypotension and cardiac arrest in infants, it is not used in children.

Finally, **class 5** agents are a miscellaneous group and include adenosine and digoxin. Adenosine is an endogenous purine nucleoside with a very rapid half-

life (less than 5 seconds). It acts by depressing slow calcium channels and enhancing potassium conduction. Its main effects are to depress sinus node and AV node activity with a shortening of atrial refractoriness. Because of its effect in blocking AV nodal activity, adenosine is superbly suited to block re-entry phenomena, the most frequent cause of SVT in children. Series show that adenosine terminates 90–100% of SVTs, but the arrhythmia re-initiates in approximately 25%. Other atrial tachycardias like multifocal atrial tachycardia, atrial flutter or atrial fibrillation are almost always resistant to adenosine. Adenosine is given in increasing doses from 50 mcg kg^{-1} to 200 mcg kg^{-1} every 2 minutes, using a two-syringe technique so that it can be flushed rapidly into the circulation. Side effects can be facial flushing, bronchospasm or sinus arrest. Adenosine is essentially very safe, although there is a single case report of VF occurring after a dose of adenosine in a neonate with a concealed Wolff–Parkinson–White (WPW) syndrome previously treated with digoxin.

Although adenosine is unlikely to be controlling in SVTs other than re-entrant SVT, the acute injection of adenosine may transiently slow AV nodal activity and allow the flutter wave or P wave morphology to be assessed. This may be very valuable in diagnosing the specific rhythm. In a haemodynamically stable wide complex tachycardia of uncertain origin, the differential diagnosis is primarily between VT and SVT with aberrant conduction. Adenosine administration is a way of making this distinction (i.e. effective in the latter but not in the former). In the adult literature, unless SVT origin is strongly suspected, this practice is discouraged because of the potential for brief hypotension and accelerated accessory pathway conduction following adenosine injection.

Digoxin is still very commonly used parenterally, acutely, and as a maintenance medication, primarily to slow AV conduction and decrease the ventricular response to atrial dysrhythmia like flutter or fibrillation. It provides rate control and sometimes converts SVTs. There is an important caveat with digoxin–that it may shorten accessory pathway refractoriness and increase the resulting ventricular response. Therefore, it should not be used in accessory pathway dysrhythmias like WPW syndrome. Virtually any arrhythmia can arise from intoxication with digoxin. This risk is greater with hypokalaemia, and the acute treatment is IV digitalis antibody.

Pathogenesis of arrhythmias

Bradyarrhythmias

Two mechanisms are responsible for bradyarrhythmias:

1. Some form of sinus node dysfunction
2. Conduction system block.

Tachyarrhythmias

There are three fundamental mechanisms proposed for the generation of tachyarrhythmias:

1. Re-entry
2. Enhanced automaticity
3. After depolarisations (triggered arrhythmias).

Re-entry

Re-entry exists when a closed loop of specialised conducting tissue allows an electrical impulse to travel in a circular fashion and permits atrial or ventricular electrical activation with each pass around the circuit. Re-entry may occur on a large (macro) or small (micro) scale. Atrial flutter and ventricular fibrillation are examples of micro re-entry; paroxysmal SVT is an example of macro re-entry.

Macro re-entry usually involves the participation of an accessory conduction pathway. Accessory pathway conduction characteristics vary widely among patients. The accessory pathway in some patients conducts antegrade during sinus rhythm, whereas in others it conducts only retrograde during tachycardia. When anterograde conduction is possible down the accessory pathway the standard ECG will show pre-excitation with a short PR interval and wide QRS, including an initial delta wave resulting from the pre-excitation of the ventricle from the sinus impulse conducting through the accessory pathway before the impulse has passed through the normal conducting system. Patients with WPW usually manifest orthodromic tachycardia (forward excitation through AV node and rapid retrograde conduction via accessory pathway to create a circuit) but also (rarely) display antidromic tachycardia involving retrograde conduction up the usual atrioventricular route or via an alternative accessory connection.

SVT is the most common sustained tachyarrhythmia in children and almost always has a re-entry mechanism. Most commonly, particularly in children

younger than 12 years of age, this re-entry is caused by an accessory AV connection (resulting in AV re-entrant tachycardia). In adolescents, AV node re-entry is the mechanism in up to one-third of patients. The re-entry circuit involves tissue, most likely atrial muscle, more than several millimetres outside the compact AV node.

The commonest mode of accessory connection-mediated re-entry tachycardia is orthodromic tachycardia, with the circular movement of the electrical impulse going antegrade through the AV node and then retrograde up the accessory connection.

Enhanced automaticity

The primary pacemaker cells of the SA and AV nodes usually demonstrate spontaneous depolarisation as well as having somewhat slower action potential propagation. The cells of the conduction system have more rapid action potential propagation but are also capable of spontaneous action potential generation if the opportunity arises. These cells are, however, normally overridden and kept refractory by the dominant (faster) pacemaker of the SA node. Under certain adverse physiological conditions (e.g. hypokalaemia, hypoxia) the threshold for spontaneous automaticity for these conducting cells may be altered. This potentially creates a situation of enhanced automaticity and secondary enhanced pacemakers. One example of a tachycardia due to enhanced automaticity is multifocal atrial tachycardia.

After depolarisations

After depolarisations are due to oscillations of the membrane potential during re-polarisation that has reached the threshold membrane potential and triggered a second complete depolarisation. This process may become self-perpetuating. *Torsades-de-pointes* is the classic example of such a triggered arrhythmia.

General principles for arrhythmia management

1. Direct primary attention to assessment and correction of airway, breathing and circulation problems.
2. If patient has an acute arrhythmia with haemodynamic compromise (e.g. shock or loss of consciousness), assess whether the rhythm is fast and/or disorganised or slow and/or irregular. If the rhythm is fast or

disorganised, cardioversion or defibrillation should take priority. If slow and/or irregular, cardiopulmonary resuscitation should commence.
3. Obtain venous access and draw blood for biochemistry, specifically Na, K, Ca, Mg and blood sugar level.
4. Record a 12-lead ECG as soon as possible.

Bradyarrhythmias

Normal heart rate varies as a function of age. Pathological bradyarrhythmia may present as fatigue, light-headedness or syncope in an otherwise well child or inappropriate absence of tachycardia in a critically unwell child under stress.

Sinus bradycardia

Sinus bradycardias can be caused in children by:

- parasympathetic stimulation (e.g. suctioning)
- metabolic disturbances (e.g. hypoxia, asphyxia, hypothermia)
- poisoning (e.g. β-blocker, calcium channel blocker)
- raised intracranial pressure
- surgical injury to the sinoatrial node (e.g. following Mustard or Senning procedures for transposition, Fontan, closure of atrial septal defect or correction of total anomalous pulmonary venous drainage)
- cardiomyopathy.

Sinus node dysfunction

Sinus node dysfunction may manifest as sinus pauses (a transient interruption of normal sinus mechanism) or sinus exit block (abnormal propagation), either of which may occur with or without an associated escape rhythm and may evolve to sinus arrest.

Management

1. Treatment is required if an adequate cardiac output is not being maintained.
2. If possible, remove the cause (i.e. suction catheter).
3. Ensure adequate ventilation.

4. If life threatening, commence compressions and administer atropine 20 mcg kg^{-1} IV (minimum dose 100 mcg).
5. If this is ineffective, or bradycardia recurs, use β-adrenergic agonist or adrenaline bolus. Commence adrenaline or isoproterenol by infusion while urgent preparations are made for temporary external or transvenous cardiac pacing.

Conduction disturbances: atrioventricular block

This involves delayed or incomplete conduction through the AV node. Causes include a congenital form (maternal systemic lupus erythematosus) and an acquired form. Acquired AV block occurs in poisoning, metabolic disturbances, myocarditis, rheumatic fever, Lyme disease, fibrosis in the area of the conduction system associated with previous cardiac surgery (particularly ventricular septal defect or atrioventricular septal defect closure, tetralogy repair or aortic valve replacement) and inferior myocardial infarction:

First-degree AV block is present when the PR interval is longer than usual for age (see Table 5.10.1), but normal sinus rhythm and 1:1 AV conduction are maintained. This is a normal feature of the ECG of endocardial cushion defects. It may occur in a normal heart during parasympathetic stimulation or digoxin treatment and usually does not require treatment.

Second-degree Mobitz type I AV block (Wenckebach) is characterised by progressive prolongation of the PR interval, culminating in a single non-conducted beat. This usually is a benign normal variant but may represent a temporary pathological prolongation of the atrioventricular node refractory period.

Second-degree Mobitz type II AV block manifests as a regular intermittent failure of P wave conduction while the PR interval remains constant. This is more likely attributable to a block within the His conduction system and as such has greater potential to progress to complete (third-degree) AV block.

Third-degree atrioventricular block (or complete heart block) represents complete failure of atrial depolarisation to propagate to the ventricle. Anatomically the block may occur at AV node or at infranodal level. ECG will show complete dissociation of P waves and QRS

complex. A narrow QRS junctional escape rhythm implies a nodal block whereas a widened slower QRS escape rhythm suggests an infranodal site for the block.

Clinical features

Patients may be asymptomatic but will usually demonstrate an inadequate cardiac output, particularly in episodes of AV block associated with slower heart rate. In third-degree AV block, cannon ‘A’ waves are visible in the neck, and a slow cardiac rhythm with variable first heart sound is present on auscultation.

Management

Emergency treatment is only required if cardiac output is inadequate. Optimise ventilation and initiate β-agonist by infusion while organising temporary external/transvenous cardiac pacing. This is definitely indicated in symptomatic children with Mobitz type II second-degree AV block and third-degree AV block.

Bundle branch block

Bundle branch block patterns are unusual in paediatrics but occur when impaired conduction is present in the specialised intraventricular conduction tissue, resulting in delayed right or left ventricular depolarisation and a resulting broad aberrant QRS complex. Right bundle branch block (wide QRS with rSR pattern in right ventricular leads) may be a normal variant but also occurs in congenital heart disease (especially involving right ventricular hypertrophy), cor pulmonale and acute pulmonary embolism. Left bundle branch block (wide QRS with RR pattern in left chest leads) is associated with left ventricular (LV) strain or hypertrophy or operated congenital heart disease.

Tachyarrhythmias

There is a wide range of tachyarrhythmias. Immediate diagnosis and management are best based on the width of the QRS complex when compared to age-based normal values.

Wide complex tachyarrhythmia

The differential diagnosis of a wide or broad complex tachycardia (i.e. QRS

duration greater than normal for age, usually >0.08 seconds) is between the following:

1. VT
2. SVT with intraventricular aberrant conduction (e.g. pre-existing bundle branch block)
3. Atypical or antidromic SVT (where during the tachycardia antegrade conduction occurs down the fast accessory pathway and retrograde component goes via the AV node)
4. Atrially originated arrhythmias conducted via an accessory connection to the ventricular muscle.

VT in children usually presents as a wide complex rhythm between 120 and 220 per minute. There is usually AV dissociation, but retrograde VA conduction may occur. It may be mono- or polymorphic, sustained or non-sustained. Causes include metabolic abnormalities, poisoning, myocarditis, cardiomyopathy, ventriculotomy, ventricular tumours and congenital or acquired long QT syndrome.

Most children with VT are symptomatic with lethargy, symptoms of pulmonary congestion, poor circulation and possibly palpitations.

The other three differential diagnoses outlined above are unusual causes of a wide complex tachycardia in children. The most important differential to think about is SVT with aberrant conduction, because it should respond to vagal manoeuvres or adenosine administration. A previous ECG is of great value in determining the presence of an accessory pathway or a pre-existing bundle branch block in that these features point to SVT with aberrant conduction. Classically, VT is a wide complex tachycardia with a left access deviation and more frequently a left bundle branch block pattern, whereas a right bundle branch block pattern (with rSR in V_1) is more commonly seen in SVT. The P wave morphology is crucial, and if P waves can be distinguished, the loss of a one-to-one relationship between P wave and QRS complex is highly suggestive of VT. Sometimes, however, in VT there still may be a one-to-one ventriculoatrial relationship. The presence of fusion beats indicating an ectopic focus below the level of the AV node is also strongly suggestive of VT.

If there is strong evidence supporting a supraventricular origin and preserved LV function, vagal manoeuvres and/or adenosine administration can be tried. The caveat is that, in doing this, there is a small but important potential for

hypotension and accelerated accessory pathway conduction from the adenosine administration that has been responsible for conversion of SVT to VT or even worse on occasions.

If there are no pointers towards a supraventricular origin, or the above manoeuvres are unrewarding, the diagnosis is most likely VT.

If the patient is severely compromised, direct current cardioversion is clearly indicated. It is debated whether this is better synchronous or asynchronous, but most authorities recommend starting with synchronous cardioversion if there is a pulse, using a 1 J kg shock from a biphasic defibrillator.

If the patient has reasonable perfusion, the recommended drug depends on the appearance of the VT. If the VT is monomorphic and the patient has preserved LV function, IV sotalol is the agent of choice, with amiodarone as a second-line agent. Amiodarone is definitely the drug of choice if LV function is impaired and is thus commonly used as a first-line agent in VT when LV function status is uncertain. If the rhythm is polymorphic, there should be some further assessment of the trace. If the trace is *torsades*-like with complexes that vary in height and appear to twist around the baseline, IV magnesium is clearly recommended. If the QT interval is prolonged, IV magnesium followed probably by IV lignocaine is recommended. If the QT interval can be seen not to be prolonged, β-blockade is the primary approach, with amiodarone again the agent of choice if LV function is impaired.

Narrow complex tachyarrhythmia

A narrow complex tachycardia is defined as one with a QRS duration normal for age (approximately ≤0.08 seconds). The narrow QRS complex almost always indicates that these tachyarrhythmias are supraventricular.

SVT refers to a family of tachyarrhythmias requiring the atrium, AV node or both for their perpetuation. SVT accounts for 90% of all significant tachyarrhythmias in children. It is most useful to subclassify these tachyarrhythmias on the basis of site of origin and mechanism into primary atrial tachycardias (including multifocal atrial tachycardia (MAT), atrial fibrillation and atrial flutter), AV reciprocating, AV nodal re-entry and junctional ectopic tachycardia. Primary atrial and junctional ectopic tachycardias will be dealt with later. AV reciprocating and nodal re-entry tachycardias are the commonest forms of SVT in childhood.

AV reciprocating tachycardia occurs due to the presence of an accessory

conduction pathway setting up a re-entry circuit. In the commonest variety (*orthodromic reciprocating*), the impulse travels antegrade via the AV node to the ventricles (as usual) and then retrograde via the accessory pathway back to the atria. If the accessory pathway is capable of conducting antegrade, *antidromic reciprocating* tachycardia can occur, with the impulse travelling in the opposite direction, i.e. forward down an accessory pathway, returning retrograde via the AV node.

AV nodal re-entrant tachycardia. In this tachyarrhythmia the two conduction pathways are thought to be within or adjacent to the AV node. Beyond 5 years of age, this becomes the most common form of SVT. Classically, the P wave is buried within the QRS complex on ECG. In AV nodal re-entrant tachycardia, for long-term prophylaxis therapy digoxin is the preferred agent. However, β-blockers may be considered in resistant cases, either alone or in combination with digoxin.

The crucial step in distinguishing between the above-mentioned atrial, AV or nodal tachycardias is an identification of the P wave and its relationship to the QRS complex. This is sometimes best done with a 12-lead ECG with the paper run at 50 mm s^{-1}. If epicardial leads are available (i.e. post-operatively), an 'atrial ECG' can be obtained by connecting the two atrial pacing leads to the right arm and right leg electrodes of a 12-lead ECG. Alternatively, a specialised transoesophageal pacing wire can be introduced, like a nasogastric tube approximately to the level of the nipple and then connected to lead V1 to display an oesophagoatrial ECG in lead V1, with the remainder of the leads showing a normal surface ECG. Because it is being recorded directly, the P wave will usually appear larger than the QRS complex in the surface ECG, and by aligning simultaneously recorded surface and 'atrial' ECGs the position of the P relative to the QRS can be determined.

Atrially driven tachycardias will have a P wave preceding each QRS. If the P wave is upright in inferior leads, the origin of the tachycardia is high in the atrium, whereas the P wave axis will be negative in inferior leads if there is AV nodal re-entry or a low atrial ectopic focus. The P wave is usually absent in junctional ectopic tachyarrhythmia or VT. An abnormal P wave morphology usually indicates an origin other than the sinus node, and several morphologies will indicate MAT. If the P wave follows the QRS complex, the differential diagnosis involves AV nodal re-entry tachyarrhythmia and VT. If the P waves are dissociated from the QRS complexes, the diagnoses are most likely junctional ectopic tachycardia or VT.

Regardless of these diagnostic considerations, the approach is essentially the same. If the patient is acutely compromised, basic life support followed by attempted DC cardioversion with 1 J kg^{-1} of DC shock (under sedation if possible) and with continuous ECG monitoring is recommended. IV adenosine is an alternative if this is immediately available. If the patient has adequate perfusion, consider vagal manoeuvres/eliciting of diving reflex, and administer adenosine in increasing doses as above.

The importance of a continuous 12-lead electrocardiogram during this process cannot be over-emphasised. This is because just a few beats of sinus rhythm before reversion back to the tachyarrhythmia might well shed considerable light on the diagnosis. If the arrhythmia is resistant to adenosine, the next step is use of amiodarone and consultation with a paediatric cardiologist.

Failure of adenosine to convert such a tachyarrhythmia makes unusual diagnoses more likely. For multiple atrial ectopic foci, sotalol is effective but probably only as a bridge to radiofrequency ablation. For JET the agent of choice is amiodarone. In atrial fibrillation/flutter, rate control can be achieved using digoxin, but usually elective cardioversion is required to terminate the arrhythmia. In post-surgical cases recurrence is common and sotalol probably has the strongest demonstrated efficacy in reducing recurrences (and controlling ventricular rate) if they occur, although only as a bridge to possible cryoablation.

Wolff–Parkinson–White syndrome

WPW is present when the accessory AV connection allows ventricular pre-excitation by rapid antegrade conduction of the normal sinus impulse, thereby avoiding the normal delay in the AV node. This is present in 22–73% of cases of paediatric SVT and produces a shortened PR interval and slurred upstroke on the QRS complex (delta waves). It usually produces orthodromic reciprocating AV tachycardia, but in a small proportion of cases (approximately 10%) an antidromic reciprocating tachycardia can occur.

If WPW is known to be present (by past history or presence of delta waves on a non-tachyarrhythmia ECG) β-blockers (propranolol or atenolol) are the agents of first choice (slow conduction through AV node with little, if any, effect on accessory pathways). Digoxin has traditionally been the drug of choice for prophylaxis against SVT in WPW but has pro-arrhythmia risks, particularly in antidromic reciprocating tachycardia (decreases accessory pathway refractory period, thereby facilitating accessory connection conductance of atrial

arrhythmias), and should therefore be avoided. Sotalol combines β-blocker and class 3 antiarrhythmic actions and is effective and safe in resistant SVT. There is also some evidence supporting a role for flecainide (class 1c agent) or amiodarone (class 3 agent) as maintenance treatments in resistant cases.

Atrial flutter

Atrial flutter is an atrial tachycardia that probably propagates via an intra-atrial re-entrant pathway. It is uncommon but can occur in infancy, when it is usually not associated with structural heart disease. Beyond infancy 95% of atrial flutter is associated with structural heart disease (Mustard/Senning operation for transposition of the great arteries, Fontan, repaired total anomalous pulmonary venous return).

Clinical features may include palpitations, cardiac failure or no symptoms, dependent on the rate of ventricular response. The ECG shows a characteristic saw-tooth flutter wave at 300 beats per minute, best seen in II, III and a VF with 2:1 or 3:1 AV block.

Infants without structural heart disease usually respond to low-energy (0.5 J kg^{-1}) DC cardioversion, repeated after digoxin loading if initially unsuccessful. Atrial flutter is more resistant in the older patient who usually has associated structural cardiac disease. Amiodarone or class 1a agents have been shown to be effective. Radiofrequency catheter ablation may be valuable as a more definitive therapy.

Atrial fibrillation

Unlike in adults, atrial fibrillation (AF) is a relatively rare tachyarrhythmia in infants and children.

Clinical features may include irregular, rapid palpitations with cardiac failure if a rapid ventricular response is present. ECG reveals absence of discrete P waves with an irregularly irregular narrow, rapid ventricular complex.

Causes of AF may include atrial distension or scars, rheumatic heart disease, hyperthyroidism, hypocalcaemia, poisoning, or intrathoracic pathology.

Management should focus on removing the cause if possible. Therapy is directed toward controlling the ventricular rate (primarily using digoxin or β-blockade to slow AV nodal conduction). Synchronised DC cardioversion is effective in conversion of atrial fibrillation to sinus rhythm, but because of the

embolic risk, the procedure is avoided in children with long-standing atrial fibrillation, cardiac failure and/or enlarged atria.

Ventricular fibrillation

In confirmed ventricular fibrillation, the first priority is rapid defibrillation, with effective chest compressions. See Chapter 2.3 for management.

Role of ‘molecular autopsy’ in sudden unexplained cardiac death in the young

Recent investigations have highlighted the importance of purely arrhythmogenic causes of sudden cardiac death as a significant explanation of the 30% of young patients (<40 years) experiencing sudden cardiac death where no cause of death is identified at post mortem (so-called ‘autopsy negative’ or ‘sudden arrhythmia death syndrome’). These primary arrhythmogenic disorders include familial long QT syndrome (LQTS), catecholaminergic polymorphic ventricular tachycardia (CPVT) and Brugada syndrome plus some others. In such a situation it is most important that DNA is collected from the decedent and a ‘molecular autopsy’ conducted directed at selected candidate genes responsible for the main primary arrhythmogenic causes. Importantly, more than 90% of inherited cardiac genetic disorders are inherited as autosomal dominant, and accordingly, this molecular analysis has high potential importance for first-degree relatives. Series of such cases have produced a variable but at times considerable diagnostic yield ranging from 0% to 35%. Ideally this analysis and family investigation/counselling should occur through a specialised multidisciplinary cardiac genetic service.

Controversies

1 Controversy exists around the requirement for ongoing prophylactic antiarrhythmic therapy for infants with paroxysmal SVT when the natural history of this condition suggests fewer attacks with age beyond 12 months.

2 The use of amiodarone as antiarrhythmic agent of first choice in children with shock-resistant ventricular fibrillation has been questioned and its replacement by lignocaine suggested.

3 Recent case series have suggested that extracorporeal membrane oxygenation (ECMO) for resuscitation (or ECPR) can be safely and effectively used particularly in infants and children with primary underlying cardiac diagnoses who experience in-hospital cardiac arrest. This consideration applies particularly to individuals with refractory shock in the context of arrhythmias (usually tachyarrhythmias). In such a situation the aim is to optimise survival, recovery and neurological outcome while minimising the potential adverse effects of extracorporeal support and avoiding reliance on catecholamine infusions while allowing time for correction of metabolic derangements, both factors which may be pro-arrhyhthmic. There is currently some evidence, which admittedly is confounded by selection bias, showing potential value of ECPR in patients with cardiac surgical diagnoses, but optimal timing, technique, patient selection and use of co-interventions remain to be clarified.

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SECTION 6

Respiratory

OUTLINE

6.1

Stridor and noisy breathing

David Armstrong

ESSENTIALS

1 Stridor is a symptom of upper airway obstruction and can usually be distinguished from other respiratory noises (such as wheeze, stertor and snoring) by careful examination.

2 A detailed history and examination are paramount in diagnosing the aetiology of stridor in children.

3 The most common cause of acute stridor in children is viral laryngotracheobronchitis (croup); other infectious causes include bacterial tracheitis and epiglottitis. Other possible (non-infectious) causes include vocal cord dysfunction (VCD), anaphylaxis and inhaled foreign body.

4 Chronic stridor in children is most often due to anatomical abnormalities of the upper airway, which can occur at the level of the larynx (e.g. laryngomalacia and vocal cord paralysis), subglottis (including subglottic stenosis, web and haemangioma) and trachea (tracheomalacia and external compression from a vascular ring).

Introduction

The term *stridor* refers to a high-pitched respiratory sound that indicates upper airway obstruction.[1] The site of the obstruction can be at any level of the extra-thoracic airway. Although most commonly heard on inspiration, there may also be an expiratory component, particularly if the site of the obstruction is below

the level of the vocal cords (e.g. subglottic haemangioma).

Stridor can usually be distinguished from other respiratory noises by careful examination. For example, **wheeze** is a high-pitched, continuous sound usually heard over the chest wall on expiration. It is caused by turbulent airflow in the small- to medium-sized intra-thoracic airways. **Stertor** is a low-pitched, wet inspiratory sound, that is heard when the child is awake and indicates obstruction at the level of either the nasopharynx (e.g. adenoidal hypertrophy), oropharynx (e.g. macroglossia, tonsillar hypertrophy) or hypopharynx (e.g. tongue base mass). **Snoring** is a low-pitched inspiratory sound caused by vibration of the soft tissue of the pharynx and is heard only during sleep.[2]

Initial assessment

The presentation of a child with stridor may constitute a medical emergency due to life-threatening airway obstruction, e.g. anaphylaxis or laryngeal foreign body. It is therefore essential that emergency physicians are familiar with the most common and important presentations of stridor in children, to enable prompt diagnosis and appropriate management. As is the case for many paediatric problems, a careful history and examination will often provide the likely diagnosis and enable appropriate referral for definitive diagnosis, which often involves airway endoscopy.

History

The three most important aspects of the history are the *age* of the child at presentation, the *age of onset* and *duration* of the stridor. The physician can then determine whether the stridor is acute or chronic.

In acute stridor, the presence of associated symptoms such as fever, cough, coryza, drooling of saliva, difficulty swallowing and respiratory distress suggests an infectious cause. Alternatively, if acute stridor is accompanied by a rash, facial or peri-orbital oedema, collapse or syncope, wheeze or vomiting, anaphylaxis should be suspected. The sudden onset of stridor with marked respiratory distress in an adolescent female athlete is suggestive of vocal cord dysfunction (VCD). A history of choking on food, followed by stridor and respiratory distress may indicate the presence of a laryngeal foreign body.

For children with chronic stridor present since early in life, the age of onset of stridor should be determined as accurately as possible. Information should also

be sought as to whether the stridor is continuous or intermittent. Details of the perinatal history should be obtained, including premature delivery, whether endotracheal intubation was needed and if any underlying congenital abnormalities are present (e.g. trisomy 21).

Further details should be obtained regarding any factors that make the stridor better (e.g. sleeping, prone positioning) or worse (e.g. increased physical activity, crying, feeding). Feeding should be assessed and longitudinal weights plotted (if available), as some children with chronic stridor have an increased caloric intake due to ongoing increased work of breathing, which results in sub-optimal weight gain.[2]

Depending on the age of the child, an assessment should be made as to whether the stridor is improving or worsening over time and whether there is any accompanying dysphagia or choking, particularly on solids.

Examination

Timing of the stridor in relation to the respiratory cycle is crucial, as the causes of a predominantly inspiratory noise differ from those in which the stridor is biphasic (present on both inspiration and expiration). Signs of increased work of breathing (including nasal flaring and head bobbing in neonates, and tracheal tug, intercostal and subcostal recession in infants) indicate more severe obstruction. The skin should be examined for the presence of cutaneous haemangiomata, particularly those in a ‘beard’ distribution.

Common causes of acute stridor in children

Acute stridor is a common paediatric presentation to the emergency department. The usual cause is acute viral laryngotracheobronchitis (croup), a clinical diagnosis that is usually straightforward when a 6–36-month-old previously well child presents with fever, acute inspiratory stridor, a barking cough and dyspnoea that follows symptoms of an acute upper respiratory tract infection. Croup is discussed in detail in Chapter 6.4.

Bacterial tracheitis is relatively uncommon but can present with symptoms identical to croup, which fail to improve with standard treatment (corticosteroids ± nebulised adrenaline). Affected children are usually aged 4–6 years. High fever and a toxic appearance are typical, although some cases can present without toxic signs.[3] The diagnosis can be made during emergent intubation for

worsening respiratory distress (with evidence of marked tracheal secretions on laryngoscopy) or by planned acute airway endoscopy, which shows marked swelling of the tracheal mucosa, with a purulent exudate. Antibiotic therapy that covers the common causative bacterial pathogens (*Staphylococcus aureus*, *Streptococcus pyogenes* and *Streptococcus pneumoniae*) is the treatment of choice.

Epiglottitis is now a rare cause of acute stridor, following the introduction of universal infant immunisation against *Haemophilus influenzae* type B. A short duration of illness (12–24 hours) is typical, and the most prominent clinical features are those of a febrile and toxic appearing infant with severe upper airway obstruction, characterised by marked stridor, dyspnoea and drooling of saliva. Cough is usually minimal or absent. Epiglottitis is a medical emergency due to the high risk of complete airway obstruction. Procedures likely to upset the infant (such as examination of the throat, venepuncture and IV insertion) should not be undertaken. Examination of the airway under general anaesthetic by gas induction should be arranged immediately. Once the airway has been secured by endotracheal intubation, treatment with intravenous ceftriaxone usually results in rapid resolution of epiglottic oedema, and extubation can usually be undertaken within 24 hours.[4]

Vocal cord dysfunction (VCD) refers to the paradoxical adduction of the vocal cords on inspiration. The symptoms of VCD are inspiratory stridor, dyspnoea and a sensation of difficulty taking a full inspiration. VCD is most common in adolescent females.[5] Exercise is a common trigger, and affected individuals describe a choking sensation in the throat or upper chest on inspiration. Adolescents with exercise-triggered VCD are often misdiagnosed with asthma[6] and may present with ongoing symptoms despite high doses of anti-asthma medication. Many affected individuals are high achievers either academically or athletically and may have symptoms of anxiety.

The presentation of VCD can often be dramatic, with loud inspiratory stridor, severe agitation and marked use of accessory muscles. Emergency management involves a simple explanation of VCD and instructing the patient to switch to abdominal breathing. The patient is asked to clasp both hands together in the epigastrium and feel their hands move outward on inspiration due to descent of the diaphragm. The patient is also encouraged to consciously slow his/her breathing rate and to concentrate on expiration rather than inspiration. Finally, patients are often reassured by normal pulse oximetry.

Once the diagnosis is made and inappropriate treatments (such as inhaled

bronchodilators) stopped, breathing exercises to encourage vocal cord abduction on inspiration are almost always curative.[7,8]

Anaphylaxis (Chapter 22.5) is a severe multi-system generalised allergic reaction that often involves the respiratory system. In children, the most common triggers are foods (including cow's milk, egg, peanuts, tree nuts and seafood). The peak age of admission for anaphylaxis is the 0–4 age group; however, recent data suggest the greatest rate of increase in admissions for anaphylaxis in the past 10 years is in the 5–14-year-old age group.[9] Respiratory symptoms of anaphylaxis include inspiratory stridor, hoarseness, dyspnoea and wheeze and can develop rapidly following exposure to the relevant antigen. Early recognition and prompt administration of subcutaneous adrenaline (epinephrine) (repeated as often as necessary) are the appropriate treatments.

Aspiration of a foreign body into the airway (see Chapter 6.3) is most common in children less than 2 years of age. The diagnosis can be made easily if a history of choking followed by respiratory symptoms is obtained; however, the diagnosis is often missed if the aspiration event is not witnessed. Most airway foreign bodies lodge in the bronchi, but on rare occasions they can cause partial obstruction of the larynx.[10] Typical symptoms include cough, stridor and hoarseness. Large objects lodged in the oesophagus may also cause stridor – in these cases, children will also exhibit dysphagia and drooling of saliva.

Common causes of chronic stridor in children

Conditions causing chronic stridor in children can be characterised by the anatomical level of the abnormality – the larynx, subglottis and trachea (both extra-thoracic and intra-thoracic).

Larynx

Laryngomalacia (or floppy larynx) is the most common cause of congenital stridor. Affected infants present in the first few days or weeks of life with a high-pitched 'cog-wheel' inspiratory stridor that may be intermittent or persistent. Symptoms are typically worse when the infant is upset, feeding or in the supine position. The noise is often less prominent during sleep. There is usually no expiratory component. Marked recession at rest is a sign of severe obstruction, particularly if accompanied by poor weight gain. Definitive diagnosis is made using naso-endoscopy during spontaneous breathing, which demonstrates

prolapse of an omega-shaped epiglottis and arytenoid cartilages into the supraglottis on inspiration. Provided that the infant is thriving, parents can be reassured that the symptoms will resolve spontaneously between 1 and 2 years of age. If there are concerns regarding poor weight gain, apnoea or desaturation during sleep, referral for overnight oximetry and/or polysomnography may be appropriate.

Vocal cord paralysis (VCP) is the second most common congenital laryngeal abnormality and can be bilateral or unilateral. The most common causes are iatrogenic (43%), idiopathic (35%) and neurologic (16%).[1] Bilateral VCP presents with inspiratory stridor and a weak cry in the neonatal period. The cords are usually fixed in abduction. Unilateral VCP presents with stridor, dysphonia, feeding difficulties and aspiration. Airway endoscopy is required to confirm the diagnosis.

Subglottis

Subglottic narrowing due to a stenosis, web or cyst presents with biphasic stridor and respiratory distress. The most common cause for these lesions is iatrogenic, due to prolonged and/or repeated endotracheal intubation in a premature infant.[1] The diagnosis should be suspected in an infant with prolonged ‘croup’, particularly if aged less than 6 months with a history of premature birth and neonatal intubation. Airway endoscopy confirms the diagnosis. Surgical treatment including tracheostomy is often required.

A subglottic hemangioma is asymptomatic at birth and presents from the age of 2–8 weeks with worsening biphasic stridor and respiratory distress. Cutaneous hemangiomata are present in approximately 50% of cases. The diagnosis is made endoscopically. Airway hemangiomas increase in size until 6–12 months of age, before undergoing spontaneous involution at 18–24 months. In the past, tracheostomy was the standard management; however, all patients are now treated with propranolol, which results in rapid and sustained reduction in the size of the hemangioma, eliminating the need for tracheostomy.[11] Treatment is usually required for 6–12 months, depending on clinical response.

Trachea

Congenital tracheomalacia is caused by softness of the tracheal cartilage and typically presents at birth or within the first few weeks of life with noisy rattly

breathing.[1] This is often accompanied by a wet cough, tachypnoea and a variable degree of respiratory distress. Primary tracheomalacia is most often idiopathic but is also present in oesophageal atresia with distal fistula and Down syndrome. The diagnosis of tracheomalacia in an otherwise well child is clinical; however, airway endoscopy will confirm the diagnosis. Although symptoms always improve with time, noisy breathing (particularly with upper respiratory infections) may persist until the age of 5–6 years in severe cases.

The most common cause of secondary tracheomalacia is external compression from a vascular ring (the two most common are double aortic arch and right-sided aortic arch with aberrant left subclavian artery).[12] The diagnosis should be suspected in a child aged 6–12 months of age with an increasing biphasic stridor or wheeze, with worsening respiratory distress, who then develops dysphagia or choking, particularly with solids. A plain chest X-ray may be notable for absence of a left-sided aortic arch, and a barium swallow shows indentation of the oesophagus in both PA and lateral views. Definitive diagnosis is most accurately made by 2D echocardiography and chest MRI. Surgical division of the ring is the treatment of choice.

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6.2

Upper respiratory tract infections

Peter L.J. Barnett

ESSENTIALS

1 Over 95% of childhood upper respiratory tract infections (URTIs) are viral and do not require antibiotics.

2 URTIs are common; children have between six and eight URTIs per year.

3 In the child with presumed URTI who has significant constitutional symptomatology one needs to consider alternative diagnoses.

4 Symptomatic nasal obstruction in infants should be treated with saline drops and gentle bulb aspiration. It is uncommon for simple URTIs to compromise feeding in infants; in this situation one needs to consider lower respiratory tract involvement.

5 Stomatitis in children is usually caused by herpes simplex or coxsackie virus, and symptomatic treatment to aid fluid intake is all that is required.

6 Tonsillitis in children under 4 years is usually viral in aetiology, despite the presence of pus on the tonsils.

7 Tonsillitis is a feature of Epstein–Barr virus (EBV); thus always examine for node enlargement and organomegaly and features in the pharynx to suggest possible EBV.

Introduction

Infections involving the upper respiratory tract are the most common infections

seen in children and the most frequent reason for presenting to emergency departments (EDs). These infections may involve anatomical structures including the nasopharynx, mouth, ear and upper airway. Specific diagnosis needs to be made to decide whether antibiotic treatment is necessary. Most of these infections are mild and self-limiting, but complications can occur, leading to more serious disease.

Nasopharyngitis

Introduction

Nasopharyngitis or the common cold is a viral illness of the upper respiratory tract. This is commonly called an upper respiratory tract infection (URTI) even though it only affects part of the upper respiratory tract. A URTI is usually caused by rhino- and coronaviruses, but during the winter season, parainfluenza, respiratory syncytial virus (RSV) and metapneumovirus are also common. These later viruses may progress to croup, bronchiolitis or pneumonia. Other specific infections may begin with URTI symptoms and progress to involve the lower respiratory tract, e.g. influenza, *Bordetella pertussis*. If symptoms are persistent, an open mind regarding alternative diagnoses is required.

In their first 6 years, children generally have between six and eight URTIs per year, and children attending day care may have more. Not infrequently these may occur one after the other, leading to the impression of persistent symptoms rather than two to three separate infections. Breast-feeding of babies may offer some protection, particularly in the first 6 months.

History

This illness is characterised by mild to moderate fever, blocked or runny nose, sneezing, mild cough (generally dry in the first few days) with an irritating sore throat if the child is able to verbalise this symptom. Infants may also become restless and irritable. Nasal congestion may interfere with feeding in infants under 6 months of age due to mechanical obstruction.

Over the first few days the watery nasal discharge will become thicker and more mucopurulent. Nasal obstruction leads to mouth breathing and increased throat discomfort. The characteristic of the cough may change, becoming moist after a few days. Sputum production occurs but in small children is generally swallowed rather than expectorated and hence will not be reported by parents.

The child is not particularly unwell. If a child has significant constitutional symptoms, one needs to consider alternative diagnoses such as lower respiratory infection or an influenza-like illness. Other family members may have or be recovering from cold symptoms.

Symptoms usually peak around 1–3 days and disappear by 7–10 days, although the cough may persist (up to 8 weeks – postviral cough). Bacterial complications are uncommon but should be considered if fever persists for longer than 3–5 days, there is ongoing mucopurulent nasal discharge for more than 2 weeks or the child appears unwell or has lower respiratory tract signs (e.g. tachypnoea, grunting, wheeze, O_2 requirement, focal signs).

Examination

The child appears well with nasal discharge, which may cause obstruction (particularly in babies). Mucopurulent discharge does not indicate a bacterial cause. The tympanic membranes may be slightly dull or pink in appearance with no evidence of fluid in the middle ear. The throat may be red but not associated with exudate or cervical lymphadenopathy. The chest is clear to auscultation, although there may be transmitted upper airway sounds originating from the nasal passages. The cough may be dry or moist depending on the length of symptoms.

Investigations

There is no indication for investigations in a child with the common cold.

Treatment

Therapy is supportive, with explanation and a management plan for parents. Parents need to ensure the child has adequate rest and fluids to maintain hydration. Nasal congestion/obstruction in infants may be improved with saline drops (one teaspoon salt in one cup boiled water and allowed to cool and kept in the refrigerator for a few days only or commercial saline preparations) into the nares prior to feeds or sleeping. In addition, aspiration of mucus using a bulb suction device is generally helpful. While feeding difficulty may occur due to nasal obstruction, infants with a presumed simple URTI who are unable to take normal feeds need to be carefully checked for features of lower respiratory tract

involvement.

The routine use of oral decongestants in infants/children is generally unhelpful and may cause side effects.

There is no indication for antibiotics in this setting. Pharyngitis or tonsillitis associated with a URTI is viral in nature and will not respond to antibiotics. The inappropriate use of antibiotics in these patients may be a contributory factor to antibiotic resistance. Herbal remedies such as echinacea, vitamin C, or zinc have not been shown to benefit resolution of symptoms. Honey may be of benefit for the cough, but it is not recommended in children less than 12 months of age because of the small risk of infant botulism.

An explanation to parents of the expected natural history may decrease the likelihood of inappropriate seeking of antibiotics. Early review should be encouraged if their child's course deviates from that expected. Paracetamol is indicated if a child is symptomatic of fever (e.g. lethargy, very unsettled, etc.) or to diminish the discomfort of a sore throat, not simply for fever control alone.

Stomatitis

Introduction

Children with stomatitis often present to emergency due to difficulties in drinking. In children it is most commonly caused by herpes simplex virus (gingivostomatitis) or Coxsackie virus (e.g. hand, foot and mouth [HFM]). These viral infections cause vesicular lesions, which may involve the buccal mucosa, gingiva, tongue, palate and pharynx. Fungal infection with *Candida albicans* (thrush) may be seen in neonates or immunosuppressed children.

History

Primary herpes simplex produces a severe gingivostomatitis involving most of the mouth and characteristically has a few lesions on the outer lip area. Fever is prominent and can last for 7–10 days. The associated mouth pain can result in drooling, decreased oral intake and subsequent dehydration. Oral herpes can be spread to the digits in patients who suck their fingers, leading to a herpetic whitlow. Coxsackie virus can produce ulcers involving the oral mucosa and skin areas (hands, feet and buttock areas). Thrush, generally seen in infants under 3 months of age, involves the tongue and buccal mucosa. It can cause feeding difficulties if severe, but other problems need to be considered if mild thrush is

associated with feeding issues.

Examination

Children may be distressed by mouth pain and have a high fever. Hydration should be assessed. Herpes causes multiple shallow ulcers of the oral mucosa and associated gingival inflammation. Hence, the oral features of herpes simplex virus infection tend to be more anterior on the gums, inner lips and tongue initially and progress in a posterior direction. Coxsackie tends to cause fewer ulcers in the mouth and more on the palatal mucosa and is less likely to cause gingivitis. It is classically associated with vesicular lesions on the hands, feet and buttocks (HFM). Isolated pharyngeal ulcers with high fever are also typical of Coxsackie virus. Oral thrush presents with characteristic white plaques on the buccal mucosa and tongue. Clinical confusion may arise in the baby who has recently taken milk, which may give a similar appearance. Differentiation can be made by trying to scrape off the plaques, which either causes bleeding or they are unable to be removed.

Investigation

Diagnosis is clinical and usually requires no investigations. Immunofluorescence for herpes simplex may be required for isolation purposes or in the immunosuppressed.

Treatment

Viral stomatitis is self-limiting, and symptomatic treatment to reduce pain and allow eating and drinking is the mainstay of therapy. Topical anaesthetic treatment such as viscous lignocaine (lidocaine) or gel (2%) applied sparingly to the lesions, half an hour prior to drinking, may be beneficial, although a recent study disputes this. In addition, oral analgesics such as paracetamol ± ibuprofen should be used regularly. Icy poles (ice lollies), cool drinks or a soft diet may also be an effective way to achieve an adequate fluid intake.

Frequent review is required to assess a patient’s hydration status. The child with severe stomatitis, who is dehydrated, may require admission for intravenous fluids and consideration of IV acyclovir. Oral aciclovir has been used to hasten the resolution by 24 hours if started within 72 hours of onset but does not

diminish recurrences, and is not currently on PBS for this reason. Treatment for oral thrush is either topical antifungal gel (e.g. miconazole) or nystatin oral drops four times a day for 10 to 14 days. Drops should be applied after a feed and inserted onto the buccal mucosa, not into the general oral cavity. Systemic antifungals are only indicated in immunocompromised patients.

Pharyngitis/tonsillitis

Introduction

Pharyngitis/tonsillitis is a common infection involving the throat, including the tonsils. In children, particularly in the first 4 years, a viral aetiology is most likely. Viruses that can cause pharyngitis include adenovirus, enterovirus, parainfluenza, or Epstein–Barr virus (EBV).

Group A *Streptococcus* is the most common bacterial infection of the throat but accounts for only up to 20% of tonsillopharyngitis. The incidence is age related and is uncommon in children under 4 years of age. Twenty percent of children are colonised by group A *Streptococcus*; hence, a positive throat swab may be incidental in a child with an acute viral infection. The clinical dilemma is distinguishing viral from acute bacterial infections.

History

Older children may complain of sore throat, headache or dysphagia or have referred abdominal pain. Younger children may present with less localising symptomatology with fever, decreased oral intake, drooling or clinging to parents. Patients presenting with associated coryzal symptoms (e.g. cough, nasal congestion, etc.) are unlikely to have a bacterial cause for their pharyngitis. In high-risk children (e.g. indigenous), one needs to consider the potential of post-streptococcal glomerulonephritis as a sequela.

Examination

Group A *Streptococcus* is most likely in the older child with fever, swollen tender cervical nodes, isolated sore throat and florid exudative tonsillopharyngitis. The presence of a scarlatiniform rash (red, sandpaper-textured skin) or a strawberry tongue is supportive of streptococcal infection.

Table 6.2.1

Features of viral and bacterial tonsillitis

Viral	Bacterial
Age <4 years	Age >4 years
Associated cough, coryza, conjunctivitis, diarrhoea	Tender cervical lymphadenopathy
Lymphadenopathy, splenomegaly (e.g. Epstein–Barr virus [EBV])	Scarlatiniform rash, oedematous tonsils or exudates
Oropharyngeal features of EBV	Absence of cough
Ulcerative lesion on palate/tonsils (Coxsackie)	

Enterovirus can cause, in infants/toddlers, an ulcerative or exudative pharyngitis associated with high fever and drooling and may be associated with vomiting/diarrhoea or a fine macular rash.

EBV (infectious mononucleosis) is common in adolescents and can have a similar pharyngeal appearance to bacterial infection. The patient may have general swelling of the neck and face due to marked cervical lymphadenopathy and enlargement of the spleen and liver (Table 6.2.1).

Investigations

The use of throat swabs in the ED remains controversial due to the *Streptococcus* A colonisation that occurs in up to 20% of children and since treatment with antibiotics only decreases symptoms by 1 day. A rapid strep test is very specific for *Streptococcus* A infection but not very sensitive. If a rapid strep test is positive, it obviates the need for further testing.

If one suspects EBV, a full blood examination (atypical lymphocytes) or monospot or EBV serology may be helpful, although the early monospot (within 1 week of onset of symptoms) can be falsely negative, particularly in younger children.

Treatment

Treatment of confirmed group A streptococcal infection with antibiotics will decrease the length of symptoms approximately by 1 day (3 vs. 4 days) and decreases the likelihood of rheumatic fever. Oral penicillin is the drug of choice, either 250 mg bd (age <10 years) or 500 mg bd (age >10 years) given for 10 days. A long-acting parenteral penicillin (benzathine benzylpenicillin) is also

effective in children who are refusing oral medication.

Most children with a sore throat have a viral infection and do not require antibiotics. Symptomatic relief using paracetamol or soluble aspirin/salt-water gargles may be helpful. Topical anaesthetic (gels or sprays) and oral steroids have been shown to decrease the throat pain. Ensuring adequate hydration improves the patient's general well-being. In those unable to drink and who are dehydrated, admission for intravenous fluids may be required. Children with severe tonsillomegaly and neck swelling from EBV can develop airway obstruction and may benefit from steroids. The threshold for antibiotic treatment needs to be lower in high-risk children (indigenous) who have an increased risk of glomerulonephritis.

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6.3

Inhaled foreign body

Peter L.J. Barnett

ESSENTIALS

1 The presentation to an emergency department (ED) of a child with airway obstruction due to an inhaled foreign body can be life threatening and requires rapid assessment.

2 One needs to have a prepared approach that differentiates the situation of total from partial obstruction.

3 One needs to be cautious to avoid the potential to convert partial upper airway obstruction to complete obstruction by unnecessary interventions.

4 Many children who were initially symptomatic may be asymptomatic by the time they are seen in the ED and have minimal clinical findings.

5 The majority of objects are radiolucent, and the radiological findings are those secondary to the foreign body's physical presence in the airway.

6 Indication for bronchoscopy should be based on history, examination and radiological investigations. When in doubt, consult a paediatric respiratory physician.

Introduction

Foreign body (FB) aspiration usually occurs in children less than 3 years of age. Exceptions to this can exist, particularly in older children with developmental or

neurological issues. FBs may lodge at any place along the airway from the hypopharynx to the segmental bronchus. Upper airway obstruction due to FBs causes a small but significant number of deaths each year. The reason toddlers are more prone to aspiration is due to their general inquisitive nature, the presence of small food or non-food objects in the general home environment and their inability to efficiently grind food objects (due to lack of molar teeth).

The most common inhaled FBs are food materials (e.g. peanuts, carrot or apple pieces) and small pieces of toys. Inhaled FBs are rarely radio-opaque and therefore often not visible on a plain X-ray. FBs in the upper airway account for only 5–10% of all inhaled objects but have a higher mortality and morbidity due to their potential to cause high-grade obstruction of the airway proximal to the carina.

Upper airway foreign bodies

History

The presentation of a child with upper airway obstruction due to an inhaled FB can be life threatening and require a rapid careful assessment and management. These children present with acute onset of upper airway obstruction – which may be partial, total or fluctuating in severity. Oesophageal FBs can cause upper airway obstruction due to adjacent airway compression, usually characterised by partial obstruction with significant drooling and saliva intolerance.

There is generally no preceding history suggesting an infective cause such as croup or epiglottitis or allergic phenomena. Occasionally, the aspiration incident is witnessed as a choking episode, but often not, and a child is only discovered with breathing difficulty following an unwitnessed event.

Examination

A foreign body in the upper airway presents with acute signs of obstruction and respiratory distress. The signs reflect the degree of airway obstruction. In life-threatening cases there may be respiratory arrest with apnoea and cyanosis. Less urgent cases may have stridor, wheeze, cough and abnormal voice or cry. The child may be drooling.

Investigations

In a stable child with partial obstruction, a soft tissue X-ray may be useful to localise the FB, if radio-opaque. This is best performed portably in the emergency department (ED) so that the airway is clinically monitored, with the ability to intervene if necessary.

Radiographs should not be attempted in an unstable child with airway obstruction, due to the potential to cause distress, which may worsen the obstruction or precipitate complete airway obstruction and respiratory arrest.

Treatment

Total obstruction

Untreated, *total* airway obstruction will rapidly lead to hypoxia, loss of consciousness and subsequently cardiorespiratory arrest. In the child presenting with complete obstruction, basic life support should be commenced (see Chapters 2.2 to 2.4). The performance of back blows and chest thrusts to dislodge the FB should be followed by attempted ventilation with bag and mask. If these methods are unable to remove the FB and allow ventilation, then direct visualisation of the larynx and Magill forceps extraction of the foreign body should occur if possible. In the unlikely event of an FB visualised below the cords, suctioning may expedite its removal, or, if not possible, advancing the object past the carina with an endotracheal tube or bougie may allow life-saving ventilation as a temporising measure. A surgical airway (Chapter 24.5) may be required if unable to intubate or ventilate the patient.

Partial obstruction

The approach to the child with partial airway obstruction, but who is able to ventilate him/herself to maintain adequate oxygenation, needs to be a cautious one. One needs to avoid the potential to convert the situation to one of complete obstruction by unnecessary interventions. It is best to leave the child undisturbed and as comfortable as possible. Small children are best kept in a parent’s arms. Oxygen should be administered if required and tolerated.

Causing distress and crying will usually worsen the degree of airway obstruction. The child needs to be carefully transferred from the ED to an appropriate environment where the object can be safely removed with anaesthetic, endoscopic and surgical facilities. DO NOT attempt to remove the object by the methods mentioned above as this may convert the partial

obstruction to complete obstruction.

Lower airway foreign body

History

The majority (70%) of patients presenting with an inhaled foreign body have a clear history to suggest an inhalation. However, up to 30% have a delayed presentation and/or no history of FB aspiration.

Patients generally present with sudden onset of coughing and choking, occasionally associated with a brief period of cyanosis. After the initial coughing episode, the child may have an audible wheeze and/or a persistent dry cough. However, many children who were initially symptomatic may be asymptomatic by the time they are seen in the ED. Depending on the size of the FB the symptoms may be mild. FBs tend to lodge in the right main stem bronchus more than the left, due to its less acute deviation from the lumen of the trachea. A patient may also suddenly deteriorate as the FB is coughed from one bronchus to the other, causing an increasing degree of lower airway obstruction.

There may be a history of persistent cough or ‘chestiness’ after a remote choking event. Recurrent lower respiratory tract infections with radiological changes in the same area may be an indication of a retained FB – thus a careful history is needed, particularly for the start of the illness.

Examination

Examination usually reveals a well child with mild to moderate respiratory distress. The patient may have a cough. Auscultation may reveal diminished air entry on one side compared to the other with or without unilateral wheeze. In delayed presentations, there may be a fever and signs of collapse or consolidation if an FB has resulted in a segmental infection. However, in some cases the inhaled object will be occult and the chest examination may be completely normal. A high index of suspicion is required, particularly if the history is suggestive.

Investigations

The initial investigation of the child with suspected inhaled FB should include a plain chest X-ray. The majority of objects are radiolucent, and the radiological

findings are those secondary to the physical presence of the FB in the airway. This may be normal or may show signs of unilateral hyperinflation.

Good-quality inspiratory and expiratory or right and left decubitus X-rays are helpful. If there is air trapping, then the affected lung or part of the lung remains inflated in expiration or on lying on that side (e.g. left side down). There may be segmental or lobar collapse distal to the FB, particularly with delayed presentation. Fluoroscopy, if available, shows decreased movement of the diaphragm on the affected side.

Persistent changes on X-ray or recurrent pneumonia in the same place (particularly the lower lobes) may signal an occult FB.

Treatment

Initially, the patient's airway and breathing should be assessed and oxygen given if required. Referral for bronchoscopy is the definitive treatment for confirmed or suspicious cases. Indications for bronchoscopy should be based on history, examination and radiological investigations '2 of 3 rule' (Box 6.3.1).

Patients should then be referred to an appropriate service that can perform a rigid bronchoscopy to remove the FB. When in doubt, consult with a paediatric respiratory consultant.

Children who have had a minor choking episode but no ongoing symptoms or signs and a normal X-ray can be discharged. Parents need to be instructed to return should any relevant symptoms evolve (wheeze, persistent dry cough, etc.).

If a child re-presents with ongoing symptoms after previous referral to a tertiary centre, clarify that an adequate assessment was made to rule out an FB at that review.

Box 6.3.1 Indications for bronchoscopy

Two of the following:

History
- Coughing and choking episode and cyanosis or persistent cough after choking episode

Examination
- Unilateral wheeze or unilateral decreased air entry

Investigations
- Hyperinflation on expiratory chest X-ray or subsegmental

atelectasis or positive fluoroscopy

Prevention

Children under 3 years should be protected from access to toys with small parts or foods requiring molar function, as they are potential inhalation risks (e.g. peanuts, hard foods and fruits with skin on).

Further reading

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6.4

Croup

Gary Geelhoed

ESSENTIALS

1 Croup is a common childhood problem with a peak incidence of 60 per 1000 child years in those aged between 1 and 2 years.

2 The loudness of stridor is variable and is not a reliable indicator of the severity of the airway obstruction.

3 All children who present to emergency departments with croup should be treated with steroids.

4 The steroids of choice are oral dexamethasone 0.15 mg kg^{-1} or prednisolone 1 mg kg^{-1}.

5 A compromised but functioning airway should never be made worse by upsetting the child.

6 Children who require adrenaline (epinephrine) may be sent home safely if they have also received steroids and have improved over a number of hours to have no stridor at rest.

Introduction

The term *croup* describes an acute clinical syndrome of hoarse voice, barking cough and inspiratory stridor usually seen in young children. Croup results from swelling of the upper airway, in and around the larynx, usually as a result of a viral infection. Croup occurs seasonally, peaking in winter months due to the epidemics of upper respiratory viruses. Parainfluenza virus type 1 accounts for around half the cases during winter, with parainfluenza type 2, influenza type A,

adenoviruses, respiratory syncytial virus, enteroviruses, and possibly *Mycoplasma pneumoniae* causing most of the other cases. Some of the viral exanthems, such as varicella, can cause concomitant croup by involvement of the upper airway in small children. Croup is a common childhood problem, with a peak incidence of 60 per 1000 child years in those aged between 1 and 2 years, although it may be seen up to the teen years. As such, it is by far the most common cause of acute upper airway obstruction likely to present to emergency departments (EDs).

The respiratory distress caused by obstruction tends to be most marked in younger children due to the small size of their larynx, the presence of loose submucous tissues, and the tight encirclement of the subglottic area by the cricoid cartilage. In children under 8 years of age, this is the narrowest region of the airway, hence any inflammatory swelling in this area results in a significant impingement of the airway. The younger child, who has a smaller diameter airway, requires an increased vigilance to assess the degree of airway compromise.

The lower airway involvement of laryngotracheobronchitis may also cause younger children to manifest wheeze due to concurrent inflammation producing mucus in the smaller peripheral airways. Likewise, occasionally older children known to have asthma may exhibit signs of asthma in the setting of croup.[1]

Presentation

History

The typical presentation of croup is in a preschool-aged child with a history of a recent onset of upper respiratory tract infection. The child subsequently develops a barking or seal-like cough, a hoarse voice and, if obstruction is severe enough, stridor. The stridor may initially be apparent only when a child is distressed, such as during crying. During crying or forced expiration the diameter of the upper airways physiologically narrows and, hence, stridor will manifest. Stridor, which is initially inspiratory, indicates obstruction at the laryngeal level or higher (i.e. upper airway). Expiratory stridor or biphasic stridor indicates more severe laryngeal obstruction or alternatively an obstruction occurring lower in the airway. The natural history of airway obstruction, when unmodified by steroids, is to increase slowly to peak over 24–48 hours. The airway compromise usually then resolves over a few days, although the laryngeal cough may persist

longer.

Less common than infectious croup, but usually more sudden in onset, older children may present with *recurrent or spasmodic croup* with no viral prodrome. This is thought to be allergic in origin. These children may have a history of atopy and suffer from asthma more than the general population. They should, however, be treated in the same manner as 'viral' croup. In the smaller child, particularly infants, problems with feeding, swallowing difficulties and whether the child has been cyanosed should be ascertained.

It is important to enquire whether or not the child has had croup or other airway problems in the past and, specifically, whether the child has had any persistence of mild stridor in between acute attacks. This is important, as any child who has a pre-existing narrowing of the airway (infantile floppy larynx, laryngomalacia or other upper airway anatomical abnormalities) is more likely to proceed to severe obstruction with a superimposed acute obstruction. These children need to have a lower threshold for a period of observation as their obstruction may be more severe or persistent.

An immunisation history is important to check whether the child has had Hib vaccination if there is any suggestion that the condition could be epiglottitis or, likewise, the very rare occurrence of diphtheria in the non-immunised.

Examination

Croup in children can generally be classified as mild, moderate or severe (Table 6.4.1).

Most children with mild croup are not distressed and have only a barking cough with no stridor at rest or stridor audible only with physical activity, crying, or agitation. Crying causes physiological narrowing of the airway and will increase the respiratory distress. Hence the distressed, crying child's obstruction will often defervesce by allowing the child to be cuddled in the parent's arms. There may be signs due to viral illness, such as mild fever and nasal discharge. Children with mild cases can have their throats examined, but this should be deferred in more severe cases. A compromised but functioning airway should never be made worse by upsetting the child.

In more severe cases, the child may have a more pronounced stridor at rest. As airway obstruction progresses, increased work of breathing ensues and the child exhibits increasing substernal, intercostal, and subcostal retractions. Subtle signs of hypoxia causing altered consciousness may be reflected as anxiety or

restlessness in a child. An obviously fatiguing child is a worrying sign. The child manifesting decreased air entry and respiratory effort, extreme pallor and cyanosis requires immediate intervention.

The child's preferred position may also give clues as to the severity of obstruction or to a diagnosis other than croup. Hyperextension or other abnormal positioning of the neck may suggest epiglottitis or a retropharyngeal abscess. It is unusual for the child with croup to be saliva intolerant or have any tenderness of the neck.

The use of a croup severity score may be helpful for less experienced staff to assess children with croup and communicate findings with a colleague when seeking advice. As previously mentioned, some children may have concomitant wheeze in addition to the upper airway stridor, and hence their pattern of breathing may be 'gas trapped' and cause hyperinflation of the chest with a slow expiration phase, in contrast to those with pure croup alone.

Investigations

Croup is usually an easy 'spot diagnosis' and requires no diagnostic tests.

Oximetry is of limited value, as children may maintain near-normal oxygen saturations even when they have significant airway obstruction.[3] While it may have a role in cases of severe croup, this must be balanced with the distress caused by the monitoring probe in small children. In stable cases, where the diagnosis is unclear, a lateral soft tissue X-ray of the neck may be helpful to distinguish croup from epiglottitis or retropharyngeal abscess. However, the possible benefits of an X-ray need to be weighed against the risks of moving or disturbing the child when the obstruction is more than mild.

Table 6.4.1

Croup severity

Sign or symptom	Mild	Moderate or severe
Stridor	None or only if agitated	Stridor at rest
Respiratory rate	Usually normal	May be decreased
Retractions	None	+ to +++
Air entry	Normal	Normal to decreased
Colour	Normal	May be pallor
Cyanosis	None	Late sign only

Conscious state	Normal	Restless or decreased

After Marks et al. 2003.[2]

Differential diagnosis

It is important to establish that other, more sinister, causes of acute upper respiratory tract obstruction masquerading as croup are not present (Box 6.4.1 and Chapter 6.1). Especially in the younger child, one should enquire regarding longer term symptoms preceding the present episode, such as low-grade stridor. This might suggest underlying congenital airway or vascular anomaly (e.g. tracheomalacia, subglottic stenosis, bilateral cord paralysis, laryngeal web, or vascular ring compression of the trachea). One should also enquire as to possible airway trauma or toxic ingestion. Dysphagia and drooling may suggest epiglottitis, peritonsillar or retropharyngeal abscess or foreign body in the airway or oesophagus.

Classic croup and epiglottitis are hard to confuse, as the latter usually presents as a pale, toxic, drooling child with a rapidly progressing course. Cough is generally not a prominent feature in epiglottitis. Children with epiglottitis may sit forward, drooling saliva and holding their neck in extension. In a child presenting with early epiglottitis, however, the distinction may be more difficult. Immunisation in developed countries has made this distinction largely academic.

Allergic angio-oedema may mimic croup after exposure to an allergen such as peanut. A child with ‘severe croup’ with a high fever, who does not respond to adrenaline (epinephrine) and steroids, may have tracheitis and will need consideration of a diagnostic laryngoscopy to provide a clear diagnosis.

Likewise, the possibility of an inhaled foreign body should be kept in mind for children who don’t respond to treatment or have a prolonged course. While usually parents will volunteer a history of an acute obstruction or a sudden coughing fit, the history of an inhaled object may not always have been observed and therefore reported. A definitive diagnosis may need to be made by directly viewing the upper airway, but this should be performed only by an experienced paediatric anaesthetist, intensivist, or emergency physician in an appropriate clinical setting (see below).

Box 6.4.1 Differential diagnoses of croup

- Epiglottitis
- Bacterial tracheitis
- Foreign body
- Congenital: laryngomalacia, subglottic stenosis, vascular ring, cord paresis
- Retropharyngeal abscess
- Allergic oedema
- Airway trauma

Treatment and disposition

Mild or moderate croup

All children who present to an ED with croup should be treated with steroids.[4] The mandatory use of steroids for croup in EDs results in a reduction in the relapse rate of those sent home,[5] a decrease in the average length of stay in hospital and a dramatic reduction in the number of children needing intensive care and intubation[6,7] Prior to the regular use of steroids, a general rule of thumb was to admit children with stridor at rest (moderate) to hospital for observation, while allowing those with occasional stridor and barking cough only (mild) to be managed at home. As many children will improve within a few hours of taking steroids, often they may be discharged home after a short stay in the ED or an observation ward. Factors such as the distance from medical care, the availability of transport, the time of day, the child's past history with regard to severe airway obstruction and parental concern and attitude all need to be taken into account when making the decision to admit (Box 6.4.2).

Recommended steroid doses are oral dexamethasone in a one-off dose of 0.15 mg kg^{-1} [8] or an equivalent dose of prednisolone of 0.75 mg kg^{-1}. Most children with croup will require only one dose, but if the upper airway obstruction symptoms persist (as opposed to upper respiratory tract infection symptoms), a further dose may be given 18 to 24 hours later. It is often more convenient to use prednisolone (rounded off to 1 mg kg^{-1}) in the community, as it is more readily available. While one study[9] suggested that children treated with prednisolone may re-present more commonly than those treated with dexamethasone, Fifoot et al.[10] did not confirm this finding. Steroids may be administered

intramuscularly or intravenously in the child with severe obstruction, when there is concern that the child may aspirate or vomit, given the degree of respiratory difficulty. There is little evidence to inform parenteral dosing of dexamethasone, but it is usually given in the range of 0.15–0.6 mg kg^{-1}.

Box 6.4.2 Possible indications for admission

- Upper airway obstruction at rest: fails to respond to steroids or adrenaline (epinephrine) and period of observation (4 hours)
- Severe croup or moderate croup that does not respond to treatment
- Infants <6 months
- Previous severe croup
- Underlying airway problem
- Inadequate social factors, i.e. transport or lack of follow-up
- Recurrent presentation

Oral dexamethasone has been found to be as effective as inhaled steroids such as budesonide[11] and to work as fast at a fraction of the cost. A blinded randomised trial of dexamethasone 0.15 mg kg^{-1} compared to placebo that looked at croup scores at 10-minute intervals after administration showed a significant difference at 30 minutes.[12] Combining dexamethasone and budesonide is no more effective than dexamethasone alone.[11,13] Fernandes et al., in a wide-ranging review of the use of systemic corticosteroids for acute respiratory conditions in children, found no evidence of harm compared with placebo.[14] There is no place for antibiotics in a typical case of croup. The use of steam or humidified air is unproven,[15,16] despite its once common usage. The anecdotal report by parents of their child improving in the steam-filled bathroom at home is due to the defervescing of crying that occurs from cuddling in the room by the parent rather than any steam effect.

Severe croup

Children with manifestations of severe obstruction should be given nebulised adrenaline. It is generally considered that adrenaline does not change the natural history of croup, such as length of stay in hospital or need to intubate, due to its

short-lasting effects. It will, however, buy time while waiting for the effect of steroids to occur. Rarely, in a worst-case scenario, adrenaline can be a useful temporising measure while organising the facilities and appropriate personnel for a child who may require intubation. The recommended dose (independent of age and weight) is 5 mL of 1:1000 adrenaline (1 mg per 1 mL), nebulised with oxygen, which can be used for all children. This may be repeated after 10 minutes if needed and may help avoid the need for intubation in children who respond to steroids. 'Rebound' phenomena may occur, where the upper airway obstruction may recur as the effect of the adrenaline wears off after 1–2 hours. While in the past it was recommended that any child who received adrenaline for croup should be admitted, a number of studies have now shown that children may be sent home safely if they have also received steroids and have improved to have no stridor at rest over a number of hours.[17,18] Children receiving nebulised adrenaline require close clinical monitoring of their response, particularly the change in air entry, in order to detect any deterioration.

Helium-oxygen (heliox) inhalation has been used during emergency transport of children with severe croup, and anecdotal evidence suggests that heliox relieves respiratory distress. While theoretically attractive the problems of specialised equipment, lack of evidence and the very fast response to steroids has resulted in very little use of this mode of treatment.

Intubation needs to be considered in the child who has increasing upper airway obstruction, hypoxia, decreasing conscious state or fatigue despite nebulised adrenaline. These children should be discussed early with a paediatric intensivist in order to optimise management. The ideal setting for this to occur is in theatre or a paediatric intensive care unit environment via gaseous induction, using an endotracheal tube 1.0 mm smaller than predicted by the child's size (see Chapter 2.3 and Chapter 24.3).

Prognosis

Most children with croup have mild symptoms, do not need hospitalisation and will recover within a few days. Their symptoms will be shortened even further with the use of steroids. It should be pointed out to parents that steroids will have no effect on the duration of any underlying viral symptomatology. Despite the substantial impact of steroids, the occasional child will still follow a prolonged course of cough and marked stridor for many days. While other diagnoses, such as foreign body, need to be considered, most cases will settle with time.

Prevention

For most children, croup is a one-off episode and well tolerated, especially if steroids are used. Children who suffer repeated episodes of recurrent croup, as described above, may benefit from steroid use at home at the first sign of croup symptoms. Although no trials have evaluated this approach, anecdotal evidence suggests that this practice appears to have benefit.

Controversies and Future Research

1. Although once controversial, steroids are now generally accepted for all children who present to emergency departments with croup.
2. It is clear that both prednisolone and dexamethasone are effective in the treatment of croup; however, two direct comparisons have not resolved the question on whether the shorter half-life of prednisolone results in more ‘relapses’ at home. While a once-only dose of dexamethasone is sufficient for the vast majority of children with croup, a second dose of prednisolone 24 hours later may be needed in some cases.
3. Children with recurrent croup theoretically should benefit from early treatment at home, but no formal study to date has confirmed this.

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6.5

Acute asthma

Colin Victor Eric Powell

ESSENTIALS

1 Acute asthma is one of the most common paediatric presentations to an emergency department.

2 It is important to understand the patterns of asthma in children – infrequent episodic, frequent episodic, and persistent.

3 There are three major groups of factors contributing to ‘high-risk’ asthma in children: previous severe asthma, management issues and psychological factors.

4 The most important parameters in the assessment of the severity of childhood asthma are the general appearance/mental state and work of breathing.

5 Life-threatening asthma is characterised by silent chest, cyanosis, poor respiratory effort, exhaustion and altered mental state.

6 The main therapies for paediatric asthma include oxygen, inhaled bronchodilators and steroids modified according to severity.

7 Discharged patients should be given a clear asthma action plan and have appropriate follow-up organised.

8 There are many areas of contention regarding the management of asthma in children, and the recommended guidelines vary among countries. One should be familiar with the local paediatric guidelines.

Introduction

The *Australian Asthma Handbook* was last updated in October 2016 [1] and forms the basis of this chapter. However, there are other best practice guidelines[2,3] and national guidelines,[4–7] and these are important resources for cross-reference and comparison to highlight the controversies.

The most recent revised global estimate of asthma in 2014 suggests that as many as 334 million people have asthma and that the burden of disability is high.[8] Acute asthma is one of the most common reasons for presentation to an emergency department (ED) and admission to a hospital.[9] A review of admissions to nine paediatric EDs in Australia and New Zealand, examining over 300,000 presentations, demonstrated that acute asthma was the fourth most common presentation, accounting for 3.5% of the total number of presentations.[10]

It is well recognised that in many cases admission to hospital may be preventable[9] if asthma is managed effectively by the family and medical team involved with a child's care. Management and clinical practice are highly variable, particularly for severe to critical acute asthma.[10–14]

Diagnosis of asthma

Consider acute asthma when a child presents with signs of increased work of breathing, widespread wheezing and shortness of breath. Asthma flare-ups, exacerbations or attacks have been defined by the Global Initiative for Asthma (GINA) [6] as '*an acute or sub-acute worsening of symptoms and lung function compared with the patient's usual status*'.

There are other causes to consider such as *Mycoplasma pneumoniae*, aspiration, inhaled foreign body, and cardiac failure (Table 6.5.1). In the setting of a child with a previous history of asthma or where asthma seems the most likely diagnosis, one can perform a primary assessment of severity and institute the initial treatment at the onset of history taking.

Table 6.5.1

Differential diagnosis of asthma

Acute	Chronic
Bronchiolitis, mycoplasma	Cystic fibrosis
Allergy	Cilial dyskinesia
Aspiration	Immune deficiency

Heart failure	Bronchiectasis
Foreign body	Airway abnormalities

A large number of epidemiological studies around the world have highlighted the variable phenotypic patterns of wheezing.[1] The preschool child in particular has continued to be a diagnostic challenge.[15] Some of the treatment dilemmas will be highlighted in this chapter.

The *Australian Asthma Handbook* classifies asthma into two age groups, 0 to 5 years and 6 years and over. The definitions used for asthma patterns in children not taking a regular preventer are outlined below.

Infrequent intermittent asthma

Infrequent intermittent asthma is the most common pattern, accounting for 70% to 75% of children with asthma. A child is symptom free for at least 6 weeks at a time, and there are no symptoms between flare-ups. They require management of the individual episode only, and regular preventive therapy is unnecessary. Within this group there is a wide range of severity. Most are mild, but this group accounts for up to 60% of paediatric hospital admissions for asthma.[1]

Frequent intermittent asthma

Frequent intermittent asthma accounts for approximately 20% of childhood asthma. This pattern is similar to infrequent intermittent asthma, but the interval between episodes is shorter, less than 6 to 8 weeks, but there remain no symptoms between flare-ups. These children may benefit from preventive therapy such as low-dose (maximum 400 mcg per day) inhaled corticosteroids or leukotriene antagonist. [1]

Persistent asthma

Persistent asthma accounts for 5–10% of childhood asthma and can be split into three levels of severity: mild, moderate or severe.

a. **Mild persistent asthma** presents with at least one of the following:
   - Daytime symptoms more than once per week but not every day
     or
   - Nighttime symptoms more than twice per month but not every

week.

b. **Moderate persistent asthma** presents with at least one of the following:

- Daytime symptoms daily
- Nighttime symptoms more than once per week

Table 6.5.2

Rapid primary assessment of acute asthma in children [1]

Mild/moderate	Severe	Life threatening
Can walk, speak whole sentences in one breath (For young children: can move around, speak in phrases) Oxygen saturation >94%	**Any** of these findings: Use of accessory muscles of neck or intercostal muscles or 'tracheal tug' during inspiration or subcostal recession ('abdominal breathing') Unable to complete sentences in one breath due to dyspnoea Obvious respiratory distress Oxygen saturation 90–94%	**Any** of these findings: Reduced consciousness or collapse Exhaustion Cyanosis Oxygen saturation <90% Poor respiratory effort, soft/absent breath sounds

Used with permission.

Source: National Asthma Council Australia. *Australian Asthma Handbook*, Version 1.2. National Asthma Council Australia, Melbourne, 2016. http://www.asthmahandbook.org.au.

- Symptoms sometimes restrict activity or sleep.

c. **Severe persistent asthma** displays any of the following:

- Continual daytime symptoms
- Frequent nighttime symptoms
- Frequent flare-ups
- Symptoms frequently restrict activity or sleep.

Some children may be readily controlled with low-dose corticosteroid preventive therapy, but many with frequent severe symptoms and abnormal lung function may require additional therapy including long-acting beta-agonists and other medications.[1]

Risk factors for mortality

Mortality from acute asthma is fortunately rare. A confidential enquiry into

deaths from asthma suggests that there are three major factors contributing to the death:[9]

- **The severity of the disease:** most children who have died from asthma have persistent asthma; however, a minority of children who have died have only mild to moderate disease.[1,9] Previous near-fatal asthma, previous admission in the past year, previous admission to paediatric intensive care unit (PICU), heavy use of β_2-agonists and repeat attendances to the ED are all risk factors associated with risk of a severe attack, near-fatal asthma or death.[1,9]
- **Medical management:** inadequate treatment with steroids, heavy or increasing use of β_2-agonists, inadequate monitoring of the asthma, underuse of written asthma plans and delay in seeking help have all been associated with deaths from asthma.[1,9]
- **Psychological factors:** non-adherence, poorly perceived symptoms, failure to attend clinics, conflict between child and parent or medical staff, and family dysfunction are all risk factors associated with risk of death from asthma.[1,9]

A child should be considered as high risk when attending the ED with an acute exacerbation of asthma when there is a combination of features of previous severe asthma and one or more adverse psychological factors.

Clinical assessment

Examination

The most important parameters in the assessment of the severity of acute childhood asthma are general appearance/mental state and work of breathing (accessory muscle use, recession), as indicated in Table 6.5.2. Initial SaO_2 in air, heart rate and ability to talk are helpful but less reliable additional features. Wheeze intensity, *pulsus paradoxus* and peak expiratory flow rate are not reliable.[2] Clinical signs of acute asthma correlate poorly with the severity of the asthma attack, and none of the signs in isolation is predictive of severity.[4]

The *Australian Asthma Handbook* has simplified the classification of acute asthma into three categories; mild/moderate, severe and life threatening (see Table 6.5.2.) but acknowledges that these may differ from those used in

published clinical trials and other guidelines that focus on, or are restricted to, the management of acute asthma within EDs or acute care facilities. The severity category may change when more information is available (e.g. pulse oximetry, spirometry) or over time. Oxygen saturation levels are a guide only and are not definitive; clinical judgment should be applied.

Investigations

Chest X-ray is not generally required in children with asthma, unless one has the suspicion of an alternative diagnosis or complication (air leak or atelectasis). Arterial blood gas and spirometry are rarely required in the assessment of acute asthma in children.

Differential diagnosis

During an acute episode of wheezing, asymmetry on auscultation is often found due to mucous plugging but warrants consideration of foreign body. Unless the child is particularly unstable, a trial of bronchodilators will often resolve asymmetrical breath sounds.

Consider other causes of wheeze (e.g. bronchiolitis, mycoplasma, aspiration, heart failure or foreign body). Chronically wheezy children may have a diagnosis other than asthma, such as cystic fibrosis, cilial dyskinesia, immune dysfunction, developmental/congenital abnormality, upper airway problems or bronchiectasis. There may be clues in the family or perinatal history or symptoms and signs that may suggest an alternative diagnosis to asthma.[4]

Treatment

The crucial aspect of managing an acute flare-up is to review regularly and assess response to treatment. The *Australian Asthma Handbook* suggests the following approach:

1. Initial assessment and categorise and commence treatment based on clinical features and oxygen saturation in air (see Table 6.5.2) (rapid primary assessment)
2. Reassess severity within minutes (Tables 6.5.3 and 6.5.4) (secondary features)
3. Reassess and review response after the first hour of treatment and

escalate or de-escalate according to response.

Treatment – mild/moderate

- Need for O_2 should be assessed. Give supplemental O_2 to maintain SpO_2 of at least 95%. Salbutamol by MDI/spacer three doses, every 20-minutes; review 10–20 minutes after third dose and decide on admission or discharge
- Oral prednisolone (2 mg kg^{-1} daily for 3 days up to 60 mg). This may be omitted in children aged 0–5 years with a good response to initial bronchodilators
- The few children of moderate severity who can go home must be discussed with a senior doctor and should not leave the ED until at least 1 hour after their last spacer treatment
- Ensure device/technique appropriate
- Provide written advice/action plan on what to do if symptoms worsen
- Consider overall control and family's knowledge. Arrange follow-up as appropriate (see discharge pack).

Table 6.5.3

Secondary severity assessment of acute asthma in children 0–5 years[1]

	Mild/moderate	Severe	Life threatening
Speech Posture Breathing	Can talk or vocalise Can walk or crawl No distress	Not applicable; may be the same as moderate Lethargic Paradoxical chest wall movement: inward movement on inspiration and outward movement on expiration (chest sucks in when person breathes in) or Use of accessory muscles of neck or intercostal muscles or 'tracheal tug' during inspiration or Subcostal recession ('abdominal breathing')	Unable to vocalise due to dyspnoea Collapsed or exhausted Severe respiratory distress or exhausted
Consciousness	Alert	Not applicable; may be the same as moderate	Drowsy or unconscious
Skin colour	Normal	Not applicable; may be the same as moderate	Cyanosis
Respiratory rate*	Normal	Tachypnoeic	Bradypnoea (indicates respiratory exhaustion)
Heart rate*	Normal	Tachycardia	Bradycardia (may occur just before respiratory arrest)
Chest auscultation	Wheeze or normal	Not applicable; may be the same as moderate	Silent chest or reduced air entry
Oxygen saturation (pulse oximetry)	>94%	90–94%	<90% or Clinical cyanosis

*	Normal heart rate (beats/minute)	Normal respiratory rate (breaths/minute)
<1 year	110–160	30–40
1–2 years	100–150	25–35
2–5 years	95–140	25–30

Source: National Asthma Council Australia. *Australian Asthma Handbook*, Version 1.2. National Asthma Council Australia, Melbourne, 2016. http://www.asthmahandbook.org.au.

Table 6.5.4

Secondary severity assessment of acute asthma in children 6 years and over[1]

	Mild/moderate	Severe	Life threatening
Speech Posture Breathing	Can finish a sentence Can walk No distress	Can only speak a few words Unable to lie flat or hunched forward Paradoxical chest wall movement: inward movement on inspiration and outward movement on expiration (chest sucks in when person breathes in) or Use of accessory muscles of neck or intercostal muscles or 'tracheal tug' during inspiration or Subcostal recession ('abdominal breathing')	Can't speak Collapsed or exhausted Severe respiratory distress or exhausted
Consciousness	Alert	Not applicable; may be the same as moderate	Drowsy or unconscious
Skin colour	Normal	Not applicable; may be the same as moderate	Cyanosis
Respiratory rate*	< or = to 25 breaths per minute	>or = to 25 breaths per minute	Bradypnoea (indicates respiratory exhaustion)
Heart rate*	Normal	Tachycardia	Bradycardia (may occur just before respiratory arrest)
Chest auscultation	Wheeze or normal	Not applicable; may be the same as moderate	Silent chest or reduced air entry
Oxygen saturation (pulse oximetry)	>94%	90–94%	<90% Clinical cyanosis

*	Normal heart rate (beats/minute)	Normal respiratory rate (breaths/minute)
5–12 years	80–120	20–25
12–18 years	60–100	15–20

Used with permission.

Source: National Asthma Council Australia. *Australian Asthma Handbook*, Version 1.2. National Asthma Council Australia, Melbourne, 2016. http://www.asthmahandbook.org.au.

Treatment – severe

- Oxygen
- Salbutamol by MDI/spacer or nebuliser – three doses every 20 minutes; review ongoing requirements 10–20 minutes after third dose; if improving reduce frequency, if no change continue every 20 minutes, if deteriorating at any stage treat as life threatening
- Ipratropium bromide by MDI/spacer or nebuliser. Three doses in the first hour every 20 minutes
- Consider nebulised magnesium sulphate 154 mg in 2.5 mL saline – three doses over 1 hour mixed with the salbutamol and ipratropium bromide if both nebulised
- Oral prednisolone (2 mg kg^{-1} daily up to 60 mg); if vomiting give intravenous (IV) methylprednisolone 1 mg kg^{-1} or hydrocortisone 4 mg kg^{-1} every 6 hours

- Involve senior staff
- Arrange admission after initial assessment.

Treatment – life threatening

- Oxygen
- Continuous nebulised salbutamol (0.5% undiluted)
- Nebulised ipratropium 250 mcg three times in first hour (every 20 minutes, added to salbutamol)
- Consider nebulised magnesium sulphate 154 mg in 2.5 mL saline three times in first hour added to salbutamol and ipratropium. Methylprednisolone 1 mg kg^{-1} or hydrocortisone 4 mg kg^{-1} every 6 hours
- Magnesium sulphate 50% 0.1 mL kg^{-1} (50 mg kg^{-1}) IV over 20 minutes, then 0.06 mL kg^{-1} hr^{-1} infusion with serum level 1.5–2.5 mmol L^{-1}
- If deteriorating give IV salbutamol 15 mcg kg^{-1} over 10 minutes, then 1–5 mcg kg^{-1} min^{-1}
- If poor response to IV salbutamol, consider aminophylline 10 mg/kg IV (maximum dose 250 mg over 60 minutes). If currently taking oral theophylline, do not give IV aminophylline in ED – take serum level. Following loading dose give continuous infusion (1–9 years: 1.1 mg kg^{-1} hr^{-1}, 10+ years: 0.7 mg kg^{-1} hr^{-1}; commence in ED if there will be >2 hours before transfer to ward). Involve senior staff, discuss with local PICU
- If conscious state permits, ventilatory support (if required) may be attempted with non-invasive bi-level positive pressure ventilation. This may be facilitated by sub-dissociative doses of intravenous ketamine (usually 0.5 mg kg^{-1})
- Where intubation is indicated, ketamine is the induction agent of choice due to its bronchodilator action. Important considerations include intravenous fluid loading, good pre-oxygenation, placement of a cuffed endotracheal tube and avoiding hyperventilation. Ventilator settings should normalize oxygenation but allow permissive hypercapnoea.

Discharge from hospital

A child should be ready for discharge when it is considered that he/she can be

stable on 3–4-hour inhaled bronchodilators.[16] This is often a subjective decision and does not necessarily require the child to be observed in the ED for 3–4 hours prior to discharge.

Advise parents to seek further medical review should their child's condition deteriorate or if there is no significant improvement within 48 hours. At discharge all patients should have follow-up arranged with a paediatrician or their local doctor. Parents should be informed of other sources of information about asthma such as the Asthma Foundation.

The concept of an asthma discharge pack is useful to ensure all aspects of discharge are considered. Adult data suggest that self-monitoring, regular review and written action plans can improve outcomes.[17] Two paediatric studies suggest that an intensive nurse-led discharge concentrating on education, written action plans and inhaler technique and appropriate follow-up with discharge prescription for steroids can reduce readmissions and following morbidity.[18,19]

Discharge pack

1. *Review need for preventative treatment.* Consider preventative treatment if there are wheezing attacks less than 6 weeks apart, the attacks are becoming more frequent and severe or there are increasing interval symptoms. Initial preventative treatment for frequent episodic asthma is inhaled steroids.
2. *Check inhaler technique.* Emergency attendance or admission should provide the patient and family with the opportunity to use an appropriate size spacer device and pMDI. Make sure the child and family can use the device adequately and know the importance of using it for all preventative therapy and treatment for significant exacerbations.
3. *Family education.* On discharge from the ED or ward it is important that families understand the immediate management of their child's asthma. It is not appropriate to educate them on all aspects of asthma during an acute episode. This is best reserved for a visit to an outpatient clinic or local doctor at a time remote from the acute episode when a reasonable amount of time can be allocated and it is more likely that the information will be understood and retained.
4. *Prescription.* A prescription for all medications should be provided at the time of discharge.
5. *Follow-up.* All patients should have a clear follow-up plan. For some it

will be appropriate that they visit their local doctor for an early review, particularly if their condition deteriorates or fails to improve significantly within 48 hours. At discharge all patients should have an outpatient appointment or appropriate follow-up arranged with a paediatrician or local doctor within 4 to 6 weeks. This visit will be used for medical review and, most importantly, appropriate education about asthma management.

6. *Written action plan.* All patients should have an individual written action plan, and the discharging doctor should spend time going over the plan with the family.
7. *Communicate with the family doctor.* For every emergency attendance or discharge, there should be communication with the patient's local doctor. The local doctor should receive a copy of the action plan.

Prognosis

Those with episodic asthma tend to improve throughout childhood, with asthma resolving by their adult years in approximately two-thirds. Those who continue to wheeze tend to have very mild asthma and maintain normal lung function. On the other hand, those with persistent asthma in childhood are more likely to continue to wheeze in their adult years (about two-thirds), with some impairment of lung function. Available evidence suggests that treatment does not influence the natural history of childhood asthma.[1]

Prevention

There are two areas of prevention to consider (primary and secondary) but the evidence is mostly inconclusive and confusing.[4] Breast-feeding and avoiding parental cigarette smoking are proven to be effective primary prevention of asthma. For secondary prevention, avoiding tobacco smoke is important. In committed families, house dust mite avoidance by use of bed covers, removal of carpets and soft toys, dehumidification, high-temperature washing of bed linen and use of acaricides on soft furnishing may reduce morbidity from asthma.[4]

If an organised discharge from hospital is completed as per recommendations (as above), then this is more likely to prevent readmission or re-presentation and reduce morbidity. A number of preventable factors associated with admission have been identified, and these issues should be addressed at discharge:

adherence issues, prophylactic treatment, action plan use and advice to prevent delay in seeking medical advice.[9]

Controversies

1. **Bronchodilator aerosol delivery.** Pressurised metered dose inhalers (pMDI) and spacers are an effective way of delivering inhalation medication to treat mild to moderate and probably severe attacks of asthma.[20] There is still debate as to cost in different countries and whether the pMDI/spacer combination use in adult acute asthma is as effective as nebulisers.[20] However, it is becoming more apparent that there is a danger of salbutamol toxicity (metabolic acidosis) especially in the younger children that will make them tachycardic and tachypnoeic and so the *Australian Asthma Handbook* recommends lower dose inhaled treatment for children between the ages of 0 and 5 years.[1] There is little evidence for any benefit for continuous nebulised (0.5% undiluted salbutamol) treatment compared to frequent intermittent doses of treatment,[4] although many CPG recommend use in life-threatening exacerbations.[1–3]
2. **Ipratropium bromide.** Repeated early doses of ipratropium bromide (IB) during the first 1–2 hours of presentation are associated with improved outcome, and efficacy and safety have been demonstrated particularly in those with a more severe attack, lowering the risk of hospital admission.[21] Standard doses of 250–500 mcg per dose for nebulised treatment seems universal.[1–7] Repeated doses of ipratropium bromide should be given early in children who are not responsive to β_2-agonists.[4] It is not clear whether there are any benefits from continued use after the attack has shown response to treatment. Once hospitalised, the addition of ipratropium to salbutamol and steroids appears to add no benefit.[22] The British Thoracic Society (BTS) recommends that the dose should be weaned down to every 4–6 hours or stopped once admitted.[4]
3. **Nebulised magnesium** ($MgSO_4$) does not have a role in adult asthma.[23] The MAGNETIC study showed that there was a statistically significant benefit with improved asthma severity score in the first hour when adding nebulised $MgSO_4$ to the initial three nebulised treatments, mixed with salbutamol and ipratropium in the first hour in those

children with more severe exacerbations and those with a short history of symptoms. It did not show a reduction on admissions, need for intravenous treatment, PICU admissions or length of stay.[24] A number of guidelines suggest considering adding this to the initial nebulised treatment to maximise the inhaled therapy.[4,6,25] There is an ongoing study in Canada examining the role further but is using a much higher dose and only in those children with a severe exacerbation.[26]

4. **Intravenous bronchodilators.** The main problem with deciding which intravenous bronchodilator to use (aminophylline, $MgSO_4$ or salbutamol/terbutaline) is that there are no good direct comparisons with the three intravenous treatments commonly used. Thus the referenced guidelines vary with recommendations,[1–7] and practice varies extensively, especially on drugs chosen and dose regimen used.[13,14] Each unit will have its own locally agreed guidelines, and it is important to understand and follow those; it is important that staff are familiar with the drugs chosen as regards preparing and administering the drug:

salbutamol. There are few clinical studies on the effectiveness of intravenous salbutamol therapy, and there have been major concerns about intravenous salbutamol and toxicity producing a lactic acidosis and hypokalemia especially in children <5 years.[27] Some centres do not use it routinely because of this concern.[2] Some Australian guidelines suggest that intravenous salbutamol should be used as a third-line intravenous treatment if there is no response to aminophylline and $MgSO_4$.[2]

Aminophylline. There is no evidence that there is a role for aminophylline in children with mild to moderate exacerbations of their asthma. However, if a child is unresponsive to maximal inhaled therapy as above, then in the more severe and life-threatening attacks, aminophylline may reduce intubation rates.[28] This remains a controversial area. There are few data on what should be a safe loading dose for aminophylline because of concerns of adverse events such as vomiting and arrhythmias.[29] Many guidelines suggest loading at 5 mg/kg[4], whereas the data suggest the dose should be 10 mg/kg.[28] There is no doubt that vomiting is a serious side effect from the higher doses of aminophylline, and this may be a reason not to use it.[29]

Magnesium. Intravenous magnesium relaxes smooth muscle and causes

bronchodilatation. It is very commonly used and in many cases the initial intravenous bronchodilator utilised in children.[13,14] It is considered first-line intravenous therapy in the *Australian Asthma Handbook*.[1] This is probably due to its ease of use and good safety profile. Doses of 40–100 mg kg^{-1} as a 20-minute infusion have been used with varying effects on lung function and asthma severity scores when compared to placebo.[30] Australian guidelines recommend using 50% 0.1 mL kg^{-1} (50 mg kg^{-1}) IV over 20 minutes for severe and life-threatening episodes, followed by infusion of 0.06 mL kg^{-1} hr^{-1} (30 mg kg^{-1} hr^{-1}): target serum levels 1.5–2.5 mmol L^{-1}.[1] The use of intravenous magnesium sulphate in severe and critical exacerbations is discussed in the BTS guidelines but is not fully endorsed,[4] and GINA[6] does not recommend its use routinely in children but mentions it may be of benefit in severe asthma (no dose discussed). There is still very limited evidence that it has a role; a recent Cochrane review including five studies with only 182 children was that there was very little evidence of benefit.[30]

Adrenaline. This bronchodilator is not mentioned in most guidelines. GINA suggests it may be of use.[6] It is likely that some children presenting with asthma may have anaphylaxis and therefore adrenaline is potentially useful;[31,32] IM adrenaline may well have a role. It is very rarely used in a paediatric population.[13,14]

5. **Corticosteroids.** There is no doubt that early use of steroids for an acute attack reduces not only the need for admission but also morbidity.[33] There are a number of different dosing regimens suggested, but none has been shown to be superior and so there can be no clear recommendations about prednisolone or dexamethasone as a first-line oral steroid.

   There has been recent evidence that systemic steroids may not have a role in acute wheezy episodes in children <5 years of age who may have simple viral-induced wheeze rather than asthma.[34] Practice varies among hospitals so be familiar with local guidelines:

   Inhaled steroids. Currently there is insufficient evidence in children that inhaled steroids should replace oral or systemic steroids in acute exacerbations.[1–7]

6. **Leukotriene antagonists.** Initiating oral leukotriene antagonists in primary care early at the onset of an exacerbation can result in reduced

symptoms, time off school, healthcare resource utilisation and parental time off work in children with intermittent asthma.[35] There is no clear evidence of a role as intravenous therapy in acute asthma exacerbations; only adult data exist.[36] Current studies are reviewing the use of leukotriene antagonists in the management of acute asthma in children.

7. **Children less than 2 years of age.** This age group can be difficult to assess, and the different phenotypes of acute wheezing and indeed different labels used in different countries can cause problems when examining the literature on appropriate treatment. There is little evidence that β_2-agonists or ipratropium bromide have an impact on wheezy children of this age in regard to their need for hospitalisation or length of stay.[37] Steroid therapy may have a role, but this is still not clear.[33,34] There are problems differentiating between bronchiolitis and asthma in younger infants, but this is outside the scope of this chapter.
8. **Intensive care management.** The *Australian Asthma Handbook* and BTS recommend that once a child has severe enough asthma to require intravenous treatment, he/she should be referred to a PICU or an PHDU even if he/she does not require intubation.[1,4] There are a number of indications for intensive care admission: deteriorating lung function, persisting or worsening hypoxia, hypercapnia, exhaustion, drowsiness, confusion, coma or respiratory arrest.[1,4] These features are clearly all part of a spectrum, and the overall picture, plus lack of response to treatment, should indicate that the child should be admitted to a high dependency or intensive care area. They may not necessarily require ventilation. There are no absolute criteria, and the decision needs to be made by an experienced physician.[1–7]

   Non-invasive ventilation has had some success in adults, but none of the paediatric guidelines recommends its use. Bi-level positive airway pressure is being used in paediatrics.[13,14] It has been shown in adults and children to improve gas exchange and clinical improvement with or without sedation, which may include ketamine or benzodiazepines.[38] High-flow oxygen delivery is also another treatment that is evolving but there are no good data yet to support its use.[39] Detailed discussion about ventilating a child with asthma is beyond the scope of this chapter. Helium and oxygen mixtures have been used in adults but again are not endorsed in the paediatric guidelines on current

evidence.[1–7]

Future directions/research

The following is a list of themes needing further development in acute asthma – it is by no means exhaustive:

- Prevention of developing asthma and secondary prevention – can we reduce asthma morbidity?
- Patient education and asthma care delivery – how best to deliver asthma care and how best to improve patients' and families' knowledge?
- Gaps in practice and delivery of care – how best to audit asthma care and improve implementation of guidelines?
- Improved assessment of acute asthma – what is the best predictor of asthma severity, need for treatment and hospital admission?
- What should be the core outcomes measured for acute asthma research in children?
- Are there different phenotypes of acute asthma with potentially different treatment modalities?
- Do long-acting β_2-agonists have a role in treating acute asthma?
- Pharmacogenomics – are there different genotypes with different responses to treatment?
- Which is the most effective intravenous bronchodilator?
- What is the best mode of delivery for aerosolised treatments?
- Non-invasive ventilation – where does this have a role?
- What is the role of inhaled magnesium, intravenous leukotriene antagonists and inhaled helium mixture?

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6.6

Pertussis

Jim P. Buttery, and Nigel W. Crawford

ESSENTIALS

1 Pertussis infection is most severe in infants less than 6 months of age.

2 All children less than 6 months with pertussis should be considered for a period of observation in order to assess the risk of significant apnoea.

3 Three stages of illness occur – catarrhal, paroxysmal and convalescent.

4 The mainstay of treatment is supportive. Admission allows for apnoea monitoring, oxygen, and overcoming feeding difficulties.

5 Respiratory complications are the most common, with pneumonia the major cause of mortality (0.5–1%) in infants <6 months.

6 Treatment is with macrolide antibiotics to eradicate carriage rather than modify illness (e.g. clarithromycin for 7 days), best if given early in the course of the illness.

7 Prevention is by universal immunisation with pertussis vaccine during pregnancy, childhood, plus a booster in adolescence/adulthood.

Introduction

Pertussis or ‘whooping cough’ is a bacterial infection of the respiratory tract caused by the gram-negative coccobacillus *Bordetella pertussis*. The word

pertussis itself means ‘intensive cough’, and the epidemic was described in 1578 in Paris.[1] Also known as the ‘100 day cough’, pertussis is the preferred term because not all cases have the classical paroxysms of coughing with an inspiratory ‘whoop’, which occurs as a massive respiratory effort forces inhaled air against a narrow glottis.

Pathophysiology

Humans are the only reservoir, and the incubation period is approximately 7–14 days. Transmission is primarily by direct contact with droplets via the airborne route. The organism is highly communicable early in the illness, with attack rates of 75–100% from symptomatic individuals to susceptible contacts. Every case infects approximately 5.5 other people.[2] Pertussis is associated with a significant morbidity and mortality, particularly in young infants.

Epidemiology

With the advent of universal vaccination programmes in the 1940s the incidence of this disease decreased markedly. Due to concerns regarding immunisation, vaccination levels waned in the late 1970s, and there was a resurgence in the number of cases. Worldwide there are approximately 250,000 deaths attributed to pertussis per year. There have been epidemics every 3–4 years in Australia, with epidemiological data showing an estimated incidence of 140 cases per 100,000 per year during 2008–2011.[3]

History

The possibility of pertussis should be considered in a child presenting to the emergency department (ED) with prominent coughing episodes, as well as young infants with apnoea. The diagnosis of pertussis is usually made on the basis of a suggestive clinical history, confirmed by isolation of the organism.

There are three characteristic stages of the illness, and presentation is most commonly in the paroxysmal phase.

Stage 1: Catarrhal (1–2 weeks)

Upper respiratory tract infection symptoms, e.g. rhinorrhoea, conjunctivitis, malaise and low-grade fever.

Stage 2: Paroxysmal (2–6 weeks, can be longer)

Consists of paroxysms of coughing that may be followed by an inspiratory ‘whoop’. Facial suffusion, with prominent eyes and protrusion of neck veins, may be seen during these paroxysms. The paroxysms can cause fatigue, which may impair an infant’s ability to take feeds. Post-tussive vomiting commonly follows the coughing episode, but between times the child may appear quite well. It is important to note that young infants may present with apnoea as the only symptom. This can result in a presentation of sudden collapse.

Stage 3: Convalescent (1–2 weeks)

The paroxysms of coughing, whooping and vomiting decrease in number and severity. The cough may persist for a number of weeks/months, and future episodes of upper respiratory tract infections may restimulate the coughing paroxysms.

Pertussis is most severe in infants younger than 6 months of age, particularly the non-immunised. It is these high-risk infants who particularly need to be considered for a period of observation to exclude significant apnoea events. Maternal antenatal vaccination in the third trimester is safe and has an estimated 91% effectiveness in preventing admission to hospital with pertussis in infants up to 3 months of age.[4] In infants, the clinical manifestations can mimic those of bronchiolitis or other infective pneumonitis. In partially immune adults and adolescents, who have waning immunity, the presentation is less typical and can be that of a persistent cough.

Examination

The clinical findings depend on the stage of the illness when the child presents. Some children will kindly display a typical coughing paroxysm in the ED and reinforce the description given by parents. This may include features of facial colour changes or a whoop that will suggest pertussis as the likely cause.

Other children may have little to find on examination, apart from mild coryza. In these, the diagnosis is based on a suggestive history and subsequent isolation of the organism. There may be visible mechanical sequelae, due to coughing and vomiting, such as petechiae or subconjunctival haemorrhages. Unless a co-infection or atelectasis due to plugging has occurred, the chest examination is generally unremarkable.

Investigations

The clinical diagnosis is typically confirmed by polymerase chain reaction (PCR) assay of a nasopharyngeal swab. These are more sensitive than traditional culture or antigen detection assays. Serology testing includes anti-*B. pertussis* IgA (suggests present or past infection), IgM or IgG (infection or vaccination).[5]

The full blood examination, at the end of the catarrhal or early in the paroxysmal phase, can have a leukocytosis with a predominant lymphocytosis.

A chest X-ray may show perihilar infiltrates, interstitial emphysema or evidence of a secondary pneumonia.

Differential diagnosis

A ‘pertussis-like syndrome’ may be caused by other organisms, including *Bordetella parapertussis*, adenoviral infections, *Mycoplasma pneumoniae* and *Chlamydia pneumoniae*. They often lead to a milder, shorter clinical course and can be isolated by a nasopharyngeal sample, or specific serology can be performed. In the first few months of life, viral bronchiolitis may cause a very similar clinical picture to that of pertussis and only be distinguished on the basis of the microbiology result. Wheeze is present in pertussis only if there is co-infection.

Complications

Respiratory complications are frequent, with pneumonia the most common. It may be due to a secondary bacterial infection. Occasionally, the pneumonia can be a severe necrotising form, which is the major cause of death. Atelectasis is common, resulting from mucous plugging, and air leaks (pneumothoraces, interstitial emphysema or pneumomediastinum) may occur secondary to ruptured alveoli. Bronchiectasis is a rare late sequela.

Subconjunctival haemorrhages, rectal prolapse or inguinal hernia may occur due to increased intraabdominal pressure. Cerebral anoxia with convulsions can occur in young children and encephalopathy is seen in approximately 1 in 10,000 cases.[6,7] Mortality is estimated at 1 in 200 infants diagnosed with pertussis under the age of 6 months.

Treatment

All infants <6 months of age with suspected pertussis should be considered for admission with isolation, due to the risk of apnoea and other complications. This allows for a period of observation of coughing episodes and monitoring for apnoea and desaturations. Older children will require admission should they have significant apnoea/cyanosis or feeding problems. Treatment is mainly supportive via monitoring and the provision of oxygen and fluids.

Treatment with macrolides is most effective if given early in or before the catarrhal phase and has little effect in the paroxysmal phase. Newer macrolides are preferred to erythromycin, and a 5-day course of azithromycin or 7 days of clarithromycin is effective at eradicating carriage.[8] Co-trimoxazole is the drug of choice if macrolides are contraindicated. There is minimal chance of transmission after 5 days of treatment. Consider broader spectrum antibiotics if there is evidence of superadded bacterial infection. The index case should be excluded from nursery/school until 5 days of treatment is completed.[7,8]

Chemoprophylaxis of contacts

Contact tracing is difficult, but the family and close household contacts should have a course of antibiotics regardless of immunisation status. Pregnant women exposed around the time of delivery and their newborn infants should also receive chemoprophylaxis. The contacts should also have vaccination at the same time, particularly those under 8 years who have not received their five DTPa vaccinations. There is no benefit from passive immunisation prophylaxis with human pertussis immunoglobulin.[7]

Prevention

Immunisation first commenced in the late 1940s with the triple vaccine, diphtheria tetanus pertussis (DTP). This was initially a whole-cell vaccine, but since the 1990s an acellular vaccine has been available in Australia. It has been shown to be almost as effective, preventing disease in 85% of cases with fewer side effects, and is now recommended.

Current immunisation schedule

DTPa: 6 weeks; 4, 6 and 18 months; and 4 and 13–15 years. Also recommended in the third trimester of pregnancy.

There are also multiple-combination vaccines available: e.g. DTPa-hepB. An adult-formulation pertussis-containing vaccine (dTpa) suitable for boosting adults and adolescents is recommended as a single dose at 13–15 years of age.[7]

Complications of the vaccine may potentially present to ED and include fever >38°C (uncommonly leads to a febrile convulsion), erythema at the injection site, persistent crying, drowsiness, vomiting, anorexia, systemic allergic reaction and hypotonic–hyporesponsive episodes (rare, with no long-term sequelae).

Prognosis

The severity is directly related to age, with the illness normally milder in older children. Morbidity and mortality (0.5–1%) are still significant in infants under 6 months of age.

Controversies and Future Directions

1. An encephalopathy has been controversially linked with the pertussis vaccination in the past, but the evidence is inconclusive, as the event is so rare and no causal link has been proven.[6] Pertussis vaccine does not cause infantile spasms, epilepsy or sudden infant death syndrome (SIDS).
2. The drop in vaccination rates in the 1970s and the subsequent increase in the incidence of this preventable disease highlight the need for ongoing surveillance to monitor the uptake and efficacy of vaccinations, the frequency of adverse reactions and the incidence of epidemics. Genomic analysis of circulating *B. pertussis* isolates has identified potential vaccine ‘escape’ strains but also may point the way to better vaccines.[9] There is also a need for more studies on equivalence testing of different antibiotics for treatment of pertussis.

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6.7

Community-acquired pneumonia

Mike Starr

ESSENTIALS

1 Viruses are the most common cause of community-acquired pneumonia in children, although up to 40% of cases represent mixed infection.

2 *Streptococcus pneumoniae* is the most common bacterial cause.

3 Routine chest X-rays are not necessary for the confirmation of suspected in children well enough to be managed as outpatients.

4 Blood tests, including full blood count, inflammatory markers and blood culture are generally unhelpful.

5 Oral amoxicillin is appropriate for empiric therapy of most children (outpatient and inpatient) with pneumonia. Intravenous benzylpenicillin can be reserved for those who cannot tolerate oral antibiotics.

6 The routine use of third-generation cephalosporins provides no additional benefit over the penicillins.

7 Follow-up chest X-rays are not required for most children with pneumonia.

Introduction

Pneumonia is a common condition with significant morbidity and mortality. It is the leading infectious cause of death among children under 5 years of age globally and is responsible for 18% of deaths in this age group.[1] Even in

developed countries, pneumonia remains a major cause of acute morbidity and one of the most common reasons for paediatric hospital admissions.[1] The incidence of pneumonia is approximately 40 per 1000 children per year in those under 5 years of age, and 15 per 1000 children per year in 5–14-year-olds.[2,3]

In spite of how common it is, there is no single clinical or radiological definition that is widely accepted for pneumonia. It is investigated with a range of tests, the usefulness of which is known to be incomplete, and it is then often treated without knowledge of the aetiology. Fortunately, most children recover completely with empiric treatment.

Definition

There are various definitions that can be used for pneumonia. From a pathological point of view, it is defined as inflammation or infection of the lung parenchyma. In the clinical setting, the diagnosis is typically made on the basis of a constellation of clinical features, including fever, cough, tachypnoea and auscultatory findings, where clinical wheezing syndromes have been ruled out. Although radiological confirmation has often been held up in the past as the 'gold standard', routine chest X-rays do not affect the clinical outcome,[4] and many guidelines do not include radiographic changes in the definition.[5–7]

Aetiology

In the majority of cases of childhood pneumonia, the causative pathogen is not identified. Blood cultures are positive in under 5% of cases of pneumonia.[3,8] Transthoracic lung aspiration yields a cause in up to 69% of cases[9,10] but is invasive. It is difficult to obtain adequate sputum for microscopy and culture in young children. Other indirect methods of identifying a cause, such as serology, or immunofluorescence, culture or polymerase chain reaction analysis (PCR) of nasopharyngeal swabs, are neither sensitive nor specific.

Although an alveolar or lobar infiltrate on chest X-ray is considered by some to be suggestive of bacterial infection, chest X-ray changes cannot reliably predict aetiology.[11,12] Nor is any radiological pattern pathognomonic for viral or *Mycoplasma pneumoniae* infection.

Age is the best predictor of aetiology of pneumonia. In neonates, where bacterial causes predominate, group B streptococci and *Escherichia coli* are the most common pathogens. Viruses, particularly respiratory syncytial virus (RSV),

parainfluenza, influenza, metapneumovirus and adenovirus, are the most common causes overall, particularly in young children. The occurrence of recent local outbreaks and the clinical pattern may give a clue to the likely causative virus. These viruses appear to be responsible for approximately 40% of cases of community-acquired pneumonia in children who are hospitalised, particularly in those under 2 years of age, whereas *Streptococcus pneumoniae* is responsible for 27% to 44% of cases of community-acquired pneumonia.[13] Up to 40% of infections are mixed.[3]

Infection with *M. pneumoniae* and *Chlamydia pneumoniae* is usually considered to cause pneumonia in children of school age and in older patients, although more recent studies suggest that preschool-aged children have as many episodes of atypical bacterial pneumonia as older children.[13] Staphylococcal and group A streptococcal pneumonia are uncommon but should be considered in children who are severely unwell with invasive disease. These infections are more likely to be seen in indigenous and Pacific Islander children. More recently, infections with community-acquired multiresistant *Staphylococcus aureus* (CAMRSA) have emerged. CAMRSA often results in a necrotising fulminant pneumonia with increased morbidity and mortality.

Gram-negative pneumonia is uncommon in children; non-typeable *Haemophilus influenzae* is mainly seen in children with underlying lung disease, such as cystic fibrosis and bronchiectasis.

The presence of a pleural effusion does not necessarily indicate more severe disease; *S. pneumoniae* remains the most common bacterial cause, with or without effusion.

Clinical findings

Pneumonia should be considered in any infant or child presenting with fever, cough, difficulty breathing, tachypnoea, increased work of breathing (nasal flaring, lower chest indrawing or recession) and auscultatory findings consistent with consolidation or effusion. However, the auscultatory findings may be unreliable in young children, particularly in those under 1 year. Infants may present with symptoms not obviously related to a lower respiratory tract infection, such as lethargy, vomiting, poor feeding, grunting or poor perfusion. Children less than 3 months of age may present with apnoea. Tachypnoea is a valuable sign, and it may be the only clue in some children.[14,15] There is a significant correlation between respiratory rate and oxygen saturation.[14]

However, work of breathing is more indicative of the likelihood of pneumonia.[7] Auscultatory findings, including asymmetrical breath sounds, crackles and bronchial breathing are less predictive. The presence of coryza or wheeze (particularly bilateral) suggests that bacterial pneumonia is unlikely.[7,8] Pneumonia located in the apex of the lung may present with neck pain and be confused with meningitis, whilst basal pneumonia may cause abdominal pain, suggestive of an acute abdomen. Conversely, an infant with grunting respirations may give the initial impression of an intraabdominal problem.

The presentation of pneumococcal pneumonia is generally abrupt, whilst *M. pneumoniae* and viral infections more often present with a more indolent course, less fever, and other symptoms such as malaise, headache, arthralgia and rash.

Investigations

Posteroanterior chest X-rays do not need to be performed routinely in children with mild disease where the diagnosis may be made clinically. Chest X-rays performed in children with a history and examination consistent with pneumonia do not lead to changes in diagnosis or use of antibiotics in 75% of those seen in a clinic or emergency department (ED) setting, and they rarely affect decisions regarding hospitalisation.[4,16,17] Chest X-rays should therefore be reserved for patients who require admission or if severe or complicated pneumonia is suspected. Lateral X-rays do not confer additional information in most cases.

Although it is not possible to reliably predict aetiology or differentiate bacterial from viral pneumonia on chest X-ray, pneumococcal pneumonia typically presents with a lobar infiltrate or round pneumonia. Pneumatoceles, abscesses and cavities are associated most frequently with staphylococcal pneumonia, but they are also seen in pneumonia caused by other bacteria. Bronchopneumonia is more typical of viral or other aetiology.

Large effusions may be difficult to differentiate from empyema. In some cases, ultrasound may be helpful in determining whether loculations are present.

Radiological changes lag behind clinical signs, and may persist for 4–6 weeks. Follow-up chest X-rays are unnecessary in children with uncomplicated pneumonia but should be considered if symptoms and signs are persistent following treatment.

Most other investigations to determine microbiological aetiology are not particularly helpful for dictating immediate medical management in the ED setting; they should only be considered in children with severe pneumonia or

complications. Testing of respiratory secretions for RSV and other respiratory viruses does not usually alter management. Testing for atypical bacteria, including PCR testing of respiratory secretions for *M. pneumoniae* or acute and convalescent serology, is not helpful in guiding management, as testing cannot differentiate between asymptomatic carriage and symptomatic infection.

Other blood tests are only helpful in more unwell children and those receiving intravenous fluids, e.g. full blood examination and urea and electrolytes. Acute phase reactants (particularly C-reactive protein) cannot distinguish between a viral or bacterial cause and are not recommended.

Management

Most children can be managed as outpatients with oral amoxicillin 30 mg kg^{-1} (max 1 g) three times daily for 3 to 5 days; a short course (3 days) of antibiotic therapy is as effective as a longer treatment (5 days) for non-severe pneumonia in children under 5 years of age.[18]

Children under 6 months of age, and those who are more unwell, hypoxic or dehydrated may require inpatient management[6] (Box 6.7.1). Patients should be stabilised as necessary with oxygen therapy, ventilatory support and/or fluid resuscitation. If ongoing nasogastric or intravenous fluids are required, fluid intake should be limited to approximately one-half to two-thirds of normal maintenance fluids to avoid fluid overload and pulmonary oedema. Admitted patients can still be treated with oral amoxicillin, unless they cannot tolerate oral intake. If this is the case, appropriate therapy is intravenous (IV) benzylpenicillin 50 mg kg^{-1} (max 1.2 g) every 6 hours. Infants less than 3 months of age should also be given IV gentamicin 7.5 mg kg^{-1} daily.

Antibiotic resistance among pneumococci is not uncommon. However, there is no difference in outcome between cases of pneumonia caused by susceptible and resistant strains, and amoxicillin or benzylpenicillin remains the treatment of choice.[19,20] The use of third-generation cephalosporins provides no additional benefit over these penicillins. They should be reserved for severe pneumonia where it may be important to cover beta-lactamase producers and gram-negative bacteria.

Children presenting with coryza, wheeze, diffuse crackles and minimal chest X-ray changes may have viral pneumonitis. Admission may be necessary for supportive care, but antibiotics should be withheld. A trial of inhaled bronchodilator therapy may be useful in children who appear to have significant

associated bronchospasm. A Cochrane review found there was insufficient evidence to demonstrate that antibiotic therapy improves outcomes for children with lower respiratory tract infections caused by *M. pneumoniae*. [21]

Only children who are severely unwell (e.g. severe systemic toxicity and rapidly progressive pneumonia) require broader spectrum antibiotics to include cover for *S. aureus* and gram-negative bacteria: ceftriaxone 50 mg kg^{-1} (max 1 g) IV daily or cefotaxime 50 mg kg^{-1} (max 1 g) IV every 6 hours plus flucloxacillin 50 mg kg^{-1} intravenously (IV) (max 2 g) every 6 hours. In settings or populations where CAMRSA is more prevalent, vancomycin 15 mg kg^{-1} (max 750 mg) every 6 hours should be added. The addition of azithromycin 10 mg kg^{-1} (max 500 mg) IV daily may be considered if the pneumonia is progressing despite this therapy.

Box 6.7.1 Possible indications for admission

Hypoxia, apnoea
- Toxic appearance, poor feeding, dehydration
- Age <6 months
- Underlying lung disease or immunodeficiency
- Extensive consolidation
- Failed response to oral therapy

Follow-up chest X-rays are not required for those who recover uneventfully; for those with complicated pneumonia, or persistent signs after 4 to 6 weeks, a repeat chest X-ray is indicated.

Complications

A child who remains very unwell and febrile after 48 hours of parenteral treatment should be reassessed for the possibility of empyema or, more rarely, lung abscess. In these cases, input from infectious diseases and respiratory specialists is advised.

Prevention

The introduction of conjugate *H. influenzae* type b (Hib) and *S. pneumoniae* vaccines into the routine immunisation schedule in many countries has led to a reduction in the burden of pneumonia caused by these two organisms.[22]

Conclusion

Pneumonia is a common illness in children. Viruses are the most common cause, and many children will not require any specific treatment. *S. pneumoniae* remains the most common bacterial cause, and amoxicillin remains the antibiotic treatment of choice.

Controversies and Future Directions

There is increasing interest in the use of clinician-performed bedside ultrasound for the diagnosis of pneumonia in children. Potential benefits include the lack of ionising radiation and improved sensitivity compared to chest X-ray. However, the clinical utility of ultrasound is yet to be established. Most pneumonia – particularly in young children – is viral, and ultrasound has not been shown to reliably distinguish this from bacterial infection. In addition, many children with pneumonia need no imaging and can be managed as outpatients. Until further studies demonstrate clinical benefit from the use of ultrasound, it should not become a routine part of the ED assessment of children with potential pneumonia.

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6.8

Bronchiolitis

Tom Beattie, and Elizabeth Cotterell

ESSENTIALS

1 The diagnosis of bronchiolitis is clinical, based on history and examination.

2 Bronchiolitis typically affects children under the age of 12 months but may occur in children up to 2 years of age.

3 High-risk patients include those with underlying chronic lung disease, congenital heart disease or corrected age less than 2 months.

4 In the very young infant, apnoea may be the predominant symptom with few respiratory signs. Likewise, in neonates a non-specific sepsis-like picture with collapse may occur.

5 As feeding by an infant is an important index of bronchiolitis severity, a careful history of a change in feeds is paramount.

6 Oxygen saturations alone should not be used as a measure of disease severity.

7 The decision to admit to hospital is based mainly on considerations of the child's risk factors for disease severity as well as need for supplemental oxygen, fluids or cardiorespiratory monitoring for apnoea.

8 Cohorting patients on the basis of viral testing is not recommended.

9 Supportive care is the mainstay of treatment of bronchiolitis.

10 The role of chest radiography is limited and only indicated if the diagnosis is unclear or in severe cases.

Introduction

Epidemiology

Bronchiolitis is a common presentation to emergency departments (EDs), with a seasonal pattern. It typically affects children under the age of 12 months but may occur in children up to 2 years of age. The peak age is between 2 and 8 months of age, with males more commonly affected. Approximately 1% of children will require admission for bronchiolitis, which is the leading cause of admission for children with lower respiratory tract disease in the Western world.[1]

Epidemics of bronchiolitis occur during each winter, with the peaking of respiratory viruses. While respiratory syncytial virus (RSV) is the most common organism responsible for bronchiolitis, others include parainfluenza virus, adenovirus, rhinovirus and influenza. It is estimated that by the age of 2, 70% of children have been exposed to RSV. Despite the frequency, mortality is low at less than 1% of hospitalised babies.

High-risk patients include those with underlying chronic lung disease, congenital heart disease, neuromuscular disorders or corrected age less than 2 months. Some indigenous populations are prone to more severe disease.[2,3] Exposure to cigarette smoke and breast-feeding for less than 2 months are also associated with more severe disease.

Pathophysiology

Infection with RSV is associated with direct invasion of epithelial cells in the respiratory tract. Primary infection in young children and infants involves the lower respiratory tract. The bronchiolar epithelium is predominantly affected and an inflammatory response follows. Lymphocytes infiltrate the affected areas and oedema develops in the submucosa. Smooth muscle spasm ensues. The net result is that small airways become narrowed by the combination of oedema and muscle spasm, giving the typical clinical picture of bronchiolitis.

How this clinical picture emerges is still unclear. The role of pro-inflammatory regulators interleukin (IL)-6, IL-8, interferon-γ, and macrophage inflammatory protein-1β, as well as of the regulatory cytokine IL-10 in causing the disease as we know it, as opposed to facilitating healing and repair still remains to be elucidated.[4–6]

The incidence of concomitant or secondary bacterial infection is low, although otitis media may occur. There is an increased incidence of urinary tract

infections in infants with bronchiolitis aged less than 2 months of age presenting to hospital or hospitalised with a temperature over 38°C.[7]

In the very young infant, apnoea may be the predominant symptom, with few other respiratory signs. Likewise, in neonates a non-specific sepsis-like picture with collapse may occur. In reinfection with RSV or primary infection in older children, the symptoms are more limited to the upper respiratory tract, causing signs consistent with a severe common cold.

Clinical assessment

History

Bronchiolitis typically presents with a prodrome of upper respiratory tract infection over 1–2 days.

When the lower respiratory tract becomes involved, the hypersecretion of mucus causes the moist cough, onset of respiratory distress and resultant feeding difficulties. As the ability to feed in an infant is an important index of bronchiolitis severity, a careful history of a change in feeds is paramount.

Examination

Examination of the child will reveal a combination of signs of upper respiratory tract infection (URTI), along with signs indicative of lower respiratory tract infection (LRTI), which may fluctuate between examinations. The fever is usually low grade. A moist cough is common, and wheeze may be audible at the bedside.

Tachypnoea and tachycardia are usually in proportion to the illness severity.

Chest examination may reveal hyperinflation and recessions of the chest wall due to increased work of breathing. Paradoxically, as an infant fatigues, the recessions will decrease. In this situation the diminishing air entry signifies progressive disease. Auscultation reveals wheezes that are generally symmetrical. There may be inspiratory crackles. The auscultation findings are dynamic as coughing will move secretions to more proximal airways, with resultant temporary clearing of the wheeze.[8]

Oxygen saturations should be monitored to determine if they are sustained below 92% and indicate a need for supplemental oxygen. Decisions regarding managementshould not be based on oximetry alone, and continuous oximetry is not required for management of non-hypoxic infants (saturations >92%) or

stable infants receiving oxygen.[9] McIntosh graded severity of bronchiolitis by simply documenting children as needing no oxygen, requiring oxygen and needing ventilation.[10]

Table 6.8.1

Bronchiolitis severity and management

Severity	Signs	Management
Mild	Alert Feeding >50% normal Mild respiratory distress SaO_2 ≥92% NOT high-risk patient Age >6 weeks	Discharge home Smaller/frequent feeds Review in primary care
Moderate	Lethargic, tired Feeding <50% normal Marked respiratory distress Dehydrated SaO_2 <92% High-risk patient (see text)	Admit O_2 to keep SaO_2 92% Minimise handling Consider NG or IV fluids Close observation
Severe	As above but with: Increasing O_2 requirement Fatigue Signs of CO_2 retention Apnoeic episode	Cardiorespiratory monitor Consider blood gas measurement Liaise with PICU

ABG, arterial blood gas; NG, nasogastric; PICU, paediatric intensive care unit.

Assessment

In assessing an infant with bronchiolitis and the likely subsequent course, one needs to determine the onset of the respiratory distress or poor feeding phase of the illness. Most children have increasing work of breathing for 48–72 hours due to increasing secretions, before a plateau phase followed by resolution over 3–7 days. The cough may persist for a further 7–10 days after resolution of the respiratory distress.

In this way one can determine if a child is likely to deteriorate further, is probably stable at the peak of severity or improving at the time of the emergency department (ED) visit. It is the tiring consequence of the tachypnoea of bronchiolitis that impairs feeding ability, which is an important determinant of whether a child warrants interventions such as oxygen or fluids (nasogastric or intravenous).

Assessment of the child with bronchiolitis requires several components to be considered. Several scoring systems for bronchiolitis have been developed to determine the severity of the disease, but there is limited evidence of benefit for clinically relevant outcomes such as length of stay, need for admission or ICU admission or representation after discharge from the ED.[11] Table 6.8.1 shows the criteria that can be used to help determine severity and guide management.

Box 6.8.1 Differential diagnosis of bronchiolitis

Asthma in older infant
Cardiac failure
Pneumonia
Neonatal sepsis – presenting as collapse
Happy wheezer – persistent wheezing in undistressed baby

Differential diagnosis

The differential diagnosis of bronchiolitis includes cardiac failure, asthma in the young child and pneumonia (Box 6.8.1).

Cardiac failure can present with many of the features of bronchiolitis – dyspnoea, tachypnoea, tachycardia, crepitations and palpable liver. Feeding may also be poor. However, infants with bronchiolitis will usually have the URTI prodrome, and the onset of poor feeding is acute. The feeding difficulty in children with cardiac failure is less acute, leading to poor weight gain. Additional signs, such as a gallop rhythm or murmur, suggest an underlying cardiac abnormality.

Recurrent episodes of wheeze associated with URTIs, particularly in older infants, can be difficult to differentiate from asthma. Pneumonia in infants can mimic bronchiolitis, and the differentiation can be difficult. Some infants have persistent wheezing, which does not compromise activity or feeds, and is unresponsive to inhaled bronchodilators (‘fat happy wheezer’).

Investigations

The diagnosis of bronchiolitis is clinical, based on history and examination. The role of chest radiography is limited and only indicated if the diagnosis is unclear

or in severe cases. The typical radiological findings are hyperinflation of the lung fields, with bilateral increase in interstitial markings (particularly in the perihilar regions). There may be patchy atelectasis secondary to plugging or, in severe cases, collapse. Children with high fever or a clinical impression of sepsis may have superimposed bacterial infection, and a chest X-ray will aid exclusion of this diagnosis.[12]

Nosocomial infection and cross-infection can occur during bronchiolitis outbreaks. There is lack of evidence of any benefit of routine use of viral testing for cohorting of bronchiolitis.[13] The rapid testing for RSV may be useful in neonates to help with decision making where a child presents with fever, collapse or apnoea.

Other tests such as full blood examination are not useful in diagnosis. Severe cases may demonstrate CO_2 retention on arterial or venous blood gases, or electrolyte disturbance.

In febrile infants less than 2 months of age, consider collecting a urine sample for microscopy and culture, looking for the concurrent presence of UTI.[7]

Treatment

Supportive care is the mainstay of treatment of bronchiolitis. The decision to admit to hospital is based mainly on considerations of the child's need for oxygen, fluids, or cardiorespiratory monitoring for apnoea. Treatment options depend primarily on the severity of the disease.

Mild cases require an explanation to parents and advice regarding feeding to ensure adequate hydration. Smaller volume feeds offered more frequently to the child will usually ensure adequate hydration. Hence, the child who feeds every 3–4 hours normally should be offered feeds every 1.5–2 hours. Parents should monitor urine output (wet nappies) and should be advised to seek review should feeding deteriorate or urine output fall significantly. Some parents become exhausted by the constant demands of infants with bronchiolitis, and an assessment of parental coping should form part of the clinical picture. If there are concerns about parental ability to provide the increased frequency of feeding, admission should be considered.

Children with moderate bronchiolitis need to be admitted. The infant will need to be monitored clinically, with regular observation of heart rate, respiratory rate, ability to feed and level of fatigue. Oxygen saturation should be monitored, and supplemental oxygen should be provided to those with sustained saturations

below 92%. Oxygen may be administered by nasal cannula or mask. The choice of method of oxygen delivery needs to be tailored to the individual case.

The use of heated humidified high-flow nasal cannula oxygen for infants with moderate bronchiolitis has benefits in reduction of work of breathing, demonstrated by decrease in heart rate and respiratory rate, as well as reduced need for ICU admission.[14,15] Flow rates for initiation and escalation of treatment between 1 and 2 L mg min have been used, but evidence is limited for optimal rates.

In some infants who have self-limiting apnoeic episodes, continuous positive airway pressure may help buy time until the disease ameliorates. However, any persistence of apnoea or failure to maintain oxygenation despite escalation of treatment would warrant consideration of ventilation using endotracheal intubation and admission to an intensive care setting. Clearly, the choice of treatment will depend on available expertise and equipment. In all situations an increase in O_2 requirement to maintain saturations indicates increasing severity and the need to escalate therapy accordingly.

Feeding can be continued orally initially in those with moderate bronchiolitis, with small frequent feeds as above. If the child cannot tolerate this, nasogastric feeds or intravenous fluids should be considered. Volumes of fluid given should be judicious given risk of fluid overload, and intravenous fluids should be isotonic.

There is no indication for antibiotic use, including azithromycin,[16] for bronchiolitis unless there is good evidence of secondary bacterial infection.

Children with severe disease need to be admitted to a facility where they can be continuously monitored by appropriate staff so that ventilation can be considered should they deteriorate or have significant apnoea. Features of concern include the infant who is tiring, has escalating oxygen requirement or develops repeated and/or prolonged apnoeic episodes. Early discussion with the local paediatric intensive care unit staff will help to determine this need and to organise appropriate transfer or retrieval if required. Fluids should be given intravenously and adjusted according to volume status, urine output and electrolyte results. Children with underlying disease such as heart disease may need additional specific therapy.

Drug therapy

The role of various drug therapies in bronchiolitis is controversial and still

undergoing research.

Infants with bronchiolitis should not be administered $beta_2$-agonists as there is high-quality evidence that their use does not result in any change in the rate of hospitalisation, length of stay or oxygen saturation compared to placebo; they also cause adverse effects including tachycardia, hypertension and tremor.[17] Nebulised adrenaline (epinephrine) has similarly not been shown to provide benefit over a sustained timeframe with similar adverse effects.[18,19]

Systemic or inhaled glucocorticoids are widely used in some parts of the world, but the evidence for their use is variable and has not been shown to result in different rates of hospitalisation or length of stay.[20] The role of ipratropium bromide is equally unclear, and there is no clear-cut benefit to use.[21]

Nebulised adrenaline (epinephrine) and dexamethasone in combination have been shown in a multicentre trial in Canada to have some potential beneficial effect on the severity of the illness as determined by hospital admission[22]; however, adjustments for the study design showed no significant difference in admission rate.[23]

Nebulised hypertonic saline may be considered in infants with bronchiolitis presenting to the ED with evidence of reduced admission rates and no increased risk of adverse effects or readmission rates.[24] However, there are conflicting results from more recent meta-analyses suggesting no benefit on length of stay for hospitalised children and low-quality evidence for a reduction in admission rates.[25,26] Therefore, nebulised hypertonic saline is not currently recommended.

Ribavarin and antiviral immunoglobulins are not used in the ED setting but may have a role in high-risk groups in intensive care. Ribavirin is expensive and has only marginal benefit when given aerosolized to high-risk patients with severe disease.[27]

The role of immunisation against RSV is still under investigation. Palivizumab, a monoclonal antibody, has a limited role for prophylaxis in high-risk infants with, e.g., extreme prematurity and chronic lung disease of prematurity and some infants with congenital heart disease.

Prognosis

The vast majority of children with bronchiolitis will recover over 7–10 days. The cough may persist for some weeks after the resolution of the respiratory distress. A small number may continue to wheeze and cough for several months, and an indeterminate number will develop persistent wheeze and/or asthma. The exact

relationship between these events is unclear. Further study is needed to elucidate the role of RSV in the development of subsequent reactive airway disease.

Prevention

The transmission of RSV occurs with contact with infected secretions. In the ED setting, attention to hand washing, stethoscope hygiene and cubicle use is important to prevent cross-infection between children.

Controversies

Treatment of bronchiolitis has changed little over the years, with the mainstay being supportive care. A considerable amount of research needs to be done on the classification of the severity of disease based on sound, objective data, and agreement on important outcome measures for clinical trials. Without this, the debate on treatment with β-agonists, glucocorticoids, anticholinergic drugs and other therapies will persist, with anecdote continuing to supplant scientific data.

The indications for supplemental oxygen and high-flow oxygen therapy need to be determined to support decisions around initiation and discontinuation with effect on discharge practices.

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SECTION 7

Gastroenterology and Hepatology

OUTLINE

7.1

Abdominal pain

Jason Hort

ESSENTIALS

1 Only a small percentage of children presenting with abdominal pain have an underlying surgical cause.

2 The age of the child with abdominal pain significantly influences the diagnostic possibilities.

3 The younger the child, the less reliable are the clinical signs of acute appendicitis.

4 The abdominal examination findings, rather than blood tests, are the most important contributor in assessing the need for surgical consultation.

5 Providing adequate analgesia to the distressed child and an unrushed gentle approach improves the reliability of physical findings and does not mask the detection of peritoneal findings.

6 Abdominal pain in children may have intra- or extra abdominal causes.

7 Warning bells for a surgical problems include pain that genuinely requires opiate analgesia, constant pain over a number of hours, inconsolability, a child who remains motionless despite severe pain (likely peritonism), associated pallor or shock, reproducible localised tenderness, guarding, rebound, abdominal distension, rigidity, bilious vomiting and representation.

8 Close observation with serial examination remains very useful in indeterminate cases.

9 It is not always possible to make a definitive diagnosis in the

emergency department. A practical, systematic approach is required to exclude serious underlying possible causes. Discharged children in whom the diagnosis remains unclear need to have a clear action plan with early follow-up organised.

Introduction

Abdominal pain is a common reason for children to attend an emergency department (ED), occurring in up to 5% of all presentations in some institutions.[1] Most commonly the underlying cause is non-surgical, and surgery is required in only 1–7% of children who present with abdominal pain.[1,2] It is not possible to make a definitive diagnosis in all children with abdominal pain. In one study as many as 15% of children presenting to the ED with abdominal pain did not have a specific diagnosis at their discharge.[1] Some children arrive at the ED soon after the onset of symptoms, and it may take time, expectant management and review before a diagnosis becomes clearer or the symptoms of a self-limiting cause resolve. It is important, however, to exclude causes of abdominal pain that may require early surgical consultation, observation or investigations within the ED.

The priorities in managing children presenting with abdominal pain are:

- triage and early appropriate analgesia
- resuscitation with attention to ABCs as required
- diagnosis formulation using history, examination and investigation
- consideration of surgical review and management according to the likely diagnosis.

If a clear diagnosis cannot be reached in the ED, then exclusion of serious/life-threatening diagnoses is the priority. Subsequent disposition and follow-up are dependent on various factors including likelihood of a serious diagnosis; severity of the pain; availability of review; and psychosocial factors that may be contributory.

Pathophysiology

The sensation of abdominal pain is transmitted by either somatic or visceral afferent fibres.[3] Visceral pain from visceral peritoneum is poorly localised,

whereas somatic pain arising from parietal peritoneum or the abdominal wall is more localised. Referred pain also occurs due to visceral and somatic pathways converging in the spinal column. Two examples of referred pain are diaphragmatic irritation leading to pain at the shoulder tip due to convergence of visceral and somatic pathways at C4 and somatic pain from pneumonia leading to T10–11 pain sensed in the lower abdomen.[3] Abdominal pain may occasionally be found to be psychosomatic in origin after a thorough assessment of alternative causes.

Aetiology

There is a broad range of causes of abdominal pain in children, and one needs to initially keep an open mind regarding the diagnosis (Box 7.1.1). The age and sex of the child need to be considered, as well as features of the abdominal pain and associated symptoms and examination findings to determine the diagnostic possibilities.

Assessment

Each child needs initial assessment and triage with attention to securing the ABCs and providing appropriate early analgesia. This will help achieve a more reliable examination. A detailed history and thorough, gentle, and age-appropriate examination need to be performed.

Box 7.1.1 Causes of acute abdominal pain in children

Inflammatory gastrointestinal

- Appendicitis
- Meckel's diverticulum
- Mesenteric adenitis
- Gastroenteritis
- Food poisoning
- Peritonitis
- Peptic ulcer, gastritis
- Hepatitis

Pancreatitis
Inflammatory bowel disease

Non-gastrointestinal

Tonsillitis, pharyngitis
Pneumonia (especially basal)
Pericarditis
Serositis
Pyelonephritis, cystitis
Pelvic inflammatory disease
Intra abdominal abscess
Epididymitis

Generalised

Infectious mononucleosis
Acute rheumatic fever
Herpes zoster

Intestinal obstruction

Intussusception
Volvulus
Adhesions
Incarcerated hernia
Foreign body

Abdominal trauma

See Chapter 4.5

Gallbladder

Cholecystitis, cholelithiasis

Haematological

Leukaemia, lymphoma
Haemolytic crisis
Sickle cell disease
Neuroblastoma, Wilms' tumour

Endocrine

Diabetic ketoacidosis, hypoglycaemia
Adrenal insufficiency
Hyperparathyroidism

Vasculitic

Henoch–Schönlein purpura
Periarteritis nodosa
Kawasaki disease

Renal

Renal colic
Hydronephrosis
Nephrotic syndrome

Miscellaneous

Constipation
Colic
Toxic ingestion, e.g. lead
Torsion–testicular/ovarian
Ectopic pregnancy
Dysmenorrhea, Mittelschmerz pain
Mesenteric artery occlusion
Hypokalaemia
Acute intermittant porphyria
Familial Mediterranean fever
Abdominal migraine
Psychosomatic – including abuse

Source: Adapted from Rudolph 1996.

History

In considering a child who has presented with abdominal pain with no history of trauma, five important questions have to be addressed:

1. The age of the child

The age of the child helps narrow the diagnostic possibilities. The most common diagnoses to consider according to age are as follows.

Neonates and infants

They usually present with a change in behaviour to signify pain.[4] This may be persistent crying, irritability, inability to be consoled, fussiness, sleeplessness, and poor feeding.[4] Serious or potentially life-threatening conditions not to miss in this age group are listed in Table 7.1.1.

The diagnoses of acute gastroenteritis or ‘colic’ need to be made after excluding more serious causes.

Preschool

Common conditions include acute gastroenteritis, urinary infection, appendicitis, viral illness (mesenteric adenitis), pneumonia, constipation and trauma-related abdominal pain.

School age

Children of school age with abdominal pain usually have acute gastroenteritis, urinary infection, trauma, appendicitis, constipation, viral-related, psychosomatic pain or inflammatory bowel disease. In older females also consider gynaecological causes.

In the preschool and school-aged child, a common cause of abdominal pain in the primary care setting is constipation.[5] The diagnosis can be made with a careful history and examination and is suggested by fewer than three stools weekly, faecal incontinence, large palpable stools, retentive posturing, or painful defecation.[5] It is discussed in more detail in Chapter 7.13.

Table 7.1.1

Serious conditions not to miss in neonates and infants

Surgical causes	Medical causes
Testicular torsion	Diabetic ketoacidosis
Appendicitis	Toxic, e.g. iron ingestion
Peritonitis	Sepsis
Necrotising enterocolitis	Haemolytic uraemic syndrome
Volvulus	Urinary tract infection
Intussusception	
Hirschsprung disease	
Incarcerated hernia	

Adolescents

The causes of abdominal pain in adolescents expand to include various diagnoses of adulthood. In the female, pregnancy-related conditions must be considered, as well as other gynaecological conditions (see Chapter 15.2 on Adolescent gynaecology).

2. Whether this is the first episode or recurrence of abdominal pain

A history of similar pain in a child may help to clarify the diagnosis. However, it is important to remember that children with chronic abdominal pain due to myriad causes may develop an acute abdomen, and this fact needs to be considered at each presentation.

3. Whether there are other associated symptoms

Generally children with abdominal pain have other associated symptoms. A full symptom review is required, with particular reference to gastrointestinal symptoms. This includes vomiting, and whether it is bilious or blood stained, and the timing and quality of stool, including the presence of blood or mucus. The child with fever, voluminous diarrhoea and vomiting is likely to have gastroenteritis. However, particularly in young children, one must keep an open mind to other possibilities that can mimic or complicate gastroenteritis (see

Chapters 7.2, 7.8 and 7.12).

More general questions should assess the child's constitution – whether the child is febrile, lethargic, irritable, anorexic or has associated pallor. It is unusual for children with appendicitis to be hungry. The child with intussusception is often lethargic and pale. The presence of upper respiratory tract infection (URTI) or viral symptoms may suggest mesenteric adenitis. Dysuria or frequency may be reported in older children with a urinary tract infection. A typical rash or sore joints may indicate Henoch–Schönlein purpura as the cause of related vasculitic abdominal pain.

4. Whether there are any relevant pre-existing conditions

The child's past medical and surgical history should be fully explored. In older females an adolescent approach (see Chapters 15.2 and 30.1) and a menstrual and sexual activity history may be important. Family history and racial background may be relevant, along with a psychosocial history that may contribute if there is a suggestion of somatisation.

5. What are the characteristics of the pain?

These include location (generalised or localised), radiation, severity, quality (constant, episodic or of a colicky nature), and what improves and worsens the pain, along with the timing of the pain – gradual or sudden onset and whether the pain wakes the child. Direct questioning of the verbal child often augments the description of the characteristics of the pain as reported by a parent.

Pain that has a sudden onset may occur with perforated viscus, intussusception or torsion. The pain from appendicitis has a more insidious onset and usually increases over a period of hours. In both intussusception and mesenteric adenitis, the pain may cause episodic distress. Constant pain over a number of hours in a child usually suggests a significant underlying cause.

Examination

The abdominal examination of the young child with abdominal pain needs to be performed in an unhurried and gentle fashion. Toddlers may be better examined on the parent's lap. It is important to note the child's general appearance and any

features of toxicity. Children with colicky pain often writhe around, whereas the child with peritoneal irritation usually remains still as movement exacerbates the abdominal pain. Providing adequate analgesia improves the reliability of physical findings and does not mask the clinical detection of peritoneal findings. It is best to very gently approach the painful quadrant of the abdomen last in distressed children after pain has subsided post analgesia and the child has relaxed and become accustomed to the examination.

One of the keys to the assessment is to determine the presence of focal tenderness or true peritoneal irritation. In children this can sometimes be difficult, as many voluntarily guard the abdomen when examined, irrespective of the cause of the abdominal pain. This is expected when a previous examination may have been distressing. Differentiating the presence of true peritoneal signs may be helped by distraction techniques or by serial gentle examination over a period of time to determine true reproducibility of findings.

Eliciting rebound should begin with gentle palpation to avoid distress and resultant voluntary guarding. Signs consistent with peritonitis include refusing or being unable to walk, slow or stooped walking, or increased pain on coughing or movement, or the child lying motionless on the bed. Likewise, peritoneal irritation may be detected by asking if the child can expand or contract his/her abdominal wall by asking him/her to ‘suck tummy in, then let it out’. The younger the child, the less likely he/she is to have reliable localising signs of appendicitis, and the threshold for surgical review or observation needs to be adapted accordingly.

Important features of examination include the following:

- General appearance of the child
- Temperature and vital signs
- Jaundice
- Skin markings, e.g. abdominal bruising
- Abdominal distension
- Abdominal tenderness
- Guarding or rebound and/or rigidity
- Abdominal mass
- Bowel sounds
- Palpation of testes (in the male infant)
- Presence of inguinal herniae
- Urinalysis.

Rectal examination in children, when rarely indicated, should be performed once and ideally by the surgeon who may require the information. The interpretation of localised rectal tenderness is often difficult in children, as it is uncomfortable in all children and therefore does not often add to the assessment. The inguinoscrotal regions should always be checked for an otherwise occult hernia or torsion referring pain upwards to the abdomen. A sensitively performed, private, and chaperoned pelvic examination may be indicated in pubertal females.

The cause of abdominal pain in children may be extra abdominal. The child with ileus may have intra- or extra abdominal pathology including sepsis, urinary tract infection, pneumonia, or meningitis.

Important features in the examination of other systems that may present with abdominal pain include:

- respiratory – signs of basal pneumonia
- ENT – URTI, tonsillitis or adenopathy
- neurological – meningitis
- endocrine – blood glucose level in diabetic ketoacidosis
- haematological – pallor, lymphadenopathy
- dermatological – rash, particularly purpura/petechiae of Henoch–Schönlein purpura, zoster
- renal – oliguria, haematuria, hypertension in haemolytic uraemic syndrome.

Investigations

Many children have the diagnosis clarified by a physical examination alone. The need for investigations should be tailored to the individual case, where the diagnosis is unclear and the result of the test is likely to ‘rule in or rule out’ significant pathology.

Pathology

A full blood count may demonstrate leucocytosis, anaemia or film changes of haemolysis. However, the white cell count/differential is not particularly useful to determine if a child has acute appendicitis. Electrolytes, urea and creatinine assess changes resulting from fluid losses and impaired renal function. This may

be indicated in the child who has had significant losses from diarrhoea or vomiting. Liver function tests are indicated in children with potential hepatic and gallbladder pathology. The possibility of pancreatitis can be assessed by serum lipase levels. A blood glucose level and venous blood gas exclude diabetic ketoacidosis in the child presenting with abdominal pain who appears acidotic or has glucose and ketones in the urine. Females of reproductive age should have a βhCG performed and consideration of urine PCR testing for gonorrhoea and chlamydia. Urinalysis, microscopy and culture are necessary to exclude renal pathology.

Imaging

The routine use of an abdominal X-ray is unhelpful and not recommended to 'screen' children with abdominal pain. Specific situations where it may be helpful include demonstrating the signs of bowel obstruction; free air, suggesting perforation (although a chest X-ray may be more useful); and an ingested foreign body. Other abnormalities occasionally seen include calcification, which may represent a faecolith in the appendix and Meckel's diverticulum; 'thumbprinting', suggesting gut ischaemia or a sentinel loop adjacent to inflammation; or a soft tissue mass. Calcification may also represent renal stones, neuroblastoma or teratoma.

An erect chest X-ray may be indicated to demonstrate gas under the diaphragm or evidence of a basal pneumonia.[3]

An ultrasound study can help evaluate the liver, gallbladder and kidneys, as well as detect intussusception, features of appendicitis (if able to be visualised) or evaluate a palpable mass. Rarely, an abdominal CT scan may be indicated in selected cases; however, it is more often performed in the inpatient setting.

Point of care ultrasound (POCUS) by emergency physicians may have a role in the evaluation of suspected appendicitis and suspected intussusception, but further studies are required.[6]

Management

The initial management of the child with severe abdominal pain should include assessment and securing of ABCs and administration of appropriate analgesia to relieve the child's distress. Children with similar pathophysiology can have markedly varied distress levels, and analgesic administration needs to be

individualised.[4] Concurrent anxiety may increase painful stimuli, and this can be lessened by involving parents in providing comfort and using a child-friendly environment to distract and help calm the child. Using a visual analogue scale to evaluate severity of pain may be helpful to assess response to analgesia.

There is no contra-indication to providing adequate analgesia for any child presenting in pain. It is much easier to perform a reliable examination on a child who is made comfortable. For severe distress, intravenous morphine titrated in increments controls most children's pain and will not mask the abdominal signs. Intra-nasal fentanyl is a useful alternative for rapid onset analgesia (2–3 minutes) with the advantage of not requiring venous access, but its short duration of action (30–60 minutes) means longer-acting analgesia will be required if pain is ongoing.[7] Oral agents such as paracetamol, codeine, oxycodone or ibuprofen may be used in less severe pain. Serial examination of the child's abdomen and observation of vital signs over a period may be important to exclude significant pathology. Children with a potential surgical cause should be kept nil by mouth, until surgical review.

Disposition

Children with significant abdominal findings need to have a surgical consultation. Not all children who present with abdominal pain will have a clear definitive diagnosis after their assessment has excluded a serious cause or the need for a surgical review. Some children with severe pain, but negative physical examination findings, where the diagnosis is not clear, will warrant admission for ongoing observation. Children who are discharged home need to have clear instructions given to parents regarding returning, should they deteriorate, and have a planned timely follow-up by the local doctor.

Acute Appendicitis

Introduction

Appendicitis is the most common non-traumatic surgical emergency in children. It occurs slightly more commonly in males at any age, although it is uncommon in those under 2 years and very rare in neonates. The peak incidence is at 9–12 years.

Clinical features

The clinical features of classic appendicitis are well known. Pain is felt initially in the periumbilical region due to visceral pain from obstruction of the appendix. There is often associated nausea, vomiting, anorexia and a low-grade fever. Later, there is migration of the pain to the region of the appendix. This more intense right lower quadrant pain results from irritation of the parietal peritoneum. Up to 50% of adults have this progression of symptoms, but it is less common in children.

Importantly, some children may have false-localising diarrhoea or dysuria caused by irritation from an inflamed appendix. Fever is generally below 39.5°C, unless perforation has occurred. Asking the child to walk or hop to demonstrate pain with right leg movement may be useful in indeterminate cases to reveal the presence of true peritoneal irritation. Likewise, manoeuvres such as the iliopsoas (extension of the right hip), obturator (internal rotation of the right thigh) or Rovsing's (pain in right lower quadrant with palpation of left lower quadrant) signs may help confirm suspicion of appendicitis.

In general, rectal examination in children should not be performed by ED staff. In children with clear signs of appendicitis, the rectal examination adds little value, is distressing for a child and does not alter management.[8] In children without clear signs of appendicitis, a rectal examination is insufficiently sensitive or specific to be useful.

Young children are less likely to present with a typical clinical picture and are at risk for delayed diagnosis and high (>80%) perforation rates. Under the age of 2 years, vomiting (85–90%) and pain (35–77%) are the most common symptoms, with diarrhoea (18–46%) and fever (40–60%) less common. Sometimes grunting respirations (8–23%), cough or rhinitis (40%) and right hip symptoms (3–23%) may be misleading. Right lower abdominal tenderness is present in less than 50%.

As children become older, right lower abdominal tenderness becomes more common (age 2–5 years, 58–85%), up to nearly all children in the school-age group, with some (15%) having generalised tenderness without perforation. In children 6–12 years, vomiting occurs in 68–95% of children, anorexia in 47–75%, diarrhoea in 9–16%, constipation in 5–28% and dysuria in 4–20%.

Differential diagnoses

These include mesenteric adenitis (less severe pain without peritoneal signs is usual, and it is rare under 3 years of age), bacterial enterocolitis, pelvic inflammatory disease, urinary tract infection, Meckel's diverticulitis, intussusception and right lower lobe pneumonia.

Investigations

Children with a clear clinical diagnosis of acute appendicitis do not require investigations and delay of surgical consultation. In equivocal cases, imaging of the appendix may be helpful or demonstrate alternative causes of the pain.

No single test is diagnostic in appendicitis, with the white blood cell count insensitive and non-specific.[9] C-reactive protein (CRP) levels >10 mg dL^{-1} have varying reported sensitivities (48–75%) and specificities (57–82%) in different studies on appendicitis. Normal CRP values do not exclude acute appendicitis in children.[9] On urine microscopy more than five white blood cells per high power field or the presence of red blood cells is found in 7–25% of children with appendicitis. Abdominal X-rays may show other pathology (e.g. right lower lobe pneumonia) and occasionally an appendiceal faecolith but are also insensitive and non-specific for diagnosing appendicitis. Note the rare presence of an appendolith can give a more colicky nature to the pain.

New biomarkers are being investigated to aid in the diagnosis of appendicitis including leucine rich alpha 2 glycoprotein (LRG), granulocyte colony stimulating factor, leukocyte gene expression profiles, and plasma cytokine levels. None are in routine use, but some show promise.[10]

Ultrasound

Ultrasound has reported sensitivity of 71–92% with specificity of 86–98% and is often used when there is initial diagnostic doubt. However, in one series, patients undergoing sonography before appendicectomy, compared to those with clinical assessment alone, had a longer delay before operation, a higher rate of misdiagnosis, and more post-operative complications.[11] It is not uncommon for the appendix to be difficult to visualise (up to 10% of cases). High positive likelihood ratios and moderate negative likelihood ratios suggest that ultrasound can be used to confirm but not exclude a diagnosis of appendicitis.[9]

Point of care ultrasound (POCUS) is an area of current interest, but there is little published evidence yet to clarify the role of POCUS for suspected appendicitis in children.[6]

CT scan

Abdominal helical CT scan in diagnosing appendicitis shows sensitivities from 87% to 100% and specificities from 83% to 97% with signs of distension of the appendix, faecolith, focal caecal wall thickening and fluid collections in ruptured appendicitis. Some institutions have used a protocol incorporating ultrasound and subsequent CT scan with patients whose diagnosis is equivocal. They have shown this to be an accurate and cost-effective approach when compared with a negative appendicectomy rate of 23%. The American College of Radiology in general advocates the use of ultrasound over CT scan. Concerns persist over radiation exposure to children from abdominal CT.[10]

Magnetic resonance imaging

Recent studies suggest high sensitivity and specificity in adult patients with appendicitis.[12] However, several disadvantages including high cost, long duration of study, and limited availability mean MRI has a limited role at the moment. It appears potentially useful in pregnant patients with suspected appendicitis in whom ultrasound is inconclusive.[9]

Active observation

Active observation with hospital admission and frequent review by a surgical doctor with observations, examination and analgesia requirements have also been used without investigation, with a positive predictive value of 97.9% in one series and a normal appendicectomy rate of 2.6%.[13]

Management

The standard management of acute appendicitis is appendicectomy. Some children require intravenous rehydration, ongoing analgesia and antibiotics, if perforation is suspected. Recent studies have suggested medical management may be an alternative to operative appendicectomy in children with uncomplicated appendicitis. Further study results are awaited to clarify the role of a non-operative approach in children.[10]

The management of the child in whom the diagnosis is not clear, but may possibly be early appendicitis, is more difficult, and one needs to have a clear approach.

In some children it may be useful to perform ultrasound imaging of the appendix, particularly in older females who may have ovarian pathology as a

cause of their pain. An expectant approach is appropriate in some children with a short history, with a clear plan of organised review over the next 6–12 hours. This may be achieved by actively observing the child in hospital or, if discharged, by arranging a definite early clinical review. Parents need clear instructions to return should their child's symptoms progress.

Recent literature[14–16] has evaluated the use of scoring systems (such as the Pediatric Appendicitis Score [PAS], Alvarado score, Children's Appendicitis Score [CAS]) as decision rules in estimating the likelihood of appendicitis. These rules, along with the incorporation of ultrasound into clinical pathways, may assist in refining ED management for children with possible appendicitis.

Meckel's Diverticulum

Introduction

Meckel's diverticulum is a vestige of the omphalomesenteric duct, occurring in 2% of the population, with 2% of those with Meckel's diverticulum manifesting symptoms. The diverticulum is usually 60 cm (2 feet) proximal to the terminal ileum. Of symptomatic patients, 45% are under the age of 2, but it can present at any age. These findings are known as Meckel's rule of twos. The majority of Meckel's diverticuli contain gastric mucosa and may secrete acid.

Clinical features

The presenting features of a Meckel's diverticulum vary and may include gastrointestinal haemorrhage (40%), bowel obstruction (30%) and diverticulitis (20%) leading to abdominal pain or perforation.[17]

Meckel's diverticuli are the most common cause of significant lower gastrointestinal bleeding in children, usually from peptic ulceration within the diverticulum or adjacent ileum. It is classically painless bleeding, with stools either bright red or tarry, depending on the site and briskness of bleeding.

Diverticulitis causing crampy lower quadrant pain can occur in older children. A Meckel's diverticulum can be a lead point in intussusception, as well as causing intestinal obstruction by the formation of intraperitoneal bands, which can lead to the possibility of a volvulus or internal herniation.

Differential diagnoses

These include intestinal polyps, intussusception, anal fissures, midgut volvulus and bacterial enteritis.

Investigations

A Meckel's scan using ^{99m}technetium can be used, which is 75–85% sensitive and 95% specific[18] at demonstrating the ectopic gastric tissue, when bleeding is present.

Management

Management is intravenous fluid support with blood transfusion for massive bleeding. A Meckel's scan can be done in stable bleeding patients, but if unstable or with peritoneal signs, urgent surgical consultation must be obtained with a view to operative intervention. Definitive treatment is surgical excision, which may be done laparoscopically.

Chronic Abdominal Pain

Introduction

Chronic abdominal pain is defined as the presence of at least three discrete episodes of pain occurring over a period of 3 months or longer.[3] The reported prevalence of abdominal pain interfering with activities is 10–15% in children between 5 and 14 years. Causes of chronic abdominal pain are diverse and are listed in Box 7.1.2.

Signs and symptoms suggesting *organic disease* causing chronic abdominal pain in school-aged children include:

- persistent fever
- poor weight gain or weight loss
- child awakened from sleep
- pain away from the umbilicus
- radiation of pain to back, shoulder or lower extremities
- persistent regurgitation, vomiting or dysphagia
- bloody emesis or stools
- associated altered bowel pattern
- perianal disease

- sleepiness following pain attacks
- positive family history of peptic ulcer or inflammatory bowel disease.

Assessment

The assessment of the child with chronic abdominal pain involves a detailed history and examination. Parents often present to hospital in an attempt to reach a diagnosis, may have seen many different doctors, and may be frustrated with the lack of a clear explanation for their child's pain.

Blood pressure should be measured. The child who has no features to suggest an organic origin requires no testing, although some clinicians opt to perform a limited screen involving urinalysis, full blood count and erythrocyte sedimentation rate and stool testing for ova and parasites and occult blood.

Diagnosis

The diagnosis of recurrent functional abdominal pain is based on history and a normal physical examination. Usually the patient has no worrying features (as listed above), has periumbilical or mid-epigastric pain, and rarely wakes at night from the pain. Psychosocial stressors may be evident, including bullying, illness of a family member, moving school, or parental separation. There may be secondary gain from the child's abdominal pain.

Management

Reassurance is the treatment of choice, although it is important to acknowledge that the child does experience pain. Cognitive–behavioural therapy may be useful for children who clearly have recurrent functional abdominal pain.[19]

Box 7.1.2 Causes of chronic abdominal pain

Gastrointestinal causes

Chronic recurrent functional abdominal pain
Peptic ulcer disease
Irritable bowel syndrome
Inflammatory bowel disease
Chronic or recurrent pancreatitis

Biliary colic
Appendiceal colic
Constipation
Partial bowel obstruction
Parasitic infection

Endocrine disease

Hyperparathyroidism
Addison’s disease
Diabetes mellitus

Cardiovascular disease

Superior mesenteric artery syndrome
Coarctation of aorta

Neurological disease

Abdominal migraine
Migraine headaches
Familial dysautonomia

Haematological disease

Sickle cell disease
Porphyrias

Gynaecological disorders

Cystic teratoma of ovary
Endometriosis
Haematocolpos
Mittelschmerz

Musculoskeletal disorders

Discitis
Linea alba hernia
Painful rib syndrome
Muscle wall sprain

Other

Uteropelvic junction obstruction
Familial Mediterranean fever
Hereditary angioneurotic oedema

Source: Adapted from Rudolph A. *Rudolph's Textbook of Paediatrics*. 20th ed. New York: Appleton and Lange; 1996.[3]

In the ED situation, the management of chronic abdominal pain depends on the possible diagnoses. Follow-up should be ensured, particularly in the child with the suggestion of organic disease or the child missing a significant amount of school. Encourage non-pharmacologic methods of pain relief and a symptom diary and recommend that the parents persist with a single medical point of contact – a trusted GP or paediatrician.

Controversies

1 The best approach to investigations in appendicitis has not yet been settled.
2 There is probably little need for a rectal examination in children with abdominal pain, but more research is needed before making definitive conclusions.

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7.2

Vomiting

Christopher Webber, and Andrew John Anderson Holland

ESSENTIALS

1 ‘ABC fluids in and out’ is a useful tool in taking a history.
2 Not all vomiting and diarrhoea are gastroenteritis.
3 Vomiting may be a non-specific symptom of a more serious medical problem.
4 The differential diagnoses are extensive.
5 Young infants with vomiting, especially with fever, must be managed with caution.
6 Bile-stained vomiting generally indicates intestinal obstruction due to surgical pathology.
7 Early referral to a paediatric surgeon is essential for bile-stained vomiting.
8 Rarely bile-stained vomiting may have a non-surgical, medical aetiology.
9 Malrotation with volvulus usually presents in the first few weeks of life but can occur at any age and is always a surgical emergency.
10 Small bowel obstruction secondary to adhesions is less common in children than in adults but rarely settles with non-operative management.

Non-Surgical Vomiting

Introduction

Vomiting is a common symptom that affects infants and children. It may occur in isolation or with diarrhoea. Clinicians must exercise caution when evaluating infants presenting with fever and vomiting as the list of differential diagnoses is extensive, including some conditions with significant morbidity and mortality.

By far the commonest condition causing vomiting and diarrhoea in children is gastroenteritis (Chapter 7.12). A urinary tract infection (UTI) must be considered in all infants with vomiting, with or without fever (Chapter 16.4). In older children dysuria would assist identifying those children likely to have a UTI. The presence of diarrhoea makes a urine infection much less likely, and in this situation the diagnosis is more likely to be gastroenteritis.

Definitions

Vomiting is an active process involving muscular contraction expelling the stomach contents orally. In comparison gastro-oesophageal *reflux* (Chapter 7.4) is the passive regurgitation of gastric contents, most commonly liquid. In the newborn and young infant gastro-oesophageal reflux is common and may be considered normal if it is not associated with consequences like pain, weight loss or failure to thrive.

Bile-stained vomitus generally indicates intestinal obstruction. Bile in the gallbladder is amber coloured, like golden syrup'. It changes colour and becomes green in the small bowel; thus vomiting bile that is green coloured is highly suggestive of intestinal obstruction.

Diarrhoea refers to increased stool frequency and the consistency of stool being loose or liquid. The stool may also contain blood and mucous, both of which are abnormal.

Fever is present when the body's temperature is elevated above normal. Actual definitions of fever vary, but a core temperature of about 36.5–37.2°C is normal. The site used to make the measurement also affects measured temperature. Generally aural (tympanic) temperatures are less accurate. Per-axilla temperatures are lower than rectal temperatures by about 0.5–1°C. Although rectal temperatures more closely reflect the body's core temperature, per-axilla temperatures are more convenient to measure and socially acceptable to the patient. Needless to say, an infant with cardiovascular compromise and peripherally 'shutdown' should have a rectal temperature checked, as a per-axilla

temperature may not accurately reflect core temperature and falsely reassure the physician.

Clinical evaluation

History

Presenting complaint and past history

Generally, the parents or carers know the infant or child best, and physicians are unwise to ignore their concerns. When taking a history consider the age of the infant or child. Explore the symptoms described in further detail if necessary. Is there blood in the vomitus or blood or mucus in the stool? Is the infant breast or formula fed? If so, which formula? What is the diet of the older child? Has there been a recent change to the milk or diet and why? Sometimes important associated symptoms are not volunteered by parents and are only alluded to when directed by a focused history. In the child with a possible infectious condition, it may be important to clarify whether there have been infectious contacts or recent overseas travel. Review of the past history should include any significant medical problems or surgical procedures and whether the child is thriving.

ABC – fluids in and out

This approach is useful for both clinicians and parents. Alertness and activity (**A**) provides information about the neurological state of the child. Lethargy and poor interactivity are concerning symptoms especially in a young child or infant. The behaviour of the baby whilst feeding, noting the quality of the suck and the alertness of the baby, provides helpful information. Breathing (**B**), specifically rapid or laboured breathing, and poor circulation (**C**), as indicated by mottling and coolness of the peripheries, are also worrying signs.

The child's fluid intake and urine output as a percentage of his/her normal provide very important markers of the unwell infant or child. The number and 'wetness' of the nappies, often indicated by the perceived weight of the nappy, is used as the indicator of urine output. Thus, using an evaluation of 'fluids in and out', one should be concerned when these are less than 50% of normal for the child.

What is the nature of the vomitus? Blood staining may be coffee grounds or fresh blood. Vomiting bile that is green coloured is highly suggestive of

intestinal obstruction. Severe retching or repeated vomiting followed by yellow-coloured vomitus may well be refluxing bile from the second part of the duodenum into the stomach and not necessarily reflect intestinal obstruction.

Examination

General observation

In the absence of a life-threatening emergency, observe the child's reactions whilst talking with the carer or to the child. General observation is invaluable in the paediatric assessment, particularly in young children. The ABCD approach (airway, breathing, circulation, disability) can be utilised, even subconsciously, for the child who isn't desperately unwell.

Assessment of the state of hydration is critical. Accurate premorbid and current weight would aid this assessment. As a consequence of clinicians tending to overestimate the degree of dehydration, the guide for the assessment of dehydration has been revised to:

- <3% – no clinical signs; reduced urine output
- 3% – mild dehydration; mild tachycardia, dry mucous membranes
- 5% – moderate dehydration; lethargy, tachycardia, reduced skin turgor, sunken eyes/fontanelle
- 10% – severe dehydration; clinical signs of shock (tachycardia, thready pulses, reduced perfusion, particularly centrally).

Always check carefully for a rash, as petechiae and purpura may be inconspicuous or subtle. Examination of the ears, nose and throat is frequently left until the end of the examination, to avoid potential distress impairing the remainder of the examination.

Cardiovascular and respiratory status

It is important to evaluate the cardiovascular and respiratory systems in the infant with vomiting for the following reasons:

- Shock may be present due to either septicaemia or fluid loss from vomiting and/or diarrhoea
- Septicaemia may be the cause of the symptoms

- Pneumonia or other respiratory infections may cause vomiting
- Effortless tachypnoea may indicate a metabolic acidosis.

The pulse rate and pulse volume may identify a rapid thready pulse indicating poor perfusion. Capillary refill is often thought to be a poor indicator of circulatory status, as the peripheral perfusion (hands and feet) may be affected by environmental temperature. Comparing central (anterior chest) and peripheral capillary refill with other data like heart rate, pulse volume and conscious state allows for a more informed assessment of the adequacy of the circulation.

The presence of shock indicates inadequate tissue perfusion. It is present when there is a rapid, thready pulse, delayed capillary refill, especially if central (≥3 seconds), and abnormal neurological status including agitation, lethargy, or coma. The diagnosis of shock is not reliant on the presence of hypotension, particularly in children. Delay in identifying shock until the child is hypotensive risks severe compromise and potential progress to cardiac arrest.

Respiratory examination of the infant with fever, cough and vomiting may identify tachypnoea and grunt that alert the clinician to the possibility of pneumonia. Grunting is an expiratory noise, usually intermittent, and generates PEEP (positive end-expiratory pressure) by partially closing the glottis during expiration. Percussion note for dullness may be more valuable than auscultation for crackles or bronchial breathing. Be aware of the child with vomiting, abdominal pain and grunt as he/she may have lower lobe pneumonia.

Abdominal examination

The abdomen is examined for distension, scars and any skin changes, including rash or signs of trauma. Palpate for tenderness and guarding. Be mindful of pain and tenderness and observant of the child's resting posture and how he/she moves into and out of bed or walk. A gentle, unrushed examination provides more information. Watch the small infant's face as you examine the abdomen looking for subtle grimacing indicating tenderness.

Presence of guarding and rebound can be elicited with gentle flexion/relaxation of the fingers of the examining hand (at the metacarpophalangeal joint). Note any organomegaly of the liver, spleen or kidneys. Are there any masses? The pale, vomiting child may have intussusception, even without the classic intermittent, severe, cyclical pain. Examine for anal fissures and other perianal abnormality that may indicate

inflammatory bowel disease. Examine the testes for torsion and the groins for herniae.

Neurological examination

The child may have altered consciousness caused by systemic illness, shock or a primary neurological problem. The unwell infant who is vomiting may have meningitis. Likewise, raised intracranial pressure (ICP) may cause vomiting. Cushing's triad includes bradycardia, hypertension and abnormal respiration but is generally associated with a marked elevation of the ICP. On neurological examination, check for any asymmetry of movement, tone and reflexes, as a cerebral abscess may cause fever and vomiting. Focal seizures generally indicate focal pathology and should be considered the same as a focal abnormality on examination.

Temperature

Always measure the temperature of the vomiting child. The critically unwell child should have his/her temperature continually monitored with a rectal probe. The body's temperature varies over time, and in the unwell patient it is crucial to continue to monitor for fever over a period of time. Remember that some modes of measurement either underestimate the core temperature or are inaccurate. Tympanic measurements are generally unreliable. Although checking for fever, the child with overwhelming sepsis may indeed be hypothermic.

Differential diagnoses

This list is extensive and varies depending upon the combination of symptoms, vomiting and/or fever and/or diarrhoea, as well as the examination findings:

- Gastro-oesophageal reflux
- Viral illness:
    - Gastritis/enteritis
    - Non-specific, generalised viral illnesses
- Gastroenteritis – viral, bacterial
- Otitis media/URTI/tonsillitis/pharyngitis/stomatitis
- Urinary tract infection (UTI)
- Septicaemia

- Meningitis/encephalitis
- Pneumonia
- Appendicitis
- Metabolic/endocrine problems
- Intestinal obstruction including malrotation (± volvulus)
- Intussusception
- Cyclical vomiting.

The child with recurrent, non-bile-stained vomiting warrants particular attention, with consideration of diagnoses like malrotation, UTIs and cyclical vomiting. An abdominal X-ray (AXR) may be helpful, but after surgical consultation an upper gastrointestinal contrast study may be required. Be mindful that a normal AXR does not exclude surgical pathology.

A child thought to have cyclical vomiting should be referred to a paediatrician for follow-up. Beware of children with a pre-existing condition (or diagnostic 'label') such as cyclical vomiting as they may develop other acute conditions. These children require careful reconsideration at each presentation before concluding 'another vomiting episode'.

Investigations

The general principle of only ordering tests that aid the decision making and management of the patient is sound. Results outside the normal range may not necessarily indicate abnormality or pathology but just reflect standard deviation of the test and the normal population. Consequently abnormal results may inadvertently lead the clinician to order more unnecessary tests.

Not every child who is medically assessed requires investigations. The tests required depend on the possible differential diagnoses and the severity of illness (Box 7.2.1). Don't forget simple bedside tests like urine analysis (UA) and bedside blood glucose. This information can be invaluable in assessing the child with diarrhoea and vomiting. In this context, check the UA for the specific gravity and for ketones. Glycosuria and ketonuria may indicate diabetic ketoacidosis. If the UA is positive for nitrites or leucocytes, then an appropriately collected urine specimen should be cultured if a UTI is possible. The specimen to be cultured should be collected attempting to minimise contamination, even if re-collection is required.

Box 7.2.1 Tests that may be useful in the child with vomiting or diarrhoea

Common tests:

- Urine analysis
- Urine culture
- Stool:
    - Microscopy, culture
    - Rotavirus antigen
    - *Giardia* (often tested by RIA, avoiding the need for multiple samples for ova and parasites)
- Biochemistry – electrolytes, urea/creatinine, blood glucose
- Full blood count

Less common tests:

- Liver function tests
- Blood gas (venous or arterial)
- Blood culture
- Lipase
- Insulin, growth hormone, cortisol (if hypoglycaemic)
- Lactate
- Urine metabolic screen
- Chest X-ray to exclude pneumonia
- Abdominal X-ray to exclude intestinal obstruction

RIA, radioimmunoassay.

In acute gastroenteritis (i.e. when there is diarrhoea) urine culture is generally unnecessary but is mandatory in the infant or baby with acute vomiting and fever. Urinary symptoms like dysuria and frequency can be expected in children from about 2 or 3 years of age, allowing a more selective approach to those who require urine culture. ‘Bag urines’ (collected in an adhesive, sterile plastic bag) have a high contamination rate and are best avoided when performing a urine

culture but are satisfactory for a simple UA. Although more difficult to collect, a ‘clean catch urine’ provides a sample less likely to be contaminated, especially after perineal cleaning. This is collected with an open nappy, a poised parent, with an open sterile container, waiting patiently to ‘catch’ the spontaneously voided urine. Those babies and infants deemed to be at higher risk of a UTI or who are more seriously unwell may require more rapid testing by either a suprapubic aspirate (‘bladder tap’) or an in–out catheter urine collection. Any child who is toilet trained and requires a urine culture should have mid-stream urine collected.

The child who is previously well and thriving, and usually tolerates illness well, who presents with vomiting, hypoglycaemia and ketonuria is likely to be hypoglycaemic from starvation. In the absence of ketonuria/ketonaemia, metabolic and endocrine abnormalities need to be considered. Critical blood tests and specific urine tests are required. Chapter 10.2 describes a detailed approach to the assessment and management of a non-diabetic child presenting with hypoglycaemia.

Table 7.2.1

Antiemetic medications for children

Medication	Dose	Comments
Ondansetron (wafer or orally dissolving tablet)	2 mg if 8–15 kg 4 mg 15–30 kg 8 mg if >30 kg Administer 6–12 hourly	Not recommended for children aged <6 months or weighing <8 kg Should not be administered repeatedly without medical review Can cause headache, constipation, prolonged QT interval Dystonic reactions rare
Metoclopramide	0.15–0.3 mg kg (adult 10–20 mg) 6 h IV, IM or oral	High risk of akathisia and dystonic reactions
Promethazine	0.2–0.5 mg kg (adult 10–25 mg) 6–8 h IV, IM or oral	Sedation, reduced seizure threshold, anticholinergic and extrapyramidal effects
Droperidol	0.02–0.05 mg kg (adult 1.25 mg) 4–6 h IM or slow IV	Sedation and extrapyramidal effects
Prochlorperazine	Oral: 0.2 mg kg (adult 5–10 mg) 6–8 h IM or IV: 0.2 mg kg (adult 12.5 mg) 8–12 h	Sedation, prolonged QT interval, extrapyramidal and anticholinergic effects

Source: Royal Children’s Hospital, Melbourne, Australia, Clinical Practice Guideline on Drug

Doses, [Internet; cited 18/12/2016], Available from: http://www.rch.org.au/clinicalguide/index/cfm.

In a young, breast-fed infant with blood in vomitus or stool it is important to distinguish swallowed maternal blood from infant's blood. An Apt test distinguishes between the two due to presence of HbF in the infant's blood and absence in the maternal blood. To perform the test, add the bloody stool/vomitus to a test tube with about 5 mL of tap water. This lyses the red blood cells. After allowing the liquid to settle, the 'supernatant' should be pink. If so, add 1 mL of 1% sodium hydroxide. Remaining pink indicates infant's blood (HbF) whereas changing to a yellow–brown colour over 2 minutes indicates maternal (adult) blood.

Management

Antiemetic medication

Treatment of the underlying cause is essential. However, the administration of medication to relieve the symptoms of nausea and vomiting will make the child (and carers) feel better and allow the child to cooperate more fully with assessment. Options for antiemetic management in children are presented in Table 7.2.1.

Most antiemetics have side effects and are not considered suitable for children aged less than 6 months. The risks of antiemetic administration (masking a potentially significant progression of the child's underlying illness or medication-induced side effects) need to be weighed up against the potential symptomatic relief.

Intravenous fluids

Fluid management will depend on the need for immediate resuscitation and, subsequently, the provisional diagnosis. A **fluid bolus** is used to restore circulating volume and is therefore warranted when signs of shock are present. This must not be confused with strategies of 'rapid rehydration' in dehydrated children, generally used in children with gastroenteritis. Intravenous fluids containing dextrose must not be used as volume expanders. Crystalloid fluids for a bolus include **0.9% sodium chloride (normal saline), Hartmann's solution and Plasma-Lyte 148®**.

In a dehydrated child who does not have shock, it may be safer to commence

initial intravenous rehydration with fluids that contain at least 140 mmol L^{-1} sodium. Hypoglycaemia is not infrequent, both at presentation or subsequently; thus glucose-containing fluids are appropriate in all children, particularly in the young infant or those who have had insufficient caloric intake and are ketotic. In this context, **0.9% NaCl + 5% glucose** or **Plasma-Lyte 148 + 5% glucose**® are appropriate intravenous fluids to use.

Be aware of the potential of SIADH in unwell children and *avoid administration of hypotonic fluids* (such as 0.45% NaCl + 5% glucose). The rate of fluid administration may need to be altered depending upon the initial biochemistry, the diagnosis and the clinical progress of the child. Monitoring fluid balance is essential in the unwell infant or child.

Nasogastric rehydration is commonly used in dehydrated infants with gastroenteritis (see Chapter 7.12).

Antibiotics

Any specific management like administration of antibiotics depends on the working diagnosis and the severity of the illness, e.g. meningitis, UTI and septicemia. Clearly it is preferable, but not always feasible, that appropriate cultures be taken prior to administration of antibiotics; although in the severely unwell, febrile patient broad-spectrum antibiotics should not be delayed whilst awaiting collection of all cultures. Even in this setting, however, it is highly preferable, but not essential, that at least blood cultures be undertaken prior to antibiotic administration.

Consultation

Consultation with more experienced staff, either emergency physicians or paediatricians, should be encouraged, especially for complex or severely ill infants or children. Assistance may be required for clinical evaluation and technical procedures in critically ill children.

Transfer

Local, experienced, senior clinical consultation and review should occur prior to transfer. The child who requires transfer to a facility that provides a higher level of care needs an experienced evaluation regarding the level of escort, urgency

and mode of transport required to safely achieve the transfer.

Conclusions

Differential diagnostic possibilities

For each patient the differential diagnoses vary depending on the constellation of symptoms (vomiting, diarrhoea or others). Keep an open mind, and always re-evaluate clinical data. Don’t be distracted by a child’s previous diagnoses or labels.

General approach

‘ABC fluids in and out’ is a useful tool in taking a history. Caution must be exercised in the infant who has fever and vomiting, as differential diagnoses include serious illnesses, like meningitis. The younger the infant, the more challenging the evaluation and the less reliable the clinical examination are. If in doubt, seek consultation from a colleague. The telephone is a useful and powerful tool for clinicians, and the value of consultation should not be underestimated.

Surgical (Bilious) Vomiting

Introduction

Vomiting in a child remains a common symptom familiar to all parents, caregivers and medical providers. In many, if not the majority, a benign, self-limiting illness represents the likely cause. It is important to quickly determine the nature, duration and frequency of vomiting, as this will significantly impact on the likely aetiology and need for and timing of further investigation and treatment. It is especially important to rapidly ascertain whether the vomiting has been bilious or non-bilious, with the former more likely to require prompt surgical assessment and intervention.

Any child presenting to the emergency department (ED) with bilious vomiting has a surgical cause of intestinal obstruction until proven otherwise. Bilious vomiting occurs when the vomitus contains bile. Typically this is indicated by its bright green colour, although the discolouration may vary from a pale yellow to a dark greenish brown. Often the carer is aware of the importance of this sign

and will have preserved a towel or item of clothing stained with the vomitus for inspection. For practical purposes, *bilious vomiting indicates an intestinal obstruction*. Some indication of the likely aetiology and level of the obstruction may be determined from the age of the child, the past medical history and length of illness prior to onset of vomiting. Rapid onset of bilious vomiting and a non-distended abdomen suggest a proximal obstruction, whereas, late onset of bilious vomiting and abdominal distension, a distal obstruction. Immediate referral to a paediatric surgeon is indicated, as urgent surgical intervention may be required.

Causes

The possible causes of bilious vomiting are as shown in Box 7.2.2 and are discussed below.

Intestinal atresia

The term *atresia* implies maldevelopment of a lumen or opening that is normally patent. If there is just a narrowing of the lumen, this is termed a *stenosis*. Atresia of the intestinal tract can occur at any level, but in the context of bilious vomiting the level of obstruction is distal to the ampulla of Vater in the second part of the duodenum.

As the lesion is congenital, the child will present in the neonatal period, although increasingly, some of these infants are diagnosed on antenatal ultrasound scans.[1] Proximal lesions, such as duodenal or jejunal atresia, will usually present in the first 24 hours of life whereas more distal lesions, such as ileal or colonic atresias, can present later. Duodenal atresia may be associated with Down syndrome and cardiac anomalies in up to 30% of cases.[2]

Box 7.2.2 Causes of bilious vomiting

Intestinal atresia
Anorectal anomalies
Meconium ileus
Hirschsprung disease
Malrotation with volvulus
Irreducible inguinal hernia
Intussusception

Inflammatory
Meckel’s diverticulum
Adhesions
Non-surgical

Anorectal anomalies

Typically the anus is imperforate, and the rectum communicates with the urinary tract or perineum via a fistula. It may occur as part of the VACTERL association of anomalies (vertebral, anorectal, cardiac, tracheo-oesophageal, renal and limb).[3] The diagnosis should normally be made as part of the routine assessment of a neonate following delivery but is occasionally missed, leading to a complete or partial distal obstruction and late-onset bilious vomiting. Infrequently, the anomaly may present outside the neonatal period once the child commences solids.

Meconium ileus

This condition occurs in about 10–20% of neonates with cystic fibrosis (CF), although uncommonly it may occur in the absence of CF.[4] There may be a family history of CF. The condition results from the occlusion of the bowel lumen by abnormally viscous enteric secretions in the small bowel and may be complicated by an atresia.

Hirschsprung disease

In this condition the enteric nervous system is abnormal, leading to a distal physiological obstruction. The condition is more common in males by a ratio of 4:1.[5] In 75% of cases, so-called ‘classical’ Hirschsprung disease (HD), a variable extent of the rectum and sigmoid colon is involved, leading to a distal colonic obstruction.[6] In the remaining 25% of cases, bowel proximal to the sigmoid colon will also be aganglionic. About 10% of children with HD will also have Down syndrome.[7] The cardinal feature of HD is the failure to first pass meconium within 24 hours of birth in a term infant, with the later development of bilious vomiting in the first few days of life.

Malrotation with volvulus

The mid-gut normally develops within a physiological hernia in utero. As this reduces towards the end of the first trimester, the mid-gut undergoes an anticlockwise rotation around the axis of the superior mesenteric vessels.[8] Failure of this process to occur results in malrotation. The onset of bilious vomiting usually indicates that the malrotated small bowel has obstructed as a result of twisting around its pedicle, although occasionally, this may occur as a result of associated congenital bands. There is a high risk of intestinal ischaemia, so urgent surgical intervention is required once the diagnosis has been confirmed.[9] Although malrotation most commonly presents in the first month of life, it can present at any time and even in adult life.[10]

Irreducible inguinal hernia

An irreducible inguinal hernia, particularly in a male, often contains bowel. The resultant obstruction of the bowel at the level of the external inguinal ring may produce bilious vomiting. Untreated, the irreducible hernia is at risk of producing intestinal ischaemia.

Intussusception (Chapter 7.10)

In intussusception the proximal bowel (the ‘intussusceptum’) invaginates or telescopes into the distal, receiving bowel (the ‘intussuscipiens’). Initially this involves the ileum invaginating into itself but then progresses to involve the colon. Most commonly, it presents in infants or toddlers with episodic colicky abdominal pain, which may be associated with vomiting or pallor. As the disease progresses, vomiting may become bilious, resulting from intestinal obstruction. The ‘classical’ re-dcurrant jelly stool of intussusception is an uncommon, typically late phenomenon, but occult blood can often be detected earlier.[11]

In older children above 3 years of age, intussusception is usually associated with a pathological lead point such as a Meckel’s diverticulum (MD) or small bowel polyp.[12] It may also complicate CF and Henoch–Schönlein purpura.

Inflammatory

Complicated acute appendicitis, Meckel’s diverticulitis or inflammatory bowel disease leading to the development of an inflammatory mass may result in

intestinal obstruction. This presentation usually occurs when there has been a diagnostic delay of the primary pathology, often in children under 5 years of age.

Meckel's diverticulum

This represents a remnant of the omphalomesenteric duct and is present in about 2% of the population.[13] For reasons that are unclear, complications of a MD are more common in males.[13] MD may lead to a bowel obstruction either as a result of acting as the lead point for an intussusception, due to an associated band adhesion with volvulus or an internal hernia, or rarely an inflammatory mass due to Meckel's diverticulitis.

Adhesions

Bowel obstruction due to adhesions is uncommon in children following abdominal surgery, at least in childhood.[9] This is fortunate as, unlike adults, the obstruction rarely settles with non-operative intervention.

Non-surgical

A wide variety of medical conditions may occasionally present with bilious vomiting, together with a variable degree of abdominal distension. Severe gastroenteritis with prolonged vomiting, sepsis, pyloric stenosis, hypothyroidism, meconium plug syndrome and the cyclical vomiting syndrome may all lead to a clinical picture similar to intestinal obstruction.[14] Any child presenting with bilious vomiting, however, requires paediatric surgical consultation to exclude a surgical cause.

Complications

The major complication of bilious vomiting due to intestinal obstruction is the potential for intestinal ischaemia if the diagnosis is delayed. Even short periods of ischaemia may lead to loss of the normal gut barrier function and a prolonged paralytic ileus. If there has been diagnostic delay with irreversible bowel ischaemia, perforation with peritoneal contamination can occur. In cases requiring extensive resection, short-bowel syndrome can result. In unrecognised cases, overwhelming septicaemia and even death can result.[9]

Investigations

A variety of investigations may be required in the child with bilious vomiting but should not replace a focused history, clinical examination and surgical assessment.

Haematological and biochemical investigations are often normal unless the child is moderately to severely dehydrated. Acid–base derangement may occur secondary to the volume and content of the vomitus. If there is intestinal ischaemia, there is usually a marked leukocytosis and lactic acidosis. The blood glucose level should always be checked and the urine screened for sepsis.

The initial radiological investigations should be directed towards demonstrating obstruction and include plain films of the abdomen and chest. Free gas from a perforation can be seen on an erect chest, erect abdomen or decubitus abdominal views. Fluid levels consistent with intestinal obstruction may be seen on an erect or decubitus abdominal film but can be normal and may occasionally occur in medical causes of bilious vomiting. Dilated small bowel is identified by the presence of the valvulae conniventes of Kirkring passing across the entire lumen, as opposed to the incomplete haustral markings in the colon. In neonates, these markings are usually not visible. There may be features to suggest intussusception (see Chapter 7.10 on Abdominal pain).

The use of further investigations will be directed by the likely pathology and surgical consultation. Contrast studies, either upper or lower, may be both diagnostic of the surgical cause and, in the case of intussusception, potentially therapeutic, although now generally replaced by the safer air enema.[8,11] Ultrasound (US) has a limited role in the investigation of bilious vomiting but may be useful as a screening tool in children with suspected intussusception and the detection of pathological lead points.[15] Whilst US may be diagnostic of malrotation, it cannot be relied upon to exclude the diagnosis, with an upper gastrointestinal contrast study the optimal investigation for this pathology.[16]

Treatment

The treatment of intestinal obstruction in children involves analgesia, fluid and electrolyte resuscitation and definitive surgical management.

The child should be fasted. Neonates and infants require a warm environment to ensure temperature stability. Often large volumes of fluid have been lost from the intravascular space, and 10–20 mL kg^{-1} fluid boluses of warmed crystalloid

solution may be required to treat shock. In addition, calculated maintenance and deficit fluid using normal saline with dextrose should be given. A gastric tube, at least 10 or 12F in size should be placed and regularly aspirated to decompress the stomach. Ongoing fluid losses should be charted and replaced on a mL for mL basis with intravenous normal saline. Hypokalaemia, if present, needs to be treated appropriately (see Chapter 10.6 on fluids and electrolytes). If required, intravenous analgesia should be given in the form of morphine 0.1 mg kg^{-1} per dose and titrated to the response of the patient.

Most surgical causes of bilious vomiting will require operative treatment following further investigation.[9] Although adhesive obstruction may initially be treated non-operatively by a 24- to 48-hour period of gut rest, only rarely is this effective in children.

Controversies

1 Radiological diagnosis of malrotation. False positives and negatives may occur with both contrast studies and US assessment.[17] Ultimately the clinician needs to make an assessment based on the patient's signs and symptoms in conjunction with information obtained from radiological studies.

2 The role of laparoscopy in the assessment of small bowel obstruction in children.[18] There may be a role for laparoscopy as the first manoeuvre in the operative management of children with adhesive intestinal obstruction.

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7.3

Gastrointestinal bleeding

Vered Schildkraut

ESSENTIALS

1 The causes of gastrointestinal (GI) bleeding in infants and children fall into age-specific diagnostic categories.

2 The majority of GI bleeding in children ceases spontaneously, does not require emergent treatment and can be managed on the basis of the presumptive diagnosis.

3 A good history is important in determining the likely site of GI bleeding, the cause, the significance and acuity of the bleeding and will guide appropriate investigations.

4 Initial assessment is for signs of haemodynamic compromise, recognising the severity of the bleed, the patient's risk factors, determining emergency management and timing of endoscopy.

Introduction

Gastrointestinal (GI) bleeding in infants and children is a concerning and relatively uncommon presentation to the emergency department (ED),[1,2] with higher rates in those admitted to ICU.[3] A proper understanding of the age-related aetiologies and available treatment modalities is essential. Although many causes of GI bleeding are common to children and adults, the frequency of specific causes differs greatly, and some lesions, such as necrotising enterocolitis, are unique to children. The causes of GI bleeding in children range from common and benign, as seen in a toddler with an anal fissure, to potentially

life-threatening oesophageal varices requiring urgent therapy. Assessment aims to differentiate between the causes of GI bleeding that require urgent treatment and those that resolve with conservative therapy.

Definitions

Haematemesis is the passage of bloody or dark brown material from the mouth, usually associated with GI bleeding proximal to the ligament of Treitz. '**Coffee-ground'** vomiting suggests that the bleeding has stopped some time ago.

Melaena is the passage of black, tar-coloured stool. This is usually associated with proximal bleeding (e.g. upper GI, small bowel, proximal colon) and caused by bacterial degradation of haemoglobin.

Haematochezia is the passage of bright red blood or maroon-coloured material per rectum. It usually indicates a source of blood from the lower GI tract, distal small bowel or colon. It can, however, be derived from an upper GI source, especially in infants, due to rapid colonic transit times.

Occult GI bleeding is sub-acute bleeding that is not clinically visible.

Obscure GI bleeding is bleeding from a site that is not apparent after routine endoscopic evaluation.

Aetiology

The aetiology of GI bleeding is best considered within defined age groups, with some overlap between groups, as guided by history and examination (Tables 7.3.1 and 7.3.2). The aetiology is also different between hospitalised and ambulatory patients. In hospital, gastritis and stress ulcers are more common, as well as drug-induced injury due to non-steroidal anti-inflammatory drugs (NSAIDs). In a recent study of 103 children admitted to hospital with upper GI bleeding, 34% had erosive gastritis, 15% oesophagitis, 12% duodenitis and 11% a duodenal ulcer.[4]

Table 7.3.1

Causes of upper gastrointestinal bleeding (causes listed most common to rare)

Neonates (<1 month)	Infants (1 month to 1 year)	Toddlers and school age	Adolescent
Swallowed maternal blood	Oesophagitis	Oesophagitis	Oesophagitis
Gastritis and gastric erosions	Gastritis and gastric erosions	Gastritis	Mallory–Weiss tear
Vascular malformations	Mallory–Weiss tear	Mallory–Weiss tear	Gastritis
Bleeding disorders Haemophilia Vitamin K deficiency	Foreign body ingestion	Drug-induced ulcers, NSAID use	Peptic ulcer disease
Duplication cyst	Peptic ulcer disease	Foreign body ingestion	Oesophageal varices and portal hypertensive gastropathy
Maternal NSAIDs	Vascular malformation	Peptic ulcer disease	Drug-induced ulcers and NSAID use
Trauma (NG tube, nasal suction)	Bleeding disorders	Oesophageal varices and portal hypertensive gastropathy	Foreign body ingestion
	Oesophageal varices and portal hypertensive gastropathy	Bleeding disorders	Bleeding disorders

NSAID, non-steroidal anti-inflammatory drug.

Table 7.3.2

Causes of lower gastrointestinal bleeding (causes listed most common to rare)

Neonates	Infants	Toddlers and school age	Adolescent
Swallowed maternal blood	Anal fissure	Anal fissure	Anal fissure
Necrotising enterocolitis	Protein sensitive enterocolitis	Polyps	Polyps
Protein-sensitive enterocolitis	Hirschsprung enterocolitis	Infectious colitis	Inflammatory bowel disease
Hirschsprung enterocolitis	Infectious colitis	Meckel's diverticulum	Infectious colitis
Ischaemic enterocolitis	Ischaemic enterocolitis	Rectal prolapse	Intussusception
Infectious gastroenteritis	Meckel's diverticulum	Inflammatory bowel disease	Meckel's diverticulum
Congenital bleeding disorders	Intussusception	Intussusception	Haemorrhoids
Malrotation with volvulus	Haemolytic–uraemic syndrome	Henoch–Schönlein purpura	Ischaemic enterocolitis
Duplication cyst	Bleeding disorders	Bleeding disorders	Vascular malformation
Vascular malformation	Vascular malformation	Haemolytic uraemic syndrome	Bleeding disorders
Maternal NSAIDs	Inflammatory bowel disease		Solitary rectal ulcer
	Vascular malformation		Typhilitis/neutropenic colitis

NSAIDs, non-steroidal anti-inflammatory drugs.

The term newborn infant who is breast-fed and has GI bleeding is most likely to have ingested maternal blood either at the time of delivery or from breast-feeding from cracked nipples. Premature infants are at an increased risk of necrotising enterocolitis (NEC). Sick infants, compromised by hypoxia or hypotension, are at risk of GI bleeding from stress ulcers. Haemorrhagic disease of the newborn, associated with vitamin K deficiency, can present with GI bleeding. Cow's milk protein intolerance can cause enteropathy and rectal bleeding within the first few weeks to months of life in formula-fed as well as breast-fed infants. The older child with cerebral palsy may have gastro-oesophageal reflux related upper GI bleeding.[1] History of aspirin and NSAIDs use is an increasingly common risk factor for GI bleeding.[4]

History

Initial questions for all patients presenting to the ED with GI bleeding aim to assess the need for an immediate intervention and should include information about the location, duration, quantity and appearance of the bleeding. It is important for the ED physician to identify the risk factors for subsequent

complications, including rebleeding, the need for surgery, transfusion and death. **High-risk GI bleeding** is associated with existing chronic medical comorbidities, including coagulopathy, portal hypertension, respiratory and cardiac failure, use of medications (aspirin, NSAIDS, anticoagulants, steroids and chemotherapy) and previously diagnosed peptic ulcer.[5] There are two causes that are associated with high mortality rates in children if not treated: oesophageal varices and **aortoenteric fistula**. The latter is very rare but rapidly fatal and in children can be related to oesophageal foreign bodies, particularly button batteries. **Gastro-oesophageal varices** form in children with intrahepatic and extrahepatic causes of portal hypertension and rarely in association with congenital heart disease or vascular malformations. Cirrhosis is the most common cause of paediatric portal hypertension.[1]

Further history should provide clues to the location of the bleeding (e.g. upper or lower GI), whether the amount of bleeding is significant (e.g. the appearance of the blood in the toilet bowel or streaks on the toilet paper) and to assess a possible cause based on associated symptoms.

It is important to consider conditions that may mimic GI bleeding such as ingestion of **substances that may colour** the stool or emesis (food colouring, candy, beets or iron supplements) or that the bleeding is not from the GI tract. **Non-GI sources** that mimic upper GI bleeding include epistaxis, dental bleeding, recent ENT or oral surgery or hemoptysis. Vaginal bleeding, gross haematuria, and partially digested foods may be mistaken for haematochezia.[5]

A positive family history may be helpful, including a history of familial polyps, peptic ulcer disease and *H. pylori* infection, inflammatory bowel disease, coagulation disorders, haemorrhagic telangiectasia and Hirschsprung disease.

Certain diagnoses also have a recognised age pattern, such as juvenile polyps (peak incidence of 1 to 6 years of age), intussusception (peak incidence of 5 to 18 months), and inflammatory bowel disease that more commonly presents in adolescence, although can occur at any age. Inadvertent ingestion of foreign bodies, caustic agents and rodenticides containing warfarin-type agents should be considered in preschool-age children. Perianal streptococcal dermatitis caused by group A beta-hemolytic streptococci infection occurs mainly in children between 6 months and 10 years of age.[1] Table 7.3.3 outlines the historical features of the different causes of GI bleeding.

Examination

On initial evaluation, physical examination should focus on the patient's vital signs and assess for the presence haemodynamic compromise. Examination should be directed to assess circulatory adequacy (vital signs, orthostatic vital signs and urine output) and to find evidence of brisk bleeding in the oral cavity, rectum, on the clothes or in the nappy. This will allow rapid determination of whether the child is well or ill.

Specific physical examination findings may establish potential causes for the GI bleeding. Stigmata of chronic liver disease and signs of portal hypertension suggest oesophageal varices. Skin and mucous membrane examination may reveal evidence of an underlying bleeding diathesis. Lip and buccal hyperpigmentation suggests Peutz–Jeghers syndrome. Perianal inspection may identify an anal fissure associated with chronic constipation, perianal erythema caused by beta-haemolytic *Streptococcal* infection or features of perianal Crohn's disease (Box 7.3.1).[6,7]

Table 7.3.3

Suspected source of gastrointestinal bleeding as suggested by the patient's history

Suspected source of bleeding	Patient history
Maternal blood	Newborn, cracked maternal nipples in breast-fed infants
Oesophageal ulcerations	Reflux, heartburn, odynophagia, drugs, foreign body
Mallory–Weiss tear	Recurrent retching, prior vomiting*
Cameron's lesions	Large hiatal hernia
Oesophageal or gastric varices Portal hypertensive gastropathy	Chronic liver disease, cirrhosis, portal hypertension
Peptic ulcer, gastritis	Epigastric pain, aspirin, NSAIDs use, alcohol exposure, family history of peptic ulcers or *Helicobacter Pylori* infection
Aorto-oesophageal fistula	Foreign body, disc battery ingestion
Bile ducts	Recent liver biopsy
Pancreatic ducts	Pancreatitis, pseudocyst
Meckel's diverticulum	Painless, unexplained GI bleeding, melaena, dark red stools
Small intestinal or colonic ulceration	Use of aspirin, NSAIDs
Cow milk protein enteropathy	Infant irritability, poor feeding, vomiting
Intussusception	Paroxysmal abdominal pain, lethargy, currant jelly stool
Infectious colitis†	History of travel, exposure to sick contacts, acute crampy abdominal pain diarrhoea with mucus and fresh blood
Necrotising enterocolitis (NEC)	Preterm infant, apnoea, poor feeding

Ulcerative colitis	Bloody diarrhoea, chronic abdominal pain, family history of IBD[‡]
Crohn's disease	Bloody diarrhoea, chronic abdominal pain, family history of IBD[‡]
Polyps	Family history of hereditary GI cancer syndromes, fresh blood coating the stool
Anal fissure	Chronic constipation, fresh blood coating the stool, pain when passing hard large bowel motions, straining at stool
Haemorrhoids	Dripping of blood with bowel motions
Perianal streptococcal dermatitis	Perianal rash, blood-streaked stools
Henoch–Schönlein purpura	Typical palpable purpura on extremities, abdominal pain
Hereditary haemorrhagic telangiectasia (Osler-Weber-Randu disease)	Frequent nosebleeds, family history

GI, gastrointestinal; IBD, irritable bowel disease; NSAIDs, non-steroidal anti-inflammatory drugs.

[*] A Mallory–Weiss tear may occur in the absence of prior symptoms.

[†] Bacteria causing infectious colitis: *Campylobacter*, *Shigella*, *Salmonella*, *E coli, Clostridium difficile* and *Yersinia*.

[‡] IBD inflammatory bowel disease.

Investigations

Laboratory testing

Diagnostic tests should be directed by the history and physical examination, as well as the severity of the bleeding. If the history is suggestive of a more benign cause of bleeding, such as an anal fissure or cow's milk protein enterocolitis, limited laboratory studies are required, and a full blood examination (FBE) may suffice. For more significant GI bleeding, FBE, coagulation studies, liver function tests (LFTs), urea, creatinine and electrolytes in addition to blood type and screen are the first-line blood tests (Box 7.3.2).

The hematocrit value immediately after the onset of bleeding may not accurately reflect blood loss. A low MCV may indicate chronic blood loss and iron deficiency. A low platelet count suggests chronic liver disease, and blood urea nitrogen increases in upper GI bleeding as a result of breakdown of blood proteins by the intestinal bacteria.[7]

In a breast-feeding newborn with streaks of fresh blood in the vomitus, an Apt test distinguishes between swallowed maternal blood and infant's blood.[6]

Box 7.3.1 Physical examination

Vital signs: orthostasis, pulse pressure, urine output
General appearance: well or ill, fever, mental status[*]
Head, ears, nose and throat: trauma, scleral injection, petechiae, lip pigmentation[†], mouth ulcers[‡], epistaxis, erythema or burns to posterior pharynx, blood in the nose or oral cavity, lip swelling (orofacial granulomatosis)[‡]
Skin: pallor, jaundice[*], rash, arteriovenous malformation, bruising, petechiae, palmar erythema[*], spider angiomas[*], clubbing, telangiectasia[§‡]
Chest/cardiovascular: tachycardia, murmur, capillary refill
Abdomen: tenderness, mass, distension, splenomegaly[*], hepatomegaly[*], caput medusae[*], ascites[*]
Perianal, rectal: gross blood, melaena, tags[‡], tenderness, fissure, fistula[‡], swelling[‡], mass[‡]

[*] chronic liver disease
[‡] Crohn's disease
[†] Peutz–Jeghers syndrome
[§] hereditary haemorrhagic telangiectasia (HHT, Osler-Weber-Randu disease).

Stool testing

Stool samples for microscopy, looking for red and white blood cells, and culture should be sent when bacterial enterocolitis is suspected.[6]

Imaging

Imaging studies will depend on the apparent or suspected source of bleeding. Chest X-ray is warranted when a pulmonary source or a foreign body is suspected. An abdominal X-ray may be indicated for the evaluation of possible obstruction, bowel perforation or bowel wall pneumatosis in NEC.

Plain films of the neck, chest and abdomen should be performed in preschool children with unexplained bleeding to look for an ingested foreign body, particularly button batteries.

Ultrasound is useful to identify portal hypertension or an intussusception.[6] A Technetium pertechnetate scintigraphy (a 'Meckel scan') can identify ectopic gastric mucosa in a Meckel's diverticulum with overall accuracy of about 90% in

children and is indicated in children with unexplained painless GI bleeding.[8]

Angiography and radionuclide imaging (Technetium-99m-labelled red blood cells or 'red-cell scan') are rarely indicated or used in children with GI bleeding. Although these may be used to diagnose and treat severe bleeding when the cause cannot be determined endoscopically, they are limited by the rate of bleeding (>1 mL min in angiography and >0.1 mL min in red cell scan).[6]

Capsule endoscopy

Capsule endoscopy (CE) has been used to identify bleeding sites in the small intestine, which is otherwise inaccessible by upper and lower endoscopy. The capsule is either swallowed or placed endoscopically. Its disadvantages are that it is too large for use in most children aged less than 2 years and it cannot obtain biopsy specimens or perform therapeutic interventions. There is emerging adult data about CE use for acute GI bleeding; however, this has not been studied in children and cannot substitute for upper endoscopic evaluation.[9]

Box 7.3.2 Laboratory studies

Full blood count (chronic anaemia, thrombocytopenia)
PT/INR (vitamin K deficiency, liver disease)
PTT (factor deficiency)
Liver function tests (chronic liver disease)
Urea, creatinine, electrolytes (increased urea from amino acid catabolism secondary to blood digestion)
Blood type, screen, cross
Stool culture (if indicated by history)
ESR, CRP (if indicated by history)
Hemoccult blood test (presence of blood)
APT test (swallowed maternal blood)
Serial FBE, electrolytes and fibrinogen (rate of bleeding)

APT, alkali denaturation test; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; FBE, full blood examination; INR, international normalised ratio; PT, prothrombin time; PTT, partial thromboplastin time.

Initial medical therapy

Fluid resuscitation and blood transfusion

Cool peripheries, delayed capillary refill and tachycardia will be the first detectable signs of haemodynamic compromise in significant GI bleeding. Hypotension or postural hypotension occurs late.

At least one large-bore intravenous catheter should be placed in the patient with significant bleeding. Two intravenous (IV) catheters should be placed in the ill patient or the patient who has ongoing bleeding after initial fluid resuscitation. Give 20 mL kg fluid boluses with crystalloid solution (0.9% sodium chloride). Children with significant GI bleeding require ongoing monitoring of heart rate, blood pressure, and urine output in the ED. These observations combined with frequent clinical assessment will help guide ongoing fluid therapy.

Packed cell transfusion (Chapter 11.1) is indicated for moderate to severe bleeding with haemodynamic instability unresponsive to initial fluid boluses. The haemoglobin level that indicates transfusion differs according to the clinical evaluation and the cause of bleeding. In patients over the age of 18 years with upper GI bleeding, a restrictive transfusion strategy (transfusion when haemoglobin level was <7 g dL rather than <9 g dL) was associated with higher survival rate.[10] Correction of identified coagulopathy with fresh frozen plasma, activated factor VII or platelet infusions may be needed to control the GI bleeding in patients with coagulopathy.

Nasogastric aspiration and lavage

A nasogastric (NG) lavage and aspiration have previously been considered useful for significant upper GI bleeding. Reasons included providing evidence of ongoing bleeding, estimating blood loss, clearing gastric blood for better endoscopic visualisation and minimising the risk of aspiration. However, they have poor sensitivity and are generally *not indicated* for evaluation of GI bleeding.[11,12]

Emergency endoscopy

Upper GI endoscopy in children with melaena or haematemesis is the preferred diagnostic procedure to find the source of bleeding and perform endoscopic haemostasis. The diagnostic yield of endoscopy is best if performed as soon as the patient becomes stable and preferably within 24 hours of the onset of bleeding. Upper endoscopy allows direct visualisation of the mucosal lining,

identification of bleeding lesions and provides important information regarding the risk of recurrent ulcer bleeding. In infants with gastrointestinal bleeding, the use of endoscopy is limited by the ability to apply interventional modalities through a smaller endoscope. Endoscopic haemostasis techniques include epinephrine injection, thermal probes, endoscopic haemostatic clips and Argon Plasma Coagulation (APC laser).[1,6] The source of upper GI bleeding may not be identified in upper endoscopy in approximately 25% of patients, while procedures performed more than 48 hours after the bleeding event have a lower diagnostic yield.[4]

Most cases of acute lower GI bleeding in children will stop spontaneously, allowing non-urgent management; an urgent colonoscopy is very rarely indicated. During a colonoscopy, identified polyps can be removed endoscopically and retrieved for histologic examination.

Surgery

In selected patients with severe, ongoing GI bleeding, in whom the diagnosis is not made by endoscopy and the bleeding either cannot be controlled by endoscopic methods or is severe, recurrent and obscure, a laparoscopy or laparotomy may be indicated. A study of 17 children with GI bleeding and no identifiable source after upper endoscopy and colonoscopy found that eight patients had Meckel's diverticulum at laparoscopy, five patients had other GI pathology that accounted for symptoms (intestinal duplication, lymphoid hyperplasia and vascular enteritis) and four patients had normal findings.[13]

Treatment

Acute upper gastrointestinal bleeding

After the initial assessment and stabilisation, consideration of the probable cause of bleeding, risk factors and gastroenterology consultation, the emergency physician is likely to have decided whether pharmacologic treatment is indicated and/or emergency endoscopy is imminent.

For all patients with a significant upper GI bleeding:

- keep the patient nil by mouth, as an endoscopic procedure may be required
- who have required resuscitation, an intensive care or high dependency

unit admission may be needed
- start acid suppression therapy.

Pharmacological treatment (Table 7.3.4)

Both proton pump inhibitors (PPIs) and H_2-receptor antagonists may be used to control acid secretion. PPIs are more effective and are used for significant bleeding.[6] PPIs have been shown to reduce the risk of re-bleeding and need for surgery for peptic ulcers, and intravenous treatment should be commenced in acute upper GI bleeding.[14] The risk of subsequent bleeding is greatest in the first 3 days after the initial bleeding episode, and IV PPI is usually continued during this time. In less significant bleeding, an initial, twice daily IV dose can be converted to an oral PPI after 24–48 h. Children with gastroduodenal ulceration associated with *Helicobacter pylori* infection are treated with antibiotic eradication therapy.[1,6]

Variceal bleeding

In children with variceal bleeding, administer IV vitamin K (2–10 mg) and broad-spectrum antibiotics (a third-generation cephalosporin), as variceal bleeding is associated with bacterial infection. In a stable patient, upper GI endoscopy should be arranged as soon as possible. Intravenous somatostatin or octreotide has been shown to be effective for uncontrolled bleeding from oesophageal varices by decreasing splanchnic blood flow[1] (see Table 7.3.4).

Table 7.3.4

Medications used for gastrointestinal bleeding

Agent	Drug category	Oral administration	IV administration
Ranitidine	Histamine-2 antagonists	2–4 mg kg dose 2–3 times a day (maximum, 300 mg)	Bolus: 1 mg kg dose (max 50 mg dose) every 6–8 hours Continuous infusion (after bolus): 2–4 mg kg d
Pantoprazole	Proton pump inhibitor	1–1.5 mg kg d 1–2 times daily (maximum, 40 mg d)	Intermittent bolus dose: 0.5–1 mg kg day in two divided doses Continuous infusion (including initial bolus): 1 mg kg followed by infusion dose of 0.1 mg kg h (to maximal adult dose) Adults: Bolus 80 mg followed by 8 mg h
Esomeprazole	Proton pump inhibitor	Infants: 3.5–5 kg: 2.5 mg d 5–7.5 kg: 5 mg d Children 1–11 years old: <20 kg: 10 mg d >20 kg: 20 mg d Children >12 years old: 20–40 mg d	Intermittent bolus dose: Infants: 0.5 mg kg d Children 1–17 years old: <55 kg: 10 mg Children >55 kg: 20 mg Continuous IV infusion: 1 mg kg (up to 80 mg) followed by infusion dose of 0.1 mg kg h (up to 8 mg h) Adults: Bolus 80 mg followed by 8 mg h
Omeprazole	Proton pump inhibitor	1–1.5 mg kg d 1–2 times daily (maximum, 40 mg d)	
Lansoprazole	Proton pump inhibitor	1–1.5 mg kg d 1–2 times daily (maximum, 60 mg d) Adult dose: 15–30 mg once daily (maximum 180 mg day in two divided doses)	
Octreotide	IV vasoactive agent Somatostatin analogue		1 mcg kg bolus (maximum, 50 mcg) followed by: IV infusion: 1 mcg kg h may increase to 4 mcg kg (maximum, 250 mcg dose every 8 hours) When bleeding controlled, taper dosing: by 50% every 12 hours for 1–2 days
Vasopressin	IV vasoactive agent Antidiuretic hormone		0.002–0.005 units kg min for 12 hours, then taper for 1–2 days (maximum, 0.2 units min)

∗Doses of these medications are often not well studied in children. Dose adapted from adult dosing.

Table adapted from Neidich GA, Cole SR. Gastrointestinal bleeding. *Pediatrics in Review* 2014;**35**:243–54.

Endoscopic management of varices includes endoscopic sclerotherapy and endoscopic band ligation. A **Sengstaken-Blakemore** tube is used for bedside balloon tamponade and should be considered only in exsanguinating patients with variceal bleeding when endoscopy is not immediately available.[5]

Lower GI bleeding is common, but usually in children it is of modest severity. Most of the patients can be managed on an outpatient basis, and the bleeding will resolve spontaneously. Bacterial enterocolitis is usually self-limiting and does not require empiric antibiotic treatment. Children with bleeding from an anal fissure may require laxatives to ensure regular soft stools.

When cow's milk protein intolerance is suspected, consider maternal elimination diet in breast-fed infants or extensively hydrolysed and amino acid formula in formula-fed infants, with expected improvement of bleeding within days to weeks.[15]

Dispositon

The initial goals when managing a patient with GI bleeding are to establish the extent, the severity and source of bleeding. The decision to admit or discharge the infant or child with GI bleeding from the ED is influenced by the clinical assessment, risk stratification and presumptive diagnosis. Given that the majority of GI bleeding events in children are self-limiting or respond readily to treatment, most can be managed on an outpatient basis. Sensible diagnostic testing, utilising a variety of modalities, can be used to establish the diagnosis. GI endoscopy and newer tests such as capsule endoscopy are often diagnostic and direct management. Mortality in children with GI bleeding is low, and fatalities are reported rarely, mainly in critically ill patients. Emergency physicians should be familiar with the common, uncommon, and comorbid conditions that cause GI bleeding in children and the available treatments.

Controversies

1 Treatment guidelines including pharmacologic treatment and endoscopy are based on adult studies with poor availability of data in children.

2 The lack of evidence of benefit from the use of H_2 antagonists and PPIs for acute gastrointestinal bleeding in children.
3 Limitation of use of endoscopic modalities in children because of size restrictions.
4 The emergency management of upper GI bleeding often involves paediatric endoscopy, which is not readily available in every centre.

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7.4

Gastro-oesophageal reflux

Daryl Efron

ESSENTIALS

1 The infant presenting with regurgitation/vomiting needs to have alternative causes considered, before concluding that the diagnosis is gastro-oesophageal reflux (GOR).

2 GOR is a normal physiological phenomenon to which infants are particularly prone.

3 Infants with benign GOR appear healthy and thriving. If the child appears unwell, another cause should be considered.

4 Infants and children with uncomplicated GOR do not need any investigations.

5 Infants with uncomplicated GOR do not need any intervention, e.g. changes to formula or medications.

6 Complications of GOR include failure to thrive, reflux oesophagitis, feeding refusal, and recurrent aspiration pneumonia.

7 Acid suppression can be helpful in reflux oesophagitis.

Introduction

Gastro-oesophageal reflux (GOR) is the regurgitation of gastric contents into the oesophagus. It is a normal physiological phenomenon at all ages and occurs up to 30 times per day in healthy infants, with a peak at around 4 months. The refluxed gastric contents are generally rapidly cleared from the oesophagus without causing any significant clinical symptoms. In most cases GOR is a

benign condition which usually resolves by 12–15 months. Complications warranting medical intervention are uncommon. When complications do occur, it is referred to as GOR disease (GORD).

Pathophysiology

During periods of raised intraabdominal pressure such as straining, there are a number of protective mechanisms which help to prevent GOR. These include contraction of the diaphragmatic crura around the lower oesophagus in the region of the lower oesophageal sphincter (LES), as well as compression of the intraabdominal segment of the oesophagus. Many episodes of GOR are associated with transient relaxations of the LES.

In infants the LES is located above the diaphragm, rendering the above protective mechanisms ineffective. Therefore, infants are especially prone to GOR, particularly in the early months of life. However, with anatomical and physiological maturation, the natural history is of symptom resolution by late infancy in the vast majority. GOR is unusual in children older than 18 months.[1]

Some children are particularly predisposed to develop GORD. These include premature infants, children with neurological conditions such as cerebral palsy or neurodegenerative disorders, and children with anatomical anomalies such as tracheo-oesophageal fistula. These children have poor oesophageal motility with reduced clearance of refluxed gastric contents.

History

The cardinal symptom of GOR is visible regurgitation of milk post-feeds. This is often called vomiting, although in GOR the expulsion of gastric contents is generally by an effortless spill, whereas true vomiting involves forceful contraction of abdominal wall musculature.

The history is crucial in considering potential differential diagnoses and in identifying complications. Careful questioning regarding the relationship of vomiting to feeds, the content of regurgitated material (e.g. is there blood or bile?), associated distress, and feeding behaviour is essential. Projectile vomiting, in which milk propels some distance, raises the possibility of pyloric stenosis. Episodic irritability related to feeds may indicate GOR, although the association between irritability and GOR in infants is weak. The presence of eczema, rectal blood or mucus (allergic proctocolitis) and a family history raise

the possibility of cow's milk protein allergy. A history of associated fever suggests an infective cause such as urinary tract infection (UTI). In a young infant with known GOR, a change in the character of the vomiting may indicate another diagnosis on a background of GOR, such as pyloric stenosis or a UTI. Seizures raise the possibility of a neurological disorder. Inquiry about associated symptoms such as feeding aversion, poor weight gain and respiratory symptoms such as chronic cough or wheezing is important to screen for potential complications. Onset of vomiting after 6 months of age is unusual in GOR and suggests an alternative cause.

Examination

Infants with benign GOR appear well. If the child appears unwell or lethargic, another cause for the symptoms needs to be carefully considered. The presence of abdominal distension, abnormal posture or tone, or hepatosplenomegaly suggests alternative diagnoses including anatomical anomalies, neurodevelopmental dysfunction or metabolic disorders. A pyloric tumour should be specifically palpated for in young infants. Serial weight measurements should be plotted on the child's growth chart. Respiratory examination is important to identify signs of bronchospasm or chronic lung disease, which may complicate GOR. Pallor may indicate anaemia, which can occur secondary to reflux oesophagitis.

Differential diagnosis

There are many causes of vomiting in children, and it is important to consider alternative causes that may mimic GOR (see Chapter 7.2 on vomiting). These include cow's milk protein intolerance; infection, e.g. urinary tract infection, gastroenteritis; surgical conditions, e.g. pyloric stenosis, malrotation and volvulus; intussusception; metabolic disorders, e.g. inborn errors, diabetes; food allergy; and raised intracranial pressure, e.g. hydrocephalus, posterior fossa tumour, subdural haematoma.

Complications

GOR may result in complications and is then referred to as GORD.

Failure to thrive

Some infants with GOR have poor weight gain, which may extend to failure to thrive (crossing two major centiles on a growth chart). This may result from reduced intake – either related to dysphagia or parental reduction of feed volumes offered in an attempt to minimise regurgitation – or from inadequate absorption of ingested food due to excessive losses.

Respiratory

A number of respiratory complications may occur with GOR. Recurrent aspiration (often silent) results in chronic wheeze or cough, and alveolar disease may develop, with signs of tachypnoea and increased work of breathing. Exposure of the oesophageal mucosa to acid can trigger reflex bronchospasm, and persistent asthma symptoms may occasionally be related to GOR.[2] Intermittent (or even isolated) episodes of aspiration can cause pneumonia. Reflux of gastric contents into the upper airway can result in laryngospasm, presenting as an obstructive apnoea or 'brief resolved unexplained event' (BRUE).[3] However, it is unusual to be able to demonstrate an association between GOR and BRUE.

Oesophagitis

Reflux oesophagitis has become a popular clinical diagnosis given to infants presenting with excessive crying behaviour (so-called 'silent reflux'). There are many causes for infant distress (including temperamental factors, food allergy), and peptic oesophagitis is only responsible for a minority of cases.[4] These infants may have episodes of blood in the vomitus or develop iron deficiency anaemia due to blood loss. Rarely, an infant may develop feed aversion due to the distress of GOR.

Infants with Sandifer syndrome exhibit back arching, neck torsion and chin lifting in response to oesophageal pain due to refluxed gastric acid. These uncommon episodes may be confused with seizures.

Investigations

In the majority of cases, a careful history and examination will clarify the likely diagnosis of GOR. Well and thriving infants and children with uncomplicated

GOR should not be subjected to any investigations. Investigations may sometimes be required to exclude differential diagnoses.

Serum biochemistry, including an acid–base assessment to exclude the evolving hypokalaemic, hypochloraemic metabolic alkalosis of pyloric stenosis, and urine microscopy and culture are relevant first-line tests in the vomiting infant. If the history or biochemistry is suggestive, a pyloric ultrasound should be performed to rule out pyloric stenosis. Some children may require admission for observation of feeding and the regurgitation pattern and consideration of further tests to clarify the diagnosis.

An upper GI contrast study is used to exclude anatomical problems such as malrotation or antral web. The observation of GOR merely demonstrates that the infant experienced an episode of reflux at the time of the study. The frequency of GOR episodes, their correlation with clinical symptoms, or the presence of complications cannot be determined from a contrast study.

Oesophageal pH monitoring with multi-channel intraluminal impedance can be used to quantify GOR but has limited clinical utility. Infants often have abnormal oesophageal pH findings in the absence of histological features of oesophagitis on biopsy, and, conversely, infants with oesophagitis may have no evidence of significant GOR on pH study.[4] These infants presumably have an alternative cause for their oesophagitis, such as allergy. Oesophageal pH and impedance studies may be helpful to confirm the pathophysiology prior to embarking on antireflux surgery, and there may be occasional other clinical scenarios where it provides useful information, e.g. in conjunction with monitoring of respiratory rate, heart rate and oxygen saturations in the investigation of apnoea or episodic hypoxaemia/bradycardia.

Oesophagoscopy and biopsy may be indicated to evaluate for oesophagitis, (peptic or eosinophilic) if the infant is refusing feeds, underweight or displaying severely distressed behaviour.

Treatment

If there is any doubt about the diagnosis, then a paediatric review should be arranged.

Simple measures

It is important to ensure that bottle-fed infants with GOR are not being over-fed.

As a guide, a newborn baby needs about 150 mL kg day, and this requirement decreases to about 120 mL kg day at 3 months and 100 mL kg day from 6 months. Conservative measures such as posturing (upright posturing after feeds, elevation of cot head ~30 degrees), and thickening of feeds are commonly used to reduce the symptoms of GOR;[5] however, these have not been demonstrated to modify clinical outcomes.[6] Breast-feeding should not be stopped to thicken feeds. Prone sleeping position is best for reducing GOR[7] but is not recommended as it is associated with sudden infant death syndrome.

A trial of cow's milk and soy exclusion from the maternal diet in breast-fed infants, or extensively hydrolysed formula, may be indicated if GOR is associated with failure to thrive, rectal bleeding, eczema, or a family history of atopy. Any such trial should be commenced in consultation with a paediatrician and continued for at least 2 weeks, with careful evaluation of symptomatic response. If there is no response after 2 weeks then a trial of an amino-acid based formula is indicated. If a change in feed is successful then this is normally continued until at least 12 months of age. If weight gain is suboptimal in bottle-fed infants, more concentrated feeds may be given. Thickening feeds with cereals also increase caloric intake.

Pharmacological

Pharmacological treatment of infants with uncomplicated GOR is not indicated.[8]

H_2-receptor antagonists [9] and proton pump inhibitors (PPIs)[10,11] have been shown to improve both histological changes and symptoms in reflux oesophagitis. However, acid suppression with PPIs is not effective in reducing regurgitation or irritability in infants with GOR.[12] Furthermore there are safety concerns with chronic use of PPIs, including an increased risk of pneumonia, diarrhoea and micronutrient deficiencies.

High-dose antacids can be effective in treating oesophagitis;[13] however, prolonged use can be associated with aluminium toxicity and is not recommended. Prokinetic agents do not have clear evidence for symptom reduction;[14] however, a trial of domperidone or erythromycin may occasionally be indicated in severe GORD under gastroenterology supervision.

Surgical

Fundoplication surgery has a limited role in selected cases of severe GORD,

such as neurologically abnormal infants with recurrent aspiration pneumonia. Complications include breakdown of the wrap, dysphagia, bloating and gagging. Some surgeons are now performing fundoplications laparoscopically.

Follow-up

Children diagnosed with GOR need follow-up organised with their GP or paediatrician to ensure no alternative diagnosis is missed and to monitor for complications. Any changes to maternal diet or infant formula need to be continued for at least 2 weeks in order to determine effectiveness. Parents need an explanation of the symptoms and to be reassured that the natural history is for spontaneous resolution in infancy.

Controversies

1 Infant distress has a range of contributing factors, and peptic oesophagitis is uncommonly the cause.
2 Acid suppression medications are not indicated for uncomplicated GOR and are probably being overprescribed in infancy.
3 Prokinetic agents have an extremely limited role in cases of severe GORD.

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7.5

Pyloric stenosis

Kim Lian Ong, and Ramesh Nataraja

ESSENTIALS

1 Hypertrophic pyloric stenosis (HPS) commonly presents at 2–8 weeks of age and is rare outside of that age range.

2 Projectile vomiting is pathognomonic, occurring approximately half an hour after a feed. The infant remains hungry after each bout of vomiting. Characteristic metabolic derangements with a hypochloraemic, hypokalaemic metabolic alkalosis occur as a result of prolonged vomiting.

3 Other symptoms include weight loss, failure to thrive, clinical signs of dehydration (e.g. increased capillary refill time [CRT], decreased skin turgor, sunken fontanelle, decreased urine output), and also possible jaundice.

4 The diagnosis can be made by the characteristic clinical manifestations and the finding of a pathognomonic palpable olive-shaped mass in the right upper quadrant.

5 Ultrasonography is the gold standard for diagnostic investigation.

6 Treatment consists of two stages: correction of metabolic derangements via intravenous fluid resuscitation and definitive surgical intervention with a Ramstedt pyloromyotomy.

Introduction

Hypertrophic pyloric stenosis (HPS) is a common cause of infantile gastric

outlet obstruction and is one of the most common emergency surgical conditions of infancy.[1] It is caused by idiopathic diffuse hypertrophy and hyperplasia of the circular muscle fibres of the pylorus with proximal extension into the gastric antrum. In response to this outflow obstruction and vigorous peristalsis, the stomach musculature becomes uniformly hypertrophied and dilated.

Epidemiology

Pyloric stenosis has an incidence of 2 to 4 per 1000 live births in Western populations[2] and is less common in infants of African and Asian backgrounds. Males are affected four to five times more than females.[3] The exact cause of pyloric stenosis is unknown, although there are a number of candidate genes involving smooth muscle function and regulation including the nitric oxide synthase gene NOS1. These are located on chromosome X accounting for the increased male incidence.[4] There is also evidence for a strong environmental trigger due to the typical age range of presentation. Offspring of parents with this condition have a higher risk of developing HPS, and in many series first-born males are more frequently affected than the other siblings.[5]

Clinical presentation

The hypertrophy and hyperplasia of the pyloric muscle develops gradually over time. Therefore the symptoms progress and become more severe with increasing gastric outflow obstruction. In the early phase, there may just be regurgitation or the occasional non-projectile vomiting. It commonly presents at 2–8 weeks of age; however, it is reported in premature babies prior to this corrected age. With progression of the gastric outflow obstruction to near complete, the vomiting becomes more projectile. Increased levels of obstruction shorten the time before the feed is vomited.

Projectile vomiting is defined as the sudden involuntary forcible ejection of gastric contents so that it is projected a distance from the infant. This is compared to posseting, which is the normal effortless small quantity vomiting of infancy when the vomitus comes down the infant’s chin.

Shortly after vomiting, the infant is often hungry and will want to take another feed immediately. The vomit is non-bilious, but due to the frequency and force of vomiting, it may contain altered blood from oesophageal Mallory–Weiss tears. The amount of stool passed may be variable. There is always a degree of

clinical dehydration, but once there is an established pyloric tumour this may progress quickly to include severe metabolic alkalosis.

It is important to note that it may take some time for the clinical features, metabolic disturbance and ultrasound findings to become established. A diagnosis of HPS should therefore not be excluded if there are variable symptoms and a relatively normal examination in an infant presenting early. Clinical features may evolve, and a planned review or a period of observation in hospital is appropriate.

Infants with a complete HPS will anatomically have severe gastro-oesophageal reflux (GORD). However, HPS may also develop in infants already being treated for GORD. Therefore, a detailed history from the parents is vital to detect a change from a benign ‘posseting’ or ‘refluxy’ baby, whose regurgitation of milk has ‘changed’ in intensity or frequency and become more projectile.

Examination findings

With an established HPS, the infant fails to thrive due to calorie losses, becomes dehydrated due to fluid losses and appears ‘hungry’ unless significant dehydration or alkalosis has started to affect the infant’s activity level. The infant usually appears ‘bright and active’ unless significantly dehydrated, compared to the infant with a urinary tract infection who may be systemically unwell. With severe dehydration – which may occur when the infant presents late to a medical facility – there will be typical features such as dry mucous membranes, decreased skin turgor, sunken fontanelle and increased CRT.

On physical examination, gastric distension or visible peristaltic waves may be seen moving from the left upper abdomen toward the epigastrium and then to the right.[6] The palpation of a firm, mobile and non-tender ovoid mass (‘olive’) is diagnostic. This is located either to the right of the epigastrium or in the midline, deep to right rectus muscle and under the liver edge. This clinical finding requires patience, as success is dependent on an empty stomach and a relaxed anterior abdominal wall in a non-crying settled infant. If the stomach is significantly distended during palpation, aspiration of gastric contents using a nasogastric tube may be helpful to increase the likelihood of feeling the ‘olive’. A test feed, consisting of warm milk or water, can also be used and may allow a previously non-palpable hypertrophied pylorus to be felt during peristaltic contractions. The test feed should be aspirated afterwards. The best position for palpation is on the infant’s left side, potentially with the infant on his/her

mother’s lap. The inability to palpate an olive-shaped mass does not exclude the diagnosis of HPS, and often an ultrasound is needed to clarify the diagnosis.[7]

With an established HPS, prolonged vomiting of gastric contents leads to depletion of sodium, potassium and hydrochloric acid, which results in the characteristic finding of hypokalaemic, hypochloraemic metabolic alkalosis.[8] The kidneys conserve sodium at the expense of hydrogen ions, resulting in a paradoxical aciduria. With the increasing degree of dehydration, renal potassium losses are accelerated in an attempt to retain sodium and fluid.

Imaging studies

Historically the palpation of an olive-shaped mass may obviate the need for a confirmatory imaging study, as a positive examination has high specificity.[9] Plain radiographs will often have been performed, although they often are of no diagnostic value. They may demonstrate a dilated stomach with minimal distal air when there is a complete gastric outflow obstruction.

Ultrasonography (USS) is now the diagnostic test of choice as it can be performed quickly and without radiation exposure. The accuracy is close to 100% when performed by experienced personnel,[10] having a sensitivity and specificity of 99.5% and 100%, respectively.[11] The sonographic finding of HPS is characterised by a classic ‘target’ sign on transverse view. The measurements most commonly used are pyloric muscle thickness, pyloric muscle length and pyloric diameter. A muscle thickness of the pylorus greater than 4 mm and a pyloric channel length of greater than 17 mm yield a positive predictive value of greater than 90%. For infants less than 30 days of age, these limits may be lower.[12] Other findings include the failure of the channel to open during a 15-minute period of scanning, retrograde or hyperperistaltic contractions and an antral nipple sign.

As USS is widely available and routinely ultilised, a contrast upper gastrointestinal study is rarely needed. Its remaining diagnostic role is either when USS is not available or when there is a possibility of incomplete pyloromyotomy in the post-operative period. Positive contrast findings include an elongated pylorus with antral indentation from the hypertrophied muscle. The pathognomonic finding is the appearance of a ‘railroad track’ sign caused by two thin parallel streams of contrast traversing the pylorus. There is also a vigorously peristaltic stomach with delayed or no gastric emptying.

Upper GI endoscopy is performed on the very rare occasions when other

imaging modalities are inconclusive, and, although it would demonstrate pyloric obstruction, it would be difficult to differentiate it from pyloric spasm.

Differential diagnosis

The diagnosis is made by the characteristic clinical manifestation of projectile non-bilious vomiting and the finding of a pathognomonic pyloric mass. Other causes of vomiting, especially in early life, include severe GORD, duodenal stenosis or web, malrotation with partial volvulus or the presence of an annular pancreas. GORD may have a similar presentation of persistent vomiting after feeding. Other non-pathological causes include inexperienced feeding technique and overfeeding.

Metabolic causes may mimic pyloric stenosis. Adrenal insufficiency results in profuse non-bilious vomiting, but it is likely to result in metabolic acidosis rather than alkalosis. Unlike pyloric stenosis, the serum potassium and urinary sodium concentration are also elevated. Recurrent emesis with alkalosis or acidosis may be caused by some inborn errors of metabolism, but usually there will be other associated clinical features, such as hypoglycaemia, coma or seizures to suggest a metabolic cause. Any vomiting infant needs to have a clean urine check to exclude infection, as it is common for a urinary tract infection to present with vomiting.

Management

The definitive treatment is surgical intervention with a Ramstedt pyloromyotomy. This should only occur once the metabolic derangement has been corrected, preventing the anaesthetic respiratory complications of chronic hypercapnia. In this operation the hypertrophic muscle is incised from the prepyloric vein of Mayo onto the gastric antrum. The muscle is split leaving the mucosal layer intact. Traditionally a right upper quadrant transverse incision was used, but currently the majority of procedures are performed either via an open supra-umbilical or laparoscopic technique. Both approaches have been demonstrated to be equivalent in outcome.[13]

The preoperative treatment is directed toward correction of fluid, electrolyte and acid–base imbalance, which is vital to achieve prior to anaesthesia. The amount of fluid resuscitation is based on the degree of dehydration. Correction of fluid and electrolyte imbalance can usually be achieved within 24–48 hours.

Resuscitation with a bolus dose of intravenous normal saline is required for moderate to severe dehydration and is given at 10–20 mL kg^{-1}. This is followed by rehydration with 0.9% saline + 5% dextrose solution at 150 mL kg day.

Adequate amounts of both chloride and potassium are necessary to correct metabolic acidosis. The correction of potassium can be achieved by adding 10–20 mEq of KCl per 500 mL of intravenous fluid in patients with normal renal function. In severe dehydration, the kidney function should be assessed prior to adding potassium to the intravenous fluid. Chloride can be adequately replaced by normal saline with 5% dextrose. During resuscitation, urine output and electrolyte determinations should be performed regularly. In general, correction of chloride level to 90 mEq L^{-1} or greater is adequate for surgery to be performed.

Complications

Pyloromyotomy is associated with a low incidence of morbidity and mortality. A retrospective review of a large historical cohort of patients has revealed a decreasing complication rate to less than 5%.[14] Potential complications include an incomplete pyloromyotomy or a perforation in the duodenum or stomach. The mortality associated with this procedure is less than 0.4% in most major centres.[15]

Controversies

1 There are different thresholds for investigation of a vomiting infant. This varies between centers and has also been impacted by the increased use of USS. A USS performed too early in the hypertrophy process may be falsely reassuring with an equivocal pyloric measurement.

2 It is an acquired skill to palpate a pyloric mass in infants, and patience is required. With reliance on USS diagnosis this clinical skill is gradually being lost.

3 There is some evidence that the laparoscopic approach leads to more incomplete pyloromyotomies, but this is being further investigated.[16]

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7.6

Ingested foreign bodies

Scott Pearson

ESSENTIALS

1 Foreign bodies in the stomach or intestines are expected to pass spontaneously about 99% of the time if the child has normal anatomy.

2 Most complications occur when a foreign body is lodged in the oesophagus. Patients with oesophageal foreign bodies should be assessed urgently.

3 Children who ingest a metallic foreign body and attend an emergency department (ED) should have X-ray localisation of the object. For coin ingestions, an alternative is localisation with a hand held metal detector.

4 Removal of disc batteries lodged in the oesophagus should occur urgently as mucosal injury can occur within 2 hours.

5 Serious complications can occur in a delayed fashion in children who have had oesophageal mucosal injury from disc batteries.

Introduction

Ingestion of foreign (non-food) material is common in early childhood and often goes undetected whilst the child is playing and may not prompt a physician visit. The exact frequency of reported foreign body ingestions is uncertain. The literature in this field can be divided into three areas: descriptive studies of fairly large numbers of ingestion cases; studies primarily or exclusively about coin

ingestion; and studies about disc batteries.[1] The rise of serious complications and death from button battery ingestion is particularly alarming and has led to updated guidelines. Common foreign bodies ingested that come to medical attention include coins, bones (fish, chicken), other metallic objects (pins, screws, keys, batteries, magnets), toys, crayons, and plastic and rubber foreign bodies. In one large series of 1265 reported cases of foreign-body ingestions, age ranged from 7 months to 16 years with a mean of 5.2 years.[2] The peak ages for paediatric foreign body ingestions are 1–4 years. Most foreign bodies pass through the gastrointestinal tract without complications. The emergency physician should be aware of the few instances when emergent or semi-urgent intervention is indicated. Parents need to have clear guidelines regarding the treatment plan for children who are discharged to outpatient follow-up.

History

In most instances, a thorough history can be obtained from parents or caregivers before a requirement for intervention. The nature of the ingested item is obviously imperative, as is the time of ingestion. The ingestion may have been witnessed or may have been reported (by an older child) or implied by the child's environment at the onset of symptomatology. It can be extremely useful if a replica of the foreign body can be easily obtained, especially in determining the type and size of disc batteries. Determining the immediate environment of the child at the time of ingestion can assist in revealing the possibility of any likely co-ingestants.

The symptoms experienced by the child since ingestion help determine the likely site of the foreign body, but this has limitations. Many children are asymptomatic at presentation, which usually (but not always) suggests that the foreign body is lying in the stomach or more distal part of the gastrointestinal tract. Symptoms of vomiting, chest or throat pain or discomfort on swallowing, drooling, irritability and refusal to take food or fluids, coughing or gagging when eating or drinking may occur and suggest oesophageal foreign body. Several reports note that some children will be asymptomatic with foreign bodies lodged in the oesophagus, especially the distal oesophagus.[3,4] Even in the context of sharp fish bones, a prospective study found that symptoms were a poor predictor of the presence of fish bones, except for a sharp pricking sensation on swallowing.[5] Reports of abdominal pain or blood in the bowel motions should be noted. A history of previous oesophageal or other gastrointestinal disease is

significant in determining a management plan and alerting one to potential complications. Significant developmental/intellectual delay has been associated with major morbidity and mortality after foreign body ingestion.[2,6] This is often due to the vague symptomatology and delay in presentation.

Examination

For the majority of children the foreign body will have negotiated the oesophagus, and general examination will be unremarkable. Vital signs should be recorded but once again are unlikely to be abnormal unless there is delayed presentation and the development of a complication.

Palpation of the abdomen will usually be normal. If the history alerts one to abdominal symptoms, the examiner may find localised tenderness or peritonism suggestive of intestinal perforation. This is rare.

Some children will have symptoms of proximal oesophageal or pharyngeal foreign body. In the older child or adolescent where this is due to bony ingestion (e.g. fish or chicken) there should be a careful examination of the oral cavity and pharynx by either indirect or direct laryngoscopy. Younger children are unlikely to cooperate with these procedures without sedation and an experienced examiner.

Investigations

All children with a history of ingestion of coins or batteries (or other radio-opaque foreign bodies) should have an X-ray performed to localise the foreign body. Care should be taken to ensure the X-ray includes neck and chest. There is some controversy on this issue in relation to the need for X-rays in children who have ingested coins. Some authors (mainly hospital-based) note that even previously healthy children can be asymptomatic with an oesophageal coin and advocate early removal of oesophageal coins to prevent serious sequelae.[3,4] Other authors (notably in primary care) believe that routine X-rays in children having ingested a coin are unnecessary given that asymptomatic coin ingestion is rarely, if ever, associated with complications in otherwise healthy children.[7]

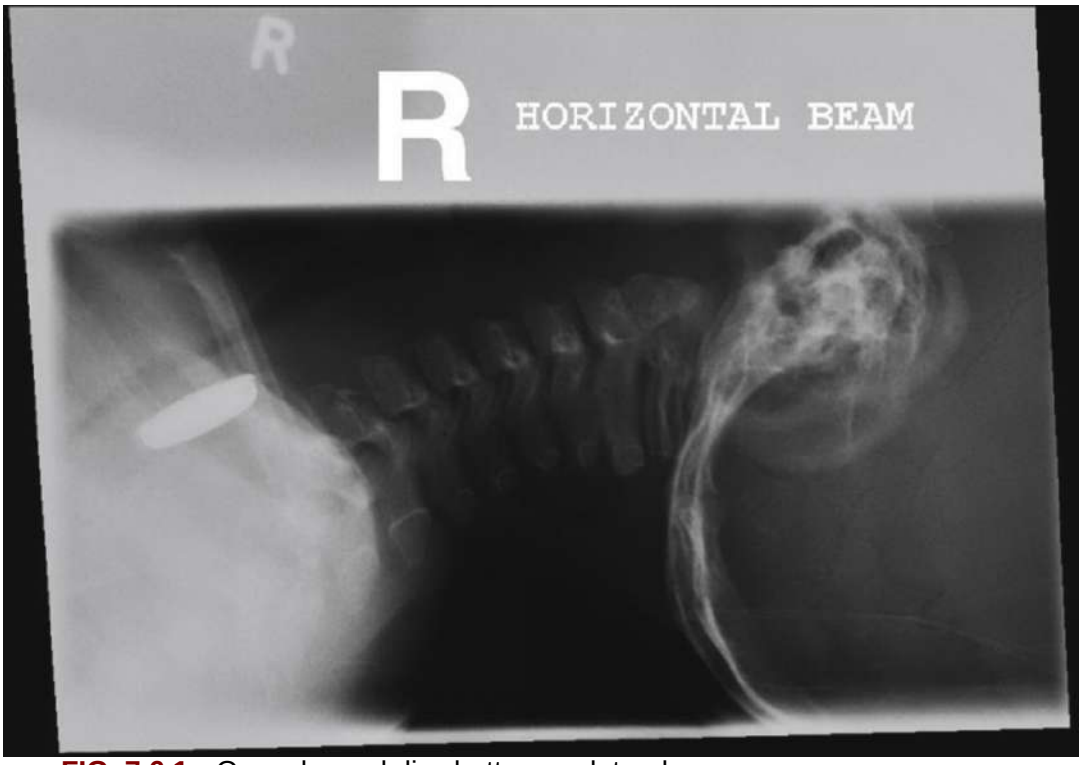

FIG. 7.6.1 Oesophageal disc battery on lateral.

It has been recognised that those patients presenting to an emergency department (ED) are a selected group and would be expected to have greater severity of symptoms and higher frequency of complications.[8] Hence the recommendation that all children with a history of coin ingestion *who attend an ED* should have X-ray localisation of the coin performed.

An alternative approach for coin ingestions is the use of a handheld metal detector for localisation of the coin. Several authors have confirmed the safety of this approach following a clear algorithm. Handheld metal detectors are not reliable at detecting metal foreign bodies other than coins.[9,10]

If the ingestion is unwitnessed and the object looks like a coin on anteroposterior X-ray, the clinician should be aware that a disc battery may have a similar appearance (Fig. 7.6.2). In this situation, an additional lateral view may reveal an asymmetry (Fig. 7.6.1), as the two sides of the disc battery have slightly different diameters. However, this is unreliable as there are some slimmer designed button batteries now available that are indistinguishable from a coin on lateral image. On the magnified anteroposterior view one may also see the ‘double ring or halo effect’ of both circumferences of the battery (Fig. 7.6.2).

Treatment

Coins

Coins that reach the stomach almost always pass through the gastrointestinal tract without incident, and further management is unnecessary unless symptoms arise. Coins that lodge in the oesophagus and have been unrecognised can cause complications such as oesophageal perforation, mediastinitis, acquired tracheo-oesophageal or aorto-oesophageal fistula formation – and death has been reported.[11] All children who are symptomatic should have a procedure to remove the coin. Asymptomatic children with proximal or middle-oesophageal coins should also have the coin removed as it is unlikely to pass spontaneously (20–30% in one series).[3] If there is a significant delay before the procedure (of more than a few hours) the X-ray should be repeated to ensure that spontaneous passage into the stomach has not occurred, or a metal detector should be used to check if the position has clearly changed. Asymptomatic children with distal oesophageal coins can safely be observed as outpatients (depending on social circumstances) for 12–24 hours. The chance of spontaneous passage into the stomach has been reported to occur in 37–60% of patients.[3,4] If the distal oesophageal coin has not progressed into the stomach by 12–24 hours, it should be removed endoscopically.

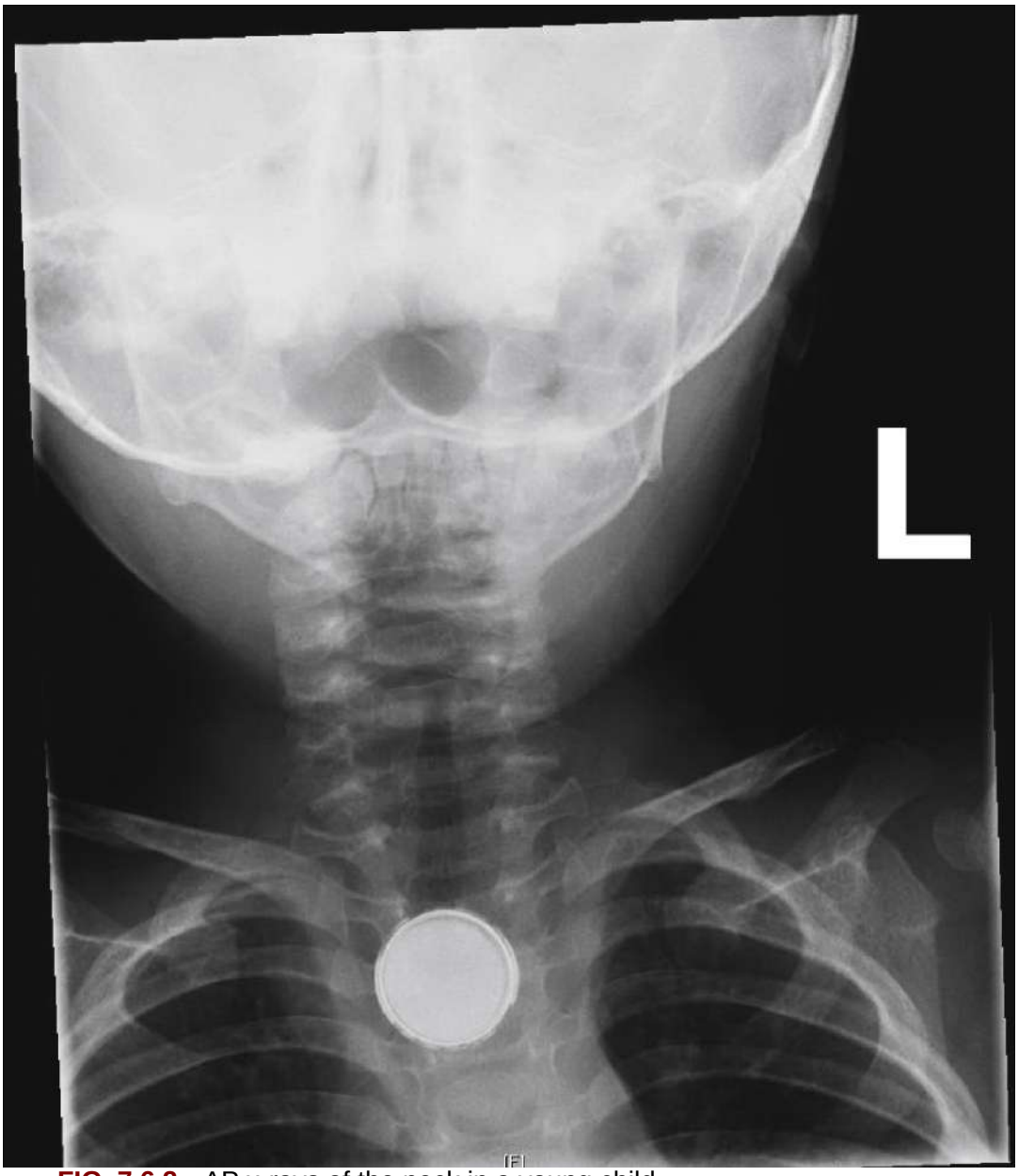

FIG. 7.6.2 AP x-rays of the neck in a young child.

Glucagon has been found to be ineffective when studied prospectively in the management of oesophageal coins. Children should be referred to a specialist with expertise in paediatric endoscopy for endoscopic removal under appropriate sedation or anaesthesia. This may require transfer of the child to another institution, as local resources dictate. Other techniques have been described, such as oesophageal bougienage or Foley catheter extraction, but most emergency physicians will not manage sufficient children with this problem to develop expertise and appropriate safety of the procedure.

Magnets

Small magnets used in toys may be ingested and can cause bowel injury, primarily by the potential for enteroenteric fistula formation between magnets in adjacent loops of bowel. Recent guidelines support the urgent removal of multiple magnets by endoscopic techniques.[12] Close follow-up with regular X-rays and laparoscopic intervention as needed may be necessary if the magnets are too distal for endoscopic retrieval.

Disc (button) batteries

Disc batteries are used for hearing aids, electronic devices and children's toys. Over the last 10 years there has been a dramatic increase in severe injuries and deaths associated with children ingesting button batteries. Smaller batteries will often pass through the gastrointestinal tract uneventfully. However batteries 20 mm and larger can lodge in the oesophagus more easily and lead to serious complications and death.

Battery size, voltage, location and local environment can all affect the speed of injury. The larger 20 mm batteries have a higher (3V) voltage. These are often lithium cells. Hydroxide ions are generated at the negative pole of the battery caused by the current created through the adjacent tissue. The hydroxide ion accumulation is equivalent to an alkaline caustic injury leading to tissue liquification and necrosis. Tissue pH >10 can rapidly occur causing a powerful alkaline burn. Higher voltages cause a faster injury.[13]

FIG. 7.6.3 New Zealand 10 cent coin compared to 20mm disc battery.

The speed of injury should be emphasised. **An oesophageal battery needs urgent removal** in a centre capable of paediatric endoscopy. This may require urgent transfer of the child to another facility. Other techniques of removal are inadvisable as they do not allow inspection of the mucosal injury and can cause inadvertent injury. The location of the most severe mucosal injury can be remembered by the 3 ‘N’s- Negative-Narrow-Necrotic. The *negative* battery pole which is the *narrowest* on lateral X-ray causes the most severe *necrotic* injury.[13,15]

Complications include local mucosal injury through to perforation and mediastinitis, stricture formation, vocal cord paralysis, tracheo-oesphageal fistula, spondylodiscitis, or major arterial haemorrhage from aorto-oesophageal fistula. Complications frequently develop after battery removal suggesting ongoing mucosal injury and inflammation. Fatal bleeding from aorto-oesophageal fistula has been described 28 days after battery removal. Strictures may take some months to develop. Use of other modalities such as contrast oesophagram, MRI and CT angiography are used to define adjacent vascular structures and the extent of inflammatory changes after battery removal. These are usually the domain of those providing ongoing care of the child. The emergency physician should be aware of the risk of delayed potentially life-threatening complications should a child re-present with symptoms after recent

button battery ingestion.

Once button batteries have reached the stomach, they usually pass through the gastrointestional tract uneventfully. Guidelines from the National Battery Ingestion Hotline (NBIH)[15] (Fig. 7.6.4) and Button Battery Taskforce[13] currently advocate abdominal X-rays and observation in asymptomatic patients with batteries distal to the oesphagus. In higher-risk patients (<6 years of age and battery ≥15 mm), a repeat X-ray is recommended in 4 days, while a repeat X-ray is recommended in 10–14 days in low-risk patients. Endoscopic removal is recommended in high-risk patients at 4 days if the battery remains in the stomach. However, other guidelines[12] recommend routine endoscopy even in asymptomatic patients who are high risk (<5 years of age and battery ≥20 mm). This recommendation is based on concern of oesophageal injury occurring before the battery has reached the stomach. Endoscopy is therefore primarily to facilitate inspection of the oesphageal mucosa.

Other metallic foreign bodies

Foreign bodies should be localised by X-ray and removed if lodged in the oesophagus. Sharp objects that have passed into the stomach, including open safety pins, may cause intestinal complications and patients should be referred for consideration of removal, usually endoscopically. An observational approach may be appropriate with some small sharp objects, but consultation is advisable. In certain situations, where the object is lodged in the stomach, it may be appropriate to repeat an X-ray to ensure passage into the intestine. In many situations repeat X-rays are unnecessary unless symptoms develop. Larger blunt objects longer than 3 cm (in children <12 months of age) or 5 cm (in children over 12 months of age) should be removed as they are unlikely to pass the pylorus.[14] A more cautious approach, and consultation, is advisable when there is abnormal gastrointestinal anatomy.

Non-metallic foreign bodies

Sharp foreign material such as fish or chicken bones will usually impact in the pharynx or oral cavity and may be removed under direct vision in the ED. Foreign body removal often needs to be facilitated by good lighting, appropriate equipment, local anaesthetic spray and sometimes careful procedural sedation. Consultation with ENT for consideration of nasoendoscopy may be required for

foreign bodies which are unable to be visualised by ED clinicians.

X-rays may assist in localisation when the ingested object is unable to visualised in the oral cavity or pharynx, but many foreign bodies are not radio-opaque. Intervention is dictated by the presence or persistence of symptoms. The vast majority of other objects pass through uneventfully. Plastic bread-bag tags have been associated with small bowel obstruction and perforation. If there are symptoms of lodgement in the oesophagus the foreign body should be removed by gastroscopy; otherwise an observant approach is adequate, with the parents or caregiver receiving clear advice on when to return to the hospital.

Disposition

Most children will be managed in the ED and discharged. A minority will be asked to return for further X-ray imaging, but many will be advised to return only if symptoms (abdominal pain, fever, vomiting) or complications (bleeding) develop. Parents should not be routinely encouraged to examine the child's stool to ensure passage of a foreign body.

Where admission is required this should be to a facility that is skilled in the endoscopic and surgical management of these children.

Prevention

Together with child-proofing the home to ensure that children are protected from access to potential poisons, this should extend to protection from small items that pre-school-age children can ingest. Battery compartments in equipment and toys are now made increasingly secure by manufacturers. Disc battery design is moving towards design of batteries that are inactive outside their device.[13]

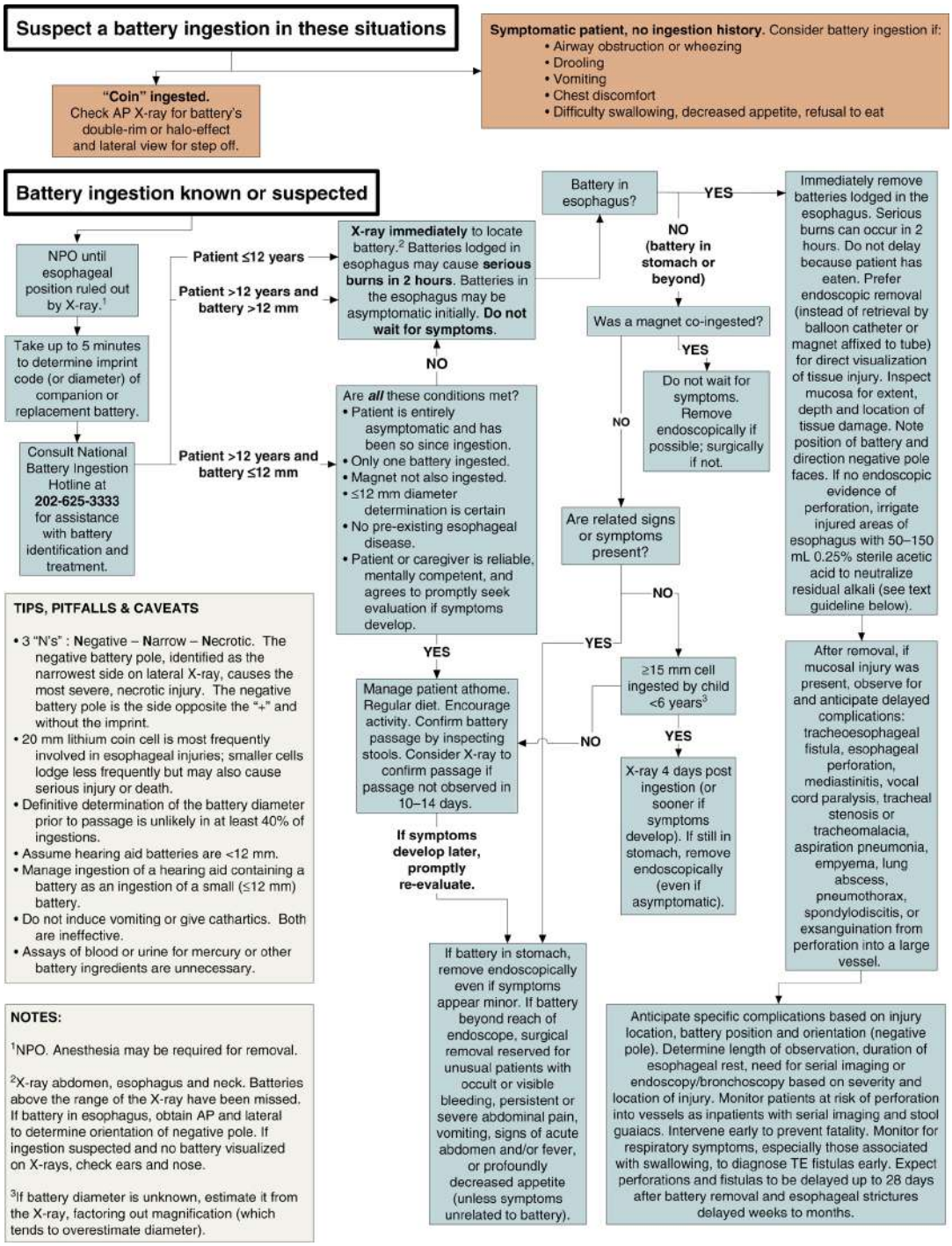

FIG. 7.6.4 NBIH triage and treatment algorithm for battery ingestions. Reproduced with permission from http://www.poison.org/battery/guideline and adapted from: Litovitz T, Whitaker N, Clark L, et al. Emerging battery-ingestion hazard: clinical implications. *Pediatrics* 2010;**125**(6):1168–77.

Controversies

Methods of removal of oesophageal coins. There are some advocates of Foley

catheter removal or oesophageal bougienage by emergency physicians or surgeons. Most centres utilise endoscopic removal by trained endoscopists.

Most children who are asymptomatic with sub-diaphragmatic button batteries require no further intervention. However, there are some higher-risk groups based on age and button battery size that may require endoscopy to enable inspection of the oesphageal mucosa.

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7.7

Acute liver failure

David M. Krieser

ESSENTIALS

1 Acute liver failure implies evidence of hepatocellular failure. Synthetic, excretory and metabolic processes may be affected.

2 Acute liver failure is a rare but serious problem in the paediatric population.

3 Consider the following causes: infectious, toxin-mediated, congenital (structural or metabolic) or infiltrative (malignancy, storage disease).

4 Infectious hepatitis is the most common cause worldwide; paracetamol toxicity the most common in the developed world.

5 Clinical information is vital to target appropriate investigation and treatment.

6 Management is largely supportive and aims to maintain homeostasis until hepatic recovery or transplantation occurs.

7 Liaison with, and transport to, a paediatric liver transplantation centre is usually required. Consultation should occur early.

Introduction

Acute liver failure (ALF) is a rare but devastating presentation in children. The liver performs synthetic, metabolic and excretory functions. Synthetic functions include production of coagulation factors and albumin. Metabolic and excretory functions include glucose metabolism and waste product processing (e.g. bilirubin, nitrogenous compounds, drug elimination). The manifestations of

coagulopathy, hypoglycaemia, jaundice, encephalopathy and hypoalbuminaemia reflect disturbances of liver function.[1] Examples of the causes of ALF in children appear in Table 7.7.1. ALF may be immediately life threatening or a subacute process, with a spectrum of severity between those extremes. Medical management must support vital functions while hepatic recovery occurs or liver transplantation can be performed.

ALF has been defined in adults by clinical and laboratory criteria:

1. Hepatocellular dysfunction (e.g. jaundice, coagulopathy) of rapid onset
2. Encephalopathy within 8 weeks of jaundice.

The Pediatric Acute Liver Failure Study Group (2) has defined ALF in children as:

1. biochemical evidence of liver injury (usually of less than 8 weeks' duration)
2. no history of chronic liver disease
3. coagulopathy that is not corrected with vitamin K
4. an international normalised ratio (INR) greater than 1.5 if accompanied by encephalopathy or greater than 2 if not accompanied by encephalopathy.

The paediatric definition, with less emphasis on encephalopathy, developed because the identification of encephalopathy, especially in infants and young children, can be very difficult. In addition, the onset of the illness may not be clear, particularly in metabolic disorders. Bhaduri and Milli-Vergani have provided a definition for clinical and research purposes: 'fulminant hepatic failure is a rare multisystem disorder in which severe impairment of liver function, with or without encephalopathy, occurs in association with hepatocellular necrosis in a patient with no recognized underlying chronic liver disease'.[3] For children with chronic liver disease who present with features of ALF, the management principles are similar; although where specific therapy is available for an underlying disease, then this should be considered as well.

ALF classification, using the time interval between the onset of jaundice and encephalopathy, has aetiological and prognostic importance (Table 7.7.2), despite the difficulties in identifying encephalopathy. O'Grady et al.[4,5] and Poddar et al.[6] found that, in comparison with patients suffering acute or subacute

liver failure, those with hyperacute liver failure had a better prognosis. Mortality is associated with: age under 1 year, INR over 4, plasma ammonia over 200 micromol/L, dialysis requirements prior to transplantation and stage 4 encephalopathy (where identifiable).[7]

Table 7.7.1

Aetiology of liver failure

Neonates and infants		Older children	
Cholestasis	Biliary atresia Choledochal cyst Intra-hepatic bile duct paucity (Alagille syndrome) Inspissated bile syndrome	Infection: viral	Hepatitis A–D Enteroviruses Epstein–Barr virus (EBV) Varicella
Idiopathic neonatal hepatitis			Adenovirus
Cystic fibrosis			Cytomegalovirus (CMV)
Endocrine	Hypopituitarism Hypothyroidism		Herpes simplex Rubella
Neonatal haemochromatosis		Infection: bacterial	*Listeria* Tuberculosis
Infection: viral	CMV Herpes simplex virus/human herpesvirus 6/varicella-zoster virus EBV Parvovirus Rubella Reovirus type 3 Adenovirus Enterovirus	Infection: parasitic	Toxoplasma
		Toxins and drugs	
		Malignancy	Leukemic infiltration Lymphoma Neuroblastoma Primary hepatic tumours
		Metabolic	Wilson's disease
		Hepatic venous occlusion	
Infection: bacterial	Bacterial sepsis Urinary tract infection	Fatty liver	Obesity Pregnancy
	Tuberculosis Syphilis	Hepatic hypoperfusion	Cardiogenic shock Hypovolemic shock
Infection: parasitic	Toxoplasma		Septic shock
Metabolic disease	Peroxisome function abnormality (Zellweger's) α1-Antitrypsin deficiency Bile acid metabolism Urea cycle abnormalities Amino acid metabolism abnormalities Lipid metabolism abnormalities (Gaucher, Wolman, Niemann Pick C) Carbohydrate metabolism abnormalities (galactosaemia, fructosaemia, type IV glycogen storage disease)		
Toxins	Paracetamol TPN Hypervitaminosis A		
Tumour	Intra- and extra-hepatic		

Aetiology

The aetiology can be grouped according to onset prior to, or after, the first year of life (see Table 7.7.1). In broad terms, infection, immune dysregulation, toxicity (including medication), infiltration, and inborn errors of metabolism are the causative pathways that may lead to ALF. Cases where the cause is not determined predominate in children under 3 years.

In 80 infants under 12 months with ALF, inherited metabolic conditions were responsible for 42.5% of cases: neonatal haemochromatosis 16%, acute viral hepatitis 15%, and miscellaneous causes (toxins, autoimmune, malignancy) 10%. Sixteen percent of neonatal cases were undetermined.[8] Sundaram[9] reported on a cohort of 148 infants under 90 days of age in the PALF study group. Five diagnostic groups were defined in this report: metabolic disease, neonatal

haemochromatosis, viral infection, other and indeterminate. The Asia-Pacific Association for the Study of Liver Diseases[10] described 499 children with chronic liver disease presenting with acute-on-chronic liver failure; Wilson disease (42.8%) and autoimmune hepatitis (32.1%) predominated. Of note, 30% of the children with underlying chronic liver disease deteriorated due to acute viral hepatitis.

Table 7.7.2

Classification of acute liver failure

Interval between onset of jaundice and encephalopathy	Classification
7 days or less	Hyperacute
8 to 28 days	Acute
5 to 12 weeks	Subacute

In the neonatal population specifically, four causes predominate: gestational alloimune liver disease-neonatal haemochromatosis, viral infection, hereditary haemophagocytic histiocytosis and mitochondrial disorders.[11] There are some classification differences here that may influence this classification. Taylor and Whitington[11] describe gestational alloimmunity leading to liver insult and iron infiltration as the most common causes of neonatal ALF. Neonatal haemochromatosis is described by Bonilla et al. as a ‘phenotype’ where iron overload and tissue siderosis result from impaired control of maternofetal iron exchange.[12]

Infectious hepatitis

Worldwide, infectious hepatitis, due to five RNA viruses (hepatitis A, C, D, E and G) and one DNA virus (hepatitis B), is the greatest cause of ALF. Transmission of hepatitis A and hepatitis E is via the faecal-oral route. The others are transmitted via body fluids. Acute viral hepatitis is a clinical syndrome with systemic symptoms occurring after a virus-dependent incubation period. Jaundice ensues after hepatocyte necrosis reduces the liver’s capacity to metabolise bilirubin.

Fulminant hepatitis occurs in less than 1% of children with hepatitis A and in 1–2% of cases of hepatitis B. Fulminant disease occurs with hepatitis D in approximately 10% of cases. It is more likely in those with chronic hepatitis B

who subsequently become infected with hepatitis D (superinfection), than in patients who acquire both viruses simultaneously (co-infection). Hepatitis C can cause acute, chronic and, rarely, fulminant hepatitis. Severe acute hepatitis E infection is a leading cause of ALF in the tropics.

Epstein–Barr virus, cytomegalovirus, herpes simplex virus, varicella zoster virus, human herpes virus 6 and parvovirus B-19 are non-hepatotropic viruses that can rarely cause ALF. Consideration of, and investigation for, these viruses is important, because specific therapy with antiviral medication (e.g. aciclovir for herpes simplex virus) is available for some of these pathogens.[13]

Toxins and medication

Paracetamol

Paracetamol toxicity is the most common cause of ALF in the developed world. In Australia and New Zealand, paracetamol was the cause of ALF in 25.9% of children admitted to New Zealand and Queensland Paediatric Liver Transplant Services.[14] Paracetamol is metabolised by the hepatocyte and toxicity exhausts hepatic glutathione stores. Generation of toxic metabolites leads to centrilobular necrosis. Toxicity is unlikely with single doses under 150 mg/kg. Children are often given multiple doses of paracetamol and this can lead to toxicity if the cumulative daily dose is greater than 60 mg/kg/day. Factors predictive of hepatotoxicity include: age (lower incidence in children under 5), genetics (cytochrome isoenzyme polymorphisms are inherited),[15] alcohol and tobacco use (relevant in adolescents), other medications and nutritional status.[16] Treatment of toxicity is discussed elsewhere in the text (Chapter 21.2).

Anticonvulsants

Genetic predisposition has been proposed for anticonvulsant induced hepatotoxicity.[2] Sodium valproate causes intracellular fat accumulation within the hepatocyte, and may be related to a primary defect of respiratory chain enzyme function.[2] Impaired metabolic functions within the cell may lead to necrosis. Children under 2 years and those on multiple medications are at highest risk. Carbamazepine may cause hepatitis and/or cholestasis during the first months of therapy. Clinically significant hepatotoxicity is rare.

Aspirin and Reye’s syndrome

Mitochondrial dysfunction leading to acute encephalopathy, selective hepatic dysfunction and visceral fatty infiltration has been called Reye's syndrome.[17] Mitochondrial oxidative phosphorylation and fatty acid β-oxidation are the metabolic pathways affected in Reye's syndrome. Metabolic disorders have been later identified in some children initially diagnosed with Reye's syndrome. Preceding viral infection (classically varicella), immune mediators and aspirin (or its metabolites) can limit normal functioning of these pathways. The association of aspirin with this disorder remains unclear; however, Forsyth et al.[18] identified a dose–response relationship, and population studies demonstrate that the decline in Reye's syndrome mirrors a decline in aspirin usage.[17]

Mushrooms

Edible and inedible mushrooms can be difficult to distinguish. *Amanita phalloides* produces amatoxin, which is hepatotoxic. The toxin is a heat stable octapeptide. After a period of 6–48 hours vomiting, abdominal pain, and diarrhoea usually precede neurological symptoms (coma, seizures) and ALF. Mortality of 30% was reported[19] in Australia. A cohort study from The American Acute Liver Failure Study Group reported a mortality of 15%.[20] Cholinergic symptoms via muscarinic receptors may also occur and respond to atropine. Charcoal may be given to reduce absorption. Silibinin and high-dose penicillin G may assist in limiting hepatic damage.[21] Identification of the mushroom is important and may require referral to local botanists or mycologists.

Metabolic diseases associated with liver failure are summarised in Table 7.7.3.

Structural anomalies

Alagille syndrome

This autosomal dominant condition leads to intra-hepatic biliary hypoplasia in association with cardiac, renal, ocular, facial, and skeletal abnormalities. Alagille syndrome has an incidence of 1:100,000 live births and is associated with abnormalities of chromosome 20. Peripheral pulmonary arterial stenosis, with or without pulmonary valvular stenosis, is the most common cardiac defect. Vertebral anomalies described as 'butterfly vertebrae' constitute the most common skeletal defect. Retinal pigmentation and posterior embryotoxon (requiring slit lamp examination) are present in the eye. The facies are

‘triangular’ with a broad forehead and a pointed mandible. Failure to thrive and jaundice are common presentations. Hepatic cirrhosis can develop in up to 50% of children.

Table 7.7.3

Metabolic disease associated with liver failure

Disease	Chromosome(s) involved	Incidence / inheritance	Clinicopathological features
Zellweger's (cerebrohepatorenal) syndrome	Chromosomes: 1, 2, 6-8, 11, 12, 22; involving multiple PEX genes	1:50,000 live births Autosomal Recessive	Biopsy demonstrates mitochondrial abnormalities and peroxisomal absence leading to abnormal fatty acid oxidation Multi-organ involvement (cardiac, pulmonary, neurological, renal), failure to gain weight, hypotonia Jaundice in 50%
α-1 antitrypsin deficiency	Chromosome 14	1:500–3500 in Caucasians; rare in Asians Autosomal Recessive	Isoelectric phenotyping of protease inhibitor (Pi) Pi ZZ most likely to have liver disease Others: Pi SZ, SS, FZ and MZ (carrier) Neonatal cholestasis, failure to gain weight, intrauterine growth retardation, coagulopathy (2%) Lung disease (bronchiectasis) is rare in children Liver transplant leads to phenotypic cure
Tyrosinaemia	Chromosome 15	1:100,000 worldwide (1:16,000 in Quebec) Autosomal Recessive	Deficiency of fumarylacetoacetase within the phenylalanine pathway Progressive liver parenchymal damage and renal tubulopathy (aminoaciduria) Rapid progression in infancy or indolent course with hepatic cell carcinoma in 37% of children over 2 years
Galactosaemia	Chromosome 9	1:40,000 live births Autosomal Recessive	Deficiency of galactose-1-phosphouridyl-transferase Accumulated galactose-1-phosphate in liver, brain and renal tubules Progressive jaundice, liver failure, hypoglycaemia, cataracts, gram-negative sepsis (increased susceptibility) Encephalopathy caused by liver failure and hypoglycaemia Diet control is required to manage and routine newborn screening is performed in many countries
Wilson's disease	Chromosome 13	1: 30,000 live births Autosomal Recessive	Disorder of copper metabolism Liver disease including acute liver failure (ALF), chronic active liver disease, cirrhosis Neuropsychiatric symptoms including behaviour change, tremor, dysarthria, learning difficulties Renal tubulopathy, haemolysis Kayser-Fleischer rings are brown bands seen at the corneal-iris border

Data Sources: https://www.omim.org/ and https://rarediseases.org/

Biliary atresia

Complete absence of extra-hepatic biliary structures leads to cholestasis, and a clinical picture of jaundice, pale (acholic) stools and dark urine. Erlichman and Loomes describe three catagories of biliary atresia:[22]

- Biliary atresia without associated anomalies:
    - 70–85% of infants with biliary atresia
    - gradual onset of jaundice, pale stools and dark urine within 2 months of birth
- Biliary atresia associated with laterality malformations:
    - 10–15% of infants with biliary atresia
    - associations include: situs inversus, asplenia/polysplenia, malrotation, and certain cardiac and vena cava anomalies
- Biliary atresia associated with other malformations:
    - 5–10% of infants with biliary atresia
    - associations include: intestinal atresia, imperforate anus, renal anomalies, and certain cardiac anomalies.

Biliary atresia occurs in approximately 1:10,000 to 1:20,000 live births with equal gender incidence. Jaundice will usually be the presenting feature and conjugated hyperbilirubinaemia identified after blood is sent. Conjugated hyperbilirubinemia is always abnormal. Following diagnostic testing (ultrasonography, nuclear medicine hepatic scan, occasionally liver biopsy), management is by surgical hepatoportoenterostomy (Kasai procedure) prior to 8 weeks of age, if possible, as earlier surgery improves outcomes.[23] Biliary atresia, in association with subsequent complications (cholangitis and portal hypertension) often leads to acute-on-chronic liver failure and transplantation. Biliary atresia is the most common indication for liver transplant in children.[24]

Pathophysiology

Exposure to hepatotoxic agents such as: drugs, products of metabolism or infectious particles, in addition to immune responses, initiates hepatocyte injury that may progress to necrosis. Biopsy, when performed, reveals multilobular or bridging necrosis with reticulin framework collapse. Patterns related to aetiology can be seen, such as centrilobular necrosis in paracetamol toxicity or with circulatory shock. When normal regenerative processes do not occur, liver failure follows.[7] Astrocyte oedema may be seen and may be due to altered cell-wall permeability, glutamate, ammonia, and neurotransmitter balance.[9]

Hepatic encephalopathy occurs through the interplay of three factors:

1 Reduction in synthesis of substances essential for normal brain function
2 Production of substances that are neurotoxic
3 Reduced elimination of neurotoxins.

Contributions from ammonia, inflammatory cytokines, benzodiazepine-like compounds and manganese[25] lead to neuronal dysfunction and altered interaction of astrocytes with neurons. The clinical manifestations of hepatic encephalopathy follow. The balance of inhibitory (e.g. GABA) versus excitatory (e.g. glutamate) neurotransmission is altered in hepatic encephalopathy.[17] Ammonia appears to augment inhibitory neurotransmission. In addition, the role of sepsis via systemic immune response,[26] via lipopolysaccharides,[27] with hypoglycaemia and/or raised intracranial pressure are important factors in the development of encephalopathy.

Presentation

History

History and examination findings are significantly influenced by the age of the child. Neonates may present with jaundice and care must be taken to differentiate physiological from pathological causes. The presence of acholic (pale) stools is characteristic of cholestasis (e.g. biliary atresia). Investigation (see below) will be directed at identifying the cause of cholestasis. Jaundice in the context of dysmorphic features, cardiac murmur and ocular abnormalities suggest Alagille syndrome or biliary atresia with associated congenital abnormality. The development of jaundice after a change in diet may suggest metabolic abnormalities of carbohydrate metabolism such as galactosaemia or hereditary fructose intolerance.

Infants and older children may present with a history of loss of appetite, vomiting, fevers, or abdominal pain, prior to the development of jaundice. Infectious hepatitis (e.g. hepatitis A, Epstein–Barr Virus [EBV] or cytomegalovirus [CMV]) is the most likely cause in this situation. EBV infection may be suggested by a history of sore throat and lymphadenopathy. A dietary and travel history may be relevant (hepatitis A). Hepatitis B and C infection needs to be considered if a history of exposure is obtained. In adolescents, sexual activity and the use of illicit drugs must be discussed while maintaining confidentially. In areas where strict screening is performed, children who receive blood product transfusions (e.g. malignancy, renal failure, haemophilia, haemoglobinopathy) are at very low risk for the acquisition of hepatitis B and C. An infant presenting with encephalopathy and jaundice may have a metabolic disease (fatty acid oxidation, carbohydrate metabolism). Note consanguinity as part of the family history. Previous surgery for a choledochal cyst or for biliary atresia is important in the context of a child presenting with jaundice and/or hepatic failure. Exposure to hepatotoxins such as paracetamol, anticonvulsants, aspirin or mushrooms must be identified, as specific treatments can be implemented. Pruritus with jaundice, dark urine and pale stools may be the presenting features of cholestasis in an older child.

Examination

Hepatomegaly and jaundice are the most frequent findings. The character of the liver edge may offer additional information:

- A firm, nodular surface suggesting cirrhosis or fibrosis
- Tenderness suggesting acute hepatitis
- Liver enlargement:
    - Note: in cirrhosis liver span may be reduced
- Splenomegaly suggests portal hypertension or infiltration (e.g. storage disease, extramedullary haematopoiesis, malignancy)
- Ascites.

As discussed above, specific diseases associated with liver failure will have other features. The phenotypes of galactosaemia, Alagille syndrome, α_1-antitrypsin deficiency and Wilson's disease may be identified by careful clinical examination. Neurological evaluation is essential and the presence of asterixis important in a child with liver failure in order to assess for, and classify a stage of, hepatic encephalopathy (see Table 7.7.4). In young children encephalopathy, which occurs late, is difficult to detect. Cutaneous features such as bruising, petechiae or bleeding may indicate an associated coagulopathy. In children with chronic liver disease there may be signs such as spider naevi, caput medusae or finger clubbing.

Investigations

Investigation of the child with ALF focuses on identifying the extent of liver dysfunction and on identifying the cause. History and examination findings must guide investigation. ED screen should include:

- full blood examination
- liver function tests including conjugated and unconjugated bilirubin
- blood glucose
- coagulation screen
- renal function
- blood gases: arterial or venous
- serum ammonia.

Table 7.7.4

Stages of hepatic encephalopathy

	I	II	III	IV	
				IVa	IVb
Symptoms	Lethargy, euphoria, poor concentration	Drowsiness, erratic behaviour, disorientation	Stuporous but rousable, incoherent speech	Responsive to pain	No response
Signs	Reduced cognitive performance (drawing figures, memory)	Asterixis, incontinence, fetor hepaticus	Asterixis, hyper reflexia, rigidity	No asterixis, areflexia, flaccidity	
EEG	Normal	Generalised slowing, theta waves	Markedly abnormal, triphasic waves	Markedly abnormal bilateral slowing, delta waves, cortical silence	

Source: Modified from Suchy FJ 2000 Fulminant hepatic failure. In Behrman RE (ed) Nelson Textbook of Pediatrics, 16 edn. Philadelphia: Elsevier.

Further potentially relevant investigations may be discussed with a hepatologist:

- Viral serology
- Copper level
- Caeruloplasmin levels
- Alpha1-antitrypsin phenotyping (Pi typing)
- Lactate level
- Drug screen:
    - Especially for paracetamol, or where history of ingestion is unclear
- Urine metabolic screen
- Relevant imaging (ultrasound, CT, MRI, nuclear medicine).

Note that the INR for coagulation and the serum bilirubin concentration are not predictive of post-transplant survival, while renal dysfunction requiring dialysis is associated with a higher mortality.[7] Escudie et al.[28] found a prothrombin index below 10% (INR approx 6) 4 days after Amanita phalloides ingestion was predictive of fatal outcome.

Management

The management of acute liver failure involves supportive care, complication management and specific treatment modalities where they exist. No intervention will enhance hepatic regeneration or hepatocyte injury.[7] Therapy needs to be initiated in the ED prior to transfer to a liver transplantation centre with paediatric intensive-care services. The initial management issues are listed in Table 7.7.5.

Intubation and ventilation may be required due to coma or respiratory failure. Respiratory failure itself is multifactorial: altered cardiac output; capillary leak; possible oliguria; and significant fluid requirements contribute.

Fluid management may be complex in the face of renal failure and electrolyte imbalances. Vitamin K, fresh frozen plasma or cryoprecipitate may be required to correct symptomatic coagulopathy. Intravenous fluids containing 10% dextrose, or more, correct and then maintain normoglycaemia.

Lactulose is given in hepatic encephalopathy to reduce absorption of nitrogenous wastes. Neomycin and/or metronidazole, given enterally, reduce the enteric bacterial load and the production of nitrogenous wastes.

Table 7.7.5

Initial management tasks

Management tasks	
Observation of:	Vital signs Pulse oximetry Level of consciousness Urine output (may need indwelling catheter)
Monitoring of:	ECG Blood glucose Acid–base status (blood gases) Coagulation Liver function tests Serum electrolytes
Insertion of:	Vascular access (peripheral, central, intraosseous) Nasogastric tube (for gastric drainage and administration of neomycin and lactulose)
Supplementation of:	Glucose Potassium Albumin Coagulation factors (vitamin K, frozen plasma, cryoprecipitate, platelets) Oxygen (may require endotracheal intubation) Intravascular volume if required
Identification of:	Treatable cause if present (toxic, infectious, metabolic) Evolving encephalopathy Raised intracranial pressure

Proton pump inhibitors, H_2-receptor blockers and/or sucralfate are given to limit the risk of gastric ulceration in the context of coagulopathy.

Sepsis is a common and serious complication, as it exacerbates liver failure. Aggressive antimicrobial therapy is required.

Raised intracranial pressure and, more specifically, reduced cerebral perfusion pressure is an important complication of ALF. Therapeutic cooling (32–33°C) reduces brain energy metabolism, normalises cerebral blood flow, reduces ammonia delivery, reduces oxidative pressure on astrocytes and reduces brain glutamate. [7]

A bridge to liver transplantation may be created via the use of liver support devices. Artificial systems using filtration, dialysis or ion exchange or bioartificial systems (human or non-human hepatocytes within an artificial framework) are available.[29] Anecdotal reports of these systems include reduction in serum copper in Wilson's disease induced ALF that allowed stabilisation prior to transplantation see p209.[30]

The development of liver transplantation has allowed children with irreversible liver failure to survive. The paucity of available donors and

contraindications to transplantation limit the number of children able to receive liver transplants in the acute setting. Contraindications to transplantation include: uncontrolled systemic infection, extrahepatic metastasis in liver tumours, irreversible neurological injury, and multiorgan failure.

Living donor transplantation has increased the number of liver transplants in children. Auxiliary partial orthotopic transplantation allows the transplanted liver segment to function while a diseased native liver recovers and regenerates. This technique also may allow for the discontinuity of antirejection medication once the native liver recovers.[31]

For paracetamol toxicity, disease-specific treatment, using N-acetylcysteine (NAC) in addition to supportive measures, is available. Herpes virus-induced fulminant hepatitis may respond to aciclovir. Children with Wilson's disease may respond to chelation therapy pending transplantation. Certain metabolic diseases can be managed via special diets or metabolic pathway manipulation.

Disposition

Management of the child with ALF requires a multidisciplinary team of physicians, nursing staff and allied health personnel. The invasive nature of monitoring and maintaining such patients necessitates transport to a paediatric intensive-care unit (PICU). Transportation will require stabilisation and continued monitoring of the above parameters, with intervention occurring en route, if necessary. Liver transplantation will require the involvement of surgeons and anaesthetists in addition to those already involved within the PICU. The transplant service should be consulted early in such cases.

Table 7.7.6

The King's College criteria

Features associated with a poor prognosis
• Unknown aetiology • Toxin associated (other than paracetamol) • Age under 10 years • Age over 40 years • Jaundice for more than 1 week prior to encephalopathy • Serum bilirubin over 300 μmol L^{-1} • International normalised ratio over 3.5 • pH less than 7.3 • Serum creatinine over 300 μmol/L

Source: Data from O'Grady et al 1989 Gastroenterology 97: 439–445.

Prognosis

The Pediatric Acute Liver Failure Study Group report that among 769 patients, mortality rates were linked to the presence of hepatic encephalopathy.[32] Overall 11% of children died, with a mortality of 55% in those with persistent grade III or IV hepatic encephalopathy. Seventy-nine percent of children without hepatic encephalopathy survived. Sepsis is the cause of death in approximately 10% of children with ALF.

Earlier work in establishing the Kings' College criteria (Table 7.7.6), provided clinical and laboratory parameters predictive of mortality. A single adverse factor was associated with 80% mortality; three adverse factors were associated with 95% mortality. Paracetamol toxicity was associated with a better prognosis. If paracetamol toxicity was the reason for ALF then the presence of a single adverse risk factor places mortality risk at 55%.

The United Network for Organ Sharing (UNOS) developed the Model for End-Stage Liver Disease (MELD) for patients over 12 years. The Pediatric End-Stage Liver Disease (PELD) score is for patients under 12. Both assess risks of death while waiting for transplantation in order to prioritise available organs. The PELD score uses: albumin, bilirubin, INR, growth failure and age at listing (for transplantation) while the MELD score uses creatinine, bilirubin and INR in predictive modelling (www.unos.org).

Prevention

Viral hepatitis requires public health and legislative intervention to be controlled. Immunisation against hepatitis A and B, improved hygiene, reduced over-crowding and the promotion of harm-minimisation through 'safe sex' and reduction in the sharing of needles among intravenous drug users may have impacts on the incidence of viral hepatitis. Vaccines for hepatitis A and B are available but expense and distribution problems act as barriers in the developing world.

The widespread availability of paracetamol contributes to its position as the leading cause of ALF in the developed world. Limiting packet size, 'child-proofed' containers and even reducing distribution to pharmacies alone may

reduce the incidence of paracetamol poisoning.

Screening of embryos or parents after the identification of a metabolic disease will assist in reducing the incidence of such diseases, if parents consider termination of pregnancy an option. Genetic counseling is mandatory in such cases. Direct gene therapy for such diseases remains elusive.

Controversies and Future Directions

MARS and other liver-replacement therapies are biological, partly biological or artificial. These systems use combinations of haemodialysis, haemofiltration and plasma exchange to remove wastes. Systems using implanted hepatocytes exist. While improvements in isolated liver function test parameters have been reported,[20] overall survival to successful transplantation has been elusive. [33] Transplantation remains the gold standard therapy and case reports of auxiliary partial orthotopic liver transplantation demonstrated that a transplanted liver lobe functioned satisfactorily until native liver recovery occurred in a case of mushroom toxicity. [31]

Cerebral cooling for management of raised intracranial pressure associated with ALF may offer some benefits and requires more research.

Prognostic scores have been developed according to aetiology of ALF and need more prospective research. In paracetamol hepatotoxicity arterial pH, serum lactate, prothrombin time and creatinine are used to predict outcome. In mushroom toxicity prothrombin time and creatinine appears more predictive. A patient with Wilson's disease requires transplantation if encephalopathy is present, although serum bilirubin, INR, aspartate aminotransferase and white cell count are important predictors of the need for transplantation. In general, predictors of poor outcome include: poor renal function, a PELD score >25, age under 24 months, and onset of encephalopathy within 7 days of onset of jaundice.[5]

Children with metabolic disease may benefit from liver transplantation, even if complete resolution of their disease cannot be expected. The improvements in quality of life have to be balanced against the risk of transplantation, the availability of donor liver and bioethical principles.

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7.8

Diarrhoea

Christopher Webber

ESSENTIALS

1 ‘ABC fluids in and out’ is a useful tool in taking a history.
2 Diarrhoea may or may not be associated with vomiting
3 Not all diarrhoea and vomiting is gastroenteritis.
4 The differential diagnoses are extensive and vary if acute or chronic.
5 Young infants with diarrhoea must be managed with caution.

Introduction

Common symptoms

Diarrhoea, especially associated with vomiting, is a common symptom that affects many infants and children. It may be acute or chronic.

Definitions

Diarrhoea refers to increased stool frequency and the consistency of stool being loose or liquid. The stool may also contain blood and/or mucous, both of which are abnormal.

Fever is present when the body’s temperature is elevated above normal. Actual definitions of fever vary, but a core temperature of about 36.5–37.2°C is normal. The site used to make the measurement also affects measured temperature. Generally aural (tympanic) temperatures are less accurate. Per-axilla temperatures are lower than rectal temperatures by about 0.5–1°C.

Clinical evaluation

History

Presenting complaint and past history

Always listen to the parents or carers of the child. Generally, they know the infant or child best and physicians are unwise to ignore their concerns. Explore the symptoms further and in more detail, if necessary. Understanding the normal stooling pattern of the infant or child is important. Check for blood or mucus in the stool, as well as associated symptoms like vomiting or fever. Sometimes important associated symptoms are not volunteered by parents, and are only alluded to when directed by a focused history.

Exploring the dietary intake, usual as well as current is important. Breastfeeding or the type of formula in the baby or infant is important, especially if the intake is lactose containing. In addition the persistent use of an oral rehydration solution after acute gastroenteritis without the reintroduction of diet helps refine the differential diagnoses.

In the child with a possible infectious condition, it may be important to clarify whether there have been infectious contacts or recent overseas travel.

Review of the past history should include any significant medical or surgical problems and whether the child is thriving. Failure to thrive and chronic diarrhoea will have a different constellation of differential diagnoses compared to acute diarrhoea.

ABC – fluids in and out

This schema provides a simple structure for obtaining the history, and may be used to advise parents about concerning symptoms that may develop subsequently. Alertness and activity (**A**) provides information about the neurological state of the child. Lethargy and poor interactivity are concerning symptoms in a young child. The behaviour of the baby whilst feeding is also important. Parents may note poor suck and sleepiness during feeding; and the breast-feeding mother may comment more precisely about the quality of the infant's suck. Breathing (**B**), specifically rapid or laboured breathing, and poor circulation (**C**), as indicated by mottling and coolness of the peripheries, are also worrying signs.

The child's fluid intake and urine output as a percentage of usual or normal provide very important markers of the unwell infant or child. The number and

‘wetness’ of the nappies is used as the indicator of urine output. Thus, using an evaluation of ‘fluids in and out’, one should be concerned when these are less than 50% of normal for the child.

Examination

General observation and examination

In the absence of a life-threatening emergency it is always worthwhile to make a careful observation of the child, either whilst talking with the carer or to the child. This general observation phase is invaluable in the paediatric assessment, particularly in young children. During this observation look and listen for the following:

1. Airway noises (stridor, stertor, grunt or wheeze)
2. Tachypnoea (measure the respiratory rate)
3. Colour (centrally and peripherally, look for cyanosis and mottling)
4. Alertness and activity (or inter-activity, particularly with child’s carer/s).

Note that the ABCD approach (airway, breathing, circulation, disability) is utilised.

Assessment of the state of hydration is critical. Accurate premorbid and current weight may aid this assessment. It is well recognised that clinicians tend to overestimate the degree of dehydration, consequently excessive amounts of intravenous fluid may be administered. Therefore, clinical assessment recommendations for dehydration have been revised to:

- <3% – no clinical signs; reduced urine output
- 3% – mild dehydration; mild tachycardia, dry mucous membranes
- 5% – moderate dehydration; lethargy, tachycardia, reduced skin turgor, sunken eyes/fontanelle
- 10% – severe dehydration; clinical signs of shock (tachycardia, thready pulses, reduced perfusion, particularly centrally).

Always check carefully for a rash, as petechiae and purpura may be inconspicuous or subtle.

Measure and plot the centiles (head circumference, length/height and weight). Serial measurements provide significantly more information than a single set of

measurements.

Cardiovascular and respiratory status

It is important to evaluate the cardiovascular and respiratory systems in the infant with severe diarrhoea, especially if there is concurrent vomiting, for the following reasons:

- Shock may be present due to either septicaemia or fluid loss from vomiting and/or diarrhoea.
- Septicaemia may be the cause of the symptoms.
- Pneumonia or other respiratory infections may cause abdominal pain and vomiting.
- Effortless tachypnoea may indicate a metabolic acidosis.

The pulse rate and pulse volume may identify a rapid thready pulse indicating poor perfusion. Capillary refill is often thought to be a poor indicator of circulatory status, as the peripheral perfusion (hands and feet) may be affected by environmental temperature. The comparison of central (anterior chest) and peripheral capillary refill, with other data like heart rate, pulse volume and consciousness state, allows an assessment of the adequacy of the circulation. The presence of shock indicates inadequate tissue perfusion. It is present when there is a rapid, thready pulse, delayed capillary refill, especially if central (≥3 seconds), and abnormal neurological status including agitation, lethargy, or coma. The diagnosis of shock is not reliant upon the presence of hypotension, particularly in children. Delay in identifying shock until the child is hypotensive risks severe compromise and potential progress to cardiac arrest.

Septicaemia, with or without meningitis, may cause fever, vomiting and diarrhoea. Some bacterial pathogens that cause gastroenteritis may also cause septicaemia, such as *Salmonella* and *Shigella*.

Abdominal examination

The abdomen is examined for distension, tenderness and guarding. Take note of the child’s resting posture and how they move spontaneously or on request, both in the bed or walking. A gentle, unrushed examination provides more information.

Presence of guarding and rebound can be elicited with gentle flexion/relaxation of the fingers of the examining hand (at the metacarpophalangeal joint). Evaluate for organomegaly – liver, spleen and kidneys. Are there any masses? Examine for anal fissures and other perianal abnormality that may indicate inflammatory bowel disease.

Neurological examination

The child may have impaired consciousness either due to systemic illness, shock or a primary neurological problem. The unwell infant may have meningitis.

Temperature

Always measure the temperature of the child. If the child's circulation is compromised or the child is particularly unwell, then consider continually monitoring the temperature with a rectal probe. The child with overwhelming sepsis may not mount a fever and, indeed, may be hypothermic.

Differential diagnoses

This list of pathogens causing diarrhoea is extensive. The differential diagnosis list varies depending upon the combination of symptoms: diarrhoea alone, or with vomiting and/or fever. The clinical examination findings may also refine the differential:

- Gastroenteritis viral, bacterial (Chapter 7.12):
    - Secondary lactase deficiency
- Viral illness:
    - Gastritis/enteritis
    - Non-specific, generalised viral illnesses
- Antibiotic-associated diarrhoea
- Constipation with overflow
- Septicaemia
- Meningitis/encephalitis
- Appendicitis
- Intussusception
- Malabsorption including cows' milk protein enteropathy, lactose intolerance

- Colitis:
    - Inflammatory bowel disease (Crohn's disease, ulcerative colitis)
    - Pseudomembranous colitis due to *Clostridium difficile*
- Immunodeficiency
- Endocrine disorders (adrenal insufficiency, hyperthyroidism, hypoparathryoidism).

In infants, non-IgE mediated cow's milk protein allergy may present with eczema, gastro-oesophageal reflux and/or diarrhoea, which may have associated mucous and blood.

Investigations

Remember, only order tests that aid the decision making and management of the patient. Reference ranges are frequently based on the standard deviation in a population, so a result outside the range may not necessarily indicate abnormality or pathology. This may inadvertently lead the clinician to order more unnecessary tests.

Not every child who is medically assessed requires investigations. The tests required depend on the possible differential diagnoses and the severity of illness (Box 7.8.1). Don't forget simple bedside tests like urine analysis (UA), blood glucose or stool reducing substances, all of which can be completed at the bedside. This information can be invaluable in assessing the child with diarrhoea. In this context, check the specific gravity and for ketones. Glycosuria may indicate diabetic ketoacidosis. If the UA is positive for nitrites or leucocytes, firstly verify that the specimen was appropriately collected as a contaminated specimen may cause this result, probably even more so than a urinary tract infection in the infant with diarrhoea If a urine specimen is to be cultured, it should be collected attempting to minimise contamination, even if re-collection is required.

Box 7.8.1 Tests that may be useful in the child with vomiting or diarrhoea

Common tests:

- Urine analysis

- Urine culture
- Stool:
    - Microscopy, culture
    - Rotavirus antigen
    - *Giardia* (often tested by RIA, avoiding the need for multiple samples for ova and parasites)
    - *Clostridium difficile* toxin detection (difficult to interpret in young children)
    - Stool reducing substances (carbohydrate malabsorption)
- Biochemistry – electrolytes, urea/creatinine, blood glucose
- Full blood count

Less common tests:

- Liver function tests
- Blood culture

RIA, radioimmunoassay.

If the infant has persistent diarrhoea while feeding on a lactose containing formula or breast milk, then testing fluid stool for **reducing substances** using a Clinitest® tablet, with stool instead of urine, can quickly identify lactose intolerance. Remember for the test to be positive in lactose intolerance, there must be lactose in the diet at the time of the test. If access to Clinitest® tablets is problematic, then stool can be sent to the laboratory for faecal reducing substances, sometimes referred to as faecal sugars or faecal carbohydrates.

The microscopy of the diarrhoeal stool may be important. The presence of stool white cells, without a viral or bacterial pathogen, in the young infant, may indicate cows' milk protein intolerance. Viral testing of stool and bacterial culture or PCR should be selective and are not routinely required for all children. The role of faecal multiplex testing is unclear, as their very high sensitivity will often demonstrate 'positive' results of questionable significance.

Coeliac disease is possible in a child with symptoms of persistent diarrhoea, decreased appetite, abdominal pain and/or bloating, poor weight gain or weight loss. If the child is on solid feeds containing gluten, coeliac serology should be sent in the appropriate clinical context.

Further tests in the setting of chronic diarrhoea should occur in consultation

with the paediatrician who will be following up the child.

Management

Intravenous fluids

Intravenous fluid boluses of 0.9% sodium chloride should be administered to a child shocked from diarrhoea. Once resuscitated, further fluid management should consist of isotonic fluids with glucose added (e.g. 0.9% sodium chloride + 5% dextrose) aiming to replace deficits and keep up with ongoing losses, with potassium replacement guided by electrolyte measurement. Chapter 10.6 provides a detailed discussion of fluid and electrolyte management.

Nasogastric rehydration is commonly used in dehydrated infants with gastroenteritis (see Chapter 7.12).

Dietary modification

Some infants may present with symptoms consistent with non-IgE mediated cow's milk protein allergy. If suspected, there are no specific tests which are helpful. Rather, exclusion of cow's milk protein and soya protein (due to the risk of cross-reactivity) from the baby's diet is recommended. Breast-feeding infants can continue to breastfeed with their mothers excluding cow's milk and soy with assistance from a dietician and under GP supervision. Formula-fed infants should be changed initially to an extensively hydrolysed formula.

With elimination of cow's milk protein, symptoms should resolve within 2–4 weeks. At this time, the diagnosis can be confirmed with brief reintroduction of cow's milk into the diet. If symptoms recur, then the elimination diet should continue until 1 year of age.

Antibiotics

Any specific management such as the administration of antibiotics depends on the provisional diagnosis, and the severity of illness. Most acute diarrhoeal illnesses are viral and self-limiting and therefore do not require antibiotics. Most bacterial causes of diarrhoea will also clear, and do not necessarily require antibiotics. Those which require specific antibiotics often require stool culture to determine the likely pathogen prior to initiating treatment.

Consultation

Consultation with more experienced staff, either emergency physicians or paediatricians should be encouraged, especially for complex or severely ill infants or children. Assistance may be required for clinical evaluation and technical procedures in critically ill children.

In children with more chronic presentations, discussion with a paediatrician is recommended, particularly if considering diagnoses unfamiliar to emergency practitioners (such as coeliac disease, malabsorption, or cow's milk protein enteropathy), or prior to recommending a change in diet. Infectious diseases consultation may be required particularly if considering antibiotics for gastroenteritis.

Conclusions

Differential diagnostic possibilities

For each patient the differential diagnoses vary depending on the constellation of symptoms (fever, vomiting, diarrhoea or others; acute vs. chronic). Keep an open mind and always re-evaluate clinical data. Don't be distracted by a child's previous diagnoses or labels.

General approach

'ABC fluids in and out' is a useful tool in taking a history. The younger the infant, the more challenging the evaluation, and the less reliable the clinical examination. If in doubt, seek consultation from a colleague. The telephone is a useful and powerful tool for clinicians and the value of consultation should not be underestimated.

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7.9

Management of acute hepatitis in children presenting to the emergency department

Sheena Kaul, Franz E. Babl, and Rupert Hinds

ESSENTIALS

1 Mild hepatitis is common, self-limiting, and is usually secondary to a generalised viral syndrome. Most children can be followed as outpatients but may require admission for supportive care depending on severity of illness.

2 Acute hepatitis due to the various hepatotropic viruses is clinically indistinguishable.

3 Severe hepatitis, especially with synthetic dysfunction including prolonged international normalised ratio (INR), should be urgently referred to paediatric gastroenterology.

4 Chronic liver disease may present in a manner resembling an acute hepatitis (autoimmune hepatitis, Wilson's disease). Early referral to paediatric gastroenterology is essential if this is suspected.

Introduction

Hepatitis, defined as inflammation of the liver, is characterised by a wide variety of clinical and histologic manifestations, and ranges in severity from mild and self-limiting to fulminant hepatic failure. Jaundice implies a yellow discolouration of skin, sclera and mucous membranes due to

hyperbilirubinaemia which may or may not be caused by hepatitis. In children, the majority of hepatitis cases are related to a viral infection.

Aetiology

- Infection:
    - Hepatotropic viruses, e.g. hepatitis A, B, C, E
    - Non-hepatotropic viruses, e.g. Epstein–Barr virus (EBV), cytomegalovirus (CMV), herpes simplex virus (HSV), enteroviruses, adenovirus, parvovirus B19
    - Bacterial infections with hepatic involvement, e.g. leptospirosis, brucellosis, Q fever, cat-scratch disease, or associated with septicaemia
- Drug- and toxin-related liver injury
- Chronic liver disease presenting ‘acutely’, e.g. autoimmune hepatitis, Wilson’s disease.

History

Children with acute hepatitis generally present with non-specific symptoms of nausea, vomiting, low-grade fever, anorexia, and malaise. There may be more specific signs and symptoms including jaundice, icterus, localised hepatic pain and/or tenderness, hepatosplenomegaly, and pruritus. A history of acute drug or toxin ingestion (particularly paracetamol) or therapeutic use of potentially hepatotoxic agents should be elicited. The history should also explore the possibility of foreign or rural travel. Consideration should also be given to the possibility of acute presentations of chronic liver disease.

Examination

Although jaundice is the hallmark of hepatitis, many children will present anicteric. In addition to jaundice, physical examination in acute hepatitis commonly reveals tender hepatomegaly. Signs of chronic liver disease should be specifically looked for including clubbing, palmar erythema, spider naevi, bruising, scratch marks, ascites, prominent abdominal veins, oedema and signs of fat-soluble vitamin deficiency.

Investigations

Extensive laboratory tests for hepatitis may not be clinically useful in the emergency setting. Initial studies should include a full blood count with platelets, electrolytes, urea, creatinine, blood sugar level, total and fractionated serum bilirubin, aspartate aminotransferase (AST), alanine aminotransferase (ALT), gamma glutamyl transferase (GGT), alkaline phosphatase (ALP), albumin and prothrombin time (PT)/international normalised ratio (INR).

PT/INR is the most useful indicator of synthetic function.

If viral hepatitis is suspected, a hepatitis antibody/antigen panel should be done (Box 7.9.1). If no viral cause can be identified, metabolic and systemic causes of liver dysfunction should be investigated as clinically indicated. If chronic liver disease is suspected, this should be discussed with a paediatric gastroenterology unit.

Diagnostic imaging is not specific in most acute viral hepatitis. In cases of suspected structural liver disease, imaging would initially include ultrasonography.

Box 7.9.1 Investigations for acute hepatitis

Haematologic tests

Full blood count
INR or prothrombin time
Blood group and hold

Biochemical tests

Liver function tests (bilirubin fractions, AST, ALT, GGT, ALP, albumin)
Serum electrolytes
Serum calcium, magnesium
Blood sugar level
Acid–base status
Consider lactate and ammonia if concerns around the child's neurological status

Serologic tests

Hepatitis A/B/C serology, hepatitis E (if travel history)
CMV
EBV
Consider if indicated by other clinical features

Imaging

Abdominal ultrasound

Further investigations (on discussion with paediatric gastroenterology team) – particularly if the child has features of chronic liver disease

Immunologic tests

Autoantibodies:
ANA
anti-SMA
anti-LKM1
Serum immunoglobulins

Urine

24-h urinary copper

Others

Serum caeruloplasmin/copper

ALP, alkaline phosphatase; ALT, alanine aminotransferase; ANA, antinuclear antibodies; anti-LKM1, liver-kidney microsomal antibodies; anti-SMA, anti-smooth muscle antibodies; AST, aspartate aminotransferase; CMV, cytomegalovirus serology; EBV, Epstein–Barr virus; GGT, gamma glutamyl transferase; INR, international normalised ratio.

Box 7.9.1 highlights the important tests to consider when investigating the cause for acute hepatitis, but the age of the child and severity of the child's illness should be considered when requesting the tests.

Viral hepatitis

The agents identified to cause hepatitis as their primary disease manifestation include hepatitis A virus (HAV), hepatitis B virus (HBV), hepatitis D virus (HDV) (only in the presence of HBV), hepatitis C (HCV) and hepatitis E (HEV). HAV and HEV are not known to cause chronic disease whereas the others can develop chronic infection. Many other viruses can cause hepatitis as part of their clinical spectrum, such as EBV and CMV.

All viruses can cause fulminant hepatitis with acute liver failure. A child with evidence of synthetic dysfunction (prolonged prothrombin time, hypoglycaemia, hyperammonaemia, lactic acidosis) with or without positive viral serology should be urgently referred to paediatric gastroenterology.

Hepatitis A

HAV is a common viral illness worldwide, although the incidence in developed countries has diminished due to extended immunisation in targeted groups. Hepatitis A virus (HAV) is an RNA virus and the most common mode of transmission is by the faecal–oral route. The incubation period is 15–50 days. Hepatitis A is an acute self-limiting illness associated with fever, malaise, jaundice, anorexia and nausea. Children under 6 years of age are often asymptomatic.

Fulminant hepatitis is rare, but possible.

Hepatitis B

HBV infection is one of the most important causes of hepatitis worldwide and its prevalence varies markedly from country to country, but the incidence has reduced in Australia due to universal immunisation programmes and the implementation of blood-donor screening.

HBV is transmitted through contact with infected blood or body fluids. The incubation period is 45 to 160 days. HBV infection causes a wide spectrum of diseases ranging from asymptomatic seroconversion, clinical hepatitis with jaundice, and fulminant hepatitis.

Follow-up laboratory studies are important to assess the development of carrier state and chronic liver disease.

Hepatitis C

HCV is a small single-stranded RNA virus with multiple genotypes. HCV is primarily acquired by exposure to blood and blood products (screening of blood donors and blood products and inactivation procedures for HCV have virtually eliminated this risk), persons with high-risk sexual behaviour and in children vertical transmission. Incubation time is 2 weeks to 6 months.

Symptomatic acute hepatitis is very unusual in children. Most children with chronic HCV infection are asymptomatic, but chronic liver disease and cirrhosis can follow.

Hepatitis E

Hepatitis E virus (HEV) is an RNA virus. Transmission of HEV is by the faecal–oral route. HEV in Australia has been reported most commonly in returning travellers from endemic regions, such as Asia. HEV causes a self-limited acute illness with jaundice, malaise, anorexia, fever, abdominal pain and arthralgia.

Viral hepatitis due to other viruses

Many other viruses including EBV, CMV, parvovirus B19, varicella zoster, adenoviruses and enteroviruses, as well as yellow fever and dengue fever in tropical and subtropical areas, can also cause acute hepatitis in children. EBV and CMV are ubiquitous and most persons are infected by the time they reach adulthood. If tests for hepatitis A, B and C are negative, EBV and CMV are among the most likely agents to consider. Hepatic involvement with these viruses is usually only one component of a multisystem disease.

The majority of EBV infections in children are asymptomatic, and primary infection in adolescents results in acute infectious mononucleosis syndrome –'glandular fever'. Most cases present with asymptomatic or mild illness accompanied by a mild elevation of hepatic enzymes that generally returns to normal within 2–6 weeks.

Drug- and toxin-induced liver injury

The liver is the main site of drug metabolism and is particularly susceptible to injury from drugs and toxins. The clinical spectrum varies from asymptomatic biochemical abnormalities to fulminant liver failure. Clinicians need to consider

all drugs taken in the previous months including prescription medications, over the counter medications and complementary medicines. Treatment is mainly supportive, although N-acteylcysteine is an important therapy in paracetamol-induced liver toxicity.

Chronic liver disease presenting as acute hepatitis

Autoimmune hepatitis

Autoimmune hepatitis (AIH) is a chronic disease of uncertain aetiology characterised by hepatocellular inflammation. Progressive destruction of hepatocytes may occur with possible progression to cirrhosis. The clinical features of autoimmune hepatitis vary widely; for many children the disease has an insidious onset with non-specific symptoms such as abdominal pain, fatigue, anorexia and weight loss. However, about one-third of children present with symptoms of acute hepatitis. Blood test abnormalities at presentation commonly include elevated transaminases, raised immunoglobulins and positive autoantibodies including antinuclear antibodies (ANAs), smooth-muscle antibodies (SMAs), liver-kidney microsomal type 1 (LKM-1), anti–liver cytosol 1 (anti-LC1) antibodies, and antibodies to soluble liver antigen (SLAs).

Wilson’s disease

Wilson’s disease is an autosomal recessive disorder caused by a defect in biliary copper excretion. Excessive intrahepatic accumulation of copper causes liver cell damage followed by fibrosis and cirrhosis. Extrahepatic accumulation of copper in brain, cornea and kidney accounts for extrahepatic manifestations of neurological abnormalities (in particular, dystonia, dysarthria and cognitive decline), Kayser–Fleisher rings, and renal effects of nephrocalcinosis and renal tubular acidosis. Initial presentation of Wilson’s disease may resemble acute viral hepatitis with variable extrahepatic findings, including Coombs negative haemolysis.

Both of these conditions may present with features of acute liver failure and if either is suspected children should be urgently referred to paediatric gastroenterology.

Other causes of chronic liver disease presenting in childhood

Many rare metabolic conditions can present with liver disease in childhood, particularly infancy, but it is unusual for them to present in a manner resembling acute hepatitis. More commonly, they present with metabolic acidosis, encephalopathy and/or hypoglycaemia. These conditions are discussed in more detail in Chapter 10.1.

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7.10

Intussusception

Kim Lian Ong, and Ramesh Nataraja

ESSENTIALS

1 A high index of suspicion is needed to make an early diagnosis, especially with the presence of inconsolable crying. The morbidity and potential mortality is increased by a delayed diagnosis.

2 The majority of cases are idiopathic.

3 Intussusception is the most common cause of bowel obstruction in children between 3 months and 3 years of age.

4 Paroxysmal inconsolable colicky abdominal pain/distress is the most common symptom. The classical triad of abdominal pain, vomiting and redcurrant jelly stools occurs in less than 50% of patients.

5 Profound lethargy may be the presenting feature in 10% of cases.

6 Bilious vomiting and redcurrant stools are late signs.

Introduction

Intussusception is a common cause of bowel obstruction in children less than 2 years old. It occurs when a section of bowel (usually the small intestine) telescopes into the lumen of a more distal portion of bowel. The intussuscepted segment (intussusceptum), is carried distally by peristalsis while the mesentery and vessels are trapped within the engulfing segment (intussuscipiens). The resulting venous congestion causes oedema that may then progress to ischaemia with arterial compromise and necrosis. This leads to the mucosa ‘sloughing off’ and the classic ‘redcurrant jelly’ stool. Intussusception occurs most commonly at

the terminal ileum when it is carried through the ileocaecal valve into the colon (ileocolic ~90%) and in some instances the telescoping small bowel may even reach the rectum.

Aetiology

Most cases of intussusceptions are *idiopathic* without any mass lesion acting as a lead point or an apex of the intussusceptum. The lead point is thought to be hypertrophied Peyer's patches in the ileal wall secondary to either adenovirus or rotavirus infection. This accounts for the seasonal nature of intussusception presentations in spring and winter. There is also a significant rise in the incidence of intussusceptions with a recent rotavirus vaccination. In *pathological intussusception*, the following may act as lead points:

1. Meckel's diverticulum
2. Intraluminal polyps (associated with familial adenomatous polyposis or Peutz Jeghers syndrome or other syndromes)
3. Lymphoma and leukaemia involving the bowel wall, although this is rare
4. Haemolytic–uraemic syndrome
5. Cystic fibrosis with inspissated bowel content
6. Foreign bodies
7. Henoch–Schönlein purpura (HSP) with intramural haemorrhage.

Epidemiology

Most children with intussusception are younger than 1 year of age with a peak incidence in infants between 5 and 10 months. Patients who are under 3 years of age are unlikely to have a pathological lead point. With recurrent intussusceptions outside of the normal age range, which resolve with non-operative management, patients should be investigated with USS or laparoscopy.

The estimated incidence is 1 to 4 per 1000 live births. There is an overall male preponderance, with a male-to-female ratio of approximately 3 to 1. Mortality with appropriate medical intervention is rare.

Clinical

Clinically, the **classical symptom triad** of abdominal pain, vomiting, bloody (redcurrant jelly) stool described in patients with intussusception is present in

less than one-half of patients with the disease.[1,2] This may lead to delays in diagnosis, and in these patients intestinal obstruction is often the primary presenting symptom.

The patient usually presents acutely with no previous medical history of note. The presentation is one of sudden onset of intermittent colicky abdominal pain, manifesting as episodic bouts of crying. During these episodes the infant is inconsolable, even by the mother, and this should raise a high index of suspicion. The episodes typically last for 20–30 minutes with respite for a similar time period. One possible description by the caregivers is the drawing up of the legs to the child's abdomen and then kicking the legs in the air. Often the child will appear **pale** due to increased vagal tone caused by the telescoping bowel. Between the episodes, the child may be flat, lethargic or fall asleep exhausted, whereas some children will resume normal activity.

There is associated poor feeding, vomiting, and there may be the passage of loose or watery stools. The symptoms are often vague as there is usually a preceding viral illness (gastrointestinal or respiratory) leading to the hypertrophy of Peyer's patches. Diarrhoea may be present as an existing symptom. The mixture of mucus and blood described as 'redcurrant jelly' is a **late sign**. Initially the vomiting is non-bilious but it becomes green/bilious once intestinal obstruction occurs.

Most children appear pale, with variable degrees of clinical dehydration. Abdominal examination may not reveal any clinical signs, although tenderness, guarding, distension or a right hypochondrium/mid-abdominal sausage-shaped mass may be palpated. The nappy should be checked for any blood or stools containing altered blood. Rarely the intussusception may prolapse rectally. The presence of peritonitis, with the physical signs of percussion tenderness and involuntary guarding, or pyrexia and leukocytosis indicate potential intestinal transmural gangrene and infarction.

There is a small subset of 'encephalopathic' intussusceptions that present very rarely without gastrointestinal symptoms ('painless presentation'). These children will present with lethargy, sweating and pallor which may be episodic.

Morbidity is increased by a diagnostic delay and is secondary to bowel wall necrosis and hence potential intestinal perforation. The associated intestinal obstruction with persistent vomiting results in fluid depletion and electrolyte imbalance. Established intussusceptions are less likely to respond to non-operative treatment as the area of necrosis prevents reduction by air enema or hydrostatic reduction. Pathological intussusceptions may resolve with non-

operative intervention therefore recurrence should trigger further investigation for an underlying pathological lead point.

Investigations

All children should be resuscitated and be haemodynamically stable prior to any imaging. It is therefore essential to obtain intravenous access and administer intravenous fluids. Intravenous antibiotics should only be given once the diagnosis is confirmed and an enema reduction is planned.

Abdominal X-ray

A plain abdominal radiograph may be performed, although this is not routine practice. Possible findings are:

1. an absence of air in the right upper quadrant
2. a right-sided soft tissue shadow giving an impression of an intracolonic mass or a crescent sign
3. absence of intestinal air or fluid levels in the caecum
4. distal intestinal obstruction with dilated intestinal loops
5. intra-peritoneal free air (Rigler's sign, football sign or air under the diaphragm)
6. unremarkable gas intestinal pattern.

The presence of stool and air fluid levels in the caecum makes the diagnosis of intussusception less likely.[3,4] The accuracy of plain radiography in diagnosis or exclusion of intussusception ranges from 40% to 90%. The presence of any free air indicates intestinal perforation and warrants urgent surgical intervention.

Historically, the diagnosis of intussusception is made by the use of an enema, either using air or barium. This has the advantage of also being potentially therapeutic. Barium has traditionally been used but perforation can result in barium and faecal peritonitis.[5–7] The availability of near-isotonic water media and the use of air as a contrast medium[8,9] have changed this traditional therapeutic approach. The advantages of air reduction are its rapidity and safety compared with barium and decreased radiation dosage. In the case of perforation during the procedure, air enema has been shown to result in a smaller tear than hydrostatic enema with markedly less spillage of faeces.[10] In addition, air has no

deleterious consequences within the peritoneal cavity.[11,12]

Findings on contrast examination include the classical ‘coiled spring sign’ (contrast tracking around the lumen of the oedematous intestine) and the ‘meniscus sign’ (rounded apex of the intussusceptum protruding into the column of contrast material).[13]

Ultrasound

This is the gold standard for diagnostic imaging, as it is highly sensitive and specific for the diagnosis of intussusception. It also involves no radiation exposure. Some studies report a sensitivity of nearly 100%, even in relatively inexperienced hands.[14–16]

The classical ultrasonographic signs include; the ‘target sign’ on the transverse plane (circular mass with a ‘donut’ appearance) and ‘pseudokidney sign’, on the longitudinal plane (reniform appearance). Both have a central hyperechoic area representing mesenteric fat pulled with the vessels and lymph nodes into the intussuscipiens, while the hypoechoic outer layer is the oedematous wall of the intussuscepted intestinal head.[17]

Management

The treatment in the emergency department is to initially provide resuscitation according to the APLS guidelines, establish intravenous access and consult with a paediatric surgeon. Fluid therapy should include intravenous resuscitation (20 mL kg^{-1} 0.9% saline bolus) and subsequent maintenance as required. The stomach should be decompressed by use of a nasogastric tube in the presence of vomiting or suspected intestinal obstruction. If there are clinical signs of peritonitis or sepsis, broad spectrum antibiotics should be administered intravenously and preparation for emergency operative intervention should occur.

Non-operative reduction by air enema is now generally accepted as the optimal initial modality of treatment. Absolute contraindications include peritonitis, perforation or profound shock.[18]

Operative intervention is traditionally performed with a laparotomy via a right upper or lower quadrant incision. With this approach the intussusception is carefully milked back from distal to proximal avoiding inadvertent trauma to the oedmatous friable intestines. Any areas of necrosis are then resected with an

end-to-end anastomosis, although occasionally a stoma has to be fashioned. The laparoscopic approach has gained popularity in many centres with comparable results in recent times.

Outcome

Air enema reduction rates are between 80% and 90%, with a 1% perforation rate.[18–20,26–28] Factors that are associated with either a lower successful reduction or higher perforation rate are:[20–25]

a. Patient's age: younger than 3 months or older than 5 years
b. Long duration of symptoms, especially if more than 48 hours
c. Passage of rectal blood
d. Significant dehydration
e. Small bowel obstruction
f. Presence of dissection sign on contrast study.

Surgical reduction is indicated after failure of air enema reduction or if there is a contraindication to non-operative reduction. If there is only the absence of reflux of air into the small bowel, in a clinically well patient, then a second air enema may be attempted after 6 hours.

There is an immediate recurrence rate of 5%, with 33% of these occurring in the first 24 hours after reduction and the majority within 6 months. These patients need to be monitored after successful air enema reduction for 24 hours and investigated for any recurrence of symptoms.

The overall mortality rate of intussusception is less than 1%.[18] Mortality rates observed among children in industrialised countries are lower than those in developing countries.[1,28–33] Some of these deaths are preventable and may be related to reduced access to or delays in seeking health care, factors known to be associated with mortality in children with intussusception.[31–33] Therefore, early diagnosis and management play an important role in the reduction of mortality.

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7.11

Herniae

Andrew John Anderson Holland

ESSENTIALS

1 The key diagnostic steps are detailed history from the carer and a careful clinical examination.

2 Radiological investigations have little, if any, role in the management of inguinal herniae in children.

3 A hernia that is not reducible may be associated with ischaemia and an urgent surgical opinion should always be sought.

4 In children, unlike in adults, it is the gonad rather than the bowel structures that is most at risk from an inguinal hernia.

Introduction

A hernia is defined as the protrusion of a viscus or part of a viscus into an abnormal location. The most common herniae occurring in children are inguinal and umbilical – femoral herniae are rare. Various descriptive terms are applied to herniae in these locations, often incorrectly. This may lead to diagnostic confusion and inappropriate delays in referral.

Herniae are usually classified based on two characteristics: anatomical location and whether or not the hernia is reducible. An irreducible hernia may result in ischaemia of either the contents of the hernia or adjacent structures. Treatment of an irreducible hernia is thus a surgical emergency. The use of the terms incarceration (irreducible) and strangulation (arrest of circulation) should be avoided, as all irreducible herniae may be associated with ischaemia

(suggested by pain and/or marked tenderness with overlying erythema) and therefore require urgent treatment.

Types of herniae

Inguinal

The incidence of inguinal herniae in children has been reported to be between 0.8% and 4.4%,[1] rising to 18.9–30% of preterm infants.[2,3] Inguinal herniae are six times more common in boys and are more common in twins.[1,4] Around 1 in 10 inguinal hernias are non-reducible at presentation, although a careful history from the parents will often elucidate earlier signs and symptoms. In this group, over two-thirds are under 1 year of age.

In children, inguinal herniae are almost always indirect.[5] The hernia exits the peritoneal cavity via the internal inguinal ring to enter the inguinal canal, leaving the canal via the external inguinal ring. The sac is intimately related to the contents of the spermatic cord. It is compression of the testicular vessels by hernial contents that may render the testis ischaemic. In contrast, in females the risk of ischaemia to the ovary and adnexae usually occurs as a result of torsion of these structures within the hernial sac.

Rarely, direct inguinal herniae may occur, with some series reporting an incidence of up to 5%.[6] Typically these children have either had previous inguinal surgery, a connective tissue disorder or were delivered at less than 30 weeks' gestation. The clinical management and surgical approach remains similar to indirect inguinal herniae.

Inguinal herniae usually present as a swelling in the inguinal region first noticed by the carer when changing or bathing the child, especially if crying or straining.[7] The swelling may extend into the scrotum in boys or the labia majora in girls. Persistent tachycardia, overlying erythema and marked tenderness suggest an irreducible hernia complicated by ischaemia.[8] Occasionally an irreducible hernia may present with vomiting and abdominal distension as a result of intestinal obstruction.

A hernia can be differentiated from a hydrocele as the latter transilluminates and does not usually extend into the inguinal region. In neonates, these differences may be less obvious and a hernia may appear to transilluminate. Encysted hydroceles of the cord and, rarely, testicular torsion may also cause diagnostic confusion. Where there is any doubt regarding the diagnosis, a

paediatric surgeon should be consulted.

Femoral

Femoral herniae are rare in children, accounting for between 0.4% and 1.1% of all groin herniae.[1,9] They are often not diagnosed prior to surgery and are one cause of recurrent ‘inguinal’ herniae in children. Typically most present between 4 and 10 years of age and there is an equal sex incidence. Clinically the hernia presents as a swelling that is inferior and lateral to the pubic tubercle. As in adults, femoral herniae are associated with a high incidence of complications, so prompt surgical intervention is required.

Umbilical and paraumbilical herniae

A true umbilical hernia occurs through the umbilical ring as opposed to a paraumbilical hernia that results from a defect *adjacent* to the umbilical ring. In most cases careful clinical examination can distinguish between them.

Umbilical herniae are very common, occurring in up to 18.5% of infants under 6 months of age.[5] They are more common in Afro-Caribbean children and in premature infants, when the incidence rises to between 41.6% and 75%.[10,11] The vast majority of umbilical defects appear to close spontaneously with increasing age, largely irrespective of the size of the defect. The risk of an umbilical hernia becoming non-reducible remains very low. Surgical repair is only indicated for those herniae that persist beyond 3 to 5 years of age, or in the very few that present with symptoms.[12]

Paraumbilical defects do not close with increasing age and should be referred for elective repair on diagnosis.

Epigastric herniae

These herniae occur as a result of a defect, often only a few millimetres in size, in the linea alba between the umbilicus and the xiphisternum. The child presents with a small swelling in this region that may be associated with pain, usually due to entrapment of extraperitoneal fat. Once it is diagnosed elective surgical repair is required.

Complications

Complications of an irreducible hernia are more likely to occur if the child presents late or there is diagnostic delay. The most common adverse sequelae include gonadal ischaemia and bowel obstruction. Enteric ischaemia, perhaps leading to perforation, remains rare.

Treatment

All inguinal hernias require surgery due to the risk of bowel ischaemia and compression of adjacent gonadal structures. If the hernia is reducible, the patient should be discussed with a paediatric surgeon regarding the timing of follow-up. Neonates and infants should be assessed within 1 week of presentation and scheduled for early elective surgery because of the high risk of the hernia becoming irreducible in this age group. Carers should be advised to re-present urgently if signs and symptoms of an irreducible hernia develop.

Indirect inguinal hernias have traditionally been treated by a herniotomy, with removal of the hernial sac following its ligation at the internal ring. This procedure has a high success rate with a low incidence of important complications such as injury to the vas and subsequent testicular atrophy.[6] Laparoscopic correction involves suture ligation of the hernial sac from within the abdominal cavity and appears to have a slightly higher recurrence rate than open surgery.[13] Direct hernias are generally repaired primarily without the use of mesh.[6]

If the hernia does not spontaneously reduce, a paediatric surgeon should be contacted urgently. In the absence of signs or symptoms suggestive of ischaemia, manual reduction by an experienced clinician may be attempted. The child should be given appropriate analgesia. A two-handed technique is required: one hand from above disengages the hernia from the external ring and the other from below reduces the hernia by gentle direct pressure. If there are signs suggestive of ischaemia, emergency surgery is required.

Controversies

1 The role of routine contralateral inguinal exploration in children with a unilateral inguinal hernia remains controversial. A recent systematic review suggested that, as the overall risk of developing a metachronous hernia in childhood was 7.2%, routine contralateral exploration was not warranted.[14]

2 The use of intraoperative laparoscopy for the assessment of the presence of a contralateral inguinal hernia has been advocated but even if positive may not necessarily correlate with subsequent development of a symptomatic hernia on that side.

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7.12

Gastroenteritis

Susan Phin

ESSENTIALS

1 Most children with gastroenteritis and mild-to-moderate dehydration can be successfully rehydrated with oral rehydration solutions either by mouth or nasogastric tube.

2 In dehydrated children, oral rehydration solutions (ORS) should be used for oral rehydration, not fruit juices, soft drinks or sports drinks.

3 It is important to use appropriate intravenous fluids for bolus or maintenance phases of rehydration.

4 Careful instruction given to parents on how to give oral fluids appropriately is vital.

5 Antidiarrhoeal and most antiemetic medications should not be used in children.

6 Early refeeding with age-appropriate foods should be encouraged.

7 Timely reassessment is vital and, if the child does not progress as anticipated, other diagnoses and complications should be considered.

8 Other diagnoses should be considered especially in the presence of:

- an infant less than 6 months of age
- high-grade fever
- bilious vomiting
- significant abdominal pain
- no diarrhoea
- blood in vomitus or stool

- drowsiness/reduced level of consciousness.

Introduction

Acute gastroenteritis is an inflammation of the gastrointestinal tract. It is one of the commonest reasons for children to present to an emergency department (ED). Most children under 5 years of age have experienced an episode of gastroenteritis and most can be successfully managed without admission to hospital. Worldwide, however, gastroenteritis still remains a significant cause of morbidity and mortality.[1]

Aetiology

Gastroenteritis is caused by a wide range of pathogens including viruses, bacteria and parasites (as shown in Table 7.12.1). In developed countries, the majority of episodes are due to viruses, with rotavirus being by far the most common pathogen. The most common bacterial causes are *Salmonella* and *Campylobacter. Shigella*, *Yersinia* and *Escherichia coli* are less common, while *Vibrio cholerae* is seen in developing countries. Parasites such as *Giardia* and *Cryptosporidium* are sometimes the infective agent.

In general a bacterium is more likely to be the causative agent if:

1. there is blood or mucus in the stool
2. there is significant abdominal pain
3. there is a high-grade fever.

These infective agents cause gastroenteritis by various mechanisms. Some directly invade the bowel wall, e.g. *Salmonella* or *Shigella,* some produce toxins prior to ingestion, e.g. *Staphylococcus aureus*, and some multiply and produce toxins within the gastrointestinal tract, e.g. *Shigella.*

In an uncomplicated acute bout of gastroenteritis, determining the causative pathogen is usually unnecessary, as this usually does not alter the management.

History

Most children with acute gastroenteritis present with a history of diarrhoea and

vomiting. Abdominal pain and fever are sometimes accompanying symptoms. It is important to ask for specific details about these symptoms, to increase the certainty of the diagnosis of gastroenteritis and also to help assess the risk of dehydration in the child.

Diarrhoea refers to loose or liquid stools. The frequency, volume (small to voluminous) and consistency (semisolid to watery) and the presence of blood or mucus are all important. Similarly, the frequency and nature (bilious or non-bilious) of the vomiting are also important. The words ‘my child is vomiting everything he tries to drink’ can be used by parents to describe a child who has had either two or 20 vomits in the last 24 hours. It is important, therefore, to be specific in questioning.

The abdominal pain that can be associated with gastroenteritis is crampy in nature and commonly more pronounced in bacterial gastroenteritis. It is not accompanied by focal or significant findings on abdominal examination. If fever is a symptom, the height of the fever can be helpful. Although children with rotaviral gastroenteritis are often febrile, it is not usually high-grade. Such fevers are more likely in bacterial gastroenteritis or another diagnoses altogether.

Specific details about the amount of fluid tolerated by the child are vital in assessing the risk of dehydration. Parents are usually able to estimate whether, over a 24-hour period, the child has taken one-quarter, one-half or three-quarters of normal intake. Intake that is consistently less than half of normal is of concern.

Specific questions about urine output (i.e. heaviness and frequency of wet nappies) are also important. The production of fewer than four wet nappies in 24 hours is of concern when assessing hydration status.

Questions about the level of alertness and activity of the child are also important. Lethargy may simply be an indication of significant dehydration in a child. However, more severe lethargy and drowsiness (particularly if out of proportion to that expected for the dehydration) may indicate another more serious diagnosis as the cause of the child’s symptoms, such as enteroviral meningitis.

Table 7.12.1

Common causes of acute gastroenteritis

Viruses	Bacteria	Parasites
Rotavirus	*Salmonella*	*Cryptosporidium*

Norwalk virus	*Campylobacter jejuni*	*Giardia lamblia*
Enteric adenovirus	*Shigella*	*Entamoeba histolytica*
	Escherichia coli	
	Yersinia enterocolitica	
	Vibrio cholerae	

Table 7.12.2

Standard assessment scale for severity of dehydration

Variable	Mild	Moderate	Severe
Thirst	+	++	+++
Dry mucous membranes	+	++	+++
Reduced urine output	+	++	+++
Lethargy	–	+	+++
Sunken eyes	–	+	++
Reduced skin turgor	–	+	++
Tachycardia	–	+	++
Poor perfusion	–	+	++
Tachypnoea	–	–	+
Hypotension	–	–	+

Examination

The aim of the clinical examination is to exclude signs of an alternative cause of the symptoms, other than gastroenteritis (see Chapters 7.2 and 7.8) and to assess the degree of dehydration.

Occasionally pathogens causing gastroenteritis can cause extraintestinal disease. This can occur particularly with *Shigella* and *Salmonella*. *Shigella* can cause central nervous system irritation, which can manifest as encephalopathy or seizures. This may occur prior to the onset of diarrhoea. *Salmonella can* cause a bacteraemia, which can lead to focal infections including osteomyelitis and meningitis. The infant under 6 months of age is particularly at risk.

Assessment of dehydration

The accurate assessment of dehydration is difficult. Studies have shown that medical personnel tend to overestimate the degree of dehydration.[2] The gold standard in assessment of dehydration is a loss of weight compared with a recent pre-illness weight. For example, a 1-year-old weighing 10 kg a week ago who presents with gastroenteritis for 3 days and now weighs 9.5 kg, is approximately 5% dehydrated. However, a recent weight is rarely available in the ED. Therefore, tables such as that shown in Table 7.12.2 use a combination of

clinical symptoms and signs to estimate the degree of dehydration.

It is important to remember that some of the signs and symptoms of dehydration can be affected by other factors. Mouth breathers have dry mucous membranes. Watery diarrhoea can make it difficult to assess if the child has a wet nappy from urine output. Crying can cause a sunken fontanelle to appear full. Tachycardia may be caused by a crying, anxious or febrile child; therefore it is the context and trend of the pulse that is important.

Various studies have attempted to validate combinations of these signs and symptoms with varying degrees of standardisation and scientific validity.[2–5] Difficulties arise as some of the signs are subjective and gold standards differ.

Gorelick et al.[5] examined the 10 clinical signs shown in Box 7.12.1. Using a gold standard of serial weight gain, they found that <3 signs correlated with <5% dehydration, 3–6 signs correlated with 5–9% dehydration and >7 signs correlated with >10% dehydration.

Box 7.12.1 Ten clinical signs of dehydration

1. Decreased skin elasticity
2. Capillary refill >2 seconds
3. General appearance (ill-appearing, irritable, apathetic)
4. Absent tears
5. Abnormal respirations
6. Dry mucous membranes
7. Sunken eyes
8. Abnormal radial pulse
9. Tachycardia (HR >150)
10. Decreased urine output

Source: Gorelick MH, Shaw KN, Murphy KO. Validity and reliability of clinical signs in the diagnosis of dehydration in children. *Paediatrics* 1997;**99**(5):E6.

Laboratory investigations in the assessment of dehydration

There are few data to support the usefulness of laboratory tests in the assessment of dehydration due to gastroenteritis. Dehydration is thought to typically cause a metabolic acidosis. It is true that vomiting can cause a metabolic acidosis by

several mechanisms including volume depletion, lactic acidosis and starvation ketosis. However, isolated vomiting can also cause a metabolic alkalosis through loss of gastric acid. In addition, isolated diarrhoea can cause a metabolic acidosis through loss of bicarbonate in the stool, but the child who can increase oral intake to keep pace with the diarrhoeal losses may not actually be dehydrated. Despite these confounding factors, it has been shown that serum bicarbonate is significantly lower in children with moderate or severe dehydration (mean 14.5 mEq L^{-1} and 10.3 mEq L^{-1}) than in children with mild dehydration (mean 18.9 mEq L^{-1}).[3] Raised serum urea levels have also been thought to reflect dehydration, but with even less evidence.

From a practical point of view, it is not vital to put a specific percentage on the degree of dehydration, particularly when the accuracy of such a specific percentage is questionable. What is important is to allocate the child into a broad category of dehydration, for example, mild, moderate or severe, which determines what treatment should be commenced, and then to reassess the child and the degree of dehydration within a specific timeframe.

Differential diagnosis

Many other conditions can masquerade as gastroenteritis by presenting with a combination of vomiting, diarrhoea, fever, or abdominal pain. These include:

- other infections, e.g. urinary tract infection, meningitis
- surgical diseases, e.g. intussusception, bowel obstruction, appendicitis
- raised intracranial pressure, e.g. blocked ventriculoperitoneal shunt
- metabolic diseases, e.g. diabetic ketoacidosis, inborn errors of metabolism.

Features of concern to suggest the possibility of another cause include:

- age less than 6 months
- high-grade fever
- bilious vomiting
- significant abdominal pain
- no diarrhoea
- blood in vomitus or stool
- drowsiness/reduced level of consciousness.

See Chapter 7.2 (vomiting) and Chapter 7.8 (diarrhoea), for more details on differential diagnosis.

Investigations

In uncomplicated acute gastroenteritis, it is usually not necessary to perform any investigations, as they will not alter management. Indications to consider performing investigations include:

- uncertainty about diagnosis
- severe dehydration
- the child's clinical course not progressing as anticipated
- commencement of intravenous rehydration.

Stool cultures do not change management in simple gastroenteritis; therefore they are not usually warranted. If bacterial gastroenteritis is suspected a stool sample may be performed, although this will not change management in the majority of cases. A stool sample may be taken for public-health reasons, or to reassure the doctor that there is not another cause for the blood and mucus in the stool.

Urine culture is usually unnecessary when the diagnosis of gastroenteritis is clear. However, if there is little diarrhoea, particularly if the child is under 6 months of age, it may be necessary to exclude a urinary tract infection. It is important that a *clean* specimen is collected to avoid contamination by stool contents.

Electrolytes, urea and creatinine are usually unnecessary in children with gastroenteritis who are mild to moderately dehydrated and who are to receive oral fluids, as they will not usually alter management. If, however, the child does not improve as expected, or deteriorates despite rehydration, or the child was initially severely dehydrated, these tests, along with a blood glucose level and perhaps other investigations, will be required.

A full blood count and blood culture are unnecessary in acute gastroenteritis. However, if the diagnosis is uncertain it may be reasonable to perform these tests. In the younger infant, particularly the <3 months age group, if bacterial gastroenteritis is likely the possibility of systemic *Salmonella* infection should be considered and blood culture and intravenous antibiotics considered.

If significant abdominal pain is present and surgical pathology such as

intussusception or bowel obstruction seems more likely than simple gastroenteritis, other investigations such as abdominal X-rays, ultrasound or air enemas may be required.

Treatment

Mildly dehydrated

Children in developed countries presenting to EDs with gastroenteritis are often only mildly dehydrated or not dehydrated at all. Most can be managed as outpatients with oral fluids alone. Educating the parents on how to give fluids and observing them giving a trial of fluids to the child are important to empower the parents to provide ongoing oral rehydration at home.

The essential things to ensure are that:

- the parent knows the appropriate oral fluids to give
- the parent knows how, and is able, to give the oral fluids correctly to optimise success
- the parent will have the child reviewed by the local doctor within 48 hours
- the parent knows what worrying signs and symptoms to look for that would require representation.

If the above criteria cannot be satisfied, the child may need to be admitted. This is particularly so in the younger infant or those with profuse gastrointestinal losses, as they have a higher risk of becoming dehydrated. See ‘moderately dehydrated’ for further treatment options in those patients.

Appropriate oral fluids

Oral rehydration solutions (ORSs) are the appropriate fluids to rehydrate children with gastroenteritis. These are specifically designed fluids that contain the correct amounts of sodium, glucose and other electrolytes and are of the appropriate osmolality to maximise water absorption from the gut. They use the principle of glucose-facilitated sodium transport, whereby glucose enhances sodium and secondarily water transport across the mucosa of the upper intestine. The sodium and glucose concentrations and the osmolality are of vital importance.

The WHO ORS has a sodium concentration of 90 mmol L^{-1}. In developed countries with non-cholera diarrhoea, it is generally thought that 90 mmol L^{-1} is a little high, as non-cholera gastroenteritis does not result in the same sodium losses that are seen in cholera. Many different ORSs with varying sodium concentrations have been developed. It has been shown[6] that water absorption across the lumen of the human intestine is maximal using solutions with a sodium concentration of 60 mmol L^{-1} and this is the concentration recommended by the European Society of Paediatric Gastroenterology and Nutrition.[7] Studies have also shown that hypo-osmolar solutions are most effective at promoting water absorption.[8–10]

Studies have also examined rice-based ORSs and their effect on stool output and duration of diarrhoea when compared with glucose-based ORSs. Rice-based ORSs appear to have benefits in cholera diarrhoea but not in non-cholera diarrhoea.[11]

The composition of various ORSs and other fluids is shown in Tables 7.12.3 and 7.12.4. Fruit juices and soft drinks are inappropriate for dehydrated children because of the minimal sodium content and the excessive glucose content and hence excessive osmolality. Diluting these solutions will not address the grossly inadequate sodium content, nor will it result in an optimal glucose concentration or osmolality. Sports drinks are also inappropriate, with too low sodium levels and too high glucose levels and osmolalities.

There is some recent evidence[12] that children with mild gastroenteritis and minimal dehydration may have fewer treatment failures (such as need for intravenous rehydration) if they are given dilute apple juice/preferred fluids instead of an electrolyte maintenance solution.

Method of giving oral fluids

The important message is to give small amounts of fluid frequently, for example 0.5 mL kg^{-1} every 5 minutes. The fluid can be measured in a syringe and given to the child either by syringe, teaspoon or cup. The child is far more likely to tolerate these small amounts of fluid than a whole bottle at once. If the child tolerates this fluid the parent can gradually increase the volume and decrease the frequency of the fluid offered. Success can be optimised in the ED setting by giving the parents a documentation chart to fill in, which shows the fluid given and any vomits or diarrhoea or urine passed.

It is important to educate the parents that seeing a doctor will not cure the vomiting and diarrhoea. Small, frequent amounts of fluid will hopefully

minimise the vomiting, but will not reduce the diarrhoea. The aim is for the input to exceed the output enough to rehydrate and then maintain hydration. Parents will know if they are succeeding by observing their child for the following symptoms and signs.

Indications for re-presentation

Parents should be given some indication of symptoms and signs that will prompt re-presentation. These need to be relatively easy for a parent to detect. They will be related to two things:

Table 7.12.3

Composition of oral rehydration solutions

ORSs	Na^+ (mmol L^{-1})	Carbohydrate			Osmolality (mOsm L^{-1})
			(mmol L^{-1})	(%)	
WHO	90	G	111	2	331
Gastrolyte	60	G	90	2	240
Gastrolyte-R	60	RSS	6 (g L^{-1})	2.5	226
Hydralyte (solution or iceblock)	45	G	90	2.5	240

G, glucose; RSS, rice syrup solids.

Table 7.12.4

Composition of oral fluids

Oral fluids	Sodium (mmol L^{-1})	Carbohydrate (mmol L^{-1})	Osmolality (mOsm L^{-1})
Apple juice	3	690	730
Sports drinks	20	255	330
Soft drinks	2	700	750

1. The child is, or is at risk of becoming, more dehydrated:
    - Persistent vomiting despite small amounts of oral fluids frequently
    - No wet nappy for 8 hours
    - Increasing lethargy
2. Gastroenteritis may not be the correct diagnosis:
    - Bilious vomiting
    - Significant abdominal pain
    - Drowsiness.

Refeeding

- Breast-fed infants – breast-feeding should continue throughout the

episode of gastroenteritis, including the rehydration phase.

- Formula-fed infants – full-strength normal formula should be started as soon as the infant is rehydrated. This can be supplemented with an ORS for ongoing losses, for example offer 10 mL kg^{-1} after each diarrhoea. However, children with no dehydration and mild gastroenteritis are the least likely to take an ORS because of the salty taste. In this instance increased amounts of normal fluids in conjunction with age-appropriate feeding may be sufficient.
- Early refeeding – studies have shown that early refeeding does not worsen or prolong the duration of diarrhoea, nor increase vomiting or lactose intolerance, and leads to a significantly higher weight gain after rehydration.[13] Early reintroduction of age-appropriate foods is now recommended.

Moderately dehydrated

Some children who present to EDs with gastroenteritis are moderately dehydrated. These children can be rehydrated in several ways. Some are successfully rehydrated with oral fluids alone, as previously described. Some fail this and require additional intervention. Increasing numbers of hospitals in developed countries are using oral rehydration therapy (ORT) via continuous nasogastric infusion.[14,15] This is where an ORS is infused continuously down a nasogastric tube with a pump such as a Kangaroo pump. Nasogastric infusions should not be used when the child has an ileus or is comatose. Oral rehydration therapy has been the method of choice in developing countries since the 1970s. However, it has taken longer to become accepted in developed countries. This is despite numerous studies that show that it is as effective as intravenous rehydration but less expensive[14–22] and reduces lengths of hospital stay.[14]

Different regimens are used for continuous nasogastric rehydration. The European Society of Paediatric Gastroenterology and Nutrition recommends calculating the fluid deficit and replacing that over 4 hours.[23] The American Academy of Pediatrics recommends that mildly dehydrated children receive 50 mL kg^{-1} over 4 hours and moderately dehydrated children receive 100 mL kg^{-1} over 4 hours.[24] Other regimens recommend a fixed volume, for example 40 mL kg^{-1} over 4 hours for all mild to moderately dehydrated children followed by a reassessment and retrial of oral fluids.[25] This takes into account the tendency for medical officers to overestimate the degree of dehydration[2] and the desire to

avoid subsequent over-hydration. The important thing to remember is that, whatever regimen is used to rehydrate the child, fluid status should be regularly reassessed. An example of recommended nasogastric fluid rehydration rates can be found in Chapter 10.6.

Intravenous rehydration

Intravenous rehydration should be used to rehydrate children with gastroenteritis who fail nasogastric therapy or deteriorate.

The volume (mL) of replacement fluids is calculated using the formula: weight (kg) × dehydration (%) × 10. Maintenance fluids are then calculated, with one method being to give 100 mL kg^{-1} for the first 10 kg, 50 mL kg^{-1} for the second 10 kg and 20 mL kg^{-1} for each subsequent kilogram. The volumes for rehydration and maintenance are then added together and divided by 24 to calculate the hourly rate.

The recommended fluid used to rehydrate children with gastroenteritis has changed over recent years. It is now recommended that for children, excluding neonates, **0.9% (150 mmol L^{-1}) sodium chloride + 5% glucose +/- 20 mmol/L potassium chloride should be used**.[26,27] Studies have shown that children with gastroenteritis have inappropriately high levels of antidiuretic hormone and increased incidence and risk of hyponatraemia[28] and that low sodium solutions cause hyponatraemia while solutions with a sodium content of 130–154 mmol L^{-1} are protective.[29] As in nasogastric rehydration, it is important to regularly reassess the child's fluid status. If the child remains on intravenous fluids for >24 hours, it is important to recheck the electrolytes.

If the child is significantly hypernatraemic the rate of infusion needs to be reduced to rehydrate the child over 48–72 hours. The aim is to avoid a rapid fall in the serum sodium level as this can precipitate cerebral oedema. It is still reasonable to start with 0.9% (150 mmol L^{-1}) sodium chloride + 5% glucose +/- 20 mmol/L potassium chloride to prevent a rapid initial fall in serum sodium, but this may need to be changed depending on progress of the electrolytes, which need to be rechecked frequently. If the child is hyponatraemic, 0.9% (150 mmol L^{-1}) sodium chloride + 5% glucose +/- 20 mmol/L potassium chloride should be used. Senior paediatric advice should be sought if the serum sodium level is significantly abnormal.

Rapid intravenous rehydration

Interest in rapid intravenous rehydration for children with gastroenteritis has

emerged in recent years in developed countries. It has long been used in developing countries.[30,31] There have been a number of studies in developed countries over the last several years that have looked at this issue[25,29,32–34] and shown that this seems to be a safe and effective way to rehydrate children with gastroenteritis who require intravenous therapy. An example of this type of regimen is giving 0.9% (150 mmol L^{-1}) sodium chloride + 5% glucose at 10 mL kg^{-1} hr^{-1} for 4 hours.[35] It is important that lower sodium-containing fluids and potassium-containing fluids are **not** used. Rapid intravenous rehydration is **not** to be used if the child is hypernatraemic or hyponatraemic.

Severely dehydrated

Although this is a relatively rare occurrence in developed countries, it is vital to diagnose and institute immediate treatment, as this is a medical emergency. The child is usually shocked and needs immediate intravenous access and **resuscitation with a 20 mL kg^{-1} bolus of 0.9% sodium chloride (normal saline) or Hartmann's solution**. If the child remains shocked the bolus should be repeated. Senior advice should be sought about any child presenting in this condition. During insertion of the cannula, blood should be taken for electrolytes, urea, creatinine, glucose, full blood count and venous blood gas. Blood cultures can also be taken at the same time if indicated.

Glucose-containing fluids such as 0.45% (75 mmol L^{-1}) sodium chloride + 2.5% glucose or 0.225% (37.5 mmol L^{-1}) sodium chloride + 3.75% glucose *should never be used for resuscitation boluses.* Although these solutions are technically isotonic, the glucose is rapidly metabolised, so effectively it is as if a hypotonic solution has been given, with the resultant risk of rapid fluid shifts and cerebral oedema. It is important to differentiate these resuscitation boluses from rapid rehydration therapy. The resuscitation boluses are given over several minutes and *do not* contain glucose. The rapid rehydration fluids are given over several hours and *do* contain glucose.

After the initial resuscitation, replacement plus maintenance fluids are given as per above. It is important to reassess the child's hydration status regularly and look for any indication that another diagnosis is the cause of the child's condition. Electrolytes, urea and creatinine need to be regularly repeated while the child remains significantly unwell.

Other treatments

Antibiotics

Antibiotics are rarely indicated in gastroenteritis, and should not be prescribed without strong evidence of a specific pathogen. They are of proven benefit for treatment of only the following situations:

- *Shigella*:
    - Intravenous ampicillin in severe disease; oral ampicillin or trimethoprim-sulfamethoxazole in milder disease
    - Shortens illness duration, eradicates organism
- Invasive *Salmonella* infections – e.g. septicaemia, osteomyelitis, meningitis:
    - Intravenous cefotaxime or ceftriaxone
    - Antibiotics prolong excretion in gastroenteritis.
- Traveller's diarrhoea:
    - Azithromycin
- *Campylobacter* septicaemia:
    - Erythromycin or gentamicin.
    - Not indicated in uncomplicated gastroenteritis.

Anti-diarrhoeal and anti-emetic medications

Anti-diarrhoeal medications are *not indicated* in children with gastroenteritis. Most anti-emetic medications are also *not indicated*. There is little evidence of efficacy and a high incidence of side effects such as dystonic reactions and sedation in infants and young children.

There have been several studies over the last 20 years looking at the role of ondansetron in the treatment of children with acute gastroenteritis.[36–38] One of the more recent is a Cochrane review published in 2011[39] which concluded that oral ondansetron increases the proportion of children with acute gastroenteritis who cease vomiting, reduces the number requiring intravenous rehydration and reduces the immediate need for hospitalisation. There was no difference in adverse events, although there was an increase in diarrhoea in some studies. A single dose of oral ondansetron has been incorporated into the state guidelines for the management of children with acute gastroenteritis in WA,[40] Victoria[41] and NSW.[42]

Ondansetron is not recommended if the child weighs <8 kg or is <6 months of age. The recommended dose for ondansetron is:[41]

2 mg for children weighing 8–15 kg
4 mg for children weighing 15–30 kg
8 mg for children weighing >30 kg.

Disposition

In the past, many children were admitted to hospital with gastroenteritis when perhaps they did not need to be.[43] With the emergence of 'Short Stay Wards' or 'Emergency Observation Units', where patients can be admitted to a special area within the ED for a finite time of less than 24 hours, hospital inpatient admission rates for children with gastroenteritis have fallen.[24] In these units children can be rehydrated (orally or via rapid nasogastric or intravenous infusion) over a period of a few hours and then sent home after appropriate education and advice. This is provided that the medical officer is confident about the diagnosis of gastroenteritis and that the criteria set out in the 'mildly dehydrated' section are fulfilled. Care needs to be taken especially with young infants, as they become dehydrated more rapidly than older children and are more likely to have another diagnosis.

Paediatric staff in non-children's hospitals can potentially be utilised to help manage these patients in conjunction with ED staff. An admission to, and discharge from, these 'Short Stay Wards' causes much less disruption to the child and family and allows hospital resources to be used for other patients requiring inpatient admissions. Some children, however, still require an inpatient admission for successful management.

Prognosis

Gastroenteritis in children is generally a benign, self-limiting disease, with dehydration being the major potential complication. If this is recognised early and the child is rehydrated appropriately, the child should recover completely with no adverse sequelae.

Controversies and future directions

1 Interest has arisen in the use of probiotics such as *Lactobacillus* GG in the treatment of acute gastroenteritis. Early studies seem to suggest that it may shorten the diarrhoeal illness.[44] Further research is necessary.

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7.13

Constipation

Bruce Fasher, and Simon Craig

ESSENTIALS

1 It is important to understand the physiology and development of gut transit to recognise the normal (often regarded as abnormal).

2 Constipation is a common reason for children to present to emergency departments (EDs) with abdominal pain.

3 Management depends on the child's age and whether the problem is acute or chronic.

4 The patient's and parents' real concerns need to be acknowledged.

5 Issues to be addressed include:

- natural history
- the aims of treatment (empty the bowel and establish pharmacological rhythm and then wean to biological rhythm)
- the likely duration of treatment
- the anticipated setbacks
- the need for a coordinator clinician (normally unavailable in an ED).

6 Treat from the top, once at night and titrate the dose according to the response. Avoid treatment per rectum if at all possible, to draw attention away from any anal obsession to achieve bowel actions.

7 Be able to refer to a resource that is known to be interested in managing the condition and to ensure appropriate ongoing review and support.

Introduction

Constipation in childhood engenders in clinicians unwarranted anxieties about management. Defining terms and understanding normal physiology and its variants, the frequent blurring in paediatric medicine between physical and emotional factors and their impact on normal development along with evidence-based data can provide the clinician with an algorithm for management.

Management varies according to age and whether constipation is acute or chronic. The emergency department (ED) is a difficult place from which to manage constipation, especially chronic constipation, for success requires ongoing maintenance therapy and contact with a committed and interested clinician. This is best achieved through the child's local doctor, with input from a sympathetic paediatrician in difficult cases if deemed appropriate.[1,2]

Definitions

1. *Constipation* is delay or difficulty in defecation, sufficient to cause significant distress.
2. *Encopresis* is the passage of a normal stool in an abnormal place.
3. *Soiling* is the frequent involuntary passage of loose or semi-loose stools in clothing. This is usually overflow incontinence as the liquid stool escapes around an impacted rectal facility.

Pathophysiology

The rectum and anal canal have two tasks: to store faeces temporarily and to evacuate at a socially convenient time. The distending rectum evokes a wave of contraction with inhibition of the smooth muscle tone of the internal anal sphincter, resulting in a sensation of the urge to defecate. An urgent desire to defecate occurs as the stool stretches the sensitive zone of the upper anal canal. This urge is overcome by the voluntary contraction of the external sphincter and the levator ani muscles. Eventually the rectum habituates to the stimulus of the enlarging faecal mass and the urge to defaecate subsides. With time this retentive pattern can become automatic.

It is understandable, therefore, that the child who is afraid to use the non-private, wet, smelly school lavatory, and allows his or her rectum occasionally to overcome the external sphincter, as he is relieved to arrive home, albeit with

‘poo in the pants’ – is really a normal variant rather than true encopresis.

There is a wide variation of physiology and normal development as can be seen in the age range of successful potty training. To produce a stool at will is one of the child’s first major achievements and most gain satisfaction from framing their success in a pot. If too much persuasion is provided, especially if full control has never been attained, the child’s profound disappointments are compounded by disapproval and hostility from the parents. Like most adults, most children seek solitude to defecate.

Management basics

1. Be interested. The patient has often been pushed from pillar to post, with a ‘quick fix’ and minimal supportive follow-up. Recognise that the parents and the patient have a legitimate concern, which can be the cause of major family dysfunction.
2. Endeavour to treat from the top, orally, rather than continue to direct attention to the rectum and anus with suppositories and enemas. The exception is when a fissure needs managing with ointment or local anaesthetic ointment if defecation can be anticipated.
3. Develop a pharmacological armamentarium of stool softeners or osmotic aperients. Stimulants (e.g. senna) may cause abdominal discomfort with colic in infants.

Table 7.13.1

Options for oral laxatives in children

Trade name	Active ingredient/class	Dosage	Tips
• Parachoc™ • (chocolate) • Agarol™ (vanilla) • Plain paraffin oil	• Paraffin oil • Stool softener	• 1–6 years old 10–15 mL day • 6–12 years old 15–20 mL day • >12 years old up to 40 mL day	• Can cause orange oil seepage in underwear • Can mix in foods, mixes well in ice-cream • Avoid in children with swallowing problems due to aspiration risk
• Osmolax™ • Clearlax™	• Macrogol 3350 • Iso-osmotic laxative	• 4–5 years old 1 large scoop day • 6–12 years old 1.5 large scoops day • >12 years old two large scoops day	• Tin with double ended scoop – large (17 g) and small (8.5 g) • Mix 17 g scoop with one cup of hot or cold liquid • Same active ingredient as Movicol without electrolytes (no salty taste) • PBS listed (authority not required)
• Movicol™	• Macrogol 3350 + electrolytes • Iso-osmotic laxative	• 2–5 years old: one sachet Movicol™ half day • 6–11 years old: one full strength day • >12 years old one to three full-strength day	• Movicol™ full strength 13 g (lemon-lime/choc/ flavour free) • Movicol™ half 6.9 g (lemon-lime) • Movicol™ junior 6.9 g (flavour free) • Dissolve full strength sachet in one-half cup liquid • PBS listed (authority not required)
• Actilax™	• Lactulose • Osmotic laxative	• 6 months old–1 year old: 3–5 mL day • 1–6 years old 5–10 mL day • 7–14 years old 10–15 mL day	• Can mix with water, milk or juice • Can cause bloating/ abdominal discomfort
• Coloxyl™ drops	• Poloxamer • Stool softener	• <6 months old 0.3 mL tds • 6–18 months old 0.5 mL tds • 18 months old–3 years old 0.8 mL tds	• Can mix in formula or juice (for Coloxyl+Senna, Senna is the stimulant component and should be avoided unless stools are soft)
• Dulcolax™ drops or tablets	• Sodium picosulfate/ bisacodyl • Stimulant	• 4–10 years old: 5–10 drops nocte • >10 years old: 10 drops nocte or 1–2 tablets nocte	• Useful for patients who cannot tolerate large volumes of liquid • Avoid if impacted • Can cause abdominal cramps • Do not use long term

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4. Be aware that ongoing management by a single interested clinician is desirable as the constipation will often relapse, and gains are often small (three steps forward, two steps back).
5. This makes constipation difficult to manage in EDs. Such departments need to have appropriate and willing referral resources. Outpatient appointments rarely work for the same reasons – difficulties with continuity of care and the ability to be seen relatively urgently (albeit that encouragement with management, behavioural modification and aperient dose titration is often all that is necessary and can often be done by phone by the interested clinician).
6. The significant risk of relapse dictates that a period of maintenance therapy is necessary before weaning off medications. It might be appropriate to let the patient and family know that if it has taken 2 years to arrive at this stage it is highly likely it will take 2 years or so to regain the rectum's natural motility and sensitivity.

Constipation in babies

Never again in life will the stooling pattern, and indeed the stools, be so closely examined as in the period of nappies being changed prior to toilet training. Straining at stool is often marked and then misconstrued as constipation whereas it is a simple reflection of the urge to defecate sensation. Plantar flexion of the toes is a similarly objective sign (at all ages).

Breast-fed infants may pass a stool after each feed or as infrequently as once every few weeks. As long as the stool is of normal quality (often referred to as scrambled eggs) and of great volume there is no reason for concern. For a couple of days prior to defecation the infant may be, not unreasonably, somewhat unsettled.

Just weaned, bottle-fed infants may produce dry, hard stools with difficulty and sometimes traces of fresh blood. Attention to water intake, perhaps addition of extra sugar (brown sugar is better) or sorbitol (1–3 mL kg^{-1} nocte) will help soften the stool. Formula switching is usually unproductive, although it is recognised that some forms of cows' milk intolerance may present with persistent constipation.

Should constipation persist and the clinician is confident that mother's description is of constipation, red flags should alert other considerations.

Fever, vomiting, bloody diarrhoea, failure to thrive, anal stenosis, abdominal

distension, history of delayed passage of meconium, polydipsia or polyuria should prompt a search for physical causes. It is important to assess for evidence of occult spina bifida (tuft of hair or deep sacral cleft) and any lower limb neurological abnormalities. A rectal examination is not required routinely; however, the anal area should be visualised to exclude fissures or obvious stenosis.

Box 7.13.1 Conditions possibly causing persistent constipation

- Meconium ileus
- Anal anomalies (stenosis, stricter, malposition)
- Spinal anomalies (spinal dysraphism, sacral agenesis)
- Hirschprung's disease
- Hypothyroidism
- Metabolic – hypercalcaemia, hypokalaemia
- Coeliac disease
- Cows' milk intolerance
- Lead poisoning
- Intellectual impairment
- Child abuse
- Cystic fibrosis
- Dietary – inadequate roughage, excessive cows' milk intake
- Psychogenic – parent-child and environment issues
- Drug related – anticholinergic, sympathomimetics, codeine.

Conditions to consider are listed in Box 7.13.1.

Acute constipation

Acute constipation often occurs after febrile illnesses, change in diet or environment, especially when intake has been low (e.g. post-operatively). A fissure in ano, usually from the passage of a hard faecolith, may cause pain, sphincter spasm, withholding and the start of a vicious cycle leading to chronic

constipation with megacolon and overflow incontinence.

Most acute constipation resolves spontaneously but given the risk of chronic constipation, especially if there has been a previous tendency to constipation, it may be prudent to commence gentle oral therapy early.

Chronic constipation

Chronic constipation is defined as persistent delay and difficulty in defecation often associated with soiling (overflow incontinence). Symptoms may be temporarily relieved by laxatives but relapse rapidly. This is why the ED is not the ideal place for long-term management, but may offer acute intervention for immediate relief of unpleasant symptoms and to gain the family's confidence by explaining management and natural history.

These children usually present in the school-age group. There may be a history of fissure, pain and withholding then ongoing constipation for months or years. The colon, rectum and upper anal canal become loaded with faeces, insensitive and hypotonic. Soiling develops with loose faeces escaping around an inspissated hard faecolith. Faeces are often palpable per abdomen. Social rejection in the class, due to the malodorous state, is often the first impetus for the patient themselves to seek cure. This, with loss of ability to defecate at will, takes a toll on the child's emotional stability. They have often undergone multiple management regimens from dietary manipulation to manual disimpaction, delivered by a variety of healthcare workers.

Investigations

Abdominal X-rays are not recommended as a routine part of the assessment of children with constipation.[3] They have poor inter-observer agreement for the diagnosis of constipation,[4] rarely change management, and a 'positive' diagnosis (demonstration of stool within the bowel) is associated with delayed diagnosis of other pathology.[5]

If constipation is persistent despite appropriate ongoing non-pharmacologic treatment and laxative therapy, it may be appropriate to investigate for medical causes (Box 7.13.1).

Management

The management of acute versus chronic constipation varies mainly in nuance and the extent and duration of treatment.

The aim of treatment is that in gaining the family's confidence the clinician recognizes that there is a very real concern, and a plan can be instituted to:

1. exclude physical treatable causes with careful history, examination and investigations as warranted
2. empty the rectum, preferably 'from the top' i.e. through *oral agents* rather than per rectum
3. establish a pharmacological bowel rhythm and pattern, which may need support for several years
4. allow the enlarged rectum/colon to re-establish its own inherent physiological bowel pattern and tone and to regain its normal sensation
5. continue maintenance for some time before considering slowly weaning off pharmaceuticals.

This will involve:

- enthusiasm on part of managing clinicians
- non-punitive behaviour on part of the parents
- adjunctive behavioural modification interventions with achievable goals, including motivational praise with star and reward charts, a stooling 'diary' and establishing toileting patterns such as 5 minutes on the potty after breakfast every day (tailored to the child's needs)
- acknowledging the need for patience, determination and resolution as the most chronic cases may take years to resolve.

Medications

Oral medication

Reference to the Bristol Stool Chart (Fig. 7.13.1) will help parents manage titrations of aperients to achieve the desired normal stool quality.

The common medications are listed below. Osmotic and lubricant laxatives can be used safely for months to years. Titrate dose to response, usually a single dose at night, aiming for a stooling pattern resembling Bristol Chart type 4 or 5 stools regularly.

In infants less than 6 months of age, poloxamer (Coloxyl™ drops) are

appropriate first-line therapy. These are also reasonable in those aged up to 12 months, although lactulose is another option in this age group. For older children macrogol or paraffin oil should be used initially.[6]

Bristol stool chart	
Type 1	 Separate hard lumps, like nuts (hard to pass)
Type 2	 Sausage-shaped but lumpy
Type 3	 Like a sausage but with cracks on its surface
Type 4	 Like a sausage or snake, smooth and soft
Type 5	 Soft blobs with clear-cut edges (passed easily)
Type 6	 Fluffy pieces with ragged edges, a mushy stool
Type 7	 Watery, no solid pieces. **Entirely liquid**

FIG. 7.13.1 Bristol Stool Chart

Disimpaction

If constipation is severe, the bowel may need disimpaction before maintenance treatment can be commenced. There are various options for bowel disimpaction, including oral, rectal and occasionally nasogastric.

- Oral outpatient faecal disimpaction

This is the preferred method for clearing the bowel, as it is effective in most cases, and much less distressing than the rectal or nasogastric approaches. Macrogol (international nonproprietary name for polyethylene glycol) 3350 sachets can be mixed in liquid and may be better tolerated if cooled in the fridge. They are administered in an escalating dose, and should be continued until the rectal effluent is clear.

Suggested dosing regimens (using full strength/'adult' sachets) include:

- Age 2–5 years: one sachet on day 1, two sachets on days 2 and 3, three sachets on days 4 and 5, four sachets on days 6 and 7
- Age 5–11 years: two sachets on day 1, three sachets on day 2, four sachets on day 3, five sachets on day 4, then six sachets daily
- Age 12 or more years: eight sachets daily.

Rectal medication for disimpaction

This should be avoided when possible, and used only if there is significant acute distress relating to a large impacted stool in the rectum. Rectal medication administration is very uncomfortable – consider using procedural sedation with nitrous oxide or midazolam.

Options include:

- Glycerine suppositories, which are particularly helpful in acute constipation
- Sodium citrate 5 mL enemas (Microlax™)
- Phosphate enemas should be avoided in children less than 2 years old.

Their persistent use may lead to problems with hyperphosphataemia, hypocalcaemia and tetany.

Inpatient disimpaction

This is usually a planned admission involving administration of bowel preparation fluid via a nasogastric tube. This usually occurs at a rate of 25 mL kg hour (up to 400 mL hour), along with maintenance oral fluids. Risks of this therapy include nausea, bloating, cramps, aspiration pneumonitis and dehydration.

Maintenance therapy

This focuses on the prevention of recurrence and *should be instituted straight after successful disimpaction*. It involves, as above, dietary interventions, behavioural modification, laxatives, patience and enthusiasm. Explain the natural history of paediatric constipation, and establish realistic expectations: there is no ‘quick fix’, and the child may be on laxatives for many months. Ensure appropriate follow-up with the child’s GP.

Dietary changes

Those commonly advised include increased fluid intake, absorbable and non-absorbable carbohydrate (sorbitol is found in prune, pear and apple juice). No randomised controlled study confirms the benefit of this. Forceful implementation would therefore seem undesirable.

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7.14

Inflammatory bowel disease

Ed Giles

ESSENTIALS

1 Inflammatory bowel disease (IBD) often presents in adolescence with symptoms depending on site and severity of intestinal involvement.

2 Crohn's disease can present as asymptomatic growth failure.

3 Faecal calprotectin is a good screening test for IBD, but is elevated in all intestinal infections/damage, so is a poor test in the acute setting.

4 In ulcerative colitis (UC), immunosuppressant medication may mask physical signs of toxic megacolon.

5 Gastrointestinal infections, including *C. difficile*, are increased in patients with IBD, and can mimic disease flare.

Introduction

Inflammatory bowel disease (IBD), comprised predominantly of Crohn's disease (CD) and ulcerative colitis (UC), is a chronic inflammatory condition of unknown aetiology, thought to be an aberrant immune response to commensal microbiota in genetically susceptible hosts.[1] It affects approximately 1 in 250 people aged 5–40 in Australia, with an increasing incidence. Over 25% of patients with IBD present <18 years of age; [2,3] most commonly they are older than 10. In patients under 6 years with diarrhoea and poor weight gain, primary immunodeficiences must also be considered.

Broadly, emergency department (ED) management of IBD comprises either diagnosis of new patients or management of flares of patients with existing disease on the background of their current treatments.

New diagnoses

Definitive diagnosis of IBD is made with a combination of clinical, radiological, endoscopic and histological features. However, there are patterns of disease that point to either subtype (Table 7.14.1).

Presentation

Chronic diarrhoea is defined by the presence of diarrhoea for more than 2 weeks. Acute presentations of IBD may be shorter; however, it is particularly important to consider infection – a much more common cause of acute diarrhoea. Important historical features include travel, infectious contacts, family history of IBD and any extra-intestinal manifestations (Table 7.14.2), which can occur with either UC or CD.

The distribution of the disease pathology within the gut will determine the symptoms. Colonic inflammation (in CD or UC) presents with diarrhoea, usually with blood, whereas small bowel disease can be subtle, even asymptomatic. Distal colonic disease/proctitis commonly presents with urgency and tenesmus. Abdominal pain may be present with either condition, although significant pain consistently after eating suggests a stricture (see Table 7.14.2).

Examination

In mild/moderate IBD cases the examination is usually unremarkable. Extraintestinal features (Table 7.14.2) may be present. Abdominal tenderness can be a sign of the severity of intestinal inflammation. A mass, point tenderness or peritonism can suggest an abscess or other complication of CD, which is most often found in the right iliac fossa (terminal ileum). Growth parameters are important, but are difficult to interpret without serial measurements.

Table 7.14.1

Common clinical features of Crohn's disease and ulcerative colitis

Clinical feature	Crohn's disease	Ulcerative colitis
Diarrhoea	++	+++
Bloody diarrhoea	+	+++
Urgency/tenesmus [6]	+	+++
Abdominal pain [7]	+++	++
Growth retardation	+++	+
Perianal disease	May be present	No
Extra-intestinal features	May be present	May be present

Table 7.14.2

Clinical features of the more common or classic extra-intestinal features of inflammatory bowel disease

Extra-intestinal manifestation	Presentation
Erythema nodosum	Painful/tender red rash classically on anterior leg. Large differential diagnosis but often associated with IBD
Pyoderma gangrenosum	Rare necrotising ulcers typically on the legs
Arthritis	Arthralgia common but true arthritis less so. Usually monoarticular. Can be presenting symptom of IBD
AIH/PSC/overlap syndrome	Approximately 10% of IBD patients have liver involvement (abnormal LFTs)
Orofacial granulomatosis	Significant mouth and/or lip swelling with abundant granulomas on biopsy, related to CD
Others	Uveitis/iritis/episcleritis Ankylosing spondylitis Pancreatitis Nephrolithiasis

AIH, autoimmune hepatitis; CD, Crohn's disease; IBD, inflammatory bowel disease; LFT, liver function test; PSC, primary sclerosing cholangitis.

Table 7.14.3

Investigations for known or suspected inflammatory bowel disease patients comparing Crohn's disease and ulcerative colitis (other tests depending on the clinical circumstance would include stool M/C/S and *C. difficile* toxin)

Investigations	Crohn's disease	Ulcerative colitis
FBC +/- iron studies	Iron deficiency anaemia; high platelet count; rarely normal	Iron deficiency anaemia; high platelet count; often normal in mild-to-moderate disease
Inflammatory	Raised in active disease	Often normal in mild-to-moderate disease

markers (CRP/ESR)		
LFT incl Albumin	Albumin low in active disease; can be co-existent liver disease (AIH/PSC)	Albumin only low in severe disease; can be co-existent liver disease (AIH/PSC)
Faecal calprotectin*	High ++	High +++

AIH, autoimmune hepatitis; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; FBC, full blood count; LFT, liver function test, PSC, primary sclerosing cholangitis.

* Faecal calprotectin not useful in the acute setting.

CD can present with perianal disease – tags, fissures, fistulas and/or abscesses. In general patients will complain of symptoms in this area but given the patients are usually adolescents they are often embarrassed. It is important to visually examine this area, although a PR is rarely indicated.

Investigations in the emergency department

There are no specific tests for diagnosing IBD in the ED. Table 7.14.3 shows which tests are suggested for both potential new patients and known IBD patients, and shows the differences between CD and UC. Specific testing for serological markers should only be done in discussion with a paediatric gastroenterologist.

In addition, any patient with new onset chronic (>2 weeks) or bloody diarrhoea should have stool M/C/S and *Clostridium difficile* toxin sent, and consideration for tests for parasites.[4] Asymptomatic *C. difficile* colonisation is very common in children aged <2 years; routine testing is not recommended without discussion with gastroenterology or infectious disease teams. Faecal multiplex tests often provide positive results of questionable significance due to their extremely sensitive nature, and must be interpreted with caution.

Importantly, when patients are in complete ('deep') remission from their IBD all of their investigations (blood tests, faecal calprotectin, even endoscopy) would be normal. Plain X-rays are rarely helpful.

Differential diagnosis

Even excluding infections (including parasites), the list of differential diagnoses for patients presenting with symptoms of possible IBD is very broad (Table 7.14.4). Determining between IBD and IBS in particular can be difficult but it is not the role of the emergency physician to make a definitive diagnosis.

Other rarer differentials include Behçet's disease, allergic gastroenteritis, food

protein-induced enterocolitis syndrome (FPIES), Yersinia infection, intestinal tuberculosis (TB), Meckel's diverticulum, immunodeficiency including chronic granulomatous disease (CGD), intestinal lymphangiectasia, Henoch-Schonlein purpura (HSP), graft versus host disease and ischaemic enterocolitis.

Diagnosis/management

It is virtually impossible to make a certain new diagnosis of IBD in the ED; however ruling out certain differentials can be extremely useful in consultation with a paediatric gastroenterology team. In particular, sending off stool investigations can prevent diagnostic delay.

Broad spectrum antibiotics including anaerobic cover should only be commenced if there is clinical concern about perforations or collections. In the absence of severe sepsis, antibiotic administration should occur in discussion with the accepting specialty unit.

The decision on whether patients need admission will depend on the severity of their symptoms. In particular, patients with significant bloody diarrhoea, especially those at risk of toxic megacolon (see later) must be admitted.

Table 7.14.4

Comparison between inflammatory bowel disease and differentials in clinical presentation and basic investigations

Differential	Clinical comparison to inflammatory bowel disease	Investigations
Irritable bowel syndrome	May have a range of bowel and other symptoms, but constitutionally well with normal growth	Normal blood tests, normal faecal calprotectin
Appendicitis/other surgical abdomen	Usually shorter history and more localised signs/peritonism	May be similar, no chronic disease (e.g. no iron deficiency)
Coeliac disease	Variety of GI and other symptoms and affects growth but no bleeding	Normal inflammatory markers. Coeliac screen
Intestinal polyps	PR bleeding that is persistent but otherwise well child	Normal inflammatory markers
Helicobacter pylori infection	Epigastric pain, may cause bleeding ulcers, demographic risk factors	Normal inflammatory markers, *H. pylori* stool antigen/breath test/gastroscopy
Eating disorders	Usually less bowel symptoms but can be difficult especially compared with TI CD	Normal inflammatory markers, normal faecal calprotectin.

GI, gastrointestinal; PR, per rectum; TI CD, terminal ileal Crohn's disease.

Known IBD patients

As a relapsing and remitting condition, with varied and often unknown triggers, patients with IBD will present to the ED with various complaints, including complications of therapies.

Crohn's disease

CD can affect anywhere from mouth to anus but ileo-colonic disease is most common in children. Inflammatory flares are usually associated with diarrhoea +/- blood +/- abdominal pain. These patients must be assessed for any differential diagnoses, particularly infection. Depending on the situation patients may require admission for analgesia, rehydration, and/or bowel rest. Any decisions regarding IBD medications (such as steroids or specific immunosuppressive therapy) should be made in consultation with the treating gastroenterology team.

Strictures most commonly affect the distal ileum but can affect any area of the bowel. This is usually a result of longstanding disease, but can be found at initial presentation. Patients will have symptoms of obstruction or partial obstruction. Treatment is supportive, with nasogastric tube insertion and early surgical review, although surgery is not always indicated.

Fistulising disease (apart from perianal disease) is rare in paediatrics, but can occur. This may lead to faeculant discharge on the abdominal wall, or entero-vesical fistulae with faeces/air passed per urethra. Most commonly fistulae present infected with an abscess and, even if IBD diagnosis is not known, a right iliac fossa abscess (often thought to be appendicial in origin) may turn out to be complicated CD.

Perianal disease

Perianal disease is less common in paediatric than adult CD but still occurs, and can be the first presentation of CD.

Perianal disease is classically fistulae between the lower rectum and the skin, which are easily infected. They will present with discharge which will become purulent and increase with infection, with subsequent pain due to abscess formation. Once there is a significant abscess only surgical management will be successful, although small collections and discharge can be managed with prolonged courses of anti-anaerobic antibiotics.

Paediatric patients who present with perianal disease are often teenagers who can be extremely embarrassed and must be approached considerately. Any patient with repeated fissures, complex fistulae or large anal skin tags should be referred to a paediatric gastroenterologist for consideration of possible CD.

MRI is the best imaging modality for perianal disease, particularly if it is unclear whether surgical management is needed. However, any advanced imaging should be arranged in consultation with the admitting unit.

Ulcerative colitis

UC patients typically present with bloody diarrhoea, but once again infection must always be considered. Indeed, in a sudden flare it may be more reasonable to treat with antibiotics, such as ciprofloxacin and metronidazole, until stool M/C/S and *C. difficile* toxin results are available. Beyond this, the severity of symptoms can be quantified using the paediatric UC activity index (PUCAI), a 6-item questionnaire assessing pain, bleeding, stool consistency and frequency, nocturnal stools and activity level.[5] This guides need for admission and gives a baseline to judge response to therapy, and possible need for escalation of treatment or colectomy.

Toxic megacolon

The most life-threatening complication of IBD is toxic megacolon, which is rare in children. Adult guidelines for the definition of toxic megacolon are not validated in children but are likely to be similar, particularly in teenagers. In general, patients who are persistently tachycardic, febrile, have abdominal distension with guarding, or are shocked should have an urgent surgical opinion and X-ray (Fig. 7.14.1). Importantly, in known UC patients who are already on immunosuppression, some of the clinical signs (such as tenderness/guarding) may be affected by the treatment, and so should be assessed even more carefully, with a low threshold for imaging.

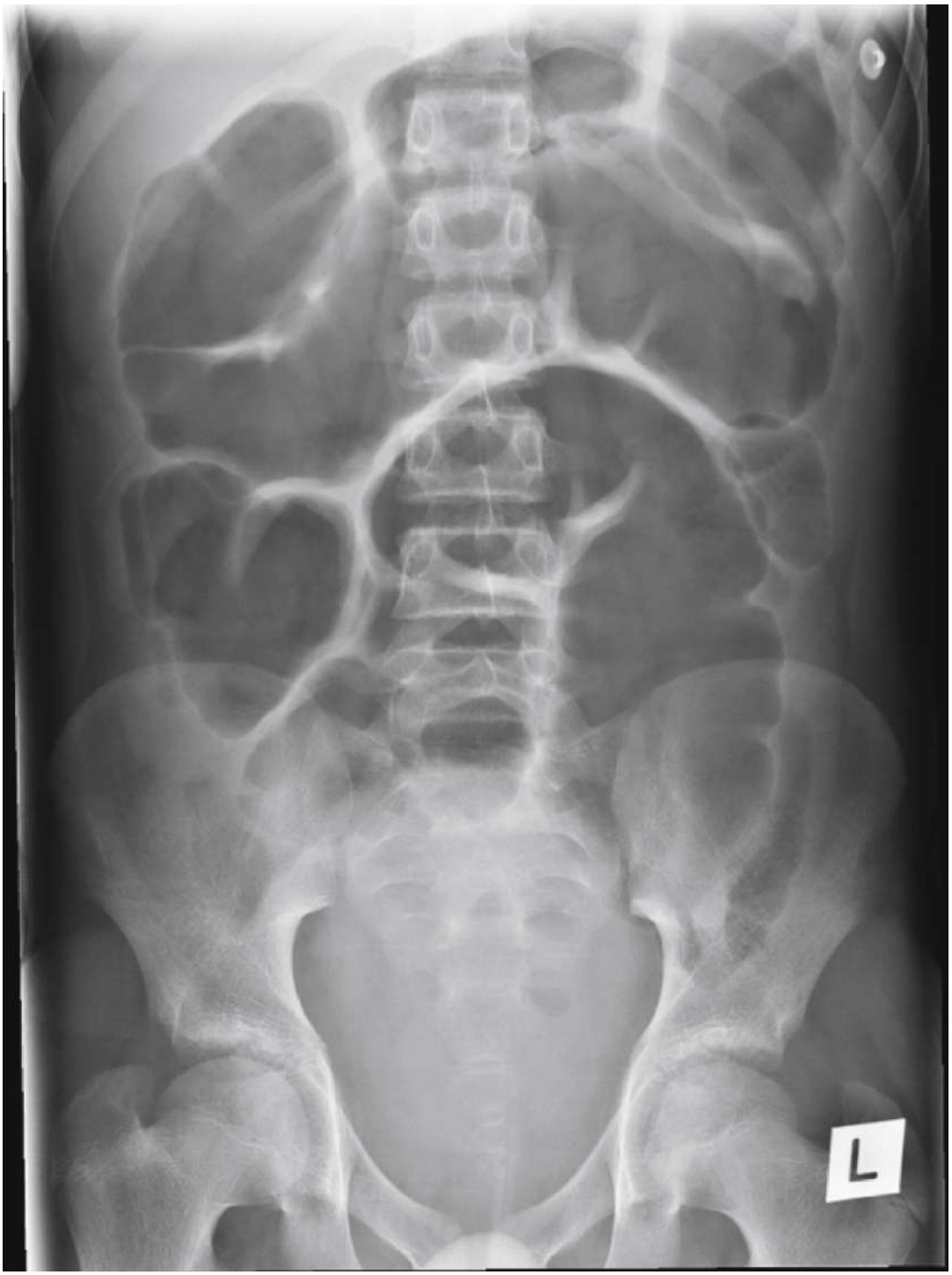

FIG. 7.14.1 Example of toxic megacolon with grossly dilated colonic loops.

If toxic megacolon is identified or strongly suspected, the patient should be stabilised and appropriately fluid resuscitated. Urgent surgical assessment is indicated, with the patient remaining nil by mouth and given broad spectrum IV

antibiotics including anaerobic cover (see Fig. 7.14.1).

Common treatments and their complications

Corticosteroids remain a mainstay of therapy for induction of remission in both moderate and severe CD and UC, and should be initiated in consultation with a gastroenterologist.[6] For CD, an alternate and equally effective therapy is exclusive enteral nutrition (EEN).[7] EEN is where patients cease all food intake for a period of 6–8 weeks and drink a nutritionally complete feed. This is a difficult therapy to adhere to but has the advantages of no side effects, and improving the nutritional state of the patient.

The majority of paediatric IBD patients are on long-term immunosuppressive therapy. Not only does this put them at risk of infections, but they may also present with side effects from the medications themselves, particularly in the weeks after a new drug has commenced. Azathioprine is the usual first choice maintenance treatment with complications of bone marrow suppression, liver dysfunction and pancreatitis. Many patients are now treated with biological therapies, particularly mono-clonal antibodies to TNFα (e.g. infliximab) and there will be more new treatments in the years ahead.

Controversies

1 The use of antibiotics in acute flares of IBD. There is modest evidence for the use of ciprofloxacin and/or metronidazole in flares of Crohn's disease in adults but not in UC. However, they are often used in adults and children in IBD even when infection has been excluded.

2 Use of serological markers. pANCA, ASCA and other auto-antibodies can be measured to predict type and severity of IBD but there is no consensus on their utility.

3 The use of DVT prophylaxis in acute severe flares is well established in adults but is less clear in children.

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SECTION 8

Neurology

OUTLINE

8.1

Cerebrospinal fluid shunt complications

Richard Lennon

ESSENTIALS

1 Emergency physicians should consider the possibility of a shunt complication in any child with a cerebrospinal fluid (CSF) shunt presenting to an emergency department because symptoms and signs of complications may be non-specific, subtle and of gradual onset.

2 The most common shunt complications are malfunction (undershunting and overshunting) and infection.

3 Shunt infection is much more likely to occur in the 6 months following insertion or revision.

4 CT scan and plain X-rays of shunt hardware ('shunt series') are the preferred imaging methods for detecting shunt malfunction; however, these tests are not 100% sensitive, and patients should be referred to the neurosurgical service if clinical suspicion persists. Rapid acquisition MRI scans may be a radiation-sparing alternative to CT scans.

5 Before performing any procedure on a shunt, it is best to consult with the treating neurosurgeon.

Introduction

The intervention of cerebrospinal fluid (CSF) shunting for CSF accumulations

has brought about long-term survival and avoidance of disabilities in children suffering from hydrocephalus. Unfortunately, the insertion of inert non-growing hardware in infants and children is associated with a high rate of shunt complications. Some studies find from 33% up to 60% or more shunts need revising after several years.[1] The task of the emergency physician is to diagnose those complications, commence time-critical treatments and refer to a neurosurgical service when appropriate.

Types of shunt

Many types of CSF shunts may be encountered in paediatric emergency practice. There are also many different types of shunt hardware; however, most have the same basic structure, which comprises a proximal tube that takes CSF, usually from the lateral ventricle through brain tissue and a small burr hole, to the outer surface of the skull.

At or near this point there is usually a silicone reservoir dome that can be used for sampling and/or pumping CSF; then there is a subcutaneous one-way valve that is set to open at a certain pressure differential in order to avoid overshunting. Some of these valves are ‘programmable’, meaning that the opening pressure can be adjusted by using a magnet. These settings may be accidentally changed by strong magnetic fields (e.g. MRI) or by G-forces at amusement park rides;[2] therefore a history of such exposure should be elicited in these cases. As well as this, there may also be an antisiphoning device which increases its opening pressure in the erect posture or when pressure falls below atmospheric to avoid overdrainage.

The distal tubing is tunnelled under the skin to the drainage site, which is most commonly the peritoneal cavity. The distal catheter usually contains valves that prevent back flow.

Variations on the positioning of the proximal tubing include placement in the subdural or subarachnoid space and placement in posterior fossa cystic malformations such as in the Dandy– Walker syndrome, and also in the spinal canal as in lumboperitoneal shunts.

Variations in the placement of the distal catheter include the right atrium, the pleural cavity and the gallbladder. By far the most common is the peritoneal cavity, with these alternatives only being used if the peritoneal site is contra-indicated. Some or all of the tubing may be impregnated with antibiotics; evidence suggests that this decreased the risks of infection.[3]

There is another treatment of hydrocephalus called endoscopic third ventriculostomy. In this situation an endoscope is passed through a burr hole, brain tissue and a lateral ventricle into the third where a hole is made in the floor of the third ventricle communicating with the prepontine subarachnoid space so that CSF may bypass any obstructions below the third ventricle. This is not so much a shunt as a diversion, and no artificial hardware is left in the patient. The main complications are bleeding, infection, short-term memory problems and endocrine abnormalities in the early post-operative period. After this time, the most common complication is blockage of CSF flow and recurrence of hydrocephalus. This is the most likely complication to present to ED and should be managed as per a blocked shunt.

Shunt problems that may present to an emergency department (ED) are listed in Box 8.1.1.

Clinical presentation

The developmental stage of the child with a CSF shunt can cause considerable variation in the clinical presentation of shunt complications. The presence of an open anterior fontanelle in infants up to the age of approximately 9 to 18 months allows the estimation of intracranial pressure by simply looking and feeling the fontanelle. A bulging fontanelle is a highly specific but not very sensitive sign of under-shunting; a sunken fontanelle can be a sign of overshunting.

The cranial sutures in children are not fused and can undergo diastasis due to raised intracranial pressure. This means that the head circumference will rapidly increase when there is inadequate shunting. Therefore it may be useful to measure the occipitofrontal circumference and compare it with previous records if available and plot it on growth centile charts. A head circumference that is rapidly crossing centiles in an upward fashion or a head circumference that is in the very high centile range, especially when the other parameters, weight and length, are not, is an indicator of undershunting. Another implication of unfused cranial sutures is that because this allows an increase in cranial volume it retards the rise in intracranial pressure. This may be the reason that small infants present with more non-specific signs and symptoms than older children.

Box 8.1.1 Complications of cerebrospinal fluid shunts

Infection
Malfunction:
- Blocked proximal catheter
- Blocked distal catheter
- Loculation of lateral ventricle
- Valve dysfunction
- Overshunting/slit ventricle syndrome
- Disconnection

Abdominal pseudocyst
Migration of distal catheter
Invasion of abdominal organs (VP shunt)
Peritonitis, ascites (VP shunt)
Bowel obstruction (VP shunt)
Pulmonary emboli (VA shunt)
Glomerulonephritis (VA shunts)
Brain tumour metastases
CSF ascites
Pleural effusion (pleural shunts)

The most common question an emergency physician will have to answer when confronted with a child who has a CSF shunt is 'Does this patient have a risk of shunt complication such that I should refer this patient to the neurosurgical service?' A study by Piatt et al.[4] attempted to quantify the power of various symptoms and signs to predict shunt malfunction or infection. Box 8.1.2 shows the strongly predictive signs and symptoms that allow referral to be made on the basis of that single finding. This may be done even before CT scanning, as the neurosurgeon may prefer to have the scan done locally to allow for easier comparison with previous scans (see discussion of CT scanning below).

Box 8.1.3 shows the signs and symptoms with strong positive predictive power but not strong enough to warrant immediate referral if just one feature is present on its own. Thus in a patient with a ventriculoperitoneal (VP) shunt who presents with fever alone, an initial general workup for a cause of the fever is warranted and then consideration for referral to neurosurgery made if no definite cause for the fever is found. However, fever with another feature listed in Box 8.1.3, such as headache, warrants early neurosurgical referral.

The question of whether to send the child home is more difficult. The absence

of any of the symptoms and signs listed in Boxes 8.1.2 and 8.1.3 does not rule out shunt malfunction or infection. If concerned about shunt infection, this is much less likely if the patient is more than 6 months from the last shunt insertion or revision. However, this does not exclude shunt malfunction. Where a symptom or sign that is not of high predictive power is adequately explained by another diagnosis (e.g. vomiting with diarrhoea and recent contact with a case of gastroenteritis) CSF shunt complication is very unlikely. For cases where CSF shunt complications are neither ruled in nor out on historical and examination findings, one must resort to investigations and/or observation and/or neurosurgical consultation.

Box 8.1.2 Signs and symptoms that on their own warrant immediate referral to neurosurgical service

Bulging fontanelle
Decreased level of consciousness
Fluid tracking around shunt tubing
Loss of upward gaze (sunset eyes)
Signs of local infection (usually <6 months post-operative):
- Erythema of site
- Erosion/ulceration
- Cerebrospinal fluid leak
- Purulent drainage

Meningismus
Peritonitis

Box 8.1.3 Symptoms and signs may warrant neurosurgical consultation (see text)

Abdominal pain
Fever
Nausea/vomiting
Irritability
Headache
Abnormal shunt pump test

Accelerated head growth

History

Parents may present very early, reporting vague and non-specific symptoms, particularly if the child has had a similar presentation of shunt malfunction previously. It is important to take parental concerns seriously. They have spent the most time with the child and will be able to pick up subtle behaviour changes that an ED clinician, who may have never seen the child before, cannot.

The most common presentation of children with shunt obstruction is headache, vomiting and/or drowsiness. It is notable that seizures occur on neither of the above lists. This is because many children with shunts also have epilepsy; therefore seizure alone has a poor correlation with shunt complication. It is important to remember that distal shunt problems may present with abdominal symptomatology such as pain and distension.

Symptoms suggesting possible infection include fever, lethargy, irritability, or features of meningism. There may be concurrent symptoms of shunt obstruction.

Examination

For examination features of raised intracranial pressure see Chapter 8.2. Features of infection, e.g. fever, rigors, lethargy, localised redness, swelling and peritonism (ventriculoperitoneal shunt), pleurisy (ventriculopleural shunt), or bacteraemia (ventriculoatrial shunt), should be sought.

Shunt evaluation

The extent of the shunt course itself should be examined thoroughly. The older child is usually the best at finding the shunt hardware under his/her hair. This should be palpated and, if possible, inspected for inflammation of the skin along the shunt route. Many have a silicon-pumping bulb, which is used both for checking the patency of the shunt and for access to sample CSF.

Most reservoirs can be compressed easily and rapidly refill in a few seconds. Incompressibility of the bulb is usually due to obstruction of the distal catheter. This is the less common site of obstruction. If the bulb compresses easily but does not refill, then the proximal catheter is blocked. This is the ‘shunt pump

test’. An abnormality of the test did not perform well enough to warrant immediate referral in Piatt et al.’s study.[4] This is because it can be difficult to tell if a test is abnormal or not. The return of the chamber after depression can take a number of minutes and may take longer if the choroid plexus is drawn into the catheter causing partial or complete obstruction. This is the reason why shunt pump tests should be kept to a minimum as multiple tests can cause blockage not only with choroid but also other debris and the ventricular wall and can also cause low pressure headache.

CSF tracking around the proximal catheter can form a fluctuant swelling around the burr hole in the skull where the shunt enters. This indicates a blocked proximal catheter or disconnection, and the child requires neurosurgical consultation.

The entire subcutaneous catheter should be carefully inspected and palpated for continuity and the abdomen examined for evidence of peritonitis or pseudocyst formation.

Investigations

On rare occasions, insertion of a needle into a shunt pumping chamber may be useful for both diagnostic and therapeutic reasons but should generally be performed by our neurosurgical colleagues.

The exception is in the rapidly deteriorating child, where contact with the neurosurgeon is delayed or where in remote locations after discussion with the treating neurosurgeon it is considered important to get some information to help with a decision to transport. In this age of modern communication it would usually be possible to get a neurosurgeon to ‘talk you through’ the procedure in real time.

In the case of a moribund child, an emergency physician may relieve the raised intracranial pressure by inserting a 23-gauge butterfly needle into the bulb/pumping chamber at 45 degrees to the skin under strict aseptic technique.[5] It is important not to advance too deeply as the needle can damage valve mechanisms so badly that replacement is necessary.

Once the needle is in the reservoir there are several diagnostic manouevres that can be done, and it would be good to ask the neurosurgeon who knows the child and the hardware best what he/she would prefer. One technique is to allow the end of the butterfly needle to drain freely while held 5 cm below the valve and to time the interval between drops. In one study a cutoff of >20 seconds was

95% sensitive and specific for proximal shunt malfunction.[6] Some authorities attempt to aspirate, and then the opening pressure can be measured with a similar technique to that used in a lumbar puncture. If it is high (e.g. >20 cm H_2O) and the child is moribund with raised intracranial pressure then CSF can be drained off until the pressure is 10–15 cm H_2O, depending on the opening pressure of the valve, and the clinical condition improves significantly. Then closing pressure can be measured.

The CSF should be sent for analysis as with a lumbar puncture specimen; however, cell counts and chemistry may be deceptively low even in the presence of infection, and culture is the gold standard for ruling out infection with bacterial PCR being a useful more rapid adjunct.[7]

In the more stable child, an X-ray of the entire shunt ('shunt study') may demonstrate a disconnection or kinking causing blockage. Several studies, however, have noted that the number of positive shunt studies is small (about 1%), and they are not associated with subsequent surgical revision. It would seem that most neurosurgeons would not operate on the basis of an abnormal shunt study alone. Therefore some EDs do not routinely do shunt studies but rather reserve them for those cases where the decision to operate has already been made on the basis of the neuroimaging for the purpose of planning the operation.[8,9]

The CT scan is the usual preferred method of imaging, because it provides clear images in a short space of time. However, difficulties associated with obtaining a CT scan need to be considered. One difficulty is keeping the child still long enough to obtain adequate images even with rapid CT scanners. In infants, firm wrapping and sucrose on a dummy or pacifier may be sufficient to keep them still. When these fail procedural sedation or general anaesthesia is required. This is less desirable because of anaesthetic risk, longer delays, use of resources, and the potential to temporarily obscure one of the important signs of raised intracranial pressure (ICP) and decreased level of consciousness.

Another consideration is the harmful effects of the radiation exposure. A protocol designed to minimise radiation exposure in children should be used and the head angulated to avoid radiation exposure to the eyes. This reduces the risk of premature cataract formation. These precautions may not be routine at institutions that do not frequently scan children, and the clinician may have to ensure that these things are done.

Some studies have examined MRI as an alternative to CT scanning. New rapid acquisition MRI protocols make it more feasible to use this form of imaging.[10] In

some cases (e.g. a stable child seen in a peripheral hospital for definite transfer) it might be better, after discussion with the treating neurosurgeons, to get the imaging done at a tertiary children's hospital where lower-dose CT or rapid cranial MRI is more likely to be available.

Enlarged ventricles on scanning may indicate undershunting due to obstruction. However, this may be chronic, and comparison with previous CT scans is necessary for accurate interpretation. Obliteration of the perimesencephalic cistern is a particularly worrying sign and mandates urgent neurosurgical consultation. The scan may also show the reason for malfunction (e.g. catheter tip embedded in brain tissue), or it may show a ventriculitis when the lining of the ventricles enhances with contrast. Alternatively, neuroimaging may reveal small 'slit-like' ventricles. These are thought to be due to chronic overshunting and may be non-compliant. This leads to sharp fluctuations in ICP, with very little change in CT appearance. The management of this complication is a neurosurgical challenge. The emergency physician needs to be aware that small ventricles do not necessarily mean a normal ICP.

Nuclear medicine shunt function studies are also used in some centres for diagnosing shunt blockage. An abdominal ultrasound may be useful to identify a CSF pseudocyst.

Aspects of some cerebrospinal fluid shunt complications

Infection

The diagnosis of shunt infection is not always straightforward. In a retrospective case series[3] the presenting symptoms were as listed in Table 8.1.1.

The positive predictive value of these symptoms was even greater in the first 9 months following insertion of the shunt because 80% of all shunt infections occur in this time. There is also an increase in VP shunt infections after laparotomy. Case reports also mention other symptoms such as rigors whenever a ventriculoatrial shunt is manipulated. The most useful investigation is initially the CSF white cell count, which is elevated in approximately 70% of cases. The blood white cell count is elevated in only 30% of cases. A positive CSF culture is the gold standard, although this may be negative in those children who have been on antibiotics. Bacterial antigen detection and polymerase chain reaction testing may be helpful in this situation.

Table 8.1.1

Shunt infection presenting symptoms[2]

Symptom	Percentage of cases of infection with symptom
Shunt malfunction	33%
Fever	26%
Localised wound or shunt tract inflammation	22%
Abdominal pain or pseudocyst	19%

Staphylococcus epidermidis is the most common bacterium isolated, followed by other coagulase-negative staphylococci and *S. aureus.* Less commonly gram-negative bacteria (such as *Propionobacterium),* *Streptococcus pneumoniae* and *Candida* infections have been reported.

Initial antibiotic therapy can be tailored to findings on Gram stain from a CSF tap and/or cultures from previous infections. If the patient has no contraindications, flucloxacillin or dicloxacillin should be in all empirical regimens with gram-negative cover considered after consultation with neurosurgeons and an infectious-disease specialist. In the majority of cases the shunt will have to be removed and replaced at a later date, with some temporary measure in the meantime.

Early post-operative complications

Wound dehiscence and purulent discharge are much more likely in the first few weeks after shunt insertion. If there is early overshunting this can lead to such a degree of brain shrinkage that the subdural bridging veins are broken and subdural haematomas form. Also, the presence of blood or fibrin in the CSF increases the risk of proximal blockage. At the other end of the shunt, peritonitis and wound infection are more likely in the early post-operative period.

Migration and penetration of shunts

In various cases the distal end of VP shunts has migrated into the thorax, liver, bowel lumen and even out of the anus and mouth. Several liver abscesses have been reported. Treatment involves removing the distal part the tube, possibly repairing damaged organs and often a temporary alternative shunting method.

Glomerulonephritis

This is a complication of ventriculoatrial shunts, which are rarely used today. This is usually a gradually progressive illness, which requires a high level of suspicion for diagnosis. Treatment is usually to remove the shunt and replace it with an alternative.

Trauma in children with a cerebrospinal fluid shunt

A recent multicentre prospective cohort of children with VP shunts who presented to EDs after trauma and a GCS of 14 or 15 had a reassuringly low incidence of clinically important brain injury (1%) but a higher rate of CT scan than the non-VP shunt cohort (46% vs. 35%). Overall this was a rare presentation with only 1 in 500 such head injuries having a VP shunt.[11] Therefore as far as head injury is concerned, the child with a CSF shunt should be assessed as any other child initially but with the following additional considerations:

1. *Was there a direct impact upon the shunt hardware?* Relatively minor trauma to the shunt may cause breakage and malfunction which may not become manifest until some days or weeks following the trauma; also, a direct blow may cause shunt movement and damage to tissues surrounding the shunt. Intracranial haemorrhage following moderate impact in sporting events has been reported. Impacts on distal tubing may cause shunt penetration of abdominal viscera.
2. *Is there an open wound that may communicate with the shunt or the CSF?* This would increase the risk of infection. Thus a scalp wound near the entry point of the tubing into the cranial cavity may cause pneumocephalus and may lead to meningitis. At the other end, a bowel perforation or penetrating trauma may cause a distal shunt infection.

In addition to these questions there is the possibility that the presence of the shunt and/or the underlying pathology may make the child more likely to have some complication of trauma. The most common example of this is subdural haematoma from rupture of bridging veins that have been stretched by brain shrinkage after shunt placement. Notwithstanding this, the latest data[11] do not

support excessive CT scanning in these children, and a period of observation with neurosurgical consultation may be a better course of action.

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8.2

Raised intracranial pressure

Richard Lennon

ESSENTIALS

1 Acute severe elevation of intracranial pressure (ICP) is a true emergency often requiring intubation and ventilation, elevation of the head of the bed 30 degrees, infusion of mannitol or hypertonic saline, treatment and prevention of fever and seizures, and early consultation with neurosurgeons.

2 The open anterior fontanelle allows direct palpation of ICP in children up to 9–18 months of age.

3 A rapidly expanding head circumference in an infant prior to suture fusion is an important sign of raised ICP.

4 In rare circumstances an emergency procedure, such as a subdural or a ventricular tap, can be performed through the anterior fontanelle in infants under the guidance of a neurosurgeon.

5 A brain CT scan or MRI can rule out many causes of raised ICP but does not exclude raised ICP itself and thus should not be the sole basis for deciding whether or not it is safe to perform a lumbar puncture.

Introduction

Normal physiology

Normal (10–90th centile) intracranial pressure (ICP) is 11.5–28 cm H_2O[1] in

children, and lower (3–8 cm H_2O) in newborns.[2] It is the product of the intracranial contents mainly blood, brain and cerebrospinal fluid (CSF), as well as CSF production rate and resistance to its flow and the resistance of the cranial vault.

Normal ICP has a diurnal cycle that is higher in the early hours of the morning when one is normally supine during sleep. Therefore the symptoms of raised ICP, such as headache and vomiting, are usually worse in the morning. Intracranial pressure may be raised by anything that can cause an increase in the volume of its contents or a decrease in the size of the cranial cavity (Box 8.2.1). This section will not cover the diagnosis and management of raised ICP associated with trauma. Those issues are covered in Chapter 4.2.

Most CSF is made by the choroid plexus in the lateral third and fourth ventricles. A normal child makes approximately 0.15–0.30 mL min^{-12}, and the total volume is 50 mL in an infant, rising to 150 mL in an adult. It flows through the foramen of Monro into the third ventricle, then down the aqueduct of Sylvius, which is usually only 2 mm wide and 3 mm long in a child. This leads into the fourth ventricle and from there via the foramina of Luschka and Magendie to the basal cisterns. From there the CSF flows over the surface of the cerebellar and cerebral hemispheres to be reabsorbed through the arachnoid villi on the superior sagittal sinus. CSF can also be reabsorbed through several other channels, including a small amount through the choroid plexus and via lymphatics. ICP will be raised whenever there is obstruction to the flow or reabsorption of CSF or in the rare circumstance where CSF production is increased.

Pathophysiology

The Monro-Kellie doctrine states that the intracranial volume is fixed and that the contents have three compartments: the blood, the brain and the extracellular fluid volume with CSF. In older children and adults if one of these compartments increases the others will shrink to maintain normal ICP. This usually means less cerebral blood volume and/or CSF. Once these compensatory mechanisms have been exhausted, ICP will rise.

Maintenance of adequate cerebral blood flow is mainly dependent on adequate cerebral perfusion pressure (CPP). This is equal to the mean arterial pressure (MAP) minus the mean ICP. Minimum values for CPP vary with age from 40 mmHg in 0- to 5-year-olds and 50 mmHg in 6- to 17-year-olds.[2] When ICP rises

blood pressure will rise in order to maintain CPP; this is often associated with reflex bradycardia and irregular respirations and is called the Cushing's response.

Box 8.2.1 Causes of raised intracranial pressure

Increased cerebrospinal fluid (hydrocephalus)

1. Decreased absorption:
    - Obstructive
    - Communicating
2. Increased production

Swollen contents

1. Meningitis
2. Encephalitis
3. Cerebral oedema:
    - Metabolic (hyper/hypo-osmolarity-natremia uraemic or hepatic encephalopathy)
    - Vasogenic
    - Post-ischaemic
    - Post-traumatic/diffuse axonal injury
4. Increased venous pressure:
    - Cerebral venous sinus thrombosis
    - Right heart failure
    - Jugular vein/superior vena cava obstruction

Space-occupying lesion (SOL)

1. Tumours:
    - Primary
    - Secondary
2. Haematomas:
    - Extradural
    - Subdural
    - Intracerebral/intraventricular

3. Abscesses/cysts
4. Arteriovenous malformation
5. Congenital cysts

Decreased or fixed intracranial volume

1. Depressed skull fracture
2. Premature fusion of cranial sutures (craniosynostosis)

Idiopathic intracranial hypertension (aka pseudotumour cerebri)

Measurement of intracranial pressure

Although some non-invasive methods of measuring ICP have been proposed (e.g. transcranial Doppler, optic nerve sheath diameter), none of these have so far made it into generally accepted clinical practice. Therefore ICP is still most commonly estimated from lumbar puncture (LP) manometry, when performed on a child lying in the lateral decubitus position in a relaxed posture.

Measuring CSF pressure in a screaming child is likely to be inaccurate because of the temporary rise caused by high venous pressure. Similarly, the person holding the child in the fetal position for the LP should relax his/her grasp and allow the child to stretch out temporarily while the pressure is being measured. If the LP is being done under a general anaesthetic, attempts should be made to normalise the pCO_2 so that the pressure reflects awake ICP.

If a lumbar puncture is performed on a child in a sitting posture, measurement of hydrostatic pressure is likely to be elevated compared to ICP. Adjustment of the sitting posture LP pressure to reflect ICP is difficult because multiple factors affect the relationship, including the height of the child, his/her posture and his/her emotional state. Nevertheless, if the child is very cooperative, pressure measurement can still be attempted by carefully laying the child on his/her side straight after the needle has entered the subarachnoid space.

Patients who have a CSF shunt in place may be able to have ICP measured by putting a needle into the reservoir chamber (see Chapter 8.1); however, this is usually performed by neurosurgical colleagues.

Particular issues in children

Infants

An infant's inability to communicate and smaller repertoire of behaviours make raised ICP more difficult to diagnose, and one needs to have a high index of suspicion as the presentation may be subtle.

The infant does not have a rigid cranial vault until fusion of the cranial sutures. This is a gradual process occurring throughout childhood; therefore ICP can cause diastasis of the cranial sutures in children. However, this is rare in children over the age of 7 years and even rarer now that CT scans and MRI allow for earlier diagnosis. Because of this flexibility, increasing intracranial contents in the infant will cause a lesser increase in the ICP and a greater increase in the head circumference than it would in older children or adults. This is one of the reasons expanding intracranial lesions have more subtle clinical manifestations in infants.

In less urgent cases the measurement of head circumference (occipitofrontal) is a useful means of detecting intracranial pathology. The head circumference should be plotted on centile charts, using prior measurements, if available, to determine if there has been a trend to cross percentiles. The child's length and weight should be plotted concurrently, to evaluate if the head is disproportionately large or small. When the cranial sutures separate due to expansion, percussion of the skull makes a sound similar to that of a 'cracked pot'. This is known as Macewen's sign.

Also, the open anterior fontanelle allows direct palpation of ICP up to the age of 9 to 18 months. It is highly recommended that one closely observes the fontanelles of normal infants, to help one identify abnormalities in clinical practice. The normal fontanelle will bulge slightly when the infant is lying down. It will become depressed when the child is sat up. It will bulge more prominently when the infant is crying or straining. The normal fontanelle will have an arterial pulsation more apparent when the infant is upright.

The fontanelle also provides access for emergency procedures such as the draining of a traumatic subdural or a ventricular tap. Fortunately the need for these procedures in the emergency department (ED) rarely arises. When it does, however, it should be done by a neurosurgical specialist. In centres where such help is not rapidly available, over-the-phone advice from a neurosurgeon may help with both the decision to do the procedure and the technique.

Clinical features of raised intracranial pressure

The symptoms and signs that lead to a diagnosis of raised ICP will vary with the age, severity and rate of development. In slower onset conditions such as brain tumours, the most common scenario in infants is the gradually progressive onset of drowsiness/lethargy, morning irritability and vomiting, with an expanding head circumference. In older children there are progressive early morning headaches but no dramatic increase in head circumference.

Brain tumours may also present with focal neurological signs before there is a significant rise in ICP due to direct invasion of neural pathways. Other symptoms may include a head tilt, which is due to unilateral 4th cranial nerve palsy causing a vertical strabismus. The child will compensate for this by tilting the head. This often occurs with posterior fossa tumours. However, there are other causes of head tilt such as sternomastoid 'tumour' in the newborn and benign torticollis. In obstructive hydrocephalus, paralysis of upward gaze is common due to third nerve dysfunction. This leads to the classic picture in the infant of a big head and 'sunset' eyes. Some infants will become irritable on watching TV or looking at books because of diplopia. Parents may notice strabismus, and older children will complain of diplopia.

Often there will be regression in motor milestones due to ataxia and/or weakness. Personality changes may occur.

As pressure increases, the pressure itself may cause focal neurological symptoms and signs. These may be due to several mechanisms, which include:

- impingement on, or disruption of, a cranial nerve, cranial nerve nucleus or higher centre
- impingement on a blood vessel supplying any of the above
- in hydrocephalus, stretching of corticospinal tract fibres around enlarging ventricles causes upper motor neuron signs in the legs
- focal seizures may accompany raised ICP and leave an infant with a Todd's paresis.

In the more rapid onset conditions, such as intracranial haemorrhage, sudden drowsiness and vomiting, with or without focal neurology and headache, are the rule. When there is a large intracerebral pressure differential across a fixed structure, such as the tentorium, the brain will herniate. There are several herniation syndromes.

The central herniation syndrome

Generalised midline or bilateral swelling above the tentorium cerebelli causes the midbrain to herniate through the tentorium. This leads to dysfunction of the midbrain and higher centres due to compression, causing:

- drowsiness
- initially *small* reactive pupils
- decorticate posturing
- as the process worsens pupils become midrange
- posture becomes decerebrate
- bilateral 6th cranial nerve palsies may become apparent when attempting the doll's-eye reflex
- these are accompanied by Cushing's triad, i.e. hypertension, bradycardia and abnormal breathing patterns (alternating tachypnoea/bradypnoea and fluctuating depth of respiration).

The lateral mass herniation syndrome

A unilateral supratentorial swelling will give rise to a pattern of events formerly known as the uncal herniation syndrome. Imaging studies now show that herniation of the uncus happens very late in the process and is not responsible for most of the signs. Features in approximate sequence are:

- contralateral hemiplegia
- drowsiness
- ipsilateral pupillary dilatation (partial 3rd nerve palsy)
- complete 3rd nerve palsy as pressure increases (can be bilateral)
- ipsilateral hemiplegia (midbrain being pushed against edge of tentorium)
- Cushing's triad.

Cerebellar tonsillar herniation syndrome

A wide variety of signs and symptoms arise from posterior fossa masses. However, sometimes as the cerebellar tonsils are pushed through the foramen magnum and the medulla with its respiratory control centres is compressed, the following sequence occurs:

- Patient complains of a stiff neck

- Drowsiness
- Nystagmoid eye movements
- Apnoea.

Other examination findings in raised intracranial pressure

General observation

In the uncooperative irritable child, general observation from a distance is a vital part of the examination. It may reveal a large and/or an asymmetric head, evidence of trauma, gait, speech or visual disturbance. Watching a playful child from a distance will yield a lot more useful neurological information than an attempted formal examination of a screaming unhappy one.

Fundi

Although often difficult to visualise in children, fundal examination is of vital importance. Using a parent as a visual distraction will often help. New ophthalmoscopes also make it easier. With patience, the disc can usually be visualised through an undilated pupil. One may be able to visualise retinal venous pulsations. The implication of these pulsations is that ICP is less than peak venous pressure. Papilloedema is less common in infants with raised ICP due to the ‘decompressing’ presence of an open anterior fontanelle. Examination of the fundi is less useful in acute severe raised ICP, as papilloedema takes days to evolve. Other retinal findings associated with raised ICP include retinal haemorrhages, which have a strong association with non-accidental injury, aneurysms and arteriovenous malformation that may be associated with similar intracranial lesions, and subhyaloid haemorrhages, which may be seen in patients with subarachnoid haemorrhages. In some instances it may be useful to ask an ophthalmologist to examine the child through dilated pupils.

Peripheral neurological signs

The development of ‘handedness’ is not apparent in normal infants under 1 year of age. If handedness is apparent in infancy, it is usually due to a neurological or musculoskeletal lesion, some of which are associated with raised ICP.

Hydrocephalus may produce lower limb signs with gait disturbance in the ambulant child. In infants, this may be apparent when the child is held up by hands around the chest with the legs suspended in mid-air. Spasm of the adductors will cause scissoring of the lower limbs.

Investigations

Imaging the brain will often reveal the underlying pathology in children with raised ICP. However, there are several drawbacks that must be borne in mind when deciding to obtain a scan.

One must be sure that it is safe to move the child to the scanner room where facilities for resuscitation and access to the patient are less than ideal. CT or MRI scans may be falsely reassuring, especially in the case of idiopathic intracranial hypertension or meningitis, where the appearance may be normal even when the pressure is dangerously high.

To keep an uncooperative child still may require a general anaesthetic, with its concomitant risks. However, as scanning technology improves, the time required for a scan is getting dramatically shorter. Many infants can often be successfully scanned with the use of a ‘dummy’ (comforter) dipped in a sucrose solution and firm wrapping. Likewise, in older children, the presence of a parent in the scanner room may permit the avoidance of anaesthesia. Otherwise the decision to use general anaesthesia or procedural sedation depends on the clinical situation, local policy and skill mix, with safety always being the first consideration.

An MRI scan is more likely to require anaesthesia in younger children, as the scans are very noisy, the child needs to be still for a relatively long time, and patient access is more difficult.

When obtaining CT scans, one must also consider the risks of radiation causing cancer, which in a CT scan of the head in a child is estimated to be 1:1000–1:10,000 over the lifetime of the child.[3,4]

Head ultrasound through the open fontanelle is an option for initial imaging in infants. This is good at imaging the lateral ventricles and surrounding structures but dependent on operator experience. However, ultrasound is less able to image the subdural space around the vertex and the posterior fossa region which makes it more of a ‘rule in’ investigation. To rule out pathology in these regions CT or MR may be necessary.

Management of raised intracranial pressure

Acute severe increases in ICP are a true emergency because a rise in ICP without a concomitant rise in MAP leads to a decrease in the CPP and eventually cerebral hypoperfusion with neuronal cell death. Therefore, children in the ED with acute severe raised ICP should be treated in the resuscitation area of the ED or taken urgently to the operating theatre. A neurosurgeon should be consulted as early as possible.

In this situation efforts should be directed towards maintaining cerebral oxygenation and perfusion by supporting the child's ventilation and circulation, if necessary using rapid sequence intubation (RSI) and mechanical ventilation. There is considerable controversy over which sedatives and muscle relaxants to use because some drugs used in RSI and the manipulation of the airway itself may cause a further transient rise in ICP.

Several strategies have been used to avoid this transient rise including administering an IV dose of lignocaine (lidocaine) just prior to RSI and giving a 'defasciculating' dose of a non-depolarising neuromuscular blocking agent just before using succinylcholine (a depolarising muscle relaxant that causes increase in ICP in some studies) or using a non-depolarising agent such as rocuronium with a reversal agent available, such as sugammadex.

With regard to sedative agents, there are a number of recommended options including propofol, thiopentone, etomidate, morphine and fentanyl. Ketamine was thought to raise ICP in these situations; however, recent evidence has contradicted this, and its use, particularly in neurotrauma, is increasing.[5]

There is little or no evidence looking at what effect the use of these various strategies has on long-term patient outcome. However, there is evidence that hypotension and hypoxia during RSI are harmful. Therefore it would seem wise to maintain an evidence-based departmental-wide strategy which is most likely to prevent complications and delays in the RSI process.

Endotracheal tubes should be secured with tape to the face of the patient and not with ties that encircle the neck as this has been shown to increase ICP. ICP can be reduced by giving intravenous mannitol and nursing the patient in a 30 degree head-up position. Mannitol should not be given if the patient has circulatory failure (i.e. hypotension or hypoperfusion) as the osmotic diuresis that it causes could exacerbate the hypoperfusion. Hypertonic saline (HS) may be a better alternative to mannitol because it increases blood pressure as well as decreasing cerbral oedema. HS is the fluid of choice in treating acute severe

raised ICP due to hyponatraemia. Just as in trauma so in medical illnesses C (circulation) comes before D (disability); meaning that a perfusing blood pressure must be maintained if there is to be any CPP.

There is strong evidence against prolonged hyperventilation because vasoconstriction impairs cerebral perfusion despite the reduction in ICP. pCO_2 should be kept between 35 and 40 mmHg unless the above measures have been unsuccessful or herniation is progressing, in which case the pCO_2 can be taken down to 25 mmHg for a brief period while further measures are commenced.

Seizures should be aggressively treated and consideration given to seizure prophylaxis in conditions that could be epileptogenic, as seizures greatly increase the cerebral metabolic demands and cause cerebral vasodilatation which will further increase ICP. Similarly, fevers should be avoided in this situation.

Whilst these measures are taking effect consideration and management of the underlying cause should be undertaken. This will be aided by a concise history and some rapid bedside testing such as blood gas, which may reveal conditions such as hyponatraemia or hypercarbia followed by urgent neuroimaging (CT scan or MRI) if the patient is stable enough.

Concurrent with this neurosurgical advice should be sought to consider definitive treatment. If an infant with an open fontanelle continues to deteriorate despite the above measures, one may perform a ventricular or subdural tap via the open fontanelle. This should be done either by the neurosurgeons or in rural and remote situations under instructions from them via video or telephone link.

In older children who continue to deteriorate, the options depend on the clinical situation and the underlying cause. Therapeutic options include treating an underlying cause urgently (e.g. high-dose dexamethasone for vasogenic oedema or hypertonic saline for hyponatraemia) or maintaining cerebral perfusion pressure by maintaining mean arterial blood pressure at above normal levels with fluids and inotropes and reducing cerebral metabolic demand with drugs such as thiopentone.

Other neurosurgical interventions include craniotomy, insertion of external ventricular drain, and insertion of a CSF reservoir or a complete CSF shunt. Where a shunt is already present this may be tapped (see Chapter 8.1 on shunt complications). Once initial stabilisation is under way and neurosurgeons are involved, referral to a paediatric ICU is mandatory.

Children with less severely raised ICP usually need investigation and treatment of the cause, usually under the care of a paediatric neurosurgeon whilst avoiding interventions that may increase ICP. One should avoid the infusion of

hypotonic fluids. Children with hydrocephalus will usually require admission for consideration of a shunt or 3rd ventriculostomy.

Some particular causes of raised intracranial pressure

Iatrogenic

Any child receiving intravenous fluids, especially for treatment of diabetic ketoacidosis (DKA), who develops headache and/or drowsiness and/or seizures during therapy should be considered to have raised ICP until proven otherwise. A thorough assessment should be made and electrolytes checked for disturbances that may lead to cerebral oedema. Imaging should be considered. If cerebral oedema is confirmed or not excluded then the child should be treated as above and observed in a paediatric intensive care unit.

Idiopathic intracranial hypertension (aka pseudotumour cerebri or benign intracranial hypertension)

This condition is characterised by sustained raised ICP causing symptoms similar to a cerebral tumour but with no anatomical abnormality on neuroimaging. It is usually due to decreased CSF re-absorption. Idiopathic intracranial hypertension (IIH) has many causes (Box 8.2.2). It is more prone to occur in overweight pubescent girls, in whom no cause is found. The presentation is usually with headaches and vomiting, which are worse in the morning. Some patients may complain of transient visual obscuration with blurring or darkening of vision that lasts less than 30 seconds. Later there may be unilateral or bilateral 6th nerve palsies causing diplopia on lateral gaze. Papilloedema is often the only positive physical finding. The most significant complication of IIH is loss of vision. This begins with an enlargement of the blind spot associated with papilloedema and later progresses with erosion of the peripheral visual fields. Left untreated the patient with persistent IIH will eventually develop optic atrophy and blindness.

Investigations include CT scan or MRI to rule out a space-occupying lesion, followed by lumbar puncture to measure opening pressure. Lumbar puncture may be both diagnostic and therapeutic. The CSF has a high pressure, but

analysis reveals normal protein, glucose, and cell count and no microorganisms.

Therapeutic CSF taps should aim to decrease the ICP by 50% under the guidance of a paediatric neurologist. This may be curative. However, in most cases repeat lumbar punctures and/or acetazolamide are required. It is believed that acetazolamide works by reducing CSF production. In severe cases a lumboperitoneal shunt or optic nerve sheath fenestration may be required. One needs to exclude any treatable underlying cause. Anticoagulants should be given for venous thrombosis, and drugs such as glucocorticoids should be ceased or weaned if possible. Referral to a paediatric neurologist is mandatory.

Box 8.2.2 Causes of pseudotumour cerebri

1. Idiopathic
2. Venous obstruction
3. Metabolic:
    - Hypervitaminosis A
    - Hypoparathyroidism
    - Addison's disease
    - Obesity
    - Pregnancy
    - Galactosaemia
4. Drugs:
    - Oral contraceptive pill
    - Glucocorticoids
    - Tetracyclines
    - Isotretinoin
    - Nalidixic acid
    - Nitrofurantoin
5. Haematologic:
    - Anaemia
    - Polycythaemia
6. Infections:
    - Roseola infantum
    - Chronic complicated otitis media
7. Others:
    - Guillain–Barré syndrome

Fever and raised intracranial pressure

Children who have clinical signs of acutely raised ICP and fevers must be treated as if they had until proven otherwise (Chapter 8.7). This includes giving dexamethasone before or at the same time as the antibiotics to diminish brain swelling and prevent further rises in ICP.

If the patient has been recently exposed to mosquitoes in malaria endemic areas antimalarials should be considered.

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8.3

Seizures and non-epileptic events

Jeremy Furyk, and Stuart Dalziel

ESSENTIALS

1 Seizures are a common life-threatening presentation to emergency departments requiring immediate management.

2 The diagnosis of a seizure is based on a careful history.

3 There are many paroxysmal events or ‘funny turns’ in children, which can mimic seizures.

4 Bedside blood glucose testing should be performed on the child who is having a seizure. First-ever seizures should have blood sent for formal glucose, electrolytes, calcium, magnesium and phosphate levels.

5 The acute management of the child having a seizure involves supporting airway and breathing and the graduated use of anticonvulsant drugs to terminate the seizure.

6 Parental reassurance and education regarding seizures, basic life support and safety issues are an important aspect of management in the child presenting with a first convulsion.

Introduction

A seizure can be defined as a disturbance in neurological function, resulting from uncontrolled excessive neuronal activity in the brain.

The subject of seizures and their differential diagnosis is large. This chapter provides only essential information that may be useful in formulating diagnosis

and treatment in the emergency department (ED). The terminology used by neurologists to describe seizure types has changed over time; excellent up-to-date resources are available online (www.epilepsydiagnosis.org) from the International League Against Epilepsy.

General comments

There are a number of conditions that must always be considered in children experiencing ‘unusual events’. Hypoglycaemia can cause both partial and generalised seizures; it may also rarely produce focal neurological signs. Abnormalities of electrolytes, calcium, magnesium and phosphate can present in a similar fashion. Syncope can be easily misdiagnosed as a seizure; both benign and potentially life-threatening cardiac causes can be associated with anoxic seizures or so-called ‘convulsive syncope’. They are usually brief and not associated with a post-ictal period. Syncope is discussed in more detail elsewhere (see Chapter 5.3). Taking a comprehensive history – particularly from eyewitnesses – is vital in obtaining the correct diagnosis.

Some variants of migraine share clinical features with epilepsy syndromes and may be difficult to distinguish from seizures. Diagnosis is often difficult or impossible to make on the first presentation and without further investigation.

Non-epileptic seizures (previously known as non-epileptic attacks, psychogenic seizures and pseudoseizures) need to be considered in the differential diagnosis of atypical epileptic seizures. Some specific clinical clues include a prominence of a waxing and waning pattern to jerking movements, variability in rate and direction of jerking movements, horizontal movement of the head, crying during or post ‘seizure’, and resistance to passive eye opening during and post ‘seizure’. Munchausen's syndrome by proxy is also a consideration, particularly in infants and young children. Again, the diagnosis is often difficult, and the ED is usually not an appropriate setting to make a confident diagnosis of either of these conditions on the first presentation.

Classification of seizures

Seizures are conceptually divided into *generalised seizures*, where the onset is from all brain regions simultaneously or rapidly involves bilateral neuronal networks, and *focal seizures*, where the seizure arises from one area in the brain. Focal seizures can occur with or without impaired consciousness or awareness.

Occurrence of a seizure does not equate with the child having epilepsy. Focal seizures may spread and become secondarily generalised.

Status epilepticus has traditionally been defined as a seizure that lasts more than 30 minutes or when there is incomplete recovery between seizures over this time period. Recently a revised operational definition of 5 minutes of continuous seizure activity has been proposed based on the time frame in which the majority of seizures would have resolved spontaneously and when one would be expected to commence treatment. In practical terms, any child who arrives in the ED still fitting should be regarded as a medical emergency. Status epilepticus can occur in both convulsive and non-convulsive forms.

Febrile seizures

Febrile convulsions are a frequent cause of children presenting to EDs with seizures. Typically they occur in children between 6 months and 6 years of age and are not regarded as a form of epilepsy. They are common, affecting around 3% of children. Most children who have febrile convulsions have only one, but 25–30% will have a recurrence. The risk of recurrence is greater in infants less than 12 months, where it may be as high as 50%. Febrile convulsions are classified as simple or complex. Classification as a complex febrile convulsion requires any of the following: a prolonged duration (>15 minutes), focal features, recurrence during the same febrile episode, or prolonged recovery post seizure (>1 hour).

Simple febrile convulsions most commonly occur in the setting of viral illnesses such as an upper respiratory tract infection, pharyngitis, gastroenteritis, or an exanthem such as roseola infantum. Much less commonly, pneumonia or a urinary tract infection may be the underlying cause. Febrile seizures typically occur relatively early in the course of an infectious illness, and sometimes the convulsion is the first sign that the child is unwell.

Most febrile seizures are generalised and brief, lasting less than a few minutes. The child returns to normal after a short (usually less than 30 minutes) post-ictal period. The seizures may be clonic, tonic–clonic or atonic where the child may simply seem to stop breathing. Assessment of the child with simple febrile convulsion should be directed at determining the cause of the fever and should be similar to the assessment of the febrile child presenting in the absence of a seizure. The possibility of central nervous system infection should always be considered, particularly in young children (<6 months) and unvaccinated

children. Most children with an uncomplicated febrile seizure who have completely recovered require no investigations, apart from those that would be otherwise indicated if they presented with fever alone.

Complex febrile seizures are a frequent cause of paediatric status epilepticus. While most recover, underlying meningitis/encephalitis must always be considered in the child who fails to return to normal, has multiple or prolonged seizures or has residual neurological signs. The threshold to perform a lumbar puncture is lower when the child is <12 months of age or has been on antibiotics, which may mask the usual signs of meningism. If there is concern regarding the safety to perform a lumbar puncture, it may be prudent to institute treatment for meningitis/encephalitis and delay the procedure until the child is stable.

Most children with a febrile seizure are able to be discharged home after a brief period of observation in the ED. Parents often require considerable reassurance and explanation of first-aid measures and the natural history of the condition (around one in three children will have at least one further febrile seizure), which should be supplemented by appropriate written advice. There is no evidence that antipyretics prevent recurrent febrile convulsions.

Presentation to Emergency Department

Children with seizures present to the ED in two broad categories:

- The child who presents after an event. With a key question being ‘was the event a seizure?’
- The child who presents while actively fitting.

Presentation post a possible seizure

History

The diagnosis of seizures is based almost entirely on the history, although many children are now presenting to the ED with video footage of their seizures taken on a carer’s smartphone.

The parents or any witnesses of the event should be taken through the episode systematically. An exact description of what took place is crucial in making a correct diagnosis. What was the child doing when it started? What was the very first sign of a problem, and what followed? If abnormal movements were present, did they principally involve one side of the body? How long did the

event last? What was the child like afterwards? How long before the child was back to normal? Was there any faecal or urinary incontinence or tongue biting? Was there any apnoea or colour change?

Witnessing a sudden loss of consciousness in a child is an extremely upsetting event for any observer, particularly the parents. Many parents report that they thought their child had ‘stopped breathing’ and commenced various resuscitation techniques. There are obvious limitations to the history, but it remains the single piece of information most likely to provide a diagnosis. It is also worth talking to the verbal child about what happened. Even young children can sometimes give very helpful accounts if spoken to soon after the event.

It is important to establish whether there may have been provocative factors, such as sleep deprivation, a febrile illness, head trauma or potential exposure to epileptogenic medications. A past history of similar events should be sought. It is also important to ask about a family history of seizures, sudden death, or unusual turns and to review the child's developmental milestones.

Examination

The physical examination is often normal, but it must be carefully performed, looking for fever, evidence of intercurrent illness and abnormal neurological signs. Other important diagnostic clues include a Todd's paralysis, skin lesions of tuberous sclerosis and neurofibromatosis, or morphological features of a chromosomal disorder or other underlying neurological abnormality associated with a seizure disorder. The finding of lateral tongue trauma or incontinence supports a suspicion of seizure when the diagnosis is unclear. The febrile child needs to have underlying meningitis or encephalitis excluded.

In infants, the sudden onset of frequent seizures and failure to regain consciousness is a common presentation of non-accidental injury and a careful examination must be performed, looking for additional signs of non-accidental injury, e.g. retinal haemorrhages and unexplained bruising.

Differential diagnosis and specific seizure syndromes

Seizure classification is complex and is well described in other resources and texts (e.g. www.epilepsydiagnosis.org).

Although there is overlap between the various paediatric age groups, it is

useful to approach the aetiology and differential diagnosis of paediatric seizures by age of presentation. For neonatal seizures refer to Chapter 3.4. There are many syndromes, the specifics of which are generally not required for the acute management. The most common are briefly reviewed here along with important seizure mimics or non-epileptic events.

Infancy

Seizure types

Familial (and non-familial) infantile epilepsy

This syndrome is characterised by frequent and intractable seizures that start between 3 and 20 months. Children have otherwise unremarkable past history and examination, development continues normally, and seizures usually resolve within a year.

West syndrome (infantile spasms)

West syndrome is characterised by epileptic ‘spasms’, usually first occurring at 3 to 12 months of age. Spasms are a sudden, brief flexion or extension of proximal and truncal muscles and can occur in clusters. Development is usually delayed, and the syndrome is associated with various structural, genetic and chromosomal disorders.

Dravet syndrome

Previously called severe myoclonic epilepsy of infancy, Dravet syndrome presents in the first year of life in a previously normal child, with a variety of different seizure types. The majority of patients have abnormalities of the sodium channel genes. Development is usually delayed from beyond 12 months of age.

Non-epileptic events of infancy

The following events can mimic seizures in young children and are usually distinguished by a careful history.

Breath-holding spells

These are always provoked by an unpleasant stimulus. This is usually minor trauma, e.g. a bump on the head, but the child being scolded or frustrated may

provoke them. Arbitrarily, breath-holding attacks are divided into the following:

1. Pallid breath-holding attacks, where after a short cry, the infant loses consciousness and becomes pale, and there may be tonic stiffening.
2. Cyanotic breath-holding attacks, where after vigorous crying, there is breath-holding in expiration and loss of consciousness.

As in syncopal episodes there may be brief clonic jerking after an episode. The story of provocation is usually clear, but at times there can be diagnostic confusion; for example, when a child who is playing happily is found unconscious on the floor and no precipitating event was observed.

Benign neonatal sleep myoclonus

This is a normal sleep occurrence characterised by myoclonic jerks, which can be quite violent and asymmetric but are confined to sleep. The infant is otherwise normal.

Jitteriness

Jitteriness is common in the newborn period; infants usually appear awake and behave normally afterwards. Newborn jitteriness is usually a transient self-limiting finding. Pathological causes include hypocalcaemia, hypoglycaemia and neonatal withdrawal syndrome. Jitteriness can often be distinguished from seizure activity in that it may increase when the infant is startled or unwrapped and decrease when the infant is swaddled or the affected limb held.

Self-stimulatory episodes

These are seen in girls. The thighs are generally clenched tight, with the legs crossed at the ankles. There are pelvic undulatory movements. The infant may look extremely flushed and upset and may be sleepy afterwards.

Stereotypies

These are repetitive movements such as hand-flapping or body rocking. These are usually repeated in exactly the same way each time. Although common in children with autism, they also occur in children who are otherwise normal.

Daydreaming

This is common at all ages in childhood. The child may stare straight ahead and

at times not respond to being called.

In daydreaming, stereotypic behaviours and self-stimulatory episodes, the event can usually be immediately stopped by physical interaction with the child, e.g. by tickling him/her.

Childhood

Seizure types

Childhood absence epilepsy

Childhood absence epilepsy usually consists of brief staring spells characterised by a sudden loss of consciousness and an abrupt end to the seizure. There is no post-ictal period. With longer absences, automatisms may occur. The events usually last less than 10 seconds, and multiple episodes may occur per day.

Panayiotopoulos syndrome

Characterised by onset in early childhood of often prolonged autonomic seizures, with pallor, nausea and vomiting, progressing to eye deviation and tonic–clonic movements. Onset is usually between 1 and 14 years, seizures are infrequent and the condition is usually self-limiting.

Childhood occipital epilepsy (Gastaut type)

This is a self-limiting epilepsy of childhood where seizures are usually well controlled with medications, and remission frequently occurs.

Lennox–Gastaut syndrome

In Lennox–Gastaut syndrome there are multiple seizure types, including atypical absences, myoclonic seizures, drop attacks, nocturnal tonic seizures and generalised tonic–clonic seizures. In the early phases, which can be seen in late infancy, the child may present with atypical absences. These have somewhat slower onset than typical absences, and there may be obscuration rather than complete loss of consciousness. Great caution should be taken with any child presenting in the first 2 years of life with ‘absences’, as this may be an early manifestation of Lennox–Gastaut syndrome.

Non-epileptic events

Night terrors

Usually occur within the first few hours of the child going to sleep. Typically the child screams and is found sitting up in bed with widely dilated pupils and appears to be extremely fearful. This may last for half an hour or more. The child is eventually comforted and goes off to sleep. The next morning the child has no memory of the event.

Nightmares

In contrast, these usually occur in the second half of the night. The child goes into the parent's bedroom afraid because of a ‘bad dream’. The child usually has a good memory of the dream, although he/she may not want to discuss it.

Long QT syndrome

Long QT syndrome may present as a seizure mimic. All first presentations of a seizure should have an electrocardiogram (ECG) performed and the QTc interval calculated.

Adolescence

Seizure types

Juvenile myoclonic epilepsy

This usually develops in the early teenage years. Typically there are myoclonic jerks in the morning soon after awakening. These may not be mentioned to the parents. The first presentation is often with an early morning generalised tonic–clonic seizure. A history of early morning myoclonus should always be sought in this situation.

Non-epileptic events

Syncope

Is usually a vasovagal response provoked by a stressful situation, such as seeing blood or standing for a prolonged period in hot weather. There is a feeling of light-headedness, nausea and then a progressive fading out of vision. As mentioned above, there may be brief stiffening and clonic jerks in the course of syncopal episodes. Unusual forms of syncope in adolescence include ‘stretch syncope’ provoked by neck hyperextension and shoulder abduction while

stretching and ‘hair grooming syncope’, which seems to be provoked by strong pulling of the hair while combing or brushing. Children with intellectual handicap may repeatedly provoke episodes of loss of consciousness by hyperventilating and then performing the Valsalva manoeuvre. See Chapter 5.3 and 5.10 long QT syndrome above.

Migraine

Migraine with aura and familial hemiplegic migraine may also present as a seizure mimic.

Hypnogogic jerks

Common at any age and are normal phenomena at sleep onset. Can be more common in children with motor and developmental disorders and can be confused for myoclonic seizures or spasms.

Rage attacks

These consist of sudden episodes of directed anger, usually provoked by being thwarted in some way. A careful history will usually differentiate a rage attack from non-directed aggression, which can occur in the post-ictal state. This usually occurs when a patient in a confused post-ictal state is restrained or surrounded by a crowd of people.

Presentation of a seizure

Convulsive status epilepticus

When a child is fitting on arrival at the ED, the seizure is likely to have continued for at least 5 minutes and fulfil the current definition of convulsive status epilepticus. The management of a child presenting in status epilepticus involves stopping the seizure, identifying the likely aetiology and supporting Airway, Breathing and Circulation. The child should be taken directly to the paediatric resuscitation area with ready access to resuscitation, airway equipment and monitoring.

The child should receive high-flow oxygen via mask or assisted by bag and mask, if ventilation is inadequate. Basic airway manoeuvres and suctioning may be required. Oxygen saturation should be monitored continuously. Intravenous (IV) access should be gained and, if not possible, intraosseous (IO) access

obtained. Immediate blood glucose testing should be undertaken to exclude reversible hypoglycaemia. Blood should be taken for glucose, electrolytes, calcium, magnesium and phosphate and full blood count. Venous blood gas analysis and point of care electrolyte testing might be useful if available. Children on certain maintenance anticonvulsants should have their drug level taken if possible and appropriate.

At the same time, a history should be rapidly obtained regarding:

- the duration of the seizure
- previous episodes (and if there is a particular drug most likely to be effective or not effective)
- precipitating events
- medications the child is taking
- medications and illicit drugs that may be in the child's environment
- whether drugs have already been given for the seizure
- the presence of underlying neurological problems.

Drug therapy

If the child has received no treatment for the seizure prior to arrival, international guidelines advocate two doses of benzodiazepines, followed by a 'second line' agent/s, followed by induction of anaesthesia if seizures continue. Management is often commenced in the pre-hospital setting by paramedics or parents, and doses administered should be incorporated into the algorithm that follows. Management with first-line benzodiazepines is supported by randomised controlled trials. Management beyond first-line benzodiazepines is supported only by non-experimental evidence and expert opinion.

First line

Initial treatment is with benzodiazepines, which are rapidly acting. IV diazepam 0.25 mg kg^{-1} or IV midazolam 0.15 mg kg^{-1} are reasonable options. Midazolam can be given by other routes (IO, intramuscularly, intranasal or buccal) if IV is not available, while diazepam can be given IO or per rectum. If the seizure has not stopped within 5 minutes the dose can be repeated. The potential side effects of benzodiazepines, respiratory depression or apnoea, need to be anticipated.

Second line

If after another 5 minutes the seizure continues, IV phenytoin 20 mg kg^{-1} as an infusion over 20 minutes (or IV phenobarbitone 20 mg kg^{-1} as an infusion over 20 minutes) should be given. These second-line agents need to be given slowly by IV infusion and therefore will take time to control the seizure. Paraldehyde per rectum is efficacious; however, it is difficult to give and is infrequently used. Alternative second-line agents such as levetiracetam, sodium valproate and lacosamide have increasing non-experimental evidence supporting their use and are currently being evaluated in clinical trials. If at any stage continued seizure activity is causing respiratory compromise, or it is felt that an unprotected airway may result in brain injury, it may be necessary to escalate care.

Third line

If there has been no response after second-line agents, induction of anaesthesia using rapid sequence intubation is the next step. The decision to intubate to control a seizure needs to involve consideration not only of the length and severity of the seizure but also the degree of respiratory embarrassment to the child. Mild seizure activity that is not compromising a child can be tolerated longer than a major convulsion that is impairing oxygenation. These children will need to be subsequently managed in a paediatric intensive care unit. There is no consensus on the optimum induction agent in this situation, and clinicians must be cognizant that muscle relaxation will remove clinical monitoring of ongoing seizure activity or recurrence.

In individual patients with frequent bouts of status epilepticus along with other neurological disabilities, where there is a desire to avoid intubation, alternatives to the above general approach may be appropriate.

Absence, atypical absence and complex partial status are much less common than convulsive status but should be considered in children who are obtunded or have bizarre behaviour without obvious cause. If there is doubt an electroencephalogram (EEG) should be performed, although this is not routinely available in most EDs. If an EEG is unavailable, ‘waking up’ soon after an IV dose of a benzodiazepine is a useful diagnostic clue of non-convulsive status.

Investigations

These depend on the clinical setting, but some general rules apply.

Blood tests

All children having a seizure should have a bedside glucose testing followed by a formal blood glucose to exclude hypoglycaemia. At the same time full blood count, electrolytes, calcium, magnesium and phosphate levels should be checked in order to exclude a metabolic cause. These are exceedingly rare beyond the infant age group in the child who has returned to normal following a seizure and the yield is negligible, unless there are other clues to suggest an electrolyte disturbance. Blood gas analysis and point of care electrolyte analysis may imply either cause (such as hyponatraemia) or complications associated with ongoing seizure activity (such as acidosis and inadequate ventilation), which may change immediate management.

Neuroimaging

As a general rule, patients with focal seizure or examination findings, features suggesting raised intracranial pressure, or seizures in the setting of trauma should have urgent neuroimaging. The CT scan of the brain has the advantage of relative ease of access, but it should be remembered that the MRI scan is a better test in almost all instances except traumatic brain injury.

EEG

An urgent EEG is very useful in cases where convulsive status epilepticus has been treated but the patient remains unconscious and there is a suspicion of continuing non-convulsive status. It is also helpful in the child presenting with an altered state of consciousness and absence or when focal status epilepticus with impaired consciousness is suspected.

ECG

For patients presenting with their first generalised afebrile convulsion an ECG should be performed and the QTc interval calculated, to screen for long QT syndrome. This investigation is particularly relevant in families with a history of sudden death in children and young adults.

Disposition

Children who have prolonged or multiple seizures, focal seizures, or abnormal neurological examination or where the diagnosis is unclear should be admitted for observation and paediatric or neurological review. Children who are having

increased frequency of seizures or are known to have prolonged or clusters of seizures warrant admission and observation until stabilised.

Not all children presenting with an uncomplicated first seizure need to be admitted to hospital. Those who have a first generalised seizure, with normal neurological examination and initial investigations, can be followed up on an outpatient basis. Children who present with their first afebrile seizure should have an EEG prior to follow-up in outpatients if possible. Children presenting with a febrile convulsion do not require an outpatient EEG.

Long-term anticonvulsant management is usually not indicated after the first presentation with an afebrile seizure. Risk of recurrence is 30%, increasing considerably following a second afebrile seizure.

First-aid measures should be discussed with the parents so that they have a clear plan should their child have a further seizure at home. The child should be placed on the floor away from objects that may cause harm; forceful attempts should not be made to open the child's mouth; phone for an ambulance if the seizure lasts longer than a few minutes or if concerned at any stage; and when post-ictal, place in the left lateral recovery position.

The parents should be warned that further seizures can occur. It is important to take care that the child is not in a potentially dangerous situation should this happen. Risky activities to be avoided include being unsupervised around water (both swimming and bathing); bike riding in heavy traffic; and climbing to heights of greater than a metre. This safety message should be reinforced at follow-up.

Children who are discharged after a febrile convulsion should be reviewed by their local doctor within 24 hours to follow progress and reinforce the safety issues.

Controversies and Future Directions

1. Newer second-line agents are currently being evaluated in large clinical trials internationally. Results are very likely to influence practice.
2. Given the efficacy and safety of midazolam by the IM or IN route and the increasing use of the IO route, many clinicians argue that there is no place for rectal diazepam, given its unpredictable absorption by this route.
3. Further research may evaluate various strategies for induction of anaesthesia, e.g. optimal timing and agents to use.

4. Utility of real-time video and EEG assessment in the ED may lead to advances in diagnosis and management.

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Further reading

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8.4

Acute weakness

Richard Lennon

ESSENTIALS

1 The fact that a child has acute weakness may be obscured by an inability to communicate and a smaller repertoire of activities. The possibility of a neuromuscular problem must be consciously considered by the clinician.

2 Conversely, a small child may not move a limb because of pain rather than weakness. A history and examination for evidence of trauma should be performed and appropriate diagnostic imaging considered.

3 Lower limb weakness in small children may resemble ataxia.

4 Respiratory insufficiency due to weakness may occur with surprising rapidity and may be compounded by aspiration. Careful assessment and close monitoring for this complication must be made.

5 Abnormal reflexes are important in localising the cause of acute weakness. If some are *absent* especially distally it suggests a lower motor neuron dysfunction. If all are *present* it suggests muscle dysfunction, and if they are *increased* it suggests an upper motor neuron weakness.

6 The presence of a sensory level indicates a spinal cord lesion. One should consider investigating this with an urgent MRI to determine whether or not urgent surgery is needed.

Introduction

The acutely weak child is challenging to the emergency physician because she/he can present in many different ways with many different diagnoses. In particular, the preverbal child may present with only regression in motor milestones or bruising on the face from increased frequency of falls. The diagnosis in such children can range from a tick bite to inherited neuromuscular disorders.

Presentation

In infants of less than 3 months, parents are usually quite sensitive to abnormalities of the infant's behaviour. They may bring a baby in complaining of poor feeding, decreased activity or being 'floppy'. The older infant may be noted to lose milestones such as ceasing to crawl or walk. 'Ataxia' may be the predominant feature in preschool and school-age children. Sometimes children may present with an associated feature of illness, such as the rash of dermatomyositis or an obvious engorged tick. In general, the older the child is, the more typical the presentation. However, even in this group, as in adults, there is a wide range of presentations, e.g. cranial nerve palsies in the Miller–Fisher variant of Guillain–Barré syndrome (GBS).

Trauma masquerading as weakness

Trauma is dealt with in detail in other chapters of the book. However, it should be noted that non-accidental injury (NAI) may present as 'acute weakness' in the infant and young child. 'Shaken baby syndrome' may present as a lethargic irritable child with little or no bruising. The apparent focal weakness may be due to an underlying fracture. NAI should be considered in the differential diagnosis of the acutely weak child (see Chapter 18.2).

Primary survey approach

General inspection

This is done by looking for both cause and effect of the weakness. An abnormality of the primary survey severe enough to cause generalised weakness (e.g. respiratory distress) is usually the obvious presenting problem. It is

important in the ‘weak’, floppy infant to consider the possibility of intussusception or other intraabdominal pathology. For example, in an infant with intussusception, prostration may mimic severe ‘weakness’, and lethargy may be the only presenting sign.

Exposure in the primary survey of the weak child includes a thorough search for ticks (don’t forget to look in deep skin folds), venomous bites and stings, and rashes, such as those caused by enteroviruses. Enteroviral rashes are usually scattered maculopapular but may be petechial or a hand foot and mouth rash. A close inspection should be made for bruises and other suggestive signs of NAI. A quick assessment of the child’s nutritional state and the appropriateness of weight for age should be estimated. The size of the child’s head and its proportion to the body should be noted, and centiles should be plotted later in the detailed examination phase.

ABC

The effects of weakness may include airway compromise from bulbar palsy (e.g. hoarse voice, stridor, and aspiration of secretions). Severely impaired ventilation will be clinically apparent; however, mild or moderate impairment may be subtle and rapidly progress to cause respiratory embarrassment. Therefore respiratory function tests should be serially performed on all children presenting with acute neuromuscular weakness who are able to cooperate with the test (i.e. developmental age of about 7 years). Arterial or venous blood gases should also be considered in this assessment. (see Box 8.4.1 for predictors of needing intubation and mechanical ventilation.) Circulatory defects may arise from disturbance of the autonomic nervous system (dysautonomia). This is characterised by labile blood pressure, heart rate and postural hypotension.

DEFG

The neurological part of the primary survey can give early hints as to the site of the lesion(s) involved. The classic description of peripheral neuromuscular weakness is an alert but anxious-looking child with paucity of limb movements, although if respiratory failure or facial weakness has intervened this appearance will not be present. Conversely, intracranial causes of weakness are usually associated with some degree of obtundation.

The weak infant classically lies in a frog leg posture with all limbs lying on

the bed, abducted at the hips and flexed at the knees. Asymmetry may be apparent in those with a focal weakness. With central causes of weakness (e.g. acute hydrocephalus) extensor posturing may be seen, with scissoring adduction of lower limbs. Autonomic dysfunction may lead to associated fever or hypothermia. One should never forget the bedside glucose test in the acutely weak child because hypoglycaemia in a child can present as weakness and in rare circumstances even focal weakness.

Once the primary survey has been completed and any abnormalities corrected, then a detailed history and examination should be performed.

History

If, on questioning, the weakness has been chronic then the list of possible diagnoses is extensive and is beyond the scope of this text. Such children should be stabilised and referred to the appropriate paediatric service for diagnosis. However, it is possible that a child with chronic weakness could undergo an acute deterioration, such as influenza in a child with a Duchenne muscular dystrophy. In infants the distinction between acute and chronic weakness is less relevant. In those children with conditions covered by this chapter, the history needs to focus initially on finding treatable causes. Detailed enquiry should be made about any possible tick bites or other venomous bites or stings. Enquiry should also be made about the availability of various medications and poisons around the house.

Box 8.4.1 Predictors of the necessity for intensive care unit/high dependency unit admission

Bulbar palsy
Vital capacity <20 mL kg^{-1}
>30% reduction in vital capacity from baseline
Flaccid quadriparesis
Rapidly progressive weakness
Autonomic cardiovascular instability

A precise time course for the illness should be obtained, along with the pattern of evolution of the weakness. Whether it is *ascending* from lower limbs up,

whether it is *lateralised* and whether it has *progressed* or not. A sudden onset may suggest a vascular or epileptic event. Rapid onset weakness may follow an intoxication or envenomation. Other causes are more subacute and may have progressed over weeks. The family history should be reviewed and whether there is consanguinity. A history of recent infectious illnesses may be relevant. Immunisation history regarding polio vaccination is important, and diphtheria is still an occasional cause of weakness in third world countries. Any overseas travel should be noted. Symptoms of pain in the limbs should be elicited (myositis, rhabdomyolysis). History of recent anaesthetic or medication use should be obtained.

Examination

A detailed but focused examination is then performed that attempts to establish the level and the nature of the lesion. The following lists the important distinguishing pathological features of the different levels of the neuromuscular system which may present with weakness:

1. Muscle:
    - Usually proximal more than distal weakness
    - Reflexes preserved until very late
    - May have tender muscles
    - Often have a positive Gower's sign (see below).
2. Neuromuscular junction:
    - Fatigability
    - Reflexes are preserved if the muscle is not fatigued or the disease is not severe.
3. Lower motor neuron weakness:
    - More distal than proximal weakness (although about 15% of children with GBS present with proximal weakness)
    - Reflexes are lost early in the illness.
4. Upper motor neuron weakness:
    - Apart from an initial flaccid phase, tone and reflexes are usually increased
    - Spinal cord lesions are usually associated with a sensory level
    - Intracranial problems are associated with features of encephalopathy, decreased level of consciousness, speech

dysfunction, ataxia, and bulbar dysfunction. Fundoscopy may reveal papilloedema.

It may be difficult to perform a formal neurological examination in an uncooperative infant or small child. Often in these situations, one can get a lot of information just by watching the child play. Gait examination is important including testing for Gower's sign. The child is laid on his/her back on a firm surface and encouraged to stand. A child with proximal muscle weakness will not be able to sit up but will roll on to his/her abdomen, get up on all fours and then 'climb up his/her legs' using his/her hands.

Cranial nerve palsy can often be established just by careful observation of eye and facial movements. Palpation of muscles reveals tenderness in acute myositis.

Examination of reflexes is important. Upper motor lesions will usually have increased reflexes with increased tone, although immediately after a spinal cord insult there may be a flaccid paralysis below the level of the lesion with absence of reflexes. In transverse myelitis, there may be patchy combinations of upper and lower motor neuron signs. GBS is associated with absent reflexes, although early in the disease process the reflexes may be preserved. In acute myositis reflexes are preserved; however, very late in myopathic weakness distal reflexes may be lost.

Investigations

Laboratory

Tests that may be useful in the acutely weak child include:

- *a full blood count*, which may reveal anaemia, nutritional disorders, leukaemia, or signs of infection
- abnormalities of electrolytes especially potassium, calcium, phosphate and sodium may cause weakness
- blood glucose
- urea and creatinine – renal failure may present as generalised weakness but more likely cause an abnormality of electrolytes
- creatine kinase is often raised in muscular causes of acute weakness
- endocrine function tests – especially thyroid and adrenal (see below)
- lumbar puncture (see below)
- electrophysiological studies (electromyogram [EMG], nerve conduction

studies).

Imaging

- Urgent MRI of spine (where progressive spinal cord lesion is suspected)
- CT or MRI of brain (where central cause is suspected)
- Chest X-ray (e.g. for suspected aspiration).

Specific conditions causing acute weakness

Though not exhaustive, the following sections give some detail on the more common causes of acute weakness seen in children and also the rarer ones that must be diagnosed and treated emergently.

Guillain–Barré syndrome

Introduction

GBS is an acute polyradiculopathy. It is now considered a group of diseases with two main pathological mechanisms: demyelination and axonal degeneration. It is uncommon in the paediatric population estimated at less than 1/100,000 person years. It is primarily a lower motor neuron disease affecting the myelin sheath with variable damage to axons. It is an immune-mediated reaction, usually to a recent infection. Antibodies to various gangliosides are found in the serum of patients with GBS, and matching antigens are often found in the preceding infectious agent. Infections known to be associated with GBS include *Campylobacter*, *Mycoplasma*, Epstein–Barr virus, Coxsackie viruses, influenza viruses, echoviruses and cytomegalovirus. The greater the axonal damage, the longer and the less complete is the recovery.

History and examination

Children usually present with weakness, falls, regression of motor milestones or ataxia. They also complain of muscle pain in the early part of the illness. Cranial nerves are involved in 40–50% of cases, with the facial nerve most commonly involved. The Miller–Fisher variant presents with oculomotor palsies, ataxia and areflexia.

On questioning, the parents often give a history of a generalised viral,

respiratory or gastroenteritis illness in the preceding 2 weeks. In the early part of the illness the child may have paraesthesiae. Classically, the paralysis is ascending and symmetrical. The majority present with mostly distal weakness; however, about 15% have extensive proximal muscular involvement.

Papilloedema is rare but may occur in GBS and is associated with raised intracranial pressure. Paralysis of the respiratory muscles is common and must be monitored carefully. Sympathetic nervous system involvement can produce profuse sweating, hypertension, postural hypotension and disturbances of sphincter function. Fatal cardiac arrhythmias have been reported in association with these signs. Although primarily a motor problem, sensory disturbance does occur, especially impairment of position sense. As mentioned above, reflexes are usually absent though increased reflexes and extensor plantar responses are occasionally found in the early phase of the illness.

The weakness may evolve rapidly within hours. However, it usually takes 1–2 weeks to reach the maximal weakness. Then, in the 2nd to 4th week of the illness, recovery is apparent and most children have recovered by 2 months, although some take as long as 18 months. Rarely, GBS will present in the newborn and is known as congenital GBS. They present as floppy babies that are areflexic and have elevated cerebrospinal fluid (CSF) protein.

Laboratory findings

An isolated elevation in CSF protein is the characteristic clinical finding of GBS. The CSF cell count is usually normal, although 5% of children may have a pleocystosis of 100 or so cells. Approximately 10% of children will have a normal CSF protein. The protein may be raised if a lumbar puncture is done later in the illness as it rises to a maximum after 4–5 weeks. The nerve conduction studies characteristically show increased latency and conduction block.

Differential diagnosis

As laboratory tests may be normal in early GBS and signs may be variable, it is important to exclude other causes of acute weakness (Table 8.4.1).

Treatment

The main role of the emergency physician in the treatment of GBS is in monitoring, prevention and treatment of cardiovascular and respiratory complications. In addition to monitoring of vital signs and cardiac rhythm,

frequent examinations and (if possible) bedside lung function tests should be performed. As noted above, progression to respiratory failure can be surprisingly rapid, and the indicators for intubation listed in Box 8.4.2 should be actively sought to avoid respiratory arrest.

The other less emergent treatments of GBS are either intravenous immunoglobulin or plasma exchange. This is indicated if the child cannot walk unaided, has bulbar palsy, has rapidly progressive weakness, or has worsening respiratory status. The immunoglobulin dose is usually 2 g kg^{-1} given in divided doses with variable regimes.

Prognosis

Prognosis is usually good in those treated and supported appropriately. Most children reach maximum weakness in 2 to 4 weeks then remain stable for 1–3 weeks and then recover fully over a period of time that varies from 6 weeks to many months. Approximately 85% of children recover completely. Mortality is 2% to 4% and is due to cardiovascular and respiratory complications.

Table 8.4.1

Differential diagnosis of Guillain–Barré syndrome

Diagnosis	Features/action to exclude
Puffer fish, shellfish and blue ringed octopus poisoning, ciguatera	History of ingestion or bite often a descending paralysis
Tick paralysis	Thorough examination of hair/skin creases
Snake envenomation	History; check for bite site; coagulation screen, creatine kinase
Spinal cord lesion	Look for upper motor neuron/mixed features and a sensory level; if suspected perform MRI
Periodic paralysis	Usually a sudden or very rapid onset, reflexes are diminished but preserved; check family history; measure serum potassium; do ECG
Infant botulism	Almost exclusively in infants; ask for history of eating honey; culture stools for *Clostridium botulinum* and test for botulinum toxin
Poisoning (e.g. organophosphate, lead)	History of exposure; toxidrome; check levels where suspected
Myasthenia gravis	Often cranial nerves involved; fatigability; look for antibodies; perform Tensilon test
Vasculitis (e.g. polyarteritis nodosa)	Check urinalysis; look for autoantibodies if suspected
Myositis (e.g. dermatomyositis)	Reflexes preserved; no ophthalmoplegia; look for characteristic rash; check creatine kinase; EMG
Poliomyelitis, diphtheria, other enteroviruses	Usually has fever and sore throat in diphtheria; ask about immunisations; no sensory changes

Box 8.4.2 Predictors of the necessity for intubation and ventilation in Guillain–Barré syndrome

Vital capacity ≤20 mL kg^{-1}
Maximum inspiratory pressure ≤30 cm H_2O
Maximum expiratory pressure ≤40 cm H_2O
Tidal volume <5 mL kg^{-1}
A sustained increase of pCO_2 to ≥50 mmHg
An increasing respiratory rate
Increasing oxygen requirement
An increased use of accessory muscles and paradoxical diaphragm movements; these reflect restrictive lung-chest wall movement and low lung volumes

Disposition

Children with suspected GBS should be admitted and closely observed for the evolution of severe weakness that can occur. The findings outlined in Box 8.4.1 will assist in deciding who should go to an intensive care unit (ICU)/high dependency unit and who should go to the general ward. Children with any one of these features should be monitored in a paediatric ICU.

Tick paralysis

Ticks that can cause paralysis are found throughout the world. In Australia *Ixodes holocyclus* is the main paralysing tick; it is found on the east coast. Other ticks have been known to cause paralysis, but they are rare. The toxin previously called holocyclotoxin is now thought to be several toxins. The toxins inhibit the release of acetylcholine from motor end plates. The paralysis usually occurs 5 to 7 days after the tick attaches. The paralysis can resemble GBS in the form of an ascending paralysis and is an important differential diagnosis because removal of the tick is necessary for recovery.

The envenomation most frequently presents with cranial nerve palsies. Although the tick is often located in the vicinity of the palsy, this is not always the case, and in all cases a thorough examination should be made for multiple

ticks. The paralysis can be severe and require ventilation.

Australian tick paralysis may worsen in the 48 hours following tick removal, and children should be monitored carefully during this time. Conversely, with children presenting with paralysis one should ask if a tick was removed in the last 48 hours.

Deaths have usually been due to respiratory paralysis; however, there have been reports of myocarditis and autonomic effects on the heart.

Treatment includes supportive therapy then careful removal of the tick(s) ensuring that the mouthparts are removed along with the body. This can be made easier by applying a pyrethrum spray, which suffocates the creature and makes it loosen its grip. Squeezing the tick is not recommended as this may inject more toxin. Human tick antivenom is no longer available. The treatment of tick paralysis is supportive; this may well include intubation and ventilation for several days after tick removal.

Other envenomations

Snake and spider bites are discussed in detail elsewhere in the text (see Chapter 22.1). Usually the history and accompanying symptoms will give the diagnosis. Death adder envenomation is usually a purely neurological presentation, so in the weak non-verbal child a thorough look for a bite site is indicated.

Botulism

Poisoning with botulinum toxin can present in three ways:

1. Infant botulism
2. Food-borne botulism
3. Wound botulism.

Infant botulism

Although rare, several cases of infant botulism have been reported in Australia in the last few years. Infant botulism is caused by release of botulinum toxin into the bloodstream from *Clostridium botulinum* bacteria colonising the intestines. Part of the toxin enters the terminal bouton of cholinergic motor nerves and enzymatically disables the mechanism by which acetylcholine-containing vesicles attach to the cell membrane. Risk factors for babies contracting this disease include exposure to honey in the first 6 months (honey is not

recommended for babies under 1 year), decreased frequency of stooling and lack of breast-feeding.

Diagnosis of this condition is characterised by:

- age group – the infants are almost always less than 6 months; however, there are reports in infants up to 1 year and older children with abnormal bowel anatomy
- nature of onset – the infants always start with bulbar palsies because that is where the blood supply is greatest; this is a descending paralysis
- fatigability but lack of reversibility with edrophonium or neostigmine
- absence of fever
- absence of an altered mental status
- absence of sensory defects
- normal CSF.

Clostridium botulinum may be cultured from the stools, and the toxin may be found in stools or serum through polymerase chain reaction or enzyme-linked immunosorbent assay tests. Mouse bioassay is also available where sterilised stools are injected into a mouse to see if it becomes paralysed. If suspicion is high, foods in the child's residence can be tested for *C. botulinum.* The EMG is characteristic.

The difficulty is in early diagnosis. Constipation is common. A weak suck is often put down to generalised illness, and it is often only the mother who notices the lack of expression on the infant's face. Dilated sluggishly reactive pupils may be found. A high index of suspicion needs to be maintained.

Treatment is by supportive care and administration of botulinum antitoxin. Antibiotics are not recommended unless there is secondary infection (e.g. pneumonia). This is because they may increase toxin release when the bacteria lyse and divulge their contents. Tube feeding is recommended as it restores peristalsis, which is essential for clearing *C. botulinum* from the gut. If antibiotics are used, aminoglycosides should be avoided because these worsen the paralysis. About half of the patients end up needing intubation and ventilation. Hospital stay is an average of 1 month but varies widely. Infants seem to recover completely.

Botulism immune globulin (BabyBIG), a human-derived hyperimmune globulin, which in a recent randomised controlled trial reduced hospital stay from 6.6 weeks to 2.6 weeks, is now available in Australia from the United

States on a case-by-case basis through the Therapeutic Goods Administration's Special Access Scheme. There is also an equine-derived antitoxin, a small amount of which is held in Australia in the Commonwealth Serum Laboratories. This is not effective in infant botulism.

Food-borne botulism

This is due to consumption of food in which there is preformed toxin. It is associated with home-canned foods. It differs from infant botulism in that it can occur in any age group and one-third of cases have gastroenteritis-like symptoms. The illness usually begins about 18 to 36 hours after ingestion, but onset can range from 2 hours to 8 days. Presentation is similar to infant botulism but more obvious because of the patients' greater age. The treatment is similar.

Wound botulism

This is perhaps the rarest form. The differences from the other forms are the presence of a wound, which may be obviously infected, and the presence of fever. The incubation period is 4–14 days. This requires aggressive treatment with antibiotics and antitoxin.

Spinal cord lesions

These are usually distinguished from peripheral nerve disease by upper motor neuron signs. However, in many cases initially the reflexes and tone are diminished (e.g. 'spinal shock' after trauma). In other cases, there may be patchy upper and lower motor neuron involvement such as in transverse myelitis. Therefore spinal cord lesions need to be considered in the differential diagnosis of the weak child. The key to spinal cord lesions is the presence of a sensory level. However, in transverse myelitis and in a preverbal child this may be difficult to establish. If a spinal cord lesion is suspected, the investigation of choice is an urgent MRI. This is time-critical as a space-occupying lesion in the narrow canal can rapidly cause permanent damage to the surrounding cord.

Transverse myelitis

The aetiology of this acute spinal cord inflammation is still uncertain. Hypotheses include microbial antigen cross-reaction with neural elements, bacterial superantigen inflammation and direct microbial invasion. Rarely, it is

associated with systemic diseases such as systemic lupus erythematosus and multiple sclerosis. It is likely that transverse myelitis (TM) will be found to be several diseases, and perhaps treatment will need to be tailored to the specific aetiology.

This disease usually has a rapid onset of predominantly lower limb weakness and altered sensation. Neck stiffness and fever are present early in most cases along with low back pain or abdominal pain. The sensory level is usually around the mid-thoracic region below which pain, light touch and temperature sensation are impaired. However, joint position and vibration sense are more preserved. Bladder and bowel disturbance is common, although this may be difficult to determine in a child in nappies. Tone is usually flaccid early in the illness with decreased reflexes, followed by increased tone and hyperreflexia as the disease progresses to its peak over the next 2–3 days. Sixty percent of patients recover fully over weeks to months.

Urgent MRI usually shows fusiform oedema around the site of the sensory level. CSF shows a moderate lymphocytosis and mildly raised protein.

Treatment is controversial, with case-control studies indicating a benefit with glucocorticoids, whereas prospective trials show no benefit. Treatment decisions should be made in consultation with a paediatric neurologist.

Spinal cord trauma

This is discussed in Chapter 4.3.

Spinal cord space-occupying lesions

These include epidural abscess, tumours, syringomyelia and arteriovenous malformations. Since syringomyelia is almost always a chronic condition it will not be dealt with here.

Tumours

These may be intramedullary (e.g. low-grade astrocytoma), extramedullary intradural (e.g. meningioma) and extradural (e.g. lymphoma). They usually have a more gradual onset than the other diseases mentioned in this chapter, but the early signs may be missed, and the child may present when signs are rapidly evolving due to high intramedullary pressures. Progressive gait and bladder disturbance with back pain and absence of fever are characteristic signs. Recent onset of scoliosis may be a feature. There may be a mixture of signs with upper

motor neuron signs in the lower limbs and lower motor neuron signs in the upper limbs.

Urgent MRI is the investigation of choice, followed by consultation with surgeons and/or radiation oncologists. If MRI is unavailable, CT or plain X-ray may show abnormalities. However, these should only be done if they do not delay transfer to an MRI capable centre.

Arteriovenous malformations

These usually occur in the thoracic region. They may cause symptoms as a space-occupying lesion causing compression or by stealing circulation from the nearby cord. The history is usually subacute, unless a haemorrhage or ischaemia has intervened. Clues to the diagnosis on examination include a cutaneous angioma over the region and a bruit on auscultation. MRI/MR angiogram will give the diagnosis, but angiography is usually required to define the lesion fully and to decide on whether to treat with surgery or embolisation.

Epidural abscess

This is a rare disease in children, who usually present with back pain and rigidity, fever, leucocytosis and a raised erythrocyte sedimentation rate. These symptoms may be followed by spinal neurological signs. MRI is the investigation of choice, but there is controversy over whether treatment should be by surgery or antibiotics alone.

Tethered cord/diastematomyelia

These are two conditions that tend to fix the cord of the child, which has to ‘move up’ the canal as the child grows. They rarely present with acute weakness, although there may be sudden exacerbations on flexion and extension. Examination over the spine may reveal sentinel lesions such as a tuft of hair, a sacral pit, a lipoma, or a cutaneous haemangioma.

Myasthenia gravis

This condition, which is usually due to acetylcholine receptor autoantibodies, leads to easy and rapid fatigability of muscles. This may occur in children in three forms.

Transient myasthenia of newborns

This occurs where the mother has myasthenia. Maternal antibodies to the acetylcholine receptor (AChR) cross the placenta, and the baby is born with fatigable muscle weakness. These babies may show very little motor activity for days after birth. Alternatively, they may just have feeding difficulty that worsens during the day. This illness improves within weeks as the maternal antibody levels diminish in the child's blood. The only treatment is supportive by tube feeding and intubation and ventilation, if severe. The babies recover completely and have no increased risk of myasthenia gravis (MG).

Congenital myasthenia

This is a lasting condition due to genetically inherited abnormalities in the ACh receptor. It is not due to antibodies to the AChR.

Acquired myasthenia

The incidence of this is low, though not insignificant. In an Italian study the annual incidence was 3.3 per million children <15 years– about 1% of total MG cases. The presentation is usually with ptosis, ophthalmoplegia or bulbar weakness. The clue on history is the worsening of symptoms as the day progresses due to fatigue. Peripheral muscles, especially limb girdle and hand muscles, are also involved. Reflexes are preserved but diminished. Clinical tests for fatigability such as getting the child to look up for 60–90 seconds and watching for ptosis or to flap one arm 'like a bird' for a minute and then comparing its strength with the other arm are very useful.

Diagnosis can be made with three tests. First, an anticholinergic drug may be given. Edrophonium, the usual drug used in adults may cause cardiac arrhythmias in small children, so neostigmine is preferred. Atropine is given beforehand to block the muscarinic effects. These drugs should abolish the fatigability. Second, the EMG is characteristic and usually obviates the need for muscle biopsy. Third, AChR antibodies can be measured in the blood. Early diagnosis is important because, if untreated, this disease will often progress to life-threatening severity.

Associations with MG commonly found in adults, such as thymoma, are rare in children. Differential diagnosis includes botulism, chronic low-grade organophosphate toxicity and tick paralysis.

In the ED the child may present as an initial episode or because of a crisis of

weakness. These crises may be myasthenic, due to exacerbation of the underlying condition or cholinergic due to excessive anticholinesterase treatment, which leads to overstimulation and exhaustion of receptors. Classically, a cholinergic crisis has the cholinergic toxidrome features of hypersalivation, pulmonary oedema and muscle fasciculation. However, in someone with myasthenia the cholinergic crisis may only be manifest by weakness. Distinguishing between a myasthenic and cholinergic crisis may be difficult and early consultation with a paediatric neurologist, especially one who knows the child well, is recommended. History may give a clue if medications have been missed or an overdose of pyridostigmine has been taken. A therapeutic trial of a cholinergic agent may help but should not be undertaken if there is significant risk of a cholinergic crisis. In the latter case supportive treatment and measurement of blood cholinesterase activity may be the only option.

Many drugs exacerbate weakness in myasthenia gravis including common antibiotics such as erythromycin – a careful drug history should be obtained.

Long-term treatment of MG comprises anticholinesterases and a variety of immunosuppression, IgG infusion or thymectomy.

In MG:

- Do not give neuromuscular blocking agents (e.g. suxamethonium, vecuronium) as these may paralyse the MG patient for days or even weeks.
- Do not give aminoglycoside antibiotics as these exacerbate the weakness.

Poliomyelitis and other enteroviral infections

Poliomyelitis is now exceedingly rare; however, one should always ask about immunisation status in the acutely weak child. If the child is not immunised one should ask about contact with infants recently immunised with Sabin (oral weakened live poliovirus) vaccine. Infants excrete the virus after the immunisation and this is where most recent cases of poliomyelitis have come from. The other source of infection is in developing nations, where immunisation rates may be low.

The illness itself usually presents with symptoms related to the virus’s portal of entry through the gut and upper respiratory tract. Patients have fever, sore

throat, anorexia, nausea, vomiting, generalised non-specific abdominal pain, malaise and headache. The great majority of poliovirus infections end here or are asymptomatic. In those patients who progress there is often an asymptomatic period of 1–2 days followed by symptoms of aseptic meningitis with neck and often entire spine stiffness. There may be mild transient neurological deficits such as bladder paralysis and loss of abdominal and anal reflexes. Those who progress to paralytic polio will usually do so 8–12 hours after the superficial reflexes are lost. Poliovirus can infect and destroy neurons from the motor cortex down to the anterior horn cells. However, most commonly the paralytic form presents with patchy asymmetrical lower motor neuron weakness. Bulbar weakness is also a frequent presentation, and often the picture is mixed. Rarely there are also encephalitic and ataxic presentations.

Diagnosis is based on the immunisation history, the clinical picture and a lumbar puncture showing a moderate pleocytosis, initially of neutrophils but then changing to monocytes. Serology and culture of stool, throat swab and rarely CSF will often reveal the organism. Treatment is supportive only. Infection control authorities need to be informed.

Bell’s palsy

A sudden or rapid onset of a unilateral, lower motor-neuron paralysis is not a rare occurrence in children. Estimated annual incidence varies from 3 to 10 per 100,000 children per year. Its aetiology is uncertain. The disease commonly begins 2 weeks after an infectious illness, which suggests a post-infectious autoimmune or allergic aetiology. Lyme disease has been associated, and serology should be done if the patient has been in an endemic area. Epstein–Barr, mumps and herpes simplex viruses have also been associated with this disease. An association with hypertension has suggested aetiology related to pressure necrosis of the nerve due to swelling in the narrow facial canal.

The patient often presents with pain around the ipsilateral ear and may also complain of abnormal hearing. About half have loss of taste sensation to the anterior two-thirds of the tongue, and there may be hemifacial ‘dysaesthesia’ due to the proprioceptive fibres to the facial muscles in the facial nerve.

The differential is extensive, but the diagnosis can be determined by a thorough history and clinical examination. One should look for evidence of trauma (be aware of non-accidental injury), central nervous system dysfunction, aural vesicular lesions (e.g. the Ramsay Hunt syndrome), other cranial nerve

dysfunctions, hypertension and GBS. Acute lymphoblastic leukaemia and even sarcoidosis have been reported.

Treatment is controversial, as the prognosis in children is better than in adults. Complete recovery occurs in 60% to 80% of patients, with near complete recovery in the remainder. In one study average time to recovery was about 7 weeks with a range of 9 days to 7 months. Uncontrolled studies have claimed a benefit for early corticosteroid therapy. However, controlled studies in adults have shown a benefit for oral glucocorticoids in Bell's palsy. This and the less reliable evidence in children prompt this author to recommend glucocorticoids in all but the mildest cases of Bell's palsy. There is even less evidence for antiviral agents in the absence of apparent viral infection (e.g. the Ramsay Hunt syndrome). If the eyelid does not completely close steps should be taken to protect the cornea from exposure keratopathy, i.e. artificial tears and eyeglasses during the daytime, ointment and a protective eye chamber at night.

Toxic neuropathies

A long list of substances can cause acute weakness. Some of the more common ones are listed here.

Anticholinesterases

The organophosphates and carbamates are commonly used insecticides, which can cause poisoning through skin, oral or pulmonary exposure. They inhibit cholinesterases, allowing acetylcholine to persistently stimulate the nicotinic and muscarinic receptors, which then can become refractory and thus cause weakness. This weakness is usually accompanied by the cholinergic toxidrome (see Chapter 21.2 on Toxicology), and, indeed, it is usually the respiratory and cardiac features that predominate.

However, there is an *intermediate syndrome* where 12 hours to 7 days after the initial poisoning, one finds proximal limb weakness that is unresponsive to atropine or pralidoxime. There may also be respiratory and bulbar paralysis. Recovery from this is usually complete.

Late neurotoxicity arises 4–21 days after the acute exposure, causing a mixed sensory motor deficit, which may take weeks or months to recover or may be permanent.

Diagnosis is by identification of the poison at source or in urine or serum drug screens and also by measuring serum cholinesterase activity level. Treatment is

supportive and with antidotes atropine and pralidoxime. Muscle relaxants will have a prolonged effect and should be avoided if possible.

Lead and other heavy metals

Lead, mercury and arsenic are all known to cause neuropathies, often taking the form of mononeuritis multiplex. It is uncommon for these to present as acute weakness. Nevertheless, a history of exposure should be sought and appropriate specimens taken if the cause of the acute weakness is unclear.

Chemotherapeutic agents

Vincristine, vinblastine and cisplatin are known to cause a peripheral neuropathy. Any child who is on, or has recently had, chemotherapy needs to have this considered as a possible cause.

Hereditary neuropathy

Rarely, a patient with an undiagnosed hereditary neuropathy presents with an acute exacerbation. A thorough history will reveal the chronicity of the problem. Such patients should be referred to an appropriate paediatric neurology service.

Muscular disorders

Infectious myositis

Viral infections such as influenza can cause a myositis that may in its severest form lead to rhabdomyolysis and myoglobinuria. Children frequently stop walking due to associated myalgia. These usually present with the other features of the infection and myalgia, rather than acute weakness. The weakness is only found on close examination, if the myalgia permits. The creatine kinase is raised. Various bacteria including *Streptococcus* and *Mycoplasma* species have also been reported to cause a focal myositis with tender muscles. The prognosis for viral myositis is generally good, although permanent muscle damage may ensue in focal bacterial myositis.

Acute presentations of children with isolated calf pain and tenderness with reduced mobilisation following a viral illness may not need any investigation. The child should be encouraged to drink plenty of fluids, and the parents should be reassured that the calf pain should resolve within a few days. They should

return to hospital if the child develops severe pain, dark urine, or fails to improve within 1–2 days.

Juvenile dermatomyositis

Juvenile dermatomyositis is a systemic vasculitis thought to be triggered by infection. Both enteroviruses and Group A *Streptococcus* have been implicated. Its peak age of incidence is 6 years. Because of its gradual onset this uncommonly presents as acute weakness. The child may well present with the rash before weakness has become apparent.

The rash appears on sun-exposed areas, especially the malar region of the face, and a purple discolouration of the eyelids is apparent (heliotrope rash). The rash may also be found on the extensor surfaces of the arms and legs, thorax, ankles and buttocks. The fingers develop thickening of the skin over joints, called Gottron's papules. The weakness comes on about 2 months after the rash and is usually very slow in onset. Small children may be noted to become gradually inactive, and older children have increasing difficulty with sport. Proximal muscle activities, such as climbing up stairs, reveal the weakness first. The vasculopathy can affect any muscle group, and children may present with aspiration, dysphagia or hoarse voice due to pharyngeal muscle weakness. The affected muscles are often tender and sometimes swollen.

The vasculitis can affect any organ system. Subclinical myocarditis and conduction defects are often found at diagnosis. Less common effects include renal dysfunction, hepatosplenomegaly, retinitis, iritis, seizures, depression, bowel dysfunction and pulmonary disease.

Diagnosis is made on the clinical picture associated with a raised serum creatine kinase. There are typical changes on muscle biopsy and electromyogram. There may also be elevated autoantibodies, liver function tests and abnormalities on MRI.

Complications include calcinosis of muscles, subcutaneous fat and fascia. These are more likely if the illness is prolonged and are decreased by aggressive treatment. Calcinosis lesions may become infected and lead to septicaemia.

These children may require judicious use of pain relief and require referral to a specialist paediatric unit. Treatment options range from sunscreen for the rash through to immunosuppression with glucocorticoids and chemotherapeutic agents, depending on the severity of the disease. Prognosis is generally good in children, with approximately 80% making a good recovery.

Metabolic/endocrine myopathies

Most endocrinopathies can produce weakness by several mechanisms. Often this is just myalgia and fatigue, but true myopathies can develop. This weakness usually responds to the treatment of the underlying endocrine disorder. Endocrinopathies can be associated with electrolyte disturbances that lead to weakness (e.g. hypokalaemia in Conn's syndrome) or with primary muscle diseases, such as hypokalaemic periodic paralysis with thyrotoxicosis.

Periodic paralysis

The periodic paralyses are a series of genetic ion channel disorders that lead to acute episodes of weakness lasting from 1 hour up to more than a day. They often come on after rest, during sleep or immediately following exercise but never during exercise. Diagnosis is by measurement of electrolytes during an attack, response to a metabolic challenge or by gene mutation identification. Hypokalaemic periodic paralysis usually presents in adolescence. The patient will have low potassium during attacks. Conversely, hyperkalaemic periodic paralysis usually presents in early childhood. The attacks are more frequent, occurring up to several times a day, and are associated with an elevated or normal potassium. The identification of these diseases is important, as they are treatable.

Somatisation disorders/malingering

Not infrequently children may present with a psychogenic cause of weakness. The findings of a careful neurological examination will demonstrate inconsistencies, and the child will be able to perform manoeuvres that require more strength than normal walking (e.g. knees giving way but not falling). Reflexes will be normal. Usually, no laboratory tests or imaging is necessary. However, if there is any doubt, consultation and investigation should be undertaken.

Once you have made the diagnosis it is best to take the parent or carer aside and discuss the diagnosis with him/her. Confrontation of the child is rarely helpful, and it is often best to reassure the child that this is not serious and that he/she will get better. This gives the child an exit from the medical review with dignity intact. An effort should be made to find the underlying psychological reason for the presentation, which may be anything from school avoidance to

sexual abuse. Consultation with social work or psychiatry may be indicated for ongoing counselling.

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Further reading

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8.5

Acute ataxia

Joanne Grindlay

ESSENTIALS

1 Acute ataxia (<72 hours) is a benign condition in most children.

2 The most common causes of acute cerebellar ataxia in children are post-infectious (acute cerebellar ataxia), drug intoxication and posterior fossa tumours.

3 Assessment focuses on identifying those children with features suggestive of a serious underlying cause: altered conscious state, fever, headache, focal neurological features or behavioural changes.

4 Ataxia is not uncommonly a presenting symptom of poisoning or excessive therapeutic drug levels, particularly older anticonvulsants.

5 Weakness may cause an unsteady gait and mimic ataxia.

6 Investigations in the child with ataxia are directed by the history and clinical features detected on examination.

7 While ataxia may be seen in post-concussion syndrome, conditions requiring acute intervention should be sought by CT scan or rapid MRI.

8 Most children with significant ataxia require admission for investigation and observation.

Introduction

Ataxia is an uncommon but important paediatric presentation to the emergency department (ED). Ataxia is a disorder of movement manifest by the loss of

coordination, most apparent as a disturbance of gait, with intact muscle strength. It may be associated with a disturbance of balance.

Ataxia is most often caused by a loss of function of the cerebellum, which controls the coordination of movement. Disease of the peripheral sensory nerves or the spinal column, particularly affecting proprioception, may also lead to ataxia as a result of abnormal inputs into the cerebellum. Cortical ataxia results from cerebral cortical dysfunction, particularly of the frontal lobe, while vestibular ataxia results from disease of the inner ear. Rarely, psychiatric causes of 'ataxia' may also be seen as a manifestation of conversion reaction.

The most common diagnosis of acute ataxia in children is a post-infectious ataxia called acute cerebellar ataxia (see below). This is a diagnosis of exclusion, made after consideration of other causes (Box 8.5.1). These include poisoning, metabolic disorders and organic brain lesions.

There are numerous hereditary conditions causing chronic ataxia that may present in childhood. These include Friedreich's ataxia, hereditary cerebellar ataxia, spinocerebellar ataxia and ataxia telangiectasia. The progressive nature of these conditions and their associated signs differentiate these from the acute ataxias.

Pathophysiology

Cerebellum

The cerebellum consists of two lateral hemispheres, divided into anterior and posterior lobes by the primary fissure. The hemispheres are joined by the vermis in the midline. The cerebellum connects to the cerebral cortex and brainstem via three paired sets of peduncles, the superior, middle and inferior cerebellar peduncles.

Cerebral hemispheres and vermis

There are three functional parts of the cerebellum. These are the archaeocerebellum, the palaeocerebellum and the neocerebellum.

The *archaeocerebellum* is formed by the flocculus, nodule and lingula of the vermis. It has only vestibular connections, which travel in the inferior cerebellar peduncle. The archaeocerebellum controls balance. Dysfunction leads to truncal ataxia characterised by a drunken gait, with swaying of the trunk and titubation when sitting, standing or walking. Typically there is neither alteration in fine

movements nor nystagmus. Reflexes are normal, and there is no tremor.

Box 8.5.1 Causes of acute ataxia in childhood

- Post-viral – acute cerebellar ataxia
- Poisoning/drug intoxication
- Tumours:
    - Posterior fossa, brainstem
    - Paraneoplastic syndrome
- Trauma including non-accidental injury:
    - Haematoma
    - Post-concussion
- Metabolic:
    - Hypoglycaemia
    - Hyponatraemia
    - Hyperammonaemia
    - Inborn errors of metabolism
- Infections:
    - Meningitis – bacterial, viral
    - Cerebral abscess
    - Malaria
    - Labyrinthitis
    - Encephalitis
- Vascular:
    - Stroke
    - Vasculitis
- Immune:
    - Multiple sclerosis
    - Acute disseminated encephalomyelitis

The vermis and surrounding nuclei of the cerebellum form the roof of the 4th ventricle. An expanding lesion may obstruct the cerebrospinal fluid (CSF) flow, with resultant hydrocephalus and truncal ataxia.

The anterior lobes of the lateral hemispheres, together with uvula and pyramid

of the vermis, form the *palaeocerebellum*. The palaeocerebellum connects to the spinocerebellar tracts and is involved with postural reflexes. Dysfunction leads to postural imbalance and increased reflexes.

The posterior lobes of the lateral hemispheres form the *neocerebellum*, which connects to the cerebral hemispheres, basal nuclei and the pontine nuclei, via the middle cerebral peduncle. It is involved in the coordination of fine, voluntary movements. Dysfunction leads to nystagmus, intention tremor, dysdiadochokinesis, hypotonia and decreased or pendular reflexes.

Cerebellar peduncles and connections

The cerebellum receives inputs from the vestibular and peripheral nervous systems. Outputs, which coordinate muscle movements by modifying tone and contraction, travel in three sets of peduncles (superior, middle, and inferior) between each cerebellar hemisphere and the brainstem.

Differential diagnosis

The most common cause of acute ataxia in children is post-infectious, acute cerebellar ataxia. Drug intoxication and posterior fossa tumours are less common.

Acute cerebellar ataxia

Acute cerebellar ataxia is the most common diagnosis in acute ataxia in children, particularly between 2 and 7 years of age. It is a diagnosis of exclusion, after consideration of more sinister causes such as tumours. An autoimmune aetiology is likely, with autoantibodies demonstrated in acute cerebellar ataxia following infections with varicella,[1,2] Epstein–Barr virus (EBV),[3] mycoplasma and human parvovirus B19.[4] The clinical presentation is of a prodromal illness, frequently non-specific, with or without an exanthema, 5–10 days prior to the onset of acute ataxia, though the timing may show considerable variation. If a specific aetiology is present it is historically most commonly varicella,[5] though a number of other viruses have been implicated (Box 8.5.2).[6–22]

Acute cerebellar ataxia usually presents with sudden onset of severe gait ataxia, though a small number of cases have an insidious onset. Most have dysarthric speech. Mild horizontal nystagmus occurs in 50% of cases. Findings

of intention tremor, dysdiadochokinesis, hypotonia and decreased or pendular reflexes are seen in two-thirds of cases but are less pronounced than the gait disturbance. Truncal ataxia is uncommon. Unlike acute disseminated encephalomyelitis (ADEM) or multiple sclerosis, there are no focal neurological signs.

Box 8.5.2 Causes of post-infectious acute cerebellar ataxia

- Varicella[6] and varicella vaccine
- Coxsackie A9[10]
- Epstein–Barr virus[8]
- Mycoplasma[9]
- Mumps[10]
- Poliomyelitis[11,12]
- Typhoid[13]
- Echovirus[14]
- Pertussis[15]
- Human parvovirus[16]
- Measles[15]
- Herpes simplex virus[17]
- Enterovirus 71[18]
- Malaria[19]
- Immunisation[20,21]
- Human herpesvirus-6[22]

Evaluation is aimed at excluding an alternative diagnosis, as there is no specific diagnostic test for acute cerebellar ataxia. The CT scan is normal in acute cerebellar ataxia; however, MRI may be abnormal. In one series, inflammatory changes were seen in the cerebellum of one of nine children.[23,24] There is a slight elevation of CSF cell count by 4–50 cells per microlitre in 32%, though occasionally (in 8%) the elevation is higher. There may also be slightly elevated protein 410–900 mg L^{-1}.[25]

Acute cerebellar ataxia usually begins to improve within a few days, but full recovery may take from 10 days to 2 months. Patients who have a slower recovery are still likely to recover fully. In one series, 91% recovered fully from their ataxia, including all those with varicella, EBV or post-vaccination, but 8% had sustained learning problems. Varicella-associated ataxia recovered quicker than non-varicella.[24]

Poisoning

Ataxia is not uncommonly a presenting symptom of ingestion or excessive therapeutic drug levels, particularly anticonvulsants.

Accidental poisoning occurs most commonly in 1- to 4-year-old children. A wide variety of compounds are implicated, including alcohols, substances of abuse and essential oils, as well as medications (Box 8.5.3). As distinct to acute cerebellar ataxia, these children usually have altered mental status and may have nystagmus and vomiting.

Anticonvulsants

Phenytoin toxicity with serum levels of >20–30 mcg mL^{-1} may produce signs of ataxia, nystagmus on lateral gaze and drowsiness. Onset of symptoms following an acute ingestion is usually within 1–2 hours and may persist for 4–5 days. At >30 mcg mL^{-1}, the ataxia and drowsiness become more marked and the nystagmus vertical.[26–28]

Box 8.5.3 Drugs causing acute ataxia

- Anticonvulsants:
    - Phenytoin
    - Carbamazepine
- Benzodiazepines
- Alcohols:
    - Ethanol
    - Isopropanol
    - Ethylene glycol
- Essential oils:
    - Eucalyptus oil

- Tea tree oil
- Pine oil

- Cough suppressants:
  - Codeine
  - Dextromethorphan

- Drugs of abuse:
  - PCP
  - Solvents, petrol, glue

Carbamazepine toxicity may also lead to ataxia. There are usually associated findings of drowsiness and nystagmus. There may be progression to seizures and coma, particularly if the level is >100 micromol L^{-1}.[29]

Benzodiazepines

Ataxia may be the sole presenting feature of benzodiazepine ingestion in children. In one series, it was an isolated finding in one-fifth of the cases who demonstrated ataxia. Other findings included lethargy (57%), GCS <15 (35%) and respiratory depression (9%).[30]

Alcohols

Ethanol intoxication produces ataxia, disinhibited behaviour and slurred speech. A serum or breath ethanol level will aid clarification of the diagnosis.

Ethylene glycol is the main component of antifreeze. Early symptoms of ingestion are similar to those of ethanol intoxication. Delayed features include cardiopulmonary distress and nephrotoxicity. Ethylene glycol produces a raised anion gap metabolic acidosis with an osmolal gap. Serum levels >20 mg dL^{-1} are toxic. The ethanol level will be zero.

Isopropanol toxicity is evident between 0.5 and 2 hours post ingestion and may include vomiting, ataxia, nystagmus, and altered mental state. Coma and apnoea may occur in severe poisoning.[31]

Essential oils

Eucalyptus oil is not an uncommon ingestion by children. In one series of 109 admitted children, 41% were asymptomatic. Those who were symptomatic demonstrated decreased conscious state (28%), vomiting (37%), ataxia (15%)

and pulmonary disease (11%). There was a correlation between ingested dose and toxicity. An ingestion of >5 mL 100% oil was associated with a significantly decreased conscious state, whereas <2–3 mL was associated with minor depression of consciousness.[32] A second series of 41 presentations, however, only demonstrated effects in 20%, with no correlation with presumed dose. The clinical effects included ataxia (5%), decreased conscious state (10%) and gastrointestinal symptoms (7%).[33]

Other essential oils that may produce nystagmus include tea tree oil[34] and pine oil. In one small series of pine oil cleaner (35% pine oil, 10.9% isopropyl alcohol), symptoms developed within 90 minutes of ingestion. Lethargy was present in all symptomatic children and ataxia in four of five cases of children.[35]

Cough suppressants

Codeine is contained in a number of cough medicines as well as in analgesics. In one series of 430 children, ataxia was reported in 9% receiving codeine. Associated symptoms are somnolence 67%, rash 39%, miosis 30%, vomiting 27%, itching 10%, angio-oedema 9%.[36] Dextromethorphan is a common component of cough and cold medications, which acts through opiate receptors in the medulla. It may cause opisthotonus, ataxia and bidirectional nystagmus. Fatality is highly unlikely, even with one hundred-fold the therapeutic dose.[37,38]

Substances of abuse

Phencyclidine (PCP) and lysergic acid diethylamide (LSD) are rare causes of ataxia in children. 1,4-butanediol, which is metabolised to gamma-hydroxybutyrate (GHB) upon ingestion, was responsible for ataxia, vomiting and seizures in several children following the ingestion of Aqua Dots, from toy craft kits.[39]

More common, however, is the abuse of hydrocarbon solvents; toluene in glue, spray paints and petrol. Hydrocarbons may produce ataxia as a component of acute intoxication and also as a chronic central nervous system effect.

Tumours

Cerebellar lesions may present with an acute rather than insidious onset of ataxia as a result of either haemorrhage into a tumour or as a result of hydrocephalus. The ataxia may progress to chronic ataxia. Paraneoplastic syndromes including paraneoplastic cerebellar degeneration and opsoclonus-myoclonus-ataxia

syndrome are uncommon causes of acute paediatric ataxia.

Clinical features suggestive of a brain tumour, and hence further investigation, include headache, vomiting, behavioural changes (particularly with frontal lobe lesions), papilloedema or cranial nerve dysfunction. The presence of opsoclonus (rapid, irregular eye movements) is an indication for imaging.

Posterior fossa tumours include medulloblastoma, astrocytoma and ependymoma.

Medulloblastoma (20–25% of posterior fossa tumours) usually presents in a child of less than 6 years with symptoms of ataxia (which may be truncal), headache, irritability or vomiting. The tumour may be located in the 4th ventricle or vermis.

Cerebellar astrocytoma (10–30% of paediatric brain tumours) is located in one of the cerebellar hemispheres. It is seen in primary-school-aged children who may display ipsilateral limb ataxia, headache and vomiting. The head may be held tilted to one side. Associated raised intracranial pressure may be life threatening.

Ependymoma (8–10%), located in the 4th ventricle, causes obstruction of CSF flow and may present with headache, vomiting and ataxia, which may be truncal.

Brainstem gliomas (10–15% of paediatric brain tumours) develop in the pons or medulla. They typically occur in early primary-aged children. Presenting symptoms include cranial nerve palsies, ataxia and vomiting.

Hydrocephalus may present with ataxia due to stretching of frontopontocerebellar fibres. Associated features are headache, vomiting and the late signs of raised intracranial pressure – altered conscious state, raised blood pressure and decreased pulse. Supratentorial tumours may also present with ataxia through involvement of the frontopontocerebellar fibres.

An occult neuroblastoma may present with a triad of acute ataxia, opsoclonus (jerky, random, chaotic eye movements) and myoclonus (severe myoclonic jerks of the head, trunk or limbs). The most common site for the tumour is in the abdomen.[40] This triad may also be seen with viral infections, including meningitis, particularly mumps, hence a lumbar puncture should be considered. Neuroblastoma may present with isolated ataxia and should be considered in cases of persistent or recurrent ataxia.[41]

Trauma

Head trauma is a frequent cause of ED presentations. While ataxia may be seen

in post-concussion syndrome, intracranial injuries (including haemorrhage and cerebral oedema that require acute intervention) should be sought by CT scan or MRI. A base-of-skull fracture may cause ataxia by direct damage to the vestibular apparatus. Non-accidental injury should be considered in all cases of paediatric trauma.

Infections

Meningitis, encephalitis and cerebellar abscess may all have ataxia as a presenting feature.[42,43] Other features to suggest an infective cause, such as headache, vomiting, fever and neck stiffness in the older child, will usually be evident.

In labyrinthitis and vestibular neuronitis, vertigo and vomiting are prominent. Apart from nystagmus and hearing alterations, neurological examination is normal.

Vascular conditions

Vertebrobasilar stroke is an uncommon cause of ataxia in children. Ataxia will not be an isolated finding. Other findings include ipsilateral cranial nerve palsies, contralateral weakness, vertigo and diplopia. Systemic lupus erythematosus vasculitis may rarely present with ataxia.[44]

Other neurological conditions

In isolated ataxia there is preservation of muscle strength. A number of neurological conditions may present with an unsteady gait (pseudo-ataxia) because of weakness.

These include Guillain–Barré syndrome in which areflexia and ophthalmoplegia (in Miller–Fisher variant) distinguish it from acute cerebellar ataxia. Tick paralysis should also be considered in cases where the patient has been in an appropriate area.

Multiple sclerosis/transverse myelitis is usually not seen until adolescent years. It may present with ataxia, optic or retinal neuritis, regional paraesthesias or weakness.[45]

ADEM (acute disseminated encephalomyelitis), a treatable post-infectious encephalomyelitis, where host myelin components become immunogenic, may

have ataxia as one abnormality within a cluster of multifocal signs, which may include altered conscious state, hemiplegia or optic neuritis. The diagnosis is based on MRI findings of multiple areas of demyelination in the brain and spine.

Other neurological conditions that may present with ataxia include seizures and complex migraine phenomenon. Because of the episodic nature of these conditions, there will be a recurrent nature to the ataxia.

Basilar artery migraine may demonstrate associated headache, blurred vision, visual field defects and vertigo. It tends to have a recurrent course.

Epilepsy uncommonly presents with ataxia. Ataxia may be seen post-ictally or in minor motor status or partial complex seizures. Other clues to seizure disorders include altered consciousness or motor manifestations.

Benign positional vertigo is uncommon in children. Ataxia accompanies severe, reproducible vertigo, nausea and vomiting. Cranial nerve examination is normal apart from nystagmus.

Metabolic disorders

A large number of metabolic disorders may have ataxia as a feature and should be kept in mind. Hypoglycaemia and hyponatraemia may present with ataxias, but other signs will also be present. Acute ataxia may be a presenting feature of vitamin B^{12} deficiency.[46]

The inherited metabolic diseases will likely demonstrate episodes of ataxia, together with the other features of the conditions, which will point towards the diagnosis. There may be a progression to chronic ataxias. Other inborn errors are listed in Box 8.5.4.

Chronic ataxia

Chronic ataxia usually has an insidious onset, but it may present as a progression of acute ataxia or as recurrent episodes. Causes include fixed deficits as in ataxic cerebral palsy, which makes up 10% of cerebral palsy and progressive diseases such as the hereditary ataxia, inborn errors of metabolism and tumours. Some causes are amenable to treatment. Box 8.5.4 lists some of the causes.

Box 8.5.4 Causes of chronic ataxia

- Brain tumours:

    - Cerebellar astrocytoma
    - Brainstem glioma
    - Medulloblastoma
    - Ependymoma
    - Cerebellar haemangioblastoma
    - (Von Hippel–Lindau)
- Hereditary ataxia:
    - Friedreich's ataxia
    - Ataxia telangiectasia
    - Dominant hereditary ataxia
    - Olivopontocerebellar degeneration
    - Roussy–Levy syndrome
- Hydrocephalus
- Congenital malformations:
    - Cerebellar aplasia/hypoplasia (autosomal recessive)
    - Vermal aplasia, including Dandy–Walker malformations
    - Arnold–Chiari malformations
- Inborn errors of metabolism:
    - Hartnup's disease
    - Wilson's disease
    - Refsum's disease
    - Abetalipoproteinaemia
    - Maple syrup urine disease
    - Argininosuccinicaciduria
    - Ornithine transcarbamoylase deficiency
    - Multiple carboxylase deficiencies – biotinidase deficiency
    - Pyruvate dehydrogenase deficiency
    - Juvenile GM2 gangliosidosis
    - Juvenile sulfatide lipidosis
    - Leigh's syndrome
    - Sphingolipidoses
    - Neuronal ceroid-lipofuscinosis
    - γ-Glutamyl cysteine synthetase deficiency
    - Triosephosphatase isomerase deficiency
    - Glucose transporter 1 deficiency syndrome
    - Vitamin B_{12},[46] E or folate deficiency

Hereditary ataxias/spinocerebellar degenerative

Friedreich's ataxia is an autosomal recessive condition, which manifests in a child less than 10 years of age with ataxia and nystagmus. There is usually rapid progression. On examination there is impaired position and vibration sense, positive Romberg's sign and absent tendon reflexes. The plantar response is up-going. Dysarthria is present. Kyphoscoliosis, distal muscle wasting and contractures, and cardiomyopathy may develop.

Ataxia telangiectasia is also an autosomal recessive condition with neurocutaneous manifestations. Ataxia is predominantly truncal and becomes evident in early childhood. Ocular and cutaneous telangiectasia become evident between 2 and 6 years of age. Developmental delay, increased susceptibility to infection (due to thymic atrophy and IgA and IgE deficiency), and an increased incidence of neoplasia are seen.

Congenital malformations

Cerebellar aplasia/hypoplasia, Dandy–Walker malformations, Arnold–Chiari malformations or vermal aplasia may result in ataxic cerebral palsy which accounts for 10% of all cerebral palsy. Signs include, but are not limited to, ataxia, hypotonia and tremor.

Clinical evaluation of the patient

The ED assessment includes history and a thorough examination to detect life-threatening conditions and to identify reversible factors. Investigation is dependent on the formulation of a differential diagnosis.

History

Important aspects of the history include the timing of the ataxia. Is it acute, recurrent or chronic?

What is mainly affected; is it the trunk or limbs? Are there other symptoms, such as headache, vomiting, blurred vision, altered mental status or nausea suggestive of a tumour or meningitis?

The antecedent history is important; for example, a recent viral illness may

suggest acute cerebellar ataxia, or there may be a history of trauma or possible drug ingestion.

Other symptoms, for example paraesthesia, may suggest an alternative diagnosis such as ADEM.

Examination

This should include a general examination to look for conditions in need of urgent intervention, for example signs of meningitis, shock, hypoglycaemia, raised intracranial pressure (ICP) or head injury. A complete examination is required to detect signs that may aid in the diagnosis, such as abdominal masses, nystagmus, opsoclonus and myoclonus in neuroblastoma, or signs of infection or vasculitis.

A complete neurological examination should be performed and documented. Cerebellar signs should be carefully assessed, initially by careful observation. Dysmetria and intention tremor can be assessed in the younger child by playing a game which involves pointing to items on a screen. Finger–nose test or dysdiadochokinesis may, however, be difficult to perform.

Gait should be assessed – abnormalities include a wide-based, staggering gait or, in younger children, refusal to walk. If only the anterior lobe or the vermis is involved, gait may be affected but the upper limbs spared. With truncal ataxia the child may have difficulty keeping balance whilst seated. This imbalance increases if sitting cross-legged, standing or walking. Heel–toe walking tests are useful in detecting cerebellar problems, but unsteadiness may also be due to weakness or sensory deficits.

Sensory ataxia is differentiated from cerebellar ataxia by the presence of impaired position and vibration sense and a positive Romberg’s test.

Lesions of the ipsilateral cerebellar hemispheres cause ataxia prominent in one direction. The child will fall towards the side of the lesion. Dysmetria and hypotonia are seen on the side of the lesion.

Cerebellar dysarthria (scanning speech) may be detected by repeating ‘sizzling sausage’. The speech displays a slow onset and is a slurred, jerky sound with an explosive nature.

Reflexes are often decreased in cerebellar lesions and are absent in Guillain–Barré syndrome. Pendular knee jerks are seen in severe cases of cerebellar dysfunction and in those with associated pyramidal tract defects. Decreased tone is seen in cerebellar lesions with drift and static tremor on holding up the arms.

Rebound may also be detected.

Nystagmus is horizontal in cerebellar lesions and maximal to the side of the lesion. It may be positional. In phenytoin intoxication, nystagmus is initially horizontal but becomes vertical at higher levels. Nystagmus in vestibular neuronitis is typically rotational. The cranial nerves should be carefully examined to look for brainstem involvement.

Investigations

Investigations are directed by the history and clinical features detected on examination. Acute cerebellar ataxia is primarily a diagnosis of exclusion, and investigation may be required to detect alternative conditions where urgent intervention is required. However, where there is a clear history of a viral prodrome, with normal and isolated cerebellar signs, no investigations may be required provided that close follow-up can be assured.

Toxicology screening, including for ethanol, ethylene glycol, anticonvulsants or benzodiazepines, should be considered, particularly if there is altered mental state.[47] If the drug screen is positive, and in an appropriate clinical setting, no other tests may be required.

Imaging by CT or rapid MRI is indicated urgently where there are signs of trauma or raised intracranial pressure. In the absence of a clear diagnosis the risk/benefit of an MRI, which may require general anaesthesia or sedation, should be considered. MRI is superior for identifying posterior fossa lesions; however, there is a low yield (<0.7%) from MRI in those children 3 years or younger, with symptoms of less than 72 hours' duration,[48] and a normal MRI does not reliably predict outcome.[49]

A lumbar puncture should be considered in children with fever, meningism or altered conscious state, where meningitis or encephalitis is possible. Differential of a space-occupying lesion or raised intracranial pressure may indicate that a CT scan be performed prior to the lumbar puncture. Treatment with antibiotics and antivirals should not be delayed whilst awaiting imaging and lumbar puncture results.

There is insufficient evidence to support screening for metabolic causes.[47] In recurrent ataxia, or where there are features of metabolic disease, further metabolic screening should be performed in consultation with a paediatric neurologist or metabolic physician. This may include blood gas, ammonia, electrolytes, urinalysis, liver function tests, thyroid function tests, lactate in

blood and CSF, pyruvate, cholesterol and lipoproteins. If occult neuroblastoma is suspected, urinary homo-vanillic acid and vanillylmandelic acid levels are measured. Other investigations may include an EEG if seizures are suspected.

Management

Treatment is aimed initially at detecting and treating emergency conditions. For example, in hydrocephalus and tumours, raised intracranial pressure is treated in consultation with the neurosurgical unit. In meningitis, urgent antibiotics are given. Hypoglycaemia and other reversible metabolic causes are treated. Intoxications and poisonings are generally managed by supportive care.

Acute cerebellar ataxia usually resolves over weeks to months. Occasionally there is a persistent movement disorder or behavioural and speech disorders. There is insufficient evidence to recommend either immunoglobulin or steroid therapy, and treatment is primarily supportive. Physiotherapy, occupational therapy and a wheelchair for mobility may be required.

Disposition

Most children with significant ataxia require admission for investigation and observation under a paediatrician. Some children who have mild ataxia, clearly due to post-infectious cerebellitis, are appropriate to be followed as an outpatient.

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8.6

Headache

John Cronin, and John M. Ryan

ESSENTIALS

1 The primary role of an emergency physician is to exclude a significant underlying cause for a headache, most urgently that of infection, raised intracranial pressure or an intra-cerebral bleed.

2 While the majority of children with headaches presenting to the emergency department (ED) are likely to be benign, appropriate investigation and follow-up of patients will ensure that serious causes are unlikely to be missed.

3 A thorough history and examination are most likely to lead to the diagnosis in the majority of children presenting to an ED with a headache.

4 *Red flags* prompting further investigation of headache include occipital headache; meningism; focal neurological signs; seizures; progressive nature; papilloedema; persistent vomiting; ataxia; presence of a ventriculoperitoneal shunt; age younger than 3 years; or early morning headaches, especially those that wake the child from sleep.

Introduction

The aim of this chapter is to provide the reader with a basic understanding of the aetiology of headaches in a paediatric population presenting to an emergency department (ED) while offering a functional and safe approach to their

management. Some key points should be remembered from the outset:

- Children may require different approaches depending on their age.
- Children under 3 years rarely complain of headache. This should prompt consideration of an organic cause.
- The classical history of migraine from a teenager may be more easily elicited than the history of symptoms from a shy 5-year-old.

Incidence

Perhaps surprisingly, headache is one of the most common presentations to the paediatric ED, comprising approximately 1% of all presentations.[1–3] Headaches in children are very common with up to 75% of children having had a headache of some form by the age of fifteen.[4] It has been reported that the prevalence of headache up to the age of 20 years is approximately 58% with a 1.5:1 female to male ratio.[5] Despite the frequency, very few paediatric patients with headaches ever consult their family physician or an ED. However, this does not take account of patients who present with a different complaint, such as a temperature, who might also have a headache as part of a concomitant illness.

Pathophysiology

The causes of headache are myriad, but the primary aim of the emergency physician should be to differentiate the patient with a headache that will run a relatively benign course from that which may be a symptom of significant underlying pathology with immediate health implications.

The overwhelming majority of headaches will be diagnosed on history and examination alone, with little additional information arising from investigations.[3,4,6] Furthermore, the vast majority of headaches that children present with to the ED are likely to be benign, but those that are not have the potential to be life threatening.

The classification of headaches is based on the underlying aetiology.[7] The International Headache Society has developed a classification of headache, the third edition of which was published in 2004 in *Cephalgia* and is also available on their website.[8] This classifies headache into three broad categories: **primary** and **secondary headaches** and **painful cranial neuropathies, other facial pains** and **other headaches** (Box 8.6.1).

The causes of some headaches will be dealt with in other chapters, e.g. Chapter 8.7 on meningitis, while some specific causes of headache will be discussed in more detail later in this chapter. We recommend an approach whereby the emergency doctor approaches each case by initially excluding the most sinister causes of the headache (Boxes 8.6.2 and 8.6.3).

Box 8.6.1 Summary of International Headache Society classification of headaches (ICHD-3)

Primary headaches

Migraine
Tension-type headache
Trigeminal autonomic cephalgias
Other primary headache disorders

Secondary headaches

Headache attributed to trauma or injury to the head and/or neck
Headache attributed to cranial or cervical vascular disorders
Headache attributed to non-vascular intracranial disorder
Headache attributed to a substance or its withdrawal
Headache attributed to infection
Headache attributed to disorder of homeostasis
Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
Headache attributed to psychiatric disorder

Painful cranial neuropathies, other facial pains and other headaches

Painful cranial neuropathies and other facial pains
Other headache disorders

Box 8.6.2 Causes of headache in children

Infection

- Meningitis, encephalitis, abscess
- Influenza
- Systemic infection
- Sinusitis
- Dental infection

Vascular

- Migraine
- Intracranial haemorrhage
- Hypertensive encephalopathy

Post lumbar puncture

Raised intracranial pressure

- Brain tumour
- Hydrocephalus
- Benign intracranial hypertension
- Trauma – SDH, concussion

Toxic

- Carbon monoxide poisoning
- Lead poisoning

Functional

- Tension or cluster headache

Psychogenic

Box 8.6.3 Important causes of non-benign headache

- Brain tumours and hydrocephalus
- Idiopathic intracranial hypertension
- Intracranial haemorrhage
- Meningitis, encephalitis or abscess
- Hypertensive encephalopathy

Clinical assessment

History

The first step in any medical assessment is the history, and this is no less important in the case of headache. Depending on the age of the child, the history may be taken from the child, the parents or guardians, or a combination of the two.

Site

Although classically unilateral in adults, migraine in children is more frequently bilateral, and children tend to have shorter attacks.[9] The headache of meningitis may involve neck pain and/or stiffness. Occipital headaches are relatively uncommon in children and have been reported to be an independent predictor of underlying pathology, particularly of the posterior fossa.

Nature

Headache of a throbbing nature is suggestive of migraine, while tension-type headache (TTH) is pressing/tightening and non-pulsating in quality and of mild or moderate intensity.[7] TTH tends not to be aggravated by physical activity, whereas migraine does. It has also been suggested that the inability of a child to describe the nature of the headache may in itself be a predictor of underlying pathology.[4] Severity of headache should be quantified using a pain rating scale (e.g. the visual analogue scale) appropriate for the age and cognitive levels of the child or adolescent.[10]

Onset of the headache

Abrupt onset of worst ever headache is a red flag for subarachnoid haemorrhage. However, this is rare in children, and a headache of this nature is most frequently

caused by an upper respiratory tract infection with fever, by sinusitis or by migraine.[2–4,10] There is a significantly higher proportion of underlying pathology in cases of acute headache, compared to chronic headaches.

A history of any recent head injury is important as concussion can be associated with a prolonged headache as well as other symptoms such as nausea, dizziness, malaise and emotional lability. A mild post-concussion headache may persist for a number of days or even weeks after a head injury but may require imaging to exclude a possible subdural haematoma, particularly where the headache is severe, responds poorly to analgesia, is associated with vomiting or is prolonged.

It should also be noted that the investigation of headache of acute onset is more properly the role of the emergency physician, while chronic headaches may be best investigated by the child's general practitioner or paediatrician.

Progression

The temporal progression is of relevance in children with headaches. For example, a classic migraine will last between 1 and 72 hours in a child, but attacks tend to be shorter in children compared to adults. A child with a chronic headache that is becoming progressively more severe may well have an underlying organic cause. This should prompt the emergency physician who encounters a child with such a pattern, even if incidentally, to ensure that neuroimaging is performed and that urgent appropriate follow-up is arranged.[6]

The persistence of headache should prompt the consideration of further investigation.

Pyrexia

The presence of a fever will often be reported by a parent and should immediately raise the suspicion of an infectious origin. An upper respiratory tract infection will be the most common, but meningitis and encephalitis need to be considered given the potential morbidity and mortality associated with them. The latter can be associated with confusion and clouding of consciousness. Other inflammatory disorders may also cause pyrexia. Equally, the absence of pyrexia does not exclude infection.

Behavioural change and avoidance behaviour

This is often noted in a collateral history. While entirely non-specific, it is

particularly important in raising suspicion of other causes of a headache, such as a school phobia or drug misuse in the adolescent, or indeed it may be a pointer towards sexual assault.[11]

Sleep disturbance

Benign causes of headache rarely wake patients from their sleep, and if a child is being woken by a headache this should prompt the consideration of further investigation or referral.

Postural symptoms

A headache that is worse on lying flat or exacerbated by coughing may be related to raised intracranial pressure. Early morning headaches after recumbency of sleep may occur with raised intracranial pressure or space-occupying lesions.

Neurological deficit

A history of neurological deficit, even if transient, is a red flag. This is particularly important, as some children may have subtle objective neurological findings that could easily be overlooked in a hurried examination. Parents may have noticed an unusual posture or limp but may not immediately mention it unless prompted. The importance of this is reinforced by the evidence that it may take an average of 7 months for a brain tumour to be diagnosed, with as many as three different consultations with a physician. This is despite the fact that a significant proportion of children with tumours have abnormal neurological examinations.[12]

Family history (especially of migraine)

Approximately 70–90% of diagnosed cases of migraine have a positive family history. Therefore diagnosing a migraine in the absence of a family history should be done with caution.

Analgesic use

Abolition of a headache with simple analgesics or restoration of normal activities is more common in those with upper respiratory tract-associated headaches. Chronic daily use of analgesics can in itself be a cause of headaches.[13]

Examination

A number of specific points should be considered when examining the child with a headache. Headache may be a secondary symptom of a more generalised illness. A comprehensive physical examination is therefore required. Observation of a child from a distance may be valuable. A child lying quietly, unresponsive to the environment, should prompt earlier assessment than an active child who interacts normally with healthcare staff. One must also consider other causes of headache that are not readily discernible. In some cases there may be few clinical signs, such as with carbon monoxide poisoning from houses with poorly ventilated gas boilers.[14] Heavy metal poisoning is another rare cause. In the majority of patients with primary headache disorders, the general physical and neurological examinations are normal.[15]

A focused examination should be directed on the basis of the history. Vital signs may provide valuable keys to the aetiology. The temperature should be recorded on more than one occasion. Likewise, it is important to measure the blood pressure as headache may be the presenting feature of coarctation of the aorta or underlying hypertension. The blood glucose is relevant if the history suggests hypoglycaemia. The febrile, toxic-appearing child should be examined for evidence of nuchal rigidity and other signs of meningeal irritation, which may be present in meningitis.

A significant proportion of children who present with headache will have a diagnosis of respiratory tract infection, hence the importance of a thorough ear, nose and throat examination. Occasionally a child with tonsillitis will present with headache, without any complaint of a sore throat. Dental examination may reveal percussion tenderness of an infection that may cause a referred temporal headache. The presence of nasal discharge or respiratory symptoms should prompt the clinician to seek evidence of sinus tenderness.

It is useful to measure head circumference and plot this on centile charts, even for the older child. A disproportionately large head should prompt further investigation,[5] and the tension should be palpated in infants who have an open anterior fontanelle.

A careful examination may reveal subtle evidence of trauma, which may or may not have been sustained accidentally. Headache has been retrospectively described as being more common in victims of sexual abuse. In the child who has a ventriculoperitoneal shunt, the reservoir should be assessed by palpation (see Chapter 8.1).[11]

Ophthalmological examination including fundoscopy is particularly important as it may influence immediate management. The presence or absence of papilloedema should be specifically sought as this may be the only objective finding in cases of idiopathic intracranial hypertension (IIH). In this condition the CT scan may be normal and the diagnosis based on an elevated opening pressure >20 cm H_2O on lumbar puncture. This valuable test is frequently neglected in lumbar punctures performed in the ED but should be done if one is considering idiopathic intracranial hypertension. The cerebrospinal fluid (CSF) protein, glucose and cell counts are normal in IIH. These children may present with intermittent headache, vomiting, blurred vision or diplopia (Chapter 8.2).

Retinal haemorrhages should also be sought on fundoscopy. Their presence is considered as evidence of significant trauma in the absence of any other causes such as hypertension or diabetes mellitus. It is difficult, if not at times impossible, to achieve successful examination of the fundi and retina of the younger child. Often one will require mydriatic eye drops to dilate the pupils or seek the opinion of a suitably qualified senior colleague. A deferred formal retinal assessment by an ophthalmologist may be required in cases where the possibility of non-accidental injury exists. Eye movements must be carefully examined as subtle nerve palsies may be apparent early in children presenting with a space-occupying lesion or hydrocephalus.

The main consideration in a cardiovascular examination should be directed at the exclusion of hypertension and the palpation of femoral pulses, checking for evidence of coarctation of the aorta.

Abdominal examination is usually normal but is nevertheless important, as nausea, vomiting and abdominal pain are frequently associated with headache in children. It is recognised by the International Headache Society that ‘cyclical vomiting’ and ‘abdominal migraine’ are distinct entities and not infrequently progress to the typical pattern of migraine in adulthood. Hypertension of renal origin may be suggested by the presence of polycystic kidneys.

A child’s gait should also be assessed to check for ataxia, and a neurological examination of cranial nerves and all limbs should be done to check for focal or hemiparesis, loss of coordination and abnormal tendon reflexes.[3]

The child should be fully undressed. In particular, a rash or abnormality of pigmentation should specifically be sought. The most urgent of these is the search for the petechial rash of meningococcal disease, but examination may also reveal neurofibromas and café-au-lait spots associated with neurofibromatosis or a pigmented patch associated with tuberous sclerosis.

Bruises or evidence of other unexplained injuries of different ages may be seen in cases of suspected non-accidental injury.

In addition, a psychiatric review of children and parents should be performed when needed.

Investigation

The issue of how to investigate a child with a headache in the ED is not clearly defined and remains contentious. It is clear that in the majority of cases the cause of children presenting to the ED with a headache is a respiratory tract infection or primary headache syndrome. The key role of the emergency physician is to diagnose those headaches that may have significant underlying pathology. Investigation should be driven by the history and by findings on examination, as the majority of diagnoses should be suggested by these and confirmed by appropriate diagnostic tests.

Blood tests

Laboratory-based testing has a very low yield in children with headaches. Occasionally they can alert the clinician to more serious underlying pathology, but their value is diagnostically limited in the emergency setting. They should not delay the progression to more specific investigation and treatment. Indeed, much of the useful information can be obtained at the point of care with the use of a venous blood gas, which provides information on pH, electrolytes, haemoglobin and even carboxyhaemoglobin levels.

Elevated inflammatory markers, such as white cell count and C-reactive protein, may help point to an infective aetiology. However, normal values should not be used to exclude meningitis, encephalitis or other conditions such as tonsillitis or middle ear infections if they are suspected clinically.

Imaging

Numerous studies have assessed the value of investigating a child with headache, with most focusing on the relevance of neuroimaging.[4,16–18]

Subjects who have any signs or symptoms of focal/progressive neurological disturbances should be investigated with CT scan of the brain or MRI.[10,18]

A CT scan cannot be considered a benign test as it carries with it a risk in relation to the sedation and transfer of a child and radiation exposure. A CT head scan carries a 1:1500 risk of developing a subsequent cancer directly related to

that radiation exposure.[19]

In the setting of an acute atraumatic headache, the absence of focal neurology or other 'red flags' (as listed later in this chapter), a CT scan is unlikely to be of significant value (Box 8.6.4).

A guide to CT scanning in the context of trauma is considered separately (Chapter 4.2).

In the context of a chronic, *non-progressive* headache with a normal neurological examination, CT is unlikely to be of value. This is different to a chronic *progressive* headache, which is a concerning feature and more likely to harbour underlying pathology, and an MRI would be indicated.[16,20] However, this cohort of patients is less likely to be seen in the ED, and, in the absence of an acute deterioration, their investigation may be better performed by their family physician, paediatrician or paediatric neurologist.

Box 8.6.4 Red flags prompting further investigation of headache

Occipital headache
Meningism
Focal neurological signs
Chronic progressive headaches
Persistent vomiting
Seizures
Papilloedema
Focal neurological symptoms
Ataxia
Presence of a VP shunt
Age younger than 3 years
Abnormal eye movements
Early morning headaches especially those that wake the child from sleep

VP, ventriculoperitoneal.

Lumbar puncture

Lumbar puncture is mandatory in any case of suspected meningitis or encephalitis. It is a reassuring test when normal and often diagnostic when

abnormal. It should be noted, however, that post lumbar puncture headache is also a recognised complication in children, as it is in adults.[21] Such an invasive procedure may often be emotionally traumatic for a child and should be performed with appropriate procedural sedation. Given the invasive nature of the test, it is not recommended as a routine investigation in the diagnosis of headache. However, a lumbar puncture with measurement of an elevated opening pressure is diagnostic in children with benign intracranial hypertension (Chapter 8.2).

Other investigations

An EEG, if available, may have a role if there is a history of a suspected seizure.[3,10,22] Other investigations such as electromyogram, SPECT, positron emission tomography, autonomic testing and transcranial Doppler have not clearly demonstrated their utility in the acute setting.[17]

Management

Symptomatic treatment in the ED of most headaches in children can be achieved by use of oral analgesics such as paracetamol, codeine or ibuprofen. It should be noted that, in most children, headaches due to uncomplicated upper respiratory tract infections can be expected to settle with simple analgesics. Headaches that require stronger analgesics to control pain are an indication for admission for further evaluation.

Disposition

The disposition of a patient from the ED should be considered on an individual basis, though some guidelines may be helpful in minimising the risk of missing a significant diagnosis. The increasing use of observation wards and short-stay units can prove helpful in selected cases. Such a facility is most useful in the case where an infective cause of the headache is suspected.

All patients who present to the ED with headache and are discharged should have a follow-up appointment in the short term with either their family physician or a paediatrician, depending on each case. This will ensure that not only are high standards of care maintained but unnecessary acute investigations can be avoided that would otherwise be ordered in order to exclude every possible cause of headache. Furthermore, there may be psychosocial aspects of relevance

to childhood headaches that may be more appropriately dealt with by the family physician.

Migraine

Essentials

1 Family history of migraine is very common, such that a diagnosis of migraine in the absence of a family history should be carefully considered.
2 Unlike adult migraine, simple analgesics are often effective in the context of childhood migraine.
3 Childhood migraine is often bilateral.
4 Childhood migraine may be shorter lived than in adults (1–72 hours).
5 Migraine is frontotemporal. Occipital headache suggests a sinister cause of headache.

Childhood migraine is a common condition presenting to the ED. The prevalence of migraine headaches in children varies from approximately 3% in younger children to 15% in adolescents.[23–25] In younger children, boys tend to have more migraines than girls, but that this reverses at puberty.[5,26,27] The diagnosis is based on a good history, but it can be a little more difficult to make in the younger age group. The emergency physician should not diagnose migraine in children less than 3 years old. Childhood migraine also has a wider spectrum of symptoms than adult migraine and should be considered in a patient who has recurrent abdominal pain, with normal investigations. It is important to remember that, although the diagnosis of migraine is common, it is principally a diagnosis of exclusion.

Pathophysiology

The exact mechanism of migraine is complex but is thought to occur as a result of a cascade of events that result in neurovascular vasodilatation.[23,28,29] The primary dysfunction is believed to be related to centres in the brainstem that regulate vascular tone and pain sensation.[30] 5-hydroxytryptamine (serotonin) (5-HT) has an inhibitory role in the central nervous system (CNS) preventing vasodilatation, hence the use of serotonin agonists such as triptans in the acute

treatment of migraine. It is likely also that a hyperexcitable cerebral cortex also has a role, leading to cortical spreading depression (CSD) and in turn to activation of the trigeminal nerve and its associated vessels. CSD can lead to areas of oligaemia of the cortex and may be responsible for the aura associated with migraine. Following this, the trigeminal nerve afferents appear to be sensitised and may account for the fact that ordinary activities can significantly exacerbate the migraine.[31]

Clinical features

Migraine without aura is more common than migraine with aura in children.[32] A number of key features are listed below.

International Headache Society diagnostic criteria of migraine without aura:[8]

A. At least five attacks, fulfilling criteria B–D
B. Last 4–72 hours (untreated or unsuccessfully treated)
C. Headache has at least two of the following four characteristics:
    1. Pulsating in quality
    2. Unilateral or bilateral (unlike adult migraine which is typically unilateral), *not* occipital
    3. Aggravation by routine activities (walking or climbing stairs)
    4. Moderate or severe pain intensity
D. At least one of the following during the headache:
    1. Nausea and/or vomiting
    2. Photophobia or phonophobia (may be inferred from child's behaviour).

Not better accounted for by another ICHD-3 diagnosis.

Occipital headache is rare in children and suggests a structural abnormality and not migraine.

In migraine with aura the headache is preceded by a period of altered perception that may take numerous forms. This period of altered perception can last up to 60 minutes. Examples are positive visual symptoms (flickering lights, spots, lines), negative visual symptoms (loss of vision, including partial), sensory symptoms (pins and needles) or fully reversible speech disturbances (dysphasia). This period of altered perception can last up to 60 minutes prior to

the onset of the headache.[8]

Assessing the severity of headache in a child can prove challenging. Children almost universally point to the unhappiest face on a visual analogue scale when describing a headache. More reliable answers may be achieved by asking about quality of life issues such as ability to continue playing. Children who can continue playing during a headache appear to have a more benign cause to their headache.

Up to 90% of patients with migraine have a positive family history, although it is noteworthy that only one migraine gene has been identified, and this is associated with only 50% of the cases of familial hemiplegic migraine, a particularly rare condition.[6]

A number of other conditions are considered to be migraine disorders in childhood, most notably cyclical vomiting and abdominal migraine. In these conditions there may be recurrent episodes of severe abdominal pain with associated vomiting, but the examination and investigation of the children are often normal. These conditions are a diagnosis of exclusion, but their consideration may lead to appropriate referral and follow-up as they carry significant morbidity.

Migraine variant disorders may cause transient abnormal neurological findings, such as hemiplegia, ophthalmoplegia or confusional state. These conditions are rare, and an abnormal examination should prompt the emergency physician to investigate further.

Investigation

Migraine as a distinct entity has no specific diagnostic test. Key to the diagnosis is the history, as described above, with a normal neurological examination. Investigations should be requested on the basis of excluding other pathology as the underlying cause of headache. It is notable that, although a significant number of migraine sufferers have abnormal EEGs, these add little diagnostic value and on current evidence should remain a research tool.[17]

Treatment

Most cases of childhood migraine can be adequately managed with simple analgesics such as paracetamol (15 mg kg^{-1}) and ibuprofen (10 mg kg^{-1}), although ibuprofen appears superior.[33,34] As nausea can often be a significant

feature, prochlorperazine (0.15 mg kg; maximum 12.5 mg) appears to be of benefit.[35] A study of 66 patients in 2004 found prochlorperazine to be superior to ketorolac at reducing pain from acute paediatric migraine treatment in the ED.

Chlorpromazine is commonly used, in particular in Australia, for severe migraines. However, the current existing evidence is weak.[36,37] The recommended dose is 0.15 mg kg in 1 litre of normal saline over 1 hour. Patients should be observed for hypotension. Chlorpromazine can prolong the QTc interval and therefore should be avoided in patients with cardiac disease, a family history of sudden death, or potassium or magnesium deficiencies (e.g. after persistent vomiting). In recent years, more evidence has emerged on the effectiveness of triptans in acute migraine in the paediatric population. Of particular relevance in children, intranasal triptans have been studied with intranasal sumatriptan (10 mg for a body weight of 20–39 kg and 20 mg for >40 kg) and zolmitriptan (5 mg) recommended in the paediatric populations over 5 and 12 years, respectively.[38,39] Placebo-controlled RCTs have shown efficacy of oral rizatriptan and of the combination therapy of sumatriptan and naproxen.[40–43] At present though, the triptan group of drugs remains unlicensed for paediatric use in many countries.

Limited but promising evidence of the use of subanaesthetic doses of propofol has been reported, but larger studies are needed before its use can be recommended.[44,45] There are very limited data regarding prophylactic treatment of migraine in children, and hence no clear recommendations can be given, other than ensuring appropriate follow-up of these cases.

Disposition

All patients with a discharge diagnosis of migraine should be followed up to ensure that the headache resolves within the expected time frame and also to ensure that it does not progress. Migraine can carry significant impairment to lifestyle, if unrecognised. Follow-up should be arranged with the primary care physician, with review of medication and parental understanding of the condition. Children with frequent migraine warrant referral to a paediatrician.

Conclusions

Patients with headaches commonly present to paediatric EDs. Those children presenting with new-onset symptoms, significant neurological symptoms/signs

or evidence of abnormal physiology should be rapidly assessed, with early referral for diagnostic facilities and neuroimaging being a priority. Children with chronic symptoms or recurrence of chronic symptoms should have a thorough examination and treatment. Follow-up should be arranged with the patient's paediatrician or GP.

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8.7

Central nervous system infections

Meningitis and encephalitis

Mike Starr

ESSENTIALS

1 Bacterial meningitis can be rapidly progressive and result in substantial morbidity and mortality.

2 A high index of suspicion of the possibility of meningitis must be maintained in any sick infant or child as the symptoms and signs are frequently non-specific, particularly with younger age.

3 Antibiotic treatment must not be delayed.

4 Careful management of fluid and electrolyte balance is critical.

5 Careful evaluation for features suggesting raised intracranial pressure should be done at the initial assessment.

6 Children with suspected meningitis should have a lumbar puncture (LP) performed, unless there is a clear contraindication.

7 Encephalitis may be infectious or immune mediated, and both may be treatable.

8 HSV encephalitis must be considered in any child with features of meningitis with encephalopathy.

9 Empiric aciclovir must be given early when encephalitis is suspected.

10 In a patient with encephalitis without an identified aetiology, corticosteroids and/or intravenous immunoglobulin (IVIG) should be considered.

Introduction

Meningitis and encephalitis are medical emergencies that require prompt assessment and treatment. Meningitis is inflammation of the meninges that surround the brain and spinal cord. Resultant inflammatory cells typically spill into the cerebrospinal fluid (CSF) from the meninges, producing an increased cell count. Encephalitis is inflammation involving the brain parenchyma, which may be infectious or immune mediated. There is considerable overlap between the features of meningitis and encephalitis, and the two may coexist. Encephalitis should be considered if a child has encephalopathy, convulsions or neurological deficits.

Meningitis

Classification

Meningitis is usually broadly classified as bacterial or aseptic. Aseptic meningitis may be the result of a localised or systemic insult but is most commonly caused by viruses. Fungal meningitis usually occurs in immunocompromised children. Tuberculous meningitis occurs in regions of high prevalence of tuberculosis.

Aetiology

Bacteria

The bacterial causes of meningitis vary with the age of the child. In infants less than 2–3 months old, organisms acquired from the maternal genital tract predominate: group B streptococci, *Escherichia coli* and *Listeria monocytogenes*. In older children and adults, the most common causes are *Neisseria meningitidis* and *Streptococcus pneumoniae*. Other causes, including *Staphylococcus* species and gram-negative bacilli, are occasionally seen in immunocompromised hosts or following trauma or neurosurgery. *Haemophilus influenzae* type b (Hib) is rarely seen as a cause now because of widespread immunisation. *Mycobacterium tuberculosis* is rare, other than in children who have had contact with local populations in countries of high prevalence.

In Australia, meningococcal disease, particularly that caused by serogroup C, has declined since introduction of the meningococcal C conjugate vaccine (MenCCV) into the National Immunisation Program (NIP) schedule. The most

common *N. meningitidis* serogroup causing invasive disease in those under 19 years is serogroup B (83%), followed by serogroup W135 (9%).[1] Although a serogroup B conjugate vaccine (4CMenB: Bexsero™) is now available in Australia, it is not included in the funded NIP.

A 7-valent pneumococcal conjugate vaccine (7vPCV) was introduced into the NIP schedule in 2001, leading to a dramatic reduction in the overall incidence of invasive pneumococcal disease (IPD) in Australia.[2] However, an increase in disease caused by non-7vPCV serotypes was also observed.[3,4] In 2011, a 13-valent vaccine (13vPCV) replaced 7vPCV in the NIP. The annual rate of IPD in children aged less than 5 years fell in the following year but has remained about the same since then, although the proportion of cases caused be 13vPCV serotypes has continued to decline.[5] Penicillin resistance remains relatively low amongst *S. pneumoniae* isolates (around 10%) and isolates with reduced susceptibility to ceftriaxone/cefotaxime remain rare (around 2%).[6] After the introduction of PCV13 in the United States, there was no change in the number of cases of pneumococcal meningitis in children, although the proportion of cases caused by PCV13 serotypes significantly decreased.[7] Moreover, isolates with reduced susceptibility to ceftriaxone significantly decreased.[7]

Table 8.7.1

Cerebrospinal fluid findings in meningitis

	White cell count		Biochemistry	
	Neutrophils ($\times 10^6$ L^{-1})	Lymphocytes ($\times 10^6$ L^{-1})	Protein (g L^{-1})	Glucose (CSF:blood ratio)
Normal (>1 month of age)	0	≤5	<0.4	≥0.6 (or ≥2.5 mmol L^{-1})
Normal (<1 month of age)	0*	<20	<1.0	≥0.6 (or ≥2.1 mmol L^{-1})
Bacterial meningitis	↑ (but may be normal)	↑ (usually <100)	↑ (but may be normal)	↓ (but may be normal)
Viral meningitis	↑ (usually <100)	↑ (but may be normal)	0.4–1.0 (but may be normal)	Usually normal
Encephalitis	↑ (usually <100)	↑ (but may be normal)	0.4–1.0 (but may be normal)	↓ (but may be normal)

* Some studies have found up to 5% of white cells in neonates without meningitis comprise neutrophils.

Viruses

Enteroviruses, including coxsackie and echoviruses, cause 85–95% of cases of viral meningitis. Parechoviruses (which are now known to be genetically distinct from enteroviruses) have recently been recognised as a cause of meningoencephalitis in infants.[8] Other viruses tend to cause encephalitis (see below).

Fungi

Cryptococcus neoformans is the most common fungal cause of meningitis, but occurs almost exclusively in immunocompromised hosts.

Clinical findings

The classical features of meningitis comprise fever, headache, vomiting, neck stiffness, photophobia and altered mentation. However, the clinical manifestations are often non-specific, particularly in infants and young children. They may include fever, irritability, lethargy, poor feeding or vomiting. Up to 58% of children with meningitis have received antibiotics before the emergency department (ED) presentation.[9] This may modify the clinical presentation of meningitis.[10] It is therefore important to consider the possibility of central nervous system (CNS) infection in any sick infant or child, particularly if he/she is already taking antibiotics.

If the fontanelle is still open, it may be bulging when examined with the infant in a sitting position. Photophobia is difficult to ascertain in young children, and other signs of meningeal irritation may be absent or difficult to elicit. Resistance to being picked up or distress on walking may be the only clues. Kernig's sign (inability to extend the knee when the leg is flexed at the hip), Brudzinski's sign (bending the head forward produces flexion movements of the legs) and nuchal rigidity may be present in older children but have been shown to have low positive and negative predictive value in adults with meningitis.[11]

Rashes may occur with any bacterial meningitis although are less common with pneumococcal infection. Petechiae or purpura is suggestive of meningococcal sepsis but may also occur in Hib and viral meningitis. Enteroviral meningitis may even be associated with florid purpura fulminans.

It is impossible to reliably differentiate between bacterial and viral meningitis on clinical grounds. However, children with enteroviral meningitis are more likely to present in summer or autumn with gradual onset of non-specific constitutional symptoms including diarrhoea, cough and myalgia, in addition to the more typical features.

Investigations

Definitive diagnosis of meningitis relies on examination of the CSF with biochemical analysis, microscopy and culture. Children with suspected meningitis should have a lumbar puncture (LP) performed, unless there is a clear

contraindication. The only absolute contraindication is raised intracranial pressure (ICP). It may be difficult to determine whether ICP is raised, but the following signs may be indicative:

- Coma (absent or non-purposeful response to painful stimulus)
- Abnormal pupillary responses
- Abnormal posturing
- Focal neurological signs or seizures
- Recent (within 30 minutes), prolonged (over 30 minutes) or tonic seizures
- Papilloedema – although this is an unreliable and late sign of raised ICP.

A bulging fontanelle, in the absence of other signs of raised ICP, is not a contra-indication to LP.

The threshold for performing an LP should be lower in young children with less specific signs or those who have been taking antibiotics prior to presentation.

Cerebral CT should not be used to decide whether it is safe to proceed with LP or not. In a prospective study of bacterial meningitis, CT findings obtained during the acute stages failed to reveal any clinically significant abnormalities that were not suspected on neurological examination.[12] Moreover, cerebral herniation can occur with a normal CT,[13,14] and the true cause of coning and relationship to prior lumbar puncture is not clearly established.

Other relative contra-indications to LP include:

- cardiovascular compromise or shock
- respiratory compromise
- coagulopathy or thrombocytopenia.

Examination of the cerebrospinal fluid

A CSF specimen should always be sent for *urgent microscopy* to help guide empiric treatment. Normal CSF is clear and contains few cells (and no neutrophils). As few as 200×10^6 cells per litre will cause CSF to appear turbid. The CSF profile may help differentiate between bacterial and viral meningitis, but the findings vary. Table 8.7.1 indicates the typical profiles in normal children and those with meningitis. However, these should always be interpreted in the context of the clinical picture. In early bacterial meningitis, the CSF cell count

may be normal. In enteroviral meningitis, there is typically a neutrophil predominance early, and this may remain so for more than 24 hours.[15]

Organisms are seen on CSF Gram stain in 60–80% of cases of bacterial meningitis, provided that prior antibiotics have not been given. The sensitivity is highest in pneumococcal meningitis. Prior antibiotics may preclude culture of the causative organism, but the biochemistry and white cell count remain abnormal for several days whether or not antibiotics have been given.

A traumatic tap occurs in 15–20% of LP in children.[16,17] Several formulae have been devised for interpretation of CSF contaminated with blood, but the safest practice is to disregard the red cell count and treat if meningitis is suspected until culture and other studies are clearly negative.

Seizures do not result in increased CSF cell count in the absence of meningitis.

Bacterial or viral DNA can be detected in blood and/or CSF using polymerase chain reaction analysis (PCR). Sensitivity and specificity are high, particularly for *N. meningitidis*, HSV and enterovirus. PCR for *N. meningitidis* is particularly useful in patients with a clinical picture consistent with meningococcal meningitis who have received prior antibiotics.

Latex agglutination allows rapid detection of bacterial antigens in CSF and urine. However, it lacks sensitivity and specificity, other than for Hib, and is therefore not clinically useful.

Other investigations

- Culture of blood, throat swab, stool/rectal swab or of skin lesion may yield a causative organism.
- Gram stain on blood smear may be positive.
- Blood glucose should be measured at the same time as CSF glucose.
- A baseline sodium should be measured. Hyponatraemia occurs in about one-third of children with meningitis and may be due to increased antidiuretic hormone secretion, increased urine sodium losses, and excessive electrolyte-free water intake or administration.
- Full blood count and acute phase reactants (e.g. C-reactive protein) may provide supportive information but are more useful when measured serially to monitor the response to therapy.

Management

Antibiotics

Following initial fluid resuscitation, the emphasis is on prompt commencement of parenteral antibiotics (Box 8.7.1). Delay in antibiotic therapy has been associated with adverse clinical outcome in adults with bacterial meningitis,[18] although the evidence for this in children is not as clear.

Steroids

Routine administration of steroids as adjunctive therapy has been controversial in the past. The evidence that they protect against neurological (particularly audiological) complications of childhood meningitis is strongest for Hib meningitis, when dexamethasone is given before the first dose of antibiotics and when a third-generation cephalosporin is used. A large European trial in adults with meningitis showed reduction in mortality and severe morbidity with pneumococcal meningitis.[19] A Cochrane meta-analysis including adult and paediatric trials concluded that adjuvant steroids are beneficial for children with bacterial meningitis.[20] Evidence from animal studies shows that dexamethasone reduces penetration of vancomycin into infected cerebrospinal fluid.[21] Thus, there is concern that use of dexamethasone with vancomycin could compromise the efficacy of vancomycin in third-generation cephalosporin-resistant strains. Fortunately, the majority of cases of pneumococcal meningitis are still caused by strains that are susceptible to penicillin and third-generation cephalosporins.

Box 8.7.1 Empiric antibiotics for management of meningitis.

Age <3 months

Amoxicillin 50 mg kg^{-1} IV 12-hourly (week 1 of life), 8-hourly (week 2–4 of life), 4–6-hourly (>week 4 of life) ***plus***
Cefotaxime 50 mg kg^{-1} IV 12-hourly (week 1 of life), 6-hourly (>1 week of age) ***or*** Ceftriaxone* 100 mg kg^{-1} (max 2 g) IV daily or 50 mg kg^{-1} (max 1 g) IV 12 H

Age >3 months

Ceftriaxone 100 mg kg^{-1} (max 2 g) IV daily or 50 mg kg^{-1} (max 1 g) IV 12 H ***or***
Cefotaxime 50 mg kg^{-1} (max 2 g) IV 6-hourly

* Ceftriaxone should be avoided in neonates if premature, jaundiced or receiving calcium containing solutions, including total parenteral nutrition (TPN).

Accordingly, children (>4 weeks old) who are being treated for possible meningitis (who have not yet received parenteral antibiotics or who have received their first dose less than 1 hour previously) should be given dexamethasone 0.25 mg kg^{-1} IV (max 10 mg) (followed by 0.25 mg kg^{-1} 6-hourly). Steroids should preferably be given 15–30 minutes before antibiotics, although antibiotic administration should not be delayed for more than 30 minutes.

Fluids

Careful management of fluid and electrolyte balance is important in the treatment of meningitis. Over- and under-hydration are both associated with adverse outcomes.[22–24] Many children have increased antidiuretic hormone secretion, and some will have dehydration due to vomiting, poor fluid intake or septic shock. Assessment of the clinical signs of hydration, including weight, measurement of the serum sodium, documentation of urine output, and clinical assessment of the neurological state, should be monitored closely and the total fluid intake adjusted accordingly.

Initial fluid resuscitation to treat shock should be with 20 mL kg^{-1} of isotonic (normal) saline. Thereafter, isotonic fluids should be given to maintain systemic blood pressure (and thereby cerebral blood flow). Previous guidelines have emphasised the importance of fluid restriction.[22] It may be necessary to restrict fluids if the serum sodium is <130 mmol L^{-1} or if there are signs of fluid overload. However, fluid restriction does not generally improve outcome[23] and has even been associated with a worse neurological outcome.[24] Thus, most children should receive normal maintenance fluid volumes.

Prevention

Conjugate pneumococcal and meningococcal vaccines should have a continuing impact on the number of cases of childhood meningitis.

Contacts of patients with meningococcal meningitis may require chemoprophylaxis to prevent secondary spread. Those who should receive chemoprophylaxis include:

- the index case if treated only with penicillin (does not eradicate carriage)
- all intimate, household or day-care contacts who have been exposed to the index case within 10 days of onset
- any person who gave mouth-to-mouth resuscitation to the index case or had direct contact with his/her airway secretions.

Although nosocomial transmission of meningococcal infection is rare, droplet precautions are recommended until the patient has received 24 hours of antibiotic therapy. As the aetiology is usually unknown during this period, all cases of suspected bacterial meningitis should be managed in this way. Preferred placement is in a single room. However, if a spatial separation of >1 m can be achieved and curtains can be drawn between the infected patient and other patients and visitors, this may be sufficient.

Chemoprophylaxis

Rifampicin 10 mg kg^{-1} (5 mg kg^{-1} <1 month old) orally 12-hourly (max 600 mg) for 2 days *or*

Ceftriaxone 125 mg (≤12 years)/250 mg (>12 years) mg intramuscularly as a single dose *or*

Ciprofloxacin 500 mg orally as a single dose.

Complications

Bacterial meningitis is associated with a 4.5% mortality rate and intellectual, cognitive and auditory impairment in 10–20% of survivors.[25] The risk for sequelae is greatest in those who experience acute neurological complications at the time of their illness.[26]

Viral meningitis

Most cases are self-limiting, and treatment is symptomatic.

Brain abscess

Brain abscess classically presents with fever, headache and focal neurological deficit. Although rare, early recognition is vital, as early antibiotic treatment and drainage improve outcome. Diagnosis is by cerebral CT or magnetic resonance imaging (MRI). Aspiration for diagnosis and neurosurgical intervention is usually required. The most common causes are oral viridans streptococci, anaerobes, gram-negatives and *S. aureus*.

Empiric treatment is:

Flucloxacillin 50 mg kg^{-1} (max 2 g) IV 4-hourly ***plus***
Ceftriaxone 100 mg kg^{-1} (max 2 g) IV daily or 50 mg kg^{-1} (max 1 g) IV 12-hourly ***or***
Cefotaxime 50 mg kg^{-1} (max 2 g) IV 6-hourly ***plus***
Metronidazole 15 mg kg^{-1} (max 1 g) IV stat, then 7.5 mg kg^{-1} (max 500 mg) IV 8-hourly.

Encephalitis

Aetiology

Multiple infectious agents have been associated with encephalitis, but the syndrome is an uncommon manifestation of most. Viruses are the most commonly identified cause.[27] Immune-mediated aetiologies are increasingly recognised in up to one-third of cases and are important because they are often treatable. These include acute disseminated encephalomyelitis (ADEM), primarily seen in children under 15 years of age, and antibody-mediated encephalitides (e.g. anti-N-methyl-D-aspartate receptor [NMDAR] and anti-voltage-gated potassium-channel [VGKC] complex). The aetiology is not identified in the majority of cases.[27]

Viral causes of encephalitis include:

- herpes simplex viruses 1 and 2 (HSV-1 and HSV-2)
- other herpes viruses – human herpesviruses (HHV)-6, 7 and 8, varicella-zoster virus (VZV), cytomegalovirus and Epstein–Barr virus. HHV-6, 7 and 8 are usually associated with febrile seizures and with encephalitis in the immunocompromised.
- enteroviruses and parechovirus

- influenza virus
- adenovirus
- other viruses causing seasonal epidemics of encephalitis in tropical areas of Australia, e.g. Japanese encephalitis virus, Murray Valley encephalitis virus, Kunjin virus, etc.

HSV-1 and HSV-2 are perhaps the most important causes to consider, as encephalitis caused by these viruses is associated with high morbidity and mortality, which may be reduced with early treatment with antiviral medications.

Mycoplasma pneumoniae has been associated with encephalitis, although causality remains controversial without concurrent pathogen identification. *M. pneumoniae* may well cause an immune-mediated encephalitis.

Clinical findings

There is considerable overlap between the features of meningitis and encephalitis, and the two frequently coexist. Encephalitis should be suspected if there is encephalopathy, that is, a change of behaviour, altered conscious state or extreme drowsiness. Convulsions (particularly focal) and focal neurological deficits are also more common in encephalitis. Children with ADEM typically have encephalopathy associated with multifocal neurological symptoms and signs.

Investigations

- CSF:
    - Collection of 5–10 drops into each of two numbered sterile tubes is usually adequate. To enable more extensive testing in older children, up to 10 mL of CSF may be required
    - Measure opening pressure
    - Biochemistry and microscopy findings are non-specific (see Table 8.7.1)
    - PCR testing for HSV, enterovirus and VZV is sensitive and specific
    - Oligoclonal bands
    - Cryptococcal antigen.
- Serology – may be helpful to collect serum for future testing, particularly

in case IVIG is given
- Nasal swab – PCR testing for respiratory viruses may be helpful
- Stool viral testing – PCR or antigen for enterovirus, adenovirus, rotavirus
- MRI of the brain and EEG may be suggestive of HSV encephalitis, particularly if they show temporal lobe involvement. MRI brain and spine showing multiple demyelinating lesions.

Management

It is vital to consider the diagnosis of HSV encephalitis early, because **early** treatment with aciclovir may improve the outcome (Box 8.7.2). Treatment should be continued for 21 days unless HSV is excluded clinically or on subsequent investigations. Corticosteroids have a role in the management of ADEM, and IVIG is often used as adjunctive therapy for infectious and immune-mediated encephalitis. There may be a role for neuraminidase inhibitors (e.g. oseltamivir) for the treatment of encephalopathy associated with influenza infection.[28] There is no evidence that antimicrobials are beneficial in *M. pneumoniae*-associated encephalitis.[27]

Box 8.7.2 Aciclovir for treatment of suspected or proven HSV encephalitis.

Aciclovir

20 mg kg^{-1} IV 8-hourly (age <3 months)
500 mg m^{-2} IV 8-hourly (age 3 months–12 years)
10 mg kg^{-1} IV 8-hourly (age >12 years)

Complications

Overall mortality of encephalitis is approximately 10%. Without treatment, mortality of HSV encephalitis is up to 80%, and 50% of survivors have long-term sequelae.[29] Outcome is worse if coma is present initially.

Conclusion

Meningitis and encephalitis are medical emergencies that must be assessed and treated urgently to reduce the likelihood of poor outcomes.

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SECTION 9

Infectious Diseases

OUTLINE

9.1

Infectious diseases

Mike Starr

ESSENTIALS

1 Fever is one of the most common reasons for children to present to the emergency department (ED).

2 Serious bacterial infections need to be identified early and treated aggressively; these include meningitis, septicaemia, urinary tract infection (UTI), pneumonia, and bone and joint infections.

3 The majority of febrile children have a viral illness, requiring few if any investigations.

4 The risk of serious bacterial infection in well-appearing febrile children is low overall, and UTI makes up 92% of causes.

5 Most well-appearing febrile children over 1 month of age without a focus of infection do not require laboratory testing or treatment, apart from microscopy and culture of urine.

6 Provided close follow-up is assured, clinical judgment can be employed for well-appearing febrile infants, rather than investigating all such infants according to guidelines.

7 Targeted and judicious use of laboratory investigations facilitates more rapid and accurate diagnosis of causes of infection in children presenting to the ED.

8 The risk of acquisition of a blood-borne virus from a community-acquired needlestick injury is negligible.

In this section, general infectious disease issues, including the appropriate collection of microbiological specimens, guidelines for

empiric antibiotic therapy, post-exposure prophylaxis and immunisation, are addressed.

Fever

Fever is one of the most common presenting complaints in children in both the primary care and emergency department (ED) settings. Of all children's visits to the ED, up to 30% are with acute episodes of fever.[1–3] In children <1 year old presenting to EDs in Australia and New Zealand, fever without identifiable source is the diagnosis in over 3%.[3] In the first 2 years of life, children average four to six febrile episodes. Those in child care may have many more than this.

Defining and measuring temperature

There is controversy regarding the most appropriate thermometer and the best anatomical site for temperature measurement.[4] Parents often use touch to detect fever in their children. However, touch has only 50% specificity.[5] It tends to overestimate the incidence of fever and is more useful to exclude fever. Rectal temperature has long been considered the gold standard for routine measurement of body temperature, but it does not in fact reflect true core temperature within the pulmonary artery. Moreover, parents and patients generally prefer other temperature assessments. Nonetheless, rectal temperature remains the most widely used measure in infants under 3 months of age. Tympanic thermometers provide the most accurate assessment of core temperature, but the probe may be too large for an infant's auditory canal. Measuring oral temperature requires patient cooperation and is generally unsuitable for children under the age of 5 years. Axillary temperature measurement is inaccurate and insensitive.

The definition of fever is:

- 38°C (rectal or tympanic)
- 37.5°C (oral) or
- 37.2°C (axilla).

Fever: to treat or not to treat?

The drugs most commonly used for treating fever are paracetamol, ibuprofen

and aspirin. The routine use of these medications in the treatment of fever has been questioned.[6] In particular, there has been concern that the use of antipyretics may prolong viral shedding, impair antibody response to viral infection, and may increase morbidity and mortality.[6–8] Moreover, each of the commonly used antipyretics may have significant adverse effects such as hepatic dysfunction, metabolic acidosis, Reye syndrome and gastrointestinal bleeding. Treatment should therefore be focused on alleviation of discomfort or pain rather than on the height of the temperature. Either paracetamol or ibuprofen may be used. There is some evidence that both alternating and combined antipyretic therapy may be more effective at reducing temperatures than monotherapy alone.[9] However, the evidence for improvements in measures of child discomfort remains inconclusive. It is important to note that the use of antipyretics has not been shown to prevent febrile convulsions.

Paracetamol may be given orally, rectally or intravenously at a dose of 10–15 mg kg^{-1} 4–6 hourly. In an unsupervised, community setting, the total daily dose should be limited to 60 mg kg^{-1}, although up to 90 mg kg^{-1} per 24 hours can be used under medical supervision. Single doses of 30 mg kg^{-1} may be used for night-time dosing. Serious toxicity has been reported in children with chronic daily over-dosage, mostly occurring in children who have a febrile illness and associated anorexia, vomiting and/or dehydration.[10] A child should be reviewed after 48 hours if regular paracetamol has been ‘required’ for this period.

Ibuprofen can be used as an alternative to paracetamol at a dose of 5–10 mg kg^{-1} (maximum of 400 mg per dose), given 6–8 hourly (maximum daily dose of 30 mg kg^{-1} or 2 g). It is recommended that it be used alone and not in combination with paracetamol, as this practice may lead to an increase in adverse effects, including gastrointestinal bleeding, renal dysfunction and anaphylaxis.[8] A theoretical risk of aggravating concurrent asthma has also been described, although these adverse effects are refuted in large prospective studies.[10] There is also a concern that ibuprofen may be associated with an increased risk of necrotising group A streptococcal infections.[11]

Practical approach to the febrile child

The majority of febrile children will have self-limiting viral infections. The challenge is to identify those children at risk of serious illness while avoiding unnecessary investigation and treatment of children with benign viral illnesses.[12]

Fever in children may be classified into three groups:

- Fever with localising signs
- Fever without focus
- Pyrexia of unknown origin (PUO).

Fever with localising signs

A careful history and examination will identify the source of infection in most patients. These children should be managed according to the individual condition and its severity.

Fever without focus

In a small number of children presenting with fever, no focus is found. While most will have a viral infection, a more serious illness such as a urinary tract infection (UTI) (up to 8%), occult bacteraemia (<1%) or meningitis (<0.2%) may be present.

Occult bacteraemia is the presence of bacteria in the bloodstream of a febrile child who has no apparent focus of infection and looks well. Diagnosis is by blood culture and exclusion of focal infection. The incidence of occult bacteraemia in febrile children has reduced dramatically to approximately 0.25% since the introduction of conjugate pneumococcal vaccines.[13–15]

Most children who present with fever and no identifiable focus appear otherwise well. History should include details about immunisation status, infectious contacts, travel, diet and contact with animals or insects. A thorough physical examination should be performed, paying particular attention to general appearance (colour and level of activity) and vital signs (respiratory rate, pulse, peripheral perfusion and blood pressure).

It is difficult to assess whether a child is ‘septic’ or ‘toxic’. A simple and effective approach that is useful in the ED is a combination of ABC, fluids in and fluids out.[16] An infant with one or more of these symptoms or signs has a higher risk of serious illness:

- A – poor arousal, reduced alertness and reduced activity
- B – breathing difficulty
- C – poor perfusion
- Fluids in – the frequency of feeding over the 24 hours prior to presentation, <50% of normal over 24 hours prior to presentation,

suggesting dehydration
- Fluids out – significantly abnormal urine output of <50% of usual output.

Other features on examination that strongly suggest a seriously ill infant include pallor or cyanosis, purpuric rash, high-pitched scream and bulging fontanelle.

Patients with unexplained fever with a higher likelihood for serious infection include the following patient groups or conditions:

- Neonates (<1 month of age)
- Incompletely immunised children
- Immunocompromised (e.g. congenital immunodeficiency, human immunodeficiency virus (HIV), neutropenic and other oncology patients, cytotoxic drugs and steroids)
- Asplenic children (congenital, post splenectomy or functional, e.g. sickle cell disease)
- Patients who have received prior oral antibiotics; many of these patients have a viral infection, but meningitis or other serious bacterial infection (SBI) must be considered
- Children with fever and prolonged convulsion
- Children with underlying medical conditions (e.g. cystic fibrosis, structural cardiac defects, etc.)
- Children with central venous devices, shunts or other foreign material.

When considering management strategies for febrile infants, three age groups are generally assigned: <1 month of age, 1–3 months and >3 months (Table 9.1.1). In infants less than 1 month of age and those with any of the risk factors above, there should be a low threshold for performing investigations including full blood count, culture of blood, urine and cerebrospinal fluid (CSF), and a chest X-ray if indicated. Empiric antimicrobial therapy should be based on the patient's clinical illness, risk factors, and the local epidemiology of potential pathogens and their antibiotic susceptibility.

Many guidelines for the management of infants with fever include the use of various markers to assist in identifying those at low (or high) risk of SBI. White cell count (WCC) is used in most guidelines as a screening tool. However, studies performed since the introduction of conjugate pneumococcal vaccine have shown that WCC is neither sensitive nor specific as an indicator of

bacteraemia and other SBI in infants. A WCC threshold of 15 x 10^9 L misses half of all SBI while misclassifying a quarter of self-limiting illnesses. Van den Bruel et al. performed a systematic review of the diagnostic value of various laboratory tests in identifying SBI in febrile children. They found that the tests providing most diagnostic value were C-reactive protein (CRP) and procalcitonin (PCT). However, they found few studies, and none were of high methodological quality. Moreover, neither CRP nor PCT was found to have sufficient diagnostic value to either confirm or exclude SBI; the results must be interpreted in the light of clinical findings.

Clinical scores, such as the Rochester, Philadelphia and Boston criteria, have been devised to identify children at low risk of serious bacterial infection.[17] However, their utility has been questioned in the era of widespread conjugate Hib, pneumococcal and meningococcal vaccination.

Febrile infants between 1 and 3 months of age who appear well and do not have risk factors may not require blood tests or a lumbar puncture, although urine microscopy and culture are advisable. Those over 3 months of age do not routinely require laboratory testing or treatment, although urine microscopy and culture may still be appropriate.

Table 9.1.1

Management of well-appearing febrile child without focus

Age	Investigation	Management
<1 month	Low threshold for blood, urine and CSF cultures; FBC; CXR	Admit Empiric IV antibiotics: Amoxicillin plus 3GC If herpes simplex infection is suspected, add aciclovir
1–3 months	Urine culture	Consider admission and observation Discharge with arranged review
>3 months	Consider urine culture	Discharge with arranged review

CSF, cerebrospinal fluid; FBC, full blood count; CXR, chest X-ray; 3GC, third-generation cephalosporin.

There is no evidence that oral or parenteral antibiotics prevent the rare occurrence of focal infections from occult bacteraemia; instead, they result in delayed diagnosis, drug side effects, additional costs and the development of resistant organisms. What are required are a careful clinical assessment, parental education and review within 24 hours.

As UTI is the most common serious bacterial infection among febrile infants

and children, urine microscopy and culture should be included in the investigation of most such children. In infants, a urine sample should ideally be obtained via suprapubic aspiration or catheter. A negative urinalysis does not exclude a UTI, which may occur in the absence of pyuria.[18,19] In well-appearing infants, there is generally no need to perform lumbar puncture once a UTI has been diagnosed.[12]

Other rarer causes of fever should also be considered:

- Kawasaki disease (see Chapter 5.9)
- Connective tissue disease (e.g. juvenile arthritis)
- Inflammatory bowel disease
- Malignancy (e.g. leukaemia).

Pyrexia of unknown origin

Pyrexia of unknown origin (PUO) is defined as prolonged fever (usually greater than 10 days) where history, examination and 'routine' investigations have not identified a cause. Occasionally, fever appears to occur in a repetitive or periodic pattern. In either case, a chronic or non-infectious condition should be considered, such as juvenile idiopathic arthritis and other connective tissue diseases, inflammatory bowel disease or malignancy. Infectious causes include systemic viral syndromes (such as infectious mononucleosis), upper or lower respiratory infections (e.g. sinusitis), UTI, central nervous system infection, bone infection, tuberculosis, abscess (e.g. parameningeal, intra-abdominal), endocarditis and enteric infections (e.g. typhoid fever).

More extensive investigations are often required, including specific investigations for mycobacterial, fungal or viral infections. Imaging may be required, looking for occult abdominal or central nervous system collection, for osteomyelitis or endocarditis.

Kawasaki disease (see Chapter 5.9) is an important consideration in an infant or child presenting with prolonged fever, and diagnosis is often delayed. There is a degree of urgency in diagnosis, because treatment within 10 days of onset of fever with intravenous immunoglobulin and aspirin reduces the incidence of coronary artery lesions from around 20% to around 5%.[20]

Empiric antibiotic therapy

With the possible exception of bacterial meningitis, where Gram stain results may guide therapy, the most appropriate antibiotic therapy in children must be based on epidemiological grounds. The most important factors determining the likely pathogens, which should be targeted by empiric therapy, are:

- age
- presumed focus of infection
- presence of underlying disease or anatomical abnormality
- whether the infection is hospital or community acquired.

In addition, the site of infection may have implications for the expected penetration of the antibiotic chosen (e.g. aminoglycosides do not penetrate into abscess cavities and are inactive in an anaerobic environment).

For presumed bacterial infection (including meningitis) in the first 3 months of life, empiric treatment must cover Group B streptococci, *Escherichia coli* and *Listeria monocytogenes* infections. Recommended antibiotics are amoxicillin plus a third-generation cephalosporin (cefotaxime or ceftriaxone). Ceftriaxone should be avoided in neonates, particularly if <41 weeks, gestation, jaundiced or receiving calcium containing solutions, including total parenteral nutrition.

Amoxicillin	50 mg kg^{-1} per dose intravenous (IV) 12 hourly (week 1 of life), 6 hourly (week 2–3 of life), 4–6 hourly thereafter.
Cefotaxime	50 mg kg^{-1} per dose (max 2 g) IV 8 hourly (week 1 of life), 6 hourly (week 2–4 of life), 4–6 hourly thereafter.
Ceftriaxone	100 mg kg^{-1} per dose (max 2 g) IV/IM once daily or 50 mg kg^{-1} per dose 12 hourly.

Herpes simplex virus infection should be considered in febrile neonates who present with seizures, vesicular rash or septic shock, and aciclovir added to the empiric antimicrobial regimen.

For presumed bacterial infection (including meningitis) after 3 months of age, potential pathogens include *Neisseria meningitidis*, *Streptococcus pneumoniae*, Group A streptococci and *Staphylococcus aureus*. Recommended empiric therapy is flucloxacillin plus a third-generation cephalosporin.

Flucloxacillin	50 mg kg^{-1} per dose (max 2 g) IV 12 hourly (week 1 of life), 8 hourly (week 2 to 4 of life), 6 hourly thereafter.
Cefotaxime/Ceftriaxone	As above

If meningitis has been excluded, recommended antibiotics are flucloxacillin plus gentamicin.

Gentamicin	7.5 mg kg^{-1} 24 hourly (≤10 years), 6 mg kg^{-1} per dose 24 hourly (>10 years)

Antibiotic choice should also be modified once relevant culture results become available.

Antimicrobial resistance

Antimicrobial resistance is an increasing worldwide problem.[21,22] Resistance of bacteria to antibiotics can conveniently be divided into two categories:

1. Intrinsic – resistance due to inherent properties of the organism or antibiotic
2. Acquired – resistance gained by the organism either before or during therapy.

Important examples of intrinsic resistance (which affects 100% of organisms) include enterococci to cephalosporins, enteric gram negatives to penicillin and flucloxacillin and anaerobes to aminoglycosides and quinolones. A few important examples of increasing acquired resistance include:

- penicillin (and cephalosporin)-resistant *Strep. pneumoniae* (PRP) – of note, resistance rates have fallen since introduction of the conjugate vaccine
- methicillin (multidrug)-resistant *Staph. aureus* (MRSA)
- *Staph. aureus* with intermediate sensitivity to vancomycin and teicoplanin (GISA)
- community-acquired non-multiresistant MRSA (CA-MRSA)
- vancomycin-resistant *Enterococcus* (VRE)
- bacteria that produce extended-spectrum β-lactamases (ESBL), e.g. some *E. coli* and *Klebsiella* spp., which are associated with cephalosporin (and often gentamicin) resistance
- carbapenem-resistant Enterobacteriaceae.

Common infectious exanthems

Most frequently, the cause of fever in children is a viral illness. This usually occurs in a seasonal pattern – in Australasia particularly the months of April through to September (autumn and winter) when there is an increase of acute

infections in the community. Most of these are due to respiratory and gastrointestinal pathogens, such as respiratory syncytial virus (RSV) and rotavirus, respectively. However, there are several viral infections that commonly present with fever and rash which emergency physicians need to be familiar with.

A rash or other cutaneous manifestation accompanies a large number of presentations for childhood infectious disease. An exanthem is an acute infectious disease accompanied by a rash. The most common childhood exanthems are scarlet fever, measles, rubella, erythema infectiosum and roseola infantum. Rash and fever may be associated with many other viruses, bacteria, and even parasitic infections. In addition, a rash and fever may also be associated with a wide variety of non-infectious processes. See Chapter 12 on Dermatology for further details.

What specimens, when should they be ordered and what tests?

Targeted and judicious use of laboratory investigations facilitates more rapid and accurate diagnosis of causes of infection in children presenting to the ED. Microscopy, culture, biochemical testing and antibody detection by enzyme immunoassay (EIA) and enzyme-linked immunosorbent assay (ELISA) are a few of the first-line investigations that have been used for some time to diagnose infections. However, a number of emerging technologies enable the detection and quantification of pathogens with enhanced speed, sensitivity, and ease of use;[12,23] real-time polymerase chain reaction (PCR) and other nucleic acid–based amplification technologies (NAATs) detect microbial constituents with high sensitivity and specificity. Molecular tests are now used routinely for the diagnosis of many infections including RSV, enterovirus, herpes simplex virus, *Bordetella pertussis* and *Neisseria meningitidis*.

Multiplex PCR platforms are designed to probe respiratory specimens, stool samples and even blood for an array of pathogens; these are increasingly being used for point of care testing. Matrix-assisted laser desorption ionisation: time of flight mass spectrometry (MALDI-ToF MS) is a newer technology being increasingly used for the rapid identification of bacteria, fungi, mycobacteria, and parasites.[24] Bacterial whole-genome sequencing is on the horizon as a rapid diagnostic microbiology tool.

Collection of microbiological specimens

It is imperative that microbiological specimens be collected appropriately, taking care not to contaminate samples during collection. Samples should also be transported to the laboratory promptly, stored in appropriate laboratory environments and processed without undue delay.

Blood cultures

Collection

Proper hand-washing technique and use of gloves will avoid contamination of the sample. Peripheral blood cultures are usually collected from veins, either direct puncture or immediately aspirated from a cannula after insertion. Arterial samples may also be used, but heel prick samples are inappropriate. It is imperative to disinfect the patient site for blood collection, usually with topical aqueous chlorhexidine or large alcohol swabs and to allow the disinfecting agent to dry, which is an important part of the disinfection process. Swab the rubber bung on the bottle with an alcohol swab, and allow drying time before introducing blood into the bottle. Blood for cultures should be collected first, before placing blood into other, non-sterile bottles for additional investigations. Avoiding needle changes to inoculate the culture bottle minimises the risk of an accidental needlestick injury.

Volume of blood and collection media

It is important to remember that a minimum amount of blood must be collected but also that more than a maximum amount of blood will decrease the sensitivity of the culture and thus the likelihood of identifying a microorganism. The optimum blood-to-culture broth dilution is 1:10 in most blood culture systems, and up to 5 mL of blood can be inoculated into older systems but from 0.4 mL to 4 mL in newer collection media. Samples for anaerobic bacterial cultures are not routinely collected, with few exceptions:

- Febrile neutropenic, immunocompromised patients
- Patients with suspected intra-abdominal or pelvic sepsis
- Neonates with a high risk of anaerobic sepsis, i.e. those with prolonged rupture of membranes, offensive liquor, or maternal fever in labour.

Blood culture bottles should be correctly labelled and optimally stored in an appropriate incubator (never in a fridge).

Cerebrospinal fluid

See Chapter 8.7 and also Chapter 23 on Common procedures.

Urine

A sterile urine sample enables a more accurate diagnosis of UTI, and as such the method of collection significantly influences the accuracy of the microscopic results obtained. If UTIs are overdiagnosed, other important diagnoses may be missed, and children may be subjected to unnecessary further investigations. A negative urinalysis does not exclude a UTI, and up to 10% of UTIs may be missed. The presence of white or red blood cells or protein does not either confirm or refute the diagnosis. All urine specimens in infants <2 months of age or those with positive urinalysis (either nitrates or leucocyte esterase positive) or in those with significant symptoms should be sent for microscopy and culture if a UTI is suspected or if obtained via catheter/suprapubic aspirate in non-toilet-trained children. The laboratory should always be informed of the collection method as this influences the interpretation of results.

Bag urines

Urine collected is usually contaminated, and testing is neither sensitive nor specific for culture-positive UTIs.

Clean catch and midstream collection

Clean catch and midstream collection are both non-invasive, and contamination is less likely, but care must be taken to reduce contamination. The ‘best’ clean catch possible is obtained from boys with the foreskin retracted and girls with the labia spread after cleansing with soap and water and gentle drying. The patient or parent should avoid touching skin, and the specimen should be collected in midstream.

Suprapubic aspiration

This is the most reliable means of collection of a sterile specimen, but the infant

usually needs to have a reasonably full bladder. Bladder scanning/ultrasound improves the yield. The technique is safe.

Catheterisation

Quick ‘in-out’ urinary catheterisation is a reasonably reliable collection method and does not require the bladder to be filled. However, contamination of the specimen can occur. It is very unlikely that infection can be introduced with an in-out procedure that is done with sterile technique. Only mild discomfort is usually experienced. Catheterisation should not be attempted in girls with significant labial adhesion nor in boys with phimosis that prevents vision of the urethral meatus.

Interpretation

Reagent-impregnated dipstick

A dipstick urinalysis can be used to detect the presence of nitrite-forming bacteria or estimate the presence of pyuria using a leucocyte esterase test strip, thereby allowing a quick presumptive diagnosis of a UTI and early treatment in an unwell child. Blood, protein and ketones can also be detected. Dipstick urinalysis positive for either nitrates or leucocyte esterase has a sensitivity of 93% (90–100%) and a specificity of only 72% (58–91%).[18] Non-nitrite-forming bacteria such as *Enterococcus* spp. may also cause UTIs, and pyuria is not always present in UTIs, which makes a UTI infection still possible following a negative urinalysis.

Microscopy

A urinary white cell count $>10 \times 10^6$ L^{-1} together with a positive leucocyte esterase is a very sensitive screening test for UTIs but still has a false-negative rate of 15%. The presence of >10 leucocytes mm^{-3} on direct microscopy has been shown to have a low positive predictive value (56%) for UTI, but, combined with the presence of bacteria, this constitutes the most accurate screening test in detecting positive urinary cultures.[19]

Culture

UTI is ultimately diagnosed on demonstration of significant bacteriuria, which implies counts of $>10^8$ L^{-1} of a single pathogen in a fresh, uncentrifuged clean catch or midstream urine sample. Any growth on a sample obtained by urinary

catheter or suprapubic aspirate is also diagnostic.

Stool specimens

Viral gastroenteritis is the most common cause of paediatric hospital attendance during the spring and early summer months. The organism involved is usually rotavirus, which is usually self-limiting and is readily detected by enzyme-linked immunoassay or latex agglutination. Laboratory analysis is usually not warranted during seasonal endemic periods as most causes of diarrhoea are usually self-limiting, and identification will not alter management in most cases. Indications for stool analysis are:

- persistent diarrhoea >7 days
- blood in stool
- recent overseas travel, particularly to typhoid endemic areas
- immunocompromised children.

Collection

Diarrhoeal stool (5–10 g) should be collected in a sterile container with a secure fitting lid. Testing the stool pH using a reagent strip may be a useful indicator for rotavirus gastroenteritis as it causes an acidic stool (pH <5). Similarly, testing stool for reducing substances may be positive in cases of secondary lactose intolerance.

Throat swab

Throat swabs may be performed both for bacterial and viral culture, although for the latter, a nasal swab or nasopharyngeal aspirate is usually more helpful. Guidelines exist for those population groups at risk of rheumatic fever (see Chapter 5.7).

Nasopharyngeal aspirate

Nasopharyngeal aspirate (NPA) was the preferred method for recovering viruses from the upper respiratory tract for many years. However, the test was distressing for infants (and parents), and the results rarely affect patient management. Newer molecular techniques are performed on nasal swabs.

Interpretation

Most respiratory viruses can be detected by direct immunofluorescence of exfoliated respiratory epithelium, as viral antigens are expressed on the cell surface. If available, this allows rapid diagnosis of infection. Viruses can also be cultured. The positive and negative predictive values of both NPA and nasal lavage in diagnosing RSV infection are >90%. *Bordetella pertussis* may be identified by PCR if a definitive diagnosis is required before culture results of NPA are available.

Nasal swab

A nasal swab is the classic method for collection of diagnostic specimens for molecular diagnosis of pertussis and a number of other respiratory pathogens.

Collection

A small-tipped nasopharyngeal swab is passed into the posterior nasopharynx. Do not use a swab with a cotton tip as this may be inhibitory to the organism. The swab is left in place for at least 30 seconds, if possible.

Infection control in the emergency department

Adhering to strict universal precautions (i.e. treating all blood and body fluids as potentially infectious) is an integral part of containing the spread of infections in any healthcare setting. This is essential nowhere more so than in paediatric EDs, where communicable viral respiratory and enteric infections are the most common causes of presentations. Cross-infection can be eliminated by strict hand-washing measures by both staff and parents, as well as avoiding toy sharing by patients in ward situations. Prophylaxis with normal or specific human immunoglobulin is sometimes indicated following significant exposure to communicable diseases, such as hepatitis A and B, measles and varicella, and antibiotic prophylaxis may be indicated for significant exposure to meningococcal and *Haemophilus influenzae* type b disease.

Needlestick injury

The child presenting with a (community)

needlestick injury

The risk of seroconversion to HIV, hepatitis B virus (HBV) or hepatitis C virus from a community-acquired needlestick injury (NSI) is almost zero.[25] Exposed individuals should be reassured. Immunity to hepatitis B should be confirmed and, if incomplete, hepatitis B vaccine should be given. Unless the injury is considered to be particularly high risk, no further management is required at the time. Follow-up should be arranged for counselling and serology if required.

Hospital staff exposure to blood-borne viruses

In general, NSIs occur more often in the hospital environment, where a lack of universal precautions in handling or discarding contaminated needles and sharps has occurred. It is advisable that all staff working in the hospital environment should be adequately immunised against hepatitis B and that serological confirmation of immunity is performed. If significant exposure (percutaneous, ocular, or mucous membrane) to blood or potentially blood-contaminated secretions has occurred in someone who is unimmunised, testing of the source for hepatitis B surface antigen (HBsAg), hepatitis C virus antibody and human immunodeficiency virus antibody should be performed following appropriate consent. The recipient should have blood taken for hepatitis B surface antibody (HBsAb) and for storage for future baseline testing for other blood-borne viruses if required. A single dose of hepatitis B immunoglobulin (HBIG 400 IU for adults and 100 IU for children) is then offered to the recipient if HBsAb negative, or if the source is HBsAg positive or cannot be identified. This should be given within 72 hours after exposure.

Immunisation

Immunisation of staff

It is in the best interests of all healthcare workers, and their patients, that staff in paediatric centres have a serologically proven immunisation record against communicable illnesses such as measles, mumps and rubella, varicella and HBV and receive the relevant immunisation as appropriate. Pertussis-containing booster and annual influenza vaccine should also be given.

Opportunistic immunisation

The ED visit also presents an invaluable opportunity to monitor the immunisation status of children and offer ‘catch-up’ immunisation to those who have missed vaccinations or commence the appropriate vaccination schedule.

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SECTION 10

Metabolic Emergencies

OUTLINE

10.1

Inborn errors of metabolism

Drago Bratkovic, and Niki Talić

ESSENTIALS

1 Inborn errors of metabolism are individually rare but as a group are regularly seen in the emergency department (ED).

2 The possibility of an inborn error of metabolism needs to be considered in any child with unexplained hypoglycaemia, acidosis, altered conscious state, neurological presentation or vomiting.

3 Prompt recognition is important to allow appropriate therapy and to avoid further decompensation.

4 Precise identification of the defect in the ED is not generally important so long as appropriate initial therapy is commenced, and this usually consists of dextrose.

Introduction

Inborn errors of metabolism (IEM) are a diverse group of disorders that result from a defect or absence of an enzyme or transport system. The presenting symptoms and signs are equally diverse; however, there are a number of common features that can alert the clinician to their presence. This chapter will concentrate on those IEM that present acutely to the emergency department (ED) but will also touch on those with more chronic presentations which could easily go unrecognised.

The majority of patients presenting with acute IEM to the emergency department will have already been diagnosed, particularly given the advent of

extended newborn screening (see Extended newborn screening at the end of chapter). However, we are unable to screen for all acutely presenting IEM, and some will be missed by screening, thus children and, indeed, adults still present acutely with an undiagnosed IEM.

Physiology and pathogenesis

The process by which living matter is built up (anabolism) or broken down (catabolism) is termed *metabolism*. Strictly speaking, an IEM is an inherited defect in a metabolic pathway or enzyme. Most commonly IEM result from a deficiency in an enzyme that catalyses the conversion of one organic (substrate) compound to another (product); this can be quantitative (less enzyme), qualitative (lower activity of the enzyme) or both. However, it is important to remember that not all defects in metabolic pathways give rise to pathology. Fig. 10.1.1 shows a typical enzymatic reaction where compound A is converted to B. A reduction in the activity of the enzyme can cause pathology via a number of mechanisms:

1. There is accumulation of the substrate A, which is toxic onverted to B. A reduction
2. There is a deficiency of the product B A, which is toxic converted to B. A reduction in t fatty acid oxidation disorders.
3. Excess substrate A is converted to another compound C via an alternate pathway which is toxic acid oxidatyrosinaemia type 1.
4. If earlier reactions are reversible then accumulation of substrate A can also result in accumulation of an earlier metabolite, E, in the pathway, which is toxic – example: maple syrup urine disease.
5. Excess A is removed by reacting/conjugating with compound D, which results in deficiency of D lation of an earlier metabolite, E, in the pathway, which is tts.

The pathology seen in an IEM can result from any of the processes outlined in Fig. 10.1.1, and in reality most IEM are the result of more than one mechanism. It is also worth remembering that in some situations an IEM is not caused by mutations in the genes for the enzyme but in genes that support the function of the enzyme such as transport proteins, chaperones and assembly proteins, which ultimately result in enzyme deficiency. The best examples of these are the

mitochondrial respiratory chain disorders where mutations in genes have been found in a number of these support processes.

Clinical features

The clinical features and presentations of IEM are many and varied due to the diverse nature of the enzymes and processes affected; however, there are a number of common features that should alert the clinician to the possibility of an IEM.

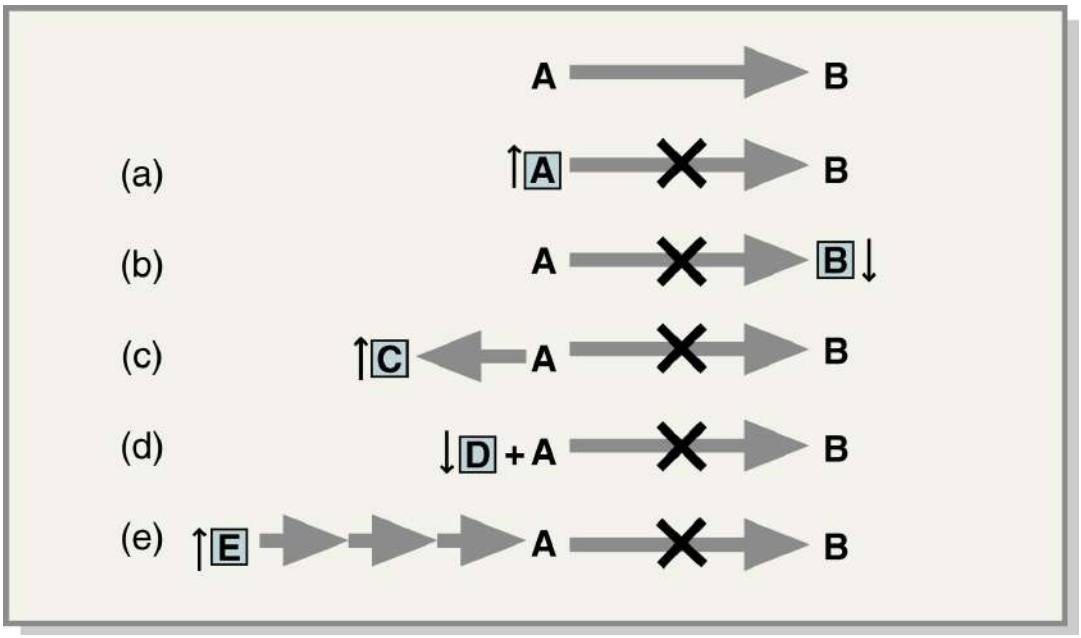

FIG. 10.1.1 Pathogenic mechanisms in inherited errors of metabolism; refer to text for a full explanation.

Table 10.1.1

Groups of inherited errors of metabolism that present to the emergency department, their common clinical and biochemical features, and further recommended investigations

Defect/disorder	Clinical clues	Biochemical clues*	Further investigations
Organic aciduria – IVA, PA, MMA	Vomiting Encephalopathy Seizures (late)	Metabolic acidosis – variable Elevated NH3 – mild to moderate (Hypoglycaemia)	Plasma AA, UO&AA ACP
Maple syrup urine disease	Vomiting Encephalopathy Maple syrup-like smell Seizures (late)	(Metabolic acidosis) (Hypoglycaemia)	Plasma AA
Urea cycle disorder – OTC, ASA, citrulinaemia	Vomiting Encephalopathy Hyperventilation Seizures (late)	Ammonia >150 mmol L Respiratory alkalosis - variable	Plasma AA UO&AA Urine orotate
Fatty acid oxidation disorders – MCAD, VLCAD, LCHAD	Previously well Muscle pain Encephalopathy Cardiomyopathy	Hypoketotic hypoglycaemia Elevated CK – VLCAD, LCHAD Elevated LFTs – acute	UO&AA ACP
Disorders of ketone utilisation Ketotic hypoglycaemia	Previously well Vomiting – infectious cause	Metabolic acidosis – pH <7.2 β-hydroxybutyrate >4.0 mmol L (ward test)	Plasma ketones, UO&AA ACP
GSD (liver) - I, III, IV, VI and IX	Hepatomegaly Frequent feeding Seizures/drop attacks Delayed development	All - hypoglycaemia GSD I – lactic acidosis GSD III – elevated CK GSD IV, VI, IX – ketotic hypoglycaemia	Urate Triglycerides Cholesterol Genetic studies
GSD (muscle) - V, VII, X	Muscle pain Exercise intolerance Dark urine	Acute rhabdomyolysis with grossly elevated CK	Genetic studies
Mitochondrial respiratory chain disorder	Encephalopathy Cardiomyopathy Stroke-like events	Lactic acidosis (Elevated LFTs) (Elevated ammonia)	UO&AA, pre- and post-prandial lactate
Tyrosinaemia type 1	Ascites Bruising	Elevated LFTs Coagulopathy Hypophosphataemia	UO&AA, Urine succinylacetone
Galactosaemia – neonatal only	Poor feeding/weight loss Ascites Bruising Jaundice (galactosemia) Cataracts	Elevated LFTs Coagulopathy Hyperbilirubinaemia	UO&AA, urine-reducing substances

ACP, acylcarnitine profile; ASA, argininosuccinic aciduria; BSL, blood sugar level; CK, creatine kinase; GSD, glycogen storage disease; IVA, isovaleric acidemia; LCHAD, long-chain 3-hydroxacyl-CoA dehydrogenase; LFTs, liver function test; MCAD, medium-chain acyl-CoA dehydrogenase; MMA, methylmalonic acidemia; MSUD, maple syrup urine disease; OTC, ornithine transcarbamylase deficiency; plasma AA, plasma amino acids; PPA, propionic acidemia; UO&AA, urine organic and amino acids; VLCDA, very long-chain acyl-CoA dehydrogenase.

Those in ( ) – sometimes seen.

* Biochemical panel of venous acid–base, lactate, glucose, ammonia, electrolytes, liver function, ward ketones.

IEM are broken down into a number of groups of conditions, all of which affect a common metabolic pathway. The groups of IEM that are likely to present to the ED are outlined in Table 10.1.1, including common presenting features, biochemical features, and suggested further investigations.

Table 10.1.2 can be used as a guide to identifying an IEM in the ED.

An IEM should be considered if there is both a clinical and a biochemical feature from each of the lists; however, considering an IEM should never take the place of the workup and treatment for more common causes of the above presentations, the most important being sepsis.

Investigation

The metabolic markers of many IEM *are present only at the time of presentation* and may disappear with treatment. Thus, the timing of investigations is

extremely important; if done incorrectly they could result in an incorrect or no diagnosis, with risky provocation testing, such as fasting or loading studies being the only option.

The following investigations are a good starting point when considering an IEM. They are all standard laboratory investigations with rapid turn-around times and are thus likely to be available to the emergency physician (Table 10.1.3).

Blood	Venous blood gas Urea, creatinine, electrolytes, liver function tests Laboratory glucose Ammonia Lactate Ward ketones (beta-hydroxybutyrate) Calculated anion gap ([Na + K] – [C1 + HCO_3])

In *all* cases of hypoglycaemia the following additional investigations are recommended and need to be collected *before* the hypoglycaemia is treated. Most tertiary paediatric emergency departments will have a hypoglycaemia investigation kit for use in this situation.

Blood	Insulin Cortisol Growth hormone, adrenocorticotropic hormone (ACTH) Free fatty acids (FFA) Ketones (beta-hydroxybutyrate and acetoacetate) Acylcarnitine profile (collect a newborn screening card)
Urine	Organic and amino acids

Once the results of the initial investigations are available, a possible diagnosis and further investigations may be suggested, depending on the profile (Table 10.1.1 and Fig. 10.1.2). However, if this is not helpful, all of the following second-line investigations should be performed:

Table 10.1.2

Guide to identifying an inherited error of metabolism in the emergency department

Clinical features	Biochemical features
Overwhelming illness with no clear focus Recurrent vomiting Coma or encephalopathy Apnoea Seizures	Acute acidosis (with raised anion gap) Hypoglycaemia Lactic acidosis (normal perfusion) Ketoacidosis (non-diabetic) Acute hepatic dysfunction

Muscle pain – rhabdomyolysis Not responding to usual treatment FH of neonatal or infant death, SIDS or acute life-threatening event Presence of an unusual odour	Raised CK

CK, creatine kinase; FH, fetal haemoglobin; SIDS, sudden infant death syndrome.

Table 10.1.3

Biochemical profiles of the different inherited errors of metabolism

Defect/disorder	Metabolic acidosis	Lactate	Ketones	NH_3	AST/AST	BSL	Further investigations
Mitochondrial respiratory chain disorder	+ to +++	↑ to ↑↑↑	N to ↑↑	(↑)	N to ↑↑	N or ↓	UO&AA, pre- and post-prandial lactate
Organic aciduria	+ to +++	N or ↑	↑↑	↑ to ↑↑	N or ↑	N or ↓	Plasma AA, UO&AA, ACP
Ketolytic defect	++ to +++	N	↑↑↑	N	N	↓ or N	Plasma ketones, UO&AA, ACP
GSD I	+ to ++	↑ to ↑↑	N	N	N to ↑↑	↓↓↓	Urate, triglycerides, cholesterol
GSD III	+	N	↑ to ↑↑	N	N to ↑↑	↓↓↓	CK, triglycerides, cholesterol
Fatty acid oxidation	– to ++	N to ↑↑	N or ↑	(↑)	(↑↑)	↓↓↓	UO&AA, ACP, CK
Urea cycle defect	(+) OR alkalosis	N	N	↑↑↑	N or ↑	N	Plasma AA
MSUD	– to +	N	↑↑	N	N	N	Plasma AA
Galactosaemia, tyrosinaemia type 1	(+)	N or ↑	N or ↑	N	↑↑↑	N or ↓	UO&AA, urine reducing substances

–, absent; +, present; ( ), sometimes; N, normal; ↑, elevated; ↓, decreased; ACP, acylcarnitine profile; CK, creatine kinase; BSL, blood sugar level; GSD, glycogen storage disease; MSUD, maple syrup urine disease; plasma AA, plasma amino acids; UO&AA, urine organic and amino acids.

Blood	Plasma amino acids Plasma ketones Acylcarnitine profile Creatine kinase Urate
Urine	Organic and amino acids

Many of these investigations can take days for a result, and it is suggested that all suspected metabolic cases be discussed with a metabolic physician, so a treatment plan can be developed to keep the child stable while awaiting further results.

The single most useful investigation in the suspected IEM workup is the urine organic and amino acids, which in some centres is referred to as the ‘metabolic urine screen’. This is preferably the first urine sample passed after presentation, however, as many of the metabolites being measured will continue to be excreted for some time, any sample within the first 24 hours will be helpful. Testing can be performed even on a small non-sterile urine sample; however, for technical reasons most laboratories will not perform the test if there is faecal contamination.

Management

The management of a child suspected of having an acute presentation of an IEM has three primary goals:

1. Correction of altered homeostasis
2. Reduction of toxic compound production
3. Removal or enhancement of excretion of toxic compounds.

Correction of altered homeostasis

Hypoglycaemia and metabolic acidosis are the two most common biochemical abnormalities seen in IEM.

Hypoglycaemia should always be corrected immediately after collection of critical blood samples with an initial infusion of 5 mL kg^{-1} of 10% dextrose. This should be followed with an infusion of fluids containing 10% dextrose such as ½ N. Saline + 10% dextrose or N. Saline + 10% dextrose, depending on the clinical situation and the patient's electrolytes. If run at maintenance rates this will equate to basal glucose requirements of 5–6 mg kg^{-1} min^{-1} of glucose; however, in some conditions where the aim is to suppress catabolism, higher rates of up to 10 mg kg^{-1} min^{-1} of glucose may be required. If infusion rates of dextrose higher than basal requirements (5–6 mg kg^{-1} min^{-1}) are required to maintain normoglycaemia then this suggests hyperinsulinism.

Blood sugar level (BSL) monitoring following hypoglycaemia will depend upon the clinical situation. In the undifferentiated patient, the BSL should be checked within 10 minutes of a corrective dextrose bolus and then at 30 minutes once IV fluids have been commenced in case of hyperinsulinism in which case the patient will require higher rates of glucose infusion. Once the BSL is shown to be stable and the patient is receiving adequate glucose then frequent BSL monitoring is no longer required.

Persistent metabolic or lactic acidosis can be problematic; however, the management strategies outlined later in this section will help reduce acidosis as the patient recovers; occasionally sodium bicarbonate may be required; however, the effect of the associated sodium load needs to be considered and may make other clinical problems such a cerebral oedema worse. Monitoring of venous acid–base depends on the clinical situation. In the undifferentiated patient with metabolic acidosis, a repeat venous acid–base should be repeated at 2 hours.

Reduction of toxic compound production

This is generally achieved through dietary means and varies with the metabolic condition. A feature common to all dietary interventions in the acutely unwell patient with an IEM is the provision of increased calories. Many of the IEM are in catabolic pathways, and thus measures that help suppress catabolism, such as increased caloric intake, help reduce the production of toxic compounds.

The source of calories will vary depending on the condition (carbohydrate vs. fat vs. protein); however, in the acute emergency setting, extra calories from glucose are considered safe and preferable in the majority of IEM. However, protein-containing calories, which include all regular infant formulas, should be avoided in the undiagnosed IEM patient, given that most children presenting acutely with IEM are in pathways of amino acid degradation. Fat sources such as MCT oil and intralipid are dense sources of calories; however, they should only be used when a fatty acid oxidation disorder has been excluded.

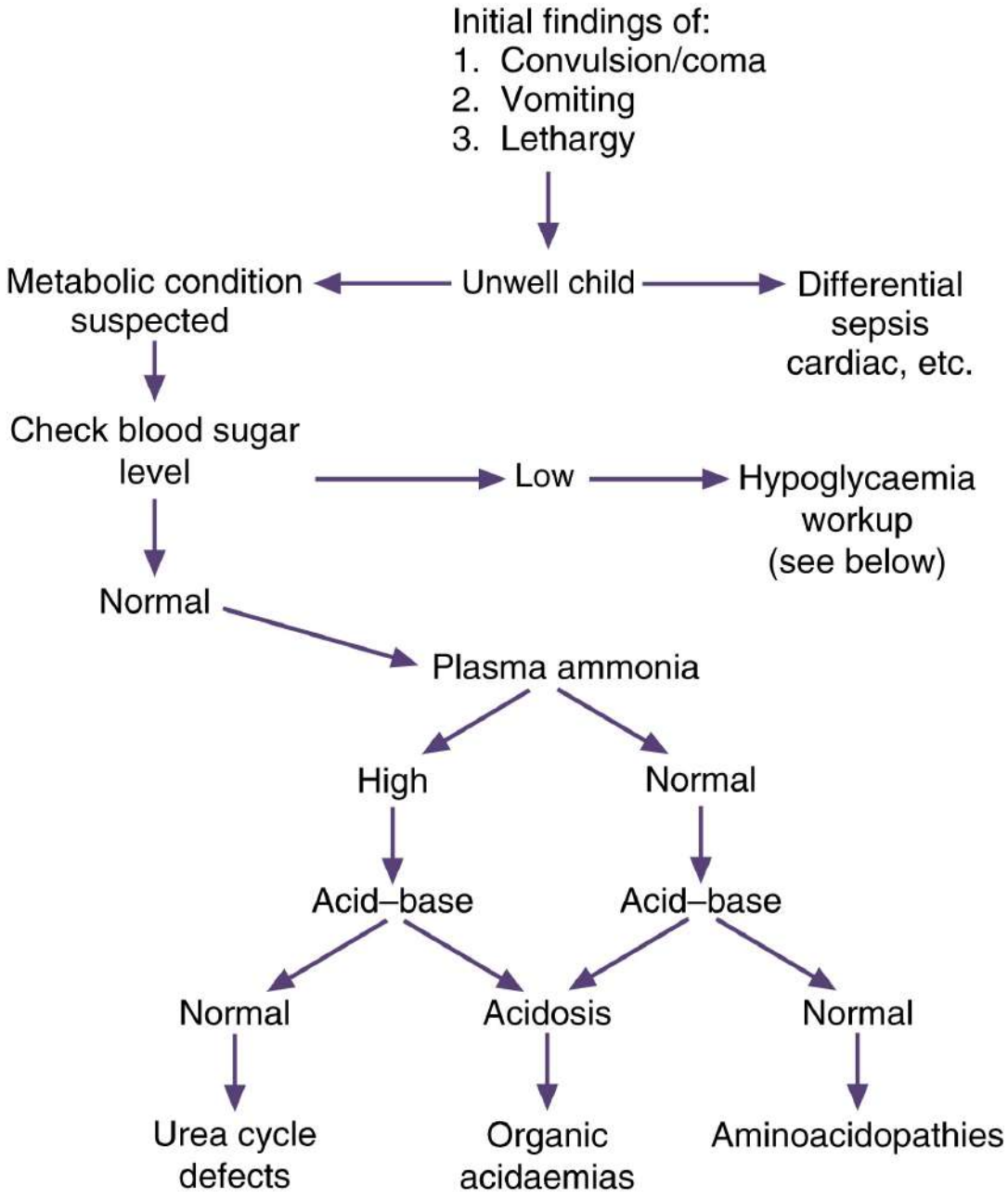

FIG. 10.1.2 Clinical approach for the child with a suspected metabolic disorder.

Table 10.1.4

Chronic presenting signs in inherited errors of metabolism, the associated condition and appropriate investigation

Disorder(s)	Signs	Test

Mucop0lysaccharidoses (MPS) Oligosaccharidoses	Recurrent hernia, especially umbilical Spinal deformity in infant or toddler Recurrent otitis media Persistent nasal discharge Hepatosplenomegaly Coarse facial features	Urine MPS screen Urine oligosaccharides
Fabry disease	Recurrent pain attacks, particularly with fevers Proteinuria	Lysosomal enzymes
Pompe disease (GSD II)	Cardiomegaly Proximal myopathy Short PR interval Raised CK Respiratory failure out of keeping with lung disease	Lysosomal enzymes
Gaucher disease	Recurrent or bilateral avascular necrosis of the femoral head Hepatosplenomegaly	Lysosomal enzymes

CK, creatine kinase.

Thus in the suspected IEM, dextrose at a high rate of between 5 and 10 mg kg^{-1} min^{-1} is the safest and best option while awaiting results and discussion with a metabolic clinician.

Removal/enhancement of excretion of toxic compounds

There are a number of pathways through which toxic metabolites are removed from the body in IEM. Renal excretion, usually conjugated to carnitine, is a common pathway, but in some conditions there is no excretory pathway, and the only treatment option is haemofiltration or dialysis. This is particularly the case in organic acidaemias such as maple syrup urine disease in order to remove the neurotoxic amino acid, leucine, and in the urea cycle disorders to help remove ammonia.

In the acute presentation of IEM, prompt institution of appropriate management is essential to achieve a good outcome for the patient; previously diagnosed patients should have a care plan for acute presentations and in some centres pre-made supplements and formulas available for use. Thus, patients with a known IEM should always be seen promptly (ATS 2) and management as per their care plan instituted as soon as possible; do not wait for topical anesthetics agents to work before putting in an IV, and immediately defrost and put up any pre-made supplements. Those with a suspected IEM should be discussed with a metabolic physician as soon as the suspicion is first raised.

Chronic presentations

There are a number of IEMs that do not present with acute symptoms but in a progressive degenerative manner, frequently multisystemic. The early symptoms and signs in these conditions can be common presentations or incidental findings in the ED (Table 10.1.4). Appreciation of their significance can allow for early diagnosis and in a growing number of conditions can improve the outcome for the patient, through innovative new therapies such as enzyme replacement therapy (ERT), haematopoietic stem cell transplantation (HSCT) and gene therapy.

Extended newborn screening

Many newborn screening programmes throughout the world now use a technique known as tandem mass spectrometry or MS/MS to screen for a large number of IEM from a blood spot. The conditions screened vary between programmes but generally cover most of the organic acidaemias, aminoacidopathies and fatty acid oxidation defects. The conditions a local programme screens for affect the types of IEM that present both diagnosed and undiagnosed to the ED, remembering that these are population-based screening tests and will inevitably miss the occasional case.

Conclusion

The child with a metabolic condition may present in a variety of ways to an ED, and the diagnosis should be considered in all children with an unexplained serious illness. There should be a low threshold for performing blood glucose in the young and initial investigations, and management should be instituted expeditiously.

10.2

Hypoglycaemia in the non-diabetic child

Drago Bratkovic, and Niki Talić

Introduction

Hypoglycaemia is an unfortunately common occurrence in the extremely sick child or neonate. Young children and infants are less able to tolerate prolonged fasting due to decreased glycogen storage and immature metabolic pathways. Blood sugar levels should therefore be checked in any unwell child and hypoglycaemia corrected as soon as it is detected.

The complicating issue is that hypoglycaemia can be the manifesting sign in a number of metabolic and endocrine conditions; prompt and appropriate investigation for these conditions is essential so that a diagnosis is made and management instituted to avoid further episodes of hypoglycaemia and potential morbidity.

Many of these investigations need to be performed *prior* to the treatment of the hypoglycaemia, given that some change almost immediately once treatment has been instituted and if performed later may miss an underlying cause.

The precise definition of what constitutes hypoglycaemia is controversial and therefore varies at different ages and between different clinical centres. A clinical approach is to define hypoglycaemia as a plasma glucose concentration low enough to cause symptoms and/or signs of impaired brain function which improve with administration of glucose. This is practical only in patients who are able to communicate their symptoms.[1]

For younger children current recommendations are for evaluation and management of children who have a documented *laboratory quality* assay showing a plasma glucose concentration of less than 3.3 mmol L^{-1}.[1]

Bedside glucometers are less accurate (±0.6–0.8 mmol L^{-1}) in the presence of hypoglycaemia,[1] and other references suggest that a bedside blood glucose of

less than 2.6 mmol L^{-1} should trigger a workup and treatment of hypoglycaemia.[2,3]

The approach to hypoglycaemia in the child with diabetes is addressed in Chapter 10.3.

The hypoglycaemia screen

We strongly recommend that all emergency departments that see paediatric patients have on hand a premade **hypoglycaemia investigation kit**, including all the required tubes and a pre-written request form. Collecting sufficient blood at the time of venous access being established can be difficult.

Causes of hypoglycaemia

Endocrine and metabolic causes

The most common cause of hypoglycaemia in the non-diabetic child is idiopathic ketotic hypoglycaemia (IKH). This diagnosis can only be made once the episode of hypoglycaemia has been fully investigated and other causes excluded.

Non-diabetic endocrine causes of hypoglycaemia include hyperinsulinism, adrenal insufficiency, growth hormone deficiency and combined endocrine deficiencies such as hypopituitarism.

Hyperinsulinism tends to present in the newborn period and is the working diagnosis in any child that requires greater than 8 mg kg min of dextrose to maintain a normal BSL. The other important clue to hyperinsulinism is the absence of ketones, due to suppression of fatty acid oxidation. Hyperinsulinism poses a significant risk to the brain given the lack of ketones as an alternative fuel source and needs to be identified in order to avoid further events. Rarely hyperinsulinism can be seen in the older child and may be associated with protein ingestion and hyperammonaemia.

Adrenal insufficiency may present with other signs consistent with an adrenal crisis such as hypotension, hyponatremia and hyperkalemia; also look for hyperpigmentation. An important inherited error of metabolism (IEM) associated with adrenal insufficiency is X-linked adrenoleukodystrophy, which is diagnosed on very long-chain fatty acids (VLCFAs).

Metabolic causes of hypoglycaemia can be broken down into those involved with glycogen breakdown (glycogen storage disorders), gluconeogenic disorders

(endogenous glucose production) and fatty acid oxidation disorders. Each group has their own characteristic biochemical and clinical signature (Fig. 10.2.1). Knowing the duration of fasting preceding the event is also important in helping determine the cause.

Glycogen storage disorders (GSDs) are characterised by hepatomegaly and a short duration of fasting prior to hypoglycaemia. The more severe GSDs (I and III) have fasting tolerances of 4–6 hours, whilst the milder hepatic GSDs (IV, VI and IX) can fast for longer (10–12 hours). Hyperketosis is seen in all the hepatic GSDs except for GSD I, where lactic acidemia is seen instead.

Gluconeogenic disorders are rare amongst the IEM, and they are characterised by similar fasting durations to the GSDs but are differentiated by the lack of hepatomegaly and the presence of lactic acidosis. Strictly speaking GSD I is a gluconeogenic disorder; however the other important gluconeogenic disorder is fructose 1:6 bisphophatase deficiency which can be triggered or worsened by fructose ingestion.

Fatty acid oxidation disorders are characterised by longer fasting tolerance, given that fat is the last energy source to be utilised in the fasted patient. The hallmark biochemical presentation is hypoketotic hypoglycaemia; however, milder forms of these conditions can still produce ketones, and it is the relative hypoketosis for the clinical presentation that is the clue. The commonest is medium-chain acyl-CoA dehydrogenase deficiency (MCAD), which does not have any other clinical clues beyond hypoglycaemia and encephalopathy. Longer chain defects such as VLCAD and long-chain 3-hydroxyacyl-coenzyme A dehydrogenase (LCHAD) deficiency can also involve the heart (cardiomyopathy) and skeletal muscle with rhabdomyolysis.

Pharmacological and toxic causes

Potential toxic ingestion should be considered as a differential diagnosis in all patients with unexplained hypoglycaemia, particularly in those presenting with access to medication and a lapse in supervision.

There are many commonly used pharmacological agents that can cause significant hypoglycaemia in children. These could be as a result of an accidental overdose, such as insulin in children with type 1 diabetes or recreational drugs of abuse such as ethanol.

Hypoglycaemia is the most common acute complication of type 1 diabetes due to excess insulin or concurrent intercurrent illness. Both insulin and

sulfonylureas have a narrow therapeutic index. Small doses can cause hypoglycaemia which may be delayed or persistent. Insulin overdose causing hypoglycaemia is most commonly seen in type 1 diabetes but should also be considered in any child with access to insulin at home. Sulfonylurea ingestion can cause prolonged hypoglycaemia, which may be difficult to treat with glucose alone.[4]

Hypoglycaemia associated with alcohol ingestion may be seen in young teenagers and adolescents. The suggested mechanism of alcohol-induced hypoglycaemia in a fasting/intoxicated patient is that of gluconeogenesis inhibition and depleted glycogen stores. It is important to remember that symptoms of hypoglycaemia can be mimicked by mild alcohol intoxication, and early blood sugar monitoring is an important investigation in these patients. Very young children are especially sensitive to alcohol-induced hypoglycaemia, and many require a dextrose infusion following initial treatment.

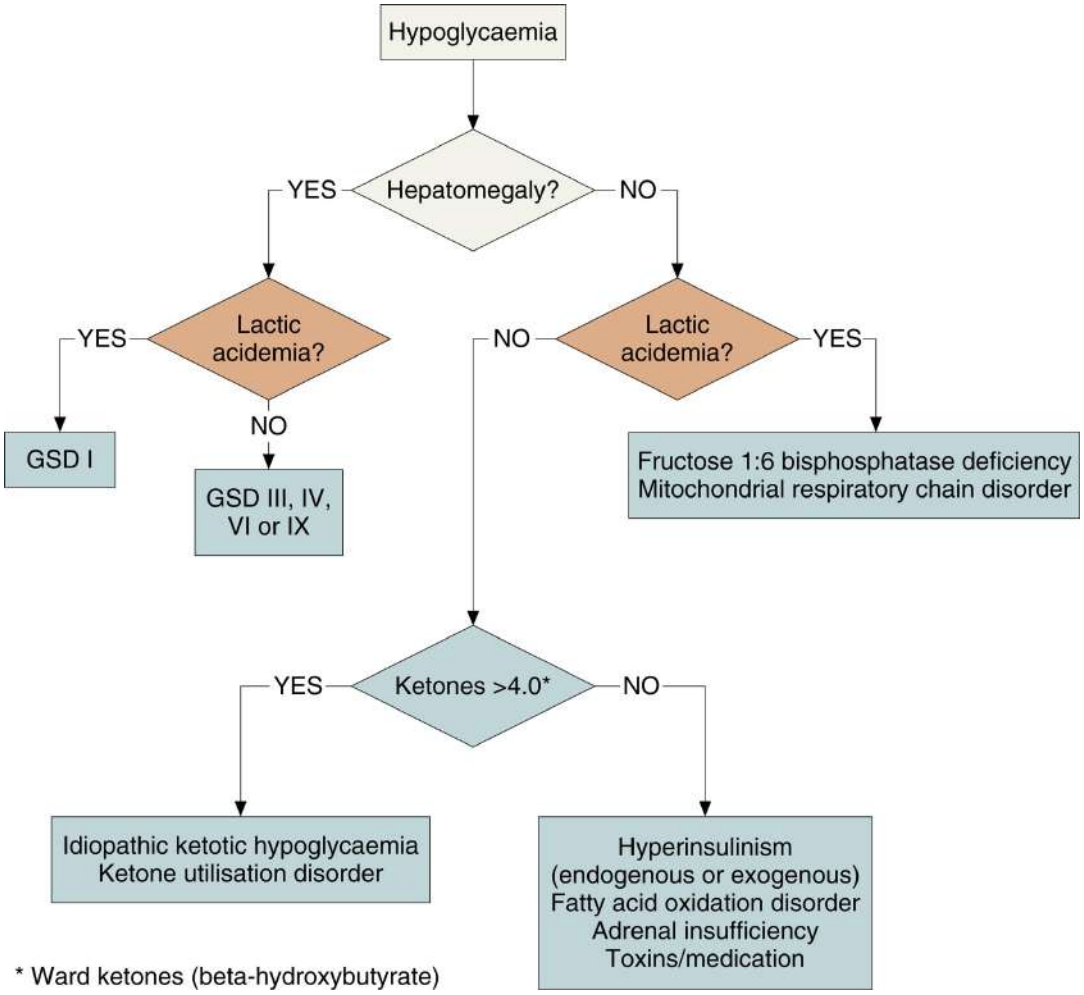

FIG. 10.2.1 Metabolic causes of paediatric hypoglycaemia.

Beta-blockers such as propranolol can cause hypoglycaemia when ingested accidentally. This occurs by inhibition of hepatic glucose production, which is usually promoted by sympathetic nervous stimulation. Adrenergic counter-regulation is diminished resulting in a reduction in glycogenolysis. Cardioselective beta-blockers such as atenolol and metoprolol are less likely to cause hypoglycaemia than non-cardioselective agents such as propranolol.

Other toxicologic causes of hypoglycaemia include salicylates, valproic acid, quinine and chloroquine.[4]

Treatment of hypoglycaemia

Immediate management of hypoglycaemia (*once critical blood samples have been collected*) in the unconscious child is a **bolus of intravenous (IV) 10% dextrose at a dose of 2 mL kg**.

If IV access cannot be obtained then buccal glucose gel (or honey) can be used initially. Glucagon can be used; however, in the non-diabetic child and especially one with a history of prolonged fasting it is unlikely to increase the BSL due to glycogen depletion.

In the awake non-vomiting child, a reasonable approach is the provision of 10–20 g of oral glucose. Juice and most sweetened non-diet carbonated drinks contain 10% carbohydrate, thus 100 mL will equate to 10 g of carbohydrate.

BSL and clinical response should be checked within 10–15 minutes, and if hypoglycaemia persists, or the patient has not recovered, then a dextrose infusion of 10% dextrose at maintenance rates should be commenced. This will provide approximately 5 mg kg min of dextrose.

An IV infusion containing dextrose following initial treatment should be strongly considered in children who are unlikely to eat or drink or where hyperinsulinism is suspected.

In the child with suspected adrenal insufficiency, hydrocortisone must be administered along with correction of the hypoglycaemia with dextrose. Further details on management of adrenal insufficiency are available in Chapter 10.5.

In the symptomatic patient with severe persistent hypoglycaemia after sulfonylurea or quinine ingestion, octreotide is recommended.[5] Experience in children is limited, and advice should be sought from a toxicologist. The recommended starting dose is 1 mcg kg^{-1} subcutaneously or IV followed by an infusion of 1 mcg kg^{-1} $hour^{-1}$. Once stable, tolerating food, and no longer

requiring octreotide, the child should be observed for at least an additional 12 hours or overnight.

Prior to discharge home, children with hypoglycaemia need to be eating and drinking and able to maintain blood sugar levels between meals. This will usually require a period of hospital admission, during which time a working diagnosis will be determined and an appropriate follow-up plan put in place.

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10.3

Diabetic emergencies in children

Kam King Sinn

Diabetes mellitus is one of the most common chronic diseases in childhood, with an increasing incidence. As obesity becomes more prevalent in childhood, type 2 diabetes has started to present in younger and younger adolescents. However, type 1 diabetes remains the major (>90%) cause of childhood diabetes.

Diagnosis

The classic symptoms of polyuria, polydipsia and weight loss may be present for a few weeks before parental concern is raised.

The diagnosis should be confirmed by an elevated random laboratory blood glucose level (>11 mmol L), in addition to urine analysis for glucose and ketones.

Once the diagnosis is confirmed, initial management is dictated by the severity of dehydration, presence of shock, degree of acidosis, hyperglycaemia, ketosis, and osmolality.

In a child with no past history of diabetes, the initial diagnosis may be misled by non-specific symptoms, such as abdominal pain, weight loss, drowsiness, fever, secondary enuresis and dyspnoea. Beware of tachypnoea due to metabolic acidosis, intercurrent infection in a new diabetic, abdominal pain related to diabetic ketoacidosis (DKA), and drowsiness. In such children, diabetes should be excluded as a possible cause with a random blood glucose.

Diabetic ketoacidosis

Diabetic ketoacidosis is the major cause of mortality in diabetic children. It often presents in newly diagnosed type 1 diabetic children. In established diabetics, it occasionally presents in the midst of intercurrent febrile illness, poor adherence

to management, or problems with their insulin pump.

Diabetic ketoacidosis is caused by insulin deficiency, leading to hyperglycaemia, osmotic diuresis, hyperosmolar dehydration, lipolysis, ketosis and acidosis.

It may be defined by the combination of:

hyperglycaemia (BSL >11 mmol L)
ketosis and ketonuria
acidosis (pH <7.3, bicarbonate <15 mmol L)
dehydration.

Management starts with rapid assessment, resuscitation, meticulous replacement of fluid and electrolytes, and an insulin infusion.

Like all medical emergencies, assessment of Airway, Breathing and Circulation (ABC) is vital. Ketotic breath, degree of tachypnoea and respiratory distress should be noted. Degree of shock or dehydration should be assessed. Initial level of consciousness should be noted and hourly neurological observation commenced. It is also important to look for a focus of infection and sepsis.

Initial investigations should include venous blood glucose, bedside blood ketone measurement, electrolytes, urea, creatinine, venous blood gases and full blood count. If sepsis is suspected, blood culture, urine culture and chest X-ray may be required, followed by targeted antibiotic therapy.

Resuscitation

Intravenous access should be established–ideally two intravenous (IV) cannulae to allow for repeated venous sampling of blood glucose, venous blood gases and electrolytes.

In children with shock, noted to be hypotensive and poorly perfused, resuscitation should start immediately with supplemental oxygen and an intravenous fluid bolus of normal saline (0.9% sodium chloride) 10 mL kg. The fluid bolus should be repeated if the child remains shocked upon reassessment in 15 to 30 minutes. In the absence of shock or poor perfusion, a fluid bolus is unnecessary.

Careful and frequent monitoring should continue for the next 24 to 48 hours. Monitoring should include all vital signs, including neurological assessment,

urine output and ECG monitoring.

Fluid

After the initial resuscitation, IV fluid consisting of maintenance fluid and deficit replacement should be calculated and replaced evenly over 48 hours. The child's degree of dehydration should be assessed clinically, including an accurate weight.

Hospital protocols commonly provide recommended infusions rates for intravenous fluids according to estimated degree of dehydration and body weight. These should form the basis of fluid therapy in DKA.

Maintenance and deficit replacement should be given as normal saline (0.9% NaCl) until the blood glucose falls to 12–15 mmol L. Once the blood glucose level (BGL) falls to 12–15 mmol L, the IV fluid should be changed to saline solution with added glucose.

The choice between normal saline with 5% glucose (0.9% sodium chloride and 5% glucose) and half normal saline with 5% glucose (0.45% sodium chloride and 5% glucose) depends on the rate of fall of serum sodium. If serum sodium is dropping rapidly, then normal saline with 5% glucose is preferred.

Beware of giving deficit replacement too rapidly; this decreases intravascular osmolality and may contribute towards cerebral oedema.

Insulin

Insulin infusion should only be started after shock (if present) has been resuscitated. Fluid replacement alone – particularly if given in rapid fashion – will produce a reduction in blood glucose.

Insulin infusion with short-acting insulin (soluble or regular) should be started with maintenance IV fluid. An initial bolus of insulin is not recommended, as it may reduce the BGL and osmolality too rapidly.

The infusion is made up by diluting soluble or regular insulin (e.g. Actrapid^{TM}) in normal saline to a concentration of 1 unit mL, to be given in an IV syringe pump. The infusion is started at a rate of 0.05 to 0.1 units of insulin per kg per hour, titrated to achieve a gradual fall in blood glucose of 4–5 mmol hour.

(For example, a 20 kg child should commence with an insulin infusion of 1–2 units hour.)

Insulin is required to suppress lipolysis and clear ketosis. Even once the blood glucose falls to the normal range (5–10 mmol L), *the insulin infusion should continue until the ketosis is cleared*, with additional dextrose (for example, 10% dextrose) added to intravenous fluids to maintain normoglycaemia.

The change-over from IV insulin to subcutaneous insulin is often most conveniently performed at mealtime–when the child is no longer acidotic, is alert and able to tolerate an oral meal. At such time, rapid- or short-acting insulin can be given subcutaneously before a meal. The insulin infusion should continue during the meal and then cease 30 minutes after the meal.

Local hospital protocols should be followed, and advice may be obtained from local paediatricians/paediatric endocrinologists.

Potassium

The initial serum potassium may be high, normal or low, despite low total body potassium. Potassium replacement should be given intravenously once resuscitation is completed and there are no ECG changes of hyperkalaemia, anuria or severe renal impairment.

The measured serum potassium goes up by 0.6 mmol L for every 0.1 drop in pH. As the acidosis gets corrected with fluid resuscitation and insulin, serum potassium will drop rapidly in exchange for H^+ ions.

The potassium requirement may be as high as 4–5 mmol kg day in the first 24 hours. Initially, potassium chloride 40 mmol is usually added to each litre of normal saline for rehydration. The maximum rate of IV potassium replacement is usually up to 0.5 mmol kg hour.

It is important that serum electrolytes are checked frequently, every 2 to 4 hours in the first 12 to 24 hours of DKA management.

Sodium

Serum sodium is a major determinant of osmolality. Rapid change and fall in serum sodium should be avoided to minimise the risk of cerebral oedema. The measured sodium is initially affected by hyperglycaemia.

The corrected sodium can be calculated by:

$$\text{Corrected } Na^{+} = \text{measured } Na^{+} + (\text{blood glucose}/3)$$

If there is hypernatraemia with a corrected serum sodium >150 mmol L, the rate of rehydration should be slowed down further to 72 hours, to minimise rapid changes in osmolality and fluid shift.

Acidosis and bicarbonate

The severity of DKA at presentation can be determined by the initial acid–base status as follows:

Mild:	pH 7.2 to 7.3	bicarbonate 10 to 15 mmol L
Moderate:	pH 7.1 to 7.2	bicarbonate 5 to 10 mmol L
Severe:	pH <7.1	bicarbonate <5 mmol L

Treatment with bicarbonate is rarely indicated, even in the presence of severe metabolic acidosis. Bicarbonate may increase central nervous system acidosis and worsen hypokalaemia and hypernatraemia.

As the ketoacidosis improves with insulin infusion, the pH gradually improves towards normal over 6 to 24 hours, depending on the initial severity of the DKA.

Blood gas should be repeated at frequent intervals if the initial pH is below 7.1. Bedside point of care measurements of beta-hydroxybutyrate provide accurate and timely assessment of ketosis. Measurement of BSL and ketones should be initially performed hourly to every 2 hours in severe DKA.

Phosphate

Serum phosphate is often low initially. It is controversial whether replacement of phosphate makes any difference to outcome.

Complications

Careful and close observation of vital signs, frequent review of clinical status and laboratory values of BGL, electrolytes, ketone and acid–base status are vital in the first 24–48 hours of DKA to minimise complications.

Hypoglycaemia and hypokalaemia can be minimised by frequent monitoring of BGL and electrolyte, followed by the adjustment of rehydration fluid and

insulin infusion.

Approximately 0.5% children develop **cerebral oedema** as a complication of DKA. The warning signs are changes in level of consciousness, irritability, headache, cranial nerve palsies and seizures. Treatment includes slowing the rehydration infusion, IV mannitol, intubation and assisted ventilation.

Disposition

Many children with DKA are able to be admitted to a standard ward environment once stabilised. Local hospital protocols often have specific cut-offs suggesting which children require transfer to a higher level or care and/or PICU admission.

Common indications for PICU include severe acidosis (pH <7), altered conscious state or suspected cerebral oedema, haemodynamic instability, or very young children who would be challenging to manage with frequent blood tests in a ward environment.

Apart from addressing the clinical and metabolic complications of DKA, hospital admission provides an opportunity for intensive education and support, which may reduce the risk of recurrence.

Hypoglycaemia

Hypoglycaemia is a common complication in all diabetics. Education and ongoing support for the diabetic child and his/her family is the key to preventing and managing hypoglycaemia when it does occur.

Hypoglycaemia can be caused by insulin excess (e.g. ‘honeymoon period’), overdose of oral hypoglycaemic (in type 2 diabetes), intercurrent illness (e.g. gastroenteritis), vigorous exercise and other metabolic/endocrine disorders (e.g. Addison’s disease). Chapter 10.2 provides a discussion of the approach to hypoglycaemia in the non-diabetic child.

Hypoglycaemia is often defined as a BGL <2.5 mmol L. However, symptoms of hypoglycaemia may appear with a BGL <4.0 mmol L in a diabetic. Therefore, the level of BGL in a child with diabetes should be maintained above 4 mmol L.

Hypoglycaemia may cause symptoms related to neuroglycopenia (weakness, fatigue, dizziness, odd behavior, confusion, coma) and autonomic activation (sweating, tachycardia, anxiety and hunger).

In mild to moderate cases where the patient remains conscious, treatment

includes ingestion of rapidly absorbed simple carbohydrates such as sugar or fruit juice, followed by more complex carbohydrates and medical review of the cause of the hypoglycaemia.

In severe hypoglycaemia, the patient may present with seizure or coma. Treatment includes stabilisation of airway, breathing and circulation:

If IV access is available, 2 mL kg of 10% dextrose can be given as an IV bolus.

If IV access is not available, intramuscular or deep subcutaneous injection of glucagon should be given. The dose of glucagon is 0.5 units (0.5 mg) for children up to 8 years of age and 1 unit (1 mg) for older children and adolescents.

In the recovery phase of severe hypoglycaemia, close monitoring of blood glucose, medical review of insulin dosage and diabetic control are required. In addition, further carbohydrates will need to be administered, preferably by ingestion of oral complex carbohydrates but occasionally with intravenous glucose-containing solutions.

In all children with diabetes, education for the parents, patient, teachers and other carers on symptoms and management of hypoglycaemia, sick-day management, and the availability and use of glucagon is vital.

Some form of wearable identification (e.g. Medic-Alert bracelet) for the child would also help in the management of hypoglycaemic coma.

The child with an insulin pump

After initial diagnosis and stabilisation of diabetes, most children are started on management with either multiple daily subcutaneous injections of insulin or continuous subcutaneous insulin infusion (CSII) with an insulin pump. CSII has been used increasingly in recent years, particularly in young children, as a more physiological insulin replacement therapy.

The insulin pumps operate on the basal bolus principle with continuous subcutaneous insulin infusion. The bolus insulin dose is given at meal time, calculated based on food (carbohydrate) consumption and the current blood glucose reading. Many pumps allow programming algorithms to calculate meal and correction boluses based on insulin to carbohydrate ratios, insulin sensitivity and blood sugar level. Rapid-acting insulin analogues are used in most pumps,

with the infusion tubing and cannula changed every few days.

With a small depot of rapid-acting insulin used in CSII, any interruption in the insulin supply can cause a rise in blood glucose and ketones. Short-lived disconnection of the pump gives a blood glucose rise of 1.5 mmol L per 30 minutes. Children on CSII should have check blood glucose regularly at home and also check ketone levels when the blood glucose level is high.

In the event of suspected pump failure, children and their parents/carers should be educated on the use of insulin injections via syringes or insulin pen device.

The most common causes of hyperglycaemia in children on pump therapy include malfunction of the infusion set, intercurrent medical or surgical illness, and pump malfunction.

When a child on insulin pump therapy has hyperglycaemia but no ketosis, a predetermined bolus insulin dose should be given and the blood glucose rechecked in an hour.

If there are hyperglycaemia and ketosis but no acidosis, give 20% of the patient's total daily dose of insulin as a subcutaneous injection of rapidly acting insulin. After this, ask the patient or family to resite the pump cannula and recommence insulin delivery at the usual settings, aiming to normalise blood glucose and reduce ketones to less than 0.6 mmol L.

The blood glucose and ketones should be rechecked in 2 hours. Further management should be discussed with the on-call paediatrician and/or paediatric endocrinologist.

At times of intercurrent illness with stress hormone release, hyperglycaemia may need to be treated with increase in total daily dose of insulin. The basal rate may need to be increased by 10–20%, with the correction bolus increased by 10–20% throughout the duration of the intercurrent illness.

In contrast, at times of poor oral intake (e.g. with gastrointestinal illness), the total daily dose may need to be reduced by 10–20%, based on blood glucose and ketones level.

The child with diabetes and intercurrent illness

There are a few general principles in sick-day diabetes management:

- Do not stop insulin
- More frequent blood glucose and ketone monitoring

- Maintain oral intake and hydration
- Treat the underlying cause of the intercurrent illness
- Emergency contact with paediatric team for advice.

Some febrile illnesses may cause an increase in blood glucose and ketones, due to high level of stress hormone production, causing insulin resistance. In contrast, some illnesses associated with vomiting, anorexia and diarrhoea may lower blood glucose with risk of hypoglycaemia, due to poor oral intake and poor nutrient absorption. As a result, the need for insulin may increase or decrease during the intercurrent illness, which may last for from hours to days.

Even in a fasting state in a type 1 diabetic, basal insulin is required and may need to be increased during an acute illness.

Frequent blood glucose monitoring should be performed at least every 3–4 hours during acute illness. The aim is to maintain blood glucose between 5 and 10 mmol L.

If there is hyperglycaemia with a negative or small amount of ketones, an additional 5–10% of the total daily dose of insulin may be given as rapid-acting insulin subcutaneously, usually as 0.05 to 0.1 unit per kg. This supplemental bolus dose of insulin may be repeated based on blood glucose level in 2 hours.

If there is hyperglycaemia with moderate to high ketones, an additional 10–20% of the total daily dose of insulin may be given as rapid-acting insulin subcutaneously, usually as 0.1 unit per kg. The blood glucose and ketones should be checked again in 1–2 hours to assess response.

In children with a poor appetite, normal meals may need to be replaced with easily digestible food and a sugar-containing solution. Oral rehydration solution used for gastroenteritis can be used to maintain hydration and electrolyte balance at times of nausea and vomiting.

Attention to oral intake, outputs and body weight changes during this time is vital. Frequent blood glucose and ketone monitoring provides a guide to the need for insulin dose adjustment.

If the blood glucose falls below 5 mmol L, with moderate to high ketones, the ketosis is likely due to poor oral intake with ‘starvation’ ketone production. An extra simple carbohydrate is required to maintain blood glucose and suppress ketone production. In patients managed on basal bolus insulin regime, the basal insulin dose may need to be reduced by 10–20% and the bolus doses to be adjusted, based on oral intake.

On sick days when the oral intake can be erratic, the bolus rapid-acting insulin

may be given with or after food when needed, especially when it is unclear how much carbohydrate the child can tolerate.

Diabetic child and surgery

The aim in a diabetic child requiring surgery is to maintain blood glucose in the range of 5 to 10 mmol L and to prevent ketoacidosis.

For emergency surgery, the surgical team and the child's paediatrician and/or endocrine specialist should have close consultation and plan before the operation. If ketoacidosis is present, it should be treated and stabilised before surgery.

If there is no ketoacidosis, the child should be fasted and commenced on maintenance intravenous fluid with 0.9% normal saline and 5% dextrose with potassium chloride (KCL) and a continuous IV insulin infusion. The initial insulin infusion rate may be started at 0.02 to 0.03 units per kg per hour, to be titrated with blood glucose level hourly, immediately preoperatively and intraoperatively.

Transition back to subcutaneous insulin in the post-operative period should be managed individually, based on the patient's usual insulin treatment regime and ability to tolerate an oral diet or gastric feeds.

For elective surgery, the surgery should be planned for the morning list. For a child on a basal bolus regime, the usual basal long-acting insulin can be given on the day before surgery. On the day of the surgery, blood glucose should be monitored closely before, during and after surgery. Based on the blood glucose level, additional short- or rapid-acting insulin can be given while the child is treated with maintenance glucose-containing IV fluid (such as 0.9% sodium chloride and 5% dextrose).

Long-term management

The key to good long-term management of children with diabetes is an enthusiastic team providing a holistic approach to education for the affected child and his/her parents.

Most diabetic children are treated with multiple daily subcutaneous injections of insulin or subcutaneous insulin given via insulin pump. Common subcutaneous injection regimens include either twice-daily injections of a mixture of short- and intermediate-acting insulins, or multiple daily injections of

insulin, using a long-acting insulin at night and pre-meal injections of rapidly acting insulin.

Regular medical review of insulin dosage, diet, exercise, medium-term control and complications should be undertaken with a paediatrician, diabetic educator and dietician. It is beyond the scope of this chapter to deal with the details of long-term management of childhood diabetes.

Further reading

www.ispad.org.

10.4

Thyroid emergencies

Malcolm Higgins

ESSENTIALS

1 Life-threatening paediatric thyroid emergencies are rare.

2 Children with undiagnosed thyrotoxicosis or hypothyroidism may present to the emergency department with a range of acute symptoms and signs.

3 The identification, appropriate management and referral of children with congenital hypothyroidism are important as this condition may cause severe neurological impairment if untreated.

4 The onset of clinical hypothyroidism in Hashimoto's thyroiditis may occur in adolescents.

Thyrotoxicosis

Hyperthyroidism is generally a disease of adult women but occasionally may present in adolescents. Although there are a number of causes, the most common is Graves' disease, which is thought to have an autoimmune basis. Thyroid-stimulating hormone (TSH)-receptor stimulating immunoglobulins stimulate the thyroid and are also thought to be responsible for the ophthalmopathy and dermopathy associated with this condition. In approximately 1–2% of cases the presentation of hyperthyroidism is acute and severe. This has been called the 'thyroid storm' and is potentially life threatening. Other causes of hyperthyroidism in children include thyroiditis, iodine-induced hyperthyroidism, TSH hypersecretion, excessive ingestion of thyroid hormone and thyroid

neoplasms. Hyperthyroidism is one of several endocrinopathies associated with McCune–Albright syndrome.

Clinical features

Raised levels of circulating thyroid hormone have predictable clinical effects depending on the organ system and are similar to the symptoms and signs of catecholamine excess (Box 10.4.1). The onset of symptoms in Graves' disease is usually insidious and variable in severity between patients. Children rarely develop severe ophthalmopathy or dermopathy, but neuropsychiatric symptoms and effects on growth and development are more commonly seen. Most patients with Graves' disease will have a goitre characterised by diffuse, non-tender and symmetrical enlargement of the thyroid gland.

The clinical presentation of thyroid storm is of abrupt onset of high fever and marked tachycardia and hypertension with exaggerated features of hyperthyroidism (see Box 10.4.1). Altered mental state is invariably present and may progress to seizures and coma.

Diagnosis

In most cases, low TSH with raised free T_4 and T_3 will be diagnostic of hyperthyroidism. Other investigations such as thyroid autoantibodies and radionuclear scans are not usually part of the emergency department (ED) assessment.

Thyroid storm is a clinical diagnosis, and treatment should not be withheld while waiting for laboratory results.

Treatment

Further investigation and management of hyperthyroidism in children and adolescents will optimally occur following referral to a paediatric endocrinologist or general paediatrician. Therapy will depend on the cause but may include antithyroid drugs such as carbimazole or propylthiouracil. Both can cause agranulocytosis, so patients discharged on these medications should be counseled regarding important symptoms (such as fever or sore throat), which indicate the need to seek urgent medical advice. Radioactive iodine and surgical treatment are less commonly used in childhood. β-Blockers (usually propranolol)

are useful for initial control of symptoms, particularly in thyroid storm.

Box 10.4.1 Symptoms and signs of thyrotoxicosis

Symptoms

Nervousness/irritability
Heat intolerance and increased sweating
Weight loss
Behaviour problems and poor school performance
Fatigue
Restless sleep/insomnia
Palpitations
Diarrhoea
Menstrual irregularities

Signs

Tachycardia, hypertension
Thyroid enlargement with bruit or thrill
Tremor
Warm, moist skin
Muscle weakness
Eyelid lag and retraction
Brisk tendon reflexes
Growth acceleration

Thyroid storm is a medical emergency. Attention to airway, breathing and circulation is the initial priority. Therapy should be individualised in conjunction with a paediatric endocrinologist and will include antithyroid medications, β-blockers and glucocorticoids.

Neonatal thyrotoxicosis

This rare condition is usually due to the transplacental transfer of thyroid-stimulating antibodies from a mother with autoimmune hyperthyroidism.

Importantly, antibody levels sufficient to cause neonatal thyrotoxicosis may not cause clinical hyperthyroidism in the mother.

The infant may develop clinical features of hyperthyroidism. Poor weight gain, cardiac dysfunction and hepatosplenomegaly may also be present. Cardiac failure and airway compression from the goitre may occur. Onset of symptoms may be delayed if the mother is on antithyroid medication. Although symptoms usually resolve spontaneously, this condition is associated with significant morbidity and mortality.

Hypothyroidism

Insufficient thyroid hormone leads to slowing of bodily functions and can impair the function of many organ systems. The most important causes in the paediatric patient are congenital hypothyroidism and autoimmune (Hashimoto's) thyroiditis.

Congenital hypothyroidism

Inadequate thyroid hormone production in newborn infants can occur from anatomical defects of the thyroid, an inborn error of thyroid metabolism, or from maternal iodine deficiency. Thyroid hormone is vitally important to brain growth and development. Profound mental retardation is the most serious effect of untreated congenital hypothyroidism.

Fortunately, the majority of infants with congenital hypothyroidism are diagnosed soon after birth via the newborn screening programme. A small number of cases will be missed and present for medical attention because of symptoms of hypothyroidism that develop over the first few weeks of life. Suggestive symptoms and signs are listed in Box 10.4.2.

Box 10.4.2 Symptoms and signs of congenital hypothyroidism

Poor feeding, growth and development
Constipation
Umbilical hernia, enlarged fontanelle
Enlarged, protruding tongue
Prolonged neonatal jaundice

Hypotonia
Hoarse cry
Bradycardia, cool extremities

Diagnosis of congenital hypothyroidism is confirmed by demonstrating decreased levels of serum thyroid hormone (total or free T_4) and elevated levels of TSH. Urgent referral for further investigation and initiation of therapy is important.

Box 10.4.3 Symptoms and signs of Hashimoto's thyroiditis

Weakness and lethargy
Declining growth velocity/short stature
Weight gain
Cold intolerance
Constipation
Depression, emotional lability and personality changes
Forgetfulness and poor concentration
Hoarse/husky voice
Goitre
Delayed puberty, menorrhagia and menstrual irregularity
Bradycardia
Hypothermia
Delayed relaxation of deep-tendon reflexes

Hashimoto's thyroiditis

Autoimmune thyroiditis is generally a condition of adult women. Adolescents may be affected with profound effects on growth, pubertal development and school performance. The clinical features are of insidious onset (Box 10.4.3). Diagnosis is confirmed with a TSH assay supplemented by thyroid hormone levels, thyroid autoantibodies and a thyroid scan. Referral to an appropriate specialist is highly recommended.

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10.5

Adrenal emergencies

Yuresh Naidoo

ESSENTIALS

Adrenal Crisis

1 The prompt recognition of the possibility of adrenal crisis or the risk of adrenal insufficiency is paramount to early and appropriate management.

2 Adrenal crisis should be considered as a possible contributor in any child with acute severe cardiovascular collapse.

3 In children, most cases are due to primary adrenal failure, with congenital adrenal hyperplasia the most common cause.

4 A crisis can be precipitated in a child with known adrenal insufficiency who develops an intercurrent illness or other physiological stress.

5 Signs of glucocorticoid deficiency include hypoglycaemia, hypotension (absolute and postural) and refractory shock.

6 Signs of mineralocorticoid deficiency include dehydration (often out of proportion to estimated fluid losses), hyperkalaemia, hyponatraemia, acidosis and pre-renal failure.

7 Patients are at risk of hypoglycaemia.

8 The management of adrenal crisis involves immediate fluid resuscitation, replacement of corticosteroid and treatment of hypoglycaemia.

9 The differential diagnosis of a collapsed neonate (particularly male) in the first 14 days includes adrenal insufficiency.

10 Prevention of adrenal crisis in susceptible children may be possible by following a predetermined action plan during intercurrent illnesses.

Adrenal Excess

1 Cushing's disease (CD) is the commonest form of corticosteroid excess and is a rare clinical diagnosis in paediatric and adolescent patients.

2 Key presenting features of Cushing's syndrome in children include weight gain, a change in facial appearance and growth failure.

3 Exogenous glucocorticoids are the most common cause of corticosteroid excess in childhood.

4 Investigation of potential Cushing's syndrome should occur in consultation with a paediatric endocrinologist and usually commences with a 24-hour urine collection.

Adrenal Crisis

Introduction

Adrenal crisis is a life-threatening emergency caused by acute insufficiency of the adrenal hormones cortisol and aldosterone. This can occur in situations of stress where the adrenal gland would normally respond by increasing glucocorticoid secretion. A crisis can be precipitated in a child with known adrenal insufficiency, who develops an inter-current illness or other physiological stress (e.g. burns, surgery, trauma, and sepsis). In this situation, the increased cortisol requirements of the stress or the altered oral intake of normal replacement therapy results in a relative insufficiency and rapid clinical deterioration. Alternatively, the emergency department (ED) visit may represent a new presentation of adrenal insufficiency in a child previously unrecognised to have the subtle, often non-specific symptoms of lack of adrenal hormones or a child who is at risk due to suppression by prolonged steroid therapy. Prompt recognition of the possibility of adrenal crisis or the risk of adrenal insufficiency is paramount. These children are also at risk of hypoglycaemia, and this needs to be anticipated and managed accordingly.

Adrenal insufficiency can be primary, due to a failure of secretion of the adrenal cortex, or secondary to hypothalamic or pituitary dysfunction. In

children, most cases are due to primary adrenal failure, with congenital adrenal hyperplasia the most common cause. The incidence of neonatal congenital adrenal hyperplasia in Australia is estimated at 5.9 cases per 100,000 births.

1. *Primary* causes include:
    - congenital adrenal hyperplasia
    - Addison's disease (autoimmune)
    - adrenal aplasia/hypoplasia
    - adrenal infarction secondary to haemorrhage/sepsis
    - other – trauma, tumour, post-surgical.
2. *Secondary* causes include:
    - central nervous system (CNS) tumour or trauma
    - idiopathic
    - exogenous steroid therapy – including inhaled corticosteroids.

Clinical presentation

Clinical features may be subtle; however, the diagnosis should be considered in all children with cardiovascular collapse. Features to look for in history, examination and investigation findings are listed below, with differential diagnosis.

History

- Prior steroid use, including inhaled steroids for asthma or other condition
- Known congenital adrenal hyperplasia or other adrenal insufficiency on replacement therapy
- Severe physiological stress (sepsis, trauma, burns, surgery)
- On anticoagulants, haemorrhagic diathesis
- Neonate with collapse and/or hypoglycaemic event or ambiguous genitalia
- Symptoms of glucocorticoid deficiency – weakness, fatigue, lethargy, anorexia, vomiting, diarrhoea, weight loss.

Examination

- Signs of glucocorticoid deficiency – hypoglycaemia, hypotension (absolute and postural), refractory shock
- Signs of mineralocorticoid deficiency – dehydration (often out of proportion to estimated fluid losses), hyperkalaemia, hyponatraemia, acidosis, pre-renal failure
- Signs of excess adrenocorticotropic hormone secretion – pigmentation of skin, lips, nipples, skin creases
- Signs of glucocorticoid therapy – Cushing's syndrome
- Signs of associated hypothalamic/pituitary abnormality – growth abnormality, midline defects, hypogonadism, diabetes insipidus, hypothermia.

Investigations

- Bedside: finger-prick glucose (hypoglycaemia), ECG (hyperkalaemia)
- Biochemical: glucose, urea and electrolytes, blood gas, save clotted blood for cortisol level and 17-hydroxyprogesterone, if no underlying diagnosis is known.

Differential diagnosis

- Other causes of hyponatraemia – syndrome of inappropriate antidiuretic hormone secretion, nephrogenic or cerebral salt wasting, gastrointestinal or urinary losses
- Other causes of shock – septicaemia, profound dehydration, duct-dependent cardiac lesion.

Treatment

The management of adrenal crisis involves immediate fluid resuscitation, corticosteroid replacement and treating hypoglycaemia. The underlying illness causing the stress needs to be treated on its merits. Potential complications, such as hyperkalaemia, may require intervention. In children not previously known to be adrenal deficient, blood (prior to administration of glucocorticoid) should be held for analysis to determine if an underlying condition exists.

Fluid management

Patients known to have adrenal deficiency who are not dehydrated or shocked can have a trial of enteral fluids in the ED. They should be considered to have vomited or potentially not absorbed normal replacement adrenal medication and should be administered intramuscular (IM) hydrocortisone 2 mg kg^{-1} to ensure delivery.

Children who are dehydrated or shocked require intravenous resuscitation with crystalloid. Initial boluses of 10–20 mL kg^{-1} normal saline should be titrated to restore peripheral circulation. Remaining estimated deficit plus maintenance volumes should then be replaced with a 5% dextrose in normal saline mixture over a 24-hour period. The maintenance fluid requirements are 1.5 times normal in this setting, and clinical and biochemical (electrolytes and glucose) reassessment is required to tailor fluid therapy to the individual.

Replacement of corticosteroid

Hydrocortisone should be given intravenously, unless access is significantly delayed, where it should be administered IM as an interim alternative. The appropriate dose can be determined by age:

- Neonate: 25 mg stat., then 10–25 mg 6-hourly
- 1 month–1 year: 25 mg stat., then 25 mg 6-hourly
- 1–3 years: 50 mg stat., then 50 mg 6-hourly
- 4–10 years: 75 mg stat., then 75 mg 6-hourly
- >10 years: 100 mg stat., then 100 mg 6-hourly.

Maintenance doses of glucocorticoid and mineralocorticoid are introduced after the child has been stabilised as an inpatient. Generally, glucocorticoid replacement dose is 10–15 mg m^{-2} day^{-1} orally, and mineralocorticoid dose is 0.1–0.2 mg day^{-1} for salt-wasting children.

Hypoglycaemia

Hypoglycaemia is treated in the routine fashion using a lower concentration of intravenous dextrose in smaller children. Maintenance fluid will generally require 5–10% dextrose solution added to normal saline:

- Neonate/infant: 5 mL kg^{-1} of 10% dextrose stat.
- Older child: 2 mL kg^{-1} of 25% dextrose or 5 mL kg^{-1} of 10% dextrose stat.
- Maintenance fluid: 5–10% dextrose-containing solution with normal saline (0.9% NaCl).

Hyperkalaemia

Hyperkalaemia should be treated if K^+ >7 mmol L^{-1} with ECG changes:

- 0.5 mL kg^{-1} of 10% calcium gluconate over 3–5 minutes
- 0.1 unit kg^{-1} $hour^{-1}$ of insulin + 5 mL kg^{-1} $hour^{-1}$ of 10% dextrose infusion.

Disposition

All children with an established adrenal crisis require inpatient admission.

Discharge may be considered in milder cases, after a period of 6 hours observation, for those patients who respond well after intramuscular hydrocortisone and can tolerate oral fluids.

Prevention

An action plan for parents may prevent an adrenal crisis in at-risk children with an intercurrent illness:

- If moderately unwell and/or temperature is 38–39°C give 3× oral dose of oral hydrocortisone
- If more unwell and/or temperature >39°C give 4× dose of oral hydrocortisone
- If vomiting give IM hydrocortisone 2 mg kg^{-1}
- If gastroenteritis or diarrhoea give 4× dose of oral hydrocortisone.

NB. Only the dose of hydrocortisone should be increased, not fludrocortisone. When the stress is over the previous dose should be resumed without tapering.

Cushing’s Syndrome

Introduction

Endogenous Cushing's syndrome (CS) is a rare life-threatening disorder caused by prolonged exposure to excess glucocorticoid hormone concentrations. CS can be divided into adrenocorticotrophic hormone (ACTH)-dependent and ACTH-independent aetiological categories. Approximately 10% of new CS cases each year occur in children aged up to 18 years, and Cushing's disease (CD) caused by an ACTH-secreting pituitary adenoma is responsible for 75–80% of cases.

Once CS is suspected, the paediatric patient requires investigation using a formal protocol to ensure an accurate diagnosis and definition of the aetiology. Once CD has been diagnosed, the primary aim of treatment is rapid normalisation of serum cortisol, which is particularly important in children due to the adverse effects of prolonged hypercortisolaemia on growth and development.

Presenting symptoms

Early recognition of the salient features of CD is crucial to allow prompt diagnosis and effective treatment. Therefore, five key features, namely a change in facial appearance, weight gain, growth failure (or deceleration) associated with weight gain, elevation of body mass index (BMI) and the presence of genital virilisation, should alert the clinician to the possibility of CD and initiate laboratory evaluation.

Reduction in height velocity and increased weight have a high sensitivity and specificity for CD.

Although there is a spectrum of features, most children and adolescents have a typical Cushingoid appearance.

A further important aspect of the physical assessment in paediatric patients with CD is examination of secondary sexual development. Most children show signs of abnormal virilisation with advanced pubic hair and genital growth in boys in association with prepubertal testicular volumes or pubic hair growth in girls with prepubertal breast development. These features indicate abnormal exposure to adrenal androgens combined with gonadotrophin deficiency.

Striae and acne are present in 49% and 44% of patients, respectively, and are more common in adolescent patients. Young children with CD may present with obesity and growth failure alone, without other classical features such as plethora, hirsutism, acne and striae. Additional features commonly reported

include emotional lability (60%), fatigue (60%) and hypertension (49%). Muscle weakness and easy bruising are rare.

Diagnostic testing

Before embarking on biochemical evaluation to confirm a diagnosis of CD, it is important to exclude other causes of CS such as excess glucocorticoid use (oral, nasal, inhaled, nasal spray and topical treatments), as exogenous CS is much more common than the endogenous form.

The algorithm for investigations in children should be based on that performed in adults and consists initially of confirmation or exclusion of the diagnosis of CS followed by investigations to determine the aetiology. The diagnosis of an ACTH-secreting pituitary adenoma follows from the investigation of suspected CS and the demonstration of ACTH-dependent hypercortisolaemia.

If the diagnosis of CS is suspected on clinical grounds, the most useful next step in the ED is to refer the patient to the local paediatric team.

Specific investigations are usually arranged in consultation with a paediatric endocrinologist. The most sensitive test to check for the possibility of this disease is to measure the amount of urinary cortisol excreted during a 24-hour period. Cortisol is normally secreted in different amounts during the day and night, so this test usually will be repeated once or twice to eliminate the variability that is normally seen. This normal variability is why simply checking the amount of cortisol in the blood is not a very reliable test. A 24-hour free urinary cortisol level greater than 100 mcg is highly suggestive of Cushing's syndrome.

The second test that helps confirm this diagnosis is the low-dose dexamethasone suppression test, which measures cortisol secretion following the administration of a powerful synthetic steroid that will shut down steroid production in everyone with a normal adrenal gland. Subsequent tests will distinguish whether the disease is due to an ACTH dependent or independent cause.

Invariably, once the diagnosis is made, patients will undergo imaging of the adrenal glands to look for tumours in one or both of the glands. If the laboratory tests suggest a pituitary origin, imaging of the brain (and possibly of the chest) will be performed.

Treatment

CD is extremely rare in the paediatric age range, and for this reason, collaboration with a specialised adult endocrinology unit is essential. Definitive treatments such as surgery and/or pituitary radiotherapy, rather than long-term medical therapies, are currently recommended.

Obviously, the treatment of this disease depends on the cause. Pituitary tumours are usually removed surgically and often treated with radiation therapy. Neurosurgeons and some ENT surgeons specialise in these tumours. If the cause is determined to be within a single adrenal gland, this is treated by surgical removal. If the tumour has characteristics of cancer on any imaging tests, then a larger, conventional operation is in order. If a single adrenal gland possesses a small, well-defined tumour, it can usually be removed by laparoscopic adrenalectomy.

Controversies and future directions

1 In Australia there is currently no screening programme in place for congenital adrenal hyperplasia (CAH). Research indicates that early screening may reduce the morbidity or mortality associated with adrenal crisis. National newborn screening awaits the development of a more cost-efficient test.

2 New research suggests that critically ill children with adrenal crisis may be best managed with a single intravenous bolus dose of hydrocortisone followed by a constant rate infusion. This is currently used in preference to the 6-hourly boluses.

3 A further recent research development is that CAH can be diagnosed and treated prenatally if a mother has previously had a child with CAH.

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10.6

Disorders of fluids, electrolytes and acid–base

Wayne Hazell, and Sarah Davidson

ESSENTIALS

1 Safe use of intravenous fluids requires accurate prescribing and monitoring.

2 Isotonic or near-isotonic fluids such as 0.9% saline, 4% albumin in 0.9% saline, blood products and Hartmann's solution are appropriate for volume resuscitation. Hypotonic solutions including 5% glucose and 0.18%, 0.225% or 0.45% saline are inappropriate.

3 The most common causes of hypovolaemic shock in paediatric patients are sepsis, dehydration and trauma.

4 Timely restoration of the circulating volume reduces mortality in hypovolaemic shock.

5 In situations not requiring resuscitation, fluids should be administered by the enteral route whenever possible.

6 After resuscitation of circulating volume, residual dehydration should be corrected slowly with physiological solutions, such as Hartmann's solution or 0.9% saline. 0.45% saline may be used in the presence of hypernatraemia.

7 The degree of dehydration is easily overestimated.

8 The concept of maintenance fluids applies only after resuscitation of circulating volume and repair of dehydration or water overload. Even then, fluid therapy must be individualised to the patient's particular needs.

9 Hyponatraemia generally reflects water excess and creates a risk for cerebral and pulmonary oedema. Hypotonic fluids are contra-indicated until the plasma sodium is corrected. Cerebral oedema may develop rapidly in hyponatraemic children, especially after administration of hypotonic fluids.

10 Rapid correction of hyper- or hyponatraemia is contraindicated.

11 Hyperkalaemia with electrocardiogram changes requires urgent potassium-lowering treatment.

12 Metabolic acidosis is common in sick children. The anion gap remains useful in determining the cause.

13 Treatment of acidosis is aimed at correcting the underlying cause. Alkali therapy is rarely indicated.

14 Metabolic alkalosis is caused by vomiting, especially pyloric stenosis, or as a compensation for chronic respiratory failure.

Introduction

Disorders of blood volume, body fluids, sodium, potassium and acid–base are common in acutely ill children. Always manage the airway and ventilation first, the 'ABC' principle of resuscitation, even in acute illness where shock and dehydration are dominant. In resuscitation of the circulation, vigorous replacement of blood volume deficit is urgent, but correction of residual dehydration is not. Maintenance fluids are the last consideration.

Physiology

There are many physiological differences between adults and infants, and a few specific features must be taken into account in managing fluid therapy. Infants have greater total body water, up to 70% of body weight compared with 60% in adults, the extra fluid being mostly extracellular.[1,2] The ionic composition of intracellular and extracellular fluid is shown in Table 10.6.1. Small children drink more to accommodate a higher metabolic rate and excrete a higher solute load, and thus urine volume is greater.

The following physiological differences apply to children:[1–5]

- Infants are more dependent on heart rate for cardiac output.
- Infants have less ability to increase myocardial contractility.
- The infant's ventricle is less compliant. Higher atrial pressures are required for the same degree of ventricular filling.
- The child is more dependent on the extracellular concentration of calcium for myocardial contractility.
- Storage and release of calcium from the sarcoplasmic reticulum in the infant's myocardium are less efficient.
- The infant tolerates more tachycardia and hypotension.
- Degenerative diseases are uncommon in children.
- The capacity of the infant's brain and heart for anaerobic metabolism is greater than the adult.
- The infant's kidney is less able to concentrate urine and to handle a sodium load in the first 2 years of life. Thus infants are less able to compensate for fluid loss.
- Renal blood flow and glomerular filtration rate are less per unit body surface area.
- Infants have less capability to acidify the urine and are thus less able to deal with a hyperchloraemic acidosis caused by too much chloride administration.
- Total body water and extracellular fluid are proportionally higher in the first 6 months of life. In later childhood extracellular fluid declines, and intracellular fluid increases.
- Limited glycogen stores exist. Hypoglycaemia frequently occurs in periods of stress in infants and young children.

Clinical assessment

In assessing the degree of dehydration, sequential body weight measurement is the most accurate measure of water loss. However, previous normal body weight is seldom available in the emergency department (ED).

A recent systematic review of 21 studies in 1915 children regarding capillary refill testing was undertaken by Fleming et al. The authors recommended the use of the following standardised capillary return testing method of measurement: press on the finger for 5 seconds using moderate pressure at an ambient temperature of 20–25°C. A capillary refill time of 3 seconds or more should be considered abnormal.[6] However, ambient temperature affects capillary return.[7] A

normal capillary refill time of ≤2 seconds is associated with superior vena cava oxygen saturations of greater or equal to 70%,[8] suggesting normal oxygen circulation and delivery.

Table 10.6.1

Ionic distribution of body fluid compartments

Cations and anions	Intracellular fluid (mEq L^{-1})	Interstitial fluid (mEq L^{-1})	Plasma (mEq L^{-1})
Sodium (Na^+)	10	145	140
Potassium (K^+)	150	4	4
Magnesium (Mg^{++})	30	2	2
Calcium (Ca^{2+})	0	5	5
Chloride (Cl^-)	5	114	104
Bicarbonate (HCO_3^-)	10	24	24
Protein (Pr^-)	65	6	15
Phosphate (PO_4^-)	95 (organic)	4 (inorganic)	4 (inorganic)
Sulfate (SO_4^-)	20	1	1

Table 10.6.2

Clinical assessment of the severity of dehydration

Signs	Mild	Moderate	Severe
General condition	Thirsty, restless, agitated	Thirsty, restless, irritable	Withdrawn, somnolent or comatose; rapid deep breathing
Pulse	Normal	Rapid, weak	Rapid, weak
Anterior fontanelle	Normal	Sunken	Very sunken
Eyes	Normal	Sunken	Very sunken
Tears	Present	Absent	Absent
Mucous membranes	Slightly dry	Dry	Dry
Skin turgor	Normal	Decreased	Decreased with tenting
Urine	Normal	Reduced, concentrated	None for several hours
Weight loss	4–5%	6–9%	Greater than 10%
Extremities	Warm, normal capillary refill	Delayed capillary refill	Cool, mottled

Adapted from Burkhart DM. Management of acute gastroenteritis in children. *Am Fam Phys* 1999;**60**:2555–63 and Australian Government Department of Health Emergency Triage Education Kit.[12]

In children less than 4 years old clinicians overestimate the degree of dehydration by 3.2%.[9] Other studies have suggested that the sensitivity of clinical examination for diagnosing dehydration is variable.[10] See Table 10.6.2 for clinical signs associated with dehydration.[11,12]

Plasma bicarbonate concentration may be the single most useful laboratory test; a level less than 17 mmol L^{-1} indicates moderate or severe dehydration. Addition of this to a clinical assessment scale in one study improved the sensitivity of diagnosing moderate and severe dehydration to 90% and 100%, respectively. Plasma bicarbonate was a better predictor of dehydration than plasma urea and creatinine.[10]

Shock is a disorder characterised by a decrease in end-organ oxygenation and/or perfusion. This does not solely depend on blood pressure and pulse rate.

Blood pressure can be maintained in the infant with shock being present. Hypotension is a pre-terminal sign.

End-organ perfusion can best be assessed by the conscious state, capillary return, urine output and degree of metabolic acidosis.

Haemorrhagic shock

Clinical signs are of value in assessing degree of haemorrhage[13] (Table 10.6.3). Hypotension is a pre-terminal sign.

Fluid deficit

In the case of dehydration, once the percentage loss of body weight (PLBW) is estimated from clinical and laboratory tests the fluid deficit can be estimated:

Table 10.6.3

Classes of haemorrhagic shock

Class of haemorrhage	Blood volume lost (%)	Signs
I	<15	Minimal, slight tachycardia
II	15–30	Tachycardia, tachypnoea, diminished pulse pressure, systolic BP unchanged, prolonged capillary refill, minimal decrease in urine output, anxiety
III	30–40	Tachycardia, tachypnoea, decreased BP, decreased urine output, mental status changes
IV	>40	Hypotension, anuria, loss of consciousness

Morgan WM, O'Neill JA. Hemorrhagic and obstructive shock in pediatric patients. *New Horiz* 1998;**6**:150–4.

Table 10.6.4

Most common crystalloid intravenous fluids

	Isotonic		Hypotonic	
Content per litre	**0.9% NaCl +- 5% glucose**	**Hartmann's solution (similar to Ringer's lactate)**	**0.45% NaCl +- 5% glucose**	**5% glucose**
Content per litre	**0.9% NaCl +- 5% glucose**	**Hartmann's solution (similar to Ringer's lactate)**	**0.45% NaCl +- 5% glucose**	**5% glucose**
Na^+ (mmol L^{-1})	154	131	77	
K^+ (mmol L^{-1})		5.4		
Cl^- (mmol L^{-1})	154	111	77	
Ca^{2+} or Mg^{2+} (mmol L^{-1})		Ca^{2+} 2		
Buffer (mmol L^{-1})		Lactate 29		
Glucose (g L^{-1})	+- 50		+- 50	50

$$\text{Fluid deficit (litres)} = \text{PLBW\%} \times \text{estimated premorbid body weight (kg)}$$

Thus a 10 kg, 1-year-old who is 10% dehydrated has a fluid deficit of:

$$10\% \text{ of } 10 \text{ kg} = 1 \text{ kg} = 1000 \text{ g} = 1 \text{ litre or } 1000 \text{ mL}$$

As this is an estimate, ongoing clinical parameters must be assessed including aiming for a urine output of >1.0 mL kg^{-1} $hour^{-1}$.

Oedema

Oedema suggests that considerable water and salt retention has occurred. Mild sodium and water retention does not cause clinical oedema nor does water retention without sodium retention (the syndrome of inappropriate antidiuretic hormone secretion rarely causes oedema). Oedema is most obvious in dependent and distensible subcutaneous tissue such as the genitalia, eyelids, lower legs and lower back. Firm pressure for at least 1 minute is needed to detect subtle cases. Causes include:

- heart failure:
    - structural cardiac disease
    - myocarditis

    - cardiomyopathy
    - arrhythmias, especially supraventricular; tachycardia
    - pericardial effusion
- liver disease:
    - acute hepatic failure
    - chronic hepatic failure
- renal disease:
    - nephrotic syndrome
- protein-losing enteropathy
- hereditary angioedema
- excessive water and sodium intake
- other causes of hypoalbuminaemia.

Investigations

Most children with gastroenteritis do not warrant an IV line or investigations. However, the following biochemical tests should be performed in children presenting with shock or significant problems of water and electrolyte imbalance that require intravenous treatment or where such disorders are expected, e.g. respiratory disease, renal disease, liver disease and encephalopathy:

- Plasma sodium, potassium, urea, creatinine, osmolality, glucose, bicarbonate, lactate
- Plasma calcium, magnesium, phosphate, liver function tests, albumin
- Arterial and/or venous gases if shocked (venous or intraosseous blood samples give a satisfactory measurement of electrolytes and metabolic acid–base status)
- Urine sodium, potassium, urea, creatinine, osmolality, glucose, and dipstick test for protein, red cells and casts (random spot sample).

A urine sample aspirated from a cotton wool ball placed at the perineum is satisfactory for all measurements except urine calcium.

Plasma osmolality can be calculated as well as measured in order to detect an osmolar gap:

$$\text{Osmolality}\ (\text{mosmol kg}^{-1}) = 1.86 \times (\text{Na} + \text{K}) + \text{glucose} + \text{urea} + 10\ (\text{all in mmol L}^{-1})$$

If measured osmolality is 5 mosmol kg^{-1} or more, then an unmeasured solute is present. The most frequent causes are alcohols, such as ethanol or ethylene glycol. Ingestion of alcohols may cause a high anion gap ketoacidosis and hypoglycaemia.

Accurate timed urine collections are rarely possible in the ED. A useful indication of urine flow rate can be obtained from a spot urine sample, based on the relatively constant creatinine excretion between individuals. For example, a urine creatinine of 2000 μmol L^{-1} represents urine flow of 2–4 mL kg^{-1} $hour^{-1}$, and 8000 μmol L^{-1} represents 0.5–1 mL kg^{-1} $hour^{-1}$.

Fractional sodium excretion (FE_{Na}) is a useful diagnostic tool. It represents the proportion of filtered sodium that is not reabsorbed and is <1% in health. It is given by the formula:

$$FE_{Na} = \frac{\text{Urine Na}}{\text{Urine creatinine}} \times \frac{\text{Plasma creatinine}}{\text{Plasma Na}}$$

A high FE_{Na} suggests any cause of natriuresis, including acute renal failure, a renal salt-wasting disorder or diuretics. Hyperosmolar urine with high urine creatinine and low FE_{Na} suggests dehydration of non-renal cause. Diabetes insipidus causes hypo-osmolar urine (often <100 mosmol kg^{-1}), low urine creatinine (often <1000 μmol L^{-1}) and plasma hyperosmolality. Plasma urea is high in most cases of dehydration because of reduced urea clearance.

Treatment

Replacement of circulating volume

Also called volume resuscitation, this is an urgent priority in any cause of hypovolaemic shock, e.g. haemorrhage, sepsis, burns, anaphylaxis and dehydration. It requires isotonic fluids (Table 10.6.4). Hypotonic fluids are inappropriate. Crystalloids are inexpensive and readily available.

Colloids have a theoretical advantage of increasing the colloid oncotic pressure of plasma, thus helping to maintain fluid in the vascular space.

However, no significant benefit has been found in acute resuscitation. A problem with saline is that it has no bicarbonate and relatively high chloride, so it can lead to hyperchloraemic acidosis. Hartmann's and Ringer's solutions are more physiological, containing buffer and calcium.[14]

In dehydration, the loss of fluid and electrolytes is similar to the composition of extracellular fluid; this is predominantly where the loss comes from. Thus the deficit is best replaced with a solution that approximates extracellular fluid, i.e. 0.9% saline (normal saline).

However, as fluid replacement is not an exact science, the general consensus is that maintenance fluids should be with saline (0.9%) and 5% dextrose, with added electrolytes as required. Rarely other fluids are needed, for example: saline (0.45%) with 5% dextrose (hypernatraemia); hypertonic saline (head injuries); or saline (0.9%) and 10% dextrose (neonates). Clinical parameters and serum biochemistry are sequentially measured, and the fluid is adjusted accordingly.

$$\text{Overall 24-hour fluid requirement} = \text{fluid deficit} + \text{maintenance} + \text{ongoing losses}$$

An initial bolus or boluses of 10–20 mL kg^{-1} of normal saline to treat shock may be part of this initial fluid deficit. Glucose may then be added to the sodium chloride preparations for ongoing fluid replacement.

How much fluid?

Enough fluid should be given in shock to result in the improvement and disappearance of the signs of shock.

The blood volume of an infant is approximately 70–80 mL kg^{-1}. In the setting of blood loss, decompensation starts to occur in class 3 haemorrhagic shock. At this stage it can be presumed that at least 30% or approximately 20 mL kg^{-1} of blood has been lost. Thus a bolus dose of 10–20 mL kg^{-1} is a logical dose of fluid with which to begin resuscitation.

In haemorrhagic shock, boluses of 10 mL kg^{-1} of whole blood or packed cells may be given. If fresh frozen plasma (FFP) is required this can also be given in 10 mL kg^{-1} aliquots.

In dehydration and sepsis, much larger fluid losses and shifts have occurred,

and the total body water and extracellular fluid are likely to be depleted far more than 20 mL kg^{-1}. If crystalloid is given, not all of the fluid stays in the intravascular space. Capillary leakiness and third space losses in sepsis contribute to the ongoing loss of fluid from the vascular space.

Thus in dehydration, repeated boluses may be necessary. Give a bolus, wait 10 minutes and reassess the patient again. Do not wait 10 minutes if it is clear that more than 10–20 mL kg^{-1} will be required.[13–15] In severe sepsis, such as meningococcaemia, 80–100 mL kg^{-1} may be required.[15]

However, there is now a growing body of evidence that shows that overzealous fluid resuscitation in adults and children, leading to a positive net fluid balance, is associated with a number of negative effects. These include worsening renal function, acute respiratory distress syndrome (ARDS), prolonged ICU stay, increased hospital length of stay and increased mortality.[16–18]

The volume and the rapidity of resuscitation may be more important than the type of fluid used. The key to this is regular reassessment and evaluation of fluid requirement.

How to administer fluid in shock

Intravenous access is often difficult in a shocked young infant, but it is essential that this is achieved as soon as possible. If intravenous access has not been achieved within 90 seconds and/or three failed attempts have occurred, intraosseous access should be gained.

Poiseuille’s law states that the flow through a hollow tube is proportional to the radius to the power of four and inversely related to the viscosity of the fluid and length of the tube. The small radius can be overcome by applying a larger pressure differential across the tubing. Fluid can be drawn up in a 20 or 50 mL syringe and injected rapidly. It is harder to push fluid through a small cannula with a 50 mL syringe.

A three-way tap can be connected to a normal giving set attached to the IV fluid of choice. The tap can be turned off to the patient allowing fluid to be drawn into the syringe. The tap can then be turned off to the giving set, and fluid can rapidly be injected into the patient.

Ideally, blood should be given through the largest cannula that can be inserted. As well as the usual complications of blood transfusion, the paediatric patient is at risk of hyperkalaemia from cell lysis through the small cannula.

Infants have a higher surface area to body weight ratio and lose heat more quickly. Overhead radiant warmers or fluid warmers are recommended for large rapid volume replacement when hypothermia is a risk.

Investigation and management of fluids in different conditions

Dehydration

Dehydration is a common presenting feature in children. Causes are outlined in Box 10.6.1.

Enteral rehydration either by mouth or nasogastric tube is the method of choice for rehydration unless shock exists. It is safe, cheap and effective. Oral rehydration can be effective even in the presence of vomiting.[19–22]

Box 10.6.1 Causes of dehydration

Gastrointestinal loss

- Diarrhoea
- Non-obstructive vomiting
- Obstructive vomiting
- Third space loss into bowel wall and peritoneum

Renal loss

- Renal tubular disease
- Acquired tubulopathies
- Chronic renal failure including obstructive uropathy
- Adrenal failure
- Congenital adrenal hyperplasia (salt-losing)
- Diabetes insipidus (central and renal)
- Osmotic diuresis (e.g. diabetic ketoacidosis)
- Diuretics

Insensible loss

- Burns
- Sweating
- Hyperthyroidism
- Hyperventilation with reduced intake
- Ichthyosis, e.g. Netherton syndrome

Reduced intake (rarely causes dehydration)

Burkhart DM. Management of acute gastroenteritis in children. *Am Fam Phys* 1999;**60**:2555–63.

Lemonade, homemade oral rehydration solutions (ORS) and sports drinks are not appropriate fluids for rehydration. These may be high in carbohydrate and osmolality and lead to osmotic diarrhoea. Hyponatraemia may occur due to their low sodium content.[11,23]

Oral rehydration should occur with one of the many recommended and/or commercially available ORS such as Gastrolyte™, HYDRAlyte™, or Pedialyte™. Aim for 10–20 mL kg fluid over 1 hour with frequent small amounts. This is often easily administered by parents in the ED with the use of a cup, an oral syringe, and plenty of encouragement. If possible, continue breast-feeding. Early feeding (as soon as rehydrated) reduces stool output and aids gastrointestinal tract recovery.[23] Some trials suggest frozen oral rehydration solution is better tolerated than the liquid form.[24]

Nasogastric rehydration is a safe and effective way of rehydrating most children with moderate dehydration, even if the child is vomiting. It is preferred over the IV route. Most children stop vomiting after NGT fluids are started. If vomiting continues, consider ondansetron, and slow the NG fluids temporarily. However, this is not applicable to children with dehydration from respiratory illnesses (e.g. bronchiolitis) or with hypernatremia who require a tailored rehydration plan. Rapid nasogastric rehydration can be suitable for the majority of patients with gastroenteritis and moderate dehydration**.** This is done at 25 mL kg hour for 4 hours (Table 10.6.5).[23]

Table 10.6.5

Recommended hourly rate for rapid nasogastric rehydration (not intravenous rehydration)

Weight on admission	mL hour	Total infusion time
7 kg	175	4 hours
8 kg	200	4 hours
9 kg	225	4 hours
10 kg	250	4 hours
12 kg	300	4 hours
14 kg	300	4.5 hours
16 kg	300	5 hours
18 kg	300	6 hours
20 kg	300	6.5 hours

Source: Royal Children's Hospital, Melbourne, Australia, Clinical Practice Guideline on Gastroenteritis, [Internet; cited 5.01.17], Available from: http://www.rch.org.au/clinicalguide/index.cfm.

Hypernatraemia and hypernatraemic dehydration

Hypernatraemia is usually caused by water depletion. It also occurs where salt and water are lost but with relatively greater water than sodium losses. Most causes of dehydration can cause hypernatraemia. Rarely, hyperaldosteronism or salt poisoning causes sodium excess.

The clinical features are usually those of dehydration. Cutaneous signs may include warm, 'doughy' texture and possibly decreased skin-fold tenting in severe dehydration, thereby giving the appearance of a lower level of dehydration.[11] Coma and seizures may occur, especially if plasma sodium is >160 mmol L^{-1} at any time and if there are rapid changes in circulating volume or plasma chemistry. Some neurological deficits may result from the

encephalopathy of severe hypernatraemia.

Fractional sodium excretion is low because of a normal physiological response to dehydration but may be high if the cause is an osmotic diuresis, excessive diuretic use or in salt poisoning. The urine is concentrated (osmolality >600 mosmol kg^{-1}) except in diabetes insipidus.

Shock is treated with isotonic fluids until haemodynamically stable, then 0.45% saline, with potassium as needed (40 mmol L^{-1} if not hyperkalaemic). The estimated dehydration should be corrected over 48 hours with slow correction of plasma sodium at no more than 0.6 mmol L^{-1} $hour^{-1}$ (15 mmol L^{-1} day^{-1}). Electrolytes should be measured hourly initially until the rate of change with treatment becomes clearer.

FE_{Na} is high, often >10%. If there is salt excess without dehydration, then give less than maintenance water with no sodium, aiming for a very slow correction of plasma sodium, <0.6 mmol L^{-1} $hour^{-1}$. A diuretic may be considered and, in extreme cases, dialysis.[25–27]

Diabetes insipidus

Diabetes insipidus should be treated with glucose/water solutions. Hyperglycaemia may result. An intravenous vasopressin infusion may be needed. Consider admission to a ward or ICU and consulting an endocrinologist.

Hyponatraemia and hyponatraemic dehydration

Hyponatraemia may be caused by water excess (e.g. water intoxication, inappropriate antidiuretic hormone (ADH)), water and salt retention where the water excess is greater than the sodium excess (e.g. nephrotic syndrome, heart failure, renal failure or liver failure), dehydration where sodium depletion is greater than the water depletion (e.g. renal loss, gastrointestinal loss, third-space loss, congenital adrenal hyperplasia, acute adrenal failure), or abnormal solute in extracellular fluid (e.g. glucose in uncontrolled diabetes) causing water shift from the intra- to extracellular space.

Hyponatraemia may cause nausea, lethargy, depressed consciousness, raised intracranial pressure and seizures, especially if of rapid onset.

Urine osmolality and plasma urea are high in dehydration but low in water intoxication. FE_{Na} is high in salt-losing states, inappropriate ADH and often in acute renal failure.

Treatment of shock is according to standard guidelines and can be instigated

safely with normal saline. Severely symptomatic patients with plasma sodium less than 120 mmol L^{-1}, such as those with seizures or coma (which is likely to be associated with cerebral oedema), should have the sodium corrected rapidly to 125 mmol L^{-1} (but no higher), using 3% saline (0.5 mmol mL^{-1}).

The following formula may be useful:

$$\text{Sodium required (mmol)} = (125 - \text{plasmasodium (mmol/L)}) \times 0.6 \times \text{weight (kg)}$$

One millilitre of 3% saline contains 0.5 mmol of sodium. Therefore, the *volume of 3% sodium required* to achieve a plasma sodium of 125 mmol L^{-1} is:

$$\text{Volume of 3\% sodium chloride required (mL)} = (125 - \text{plasma sodium (mmol/L)}) \times 1.2 \times \text{weight (kg)}$$

A starting dose of 3 mL kg of 3% saline may be reasonable in severe neurological deficit or seizures while precise calculations are ongoing.

Excessive rapid correction of hyponatraemia can be associated with pontine or extrapontine myelinolysis, but it is not clear whether this relates to the severe hyponatraemia itself or the rapid correction. Convulsions and decreasing level of consciousness may occur at any time in treatment, especially if there are rapid changes in circulating volume or plasma chemistry. Some neurological deficit may result from the encephalopathy of severe hyponatraemia.

After treatment of shock, residual dehydration should be managed by replenishing the extracellular space with 0.9% sodium chloride or one of the buffered isotonic solutions, slowly over 48 hours. Residual hyponatraemia should correct itself slowly.

Water intoxication needs water restriction to half maintenance and sodium supplementation if there is evidence of natriuresis.

Syndrome of inappropriate antidiuretic hormone

secretion

This condition is overdiagnosed. Plasma concentration of antidiuretic hormone, vasopressin, is elevated, despite physiological conditions which should suppress it, namely hypo-osmolality and water overload. Hyponatraemia and hypo-osmolality are accompanied by marked oliguria but with high fractional sodium excretion and very high urine osmolality and sodium (often >200 mmol L^{-1}). The child is sodium depleted but water overloaded, so the management of choice is water restriction, the IV fluid being given as 0.9% saline. Furosemide causes a diuresis in syndrome of inappropriate antidiuretic hormone secretion (SIADH) and may contribute to management.

Pyloric stenosis

Shock is treated with 10–20 mL kg^{-1} boluses of 0.9% saline. Saline (0.9%) with 40 mmol L^{-1} of KCl is the appropriate rehydration solution. Glucose needs to be added. This remedies the loss of chloride and potassium and slowly corrects the alkalosis.

Sepsis and meningococcal disease

There is no compelling argument for crystalloid or colloid.[14] There is current controversy regarding the amount of fluid (see FEAST study[16]) and the timing of inotropes. However, inotropes should be considered after the second 20 mL kg^{-1} bolus. The inotropes used are centre dependent, but noradrenaline (norepinephrine) is generally acceptable; ECMO should be considered early in children who are not responding. For further discussion of the management of severe sepsis and septic shock, see Chapter 2.6.

Haemorrhagic shock

Initial resuscitation can take place with crystalloid or colloid. Whole blood at 10 mL kg^{-1} can be given and repeated as necessary until blood pressure is restored. Massive transfusion protocols (MTP) are now commonly available, and these should be referenced at individual sites. FFP only comes in adult-sized bags, and doses of 10 mL kg^{-1} are appropriate with repeating of coagulation times.[13] The use of blood products in children is further discussed in Chapter 11.1.

Head injury

Vigorous treatment of shock should be undertaken to maintain cerebral perfusion pressure.[28] Interest has been generated in the use of 7.5% saline in head-injured patients.[28–30] More evidence is required, and it is not in common use in EDs.

Burns (Chapter 4.6)

The original Parkland formula of 3–4 mL kg^{-1} per percentage burn is still applicable to burns patients. This is over the first 24 hours and starts at the time of the burn, not admission to ED.

Originally, 50% of this was to be given in the first 8 hours and the rest over the remaining 16 hours. However, recent evidence suggests that restoration of urinary output and vital signs occurs earlier if 50% is given in the first 4 hours. Many burns patients require more fluid than is provided by the Parkland formula. Aim for urine output greater than 1 mL kg^{-1} $hour^{-1}$.[31]

Diabetic ketoacidosis (Chapter 10.3)

Treatment of shock should include boluses of 10–20 mL kg^{-1} normal saline. Rehydration over 48 hours is desirable so as to minimise the risk of cerebral oedema. Cerebral oedema is almost exclusively a condition of the newly diagnosed young diabetic, with 95% of cases occurring under 20 years of age.

Table 10.6.6

Hyperkalaemia treatment example for a 10 kg child

Agent	Dose	Dose for a 10 kg child	Route of administration
Calcium gluconate 10%	0.5 mL kg^{-1}	5 mL	slow IV push
Calcium chloride 10%	0.2 mL kg^{-1}	1 mL	slow push (central line only)
Salbutamol		2.5 mg	nebulised
Dextrose 10%	5 mL kg^{-1}	50 mL	to be given with insulin over 1 hour
Insulin (Actrapid)	0.1 units kg^{-1}	1 unit	to be given with 10% dextrose over 1 hour
Sodium bicarbonate 8.4%	1–3 mL kg^{-1}	10–30 mL	not to be given in the same line as calcium
Calcium resonium	1 g kg^{-1}	10 g	rectal but slow acting

The ongoing fluid of choice is controversial. The Royal Children's Hospital Melbourne recommends normal saline and then adjusts depending on sodium and potassium measures. Potassium replacement should be started when the urinary output is adequate and there is no hyperkalaemia.[32]

Hyperkalaemia

Hyperkalaemia may be suggested by ECG changes and may be associated with renal failure, hypoxia-ischaemia or acidosis. There may not be potassium excess but rather potassium shift from intra- to extracellular fluid.

Mild hyperkalaemia may be asymptomatic, but potassium levels >6 mmol L^{-1}, especially when acute, may cause weakness, peaked T waves and wide PR interval on ECG and then loss of P wave, heart block and asystole.

Investigations should include a blood gas, creatine kinase (CK) and glucose.

Any plasma potassium >6 mmol L^{-1}, or <6 mmol L^{-1} but rising fast, demands urgent treatment. If K^+ >7 mmol L^{-1} or there are ECG changes, consider giving calcium 0.1–0.15 mmol kg^{-1}, equating to 0.5 mL kg^{-1} of 10% solution of calcium gluconate or 0.2 mL kg^{-1} of 10% calcium chloride, which does not change plasma potassium but protects cell membranes. Beware of subcutaneous infiltration of the infusion.

Nebulised salbutamol 2.5–5 mg promotes potassium entry into cells as does a glucose/insulin infusion (0.1 units kg^{-1} Actrapid insulin plus 5 mL kg^{-1} of 10% dextrose, over 30–60 minutes). Sodium bicarbonate may also be given, especially if the child is acidotic, at a dose of 1–3 mmol kg^{-1} equating to 1–3 mL kg^{-1} of 8.4% sodium bicarbonate, over 30–60 minutes.

Resonium, 1 g kg^{-1} every 4–6 hours orally or rectally, may eliminate 1–2 mmol kg^{-1} of potassium but is more slowly acting. Consider arranging early dialysis. Continuous electrocardiographic monitoring is required as well as discussion with nephrology and intensive care services.[33] See Table 10.6.6 for an example of doses required for management of hyperkalaemia in a 10 kg child.

Hypokalaemia

Hypokalaemia usually represents with potassium depletion rather than potassium shifts. Causes of potassium depletion include vomiting, diuretics, secretory diarrhoea, ureterosigmoidostomy, renal tubular acidosis, hyperaldosteronism, anorexia nervosa and diabetic ketoacidosis. Causes of hypokalaemia without depletion include salbutamol use, alkalosis and familial periodic paralysis.

Mild hypokalaemia is often asymptomatic. This may not need to be actively treated as it will self-correct with illness resolution. Hypokalaemia associated with alkalosis usually corrects itself as the pH corrects. However, more severe hypokalaemia can cause tachyarrhythmias, ileus, and weakness and rhabdomyolysis may occur.

If potassium depletion requires treatment, it should be by slow potassium

supplementation. Depending on the situation and severity, oral potassium can be given at maximum 1 mmol kg^{-1} (<5 years) or 0.5 mmol kg^{-1} (>5 years). IV potassium can be given at a maximum rate of 0.4 mmol kg^{-1} hour for 4–6 hours. ECG monitoring and frequent repeat measurement are required. Concentration >40 mmol L^{-1} of infusion fluid should be given into a central venous catheter.[33]

Maintenance fluids

The concept of maintenance fluids refers to healthy children, where the kidneys are able to conserve or excrete water and salt over a wide range, in each case according to intake and non-renal losses. 'Maintenance' is a volume of water intake which maintains urine output in the middle of the normal range with an osmolality about that of extracellular fluid. However, changes in total body water and sodium and other electrolytes are common in many diseases. Insensible skin loss of water may be high because of fever and the higher surface area:weight ratio in infants. Maintenance is only relevant after restoration of circulating volume and total body water and is therefore relevant to ongoing rather than ED care. A maintenance amount should be a starting amount. It may be excessive for any sick child where there may be diminished ability to excrete water. Maintenance fluids should initially be with isotonic fluids, such as **saline (0.9%) and 5% dextrose**, and then tailored to the child's needs. Hypotonic solutions should not be used for initial maintenance fluids.

A common formula for calculating 'maintenance' water requirements in a healthy child is as follows:

- For the first 10 kg of body weight: 100 mL kg^{-1} day^{-1} or 4 mL kg^{-1} $hour^{-1}$
- For the second 10 kg of body weight: 50 mL kg^{-1} day^{-1} or 2 mL kg^{-1} $hour^{-1}$
- For every subsequent kg of body weight: 25 mL kg^{-1} day^{-1} or 1 mL kg^{-1} $hour^{-1}$.

Remember to not include the dehydration deficit in maintenance; consider this separately and use 0.9% saline or a buffered isotonic solution for replacement. Fluid input from coexisting drug administration and oral intake should also be taken into consideration. Constant monitoring and re-assessment are the key to safe maintenance fluid prescribing.

Unwell children are more likely to release ADH as a response to illness; this

in turn reduces the ability of the kidneys to excrete water. Therefore maintenance fluids should be reduced by a one-third initially or even one-half if there is risk of cerebral oedema, e.g. in meningitis or brain injury, especially if hyponatraemia already exists.

Acid–base disorders

Disorders of physiological control of acidity of body fluids are common in acutely ill children. The system of defining acid–base state by changes in pCO_2 (respiratory) and standardised base excess (metabolic) according to the Copenhagen school remains the most familiar way of analysing acid–base disorders. Base excess is mostly bicarbonate deficit but includes a small amount of buffering by albumin and a larger amount by haemoglobin. Standardised base excess is provided by blood gas machines derived by microprocessor rather than the original nomograms. The philosophy of base excess has been challenged because it does not take into account the actual plasma albumin concentration and assumes a notional haemoglobin concentration of 50 g L^{-1} across blood and extracellular fluid in all patients. It can be argued that the bicarbonate concentration alone is a sufficient measure of the degree of metabolic acidosis or alkalosis.

Table 10.6.7

Causes of acidosis

High anion gap	Normal anion gap
M – Metformin U – Uraemia/renal failure D – Diabetic ketoacidosis P – Phenformin, pyroglutamic acidosis I – Infection, inborn errors of metabolism, isoniazid L – Lactic acidosis including sepsis, hepatic failure, hypoxia, shock, leukemia, lymphoma and G6PD deficiency E – Ethanol, ethylene glycol, ethanol ketoacidosis S – Salicylates (especially in babies), solvents starvation/ketosis	U – Ureteroenterostomy S – Small bowel fistula E – Excessive sodium chloride administration, early acute renal failure D – Diarrhoea C – Carbonic anhydrase inhibitors A – Addison's disease R – Renal tubular acidosis P – Pancreatic fistula

The anion gap remains a useful tool in determining whether any bicarbonate deficit is caused by organic acid (high anion gap) or by chloride excess (normal anion gap). It is obtained from the formula:

$$\text{Anion gap} = Na^+ + K^+ - Cl^- - HCO_3^- \ (\text{all in mEq } L^{-1})$$

The normal is 16 mEq L^{-1} and is essentially the negative charge on albumin and phosphate. If the potassium is normal this can be deleted from the equation with the normal now being 12 mEq L^{-1}. Any excess is accounted for by abnormal unmeasured acid and/or lactate. Most modern laboratories, including blood gas machines, measure lactate.

A more accurate way of determining the unmeasured component is from the formula:

$$\text{Unmeasured anion} = Na^+ + K^+ + Mg^{2+} + Ca^{2+} - Cl^- - HCO_3^- - \text{albumin} \times (0.123 \times \text{pH} - 0.631) - \text{phosphate} \times (0.309 \times \text{pH} - 0.469) - \text{lactate}^- \ (\text{all in mEq } L^{-1})$$

The normal value for unmeasured anion is 5–6 mEq L^{-1}, including 1 mEq L^{-1} of lactate. Any excess is abnormal. This may be lactate in hypoxia–ischaemia, acetoacetate or β-hydroxybutyrate in diabetic ketoacidosis (DKA), ketones in starvation, organic acids in inborn errors of metabolism, or alcohol poisoning or toluene inhalation.

Most metabolic acid–base disturbances do not need treatment. Compensating mechanisms should generally not be treated, otherwise the primary disturbance is exacerbated. It is not necessary to restore pH to normal.

Metabolic acidosis

Normal physiology maintains extracellular pH close to 7.4 in health but permits metabolic acidosis at times of anaerobic metabolism without danger. Metabolic acidosis may be advantageous because it is thought to protect cells against the effects of hypoxia and assists oxygen unloading from haemoglobin by shifting the oxygen/haemoglobin dissociation curve to the right.

When a metabolic acidosis is present as indicated by a pH <7.4 associated with a bicarbonate of less than 24 mmol L, an anion gap should be calculated as above. If there is a raised anion gap the mnemonic ‘MUDPILES’ may be useful, and if the anion gap is normal the mnemonic ‘USEDCARP’ may be useful

(Table 10.6.7).

It is often useful to also perform a delta ratio and gap. This can inform a clinician as to whether there is one or more types of metabolic acidosis occurring. Delta ratio equals the increase in anion gap divided by the decrease in bicarbonate. This is done by the following equation:

$$\text{Calculated Anion gap} - 12^{*} / \text{Bicarbonate} - 24$$

∗(if not using K and it is normal, otherwise use 16 instead of 12)

A guide to interpreting the delta ratio value is in Table 10.6.8. Clinical judgement is needed in conjunction with these values as guides[34].

Acidosis is said to cause negative inotropy or failure of inotropes to work at pH <7.2, but this has little experimental support. Acidosis causes pulmonary vasoconstriction and may predispose to arrhythmias caused by other electrolyte abnormalities.

In metabolic acidosis, bicarbonate deficit (mmol kg^{-1}) is given by:

$$\text{Weight (kg)} \times (24 - \text{plasma bicarbonate } [\text{mmol}\,\text{L}^{-1}]) \times 0.5$$

Plasma lactate estimation should be performed if there is significant base deficit or high anion gap. For suspected inborn error of metabolism (Chapter 10.1) if the anion gap is greater than 20, send the urine for metabolic screen.

Table 10.6.8

Guide to interpreting the delta ratio

Delta ratio	Assessment
<0.4	Hyperchloraemic normal anion gap acidosis
0.4–0.8	Consider combined high anion gap (AG) and normal AG acidosis BUT note that the ratio is often <1 in acidosis associated with renal failure

1–2	Usual for uncomplicated high-AG acidosis Lactic acidosis: average value 1.6 Diabetes ketoacidosis more likely to have a ratio closer to one due to urine ketone loss (especially if patient not dehydrated)
>2	Suggests a pre-existing elevated HCO_3 level so consider: a concurrent metabolic alkalosis or a pre-existing compensated respiratory acidosis

Source: Brandis K. Acid–Base Physiology. http://www.anaesthesiamcq.com/AcidBaseBook/ab3_3.php.

First treat the underlying disease (Table 10.6.7), including general management of renal, hepatic failure, cardiac failure, hypoxia, shock and hypovolaemia. Alkali is not usually needed for mild acidosis (pH >7.2) because the acidosis itself does not cause any compromise. Even extreme acidaemia (pH <6.8) can be followed by full recovery, and bicarbonate therapy is only indicated if there is severe hyperkalaemia, tricyclic antidepressant poisoning or some sodium-channel-blocking toxicities.

Sudden changes in acid–base status should be avoided. Sodium bicarbonate has many adverse effects, especially when given rapidly, such as hypokalaemia, decreased plasma ionised calcium, sodium load, osmolar load, increased haemoglobin/oxygen affinity, exacerbation of effects of hypophosphataemia, and late metabolic alkalosis. There is no evidence for increased intracellular acidosis. Sodium bicarbonate is especially unhelpful in lactic acidosis, resulting in sodium overload with a metabolic alkalosis as the lactic acid is metabolised to bicarbonate during recovery.

Slow sodium bicarbonate treatment may have a role in the management of normal anion gap acidosis where excessive chloride therapy would exacerbate hyperchloraemia. A suitable amount is 2 mmol kg^{-1} day^{-1}. In diabetic ketoacidosis bicarbonate is not recommended acutely, even in severe acidaemia with pH <7.0.

Inborn errors of metabolism may present with severe metabolic acidosis and sometimes hypoglycaemia. Hypoglycaemia should be corrected, after taking critical blood samples (Chapter 10.2). Bicarbonate therapy is rarely required. Initial fluid therapy should include 0.9% saline and 10% dextrose at maintenance rates. Further management of metabolic disorders is described in Chapter 10.1.

Metabolic alkalosis

Metabolic alkalosis may be caused by chronic potassium and/or chloride depletion, e.g. vomiting, especially pyloric stenosis (accompanied by volume

depletion) or renal (including diuretic use). It may be compensatory (chronic renal bicarbonate retention) in chronic respiratory failure, in which case it should not be treated. Acute alkalosis causing tetany by lowering plasma ionised calcium should be treated.

After correction of dehydration, chloride deficit is approximately 6 mmol kg^{-1} for every 10 mmol L^{-1} fall in plasma chloride. A suitable fluid for treating alkalaemic patients is 0.9% or 0.45% sodium chloride plus 40 mmol L^{-1} potassium chloride with added glucose. Hypotonic solutions, which may exacerbate hyponatraemia, should not be used.

Intravenous hydrochloric acid (or arginine hydrochloride) is indicated in rare severe cases when alkalosis may be depressing respiratory drive and when the chloride deficit is not accompanied by sodium deficit. This should be only given in an intensive care setting.

Acidifying diuretics such as acetazolamide may be indicated in metabolic alkalosis where there is sodium and water retention.

Controversies and future directions

1 Volume resuscitation and severe sepsis.
2 Clinical signs are the current standard in assessing the response to fluid in paediatric patients, but this process is inaccurate. Further research is needed into more accessible avenues of monitoring, e.g. ultrasound or tissue oxygen saturation.

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SECTION 11

Haematology and oncology

OUTLINE

11.1

The use of blood products in children

Joseph Ting, and Kottayam Radhakrishnan

ESSENTIALS

1 Blood components are appropriately used when clinical benefits of administration outweigh potential hazards, provided the indication is appropriate. Although blood product administration is relatively safe, potential risks remain.

2 Use specific component(s) appropriate to a clinical problem whenever possible. Determine and document urgency for transfusion, the specific blood product to be administered, quantity and duration of transfusion and special instructions (e.g. premedication).

3 For safe use of blood products, hospitals must have a policy for blood product administration, including the staff responsible, credentialing, documentation, checking and administration procedures such as identity checks. The majority of adverse events result from administrative errors.

4 Blood loss with poor perfusion and/or haemodynamic derangement requires packed red blood cell (PRBC) transfusion, with or without platelets and coagulation factors, while emergent decisions are made regarding the need for definitive surgical or interventional radiology haemostasis.

5 Anaemia that does not pose a hypoxic or metabolic risk to the asymptomatic child may not require transfusion; current guidelines use a Hb threshold <70 g L^{-1} in critically ill children, although well children without symptoms and/or where alternative therapy is

available (e.g. erythropoietin) may tolerate lower levels.

Introduction

Successful and safe transfusion practice depends on administering a quality blood component of the right type, in the right amount, in the right way, at the right time to the right patient.[1]

Blood components should only be given when the expected benefits to the child outweigh the potential hazards. A range of clinical signs and symptoms viewed within the context of a clinical need is essential for the decision to transfuse. Transfusion triggers include both clinical (symptoms, signs, comorbidities) and laboratory indications; benefit–risk of blood product use requires careful consideration.

Emphasis on blood component safety, standardisation of appropriate guidelines for use of blood components and informed consent for blood component administration have led to a substantial reduction in the potential risks and complications of their use as well as increasing their appropriate usage. Informed consent with regards to the risks and benefits of blood component therapy needs to be obtained in the light of community concerns about transfusion safety, particularly the potential for infection transmission. The indication, risks, benefits, alternatives to transfusion, parental consent, response to treatment and any adverse event should be clearly documented.

Clinical guidelines for use of blood products in children are standardised and consensus and evidence based in Australia.[1] It is recommended that intravenous (IV) access be 22–24G or larger for children receiving blood products through a standard blood administration set primed with normal saline or the blood component. Very slow rates are recommended in small children if rapid volume expansion is not required. Blood products should not be warmed to above 41°C.

The rate of administration should not be >5 mL min^{-1} in the first 15 minutes as severe reactions are most likely to occur then; all blood components should be infused within 4 hours unless fluid overload is a risk. The child and infusion need to be monitored during blood product administration, more closely if unconscious or anaesthetised. A severe reaction requires suspension of blood product administration pending further incompatibility or bacterial contamination checks, consideration of antihistamines/steroids, and should be reported to the Australian Incident Monitoring System in Australia or the New

Zealand Blood Service.

Use of blood products in resuscitation[1, 2]

In neonatal and paediatric patients with critical bleeding requiring massive transfusion (>40mL kg of blood products within 12 hours), use a critical bleeding protocol, which includes weight adjustments to guide blood product supply and administration.

During critical bleeding, the clinician is required to:

- identify and aggressively control the cause of bleeding including surgery and angiography as needed
- restore or maintain normal coagulation
- avoid hypothermia
- avoid excess crystalloid
- tolerate permissive hypotension until bleeding actively controlled, especially if penetrating trauma.

The principles of damage control trauma resuscitation include early diagnosis of at-risk patients, maintaining a low ratio of PRBC to FFP and platelets within the governance of a massive transfusion protocol, as well as minimising crystalloid use.

Haemoglobin should not be used alone as a transfusion trigger in hypovolaemic shock. Massive transfusion is likely in children with any of the following features: base deficit >6, INR >1.5, sustained hypotension, reduced haematocrit before treatment and hypothermia.[2]

Civilian adult and military studies suggest improved survival in massive transfusion with reconstituted equivalent of whole blood 1 PRBC: 1 FFP:1 platelet unit. However, some would advocate ratios of up to 1:3.

For children suffering, or at risk, from massive bleeding, pack one of a massive transfusion protocol could comprise the following:[2]

- Child <20 kg: 2 units of RBC, 3 units of FFP
- Child 20–40 kg: 3 units of RBC, 4 units of FFP
- Child >40 kg: 4 units of RBC, 6 units of FFP
- Consider giving cryoprecipitate 1 unit kg if ongoing features of coagulopathy despite FFP usage, especially if there is ongoing bleeding

and packed red blood cell (PRBC) requirement in the trauma setting.

There remain many controversies, including fibrinogen replacement, the role of antifibrinolytics and recombinant VIIa, the optimal ratio of FFP/PRBC/platelets and the use of point of care coagulation testing.

The adverse effects of massive transfusion include hypothermia, electrolyte abnormalities (hyperkalaemia, hypocalcaemia), acidosis, citrate toxicity, transfusion-related lung injury and multiorgan dysfunction.

Packed red blood cells[1]

Indications

In children, including those who are critically ill, a restrictive transfusion strategy is suggested.

In haemodynamically stable children (excluding neonates), if Hb <70 g L, RBC transfusion is often appropriate. Transfusion guidelines should use age-specific Hb reference ranges and close monitoring of volume and rate of administration.

In children the decision to give a RBC transfusion should not be dictated by Hb concentration alone. The decision should also be based on assessment of the patient's underlying condition, anaemia-related signs and symptoms, and response to previous transfusions. Underlying conditions that influence the decision to transfuse include congenital and acquired cardiac disease, and severe respiratory disease. Non-resuscitative PRBC transfusion should be discussed with the treating paediatric team.

Recommended initial dose of packed red blood cell transfusion

One formula for determining the appropriate initial volume of PRBC for transfusion in children is:

$$\text{Transfusion volume (mL)} = \text{patient's weight (kg)} \times (\text{desired Hb[g/L]} - \text{patient's Hb[g/L]}) \times \text{transfusion factor (0.5)}$$

Example : 12 – *month* – *oldterminfantwithcriticalnon* – *trauma* – *relatedanaemia*, *Hb*45, *tachypneaandlethargy*

$$TargetHb = 90\ \text{gL}$$

$$CurrentHb = 45gL$$

$$Calculatedweight = (age + 4)\ \times 2 = 10\,kg$$

$$Transfusionvolume(mL) = 10kg \times (90 - 45g\ L)\ \times 0.5 = 225mL$$

Alternatively, administration of 10–20 mL kg of PRBC is a reasonable starting dose. A transfusion of 10 mL kg is often sufficient and will increase Hb by approximately 20 g L.

Clinical reassessment to determine the need for further transfusion is then appropriate. The child's Hb level, patient factors, signs and symptoms of hypoxia, ongoing blood loss, risk of anaemia and risk of transfusion should be considered.

Adverse reactions

Fatal acute haemolytic reactions occur in 1:250,000–1:1000,000 transfusions, with half caused by ABO incompatibility as a result of mismatching the blood with the patient or other administrative errors.

Another 1:260,000 patients have a haemolytic reaction due to minor red-cell

antigen incompatibility and 1:1000 patients experience delayed haemolytic reactions.

Platelets

Indications

General

In children undergoing chemotherapy and haematopoietic stem-cell transplantation, the recommended strategy for prophylactic use of platelets is at a platelet count of $<10 \times 10^9\ L^{-1}$ in the absence of risk factors and at $<20 \times 10^9\ L^{-1}$ in the presence of risk factors. Platelets are appropriate in bleeding children in whom thrombocytopenia is a major contributory factor. In haemorrhagic shock, platelet infusion should be considered in massive transfusion associated with platelet count less than $50 \times 10^9\ L^{-1}$.

Platelet transfusion may be appropriate in children with failure of platelet production and platelet count less than $10 \times 10^9\ L^{-1}$, as there is a risk of intracerebral bleeding. Platelets may be given prophylactically in children undergoing invasive procedures or surgery, to maintain platelet count $>50 \times 10^9\ L^{-1}$.

In children with platelet dysfunction (inherited or acquired), platelets are administered to treat active bleeding. Platelets are indicated in thrombocytopenic premature infants with active bleeding or prior to an invasive procedure.

Immune thrombocytopenia (Chapter 11.4)[5]

Most children with idiopathic thrombocytopenic purpura (ITP) do not require platelet-enhancing therapy including transfusion. The watch and wait strategy is used in most patients with mild disease. Steroids, intravenous immunoglobulin (IVIG) and anti-D Ig are current first-line measures for children at risk of severe bleeding.

Chemotherapy-induced thrombocytopenia[6]

The risk of bleeding in chemotherapy-induced thrombocytopenia (CIT) increases as platelet count decreases. The American Society of Clinical Oncology 2001 guidelines recommend platelet transfusion threshold for CIT to be $<10 \times 10^9\ L^{-1}$. Risk factors for bleeding with CIT include specific age groups (neonates, ages 0–5 and 6–12 years), signs of haemorrhage, high fever, hyperleucocytosis, rapid

platelet fall, associated coagulopathy, acute promyelocytic leukaemia, critical illness, and patients undergoing invasive procedures.

Administration

Each paediatric unit is 40–70 mL and contains at least 5.5×10^{10} platelets.

Infusion of 5–10 mL kg^{-1} of platelets will lead to a rise in platelet count of 50–100 $\times 10^9$ L^{-1}.

Adverse reactions and other problems

Platelets are stored at room temperature to maintain their ability to aggregate, and this promotes bacterial growth. Compared with other blood components, the risk of bacterial contamination of platelets is relatively high at 1:2000 units, and sepsis is the most frequent severe complication of platelet use. Platelets older than 5 days lose their ability to aggregate and are therefore less effective. Alloimmunisation due to the development of platelet alloantibodies from repeated transfusions may lead to refractory thrombocytopenia.

Fresh frozen plasma

Fresh frozen plasma (FFP) contains all coagulation factors, except VII, including approximately 150 units of factor VIII. FFP is used to treat bleeding related to coagulopathy associated with cardiac surgery or massive transfusion for haemorrhagic shock when 50% or more of circulating blood volume has been replaced. FFP may be considered for treatment of acute disseminated intravascular coagulation (DIC) and thrombotic thrombocytopenic purpura (TTP).

Less urgent indications include children with coagulopathy undergoing invasive procedures/surgery, liver disease-related coagulopathy, replacement of single coagulation factor deficiency where a specific factor concentrate is not available, and reversal of life-threatening bleeding due to warfarin.

FFP should not be used for volume expansion, plasma-exchange procedures or treatment of immunodeficiency states. Specific recombinant coagulation factors rather than FFP are used to treat specific inherited coagulation disorders.

Administration

In life-threatening exsanguination requiring massive transfusion, FFP may be required to minimise dilutional coagulopathy. One unit of FFP is required for every four units of packed cells but titrate FFP use against the coagulation profile if possible. FFP contains all coagulation factors including approximately 1 unit of factors VIII and V for each mL of FFP. FFP is administered at an initial dose of 10–20 mL kg^{-1}. This dose raises coagulation factor level by 20% immediately. After thawing, FFP needs to be used immediately or stored at 2–6°C for up to 24 hours.

Cryoprecipitate

Cryoprecipitate is appropriate to use when clinical or potential bleeding (invasive procedure, trauma or DIC) is attributable to fibrinogen deficiency. It contains factors VIII, XIII, fibrinogen, von Willebrand factor and fibronectin. Approximately 200 units of each of these factors are contained in a 15 mL bag. This allows more rapid administration than FFP, whilst reducing the risk of fluid overload. In infants, a dose of 5–10 mL kg^{-1} is sufficient to provide adequate amounts of the appropriate coagulation factors. Cryoprecipitate is not used to treat von Willebrand disease, haemophilia, or deficiencies of factor XIII or fibronectin.

Clotting factor concentrates

Bleeding or thrombosis in children with proven coagulation or antithrombotic factor deficiency may require specific factor replacement. These factors have become safer with the advent of synthesis using recombinant genetic technology. Recombinant factor VIII concentrate is used to treat haemophilia A, whereas recombinant factor IX is the treatment of choice for haemophilia B. Antithrombin is used to treat patients with specific factor deficiencies associated with prothrombotic disease.

Albumin

Indications

Albumin is derived from volunteer human plasma pools and is indicated for rapid volume expansion in children with evidence of shock or poor perfusion. In paediatric emergency practice, albumin may be used in the initial fluid

resuscitation for hypovolaemic shock, although it is no better than crystalloids or other colloids in this setting in adults. Other indications include the treatment of hypoproteinaemia, diuretic-resistant nephrotic syndrome, large volume paracentesis, severe burns after the first 24 hours and plasma exchange. There is no evidence for albumin use as a nutritional supplement or for the treatment of ascites or oedema related to portal hypertension.

Administration

Albumin 4% (40 g L^{-1}) at 10–20 mL kg^{-1} is used for volume expansion.
Albumin 20% (200 g L^{-1}) is used for plasma albumin repletion.

Adverse effects and risks

There is evidence that albumin is associated with increased mortality and morbidity in critically ill adults but no such evidence in young children. Complications of albumin use include circulatory and sodium overload, with the relative risk of viral disease transmission being less compared with cellular blood components.

Normal human immunoglobulin

Human immunoglobulin affords passive immunity against many infectious agents for several weeks. Normal human immunoglobulin (NIGH) is derived by Cohn fractionation from pooled plasma and contains antibodies to viruses that are prevalent in the general population. The ‘new’ purified commercial products are all a little different, with different minor reaction rates.

NIGH is administered intramuscularly (IM) for pre- or post-exposure prophylaxis against measles, varicella zoster and hepatitis A.

IV NIGH (either Intragam or Sandoglobulin) is used to treat ITP, Kawasaki disease, perinatal acquired immune deficiency syndrome, Guillain–Barré syndrome, demyelinating disease, immunological disorders associated with antibody deficiency (such as primary hypogammaglobulinaemia) and sepsis with secondary immunodeficiency.

Allergic reactions and, rarely, anaphylaxis may occur. NIGH is contraindicated in children with selective IgA deficiency, and evidence for its role in the treatment and prevention of neonatal sepsis is conflicting. Immune

response to live virus vaccines (with the exception of yellow fever vaccine) may be inhibited by NIGH; timing of further doses depend on the particular immunoglobulin administered.

Hyperimmune immunoglobulins

Maternal administration of rhesus (Rh) Ig prevents the development of Rh antibodies in an Rh-negative mother who gives birth to an Rh-positive baby. This reduces the risk of maternal Rh sensitisation and maternal-neonatal Rh incompatibility in subsequent pregnancies.

Cytomegalovirus (CMV) Ig is used in the prevention and treatment of CMV in children at risk of adverse sequelae from CMV infection, such as with immunodeficiency and following renal and bone-marrow transplantation.

Zoster Ig (ZIG) is advisable within 72 hours after varicella exposure in children with cellular immune deficiency, immunosuppression, neonates whose mothers are susceptible to primary varicella infection and infants who are less than 28 weeks' gestation at birth.

Infants born to HbsAg-positive mothers should receive hepatitis B Ig (HBIG) and commence primary hepatitis B vaccination within 12 hours of birth.

Respiratory syncytial virus (RSV) Ig is used prophylactically in infants at high risk of severe RSV infection, including those with bronchopulmonary dysplasia.

Tetanus Ig passively protects non-tetanus immune children who sustain a tetanus-prone wound, and a tetanus Ig infusion is used to treat clinical disease.

Risks of blood component use[7]

If a major reaction occurs, blood product administration needs to be discontinued immediately. Reactions are categorised as acute or delayed, haemolytic or non-haemolytic, allergy based or not.

Mild non-haemolytic febrile reactions can occur with any blood component and need to be distinguished from major reactions. If a mild non-haemolytic febrile reaction occurs (fever, chills, headaches and/or rigors), stop administering blood products, administer paracetamol and review the patient. If the child is stable, the transfusion can be recommenced.

If a major reaction occurs, the transfusion is discontinued, and threats to the airway, breathing and circulation are attended to. Specific treatment for anaphylaxis, sepsis and blood group incompatibility may be required, in

consultation with a haematologist. Recipient and donor blood samples are sent for immunological and microbiological testing. Critical adverse incidents should be notified to the blood bank and followed up as a quality assurance activity with appropriate adjustments made to hospital transfusion protocols to reduce risk of recurrence.

Infections

Even though blood components are the safest they have ever been, infection transmission risk remains emotive and highly publicised, especially for human immunodeficiency virus (HIV). Infection acquired from a blood component is a rare occurrence when compared with non-infectious complications.

The estimated risks per actual blood unit transfused are less than 1 in 1 million for HIV, HCV, and HTLV 1 and 2.[3] The risk of transfusion related HBV is 1 in 5570,000.[4] The risk of transfusion-transmitted viral infection is now so low that mathematical modelling is required to estimate the residual risk. The small residual risk for blood product recipients is due to the window period between infection and the first detectable viral marker. Disease transmission occurs primarily during the window period when a blood donor is infectious and the infection is immunologically silent and therefore undetectable on screening tests.

There have been no reported cases of transmission by transfusion of classical Creutzfeldt–Jakob disease (cCJD), and retrospective studies suggest the possibility of such transmission of cCJD is remote. To date there have been no reported cases of cCJD in Australia. In the UK, there have been a small number of reported cases of putative transfusion transmission since 2004. In Australia, as a precaution, people who have spent a cumulative period of 6 months in the UK between 1 January 1980 and 31 December 1996 and/or had a transfusion in the UK between 1 January 1980 and the present time are not accepted as blood donors.[7]

Transfusion-related acute lung injury

Acute respiratory distress syndrome with non-cardiogenic pulmonary oedema occurs within 4 hours in 1:5000 transfusions, with 90% of patients recovering. This reaction may be difficult to distinguish from fluid overload and is related to immune-mediated increased pulmonary capillary permeability.

Transfusion-mediated immunomodulation

Allosensitisation, disturbed immunomodulation (both due to contamination by donor leucocytes) as well as infection transmission may be reduced by leucocytodepletion, psoralens and UV irradiation of blood components and using non-pooled blood components from a single donor. Immunomodulation is related to decreased cell-mediated immunity, with increased risk of reactivated viral infection, solid tumour recurrence and post-operative sepsis.

Transfusion-associated graft-versus-host disease

This occurs as a result of transfused lymphocytes engrafting and proliferating in the transfusion recipient. Transfusion-associated graft-versus-host disease (TA-GVHD) is fatal in 90% of cases because of induction of marrow hypoplasia. Irradiation of cellular blood components is effective prevention.

Other transfusion-related adverse reactions

These include donor-recipient incompatibility, allergic reactions, anaphylaxis or anaphylactoid reactions, circulatory overload, haemolysis (acute/chronic, intra-/extravascular, associated fever/no fever), complications associated with massive transfusion (hypothermia, dilutional coagulopathy and thrombocytopenia) and post-transfusion thrombocytopenia.

Controversies and future directions

1 The threshold for red cell transfusion at which benefits outweigh risk in stable children and non-critically ill adults remains unclear.
2 The role and potential harm of albumin in adult resuscitation have not been investigated in paediatric resuscitation, and the role of immunoglobulins in severe sepsis remains uncertain.
3 Oxygen-carrying red cell substitutes such as modified haemoglobins and perfluorocarbon emulsions may offer the same benefits as red blood cells without the infection–transmission risk but are not in clinical use at present.
4 The role of non-blood alternatives such as autologous blood donation,

perioperative blood salvage and directed parental transfusion is not clearly defined in children.

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11.2

Anaemia

John Craven, and Michael Osborn

ESSENTIALS

1 The normal haemoglobin concentration and type are dependent on age and gender.

2 Clinical effects of anaemia depend on the time over which it develops, the magnitude of the anaemia and the underlying cause.

3 Iron deficiency is the commonest cause of microcytic, hypochromic anaemia and is treated with oral iron replacement in nearly all cases.

4 Haemolytic anaemia can be classified as hereditary or acquired, with acquired causes being subclassified as immune mediated or non-immune mediated.

5 Autoimmune haemolytic anaemia (AIHA) is commonly caused by a variety of infective agents, although it can be triggered by a number of medications and systemic illnesses.

6 Acquired non-immune haemolytic anaemia can be due to infective agents, medications, chemical exposures, or mechanical damage (e.g. haemolytic uraemic syndrome, prosthetic heart valves).

7 Haemolytic uraemic syndrome (HUS) is the commonest cause of acute renal failure in children, most frequently secondary to infection with *E. coli.*

8 Thrombotic thrombocytopenic purpura has clinical features that overlap with HUS, as well as exhibiting neurological abnormalities.

9 G6PD deficiency is an X-linked enzymatic inborn error in which haemolysis is triggered by exposure to an oxidant.

10 Thalassaemia is a relatively common inherited defect in the synthesis of the globin chain.

11 Sickle cell disease is an autosomal recessive inherited condition with a globin chain that is unstable in the deoxygenated state leading to vaso-occlusive complications.

Introduction

Anaemia is defined as a reduction in the red blood cell (RBC) volume or haemoglobin concentration below normal values. The normal haemoglobin level varies according to age and gender, and racial differences exist. In the first week of life, the normal range of haemoglobin is 168–228 g L. This falls to a trough level of 90–114 g L, or lower, in premature infants between 8 and 12 weeks of age before rising over the course of childhood toward the normal adult ranges of 115–165 g L (non-pregnant female) or 135–180 g L (male).

RBC production is regulated by erythropoietin. This hormone is initially produced in the fetal liver, but after birth production shifts to the renal peritubular cells. Erythropoietin stimulates the differentiation of committed progenitor cells in the bone marrow into RBCs. During this process, there is a condensation of the cell's nuclear material, with continual production of haemoglobin until it comprises 90% of the mass of the RBC. The nucleus is then extruded, leaving the RBC with no synthetic or replicative ability. Mature RBCs have a life span of 120 days, although disease may reduce this, before being removed from circulation by the reticuloendothelial system, including the liver and spleen.

Haemoglobin is the functional constituent of the RBC that binds oxygen for transport, with the RBC acting as the carrier through the cardiovascular system. Haemoglobin is a complex protein consisting of iron-containing haem groups and polypeptide chains, known as globin. The haemoglobin molecule is made up of two pairs of polypeptide chains each coupled with a haem group. Chemical variations in the polypeptide chains lead to different types of haemoglobin being produced. At various stages of life, from the embryo to the adult, there are different types of haemoglobin normally present. The normal adult haemoglobin, HbA, is comprised of two α-polypeptide chains and two β-chains ($\alpha_2\beta_2$). Haemoglobin F, or fetal Hb (HbF), is comprised of two α-chains and two γ-chains ($\alpha_2\gamma_2$). This constitutes 70% of the haemoglobin present at birth but

reduces to trace levels by 6 months of age. Minor amounts of HbA_2 ($\alpha_2\delta_2$) are present at all ages. Pathological variations in the polypeptide chains can produce disease states known as haemoglobinopathies (see below).

Anaemia is a clinical finding rather than a disease process in its own right and occurs as a result of a variety of pathological processes. The anaemias of childhood can be pathologically divided into two groups: those caused by inadequate production of RBCs or haemoglobin and those due to increased destruction or loss of RBCs. Boxes 11.2.1 and 11.2.2 list the major causes of childhood and neonatal anaemias. From a practical perspective, anaemia is usually classified based on the size of RBCs, as measured by the mean corpuscular volume (MCV). Anaemia can be microcytic (MCV typically less than 80 fL – although this varies with age), normocytic (80 to 100 fL), or macrocytic (greater than 100 fL). Fig. 11.2.1 outlines an algorithm for evaluating anaemia based on MCV.

Acute management

Anaemia is more likely to be an incidental finding in the emergency department (ED); however, severe and life-threatening anaemia does occur. This needs to be recognised and treated urgently. The principles of management of life-threatening anaemia in the ED are:

1. resuscitate the patient with circulatory collapse
2. remove or treat the precipitant if known
3. treat the cause if treatment can be initiated in the ED.

Causes of life-threatening anaemia include uncontrolled haemorrhage, acute intravascular haemolysis in glucose-6-phosphatase deficiency (G6PD) or autoimmune haemolytic anaemia (AIHA), sequestration crisis in sickle cell disease (SCD), and acute decompensation in chronic anaemia.

Initial assessment is directed at respiration and circulation. The anaemic patient may be tachypnoeic with tissue hypoxia. The adequacy of ventilation should be ascertained and supplemental oxygen provided. Circulatory compromise in acute haemorrhage and acute haemolysis requires cardiorespiratory monitoring and intravenous access for fluid resuscitation (see Chapter 2.5 on shock).

Box 11.2.1 Causes of childhood anaemia

Production defect

1. Primary bone-marrow failure:
    - Aplastic anaemia (congenital or acquired)
    - Diamond–Blackfan anaemia
    - Transient erythroblastopenia of childhood (TEC)
    - Marrow infiltration, e.g. malignancy or osteopetrosis
2. Erythropoietin production problem:
    - Chronic renal disease
    - Hypothyroidism
3. Abnormal cytoplasmic maturation (microcytic anaemias):
    - Iron deficiency
    - Sideroblastic anaemia (congenital or acquired)
    - Lead toxicity
    - Thalassaemia syndromes
4. Megaloblastic anaemia:
    - Vitamin B_{12} deficiency
    - Folate deficiency
    - Thiamine-responsive megaloblastic anaemia
    - Orotic aciduria (rare, associated with megaloblastic anaemia)
5. Others
    - Anaemia of chronic disease
    - Congenital dyserythropoietic anaemias
    - Erythropoietic protoporphyria

Increased RBC turnover

1. Hereditary haemolytic anaemias:
    a. Haemoglobin variants:
        - Thalassaemia syndromes
        - Sickle cell disease
    b. Abnormal red blood cell (RBC) membrane:
        - Hereditary spherocytosis
        - Hereditary elliptocytosis

- Hereditary stomatocytosis

c. Abnormal RBC enzymes:
- Glucose-6-phosphate dehydrogenase (G6PD) deficiency
- Pyruvate kinase deficiency

2. Acquired haemolytic anaemias:

a. Autoimmune haemolytic anaemia (AIHA):
- Warm antibody type (primary or associated with autoimmune disease, immunodeficiency, or lymphoproliferative disease)
- Cold antibody type (paroxysmal cold haemoglobinuria or cold haemagglutinin disease)

b. Alloimmune haemolytic anaemia (e.g. haemolytic disease of the newborn, haemolytic transfusion reactions, drugs)

c. Non-immune haemolytic anaemia:
- Infection (e.g. malaria, *C. perfringens*)
- Chemicals, drugs, and physical agents (e.g. burns)
- Fragmentation haemolysis (e.g. HUS/TTP, prosthetic heart valves, disseminated intravascular coagulation)
- Paroxysmal nocturnal haemoglobinuria

3. Haemorrhage

Box 11.2.2 Causes of anaemia in the neonate

1. Physiological anaemia
2. Abnormal RBC production:

a. Nutritional deficiency

b. Secondary to infection

3. Decreased RBC survival – blood loss:

a. Overt blood loss:
- Iatrogenic during delivery
- Obstetric complications (e.g. ruptured cord, placental laceration)

b. Occult blood loss:
- Twin-to-twin transfusion

- Fetoplacental haemorrhage
- Fetomaternal haemorrhage

4. Decreased RBC survival – haemolysis:
    a. Immune related: e.g. haemolytic disease of the newborn
    b. Infection related
    c. Hereditary haemoglobin variants, enzyme abnormalities, membrane abnormalities

Box 11.2.3 Ethnicity and anaemia

1. Thalassaemia syndromes:
    - α-Thalassaemia – descendants from China, Malaysia, Indonesia and Africa
    - β-Thalassaemia – descendants from Africa, Middle East, Mediterranean, India, Pakistan, China
2. Sickle cell disease:
    - Descendants from Africa, southern Arabia, Turkey, Greece, central India
3. Hereditary spherocytosis:
    - Descendants from northern Europe
4. G6PD deficiency:
    - Descendants from Africa, China, southeast Asia and the Mediterranean

Packed red blood cell transfusions (Chapter 11.1) are required if haemorrhage or haemolysis is life threatening or there is severe anaemia with cardiovascular compromise. A dose of 4 mL kg of packed RBCs raises haemoglobin concentration by approximately 10 g L. This dose can be given over an hour or more slowly in the presence of cardiac failure. Frusemide 1 mg kg may be given if there is evidence of volume overload. If tissue oxygenation is not critically affected, the circulatory volume should be sustained with intravenous fluid until group-specific or cross-matched RBCs are available. Acute transfusion may not be required if the bleeding or haemolysis is controlled, further RBC loss is unlikely, circulation is adequate and the haemoglobin concentration is greater than 70 g L. However, a cross-match should be performed regardless, and the

packed RBCs should be held in reserve for at least 24 hours. Early consultation with a haematologist is important in acute haemolytic anaemia, especially if the underlying cause has not been diagnosed.

Symptoms of anaemia depend on the time over which it develops, the severity and the underlying cause. The presentation is frequently not specific to anaemia. Common presenting features consistent with insidious onset anaemia include:

- reducing exercise tolerance
- lethargy, irritability, sleep or settling difficulty
- pallor or yellow skin hue
- loss of appetite ± weight loss ± pica
- bruising, bleeding or infection (if other cell lines are also involved).

Anaemia is confirmed by low haemoglobin concentration and decreased RBC count. An age-related reference range must be used. Abnormalities in the other cell lines should be noted. The mean cell volume (MCV) may suggest an underlying cause. The reticulocyte count is expressed as a percentage of the circulating RBC mass with a normal range of 0.5–1.5%. A low reticulocyte count is found in abnormalities of RBC production, and a high reticulocyte count is found in increased destruction or from RBC loss. Reticulocytosis will be absent if there is concurrent bone-marrow pathology and may not yet be evident if the anaemia has developed acutely. Morphological features of RBCs may be diagnostic of the underlying cause as shown in Table 11.2.1.

There are many additional investigations that may assist in identification of the underlying cause. Some that are undertaken in the ED include:

- bilirubin, lactate dehydrogenase and haptoglobin to detect haemolysis
- urea and creatinine, liver function and thyroid function to detect underlying medical conditions
- iron studies (ferritin, serum iron, transferrin, transferrin saturation)
- direct antiglobulin test (DAT) to identify immune-related haemolysis
- haemoglobin electrophoresis.

Identification and treatment of the underlying precipitant, where appropriate and possible, are a crucial part of the management of anaemia in the ED. Involvement of a paediatric haematologist is recommended for all symptomatic anaemias; however, simple nutritional anaemias can initially be managed in

liaison with the child’s regular doctor.

Neonatal anaemia

Physiological

In the term neonate, the haemoglobin at birth ranges between 159 and 191 g L and rises in the first 24 hours. Subsequently it falls to a trough level of 90–114 g L (or lower in premature neonates) between 8 and 12 weeks of age. This decline is generally referred to as physiological anaemia of infancy and is an expected normal occurrence. The falling haemoglobin is due to a relative haemodilution in the first 3 months of life from rapid weight gain and blood volume expansion, combined with the shorter life span of the fetal RBC, and a sharp decline in erythropoiesis that occurs a few days after birth due to increased arterial oxygenation and a reduction in the production of erythropoietin. Following the nadir in haemoglobin, erythropoiesis increases and the haemoglobin rises again.

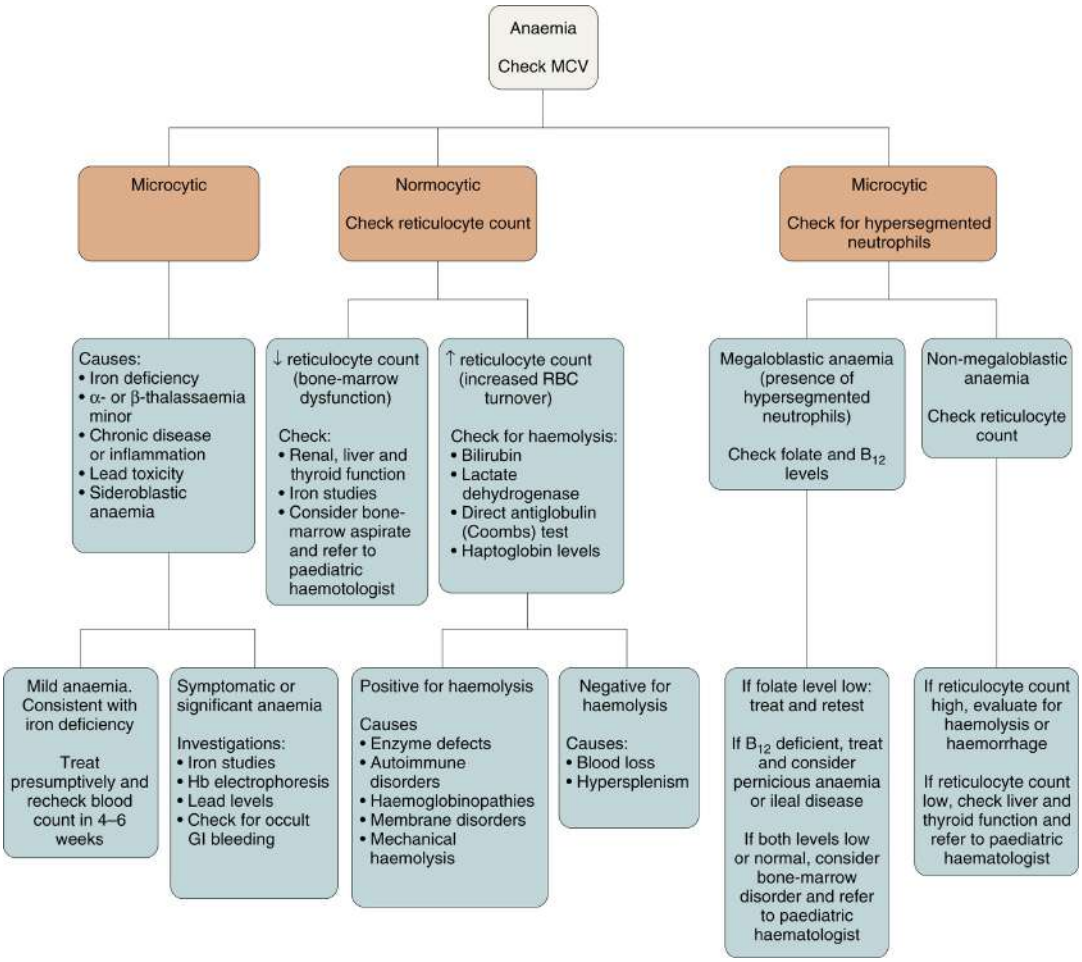

FIG. 11.2.1 Evaluation of anaemia.

Table 11.2.1

Morphological features of red blood cells to assist the diagnosis of anaemia

Schistocytes	Erythrocyte fragmentation syndromes
Sickle cells	Sickle cell disease
Spherocytes	Immune-mediated haemolytic anaemiaHereditary spherocytosis
Blister or bite cells	G6PD deficiency

Immune related

Immune-related haemolysis is the commonest cause of anaemia in the neonate. Haemolysis occurs when maternal antibodies targeting fetal RBCs pass through the placenta. The commonest antigen responsible for haemolytic disease of the

newborn is the RhD antigen on the RBC surface, followed by ABO incompatibility. Sensitisation of the RhD-negative mother occurs when RhD-positive fetal cells gain access to the maternal circulation. Fetal RBCs can be found from 8 weeks' gestation onwards. As little as 0.05–0.10 mL is enough to cause primary sensitisation, but the volume and risk increase with obstetric procedures, miscarriages and pre-eclampsia. Fetal harm occurs during subsequent pregnancies with an RhD-positive fetus in an RhD-negative mother. The resultant intravascular haemolysis in the fetus can be devastating. The clinical significance is due both to the severity of the anaemia and kernicterus from the hyperbilirubinaemia. ABO incompatibility produces less severe jaundice and anaemia during the second week of neonatal life, while many other maternal antibodies are also associated with haemolytic disease of the newborn.

Prevention relies on identification of the Rh status of all pregnant women. This is an essential task for any doctor involved in antenatal care. Prophylactic therapy using anti-D immunoglobulin (Ig) is given to all RhD-negative women with vaginal bleeding or requiring any obstetric procedures that may lead to fetomaternal haemorrhage. The recommended dose is 250 IU during the first trimester and 625 IU beyond this gestation period. This passive immunisation is given intramuscularly. If required in the postpartum period the immunoglobulin is administered intravenously.

Neonatal infection

Congenital infections cause anaemia by producing bone-marrow suppression of erythropoiesis or by haemolysis. The infections of relevance in the neonate are cytomegalovirus (CMV), rubella, toxoplasmosis and congenital syphilis. Human parvovirus may lead to spontaneous miscarriage in early pregnancy and in late pregnancy produces severe fetal anaemia by selectively depressing erythropoiesis. In the older child, parvovirus is the cause of erythema infectiosum (fifth disease), which can be complicated by aplastic crisis or chronic haemolysis. Vertical transmission of malaria is a major cause of neonatal anaemia in endemic regions. Human immunodeficiency virus (HIV) infection in the neonate is predominantly acquired via perinatal transmission from the mother. Without treatment, the transmission rate is in the range of 20–40%, although appropriate medical therapy decreases this to <2%.

Haemoglobinopathies and enzymopathies

G6PD deficiency is an important global cause of neonatal jaundice. Haemolysis from beta globin variants, such as β-thalassaemia major, and SCD is uncommon in the neonatal period as these conditions only become apparent when the proportion of fetal haemoglobin falls. Conversely, α-thalassaemia syndromes may manifest clinically in the newborn period. It is important to consider congenital causes of anaemia in children with a family history of hereditary anaemia or persisting neonatal jaundice.

Blood loss

Blood loss may arise from twin-to-twin transfusion, obstetric procedures, or as part of the delivery of the neonate in the perinatal period. Haemorrhage from an underlying bleeding disorder must be considered. Newborns are at increased risk of vitamin K deficiency, so vitamin K prophylaxis is routinely given to all infants at birth to prevent bleeding. Breast milk has only a small amount of vitamin K, and in the breast-fed baby, maternal medications such as warfarin, phenytoin, rifampicin and isoniazid may compound the deficiency. Nutritional vitamin K deficiency has been associated with prolonged breast-feeding without supplementation, malabsorption, chronic diarrhoea, and prolonged use of oral antibiotics. Vitamin K prophylaxis is normally given at birth. Severe haemophilia can occasionally cause spontaneous haemorrhage in the newborn.

Anaemias of childhood

Iron deficiency

In the normal term infant, total body iron changes little during the first 4 months of life. Iron deficiency is uncommon during this period due to the efficient recycling of iron from the haem component of haemoglobin and an overall drop in haemoglobin concentration. However, iron stores fall after 6 months of age if the infant is not introduced to adequate dietary iron. Iron deficiency occurs earlier in the preterm and low birth weight neonate, often at the time of doubling of the birth weight. Iron deficiency is found in 20% of 6–12-month-old Australian infants, with associated anaemia in 3%. In the second year, iron deficiency is found in 35% and anaemia in 9%. It is even more common in indigenous children and children from non-English speaking backgrounds. In school-age children, iron deficiency is present in 1–2% but is found in up to 9% of adolescent girls.

Stages

The first stage of iron deficiency is a low serum ferritin level. In stage 2, iron-dependent body functions, including erythropoiesis, are affected. Transferrin increases, while transferrin saturation and serum iron decrease. The third stage is the development of microcytic hypochromic anaemia.

It is important to note that serum iron values can be quite variable, making ferritin a more reliable measure of body iron stores. However, iron deficiency can be masked in unwell children presenting to the ED because ferritin is an acute phase reactant which rises with inflammation or chronic disease. Sick children with an incidental finding of microcytosis should have their iron studies performed or repeated once they have recovered from their acute illness. Alternatively, an empiric trial of iron supplementation could be considered, with appropriate follow-up from the child's usual doctor.

Causes

The most common cause globally of iron deficiency anaemia is insufficient intake of dietary iron. In developing countries, the two most significant factors are the failure to obtain iron-rich nutrients and malabsorption from intestinal pathology.

The aetiology is different in developed countries, such as Australia, where prolonged breast-feeding beyond 6 months of age without supplementation with iron-rich foods is a common finding. Breast milk has easily absorbable iron, but it is present only in low concentration, and the quantity is inadequate for the rapid growth that occurs in the second 6 months of life.

Cows' milk is not recommended under the age of 12 months. It has minimal iron content, and the iron present is in a poorly absorbable form. In addition, cows' milk can cause occult gastrointestinal bleeding in the immature gut, and children who consume excessive volumes of cows' milk often have a lower appetite for other foods. Children aged 1 to 5 years should not drink more than 600 mL of milk per day. Iron-fortified cereals should be introduced from 5 months of age.

Adverse effects

While most children with mild to moderate iron deficiency are asymptomatic, iron deficiency before 24 months can lead to impaired development of cognitive and psychomotor skills. This may be only partially reversible when the

deficiency and associated anaemia are corrected. Some parents report loss of appetite, irritability, and inability to concentrate in affected children. Pica may be present. Iron deficiency affects neutrophil and lymphocyte function in vitro and is indicated as one of the investigations of recurrent infections.

Diagnosis

Iron deficiency is the commonest cause of microcytic hypochromic anaemia and is characterised by a low serum iron, high transferrin and low transferrin saturation. The reticulocyte count is less than 2% unless iron has been reintroduced into the diet. Treatment with iron supplementation leads to a reticulocytosis within 1–2 weeks and a rise in the haemoglobin over 2–4 weeks. Other causes of microcytic anaemia must be considered if microcytosis persists despite iron supplementation, although the most common reason for treatment failure is poor adherence.

Treatment

Dietary advice and education are the basis of prevention and treatment in developed countries. However, while increased dietary iron helps maintain iron stores, it is usually insufficient to replenish diminished iron stores. Children with proven or suspected iron deficiency should be given oral iron supplementation as syrup or tablets at a dose of 2–3 mg kg day of elemental iron.

In young children, Ferro-Liquid™ (contains 6 mg elemental iron per mL) can be prescribed 0.3–0.45 mL kg day. If iron deficiency is severe, then bd or tds dosing can be utilised to administer a total daily dose of up to 0.9 mL kg day. Side effects of nausea, vomiting and constipation are dose related. Supplementation will need to be continued for at least 3 months, and reticulocyte count and ferritin levels should be checked 4 weeks after treatment commencement.

Even in severe nutritional iron deficiency anaemia, transfusion is only ever required if there is cardiovascular compromise. Intravenous iron infusions can be useful in moderate to severe cases that have failed oral iron therapy. Parents should be informed of the toxicity of iron in overdose so that they store the medication safely.

Haemolytic anaemias

RBCs exist in the circulation for approximately 120 days with up to 1% of the

RBC population being removed by haemolysis and replaced per day. Pathological haemolysis is that which occurs outside of this normal process leading to reduced RBC survival. Bone-marrow activity increases in response to increased haemolysis, which leads to an increased reticulocyte count of >2%. In older children, the bone marrow's capacity to increase erythropoiesis can compensate for mild pathological haemolysis, but in the infant and young child, the bone marrow is maximally active with minimal ability to increase erythropoiesis, and anaemia accompanies pathological haemolysis.

The haemolytic anaemias of childhood may be classified as hereditary or acquired (see Box 11.2.1). The hereditary haemolytic anaemias are caused by inherited traits that lead to abnormalities of either haemoglobin chains (haemoglobinopathies), RBC membranes (membranopathies) or RBC enzymes (enzymopathies). The acquired haemolytic anaemias may have an immune or non-immune cause or occur from structural damage as a result of fragmentation syndromes.

The most helpful investigation to distinguish AIHA from non-immune causes is the direct antiglobulin test (DAT) (also known as the Coombs test). This test detects a coating of immunoglobulins or complement components on the RBC surface.

Several other laboratory tests support a diagnosis of haemolytic anaemia. Lactose dehydrogenase (LDH) is elevated due to release from lysed RBCs, and an unconjugated hyperbilirubinaemia is observed. During haemolysis, free haemoglobin is liberated in plasma and combines with haptoglobin (causing reduced serum haptoglobin levels), and the resulting complex is cleared mainly by the liver. If there is intravascular haemolysis, free haemoglobin is released and excreted into the urine, resulting in haemoglobinuria. The urine colour may vary from rosé through brown to almost black. Urine dipstick testing does not differentiate haemaglobinuria from haematuria, but the absence of RBCs on urine microscopy supports the former.

Acquired haemolytic anaemias

Autoimmune haemolytic anaemia

Autoimmune haemolytic anaemia (AIHA) can be primary or secondary, with infections such as Epstein–Barr virus, CMV, parvovirus, and mycoplasma being a common cause of secondary AIHA in children. While a history of recent viral infection is frequently described, the specific organism is not always identified.

Other diseases that trigger autoantibodies include systemic lupus erythematosus, rheumatoid arthritis, thyrotoxicosis, ulcerative colitis, malignancy and immunodeficiency syndromes.

Children presenting with AIHA are often acutely unwell with rapid development of pallor, weakness and, sometimes, dark urine. While many cases are well compensated, a life-threatening anaemia may develop with circulatory failure and hypoxia. Haemoglobin drops to less than 60 g L in over 50% of cases. Most children recover completely within 3 months, but some may follow a chronic and relapsing course which confers a more guarded prognosis.

The blood film usually shows spherocytes and polychromasia in warm AIHA and RBC agglutination in cold AIHA. Most cases exhibit reticulocytosis, but 10% may have reticulocytopenia. Associated immune thrombocytopenia suggests a diagnosis of Evan's syndrome. A positive DAT confirms an immune aetiology, and subsequent laboratory investigations distinguish warm versus cold autoantibodies, thereby guiding a more specific diagnosis. Antibodies that are maximally active at 37°C belong to the IgG class and are known as warm antibodies. Antibodies that are maximally active between 0°C and 30°C belong to the IgM class and are known as cold antibodies. Serology may be useful in identifying the infective agent.

Therapy depends on the type of AIHA and the severity. Warm AIHA responds to steroid therapy, and RBC transfusion is not always necessary. Transfusion should be considered for children with cardiovascular compromise, but often cross-matching can be particularly difficult for the laboratory, and occasionally transfusion can result in severe haemolysis. Steroid therapy is not effective in cold agglutinin (IgM induced) disease, which may require plasmapheresis or exchange transfusion if severe. Splenectomy may be required for chronic AIHA. However, this should be avoided in young children due to the risk of sepsis with encapsulated organisms, while older children should be vaccinated against *N. meningitidis*, *S. pneumoniae* and *H. influenzae* pre-splenectomy and continue long-term penicillin prophylaxis post-splenectomy. A haematologist should always be consulted regarding specific treatment.

Non-immune haemolytic anaemia

Non-immune haemolytic anaemia can be caused by a variety of infectious agents (malaria, gram-positive and gram-negative organisms), medications (salicylates, sulfasalazine, nitrofurantoin) and chemicals (naphthalene). Clinical features are

similar to AIHA, but DAT is negative. Treatment is supportive, focusing on the identification and treatment of the underlying cause.

Erythrocyte fragmentation syndromes

These are haemolytic anaemias due to direct physical or mechanical damage to the erythrocytes. They are characterised by red cell fragmentation with schistocytes in the peripheral blood film and features of intravascular haemolysis. There are two groups:

Cardiac haemolytic anaemia

Prosthetic heart valves, patches and grafts can be associated with fragmentation haemolysis. Damage is from a combination of shearing forces from turbulent flow and interaction between the RBCs and the abnormal surfaces and is less likely with xenografts or prostheses that are covered by endothelium. The anaemia is usually mild, and severe haemolysis is uncommon.

Microangiopathic haemolytic anaemia

Microangiopathic haemolytic anaemia (MAHA) occurs when RBCs sustain damage as they pass through diseased or partially occluded arterioles and capillaries. It is often acute and severe and associated with thrombocytopenia. The most common causes are haemolytic uraemic syndrome (HUS) and thrombotic thrombocytopenic purpura (TTP). MAHA is also seen in disseminated intravascular coagulation, certain connective tissue diseases, haemangiomas and HELLP syndrome. Haemolytic uraemic syndrome is further discussed in Chapter 16.5.

Thrombotic thrombocytopenic purpura

Thrombotic thrombocytopenic purpura (TTP) is a rare, life-threatening thrombotic microangiopathy which causes significant morbidity and mortality unless promptly recognised and treated. TTP is caused by a deficiency of ADAMTS13 activity, either due to congenital deficiency or development of autoantibodies. ADAMTS13 is a metalloprotease synthesised in the liver that cleaves ultra-large von Willebrand factor multimers, which otherwise cause spontaneous aggregation of platelets in small blood vessels, resulting in microvascular thrombosis and damaged RBCs.

While hereditary TTP can occur at any age, this entity is extremely rare.

Acquired TTP is usually not seen prior to adolescence. TTP was originally characterised by a ‘pentad’ of thrombocytopenia, MAHA, neurological abnormalities, renal impairment and fever. However, the majority of patients do not have all five clinical features, and TTP should be suspected in the presence of MAHA (red cell fragmentation on blood film) and thrombocytopenia alone. Neurological findings may be as subtle as confusion or headache, with major neurological features, such as hemiparesis, hemiplegia or seizures, occurring in only 35% of cases.

Treatment is with plasma exchange, which has reduced mortality rates of TTP from over 90% to 10–20%. It allows removal of autoantibodies and replenishes ADAMTS13. Delay in initiation of plasma exchange leads to preventable early mortality and is recommended to commence within 4–8 hours. Other treatment options include high-dose corticosteroids and the anti-CD20 antibody rituximab.

Hereditary haemolytic anaemia

Glucose-6-phosphatase dehydrogenase deficiency

Glucose-6-phosphatase dehydrogenase (G6PD) plays an important role in protecting cells from oxidative damage by enzymatically reducing nicotinamide adenine dinucleotide phosphate, which in turn maintains glutathione in its reduced form. Reduced glutathione is an essential factor in the degradation of cellular peroxides that may otherwise damage cellular proteins including haemoglobin. Consequently, G6PD deficiency renders the RBC vulnerable to haemolysis when exposed to oxidative agents. The enzyme activity is higher in the younger RBCs but deteriorates as the cell ages, so older RBCs are more prone to haemolysis.

G6PD is an X-linked recessive condition with wide variability of expression which is dependent on the biochemical type, the oxidative stress and the gender of the patient. Homozygous females are rare, and the disease is predominantly found in males. The severity of the disease varies, and levels of enzyme at or above 40% of normal will rarely result in clinically significant haemolysis. A number of different molecular variants are found in patients of African, Chinese and Mediterranean descent.

Acute haemolysis can be precipitated by viral or bacterial infections as well as oxidant drugs and chemicals, including naphthalene, sulphonamides, antimalarials, nitrofurantoin, diazoxide and dapsone. Favism refers to the acute haemolysis resulting from the ingestion of broad bean (*Vicia faba*) or inhalation

of the pollen. The haemolytic agent in the broad bean can be passed to the infant by breast milk.

Most of the time, individuals with G6PD deficiency are asymptomatic. However, following exposure to a precipitant, dark-coloured urine, jaundice and symptoms of anaemia develop within 3–36 hours and usually last for 2–6 days. Death can occur within 24 hours of severe haemolysis. The degree of anaemia is moderate to severe with an associated brisk reticulocytosis, and the blood film reveals characteristic bite cells. Definitive diagnosis of G6PD deficiency is via a quantitative assay of the enzyme, but the results of a screening test are often available in a more timely fashion. Children with a haemoglobin of less than 70 g L, or less than 90 g L with ongoing haemoglobinuria, should be transfused promptly. Prevention is the cornerstone of management, with avoidance of precipitants and prompt treatment of infections.

Hereditary spherocytosis

Hereditary spherocytosis (HS) is a heterogeneous group of genetic disorders most common in individuals of northern European ancestry and is generally inherited in an autosomal dominant pattern. Inherited abnormalities in various RBC membrane proteins, such as spectrin and ankyrin, result in a progressive loss of membrane surface area, forming a spherical RBC rather than the usual biconcave disc shape. Abnormal spherocytes are trapped in the spleen and destroyed prematurely. The eosin-5-maleimide binding test has largely replaced the osmotic fragility test for diagnosing HS.

Clinically, HS is characterised by anaemia, jaundice (including prolonged neonatal jaundice) and splenomegaly, but the severity is variable with most patients having a well-compensated haemolytic anaemia. Some individuals are asymptomatic, whereas a minority have severe haemolytic anaemia requiring frequent RBC transfusions. Common complications are cholelithiasis, haemolytic crises (increased haemolysis in response to viral infections), and aplastic crises (severe anaemia secondary to parvovirus B19). Splenectomy is curative but should only be undertaken after careful assessment of the risks and benefits. It is usually reserved for patients with moderate to severe HS and, where possible, is deferred to age 5–9 years.

Haemoglobinopathies

Haemoglobin is a protein consisting of two pairs of globin chains, each with a

haem molecule. Adult haemoglobin (HbA), which predominates from about 1 month of age, contains two α chains and two β chains ($\alpha_2\beta_2$). Abnormalities of the globin genes or globin chain production may result in significant disease.

Thalassaemias

Thalassaemia refers to a heterogeneous group of inherited disorders of haemoglobin synthesis. Reduced or absent synthesis of one pair of globin chains results in imbalanced globin chain production. The unaffected chains tend to aggregate, precipitate and cause damage to the RBCs. The nomenclature depicts the missing (or reduced) chain; for example, β-thalassaemia refers to a deficiency of the β-chain. Inheritance is in a Mendelian recessive manner. Thalassaemias are found commonly in people originating from the Mediterranean, Middle East, Indian subcontinent, Africa and southeast Asia. Haemoglobin electrophoresis defines the haemoglobin variants.

β-Thalassaemia minor

Individuals with β-thalassaemia minor (heterozygous β-thalassaemia) carry one abnormal gene only and produce enough β-globin chains, and hence normal haemoglobin, to avoid symptomatic anaemia. The RBCs are microcytic and hypochromic, and target cells may be observed. The life span of the RBCs is slightly decreased, and the haemoglobin is typically between 90 g L and 110 g L. Elevated HbA_2 level in the range of 3.5–7.0% is almost diagnostic of the β-thalassaemia trait. These individuals are asymptomatic, and no treatment is required, but genetic counselling is very important to minimise the risk of having children with homozygous β-thalassaemia.

β-Thalassaemia major

Affected individuals are homozygous for abnormal β-globin genes, receiving one from each carrier parent. Production of β-chains is reduced or absent. Symptoms develop as levels of fetal haemoglobin, HbF ($\alpha_2\gamma_2$), fall in the first month of life and there is increased reliance on adult haemoglobin, HbA ($\alpha_2\beta_2$). Excess unpaired α-chains aggregate to form an unstable tetramer, which is functionally useless and leads to haemolysis. Without treatment, severe chronic anaemia develops leading to growth retardation and extramedullary haematopoiesis which results in hepatosplenomegaly and abdominal distension. Bone-marrow expansion results in skeletal deformities and a characteristic facial

appearance. Regular transfusions will ameliorate all of these problems, and an adequately transfused and chelated child will grow and develop normally.

The peripheral blood film demonstrates a marked microcytic hypochromic anaemia with striking poikilocytosis and anisocytosis, target cells and nucleated red cells. Basophilic stippling may be seen in the RBCs.

Treatment is regular (usually monthly) blood transfusions aimed at suppressing the ineffective haematopoiesis and maintaining a functional haemoglobin level. The major complication of regular transfusions is iron overload. Without adequate iron chelation, transfusion haemosiderosis is associated with cardiac and hepatic failure, diabetes and other endocrine dysfunction, and death in young adulthood. While subcutaneous iron chelation with desferrioxamine very effectively prevents these complications, the onerous infusion schedule impacts upon adherence. Newer oral iron chelators have substantially improved the quality of life of transfusion-dependent patients. HLA-matched bone-marrow transplantation has been curative but is infrequently performed for this indication in Australasia.

α-Thalassaemia

Two genes on chromosome 16 code for the α-chain, producing four possible phenotypes. Deletion of one α-gene is asymptomatic (silent carrier: - α/α α), with a minority having a microcytic hypochromic anaemia. Deletion of two α-genes produces mild anaemia (α-thalassaemia trait: - α/- α or --/α α). Deletion of three α-genes is known as haemoglobin H disease (--/-α), where aggregation of β-chains results in unstable haemoglobin. Chronic haemolysis occurs with anaemia in the range of 70–110 g L. Jaundice and hepatosplenomegaly are common. Cholelithiasis and leg ulcers may develop. Deletion of four α-genes precludes the production of any α-chains and is known as haemoglobin Bart or hydrops fetalis. It generally results in intrauterine death and stillbirth, usually after 25 weeks' gestation. Live births have severe anaemia, gross oedema and congestive cardiac failure and typically succumb soon thereafter. The haemoglobins present in this condition are HbH(β_4), Hb Bart (γ_4) and small amounts of Hb Portland ($\zeta_2\gamma_2$). None of these is functional.

Sickle cell disease

Sickle cell disease (SCD) is an autosomal recessive inherited condition in which glutamine is replaced by valine in the sixth position of the globin chain. In the

homozygote with HbSS, the haemoglobin is unstable in the de-oxygenated state and precipitates in the RBC, with the normal biconcave disc of the RBC changing into a shape resembling a sickle. This change is initially reversible but becomes irreversible with time and causes multiple detrimental consequences. Sickle cells have a reduced life span of 10–20 days and occlude the microvasculature, leading to end-organ ischaemia. The physiological precipitants for the sickling are tissue hypoxia or acidosis, dehydration, vascular stasis or increased levels of 2,3-diphosphoglycerate (2,3-DPG) in the RBC. Clinically, acute crises may occur spontaneously or may be precipitated by infection, dehydration, hypoxia, sedatives, local anaesthetics and surgery. In addition, all patients become functionally asplenic and are at risk for infection, particularly by encapsulated organisms such as *S. pneumoniae*, *N. meningitidis* and *H. influenzae*. Patients with SCD may present acutely with painful vaso-occlusive crises, fever/sepsis, acute chest syndrome, acute splenic sequestration, aplastic crisis, stroke, or priapism. Many of these presentations require urgent treatment, and it is strongly recommended that patients are discussed with a paediatric haematologist. Many patients are aware of their condition, but not all are, and an acute complication of undiagnosed SCD should be considered as a differential for any child of African, Mediterranean or Middle Eastern descent who presents with fever, splenomegaly, pain, breathlessness or new neurological symptoms.

Vaso-occlusive crises are acute episodes of severe pain from tissue infarction resulting from vessel occlusion by the sickled cells. The major organs involved are bones, lungs, liver, spleen, brain and the penis. Painful bone crisis is the commonest, with minimal signs on examination. Treatment of pain should be rapidly initiated with non-steroidal anti-inflammatory drugs (NSAIDs) and opioid analgesics. Intravenous (IV) fluids should be utilised to maintain euvolaemia. Frequent recurrent episodes may be reduced by using regular hydroxyurea. Chest pain should be assumed to be acute chest syndrome rather than vaso-occlusive crisis.

Acute chest syndrome (ACS) is an acute illness characterised by fever and/or respiratory symptoms, accompanied by a new pulmonary infiltrate on chest X-ray. In some cases ACS may be difficult to distinguish from a standard respiratory infection or asthma, but ACS may follow a severe clinical course and can progress rapidly from mild hypoxia to respiratory failure and death. Due to the potential severity of illness, ACS needs to be recognised early and managed aggressively. Patients should be treated with antibiotics (third-generation cephalosporin and oral macrolide) and given supplemental oxygen, IV fluids and

analgesia as required. Consideration of a blood transfusion should be undertaken in consultation with a paediatric haematologist.

Fever should be managed aggressively in patients with SCD as they are at increased risk of serious bacterial infection from reduced splenic function. Take blood cultures and commence empiric broad-spectrum IV antibiotics in all children with SCD who have a temperature ≥38.5°C).

Stroke should be considered in any child with SCD presenting with severe headache, altered conscious state or new neurological abnormalities. Urgent neuroimaging should be performed, and, in those with confirmed stroke, exchange transfusion should be arranged with a paediatric haematologist.

Priapism is a common complication of SCD, affecting 35% of boys and men. It is usually of the low-flow ischaemic type and characterised by pain and a soft glans. Prompt recognition of priapism and initiation of conservative medical management may lead to detumescence and limit the need for more aggressive and invasive intervention. Delayed diagnosis and therapy can result in impotence. Administer aggressive oral or IV hydration and oral or intravenous analgesia for episodes of priapism lasting 4 hours or longer. Consult with a urologist for an episode of priapism lasting 4 hours or longer, and consult with a haematologist for possible preoperative transfusion if surgical intervention is required. Do not use transfusion therapy for immediate treatment of priapism associated with sickle cell disease.

Dactylitis is frequently the first manifestation of pain in children with SCD, occurring in 50% of children by 2 years of age. Children present with symmetric painful swelling of the hands and/or feet and require careful management with pain medication. Differential diagnosis of osteomyelitis should be considered carefully if the presentation is unilateral.

In *aplastic crisis*, the reticulocyte level falls to less than 1%. The haematocrit may fall as rapidly as 10–15% per day. The precipitant is usually an infection. Spontaneous recovery is usual. Supportive therapy may involve transfusion of RBCs.

Acute splenic sequestration (ASS) is a life-threatening complication of SCD defined as a sudden enlargement of the spleen and a reduction in haemoglobin by at least 20 g L below baseline. It occurs in 7–30% of patients under the age of 2 years with homozygous SCD and has also rarely been reported in heterozygous cases. ASS can occur suddenly with acute pallor, weakness, abdominal pain and distension with features of hypovolaemic shock. The precipitant is usually an intercurrent infection. The key finding is circulatory shock in a child with

atraumatic abdominal pain and distension. A massive tender spleen is unmistakable. This is a medical emergency, and intravenous fluid resuscitation must be commenced immediately. Transfusion should then promptly be discussed with a paediatric haematologist, as well as treatment of the potential infective precipitant and analgesia. ASS is recurrent in 50% of cases, with the interval between episodes being less each time. Splenic sequestration of RBCs affects those SCD sufferers who are young enough to still have a functional spleen, as autosplenectomy from recurrent infarction commonly occurs. Hepatic sequestration may also occur, but the volume of trapped RBCs is unlikely to lead to devastating hypovolaemia due to the tight hepatic capsule.

Sickle cell trait

Sickle cell trait denotes the heterozygous or carrier state. These patients usually have blood count that is within the normal range. Haemoglobin analysis is diagnostic, with HbS of about 40% and HbA 60%. The RBCs have a normal life span. Serious complications in sickle cell trait are very rare but may include sudden death during rigorous exercise, splenic infarcts at high altitude, haematuria and bacteriuria. The life span of people with sickle cell trait is normal, and children with sickle cell trait should not have any restrictions placed on activities.

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Further reading

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11.3

Disorders of coagulation

Lalith Gamage

ESSENTIALS

1 Although haemorrhagic disorders due to abnormalities of coagulation are relatively uncommon, their early recognition and accurate diagnosis are important, as many of them require specific treatment.

2 Haemophilia A and B and von Willebrand disease account for the majority of inherited coagulation disorders.

3 The best screening test for a bleeding disorder is a comprehensive history and physical examination.

4 Always consider 'hidden' potentially life-threatening bleeding: intracranial bleeding is a major cause of mortality in the haemophiliac patient.

5 If bleeding or bruising raises concerns for non-accidental trauma, careful history, physical examination and detailed description of physical findings are warranted.

Haemophilia

Introduction

Haemophilia A and B are X-linked recessive disorders (70% of cases) characterised by deficiencies of coagulation factors VIII and IX but can occur as spontaneous mutations (30% of cases). Acquired haemophilia is rare and occurs when autoantibodies develop against factor VIII.

Clinical presentation

Haemophilia A and B are clinically indistinguishable. The severity of haemophilia is based upon the concentration of FVIII and FIX. In severe haemophilia (factor levels less than 1%), bleeding episodes are frequent and often spontaneous in the absence of trauma. Moderate haemophilia (factor levels 1–5%) typically presents with bleeding symptoms after minor trauma; spontaneous bleeding such as haemarthroses is not as frequent as in severe haemophilia unless precipitated by trauma. Mild haemophilia (factor levels 5–40% present) typically manifests as bleeding symptoms with surgery or significant trauma.

Intracranial haemorrhage (ICH) is relatively rare compared with other sites of bleeding, but it is a life-threatening event in individuals with haemophilia. Spontaneous ICH occurs in infants as well as adults. Importantly, many neonates with spontaneous ICH do not have a known family history of haemophilia. ICH can occur immediately after trauma or as a delayed complication days to weeks later.

Hemarthrosis is the most common site for bleeding in ambulatory patients, representing up to 80% of haemorrhages. Forehead haematomas (‘goose-eggs’) have been reported as a common presenting finding of children with haemophilia.

Bleeding can occur from numerous oropharyngeal sites such as the nose, oral mucosa and gingiva; sometimes this type of bleeding follows minor trauma or dental procedures. Frenulum and oral injuries are also common sites in young toddlers.

Haematomas of the bowel and abdominal wall and retroperitoneal bleeding can also occur, producing symptoms that mimic acute appendicitis or produce obstruction or intussusception. Haematuria is a frequent manifestation of severe haemophilia.

Investigations

A complete blood count and evaluation of the peripheral blood picture usually constitute the first step. Screening patients with haemophilia usually demonstrates a prolonged aPTT with normal PT/INR value. Diagnosis can be confirmed by demonstrating a low or absent FVIII or FIX coagulant activity.

Specific imaging is required in particular clinical settings. This can include

CT scans for suspected intracranial bleeding, ultrasound for suspected retroperitoneal bleeding, and plain films to exclude fractures associated with traumatic haemarthroses.

Treatment

Treatment needs to be administered expeditiously in the case of acute bleeds to prevent both short-term complication and long-term disability. Early consultation with a paediatric haematology team to guide treatment is recommended.

Factor replacement therapy using FVIII and FIX concentrates are the backbone of management for patients with severe haemophilia. The dose of concentrate to be administered is calculated based on the haemophilia subtype, baseline factor activity, and desired increase in the factor level. Factor concentrates are available as virus-inactivated plasma-derived products and recombinant factor concentrates. When availablilty and expense allows, recombinant factor concentrates should be used.

The half-life of FVIII is 8 to 12 hours, and that of FIX is 12 to 24 hours. Factor concentrates may be administered as bolus doses or continuous infusion.

Mild superficial bleeding in patients with mild to moderate haemophilia may be managed with local measures. Compression, use of a gelatin sponge or gauze soaked in tranexamic acid may be tried for superficial wounds. Epistaxis may be managed by nasal packing and use of topical thrombin and/or fibrin sealant gel to provide an immediate scaffold for clot formation.

Desmopressin (DDAVP), a synthetic analogue of the antidiuretic hormone vasopressin, exerts its pro-coagulant effect by causing the release of stored vWF and FVIII from endothelial cells into plasma thus increasing the circulating level of FVIII and VWF. For patients who respond, DDAVP may be used to treat minor bleeds and for prophylaxis before dental and minor surgical procedures.

Lysine analogues, such as tranexamic acid, are antifibrinolytic agents usually used in combination with other therapeutic modalities (FVII or FIX concentrate or DDAVP). They are particularly useful in patients with epistaxis, gingival bleeding, and menorrhagia and may be used for prevention of bleeding after minor surgical and dental procedures.

Development of inhibitors is currently the most serious complication of haemophilia. Inhibitors are neutralising alloantibodies that develop in 30% or more of patients with haemophilia A and about 2% to 5% of patients with haemophilia B. Acute bleeds in patients with low-titer inhibitors may be

managed by giving high doses of FVII or FIX. Permanent eradication of inhibitors can be achieved using immune tolerance induction.

von Willebrand disease

Introduction

von Willebrand disease (vWD) is the commonest inherited disorder of coagulation and is present in 1% of the population in screening studies, although the prevalence of clinically symptomatic vWD is probably closer to 1 in 1000. In most cases inheritance is autosomal dominant with variable penetrance and expressivity, though autosomal recessive inheritance also occurs (type 2N and 3). Rarely, vWD may be acquired and has been described in conditions such as hypothyroidism and SLE. von Willebrand factor (vWF) is synthesised by vascular endothelial cells and megakaryocytes and plays an important role in both the primary and secondary haemostatic pathways; it facilitates platelet adhesion and stabilises FVIII in circulation, protecting it from degradation by activated protein C. Platelet aggregation is mediated by both vWF and fibrinogen.

Three types of vWD and three subtypes have been described: type 1 (70–80% of cases) and type 3 (rare) are characterised by partial and virtually complete deficiency of vWF, respectively; type 2 vWD (subtypes 2A, 2B, 2M, 2N) reflects qualitative defects in vWF function.

Clinical presentation

Clinical presentation varies substantially, depending on the subtype and severity, and manifestations range from mild mucocutaneous bleeding to haemarthrosis. Abnormal function of vWD mostly affects platelet plug formation; thus, the clinical manifestations of disease are similar to those seen in platelet disorders such as easy bruising, skin bleeding and prolonged bleeding from mucosal surfaces.

Haemarthroses and intracranial and intraabdominal bleeds are much less common but can occur in more severe types 2N and 3 vWD. vWD is diagnosed in 10–20% of women and adolescents with menorrhagia and in 15% of children with recurrent epistaxis.

Investigations

vWD is diagnosed most often if the patient satisfies three criteria: a positive bleeding history, reduced level of vWF activity, and a positive family history suggestive of vWD. A coagulation screen may be normal and does not rule out vWD. In most patients aPTT is normal, though a prolonged aPTT can occur in types 2N and 3, with reduced factor VIII levels. Bleeding time may be prolonged. Screening tests for vWD include vWF assay, ristocetin co-factor assay and FVIII activity.

Because vWF is an acute-phase reactant, concentration increases with stress, exercise, acute inflammation, in pregnancy and during the menstrual cycle. The plasma concentration of vWF is also modified by other determinants such as blood group and race.

Treatment

Two primary options are available to treat spontaneous bleeding episodes and for bleeding prophylaxis in vWD: DDAVP and therapy with plasma-derived vWF and FVIII products. Treatment is usually given in response to bleeding or prophylactically prior to surgical or dental procedures.

DDAVP, a synthetic analogue of vasopressin, releases FVIII and vWF from endothelial storage sites, causing a transient rise in levels. Over 80% of patients with type 1 vWD will increase plasma levels of vWF between two to five times from baseline within 30–60 minutes as a response to DDAVP, although the response in type 2 vWD is less predictable. DDAVP is not beneficial in type 3 vWD, and given the risk of exacerbating the thrombocytopenia, it is usually contraindicated in type 2B vWD. Similar to haemophilia, it is important to perform a DDAVP challenge test before using it in a clinical setting.

The dose of DDAVP is 0.3 μg kg (maximum 20 μg) given intravenously/subcutaneously or an intranasal dose of 150 μg for children weighing <50 kg or 300 μg for children weighing ≥50 kg. The most common adverse effects are facial flushing, headache, tachycardia and hyponatraemia, which usually respond to slowing the infusion. Repeat doses can be given at 12–24 hours if needed, but tachyphylaxis occurs after approximately 48 hours.

Due to the risk of hyponatraemic seizures, DDAVP should be used with utmost caution in young children (<2 years of age) and children undergoing surgical procedures associated with significant blood loss.

Simultaneous use of antifibrinolytic agents (ε-aminocaproic acid, 50 mg kg q 6 hours or tranexamic acid 25 mg kg q 4 to 6 hours) prevents plasmin-medicated clot lysis and leads to clot stabilization. Oestrogen-containing oral contraceptive pills may be useful for adolescent females with menorrhagia.

Controversies and future directions

The management of haemophilia is likely to change with the development of long-acting FVIII and FIX concentrates and gene therapy. Further studies evaluating the safety and long-term efficacy of gene therapy are under way.

vWD disease treatment options for the future include recombinant vWF, which has shown promising early results in animal trials. Interleukin-11 has also been shown to increase vWF, but human trials are needed. Oestrogen ointment has been used internationally on a prophylactic basis for paediatric patients with recurrent epistaxis with some success.

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11.4

Platelet disorders

John Craven, and Kottayam Radhakrishnan

ESSENTIALS

1 Platelet dysfunction clinically mostly manifests as mucocutaneous bleeding, including abnormal bruising, petechiae, purpura or epistaxis.

2 Platelet disorders can be qualitative, due to disorders of platelet function, or quantitative due to deficiency in platelet numbers.

3 Immune thrombocytopaenia (ITP) is the most common quantitative platelet disorder in childhood but only requires treatment under specific circumstances.

Introduction

Children with platelet disorders present with symptoms and signs of mucocutaneous bleeding including bruising (petechiae or purpura), epistaxis, bleeding from the oropharynx or gastrointestinal tract, and menorrhagia. Accurate assessment and diagnosis are vital to ensure that management of the child is appropriate.

A normal platelet count is 150–400 × 10^9 L^{-1}, and platelets usually exist within the circulation for 5–7 days. In normal haemostasis, blood loss is initially limited by the formation of a platelet plug followed by cross-linking of fibrin at the site of injury. In platelet disorders the platelet plug is not formed or is ineffective, and abnormal bleeding occurs. Platelet abnormalities can be either quantitative due to deficiency in platelet numbers, or qualitative, due to disorders of platelet function.

The most common platelet disorder in children is thrombocytopaenia (low platelet count), which may be acute or chronic. Bleeding complications usually only occur at levels of less than 50×10^9 L^{-1}, with spontaneous bleeding more likely at levels below 20×10^9 L^{-1}. Falsely low platelet counts (pseudothrombocytopaenia) can result from the platelet clumping that can occur in ethylenediaminetetraacetic acid (EDTA) collection tubes. The platelet clumping can be confirmed by direct inspection of the peripheral blood smear. Testing blood collected in tubes with citrate as the anticoagulant can overcome this problem.

In qualitative disorders of platelet function, abnormal bleeding occurs despite a normal platelet count. Platelet dysfunction may be acquired in hepatic failure, chronic renal failure, myeloproliferative disorders, and with some drugs (e.g. aspirin) and occurs in rare inherited disorders of platelet function (Chédiak-Higashi, Glanzmann thrombasthaenia, Hermansky–Pudlak, May–Hegglin and Bernard–Soulier syndromes). Assessment of platelet function is complex, and advice from a paediatric haematologist should be sought as to the most appropriate tests.

Thrombocytosis (elevated platelet count) may be reactive as is seen in inflammatory conditions including infection, Kawasaki disease, malignancy, inflammatory bowel disease and in non-inflammatory conditions including iron-deficiency anaemia, post-splenectomy and certain medications (e.g. vincristine). A rare cause of thrombocytosis in children is myeloproliferative disorders, such as essential thrombocythaemia, where there may be extreme thrombocytosis with platelet counts >1000 x 10^9 L. Reactive thrombocytosis, irrespective of the platelet count, rarely causes thrombosis.

Immune thrombocytopaenia

Introduction

Immune thrombocytopaenia (ITP), previously known as idiopathic thrombocytopenic purpura, is an autoimmune disorder most commonly occurring in response to an unknown stimulus. ITP is caused by increased platelet destruction at a rate that exceeds production by the bone marrow, and there is evidence that platelet production is also decreased in many patients. ITP may occur in isolation (primary) or in association with other disorders (secondary). Secondary causes include autoimmune diseases (particularly the

antiphospholipid antibody syndrome), viral infections (including hepatitis C virus and human immunodeficiency virus), certain drugs and following vaccination, particularly the MMR vaccine. Rather than using the terms ‘acute’ and ‘chronic’, the recommended nomenclature defines ITP as newly diagnosed (diagnosis to 3 months), persistent (3 to 12 months from diagnosis), or chronic (lasting for more than 12 months). In the majority of children (75–80%), ITP is expected to resolve spontaneously within 6 months and, while it can occur at any age group, is most common between the ages of 2 and 6 years. Adolescents and girls with ITP are more likely to develop chronic ITP.

Clinical presentation

ITP typically presents up to 2–6 weeks following a viral infection or vaccination. There is generally a history of unexplained bruising, non-blanching rash (petechiae or purpura) or mucosal bleeding in an otherwise well child. The mucosal bleeding is usually from the gums or nose. Haematuria or gastrointestinal bleeding may less commonly occur. Physical examination should be otherwise normal, with no hepatosplenomegaly or lymphadenopathy.

Diagnosis

The diagnosis of ITP relies on the presence of laboratory-demonstrated thrombocytopaenia (defined as a platelet count less than 100 x 10^9 L) and a history, clinical examination and full blood count and blood film that do not suggest another cause.

Although a few large platelets may be seen on the film in ITP, significant numbers of large or giant platelets should raise a suspicion for inherited/congenital thrombocytopaenia. The blood count and film are otherwise usually normal. A normocytic anaemia may be present, dependent on the extent of previous haemorrhage. Eosinophilia may occur but is not a consistent sign. A few reactive lymphocytes may be noted. Coagulation studies, although not routinely indicated, will be normal with the exception of a prolonged bleeding time.

ITP is a diagnosis of exclusion as there are no specific diagnostic tests. Currently available platelet antibody tests are not indicated since they are neither sensitive nor specific for ITP. Routine testing of antiplatelet, antiphospholipid, and antinuclear antibodies, thrombopoietin levels or platelet parameters is not

generally recommended. Bone-marrow aspiration is no longer recommended for diagnosis but may be performed if there is concern for underlying malignancy, bone-marrow abnormality or in chronic ITP.

Other causes of thrombocytopaenia should be considered if there are additional symptoms. An underlying lymphoproliferative disorder should be considered in the presence of limb pain, hepatosplenomegaly or lymphadenopathy. NAI must be considered in infants with bruising. In Wiskott–Aldrich syndrome, eczema is present in the first year of life, and platelets are smaller than normal. The presence of other congenital abnormalities, particularly those of the skeleton (short stature), skin (pigmentation) and other systems, might be a clue to Fanconi's anaemia. A systemic autoimmune disorder such as lupus or antiphospholipid syndrome should be considered particularly in older children with a chronic course. Additional testing should be performed in consultation with a paediatric haematologist.

Management

Management is directed by bleeding symptoms rather than specific platelet counts. Most children have only cutaneous or mild mucosal bleeding and recover spontaneously within 6–8 weeks. They require no treatment and can be managed as ambulatory patients. Parents should be provided with good written advice on the condition and given a telephone contact name and number. Written advice should include information about avoiding contact sports, risk of head injury, aspirin and other drugs that interfere with platelet function. They should watch for bleeding and report to hospital if new episodes occur.

In the initial stages the platelet count can be reviewed weekly, but this can be extended to every 2–4 weeks once it is improving. By 6 months around 75–80% will have a normal platelet count. Patients with platelet counts greater than $150 \times 10^9\ L^{-1}$ can be discharged from follow-up but warned that occasional relapses are seen and to re-present if symptoms recur.

Admission should occur for all patients with active bleeding, particularly of the gastrointestinal or renal tracts, and should be considered for patients with initial platelet counts below $20 \times 10^9\ L^{-1}$. Serious bleeding is rare in ITP, with intracranial haemorrhage occurring in 0.5–1% of patients, most often with platelet counts less than $20 \times 10^9\ L^{-1}$. The decision to admit may be influenced by social circumstances and parental care issues.

Current treatments

Over the past decade, recommendations for initiation of treatment in ITP have changed with the advent of new treatment options and greater evidence for them.

Current treatments

Asymptomatic children with acute ITP and platelet count $<20 \times 10^9$ L^{-1} should be carefully observed. A single dose of intravenous immunoglobulin (IVIG) or a short course of corticosteroids is the recommended first-line treatment for patients with significant bleeding symptoms.

IVIG infusion (0.8–1 g kg) will generally lead to a more rapid platelet count rise than corticosteroids, usually within 48 hours, lasting for 2–4 weeks. As a pooled blood product IVIG carries the risk of viral transmission and is expensive. Various side effects have been reported.

Oral corticosteroids are an effective treatment option; however, there is no evidence to support one dose or duration of treatment over others. A common treatment regime is prednisolone 2 mg kg day for 2 weeks after which it is tapered. Long-term use should be avoided due to the association with many adverse effects.

Anti-D has been shown to be effective in Rh-positive children; however, the platelet increase is slower, taking 48–72 hours. Anti-D is recommended only in patients who are Rh positive, who have a negative direct antiglobulin test (DAT), and who have not undergone splenectomy. Anti-D use is associated with intravascular haemolysis, and while there is currently little evidence to support a specific dose, the currently recommended dose is 50–75 mcg kg.

Second-line treatments are considered for patients with newly diagnosed ITP with severe bleeding, unresponsive first-line treatments or with chronic or persistent ITP. Rituximab or high-dose dexamethasone would be more commonly recommended, but numerous agents such as azathioprine, danazol, interferon, mycophenolate mofetil, cyclosporine and anti-CD52 monoclonal antibody have been used with scant evidence. Among the newer agents, thrombopoietin receptor agonists (TRAs) are the most promising and are now being used in clinical trials.

It is now recommended that splenectomy, the long-recognised gold standard second-line therapy, be delayed for at least 12 months unless the child has severe and unresponsive disease or quality of life concerns that mandate more definitive

therapy. Post-splenectomy patients have an increased risk of sepsis despite pneumococcal vaccination and antibiotic prophylaxis.

Medical emergencies

The risk of intracranial haemorrhage is less than 1% but persists as long as the platelet count is very low. Children with ITP and severe headache with neurological signs, especially in the setting of head trauma, require urgent investigation with cranial CT scan and, if indicated, immediate treatment with IVIG and IV high-dose corticosteroids. If clinical suspicion of intracranial haemorrhage is high, treatment should be started immediately while waiting for radiological confirmation. Life-threatening haemorrhage is the only instance where platelet transfusions should be used, as the antibodies in ITP target all platelets including donor transfusions. As a result the benefit of any transfusion is very short lived. Emergency splenectomy may be used as a last resort.

Chronic immune thrombocytopaenia

Around 15–20% of children will go on to develop chronic ITP; however, remission will occur in up to two-thirds of these patients in subsequent years. A chronic course is most often seen in older children, particularly adolescent girls, or in secondary ITP with underlying conditions such as lupus or antiphospholipid syndrome. Most do not have significant bleeding problems or a need for regular treatment.

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11.5

Vasculitis

Evelyn Doyle

ESSENTIALS

1 Vasculitic syndromes are rare in childhood. The two most common are Henoch–Schönlein purpura (HSP) and Kawasaki disease (KD), which are discussed in separate chapters of this book.

2 Childhood vasculitis can be a primary disease process or secondary to an underlying disease, such as infection, inflammatory disorders, drug reaction or malignancy. Diagnosis and treatment of the underlying cause are important.

3 Presentation of vasculitis in childhood can vary widely, and features may include fever, malaise, weight loss, skin rash, arthritis, myalgia, hypertension, proteinuria and abdominal pain, making the diagnosis difficult.

4 Vasculitis should be considered in the differential diagnosis in children with a non-specific febrile illness, with systemic manifestations of disease and raised inflammatory markers without other explanation.

Introduction

Childhood vasculitides are a heterogeneous group of disorders that manifest in multisystem disease characterised by inflammation of blood vessels and may lead to tissue ischaemia and necrosis. Most forms of vasculitis are rare in childhood; primary systemic vasculitis (PSV) affects less than 25 per 100,000

children, with Henoch–Schönlein purpura (HSP) and Kawasaki disease (KD) being the two commonest.

Other forms of vasculitis – although rare – are important as they are often associated with significant morbidity and mortality mainly due to progressive renal failure or aggressive respiratory involvement. They include polyarthritis nodosa (PAN), Takayasu's arteritis (TA) and the ANCA-associated vasculitis (AAV), such as granulomatosis with polyangiitis (GPA) (previously known as Wegener's granulomatosis), microscopic polyangitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA, previously known as Churg–Strauss) (Box 11.5.1). With the wider use of immunosuppressive agents, both mortality and morbidity rates have improved. However, many patients still develop significant treatment- and disease-related morbidities, and mortality remains high.

Box 11.5.1 Classification of childhood vasculitis

Predominantly large-vessel vasculitis:
- Takayasu arteritis

Predominantly medium-sized-vessel vasculitis:
- Polyarteritis nodosa
- Cutaneous polyarteritis
- Kawasaki disease

Predominantly small-vessel vasculitis

Granulomatous:
- Granulomatosis with polyangiitis
- Eosinophilic granulomatosis with polyangiitis

Non-granulomatous:
- Henoch–Schönlein purpura
- Microscopic polyangiitis
- Hypocomplementaemic urticarial vasculitis

Other vasculitides:
- Secondary vasculitis (due to infection, connective tissue disease, drugs and malignant disease)
- Behçet disease
- Central nervous system vasculitis
- Cogan syndrome

Unclassified

Modified from Ozen et al.

Clinical presentation

The presentation of vasculitis varies widely according to the size of the affected blood vessel and the extent and nature of organ involvement. Nonspecific constitutional symptoms are common to all vasculitides and include fever, weight loss, malaise, irritability, arthralgia and myalgia. Skin manifestations are frequent and often give a clue to the size of the vessel involved; lesions are commonly purpuric but may be urticarial, ulcerative and vesicular or resemble erythema multiforme. In severe vasculitis, vessel destruction leads to tissue infarction and can result in ischaemia and gangrene. The other symptoms depend on the organs involved: joints, respiratory, renal and gastrointestinal systems are commonly affected.

Classification

The primary vasculitic disorders can be divided according to the size of the vessel involved and subdivided into granulomatous, non-granulomatous, and ANCA-associated vasculitis (AAV).

Small-Vessel Vasculitis

Henoch–Schönlein purpura

Small-vessel vasculitis includes Henoch– Schönlein purpura (HSP), a systemic small-vessel vasculitis involving the skin, kidney, joints and gastrointestinal tract. It is the commonest systemic vasculitis in childhood. The aetiology is unknown; however, HSP is believed to be an immune-complex-mediated disease characterised by the presence of polymeric IgA-containing immune complexes predominantly in dermal, gastrointestinal and glomerular capillaries. It is discussed in detail in Chapter 16.7.

Anti-cytoplasmic-antibody-associated vasculitis

This group of vasculitides are usually positive for anti-cytoplasmic antibody (ANCA) and includes GPA, MPA and EGPA. The incidence of ANCA-associated vasculitis (AAV) in children is extremely low. GPA is the commonest

of the AAV in children and has an incidence of approximately 0.3–0.6 per 100,000/year. MPA and EGPA are extremely rare in childhood.

Granulomatosis with polyangiitis

GPA is a vasculitis with necrotising granulomatous inflammation affecting predominantly small vessels. Typical age of onset is early adolescence, with a female preponderance. Relapses are frequently observed in GPA. In a recent review, the paediatric mortality in GPA and MPA was found to be 7%, mostly from disease-related and infectious complications. Respiratory complications were the leading causes of disease-related death in children with GPA.

Classification criteria for childhood GPA are met when at least three of the six following features are present: (1) histopathology (granulomatous inflammation and/or necrotising pauci-immune glomerulonephritis); (2) upper airway involvement (nasal discharge, epistaxis, granulomata, nasal septum perforation or saddle nose deformity, sinus inflammation); (3) subglottic, tracheal or endobronchial stenosis; (4) pulmonary involvement (abnormal chest X-ray or chest CT scan); (5) ANCA positivity; and (6) renal involvement (haematuria and/or significant proteinuria or other renal disease).

Clinical presentation

In GPA, disease onset and severity vary between patients and earlier diagnosis, and treatment can prevent life-threatening complications. It mainly affects the respiratory tract (sinuses, nose, trachea, lungs) and kidneys, but the disease can involve any organ system. Among children with GPA constitutional symptoms such as lethargy, weight loss, and fever are very common.

Inflammation of the upper respiratory tract may include sinusitis, discoloured or bloody discharge from the nose and nasal ulcers. Nasal ulceration may result in a hole developing in the cartilage of the nose, which can lead to collapse (saddle-nose deformity). Granulomas may be seen on inspection of the nasal mucosa. A common sign of the disease is almost constant rhinorrhea that does not respond to usual treatment. The eustachian tubes may become blocked, causing chronic ear problems and hearing loss.

The lungs are affected in most people with GPA, although no symptoms may be present. If symptoms are present, they include cough, haemoptysis, shortness of breath, and chest discomfort. Subglottic airway obstruction is more commonly seen in children than adults.

In children with GPA, renal disease occurs in about 75%; usually it is asymptomatic, but renal disease may progress to kidney failure. Of 117 children described by Cabral et al., 42% had significant renal involvement, 19% severe respiratory involvement requiring continuous oxygen therapy, and 11% required mechanical ventilation.[1]

Nearly half of children with GPA develop skin lesions. These often have the appearance of small red or purple raised areas or blister-like lesions, ulcers or nodules that may or may not be painful. Myalgia and arthralgia affect two-thirds of children with GPA.

Diagnosis

A positive ANCA has a sensitivity of 93% and specificity of 90% for GPA in childhood. There are two main types of ANCA that differ according to the patterns of immunofluorescent staining. ELISA should always be performed in patients with positive results on immunofluorescence to identify the specific antigen target; presence of c-ANCA with anti-proteinase 3 (anti-PR3) is highly suggestive of GPA.

Targeted imaging of symptomatic patients may show evidence of sinus inflammation (CT sinuses) or lung nodules, cavities or fixed infiltrates in GPA (chest X-ray or CT).

Renal involvement may be present and suggested by proteinuria, haematuria and necrotising pauci-immune glomerulonephritis. Biopsy and histopathology with granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area may be seen in GPA.

Microscopic polyangiitis

Microscopic polyangiitis (MPA), an ANCA-associated vasculitis, is extremely rare in childhood, and its pathogenesis is poorly understood. The multisystem nature of MPA results in considerable variability in the clinical presentation, and diagnostic delay is common. MPA in childhood has a female predominance of 80% and is usually diagnosed in adolescence. Relapses in MPA are less frequent than GPA and are seen in less than half of patients. The use of glucocorticoids, cyclophosphamide and other immunosuppressive agents has favourably changed the prognosis of MPA, but mortality remains high.

Renal disease is observed at onset of illness in almost all children with MPA, and 20% have progression to ESRD (end-stage renal disease) or a need for

dialysis during follow-up. In MPA systemic features are also frequent and seen in three-quarters of patients at presentation, and 50% of patients present with musculoskeletal symptoms. ENT involvement is rare in MPA, with sinusitis the only feature observed; this may help to distinguish between GPA and MPA.

MPA causes pauci-immume necrotising vasculitis (few or no immune deposits) affecting small vessels, necrotising glomerulonephritis, and pulmonary capillaritis may occur with the absence of any granulomatous inflammation ANCA is usually positive for perinuclear ANCA (pANCA), which is typically directed against myeloperoxidase (MPO-ANCA).

Eosinophilic granulomatosis with polyangiitis (formerly Churg-Strauss)

Eosinophilic granulomatosis with polyangiitis (EGPA) is the rarest of the ANCA-associated vasculitides and primarily affects small vessels. There is not a paediatric classification system due to paucity of data in children. In adults the majority have an established diagnosis of asthma or develop it within 6 months of diagnosis of EGPA. Other clinical features seen are peripheral neuropathy and ENT signs in about half of patients; skin lesions and lung infiltrates also occur commonly as does cardiomyopathy in about 15%. Among adult patients about one-third are ANCA positive. ANCA positivity is associated with more frequent ENT manifestations, peripheral neuropathy, and/or renal involvement but less frequent cardiac manifestations. ANCA positivity is also associated with a higher relapse rate but a lower mortality rate than with ANCA-negative disease. Eosinophilia >10% on differential white blood cell count and biopsy containing a blood vessel with extravascular eosinophils are diagnostic criteria in adults.

Medium-Vessel Vasculitis

Kawasaki disease

Kawasaki disease (KD) was first described by Kawasaki et al. in 1974 and is an acute systemic vasculitis that mainly affects small to medium vessels with a predilection for the coronary arteries.[2] It mainly affects children less than 5 years of age and in developed countries is the leading cause of acquired heart disease in children and is discussed in more detail elsewhere (Chapter 5.9).

Polyarthritis nodosa

Polyarthritis nodosa (PAN) was first described in 1866 by Kussmaul and Maier and is a necrotising vasculitis associated with aneurysm formation in medium-sized muscular arteries.[3] Although rare in childhood, it is the most common form of systemic vasculitis after HSP and KD, and onset in childhood is usually between 7 and 11 years of age, with a male preponderance.

Clinical presentation

PAN can have a wide spectrum of clinical presentations depending on the organs involved. The mortality figures for PAN in children have been quoted as approximately 10%.

The main clinical features seen in PAN include constitutional symptoms common with all vasculitides presentations: malaise, fever and weight loss are very common, as are skin rash, abdominal pain, arthralgia, arthritis and myalgia.

Skin manifestations include painful subcutaneous nodules especially of the feet, palpable purpura that may resemble HSP or erythema multiforme, livedo reticularis, necrotic lesions and peripheral gangrene.

Other manifestations depend on the organs involved. Renal disease can manifest as hypertension, proteinuria and haematuria and less commonly a rapidly progressive nephritis. Abdominal pain is common and thought to represent visceral ischaemia, and testicular pain may be a feature in boys. Rupture of arterial aneurysms can cause retroperitoneal and peritoneal bleeding, with perirenal haematomata being a recognised manifestation.

Cardiac involvement may present as ischaemic heart pain but occurs less commonly. Respiratory and neurological features also occur less frequently. Neurological involvement may present as a peripheral neuropathy and less commonly with central nervous system features, such as seizures or stroke, visual loss and psychosis.

There is a cutaneous form, known as benign cutaneous polyarteritis nodosa (BCPAN) or cutaneous polyarteritis, which has a much better prognosis and may represent one spectrum of the same disease.

The paediatric classification criteria for PAN requires histopathological evidence of necrotising vasculitis or angiographic abnormalities (aneurysms, stenosis or occlusions) in a small-/medium-sized artery as a mandatory criterion plus one of the following five systemic features: skin involvement, myalgia or

muscle tenderness, systemic hypertension, peripheral neuropathy and renal involvement.

Investigation and treatment

Characteristic histopathological evidence of necrotising vasculitis or angiographic or MR angiographic evidence of aneurysm, stenosis or occlusion in affected vessels is a requirement for PAN.

Investigations reveal elevation of inflammatory markers (ESR and CRP) and may include leukocytosis, thrombocytosis, anaemia, proteinuria and haematuria.

A positive ANCA is sometimes seen in PAN, but its presence usually raises the suspicion of a different diagnosis of microscopic polyangiitis. A recent multicentre survey found 6 out of 47 children with PAN had a positive ANCA.

In childhood systemic PAN, remission induction with high-dose steroid and cyclophosphamide is used but has not been tested in a controlled trial. Plasma exchange has been used in severe life-threatening cases; however, effectiveness remains unproven without larger studies. A retrospective review concluded that a good response occurred in four out of five PAN cases treated with plasma exchange. Aspirin has also been prescribed empirically as an antiplatelet agent.

Once remission is achieved, agents used to maintain it include steroids and azathioprine for up to 18 months. Other maintenance agents include methotrexate, mycophenolate mofetil and cyclosporine. More recently, successful treatment with biologic agents such as infliximab or rituximab has been reported for those unresponsive to conventional therapy.

Large-Vessel Vasculitis

Takayasu disease

Takayasu disease is a large-vessel vasculitis of unknown aetiology mainly involving the aorta and its proximal branches. It is a rare vasculitis with a reported incidence of 1.2 to 2.6 per 1 million per year in Caucasians and a 100-fold higher incidence in East Asians. In adults it occurs 10 times more commonly in young women than men. It is rare in children but causes significant morbidity and mortality.

Clinical presentation

Hypertension is the most common presenting symptom in children, warranting careful examination of pulses and blood pressures in all four extremities, with a search for asymmetry and bruits essential.

Morbidity results from stenosis, ischaemia and aneurysmal dilatation of affected vessels. Presentation may be non-specific with constitutional symptoms or specific symptoms relating to inflammation of large vessels, including congestive heart failure, angina, myocardial infarction, stroke, limb claudication and renovascular hypertension found.

The paediatric classification criteria for TA require typical angiographic findings of the aorta or its main branches (including CT or MRI imaging) for the diagnosis and thus a mandatory criterion, plus at least one of the following five features: (1) decreased peripheral artery pulse(s) and/or claudication of extremities; (2) blood pressure difference >10 mmHg in any limb; (3) bruits over the aorta and/or its major branches; (4) hypertension; and (5) elevated acute-phase reactants.

Secondary Vasculitis and Vasculitis Mimics

Secondary vasculitis (SV) is important to distinguish from a primary vasculitis as the treatment will differ significantly. This can be difficult due to the marked variability observed in symptomatology even within different cases of the same form of vasculitis. SV may be seen in a wide array of medical disease; thus it is important to have a wide differential diagnosis when assessing for suspected vasculitis.

Many infectious diseases have been associated with SV, predominantly with cutaneous manifestations. Papulovesicular acrolocated syndrome (PALS or Gianotti–Crosti syndrome) is a vasculitis associated with multiple viral and other triggers, including hepatitis B antigenaemia, Epstein–Barr virus, cytomegalovirus, human immunodeficiency virus, rickettsial diseases and streptococcal infections.

Connective tissue disorders including rheumatoid vasculitis, systemic lupus erythematosus, sarcoidosis, Sjögren’s syndrome, inflammatory bowel disease, drug reaction or malignancy may also present with SV. There is an increased association with SV in patients with familial Mediterranean fever, with several patients described with PAN and HSP features.

Hypersensitivity angiitis is a serum-sickness-like reaction in children that includes a rash, arthropathy and fever. It commonly follows the use of penicillin

or cephalosporins (particularly cefaclor). Skin manifestations include oedema of the dorsum of the hands and feet and a nodular, purpuric, urticarial or erythema-multiforme-like rash. Arthralgia and arthritis are common and respond to non-steroidal anti-inflammatory drugs, with short-term oral steroids reserved for the more severe cases. The condition is self-limiting but recurs on re-exposure to the same agent.

Also any process that causes occlusion of a blood vessel can mimic vasculitis. The clinical appearance of subacute bacterial endocarditis resembles a primary vasculitic process but is secondary to septic emboli.

Controversies and future directions

A paediatric vasculitis activity score (PVAS) has been developed as a tool for the evaluation of systemic vasculitis in children. It is a validated disease activity tool specific to paediatric vasculitis and is the only tool currently used as an outcome measure in paediatric vasculitis clinical trials. This score will be important for the ability to research new therapies and assess a patient's response to therapy in the future.

Genetic markers in vasculitis continue to be discovered and may provide more clues to the pathogenesis. This may assist with treatment options as we learn more about these rare conditions.

Prospective cohort studies with novel therapies are crucial to try to improve management of this complex group of patients.

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11.6

Acute leukaemia

Joseph Ting, and Kottayam Radhakrishnan

ESSENTIALS

1 At its most severe, acute leukaemia leads to diffuse bone-marrow infiltration and failure. Pancytopenia, including neutropaenia, anaemia and thrombocytopenia, as well as extramedullary haematopoiesis, gives rise to classical symptoms of infection susceptibility, easy fatigability, bleeding diathesis and hepatosplenomegaly. Hepatosplenomegaly related to acute leukaemia at presentation is due to infiltration by blast cells and less often due to extramedullary infiltration.

2 Emergency department supportive treatment in conjunction with a paediatric haematologist may include intravenous fluids to reduce the risk of tumour lysis syndrome, corticosteroids for spinal cord compression, broad-spectrum antimicrobial administration and/or sepsis resuscitation for infection, packed red blood cell transfusion for symptomatic or critical anaemia and platelet replacement if the child has high potential or actual risk of thrombocytopenia-related bleeding.

Introduction

Leukaemia results from the uncontrolled proliferation of immature and abnormal white cells and abnormal white blood cell precursors of varying haematopoietic lineage and aberrant differentiation, which results in death within 1–6 months without treatment.[1] Leukaemia originates in the bone marrow, where normal

blood cells are replaced by leukaemic cells. One-third of all cancers in children are leukaemias, with incidence peaking at 2–5 years of age.

Pathogenetic factors include chromosome instability/fragility, immunodeficiency and certain environmental exposures (radiation, chemicals, viruses). The occurrence in all age groups during childhood, grouped by type of leukaemia is as follows:

- 75% acute lymphoblastic leukaemia (ALL)
- 20% acute myeloid leukaemia (AML)
- 5% undifferentiated acute leukaemia and chronic myeloid leukaemia (CML).

Leukaemia differs in the lineage and degree of differentiation of cells involved, being broadly divided into two groups: lymphoid and myeloid. These two categories are further subdivided into an acute form that progresses more rapidly than the chronic disease, which is relatively indolent. Preconceptional germ cell and postnatal environmental exposure to electromagnetic radiation and carcinogens may play a role,[1–4] with higher risk in children with congenital neutropaenia, Down's syndrome and Fanconi anaemia.[5] Previous chemotherapy and radiotherapy are associated with increased risk of a secondary malignancy.

Classification

The classification of leukaemia is complex. Leukaemogenesis represents clonal proliferation of white blood cells arrested at various stages of differentiation, lending itself to diagnostic and prognostic classification based on the cell line affected, degree of differentiation, immunophenotyping, cell receptor, cell antigen and chromosomal studies. The WHO classification of neoplasms of the haematopoietic and lymphoid tissues, published in 2001 and updated 2008, represents a worldwide consensus on the diagnosis of these tumours.[6]

The major principle of the classification is the recognition of distinct diseases according to a combination of morphology, immunophenotype, genetic, molecular and clinical features. The disease entities are stratified according to cell lineage and derivation from precursor or mature lymphoid cells.

Clinical presentation

Signs and symptoms in ALL and AML are similar and depend on the degree of marrow infiltration and extent of extramedullary involvement. The first symptoms are non-specific including lethargy, bone pain and loss of appetite. Classic findings in acute leukaemia include fever/infection, pallor/lethargy/easy fatigability and bruising/mucosal bleeding due to febrile neutropaenia, anaemia and thrombocytopenia, respectively.

Marrow infiltration leads to bone pain, and extramedullary spread leads to painless adenopathy (including mediastinal), hepatomegaly, splenomegaly, skin or periorbital infiltrates and rash. **Bone pain is more common in children with ALL than AML.** Difficulty with diagnosis arises in differentiating early acute from chronic disease, when symptoms (night sweats, arthralgia, bone pain, constitutional symptoms) and signs (enlarged liver and spleen, lymphadenopathy) are variable and non-specific.

Central nervous system (CNS) involvement occurs in 4% of children (cranial nerve palsy, cord compression) and testicular leukaemia in 10% of boys (painless enlarged testes). **In children, CNS involvement at diagnosis occurs more frequently with ALL (4%) than AML. Testicular leukaemia at presentation rarely occurs in AML.**

Hyperviscosity results in tissue infarction (CNS, pulmonary) and rarely priapism results from leucocyte sludging and cavernosal obstruction.

Differential diagnosis

Pancytopenia may result from acute leukaemia, aplastic anaemia, marrow infiltration by non-haematological malignancy and collagen-vascular disease. However, hepatosplenomegaly and adenopathy are unusual with the alternative diagnoses.

Viral infections may lead to lymphadenopathy and hepatosplenomegaly accompanied by atypical lymphocytes rather than blast cells.

Leukaemoid reactions with leucocyte counts $>50 \times 10^9\ L^{-1}$ may rarely occur with acute infections and inflammatory disease such as rheumatoid arthritis.

Marked lymphocytosis without leukaemic cells occurs in pertussis.

Isolated thrombocytopenia in childhood may follow a viral infection or be associated with idiopathic or thrombotic thrombocytopenic purpura rather than leukaemia.

Investigations

Full blood count reveals anaemia and thrombocytopenia in 80% of cases. The majority of children have leucocyte counts below $20 \times 10^9\ L^{-1}$. Those with leucocyte counts up to $900 \times 10^9\ L^{-1}$ usually **have blast infiltration leading to hepatosplenomegaly and lymphadenopathy.** The neutrophil count is often depressed to below $1.0 \times 10^9\ L^{-1}$. **Acute leukaemia presenting with isolated thrombocytopenia without other cytopenia, clinical or blood stream abnormalities is extremely rare.** Circulating blast cells are frequently present on blood film examination. Blood typing is necessary if red-cell transfusion is anticipated. A screen for atypical antibodies is warranted if there have previously been multiple occasions of blood product use.

Cultures of blood and other potential infection sites are obtained prior to antimicrobial administration. Positive blood cultures are uncommon in newly diagnosed acute leukaemia. Fever at presentation is commonly a manifestation of leukaemia itself rather than underlying sepsis.

Tumour lysis syndrome due to massive cell death during treatment causes raised lactate dehydrogenase (LDH), liver enzymes, hyperkalaemia, hyperuricaemia, hyperphosphataemia, hypocalcaemia and acute renal failure.

Anterior mediastinal masses are visible on chest X-ray in 5–10% of cases, and pneumonia may require exclusion.

Bone-marrow aspiration and trephine are essential for specific diagnosis and prognostication, including complex tests, such as immunophenotyping, cytogenetics, molecular studies and cell-cycle kinetics.

AML is, rarely, complicated by a coagulopathy that progresses to disseminated intravascular coagulation so that a coagulation profile and fibrinogen level may be helpful.

Prognosis

The overall survival rate in ALL is 80%,[7] compared with only 30–50% for AML.[8] High-risk features in paediatric ALL include age <1 year or >10 years, high initial WCC, CNS involvement at presentation, unfavourable cytogenetics and suboptimal induction response (induction failure or positive minimum residual disease).

Poor-risk AML has a dismal outlook, with less than 20% achieving long-term remission. Adverse prognostic factors include chromosome 5 and 7 abnormalities as well as complex cytogenetic abnormalities.

Complications

Hyperleukocytosis

Leukostasis is the sludging that develops in the microcirculation of the central nervous system (CNS) and lungs in the setting of hyperleukocytosis, which can lead to life-threatening complications. The risk of complications related to leukostasis increases with increasing white blood cell (WBC) count. Prompt initiation of chemotherapy is the most effective approach to the treatment of hyperleukocytosis.

Tumour lysis syndrome

Hyperkalaemia, hyperphosphataemia and hyperuricaemia occur with lysis of a huge burden of malignant cells. Risk factors for tumour lysis syndrome are large tumour burden, pre-existing hyperuricaemia, renal impairment and dehydration. The mainstay of treatment is **aggressive hydration** to prevent tumour lysis syndrome (TLS) prior to commencement of chemotherapy. The goal is to induce high urine output to minimise uric acid or calcium phosphate precipitation in the renal tubules. A 2008 International Expert Panel on TLS recommended that both adults and children receive 2 to 3 L m^2 per day of IV fluid, to facilitate 2 mL kg^{-1} $hour^{-1}$ of closely monitored urine output.[9]

Allopurinol, which competitively inhibits xanthine oxidase and reduces uric acid production, is used to treat children at intermediate risk of TLS. Rasburicase is safe and effective for the prevention of TLS in children.[10]

Superior vena cava and superior mediastinal syndrome

In patients with cardiovascular or respiratory compromise, emergency corticosteroid administration may be necessary before a diagnosis is established.

Spinal cord compression

This occurs in 3–5% of children with cancer, with symptoms including motor deficits, diplegia or quadriplegia, sensory deficits, sphincteric abnormalities and back pain. When spinal cord dysfunction is suspected, dexamethasone 1–2 mg kg should be given immediately.

Management

Management requires close collaboration with a paediatric haematologist. Emergency department supportive treatment focuses on the sequelae of acute leukaemia: febrile neutropaenia and sepsis (broad-spectrum antimicrobial agents), actual or potential bleeding from thrombocytopenia (platelet transfusion) and correction of acute or symptomatic anaemia, which are described in Chapter 11.1. Blood products should ideally be irradiated and cytomegalovirus negative and screened for atypical antibodies, more so for repeated transfusions.

Chemotherapy may give rise to bone-marrow suppression, similar to the sequelae of acute leukaemia. Children who relapse or do not achieve remission are candidates for bone-marrow transplantation. Barrier nursing, isolation, antimicrobial and antiviral chemoprophylaxis, vaccination and post-exposure administration of immunoglobulin (e.g. varicella zoster virus) and careful monitoring for early infective complications help prevent complications related to immunosuppression. Psychosocial support of the child and family is important.

Controversies and future directions

1 The future management of leukaemia will be influenced by new and improved risk-stratified therapy to improve outcomes and minimise adverse effects. High-risk patients could be identified by molecular analysis, as well as in vitro pharmacodynamic, pharmacogenetic, pharmacogenomic and drug-resistance studies to receive more intensive and targeted therapy.[11] Identifying new biomarkers for risk and target identification may make personalised medicine a reality in the near future.

2 The increasing use of severely myelotoxic chemotherapy and unrelated bone-marrow transplantation has improved prognosis in acute leukaemia but has given rise to complications related to marrow suppression. Chemotherapy frequently causes myelosuppression, mucositis, and nausea and vomiting, which are particularly problematic in children.

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11.7

Febrile neutropaenia

Joseph Ting, and Kottayam Radhakrishnan

ESSENTIALS

1 Aggressive and early resuscitation of the child with overt or incipient septic shock is vital to avert impaired perfusion, even in the presence of normal mental state and blood pressure.

2 Careful physical examination and broad microbiological/radiological investigation to detect and eradicate the source of infection are essential.

3 Early institution of broad-spectrum antimicrobial therapy, followed by targeted culture-guided treatment, contributes to improved outcome.

Introduction

Infection is a major cause of death in children with haematological malignancies following bone-marrow or solid organ transplantation, iatrogenic immunosuppression, and chemotherapy. The major factor predisposing to infection in these patients is neutropaenia, which is defined as decreased circulating neutrophils in the peripheral blood.

Neutropaenia is defined as an absolute neutrophil count <500 mm^3 OR <1000 mm^3 with predicted decline to <500 mm^3 over the next 48 hours.[1] Febrile neutropaenia requires an associated fever ≥38.5°C or ≥38.0°C for at least 1 hour.[1] This chapter focuses on neutropaenia associated with paediatric cancer or its treatment as this is the more frequent type encountered in the emergency department (ED). *Congenital neutropaenia* is managed in a similar way to cancer chemotherapy-induced neutropaenia.

Acquired non-cancer neutropaenia (autoimmune, drug induced, infection associated) requires an individualised – often less aggressive – approach in discussion with a paediatric haematologist.

Neutropaenic children receiving chemotherapy for leukaemia or undergoing bone-marrow transplantation (BMT) are at significant risk from Gram-negative sepsis, including *Pseudomonas aeruginosa* and *Escherichia coli*. In BMT patients, half the infections are bacterial, evenly split between gram-positive (such as *Staphylococcus aureus*) and gram-negative organisms; 40% are due to viruses like cytomegalovirus; and 10% due to fungi, with up to one-third of these episodes due to life-threatening systemic fungaemia. Vascular catheter-related infections are usually related to gram-positive skin organisms. Although the overall mortality rate due to neutropaenia-associated infection is 1%, children undergoing BMT with febrile neutropaenia have a startling mortality of up to 80%.[2]

Management priorities are:

- early recognition of the child with compensated septic shock before onset of hypoperfusion
- aggressive and early resuscitation of the child with overt or incipient septic shock
- careful physical examination and broad microbiological/radiological investigation to detect and eradicate the source of infection
- obtaining blood and other cultures prior to early institution of broad-spectrum antimicrobial therapy, followed by targeted culture-guided treatment
- achieving optimal microcirculatory, metabolic and circulatory conditions using a combination of intravenous (IV) fluids, vasopressors and/or blood products.

Presentation

Young children compensate for impending shock and remain normotensive in the early stages of systemic sepsis. The earliest reliable sign of vascular redistribution is delayed capillary return, because tachycardia and tachypnoea can be secondary to the fever. Neutropaenic or immunosuppressed children may have attenuated or altered signs of infection. The absence of neutrophils to localise an infection makes it difficult to determine the source of infection by

physical examination alone. Pay special attention to body regions vulnerable to bacterial infection, such as the oropharynx, ears, skin, and perianal area. Evidence of respiratory, circulatory, renal and metabolic derangement indicates severe sepsis.

After urgent first-dose antibiotics, a full examination includes:[1]

- upper respiratory tract
- oropharynx
- lower respiratory tract (chest X-ray only if signs and symptoms of pneumonia are present)
- abdomen for localising tenderness including colitis
- perineum and perianal area (without rectal examination) for anal fissure, cellulitis and abscess
- skin for cellulitis and vesicular lesions
- central vascular access device for signs of tunnel/exit site infection.

Investigations

Microbiological samples from potential infection sources such as blood, urine, cerebrospinal fluid, throat, skin and central access devices should ideally be obtained prior to, but not delay, treatment. Similarly, radiological investigation to localise infection source should not delay treatment commencement. Although no microbiological source is identified in one-half of febrile neutropaenic children with cancer, a high proportion have culture-positive bacteraemia, respiratory tract infections and central access device infections.[2]

Initial investigations include:

- Full blood examination (FBE) and differential
- Electrolytes and liver function test (E/LFT)
- Two blood culture sets taken from several peripheral sites or taken from separate draws from one peripheral site
- VBG and blood lactate
- Group and hold
- Urine midstream clean catch (MCS).

A chest X-ray is indicated in the evaluation of the newly febrile neutropaenic paediatric oncology patient only when respiratory signs or symptoms are present

in addition to the high fever.[3]

Additional diagnostic tests may include:

Respiratory:

- Nasal swab or NPA for respiratory virus panel
- Sputum MCS in older children

Diarrhoea:

- Stool for MCS, viral studies
- *C. difficile* toxin assay if recent antibiotics

Skin, CVAD site or mouth lesions:

- Bacterial swab MCS
- Viral swab of vesicular lesions or mouth ulcers for herpes simplex virus (HSV) and varicella zoster virus polymerase chain reaction (VZV PCR).

If central nervous system symptoms are present, neuroimaging and lumbar puncture are reasonable after correction of coagulopathy or thrombocytopenia. If febrile neutropaenia is prolonged for >72 hours, despite broad-spectrum antibiotics, evaluate and consider treatment for invasive fungal infection.

Treatment

Repeated venesections and difficult venous access in paediatric haematology and oncology are a feared painful procedure in children.[4] Many children have CVAD ports, which require trained staff to cannulate for IV use or obtaining blood tests. Despite their advantages, CVAD ports may become a source of systemic infection, so great care should be taken with sterile technique, and access should only be by appropriately trained staff.

If <7 days of access is required, a peripheral cannula is appropriate. Consider a sampling IV line if repeated blood testing is required during admission.

Distress from IV access is reduced in infants <3 months by using oral sucrose with a pacifier or encouraging a feed during the procedure. In non-urgent situations, older infants and children benefit from the application of topical local anaesthetic cream. Nitrous oxide or oral midazolam (0.5 mg kg up to 20 mg) is another option to reduce anxiety during IV cannulation. After two failed attempts at peripheral cannulation, a senior doctor should be called upon to assist and consideration made for intraosseous or central venous access if the clinical

situation is emergent.

IV fluid resuscitation and vasopressor support may be required in septic shock (see Chapter 2.6). Urgent empiric intravenous therapy with combination broad-spectrum antimicrobials that have excellent gram-negative cover is essential in febrile neutropaenia. The pattern and severity of infection vary according to location, clinical context, and level of immune suppression. Regularly reviewed institution- and scenario-specific treatment protocols, developed in conjunction with infectious diseases physicians and microbiologists that take into account local patterns of infection and their susceptibilities, help to deliver and standardise optimal treatment.

The Royal Children's Hospital Melbourne[1] recommends the initial treatment for all children with no known allergies to be ***piperacillin/tazobactam*** *100 mg kg (max 4G) IV q 6 h.*

In the presence of systemic compromise or high-risk cancer treatment or inpatient with onset of febrile neutropaenia (FN), it is recommended to add ***amikacin*** *22.5 mg kg (18 mg kg if >10 years) (max 1.5G) IV q 24 h.*

If there is life-threatening immediate penicillin/beta-lactam hypersensitivity, urgent administration of ***ciprofloxacin*** *10 mg kg (max 400 mg) IV 12 h* AND ***vancomycin*** *15 mg kg (max 500 mg) IV 6 h* is required.

All patients must be given the first antibiotic within 60 minutes (preferably 30 minutes if systemic compromise/sepsis). Time to antibiotic delivery <60 minutes has been shown to be associated with significantly reduced need for ICU consultation or admission.[5] All children must be discussed with their treating paediatric oncology team (or a consultant paediatrician in regional centres) after the first dose of intravenous antibiotic.

Fluid resuscitation in sepsis includes initial 10 mL kg normal saline IV bolus over a maximum of 10 minutes (not through an infusion pump) and considering additional fluid boluses to a maximum of 40 mL kg if only transient improvement occurs.

Controversies and challenges

1 Although outpatient management of febrile neutropaenia has been used in adults, its role, safety and efficacy in paediatric oncology remains contentious. Further research is required to validate the use of outpatient oral antibiotic treatment of selected low-risk children with febrile neutropaenia.

2 The emergence of highly resistant and virulent organisms is increasingly recognised as a barrier to improved outcomes, especially in the context of immunosuppression.

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11.8

Emergencies in paediatric oncology

Peter Downie

ESSENTIALS

1 Paediatric oncologic emergencies can be part of the initial disease presentation, as a consequence of therapy, or at the time of disease progression or relapse.

2 Fever – particularly with neutropaenia – suggests risks for bacteraemia, sepsis and septic shock.

3 Cardiothoracic emergencies include mediastinal masses (superior mediastinal syndrome, superior vena cava syndrome), pericardial effusion and cardiac tamponade.

4 Hyperleukocytosis requires aggressive management to avoid serious morbidity and mortality.

5 Blood product transfusions should be evidence based, and irradiated blood products are necessary to avoid graft-versus-host disease (GVHD).

6 Renal emergencies include obstructive uropathy and nephrocalcinosis.

7 Metabolic emergencies include tumour lysis syndrome, hypercalcaemia, hyponatraemia and syndrome of inappropriate secretion of anti-diuretic hormone (SIADH).

8 Abdominal emergencies include typhlitis, pancreatitis, obstruction, intussusception and drug-induced ileus.

9 Neurological emergencies include seizures, confusion and altered mental state.

10 Brain and spinal cord tumours often have a delayed diagnosis. Awareness of specific signs and symptoms is essential for emergency physicians.

Emergency Complications of Paediatric Malignancy

Introduction

Cancers in children are histologically diverse and can arise in almost any anatomical site. Approximately 1 in 500 children less than 16 years of age is diagnosed with cancer every year. Worldwide, this equates to more than 180,000 children. Childhood cancer is the single most common cause of death in children aged up to 14 years. Table 11.8.1 provides an overview of common types of childhood cancer at various ages.

Children and adolescents with cancer are at increased risk for life-threatening complications either due to their malignancy or its treatment. Early identification, assessment and appropriate treatment of these potentially serious events are important to reduce morbidity and mortality.

Fever and infection

There are three time points to consider when a child with cancer presents with a fever.

1. **At diagnosis.** Children and adolescents with cancer often present with fever; it is either part of the disease process with the release of cytokines (neuroblastoma, Hodgkin disease, leukaemia) or due to infection, secondary to impaired immunity or pancytopaenia (leukaemia, non-Hodgkin lymphoma [NHL], Hodgkin disease [HD]).
2. **During treatment.** A common presentation of the cancer patient to the emergency department is fever in association with neutropaenia (Chapter 11.7).
3. **Relapse.** Similar to the time of initial diagnosis, relapse of malignancy can release cytokines, leading to fever.

Important considerations in children with malignancy and possible infection include the following:

- Patients receiving steroids may have infection without fever.
- The chemotherapy schedule, timing of the drugs, and where the patient is in terms of his/her diagnosis influence the risk for serious infection. For example, acute lymphoblastic leukaemia (ALL) therapy induction is a high-risk period, and the patient will be on steroids, whereas the maintenance phase is relatively low risk.
- Fever should not be assumed to be due to a benign cause such as a simple viral illness.
- Oncology patients are often anaemic and are more sensitive to vasodilation from sepsis, plasma loss from hypoproteinaemia, and free water loss from vomiting and diarrhoea.

Children and adolescents with cancer are prone to infection, particularly during periods of neutropaenia and secondary to immunodeficiency. Patients with fever and neutropaenia can be stratified into risk groups, based on diagnosis and time of treatment, type of chemotherapy given and the absolute neutrophil count. An absolute neutrophil count (ANC) $<500 \times 10^6\ L^{-1}$ is associated with an increasing risk of serious bacterial infection (Chapter 11.7). A significantly higher risk of serious infection occurs during leukaemia induction and post stem-cell transplantation of any type.

Gastrointestinal emergencies

Gastrointestinal obstruction

This is an uncommon presentation and is usually due to one of the following mechanisms:

1. Intussusception is the presenting finding in around 25% of children with abdominal Burkitt's lymphoma.
2. Paralytic ileus can be due to vinca alkaloids (vincristine, vinblastine) or opiates.
3. Mechanical obstruction due to adhesions and strictures, including previous surgery and/or previous perforation.
4. Direct tumour effect is seen in germ cell tumour of the sacrum and

massive hepatomegaly seen in infant neuroblastoma. In the latter, obstruction can impair diaphragmatic function leading to respiratory failure.

Diagnosis is through clinical findings and imaging. Symptoms of bloody stools, intractable vomiting, colicky abdominal pain and the sign of a palpable mass indicate bowel obstruction. Never assume that vomiting is due to previous chemotherapy.

Table 11.8.1

The common malignancies in childhood and adolescence

Age	<12 months	1–4 years	5–9 years	10–14 years	15–19 years	Total
Cancer category	Numbers = percent of total cancers					
Leukaemia – all types	20	33	40	37	23	32
Lymphoma including Hodgkin disease	3	3	6	10	21	11
CNS tumours	13	17	17	21	17	18
Neuroblastoma	21	15	10	4	0.5	6
Wilms tumour	5	9	8	5	0.5	5
Bone tumours	-	-	2	5	10	4
Soft tissue sarcomas	3	3	3	6	8	6
Germ cell tumours	15	5	3	3	8	5
*Other cancers	20	15	11	8	9	13

NB. The most common cancers diagnosed in children and adolescents; pooled data. Numbers are percentages of the total.

* Other cancers include rare tumours – i.e. hepatoblastoma and retinoblastoma in infants, thyroid cancer and an increasing incidence of melanoma in older adolescents.[1,2]

Perforation

Perforation may be due to obstruction that has progressed, ulcers (steroid therapy), erosion from the primary tumour (NHL, sarcoma) or typhlitis (see below).

Children with Burkitt's lymphoma are at particular risk of perforation due to necrosis from pressure of the tumour on the underlying bowel, steroid therapy and especially if there is associated invasive fungal infection.

Gastrointestinal bleeding

If the presentation is with haematemesis or melaena, think of:

1. swallowed blood from epistaxis and thrombocytopaenia or coagulopathy
2. corticosteroid-induced ulcers, either due to induction chemotherapy for

various haematologic malignancies or dexamethasone for raised intracranial pressure (ICP) in brain tumours

3. Mallory-Weiss tears from chemotherapy-induced vomiting (+/- thrombocytopaenia)
4. Oesophageal varices, while rare, are associated with fibrosis from radiation, tumours compressing the portal vein, and infiltration with Langerhans cell histiocytosis.

Pancreatitis

Pancreatitis commonly presents with central abdominal pain and vomiting and is caused by asparaginase, steroids and rarely with administration of 6-mercaptopurine in ALL maintenance therapy. Serum lipase is elevated. Treatment includes IV fluid replacement, bowel rest, nasogastric tube insertion for vomiting and analgesia. As there is often associated neutropaenia, antibiotics may be necessary, particularly if necrosis or perforation is possible.

Veno-occlusive disease of the liver

This is often of rapid onset, with right upper quadrant pain, tender hepatomegaly, ascites and jaundice. It is most commonly seen as a side effect of conditioning regimens for bone-marrow transplant but may also be seen from chemotherapy for various malignancies.

Infection and inflammation

Neutropaenic enterocolitis (typhlitis)

This is a life-threatening, necrotising enterocolitis due to chemotherapy effects on the gastrointestinal tract mucosa, decreased host defences, and invasion of micro-organisms into the bowel wall. It consists of neutropaenia, fever, abdominal pain and diarrhoea, primarily in patients with leukaemia but also in others treated with high-dose chemotherapy. It is strongly associated with oral mucositis.

Disease is centred around the caecum, although it often extends into the ascending colon and terminal ileum. Bacterial or fungal invasion of the bowel wall can progress to full-thickness inflammation, infarction and perforation.

Typhlitis, although difficult to distinguish from appendicitis, must be

considered in the differential diagnosis of any patient with ANC <500 x 10^6 L^{-1} who presents with fever and right lower quadrant pain. Symptoms frequently appear around 10–14 days after high-dose chemotherapy.

It is diagnosed on the basis of clinical suspicion and CT scan. Look for pneumatosis coli, bowel wall thickening, bowel dilatation and mucosal enhancement. Antibiotic treatment should commence with piperacillin-tazobactam and metronidazole.

Surgical intervention is indicated for (1) persistent bleeding despite correction of thrombocytopaenia and coagulopathy; (2) evidence of perforation (i.e. abdominal free gas); and (3) severe sepsis thought to be due to bowel infarction.

Perirectal abscess

Pain on defaecation with constipation and local tenderness may indicate a perirectal abscess. It occurs most commonly during prolonged periods of neutropaenia. The signs may be restricted to indurated tenderness, with little redness; perianal pain and redness are often worse with neutrophil recovery, before resolution.

Gastrointestinal complications of stem-cell transplantation

Cytomegalovirus (CMV) colitis usually presents with abdominal pain, diarrhoea and gastrointestinal bleeding. GVHD may also cause gastrointestinal symptoms, often with associated skin, oral mucosa and liver involvement.

Blood product use in oncology

Irradiation of blood products

Irradiated blood products are necessary for *all* patients who are on chemotherapy due to the risk of transfusion-associated GVHD. Transfusion-associated GVHD occurs as a result of the immunosuppressed patient being unable to reject donor T-cell activation. It has a very high mortality.

Practice guidelines for platelet transfusion

Severe haemorrhage associated with thrombocytopaenia is uncommon, despite many children being treated with drugs that cause myelosuppression. Based on several studies, consensus recommendations for platelet therapy in paediatric

oncology patients are presented in Table 11.8.2.

Cardiothoracic emergencies

Presentation is with respiratory distress, fever and chest pain. Respiratory distress is often the most prominent symptom and may occur in isolation.

Superior vena cava syndrome and superior mediastinal syndrome

Superior vena cava syndrome (SVCS) occurs when there is compression, obstruction or thrombosis of the SVC. Superior mediastinal syndrome (SMS) occurs when there is the addition of tracheal compression. In children and adolescents the terms are interchangeable.

Table 11.8.2

Recommendations for platelet transfusion in paediatric oncology patients

Clinical	Situation	When to transfuse (platelet count)
Well	In leukaemia In solid tumour patients where recovery is unlikely in the next 3 days	<10
For procedures	LP – standard LP diagnosis and to avoid introduction of blasts Insertion of CVC Major surgery Bone-marrow aspirate	10–20* 50–100 50 >100 Unnecessary
Signs and symptoms	Epistaxis or mucosal (mouth, gums) Fever Significant bleeding Tumour necrosis CNS tumours APML induction	10–20 20 75–100 75–100 30–50 50
Other	Hyperleukocytosis DIC/sepsis/coagulopathy	20 50

* Can be lower or without transfusion in properly sedated child and with experienced operator.
APML, acute promyelocytic leukaemia; CVC, central venous catheter; DIC, disseminated intravascular coagulation; LP, lumbar puncture.

The most common symptoms are cough, dyspnoea, orthopnoea; the signs are

tachypnoea, suffusion and oedema of the head and neck, distended neck veins, stridor, wheeze and signs of pleural and/or pericardial effusion.

These symptoms and signs often progress rapidly; symptoms are usually worse when supine, and the child may present sitting up and leaning forward to prevent compression of the trachea.

More than 90% of children with SVCS will have a malignant mass in the anterior superior mediastinum. Common causes include ALL and NHL in children under the age of 10 years and Hodgkin disease HD and germ cell tumours (GCT) in adolescents. Sarcomas (Ewing's sarcoma and rhabdomyosarcoma) and neuroblastoma more often present with a posterior mediastinal mass, which is less likely to cause SVCS. NHL is commonly associated with pleural and pericardial effusions.

A useful mnemonic for causes of a mediastinal mass (and in evaluating SVCS) is GnNASH-T; **G**erm cell tumour, **n**on-Hodgkin lymphoma, **N**euroblastoma, **A**cute leukaemia (T cell), **S**arcoma, **H**odgkin lymphoma.

In the child who presents with SVCS without a mass, think **T**hrombosis (Table 11.8.3). This is a medical emergency. Immediate measures for treatment are:

1. elevation of the head and proper positioning of the patient to minimise the obstruction; usually sitting upright or leaning forward
2. oxygen
3. maintenance of a calm and quiet environment
4. start fluids in hyperleukocytosis, but also diuresis, as there is a risk of pulmonary oedema; beware of tumour lysis syndrome (see below)
5. admission to ICU.

Thrombotic obstruction can occur as a complication of long-term central venous catheters (CVCs). Think thrombosis in the child or adolescent with SVCS and no mediastinal mass. Look for signs of venous congestion in the chest and neck, dilated neck veins, dilated superficial chest veins and oedema of the arms and face. There is no established role for thrombolytics in children. The CVC should be removed and low molecular weight heparin should be started.

Haemoptysis

Haemoptysis is an uncommon presentation, usually caused by invasive aspergillus infection in immunocompromised hosts.

Pneumonia

Lung infections can rapidly lead to respiratory failure in immunocompromised children. In patients without neutropaenia, viral infection is most common; if there is pulmonary infiltrate seen, also think plasma cell pneumonia.

Pleural and pericardial effusions

In children and adolescents, malignant pleural or pericardial effusions are due to cancers with a high turnover rate. These are usually lymphoma (both NHL and HD), any leukaemia with a very high white cell count and germ cell tumours. Bedside ultrasound can readily demonstrate fluid in the pericardial or pleural spaces:

Pleural effusion: Symptoms are dyspnoea, orthopnoea, tachypnoea, chest pain and cough. Large pleural effusions result in respiratory distress; diagnosis is based on clinical findings and chest X-ray. Look for dull percussion note, increased vocal fremitus and resonance, and decreased localised air entry. The patient is often sitting up; look for signs of SVCS as previously described. If neutropaenic, the patient must be started on broad-spectrum antibiotics, as the cause of the effusion may be infective.

Pericardial effusion and cardiac tamponade: Symptoms are cough, chest pain, dyspnoea, hiccoughs, cyanosis and sometimes abdominal pain. Look for Beck's triad (elevated jugular venous pressure, muffled praecordial sounds, hypotension), and pulsus paradoxus (an increase of systolic pressure of >10 mmHg on inspiration). Listen for a friction rub and diastolic murmurs, and check for arrhythmia. Chest X-ray shows a typical 'water-bottle' cardiac silhouette; ECG shows flat T waves, electrical alternans, and low-voltage QRS complexes.

Once a blood count and differential have been done, the diagnosis of leukaemia may then be made; also, if paracentesis is needed, the full blood examination (FBE) will give the platelet count, which should be checked before doing any procedure.

In children, in the absence of severe haemodynamic compromise, pericardial and/or pleural fluid is usually drained under general anaesthetic in the operating theatre. Emergency pericardiocentesis is described in Chapter 24.12, while

pleural drainage is described in Chapter 24.6.

Remember to send the fluid for diagnostic cytology and flow cytometry. Exudates are caused by malignancy and inflammation; there is a high cell count, high protein and high specific gravity. Transudates are due to a sympathetic response to tumour, fluid overload, heart failure or hypoproteinaemia; there is a low cell count, low protein and low specific gravity.

Metabolic emergencies

Tumour lysis syndrome

Tumour lysis syndrome (TLS) is seen in conditions where there is a high cell turnover, particularly lymphoma and acute leukaemia. Rapid breakdown of tumour cells releases large amounts of intracellular contents (phosphate, potassium, nucleic acids and uric acid) into the blood. The metabolic consequences are hyperphosphataemia, secondary hypocalcaemia, hyperkalaemia, hyperuricaemia and, if not corrected, acute kidney damage.

Principles of treatment include generous fluid therapy, reduction of serum uric acid levels using either allopurinol (which blocks xanthine catabolism to uric acid but may lead to accumulation of xanthine) or rasburicase (recombinant urate oxidase) and correction of electrolyte abnormalities (Table 11.8.4).

Table 11.8.3

Clinical and demographic features and recommended initial investigations for mediastinal masses in children

Disorder	Age	Clinical	Tests to order in the emergency department
Germ cell tumour	Older children and adolescents	Dyspnoea, wheeze, SVCS, chest pain, hoarse voice and tracheal deviation Examine boys for Klinefelter syndrome	AFP and β-hCG CXR – may show calcification Chest CT – in consultation with the oncologist
Non-Hodgkin lymphoma	Any	Short history, relatively rapid onset, dyspnoea, orthopnoea, chest pain, SVCS and pleural and pericardial effusion Weight loss, fever – less common	CXR FBE and differential LDH, uric acid, renal function and LFTs DO NOT arrange CT scan: can precipitate tracheal obstruction DO NOT give anxiolytics or conscious sedation Echocardiogram
Neuroblastoma	Young children; mean age 18 months	Horner syndrome (unilateral ptosis, myosis, anhidrosis), periorbital bruising, FTT, skin nodules, abdominal mass, fever, potential neurological signs related to thoracic spinal cord compression (paraplegia, bladder/bowel problems), radicular pain and hypertension	CXR, CT and MRI (spinal infiltrate) FBE, ferritin and NSE Urinary catecholamine analysis Plasma metanephrines
Acute leukaemia	Any	Usually T-cell subtype: dyspnoea, wheeze, SVCS and effusions Headaches – especially with CNS involvement Fundoscopy for papilloedema, infiltrates Look for hepatosplenomegaly, lymphadenopathy, testicular involvement, fever, bruising and bleeding Can be (rarely) AML	CXR FBE and differential: T-cell ALL often presents with a high WCC but relative sparing of Hb and platelets Coagulation screen LDH, uric acid, renal function and LFTs DO NOT arrange CT scan: can precipitate tracheal obstruction DO NOT give anxiolytics or conscious sedation Echocardiogram
Sarcoma	Any: usually older children with Ewing's sarcoma	Cough, dyspnoea, pain (may fluctuate), paraesthesia Slower onset; longer history Neurological signs of thoracic spinal compression	CXR, CT chest and MRI (spine) FBE and ESR
Hodgkin lymphoma	Adolescents, older children	Cough, dyspnoea, wheeze, hoarse voice, central chest pain, orthopnoea, stridor and SVCS Pleural and pericardial effusions are much less common than in NHL Pruritus, weight loss, low-grade fever, hepatosplenomegaly and cervical lymphadenopathy.	CXR and CT (chest, neck, abdomen) FBE and ESR (anaemia and lymphopaenia), LDH, CRP and ferritin

AFP, alpha-fetoprotein; b-hCG, beta human choriogonadotropin; CRP, C-reactive protein; CT,

computed tomography; CXR, chest X-ray; ESR, erythrocyte sedimentation rate; FBE, full blood examination; Hb, haemoglobin; LDH, lactate dehydrogenase; LFT, liver function test; MRI, magnetic resonance imaging; NHL, non-Hodgkin lymphoma; NSE, neuron specific enolase; SVCS, superior vena cava syndrome; WCC, white cell count.

Table 11.8.4

Tumour lysis syndrome: management summary

Hydration and diuresis	N/2 saline: no added K^+ to start Run at 125 mL m^2 hour Maintain urine output at >100 mL m^2 hour Prevent hypovolaemia: use frusemide 1 mg kg if urine output is low
Alkalinisation	Only needed in patients with metabolic acidosis If started ONLY when urate is high: stop when hyperphosphataemia occurs If used, aim for urine pH >7.0 Cease $NaHCO_3$ if urinary pH >8.5 NB. If rasburicase is used, no need to alkalinize urine
Uric acid management	Standard-risk patients: start allopurinol 10 mg kg day High-risk patients: give rasburicase 0.2 mg kg. Usually only one dose is needed. May need a second dose, if urate is increasing above the ref range NB. Test for G6PD deficiency
Electrolytes	Monitor Ca, PO_4, K, urea and creatinine every 12 hours for 48 hours
Hyperphosphataemia and hypocalcaemia Hyperkalaemia	Aluminium hydroxide 15 mL BD Calcium gluconate IV SLOWLY and ONLY if symptomatic Oral binders if needed. If severe, insulin 0.1 unit kg with 25% dextrose 2 mL kg
Indication for dialysis	Renal failure Uncontrolled hyperphosphataemia, hypocalcaemia (symptomatic), hyperuricaemia

Hyperleukocytosis

This is variably defined as a peripheral blood white cell count more than 50–100 x 10^9 L^{-1}. The risk of complications increases when the white cell count is >100 x 10^9 L^{-1}. This occurs in 10% of children with ALL, up to 25% of patients with acute myeloid leukaemia (AML) and in virtually all children with chronic myeloid leukaemia (CML).

The interaction between cytokines released from the endothelium and leukaemic blasts increases leukostasis. This is an oncologic emergency; death can occur due to central nervous system (CNS) thrombosis and haemorrhage, respiratory failure from pulmonary leukostasis and tumour lysis syndrome preceding therapy.

With CNS leukostasis, look for headaches, blurred vision, cranial nerve and long tract signs, retinal vein distension, papilloedema and any mental state

changes. With pulmonary leukostasis, check for haemoptysis, tachypnoea, dyspnoea and hypoxia. In boys, check for priapism and in girls, clitoral engorgement.

Leukostasis is a medical emergency. Start IV fluids as for TLS immediately. Packed cells increase viscosity and should be used with extreme caution. Platelets, however, do *not* increase viscosity and should be given to decrease the risk of CNS haemorrhage. Coagulation abnormalities should also be corrected. Fluids alone will reduce the count; leukapheresis may need to be performed. However, the only way of effectively resolving hyperleukocytosis is to start systemic chemotherapy as soon as possible.

Sodium abnormalities

Hypernatraemia is caused by dehydration (vomiting due to tumour effect or chemotherapy) or diabetes insipidus (DI) caused by Langerhans cell histiocytosis involving the pituitary or suprasellar tumours.

Hyponatraemia is most commonly iatrogenic. In disease, it is caused either by cerebral salt wasting (CSW) or by SIADH. SIADH is associated with (1) some chemotherapeutic agents; (2) CNS injury/disease (surgery, tumour, or cranial irradiation) and rarely ADH-secreting tumours (lymphoma); and (3) pulmonary infection.

Useful initial tests include renal function; liver function; serum osmolality; and urine specific gravity, sodium, creatinine and osmolality.

The management of sodium abnormalities is discussed in more detail in Chapter 10.5.

Hypercalcaemia

Hypercalcaemia is uncommon in childhood cancer. It may be part of the initial presentation in leukaemia, due to a paraneoplastic syndrome (usually NHL), or secondary to bony metastases.

Hypercalcaemia treatment may result in TLS, with increasing phosphate, and subsequent nephrocalcinosis. Increased hydration (125 mL m^2 hour of normal saline) with frusemide will increase renal excretion of calcium. This approach will also help the hyperphosphataemia seen in TLS. Avoid alkalinisation, as it will increase precipitation of $CaPO_4$ in renal tubules.

Genitourinary emergencies

Genitourinary emergencies may be caused by mass effects of tumour with ureteric obstruction, rapid tumour lysis, as discussed above, or from the nephrotoxic effects of treatment such as haemorrhagic cystitis. In neutropaenic patients, infection is a significant factor. Septic shock is the most common cause of acute renal injury in paediatric cancer patients.

Urinary tract obstruction

Extrinsic compression:

1. Ureters – Wilms tumour, neuroblastoma – especially involving the coeliac axis
2. Bladder – Pelvic sarcoma, germ cell tumours, ovarian tumours, pelvic nodes with NHL
3. Urethra – Prostatic rhabdomyosarcoma.

Intrinsic obstruction:

1. Bladder rhabdomyosarcoma
2. Blood clots due to haemorrhagic cystitis from cyclophosphamide or ifosfamide.

Boys with rhabdomyosarcoma of the pelvis (prostate, base of the bladder) will present with lower abdominal pain, which can mimic appendicitis, and with urinary obstruction.

In children on treatment, obstructive uropathy has to be differentiated from urinary retention due to vincristine, varicella zoster infection of the sacral nerves with bladder dysfunction, spinal compression and opiate analgesics.

In all cases, look for decreased urine output, abdominal pain, hypertension and a rising serum creatinine. Feel for masses in the pelvis and abdomen. Arrange an urgent abdominal and pelvic ultrasound.

Consultation with the paediatric oncologist and urologist together will facilitate decompression and draining, as well as diagnosis and treatment.

In the patient on treatment, chemotherapy-induced vomiting, severe diarrhoea, poor oral intake due to nausea or mucositis, and electrolyte disorders result in intravascular depletion and consequent renal impairment.

Other renal complications

1. **Urate and $CaPO_4$ deposition and nephropathy** following tumour lysis syndrome can result in bilateral renal obstruction.
2. **Hypertension:** due to Wilms tumour (direct compression of the renal artery, renin production with secondary hyperaldosteronism) and neuroblastoma (production of adrenaline [epinephrine] and noradrenaline [norepinephrine] – which causes skin blanching and hypertension), steroid treatment, cyclosporine therapy, increased ICP from brain tumours and pain.
3. **Direct renal toxicity** from chemotherapy (cisplatin, carboplatin and methotrexate)
4. **Haemorrhagic cystitis.** Most commonly due to cyclophosphamide and ifosfamide. It can occur within 24 hours to several weeks after treatment. It needs to be differentiated from adenovirus, John Cunningham (JC) virus, BK virus and CMV infection. Treatment is with hyper-hydration, correction of thrombocytopaenia, and potentially catheterisation if clots are causing ongoing obstruction.

Neurological emergencies

Seizures

Seizures are most commonly due to focal cortical irritation and may be due to the cancer, the direct toxicity of treatment, metabolic effects of treatment or infection. Also consider usual causes of seizures in childhood (see Chapter 8.3).

Cancer treatment can induce seizures in various settings:

1. l asparaginase; especially in relation to sagittal sinus thrombosis or intracranial haemorrhage. PEG-asparaginase is long acting, and effects may be seen weeks after administration.
2. Intrathecal or high-dose intravenous chemotherapy (methotrexate or cytarabine)
3. Vincristine may cause SIADH and hyponatraemia; vinca alkaloid toxicity is more pronounced with azole antifungals.
4. Cranial radiation may induce seizures.
5. CNS infection due to chemotherapy-induced immunosuppression

6. CNS haemorrhage due to chemotherapy-induced thrombocytopaenia.

Assessment should focus on identification of fever and potential CNS infection, electrolyte abnormalities (sodium, glucose, calcium and magnesium) and bleeding risk (activated partial thromboplastin time [APTT], prothrombin time [PT], international normalised ratio [INR] and platelet count).

Imaging is dependent on what the clinical scenario entails; CT will show whether there is intracranial haemorrhage or thrombosis and will be a useful first-line modality for identifying causes of raised intracranial pressure (ICP).

Treatment

Seizures in paediatric cancer patients are usually brief and self-limited. Starting anti-convulsants is often needed while the underlying cause is being evaluated. Seizures associated with intrathecal chemotherapy seldom recur, even when the drugs are given a second time.

Acute alterations in consciousness

There are many causes of acute alterations in consciousness (AAC) in children with cancer. They can be focal or diffuse. Focal causes include tumour, infarction, haemorrhage, metastases and abscess. Diffuse causes include metabolic derangements, encephalitis, drug toxicity (cytotoxic chemotherapy and/or benzodiazepines or narcotics), postictal state, meningeal leukaemia, leukostasis/hyperleukocytosis and post-radiation somnolence syndrome.

Specific neurological conditions that are unique to children with cancer and need to be considered are as follows:

Steroids: in high doses (especially dexamethasone) can cause personality changes and psychosis. This is more common in adolescents, during induction or re-induction phases of ALL.

Acute methotrexate toxicity: can be from intrathecal or high-dose IV administration. This is a transient encephalopathy, caused by direct toxicity to the brain. There can also be focal signs that mimic stroke. In high-dose IV therapy, check the serum methotrexate (MTX) level, increase hydration and give leucovorin (folinic acid) according to the MTX nomogram.

High-dose cytarabine (Ara-C): given as an infusion is associated rarely

with cerebellar dysfunction (ataxia, speech disturbance), seizures and coma. It can occur several days after therapy has been completed. It is usually self-limiting once the drug has been ceased; but in 20% recovery is incomplete.

Nelarabine (Ara-G): used in T-cell disease. Causes weakness, cranial nerve palsies, ataxia and confusion.

Ifosfamide: can cause self-limiting dose-related neurotoxicity, usually in older children. Symptoms can range from drowsiness or confusion to unconsciousness. Occasionally, there are movement disorders (chorea and hemiballismus).

Somnolence syndrome associated with cranial irradiation: typically occurs around 6–8 weeks after whole brain irradiation and can be severe, with somnolence up to 18 hours per day, nausea, malaise, low-grade fever and weight loss. It can be confused with depression. Short-term steroids are helpful; often, recognising the syndrome and reassurance are all that are needed.

Retinoids (ATRA): given in acute promyelocytic leukaemia and neuroblastoma, can cause pseudotumor cerebri.

CNS lesions: tumour mass or cerebrovascular accident: can cause focal neurological signs and/or impaired consciousness due to invasion of brain tissue, haemorrhagic stroke, cerebral venous sinus thrombosis, cerebral oedema or mass effect.

Posterior reversible encephalopathy syndrome (PRES): a clinical syndrome characterised by headaches, altered consciousness, visual disturbances, and/or seizures. MRI may demonstrate characteristic symmetrical white matter oedema in the posterior cerebral hemispheres. PRES is associated with hypertension and cyclosporine therapy for prevention of GVHD after stem-cell transplantation. Treatment is supportive, with control of hypertension and seizures and strict fluid management.

Brain and Spinal Cord Tumours in Children

Early signs of CNS tumours in children can be subtle, and often there is a significant delay between initial presentation of symptoms and definitive diagnosis. The HeadSmart campaign in the UK (www.headsmart.org.uk) is an example of the importance of recognising symptoms and signs and how early

diagnosis can potentially make a difference to overall outcome, especially cognitive and developmental problems.

Brain tumours

Brain tumours account for around 20% of all childhood cancer and are the second most common paediatric cancer diagnosis, behind the leukaemias. The majority are infratentorial, which is reflected in the presenting symptoms and signs.

This knowledge should help the emergency doctor make a list of differential diagnoses, based on the type of tumours that occur in anatomical locations.

Infratentorial brain tumours

Posterior fossa tumours

Medulloblastoma, astrocytoma, ependymoma

Signs and symptoms relate to obstruction of CSF with raised ICP and involvement of the brainstem and cerebellum. Any child who complains of headache needs a thorough neurological examination. Increased ICP causes headache, vomiting, diplopia secondary to VIth nerve palsy, changes in behaviour, especially developmental milestone regression, and in children under 12 months, widening of sutures. The ‘setting sun’ sign of increased ICP is rare.

If vomiting is severe, think ependymoma: it is due to compression of the area postrema, which is an emetogenic centre in the dorsal medulla.

Papilloedema may be seen but is generally a late sign.

Cerebellar involvement causes truncal ataxia and vomiting from raised ICP. Lateral cerebellar involvement results in dysmetria, ataxia, nystagmus, slow sometimes ‘staccato’ speech, clumsiness, worsening handwriting and difficulty with running or hopping.

Tumours of the cerebello-pontine angle cause facial weakness, absent corneal reflex and hearing loss. Diplopia can be a false localising sign. It may be from direct compression of the abducens nerve within the pons (abducens palsy) or secondary to raised ICP.

Brainstem tumours

Astrocytoma, glioma

These may be low grade or high grade and most commonly occur in the pons and upper medulla.

The symptoms and signs are often insidious and include head tilt and squint (VIth nerve palsy), facial weakness (VIIth nerve), weakness/hemiparesis on the contralateral side, dysarthria and dysphagia, with drooling and an absent gag reflex (IXth and Xth nerve palsies).

Midline tumours

Optic and chiasmic glioma, craniopharyngioma, pineal tumours, germ cell tumours (germinoma, yolk sac tumour choriocarcinoma)

The diagnosis of midline brain tumours can be difficult. Careful history taking will show diabetes insipidus (craving water, nocturnal enuresis and weight loss). Look for precocious puberty and ask about school performance, which is usually regressing. Look for Parinaud's syndrome, which indicates pineal involvement (paralysis of upward gaze, pupils reacting to accommodation better than light, lid retraction and convergent nystagmus).

Yolk sac tumour produces alpha-foetoprotein, and choriocarcinoma produces beta-hCG. Germinomas are common and do not produce hormone markers. They can present with growth failure but also raised ICP, with diplopia and rapid progress to obtundation. Optic pathway tumours are strongly associated with NF-1; look for clinical signs of café-au-lait skin lesions, axillary freckling, and neurofibromata. In contrast, the much less common NF-2 is linked with vestibular schwannomata and deafness.

Supratentorial brain tumours

The most common signs and symptoms are seizures, long tract signs (haemiparesis, hyperreflexia and haemisensory loss) and vision problems.

Spinal cord compression

This may be insidious or acute. Symptoms may be broad, such as constipation or non-specific pain. Spinal cord compression is a medical emergency, as early diagnosis and intervention are critical to preserving neurological function. Tumour compression can disrupt the cord directly or by associated vasogenic

oedema of the cord. Around 80% of children with spinal compression will have back pain; look for weakness, sensory deficits, loss of bladder function, constipation and gait abnormalities.

If a younger child stops walking, think neuroblastoma or yolk sac tumour of the lower spine. In yolk sac tumours, children often present with frequent falls, clumsiness and a discoloured swelling at the base of the spine. Many parents assume this to be bruising from frequent falling; it is not. In older children and adolescents, think non-Hodgkin lymphoma and Ewing's sarcoma. Fifty per cent of spinal compression in childhood cancer is attributed to neuroblastoma and Ewing's sarcoma.

MRI of the spine is the best investigative test. It should be done on any patient with symptoms and/or physical findings suggestive of spinal cord compression. Plain films are of no specific benefit. Other investigations depend on the underlying diagnosis (i.e. FBE and film in leukaemia, plasma metanephrines in suspected neuroblastoma). Lumbar puncture is absolutely contraindicated.

Treatment

Early consultation with a paediatric neurosurgeon and paediatric oncologist is essential. Dexamethasone 1–2 mg kg administered IV should be given as soon as the diagnosis is considered; in the case of NHL, there may be more widespread disease and therefore a risk of tumour lysis syndrome, hence the need for early consultation.

Because spinal compression presents with radicular pain, children with a known cancer diagnosis and who present with sudden onset back pain should be emergently investigated.

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SECTION 12

Dermatology

OUTLINE

12.1

Dermatology

Roderic Phillips, Mike Starr, David Orchard, and Diana Purvis

ESSENTIALS

1 Look at the rash:

• Are there any blisters? Finding fluid-filled lesions greatly narrows the range of possible diagnoses.

• Is the rash red? Redness is from haemoglobin. Most red rashes blanch, i.e. the redness disappears with pressure. If not, the haemoglobin is outside the blood vessels giving petechiae or purpura.

• Is the rash scaly? If so, the epidermis is involved in the disease process. The epidermis may be broken to give weeping, crusting or fissures (‘eczematous rash’), or it may be intact (‘red scaly rash’).

2 Examine all the skin including the anogenital region.

3 Do not ignore a skin condition even if unrelated to the emergency visit. You may not deal with it yourself but you can organise appropriate follow-up.

Introduction

Approximately 90,000 children a year attend the emergency department (ED) at the Royal Children’s Hospital in Melbourne with 20,000 of these having some skin findings relevant to their visit. For 5000 children (6%), skin findings are a primary reason for their visit. These figures are likely to be representative of other tertiary teaching hospital EDs in Australasia. Diagnosing and managing

these children require a careful history and astute observation, particularly focusing on the appearance, site and development of their rashes.

For children who present to the ED with the primary symptom in another organ, the physician has an opportunity to observe cutaneous signs of disease, which may be relevant or may be coincidental to the child's presentation. For example, in a 5-year-old boy (Fig. 12.1.1) who presents with gastroenteritis a mild comedonal rash is noticed on his forehead; examination shows that he is tall for his age and has inappropriate penile (but not testicular) growth; thus his hyperadrenergic state can be investigated and treated years before it might otherwise have been recognised.

The discussion of diseases in this chapter is based on the features of the presenting rash and follows the algorithm given in Box 12.1.1. Diseases are grouped under their main morphology, e.g. vesicular, papular, eczematous or purpuric. Mouth, anogenital, hair and nail problems are considered separately. Neonatal skin problems are discussed in a separate chapter (see Chapter 3.3 Neonatal dermatology).

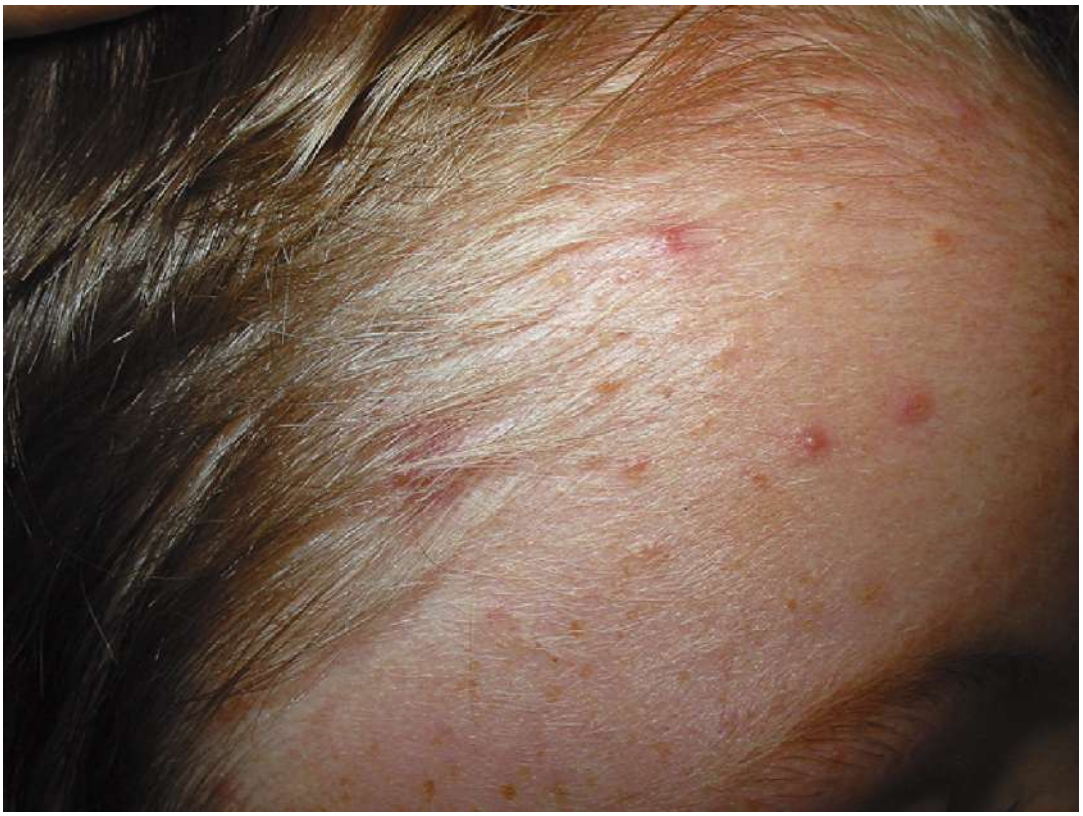

FIG. 12.1.1 Mild prepubertal acne on the forehead of a 5-year-old boy who presented with the unrelated problem of gastroenteritis. Examination

and investigation confirmed that he had a variant of congenital adrenal hyperplasia requiring treatment.

Erythroderma and skin failure

Erythroderma is one form of skin failure and refers to any condition that causes most of the skin surface to become red, often with some degree of scaling. Skin failure is rarely taught as a concept but is analogous to failure of any other organ system. Skin failure refers to the inability of the skin to adequately perform its functions, such as fluid and electrolyte balance, temperature control and protection from infection. Any child presenting with extensive areas of skin pathology or skin loss has some degree of skin failure. This may be well tolerated in otherwise healthy children but may be life threatening, especially in infants and those children with associated disorders.

Children presenting with erythroderma will usually be found to have one of the following: atopic eczema (with or without secondary staphylococcal infection), an allergic reaction, or sepsis with toxin-mediated skin involvement. Less common causes include psoriasis, pityriasis rubra pilaris, several forms of inherited ichthyosis, some other genodermatoses and internal lymphoid malignancies. In 10–20% of children with erythroderma no cause can be found.

Management

- The aim is to restore and maintain the core functions of skin. Many sensible management decisions can be made before the diagnosis is clear.
- Have a low threshold for admission to hospital.
- Restore skin-barrier function by using an ointment emollient frequently.

Box 12.1.1 Differential diagnosis of dermatological presentations

Vesicobullous eruptions

Infections

Viral: varicella, zoster, herpes simplex, hand, foot

- and mouth
    - Bacterial: impetigo, staphylococcal scalded skin syndrome
- Inflammatory
    - Erythema multiforme
    - Stevens–Johnson syndrome/toxic epidermal necrolysis
    - Acute contact dermatitis
    - Insect bite hypersensitivity reaction/spider bite
    - Burns, scalds
- Immune mediated
    - Dermatitis herpetiformis
    - Linear IgA dermatosis
    - Pemphigoid, pemphigus
- Photosensitive dermatoses
    - Primary photosensitive disorders: polymorphic light eruption, juvenile spring eruption, hydroa vacciniforme, solar urticaria
    - Inherited disorders: porphyrias
    - Systemic diseases: systemic lupus erythematosus (SLE), juvenile dermatomyositis
    - Drug induced
- Genetic
    - Epidermolysis bullosa
    - Epidermolytic ichthyosis
    - Incontinentia pigmenti

Pustular eruptions

- Infectious
    - Folliculitis
    - Boils
    - Tinea

- Scabies
- Inflammatory
  - Acne
  - Pustular psoriasis
- Drug induced
  - Acute generalised exanthematous pustulosis

Papular eruptions

Widespread

- Infectious
  - Scabies
  - Molluscum
  - Folliculitis
- Inflammatory
  - Papular urticaria
  - Papular acrodermatitis
  - Keratosis pilaris

Localised

- Infectious
  - Molluscum
  - Warts
- Tumours
  - Adnexal tumors
  - Langerhans cell histiocytosis
  - Juvenile xanthogranuloma
  - Neurofibromas
  - Angiofibromas
- Inflammatory
  - Granuloma annulare
  - Lichen planus

Red scaly (papulosquamous) eruptions

- Inflammatory
  - Eczema
  - Psoriasis
  - Seborrhoeic dermatitis
  - Contact dermatitis
- Infectious
  - Pityriasis rosea
  - Tinea corporis

Red blanching (erythematous) eruptions

- Infectious
  - Viral: erythema infectiosum, roseola infantum, enteroviruses, infectious mononucleosis, measles, rubella, unilateral thoracic exanthem
  - Bacterial: scarlet fever, toxic shock syndrome
- Inflammatory
  - Kawasaki disease
  - Urticaria
  - Serum sickness
  - SLE
  - Dermatomyositis
- Vascular
  - Capillary malformation
  - Early haemangioma (macular phase)
- Drug
  - Morbilliform drug eruption
  - Drug hypersensitivity (drug reaction eosinophilia systemic symptoms [DRESS])

Petechiae and purpura

- Infectious
  - Septicaemia: meningococcal, Staphylococcus, Streptococcus, Haemophilus, gram negative
- Inflammatory
  - Vasculitis, Henoch–Schönlein purpura
- Haematological
  - Thrombocytopaenic purpura
  - Leukaemia
  - Coagulopathy
- Trauma

Vascular lesions

- Tumours
  - Infantile haemangioma
  - Kaposiform haemangioendothelioma
  - Others
- Malformations
  - Capillary
  - Venous
  - Arterial
  - Lymphatic
  - Combination

Hyperpigmentation

Widespread

- Endocrine: Addison, pituitary Cushing, acromegaly, hyperthyroidism
- Haemochromatosis
- Renal failure
- Lipoidoses

Localised

Macular: café au lait macule, pigmentary mosaicism, nevus of Ota, mongolian spot, post-inflammatory pigmentation
Raised: congenital and acquired melanocytic nevi, acanthosis nigricans, lichen planus

Hypopigmentation

Pityriasis versicolor
Pityriasis alba
Vitiligo
Tuberous sclerosis
Pigmentary mosaicism
Post-inflammatory hypopigmentation

Anogenital rashes

Inflammatory
- Irritant napkin dermatitis
- Anogenital psoriasis/seborrhoeic dermatitis
- Lichen sclerosis
- Nutritional – zinc
- Metabolic diseases
- Langerhans cell histiocytosis

Infectious
- *Candida*
- Perianal streptococcal infection
- Molluscum
- Perianal warts
- Herpes simplex virus
- Varicella zoster virus
- Threadworms
- Syphilis

Hair and scalp

- Itchy
  - Head lice
  - Tinea capitis/kerion
  - Eczema
  - Psoriasis
  - Seborrhoeic dermatitis
- Hair loss
  - Alopecia areata
  - Traumatic alopecia, trichotillomania
  - Tinea capitis
  - Telogen effluvium
  - Chemotherapy and drug induced

Nails

- Painful
  - Acute and chronic paronychia
  - Herpetic whitlow
  - Ingrown toenail
  - Subungual haematoma
- Discoloration
  - Onychomycosis
  - Nail psoriasis
- Congenital nail disease

- Monitor temperature, as the child may readily become hypothermic or, less commonly, hyperthermic.
- Monitor fluids and electrolytes.
- Monitor cardiac output, particularly if the child has known cardiac pathology.
- Skin and/or systemic sepsis may have caused the skin failure or may be

secondary to it. Temperature control may be affected, so fever may not be present. Have a low threshold for giving systemic antibiotics, particularly to cover *Staphylococcus aureus* and streptococcal species.

- Local or systemic pain relief may be required.
- Cease all non-essential drugs.
- Identify and treat the underlying cause.
- Manage any associated conditions (e.g. diabetes, renal failure, congenital heart problems, metabolic disorders).

Vesiculobullous rashes

Vesicles are usually caused by infections (herpes simplex virus, varicella zoster virus, enterovirus, tinea, scabies or impetigo) or contact dermatitis. Consideration should be given to drug reactions, erythema multiforme and photosensitivity. Dermatitis herpetiformis is a rare cause of itchy vesicles and papules. Larger blisters occur with staphylococcal infections, tinea, erythema multiforme, Stevens–Johnson syndrome, immune-mediated blistering disorders, arthropod bites, contact dermatitis, fixed and bullous drug reactions, mastocytosis, burns or trauma.

Many vesicles and blisters are fragile and rupture easily. A vesicular disease may present with a number of shallow monomorphic erosions but no vesicles. A bullous disease may present with several larger shallow erosions with surrounding loose epithelium but no bullae. Vesicles that present for more than a couple of days become cloudy and vesiculopustular.

Varicella (chickenpox) (also see Chapter 3.3 Neonatal dermatology)

Varicella is caused by infection with varicella zoster virus (VZV). It usually occurs in children less than 15 years of age and is highly contagious. The incubation period is 2–3 weeks. The contagious period begins 2 days before the rash and continues until the lesions are crusted. Clinical disease begins with a fever and malaise, but these are usually mild. Lesions on the skin appear initially as erythematous macules, rapidly becoming small vesicles and then crusting. Over the next few days, anything from a few to hundreds of lesions may develop, initially on the face and trunk and gradually spreading onto the extremities. Superficial ulcers may be seen on all mucosal surfaces. Itch is not

always present but may be marked. Vaccinated children may develop attenuated disease with fewer lesions and without constitutional symptoms. Diagnosis is clinical but can be confirmed by serology or polymerase chain reaction (PCR) analysis of vesicular scrapings or fluid for VZV deoxyribonucleic acid (DNA). Complete resolution occurs in most children, although some scarring is common.

Children with varicella usually remain systemically well and afebrile after the first few days. If persistent fever and malaise are present, search for complications. In previously well children, the most common complications are:

- bacterial skin infection with group A streptococci or *Staphylococcus aureus*. Look for bullous, indurated, spreading or cellulitic lesions. Treat with oral antibiotics.
- central nervous system (CNS) dysfunction. Encephalitis, meningitis, transverse myelitis, Guillain–Barré syndrome or Reye's syndrome (sudden onset of vomiting, coma and liver abnormalities) may occur. Cerebrospinal fluid (CSF) analysis, including PCR for VZV DNA may be useful in suspected encephalitis.
- pneumonia. Cough and respiratory distress begin a couple of days after onset of the rash.
- cerebellar ataxia, which may appear as the rash is clearing and resolve within weeks.

In neonates, immunocompromised children and adolescents, varicella can be more severe and occasionally fatal.

Management

- Treatment of otherwise well children with varicella is supportive with bland emollients and oral antihistamine if necessary. If the child is unwell, look for secondary complications.
- Avoid aspirin or other salicylates because of the association with Reye's syndrome.
- Treat secondary skin infection with cephalexin 30 mg kg^{-1} (max 500 mg) three times daily.
- Immunocompromised children should be treated with intravenous (IV) aciclovir 20 mg kg^{-1} per dose (2–12 weeks), 500 mg m^{-2} per dose (12 weeks–12 years), 10 mg kg^{-1} per dose (adult) every 8 hours given over

1 hour.

- Premature neonates (less than 1 month old) with varicella should be treated with IV aciclovir.
- Term neonates should be admitted and treated with aciclovir if they have severe disease.
- Give zoster immune globulin to at-risk contacts (immunocompromised children, those on prednisolone 2 mg kg^{-1} day^{-1} or more, newborns of mothers who have varicella any time from 5 days before until 2 days after delivery). Give within 4 days of exposure (6 mL for adults, 4 mL for children 6–12 years of age, 2 mL for children up to 5 years of age).
- Exclude from school until all lesions are crusted.
- Offer varicella-zoster vaccine to non-immune contacts; the vaccine has been shown to prevent or attenuate infection if given within 5 days of exposure.

Zoster

Zoster is uncommon in childhood but can occur at any age, even in the neonatal period. It occurs in children who have previously had primary varicella infection. Zoster in a young child is usually associated with primary varicella having occurred in infancy. The primary infection may have been mild and may even have gone unnoticed because of maternally acquired IgG.

Radicular pain may be the first symptom of zoster, but pain is less common than in adults and may not be present. Vesicles on an erythematous background appear in one or more groups aligned in a dermatomal distribution, frequently with a striking midline cut-off. The affected area can be an isolated group of a few vesicles suggestive of herpes simplex virus infection. Alternatively, there may be extensive involvement of one or more dermatomes. Lesions continue to appear for a few days and then resolve over 2 weeks, generally without sequelae. Post-herpetic neuralgia is very uncommon after childhood zoster.

Complications include generalised dissemination during the first week of the rash, sometimes with pulmonary or brain involvement. This can be seen in both normal and immunocompromised children. Zoster on the head is occasionally accompanied by an aseptic meningitis, which resolves fully without treatment. Zoster lesions on the tip of the nose imply involvement of the nasociliary branch of the ophthalmic nerve and may accompany ocular involvement including keratitis and conjunctivitis. Facial nerve palsy and ear involvement can occur

(Ramsay Hunt syndrome).

Immunocompromised children are much more likely to develop zoster. In such cases, zoster may be severe, extensive and prolonged. Significant dermatomal scarring can result.

Management

- If the diagnosis is unclear, laboratory confirmation is worthwhile. PCR requires collection of a dry swab rolled over a vesicle. Care needs to be taken to collect sufficient cells for analysis. Yield may be improved by collecting epithelial cells from the base of a vesicle for viral culture.
- In an otherwise well child, the occurrence of typical zoster does not require investigation for underlying immunodeficiency states. However, unusually severe or extensive zoster should raise this possibility.
- If a child has underlying chronic illness or immunodeficiency, give IV aciclovir or oral famciclovir or valaciclovir. Oral antiviral agents can be considered in older adolescents but are not generally indicated in an otherwise well child as the risk of significant symptoms or post-herpetic neuralgia is very small.

Hand, foot and mouth disease

Hand, foot and mouth disease is one presentation of enteroviral infection, usually Coxsackie A16 virus. There may be a mild prodrome of fever, malaise and sore throat. Discrete vesicles about 6 mm in diameter on an erythematous base appear first in the mouth, sparing the lips and usually sparing the gingiva. Vesiculopustules (3–7 mm) on an erythematous base appear on the palms, soles and around the fingers and toes 1–2 days later. Lesions are also often found on the buttocks. They are often greyish and oval rather than circular in shape. In some cases, the mouth, hands or feet may not be involved. Resolution occurs within a week. The incubation period is about 5 days, and epidemics are common. Virus is excreted in the faeces for weeks, and exclusion is not recommended. Supportive care leads to complete recovery.

Herpes simplex infection

Herpes simplex virus (HSV) infection in childhood is common. Intrauterine,

intrapartum and neonatal infections are considered elsewhere. Primary herpes infection after the neonatal period may be either primary herpetic gingivostomatitis or primary cutaneous herpes simplex. In children with impaired immune function, HSV infection can present as indolent, slowly growing, irregular ulcers. Most HSV infections in children are caused by HSV-1, but HSV-2 may also cause gingival and cutaneous infection.

Primary cutaneous herpes simplex can present at any age. There may be a history of a family member with a recently activated herpes simplex lesion on the lip (‘cold sore’). However, the virus is ubiquitous, and transmission more often occurs from an unknown source. At least 50% of the population has HSV-1 antibodies, and many will have never had an obvious infection. Painful grouped vesicles on an erythematous base can appear at any site. Multiple lesions rupture, crust and coalesce into larger erosions with scalloped edges. There may be associated fever, malaise and lymphadenopathy.

Management

Management consists of the following:

- Supportive care
- Bathing of crusts
- Analgesia
- Aciclovir is not routinely indicated unless risk factors for complications exist (e.g. underlying disease, immunosuppression).

Recurrent cutaneous herpes simplex can occur weeks or months after the primary episode. Recurrences usually get milder and less common with time, but exacerbations can occur throughout childhood and later life (Fig. 12.1.2). Recurrences can lead to significant time off school. Antiviral prophylaxis may reduce the frequency and severity of recurrences and should be used if recurrences are frequent or debilitating.

HSV infection can involve the fingers, especially the thumbs and index fingers (herpetic whitlow). At these sites, the thick skin does not readily rupture, and children present with painful, coalesced pustular collections, often misinterpreted as bacterial abscesses. Any history of ‘abscesses’ occurring at the same site on a finger on more than one occasion is suggestive of herpes simplex. Management includes taking a swab for PCR and viral culture to confirm the diagnosis, and analgesia and supportive care.

Eczema herpeticum

Herpes simplex infection in children with eczema is quite common. It occurs in children with any severity of eczema including children with mild eczema under excellent control. Many cases are misdiagnosed either as an exacerbation of the eczema or as bacterial infection (Fig. 12.1.3). Grouped vesicles may be prominent, but more often vesicles are rudimentary or absent, and the infection may present as a group of shallow 2–4 mm monomorphic erosions on an inflamed base. In more severe cases, evolution is rapid, and large crops of vesicles can arise daily. Ulcers may coalesce into larger erosions with scalloped edges. The infected area may not be painful or itchy.

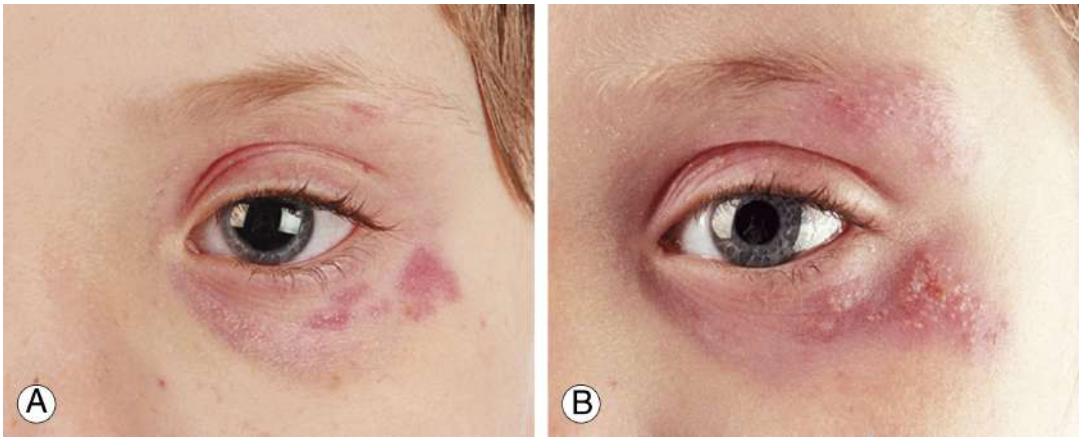

FIG. 12.1.2 (A) This erythematous, slightly vesicular rash around the eye of a 4-year-old boy had appeared over 12 hours. Because his other eye was blind, he was treated with oral aciclovir. Herpes simplex virus was confirmed by polymerase chain reaction. (B) One day later.

In an atopic child, a high index of suspicion is needed about any patch of skin where small erosions or crusts are present and not responding to standard eczema therapy with moisturisers and topical corticosteroid (CS) preparations. One reason for the under diagnosis of this condition is that even without treatment, resolution usually occurs in 1–4 weeks. However, dissemination may occur, leading to multifocal and extensive erosions, malaise and secondary bacterial infection. In the past, disseminated eczema herpeticum had a significant mortality, but this has declined with the recognition and aggressive treatment of secondary bacterial infections.

After complete resolution, recurrences may occur at the same or different sites, sometimes as often as every few weeks. Recurrences can be widespread

and severe but usually decrease in severity over 1 to 2 years.

Management

- Clinical differentiation between herpetic and bacterial infection may be difficult; investigation and empiric treatment for both may be necessary.
- Swab vesicles for HSV PCR and viral culture.
- Local stable disease in an otherwise well child requires regular observation but does not need antiviral therapy.
- Milder cases demonstrating progression or facial involvement can be managed with oral aciclovir.
- Admission to hospital and treatment with IV aciclovir (20 mg kg^{-1} per dose [2–12 weeks], 500 mg m^{-2} per dose [12 weeks–12 years], 10 mg kg^{-1} per dose [adult] every 8 hours) should be considered for children with fever, multiple sites of cutaneous herpes infection, widespread eczema, eye involvement, immunosuppression or age less than 6 months.

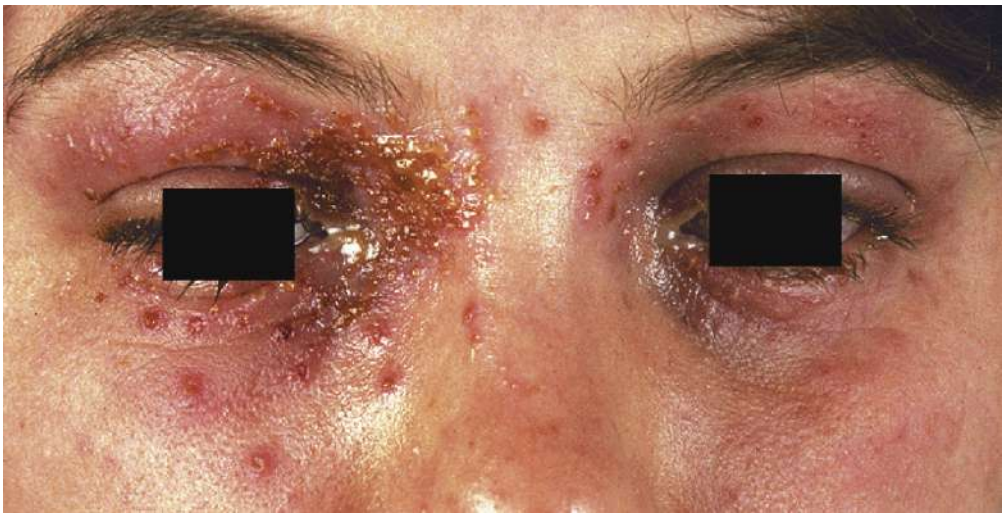

FIG. 12.1.3 Eczema herpeticum. Typical monomorphic vesicles can be seen away from the central weeping area.

- Secondary bacterial infection is common. Swabs for microscopy and culture are usually unhelpful. If there is any suspicion of bacterial infection, add IV flucloxacillin 50 mg kg^{-1} (max 2 g) every 6 hours or oral cephalexin 30 mg kg^{-1} (max 500 mg) three times daily. Soak to remove excessive crusts.

- Monitor for eye involvement, particularly scleral redness. If present, this should be managed with topical or systemic aciclovir, or both, and urgent review by an ophthalmologist.
- The underlying eczema can be treated with moisturiser or wet dressings until the herpetic scabs have been removed, and then it is important to restart topical CS treatment.
- Antiviral prophylaxis may reduce the frequency and severity of recurrences and is often warranted in older children with recurrent eczema herpeticum.

Impetigo (School sores)

This is caused by *Staphylococcus aureus* or group A *Streptococcus* or both. Non-bullous impetigo presents initially as small erythematous vesicles that rapidly rupture to form yellow-crusted lesions, commonly on the face. Bullous impetigo is due to *Staphylococcus aureus* and presents as flaccid blisters on normal skin. Lesions are rounded and well demarcated and may be single, grouped or widespread. Their onset and spread may be rapid or occur over days. New blisters appear, and existing blisters rupture to give shallow moist erosions that can be many centimetres in size. There is often loose epithelium and/or brown crusting peripherally and some degree of central healing (Fig. 12.1.4). In more chronic cases, lesions may appear annular.

Impetigo is often secondary to itchy conditions such as scabies infection (especially hand impetigo), atopic eczema and head lice.

Sequelae include post-streptococcal glomerulonephritis, which may occur in the ensuing 2 months, and chronic streptococcal impetigo also has been identified as a precursor of rheumatic heart disease in communities where medical conditions are poor and skin hygiene is suboptimal.[1]

Management

- Swab for microscopy, and culture if the diagnosis is unclear or in widespread or complicated disease.
- Bathe off crusts.
- Apply topical mupirocin 2% ointment every 8 hours if very localised, or oral cephalexin 30 mg kg^{-1} (maximum 500 mg) three times daily if extensive. Note that resistance to mupirocin is becoming more

widespread, and use should be restricted.

- Isolate the child from other children or from sick adults unless all lesions are covered or antibiotic treatment has commenced.
- Treat any underlying condition such as scabies or eczema.
- Blisters or erosions often continue to occur for a couple of days after antibiotic therapy. If new blisters continue to form after this time, they may be caused by resistant *Staphylococcus aureus* infection, or the child may have an immune-mediated blistering disorder rather than impetigo.
- Children with chronic impetigo should be examined for any signs of heart disease. Families should be aware that rheumatic heart disease may be delayed by some years in this setting. Older siblings are likely to have had chronic impetigo in the past and therefore to also be at risk of heart disease and should be examined.
- In any child with chronic impetigo where ongoing care may be suboptimal, review with urinalysis to monitor for glomerulonephritis is warranted. In other cases, families should be aware of the possibility of renal involvement.

Staphylococcal scalded skin syndrome

This is usually seen in younger children or children with renal impairment. It is mediated by an epidermolytic toxin released from an often insignificant staphylococcal focus (e.g. eyes, nose or skin). Fever and tender erythematous skin are early features. Exudation and crusting develop, especially around the mouth. Wrinkling, flaccid bullae and exfoliation of the skin are seen and can be extensive. Nikolsky sign is present (‘normal’ skin separates if rubbed). Blisters are very superficial and heal without scarring.

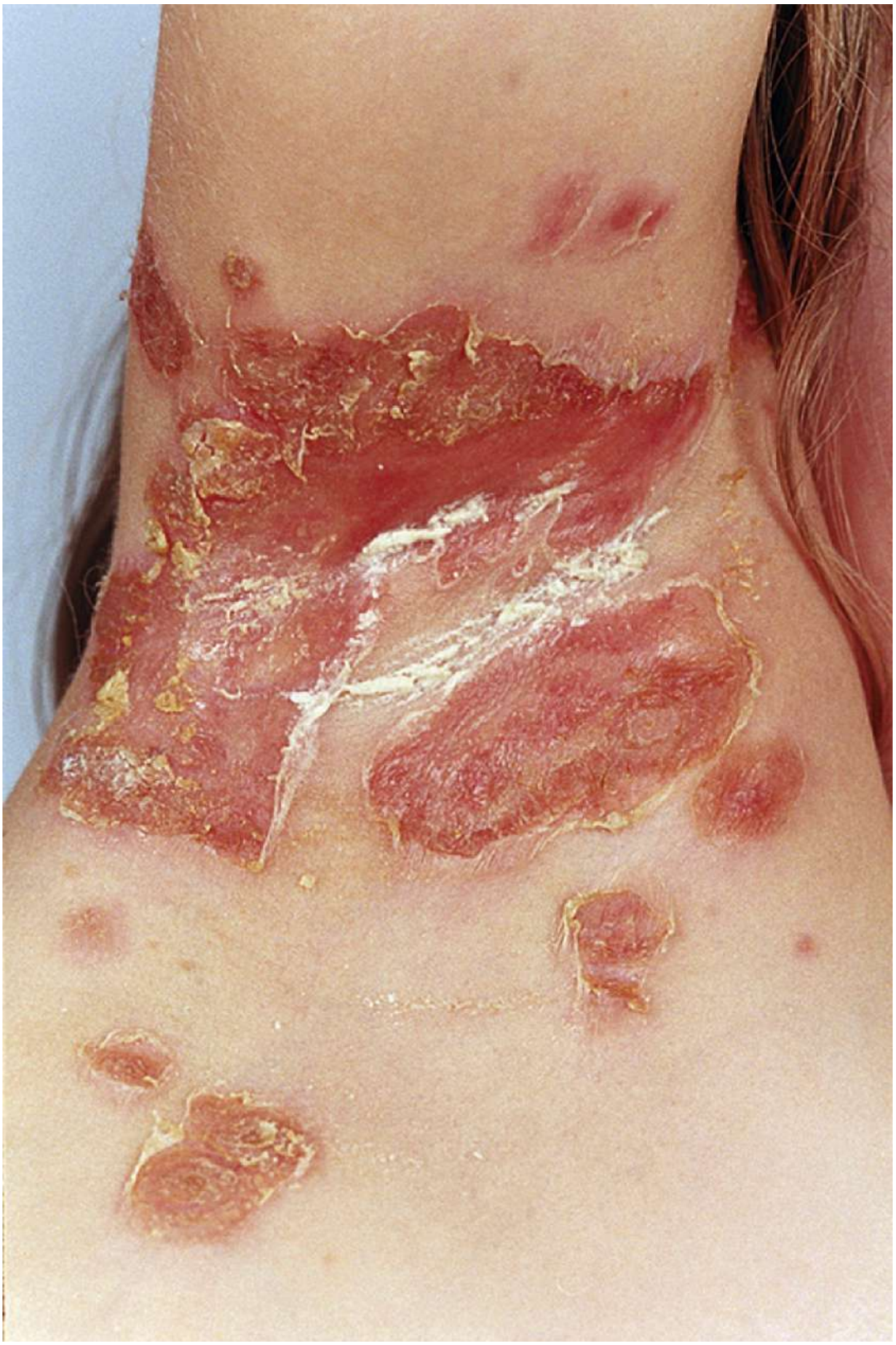

FIG. 12.1.4 Bullous impetigo in a 7-year-old girl.

Management

- Admit to hospital. Monitor temperature, fluids and electrolytes if large areas are involved. Increase oral (or IV) fluids to increase toxin excretion. Give analgesia.
- Consider alternative diagnoses including Stevens–Johnson syndrome and toxic epidermal necrolysis. If the diagnosis is unclear, a skin biopsy will confirm the diagnosis by showing the split is in the granular layer in the upper epidermis.
- Flucloxacillin 50 mg kg^{-1} (maximum 2 g) IV every 6 hours if there is evidence of sepsis or systemic involvement.
- Look for a focus of infection. Drain any foci of pus if present.
- Handle skin carefully, and use an emollient ointment.

Erythema multiforme

This is a specific hypersensitivity syndrome that occurs at any age, often preceded by facial HSV infection. It is over-diagnosed in EDs. Most children diagnosed with erythema multiforme actually have urticaria, often with large lesions with annular or polycyclic borders. In erythema multiforme, the primary lesions are red papules, usually symmetrical and involving the forearms, palms, feet, face, neck and trunk. They can be found anywhere. There may be few or many lesions. At least some of the papules will form classical target lesions – these have an inner zone of epidermal injury (purpura, necrosis or vesicle), an outer zone of erythema and sometimes a middle zone of pale oedema (Fig. 12.1.5). These papules and target lesions are not migratory. The involvement of mucous membranes is common but, unlike Stevens–Johnson syndrome, is limited to isolated patches. Most cases are caused by HSV, often without prior symptoms. Drugs are an uncommon cause. Erythema multiforme does not evolve into Stevens–Johnson syndrome; they are different conditions.

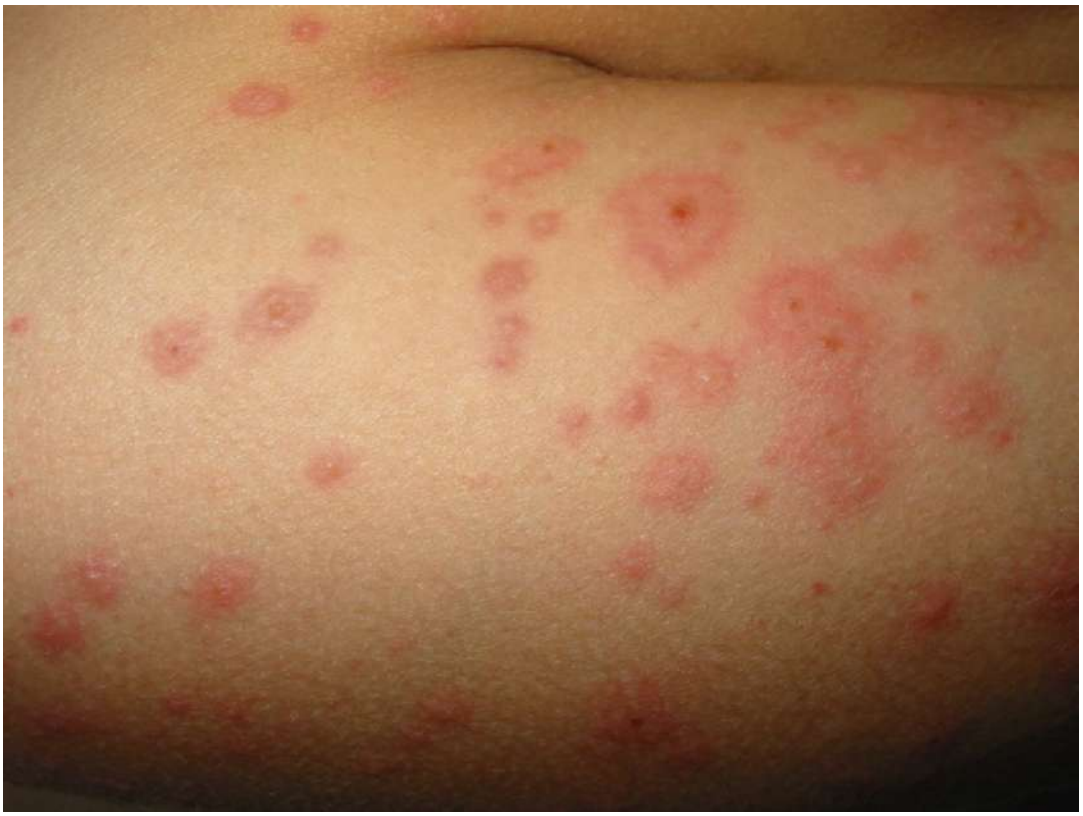

FIG. 12.1.5 Erythema multiforme target lesions.

Management

- Fluid maintenance
- Analgesia if required
- Apply emollient ointment to the lips, if needed
- If the condition is recurrent, it is highly likely to be related to HSV. Prophylactic aciclovir prevents recurrences and should be considered if recurrences are frequent, severe and affecting the quality of life
- Prednisolone alleviates symptoms but probably prolongs the duration of the condition and may cause recurrence of HSV infection.

Stevens–Johnson syndrome/toxic epidermal necrolysis

Stevens–Johnson syndrome and toxic epidermal necrolysis are variants of one condition. Most cases are thought to be triggered by medications, especially

antibiotics (sulfonamides, penicillins), anticonvulsants (lamotrigine) and non-steroidal anti-inflammatory drugs. *Mycoplasma* and other infections may be the cause in some children. Fever, myalgia, arthralgia, headache and other organ involvement are usually present as a prodromal illness for several days. The rash evolves suddenly, characterised by widespread blisters on an erythematous or purpuric macular background, often with extensive mucous membrane haemorrhagic crusting. Lesions are usually on the face and trunk and generalised rather than acral, and typical target lesions are not seen. There may be tender erythematous areas with a positive Nikolsky sign (‘normal’ skin separates if rubbed). Mucous membrane involvement can be extensive, severe and painful. Conjunctivitis, corneal ulceration and blindness can occur. Anogenital lesions can lead to urinary retention.

Management

- See general advice under ‘skin failure’.
- Admit to hospital. A multidisciplinary approach including dermatology, paediatrics, ophthalmology, surgery and paediatric intensive care is often required. Good nursing care is paramount. In severe cases, nurse in a specialised burns unit.
- Apply emollient ointment to the skin, lips and anogenital areas – this may be required many times a day.
- Regular eye examination with specialist review is required. Topical steroid eye drops may be needed.
- Give IV immunoglobulin 2 g kg^{-1}. Commence cyclosporin (5–6 mg kg^{-1} day^{-1} orally for a few days, then taper to 3–5 mg kg^{-1} day^{-1} for 2–3 weeks). Data about the efficacy of any treatment regimens are weak. Stevens–Johnson syndrome has a significant mortality and has a high rate of permanent visual scarring, including blindness.
- Maintain good nutrition, with nasogastric feeding if needed.
- Intubation may be needed if the child is unable to protect the airway because of severe oropharyngeal involvement.

Stevens–Johnson syndrome is *not* severe erythema multiforme. They are distinct conditions. Permanent sequelae are rarely seen in severe erythema multiforme, and concurrent drug use is unlikely to be the cause. Skin lesion distribution and morphology are the best discriminating factors. Mucous

membrane involvement can be seen in both conditions but is confluent in Stevens–Johnson syndrome.

Dermatitis herpetiformis

Dermatitis herpetiformis is an uncommon autoimmune blistering disease that may present at any age as itchy papules or vesicles. Rapid excoriation means that intact vesicles are rarely seen. Lesions occur on extensor surfaces of the limbs, buttocks, trunk and scalp. Most patients have gluten enteropathy (coeliac disease) and may have abdominal discomfort, diarrhoea or anaemia. Some degree of villous atrophy will be seen on small bowel biopsy in most cases, but coeliac disease is often asymptomatic at the time of presentation of dermatitis herpetiformis. Growth retardation has been reported with dermatitis herpetiformis, probably secondary to intestinal involvement.

Recurrence some years after successful treatment has been reported.

Management

- Perilesional skin should be biopsied for immunofluorescence to demonstrate the diagnostic granular IgA deposition.
- Investigate for coeliac disease with total IgA, anti-tissue transglutaminase IgA and anti-deamidated gliadin IgA. Anti-transglutaminase antibodies are the most specific for dermatitis herpetiformis. A small bowel biopsy is required but may be equivocal early in the disease course.
- Anaemia, usually megaloblastic secondary to malabsorption, may be found.

Skin lesions (but not intestinal symptoms) respond well to dapsone, but haemolysis and other side effects can limit its use. A gluten-free diet normalises gut function and, in most children, leads to eventual clearing of skin lesions.

Other immune-mediated blistering disorders

As with dermatitis herpetiformis, the many diseases in this group are characterised by circulating autoantibodies directed against one or other of the structural proteins that give the epidermis its integrity. All are uncommon in

childhood, linear IgA dermatosis (chronic bullous disease of childhood) being the most common. An immune-mediated blistering disorder should be considered in an otherwise well child if vesicles, blisters or crusted erosions continue to appear for more than a couple of weeks.

If there is a split below the basement membrane, lesions may be tense, long-lasting bullae, as seen in dermatitis herpetiformis, linear IgA dermatosis, systemic lupus erythematosus (SLE) and pemphigoid. If the split is higher in the epidermis, lesions are flaccid, short-lived blisters that rapidly give crusted erosions with loose epithelium around the edges. These are seen in pemphigus variants. Lesions can mimic local or generalised impetigo. Mucous membranes may be involved. Despite the widespread and often dramatic appearance of the rash, the child is usually well. Accurate diagnosis always requires histology and immunofluorescence in addition to clinical findings. Management requires immunosuppressive therapy and long-term follow-up.

Sunburn and photosensitivity

Excessive solar radiation to the skin causes erythema and tenderness commencing at least half an hour after the beginning of the exposure. Tenderness worsens for a day and resolves in 4 days. In more severe cases, oedema and blistering may be widespread. Healing is accompanied by desquamation and intense itch. Sunburn occurs more rapidly when the sun is overhead (summer, closer to the equator, 11 a.m. to 3 p.m.), and when the skin is less pigmented, but can occur with any skin type. Children with sensitive skin can burn significantly within 15 minutes of midday exposure in summer.

Any child presenting to the ED with sunburn or with a rash in sun-exposed areas may have an underlying photosensitivity disorder. These can be considered in four groups:

1. Primary photosensitivity disorders
2. Inherited disorders including porphyrias
3. Diseases with a photosensitive component (SLE, dermatomyositis)
4. Exogenous photosensitivity from drugs or plants.

A high index of suspicion is needed to diagnose photosensitivity. These diseases are common, but diagnostic traps abound. For example, sun-induced rashes may develop in spring with the first sun exposure of untanned skin after

winter but not later in summer. The rash may develop on areas usually covered in winter such as the neck and arms and not on the face. Sun-induced rashes may require a few minutes or several days of sun exposure and may commence days after the exposure. In erythropoietic protoporphyria, sun exposure induces pain without skin changes initially. Viral exanthems may occur exclusively or mainly in areas of sun-exposed skin. Children with solar urticaria develop urticaria on sun-exposed areas.

Management

- Assess for associated causes (below) of increased sun sensitivity. Many children presenting to the ED with ‘sunburn’ have an associated photosensitive trigger. Any child who experiences symptoms within half an hour of sun exposure should be assumed to have an underlying condition.
- Assess the family situation, especially with younger children. Significant sunburn in a young child can occur in the setting of suboptimal child care, although usually it is due to an oversight by otherwise caring parents.
- Within the first few hours of exposure, before blistering has developed, application of topical potent CS can considerably reduce the severity and duration of symptoms.
- Cool compresses and wet dressings will alleviate symptoms. Topical anaesthetics should not be used. Admission and oral analgesia may be required in severe cases. Maintain adequate fluid intake.
- Educate about prevention.

Prevention

Prevention of sunburn is important. Episodes of sunburn are linked to later development of naevi and melanoma.

Parents need to balance the psychological and physical benefits of activities associated with sun exposure (including increased fitness, increased independence, healthier bones and decreased obesity) with the increased risk of skin cancers of all types. In summer, children should minimise sun exposure during the middle 4 hours of the day, wear broad-brimmed hats and long-sleeved shirts in the sun, and use a sunscreen with SPF 30 or greater on exposed parts.

Sunburnt infants regularly present to the ED, partly because of widespread

advice that sunscreens are not recommended below the age of 6 months. This recommendation is not based on any evidence of problems in infants using sunscreen. It is based on the premise that it is usually easy to protect young babies who can't crawl by keeping them out of the sun and appropriately dressed. Thus, if a young baby is going to be in the sun, sunscreen should be used.

Primary photosensitivity disorders

Polymorphous light eruption

This presents in spring or early summer as skin-coloured or erythematous itchy papules or vesicles on the face, ears, neck or arms. It is recurrent. Recurrences usually occur at the same sites in that child.

Juvenile spring eruption

This is a specific variant of polymorphous light eruption (PMLE) and is common in 4- to 12-year-old boys as a recurrent blistering of the ears each spring.

Hydroa vacciniforme

This is rare and can begin at any age in childhood as a sun-induced vesicular eruption on the cheeks during spring and summer. The cheeks, ears, nose, dorsum of the hands and rarely the eyes can be involved. Recurrences occur for many years.

Diagnosis can be difficult and may require formal evaluation in a photobiology unit including assessment of the response to different wavelengths of light. Management requires sun avoidance and often both ultraviolet (UV)A and UVB sun protection.

Solar urticaria

This presents as urticaria within minutes or hours of exposure and settles within 1 day. Sharp margins are seen at the edges of clothing. Skin chronically exposed to the sun, such as face and hands, is often spared.

Porphyrias and other inherited disorders with photosensitivity

The porphyrias are a group of inherited enzymatic defects in haem synthesis leading to increased levels of porphyrins, some of which cause photosensitivity.

Erythropoietic protoporphyria

This is the most common childhood porphyria. Infants and young children present after brief sun exposure with acute discomfort, burning sensations, itching, erythema, oedema, urticaria and occasionally vesicles, particularly on the face and dorsum of the hands. Episodes are recurrent, and with time affected skin appears prematurely aged. Erythrocyte protoporphyrin levels are elevated.

Congenital erythropoietic porphyria

This is rare and presents in early childhood with extreme photosensitivity leading to painful blisters filled with red fluid on the face and dorsum of the hands. Progressive scarring can be severe.

Familial porphyria cutanea tarda

This presents in childhood with chronic blistering on hands, arms and face leading to poorly healing ulcers, atrophic scarring and mottled hypo- and hyperpigmentation.

If a porphyria is suspected, measure blood, urine and faecal levels of porphyrins. Management requires intensive avoidance of solar radiation including UVA and UVB. β-Carotene is useful in erythropoietic protoporphyria.

Many genodermatoses including xeroderma pigmentosum, Cockayne syndrome, Rothmund-Thomson syndrome and trichothiodystrophies are associated with photosensitivity and increased photo-damage to skin but do not usually present with blistering.

Photosensitivity and bullous reactions to drugs

Blisters may be the presenting feature of a number of different types of drug reactions. A high index of suspicion is required to diagnose these conditions. Families must be asked about all ingested and topical products including herbal, recreational and prescription drugs and unusual food patterns (e.g. daily cups of celery juice). Families will occasionally not remember the causative drug and deny taking anything, only to recall the vital information days later.

Many medicines can cause increased sun sensitivity leading to erythema, oedema and blistering on sun-exposed areas. Doxycycline (particularly at doses

higher than 100 mg day^{-1}), tetracyclines, griseofulvin, isotretinoin, non-steroidal anti-inflammatory drugs, sulfonamides, fluoroquinolones and diuretics are typical causes in children. In one series of children receiving naproxen, 12% developed photosensitivity reactions on the face. Other causes include tars, perfumes, cosmetics, sunscreens, artificial sweeteners and many dyes. As with other photosensitivity syndromes, the rash can occur rapidly or days after the sun exposure and is most prominent on sun-exposed areas. The photosensitivity can persist for up to 3 months after withdrawal of the medication. Prominent hyperpigmentation may persist for months after resolution.

Fixed drug eruptions in children are usually caused by paracetamol, non-steroidal anti-inflammatories or sulfonamides. They are quite common and present as single or multiple, usually circular red patches that may blister. With subsequent exposures, eruptions recur at the same site and sometimes at other sites. Post-inflammatory hyperpigmentation is prominent and may be the only sign of the drug reaction.

Many plants and herbal products contain chemicals that can induce drug reactions including fixed drug reactions, bullous drug reactions and photosensitivity reactions.

Management

- Recognise and stop the causative drug.
- Institute strict sun protection measures. Photosensitivity can persist for 3 months after cessation of the drug.
- If the patient is taking multiple drugs, the timing of the eruption and the frequency of reactions with each of the medications may suggest the causative drug.
- If continued drug ingestion is necessary, reducing the dosage may prevent the reaction.

Photosensitivity reactions to plants

Many plants contain furocoumarins, which are naturally occurring psoralens. UVA light induces covalent bonding of psoralen into DNA, leading to cell death. Typically, a child is playing outside in spring or summer and comes into close contact with a psoralen-containing species (e.g. species of celery, parsley, parsnip, fig, hogweed, limes and other citrus fruits). Several hours later,

erythema, oedema and occasionally blistering develop. While contact dermatitis from plants can also present with streaky, often linear vesicular eruptions on areas of contact after exposure, photosensitivity reactions are more often painful, are usually sharply limited to sun-exposed areas, and typically heal to leave striking patterns of hyperpigmentation that can persist for months or years.

Management

- Sun protection measures including a UVA sunscreen will prevent phytophotodermatitis.
- Offending plants can be removed.
- Treat as for sunburn.

Contact dermatitis – plants

In children, contact dermatitis reactions from agents other than plants usually do not blister, and these conditions are discussed under eczematous rashes (p. 339).

Many plants, including *Rhus* and *Grevillea* species, can cause an allergic contact dermatitis. Some species only cause reactions at specific times of the year, and contact with the plant at other times does not cause a rash. Erythema, oedema and vesiculation develop at sites of contact 1–3 days after exposure, often in linear distribution. Lesions may persist for 3 weeks. Periorbital erythema and vesiculation are often misdiagnosed in the ED as cellulitis needing antibiotics. Clues to the correct diagnosis are that pruritus is the main symptom; the degree of pain and tenderness is much less than would be expected for cellulitis; systemic features, such as fever, are absent; the outline of the rash often has irregular patterns corresponding to the contact areas and a careful examination will often reveal other lesions on sensitive parts of the body. In particular, genital skin may develop lesions from secondary spread of the allergen via the fingers. Patients with allergic contact dermatitis may be extremely uncomfortable. Treatment often requires a few days of oral prednisolone, potent topical CSs, cool compresses and supportive care. Unlike plant-induced photosensitivity, contact dermatitis from plants does not induce long-term hyperpigmentation.

Irritant contact dermatitis to components of stinging nettles, chilli peppers, mustard, horseradish and other plant products can lead to irritation, stinging and occasionally blisters. Burning, oedema and blistering in the oral cavity can occur

in small children after chewing irritant plants.

Contact dermatitis – id reactions

Repeated exposure to an allergen can result in a papulovesicular eruption at sites distant from the area of contact. This is thought to be a result of systematised contact sensitisation. The id reaction may be local or generalised. Examples include non-infective blisters erupting on the hands of a child with chronic tinea infection of the feet and a severe generalised itchy vesicular rash on an adolescent using topical neomycin ointment.

Isolated blisters

For a child who presents with a single blister or a few blisters as an isolated finding, consider:

- mastocytoma. Usually in an infant with a history of recurrent blistering or crusting at the site of a brownish macule, often misdiagnosed as recurrent localised impetigo. There may be one brownish lesion or as many as hundreds.
- insect bite. There may be a few lesions in one area. Non-blistered papules may have a tiny central red bite punctum.
- irritant contact dermatitis
- spider bite. These can grow over days to become a non-tender blister with a diameter of many centimetres. They are uncommon. Regular dressings may be required. Debridement is rarely necessary. White-tailed spiders and huntsman spiders do not cause chronic necrotic ulcers.
- scalds or burns from cigarettes or other hot objects. Look for characteristic patterns in the burn, as well as other factors on the history or examination suggestive of non-accidental injury or suboptimal caring practices.
- fixed drug eruptions, e.g. from tetracyclines, sulfonamides, non-steroidal anti-inflammatory drugs or paracetamol
- friction. If minor friction appears to cause more blistering than expected on hands and/or feet in children, consider the possibility of a mild inherited blistering disorder (e.g. mild epidermolysis bullosa simplex).

There may be a positive family history of childhood blistering that settled in later life.
- artefactual lesions caused by either the child or a carer
- Bullous sweet syndrome or pyoderma gangrenosum.

Chronic erosions or ulcers

Several primary skin conditions can cause chronic erosions or ulcers in children, including itchy conditions such as scabies and papular urticaria. Also consider:

- immunodeficiencies. Recurrent boils can be seen in chronic granulomatous disease and hyper IgE syndrome. Poor wound healing is a feature of leucocyte adhesion defects.
- skin fragility syndromes. In junctional and dystrophic forms of epidermolysis bullosa, chronic ulcers related to minimal skin trauma may be seen in association with failure to thrive, anaemia and gastrointestinal tract involvement.
- porphyria. Several enzyme defects in haem metabolism are associated with chronic erosive lesions, photosensitivity and hyperpigmentation. Episodes of acute pain or neurological dysfunction are rarely seen in childhood porphyrias.
- artefactual lesions caused by either the child or a carer
- pyoderma gangrenosum
- *Mycobacterium ulcerans* infection.

Neonatal vesicles (see Chapter 3.3 Neonatal dermatology)

Pustular rashes

Consider acne, folliculitis, drug reactions, pustular psoriasis, scabies, perioral dermatitis, tinea and localised bacterial infection. All vesicular rashes can become pustular if the vesicles persist for more than a couple of days. Vesicles on areas of thick skin can be white and look like pustules, e.g. on the toes in hand, foot and mouth disease. If in doubt, prick one lesion to reveal the clear fluid within.

Acne

Acne mainly affects the forehead and face but can involve other sebaceous gland areas (neck, shoulders and upper trunk). Early lesions include blackheads, whiteheads and papules. In more severe cases there may be pustules or inflammatory cysts that can lead to permanent scarring. Acne is common in adolescence. It is treatable, and no person with acne should be told it is an inevitable part of adolescence. Several topical and oral acne therapies are available and should be used to treat the disease. Under-treated acne is a major medical cause of significant morbidity in adolescents and has a recognised mortality as a factor in teenage suicide. It can also lead to permanent scarring in 10% of untreated cases (Fig. 12.1.6). Significant acne in an adolescent who is attending the ED for another reason should not be ignored. The adolescent should be offered information, initial treatment and referral for ongoing management.

Management

- Most cases are mild and either require no intervention or can be managed with topical treatments. A first-line topical treatment is benzoyl peroxide 2.5% applied once or twice a day. Warn the adolescent that improvement with topical therapy occurs steadily over 1–4 months (not in a few days). Other topical agents include topical antibiotics for inflammatory lesions (clindamycin, erythromycin or tetracycline), topical retinoids (isotretinoin, adapalene) or azelaic acid. These can be used singly or in combination with combination preparations becoming more available. All of these topical agents have the potential to cause irritation.

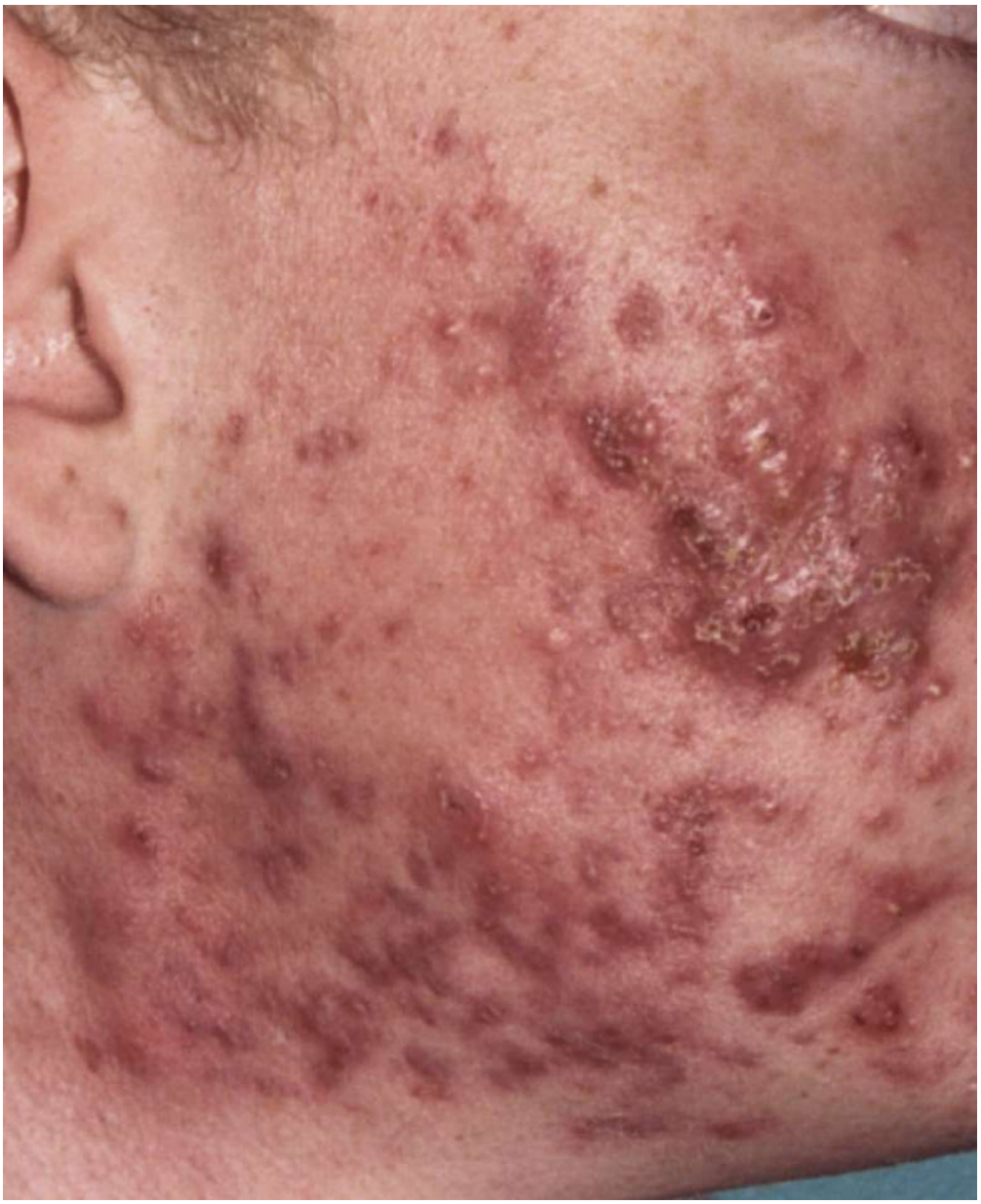

FIG. 12.1.6 Cystic acne. Acne should be treated with isotretinoin before reaching this stage.

- If there are prominent red papules or pustules, a course of oral antibiotics for 3–6 months in combination with a topical treatment is warranted (e.g. tetracycline 500 mg twice daily, erythromycin 500 mg twice daily, doxycycline 50–200 mg day^{-1}). Oral hormone therapy can help female patients.
- If the acne is severe, or if antibiotics and topical treatment have not resulted in considerable improvement in 3 months, oral isotretinoin (Roaccutane) is indicated. Prescription of isotretinoin is tightly

controlled because it is expensive and because it is teratogenic, requiring absolute avoidance of any pregnancy risk in girls. In boys, and in girls who do not become pregnant, it is safe and highly effective. Most adolescents with troublesome acne can be cleared with oral isotretinoin.

- Some acne sufferers develop scarring. This can be quite subtle and can occur with otherwise fairly mild disease. Any scarring is an indication for oral antibiotics and early consideration of oral isotretinoin. Similarly, the presence of any cysts should alert you to the likelihood of scarring and the need for oral therapy.

Acne and depression

Both adolescence and acne can be associated with depression. Significant depression is an indication to consider oral isotretinoin therapy. For many years, the issue of whether isotretinoin can trigger depression and suicide has been investigated. Current medical evidence suggests that treatment of severe acne with oral isotretinoin may decrease the risk of suicide. Assessment and treatment of any concurrent depression are required.

Acne fulminans

Acne fulminans typically occurs in young males being treated for acne. There is sudden onset of fever, malaise, arthralgia, myalgia, lymphadenopathy and/or hepatosplenomegaly in association with a rapid worsening of acne over the trunk and shoulders. Many painful cystic lesions develop and become haemorrhagic and ulcerated. Many laboratory abnormalities have been noted, including elevations of white cell count, erythrocyte sedimentation rate, C-reactive protein and liver enzymes. The cause is unknown but may be an abnormal immunological response rather than a primary bacterial infection. The response to oral antibiotics is slow, and indeed these adolescents are often taking oral antibiotics at the time of onset.

Management

- Oral prednisolone 0.5–1 mg kg^{-1} daily for 4–6 weeks (thereafter slowly reduced to zero)

- Oral isotretinoin being added to the regimen at the 4th week, initially at 0.5 mg kg^{-1} daily and gradually increased to achieve complete clearance.

Acne with Gram-negative folliculitis

This can develop in patients treated for acne with long-term antibiotics. It is more common in adolescents with an oily complexion. It presents as rapidly worsening acne with pustular lesions on the face. Several common gram-negative pathogens may be involved. Treat with oral trimethoprim and oral isotretinoin.

Early onset acne

Acne normally appears after the onset of pubertal changes such as testicular enlargement, pubic hair and breast development. However, comedonal and occasionally papular lesions may be the first manifestations of puberty in a child. These changes can begin up to 2 years before other signs of puberty. Early onset of comedonal lesions, even if mild, is associated with more severe acne several years later.

If comedones or acneiform papules are noted before puberty (see Fig. 12.1.1), you should consider the possibility of pathological androgen secretion (see below). Look for increased growth, genital development and advanced radiological bone age.

Atypical acneiform rashes

Acne that is atypical in age of onset, distribution, morphology or severity may be associated with systemic disease. Consider:

- glucocorticoid excess, either exogenous or endogenous. This can give a monomorphic acneiform rash on the face and trunk. Other cushingoid stigmata may be present. In children on corticosteroid treatment, the rash may appear as the dose of corticosteroid is being weaned. Glucocorticoid acne may be due to *Malassezia* folliculitis. If there is no response to tetracycline, treatment of *Malassezia* folliculitis with topical or oral antifungals may be required (see Folliculitis below).
- androgen excess. Other features depend on the age and sex of the child.

In younger children accelerated growth, body odour, pubic hair, clitoromegaly (but not breast development) and penile (but not testicular) enlargement may be present. In teenage females, polycystic ovary syndrome is commonly the cause. Other causes include late-onset congenital adrenal hyperplasia, and virilising tumours. Investigations may include X-rays for bone age, pelvic ultrasound, plasma androgens, fasting glucose and insulin, and response to dexamethasone suppression.

- precocious puberty
- drug induced. An acneiform rash may present with monomorphic papules and pustules in post-pubertal children. The distribution is different from acne vulgaris, often involving arms, legs or trunk, and comedones are not present. Affected individuals often have a history of troublesome acne vulgaris. Possible causative agents include anti-epileptic medications, especially phenytoin and phenobarbital, glucocorticoids, testosterone, isoniazid, lithium and iodides.
- Apert syndrome and related craniofacial syndromes can present with more severe widespread acne.

Folliculitis

Infection of the hair follicles is usually due to *Staphylococcus aureus*. Predisposing factors include a moist environment, a heavy bacterial load (e.g. adjacent to wounds or anal region) and excessive occlusive ointment, particularly on the trunk. Treatment involves avoidance of the predisposing factors, use of topical cleansers such as chlorhexidine and, rarely, oral antibiotics. Folliculitis often occurs repeatedly over a period of months and may require continued topical measures for 3–6 months. In resistant cases, daily use of topical benzoyl peroxide lotion may be helpful.

Folliculitis can occur in a child with moderate or severe eczema. Oral antibiotics are usually beneficial in this setting.

In adolescents, *Malassezia* folliculitis is common and presents as a persistent itchy rash with many tiny monomorphic erythematous papules and pustules on the back, shoulders and upper trunk. It is often associated with occlusion or sweating. Daily application of ketoconazole 2% shampoo is effective in gaining control. For resistant cases, use oral ketoconazole 400 mg weekly for 6 weeks taken with a glass of grapefruit juice followed by indefinite weekly shampooing

with selenium sulfide 2% shampoo.

Occasionally, pruritic papules and pustules can develop many hours after soaking in a hot spa. Lesions usually occur under the bathing costume and are caused by *Pseudomonas aeruginosa*. Oral antibiotics are not usually needed.

Acute generalised exanthematous pustulosis

Acute generalised exanthematous pustulosis (AGEP) appears as a rapidly developing facial or truncal erythema with hundreds of tiny sterile pustules. Oedema is often seen in the affected areas. Fever and malaise may be present. Healing is associated with extensive superficial peeling. In some cases, children presenting with AGEP have an underlying psoriatic tendency.

Many common medications, including amoxicillin, erythromycin, azithromycin, sulfonamides, anticonvulsants (especially carbamazepine) and non-steroidal anti-inflammatory drugs can cause this. AGEP drug reactions often begin within hours of commencing the drug. Other causes include group A streptococcal infection.

Treat by stopping any offending medication. Give oral antibiotics for underlying streptococcal infection if required.

Pustular psoriasis

Psoriasis can present with pustules on an erythematous background. The pustules are sterile. The affected area may be limited to fingers, palms and soles, or local areas of skin, often with an annular arrangement of pustules. Alternatively, there may be erythroderma, high fever, malaise and arthralgias with widespread pustules coalescing into sheets of pus. Local pustular psoriasis can be treated topically (see psoriasis). Generalised pustular psoriasis requires admission to hospital, rest, skin emollients and/or wet dressings, monitoring of fluid balance, electrolytes, renal and cardiac function and oral therapy (see erythroderma and psoriasis).

Neonatal pustules (see Chapter 3.3 Neonatal dermatology)

Neonatal pityrosporum folliculitis (see Chapter 3.3 Neonatal dermatology)

Papular (raised) rashes

If a child has itchy papules, consider scabies, urticaria, serum sickness, papular urticaria, molluscum, *Malassezia* folliculitis, dermatitis herpetiformis or Langerhans cell histiocytosis. If papules are not itchy, consider urticaria, molluscum, warts, acne, skin appendageal tumours, melanocytic naevi, Spitz naevi, pilomatricomas, keratosis pilaris, vasculitis and papular acrodermatitis. For raised red circles or rings, consider urticaria. For softer red, purple or blue swellings, consider haemangiomas or vascular malformations. For haemorrhagic papules, consider Henoch–Schönlein purpura and other causes of vasculitis. If papules are yellowish when blanched, consider juvenile xanthogranulomas or xanthomas.

Papules or nodules may also occur in a number of disorders including acute rheumatic fever, juvenile chronic arthritis, SLE and neurofibromatosis but are rarely the presenting feature.

Scabies

Scabies infection occurs as a result of close, usually repeated, contact with an infected individual. Scabies mites eat into the upper skin forming burrows a few millimetres in length around the fingers, palms, wrists, elbows, axillae, nipples, penis and soles. Early burrows may be vesicular. Usually only a few mites are present.

An intensely itchy secondary papular eruption develops 2–6 weeks after first exposure to the *Sarcoptes scabiei* mite or 1–4 days after subsequent reinfestation. This secondary eruption represents an immune response to the scabies antigen and does not mean that mites are spreading all over the body. Papules can occur anywhere, including the palms, soles, axillae and genitalia but are most prominent on the abdomen, buttocks and thighs. The scalp and head may be involved in infants and young toddlers. Inflammatory nodules may also develop, especially on covered areas. Excoriations and secondary impetigo may be present. Scabies is pandemic and affects both adults and children.

Not all itchy parasitic rashes are scabies. Many species of parasite can cause small itchy papules on the skin, in some cases hundreds of lesions. Bird mites, fleas, body lice, mosquitoes, sand flies, horse flies, bed bugs, ticks, chiggers, midges and harvest mites can all masquerade as scabies. Parasites can be

collected from the skin on transparent adhesive tape. The Commonwealth Scientific and Industrial Research Organisation Australian National Insect Collection provides an identification service on http://www.csiro.au/services/InsectID.html.

Management

- Treatment of scabies is expensive and upsetting. If there is any doubt about the diagnosis, confirm by scraping to find a scabies mite, identifying through a dermatoscope (all dermatologists and some GPs will have this) or refer before treating.
- Permethrin 5% cream
- Apply to dry skin (not after a bath) from the neck down to all skin surfaces. For infants, apply to the scalp as well (not face). Use mittens if necessary to prevent finger-sucking.
- Leave the cream on for at least 8 hours.
- Wash the cream off. Wash clothing, pyjamas and bed linen at this time.
- Remove soft toys from the bed. The scabies mite cannot live for long periods away from the body. Insecticide sprays, and cleaning of furniture and carpets, are not warranted.
- Exclude from school until after treatment has commenced.
- Treat all family members and any other people who have regular close skin contact with the affected individuals.
- An alternative, recommended for pregnant or neonatal cases, is sulfur 6% in yellow soft paraffin. This is unpleasant and has no proven safety advantages over permethrin in these groups. The following are *not* recommended: lindane 1% (contraindicated in infants or women who are pregnant or breast-feeding) or benzyl benzoate 25% (too irritant for children and ineffective if diluted).
- The itch takes a week or two to settle and can be treated with a topical or oral corticosteroid. Nodules may take months to resolve despite successful treatment of the scabies infestation.
- Reinfestation is common. To minimise the risk of reinfestation, the family should notify all social contacts (e.g. crèche, school or close friends) to ensure that all those infected receive treatment.
- Treat secondary impetigo with oral cephalexin 30 mg kg^{-1} (max 500 mg) three times daily.

- Post-streptococcal glomerulonephritis may be seen after chronic impetigo secondary to scabies in the indigenous population. Oedema, hypertension and haematuria may occur over the next 2 months. In a child with long-standing infected scabies where ongoing care may be suboptimal, review and urinalysis to monitor for renal involvement are warranted. Examination of the child and siblings for signs of rheumatic heart disease is also warranted in this patient population.

Papular acrodermatitis of childhood

Papular acrodermatitis of childhood usually occurs in children aged 1–3 years but can occur outside this range. It is a reaction pattern to many infectious agents, including coxsackie viruses, echoviruses, *Mycoplasma* species, Epstein–Barr virus, adenovirus, respiratory syncytial virus, rotavirus, cytomegalovirus, hepatitis B virus and others. It has also been reported after all standard childhood vaccines. It is common and a regular source of confusion in EDs.

Papular acrodermatitis of childhood is characterised by the acute onset of monomorphic, red or skin-coloured papules mainly on the limbs, buttocks and face, with striking sparing of the trunk (Fig. 12.1.7). In any given patient, the papules tend to all look the same, typically firm and dome shaped, measuring 2–4 mm in diameter. However, in different patients, the papules can vary from tiny, skin-coloured papules to larger urticarial plaques. Lesions may coalesce into patches on the extensor surfaces of the elbows and knees. Lesions may be papulovesicular (particularly on the limbs) or purpuric (particularly on the face). Hundreds of lesions may appear over several days. It is usually asymptomatic but may be itchy. Complete resolution occurs in 4–8 weeks.

Affected children may be otherwise well or may have features of the underlying infection, such as mild fever, malaise, coryza, sore throat, lymphadenopathy and splenomegaly. Lymphopenia or lymphocytosis may be present. In some cases, no history of a preceding or intercurrent illness can be found.

Papular acrodermatitis of childhood was formerly known as Gianotti–Crosti syndrome and was initially described as a manifestation of hepatitis B infection in Italy. This association has not been shown to be of general relevance, and papular acrodermatitis of childhood has been recognised as benign and much more common than previously thought.

Management

- Reassure and advise that clearing can take a few weeks.
- Children with papular acrodermatitis of childhood do not routinely require investigation to determine the underlying cause.
- Itch may require topical mid-potency CSs and/or oral antihistamine.
- No exclusion from school or crèche is needed.

Papular urticaria

This is a clinical hypersensitivity to insect bites and may occur in just one child within a household, even though all family members may be bitten. New bites appear as crops of asymmetrical, small, red papules, usually in warmer weather. Older bites appear as 1–5 mm papules, sometimes with surface scale or crust, or with surrounding urticaria. Vesicles or pustules may form in the centre of lesions. Individual lesions may resolve in a week or, if scratched, may last for months and may repeatedly flare up after fresh bites elsewhere. Itch is often intense, and secondary ulceration or infection can occur. Full resolution usually occurs within 9 months. However, scratching can lead to secondary changes, with non-healing erosions, ulceration, scarring and nodule formation. These lesions remain itchy, and a cycle of scratching and skin destruction can persist for years or decades if not treated. Improvements occur with new treatments, but relapses are common without considerable medical support.

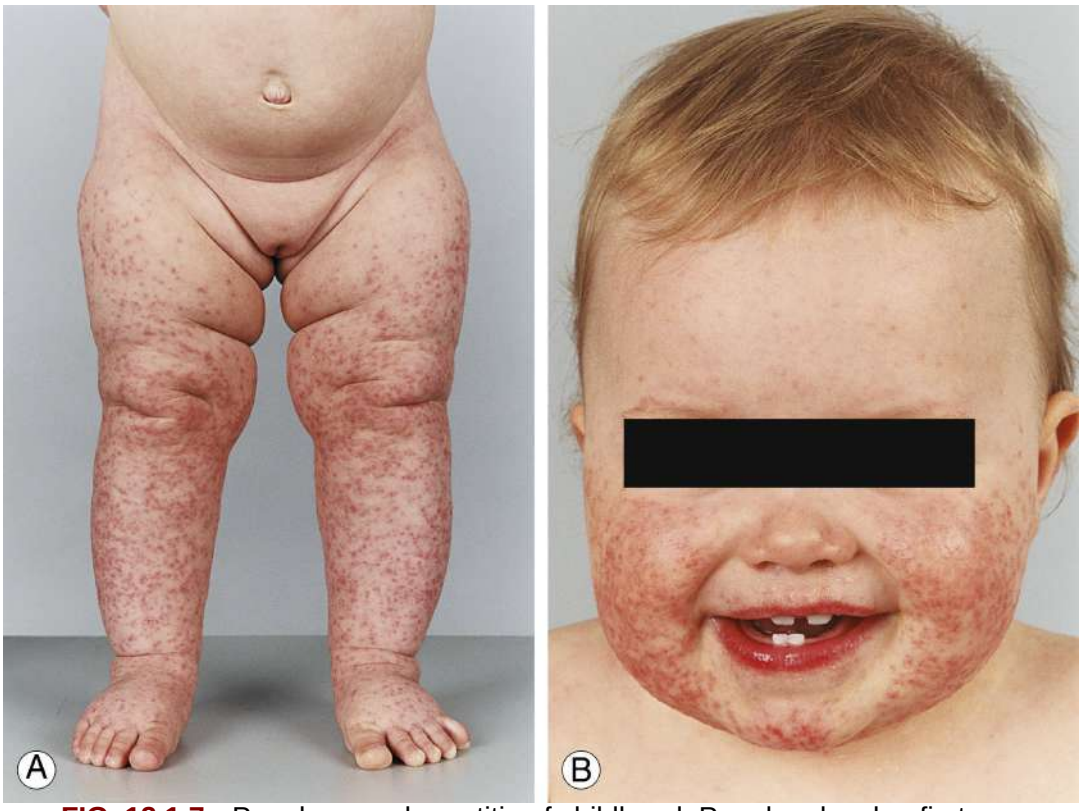

FIG. 12.1.7 Papular acrodermatitis of childhood. Papules develop first on the thighs and buttocks (A), then on the outer aspects of the arms, and finally on the face (B). The trunk is almost completely spared.

Management

- If the diagnosis is unclear, skin biopsy is useful as the typical histological findings are specific for insect-bite reactions.
- Prevent bites, e.g. by adequate clothing, modifying behaviour that leads to exposure, occasional use of repellent, and the treatment of pets and house for fleas and mites if necessary. The commonest source of bites is from mites in roof-dwelling animals such as birds, rats and possums. Surface spray of ceiling vents can be very useful.
- Treat the itch with an agent such as aluminium sulfate 20% (Stingose), liquor picis carbonis 2% in calamine lotion, potent steroid ointment or antihistamines.
- Protective dressings (e.g. Duoderm) can speed the healing of lesions. In older children, intralesional injection of corticosteroid is effective.
- Treat secondary infection with oral cephalexin 30 mg kg^{-1} (max 500 mg)

three times daily.

- For more persistent and severe lesions, referral for an intensive regimen may be necessary, including inpatient admission for moisturiser, potent topical steroid ointment and wet dressings, combined with continued intensive care of any relapse for weeks or months. Treatment needs to be modified according to response.

Molluscum

Molluscum is caused by a pox virus and is very common. Most children get at least a few molluscum at some stage between the ages of 1 and 12 years. Uncomplicated molluscum lesions are easily recognised as firm, pearly, 1–4 mm dome-shaped papules with central umbilication. They are sometimes misdiagnosed as vesicles on initial examination. A child may develop a few or a great many lesions, and individual lesions may last for months. Lesions can come and go for up to 3 years without causing any problems in most children. Complete resolution will not happen until an immune response develops, which may take from 3 months to 3 years.

Rarely, individual lesions can grow to over 1 cm in diameter. Individual lesions can become inflamed and, uncommonly, can develop secondary abscesses. More commonly, presentation to the ED is triggered by the development of widespread itchy and excoriated eczematous lesions in surrounding skin, usually on the lateral chest and axillary region or between the thighs. In such cases, recognition can be difficult, as the secondary changes can obliterate the primary lesions. A carefully taken history of the initial lesions is usually diagnostic.

Molluscum is common in the anogenital area, and occurrence at this site does not suggest child sexual abuse. Molluscum is common on the face, and occurrence at this site does not suggest underlying undiagnosed immunodeficiency.

Molluscum may occur on the eyelid margin or adjacent area. This can cause a recurrent or persistent unilateral conjunctivitis. The molluscum may be isolated and subtle and go unnoticed for months during which the child may receive multiple courses of antibiotic treatment for conjunctivitis (Fig. 12.1.8).

Management

- Most children with molluscum require no treatment.
- Children with molluscum should not share towels but should not be restricted otherwise in their activities, including no restriction of swimming. Although, it is possible that spread to other parts of the body and to other children may be reduced by advising the child not to soak in warm water (baths, spas, heated pools).
- Any child having problems with molluscum should be treated.
- The treatment depends on the age of the child, the location of the lesions and any secondary changes. Uncomplicated lesions not causing problems and not spreading can be left alone.

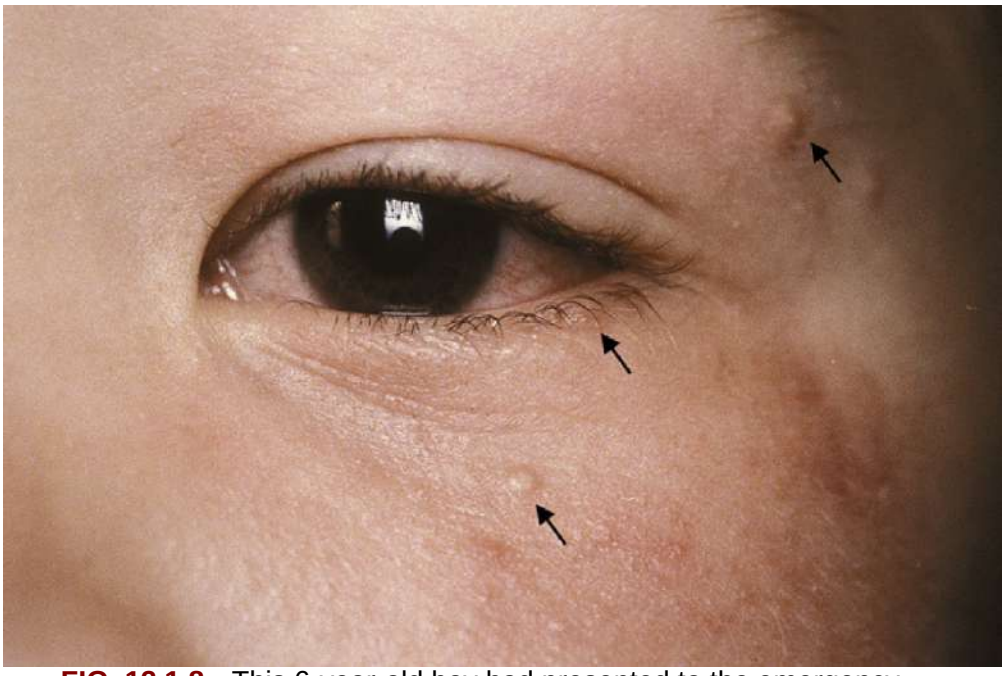

FIG. 12.1.8 This 6-year-old boy had presented to the emergency department four times in 6 months with recurrent unilateral conjunctivitis. Molluscum can be seen on the surrounding skin, and a small papule towards the lateral end of his lower lid margin is also a molluscum. Treatment of the molluscum cured the conjunctivitis.

- If the presenting problem is the itch from secondary eczema, treatment of the eczema with mild or mid-potency topical CS ointment may be all that is required.
- It is not difficult to destroy molluscum lesions. Simply deroofing the central umbilication is almost painless and leads to the papule becoming inflamed and resolving. In older children, this can be achieved with

superficial needle pricking or light cryotherapy. Benzoyl peroxide 5%, a variety of chemical irritants and tape stripping can all be used, but these can tend to flare any surrounding eczema. 10% Potassium hydroxide solution can be applied at home on lesions away from the mouth and eyes in cooperative children.

- Topical cantharidin (e.g. Cantharone) is a rapid and effective way of dealing with molluscum in many situations. This is painless on application and causes small blisters at the site over the next several hours. In Australia and New Zealand, cantharidin is difficult to obtain and is not warranted unless you see many children with troublesome molluscum (i.e. paediatric dermatologist).
- Imiquimod 5% cream nocte for several days may be useful for small numbers of lesions at difficult-to-treat sites.
- Several treatment options exist for molluscum around the eye and causing conjunctivitis. One or two lesions in older non-anxious children can be treated physically as for elsewhere on the face. Imiquimod 5% cream can be tried. If there are many lesions elsewhere, treating all the other lesions away from the eyes often induces rapid resolution of the eye lesions as well. In anxious children with significant problems with conjunctivitis, sedation or a brief general anaesthetic may be warranted.
- Inflamed lesions rarely warrant antibiotic treatment or drainage.

Adnexal tumours – pilomatricoma

Many different benign tumours of the various cell types in hair follicles, sebaceous glands and sweat glands can present as papules during childhood. The most common by far are pilomatricomas. These present at any age in childhood as a slow-growing papule on the head, face or neck (or occasionally elsewhere) that can be 4–40 mm in size. They may be skin coloured, white or bluish and are usually firm or hard, often due to calcification within the lesion. They may be lobulated. The main differential is an epidermal cyst. An ultrasound may confirm the diagnosis by demonstrating calcium shadows, which are not present in epidermal cysts. Pilomatricomas do not usually regress. Surgical removal is usually recommended because of the appearance but is not urgent.

Keratosis pilaris

This is a rough, somewhat spiky papular rash, mainly on the upper outer arms, thighs and/or cheeks with variable erythema. It affects 50–80% of all adolescents and approximately 40% of adults. About 30–50% of patients have a positive family history. Autosomal dominant inheritance with variable penetrance has been described. It appears in infancy and persists throughout life. However, it tends to improve towards adulthood. Rarely, the erythema and papules are florid and distressing and treatment is warranted.

Management

Reassure the patient that this is rarely a problem. Soap avoidance and moisturisers can improve the spiky feeling. Most children need no further treatment. Steroids do not help.

Granuloma annulare

Granuloma annulare is not common in children. It begins as a small skin-coloured or red papule that spreads outward over many weeks or months to give an annular, asymptomatic ring, usually on the hands or feet. Although often misdiagnosed as tinea, the epidermis is not scaly, and discrete subcutaneous papules can be felt all around the ring. It is slow growing and takes months to reach a size of 2 cm. Resolution usually occurs within a year or two. No investigation is necessary. Most children should be reassured and don’t need treatment.

Langerhans cell histiocytosis

Langerhans cell histiocytosis may present in several ways and should be considered as a possible diagnosis in any unusual, non-healing rash in infancy.

In neonates, Langerhans cell histiocytosis may present as a congenital self-healing form. One or a few papules are present at birth or shortly thereafter. They can be many millimetres in diameter and classically have a raised border and central necrosis. The papules usually show complete regression within months. Recurrence, years later, with visceral involvement or disseminated disease, has been reported, and long-term monitoring is warranted.

In infants, Langerhans cell histiocytosis can present as recalcitrant ‘cradle cap’ with scaly, papular and petechial scalp lesions, often mistaken initially for severe seborrhoeic dermatitis. Flexural areas, especially the groin and anogenital area,

can be involved, presenting as a chronic weeping ‘nappy rash’. The presence of papules and/or petechiae, sometimes with ulceration, and the lack of response to treatment for napkin dermatitis should raise suspicion.

In infants and older children, there may be widespread, often itchy, small haemorrhagic papules mimicking a vasculitis. Erythematous and purpuric scaling and papules may be present on the hands and feet, and nails may become dystrophic. Mucosal involvement can lead to ulceration on the gums or palate.

Extracutaneous disease can involve the liver, lymph nodes, marrow, bone and CNS. Diabetes insipidus can occur.

Management

- Confirm the diagnosis with a skin biopsy.
- Investigate for other organ involvement with skull, chest and skeletal X-rays, dental examination, full blood examination and liver function tests.
- Skin lesions can be treated with topical CS application.
- While Langerhans cell histiocytosis can be self-limited; it can also be rapidly progressive with progressive organ involvement and a poor prognosis, requiring intensive chemotherapy.

Juvenile xanthogranulomas

Juvenile xanthogranulomas are the commonest cause of yellow papules in children. Occurring in early childhood, lesions appear as low, dome-shaped papules 2–5 mm in diameter. They may be yellow or red-brown lesions that become yellow when blanched. They are often on the scalp but can occur elsewhere. There may be one or multiple papules appearing over weeks or months. Biopsy may be required to confirm the diagnosis. Resolution without treatment usually occurs within a few years. Juvenile xanthogranulomas are not associated with lipid disorders.

Children with neurofibromatosis have a considerably increased chance of having juvenile xanthogranulomas. The known increased risk of myelomonocytic leukaemia in a child with neurofibromatosis is probably not changed significantly if juvenile xanthogranulomas are also present, despite statements to the contrary in many texts.

Children less than 2 years old with multiple juvenile xanthogranulomas should

have an ophthalmology assessment. Ocular juvenile xanthogranulomas leading to glaucoma have been reported in this subgroup of patients.

Xanthomas

True xanthomatous papules associated with elevated cholesterol levels are rare in childhood. They may present as multiple eruptive xanthomas, tuberous xanthomas (usually distributed over the elbows, knuckles, buttocks, knees and heels) and tendinous xanthomas (usually on tendons around the elbow, wrist, hand or ankle). Investigation to exclude hyperlipidaemias is mandatory. Consider:

- primary hyperlipoproteinaemias. Skin lesions may present between 5 and 15 years of age, sometimes as tendinitis or tenosynovitis. Look for signs of atherosclerotic disease and a family history of high cholesterol, early myocardial infarcts or strokes.
- secondary hypercholesterolaemia. This can occur in hypothyroidism, biliary cirrhosis, diabetes mellitus, glycogen storage disease and nephrotic syndrome.

Angiofibromas in tuberosclerosis

Facial angiofibromas may be the first sign of tuberosclerosis. They appear as small, red-brown papules (not pustules) on the cheeks from about 5 to 6 years of age and can be misinterpreted as flat warts or early-onset acne. Facial angiofibromas can be treated with topical rapamycin or laser.

Red scaly (papulosquamous) rashes

Redness and scale indicate a combination of vasodilatation and epidermal involvement. Atopic eczema is the commonest cause of red scaly rashes in children. Other red scaly eruptions include psoriasis, tinea corporis, pityriasis rosea and pityriasis versicolor. If itch is present, consider any of the causes of eczematous rashes (p. 339).

Psoriasis

Psoriasis can occur at any age. Every year in a city of 2000,000 people, 200

children will present with psoriasis for the first time. Lesions typically begin as small, red papules that develop into circular, sometimes itchy, sharply demarcated, erythematous plaques with prominent silvery scale. However, psoriasis can present in many ways, including isolated thick scaly scalp lesions, scaly plaques on the hairline and behind the ears, annular lesions, pustular lesions, palmoplantar psoriasis, guttate psoriasis and flexural psoriasis. Therefore, psoriasis must be considered in the differential diagnosis of any red, scaly rash, particularly if well demarcated and not particularly itchy.

Guttate psoriasis describes the eruption of hundreds of small, scaly papules on the trunk and limbs, often following a streptococcal throat infection.

Flexural psoriasis can involve any of the skin folds, particularly the anogenital area, and is more common in children than in adults. Psoriasis at flexural sites presents as moist, non-scaly erythema, often painless but sometimes with secondary fissuring or streptococcal infection.

Minor nail pitting is often seen in childhood psoriasis.

Management

- Presentation to the ED is often precipitated by the onset of guttate psoriasis or by secondary problems with long-standing anogenital psoriasis. The treatment depends on the site and extent of disease and the age of the child.
- Minimise skin trauma, and use regular moisturiser.
- Guttate psoriasis is often most easily managed with an extemporaneous tar cream, e.g. liquor picis carbonis (LPC) 3% and salicylic acid 2% in Sorbolene cream, 500 g. Adolescents are less tolerant of tar creams. Swab and treat for streptococcal throat infection.
- Treat isolated skin plaques with either topical CSc (e.g. mometasone) or topical calcipotriol/betamethasone for 4 weeks, with clinical monitoring. Topical CSs are not used for large areas in childhood psoriasis because of the possible development of rebound pustular disease. Thick scalp plaques can be softened overnight with a tar cream and removed with a tar shampoo. Generally avoid tar cream on the face, flexures and genitalia as it can be irritating.
- Use hydrocortisone 1% or pimecrolimus 1% cream on the face, flexures and anogenital region.
- Palmoplantar psoriasis can be resistant to treatment. Initially treat with

topical potent CS or calcipotriol/betamethasone ointment.

- Secondary infection including perianal streptococcal infection may require treatment with oral penicillin. Recurrent or persistent streptococcal infection or carriage should be considered but is uncommon.
- A mild normocytic (occasionally microcytic) anaemia of chronic disease can occur in more severe cases.
- Appropriate medical follow-up is essential. Psoriasis will recur. Widespread or resistant psoriasis may need treatment with one or more of dithranol, UV therapy, acitretin, methotrexate, cyclosporin, or biologics, all of which may be effective.
- Widespread involvement and pustular psoriasis may lead to erythroderma with metabolic and other complications (see erythroderma).
- Psoriasis is associated with an increased risk of many problems including obesity, cardiovascular disease and arthritis. Ongoing follow-up and monitoring should include a broad-ranging assessment of health, weight and general well-being.

Tinea corporis

Tinea corporis (often called ringworm) is a common skin disorder, especially among children, but it may occur in people of all ages. It is caused by mould-like fungi (dermatophytes). Tinea infections can be transmitted by direct contact with affected individuals or by contact with contaminated items such as combs, clothing, shower or pool surfaces. They can also be transmitted by contact with pets that carry the fungus (cats are common carriers). The typical lesion is a slow-growing erythematous ring with a clear or scaly centre. Scale is usually most prominent on the outside of the annular ring. However, tinea corporis can present in a wide variety of ways. It can be pustular or vesicular, particularly on the soles (Fig. 12.1.9), or it can spread to many sites within days. Between the toes, it presents as an itchy, white, scaly and macerated rash (athlete’s foot). Previous treatment with steroid ointments often leads to partial improvement in symptoms but causes spreading of the rash, the development of papules and the masking of diagnostic features. Tinea should be considered in any red, scaly rash where the diagnosis is unclear, particularly if there is a gradually spreading eruption with inflammatory edges.

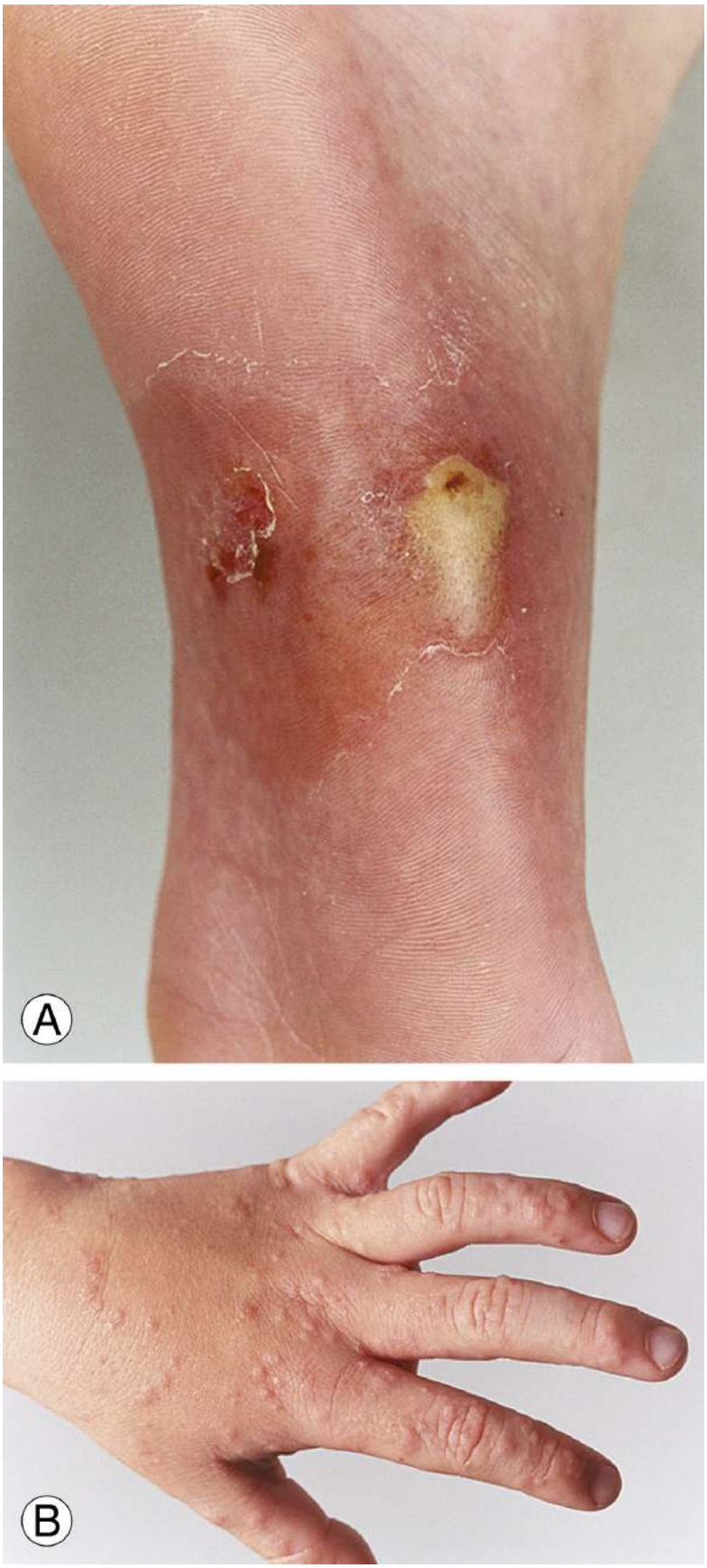
A
B

FIG. 12.1.9 Inflammatory bullous fungal infection. (A) The foot with (B) secondary id reaction involving the hands.

Management

- Confirm the diagnosis by scraping the scale for microscopy and culture.
- The family should identify and treat the source animal if any.
- Treat focal lesions with terbinafine cream (twice daily for 1 week) or an imidazole cream (e.g. clotrimazole, miconazole or econazole two to four times daily for 4 weeks).
- Rapidly spreading or widespread lesions or involvement of hair-bearing areas usually requires oral treatment, e.g. terbinafine (62.5 mg od 10–20 kg body weight, 125 od 20–40 kg, 250 mg od >40 kg) for 4–6 weeks for tinea corporis. Treatment for scalp infection is longer – see later in chapter.
- Combined steroid and antifungal ointments (e.g. Kenacomb) can be ineffective for clearing tinea corporis.
- Exclude from school until 1 day after treatment has commenced.

Pityriasis rosea

Pityriasis rosea is common from 1 to 10 years of age. The cause is uncertain but is thought to be viral. Human herpesvirus types 6 and 7 (HHV-6, HHV-7) have been implicated. A similar eruption may occur in response to a number of drugs. A pink scaly patch 2–4 cm in diameter near the shoulders or hips (herald patch) is the first sign in about 50% of children and is often misdiagnosed as tinea corporis because of its central clearing. However, unlike tinea, the scale is usually most prominent on the inside edge of the inflammatory ring. A few days or weeks later (or as the first feature if no herald patch is present), many pink/red, scaly, oval macules appear, mainly on the trunk, arms and thighs. The face, palms, lower legs and soles are largely spared. There may be thin scale within the lesions, and oval lesions may align with skin lines to give a ‘Christmas tree’ pattern on the back and trunk. Pityriasis rosea usually persists for 1–2 months. It may be mildly itchy but is often asymptomatic.

Pityriasis rosea may be atypical. Lesions may be papular, crusted, vesicular or purpuric. The distribution may involve neck and extremities.

Secondary syphilis can appear in a similar fashion, but mucosal and acral lesions are usually present. Secondary syphilis should be excluded in any adolescent at risk of sexually transmitted disease who presents with pityriasis rosea, particularly if palms and soles are involved.

Management

- Reassure the patient.
- Sunlight (or UV light in troublesome cases) will hasten clearing. Topical CSs do not help. Emollients, or occasionally oral antihistamine, are useful for itch.
- In an adolescent at risk for sexually transmitted diseases, arrange syphilis testing (venereal disease research laboratory [VDRL] test).

Secondary syphilis

Secondary syphilis presents 1–2 months after the primary chancre (which may be unreported). Erythematous, slightly scaly, macules or papules appear mainly on the trunk. There may be darker red macules on the palms and soles, as well as smooth or eroded papules on the tongue and oral mucosa, moist perineal papules (condylomata lata), annular lesions and pustules. Lymphadenopathy is usually present. Secondary syphilis can mimic pityriasis rosea and other red scaly disorders.

Seborrhoeic dermatitis

The term ‘seborrhoeic dermatitis’ has been used to describe a number of different clinical entities. It is most widely used for a particular dermatitis that occurs in areas of skin that have a high density of sebaceous glands, namely the scalp, the central ‘T-zone’ of the face, and the upper chest and back (not the anogenital area). It is due to a reaction to *Malassezia* (previously known as *Pityrosporum*) yeast, which is part of the normal flora at these sites.

As true seborrhoeic dermatitis can only occur in the setting of active sebaceous glands, the diagnosis should only be made after puberty has begun. The sites of predilection are the scalp, inner eyebrows and paranasal folds. The quality of the scale is more greasy than that of an eczema at other sites, and the degree of pruritus and discomfort is usually minimal. The degree of erythema

varies, and mild cases present as adolescent dandruff.

Infantile seborrhoeic dermatitis, resulting in greasy scales on the scalp and forehead and cradle cap during the first 3 months of life, has been described as a subtype of eczema; however, international opinion is divided as to whether this is the case or if it is a separate entity.

Atopic eczema is common on the scalp of infants and is differentiated from cradle cap by increased pruritus and discomfort, a harsher drier scale and occurrence after the age of 2 months. It is important not to diagnose scalp eczema as infantile seborrhoeic dermatitis as eczema will be further irritated by shampoos and ‘cradle cap’ creams.

Management

- Gently debride any built-up crust with a non-irritating product such as olive oil, bath oil or a soap substitute. Only if very thick scale is present should salicylic acid creams be used.
- Settle erythema with 1% hydrocortisone ointment.
- The use of an anti-yeast shampoo (e.g. 2% ketoconazole) may be helpful in more recalcitrant cases.
- Adolescents with seborrhoeic dermatitis may have to wash their hair more frequently.

Lichen striatus

Lichen striatus usually presents on a limb as a unilateral linear eczematous rash following the developmental lines of the skin. On close inspection, there are often collections of tiny flat-topped papules, coalescing into scaly lines that may stretch the full length of the limb. It may remain static for a period of months or a couple of years before spontaneously resolving. It may be pruritic but usually not significantly so.

The cause of the eruption is an inflammatory reaction in a streak of skin that has a genetic abnormality (mosaicism). The genetic difference of the streak is so subtle that the skin is phenotypically and functionally normal until a trigger, mostly a viral infection, induces an inflammatory reaction against the abnormal skin. As the condition is self-limiting, no treatment is necessary if the condition is asymptomatic. Moderate-strength to potent topical CS can help with any pruritus.

Eczematous rashes

The term ‘eczematous rashes’ covers several common conditions that are characterised by erythema, itching and disruption to the epidermis with oozing, crusting, fissures or excoriations. The degree of epidermal disruption may be minimal so that the presentation of atopic eczema may be with just red dry patches. Alternatively, the degree of epidermal disruption may be severe, with oedema and widespread vesiculation (e.g. plant contact dermatitis, which is discussed under vesiculobullous rashes, p. 327).

Atopic eczema – general issues

The term ‘atopic eczema’ covers a range of presentations that depend on the age of the child and on the child’s individual sensitivities. Several of these presentations are best seen as separate diseases. Clinical features and specific management suggestions are discussed under the individual headings below. However, some generalisations apply to most children with atopic eczema subtypes.

Atopic eczema usually begins in infancy. It commonly involves the face and often the trunk and limbs as well. In older children, the rash may be widespread but is often localised to flexures. Erythema, weeping, excoriation and, rarely, vesicles may be seen in acute lesions. Chronic lesions may show scale, lichenification and pigmentary changes. In some children, the lesions are more clearly defined, thickened discoid areas that may intermittently be itchy. There is usually a cyclical pattern of improvement and exacerbation.

Families who present to the ED with atopic eczema often have children with chronic and severe disease requiring frequent, time-consuming applications of topical medicines and wet dressings. These children may have recurrent infections, particularly with *Staphylococcus aureus* or HSV. Failure to thrive may also be present. A mild, normocytic (occasionally microcytic) anaemia of chronic disease can occur. Severe psychosocial and behavioural problems are common in these children although often hidden from medical staff. Parents may be under enormous stress with sleep loss, financial, marital and other problems secondary to the child’s eczema. These problems need to be identified and addressed.

Atopic eczema – general management principles

- *Education.* Parents need to know that treatments are effective in controlling the disease. They should be aware of the principles of avoiding relevant triggers and settling the inflammation.
- *Avoid irritants.* The following may worsen atopic eczema: soaps, bubble baths, prickly clothing, including clothing worn by adults carrying babies, seams and labels on clothing, car seat covers, sand, carpets, overheating or contact with pets. Smooth cotton clothing is preferred.
- *Avoid heat.* Overheating increases itch. Parents may overdress young children; outer clothing should be removed when entering warm environments. Bedding and baths should be kept comfortably cool.
- *Keep the skin moist.* Use a moisturiser such as sorbolene with 10% glycerine, aqueous cream or paraffin ointment (50:50 white soft paraffin/liquid paraffin) as often as four times a day if necessary. If a funded moisturiser is available use that.
- *Treat inflammation.* In mild or moderate cases, topical CS can be used with good effect. Hydrocortisone 1% is usually adequate for the face or mild disease in infants. For the body in infants and children moderate potency (e.g. methylprednisolone aceponate 0.1%, hydrocortisone butyrate 0.1%) or potent (e.g. mometasone 0.1% or betamethasone valerate 0.1%) ointment can be used for exacerbations. Topical CS should be applied once a day to all areas affected by eczema in sufficient quantities to leave a sheen on the skin. Topical CS can be applied to broken skin. The finger tip unit (i.e. the amount squeezed from a tube from an adult distal interphalangeal [DIP] crease to the finger tip) is 0.5 g and will cover an area approximately the size of two adult palms. To treat generalised eczema in a 1 year old will require approximately 7.5 g per application, for an adolescent approximately 30 g per application. Prescribe sufficient quantities to last until planned review.
- *Topical CS side effects:* It is very unusual to see side effects from topical CS in childhood; side effects from under treatment of eczema, however, are very common. Skin thinning has not been demonstrated in children using potent topical CS on the body for up to 9 months. Systemic side effects are unusual but most likely with long-term use of high potency topical CS under occlusion (e.g. nappies or wet wraps). Striae distensae may occur with use in the flexures or groin.
- *Control itch.* Advise parents to avoid saying ‘stop itching’ all the time

and to distract the child instead. Treating the eczema more effectively is helpful including use of wet bandaging if warranted. Antihistamines are often unhelpful, but if the itch is not controlled by other measures, they may be tried.

- *Treat infection.* Weeping and yellow crusted areas that do not respond to therapy may indicate secondary bacterial or herpetic (see p. 328) infection. Take cultures and treat with simple wet dressings and oral cephalexin 30 mg kg^{-1} (max 500 mg) three times daily. For recurrent bacterial infection use bleach baths, e.g. White King Bleach (4.2%) 3 tablespoons (= 45 mL) in a quarter full bath (approx 60 L) twice a week, and treat the eczema more aggressively.
- *Wet dressings.* Wet dressings are bandages applied over topical moisturiser or CS ointments two to four times a day. They cool the skin and are effective in controlling flares of eczema, settling troublesome focal patches of eczema and reducing the need for CS. They are well tolerated by young children, although cumbersome to apply. If possible, wet dressings should ideally be commenced by trained staff in hospital or with support from experienced nursing staff in the community.
- *Follow-up.* Children presenting with an acute flare who do not require admission should have a planned follow-up with their GP, paediatrician or dermatologist within 2–4 weeks to ensure improvement and form a longer-term management plan. Severe generalised or infected eczema or those with significant psychosocial complexity may require hospitalisation for wet wrap therapy and education.
- *Severe cases.* Do not accept that nothing more can be done. Children with chronic severe eczema need to be referred for psychosocial support and for dermatologist assessment as to whether they warrant admission to hospital, wet dressings, narrow-band UVB therapy or systemic immunosuppressive therapy (e.g. low-dose methotrexate).[2]

Atopic eczema – dietary principles

A normal diet is indicated in most children with eczema. If a child has immediate urticarial reactions to a particular food, that food should be avoided. No alteration should otherwise be made to an infant's or child's diet in an ED setting. Restrictive diets without professional supervision should be avoided.

Babies with a first-degree relative with eczema have a 50% chance of

developing eczema. For these babies, no modifications to the mother's diet are recommended during pregnancy and breast-feeding.

Atopic eczema – use of topical corticosteroid preparations

The use of topical CS remains a pivotal component of eczema management. It is important to settle the eczema and restore the natural integrity of the skin. If eczema is left untreated, the natural barrier of the skin remains disrupted, and the skin is more prone to react to other eczema triggers. Any scratching will further flare the skin, and a worsening cycle is created. Topical CS preparations are used to break this cycle and heal the skin. They should not be used as prophylaxis on normal skin. The preparations are usually best prescribed in ointment rather than cream formulation as these are more moisturising. As a general rule, topical CS use is extremely safe. Side effects are very uncommon if preparations are used appropriately.

Always ask parents if they are treating their children with herbal creams. Any herbal cream that gives dramatic clearing should be assumed to contain CS products until proven otherwise. A British study in 2003 looked at a number of herbal creams given to children for eczema.[3] Over 80% of those tested had illegal potent or very potent CS additives. Similar results have been found in previous studies.

Systemic effects of topical steroid use

Suppression of the pituitary-adrenal axis is very uncommon but potentially may occur if potent topical CS is used over most of the skin surface continuously for many weeks. If the child still has persistent eczema requiring ongoing use of large quantities of topical CS then consideration should be given to phototherapy or systemic treatments. If a child is using large amounts of topical CS in combination with inhaled corticosteroids and/or occasional oral cortisone therapy, you should consider adrenal suppression. Steroid therapy should not be abruptly stopped in this situation.

Local effects of topical steroid use

Irritation or allergy to one of the constituents of the topical CS occurs in a few children. Apart from this, local side effects rarely occur. Ongoing (months to years) of inappropriate use can lead to atrophy, striae, telangiectasia, purpura,

cataracts and juvenile rosacea. Care is required when treating the face, anogenital areas, flexures and any areas of rapid growth where the skin is already under tension (e.g. breasts in adolescent females). In these situations, permanent striae can occur within weeks. It should be explained to patients that once the eczema has cleared, the topical CS should be ceased. The topical calcineurin inhibitors, tacrolimus and pimecrolimus, can be used as steroid-sparing agents at specific body sites.

Atopic eczema – acute flare

Children with eczema often present to the ED with an acute worsening of previously stable eczema. This should prompt a search for an underlying cause. Look for any evidence of scabies, which can easily be overlooked amongst the background eczema – other family members may be itchy. Look for secondary bacterial infection. Consider HSV infection (p. 328). Small papules in widespread patches of eczema may be from subtle molluscum lesions. The child may have developed an irritant or allergic contact sensitivity to his/her topical treatments or other agents. Search for and treat any exacerbating factors. Arrange early outpatient paediatric or dermatology follow-up for management of the underlying eczema, or admit if severe.

Atopic eczema – admission to hospital

Children with eczema who attend EDs often do so because the parents have become increasingly desperate to get their child's skin under control. Admission to hospital can be very helpful. You should admit the child if either of the following circumstances apply:

- A child has severe impetiginisation or you have concerns about sepsis
- A child has widespread eczema herpeticum.

You should consider admitting the child or arrange dermatology outpatient review within 1–2 weeks if any of the following circumstances apply:

- A child is missing school because of atopic eczema.
- The family has social, financial or mental health issues that make home treatment difficult.

- Education of the parents in an emergency setting is difficult because they speak a language other than English.
- The parents exhibit significant stress or impending breakdown.
- An adolescent has widespread chronic eczema that affects his/her lifestyle (wears long clothing in summer, won't swim, etc.).

Atopic eczema – generalised infantile

Some infants present in the first 6 months of life with red, scaly and excoriated lesions covering much of the trunk and often the limbs, scalp and face as well. These infants are typically irritable and often sleeping and feeding poorly. The parents are often highly stressed by the difficulties with managing the child. Appropriate treatment usually leads to a dramatic change in the baby's behaviour and the family dynamics.

Unless there is a history of immediate hypersensitivity reactions (urticarial or anaphylaxis) no dietary changes should be advised in the ED setting.

Consider whether admission for wet dressings is required. Consider whether secondary bacterial or HSV infection is present. Treat with a bland emollient twice daily. Use hydrocortisone 1% ointment on the face and a moderate potency steroid to the body daily to gain rapid control of the skin. Arrange outpatient or GP follow-up within 1 or 2 weeks.

Atopic eczema – facial

Some infants will present with facial lesions. There may or may not be eczema elsewhere. Facial eczema may be secondarily infected with *Staphylococcus aureus*, resulting in weeping and crusting (Fig. 12.1.10). HSV infection needs to be considered (look for the typical monomorphic erosions, see p. 328). Saliva is often a significant exacerbating factor. Treat with ointment moisturiser several times daily and, if indicated, oral cephalexin 30 mg kg^{-1} (max 500 mg) three times daily for 1 week. Use 1% hydrocortisone to settle inflammation (or in severe cases a moderate potency steroid for the first few days, e.g. methylprednisolone aceponate 0.1%). Avoidance of irritating factors, such as napkin wipes being used on the face, and general management measures are required to prevent relapse. Arrange follow-up within 1–2 weeks.

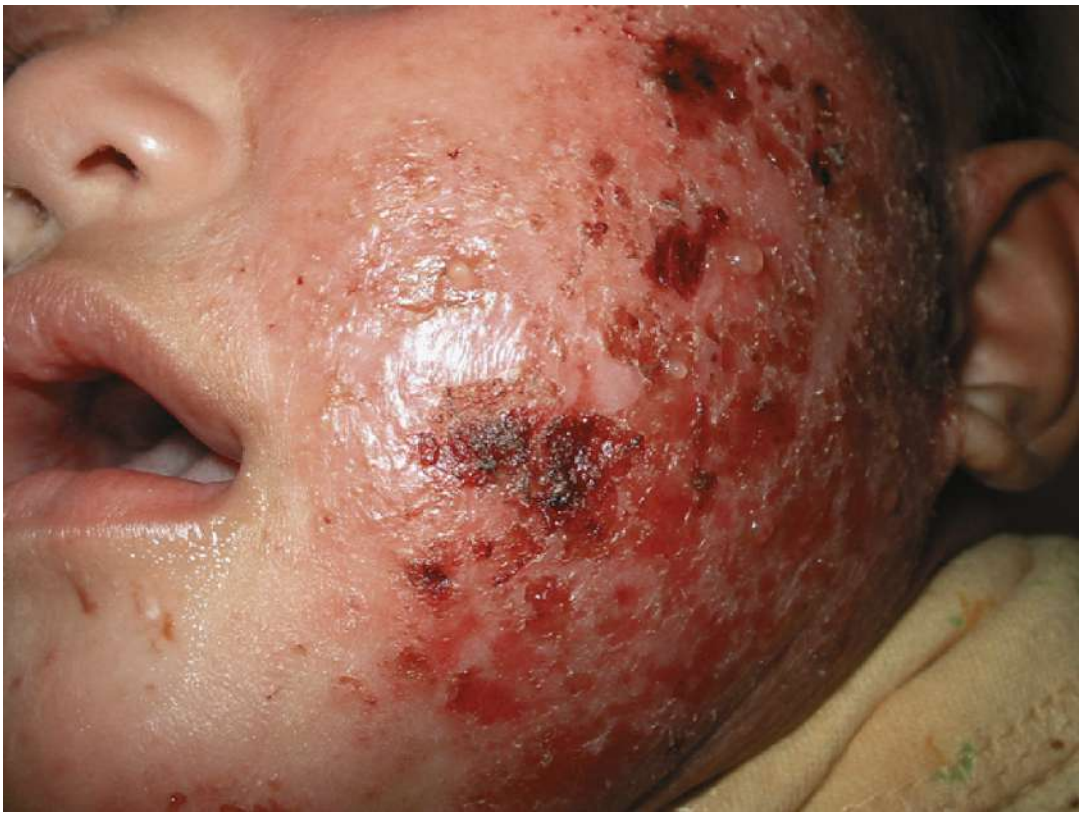

FIG. 12.1.10 Facial eczema with secondary impetiginisation in a 10-month-old boy.

Atopic eczema – perioral eczema versus juvenile rosacea

Perioral eczema

This refers to eczema round the mouth. This is common in infancy and early childhood. Irritation from saliva is the main cause. Occasionally, intolerance or allergy to foods can play a role in the older child. Confluent patches of erythema with weeping and superficial erosion occur in acute eczema. Management involves settling the active patches with a brief period of topical CS ointment and then trying to minimise irritation from saliva. This is difficult in the dribbling infant. Thicker moisturisers such as 50% white soft/liquid paraffin (Dermeze) or plain Vaseline may be used both as a moisturiser and as a barrier on the skin. In persistent cases use pimecrolimus 1% cream or tacrolimus 0.03% ointment. Improvement usually occurs between 18 and 24 months of age.

Juvenile rosacea

This is often called ‘perioral dermatitis’ and confused with perioral eczema, but it is not a true dermatitis. It is better thought of as a subset of rosacea. The most common clinical setting is following the use of topical CSs in a young child who is genetically predisposed to develop this condition. The morphology is quite different from perioral eczema. Erythematous papules occur around the mouth but spare the skin immediately adjacent to the lips. Other facial areas may be involved. The papules are essentially distinct from each other but may coalesce at times. Topical CSs tend to temporarily settle the inflammation and then cause a rebound flare, and patients are often under the false impression that the topical CSs are the only effective therapy. Treatment involves the cessation of all topical CSs on the skin in this area and the introduction of an appropriate antibiotic. Use erythromycin in children or tetracyclines in adolescents. Pimecrolimus 1% cream may give added benefit. Resolution usually takes 3–4 weeks of therapy. It is important to warn patients who have been actively using topical CSs that there will be a rebound flare before they begin to see improvement.

Atopic eczema – periorbital

Periorbital eczema is usually associated with sensitivity to airborne allergens, especially house dust mites, cat dander and pollens. Consideration of the occasions when the itch is worst will often indicate the causative agent. It often occurs in conjunction with allergic rhinoconjunctivitis, and treatment with antihistamine eye drops may be of benefit. Formal skin prick testing, allergen avoidance measures, moisturiser, 1% hydrocortisone ointment or pimecrolimus 1% cream and follow-up are required. In atypical cases, consider allergic contact dermatitis, irritant contact dermatitis and molluscum.

Atopic eczema – molluscum

Molluscum infection can trigger a marked local or generalised reaction in children. Presentation to the ED follows the development of widespread itchy and excoriated eczematous lesions in surrounding skin, usually on the lateral chest and axillary region or between the thighs. If the child has a previous history of atopic eczema, the molluscum lesions may be overlooked or hidden in the eczema. A careful history may reveal that clear, firm papules were present at some stage. A careful examination may reveal some typical molluscum or some

residual papules in the centre of eczematous patches. Treat both the eczema (as above) and the molluscum (see p. 335).

Atopic eczema – discoid

Discoid eczema is characterised by chronic focal circular crusted lesions 1–5 cm in diameter. The discoid lesions are thickened, oozing and itchy. They are relatively resistant to therapy and often require potent topical steroids. Secondary staphylococcal infection is often present. Intensive and prolonged therapy may be needed to break the cycle of itch, lichenification and infection. Oral cephalexin 30 mg kg^{-1} (max 500 mg) three times daily for 7 days to treat infection, two to three times daily moisturiser, and potent topical steroid ointment may be adequate, but therapy may need to be prolonged. Follow-up is mandatory. Hospital admission for wet dressings, ultraviolet light therapy or oral immunosuppressants may all be considered.

Atopic eczema – juvenile plantar dermatosis

Juvenile plantar dermatosis is characterised by erythema, dryness and cracking of the anterior sole and the under surface of the toes. It is usually seen in mid-childhood and tends to occur in children with a tendency for increased sweating. The child may or may not have atopic eczema elsewhere. Both feet are usually symmetrically involved in the weight-bearing skin on the soles. Juvenile plantar dermatosis may persist for years, and exacerbations may be associated with painful fissures. The differential includes tinea (which should be excluded by culture), psoriasis and contact dermatitis, which can all present with red scaly lesions on the soles. Cotton socks, leather shoes and regular ointment moisturiser three times daily should be used. Cotton socks can be worn over the moisturiser. Minimise exposure of cracked skin to carpets and other irritants. Potent topical CS ointments may be necessary. Follow-up is needed.

Atopic eczema with systemic associations

Eczematous skin lesions can also be associated with:

- immunodeficiency syndromes. Neonatal erythroderma can be due to forms of severe combined immunodeficiency. Netherton syndrome should be considered if neonatal-onset eczema is associated with failure

to thrive and sparse hair. Hyper IgE syndrome, DOCK8 deficiency and other immunodeficiencies may present with eczema, failure to thrive, recurrent infections, and diarrhoea. Wiskott-Aldrich is associated with eczema, thrombocytopenia and petechiae.

- multiple food allergies with consequent dietary restrictions and secondary nutritional and psychosocial problems
- metabolic or nutritional disorders. Phenylketonuria often presents with eczema. Less typical eczematous lesions, more prominent in periorificial areas, are seen in biotin, essential fatty acid and zinc deficiency syndromes. Malnutrition and organoacidaemias, e.g. methylmalonic acidaemia, can result in similar lesions.

Irritant contact dermatitis

The most common forms of irritant contact dermatitis in children are irritant napkin dermatitis (p. 355), juvenile plantar dermatosis (see above) and lip licking. Lip licking refers to a dermatitis caused by chronic exposure to saliva. Usually the application of saliva by the child is presumed by the carers to be secondary to the dermatitis rather than causative and may not be mentioned. Lip licking often presents with a clearly defined ring of dermatitis around the mouth, but the pattern of involvement may vary considerably depending on the behaviour of the child. Any habit that spreads saliva in a particular direction will affect the pattern of the dermatitis. Acidic foods, artificial colours, preservatives and toothpaste may contribute to the irritation. Treatment requires education, frequent topical ointment barrier applications (e.g. Vaseline) and immediate treatment of any relapse. A short period of topical 1% hydrocortisone ointment or pimecrolimus 1% cream may be needed. Secondary bacterial or candidal infection is rarely present.

Allergic contact dermatitis

Acute allergic contact reactions (e.g. to plants) often cause vesiculation and are considered under vesiculobullous rashes (p. 327). Chronic exposure to a contact allergen presents with erythema, itch and lichenification. The site of reaction usually suggests the cause: axillary rashes from deodorants, ears from earrings, lips and eyes from make-up, lower abdomen from nickel buttons on pants, wrist from watch, perioral from toothpaste, black henna from tattoos and so on.

However, the trigger may be overlooked. For example, an itchy eczematous patch on the scalp may be due to a metal stud in a cap that is not being worn at the time of presentation to the ED. Consider allergic contact dermatitis if a child has persistent eczema in an unusual distribution.

Generalised dry skin – ichthyosis

Ichthyosis refers to generalised scaly skin. There are several forms. Some ichthyoses only affect the skin, but several have associated abnormalities in other organs. Eczema does not usually present as generalised dryness in babies. Unless there is a clear history of familial ichthyosis vulgaris, an infant with significant dryness should be referred for assessment by a skin specialist.

Ichthyosis vulgaris is the commonest of the ichthyoses. It has autosomal dominant inheritance and occurs as an isolated finding in 1 in 200 children.

X-linked ichthyosis is less common and may be undiagnosed for years or decades. About one-third of affected boys are diagnosed for the first time by an observant clinician during an emergency attendance for an unrelated problem. A fine scaling at birth is later replaced by larger brown scales. Diagnosis is confirmed by enzyme analysis for the affected steroid sulfatase gene or by genomic analysis. Boys with X-linked ichthyosis are usually otherwise normal but need to be monitored for hypogonadism, cryptorchidism, anosmia, short stature and mental retardation. Female carriers may have obstetric difficulties leading to prolonged labour.

In Sjögren–Larsson syndrome, a yellow-brown lichenified appearance is present in infancy. This evolves into a more florid scaling with a symmetric spastic paralysis and mental retardation. In some trichothiodystrophies, ichthyosis is associated with brittle hair and mental and growth retardation. Several ichthyoses are linked with deafness, cataracts or other eye problems. Mild, generalised scaling may be the first feature in later-onset Refsum disease, before development of multiple visual problems, deafness and neuropathy.

Generalised dry skin – ectodermal dysplasias

Ectodermal dysplasias are a heterogeneous group of conditions characterised by congenital, non-progressive abnormalities of hair, teeth, nails and sweat glands. The skin is dry and may be hyperpigmented. Deficient sweating may cause overheating in infants at any time and in older children in summer. Children may

present with undiagnosed recurrent fevers or with heat prostration, collapse or death. Teeth are often poorly developed or absent. Cleft lip and palate, limb abnormalities and mucous retention in the upper airway and ear may be present. Accurate diagnosis, genetic counselling, dental and audiological review, appropriate moisturiser and topical cortisone use if needed, education about avoiding overheating and regular follow-up are required.

Red blanching rashes (erythematous)

Erythematous rashes are common in children. They are most commonly caused by viral infections (e.g. Coxsackie virus, echovirus, Epstein–Barr virus, adenovirus, parainfluenza, influenza, parvovirus B19, HHV-6, rubella and measles) or by a drug reaction. Consider also septicaemia, scarlet fever, Kawasaki disease and *Mycoplasma* infection.

Fever and exanthem

Fever and rash are common presentations to the ED. The most common cause is a viral illness. Some infections have specific clinical features that aid diagnosis, e.g. measles and erythema infectiosum. However, in most instances, a specific viral diagnosis cannot be made with certainty. To manage such a child, consider the following:

- Is the child sick? Is the child lethargic or peripherally cold? Is he/she young? Consider meningococcal disease, other bacterial sepsis, and Kawasaki disease. These children require urgent investigation and appropriate treatment guided by the findings, and resuscitation may be needed.
- Is the child taking medications? Consider ceasing any medication.
- Are there other people at risk? If the child has any relatives that are immunosuppressed or pregnant, consider testing the child with serology, viral swabs and culture and advising the at-risk person to consult his/her doctor.
- Is the rash papular? Consider papular acrodermatitis (p. 335).
- Is the rash itchy? If so, it may be primarily urticaria or a dermatitis.
- If the answer to all the above is ‘no’, reassurance and review are probably appropriate.

Scarlet fever

Group A streptococci cause a variety of diseases including scarlet fever, pharyngotonsillitis, impetigo, cellulitis, otitis media, streptococcal toxic shock syndrome, necrotising fasciitis, glomerulonephritis and rheumatic fever. Group A streptococcal infection usually occurs in school-age children.

Scarlet fever begins with a prodrome consisting of sudden onset of high fever, vomiting, malaise, headache and abdominal pain. This is followed within a few hours by rash. The typical rash is a diffuse, pink-red generalised 'flush' with pinhead spots that feels like sandpaper. The rash blanches. It is first noted on the upper chest, before spreading to the trunk, skin folds, neck and extremities. The face is often flushed with circumoral sparing. It does not usually involve the palms or soles. Other features include strawberry tongue (initially white, then red days 4–5), pharyngotonsillitis and tender cervical and submaxillary nodes.

Confirm diagnosis by throat swab and serology. Notification is not required. Transmission occurs by direct contact. Isolate the child from school or crèche for 3 days after the start of treatment. Treat with phenoxymethylpenicillin (penicillin V) 250 mg orally (under 10 years), 500 mg orally (over 10 years) twice daily for 10 days. For patients with penicillin hypersensitivity, use cephalexin.

Toxic shock syndrome

Toxic shock syndrome is an acute, febrile illness with a characteristic rash and multiple system involvement. It has potential complications that include shock, renal and myocardial failure, coagulopathy and respiratory distress syndrome. Toxic shock syndrome has been associated with *Staphylococcus aureus* and group A streptococcal infection.

Presentation is initially similar to that of scarlet fever, but other features include myalgia, profuse diarrhoea, conjunctival injection and hypotension. Desquamation of digits, palms, soles and perianal region occurs 7 to 21 days later.

Initial management should focus on resuscitation. Antibiotic treatment should include IV flucloxacillin 50 mg kg^{-1} (max 2 g) every 4–6 hours. Clindamycin 5–10 mg kg^{-1} every 6–8 hours oral or IV should be added as it has been shown (*in vitro*) to inhibit the release of toxin. As gram-positive toxic shock syndrome may be indistinguishable from gram-negative sepsis at presentation, empiric cover should be broad.

Kawasaki disease

Kawasaki disease (see Chapter 5.9 Kawasaki disease) is mainly seen in children between the ages of 6 months and 5 years. It is an acute, self-limiting vasculitic illness and a major cause of acquired coronary artery disease in children in high-income countries. An understanding of the timing of clinical features aids diagnosis. The cardinal feature is a high fever persisting for 1–4 weeks with unusually severe irritability unresponsive to antipyretics. Dilatation of conjunctival vessels is seen within a few days of the onset of fever in 90% of children but may be subtle and evanescent, and there is no conjunctivitis or purulent discharge. In 60% of children, cervical lymph nodes are enlarged as a firm mass at the onset of fever. In older children, there may be striking, unilateral tender lymphadenopathy thought to be cervical adenitis but unresponsive to antibiotics.

Within a few days of onset of fever, 90% of children develop a widespread erythematous rash. This may include urticarial and maculopapular lesions that increase to several centimetres in diameter. There is usually involvement of the anogenital area. There are no vesicles or crusting, but there may be a few pustules on elbows or knees. The rash lasts for 1–7 days.

Within a day or two of the rash, 90% of children develop changes of the lips and mouth. The lips become red, dry, fissured and occasionally crusted and bleeding. The oral mucosa is red without ulceration. These changes can persist for 2–3 weeks.

Within a day or two of the development of a rash, about 95% of children develop erythema of the palms and soles, often with associated oedema to give a shiny swollen appearance to the hands. The swelling can persist until the fever resolves. About 2 weeks after the onset of fever, desquamation begins at the fingertips and spreads to involve the palms, followed a few days later by desquamation of the toes then soles. Fingertip desquamation will usually be seen even if no palm or sole involvement was noted earlier. This is an important feature as palm and sole involvement in a widespread eruption is otherwise uncommon.

Less common features include arthritis, diarrhoea, vomiting, coryza, cough and hydrops of the gallbladder.

In babies under 6 months of age, and sometimes in older children, Kawasaki disease may present as prolonged fever with only one or two of the above features. Untreated, about 25% of children with Kawasaki disease develop

coronary artery changes. These can occur up to 6–8 weeks after the onset of fever and may ultimately be fatal. Any child with persistent fever of unknown cause should be carefully screened for any manifestations of Kawasaki disease. Investigations should include full blood count, erythrocyte sedimentation rate, liver function tests and group A streptococcal and other serology to exclude possible differential diagnoses. If Kawasaki disease is suspected, echocardiography should be performed at least twice: at presentation and again at 6–8 weeks. Treatment with IV immunoglobulin (2 g kg^{-1}, repeated if fever persists) and aspirin (10 mg kg^{-1} every 8 hours while febrile, then 3–5 mg kg^{-1} orally daily for at least 6–8 weeks) may be life-saving. Regardless of the initial echocardiogram result, treatment should be given as early in the course of the illness as possible, to minimise the risk and severity of cardiac problems.

Erythema infectiosum

Erythema infectiosum, also known as slapped cheek disease or fifth disease, is caused by parvovirus B19. It begins with a non-specific prodrome of fever (15–30% of cases), malaise, myalgia and headache. The distinctive rash has three stages:

1. Slapped cheek appearance (1–3 days)
2. Maculopapular blanching rash on the proximal extensor surfaces, flexor surfaces and trunk. This fades over several days with central clearing and then forms a reticular pattern (after 7 days).
3. Reticular rash reappears with heat, cold and friction (weeks/months).

Arthralgia and arthritis occur infrequently in infected children but are common in adults. Other potential complications include aplastic crises in children with haemoglobin abnormalities and chronic anaemia in children with human immunodeficiency virus (HIV) infection.

Treatment is supportive. Children with erythema infectiosum are highly infective before the onset of illness and are probably not infective once the rash appears. They do not need to be excluded from school or childcare. Seek specialist advice if the child is known to have a blood disorder or to be immunosuppressed.

Pregnant women who contract parvovirus B19 have a small risk of fetal anaemia and death but do not have an increased risk of fetal abnormalities. It is

not practicable to prevent exposure at home, and exclusion of pregnant women from work is not recommended.[4] Pregnant women in contact with a child with parvovirus B19 infection should be advised to consult the doctor supervising their pregnancy.

Roseola infantum

This common viral exanthem is caused by HHV-6 and in some cases by HHV-7. Ninety-five per cent of children have been exposed to HHV-6 by the age of 2 years. Up to 30% of all infants will present with the clinical features of roseola. Typically, an infant has a high fever for 2–4 days and is often prescribed oral antibiotics. Despite the high fever, the child remains well and active. Occipital and cervical lymphadenopathy is frequently present. The fever then disappears, but at the same time a widespread erythematous rash appears on the face and trunk. This commonly leads to presentation to the ED. Recognition of this condition is important so that both doctor and family realise that the rash is not a reaction to the antibiotics. After the appearance of the rash, the child remains well, and no isolation is necessary.

Enteroviruses

Coxsackie A, B and echoviruses are all enteroviruses that cause a range of childhood illnesses, particularly in the summer months. They can cause several types of exanthem, including maculopapular, erythematous, vesicular and petechial rashes. Hand, foot and mouth disease represents one particular syndrome, usually caused by Coxsackie A16 virus and less commonly by other group A and group B Coxsackie viruses and enterovirus 71 (EV71) (see p. 328). Pharyngitis is a frequent feature of enteroviral infection. Encephalitis is a less-common accompanying presentation.

Infectious mononucleosis

This syndrome is most commonly caused by Epstein–Barr virus but may also be associated with cytomegalovirus and other viruses. Clinical features include fever, generalised lymphadenopathy, exudative tonsillopharyngitis, palatal petechiae and hepatosplenomegaly. Children usually have milder disease than adults.

Rash occurs in up to 20% of children in the first few days of the illness. The rash may be erythematous, maculopapular or morbilliform. There is an increased incidence of rash in children with infectious mononucleosis if treated with amoxicillin or other penicillins. This rash is typically maculopapular and pruritic and occurs mainly over the trunk.

Epstein–Barr virus has a number of other less-common dermatological manifestations, including papular acrodermatitis, oral hairy leukoplakia and cutaneous lymphoproliferative disorders.

Diagnosis may be suggested by absolute and relative lymphocytosis and an increased proportion of atypical lymphocytes on full blood examination and abnormal liver function tests. The monospot test for heterophile antibodies is positive in 90% of cases except in those under 4–5 years of age in whom the sensitivity is poor. Diagnosis is confirmed by serology. Isolation is not required. Children with significant throat symptoms may need admission for supportive care. Oral prednisolone for 5 days may assist resolution of symptoms. Children with splenomegaly should be advised to avoid contact sports until improved.

Measles

As a result of widespread measles immunisation, this disease is now seen infrequently. However, outbreaks continue to occur in most parts of the world. The hallmarks of measles are cough, conjunctivitis and rash. The rash appears 3–4 days after the prodrome of fever, conjunctivitis, coryza, cough and Koplik spots (white spots on a bright red buccal mucosa). The rash is initially red, blanching and maculopapular. It begins around the ears and hairline and spreads to the trunk and the proximal arms and legs. It becomes confluent by the third day. High fever often persists after onset of the rash. Uncommonly, a child will develop otitis media, pneumonia or encephalitis. Subacute sclerosing panencephalitis is a rare, fatal, late complication.

Measles vaccination gives a transient mild exanthem in about 5% of cases. If a child has been previously immunised with killed measles vaccine and later contracts measles, the presentation may be unusual. This ‘atypical measles’ often has high fever and malaise, no cough, no eye signs, no Koplik spots and a rash that is more distal and often purpuric.

Measles is highly contagious and spread by airborne droplets or direct contact with infected nasal or throat secretions. Suspected cases presenting to the ED should be managed in a separate area to other patients in the department,

preferably in a negative pressure room. Diagnosis should be confirmed by serology and PCR of nose and throat swabs. Laboratory confirmation and notification to the health department are essential to prevent epidemics. The child should be excluded from school or crèche for 5 days.

If an unimmunised child over 9 months of age has contact with measles, measles infection can be prevented by MMR vaccination within 72 hours. This is because the incubation period of the vaccine strain is shorter (4–6 days) than the incubation period of wild measles virus (10–14 days). Older contacts who do not have documentation of immunisation with measles vaccine should be tested for measles IgG if this can be performed within 72 hours. If seronegative, or if serology cannot be performed, contacts should be immunised within 72 hours. Infants under 9 months old who have contact with measles should be given normal human immunoglobulin within 7 days.

Rubella

Rubella infection is asymptomatic in 25–50% of children. Affected children are usually only mildly unwell. The prodrome lasts up to 5 days and consists of low-grade fever; malaise; headache; coryza; and postauricular, occipital and posterior triangle lymphadenopathy. The rash is characterised by small, fine, discrete, pink maculopapules. It starts on the face and spreads to the chest and upper arms, abdomen and thighs, all within 24 hours.

If a child is over 1 year of age and known to have received rubella vaccination, a subsequent exanthem is highly unlikely to be rubella, and investigation is not required. If rubella is suspected in younger children or you cannot confirm that the child has been previously vaccinated, PCR of a nasal or throat swab should be performed. If diagnosis is confirmed, notification to state health departments is required in most states. The child should be excluded from school or crèche for 5 days. Pregnant women in contact with a child with rubella infection must consult the doctor supervising their pregnancy without delay.

Unilateral laterothoracic exanthem

Unilateral laterothoracic exanthem has only been well recognised in the past decade but is another common presentation to the ED. It usually occurs between the ages of 1 and 4 years. There is often a mild fever with gastrointestinal or upper respiratory symptoms 1 to 3 weeks before the onset of rash. The rash

begins in one axilla or on one side of the chest as erythematous, urticarial, eczematous or papular lesions. Uncommonly, the rash may start in the inguinal region or on the thigh. Over a period of a week, the rash spreads to give a strikingly unilateral involvement of the lateral chest, axilla and arm. Over the next 2 weeks, there may be some spread to the other side and other areas. Itch may be present but is not marked. Coryza, pharyngitis and regional lymphadenopathy may occur. Complete resolution occurs without sequelae within a total of 4 to 6 weeks.

Although the epidemiology of unilateral laterothoracic exanthem suggests an infective cause, no causative agent has been identified despite exhaustive searching. No isolation or restriction of activities is needed. Moderate potency topical corticosteroids may give some relief, but often no treatment is needed.

Urticaria

Urticaria is common. It is characterised by the rapid appearance and disappearance of multiple raised red wheals on any part of the body. Circular erythematous macules or swellings, sometimes with central pallor, appear, migrate and disappear over minutes or hours (Fig. 12.1.11). Individual lesions are often itchy and resolve within 1 day. There may be central clearing to give ring lesions (these are not the target lesions of erythema multiforme, which always persist for several days).

In some children the urticarial wheals do not fully blanch with pressure. As the lesions migrate or grow circumferentially, they leave a purplish non-blanching tinge on the skin, indicating some degree of leakage of red cells from capillaries. Frank purpura is not usually present. Typically, these children are otherwise well and are reacting to the same group of possible triggers as children with normal urticaria. Occasionally, this may be a presentation of serum sickness or cutaneous or systemic vasculitis. Features that raise suspicion about an underlying vasculitis include weals lasting more than 24 hours, bruising within weals, painful lesions or associated fever, arthralgia, abdominal pain or haematuria.

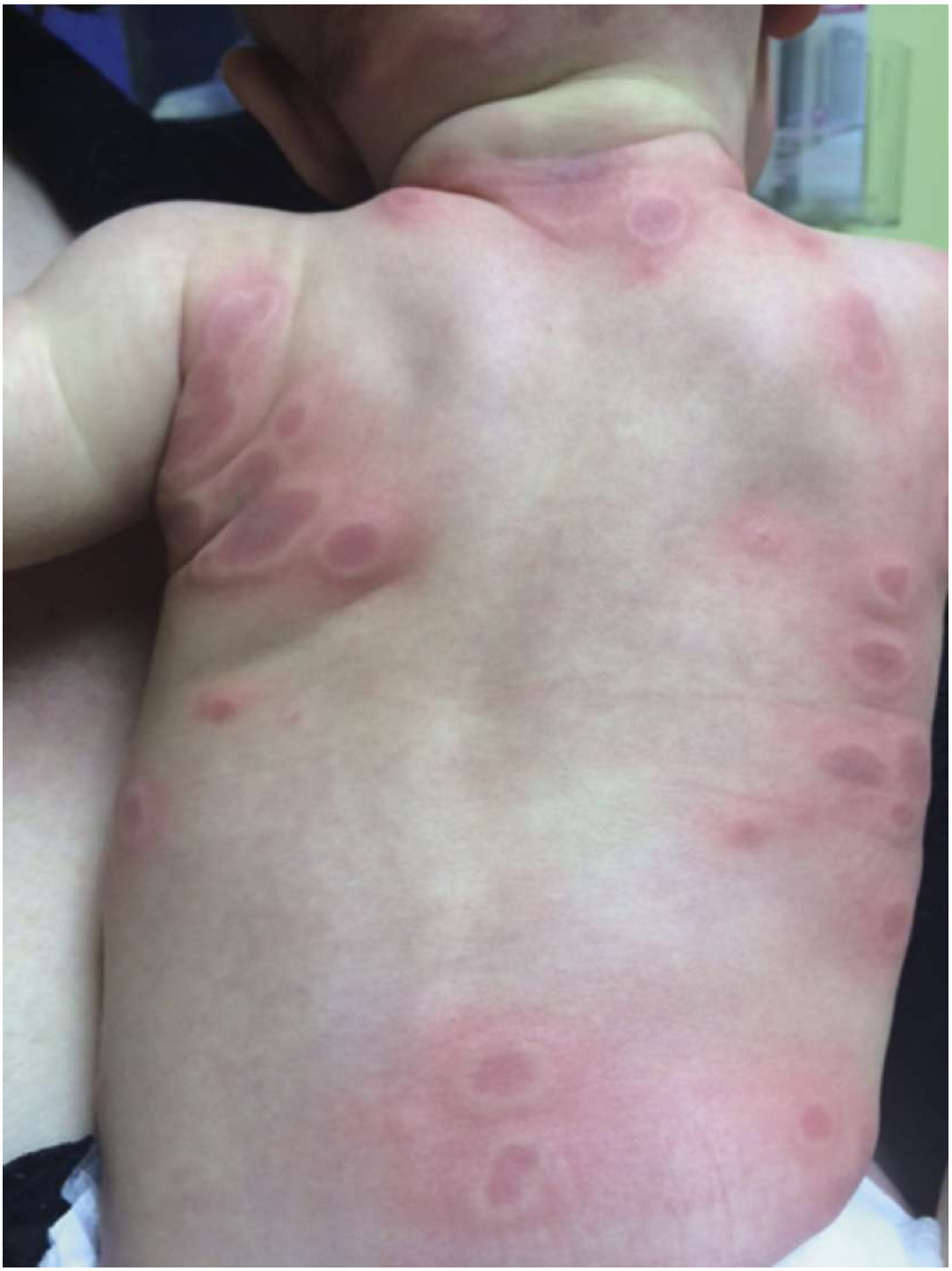

FIG. 12.1.11 Urticaria can often form annular or polycyclic patterns and be mistaken for erythema multiforme. However, the lesions are migratory and do not have the central vesiculation of target lesions.

Urticaria usually occurs in otherwise well children as an isolated finding. The most common trigger for acute urticaria is a viral infection, but it may be triggered by any environmental or food allergen or by prescription and non-

prescription drugs. In most cases of short duration the urticaria resolves rapidly, and the cause cannot be determined. Occasionally, the parents will strongly suspect some food or environmental agent that the child contacted before the onset of the urticaria.

Urticarial drug reactions in children are often associated with penicillin group antibiotics, cefaclor, other antibiotics, aspirin, other non-steroidal anti-inflammatory drugs and latex. Latex allergy is most common in children with recurrent exposure to latex such as repeated catheterisations or surgery and may present as local urticaria, oedema or anaphylaxis. Banana, avocado and kiwifruit can cross-react with latex and after ingestion, children may present with itch, oedema, urticaria and wheezing.

Urticaria can be precipitated by sunlight, pressure, water, cold, heat and other physical factors.

Urticarial episodes usually resolve over days or weeks and rarely last longer than 6 weeks. Chronic urticaria refers to lesions being present virtually every day for more than 6 weeks. Rarely, this can last for many years. Urticaria may be recurrent with individual episodes resolving quickly but occurring many times over weeks, months or years. Recurrent or chronic urticaria is usually of unknown aetiology but rarely may be related to underlying inflammatory conditions such as SLE, juvenile chronic arthritis, other vasculitic diseases, and parasitic infection. Painful urticaria may be a presenting feature of erythropoietic protoporphyria.

In a child who is very unwell with urticaria, consider anaphylaxis or Kawasaki disease. If individual lesions last longer than 2 days or are tender or purpuric, consider investigation for cutaneous vasculitis including Henoch–Schönlein purpura and urticarial vasculitis.

Management

- Urticaria may be the first sign of anaphylaxis. If there is associated angio-oedema (prominent subcutaneous swelling) or wheeze, continued observation and appropriate treatment are required (see Chapter 22.5 Anaphylaxis).
- Investigation is usually not required.
- Identify the cause if possible. Ask about illnesses, medications, recent unusual foods and environmental contacts. Document these clearly.
- Treat the itch with oral antihistamine.

- Oral prednisolone, 1 mg kg^{-1} per day (maximum 50 mg), for a few days may be warranted by the severity of the itch but is usually not needed.
- If any purpura is present or the child is significantly lethargic or less responsive than usual, assess and investigate to exclude meningococcal sepsis and other causes of purpura. Check urine for blood.
- For recurrent or chronic urticaria, look for trigger factors. Consider mast-cell-degranulating drugs (including opiates, aspirin, and other non-steroidal anti-inflammatory drugs), foods, alcohol, animals, parasitic infections, heat, cold, sunlight and physical pressure. Consider investigating with a throat swab (for group A streptococcal carriage), full blood examination (for eosinophilia and anaemia), IgE, antinuclear antibodies, urine culture for bacteraemia, nocturnal check for threadworms and a possible challenge with any suspected agent. Assessment of erythrocyte protoporphyrin is warranted in young infants with sun-induced, painful urticaria.
- For recurrent or chronic urticaria that is unresponsive to conventional H1 antihistamine antagonists, consider adding cimetidine 10–15 mg kg^{-1} (maximum 200 mg) orally every 6 hours.
- For recurrent urticaria, or for chronic urticaria where there is a clear history of regular exacerbations in the severity of the lesions, a diary recording all happenings in the 24 hours before each relapse may be rewarding in identifying the cause.

Serum sickness

Serum sickness (sometimes called ‘serum-sickness-like reactions’) consists of the triad of fever, urticaria and arthralgias. It can occur at any age. Historically, about 50% of serum sickness reactions in Australian children are idiopathic, and 50% have been associated with a recent course of cefaclor. Other medications may also be implicated.

Symptoms begin 5 to 21 days after commencement of the cefaclor and persist for 5 to 10 days. Previous courses of cefaclor may have been taken uneventfully, but more rapid onset can be seen in those children previously exposed. The child presents with an urticarial rash, which can be dramatic in appearance and cause considerable alarm to parents. The lesions may be more fixed than typical urticaria and may bruise or be tender. Arthralgia and joint swelling, possibly with effusion, are observed. Polyarthralgia occurs in up to 80%. Fever,

lymphadenopathy, malaise, nausea, vomiting and abdominal pain are less common. Symptoms resolve fully in a few days. Laboratory investigations are unrewarding and infectious studies negative.

Management (see urticaria above)

- If associated with cefaclor, or another medication, cease the medication and give written information about this to the family and treating doctor.
- Joint symptoms may require analgesia and bed rest.
- Oral antihistamines may provide symptomatic relief. A few days of oral prednisolone, 1 mg kg^{-1} day^{-1} (maximum 50 mg), may be warranted by the severity of the symptoms.
- Hospitalisation and supportive care may occasionally be needed.
- Rechallenge with cefaclor may result in a recurrence of serum sickness, although the risk of recurrence is unknown. A serum-sickness reaction to cefaclor is not a contraindication to having another cephalosporin.

Lupus erythematosus

SLE may present as erythematous, well-demarcated facial lesions, occasionally urticarial or slightly scaly. These lesions occur most commonly in a characteristic butterfly distribution over both cheeks and the base of the nose. In children, the facial rash may be the only manifestation (cutaneous lupus), or there may be systemic involvement. There is usually an epidermal and a dermal component. Therefore the lesion is usually more indurated than a dermatitis and more scaly than an urticaria.

In more widespread disease, patchy lesions may be present over the ears, neck and, less commonly, the limbs. Erythematous macules, petechiae and small infarcts can also be seen around the nail beds and on the tips of the fingers and toes. Erythematous macules may appear on the palms of the hands and soles of the feet. Florid urticaria, blistering and erythema nodosum are uncommonly seen.

Fever, arthralgias and arthritis affecting many joints imply systemic involvement. Lymphadenopathy, anorexia, weight loss, muscle weakness, pulmonary disease (especially pleuritis), cardiac disease (myocarditis and coronary artery disease), nephritis and a wide range of neuropsychiatric symptoms can be seen in childhood SLE.

If lupus is suspected, a detailed history and examination should look for systemic organ involvement, and investigations should include antinuclear and anti-double-stranded DNA antibodies, erythrocyte sedimentation rate, full blood count, renal assessment and/or brain imaging as indicated. Localised cutaneous lesions generally respond well to moisturiser, sun protection and potent topical corticosteroid preparations. Systemic disease requires oral corticosteroid therapy initially. Regular follow-up and monitoring for renal and other organ involvement are essential in all cases.

Neonatal lupus erythematosus (see Chapter 3.3 Neonatal dermatology)

Dermatomyositis

Childhood dermatomyositis affects skin and muscle. Children may present with skin changes years before any muscle involvement or may present with acute or subacute development of fever, rash, pain and weakness. The earliest features in many children are an erythematous, sometimes violaceous, rash on the eyelids, periorbital oedema, a malar erythematous rash, and erythematous papules over the extensor surfaces of the joints of the hands. In more severe cases, there may be widespread desquamation and ulceration around the eyes, fingers and skin folds.

Muscle weakness may not have been noticed at presentation but may be present on testing. Investigations should include erythrocyte sedimentation rate, full blood count and creatine kinase. Magnetic resonance imaging is used to assess the presence and extent of muscle inflammation. Therapy involves high-dose oral corticosteroid therapy initially, maintenance immunosuppression for a couple of years, and regular follow-up.

Juvenile chronic arthritis

An erythematous maculopapular rash is often seen in the systemic form of juvenile chronic arthritis. This rash usually has a salmon pink colour and tends to come and go, being particularly evident at the time when the fever is at its height. It may be urticarial.

Erythema nodosum

Erythema nodosum can occur at any age and presents with the fairly abrupt onset of painful and tender subcutaneous erythematous lesions up to 5 cm in diameter, mainly on the anterior lower legs. The arms, soles and trunk may be affected. Malaise, fever, and arthralgia may be present. Histologically there is a septal panniculitis.

In about 50% of children, erythema nodosum occurs in association with another condition, either as the presenting feature or as part of the evolution of an already diagnosed disease. Causes include chronic streptococcal disease, tuberculosis at any site, inflammatory bowel disease, chronic gastrointestinal infections, sarcoidosis, *Mycoplasma* infection, lymphoma, secondary syphilis, deep fungal infections and the oral contraceptive pill.

Lesions pass through colour changes similar to aging bruises. Resolution occurs in 3 to 6 weeks in most cases. Chronic or recurrent erythema nodosum can persist for months or years, particularly if an underlying cause has not been removed.

Management

- Search for any underlying cause.
- Investigations may include throat swab, full blood examination, erythrocyte sedimentation rate, serology for group A *Streptococcus*, *Mycoplasma* and Epstein–Barr virus, stool culture, Mantoux testing and chest X-ray.
- Depending on the severity of symptoms, bed rest or limitation of activities for a few days may be required.
- Non-steroidal anti-inflammatory treatment will help settle inflammation and pain. The use of oral prednisolone is controversial.
- In troublesome, prolonged or recurrent cases, potassium iodide (10% solution, 2.5 mL = 250 mg three times daily for older children, taken with milk or juice for 2 weeks) is effective.[5]

Necrobiosis lipoidica

This presents as large, irregularly shaped, red-yellow patches on the lower legs, usually in adolescents. With time, the central area may become atrophic or sclerosed. Necrobiosis lipoidica may be the first sign of diabetes in childhood, and a fasting serum glucose should be arranged with follow-up.

Palmoplantar hidradenitis

Palmoplantar hidradenitis (previously known as childhood neutrophilic eccrine hidradenitis or plantar erythema nodosum) occurs in otherwise well children, typically aged 2 to 14 years, during spring or autumn. There may be a history of considerable physical exertion in the days before the lesions appear, often with exposure to cold or water. Erythematous, dusky tender lesions appear on the soles (Fig. 12.1.12) and sometimes on the hands. *Pseudomonas* infection within the sweat glands may be the cause in some or most cases. Antibiotics are not needed, and spontaneous resolution occurs within 3 weeks. If you are confident of the diagnosis, no investigation is necessary.

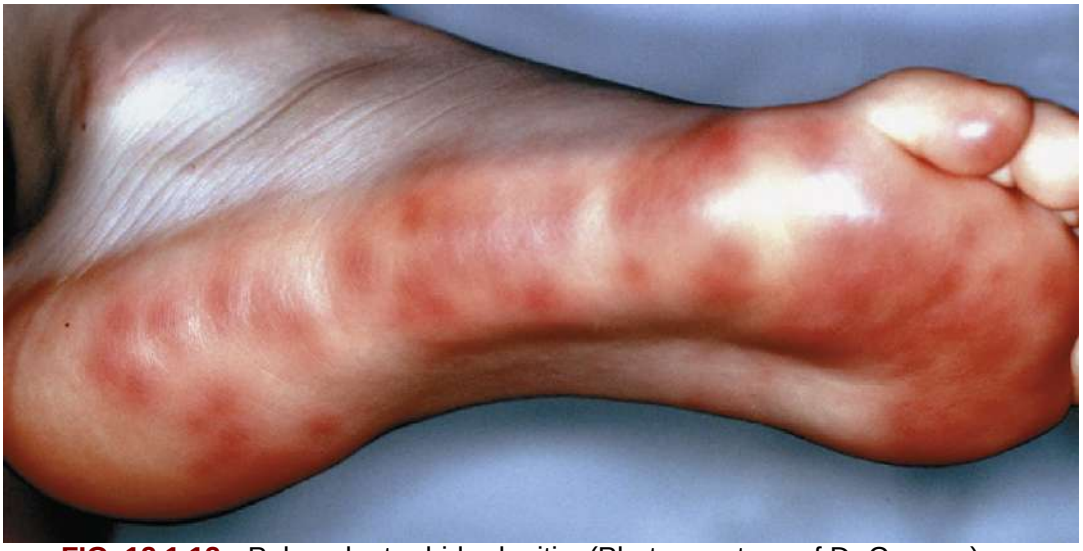

FIG. 12.1.12 Palmoplantar hidradenitis. (Photo courtesy of Dr Cremer.)

Pernio (chilblains)

Pernio usually presents after exposure to cold, such as playing an outdoor sport in winter. Tender red-purple swellings occur on the fingers and/or toes. Initially there is no epidermal change, but lesions can persist for weeks, and dusky scaling can develop. Symptoms are usually mild. Most children presenting with pernio are otherwise well and do not routinely need investigation. In persistent and troublesome cases, baseline investigations may include anti-Ro, antinuclear and anti-phospholipid antibodies, rheumatoid factor and cryofibrinogens. Recurrences are common. Manage by avoiding cold exposure to hands and using

adequate clothing to stay systemically warm.

Spider telangiectasia

A spider naevus presents as a tiny red macule or papule with a surrounding network of fine telangiectasias, best appreciated by compressing the lesion to make it disappear then watching it reappear in the characteristic centripetal pattern, filling from the centre. These appear during mid-childhood in about 50% of children, typically on the face, hands or arms. Most resolve within a few years. Investigation is not needed. If the cosmetic concern is severe, removal by electrodessication or laser can be arranged.

Multiple spider naevi are of no medical significance in healthy children. Multiple telangiectatic vessels of non-spider type may raise suspicion of hereditary haemorrhagic telangiectasia or a photosensitivity syndrome.

Other erythematous rashes

Many conditions that present with red, scaly or purpuric rashes may appear erythematous initially, including guttate psoriasis, pityriasis rosea and Henoch–Schönlein purpura. Most of the lesions may be erythematous, but sometimes a few may reveal the more typical features of the underlying condition, such as scale or purpura.

Many haemangiomas (vascular tumours) will initially present as erythematous macules. This possibility should be borne in mind in any infant presenting with an erythematous macule on the face. Early review may show some progression of the lesion. If so, urgent referral for treatment may minimise potentially severe long-term problems from florid facial haemangiomas (see p. 348).

Purpuric rashes

Purpuric rashes in childhood may be due to vascular dysfunction, coagulation disorder or a low platelet count. Fever may or may not be present. Purpuric rashes are associated with several life-threatening diseases and require urgent assessment.

Fever and petechiae

Petechiae are pinpoint, non-blanching spots, less than 2 mm diameter. There are

many causes of fever and petechiae. These include:

- viral infections including enteroviruses and influenza
- *Neisseria meningitidis* (meningococcal) disease
- other causes of bacteraemia including *Streptococcus pneumoniae* and *Haemophilus influenzae*
- other diseases including Henoch–Schönlein purpura, idiopathic thrombocytopenic purpura and leukaemia
- illness associated with vomiting or coughing causing petechiae around the head and neck.

The majority of children with fever and petechiae do not have a cause identified – they are presumably due to viral infections. Few children with fever and petechiae (<5%) will have meningococcal disease. Recognition and early treatment of these children with meningococcal disease are paramount. Clinical signs and laboratory investigations will help determine those who should be treated for suspected meningococcal disease. All children with fever and petechiae should be reviewed by a senior doctor.

Fever and petechiae in an unwell child (including meningococcal sepsis)

If a child with fever and petechiae meets any of the following criteria, he/she should be presumed to have meningococcal sepsis until proven otherwise and treated accordingly (see meningococcal and other septicaemia below):

- Impaired conscious state (lack of alertness, lethargy, irritability)
- Abnormal signs (tachycardia, tachypnoea, desaturation in air, widened pulse pressure)
- Poor perfusion (cold extremities)
- Any purpuric lesions greater than 2 mm (unless the clinical picture is suggestive of Henoch–Schönlein purpura, see below).

Fever and petechiae in a well child

If a child with fever and petechiae is well and does not meet any of the criteria for meningococcal sepsis in the preceding paragraph, the petechiae may be due

to an obvious mechanical cause. These children do not require investigation and may be discharged with review planned within 12–24 hours. Mechanical causes of petechiae include:

- coughing or vomiting leading to petechiae around the head and neck
- local physical pressure such as a tight tourniquet or being held tightly for procedures
- the practice seen in some ethnic groups of treating a febrile child by rubbing or suctioning the skin with a variety of implements, producing bizarre circular and linear patterns of petechiae.

If no mechanical cause is identified in a well child with fever and petechiae, the child is likely to have a viral infection and should be managed as follows:

- Investigate with a full blood count, and consider a C-reactive protein and blood cultures
- Observe for 4 hours
- If the initial test results are normal and there is no clinical deterioration or progression of the rash over the 4 hours, discharge with review the next day.

Children who have received antibiotics prior to presentation can be managed the same way, but the possibility of partially treated meningitis or sepsis may lead to a lower threshold for admission or early review.

Meningococcal sepsis and other septicaemia

Septicaemia with purpura is usually due to meningococcal disease. There may be an upper respiratory prodrome followed within hours by high fever, malaise and headache. Skin lesions may begin as tender erythematous macules or as petechiae on the skin and mucous membranes. Lesions develop purpuric centres, and large haemorrhagic areas may form. Purpura may also be due to sepsis with *Haemophilus influenzae*, group A streptococci, *Staphylococcus aureus* and some gram-negative organisms. Infective endocarditis, typhus, typhoid fever and certain viral haemorrhagic fevers may cause purpuric lesions.

Early management (a detailed protocol for meningococcal management can be accessed online[6]):

- Give oxygen
- Gain IV or intraosseous access
- Immediate investigations (if blood can be obtained without delay) including blood culture, blood smear, blood PCR, full blood count/differential, glucose, urea and electrolytes, and clotting, if appropriate
- Administer cefotaxime 50 mg kg^{-1} (max 2 g) IV 6H ***or*** ceftriaxone 100 mg kg^{-1} (max 2 g) IV daily
- Meningococcaemia is often associated with hypovolaemia: give 20 mL kg^{-1} of normal saline. More fluid will often be needed to improve blood pressure and peripheral perfusion (40 mL kg^{-1} in the first hour is common)
- Isolate cases (if possible) until they have had >12 hours antibiotic treatment
- Subsequent investigations include throat swab, lumbar puncture (unless contraindicated). Urinary or CSF 'rapid antigen' testing is not recommended because of poor sensitivity and specificity.

Vasculitis and Henoch–Schönlein purpura

Vasculitis is a reaction pattern that presents as non-itchy, painless macules, papules or urticarial lesions with purpuric centres. There are many causes of vasculitis including infections (streptococcal, hepatitis viruses and others), autoimmune diseases (SLE, Behçet disease), allergy (drugs) or idiopathic.

Henoch–Schönlein purpura is an IgA-mediated condition and is the commonest cause of vasculitis in children. It can occur at any age but is most common between the ages of 2 and 10 years. There is sometimes a preceding streptococcal or non-specific viral illness. Over a couple of days, lesions occur in a symmetrical distribution, mainly on the buttocks and lower legs and occasionally on the genitalia, arms and elsewhere (Fig. 12.1.13). In many children, most lesions are purpuric, ranging from pinpoint petechiae to 2 cm in diameter. In other children, only a few purpuric areas may be noted amongst many erythematous or urticarial lesions. There may be associated abdominal pain, arthralgia, arthritis or haematuria. Renal involvement leading to chronic renal failure is rare but can occur irrespective of the severity of the rash and other symptoms and may be delayed until weeks or months after the onset of the illness.

A child with Henoch–Schönlein purpura may warrant a full blood examination to exclude thrombocytopenia. Check blood pressure and urine for blood or protein. Bed rest and analgesia may be needed if discomfort is considerable. Oral prednisolone may reduce the duration of abdominal pain and may reduce the likelihood of renal problems. Severe renal involvement may require immunosuppression, plasmapheresis or hypertensive therapy, but this is rare. In children with no renal involvement at presentation, blood pressure and urinalysis should be done weekly for 1 month, then bimonthly for 6 months. Children who have not developed urinary abnormalities within 6 months do not require further regular follow-up.

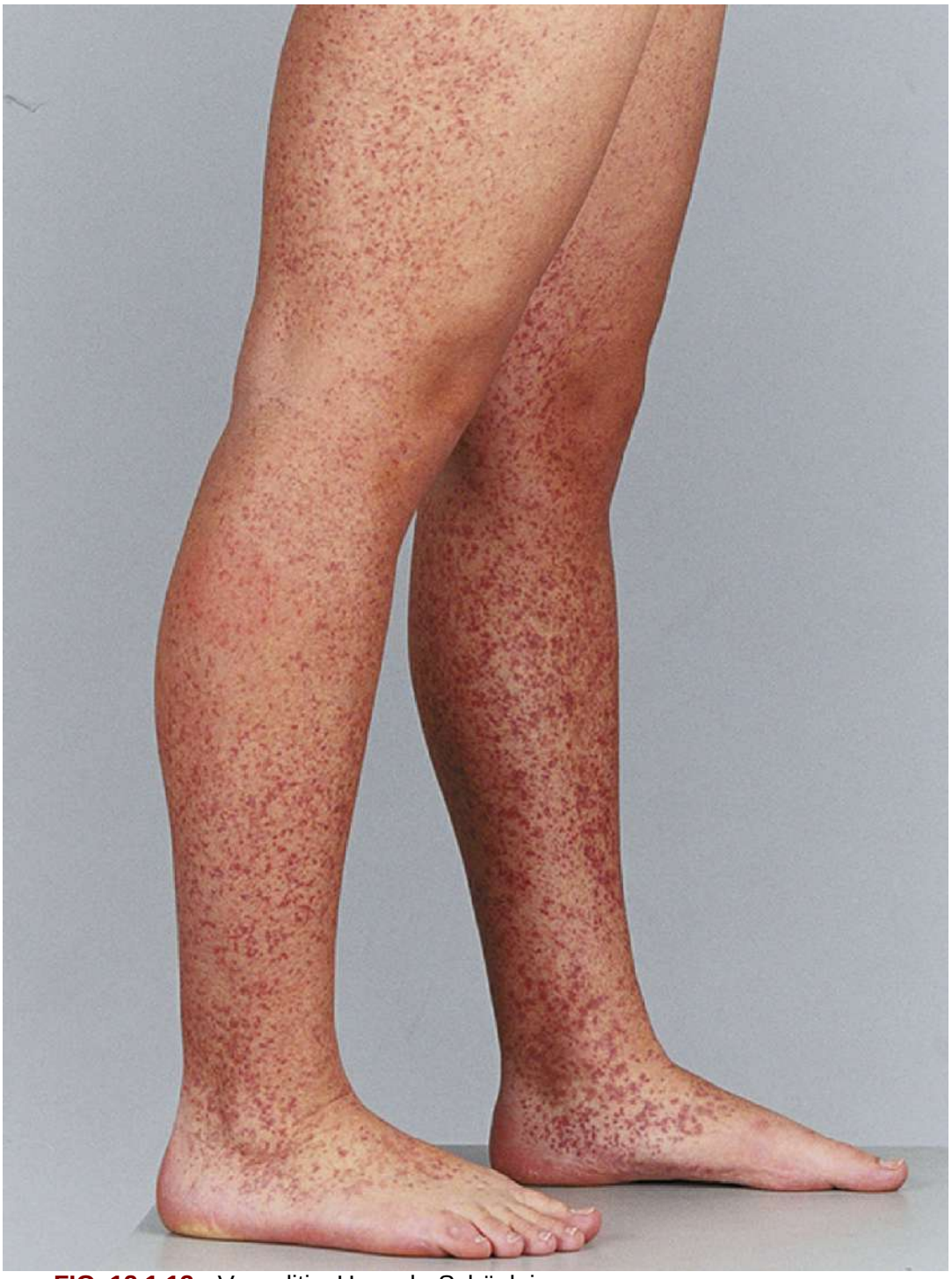

FIG. 12.1.13 Vasculitis. Henoch–Schönlein purpura.

Thrombocytopenic purpura

The most common cause of purpura due to thrombocytopenia is idiopathic

thrombocytopenic purpura (ITP). This is an acquired thrombocytopenia due to immune-mediated shortened circulating platelet survival in the absence of other disturbances of haemostasis or coagulation. Most children present with bruising and petechiae alone. Oral bleeding, epistaxis, rectal bleeding or haematuria are less common. There is often a history of a recent viral infection. The child is otherwise well. More serious causes, such as leukaemia or aplastic anaemia, should be excluded. Full blood examination will be normal apart from a low platelet count.

Thrombocytopenia may be drug induced (e.g. secondary to chloramphenicol, antithyroid medications).

Leukaemia

Leukaemia should be suspected in a child with generalised petechiae or purpura in the absence of trauma. Features may include tiredness, pallor, limb pain, malaise and gum hypertrophy. Thrombocytopenia is usually present. An urgent full blood examination should be obtained (see Chapter 11.6 Acute leukaemia).

Two subtypes of acute myeloid leukaemia typically present with skin lesions. Acute monocytic leukaemia may present with skin infiltrates and gum hypertrophy. Acute promyelocytic leukaemia often presents with a purpuric rash anywhere on the body in association with disseminated intravascular coagulation.

Coagulation disorders

Extensive purpuric lesions in a well child without a history of significant trauma may be due to an underlying coagulation disorder. This may be inherited or acquired. There may be a history of joint pain or swelling or bleeding from other sites.

Child abuse

Twisting, compression, pinching and hitting can all cause petechial or purpuric lesions. Look for bruises of bizarre shapes and different ages or evidence of fractures or other injuries. Assess the child’s affect and relationship with carers. A complete examination is mandatory. If child abuse is suspected, admission or immediate referral to a paediatrician/child protection service may be needed for

complete assessment and further investigations (see Chapter 18.2 Child abuse).

Artefactual purpura

Older children sometimes present with self-induced purpuric lesions. These are often in bizarre, non-physiological shapes, sometimes with obvious marks of finger trauma. They occur in exposed areas. Lesions may have recurred for months or longer in an otherwise well and generally unconcerned child or adolescent. The history typically has inappropriate or contradictory details and a lack of concern about the lesions. The diagnosis and management of artefactual disease in children may be difficult and require confidence in interpretation of the skin findings and ongoing paediatric and psychological follow-up of the child and family. Occasionally, admission to hospital may be required.

Papular-purpuric gloves and socks syndrome

Papular-purpuric gloves and socks syndrome is caused by parvovirus B19 and probably by several other common viruses. Adolescents develop vague symptoms of fever, fatigue and pains. Redness and swelling of the palms and soles progress rapidly to petechial and purpuric lesions that do not extend onto other body parts. There may be oral vesicles or erosions. The adolescent remains generally well, and the eruption clears in 2 weeks. Interestingly, unlike erythema infectiosum, adolescents with papular-purpuric gloves and socks syndrome are thought to be infective during the period of rash.

Dusky purple nodules on hands and feet

For tender red-purple areas (not usually purpuric) on hands and feet in otherwise well children, consider pernio (see p. 346) and palmoplantar hidradenitis (see p. 346).

Chronic pigmented purpura

Chronic petechiae in childhood are usually due to chronic pigmented purpura. This typically presents as several 2–3 cm patches of petechiae. In each patch, both new and old petechiae can be seen giving red, purple and brown dots (like cayenne pepper). Patches may be subtle, the child is otherwise well, the condition is benign, but lesions recur for months or years.

Other causes of childhood purpura

Unusual patterns or presentations of purpura in childhood may be associated with vasculitic diseases, glucocorticoid excess, any cause of abnormal skin elasticity and scurvy. In children with scurvy, irritability, bone pain, gum sponginess and bleeding may be present. Wrist X-rays are diagnostic.

Langerhans cell histiocytosis may present with persistent anogenital or scalp petechiae in infants or with widespread, itchy, haemorrhagic papules, including on the hands and feet (see p. 336).

Neonatal purpura (see Chapter 3.3 Neonatal dermatology)

Vascular tumours – haemangiomas and haemangioma variants

Haemangiomas of infancy

Haemangiomas are tumours of vascular endothelial cells. While haemangiomas may be present at birth, most appear in the first days or weeks of life. The appearance depends on whether the haemangioma involves the skin. If the skin is involved, the first sign is often a patch of pallor, followed by the development of an erythematous macule. Over a few weeks, this may grow and thicken into a firm, partly compressible, well defined, crimson or purple nodule (the so-called ‘strawberry haemangioma’). There may be a substantial subcutaneous mass. Haemangiomas that do not involve the skin are often first noticed as firm, blue or skin-coloured swellings at a few months of age. Haemangiomas can occur anywhere on the body and may involve internal organs such as the liver or trachea.

Haemangiomas vary in size, anywhere from tiny 1 mm lesions or extensive lesions involving large areas of the body. Growth may be slow but can be rapid and frightening, even with intensive treatment.

Haemangiomas usually grow for 3–9 months and then slowly involute over the next 2–10 years. Involution is first noticed as a decrease in the intensity of the surface crimson colour with development of small islands of greyish skin. The haemangioma becomes softer and gradually shrinks in size. Half resolve by age 5 years and 90% by age 9 years. Resolution may not be complete, and many haemangiomas leave permanent changes such as telangiectatia, fibro-fatty tissue, atrophy or redundant skin. Some haemangiomas show little resolution.

Most haemangiomas never cause problems and are best left alone and allowed to involute spontaneously. However, haemangiomas can rapidly lead to problems such as extreme disfigurement, ulceration, blindness, destruction of cartilage, respiratory obstruction, coagulation failure or death. Urgent assessment by a clinician experienced in this field is needed in the following circumstances:

Association with stridor

Stridor, especially increasing stridor of recent onset in a child of a few months of age, may be due to a laryngeal or tracheal haemangioma. This risk is greatest in children with a superficial haemangioma involving the beard area. About half of all children with an airway haemangioma will have a visible superficial haemangioma.

Eye involvement

Even quite small haemangiomas on the eyelid or adjacent to the globe of the eye can cause impaired vision on that side, either by directly obstructing the visual axis or more commonly by pressing on the globe to cause astigmatism. In both cases, amblyopia may result and may lead to permanent blindness if not treated.

Involvement of facial structures

Haemangiomas may deform structures such as the lip, ear cartilage, nasal cartilage, dentition or jaw growth. This deformity will remain after resolution of the haemangioma. Deforming lesions require aggressive treatment.

Ulceration

Ulceration is more common in haemangiomas at sites exposed to trauma, including areas of friction, anogenital lesions and lesions involving the lips. Ulcers may be very painful and can spread within days to give necrotic lesions several centimetres in size. There may be full tissue loss of lips, nose and eyelids with permanent disfigurement and loss of function.

Macular facial lesions

A macular (flat) capillary lesion on the face of a neonate is likely to be a capillary malformation, not a haemangioma, and does not require urgent treatment. However, it may be the first sign of an extensive facial haemangioma. This can commence as a large macular capillary lesion that is indistinguishable from a capillary malformation until thickening and rapid growth occur (Fig.

12.1.14). These haemangiomas can cause marked disfigurement, and treatment should be begun early, within the first few weeks. If you are suspicious about a large macular capillary lesion in a neonate (e.g. onset after birth, changing colour, any sign of thickening), monitoring and/or referral should be done with an awareness of the possible urgency of treatment.

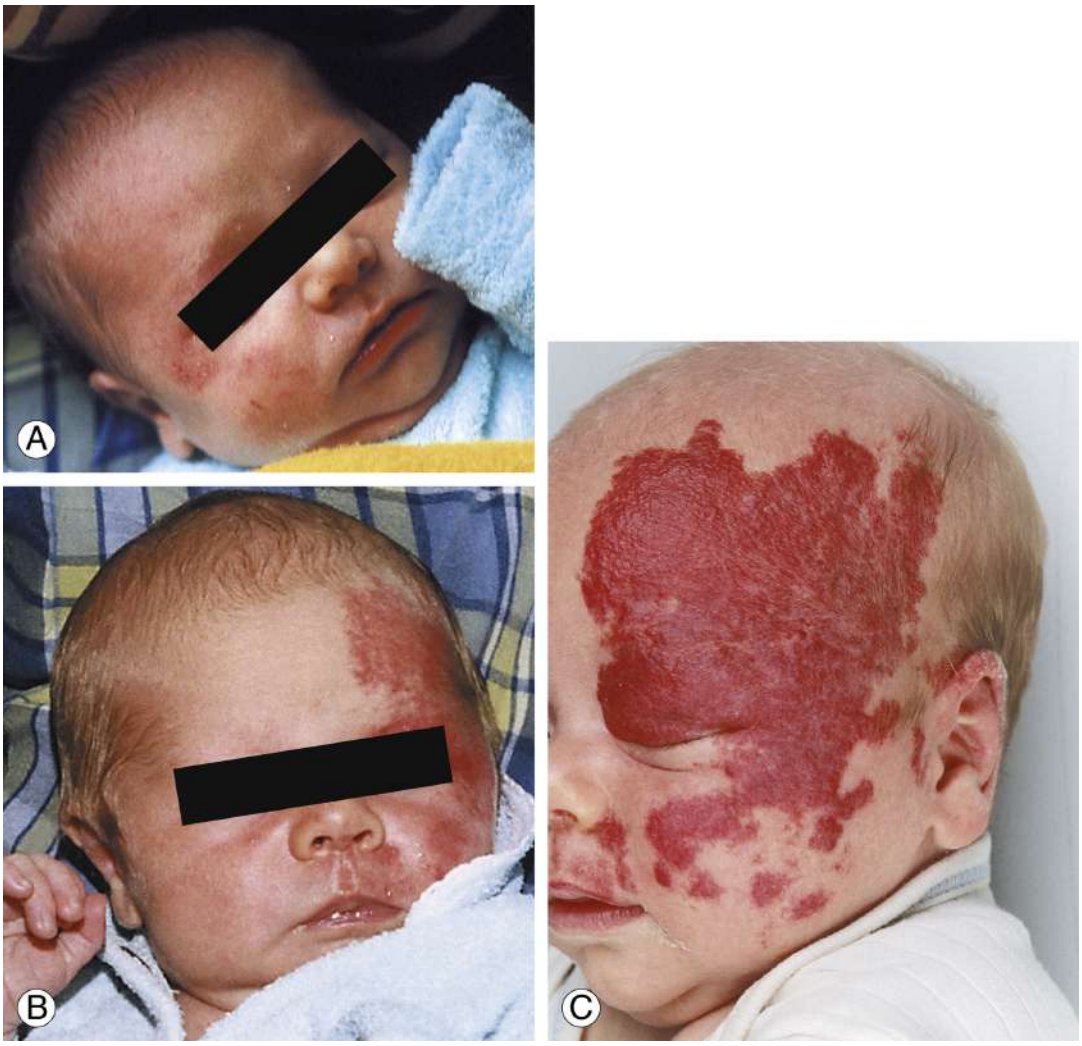

FIG. 12.1.14 Untreated segmental facial haemangioma. (A) At 1 week; (B) at 2 weeks (easily misinterpreted as a capillary malformation); and (C) at 6 weeks, by which time thickening has occurred. Urgent assessment for possible laser and oral propranolol treatment can minimise morbidity.

Sudden onset of swelling and bruising in a large vascular lesion

This may indicate the development of a consumptive coagulopathy in another type of infantile vascular tumour – see Kaposiform

haemangioendothelioma/tufted angioma (see p. 349).

Multiple haemangiomas (diffuse neonatal haemangiomatosis)

Infants with five or more cutaneous haemangiomas have a higher risk of haemangiomas at other sites, including liver.

Cardiac failure with a hepatic mass

Infants who present with cardiac failure and are found to have one or more highly vascular hepatic masses on ultrasound are likely to have either a single large congenital haemangioma or multiple infantile haemangiomas. These are separate types of lesion. Both can present with high-flow cardiac failure, but the treatment of the haemangioma is different in the two cases. Treatment of congenital haemangiomas is supportive as they do not respond to propranolol (see below).

Management

- Most parents just need education and reassurance about the inherently benign nature of these lesions.
- Treatment with timolol maleate 0.5% gel forming drops, one drop, applied to the lesion twice a day may benefit some small flat superficial haemangiomas. Topical therapy is less effective and should not replace systemic therapy (see below) in patients who have a clear indication for treatment. Systemic absorption of topical timolol may be significant with ulcerated or large lesions, especially in premature or small infants.
- If complications of the haemangioma, as described above, are considered likely, a thriving healthy infant can be treated with oral propranolol (or other appropriate beta-blocker) for 3–24 months. A typical regimen uses 5 mg mL syrup, giving 0.5 mg kg^{-1} bd for several days and then increasing to 1 mg kg^{-1} bd. Early treatment will prevent or minimise complications in most cases.
- Infants who are premature, small, unwell, or failing to thrive are at an increased risk of complications from propranolol, such as hypoglycaemia. Treatment dosing regimens in these infants should be lower with more gradual dose increasing and with closer clinical monitoring.
- Most haemangiomas show immediate cessation of growth on starting

propranolol and some shrinking with weeks. Not all haemangiomas respond as well. Intralesional corticosteroids, IV vincristine, vascular laser or surgery may all be warranted in individual patients.

- Any infant with a large facial or head haemangioma has a greatly increased risk of posterior fossa anomalies, intracranial arterial dysplasia, eye abnormalities and aortic anomalies (PHACES syndrome). Consider echocardiogram, magnetic resonance imaging/angiography (MRI/MRA) of the head and ophthalmology assessment. If feasible, brain imaging should be done before commencement of propranolol because of the theoretical very small risk of propranolol causing stroke if the brain arterial anatomy is compromised.
- Any infant with a large lumbosacral or anogenital haemangioma or other vascular malformation should be checked for genitourinary and spinal abnormalities. Consider a renal ultrasound. To image the spine prior to 3 months of age, an ultrasound examination is generally satisfactory. In older children, a MRI examination is warranted.
- Most haemangiomas treated with propranolol do not clear completely. Residual redness, atrophy or other change is commonly still visible in later childhood.

Rapidly involuting congenital haemangioma

Rapidly involuting congenital haemangiomas are a distinct type of haemangioma. They are not infantile haemangiomas. They are present at birth as raised violaceous vascular tumours, often many centimetres across. They can be associated with high-flow cardiac failure and can be mistaken both on clinical and radiological assessment for arteriovenous malformations or vascular sarcomas. The distinction is important because they resolve within 12 to 18 months without treatment. They do not respond to propranolol therapy.

Kaposiform haemangioendothelioma/tufted angioma

These tumours may be present at birth. Growth can be rapid and extend over much of a limb and/or trunk. The texture is often firmer than felt with typical infantile haemangiomas. Kaposiform haemangioendothelioma and tufted

angioma are associated with platelet trapping and a consumptive coagulopathy (Kasabach–Merritt syndrome). This has a significant mortality if untreated. A platelet-trapping crisis causes rapid swelling of the lesion, with the lesion appearing bruised and becoming hard and tender. Assess platelet count, clotting profile and fibrinogen level. Oral sirolimus and/or high-dose oral corticosteroids may be effective. If not, vincristine for many months may help achieve and maintain control and reduction in tumour size.

Pyogenic granuloma

Pyogenic granulomas are benign acquired vascular tumours that often present to EDs with a history of bleeding. They usually occur after infancy, on the face and neck, hands, or elsewhere on the body. There may be a history of preceding trivial trauma to that site. Over several days, a dark red/bluish papule appears and grows to 10 mm in height. The surface may be moist, scaly or warty. Bleeding is common and may be recurrent.

Management

Bleeding will stop with the application of pressure. Topical imiquimod applied three times a week usually results in resolution in about 3–4 weeks. Lesions that persist at 4 weeks require surgical removal for histological confirmation and definitive treatment.

Vascular malformations

Vascular malformations are developmental lesions composed of dilated blood vessels and/or lymphatic channels. They are present at birth but may not become evident until infancy or occasionally later in childhood. Malformations include common capillary malformations on the face (port-wine stain), more extensive capillary lesions, localised venous malformations, venous malformations involving large areas with associated overgrowth and other changes, arteriovenous malformations, lymphatic malformations (e.g. cystic hygroma) or any combination of these.

They are not tumours and do not have a growth phase or resolution phase as haemangiomas do. They are generally static lesions. However, changes can occur over time, leading to increasing problems. Particularly at puberty, there may be growth of the malformation leading to its appearance for the first time or

leading to problems such as pain in a previously asymptomatic lesion.

The appearance depends on the nature and site of the malformation. Purely capillary malformations appear as red macules on the skin. They blanch partially or fully. Venous involvement manifests as dilated veins or soft compressible bluish lumps that empty when elevated. Phleboliths are common and may be palpable. Significant arterial involvement may allow a thrill to be felt on examination. Lymphatic lesions give diffuse swelling of a limb or soft subcutaneous masses. Lymphatic involvement on the skin surface results in a mass of firm warty 2 mm papules that may be clear but often are dark blue or black because of haemorrhage into the lesions.

Vascular malformations can be associated with many chronic and acute problems that can precipitate attendance at an ED. Chronic problems include concerns about the appearance, psychological problems, social isolation, pain, deformity, dental problems, overgrowth, leg length discrepancy, arthritis and bone erosion. Acute problems may be related to any of these and also include pain, haemorrhage, infection, thrombosis and pulmonary emboli.

Neonatal vascular malformations

A neonate with a substantial vascular malformation should be examined thoroughly for associated malformations, for high-flow cardiac compromise and for other abnormalities in any system. Investigation includes ultrasound and radiological imaging.

Facial capillary malformations (‘port wine stain’)

Facial capillary malformations are usually present at birth and persist throughout childhood, becoming darker and thicker during adolescence. Treatment with vascular laser in early childhood leads to clearing of much of the lesion in most children.

Children with vascular malformations around the eye should be screened for glaucoma and other eye abnormalities. Children with capillary malformations involving the forehead and upper eyelid have a 10% risk of intracranial involvement and a smaller risk of developing epilepsy, strokes, hemiplegia or mental retardation (Sturge–Weber syndrome).

Beware of any facial ‘capillary malformation’ that appears after birth or becomes darker or thicker in the first weeks of life. This may actually be a

haemangioma requiring urgent treatment (see macular facial lesions, p. 348).

High-flow lesions

Arteriovenous malformations are dangerous. Any vascular lesion with a thrill should be presumed to be an arteriovenous malformation. They can present in many ways to the ED – as a new swelling, an area of skin necrosis, skin ulceration, bleeding or with the sequelae of internal bleeding, including intracranial bleeding. They can expand and erode surrounding structures. If a high-flow lesion is suspected, Doppler ultrasound should be arranged to confirm this. Expert follow-up is essential.

Pain, swelling

Chronic discomfort and pain are common with large venous malformations, particularly those involving the legs. Pain is worse after standing for prolonged periods. Sudden onset or worsening of pain is also common. In venous malformations, it may arise from localised microthrombi and microinfarction. In venous and lymphatic malformations, it may be due to secondary infection. Sudden pain may be accompanied by a sudden increase in swelling and may lead to a rapid compromise of a body function, depending on the site. Sudden, severe pain in an adolescent, with or without fever, may be the presenting feature of Fabry disease (see below). Complex regional pain syndrome should also be considered.

Bleeding, coagulopathy

Large venous malformations are associated with chronic consumption of clotting factors, low fibrinogen, somewhat reduced platelet count and bleeding. (This is not Kasabach–Merritt syndrome, which is associated kaposiform haemangiomendothelioma, see p. 349.)

Chest pain or shortness of breath

A child with a venous malformation who presents with chest pain or shortness of breath may be having chronic and/or acute pulmonary emboli. Investigate as indicated by the clinical situation.

Intestinal bleeding, anaemia

Multiple cutaneous and visceral vascular malformations may cause intestinal bleeding and chroni anaemia (previously known as ‘blue rubber bleb syndrome’). In hereditary haemorrhagic telangiectasia, telangiectasias usually appear on the face, mouth and nose. Nose bleeds become frequent in late childhood, and gastrointestinal bleeding occurs in adult life.

Bladder or bowel dysfunction in older children

Congenital lesions over the lumbosacral area may be associated with occult spinal abnormalities such as a tethered cord. These spinal anomalies may not cause problems until later in childhood when they can present insidiously with irreversible bladder, bowel or limb dysfunction. These problems can be prevented by earlier screening with MRI and surgical correction. Congenital lesions that have been associated with underlying spinal problems include haemangiomas, capillary malformations, lipomas, dimples, sinuses and hairy patches.

Leg length discrepancy

Many vascular malformations can be associated with uneven leg lengths. This can occur even when the vascular malformation does not affect either leg. Leg length discrepancy usually presents as an asymptomatic finding but can be noticed as a limp or as a secondary problem including scoliosis, back pain, joint pain and headache. The difference in leg lengths can be progressive and can reach more than 10 cm. Treatment by specialist centres is very effective at reducing the adult-limb length difference to insignificant levels provided that detection and treatment planning begin in childhood.

Multiple telangiectatic vessels

Multiple telangiectatic vessels of non-spider type may raise suspicion of hereditary haemorrhagic telangiectasia or some photosensitivity syndromes. Multiple spider naevi do not require investigation.

Fabry disease

Fabry disease is X linked and primarily affects males. Females can be variably affected. Angiokeratomas (flat or raised, slightly warty, red/purple lesions) appear in mid-childhood on the lower trunk, pelvis and thighs. Tiny vascular lesions may be seen on the lips or in the mouth. Corneal opacities are invariably present. Children or adolescents may present to the ED with paraesthesia of the hands and feet or with sudden severe pain involving the limbs. Renal, cardiac and central nervous system problems occur later.

Management of vascular malformations

- Management of vascular malformations can be demanding and complex. For example, interventional embolisation of a large venous malformation of a limb may achieve an excellent result but only after perhaps 20 or more general anaesthetics each of a few hours duration.
- Management requires a multidisciplinary approach using expertise from surgical, paediatric, dermatological, radiological and psychological fields.
- Children with lumbosacral vascular malformations should have MRI to exclude a tethered spinal cord.
- Compression stockings may reduce chronic pain in venous malformations. Low-dose aspirin may reduce episodes of pain in venous malformations. Heparin will minimise the coagulopathy from large venous malformations (but does not help the coagulopathy associated with Kasabach–Merritt syndrome seen with some vascular tumours).
- Partial or complete surgical resection may be needed for venous, lymphatic or arteriovenous malformations.
- Orthotic or surgical correction may assist with a leg length discrepancy.

Hyperpigmentation

Diffuse hyperpigmentation

Diffuse hyperpigmentation is rare in children, and investigation to identify the cause is required. Generalised darkening of the skin is often most obvious on the palmar creases, linea alba and areola, on the buccal mucosa and on the sun-exposed areas of the face, neck and extremities. Consider:

- endocrine disease. Addison disease (also thickening of the skin and signs of increased androgen production), Cushing syndrome of pituitary origin (also acne, hirsutism, striae, purpura), exogenous adrenocorticotropic hormone administration, acromegaly (thickened, greasy, more hairy and often with many skin tags) and hyperthyroidism can all cause hyperpigmentation.
- renal failure may cause greying of the skin.
- haemochromatosis. In children, this is usually secondary to transfusions.
- lipoidoses. A yellow-brown darkening of skin, most prominent in sun-exposed areas, can occur in the Niemann–Pick diseases. Look for waxy indurated skin, purpura, hepatosplenomegaly and neurological deterioration. Although a similar colour can be seen in adult-onset Gaucher's disease, it is not a feature of the earlier onset forms.
- pellagra. Hyperpigmentation and erythema on sun-exposed areas, cheilitis, perineal inflammation and diarrhoea may be seen in niacin deficiency.

Localised macular hyperpigmentation – including café-au-lait macules

Pigmentation of the skin can be divided into epidermal and dermal pigment, and these are usually able to be distinguished clinically. Pigment in the epidermis will be brown to black in colour whereas pigment in the dermis, purely because of the depth in the skin, will have a blue to grey shade.

With regards to epidermal pigment, up to 2% of healthy children have one or two well-defined pigmented macules, generally not present at birth but appearing in the early years. In a child with brown-pigmented macular lesions of normal texture, consider:

Neurofibromatosis

Six or more café au lait spots greater than 0.5 cm in diameter are strong evidence for neurofibromatosis. Examine for other features including axillary 'freckling' (usually not seen until mid-childhood), pigmented or thickened skin over plexiform neurofibromas (present in early childhood), iris pigmentation (Lisch nodules), optic tumours, skeletal abnormalities, short stature, skin neurofibromas, hypertension, macrocephaly and learning difficulties. The diagnosis may be uncertain early in life, but full penetrance of neurofibromatosis

is usually seen by 8 years of age. It is appropriate therefore to have children with multiple café au lait macules assessed and followed by clinicians familiar with neurofibromatosis and related syndromes. Regular follow-up is needed, including assessment of intellectual progress and audiological and ophthalmological review. In children under 5 years old, MRI of optic tracts to exclude optic glioma is warranted. Other investigations are not required unless suggested by clinical findings. The risk of myelomonocytic leukaemia in a child with neurofibromatosis is several hundred times higher than in other children. It is still sufficiently low (0.05%) that routine testing is not warranted, but any unexplained hepatomegaly, splenomegaly, lymphadenopathy, pallor or infiltrative skin lesions require investigation. Most children with neurofibromatosis never develop any of the significant medical associated features.

McCune–Albright syndrome

A child with one or more large unilateral brown macules may have McCune–Albright syndrome. The macules will have midline cut-offs and have ‘geographical’ borders. Endocrine or bony abnormalities should be looked for on examination. A bone scan should be performed at 3 years of age, and the child should be monitored for any signs of increased hormonal secretion. Routine endocrine investigations are not warranted in the absence of abnormal clinical signs.

Incontinentia pigmenti

If the earlier phases have occurred *in utero*, hyperpigmented linear streaks may be present at birth.

Peutz–Jeghers syndrome

Small, pigmented macules present on the lips and mucosa from birth are associated with intestinal polyposis. Care must be taken when assessing any episodes of abdominal pain as these children are at higher risk of intussusception and collapse. Intestinal bleeding may be the presenting feature.

Segmental pigmentary disorder (including linear and whorled hyperpigmentation and hypopigmentation of Ito)

Streaks, lines and whorls of hyperpigmentation and/or hypopigmentation may be present from birth. These patterns reflect mosaicism. Usually these children are

otherwise normal, but a wide range of associated malformations in other systems have been reported, particularly eye, teeth, brain and skeletal malformations. These areas of altered pigmentation often will present many months or even years after birth. Small, localised birthmarks do not require additional assessment, but for larger, more extensive involvement, audiological and ophthalmological examination is appropriate. Other investigations are not required unless suggested by clinical findings, but these children should be reviewed until settled in school.

Mastocytosis

Childhood mastocytosis (also known as urticaria pigmentosa) will usually present within the first months of life with one or multiple red-brown patches or plaques due to collections of mast cells in the skin. Rubbing, heat or immunological stimulus can cause these to urticate and even blister. Diagnosis can be made clinically or by skin biopsy. Unlike mastocytosis in adults, disease presenting in the first decade of life is usually benign and not associated with haematological malignancy. First-line treatment is with oral antihistamines if symptomatic.

Presentations with dermal (blue/grey) pigmentation

Post-inflammatory hyperpigmentation

This occurs particularly in dark-skinned people. Many inflammatory skin disorders, particularly those involving the dermo-epidermal junction, may heal leaving diffuse, hyperpigmented macules that can persist for months. Inflammatory conditions to be considered include cutaneous lupus, erythema multiforme, fixed drug eruptions and lichen planus.

Mongolian spots

Mongolian spots are another manifestation of dermal melanocytosis. They are extremely common and can be found on most Asian and dark-skinned babies as poorly circumscribed, blue-black patches. The most common site is on the lower back, but they can occur elsewhere and may be multiple and widespread. They are benign and usually fade over many years. No treatment is required. When at unusual sites, they can be mistaken for bruises.

Naevus of Ota

Naevus of Ota is present at birth in 50% of cases and appears at puberty in most others. It can occur in any racial group and is one manifestation of dermal melanocytosis. It presents as a unilateral, well-demarcated, blue-black patch of skin on the cheek, forehead and periorbital area. The sclera of the associated eye may also be pigmented. Cover-up cosmetics or laser depigmentation has been used to treat the discolouration.

Glaucoma has been reported with naevus of Ota.

Localised raised hyperpigmentation

If local areas of hyperpigmentation are roughened, raised or warty, consider:

- congenital and acquired pigmented naevi (see below)
- genodermatoses, such as epidermal naevi or dyskeratosis congenita. Congenital nail dystrophy, pancytopenia, skeletal and eye anomalies may be present.
- acanthosis nigricans. Rough ‘dirty’ skin on the neck or axillary folds is associated with obesity, polycystic ovary syndrome, other insulin resistance syndromes and hypothyroidism. Early intervention and follow-up are required.

Congenital pigmented naevi

Congenital melanocytic naevi are thickened, sharply defined, tan, dark brown or black birthmarks. They range in size from common small lesions less than 2 cm in diameter to rare giant lesions that may cover entire parts of the body, such as the trunk. Larger lesions may have irregular colour, texture and hairiness with areas of thick redundant skin. Large congenital melanocytic naevi often are associated with many smaller ‘satellite’ lesions elsewhere on the body, some of which appear in the first 2 years of life.

Management (see Chapter 3.3 Neonatal dermatology)

- The risk of malignancy in isolated small congenital melanocytic naevi is very small, and lesions do not require surgical excision. Most melanomas develop in normal skin rather than from pre-existing naevi,

and so decisions about removal are based purely upon the site and potential effects of the child's psychosocial development.

- Giant congenital melanocytic naevi are associated with major psychosocial problems. Early dermabrasion can markedly decrease the colour of the naevus and may be considered. Contact with a plastic surgeon experienced in this area is necessary. Pigment lasers have also been used and may be useful for some lesions.
- A second concern is malignancy. The lifetime risk of melanoma in a giant congenital melanocytic naevus is small (less than 5%). Parental instruction about reporting changes in the naevus will allow early biopsy of changing areas.
- Giant congenital melanocytic naevi of the scalp or spine can be associated with neurocutaneous melanosis and other CNS abnormalities. Most of these abnormalities remain asymptomatic, and screening with MRI is warranted primarily for prognostic information if large axial naevi and/or multiple satellite naevi. Early imaging in the first months of life allows the use of MRI without the need for a general anaesthetic.
- Regular follow-up is needed throughout childhood to monitor the naevi and psychosocial development.

Acquired pigmented naevi

During childhood, most children develop multiple acquired melanocytic naevi. Sun exposure in white children is associated with the development of an increased number of naevi. Acquired melanocytic naevi begin as small, flat, well-demarcated pigmented lesions (junctional melanocytic naevi). A small percentage of junctional melanocytic naevi become raised and dome shaped (compound melanocytic naevi) often with a darker centre. Enlargement and darkening of naevi just before and during puberty are common.

Melanoma is very rare in childhood and adolescence. Children and adolescents continue to acquire new melanocytic naevi until age 20. Therefore, the presence of a new naevus is not in itself suspicious. You do need to be concerned if atypical features are present, particularly in adolescents. Any rapid change over a few weeks leading to an increase in asymmetry, an irregular border, multiple colours especially with non-brown areas within brown areas, and growth beyond 10 mm may suggest malignancy.

Immune-suppressed children and those who have had chemotherapy are at greater risk of skin malignancy and should have regular skin review.

Children with a great many lentigines (small, dark brown 1–2 mm macules) may have associated findings (e.g. LEOPARD syndrome) and require screening for hearing and cardiac problems and regular follow-up.

Dysplastic naevi

These are a subtype of acquired pigmented naevi. Dysplastic naevi are usually macular and greater than 5 mm in diameter, with a less-regular border, some erythema, and some variability in colour. They may develop during childhood or after puberty. The presence of multiple dysplastic naevi is associated with a significant increased risk of melanoma. However, the great majority of melanomas that occur in this population are found in unaffected skin or normal moles rather than in pre-existing dysplastic naevi. Removal of a dysplastic naevus is only indicated if there is concern that a naevus may represent a melanoma. Regular photography and follow-up are indicated to aid in the detection of melanoma at an early stage.

Children with a family history of malignant melanoma and/or multiple dysplastic naevi are at a higher risk of developing melanoma and require regular skin review.

Halo naevi

Halo naevi appear as a sharply defined area of depigmentation 5–15 mm in diameter, centred on a regressing mole. They are common in childhood and benign. Most often the central naevus will eventually disappear, and the colour will slowly normalise.

Spitz naevi

Spitz naevi are fairly common, benign melanocytic lesions. They usually present prior to puberty as single, red or red-brown, symmetrical papules that can grow to 1 cm in diameter. They are vascular and blanch to reveal the underlying pigmentation.

Management is controversial as they essentially are a histological diagnosis, and it is difficult to be sure of the exact diagnosis of a particular naevus until it is

excised. It is thought that Spitz naevi are benign but usually do not resolve. Removal may be indicated because of uncertainty about the clinical diagnosis or for cosmetic reasons. Histological interpretation is sometimes difficult with findings that overlap with malignant melanoma. Assessment by an experienced dermatopathologist is required.

Hypopigmentation

Localised patches of depigmentation (total loss of pigment of skin or hair) can be seen in vitiligo, halo naevi, naevus depigmentosis and piebaldism. Localised patches of hypopigmented skin may be due to pityriasis versicolor (usually in adolescence), pityriasis alba (representing post-inflammatory change following mild eczema, usually in mid-childhood) and other post-inflammatory loss of pigment. Focal pale patches may be the only marker of leprosy in an individual from an endemic area.

Pityriasis versicolor

This is common in adolescents, as there is a requirement of background sebaceous activity, and caused by an increased activity of commensal yeasts. Multiple oval macules, 3–10 mm in diameter and usually covered with fine scale, appear on the trunk or upper arms. The lesions coalesce and may appear paler, redder or darker than the surrounding skin, hence the term *versicolor*. Treat with topical imidazole creams or with anti-yeast shampoos. For example, apply selenium sulfide 2% or ketoconazole 2% shampoo. Leave on for 30 minutes, if tolerated, rinse, and treat daily for 1 week and then monthly. If there are cosmetic concerns, a short course of oral ketoconazole followed by the above measures may be warranted. The yeast is readily treated, but the post-inflammatory hypopigmentation takes months to resolve. Relapses are common unless ongoing monthly treatments are used for a year or so.

Pityriasis versicolor is extremely uncommon in young children, but, when it occurs, lesions may be on the face or neck. If found in a young child, examine the child for other markers of an underlying congenital adrenal hyperplasia.

Pityriasis alba

This condition is common in prepubertal children, and it not a separate entity as

it simply represents post-inflammatory hypopigmentation secondary to mild eczema in darker skinned children. Single or multiple, poorly demarcated hypopigmented (but never completely depigmented) 1–2 cm macules are seen on the face or upper body. Lesions are not itchy but often have a fine scale. Treatment is the same for eczema, keeping in mind that the hypopigmentation takes many weeks to normalise.

Vitiligo

This condition is characterised by sharply demarcated, often symmetrical areas of complete pigment loss. Eventual repigmentation in childhood vitiligo is common and is helped by topical steroids or pimecrolimus cream. In troublesome cases, corrective cosmetics or UV therapy may be required. The association in children between vitiligo and other organ-specific autoimmune conditions (e.g. diabetes, thyroid disease) is small. Investigations for these are not necessary unless there is a clinical suspicion of an associated disorder.

Post-inflammatory hypopigmentation

This condition occurs particularly in dark-skinned people. Many inflammatory skin disorders may heal leaving diffuse, hypo- or hyperpigmented macules that can persist for months. Repigmentation eventually occurs.

Linear and whorled hypopigmention (‘hypomelanosis of Ito’)

See Segmental pigmentary disorder (including linear and whorled hyperpigmentation and hypopigmentation of Ito) p. 351.

Tuberous sclerosis

Hypopigmented patches may be the first sign of tuberous sclerosis. These patches can be regular or irregular in shape. One or two isolated hypopigmented patches in a young child are far more likely to be simple achromic naevi than to be the first sign of tuberous sclerosis. Tuberous sclerosis becomes much more likely if the child has a history of epilepsy. Look also for forehead plaques (pink, initially flat but later slightly raised) and shagreen patches (rough, slightly

thickened skin usually over the back). Other skin findings such as periungual fibromas and facial angiofibromas usually appear in older children. The diagnosis of tuberous sclerosis may require imaging of eyes, brain, heart and kidneys and investigation of relatives. Regular paediatric follow-up is required.

Generalised hypopigmentation

Generalised hypopigmentation, blonde hair and grey-blue eyes are seen in several genodermatoses involving chromosomes 11 or 15. The clinical presentations vary from mild to complete loss of pigmentation, and individuals may go undiagnosed unless compared to their siblings and parents. Early diagnosis and investigation are important to ensure early ophthalmological intervention and rigorous sun protection. Look for:

- poor vision, photophobia and nystagmus. Type 1 oculocutaneous albinism usually results in more severe disease than type 2
- bleeding diathesis, due to a platelet defect in Hermansky–Pudlak syndrome
- recurrent infections in Chédiak–Higashi syndrome
- mental retardation and obesity. Both Angelmans and Prader–Willi syndromes can present with albinism.

Skin texture

Lax skin

Lax, hyperextensible skin is seen in the several Ehlers–Danlos syndromes. There may be bruising, scarring at sites of minor trauma, joint hyperextensibility and arthritis, and recurrent urinary infections.

Firm or thickened skin

Scleredema (do not confuse with scleroderma) is a rare condition in childhood, which presents over some weeks with gradual thickening of the skin of the head, neck and upper trunk. It may be related to preceding streptococcal infection. Diagnosis can be confirmed by skin biopsy showing mucopolysaccharide intradermal staining. Resolution usually occurs over months.

Unusually firm skin is an early feature of systemic sclerosis. In children, there

is usually widespread skin involvement. Raynaud phenomenon may be present, and involvement of lungs, heart, kidneys and gastrointestinal tract usually occurs within a few years.

Waxy indurated skin may accompany neurological degeneration, and hepatosplenomegaly in type A Niemann–Pick disease and can be a feature of other genetic conditions such as premature aging syndromes and stiff skin syndrome. Thickened skin may also be seen in acromegaly and Addison disease.

Mouth disorders

Most of the conditions covered in the section on vesicular disease can involve the oral mucosa. Oral vesicles break rapidly so that usually only erosions or small ulcers are seen. Oral involvement is often the first feature of hand, foot and mouth disease, manifesting as a few lesions involving the tongue and palate, followed a day or two later by vesicles on the extremities. Oral erosions may be seen occasionally in varicella and rarely in zoster.

Primary herpetic gingivostomatitis

Primary herpetic gingivostomatitis (see p. 328) presents in infants and young children with malaise, high fever, soft-tissue swelling, lymphadenopathy and vesicles and erosions on the buccal mucosa, gingivae, lips and adjacent face. Drinking and eating are painful. Management includes supportive care, bathing of crusts and analgesia. Aciclovir is not routinely indicated unless risk factors for complications exist (e.g. underlying disease, immunosuppression).

Herpangina

Several echoviruses and Coxsackie viruses can cause the sudden onset of malaise, high fever, sore throat and vomiting, sometimes with abdominal pain. Examination of the pharynx and posterior mouth reveals a large number of small, 2 mm vesicles or erosions coalescing on an erythematous background to form painful ulcerations. Diagnosis can be confirmed by throat or faecal swab. Complete resolution occurs with supportive care over 5 days.

Transient lingual papillitis

Transient lingual papillitis is probably the same condition as eruptive familial

lingual papillitis. It was first described in 1996 but is much more common than generally realised. Most reported cases have been in the families of hospital staff. In one survey of 200 hospital staff, 50% of families appeared to have experienced this condition.[7] In most cases, an infant appears to be the index case, and many other family members are then affected. Older children present with a burning sensation on the tip or sides of the tongue. Younger children present with irritability and poor feeding of unknown cause. The tongue is normal apart from small papules or ‘scalloping’ along the anterior and lateral tongue margins without ulceration. Affected individuals are otherwise well without any fever, lethargy, respiratory or gastrointestinal problems. Symptoms and signs last from 2 to 20 days. Recurrent episodes may occur for many weeks and again years later. Manage with symptomatic treatment and education.

Angular cheilitis

Angular cheilitis can occur during childhood and is common in older children. It presents as persistent erythema, scaling and fissuring at the corners of the mouth and surrounding skin, sparing the buccal mucosa. Discomfort and pain may be worsened by stretching, during yawning for example, and by salty or tart foods. Most cases are primarily attributable to atopic eczema but are multifactorial. Contributing factors include saliva, licking, drooling at night, significant dental malocclusions, irritant and allergic dermatitis (e.g. certain acidic foods, cosmetics, medicated lip balms, sunscreen lip creams, toothpaste, fluoride and sucked or chewed lollies). *Staphylococcus aureus*, *Candida albicans* or streptococcal species may all be cultured from affected skin.

Rarely, angular cheilitis may be the presenting feature of nutritional deficiencies including riboflavin, iron, zinc, pyridoxine, biotin and protein metabolic disorders. Malabsorption, malnutrition, oral corticosteroids and oral antibiotics may contribute:

- Management requires avoidance as much as possible of any factors that may be contributing.
- Minimise acidic foods such as tomatoes and citrus fruit.
- A trial of non-fluoride toothpaste and supervised rinsing after teeth cleaning helps in some children.
- Topical unmedicated ointment or paste up to several times a day will assist recovery. Topical 1% hydrocortisone ointment is useful. If

microscopy reveals evidence of *Candida* or bacterial pathogens, topical miconazole or mupirocin, respectively, can be added.
- Allergy to a component of the treatment schedule should be suspected if the condition persists despite treatment, and patch testing may be warranted.
- In difficult cases, consider an uncommon underlying cause as above and a trial of iron, vitamin and mineral supplementation.

Geographic tongue

Geographic tongue is typically seen under 4 years of age but can occur in older children. Most children are asymptomatic, but some complain of tenderness or pain with salty foods. Irregular smooth patches are seen on the tongue in a pattern that changes from day to day. The cause is unknown. Geographic tongue is more common in psoriasis, but most children (>90%) who have geographic tongue never develop psoriasis. No treatment is needed unless some foods cause discomfort, in which case avoidance of those foods for a while is usually sufficient. If needed, topical or inhaled corticosteroid can alleviate symptoms.

Recurrent mouth ulcers

These are most often due to aphthous stomatitis. Aphthous ulceration usually presents as a few painful, 4–8 mm ulcers on an erythematous base on the non-keratinised mucosa (the ‘softer’ areas) rather than the gums or hard palate. They resolve in a few days but for some healthy children can be recurrent and bothersome. Less commonly, ulcers can be larger and persist for weeks. Any atypical features should lead to a consideration of less-common causes of recurrent mouth ulcers including the following:

- Iron, folate or vitamin B_{12} deficiency
- Recurrent erythema multiforme
- Pemphigus, pemphigoid and other immune-mediated blistering conditions
- Gastrointestinal disorders. Coeliac disease, Crohn’s disease and ulcerative colitis are all associated with mouth ulceration. Recurrent abdominal pain, intermittent diarrhoea and failure to thrive may be present

- Connective tissue disorders. Patients with Behçet's disease usually present in late childhood with ulcers at one site (mouth or genitals), and it may be many years before a second site is involved. SLE and juvenile rheumatoid arthritis may cause recurrent mouth ulcers
- Immunodeficiency states, HIV infection, cyclic neutropenia and periodic fevers
- Malignancy and chemotherapy. Lymphoma and Langerhans cell histiocytosis can present with non-healing mouth ulcers
- PFAPA syndrome of periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis
- Troublesome episodes of aphthous ulceration can be managed by:
    - investigating for an underlying cause as above
    - topical anaesthetic gels
    - topical steroid paste (e.g. triamcinolone acetonide)
    - oral colchicine.

Anogenital rashes

Anogenital rashes are common in infancy. Most anogenital rashes seen in infants that wear nappies are primarily caused by reaction to urine or faeces (irritant napkin dermatitis). Soaps, detergents and secondary yeast infection may contribute. In atypical or persistent anogenital rashes in infancy, consider psoriasis, Langerhans cell histiocytosis, zinc deficiency and malabsorption syndromes. In older children, consider perianal streptococcal infection (if painful), psoriasis, lichen sclerosis and atopic eczema. Kawasaki disease may be associated with a tender anogenital rash in an unwell child.

Treatment of rashes in the anogenital area requires caution. The moistness and occlusion of these areas increase the penetration and effectiveness of topical agents. For example, the absorption of topical hydrocortisone per square centimetre of anogenital skin can be several times greater than trunk skin. Adrenal suppression may occur after use of excessive potent topical cortisone preparations under the nappy area in small infants. Anogenital skin is also more likely to develop local steroid side effects, such as atrophy and striae.

Irritant napkin dermatitis

Irritant napkin dermatitis is the most common cause of napkin dermatitis in

infants and typically presents as confluent erythema that spares the groin folds. Variant presentations include multiple erosions in the natal cleft secondary to diarrhoea, scaly or glazed erythema, and satellite lesions at the periphery. Satellite pustules are suggestive of secondary *Candida* infection, although *Candida* infection is much less common with breathable disposable nappies.

Gluteal granulomas are purple-brown papules or nodules, often oval shaped, which can develop around the skin folds in infants with troublesome napkin dermatitis. Most cases are associated with the inappropriate use of potent topical CSs in this area. These lesions slowly resolve over several months provided the underlying irritant napkin dermatitis is treated.

Irritant or traumatic anogenital rashes may be seen in circumstances of suboptimal care, emotional abuse or physical abuse.

Management

- Keep the area clean and dry. Leave the nappy off whenever possible.
- Clean the anogenital area by hand under warm water or with diluted bath oil on cotton wipes. Avoid commercial ‘wipes’ which can be irritating or uncommonly cause allergy.
- Gel-based disposable nappies are preferred to cloth-based nappies.
- Use topical zinc cream or paste for mild eruptions. This functions as a barrier and should be applied thickly so that some is still present at the next nappy change. The cream or paste does not need to be completely removed at each change.
- Add hydrocortisone 1% ointment if inflamed. Do not use stronger steroids.
- Antifungal therapy is usually not needed even if *Candida* is present.

Candida napkin dermatitis

This occurs secondary to irritant napkin dermatitis and antibiotic use, leading to erythema and white material in the folds and satellite pustules. Treat the underlying cause as above, and add topical imidazole cream if necessary.

Anogenital psoriasis

Anogenital psoriasis can present at any age from infancy to adolescence. Sharply

demarcated, non-scaly, brightly erythematous plaques can be seen and may involve any or all of the perianal and intragluteal region, inguinal folds and the genital area in both boys and girls (Fig. 12.1.15). Symptoms are often surprisingly minimal. If pain and/or fissuring are present, swab for secondary streptococcal infection. Anogenital psoriasis may be an isolated finding, or there may be similar lesions in other intertriginous areas, such as the axillae, or more typical signs of psoriasis elsewhere. Occasionally, in infants, anogenital psoriasis will lead to a few lesions on the lower trunk and then a rapid eruption of lesions elsewhere including scalp, face and trunk.

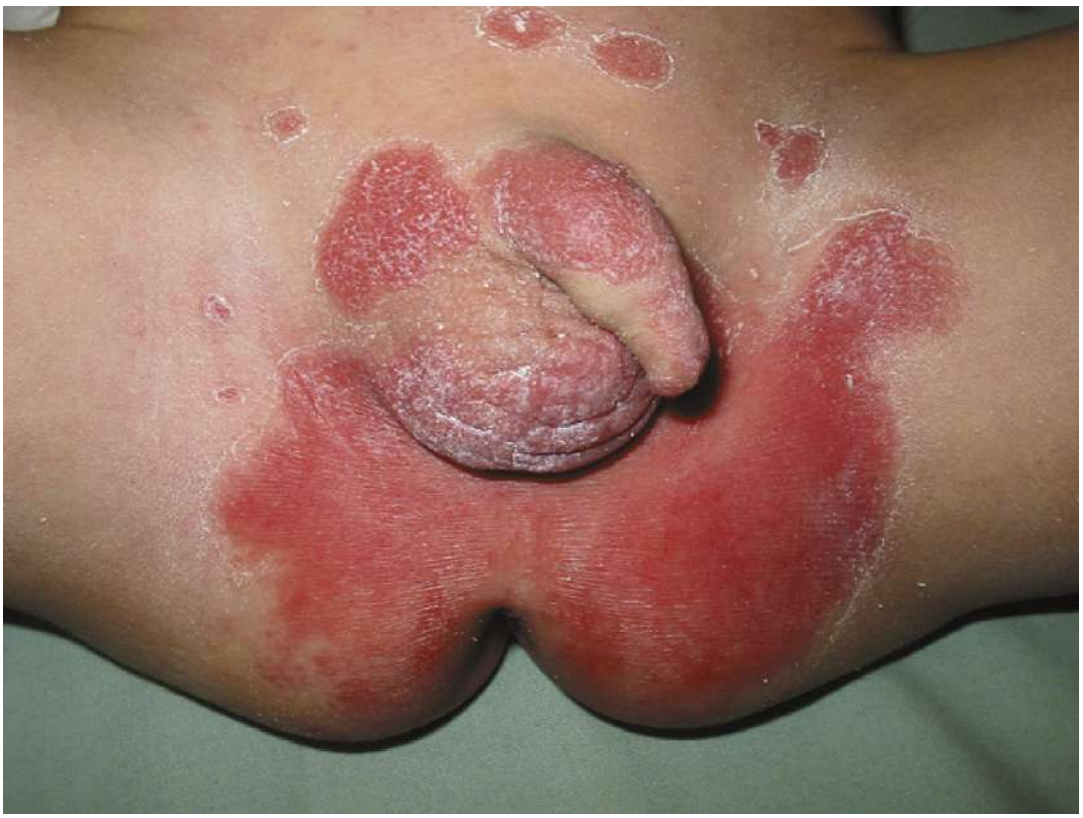

FIG. 12.1.15 Anogenital psoriasis in a 1-year-old boy.

Manage by treating streptococcal infection if present, and then treat as for irritant napkin dermatitis (see above). Medical follow-up is essential to ensure clearing, as children and adolescents are often unaware of quite significant asymptomatic anogenital psoriasis or fail to report it because of embarrassment.

Perianal streptococcal dermatitis

A painful, red, perianal rash in a child between the ages of 1 and 10 is likely to be due to perianal streptococcal dermatitis. The usual presentation is a child who has had a sore anal region for weeks or months. Defecation is painful, and secondary constipation is usually present, but treatments aimed at relieving the constipation have not helped. Bright red blood may be present on the stool or toilet paper. On examination, a localised, well-demarcated erythema that covers a circular area of 1–2 cm radius around the anus can be seen. Fissures or macerated skin may be seen.

Concurrent pinworm infestation should be considered. Constipation usually follows painful defecation, rather than being the cause of it. It takes a long while for the child to recover confidence in defecation after successful treatment.

Management

- Take perianal and throat cultures to confirm the presence of group A *Streptococcus*.
- Apply paraffin ointment three times daily to the perianal area for symptomatic relief.
- Topical mupirocin three times daily has a role. Note that resistance to mupirocin is becoming more widespread, and use should be restricted.
- Treat with phenoxymethylpenicillin (penicillin V) orally bd 250 mg (under 10 years), 500 mg (over 10 years) for a minimum of 3 weeks, although at times several weeks of therapy may be required.
- Consider treatment for pinworm with mebendazole or pyrantel.
- Keep stools soft with oral liquid paraffin.
- Perianal emollient ointment and stool softeners should be continued for a further 2 months after resolution of symptoms to prevent relapse, which is otherwise common.

Staphylococcal-mediated anogenital rashes

Bullous impetigo may present in the anogenital area as small vesicles or pustules that coalesce to give areas of peeling skin with peripheral margins of loose epidermis. These may be asymptomatic, and the infant is otherwise well. Provided the baby is well, the disease is localised and the parents will follow instructions regarding follow-up, an alternative is to treat with topical mupirocin 2% and general measures for napkin care. Oral cephalexin 30 mg kg^{-1} (max 500

mg) three times daily may be required for more widespread disease.

Herpes simplex virus

Primary HSV infection and eczema herpeticum can both occur in the anogenital area. Initially, small vesicles on an erythematous base may be seen, but these rapidly become small monomorphic erosions, crusting and coalescing, sometimes into an extensive, painful, erosive rash. HSV infection in the anogenital area of young children is usually innocent, contracted from a cold sore on a carer, but the possibility of child sexual abuse should be considered. The possibility of child sexual abuse is higher in older children. For management see p. 328.

Varicella (see p. 327)

Varicella lesions may initially be localised to the anogenital area in young children. Laboratory confirmation by PCR may be required to avoid an incorrect diagnosis of herpes simplex.

Threadworms

In older children, threadworms (*Enterobius vermicularis*) are a common cause of an itchy anogenital rash. Look for the worms at night, and treat with oral mebendazole 50 mg (<10 kg), 100 mg (>10 kg) (not in pregnancy or less than 6 months of age) or oral pyrantel 10 mg kg^{-1} (maximum 500 mg) as a single dose. A repeat dose 2 weeks later helps reduce the high rate of reinfestation. Emphasise the importance of wearing clean underwear at night, preventing perianal scratching at night, hand washing immediately on awakening, avoidance of thumb-sucking and good toileting hygiene.

Lichen sclerosis

Lichen sclerosis presents with chronic itch, discomfort or pain in the vulval area of girls aged 3 years or older. On examination, there may be an area of atrophy with white shiny skin, purpura or telangiectasia in the perivulval region. Cases have been misdiagnosed as sexual abuse. Management is with moisturisers and courses of moderately potent or potent steroid ointment. Treatment should be continued for many months or longer. Relapses after successful treatment are

common, and long-term follow-up is required to avoid long-term scarring and or neoplastic disease.

Vulval itch/vulvitis in prepubertal girls

Prepubertal girls with vulval itch usually have an identifiable dermatological condition. Girls between the age of 2 and 10 present with months or years of vulval itch, occasionally with stinging or pain on micturition. On examination, the labia majora may appear normal or may show erythema, scale, lichenification and excoriation. The introitus is normal. The great majority of these girls have atopic eczema. There may be signs of this elsewhere, or signs may be limited to the vulval area. Management is as for atopic eczema at other sensitive sites and involves avoidance of chemicals, soaps, bubble baths and other irritants and use of cotton underwear and ointment moisturiser. Use of topical CSs or pimecrolimus creams is appropriate to settle the erythema and itch. Measures to ‘oestrogenise the vagina’ are either irrelevant or counterproductive, and oestrogen creams should not be used.

Less common causes of vulval itch include psoriasis, threadworms, lichen sclerosis, staphylococcal folliculitis or infection with group A streptococci. In all these cases, clinical signs suggestive of the associated condition are likely to be present.

Zinc and other nutritional deficiencies

Zinc deficiency may be due to an inherited defect in zinc handling or may be an acquired deficiency as a result of prematurity, malabsorption syndromes or abnormally low maternal breast-milk zinc levels. Breast milk normally provides some protection from the inherited form, so presentation of inherited zinc deficiency is usually after breast-feeding has ceased. By contrast, acquired zinc deficiency can present earlier, in the neonatal period.

Zinc deficiency causes acrodermatitis enteropathica. A sharply defined, red, often extensive, anogenital rash is an early and prominent manifestation. Look for perioral (Fig. 12.1.16), perinasal and acral (hand and foot) dermatitis, alopecia, diarrhoea, and failure to thrive. Serum zinc levels do not correlate well with body zinc status. Oral zinc leads to a rapid improvement of the rash within days. Long-term follow-up to monitor the child’s zinc and copper status is essential.

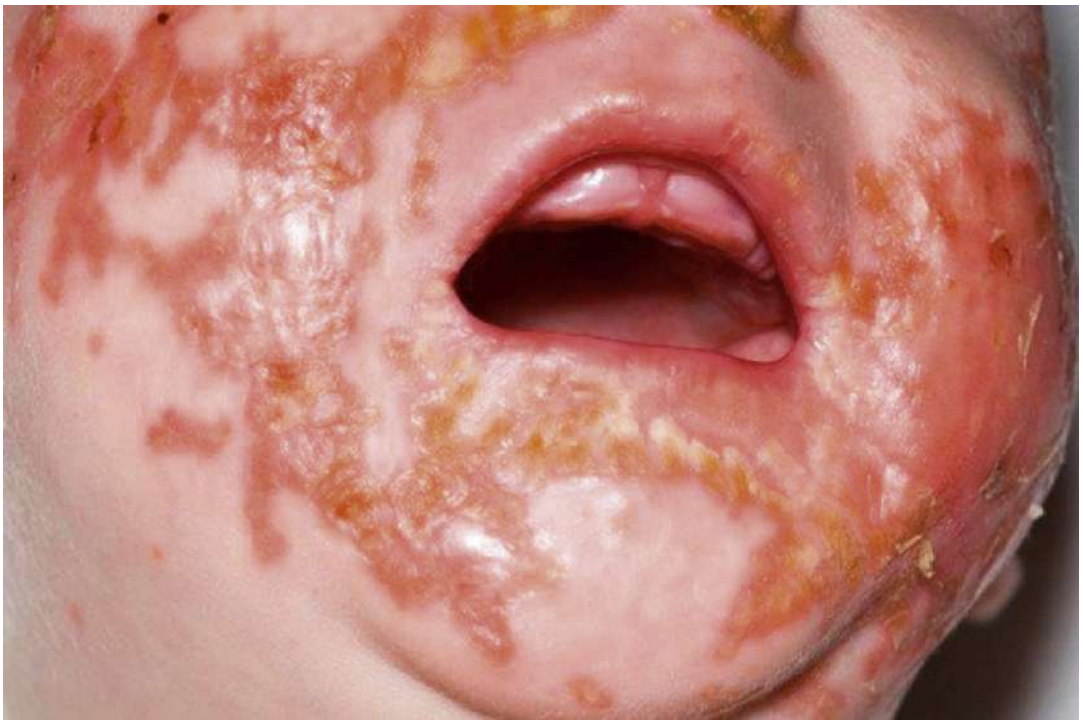

FIG. 12.1.16 Zinc deficiency. Erythematous, scaly, crusting plaques on the cheeks and perioral area.

Biotin deficiency and various other metabolic deficiency syndromes can give a similar picture, although usually in the setting of an unwell acidotic baby.

Malabsorption

Malabsorption from any cause (e.g. cystic fibrosis) can present with diarrhoea, erosive dermatitis and failure to thrive. There may be a progressive intractable napkin rash contributed to both by the diarrhoea and by secondary nutritional deficiencies. If malabsorption continues, the rash may become glazed and generalised in association with oedema and malaise. Identify and treat the cause of the malabsorption. Manage the anogenital area as for irritant napkin dermatitis. Topical pastes are thicker than ointments and may be required.

Langerhans cell histiocytosis

Langerhans cell histiocytosis may present in infancy as a chronic inguinal or anogenital rash, with brownish-red scale and petechiae, which is often erosive and unresponsive to treatment. A scaly, papular eruption on the scalp or trunk may appear. Purpura, fever, diarrhoea or hepatosplenomegaly may be present.

Constipation

Constipation by itself does not cause perianal erythema and pain. Look for another cause, e.g. eczema, psoriasis or streptococcal infection.

Anogenital papules and lumps

Anogenital warts

These are soft fleshy warts that occur at the mucocutaneous junctions, especially around the anus. They may be isolated, flesh-coloured nodules or may coalesce into large cauliflower-like masses. Management options include awaiting resolution, topical podophyllotoxin, imiquimod, curettage and diathermy and carbon dioxide laser.

The presence of genital warts in a young child is not an indication for mandatory reporting to government protective services. Genital warts in young children should prompt consideration of the possibility of sexual abuse, but transmission is usually by normal intimate parent–child contact, including at birth. Unless there are other risk factors for sexual abuse on history or examination, referral for sexual abuse investigation is inappropriate. If in doubt, seek paediatric advice. In children older than 5 years, the association between anogenital warts and sexual abuse increases, and assessment by a paediatrician should be considered.

Molluscum

In the anogenital area are often misdiagnosed as warts. Close examination of all lesions usually reveals a few to have the classical umbilicated pearly appearance. Molluscum in the anogenital region are common and are not a marker of child sexual abuse.

Congenital syphilis

Erosions and moist warty lesions in the perianal area may be seen in infants with congenital syphilis. There may be erythema on the palms and soles, fever, failure to thrive and hepatosplenomegaly.

Immunodeficiency states

Immunodeficiency from any cause may present in infancy as diarrhoea with a chronic erosive napkin dermatitis, widespread seborrhoeic dermatitis or neonatal erythroderma. Secondary infection with bacteria and/or viruses can complicate the clinical appearance. Look for other features of immunodeficiency including failure to thrive, erythroderma, lymphadenopathy, unusual infections and petechiae with eczema.

Hair problems

For an itchy scalp, consider tinea capitis, atopic eczema and head lice. For patches of hair loss, consider alopecia areata, traumatic alopecia, tinea capitis and kerion. For diffuse hair loss, consider telogen effluvium and nutritional causes. Long-standing sparse hair can be associated with ectodermal dysplasias and with many primary genetic abnormalities of hair and hair anchoring.

Head lice

Head lice infestation is common. Epidemics of head lice regularly sweep through primary schools in all areas. Nits (eggs) can be seen as small white specks firmly attached (unlike dandruff flecks) to hairs. Adult lice may occasionally be seen as small brown insects, about 4 mm long, walking across the scalp. Itching and excoriation are common. Presentation to an ED usually follows secondary eczematisation and infection causing extending weeping and crusted scalp lesions and neck lesions.

Suitable treatments include pyrethrin 0.165% (e.g. Pyrifoam), maldison 0.5% and permethrin 1% (e.g. Nix and Lyclear cream rinse). Topical ivermectin preparations are becoming available and may be more effective. Thoroughly moisten the hair with the treatment, and leave for 10 minutes. Rinse well, and comb out with a fine-toothed lice comb; physical removal of lice and eggs is an important part of therapy. Reapply 1 week later to kill any eggs that have subsequently hatched. Reinfestation is common. A regular physical inspection, use of conditioner and combing of the hair are as important as chemical treatment. A shorter hair cut can make this more manageable.

Tinea capitis

In Australia, tinea capitis can be caused by *Microsporum canis* contracted from

cats or dogs or by *Trichophytum* species contracted from other infected children or animals. The incidence of tinea capitis is much higher in children with racially determined tightly curled black hair. Any child of African background with an itchy scalp should be assumed to have tinea until proven otherwise. Apart from itch, tinea capitis is characterised by patches of hair loss with some short, lustreless, broken hairs a few millimetres in length. Redness and scaling are present in the patch. Occasionally, inflammatory pustular swellings may be present (see kerion below). Hair loss without any epidermal changes is not likely to be fungal.

Confirm the diagnosis by greenish fluorescence of the hair shafts (not present with some fungi) with Wood's light and with microscopy and culture of hair and scale. Treatment usually comprises griseofulvin orally 20 mg kg^{-1} in divided doses (maximum 1 g, after meals with milk) daily for 8 to 12 weeks or until non-fluorescent. Terbinafine and itraconazole are also effective. Ketoconazole 2% shampoo every 2 weeks can be used by all family members for 6 weeks to reduce cross-infection. Exclude from school until 2 days after treatment has commenced.[2]

Kerion (inflammatory ringworm)

This represents an inflammatory scarring immune response to tinea capitis. It is an erythematous, tender, boggy swelling, often several centimetres in diameter, which discharges pus from multiple points. Skin incision and drainage should be avoided as it leads to delayed healing and increased scarring. Scrapings and/or some extracted hairs should be sent for microscopy and culture. Treatment is as for tinea capitis combined with a brief course of oral steroids to suppress the inflammation. Antibiotics for secondary infection may occasionally be useful. Other inflammatory granulomas can mimic kerions. Exclude from school until 2 days after treatment has commenced.

Alopecia areata

Typically one or more oval patches of hair loss develop over a few days or weeks. These patches are usually completely bald, but some hairs may remain within the patches. Rarely, the hair loss is diffuse. The scalp appears normal and does not show scaling, erythema or scarring. Most cases in childhood resolve spontaneously, but progression to total scalp or body-hair loss or recurrent

alopecia can occur. Regrowth can occur up to decades later.

Management

- For isolated small patches present for weeks without further progression, no treatment is needed.
- In older children who are bothered cosmetically, intralesional corticosteroid injections are the treatment of choice.
- For recent or progressive hair loss, treatment with potent topical steroids may be trialled but is rarely useful as penetration into the deeper dermis to the hair follicle bulb is small. In difficult cases, other therapies, including contact sensitisation, irritant agents and pulsed corticosteroids, need to be considered.

Traumatic alopecia

This condition is usually caused by rubbing (as on the occiput of many babies), cosmetic practices (e.g. tight braiding) or hair pulling as a habit (trichotillomania). Trichotillomania may be largely nocturnal, and parents are often unaware of it. The affected areas are usually angular and on the anterior or lateral scalp. The areas contain hairs of different lengths and are never completely bald, unlike alopecia areata. Presentation to EDs may be triggered by the hair loss or by the sequelae of eating the hair, including acute intestinal obstruction (trichobezoar) in severe cases.

Management

- Recognition of the problem and a careful explanation to the family are often sufficient. Alert the family to the risk of trichobezoar.
- Trichotillomania in younger children does not usually indicate that significant psychological problems are present. It is a habit similar to thumb-sucking or nail-biting, and a low-key approach similar to that used in those conditions is appropriate. In older children, it may be part of an obsessive trait or other psychological problems that require professional and pharmacological support.

Diffuse hair loss

Diffuse hair loss in children is usually from telogen effluvium. This is characterised by an increased number of hairs going synchronously into the resting (telogen) phase and subsequently being shed several weeks later. Shed hairs have a small club-like appearance at the proximal end. Telogen effluvium presents as diffuse, often quite dramatic hair loss 4 to 16 weeks after the causative event. In children, this is commonly a high fever for a few days. Other causes include car accidents, operations and acutely stressful events. Complete regrowth always occurs.

Dramatic widespread diffuse hair loss may be from alopecia areata.

Diffuse hair loss occurs after cancer chemotherapy. More chronic, diffuse hair loss without an obvious cause may require investigation to exclude treatable causes, such as zinc or iron deficiency, malnutrition, excess vitamin A, oral retinoids for acne, hypothyroidism or antithyroid medications, hypopituitarism, hypoparathyroidism, diabetes and thallium poisoning.

Long-standing sparse or irregular hair is a feature of many genetic diseases. In some, such as monolethrix, the child is otherwise normal. In others, such as ectodermal dysplasias, many associated features may be present. These include decreased or absent sweating, pointy or missing teeth, dysplastic nails and dry skin. Clinical findings may be subtle. In any young child with unusual nails, skin or teeth who presents with fever and collapse, ectodermal dysplasia with inability to sweat and dysfunctional temperature control should be considered.

Hypertrichosis

Generalised hypertrichosis (increased hair in all areas) may be an isolated finding or may be related to inherited syndromes (including Hurler’s and De Lange’s syndromes), medications (especially minoxidil, phenytoin and cyclosporin), gastrointestinal disease (including coeliac disease), anorexia nervosa, hypothyroidism and porphyria (look for photosensitivity and blisters).

Local patches of hypertrichosis can be seen as an isolated finding or in association with lichenified eczema (temporary), pigmented lesions (congenital pigmented naevi, acquired naevi, Becker’s naevi), plexiform neurofibromas, other naevoid lesions, chronic rubbing or over the sacral area in association with underlying spinal anomalies (‘Faun’s tail’). A small degree of increased hair growth over the sacral area is common in many racial groups, and this is not a marker of spinal anomalies.

Hirsutism

Hirsutism refers to an increase in male pattern terminal hair. Increased pubic or axillary hair in young children may be due to adrenal, gonadal or CNS disease and requires investigation. Hirsutism in adolescent females may be an isolated finding or may be seen with obesity and amenorrhea in polycystic ovary syndrome. Cushing syndrome, mild congenital adrenal hyperplasia, virilising adrenal and ovarian tumours, exogenous androgens and thyroid dysfunction may cause hirsutism.

Nail problems

Infants' nails are often thin and exhibit a degree of koilonychia (spoon shape) that requires no investigation. Congenitally abnormal nails are usually atrophic and can be the presenting feature of rare inherited conditions, such as ectodermal dysplasias (these children may present with fever and heat prostration), dyskeratosis congenita, pachyonychia congenita (thickened nails), congenital malalignment of the great toenails and the nail-patella syndrome. Nails may have appeared normal at birth and become dystrophic over the first year of life.

Acquired nail disease is usually a result of paronychia, fungal infection, psoriasis, ingrown toenails or 20-nail dystrophy. It may also be seen in association with diseases, such as alopecia areata and lichen planus. Nail-biting and picking can lead to marked deformity of involved nails.

Paronychia

Acute paronychia (inflammation of the nail fold)

Is usually caused by *Staphylococcus aureus* or group A *Streptococcus*. It can be seen during therapy with oral isotretinoin for acne. One or more nail folds become red, painful and swollen. Pus may be present at the nail margin. Any cause of prolonged wetness or minor trauma (e.g. finger sucking, nail picking, finger eczema, preceding chronic paronychia) predisposes to acute infection by breaching the cuticle between nail and skin. Treatment requires identification and avoidance of the trigger factors, warm compresses, drainage if needed, oral cephalexin 30 mg kg^{-1} (max 500 mg) three times daily, topical paraffin ointment and ongoing nail care. If due to oral isotretinoin, dose reduction and treatment as above usually resolve the problem.

Chronic paronychia

Is traditionally associated with *Candida albicans*, which can be found in specimens from most cases. However, *Candida* is now seen by many authorities as a secondary feature that may not be particularly relevant to the onset and maintenance of the condition. The primary event is disruption of the nail cuticle resulting in inflammation of the proximal nail fold. Chronic paronychia may be a primarily eczematous condition caused by many factors including finger sucking, nail picking, finger eczema and any increased exposure to moisture. Affected nail folds are often dusky red or purple with loss of the cuticle and without significant pain. The nail often has several horizontal ridges corresponding to episodes of inflammation of the proximal nail fold. The nails may become increasingly dystrophic with time but do not tend to thicken. Chronic paronychia may be present for months or years with occasionally acute exacerbations that respond to oral antibiotics without clearing the underlying problem.

Management

- Management requires potent topical CS application daily for several weeks.
- Avoid all factors that predispose to continuing irritation or allergen exposure. Nails should be kept dry. Repeated applications of Vaseline or even a hydrocolloid dressing over the cuticle can assist in this. Cotton gloves under rubber gloves should be used for wet work.
- In difficult or unresponsive cases, consider an unusual infection or rare underlying cause such as mucocutaneous candidiasis (especially if oral *Candida* infection is present), hypoparathyroidism and malignancy.

Nail psoriasis

Psoriasis can present as isolated nail disease and can mimic acute or chronic paronychia and onychomycosis. Features of nail psoriasis include pitting, thickening and brownish subungual discolouration. There may be repeated episodes of pain associated with the collection of pus under the nail. Drainage leads to relief, but antibiotics don’t help. Cultures are sterile. Detailed history and examination of the rest of the child may reveal other features of psoriasis. Initially treat with potent topical steroid ointments.

Ingrown toenail

The nail of the large toes can grow into the lateral nail fold and cause pain, redness, swelling and occasionally pus secondary to *Staphylococcus aureus*. The chronic inflammatory process leads to progressive granulation tissue formation. Management involves avoidance of shoes where possible, avoidance of tight-fitting shoes, careful cutting of the nails and use of pledgets between the nail and lateral nail fold. Oral antibiotics are not usually required. Surgical removal of granulation material is helpful. Trimming of the lateral nail alone usually results in recurrence of the problem. In troublesome cases, surgical ablation of the lateral nail plate may be warranted.

Tinea unguium (onychomycosis)

Dermatophyte infection may affect one or more nails. White or yellow patches develop at the distal and lateral nail edges. The rest of the nail may become discoloured, friable and deformed with accumulation of subungual debris. Multiple nails may become involved over years. Tinea may also present on the adjacent skin, particularly in the interdigital folds. It can be difficult to clinically distinguish between psoriasis and fungal infection in nails. Always confirm the diagnosis by microscopy and culture of nail clippings before commencing treatment. If cultures are negative, repeat the cultures, but do not start oral treatment. In very mild cases only, treatment with physical debridement and antifungal nail lacquer (e.g. amiolfarone) may be effective. Most cases require oral therapy with terbinafine or griseofulvin for months. School exclusion, although specified in some states, is not medically warranted or appropriate.

Itch without rash

Some children present with an itch without an obvious rash. Other children may have multiple excoriations without obvious cause, or they may have much more severe pruritus than would be expected for the eruption present. All such children need to be evaluated for:

- atopy
- scabies
- symptomatic dermographism
- dermatitis herpetiformis

- iron deficiency
- food allergy
- drug use; most pharmaceutical, recreational and herbal drugs can cause itch
- systemic disease, including hepatitis C infection, diabetes, uraemia, conjugated hyperbilirubinaemia, paraproteinaemia, polycythaemia and thyroid disease.

Collection of specimens

Bacterial swabs (Gram stain and culture)

- Swab lesion with a plain dry swab and roll over a glass slide. Discard swab and air dry slide.
- Swab lesion with second swab. Wet swab with sterile saline if lesion is dry. Place swab in charcoal medium (maintains organism viability).

Swabs for herpes simplex virus or varicella zoster virus

- Roll dry flocked swab over suspicious lesion for PCR. To collect cells for culture, base must be swabbed vigorously, which may cause discomfort. Swab should be cut off and placed in viral transport medium for culture.
- Fluid from vesicles may be collected into viral transport medium for culture and PCR.

Swabs for polymerase chain reaction (for pertussis, varicella zoster virus, herpes simplex virus, *Mycobacterium ulcerans*, etc.)

- Dry (flocked) swab.

Swabs for *Chlamydia trachomatis*

- Dry swab for PCR
- For culture, swab lesion with two dacron or rayon-tipped swabs. Smear one onto ‘microtrak’ well, and break tip of other into *Chlamydia*

transport medium.

Skin scrapings for fungal/dermatophyte culture

- Use scalpel blade to scrape the surface of the lesion. Collect flakes of skin in specimen jar.

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SECTION 13

Eyes

OUTLINE

13.1

Ophthalmological emergencies

Nader Beshay, and Jennie Martin

ESSENTIALS

1 The consequences of missing ocular pathology are more serious in children due to the risk of amblyopia.

2 Observation alone will often reveal the key information needed to reach a differential diagnosis.

3 An asymmetric red reflex warrants urgent review by an ophthalmologist.

4 Significant pain relief after instillation of topical anaesthetic localises the pathology to the conjunctiva and/or cornea.

5 Ensure that no pressure is applied to the globe until an open globe injury has been excluded.

6 Lid swelling and concurrent sinusitis should raise the suspicion for orbital cellulitis.

Introduction

Few ophthalmological emergencies relate exclusively to children. However, their pattern of incidence is variable, depending on the age of the child. Whilst the pathology is similar to that seen in adults, the approach in assessing the child and reaching the diagnosis may be quite different. Furthermore, because the visual cortex is still developing in children, the consequences of missing an important diagnosis and delaying treatment can lead to permanent visual impairment (amblyopia), even if the pathology is identified and treated later on, after the so-

called ‘critical period’.

History

The history may be obvious or concealed. The most common presentation is a red eye (or eyelids) that may or may not be associated with pain. If the parent/carer reports a sudden red eye that has appeared without obvious cause and the child is preverbal, consider trauma or foreign body.

A general medical history as well as family history can provide clues to the diagnosis, in addition to the more focused ophthalmic history. Enquire about sick contacts as viral conjunctivitis is very contagious and commonly affects multiple siblings simultaneously.

A sexual history may be relevant in teenagers because of the possibility of sexually transmitted disease. Any patient with a red eye and a history of contact lens use should be reviewed by an ophthalmologist within 24 hours.

Younger children, especially preverbal infants, will not complain of decreased vision. Ask the parent/carer whether he/she feels the child is seeing well. Any child in whom a decreased visual acuity is suspected or confirmed on examination needs to be reviewed urgently by an ophthalmologist. Nevertheless, most children presenting to the emergency department (ED) will present with a red eye as the chief complaint.

Examination

This is perhaps the most daunting aspect of the assessment. Trying to perform a routine ocular examination on a healthy child can be challenging, let alone a child who is acutely distressed. Most children with a painful red eye will be very reluctant to have their eyes/eyelids physically touched. As such, begin by performing as much of the examination with observation alone.

Every child presenting with an eye complaint should have some form of visual acuity (VA) assessment performed. Infants should be able to fix and follow by 3–4 months. Toddlers may be able to perform matching tests where a chart is shown and they are asked to match the corresponding shape or letters with the chart on their lap. If this is not available, use the illiterate E chart. Test each eye individually if possible. Pinhole testing should be attempted in school children, as this will indicate whether any visual impairment is due to a refractive or other problem.

The red reflex provides useful information. Dim the lights, and use the direct ophthalmoscope from a distance (arm's length) to assess the reflex in each eye. They should be roughly equal. An equal red reflex rules out a number of important conditions, including a large retinal detachment, a large vitreous haemorrhage, a dense cataract and retinoblastoma. Furthermore, a foreign body on the surface of the cornea may be seen as a black dot that blocks a small part of the returning red reflex. It is helpful to keep in mind that the 'red reflex' is only truly red (or deep orange) in fair-skinned children. In darker children, it will have a pale orange appearance. The key to identifying an abnormal red reflex is to look for asymmetry. Any child with an asymmetric red reflex should be referred to an ophthalmologist.

Visual fields can be tested in younger children by using two toys – one large toy at the centre and one smaller toy that is brought in to the visual field from the periphery. The child should switch fixation from central to peripheral when the object is in view. Older children should cooperate with the usual confrontational method of counting fingers in each quadrant.

Next, assess the pupils. This can be challenging in children with a dark iris. The direct ophthalmoscope can be used to get around this problem. Once again, dim the lights, and look through the direct ophthalmoscope. Direct pupillary response and a relative afferent pupillary defect (RAPD) can be detected using this technique.

Examine the child's eye movements, looking for evidence of ophthalmoplegia. Strabismus is quite common in children, and the parents should be asked if this is something new. Although uncommon, any child with a third, fourth or sixth nerve palsy should be seen urgently by an ophthalmologist, especially when in the presence of other symptoms suggestive of elevated intracranial pressure, such as nausea, vomiting and lethargy.

A topical anaesthetic such as amethocaine can be used as both a therapeutic and diagnostic agent. Pain or irritation that apparently goes away after instillation of anaesthetic effectively isolates the pathology to either the conjunctiva or surface of the cornea. In children who refuse to open their eyes for the anaesthetic, apply a drop onto the medial canthus and when they open their eyes, the drop should flow in. Fluorescein (which does not sting) can then be applied to look for epithelial defects in the cornea. 2% fluorescein should be used when performing Seidel's test if an open globe injury is suspected (see chapter 13.3 on ocular trauma).

Some children may cooperate with a slit-lamp examination, which is essential

if you are looking for cells in the anterior chamber, cells within the vitreous and just a more detailed look at the structures of/around the eye.

Intraocular pressure is difficult to measure accurately in the adult, let alone the child. An acute rise in intraocular pressure should be suspected if:

- there is a history of blunt trauma to the eye a few days ago
- there are accompanying symptoms of nausea, vomiting and headache
- the cornea looks oedematous (if the details of the iris are difficult to appreciate due to a hazy cornea)
- there is conjunctival injection around the limbus
- the pupil is poorly responsive to light.

Performing direct fundoscopy is challenging and is virtually impossible in the distressed child. A view of the optic disc alone may be all that is achieved and will most likely only be possible if the pupils have been dilated. In children, it is safe to use one drop of phenylephrine 2.5% and one drop of tropicamide 1%, if no contraindications exist. In all children with suspected trauma, ensure that no pressure is applied to the globe until an open globe injury has been excluded.

If unable to perform an adequate examination due to an uncooperative and/or severely distressed child, it may be necessary to use sedation. This is particularly important in settings where review by an ophthalmologist is not feasible within a reasonably urgent timeframe and a high level of suspicion exists for serious ophthalmic disease/injury.

It is imperative that other serious injuries (such as intracranial injury) are excluded prior to administration of sedation and that the risks of sedation are discussed with the parents. Although nitrous oxide is useful when sedation is required for minor paediatric procedures, it should never be used in the setting of paediatric ocular trauma to avoid the risk of inducing vomiting (which can lead to extrusion of intraocular contents in the setting of an as yet undiagnosed open globe injury). However, it may be considered for the repair of simple eyelid lacerations once globe injury has been excluded.

The red eye in paediatrics

A red eye is the most common ED presentation. The flowchart below (Fig. 13.1.1) can be used to help reach a likely diagnosis. There is some overlap of symptoms between conditions, but this flowchart is based on the most prominent

symptoms for each condition.

The red eyelid

Preseptal versus orbital cellulitis

Preseptal cellulitis is one of the most common conditions seen in the paediatric ED. It usually occurs secondary to a nearby wound or insect bite or may even occur from an infected chalazion, hordeolum (stye) or dacryocystitis. The child usually appears well with no systemic features, despite grossly swollen lids (sometimes difficult to part).

The keys to differentiating preseptal from orbital cellulitis are:

- the absence of systemic features
- the absence of concurrent sinusitis
- a normal VA with no RAPD
- a white eyeball
- normal eye movements with no proptosis or diplopia.

As with all conditions, the threshold for admitting should be reduced the younger the patient. Treatment is with antibiotics. In the case of preseptal cellulitis, admission may be warranted for a younger child or infant as the orbital septum is less well developed, and preseptal cellulitis may progress to orbital cellulitis. If the child is not admitted, ensure close follow-up with his/her GP.

Orbital cellulitis is a sight-threatening and potentially life-threatening condition. Infection can drain through the valveless venous system of the midface, leading to cavernous sinus thrombosis, meningitis, brain abscess or frank sepsis. Always be certain this has been ruled out before discharge. Patients usually require an orbital CT scan, systemic antibiotics and urgent ophthalmology review.

Blepharitis

Blepharitis is an inflammatory process of the eyelids that is characterised by crusting at the base of the eyelashes. There may be oedema of the lids themselves, and occasionally the conjunctiva is also injected. Staphylococci are the usual organisms. Treatment should involve removal of the crusts using a warm flannel, cleansing with dilute baby shampoo and regular massage of the lids in the direction of the lashes.

Hordeolum (stye)

A hordeolum is an acute infection, usually staphylococcal, of an eyelash follicle. It causes a small, tender swelling, often with a head of pus, which is seen at the lid margin. Treatment is with warm compresses and chloramphenicol ointment until resolution occurs.

Chalazion (meibomian cyst)

This is caused by inflammation of the lipid-secreting meibomian glands of the lid, probably due to ductal obstruction. It presents as a slowly progressing, mildly painful red lump in the eyelid. Most settle with conservative treatment, namely warm compresses for 10 minutes, four times a day with light massage and chloramphenicol ointment for up to 4 weeks. Rarely, if this treatment fails, or the cyst is excessively large causing visual disturbance, it can be incised. It may become complicated by preseptal cellulitis that will require systemic antibiotics.

Dacryocystitis

This is defined as infection of the nasolacrimal duct and presents with pain, redness, swelling and tenderness of the overlying skin and the adjacent nasoperiorbital area. There may also be tearing, discharge and fever. Pressure over the lacrimal sac may express pus from the punctum. Any discharge should be sent for culture. *Staphylococcus aureus* and *Staphylococcus epidermidis* are the commonest organisms. Treatment should be with cephalexin orally or cephazolin intravenously if unwell. Gentle massage in the more minor cases may well aid in clearing the infection and unblocking the duct, as dacryocystitis is invariably associated with ductal obstruction.

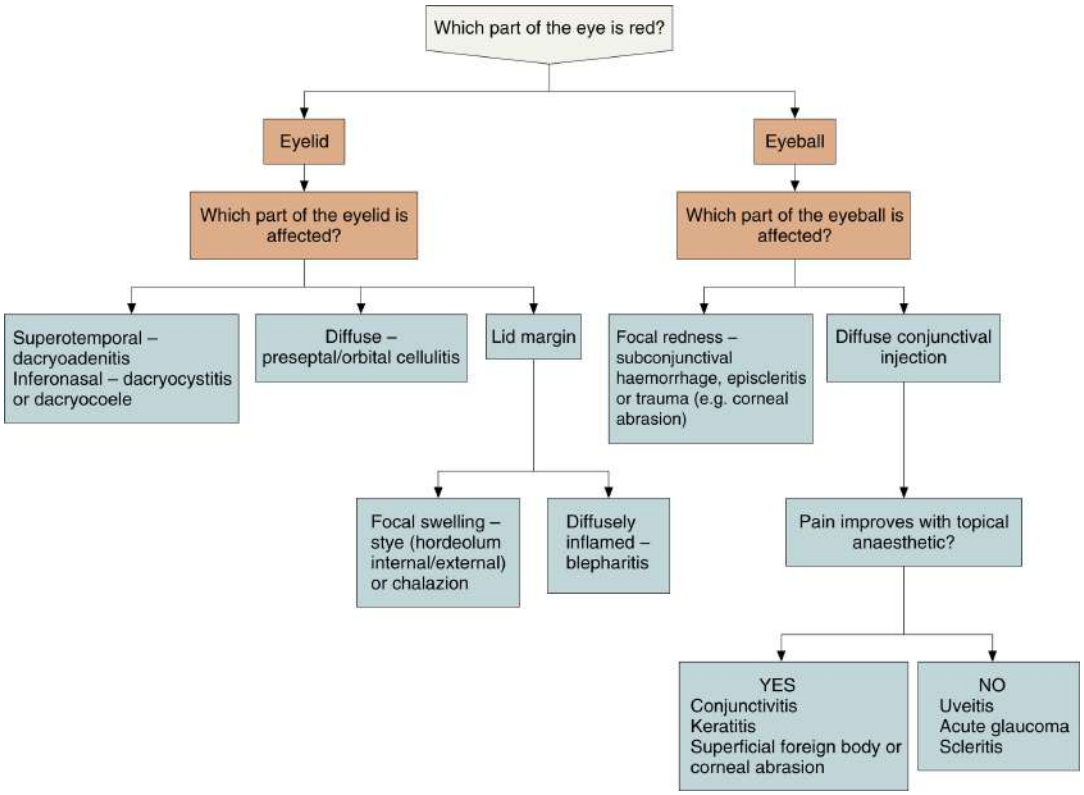

FIG. 13.1.1 Diagnosis flowchart for red eye in paediatrics.

The red eyeball

Conjunctivitis

Neonatal conjunctivitis

Neonatal conjunctivitis (ophthalmia neonatorum) is defined as conjunctivitis occurring within the first 30 days of life. These infants require urgent review by an ophthalmologist. The maternal history, particularly gynaecological and obstetric history, should be obtained and relayed to the consulting ophthalmologist.

The causative organisms are usually passed from mother to foetus during passage through the birth canal, commonly *Neisseria gonorrhoeae*, *Chlamydia trachomatis*, staphylococci, streptococci or herpes simplex.

Gonococcal infection classically presents within 24 to 48 hours of birth (usually within 5 days) with acute eyelid oedema, bulbar conjunctivitis, chemosis and a profound purulent discharge. Both eyes are usually affected. As the presentation may overlap with other infectious agents, a swab should be

taken before treatment is commenced. An urgent Gram stain may show gram-negative intracellular diplococci. Treatment should not be delayed, as there is a risk for rapidly progressive corneal ulceration and perforation. Ceftriaxone 50 mg kg intravenous (IV) should be commenced with the addition of erythromycin orally to cover for infection with *C. trachomatis* until cultures are negative. Irrigating the eyes regularly with normal saline to eliminate the ocular discharge is also recommended.

Chlamydial infection is classically associated with a watery then mucopurulent discharge 5 to 14 days after delivery. There is also palpebral conjunctival injection but less lid oedema than with gonococcal infection. Presentation ranges in severity from mild conjunctival redness with minimal watery discharge to severe redness, swelling of lids and purulent discharge. Perform swabs for culture and polymerase chain reaction (PCR) and then commence treatment with oral erythromycin 10 mg kg QID for 21 days or alternatively azithromycin 10 mg kg OD for 5 days. Concurrent treatment with topical erythromycin may also be beneficial. This disease is complicated by pneumonitis in 10–20% of cases, characterised by nasal discharge, cough and tachypnea. For both gonorrhoeal and chlamydial conjunctivitis, the mother will need treatment, and her partner will need to be screened.

The same organisms that affect older children can also cause neonatal conjunctivitis. These organisms typically present from days 5 to 7. Clinical findings do not distinguish the pathogen, so cultures should be taken and treatment commenced with broad-spectrum antibiotic ointment.

Herpes simplex virus (HSV) conjunctivitis is the exception and should be suspected if there is a maternal history of infection, vesicular blepharitis or dendritic ulceration. Neonatal HSV infection usually presents with a systemic meningitic-like illness. Treatment should be with topical and systemic acyclovir and urgent ophthalmological consultation sought.

Neonatal conjunctivitis is at times confused with congenital nasolacrimal duct obstruction. Congenital nasolacrimal duct obstruction occurs in up to 10% of newborn infants and resolves spontaneously in 95% of patients by the age of 1 year. It presents as watery or sticky eyes in the first 1 to 2 weeks of life. The eye itself is neither red nor inflamed differentiating congenital nasolacrimal duct obstruction from neonatal conjunctivitis. Management consists of using warm water or breast milk to remove sticky discharge. Massage of the lacrimal sac has not been shown to be beneficial.

Conjunctivitis in older children

Viral conjunctivitis is the most common condition in children presenting to the ED with a red eye. It is characterised by a mostly clear discharge, gritty eyes and often preceded by a viral upper-respiratory-tract infection (URTI). There may be pre-auricular lymphadenopathy, which if present ‘seals’ the diagnosis.

Bacterial conjunctivitis often produces a more mucopurulent discharge, with the eyelids often sticking together. In cases of suspected bacterial conjunctivitis the eye should be swabbed and treatment commenced with chloramphenicol eye drops every 2 hours and review organised for the next day. Referral to an ophthalmologist should occur immediately if vision or cornea is affected, if there is no improvement or worsening after 2 days and if symptoms persist after 5 days of treatment. Sexually active teenagers should be swabbed for chlamydia and treated systemically as well as topically if sexually transmitted infection (STI) is suspected. Enquire about co-existing urogenital symptoms, as this is common with chlamydial conjunctivitis. Gonorrhoea produces a copious purulent discharge and represents a true ophthalmological emergency as corneal ulceration and perforation can occur rapidly. Immediate treatment and consultation with an ophthalmologist are required.

The hallmark of allergic conjunctivitis is itch, and one should think twice before making the diagnosis in the absence of this symptom. It is bilateral, chemosis is often present and symptoms also include burning red eyes and watery discharge. There is often a history of atopy (allergic rhinitis, asthma, atopic dermatitis and food allergy). Allergic conjunctivitis may also be a reaction to eye drops so ensure an ophthalmic medication history is recorded. For moderate symptoms, oral antihistamines and topical allergy eye drops such as Patanol may help.

Keratitis

Keratitis is defined as inflammation of one or more layers of the cornea. It appears as a focal white corneal opacity without an overlying epithelial defect. If ulceration is also present it will stain with fluorescein. Symptoms include pain, redness, photophobia and poor vision. Epiphora, conjunctival injection and cells in the anterior chamber may all be seen on slit-lamp examination. When a large amount of white cells are present in the anterior chamber, they will settle to form a hypopyon.

Bacterial infection is the commonest cause of keratitis and is considered to be

one of the leading causes of blindness in the developing world. Organisms include gram-positives (staphylococci, streptococci and bacilli), as well as gram-negatives. Of these, *Pseudomonas aeruginosa* is of significance as it is the commonest cause of keratitis in the contact lens wearer and can rapidly lead to corneal perforation. Other organisms include the following:

- Fungi; particularly after trauma from vegetable matter. The course is usually more gradual in comparison to bacterial keratitis.
- Acanthamoebic keratitis; a parasitic infection seen in contact lens wearers, especially if they have poor lens hygiene. It is also a complication of a corneal injury contaminated by soil. Patients often present with pain out of proportion to early clinical findings which progress over several weeks.
- Herpes simplex virus (HSV); may cause punctate keratitis before the classical dendritic ulcer manifests. It may affect both eyes, and corneal scarring may be permanent.
- Varicella zoster virus (VZV); associated with a dermatomal skin rash. Look for vesicles at the tip of the nose (Hutchinson's sign), which indicates involvement of the nasociliary branch of V1 and mandates urgent ophthalmological review.

All patients with suspected infectious keratitis should be urgently referred to an ophthalmologist for culture by corneal scraping before starting antibiotic treatment.

Recurrent corneal erosion

This manifests with recurrent attacks of pain, photophobia, grittiness and epiphora, which occur on waking or rubbing the affected eye. There is often a prior history of corneal abrasion, which creates an area of epithelial 'weakness' that becomes vulnerable to detachment in the future. Examination will often reveal a corneal defect, but minor epithelial changes may have resolved by the time the patient is reviewed. Treatment consists of antibiotic ointment and then artificial tears once the corneal defect is healed. Ophthalmic follow-up is recommended.

Uveitis

Uveitis refers to inflammation of any part of the uveal tract (iris, ciliary body or

choroid). In most cases, it is limited anteriorly (iris or ciliary body), and in 50% of cases the cause is unknown (idiopathic). The predominant symptom of uveitis is photophobia since pupillary constriction in response to light necessitates movement of the iris and/or ciliary body, which is inflamed in this condition. Thus cycloplegics provide significant relief. There may be a clear watery discharge, but the pain does not subside with instillation of local anaesthetic. There will be conjunctival injection, but this will be most pronounced around the cornea (‘ciliary flush’).

Traumatic iritis is the most common cause of uveitis in a child presenting to the ED and often occurs 1–3 days after blunt trauma to the eye, so enquire about a recent history of trauma (refer to chapter on ocular trauma).

Acute anterior uveitis is an uncommon but serious condition in infants as it is often associated with a systemic vasculitis. Uveitis in older children is also associated with a number of systemic inflammatory conditions, including juvenile chronic arthritis, inflammatory bowel disease and psoriasis. Less common conditions causing uveitis in the child include sarcoidosis, tuberculosis, herpes viruses (HSV and VZV) and syphilis.

All patients with suspected uveitis should be referred to an ophthalmologist for detailed posterior segment examination and further testing (such as HLA-B27).

Episcleritis and scleritis

Episcleritis presents with acute redness to a section of the sclera. There is usually minimal discomfort or tenderness, and it is only the more superficial episcleral vessels that are engorged. Although it is associated with connective tissue disorders, HSV, VZV and inflammatory bowel disease, most presentations are idiopathic in nature.

Treatment of episcleritis is symptomatic with artificial tears and oral non-steroidal anti-inflammatory drugs (NSAIDs), but if severe, topical steroids can be used. Ophthalmic follow-up should be arranged to ensure resolution.

Scleritis is a more serious disorder that presents with pain that gradually becomes severe and a diffuse redness due to injection of the scleral, episcleral and conjunctival vessels. When viewed in daylight, the sclera may have a bluish hue, and vision may be reduced. There is a strong association with connective tissue disorders. Treatment consists of oral NSAIDs and steroids and should be commenced by an ophthalmologist.

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13.2

Congenital, developmental and neoplastic conditions of the eye

Greg Stevens

ESSENTIALS

1 These conditions are most likely to present with a concern from a parent or caregiver who has noted that there has been a change in the appearance of his/her child's eyes or that he/she is not responding to visual cues in an expected way.

2 A detailed examination will help to further define the abnormalities or deficits noted. These conditions require specialised investigations and management, and as such consultation with a paediatric ophthalmologist is advised.

Deficits in visual acuity

Deficits in visual acuity (VA) may reflect a deficit anywhere from the visual cortex to the cornea, and a careful examination including pinhole testing in children old enough to cooperate will help localise this and guide appropriate referral.

Strabismus

Abnormalities of the binocular alignment of the eyes, strabismus, may present with a concern from a parent that his/her child has a squint, and this may be confirmed by careful examination of the corneal reflex and use of the cover-

uncover test (observation of deviation of affected eye with uncovering while focusing on near or distant object).

Strabismus may be convergent (esotropia), divergent (exotropia), upwards (hypertropia) or downwards (hypotropia).

Strabismus may be a dynamic process, occurring in relation to alterations in the accommodative reflex with refractive errors.

The importance of strabismus lies in the development of amblyopia, which is the decrease in VA occurring in visually immature children due to the lack of a clear image provided to the retina. It is usually unilateral. Strabismus may also be an early sign of significant visual pathology, e.g. retinoblastoma and Coats' disease (exudative retinitis/retinal telangiectasis).

Paediatric cataracts

These can be detected as an abnormality of the red reflex. There is an association with several chromosomal, metabolic and intrauterine infective causes.

Congenital nasolacrimal duct obstruction

This is a common disorder, occurring in up to 6% of children. It presents with excessive overflow of tears. It has a good outcome, with 96% resolving by the age of 1 year (see Chapter 13.1).

Infantile glaucoma

Glaucoma in children is rare but if undetected may cause blindness.

Because of the elasticity of the paediatric eye, significant enlargement of the cornea may occur, with subsequent clouding of the cornea. Excessive lacrimation and photophobia may be present. It is a cause of irritability in the infant.

The treatment is usually surgical.

Ocular tumours

Tumours may present with an evident deformity, with proptosis, or with an alteration in the visual axis. Any of the tissues in the orbit and eyelids may give rise to tumours. Children treated in infancy with radiotherapy for an intraorbital tumour are at increased risk of developing malignancies in the radiated area in

later life.

Retinoblastoma

Retinoblastoma is the most common primary intraocular tumour in childhood with an incidence of 2 to 5 per million children. Ninety per cent occur before the age of 5 years. More than half present with leukocoria, with strabismus being the next most common presenting complaint.

One in three is congenital, typically multifocal and bilateral. In 75% a germline mutation of the *RB1* gene occurs for the first time *in utero* without inheritance from a parent. There is an association with other extra-ocular tumours such as pineoblastomas, and new tumour formation after initial treatment is common. When radiotherapy is used as part of the treatment, there is a lifetime increased risk of developing other cancers.

Two-thirds are due to a sporadic mutation of the *RB1* gene. These are typically unilateral and do not have the same association with other extra-ocular tumours.

When the tumour is confined to the eye there is a good survival rate.

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common primary intraorbital tumour in childhood and usually occurs in the first decade. Presentation is usually with rapid proptosis or displacement of the globe, but may be associated with eyelid oedema, and can lead to confusion with an infective process.

Current treatment regimens have a good survival rate, but recurrence is not uncommon.

Neuroblastoma

The orbit is a common site of metastasis of neuroblastomas. The primary is usually adrenal or abdominal and usually known at the time of orbital metastasis. Most present in the first 5 years of life with periorbital ecchymosis, proptosis, periorbital oedema, strabismus, Horner syndrome, or heterochromia iridis.

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13.3

Ocular trauma

Jennie Martin, and Nader Beshay

ESSENTIALS

1 There are unique challenges to obtaining an ocular history and examination in a child.

2 Anxiety is pronounced in the carer: be calm in your approach.

3 Consider hidden injury.

4 Never apply pressure to an eye that may be ruptured or penetrated.

5 Chemical injury: commence immediate treatment, prior to history and examination.

Introduction

Injury is the leading cause of visual disability and blindness in children and has the following features:

- Severe eye injury is not uncommon.
- All age groups may be affected, most commonly toddlers and school age children, with a male predominance after infancy.
- The mechanism is usually unintentional.
- Causes include sports activities (ball strikes or head clashes); sharp tools, knives and scissors; thrown projectiles (paintballs); plant thorns; pet bites/scratches; elastic/bungee cords or fireworks.
- Education is important as up to 90% of injuries are preventable with the majority of injuries occurring at home.

• Visual impairment results in an emotional, social and economic cost.

Trauma to the eye often engenders a marked anxiety reaction in parents/carers, particularly with concern about long-term visual impairment. Use a careful and calm approach to enable cooperation, ensuring the parent/carer is close so the child can be reassured, and a thorough examination may be performed.

Always consider what may lie beneath an injury that appears to be superficial. Even in an uncooperative child, extensive information can be obtained by observation alone. Uncommonly, mild sedation may be required to assist with examination. However, when there are genuine concerns, referral for general anaesthesia to enable adequate examination may be required. If gentle restraint does not allow adequate examination or a particular procedure, repeated and forcible restraint should not be performed.

Begin by taking a careful history. In addition to aspects of history common to all presentations, ask specifically about existing eye disorders, glasses or contact lens wear, the mechanism of injury and subsequent events.

Often the injury is unwitnessed or the child may be too frightened to reveal details, and so the history may be vague or concealed. Have a high index of suspicion for hidden injury. Specifically ask for visual symptoms of reduction or change in vision (diplopia, floaters or flashing lights) which children are less likely to spontaneously report or notice. Children are prone to the oculocardiac reflex, and a history of bradycardia, nausea, somnolence or syncope strongly suggests orbital fracture, with or without muscle entrapment, or a globe injury.

Perform the non-invasive aspects of the examination first. Reassure the patient (and the carer) that you will not hurt him/her. Dim the room lights if possible, keeping the ophthalmoscope light to a minimum. Be systematic and touch last. Importantly, know when to stop prior to upsetting the child and if concerns or incomplete examine refer for ophthalmological opinion. If appropriate, relieve pain with topical anaesthesia early to assist examination.

Document the visual acuity (VA) in each eye (monocular testing). VA provides an indication of prognosis at presentation. VA testing should be adjusted to the age and ability of the child. Fix and follow testing of a colourful object or your face should confirm any defects in eye movements; test ability to reach for a small toy, or small object like 100s and 1000s, in each eye, with the other eye covered. In the verbal child use an Allen chart using pictures (allow the child to identify the pictures closely first), a Tumbling E chart (described as table legs pointing in different directions) or a formal Snellen chart. A difference of two or

more lines between the two eyes is clinically significant. Remember children often have low attention spans, so do not insist on them reading every line. If acuity is markedly reduced, use finger counting at a specified distance (CF), hand movements (HM) or light perception (LP) at close range. If the patient is totally blind this is referred to as no light perception (NLP).

Examine and document:

- red reflex
- pupillary shape, size and reaction to light
- assessment for a relative afferent pupillary defect (RAPD)
- ocular movements in all directions of gaze (nystagmus, diplopia, pain or limitation of movement)
- inspect the lids
- palpate orbital rims for steps or tenderness
- test for infraorbital nerve sensation (lower lid, medial cheek and side of the nose, upper lip)
- the position of the globes (exophthalmos, enophthalmos)
- the conjunctival surface of the eyelids (the deep superior fornix requires lid eversion for viewing)
- conjunctiva and sclera for laceration or foreign body
- corneal surface (use fluorescein staining where possible)
- examine the anterior chamber depth and clarity.

Globe trauma

Subconjunctival haemorrhage presents as a painless red eye with a collection of bright, smooth blood confined to a sector of the bulbar conjunctiva. This is sharply demarcated at the limbus. VA is not affected.

Causes include:

- trauma
- idiopathic
- hypertension
- bleeding diathesis
- valsalva (cough, heaving lifting, straining).

Pain with extra-ocular movement, reduced vision, hyphaema, pupil

abnormality and/or bloody chemosis raises suspicion of a more complex globe injury. A 360 degree subconjunctival haemorrhage should be referred urgently for ophthalmological review as the globe may be ruptured posteriorly. No specific treatment is required for an isolated subconjunctival haemorrhage, though patients and parents/carers may find the sudden appearance alarming. The haemorrhage will clear spontaneously within 2–3 weeks, with the blood turning from red to brown to yellow before fading away.

Conjunctival lacerations present as a red eye with a foreign body sensation and usually a history of trauma. Conjunctival lacerations may be associated with subconjunctival haemorrhages. The conjunctival edges can be separated gently with a moistened cotton tip following topical anaesthesia to assess the depth of injury. If the diagnosis is uncertain, refer for ophthalmological opinion to ensure a scleral perforation or subconjunctival foreign body is excluded. If the conjunctival laceration is isolated, treatment with antibiotic ointment or drops is required for 4–7 days. They rarely require repair.

Corneal abrasions are usually very painful. They may present with:

- tearing (epiphora)
- a foreign body sensation
- photophobia
- VA may be reduced, dependent on the size of the abrasion and the position over the visual axis
- conjunctival injection, iritis and eyelid swelling may be associated.

The application of a topical anaesthetic provides temporary pain relief and will assist in allowing the eye to be opened for examination. In a non-compliant child, place a drop at the medial canthus, and when the eye is opened, the drop flows in. The non-verbal child may present simply with undifferentiated distress with or without refusal to open the eye, and topical anaesthetic may be diagnostic.

To date high-quality evidence for safety of short-term topical anaesthesia use at home is lacking, and patients should not usually be provided with this. Continued instillation of topical anaesthesia and prolonged home use may impair healing, inhibit protective reflexes and permit further injury.

Corneal abrasions may be associated with a foreign body on the lid conjunctiva, which must be everted to be examined fully. An upper lid foreign body is suggested by linear vertical abrasions on the cornea. The inner surface of

the upper lid is examined by asking the patient to look down, applying a cotton bud tip to the lid crease and applying light pressure. Use the eyelashes to pull the everted lid over the bud tip, away and up from the globe. Hold the lashes against the orbital rim to keep the lid everted. To return the lid, release the pressure, and ask the patient to look up. The lower fornix is easily inspected by applying downward pressure to the lower lid while the patient looks up.

Corneal abrasions are diagnosed by demonstrating a staining defect with fluorescein using either an ophthalmoscope (+12 magnification) or a slit lamp. Use only a small amount of fluorescein as excessive dye can mask a defect. Abrasions appear bright green when viewed under a cobalt blue light (available on a slit lamp or ophthalmoscope). Abrasions are managed similarly to defects remaining after foreign body removal.

Corneal foreign bodies should be assessed for and referred for ophthalmological opinion – those with an intraocular foreign body immediately (see penetrating eye injury). Always document the VA. Topical anaesthesia is usually required to relieve pain and blepharospasm, enabling examination.

Foreign body removal requires adequate magnification and illumination. Foreign bodies may be removed by irrigation, a cotton-tipped applicator or needle removal. Needle removal of a superficial foreign body must occur under magnification at the slit lamp and requires a cooperative child. Approaching from the temporal side, use a 25-gauge needle attached to a 1 mL or 3 mL syringe, bevel angled away from the eye, to gently scrape the foreign body from the cornea. Ophthalmological referral should occur if the foreign body is central or deep or if the child is uncooperative. Post removal, complete an examination of the eye using fluorescein to ensure no underlying open globe injury exists.

If a rust ring or residual foreign body remains, next-day ophthalmological review should be arranged. Refer patients with central or large corneal abrasions for daily review. Topical antibiotic treatment should be commenced. A topical non-steroidal anti-inflammatory drug (NSAID) provides effective analgesia, and if there is severe pain, a cycloplegic (tropicamide 0.5–1%, cyclopentolate 0.5–2%) may be prescribed to relieve ciliary spasm (avoid in infants). As with all wounds, ensure tetanus prophylaxis is administered. Eye patching does not reduce pain or aid healing and may cause difficulty with walking in children and thus should be avoided.

Contact lens wearers should be referred for follow-up and require pseudomonal coverage (tobramycin or ciprofloxacin). The lens should not be worn until the defect has been healed for a week.

Corneal lacerations may be partial (not into the anterior chamber) or full thickness (penetrating eye injury). Complete examination is required to exclude a penetrating injury of the cornea or sclera. Ensure the anterior chamber is of normal depth and there is no hyphaema. Superficial partial-thickness lacerations will heal spontaneously with antibiotic cover; however, daily review is necessary to exclude the development of infection until healing is complete. Deep partial-thickness lacerations should be referred for consideration of repair. Seidel's test can be used (see open globe injury). For management of full-thickness lacerations refer to open globe injury.

Conjunctival foreign bodies (palpebral/lid or bulbar/ocular) should be removed after topical anaesthesia, with gentle irrigation or a moist cotton-tipped swab.

Hyphaema is blood in the anterior chamber, which may appear as pooling. This is usually a result of blunt or penetrating trauma. Hyphaema presents with pain, photophobia, mydriasis/miosis, reduced VA or blurred vision. The red reflex will be intact, unless a vitreous haemorrhage or retinal detachment has also occurred. If the pupil is dilated, it is important to differentiate between traumatic mydriasis (present only in the affected eye) and a RAPD (use the swinging torch test – in a positive/abnormal test there is paradoxical initial dilatation of the affected pupil when light is shone quickly from the unaffected to the affected eye); this may suggest an optic nerve or severe retinal injury. Hyphaemas may vary in size from microscopic hyphaema (only visible using a slit lamp) to blood involving the whole anterior chamber (so-called '8-ball hyphaema'), though most will involve <50%. Urgent ophthalmological referral is required for any hyphaema in children. The aim of treatment is to prevent raised intraocular pressure (IOP), secondary hemorrhage and corneal staining. Treatment includes restricted activity, eye shield, anti-emetic, cycloplegic and topical steroid in some children. The child should not lie flat, even when asleep, as doing so will cause blood to collect within the entire 360 degrees of the drainage angle, blocking the outflow of aqueous and precipitating an acute rise in IOP. NSAIDs and aspirin should be avoided. Always consider non-accidental injury in young infants. Rebleeding occurs in up to one-third of patients, usually after a few days.

Open globe injury (globe rupture and penetrating eye injury)

A ruptured globe occurs when the integrity of the sclera or cornea is disrupted by blunt trauma, while a penetrating eye injury occurs due to direct perforation by a sharp object. There may be surprisingly few signs. Suspect if there is a peaked pupil (apex of the teardrop points to the perforation), which may be the only clue to occult open globe injury. Chemosis overlying the laceration, subconjunctival haemorrhage, corneal or scleral laceration, loss of the red reflex, a RAPD, distortion of the anterior chamber (deep or shallow), bubbles in the anterior chamber, extrusion of the intraocular contents, hyphaema, or loss of ocular motility may be indicative. A Seidel's test can be used if the diagnosis is unclear. Apply a moistened fluorescein strip over the potential site of perforation. Use the blue light of the slit lamp, and the leak from a perforation will manifest as a green dilute aqueous stream within the darker, concentrated orange dye.

Once an open globe injury is suspected, further emergency department (ED) examination is unnecessary. Contact the ophthalmologist urgently:

- DO NOT apply pressure to the globe if penetration or rupture is suspected.
- NEVER attempt to remove a protruding foreign body from the globe.
- NEVER force the lids open.
- Do not use further eye drops and never use eye ointment on an open globe.

It is important to appreciate that iris material that has prolapsed through an open globe injury wound can appear like foreign material sitting on the surface of the cornea/sclera. Do NOT attempt removal of this material, as this may still be viable tissue that can be saved during surgery.

Rest the child in bed, head up. Protect the eye with a rigid eye shield. If a formal eye shield is unavailable, one can be created from the base of a polystyrene cup. Keep nil by mouth. Commence an anti-emetic and analgesia, and ensure tetanus prophylaxis and systemic antibiotic prophylaxis. If an intraocular foreign body or globe rupture is suspected CT will usually be required.

Eyelid trauma

A laceration to the eyelid may be partial or full thickness and may involve the lid margins, canthal tendons, levator complex or canalicular system.

Perform a thorough and complete eye examination to exclude an injury to the globe. An open globe injury should be presumed, until specific examination has excluded this. Pressure exerted by attempts at cleaning and repair may apply pressure to a potentially open globe. Children who are unable to cooperate enough to allow accurate assessment of wound depth should be referred for examination under anaesthesia. If an open globe injury is suspected, refer immediately and follow the instructions above.

The mechanism of injury should be determined to assess the risk of a foreign body (e.g. windscreen shattering) and whether significant contamination may have occurred. The nature of a bite, whether human or animal, should be established to ensure appropriate antibiotic coverage.

Indications for emergency ophthalmologic consultation include:

- bites and those with significant contamination requiring debridement
- lacerations involving the eyelid margin (to prevent notching)
- lacerations to the medial third of the eyelid (due to the risk of injury to the canalicular system)
- lacerations with involvement of the levator aponeurosis of the upper eyelid (which produces ptosis) or the superior rectus muscle
- exposure of orbital fat. This suggests the orbital septum has been lacerated as there is no subcutaneous fat in the lids themselves
- lacerations with significant tissue loss.

Wounds requiring referral should be cleaned with normal saline and have foreign material removed as much as possible. Following cleansing, the wound should be covered with a saline-soaked dressing, prophylactic antibiotics commenced for bites or significantly contaminated wounds and tetanus status considered.

If the laceration is suitable for repair in the ED, the eyebrow should not be shaved as long-term cosmetic alterations may result, and the hair direction assists in correct alignment of the wound. Tissue should not be removed, as the good blood supply of the eyelid generally ensures viability. Partial-thickness lacerations should be repaired with 6/0 synthetic suture, and full-thickness lacerations should be repaired by an ophthalmologist. In general, non-absorbable sutures should be removed in 4–7 days. If procedural sedation has been required for repair, use rapidly absorbing sutures (e.g. fast gut or vicryl rapide) so re-sedation for removal of sutures is not required. Tissue glue is not advised due to

proximity to the lashes and cornea.

Ecchymosis

A black eye is a common injury and may be limited to a minor ecchymosis of the lid or extend to a periorbital haematoma with significant oedema. Always consider a blowout fracture of the orbit, globe injury and/or base of skull fracture or anterior fossa fracture (suggested by a subconjunctival haemorrhage without posterior limit). Always document the ocular motility. Care must be exercised when opening the lids so that pressure is not exerted on the globe until an open globe injury is excluded. If the eyelids are markedly swollen, examination of the eye is difficult. By placing one's thumb on the infraorbital and supraorbital rims, swollen eyelids can often be separated without placing pressure on the globe. The eyelids should not be forcibly opened. If a strong index of suspicion exists, place a rigid shield over the eye, and refer for an ophthalmological opinion. When swelling limits direct visualisation of the eye, ocular ultrasound is used in some centres to allow non-invasive assessment.

After other injury is excluded, eyelid haematomas require no specific management. Cold compresses may provide analgesia for the first 24 hours. The eye should be re-examined in 24 hours to ensure an injury has not been missed. Swelling and bruising may appear to extend down the cheek or to the other eye.

Orbital trauma

Suspect an orbital fracture when there is tenderness of the orbital rim or crepitus of the lid indicating a sinus fracture. There is often an associated haematoma of the eyelid. 'Blowout' fractures (fracture of the orbital floor ± the medial wall) from blunt trauma are typically seen in adolescents. Orbital fractures in children are typically trap-door type floor fractures which cause entrapment of the extra-ocular muscles and orbital fat more commonly than in adults.

Look for:

- restricted ocular movements which may produce diplopia, usually on upward gaze
- enophthalmos
- infraorbital nerve anaesthesia.

Nausea and vomiting may occur. Facial CT with axial and coronal views and ophthalmological and faciomaxillary referral are required. Ensure a globe or intracranial injury is not present, as these will have greater treatment priority. Eye injury may occur in over 50% of orbital fractures, with globe rupture in 5–10%. If prominent symptoms of the oculocardiac reflex are present, immediate surgical repair is indicated. If not, surgery may be arranged non-urgently. Instruct the patient not to blow the nose, as orbital emphysema may occur or worsen if already present.

Chemical burns require urgent immediate irrigation as early effective treatment affects outcome. Chemical burns are unusual in childhood but may be very serious. Anaesthetise the eye/s with local anaesthetic drops (procedural sedation may be required in young children), and then copiously rinse the eye with at least 2–4 litres of fluid (preferably Hartmann's, warmed if possible) for *at least 30 minutes*. Use IV tubing connected to the bag of solution, and direct the flow from medial to lateral onto the conjunctiva, not cornea. Aim to have the giving set regulator fully open. Ensure the fornices are irrigated by everting the lid. A sweep of the conjunctival fornices with a moist cotton-tipped applicator ensures the removal of debris or foreign body from this area. A Morgan lens (a moulded lens applied to the eye) allows continuous irrigation if available but may trap foreign material if not cleared prior to insertion. Irrigate for 30 minutes prior to history and examination. Five minutes after irrigation has been completed, check the pH with litmus paper, and continue irrigation until a pH of 6–8 is achieved. A urine dipstick can be used (trim with scissors to retain the pH section). Check the pH again 20 minutes after irrigation to ensure there is no drop in the pH. Once irrigation is completed, examine for retained foreign bodies. Particulate matter requires removal (moist cotton bud or toothed forceps), and this may require general anaesthesia. Alkali burns (dishwasher liquid, oven cleaner) produce liquefactive necrosis and are often more severe than acid burns.

Only once irrigation has been completed, assess injury severity, and document VA. Assess with a slit lamp and fluorescein stain. Moderate or severe burns are suggested by significant epithelial loss, chemosis, corneal oedema or haziness, blanching of the conjunctival vessels or opacification. Refer immediately for ophthalmological review. All patients require treatment with antibiotic drops/ointment and analgesia ± cycloplegics and ophthalmological follow-up.

Thermal burns are managed similarly to abrasions. UV/radiation keratitis may result from excessive sunlight. The symptoms develop several hours after

exposure with pain, tearing and red eye. There are usually bilateral superficial corneal defects seen on fluorescein staining. Treatment is with topical antibiotics ± a cycloplegic.

Traumatic iritis presents with the onset of a dull, aching pain, photophobia, and tearing within 3 days of trauma. Possible signs include a small pupil; perilimbal injection of the conjunctiva and pain in the affected eye when a light is shone into either the affected or non-affected eye; reduction in VA; and hyphaema. White blood cells and a flare are seen within the anterior chamber when examined under the slit lamp. This is best seen by placing the slit lamp beam at 45 degrees with full intensity and a short narrow slit (1 mm x 1 mm). The appearance is like dust in a room illuminated with a torch. Refer immediately for ophthalmological review.

Non-accidental injury may result in any eye injury. A high index of suspicion is required if the injury is inconsistent with the given history or the history is inadequate, particularly in young infants.

Further reading

New South Wales Department of Health. *Eye Emergency Manual. An Illustrated Guide* 2nd ed. Sydney: NSW Department of Health; 2009.

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SECTION 14

Ent and Dental

OUTLINE

14.1

The ear

Loren Sher, and Laura Graley

ESSENTIALS

1 Otitis externa usually results from excessive exposure to heat and moisture and is usually very painful. Treatment involves cleaning, keeping dry and topical antibiotics.

2 Acute otitis media is a very common emergency presentation, but not all red eardrums are due to otitis media. Management with adequate analgesia is essential. However, antibiotic use should be restricted to specific circumstances.

3 Chronic suppurative otitis media usually presents with painless and offensive discharge. Treatment is with ear toilet and topical antibiotics. Oral antibiotics have little or no role.

4 Otitis media with effusion is very common in children, but treatment is unnecessary in the majority, with resolution over 3 months.

5 Mastoiditis continues to be a problem in the antibiotic era and in many cases is the first presentation of ear disease. Admission, myringotomy and intravenous antibiotics constitute the mainstays of medical management.

6 Ear trauma is uncommon. Accidental ear injuries are usually unilateral and isolated. Ear trauma is rare in the first year of life and may indicate non-accidental injury. Haematomas should be removed by aspiration or excision.

Otitis externa

Introduction

Otitis externa includes various conditions from the most common acute diffuse otitis externa (swimmer's or tropical ear) to otomycosis, localised (furunculosis) or chronic otitis externa. It occurs commonly in hot, humid climates or in the summer of temperate climates. It predominantly affects children between the ages of 5 and 14 years but can affect any age group.

Risk factors include swimming and other water exposure, local trauma, loss of the protective coating of the ear canal, including cerumen, and obstruction of the ear canal. Occlusive ear canal devices, such as hearing aids or earplugs, are additional predisposing factors.

Chronic otitis externa may be a sign of an underlying dermatological disease, such as seborrhoeic or atopic dermatitis.

Important differentials include foreign body, suppurative otitis media, cholesteatoma and contact dermatitis, e.g. shampoo.

History

It may initially present with aural fullness or itch but usually progresses to pain with or without discharge. The pain is often severe and is worse with chewing. There may be associated hearing loss.

Otomycosis or fungal otitis externa makes up 10% of cases and has a more insidious onset.

Examination

Oedema and erythema of the canal with serous or purulent discharge are usual. The tragus is tender to manipulation. With increased severity, the canal becomes occluded with periauricular oedema and may progress to otitis externa with cellulitis when the child becomes febrile with a toxic appearance.

Differentiation from acute otitis media with perforation or chronic suppurative otitis media is important. Usually in these conditions, the tragus and canal are not tender, and there are no erythema and oedema of the canal.

Acute localised otitis externa (furunculosis) occurs in the posterosuperior aspect on the ear canal.

Otomycosis has only mild canal wall inflammation and thick otorrhoea. Deep-

seated itching is the most prominent feature. Different fungal species have characteristic appearances on otoscopy. *Aspergillus*, for example, may appear as fine coal dust in the ear canal. *Candida* infections have a soft white sebaceous material and a pseudomembranous lining.

Investigations

Investigations are largely unnecessary and rarely alter empiric treatment. The organisms found in diffuse otitis externa are most commonly *Pseudomonas aeruginosa* and *Staphylococcus aureus*. Furunculosis is usually *S. aureus* and otomycosis – *Aspergillus* or *Candida* species. Consideration of cultures – aerobic, anaerobic and fungal – is worthwhile in cases resistant to routine therapy, in recurrent disease and if there is more extensive disease – such as associated cellulitis.

Treatment

Acute diffuse otitis externa is managed with frequent gentle cleaning of the canal using rolled tissue spears or similar until the external canal is dry. Submersion in water should be avoided (swimming prohibited).

Ototopical medications are the mainstay of treatment and should be administered after aural cleaning. Topical antibiotics such as framycetin/gramicidin/dexamethasone combinations are commonly used. Ciprofloxacin is also effective. Most patients require a 7-day course of treatment and should expect their symptoms to improve within 48–72 hours. Symptoms beyond 7 days should prompt continuation of treatment for an additional 7 days. Persistence beyond 14 days should be considered treatment failure, requiring cultures and alternative therapy.

Insertion of a wick or ribbon gauze can facilitate topical medication application. The ear wick should be changed every 2–3 days. Close follow-up for repeated cleaning with or without wick reinsertion may be required.

Furunculosis treatment is by local heat application and oral antibiotics (flucloxacillin or cephalexin) or incision and drainage.

Otomycosis will require canal cleaning and antifungal drops.

Prevention

Keeping the ear canal dry and avoidance of trauma to the canal are the mainstays of prevention.

Complications

Progression to cellulitis of the nearby skin/soft tissue and/or lymphadenitis may occur. Oral antibiotics are usually then indicated, with cephalexin a reasonable first choice, but antipseudomonal antibiotics may be necessary. Progressive cellulitis and a toxic-appearing child will require admission for intravenous antibiotics, including pseudomonas cover. Less commonly, involvement of the parotid gland, temporomandibular joint or base of skull may occur.

Acute otitis media

Introduction

Acute otitis media (AOM) is one of the most common primary care paediatric presentations. It is most prevalent between 6 and 24 months of age and then declines with a small increase at 5 and 6 years of age.

It occurs as a result of infection of the middle ear cavity by both viral and bacterial organisms. It is frequently over diagnosed and remains a common cause for excessive antibiotic use. Adequate analgesia and a selective approach to antibiotic use are the mainstays of management.

History

Classic symptoms include fever, malaise and ear pain. The pain can be severe and during the night may wake the child. Other systemic features such as nausea and vomiting can occur. In younger children, presentation is often non-specific, with crying and irritability. Eardrum perforation and otorrhoea may occur (see complications). These symptoms may present as a primary complaint or frequently occur in the course of an upper respiratory infection.

Examination

A red, bulging, non-mobile tympanic membrane and middle ear effusion are the most reliable constellations of signs. Redness of the eardrum is a non-specific finding and may be seen with a high fever or following crying. Alone, therefore,

it is not diagnostic of otitis media and is an inadequate finding to make the diagnosis.

Investigations

The diagnosis of otitis media is made solely on clinical grounds, and investigations are rarely performed. In cases where tympanostomy has been performed, two-thirds are bacterial-culture positive, with a predominance of *Streptococcus pneumoniae*, *Moraxella catarrhalis* and *Haemophilus influenzae*.

Treatment

The administration of adequate analgesia is paramount to the management of acute otitis media. Paracetamol alone may not be adequate, and the combination with ibuprofen may be required. Topical instillation of lidocaine (lignocaine) 2% has been shown to be a useful adjunct for rapid pain control but should be combined with longer-acting oral analgesia. Decongestants and antihistamines have not been shown to be effective and are not recommended.

The majority of cases of otitis media will resolve spontaneously. However, antibiotics continue to be widely used. In an otherwise healthy child over 2 years, most authorities now recommend deferring antibiotic use for 2 to 3 days and to commence treatment only if the child remains symptomatic at review. Approximately 80% of children will avoid antibiotic use with this approach. Provision of a prescription upfront, with advice to commence antibiotics in 2 to 3 days if the child remains unwell, has been shown to result in approximately 50% of children avoiding antibiotics. Both strategies are reasonable, with the latter chosen in cases where access to timely medical review is uncertain. Early antibiotic therapy continues to be advocated in children less than 2 years of age or in any age group presenting with severe symptoms: vomiting, high-grade fever, perforation or bilateral disease. All indigenous children should be commenced on antibiotics at the initial presentation.

Amoxicillin is a reasonable first-line antibiotic choice. Amoxicillin + clavulanate is the next choice for poor responders. Cefaclor has a significant rate of serum sickness reactions in children and should not be used.

Complications

The most common complication is perforation of the drum and otorrhoea. Other complications are very unusual but potentially severe. Most are due to bacterial spread and include extracranial complications such as mastoiditis, cholesteatoma and facial nerve paralysis and intracranial complications such as epidural abscess, meningitis and lateral sinus thrombosis.

Persistent middle ear effusions are almost universal after an episode of acute otitis media and should not be viewed as a complication. Complete resolution over several months occurs in 90% of cases.

Prevention

Prophylactic antibiotics confer a small decrease in recurrence at best and are likely to contribute to increasing antibiotic resistance and generally are not recommended. More appropriate means of reducing recurrence include avoidance of passive smoke exposure, reducing day-care attendance and immunisation (pneumococcal and influenza).

Discharging otitis media – chronic suppurative otitis media

Introduction

Persistently discharging otitis media is most common in developing countries and certain high-risk populations in developed nations, such as Aboriginal Australians. It generally occurs following perforation of the eardrum from acute otitis media. It may also occur as a complication of tympanostomy tube (grommet) placement.

History

There is usually an absence of pain and a variable history (often weeks) of discharge that is purulent and offensive. Many authors require 6 weeks of discharge to fulfill diagnostic criteria. This condition is often recurrent.

Examination

The canal is usually non-tender, and there is usually no inflammation or some chronic inflammation. If the tympanic membrane can be visualised, usually only

after ear toilet, there will be a perforation or tympanostomy tube in situ.

Investigations

Investigations are largely unnecessary. If performed, the organisms found on ear swabs are most commonly *Pseudomonas aeruginosa* and *Staphylococcus aureus.*

Treatment

Ear toilet (using a dry tissue spear) and topical antibiotics, particularly quinolones (ciprofloxacin), have been demonstrated to be effective in acute resolution of otorrhoea. Systemic antibiotics alone are not as effective, and addition to topical treatment does not improve outcome.

Long-term outcomes are still to be determined. Patients should be referred for audiological evaluation and ear, nose and throat (ENT) specialist review.

Complications

Chronic perforation and discharge are the main issues, although hearing impairment may be a problem. Cholesteatoma occurs in a small proportion of affected children.

Otitis media with effusion

Introduction

Otitis media with effusion (OME) is the presence of a middle ear effusion in the absence of acute inflammation. It is unlikely to be a presenting complaint in an emergency department (ED) setting. It is more likely to be an incidental finding. It is extremely common, particularly in pre-school children. Its significance is in relation to its effect on conductive hearing.

History

Older children may present with aural fullness or reduced hearing. OME is usually asymptomatic in young children but may impact language and learning.

Examination

The eardrum may appear dull and non-erythematous, and the effusion is most easily recognised by the presence of bubbles or a fluid level. If these are not present, pneumatic otoscopy will demonstrate impaired mobility.

Investigations

No acute investigations are indicated. In persistent cases, referral for audiology is recommended to determine any significant hearing loss.

Treatment

As the majority of OME cases will resolve spontaneously (90% by 3 months) a period of observation is recommended. Persistent OME is more likely to follow acute otitis media in the first year of life.

Trials of many different treatments including antibiotics, nasal decongestants, nasal insufflation, and corticosteroids have failed to show benefit. Persistent OME with concerns of significant hearing loss is an indication for audiology and referral to an ENT surgeon for consideration of tympanostomy tubes (grommets).

Complications

The principal concern for persistent OME is conductive hearing loss and potential impact on language and cognitive development. There are potential long-term changes to the tympanic membrane and middle ear that may cause hearing loss (e.g. tympanosclerosis).

Mastoiditis

Introduction

Mastoiditis is the infection of the mastoid air cells. It is an infrequent illness with a rate of between 1.2 and 4.2 per 100,000 person years in developed nations. Presentation can occur at any age, with a median of 12–48 months. There is some evidence that decreased use of antibiotics for AOM has resulted in a small increase in cases of mastoiditis. However, it is estimated that approximately

5000 children with otitis media would need to be treated with antibiotics to prevent one case of mastoiditis.

History

Symptoms at presentation with mastoiditis are very similar to AOM, with pain, irritability and fever. Mean time from onset of illness to signs of acute mastoiditis has been reported as just over 4 days.

Examination

Examination findings differentiating mastoiditis from AOM include protrusion/displacement of the auricle, post-auricular inflammation and tenderness, and narrowing of the external auditory canal. The tympanic membrane is usually abnormal and may be perforated. On average, 80% of children have AOM at time of presentation.

Investigations

Increased routine use of computerised tomography scanning is due to the difficulty in diagnosis of subperiosteal abscess by clinical examination alone. Magnetic resonance imaging may also be valuable. Bacteriological diagnosis can be made at the time of operative treatment. Cultures show *Streptococcus pneumoniae, Strep. pyogenes* and *Staph. aureus* to predominate, but many organisms are possible. *P. aeruginosa* can be seen in chronic or recurrent cases.

Treatment

Management has historically been cortical mastoidectomy. However, a number of series report successful treatment in the majority of uncomplicated cases with myringotomy, intravenous antibiotics and insertion of tympanostomy tubes. Broad-spectrum antibiotics, such as third-generation cephalosporins are generally recommended, although antipseudomonal antibiotics may be required. Complicated disease will have similar management with inclusion of mastoidectomy.

Complications

Complication rates are significant (13–35%) and include subperiosteal abscess, facial nerve paralysis, sigmoid sinus thrombosis, epidural abscess and meningitis.

Trauma

Introduction

Paediatric ear trauma is an uncommon presentation to an ED. Accidental ear trauma is almost always unilateral. There is the usual male predominance, with a majority between 1 and 7 years of age. Accidental ear trauma is rare in the first year of life, and such presentations should be assessed for possible non-accidental (inflicted) injury. Other suggestive findings are bilateral ear injuries and associated injuries, particularly retinal haemorrhages and subdural haematoma.

History and examination

The most common mechanism is a fall, followed by blows from an object and self-inflicted penetrating injuries, which may result in perforation of the tympanic membrane. The most common objects inserted are cotton buds. Dog bites of the ear also occur. Lacerations are the most common injury, followed by bruising, abrasions and haematomas. Blood in the canal is a common finding in the penetrating injuries, making assessment of the eardrum difficult at the time of initial presentation. Burns are rare and are likely to be associated with more extensive burns. Barotrauma from explosions and loud noises are uncommon compared to industrial injuries in adults.

Assessment should include assessment of the facial nerve and hearing.

Investigation

Acute investigations are rarely required. However, audiology and ENT referral is indicated in penetrating and barotrauma injuries.

Treatment

Minor lacerations may be managed with steristrips, glue or suturing under local anaesthesia. Complex or larger lacerations will often require a general

anaesthetic and surgical repair. Haematomas can lead to cartilage necrosis, infection and chondritis or fibrous organisation. All of these have potential to cause significant deformity. Therefore removal of the haematoma is indicated. Smaller haematomas can be aspirated, and larger or recurrent collections should be evacuated. Appropriate contoured pressure dressings are then required to avoid re-accumulation. Penetrating injuries will often require ENT referral. Unless minor, burns will require evaluation by a burns specialist.

Complications

The main concerns are cosmetic deformity from haematomas, lacerations and burns, and hearing loss from penetrating injuries.

Controversies and future directions

1. Acute otitis media:
    - Antibiotic usage for acute otitis media in Australia, the UK and USA remains high. Due to concerns over the emergence of antibiotic resistance, the Netherlands has limited antibiotic use (31%), with good results leading to a review of use in high-prescribing countries. The concern about lower rates of antibiotic use is the potential for increased complications (see mastoiditis).
2. Discharging otitis media – chronic suppurative otitis media:
    - Concern has arisen with the use of potentially ototoxic antibiotic ear-drops in the presence of a tympanic-membrane perforation, despite routine use by ear, nose and throat specialists with few reported cases of ototoxicity.
    - A series of nine cases of iatrogenic topical vestibulotoxicity, all secondary to gentamicin-containing eardrops, have been reported. As a result, agents such as ciprofloxacin are being increasingly used.
3. Otitis media with effusion:
    - Ventilation-tube (grommet) insertion remains one of the most common surgical procedures performed in children.
    - It is effective at improving hearing in the short term. There has been conflicting evidence for its long-term benefit on

language and cognitive development.

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14.2

The nose

Loren Sher, and Laura Graley

ESSENTIALS

Rhinitis and sinusitis

1 Acute rhinitis is common and most commonly due to viral infections.

2 Mucopurulent discharge does not necessarily mean bacterial infection.

3 Antibiotics might be indicated in prolonged cases of nasal discharge (>10–14 days duration), indicating possible sinusitis.

4 Constant nasal discharge may be due to perennial allergic rhinitis.

Epistaxis

1 Epistaxis is common and largely responds to simple first aid – compression of the nose.

2 Severe cases are uncommon but are a medical emergency, requiring nasal packing and urgent ear, nose and throat (ENT) referral.

3 Underlying bleeding tendency should be excluded in cases with a family history of bleeding tendency or those with severe recurrent epistaxis.

Nasal trauma

1 Nasal trauma is common.

2 Most injuries will require little or no intervention. However, a careful examination to exclude septal haematoma is essential.

3 Septal haematoma requires urgent ENT referral.

4 Acute assessment can be difficult, and a review, once the swelling

has settled, is required to manage any deformity.

Rhinitis and sinusitis

Introduction

Rhinitis is inflammation of the mucosal lining of the nose, commonly caused by either viral infections or allergy. Sinusitis refers to inflammation of the sinuses. The proximity of the nasal passages and the sinus cavities, as well as the continuous respiratory mucosa, leads to frequent simultaneous involvement of both structures. Hence the term *rhinosinusitis* is frequently used to encompass inflammation of both.

The sinuses develop at different ages. The ethmoid and maxillary sinuses are present at birth, whereas the sphenoid sinus becomes pneumatized at 5 years old, and frontal sinuses appear at 7–8 years of age, although these are not completely developed until adolescence.

Viral upper respiratory tract infections causing rhinitis and inflammation of the upper respiratory tract are a frequent cause of febrile illness presentations amongst children to emergency departments. Children will commonly experience three to eight of these infections per year, even more if attending day care. After exclusion of more significant illness, education and symptomatic treatment are all that is required. Despite recommendations against such practice, many children will have been prescribed unnecessary antibiotics. Bacterial sinusitis has been estimated as complicating only 0.5–5% of upper respiratory tract infections.

Allergic rhinitis can begin at any age and is often under recognised. Environmental factors, such as pollution and smoking, especially when there is also a genetic predisposition, contribute to the development of acute allergic rhinitis.

Allergic rhinitis is more common in older children as opposed to viral rhinitis, which is more frequent in younger children.

History

The common cold usually commences with a throat irritation and clear, thin nasal discharge, eventually progressing to a thick, mucopurulent discharge after

a few days. The colour of the discharge is not indicative of the type of infection. Sneezing, cough, nasal obstruction and systemic features such as malaise and low-grade fever are common. Generally the duration is about 7 days.

Persistence of mucopurulent nasal discharge without improvement for longer than 10–14 days or more severe acute symptoms, such as high fever, headache, facial pain and/or swelling, are more indicative of bacterial sinusitis.

Persistent clear nasal discharge is more likely due to perennial allergic rhinitis. This may be associated with sneeze and itchy eyes and/or nose.

Unilateral nasal discharge, particularly offensive, is suggestive of a nasal foreign body.

Examination

In rhinosinusitis, nasal discharge is the usual finding. This may be clear or mucopurulent and is not indicative of cause. A red throat is a common association. In allergic rhinitis, the nasal mucosa will be pale and swollen.

Facial swelling, such as orbital/periorbital and/or facial tenderness, may occur in sinusitis. During the examination it is important to rule out possible serious complications of bacterial sinusitis.

Atopic children may give the ‘allergic salute’. This is in the form of a crease above the nasal bridge as a result of persistently rubbing their nose upwards from irritation. Allergic ‘shiners’ (purple–blue shadow beneath the eye) may also be present, especially if there is a history of severe or chronic allergy. Frequent throat clearing is frequently present.

Investigations

Investigations are not indicated for the common cold. Sinusitis is also usually a clinical diagnosis. Radiological investigations for acute sinusitis are difficult to interpret, as similar findings may be found in the common cold or even asymptomatic children. Additionally, the sinuses of young children are poorly developed, with frontal and sphenoid sinuses not appearing till 5–6 years of age. Radiological investigations should be reserved for those cases with suspected complications of orbital/intracranial extension or possibly in recurrent cases.

If aspirated under general anaesthesia, sinusitis fluid most commonly cultures *Pneumococcus*, followed by *Haemophilus influenzae*, *Moraxella catarrhalis* and then viruses. Nasopharyngeal cultures are not predictive and therefore not useful.

Treatment

Antibiotics are not indicated for the common cold and have been shown neither to alter the course of the illness nor prevent the development of complications.

Perennial allergic rhinitis responds best to allergen avoidance. Consideration for nasal or oral antihistamine and possible addition of intranasal corticosteroids can be used in children greater than 2 years. Nasal antihistamines can only be utilised in children over the age of 5 years old.

Even in bacterial sinusitis, the majority of cases will resolve spontaneously. There are no data to determine whether nasal decongestants are of benefit. If considered, they should not be used for longer than 3 days in order to avoid rebound nasal congestion. They should be avoided in children less than 6 years of age and used with caution in children between 6 and 12 years old.

Antibiotics should be considered if symptoms of rhinosinusitis last longer than 7 days or if there is purulent nasal discharge, sinus tenderness (particularly unilateral) or maxillary toothache. In addition, commence antibiotics if there are severe symptoms and high fever (39°C or higher) at the onset of illness and these symptoms last longer than 3 days or if there are worsening symptoms after initial improvement (‘double sickening’).

If antibiotics are used, amoxicillin is recommended as first-line treatment for 10–14 days duration. Amoxicillin + clavulanate can be reserved for those cases failing to respond in 48–72 hours.

Complications

Bacterial complications of the common cold, such as otitis media, do occur but cannot be prevented by antibiotic use. Asthma is frequently precipitated by upper respiratory tract infections.

Ethmoid sinusitis can spread causing orbital cellulitis. Intracranial extension can lead to abscess formation or cavernous sinus thrombosis.

Prevention

Breast-feeding has a protective effect on the number of upper respiratory tract infections experienced by young children. Day care attendance and environmental tobacco exposure increase the number of episodes.

Epistaxis

Introduction

Although epistaxis is common in childhood (affecting about 6–9%), it is usually mild and usually does not require any medical intervention. Epistaxis is rare in children younger than 2 years of age and should prompt consideration of trauma or serious illness, e.g. thrombocytopenia. In young babies non-accidental injury (NAI) should always be considered.

History

It is important to establish the frequency and severity of episodes, as well as first-aid measures that have already been undertaken. A history of abnormal bleeding/bruising and a family history of bleeding disorders should be elicited. Additionally, a history of infections, trauma (nose picking, blunt trauma), foreign body (especially if it is unilateral with foul-smelling nasal discharge), allergy, chronic rhinitis or use of anticoagulant/anti-inflammatory medications should be sought.

Examination

In the majority of cases, positive examination findings are limited to the nose. Bleeding can be unilateral or bilateral. Anterior bleeds are more common. Crusting and vessels in Little's area are commonly seen. Failure to identify a source may indicate posterior bleeding.

Application of a topical vasoconstrictor and/or anaesthetic agent can be considered in children 2 years and older to facilitate assessment.

A thorough systemic examination should exclude hypertension, abnormal bruising or petechiae, evidence of anaemia and findings of lymphadenopathy or hepatosplenomegaly. In the instance of trauma, visual acuity and extra-ocular movement should be tested. Evidence of NAI should be sought in children less than 2 years of age.

Investigations

Investigations are generally not required. The following should prompt investigation for a bleeding disorder: directly observed prolonged epistaxis

greater than 30 minutes despite active pressure, children less than 2 years old with no history of direct trauma, or a suspicion of bleeding diathesis on history or examination. A history of more severe epistaxis is suggestive of a bleeding disorder and warrants appropriate testing.

Treatment

First-aid treatment of epistaxis is firm compression of the alar nasae, applying pressure over Little's area for at least 5 minutes. This will result in cessation of bleeding in the majority of cases within 5–10 minutes. The use of anaesthetic/vasoconstrictor spray or soaked pledgets may assist in haemostasis. If these measures fail, nasal cautery (chemical or electrical) can be used for a well-identified bleeding site or an anterior nasal pack inserted. Purpose-made self-expanding nasal packs have made this procedure much easier.

Children with epistaxis that require nasal packing should be considered for investigation of underlying bleeding problems. A review is required within 48 hours, and an ear, nose and throat (ENT) referral should be made. Posterior nasal packing or other more invasive procedures are rarely required in children but may be needed in severe epistaxis in the context of resuscitation and urgent ENT consultation.

Complications

Rarely, recurrent epistaxis is sufficient to cause anaemia. Care should be taken when applying nasal cautery to avoid damage to the underlying septal cartilage. Bilateral cautery should be avoided for this reason.

Nasal trauma

Introduction

Nasal trauma is common in children. It can occur at any age but is most frequent in early primary-age groups. Like most injuries, there is a male predominance.

History and examination

The most common mechanism of injury is a fall, usually at home, followed by sports injuries. Child abuse (inflicted injury) should be considered, particularly

in the younger (non-mobile) child. A careful history is therefore required.

The types of injury are varied, from abrasions and lacerations of the soft tissues to fractures, dislocations and cartilage damage. More than minor epistaxis is unusual. Due to swelling it is often difficult to make a thorough assessment of the extent of injury in the acute setting – particularly for deformity. Examination must include an examination of the nasal cavities. Deviation, swelling and/or discolouration of the nasal septum may indicate a septal haematoma, and urgent ENT consultation for evacuation is indicated. Other facial injuries, including orbital injuries and frontal sinus fractures, should be actively excluded on examination.

Investigations

Nasal X-rays offer no benefit. A young child's nose is predominantly cartilage, and even when a fracture of the nasal bones is identified, this information does not influence management. Facial X-ray or CT should be limited in cases where more serious facial injury is suspected.

Treatment

Most nasal injuries that have minimal swelling and deformity and no septal deviation or haematoma require no acute intervention beyond analgesia. In some cases, where there is more severe swelling or suspicion of deformity/underlying fracture, specialist follow-up will be required. The assessment of the extent of the deformity should be done 3–5 days later once the oedema has resolved so that appropriate timely intervention can take place. After 7 days the fracture elements become difficult to mobilise.

Complications

Septal haematoma with or without abscess formation is the most serious complication that causes long-term damage and is therefore a concern. It may present later down the line and may have been missed on earlier assessment. It is therefore important to advise the family of the typical symptoms of an evolving nasal septal haematoma/abscess. These include progressive nasal obstruction, persistent or worsening pain, rhinorrhoea and fever.

On its own, a septal haematoma can cause nasal cartilage damage, including

aseptic necrosis. However, the development of an abscess is almost always associated with cartilage damage and potentially severe nasal deformity. Suspected septal haematoma/abscess requires urgent specialist referral for drainage. Meningitis or cavernous sinus thrombosis may also complicate a septal abscess. *Staphylococcus aureus*, *Streptococcus pneumoniae* and *Strep. pyogenes* have all been cultured.

Controversies

Epistaxis

The choice of which children to test and extent of investigation for bleeding disorders is still uncertain. For the severe and recurrent cases, thorough coagulation testing, including for von Willebrand disease, is appropriate.

Nasal trauma

Open rhinoplasty for nasal deformities may affect nasal growth in itself and should be used as conservatively as possible.

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14.3

The mouth and throat

Loren Sher, and Laura Graley

ESSENTIALS

Stomatitis

1 Acute herpetic gingivostomatitis is the most common cause of stomatitis in young children.

2 Early treatment with aciclovir is effective if commenced in the first 72 hours.

3 Aphthous stomatitis or ulcers are more common in young adults but do occur in children.

Pharyngitis/tonsillitis

1 Most sore throats in children are due to viral infections.

2 Bacterial tonsillitis is more likely in older children with isolated sore throat, high fever and tender cervical lymphadenopathy.

3 Antibiotics should be reserved for high-risk patients.

Peritonsillar abscess

1 Peritonsillar abscess is the most common abscess of the head and neck.

2 Diagnosis is not always straightforward.

3 Management is analgesia, antibiotics, fluids and draining the abscess.

Post-tonsillectomy haemorrhage

1 Tonsillectomy remains a relatively common procedure.

2 Post-tonsillectomy bleeding occurs in about 0.1–3%, and, although

most may be managed conservatively, for some, transfusion and/or surgery are necessary.

Oral/dental trauma

1 Oral/dental trauma is common in children.

2 A careful examination of the orofacial structures is required.

3 Dental consultation is indicated for all but minor cases.

Oral/dental infection

1 Dental infections are the most common cause of facial cellulitis.

2 There are usually underlying dental caries, and tooth extraction is often required.

Stomatitis

Introduction

Acute herpetic gingivostomatitis is the most common cause of stomatitis in young children (1–3 years). It is also the most common clinical presentation of primary herpes simplex infection in young children. It can also occur in older children and adults. Untreated, the course of the illness is 10–14 days. Aphthous ulcers usually occur as single ulcers and are often recurrent.

History

The incubation period is usually 1 week, and the contact case is often asymptomatic. It normally has a prodrome of 4 days – initially presenting with fever and irritability. Parents may confuse this with teething. Mouth pain, often severe, plus drooling and refusal to eat will usually follow. Dehydration can occur if the child refuses to drink. Gingival bleeding may occur. Mild lesions should resolve in a week, but healing may require up to 3 weeks.

Aphthous ulcers present as recurrent painful lesions of the oral mucosa, usually single and less than 1 cm diameter.

Examination

Early herpetic lesions are vesicles but may not be seen due to early rupture.

Multiple ulcers up to 1 cm then occur on any part of the oral mucosa and are initially covered with a yellow–grey membrane. The lips and perioral skin are affected in approximately two-thirds of patients. Associated gingivitis is usual.

Investigations

The diagnosis is clinical. Viral swabs and immunofluorescence or viral culture for herpes simplex can confirm the diagnosis if required.

For recurrent aphthous ulcers, checking a neutrophil count to exclude cyclical neutropaenia is appropriate.

Differential diagnosis

Initial presentation is non-specific and can be confused with a general viral infection. If the tonsils are involved early, acute tonsillitis or herpangina may be suspected. Some cases are misdiagnosed as oral candidiasis. An important condition to also consider is Stevens–Johnson syndrome.

Aphthous ulcers are easily distinguished, as these are usually single. Recurrent aphthous ulcers can be seen in cyclical or congenital neutropaenia and periodic fever adenitis pharyngitis aphthous ulcer (PFAPA) syndrome (fever, malaise, aphthous stomatitis, tonsillitis, pharyngitis and cervical adenopathy). Any ulcer that lasts beyond 3 weeks is unusual.

Treatment

Traditionally, treatment has been symptomatic, in the form of analgesia and hydration. Analgesia usually requires a combination of oral analgesics, such as paracetamol or ibuprofen. Topical lidocaine (lignocaine) gel can be very effective, but its use is controversial. Clinical studies have failed to show clear benefit, there is potential for toxicity and there is no standardised regime for administration. Topical healing methods such as rinsing the mouth with salt solution, applying wet tea-bags, topical steroids and tetracycline suspension rinses are commonly advocated, but without firm evidence.

If there is still insufficient oral intake after adequate analgesia, the child may require nasogastric or intravenous fluids for rehydration.

Studies have demonstrated the efficacy of oral aciclovir, if commenced in the first 72 hours of the illness. They have shown a significant reduction in duration

of fever, feeding difficulties and viral shedding. This should be considered in severe cases requiring admission.

Aphthous ulcers with an adherent/dental base can be treated with topical corticosteroids (e.g. triamcinolone).

Complications

Acute dehydration has been mentioned. Secondary bacterial infection is uncommon. Primary herpetic infection may progress to generalised vesicular eruption. Autoinoculation can also occur, particularly to the eye.

Recurrent labial herpes (cold sores) is common and more of an inconvenience. It occurs following exposure to sunlight, stress, trauma or cold. Topical aciclovir may be of use when applied with the first evidence of symptoms.

Pharyngitis/tonsillitis

Introduction

Sore throat is an extremely common presentation and is predominantly viral in cause. Group A streptococcal infection (GAS) accounts for 0–40% of cases dependent on setting and is more prevalent in school-aged children (5–15 years of age). The concern over potential complications from streptococcal infection, such as rheumatic fever and glomerulonephritis, has led to many children being prescribed unnecessary antibiotics.

History

Older children will present with a complaint of sore throat, while younger children may be non-specifically unwell. A reluctance to take food or drinks may indicate a sore throat. Associated symptoms include fever, headache, vomiting and abdominal pain but are not predictive of either bacterial or viral aetiology.

Examination

Ulcerative pharyngitis (herpangina) is a helpful finding indicating viral infection, such as Coxsackie. Otherwise, it is difficult to differentiate between bacterial and viral causes. Clinician judgment and Centor score are inadequate tools for clinical decision making for children presenting with sore throat. Features that

suggest bacterial infection are tender cervical lymphadenopathy and absence of other symptoms such as coryza, cough, conjunctivitis and diarrhoea. Tonsillar exudate is not an accurate predictor of bacterial infection and is frequently seen in viral causes such as Epstein–Barr virus (EBV) and adenovirus. EBV infection may be suggested by the presence of more widespread lymphadenopathy, particularly if there is associated splenomegaly. Palatal petechiae can be seen in GAS and EBV infection.

Scarlet fever is suggested by a widespread, fine, maculopapular rash with a sandpaper-like feel, with associated pharyngitis and possibly a ‘strawberry-tongue’. Although the scarlatiniform rash is highly specific for streptococcal infection, it only occurs in a minority of cases.

Investigations

Throat swab and culture may be used to assist differentiation between viral and bacterial aetiology. Many children are normal carriers of GAS. Swabs should only be performed in high-risk patients (indigenous communities, previous acute rheumatic fever, immunosuppressed and suspected scarlet fever) or those patients being considered for antibiotic treatment. Adjunctive point-of-care (POC) testing may provide sufficient accuracy to guide antibiotic prescription on first presentation in settings where GAS infections are more common.

Blood tests are usually done for investigation of infectious mononucleosis, although in children, the monospot/monotest has a high false-negative rate, and serology is more reliable.

Treatment

For the majority of sore throats due to pharyngitis/tonsillitis, no antibiotics are necessary. Antibiotic treatment shortens the duration of symptoms from GAS infection by only 16 hours. When weighing the risk benefits of antibiotic treatment, considering the extremely low prevalence of non-suppurative complications of GAS in Australia, antibiotics should be reserved for high-risk patients. Analgesia in the form of paracetamol or ibuprofen will provide symptomatic relief. For patients where antibiotic treatment is indicated, phenoxymethylpenicillin twice daily for 10 days is recommended.

Complications

Complications from sore throats are uncommon. Suppurative complications of streptococcal infection include peritonsillar abscess, sinusitis and otitis media. The principal concerns, however, are with the non-suppurative complications: rheumatic fever and glomerulonephritis. Acute tonsillitis can cause airway obstruction, particularly with pre-existing tonsillar hypertrophy, and may even warrant admission for monitoring.

Peritonsillar abscess

Introduction

Peritonsillar abscess is the most common deep space head and neck infection in children. It is likely to be an extension of acute tonsillitis. However, it has been suggested that some cases may arise from obstruction and infection of Weber's glands (mucous glands located in the superior tonsillar pole). It can occur at any age; however, it presents more frequently in adolescents.

History and examination

Presentation is usually with unilateral sore throat or neck pain, which is often severe. Odynophagia, dysphagia, and fever are also common. A muffled (or hot potato) voice, trismus and ipsilateral ear pain help to differentiate peritonsillar abscess from severe pharyngitis/tonsillitis. Only a minority will have a prior history of tonsillitis (20–30%).

Examination findings include cervical lymphadenopathy, unilateral tonsillar erythema, bulging of the superior aspect of the tonsil and uvular deviation to the opposite side. The differential is peritonsillar cellulitis, where there is no abscess. Clinical differentiation between the two is difficult. Bilateral peritonsillar abscess is rare.

Investigations

The majority of cases are diagnosed clinically with definitive confirmation on needle aspiration of pus. CT scan may assist when the diagnosis is uncertain and other deep neck infections are being considered. Microbiological identification of abscess contents probably does not alter management. If performed, the predominant organism found is *Streptococcus pyogenes.*

Treatment

Initial treatment is rehydration, analgesia and antibiotics (penicillin). Acute drainage is generally recommended, with intra-oral drainage, abscess tonsillectomy or needle aspiration. Needle aspiration alone would appear to be effective in a majority of cases (>90%). This should be performed by an appropriately trained specialist owing to the risk of complications, including puncture of the carotid artery.

Abscess tonsillectomy should be reserved for recurrent tonsillitis abscess, but many of these can still be managed with aspiration and delayed tonsillectomy.

Complications

Dehydration is common and will require medical intervention. Uncommon but dangerous complications include infection extension to the parapharyngeal space, airway obstruction and aspiration of pus causing pneumonia.

Post-tonsillectomy haemorrhage

Introduction

Tonsillectomy remains a very commonly performed procedure. The most common complication of tonsillectomy is bleeding. Primary haemorrhage occurs within 24 hours of surgery and occurs in 0.2–2.2 per cent of patients. Secondary (delayed) haemorrhage occurs after the first 24 hours following surgery and complicates about 0.1–3% of tonsillectomies. These patients tend to present 5–10 days postoperatively. Haemorrhage is caused by premature separation of the eschar. This is usually associated with infection or dehydration.

Risk factors include bleeding tendencies and patients who have a history of chronic tonsillitis, preceding surgery.

Rarely, haemorrhage is catastrophic and life threatening.

History

Bleeding is usually obvious, although occasionally will be swallowed and not immediately apparent. A history of bleeding or bruising tendency should be sought although ideally will have been identified prior to surgery.

Examination

Initial assessment should focus on signs of shock or haemodynamic compromise. Subsequent examination of the tonsillar fauces for evidence of active bleeding is then important.

Investigations

Full blood count examination to monitor for a drop in haemoglobin and taking blood for cross match is recommended in any significant post-tonsillectomy bleed. Coagulation profile testing rarely demonstrates abnormality but is warranted in major haemorrhage.

Treatment

Postoperative bleeding often stops spontaneously. Small recurrent haemorrhages increase the risk of a major bleed. Patients with small haemorrhages should therefore be discussed with an ENT service and admitted.

Patients with active haemorrhaging should be managed in a high acuity area in an ABC approach. They require immediate, preferably large bore intravenous (IV) access, and blood should be sent for full blood count (FBC) and cross match.

Patients may require fluid resuscitation. Transfusions have been reported as being required in 10–12% of patients with secondary haemorrhage. Early involvement of an ear, nose and throat (ENT) surgeon for children with active bleeding is indicated. Severe bleeding will require removal of the clot to allow application of adrenaline (epinephrine)-soaked gauze (1:10,000) directly onto the bleeding point. The gauze is held in place with Magills forceps, and pressure is applied laterally to the wall of the mouth. This should be continued until bleeding ceases or specialist help arrives. In many instances antibiotics will be commenced and should be guided by local practice.

Complications

Post-tonsillectomy bleeding is potentially life threatening, largely from hypovolaemic shock. Massive bleeding may also cause airway obstruction.

Oral/dental trauma

Introduction

The most common reasons for infants and children to present to the emergency department (ED) with oral/dental trauma are falls and sporting injuries. Most of these are relatively minor. Occasionally, higher impact trauma including motor vehicle accidents will produce severe injury requiring maxillofacial reconstructive surgery and early problems of haemostasis and airway compromise.

History

Make specific enquiry with regard to the time and mechanism of trauma and the nature of the dentition prior to injury. This includes the number of deciduous teeth that were present and the presence of any secondary dentition. Determine whether any teeth have been avulsed and their current location and method of storage. Consider the possibility of aspiration if a tooth is missing.

Examination

The physical examination should review all orofacial structures with careful scrutiny, digital palpation and observation of normal function. Externally and internally the orofacial region should exhibit consistent symmetry, and any departure from this, be it an area of altered facial soft tissue architecture associated with a swelling or altered bony architecture associated with a fracture, will be important. For example, chin point trauma is a common injury associated with mandibular condylar fracture. Assessment of mandibular opening and malocclusion is vital. In cases where the child has fallen with an object in his/her mouth, careful assessment for palatal or pharyngeal penetrating trauma is required.

The key points in recognition of a facial fracture are pain, facial swelling, stepping (of the bony border), limited jaw opening, palatal or sublingual haematoma, malocclusion and paraesthesia. The need for tetanus prophylaxis and antibiotics should be considered. Usually, antibiotics (penicillin and metronidazole) should be given for a compound fracture in the mouth.

A common error when assessing a child’s occlusion is to fail to recognise that an anterior open bite (associated with a prior digit or object-sucking habits) is a

normal anterior relationship for that child and not necessarily evidence of a fracture in the maxilla/mandible. Check the posterior teeth for appropriate occlusion in this situation. The older child will usually be able to tell you if the teeth ‘feel right’ when he/she bites.

Investigations

The most relevant extra-oral radiograph is an orthopantomogram (OPG). This radiograph will give an excellent view not only of the body of the mandible, the dentoalveolar area, but also of the temporomandibular joints (TMJ). However, the child’s cooperation is required. If a TMJ view is specifically required it will be important to notify the radiographer. A chest X-ray (CXR) should be performed if there is a possibility of an aspirated tooth.

Treatment

Management will depend upon the exact diagnosis, which may include the following six factors:

Luxation

This is defined as a slight movement of the teeth as a result of trauma, with or without associated gingival bleeding. These injuries can occur in either the primary or permanent teeth and can be in any direction, i.e. internal, external or lateral. Active intervention may or may not be needed, depending on the extent of movement and interference with the occlusion or risk to the development of the underlying secondary dentition.

Minor luxation may be treated conservatively or involve extraction of the damaged primary teeth. Repositioning of displaced permanent teeth may be required with placement of a dental splint and associated suturing of damaged soft tissues.

Avulsion

This is defined as the complete loss of a tooth, primary or permanent, and is considered a dental emergency. Permanent teeth are always re-implanted, and re-implantation within 15 minutes by a competent provider allows for the best chance of survival. If obviously soiled, the tooth should be rinsed briefly in cold water/saline (less than 10 seconds). It is crucial not to handle the root of the

tooth. Manipulate using the crown only. This avoids damage to the periodontal ligament cells, which line the root surface and are critical in re-establishing the tooth back in the mouth as a functioning unit. Once in place the teeth will need to be secured by a temporary dental splint. Options include molded aluminium foil or use of an emergency dental kit. The patient should have his/her tetanus status assessed, and all patients require antibiotics and referral to a dentist.

If immediate re-implantation is not achievable or there is any doubt about the nature of the tooth (that is, whether it is primary or not), the avulsed tooth should be placed in milk. Saliva or saline is a reasonable alternative. The tooth can survive up to 6 hours in milk but only 1 hour in the alternative solutions. Water should be avoided as it accelerates cell death in the periodontal ligament. At a minimum it is crucial to keep the tooth moist by wrapping it in cling film or a gauze that is kept moist with saline or the patient's saliva.

As a general rule, the primary teeth are not re-implanted. Reasons include the potential damage to the developing permanent teeth, difficulties in securing the tooth/teeth in place and the level of cooperation that is required. Patients should be referred to a dentist for formal management.

Intrusion

This occurs when a tooth is pushed up into the socket. It warrants urgent evaluation by a dentist as intruded teeth may require surgical or orthodontic repositioning depending on the degree of intrusion.

Root fractures

The root of the tooth, which is buried in the alveolar bone, can also be fractured, often with minimal damage to the crown of the teeth. This may present at review, if not initially obvious. A particularly mobile or loose tooth/teeth that are slightly extruded are classic signs of associated root fractures.

Hard tissue injuries

This involves a chip off the tooth, whether it is just enamel or involves the deeper layers – such as the dentine. It may involve all three layers of the tooth and 'expose' the dental pulp. If a tooth is exposed, then it is important that the exposure is treated immediately so as to preserve the vitality of the dental pulp and thus enable the root to grow to its full length and thickness. This is very important for the tooth's long-term maintenance as a useful member of the

dentition.

Soft tissue injuries

The soft tissues of the mouth involve the gingival tissues, mucosa and muscle. Soft tissue lacerations, especially of the gingival tissues, need to be carefully assessed for degloving which requires operative repair. Where teeth are significantly displaced there will be associated displacement and laceration of soft tissue, and it is very important for the long-term periodontal health of the traumatised teeth that displaced soft tissues are adequately repositioned and sutured. Alveolar bone should not be left exposed to granulate over.

Prevention

Advice on prevention is never misplaced, and recommendation of the use of well-made and fitted mouth guards is encouraged.

Oral/dental infection

Introduction

Dental infections are the most frequent cause of acute facial cellulitis, accounting for about 50% of all cases. Therefore, for children presenting with a facial swelling associated with fever and general malaise, a dental cause must be high on the list of differentials.

History

Facial cellulitis will present as above, with facial redness and tenderness. The upper face is affected more frequently, particularly in younger children. The lower face is affected relatively more often in older children, probably related to caries patterns.

Examination

A thorough examination, particularly intra-orally, is required, and early dental involvement is very helpful in determining a dental source.

Investigations

Appropriate radiographs are frequently indicated to determine a dental aetiology. These may be extra-oral, e.g. an OPG and/or intra-oral.

Treatment

The usual treatment involves the use of antibiotics (usually penicillin will be adequate) and repair or removal of the offending tooth/teeth. Metronidazole may be added in cases of significant infection requiring hospitalisation.

Prevention

A significant cause of tooth decay in infants and young children is inappropriate use of night-time feeding bottles containing sweetened liquids. This results in extensive dental damage. Advice should be given to parents to go from breast to cup with their babies or to wean from the bottle by 12 months. Infants should not be put to bed with a bottle.

Other dental issues

Spontaneous oral haemorrhage

Oral bleeding is still a very important sign for the diagnosis of underlying generalised bleeding/platelet disorders. It is important to determine the actual site of the bleeding, and consequently a through oral examination will be required. Try to rinse the child's mouth with water or saline, and use gauze to remove any blood clots and identify the source of the oral bleeding. In many instances this may be a tooth socket associated with a recent extraction. However, life-threatening disorders such as haemangiomas and arteriovenous malformations may present in the same fashion.

If a tooth-extraction socket is identified as the cause of bleeding, local measures to control the bleeding will usually be sufficient. This involves applying digital pressure to the bleeding socket or having the child bite down on a gauze pad for 15 minutes. Bleeding disorders such as von Willebrand disease and haemophilia should be considered in cases that continue to bleed.

Controversies

Pharyngitis/tonsillitis

Although antibiotics have a protective effect in the prevention of rheumatic fever, the evidence does not support a similar effect on prevention of glomerulonephritis.

Tonsillectomy

Antibiotics post-tonsillectomy do lessen morbidity, in particular pain, analgesic use and delay in resumption of oral diet. Amoxicillin and amoxicillin + clavulanate have been shown to be effective. The studies have not had the numbers to demonstrate whether this is also true for post-tonsillectomy haemorrhage.

Oral/dental trauma

1. The management of avulsed permanent teeth centres on the prescription of routine antibiotics post re-implantation in an effort to limit the unwanted side effects associated with re-implantation, such as an ankylosis of the teeth and inflammatory root resumption. This prescription of antibiotics is not universally accepted.
2. The immediate disimpaction of the traumatically intruded permanent tooth/teeth using orthodontic appliances is proposed as the most appropriate care, especially when trying to limit the unwanted sequelae of poor alveolar bone height and gingival attachment. However, there is a school of thought that proposes to partially disimpact surgically and then complete the job of realignment orthodontically.

Oral/dental infection

Antibiotic choice is not universally agreed in either delivery (oral vs. intravenous) or type (penicillin vs. cephalosporins).

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14.4

Retropharyngeal abscess

James Tilleard

ESSENTIALS

1 Ensuring a safe and secure airway is the immediate treatment priority.

2 Retropharyngeal abscess should be considered in children with fever, dysphagia and restricted neck movement.

3 Early antibiotic treatment will minimise the risk of serious complications.

4 Management should be in a hospital equipped to manage the potential complications with paediatric medical; ear, nose and throat; intensive care and anaesthetic services.

Introduction

Retropharyngeal abscess is an uncommon but important paediatric infection. The risk of airway compromise is the most important complication for immediate consideration. The retropharyngeal space lies between the pharynx anteriorly, the prevertebral fascia posteriorly and is contiguous laterally with the parapharyngeal space. The retropharyngeal space contains lymph nodes that have the potential to become inflamed with progression from adenitis and cellulitis to phlegmon and finally abscess formation. This sequence may follow upper respiratory tract infections and less commonly pharyngeal trauma or infection of adjacent structures. Retropharyngeal, parapharyngeal and peritonsillar abscesses are known as deep space neck infections (DNI).

Retropharyngeal abscesses are frequently polymicrobial. Causative organisms include *Streptococcus pyogenes*, *Staphylococcus aureus* (an increasing incidence of MRSA is reported), anaerobes and other respiratory pathogens.

Incidence is highest in children aged less than 5 years with boys more commonly affected in all age groups and a winter-spring seasonality. Presenting symptoms of retropharyngeal abscess are variable, and diagnosis may be difficult especially early in the clinical progression. Early diagnosis and treatment are essential to the prevention of complications.

History

Children with retropharyngeal abscess may present with a history of recent upper respiratory tract infection or less commonly trauma or other infective focus of the head and neck. Typical symptoms include sore throat, fever, odynophagia and torticollis. Presentations may also be subtle with irritability, headache, decreased oral intake, dysphagia and reduced neck movement. More advanced presentations may have systemic toxicity, drooling, voice change, chest pain, trismus or obstructed breathing.

Examination

Examination may be non-specific especially early in disease progression. Visible or palpable external swelling is unusual, but cervical adenopathy may be found, and swelling of the posterior pharyngeal wall may occasionally be noted. Restricted neck movement, torticollis and fever may be present. Eliciting pain on moving the larynx from side to side (tracheal rock manoeuvre) is a non-specific sign. Signs of complications, including airway obstruction, mediastinitis, thrombophlebitis, carotid artery aneurysm, aspiration pneumonia and signs of systemic sepsis may also be present.

Investigation

Contrast CT imaging is the investigation of choice if retropharyngeal abscess is suggested clinically. CT establishes the diagnosis, may reveal complications and directs treatment toward medical or surgical options on the basis of detection of a drainable collection. Prior to investigation it is essential to ensure the patient is stable and safe with particular attention to the airway.

Lateral soft tissue neck X-rays have poor sensitivity and specificity but may reveal prevertebral swelling, gas, foreign body, air fluid level, bony erosion or evidence of an alternative disease process. Ultrasound requires experience for interpretation, may not define deep structures and is operator dependent. MRI when available has the advantage of excellent soft tissue resolution and avoids radiation, but duration of the procedure and isolation are problematic in the paediatric population especially considering the potential for airway or other complications.

Laboratory studies are typically consistent with infection revealing raised white-cell count and elevated inflammatory markers. Microbiology specimens including blood cultures, throat swabs and pus gained at the time of surgery may guide subsequent antibiotic therapy.

Treatment

The initial treatment priority is to ensure a safe and secure airway. If the airway is compromised or at risk, it should be secured, ideally in the operating theatre with both anaesthetic and ear, nose and throat (ENT) support.

Once initial resuscitation is complete antibiotics should be commenced as soon as possible. Recommended empiric antibiotic regimens vary but should cover the most likely organisms as above. An appropriate initial choice would be piperacillin-tazobactam in the absence of penicillin allergy or cefotaxime ± metronidazole with the addition of vancomycin if there is concern about MRSA. Early infectious diseases consultation is recommended with treatment modified according to advice and results of microbiological testing.

Subsequent management may be medical, with antibiotics and close observation for the development of airway or other complications, or surgical. This decision is made in consultation with the ENT service, and surgery is more likely if CT reveals an organised abscess. Children with retropharyngeal abscess are best managed in larger centres equipped with appropriate paediatric medical, intensive care, ENT and anaesthetic services.

Controversy and future directions

Current debate revolves around the criteria for surgical or medical management, the role of steroids in reducing inflammation and appropriate antibiotic selection especially considering the incidence of MRSA.

Two national level studies of DNI in the United States have shown a

significant increase in the incidence of retropharyngeal abscess.

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14.5

Foreign bodies and caustic ingestion

Loren Sher, and Laura Graley

ESSENTIALS

Nasal foreign bodies

1 Foreign body removal from the nasal passage requires good preparation and lighting.

2 The appropriate method of removal depends upon the type of object and its location.

Aural foreign bodies

1 Foreign bodies in the medial two-thirds of the ear canal are difficult to remove and frequently require sedation or general anaesthesia.

Caustic ingestion

1 Caustic ingestion is usually accidental in children.

2 The children are often asymptomatic and are at minimal risk of complications such as stricture.

3 Airway management is the priority in resuscitation.

4 There is no proven treatment to prevent stricture formation.

Nasal foreign bodies

Introduction

Nasal foreign bodies are a relatively common presentation to a paediatric emergency department (ED), although there is little published literature on the

subject. They are not life threatening but can cause significant morbidity.

Button batteries and paired disc magnets can cause significant damage to nasal structures and require urgent removal.

History

Nasal foreign bodies predominantly present in children from 2 to 5 years of age. Children with nasal foreign bodies are usually brought in within 24 hours of insertion/impaction. Delayed presentation is mostly with unilateral purulent nasal discharge, which is usually offensive. Other possible symptoms include foul breath, nasal pain, sneezing, snoring and mouth breathing.

Examination

The type of foreign body is extremely variable, although it is most commonly a plastic toy or bead. The foreign body is usually readily visible. It is essential to examine both nostrils and ears to ensure that no other foreign bodies are present.

Investigations

Investigations are not usually required. Imaging is usually indicated for other possible causes such as tumour.

Treatment

Except for button batteries (see also Chapter 7.6) and paired magnets, removal of the foreign body is not urgent. Most foreign bodies can be successfully removed in the ED with adequate preparation and planning. Concern for migration of an inert nasal foreign body with aspiration into the trachea is unlikely in a healthy patient with intact airway reflexes. There is a greater risk of aspiration when extraction is attempted by inexperienced clinicians or when there is lack of preparation for the procedure.

Prior to the attempted removal, a topical anaesthetic nasal spray, such as lidocaine (lignocaine) plus phenylephrine, is recommended. Sedation and/or appropriate restraint of the child may be required. A good light source and an assistant to hold the child's head still are essential.

The appropriate procedure for removal will depend upon the foreign body size, shape, consistency and location in the nares. These include the following:

- Right-angle hooked probes can be passed alongside and past larger objects, rotated and then drawn back gradually, removing the object.
- Forceps (alligator or bayonet) are useful for smaller objects near the anterior nares.
- Positive pressure ventilation using bag–valve–mask is effective, particularly for larger smooth objects that occlude the nasal cavity. An alternative to a bag–valve–mask is for the parent to blow into the child's mouth with his/her own. Air is 'bagged' or blown rapidly through the mouth while the unaffected nostril is occluded with a finger. The high pressure generated in the upper airway expels the foreign body from the nose. There is an unreported theoretical risk of barotrauma.
- Suction catheters can be used. Generally, a soft, pliable end is required, and this can be made up using soft tubing, although commercially available kits are now available.
- A balloon catheter can be used if passed beyond the foreign body, inflated and drawn back. This technique is best suited to small round objects that are difficult to grasp with forceps.
- Application of cyanoacrylate glue via a wooden swab stick or similar is described but requires patience and a very steady hand. It may adhere to the mucosa.

General anaesthesia and removal by ear, nose and throat (ENT) staff is required for the failed removal and the uncooperative child.

Complications

Possible complications include epistaxis and local and more widespread infection. Uncommonly, septal perforation has occurred secondary to a button battery. Mucosal injury may occur within hours.

Aural foreign bodies

Introduction

Aural foreign bodies in children are also a relatively common presentation to EDs. Again morbidity can be significant.

History

A greater proportion of children present beyond 24 hours from insertion compared with nasal foreign bodies, and in many cases the time frame may not be determined. It may be a result of the event being witnessed, reported by the child due to irritation or pain, or as an incidental finding. The object may have been placed due to existing ear canal irritation due to wax impaction or otitis. Live insects are not uncommon and cause marked discomfort requiring more urgent attention. The use of cotton tips or cotton wool in the external ear canal and silicone ear plugs should be discouraged.

Examination

Visualisation of the foreign body is usually by otoscopy. The type and location of the object have a significant role in the difficulty of removal. Both ears and nostrils should be examined for other foreign bodies.

Investigation

Investigations are not indicated.

Treatment

Removal of foreign bodies from the lateral one-third of the external auditory canal is much easier and more successful than from the medial two-thirds. The latter is the osseous portion and is narrower, more vascular and very tender. Therefore, removal is more likely to require sedation or general anaesthesia and ENT expertise.

- Adequate preparation, appropriate restraint of the child and a good light source are always necessary. The first attempt at removal is critical as success rates markedly decline after the first failed attempt.
- Sedation may have a role in some children.
- Choice of method of removal will depend upon the object and its location.
- Irrigation is the simplest method but is contraindicated if there is a tympanic perforation or for soft objects or vegetable matter that may swell. This technique is not practised in the specialist setting with the

availability of magnified direct vision, microscopy, suction and specialised instrumentation such as wax hooks or curettes. Although often successful for inorganic material, syringing must be practised cautiously, particularly in the presence of irregular objects which may harm the tympanic membrane or organic material which may expand, irritating the canal and making later extraction difficult.

- Suction via a small catheter can be useful for friable foreign bodies as other techniques will tear the object. The noise, however, is often distressing to the child, and sedation is usually required.
- Alligator or Hartmann forceps can be used for irregular objects with an easily visible edge.
- A right-angled hook, passed beyond the object and pulled out, is particularly useful for smooth and spherical objects.

Button batteries are again a risk (see also Chapter 7.6), causing necrosis of the ear canal or tympanic membrane and should be removed as soon as possible. A live insect should be killed or immobilised with microscopic immersion oil, mineral oil or local anaesthetic solutions (2% lidocaine [lignocaine]). It can then be removed using irrigation or forceps. Putty is often very difficult to remove and may require otomicroscopic removal. Sharp objects also usually require ENT removal.

Failure to remove in the ED will require ENT referral. Inspection following removal is advised to ensure there is no persisting foreign body and to assess for trauma or inflammation. Aural antibiotic drops with steroid are often recommended, particularly if there is evidence of trauma or inflammation.

Complications

Trauma to the ear canal is common as a result of the foreign body and/or its removal. More seriously, trauma to the tympanic membrane or ossicles can occur. Otitis externa can occur as a result of trauma.

Caustic ingestion

Introduction

Caustic ingestion in children is an uncommon presentation, and significant injury with stricture formation is rare because the volume ingested is usually

small. The ingested agents are variable and include alkalis (more common) and acids in liquid, granule or solid form. The likelihood of significant injury is dependent upon how alkali or acidic the agent is (i.e. its pH). Strong liquid alkali is particularly dangerous as it causes deeper tissue penetration and more extensive burns.

History

In children, in contrast to adults, ingestion is largely accidental and, as a result, significant injury far less common. Most cases occur in children less than 3 years of age and, like accidents in general, occur in boys more frequently than girls. Most ingestions occur at the home of the child with commonly found household products. Button batteries have a significant corrosive and burn risk if lodged or impacted in the oesophagus (see Chapter 7.6).

Many children are asymptomatic. If they do occur, symptoms include vomiting, drooling, pain on swallowing and dysphagia, chest pain and refusal to drink. Less commonly, stridor may occur secondary to upper airway burns and obstruction.

In one series, the most predictive symptoms of significant injury and scar formation were prolonged drooling and dysphagia (100% sensitivity and 91% specificity). In another, haematemesis and respiratory distress were always associated with severe injury.

Examination

Many children will have normal examination findings. Oral burns may be demonstrated by inflammation, oedema or white areas. The presence or absence of oral burns is a poor predictor of oesophageal injury. Signs of upper airway obstruction warrant urgent early intervention.

Investigations

Chest X-ray has demonstrated pulmonary irritation in up to 13% in some series. However, there is evidence that radiological investigations have little impact upon management and therefore cannot be recommended routinely, except to localise a button battery.

Oesophagoscopy may be performed according to symptoms, particularly

ongoing drooling and dysphagia, vomiting/haematemesis and stridor. The type of caustic substance, its form and pH should be considered.

Treatment

Resuscitation, involving airway and breathing management, is the priority in severely affected cases. Upper airway obstruction may warrant intubation.

Dilution using water or milk is no longer recommended as it may induce vomiting, leading to further complications.

Symptomatic children should be referred to a specialist centre for endoscopic examination and further management. Children who remain asymptomatic may be discharged. Lodged button batteries require urgent endoscopic removal, followed by a period of observation.

There is no proven effective treatment for oesophageal burns, especially in the prevention of stricture development. Antibiotics and corticosteroids have been commonly used but have not been shown to be of any value.

Complications

Stricture formation is rare and, in a combination of nine series giving a total of 1961 children who had caustic ingestions, occurred in only 3.2%. This was despite a mean incidence of oesophageal burns of 21.4% and of deep burns of 4.7%. Upper airway obstruction has been mentioned and is potentially life threatening.

Prevention

Labelling, formulation, and child-proof containers and caps have proved effective in prevention, when legislated. Storage of strong caustics should be in locked cupboards or similar, and handling should occur out of the reach of children. More hazardous caustics are available in developing countries, and severe injury occurs more frequently.

Controversies

In caustic ingestion, acute oesophagoscopy has been advocated as critical to the early diagnosis and management of burns. This may help to assess the severity of injury and likelihood of stricture formation. However, there are

few data to determine the ideal timing for this procedure.

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SECTION 15

Obstetrics and Gynaecology

OUTLINE

15.1

Paediatric gynaecology

Sonia R. Grover, and Christine Brabyn

ESSENTIALS

Infants

1 Vaginal discharge in neonates can be physiological, usually resolving by 3 months of age.

2 Ensure normal external genitalia.

3 Labial fusion secondary to mild inflammation is a common presentation.

4 Vaginal bleeding in the prepubertal female is abnormal after 3–4 weeks of age.

Prepubescent girls

1 External examination with gentle labial separation usually reveals as much information as internal vaginal examination. Internal vaginal examination, where indicated, should be performed under anaesthetic. Pelvic ultrasound may provide further information.

2 Vulvovaginitis is the most common gynaecological problem in prepubertal girls, and in most cases a specific infectious cause cannot be identified.

Adolescents

1 Sexually transmitted infections among adolescents is increasing. Chlamydia and gonorrhoea testing should be considered in all sexually active patients <25 years of age.

2 Pregnancy-related conditions need to be considered for all adolescents with pelvic pain or abnormal vaginal bleeding.

3 Abnormal vaginal bleeding may be secondary to an underlying haematological condition.

Infant and prepubescent gynaecology

Vaginal discharge

Introduction

Vaginal discharge in neonates and prepubertal girls can be physiological. In neonates, a white mucoid vaginal discharge is present in most baby girls and is due to the effects of maternal oestrogens; it disappears by about 3 months of age. The second period where physiological discharge may occur is at the time ovarian activity commences with the onset of puberty.

Other conditions that cause vaginal discharge in the prepubertal girl include vulvovaginitis, lichen sclerosis, foreign body, eczema and pinworms.

Vulvovaginitis is the most common gynaecological problem in childhood, usually occurring in girls aged between 2 years and prior to the onset of puberty.[1–3] Symptoms include itch, offensive vaginal discharge, erythema, soreness, dysuria and bleeding. Consideration of urinary tract or bowel symptoms must also be considered when a child presents with vaginal or vulvar complaints. These symptoms may fluctuate in severity. The majority of vulvovaginitis in prepubertal children is nonspecific in a etiology. Contributing factors to vulvovaginitis include unoestrogenised thin mucosa, lack of labial development, poor hygiene, soaps, obesity, foreign bodies and choice of clothing (tights, leggings, leotards). In addition, lack of protective labial hair and fat pads and the presence of bowel flora, which is the normal flora in the atrophic vagina, may contribute to skin irritation and the other related common symptom of offensive smell.

Infectious vulvovaginitis is less common and is due to an overgrowth of one organism, for example group A streptococcus, *Staphylococcus*, enterococci and *Escherichia coli*. In this instance, profuse discharge is usually present with marked skin inflammation with demarcation that is often beyond the contact surfaces of the labia.[3] Isolation of an organism that has strong sexual transmission, such as *Neisseria gonorrhoeae* or *Chlamydia trachomatis*, generally indicates sexual abuse or sexual activity and therefore warrants further investigation.[3,4]

Lichen sclerosis is an uncommon condition of unknown aetiology. It may present in childhood with vulval irritation, pruritus, dysuria or bleeding.[1,2] Examination reveals pale atrophic patches on the labia and perineum. The patches can be extensive and coalesce and with scratching lead to chronic inflammation and purpuric haemorrhage into the skin. The condition usually persists with intermittent exacerbations. Most resolve before puberty, although some may continue to have problems into adult life.[5]

Eczema may contribute to the symptoms of vulvovaginitis with the addition of itch. In these cases eczema is usually present elsewhere on the body and can be superimposed on the irritation due to the discharge.

Foreign bodies are a potential cause for a persistent, unresolving, often bloodstained offensive discharge.

Candida is very uncommon in the prepubertal girl unless there has been significant antibiotic use or she is still in nappies.[2,4] (Thrush thrives in an oestrogenised environment, not in the atrophic setting). In this age group, recurrent or unexplained *Candida* requires exclusion of diabetes mellitus or other causes of diminished immune function.

History

A general medical history is required, with specific history of the symptoms and their duration including nature of discharge as well as any previous treatments. Also important is past history of urinary tract infection, encopresis, constipation, enuresis, the presence of skin disorders, and any other illness, including antibiotic use in the previous 4 weeks. Although a history of perineal hygiene (e.g. wipe front to back, frequency of bathing, type of underwear/clothes, specific irritants, such as bubble baths or use of feminine hygiene sprays in the adolescent population, etc.) should also be established, there is limited evidence to support the role of this in the pathogenesis of vulvovaginitis.[3,5] Where itch is the dominant symptom, pinworms should be considered and questions asked about family symptoms. In younger children itch may be difficult for them to describe, and they may present with discomfort/pain at night.

Although uncommon, the possibility of sexual abuse should always be considered in a child that presents with genital symptoms, and check for other signs that may be present including alterations in behaviour, such as phobias and eating or sleeping disorders.

Examination

A general examination including sexual development is required. Perineal, vulval and introital examination may be required for the above conditions. Attempts must be made to ensure it is not a traumatic event for the child. The perineum is best examined either with the girl supine with heels together and knees flexed and hips abducted or in the lateral position with knees drawn up to the chest.[6] Vaginal examination is inappropriate in paediatric patients and usually provides little further information. Specific external examination of the perineum usually reveals mucoid discharge and reddened introitus, particularly on the contact surfaces between the labia.

The presence of a profuse discharge or marked skin inflammation, especially if it extends beyond the contact surfaces of the labia, suggests an infectious cause. An offensive discharge can occur with vulvovaginitis or foreign body. A bloody discharge can occur with vulvovaginitis (particularly with *Shigella* or group A streptococci).[6] The presence of perianal excoriation suggests pinworm.

Investigations

Swabs are generally not required. If taken in mild cases, they usually reveal a growth of mixed coliforms.[3] If discharge is visible or profuse or marked erythema is present, introital swabs should be taken for culture. Vaginal swabs are painful and distressing and are not required.

If urinary symptoms are present, the urine (midstream urine [MSU]) should be checked to exclude urinary tract infection. A pinworm test may be considered if itch is a prominent symptom. This requires briefly placing clear sticky tape on the perianal skin in the morning and viewing under a microscope for eggs. Otherwise worms may be seen in the perianal region at night.

Pelvic ultrasound may provide further information, especially if there is a possibility of foreign body; if necessary an examination under anaesthetic may be appropriate.

Management

Vulvovaginitis

Management consists of the following:

1. Explanation and reassurance
2. Toileting/hygiene advice:
    - Avoidance of potential irritants such as soaps and bubble baths

and other causal factors, such as tight/synthetic clothing and wet bathers
- Vinegar (add one cup white vinegar in a shallow bath and soak)
- The use of a simple barrier cream to the labial area (e.g. zinc-castor oil or nappy rash cream).

Most non-specific vulvovaginitis resolves with these supportive measures within 2 to 3 weeks. The natural history of vulvovaginitis is for recurrences to occur up until the age when oestrogenisation begins. If a primary bacterial cause is suspected, cultures should be taken and treatment commenced with the appropriate antibiotics, e.g. initial amoxicillin, and adjusted when culture results are available. Alternative regimes include topical metronidazole or topical clindamycin.

A short course of oestrogen-containing cream (e.g. Premarin® cream [conjugated oestrogen]) can thicken the vaginal mucosa and make it more resistant to recurrent non-specific infections. It is always important to consider the differential diagnosis.[7]

Where itch is a dominant symptom consider the following:

- Pinworms, especially if perianal excoriation is present, and treat the child and her family with pyrantel 10 mg kg^{-1} (maximum 500 mg) orally or mebendazole 50 mg (6 months to <10 kg), 100 mg (>10 kg) orally (contraindicated in pregnancy) as a stat. dose, and repeat 2 weeks later.[8]
- Lichen sclerosis in the asymptomatic patient does not require treatment other than reassurance. In symptomatic patients management should consist of avoidance of irritants, improved hygiene, reduced trauma and barrier ointments, e.g. nappy rash creams. In more severe cases, a brief course of topical steroid (1% hydrocortisone) and referral to a dermatologist or paediatric gynaecologist are required.
- Eczema, especially if skin disease is present elsewhere. Combined treatment of the vulvovaginitis (as above) and hydrocortisone or betamethasone may be indicated.

Foreign body

If foreign body is suspected then an examination under anaesthesia with vaginoscopy is usually required. If the presence of a foreign body is questionable then an ultrasound may be beneficial to visualise the object. The most common

foreign bodies are tiny pieces of tissue paper. The possibility of sexual abuse should be borne in mind.

Vaginal bleeding

Introduction

Vaginal bleeding in the prepubertal child is abnormal after 3 to 4 weeks of age.[6]

Causes of vaginal bleeding in the prepubertal girl can be classed as hormonal and non-hormonal.

Hormonal causes

Neonates

A withdrawal bleed from maternal oestrogens is common and does not require investigation or treatment. This will cease after approximately 4 weeks of age.

Onset of menstruation

Consider as premature (precocious puberty) if this occurs at less than 8 years of age.[8]

Non-hormonal causes

Vulvovaginitis

In prepubescent girls, vaginal bleeding in this age group is most commonly due to more severe vulvovaginitis (the most common pathogens are *Shigella* or group A β-haemolytic streptococci).[6] The discharge is brown and often offensive (see above under vaginal discharge for management).

Trauma

Accidental injury usually results from straddle injuries where the girl falls on a narrow object (e.g. bicycle cross bar, jungle gym). Non-accidental injury/sexual abuse should be considered in trauma, especially where there is a hymenal tear without a history of accidental penetrating injury.

Foreign body

Vaginal foreign body may also result in bloodstained discharge and should be

considered if no infectious cause is identified or there is failure to respond to appropriate treatment. In bleeding secondary to excoriation from pinworms, lichen sclerosis or eczema, examination should demonstrate eczema and lichen sclerosis. The presence of anal and perineal excoriation suggests pinworms and treatment as described above.

Tumours

Although rare, the possibility of a genital tumour (endodermal sinus tumours and rhabdomyosarcomas) should be considered when there is chronic genital ulceration, non-traumatic swelling, tissue protruding from the vagina or foul-smelling, bloody, vaginal discharge. Benign papillomas can mimic these tumours. Vaginal bleeding associated with genital enlargement, premature sexual maturation or virilisation may be associated with a hormonally active ovarian or adrenal tumour.

Non-vaginal causes of bleeding

Haematuria may stain the underwear or nappy and be reported as vaginal bleeding. This will become evident upon urinalysis. Urethral prolapse often presents with bleeding thought to be vaginal in origin. The peak age is 5 to 8 years. Bleeding disorders are an uncommon cause of vaginal bleeding but should be considered when there are other systemic signs of a bleeding tendency, e.g. bruising and petechiae.

History

A general medical history including family and past history should be taken.

The history of vaginal bleeding should focus on the amount and circumstances of bleeding, the times of recurrences and the presence of associated pain. A history regarding any perineal trauma should be elicited as well as the possibility of a foreign body. Other medical disorders and symptoms of blood dyscrasias, such as epistaxis and bruising, should also be noted.

Examination

Vital signs and haemodynamic stability should be established. General examination, including sexual development (Tanner staging), should be performed. On examination, excessive bruising or petechiae and generalised skin disorders, such as eczema, should be noted.

Specific examination requires abdominal and vulval/perineal examination. Abdominal examination should note abdominal tenderness and the presence of a mass or organomegaly. Perineal examination should note the presence of vulvovaginitis, eczema or traumatic injury.

The common perineal injuries in trauma are bruising/haematoma of vulva and periclitoral folds and superficial lacerations of the labia minora and periurethral tissue. If a hymenal tear is present, a history of penetrating injury, e.g. broom handle or bed post, should be sought. If there is no history of penetrating trauma then sexual abuse needs to be considered.

A foreign body in the vagina may be evident on examination of the perineum.

A urethral prolapse will be evident on examination of the perineum as a friable, red–blue annular mass. The distal end of the urethra can prolapse partially anterior or posterior or in a complete circumferential (donut-like) fashion.

Investigations

Full blood examination should be performed if the child appears clinically anaemic or bleeding has been prolonged. Coagulation studies may be indicated. Perineal swabs should be taken for microscopy and culture where there is evidence of severe vulvovaginitis.

In trauma, where there is a history of penetrating injury or where it is not possible to perform adequate perineal examination, an examination under anaesthesia is required.

Midstream urine for micro and culture should also be performed where haematuria is present.

In the child less than 8 years of age who has signs of puberty evident, referral to a paediatric endocrinologist or gynaecologist is required.

Management

Trauma

A warm bath several times a day to help voiding as well as assist in keeping the perineum clean is all that is required. The girl may find voiding in a bath more comfortable. Antibiotics are rarely required. Examination under anaesthesia is only required if there is persistent bleeding, uncertain origin of bleeding, likely requirement for sutures, massive bruising or history of a fall on a potentially penetrating object. Suspected sexual abuse must be referred.

Urethral prolapse

The management is conservative as long as voiding is normal. Warm baths may be of assistance. Topical oestrogen cream assists in the resolution (Premarin® cream [conjugated estrogen] or Estrance® cream [estradiol, USP, 0.01%]).

Labial adhesions

Labial adhesions are seen in infancy, and usually resolve by about 8 years, although they may occasionally persist through to puberty when they will resolve around the time of menarche. Peak incidence is 3% of girls in their second year of life. The adhesion is not congenital, but it is acquired from a secondary adherence of the atrophic surfaces of the labia minora, presumably as a result of irritation.[1,2,9]

Labial adhesions are usually asymptomatic in children and do not need to be divided as long as the child is able to void. When symptoms occur they usually relate to difficulty with urination, recurrent urinary tract infections or irritation secondary to pooling of urine behind fused labia.[1,2,10] Parents should be reassured that separation of the labia will occur when oestrogenisation commences as the child grows. If symptomatic then labial adhesions may be treated with topical oestrogen or oestradiol or betamethasone (0.05%). Very rarely manual or surgical separation may be required for patients with severe urinary flow obstruction or recurrent urinary infections.

Associated nappy rash or vulvovaginitis is managed as described above.

Distressing vaginal or perineal pain

Severe perineal or vaginal pain, often described as a vaginal shooting pain that is very distressing, is reported in prepubertal girls. It predominantly occurs in the evening – waking from sleep. The likely cause for this is pinworms (*Enterobius vermicularis*). The worms, usually found in the perianal region, can become ‘lost’ within the vagina. When they crawl onto the thin atrophic hymen in the prepubescent girl they cause the distressing pain. Treatment with mebendazole, with repeated treatments (usually ×3 at weekly intervals), is required to ensure clearance.

Vaginal pain may also occur in conjunction with urethritis. External examination of genitalia may reveal herpetic lesions, discharge or inflammation. Herpes simplex (HSV) lesions may be a result of autoinoculation if herpetic

whitlow is present or oral HSV. Sexual contact must also be considered as the cause of HSV lesions, and this would raise the concern for sexual abuse.

Adolescent gynaecology

Vaginal discharge and sexually transmitted infections

Introduction

Vaginal discharge in an adolescent female may be physiological, a symptom of vaginal or cervical infection, or secondary to a vaginal foreign body. Under the influence of oestrogen there is an increase in the glycogen production by the vaginal epithelial cells, supporting the growth of lactobacilli in the vagina and lowering the vaginal pH to 3.8 to 4.5. The acidic environment helps to inhibit the growth of bacteria seen in the prepubertal female. Oestrogen also influences the cervix, resulting in increased mucus production, which is largely responsible for the physiological vaginal discharge.[11]

Sexually transmitted infections (STI) among adolescents are increasing and are frequently asymptomatic. Chlamydia and gonoccocal testing should be considered in all sexually active patients <25 years of age. Left untreated STIs lead to a range of complications including vaginal discharge, urethritis, infertility, pelvic inflammatory disease (PID) and pelvic pain.

Vaginal discharge may represent infections such as *Candida* or bacterial vaginosis. STIs leading to vaginal discharge include: trichomoniasis, chlamydia, gonococcus or syphilis disease, and appropriate swabs and cultures should be taken.

Non-specific urethritis in males is a diagnosis of exclusion. When the male urethra is inflamed, there may be penile discharge, and a diagnosis of chlamydia or gonorrhoea needs to be excluded.

History

Diagnosis can often be suspected from the history and the appearance of the discharge:

- *Candida* infections typically present as a cheesy white discharge and are associated with vulval and vaginal itching, dysuria or superficial dyspareunia in sexually active girls. They are commonly associated

with antibiotic use, stress and diabetes mellitus. Approximately 10–20% of women in reproductive years may be colonised with *Candida* and only require treatment if symptomatic.

- Bacterial vaginosis (BV) generally presents as a homogenous white discharge associated with a fishy odour. Symptoms of vaginal irritation are uncommon. The exact mechanism for onset of BV is unclear; however, it is associated with reduction in lactobacilli and hydrogen peroxide production and a rise in vaginal pH leading to an overgrowth of BV-associated organisms. It is not considered to be an STI but can be brought on by anything that changes the balance in the vagina.
- Trichomoniasis is an STI and typically presents with a frothy, green vaginal discharge. Fifty per cent of men and women infected with trichomonas will be asymptomatic. If symptomatic discharge may be yellow–green and frothy, and vaginal irritation may occur.
- Chlamydia and gonorrhoea infections may present as vaginal discharge, intermenstrual or post-coital bleeding, urethritis or PID. Chlamydia and gonorrhoea may also be identified incidentally due to the high incidence of asymptomatic infections. The presence of lower abdominal pain and/or dyspareunia in the history increases the likelihood of PID, with a positive and negative predictive value of 17% and 100%, respectively.[12] STIs may be contracted by oral or digital contact and do not necessarily require penetrative intercourse for transmission. Chlamydia testing should be considered in all patients found to have a sterile pyuria as well as women presenting with breakthrough bleeding while taking the oral contraception pill. It is important to remember that chlamydia is asymptomatic in approximately 70% of all people.
- Syphilis (*Treponema pallidum*) has been increasing since 2002. Although the number of overall cases still remains low in comparison to other STIs, it is important to test for and screen for. Syphilis can enter the body by sexual contact through the skin lining in the genital tract, anus or mouth.
- Genital warts are the most common viral infection diagnosed in sexual health clinics. In the last 5 years there has been a 9.5% increase in people presenting with genital warts. Human papilloma virus (HPV) subtypes 6 and 11 are the cause of 90% of genital warts. This is important as subtypes of HPV (16 and 18) are linked with genital cancer (cervical, penile and anal). HPV vaccination amongst adolescents helps

prevent HPV infection and is now publically funded in most developed countries.

- Genital herpes is caused by HSV-1 and HSV-2. Traditionally HSV-1 was thought to be primarily the cause of orolabial herpes and HSV-2 of genital herpes; however, considerable crossover is well documented.
- The most common vaginal foreign body in adolescent girls is a retained tampon. The presenting symptom is often a malodourous discharge.

Examination

Inspection of the external genitalia should be performed, specifically looking for evidence of warts, herpetic lesions, the nature of the discharge and evidence of excoriation.

Adolescent girls who have never been sexually active and/or who have not used tampons should not be examined internally. If there is a suspicion of a vaginal foreign body, vaginal trauma or PID in a girl who is sexually active or uses tampons, a gentle vaginal examination may be considered. However, even sexually experienced girls may be unable to proceed with the examination, and alternative forms of assessment may need to be considered.

Investigations

Microscopy, culture and wet-preparation analysis of a vaginal discharge will help to confirm the diagnosis of candidiasis, bacterial vaginosis and trichomoniasis.

The gold standard for diagnosis of both chlamydia and gonorrhoea is culture from a cervical sample. However, both infections can readily be detected with a high degree of sensitivity using a molecular replication technique (either polymerase chain reaction [PCR] or ligase chain reaction [LCR]) performed on samples collected from either the cervix or a first-catch urine. Women can perform self-taken lower-vaginal swabs, and men can collect a first-void urine for chlamydia testing. First-void urines in men and self-taken vaginal swabs in women are ideal tests that can be easily done for screening for chlamydia which is often asymptomatic.

Management

- *Candida albicans* is the most common cause of vaginal thrush. One

treatment option is clotrimazole 10% cream inserted vaginally as a single dose at night. An alternative, if the patient is intolerant of topical therapy and not pregnant, is fluconazole 150 mg orally as a single dose.[13]

- Bacterial vaginosis should be treated in symptomatic patients and in those at high risk of acquiring other STIs. Oral metronidazole 400 mg bd for 5 days is recommended in non-pregnant patients.[13]
- Trichomoniasis can be treated with metronidazole 2 g orally as a single dose.[13]
- Chlamydia treatment is with either a 7-day course of doxycycline (100 mg bd) or a single oral dose of 1 g azithromycin.[13]
- Gonorrhoea has developed resistance to a number of antibiotics, and therefore treatment should be based on local sensitivities. In urban Australia a single dose of 250 mg ceftriaxone intramuscularly in combination with cover for chlamydia is currently recommended.[13]
- A retained tampon can generally be removed in the ED. However, occasionally the swollen tampon cannot be extracted due to a hymenal ring, and removal under general anaesthesia is required. If the tampon is removed in the ED it should be immediately placed into a container of water to limit exposure to the often offensive odour.

Disposition

Adolescents with an STI should have a follow-up communicable disease consultation to discuss prevention of further STIs. Ongoing contraception and contact tracing should also be addressed. Partner notification is an essential part of the management of STIs. Current recommendations include notification of all sexual contacts within the previous 60 days. If there have not been any sexual partners in the last 60 days then the most recent sexual contact should be notified up to 6 months.

Gonorrhoea and chlamydia are notifiable diseases in some jurisdictions.

Abnormal vaginal bleeding

Introduction

Abnormal vaginal bleeding may be caused by a complication of pregnancy, polyp, adenomyosis, malignancy, ovarian dysfunction, trauma, ruptured ovarian

cyst, ovarian torsion or infection, or it may be secondary to contraceptive use. The most common causes of heavy vaginal bleeding are anovulatory bleeding or an underlying haematological condition.[14]

Menstrual cycles in adolescents are often anovulatory due to the gradual maturation of the hypothalamic–pituitary–ovarian axis, which can take up to 5 years after the menarche.[14–17] Anovulatory or dysfunctional uterine bleeding generally presents as irregular, often heavy, blood loss. Anovulatory bleeding may result from a relative deficiency of either oestrogen or progesterone. Relative oestrogen deficiency is more common in thin young women who have just commenced their menstrual cycles. The relative oestrogen deficiency results in a thin, atrophic endometrium, which can bleed profusely.

In girls with higher endogenous oestrogen levels, anovulatory menstrual cycles are more likely to result in a relative progesterone deficiency due to failure of the luteal phase. In this case the unopposed oestrogen results in thickening of the endometrial lining, which can bleed erratically.

Menorrhagia, whether associated with ovulatory or anovulatory cycles, may be a marker of systemic illness. Up to 20% of patients admitted with menorrhagia have been found to have an underlying haematological disease, the most common of which is a coagulopathy, half of which are von Willebrand disease and the other half due to platelet problems or dysfunction – other factor deficiencies are very rare.[14,17]

Points to review in the history of menorrhagia include asking about volume and duration of bleeding and family history of bleeding disorders (e.g. epistaxis in fathers). A useful indicator of the amount of bleeding includes information about the number of pads/tampons changed in the last 12/24 hours. Clots with a diameter greater than 1 cm are useful to ascertain if heavier bleeding is occurring. Chronic untreated menorrhagia may also present with signs and symptoms of anaemia.

Examination

The haemodynamic status of the patient and the severity of the bleeding should be assessed. A general examination should be performed, particularly noting evidence of a haematological disorder or anaemia.

Investigations

A full blood count, iron studies and coagulation screen will assist in identifying

anaemia, an underlying coagulopathy or haematological cause for the bleeding (although caution needs to be taken in interpreting these results as stress may result in apparent ‘normalising’ of the test results, particularly in von Willebrand disease[14]). A pregnancy test should also be performed.

Ultrasound may be helpful when ovarian pathology is suspected and can sometimes be used to assess the thickness of the endometrium, although transabdominal views may be limited. An STI screen should be obtained if clinically indicated, as chlamydia in particular can cause intermenstrual or heavy bleeding.

Dilation and curettage is very rarely indicated for a diagnosis and is not a recognised treatment for dysfunctional uterine bleeding.[18]

Management

Heavy bleeding, either acute or chronic, needs appropriate assessment for hypovolemia and should be treated with fluid and blood products as clinically indicated.

Oestrogens and progestins are generally used in the management of anovulatory bleeding. Suggested regimens vary with respect to the route of administration, the dose and the type of hormone used. Currently there is no convincing evidence in favour of any particular regimen.[19]

For both the acute and non-acute bleeding tranexamic acid (1.3 g orally or 10 mg kg^{-1} IV to a maximum of 600 mg $dose^{-1}$) should be used. This can be used in combination with hormonal approaches. If using progestogens first line, regimens include oral norethisterone 5–10 mg every 2 hours for four doses, followed by 5 mg two to three times a day for 14 days.[11] Commencing the oral contraceptive pill once the bleeding has ceased may be sensible, as cessation of the progestogens will almost invariably result in bleeding recommencing.

Alternatively, treatment may include oral conjugated oestrogens 0.625–1.25 mg every 4–6 hours or oral oestradiol 1–2 mg every 4–6 hours until the bleeding stops, which is usually about 24 hours. The dose is then reduced to once or twice daily. Intravenous conjugated oestrogens are no longer available in Australia and have been associated with thromboembolic complications, consequently oral regimens are generally preferred.[20] This approach is worth taking when progestogens have already been used and failed but can be used first line. Again, the oral contraceptive pill should be commenced for ongoing control, particularly if there is a need to allow some time before further menses in the context of a low haemoglobin. Supplement with iron therapy.[21]

Menorrhagia, in the absence of a coagulation disorder or other underlying pathology, can be treated with non-steroidal anti-inflammatory agents and tranexamic acid, which reduce the menstrual loss by 33% and 50%, respectively,[14,22] or, alternatively, low-dose oral contraceptive therapy.[15]

Disposition

The identification of a coagulation disorder will necessitate review by a haematologist. This can usually be done as an outpatient with a primary care physician's input until haematology review. Pregnancy-related bleeding should be reviewed by an obstetrician/gynaecologist. Anovulatory bleeding and simple menorrhagia can be managed by either a primary care physician or a gynaecologist.

Pelvic pain

Introduction

The differential diagnosis of acute pelvic pain in an adolescent includes both gynaecological and non-gynaecological causes. Common gynaecological causes include pregnancy-related pain, dysmenorrhoea, ovulation pain (Mittelschmertz), ovarian cysts and their complications, torsion of the ovary, PID, endometriosis and congenital uterine abnormalities, such as imperforate hymen.

History

A general gynaecological history should be taken, with particular note made of the relationship of the pain to the menstrual cycle. History should also include the nature of onset of the pain and a history of similar episodes. The pain of dysmenorrhoea, Mittelschmertz, endometriosis and imperforate hymen may be cyclical. However, they may also present during the first episode or as an unusually severe exacerbation. An ectopic pregnancy is not excluded by the history of a recent menstrual period:

- Primary dysmenorrhoea is common in the adolescent population; it refers to pain associated with menstruation in the absence of underlying pathology. The pain is generally described as crampy, occurring in the lower abdomen, often with radiation to the back or to the inner thighs.

Associated features may include nausea, vomiting, diarrhoea, headache and syncope. The symptoms have been attributed, in part, to an increased release of, or increased sensitivity to, prostaglandins. The diagnosis is generally made on the basis of history and limited physical examination findings.

- Mittelschmertz is pelvic pain related to ovulation. The pain is generally dull in nature and located in either of the iliac fossae. The pain may last from a few hours to 2–3 days. The pathological basis of the pain is unclear but may be due either to distension of the capsule of the ovary prior to ovulation or to spillage of a small amount of follicular fluid at the time of ovulation.[11,23]
- Simple ovarian cysts are a normal physiological consequence of ovulation. They can be up to 6 cm in diameter. The pain may be from irritation of surrounding structures or stretching of the wall of the cyst. Corpus luteal cysts can reach up to 10 cm in size and may present as acute pain secondary to haemorrhage into the cyst. Both simple and corpus-luteal cysts can present as acute pain if they rupture. Although uncommon, a ruptured corpus-luteal cyst may continue to bleed and be associated with significant haemoperitoneum.[11] Bleeding disorders need to be considered if recurrent haemorrhagic cysts occur or a significant haemoperitoneum.
- Complex ovarian cysts include dermoid cysts, which are often asymptomatic but may cause local pressure symptoms.
- Ovarian torsion is an uncommon condition and is almost exclusively seen in association with ovarian pathology. The history is typically an acute onset of colicky iliac fossa pain, with the right being affected more than the left. There may be associated nausea, vomiting and a leucocytosis. Ultrasound has a high degree of sensitivity and specificity in making the diagnosis of an enlarged ovary; however, Doppler studies are not necessarily conclusive, and presence of flow does not exclude torsion.[24,25]
- PID can have a range of clinical presentations, depending on the location and the severity of the infection. The pain can range from mild to severe; systemic features of infection are not always present. Due to the significant complications associated with untreated PID, the isolated findings of adnexal, uterine or cervical motion tenderness in the young woman who is sexually active, for which no other cause can be found,

have been suggested as a basis for the diagnosis and treatment of PID.[26]

- Endometriosis is reputed to cause a range of menstrual disorders and pain syndromes. However, the symptoms do not appear to correlate well with the degree of disease identified at laparoscopy. The diagnosis should be made with caution due to the risk of labelling a patient with what may be seen as a sentence for chronic pain and infertility.
- Congenital obstructive abnormalities include an imperforate hymen or obstruction of one side of a double uterine, cervical or vaginal system. An imperforate hymen may present as cyclical or irregular lower abdominal pain in a girl with well-developed secondary sexual characteristics and the absence of menstruation. Unilateral obstruction in a double system more typically presents with dysmenorrhoea that commences several days after the onset of menses and persists beyond the end of the menstrual period.

Examination

Examination should include vital signs and abdominal examination. Pelvic examination can give additional information regarding the location of the pain and its relationship to pelvic organs. Uterine, adnexal or cervical motion tenderness should be sought if pelvic examination is performed. The identification of an adnexal mass is clinically imprecise. However, it should prompt consideration of such conditions as ovarian torsion, tubo-ovarian abscess or ectopic pregnancy. A pelvic examination may not be possible in the young adolescent, although ultrasound will be helpful.

Investigations

A pregnancy test and a pelvic ultrasound are the most valuable tests for excluding pregnancy-related conditions. Ultrasound is the investigation of choice for the definition of masses and in the identification of ovarian cysts.[27] If a congenital anomaly is suspected, information regarding the kidneys at the time of the ultrasound can be helpful.

Laparoscopy is reserved for pain that cannot be adequately explained or has failed to respond to appropriate treatments. Laparoscopy may be required to exclude torsion where a cyst is present combined with a suspicious history.[25] It may also have a role in the investigation of chronic pelvic pain.

Management

- Dysmenorrhoea will generally respond to antiprostaglandin agents, such as mefenamic acid 500 mg orally every 8 hours or ibuprofen 400 mg orally every 8 hours. The response is improved by commencing treatment prior to the onset of symptoms.[28] Lifestyle issues, such as increasing physical exercise and decreasing stress, may also improve symptoms.[29] For patients who fail to respond to treatment, combined oral contraceptive therapy is considered second-line therapy, and further investigation, such as ultrasound and laparoscopy, may be considered to identify other pathology, for example significant endometriosis.[28]
- Mittelschmertz and simple ovarian cysts can often be treated by reassurance. However, if this is not adequate, simple analgesia and non-steroidal anti-inflammatory drugs may be required. If the pain is recurrent then consideration should be given to the use of the combined oral contraceptive pill in an attempt to suppress ovulation. Failure of resolution of a cyst after 2 months or a simple cyst greater than 8 cm diameter may require surgical exploration.[11]
- Complex ovarian cysts, including dermoids and endometriomas, are generally treated surgically.
- Ovarian torsion requires urgent operative intervention with the aim to preserve ovarian tissue.[25]
- PID can be treated with either oral or parenteral antibiotics. However, there should be a low threshold for admission in adolescents.[30]

Disposition

Most forms of pelvic pain will require further management or investigation and should be referred accordingly.

Chronic pelvic pain will need regular input of a primary care physician once all necessary investigations have been completed. It is important that a complete social history is taken looking for underlying social stressors (including sexual abuse in the past or present and previous psychological trauma) that may be adding to a complex pain syndrome. A holistic approach is needed for the effective management of chronic pelvic pain.

Controversies

1. The management of anovulatory vaginal bleeding.
2. The diagnosis and management of ovarian torsion.
3. The diagnostic criteria and optimal management of pelvic inflammatory disease.

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15.2

Emergency contraception

Alastair D.McR. Meyer, and Jacqueline E.L. Parkinson

ESSENTIALS

1 Emergency contraception (EC) should be made available to a female adolescent who has had unprotected sexual intercourse within 72 hours prior to presentation, regardless of the stage of her menstrual cycle.

2 Pregnancy must be excluded by urinary β-hCG testing before prescribing EC.

3 Commonly prescribed treatment is levonorgestrel (LNE) 1.5 mg tablet stat.

4 Follow-up is essential.

Introduction

Emergency contraception (EC) or post-coital contraception can be defined as preventing pregnancy after unprotected sexual intercourse (UPSI). Every year over 3.5 million unintended pregnancies occur in the United States alone, mostly involving teenagers. It is believed that half of these unintended pregnancies could be avoided by the judicious use of EC. Indications for the provision of EC are shown in Box 15.2.1.[1]

Many of these unintended pregnancies are later surgically terminated. Estimates of as many as 170,000 such terminations are performed in England and Wales annually.[2] These surgical terminations are not without clinical risk and cost, as well as being a major social, religious and political issue.

Clinical assessment

EC should be made available to an adolescent who has had unprotected sexual intercourse within 72 hours prior to presentation, regardless of the stage of her menstrual cycle. Pregnancy must be excluded by urinary β-human chorionic gonadotrophin hormone (β-hCG) testing.

Depending on the age of the young person, issues of child protection and consent to treatment may need to be explored and vary by jurisdiction.

Available medicines

Historically, the Yuzpe method of EC was commonly practiced in Australasia. This involved a high-dose oestrogen/progestogen preparation. Two doses of 100 mcg of ethinylestradiol combined with 500 mcg of levonorgestrel given 12 hours apart resulted in withdrawal bleeding within 21 days for 98% of women.[3] Due to relatively higher adverse effects and lower efficacy, this regimen is no longer recommended unless it is the only option available.

Box 15.2.1 Indication for providing emergency contraception

Unprotected sexual intercourse
Barrier contraception failure
Intrauterine contraceptive device expulsion
Missed contraceptive pills
Sexual assault

Levonorgestrel (LNE) has been licensed specifically as an EC agent since the early 2000s. While the precise mechanism by which LNE prevents pregnancy is not clear, it is believed to work by preventing ovulation and altering the tubal transport of sperm and ova thus preventing fertilisation. Initially this was prescribed as two doses of 0.75 mg 12 hours apart. However, the recommended regimen for LNE EC is now 1.5 mg orally stat. This simplified single-dose EC should be taken within 72 hours of UPSI, but ideally as soon as possible, as efficacy falls dramatically over time (Table 15.2.1). LNE EC can be used 96–120 hours after UPSI, however, efficacy is unknown and unlicensed.[4] There is a

possibility that efficacy is inversely related to weight, with some reports finding EC failure is four-fold higher in obese females.[5] Although dose alteration is not recommended, detailed counselling should be provided reinforcing the importance of medical follow-up to assess contraceptive outcome.[6]

Ulipristal acetate (UPA) is a progesterone receptor modulator licensed for use within 120 hours of UPSI. It is taken as a single 30 mg dose as soon as possible after UPSI.

Table 15.2.1

Efficacy of levonorgestrel emergency contraception[7,8]

Hours post-coital	Pregnancies prevented (%)
<24	95
24–48	85
49–72	58

Medicine interactions

LNE and UPA are primarily metabolised in the liver, and their efficacy can be reduced by enzyme inducers such as anti-epileptic medicines or HIV post-exposure prophylaxis.[4] A copper intrauterine device is the most preferable option of EC if the woman is taking, or has taken within the last 28 days, liver enzyme-inducing medicines. Alternatively there is some evidence for prescribing 3 mg of LNE (2 x 1.5 mg tablets), however this is unlicensed.[9]

A full medication history should be taken prior to prescribing EC, and medicine interactions should be checked using an up-to-date interaction reference.

Medicine contraindications

LNE and UPA EC are contraindicated in unexplained vaginal bleeding, current breast cancer and pregnancy. Care should be taken with patients taking anticoagulants. Adverse reactions include fatigue, abdominal pain, gastrointestinal discomfort, dizziness, headache, breast tenderness and vaginal bleeding.

Conditions regarded as relative contraindications include: severe

hypertension; diabetes with nephropathy; retinopathy; neuropathy; ischaemic heart disease; and past history of breast cancer.

Medicine adverse effects

LNE contraception has fewer adverse effects than the traditional Yuzpe method. Only 20% of patients experience nausea, which usually responds to standard oral antiemetics. If vomiting does occur within 3 hours of UPA or within 2 hours of LNE then the dose should be repeated.

Medicine outcomes

LNE and UPA EC can result in early or late menstruation, although the majority of women will experience bleeding within 3 days of their expected date. It is essential that women have pregnancy testing if their period is more than 7 days late or very light.[6]

LNE **and UPA** EC can be used multiple times in the same menstrual cycle.[10]

There is a rapid return to fertility after using LNE and UPA EC, so women should be advised to use barrier contraception until they have had their next menstrual bleed.

Copper intrauterine devices

If unprotected sexual intercourse has occurred more than 72 hours but less than 5 days before presentation, insertion of a copper-based intrauterine contraceptive device (Cu-IUD) should occur.

These devices prevent implantation. Cu-IUDs should be inserted in consultation with the gynaecology service.

If inserted up to 5 days after predicted ovulation, Cu-IUDs can prevent 99% of expected pregnancies.[11] Relative contraindications are nulliparity and patients who are at high risk of sexually transmitted infection (STI). If testing for STIs before Cu-IUD insertion is not possible, presumptive antibiotic cover should be considered.[10]

All women presenting to emergency departments following unprotected sexual intercourse should be counselled regarding STIs, ongoing contraception and the need for follow-up. It is an essential part of care that clear follow-up arrangements are made to determine if EC has been effective.

Controversies

1. Is the emergency department (ED) an appropriate site for dispensing emergency contraception (EC), as these patients are essentially well?
2. There may be a place for non-medical providers of EC, given its safety.
3. The place of follow-up termination, should EC fail, is important to consider.

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SECTION 16

Renal

OUTLINE

16.1

Acute kidney injury

Sameer A. Pathan

ESSENTIALS

1 Acute kidney injury (AKI) equates to an acute reduction in kidney function.

2 A high or rising plasma creatinine is a widely accepted surrogate for impaired GFR, but this does not necessarily distinguish between AKI and chronic kidney disease.

3 A normal plasma creatinine does not exclude AKI.

4 The commonest causes of AKI in a paediatric population presenting to the emergency department are sepsis, haemolytic uraemic syndrome and post-streptococcal glomerulonephritis.

5 Not all AKI presentations are oliguric or anuric.

6 Most cases of oliguria are not AKI.

7 Most cases of elevated plasma urea are not AKI.

8 The most important treatment objectives are correcting circulating volume, potassium and blood pressure.

9 Hyperkalaemia in acute kidney injury is not necessarily caused by potassium retention, especially when of short duration, but by potassium redistribution from cells secondary to acidosis.

10 Posterior urethral valves should be suspected in male infants with unexplained acute kidney injury.

Introduction

Acute kidney injury (AKI), in paediatric patients, presents as a wide range of clinical manifestations. AKI is defined as an abrupt decline in kidney function, resulting in retention of nitrogen waste products and disturbed extracellular fluid and electrolyte homeostasis. The precise incidence and prevalence of AKI is unknown; however, they are generally reported as 3–10% based on the geographic location and the definition used. The incidence is higher, 25–40%, in children who are critically ill, preterm or undergoing surgical procedure, or receiving bone-marrow transplant. Commonly used definitions for paediatric AKI are paediatric Risk Injury Failure, Loss of kidney Function, and End stage renal disease (pRIFLE), Acute Kidney Injury Network (AKIN) and Kidney Disease Improving Global Outcome (KDIGO). The staging of AKI is based on change in the plasma creatinine level or urine output. KDIGO uses a modification of pRIFLE and AKIN components and defines AKI as:

- rise in the plasma creatinine by 0.3 mg dL (>26.5 μmol L) within 48 hours OR
- rise in the plasma creatinine by 50% within 7 days OR
- urine output of <0.5 mL kg h for 6 hours.

Classically, AKI has been divided into three broad categories, but pathophysiological processes overlap, and the first and second groups can lead to the third when severe or prolonged. The detailed causes are mentioned in Table 16.1.1:

1. **Pre-renal disease.** In developing countries, this is the most common type of AKI in the paediatric population caused by renal hypo-perfusion. Hypovolaemia, decreased cardiac function, reduced renal blood flow, or peripheral vasodilation causing decrease preload may cause renal hypo-perfusion. In pre-renal AKI, reduction in GFR leads to elevation of plasma urea and creatinine – to improve intravascular volume, the loop of Henle increases the reabsorption of sodium and water. Thus, oliguria is observed with higher specific gravity of urine and lower urinary sodium (<20 mEq L). It usually responds to improving renal perfusion, and the GFR returns to normal.
2. **Post-renal diseases.** Anatomical obstruction in the renal tract anywhere from the pelvi-calyceal system to the urethra may lead to AKI or chronic renal failure (CRF). Crystalopathy (obstructing the tubules) is usually

regarded as a ‘renal intrinsic’ cause.

Table 16.1.1

Causes of acute kidney injury

Pre-renal causes of AKI	
Hypovolaemia reduction in circulating volume	Dehydration, gastroenteritis, haemorrhage, burns, diuretic use, salt-wasting tubulopathies, nephrotic syndrome, SIADH
Cardiac dysfunction	Cardiogenic shock, heart failure, dysrhythmias
Renal or severe systemic vasoconstriction	Sepsis, ACE inhibitors, NSAIDs, renal artery disease
Mixed mechanism leading to hypo-perfusion	Sepsis, anaphylaxis, DIC
Intrinsic renal causes of AKI	
Renal vascular disease	HUS, vasculitis, thrombosis, TTP
Glomerulonephritides	APGN, HSP, and progressive or post-infection GN
Acute tubular injury	ATN caused by prolonged hypoxia, toxins, hypo-perfusion, infection, or immune-mediated (IgG or IgA nephropathies, SLE), or contrast-induced nephropathy
Interstitial disease	Drugs, infection, immunological response to renal parenchyma (SLE, WG, anti-GBM, complement disorders, and transplant rejection), malignant infiltrations (lymphoma, myeloma), granulomatous diseases (TB, sarcoidosis), or idiopathic interstitial nephritis
Post-renal causes of AKI	Stone, clots, sludge, pus, posterior urethral valve, strictures, neurogenic bladder

ACE, angiotensin-converting enzyme; AKI, acute kidney injury; APGN, acute proliferative glomerulonephritis; ATN, acute tubular necrosis; DIC, disseminated intravascular coagulation; GN, glomerulonephritis; HUS, haemolytic uraemic syndrome; NSAIDs, non-steroidal anti-inflammatory drugs; SIADH, syndrome of inappropriate antidiuretic hormone secretion; SLE, systemic lupus erythematosus; TB, tuberculosis; WG, Wegener’s granulomatosis.

Table 16.1.2

Glomerular filtration rate and plasma creatinine

	Children	Adult
GFR	1 mL kg min	2 mL kg min
Normal plasma creatinine (P_{Cr})	27–62 µmol L	60–110 µmol L
Formula to calculate GFR (mL kg min)	70 P_{Cr}	140 P_{Cr}
Plasma creatinine rise in complete loss of GFR	150 µmol L day	300 µmol L day

3. **Intrinsic renal disease.** It represents damage to renal parenchyma secondary to hypoxia, ischaemia, toxins, infection or inflammatory insult. The damage could be further sub-divided according to renal vessels, glomerular, tubular, or interstitial damage. The urine may have low specific gravity (secondary to loss of concentrating ability), low urine/plasma creatinine ratio <20 (loss of filtration function), and high urinary sodium (>40 mEq L) due to loss of reabsorption function.

In developed countries, the most common causes of AKI in hospitalised children are sepsis, nephrotoxins, congenital heart disease and renal ischaemia (Table 16.1.1).

Pathophysiology

The kidney filtration process has two important functions:

1. Excretion of waste products of metabolism (e.g. creatinine, urea, and non-volatile acid) and excess ingested elements (e.g. potassium).
2. Re-absorption of essential constituents such as glucose, amino acids, and most of the water, NaCl, and HCO_3.

Based on the muscle mass, creatinine is produced in the body at a constant rate and continuously filtered through the glomeruli. There is a wide variation in GFR between and within individuals, reflected as wide variations in plasma creatinine (P_{Cr}) in normal children. The approximate values for GFR and the formula are given in Table 16.1.2. Although P_{Cr} is the most widely accepted surrogate marker for GFR, there is often a delayed and variable rise with acute worsening of renal function. Therefore, a normal plasma creatinine does not exclude AKI. The P_{Cr} and the GFR values vary according to age, sex, size of the individual and the assay used to measure P_{Cr}, thereby limiting the use of interpreting isolated creatinine results. Generally, any creatinine >100 μmol L^{-1} is abnormal, but in infants values as low as >50 μmol L^{-1} should raise a suspicion of impaired GFR: in a 5-year-old >70, in a 10-year-old >80 and in a 15-year-old >100 μmol L^{-1}. More important is the rate of increase of plasma creatinine in AKI, which takes 12–24 hours to become evident. The plasma creatinine loses its value to assess the kidney function once dialysis is initiated.

Unlike creatinine, some of the urea is reabsorbed from renal filtration. Normally 20% of the renal plasma flow is filtered, which may rise up to 50% in case of intravascular hypovolaemia. Thus, in pre-renal AKI, urea to creatinine ratio may rise to >20:1 with Oliguria. In conditions such as gastrointenstinal bleeding, tissue breakdown or low muscle mass in critically ill patients, there may be a falsely high urea/creatinine ratio. In a patient with liver disease or low protein intake, low urea production may give a false normal ratio of urea and creatinine. Therefore in the diagnosis of AKI the urea/creatinine ratio has limited diagnostic utility (see Table 16.1.2).

Clinical presentation

AKI in the paediatric population has a wide range of clinical manifestations,

with non-specific presentations to anuric renal failure. One should not rule out AKI simply based on normal creatinine at the time of presentation for the various reasons discussed above. However, a high index of suspicion and a detailed history and physical examination are important to detect AKI or risk of AKI in a paediatric population (Table 16.1.3). Early diagnosis and treatment are important as in-hospital mortality ranges from 1.5–9.5% in non-intensive care settings to as high as 30–50% in patients with severe AKI. The chance of developing chronic kidney disease (CKD) or end-stage renal disease (ESRD) is up to 40–60% in the survivors of paediatric AKI.

Suspicion of glomerulonephritis or vasculitis should prompt inpatient referral for further investigation into the cause.

Urine electrolytes and osmolality are helpful in differentiating causes of AKI (Table 16.1.4).

Treatment

1. Treat established causes and complications:
   Life-threatening conditions (e.g. hypoxia, hypovolaemia, hyperkalaemia, and seizures) should be assessed and treated promptly. Ventilation may be needed for pulmonary oedema not responsive to diuretics. Patients should be connected to the monitor for continuous haemodynamic monitoring.
2. Avoid further damage and remove the cause:
   Avoid excessive fluids and electrolyte supplements (especially Na, K, and PO_4). Attempt to establish urine output in oliguria of short duration, but if no response is seen, management with Furosemide should not persist. Mannitol is not recommended as it may worsen the situation by acute intravascular expansion, especially if there is no diuresis. Avoid nephrotoxic drugs.

Table 16.1.3

History	
History of fluid loss	Gastroenteritis, burns, surgery, shock
Urine output	Oliguria or anuria (pre-renal, SIADH, or obstructive cause), polyuria (AKI, CRF, diuretics, interstitial nephritis), haematuria (renal parenchymal disease or stone)
Nephrotoxic drug or toxin exposure	NSAIDs, aminoglycosides, contrast agents, amphotericin, sulphonamide, aciclovir, cyclosporin, tacrolimus, ACE inhibitors, β-lactam group, exposure to heavy metals (lead, mercury), alcohol ingestion
Post-infection history	Symptoms of respiratory infection (post-strep GN, HSP), history of bloody diarrhoea or abdominal pain (HUS)
Chronic illness	Diabetes, hypertension, heart failure and liver cirrhosis (symptoms of water retention), congenital heart disease, connective tissue disorder (rash, joint pain or swelling, mouth ulcers)
Physical exam	
Signs of decrease circulating volume	Tachycardia, hypotension, decrease capillary refill, dry mucosa, loss of skin tone, low urine output
Signs of fluid overload	Oedema (generalised or dependent), hypertension, pulmonary oedema
Signs of electrolyte imbalance	Change in neurological function or signs of acidosis
Other systemic signs	Fever, rash, joint swelling, mucosal ulceration
Investigations	
Blood	Full blood counts, cultures (sepsis)
	Plasma Na, K, Cl, Ca, PO_4, Mg (electrolyte imbalance)
	Plasma BUN and creatinine, liver function, coagulation profile (staging of AKI)
	Plasma pH, bicarbonates, lactate, osmolality (metabolic acidosis workup)
	Glucose, uric acid, albumin, myoglobin creatinine kinase
Urine	Urine microscopy, Gram stain and culture (UTI or cell cast)
	Urine chemistry including NA, K, urea, osmolality, creatinine (help differentiate pre-renal and intrinsic AKI)
	Urine protein (intrinsic renal cause) and urate
Imaging	Ultrasonography or CT scan to rule out obstructive cause
	Chest X-ray to look for signs of infection, congestion or cardiomegaly

ACE, angiotensin-converting enzyme; AKI, acute kidney injury; BUN, blood urea nitrogen; CRP, C-reactive protein; GN, glomerulonephritis; HSP, Henoch–Schönlein purpura; HUS, haemolytic uraemic syndrome; NSAIDs, non-steroidal anti-inflammatory drugs; SIADH, syndrome of inappropriate antidiuretic hormone secretion; UTI, urinary tract infection.

Table 16.1.4

Differentiating pre-renal and intrinsic acute kidney injury from syndrome of inappropriate antidiuretic hormone secretion

	Pre-renal AKI	Intrinsic AKI	SIADH
Glomerular function	Decrease GFR with preserved nephron	Damaged nephrons	Inappropriate ADH secretions
Plasma creatinine (umol L)	Mild–moderate increase and return to normal if GF is improved	Mild increase initially and then progressive worsening	Normal or low
Plasma urea	Moderate–marked increase	High	Normal–low
FE urea (per cent)	<35	>50	–
Tubular function			
Plasma Na (mEq L)	Normal, low (heart failure), high (both Na and H_2O retention)	Normal or low (ATN)	Low (both Na loss and excess H_2O intake)
Urine Na (mEq L)	<10–20 (max reabsorption by RT)	>20–40 (tubular dysfunction in Na reabsorption)	Often >100–200 (Na loss triggered by extracellular volume expansion)
FE Na (per cent)	<1 (for infants <2)	>2 (for infants >2.5)	1–2
Urine specific gravity (concentrated urine)	>1.020	<1.012	>1.020
Urine osmolality (mosmol kg)	>400 (high Na low water in urine)	Similar to plasma	>300, often >800 (very high Na in urine)
Effect on plasma osmolality	High	Normal–high	Low

ADH, antidiuretic hormone; AKI, acute kidney injury; ATN, acute tubular necrosis; GFR, glomerular filtration rate; RT, renal tubular; SIADH, syndrome of inappropriate antidiuretic hormone secretion.

When the GFR falls below 50% of normal, most drugs excreted by the kidney will require modifications in dose or scheduling. However, there is no evidence to support the reduction of the initial dose of antibiotics in the emergency department (ED) if an infection is suspected.

3. Fluid management:
    - Hypovolaemic: Rapid volume restoration should be performed with 0.9% saline (10–20 mL kg fluid bolus, can be repeated twice) in addition to preventing further fluid loss. Urine output

should be measured to calculate hourly urine output.

- Euvolaemic: Care should be given to make up for insensible losses which can be higher in febrile patients (300–500 mL m^2 per day).
- Hypervolaemic: In critically ill paediatric patients, fluid overload in the setting of AKI was found to be an independent risk factor for mortality (3% increase for each 1% increase in fluid overload). However, the role of diuretics in established renal failure is limited in terms of improving renal or patient survival. Therefore, the main use of diuretics in AKI is to treat fluid overload and not to improve or reverse the kidney function.

$$\text{Percent fluid overload} = [\text{total fluid in (Litres)} - \text{fluid out (Litres)}]/\text{admission weight (kg)} \times 100$$

A single high-dose bolus (2–5 mg kg) can be tried in children with oliguric AKI with signs of fluid overload (i.e. oliguria of less than 24 hours). If the initial dose is effective in improving urine output, an infusion of Furosemide (0.1–0.3 mg kg h) can be started in consultation with the nephrologist. Diuretic therapy should be used as bridging and not delay the need for renal replacement therapy (RRT) (when overload exceeds 10–15%).

4. Electrolyte management:
   - Hyperkalaemia is a common and life-threatening electrolyte abnormality seen in AKI. When there is a high index of suspicion, monitoring and immediate treatment of severe hyperkalaemia (K+ >6.5 mEq L or high K+ with ECG changes) are important in preventing cardiac dysrhythmia and deaths.
   - As fluid overload is common, sodium intake should be restricted to 2–3 mEq kg per day, and caution should be taken in using hyperosmolar fluids.
   - Potassium or phosphate supplements or replacement should be

avoided unless there is a significant deficit.

- Metabolic acidosis is common, and it is secondary to impaired filtration of acids and loss of bicarbonate absorption in the kidney. Use of sodium bicarbonate is reserved to treat severe acidosis.
- Hyperphosphataemia can be treated with oral phosphate binders, and calcium gluconate is used to treat hypocalcaemia.

5. Hypertension management:
   Glomerulonephritis causing AKI in children may end up requiring dialysis in many cases. Since some patients with hypertension may also be fluid overloaded, diuretics can be considered as an adjunct to antihypertensive medications. Specific treatment is further discussed in Chapter 16.3.
6. Nutrition:
   Children diagnosed with AKI need a high-calorie, high-protein, low-sodium diet. However, these recommendations and their role in AKI are largely expert-opinion based, and further evidence is needed.
7. Renal replacement therapy:
   Renal replacement therapy (RRT) includes haemodialysis (HD), peritoneal dialysis (PD), and continuous RRT (CRRT). Generally agreed indications to initiate the RRT are as below:
   - Renal:
     Uremia/azotaemia – >uraemic encephalopathy or pericarditis or haemorrhage; consider if urea >30–35 mM (no strict cutoff)
     Fluid overload: oliguria resulting in volume overload and respiratory distress
     Metabolic acidosis due to renal failure
     Hyperkalaemia
   - Non-renal
   - Toxins/drugs: small, non-protein bound
   - Sepsis – for removal of cytokines
8. Disposition:
   All children with AKI should be admitted for inpatient management.

Acute presentation of chronic renal failure

First-time presentation of undiagnosed CRF is a common differential of AKI. Anaemia, poor growth, clinical or radiological evidence of osteodystrophy or small kidneys on ultrasound may suggest CRF (though polycystic kidneys are large). Elevated P_{Cr} rising by <40 μmol L^{-1} day^{-1} is also suggestive that the renal failure is chronic. AKI can occur in a child with CRF, which may be a flare of the original disease or genuine AKI. Most CRF patients will already be under the care of a nephrologist, who should be contacted early. Symptoms such as anorexia, nausea, vomiting, malaise and sleep disturbances are common, but one should be alert for acute deterioration in any of these.

History should be sought regarding medications (diuretics, antihypertensive agents, steroids, erythropoietin, salt supplements, alkali, vitamin D, calcium carbonate phosphate binder), type and frequency of renal support (if appropriate), other illnesses and usual follow-up. When CRF is previously undiagnosed, a family history of Alport disease, haemophilia, polycystic kidney disease, sickling or stones should be sought. Ask for family history of ‘kidney failure’, ‘dialysis machine’, deafness, and blood or protein in urine.

The investigations for the acute presentation of CRF are similar to the investigations for AKI.

Acute problems in chronic renal failure include the following:

- Hypertensive crisis, possibly hypertensive encephalopathy with convulsions and coma
- Salt depletion, especially with superimposed gastroenteritis
- Dehydration can lead to irreversible deterioration in GFR caused by hypovolaemia. Urine output cannot be used to judge circulating volume. Hypertension may be normal for the child.
- Volume overload and heart failure
- Hyperkalaemia. See hyperkalaemia protocol in Chapter 10.6.
- Arrhythmias
- Acidosis. Chronic acidosis may be managed by oral alkali administration (2–3 mmol kg^{-1} day^{-1}).
- Hyper- or hypocalcaemia
- Urinary tract infection or obstruction
- Haemodialysis catheter or shunt or peritoneal dialysis catheter malfunction or infection
- Peritonitis in a child on continuous cycling or ambulatory peritoneal dialysis

- Acute gastrointestinal haemorrhage caused by platelet dysfunction. Desmopressin (DDAVP) is a therapeutic agent that may be useful, together with platelets, fresh frozen plasma or cryoprecipitate.
- Anaemia is often tolerated very well, even down to haemoglobin levels of 30–50 g L^{-1}. Do not transfuse without consulting the nephrologist, except in emergency. Transfusion, especially of platelets, may actually worsen prognosis in HUS (see Chapter 16.7).
- Fractures secondary to renal osteodystrophy.

Acute kidney injury in the renal transplant recipient

The cause of a rising creatinine or sudden oliguria must be diagnosed without delay as a transplant carries high stakes. AKI in the transplanted kidney is often reversible with prompt management. Contact the transplant/nephrology service urgently for all transplants presenting to the ED for any cause. Pre-renal uraemia must be excluded in any intercurrent illness.

Possible causes include:

- volume depletion – the transplanted kidney compensates less well to dehydration
- delayed, antibody-mediated rejection
- cyclosporin or tacrolimus toxicity (a glomerulopathy with hyaline arteriolar thickening)
- urine leak at the ureteric-vesical anastomosis
- ureteric obstruction
- acute tubular necrosis (rare in the child presenting to the ED)
- vascular thrombosis, including microangiopathic thrombosis diagnosed only on renal biopsy
- infection
- aciclovir.

Controversies and future directions

1 Role of novel biomarkers such as kidney injury molecule-1 (KIM-1), interleukin-18 (IL-18), and neutrophil gelatinase-associated lipocalin

(NGAL), urine TIMP-2 and IGFBP7 in the diagnosis and prognosis of acute kidney injury (AKI) management.

2 Acetylcysteine has been shown to ameliorate the acute progression of some causes of AKI, but its role is not yet defined.

3 Low-dose vasopressin and dopamine A1 receptor agonists, e.g. fenoldopam, are being evaluated for a role in improving renal blood flow and glomerular filtration rate (GFR).

4 Adenosine A1 receptor agonists are being evaluated for a role in improving renal blood flow and GFR, especially in association with furosemide.

5 Atrial natriuretic peptides may improve renal perfusion, particularly in oliguric renal failure.

6 Melanocyte stimulating factor, insulin-like growth factor, epidermal growth factor and hepatocyte growth factor, anti-adhesion molecule compounds, free radical scavengers and antioxidants are also being evaluated.

7 Endothelin A receptor antagonists may in the future play a role in moderating renal injury.

8 Stem cells to produce new renal tissue.

9 More accurate markers of GFR may become available, e.g. cystatin C.

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16.2

Haematuria

Ben Lawton

Haematuria can be either microscopic or macroscopic. The distinction is often considered to be visibility of blood in the urine to the naked eye. Microscopic haematuria is a relatively common incidental finding, often associated with fever or exercise, and investigation yields no diagnosis in around 80% of cases. Macroscopic haematuria on the other hand has an identifiable cause in the majority (60%) of patients. In adult patients presenting with macroscopic haematuria, the first consideration is the exclusion of malignancy. In paediatric patients presenting with macroscopic haematuria, malignancy is the cause in less than 1% of cases, and a more commonly important distinction is that between glomerular and non-glomerular aetiologies (Box 16.2.1).

Box 16.2.1 Common artifactual causes of red urine

- Foodstuffs:
    - Beetroot
    - Blackberries
    - Food colouring
- Medications:
    - Rifampicin
    - Iron
    - Metronidazole
    - Nitrofurantoin
    - Salicylates
    - Chloroquine
- Other:
    - Urates (common in neonates)

- Lead
- Porphyria

Isolated microscopic haematuria

This is a common and usually benign phenomenon. It is necessary to exclude both urinary tract infections (UTIs) and the presence of proteinuria, which is often possible on the same dipstick that identified the haematuria. The child should be examined with a focus on identifying oedema or abdominal masses, and the blood pressure should be measured accurately (see page 403, Chapter 16.3). Exploration of the family history for end-stage renal failure or other red flags as described below is prudent. Urine microscopy and culture will definitively identify or exclude a UTI and allow quantification of the degree of haematuria. It is then reasonable to discharge the child with a recommendation for the GP to repeat urine testing in a week or two when the child is well. If this microscopic haematuria is persistent over a period of weeks to months, or if red flags are identified in the history or examination, then investigation and specialist referral are appropriate, but this can usually occur as an outpatient.

History

Associated symptoms provide valuable clues in the differential diagnosis. Dysuria suggests infection or other source of inflammation in the urethra or bladder. Flank pain suggests pyelonephritis in the presence of fever or urolithiasis in the afebrile child. Precipitating sore throat or skin infection is typical in post-streptococcal glomerulonephritis while rash and arthritis are associated with rheumatological causes such as Henoch–Schönlein purpura (HSP) or systemic lupus erythematosus (SLE). Recent diarrhoeal illness, especially bloody diarrhoea, with or without the presence of a petechial rash is concerning for haemolytic uraemic syndrome (HUS). Globally schistosomiasis and tuberculosis are important causes of haematuria though these can often be easily excluded with an appropriate travel history.

Family history may be reassuring in the case of benign familial haematuria or concerning in the case of congenital deafness (suggesting Alport syndrome) or end-stage renal failure of any cause. Hypercalciuria is one of the more common causes of macroscopic haematuria and is often flagged by a family history of

kidney stones.

Examination

A thorough general examination may suggest signs of renal disease. Oedema should be deliberately sought in dependent areas, remembering that scrotal oedema can be mistaken for hydrocoeles in small boys, and parents are often better at identifying facial oedema in their own children than we are as clinicians meeting the child for the first time. Palpable purpura in a dependent distribution with associated joint swelling is classic for HSP, while petechiae may be found with idiopathic thrombocytopenic purpura (ITP) or HUS. Abdominal mass would be a rare but significant finding mandating imaging to exclude a Wilms' tumour (or a nephroblastoma).

Investigation

Urine dipstick will identify proteinuria, confirm the presence of blood and screen for UTI.

Microscopy and culture should proceed regardless as they are the definitive tests for UTI, may identify casts and/or dysmorphic red cells suggesting upper tract disease and will allow quantification of the red cells.

Proteinuria should be quantified with a protein:creatinine ratio in the first instance. Protein in the urine suggests glomerular disease and mandates specialist consultation. (See also Chapter 16.6.)

Urine calcium:creatinine ratio will identify hypercalciuria, most commonly familial, which is a common cause of haematuria. While familial hypercalciuria is benign it is associated with increased risk of kidney stones. This risk can be minimised with good hydration and a low-sodium diet.

Urea and electrolytes provide an assessment of renal function. Urea and creatinine are raised in renal failure while calcium and phosphate abnormalities occur in chronic renal failure. Potassium abnormalities can produce critical arrhythmias in the context of renal dysfunction.

Full blood count will identify HUS, ITP or anaemia and may suggest infection.

Plasma albumin will be reduced in nephrotic syndrome. Hypoalbuminaemia suggests significant protein loss is occurring which has consequences for both coagulation and immune function in addition to causing oedema.

Streptococcal serology/anti-streptolysin O titre/anti-DNase b indicates recent streptococcal infection which is the precipitant of post-streptococcal glomerulonephritis.

Complement (C3) is reduced in post-streptococcal glomerulonephritis, mesangiocapillary glomerulonephritis and SLE.

Coagulation profile abnormalities may be a cause of haematuria or a consequence of the protein loss from glomerulonephritis.

Renal ultrasound is of low yield but given the inclusion of malignancy (notably Wilms' tumour) on the list of differentials and the lack of associated pain or radiation exposure, USS is a reasonable way of reassuring both parent and clinician that the renal tract is structurally normal.

Renal biopsy is reserved for a small subgroup of patients, usually with steroid-resistant disease, while cystoscopy is very rarely indicated in the paediatric population.

Disposition

All patients presenting with haematuria will require follow-up. Those with hypertension, proteinuria or renal failure will likely require admission but, at the minimum, warrant discussion with a specialist and a robust management plan before discharge from the emergency department. Those with more straightforward diagnoses such as uncomplicated UTI or isolated microscopic haematuria may more appropriately follow-up with their GP.

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16.3

Hypertension

Ben Lawton

Introduction

Measurement of blood pressure is part of the complete assessment of any child presenting for emergency care. It is specifically indicated in patients with cardiac, neurological or renal conditions and those who are unwell or have syndromes or other risk factors known to be associated with hypertension.

Definitions of hypertension are based on the distribution of values measured in the population corrected for age, height and gender. Unlike in adults, the correlation between specific blood-pressure values and adverse clinical outcomes is not well defined in children. This means diagnostic criteria are based on expert consensus interpretation of population norms rather than anything more robust. Commonly cited reference tables are available in the 'fourth report on the diagnosis, evaluation and treatment of high blood pressure in children and adolescents' and, more accessibly, via the guidelines pages of the Royal Children's Hospital and Starship Children's Hospital (see Further reading). Though not designed for this purpose, paediatric early-warning score charts are a useful trigger to consider the diagnosis of hypertension.

Stage one hypertension is defined as a systolic and/or diastolic blood pressure between the 95th centile and the 99th percentile plus 5 mmHg. The average value of at least three measurements is required for diagnosis, meaning the role of the emergency provider in this instance is to identify the potential for diagnosis and refer for appropriate follow-up measurement.

Stage 2 hypertension is defined as a systolic and/or diastolic blood pressure above the 99th centile plus 5 mmHg. Symptomatic patients and those with evidence of end-organ damage warrant immediate specialist consultation. Those without these complications should be referred for rapid outpatient follow-up (within 1 week).

History

The symptoms of hypertension can be vague, variable or non-existent. Specific enquiry should be made about recent headaches, seizures, visual changes, epistaxis, abdominal pain and any caffeine, medication or illicit drug use. A thorough systems review is indicated and may reveal symptoms of an underlying disease process of which hypertension is a consequence rather than a cause.

The majority of cases of hypertension in the pre-adolescent population are secondary to an underlying disease process. Renovascular diseases are the most commonly identified underlying causes and may be flagged by a history of haematuria, recurrent urinary tract infections or umbilical artery catheterisation (which may be assumed if the child had a significant NICU stay). Endocrinopathies may be associated with a history of growth failure, heat/cold intolerance or a change in weight or appetite. Palpitations may point to cardiac cause while rheumatological disease may be heralded by joint pain and swelling, myalgias and/or recurrent rashes.

As children progress through adolescence a more adult pattern of disease comes to the fore with primary (essential) hypertension the leading cause. Snoring with or without sleep apnoea is commonly associated with primary hypertension.

Examination

BP may be measured by manual (aneroid) sphygmomanometry or, more commonly, automated oscillometry (e.g. DINAMAP). Automated oscillometry measures the mean arterial pressure (MAP). It then calculates the systolic and diastolic values rather than measuring them directly. This process is relatively accurate in all but the smallest (<5 kg) infants, in whom it has a tendency to overestimate values.

Use of an appropriately sized cuff is imperative if an accurate blood pressure is to be measured. The cuff should be the widest that can be applied and must cover at least two-thirds of the upper arm, with a length sufficient to completely encircle the arm. Use of too small a cuff will result in a spuriously high blood pressure reading while the artefactual effects from too large a cuff are minimal. If there is any doubt it is better to use a cuff that is too big than one that is too small.

Ideally the child should be seated and relaxed for 5 minutes before the blood

pressure is taken on the right arm, with the cuff at the same height as the heart. While this is not always immediately possible in the emergency department (ED), measured BP should be interpreted with this ideal situation in mind and also with consideration to any medication the child may have recently received.

Examination should look for evidence of both causes and consequences of hypertensive disease.

A thorough general examination may reveal clues to the underlying cause. Plotting on a growth chart and calculation of BMI are essential. Growth failure suggests underlying systemic disease while obesity is commonly associated with primary hypertension, especially in the adolescent population. Displacement of the cardiac apex would suggest significant left-ventricular hypertrophy while radio-femoral delay is a classic finding in aortic coarctation.

The most commonly recognised manifestations of end-organ damage from uncontrolled hypertension are left ventricular hypertrophy, retinopathy and proteinuria. Initial assessment of the hypertensive child should therefore include an ECG, fundoscopy and a urine dipstick, with protein:creatinine ratio if positive. It should be acknowledged that echocardiography is more sensitive than ECG and formal ophthalmological assessment will be superior to assessment in the ED. These should therefore be arranged as follow-up, but efforts should still be made to identify end-organ damage at the time of presentation.

The symptomatic patient (headache, visual disturbance, seizure, epistaxis, abdominal pain) should be managed immediately as described under ‘severe hypertension’ below.

Emergency department management

Stage 1 hypertension

This warrants a specific follow-up plan for repeated measurements to establish the diagnosis.

Stage 2 hypertension

Assessment for end-organ damage (ECG, fundoscopy, urine dipstick), if end-organ damage is identified in the ED, then management is as per ‘severe hypertension’ below.

Without evidence of end-organ damage the following investigations should be

performed:

- Urine for microscopy and culture
- Blood for urea and electrolytes with calculation of estimated glomerular filtration rate and attention to renal function and acid–base status
- Chest X-ray
- Renal ultrasound scan (with Doppler if significant suspicion of renal artery disease, which will be the case in most pre-adolescent patients. The performance of renal artery Doppler is a specialised skill, and this request should be discussed with your local radiology service.)

If the above are abnormal, immediate specialist consultation should be sought.

If normal then the patient can be discharged home after arranging urgent specialist outpatient follow-up, an echocardiogram and a formal ophthalmological assessment.

Severe hypertension

The presence of clinical symptoms or of end-organ damage mandates immediate lowering of the blood pressure. Precipitous drops can be dangerous, and the aim is to lower the blood pressure over 24–48 hours with no more than a 25% drop in systolic BP in the first 12 hours. Consistent management plans increase the chances of a smooth reduction to acceptable levels so early discussion with the receiving paediatric intensivist and nephrologist is strongly encouraged.

Hypertensive encephalopathy usually presents with a severe headache, visual disturbance and vomiting and may progress to focal neurological deficits, seizures and impaired conscious state, with grossly elevated BP. On fundoscopy, papilloedema or retinal haemorrhages may be detected. Such patients usually have underlying chronic renal disease with many being on dialysis. The differential diagnosis includes uraemic encephalopathy and metabolic disturbance. In this setting, BP should be lowered in a controlled fashion, with anticonvulsants given for seizures.

Initial management is typically with an IV beta-blocker, either esmolol or labetolol. These are commenced as an infusion in this circumstance. Contraindications to beta-blockers include asthma and heart failure. The combination of beta-blockers and calcium channel blockers can have catastrophic negative inotropic effects. Beware of the patient with known

hypertension who presents with a symptomatic exacerbation as these patients may well be on a regular calcium channel blocker prior to presentation.

If beta-blockers are contraindicated or insufficient on their own a peripheral vasodilator such as sodium nitroprusside can be used. Cyanide toxicity is a risk with this drug in larger doses or in renal failure. Alternative vasodilators include hydralazine and clonidine. Diazoxide should be avoided due to the dangers of its rapid, large and sustained effect.

Children with known hypertension whose presentation is clearly acute may tolerate a more rapid decrease in blood pressure, and children with significant renal failure will benefit from emergent dialysis. Both these circumstances warrant immediate discussion with the patient's nephrologist.

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16.4

Urinary tract infection in pre-school children

Robert Melvin

ESSENTIALS

1 Urinary tract infection (UTI) is common and potentially serious.
2 Septicaemia may occur in babies.
3 UTI should be considered in all febrile infants and young children, as well as neonates who are non-specifically unwell.
4 In young children, UTI cannot be diagnosed reliably or excluded on clinical grounds.
5 Dipstick urinalysis is an unreliable method of ruling out UTI in infants.

Introduction

Bacterial infection of the urinary tract (UTI) is common in the paediatric age group. Its significance is greatest in young children, particularly in the first year or two of life, where the high incidence of upper tract infection (pyelonephritis) and the presence of immature kidneys lead to potential for renal scarring (reflux nephropathy).

A meta-analysis examining prevalence of UTI in febrile children (≥38°C) showed that uncircumcised male infants less than 3 months of age and females less than 12 months of age had the highest baseline prevalence of UTI.[1] Girls are much more likely to suffer from a UTI than boys, with up to 10% of girls having at least one UTI by age 10. It is important to remember that recurrences are common. UTI is more frequent in boys than girls in the first months of life,

partly because of a higher incidence of obstruction including pelviureteric junction obstruction and partly due to lack of circumcision, thereafter occurring significantly more often in girls (Table 16.4.1).

UTI is caused by organisms normally resident in the gastrointestinal tract. *E. coli* is the commonest organism (>80% of cases). It is thus an ascending infection that may affect the bladder (cystitis) or upper renal tract (pyelonephritis). Neonates are unusual as they may also develop UTI following haematogenous dissemination of organisms.

Predictors for renal scarring after first UTI include:[2]

- vesicoureteric reflux (VUR)
- abnormal renal ultrasound (either pre-natal or post-natal)
- high C-reactive protein >40 mg L
- temperature ≥39°C
- UTI caused by organism other than *E. coli*.

History and examination

In infants and young children with UTI, the clinical history is frequently non-specific and may include irritability, jaundice (neonates), poor feeding or fever without apparent source. Symptoms and signs become more specific with increasing age (Table 16.4.2).

Previous UTI, increasing pain/crying on passing urine, increasingly smelly urine, and absence of severe cough or ear pain are all suggestive of UTI.[3]

Table 16.4.1

Prevalence of urinary tract infection in febrile∗ infants and children by demographic group

Demographic group	Prevalence or pretest probability (95% CI)
0 to 3 months	**7.2% (5.8–8.6)**
Girls	7.5% (5.1–10)
Circumcised boys	2.4% (1.4–3.5)
Uncircumcised boys	20.1% (1.7–11.5)
3 to 6 months	**6.6% (1.7–11.5)**
Girls	5.7% (2.3–9.4)

Boys	3.3% (1.3–5.3)

6 to 12 months	**5.4% (3.4–7.4)**
Girls	8.3% (3.9–12.7)
Boys	1.7% (0.5–2.9)

12 to 24 months	**4.5%****
Girls	2.1% (1.2–3.6)
Boys	<1%**
<19 years with urinary symptoms and/or fever***	7.8% (6.6–8.9)

* Temperature >38°C.

** 95% confidence interval not available.

*** Most of these children were older than 2 years.

Data from Shaikh N, Morone NE, Bost JE, Farrell MH. Prevalence of urinary tract infection in childhood: a meta-analysis. Pediatr Infect Dis J 2008;**27**:302.

Table 16.4.2

Urinary tract infection symptoms by age

Age	0–2 years	2–5 years	5–12 years
Failure to thrive	✓		
Feeding problems	✓		
Screaming	✓		
Irritability	✓		
Diarrhoea	✓	✓	
Vomiting	✓	✓	
Fever	✓	✓	✓
Convulsions	✓		
Haematuria	✓	✓	
Urinary frequency	✓	✓	
Dysuria	✓	✓	
Enuresis	✓	✓	
Abdominal pain	✓	✓	
Loin pain	✓		

Don’t forget to obtain a family history of renal tract disease, in particular regarding UTIs, VUR (35% for children of an affected parent[4]) and renal impairment.

Physical examination of young children with UTI is often unremarkable or non-specifically abnormal. Loin or suprapubic tenderness with normal respiratory and ear examination should increase your suspicion of UTI. Septicaemia can occur more frequently with UTI in infancy and must be considered in babies up to around 6 months of age. Fever is the best clinical marker of pyelonephritis in infants with UTI but is non-specific.

Diagnosis

A reliable urine sample is required to establish the diagnosis of UTI. In older children this is usually accomplished by obtaining a midstream sample. Difficulties arise in children too young to have been toilet trained, and this group is also at highest risk for developing pyelonephritis and renal scarring.

In infants and toddlers, urine bag samples are unreliable (very high false-positive rate) and should not be used. Clean-catch samples are more reliable and are the preferred method for non-invasive urine collection. Urine collection pads are available too but need to be changed every 30 minutes to reduce contamination rates.[5] If samples are required urgently, bladder catheterisation or suprapubic aspiration (SPA) is necessary. The yield from SPA is markedly improved by using ultrasound to confirm a full bladder. SPA is the gold-standard method for urine culture; however, SPA has a higher failure rate than catheterisation.

Samples should be sent to the laboratory for urinalysis, microscopy and culture. Findings supportive of the diagnosis of UTI include the presence of leucocytes; organisms on microscopy; and leucocyte esterase AND nitrites on dipstick urinalysis. Organisms may be seen on Gram stain. If there will be a delay of >4 hours before culture can be performed, the urine sample should be refrigerated.

Dipstick urinalysis may be helpful in making a provisional diagnosis of UTI. However, a negative result does not rule out UTI in infancy. One study showed that urinalysis was normal in 50% of infants <8 weeks with confirmed UTI. Another study suggested that dipstick urinalysis was a reliable method of ruling out UTI only after age 2 years.

The traditional definition of pyuria is >5 white blood cells (WBC) per high-power field (centrifuged urine). Another definition is >10 WBC mm^{-3} (uncentrifuged urine).

The definition of significant bacteriuria is guided by the method by which the urine specimen was collected (Table 16.4.3), though on occasion genuine UTI may be present with lower colony counts than would usually be considered significant, especially in babies – interpret results in light of history and clinical findings.

Treatment

ED treatment recommendations for UTI vary.

Age <6 months

- Admit all for intravenous (IV)/intramuscular (IM) antibiotics (septicaemia occurs more commonly in this age group)
- Admit to medical ward (observation ward only if systemically well)
- If discharged from observation ward, arrange GP or ED follow-up at 24–48 hours
- Ensure urine culture and sensitivity results are checked at 48–72 hours.

Age 6 months to 3 years old

- If systemically unwell, admit to observation ward or medical ward for IV/IM antibiotics

Table 16.4.3

Definition of significant bacteriuria

Method of collection	Colony-forming unit count (CFU mL^{-1})
Clean catch	$>10^8$
Catheter	$>10^3$
Suprapubic	>0

- If not systemically unwell, consider single IM/IV dose of ceftriaxone or gentamicin, followed by oral antibiotics
- Arrange GP or ED follow-up at 24–48 hours
- Ensure urine culture and sensitivity results are checked at 48–72 hours.

Age >3 years old

- Treat on clinical merits, i.e. admit or allow home, parenteral or oral antibiotics
- Overnight admission to observation ward for IV/IM antibiotics may be worth considering if unwell
- Ensure urine culture and sensitivity results are checked at 48–72 hours.

Antibiotic choice[6,7]

Empirical antibiotic choice, dose and duration should be guided by local sensitivity patterns and antibiotic guidelines. However, consideration may involve the following:

Parenteral treatment (IV/IM)

- Benzylpenicillin 60 mg kg^{-1} (max 2 g) every 6 hours AND gentamicin 7.5 mg kg^{-1} (max 360 mg) once daily, or
- Ceftriaxone 50 mg kg^{-1} once daily IV (may not cover enterococci in young infants), or
- Gentamicin 7.5 mg kg^{-1} once daily IV/IM (single drug for initial therapy if penicillin hypersensitivity). Reduce gentamicin to 2.5 mg kg^{-1} as a single dose in patients with known or suspected renal impairment, and check a level before repeat dosing.

Oral treatment

- Amoxicillin + clavulanic acid (22.5 + 3.2 mg kg^{-1}) every 12 hours, or
- Cephalexin 12.5 mg kg^{-1} every 6 hours, or
- Trimethoprim + sulfamethoxazole (co-trimoxazole) (4 + 20 mg kg^{-1}) every 12 hours, or
- Trimethoprim 4 mg kg^{-1} (max 150 mg) every 12 hours
- Amoxicillin is not recommended.

Duration of treatment?

- Infants <2 years old: treat for 10 days at full dose
- Older children: generally treat for 7 days at full dose
- Courses as short as 3 days may be used in older children with uncomplicated lower tract UTI.

Management after discharge from emergency department or observation wards

Most centres will have a referral, management and investigation protocol which

should be followed. Many of these are based on the National Institute for Clinical Excellence Guidelines from the United Kingdom,[8] though significant variation exists from place to place. In the absence of local guidelines, consider the following outlined below.

If antibiotic sensitivity is known, a ‘proof of cure’ urine culture is not required.

Antibiotic prophylaxis is controversial. Generally, it is only required for recurrent UTIs or known VUR. Asymptomatic bacteriuria should not be treated with antibiotics. Co-trimoxazole (because of its long shelf life), trimethoprim or nitrofurantoin is a reasonable choice. The trimethoprim dose is 2 mg kg^{-1} at night for both drugs (expressed as the trimethoprim component in the case of co-trimoxazole). Trimethoprim suspension is unavailable commercially in Australia but is a better choice than co-trimoxazole in countries where it is readily obtainable.

Arrange a renal tract ultrasound for children <6 months old or with an atypical UTI, and refer to paediatric outpatient clinic. Ultrasound is not routinely required for uncomplicated first UTI in children >6 months old.

It is generally not necessary to book other imaging studies from the ED as consultants will differ in the investigations that they prefer, e.g. micturating cystourethrogram (MCUG) and/or dimercaptosuccinic acid (DMSA) isotope scan, though this can be expedited by speaking to the consultant or unit to whom the child has been referred.

Prognosis

A systematic review[9] involving nearly 5000 patients looked at short-term outcome of first UTI in children (<19 years) and revealed:

- 25% had VUR, 2.5% with grade IV or V- VUR
- VUR is associated with an increased risk of developing acute pyelonephritis and renal scarring, particularly with higher grade VUR
- 15% had evidence of renal scarring on DMSA scan, but the significance of this was unclear
- 8% of children had a least one recurrence.

However, most children with UTI will not suffer any long-term sequelae. The association between renal scarring and hypertension, chronic kidney disease and

end-stage renal failure remains unclear.

Prevention

The ‘basics’ of good fluid intake, regular and complete voiding, avoidance of constipation and proper (front to back) bottom wiping (in toilet-trained females) should be encouraged in order to reduce the likelihood of repeat infections. There is also some evidence of benefit from the regular use of cranberry juice (which acidifies urine and reduces bladder wall adhesiveness). Prophylactic antibiotics should be considered in cases of VUR in pre-school children,[10] recurrent pyelonephritis, recurrent symptomatic UTI in older children and in pre-school children awaiting renal tract imaging.[11]

Controversies and future directions

1. Current controversies and areas for potential investigation in paediatric urinary tract infection (UTI) include the following:
    - Parenteral or oral antibiotic treatment
    - Duration of antibiotic treatment
    - Outpatient or inpatient management
    - Post-UTI imaging protocols
    - The role of antibiotic prophylaxis
    - Diagnosis and management of vesicoureteric reflux
    - ‘Complementary’ therapies such as lactobacillus and bifidus consumption
    - Cranberry juice as prophylaxis
    - Recognition of dysfunctional voiding
    - Recognition of sphincter and detrusor dyssynergia.
2. The clinical impact of vesicoureteric reflux and its management remain uncertain and controversial; whether it may lead to progressive renal impairment and potentially end-stage renal disease is also currently being questioned.

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16.5

Haemolytic uraemic syndrome

Linas Dziukas

Introduction

As a group, the conditions that cause haemolytic uraemic syndrome (HUS) range from uncommon to rare, have many different causes but share similar clinical features because they have a common pathological endpoint of occlusive microvascular disease called thrombotic microangiopathy (TMA).

The pathological features of TMA include vessel-wall thickening, swelling and detachment of the endothelial cell from the basement membrane, accumulation of material in the subendothelial space, intraluminal platelet thrombosis and partial or complete vessel luminal obstruction (by platelets fibrin or both). In some cases there is fibrinoid necrosis of arteriole walls (Fig. 16.5.1).

The widespread microvascular changes cause mechanical damage and fragmentation of red cells, causing a microangiopathic haemolytic anaemia (MAHA). Platelets are consumed by the TMA process, so thrombocytopaenia is a consistent finding. The coagulation system is not directly impaired in TMA, so the standard clotting tests are normal.

MAHA is a non-immune haemolytic anaemia resulting from intravascular red-cell fragmentation with the following features:

- Abnormal red cell shapes (schistocytes) seen on blood film

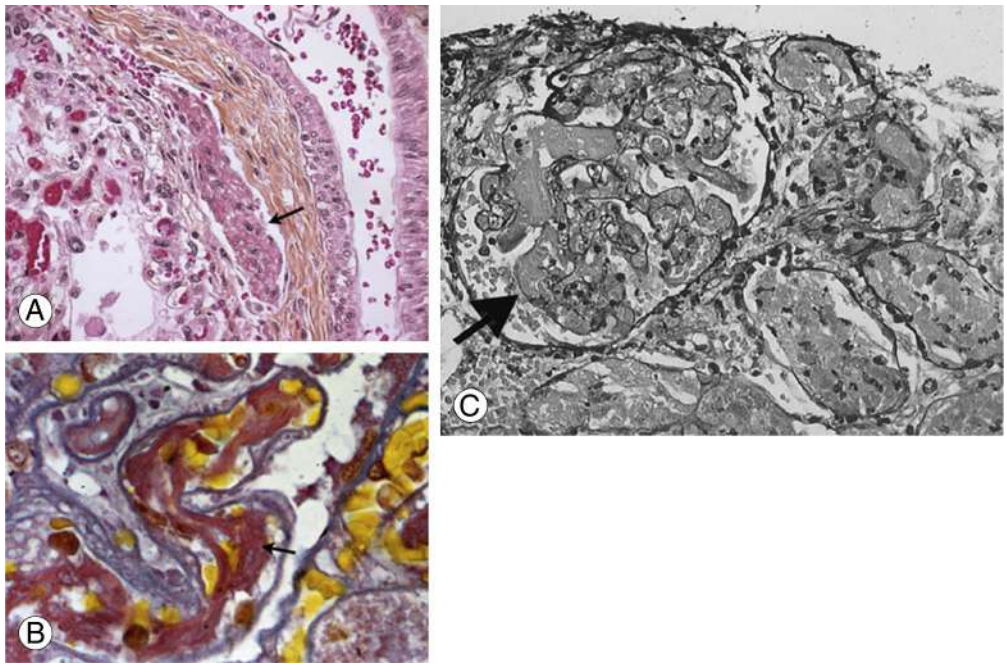

FIG. 16.5.1 Light microscopy showing thrombotic microangiopathy in pulmonary vessels (A) and in glomerular capillaries (B and C). A: Credit to Humpath.com. C: Modified from Apoptosis of renal cortical cells in the hemolytic-uremic syndrome: in vivo and in vitro studies. *Infection Immunity* 1998;**66**:636–44.

- Reticulocytosis
- Thrombocytopaenia due to consumption of platelets
- Markedly elevated lactate dehydrogenase (LDH)
- Undetectable haptoglobin levels
- Moderately elevated bilirubin concentration
- Negative Coombs test and normal coagulation tests.

Definition of haemolytic uraemic syndrome

HUS is defined by the simultaneous occurrence of:

- microangiopathic haemolytic anaemia (MAHA)
- thrombocytopaenia
- acute kidney injury (AKI).

HUS is the commonest cause of acute renal failure in infants and young children less than 5 years of age in developed countries.

Classification of haemolytic uraemic syndrome

Haemolytic uraemic syndrome can be classified into two types, depending on the presence of a diarrhoeal prodrome. The commonest form of HUS in children is the one that occurs following a prodromal illness of acute gastroenteritis with bloody diarrhoea. This diarrhoea-associated haemolytic uraemic syndrome (D+HUS) accounts for 90% of cases. D+HUS is most often caused by Shiga toxin (verotoxin)-producing *Escherichia coli* (STEC, also called VTEC). Other cytotoxin-producing bacteria such as *Shigella dysenteriae* type 1, *Salmonella typhi* and *Campylobacter* are less common causes of HUS. The annual incidence of STEC HUS is about 2 to 3 per 100,000 children aged less than 5 years old.

Non-diarrhoeal-associated HUS (D–HUS) cases account for 10% of cases. These are associated with:

- non-enteric infections, e.g. *Streptococcus pneumoniae* or human immunodeficiency virus (HIV). Pneumococcal-associated HUS may cause up to 40% of non-STEC HUS cases in children. Patients with pneumococcal-associated HUS present with pneumonia (in two-third of cases) or meningitis.
- congenital or acquired abnormalities of the complement system (complement-mediated HUS). The trigger event is usually an upper respiratory tract infection, but some cases have a diarrhoeal prodrome. Pregnancy in adolescents has also been a trigger factor. There may be a positive family history of HUS or a history of previous HUS episodes.
- immunosuppression: malignancy-associated, following bone-marrow transplant, and drug-related, e.g. mitomycin, cyclosporin and FK-506 (tacrolimus)
- inborn error of cobalamin C metabolism.

Pathophysiology of haemolytic uraemic syndrome

Shiga toxin haemolytic uraemic syndrome

Shiga toxin-producing *E. coli* (STEC) are not a normal part of the human intestinal flora but are present in the intestines of 1% of healthy beef cattle. There are over 100 different serotypes of STEC with different phage types and

subtypes. STEC O157:H7 is the commonest subtype producing disease in North America, the British Isles and Japan. It is rare as a causative agent of HUS in Australia, where other types, including O111:H, are pathogenic. D+HUS occurs in epidemics as well as sporadically. Outbreaks can be traced to contaminated food, especially undercooked hamburger and contaminated water.

STEC organisms release Shiga toxins that attach to and damage the endothelial lining of the intestine, resulting in haemorrhagic and ulcerative lesions. This causes diarrhoea which is usually self-limited. The whole digestive tract can be damaged by the Shiga toxin, causing serious complications that can include severe haemorrhagic colitis, bowel necrosis and perforation, rectal prolapse, peritonitis and intussusception.

Children infected with *E. coli* O157:H7 are symptomatic; infected adults may be asymptomatic. Only 10 to 15% of children who have STEC infection with diarrhoea develop haemolytic uraemic syndrome. Young children and older persons with altered immune response, as well as persons who have been in contact with infected farm animals, are particularly vulnerable.

In cases of HUS, Shiga toxin enters the circulation, probably facilitated by attachment to neutrophils. The toxin damages vascular endothelium and produces TMA. Human microvascular endothelial cells are more susceptible to the toxic effects of Shiga toxin than large-vessel endothelium.

STEC-HUS occurs approximately 6 days to 14 days after ingestion of contaminated food or beverage and 2 days to 6 days after the onset of enteritis. Diarrhoea occurs in over 90% of cases of STEC enteritis. It is accompanied by severe crampy abdominal pain and stools that change from watery to haemorrhagic.

Pneumococcal-associated haemolytic uraemic syndrome

The incidence of HUS following pneumococcal infection is estimated to be about 0.5%. *Streptococcus pneumoniae* (*S. pneumomiae*)-related HUS may be caused by exposure of the Thomsen-Friedenreich (TF) crypt antigen found on the surface of erythrocytes, platelets and glomerular endothelial cells. The TF antigen is normally masked by neuraminic acid. All serotypes of *S. pneumoniae* produced neuraminidase, which exposes the TF antigen. The host then produces IgM antibodies, which bind to the TF antigen. This initiates an immune response that culminates in the development of HUS.

Complement-mediated haemolytic uraemic syndrome

This rare disorder involves an abnormality of the alternative complement pathway. A trigger event or factor leads to uninhibited complement activation that damages the vascular endothelium.

History

A child who has previously been healthy presents with a history of gastroenteritis that has developed over the past 2 weeks. There is vomiting, bloody diarrhoea and cramping abdominal pain. The child is usually afebrile at presentation. The presentation may resemble an acute surgical abdomen.

Typically the patient appears to be improving from his/her gastroenteritis but then his/her condition suddenly worsens. The child then becomes pale and develops haematuria and oliguria and becomes lethargic. Loss of appetite, nausea and vomiting may be observed. Decreased urine output may be reported by some patients.

Examination

The child appears pale and lethargic and may be jaundiced. Petechiae and purpura are uncommon features**.** About one-half of patients have an elevated blood pressure. Hepatomegaly may be present, and there may be peripheral oedema.

Up to one-third of children with HUS will have some neurological symptoms. This can include irritability, altered mental state or seizures. Less common neurological complications include hemiparesis, facial palsy, pyramidal or extrapyramidal syndromes, dysphasia, diplopia, cortical blindness or coma.

Investigations

1. Features of MAHA are present (see Introduction). Anaemia is usually severe and is normochromic normocytic in type with a haemoglobin level of 50–90 g L^{-1}. Schistocytes, burr cells, helmet cells and triangle cells may be seen in the blood film. The white cell count is often raised with increased number of immature forms. The platelet count is low in

95% and can be as low as 20×10^9 L^{-1}. Subsequent increase in platelet count may be the first indication of resolution of the microangiopathic process.

2. Serum electrolyte measurement: renal impairment can cause increased concentrations of urea and creatinine, hyponatraemia, hyperkalaemia or metabolic acidosis.
3. Urinalysis shows haematuria (that can be macroscopic) and proteinuria. Urine microscopy can show dysmorphic red blood cells, and granular casts or hyaline casts or both.
4. Coagulation tests (prothrombin time, partial thromboplastin time) are normal.
5. Blood complement concentrations (C3 or C4) are normal in STEC-HUS but may be reduced in complement-mediated HUS.
6. Other blood tests: elevated inflammatory markers (CRP); raised lipase/amylase reflecting pancreatic involvement.
7. Cultures: blood cultures, urine cultures and stool culture (for Shiga toxin-producing *E. coli* O157:H7).

The degree of anaemia or thrombocytopaenia is unrelated to the severity of renal dysfunction, but an increased white blood cell count is associated with a worse prognosis.

Differential diagnosis

The differential diagnosis includes the following conditions:

- Intussusception
- Disseminated intravascular coagulation (DIC) in patients with sepsis and multiorgan failure
- Idiopathic thrombocytopenic purpura (ITP)
- Systemic lupus erythematosus (SLE) with renal involvement
- Leukaemia
- Vasculitis
- Post-infectious glomerulonephritis.

The clinical features and the findings on blood film, coagulation profile, urinalysis, stool and blood cultures assist in differentiating all these conditions

from typical HUS. There is no specific diagnostic test for HUS.

Treatment

There is no specific therapy for Shiga toxin-producing *E. coli*, and prevention of the disease is therefore of utmost importance.

Supportive therapy may include dialysis, antihypertensive therapy, blood transfusions and management of neurological complications. With supportive therapy, 85% of children recover renal function.

The cornerstone of treatment is careful attention to electrolyte and fluid balance. Once intravascular volume has been restored, the amount and type of fluid administered should be limited to ongoing losses, i.e. insensible loss, urine output and gastrointestinal losses.

No added potassium is required unless serum levels are below normal values. Hyperkalaemia must be anticipated and treated in a timely fashion (refer to management of hyperkalaemia in Chapter 10.6 on electrolytes).

Anaemia should be treated with packed red blood cell transfusion (10 mL kg^{-1}) when anaemia is severe, the patient is symptomatic or the haematocrit is falling rapidly. The Hb concentration should be maintained at an Hb greater than 70 g L^{-1} when possible.

Platelet transfusion has the potential risk of worsening the patient's clinical status. This results from the aggregation of platelets, which are a major constituent of microthrombi and thus could induce further damage. Platelet transfusion can be used if severe thrombocytopaenia is associated with active bleeding or before surgical procedures.

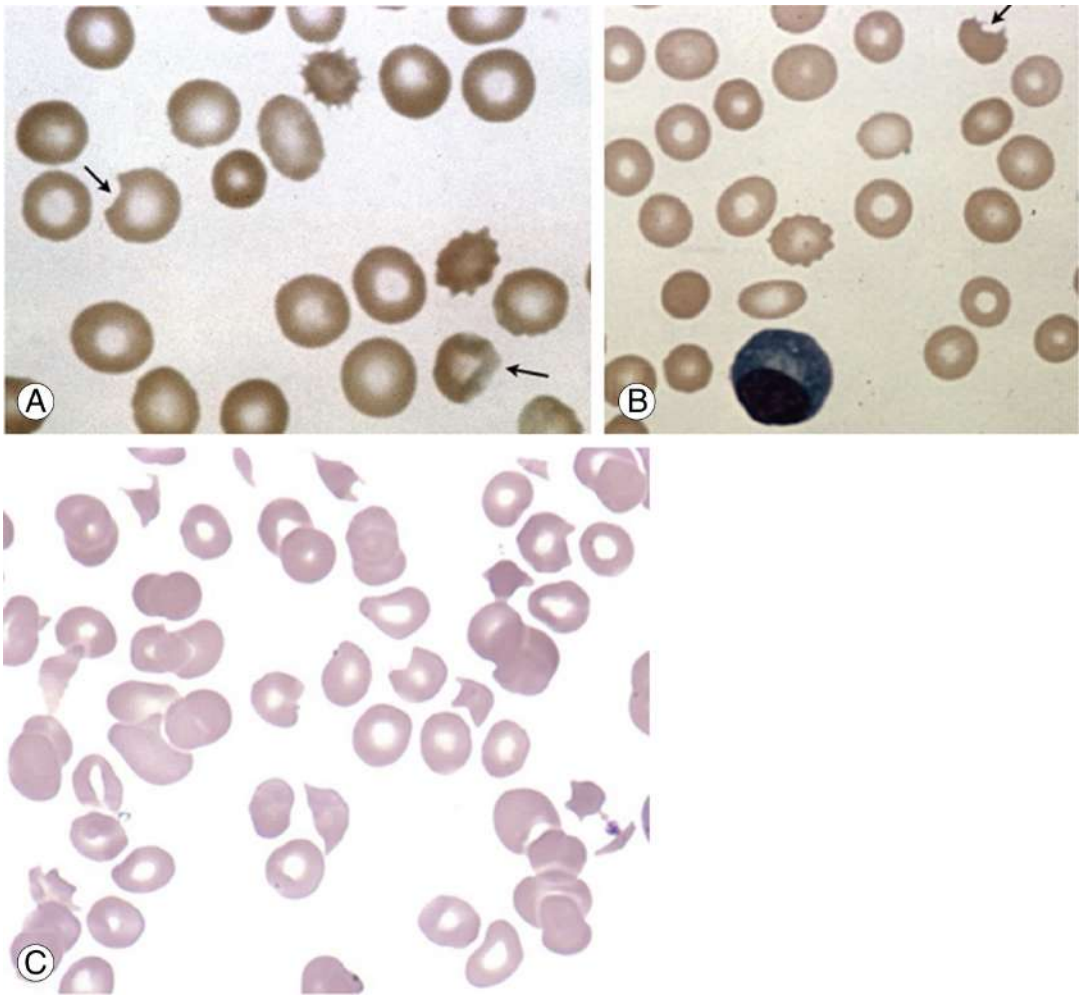

FIG. 16.5.2 Blood films showing mild microangiopathic haemolytic anaemia (MAHA) (A and B) and severe MAHA (C).
MAHA is most often caused by thrombotic microangiopathy (TMA). Other causes include disseminated intravascular coagulation, intravascular haemolysis caused by prosthetic heart valves or systemic disorders that may or not be associated with TMA. This image was originally published in ASH Image Bank. Peter Maslak; Lisa Southern. Microangiopathic hemolytic anemia, ASH Image Bank, 2009, # 00004049 © the American Society of Hematology.

Hypertension responds well to treatment with short-acting calcium channel blockers, e.g. nifedipine. Intravenous nitroglycerine can be used if oral medication is not tolerated. Labetalol by intravenous bolus or continuous infusion can also be used to manage hypertension (see Chapter 16.3). Treatment of hypertension can prevent development of encephalopathy and congestive heart failure.

Seizures should be treated with short-acting benzodiazepines initially, followed by intravenous infusion of phenytoin or phenobarbital.

There is some evidence that early dialysis may improve outcome in HUS.

Dialysis is needed in the following situations:

- Severe hyperkalaemia uncontrollable by medical means
- Fluid overload and pulmonary oedema
- Significant uraemic symptoms
- Blood urea >36 mmol L^{-1} even if electrolyte and water balances are satisfactory
- Anuria.

Peritoneal dialysis is generally used in infants and preschool-age children except in cases with severe colitis or bowel perforation.

There are no controlled clinical trials demonstrating efficacy of antibiotic therapy on the prevention and amelioration of HUS. Treatments such as plasmapheresis, anti-platelet drugs, anticoagulants, anti-thrombotic agents, steroids or Shiga toxin-binding agents are ineffective.

Nutritional requirements must be addressed aggressively, as these patients are catabolic and hypoalbuminaemic. Enteral feeds can be commenced once the diarrhoea has settled. Total parenteral nutrition is required in some cases.

Prognosis

The mortality of STEC-HUS is about 5%. About half the patients will need short-term dialysis, and about 10–30% of patients will have signs of chronic renal disease. About 5–10% of HUS patients will develop end-stage renal failure. In HUS not due to an infection nearly half develop end-stage renal disease.

The risk of renal sequelae in children with STEC-HUS is greater in males and those with hypertension, prolonged anuria and haemoglobin less than 100 g L^{-1} at onset of disease.

Other poor prognostic indicators include:

- Elevated white cell count (>20 × 10^9 L^{-1}) at presentation
- Elevated white cell count at presentation and which remains elevated
- Age <1 year or >5 years
- Central nervous system involvement
- The degree and duration of renal dysfunction.

Complications

Complications during the initial episode of HUS include stroke (occurs in 3% to 5% of patients), other central nervous system problems (seizures, coma and cortical blindness), bowel problems (hemorrhagic colitis, bowel necrosis, bowel perforation or intussusception), myocarditis, elevated serum troponin, pancreatitis, glucose intolerance or cholestatic jaundice.

Children with STEC-HUS need prolonged follow-up even after apparent full recovery as some can develop late complications. These complications include hypertension, proteinuria, reduced glomerular filtration rate and eventual development of end-stage renal disease (which can develop as late as 15 years to 25 years following recovery). HUS can recur in transplanted kidneys regardless of the aetiological agent.

Prevention

STEC-HUS is an infectious disease and the most effective prevention strategy would be to prevent ingestion of the *E. coli.* Avoidance of undercooked meat can assist in this area. There are, however, other vectors for the transmission of the *E. coli*, and these include contaminated water and beverages. Food handlers, vendors and consumers must be made aware of proper food-handling techniques.

The incubation period for *E. coli* O157:H7 is usually 3 to 4 days but can range from 1 to 8 days. The infectivity of children colonised with *E. coli* O157:H7 is up to 3 weeks in children.

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16.6

Idiopathic nephrotic syndrome

Linas Dziukas

Introduction

Nephrotic syndrome (NS) is a particular combination of clinical findings, urine abnormalities and changes in the blood. The underlying cause is a marked increase in the permeability of the glomerular filtration barrier (GFB) to protein, resulting in the following changes:

- Excretion of large quantities of protein in the urine (**marked proteinuria**), causing a decrease in the serum albumin concentration (**hypoalbuminaemia**)
- Retention of sodium by the kidney, causing an increase in the size of the interstitial compartment that causes **oedema**
- Changes in lipid metabolism with:
    - Hypercholesterolaemia
    - Fat droplets in renal tubular cells
    - Fat in the urine (lipiduria): free fat or fat within renal tubular cells.

There are other changes in the concentration of blood components that affect in particular the immune system and the coagulation system.

Measurement of the blood concentration of lipids is not as available as other blood tests, so a working diagnosis of NS is the combination of oedema, marked proteinuria and hypoalbuminaemia.

Patients without oedema but with marked proteinuria are said to have a nephrotic level of proteinuria.

Pathophysiology of proteinuria

Nephron and glomerular filtration barrier

Each human kidney is an aggregate of about one million nephrons. Each nephron has a globular enlargement that is invaginated by capillaries, forming the renal corpuscle (**glomerulus**) (Fig. 16.6.1).

The glomerulus has four structural components:

- Mesangium
- Glomerular capillaries
- Glomerular basement membrane (GBM)
- Visceral epithelial cells (VEC) or podocytes (PO).

The plasma in the glomerular capillaries crosses three structures (collectively called the glomerular filtration barrier [GFB]) to become the glomerular filtrate.

The GFB consists of:

- glomerular capillary endothelium
- glomerular basement membrane
- podocyte slit: the foot processes of the podocytes and the slit diaphragm (a specialised type of intercellular junction) connecting adjacent podocyte foot processes.

The GBM contains embedded anionic molecules that retard the filtration of negatively charged molecules in the blood (Fig. 16.6.2).

The GFB thus acts as a ‘sieve’ that allows small molecules and water to be filtered across the GBM but retards or prevents the passage of larger molecules based on their size and shape and charge.

Renal handling of albumin

Albumin is the most abundant plasma protein, accounting for 55–60% of the measured serum protein. It consists of a single polypeptide chain of 585 amino acids and has a molecular weight of 66500 Da. In solution the molecule is ellipsoid and is negatively charged (Fig. 16.6.3).

Albumin has a major role in the maintenance of the normal colloid oncotic pressure, contributing to 80% of the normal oncotic pressure. It is a flexible molecule that binds and transports many substances: long-chain fatty acids, bilirubin, calcium and magnesium. Albumin is also a secondary or tertiary

carrier for substances that have specific binding proteins, e.g. vitamin D and thyroxine.

The binding of drugs to albumin affects the delivery of the drug to tissue sites and the metabolism and elimination of the drug.

The normal serum albumin concentration is 32–45 g L, i.e. 32–45 mg mL. The amount of albumin entering the (two) kidneys (the ‘renal albumin load’) is determined by the serum albumin concentration and the renal plasma flow. The ‘renal albumin load’ in a healthy child is about 1110 mg min m^2.

The normal GFB is not completely impermeable to albumin, and the total amount of albumin filtered by the glomeruli of a healthy child is about 0.7 mg min m^2, i.e. about 1000 mg day m^2.

The total protein excreted in the urine of a healthy child is less than 100 mg day m^2. The kidneys of a healthy child thus reabsorb nearly all the filtered albumin; the main sites of reabsorption are the proximal convoluted tubule (71%), the loop of Henle and the distal tubule (23%) and the collecting duct (3%).

Mechanisms of proteinuria

There are three main processes that can cause proteinuria:

1. Overflow proteinuria:
    - Excretion of acute phase reactants or cytokines:
        - Sepsis
        - Trauma
    - Myoglobinuria
    - Haemoglobinuria
    - Immunoglobulin fragments.
2. Glomerular proteinuria:
    - Increased permeability of the GFB to proteins and large molecules.
3. Renal tubular disease (tubular proteinuria).

Urinary electrophoresis distinguishes between overflow proteinuria, glomerular proteinuria and tubular proteinuria. Urinary electrophoresis that shows a predominance of albumin in comparison to molecules of intermediate molecular weight in the NS (i.e. in glomerular proteinuria) is called a ‘selective

proteinuria'. An increased proportion of molecules of intermediate molecular weight relative to albumin in the urine is called a 'non-selective' proteinuria.

Selective proteinuria is more common in minimal change (glomerular) disease, and non-selective proteinuria is more common in other types of glomerular damage that produce the NS.

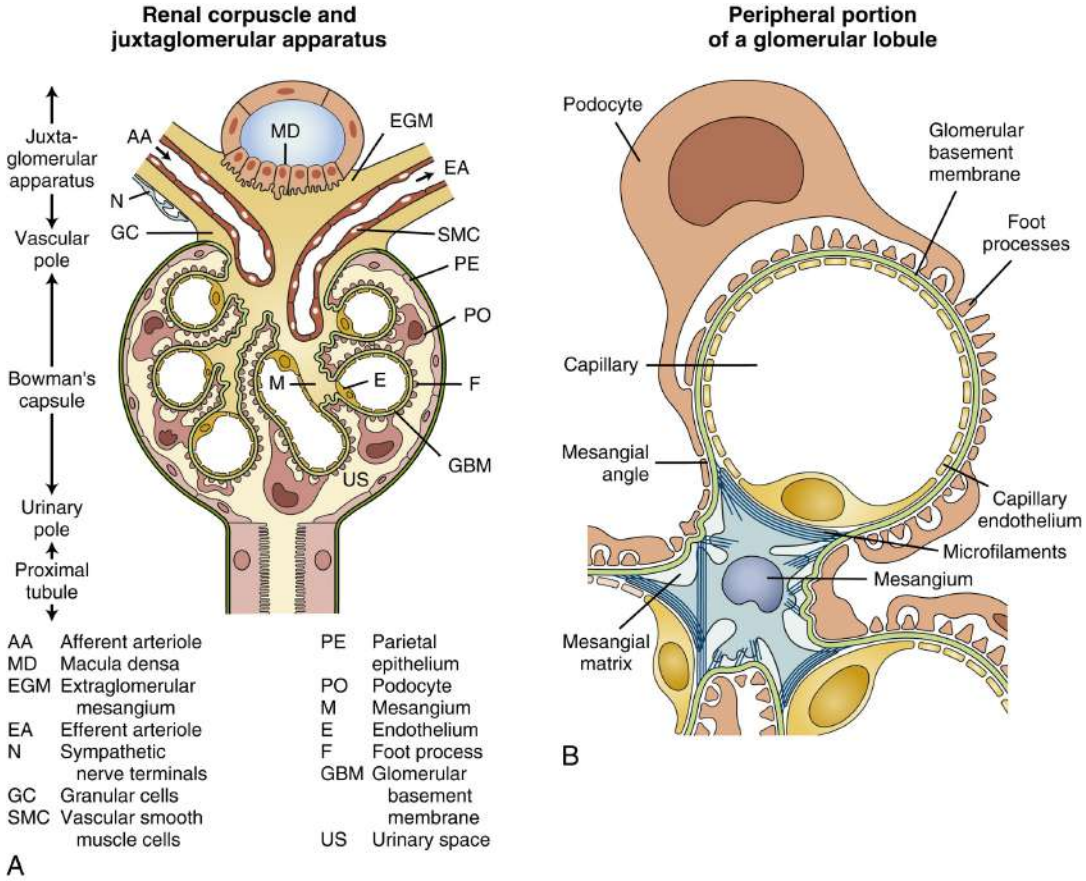

FIG. 16.6.1 A, Diagram of the structure of the glomerulus. B, Diagram of the structures forming part of the glomerulus and the glomerular filtration barrier. Modified from Feehally J, Floege J, Johnson RJ. *Comprehensive Nephrology,* 3rd ed. London: Mosby Elsevier; 2007.

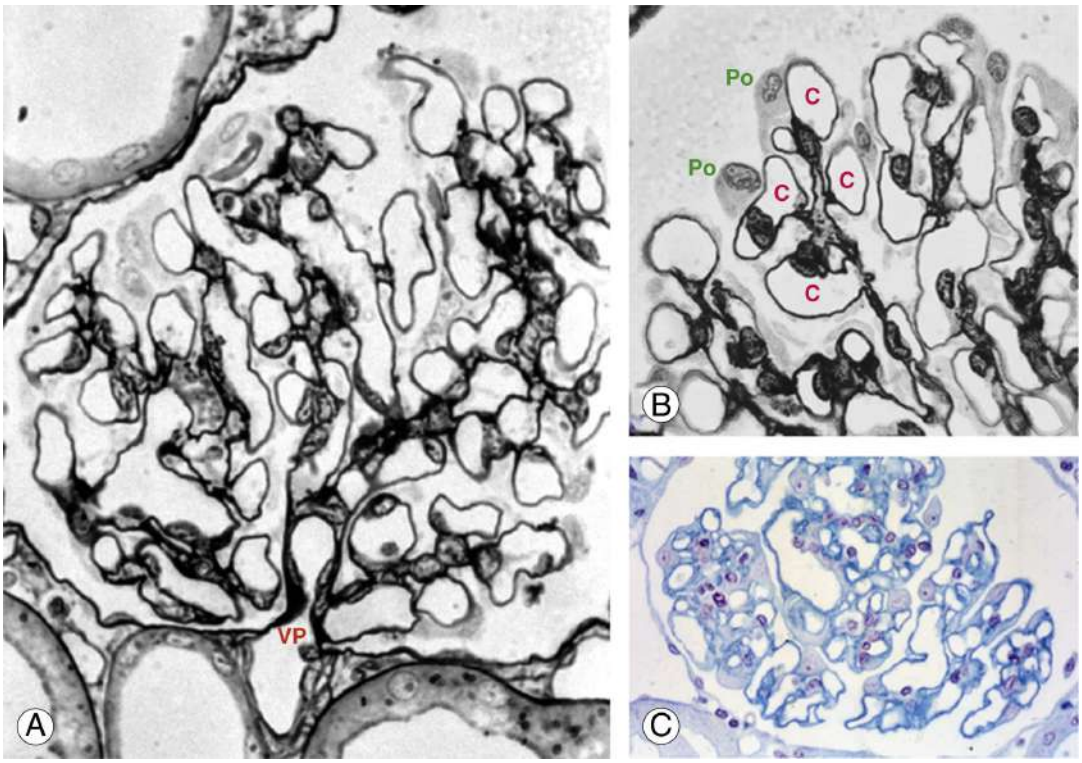

FIG. 16.6.2 Light microscopy of a normal glomerulus. A, Section passing through the vascular pole (VP), with the dominant feature being the open glomerular capillaries of the glomerular tuft (Silver stain). B, High-power view of part of a normal glomerulus, showing podocytes (Po) and glomerular capillaries (C) arranged round the mesangium (Silver stain). C, Special stain of a normal glomerulus that shows the negatively charged ions in the glomerular filtration barrier as a thin blue line (Colloidal iron stain).

Definition of nephrotic syndrome

Peripheral oedema AND marked proteinuria* AND hypoalbuminaemia** AND hyperlipidaemia

Accurate timed urine collections are difficult to obtain in children, and the protein/creatinine ratio (Pr/Cr) on an untimed urine specimen is an accepted alternative:

- Children aged 6 months to 2 years: normal Pr/Cr is <50 mg mmol
- Children over 2 years of age: normal Pr/Cr is <20–25 mg mmol

- Pr/Cr >300 mg mmol is consistent with NS.

A 12-hour or 24-hour timed urine sample with a protein excretion of more than 50 mg kg per day or 40 mg m^2 per hour indicates NS.

Classification of paediatric nephrotic syndrome

According to cause

NS in children is classed into three categories:

1. *Congenital NS* occurs within the first 3 months of life. Some cases are caused by intrauterine infections such as syphilis or toxoplasmosis. Most cases have abnormalities of genes/proteins, often affecting the podocyte. Similar abnormalities have been found in some cases of nephrotic syndrome that develop in infancy/childhood or occur in juveniles/adults.

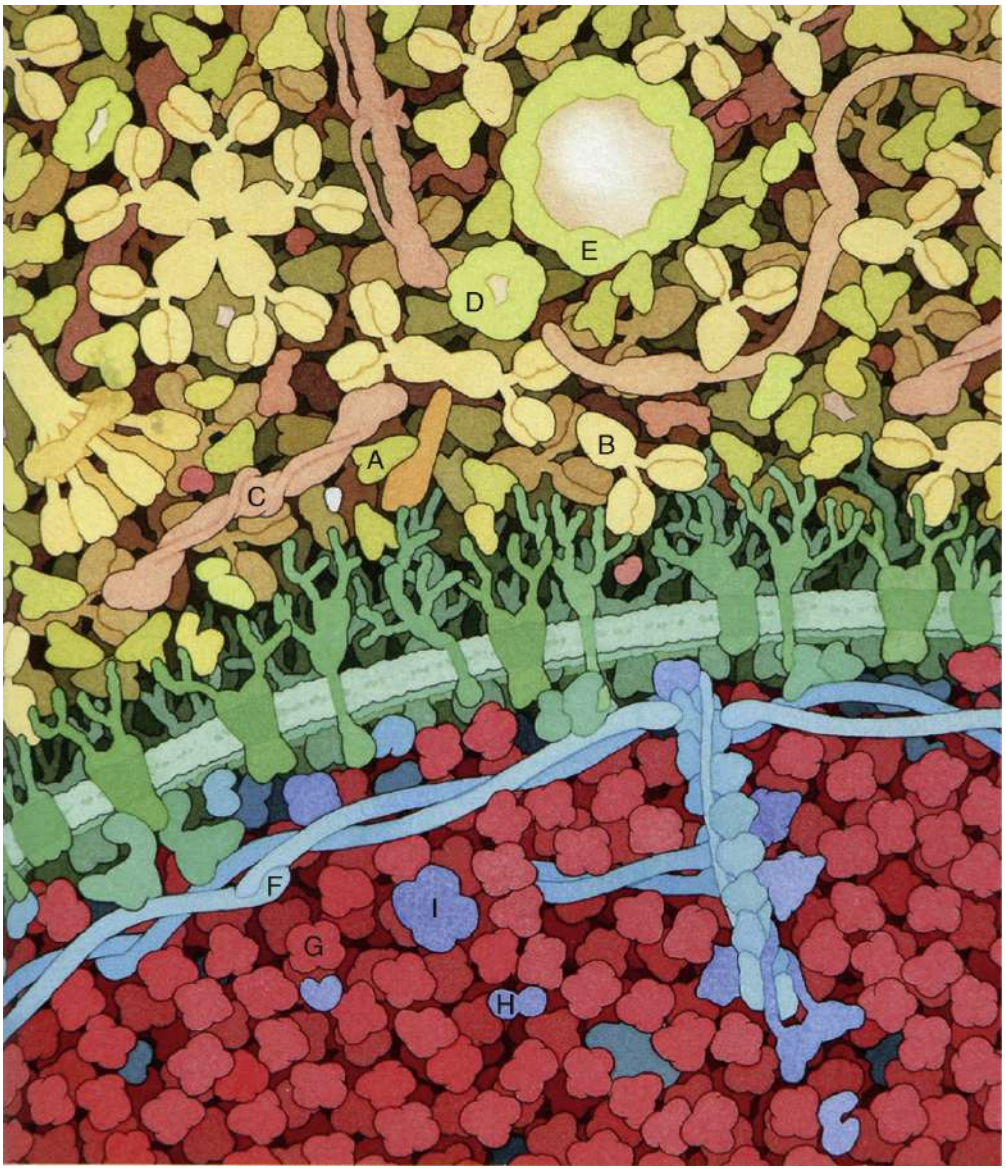

FIG. 16.6.3 The nanoscale structure of the molecules in the blood plasma and a red blood cell. The molecules are drawn to scale showing their size and shape and distribution. Source: Goodsell DS. The *Machinery of Life*. 2nd ed. New York: Springer, 2009.

2. *Secondary NS* is associated with an identifiable systemic disease or infection and is often accompanied by a nephritic component.
3. Primary NS (also termed *idiopathic NS [INS]*) is due to a disease or condition that begins in the kidney and is not related to systemic disease.

Nearly all (95%) cases of NS in children are due to a primary glomerular disease, i.e. INS. Secondary causes are seen in less than 5% of children with NS (Fig. 16.6.4).

According to renal histology

The following nomenclature describes the histological changes that may occur in the kidney in the INS:

1. Focal or diffuse: Proportion of glomeruli affected by a process:
    - Diffuse: more than 50% of all the glomeruli are involved
    - Focal: less than 50% of all the glomeruli are involved.
2. Global or segmental:
    - Global means that the entire glomerulus is affected
    - Segmental means that only part of a glomerulus is involved.

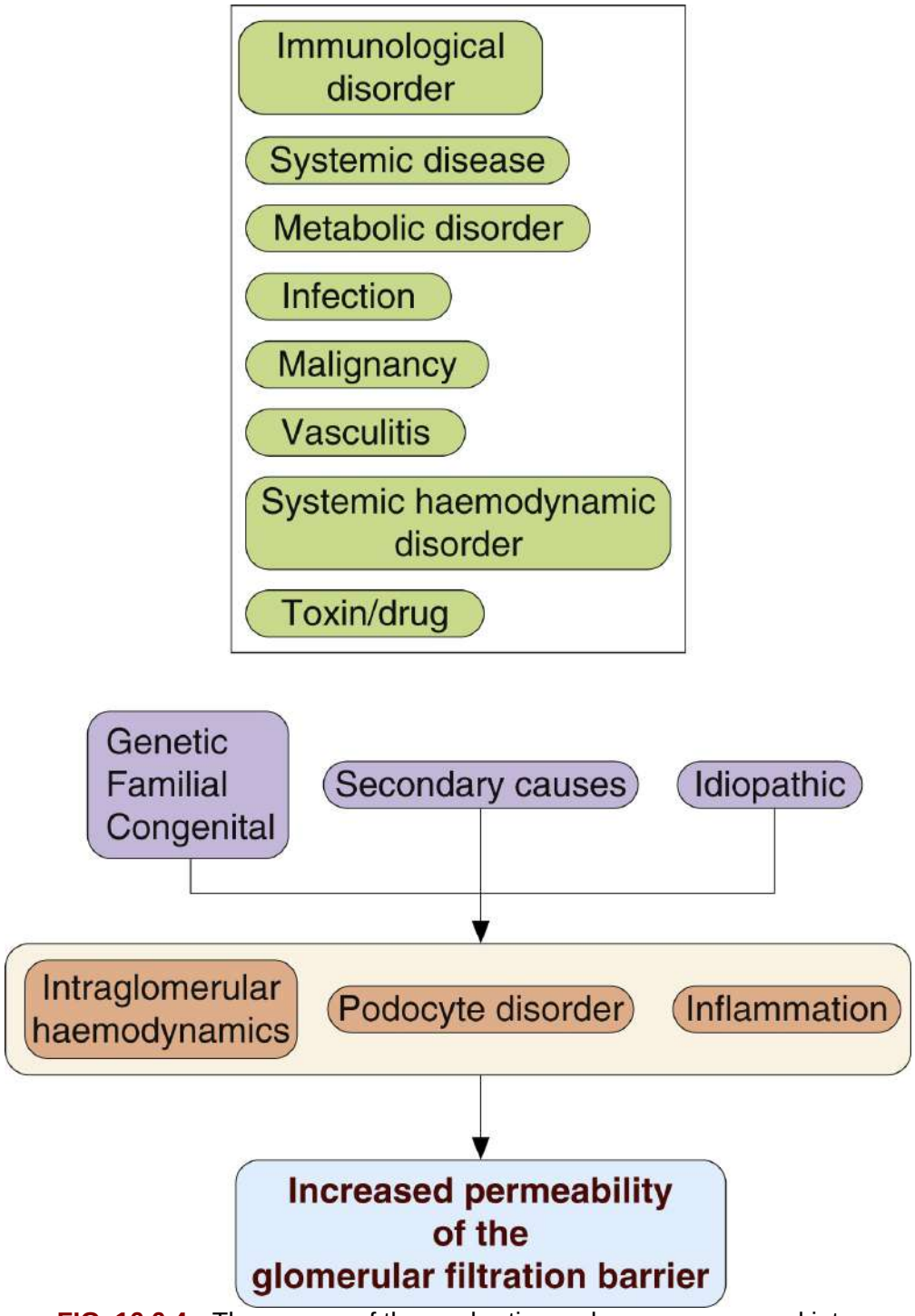

FIG. 16.6.4 The causes of the nephrotic syndrome are grouped into three main groups that will eventually have the common effect of increasing the permeability of the glomerular filtration barrier.

3. Hypercellularity:
    - Endocapillary hypercellularity means an increase in cells inside the boundaries of the glomerular capillary wall

- Extracapillary hypercellularity means an increase in cells outside the boundaries of the glomerular capillary wall.

4. Sclerosis: increase in extracellular matrix or collagen within the mesangial space, glomerular tuft or interstitial compartment.
5. Hyalinosis is an acellular, glassy and homogeneous material that contains protein (and has different staining characteristics from sclerotic lesions).
6. Deposits: protein or immune deposits within the glomerulus (Fig. 16.6.5).

The main types of primary glomerular changes that are seen in INS are:

Minimal change disease (MCD)	76–77%
Focal segmental glomerulosclerosis (FSGS)	5–7.5%
Mesangial proliferative glomerulonephritis	2.3–5.5%
Membranoproliferative glomerulonephritis	4–7.5%
Membranous nephropathy	1.5%
Others	2.5–7.5%.

In this chapter INS due to MCD and FSGS is discussed.

According to response to corticosteroids

Patients with INS can be classified according to their response to empirical glucocorticoid therapy into those with:

- steroid sensitive NS (SSNS). Ninety-five per cent of children who eventually respond to steroids do so within the first 4 weeks of treatment
- steroid dependent NS (SDNS)
- steroid resistant NS (SRNS).

Children with SSNS can be further subdivided into those who are in remission and those whose nephrotic syndrome returns or relapses – the relapses can be infrequent or frequent (Fig. 16.6.6).

About 80–90% of cases of INS respond to initial steroid therapy, i.e. they are SSNS. Children with SSNS have an 80% chance of having one or more relapses, and approximately 50% of these children will have frequent relapses.

Idiopathic nephrotic syndrome: epidemiology

The annual incidence of INS in children is about 2–7 cases per 100,000 children, and the cumulative prevalence is nearly 16 cases per 100,000. The ratio of males to females is about two to one during childhood, but the sex ratio is the same by adolescence. There is an increased familial incidence, particularly among siblings. The usual age at the onset of symptoms in patients with MCD is between 2 and 6 years; 30% of the adolescents also show MCD. FSGS may occur throughout childhood, though the median age of onset of symptoms is 6 years.

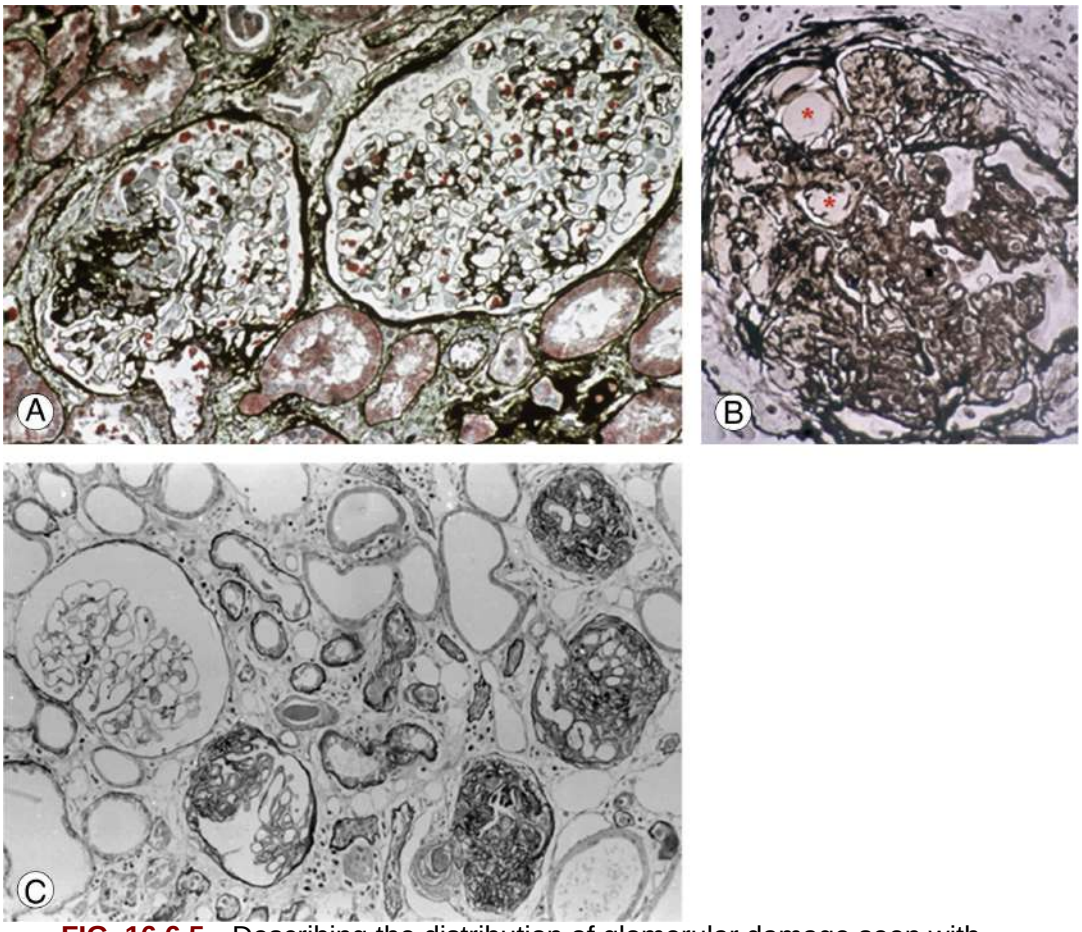

FIG. 16.6.5 Describing the distribution of glomerular damage seen with the light microscope. A, The glomerulus on the right has a normal appearance under the light microscope, while the glomerulus on the left has an area of sclerosis (irregular black stain that involves about one-quarter of the visible glomerular tuft). This histological pattern is described as a focal and segmental glomerular sclerosis (PASM stain). B, The whole glomerular tuft is sclerosed, so this is described as global sclerosis in a

single glomerulus. Hyalinosis deposits (∗) are also present (Silver stain). C, There is extensive interstitial fibrosis and tubular atrophy. Five glomeruli are seen, with the one on the left having a normal light microscopy appearance and the three glomeruli on the right showing global sclerosis. The glomerulus that is second from the left is partially sclerosed. This histological pattern is described as a diffuse and global glomerular sclerosis (PAS stain).

Minimal change disease

Terminology of minimal change disease

MCD was first called lipoid nephrosis because of the finding of lipids in the renal tubular cells as well as lipid-laden proximal tubular cells or macrophages known as oval fat bodies in the urine. The term *nephrosis* also implied that oedema is a marked clinical feature and that there is little or no haematuria. This contrasted with the term *nephritic syndrome* where oedema was less marked, there was marked haematuria and the renal function was often impaired. Another term previously used for MCD was *nil disease* (because of the normal light microscopy findings).

Pathogenesis of minimal change disease

An intriguing aspect of MCD is the combination of a markedly increased permeability of the GBM to protein and the near normal appearance of glomeruli on light microscopy, compared to other types of glomerular diseases where there can be extensive glomerular damage but much smaller amounts of protein leakage into the urine.

The exact cause of MCD is unknown, but the following factors have been linked to the development of the condition:

- Circulating permeability factors
- T-cell dysfunction
- The clinical association between MCD and chronic lymphoid neoplasms support a role for B-cell dysfunction
- Podocytopathy: visceral epithelial cells (podocytes) were previously thought to have a passive role in the process of glomerular filtration. Podocytes have now been shown to affect the structure and function of the GBM and to regulate the integrity and survival of glomerular

endothelial cells. Mutation of a protein located at the slit diaphragm (nephrin) causes the Finnish type of congenital nephrotic syndrome. This suggests that the final common pathway in the increased permeability of the GFB is MCD is a defect of the podocyte or slit barrier or both.

Pathological changes in minimal change disease (Fig. 16.6.7)

The pathological changes in MCD are:

- normal glomeruli on light microscopy ± slight mesangial hypercellularity
- immunofluorescence studies of the glomeruli are negative
- glomerular podocytes may contain protein drops
- proximal tubular epithelial cells may contain large numbers of protein drops
- the cells of the convoluted tubules contain large numbers of lipid droplets
- minimal tubule-interstitial changes (apart from fat uptake by tubular cells)
- electron microscopy findings are:
    - normal appearance of the glomerular basement membrane
    - 'fusion' of the foot processes – this is probably not a true fusion but a spreading of individual foot processes
    - 'fusion' is widespread (but not universal) and is reversible.

Primary (idiopathic) focal segmental glomerulosclerosis

Terminology of focal segmental glomerulosclerosis

FSGS describes a histological change that involves less than half the total number of glomeruli and involves less than half the volume/size of the involved glomeruli. This pattern of glomerular scarring can occur in any type of chronic kidney disease (secondary FSGS). A similar histological appearance can be seen in the INS and results from podocyte injury (primary FSGS).

Pathogenesis of primary focal segmental

glomerulosclerosis

Circulating factor(s), mutations in key podocyte genes or epigenetic factors are involved in the development of some cases of primary FSGS. Persistent proteinuria can activate inflammatory and/or pro-fibrotic pathways in tubular cells and might also injure podocytes. Progressive focal damage to glomeruli can cause compensatory changes in the intraglomerular haemodynamics of unaffected glomeruli that can damage these glomeruli.

Pathological changes in focal segmental glomerulosclerosis

The earliest glomeruli that are affected in primary FSGS are those located near the medulla (juxtamedullary glomeruli).

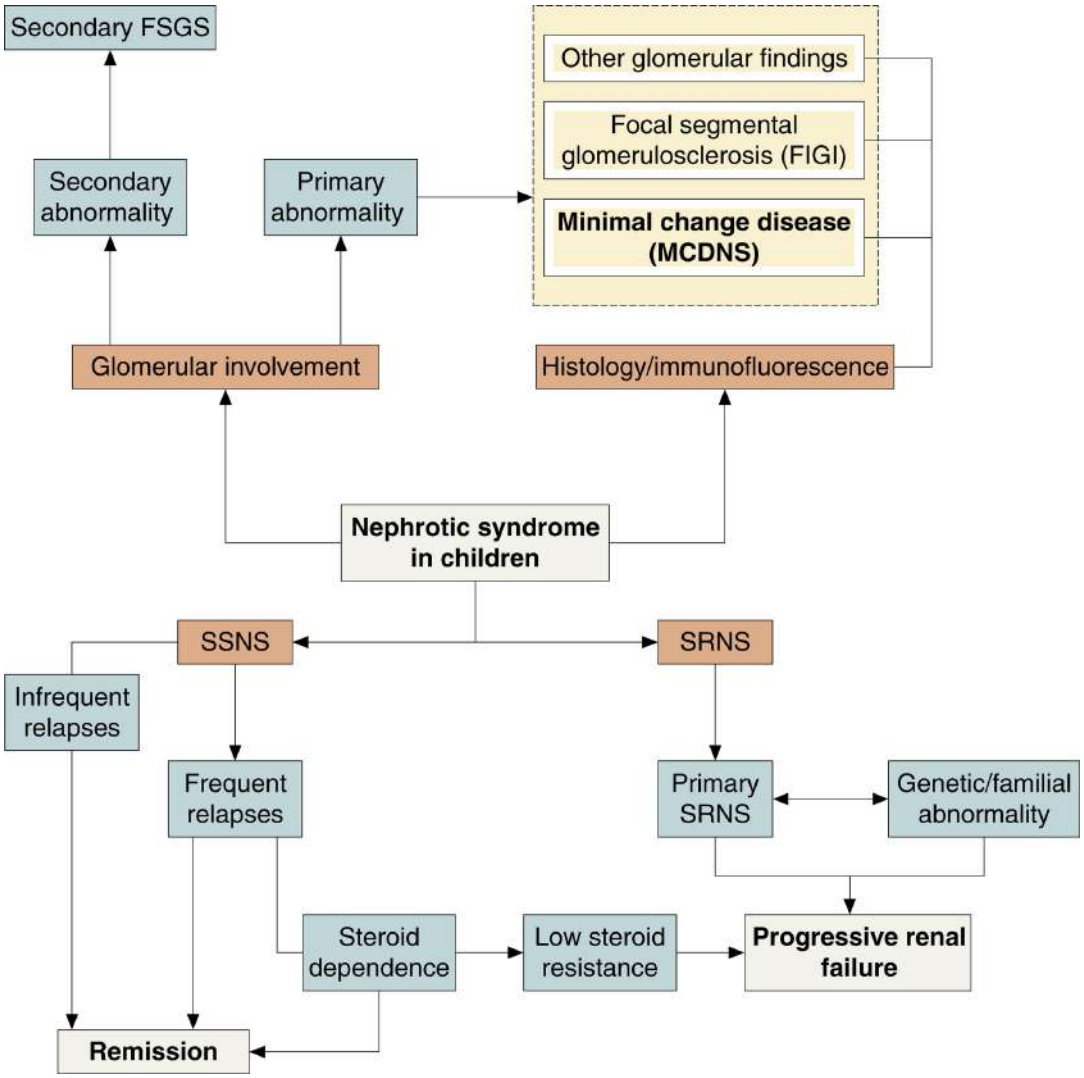

FIG. 16.6.6 Classification of nephrotic syndrome based on a combination of the initial therapeutic response to corticosteroids and the underlying histopathology.

FSGS lesions have varying morphological appearances. The light microscopy changes in a common type (FSGS – NOS [not otherwise specified]) are:

- segmental solidification of the glomerular tuft commonly affecting the vascular pole or the periphery of the glomerular tuft.
- the sclerosis process can involve or cause:
    - collapse of part of the glomerular tuft
    - adhesion (synechia) of the glomerular tuft to Bowman's capsule
- mesangial cell proliferation ± increase in mesangial matrix (Fig. 16.6.8).

Immunofluorescence usually shows focal and segmental glomerular deposition of granular IgM and C3, both within the sclerosed segment and within the mesangium.

Interstitial fibrosis and tubular atrophy are common findings. Tubules contain protein resorption droplets and lipid droplets (Fig. 16.6.9).

Clinical features of idiopathic nephrotic syndrome

There is often no precipitating cause, but sometimes a upper respiratory infection precedes the development of the NS or relapse of the disease. Other preceding events include allergic reactions to bee stings and the use of certain drugs such as non-steroidal anti-inflammatory drugs (NSAIDs). A history of allergy is present in a third of children with MCD.

Oedema

Oedema is the presenting symptom in 95% of children with INS.

The pathogenesis of the oedema is poorly understood:

- Hypoalbuminaemia is an unlikely cause as most nephrotic patients have a decreased concentration of albumin in the interstitial fluid and an increase in interstitial fluid pressure.
- The ‘under-fill’ hypothesis states that the decreased plasma oncotic pressure causes hypovolaemia and secondary renal sodium retention.
- The ‘overfill’ hypothesis states that the nephrotic syndrome causes primary renal sodium retention, leading to oedema. The intravascular volume should be normal or increased in this setting.
- The under-fill and overfill mechanisms may be present at different stages of the INS in the same person (Fig. 16.6.10).

Oedema appears initially in areas of low tissue resistance and is characteristically dependent. In the morning it is periorbital and may be misinterpreted as being due to an allergy. Over the course of a day, the oedema often distributes from the eyes to more dependent areas such as the scrotal and labial regions and the lower limbs. In severe cases oedema becomes generalised with the development of ascites or pleural effusions or both, as well as massive

oedema of the legs and face. The development of moderate or severe oedema is associated with weight gain.

Other features

Some patients have both interstitial fluid volume expansion (causing oedema) and a reduced intravascular volume. Other patients with INS have a normal (or increased) intravascular volume. The reduced intravascular volume can cause significant hypovolaemia which contributes to the acute kidney injury seen in some MCD cases.

All patients with INS should be assessed for possible intravascular depletion:

- The features of intravascular volume depletion are dizziness, abdominal cramps, peripheral hypo-perfusion (cold hands or feet, mottling, capillary refill time greater than 2 seconds), tachycardia, reduced urine output, low urinary sodium, hypotension (a late sign) (Fig. 16.6.11).

Anorexia, irritability, fatigue, abdominal discomfort and diarrhoea are common. Gastrointestinal distress can be caused by ascites, bowel wall oedema or both. Respiratory distress can occur due to massive ascites, frank pulmonary oedema, or pleural effusions or pulmonary embolism.

Patients have an increased susceptibility to cellulitis and peritonitis and may develop fever or severe abdominal pain.

The blood pressure is usually normal; an elevated blood pressure on presentation can be seen in a small proportion of patients with MCD but is more common in FSGS or in cases with a mixed nephrotic/nephritic presentation. Hypotension can be due to sepsis or hypovolaemia or both.

Investigations

Urine analysis

This will show marked proteinuria: 3+/4+ proteinuria. Microscopic haematuria is present during the first few weeks of illness in up to one-third of children with MCD. Macroscopic haematuria develops in 3% to 4% of MCD cases. Renal vein thrombosis must be considered in patients with significant haematuria.

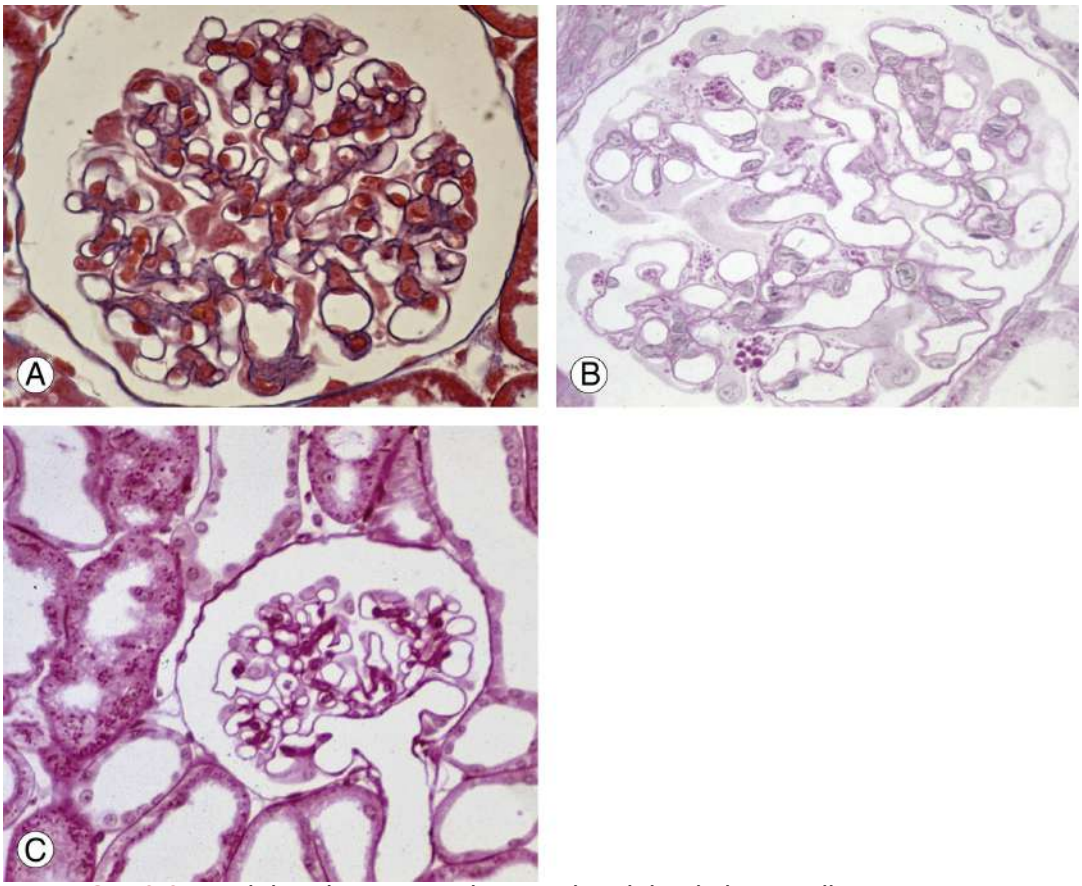

FIG. 16.6.7 Light microscopy changes in minimal change disease. A, Normal glomerulus apart from slight mesangial hypercellularity. B, Albumin-containing drops (red staining) are seen within the podocytes of otherwise normal glomerulus (PAS stain). C, Albumin reabsorption drops (red staining) are seen in the cytoplasm of tubular cells that are to the left of a glomerulus with a normal light microscopy appearance (PAS stain).

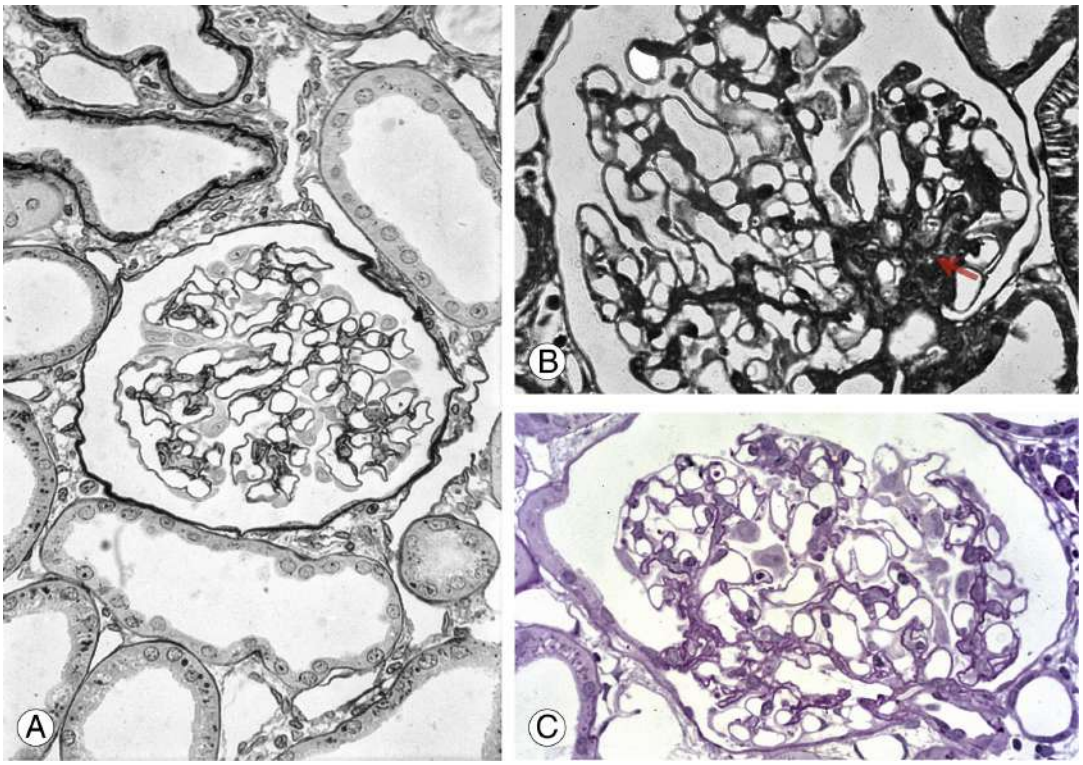

FIG. 16.6.8 Light microscopy changes in focal and segmental glomerular sclerosis (FSGS). A, Normal glomerulus, moderate interstitial change, protein reabsorption drops in tubular cells (left lower quadrant) (Silver stain). B, Segmental area of increased mesangial matrix (red arrow) near the vascular pole of a glomerulus (PAS stain). C, Increased mesangial sclerosis and an adhesion between the glomerular tuft and Bowman's capsule near the start of the proximal tubule (PAS stain).

Other investigations are urine microscopy, urine culture, urinary protein/creatinine ratio and spot urinary sodium concentration in suspected hypovolaemia. A spot urinary sodium concentration of <10 mmol L suggests hypovolaemia.

Blood tests

Blood: Full blood examination; electrolytes, urea and creatinine (Cr); liver function tests (including serum albumin albumin); immunology (serum complement levels are normal in MCD); varicella serology; clotting

Serum sodium may be decreased due to hyperlipidaemia or to water retention. Haematocrit may be increased if water depletion is present. Hypocalcaemia is caused by a low serum albumin level, but the ionized calcium concentration is normal.

Imaging

The basics are chest X-ray and abdominal ultrasound.

Differential diagnosis

INS is part of the differential diagnosis for any child presenting with new onset oedema. Features suggesting a renal condition other than MCD are:

- age of onset less than 1 year
- positive family history of nephrotic syndrome
- extra-renal disease (e.g. arthritis, rash, anaemia)
- renal failure
- nephritic urine sediment (red blood cell casts).

Complications of idiopathic nephrotic syndrome

Hypovolaemia

Some patients exhibit signs of acute renal failure (i.e. reduction in glomerular filtration rate and oliguria). These signs are usually reversed when the intravascular volume is expanded by infusion of salt-poor human albumin.

Infection

Oedema predisposes to the development of cellulitis. Patients are also at risk of other bacterial infections, the most common of which is spontaneous bacterial peritonitis (seen in 2% to 6% of patients with INS). Peritonitis used to be due mainly to *Streptococcus pneumoniae*. Vaccination has reduced infections with *S. pneumoniae*, and there has been an increase in the relative frequency of gram-negative organisms. There also has been an increase in penicillin-resistant *S. pneumoniae*.

A high degree of suspicion is needed when treating a nephrotic patient with ascites. Features suggestive of peritonitis are fever, chills, abdominal pain, ileus or diarrhoea. Peritoneal fluid must be obtained and analysed in all patients with possible peritonitis, and these patients should initially be given a combination of an aminoglycoside and ampicillin until the results of ascitic fluid analysis are back.

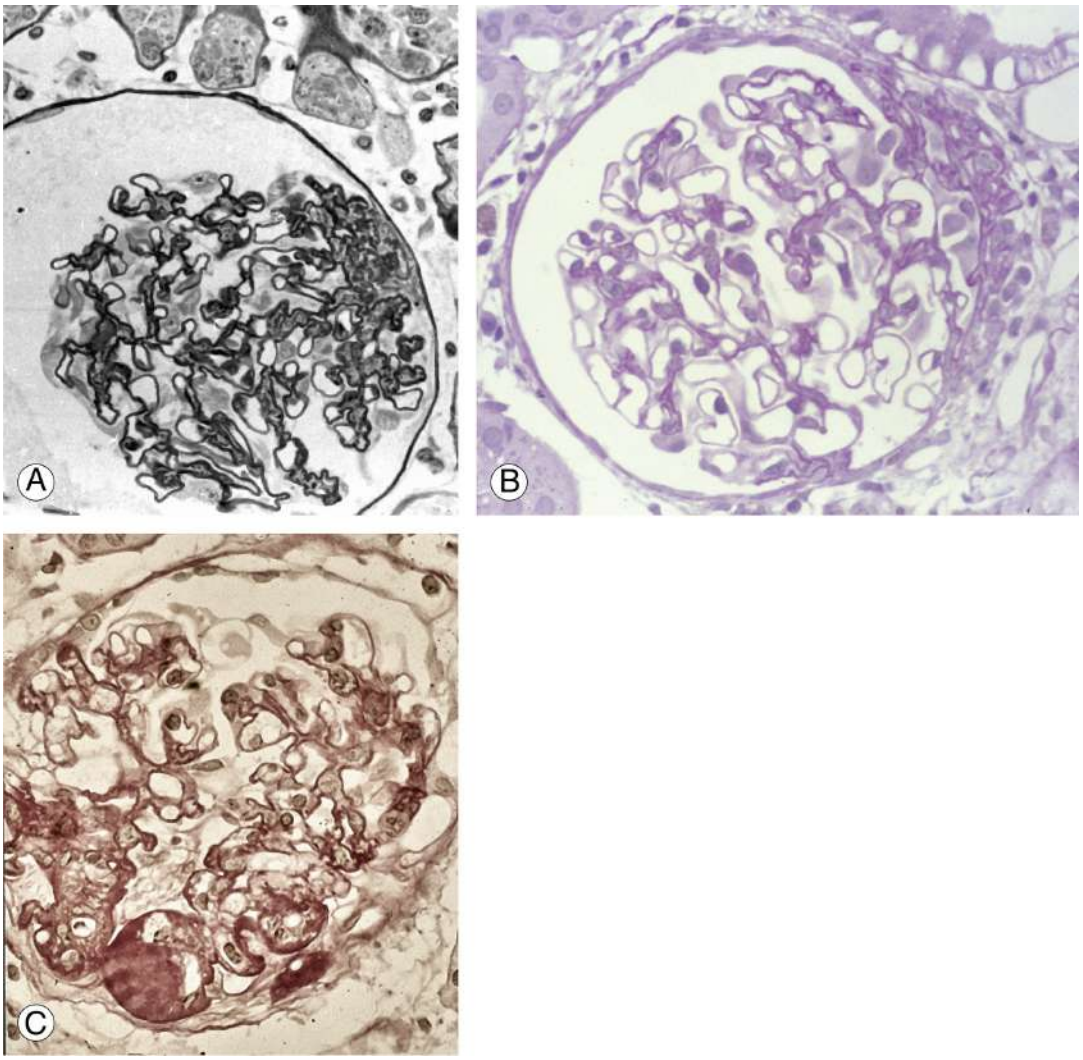

FIG. 16.6.9 Patterns of light microscopy changes in affected glomeruli in focal and segmental glomerular sclerosis (FSGS). A and B, Adhesion between the glomerular tuft and Bowman's capsule, with sclerosis and collapse of the glomerular capillaries at the adhesion. Bowman's capsule has been partially destroyed at the adhesion site in B. C, Mesangial and endocapillary swelling, with a focal area of sclerosis and hyalinosis (PAS stain).

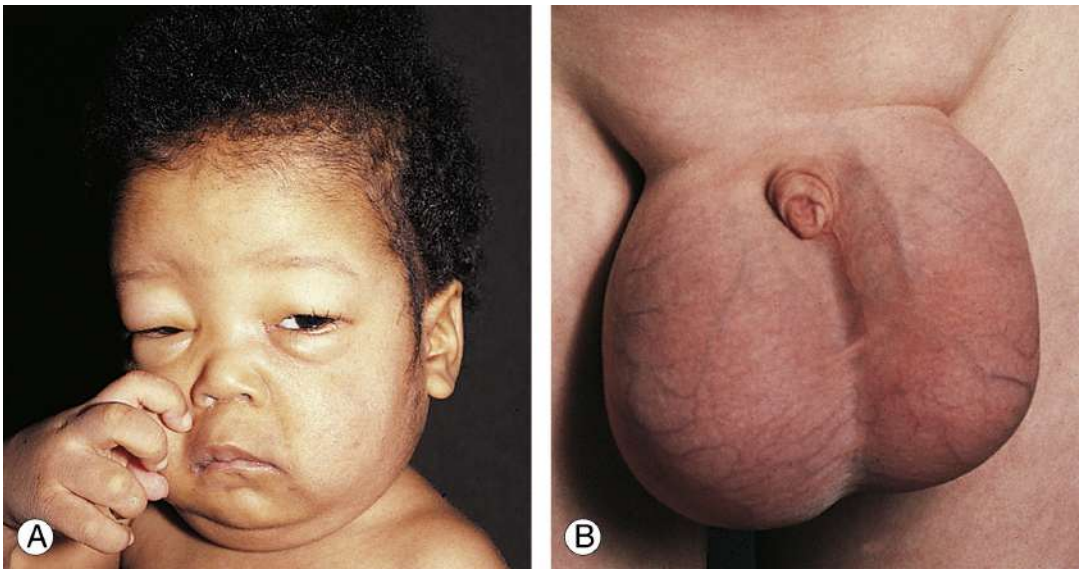

FIG. 16.6.10 Clinical features of nephrotic syndrome. A, Marked eyelid oedema in a 2-year-old boy. B, Marked scrotal oedema in a 6-year-old boy. Modified from Basil J Zitelli and Holly W Davis, editors. *Atlas of Pediatric Physical Diagnosis*. 2nd ed. New York: Mosby-Wolfe; 1992.

Thrombosis

Asymptomatic venous thrombosis may develop in up to one-quarter of children with INS. About 2% to 8% of children with INS will have a clinically apparent thrombotic complication. Thrombotic episodes are predominantly venous, but arterial thrombosis can also occur. Thrombosis of the deep leg veins is the most common site. Renal vein thrombosis should be considered if there is an unexplained deterioration in renal function or there is loin pain with or without marked haematuria. Rarely thrombosis of the sagittal sinus vein can occur.

The increased thrombotic risk is the result of multiple factors:

- Physical factors (immobility, haemoconcentration, increased blood viscosity)
- Loss of antithrombin III and protein S
- High fibrinogen level
- Elevated levels of factors V, VII, VIII, X
- Thrombocytosis
- Increased platelet aggregation
- Abnormal endothelial function (possibly related to dyslipidaemia).

Venous thrombosis is treated with heparin, followed by warfarin. Many heparin compounds are cleared renally; thus, dose adjustment may be necessary if there is renal dysfunction. Warfarin is hepatically cleared so that additional renal adjustments should not be needed. If the thrombosis is extensive thrombolytic drugs may be needed. The total recommended course of anticoagulation for a first venous thromboembolism is at least 3 months to 6 months or until the nephrotic syndrome is in remission.

Long term due to the nephrotic syndrome or treatment or both

- Muscle wasting
- Bone disease: in addition to the effect of corticosteroids on bone, urinary loss of vitamin-D-binding protein may cause vitamin D deficiency and, less commonly, secondary hyperparathyroidism
- Psychological effects
- The hyperlipidaemia in adults with the nephrotic syndrome is associated with an increased risk of coronary heart disease. The likelihood of this occurring in children with INS is not known. The chronic hyperlipidaemia in adults with renal disease may worsen the existing renal damage, but the relevance of this in children with the INS is not known.

Treatment of initial nephrotic phase

General measures

Dietary measures include salt restriction and increased protein intake. NSAIDs should be avoided.

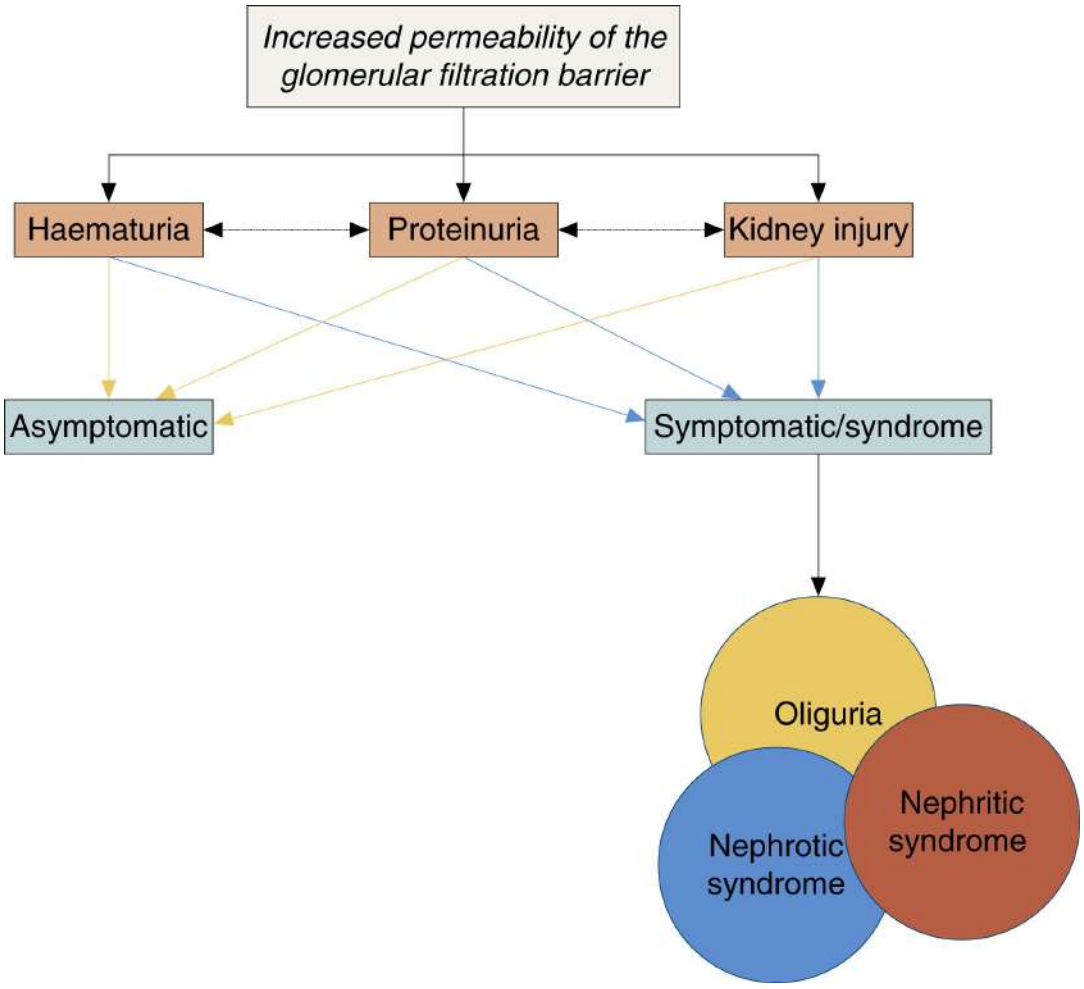

FIG. 16.6.11 Increased permeability of the glomerular filtration barrier can produce asymptomatic abnormalities or symptomatic presentations that cause the nephrotic syndrome or the nephritic syndrome or reduced kidney function with oliguria.

Immunisation

The patient's varicella status should be documented, and children who are not immune should receive varicella vaccine when the nephrotic syndrome is in remission. Pneumococcal vaccination is recommended for patients who have nephrotic syndrome. Zoster immune globulin should be given to non-immune children exposed to chickenpox while on steroids.

Corticosteroid medication

All patients with INS should be treated with corticosteroids. Longer initial

courses of prednisolone are associated with a lower incidence of relapse, and therefore the initial duration of treatment is 24 weeks. The dose of prednisolone is based on surface area:

60 mg m^2 per day as a single dose (max 60 mg day) for 4 weeks.

Then:

40 mg m^2 per alternate day (max 40 mg) for 4 weeks
20 mg m^2 per alternate day for 4 weeks
15 mg m^2 per alternate day for 4 weeks
10 mg m^2 per alternate day for 4 weeks
5 mg m^2 per alternate day for 4 weeks.

Prednisolone can be given as a single dose in the morning with food or as divided doses during the day. If prednisolone causes gastric irritation a proton-pump inhibitor can be given.

Most children with nephrotic syndrome will respond to steroid treatment within 2–4 weeks. A remission is defined as 3 or more days of trace or negative on dipstick testing. If proteinuria persists beyond the first 4 weeks of steroid treatment, then children should be referred for review by a nephrologist for possible renal biopsy.

The urine of children with steroid-responsive INS should be checked initially twice weekly, then weekly after the first episode.

Between 60% and 70% of children with INS may have one or more relapse. These are diagnosed if there is 3+ or 4+ proteinuria for 3 or more days.

Oedema

General measures include reduced salt intake, daily weights, daily urine dipstick and fluid balance measurement. Diuretic drug treatment is required when marked peripheral oedema, severe ascites or pleural effusions are present. This involves oral treatment with loop diuretics (e.g. frusemide) and thiazide diuretics (e.g. hydrochlorothiazide).

Patients with tense ascites with abdominal compartment syndrome limiting diaphragmatic excursion, severe pleural effusions or severe scrotal or labial oedema, risking skin breakdown, may require treatment with intravenous 20%

albumin (with intravenous frusemide). There is a risk of hypertension or pulmonary oedema if the infusion rate is too rapid or too much albumin is given.

A low serum albumin alone is not an indication for intravenous albumin.

Proteinuria

ACE inhibitors and angiotensin II receptor blockers can reduce protein excretion in children with proteinuric renal disorders. They may also decrease damage to podocytes and slow the progression of renal disease. The benefit of using these drugs routinely, given alone or in combination, in normotensive children with INS has not been established.

Infection

The placement of intravenous lines should be minimised. Prophylactic antibiotic (penicillin) is recommended by some centres while oedema persists. Oral penicillin V (phenoxymethyl-penicillin) is given (at a dose of 125 mg every 12 hours in children under 5 years or at a dose of 250 mg every 12 hours if over 5 years of age). There should be early use of empiric antibiotics (IV ceftriaxone 50 mg kg dose every 24 hours [max 2 g]) in a child with INS who has suspected sepsis or is very unwell.

Patient with frequent relapses of nephrotic syndrome or persistent nephrotic syndrome

Management is complex and needs supervision by a nephrologist. It will involve repeated courses of corticosteroids and the use of other immunosuppressive agents and consideration of renal biopsy. Indications for early renal biopsy include macroscopic haematuria, hypertension, persistent renal insufficiency, low C3 complement values or a failure to respond to 4 weeks of daily prednisone therapy.

A significant number of patients with MCD may have at least one relapse during adulthood. The risk of relapse is greater in those with a young age of onset or high number of relapses in childhood. Pregnancy is a predisposing factor for relapse of NS in adulthood.

Steroid-dependent patients have a prolonged course, but the risk of progression to end-stage renal failure is minimal if the patient continues to

respond to steroids.

Ultimately the response of the INS to steroid response is more prognostic than pathological diagnosis.

Most children who have NS and fail to respond to steroids have FSGS.

There is an increased risk of growth failure, osteoporosis, hypertension, cataracts or fertility problems in children on long-term steroids or other immunosuppressive agents. These children need the following monitoring:

1. Monitor BP
2. Monitor growth (including bone age and pubertal stage where appropriate)
3. Monitor weight – dietetic review where appropriate
4. Glycosuria/HbA1c
5. Bone mineral density/calcium supplements
6. Ophthalmology review.

Prognosis

The best prognostic indicator in children who have NS is steroid responsiveness. Ninety-five percent of children who eventually respond to steroids do so within the first 4 weeks of treatment:

- MCD: Frequently relapses can persist into young adulthood. However, there is no long-term renal impairment.
- FSGS**:** One-third achieve complete remission, one-third have persistent proteinuria, and one-third progress to end-stage kidney disease over 5 to 10 years.

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* The prevalence of proteinuria in asymptomatic school-children is low (0.25%), and two-thirds of those with proteinuria do not have renal disease.∗∗Hypoalbuminaemia (albumin concentrations of =25g L [=2.5 g dL]).

16.7

Henoch–Schönlein purpura

Linas Dziukas

Introduction

Vasculitis is an inflammation of vessels which damages or destroys the vessel wall, leading to haemorrhage or ischaemic damage or both. It is classified according to the size of the affected vessel (Box 16.7.1).

Box 16.7.1 Classification of childhood vasculitis

Predominantly large-vessel vasculitis:
- Takayasu arteritis

Predominantly medium-sized-vessel vasculitis:
- Childhood polyarteritis nodosa
- Cutaneous polyarteritis
- Kawasaki disease

Predominantly small-vessel vasculitis

Granulomatous:
- Wegener's granulomatosis
- Churg–Strauss syndrome

Non-granulomatous:
- Microscopic polyangiitis
- Henoch–Schönlein purpura
- Isolated cutaneous leucocytoclastic vasculitis
- Hypocomplementaemic urticarial vasculitis

Other vasculitides:
- Behçet disease
- Vasculitis secondary to infection, malignancy and drugs (including hypersensitivity vasculitis)

- Vasculitis associated with connective tissue disease
- Isolated vasculitis of the central nervous system
- Cogan syndrome

Unclassified

Vasculitis is also classified on the dominant cell mediating the inflammation (e.g. neutrophilic, granulomatous, lymphocytic or eosinophilic). Neutrophils are the main cell in small-vessel vasculitis, accumulating in a perivascular and interstitial pattern. The neutrophils undergo degeneration (leucocytoclasis), i.e. there is extravasation of erythrocytes and fibrinoid necrosis of the vessel. A variable number of eosinophils can also be seen. The vessel changes in small vessel vasculitis are called leucocytoclastic vasculitis (LCV).

Henoch–Schönlein purpura (HSP) is the most common vasculitis seen in childhood, and Kawasaki disease is the second most common cause. The vasculitis in HSP affects the capillaries, smaller venules and arterioles.

Epidemiology

HSP has an annual incidence between 10 and 20 cases per 100,000 children, of whom more than 90% are less than 10 years of age. The incidence is about 70 per 100,000 per annum in the 4 to 6 year age group. HSP is twice as common in males as in females. There is a seasonal peak during the autumn and winter months.

HSP is milder in infants and children younger than 2 years and is more severe and more likely to cause long-term renal disease in adults. With the exception of nephritis, it is generally an acute self-limited illness lasting from several days to several weeks.

Pathogenesis

The defining pathophysiological feature in HSP is a leucocytoclastic vasculitis that is associated with deposition of IgA-containing immune complexes that cause a type III (immune complex) reaction. This has resulted in suggestions that the term *IgA vasculitis (IgAV)* should replace the eponym ‘Henoch–Schönlein purpura’.

The exact aetiology of HSP is unknown, but the following are important in the development of the disease:

1. Genetic factors
2. Environmental factors/trigger factors
3. Formation of an abnormal IgA1 subtype. Normally, IgA has two isotypes (IgA1 and IgA2) that are found in the serum and in mucosal fluids. Persons with HSP have an abnormal IgA1 structure that makes the IgA1 surface more antigenic and able to induce a humoral IgG autoimmune response.
4. An altered mucosal immune system response to antigens causes increased production of the altered IgA1 and leads to the formation of circulating immune complexes (CIC) that contain the abnormal IgA1 or aggregates of IgA1 molecules.
5. The subsequent deposition of the CIC in small vessels causes vasculitis by activation of complement and increased release of cytokines. Vasculitis in the small vessels of the skin or bowel or joints causes the typical clinical features of HSP.
6. Deposition of IgA1 and CIC in the capillaries and mesangium of the glomerulus activates the complement system and causes glomerular inflammation, i.e. HSP nephritis (HSPN) (Fig. 16.7.1). Some patients develop cellular glomerular crescents that are associated with capillary wall destruction (Fig. 16.7.2).

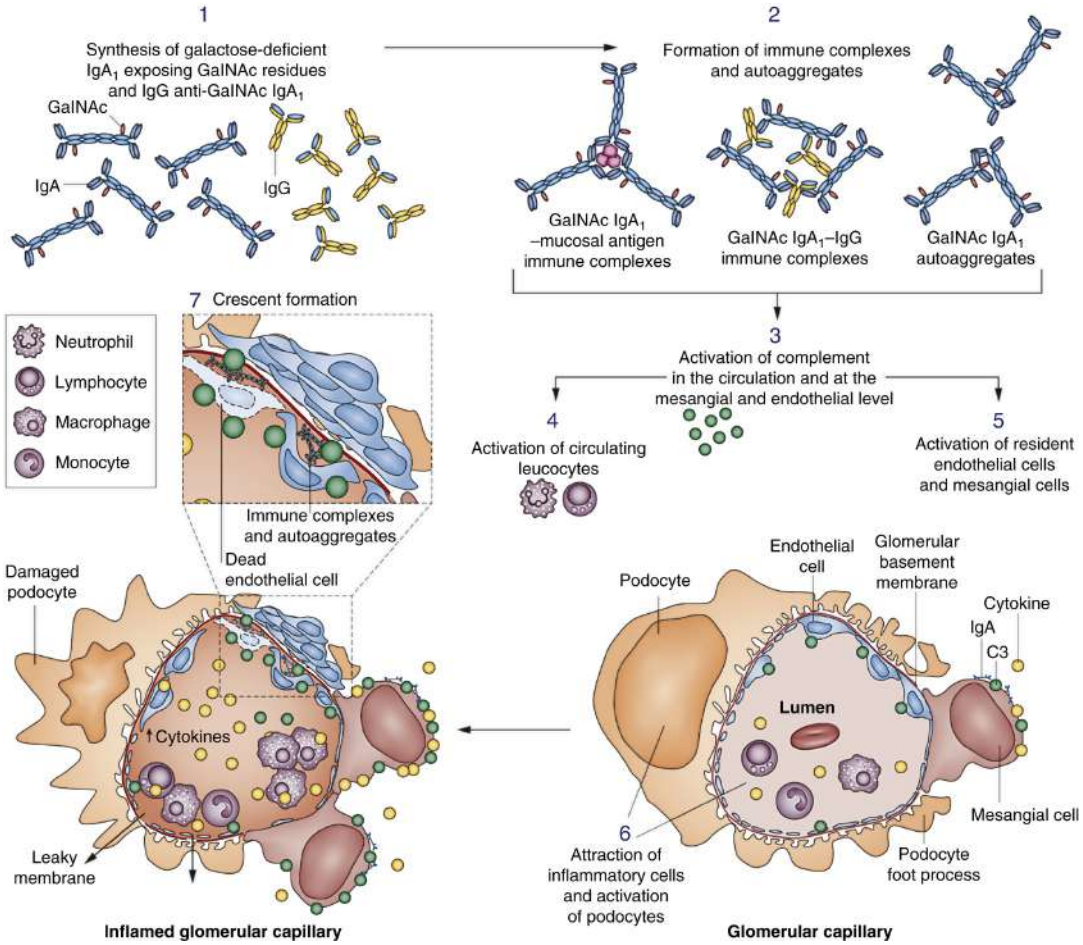

FIG. 16.7.1 Pathogenesis of Henoch–Schönlein purpura nephritis. Modified from Henoch-Schönlein purpura nephritis in children. *Nat Rev Nephrol* 2014;**10**:563–73.

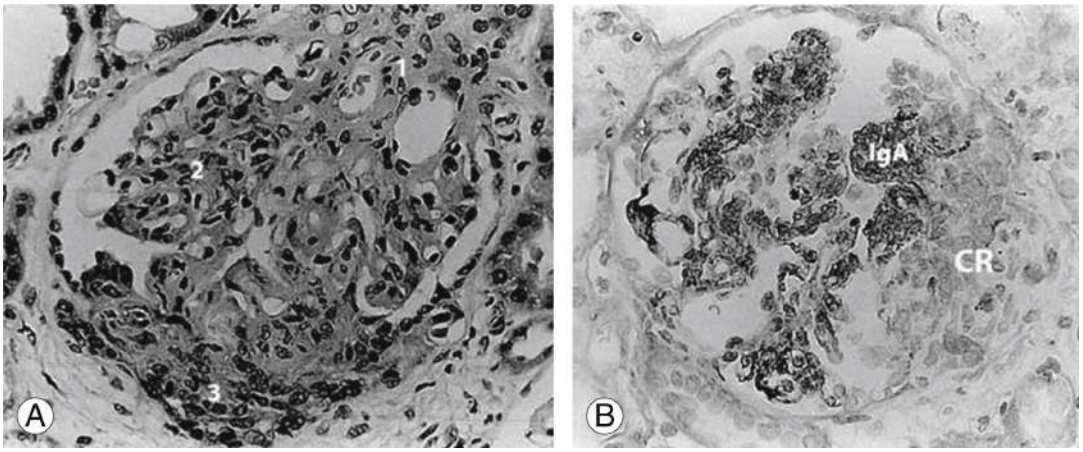

FIG. 16.7.2 Glomerular involvement in Henoch–Schönlein purpura. Modified from A fatal case of bowel and cardiac involvement in Henoch-Schönlein purpura. *Nephrol Dial Transplant* 2002;**17**:497–9.

IgA deposition in the glomerular mesangium occurs in other conditions. It may be secondary to liver disease or inflammatory bowel disease or be associated with other types of glomerulonephritis. It also occurs in primary IgA nephritis (IgAN), where the mesangial deposits also contain abnormal IgA1. Despite this shared feature there are enough differences in histological features and the clinical courses between HSPN and IgAN to regard them as separate conditions.

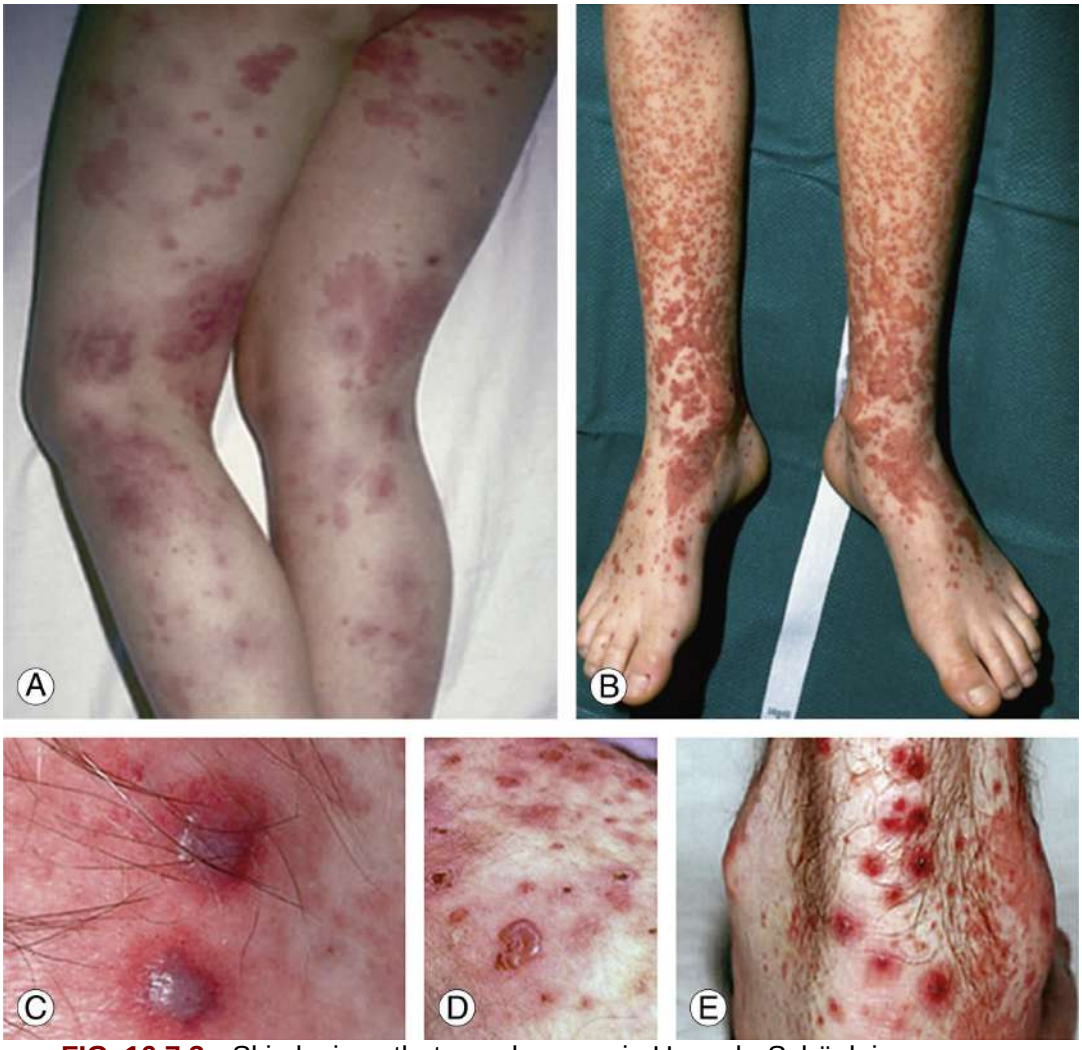

FIG. 16.7.3 Skin lesions that may be seen in Henoch–Schönlein purpura. (A) Urticarial lesions. (B) Typical appearance of palpable purpura, which is most marked on the extensor surface of the legs and forms confluent lesions with irregular edges. (C) Purpuric rash with vesicles. (D) Vesiculobullous lesions. (E) Blistering purpuric rash and swelling over the lateral malleolus due to arthritis.

HSP nephritis is typically an acute disease, and even severe glomerular lesions resolve completely if the causal event is of short duration. When the initial damage is more prolonged, or when adequate treatment is delayed, glomeruli that were not initially affected can be damaged by an increase in their single nephron glomerular filtration rate or an increase in intraglomerular blood pressure. These secondary changes can cause progression to chronic kidney failure.

Diagnostic criteria for Henoch–Schönlein purpura

Palpable purpura in a person with normal platelet count and normal coagulation who has one or more of the following:

1. Diffuse abdominal pain
2. Histopathology: typical LCV with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits
3. Arthritis or arthralgias
4. Renal involvement (proteinuria: >0.3 g 24 h or >30 mg mmol of urine albumin to creatinine ratio on a spot morning sample; and/or haematuria, red blood cell casts: ≥5 red cells per high power field or ≥2+ on dipstick or red blood cell casts in the urinary sediment).

Clinical features

In two-thirds of the cases, the disease follows an upper respiratory tract infection, with onset an average of 10 days after the start of respiratory symptoms. Despite this association, no single microorganism or environmental exposure has been shown to cause HSP. Other symptoms that can precede the disease include fever or headache. The development of HSP has also been linked to cold exposure, insect bites, drugs and vaccination.

The classic presentation is with a tetrad of symptoms or signs:

- Rash
- Arthritis
- Abdominal pain
- Kidney involvement.

The onset of purpura, abdominal pain and arthritis may be in any sequence.

All patients with HSP will eventually develop the characteristic rash, but sometimes the rash is not part of the initial presentation. Arthritis occurs in 80% of cases, and gastrointestinal involvement occurs in 50–75% of patients and is potentially the most serious initial complication of HSP due to the risk of bleeding and intussusception.

The clinical hallmark of HSP nephritis is haematuria or proteinuria or both. HSPN occurs in 50% of older children but in only 25% of children younger than 2 years.

Rash

The rash is most commonly symmetrical and is usually located in dependent areas such as the extensor surfaces of the lower extremities or on the buttocks (Fig. 16.7.3). The rash can be accentuated in areas of pressure (such as sock lines and the waistline). Lesions may also be seen on the forearms with involvement of trunk and face described occasionally in younger children.

Classic lesions consist of urticarial wheals, erythematous maculopapules and larger, palpable ecchymosis-like lesions. Petechiae and target lesions may be present as well. These lesions are typically non-blanching, as they are due to extravasation of blood into the skin**.** Blisters or ulcers or both may develop in the affected areas.

Subcutaneous oedema that is not associated with a rash may develop in the scalp, face, dorsum of the hands and feet or scrotum.

The rash can persist for 3–10 days and progresses in colour from red to purple to brown before fading. Recurrent crops of lesions occur over the next 6 to 16 weeks in up to one-third of cases and may be associated with more severe renal involvement.

Arthritis

This usually involves one to four joints, especially the ankles and knees. There is usually prominent periarticular swelling, tenderness and pain; erythema and joint effusion are uncommon. The joint symptoms are transient, may be migratory, and leave no permanent deformity. Joint symptoms may precede the rash in 25% of cases.

Gastrointestinal

The gastrointestinal manifestations are due to oedema and haemorrhage, predominantly affecting the proximal small bowel. Gastrointestinal haemorrhage is mostly confined to the mucosa and submucosa, and full-thickness necrosis and perforation of a bowel loop are rare.

Abdominal pain is the most common symptom and may mimic that of an acute abdomen. The pain is typically colicky and usually occurs about 1 week after the onset of the rash. The pain may precede the rash in up 15% of cases.

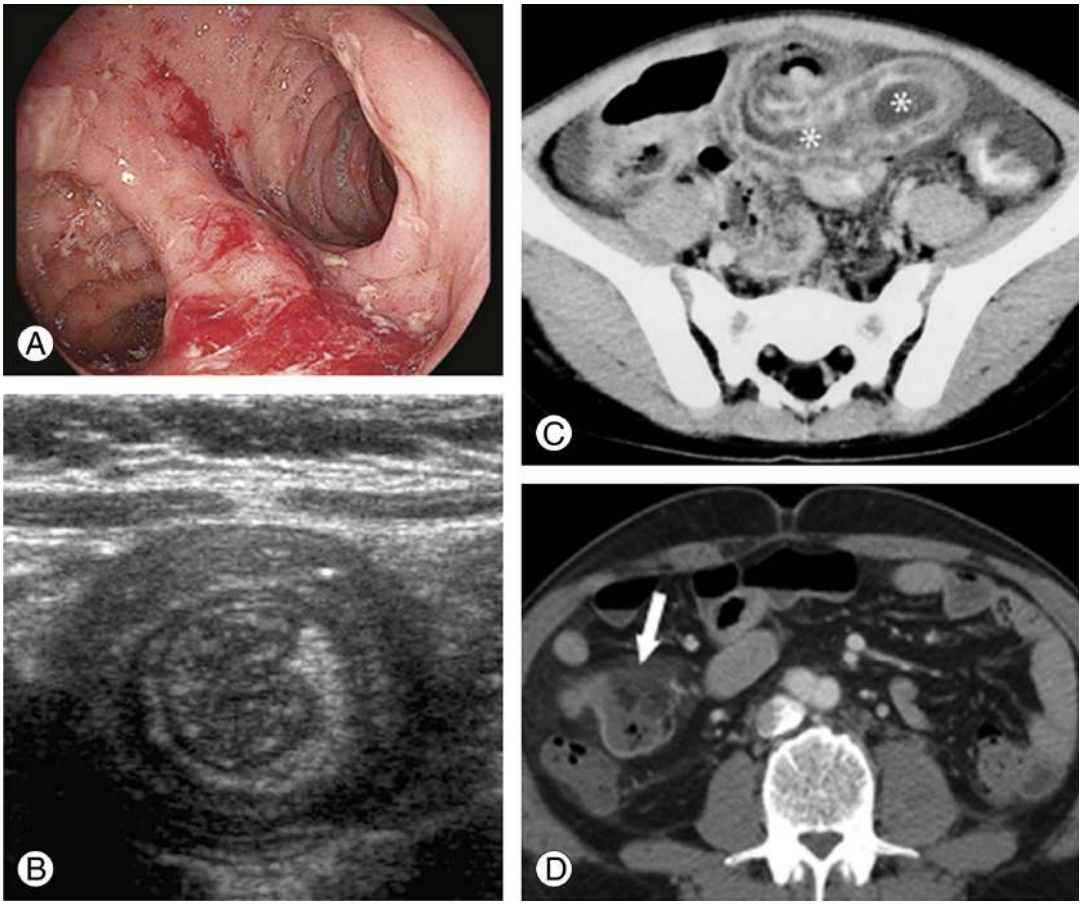

FIG. 16.7.4 Gastrointestinal involvement in Henoch–Schönlein purpura. (A) Endoscopic finding of submucosal haemorrhage and superficial ulceration. (B) Ultrasound showing intussusception. (C) Contrast-enhanced CT scan shows bowel wall thickening of the ileum (∗) with the target sign. (D) Axial contrast-enhanced CT scan showing a focal wall defect area, suggesting perforation. Modified from A: Henoch-Schönlein Purpura – A Case Report and Review of the Literature. Gastroenterology Research and Practice Volume 2010, Article ID 597648; B: Williams H. Imaging and intussusception. *Arch Dis Child Educ Pract Ed* 2008;**93**:30–6; C: Ha HK, Lee SH, Rha SE, et al. Radiological features of vasculitis involving the gastrointestinal tract. *RadioGraphics* 2000;**20**:779–4; D: Chung DJ, Park YS, Huh KC, Kim

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Gastrointestinal bleeding (occult and gross) will develop in 30% of patients. The most severe gastrointestinal complication is intussusception, affecting 3–4% of patients with HSP. The intussusception is limited to the small bowel in two-thirds of cases. Perforation of the bowel wall may occur. Pancreatitis and acalculous cholecystitis are rare complications.

Renal

In contrast to arthritis and abdominal pain, it is very uncommon for evidence of HSPN to precede the appearance of the rash. The onset of renal involvement may be delayed after the onset of other symptoms. In most cases (80% of patients with HSPN) renal involvement will develop within 4 weeks of the onset of HSP. A small number of cases of HSPN have a later onset, but nearly everyone (97%) with HSPN will develop nephritis within 3 months of the onset of other symptoms. The development of HSPN is often asymptomatic, and the diagnosis depends on urine analysis or blood tests or evidence of hypertension.

Other findings

Scrotal involvement can present as oedema or pain. Scrotal ultrasound may show scrotal wall thickening, hydrocele and inflammation of the epididymis and spermatic cord, with or without associated orchitis. Hypertension may be present in 5% of cases.

Diagnosis and investigation

Initial investigations

There is no diagnostic laboratory test for HSP, although a low platelet count will (by definition) rule out the diagnosis. Blood tests may reveal a normal or elevated white blood cell count and possible eosinophilia. The platelets count may be normal or increased, and the prothrombin time and partial thromboplastin time are normal. Factor XIII activity can be reduced and is associated with more severe disease but is not part of routine assessment.

Inflammatory markers (CRP/ESR) are usually elevated, and the initial serum creatinine is normal in nearly all (97%) cases. Autoimmune tests are either

normal (negative antineutrophil cytoplasmic antibody) or the abnormalities are non-specific (serum IgA levels are elevated in half the cases, but there is no correlation with disease severity).

Urine analysis may show isolated haematuria (40% of cases) or proteinuria (25%) or both. Macroscopic haematuria is uncommon (7% of cases), as is the presence of the nephrotic syndrome (3% of cases).

Radiology

Ultrasound is the initial imaging modality of choice for evaluation of abdominal pain or boys with scrotal oedema or testicular pain. CT scans will be needed in patients with signs of peritonitis or haemodynamic instability. On CT imaging, bowel involvement in HSP is seen as multifocal symmetric, circumferential wall thickening with a target pattern. The target pattern is seen after administration of intravenous contrast and consists of enhancing mucosal and serosal layers with an intervening hypo-dense submucosal layer due to oedema. Other abdominal findings on CT scan of HSP include free intra-peritoneal fluid, ileus of affected loop, vascular engorgement in the adjoining mesentery and non-specific lymphadenopathy.

Biopsy

Skin biopsy may be required in cases where the diagnosis of HSP is uncertain. The indications for renal biopsy include:

- acute renal impairment/nephritic syndrome at presentation
- nephrotic syndrome with normal renal function persisting at 4 weeks
- nephrotic range proteinuria (urine protein/creatinine ratio >250 mg mmol) at 4–6 weeks (if not improving spontaneously)
- persistent proteinuria – urine protein/creatinine ratio >100 mg mmol for more than 3 months.

Differential diagnosis

The diagnosis of HSP is usually obvious due to the characteristic rash. However, in the event of an atypical presentation other conditions should be considered. Patients who are suffering from sepsis, malignancy or hypersensitivity to drugs can present with leucocytosis, thrombocytosis, raised erythrocyte sedimentation

rate (ESR) or C-reactive protein (CRP) associated with a rash (urticaria, petechiae, palpable purpura), unexplained arthritis and renal impairment.

Acute haemorrhagic oedema of infancy (AHOI) is a rare type of cutaneous small-vessel vasculitis with a characteristic presentation in infants. It is not clear if it is a mild variant of HSP or a separate condition. AHOI is seen in children between the ages of 4 months and 2 years of age and usually develops after an upper respiratory infection. The child remains in good health but develops a rash on the extremities and face, especially the ears, eyelids and cheeks. The trunk and mucous membranes are usually spared. The rash consists of urticarial wheals, ecchymoses or purpura. The lesions often have a target-like appearance. Oedema may occur around purpuric lesions or involve the hands and feet and extend up the limbs. Joint involvement and renal involvement are uncommon.

The diagnosis in patients with HSP where arthritis is the presenting symptom includes juvenile rheumatoid arthritis, rheumatic fever or a reactive arthritis. The diagnosis in patients with HSP where abdominal pain precedes the rash includes surgical causes of an acute abdomen.

Kawasaki disease is an acute febrile illness with inflammation of small- and medium-sized blood vessels throughout the body, in particular the coronary arteries. The highest incidence is in children of Asian descent, especially Japanese. Eighty per cent of cases occur in children younger than 5 years of age with a peak incidence between 1 to 2 years.

A child with Kawasaki disease has a high swinging fever (greater than 39˚C), associated with marked irritability. Other symptoms and signs can include abdominal pain, diarrhoea, joint pain or arthritis. The diagnostic criteria are fever for at least 5 days, the presence of four out of five cardinal signs (peripheral extremity changes – reddening of the palms and soles, indurative oedema and subsequent desquamation; oral signs – redness within the mouth or on the pharynx, strawberry tongue and red or cracked lips; eye signs – redness of the bulbar conjunctivae; lymphadenopathy – often on one side of the neck; peripheral limb signs – firm swelling of the hands and feet, sometimes including the fingers and toes, with redness of the palms and soles; skin rash – morbilliform, maculopapular, erythematous or target-like) and the absence of any other illness to account for the signs and symptoms. The main complication of Kawasaki disease is the development of dilatation and/or narrowing of one or more coronary arteries.

Treatment

Symptomatic

Treatment is mainly supportive with hydration, rest and analgesics. Outpatient management is appropriate for many patients with HSP. Inpatient management may be required for management of abdominal pain and arthritis/arthralgia, especially if the child is unable to ambulate.

Patients with mild to moderate pain (abdominal or arthralgia) can be treated with paracetamol or non-steroidal anti-inflammatory drugs (NSAIDs). NSAIDs should be avoided if there is evidence of gastrointestinal bleeding or there is renal impairment.

Prednisolone 1 mg kg daily (maximum dose 50 mg) for 2 weeks with subsequent weaning over 2 weeks can reduce the intensity and duration of severe joint pain and severe abdominal pain.

Complications

Patients with severe abdominal pain or significant bleeding (either as haematemesis, melaena or fresh bloody stools) need close clinical monitoring of their haemodynamic status, radiological imaging, consideration of endoscopy and surgical review.

The long-term prognosis of HSP is directly dependent on the severity of renal involvement. The following observations are relevant:

- The development of HSPN can occur up to 3 months after an initial episode of HSP with non-renal symptoms and signs.
- The onset of HSPN may be asymptomatic.
- The severity and⁄or duration of extra-renal HSP symptoms and an older age are the most significant risk factors for developing HSPN.
- The clinical and histological severity at the initial HSPN episode is, in general, predictive of a long-term renal impairment. The risk of long-term renal impairment is less than 2% in patients with only isolated haematuria or proteinuria but is ten times greater in those with a nephritic syndrome or nephrotic syndrome at initial presentation.
- Children with similar presentations of HSPN can experience either complete disappearance of urinary signs or an unexpected late progression to chronic kidney disease – sometimes in spite of an

apparent long-term remission of symptoms. Hence, prolonged observation of persons with HSPN is necessary.

- Early prednisone treatment for HSP does not prevent renal disease.
- There are limited evidence-based data about the efficacy of corticosteroids, cyclophosphamide or other immunosuppressive agents in patients with established, persistent HSPN nephritis.

Prevention

Angiotensin-converting enzyme (ACE) inhibition decreases proteinuria and slows progression of renal impairment in IgAN. Given the similarities in pathology between IgAN and HSPN, ACE inhibition should be considered in the management of persistent proteinuria in HSPN and used as a first-line therapy for hypertension secondary to HSPN.

Long-term prognosis and follow-up

The duration of the disease is less than 14 days in approximately one-third of patients, 2–4 weeks in one-third and longer than 4 weeks in the remaining third.

HSP has a very good prognosis unless there are severe gastrointestinal complications (which cause early morbidity) or HSPN develops (which may cause long-term morbidity).

Patients with HSP and normal findings on urinalysis should undergo regular urine analysis for at least 1 year after the initial presentation.

Patients with HSPN that has caused only microscopic haematuria on initial presentation should be assessed regularly for several years. This group is believed to have an excellent long-term prognosis, but some have developed a late deterioration in renal function.

Patients with HSPN who have recovered fully from clinical features of the nephritic syndrome or nephrotic syndrome should be monitored closely for 5 years.

Patients with persistent haematuria or significant persistent proteinuria or both should be followed by a renal unit.

Women with a past history of HSPN are at increased risk of deterioration of renal function during pregnancy.

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SECTION 17

Psychiatric

OUTLINE

17.1

Paediatric psychiatric emergencies

Michael Fairley

ESSENTIALS

1 Emergency department intervention can reduce the risk of recurrence for self-harm and attempted suicide.

2 The presentation may be somatic but the problem psychological. The identified patient may be the child but the problem may reside in the parents or their relationship with the child.

Introduction

Paediatric patients and their parents can provide complex diagnostic and logistic challenges to staff in a busy emergency department (ED). They often present in a crisis. Thus their social situation overshadows the presenting problem, which is likely to be psychological rather than physical. Though there may have been multiple visits to previous medical practitioners, no clear diagnosis may have been made.

Assessment requires time, patience and unique clinical skills. Intent listening with directed questioning is required. It is important to avoid drawing premature conclusions. Emotional factors should be considered, even when the problem appears predominantly organic.

General approach

Assessment

Parents generally initiate medical contact for a child. In their view, the problem resides within the child. The possibility that they may have a problem to which the child is reacting or that the difficulty is in their relationship is not considered and can be unwelcome if suggested. This preconception can introduce a bias into the way information is presented. If possible, a history from both the parent and the child should be obtained. To enable a child to give his/her account, measures such as reassurance that the visit will not be painful and is not a punishment are needed. Playing and drawing may put the child at ease. If possible, the child should be seen individually.

In adolescents, self-referral for psychological problems is more likely; however, having presented they may still be reluctant to talk. Physical complaints may mask a psychological issue (e.g. headache, abdominal pain and shortness of breath). Reassurance about confidentiality can enhance rapport, but parents will need to be told about safety concerns and authorities notified of abuse. Avoid making undertakings that cannot be honoured.

History

Adolescents wish to be understood, but if they are unwilling to talk about their current problems, they may be prepared to tell about their past and gossip about their friends or family, thereby giving an indirect account. Constructing a genogram, even for a small child, can be an enjoyable activity and illuminate a complex family situation. Always have paper and coloured pens or a whiteboard. Many children communicate better with pictures than words. Children with attention deficit hyperactivity disorder (ADHD) may find it easier to talk if they have something to fiddle with or hold. Extremely restless individuals may be better interviewed in a courtyard than a small interview room.

Collateral history

1. Pre-existing psychological disorders, life experiences, medical conditions and family vulnerability
2. Precipitants – trigger for presentation
3. Presentation – why now, who is most troubled by the symptom
4. Perpetuation – factors that operate to prevent recovery
5. Positives – strengths and resources, coping with previous problems
6. Preconceptions – belief system, expectations from medical consultation.

What the child and parent *really* want (for example, the child has autism or bipolar disorder and a confirmatory letter is sought).

Examination

Examination includes obtaining vital signs, physical and mental state examination. The following points are useful headings for recording observations about mental state:

- Appearance
- Behaviour during interview
- Communication (content, themes, structure)
- Affect (mood)
- Perception and misperceptions
- Cognition (orientation level, formal thought disorder)
- Insight into present situation
- Judgment.

Investigations

Special tests should be confined to the differential diagnosis after clinical assessment. Consider a urine drug screen under appropriate circumstances.

Synthesis of assessment

It should be possible for the emergency physician to determine *why* the child is attending, *who* wants something done, and *where* the main pathology rests: in the *patient*, the *parent* or their *relationship*.

Common Paediatric Psychiatric Presentations

The acutely disturbed child

See separate chapters dealing with this presentation in adolescents (Chapter 17.2) and younger children (Chapter 17.3).

Suicidal patients

Introduction

In Australia in 2015, 3027 people died from intentional self-harm. The death rate was 12.6 deaths per 100,000 people compared to a rate of 10.2 suicide deaths per 100,000 persons in 2006. The rate for males was three times greater than that for females. Overall, suicide was the 13th leading cause of death; however, among people 15–24 years of age, suicide accounted for one-third of deaths (33.9%) and was the leading cause of death. In 5–17 year olds, the suicide rate was 2.3/100,000. The rate for males was 2.4 and females 2.2. The rate for adolescent girls has risen since 2006.[1]

The death of a child or adolescent by suicide is a tragedy. It is an irrevocable act, often in response to transient circumstances. It has an enduring effect on the family and can embolden others to follow suit.

Suicide is chosen when no other course of action remains; the person feels trapped. There is a failure of problem solving. In children and adolescents, the predicament that produces this feeling may seem trivial or easily resolved to an adult with more resources and life experience.

Even in someone determined to commit suicide, there is a struggle between the wish to die and a wish to live. The strength of these forces changes quickly. Fortunately, most suicidal crises are short lived. If lethal means are chosen, fleeting but intense suicidal urges can result in death.

Repeated acts of self-harm, such as cutting or taking overdoses, share many common features with suicide attempts, however, there is less desire for death. Supplanting mental pain by physical pain or overcoming a feeling of numbness is the main aim. Self-harm can escalate to suicide, or death can arise from misadventure.

Assessment

Suicidal patients are in emotional pain. They should be treated with kindness and sympathy. Attending to their comfort and explaining what is going to happen will improve rapport and thus the quality of assessment.[2]

A medical history and physical examination are required to identify trauma, intoxication and underlying medical conditions that may affect the presentation. Cognitive and emotional state should be noted. Toxicology screens should be collected, if indicated. Those who are intoxicated should be observed and reassessed. The patient who is suicidal while intoxicated but denies it when

sober makes assessment difficult. It should be remembered that acute or chronic alcohol abuse raises the risk of suicide, and one-third of adults who completed suicide had consumed alcohol.

Psychosocial assessment should include an examination of suicidal thoughts and behaviours, personal history, home environment and mental state. Suicidal ideation may not be volunteered. Asking about it will not incite action. Though there seems much information to be obtained, letting the patient tell his/her story, in his/her own time, enables you to discover what is most important to him/her. This process is therapeutic:

Suicidal act: Was there an intention to die? Did the wish to die persist afterwards? How lethal was the means chosen? Was the person able to assess the consequences of his/her action (e.g. due to immaturity or low intelligence)? An isolated location, low probability of intervention, precautions against discovery, detailed planning, communication of intent (e.g. social media posts, suicide note or giving away possessions) and rejection of help after discovery indicate determination and serious intent. Trying to gain help during or afterwards, an impulsive act, intoxication at the time and unawareness of irreversibility do not indicate a low risk of recurrence but rather that the person is unpredictable.

Personal history: Precipitants such as bereavement, separation, custody dispute, and disciplinary crisis. Immediate past history such as what is happening at school and home. Interpersonal relationships. Social support systems. Family medical history. Role models for suicide amongst family, friends and heroes.

Mental state examination: Current suicidal ideation, depression, pervasive hopelessness, psychotic features (delusions, hallucinations) and quality of interaction with the interviewer.

Collateral history should be obtained from family, friends, mental health professionals who know the person and police or ambulance that attended.

By the end of the assessment, there should be information that is useful in determining whether this young person has an increased risk of further suicide attempts and for making a mental health referral:

- Details of this and previous suicide attempts
- Nature and meaning of current crisis
- Current or past depression, other psychiatric disorder, drug and alcohol abuse
- History of violent behaviour
- Social disadvantage: low socioeconomic status, limited educational achievements, low income, poverty and associated factors such as itinerancy
- Indigenous
- LGBTI status
- Parental loss through separation and divorce
- Physical or sexual abuse, domestic violence
- Impaired parent–child relationships
- Parents with a mental illness such as mood disorders, harmful drug use (including alcohol) or problems involving violence
- Loss through suicide in his/her family or peer group
- Experiencing a greater number of life stresses than normal
- Personality strengths and usual coping mechanisms
- Social supports and potential source of help.

Hazards during assessment

The young person may minimise the seriousness of the event in order to avoid hospital admission so that he/she can make a further attempt, run away, obtain sharp objects for later use or make a further attempt in ED. Parents and carers may deny the seriousness of the attempt. Staff hostility, especially towards frequent attenders, may interfere with developing rapport.

Negative attitudes are common in ED staff, especially to patients who repeatedly self-harm.[3] Irritation and hostility arise from the belief that hospital is for the physically ill and these patients are attending unnecessarily. A distinction is made between illegitimate and legitimate patients based on overt responsibility. Legitimate are seen to have a greater entitlement to care even when their actions may have caused the presentation, such as ketoacidosis in a non-compliant adolescent diabetic.

Staff anxiety arises from anticipation of being hurt emotionally, feelings of failure as nothing can be done, not knowing how to speak to these patients and expectations to do something that is unrealistic.

ED staff are less likely to feel sympathy for self-harming patients than other hospital staff. Doctors hold more negative views than nurses, and those with more experience have more negative attitudes. Poor clinical practice, such as underestimation of risk through attribution of the injury to attention seeking, can result. Patient satisfaction with their ED experience and outcome measures are also affected. Staff training consistently leads to improved attitudes and understanding of the reasons for the behaviour.

Risk assessment using a checklist-based algorithm is ineffective at predicting suicide in an individual. Most who are depressed or self-harm do not commit suicide; suicidal ideation is common but in comparison, completed suicide is rare. Pokorny, in a prospective study of psychiatric inpatients, identified a 'high-risk' group using rating scales and known predictors such as suicide attempt, depression, social isolation and drug or alcohol abuse.[4] The high-risk group had a suicide rate four times higher than the group as a whole. Thus it was possible to predict a group at increased risk. Those who were admitted for a suicide attempt subsequently had a suicide rate of 1.7% per year. This was a very high rate but still of little assistance in predicting suicide in an individual. Sensitivity and specificity of risk scales were low.

False positives result in excessive restriction of those who will not commit suicide, and false negatives lead to inadequate treatment of those who will. It is still possible to define a group that is at increased risk and implement effective measures to reduce suicide. Psychiatric admission is clearly not feasible for all. Fortunately, there are effective measures that are not too onerous.

Management following an attempt

While et al. investigated measures that lower the suicide rate. Using national suicide data in England and Wales, implementation of key recommendations of the National Confidential Inquiry into Suicide and Homicide by People with Mental Illness was examined. Those services implementing seven to nine recommendations experienced a decreased suicide rate compared to no change in rate for those implementing fewer. A 24-hour crisis team, with a single point of contact providing prompt response and short-term input in the community, led to the biggest fall of suicide rate from 11.44 per 10,000 patient contacts per year to 9.32. Other measures included in the recommendations were assertive outreach in the community for those that were difficult to reach, follow-up within 7 days of discharge, dual diagnosis policy for patients with psychiatric disorder and

alcohol/substance use, multidisciplinary review and information sharing with families following suicide, training of clinical staff in the management of suicide risk, criminal justice sharing and removal of potential ligature points on inpatient wards. The greatest benefit occurred in the most deprived catchment areas and those with the most patients. Follow-up within 7 days was effective in reducing suicide in the 3 months after suicide attempt. Assertive follow-up was most effective for those who were non-compliant with medication and missed appointments.[5]

A mental health assessment should always occur. Hickey found that deliberate self-harm patients who left ED without psychiatric assessment were twice as likely to self-harm again during the next year.[6] Psychiatric admission, however, is not always desirable even when the adolescent cannot assure his/her safety. Chronically suicidal adolescents may react to efforts to keep them safe as a challenge, engaging in more ingenious and dangerous behaviours while in hospital. Relieved of the demands of everyday life, a reluctance to leave hospital can develop, leading to increased suicidal thoughts around the time of discharge. Admission is helpful when there is diagnostic uncertainty or to initiate a new treatment.

A safety plan should be developed in conjunction with the patient. Identifying suicidal thoughts, developing ways to cope with them and listing sources of help can be started in ED. Details of the follow-up appointment with a named mental health professional within the next 7 days, the telephone numbers of the 24-hour crisis service, friends and family should be on a card and entered into the patient's phone. Measures to reduce access to lethal means, such as secure storage of medication, should be discussed with parents.

Mental health follow-up, for as few as three sessions, reduces the risk of further self-harm. In a large international randomised control trial following suicide attempters, Fleischmann found that 0.2% of those who received brief intervention and contact committed suicide in the following 18 months compared to 2.2% of the controls.[7]

Suicidal patients: important considerations

In summary, the major considerations for the assessment and management of a child or adolescent presenting to the ED with suicidal ideation include the following:

1. Medical assessment for trauma, drug ingestion and other physical conditions that could affect the mental state. This may lead to admission to a medical unit
2. Assess cognitive and emotional state. Listen to the adolescent's story to discover the predicament that led him/her to the attempt. Avoid reliance on checklists especially for predicting risk
3. Safety while in ED
4. Safety planning
5. Mental health assessment in ED
6. Mental health follow-up with treatment of depression and other psychiatric condition.

Anxiety disorders

Anxiety is a complex mixture of somatic and cognitive symptoms: autonomic arousal, worry about what has happened and apprehension about what will happen. Though experienced as fear, there is no physical danger. The pulmonary, cardiac and other sensations enhance the experience of alarm. It is a ubiquitous condition that varies from the physiological to the pathological in its presentation. Panic disorder often starts in adolescence and has a lifetime prevalence of 1.5–3.5%. Cases of syncopal collapse may manifest as a panic attack. Conversion disorders present with acute paralysis, loss of vision or sensation.

Obsessive compulsive disorder (OCD) represents an attempt to reduce anxiety through extreme control of one aspect of the environment. Children are unable to resist the urge to repeat rituals such as tapping a specified number of times. They frequently attempt to enlist family members in performing rituals and become enraged when they resist. This situation may be mistaken for a behaviour disorder.

Separation anxiety presents as school refusal. The child often presents with a somatic complaint such as abdominal pain. Other anxiety diagnoses include phobias, post-traumatic stress disorder and generalised anxiety disorder. Early identification can reduce the number of medical investigations and enable mental health referral.

Acute anxiety can be treated effectively with a comprehensive approach utilising cognitive therapy, behavioural techniques, psychotherapy, counselling and medication. SSRI antidepressants are first-line treatment but do not give

immediate symptom relief. For OCD, high doses are often required, and the response may take up to 3 months. Beta-blockers effectively reduce autonomic symptoms and interrupt mounting anxiety driven by sensations usually associated with danger. They should be avoided in asthma.

Anxiety is contagious. Families present with a sense of urgency, and there is pressure to prescribe medication that provides immediate relief. Benzodiazepines and antipsychotics are often given in this situation. This should be avoided, if possible. Anxiety is chronic, but the benefits of these medications are short lived. There is the risk of dependence, and side effects are disproportionate to the benefits. There are limitations on prescription for antipsychotics that may subsequently place the GP in an awkward position or expose the family to expense. Non-pharmacological treatments, that are hard work and take time, will be eschewed in favour of something that can be taken. Arranging a referral for mental health assessment may help contain parental anxiety.

Psychosis

Hearing ‘voices’ is very common in children and often persists in those with intellectual delay. The voice is the child’s own thoughts and may say to do something naughty, thus distancing the child from the impulse. By early adolescence, the world is more complex, but the cognitive maturity may not have been achieved to maintain contradictory positions simultaneously – ambivalence. One voice may say to do something while another warns against it.

It can be difficult to differentiate thoughts from auditory hallucinations. They are often experienced within or outside the head and have distinct qualities, such as a man’s voice, that can be described. The content is almost always unpleasant and can be frightening. In depression, there are critical comments on appearance and performance–saying that the person is a burden to the family, does not deserve to live and should commit suicide. In anxiety, the voice is anticipating all the things that will go wrong. Activity and company can reduce the voices. They are often worst when alone of an evening. Self-harm, cutting and burning, can be an attempt to use physical pain to displace mental pain. There is a risk of acting on the voices’ instructions, in order to get relief.

Regular marijuana use often leads to the misperception that everyone is staring at the person, knows about him/her and is mocking. This usually settles spontaneously if consumption is ceased. Since marijuana is often used to self-

medicate dysphoria, the emergence of this symptom can result in increased use.

The underlying medical condition is usually evident in delirium. The onset is abrupt, and the mental state fluctuates. Visual and olfactory hallucinations occur as well as auditory and can predominate.

Unstable mood is one of the commonest psychiatric symptoms and can occur in any condition. It is often mistakenly equated with bipolar affective disorder. The mood swings in bipolar disorder are more prolonged. Excessively elevated mood is more likely to be the effect of antidepressant medication than bipolar disorder. Common symptoms are loud and continual talking, reduced sleep and disinhibited behaviour. A high dose of antidepressant in young children is most likely to produce this effect. Symptoms will usually settle with cessation of the antidepressant.

Psychosis is rare before late childhood. The prevalence increases through adolescence. Environmental stress precipitates illness in children with a genetic predisposition. Onset may be abrupt or preceded by a gradual withdrawal, academic decline and altered perceptions. If acute, there are more likely to be hallucinations, perplexity and fearfulness. It can resemble a confusional state. Mood symptoms are common, making the distinction between schizophrenia and bipolar disorder difficult. Schizophrenia is characterised by auditory hallucinations. Delusions may be created to account for the altered experiences (e.g. spirits coming up from underground). Thought disorder is usually prominent with the flow being blocked or disrupted connections between ideas. Idiosyncratic speech, perseveration, ideas of reference and complaints of thought control also occur. Apathy, flattened affect and withdrawal are disabling and difficult to treat.

Treatment

Combined psychotropic and psychotherapeutic treatment is required. Depression should have a mental health assessment because of the risk of suicide. There is a delay, sometimes weeks, before antidepressants improve mood so their introduction in ED is not essential, if follow-up is arranged.

Bipolar disorder and schizophrenia require antipsychotic medication. Olanzapine (initial dose 2.5–5 mg), risperidone (initial dose 0.5 mg) and quetiapine (initial dose 25 mg) are the most commonly used. They are calming, reduce aggression and promote sleep.

Acute dystonic reactions are more likely with risperidone, and all can cause

akathisia, which is distressing for the patient but may be overlooked by the clinician. As use of antipsychotic medications may be prolonged, and metabolic syndrome is a serious side effect, it is important to establish baseline height, weight and girth. Lipids, HbA1c and fasting blood glucose should be collected early in treatment.

Other psychiatric presentations

Acute dystonic reactions

Antiemetic and antipsychotic medication can induce acute muscle spasm, most commonly torticollis. Oculogyric crisis, opisthotonos, trismus and grimacing can also occur. The experience is painful and frightening to the patient and carers. Laryngeal spasm is rare but can compromise respiration. Onset is usually within days of starting the medication or following a dose increase. Young males are most at risk.

Treatment is **benztropine 0.02 mg kg to a maximum of 1 mg** intramuscularly or by slow intravenous injection. Response is usually rapid. If there is no response, repeat once after 30 minutes. If still no response, review the diagnosis.[8] To prevent recurrence, it is often recommended that the same dose be given orally and repeated twice daily for the next 2 days. If possible, the causative medication should be ceased. Occasionally, anticholinergic medication causes a delirium that can be confused with psychosis. Further antipsychotic medication should be avoided.

Night terrors

Sleep disruption is a parent's most frequent concern during the first 2 years of a child's life. Half of all infants develop a disrupted sleep pattern serious enough to warrant physician assistance. Night terror disorder is characterised by recurrent episodes of intense crying and fear. There are signs of autonomic arousal, and there is difficulty waking the child during episodes. Children do not recall a dream after a night terror and typically do not remember the episode the next morning. Night terrors are frightening episodes that disrupt family life and cause the child significant distress. Usual onset is between 3 to 12 years. The disorder generally resolves during adolescence.

An estimated 1–6% of children experience night terror episodes. Recurrent night terror episodes accompanied by significant distress and impairment are less

frequent.

Peak frequency in children younger than 3.5 years is at least one episode per week; among older children, peak frequency is one to two episodes per month.

History

The most important step toward diagnosing this disorder is to obtain a detailed history:

- Approximately 90 minutes after falling asleep, the child sits up in bed and screams. Prominent autonomic activity (e.g. tachycardia, tachypnoea, diaphoresis, flushing) occurs. The child appears awake but confused, disoriented and unresponsive to stimuli.
- Most episodes last 1–2 minutes, but the child may remain inconsolable for 5–30 minutes before relaxing and returning to quiet sleep.
- If the child awakens during the night terror, only fragmented pieces of the episode may be recalled.
- In the morning, the child typically has no memory of the experience.

Management

This consists of educating the parents that night terrors are part of the normal development of sleep. The episodes are not harmful, and it is best not to wake the child. This contrasts to nightmares, after which the child often wakes in distress and requires comforting. As lack of sleep predisposes to night terrors, ensure routine use of sleep hygiene measures.

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17.2

The treatment of the behaviourally disturbed adolescent (see Fig. 17.3.4)

Kenneth Patrick Nunn, and Meenakshi Rattan

ESSENTIALS

1 Emergency psychiatry is treating the underlying neurobehavioural processes NOT the cognitive content or psychiatric diagnosis.

2 The psychiatric ABCC is a rapid assessment of risk that can be done in a few minutes and enables decisions to be made on the need for sedation and whether sedation is cooperative or non-cooperative.

3 When thinking about management it is worthwhile to move back through the ABCC from the most disrupted young people through to the early levels of distress and dysfunction:

C – If cognitive processes are very disrupted hospital admission will be necessary irrespective of the eventual diagnosis.

C – If containment is threatened – offer cooperative sedation early, and put security on notice. If containment is actually being breached all other treatment must wait until containment is addressed voluntarily or involuntarily.

B – If behaviour is extreme – actively offer relief with calm reassurance, nursing presence and medication while consciously preparing for escalation to a containment breach.

A – If arousal is high establish whether this can be readily managed by calmness and cooperative use of medication. If calm is not forthcoming be prepared for behavioural escalation.

Introduction

Emergencies require routines and procedures that make decision making seamless, effective and professional. Psychiatric emergencies are no different.

Psychiatry, and adolescent psychiatry in particular, requires clear, well-rehearsed routines to avoid chaotic decision making. No area of medicine is more difficult to routinise *without practice* than psychiatry. No area of psychiatry punishes poorly operationalised routines as much as adolescent psychiatry.

As in all paediatrics, the first question is not ‘which illness?’ but ‘how unwell are they?’ The task of clinical stabilisation is much more important than satisfying a particular psychiatric diagnosis. Once the essentials are understood and applied, the emergency physician can provide consistent, high-quality care in triage, initial stabilisation and management of immediate risks before preparing for transfer for definitive management and ongoing treatment where needed.

This chapter is written with the underlying premise that psychiatrists, especially child and adolescent psychiatrists, will be in short supply and sometimes not trained in emergency psychiatry. For the present, outside of large paediatric hospitals, emergency adolescent psychiatry largely falls to emergency physicians and general paediatricians.

The principles of psychiatric triage

Purpose

Triage enables the early identification and treatment of factors that may threaten the immediate safety and wellbeing of the patient, others and staff in triage and the emergency department (ED). The following priorities are important in the below sequence:

1. Safety
2. Management of distress and behaviour
3. Diagnosis.

It is critical that the task of definitive diagnosis does not delay acute management. Whether or not a young person has a particular psychiatric disorder is of a lower order priority in the acute setting.

Accurately identifying the:

- *components* of threat
- *complexity* of the threat (that is the number of domains of risk)
- *severity* of the threat

- *mitigators* of threat (for risk management)

all within a rapid time frame requires training, skill and experience.

Time is risk

The more time in the ED environs taken before treatment begins, the greater the risk.

Why?

1. Impulsivity, agitation and lack of relief of distress converge with the sensory overstimulation of most ED waiting areas.
2. Time passing constitutes an escalating risk of a loss-of-control or loss-of-containment event.

Table 17.2.1

Early warning signs

Threats to safety	Syndromes of distress	Organic flags
If you feel unsafe or moved to protect others	If you feel distressed at watching	If you can’t make sense, and the child looks unwell

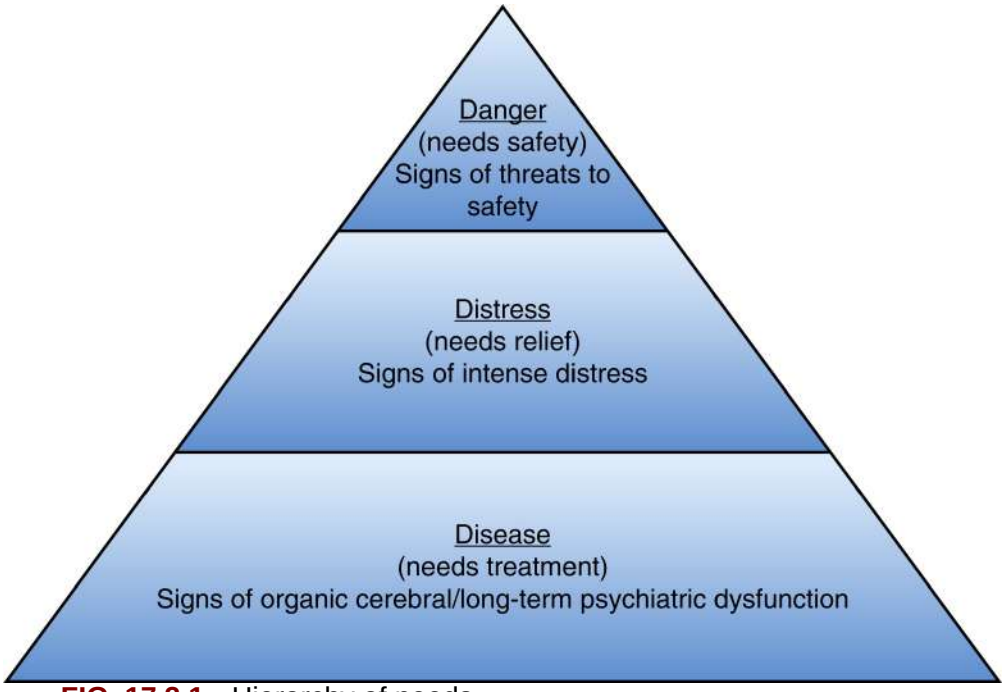

FIG. 17.2.1 Hierarchy of needs.

3. Where agitation is present, motor activity increased and cognitive processes clearly altered, the risk rises dramatically, even after short periods of waiting.
4. The slide down the spectrum of distress to disruption – anxiety, agitation, anger, together with demanding, impatient, impulsive and explosive behaviour – leaves few options once in full progress.

On the other hand, the rapid initiation of triage and the commencement of an altogether less disruptive process – seen, relieved, treated, monitored, transitioned – are each associated with a reduced risk of loss of control within the ED.

Pre-triage

An immediate threat of patients to others, themselves or to the medical safety of carrying out procedures requires immediate action analogous to a cardiac arrest.

Practicing commonly encountered scenarios with acceptance of ‘error and inefficiency’ is essential.

Early warning signs – subjective

Identifying a safety threat to staff and others?

Questions such as those listed below may all provide useful clues, if considered (Table 17.2.1). It is worthwhile acting on this as a given until reassured otherwise:

- Do I feel unsafe as a clinician in the ED?
- Am I anxious for the patient's welfare?
- Is there a sense of threat around the patient?
- Are staff members avoiding the patient because they are afraid?

Early warning signs – observed

Even before we sit down with the patient and begin to listen to his/her story, some behaviour stands out as 'out of control'.

If unusual, potentially dangerous, combative or distressing behaviour is observed:

- A louder than usual voice
- An argument between a parent and child
- A surly withdrawn father
- A seemingly drunken teenager

sitting down is a helpful first step to reducing autonomic arousal and activation in the patient.

The hierarchy of needs

All triage involves addressing a hierarchy of needs (Fig. 17.2.1).

Safety, **s**ymptom **r**elief, initial **i**nvestigation and a **p**rovisional **d**iagnosis (SSRI-PD) form the underpinnings of emergency psychological care.

Mental state monitoring and the active exclusion of medical contributors commence immediately upon completing the initial stabilisation.

Signs of threat to safety

1. Threats to self
2. Threats to others
3. Threats to the fabric and good working of the ED imperilling the

treatment of others.

Signs of intense distress

By way of analogy with the provision of acute life support, it is helpful to employ the ABCC mnemonic.

The ABCC of rapid psychiatric assessment

Arousal – autonomic fright, fight, flight arousal
Behaviour – behavioural activation and the presence of withdrawal or agitation
Containment – behavioural control within a social setting
Cognitive processes – the coherent communication of reality-based thinking and feeling.

Identifying early changes, rate of change and the extremity of change to each component of the ABCC gives a quick sense of what needs to be done.

Arousal

The triad of early signs of change in arousal, volatility of arousal and extremes of arousal is our focus. Thus, early autonomic signs of either sympathetic (crisis emotions) or parasympathetic shift (calming emotions) include pallor, flushing, tachycardia, bradycardia, tremulousness, mydriasis and tachypnoea. Marked hypervigilance and clouding of consciousness or rapid excursions between the two are very helpful in identifying the need to intervene rapidly.

Anxieties may be more difficult to assess with lack of fine social and emotional tuning with either unresponsiveness or exquisite responsiveness to the environment or a rapidly changing mixture of both.

If the arousal is sufficiently disruptive the child or young person may have a functional overflow of his/her arousal into his/her behaviour.

Behaviour

Early signs of change in behaviour, volatility of behaviour and extremes of behaviour are our focus. Sitting with one leg constantly shaking, wringing of hands or stroking of hair as part of anguish or anger all portend an imminent

deterioration in behaviour. Constant pacing or refusal to move, a loud voice or speaking very quietly, swearing excessively and particularly offensively, or refusing to talk, a broader picture of social disinhibition or extreme inhibition, reflects extremes of response and lack of fine social and emotional tuning.

If these are sustained they may overflow and have an impact on the other patients, staff and the functioning of the ED. If this impact is sufficiently disruptive, it may constitute a containment threat or demand for containment.

Containment

Behavioural control to reduce major threat and disruption, in an acute medical setting, is termed *containment*. Containment is usually a physical process which reduces the capacity of a patient to disrupt the ED. It aims to reduce risk to others, risk to self and risk to the environment and safe working of the ED. The acute and open nature of the ED means that any disruptive threat may constitute a broader threat to the provision of urgent medical treatment to the young person in question or to other patients.

Changes in arousal normally precede changes in behaviour. Changes in behaviour risk loss of containment. Early warning signs of a loss of containment event have already been covered under the headings of arousal and behaviour. Frequently these early features will have happened prior to being seen in the ED, and the demand for containment is the presenting request.

Running away, disrupting the ED, damaging property and creating an atmosphere of threat and menace would all constitute a containment threat or a containment failure. In extreme cases this may involve weapons, the police and the clearing of the ED with cessation of medical activities while a local disaster response is put into place.

In most cases, some form of containment, such as the police being in attendance, will be in place before a patient is brought to the ED. It is important that this containment is not lost in the transition process into the ED or in transfer from the ED.

Cognitive processes

Early signs of thought disruption are changes in arousal, behaviour, especially speech and containment that belie incoherence of thought, and other cognitive processes.

Other cognitive processes such as attention, executive function (planning, judgment, problem solving and insight) and perception underpin thinking and consequent action. This is not to say that thought or speech *content* is irrelevant. But the *process* of thought is more relevant to the emergency setting.

In the case of psychosis, containment failure is what is seen, and the fragmentation of thought processes is what is heard. Paranoid or self-destructive ideation is seen in the arousal, behaviour, speech and containment assessment. This can be evaluated without a formal mental status assessment.

What they say is not as critical as how they say it

The young person may be worried, fearful or self-loathing or behave in a bizarre, threatening or angry manner. However, the processes of perceived threat (in the extreme, paranoia) and the pursuit of relief (in the extreme, self-harm and suicide) are the underpinning cognitive processes foremost in any emergency assessment.

Extremes of incoherence or paranoid 'coherence' apply here as well. Jumbled, accelerated, guarded, slowed down, rigidly pre-occupied, fixedly convinced or non-communicative speech and thought are all more worrying process variables, irrespective of what or who is upsetting the patient.

Managing the ABCC including restraint and acute sedation

It is inappropriate to attempt to obtain a detailed developmental history, systems review or full exploratory psychiatric history from an acutely distressed and very ill young person.

When thinking about management it is worthwhile to move back through the ABCC from the most disrupted young people through to the early levels of distress and dysfunction:

C – If cognitive processes are very disrupted, hospital admission will be necessary irrespective of the eventual diagnosis, and ***a sedative antipsychotic such as droperidol is indicated.***

C – If containment is threatened, offer cooperative sedation early, and put security on notice. If it is actually being breached, all other treatment must wait until containment is addressed, voluntarily or involuntarily. ***Droperidol*** *will be necessary.*

B – If behaviour is extreme actively offer relief with calm reassurance, nursing presence and medication while consciously preparing for escalation to a containment breach. *Once agitation is established sedation* ***with olanzapine (voluntary) or droperidol (involuntary)*** *will be needed.*

A – If arousal is high establish whether this can be readily managed by calmness and cooperative use of medication. If calm is not forthcoming be prepared for behavioural escalation. ***Quetiapine will usually be sufficient.***

There are only **six** essential things which must be attempted:

1. ***Rapport and respect*** – even where they cannot be achieved.
2. ***Identify the immediate issue*** of concern to the young person and to others while establishing a formulation in medical terms.
3. ***Identify risks*** – self and others, including child maltreatment, medical and reputation risks.
4. ***Symptomatic relief*** – provide immediate treatment with whatever biological, psychological and social supports and resources are available. *The provision of relief is the single biggest factor in reducing risk.*
5. ***Identify psychosocial supports*** and stressors (immediate), i.e. who is the consenting authority for the young person and where he/she normally lives. None of these may be available in the acute situation. However, he/she will save a great deal of time in determining the disposition of the patient.
6. ***A plan of action*** for the next 24 hours.

Sedating the adolescent brain

When attempting to reduce hyper-arousal and behavioural agitation, to manage a breach of containment, it is helpful to consider which parts of the brain – or neurobehavioural systems – are being targeted. The neurobehavioural systems are referred to here as sub-brains as a useful shorthand.

The thinking brain **(the cortex**) – easiest to sedate, especially in children – inhibits the other two 'lower' brains. When the cortical brain is sedated, the two lower brains (disinhibition) are released. Benzodiazepines are often enough to

sedate this brain. *However, paradoxical excitement may result, so quetiapine is preferable:*

> Target: arousal and preoccupation – for example, the inability to stop thinking about distressing events despite sustained talking through and reassurance.

The feeling brain **(the limbic system)** – takes longer and more medication to sedate, especially when distress is established.

Quetiapine may be enough, but where psychosis, anger, agitation often olanzapine (voluntary) or droperidol (involuntary) will be needed:

> Target: arousal and distress – for example, the inability to find emotional relief despite sustained and skilled reassurance.

The moving brain **(the basal nuclei)** – takes longest to sedate and first to awaken from sedation.

Olanzapine or droperidol will always be needed to sedate this brain:

> Target: behaviour and agitation – for example, the inability to be able to move in an emotional paralysis or to sit still due to agitation, despite sustained reassurance.

Tracking the seven stages of sedation

1. ***Fixation***: immobilising the body to enable vascular access or intramuscular injection and a safe medical procedure.
2. ***Induction***: commencing sedation with the steady reduction of consciousness.
3. ***Disinhibition***: loss of emotional and behavioural control associated with loss of cortical inhibition before limbic and basal nuclei are similarly inhibited. This may occur after an initial settling.
4. ***Stabilisation*** of arousal depth – titrating to a level of sedation that maintains gag reflexes, pharyngeal patency and adequate breathing and oxygenation.
5. ***Maintenance***: high level observation with clear parameter thresholds and specification of appropriate response when thresholds are breached.

6. ***Emergence***: the period of decremental lowering of medication to allow a transition to the fully conscious state, with an awareness that disinhibition may occur during this process as during the establishment of sedation.
7. ***After care***: the psychological explanation and support required specifically in relation to the sedation process.

Five tips on sedating adolescents

1. Start high (without bolus), and titrate down in an emergency.
2. If there is definite motor agitation do not use a benzodiazepine alone, which may disinhibit the patient. Add droperidol, and monitor for extrapyramidal side effects, such as dystonia.
3. If re-sedation is likely, move to regular doses rather than p.r.n., 'ebb and flow prescribing'. Less medication is needed if non-p.r.n.
4. Establish sedation away from the main ED (more private and less disruptive), and maintain and monitor the sedation close to the main ED.
5. Plan for emergence risks – ensure security presence during emergence from sedation or preparedness to re-sedate.

Signs of organic dysfunction

- Elevated ***temperature*** in a psychotic patient
- ***Appearance*** – looking lost or 'out of it' with lowered eyelids, glazed eyes and 'in a world of his/her own'
- ***Behaviour*** – recent personality change with loss of social fine-tuning and reversal of sleep rhythms
- ***Speech*** – muddled speech and thinking, often with paranoid but fragmented themes
- ***Perception*** – visual illusions, i.e. misidentifying actual stimuli, e.g. shadows mistaken for people or frank visual and tactile hallucinations (especially with antihistamine or anticholinergic overdose)
- ***Ideation*** – often paranoid in thinking
- ***Cognition*** – clouded and fluctuating levels of consciousness and disorientation
- ***Judgment and insight*** – impaired markedly.

Differential diagnosis – medical

1. ***Specific toxidromes*** and syndromes of adverse drug reaction for:
    - substance abuse
    - overdose
    - accidental ingestion
2. ***Delirium*** – generalised acute brain syndrome
3. ***Frontal lobe syndromes*** (including traumatic brain injury, solvent abuse and intellectual disability)
4. ***Seizure-related disorders***
5. ***Starvation-related syndromes***
6. ***Cerebellar*** *dysfunction*:
    - slurred speech
    - ataxic gait
    - incoordination
    - substance abuse, e.g. benzodiazepine
    - toxicity from prescribed medications, e.g. anticonvulsants
    - ingestion of poisons, e.g. alcohol, phenytoin.
7. ***Dementia*** – generalised chronic brain syndrome
8. ***Neurodevelopmental:*** autism, fetal alcohol spectrum disorder (FASD) and severe attention deficit hyperactivity disorder (ADHD), intellectual disability with associated respiratory, ear, urinary or other pathology or pain (see Chapter 17.3).

Differential diagnosis – psychiatric

1. Acute on chronic self-harm or aggression in chronic complex post-traumatic stress disorder in abused, neglected and/or out-of-home-care youth.
2. Substance abuse in as yet undiagnosed mentally ill young people – especially major depression or bipolar disorder.
3. Bipolar depression treated with anti-depressants or stimulants.
4. Aggression treated with stimulants or SSRIs or both, worsening aggression. *There is no acute indication for SSRIs or stimulants.*
5. Psychosis treated with stimulants or weight-loss dopamine agonists.
6. Cryptic abuse in outwardly well-functioning family.

7. Emerging paranoid illness presenting for the first time with aggression.

The management of acute risk including medical risk

Managing threat to staff and others

- Do not ask medical or nursing staff to expose themselves to unreasonable risks.
- Offer cooperative oral sedation early such as quetiapine or olanzapine.
- Work closely with security staff, seek other help early and provide sedation backup.
- Don’t address long-term issues in an ED setting.

Behaviours that usually de-escalate aggression

Nothing works all the time, and nothing works in every case. But some of these are likely to be helpful.

Ten DOs

1. Be respectful, friendly and open (single most important strategy, especially respect).
2. Be quicker to listen than to speak.
3. Speak clearly, quietly, gently and calmly with an expectation that they will respond.
4. Use humour that shows we accept we have things about us that are not ideal, perfect or completely ‘respectable’, especially when we are derided or spoken to rudely.
5. Declare desire to be helpful even if it is not always known how to help or what it is they want.
6. Relax posture, voice and face, even if preparing internally for fight and/or flight.
7. Move slowly, predictably and with due respect for distance.
8. Distract to details that they might be interested in as well, especially things about them and what they like.
9. Acknowledge any faults in our behaviour (not someone else) that might be contributing to their being upset.

10. Acknowledge your own tiredness, a hearing problem (if they are withdrawn, hostile, talking quietly), irritability or crabbiness after a busy shift which emphasises our humanity and reduces their fear.

Ten DON'Ts

1. Try to shame them into good behaviour by telling them they are childish, silly or stupid.
2. Try to get above them (so called 'towering') physically, by position, verbally, intellectually or socially.
3. Talk without listening or 'nag'. Don't all talk at once – one person only.
4. Issue ultimatums – 'do this or else' – almost always leads to 'or else' in this population.
5. Back into corner either physically or psychologically (unless everything else has failed and safety demands we must).
6. Adopt a 'thou shalt' tone of voice instead of a reasonable request.
7. Raise voices in a counter 'arc-up' to the patients.
8. Mock, criticise or accuse the patients at any time.
9. Rush the process or create time pressure.
10. Hark back to previous behaviour or try to sort out longstanding issues, unless this is a previously agreed treatment goal.

The four main themes

1. *Communicate respect.* Many of them have not been treated with respect.
2. *Communicate our desire to keep everyone safe.* We want them, and us, to be safe. Many of our patients have not lived in safe environments.
3. *Take responsibility.* Show a willingness to acknowledge we may have contributed to their distress by what we have done or not done. Many of the young people who will present have lived with people who have never accepted blame, use denial a great deal and blame others.
4. *Appreciate their distress* even though it may not be fully understood why they feel like this or how bad they feel. Many of the patients have been told how they feel (even if they did not) and had the legitimacy of their distress invalidated. They may never have been in a situation where high distress is tolerated, validated and appreciated so long as safety is high as well.

Identify patients' threat to themselves

- If patients or those with them feel unsafe, they probably are unsafe.
- If your ABCC evaluation says they are unsafe they are unsafe.
- If a colleague outside the hospital says they have been behaving unsafely, be very wary of not honouring their assessment.

Managing a threat to themselves

- Remain calm and firm, enlist security, maintain constant observation and, if actually attempting, once you are able to ensure staff safety, intervene immediately.
- Identify treatable disorder and social predicaments while waiting for suicidality to settle (usually attenuates over hours to days NOT weeks to months).

Identify medical risks

- The broader medical needs of the patient need to be recognised.
- Psychiatric disorder does not protect against medical disorder.
- It is possible to do a great deal with a non-cooperative patient to clarify medical status.

Manage specific medical risks

- *Obtundation* – due to panic sedation in response to a previous failed sedation in adolescents who became disinhibited (see below).
- *Extrapyramidal side effects* – nuchal spasm with headache is common in the young as a form of dystonia.
- *Respiratory* – laryngeal dystonia – especially in younger patients but still rare. Use of benzodiazepines (even in reduced doses) or anticholinergics such as benztropine *with* droperidol.
- *Cardiac* – prolonged QTc, especially in poor CYP2D6 metabolisers (5% of normal population).
- *Neurological* – delirium (common, especially substance abuse), serotonergic syndrome, neuroleptic malignant syndrome (rare) (NMS).
- *Concurrent medical disorder* – asthma, diabetes, traumatic brain injury,

epilepsy, atopy and anorexia.
- *Dermatological* – lamotrigine-induced Stevens–Johnson syndrome.
- *Metabolic* – lithium toxicity – especially chronic toxicity.

Sentinel nursing observations post intramuscular or intravenous sedation

Respiratory rate, O_2 saturation, pulse, blood pressure and level of consciousness should be checked continuously during the maintenance of sedation – and documented each 15 minutes for 2 hours, then each 30 minutes for 2 hours.

An electrocardiogram to screen for prolonged QT interval should be performed as soon as possible after any sedation with zuclopenthixol acetate or droperidol.

Four tips for monitoring adolescents

1. Expect the medication to be metabolised quicker.
2. They become disinhibited more easily going into sedation and coming out of sedation.
3. Maintaining sedation intravenously (IV) should be done with a slow injection over several minutes – avoid boluses to avoid respiratory depression (benzodiazepines) or marked hypotension (droperidol).
4. Small initial doses because of timidity about sedating young people followed by large doses because of little effect leading to obtundation is to be avoided.

Voluntary/cooperative – oral agents preferred

First line

Hyper-aroused but <u>not</u> psychotic, aggressive or severely agitated: Quetiapine 5 mg kg^{-1}

Psychotic, aggressive or severely agitated: Olanzapine wafer <40 kg–5 mg; >40 kg–10 mg

Involuntary/uncooperative – parenteral agents preferred

Intramuscular injection (IMI) will often be the route of choice, and the agent

should be relatively quick, effective and safe:

Droperidol 0.1–0.2 mg kg^{-1} (max 10 mg) is first line and may be repeated after 15 minutes. Perform an ECG when able to screen for prolonged QT interval and potential arrhythmias.

Midazolam or ketamine is an option which may be used if there is an inadequate response to two doses of droperidol. The patient must be continuously monitored for respiratory depression and other potential complications.

Transfer is a potential escalation of risk

- The leaving – are they stable?
- The transfer – are they monitored with an adequate response to thresholds strategy?
- The arriving – are the recipients prepared?

Conclusion

Emergency psychiatry is treating the underlying neurobehavioural processes not the cognitive content or psychiatric diagnosis. Process psychiatry requires a different mindset. Emergency physicians are ideally trained to adopt that mindset.

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17.3

Autism and behavioural disturbance in the pre-adolescent child

Meenakshi Rattan, and Kenneth Patrick Nunn

ESSENTIALS

1 Clinicians should aim to create calm, quiet, non-pressured mini-contexts for autistic children within the sensory-overloaded, unfamiliar and sometimes frightening environment of the emergency department.

2 Recounting a history of behavioural disturbance in front of a child is likely to escalate behavioural disturbance.

3 Most autistic children do not know why they are upset. ‘Why’ questions are usually unproductive or counterproductive.

4 The non-tactile physical examination of the fully clothed uncooperative child is an essential paediatric skill pending a better available alternative.

5 Medical causes of behavioural disturbance are less common but increasingly important the younger the child, the more severe the intellectual disability, the less his/her speech and the more sensory deficits the child has.

6 Autistic children can be more difficult to restrain and sedate than most adults.

7 Deciding when to proceed to sedation and when to move from voluntary to involuntary sedation must be done calmly and with anticipation, rather than as a consequence of many failed attempts to get control of the child’s behaviour.

8 Long-term management and appropriate community follow-up are of critical importance.

Introduction

The emergency physician is often surprised to find that some of his/her biggest behavioural difficulties are encountered with young autistic children who are presented by others in a state of being ‘out of control’ or aggressive. The tiny number of children who are regarded as ‘unsedatable’ without a general anaesthetic includes many with autistic spectrum disorders. They often require more mg kg of medication than adults to achieve a safe and stable level of sedation.

Autism and the autistic spectrum of disorders (ASD) affect behaviour and experience and include the following:

1. Social and emotional agnosias (disorders of social and emotional awareness and recognition)
2. Social and emotional dysphasias (disorders of social and emotional communication)
3. Emotional and social dyspraxias (disorders of social and emotional coordination of movement).

Children with ASD have trouble making sense of their surroundings and the expectations of others. They have trouble communicating their own condition and why they are feeling what they are feeling. They have trouble implementing problem solving and converting a behavioural request into a coping strategy. The levels of autonomic arousal, both sympathetic and parasympathetic, can be profound, extreme and prolonged. Once hyper-aroused, they often have very little capacity to de-escalate themselves within a crisis and an unfamiliar setting like an emergency department (ED).

This chapter gives a simple structure to prepare for dealing with these children as a matter of routine, communicating professionalism and calm, while excluding serious medical and psychiatric disorders. Importantly, they are likely to return to the ED with repeated presentations if appropriate community follow-up to address the long-term issues of environmental reduction of threat, stimulus management and predictability of nurture are not put in place.

The underlying cause of aggression in autistic children like any other child can be multifactorial as shown in Box 17.3.1.

Underlying mechanisms in austistic spectrum of disorders/neurodevelopmental disorders

Most aggression in young people, especially autistic young people, is fear based. Frustration, especially in face of limit setting, is also common. In the rest, sensory overload or under-stimulation (Fig. 17.3.1) accounts the majority of situations where neither a medical disorder nor a discrete psychiatric syndrome is involved. Many unsuitable home settings, classrooms or residential placements have too much of the wrong sort of stimulation and too little planned activities geared to the developmental profile of the child.

Reducing fear and frustration through communication of what *will* happen over the day, what *is* happening and what *has* happened is a simple and effective preventive. Reducing sensory overload and understimulation are avoided by always having something for the child to do but never too much to do. In the acute setting reassuring that safety is our goal, that they are not in trouble and reducing the number of people dealing with the child will help much more than threatening with a ‘show of force’, saying what ‘they must not do’ and allowing everyone ‘to have go’ at settling the child with an audience looking on.

Approach to assessment

Is sedation necessary?

Try to determine the current state of agitation before determining the cause. Untreated persistent agitation in the absence of even passive cooperation is the most likely indicator that involuntary sedation will be necessary. The risk of injury/violence should be based on known past history, present agitation and whether staff and carers are feeling unsafe, until better information is available.

Physical examination

Mental state examination should assess for physiological arousal, extremes of motor agitation or withdrawal, containment breaches such as absconding or damaging property and cognitive incoherence and fragmentation (Fig. 17.3.2).

Some children will be extremely uncooperative with a full physical examination. However, a rapid but thorough examination is essential. Even in very uncooperative children, much can be gained from careful observation. Note neurological function and general health (pulling at ear, guarding the abdomen, stridor, respiratory distress, abnormal gait, pale or flushed skin).

Box 17.3.1 Differential diagnosis of acute aggression in autistic children

A). General medical conditions

Most common medical causes:

- Chest infection
- Urinary tract infection
- Otitis media and/or otitis externa
- Abdominal pathology, such as constipation, appendicitis or inflammatory bowel disease
- Any other occult cause of pain (dental pathology, fractures, inguino-scrotal conditions).

Less common but very serious:

- Ictal and peri-ictal events
- Head trauma (sometimes self-inflicted head banging, which can cause retinal detachment)
- Encephalitis, meningitis, or other inflammatory encephalopathies
- Acute delirium
- Metabolic derangement (e.g. hypoglycaemia, hyponatraemia, hypocalcaemia, liver or renal failure)
- Hypoxia.

B). Drug induced/withdrawal

Although substance addictions seem to be less common in autistic children, usage still occurs. However, this is more often a problem with adolescents than younger children. More commonly they involve the following:

- Recent commencement of stimulants, SSRIs or antihistamines

- Other drug toxicity, inappropriate ingestion or solvent abuse
- Alcohol, hallucinogens, stimulants (amphetamine-type substances and cocaine), cannabis, synthetic opioids, benzodiazepines

C). Mental health conditions

Most common psychiatric causes:

- Anxiety disorders
- Chronic complex post-traumatic stress disorder due to neglect and maltreatment combined with uncertainties of placement and future plans in the out-of-home-care child
- Agitated depression
- Dysexecutive syndromes – inability to cope with unplanned occurrences, unfamiliar settings and changes to routine
- ADHD – especially in foetal-alcohol-related disorders.

Less common but very serious:

- Mood volatility (emerging bipolar disorder)
- Psychotic disorders (emerging schizophrenia).

D). Others

- Developmental crises associated with puberty
- Psychosocial adjustment – the shift from primary school to high school
- Situational crises such as parents fighting, children teasing and loved ones leaving or dying
- Impulse control disorders – restricting behavioural addictions, such as games use or repetitive behaviour.

History

The history should be taken from a parent or carer who is familiar with the child, with particular emphasis on antecedents to and consequences of any behavioural difficulties (Fig. 17.3.3). It is important not to recount all the child's behaviour difficulties in front of the child. Looking at phone videos of behavioural crises in

the child's presence is humiliating and likely to re-escalate his/her behavioural disturbance. Detail should be sought on current medication plans, behaviour support plans, as well as any communication plans/aids and sensory considerations for the patient.

Challenges in emergency department presentations

- An ED is generally a strange, noisy and unfamiliar environment with the pervasive threat of separation and needles.
- Triage can be inappropriate (time is risk of loss of containment), and waiting times can be long.
- There can be idiosyncratic responses to standard interventions.
- Staff appreciation of the patient's level of functioning and unique ways of communicating can vary greatly.

Approach to management

The management should follow and implement the underlying principles of autism management: early identification of disturbed and/or aggressive behaviour, the use of de-escalation strategies, early effective medication use, and the minimisation of the use of manual and mechanical restraint:

- Effective use of the parent or carer
- Use of communication/visual aids if appropriate
- Team work and collaboration
- Multidisciplinary approach (emergency physician, psychiatrist, allied health, social worker, carer)
- Early recognition of escalation
- Following policies and guidelines
- Medications (avoid oral benzodiazepines due to increased risk of disinhibition)
- There is no acute indication for commencement of stimulants or selective serotonin reuptake inhibitors (SSRIs) – this should be done in the community by those following up the child if it is truly indicated.
- Proactive crisis management plan.

Management

Sedation must be carried out with a definitive plan and clear therapeutic end points. Inadequate planning of sedation can lead to a situation of '*undersedation–oversedation and obtundation*' where small doses of medication are administered, sufficient only to disinhibit any remaining controls the child has, followed by panic prescribing when these doses fail to help leading to obtundation.

Acute (Fig. 17.3.4)

The important components of management of acute behavioural disturbance include the following:

- Safety and containment
- Aggression minimisation strategies
- Verbal de-escalation
- Negotiated voluntary oral sedation
- Restraint
- Involuntary parental sedation
- Continued assessment once control of behaviour is achieved.

Avoid benzodiazepines, as in autistic children there is high risk of disinhibition, and very large doses are usually required to have any sedative effect.

If extrapyramidal side effects occur after administration of antipsychotic medication, administer benztropine 0.02 mg kg (adult dose 1–2 mg) IM or IV. If respiratory depression occurs after benzodiazepines, administer flumazenil 0.02–0.04 mg kg, titrated to respiratory effort.

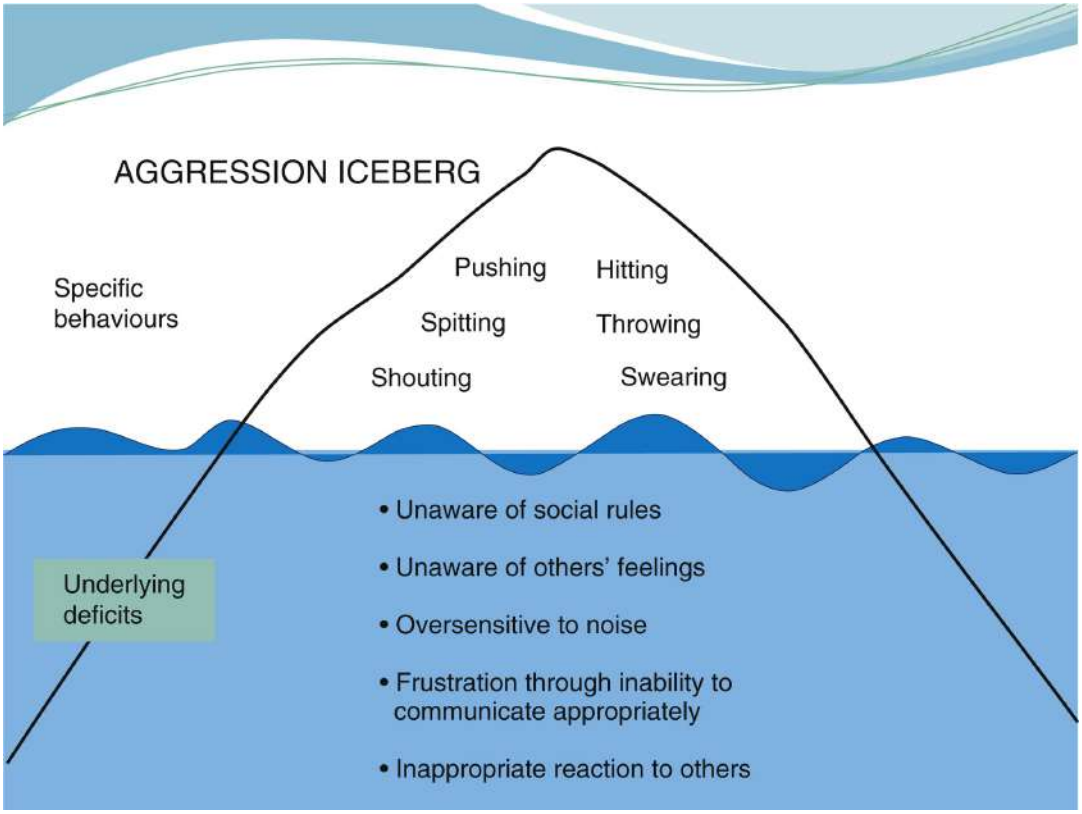

FIG. 17.3.1 Underlying causes of aggression in autism. Adapted from Schopler E. Behavioral priorities for autism and related developmental disorders. In: Schopler E, Mesibov GB, editors. *Behavioral Issues in Autism*. New York: Plenum Press; 1994, p. 55–75.

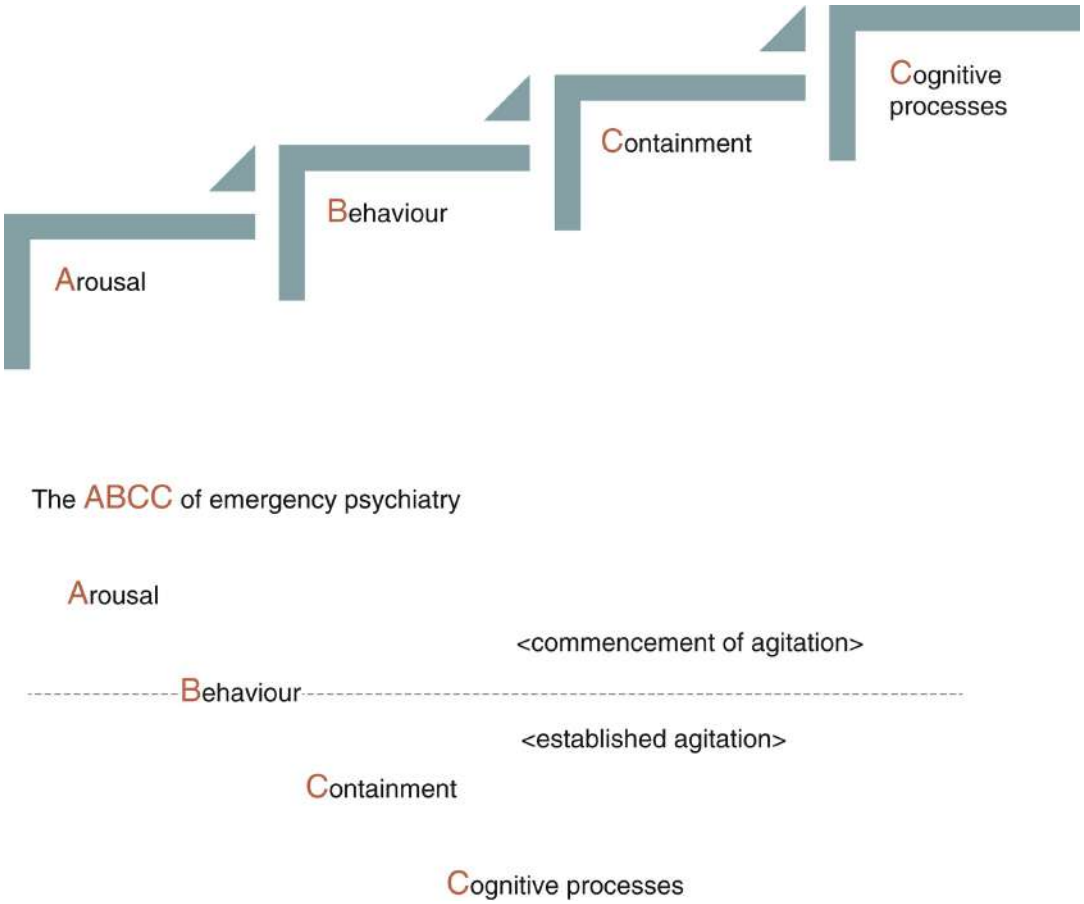

FIG. 17.3.2 ABCC (**A**rousal, **B**ehaviour, **C**ontainment, **C**ognitive processes) of emergency psychiatry.

'The unsedatable child' may very occasionally need a general anaesthetic with supported airway with or without an alpha$_2$ adrenergic agonist infusion and management in ICU for 24–48 hours. Seek specialist child psychiatry advice and involve anaesthetic and intensivist colleagues.

Restraint

- Five-point restraint (one for each limb and one for head); supine position
- Team leader should be at head end, managing airway and monitoring condition.
- Avoid prone restraint.
- Physical restraint should only be used to facilitate appropriate and effective chemical sedation.
- Explain to child and family, and try voluntary holding/cuddling when appropriate and safe.

Monitoring

Level of consciousness monitoring and pupil size monitoring are useful to assess medication response and sedation.

Monitoring of vital signs should occur every 5 minutes for 20 minutes after each dose of parenteral medication, then every 15 minutes for the next 2 hours.

Ongoing management in the community

- Behavioural modification interventions – behavioural support planning/positive behavioural reinforcement
- Applied behavioural analysis (ABA) programmes
- Picture exchange communication system (social understanding/social story)
- Functional/sensory assessment and treatment
- Disability services: Aged Care and Disability, National Disability Insurance Scheme
- ASPECT (Autism Spectrum Australia)
- Private psychologists/psychiatrists
- School counsellors
- Headspace
- CAMHS (Child and Adolescent Mental Health Service).

Conclusion

Triage of an autistic child needs to be prompt and appreciative that the whole emergency department can be locked down to deal with a behavioural containment breach if not addressed expeditiously. Autistic children represent the most common cause of ‘unsedatable’ children in EDs. This is particularly problematic if occurring during retrieval in-flight or on-road transport. General EDs, based on size and age, are prone to underestimate the chaos that is possible with these children.

Autistic children require medical assessment, including physical examination, even if the method of examining is less than ideal. It can be very reassuring to parents and carers when medical factors have been ruled out in children who cannot communicate their needs.

If sustained agitation is present and cooperation is absent, involuntary

sedation needs to occur in an organised and rapid fashion. The sedation team requires a similar level of proficiency and professionalism as one managing a cardiac arrest.

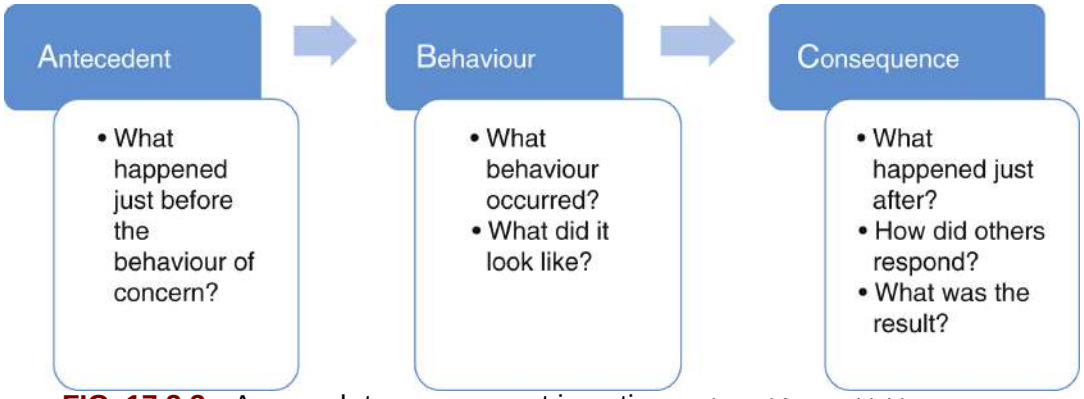

FIG. 17.3.3 Approach to assessment in autism. Adapted from Addabbo L,Bulhak-Paterson D. Behaviour Management Strategies for Individuals with Autism Spectrum Disorders, 2011, Autism Victoria, (http://www.amaze.org.au)

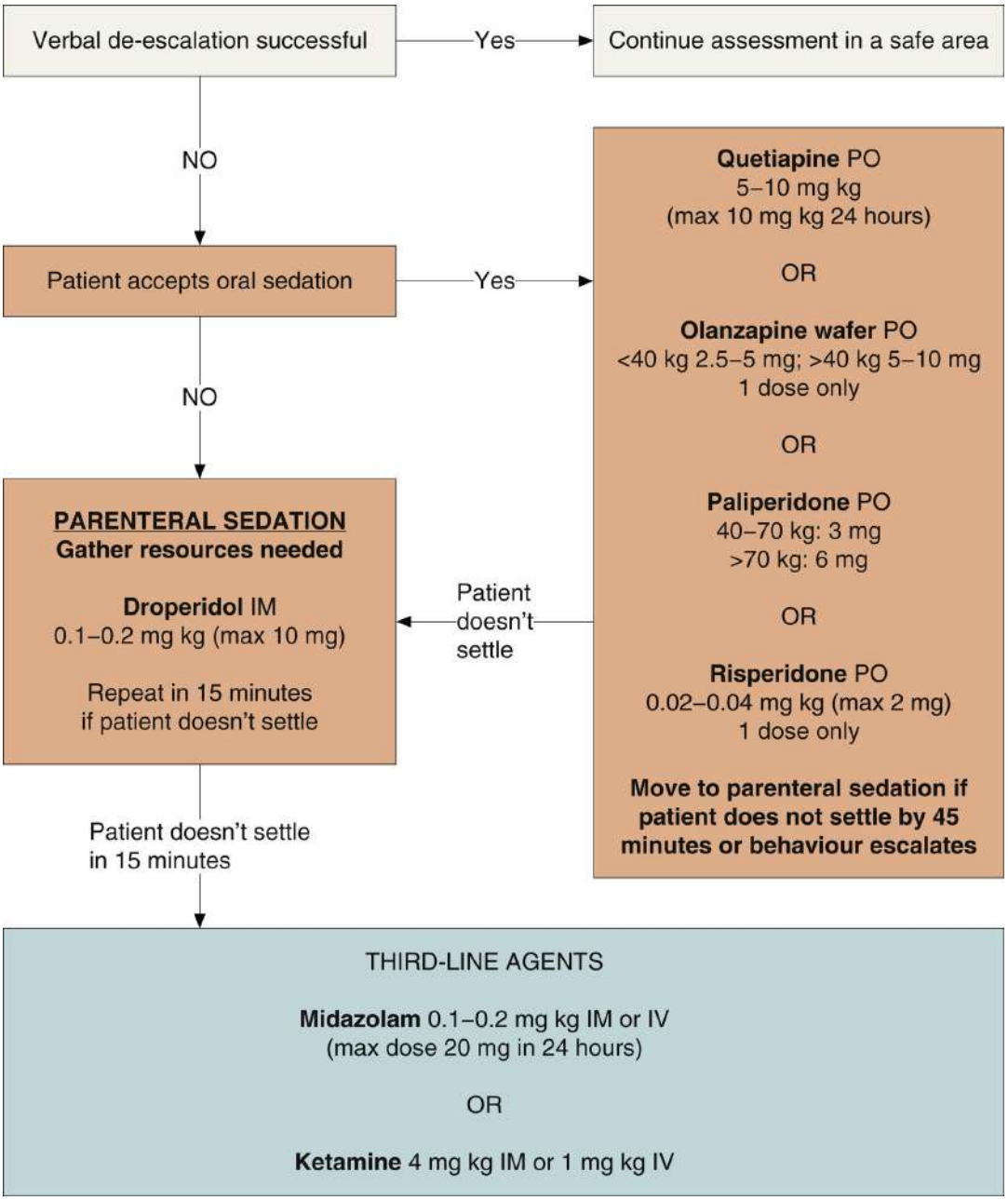

FIG. 17.3.4 Sedation algorithm for children with autism/autistic spectrum disorders and acute severe behavioural disturbance.

It is often the emergency physician/paediatrician who insists on a coherent community response to prevent future unproductive and distressing crises. Adequate and ongoing community follow-up is vital to ensure appropriate care and to reduce the risk of repeated ED visits with behavioural disturbance.

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SECTION 18

Crisis Intervention

OUTLINE

18.1

Sexual assault

Susan Marks

ESSENTIALS

1 Sexual assault occurs when a child is engaged in sexual activity that the child cannot comprehend, for which the child is developmentally unprepared and cannot give consent, and/or that violates the law or social taboos of society.

2 Sexual assault includes a spectrum of activities ranging from rape to physically less intrusive sexual activity.[1,2]

3 Assessment and management of children following alleged or suspected sexual assault is a highly specialised area and requires a multidisciplinary, multiagency team approach.

Introduction

Medical assessment of child sexual assault (CSA) requires a dedicated, well-trained and experienced doctor who is able to spend a significant amount of time making an unhurried and thorough assessment and detailed documentation of history and examination findings. The doctor must have an accurate knowledge of genital anatomy and experience in performing genital examinations. Skills and experience in this field are developed through postgraduate studies, significant case numbers, a knowledge of current literature and involvement in peer-review practices.[3,4]

Inexpert assessment of such cases may have a profound negative influence on the child and family. It may potentially lead to inappropriate removal of the child from the family or wrongful imprisonment[5] or, conversely, to fail to protect a

child from further abuse.

The roles of the emergency physician in this process are:

- recognition of the *possibility* of sexual assault
- emergency treatment of acute physical injury
- consideration of the need for urgent toxicology screen[6]
- referral to the local specialist sexual assault service (SAS) if available
- ensuring that the following services are provided if indicated (these services are usually provided by the specialist sexual assault service):
    - anogenital examination and collection of forensic evidence
    - provision of emergency contraception and/or antibiotic and antiviral prophylaxis
    - provision of appropriate psychological support to the child and family
    - protection of the child and referral to local child protection agencies.

In the majority of cases, determination of whether or not sexual assault has occurred is not possible within the emergency department (ED).

Definitions

CSA is the use of a child for sexual gratification by an adult or significantly older child/adolescent.[7] It may involve a range of activities that vary from exposing the child to sexually explicit materials to anal or vaginal penetration of the child. Central to the definition is that the child cannot provide truly informed consent for sexual activity with adults.

Sexual play between children of similar age does not fit into this description.

The term ‘assault’ is preferred over ‘abuse’ as it highlights the criminal nature of the activity and avoids minimisation of such abusive acts.

Attitudes/myths surrounding child sexual assault

The subject of CSA is an emotive one. Emergency physicians will often have strongly held opinions and attitudes on this subject. These attitudes may be shaped by past experience and/or social taboos. In order to approach CSA in a

calm, non-judgemental and objective manner it is important that emergency physicians are cognisant of their own opinions and emotional responses. In dealing with victims of CSA, expressions of anger, sadness or surprise are not helpful and potentially stigmatising and harmful to the child. With emergency physicians infrequently encountering CSA, it is useful to reflect on the following, sometimes poorly understood, statements:

- A broad range of sexual behaviours has been observed in 'normal' children.
- Most children are not abused by strangers.[8]
- As historians, children are no less reliable than adults.
- CSA is not normally an isolated incident.
- CSA uncommonly produces severe genito-anal injury.[3,9–12]
- CSA often occurs in the context of other family problems, including physical abuse, emotional maltreatment and substance abuse.[13]

Epidemiology of child sexual assault

There has been a significant increase in the recognition of CSA,[3,13,14] which has been reflected by a substantial increase in the number of reports made to child protection services across Australia and overseas.

Sexual assault of children of all ages and both sexes has been documented and is committed predominantly by men, who are commonly members of the child's family, family friends or other trusted adults in positions of authority.[13]

Sexual abuse by family members or acquaintances usually involves multiple episodes over periods ranging from a week to years.

Victims of unknown assailants tend to be older than children who are sexually abused by someone they know and are usually only subjected to a single episode of abuse.

The estimated proportion of children exposed to some form of sexual assault varies depending on the definition of sexual abuse and methodology used. In the United States, literature surveys provide estimates of 9–52% for females and 3–10% for males.[8]

In Australia, between 2010–11 and 2014–15, the rates of child maltreatment have remained steady for all types of abuse and neglect except for emotional abuse, which has increased from 2.2 to 3.4 per 1000 children over this time.[15]

Sexual abuse was the least common form of substantiated harm (or risk of

harm) from child maltreatment for 2014–15 (12.9% of all substantiations). Girls were significantly more likely to be the subject of substantiation cases of sexual abuse (16.7% of all substantiations) compared to boys (9%).[15]

Child sexual assault and emergency medicine

Children who are victims of sexual assault may present to EDs in a variety of circumstances:

1. They may be seen for an unrelated matter when routine history and physical examination produce information where sexual assault forms part of the differential diagnosis.
2. They are brought by a parent or carer to the ED for evaluation of suspected abuse.
3. They are brought to the ED by social services or the police for a medical evaluation for possible sexual abuse as part of an investigation.
4. They are brought to an ED after a suspected acute sexual assault for evaluation, evidence collection and crisis management.

Recognition of child sexual assault

Recognition of the possibility of CSA is dependent on history and examination findings, both of which are normally non-specific.

In the majority of cases, physical examination will neither confirm nor refute an allegation of sexual assault. History from the child remains the single most important diagnostic feature in coming to the conclusion that a child has been sexually abused.[16]

Signs and symptoms

Non-specific

Children who have been sexually assaulted may develop a variety of emotional and physical complaints, often unrelated to the genital area. These include:

- developmentally regressive behaviour
- deterioration in school performance
- sleep disturbances

- abdominal pain
- enuresis, encopresis
- phobias
- sexualised behaviour.

Specific

- Disclosure by child
- Genitoanal injury
- Sexually transmissible infection
- Pregnancy.

Genitoanal injury

Only 4% of all children referred for medical evaluation of sexual abuse have abnormal examinations at the time of evaluation. Even with a history of severe abuse, such as vaginal or anal penetration, the rate of abnormal medical findings is only 5.5%.[16]

The physical examination of sexually abused children should not result in additional emotional trauma.

When the alleged sexual abuse has occurred within 5 days or there is bleeding or acute injury forensic examination should be performed within an appropriate time frame by the designated sexual assault service medical officer. In this situation, protocols for CSA victims should be followed to secure biological trace evidence such as epithelial cells, semen, and blood, as well as to maintain a ‘chain of evidence’. When more than 5 days have passed and no acute injuries are present, an emergency examination usually is not necessary. An evaluation, therefore, should be scheduled at the earliest convenient time for the child, physician and investigative team.

In the child presenting with genitoanal injury or abnormality, CSA is only one of a number of diagnoses that should be considered. The differential diagnosis of genitoanal injury includes:

- accidental injury
- falls astride
- sexual assault
- medical/dermatological condition, e.g. lichen sclerosis/drug reaction.

Genital findings in children are difficult to interpret. Such interpretation is generally beyond the expertise of most emergency physicians[17]. Whilst acute trauma may be easily recognised, interpretation of such findings may be problematic for the occasional examiner.[17]

Genitoanal anatomy

Knowledge of what constitutes normal and abnormal anatomy has evolved over recent years. This has been driven partly by several highly publicised cases where misinterpretation of normal findings led to inappropriate separation of children from parents and wrongful conviction.

Hymen

There is considerable variation in the shape of the hymen. In the prepubertal girl it can be thin and relatively inelastic. In this age group, blunt penetrating trauma to the vagina may result in tearing of the hymen. Such tears when healed may manifest as a notch or defect in the hymenal tissue. It is also possible to have penetrating injury to the hymen without any abnormal findings on examination.

As an oestrogen-dependent/responsive tissue, at puberty the hymen becomes thick, irregular and elastic and distensible. It is less likely to sustain injury during penetration than in the prepubertal state.

Sexually transmitted infections

The diagnosis of a sexually transmitted infection in a child warrants a thorough assessment by a specialised sexual assault service.[18]

Diagnostic considerations

The diagnosis of CSA can often be made based on a child's history. Physical examination is infrequently diagnostic in the absence of a history and/or specific laboratory findings. Physical findings are often absent even when the perpetrator admits to penetration of the child's genitalia. Many types of abuse leave no physical evidence, and mucosal injuries often heal rapidly.

On examination, findings which are suggestive, but not diagnostic, of CSA include injuries indicative of acute or healed trauma to the genital/anal tissues. The American Academy of Pediatrics regularly updates their guidelines for the

medical assessment and care of children who may have been sexually abused including a consensus approach to Interpretation of Medical Findings in Suspected Child Sexual Abuse.[19]

Many cases of alleged sexual abuse involve parents who are in the process of separation or divorce and who allege that their child is being sexually abused by the other parent during custodial visits. Although these cases are generally more time consuming, they should not be dismissed because a custody dispute exists. Allegations of abuse that occur in the context of divorce proceedings should be reported to the child protective services agency.

Role of the emergency physician

The emergency physician (EP) should ensure that any physical injuries are detected, accurately documented and correctly treated. In most jurisdictions, medical assessment of CSA will be performed by a specialist sexual assault service.

Medical issues, such as sexually transmitted diseases and emergency contraception, should be discussed and managed.

The EP should collect, or provide opportunity for collection of, forensic specimens, ensure appropriate psychological support is provided to the child and family, and report the case to protective agencies within the legislation of the local jurisdiction.

Documentation

Because the likelihood of civil or criminal court action is high, detailed records and/or drawings should be kept.

Mandatory reporting legislation

Some form of mandatory reporting legislation exists in all Australian jurisdictions. Although this legislation varies from state to state, the basic principles are similar. Doctors are mandated to report cases where there is reasonable suspicion that CSA will occur or is occurring. The reporting practitioner has statutory protection from prosecution if a report is made in good faith.

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18.2

Child at risk

Susan Marks

ESSENTIALS

1 Concerns of child abuse or neglect arise when an adult responsible for the care of the child either harms the child or fails to protect the child from harm.

2 A child may be at risk from physical abuse, sexual abuse, emotional abuse or neglect.

3 Medical and nursing practitioners have a responsibility to protect children at risk and are often mandated by law to report suspicions that a child is at risk of significant harm

Introduction

Child abuse is increasingly recognised as a major public health and social welfare problem with important short-term and long-lasting effects for children and adolescents. As Australian and New Zealand emergency departments (EDs) provide care for many hundreds of thousands of children and adolescents each year, departments and their staff play an important role in the detection of abuse and initiation of a medical and community response. This response is aimed primarily at treating the child, minimising psychological effects and ensuring their safety.

Children and adolescents are, by virtue of their developing intellectual, emotional and physical state, a vulnerable group. The environment within which they develop is influenced by many factors outside their control: the economic and social status of their family, the personality and values of family members

and friends, and the extent of physical and intellectual stimulation that they receive may all have profound influence upon their development. The potential variability in these factors and the recognition that negative experiences often have serious short- and long-term implications for the child have led to a general acknowledgment that children and adolescents need protection.

The United Nations Convention on the Rights of the Child recognises that:

> *... for the full and harmonious development of his or her personality, [a child] should grow up in a family environment, in an atmosphere of happiness, love and understanding ....*

The Convention continues, stating in Article 19 that governments shall:

> *... take all appropriate legislative, administrative, social and educational measures to protect the child from all forms of physical or mental violence, injury or abuse, neglect or negligent treatment, maltreatment or exploitation, including sexual abuse, while in the care of parent(s), legal guardian(s) or any other person who has the care of the child ....*

It is this philosophy that has driven the creation of the social and legal framework of child protection. Doctors, nurses and other healthcare workers who deal with children are an integral part of this system that acts to protect children and adolescents. Every health professional that has contact with children needs to be aware of the possibility of child abuse, must be able to detect when it is occurring and know how to act in the best interests of the child once it is suspected.

Definition

The child at risk is not a medical diagnosis but rather a description of certain forms of behaviour displayed by adults responsible for the care of a child. The child is at risk when an adult responsible for the care of the child harms, threatens to harm or fails to protect the child from harm. Harm may be physical (e.g. inflicting an injury, causing pain or poisoning), psychological (e.g. causing feelings of being unloved or worthless; exposure to domestic violence) or neglectful (e.g. failure to meet the child's basic needs and/or medical needs, lack of adequate supervision). A child may be at risk of harm from:

- physical abuse
- sexual abuse
- emotional abuse
- neglect.

These are not exclusive, and a child may be subjected to more than one type of abuse.

Physical abuse

Many thousands of children present to EDs around Australia each year with a wide range of physical injuries, the vast majority of which are caused accidentally. It is a difficult but essential task to identify children within this group who have been injured as a consequence of abuse. While medical and nursing staff must be alert for the possibility of an inflicted injury, there are specific circumstances that may raise suspicion. These include:

- situations where there are direct allegations of violence directed against the child made by the child or any other person
- the type and pattern of injury observed at the examination
- an explanation being offered for an injury that does not fit the type or pattern of injury or does not fit with the developmental ability of the child
- delayed presentation for medical care with an injury that a reasonable person would have recognised as needing care sooner
- multiple presentations with injuries, often to different healthcare providers, seeking medical attention.

Once the possibility of inflicted injury has been raised the priorities of the treating doctor are:

1. to diagnose, treat and document the child’s injuries
2. to interpret the pattern of injury or behaviour as to the possible causes
3. to notify and involve the agency responsible for ensuring safety of the child
4. to provide a written or verbal report and advice to that agency or the police.

Presentation

Physical abuse may present to the ED in many different ways. Most commonly it will present as a child with an obvious injury and a suggestive history, but in some situations it will be more subtle, such as a younger child who presents not using a limb.

Young children who present with symptoms such as vomiting, irritability or a decreased conscious state with no obvious cause may have a head injury from a blow or a fall or a ‘shaking’ injury or may have been poisoned.

History

It is necessary to collect as much information as possible on the events that led to the child sustaining the injuries. Specifically, enquire as to when, where and how the injury happened, who was present at the time and what happened after the injury. The child’s medical, developmental and social history, with specific information on past injuries, is important.

This history must be sought in an open and non-judgemental fashion, which encourages the participants to reveal all the important information. Unfortunately, the ED is often not the ideal place to conduct a lengthy and in-depth forensic interview with the parents or carers, and it may be prudent to limit the information gathering to items that will enable specific issues to be addressed. The interview can always be completed by trained investigators at a later time.

Physical examination

Prior to commencing the examination the doctor must ensure that the parents and, where appropriate, the child are informed of the nature and extent of the examination and that valid consent has been given. In addition it is ideal to have spent some time with the child, to gain his/her confidence and thus increase the chances of keeping his/her cooperation during the examination.

Consent from the parent or legal guardian is necessary to conduct a physical examination, to perform investigations (including photographs) and to release clinical information in the form of a report to a third party. If consent is refused the protective agency or police must seek a court order. If there is an urgent medical problem that needs intervention and such intervention is clearly in the best interests of the child, then the examination and treatment should proceed and not be delayed by the lack of consent.

An adolescent may be able to give consent for his/her own examination as long as he/she is capable of understanding what the examination entails, what the results will be used for and the implications that this may have for him/her.

A thorough physical examination of the child should be performed, with observation and palpation of skin, soft tissues, bones and joints and giving specific attention to the eyes, ears and mouth. The examination should look for the following physical findings:

Bruising of the skin

Bruises are extremely common in children. In the absence of a documented bleeding tendency they are evidence of blunt trauma and may provide some information on the site, the implement or force of an impact. Accidental bruises are commonly found in children once they have learnt to crawl, occurring over bony prominences, usually on the front of the body and are directly related to a child's increasing motor activity. Babies who are not yet crawling rarely have accidental bruising.

Bruises caused by abuse may occur anywhere on a child's body. Specifically, look in places where accidental bruises are uncommon such as the mouth, behind the ears, on the inner aspect of the upper arm and around the buttocks. Observe the shape and pattern of bruises, looking for features that may suggest a blow from an open hand or single or multiple blows from an implement. Look for a pattern within the bruise that may suggest contact with a specific surface.

Whilst it is important to describe the appearance and colour of the bruising, it is not possible to be accurate about the age of a bruise. If a bruise is yellowing in colour then it is likely to be more than 18 hours old.

Laceration and abrasion of the skin

Lacerations are the tearing of tissues caused by a blow from a blunt object. Abrasions are the disruption of the outer layers of the skin caused when the skin contacts a surface at an angle. They occur at or close to the site of impact and may occur after a blow with an object or after a fall on to a surface. They are frequently associated with bruising.

Examine the wound for neurovascular and tendon injury and for foreign bodies. The presence of foreign material such as glass, dirt or gravel should be noted as they may be important in evaluating the injury.

Burns or scalds

The appearance of a burn on a child is influenced by many factors: the temperature, size and shape of the causative agent, the depth of skin at the contact site, the length of time of contact and the application of first-aid measures all have the potential to modify its characteristics. Whilst information on all of these factors should be sought from the caregivers it is often difficult to draw accurate conclusions from examination of the wound itself. Associated injuries such as bruises or fractures may help. Although classically described as associated with inflicted injury, cigarette burns are uncommon. These appear as small, deep, round burns, usually on the limbs or back.

Healing burns can sometimes be especially difficult to diagnose and interpret. Inflammation can extend beyond the margins of the burn, obscuring the shape and increasing the size of the lesion. The healing area may become flaking or exudative, causing confusion with skin conditions such as impetigo.

Scalds are a common form of accidental injury in infants, often caused by a hot liquid being tipped over the upper torso, arms or hands: some features of a scald may suggest an intentional cause. Look at the position, shape and depth of the burn. Circumferential scalds of the hands or feet may be caused by forced immersion. Small round bruises above the burn may represent forcible gripping by a hand. Scalds of the buttocks extending on to the lower back or upper thighs with sparing of the natal cleft may indicate the child has been lowered into hot water.

Fractures

Fractures of long bones, ribs and skull may occur when a child is intentionally struck, pushed, squeezed or dropped. While any fracture may be caused by inflicted injury, certain fractures have an association with abuse that should alert the ED clinician and prompt further action. Specifically, fractures in children under the age of 18 months, rib fractures, metaphyseal fractures, multiple fractures and fractures of differing ages should be carefully evaluated.

A bone scan and skeletal survey may be extremely useful in gathering evidence of multiple bony injuries when a young child, typically under the age of 2 years, presents with suspicious bruising or other features of abuse.

Eye injuries

Direct blows to the face may cause subconjunctival haemorrhages; intraocular injuries such as retinal haemorrhages may occur as a result of shaking injury. A careful eye examination including visual acuity and fundoscopy may be

necessary.

Ear injuries

Injuries such as blows to the side of the face or pinching may cause bruising of and behind the pinna. The eardrum may rupture due to the air pressure changes.

Head injuries

Head injuries are a major source of mortality and morbidity in inflicted injury. Young children have a large head-to-body size ratio, relatively weak neck musculature and compliant skull bones that predispose them to intracranial injuries. The child may be struck, dropped, thrown or shaken, producing an open or closed head wound.

Skull fractures, cerebral contusion, intracerebral haemorrhages, extradural haematoma and subdural haematoma are all possible sequelae.

Intra-abdominal injury

Blows to the abdomen may cause laceration or rupture of either solid abdominal organs, such as the liver or spleen, or of the hollow organs, such as the duodenum. There may or may not be accompanying bruising of the abdominal skin to alert you to this possibility. Liver function tests and lipase may be helpful screening investigations.

Investigations

Investigations are done for a number of reasons, including the following:

- Further assessment of the presenting injuries:
    - Primarily, the findings of the clinical examination dictate the extent and type of investigations necessary. As would be the case in the investigation of any injured child, plain X-rays, CT scans, MRI, ultrasound and other imaging should be directed at areas where there is clinical suspicion of injury.
- Assessment for possible occult injuries:
    - Other investigations, such as a skeletal survey and bone scan, are used in an attempt to detect injuries that may not be clinically apparent but which will assist in establishing the likelihood of inflicted injury. These are especially useful in children under the age of 2.

        - A urine drug screen may be indicated.
    - Investigation for underlying disease:
        - In a child with multiple bruises, the possibility of a bleeding disorder should be considered. A full blood examination and coagulation profile may be necessary in these circumstances.
        - Children with multiple fractures may need screening for underlying bone disease.

Emotional abuse

This is the commonest type of abuse that is reported to child protection agencies. Emotional or psychological abuse is difficult to define and even harder to detect, particularly in the ED, when frequently the child presents to a particular health worker on a single occasion. Five possible components of emotional abuse have been described. These are the following behaviours:

- Rejecting
- Isolating
- Terrorising
- Ignoring
- Corrupting.

The incidence of emotional abuse is unknown, but it is likely to be common and underdiagnosed.

History and examination

Presentation is subtle and depends upon the age of the child. An infant may present with sleep or feeding problems, irritability or apathy. Older children may present with attention deficit, attention seeking, aggression, school failure, truancy, anxiety, depression and psychosomatic disease. It is likely that emotional abuse accompanies other forms of abuse, such as physical and sexual abuse. Consideration of its possible role is important in all assessments of a child at risk.

Detection by all health workers looking after children, including emergency staff, requires a high index of suspicion and vigilance. Diagnosis is suggested by the consequences in the child, as above.

Assessment should include that of behavioural, emotional and physical signs

and the child–parent interaction. This will usually require time not available to emergency staff, and therefore, when emotional abuse is suspected, referral is necessary. This may be to the hospital child protection unit, to other health services for further assessment and/or to community social services.

Neglect

Neglect may also cause the child to be at risk and can be difficult to define and diagnose. A broad definition is anything that individuals, institutions or processes fail to do, which directly or indirectly harms children or damages their prospects of a safe and healthy development into adulthood. Other definitions have tended to be narrower and therefore target more severe or persistent neglect. It is important to differentiate neglect from poverty or ignorance, as these will require a different intervention.

The true incidence of neglect is unknown, and it is probably the most common reason for the child to be at risk. Its diagnosis usually only occurs when harm has occurred, but consideration should be made of potential for harm and long-term effects. Types of neglect include medical neglect, safety neglect, educational neglect, physical neglect and emotional neglect. Non-organic failure to thrive is likely to be due to a combination of a lack of calories and affection.

History and examination

Some possible features to look out for are frequent presentation and admission to hospital with accidents or illness, delay or failure to access health care, malnutrition, failure to immunise, poor physical presentation, poor compliance, behaviour disorders, developmental delay and, very importantly, failure to thrive. The assessment of growth and development is clearly an essential part of child health assessment, even in the emergency setting. Measurement of height, weight and head circumference and their plotting on standardised growth charts can be very useful in assessment and follow-up. Emotional and behavioural assessment is difficult in the emergency setting.

Treatment

The diagnosis of non-organic failure to thrive usually requires admission to hospital to assess the child’s ability to grow with adequate nutrition and an interdisciplinary approach. Other forms of neglect may require admission or, if not, referral to child protection/abuse unit or community social services.

Medical child abuse

Medical child abuse, previously known as Münchausen's syndrome by proxy (MSBP) and/or factitious illness, is an unusual presentation of the child at risk that may present to an ED. This condition occurs when an adult caregiver, usually the mother, presents a false history to the physician regarding a child. This history may cause the emergency physician to perform unnecessary diagnostic and therapeutic procedures that do not result in any specific diagnosis.

In the past, the focus of diagnosis was on an underlying psychiatric diagnosis for the caregiver. Medical child abuse is a diagnosis focused on what is happening for the child. The presentation may involve fabricated or induced symptoms or signs.

Children at risk for medical child abuse are typically aged 15 months to 6 years. The emergency physician is often confronted with baffling symptoms. Frequently, the child has been taken to many care providers before the diagnosis is finally established. Warning signs that are suggestive of medical child abuse include the following:

- Illness is multisystemic, prolonged, unusual or rare
- Symptoms are inappropriate or incongruent
- Symptoms disappear when parent or caregiver is absent
- General health of patient clashes with results of laboratory tests
- Symptoms (e.g. seizure activity) are unresponsive to conventional medical treatment and are witnessed only by parent or caregiver.

The community response to the child at risk

Responsibilities to report

For medical practitioners working in acute medicine, often the first point of contact with a child at risk is when they present for treatment. Mandatory reporting legislation brought in across all states of Australia has made it compulsory to report cases to the State Child Protection Agency where there are concerns of risk of significant harm due to suspected inflicted injury or neglect. ED staff should familiarise themselves with the mandatory reporting requirements and processes in their jurisdiction. Once notification is made a process of risk management is commenced.

What to do as the medical practitioner, in suspected cases

1. Take a concise history of events and physical examination.
2. Document injuries, and order appropriate investigations.
3. Manage injuries, as usual practice.
4. Notify relevant child protection agency. (Client's consent is not required in child protection notification.)
5. This may be facilitated by social worker, nurses and paediatric specialists.
6. Arrange appropriate ongoing medical care for the child. This may require hospital admission for further investigations and/or for child protection reasons (to ensure the child's safety and wellbeing).

Legal responsibilities

1. By law, health services staff must provide all relevant information that they have available when asked (in writing) to do so by the child protection authority.
2. Staff do not have to get permission of the client in order to forward the relevant information.
3. Protection for notifier. Typically there will be legislation safeguarding the identity of the person who makes the report.
4. Neither the report nor its contents are admissible as evidence in any proceedings against the person who made the report.
5. If, as a result of making a report, a person is threatened or fears personal violence, this should be reported to the police, who may apply for, and pursue on his/her behalf, an apprehended violence order.

What happens after notification?

1. The Child Protection Agency determines their response when someone reports that he/she thinks a child or young person under the age of 16 years has been, or is being, injured or neglected.
2. When the agency receives information about suspected inflicted injury, it makes decisions about how to go ahead with investigating the claims and how others may be able to help. For example, it may contact the

child's teacher, child-care worker, relatives or the police.
3. The police will be contacted and may become involved if the agency thinks the law has been broken.
4. Health professionals should ensure that appropriate heath care plans are in place.

Actions based on risk assessment

1. If the child is in immediate danger, steps will be taken to reduce the level of risk or move the child to a safe place. This may mean admission to hospital or foster care.
2. In many cases, this might mean giving the family practical help, such as organising child care, emergency finance, providing a referral for counselling or information on health or other services.
3. In some cases, the child protection agency takes the matter to the Children's Court. The court can order a child be placed in agency care for a period of time. The court can also order counselling and other types of support services including health services.

Further reading

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SECTION 19

Administration In EMS

OUTLINE

19.1

Managing the death of a child in the emergency department

Bereavement issues

Ioannis Pegiazoglou

ESSENTIALS

1 The death of a child under any circumstances is likely to lead to a significant crisis and grief response in parents.

2 Emergency physicians should be prepared for parental presence in the resuscitation room, anticipate their high level of distress, and ensure that they are kept informed.

3 It is important that the family knows that everything that could have been done was done.

4 Parental questions should be answered honestly and directly, allowing humanity and empathy to show.

5 Personal, compassionate and individualised support should be provided for families, respecting their cultural, religious and social values.

6 Family members are likely to have impaired decision making and communication abilities, and this needs to be taken into consideration around informed consent issues.

7 The needs of the grieving family must be balanced with the legislative requirements of the Coroner's Act when this is relevant.

8 Relatives should be allowed to spend time with the deceased child if

they want to, preferably in a quiet suite.

9 It is important to be available in the weeks following the death to clarify and answer any further questions from the family.

10 Team members need to be aware of their own likely emotional responses to the death of a child.

Introduction

Deaths occurring in the emergency department (ED) present unique challenges for the clinician, particularly if the patient is a child.[1,2]

The unexpected death of a child undoubtedly brings about the most severe and shattering grief response for the child's parents.[3] Because the loss is unexpected and involves someone so young and so intrinsically a part of self, the grief response of parents may be very painful and prolonged. The death of a child must be viewed as a tragedy for the entire continuum of family and friends. Additionally, paediatric deaths are frequently personalised by ED staff and hence have broad implications for the whole ED clinical team.

In large hospital EDs, particularly in urban areas, there is rarely a pre-existing relationship between the health professionals and the patient/family. While this facilitates the professional detachment needed for ED staff to function effectively, it creates inherent voids in the ability to support grieving relatives and friends. In smaller hospitals like those found in rural and regional communities, there may be a pre-existing relationship, potentially lowering communication barriers but bringing out other stresses and strains for ED staff.

Good communication with family members must be established early and maintained throughout. This is best left to an experienced member of the staff. There is evidence to suggest that junior medical staff do not feel adequately trained in talking with parents in regards to end-of-life care matters.[4] Furthermore, in an AAP (American Academy of Pediatrics) statement published in 2008 it was noted that 'health care communication is currently learned primarily through trial and error'.[5] Simulating scenarios of difficult discussions in paediatric emergency medicine can help the ED providers to develop specific strategies when managing those challenging events.[6] Due consideration for the comfort of the family should be at the forefront of the minds of clinical staff at all times.

Box 19.1.1 Essential components of care in the emergency department when a child dies

Clinical

Resuscitation best practice
Termination of resuscitation:
    Identifying, validating, and respecting advanced care directives

Operational

Staff training in communication
Team response (including readily available support staff such as security, child life, chaplaincy, social work)
Family presence policy
Dealing with media
Communication with medical home
Defusing/debriefing for team:
    Private location for family to be with deceased, means and location to conduct rituals

Legal and forensic

Organ donation
Autopsy
Working with police and coroner/medical examiner
Child protective services
Child fatality review team
Documentation in medical record
Preservation of evidence

Ethical

Resuscitation: how long is too long?
Prolongation of resuscitation efforts for family presence/organ donation
Practice on newly deceased

Initiation of resuscitation at the border of viability in extreme preterm birth

Spiritual and emotional

Needs of family, including saying goodbye, memory making
Needs of multidisciplinary team
Envisioning a 'good death' in the emergency department

Follow-up care for family

Helping family to know everything was done
Assisting family in explaining to siblings, family, friends
Assisting family in locating community support to address grief and bereavement
Plan for post-autopsy meeting to answer questions
Plan for scheduled follow-ups and marking of meaningful dates

Follow-up care for team

Scheduled voluntary defusing/debriefing with all members of the emergency care team who wish to participate

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Adapted from Death of a Child in the Emergency Department, Joint Statement of American Academy of Pediatrics Committee on Pediatric Emergency Medicine; American College of Emergency Physicians Pediatric Emergency Medicine Committee; Emergency Nurses Association Pediatric Committee. *Pediatrics* 2014;**134**(1):e313–30.

In summary, essential components of care in the ED when a child dies include clinical, operational, legal, ethical and spiritual layers (Box 19.1.1).[7]

The resuscitation process

Parents usually benefit from being present during the resuscitation process.[8] It is therefore unacceptable to discourage their presence unless they are interfering with, and compromising, the resuscitation itself. Family members watching monitors and seeing the trace 'go flat' experience much alarm and distress, but this should not be seen as a reason to exclude them.[9] In a study published in

2006, Mangurten et al. reported that 95% of the families they surveyed would again wish to be present during the resuscitation process and felt that it had been helpful to them, and no disruption of care was documented.[7,10] In a similar study examining pediatric trauma resuscitation efforts, there also was no difference in time to milestones of care in trauma patients with or without family members present.[7,11]

The resuscitation process can be traumatic for parents and family members, requiring ongoing communication and interpretation of events. It should be expected that parents will be visibly upset and distressed during this period. A staff member, often a social worker, should be assigned to support the family, to answer any questions about the procedures and responses, and to prevent distraught family members from impeding the resuscitation.[12] The ED medical officer in charge must communicate with this staff member and family members about the progress of the resuscitation. Viewing the resuscitation efforts allows the family to see a caring and competent staff, in control of their emotions, doing their best to save the child's life.

Where parents choose not, or feel unable, to be in the resuscitation room, it is essential that they be kept informed of progress. Panic, fear and a sense of isolation have been noted as the main responses of relatives who remain outside the resuscitation room.[9] Small, dull rooms with no windows or natural light were seen as heightening the sense of isolation, disconnectedness and fear for those family members unable to bring themselves to view the resuscitation.

It is important to be skilled in early recognition of the signs of trauma responses by parents, such as dissociation, as this can affect long-term adjustment. A social worker or other designated professional should ideally be available to provide support for parents and act as an advocate during what is likely to be an overwhelming and bewildering process. The social worker is also likely to be the main staff member to have an ongoing role after death has occurred and the family has left the hospital.

Talking to parents and families

When talking with the family about the child's deteriorating condition, give details in a simple, straightforward and accurate manner. Provide the information using appropriate language. Answer questions and be responsive to needs and concerns.

When death has occurred, or is imminent, it is essential to have identified the

relevant family members so that discussions are with the appropriate individuals. At the point of death, the medical officer in charge of the resuscitation should advise those family members present in the resuscitation room or in a private, quiet location. Research has indicated that families appreciated a high level of physician involvement.[13]

Clear, distinct and accurate information is essential, and medical jargon should be avoided. It is very important to state initially that the child has died. This is the piece of information that the parents will most want clarified. It is then desirable to provide a brief chronology of events, while reassuring the family that everything was done and that the child did not suffer pain.

Sometimes family members are not present at the time of death. If practicable it is best to delay notification of death until it can be done in person.[14] On the other hand, strict adherence to the goal of family presence at time of death pronouncement may result in the prolongation of otherwise futile resuscitative efforts. An alternative may be to designate a family surrogate, a staff member whose job is simply to be with the child, so when family members do arrive after their child has died, they can be assured that their child was not alone at the time of death.[7]

If the family cannot readily access the ED, telephone notification may be necessary. A survey of survivors suggested that if delay in personal notification was greater than 1 hour, telephone notification may be appropriate.[15] However, it is obviously difficult to be sensitive to the family's response via a telephone, and there may be limited ability to provide immediate support. Ensure that the family is safe to transport themselves and that ongoing support options have been explored for those family members unable to make it to hospital.

If family members were not present at the hospital it is likely that they will have many questions related to the process, potential suffering and any awareness by the child of the event. These may be asked either over the telephone or upon arrival. If parents arrive 'too late', this can create a further burden of guilt because they were not present.

Family members experiencing significant grief are likely to struggle with the integration of the information that they are being given and with the communication of any questions that they might have. They may need to revisit the same questions and information repeatedly in order to try to make sense of the event.[9]

It is important to allow parents and family members time to examine the implications of the loss and to begin the process of searching for some answers

and meaning in the midst of the event. It is also important to assist them to mobilise resources from their social, cultural and religious communities to help them to deal with their grief.

There can be a temptation to offer sedation to grief-stricken parents. This is often requested by relatives distressed by observing the parents' pain. Grief is a normal process, which is rarely helped by pharmacological intervention.

Junior medical staff are often involved in resuscitations, and it is essential that they have received some training/education to help them handle the unexpected death of a child. A number of programmes have been described, which have been found to be useful in preparing staff to deal with loss in an effective manner, from the perspective of both the family and staff members.[16–19]

Laying out of the child

Where parents want to 'view' or spend time with their deceased child, it is important to facilitate their wishes (having due regard for the possibility that the death may need to be referred to the coroner and hence care not to interfere with evidence). All tubes inserted during the resuscitation process (endotracheal tubes, intravenous cannulae, drains, etc.) should be removed, unless the medical officer in charge considers that the placement of a tube may have been associated with an adverse event. All wounds and cannula sites should be dressed to avoid leakage of bodily fluids. The child's face and exposed areas should be bathed/cleaned and any soiling removed.

The impact of the death can often cause an overwhelming sense of numbness and helplessness, diminishing the ability to self-advocate. Therefore it is important to be proactive with family members and ask how much they want to be involved with the bathing and laying out of the child and about any specific cultural or religious practices that they would like observed.

It can often be useful to obtain mementos of the child. Photographs, a lock of hair, or a foot/hand print may become important mementos along the grieving journey. It is recommended that hospital EDs have access to such items as a camera, memento books and bereavement packs to give to families.

There are specific requirements in place for deaths that must be referred to the coroner. These may limit the process of 'laying out' the body and require that family members may not be left unsupervised with the child. ED staff need to balance the needs of grieving family members with their legal responsibilities to the coroner.

Viewing the body – quiet suite

Most available evidence strongly suggests that seeing the body of the deceased is an important part of accepting the reality of death.[20,21] This includes not only seeing but also being able to touch and hold the loved one. It is helpful to describe to relatives what they are going to see prior to viewing the body, especially if there are trauma-related injuries.[9] Viewing the body can also relieve anxieties about mutilation, signs of trauma, or that the person was in pain when he/she died.[21] A parent or family member's preference not to spend time with the child should also be respected.

Most large paediatric hospitals have a 'quiet suite' or 'family room' to facilitate parents spending time with their deceased child. This can allow a private 'goodbye' and time to reflect. It can also allow time to create an image of the child as dead, altered from the image of the living child.[21] The importance of the family/relatives' room cannot be overemphasised – privacy and basic facilities are essential.

Subsequently, additional relatives/friends may arrive at the hospital. This can often lead to heightened distress for parents as they try to explain the events that have led to the child's death, hence taking them back to the initial traumatic stages. It can also be a useful process. By reviewing events, parents may build a clearer picture and 'fill in the blanks' as they retell their story.

The grief response

Grief is a normal reaction accompanying death. The severity of the grief response parallels the severity of the loss.

Perhaps the most well-known model of describing the process of grief is the 'stages' model with its clearly defined stages of shock, denial and isolation, anger and envy, bargaining, depression and acceptance.[22] These stages should not be seen as linear or rigid. Individuals can move back and forth between the stages or may appear 'stuck' in a stage. Although the 'stages' model is the most well known and can be a useful guide, there are a number of other models of grieving including psychodynamic,[23] attachment,[24,25] social constructionist,[26] cognitive/behavioural,[27,28] and personal construct.[29] Good practice requires being open and flexible and adapting to the needs of the grieving family as opposed to trying to fit the family into any particular model. It is important not to pathologise individuals whose grief response does not fit neatly into a particular

model of grief.[30]

The death of a child provokes the most intense form of bereavement. It is certain to alter the course of the parents' lives and their relationship with each other and with others. Losing a child is more than losing a relationship. For a parent it is losing part of his/her self, his/her present and his/her future. Many parents experience a loss of meaning in their lives and may never fully recover from the impact of their child's death.[25] A child's death is not a singular loss but produces a ripple effect overwhelming all aspects of the family and environment. Parents, and even the extended family, may feel that they have failed, irrespective of the nature of the death and level of love, nurturing and caring that existed during the child's life.[21,31,32]

The parental relationship faces severe stress following the death of a child. It can pull a dysfunctional relationship further apart or glue a functional one closer together. Adverse impacts on the relationship can occur through the real or perceived apportioning of blame by one parent to the other. This can occur where a child died while under the specific supervision of one parent or one parent was simply not present when a critical event occurred.

Siblings of the deceased child will also experience a significant grief reaction. Not only must they manage the actual loss of their deceased sibling, but they must also cope with the loss of their normal family environment. Their parents will be struggling to cope with their own grief and thus will be less emotionally available. The cognitive developmental level of a sibling has a significant bearing on his/her capacity to understand concepts of death like permanent, irreversible, inevitable, universal.[33] Regardless of how siblings understand and express their grief, it is critically important to remember that they are part of the social context in which the death has occurred. Their needs for explanation and support are just as important as the needs of their parents.

The death of a child does not occur in isolation, but rather it occurs in a social context that includes many variables. The main ones are parental coping capacity and skills, family and relationship functionality, social networks, parental physical and mental health issues, education, socioeconomic status and, importantly, any real or perceived parental responsibility in the death of the child. Thus the broader social context will have relevance to how parents and extended family members manage the impact of the child's death.[31] Any available psychosocial assessment or information, such as that provided by the ED social worker, should be factored into the management of the family.

Support of the family

Generally, parents are completely unprepared for the impact of their child's death as they have no prior knowledge or experience to draw on.[34] Arranging support is essential, and early social worker involvement is highly desirable. Parents and other family members must be provided with information about 'normal' grieving and should be linked to appropriate resources. This can take the form of written information packs that parents can take away and which they may choose to read at a later time.[35,36] Referral information should be readily available for support groups with particular expertise relating to the death of a child such as SIDS & Kids,[37] SANDS Australia,[38] Compassionate Friends,[39] and other relevant organisations. The extent of involvement of support by ministers of religion will depend on the wishes of and the religious commitment of the family. Ideally there should be a protocol that facilitates ready access to this material.

Practical assistance with arrangements at the time of the child's death, including organising family support and funeral and financial assistance, should be offered to families as appropriate, while being sensitive to the social and cultural environment of the family.

Cultural implications

Many cultures have specific rituals and practices concerning death. It is critical to listen to the family members and be guided as much as possible by their requests. Some of these rituals may require modification when the death of a child has been referred to the coroner's office. Sensitivity is essential.

It is difficult to make broad statements about the cultural practices related to death and dying in indigenous (Aboriginal and Torres Strait Islander) communities, because across Australia there are different practices and rituals. Examples of the kinds of cultural practices and rituals to be aware of include the following:

- When a child is dying many families will want any extended family present to be in attendance.
- During the grieving process pre- and post-death, loud crying/wailing may need to occur as part of the community's customs. Privacy in these circumstances is desirable.
- Senior female or male figures may wish to take a lead role in mourning

rituals after death has occurred. This can include ceremonial cleansing (washing the child's body), dressing and handling.

- Funeral arrangements may have to be organised by a specific extended family member or a senior member of the community.
- 'House smoking' (burning leaves in order to prevent the spirit from 'rising up') may need to occur relatively quickly after death has occurred. This can mean that the family will want to return to their community quickly.
- Many communities forbid the use of the deceased person's name after death (for up to 1 year). Nicknames or aliases may be used after death has occurred. A family member with the same name as the deceased may use his/her second name during the mourning period.
- After a year, there may be some ritual (i.e. tombstone opening) associated with the end of the mourning process, which requires community members to return home even if in hospital themselves.

The Maori culture of New Zealand traditionally has family members present with the body from the time of death through interment. This maintains the harmony of the child, assisting the decedent to join his/her ancestors. Family members will want to be part of the 'laying out' of the body, washing, dressing, etc.

Other practices reflecting different cultural belief systems that may need to be considered include parents needing to remain with the body 24 hours after the death, caring of the body by staff of the same gender as the deceased child, laying the body to face a certain direction (Mecca), special roles for specific religious/spiritual leaders and the burning of incense/candles.

When working with families from different cultures following the death of a child, it is important to be guided by custom, ritual, experience and the family's cultural environment.

Legal issues

Each country, state and territory will have subtle variations as to the legal requirements for the documentation and handling of the body of a deceased. An up-to-date protocol should be available to ensure that proper procedures are followed.

A life extinct form will need to be completed by one of the attending ED

medical staff. However, it will also be necessary to decide whether or not a death certificate can be completed. If the patient was known to the hospital and the death was expected, the child's usual physician may be prepared to sign a death certificate. This physician may also discuss with the parents the option of performing a hospital-based autopsy.

Usually the death of a child in the ED is not anticipated and hence becomes a coroner's case. For a coroner's case, only a life extinct form can be completed, laying out of the body will be restricted to spot cleaning, the local police must be notified, and parents must not be left unsupervised with the body. It is desirable for the family to formally identify the child's body in the presence of the police. Otherwise identification will have to be performed later and probably at the morgue, a process likely to increase family distress. All medical notes, investigations, observation sheets, etc., should be provided to the police when they depart with the child's body for the morgue. Full and accurate documentation of all events in the patient's hospital chart is essential. This should include the date and time of death, the observations that specify that the child is clinically deceased, any relevant history surrounding the circumstances of the child's death and any relevant conversations held with the parents or family members. There are potentially legal consequences following any death, and the forensic issues need to be considered. For example, child abuse remains an important cause of deaths in infancy.

Organ and tissue donation and collection

Organ donation (e.g. heart, lungs, liver, kidneys) requires intact cardiorespiratory function but brain death. Because of the preconditions required by Transplant Acts before brain death can be declared, organ donation discussions are commonly deferred until admission to the intensive care unit.[40]

Tissue donation (e.g. corneas, heart valves, skin, bone ligaments and tendons) can occur from cadavers, and hence theoretically this issue could arise for children who die in the ED. However, deaths of children in the ED are usually coroner's cases. For parents faced with the extreme distress of the sudden death of a child and the need for coroner's case status, it may be potentially too distressing to parents for ED staff to raise the further issue of tissue donation in this setting. This can come a little later at the Forensic Pathology Institute level, when parents have had a little time to regain some degree of composure and hence may be better able to give informed consent. Sometimes donation can be

perceived by families and providers alike as a way to salvage some meaning from an acute, unanticipated, and tragic loss.[7] On the rare occasion when the issue of tissue donation is spontaneously brought up by parents in the ED setting, contact with the transplant coordinator can be initiated if there are no potential medical contraindications to tissue donation. Consent by the coroner must be obtained prior to tissue removal.

When children die suddenly and unexpectedly there may be merit in considering collecting perimortem samples in order to obtain as much information as possible. This might include urine and blood for metabolic profiling, genetics screening and other possible investigations such as liver of other tissues samples that may contribute to the understanding of cause of death. This will depend on location and is more likely to be valuable in a major centre where appropriate pathology facilities are immediately available.

Debriefing and support for emergency department staff

Much of what is written about the family grief reactions applies equally to the ED staff, and due consideration of staff reactions is very important. A healthy approach is to factor the reality of day-to-day exposure of grief and loss into the culture of a busy ED. There is a paucity of literature on the reactions of staff and grief management among ED staff members. Truong et al.[41] described the ability of ED providers to normalise the abnormal events as a protective mechanism as ‘routinisation of disaster’.[41]

Identifying abnormal psychological symptomatology in ED staff (flashbacks, sleep disturbance, bad dreams, absenteeism, detachment, intensified emotions, etc.) and making ongoing psychological counselling available to affected staff are clearly important. Such symptomatology may occur as a result of either a single exposure or cumulative exposures to traumatic situations. It is important for senior ED staff to promote the concept of self-care, to guarantee confidentiality to staff experiencing problems, and to ensure staff are made aware of counselling options available to them should they experience problems.[42]

Performing an operational debriefing of the resuscitative process with a view to clarifying events for attending staff and identifying areas for potential improvement is essential. Given the fact that it is often challenging to find a time to gather those who wish to participate in a busy ED environment, there are

suggestions that even a simple acknowledgement at the bedside after the death of the patient may be beneficial to staff, given the healing potential of such a closing ritual.[41,43]

The same cannot be said for psychological debriefing sessions. It has become a popular and widespread practice to conduct single session psychological counselling for personnel attending traumatic critical incidents. ED staff in attendance at an unsuccessful resuscitation fit into this situation. A recent Cochrane Review concluded that single session psychological debriefings have not only failed to reduce the incidence of post-traumatic stress disorder but actually increased the risk of developing it.[44] In addition, there was no evidence of reduction in general psychological disturbance, depression or anxiety. This is an area where more research is required.

Collaboration with paediatric palliative care services

Studies in children with known life span–limiting conditions report that between 3% and 20% of deaths in that population will occur in the ED.[45,46] For many children receiving palliative care, advance care plans are in place, and it can be very helpful for ED staff to have an understanding, in advance, of the hopes, concerns and wishes that the child and family may have expressed.[7]

The concept of a good death

Data from the National Center for Health Statistics for the most recent year completed (2009) in the United States showed that fewer than 2% (48,000) of deaths occur in the population of children younger than 18 years.[47] This statistic is strikingly different from a century ago, when 30% of all deaths were in children younger than 5 years. On one side, these data reflect progress in child health but also underscore that child death, unlike parental or spousal death, is no longer an expected part of life.[7]

The Institute of Medicine report on childhood death provides the following definitions for good and bad deaths:

> *A decent or good death is one that is: free from avoidable distress and suffering for patients, families, and caregivers; in general accord with patients' and families' wishes; and reasonably consistent with clinical,*

> *cultural, and ethical standards. A bad death, in turn, is characterized by needless suffering, dishonoring of patient or family wishes or values, and a sense among participants or observers that norms of decency have been offended.*[48]

Aspects of what might constitute a ‘good death’ in the ED are caring for the survivors of the child’s death in a way that affirms their trust and allowing them to understand the events leading up to the death of the child and to say goodbye to their child in whatever way is meaningful to them.[7]

Conclusion

The death of a child has the most profound effect on parents, family and friends. It can also have a profound effect on staff involved in the resuscitation process. It requires the sensitivity and strength of clinical staff to help relatives through this difficult time and to assist in the initiation of a healthy grieving process. A thoughtful and sensitive approach is likely to have profound and positive long-term implications for all those impacted upon by the death of a child.

Controversies

1. There remains some controversy about actively encouraging parents to be in the resuscitation room.
2. There is little evidence to support the widespread practice of mandatory single-session psychological counselling of distressed staff who attended the child.
3. Raising the issue of tissue donation with parents in the emergency department setting is difficult.

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SECTION 20

Analgesia and Sedation

OUTLINE

20.1

Analgesia

Adrian Mark Bonsall

ESSENTIALS

1 Acute pain is one of the most common emergency department presenting problems.

2 Pain-rating scales suitable to the age and development of the child are useful in establishing a child's level of pain and assessing the adequacy of analgesia.

3 Adopt a multimodal (non-pharmacological, pharmacological) and multidisciplinary (medical staff, play therapists, parents) approach to pain management.

4 Tailor interventions to the individual child.

5 Become familiar with dose, administration and potential complications of a range of analgesics.

6 Combining drugs without detailed knowledge and training risks serious adverse outcomes.

Introduction

Analgesia is the relief of the perception of pain without sedation.

Pain is a more difficult concept to define precisely or to measure objectively. The International Association for the Study of Pain (IASP) defines pain as an unpleasant sensory and emotional experience, associated with actual or potential tissue damage, or described in terms of such damage.[1] It is the significant emotional dimension that creates considerable variability in how a painful

stimulus is experienced and thus how the interactions of physiological, psychological, developmental and situational factors can modify behaviour in both the short and long term.

Acute pain in children is one of the most common reasons for presentation to the emergency department (ED).[2] In addition to the underlying injury or illness, subsequent medical procedures may also engender pain which is often associated with increased anxiety, avoidance behaviour, systemic symptoms and parental distress. Treating pain may not only relieve acute suffering but also decrease ongoing anxiety and negative memories, facilitate medical investigations and aid cooperation with other non-painful procedures and treatments. The use of analgesia in procedural sedation is discussed in more detail in Chapter 20.2.

Pain may be classified in a number of ways, e.g. by severity, cause or pathophysiology.[3] A simple classification is shown in Box 20.1.1. Most commonly in the paediatric ED setting it is procedural and acute pain which need to be addressed.

Children with painful conditions can be difficult to assess, and their pain is often still underestimated and undertreated. Children often receive less analgesia than adults, and the administration of analgesia varies by age, with the youngest patients at the highest risk of receiving inadequate analgesia.[4,5]

Box 20.1.1 Pain classification

Procedural	Transient while stimulus is applied but before significant tissue damage occurs
Acute	Significant local tissue damage with acute inflammation but normal innervation
Intractable (chronic)	Continuing in the absence of acute inflammation and normal healing
Neuropathic	Associated with peripheral, central or autonomic nerve damage

Children's pain can be underestimated because assessment requires tools that account for the wide range of children's developmental stages. Pain is often undermedicated because of fears of oversedation, respiratory depression, addiction and unfamiliarity with use of sedative and analgesic agents in children.[5]

ED staff should be proficient in the assessment and safe management of pain in children. Early and appropriate analgesia may be best achieved by using a systematic approach with well-developed pain management educational programmes, specific pain assessment and management policies, and benchmarked standards for time-to-analgesia within the ED.

Assessment of pain

Assessment of pain should be individualised, continuous, measured and documented. Pain assessment and measurement tools have been developed that are suitable for children of different ages and developmental stages. Accurate assessment requires a detailed pain history and consideration of the complexity of the child's pain perception and the influence of situational, psychological and developmental factors. Four useful means of recognising pain in children are outlined in Box 20.1.2.

Box 20.1.2 Recognition of pain in children

1. The child's self-report of pain
2. Behavioural changes, e.g. crying, guarding, facial grimacing
3. Physiological changes, e.g. pallor, tachycardia, and tachypnoea
4. Pathophysiological process, e.g. fracture or burn

Because of its subjective nature, pain is best assessed using the child's self-report. Observational assessment scoring may be useful when the child is too young or self-report is not possible, e.g. children with cognitive impairment. Pain ratings provided by parents or regular carers may also be used.[6] However, whilst there is good correlation between the child's and the parent's assessment of pain intensity, parents tend to underscore more severe pain being experienced by their children.[7] Physiological measures (e.g. heart rate and respiratory rate) may be useful in pain assessment in non-verbal or sedated children but may be confounded by stress reactions. For example, the child who is febrile or an infant who is hungry may give inappropriately high scores.

Specific pain assessment tools employing behavioural and vital sign observations have been developed for neonates (e.g. CRIES) and non-verbal (e.g. FLACC) or cognitively impaired children (e.g. r-FLACC, NCCPC-R or COMFORT tools).[8–12] Pain scales for older children able to self-report, such as the Faces rating scale, which uses facial expressions depicting increasing pain or simple numerical or analogue scales, are commonly employed. Some of these age-dependent pain rating scales are outlined in Table 20.1.1.

Management

Optimal analgesia is achieved by the combination of both non-pharmacological and pharmacological strategies with regular pain assessments. When using drugs there should also be careful consideration of the most appropriate route and dosage and close monitoring for adverse effects.

Pain management strategies should be individualised for the child's level of pain and the anticipated discomfort of any procedure to be undertaken. Choice of agent will also depend on local resources, familiarity with doses, duration of action, adverse effects and contraindications.

Non-pharmacological methods

There are many non-pharmacological techniques that can be used to mitigate pain and distress in the ED and complement the use of pharmacological methods.[13–17] Some of these are listed in Box 20.1.3.

Providing a non-threatening, friendly environment, which might include wall decorations of animals or familiar cartoon/TV characters, can help reduce anxiety. Positioning of the child can be important. For example, babies may be more settled when swaddled; conversely, constraint in toddlers can be distressing, so allow a child to take up the most comfortable position and provide pillows to support an injured limb. Other physical techniques include slings, splints or other immobilisation methods and application of ice or cold/heat packs. Gentle, unhurried and confident handling can often minimise distress and pain.

Older children may respond to age-appropriate explanations and providing realistic expectations on the management of their pain. It is helpful to gain a rapport and to be honest when reassuring. Claims of the child feeling no pain should be avoided as they are often soon disproved and risk loss of confidence or cooperation from the child and parents.

Parents play an important role as a familiar comforter in unfamiliar surroundings. They can be included in many distraction techniques. However, overt parental anxiety can fuel a child's discomfort.

Play therapists are a good resource if available.[18] They are trained in many distraction strategies and can also be very useful in preparing a child for a painful procedure. Specific distraction strategies, which can be provided by staff or parents, may include toys, bubbles, guided imagery, music, TV and DVD/video players, hand-held game consoles, and smart phones or tablets. In infants, breast-feeding or the use of a pacifier may be comforting. Other

methods, more commonly used in the chronic setting, include reinforcement of coping behaviours, hypnosis, biofeedback, muscle relaxation and deep-breathing techniques.

Pharmacological methods

The choice of agent is dependent on acuity, severity and source of the pain, route availability, expected duration and patient factors such as age or genetic variation.

Some of the more commonly used agents discussed below have doses detailed in Table 20.1.2.

Oral analgesic agents

Apart from sucrose, these agents may also be administered via a naso- or orogastric tube and a gastrostomy tube if in a suitable liquid or crushed tablet form.

Oral paracetamol and NSAIDs

Paracetamol is usually the first line of therapy for mild to moderate painful conditions. It can also be given rectally and intravenously (IV). The rectal route is contraindicated in severely immunocompromised children. IV paracetamol is available when oral and rectal dosing is contraindicated and is dosed on lean body weight. Paracetamol may be synergistic with other analgesics.[19] It is generally safe; however, hepatotoxicity is possible in overdose or extended use. Non-steroidal anti-inflammatory drugs (NSAIDs), such as ibuprofen, are usually second-line therapy and have excellent analgesic and anti-inflammatory properties, which may enhance the effects of paracetamol. NSAIDs should be avoided if there is GI ulceration, coagulopathy or active bleeding, severe asthma, renal disease and in some orthopaedic conditions where there is a high risk that bone healing may be compromised. Both drugs should be used with caution where there is significant dehydration or liver dysfunction. In addition aspirin is not recommended as an analgesic for children due to the reported association with Reye syndrome in the context of some viral infections.[20]

Oral sucrose

Oral sucrose (24–25%) has been shown to be a simple, safe and effective means

of providing analgesia for neonates and young infants for short painful events (e.g. heel prick, venepuncture, lumbar puncture).[21–23] It stimulates endogenous opioid and non-opioid pathways in the brain and thus may be less effective if the baby is already on an opioid. It may be administered via oral syringe or on a pacifier approximately 2 minutes prior to the painful event.

Codeine and oxycodone

Oral codeine has traditionally been used for moderate to severe pain and can be found in combination formulations with paracetamol; however, it is a prodrug with little inherent pharmacological activity and must first be metabolised by the liver into morphine. There is substantial genetic variability in the activity of the cytochrome P450 hepatic enzyme (CYP2D6) responsible for this conversion. Non-metabolisers will have no analgesic benefit, while ultrarapid metabolisers run the risk of significant respiratory suppression especially if there is any obstructive sleep apnoea.[24] Oxycodone is a semisynthetic opioid that does not require metabolism to be active. Evidence from recent work shows that oral oxycodone produces greater pain relief compared with codeine and also has a better side effect profile with less itching, less nausea, and fewer allergic reactions.[25] The majority is processed in the liver by CYP3A4 to less active compounds, but a small proportion is also metabolised by CYP2D6 to the active oxymorphone which means ultrarapid metabolisers still may be at some risk. A slow release formulation of oxycodone may be used for background analgesia in more chronic pain.

Table 20.1.1

Pain scoring

CRIES pain rating scale[8]

The CRIES scale can be used in neonates and young infants up to a few months of age. It was originally designed for post-operative pain assessment; however, it can be employed for other procedures where baseline vital signs are known. Each of the 5 categories is scored from 0 to 2, and the scores are added to get a total score from 0 to 10.

	0	1	2
Crying	No	High pitched	Inconsolable
Requires O_2 for Sat >95%	No	<30% O_2	>30% O_2
Increased vital signs	HR & BP less than or equal to patient's baseline (e.g. pre-op)	HR & BP increased by <20% of patient's baseline	HR & BP increased by >20% of patient's baseline
Expression	None	Grimace	Grimace/grunt
Sleepless	No	Wakes at frequent intervals	Constantly awake

FLACC scale

The FLACC scale is a behavioural scale for scoring pain in children between the ages of 2 months and 7 years or in

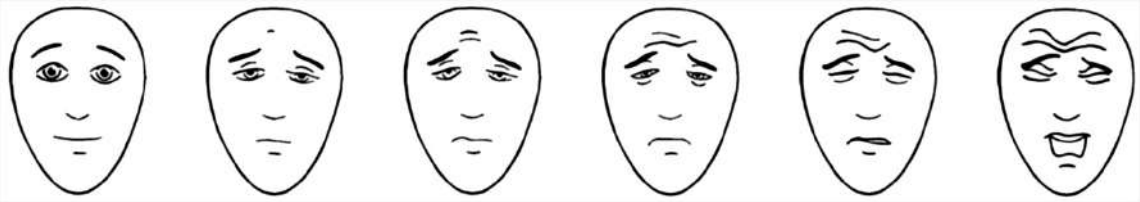
persons unable to communicate. Each of the 5 categories is scored from 0 to 2, and the scores are added to get a total score from 0 to 10.

	0	1	2
Face	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant frown, clenched jaw, quivering chin
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking, or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting back and forth, tense	Arched, rigid, or jerking
Cry	No cry (awake or asleep)	Moans or whimpers, occasional complaint	Crying steadily, screams or sobs, frequent complaints
Consolability	Content, relaxed	Reassured by occasional touching, hugging or 'talking to', distractible	Difficult to console or comfort
The FLACC behavioural pain assessment scale © University of Michigan Health System can be reproduced for clinical or research use.			

Faces rating scales (FRS)

These scales can be used with young children (as young as 4 years of age). They also work well for older children and adolescents, including those who speak a different language.

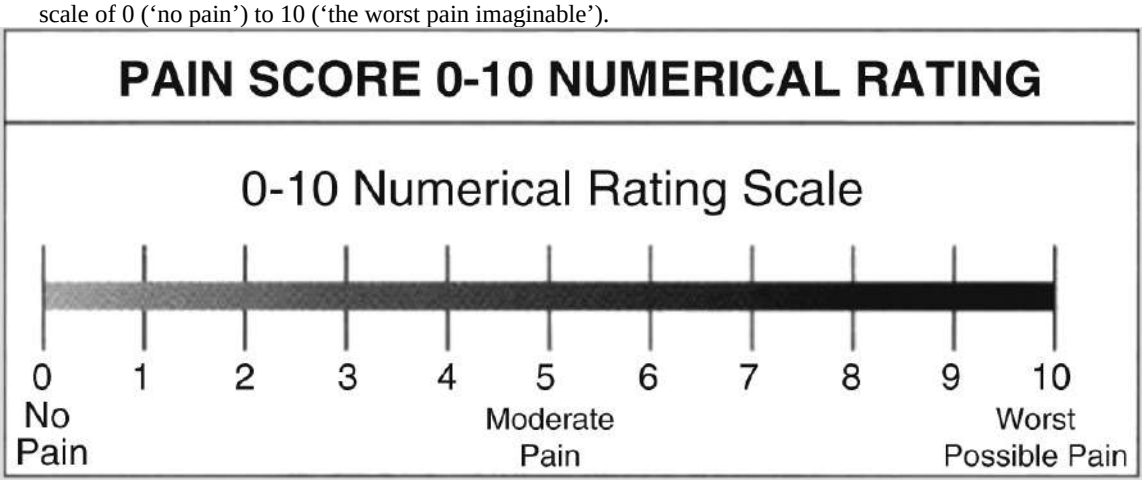
Ask the patient to choose the face that best describes how he/she feels. The far left face indicates 'no hurt', and the far right face indicates 'hurts worst'.



The Faces Pain Scale – Revised (FPS-R) can be downloaded (including instructions in multiple languages) from The International Association for the Study of Pain (IASP) website at www.iasp-pain.org/Education/Content.aspx?ItemNumber=1519&navItemNumber=577.

Numerical rating score (NRS)

This tool may be used for children over the age of 6–8 years. Instruct the patient to rate his/her pain intensity on a scale of 0 ('no pain') to 10 ('the worst pain imaginable').



BP, blood pressure; HR, heart rate; Sat, saturation.

Tramadol

Tramadol is a synthetic analogue of codeine with high oral bioavailability. It compares favourably with oral NSAIDs for pain and may be used if opioids are contraindicated, ineffective or causing side effects.[26] It is a weak central μ-opioid

agonist (30% of effect) and also inhibits noradrenaline (norepinephrine) and serotonin reuptake (70% of effect). Thus it should be used with caution with other opioids and avoided if the patient has been on serotonin re-uptake inhibitors, tricyclic antidepressants, major tranquillisers, fentanyl, pethidine or monoamine oxidase inhibitors as seizures or a serotonin syndrome can result. The most common side effects are nausea, vomiting and dizziness, however it has less sedative, respiratory depressive and pruritic effects than morphine. It can also be given by slow infusion IV over 15–20 minutes to reduce the incidence of nausea and vomiting. A ceiling effect limits tramadol usefulness to moderate pain.

Box 20.1.3 Supportive and distractive techniques

Environment

- Calm friendly non-clinical atmosphere
- Toys, mobiles, pictures and videos

Psychological

- Parental presence
- Age-appropriate communication
- Clear confident instructions

Cognitive–behavioural

- Distraction techniques
- Hypnosis and biofeedback
- Art/stories
- Music/video/TV
- Interactive computer games
- Guided imagery
- Muscle relaxation and deep-breathing techniques
- Reinforcement of coping behaviours

Physical

- Massage/rubbing
- Comfort swaddling (infants)
- Heat/cold techniques
- Immobilisation and elevation (injured part)

Breast-feeding

- Comforter (favourite blanket/soft toy)

Oral morphine

Morphine is an effective analgesic and widely used in parenteral formulations. However, it has a low oral bioavailability (30–40%), and with the advent of other synthetic opioids with higher non-parenteral route bioavailabilities and better side effect profiles, oral morphine is not often used in acute pain management in the ED.

Intranasal or inhaled options

Methoxyflurane

Methoxyflurane is a halogenated ether. Its rapid action, portability and ease of administration mean that it is most useful in the acute, pre-hospital setting, when alternatives are limited or impractical.[27] It is particularly effective for trauma pain but may also be used for brief painful procedures such as wound and burns dressings. A recent review has shown that the inhalational agent methoxyflurane is also safe and effective in the ED setting.[28] The commonly used ‘Penthrox’ inhaler is now available with an activated charcoal scavenging chamber to reduce environmental contamination.

Intranasal fentanyl

Intranasal fentanyl using a mucosal atomiser device provides safe and effective analgesia equivalent to parenteral morphine in children as young as 1 year of age.[29–32] It offers quick onset and is less invasive, and its duration of action,

although only 30–60 minutes, allows time for topical anaesthetic application prior to IV cannulation for ongoing analgesia. It is particularly useful for fractures or burns dressings, but its utilisation is spreading into other areas.

Nitrous oxide

Nitrous oxide mixed with oxygen has a potent analgesic action with rapid onset and offset. It is an excellent analgesic sedative for relatively brief painful procedures such as gaining rapid IV access, injecting local anaesthetics (LAs), performing a nerve block, laceration suturing or splinting a fractured limb.[33–35]

Entonox® (50% nitrous oxide, 50% oxygen) is usually delivered via a demand-valve system. This limits its use in younger or uncooperative children. Machines that deliver variable concentration (30–70%) nitrous oxide via a continuous flow system allow the use of this agent down to age 1 year, where it has been shown to be safe when embedded in a comprehensive sedation programme.[35]

Nitrous oxide alone is effective in achieving moderate levels of procedural sedation for a high percentage of children with painful conditions but may require the use of adjunctive analgesics for very painful procedures.[35,36]

Oxygen should be administered for 3–5 minutes after cessation of nitrous oxide to prevent diffusion hypoxia. Nitrous oxide is contraindicated in conditions involving closed air spaces (e.g. pneumothorax, bowel obstruction). The most common adverse effect is vomiting, and a fasting time of at least 2 hours is advised. Prolonged exposure has been associated with alterations in vitamin B_{12} and folate metabolism, megaloblastic bone marrow changes and subacute combined degeneration of the cord.

Parenteral analgesics

Although these may be any systemic analgesics given by a route other than the gastrointestinal tract, the common parenteral routes are IV, intramuscular (IM) and subcutaneous (SC). The IV route is often favoured for analgesia because it generally has faster onset and better efficacy than the IM and SC routes which can have variable absorption depending on perfusion. However, the IV route does require the delay and discomfort of cannulation, but that may be preferred to multiple IM injections if the need for parenteral pain relief is ongoing. Continuous IV (or occasionally SC) infusion may be used for ongoing analgesia requirements.

Patient-controlled analgesia (PCA) and nurse-controlled analgesia (NCA)

If a patient needs regular strong analgesia (most commonly opioids), a special IV pump may be set up to deliver bolus doses within pre-set dose and frequency limits to allow older, competent children to self-manage pain relief by pressing a button.[37] In children not able to use a PCA machine, NCA may be used by nurses with close monitoring and serial assessments.[38]

Parenteral opioid analgesics

Opioid analgesics are the mainstay of treatment for severe pain. Opioids are ideally administered IV in a dilute solution and titrated against response. Important side effects include respiratory depression, drowsiness, bradycardia, constipation, nausea and vomiting, and, in some, histamine release. Common choices would include morphine and fentanyl. The latter is a potent rapid-acting analgesic with a shorter duration of action. As fentanyl is metabolised in the liver to inactive metabolites, it is preferred to morphine (which is mainly excreted in the urine) in kidney impairment. Ex-premature infants and young infants (under 3 months) may require opioid dose reduction as they are at increased risk of apnoea and other side effects because they metabolise opioids more slowly.[39] In addition there is concern that early prolonged exposure to opioids may affect neurodevelopment of a number of brain systems.[40]

Ketamine

Ketamine is a phencyclidine derivative with NMDA antagonist and opioid receptor agonist properties. It is an analgesic dissociative anaesthetic that is an increasingly popular choice for procedural sedation as its action produces a trance-like state of sedation, amnesia, analgesia and some motion control with few side effects. This is discussed in greater detail in Chapter 20.2. Sub-dissociative doses of ketamine are not yet commonly used in ED for analgesia alone as the evidence base for use in children is limited. However, there are some studies that suggest equivalent efficacy to opioids.[41]

Multiple routes of administration are available. IV and IM routes are commonly used, but there is growing evidence that intranasal ketamine may be comparable to intranasal fentanyl for pain associated with limb injuries.[42] The oral route is another route that is available but not currently employed regularly in the paediatric ED.

Local anaesthetic agents

The expanded use of LAs has revolutionised the management of simple lacerations in the ED and has also greatly improved conditions for IV cannulation and lumbar puncture. These agents provide a non-invasive means of producing local anaesthesia and can be applied at triage to facilitate timely management in the ED.[43] In addition LA use in regional anaesthesia can provide timely pain relief and reduce the need for general anaesthesia in managing some fractures. However, clinicians must be mindful of the risk of overdose. LA toxicity may be heralded by dizziness, peri-oral tingling, metallic taste, altered mental status, arrhythmias and seizures and result in refractive hypotension and cardiac arrest. Methaemoglobinaemia can also complicate benzocaine, prilocaine and lidocaine administration.[44]

Table 20.1.2

Commonly used agents for analgesia

Classification	Doses	Comments
Enteral analgesics		
Paracetamol	15–30 mg kg^{-1} PO/PR (<3 months 10 mg kg^{-1})	30 mg kg^{-1} stat. only as a single dose (check no recent doses of paracetamol)
Ibuprofen (NSAID)	5–10 mg kg^{-1} PO	Avoid in dehydration, renal impairment, GI ulceration, coagulopathy, severe asthma
Oxycodone	0.1–0.2 mg kg^{-1} PO	Better analgesia and fewer side effects than codeine
Tramadol	1–2 mg kg^{-1} PO/IV	Risk of interactions with other drugs (see text)
Sucrose (24–25%)	0.1–0.5 mL PO repeated up to 2 mL per procedure and maximum of 5 mL day	Young infants. Ideally combine with non-nutritive sucking
Inhaled analgesics		
Nitrous oxide	50:50 mix inhalation agent (ENTONOX®) N_2O continuous flow variable mix 30–70% with oxygen	Rapid onset; continuous flow delivery system allows use in young children/short duration of action. Vomiting can occur; contraindicated with pneumothorax/chest injuries
Fentanyl	1.5 mcg kg^{-1} per dose (maximum 75 mcg)	
Parenteral analgesics		
Morphine	0.05–0.1 mg kg^{-1} per dose (<3 months) IV/IM 0.1–0.2 mg kg^{-1} per dose (>3 months) IV/IM Infusion (>3 months) 0.01–0.04 mg kg^{-1} h^{-1}	Cardio-respiratory depression with IV/IM doses; IV dose preferred
Fentanyl	1–2 mcg kg^{-1} per dose IV/IM	Less histamine response and better renal profile than morphine
Opioid reversal agent		
Naloxone	10 mcg kg^{-1} IV	May need to repeat doses
Local anaesthetic agents		
Topical		
Surface EMLA®	Eutectic mixture of lidocaine (lignocaine) 2.5% and prilocaine 2.5%	Requires 60 minutes post-application to achieve satisfactory dermal anaesthesia
AnGel®/Ametop®	Amethocaine 4%	Requires 30–45 minutes post-application to achieve satisfactory dermal anaesthesia
Laceraine®	Mixture of 4% lidocaine, 0.5% tetracaine and adrenaline (epinephrine) 1:1000 maximum dose 0.1 mL kg^{-1}	Requires >20 minutes of good contact with wound to provide anaesthesia; may require supplementation with small doses infiltrated LA
Infiltration		
Lidocaine 1–2%	3 mg kg^{-1} maximum dose without adrenaline 7 mg kg^{-1} maximum dose with adrenaline	Pain of injection can be minimised by using narrow-guage needles (e.g. 31G), slow infiltration, infiltrating through wound edges, pre-treatment with topical LA, buffering and warming
Regional blocks – femoral blocks		
Bupivacaine 0.25–0.5%	2 mg kg^{-1} Volume: 0.8 mL kg^{-1} 0.25% (≤20 kg) or 0.4 mL kg^{-1} 0.5% (>20 kg) Maximum 30 mL	Doses are for bolus dose block. Onset 15–30 minutes. Can last 7–14 hours
Ropivacaine 0.5%	3.0 mg kg^{-1} Volume: 0.6 mL kg^{-1} Maximum 30 mL	May be less cardiotoxic than bupivacaine.[75] Dose is for bolus dose block. Onset 15–30 minutes. Can last 7–14 hours
Intravenous regional block – Bier block		
Lidocaine 0.5% or prilocaine 0.5%	3.0 mg kg^{-1} IV Volume: 0.6 mL kg^{-1} Maximum 40 mL	Onset 5–15 minutes
Management of LA toxicity		
General	Oxygen. Use conventional therapies to treat hypotension, bradycardia and tachyarrhythmia	
Consider lipid emulsion 20% if significant toxicity	1.5 mL kg^{-1} IV over 1 min (can repeat twice every 5 min) and start infusion at 15 mL kg^{-1} h^{-1}. Double the rate any time after 5 minutes, if not stable or inadequate circulation. Maximum total dose 12 mL kg^{-1}	
Full algorithm may be downloaded from The Association of Anaesthetists of Great Britain & Ireland at http://www.aagbi.org/sites/default/files/la_toxicity_2010_0.pdf		

GI, gastrointestinal; LA, local anaesthetic; IM, intramuscular; IN, intranasal; IV, intravenous; PO, per orem; PR, per rectum.

Topical anaesthetic agents

EMLA® (a eutectic mixture of two LAs: 2.5% lidocaine [lignocaine] and 2.5% prilocaine) is a well-established topical anaesthetic for use on intact skin prior to venepuncture, intravenous cannulation or lumbar puncture. Its use in the ED is, however, limited, due to its long onset to peak effect (at least 1 hour) and its vasoconstrictive effect, which may make cannulation more difficult. EMLA® has a theoretical risk of methaemoglobinaemia and is not recommended in infants less than 3 months of age.[45]

Amethocaine, also known as tetracaine, can also be used for similar skin-puncturing procedures (e.g. Ametop® or AnGel® both contain 4% amethocaine).

It has a quicker onset of action (30–45 minutes) than EMLA, and its vasodilating effect may facilitate cannulation. Side effects reported include itching and erythema. Amethocaine has been shown to be superior to EMLA® in terms of lessening pain associated with IV cannulation and is more effective than EMLA® when application time is less than 60 minutes.[46] There is also an ophthalmic formulation (0.5–1%) for eye use.

Topical wound anaesthetics, e.g. Laceraine® (ALA: adrenaline [epinephrine] 1:1000, lidocaine 4%, amethocaine 0.5%), permit wound management with minimal to no discomfort.[47,48] It should be placed on a small wad of gauze/cotton wool inside the wound and covered with a watertight dressing for 20–30 minutes to provide sufficient anaesthesia for suturing 75–90% of scalp and facial lacerations and 40–60% of extremity wounds.[48,49] Its vasoconstrictive effect is also useful prior to application of cyanoacrylate tissue adhesive.

Cocaine-containing topical wound anaesthetics (e.g. TAC – tetracaine, adrenaline, cocaine), though effective, are more expensive, require secure storage and potentially risk the serious systemic side effects that have been reported with cocaine-containing preparations.[50–52]

Topical preparations for mucous membranes are also available. For example, xylocaine viscous, a formulation of lidocaine, has been used to encourage fluid intake when applied to painful lesions in the oropharynx, though recently this efficacy has been challenged.[53] Similarly pain from oral cavity ulcers or teething may respond to teething gels. Some of these contain choline salicylate, which has led to salicylate toxicity or Reye-like syndrome in several cases of excessive usage.[54–57]

Local anaesthetic infiltration

The pain associated with wound infiltration using an injectable LA can be lessened by buffering (e.g. add 1 mL 8.4% sodium bicarbonate to 9 mL 1% lidocaine), warming the solution to body temperature, using fine-bore needles and injecting slowly through the wound edges rather than through intact skin.[58–62] In addition, use of other analgesia, e.g. nitrous oxide or distraction techniques, during the process of injection often mitigates the pain of instillation. Adrenaline can be added to prolong the local effect of the anaesthetic and reduce bleeding. Traditionally adrenaline is not used where there are end arteries, such as the tips of the digits, pinnae, nose and penis. Although more recent evidence suggests 1:100,000 adrenaline appears to be safer than first thought, it may be prudent to avoid it in these situations in the ED until more evidence in children is

available.[63–65] It is important to consider safe dosage regimes of local anaesthetics, with and without adrenaline, and include any use of topical anaesthetic in the total dose.

Regional anaesthetic techniques

Regional nerve blocks may be used for either pain relief (e.g. femoral nerve or fascia iliaca blocks for femur fracture) or to facilitate foreign body removal, suturing, fracture or dislocation reduction (e.g. auricular, digital or metacarpal blocks).[45,66] Nerve blocks have traditionally been performed using 'blind' techniques based on anatomical landmarks with or without a nerve stimulator. The increasing availability and expertise of ultrasound in the ED has the potential to achieve equivalent or better success rates with fewer adverse events.[67]

Femoral blocks

Femoral blocks have been shown to be effective and reduce the need for opioid analgesia with femoral shaft fractures.[68,69] There are a number of methods available for femoral blocks. Femoral nerve block (FNB) with ultrasound guidance or fascia iliaca compartment block (FICB) with or without ultrasound guidance is probably the best and safest. Use of a nerve stimulator or 'blind' approach for an FNB is less ideal as there is potential for painful muscle contraction and inadvertent intravascular or intraneural injection.

The FICB is technically easier and, because it is performed away from the femoral vessels, can be safely performed without ultrasound guidance. In addition to the femoral nerve, it also blocks the lateral femoral cutaneous nerve and the obturator nerve more reliably than the FNB.[70] It is a field block that relies on volume to maximise its effect. With the maximum dose limitation, the concentration of the LA may have to be reduced in order to maintain an effective volume.

Outlines for these femoral block procedures are presented in Box 20.1.4 and suggested LA doses given in Table 20.1.2.

Bier block

This is an example of IV regional anaesthesia. It is most commonly used for cooperative older children with distal forearm fractures that require closed reduction in the ED. If appropriate local anaesthetics and LA doses are chosen,

Bier blocks are safe, effective and avoid the need for sedation or a general anaesthetic in these patients.[71–73] In order to be carried out safely, the procedure requires at least two trained doctors, a nurse, a blood pressure tourniquet capable of maintaining a pressure of at least 200 mmHg, availability of resuscitation equipment and full non-invasive monitoring. It is contraindicated if the patient has an open fracture, compromised circulation or infection of the limb, pathologic hypertension, or a bleeding disorder.

A brief outline for the procedure is presented in Box 20.1.5 and suggested LA doses given in Table 20.1.2.

Chronic pain

The usual focus in the ED is acute pain, but patients with chronic pain will also present because of their chronic condition, complications of their analgesics or an unrelated painful issue. Chronic pain may be treated in a number of ways to provide ongoing background analgesia with provision for breakthrough pain. The non-pharmacological techniques and analgesics used can be some of those described above, but drugs may be time-released formulations or delivered in different ways, e.g. fentanyl transdermal patches. Examples of other drugs used include clonidine, amitriptyline, tapentadol, gabapentin and pregabalin. Advice from a chronic pain specialist can be invaluable with these patients who often have pain management plans tailored to the individual.

Box 20.1.4 Femoral nerve blocks

General

- Ensure the patient has no contraindications – allergy to local anaesthesia (LA), local infection or bleeding disorder.
- Examine and document pre-block neurological function of limb.
- Use 0.5 mL 1% lidocaine to anaesthetise the injection site.
- Identify inguinal ligament as a line between the ipsilateral anterior superior iliac spine and pubic tubercule.
- Use short-bevelled ‘blunt’ needles for block injection attached to a 20 mL syringe.
- Use bupivacaine 0.25–0.5% or ropivacaine 0.5% without adrenaline for

the block (see Table 20.1.2 for doses).

- When correctly positioned: aspirate, inject slowly and re-aspirate every 3–5 mL to check not in a vessel.
- Should be able to inject without significant resistance, and should not elicit paraesthesia or significant pain.
- Block may take 20–30 minutes to become effective. It can last 4–8 hours depending on the LA used.

Fascia iliaca compartment block

- Injection point is ~1 cm caudal to the junction of the lateral and middle thirds of inguinal ligament.
- Insert block needle perpendicularly to skin, then angle cranially and advance slowly until 2 distinct 'pops' are felt as the needle traverses the fascia lata and then the fascia iliaca.
- Apply gentle pressure just caudal to injection site to encourage LA to flow cranially.
- If resistance is felt, may be in muscle; retract needle slightly. If still have resistance, pull back to skin and re-advance.
- If an ultrasound is available it can also be used to confirm correct placement.

Femoral nerve block with ultrasound guidance

- Palpate the femoral pulse ~1 cm caudal to the inguinal ligament.
- Using a transverse orientation of the ultrasound probe, identify and centre femoral vessels and nerve on ultrasound probe display. The femoral nerve is lateral to the pulsating artery, which itself is just lateral to the compressible vein.
- In-plane approach of block needle from lateral end of probe.
- Position needle tip just lateral to nerve.
- When injecting should see LA flood around femoral nerve.

Discharge analgesia

Lack of appropriate or inadequate dosing of discharge analgesia is an ongoing problem. Pain experienced at home after acute injury or procedure in the ED can place considerable extra burden on family physicians for pain-related issues. It is essential to include adequate discharge analgesia in ED pain management guidelines. Ibuprofen was found to be preferable to paracetamol and codeine for outpatient management for children with uncomplicated arm fractures.[74]

Box 20.1.5 Upper limb Bier block

- Place a cannula distal to the fracture.
- Insert a second cannula in the contralateral arm in case additional analgesia to tolerate the tourniquet is required or in the rare event of local anaesthetic (LA) toxicity.
- Ensure a radiographer is immediately available, and prepare materials for applying a plaster back slab.
- Elevate the affected arm for 1–2 minutes with compression of the brachial artery.
- Inflate an upper arm blood pressure cuff to ~200 mmHg, and maintain the pressure for the duration of the procedure.
- Inject LA through the ipsilateral cannula (see Table 20.1.2 for doses) and the cannula removed.
- Once anaesthesia is achieved (usually 5–10 minutes), reduce the fracture, apply a back slab and have a portable X-ray taken to confirm adequate reduction.
- The cuff may only be deflated slowly in stages after a minimum of 20–25 minutes.
- Observe for LA toxicity.

Controversies and future directions

1. The employment in the emergency department (ED) of drugs and techniques previously the province of anaesthetists and pain specialists

has been controversial in the past, e.g. ketamine. However, improvements in training programmes and experience have led to increasing acceptance of their routine use.

2. Use of quality-improvement processes and high-quality research will boost safety and efficient use of these analgesics and engender other novel agents and non-pharmacological techniques to improve the experience of children presenting to the ED with painful conditions.
3. Alternative routes of drug administration have already had a great impact on paediatric analgesia practices. The delivery of established drugs by innovative, less-invasive methods is an exciting area for future research.
4. Consensus-based recommendations for standardising the terminology used for reporting adverse will help to create a uniform reporting mechanism for future studies in this area.

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20.2

Paediatric procedural sedation within the emergency department

David M. Krieser, Shona McIntyre, and Bindu Bali

Introduction

Emergency department (ED) attendances due to acute pain are very common in children. The Paediatric Research in Emergency Departments International Collaborative (PREDICT) reported on 314,025 attendances, with abdominal pain, lacerations and forearm fractures among the ten most common ED diagnoses,[1] each inherently associated with pain. Pain can be the trigger for ED presentation but may also be the consequence of necessary medical management within the ED.

The development of procedural sedation (PS) parallels the development of paediatric emergency medicine as a subspecialty. As clinicians developed confidence in managing the sedated child, procedures previously performed in the operating theatre have migrated to the ED.[2] ED performance of PS allows more rapid discharge, reduces costs, and increases clinician skills and job satisfaction, while making unpleasant procedures more tolerable to children and their families.[3,4]

The goal of procedural sedation

The ultimate goal of PS is to safely achieve a level of consciousness that allows successful completion of the necessary procedure, a return to consciousness and discharge home within a reasonable time frame. However, it is not enough to safely sedate a child. A practitioner competent in the procedure that is deemed necessary must be available. If there is doubt, then ED PS may be inappropriate.

Collaborative planning between the ED staff and other specialists may be required. The procedure may need to be done in an operating theatre or may still

be achieved with the ED practitioner taking responsibility for safe sedation and a relevant proceduralist undertaking the procedure.[5] One clinician must remain responsible for, and focus on, safe PS, whether or not the procedure is undertaken by an ED clinician or another specialist.

Although serious adverse events are rare,[6–9] sedation providers should be prepared to intervene should complications occur. Cote[10] and Hoffman[11] demonstrated that patient selection, adherence to a prescribed process and minimisation of the number of sedating agents were important in reducing PS risks. Sedation registries have provided data accumulated over many thousands of sedation events. The Pediatric Sedation Research Consortium (www.pedsedation.org) in the United States receives data from over 30 centres that perform PS. No deaths, one cardiac arrest (requiring CPR) and one aspiration event were reported by Cravero et al.[12] in a cohort who represented 30,000 PS episodes. Unplanned intervention was needed in 336 PS episodes (111.9 per 10,000 PS episodes), with bag-valve-mask ventilation (63.9 per 10,000 PS episodes) and intubation (9.7 per 10,000 PS episodes) the most invasive.

The phases and the tasks involved in PS can be divided into pre-procedure, intra-procedure and post-procedure (Fig. 20.2.1). This division provides a framework to build one's practice upon.

Pre-procedure

Governance

Governance structures that include guideline development, education and credentialing improve practice in PS[3,13] with a reduction in adverse events.[10] This is the foundation for safe PS practice and should occur before any child is sedated. In Victoria, Australia, over the last decade, the development and dissemination of a PS programme[13–16] (handbook, presentations for trainers, practical train-the-trainer sessions and testing materials) have provided a structure for teaching and providing practical experience. Hospital networks and EDs all play a role in the establishment and maintenance of PS education and procedural systems.

Patient selection and risk assessment

Appropriate patient selection reduces the risk of PS. Assigning risks as potential

threats to airway, breathing, circulation, neurological and other factors can assist in developing a structure to assessment (Table 20.2.1). Sedation risk is higher in younger children. Those under 12 months, and especially infants under 6 months, have an increased risk of apnoea.[6,7] Airway interventions, such as bag-valve-mask ventilation and/or endotracheal intubation, if needed, may be more complicated in younger infants as well.[17] Intercurrent illness such as active asthma and upper respiratory tract infection can complicate general anaesthesia and may also have an adverse effect on PS.

As with any ED attendance a history of current medications and allergies is required. Physical examination must document patient weight, vital signs, an assessment of conscious state, airway evaluation (e.g. Malampati score) with assessment of cardiac and respiratory systems. A Malampati score of Class 3 or 4 suggests more difficult bag-valve-mask ventilation and risk of airway obstruction.[5] The Malampati score requires a certain degree of cooperation that may not be possible in younger infants.

Fasting

The association between fasting time, vomiting and aspiration is not well supported by evidence, although fasting remains a powerful principle in anaesthesia. Babl[18] noted vomiting in 162 of 2002 (8%) children undergoing PS with either nitrous oxide or ketamine, while Cravero et al., reporting on the pediatric sedation research consortium data, identified 142 vomiting events out of 30,037 PS (0.47%) episodes.[19] Bellolio et al., as part of a meta-analysis, reported 498 vomiting events out of 7865 (6.33%) PS episodes.[6] Given conflicting reports, a pragmatic sedation plan will recognise the patient's medical, surgical and anaesthetic history (where relevant), the nature of the last meal eaten and the urgency of the procedure required.[20]

Consent

Informed consent should include general risks associated with PS and the procedure being undertaken. In addition, the specific effects of the chosen sedation plan must be discussed. For example, if ketamine were the chosen pharmacological agent, discussion regarding vomiting, hyperlacrimation, hypersalivation, hiccups and emergence phenomena should be initiated. Pre-prepared information sheets for specific, and common, PS plans can be useful.

Equipment and personnel

The staff involved in a PS episode must be competent in the procedure being performed and in the management of children undergoing sedation. Depending on the sedation plan, a minimum of two practitioners should be involved. One, ‘the sedationist’, is responsible for the sedation, monitoring (especially cardiac and respiratory status) and documentation of vital signs during PS.[21] The other, ‘the proceduralist’, will be responsible for the procedure. If parenteral sedation is utilised it is prudent to have a third practitioner, to document events or assist with the procedure or the PS. In many EDs the supervising senior doctor must be notified of the procedure and the PS so that in the event of complications they can become involved.[13,14,16] Depending on the involved practitioners, the patient and the procedure, they may not be at the bedside. On some occasions this person will, however, be undertaking the PS as well.

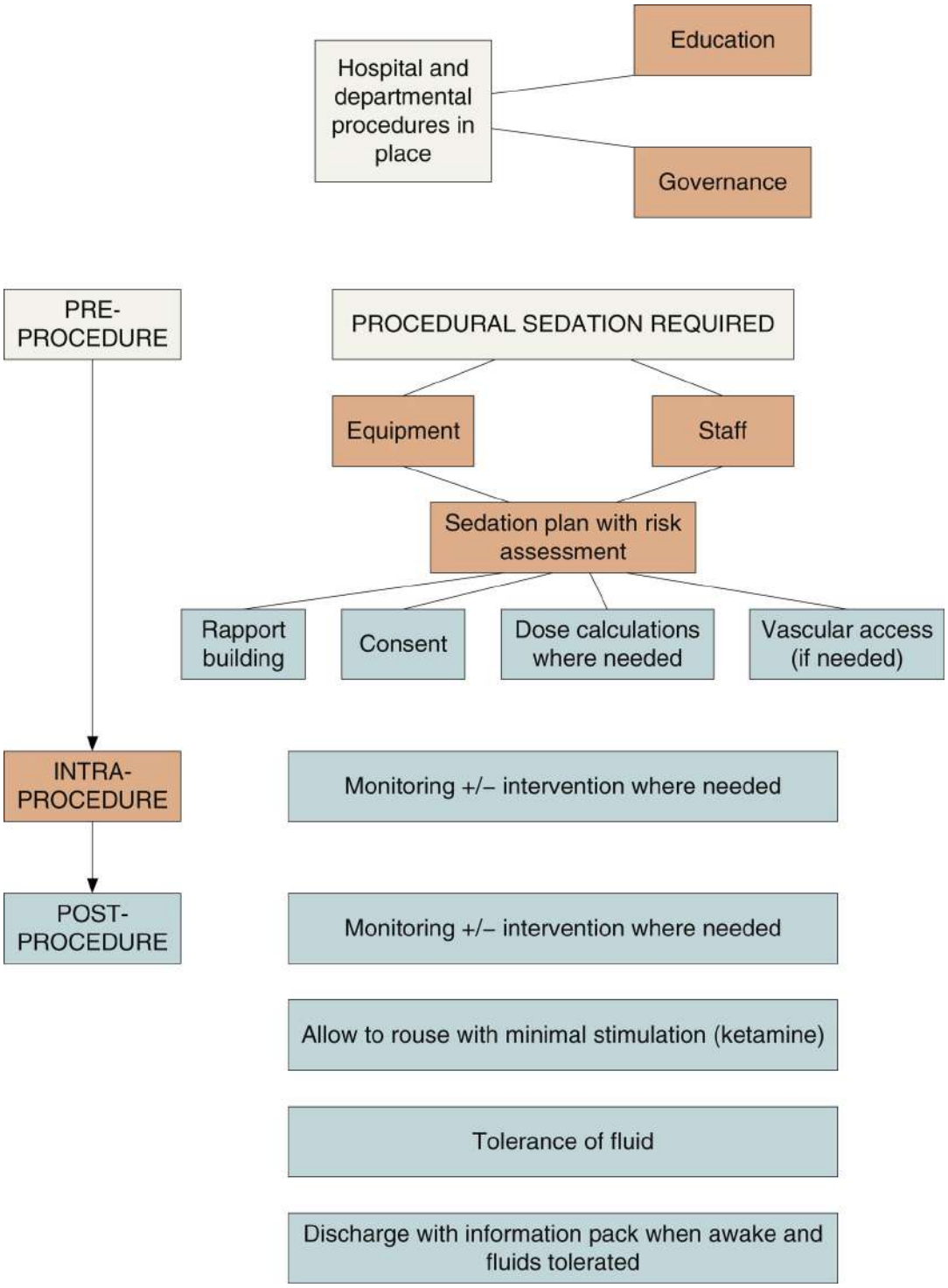

FIG. 20.2.1 The phases and tasks of procedural sedation. Krieser D, Kochar A. Paediatric procedural sedation within the emergency department. *J of Paediatr & Child Health*. 2016;52(2):197–203. Used with permission.

While intravenous (IV) access is desirable, it is not essential for safe PS.[21,22] IV access is certainly not required for nitrous oxide PS. Ketamine can be given

intramuscularly (IM), but a larger dose is needed, and sedation persists for longer and is associated with a higher incidence of vomiting.[23,24] If vascular access is not achieved prior to PS then the expertise and equipment to rapidly achieve access, including intraosseous needles, should be immediately available.

Rapport building

Developing rapport with paediatric patients and their carers is a requirement for most clinical work in paediatric emergency medicine. In PS, good rapport may mean that less pharmacological sedation is required, as the child feels less threatened. The use of distraction throughout the PS episode with items such as books, bubbles, screens (via phones, tablet computers, or televisions) or music is helpful. Prior to the procedure, a developmentally appropriate discussion should occur with the child. Play therapy,[25] guided imagery and meditation have been used to reduce pharmacological doses. Caregivers should be provided with instructions in what to say and what not to say,[26] avoiding bribery and statements that suggest the procedure is ‘nearly finished’ or that ‘it doesn’t hurt’. For nitrous oxide delivery, the use of commercially available food essences (e.g. strawberry, chocolate or orange essences) makes the use of the mask more acceptable and provides some choice where the child may perceive few choices. It also provides another distraction trigger, where the PS provider can ask, ‘Can you smell the strawberries?’ or ‘I also like chocolate’.

Intra-procedure

Real-time monitoring of airway, breathing and circulation are the mainstays of safe PS. Identified abnormalities should trigger appropriate responses from trained personnel. Equipment must be available for real-time monitoring of heart rate, respiratory rate, oxygen saturations and blood pressure (if parenteral drugs are utilised). Nasal cannulae that can monitor expired CO_2 can be useful as a rise in expired CO_2 will identify hypopnea prior to the development of hypoxia.[11,20] It is not clear if this translates to safer PS[27,28] and currently is not available in all EDs. Equipment to manage threats to airway, breathing and circulation must be available for all sizes of children. The necessary equipment should be prepared and drug doses calculated prior to the commencement of sedation.

Sedation depth can be monitored and documented; however, variations in sedation depth can be difficult to control along the continuum of sedation. This

varies by pharmacological agent and will be discussed later in the text.

Sedation can be reduced as the procedure nears conclusion. Judgment is required to balance the need for comfort for the remaining elements of the procedure with the duration of action of the pharmacological agent used. Local anaesthetic application or infiltration in laceration repair can potentially reduce the need for additional systemic PS. Nitrous oxide concentration can be weaned progressively, if possible. Rapport building prior to the commencement of the procedure may also reduce the need for additional systemic PS.[25]

Post-procedure

No patient wants to remain in the ED longer than the minimum required period, and there are quality indicators that value shorter lengths of stay. That said, following PS, a period of observation is needed.[21,29] As children emerge from pharmacological PS, vomiting may occur. Emergence dysphoria is well recognised following ketamine PS. Dysphoria can be reduced following PS by optimising the environment: dimming of lights, playing familiar music and allowing only familiar voices (i.e. family members only) while minimising medical procedures. Pharmacological treatment with a benzodiazepine, while rarely required,[29] could be used in this situation.

Table 20.2.1

Factors to consider in assessing risk for procedural sedation

	Active medical problems	Past history
Airway	Croup Foreign body Head and facial trauma	Previous airway surgery Laryngomalacia Craniofacial abnormalities Risk of vomiting (e.g. bowel obstruction, gastro-oesophageal reflux)
Breathing	Respiratory tract infection (e.g. pneumonia, bronchiolitis) Asthma	Sleep apnoea
Circulation	Shock (e.g. hypovolaemia, sepsis) Arrhythmia	Congenital cardiac disease (e.g. cardiac failure, pulmonary hypertension)
Neurological	Altered level of consciousness (seizure, meningitis, trauma) Space-occupying lesion	Unstable epilepsy Neuromuscular disease
Other	Unstable psychiatric disorder	History of sedation failure History of anaesthetic reaction Family history of anaesthetic reaction

Modified from Paediatric Procedural Sedation; ED Manual. Victorian Department of Health (ECIICN, Royal Children's Hospital, VMIA) 2013 and Daud YN, Carlson DW. Pediatric sedation. *Pediatr Clin North Am* 2014;**61**(4):703–17.

A return to pre-sedation level of consciousness is not essential for discharge, but the child should be near these levels. Vital signs should be in the normal range and documented. Persistent vomiting may preclude early discharge, with a need for longer monitoring periods and/or treatment with antiemetics and fluids.

The provision of written discharge instructions has been demonstrated to improve post-discharge care in a variety of situations,[30] and following PS such instructions should be given to families. Information should include hospital contact phone numbers, recommended levels of activity following PS, dietary guidelines and advice on when to return to the ED. Following PS it may not be appropriate to undertake activities requiring high levels of coordination (e.g. bike riding, climbing on monkey bars) for 24 hours.

Non-pharmacological methods

A balanced multidisciplinary approach using pharmacological and non-pharmacological strategies is essential to providing optimal analgesia and sedation for children.

Non-pharmacological techniques can be particularly useful in pain management (whether or not medications are used as well) as they are free of side effects and may be utilised before, during and after painful procedures.

The planning of procedures for children in the ED should include age-appropriate psychological interventions, such as distraction techniques. Involvement of play therapists or any additional team member to facilitate distraction techniques can prove very useful. Better procedure outcomes are reported for children and carers provided with age-appropriate distraction and the reduction of perceived pain, observed pain and situational anxiety.[31–37]

A child's anxiety and cooperation are affected by age, anxiety of the parent and previous medical experiences. The child and parent should be consulted about previous experiences and consideration of distraction techniques to use. Familiarisation with some equipment may help–for example, showing the child the mask to be used prior to nitrous use or asking the child to choose a 'flavour' to be placed into the mask. An informative and collaborative approach with a clear plan between all parties often produces smoother procedures.

Some useful non-pharmacological strategies that are usually used in

combination are outlined in Box 20.2.1.

Selection of agents by procedure and age

Parental preparation to obtain informed consent and provide support to the child for the procedure is essential. Discussion with the parents should include expected effects of the medication on their child and options for escalation or cessation if the procedure does not provide adequate sedation. Non-pharmacological techniques should always be utilised in synergy.

Box 20.2.1 Non-pharmacological strategies during procedures

Environment

- Calm, friendly staff (verbal and non-verbal communication) and atmosphere
- Parental/carer preparation and presence
- Age-appropriate communication
- Comforter (favourite blanket/soft toy)

Distraction techniques

- Visual, e.g. TV, electronic hand-held device, projected visual images, large story book, distraction cards, virtual reality, bubbles
- Oral, e.g. music, storytelling, singing children's songs, guided imagery
- Olfactory, e.g. flavour smell in mask, aromatherapy
- Activity, e.g. electronic game, search and find story book, arts activity

Physical

- Comfort swaddling (infants)
- Position of infant on parent so unable to see procedure, e.g. arm tucked under parent's side for IV cannulation
- Massage/rubbing
- Vibration or cold temperature to pain distract, e.g. Buzzy® for injections

- Oral sucrose/breast-feeding/pacifier for infants <6 months

Focused therapies

- Focus on breathing or encouragement of deep, relaxed breathing
- Muscle relaxation
- Hypnosis
- Biofeedback
- Acupuncture

Pharmacological methods

The choice of sedation agent is dependent on multiple factors including procedure type, duration, painfulness of procedure, fasting status and contraindications. Commonly used individual agents are discussed below. For many of these drugs coadministration of other drugs, such as opiates or benzodiazepines, may be used, but this also increases the risk of adverse effects. The specific combination of ketamine and propofol (ketofol) is discussed separately below.

Nitrous oxide

Nitrous oxide mixed with oxygen is readily available and frequently used in many Australasian EDs.[39] Its rapid onset and offset combined with analgesic and sedative properties make it ideal for short painful procedures, such as IV cannulation, bladder catheterisation, burns dressings and small wound repair. It can be safely used in procedure rooms with oxygen saturation monitoring and can be administered by nursing staff.

A fixed concentration (Entonox [50% N_2O:50% O_2]) is available with a demand-valve system making it less suitable for small or uncooperative children. The other available system uses a blender to titrate the N_2O:O_2 mix from 30% to 70% N_2O. In older children a mouthpiece allows autonomy for inhalation, whilst in a younger child a face mask is applied.

Vomiting is common (2.6%)[7] with nitrous oxide; however, there is no relationship between fasting status and emesis nor premedication with

antiemesis.[40] It also has only a mild sedative effect with few children achieving moderate to deep sedation levels even with 70% inhaled N_2O.[41]

Where there is a risk of gas expansion nitrous oxide is contraindicated. Examples of this include bowel obstruction, pneumothorax, pneumocephalus and a current acute asthma exacerbation.

The addition of opioids, such as intranasal fentanyl, increases the sedation depth but also increases the risk of vomiting.[7]

Ketamine

Ketamine is a unique, dissociative anaesthetic with analgesic properties. It provides moderate to deep sedation with preservation of airway reflexes and respiration. In addition, patients develop a relative tachycardia and hypertension rather than the hypotension more commonly seen with other sedatives. It is good for procedures requiring deeper or a longer duration of sedation, e.g. for fracture reduction or complex suturing. Patients often will have non-purposeful movements and hypertonicity of muscles making it less useful for procedures requiring the patient to remain still, such as CT, or in joint reductions where hypertonicity may make the procedure more difficult.

Ketamine can cause vomiting, although pre-dosing with ondansetron reduces the risk.[42] Transient laryngospasm and respiratory depression are less common.[43]

Emergence phenomenon includes agitation on waking, hallucinations or bad dreams. Whilst prophylactic benzodiazepines may reduce this in adults[44] there have been no positive studies for use in children. Strategies to reduce emergence phenomenon include slow IV administration of ketamine, dimmed lights, minimising sensory stimuli at the start and end of the procedure, and using guided imagery (imagining being on holiday, in a happy place, with pets, etc.) at drug administration. A small dose of midazolam can be given if the child is distressed and unable to be calmed by non-pharmacological methods.

Ketamine dosing is 1–1.5 mg kg IV as initial dose with 0.25–0.5 mg kg boluses during procedure if required. Ketamine can also be given IM as a dose of 3–4 mg kg. This has a slower onset and delayed offset and increased rates of emesis[23,24] but is safe and can avoid the additional distress of cannulation. If further doses are required during the procedure a cannula can be sited during the procedure.

Ketamine has been contraindicated in patients with raised intraocular pressure. Wadia et al. recently showed that most children do not have a clinically

significant rise in intraocular pressure during ketamine PS.[44] The patients selected in this study had no eye injury or risk factor for raised intraocular pressure, so application to patients with risk factors has yet to be studied. Traditionally ketamine has been contraindicated in patients with head injury due to the potential risk of its sympathomimetic effects increasing intracranial pressure (ICP). Studies of ketamine-sedated patients with ICP monitoring do not demonstrate rises in ICP, suggesting that the risks are less than initially thought.[45] Other contraindications include young age (<3 months absolute, <12 months relative), porphyria, seizure disorder, and intercurrent respiratory tract infection.

Propofol

Propofol is an ultrashort-acting sedative anaesthetic agent, with antiemetic properties, that is useful for procedural sedation due to its deep sedation, rapid onset and short duration of action. It does not have any analgesic properties so requires coadminstration of analgesic agent for painful procedures. Propofol is a good choice to facilitate radiological investigations. Hypotension (15.4%), desaturation (9.3%) and apnoea (1.9%) are more frequent than ketamine, necessitating access to a resuscitation cubicle with skilled and experienced clinicians. These side effects are usually dose dependent and short lived but may require intervention, such as fluid boluses, airway positioning and bag-valve-mask ventilation, to avoid adverse sequelae.

Hypotension and respiratory depression are increased with coadministration of opiates or benzodiazepines. Despite initial concerns about its use in the ED, propofol has been shown in multiple studies that it can be safely used.[46] Its very short duration of action allows patients to recover quickly, facilitating early ED discharge post procedure. An initial dose of 0.5–1 mg kg with additional titrated doses allows safe procedural sedation.

Ketafol

The combination of propofol with ketamine has emerged as an option for PS. The side effect profiles of each agent theoretically may cancel each other out.[47–49] In addition, lower doses of each may be required, which may further reduce the potential side effects. The combination has been shown to have improved user satisfaction scores compared to ketamine or propofol alone,[47,48] decreased

vomiting compared to ketamine alone[47] and decreased respiratory depression compared to propofol alone.[49] However, ketafol requires the careful balancing of two agents, and as either propofol or ketamine alone is safe there is no need to complicate dosing. A 1:1 dose ratio decreases total drug use with a starting dose of 0.5–1 mg kg of both propofol and ketamine. Further doses are titrated to effect.

Other agents

Midazolam has variable absorption and effects when given orally or intranasally and can cause paradoxical excitement in 10% of children. IV administration provides more reliable sedation, but its prolonged effect and risks of respiratory depression and apnoea combined with safer, more reliable agents means its use is not preferred.

Methoxyflurane, an inhalational agent used by ambulance services, has also shown to be safe and effective in the ED setting and is available with an activated charcoal scavenging system (Penthrox).[50]

Chloral hydrate is used in the paediatric ICU and in radiology, but no ED-specific studies have been reported. It may be used to facilitate radiological investigation in the child under the age of 18 months.[51] The achievable depth of sedation is unreliable and following administration often leads to a prolonged recovery.

Balanced sedation

A balanced approach to PS is recommended considering the four parameters of sedation, analgesia, amnesia and motion control. Table 20.2.2 provides some suggestions with regards to analgesia or sedation, which may be useful for common procedures within a paediatric ED. A child's developmental level and communication ability should be determined prior to any procedure to enable adequate assessment of pain and efficacy of sedation. Non-pharmacological methods (see Box 20.2.1) should always be used in conjunction with pharmacological techniques, with child and parental preparation essential, where possible, prior to any procedure.

Table 20.2.2

Suggestions for procedural analgesia and sedation

Agent	Mode of administration	Dose	Onset/duration of action	Adverse reactions	Notes
Nitrous oxide	Inhaled	50:50 mix (ENTONOX) 30–70% titrated to effect	Rapid onset and offset (minutes)	Vomiting	Usually provides mild sedation only Can be administered by trained nursing staff Use food essences to scent mask
Ketamine	Intravenous	1–1.5 mg kg initial dose 0.5 mg kg further doses	Onset: 1 minute Duration: 5–10 minutes	Vomiting Emergence phenomenon laryngospasm	Slow IV administration, minimising tactile stimulation and guided imagery to reduce emergence phenomenon
	Intramuscular	3–5 mg kg	Onset: 5–10 mimutes Duration: 20–30 minutes		
Propofol	Intravenous	1–2 mg kg	Onset: 1 minute Duration: 5–10 minutes	Respiratory depression Hypotension	No analgesic properties
Ketafol	Intravenous	0.5 mg kg of each initial dose Titrated additional doses	Onset: 1 minute Duration: 5–15 minutes	Respiratory depression Hypotension	

Table 20.2.3

Suggested analgesics and sedatives by procedure

Procedure	Analgesia	Sedation and considerations
Venepuncture IV cannulation	AnGel®/EMLA® Sucrose <6/12 Cold spray	Nitrous oxide
Nasogastric tube insertion	Sucrose <6/12 Topical cophenylcaine Forte®	Need to maintain airway reflexes at all times (unless intubated)
Urinary catheter	Sucrose <6/12 Lidocaine (lignocaine) gel	Nitrous oxide
Suprapubic aspiration	Sucrose <6/12 AnGel®/EMLA®	Only infants <12 months
Lumbar puncture	Sucrose <6/12 AnGel®/EMLA® Subcutaneous local anaesthetic infiltration	Nitrous oxide
Small laceration	Laceraine® (adrenaline [epinephrine], lidocaine, amethocaine mixture) Subcutaneous local anaesthetic infiltration	Nitrous oxide Tissue adhesives
Complex wound	Intranasal (IN) fentanyl Subcutaneous local anaesthetic infiltration Regional anaesthetic block	Nitrous oxide Intramuscular (IM)/intravenous (IV) ketamine May need general anaesthetic
Burn dressing	Initial – cold running water (20 minutes) Initial – Burnshield® IN fentanyl	Nitrous oxide Use non-stick dressings, e.g. Mepitel®/Mepilex®
Foreign body removal	AnGel®/EMLA® Laceraine® Subcutaneous local anaesthetic infiltration	Nitrous oxide IM/IV ketamine May need general anaesthetic

Fracture/manipulation	Splint early IN fentanyl IV morphine IV regional anaesthetic, e.g. Bier block	Nitrous oxide IM/IV ketamine IV propofol May need general anaesthetic
Joint dislocation (excluding pulled elbow)	IN fentanyl IV morphine	Nitrous oxide IM/IV ketamine IV propofol May need general anaesthetic
CT scan	None usually required	Wrapping and settling techniques for infants. Communication/distraction for older children Per os (PO)/IV benzodiazepine Chloral hydrate
MRI scan	None usually required	PO/IV benzodiazepine Chloral hydrate May need general anaesthetic
Multi-trauma	IN/IV opiates	Child-centred communication May need general anaesthetic for investigations

Angel and EMLA, topical local anaesthetic (cream based); Laceraine, topical wound anaesthetic (liquid placed onto cotton wool then on wound); Burnshield, topical Ti Tree (Melaleuca) dressing only for initial dressing.

Controversies and Future Direction

1. Sedation registries have demonstrated trends in adverse events and have potential for improving sedation safety and practice through research. Such registries require precise data definitions, funding for data entry and management and formal partnerships (e.g. research ethics and data-sharing agreements) across health services, state and national boundaries. In addition, new methods of sedation drug delivery will require research. Existing medications may be found to be effective if delivered in novel ways. New medications, with different pharmacodynamics and pharmacokinetic profiles, will require assessment before widespread use. A sedation registry could facilitate such developments.
2. Monitoring of PS with oximetry, heart rate and blood pressure is well established. Future research will assist in identifying the role of end-tidal CO_2 measurement and bispectral analysis (including EEG monitoring of the level of sedation) in improving safe sedation practice.
3. The non-pharmacological techniques used in PS are often dependent on

the individual. Is there a role for simulation or simulated patients for education regarding rapport building, age appropriate interaction and other skills that would facilitate rapport building?

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SECTION 21

Poisoning

OUTLINE

21.1

General approach to poisoning

Naren Gunja

ESSENTIALS

1 Poisoning in children is usually accidental, particularly in the under-6 age group. Deliberate self-poisoning may become apparent as they mature into teenage years.

2 Poisoning in most children runs an uneventful course, and emergency department (ED) observation is often the only management needed.

3 The potential for non-accidental poisoning (either deliberate or due to neglect) should be considered, particularly in children under the age of 1 year. Where non-accidental poisoning is suspected, the child should be referred to the relevant child protection authorities.

4 A focused history and examination should lead to a risk assessment on the likely outcome, and worst-case scenario.

5 Advice regarding management of poisoned children can be sought from Australasian Poisons Information Centres (Australia: 13 11 26; New Zealand: 0800 764 766).

6 Poisoning in children manifests clinically in a similar manner to adults. The management of poisoning in children is also similar.

7 Gastrointestinal decontamination is not necessary in the majority of cases.

8 Parents and carers should be advised to keep medicines and chemicals away from the reach of children – medicines should be stored in locked containers or cupboards at a height of at least 1.5 metres.

Introduction and epidemiology

In 2013, the NSW Poisons Information Centre received in excess of 40,000 calls from around Australia regarding paediatric exposures to pharmaceuticals, chemicals, plants and animals. There is a bimodal distribution in the frequency of exposures, with the larger peak occurring in the toddler age group (ages 1–3 years) and a much smaller peak in the mid-to-late teens. The latter peak relates to deliberate self-poisoning in adolescents. Nearly 80% of poison centre calls relating to childhood exposures are advised to stay at home as no acute management is necessary. Pharmaceuticals are by far the commonest exposure in children as per American Poison Control Center data. The top ten unintentional exposures in children (under the age of 18 years) reported to Australian Poisons Information Centres are listed in Table 21.1.1. It is important to note that paracetamol is present in many preparations as well as in combination products (e.g. with codeine, pseudoephedrine, doxylamine, dextromethorphan).

Table 21.1.1

Top 10 exposures in children

No.	Agent
1	Paracetamol
2	Ibuprofen
3	Household cleaning agents
4	Detergents
5	Dessicants (e.g. silica gel)
6	Cough and cold preparations
7	Light sticks/glow toys
8	Hand sanitiser
9	Essential oils (e.g. eucalyptus oil)
10	Bleach (containing hypochlorite)

Diagnosis

As opposed to overdose in the adult population, exposures in children are nearly always accidental or unintentional. The circumstances around the exposure or

ingestion are often unknown or difficult to elucidate. Parents and carers are usually uncertain about time of exposure or dosage of drug ingested. As such, the clear history required to make an accurate risk assessment is difficult or sometimes impossible. Regardless of whether the entire history and circumstances surrounding the exposure are available, it is prudent to plan for a ‘worst-case scenario’, assuming maximal exposure.

Important elements of the focused history include:

- age and gender of child
- agent involved – drug, chemical or plant
- approximate time of exposure
- dose ingested, including maximum possible ingestion
- circumstances around exposure
- symptoms, in particular vomiting
- first aid and pre-hospital management.

Deliberate self-poisoning in adolescents warrants further enquiry into previous ingestions, pre-existing psychiatric illness and management, drug use and social circumstances. In cases of unknown drug exposure, it is important to explore the availability of pharmaceuticals and/or chemicals to which the child may have had access. Plant and mushroom ingestion is common in children and needs to be considered in the acutely unwell child who has been outdoors.

Non-accidental (or deliberate) poisoning of a child requires mandatory reporting to child protection authorities in all jurisdictions within Australia. The index of suspicion is higher in children under the age of 1 year, or where the circumstances of the exposure do not fit the capabilities of the child in question. Rare cases of fabricated or induced illness by carers (formerly known as Munchausen’s syndrome by proxy) are also reported in the literature, involving deliberate poisoning of children by their parent/carer.

Physical examination of the potentially poisoned child is usually unremarkable, particularly in asymptomatic children or in the early stage of emergency department (ED) presentation. However, in children presenting with symptoms or patients with altered level of consciousness, a thorough physical examination is vital. Key elements of the toxicological examination include:

- vital signs: heart rate, blood pressure, temperature, respiratory rate, oxygen saturation

- odour suggesting intoxication or poisoning
- airway patency and adequacy of ventilation
- cardiovascular status and end-organ perfusion
- level of consciousness or altered mental status, presence of delirium or psychosis (including blood glucose level)
- neurological signs: abnormal tone, reflexes, clonus, seizures
- external signs of trauma, bruising, bite marks
- saliva or vomitus, pill fragments.

Children may also present with a cluster of symptoms and signs suggestive of poisoning, i.e. a toxidrome. Although most cases do not manifest the full spectrum of signs and symptoms, pattern recognition amongst clinicians may provide a clue to diagnosis. Toxidromes and corresponding causative agents commonly seen in children are listed in Table 21.1.2.

Risk assessment

Following the history and examination of the potentially poisoned child, the clinician must undertake a risk assessment of the likely exposure and probable course of toxicity, if any. This requires knowledge of the toxicodynamics and kinetics of the agents, an understanding of potential complications and experience with previous similar cases. At this stage, it is prudent to obtain advice from expert clinicians in paediatric toxicology, such as through the Australasian Poisons Information Centres (Australia: 13 11 26; New Zealand: 0800 764 766). These centres are available 24/7 and provide expert advice on the assessment and management of poisoning in children. The majority of paediatric exposures do not present to hospital, and even those that do, require observation only. However, there are a few highly toxic pharmaceuticals and chemicals that, even in small doses, can cause severe toxicity. Patients exposed to these select few agents may require close monitoring and potentially aggressive resuscitation – these are discussed in Chapter 21.2.

Table 21.1.2

Common toxidromes

Toxidrome	Agents	Clinical features
Sympathomimetic	Amphetamines	Tachycardia

	Pseudoephedrine Caffeine	Hypertension Mydriasis Sweating Agitation Delirium Fever
Anticholinergic	Atropine Hyoscine Antihistamines Plants Mushrooms	Tachycardia Mydriasis Loss of visual accommodation Flushed skin Dry skin/mouth/eyes Fever Delirium
Opiate	Opiates Tramadol Clonidine	Sedation Respiratory depression Hypotension Miosis
Cholinergic	Organophosphates Carbamates	Delirium Coma Seizures Excess secretions (DUMBELS) Weakness Fasciculations
Serotonergic	SSRIs Cyclic antidepressants Opiates Tramadol Lithium MDMA (ecstasy)	Delirium/agitation Hyperreflexia Hypertonia Tremor Clonus Diaphoresis Fever
Neuroleptic	Anti-psychotics (e.g. olanzapine)	Fever Hypertonia Muscle rigidity Rhabdomyolysis Delirium Diaphoresis

SSRI, selective serotonin reuptake inhibitor.

Investigations

The vast majority of children exposed to a substance require no investigations at all. There are some instances where a specific agent is ingested and a specific investigation may aid diagnosis and/or management. Screening tests in the poisoned child should be performed based on the risk assessment. Specific investigations, which may be invasive or time-consuming to return a result, should be discussed with an expert toxicologist prior to embarking on these tests. Table 21.1.3 summarises the potential indications for screening and specific investigations in paediatric poisoning. Baseline blood investigations (blood

counts, electrolytes, renal function test) should be performed if any of the screening or specific laboratory investigations are ordered.

Resuscitation

In the severely poisoned child, timely and effective resuscitation is the key to better outcomes. Thankfully, this scenario is uncommon in Australasia. Resuscitation of the poisoned child should follow standard advanced paediatric guidelines with regards to promoting haemodynamic stability, preventing secondary brain injury and best practice supportive care.

The majority of cases require no more than oxygen therapy and intravenous fluid boluses. In the presence of coma or cardiovascular collapse, resuscitation involves airway protection and ventilatory support as well as the potential use of agent-specific antidotes. In cases where early decontamination is required, such as exposure or ingestion of chemicals, decontamination should occur concurrently with, and not to the detriment of, active resuscitation.

Decontamination

Decontamination involves the removal of a toxic substance to which a child has been exposed in order to minimise its absorption into the systemic circulation. In a child with dermal, eye or mucosal exposure to a substance, decontamination simply involves removal of the toxic substance and irrigation or washing of the contaminated skin, eye or mucosa. Inhalational injury to toxic fumes or gases should include the removal of the patient from the source of exposure and, if necessary, administration of supplemental oxygen. In extreme situations, these patients may need advanced airway and ventilatory support. The use of universal precautions (gown, gloves, goggles) by staff during the process of decontamination is sufficient for the vast majority of poisoning situations, including hydrocarbons and organophosphate insecticides.

Table 21.1.3

Investigation of the poisoned child

Investigation	Potential toxicological indication(s)
Screening tests	
Blood glucose level	Altered mental status Deliberate self-poisoning All exposures to insulin or oral hypoglycaemic agents
β-HCG pregnancy test	Female patients of childbearing age presenting with overdose
Screening paracetamol level	Deliberate self-poisoning (of any substance)
Urine drug screen	Known or suspected exposure to illicit substances/drugs of abuse Altered mental status (delirium, psychosis, coma)
ECG	Heart rate outside normal parameters for age Haemodynamic instability or shock Poisoning with specific agents: Cardiovascular drugs Sedatives Neuroleptics Antidepressants Amphetamines and sympathomimetics Clonidine and baclofen Metals (e.g. potassium, lithium, iron)
Blood gas measurement	Known or suspected acid–base abnormality Poisoning with specific agents: Salicylates Tricyclic antidepressants Ethanol Toxic alcohols Iron Isoniazid Carbon monoxide Cyanide Metformin Methaemoglobinaemia
Specific investigations	
Chest/abdominal X-ray	Radio-opaque tablet or foreign body ingestion Known or suspected amatoxic mushroom poisoning Tube (e.g. endotracheal, gastric) placement
Liver function tests	Known or suspected paracetamol poisoning Known or suspected amatoxic mushroom poisoning Suspected hepatotoxicity from any systemic poisoning
Coagulation panel	Snake bite Suspected coagulopathy from poisoning/envenoming Poisoning with specific agents: Paracetamol Anticoagulants (e.g. warfarin, rodenticides) Salicylates Iron and heavy metals
Paracetamol level	In all cases of paracetamol ingestion (incl. deliberate self-poisoning, accidental, supratherapeutic, chronic)
Specific drug levels	Known or suspected poisoning from: Anticonvulsants (carbamazepine, valproic acid, phenytoin) Aspirin/salicylates Digoxin Metals (potassium, lithium, iron) Ethanol Toxic alcohols (ethylene glycol, methanol) Methotrexate Theophylline Phenobarbital

Salicylate levels should not be routinely ordered as their screening value is negligible. Tricyclic antidepressant levels are not useful in the management of poisoning from these agents, nor are they a useful screening test. Rarely performed investigations for specific toxins include cholinesterase levels, carboxyhaemoglobin and methaemoglobin amongst others. CT of the brain is not routinely required in the comatose child with a reliable history of poisoning; it may be warranted when the history is unclear or there is suspicion of trauma or non-accidental injury.

HCG, human chorionic gonadotrophin.

Oral exposure to pharmaceuticals, chemicals or plants requires a risk-assessment-based approach as to whether decontamination is deemed worthwhile. The need for active oral decontamination in paediatric poisoning is rare. Clinicians are advised to seek expert advice prior to decontamination in children with oral exposures.

Syrup of ipecacuanha (derived from the root of a South American plant) is no longer recommended in the management of poisoning. The induced emesis does little to prevent drug absorption and potentially can cause a myriad of

complications including protracted vomiting, aspiration and oesophageal tears and haemorrhage. Gastric lavage (‘stomach pumping’), involving the injection of fluid into the stomach via a tube and aspirating gastric contents, is also a discontinued practice which has little or no role in the management of poisoned children. Whole bowel irrigation involves the administration of a polyethylene glycol solution through the gastrointestinal tract for the purpose of promoting tablet residue in effluent and thus preventing drug absorption. It is reserved for specific ingestions such as sustained-release preparations or metals; expert advice should be sought prior to instituting whole-bowel irrigation.

Activated charcoal is a colloidal suspension of charcoal particles able to bind to most pharmaceuticals. Charcoal does not adsorb metals, hydrocarbons, corrosives or alcohols. Although the need for activated charcoal in children is uncommon, it is potentially indicated when a child ingests a highly toxic substance, which is bound by charcoal, and the charcoal can be administered within an hour post ingestion to an alert child (or in the case of intubated children, via a gastric tube). Clinicians should avoid inserting a gastric tube for the purpose of administering charcoal to a patient with altered level of consciousness. The presence of bowel sounds should always be confirmed prior to the administration of oral charcoal. When indicated, the activated charcoal dose is 1 g kg^{-1}. Expert advice should be sought regarding the use of charcoal in situations beyond an hour post ingestion or multi-dose activated charcoal (discussed below). Common side effects of charcoal administration include vomiting and the passage of black stools. Aspiration of charcoal can lead to chemical pneumonitis and potentially acute respiratory distress syndrome.

Antidotes

The need for agent-specific antidotes in children is uncommon. Knowledge of antidotes and their potential utility may be, in rare cases, life-saving. Table 21.1.4 lists selected antidotes used in paediatric poisoning and their indications. The use of these agents should be discussed with a toxicologist.

Table 21.1.4

Antidotes

Antidote or specific therapy	Dose	Potential indications
Acetylcysteine	1st: 200 mg kg^{-1} over 4 hours, 2nd: 100 mg kg^{-1} over 16 hours	Paracetamol poisoning Hepatotoxic mushroom poisoning Paraquat poisoning Patients at risk of, or with established, hepatotoxicity
Atropine	0.02–0.05 mg kg^{-1}, repeat every 5–10 minutes (use doubling regimen)	Known or suspected cholinergic toxidrome from organophosphate or carbamate pesticide poisoning. Titrate to heart rate, blood pressure and drying of chest secretions. Funnel-web spider envenoming (temporising measure)
Calcium	Calcium gluconate 10%, 0.6 mL kg^{-1} Calcium chloride 10%, 0.2 mL kg^{-1}	Calcium-channel blockers: Bradycardia or heart block Hypotension
Desferrioxamine	15 mg kg^{-1} h^{-1}	Iron toxicity: Serum iron >90 µmol L^{-1} Clinically severe systemic toxicity
Digoxin Fab antibodies	Empiric dose: Acute poisoning – 5 vials Chronic poisoning – 1–2 vials	Digoxin toxicity: Life-threatening arrhythmias Clinical signs of severe digitoxicity
Ethanol	PO/NG route preferred Loading dose: 600 mg kg^{-1} Infusion: 80–150 mg kg^{-1} h^{-1}	Known or suspected poisoning from toxic alcohols (ethylene glycol, methanol): Elevated osmolar gap Metabolic acidosis Maintain blood ethanol at 100 mg dL^{-1}
Flumazenil	0.005–0.01 mg kg^{-1} (max 2 mg)	Benzodiazepine poisoning: Reduced level of consciousness Bradypnoea N.B. Titrate to respiratory rate
Insulin (high dose + dextrose)	Initial dose 1 unit kg^{-1} Infusion: 1–2 units kg^{-1} h^{-1}	β-Blockers Calcium channel blockers: Bradycardia or heart block Hypotension
Naloxone	Bolus: 0.005–0.01 mg kg^{-1} (max 2 mg); Infusion: 0.01 mg kg^{-1} h^{-1}	Known or suspected opiate toxidrome. Titrate to respiratory rate
Octreotide	Bolus: 1 mcg kg^{-1} IV or SC Infusion: 0.5 mcg kg^{-1} h^{-1} IV	Sulfonylurea poisoning with recurrent hypoglycaemia
Physostigmine	0.02 mg kg^{-1} (max 0.5 mg)	Anticholinergic poisoning with delirium
Sodium bicarbonate	1–2 mmol kg^{-1} IV bolus (serum alkalinisation)	Sodium channel blocking agents (e.g. tricyclic anti-depressants): Cardiac arrest Wide-complex tachyarrhythmias Hypotension Seizures
Vitamin K	5–10 mg, PO 1–2 mg, IV or IM	Poisoning from warfarin (or other coumadin anticoagulants) with established, or risk of, coagulopathy. Note: higher doses may be required for poisoning from long-acting anticoagulant rodenticides.

IM, intramuscular; IV, intravenous; PO, per os; SC, subcutaneous.

Enhanced elimination

Techniques used to promote drug elimination from the body are employed in a limited number of poisonings. These methods include haemodialysis, multi-dose activated charcoal and alkaline diuresis. Various forms of haemodialysis methods are utilised in paediatric poisoning after insertion of a temporary vascular catheter. Agents that are potentially dialysable include potassium, salicylates, toxic alcohols, theophylline and carbamazepine, amongst others. Multi-dose activated charcoal is rarely used when promoting charcoal adsorption and elimination of poisons that undergo enterohepatic circulation, such as carbamazepine, phenytoin and colchicine. Urinary alkalinisation with sodium bicarbonate is indicated in salicylate poisoning to promote the renal excretion of salicylate ions. These techniques are seldom used in childhood poisonings and expert advice should be sought prior to their institution.

Supportive care

Active resuscitation of the severely poisoned child is paramount and should be followed by meticulous attention to supportive care in a high-dependency or intensive-care environment. Although specific poisoning scenarios are dealt with in the next chapter, the guiding principles of excellent supportive management of the poisoned child are likely to be more crucial.

All children with altered level of consciousness should have close glucose monitoring. Coma from drug overdose should be managed with advanced airway and ventilatory manoeuvres. In general, non-invasive ventilation does not have a role in the poisoned child. Drug-induced seizures from all causes should be treated with parenteral benzodiazepines as the first-line agent of choice. Phenytoin should be avoided as its sodium-channel-blocking properties may exacerbate the problem.

Cardiovascular collapse and asystole in the poisoned child should be managed as per standard advanced paediatric life-support guidelines. Drug-induced arrhythmias may warrant agent-specific strategies, such as antidotes. Wide QRS complex tachyarrhythmias, usually due to sodium-channel-blocking agent poisoning, should in the first instance be treated with boluses of sodium bicarbonate (1–2 mmol kg^{-1}). Drug-induced hypotensive shock may be due to central negative inotropy or peripheral vasoplegia; in these instances, echocardiography may assist in diagnosis and management.

Close monitoring and maintenance of normothermia, euglycaemia, acid–base balance and electrolyte levels are vital in the severely poisoned child. Other potential complications in this group of patients include rhabdomyolysis (seen in snake bite, prolonged coma), aspiration pneumonitis, renal impairment and persistent delirium (commonly due to anti-cholinergic drugs or plants).

Consultation and disposition

Poisoning in children, as in adults, is a symptom of an underlying issue, be it psychological, parental neglect or accidental access to harmful substances. The mainstay of management involves observation in the ED. Occasionally the child is exposed to an unusual substance or develops severe toxicity that requires input from clinical toxicologists. Clinicians should seek advice from local experts in the field and/or their local Poisons Information Centre.

The underlying issue or disease process also requires attention, such as mental health assessment or counselling. Children who are overdosed with analgesics or antipyretics may warrant investigation into the cause of pain or fever. In the case

of neglect or deliberate poisoning the child is likely to need referral to relevant child protection authorities. All carers involved in accidental poisoning should have counselling with regards to safe storage of medicines and/or chemicals in the home.

Further reading

Mowry J.B, Spyker D.A, Brooks D.E, et al. 2014 Annual report of the American Association of Poison Control Centers' National Poison Data System (NDPS): 32nd Annual Report. *Clin Toxicol*. 2015;53(10):962–1146.

NSW Poisons Information Centre Annual Report. Australia: Westmead, NSW: The Children's Hospital; 2013. www.poisonsinfo.nsw.gov.au.

21.2

Specific poisons

Naren Gunja

ESSENTIALS

1 Medically important paediatric poisoning usually falls into two categories: common ingestions and rare or dangerous toxins.

2 The emergency physician plays a crucial role in forming an appropriate management strategy based on the risk and knowledge of the relevant drug(s)' toxicity.

3 The management of common ingestions is essentially supportive, with the potential utility of a specific antidote. Gastrointestinal decontamination is rarely indicated in these cases.

4 Although most substances are harmless to children in small amounts, a few pharmaceuticals and chemicals are extremely toxic in small quantities – these substances are potentially lethal in a toddler even if only a few pills or mouthfuls are ingested.

Common poisons

Paracetamol

Paracetamol is by far the commonest paediatric poisoning presentation to the emergency department (ED), but the large majority will have no toxic effects. Absorption from the gastrointestinal (GI) tract is rapid, particularly with the liquid formulation (approx. 30 minutes). Most of the paracetamol is conjugated by the hepatic pathways of sulfation and glucuronidation to inactive metabolites, which are excreted in the urine. Children under the age of 9–12 years have a

more active sulfation pathway. Less than 5% is excreted unchanged by the kidney and 5–15% is oxidised by the hepatic cytochrome P450 enzyme CYP 2E1 to form a highly reactive intermediate metabolite – NAPQI (*N*-acetyl-*p*-benzoquinone imine), which binds to hepatocytes and leads to oxidative stress and cell death. With therapeutic dosing, NAPQI is metabolised to a non-toxic metabolite with glutathione as a sulfhydryl donor. In overdose, when glutathione reserves are depleted, NAPQI accumulates and causes hepatotoxicity. Acute toxicity from accidental ingestion is extremely rare in children. Paracetamol toxicity is more likely to be problematic in children taking standard doses of paracetamol chronically or in repeated supratherapeutic doses, rather than after a single ingestion.

Children appear less sensitive to the hepatotoxic effects of acute paracetamol overdose than adults. This may be related to metabolic differences, age-related clearance rates and to the increased propensity for children to vomit after acute paracetamol ingestion. The peak serum concentration is reached in <2 hours in the majority of children having a single ingestion of paracetamol elixir.

In the early phase, <24 hours, a child may be totally asymptomatic or complain only of mild abdominal pain, nausea and vomiting. Following a period of latency, hepatotoxicity progresses to multi-organ failure. Paracetamol can directly cause renal impairment and coagulopathy with prolonged prothrombin time. The hepatorenal syndrome can also complicate severe hepatotoxicity. Finally, fulminant hepatic failure may occur or the patient may enter the recovery phase with return to normal hepatic function within 4 weeks. Rare consequences of paracetamol toxicity include myocardial necrosis, haemolytic anaemia, methaemoglobinaemia, skin rashes and pancreatitis.

In acute overdose, a serum level should be obtained 4 hours post-ingestion in all children with potential paracetamol ingestions of greater than 200 mg kg^{-1}. Children with acute single ingestions of liquid paracetamol preparations are likely to have reliable post-peak levels earlier than 4 hours post ingestion – in these children, a 2-hour level above 200 mg L^{-1} suggests risk of hepatotoxicity. Other relevant investigations include electrolytes, renal function, liver function tests and coagulation panel.

The paracetamol nomogram (Fig. 21.2.1), based on adult toxicity profiles, is extrapolated to children and predicts the potential for significant hepatotoxicity. The nomogram is applicable when a paracetamol level is taken between 4 and 16 hours post-acute ingestion. The nomogram cannot be utilised when the time of ingestion is unknown or in the case of staggered or chronic ingestions. The

nomogram line commences at 150 mg L^{-1} at 4 hours post-ingestion and has a half-life of 4 hours.

Acetylcysteine, a glutathione precursor, is the antidote of choice in paracetamol poisoning. Acetylcysteine is indicated in the setting of acute paracetamol ingestion, where the measured paracetamol level falls above the nomogram treatment line. The traditional full course involves a loading dose of 150 mg kg^{-1} in glucose over 15–60 minutes, followed by a second infusion of 50 mg kg^{-1} over 4 hours, and finally a 100 mg kg^{-1} infusion over 16 hours. This regimen has recently been modified at several toxicology units in Australia to a 2-bag regimen (see Table 21.1.4), with increasing evidence of reduced adverse effects if acetylcysteine is administered slower (initial bolus infusion over 4 hours instead of 15–60 minutes). Acetylcysteine is preferably commenced within 8 hours of ingestion, but has been documented to be effective in adult patients when given up to 48 hours after a serious ingestion, and can even be considered later when hepatic failure is established. Anaphylactoid reactions, including rash, bronchospasm, pruritus, hypotension and tachycardia, occur in up to 15% of cases and are most common during the second infusion. These reactions are managed similarly to other hypersensitivity reactions; the acetylcysteine infusion should be temporarily ceased and recommenced at half the rate.

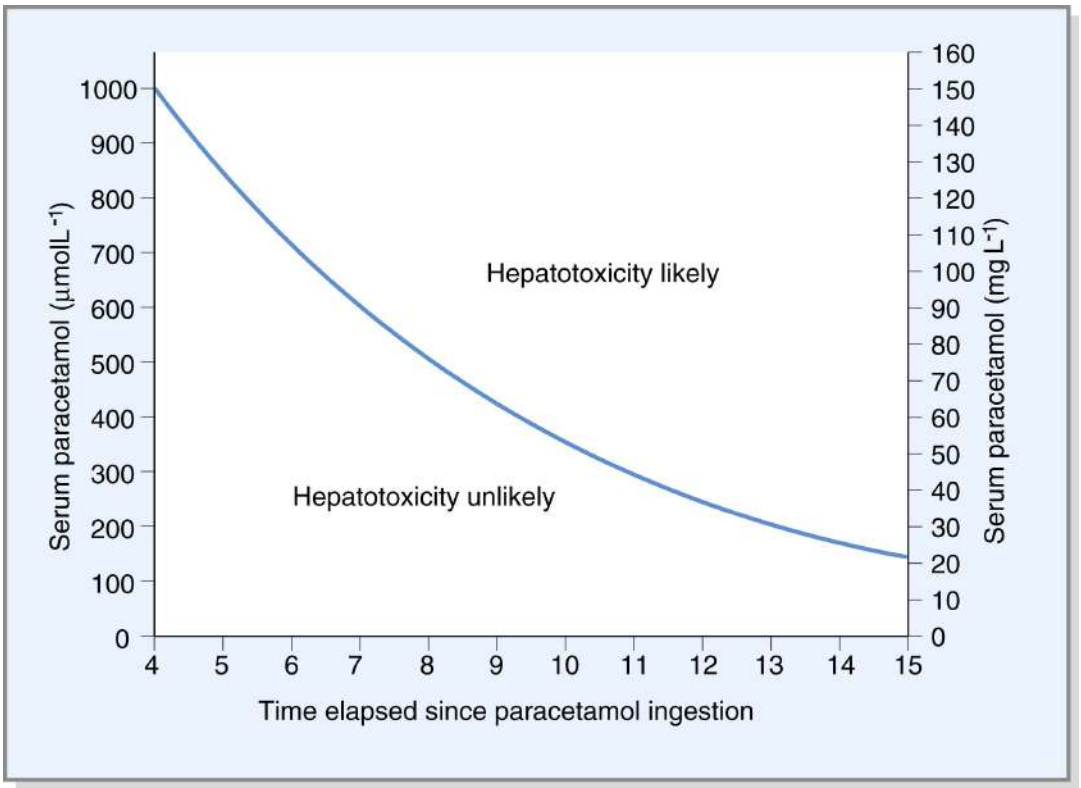

FIG. 21.2.1 Paracetamol nomogram.

As children usually ingest the elixir formulation rather than tablets, rapid absorption precludes the utility of oral activated charcoal. Activated charcoal may be indicated for potentially toxic doses of the tablet formulation, if given within 2 hours, or longer in massive ingestion or sustained-release preparations. Children who present with established hepatic failure must receive prompt resuscitation and stabilisation. Acetylcysteine is still indicated when hepatotoxicity is established. Coagulopathy and encephalopathy should be managed as from other causes of liver failure. These children are managed in the intensive care and should be assessed for potential liver transplantation.

Chronic over-dosing or repeated supratherapeutic ingestion is a significant problem. The treatment nomogram is not applicable to these situations and cases should be discussed with a toxicologist. Children with paracetamol levels below the nomogram treatment line after acute single ingestions can be cleared from a toxicological point of view. Parents or carers should be educated on correct paracetamol dosing and safe storage. Children with deliberate self-poisoning should be assessed by the mental health team.

Non-steroidal anti-inflammatory drugs

Of the many non-steroidal anti-inflammatory drugs (NSAIDs) available, some obtainable as over-the-counter medicines without prescription, ibuprofen is the most significant NSAID ingested by children. As a group NSAIDs are generally of low toxicity, producing GI upset, headache, dizziness, tinnitus and visual disturbance. Hypotension, tachycardia, hypothermia and coagulopathy have been reported when NSAIDs were consumed in large amounts. In massive overdose, electrolyte disturbances, metabolic acidosis, central nervous system depression and respiratory failure can occur.

Asymptomatic children can be discharged at 4 hours post ingestion. Symptomatic children require hospital admission. Oral fluids should be encouraged and dehydration corrected. Electrolytes, blood glucose level, renal function and acid–base status should be monitored. Activated charcoal may be indicated in massive ingestions that present early. Methods of enhanced elimination are not usually beneficial.

Benzodiazepines

Benzodiazepine overdose is commonly seen in toddlers who ingest 1–2 tablets or as part of a mixed overdose in adolescents. Benzodiazepines bind predominantly to the γ-aminobutyric acid (GABA) type-A receptor complex in the central nervous system (CNS) and enhance GABA activity to produce sedative, anxiolytic and anticonvulsant activity. The duration of sedation ranges from 4 to 36 hours, depending on the agent. Drowsiness, slurred speech and ataxia are the most common manifestations. This may progress to coma and hypotension, hypothermia and respiratory depression in more significant ingestions. Death is rare unless other CNS depressants have been co-ingested.

Management is entirely supportive. Hypotension usually responds to fluid administration. GI decontamination is not indicated in pure benzodiazepine poisoning. A blood glucose level should be checked in all children with an altered level of consciousness. Other laboratory investigations are not routinely indicated. Chest radiography is only indicated if aspiration is suspected. A qualitative urine test for benzodiazepines may provide reassurance as to the aetiology of drowsiness in the setting of an unconscious patient without a clear history of ingestion. In some clinical scenarios, flumazenil, a competitive antagonist at benzodiazepine receptors, may avert the need for intubation and mechanical support but its use should be discussed with a toxicologist. Flumazenil should be administered in boluses of 5–10 mcg kg^{-1} and titrated to

respiratory rate and effort.

Most children can be discharged after 4–6 hours if vital signs are satisfactory and the child can walk unaided.

Opioids

Opioids are frequently available where family members suffer chronic pain, abuse drugs or are on drug-rehabilitation programmes. Iatrogenic intravenous (IV) overdosing of children is commonly a result of 10-fold errors in dose calculation.

Morphine and codeine are natural opium alkaloids. Other opioids are synthetic or semisynthetic analogues. Opioids act on various receptors in the brain, spinal cord and GI tract as full or partial agonists. Opioids are well absorbed by all routes except skin, are metabolised by the liver and are renally excreted. Some opioids (e.g. pethidine and diphenoxylate) have potent metabolites. Opioids vary in their duration of action and some are available as sustained-release preparations (e.g. morphine). Toxicity is enhanced by co-ingestion of other sedative medications, which may be found in some cough remedies or analgesic preparations. Children are especially sensitive to the depressive effects of opioids.

Paediatric overdose of a parent’s methadone (long-acting opioid) syrup requires overnight admission for extended observation and monitoring. Anti-diarrhoeal preparations containing diphenoxylate (with atropine) produce delayed onset of symptoms, with numerous paediatric deaths reported. A major metabolite of diphenoxylate is more potent than the parent compound and undergoes enterohepatic circulation. Dextropropoxyphene has a membrane-stabilising effect on cardiac conducting tissue and may induce ventricular arrhythmias and heart block.

The classic features are nausea and vomiting, drowsiness, pinpoint pupils, respiratory depression, and occasionally bradycardia and hypotension. Respiratory depression may lead to hypoxia and respiratory arrest. The histamine-releasing effects of some opioids may cause urticaria and hypotension.

Management of the opioid toxidrome is essentially supportive, with attention to airway and ventilation. All children with altered levels of consciousness should have blood glucose levels checked. Activated charcoal may be considered for massive ingestions or long-acting preparations. Insertion of gastric tubes for charcoal administration is not recommended unless the child is

intubated for other indications.

Naloxone, the antidote for opiate toxicity, is a competitive antagonist at opioid receptors. Naloxone (IV 0.01 mg kg^{-1}, maximum 400 μg, as a bolus, repeated every few minutes till appropriate response) may be useful to reverse the neurological and respiratory depression, and should be titrated to respiratory rate and effort. Naloxone's short therapeutic half-life of 30–60 minutes may necessitate a continuous infusion, in order to maintain the reversal and obviate the need for mechanical ventilation, particularly for ingestions of long-acting or sustained-release opioids.

All patients should be observed for a minimum of 6 hours. Methadone, dextropropoxyphene and sustained-release opiates may cause symptoms persisting for 24–48 hours, so prolonged observation is necessary after ingestion of these agents.

Anticholinergics and antihistamines

Anticholinergic (antimuscarinic) poisoning can result from a diverse range of therapeutic substances, plants and natural remedies, many of which can be bought over-the-counter. Pharmaceuticals with prominent anticholinergic properties include first-generation antihistamines, antipsychotics and tricyclic antidepressants (TCAs). Some plants, e.g. Jimsonweed, angel's trumpet and mushrooms, contain alkaloids with potent anticholinergic effects.

The anticholinergic toxidrome is caused by competitive inhibition of the muscarinic receptor in the brain and autonomic nervous system. The classic anticholinergic toxidrome is well described by the following collection of sayings:

- Hot as a hare – hyperthermia from inability to sweat.
- Blind as a bat – dilated pupils with loss of accommodation.
- Dry as a bone – hot, dry skin and mucous membranes with paucity of secretions.
- Red as a beet – erythematous skin.
- Mad as a hatter – central anticholinergic delirium manifest by agitation, confusion, hallucinations (often visual), dystonic movements and seizures.

Other anticholinergic effects include tachycardia, GI ileus and urinary

retention.

The management of anticholinergic poisoning includes attention to the ABCs with appropriate supportive therapy and monitoring of vital signs and mental state. Symptomatic patients should have IV access, a 12-lead electrocardiogram (ECG) and continuous cardiac monitoring. Benzodiazepines are useful in managing agitation or seizures. Physostigmine is a cholinesterase inhibitor which enters the CNS and is effective at reversing central anticholinergic delirium. Due to concerns regarding adverse cardiac side effects, the use of physostigmine should be discussed with a toxicologist. The initial dose in children is 0.02 mg kg^{-1} (maximum 0.5 mg) by slow IV push; doses may be repeated every 15 minutes.

Aggressive cooling measures may be required in severe hyperthermia. Urinary retention is common and bladder ultrasound scanning is useful; urinary catheterisation is required for patients with confirmed retention. Patients with moderate or severe toxicity should be admitted to an intensive care facility.

Antihistamine poisoning in children is of concern usually for first-generation agents (e.g. promethazine), which have significant anticholinergic effects. Management is supportive, and the anticholinergic effects of antihistamines, while unpleasant, are generally not life threatening. Asymptomatic patients may be discharged after 6 hours.

Hypotension should be treated with IV fluids. Convulsions and anticholinergic delirium are best managed with benzodiazepines. Continuous ECG monitoring is advised for symptomatic children and those with persistent tachycardia. Ventricular arrhythmias should be managed with sodium bicarbonate as the drug of choice for QRS prolongation with sodium-channel-blocking antihistamines.

Corrosive ingestions

Most serious caustic ingestions involve strong acids and alkalis which account for a high number of presentations to the ED. The initial presentation and treatment are similar to other burns. Domestic bleaches and ammonia products generally cause minor injuries. Serious injuries result most often from the ingestion of drain and oven cleaners (NaOH, KOH). Dishwashing powder residue left in the dispenser of machines is a commonly accessed alkali that may cause serious injury.

The severity of burn depends on the nature, volume, pH and concentration of the agent and the duration of contact. Stomach contents may afford some

protection from injury, but pylorospasm, oesophageal reflux and vomiting may exacerbate injury. Liquids may cause a circumferential injury and powders/granules or tablets may cause prolonged contact with a mucosal surface, with potential for linear burns, deep erosion and perforation. Acids cause superficial corrosion and a coagulative necrosis, and the extent of tissue penetration is limited by eschar formation. Alkalis start to burn immediately on contact and cause a liquefactive necrosis of fat and protein, penetrating deeply into tissues. Acids typically injure the stomach while alkalis damage the oropharynx and oesophagus.

Many children will be asymptomatic, especially if low-concentration household products are involved. Pain, drooling, dysphagia, vomiting, abdominal pain and haematemesis may occur. Airway compromise with laryngeal oedema, cough and bronchospasm may be seen after ingestion of high concentration agents. CT scan or endoscopy is required to provide a guide to prognosis and management. The extent of injury is graded by the depth of ulceration and the presence of necrosis. Typically, after ingestion the mouth or oesophagus is red and ulceration follows within 24 hours. One-third of patients with oral burns have associated oesophageal lesions, whereas 10–15% of patients with oesophageal lesions have no oropharyngeal burns. Drooling and dysphagia persisting beyond 12–24 hours are reliable predictors for oesophageal injury and suggest the need for CT scan and/or endoscopy.

Oesophageal perforation and mediastinitis may be suspected by chest pain, fever, pleural rub and dyspnoea. Abdominal pain, fever, peritonism and ileus may indicate gastric or oesophageal perforation. These signs may progress to septic shock, multiorgan failure and death. Large acid ingestions may be associated with hypotension, metabolic acidosis, haemolysis, nephrotoxicity and pulmonary oedema.

Late complications are infection, achlorhydria and stricture formation in 1–3%. All patients with full-thickness and 70% with deep ulceration will develop strictures. Eighty percent of all strictures occur within 2 months of ingestion and 99% within 1 year.

The management of caustic ingestions is aimed at limiting the extent of injury and preventing strictures and other complications. Immediate management consists of rinsing the skin and mouth with water. Children with corrosive ingestion should be kept nil by mouth until assessment and consultation with a toxicologist. Acids and alkalis do not bind to charcoal. Attempts to neutralise the substance are contraindicated, but dilution with water may possibly be helpful

for acids and may reduce mucosal contact time in ingestion with particulate alkalis. Early treatment focuses on ensuring an adequate airway, IV fluid replacement, monitoring fluid balance, avoiding vomiting and adequate analgesia. Patients with deep, especially circumferential burns of the oesophagus should be admitted to an intensive care unit and may require prolonged parenteral feeding and repeated endoscopic stricture dilatations. Early surgical intervention and prophylactic antibiotics are required if perforation or penetration is suspected clinically or on endoscopy or imaging studies. Steroids have no proven benefit and may possibly increase the risk of perforation.

Patients should be advised to return if they develop respiratory difficulty, pain or dysphagia. All symptomatic children should be admitted for observation and potential CT scanning and/or endoscopy.

Ethanol

Ethanol is available in numerous household medicinals, mouthwashes and perfumery products as well as alcoholic drinks. All products marketed in Australia as ‘methylated spirits’ contain ethanol. Although frequently ingested by children, serious toxicity is uncommon.

Ethanol is well absorbed across GI mucosa and respiratory tract, mostly within 30–60 minutes, and distributes to total body water. Children metabolise alcohol faster than adults. Only very small amounts are excreted unchanged in the urine and the breath. Hypoglycaemia is caused by depressed gluconeogenesis. The potentially fatal dose of alcohol for children is 4 mL kg^{-1} of absolute alcohol (e.g. 10 mL kg^{-1} for a 40% alcohol spirit), about half the dose required for adults. Quite low serum levels (>10 mmol L^{-1}, >0.05% or >500 mg L^{-1}) may produce clinically significant effects in children.

Ethanol acts on the reticular activating system to cause CNS depression. Low concentrations result in alterations of mood and thought processes, whereas higher concentrations affect cerebellar function, causing ataxia and slurred speech. Higher levels still depress all cortical function and brainstem activity, depressing respiratory drive and protective airway reflexes. Respiratory arrest or aspiration is a frequent cause of death. Facial flushing, excessive sweating and vomiting are common.

Management depends on the time elapsed since ingestion. Assess and secure the ABCs and correct electrolyte abnormalities and dehydration. Patients with severe CNS depression are at risk of aspiration and require airway protection.

Blood glucose should be monitored. A blood alcohol level may be taken at least 1 hour post ingestion if symptoms are present, although management is generally determined by the clinical state. Hypoglycaemia should be corrected with 5 mL kg^{-1} 10% glucose. Hypotension will usually respond to IV fluids, and acidosis usually responds to correction of hypovolaemia and hypoglycaemia. Coma and prolonged lie may lead to hypothermia and rhabdomyolysis in severe cases. Haemodialysis, though rare, may be indicated in the extremely intoxicated child who potentially may take many hours to days to eliminate ethanol. Admit all children who are clinically intoxicated until asymptomatic.

Rare and dangerous poisons

Although most substances are harmless to children in small amounts, a few pharmaceuticals and chemicals are extremely toxic in small quantities. Table 21.2.1 lists drugs where ‘one pill can kill’ and chemicals where a sip or mouthful is potentially lethal.

Salicylates

The incidence of acute salicylate poisoning has declined due to improvements in medication packaging, removal of aspirin from oral paediatric formulations and paracetamol now being the favoured over-the-counter analgesic. Methylsalicylate and salicylic acid are common in many topical preparations. Oil of wintergreen containing methylsalicylate can be significantly toxic when ingested. Choline salicylate is a constituent of many teething gels.

Table 21.2.1

Potentially lethal ingestions in small amounts for a toddler

Pharmaceuticals	Chemicals
Calcium-channel blockers (diltiazem and verapamil)	Organophosphate and carbamate pesticides
Chloroquine and hydroxychloroquine	Paraquat
Clonidine	Camphor
Tricyclic antidepressants	Naphthalene
Sulfonylureas	Toxic alcohols
Amphetamines and ecstasy	Essential oils

Aspirin is rapidly absorbed from the upper GI tract, with peak plasma concentrations at 1–3 hours after therapeutic doses. An ingestion of 150–300 mg kg^{-1} may result in mild to moderate toxicity. Ingestions over 500 mg kg^{-1} can potentially cause severe toxicity.

Aspirin is hydrolysed to form salicylic acid (salicylate). In large overdoses, the potential for pharmacobezoar formation in the gut may alter absorption kinetics. In therapeutic doses, salicylate is 85–95% plasma bound, but in overdose, free salicylate concentration rises as plasma protein binding is saturated. As metabolic pathways in the liver become saturated, the kidney becomes the main route of elimination. Clearance is markedly enhanced by alkaline urine.

Salicylate poisoning leads to the uncoupling of oxidative phosphorylation and anaerobic metabolism. The resulting lactic acidosis is more prominent in young children. Early signs of salicylism include respiratory alkalosis from centrally driven hyperventilation and tinnitus. With increasing toxicity, confusion, hallucinations and seizures are reported. Metabolic derangements include temperature dysregulation, impaired glucose metabolism and transport as well as electrolyte abnormalities. Mixed picture acid–base derangement is a hallmark of severe salicylate poisoning, with serum pH below 7.2 being a late and ominous sign. Coagulopathy may result from competitive inhibition of synthesis of vitamin-K dependent factors. Acute respiratory distress syndrome (ARDS) has been reported, but the mechanism is unclear.

Investigations include baseline blood glucose level, electrolyte and renal function, acid–base status, coagulation panel and 6-hour salicylate level. Decontamination with activated charcoal is indicated in large ingestions presenting early and may be considered in late presentations of enteric-coated preparations. Meticulous monitoring of fluid balance, temperature, glucose and electrolyte levels is recommended. In particular, potassium replacement is often required, along with maintenance of urine output.

Methods of enhancing elimination should be instituted following consultation with a toxicologist. Urinary alkalinisation with IV sodium bicarbonate infusion is known to enhance renal excretion of salicylate ions. The target urinary pH is at least 7.5 with maintenance bicarbonate doses ranging from 0.25 to 0.5 mmol kg^{-1} h^{-1}. Haemodialysis is indicated in severe toxicity with acidaemia, cardiorespiratory failure, renal impairment or CNS manifestations (coma or seizures). Complications of salicylate poisoning require aggressive supportive management. The development of acute pulmonary oedema often signals the

need for invasive ventilation and haemodialysis. Seizures warrant benzodiazepine therapy and correction of any glucose or electrolyte derangement.

Observe all symptomatic children and those with ingestions of greater than 150 mg kg^{-1}. Most patients will require admission for 6–12 hours for observation of clinical state and serum salicylate level.

β-Blockers

β-Blockers have wide clinical use in the treatment of cardiac conditions, hypertension, thyrotoxicosis and prophylaxis for migraine.

β-Blockers are class II antiarrhythmics, which act by competing with catecholamines at β-adrenergic receptor sites. Different β-blockers have differing cardioselectivity, membrane-stabilising activity, partial agonist activity and lipid solubility. They are well absorbed from small intestine, with peak serum levels within 1–4 hours. The elimination half-life is less than 12 hours. They have a moderate to large volume of distribution. Highly lipid soluble drugs, such as propranolol, cross the blood–brain barrier and thus have more potent CNS effects. Propranolol is also known for its cardiac sodium-channel-blocking properties, which cause prolongation of the QRS complex.

The major clinical effects are cardiovascular with bradycardia of varying types (sinus, junctional or ventricular escape), which may progress to cardiac arrest. Hypotension results from bradycardia, myocardial depression and vasodilatation. Deterioration can be sudden and precipitous, particularly with propranolol, which can cause seizures, coma and wide complex arrhythmias. Hypoglycaemia may occur due to impaired gluconeogenesis and glycogenolysis. Bronchospasm is more likely in atopic subjects, and more prominent with non-selective agents.

Good supportive management is essential. IV access should be secured and blood glucose and electrolytes measured. ECG and BP should be continuously monitored. Cardiovascular effects should be treated with atropine, volume expansion and adrenaline. High-dose insulin enhances heart rate and myocardial contractility independently of β-receptor activation and is currently the preferred inotrope over glucagon. Doses are similar to those below in calcium-channel-blocker poisoning. Extreme measures such as transvenous pacing and cardiopulmonary bypass may be required in cases of intractable hypotension. Importantly, prolonged resuscitation and aggressive supportive care may allow

the peak toxicity to pass and improve survival. Hypoglycaemia should be treated in the usual manner with 2.5–5 mL kg^{-1} 10% glucose. In propranolol overdose, sodium bicarbonate is the antidote of choice for wide complex arrhythmias and seizures. Symptomatic children should be monitored in an intensive care setting.

Calcium-channel blockers

Calcium-channel blockers (CCBs) are widely used in the treatment of hypertension, coronary artery disease and supraventricular tachyarrhythmias.

CCBs are rapidly absorbed from the GI tract and have peak plasma concentrations ranging from 30 minutes (nifedipine) to 90 minutes (verapamil), but sustained-release preparations are associated with longer times to peak concentration and prolonged clinical effect. These agents inhibit the entry of calcium into the cells of cardiac and smooth muscle, decreasing the activity of the calcium-dependent actin-myosin ATPase. Dihydropyridines (represented by the prototype agent nifedipine) are more potent at peripheral vascular calcium channels and have little cardiac toxicity. Verapamil and diltiazem, also available in sustained-release preparations, are highly toxic drugs where a single large dose tablet can cause profound cardiogenic shock in a toddler. Dihydropyridine CCBs are unlikely to cause hypotension or cardiac conduction abnormalities in small doses. Dihydropyridines in large doses or in combination products, such as with angiotensin-converting enzyme (ACE) inhibitors, may cause profound vasoplegic shock.

The features of cardiovascular toxicity include hypotension, bradyarrhythmias, such as sinus bradycardia, varying degrees of AV block, and asystole. Dose-dependent peripheral vasodilatation with hypotension may occur. Other manifestations include nausea and vomiting, lethargy, coma, seizures, hyperglycaemia and lactic acidosis.

Good supportive management is essential. IV access should be secured, and blood glucose and electrolytes measured. ECG and BP should be continuously monitored. Activated charcoal is worth considering in patients who present early with a significant ingestion. Whole bowel irrigation with polyethylene glycol should be considered for large ingestions of sustained-release preparations. Echocardiography may assist in determining the cause of shock, either central from myocardial depression or peripheral from vasoplegia.

Calcium is the initial antidote for hypotension and bradycardia (bolus: 10% calcium chloride 0.2 mL kg^{-1} or 10% calcium gluconate 0.6 mL kg^{-1}). Atropine

is likely to be ineffective. Catecholamine infusions (e.g. adrenaline [epinephrine] commencing at 1 mcg kg^{-1} min^{-1}) may be required. Other inotropes that do not require calcium influx are potentially useful in managing intractable shock from CCBs. These include high-dose insulin (1–2 units kg^{-1} IV bolus; 1–2 units kg^{-1} h^{-1} infusion). Rarely, more extraordinary measures may be necessary such as transvenous pacing, cardiopulmonary bypass or aortic balloon pumps. Prolonged resuscitation and aggressive supportive care may allow the peak toxicity to pass and improve survival.

Symptomatic children should be monitored in an intensive care setting. Observation for 24 hours is warranted for ingestion of sustained-release formulations.

Digoxin

Digoxin is a cardiac glycoside used for management of heart failure and supraventricular arrhythmias. Some plants contain digitalis glycosides (e.g. foxglove, oleander), and poisoning from these plants should be managed in a similar manner to digitoxicity. Acute digoxin poisoning in children is more often seen in the context of toddlers who obtain access to grandparents' medication. Rarely, children with underlying cardiac disease on maintenance digoxin therapy can develop chronic toxicity, and therapeutic drug monitoring is crucial. Chronic overdosing or renal impairment can lead to chronic digitoxicity.

Digoxin is well absorbed from the GI tract and has a relatively large volume of distribution. Digoxin is predominantly excreted unchanged by the kidney, with an elimination half-life of about 36 hours. Digoxin inhibits the action of the cardiac Na/K ATPase pump and accumulation of sodium and calcium ions leads to intracellular depletion of potassium and hyperkalaemia. The slowing of conduction, as well as increased refractory period, through the AV node, enhanced automaticity of the Purkinje fibres and enhanced vagal tone have the potential of causing a multitude of arrhythmias including sinus bradycardia, sinoatrial arrest, conduction blocks, ventricular tachycardia and fibrillation.

Early signs of chronic toxicity include nausea, vomiting and diarrhoea. The child is often asymptomatic in acute poisoning, until haemodynamic instability from cardiac toxicity becomes clinically apparent. Patients can deteriorate suddenly and digitoxicity can produce both brady- and tachyarrhythmias. Assess ABCs, secure IV access and continuously monitor blood pressure (BP) and ECG. Although digoxin is well bound by activated charcoal, repeated vomiting

may preclude its use. Obtain a serum digoxin concentration and electrolytes. Serum potassium concentration should be monitored every 4 hours. Hyperkalaemia should be corrected to within upper limits of normal with sodium bicarbonate and insulin/glucose. Calcium is best avoided in digitoxicity due to the potential for myocardial tetany.

Digoxin Fab antibodies are a specific and effective antidote in digoxin poisoning. IV Fab fragments of digoxin-specific antibodies are first-line therapy for patients with cardiac arrhythmias with haemodynamic instability. The dosage should be based on total body load, estimated from the serum digoxin concentration or from the ingested dose. Alternatively, a dose estimation can be made on the presumption that one vial of 40 mg will bind ~0.6 mg of digoxin. A clinical response is seen in 20–30 minutes, with maximum effect at 2–4 hours. An empiric dose of 5 vials of digoxin Fab may be given IV over 20 minutes in severe life-threatening toxicity or digoxin-induced cardiac arrest. It is important to note that subsequent digoxin levels post treatment with antibodies are not interpretable and should not be performed.

Acute overdoses should be observed for a minimum of 12 hours or overnight. Symptomatic or haemodynamically unstable patients should be monitored in an intensive care unit or coronary care facility. Patients treated with Fab fragments should be monitored for subsequent hypokalaemia and for deterioration of any pre-existing cardiac disease.

Clonidine

Until recently clonidine was used primarily as an anti-hypertensive agent and accessibility to children was limited. The drug is now widely prescribed in the treatment of attention deficit hyperactivity disorder, conduct disorders, Tourette's syndrome and for narcotic and alcohol withdrawal symptoms. Clonidine overdose is commonly seen in children with behavioural disorders and their siblings who have access to clonidine.

Clonidine is a central α_2-adrenergic autoreceptor agonist that acts on brainstem receptors, causing inhibition of sympathetic outflow. Its stimulation of peripheral α_2-receptors on vascular smooth muscle may cause transient hypertension, but hypotension usually occurs subsequently. Clonidine also has opiate-like effects which may be mediated through μ receptors. It is rapidly absorbed and distributed with peak plasma concentrations 60–80 minutes post ingestion. The elimination half-life is 6–24 hours.

Clinical effects are seen 30–60 minutes after ingestion. Depression of the CNS with lethargy and impaired conscious state is the most frequent manifestation. Miosis and hypothermia may be observed. Symptoms are minimal with ingestions of under 10 mcg kg^{-1}, but cardiovascular compromise with hypotension and bradycardia may occur after ingestion of 10–20 mcg kg^{-1}. Respiratory depression and apnoea may be seen after ingestions of 20 mcg kg^{-1}. There have been no reports of in-hospital paediatric deaths.

Treatment is largely supportive and decontamination usually not beneficial. Hypotension should be treated with volume expansion and vasopressors are rarely required. Hypertension is usually transient and treatment unnecessary. Atropine may be useful in the treatment of bradycardia. Naloxone is worth considering in clonidine poisoning when respiratory depression is present – 5–10 mcg kg may be given titrating to respiratory rate and effort. There may be inconsistent reversal of the neurological and respiratory effects after administration of naloxone.

Maximal toxicity is expected in the first 6 hours, and children who show no symptoms at that stage can be discharged. Children with significant respiratory and cardiovascular compromise may require admission to an intensive care unit for up to 24 hours.

Iron

Iron tablets are commonly available in the homes of toddlers, but severe poisoning is uncommon.

The amount of elemental iron varies according to the formulation. Initial toxicity is due to the corrosive effects on the GI tract. Iron is absorbed in the ferrous state and after oxidation to the ferric state becomes bound to ferritin. Toxicity occurs when ferritin and transferrin are saturated and serum iron exceeds the total iron-binding capacity (TIBC). High concentrations of intracellular iron cause mitochondrial dysfunction, interfering with mitochondrial processes, causing lactic acidosis and cell death.

Ingestions of less than 20 mg kg^{-1} elemental iron usually remain asymptomatic. Significant symptoms usually only occur in ingestions above 60 mg kg^{-1}. Potentially lethal systemic toxicity may follow ingestions of greater than 100 mg kg^{-1} elemental iron. Serum iron peaks at 4–6 hours. Serum iron levels should be considered in conjunction with the clinical state.

Although four stages of iron poisoning are classically described, distinct

phases may not be apparent with severe poisoning. In the initial 6 hours the gastric irritant effects predominate, with vomiting, diarrhoea and haematemesis or melaena. Circulating free iron may damage blood vessels and cause a transudate of fluid and hypotension. There may be a quiescent phase when the patient may appear to be improving, but about 12–24 hours after ingestion the physiological processes of cells are disrupted, leading to metabolic acidosis, GI haemorrhage, altered mental state, pulmonary oedema, and cardiovascular, hepatic and renal failure. The liver is particularly vulnerable and fulminant hepatic failure may cause hypoglycaemia, coagulopathy and death. At 4–6 weeks there may be stricture formation in the GI tract due to scarring.

Symptomatic patients and those with ingestions greater than 60 mg kg^{-1} of elemental iron require laboratory investigations and an abdominal X-ray. Iron does not bind well to activated charcoal. Patients with ingestions potentially in excess of 60 mg kg^{-1} of elemental iron should have IV access and laboratory investigations including blood gas, glucose level, electrolytes, renal and liver function tests and a 4–6 hour iron level. IV fluid resuscitation may be required, and electrolytes and glucose should be monitored. Whole bowel irrigation with polyethylene glycol at 20 mL kg^{-1} h^{-1} via a nasogastric tube is reserved for massive ingestions.

Desferrioxamine (deferoxamine) binds free iron in the intravascular and extracellular space and the chelated complex is eliminated in the urine, imparting a pink–brown colour (vin-rose urine). The decision to use chelation therapy should be based on the combination of the patient's clinical condition and serum iron concentration. IV desferrioxamine (15 mg kg^{-1} h^{-1} to a maximum of 80 mg kg^{-1}/24 h) is indicated in patients with hypotension, shock, coma, convulsions or potentially if the serum iron concentration persists above 90 μmol L^{-1}. Desferrioxamine infusion is usually required for 6–12 hours. The end-points of chelation therapy are clinical improvement and a reduction in free iron levels. Acid–base and electrolyte balance should be maintained and hepatic and renal function monitored. The chelated complex can be removed with haemodialysis should renal function be significantly impaired.

Asymptomatic children with ingestions of under 60 mg kg^{-1} may be observed at home. Symptomatic patients, or those with ingestions greater than 60 mg kg^{-1} of elemental iron, require further evaluation in hospital and an admission of 12–24 hours.

Warfarin and rodenticides

Domestic rodenticides are widely available in almost every household. The majority of products contain superwarfarins, long-acting anticoagulants, as their base (e.g. brodifacoum). A limited number of rodenticides have a combination of short-acting warfarin and long-acting superwarfarin – these pose a management challenge.

Short-acting warfarin ingestion in children is of concern at doses greater than 0.5 mg kg^{-1}. Prolongation of the prothrombin time usually occurs at 12–36 hours. Treatment with vitamin K_1 (phytomenadione) is dependent on the dose ingested, prothrombin time and signs of bleeding.

Acute ingestion of a few pellets of superwarfarin (rodenticide) is usually not a problem. Coagulopathy is likely to occur in massive ingestion or repeated or chronic ingestions of rodenticides. In these cases, prothrombin time should be measured and, if prolonged, treatment with vitamin K instituted; follow-up with serial coagulation tests is usually necessary. Coagulopathy after superwarfarin ingestion may not manifest until 48 hours post exposure. A prolonged course of vitamin K may be required for large ingestions of long-acting superwarfarins.

Oral hypoglycaemics

Sulfonylurea poisoning is uncommon in children, but even ingestions of a single tablet have led to significant toxicity and mortality, and the onset of symptoms may be delayed and prolonged. Children may access the tablets in the home of diabetic relatives. In Australasia, gliclazide, glipizide and glibenclamide are responsible for the majority of poisonings. Ingestion of the newer sustained release preparations of gliclazide warrant prolonged monitoring of blood glucose levels.

Sulfonylureas induce hypoglycaemia by stimulating endogenous insulin secretion. In contrast, biguanides rarely cause hypoglycaemia but can induce severe lactic acidosis. Children are more susceptible to hypoglycaemia than adults because of their increased metabolic rate and limited ability for gluconeogenesis.

Hypoglycaemic manifestations occur with palpitations, shaking, hunger, sweating, weakness and with increasing neuroglycopenia, confusion, coma and seizures occur.

Following assessment and management of the ABCs, a blood glucose level

should be performed immediately and checked hourly. Gastric decontamination with activated charcoal is not routinely warranted. Hypoglycaemia should be treated with glucose 10% 5 mL kg^{-1} IV bolus. Glucagon is not recommended for sulfonylurea-induced hypoglycaemia.

Octreotide, a long-acting synthetic somatostatin analogue, inhibits secretion of insulin from the pancreas and may be the most appropriate method of stabilising blood glucose levels. Patients with persistent or recurrent hypoglycaemia, requiring repeat boluses of glucose, should be given octreotide (1 mcg kg^{-1} IV bolus), followed by an octreotide infusion (250 mcg in 250 mL 5% glucose at 1 mcg kg^{-1} h^{-1}, to maximum of 25 mcg h^{-1}). If octreotide is not available, a glucose 10% infusion should be commenced at 1–2 mL kg^{-1} h^{-1}. Hourly blood glucose measurements are required until octreotide and/or glucose infusions are ceased.

Asymptomatic children should be observed with glucose monitoring for at least 8 hours; this may be up to 16 hours for sustained release preparations. Symptomatic children, and those with confirmed hypoglycaemia, require intensive monitoring of blood glucose and clinical condition.

Tricyclic antidepressants

Despite the declining prescription of TCAs, the low therapeutic index and potential for lethal toxicity remain a concerning cause of paediatric morbidity and mortality.

TCAs are rapidly absorbed. TCAs have a high degree of protein binding and a large volume of distribution. Although different TCAs have different pharmacokinetic parameters, the effects in overdose are similar. Dothiepin is associated with the greatest lethality. Minor TCA toxicity is generally manifest by central and peripheral anticholinergic signs and vasoplegia from α-adrenergic blockade. More serious toxicity results from fast sodium-channel blockade in the myocardium, causing a wide complex QRS and ventricular dysrhythmias. Drowsiness from histamine blockade and seizures from GABA antagonism are hallmarks of serious TCA toxicity.

Tricyclic ingestions of <5 mg kg^{-1} result in minimal toxicity and no treatment is required. Ingestions of 5–10 mg kg^{-1} may cause drowsiness, ataxia and mild anticholinergic symptoms of dilated pupils, ileus and urinary retention but life-threatening toxicity is unlikely. Ingestions over 10 mg kg^{-1} may cause life-threatening coma, seizures and cardiac dysrhythmias. There may be acidosis, hypokalaemia and inappropriate antidiuretic hormone secretion. Onset of

symptoms is usually within 3 hours and persists for less than 12–24 hours.

The management of TCA poisoning includes attention to the ABCs, good supportive therapy and GI decontamination for potentially serious ingestions. Continuous cardiac monitoring, serial 12-lead ECGs and close observation of vital signs and mental state are required for all ingestions of >5 mg kg^{-1}. Airway protection should precede administration of charcoal if the patient is less than fully conscious.

Depressed conscious state is the best predictor of serious toxic complications (seizures, ventricular arrhythmias, hypotension and the need for mechanical ventilation) and the ECG limb lead QRS duration of 100 ms or greater is associated with an increased incidence of seizures and cardiotoxicity. Early intubation and hyperventilation to an alkaline serum pH of 7.45–7.55 may promote redistribution of TCA and reduce its binding to sodium channels. Intubate and mechanically ventilate all patients with rapidly decreasing conscious state, seizures and ventricular dysrhythmias. Hypotension should be treated with volume replacement and sodium bicarbonate; intractable hypotension may require vasopressors such as noradrenaline (norepinephrine).

Sodium bicarbonate is regarded as a specific antidote in the treatment of the cardiovascular effects of TCA toxicity. It competitively overcomes sodium-channel blockade, and its effect on serum pH appears to improve sodium-channel function. Ventricular dysrhythmias will usually respond to treatment with sodium bicarbonate (1–2 mEq kg^{-1} IV bolus, repeated till the QRS narrows or serum pH reaches 7.45–7.55). Refractory ventricular arrhythmias should be treated according to standard ACLS protocols avoiding type 1a and 1c antiarrhythmics. Lidocaine (lignocaine) appears to be safe.

Seizures may be averted or attenuated by bicarbonate therapy. Benzodiazepines are the preferred agents to treat seizures, but barbiturates may be required to treat refractory seizures. It is best to avoid phenytoin, a sodium channel blocker, which may aggravate cardiac conduction abnormalities and may have little effect in controlling seizure activity. TCAs are not amenable to removal by extracorporeal methods due to their large volume of distribution. Quantitative analysis of TCA levels does not aid acute management, but screening for other drugs should be considered in deliberate self-harm.

Patients who ingest >5 mg kg^{-1} TCA should be admitted for observation for at least 6 hours but may be discharged at that time if the ECG remains normal and the child is well. TCA ingestions with significant CNS depression, seizures or significant cardiotoxicity should be admitted to an intensive care facility.

Toxic alcohols

Ethylene glycol is encountered in antifreeze compounds and radiator additives. Methanol is found in model aeroplane fuel and in home brewing concoctions. Toxic alcohols are rapidly absorbed from the GI tract and distribute to total body water.

Methanol is oxidised to formaldehyde by the rate-limiting enzyme alcohol dehydrogenase, and then aldehyde dehydrogenase converts the formaldehyde to formic acid (formate). Approximately 2% of methanol is excreted unchanged by the kidneys, and a small amount is excreted via the lungs. The optic nerve is particularly susceptible to the toxic effects of formic acid. Lactate is produced from anaerobic glycolysis as a result of tissue hypoxia and a formate-induced inhibition of mitochondrial respiration. Ingestion of 1.5 mL of 100% methanol in a child weighing 10 kg would produce a potential peak plasma level of 6 mmol L^{-1} (0.02%, 20 mg dL^{-1}), so a single mouthful is potentially lethal. Symptoms of methanol poisoning may be delayed with a 12–24 hour latent period because of the slow metabolism to formate. The most common presentation of intoxication consists of a triad of findings related to the GI symptoms, visual disturbance and metabolic acidosis. Nausea and vomiting, epigastric abdominal pain, pancreatitis and GI bleeding may occur. Visual disturbances including blurred vision, central scotoma, yellow spots and complete blindness offer an important diagnostic clue.

Ethylene glycol depresses the CNS, but the hepatic metabolites glycoaldehyde, glycolic acid, glyoxylate and oxalate are responsible for toxicity. Formation of glycolic acid and some lactic acid is the primary cause of the delayed metabolic acidosis, which can occur 4–12 hours after ingestion. Oxalate is highly toxic, causing myocardial depression and acute renal tubular acidosis, which may progress to renal impairment. Calcium oxalate crystals may be noted on examination of the urine. The initial symptoms of an acute ethylene glycol poisoning include those of alcohol intoxication with lethargy, slurred speech, nystagmus, ataxia and vomiting. Papilloedema occurs less frequently than with methanol poisoning. An elevated anion osmol gap acidosis may occur. Seizures, myoclonic jerks and tetanic contractions reflect hypocalcaemia.

Fomepizole and ethanol preferentially bind alcohol dehydrogenase, which is involved in the metabolism of methanol and ethylene glycol to their toxic metabolites. These antidotes do not prevent the toxic effects of the acid metabolites and are only useful if an osmolar gap exists. Fomepizole, which is

difficult to source in Australasia, is expensive but easier to administer and monitor than IV ethanol, without the complications of profound hypoglycaemia, hepatotoxicity and inebriation that may occur with ethanol infusions.

The target serum ethanol concentration of 20 mmol L^{-1} (100 mg dL^{-1}, 0.1%) will fully inhibit alcohol dehydrogenase. This can be difficult to achieve in children without advanced support of airway and ventilation. Ethanol is preferably administered orally or via gastric tube. The loading dose is 7.5 mL kg^{-1} of 10% (v/v) ethanol in 5% glucose water over 30 minutes, followed by a maintenance dose of 0.8–1.5 mL kg^{-1} h^{-1} of 10% ethanol, aiming for a concentration of 0.1 g/dL. Serum ethanol and glucose levels should be monitored after the loading dose and frequently thereafter. Haemodialysis enhances elimination and is indicated for renal failure, visual impairment or severe metabolic acidosis. Children with a history of exposure to toxic alcohols should have investigation of acid–base status, blood glucose, electrolytes, renal function and determination of osmolar gap. Admit all children who are clinically intoxicated until asymptomatic.

Psychostimulants

Amphetamines and cocaine are psychomotor stimulants that promote central and peripheral sympathetic outflow. Ecstasy, 3,4-methylenedioxymethamfetamine (MDMA), is an amphetamine derivative and common drug of abuse. In children it produces typical sympathomimetic effects, such as hyperactivity, agitation, tachycardia as well as serotonergic effects like hyperreflexia and bruxism. Ecstasy has additional psychoactive effects that alter perception and mood.

Complications of amphetamine and ecstasy ingestion include coma, convulsions, arrhythmias, malignant hyperthermia, rhabdomyolysis, hypertension and multiorgan failure. Cocaine also has sodium-channel-blocking properties, which can induce ventricular tachyarrhythmias. Hyponatraemia can be seen in ecstasy ingestion, leading to intractable seizures.

Blood pressure, temperature, and ECG monitoring should be instituted. Symptomatic patients and those with persistent tachycardia should be admitted to a monitored environment. Asymptomatic children may be discharged after 12 hours. Children with suspected neglect or non-accidental poisoning should be referred to child protection services.

Patients with signs of cardiac or central nervous system toxicity require admission to the paediatric intensive care unit. Careful monitoring of

haematological and biochemical parameters is essential. Hyperthermia may respond to fluid resuscitation and simple cooling measures; however, intractable cases should receive muscle paralysis and be ventilated in an intensive care setting. Convulsions and agitation should be treated with benzodiazepines; phenytoin and neuroleptics should be avoided. Ventricular tachyarrhythmias are managed with sodium bicarbonate.

Essential oils

Essential oils are complex aromatic mixtures of alcohols, esters, aldehydes, ketones and turpenes widely used in perfumery, food flavourings, massage and alternative remedies. Eucalyptus oil is an essential oil commonly implicated in hospitalisations for childhood poisoning in Australasia. Incidents usually involve vaporiser solutions, eucalyptus oil preparations and other medicinal preparations, which are freely available over the counter. Citronella oil is used as an insect repellent. Oil of turpentine has been largely replaced by white spirit and turpentine substitutes, which are less toxic.Essential oils are complex mixtures of substances distilled from plant species, including oil of cloves, eucalyptus, citronella, lavender, peppermint, melaleuca (tea tree) and turpentine. The oils probably differ in the degree of toxicity, but comparative data are lacking. The irritant effects are manifest by vomiting after ingestion and potential aspiration causing a chemical pneumonitis.

Essential oils are potentially very toxic. The breath, vomitus, urine and faeces smell strongly of the oil. Skin irritation may occur. Oil of turpentine has been reported to cause gastrointestinal irritation, central nervous system toxicity, hepatic and renal failure and metabolic acidosis. Eucalyptus oil toxicity has been reported to involve all major body systems, including death from ingestions of less than one mouthful. CNS depression, seizures and gastrointestinal effects generally occur within 1 hour after ingestion and respiratory complications including respiratory depression, bronchospasm, aspiration pneumonitis and pulmonary oedema have been reported.

As management is entirely supportive, assess and secure the ABCs. Oral activated charcoal and gastric lavage are not recommended. A chest X-ray is indicated only if respiratory symptoms are apparent. Aspiration pneumonia is treated with respiratory support if required. Benzodiazepines are preferred for managing seizures.

With regards to eucalyptus oil, asymptomatic children can be discharged after

4 hours' observation. Patients with impaired conscious state or respiratory distress on presentation should be admitted to an intensive care unit.

Organophosphates and carbamates

Pesticide poisoning in children is a rare event in Australasia. Organophosphates and carbamates inactivate the enzyme acetylcholinesterase at cholinergic nerve terminals and neuromuscular junctions, resulting in the cholinergic toxidrome. Plasma (butyl) cholinesterase and red blood cell cholinesterase levels are surrogate markers for exposure and toxicity.

The cholinergic toxidrome involves excess secretions from muscarinic overstimulation, neuromuscular dysfunction and paralysis from nicotinic overstimulation and central effects including delirium, seizures and eventually, coma. The onset, peak and duration of toxicity vary with each organophosphate compound.

In children, lethargy, coma and hypotonia are common early features of organophosphate toxicity. Excess secretions can be absent in children. Severe cases can progress rapidly to generalised weakness, coma, convulsions and respiratory failure. Organophosphates do not off-gas, unlike nerve agents, and do not cause secondary respiratory contamination of treating staff. They do, however, warrant the use of universal precautions including gown, gloves and goggles, for protection against secretions. Pesticides are often dissolved in hydrocarbon solvents and these chemicals give the characteristic odour, as well as causing symptoms in clinicians such as headaches and dyspnoea. Staff should be rotated regularly and the patient should be placed in a well-ventilated resuscitation area.

Resuscitation and decontamination should be carried out concurrently. Vomitus and secretions should be washed off the skin with soap and water. Contaminated clothing should be removed and disposed into biohazard bins. The main treatment for organophosphate and carbamate poisoning is anticholinergic therapy with atropine. Charcoal decontamination and gastric lavage have not proven to be effective. Atropine is indicated for the muscarinic symptoms of bradycardia and excess secretions. Atropine should be administered as a doubling regime starting at 0.02 mg/kg, repeated every 2–3 minutes until the end-points of normal heart rate, blood pressure and drying of secretions are reached. An atropine infusion may be necessary and should be discussed with a toxicologist. Tachycardia, fever and dilated pupils following atropine therapy

may indicate atropine toxicity. Oxime therapy in organophosphate poisoning is controversial and unproven. Pralidoxime, a cholinesterase reactivator at the neuromuscular junction, may be effective in re-establishing respiratory muscle and diaphragmatic function in some types of organophosphate poisoning. The loading dose is 25–50 mg kg^{-1} (maximum 2 g) infused IV over 30 minutes, followed by an infusion at 10–20 mg kg^{-1} h^{-1} for up to 48 hours.

All children with organophosphate or carbamate exposure should be monitored and observed for at least 12 hours or overnight. Children with the cholinergic toxidrome or neuromuscular effects require intensive care monitoring and admission.

House fires

From a toxicological point of view, the main exposures relate to carbon monoxide (CO) and cyanide, derived from combustion products of nitrogen-containing polymers, both natural (wool and silk) and synthetic (polyurethane and polyacrylonitrile), which are used extensively in domestic furnishings. In children, carbon monoxide poisoning is often associated with other injuries, such as burns or smoke inhalation. The affinity of haemoglobin for carbon monoxide is 210 times its affinity for oxygen. Carbon monoxide dissolved in the plasma acts as a direct cellular poison reacting with other haem proteins, such as mitochondrial cytochromes, to disrupt cellular metabolism.

Although carboxyhaemoglobin (COHb) levels poorly correlate with symptoms or prognosis, patients with up to 20% of haemoglobin affected complain of headaches and nausea. At 20–40%, patients tire and become confused. COHb greater than 40% can result in ataxia, collapse, and coma. Death is preceded by cardiac arrhythmias, cerebral oedema and severe metabolic acidosis. Standard oxygen saturation monitors are unreliable in the presence of COHb, with saturations of 100% occurring in the presence of significant hypoxia. Accurate oxyhaemoglobin saturation requires measurement with a co-oximeter. Conventional blood gas analysers can also be misleading.

Cyanide binds to ferric iron ($Fe^{3}+$) in the cytochrome a-a_3 complex, inhibiting its action and blocking the final step in oxidative phosphorylation. Aerobic metabolism is halted, and carbohydrate metabolism is diverted to the production of lactic acid.

The diagnosis of cyanide poisoning requires a high index of suspicion as clinical signs are limited and made even more difficult with co-existing CO

poisoning. Cardinal features are the presence of cyanosis with severe high anion gap metabolic acidosis and elevated lactate. Complications include coma, seizures and myocardial ischaemia. Treatment usually cannot wait until definitive diagnosis is made with cyanohaemoglobin levels.

Management of CO and cyanide poisoning involves high flow oxygen, supportive care and the potential use of cyanide antidotes. Unconscious patients require airway and ventilatory support and may warrant cerebral imaging in the event of trauma. Current evidence suggests that hydroxocobalamin is the most effective cyanide antidote with the fewest side effects.

Acknowledgements

The contributions of Nicholas Cheng and Helen Mead as authors in previous editions of the textbook are hereby acknowledged.

Further reading

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SECTION 22

Environmental

OUTLINE

22.1

Envenomation

Julian White

ESSENTIALS

1 Snakebite is the most important form of envenoming globally, causing significant morbidity and mortality.

2 Envenoming can cause rapid and severe medical problems in children, such as shock, collapse, convulsions, bleeding and respiratory failure due to either neurotoxic paralysis or neuroexcitatory pulmonary oedema.

3 Stabilisation of vital systems takes priority, followed by specific antidote therapy (usually antivenom) when indicated and appropriate fluid management.

4 Not all patients bitten/stung by venomous animals develop major envenoming, so antidote (antivenom) therapy is only appropriate where significant envenoming occurs.

5 In most cases antivenom, if required, should be given IV but always with adrenaline (epinephrine) and resuscitation facilities immediately to hand.

6 Assessment of degree of envenoming is critical in determining the need for antivenom.

7 Observe all patients for long enough to exclude late-developing envenoming; duration of observation varies dependent on the type of venomous animal.

Introduction

Envenoming is a significant global problem, particularly in the rural tropics, but more temperate and urban environments are not immune. Children represent 25% or less of all cases, but because of their lower body mass, are disproportionately represented in cases of severe envenoming. In general, early diagnosis and treatment are required to optimise outcomes, but diagnosis is not always easy and specific treatments are frequently unavailable. With estimates as high as several million cases of envenoming worldwide each year, it is not surprising that deaths may exceed 100,000 per year. For most major causes of envenoming, antivenom remains the definitive treatment, when available. Dosage is based on extent of envenoming, not patient size, so there is no paediatric dosage – children receive the same dose as adults.

Snakebite

Introduction

Snakebite is the single most important cause of envenoming. Some experts have estimated more than 2.5 million venomous snakebites per year, with more than 125,000 deaths. Accurate data to confirm such estimates are unavailable, but regional studies point to the broad veracity of such statements. In India alone a recent study calculated >45,000 snakebite deaths per year. It is not just the number of fatalities that are of concern in snakebite; many survivors are left with permanent physical impairment, sometimes severe. Paediatric cases represent 20–25% of the total, but a higher proportion of fatalities.

Snakes are ectothermic (‘cold-blooded’) reptiles, comprising around 3000 species. Venomous snakes are represented in six families: Colubridae (non-front-fanged colubrids [NFFC] snakes), Natricidae (NFFC snakes), Homalopsidae (NFFC snakes), Lamprophiidae (Atractaspinae; side-fanged or mole ‘vipers’), Elapidae (elapid/cobra-type front-fanged snakes) and Viperidae (viper and pit viper front-fanged snakes) (Table 22.1.1 and Figs 22.1.1–22.1.4). Fang structures and venom types vary between families, but the common theme is a bite resulting in injection or inoculation of venom through a break in the victim’s skin. In most cases, venom is injected by fangs, paired teeth evolved to deliver venom, usually through a venom groove or enclosed channel, exiting near the tip. The act of biting can leave a variety of bite marks, which may be highly visible or almost invisible. Venom need not be injected (‘dry bites’).

Venom varies between snake families, within families, between genera, within genera, between species, within species, between individual snakes, and even over time for a particular snake. It follows that while broad patterns of envenoming can be stated, there is always the possibility of an atypical pattern of effects occurring, because of venom variability. This is just one aspect of the potential difficulties in diagnosing and treating snakebite.

Table 22.1.1

Families of venomous snakes and their principal characteristics

Family	Fang type	Common names of selected medically important species	Geographical range of family
Colubridae (colubrids)	Non-front-fanged ('Back-fanged') colubrids (NFFC snakes) (see Fig. 22.1.1) OR No fangs (aglyphs)[a]	Boomslang (Africa) Vine snakes (Africa)	Global
Natricidae	Non-front-fanged ('Back-fanged') 'colubrids' (NFFC snakes)	Yamakagashi and red-necked keelbacks (Asia)	Australasia
Homalopsidae	Non-front-fanged ('Back-fanged') 'colubrids' (NFFC snakes)	Mud snakes and related water snakes	Australasia
Elapidae (elapids)	Front-fanged (proteroglyphs) (see Fig. 22.1.2) Fangs fixed or with minimal rotation	Cobras (Africa, Middle East, Asia) Coral snakes (Americas, Asia) Kraits (Asia) Mambas (Africa) Tiger snakes (Australia) Brown snakes (Australia and New Guinea) Taipans (Australia and New Guinea) Death adders (Australia, New Guinea, and eastern Indonesia) Mulga and black snakes (Australia and New Guinea) Small-eyed snake (New Guinea) Sea snakes (Pacific and Indian Oceans)	Global
Lamprophiidae, Atractaspinae (atractaspids)	Front-fanged (proteroglyphs) (see Fig. 22.1.3) Fang placed to exit mouth through side ('side-fanged')	Side-fanged and mole vipers	Africa and Middle East
Viperidae Subfamily Viperinae (viperids) Subfamily Crotalinae (crotalids)	Front-fanged (solenoglyphs) (see Fig. 22.1.4) Fangs on mobile maxilla, with considerable rotation possible	Common vipers and asps (Europe) Puff adders and Gaboon vipers Night adders (Africa) Carpet or saw-scaled vipers (Africa, Middle East, and western Asia) Russell's vipers (Asia) Rattlesnakes (Americas) Lance-headed vipers (Americas) Bushmasters (Americas) Green tree vipers (Asia) Mamushis and habus (Asia)	Global, except New Guinea and Australia

[a] The absence of fangs in most 'colubrids' does not exclude the possibility of at least local envenoming from toxic salivary secretions inoculated into the wound in the act of biting. Note the new terminology of NFFC snake covering a variety of species across three families, all previously within family Colubridae.

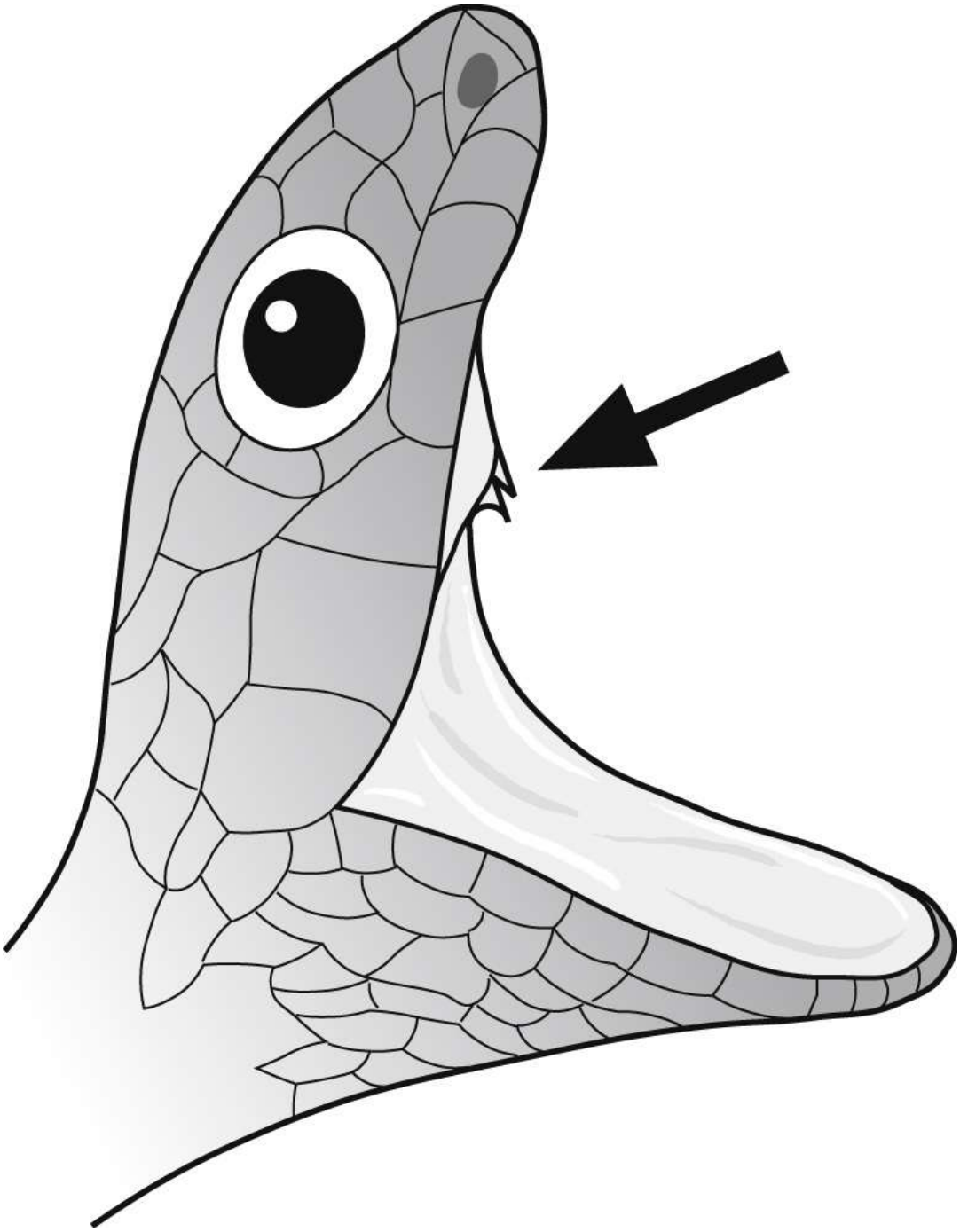

FIG. 22.1.1 Diagramatic representation of the head of a non-front-fanged colubrid snake (NFFC; previously referred to as an opisthoglyph or back-fanged snake). Reproduced with permission of Dr Julian White.

Venom has evolved from digestive juices. It has a variety of functions, which vary between species, but, in broad terms, venom has evolved to fulfil one or more of the following:

• Assist prey capture by promoting immobilisation
• Assist prey death, to avoid injury to the snake

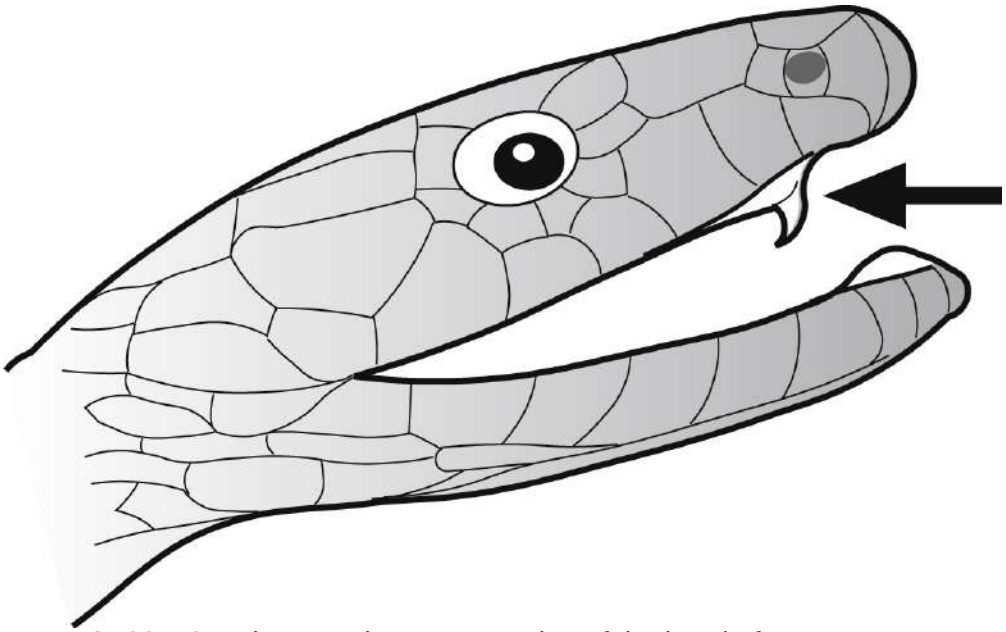

FIG. 22.1.2 Diagramatic representation of the head of a proteroglyph (front fanged) snake of the cobra type. Reproduced with permission of Dr Julian White.

• Assist prey digestion
• Act as a deterrent to predators, by causing rapid, unpleasant effects.

As humans, we tend to focus on the last function, but it is the other three functions that cause major medical problems.

Venom actions are diverse. Some major actions are listed in Table 22.1.2. From a clinical perspective, venom effects can be divided into three major groups:

1. Local effects
2. Non-specific general effects
3. Specific systemic effects.

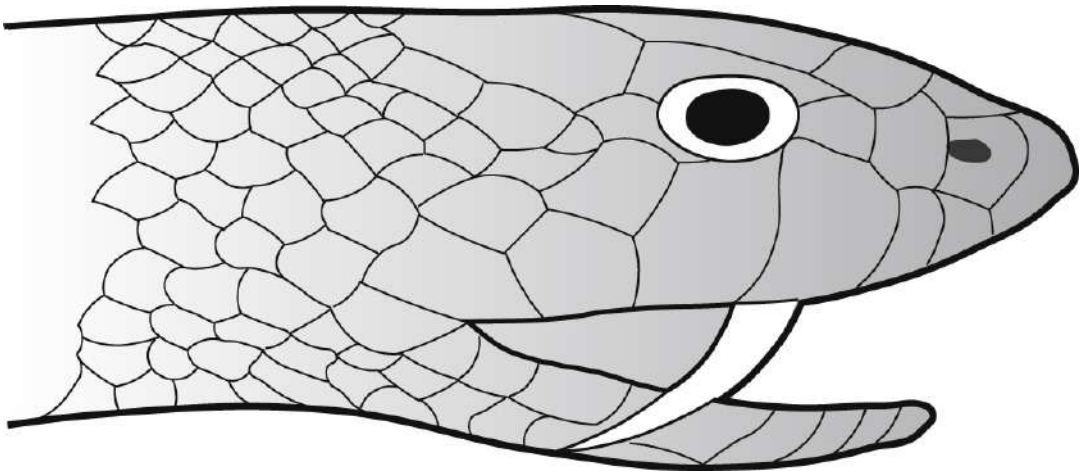

FIG. 22.1.3 Diagramatic representation of the head of a ‘side-fanged’ atractaspid snake. Reproduced with permission of Dr Julian White.

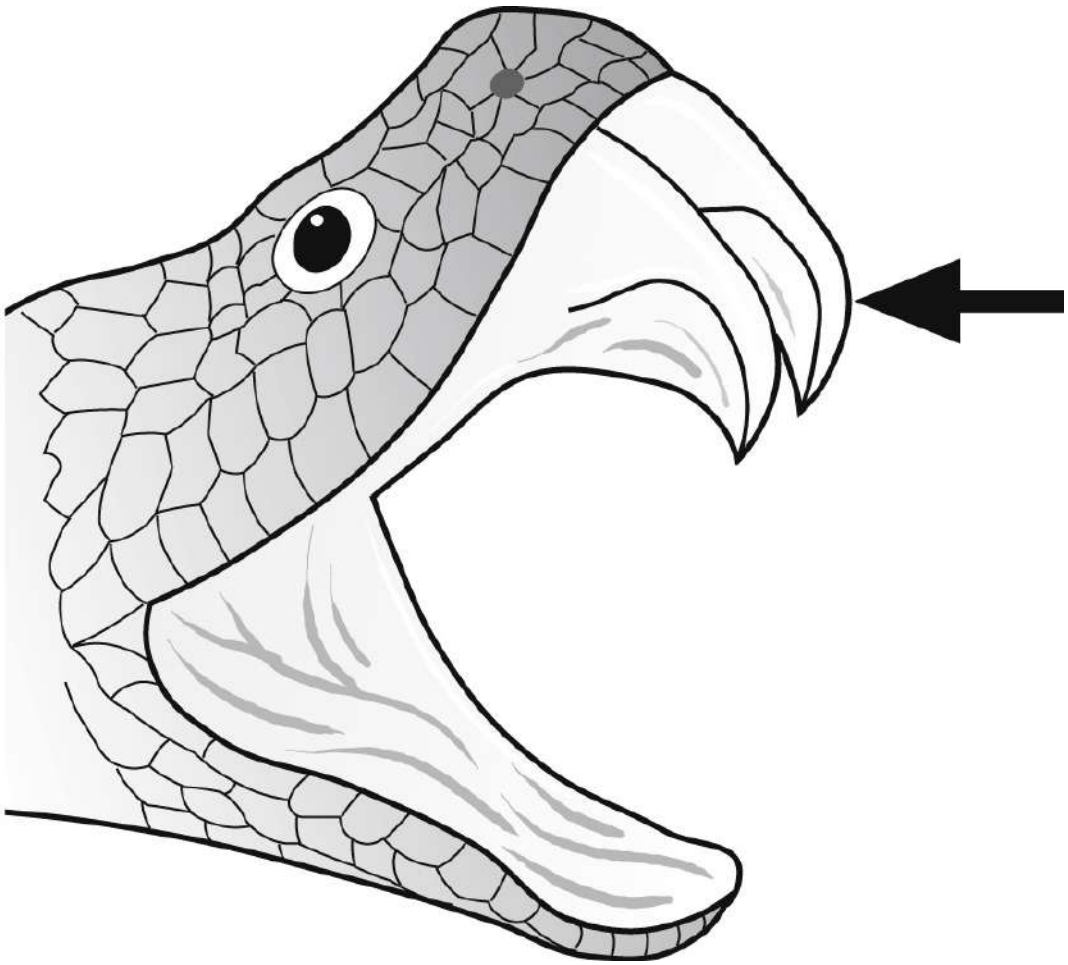

FIG. 22.1.4 Diagramatic representation of the head of a solenoglyph (front-fanged) snake of the viper type, with maxillary rotation to allow

folding of the fang against the mouth. Reproduced with permission of Dr Julian White.

Table 22.1.2

Broad overview of major clinical actions of snake venoms

Type of venom action	General site of action	Type of venom component	Clinical effects
Local toxins	Bite site and bitten limb	Necrotoxins, cytotoxins, etc.	Local effects; may include pain, swelling, blistering, bruising, necrosis
Paralytic toxins	Specific systemic (neuromuscular junction)	Neurotoxins (presynaptic, postsynaptic, dendrotoxins, fasciculins)	Progressive flaccid paralysis of skeletal muscle and diaphragm
Myolytic toxins	Specific systemic (skeletal muscle)	Myotoxins	Destruction of skeletal muscle throughout body (or locally in bitten limb only for some crotalids)
Haematological toxins	Specific systemic (interfere with haemostasis in a variety of ways, or may damage vessel walls, promote bleeding)	Procoagulants Fibrino(geno)lytics Anticoagulants Haemorrhagins Various other toxins affecting haemostasis	Varies, depending on type of toxin; may cause consumption coagulopathy, complete defibrination, active haemorrhage promoting, or even thrombosis and infarction or embolism (Martinique vipers only)
Nephrotoxic toxins	Specific systemic (kidneys)	Nephrotoxins	Renal damage, failure, or necrosis
Cardiotoxic toxins	Specific systemic (heart)	Cardiotoxins	Cardiac arrhythmias, failure or arrest

For many snakebites, in most regions of the world, local effects are a major, often the principal, medical problem. In these cases there may be local pain, swelling, which may be severe, involving much, or all, of the bitten limb, resulting in fluid shifts, secondary hypovolaemic shock and the risk of compartment syndrome. There may be local blistering, bruising or development of skin necrosis. Systemic coagulopathy may manifest locally as persistent oozing or bleeding from the bite or damaged areas of the affected limb. The extent of local necrosis may be significant, with potential, often realised, for long-term tissue injury and dysfunction. Secondary infection may develop in the injured limb. For some species (e.g. lance-headed vipers, such as *Bothrops* spp. in South America) there may be local abscess formation. Long-term disability is a frequent outcome. In some cases, amputation is required. In cases with ongoing skin damage never fully healed, skin tumours can develop after some years. This range of major local effects and secondary systemic effects can be seen following bites by many, but not all, species of viper and atractaspids but not NFFC snakes and only selected African and Asian cobras amongst the elapids. Such severe local effects are generally absent from snakebite in New Guinea and Australia, where the only medically important venomous snakes are all elapids.

The non-specific general effects of envenoming vary between species, but usually include one or more of the following:

- Nausea or vomiting
- Abdominal pain or cramps

- Headache
- Dizziness
- Non-paralytic blurred vision
- Tender or enlarged draining lymph nodes
- Brief period of collapse
- Hypertension (occasionally hypotension).

As most of these can be the result of anxiety as well as envenoming, they may not be reliable indicators of systemic envenoming.

The specific systemic effects of snake venoms are the most intensely studied, partly because they can usually be ascribed to particular venom components that can be isolated and studied in detail. An overview of these components is listed in Table 22.1.2. Specific clinical findings for the effects of these components will be discussed in the sections on ‘history’, ‘examination’ and ‘investigations’.

History

There may be a clear history of snakebite, or an encounter with a snake, where an actual bite is uncertain, or there is no history of a snake or bite. Particularly in young children, there may be no possibility of obtaining a history. Listen carefully to the story from young children, because relevant information may be disguised by rudimentary language. Some key points are listed in Table 22.1.3.

The environment and circumstances can be of great importance in deciding if a snakebite is likely. Do not assume that bites are unlikely indoors; snakes do enter houses, commonly in the rural tropics, but even in temperate urban areas such as Australian cities and country towns.

If there is a history of an encounter with a snake, note if the snake struck, how many times, if bites were through clothing, as well as the apparent length and colouration of the snake. For selected cobra attacks in Africa and Asia, the snake may spit first, particularly aiming for the eyes, before either retreating or pressing home an attack with actual bites. A chewing bite, where the snake hangs on is also important, as there is more opportunity for venom injection. Similarly, multiple bites are associated with higher rates of major envenoming.

A description of the snake and geographical location may help narrow the range of possible culprit species. This can be combined with clinical features to assist in identifying the most likely culprits using diagnostic algorithms (Figs 22.1.5 and 22.1.6).

It is important to ask about any local, general or specific symptoms that might indicate developing significant envenoming (see Table 22.1.3).

In children it may prove difficult, even impossible, to obtain any history from the child; however, parents, siblings or bystanders may have useful information. For instance, a small child seeking a parent because he/she is upset, then collapsing, having a convulsion, then recovering, but remaining miserable is a classic presentation for significant snakebite in some regions (e.g. Australia).

Examination

While examination must be thorough, time is of the essence in major envenoming. Therefore, if snakebite is suspected, examination should be directed initially to determine if there is evidence for snakebite and the extent of any envenoming. The first priority is to assess the 'ABC' (airway, breathing, circulation) and render appropriate intervention, if indicated.

It is clearly important to look at the bite site, or look for a bite, if no site is indicated from the history. Snakebites may result in single or paired fang punctures, multiple teeth punctures or even scratches, as fangs are dragged through the skin during release (Figs 22.1.7–22.1.10). If there is a bandage over the bite site, as first aid, cut a window only to inspect. Keep the removed bandage portion, if in Australia, for possible venom detection later. If venom detection is available (Australia, New Guinea), swab the bite site with the stick provided in the test kit. Do not allow anyone to clean the bite area until it has been swabbed for venom. Look for bite marks and particularly for multiple bites. Observe for local bruising, bleeding, blistering, swelling or necrosis. If there is significant local tissue injury or swelling, check pulses, etc., to exclude compartment syndrome in affected compartments. Compartment syndrome, if suspected clinically, must be confirmed by measuring intracompartmental pressure, before any consideration of surgical intervention.

Table 22.1.3

Summary of principal points in history for snakebite

Broad category	Question	Significance
Details of bite	Was snake seen?	Increases likelihood of snakebite
	Description of snake?	May assist identifying type of snake, so possible problems can be anticipated
	Size of snake?	May indicate potential for severe bite if large specimen, but beware, even juvenile snakes can inflict a severe bite
	Geographical location?	May limit types of snakes to be considered
	Environment?	May indicate likelihood of snake encounter, if no clear history of snake being seen
	Number of bites?	Multiple bites increase the likelihood of severe effects
	Was bite through clothing?	Clothing may soak up some venom, reduce the chance of an effective bite. This may also be a source for venom detection (Australia)
Details of first aid	Was first aid used?	If no first aid then nothing to impair development of effects of venom
	What type of first aid?	Some types of first aid (e.g. tourniquets, cut and suck, suction devices, snake stones, electric shock/stun guns) may make matters worse or be ineffective
		Effective first aid (e.g. immobilisation of the bitten limb, or full pressure immobilisation bandage) may delay onset of envenoming, thus the patient may present well, yet deteriorate after removal of first aid
	When was first aid applied?	If applied promptly, it may be effective, delaying envenoming If applied late or after physical activity (e.g. chasing snake, running for help) it may be ineffective
Local effects of bite	Were any bite marks, etc. noted prior to application of first aid?	If bite marks present, snakebite more likely, but absence of visible bites does not exclude snakebite (especially for some Australian elapids, notably brown snakes)
	Is there any local pain, swelling, bleeding, blistering, skin discolouration or other local effect?	May indicate likelihood of effective bite and possibly even type of bite. Absence of pain does not exclude snakebite
General symptoms	Headache, nausea, vomiting, abdominal pain?	Non-specific indicators of possible systemic envenoming (or anxiety)
	Collapse?	If in association with a definite bite, is suggestive of systemic envenoming
	Convulsion?	If in association with a definite bite is strongly suggestive of major systemic envenoming
	Blurred or double vision experienced within a few minutes of the bite?	Common effect, not likely to indicate developing paralysis
Specific systemic effects		
Paralytic effects	Presence and time of onset of paralytic symptoms? (Early ptosis may be described as heavy or sleepy eyes/eyelids)	Cranial nerves affected first, usually ptosis. Important to pick this up, before paralysis advances too far. May also help indicate the most likely type of snake
Myolytic effects	Presence and time of onset of myolytic symptoms? (Muscle pain; tenderness; weakness; urine becoming pink, red, brown or black)	Usually takes several hours to manifest. May indicate most likely type of snake
Coagulopathic and haemorrhagic effects	Presence of coagulopathy effects, such as persistent bleeding from bite site or cuts, gums, or bruising, haemoptysis, haematemesis, haematuria?	Indicates coagulopathy likely and probably significant. May indicate most likely type of snake
Renal effects	Presence of anuria or oliguria or polyuria; proteinuria	Indicates likely significant renal damage
General history		
Medications	Anticoagulants or NSAIDs?	May affect coagulation test results or increase likelihood of a major bleed if coagulopathy present
	Antihypertensives?	Though not proven for antivenoms, it is suspected that beta-blockers and ACE inhibitors may increase the chance of and severity of anaphylactic reactions to antivenom
Past history	Past bites requiring antivenom?	Past exposure to antivenom may increase the likelihood of reactions to subsequent antivenom therapy
	Past renal problems?	May increase the likelihood of envenoming causing renal damage
	Other past medical history?	Evaluate as appropriate

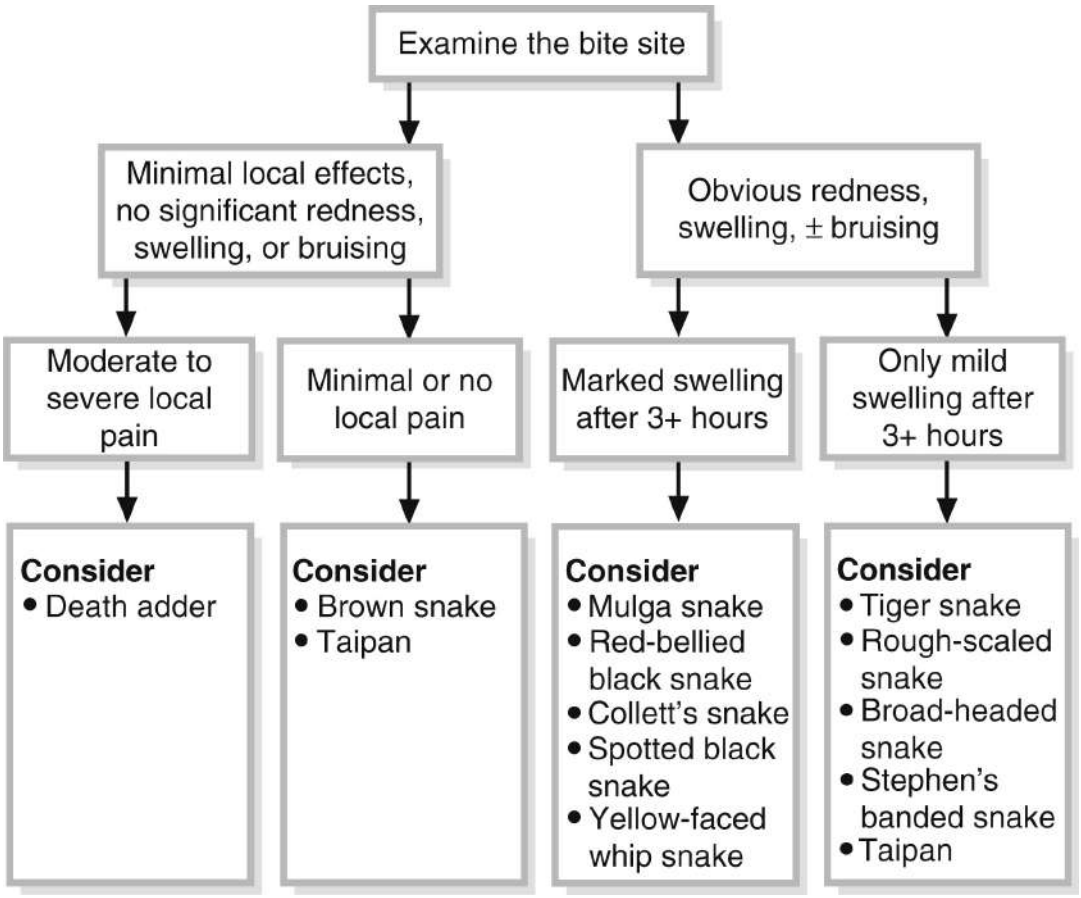

FIG. 22.1.5 Diagnostic algorithm for Australian snakes using the bite site effects. Reproduced with permission of Dr Julian White.

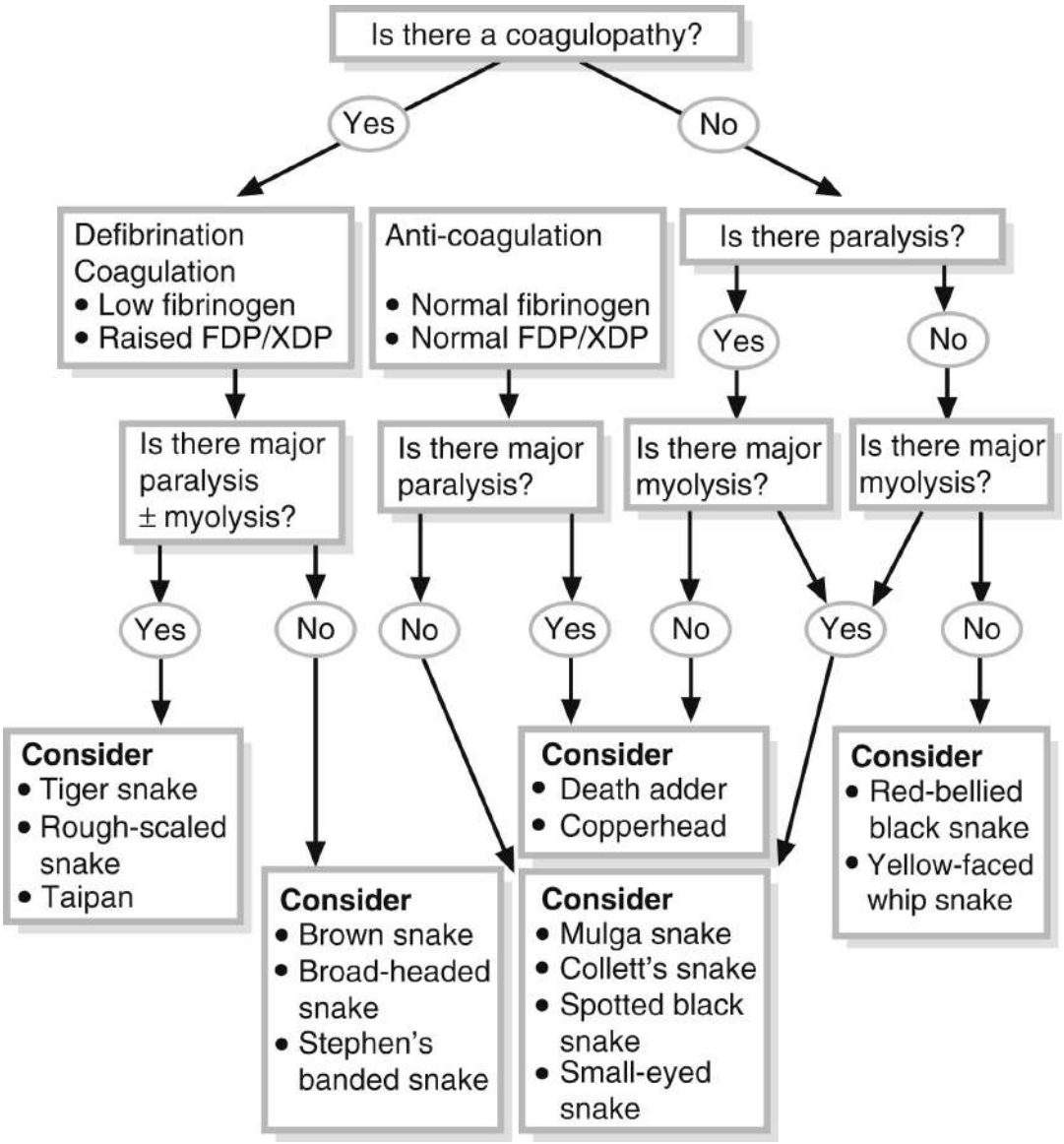

FIG. 22.1.6 Diagnostic algorithm for Australian snakes using the systemic effects of the bite. Reproduced with permission of Dr Julian White.

Check draining lymph nodes; if they are tender or swollen it may indicate venom absorption and movement.

Examine for specific effects, notably neurotoxicity (flaccid paralysis; check for cranial nerve paralysis first, starting with ptosis; Figs 22.1.11 and 22.1.12), myolysis (muscle tenderness and weakness), coagulopathy (persistent bleeding from bite site, needle punctures, gums, etc.; Fig. 22.1.13) or deep vein thrombosis (DVT) (pulmonary embolism; Martinique crotalids only), cardiotoxicity (arrhythmias), 'allergy' (angioneurotic oedema; particularly European vipers).

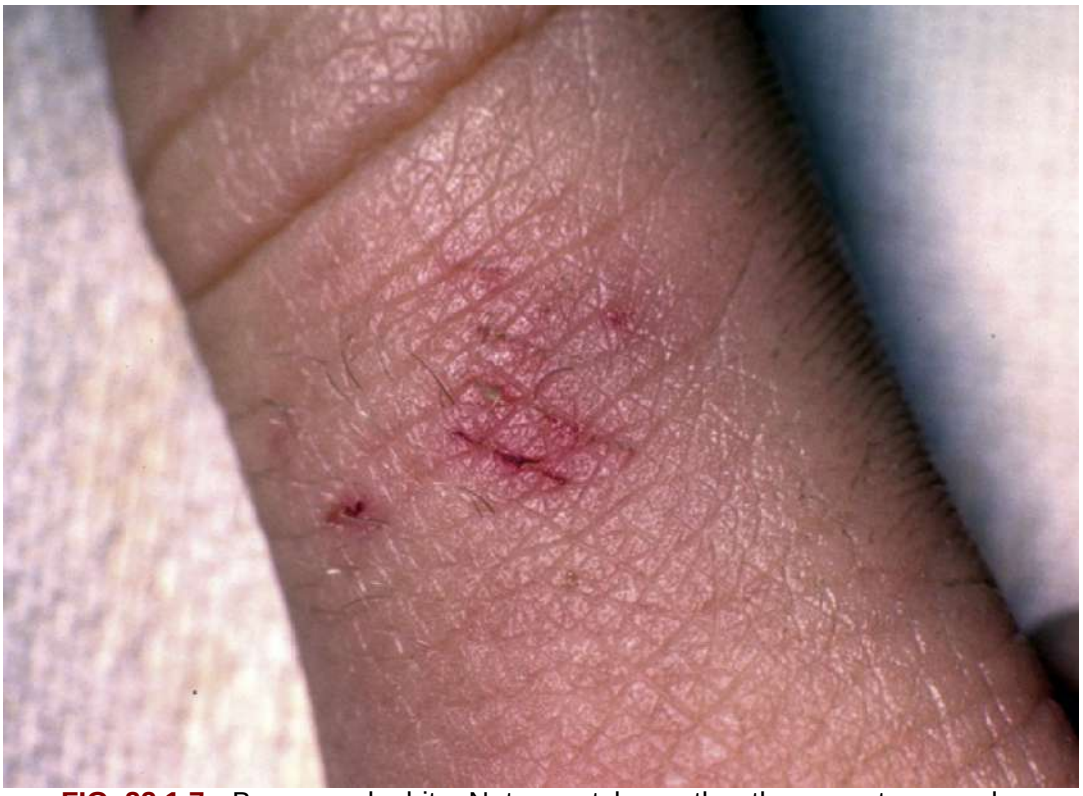

FIG. 22.1.7 Brown snake bite. Note scratches rather than punctures and lack of local reaction. Reproduced with permission of Dr Julian White.

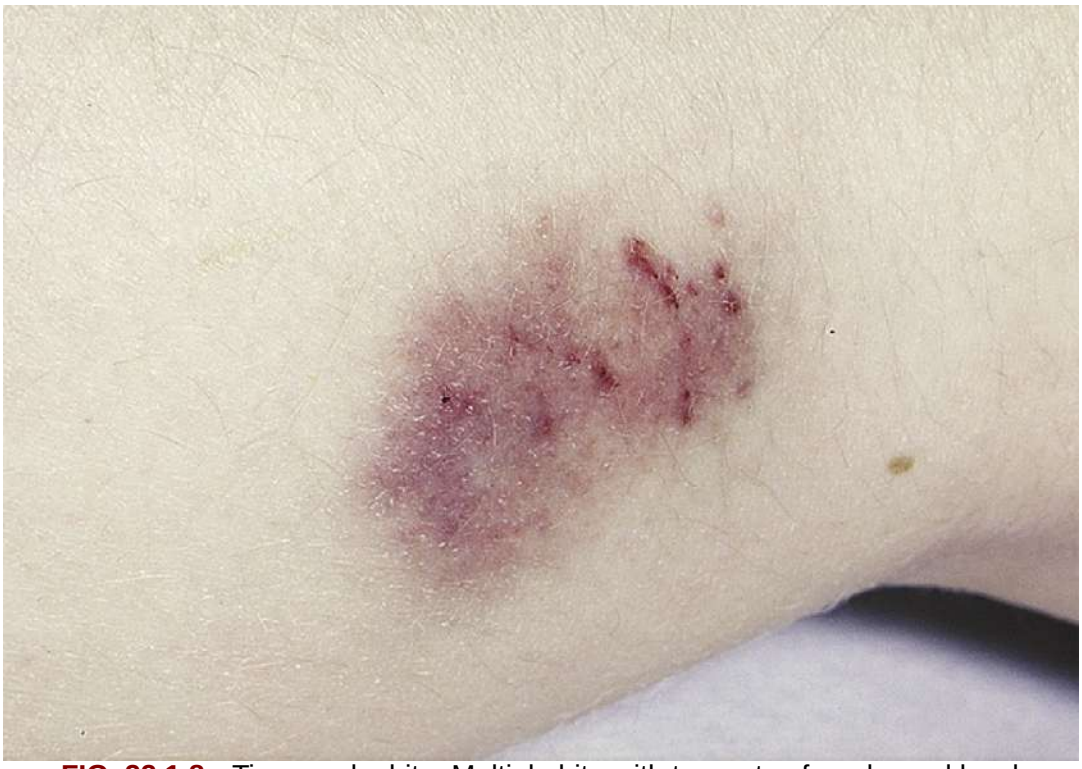

FIG. 22.1.8 Tiger snake bite. Multiple bite with two sets of marks and local bruising. Reproduced with permission of Dr Julian White.

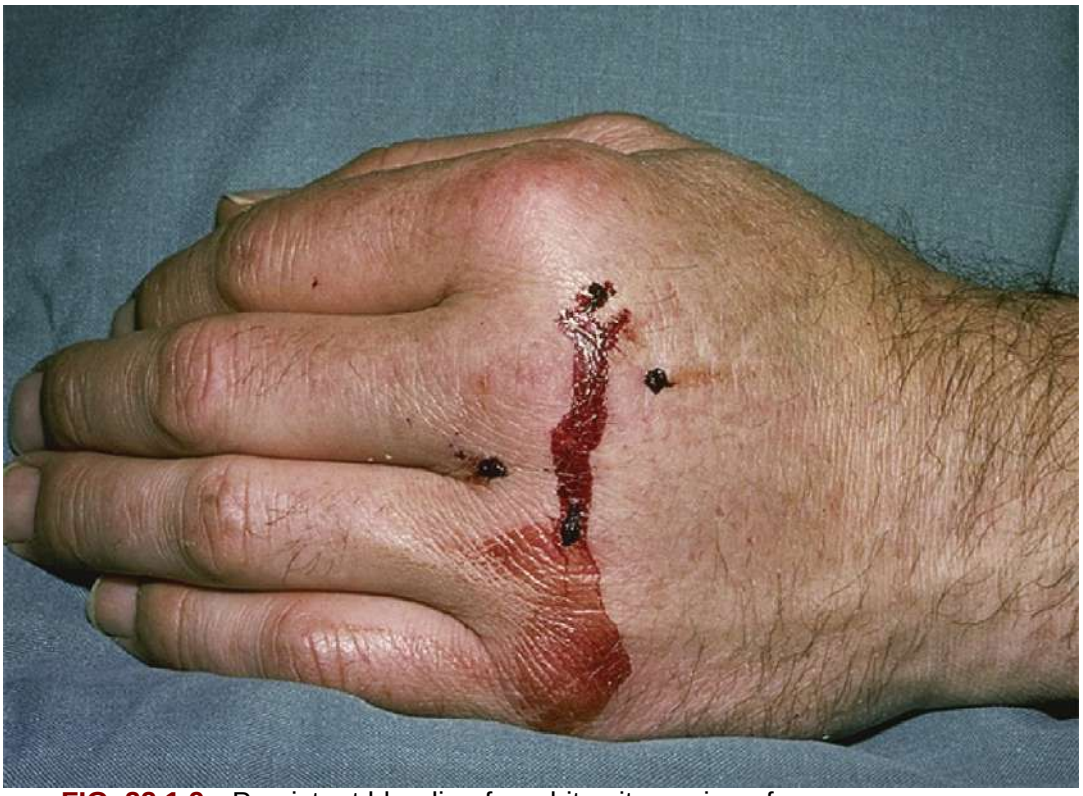

FIG. 22.1.9 Persistent bleeding from bite site, a sign of coagulopathy. Reproduced with permission of Dr Julian White.

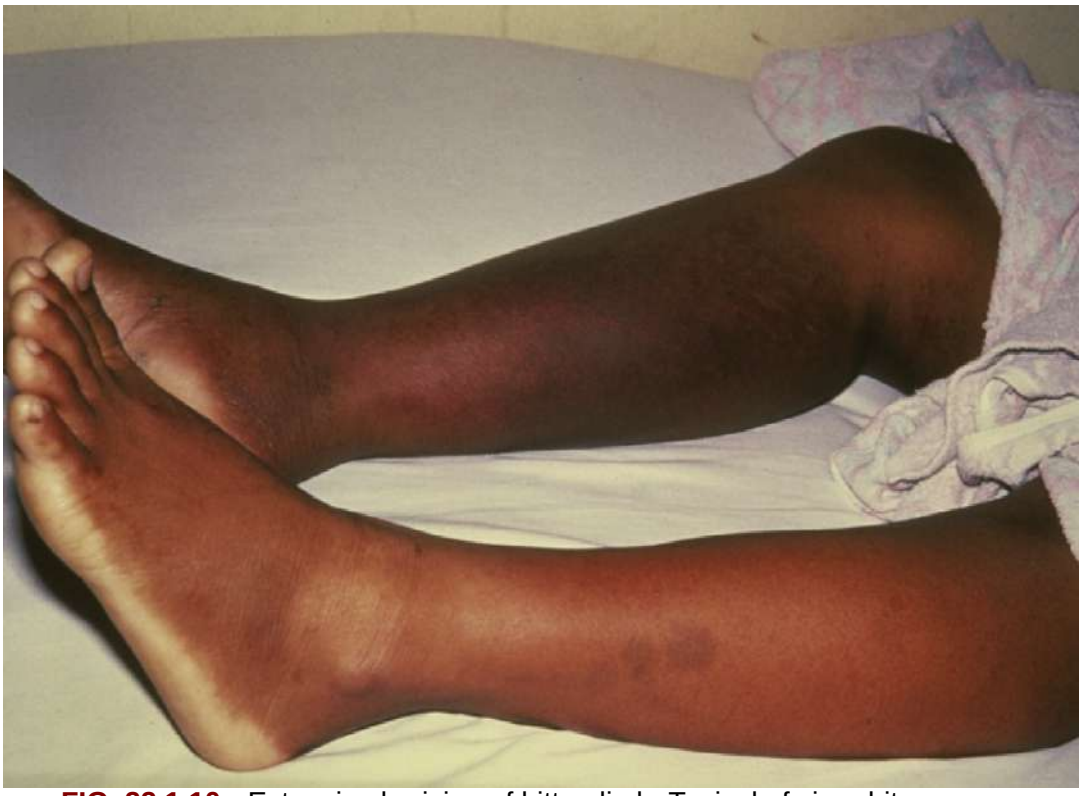

FIG. 22.1.10 Extensive bruising of bitten limb. Typical of viper bites causing coagulopathy (green pit viper bite). Reproduced with permission of Dr Julian White.

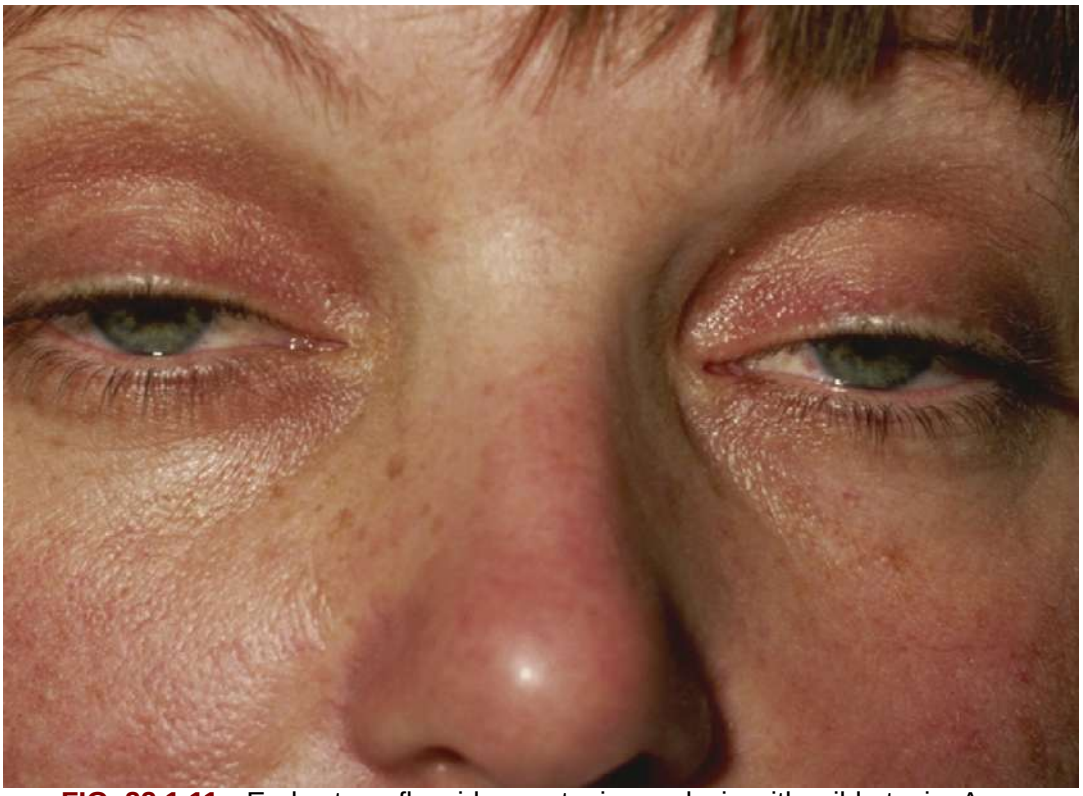

FIG. 22.1.11 Early stage flaccid neurotoxic paralysis with mild ptosis. An important early sign, easily missed (tiger snake bite). Reproduced with permission of Dr Julian White.

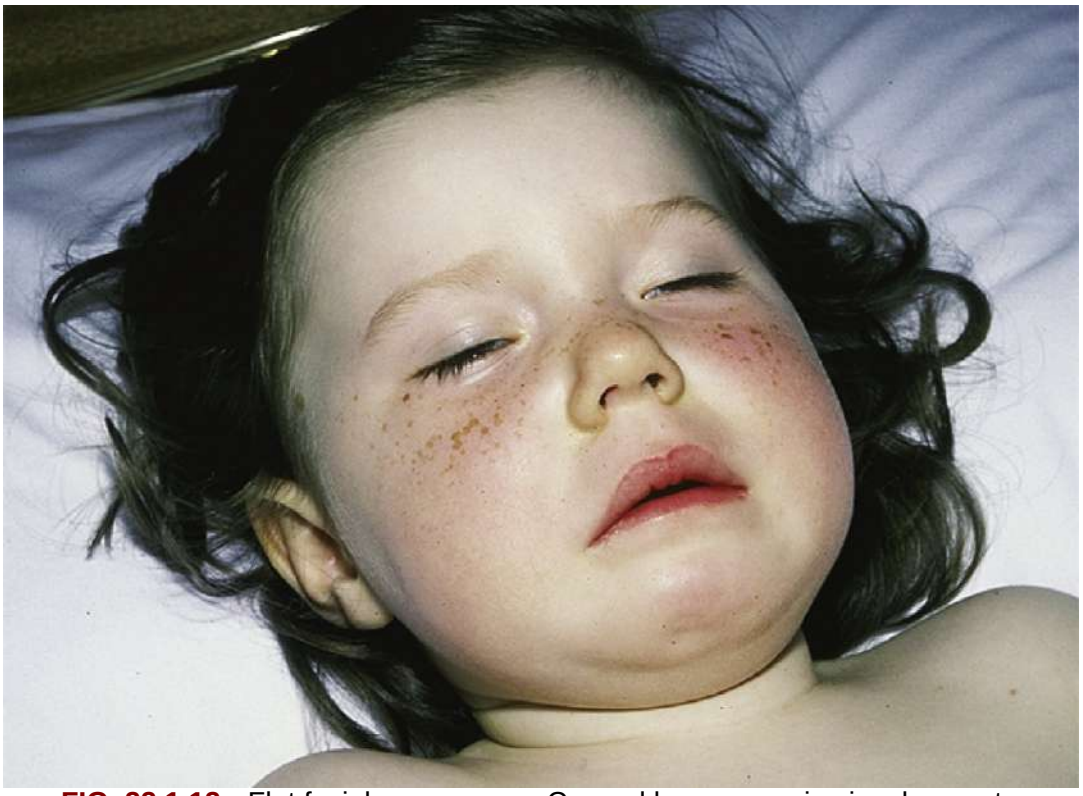

FIG. 22.1.12 Flat facial appearance. Caused by progressive involvement of cranial nerves in flaccid neurotoxic paralysis. Ptosis is also present (tiger snake bite). Reproduced with permission of Dr Julian White.

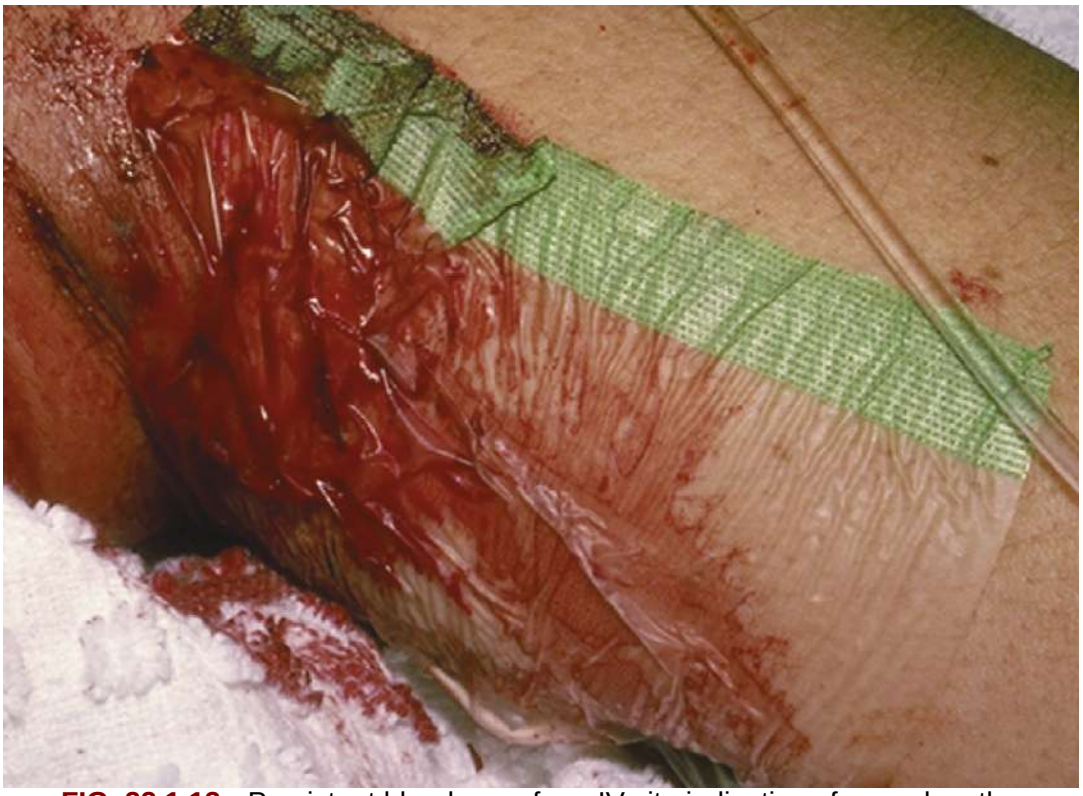

FIG. 22.1.13 Persistent blood ooze from IV site indicative of coagulopathy (taipan bite). Reproduced with permission of Dr Julian White.

Investigations

The most specific investigation is venom detection, but currently this is only routinely available in Australia (most reliable sample is bite site swab; urine can be tested if there is systemic envenoming; blood is less reliable). However, venom detection will not always provide a useful answer, even if available, so it is important to be aware of other diagnostic tools in determining the type of bite and clinical effects. These are discussed further under ‘differential diagnosis’. Venom detection is only designed to determine the type of snake and therefore the type of antivenom; it is not reliable as a screening test for snakebite or envenoming and should not be used for this purpose.

Laboratory or similar investigations are often crucial to the management of snakebite. The key areas are coagulation, renal function and muscle integrity.

Many snakes, especially vipers but also some NFFC snakes and many Australian elapids, can cause coagulopathy, which in many cases is potentially lethal. Coagulopathy can develop early or gradually over many hours. The type of coagulopathy is determined by the type of venom components, but just a few

tests are adequate in most situations to determine the extent of pathology. For rural areas or hospitals without laboratory facilities, including outback Australia, the 20-minute whole blood clotting test (20WBCT) is the only practical test. Twenty-five millilitres of venous blood is placed in a clean, glass test tube or similar and allowed to clot. If possible, the time to clot is measured. The tube is gently inverted after 20 minutes and if there is no clot present or only a tiny clot, with fluid blood, this is a positive test, indicating coagulopathy with non-clotting blood MAY be present. Normal blood should clot in under 10 minutes. If possible, perform a parallel test on blood from a normal control (e.g. relative or staff member). If laboratory facilities are available, the key tests are prothrombin time (PT) or international normalised ratio (INR), activated partial thromboplastin time (aPTT), fibrinogen titre, fibrin (ogen) degradation products (or D-dimer) titre and platelet count. In consumptive snakebite coagulopathy (defibrination, sometimes labelled ‘VICC’) in the early stages in less severe cases the D-dimer may be the first test to show an abnormality.

Renal function tests are usually urea and creatinine levels. In the absence of a laboratory, monitoring renal output is all that is practical. For bites by snakes prone to causing AKI (e.g. Russell’s viper in Myanmar, parts of India, Sri Lanka), early proteinuria or haemoglobinuria may indicate developing renal injury.

Muscle integrity relates to myolytic venoms, the best measure being creatine phosphokinase level (CK, CPK). In the absence of a laboratory, the presence of red, brown or black urine is suggestive of myolysis and myoglobinuria. However, red urine can also be caused by haematuria. If in doubt, spin down the urine and examine under a microscope, looking for evidence of red cell casts. Both haemoglobin and myoglobin test positive for blood with dipstix testing of urine.

If there is evidence of infection around the bitten area, culture and sensitivity should be performed on wound swabs.

In cases where there is clinical evidence of cardiovascular effects of envenoming, primary or secondary, or for bites by snakes known to be cardiotoxic, ECG monitoring is appropriate, but in other cases it may be unnecessary.

Chest X-ray (CXR) is only required if there are clinical grounds to suspect respiratory pathology. Similarly, arterial blood gas examination is not routinely required but could be considered if there is respiratory impairment, particularly if there is respiratory paralysis developing. In the later stages, after extensive

intravenous (IV) fluid therapy, secondary pulmonary oedema is a risk, especially in young children; if suspected, a CXR may be diagnostic.

Envenoming does not always manifest early. It is therefore appropriate to retest for coagulopathy, renal impairment and elevated CK, if the initial tests are normal. In general, a useful protocol is to retest 2–3 hours and 5–6 hours after the initial test, or earlier if symptoms or signs of envenoming develop and at 12 hours or later post bite, prior to any decision to discharge.

Differential diagnosis

A full discussion of all possible differential diagnoses for snakebite is beyond the scope of this chapter. It is important to include snakebite in the differential diagnosis for patients with unexplained collapse, convulsions, bleeding, coagulopathy, thrombosis (in Martinique, specifically), myolysis, flaccid paralysis, muscle fasciculation (mamba bites in Africa), renal failure or impairment, or local tissue injury.

Differential diagnosis can also be applied within snakebite, in determining the type of snake most likely to have caused the bite. Diagnostic algorithms have been developed for Australia (see Figs 22.1.5 and 22.1.6) and South-East Asia (Fig. 22.1.14). These are based on cases with significant envenoming and will not function if the patient is not envenomed, though this hardly matters, as such a patient will not require antivenom therapy. In some regions, notably Australia, it is important to know the type of snake involved, because antivenom therapy can be targeted appropriately. A similar situation applies in some other regions, where specific antivenoms are available. In regions such as North America this is less important, because there is only one polyvalent antivenom, which covers all venomous species except coral snakes.

Treatment

Snakebite treatment can be divided into several areas – first aid, diagnosis and treatment – the latter is further divided into specific (antivenom) and non-specific treatment.

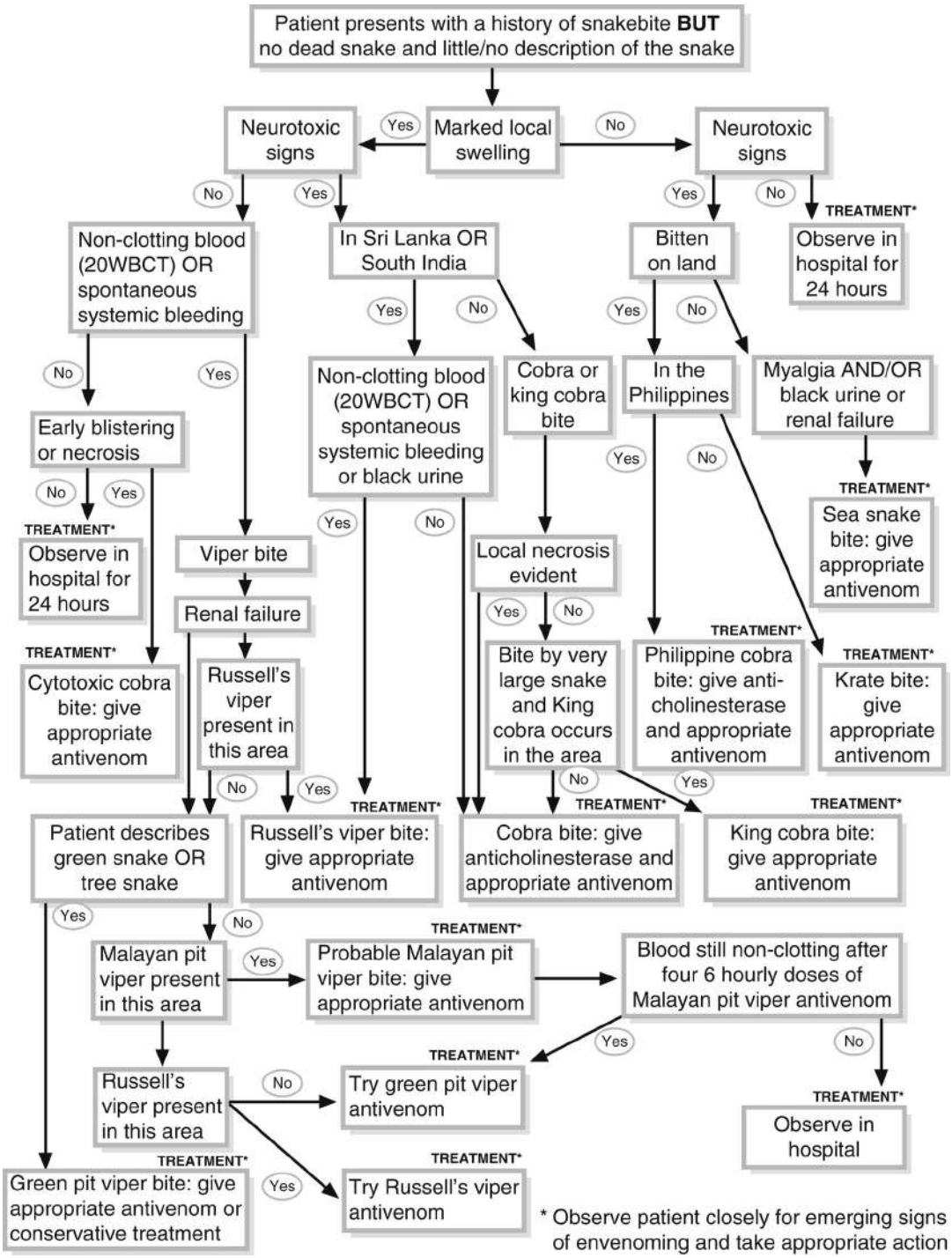

FIG. 22.1.14 Diagnostic algorithm for snakebite in southeast Asia. After Warrell et al. Reproduced with permission of Dr Julian White.

First aid for snakebite is controversial. Many techniques have been advocated and are in use throughout the world. Almost none meets the critical criteria of safety and effectiveness. For snakes not likely to cause major tissue injury in the bitten area, the Australian-developed 'pressure immobilisation' method is

appropriate. A broad bandage is applied over the bite site, then the rest of the bitten limb, at the same pressure as used for a sprain, firm but not occlusive. The limb is then immobilised using a splint. Correctly and promptly applied, this method is both safe and effective. However, for snakes likely to cause local tissue injury, even the pressure of this technique may cause further tissue damage, at least theoretically. For this reason, the pressure immobilisation method has not been recommended for all snakebites. The theoretical danger from this method has been challenged by recent research and it may be that extension of this research will show that the pressure immobilisation method is safe and effective for all snakebites.

Other popular first aid methods enjoy no such success and are either unsafe or ineffective, or both, and should never be used. Amongst these are tourniquets, ‘cut and suck’, patent venom extraction devices (suction), electric shock (‘stun guns’, etc.), application of chemicals to the bite, snake stones and ‘witch doctor’ treatments. The application of certain plant extracts is still undergoing evaluation.

Diagnosis of snakebite has been discussed earlier.

Definitive treatment for snakebite will vary depending on the type of snake, but some general principles apply.

First, not every bite will result in envenoming, but the extent of envenoming, if any, may not be immediately apparent, therefore all bites should be treated with caution.

Second, in many regions, the bulk of snake fauna is non-venomous, so many snake bites may be trivial. However, it is necessary to be sure of the snake’s identity as non-venomous before dismissing the case and identification is rarely easy, especially in paediatric cases where history is scant. An exception is Australia, where most snakes causing bites are potentially lethal.

Third, if significant envenoming has occurred, with few exceptions, antivenom, if available, is the treatment of choice and should always be given IV. Choice of antivenom will be determined by the type of snake and the availability of methods to determine snake identity. Thus, in Australia, specific antivenoms are available, together with venom detection and diagnostic algorithms, so polyvalent antivenom is often not required. In contrast, in North America the only available snake antivenom is polyvalent, covering all endemic pit-viper species, so identifying the snake is less important. In general, antivenom will be more effective than any other therapeutic agent at reversing envenoming. Used appropriately it is life saving and the old ‘wisdom’ that ‘the

antivenom is more dangerous than the venom' is outdated, inappropriate and dangerous.

Fourth, antivenom is generally disappointing as therapy for local effects of envenoming, but is still better than other therapies in most cases. Equally, do not overlook adjunctive therapies, in particular adequate IV hydration if there is massive local or limb swelling following the bite, as untreated hypovolaemic shock secondary to such fluid shifts is potentially lethal, especially in children.

Fifth, for coagulopathy caused by venom, antivenom is the best treatment to reverse effects, and factor replacement therapy, including even whole blood, is best reserved for those cases with catastrophic bleeding, or no available antivenom, or where sufficient antivenom has already been given to neutralise all venom. Giving factor replacement therapy while active venom is still circulating is to invite worsening of the coagulopathy. Heparin is generally ineffective in these cases and should be avoided.

Finally, for cases with flaccid paralysis, consider anticholinesterase therapy as an adjunct to antivenom, if a Tensilon test has shown benefit (there is likely benefit for cobra bite causing paralysis, death adders [but only some cases], and sea snakes and possibly some kraits and some coral snakes). Venoms with presynaptic neurotoxins will not show response to anticholinesterase therapy (most Australian snakes causing paralysis; except the death adder, which sometimes has only postsynaptic neurotoxins).

From the above it will be clear that antivenom is the key treatment for snakebite, when available. The latter is a real issue, because for many species and substantial areas of the rural tropics, antivenom is not available.

Prognosis

The majority of all snakebites globally prove non-fatal, but with an estimated global fatality of more than 100,000 per year, death is clearly a significant risk. This is especially true for children. Their lower body mass and often delayed application of appropriate first aid put them at greater risk. With such a wide variety of snake species, it is beyond the scope of this chapter to define prognosis for all snakes. However, some general principles apply.

The more rapid and severe the onset of envenoming, the more grave the prognosis, but this is not absolute. For instance, in Australia, a small child (under 5 years) may show early irritability, collapse, even convulsions following a snakebite (especially bites by brown snakes, tiger snakes, taipans), yet will

usually spontaneously recover consciousness. Such a presentation is indicative of major envenoming, but with correct treatment, is survivable. Thus prognosis is determined by several factors, not just the type and toxicity of the snake, but also the treatment response. It is likely that if high-quality standard treatment were universally available, the global toll from snakebite would be far lower.

For local effects of envenoming, after bites by snakes causing local tissue injury (most vipers, many African and Asian cobras), the more rapid the swelling and the more extensive the blistering, the more serious the bite. Similarly, development of discolouration of the skin with a well-demarcated edge often indicates an area of impending necrosis.

For general systemic effects, the more severe the symptoms, such as vomiting, abdominal pain, headache, often, though not universally, the more severe the envenoming.

For specific systemic effects, rapid onset often indicates severity. Early development of progressive flaccid paralysis is usually indicative of severe envenoming, while paralysis of very limited extent after 6 hours is often indicative of a less severe bite. However, this may not always be the case. Occasionally flaccid paralysis may not be evident and a patient with just ptosis at 24 hours post bite may still progress to severe paralysis without treatment. This may occur with many snakes causing paralysis, but most notably Australian death adders. This is an important reason why early discharge of an apparently well patient is ill advised. Once flaccid paralysis is extensive, with respiratory failure, the patient can still survive if ventilation is supported. However, for snakes with presynaptic neurotoxins, reversal of paralysis will not occur until the damaged terminal axons at neuromuscular junctions have recovered, which may take days to weeks. Antivenom will not reverse such paralysis. In contrast, paralysis caused by snakes with purely postsynaptic neurotoxins will usually reverse with adequate antivenom therapy. The Tensilon test will generally predict such responses.

For coagulopathy, rapidity of onset and presence of signs such as persistent oozing from the bite site and bleeding gums indicate significant coagulopathy. With appropriate treatment, this does not imply a poor prognosis but is a warning that more critical haemorrhaging is possible. Any bleeding into a vital organ, most commonly the brain, indicates a poor prognosis, with a fatal outcome most likely with intracranial bleeds. For Russell’s viper in Myanmar (Burma) and southern India, there may be haemorrhagic infarction of the anterior pituitary gland, with resulting Sheehan’s syndrome developing; though this may not be

rapidly apparent.

Myolysis is most often measured as profoundly elevated CK levels and may progress to secondary renal failure and hyperkalaemia, the latter indicating a poor prognosis, as cardiac complications may ensue. In general, the more rapid the rise in CK levels, the more severe is the myolysis, but this is not always the case. In some cases the CK rise may initially be slight, but become more significant after 24 hours, rising to high levels over several days. Early muscle pain and myoglobinuria are also suggestive of more severe myolysis.

Renal damage may be primary or secondary and can prove lethal if untreated. Rapid development of anuric renal failure indicates a poor prognosis, unless dialysis can be instituted. A slower rise in creatinine levels, without anuria, usually indicates less severe renal damage but still may take a week or more for return of normal renal function. In most cases, even acute renal failure after snakebite is reversible, over days to weeks. A small minority of cases will develop more lasting renal failure, generally because bilateral renal cortical necrosis has occurred. This severe complication with a poor prognosis is not easily predicted and is generally only discovered at renal biopsy in patients who have failed to recover from early renal failure. It has been reported after bites by only a few species, such as the Australian taipan and South American lance-headed pit vipers (jararacusu, jararaca), but other species could potentially cause this outcome.

Prevention

Prevention of snakebite can be considered in two ways. First, there is prevention of bites, by educating the population about ways to avoid contact. These will vary from region to region and are beyond the scope of this chapter. Second, there is prevention of the more severe effects or complications of envenoming, by prompt diagnosis and appropriate treatment. This commences with early application of appropriate first aid pre-hospital, to minimise the chance of severe envenoming developing before treatment can be instituted. Once in hospital, urgent triage and assessment will permit prompt IV rehydration, for cases where there is a major fluid shift into the bitten limb, or rapid commencement of IV antivenom, if indicated, before more delayed forms of envenoming (e.g. flaccid paralysis, myolysis) have progressed too far, as well as instituting any life-support measures required.

Many deaths or cases with long-term morbidity after snakebite are the result

of either delays in commencing treatment or inadequate or inappropriate treatment. The latter may be the result of poor training of health personnel. It follows, therefore, that adequate training of staff will be preventative.

Controversies

1. First aid – Possibly the most controversial aspect of snakebite management is the recommended type of first aid. Many types of first aid have been advocated and remain in widespread use, but only immobilisation, or pressure bandaging and immobilisation have consistently enjoyed both scientific and medical expert support (see earlier discussion on first aid).
2. Type of antivenom – The role of antivenom in treatment should no longer be considered controversial, as where available, if of reasonable quality, it is the treatment of choice. Less certain is the role of antivenom in treating purely local effects of envenoming, especially in preventing necrosis. Recent clinical experience suggests that high-quality antivenoms, particularly Fab′ antivenoms, are at least able to lessen the extent of local tissue injury, if used promptly. No antivenom can be expected to reverse established local necrosis. However, debate continues on the relative merits of different types of antivenom. The most recent Fab' antivenoms have proven clinically effective, but are rapidly cleared, requiring higher and repeat doses. $F(ab')_2$ antivenoms are less rapidly cleared and are effective. Whole IgG antivenoms have the highest rate of adverse reactions but are cheaper and may be the most potent. New methods of producing whole IgG antivenoms are proving cost effective and appear to significantly reduce the adverse effect profile, so there is a likely resurgence of safe, effective IgG antivenoms. The choice of animal is also contested. Horses, while traditional and easy to use in most regions, produce an antivenom with higher rates of adverse reactions than sheep, but sheep can only be used safely if raised in regions free from prion diseases, essentially limiting them to Australia and New Zealand. IgY antivenoms from egg yolk, produced by immunising chickens, are potentially easy and cheap to produce, but their safety and effectiveness are not yet established and recent research indicates both limits in effectiveness and a likely unacceptable adverse effects profile.

3. Premedication – There remains debate on the value of premedication prior to antivenom therapy to reduce the likelihood of acute adverse reactions, especially ‘anaphylaxis’. Antihistamines have been shown to be ineffective at preventing such reactions. They have adverse side effects (drowsiness and occasionally hyperexcitability) and should not be used. Hydrocortisone carries no proven benefit but no great risk. Adrenaline (epinephrine) remains the most controversial, as it can reduce the incidence of reactions for poor-quality antivenoms, but has a significant risk profile, so for most antivenoms its use has not been recommended. It may be particularly dangerous for bites likely to cause coagulopathy (e.g. many Australian snakebites, most viper bites). However, recent studies in Sri Lanka have demonstrated that, at least in that setting, such premedication with SC adrenaline (epinephrine) is associated with better patient outcomes using antivenom, compared to no premedication and, in consequence, such premedication is recommended by the WHO. The practice of pre-testing for allergy to antivenom, using a small subcutaneous dose of the antivenom is to be discouraged. It has been shown to have no reliable predictive value, but carries a significant risk, without benefit, and will delay commencement of antivenom therapy.
4. Coagulopathy – The treatment of coagulopathy remains contentious, though most experts agree antivenom is the best therapeutic choice, if available. Factor replacement therapy is often the only option if no antivenom is available, as for some NFFC snakes, but it is not without hazard. Heparin has been advocated, but most evidence suggests that it is both ineffective and dangerous in this setting.
5. Local necrosis – The treatment of local swelling should be standard, with fasciotomy reserved for those few cases with proven compartment syndrome (intracompartmental pressure measurement). Fasciotomy should be avoided, if possible, in cases with active coagulopathy. However, early fasciotomy is still practiced in some areas, often with distressing and unacceptable functional and cosmetic sequelae.
6. A clinical trial has raised controversy regarding the role of AV in Red Back Spider bite but confirmatory evidence is currently lacking and clinical experience suggests some improvement in symptoms with AV.

Future Directions

1. Even for many common species of venomous snakes, known to cause significant numbers of bites, reliable clinical studies of envenoming are scant or lacking. The medical literature on snakebite is replete with epidemiological studies that fail to relate bite effects to particular species of snakes, rendering these studies almost useless. It is essential that accurate profiles of the clinical spectrum of envenoming be documented for every species biting humans, preferably in controlled prospective studies.
2. Controlled studies to establish antivenom effectiveness and dosage are required. Use of modern techniques to measure venom and component levels in serial patient blood samples should greatly assist in such research. Antivenoms need to become safer, more effective and much more widely available, particularly in the rural tropics.

Scorpion stings

Introduction

Scorpion stings are the second most important form of terrestrial envenoming, after snakebite, with global cases probably exceeding 1,000,000 per year, and deaths numbered in the many hundreds, to possibly as high as 5000 per year, nearly all in children. Scorpion envenoming is unpleasant for adults and occasionally is severe enough to threaten life. In children, however, it can be a rapidly severe and lethal disease, with some centres still reporting paediatric fatality rates in excess of 10%.

Scorpions vary in size, with around 2000 species known. All have a sting in the ‘tail’ (telson) with associated venom glands. Most scorpions either rarely sting humans, or are too small to cause envenoming, or have venom of little potency in humans. Unfortunately, a small number of scorpions do possess potent venoms and these species predominate in parts of the world where humans exist in large numbers, often in less than affluent conditions. The combination of warm to hot evenings, sandy soils, a tendency to walk around barefoot and dwellings that do not exclude scorpions leads to the large number of stings. Major risk areas include South and Central America, particularly

Brazil (*Tityus* spp.), Mexico and adjacent USA (*Centruroides* spp.), North Africa and the Middle East (*Leiurus quinquestriatus*, *Androctonus* spp., *Buthus* spp.), Western Asia and India (*Buthus* spp., *Hottentotta* spp.) and in Iran, the unique *Hemiscorpius lepturus*.

In general, it is not the larger scorpions with robust front 'pincers' that are most concerning, but the smaller, more delicate species with unimpressive front 'pincers', because they rely on the toxicity of their venom.

Scorpion venoms contain a wide array of ion-channel toxins of great potency, causing an excitatory neurotoxic reaction (not paralysis), not dissimilar to an autonomic storm. Only a matter of minutes, not hours, may elapse from the time of the sting to major systemic envenoming. Once the systemic toxicity is established, antivenom therapy has less chance of success, though it may still save lives. In Mexico, with >280,000 cases per year, death rates in children following scorpion sting have fallen from thousands per year to a handful following the introduction of antivenom.

Scorpion venoms do not contain paralytic neurotoxins, myolysins, components affecting coagulation or renal function, nor do they contain local necrotic toxins (except for one species in the Middle East; *Hemiscorpius lepturus* in Iran).

History

Often a scorpion will have been seen. There will usually be a clear history of an immediately painful sting (except *Hemiscorpius lepturus*), followed by development of systemic envenoming with effects that may include some of the following:

- Tingling of the lips
- Nausea, vomiting
- Abdominal pain
- Collapse
- Convulsions
- Hypertension or labile blood pressure
- Increased sweating, salivation or lacrimation
- Piloerection
- Dyspnoea
- Pulmonary oedema

- Cardiac collapse
- Multiple organ failure.

Symptoms appropriate to each of these effects may be described. It is important to note the time of onset for symptoms – a rapid onset and escalation in severity indicate a severe sting.

Examination

The local effects of the sting are not generally impressive, though there may be local sweating and piloerection. It is the systemic effects that will be most important, so particularly check blood pressure, look for signs of neuroexcitation, pulmonary oedema and cardiac collapse. In small children there may be a nystagmus. The exception is *Hemiscorpius lepturus* in parts of Iran; this species causes severe local effects, plus systemic effects including intravascular haemolysis, multiorgan failure and shock, and children are particularly affected, with a significant fatality rate.

Investigations

There are no specific tests for scorpion venom, nor are there specific indicators of envenoming, but in severe cases it is important to exclude secondary effects of envenoming and multiple organ failure.

Differential diagnosis

Full differential diagnosis of scorpion sting is beyond the scope of this chapter. The ‘autonomic storm’ clinical picture seen in severe scorpion envenoming can also be caused by some other venomous animals, particularly funnel web spiders (in Australia, where major scorpion stings do not occur), banana spiders (in Brazil, typically also causing priapism in boys) and some jellyfish (irukandji type). Accidental or deliberate exposure to certain pesticides and pharmaceuticals should also be considered.

Treatment

Treatment of major scorpion envenoming is controversial, particularly centring on the role of antivenom. Most evidence suggests that antivenom use has

resulted in greatly reduced fatality rates in children, but a few doctors argue that pharmacotherapy is more effective than antivenom, particularly focusing on the cardiac failure seen in fatal cases. Prazosin, in particular, has enjoyed success and should be considered, both as an adjunct to antivenom and as first-line therapy in the absence of antivenom (i.e. in India), but currently in India, antivenom has replaced prazosin as the preferred treatment. If antivenom is available it should be used IV without delay. Dose will vary depending on product.

Prognosis

The prognosis in scorpion envenoming depends on several factors. More severe envenoming is likely in smaller children, with more rapid development of effects and a shorter window for effective antivenom therapy. If multiple organ failure develops then prognosis is generally poor.

Prevention

Most scorpion stings occur because of the patterns of human living, and thus are theoretically avoidable. It is possible to 'scorpion-proof' houses by using tiles around the lower walls thus preventing scorpion entry. Use of enclosed footwear and avoidance of sitting down or lying down outside after dusk can also reduce sting incidence.

Controversies and Future Directions

1. The major controversy in management of scorpion sting is the issue of antivenom effectiveness, as discussed earlier. Of the various types of antivenom, animal studies have indicated that Fab' antivenoms are not more effective than $F(ab')_2$ or IgG antivenoms.
2. Among non-antivenom therapies, the most controversy has surrounded the proposal to use insulin, suggested by an Indian doctor (where no antivenom was previously available). This technique is not favoured by most experts, considering it is both highly risky and of most uncertain theoretical and practical benefit.
3. Despite the frequency of scorpion stings, there are still few published studies of series of stings from particular species and few trials of

various methods of treatment. There is a need for more intensive systematic study of scorpion envenoming and controlled trials of treatment alternatives.

Spiderbite

Introduction

Spiderbite is probably very common, but most bites are trivial, with only a few species likely to cause major harm to humans (Table 22.1.4). For these species, morbidity can be significant, but mortality is low, with global deaths directly related to spiderbite probably measuring 20 or less per year. Even the world's most dangerous spiders, the Australian funnel web spiders, have only caused one known fatality in the last 20 years. As with other venomous animals, spiderbite is more likely to be severe in small children.

History

Spiderbite is not always initially painful, and spiders are small and easily misidentified, so most commonly there will be no certainty from the history about the species involved. However, particular spiders cause quite specific envenoming syndromes, making diagnosis possible even without a spider being available. The common presentations for the medically important spiders are listed below. In general, however, it is important to note the circumstances of definite or possible exposure to spiderbite, a description of the spider, if seen, and the timing of onset for any symptoms that develop.

Australian funnel web spiders

These large mygalomorph spiders are robust in appearance (Fig. 22.1.15), generally ground dwelling (there are tree-dwelling species) and are found only in eastern Australia (Fig. 22.1.16). Their large fangs and acidic venom generally cause immediate local pain on biting and they may hang on, being difficult to dislodge. Apart from pain, other local effects are not prominent. In about 10% of cases, systemic envenoming will develop, often rapidly, and can be lethal in less than 60 minutes in children. First symptoms are tingling of the lips and twitching of the tongue, followed by non-specific symptoms, which may include headache,

nausea, vomiting and abdominal pain. There is frequently evidence of neuroexcitation, with sweating, salivation, lacrimation and piloerection. Hypertension is usual and dyspnoea secondary to pulmonary oedema can develop rapidly. Without treatment this can progress to hypoxia, coma and death.

Widow spiders including Australian redback spiders

Widow spiders have a classic appearance (Fig. 22.1.17) with a globular abdomen, comparatively small cephalothorax and long thin legs. They utilise a drop-line web structure to capture prey. Humans most often receive bites when they either come in contact with the web or occasionally when touching a wandering spider (i.e. caught in footwear, clothing or similar). Widow spider venom, though neuroexcitatory, is rarely lethal in humans. A typical significant bite causes local pain, becoming more severe, sometimes with local sweating, then progressive proximal movement of pain and sweating, ultimately becoming severe regional or generalised pain, sweating, plus hypertension, nausea and malaise. Envenoming can mimic acute abdomen and cardiac chest pain. Untreated, pain may take days or weeks to resolve and often gravitates to the lower limbs, causing burning pain in the feet and legs, often with associated sweating. Rarely, severe systemic envenoming can cause pulmonary oedema or a secondary rise in creatine kinase. In infants, a generalised rash is common, the infant presenting as highly distressed and irritable, not consolable, erythematous, but with no obvious cause apparent. A widow spider may be found in the bedding or underneath the bed. Less than 20% of bites cause major envenoming.

Table 22.1.4

Medically important spider groups

Family	Genera	Common name	Distribution	Clinical effects
Hexathelidae	*Atrax* *Hadronyche* *Illawarra*	Australian funnel web spiders	Eastern Australia, from Cape York to Tasmania	Severe neuroexcitatory ('autonomic storm') envenoming; about 10% of cases develop major envenoming, which untreated, carries a significant risk of fatality
Theriidae	*Latrodectus steatoda*	Redback or widow spiders	Global	Moderate, generally non-lethal neuroexcitatory envenoming
Ctenidae	*Phoneutria*	Banana spiders	Central and South America, especially Brazil	Moderate to severe neuroexcitatory envenoming, rarely lethal
Loxoscelidae	*Loxosceles*	Recluse or fiddleback spiders	Global, particularly the Americas	Severe local tissue injury, with occasional major, potentially lethal systemic effects

FIG. 22.1.15 Male Sydney funnel web spider (*Atrax robustus*). Reproduced with permission of Dr Julian White.

Banana spiders

Banana spiders are large aggressive spiders, well known within their range. They are active hunters and invade houses, with bites occurring year round but especially in autumn. They are a common cause of bites in Brazil, accounting for 20% of all presentations in some hospitals. The neuroexcitatory venom has effects similar to widow spider bites in many ways, with pain being a predominant feature, as is sweating, hypertension and nausea. However, unlike widow spiders, there is often local swelling of the bite area and priapism is a classic feature of envenoming in boys. Death is a rare outcome, but severe cases can develop pulmonary oedema or cardiac arrhythmias.

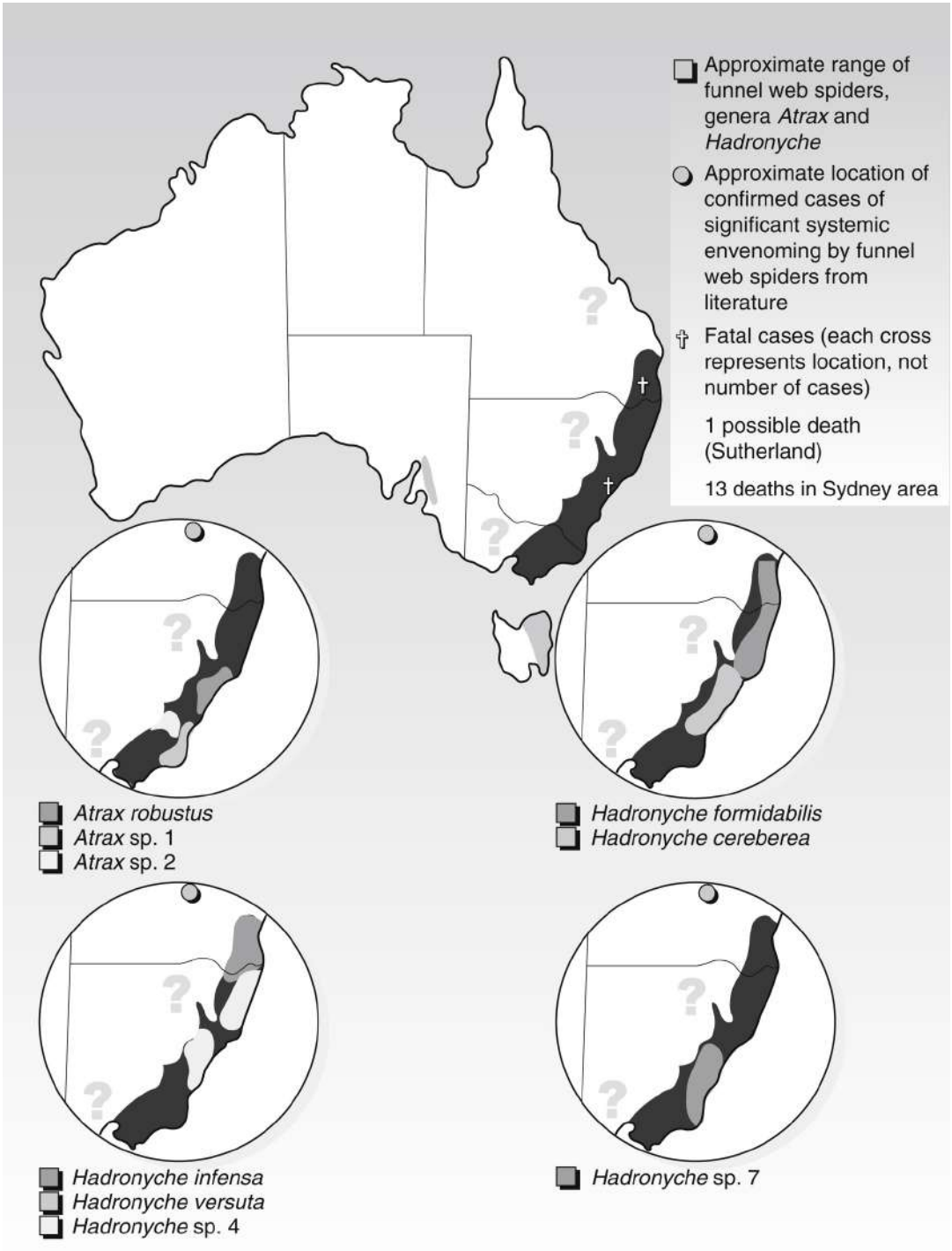

FIG. 22.1.16 Distribution of Australian funnel web spiders (*Atrax* spp. and *Hadronyche* spp.). Reproduced with permission of Dr Julian White.

FIG. 22.1.17 Female widow (redback) spider. Reproduced with permission of Dr Julian White.

FIG. 22.1.18 Recluse spider, with violin-shaped marking on the cephalothorax. Reproduced with permission of Dr Julian White.

Recluse or fiddleback spiders

These small, delicate spiders are generally brown with long spindly legs and a darker brown pattern on the cephalothorax in the rough shape of a violin (Fig. 22.1.18). They have venom that is predominantly dermonecrotic. The most common effect of bites is ‘cutaneous loxoscelism’. The bite is rarely noticed and often occurs in bed at night. However, hours later the area becomes red, painful, may blister or bruise and darken as necrosis develops over the following 4–7 days. The area finally involved can be local to extensive, with skip lesions, and is usually painful. There is commonly an associated non-specific, self-limited, systemic illness. In a few cases, a more severe systemic illness can occur, potentially lethal, with haemolysis, shock, coagulopathy or renal failure. This is ‘viscerocutaneous loxoscelism’.

Examination

Initial examination may show little locally at the bite site, depending on the type of envenoming involved. For those spiders causing predominantly regional or systemic effects, these will dominate examination findings.

Investigations

There are no clinical diagnostic tests specific for spiderbite. Most bites will result in leucocytosis. For suspected loxoscelism, it is important to look for haemolysis, disseminated intravascular coagulation and renal failure, especially in children.

Differential diagnosis

Detailed differential diagnosis is beyond the scope of this chapter, but the geographical location and pattern of local and systemic effects makes differentiation between these types of spiderbites straightforward. For severe neuroexcitatory envenoming, as seen with Australian funnel web spiders, consider pesticide or pharmaceutical poisoning in the differential. Widow spiderbite should be considered in the differential for selected cases of apparent unexplained acute abdomen or chest pain, before laparotomy is scheduled.

Treatment

Treatment for Australian funnel web spiderbite is principally the use of specific IV antivenom, available in Australia only. Initially 2–4 vials are needed, but severe cases require more. All cases with any evidence of venom spread require antivenom urgently, before life-threatening envenoming develops, which may occur rapidly in children.

Widow spiderbite only requires treatment in those cases with significant local, regional or systemic envenoming, which is a minority of cases. Antivenom is the most effective treatment and may be given intramuscularly (IM, in Australia), but is more rapidly effective if used IV (even in Australia). Experience has shown that only antivenom reliably reverses envenoming and is more effective than narcotic analgesics in treating pain. It can be effective for up to days post-bite. It is usually given as single vials (2 vials in Australia), waiting 2+ hours before giving a further dose, if symptoms warrant. Some recent research in Australia has questioned the effectiveness of antivenom, but this is in contrast to

decades of positive experience using this antivenom and there are arguably issues with the aforementioned research which, until confirmed by independent studies, should not be used to change practice away from antivenom.

Banana spiderbite is most often managed without use of antivenom, the latter reserved for severe cases, where it should be given IV, most of these cases being children under 7 years of age. In the less severe cases, local anaesthetic block is usually adequate.

Loxoscelism is difficult to treat. Specific antivenom is only available in Brazil, and its place in management is controversial, though it is widely used in Brazil and considered effective. In general, patients present late, after tissue injury has commenced, requiring good wound care. Secondary infection requires appropriate antibiotic therapy. Early surgical debridement can extend the area of injury and should be avoided. Steroids have not been shown to be effective. Dapsone, given early, can reduce injury but is toxic and not widely favoured as therapy. Hyperbaric oxygen therapy is controversial. It may benefit some patients but is unsuitable for most children.

Prognosis

Prognosis varies depending on the type of spider, but only for Australian funnel web spiders is death a likely outcome unless specific treatment is urgently instituted.

Prevention

Spiders are ubiquitous, and it is not practical to avoid human contact. In areas where the potentially deadly funnel web spiders are common, such as parts of Sydney, residents should avoid walking barefoot, leaving clothes on the ground or putting on footwear without first checking for spiders.

Controversies and Future Directions

1. Major controversies for spiderbite centre around adequate treatment, particularly the role of antivenom, with attendant risks, in envenoming unlikely to prove lethal. This concern has held sway in North America for widow spiderbites, with few cases offered antivenom therapy. As a result countless patients probably suffer prolonged periods of

eminently treatable major discomfort. The opposite situation occurs in Australia where widow (redback) spiderbites routinely receive antivenom, with apparently good results and little risk.

2. Bites by most spiders remain poorly documented and, even for major species, treatment is controversial. For these latter, controlled trials are required to establish best treatment practice.

Tick bite paralysis

Introduction

Tick bite is probably common in some regions but rarely causes major harm to humans. However, a few species of ticks have toxic saliva, containing paralytic neurotoxins, which can cause potentially lethal flaccid paralysis, especially in children. In Australia, for instance, paralysis ticks (*Ixodes* spp.) have caused more deaths than funnel web spiders.

History

There may be a clear history of a tick being found, but often in children presentation is as an unexplained progressive flaccid paralysis, first manifesting as an ataxic gait. Occasionally the paralysis may be purely local, notably a Bell's palsy. Without treatment, the envenoming may cause complete respiratory paralysis. For Australian paralysis ticks only, the paralysis may worsen for up to 48 hours after removal of all ticks. It is important to ascertain if the patient had exposure to ticks, such as walking in scrubland in eastern Australia.

Examination

Examination is crucial, both to document the extent of paralysis and to locate every attached tick. These may be hiding in the scalp, behind or in the ears or in body skinfolds.

Investigations

There are no specific investigations for tick envenoming.

Differential diagnosis

Apart from ticks, at least in Australia, flaccid paralysis can be caused by snakebite. Ataxia only can also be caused by exposure to pesticides or some pharmaceuticals.

Treatment

The principal treatment for tick envenoming is prompt removal of all ticks. Care must be taken to lever the tick off, including mouthparts, and not squeeze it between fingers, which forces in more saliva and often leaves the head embedded, when secondary infection can ensue. The previously available tick antivenom (Australia) is no longer produced. The paralysis resolves after several days, during which time ventilatory support may be needed.

Prognosis

With removal of all ticks and respiratory support, the prognosis should be optimistic. The more rapid the onset of paralytic features, the more likely is major paralysis.

Prevention

If visiting tick-infested areas, it may be difficult to exclude all tick contact risk, so routine checking for ticks after departure and their removal is advisable.

Controversies and Future Directions

The major controversy in the past was the value of tick antivenom. This antivenom is no longer available.

Jellyfish stings

Introduction

Jellyfish are numerous in all seas and oceans and stings, mostly trivial, are common. A few jellyfish can cause more severe stings and an even smaller number can cause potentially lethal envenoming. Of the many species that cause some effects, only the three groups of most medical significance will be

discussed here. All jellyfish have a common mechanism of envenoming, using individual sting organelles (nematocysts) that both produce and inject the venom. This can result in some venom directly entering small blood vessels in the skin, causing rapid envenoming. In the case of large jellyfish, like the box jellyfish, with millions of nematocysts discharging simultaneously through the skin, very rapid and severe envenoming can develop.

Box jellyfish

The Australian box jellyfish, *Chironex fleckeri*, is found in northern Australian marine waters and areas to the north, including Borneo. It is the most dangerous of all jellyfish and can even kill an adult human in less than 5 minutes, from cardiac arrhythmia and arrest. Most stings are minor, not a threat to life, and cause local pain only. In cases with extensive and severe stings, there is immediate excruciating pain, with a ladder mark present, often with adherent tentacles. Collapse may follow rapidly, either due to the pain or to cardiac effects. Respiratory failure can develop, but it is cardiac toxicity that is most likely to prove lethal.

Irukandji syndrome

This is caused by envenoming by a variety of jellyfish, including *Carukia barnesii*, some of which are very small. The initial sting may be trivial and may be from the bell rather than tentacles. However, 20–40 minutes later systemic envenoming develops, with muscle and back pain, often severe, hypertension and malaise and pulmonary oedema, consistent with an ‘autonomic storm’. While death is very rare, it has been recorded in adults as a result of intracranial haemorrhage associated with severe hypertension, though the actual contribution of envenoming is controversial.

Portuguese man-of-war

Blue bottles or Portuguese man-of-war are global oceanic ‘jellyfish’ (actually hydrozoan colony organisms), which swarm and can cause stings from their tentacles. In most cases the stings are minor, with local pain and wheal formation, but rarely more severe envenoming is reported, with a very few cases of vascular injury locally. More common is an allergic reaction to stings, occasionally resulting in lethal anaphylaxis.

History

For most jellyfish there will be a clear history of being stung while in the sea and tentacles may still be adherent. These may still be active, so caution is advisable on removal. For box jellyfish only, the first aid application of copious amounts of vinegar may inactivate the tentacles, so they may be removed safely. The geographical location is important, as is the time of day and season, as this affects the likely local jellyfish fauna.

For stings by Irukandji-type jellyfish, the presentation may be one of unexplained severe pain following a swim in the sea, at any time of the year (but only in northern Australian waters and adjacent areas). It is important to note pre-existing medical conditions and medications that might increase the risk from envenoming.

Examination

The sting area should be examined and the extent determined. For box jellyfish, stings covering half or more of one limb or an equivalent area on the trunk should be considered as potentially lethal. Such a sting area is easily achieved in small children, who are those most often involved in fatal envenoming. Evidence of systemic effects should be sought.

Investigations

There are no specific tests for jellyfish envenoming.

Differential diagnosis

Except for Irukandji syndrome, which can be confused with non-envenoming illness, jellyfish stings are usually easily diagnosed, the major differential diagnosis being other types of marine envenoming, particularly stings from venomous fish, but these usually show only a few spine penetration points, not the widespread tentacle tracks of jellyfish.

Treatment

Most jellyfish stings require either no treatment or simple symptomatic relief of pain. Hot water (45°C, usually as a hot shower) has proved the most effective

first aid for reducing local pain from jellyfish stings, though its applicability to box jellyfish stings remains untested, so for these stings a cold pack is preferred. For box jellyfish stings, where either an extensive area has been stung or systemic envenoming is evident, then box jellyfish antivenom is required urgently, either IM if pre-hospital, or preferably IV in hospital (1–3 vials). In addition full cardiorespiratory support should be instituted, where needed. Local infection of the tentacle tracks can occur, requiring antibiotic therapy. Many cases develop delayed 'allergic' reactions locally, responding to antihistamines and topical corticosteroids.

Irukandji envenoming cannot be treated with antivenom (there is none suitable), so treatment should be supportive and symptomatic. Narcotic analgesia (excluding pethidine) is often required with hypertension most often treated with glyceryl trinitrate (beware hypotension) and pulmonary oedema managed with oxygen, dopamine, adrenaline (epinephrine) and positive pressure ventilation.

Bluebottle envenoming is treated with supportive and symptomatic care only.

Prognosis

For box jellyfish envenoming the larger the area of sting, the more severe the envenoming, with half, or more, of one limb involvement being potentially lethal. For Irukandji, the prognostic indicators are less clear.

Prevention

The only sure way of preventing jellyfish stings is to avoid using the sea. Stinger suits can greatly reduce the chance of contact and will prevent potentially lethal box jellyfish stings, by limiting the area stung, but since only a small contact area is required for Irukandji jellyfish, exposed face, hands or feet may permit major envenoming. Similarly, stinger exclusion nets on beaches will prevent large box jellyfish from entering, so preventing major stings, but do nothing to prevent Irukandji stings.

Controversies and Future Directions

1. There are a number of current controversies for jellyfish sting management. For box jellyfish, while vinegar is widely accepted as first aid to inactivate unfired nematocysts, the use of pressure

immobilisation bandages in severe cases has been questioned, research showing that this technique may actually increase envenoming. As a consequence, most authorities now do not recommend using pressure immobilisation bandages for any jellyfish stings.

2. The role of antivenom for box jellyfish stings remains clouded in uncertainty, but most authorities still recommend its use. Verapamil, once suggested for treating severe cardiotoxic box jellyfish envenoming, is no longer recommended by most authorities and has never undergone clinical testing in this setting.
3. Irukandji envenoming remains problematic. There are still calls for antivenom development, but the delayed nature of envenoming and the increasing and varied range of the species of jellyfish involved make development of an effective antivenom problematic and economically unviable.
4. Research is required into the mechanisms of major jellyfish envenoming in humans, as these are still not fully understood, mirroring the difficulties in collecting and studying jellyfish venom.
5. Research is required to determine most effective treatment strategies for jellyfish envenoming. Irukandji syndrome, in particular, requires more research at all levels, starting with a concerted effort to identify and describe all likely culprit species.
6. The shift from cold pack to a hot shower as first aid for most jellyfish stings, though supported by evidence (unlike cold packs), is still questioned by a few ‘authorities’.

Venomous fish stings

Introduction

There are numerous species of fish, in both marine and freshwater environments, both bony and cartilaginous, capable of inflicting venomous stings, but envenoming, though often distressing, is rarely likely to be lethal, even in children. More concerning, in the case of cartilaginous fish, specifically stingrays, is the potential for major, even lethal, mechanical injury during the act of stinging. Stingray spines on the muscular tail can inflict major trauma, with cases of transection of vessels, nerves, tendons and direct penetration of the

chest and abdomen, including direct cardiac puncture. Such mechanical wounds can pose a great threat to life but are beyond the scope of this chapter, as envenoming is not the significant problem in such cases.

Venomous fish exist in many families, representing hundreds of species, with venomous spines in a number of different locations, depending on species, including on the back (dorsal as in stonefish), pectoral, behind the head (particularly catfish) and even on the tail. A primitive venom gland surrounds the spine and as the spine is forced into the skin by mechanical pressure (such as by stepping on a stonefish or handling a fish) the gland is compressed, forcing venom up grooves in the spine and into the victim.

Most venomous fish species have never had their venom studied. For those few that have been investigated, notably the stonefish, while the venom may contain a variety of components, in the clinical setting it is toxins causing pain and swelling that predominate. There is no evidence that the neurotoxin found in stonefish venom has any clinical effect in humans.

History

There is always a history of definite or likely exposure to a stinging fish, in a marine or freshwater environment, or of handling a fish out of water. This includes sudden pain in a foot after walking in water, such as reef walking, usually indicative of stepping on a fish, notably stonefish. The pain may be very severe, sufficient to cause collapse, but systemic symptoms are related principally to local pain, not general toxicity. There are very rare reports of pulmonary oedema following stonefish stings (in Madagascar). Portions of the sting may have been seen in the wound.

Examination

For venomous fish stings, there may be stings present in the wound(s). The number of wounds can be significant (i.e. for stonefish, as it determines antivenom dose). For stingray wounds, apart from residual sting left in the wound, careful examination to determine the extent of any mechanical trauma is essential.

Investigations

There are no specific investigations for fish sting envenoming.

Differential diagnosis

The sharp and localised pain of a fish sting is distinctive and can be separated from jellyfish stings due to the tentacle tracks caused by the latter.

Treatment

With the exception of stonefish, for which there is an antivenom, fish stings must be treated symptomatically and supportively. Both for first aid and in-hospital care, hot water immersion appears effective at reducing pain in the short term. The contralateral limb should first be immersed in water that is hot, but not so hot that thermal injury might occur (up to 45°C). The affected limb is then immersed, usually bringing rapid relief of pain. Unfortunately, pain may recur on removal from the water and if this persists, other analgesia must be considered, often a local or regional anaesthetic block in more severe cases. The wound must be examined for remnants of stings, which should be removed. The wound should be allowed to heal by secondary intention and the temptation to surgically close the wound resisted. For any wound that is extensive (i.e. many stingray injuries) a course of antibiotics should be considered. For stingray injuries with extensive trauma, surgical input on managing this injury should be paramount. Stingray venom can cause delayed local necrosis, which is particularly concerning with penetrating wounds to the chest or abdomen.

For stonefish, there is a specific antivenom in Australia, which can be given IM or IV, the dose depending on the number of stings. It is effective at reducing the severe pain and should be considered in all cases of stonefish sting with more than trivial symptoms.

Prognosis

With the exception of severe mechanical trauma from stingray injuries, the prognosis for fish stings is generally optimistic, with recovery likely. The greater the number of stings, the more likely symptoms will be severe and possibly prolonged.

Prevention

Avoidance of contact with stinging fish is the obvious preventative measure. When reef walking or on sandy bottoms where stingrays may hide, avoid sudden movements and running into water, wear strong-soled reef shoes and observe and choose carefully when placing feet. Despite all such precautions, stings may occur, particularly if reef walking, by stepping on larger stonefish.

Controversies and Future Directions

1. There are no major controversies for fish stings. There is uncertainty if stonefish antivenom is useful for stings by other types of fish. There is no formal study to validate such non-specific use, but anecdotal clinical experience suggests that this antivenom may work for some related species, including the bullrout.
2. There is great scope for studies on fish-sting venoms and for research into more targeted treatment strategies.

Venomous marine molluscs

Introduction

There are two groups of marine molluscs (snails) that are able to inflict major, even lethal, envenoming on humans. These are the blue-ringed octopus, from Australian and adjacent waters, and selected cone snails, found widely in the Indo-Pacific area. Neither is a common cause of envenoming, surprisingly, since both groups are common – the blue-ringed octopus particularly, with its various species common all around the Australian coast.

The blue-ringed octopus has a potent paralysing neurotoxin, tetrodotoxin, in its saliva. Bites, which usually occur when the octopus is removed from the water and placed in contact with skin, are often painless and may go unnoticed until 5–20 minutes or so later, when paralytic features start to develop. These may rapidly progress to respiratory paralysis and collapse, with death possible in under an hour. The paralysis is a general flaccid paralysis.

Cone snails fire poisoned ‘darts’ at their prey, or human victims, the venom being both complex and incredibly potent, immobilising fish prey in a few seconds. In humans the sting may be painless or quite painful. Systemic envenoming, with collapse and flaccid paralysis, rapidly follows.

History

There is usually a clear history of picking up and handling a blue ringed octopus or cone snail. In significant cases, which are rare, envenoming is rapid, with neurotoxic symptoms quickly evident. The earliest feature of envenoming is often tingling of the lips.

Examination

The sting or bite site may not be readily evident. The key effects to exclude are those of progressive flaccid paralysis and cardiovascular collapse, notably hypotension.

Investigations

There are no investigations specific to envenoming by molluscs.

Differential diagnosis

The onset of progressive flaccid paralysis after a marine sting or bite is uncommon and apart from mollusc envenoming, the likely differential is sea snake bite, which may manifest as either paralysis and/or myolysis. However, the development of paralysis is much less acute than with mollusc envenoming.

Treatment

There are no antivenoms for venomous molluscs, so both first aid and treatment are supportive and symptomatic. Pressure immobilisation bandaging may reduce the rate of development of systemic envenoming, if applied early enough. The key requirement is to support respiratory function and blood pressure. The latter may require pressor therapy. The former may require intubation and mechanical ventilation, but this may not be for a prolonged period, often only for 6–12 hours, unlike snakebite, where ventilation may be needed for days, weeks or even months.

Prognosis

If respiratory support is instituted early, before irreversible hypoxic organ injury

occurs, ultimate prognosis is good. Cases without evidence of significant systemic envenoming 6 hours post exposure are unlikely to develop envenoming.

Prevention

The principal preventative measure is abstinence from contact with, and certainly handling, these molluscs. This may require local education programmes, particularly directed at children, who will find these small and attractive animals tempting to pick up.

Controversies

There are no major controversies in managing marine mollusc envenoming, in part reflecting the rarity of this clinical problem.

Further reading

While there are numerous papers on various aspects of envenoming, covering different regions, there are few texts giving a detailed overview of snakebite or other types of specific envenoming or clinical toxinology. Some recent key texts are noted here. A major source of information is the Clinical Toxinology Resources Website (www.toxinology.com), a detailed site, initially developed with public funds, global in scope.

Chippaux J.-P. *Venins de Serpent et Envenimations*. Paris: IRD editions; 2002.

French handbook on global snakebite.

Covacevich J, Davie P, Pearn J. *Toxic Plants and Animals: a guide for Australia*. Brisbane: Queensland Museum; 1987.

A comprehensive textbook with a clinical focus.

Gopalakrishnakone P, Chou L.M. *Snakes of Medical Importance (Asia-Pacific Region)*. Singapore: National University of Singapore; 1990.

Covers snakes in the designate region.

Goyffon M, Heurtault J. *La Fonction Venimeuse*. Paris: Masson; 1995.

French handbook covering many aspects of toxinology.

Junghanss J, Bodio M. *Notfall-Handbuch Gifttiere: Diagnose, Therapie, Biologie*. Stuttgart: Georg Thieme Verlag; 1996.

This German textbook covers a global spectrum of envenoming with a focus on management in emergency departments.

Mebs D. *Venomous and Poisonous Animals*. Boca Raton, FL: Medpharm/CRC Press; 2002.

This textbook, beautifully illustrated in colour, covers the broad scope of venoms, the animals that produce them and their clinical effects and treatment – the focus is more on toxinology than detailed medical advice, but it is a valuable source of information.

Meier J, White J, eds. *Handbook of Clinical Toxinology of Animal Venoms and Poisons*. Boca Raton,

FL: CRC Press; 1995.

This is the standard textbook for clinical toxinology and contains chapters covering all aspects of envenoming, especially snakebite, scorpion stings, spiderbite, tick envenoming and marine envenoming – the coverage is global, but with considerable detail for most regions and animal types.

Sutherland S.K, Tibballs J. *Australian Animal Toxins*. Melbourne: Oxford University Press; 2001.

A major work covering the Australian venomous fauna.

Warrell D.A. WHO/SEARO guidelines for the clinical management of snake bites in the Southeast Asian region. *Southeast Asian J Trop Med Public Health*. 1999;30(suppl 1):1–85.

Weatherall D.J, Ledingham J.G.G, Warrell D.A. *Oxford Textbook of Medicine*. Oxford: Oxford University Press; 1996.

This standard medical text includes an extensive chapter on envenoming.

White J. .C.S.L. *Antivenom Handbook*. Melbourne: CSL; 2002.

A concise handbook on the diagnosis and treatment of envenoming by Australian fauna, which is available in its entirety on the toxinology website: www.toxinology.com.

White J. .A. *Clinician's Guide to Venomous Bites and Stings*. Melbourne: CSL; 2013.

This is the hugely revised and expanded version of the previous CSL Antivenom Handbook and covers all important Australian venomous animals.

Williamson J.A, Fenner P.J, Burnett J.W, Rifkin J.F. *Venomous and Poisonous Marine Animals: a Medical and Biological Handbook*. Sydney: University of NSW Press; 1996.

This is the major textbook covering marine toxinology.

22.2

Drowning

Simon Vincent Wood

ESSENTIALS

1 Drowning is defined as the process of experiencing respiratory impairment from immersion in liquid, regardless of the outcome.

2 Drowning is a leading cause of accidental death in Australian children. Children aged 0–4 years are the most vulnerable. The most common site of drowning in this age group is the domestic swimming pool.

3 There is no clinical or therapeutic difference between drowning in fresh or salt water.

4 The major pathophysiological consequence of drowning is hypoxic brain injury.

5 Pulmonary injury due to aspiration of water is an important clinical consideration in all drowning victims.

6 The mainstay of treatment is early effective oxygenation.

7 Whilst hypothermia may be protective in small children who drown, it does not reliably predict good outcome.

8 Response to resuscitation is the single most important predictor of outcome in children who have drowned.

9 Isolation pool-fencing with a self-locking gate has been shown to effectively reduce drowning incidents in pre-school-age children.

Introduction

Definition

Traditionally *drowning* has been defined as death due to suffocation within 24 hours of submersion in a liquid medium and *near-drowning* as survival for 24 hours or more following such an incident.[1] Considerable confusion has surrounded the use of these terms. In part this is because the distinction between drowning and near-drowning often cannot be made before 24 hours, making the terms clinically irrelevant. In addition it has been suggested that the use of a time limit for survival is not a scientific concept and is not in accordance with outcome parameters as used in the internationally accepted Utstein style.[1]

To address this issue the Utstein Taskforce on Drowning was convened in Amsterdam as part of the 2002 World Congress on Drowning. The International Liaison Committee on Resuscitation (ILCOR) has since endorsed a review of the terminology and defines drowning in the following way:

> *Drowning. Drowning is a process resulting in primary respiratory impairment from submersion/immersion in a liquid medium. Implicit in this definition is that a liquid/air interface is present at the entrance of the victim's airway, preventing the victim from breathing air. The victim may live or die after this process, but whatever the outcome, he or she has been involved in a drowning incident.*[2]

The term *submersion* is generally accepted to indicate an incident in which the victim's body is totally covered by water, while the term *immersion* refers to an incident in which the victim is only partially covered by water, although for drowning to occur the face and airway must at least be covered.[2]

ILCOR recommends that other terms such as *dry drowning* versus *wet drowning*, *active* versus *passive* versus *silent drowning*, *secondary drowning* and *near drowning* be abandoned.[2]

Epidemiology

In Australia drowning remains a leading cause of accidental death in children. Its incidence peaks in early childhood and again in adolescence. Males outnumber females in both groups. Children under 5 years of age are the most vulnerable to drowning in Australia.[3] In the period between 1 July 2015 and 30 June 2016 there were 280 drowning deaths in Australian waterways. Children aged 14 years and under accounted for 11% of deaths overall with 66% occurring in

children aged 0–4 years. Fifty-two per cent of drowning deaths in children aged 0–4 years occurred in swimming pools. In this age group 81% of drowning deaths resulted from a fall into water, while a further 14% drowned while bathing.[4]

Risk groups for childhood drowning are children aged 0–4 years, children living in cities with high swimming pool to population ratios, children living in hot climates, children living in areas with lack of isolation pool fencing, and indigenous children.[5] More toddlers drown in swimming pools than from any other cause.[6] Most children who drown in pools are out of sight for less than 5 minutes and are in the care of one or both parents.[7] Around the home small children can also drown in baths, buckets and garden ponds. Up to 8% of cases of drowning in small children in the domestic setting may be secondary to non-accidental injury.[8]

While pool-fencing legislation has proven to be effective in reducing the incidence of drowning in small children it has had little impact on rates of drowning in older children and adolescents. In this group alcohol, suicide, and risk-taking behaviours are important factors that lead to increased risk of drowning.[8]

Aetiology

Drowning is most commonly a primary event. In children it most often occurs when the victim is unable to rescue him or herself after entering the water, as in the case of a toddler falling into a swimming pool, or an infant drowning in a bath whilst unattended. In older children and adolescents fatigue while swimming may play a role, but drowning in these age groups is more likely to be secondary to other causes.

Drowning can occur secondarily to a number of underlying causes. These should be considered during assessment of the submersion victim. Individuals with seizure disorders have up to 19-times higher risk for drowning accidents, regardless of age.[3,8,9] Prolonged QT-syndrome leading to dysrhythmia has been implicated as a significant cause of drowning, although the true incidence of this condition in drowned children is unknown. Ethanol is an important risk factor for drowning injury, particularly in adolescents. Elevated serum ethanol levels are documented in 10–50% of adolescent drownings.[8] Head and cervical trauma from diving and boating-related accidents may also lead to drowning as a secondary event. Non-accidental injury is an important cause of drowning in

infants and smaller children, particularly in events that occur in the home, such as in baths and buckets. Up to 8% of drownings presenting to tertiary paediatric centres may be attributed to child abuse.[8]

The immersion syndrome is sudden loss of consciousness secondary to a bradycardia, or tachyarrhythmia induced by contact with water at a temperature of at least 5°C below body temperature. This can lead secondarily to drowning. The immersion syndrome can occur in water with temperatures as warm as 31°C, although it is more likely to occur in much colder water. Wetting the face before entering the water may reduce its incidence.[1]

Pathophysiology

The two most significant pathophysiological consequences of submersion are hypoxia from asphyxiation during the submersion itself, and aspiration of water into the lungs. It is the severity of the initial hypoxic insult that is the major determinant of outcome. If the initial hypoxic event is survived, the degree of hypoxic organ injury and pulmonary injury secondary to aspiration become the clinically important factors.

Much that is known about the sequence of events following submersion has come from animal models. Aspiration of water initially causes breath-holding or laryngospasm and the resultant asphyxiation leads to progressive hypoxia. Active and passive swallowing of water follows and, as hypoxia worsens, breath-holding and laryngospasm are terminated, resulting in aspiration of water into the lungs.[1]

Anoxia lasting 1–3 minutes can shut down both the brain and the heart, causing loss of consciousness and hypoxic cardiac arrest. Rescue and early institution of cardiopulmonary resuscitation can salvage myocardial function, but the brain is more sensitive to hypoxic injury and it is the severity of this injury that determines outcome. Effects of hypoxia on other organ systems are delayed. Profound hypoxia can cause an acute respiratory distress syndrome, which develops within hours and further worsens hypoxic injury. Post-hypoxic cerebral oedema is a major complication and can develop 6–12 hours following successful initial resuscitation from a serious submersion event. Most paediatric drowning deaths in hospital are due to hypoxic cerebral injury rather than pulmonary complications.[8]

The average volume of water aspirated in human drownings is 10–15 mL kg^{-1}. Aspiration of volumes as little as 1–3 mL kg^{-1} of water can cause profound

alterations in gas exchange and subsequent ventilatory abnormalities.[10] Laryngospasm is thought to occur in 10–15% of drowning victims, and a subset of patients who drown without evidence of significant aspiration of water at post-mortem, so-called *dry-drowning*, has been described. This concept has recently been questioned, and it has been suggested that in these cases death may have occurred prior to submersion.[1] Regardless of whether dry-drowning is a true clinical entity, or whether laryngospasm has occurred at the time of submersion, aspiration of water into the lungs remains a clinically important consideration in the management of all drowning victims.

Despite the large literature dedicated to the subject, there are no clinically or therapeutically important differences between drowning in fresh or salt water.[1,8] Pulmonary injury is related more to the amount of water aspirated than to the composition of the water itself. Both fresh and salt water cause loss of pulmonary surfactant, non-cardiogenic pulmonary oedema, impaired alveolar-capillary gas exchange, and increased intrapulmonary shunting with the potential for profound hypoxia.[1] Aspiration of water that is contaminated with particulate matter or bacteria can lead to complications from obstruction of small airways or increased risk of pulmonary infection, although neither is seen in the majority of patients.[11] If present, evidence of significant pulmonary injury due to aspiration will usually manifest or progress within hours of rescue. Delayed onset of respiratory distress and hypoxia, the so-called ‘delayed immersion syndrome’ or ‘secondary drowning’ has been refuted by recent evidence.[8]

Clinically significant electrolyte and fluid volume abnormalities are rarely seen in cases of drowning in humans despite being demonstrated in animal models.[1,3,8,11] Occasionally a mild hyponatraemia, which self-corrects without specific therapy, is observed.[8] Theoretical exceptions are drownings occurring in hypertonic solutions, such as the Dead Sea, or water contaminated with industrial waste.

Hypothermia is an important issue following drowning, particularly in small children who have a large body surface area to weight ratio. Cooling can occur at the time of submersion, but can also continue following rescue and during attempted resuscitation due to heat loss through evaporation. Hypothermia can confer some degree of protection from cerebral hypoxia, particularly in small children. Multiple case reports in the literature attest to intact survival of both children and adults following prolonged (>15 minutes) drownings in icy water (water temperature <10°C).[12] Profound hypothermia and subsequent intact survival have also been documented in children suffering drowning in non-icy

water and in temperate climates.

The mechanisms of temperature drop and cerebral protection remain unclear. Surface cooling at the time of submersion is thought to be insufficient on its own to provide central cooling of a degree that confers cerebral protection. Other mechanisms of heat loss, such as via ingestion and/or aspiration of cold water, are not supported by quantitative evidence. Some authors suggest that core temperature drop is insufficient on its own to explain the cerebral protection afforded by hypothermia.[13] The diving reflex, in which blood is shunted from the limbs and splanchnic circulation to the brain and heart alongside slowing of the heart rate and reduction of the basal metabolic rate, has been suggested as being an important mechanism for cerebral protection in children.[13] There is little clinical evidence to indicate that the diving reflex is sufficiently active in humans, even small children, to confer any benefit on its own.[1,8] It is most likely that a combination of the effects of the diving reflex initially, followed by rapid and continued cooling, is what underlies the cerebral preservation that is sometimes seen in small children who suffer submersion and who are profoundly hypothermic.

By whatever mechanism cooling occurs, and whether the diving reflex plays a significant role in cerebral protection or not, hypothermia in the drowning victim, particularly if the victim is an infant or young child, should be considered to be an indication for aggressive and prolonged resuscitation efforts. This issue is discussed further in Chapter 22.4 in the section on cold injuries.

History

Key points in history are summarised in Box 22.2.1. Broad areas in history include details of the drowning event itself, details of rescue and resuscitation, response to resuscitative efforts, possible underlying causal factors and medical conditions that may influence recovery. Features of history that alert to the possibility of non-accidental injury should be recognised. These include a history that is inconsistent or a history that is incompatible with the victim's developmental level.

Box 22.2.1 Key points in history

The circumstances leading to the submersion
The location of the submersion

Water and environmental temperatures
Duration of submersion
Time from when the victim was last seen to the time when found (if the submersion was unwitnessed)
Time from rescue to effective cardiopulmonary resuscitation
Time to first gasp or return of spontaneous circulation
Elements of basic and advanced life support employed
Symptomatology following successful resuscitation
Medical history particularly with regard to possible predisposing factors (i.e. neurological disorders, such as epilepsy)
Factors that may complicate recovery (i.e. respiratory illness, such as asthma)

Examination

Examination is dictated by the clinical condition of the victim on arrival in the emergency department (ED) and is mainly directed at assessing the degree of neurological impairment due to hypoxic cerebral injury, the severity of respiratory embarrassment due to aspiration, and cardiovascular instability due to a combination of the initial insult and/or ongoing hypoxia. In broad terms drowning victims arriving in the ED will fall into two categories: (1) those who respond to minimal resuscitation and who will generally do well with minimal complications; and (2) those 'high risk' patients who fail to respond to resuscitation and who will require ongoing resuscitation and/or monitoring.[8]

Vital signs, including oxygen saturation, a bedside blood glucose estimate, and a temperature should be recorded on all patients. It is important to detect hypothermia, particularly in patients who have failed to respond to resuscitation efforts. Small children may rapidly become hypothermic due to evaporative heat loss during resuscitation efforts and transfer to the hospital. Apparent lifelessness and severe bradycardia due to profound hypothermia need to be distinguished from asystole and brain death. Assessment of cardiac rhythm requires observation of the continuous electrocardiograph (ECG) monitor for up to a minute to detect very slow heart rates that can occur in profound hypothermia. Similarly, lack of response to painful stimulus, along with fixed and dilated pupils should not be interpreted as brain death in the presence of profound hypothermia.

Depending on the clinical condition of the victim at arrival, neurological evaluation may range from an assessment of level of responsiveness as graded by the AVPU or Glasgow coma score (GCS) and pupillary reaction, to a focused neurological examination looking for focal deficit or spinal-cord injury. An assessment should always be made for the possibility of cranial or cervical spine trauma, particularly in diving-related accidents. Trauma to other areas of the body should also be sought.

In the awake child, examination of the respiratory system can establish a clinical baseline against which subsequent deterioration can be measured. Work of breathing should be assessed, and abnormalities found on auscultation, such as crackles and wheezes, should be noted. The finding of signs on an initial examination indicates the possibility of significant aspiration and dictates close observation to identify deterioration. A clear chest initially does not exclude aspiration and a period of observation and re-examination is necessary in all children who have been symptomatic following drowning.

Investigations

Investigations will also be dictated by the clinical condition of the patient on arrival in the ED. A child who has suffered a drowning injury and who is alert and asymptomatic on arrival requires little in the way of laboratory or radiological evaluation. The more symptomatic or seriously unwell child may benefit from further evaluation with laboratory and radiological investigations.

Useful investigations include arterial blood gas (ABG) analysis, serum glucose and chest radiography. ABG analysis may demonstrate a metabolic acidosis, which confirms a significant drowning injury.[8] The severity of the acidosis reflects the severity of the hypoxic insult as well as ongoing hypoxic injury. Profound acidosis (pH <7.10) implies a poorer prognosis but needs to be interpreted in the clinical context.[14] ABG analysis can be used to guide decisions regarding oxygenation and ventilation, and serial determinations may be useful in monitoring and quantifying deterioration of pulmonary function due to non-cardiogenic pulmonary oedema secondary to hypoxia or aspiration. Determination of the blood glucose level is important in any critically ill child as hypoglycaemia can complicate physiological stress and should be actively treated. Hyperglycaemia in a comatose child, although not requiring treatment, implies a poor prognosis.[8] Initially the chest X-ray (CXR) may be normal, may demonstrate pulmonary infiltrates, or may display frank pulmonary oedema.

Abnormalities detected on an early CXR mandate close observation, and the patient should be monitored for clinical deterioration. Repeated CXR may be required but should be dictated by the patient's clinical condition.

Other investigations that may be helpful include baseline electrolytes and full blood count, although clinically or therapeutically significant abnormalities are rarely found on initial determinations. Blood ethanol levels may be relevant, depending on the age of the patient. A 12-lead ECG may be helpful in excluding prolonged QT syndrome as a cause for the drowning event.[10] Cervical spine films should be considered if cervical injury is suspected or the drowning is secondary to a diving accident.

Differential diagnosis

The major issues in differential diagnosis relate to the cause of the drowning event. Underlying medical conditions, such as epilepsy, should be considered. Trauma, either leading to or as a consequence of the drowning, should be recognised. Non-accidental injury should be suspected when there are inconsistencies in the history.

Severe hypothermia (core temperature less than 29°C) can mimic irretrievable cardiorespiratory arrest and brain death. The profound bradycardia associated with very low core body temperature can easily be confused with brady-asystole secondary to hypoxic injury and will not respond to usual resuscitation measures until hypothermia is corrected. Similarly, severe hypothermia can cause depression of cerebral function leading to unresponsiveness and fixed, dilated pupils, indistinguishable from irreversible hypoxic cerebral injury.[15] Failure of the core temperature to rise despite aggressive, active rewarming may be the only indication that death has occurred.

Treatment

Treatment of the drowning victim occurs in three major phases: (1) rescue and resuscitation at the scene; (2) initial assessment and stabilisation in the ED; and (3) subsequent observation and supportive care in the hospital ward, intensive care unit (ICU), or after discharge. Early institution of effective cardiopulmonary resuscitation (CPR) with an emphasis on providing adequate ventilation is the key task in pre-hospital management. Compression-only CPR is not the recommended resuscitation method as the primary cause of cardiac arrest in

drowning is lack of breathing.[16] Manoeuvres to drain the lungs of water have not been shown to be clinically effective and may increase the risk of aspiration of gastric contents. Emesis is common in drowning victims, both spontaneously and as a complication of resuscitation, and aspiration of gastric contents is a major potential complication following rescue. Spontaneously breathing patients should be managed and transported in the right lateral decubitus position. Cricoid pressure may reduce the risk of gastric distension and aspiration during cardiopulmonary resuscitation but requires an additional rescuer.[1]

Spinal injury with drowning is rare and is estimated to be present in less than 0.5% of cases. Cervical spine immobilisation is only indicated in cases in which head or neck injury is strongly suspected, such as accidents which involve diving into shallow water, dumping surf or watercraft.[16,17]

Hypothermia can be exacerbated by ongoing evaporative heat loss during resuscitation efforts following rescue. Wet clothing should be removed, and the patient should be dried if possible. Exposure during CPR should be minimised as much as is practicable. If hypothermia is severe at the scene (<30°C), rewarming should probably be delayed until adequate ventilation and oxygenation has been instituted.[15] Invasive rewarming techniques should be reserved for the hospital phase of management.

Treatment in the ED involves provision of adequate oxygenation, stabilisation of the body temperature, prevention of complications such as aspiration, and assessment of the patient's clinical status in order to make decisions regarding ongoing management and disposition.

Supplemental oxygen should be administered at concentrations and via delivery systems appropriate to the patient's oxygen requirement. Continuous pulse oximetry and serial estimations of respiratory rate and work of breathing should be undertaken during observation in the ED to determine worsening or improvement of the patient's respiratory status. If available, non-invasive ventilation techniques, although not extensively evaluated in paediatric drowning victims, may be considered if the oxygen requirement outstrips conventional mechanisms of oxygen delivery. Intubation to isolate the airway and provide ventilation with positive end-expiratory pressure (PEEP) may be required in patients with depressed neurological status or if respiratory compromise progresses despite supplemental oxygen therapy. Failure to maintain an SaO_2 of greater than 90% with an FiO_2 of 0.50 or higher, a $PaCO_2$ of more than 35 mmHg, an abnormally high respiratory rate (>50 bpm) or inadequate spontaneous ventilation, are all indications that mechanical

ventilation is likely to be required.[1,10] Passage of a nasogastric or orogastric tube, if cranial trauma is suspected, should be performed following intubation to decompress the stomach and facilitate mechanical ventilation.

Diuretics are not recommended in the management of non-cardiogenic pulmonary oedema.[1,8,10] Steroids have not been shown to be useful in the management of aspiration pneumonitis, and the role of antibiotics in this setting remains controversial.[1,8,10] Prophylactic antibiotics are not recommended, although they are sometimes used when there is a history of drowning in heavily contaminated or polluted water. In general, antibiotics should be reserved for patients who develop signs of pulmonary infection, such as fever, sustained leucocytosis, or persistent or new pulmonary infiltrates.[1,17] Aspiration of swimming pool water rarely results in pneumonia.[17] Fluid therapy should be judicious and should be aimed at maintaining adequate circulatory status and euglycaemia without overloading the patient.

Hypothermic patients should be warmed once adequate ventilation and oxygenation has been assured. Awake and spontaneously breathing patients with mild to moderate hypothermia (core temperature >32°C) require passive or external active rewarming techniques only. Patients with severe hypothermia (core temperature <32°C), who are obtunded or in cardiorespiratory arrest, will require invasive, active, rewarming techniques. These are discussed fully elsewhere in this text (see Chapter 22.4 in the section on cold injuries). There is increasing evidence that therapeutic hypothermia in patients with return of spontaneous circulation (ROSC) following out of hospital cardiac arrest is associated with a survival and neuroprotective benefit, although there is little evidence to support one specific temperature target over another.[18] Much of the evidence for therapeutic hypothermia after ROSC, however, is derived from adult populations. Current (January 2016) Australian and New Zealand Committee on Resuscitation (ANZCOR) Guidelines for infants and children with ROSC following cardiac arrest recommend that it is acceptable to target normothermia (36–37.5°C) or hypothermia (32–34°C) and that hyperthermia should be avoided in the post ROSC period.[19]

In the setting of cardiorespiratory arrest and severe hypothermia, resuscitation efforts should be continued until the core temperature has risen to around 32°C before a decision to cease resuscitation is made. Although there are no clear guidelines to determine how long efforts at rewarming should be continued, failure to effect rewarming despite maximal invasive efforts may be an indication that continuing resuscitation is futile. The decision to cease

resuscitative efforts in the presence of persistent severe hypothermia should follow a multidisciplinary approach involving the emergency physician, paediatrician and/or paediatric intensivist and other members of the resuscitation team, together with the parents and family. Cardiorespiratory arrest on arrival in the ED in a patient with a core temperature above 32°C carries a uniformly poor prognosis and prolonged efforts at resuscitation are generally not indicated.

Treatment in the ICU involves provision of ventilatory support to ensure adequate oxygenation, maintenance of cardiovascular stability, minimisation of secondary brain injury due to cerebral oedema, management of hypoxic organ injury, and management of the complications of pulmonary aspiration. As the clinical scenarios are similar, the ventilatory strategies used for the management of acute respiratory distress syndrome (ARDS) should be followed for patients who have drowned.[17] Extracorporeal membrane oxygenation (ECMO), if available, may be considered in patients whose pulmonary function has deteriorated such that adequate oxygenation cannot otherwise be maintained.[17] The roles of interventions such as the use of artificial surfactant, inhaled nitric oxide, and partial liquid ventilation with perfluorocarbons are yet to be established.[17] Various measures to provide cerebral resuscitation and control intracranial pressure have been evaluated in the literature and have generally been found to be not helpful in influencing outcome.[1] In general, the aim should be to maintain adequate cerebral oxygenation and to minimise causes of secondary cerebral injury such as hypotension, hypercapnia, hyperthermia and hypo/hyperglycaemia.

Disposition

Disposition is dictated by the clinical condition of the patient in the ED, the nature of the drowning event, the need for resuscitation and the presence or absence of other factors, such as trauma, suspicion of non-accidental injury and underlying or complicating medical conditions. In general, all victims of drowning will require some period of observation.

The alert, otherwise healthy, patient who is asymptomatic or who has suffered only mild, transient symptoms following a brief drowning can be safely discharged if he/she remains well after 6–8 hours of observation.[8] Patients with a history of drowning for longer than 1 minute, a period of cyanosis or apnoea, or who required pulmonary resuscitation, should be admitted for observation for 24 hours, or at least overnight, even if they are well in the ED. Although recent

evidence discredits the idea of the ‘delayed immersion syndrome’, cases of fulminant pulmonary oedema up to 12 hours after drowning have been reported as occurring in patients who appear well and display normal chest radiography in the ED.[1] Any child discharged from the ED following a drowning event should be in the care of a reliable and responsible adult. Instructions should be given to re-present for further medical assessment in the event of any change in the child’s respiratory status.

Patients who have suffered trauma may require admission for the management of their injuries. Children with underlying cardiac or respiratory disorders, those who suffer drowning secondary to a pre-existing illness and victims of suspected non-accidental injury also warrant inpatient evaluation.

Asymptomatic children and children with mild symptoms who are admitted for observation can be managed in a general ward environment, provided that there is the facility to increase the level of monitoring and care should it be required. Children who have required CPR, who have abnormal chest radiography or ABGs on arrival in the ED, or who have required ventilatory support, should be admitted to a high-dependency unit or ICU for observation and management.[1] Transfer to a facility that provides paediatric ICU should be considered for all such patients prior to clinical deterioration.

Prognosis

Most children who suffer drowning injury will either survive intact or die. Death occurs in 30–50% of drowning victims, most of these not surviving to treatment in the ED. A small proportion of victims will be left with severe neurological deficit, either persistent vegetative state or spastic quadriplegia. Mild learning deficits may occur in apparently intact survivors, although the extent of these and the impact on subsequent function have not been clearly quantified.[8] In general, however, the prognosis of children who suffer drowning events and survive to hospital admission is excellent.[20]

Despite a large body of literature dedicated to the subject, the factors that predict prognosis in the ED following childhood drowning injury remain poorly defined. There are no prospectively validated scoring systems and most of the literature is based on retrospective data. Individual case reports of children surviving prolonged resuscitative efforts with good outcome continue to arise in the popular press and medical literature.

Factors elicited in history that influence prognosis are duration of submersion,

time to institution of effective CPR, and time to first spontaneous gasp, all three reflecting the duration of the hypoxic cerebral insult. Duration of submersion can often be difficult to determine, but submersion for longer than 5 minutes is associated with a poorer prognosis.[14] Although time to institution of effective CPR can be similarly difficult to ascertain accurately, delays of longer than 10–20 minutes are also associated with poorer outcomes, while early effective CPR has been shown to be an important factor in improving survival after rescue.[14]

As already discussed, hypothermia may be protective in small children who suffer drowning, particularly, but not exclusively, if the drowning has occurred in cold or icy water. However, neither hypothermia nor water temperature can be reliably used to predict outcome.[21,22]

Response to resuscitation following rescue and prior to arrival in the ED is the single most important indicator of outcome in children who have suffered drowning. Children who arrive conscious in the ED after successful resuscitation have almost universally excellent outcomes. Similarly, lack of response to early resuscitation efforts and coma on arrival in the ED are associated with a poor outcome.[10] In a classification system based on the neurological status of the patient on arrival in the ED, it was discovered that less than 15% of patients who were unresponsive and flaccid to painful stimulation had intact survival. This is supported by the findings of other investigators that a GCS of five or less on arrival in the ED is associated with a dismal outcome, either death or severe neurological disability.[1,23] Lack of pupillary response in the ED has similarly been identified as a predictor of dismal outcome.[24] The caveat concerning the use of level of responsiveness and pupillary reaction to predict poor prognosis and withdraw treatment applies to the child who presents with severe hypothermia, which may mimic brain death. In these patients lack of response to aggressive rewarming and resuscitation becomes a surrogate indicator of poor outcome. In non-hypothermic patients who are in cardiorespiratory arrest, a lack of response to 25 minutes of effective advanced-life-support measures is almost universally associated with poor outcome.[8] It should be noted, however, that studies of outcome following out-of-hospital cardiac arrest in children indicate that children who suffer cardiac arrest secondary to drowning have significantly better outcomes than those who suffer cardiac arrest from other causes.

Prevention

Drowning remains a leading cause of accidental death in toddlers and

adolescents in Australia. Most drowning-prevention strategies are aimed at the small child who drowns after falling into the domestic swimming pool, with few aimed at older children and adolescents.

The Australian Water Safety Council (AWSC) has set out a strategy to reduce drowning deaths by 50% by 2020. This strategy follows a life-stage approach. Key objectives vary between target age-groups. For children aged 0–14 years evidence-based approaches for the prevention of drowning in children include: promotion of adult supervision, legislation for and compliance with four-sided pool fencing, promotion of community-wide rescue and CPR skills, and age appropriate water familiarisation and survival swimming skills.[25,26] For adolescents and young adults key focuses relate primarily to reduction of risk-taking behaviour and the use of alcohol and other drugs.[25] Lack of adequate adult supervision is recognised as a significant contributor to child drowning deaths.[26] The Royal Life Saving Society recommends levels of supervision for different age groups: children aged 0–5 years and all non-swimmers should receive constant active supervision within arm's reach, children aged 6–10 years may receive constant active supervision from a distance, and for children aged 11 years and above regular checks by physically looking is recommended.[26]

Isolation or four-sided fencing with a self-locking or dynamic gate has been shown to effectively reduce drowning incidents by up to 50%, in both Australia and New Zealand, and in the United States.[8] Pool alarms, pool covers and fencing that does not isolate the pool from the home are probably not effective, although evidence is lacking. Pool-fencing legislation exists in Australia but is not uniform across states. Compliance with legislation remains incomplete.[25]

Controversies and Future Directions

1. The role of therapeutic hypothermia following cardiac arrest due to drowning in children continues to be based primarily on data from adult populations.
2. The role and indications for advanced or novel therapies, such as ECMO, the use of artificial surfactant, inhaled nitric oxide and partial liquid ventilation with perfluorocarbons, are yet to be established.
3. Prospective collection of data on drowning victims is needed to generate more useful information regarding prognosis. Scoring systems for prognosis should also be prospectively validated.
4. Further research regarding preventative strategies and their effectiveness

is required. Areas that require specific attention include programmes aimed at older children and adolescents.

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22.3

Heat-induced illness

Nicholas Cheng, and Ioannis Pegiazoglou

ESSENTIALS

1 Most heat-induced illness in children is due to overheating from exogenous sources.

2 Children are particularly susceptible to heat-related illness due to their physiology.

3 Certain childhood genetic and dermatological disorders dramatically increase the risk of heat-related illness.

4 Genuine heat stroke is a true medical emergency. It is characterised by a core body temperature above 40°C in association with acute mental state changes.

5 Immediate aggressive cooling methods should be instituted for heat stroke.

6 Children account for almost one-fifth of all cases of malignant hyperthermia. Dantrolene should be given early for suspected cases. Approximately 50% of cases have a gene defect of the ryanodine receptor.

Introduction

In children, heat-related illness is a common presentation to the emergency department (ED). Fortunately, deaths are rare. Mortality figures from the USA show that in the decade prior to 2015 heat illness accounted for 276 deaths in children 1–16 years of age (0.2% of total unintentional injury deaths in this age

group in the USA during this period).

Most heat-induced illness in children is due to overheating from exogenous sources. Hyperthermia differs physiologically from fever. Fever is caused by an elevation of body temperature secondary to hypothalamic regulation. In children, high body temperature that is not a result of hypothalamic thermoregulatory mechanisms is usually due to:

- prolonged exposure to high ambient temperature (overheating)
- Increased heat production
- reduced heat loss.[2]

Children are at increased risk of heat-related illness because:

- children have a greater body surface area relative to mass and therefore are more susceptible to radiant heat
- young children have less cardiovascular compensatory mechanisms to deal with heat insult; older children, if acclimatised, show the same ability to compensate as adults[3]
- young children produce less sweat which limits their ability to lose heat through evaporation
- children depend on parents or adult caregivers to protect them from exogenous heat sources and cannot manipulate their environment to cool themselves.[4]

Certain genetic disorders such as ectodermal dysplasia and Fabry disease also put children at risk of heat-related illness. They have impaired heat dissipation through sweating.[5]

Rising temperatures and more extreme weather events due to climate change has the potential to increase the incidence of heat-related illness in children.[6]

Causes of heat-related illness

1. Overheating

   Overheating most commonly results from high ambient temperature, especially when associated with high humidity. Examples include participation/attendance at summertime sporting events and exposure during heat waves, defined in

Australia as ‘three or more days of high maximum and minimum temperatures that are unusual for that location’.[7]

Young children left in cars are particularly at risk. A review of 171 cases of heat-related car deaths found that in approximately 25% of cases the child gained access to an unlocked vehicle (and was unable to extricate him/herself) and in 75% of cases, the child was left inside a vehicle by an adult.[8]

2. Increased heat production

Metabolic derangements, such as hyperthyroidism, can increase heat production. Drugs including anticholinergics (such as atropine), sympathomimetics, aspirin, neuroleptic agents, selective serotonin re-uptake inhibitors can also increase heat production and are a feature of some classical toxidromes.[9] Amphetamines, methamphetamine and their synthetic derivatives are often used as recreational drugs and are known to induce hyperthermia.[9,10]

3. Decreased heat loss

Excessive wrapping and/or clothing (especially during hot and/or humid days) may cause clinically significant heat trapping.[11] Some chronic illnesses, such as cystic fibrosis, reduce the ability of individuals to lose heat.[12] Heart disease is a rarely encountered but potential cause for decreased heat loss. Drugs including phenothiazines and anticholinergics may affect normal physiological heat loss mechanisms.[9]

Clinical syndromes

A number of heat-related clinical syndromes may present to the ED.

The neonate/infant

Overheating, often in conjunction with excessive wrapping and/or clothing is the most common factor in the neonate and infant. It is important to distinguish the healthy infant who is overheated from the febrile infant (Table 22.3.1).[13]

Mild overheating is usually not dangerous to healthy infants though there may be some association with apnoeic episodes in premature babies. More significant hyperthermia from overheating has been associated with sudden death, particularly in families with a history of malignant hyperthermia (MH).[13,14]

Infants with heat-related illness may present with non-specific features of serious illness. The ‘ABC fluids in and out’ approach is a method of approaching this probem.[15]

Heat syncope

Heat syncope may affect children who have been standing for prolonged periods in hot/humid weather or who have undergone strenuous exertion. They present suffering orthostatic syncopal or pre-syncopal symptoms. Orthostatic measurable haemodynamic changes may not necessarily be present. The main pathophysiological mechanisms are reduced vasomotor tone and hypovolaemia.[3,16,17]

Table 22.3.1

The overheated and febrile infant

Overheated infant	Febrile infant
High rectal temperature	High rectal temperature
Warm hands and feet	Cool hands and feet
Pink skin	Pale skin
Extended posture	Lethargic
Healthy appearance	Looks unwell
Abdomen temperature exceeds hand temperature by <2 degrees	Abdomen exceeds hand skin temperature by >3 degrees

Heat cramps

Heat cramps occur in the setting of strenuous exercise in hot/humid conditions. The term is actually a misnomer because they can occur without overheating being a factor; rather, they are more likely the result of electrolyte imbalance due to excessive sweating.[18] Hypotonic fluid consumption may exacerbate symptoms. Where exercise is a factor, fatigue and neuromuscular control may also be causative factors.[17] They can be isolated or occur in conjunction with heat exhaustion.[17,18] Abdominal muscle cramps have been known to simulate an acute abdomen.[19]

Heat exhaustion

Heat exhaustion in children presents as hyperpyrexia, vomiting, headache,

lethargy and weakness with a normal mental state. The major problem is body water depletion; however, in some paediatric patients (e.g. cystic fibrosis where greater amounts of salt are lost in their sweat), heat exhaustion may occur predominantly due to salt depletion.[19]

Heat stroke

Heat stroke is defined as a core body temperature above 40°C in association with acute mental state changes.[16] There are two types: exertional and classical. In the paediatric population, exertional heat stroke is most likely to occur in the adolescent who is exercising vigorously in a hot and humid environment.[3,16] Sweating is usually present in this group. Classical heat stroke, however, may be seen in children in hot environments, such as during heat waves or when children are left in cars in hot weather.[16,19]

The primary pathophysiological mechanisms occurring in heat stroke are protein denaturation and induction of systemic inflammatory response syndrome as a consequence of endothelial damage. Disseminated intravascular coagulation occurs as a direct consequence of activation of the coagulation cascade by heat. Dehydration is present and may be severe, contributing mainly to cardiovascular and renal compromise. However, shock in heat stroke is primarily distributive, due to extreme peripheral vasodilatation.[16]

Table 22.3.2

Comparative table of the signs and symptoms of heat-induced syndromes

Condition	Symptoms	Signs
Heat syncope	Dizziness, feeling faint, may have brief loss of consciousness	Pallor, sweating, moist and cool skin, normal body temperature
Heat cramps	Painful, brief muscle cramps during or after strenuous exercise	Spasm of affected muscle group, body temperature usually normal
Heat exhaustion	Vomiting, headache, lethargy, weakness, normal mental state	Signs of dehydration, tachycardia, orthostatic hypotension, core temperature <40°C
Heat stroke	Episode of loss of consciousness common, neurological symptoms, irritability, confusion	Core temperature >40°C, acute change in mental state, sweating often present if exertional heat stroke

The organs most susceptible to heat stroke are the brain, particularly the cerebellum, and the liver. Irritability, confusion and ataxia are common.[16,19] Seizures may be seen, particularly during cooling. Unresponsiveness (‘coma’) is common at presentation; however, the child’s conscious state may improve in the

pre-hospital environment.

Other serious complications of heat stroke include cerebral oedema, liver injury, renal failure secondary to hypoperfusion and rhabdomyolysis, non-cardiogenic pulmonary oedema and disseminated intravascular coagulation. Laboratory findings include normal or elevated sodium, chloride and elevated CK.[16,19]

Table 22.3.2 presents a comparison of signs and symptoms of heat-induced syndromes.

Other heat-related syndromes

MH is most commonly encountered in the paediatric patient in the context of general anaesthesia. Seventy-five per cent of victims have no family history and 20.9% have had a previous 'normal' anaesthetic.[20] Mutations in the gene that programmes the ryanodine receptor, which is a tetrameric calcium release channel in the sarcoplasmic reticulum, are associated with MH.[21] This gene defect of the ryanodine receptor is present in 50% of cases. Approximately 18% of all cases of MH occur in children, and their mortality rate is lower as compared to adults.[22, 23]

Initially, an increase in heart rate is followed by an elevation of blood pressure. Because these children are often paralysed, tachypnoea may not be seen. Elevation of the end tidal CO_2 is also an early sign. Muscle rigidity or increased tone may become apparent and the body temperature rises at a rate of 1–2°C every 5 minutes. When succinylcholine is used, however, an acceleration of the syndrome may occur. Hyperkalaemia, hypercalcaemia, metabolic acidaemia and myoglobinuria follow.

The condition of masseter muscle rigidity (MMR) is said to be associated with MH. The peak age group is 8–12 years. It occurs after administration of succinylcholine. In approximately 50% of patients MH occurs after MMR is first seen. The rigidity can usually be overcome with effort and resolves after 2–3 minutes. Repeat doses of succinylcholine do not help.

The association between MH and skeletal abnormalities, such as osteogenesis imperfecta, is unclear.[20]

Serotonin syndrome is the clinical syndrome seen in the setting of excessive serotonin neurotransmission as a result of the ingestion of serotonergic agents. The triad of clinical features of the serotonin syndrome consists of central nervous system, autonomic and neuromuscular effects. Neuroleptic malignant syndrome (NMS) is a rare but potentially lethal syndrome, the exact aetiology of

which is unclear. It is seen in children and adolescents taking antipsychotic medication. It has been suggested that the duration of NMS was one-third shorter in children taking atypical antipsychotics when compared with older, more typical agents.[24] Methylphenidate (Ritalin) has been implicated in NMS. The anticholinergic syndrome is usually seen in the setting of deliberate self-poisoning with potent anticholinergic agents.[25] Clinically there are both central and peripheral features. The latter are an extension of the physiological effects of blocking cholinergic receptors.

Other drug-related toxidromes from amphetamine use (e.g. ecstasy at rave parties) and other drugs (e.g. salicylates, anticholinergics) can cause heat-related emergencies in the adolescent group. Vigorous activity, high ambient temperatures and ingestion of amphetamine compounds contribute to the risk of hyperthermia at rave parties.[26]

Investigations

Heat-related illness in children is a clinical diagnosis. Rectal temperature is the most accurate means to evaluate temperature in such patients. Investigations are directed at the complications of heat-related illness in the paediatric patient.[16,17,19]

Children who present with heat syncope or heat cramps usually do not require any investigations. If performed, laboratory findings in patients with heat cramps include decreased serum sodium, chloride and normal or slightly elevated blood urea.[19]

Those children with more severe heat-related illness require further workup. In heat exhaustion, basic bloods including full blood examination, urea and electrolytes, as well as serum creatine kinase and glucose should be performed. Urinalysis would also be indicated.

The paediatric patient with heat stroke or other life-threatening hyperpyrexial illness requires full investigation: full blood examination, electrolytes, liver function tests, total creatine kinase, coagulation studies, urinalysis, arterial blood gases and 12-lead electrocardiogram.[16,17,19]

Management

Heat syncope and heat cramps

For children with heat syncope and heat cramps, external cooling and oral fluids (e.g. gastrolyte) are usually sufficient. These patients usually do not require

admission.[16,17]

Heat exhaustion

The child with heat exhaustion needs fluids (oral if able to tolerate, otherwise intravenous [IV]) and rest in a cool environment. The decision to admit will depend on a number of factors.[16,17]

Heat stroke

Heat stroke in children is a medical emergency. The ABC approach is important and all children with heat stroke should be triaged to a resuscitation environment.[16]

Treatment of heat stroke focuses on two main goals: immediate elimination of hyperpyrexia and support of the cardiovascular system. As mortality and morbidity are correlated to the duration of hyperpyrexia, cooling should commence as early as possible, preferably in the field.[16,17] Rapid external cooling performed with evaporative methods is considered the most effective and practical method. Remove all the child's clothes, spraying with water and fanning (the 'wet and windy approach') and applying ice packs to the neck, groin and axillae. Initial cooling should aim for a core temperature of 38.5°C.[16,19] Shivering, which can generate heat, can be minimised by the administration of benzodiazepines. Phenothiazines do inhibit shivering but have anticholinergic properties including sweat inhibition and are contraindicated.[16] Antipyretics, such as paracetamol or ibuprofen, have no role. Cold lavage techniques (peritoneal, gastric, rectal and bladder) are invasive and evidence for efficacy is lacking. Cold immersion is impracticable. Cold IV fluids are not usually used but are being further investigated, especially for preservation of the brain.[16]

All children with genuine heat stroke require IV fluids, an indwelling urinary catheter and admission to the intensive care unit. As shock in heat stroke is mostly distributive, initial 'permissive hypotension' is practiced to avoid pulmonary oedema until cooling has substantially corrected core body temperature.[16] The fluid of choice is normal saline, as substantive sodium loss from sweating usually occurs.

The patient should be monitored for complications including non-cardiogenic pulmonary oedema, cerebral oedema, disseminated intravascular coagulation and renal/hepatic failure. Early and active treatment of these life-threatening

complications should be instituted when they occur.

Malignant hyperthermia

The principles of treatment of MH are similar to those of heat stroke. If MH has been caused by an inhalational anaesthetic, the agent should be stopped immediately. Dantrolene is the antidote, inhibiting the release of myocyte calcium. Several regimes have been suggested. Shann suggests an initial dose of 1 mg kg^{-1} min^{-1} until improvement, with a maximum dose of 10 mg kg^{-1} total.[27] Another regime suggests giving 2.5 mg kg^{-1} IV and repeating every 10 minutes to a maximum of 10 mg kg^{-1}.[21] Other manoeuvres include hyperventilation with 100% oxygen, bicarbonate in fulminant cases (2–4 mEq kg^{-1}), lidocaine (lignocaine) to treat arrhythmias and aggressive cooling of the child (see above).

Serotonin syndrome, neuroleptic malignant syndrome and anticholinergic syndrome

These conditions are rare in children as serotonin syndrome is usually seen when drug combinations are used, whereas NMS is most commonly an idiosyncratic reaction to a single agent. In patients with serotonin syndrome, most resolve within 24–48 hours with supportive management and cessation of the serotonergic agent/s. In severe cases, attention to ABCs, continuous core temperature monitoring and titrated IV benzodiazepines are indicated.[25,28] Specific serotonin antagonists have also been used such as cyproheptadine (0.1 mg kg^{-1} per dose) or chlorpromazine (0.25–1.0 mg kg^{-1} per dose). In NMS, benzodiazepines are useful in mild cases. In more severe cases, bromocriptine and dantrolene may have a role. The treatment of anticholinergic syndrome is similar. Physostigmine (0.02 mg kg^{-1} per dose up to a maximum of 0.1 mg kg^{-1}) is a centrally acting acetylcholinesterase inhibitor. Its exact role is controversial, but one review has suggested that in confirmed cases of anticholinergic poisoning in children, physostigmine might be more effective than benzodiazepines.[29]

Prognosis and disposition

Apart from children with mild heat-related illness (i.e. heat syncope, heat cramps and mild heat exhaustion), who can be managed in the ED, all others should be

admitted to hospital. Any child with suspected heat stroke should be admitted to intensive care.

Prognosis in heat-related illness is related both to the absolute temperature level and the duration of the elevation.[30,31] Persisting coma (greater than 3 hours) and multi-organ failure are poor prognostic signs. As prognosis correlates with duration of hyperthermia, immediate cooling is critical. The mortality rate for heat stroke is approximately 10%. For malignant hyperthermia mortality is 7%.

Prevention is a major intervention, particularly in babies and infants. Prevention centres on educating new parents about the risks of sun exposure, clothing and leaving children in cars in hot weather even for brief periods. Sports medicine guidelines regarding older children taking part in sporting activities in hot, humid weather need to be in place.[3,8,16,17]

Controversies

Controversies relevant to heat-related illness in children include

1. the likely effects of climate change on environmental heat-related illness in the future
2. cooling techniques used in children
3. the association between malignant hyperthermia and masseter muscle rigidity
4. neuroleptic malignant syndrome occurring with methylphenidate (Ritalin) use
5. The lack of public education about the susceptibility of infants and young children to heat-related illness compared to adults.

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22.4

Cold injuries

Nicholas Cheng, and Ioannis Pegiazoglou

ESSENTIALS

1 Hypothermia is defined as a core body temperature less than 35°C.

2 Cold injury is uncommon in Australia, but when cold injuries do occur it is more significant in children than in adults.

3 The major complications of hypothermia are altered mentation and arrhythmias.

4 Hypothermia is treated with passive external, active external, active internal and/or extracorporeal rewarming, depending on severity. In addition, hypothermia should be rigorously prevented in any patient with a severe illness.

5 The major complications of rewarming are an ‘after-drop’ in temperature and vasodilatory shock.

6 Severe local cold injuries are treated with rapid immersion rewarming, then care given as for burns.

7 In Australia patients are almost always dead, then cold. Situations where patients become cold enough to apply ‘warm and dead’ criteria are very rare.

Introduction

Cold injury is uncommon in Australia due to its warm climates. However, its significance is more important in children, as they have:

- a larger surface area to weight ratio[1]
- underdeveloped behaviour responses to cope with extreme cold (young children can't put on more clothes).

When cold injuries do occur, they can be subdivided into generalised injury (namely accidental or environmental hypothermia) and localised injury. Cold injury is also a common problem potentially complicating any other severe illness (especially trauma) and must be prevented.[2]

A recent study showed that the diagnosis of accidental hypothermia significantly increased mortality in hospitalised children in US hospitals (8.45% compared to 0.32% without hypothermia).[3]

Normal physiology: a review

Heat production is derived from basal metabolism, digestion and muscular activity, which may be voluntary (exercise) or involuntary (shivering). Emotional factors and hormonal fluctuations influence heat production. The main mechanisms by which the body compensates for low-core-body temperature are by increasing its metabolic rate, primarily through shivering and by shunting blood away from non-essential organs to preserve vital organs. The capacity to shiver is dependent on local glycogen stores and the rate of change of core and external temperature.[1, 4]

Neonates are the patients most prone to hypothermia. They are unable to shiver and have limited stores of energy. Because of this, newborn children utilise catabolism of brown fat to generate heat. This is an inefficient process that consumes oxygen, thus exacerbating hypoxia. In addition, the large surface area to weight ratio, due to a relatively large head, contributes to heat loss. At birth, neonates are covered in amniotic fluid, and evaporative losses are significant. An overhead radiant heater is not adequate to compensate for this evaporative loss.[5,6]

Heat loss from the human body is by four methods:

- Radiation
- Conduction
- Convection
- Evaporation.[1]

Radiation occurs when heat energy leaves the skin at the speed of light. Patients with more fat become more hypothermic than thinner patients, due to the former's larger surface area for radiation heat loss. In children, who have a higher surface area to weight ratio, it accounts for up to 50% of all heat loss; indeed, up to 75% in neonates. This higher number in neonates is due to a proportionally larger head increasing the surface area to weight ratio.[1,5] Radiation losses decrease when a patient is clothed.

Conduction of heat is poor in air and therefore does not contribute much to hypothermia in normal circumstances. However, water-conductive heat loss is 24 times more than that of air. It is this method by which patients suffering from water immersion become profoundly hypothermic, and how patients in wet clothes become hypothermic quickly. The surface on which the patient is lying also contributes to conduction heat loss. For instance, a patient lying on snow is likely to become more hypothermic than a person lying on sand.

Convection occurs as warm air next to the skin is replaced by cool air. This can contribute to 25% of total body heat loss in still air. In a wind of 63 km h^{-1}, this increases by 14 times. This is described as the 'wind chill factor'.

Evaporation from the skin accounts for only 7% of heat loss at rest. This may be increased in cold, dry conditions and by sweating. Evaporation from the respiratory tract removes another 7%. This can be increased by faster breathing (such as at high altitude or during exercise).

Temperature is perceived through central and peripheral mechanisms. Heat sensors in the central hypothalamus receive input from the skin, central arteries and viscera. It is this central thermostat that is reset, which causes fever. Skin receptors respond to a change in skin temperature but do not themselves indicate the patient's core temperature. A result of all this input is that the body responds by those autonomic reflexes listed below to increase or decrease core body temperature.[4]

Hypothermia

This is defined as a core temperature of <35°C.[1] Hypothermia is classified on the basis of severity. The reason for this classification is that it influences the rewarming mechanisms that are most often deployed. It is also related to the physiological ability of the patient to compensate for hypothermia. An easy way to remember these temperature ranges is in the following way:

- Acceptable low temperature: 2°C below 37°C (>35°C)
- True mild hypothermia: 3°C below that (32–35°C)
- Moderate hypothermia: 4°C below that (28–32°C)
- Severe hypothermia: anything below that (<28°C).

Table 22.4.1

Compensatory mechanisms at different severities of hypothermia

Mild (32–35°C)	Moderate (28–32°C)	Severe (<28°C)
Increased basal metabolic rate by shivering Vasoconstriction peripherally, leading to fluid shift Mild tachycardia Cold diuresis (see text) Apathy, ataxia, amnesia, dysarthria	Limits of increasing basal metabolic rate reached Shivering stops Decreasing cerebral blood flow, causing delirium and gradual decreased level of consciousness Decreased rate of neural impulse transmission, causing clumsiness and numbness Muscle rigidity due to increasing acidosis	Complete loss of thermoregulation Stupor, coma Pulseless Fixed dilated pupils Absent reflexes Dysrhythmias, initially slow atrial fibrillation then ventricular fibrillation Appearance of death at <25°C with asystole Falling blood pressure See text for more specific changes

Table 22.4.2

Findings at low body temperatures

Temperature (°C)	Findings
27	Reflexes absent, no response to pain, comatose
25	Cerebral blood flow one-third of normal, cardiac output one-half of normal
23	No corneal reflex, ventricular fibrillation risk is maximal
19	Asystole, flat EEG
15	Lowest temperature survived from accidental hypothermia

Tables 22.4.1 and 22.4.2 show the main consequences of hypothermia at a given temperature.[1,4,7] Much of our understanding of this pathophysiology comes from controlled hypothermia in cardiac surgery. Note that there is a huge variation of the onset of certain clinical signs based on temperature level. For instance, some patients may exhibit confusion at higher temperatures compared with others. Note that in children clinical manifestations of altered consciousness may be subtle.

Note that only during severe hypothermia does protection from hypoxia occur,

due to decreased demand for oxygen by tissues and even then only at extremely low temperatures (patients <20°C can tolerate anoxia for up to 60 minutes). Metabolic processes slow by approximately 6% for each 1°C drop in body temperature.[1] Thus at 28°C the basal metabolic rate is about 50% of normal. This leads to hypoventilation and hypoxia. However, at this temperature the decreased cellular metabolism affords some protection against hypoxia.

Cold diuresis is an initial brisk diuresis; this is due to decreased tubular reabsorption and also a decreased production of antidiuretic hormone. There is also an increased central blood circulating volume as blood is shunted away from the periphery, thus presenting the kidneys with an apparent increased blood volume for filtration.[1]

History

Important points in history are:

- approximate time of exposure, if known
- environment in which patient was found
- resuscitation at the scene, including duration of time with no cardiac output (‘downtime’)
- pre-existent illnesses (e.g. thyroid disease)
- drugs, medications, allergies and immunisation status.

Examination

- Full primary and secondary assessment of patient
- Accurate measurement of core body temperature.

Diagnosis

This requires only two essentials:[1]

- A thermometer able to record low core temperatures accurately
- A high index of suspicion.

Core temperatures can be measured best with oesophageal or rectal probes. The most direct method of measurement is with a cardiac catheter such as a

Swan–Ganz, but this is impractical in the emergency setting. Rectal probes are often used,[8] but care must be taken when using these. The probe must be at least 10 cm into the rectum in older children (more than 8 years old) and 5 cm in younger children. Inaccuracies may occur due to the presence of faecal material,[1] and the probe must be left in until the temperature equilibrates. Tympanic measurements are well known to be unreliable in the very young,[8] but they are a good indicator of therapy progression in the older child. Oral and axillary temperature probes are unreliable and impractical in the setting of true hypothermia.

Treatment

Pre-hospital treatment

This is mainly the realm of passive external rewarming methods (see below). Patients should be carefully removed from the precipitant cold environment to a dry, sheltered area. If clothes are wet they should be removed and the patient dried and covered with a warm dry blanket. All patients should be gently handled, especially during transport, as there is evidence that sudden movements to a body in severe hypothermia can precipitate arrhythmias, particularly ventricular fibrillation.[7] While this is occurring one should attend to the patient's airway, breathing and circulation, as per any resuscitation.

Active rewarming should be avoided until the patient reaches the emergency department (ED). This is because of the complications of rewarming, namely 'after-drop' and shock.[1,7,8]

Treatment in the emergency department

Once in the ED, the patient should be triaged to an appropriate area, which is warm. In the very young, a radiant warmer bed and heating lamps should be available when the patient presents.[5,9] Patients should have their airway, breathing and circulation reassessed and appropriate resuscitation commenced. Appropriate monitoring should be instituted, including electrocardiogram (ECG) and core temperature, either by rectal or oesophageal means. Oxygen saturation monitoring should be attempted, whilst understanding that initial vasoconstriction will give inadequate readings. Urine output should be monitored. Gentle handling should be continued to avoid precipitation of arrhythmias.[7] Patients should continue 100% oxygen on arrival in emergency.

Blood tests taken should include arterial blood gas (ABG); full blood count; electrolyte, urea, creatinine (EUC); liver function tests; amylase; comprehensive metabolic panel (CMP); glucose; thyroid function tests (TFTs); coagulations; tests for infection; and, if suspected, a screen for sedative drugs and ethanol. Hypothermia causes measured pH to fall and pO_2 and pCO_2 to be higher. It is recommended that these ABG values should not be corrected for temperature to better reflect the physiological state of the patient.[7]

A 12-lead ECG should be taken. Classic changes include the presence of a J (Osborn) wave, interval (PR, QRS, QT) prolongation, atrial dysrhythmias and ventricular dysrhythmias.[11] Other less well-known changes include abnormalities similar to myocardial infarction. Hypothermia can also blunt the ECG changes with hyperkalaemia.[7] Note that all these are not present in all hypothermic patients.

Once temperature is measured, the severity of hypothermia determines the methods of rewarming. Specific methods of rewarming are classically divided into four categories:[7,8,12]

- Passive external rewarming
- Active external rewarming
- Active internal (core) rewarming
- Extracorporeal rewarming.

Passive external rewarming

These are methods used to prevent excessive endogenous heat loss and promote the patient to self-warm to normal core temperature. They include placing the patient in a warm room to prevent excessive convection, evaporation and conduction heat losses. Sheets of foil (foil space blankets) are placed over the patient to decrease loss of heat through radiation.

Active external rewarming

These are methods used to transfer heat energy to the patient from the external environment. They include:

- warmed blankets
- chemical hot packs and warm water bottles
- forced warm air blankets
- radiant heaters and lights

- warm-water body immersion.

Active internal rewarming

These are methods used to transfer heat energy to the patient internally. They can be subdivided into simple methods (the first two methods below) and invasive methods. They include:

- warmed intravenous (IV) fluids (normal saline 40°C)
- inhalation of humidified warm oxygen (40°C)
- gastric lavage
- bladder lavage
- colonic lavage
- pleural and peritoneal lavage (10–20 mL kg^{-1} of 40°C saline).

Extracorporeal rewarming

These are methods by which the patient's blood is removed, rewarmed outside the body, and replaced. They include:

- haemodialysis
- venovenous transfer of blood
- extracorporeal (coronary bypass) circulation.

Due to the child's larger surface area to weight ratio,[1,6] emergency physicians should start to institute limited active external rewarming methods, such as radiant light warmers and forced-air warming blankets, even in mild hypothermia.[12] If the child is unable to produce extra heat, then the institution of simple active internal rewarming methods (warm humidified oxygen and warmed IV fluids) is appropriate.

In older children, asking them to drink some warm liquids (hot chocolate or soup) will in effect give them the effects of a warm gastric lavage. However, patients should only have warmed fluids if they are fully conscious, can protect their airway and have no evidence of gastroparesis.

Moderate hypothermia (28–32°C) should have all active external rewarming techniques instituted except immersion in warm bath therapy, which is limited to localised cold injury in the emergency setting (see frostbite below). It is almost impossible to adequately monitor patients while they are in an immersion bath. Forced-air warming blankets can increase core temperature by up to 1.5°C h^{-1}.

Patients should also have warmed IV fluids and heated humidified oxygen for inhalation. Together, they can increase core temperature by 1–2°C h^{-1}. Normal saline bags can be safely warmed in a microwave oven. The optimum operating system for warming 500-mL bags of crystalloid is 400-W microwave for 100 seconds or 800-W microwave for 50 seconds.[13] Alternatively, if a heat infusion pump is available, this should be used. IV infusion tubing should be as short as possible, as longer tubing loses more heat to the atmosphere.

Severe hypothermia requires institution of invasive core-rewarming techniques. All can raise core temperature by 2°C every 5 minutes.[7] In cardiac arrest, cardiopulmonary resuscitation (CPR) should be commenced until core temperature has reached 35°C, and then a further assessment of the patient done (see Controversies). Note that, according to criteria for diagnosing brain death as quoted from the Australian and New Zealand Intensive Care Society,[14] the patient must have a core temperature above 35°C, whereas other sources say 32°C.[15]

The decision of who to rewarm continues to evolve. There is a case report of a 26-month-old patient with a core temperature of 15°C who, after rewarming, recovered neurologically intact.[16] For patients without submersion, extreme duration of exposure is not incompatible with life. In immersion patients, successful recovery is very rarely seen unless patients are immersed in ice-cold water (<10°C) for long periods of time (see Chapter 22.2). However, further attempts to resuscitate after failure to restore a circulating cardiac rhythm within 30 minutes of rewarming to above 32°C are likely to be ineffective.[15] In most Australian cases, however, the adage ‘you’re not dead till you’re warm and dead’ does not apply. The above only relates to patients that are ‘snap frozen’ in snowy weather.

With the recent evidence of inducing hypothermia for out-of-hospital arrests, it would make sense for patients who present hypothermic due to out-of-hospital arrests to be warmed to 34°C. In this way, hypothermia has been treated, whilst giving the patient admitted to the paediatric intensive care unit an opportunity to recover with the best possible neurological outcome.[17]

Neonatal resuscitation

In neonatal resuscitation, a warm ambient environment for the newborn child is prepared. A heated room with an overhead warmer is required. Warmed towels and blankets are used to rapidly dry the newborn to prevent evaporative heat loss. Unwell neonates should be admitted to special care nurseries or neonatal

intensive care within humidicribs or transport cribs with radiant heat. If ventilated, humidified warm gases should be used.[5,6]

There is now increasing evidence that in neonates suffering hypoxic ischaemic encephalopathy a period of hypothermia may be beneficial (see Controversies). Some centres in Australia have already changed their practice to cool such neonates. The reader is advised to consult local guidelines and their referring neonatal intensive care unit for more information, and the latest resuscitation guidelines of their country which were released following the latest ILCOR guidelines released in late 2015.

Complications

Complications can be classified as due to the hypothermia itself and complications as a result of rewarming.

Complications from hypothermia include:[1,4]

- cardiac: arrhythmia
- haematological: platelet dysfunction, thrombocytopenia and disseminated intravascular coagulopathy
- pulmonary: pneumonia, pulmonary oedema and (adult) acute respiratory distress syndrome
- infection: immune complex suppression
- renal: acute tubular necrosis and rhabdomyolysis
- neurological: cerebral oedema, prolonged coma and slow neurological recovery (up to 6 months)
- gastrointestinal: pancreatitis
- biochemical and metabolic derangements (including glucose, sodium and potassium).

There are two main complications that result directly from the rewarming of patients: after-drop and shock.[1,7,9] After-drop is a drop in core temperature after rewarming therapies have commenced. There are two proposed mechanisms for after-drop:

1. Cold peripheral blood re-enters the circulation once peripheral vasodilatation occurs with rewarming.
2. After-drop is due to ongoing conduction of heat from the warmer core

into colder peripheries and surface layers of the body.

In reality, after-drop is most probably a combination of the two mechanisms described. This can be minimised by ensuring that rewarming only occurs over the core of the body, while the peripheries of the body are not actively rewarmed. For instance, a forced warm air blanket is placed over the patient's body, but his/her hands and feet are left outside the blanket.

Shock occurs from the same mechanism, where peripheral vasodilatation increases the intravascular space to be filled, causing a consequent drop in blood pressure and rise in heart rate. In addition, the cold diuresis experienced has already decreased the circulating blood volume (up to 35%). In immersion patients, there is also a hydrostatic squeeze effect, which further decreases blood volume. Rewarming should therefore occur only once vascular access is established and warmed normal saline is instituted if active rewarming occurs, starting with usual shock doses of 20 mL kg^{-1}. Insertion of a central venous pressure (CVP) line would be useful; however, this needs to be balanced against the increased rough handling of the body during insertion.

Medications in hypothermia behave unpredictably. Metabolism of most drugs will be slowed due to hypothermia. Some drugs have decreased effectiveness; others, such as morphine, have increased effects. Drugs not used in treatment include sodium bicarbonate, insulin, corticosteroids, empirical antibiotics and ethanol (contrary to popular belief).[4,7]

Electrical defibrillation and antiarrhythmics may be administered at any temperature, but most efforts do not succeed until the temperature reaches above 28–30°C.[4]

Disposition

A patient with mild hypothermia, once treated, may be observed in the ED for a few hours before discharge if the patient is well. Moderate and severe hypothermia are an indication for ward or intensive care unit admission, depending on if cardiac abnormalities are present during assessment. Discharge should include education of patients on prevention of further occurrences, for example the proper use of clothing, checking weather reports, etc.[1]

Localised cold injuries

Frostbite is the most severe of the injuries, but others include chilblain (perniosis), cold-induced fat necrosis (panniculitis), frost nip and trench foot.[18,19]

Frost nip is transient blanching and numbness of peripheries that resolves with rewarming. Only the skin surface is damaged. No ice crystals form within tissues, as opposed to frostbite (see below).

Trench foot, also known as immersion foot, is historically the most interesting, with the most famous cases occurring at Napoleon's failed invasion of Russia in the winter of 1812. It is due to prolonged exposure to wet and cold, most common in the feet when poorly ventilated cold shoes are worn. There is peripheral neurovascular damage but no ice crystal formation within tissues.

Perniosis or chilblain is injury due to repeated exposure to dry cold. Bullae form 12 hours post injury. These bullae burst to form ulcers, which are painful and pruritic. It is possibly due to repeated vasoconstriction. Common areas involved are the feet, lower legs and face.

Panniculitis consists of red lesions due to cold. Treatment is with non-steroidal anti-inflammatory drugs (NSAIDs), and the lesions resolve in 10–21 days.

Frostbite

Frostbite is the most dangerous of the local cold injuries. It is caused by freezing and ice crystal formation within the interstitial and cellular spaces due to prolonged exposure to freezing temperatures. It tends to occur more if the skin is directly exposed to temperatures less than –10°C. Several pathogenic phases evolve, called the frostbite injury cascade:[9,19]

1. Prefreeze phase. Superficial tissue cooling occurs, which leads to increased blood viscosity, microvascular constriction and endothelial plasma leakage.
2. Freeze phase. Ice crystals form in the extracellular space, leading to disruption of endothelium, disruption of cell anatomy and hyperosmolality within cells due to crystals osmotically drawing water out of cells. This leads to protein denaturation and DNA synthesis inhibition.
3. Vascular stasis. There is arteriovenous shunting within damaged tissue, leading to stasis coagulopathy and thrombus formation.
4. Late progressive ischaemic phase. The thrombus induces inflammation,

distal hypoxia, and anaerobic metabolism, which eventually leads to tissue necrosis.

Clinical features and diagnosis

Symptomatically, frostbite begins as an initial coldness of the skin. It then progresses to a stinging or burning pain, then to an anaesthetic limb, loss of fine motor function, loss of gross motor function and, finally, severe joint pain. Examination of the affected limb will reveal varying degrees of frostbite.[19] In the past, they have been classified similarly to burns, from first-degree to fourth-degree injury. It is much easier to classify them as superficial or deep.[9] Superficial injuries go only to the skin and subcutaneous tissues, whereas deep frostbite also affects bones, joints and tendons.

Most investigations are unhelpful. However, a full blood work up with full blood count, EUC, liver function tests, glucose and creatine kinase to look for rhabdomyolysis should be done. Urinary myoglobin should be checked. Imaging is unhelpful initially; however, there is some evidence that a bone scan will help surgeons later determine how much limb is still viable and whether superimposed osteomyelitis has developed.[9,19,20]

Treatment

Frostbite is a challenging condition with limited therapeutic options. Paediatric patients are also at risk for damage to the epiphyseal growth plates in the affected bones. Premedical treatment involves preventing further hypothermia and initiating resuscitation. As for hypothermia, removing the patient from the cold environment is paramount. Rubbing the limb to try to warm it should be avoided, as this increases tissue damage.[9,19]

Once in emergency, the mainstay of treatment is rapid immersion rewarming.[9,19] The affected limb should be placed in a whirlpool of water about 40°C for 20–40 minutes for superficial frostbite, 1 hour for deep frostbite. This procedure will be painful, sometimes exceedingly so, hence narcotic and NSAID analgesia should be started before treatment is commenced. Rewarming is complete when the distal area of the limb is flushed, soft and pliable. The patient should start to move the limb during rewarming to encourage blood flow in the limb. The main reason for suboptimal results is premature cessation of rewarming. After rewarming, the limb is dried and placed in a splint and

elevated, and dressings applied four times a day.

Blistering of the limb will occur. The blisters are usually clear in superficial frostbite and haemorrhagic blisters in deep frostbite. Controversy exists as to whether to aspirate these blisters. Haemorrhagic blisters should not be aspirated, as this increases trauma to the wound. The treatment of clear blisters is not as well defined.

All patients should have their tetanus status updated, and, as 30% of wounds become infected, IV prophylactic antibiotics may be of use. IV penicillin G is most commonly used.[19]

Negative pressure wound therapy (NPWT) has been used with success in treating early adult frostbite and a new case series of paediatric patients presented promising results.[21]

Disposition

All patients should be admitted under a specialised (burns) unit.[9,19] Patients are observed for up to 6 weeks, which is usually the time the gangrenous parts of the limb are fully delineated so that safe amputation, if necessary, will occur. Good prognostic factors are patients with superficial injuries only, clear blisters and sensation still present after rewarming.[19]

Hypothermia not due to environmental causes

There are many other causes of hypothermia,[1,4] as seen in Table 22.4.3.

Once the hypothermia is treated, then non-environmental causes should be investigated, and, if found, appropriate treatment instituted. Iatrogenic causes need to be prevented. Always prevent hypothermia in trauma patients after full assessment, as hypothermia worsens the diagnosis.

Table 22.4.3

Hypothermia not due to environmental causes

Cause	Details
Metabolic or endocrine	Hypoglycaemia
	Diabetic ketoacidosis
	Hypopituitarism
	Hypothyroidism

	Addison’s disease
	Uraemia
	Malnutrition
Toxicological	Alcohol
	Barbiturates
	Anaesthetic agents
	Carbon monoxide
	Cyclic antidepressants
	Narcotics
	Phenothiazines
CNS disorders	Head trauma
	Spinal trauma
	Subarachnoid haemorrhage
	Degenerative diseases
	Cerebrovascular accidents
	Intracranial neoplasm
Infections	Sepsis
	Meningitis
	Encephalitis
	Pneumonia
Vascular or skin	Shock
	Gastrointestinal tract haemorrhage
	Pulmonary embolism
	Burns
	Erythrodermas
Iatrogenic	Cold fluid infusion
	Exposure during treatment
	Prolonged extrications
	Exposure during transport
	Exposure post birth

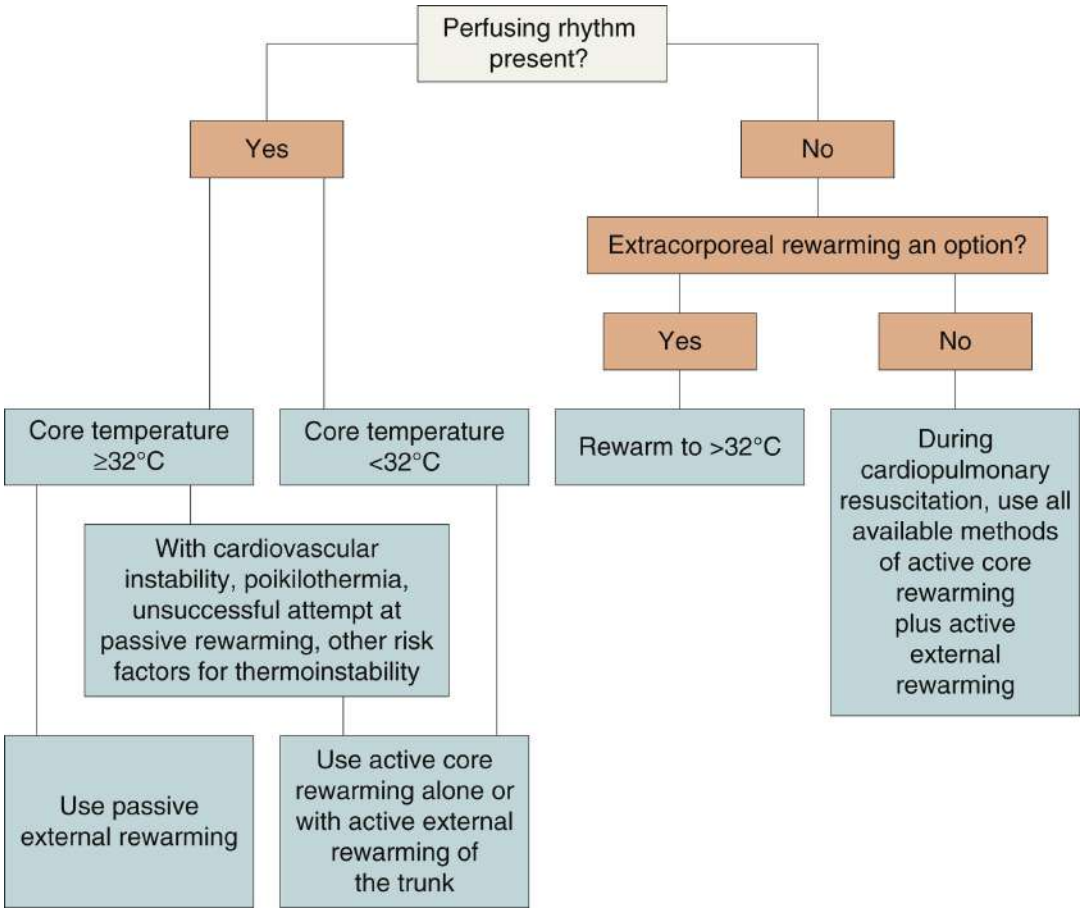

FIG. 22.4.1 Algorithm for rewarming. Adapted from Danzl DF, Pozos RS. Accidental hypothermia. *N Engl J Med* 1994;**331**(26):1756–60.

Controversies

1. In certain countries, active rewarming is commenced prior to arrival in the emergency department.[9] This is due to well-organised emergency medical systems catering for such emergencies, like in Canada. In Australia, this is more of a contentious issue, as we don't see hypothermia as often.
2. Controversy exists regarding cardiopulmonary resuscitation (CPR) causing arrhythmias when a patient loses spontaneous circulation.[1] However, the evidence that CPR causes ventricular fibrillation in hypothermic patients is at best circumstantial, and therefore the general consensus is that, to promote brain perfusion, CPR should be continued till the core body temperature reaches 35°C.[14]
3. Continuing CPR in the absence of cardiac output in a hypothermic

patient has traditionally been mandatory. However, as survival occurs only in patients who have been immersed in water <5°C prior to cardiac arrest,[1] it is more likely that patients arrest in Australia before they become hypothermic. Therefore when to stop cardiac resuscitation is an issue.

4. The use of hypothermia in treatment, particularly in trauma and head injury, has always been of some debate.[12] Hypothermia causes a reduction in oxygen consumption and is theoretically cerebroprotective. This contradicts the results that trauma patients presenting with hypothermia have a worse prognosis.[2]
5. In neonates with hypoxic–ischaemic injury, cooling of the head is now accepted in many centres as best-practice treatment, ahead of changes in the ILCOR guidelines. However, the means to do so is not fully determined. This is an area in which there are rapid developments, and guidelines may change.

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Further reading

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Douwens R. Hypothermia prevention, recognition and treatment. 2003. http://www.hypothermia.org.

An excellent website with up-to-date insight on hypothermia and future directions in management.

Meteorological Service of Canada, . Wind chill charts and tables. Ottawa: Meteorological Service of Canada; 2002. http://www.msc.ec.gc.ca/education/windchill/charts_tables_e.cfm.

An excellent article with excellent graphical depictions of wind chill effects; the reader should also peruse the rest of the website.

22.5

Anaphylaxis

Simon G.A. Brown

ESSENTIALS

1 Intramuscular adrenaline (epinephrine) is the initial treatment of choice and should be administered if there are any respiratory or cardiovascular features.

2 Multiple intramuscular doses and/or an intravenous infusion of adrenaline may be required.

3 Delayed deteriorations, including true ‘biphasic’ reactions, that need further treatment with adrenaline, are relatively common after initially severe reactions and usually occur within 10 hours of initial successful treatment.

4 If adrenaline is administered, observe for *at least* 4 hours after the last dose. If the initial reaction is severe with hypotension and/or hypoxaemia, observe for at least 10 hours or overnight if the patient presents in the evening.

5 A full history should be obtained with the aim of identifying the triggering factor, while the memory of events is fresh.

6 If further exposure to a suspected allergen is considered possible, consider provision of an anaphylaxis action plan and instruction in use of adrenaline injector.

7 An allergy referral is indicated for definitive diagnosis of precipitating factors.

Introduction

Acute allergic reactions resulting from the degranulation of mast cells present as a continuum of responses from mild cutaneous erythema and urticaria to severe hypoxaemia, hypotension, collapse and death. Different authorities include varying components of this continuum in the definition of anaphylaxis, but general consensus is that anaphylaxis is a systemic, multiple-organ allergic reaction that is potentially life-threatening.[1] Anaphylaxis is therefore usually defined as a severe acute allergic reaction that involves the respiratory tract and/or circulatory system with hypotension or clinical features such as collapse, incontinence and syncope that suggest hypotension.

Along with the increasing incidence of allergic diseases, anaphylaxis admissions to Australian, USA and UK hospitals seem to be increasing. Australian studies reported a two- to fourfold rise in anaphylaxis hospital admissions over an 11-year period from 1994.[2,3] The greatest rise was reported in children less than 5 years of age, with an almost sevenfold increase in hospital admissions from anaphylaxis, predominantly due to reactions to foods. It is difficult to ascertain how much this represents a true increase in prevalence of disease and how much is due to changes in clinical practice and data reporting. Despite the reported increase in admissions, death from anaphylaxis in childhood remains rare (the Australian mortality rate has remained stable at 1 per million population per year over an 11-year time period).[3] Thus, although anaphylaxis admissions have increased, particularly in children less than 5 years of age, death from anaphylaxis remains rare. The majority of deaths occur in teenage and adult years, rather than in early childhood.

Pathophysiology

Most of the clinical features of anaphylaxis can be explained by mast cell mediator release. The mediators with vasoactive and/or bronchoconstrictive activities (histamine and the sulfidopeptide leukotrienes) and platelet-activating factor (PAF) are principal factors in the development of anaphylaxis. Rapid systemic spread of immune activation and mediator release is also required for the development of severe anaphylaxis, but the mechanisms are unclear. Mouse models of anaphylaxis demonstrate the critical involvement of neutrophils, and recent genomic and biochemical mediator studies of human anaphylaxis provide compelling evidence for the involvement of neutrophils.[4,5] Thus a ‘mast cell-

leukocyte cytokine cascade', like that proposed in the context of allergic airway inflammation, may also apply in the context of acute anaphylaxis.

An increase in vascular permeability causes loss of fluid from the circulation into the interstitial space. The relative contributions of the mediators to the various clinical features are not completely defined. Mediators may directly compromise cardiac muscle function and the intravascular volume depletion from extravasation further compromises cardiac output. In conscious patients, bradycardia, probably a reflex response to reduced venous return and reduced heart filling, is common and accompanies the onset of hypotension.[6] Both upper and lower airways are affected. Laryngeal oedema results in a variable degree of upper airway obstruction, and bronchospasm and increased mucus secretion compromise the lower airways. Age, comorbidities and medications may also influence severity.[7] Thus, reactions may be more severe in older patients in general, the effects of bronchospasm may be more marked in asthmatic patients and cardiac pump failure may be prominent in those with already impaired cardiac function. Beta blocker therapy may worsen bronchospasm and further reduce cardiac output.

Aetiology

The triggering agent can be identified in about three-quarters of patients presenting to emergency departments (EDs).[8]

In one childhood series the triggers were:[9]

- food: 50%
- medications: 25%
- insect stings: 10%
- immunotherapy: 1%
- immunisations: 1%.

In some series, food anaphylaxis comprises most presentations,[10] but triggers vary between areas and age groups, presumably a consequence of differences in early childhood exposure (immune tolerance) and later cumulative intermittent exposures and thus the risk of developing allergy. In rural and outer metropolitan areas sting anaphylaxis is the most common cause across all age groups at 30% overall.[11] Food anaphylaxis is more common in young people, whereas sting anaphylaxis and drug anaphylaxis increase in prevalence with age.

Clinical features

Symptoms occur along a continuum, from reactions that are primarily cutaneous in nature, through mild to moderate anaphylactic reactions that may have respiratory symptoms but without tachypnoea or hypotension, to severe life-threatening anaphylaxis with hypotension and hypoxia.

In children, cutaneous (90% of cases) and respiratory (80% of cases) manifestations occur earlier and are more common than gastrointestinal and cardiovascular manifestations. In 'food' anaphylaxis, gastrointestinal symptoms are more frequent, whereas cardiovascular symptoms are rare. Gastrointestinal symptoms include abdominal discomfort and vomiting. Gastrointestinal features are associated with cardiovascular rather than respiratory manifestations. The cutaneous features of pruritus, erythema, urticaria and angio-oedema occur in nearly all children. These are commonly the first symptoms experienced, occurring within minutes following allergen exposure. Life-threatening symptoms and signs include loss of consciousness, syncope, dizziness, light-headedness, cerebral dysfunction, hypotension, hypoxaemia, stridor, cyanosis and laryngeal oedema.

Table 22.5.1 lists the frequency of the presenting symptoms and signs in children admitted to hospital for anaphylaxis.

In one series of adults and children, 38% of severe reactions (defined by either hypoxaemia or hypotension) received multiple doses of adrenaline (epinephrine) and 23% of severe reactions were noted to have a delayed deterioration where symptoms worsened after an initial improvement.[7] Many such cases reflect declining plasma adrenaline concentrations after initial treatment in the face of an ongoing reaction, but some are true 'biphasic' reactions. Biphasic anaphylactic reactions are defined as worsening of symptoms, requiring new therapy, after the resolution of anaphylaxis, and are reported to occur in 3–20% of reactions.[7,9,10] Larger studies that include all-comers report the lowest incidences of biphasic reactions. Biphasic reactions usually occur 4–10 hours after the initial event; however, it has been described up to 48 hours later. Biphasic reactions are not accurately predicted from the initial clinical features, but the more severe the initial anaphylactic event, the more likely a delayed deterioration or a biphasic reaction will occur.[7,10]

Table 22.5.1

Presenting features of children with anaphylaxis[5]

Presenting feature	Per cent (%)
Cutaneous (urticaria, angio-oedema, flushing, warmth)	90
Upper airway (throat tightness or itchiness, drooling, stridor, oropharyngeal swelling)	80
Lower airway (chest tightness, wheezing)	60
Gastrointestinal (abdominal discomfort, vomiting)	40
Cardiovascular (arrhythmias, hypotension, poor capillary refill, weak pulses)	30
Neurological (confusion, decreased conscious state)	25
Generalised (diaphoresis, tingling, an impending sense of doom)	15

Investigations

Anaphylaxis is a clinical diagnosis, and investigations do not have a role in the acute management. On occasions, it may be difficult to differentiate anaphylaxis from other cardiac, respiratory, or neurological episodes. In this situation, determination of plasma levels of mast cell mediators (histamine and mast cell tryptase) has the potential to provide some diagnostic help, but only mast cell tryptase is available outside the research setting.

Mast cell tryptase (MCT) occurs in an alpha form that is constitutively released and a beta form that is released only following mast cell activation. Peak levels of beta-tryptase occur at 1–2 hours and decline with a half-life of approximately 2 hours. If blood is collected in the initial 30 minutes after a reaction, tryptase elevation may not be detected. Tryptase is stable and can be identified in plasma or serum stored at room temperature for several days. The commercial assay measures total MCT, comprising both alpha and beta forms and thus a single measurement of a high concentration may be constitutive or due to underlying mastocytosis (itself a risk factor for anaphylaxis), rather than due to a reaction. Likewise, significant fluctuations in MCT may occur within the normal range and will be missed if a single measurement is taken. Therefore, serial measurement of MCT (for example, on arrival, 1 hour later and the next day) has been suggested, but a number of uncertainties in this approach remain.[12] The reliability of measuring mast cell tryptase post mortem has been questioned, because elevation of tryptase may be seen in control cases where death has occurred from other causes.

The investigation of allergic triggers requires referral to a consultant allergist for performance of appropriate skin prick and blood tests to determine the presence of specific IgE antibodies. Serum allergen specific IgE levels

determined via UniCAP®, above which patients have a >95% chance of having an immediate IgE-mediated reaction, have been determined for some foods (e.g. cow's milk, egg, peanut, wheat).[13] It is not possible to predict the severity of a future allergic reaction based on the skin prick test size or allergen specific IgE levels.

Treatment

Adrenaline

Adrenaline should be given via the intramuscular rather than the subcutaneous route, due to better absorption from muscle. In children, peak adrenaline levels are reached 8 minutes after intramuscular and 34 minutes after subcutaneous injection. Peak levels are 20% higher after intramuscular injection. The preferred injection site for intramuscular administration is the upper outer side of the thigh, which gives significantly better absorption as compared with the deltoid muscle:[14]

- The dose of adrenaline is 0.01 mL kg^{-1} of 1 in 1000 intramuscular injection. Improvement should be seen within minutes. The dose should be repeated after 5–15 minutes if the effect is incomplete. Approximately one-third of patients will require more than one dose of adrenaline. Although the elimination half-life of adrenaline is very short, an intramuscular bolus will provide a depot for continued absorption and therapeutic blood levels may be maintained for an hour or more.
- In situations where there is severe circulatory compromise, adrenaline will be poorly absorbed from muscle. Therefore, after the initial intramuscular dose an intravenous infusion may be required.
- Intravenous adrenaline should be given by infusion with an infusion pump per hospital protocol. Start at 0.1 mcg kg^{-1} per minute and titrate to response up to a maximum of 1 mcg kg^{-1} per minute. One easy-to-remember approach is to dilute 1 mg in 100 mL and start at 0.5–1 mL/kg/h.[6]
- The elimination half-life of adrenaline is 2–3 minutes, so a steady state will be achieved within 10–15 minutes of any infusion rate change. Beware that as the reaction resolves, a previously therapeutic infusion rate will rapidly become toxic.

- Do not give intravenous bolus doses of adrenaline unless the patient is in cardiac arrest.
- For cardiac arrest, the initial intravenous dose is 10 mcg (0.01 mg) per kg body weight of a 1 in 10,000 (wt/vol) dilution. Subsequent doses are 100 mcg kg^{-1} every 3–5 minutes and, if still refractory, the dose may be increased to 200 mcg kg^{-1}. This is higher than standard cardiac arrest doses, recognising that refractory anaphylaxis may only respond to very high doses of adrenaline.
- Adverse effects of adrenaline include transient pallor, tremor, anxiety, palpitations, cardiac arrhythmias, headache and nausea. As a general rule, tachycardia, anxiety, pallor or tremor in the setting of normal or elevated blood pressure should be attributed to adrenaline toxicity, not anaphylaxis.

Airway and breathing

- Give high-flow oxygen by mask.
- For bronchospasm, consider continuous nebulised salbutamol (0.5%) if there is no response to adrenaline.
- Nebulised adrenaline 0.5 mL of 1% may be added for upper airway obstruction that does not respond to initial parenteral adrenaline.
- Consider intubation in cardiac arrest and if upper airway obstruction or bronchospasm is severe and does not respond adequately to initial management.

Circulation

- Achieve intravascular access with large-bore cannula.
- Treat hypotension with normal saline 20 mL kg^{-1}.
- If hypotension continues, give further colloid boluses of 10–20 mL kg^{-1} along with an adrenaline infusion.
- In unrelenting hypotension due to profound vasodilation and/or cardiac (pump) failure, selective vasopressors, such as metaraminol or vasopressin, and/or mechanical circulatory support (intra-aortic balloon pump), have been described in case reports to be effective for cases of severe anaphylaxis in adults.

Supplemental treatment

- The rationales for supplementary treatment with steroid and antihistamines are less well defined. Antihistamines are unlikely to be of any value for treating life-threatening manifestations because of the many different mediators involved in anaphylaxis.
- Because of the self-limited nature of most anaphylactic reactions, steroids are unlikely to be of any benefit. They may be used for severe bronchospastic reactions that are slow to respond to treatment, e.g. methylprednisolone 1 mg kg^{-1} intravenously.
- Antihistamine: consider promethazine 1.0 mg kg^{-1} per dose (maximum 25 mg) orally for symptomatic relief of urticaria. Do not give parenterally as this may precipitate hypotension.[15]

Duration of treatment

Duration of treatment is determined by clinical response. Repeated doses of adrenaline are often required.

Admission/observation

Admit or observe for at least 4 hours after the last dose of adrenaline, extended to 10 hours or overnight for all patients with severe anaphylaxis; a biphasic reaction with deterioration may occur following the initial episode and is both more common and more likely to require treatment with adrenaline in patients with initially severe reactions.[7]

The role of H_1 and H_2 blockers

The role of antihistamines is unclear. Addition of H_2 to H_1 antagonists produces no differences in blood pressure and symptoms, but may be associated with some improvement in urticaria at 2 hours. Antihistamines are not a substitute for adrenaline.

Patients on beta-blockers

Glucagon 0.02 mg kg^{-1} intravenously has been used in an attempt to reverse beta

blockade.

Diagnosis

It is important to determine the cause of the anaphylactic reaction whenever possible.

Types of anaphylaxis

Common

Foods

Severe life-threatening reactions are predominantly due to peanut and tree nuts. In pre-school-age children, they may be due to egg and cow's milk proteins.

Drugs

Many reactions, in particular those due to antibiotics, are IgE-mediated. Non–IgE-mediated anaphylactoid reactions are clinically indistinguishable from anaphylaxis. Drugs that can cause anaphylactoid reactions include opiates, muscle relaxants, radiocontrast media, non-steroidal anti-inflammatory drugs and quinolone and vancomycin antibiotics.

Stings

Stings are most commonly due to bees and less often wasps. In Australia, jumper ant and bulldog ant (*Myrmecia* spp.) stings are a common cause of anaphylaxis in endemic areas.

Latex

Sensitisation occurs particularly in children with multiple exposures to latex-containing items during medical procedures. Children with spina bifida are particularly at risk due to multiple exposures following surgery and urinary tract catheterisation.

Less common

Idiopathic

No triggers are identified, despite full investigation. Cases may present with

laryngeal oedema as the only manifestation. The episodes can usually be controlled by regular antihistamine and, if necessary, the addition of alternate day steroids.[16] Psychogenic anaphylaxis has been classified as a variant of idiopathic anaphylaxis and should be considered in the differential diagnosis.

Uncommon

Exercise-induced

Symptoms of urticaria, angio-oedema and stridor plus or minus hypotension develop during or soon after cessation of vigorous exercise.

Food-dependent exercise-induced

In this situation, exercise induces symptoms only following ingestion of the relevant foodstuff, which has included wheat, celery, shellfish, oranges and peaches. In some but not all cases, IgE sensitisation to the relevant food can be demonstrated.

Recurrent anaphylaxis

Anaphylaxis is frequently multiple and in one series two-thirds of patients had three or more anaphylactic episodes. Efforts should be made to identify unrecognised triggers. In children, this is often due to foods (generally peanut or tree nut products) contained in manufactured or processed foods. Some cases without an identifiable cause are due to idiopathic anaphylaxis.

Differential diagnosis

Anaphylaxis should be distinguished from other presentations that may cause confusion. These include:

- acute asthma
- vasovagal syncope
- urticaria or angio-oedema
- psychogenic stridor
- cardiovascular events
- seizure disorders
- mast cell mediator release in mastocytosis

- hereditary angio-oedema.

Prevention

Advice concerning the avoidance of the triggering allergen is critical. This will usually require referral to a consultant allergist.

The provision of self-injectable or carer-administered adrenaline should be considered for all children with anaphylaxis. Inadvertent re-exposure is most likely in the case of insect stings and foods, and least likely for drugs. The prescription of a self-injectable adrenaline also requires instruction in the indications for and demonstration of use and the provision of a clear and simple written anaphylaxis action plan.

Self-injectable adrenaline is available in two fixed-dosages: (0.15 mg of adrenaline) for children 15–30 kg and (0.3 mg of adrenaline) for children greater than 30 kg. The American Academy of Allergy, Asthma and Immunology recommends the 0.3 mg dose for children >20 kg.[17]

In venom-induced anaphylaxis referral to an allergist for desensitisation should be considered for life-threatening reactions with respiratory or cardiovascular involvement if there is an appropriate reagent available, such as honey bee (*Apis mellifera*), paper wasp (*Polistes* spp.), European wasp (*Vespula* spp.), fire ant (*Solenopsis* spp.) and the Australian jack jumper ant (*Myrmecia pilosula*). This will involve a series of injections with venom for a duration of 3 or more years. In general, venom desensitisation is not recommended for children with generalised cutaneous reactions in the absence of respiratory or cardiovascular involvement, because in such cases the natural history is usually for reactivity to become less serious or negligible over time.[18]

Controversies

Currently, there are no clear guidelines on which children should be prescribed an EpiPen. The great majority of fatalities are recorded in children over 5 years of age, despite the fact that food-allergic reactions are more common in pre-school children and frequently lessen with time. As the prescription of an EpiPen is primarily concerned with risk management, it is necessary to consider the factors that point to the likelihood of developing a severe life-threatening reaction.[19] These are:

- age over 5 years

- a history of respiratory tract involvement with the initial or subsequent reactions
- a history of asthma requiring preventive medication
- peanut or tree nut sensitivity
- reactions induced by traces or small amounts of allergen
- a strongly positive skin prick test (>8 mm).

Each factor should be considered, and the greater the number that are positive, the lower the threshold for prescribing an EpiPen. In addition, these factors need to be weighed in the light of the parental wishes and environmental circumstances. Providing the parents with a rational perspective on the remote risk of death is essential.

Future Directions and Research

1. Humanised monoclonal anti-IgE antibodies increase the threshold dose of food required to trigger symptoms in food-induced anaphylaxis. Regular administration is required. Unlike desensitisation, the treatment is not allergen-specific, and therefore offers promise to individuals with life-threatening reactions to multiple allergens.
2. Characterisation of the molecular structure of allergenic epitopes may allow the construction of peptides for immunotherapy that trigger T-cell responses without binding IgE, thus significantly reducing the risks and increasing the efficacy of immunotherapy.
3. The promotion of desensitisation or tolerance in food allergy by graded oral administration of food allergens such as peanut or egg.

Acknowledgement

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SECTION 23

Ultrasound

OUTLINE

23.1

Ultrasound

Adam Bystrzycki

ESSENTIALS

Point-of-care ultrasound:

1 No ionising radiation

2 Ultrasound performed by clinicians at the bedside

3 Focused – it answers a specific clinical question

4 Can alter clinical decisions and direct therapies

5 Used to guide procedures in order to increase safety and/or effectiveness

6 Can test the effectiveness of clinical interventions.

The Australasian College for Emergency Medicine (ACEM) released policy document (P21) on point-of-care ultrasound (POCUS) in 1999.[1] The latest iteration of the policy states that ultrasound examination, interpretation and clinical correlation should be available in a timely manner 24 hours a day for emergency department (ED) patients; emergency physicians providing POCUS should be appropriately trained; and ACEM specifically supports the use of POCUS for:

> *…at least the following:*
>
> *-traumatic haemoperitoneum/haemothorax/pneumothorax*
>
> *-abdominal aortic aneurysm*

-pericardial fluid

-intra-uterine pregnancy identification

-vascular access and other procedures

-basic echocardiography in life support

-hydronephrosis and biliary tract disease

-soft tissue studies

-deep vein thrombosis…

and it:

> *…encourages all Emergency Physicians to be competent in the "core" areas of emergency ultrasound, being abdominal aortic aneurysm, EFAST, procedural guidance and echo in life support.*[1]

Several of the above clinical indications have direct relevance to paediatric emergency medicine (PEM).

The applicability of POCUS to the practice of PEM has been further bolstered by the recently released policy statement of the American Academy of Paediatrics.[2] This document exhorts PEM physicians to be familiar with the definition and application of POCUS and its utility for emergency patients.

A POCUS exam is focused and specific. Unlike a diagnostic study, which aims to image an entire body region or entire organ in a systematic manner, the POCUS exam is constrained in its approach and aims to answer a specific clinical question. Examples of this approach might include the following:

1. In this shocked patient, what is the contractility of the left ventricle?
2. In this multitrauma patient, is intraabdominal bleeding the cause of his/her tachycardia? (Figs 23.1.1–23.1.3)
3. In this septic patient, should I infuse another bolus of fluid?
4. In this dyspnoeic patient, is there ultrasound evidence of a pneumothorax or haemothorax? (Figs 23.1.4 and 23.1.5)

In each of these examples, it should be noted that the POCUS exam is used to

‘rule in’ the suspected clinical diagnosis or test condition. The utility of POCUS as a ‘rule-in’ test, as well as its poor performance when used incorrectly to ‘rule out’ conditions, has been best studied in the case of the extended focused assessment with sonography for trauma (EFAST) examination. In the haemodynamically stable blunt trauma patient, EFAST has an overall sensitivity of 43%, specificity of 99%, and accuracy of 94.1%.[3] In cases of penetrating trauma EFAST performs worse, with prevalence of a positive FAST exam after penetrating trauma fairly low, ranging from 24.2% to 56.3%. The FAST exam for penetrating trauma is a highly specific (94.1–100.0%), but not very sensitive (28.1–100%), diagnostic modality. Nevertheless, this is still much better than EFAST’s performance once organ pathology is examined with FAST, which is reported to have a sensitivity of 42% for blunt abdominal injury that can be found on CT.[4]

Ultrasound in PEM is also useful for guiding procedures. The use of ultrasound when performing procedures such as nerve blocks and central venous access device insertion improves success and reduces complication rates.[5–8] It also increases the rate of cannulation success in children assessed as having difficult veins.[9]

Ultrasound has rapidly gained ground as an important tool during resuscitation, and the severely ill or injured child is no exception. As a ‘core’ technique, Echo in Life Support (ELS) aids rapid assessment of cardiac function and detection of potentially reversible causes of haemodynamic deterioration or cardiac arrest in a non-shockable rhythm.[10,11] ELS allows differentiation of cardiac standstill (true-PEA or asystole) from pseudo-PEA (presence of electrical activity with no detectable pulse, but presence of echocardiographic organised mechanical activity). The former has a much worse prognosis than the latter.[12]

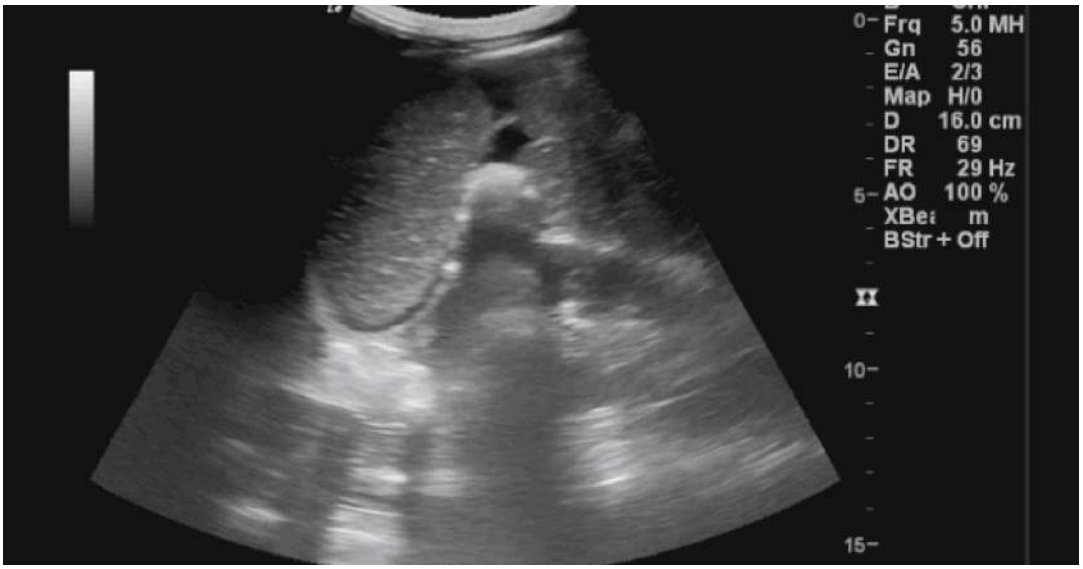

FIG. 23.1.1 - LUQ view; perisplenic free fluid.

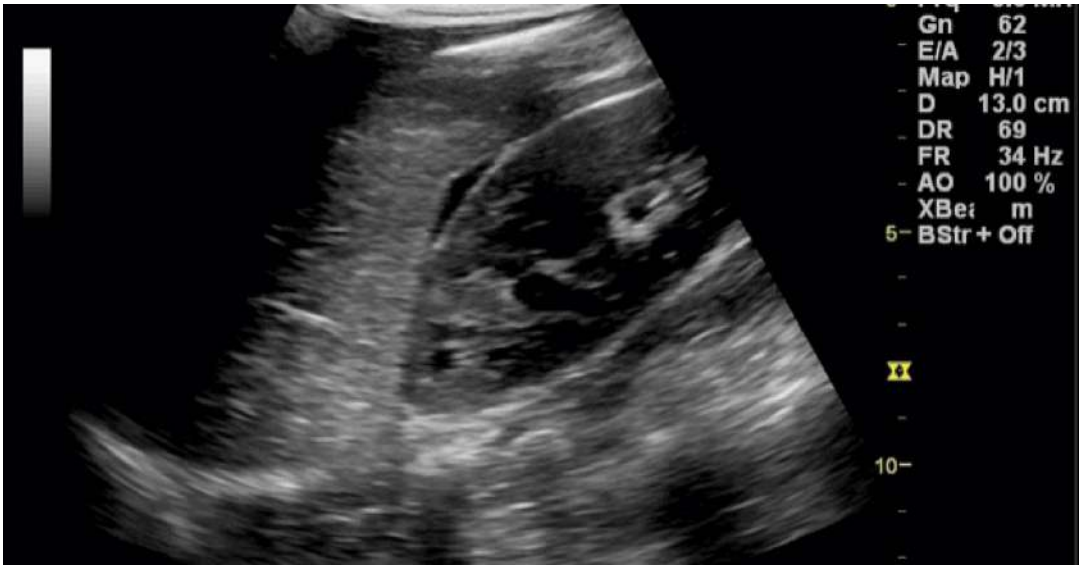

FIG. 23.1.2 - RUQ view; small amount of fluid in Morrison's Pouch.

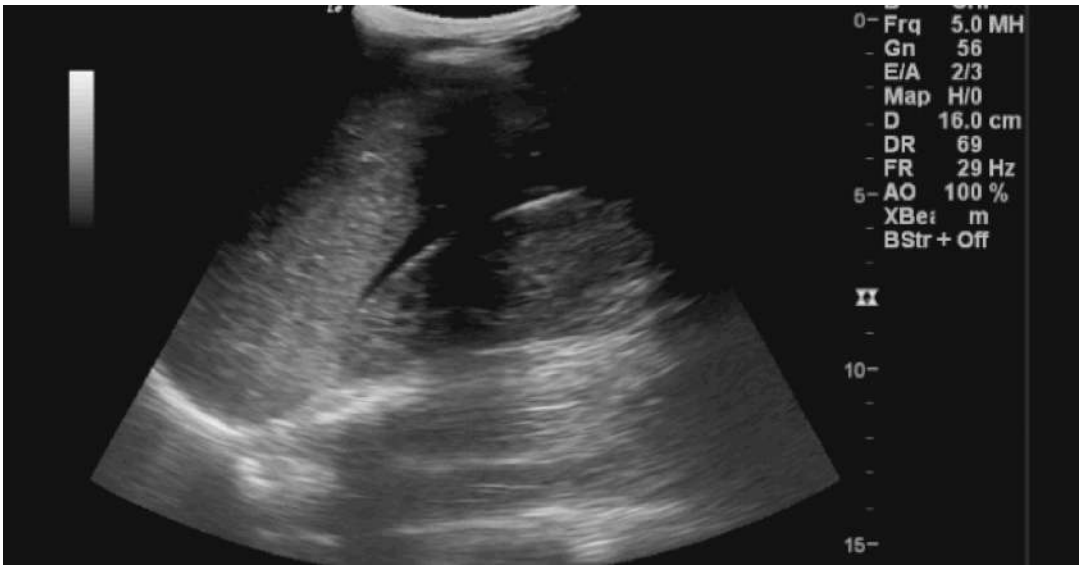

FIG. 23.1.3 - RUQ view; larger amount of free fluid.

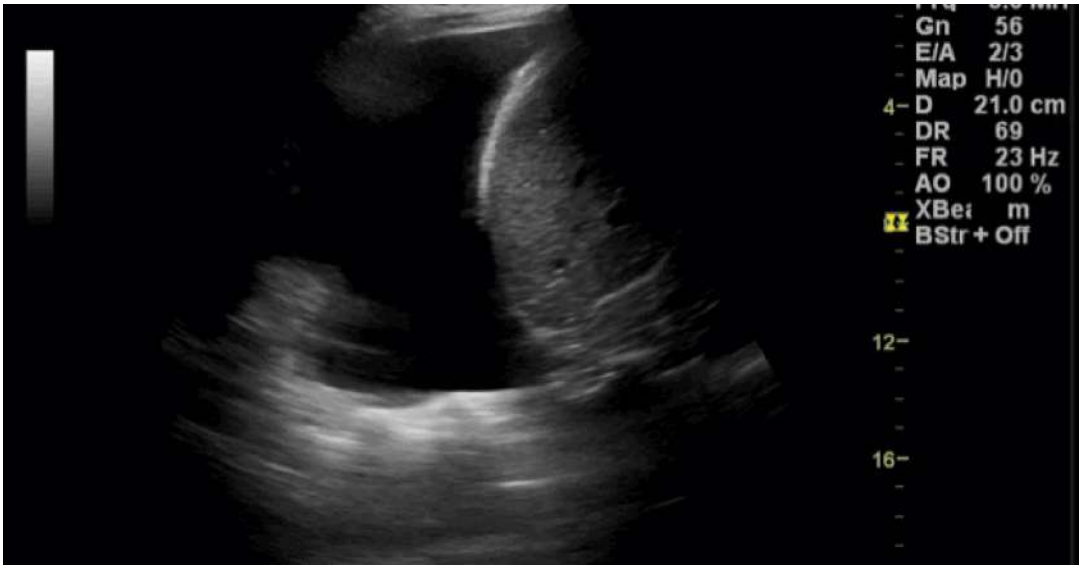

FIG. 23.1.4 - RUQ supradiaphragmatic view showing large amount of pleural fluid.

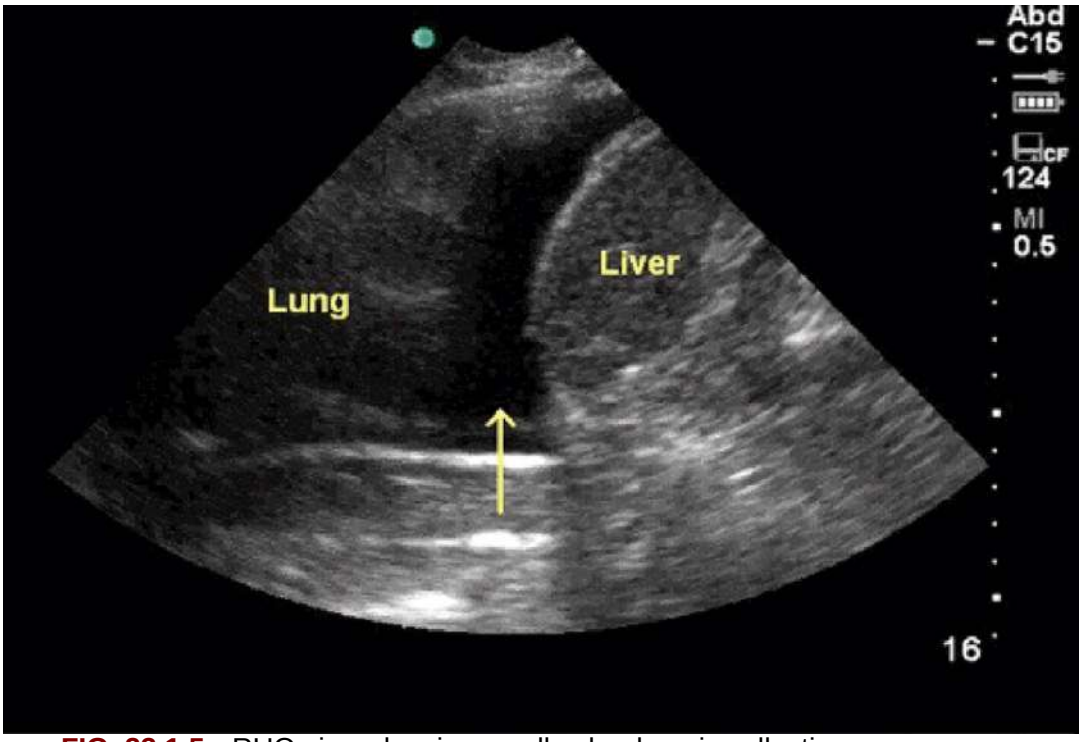

FIG. 23.1.5 RUQ view showing small subpulmonic collection.

Regardless of the reason for performing ultrasound at the bedside, it is imperative that it be done in a safe and consistent manner. Any ultrasound examination which may result in a treatment decision should be performed by a suitably trained and credentialed practitioner. This ensures performance at, and maintenance of, a minimum standard.[13] Credentialing should be specific to the study type and should expose the trainee to normal and abnormal sonographic findings.[14] While credentialing is the responsibility of the Health Service, ACEM has described the phases for an ultrasound credentialing programme[15,16]:

1. Introductory teaching phase covering theory and practice
2. Period of skill acquisition: a minimum number of scans is stipulated in patients with relevant indications. A minimum number of ‘abnormal’ or positive scans must be performed.
3. A summative assessment requiring the candidate to perform and document an examination under direct supervision.

To achieve these requirements it may be necessary for some EDs to seek the assistance of external education providers or other suitably qualified practitioners within the health service.

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23.2

Diagnostic ultrasound in paediatric emergency medicine

Adam O'Brien

ESSENTIALS

1 Diagnostic point-of-care ultrasound should enhance diagnostic decision making and patient flow.

2 Some adult diagnostic point-of-care ultrasound applications are directly transferrable to paediatrics.

3 Paediatric specific diagnostic applications of point-of-care ultrasound include intussusception, pyloric stenosis and constipation.

Many diagnostic point-of-care ultrasound (POCUS) applications used in adults can be transferred to paediatric emergency medicine (PEM), including those relating to the biliary tract, kidneys, venous thrombosis, ocular, soft tissue and first trimester pregnancy. Over the last decade there has been an increasing use of PEM POCUS with a myriad of established and emerging diagnostic applications being used. Internationally, PEM POCUS is being incorporated into diagnostic pathways. For example, the child with respiratory distress can have the number of possible diagnoses rapidly narrowed with the use of lung ultrasound at the bedside.[1]

As discussed in Chapter 23.1, when point-of-care ultrasound is used to 'rule in' and not to 'rule out', errors relating to missed diagnoses will be minimised. Answering binary clinical questions is the domain of POCUS; for example, 'Is there sonographic evidence of intussusception?' This is in contrast to comprehensive ultrasonography performed in medical imaging departments that

provide clinicians with detailed and systematic regional imaging.

Characteristics of POCUS that make it ideal for the PEM practitioner are that it is goal directed, problem oriented, limited in scope, reveals qualitative or semi-quantitative information and is time sensitive and repeatable.[2]

POCUS requires training and experience to be able to integrate the findings with the clinical history, examination and other investigations, as opposed to relying on POCUS to provide all answers. This is particularly important when used in children who may have diagnoses that have harmful effects when they progress, such as with appendicitis, intussusception and haemoperitoneum.

The haemodynamic assessment and management of a **hypotensive** patient can be enhanced by imaging the heart, inferior vena cava (IVC), lungs and bladder with POCUS. For example, if the heart is hyperdynamic, the IVC collapses with respiration, the lungs are non-oedematous, and the bladder empty, it is likely that the systolic blood pressure and tissue perfusion will be improved with a fluid bolus.

Lung ultrasound has many applications and may be one of the diagnostic POCUS applications with the most utility in PEM. As in adults, it reliably detects pneumothoraces and pleural effusions. The diagnosis of pneumonia in both adults and children has a sensitivity and specificity of 85% and 90% respectively. Some early research suggests that lung POCUS in children may be able to aid in differentiating viral from bacterial infections by measuring the depth of consolidations; when it is less than 1 cm it is suggestive of a viral infection. Other research has raised the possibility of POCUS being able to assist with predicting which infants with bronchiolitis will progress to require oxygen therapy.[3]

In the child with undifferentiated severe **respiratory distress**, POCUS can identify pneumothorax, pleural effusions, pneumonia and interstitial oedema (usually fluid overload or viral infection). A-lines in combination with lung sliding indicate normal lung at the site where POCUS is being performed. Lung sliding excludes a pneumothorax, non-ventilation and pleural fluid. A-lines are replaced with B-lines when there is interstitial oedema. In the child with pneumonia, A-lines are replaced by hypoechoic alveolar consolidation that may contain echogenic sonographic air bronchograms.[4]

Abdominal pain is a common paediatric complaint. POCUS may elucidate the cause of the pain earlier in the diagnostic workup as it has the ability to visualise bowel motility, free fluid, rectal diameter, gross organ abnormalities such as hydronephrosis, and specific findings such as intussusception.

Acute **appendicitis** is a common paediatric surgical emergency. Several POCUS studies have shown sensitivities for detecting appendicitis to be between 60% to 80% with a specificity of 90%. The normal appendix is notoriously difficult to find but an abnormal appendix draws attention to itself as its diameter is more than 6 mm, it is blind ending, non-peristaltic and non-compressible. Secondary features of acute appendicitis include the presence of an appendicolith, its attachment to the caecum, surrounding echogenic inflammatory fatty tissue or free fluid, and hyperaemia. Perforated appendicitis, however, is highly suspicious when there are dilated bowel loops, echogenic fat and complex fluid collections.[5]

Intussusception is another surgical emergency requiring timely diagnosis. POCUS novices have learnt to diagnose intussusception with ultrasound, after which there was a diagnostic sensitivity of 85%, compared to almost 100% in comprehensive ultrasonography performed in medical imaging departments.[6] A target lesion with concentric layers of colon surrounding the invaginating ileum is the pathognomonic sonographic finding of ileocolic intussusception (see Fig. 23.2.1).

In the paediatric patient with **blunt abdominal trauma** the eFAST performs less well compared with adults, with reported sensitivities for detecting intraabdominal injuries of approximately 50%.[6] However, a positive eFAST has excellent specificity and may assist in the choice and urgency of subsequent imaging and consultations. Because of the limitations of eFAST, including its lack of information about solid organ and bowel injuries, and the sensitivity threshold of 200 to 250 mL, it is important to remember that in childhood trauma, a normal eFAST examination is not a screening test and should not influence any decisions made on clinically acquired information.

Constipation is a common cause of paediatric abdominal pain. Finding a rectal diameter of greater than 3 cm with POCUS in a child without the urge to defaecate is indicative of constipation[1] and can replace the digital examination.

Fractures of all types can be detected with POCUS, seen as a breach or step in the bone cortex. The use of ultrasound for this purpose reduces the need for ionising X-rays.

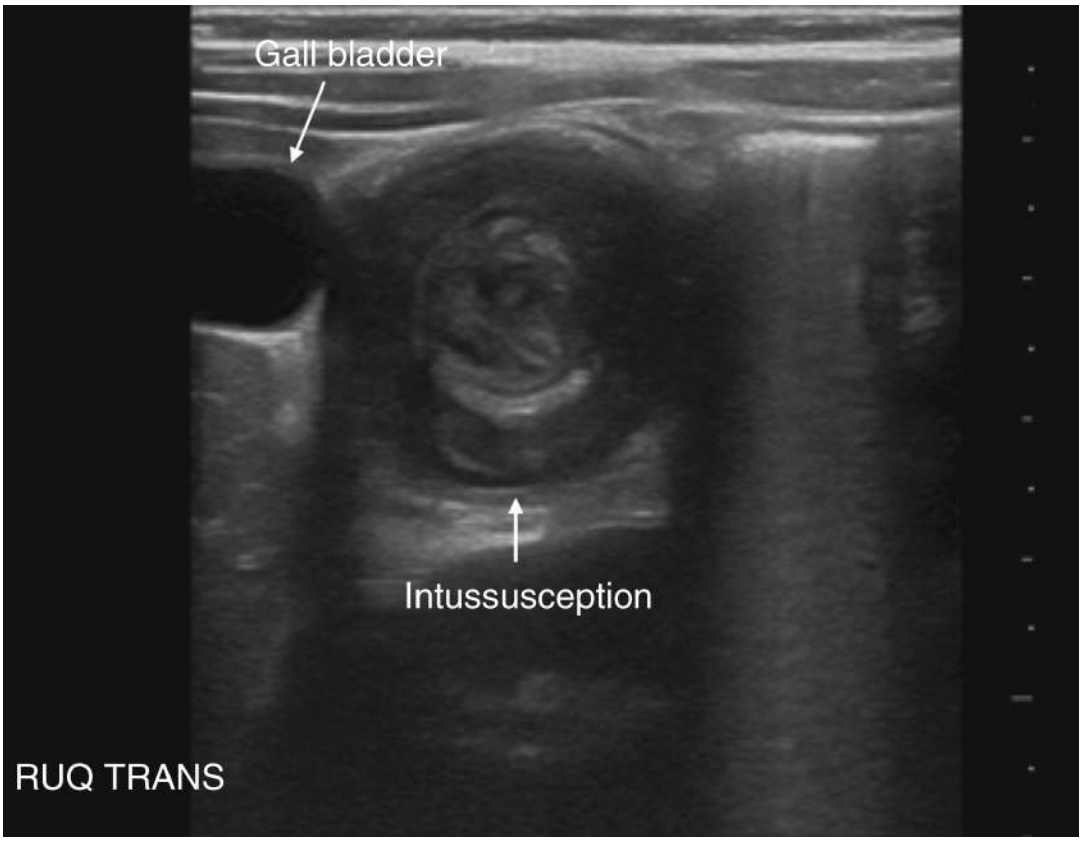

FIG. 23.2.1 Sonographic finding of ileocolic intussusception.

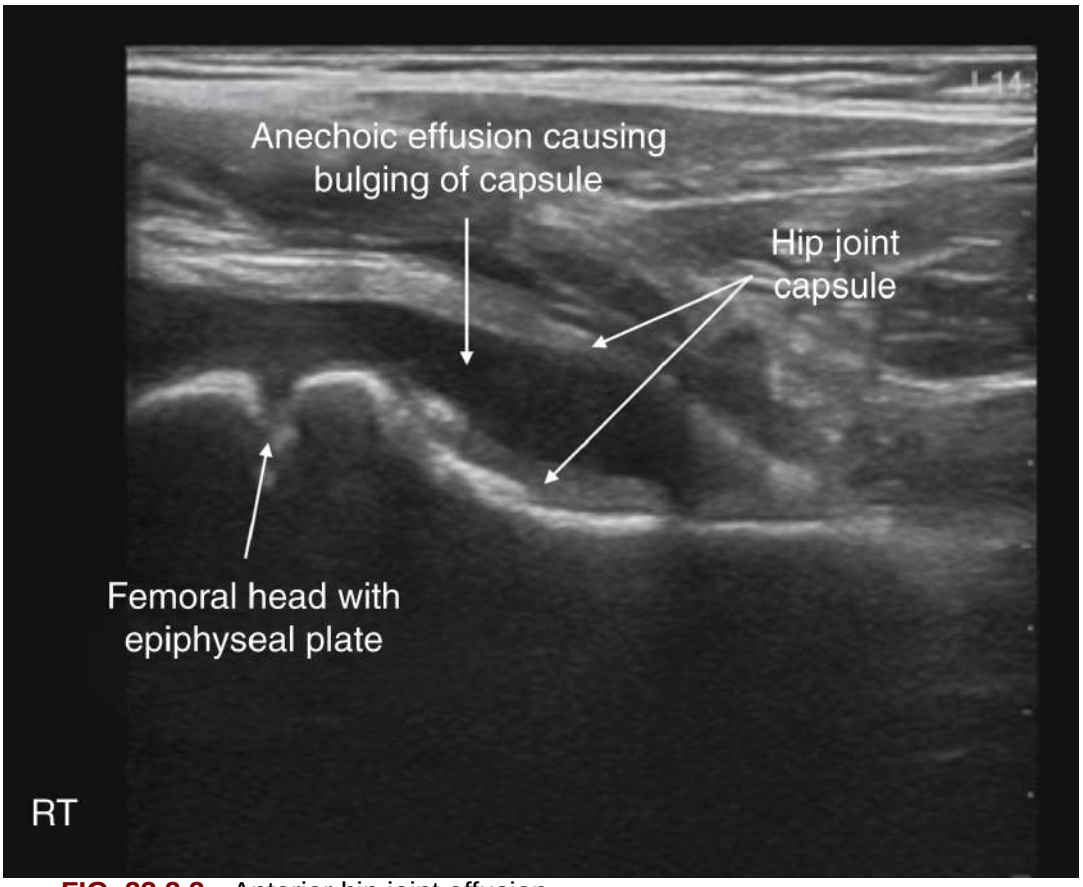

FIG. 23.2.2 Anterior hip joint effusion.

It is often difficult to clinically detect whether there is a fracture underlying a scalp haematoma in children with closed head injuries. POCUS can detect **skull fractures,**[7] with care needed to image beyond the margins of the haematoma to increase sensitivity and to differentiate fractures from normal suture lines. If a fracture is detected in a child with symptoms suspicious of intracranial injury, CT of the brain will be required to exclude significant brain injury or bleeding.[8]

Long bone fractures, particularly forearm fractures, can be identified with POCUS. Furthermore, ultrasound can be used to guide fracture reduction, once again reducing the child's exposure to procedural fluoroscopy.[6]

A young child with a **limp** has a number of possible causes for their presentation, ranging from strained ankle ligaments to a paraspinal tumour. One of the more common causes is transient synovitis of the hip joint, which can be diagnosed and managed on clinical grounds alone. In children in whom it is difficult to localise from where the limp originates, POCUS of the hip joint might demonstrate an anterior joint effusion (see Fig. 23.2.2). The sonographic appearance of a hip joint effusion will not differentiate between transient

synovitis and septic arthritis;[1] this must be done clinically with history, examination and sometimes blood tests.

There are many diagnostic applications for POCUS in paediatric emergency medicine, some of which are evidence based and others that are emerging. Indeed, there are many research papers concluding that POCUS in PEM can be performed with good sensitivities and specificities. The challenge for emergency physicians, once they have acquired the necessary kinaesthetic and image recognitions skills, is to integrate POCUS into clinical pathways that enhance the diagnosis and management of our paediatric patients.

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23.3

Ultrasound guidance for procedures

Adam O'Brien

ESSENTIALS

1 Ultrasound guidance during invasive procedures improves accuracy and complication rates.

2 Acquisition of the skills necessary for ultrasound guidance requires training and practice.

Ultrasound guidance for procedures

Using ultrasound to guide invasive procedures improves accuracy, patient safety and time taken to completion[1] compared with traditional landmark techniques. However, learning to effectively use ultrasound to guide procedures requires persistence to develop the fine motor coordination required to do it safely.

There are many procedures in adult emergency medicine that are enhanced by ultrasound guidance. Some are transferrable to paediatrics including:

- Foreign body identification and removal:
    - Glass and metal are radiopaque and are better detected on plain radiography. However, when the foreign body (FB) is a radiolucent material such as wood, plastic or another vegetable matter, ultrasound can be used as a screening test with moderate sensitivity.[2] Once a FB is detected with ultrasound its removal should be guided with ultrasound, for example, by directing a needle towards the FB to indicating the incision site.[3] The FB's position should be regularly

reassessed during its removal as its position can easily change.

- Joint aspiration:
    - Ultrasound is superior to clinical examination for detecting the presence or absence of joint effusions.[4] When there is clinical suspicion of a septic joint, paediatric patients are more likely to have a joint effusion aspirated in the operating theatre, enabling the joint to be washed out during the same procedure.
- Lumbar puncture:
    - The use of ultrasound to mark the site for lumbar puncture significantly increases the clinician's confidence in selecting the insertion site.[5] Visualisation of the anatomical structures of the spine, including the depth of the subarachnoid space and the distal end of the spinal cord (conus medellaris), ensures the lumbar puncture is performed at a safe intervertebral level[6] (see Fig. 23.3.1).
- Nerve blocks:
    - In the setting of trauma, ultrasound-guided nerve blockade reduces the need for sedating analgesics in the paediatric patient. Ultrasound guidance reduces the time to effective anaesthesia, the anaesthetic volume required, and increased blockade efficacy compared with landmark techniques.[6] The femoral nerve block is the most commonly used regional nerve block in children of all ages for isolated femoral fractures.
    - Other useful nerve blocks include:
        - mid-forearm ulna, median and radial nerve blocks for metacarpal fracture manipulations and other hand injuries[7]
        - the posterior tibial nerve block for procedures involving the plantar surface of the foot is particularly useful in children compared to the painful alternative of local anaesthetic injections[8]
        - dorsal penile nerve blocks to minimise pain during reduction of a paraphimosis reduction.[9]

Vascular access

Vascular access in children, whether peripheral or central, intravenous or intra-arterial, is often essential for their care and resuscitation. Children, however, in contrast to adults, have smaller vessels, are generally less compliant and move during the procedure, making access more difficult. These factors also make ultrasound-guided vascular access more difficult to learn in children but, once mastered, results in greater success rates, fewer skin punctures, reduced time to placement and fewer complications:

- Peripheral:
    - Ultrasound-guided peripheral vascular access in the younger mobile child is only possible using the out-of-plane approach with the vessel in short axis. The in-plane approach with the vessel in long axis can be used in the older, more cooperative child. While using the out-of-plane approach the key skill that must be learnt is to be constantly aware of the needle tip's position. Failure to do so will result in complications related to puncturing deeper anatomical structures (see Fig. 23.3.2).
- Central:
    - Central venous access should be performed with ultrasound guidance in both adults and children to improve success rates and improve patient safety.[1,10] Central venous access in children is commonly achieved using the femoral vein where ultrasound will detect anatomical variations, including when the femoral artery overlies the vein. The success of subclavian[10] and internal jugular venous access is greatly improved if ultrasound guidance is used. Once venous access has been achieved the catheter's position in the central vein can be confirmed by injecting agitated saline and observing gas bubbles entering the right side of the heart with a subcostal cardiac view.[11] This is particularly useful in children as there is a short transit time for the agitated saline to travel from the catheter to the heart.

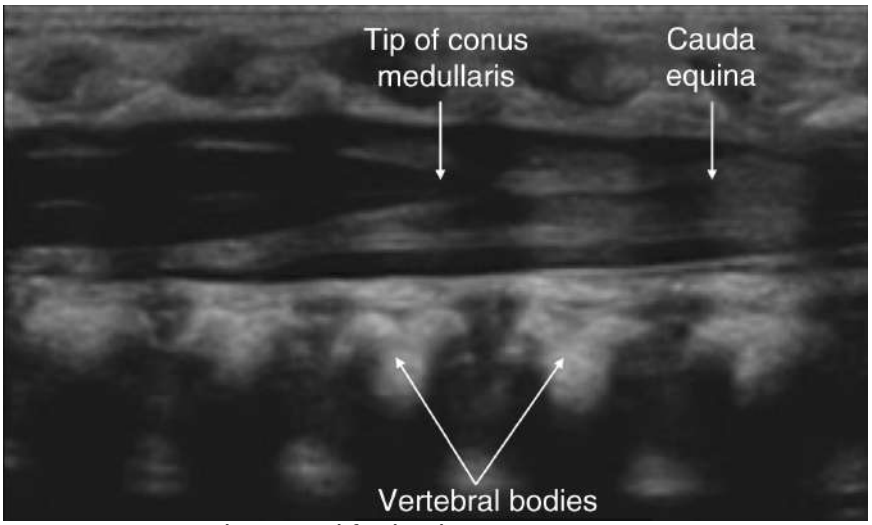

FIG. 23.3.1 Ultrasound for lumbar puncture.

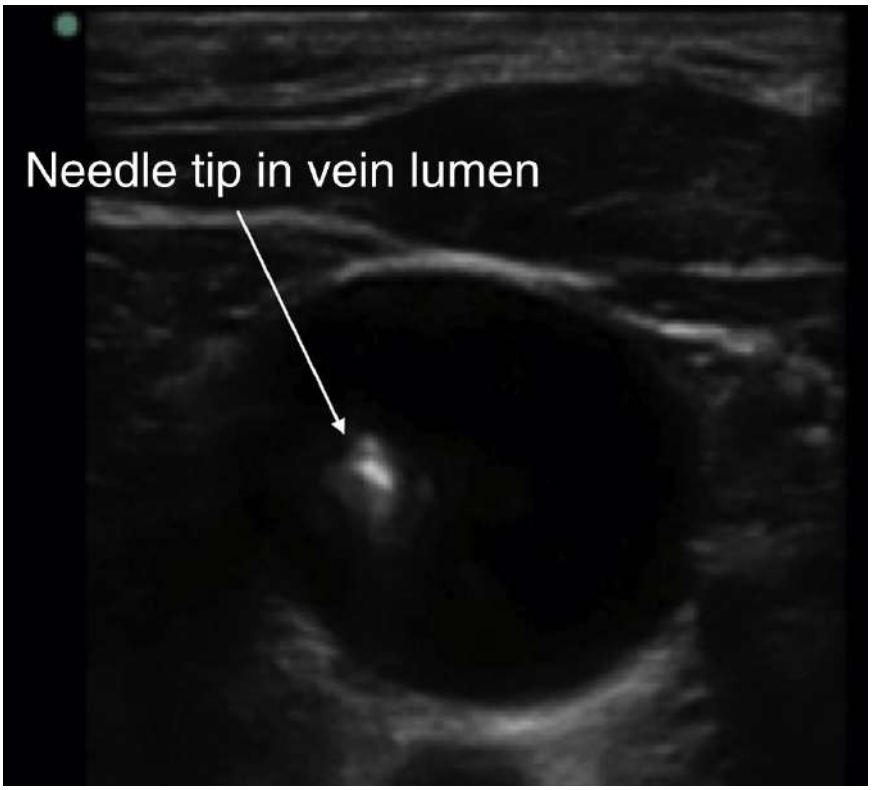

FIG. 23.3.2 Needle tip position on ultrasound.

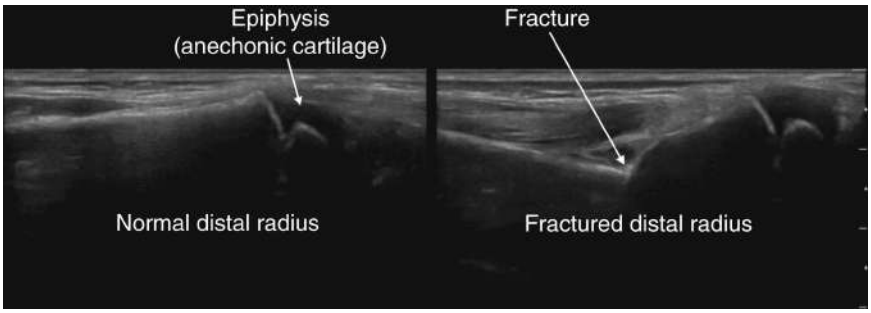

FIG. 23.3.3 Ultrasound for fracture reduction.

- Intraosseous:
    - In the severely unwell child, vascular access may not be possible and time should not be wasted attempting to achieve it. Intraosseous needle insertion must be utilised in such situations. Its correct intramedullary placement can be confirmed by observing Doppler flow within the bone medulla while flushing isotonic fluid through the needle.[12] If incorrectly placed, flow will be seen outside the bone cortex.

Aspiration

Suprapubic aspiration is the most sterile method of collecting urine for microbiological analysis. Ultrasound can be used to guide the needle into the bladder with variable success rates, with the relatively slow needle advancement often resulting in the bladder wall being indented rather than being punctured. If, however, the bladder dimensions are measured prior to aspiration and the transverse diameter of the bladder is found to be more than 2.5 cm, the success rate of ‘blind’ suprapubic aspiration of urine approaches 100%.[13] This latter technique allows swift needle advancement through the bladder wall.

Fracture reduction

Ultrasound can be used to diagnose fractures in children. It can also be used to guide the reduction of fractures, particularly distal radial fractures, and therefore increase confidence of a successful reduction (see Fig. 23.3.3). Although the use of ultrasound may not improve final patient outcomes, it can reduce the time taken for reduction and the need for radiographic imaging during the procedure.[14]

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23.4

Incorporating ultrasound into paediatric resuscitation

Robyn Brady

ESSENTIALS

Point-of-care ultrasound (POCUS) in critically ill infants and children shows promise for early diagnostic streaming in undifferentiated shock, respiratory distress and trauma as well as adding diagnostic and prognostic information in non-shockable cardiac arrest, and improving procedural accuracy and safety. As paediatric emergency medicine POCUS capabilities grow internationally, its role in paediatric resuscitation will be better defined.

Point-of-care ultrasound (POCUS) as a diagnostic tool can provide key real-time physiological and anatomical information for the emergency physician in a paediatric resuscitation, facilitating rapid diagnostic streaming at key junctions of clinico-therapeutic algorithms for respiratory distress, shock and cardiac arrest. Serial POCUS assessments of markers such as peritoneal free fluid and optic nerve sheath diameter add valuable data to our understanding of evolving pathophysiology and response to treatment.

In **undifferentiated severe respiratory distress**, POCUS can identify pneumothorax, pneumonia, pleural effusions and pulmonary oedema, with sensitivities greater than or equal to chest X-ray.[1,2] The key sonographic features in lung examination for these pathologies are outlined in Chapter 23.2. Haemothorax and pleural effusion are easily seen, can be qualitatively described

in relation to septation or loculation, and the localisation can guide drainage procedures.[3]

During intubation, transtracheal POCUS can be used in real time to observe the endotracheal tube (ETT) entering the trachea as opposed to the oesophagus with 95% sensitivity and specificity, these figures have been replicated in small number paediatric series. Checking for bilateral lung sliding can then ascertain within seconds whether the tube position is allowing bilateral ventilation (in the absence of pneumothorax or proximal obstruction) with far greater sensitivity than physical examination (95% vs 63%).[4] This schema is shown in Fig. 23.4.1. Bilateral lung sliding can also allow confirmation of continued endotracheal ventilation when, for example, CO_2 tracing is inadvertently lost. A 'lung pulse' demonstrating cardiac but not pleural movement, suggests contralateral main bronchus intubation.

Ultrasound guidance contributes to increased safety and efficacy of multiple other procedures during paediatric resuscitation, including the placement of peripheral and central venous access, nerve blocks and drainage catheters.

The use of POCUS in adult **cardiac arrest** has been paradigm-shifting in identifying that the presence of cardiac wall motion cuts across electrocardiographic categories of 'asystole' and 'pulseless electrical activity' (PEA) and better identifies both prognosis and possible causation/amelioration.[5,6] These studies demonstrate that there is cardiac wall motion in 10–30% of electrocardiographic asystole, and up to 70% of PEA, and that these distinctions relate strongly to the potential for survival. In Gaspari's multicentre study of 793 in and out of hospital cardiac arrests in which POCUS was used, the rates of return of spontaneous circulation (ROSC) and hospital discharge in those with cardiac wall motion were 51% and 3.8%, respectively, compared with 14% and 0.6% where no cardiac motion was observed. In Breitkreutz' single centre study, contributing or causative factors for PEA with cardiac motion detected were identified by POCUS in up to 80% of 38 cases of 'PEA arrest with cardiac wall motion'. These factors included hypovolaemia (haemorrhagic or distributive), obstruction (due to tension pneumothorax, cardiac tamponade, or pulmonary thromboembolus) and poor global cardiac function. These two studies included patients in whom pericardial effusions and right heart strain or thrombus were identified and treated, with several of these cases surviving to discharge. A small study of cardiac arrest in children[7] demonstrated both the feasibility of obtaining meaningful scans during the 'pulse check' of cardiopulmonary resuscitation (CPR), and the ability to identify potentially remediable factors during that time.

There is a case report of a child with cardiac arrest due to pericardial tamponade having this identified by POCUS and drained with quality survival.[8]

The use of POCUS in cardiac arrest must be limited to the 10 second pulse check, which requires significant experience, skill and practical coordination.[9] While it appears promising that the findings of an experienced operator may rapidly suggest specific contributing causes, prognoses or ameliorating interventions, further research is needed to define the place of POCUS in paediatric resuscitation. Many questions are raised including the potential for undue weighting to be given to amateur POCUS findings, and the ethical challenge of stopping resuscitation despite demonstrated cardiac wall motion.

Outcomes in **undifferentiated shock** and other peri-resuscitation scenarios in adults have demonstrated similar benefits from POCUS with regard to early diagnostic streaming and changes in patient management.[6,10,11] Various sonographic sequences with which to interrogate the shocked adult patient have been proposed, the most well-known of which is 'RUSH' (Rapid Ultrasound in Shock).[12] The RUSH concept of tank (venous capacitance), pump (heart), and tubes (large arteries and veins) relates well to Desbien's pathophysiological algorithm for PEA.[13] A proposed algorithm for undifferentiated shock or PEA (specifically non-shockable rhythm plus cardiac wall motion) in paediatrics, which combines these models and the above research findings, is outlined in Fig. 23.4.2. It is to be remembered that in some cases, clinical signs and management priorities may dictate management before sonographic confirmation, e.g. for paediatric tension pneumothorax.

Early recognition of possible congenital heart disease in infant collapse is another example of POCUS utility in paediatric shock. While detailed definition of cardiovascular anatomy requires high level echocardiographic experience, a systematic approach to shock with POCUS may reveal unexpected cardiac failure or gross chamber disproportion, triggering early cardiology review.

The **assessment of fluid status and the response to fluid therapy during shock** treatment can be aided by POCUS. Sonographic enquiries focus on respiratory variation in inferior vena cava (IVC) diameter, IVC/aorta ratio, left ventricular volume and contractility, and the presence or absence of signs of pulmonary interstitial fluid. While the optimal index (IVC diameter, IVC index, IVC/aorta ratio), technique and place of measurement have not yet been resolved, there is a well-demonstrated relationship between IVC and fluid status at the extremes.[14] This relationship is stronger in the unventilated than the patient undergoing positive pressure ventilation.[15] However, it may be useful for initial

and serial evaluation of fluid balance in undifferentiated paediatric shock. Concern about possible harm from excessive fluids during resuscitation remains unresolved in paediatric severe sepsis.[16,17] In adult critical care, there is evidence that the use of POCUS to identify preload reserve and direct fluid administration decreases renal complications and ICU stay.[18] POCUS helps to identify a specific patient's preload reserve and personal 'Starling curve', and serial POCUS helps to identify early pulmonary oedema prior to the development of clinical signs. These relationships and their sonographic findings are outlined in Fig. 23.4.3. Further research to outline the specific findings in paediatric shock in various settings is beginning.[19] and will be revelatory in our understanding of paediatric pathophysiology during different types of sepsis and its treatment.

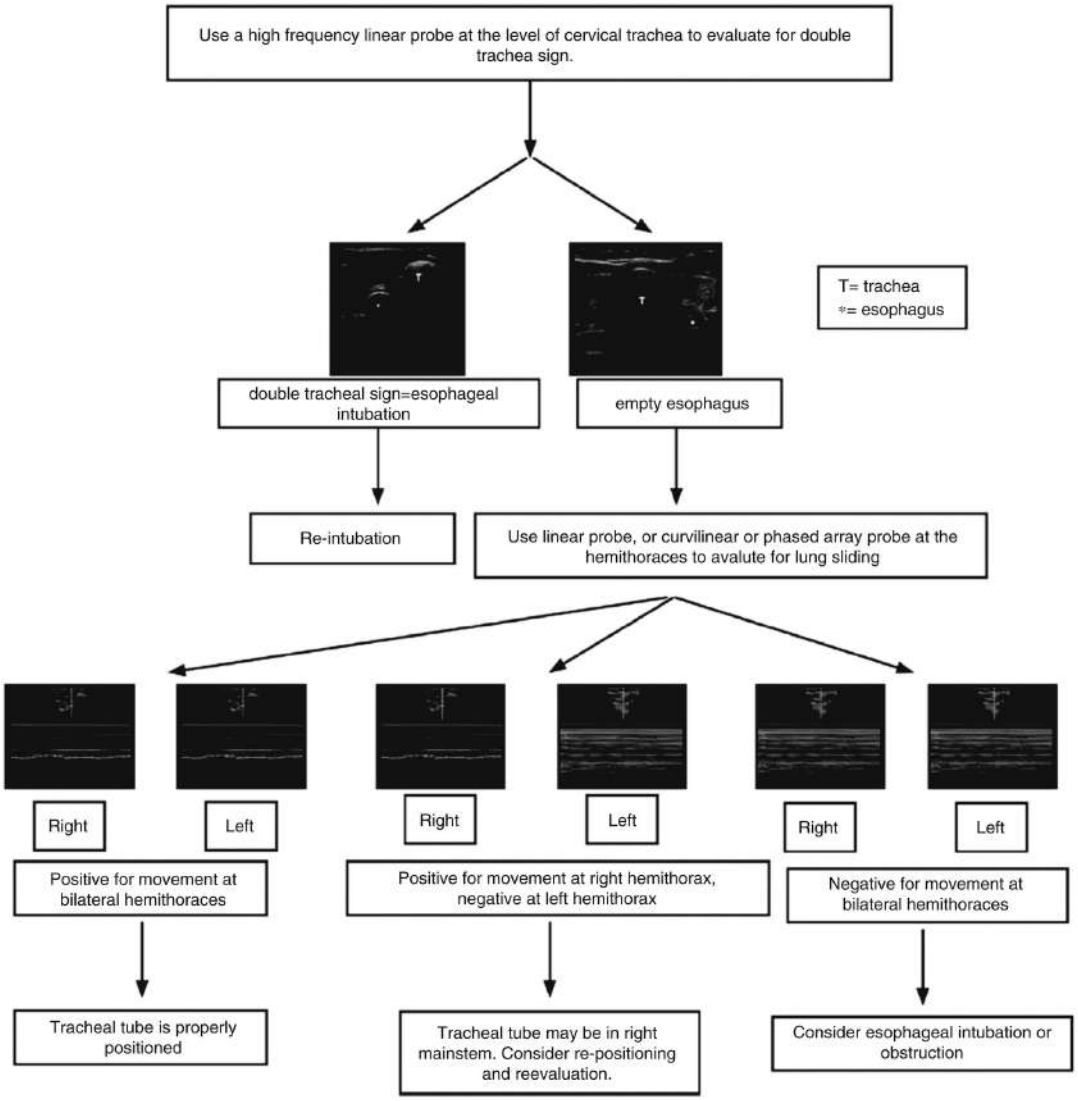

FIG. 23.4.1 LIN: Schema for point-of-care ultrasound during intubation.

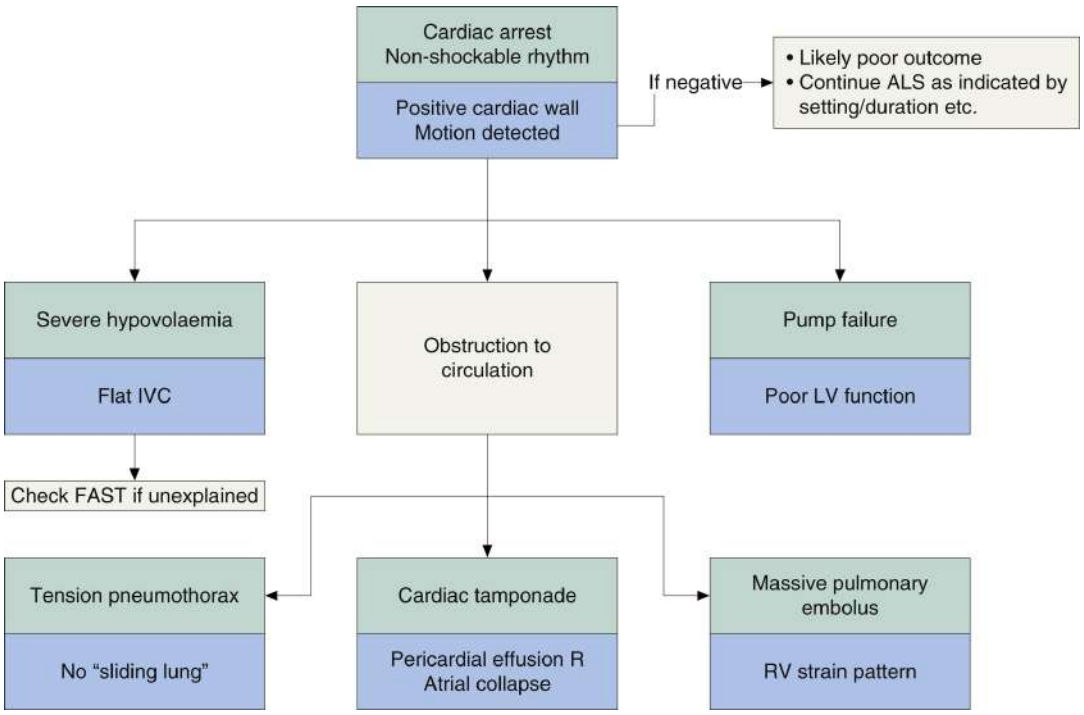

FIG. 23.4.2 Proposed algorithm incorporating point-of-care ultrasound into the pathophysiological schema for undifferentiated shock or pulseless electrical activity with cardiac wall motion.

An important added value of POCUS is the ability to perform **serial evaluations at the bedside**, incorporating a valuable additional dimension to ongoing clinical evaluations, e.g. serial FAST scans in trauma and interval fluid balance assessments in sepsis. Measurement of optic nerve sheath diameter (ONSD), which relates directly to intracranial pressure,[20] can be valuable both as a reflection of cerebral involvement in severe malaria,[21] and for the serial evaluation of intracranial pressure in patients being treated for diabetic ketoacidosis.[22]

In conclusion, studies of POCUS use in adult critical care have demonstrated that POCUS in resuscitation is feasible, more accurate for many assessments than physical examination alone, and useful in diagnostic streaming, the early initiation of goal directed therapy and serial monitoring of patient physiology. Its use in paediatric resuscitation is hampered by the current limited POCUS skill base and the paucity of paediatric resuscitations. We await the outcomes of proposed POCUS-incorporated pilot and multicenter studies with interest.

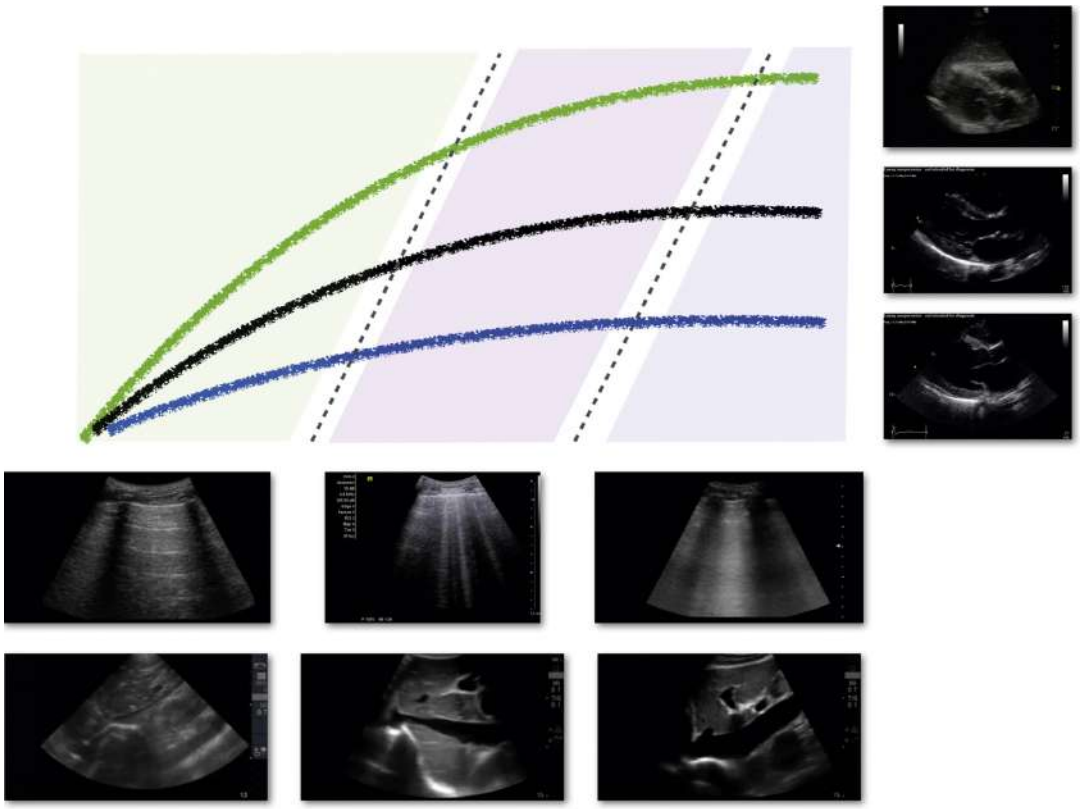

FIG. 23.4.3 Echo-guided life support: Starling x 3.

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SECTION 24

Common Procedures

OUTLINE

24.1

Estimating the weight of infants and children

Fenton O'Leary

Background

Emergency treatment of infants and children is sometimes difficult because children of different ages require different sizes of equipment, doses of medications, and volumes of fluids. Errors are common when selecting appropriate equipment and medications in critical paediatric emergencies, and mistakes are especially frequent with doses of drugs that are powerful cardiovascular agents, such as adrenaline (epinephrine).

Ideally each patient will be weighed on arrival and this measurement used to calculate his or her drug and fluid doses. Unfortunately, it is sometimes not possible to weigh patients due to the severity of their illness or their being unable to ambulate onto scales. Parents may remember a recent weight, but if not, either an aged-based or length-based tool is required to estimate the weight.

Clinicians should also consider whether drug doses should be based on total body weight (TBW) or ideal body weight (IBW). For instance, suxamethonium should be prescribed at TWB doses, but rocuronium should be prescribed at IBW, as using TBW can double its duration of action.[1]

Age-based tools to estimate body weight

If the age of the patient is known pre-arrival then important calculations can be made in the planning stage before the patient arrives and drugs, fluids and equipment prepared. Modifications can be made once the correct weight is known. Several formulas are available to estimate weight. In the general population the best guess formula has been shown to perform best and for over 5-year-olds is easy to remember.[2–4] However, in specific populations, specifically

derived weight charts may perform better.[5]

Best guess formula

Infants 1–11 months: weight (kg) = (age in months + 9)/2
Children 1–4 years: weight (kg) = 2 × age (+ 5)
Children 5–14 years: weight (kg) = 4 × age

Length- and body-habitus–based tools

In general length-based tools are better at predicting weight compared to age-based tools, and those tools that modify the weight prediction by a body habitus correction are even more useful.[2] The most common length-based tool is the Broselow tape, which is available commercially, and as well as weight estimation has recommended drug doses and equipment sizes already calculated depending on which colour band the patient's length falls into (Fig. 24.1.1). Clinicians should be aware that the Broselow tape may incorrectly classify the patient by colour band over 50% of the time, and clinicians may have to adjust doses and equipment sizes.[4]

Tips

- Use the patient's age to calculate drug dosages and equipment sizes pre-arrival.
- If possible always weigh the patient and try to obtain a bare weight in infants.
- Use length-based tools, with or without body habitus adjustment, if the patient is in the department.
- In patients who may be smaller or larger than average consider whether IBW is more appropriate than TBW to use in drug and fluid calculations.

FIG. 24.1.1 Colour-coded paediatric resuscitation tape. The Broselow® Pediatric Tape. © 2017 Vyaire Medical, Inc.; Used with permission.

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24.2

Basic airway management techniques

Scott Schofield, and Holly Smith

Background

Basic airway management skills are critically important in paediatric emergency medicine due to the high proportion of respiratory-based presentations. A high proportion of cardiac arrests in the paediatric population are related to hypoxia, necessitating familiarity and competence with airway and breathing interventions. There is a range of commonly used devices and techniques for supporting a child's breathing in addition to the application of oxygen delivery devices (see Chapter 2.3). These include oropharyngeal (OP) and nasopharyngeal (NP) airways, laryngeal mask airways (LMA), self-inflating bag-valve-mask (BVM) devices, flow-inflating bags, high-flow nasal prong oxygenation (HFNPO), T-piece device ventilation and mask continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BPAP).

Children have relatively large tongues, which may contribute to airway obstruction with loss of nasopharyngeal muscle tone, for instance, in patients who are post-ictal or overly sedated by drugs. Hypoxia and hypercarbia may develop secondary to obstruction. Non-invasive airway adjuncts, such as NP and OP airways, can maintain a patent airway even when the tongue falls back against the posterior pharyngeal wall. The LMA is designed to provide a seal around the laryngeal inlet when inserted and the cuff inflated and is used in situations involving a difficult BVM fit in an unconscious patient and as a back-up device where endotracheal intubation is unsuccessful.

Structurally, an NP airway is hollow, made of latex or a latex-like substance, and has a slight curvature to approximate the curvature of the nasopharynx. The distal end has a bevel, which helps to tunnel through the nasopharyngeal soft tissue but which may also cause inadvertent shearing trauma to the nasal septum. The proximal end of the NP airway has a wide flange to anchor the tube in place

at the nare. Patients tolerate this airway adjunct better than an OP airway because it does not trigger the gag reflex.

The OP airway is hollow, made of rigid plastic and has a slight curvature to approximate the curvature of the oropharynx. The distal end rests along the posterior tongue, which would trigger the gag reflex in a conscious patient, and the proximal end has a wide flange to anchor the tube in place at the lips.

Airway adjuncts may allow adequate spontaneous ventilation and avert the need for BVM ventilation or more invasive airway management such as endotracheal intubation.

BVM ventilation is the most important skill in paediatric airway management. This non-invasive manoeuver for assisted positive-pressure ventilation is effective treatment for most children with apnoea and hypoventilation. A child with respiratory insufficiency may require only temporary assisted ventilation with BVM. The device is widely available, and the technique for its use is simple and easily learned by all healthcare providers. Some children in respiratory failure who require prolonged ventilation or airway protection may require endotracheal intubation (Chapter 24.3).

In the BVM setup, oxygen flows into a bag reservoir, past a pressure relief valve and into a mask, which forms a tight seal around the child's nose and mouth. Squeezing the bag administers oxygen, when connected, under positive pressure to the lungs. While this technique does not fully protect the airway, as endotracheal intubation does, BVM ventilation will provide adequate emergency airway and ventilation support during initial resuscitation, transient hypoventilation or acute decompensation. The equipment is widely available in hospital settings, and the skillset is an expectation for all healthcare providers. It has been demonstrated that BVM ventilation is as effective as endotracheal intubation for airway management in the pre-hospital setting, regardless of the underlying aetiology.[1] Thus, both pre-hospital and in-hospital practitioners must be comfortable and proficient in performing BVM ventilation.

Oropharyngeal and Nasopharyngeal Airways

Indications

- Airway obstruction
- Respiratory decompensation
- Prolonged seizures or post-ictal state.

Contraindications

Nasopharyngeal airway

- Age less than 1 year old – the nares diameter is too small to introduce an NP airway.
- Nasal obstruction – attempting to introduce an NP airway into a nasal passage that is obstructed will be unsuccessful and may cause traumatic epistaxis.
- Severe facial injury or suspicion of facial or base of skull fractures – a fractured cribriform plate may allow an NP airway to traverse incorrectly into the intracranial space rather than into the posterior oropharynx.

Oropharyngeal airway

- Intact gag reflex – because the OP airway tip rests on the posterior tongue, a patient with an intact gag reflex will likely vomit and aspirate gastric contents.

Equipment

- Lubricating jelly (for NP airway)
- Tongue depressor or laryngoscope (for OP airway)
- NP airway (Fig. 24.2.1)
- OP airway (Fig. 24.2.2).

Preparation

Nasopharyngeal airway

Determine the correct NP airway size by one of three methods:

1. Follow the recommendations on a length-based resuscitation tape.
2. Choose an NP airway the length of which is equivalent to the distance from the patient's lateral nare to the tragus of the ear (Fig. 24.2.3).

FIG. 24.2.1 Nasopharyngeal airway.

3. Choose an NP airway the outer diameter of which is equivalent to the nostril's inner diameter or the diameter of the patient's fifth finger. Cut the length of the tube according to the length-measurement guide above.

Oropharyngeal airway

Determine the correct OP airway size by one of two methods:

1. Follow the recommendations on a length-based resuscitation tape.
2. Choose an OP airway the length of which is equivalent to the distance from the centre of the patient's mouth to the angle of the mandible (Fig. 24.2.4).

Positioning

1. Place the patient in a supine, neutral or 'sniffing' position appropriate for age.
2. To prevent the patient's tongue from worsening the airway obstruction, perform a chin-lift or jaw-thrust manoeuvre. Spinal immobilisation is crucial in patients presenting with trauma.

Procedure

Nasopharyngeal airway

1. Prelubricate the NP airway before insertion, ensuring the distal end is not occluded by lubricant.
2. When inserting the NP airway in the *right nostril*, the bevel already points towards the septum and is of minimal risk for nasal septal trauma. Gently introduce the NP airway directly posteriorly into the patient's nostril until the flange rests just external to the nostril orifice.

FIG. 24.2.2 Oropharyngeal airway.

3. When inserting the NP airway in the *left nostril*, the bevel is pointed

away from the septum. To avoid septal trauma, rotate the NP airway 180 degrees so that the bevel now points towards the septum. Insert the airway approximately 2 cm so that the tip passes the septal border. When resistance is felt, re-rotate the NP airway 180 degrees into its original, correct orientation. Finish inserting the NP posteriorly into the nostril until it rests just external to the nostril orifice.

Oropharyngeal airway

The most common mistake in OP airway insertion is causing damage to the soft palate or pushing the tongue farther back while inserting it and thus worsening airway obstruction:

1. Use a tongue depressor or laryngoscope blade to push the tongue inferiorly.
2. Under direct vision slide the OP airway directly over the tongue with the concave side down, i.e. with the tip pointed inferiorly.
3. Continue until the flange gently rests against the lips.

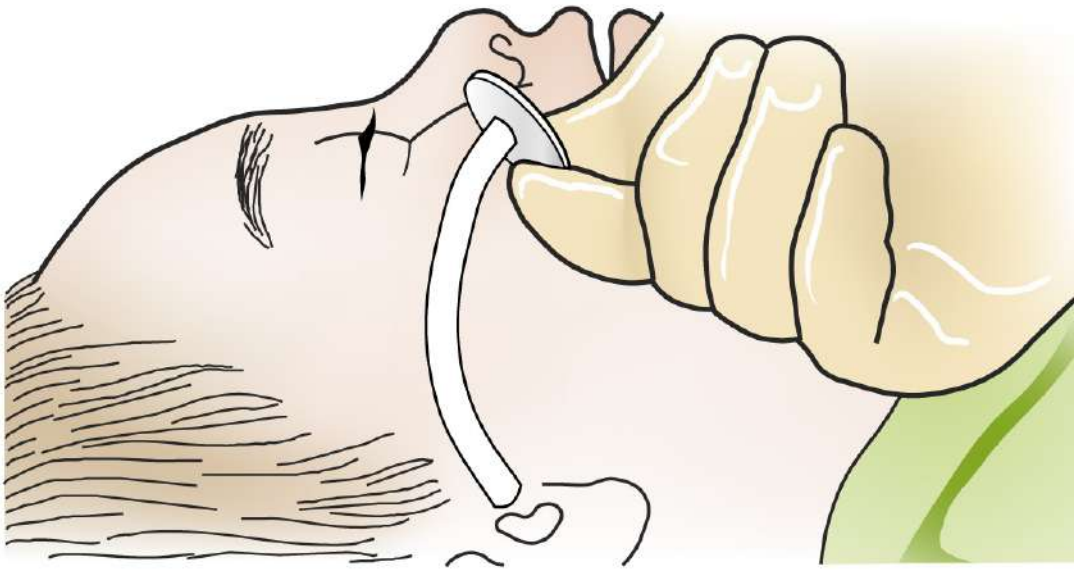

FIG. 24.2.3 Sizing the nasopharyngeal airway. Lateral border of nare to tragus of ear.

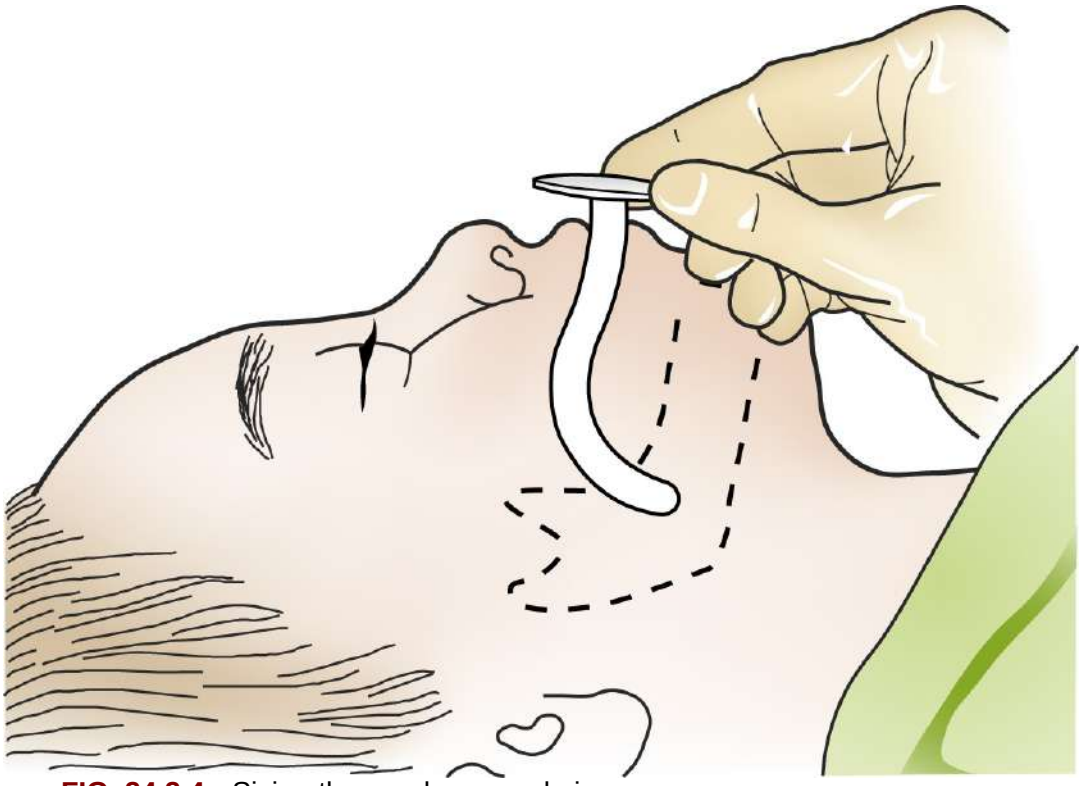

FIG. 24.2.4 Sizing the oropharyngeal airway. Center of mouth to mandibular angle.

Complications

- Triggering the gag reflex causing emesis and aspiration
- Laryngospasm
- Local trauma and bleeding to palate or nasal septum
- Worsening airway obstruction – when the OP airway is inappropriately long, it may directly occlude the posterior oropharynx; when it is inappropriately short, it may push the tongue farther back, causing more airway obstruction
- NP airway plug – the relatively small diameter of the NP airway makes it easily prone to occlusion with mucus, secretions and blood
- Intracranial placement of the NP airway.

Tips

- Insert the NP airway directly posteriorly along the floor of the nose and

not superiorly.

- Do not select too wide an NP tube, which may cause pressure necrosis to the nasal ala.
- If the flange of the NP tube is narrow and at risk of being pushed in past the nare, a safety pin can be pushed through the tube proximal to the flange to provide a wider anchor (Fig. 24.2.5).
- If an NP airway is not immediately available, an alternative is to use an endotracheal tube. First, choose the tube where the outer diameter is equivalent to the inner diameter of the patient's nostril. Second, trim the tube from the proximal end to the appropriate length as measured from the patient's nasal tip to the tragus of the ear. Leave the proximal ventilator adapter on the endotracheal tube in place so that the tube is anchored at the nasal tip and does not accidentally slip beyond the nares (Fig. 24.2.6).

FIG. 24.2.5 Use of a safety pin to anchor nasopharyngeal airway.

- Be sure the patient does not have a gag reflex before placing an OP airway.

Laryngeal Mask Airway

Indications for laryngeal mask airway

- Situations involving a difficult mask fit
- May be used as a back-up device where tracheal intubation is not successful.

Equipment

- Appropriate size LMA
- Syringe with appropriate volume for LMA cuff inflation
- Water-soluble lubricant
- Self-inflating or flow-inflating bag to ventilate
- Stethoscope to check for adequate air entry into both lungs once in place
- Tapes or other device(s) to secure position of LMA.

Preparation

- Select correct size (Table 24.2.1)
- Check the LMA by testing inflation and deflation of the cuff.
- Lubricate the back of the LMA thoroughly using a water-soluble lubricant and avoiding excessive amounts as inhalation of the lubricant can result in coughing or obstruction.

Positioning

Extend the head and flex the neck (tragus to sternal notch position) if possible, with consideration of the need for spinal immobilisation.

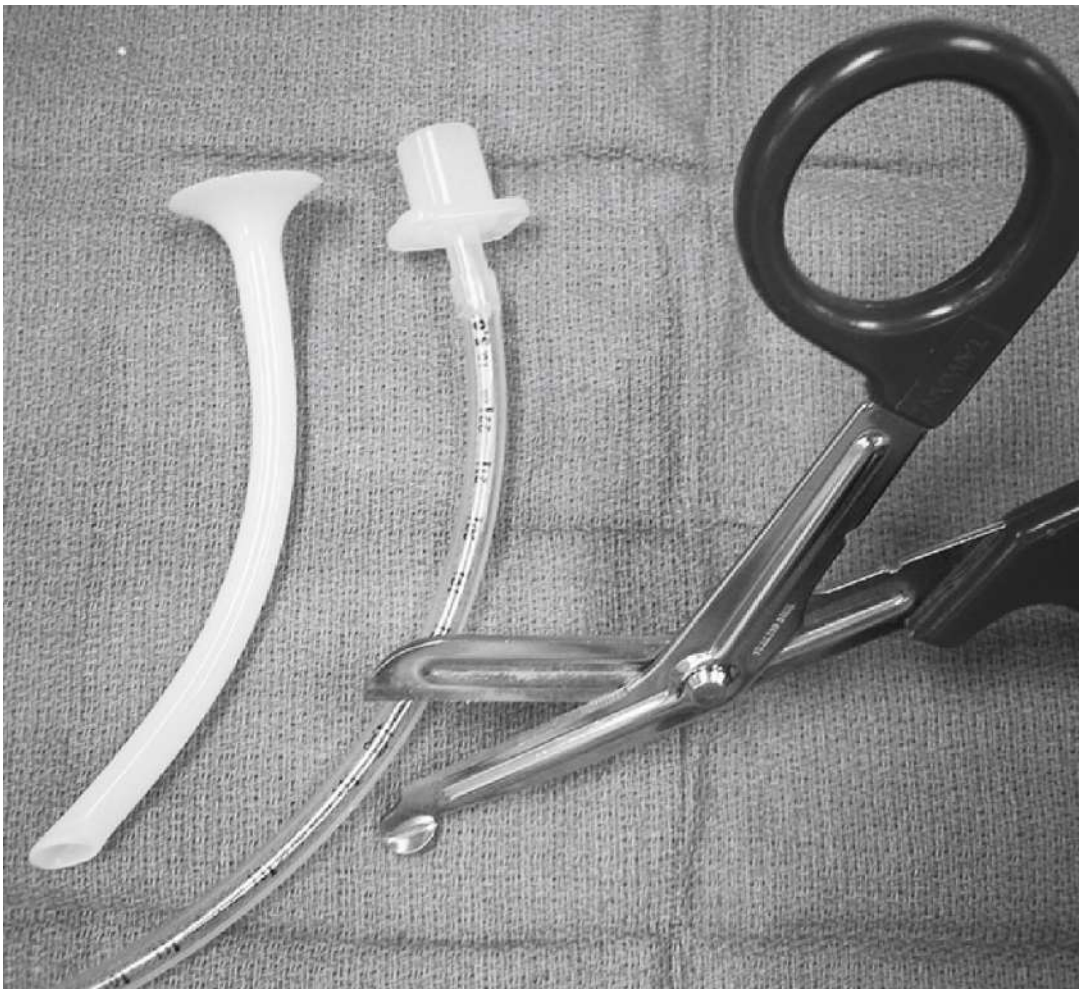

FIG. 24.2.6 Endotracheal tube cut into a nasopharyngeal airway.

Procedure

See Fig. 24.2.7.

1. The mask is held like a pen and inserted while pressing against the palate and posterior pharyngeal wall using the index finger until resistance is felt when the mask tip reaches the triangular base of the oropharynx.
2. The lumen of the LMA should be facing the patient's tongue and not the hard palate.
3. Inflate the mask with the recommended volume of air (see Table 24.2.1), avoiding over-inflation.
4. Normally the mask should be allowed to rise up slightly out of the hypopharynx as it is inflated to find its correct position.

5. Connect the LMA to a BVM device or low- pressure ventilator.
6. Ventilate the patient while confirming equal breath sounds over both lungs fields and the absence of gurgling sounds over the epigastrium.
7. Secure the LMA in position using the same techniques as for an endotracheal tube.

Complications

- Incorrect positioning with poor seal and large air leak resulting in inadequate pressures for ventilation
- Inadequate lubrication, complete deflation of the cuff or lack of pressure of the mask against the hard palate upon placement can cause the mask tip to fold back on itself. This may progress, pushing on the epiglottis and causing mechanical obstruction.

Tips

- Partial inflation of the cuff prior to insertion will allow it to hold its shape and prevent the tip of the cuff from folding upon insertion.

Table 24.2.1

Recommended weight-based sizing and inflation volumes

Weight of patient	Recommended size guidelines	Maximum air in cuff (mL)
5 kg	Size 1	4
5–10 kg	Size 1.5	7
10–20 kg	Size 2	10
20–30 kg	Size 2.5	14
30 kg–small adult	Size 3	20
Adult	Size 4	30
Large adult	Size 5	40

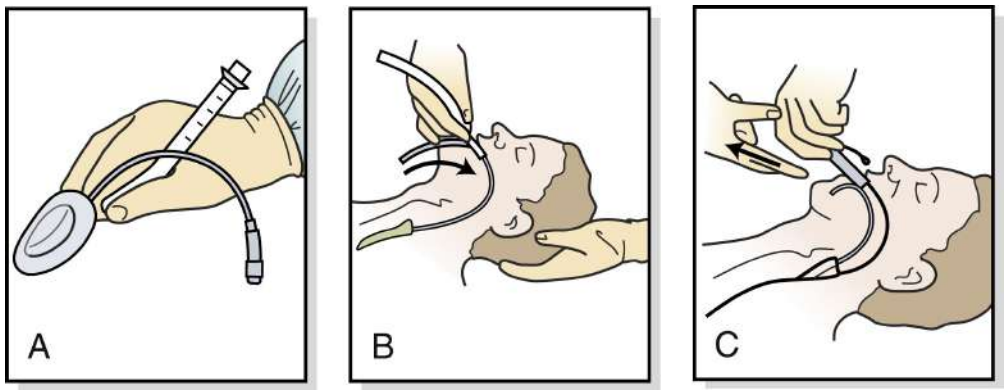

FIG. 24.2.7 Placing a laryngeal mask airway. Hold LMA with pencil grip (A). Insert while pressing against the palate and posterior pharyngeal wall using the index finger until resistance is felt when the mask tip reaches the base of the oropharynx (B). Once placed inflate device with the recommended volume of air (C).

- The insertion of the LMA by the standard technique is not always easy owing to the anatomy of the paediatric airway. Some advocate a rotational technique: with a partially inflated cuff, the mask is inserted with its lumen facing backwards and then rotated through 180 degrees, when the resistance of the posterior pharyngeal wall is felt, and then passed downwards into position behind the larynx.
- Insert a bite-block or roll of gauze to prevent occlusion of the tube should the patient bite down.

Bag-Valve-Mask and Flow-Inflating Mask Ventilation

Indications

- Hypoventilation or apnoea
- Respiratory failure
- Hypoxia despite high-flow oxygen administration via a non-rebreather mask.

Contraindications

Positive pressure ventilation should not be performed in the setting of a complete airway obstruction. If this exists, first perform airway clearance and basic life-

support manoeuvres. Then attempt removal of the obstruction (e.g. foreign body), if necessary, under direct laryngoscopy with Magill forceps.

Relative contraindication

In the presence of a congenital diaphragmatic hernia or a tracheo-oesophageal fistula, positive pressure ventilation can cause insufflation of the stomach and subsequent extrapulmonary compression of the lungs. This may compromise optimal oxygenation and ventilation.

Bag-Valve-Mask Equipment

- Appropriately sized mask (Fig. 24.2.8)
- Self-inflating bag reservoir (Fig. 24.2.9)
- Oxygen supply (optional)
- Oxygen saturation monitor.

Preparation

1. Select an appropriately sized mask. The mask should rest over the bridge of the nose superiorly and the cleft of the chin inferiorly. Masks are available for all ages ranging from preterm neonates to young children and adults. Round masks should be used for neonates and infants and triangular masks for school-aged children, adolescents and adults.
2. Select the appropriate self-inflating bag from neonatal (240–250 cc), child (500 cc) and adult (1000–1600 cc) sizes.
3. Connect the oxygen tubing to the self-inflating bag and attach to the face mask.
4. Commence oxygen flow to the BVM set up at 15 L per minute.

Positioning

Patient positioning is essential for successful BVM ventilation. Maintain a supine, neutral (infant) or sniffing (child) neck position to keep the airway patent. Because of their large occiputs, infants and toddlers are prone to hyperflexion of the neck and consequently benefit from a small towel roll under

their shoulders to achieve a neutral position. Both hyperflexion and hyperextension of the neck may contribute to airway obstruction, compromise ventilation and increase risk of spinal injury.

Procedure

1. Perform the head-tilt and chin-lift manoeuvre to open the airway and lift the tongue away from the soft palate of the oropharynx. Proper positioning and suctioning of excess secretions will often relieve the respiratory compromise without BVM ventilation.
2. In the setting of trauma where spinal injury is a consideration, do not perform the head-tilt/chin-lift manoeuvre. Instead, provide in-line spinal immobilisation, and perform a jaw-thrust manoeuvre to create a patent airway.

FIG. 24.2.8 Choosing the appropriately sized bag–mask.
Various sized masks are available for all ages. These masks have an inflated circumferential rim, which provides a tight seal to the patient's lower face. With an appropriately sized mask, the superior aspect of the mask should rest over the patient's nasal bridge, and the inferior aspect should rest over the cleft of the chin.

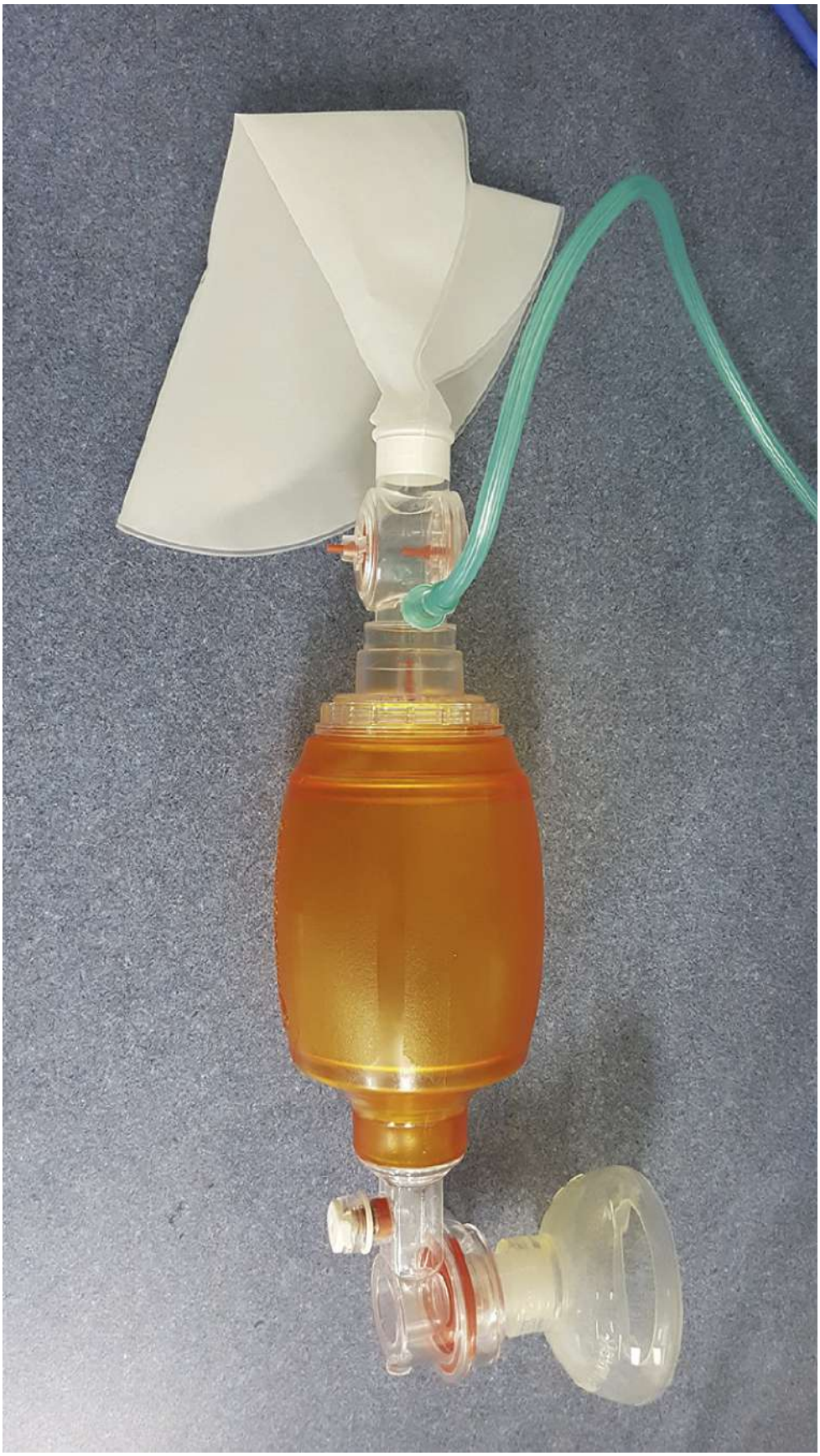

FIG. 24.2.9 Paediatric bag-valve-mask device.
Supplemental, high-flow oxygen flowing into a bag serves as the oxygen reservoir for bag–mask ventilation. A paediatric bag is adequate to oxygenate and ventilate a small child. Alternatively, if a paediatric bag is not readily available, an adult bag can also provide adequate oxygenation and ventilation. Be careful not to administer excessive tidal volumes with this larger bag.

3. If airway patency is still suboptimal, reposition and suction the patient. If still inadequate, insert an oropharyngeal or nasopharyngeal airway underneath the mask to lift the tongue from the posterior oropharynx.
4. In the *one-person BVM technique* place the mask on the patient's face and achieve an airtight seal over the mouth and nose. Using the non-dominant hand, place the thumb and index finger on the superior and inferior parts of the face mask, respectively. Cradle the tips of the other three fingers along the mandible, and lift the jaw up towards the mask to create the seal. This is the 'E–C clamp' manoeuvre, based on the E-shape of the three fingers along the jaw and the C-shape of the thumb and index finger along the face mask (Fig. 24.2.10). In creating the seal, pull the jaw anteriorly rather than push the mask posteriorly into the patient's face. To ventilate, squeeze the oxygen reservoir bag using the dominant hand.
5. In the *two-person BVM technique* place the mask on the patient's face and achieve an airtight seal over the mouth and nose. For operator #1, place the thumb and index finger of both hands on the superior and inferior parts of the face mask, respectively. Cradle the tips of the other three fingers of both hands along either side of the mandible and symmetrically lift the jaw up towards the mask to create a seal. This is a double 'E–C clamp' manoeuvre (Fig. 24.2.11). Again, pull the jaw anteriorly rather than push the mask posteriorly into the patient's face. Operator #2 ventilates the patient by squeezing the oxygen reservoir bag.
6. Provide BM ventilation at a rate of 20, 30 and 40 inspirations per minute for the child, infant and neonate, respectively. Squeeze the bag slowly and gently to avoid high pressures and flow rates, which will result in gastric insufflation. Aim for an inspiratory-to-expiratory ratio of 1:2. A common error is to over-ventilate the patient with too rapid a rate.

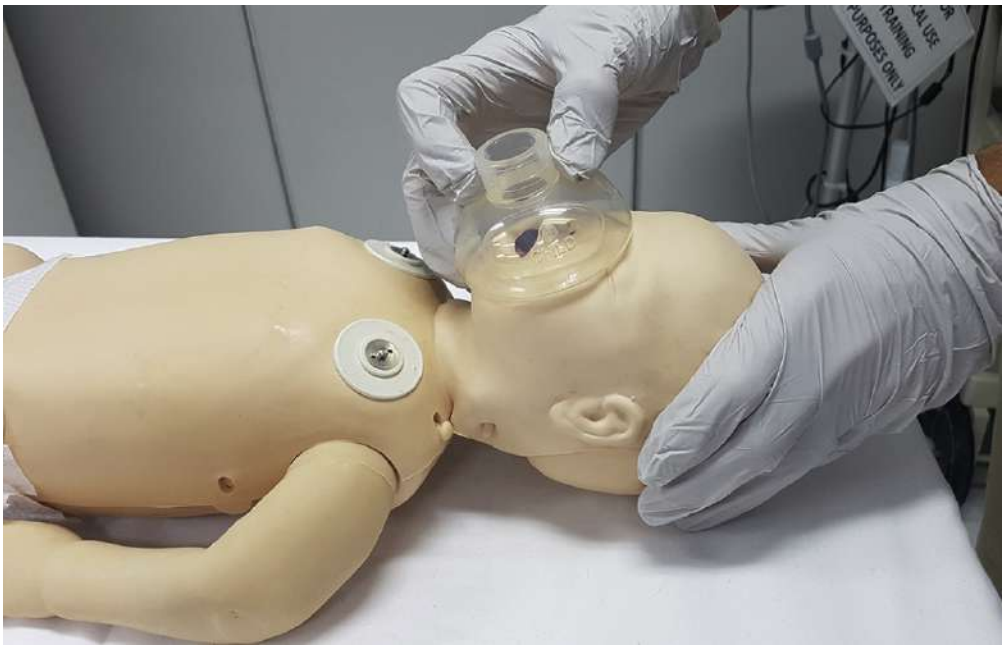

FIG. 24.2.10 E–C clamp bag-valve-mask technique.

FIG. 24.2.11 Two-handed E–C clamp bag-valve-mask technique.

7. Provide a BVM tidal volume of 8 mL kg^{-1} to oxygenate and ventilate the patient. As an effective alternative to estimate the appropriate ventilation volume, watch for bilateral chest rise and fall with each breath.
8. In addition to visualisation, auscultation of bilateral breath sounds in the

mid-axilla and improvement of the oxygen saturation both corroborate with adequate ventilation.

9. With prolonged BM ventilation, decompression of the stomach with an orogastric (OG) or nasogastric (NG) tube is essential due to the likelihood of gastric insufflation. Aspirating any gastric contents and keeping the gastric tube on free drainage reduce the risk of emesis and consequent aspiration.

Complications

- Incorrect mask sizing causing inadequate ventilation or trauma to the eyes
- Gastric insufflation and distension
- Aspiration from emesis
- Pneumothorax from poor lung compliance, excessive tidal volumes or excessive inspiratory pressure
- Hypoxia.

Tips

- In paediatric trauma cases requiring in-line spinal immobilisation, perform BVM ventilation with the two-person technique. One operator focuses on providing an adequate mask seal, while the other operator focuses on squeezing the bag and maintaining spinal alignment. In comparison, a one-person BVM technique often inadvertently extends the patient’s neck while trying to achieve an adequate mask seal.
- Incorrect technique in holding the mask can lead to irregularity or creases in the inflatable rim and a leak in the seal.
- Poorly fitting masks will prevent adequate ventilation. If the mask is too large it will cross the orbits, which may injure the eyes and cause a leak in the seal. If the mask is too small it may occlude the nares.
- When elevating the jaw anteriorly to form a mask seal with the third, fourth and fifth fingers, be sure to lift up along the bony mandible rather than the submandibular soft tissue. In addition to causing trauma, compressing the submandibular soft tissue may inadvertently occlude the airway.

- Many BVM devices have a pressure relief valve to prevent excessive positive pressure ventilation. This may inadequately ventilate a patient with low lung compliance. Occlude this valve to allow higher positive pressures during inspiration, while watching for chest rise.
- For the neonate and infant, assessing chest rise and fall for adequate BVM tidal volume is subtle. The optimal viewing angle is from the patient's side at the level of his or her bed.
- In order to prevent the common complication of over-ventilating a patient, say aloud 'squeeze – release – release' repeatedly while correspondingly squeezing and releasing the bag. This approximates the correct ventilatory rate and inspiratory-to-expiratory ratio for the patient.
- A BVM device should not be used as a source of oxygenation alone in a spontaneously breathing patient. The valve at the mask end of the device is opened during active compression of the self-inflating bag by the operator. Without active ventilation by an operator the spontaneously breathing patient would have to overcome a pressure gradient to open this valve independently, making it an inappropriate device for oxygen delivery without ventilatory support.

Flow-Inflating Bag

A flow-inflating mask is able to provide non-invasive positive pressure ventilation. Unlike the BVM it is designed to be able to provide continuous positive end expiratory pressure (PEEP). This is achieved by manually occluding or restricting the exhaust port of an expandable, soft bag whilst co-ordinating compression of the bag for inspiratory breaths. There are tradeoffs to the advantage of being able to provide PEEP. This device requires a higher degree of dexterity and practice to use effectively. The flow-inflating bag always requires a gas source to operate. It is also capable of producing very high pressures so should always be used with an inline manometer.

Equipment

- Appropriately sized mask (Fig. 24.2.8)
- Flow-inflating (anaesthetic) bag (Fig. 24.2.12)
- Manometer and T-piece (optional)

- Oxygen or medical air supply
- Oxygen saturation monitor.

FIG. 24.2.12 Flow-inflating bag and manometer.

Preparation

1. Select an appropriately sized mask
2. Select the appropriate flow-inflating bag (500 cc, 1000 cc, 2000 cc, 3000 cc)
3. Connect the circuit to a gas (oxygen or medical air) source, attach the appropriate face mask and consider connecting a manometer to the circuit
4. Commence gas flow to the circuit at a minimum of 8 L/min

Procedure

1. Ensure optimal positioning and airway opening techniques.
2. Start gas flow to the bag at a minimum of 8 L/min. The bag will only inflate when the circuit is closed by obtaining a good mask seal on the patient.

3. Partially occlude the exhaust port of the flow-inflating bag to achieve a constant PEEP at the desired pressure. This can be done in one of two ways: with the thumb and forefinger, leaving the lateral three fingers to compress the bag to deliver breaths; or with the fifth finger leaving the rest to compress the bag (Fig. 24.2.13).
4. Check PEEP and inspiratory pressures on the inline manometer.
5. To increase the pressure being delivered, apply more pressure to the occlusion of the exhaust port.

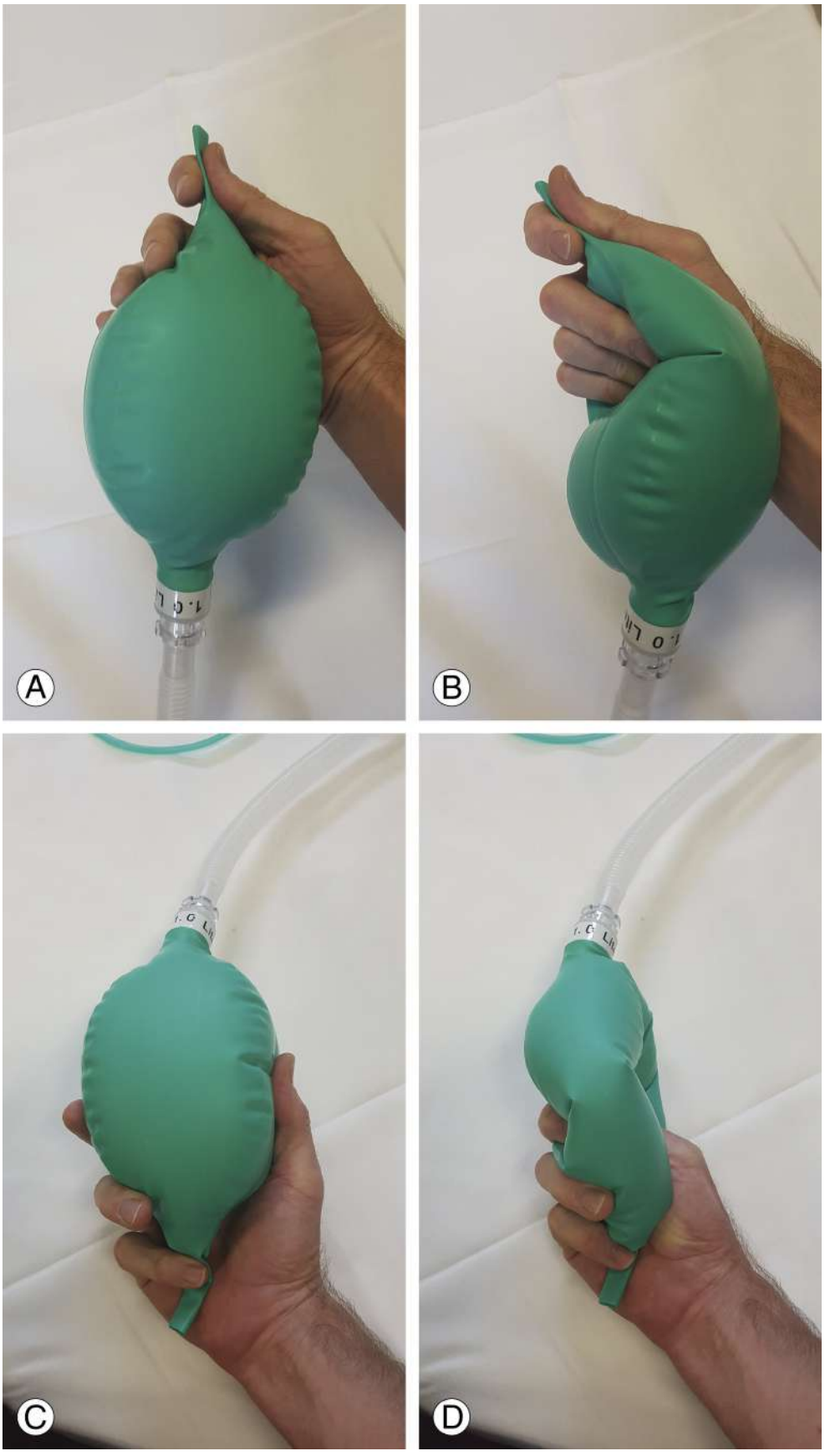
A
B
C
D

FIG. 24.2.13 Flow-inflating bag grip and technique. Partially occlude the exhaust valve with either a pincer grip (A&B) or flexed fifth finger (C&D) and intermittently ventilate by compressing the bag with the rest of the available fingers.

24.3

Non-Invasive Ventilation

Scott Schofield, and Holly Smith

Non-Invasive Continuous Positive Pressure Ventilation

Indications

- Insufficient oxygenation
- Insufficient alveolar ventilation.

Contraindications

- Cardiac/respiratory arrest
- Inability to protect airway
- Upper airway obstruction
- Untreated pneumothorax
- Persistent vomiting
- Maxillofacial surgery or base of skull fracture.

Relative contraindications

- Staff inexperience
- Patient refusal, intolerance.

Humidified High-Flow Nasal Prong Oxygenation

The administration of high-flow humidified oxygen through nasal cannula has a number of benefits for a select paediatric population. By continuously washing

out exhaled gas it reduces the anatomical dead space, it theoretically produces some positive distending pressure through snug-fitting nasal cannulae and the humidification improves mucociliary clearance and reduces some bronchoconstriction caused by the inhalation of cold, dry air.[2]

HFNPO is used most frequently for infants with bronchiolitis. It can be considered for children with moderate to severe work of breathing who do not respond to low-flow nasal prong oxygenation or bronchodilator therapy. There is limited evidence for its use outside of bronchiolitis and should therefore only be considered with senior consultation in these situations.

Equipment

See Fig. 24.3.1.

- Humidifier base unit
- Humidifier and patient circuit
- Sterile water
- Oxygen blender
- Appropriate-sized nasal cannulae (neonatal, infant, paediatric).

Procedure

Below is a guide to the general principles for setting up an HFNPO circuit. There are multiple manufacturers whose devices are capable of delivering HFNPO. Please refer to your local instructions and guidelines:

1. Connect air and oxygen hoses from gas supply to oxygen blender.
2. Set up humidifier base ensuring it has sterile water in its reservoir and a reliable power supply. It will need to be powered to heat and humidify the gas supply. Ensure correct connection of humidifier circuit to the oxygen blender, base and patient circuit.
3. Connect the patient circuit including the appropriate-sized nasal cannula. The patient circuit may include a temperature probe.
4. Fit the patient with the correct-sized nasal cannulae.
5. Commence HFNPO, titrating FiO_2 to a target oxygen saturation and flow rate based on patient weight (normal starting flow is 1–2 L/kg/min).
6. Ensure appropriate regular monitoring of respiratory rate, heart rate,

SpO_2 and work of breathing.

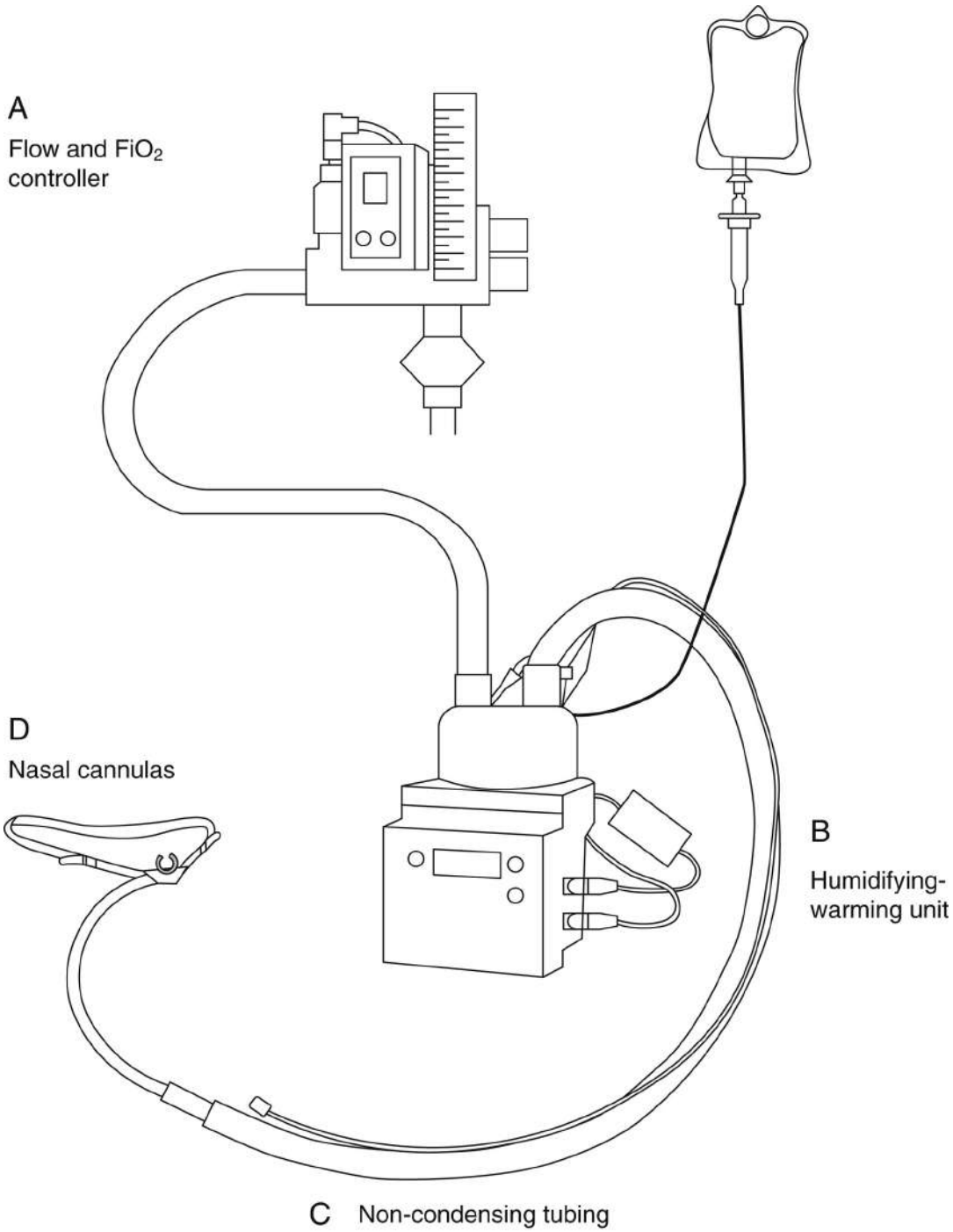

FIG. 24.3.1 High-flow nasal prong oxygenation equipment.

Complications

- Gastric distension with air is a common problem, especially in infants who continue to feed orally. Insertion of a nasogastric tube on free drainage should be considered for all patients on HFNPO >1 L/kg/min. Children on 2 L/kg/min HFNPO should be kept nil per os with supplemental intravenous fluids initially. Oral or nasogastric feeds should only be considered once a child is stable and has demonstrated an improvement of respiratory distress.

Tips

- HFNPO is often used as a bridging therapy in an attempt to avoid the need for more invasive forms of ventilation. Once commencing a patient on HFNPO it is imperative that the most senior member of your clinical team is aware and you should have a low threshold for escalating management if the therapy is unsuccessful.
- Flow can be increased up to 2 L/kg/min to assist work of breathing
- Oxygen percentage (FiO_2) can be titrated to achieve an acceptable oxygen saturation (SpO_2).
- Transfer to and management in an intensive care environment should be considered for patients who do not improve following 2 L/kg/min HFNPO or who is requiring FiO_2 of ≥ 0.6 to maintain adequate SpO_2.
- Weaning HFNPO therapy should begin with FiO_2, reducing by 0.1 per hour. Once the FiO_2 is less than 0.3 the flow rate can be weaned by 50% (e.g. 1 L/kg/h to 0.5 L/kg/h) per hour with regular reassessment.

T-Piece Ventilation Device

T-piece devices (e.g. Neopuff™, NeoPIP™, etc.) are used to provide positive pressure ventilation including positive end-expiratory pressure (PEEP) and PIP. They are commonly used in the neonatal population and are usually suitable for use in children up to 10 kg. They can also be used to provide CPAP via a face mask or endotracheal tube. The T-piece delivers a consistent set pressure for both PEEP and peak inspiratory pressure (PIP) as long as there is an adequate face mask seal or minimal endotracheal tube (ETT) leak. The inspiratory and

expiratory times are controlled by the operator by manually occluding a PEEP valve. These devices require a gas source to operate and have an associated risk of barotrauma as the operator is responsible for controlling the inspiratory volume.

Equipment

See Fig. 24.3.2.

- Gas supply (oxygen and/or medical air)
- Oxygen blender (optional)
- T-piece device (Neopuff™, NeoPIP™ or similar)
- T-piece circuit with PEEP valve
- Appropriate-sized face mask or endotracheal tube.

Procedure

- Ensure manometer reads zero prior to gas flow connection
- Connect gas supply (oxygen, air or blended oxygen source)
- Connect T-piece circuit
- Check and adjust pressure settings:
    a. Turn on gas supply at a flow of 5–15 L per min.
    b. Use provided cap (or your finger/hand) to occlude the patient end of the circuit.
    c. Check maximum pressure relief setting by occluding PEEP valve on T-piece and turning PIP control to maximum (clockwise). Then turn the maximum pressure relief control (clockwise or counterclockwise) to desired max pressure.
    d. Set PIP by occluding PEEP valve and turning the PIP control down (counterclockwise) to desired inspiratory pressure.
    e. Set PEEP by adjusting the PEEP valve to desired expiratory pressure.
- Fit an appropriate-sized face mask to the T-piece, and place over the patient's nose and mouth (device can also be connected to a correctly placed endotracheal tube)
- Ventilate by occluding the PEEP valve with a finger or thumb to allow

inspiration and removing to allow expiration (Fig. 24.3.3).

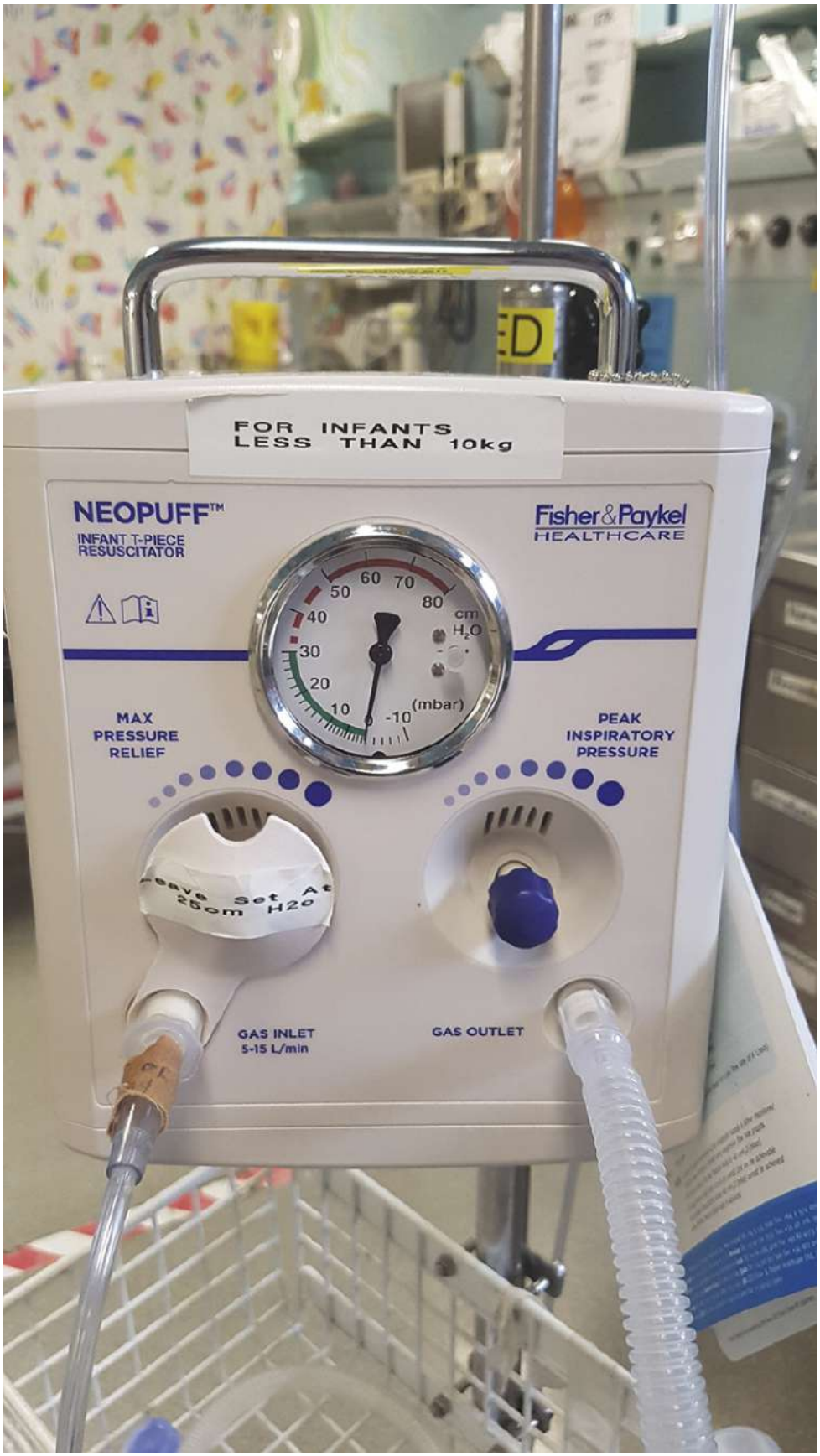
FOR INFANTS
LESS THAN 10kg
NEOPUFF™
INFANT T-PIECE
RESUSCITATOR
Fisher&Paykel
HEALTHCARE
50 60 70 80
40
30
20
10
0 -10
cm H2O
(mbar)
MAX
PRESSURE
RELIEF
PEAK
INSPIRATORY
PRESSURE
GAS INLET
5-15 L/min
GAS OUTLET

FIG. 24.3.2 Neopuff™ T-piece ventilation device.

Complications

- Barotrauma resulting from high pressure or high volume.

Tips

As guideline for starting pressures, in term newborns use a PEEP of 4–6 cm H_2O and PIP of 20–25 cm H_2O. Preterm neonates may require higher pressures if hyaline membrane disease is present.

Continuous Positive Airway Pressure/Biphasic Positive Airway Pressure

CPAP or BPAP may be required to support spontaneous ventilation for a range of conditions. CPAP provides a constant expiratory pressure and is often used to support patients with obstructive pathology (e.g. obstructive sleep apnoea, acute exacerbations of asthma). BPAP offers greater support than CPAP with inspiratory and expiratory pressures and is often used for neuromuscular diseases and respiratory failure.

There are too many models of equipment capable of providing CPAP and BPAP to effectively cover in this book. Refer to your local guidelines and equipment for specific and detailed instructions.

Equipment

- Appropriately sized nasal or facial mask
- Headgear or straps to secure mask to patient (refer to your local guidelines and equipment)
- Ventilator or CPAP device
- Oxygen and/or medical air supply

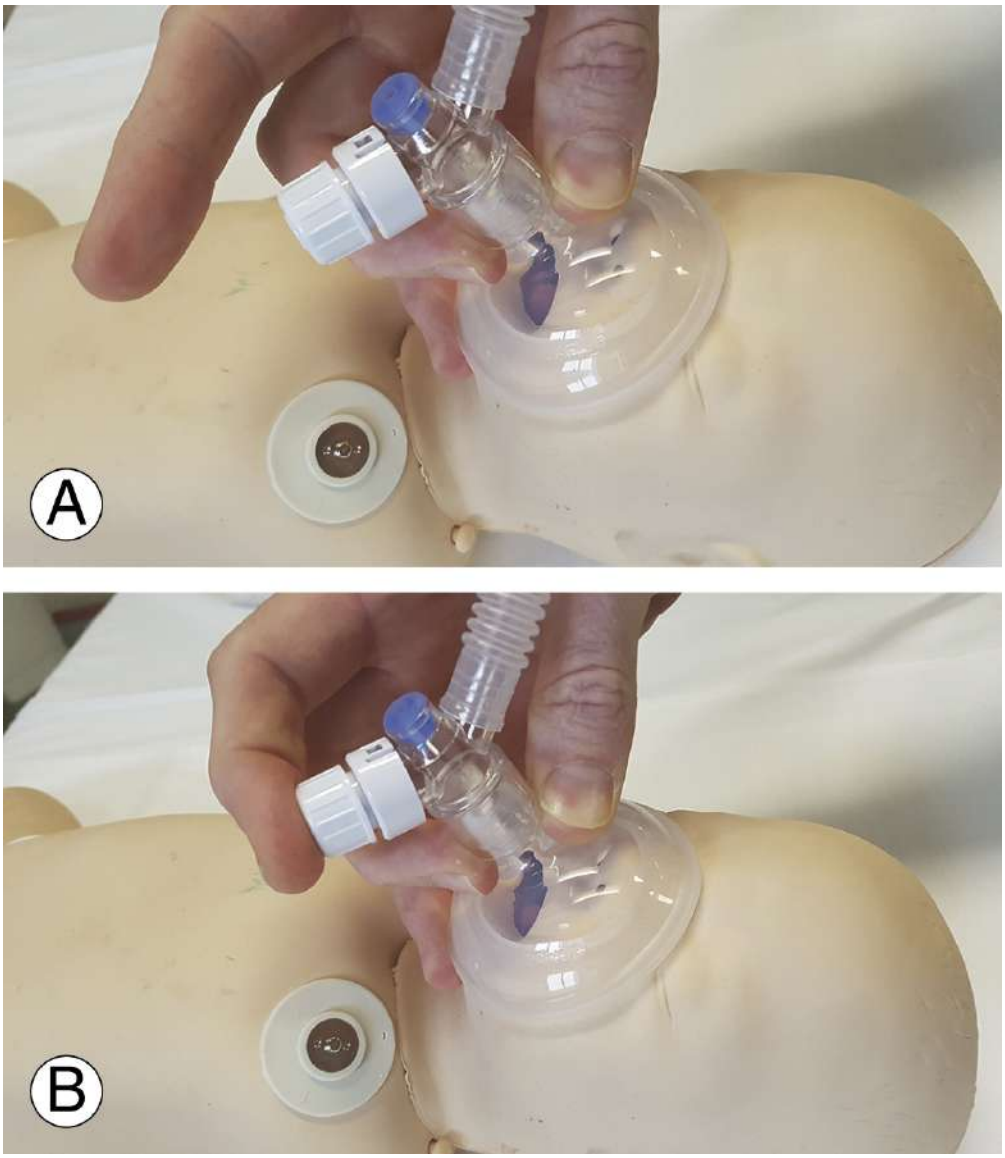

FIG. 24.3.3 Ventilation using a T-piece ventilation device. Provide positive end-expiratory pressure (PEEP) by ensuring a tight mask seal with PEEP valve open (A). Assist inspiration with peak inspiratory pressure by occluding PEEP valve (B).

- Ventilator circuit
- Cardio respiratory monitoring (including SpO_2).

Contraindications

- Apnoeic, arrested or comatose patient
- Inability to protect airway

- Oesophageal atresia/trachea-oesophageal fistula
- Gastric perforation.

Relative contraindications

- Necrotising enterocolitis
- Abdominal distension/surgery.

Procedure

1. Refer to local guidelines and equipment, and ensure senior staff are aware of the patient's need for ventilator support
2. Set up ventilator or CPAP device with desired inspiratory and/or expiratory pressures
3. Connect ventilator circuit and test pressures on a test lung
4. Select appropriately sized nasal or facial mask, and ensure fit on patient with adequate seal
5. Attach mask to circuit and fit to patient, noting that patients will often take some time to settle once pressure support mask has been applied.

Complications

- Volutrauma/barotrauma (e.g. pneumothorax)
- Atelectasis
- Acute lung injury
- GIT complications (gastric distension, stress ulcers, GIT bleed, paralytic ileus)
- Air swallowing leading to potential for vomiting and aspiration
- Oxygen toxicity
- Decreased cardiac output from reduced venous return
- Local pressure sores from mask fit.

Tips

Initial pressure settings will depend on the patient age and underlying pathology. A guideline for settings is below.

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24.4

Endotracheal intubation

Scott Schofield, and Holly Smith

Background

Endotracheal intubation (TI) provides a definitive airway. Insertion of a tube between the vocal cords and into the trachea allows optimal management of the patient's oxygenation and ventilation while also protecting the airway from aspiration. Depending on the preparation and skill of the practitioner, this procedure can be either life-saving or life-compromising. TI is the standard rescue procedure when bag-valve-mask (BVM) ventilation is ineffective, insufficient or required for prolonged periods.

The anatomy of the paediatric airway creates unique considerations during intubation as compared to adult intubation. Specifically, the differences are as follows:

1. A paediatric patient has a relatively larger tongue, making visualisation of the vocal cords more difficult. Hence, preintubation positioning of the patient plays a crucial role in TI success.
2. A paediatric patient has a wider and floppier epiglottis, which often obscures the view of the larynx.
3. A paediatric patient has more anterior and cephalad position of the vocal cords.
4. The cricoid ring is the narrowest part of the airway in patients less than 8 years old. This anatomical narrowing has traditionally been thought to adequately secure the endotracheal tube (ETT) in place at the level of the cricoid ring without an air leak and therefore negate the need for a cuffed tube. In contrast, adolescents and adults have a cylindrical-shaped rather than a funnel-shaped airway.
5. A paediatric patient has large adenoidal tissue and relatively small nares.

Consequently, nasotracheal intubation in the paediatric population is technically difficult and has a high complication rate from traumatic bleeding, aspiration and oesophageal intubation. These intubations also generally take a longer time to perform and require a patient who is awake and cooperative. The orotracheal route is the preferred approach in the emergency department.

In addition to proper positioning and equipment selection, successful emergent intubation often requires rapid sequence induction (RSI) of anaesthesia. RSI involves the administration of medications that provide transient sedation and paralysis to facilitate the procedure. Induction agents include ketamine, thipentone, fentanyl, midazolam and propofol. Neuromuscular paralysing agents include succinylcholine, rocuronium, and vecuronium. RSI is imperative prior to intubation of a conscious patient. Sedation and muscle relaxation prevent reaction to noxious stimuli, such as the laryngoscopic blade and tracheal tube insertion. This optimises visualisation of the vocal cords and successfully passing the tube. When performing RSI and intubation the practitioner should also be aware of the following autonomic responses:

1. Bradycardia – patients less than 5 years old have a higher risk for bradyarrhythmias with direct laryngoscopy because of vagal nerve stimulation. Consequently, premedicating with atropine should be considered in the RSI drug regimen.
2. Tachycardia and hypertension – induction medications attenuate this catecholamine response to laryngoscopy and intubation.
3. Gag reflex – patients requiring emergency intubation benefit from RSI medications because patients are assumed to have a full stomach, the contents of which may regurgitate during intubation. By blunting the gag reflex using RSI, the risk of oesophageal reflux and pulmonary aspiration decreases.

RSI agents must be chosen specifically for each individual patient with consideration of the medications' mechanisms of action and side effects. Practitioners must always perform a pre-intubation risk assessment including patient factors, equipment checklist and failed intubation plan. Preparation for alternative methods of ventilation is essential as following RSI the patient will be apneoic and will desaturate rapidly. Each effort to intubate should not exceed

30 seconds prior to abandoning the current attempt and re-oxygenating the patient.

Indications

- Cardiopulmonary arrest
- Respiratory failure or obstruction
- Excessive work of breathing refractory to non-invasive ventilation
- Loss of the gag reflex
- Need for prolonged ventilation or hyperventilation
- Expected clinical course (airway burns, trauma) or transport requirement.

Contraindications

- Adequate response to BVM ventilation with anticipated brief requirement for assisted ventilation
- Structural abnormalities, such as a large tongue haematoma, or massive facial injuries that require a tracheostomy or cricothyrotomy
- Functioning tracheostomy.

Equipment

1. Local intubation guideline and pre-intubation checklist
2. Cardiopulmonary and oxygen saturation monitor
3. Rigid-tip suction catheter
4. Bag-mask (BM) and self-inflating bag
5. Laryngoscope (Figs 24.4.1 and 24.4.2)
6. Tracheal tube (Fig. 24.4.3)
7. Tracheal tube stylet
8. Bougie
9. Video laryngoscope (optional)
10. Syringe for cuffed tracheal tube
11. End tidal CO_2 device for confirmation of intubation
12. Adhesive tapes or other device for securing the tracheal tube
13. Bite block or oropharyngeal (OP) airway.

FIG. 24.4.1 Curved (top) versus straight (bottom) laryngoscopic blades.

Preparation

Many institutions and organisations have developed and implemented local intubation guidelines and checklists. Become familiar with your local guideline and remember to refer to available pre-intubation checklists. If you do not have a local checklist the 'SOAP ME' mnemonic is a helpful way to remember the essential equipment required for safe intubation (Box 24.4.1).

Laryngoscope

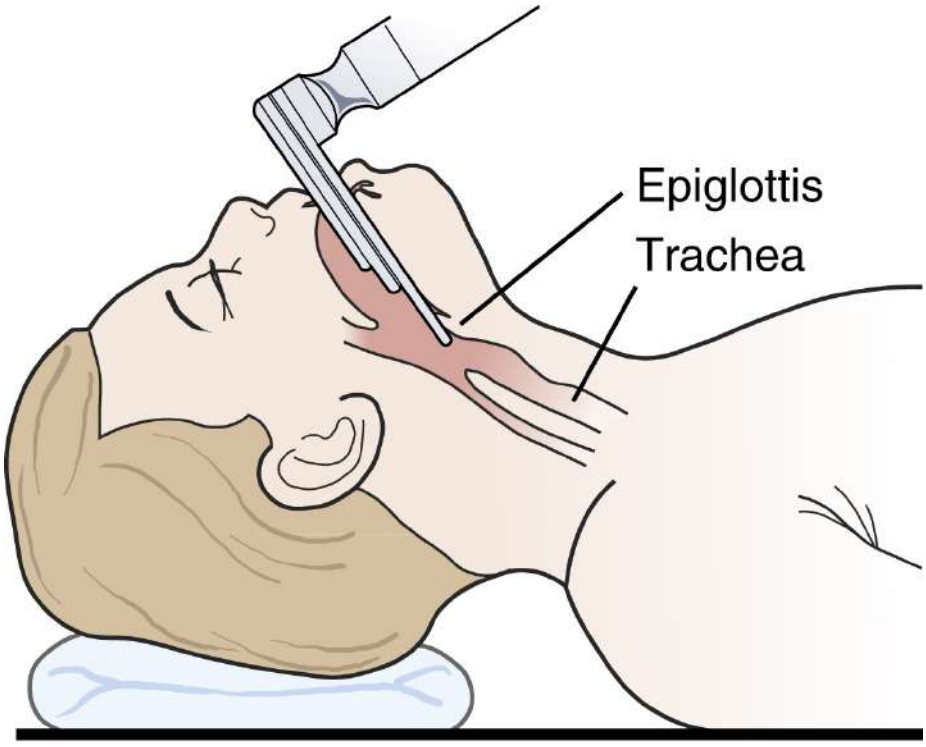
There are two main types of blades to visualise the vocal cords – straight and curved. For the neonate or infant, use a straight blade to more easily visualise the anteriorly located vocal cords. For older patients, whose anatomy more resembles that of adults, use either a straight or a curved laryngoscopic blade.

Additionally, the laryngoscopic blades come in different sizes. To determine

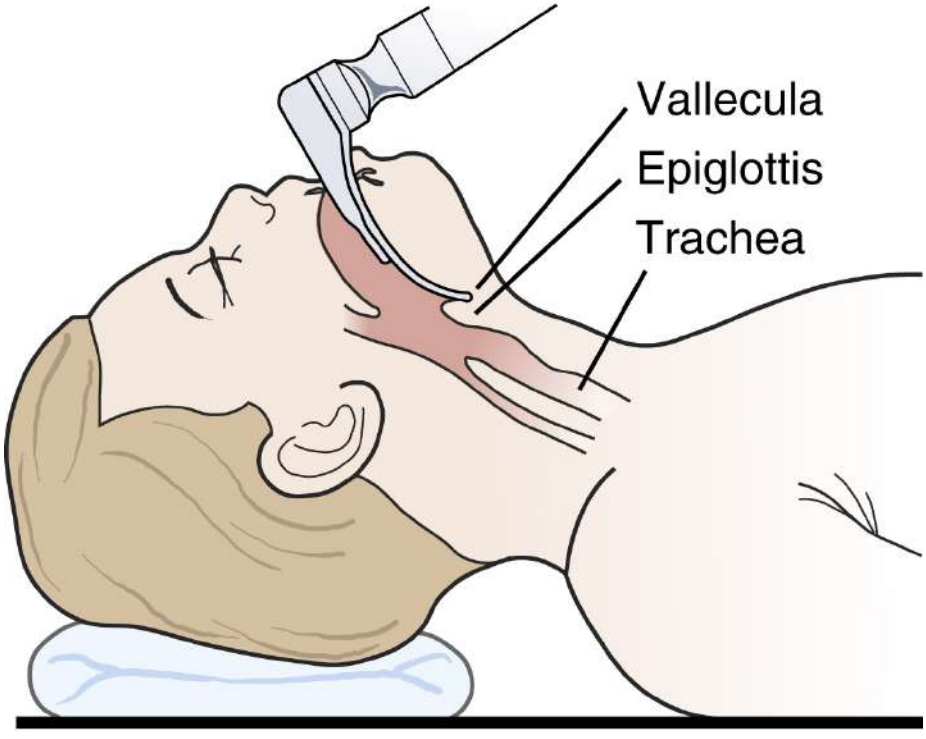
the correct size to use for a specific patient, either (1) use the length-based resuscitation tape (Chapter 24.1) or (2) use an age-based table (Table 24.4.1).

Endotracheal tube

Cuffed ETTs can assist in preventing an air leak around the tube. Because of the relatively funnel-shaped airway in paediatric patients traditional teaching has been to use uncuffed tubes in children less than 8 years old. The recent design and production of paediatric-specific cuffed ETTs (e.g. Microcuff™) with a more distal and thinner (10 μm) cuff have strengthened evidence for use of cuffed tubes in all paediatric patients weighing over 3 kg.[1,2] For older patients and adults, cuffed tubes are essential because of their cylindrical-shaped airway. Without a balloon cuff, this airway shape is susceptible to significant air leaks. An appropriately sized ETT can be selected by using a length-based resuscitation tape or using the formula (age/4 + 4). Tube sizes range from 1.0 to 9.0, representing the inner diameter in millimetres.



A



B

FIG. 24.4.2 Insertion of laryngoscopic blades.
Laryngoscopes are available with either a straight blade end (A) or a curved blade end (B). The straight laryngoscopic blade should slide just 'under' or posterior to the epiglottis and elevate the tongue and epiglottis as a unit to visualise the vocal cords. Because of their floppy and large epiglottis, neonates and infants require this straight blade technique for better visualisation of the cords. For older patients, both straight and curved blades are acceptable. When the curved laryngoscopic blade is used, the blade should rest in the vallecular space just anterior to the epiglottis and posterior to the base of the tongue. Lifting the laryngoscope up towards the ceiling elevates the vallecular space and the tongue as a unit, which indirectly lifts the epiglottis anteriorly to allow visualisation of the vocal cords.

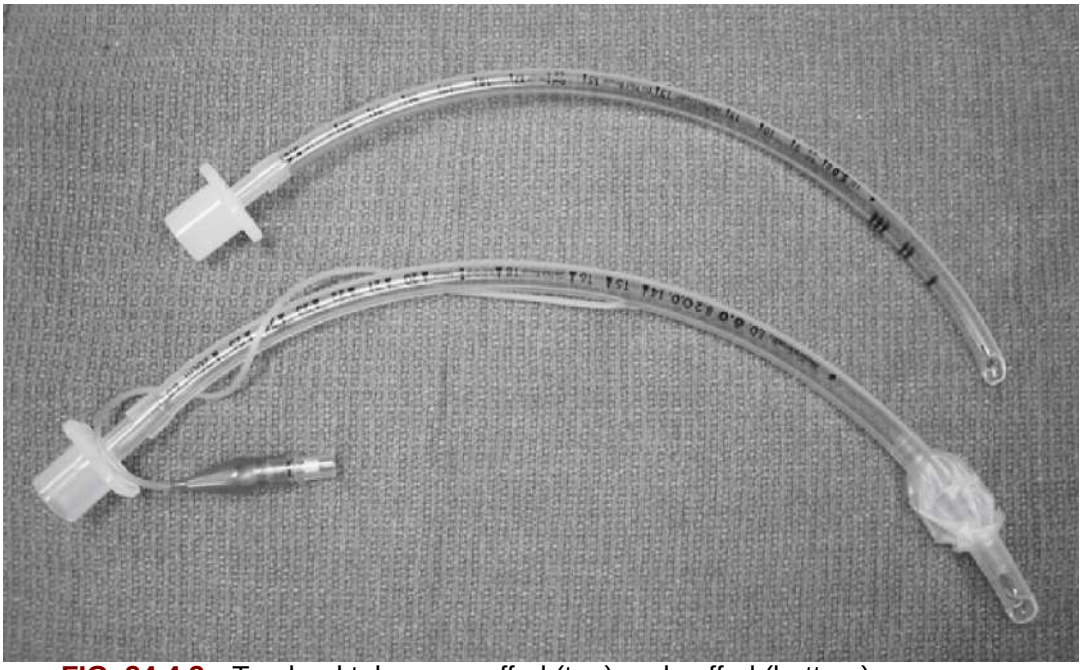


FIG. 24.4.3 Tracheal tubes, uncuffed (top) and cuffed (bottom).

Box 24.4.1 Preparing the equipment –'SOAP ME' mnemonic

1. Suction – turn the suction apparatus on and test the suction catheter.
2. Oxygenation equipment – test the self-inflating reservoir bag and bag–mask (BM) setup.

3. Airway equipment – select the appropriately sized tracheal tube and insert a stylet. Check the tracheal tube cuff integrity with a syringe, if applicable. Also obtain tube sizes slightly larger and smaller than expected, in case of unexpected anatomy. Obtain laryngoscopic blades slightly larger and smaller than anticipated, and test the blade lights. Finally, check the rescue airway devices in case of a failed intubation.
4. Pharmacological agents – prepare rapid sequence intubation drugs.
5. Monitoring equipment – turn on the cardiopulmonary monitor, oxygen saturation monitor, and tracheal confirmation device.

Table 24.4.1

Laryngoscope size selection

Age	Blade type and size
Premature infant	Straight blade (Miller) 0
Newborn to 1 year	Straight blade 1
2–12 years	Straight blade 2, Curved blade (MacIntosh) 2
Adolescent	Curved blade 3

Positioning

The patient should be placed in a supine, neutral or ‘sniffing’ position. For infants and small children, placing a small towel roll under the shoulders prevents the occiput from hyperflexing the neck and occluding the airway.

Procedure

1. Provide in-line spinal immobilisation, if necessary. Have an assistant stand at the side of the bed or kneel at the head of the bed, maintaining a neutral position of the head and neck by cupping the patient’s ears with both hands or securing one hand across the patient’s forehead (Fig. 24.4.4).
2. Pre-oxygenate the patient by BVM ventilation with 100% oxygen for 1–2 minutes. Be aware that the smaller the child, the faster the rate of oxygen desaturation during the intubation procedure. Apnoeic

oxygenation with nasal prongs at a flow rate of 8 L/min should be considered, despite limited evidence for its benefit in children.[3]

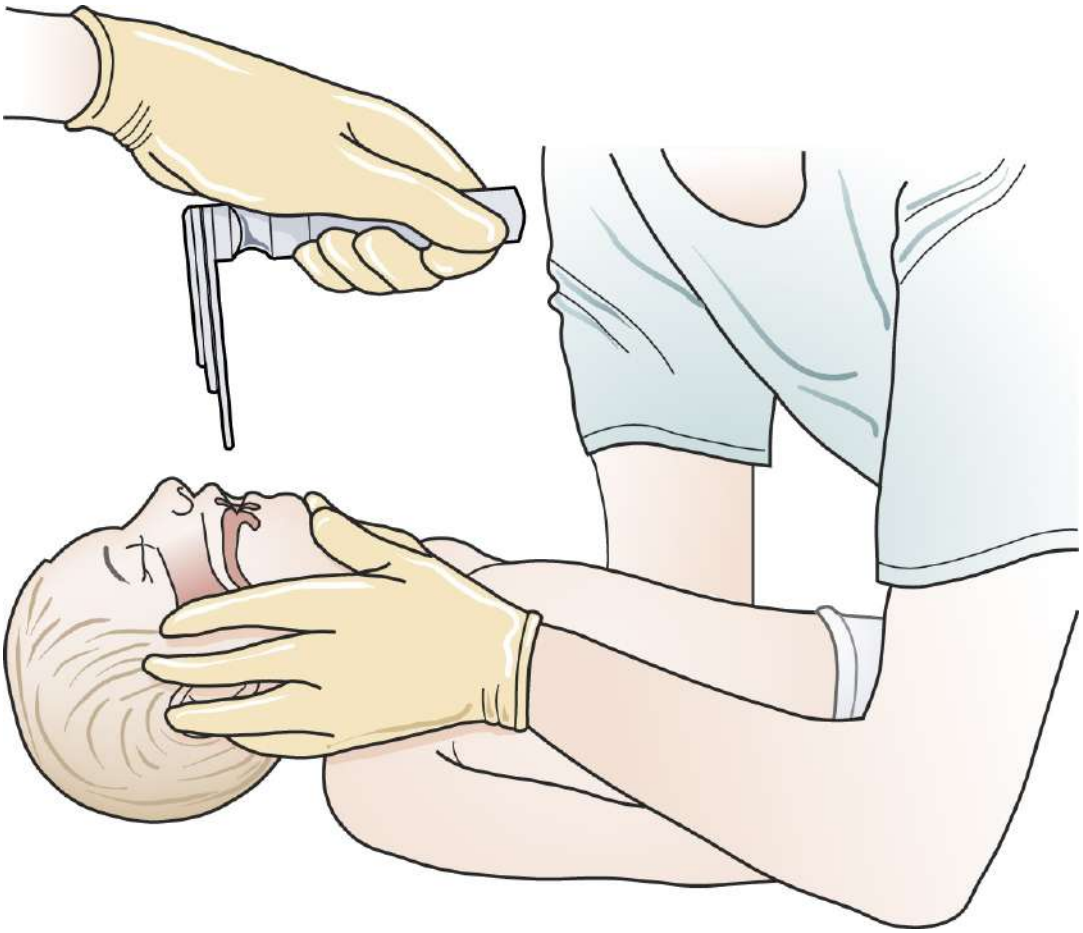


FIG. 24.4.4 Spinal immobilisation by assistant.

3. Administer RSI medications and appropriately support ventilation until neuromuscular paralysis occurs.
4. Grasp the laryngoscopic handle with the left hand, and engage the blade so that the attached light bulb illuminates.
5. Gently insert the laryngoscope, starting from the right side of the patient's mouth, and 'sweep' the tongue towards the left. A straight laryngoscope blade should be inserted midline.
6. Continue inserting the blade deep while lifting the laryngoscope anteriorly and inferiorly, trying to visualise the vocal cords. The epiglottis can be picked up directly by the blade or moved out of view

with pressure from the tip of the blade in the vallecula.

7. Suction any excessive secretions or gastric contents that are obscuring adequate visualisation of the epiglottis and vocal cords.
8. External laryngeal or cricoid manipulation may be used to improve visualisation of the cords. The pneumonic BURP refers to backward, upward, rightward pressure, which is commonly used to bring the high anterior paediatric airway into view. Traditional cricoid pressure to passively occlude the oesophagus in an attempt to prevent regurgitation of gastric contents should not be used as standard practice but considered in patients at high risk of regurgitation and aspiration.[4]
9. Once the vocal cords are identified, use the right hand to insert the tracheal tube into the right side of the mouth, and pass between the vocal cords. The use of a stylet in the ETT may improve your chances of successful intubation. Many operators will initially pass a bougie through the vocal cords and then proceed to passing the ETT over the bougie.
10. Watch the tube or bougie pass between the vocal cords and enter the trachea. The best way to determine whether a TI is successful (i.e. not an oesophageal intubation) is by direct visualisation. Other confirmatory modalities may provide false and misleading information.
11. Insert the tube to the appropriate length by inserting to the bold black line at the level of the cords, or use the formula to calculate the length at the lips (age/2 + 12).
12. Inflate the cuff (if applicable) to 10 cm H_2O.
13. Attach an oxygen source (BVM, flow-inflating bag, ventilator circuit) to the tracheal tube and ventilate the patient, watching for equal chest rise with each inspiratory effort.
14. Connect end tidal CO_2 detection device (colour or waveform capnography). This is the gold standard confirmation for correct ETT placement.
15. Listen for equal breath sounds in bilateral axilla and the absence of gurgling sounds in the epigastrium.
16. Secure the tube in place with adhesive tape.
17. Insert a bite block or OP airway to prevent the patient from biting down on the tracheal tube.
18. Insert an orogastric or nasogastric tube, aspirate gastric content and keep on free drainage to prevent regurgitation and aspiration.

19. Obtain a post-intubation chest radiograph to confirm successful tube placement, adequate depth of tube insertion and successful gastric tube insertion. The ETT should end 1–2 cm above the carina.
20. Ensure that the patient has adequate ongoing sedation, muscle relaxation and analgesia.

Complications

- Traumatic injury to lips, teeth, oropharynx, larynx, vocal cords and oesophagus
- Incorrect insertion of tube into oesophagus or intracranially
- Emesis and aspiration
- Increased intracranial, intraocular and intragastric pressures
- Cervical spinal injury
- Dysrhythmia
- Hypertension or hypotension
- Hypoxia and/or hypercarbia
- Pain and anxiety
- Dislodgement of tracheal tube during movement.

Tips

- Equipment failure is an unnecessary complication with potentially devastating consequences. Ensure availability and functionality of all equipment prior to intubation.
- Anticipating a failed initial intubation reduces the complication rate in tracheal intubation. Prepare multiple laryngoscope blades and have a spare functioning handle. Have slightly larger and smaller tracheal tubes and failed-airway alternative devices at the bedside (e.g. BVM and supraglottic device). It is useful to consider and discuss alternative airway plans with your intubation team.

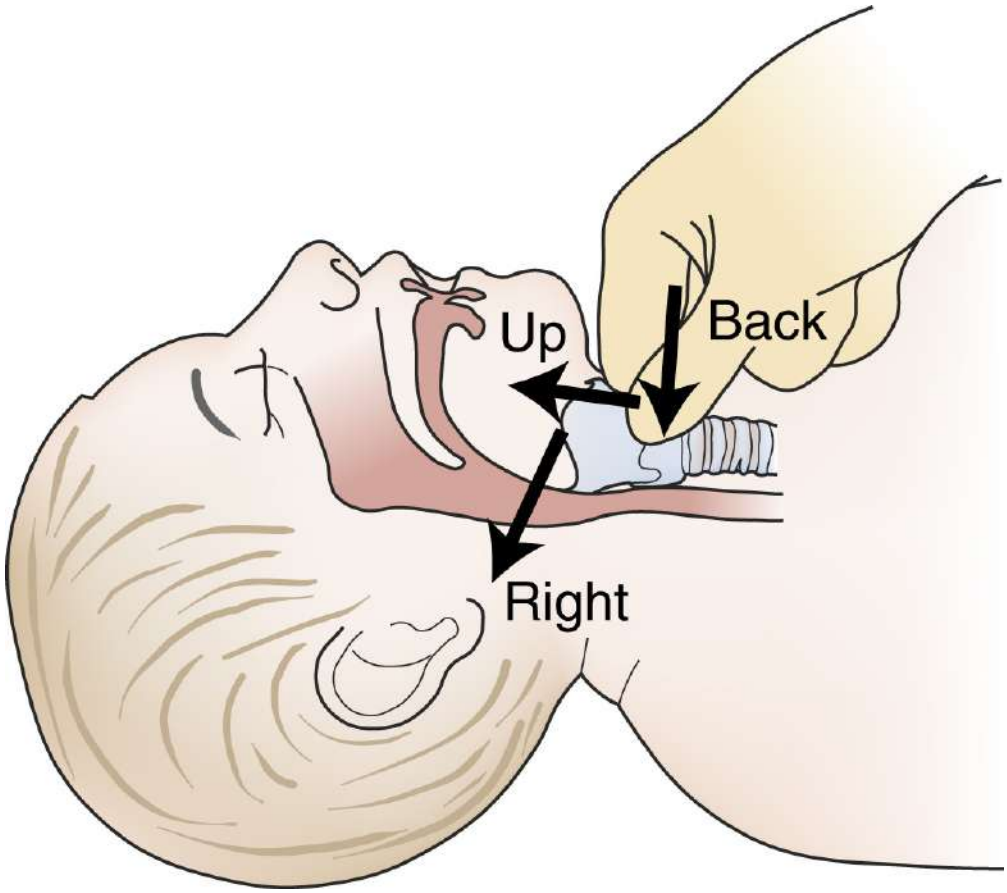


FIG. 24.4.5 Improving vocal cord visualisation – BURP manoeuvre. Using your right hand (non-laryngoscope hand), manipulate the external cricoid cartilage with Back (posterior), Up (superior), and Rightward Pressure. This essentially moves the vocal cords towards the right and out from ‘under’ the overhanging epiglottis.

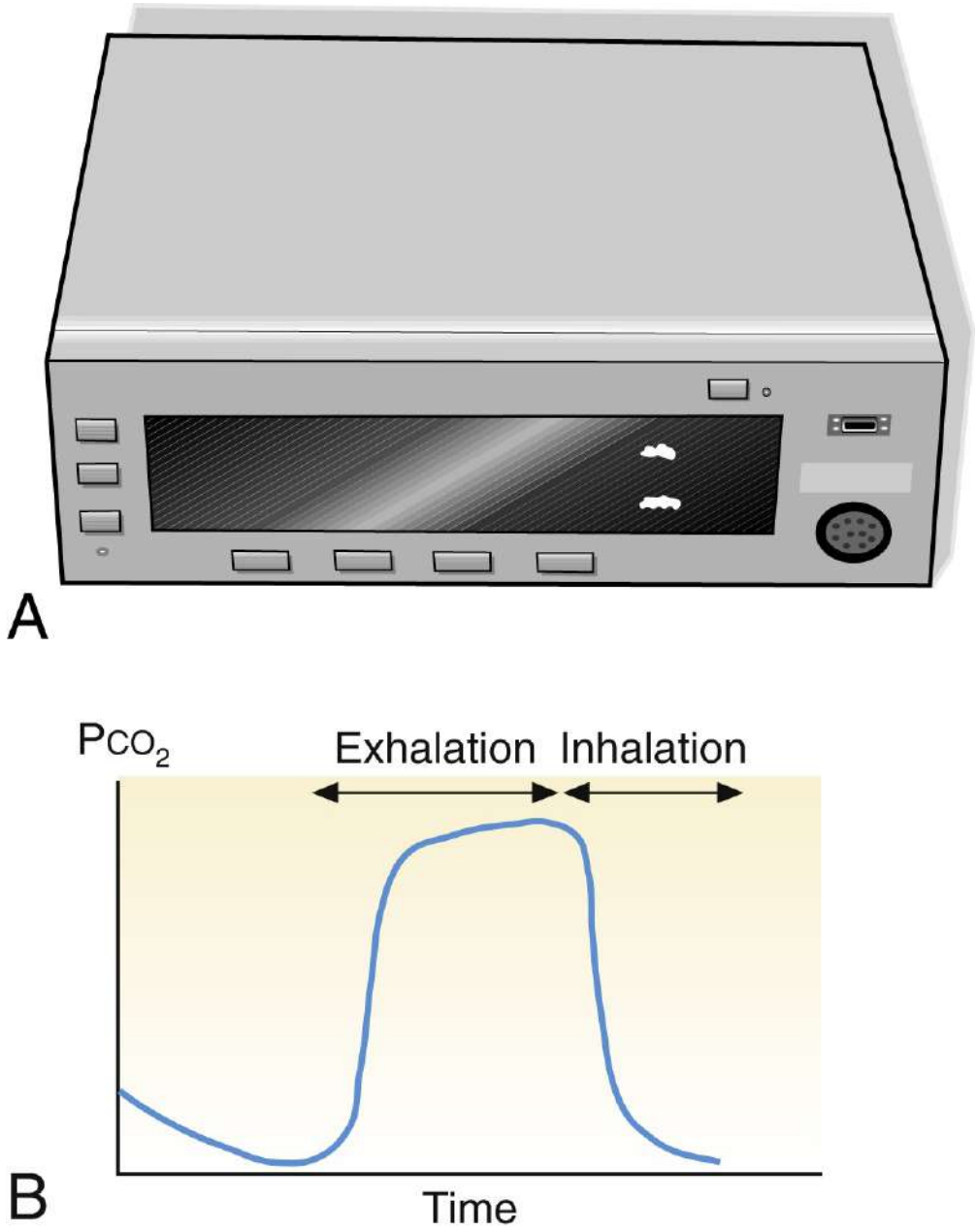


FIG. 24.4.6 Digital capnography instrument (A) and typical waveform reading (B).

- Some practitioners recommend starting laryngoscopy with epiglottoscopy. Once the epiglottis is successfully visualised it is expected that the vocal cords will be easy to find once the epiglottis is picked up directly with the straight blade or with pressure in the vallecula with the curved blade.
- While performing laryngoscopy, a common mistake is to use the patient's teeth as a fulcrum for the blade. Avoid dental injury by lifting

the laryngoscope in the plane of the handle and avoiding rotational force.

- To help visualise a patient's vocal cords, which are being obscured by a floppy epiglottis, use the BURP manoeuvre (Fig. 24.4.5).

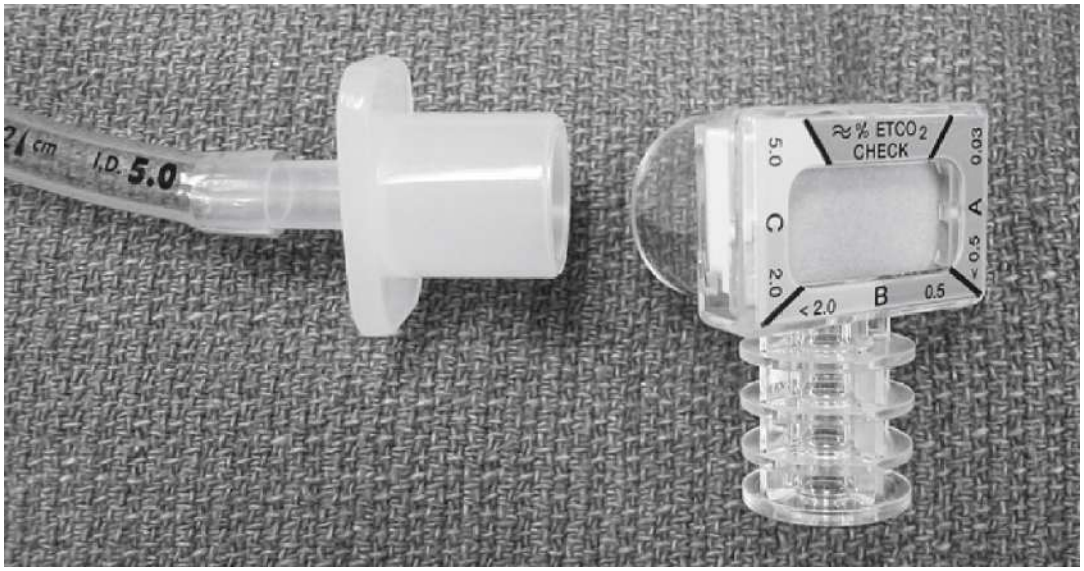


FIG. 24.4.7 Colorimetric capnometer.

- Because of the short trachea in the paediatric population, a common mistake during intubation is inserting the tracheal tube too deep, resulting in a right main stem intubation.
- Because of the short trachea in the paediatric population, the tracheal tube may easily dislodge from its position in the airway. Be vigilant about securing the tube at all times, but especially during transport or with any movements. Frequently recheck its position.

Confirmation of Intubation

Background

Confirming correct placement of the ETT is crucial because of the high morbidity and mortality of an inadvertent and unrecognised oesophageal intubation. Optimally the clinician will confidently visualise the ETT pass between the vocal cords and into the trachea. Often the view of the vocal cords is suboptimal. Regardless of operator confidence, confirmation of the ETT

placement is always required. A successful TI is likely when condensation appears in the ETT, breath sounds are heard in both axillae and not in the epigastrium, pulse oximetry is 100% and the chest cavity rises and falls with positive-pressure ventilation. At times, however, these findings can be equivocal, especially in children. The clinical exam is notoriously deceptive in determining correct ETT placement. It is imperative to use at least one of the following techniques for every intubated patient, being mindful that end tidal carbon dioxide is considered the gold standard for confirmation of placement:

1. Digital (wave form) capnography (Fig. 24.4.6). This technology continuously detects and displays the partial pressure of CO_2 within the ventilation circuit. In adult cardiac arrest patients, a pCO_2 <5 mmHg correlates with an extremely poor prognosis. Digital capnography is the gold standard for confirmation of intubations. In-line and side-stream attachments exist and are connected between the ETT and ventilator circuit (or manual ventilation device):

Box 24.4.2 End-tidal colour capnometer – interpreting the colour

- If you see Yellow, then 'Yes' – there is CO_2 return. At least 20 mmHg of CO_2 is detectable. Several studies show a 100% positive predictive value of a yellow indicator colour with a tracheal space intubation (rather than an oesophageal).
- If you see Purple, then there is a 'Problem' – there is no CO_2 return. A tracheal intubation is unlikely, unless the patient has extremely poor perfusion, such as during asystole. If a purple colour is displayed, less than 4 mmHg of CO_2 is detectable.
- If you see an intermediate 'Tan' colour, then 'Think about it', because the tube could be sitting in the trachea or the oesophagus. The capnometer is sensing between 4 mmHg and 15 mmHg of CO_2. This equivocal finding requires direct laryngoscopy re-visualisation of the tube's placement to help determine placement.

- Tracheal intubation. With each exhalation and inhalation, a characteristic waveform, showing the rise, plateau and fall of CO_2 levels, confirms correct positioning of the tube in the trachea.
- Oesophageal intubation. With each exhalation and inhalation, an unchanging, flat waveform demonstrates the absence of CO_2 and thus the misplacement of the ETT.

2. End-tidal colorimetric capnometry (Fig. 24.4.7). The capnometer attaches to the proximal end of the endotracheal tube and detects the presence of CO_2 within the tube. The capnometer will display a yellow (CO_2 present) or purple (CO_2 absent) colour in the indicator window, which generally correlates with a tracheal or oesophageal intubation, respectively. Multiple studies find that a yellow colour change has a 100% positive predictive value for correct ETT placement. When used for a poorly perfused patient (e.g. cardiac arrest), however, there will often be no yellow colour change because of low CO_2 levels. This is the primary limitation when using this modality. Colorimetric capnometry is not commonly used in hospital. Box 24.4.2 provides a mnemonic to help remember the colour scheme.

Indications

All intubated patients.

Contraindications

- Do not use an adult-sized colorimetric capnometry device on intubated patients weighing less than 15 kg, because the device adds a significant dead-space volume for the neonate to rebreathe CO_2. Instead, attach a paediatric-sized capnometer for these patients.
- Use caution with oesophageal aspiration on patients weighing less than 20 kg. Several studies, however, suggest that oesophageal aspiration may still be safe at a lower weight limit of 4 kg, because the resting lung volume is still greater than 50 cm^3, which is the typical volume in an oesophageal aspirator.

Equipment

1. Digital capnography (see Fig. 24.4.6)
2. End-tidal colorimetric capnometer (see Fig. 24.4.7)
3. Oesophageal aspiration syringe or bulb (see Fig. 24.4.8).

Preparation and positioning

- Prepare at least one confirmation device for bedside use before the patient is intubated so that immediate confirmation is available.
- Specific for the colorimetric capnometer, open the packaging for this disposable device. Check that the unit is dry and initially displays a purple colour in the indicator window to ensure proper functioning.
- Specific for digital capnography, be sure that the device is plugged in and turned on. Blowing across the capnography tubing tests the monitoring and sensing function.

Procedure

Digital capnography

1. Follow manufacturer instructions for calibration of devices prior to use
2. Connect the digital capnography between the ETT and ventilator circuit or BVM device. The air in the ETT is now contiguous with the air in this tubing.
3. Ventilate the patient and observe the real time waveforms of CO_2 return during exhalation. The absence of CO_2 indicates a failed intubation.

End-tidal colorimetric capnometer

1. Attach the capnometer between the ETT and the BVM device or ventilator circuit. Ventilation can continue through the capnometer.
2. After ventilating the patient for greater than six respiratory cycles, check the colour of the capnometer indicator window during the exhalation phase. The colour will change from purple to yellow in the event of a

successfully placed ETT.

3. Secure the capnometer in place if continuous monitoring is required. Most colorimetric devices will accurately monitor CO_2 for a period of 2 hours, after which the device must be discarded.

Complications

There are no direct complications to the patient by using these devices. Indirectly, however, the misinterpretation of their findings may cause the practitioner to incorrectly change patient management.

Tips

- Check tube placement after every intubation and after any patient movement, such as during transport, because the ETT can easily be displaced from between the vocal cords.
- Use a paediatric-sized colorimetric capnometer for patients less than 15 kg to decrease the volume of dead space in the ventilatory circuit.
- Be aware that confirmation with end tidal CO_2 (particularly colorimetric) may more difficult in a poorly perfused patient and false negative results are possible.

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24.5

The surgical airway

Holly Smith, and Scott Schofield

Background

The paediatric airway is different from that of the adult. The epiglottis is larger and floppy, while the larynx is shorter, more anterior and more cephalad. The larynx is also narrower and therefore more prone to obstruction by oedema, scarring, fluids or foreign bodies. The cartilage of the larynx is softer, which makes palpation of landmarks on the skin more difficult. The cricoid ring is the only level where cartilage encircles the paediatric trachea, and this is also the narrowest portion of the airway (Fig. 24.5.1). This is notably different from the adult airway where the narrowest point is at the level of the vocal cords.

The vast majority of paediatric airway emergencies can be readily managed with bag-valve-mask (BVM) ventilation and/or tracheal intubation (TI), with simple backup procedures to facilitate oxygenation and ventilation. Rarely, however, a child presents with complete airway obstruction or anatomic abnormalities that make oxygenation with BVM ineffective and/or TI impossible. This critical situation is referred to as ‘Can’t Intubate, Can’t Oxygenate’ (CICO), which has taken the place of the term ‘Can’t Intubate, Can’t Ventilate’ owing to the paramount importance of oxygenation over ventilation. In these situations, a method of front of neck access (FONA) may be necessary to provide life-saving oxygenation to avoid hypoxic brain injury. Methods of FONA in children include needle cricothyroidotomy, surgical cricothyroidotomy and rarely tracheostomy. Placing a needle, catheter or endotracheal tube (ETT) directly into the trachea through the neck will temporarily relieve airway obstruction or bypass anatomic abnormalities.

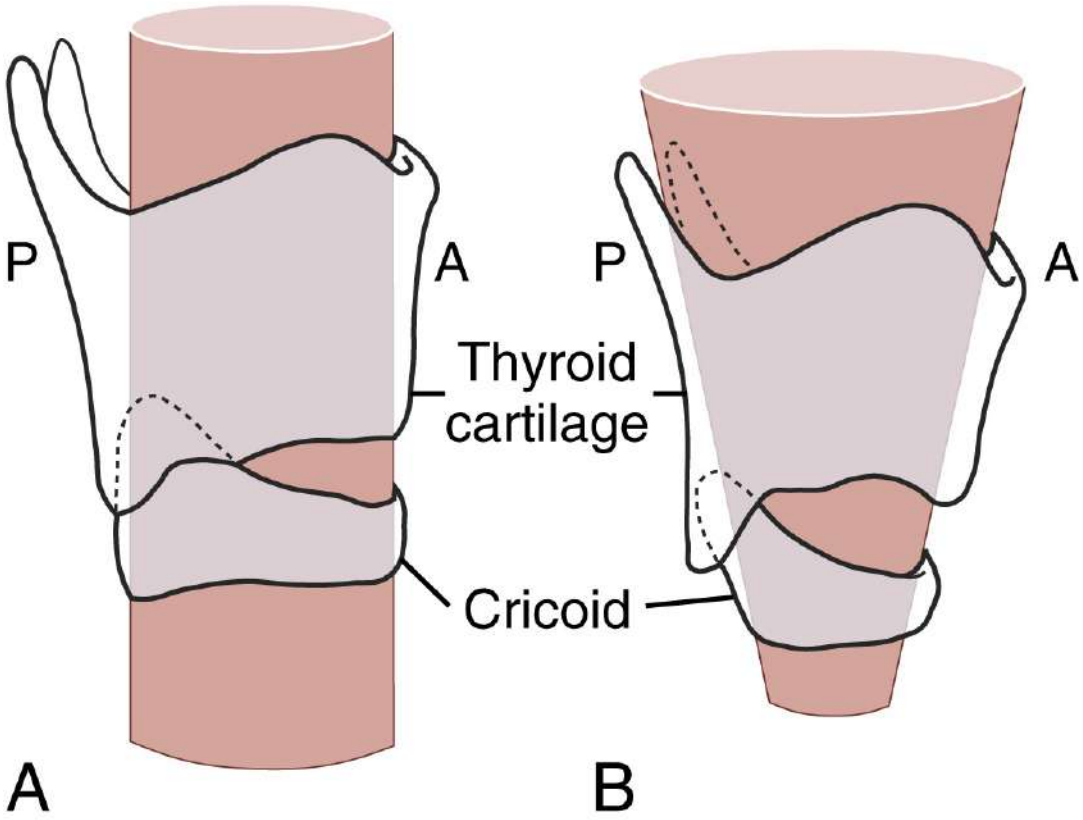

FIG. 24.5.1 The cricothyroid membrane. Comparison of adult (A) and paediatric (B) airways. Note funnel shape of paediatric airway with narrowest portion at cricoid ring.

Although this is a procedure that instils fear into most practitioners, it is important to acknowledge that in a true CICO situation, outcomes can be worse when practitioners fail to proceed to FONA when other methods have failed.

In children under the age of 5 years, the membrane between the thyroid cartilage and the cricoid cartilage, called the cricothyroid membrane (see Fig. 24.5.1), is extremely small. For this age group, needle cricothyroidotomy is easier than surgical cricothyroidotomy. While the needle technique will provide emergency oxygenation, it is unlikely to provide adequate ventilation, especially over time. Even a jet ‘ventilator’, a rescue device that attaches to the needle and provides intermittent bursts of high-pressure oxygen to simulate the normal respiratory cycle, may not allow for adequate ventilation and clearance of CO_2.

In children 5 years and over, surgical cricothyroidotomy becomes easier and more effective and creates a larger conduit to allow both immediate oxygenation and ventilation via a bag-valve device. Formal tracheotomy in a young child is extremely difficult to perform emergently and is rarely indicated in the emergency department.

Indications

- Failure to oxygenate with BVM with failed tracheal intubation
- Obstructed or disrupted larynx.

Box 24.5.1 lists situations in which these indications occur.

Contraindications

- Presence of a secure airway
- Traumatic destruction of the cricothyroid membrane
- Transection of the trachea with retraction of the distal segment
- Surgical cricothyroidotomy is *relatively* contraindicated in children under the age of 5 years. In this group, perform needle cricothyroidotomy and plan for definitive airway intervention, such as tracheostomy.

In the child who cannot be intubated from above and has a rising CO_2, but for whom oxygenation can be maintained with continuous positive airway pressure (CPAP), airway adjuncts and a high concentration of oxygen, preparation for FONA access should happen *concurrently* with active planning for definitive tracheostomy (rather than take precedence over it). Know your local referral procedures.

Needle cricothyroidotomy

Equipment

Box 24.5.2 lists the equipment for this procedure, which can be divided into three categories:

1. **Oxygen source.** A proprietary transtracheal jet ventilator device is ideal (example is the COOK Enk Oxygen Flow Modulator Set™). These provide high-pressure 100% off-wall oxygen (50 psi) in intermittent bursts (Fig. 24.5.2A). Several devices are available commercially and have been trialled with good effect. Alternatively, a setup that connects wall oxygen to the patient with an ability to modify the flow on/off. An example of this could include oxygen tubing attached to a three-way

stopcock (which would be occluded with your thumb to provide flow) (Fig. 24.5.2B). Finally, an anaesthetic bag connected to wall oxygen may provide sufficient pressure but will need an adaptor to attach to the cannula in the neck. A self-inflating bag will not generate enough pressure to overcome resistance in the small-diameter cannula.

Box 24.5.1 Situations potentially requiring cricothyroidotomy

- Trauma
    - Laryngeal fracture
    - Airway burns
    - Massive airway haemorrhage
    - Foreign body
    - Subglottic stenosis (late)
    - Burn contractures (late)
- Congenital abnormalities
    - Laryngeal atresia/stenosis/clefts
    - Tracheoesophageal fistula
    - Pierre–Robin syndrome
    - Treacher–Collins syndrome
- Cervical spine abnormalities
    - Trisomy 21 syndrome
    - Klippel–Feil malformation
    - Torticollis
- Inflammatory/infectious
    - Severe croup
    - Epiglottis
    - Bacterial tracheitis
    - Retropharyngeal abscess
    - Cricoarytenoid arthritis
- Laryngeal spasm

Box 24.5.2 Equipment for needle cricothyroidotomy

- Oxygen source
  - Proprietary jet ventilator (50 psi)
  - Oxygen tubing to 3-way stopcock attached to wall oxygen and flowmeter (up to 15 L min^{-1})
  - Anaesthetic bag-valve-mask device (T-piece) attached to wall oxygen and flowmeter (up to 15 L min^{-1})
- Adaptor
  - 3 mm endotracheal tube adaptor to be used with anaesthetic bag
- Catheter
  - 14 gauge catheter over a needle

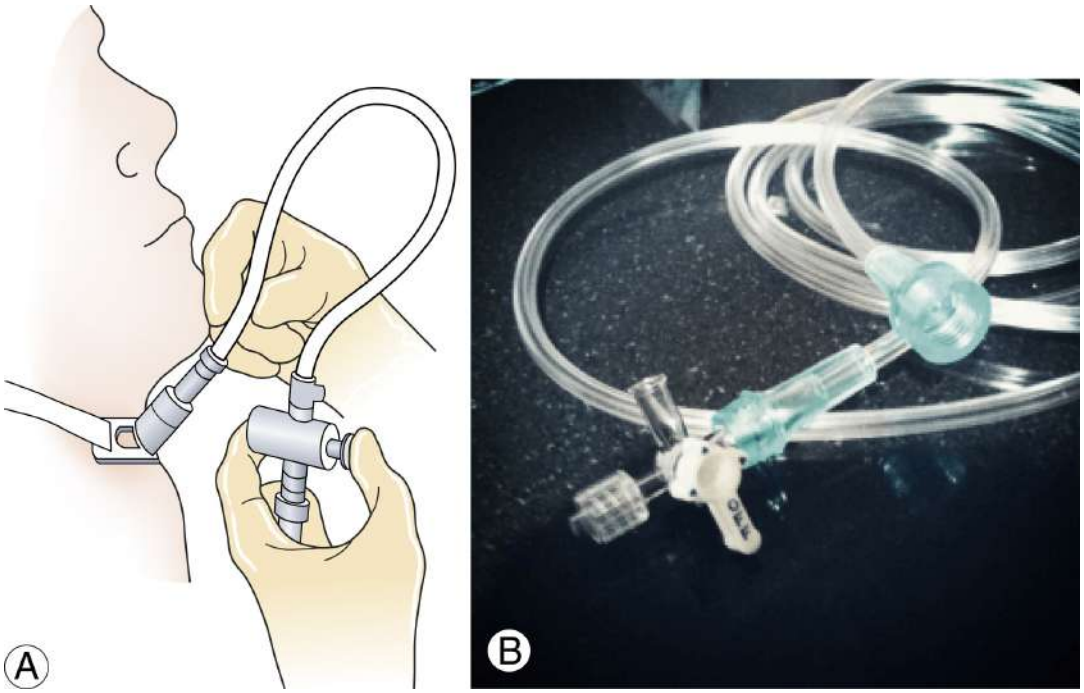

FIG. 24.5.2 Transtracheal jet ventilation setups (A) connected at neck. (B) Oxygen tubing to 3-way stopcock. EMCrit, Metasin LLC (http://emcrit.org/podcasts/cricothyrotomy-needle-or-knife/).

More important than what actual device is used in your setting, it must be accessible, and team members must know how to use the device available to them. Many online references are available regarding their use.

2. **Adaptors.** Proprietary jet ventilator devices connect directly to the catheter at the neck so no adaptor is required. Similarly, oxygen tubing to a three-way stopcock setup will attach directly to the cannula at the neck. To use an anaesthetic bag (T-piece) off-wall oxygen, attach a 3 mL Luer-Lok syringe barrel to the cannula at the neck. An 8.0 ETT adaptor will fit snugly into the barrel and connect to a bag (Fig. 24.5.3). Be careful not to dislodge the catheter in the neck.
3. **Needle/cannula** for access through the front of neck. A 14 gauge over-the-needle catheter (i.e. intravenous cannula) is suitable.

Preparation

1. Since children have relatively large occiputs, place a shoulder roll transversely to prevent hyperflexion of the neck and improve visualisation of the anterior neck anatomy (except in the setting of major trauma when the integrity of the C-spine is in doubt).
2. If time allows, prepare the skin with surgical cleaning solution.

Procedure

1. Wash hands and don gloves.
2. Palpate the hyoid bone high in the neck, and move caudally to identify the thyroid and cricoid cartilages (Fig. 24.5.4A).
3. Stabilise the cartilage using the thumb and index finger of the non-dominant hand.
4. Insert a 14 gauge over-the-needle catheter attached to a 3 mL syringe, just superior to the cricoid cartilage (Fig. 24.5.4B). Angle the catheter caudally towards the cricothyroid membrane at about a 45 degree angle to the skin.
5. Continuously aspirate on the syringe while advancing. A rush of air into the syringe confirms entry into the trachea. You may choose to fill the syringe with saline to use as an indicator. Entry into the trachea is

marked by an influx of air bubbles into the saline-filled syringe.

6. Advance the catheter into the trachea over the needle, and remove the needle and syringe. Reattach the syringe to the catheter to confirm that air still bubbles into the syringe, ensuring correct placement. Do not let go of the catheter until it is secured or an alternative airway is obtained.
7. If a proprietary jet ventilator device is available, attach it directly to the catheter. If this device is not available, attach an anaesthetic bag-valve system to high-flow wall oxygen. Place an adaptor as noted above between the catheter and the bag-valve device (e.g. a 3 mm ETT adaptor directly onto the catheter).
8. Commence with a wall oxygen flow of 1 L/min (LPM) per year of age on the flow meter. An initial occlusion of 2–4 seconds is advised, assessing for chest rise. The chest will fall slowly following an effective insufflation. Repeat occlusion/inspiration of 1 second's duration every 4 seconds (1:4 ratio). Watch for chest rise and slow fall. If no chest rise is achieved, increase the flow by 1 LPM (i.e. not by prolonging the occlusion) with each breath until chest rise is achieved. Note the risk for breath stacking and barotrauma; observe the chest diameter for evidence of stacking, watch for subcutaneous emphysema, be ready to perform needle thoracentesis. This risk, however, should not preclude you from trying to achieve chest rise and oxygenation.

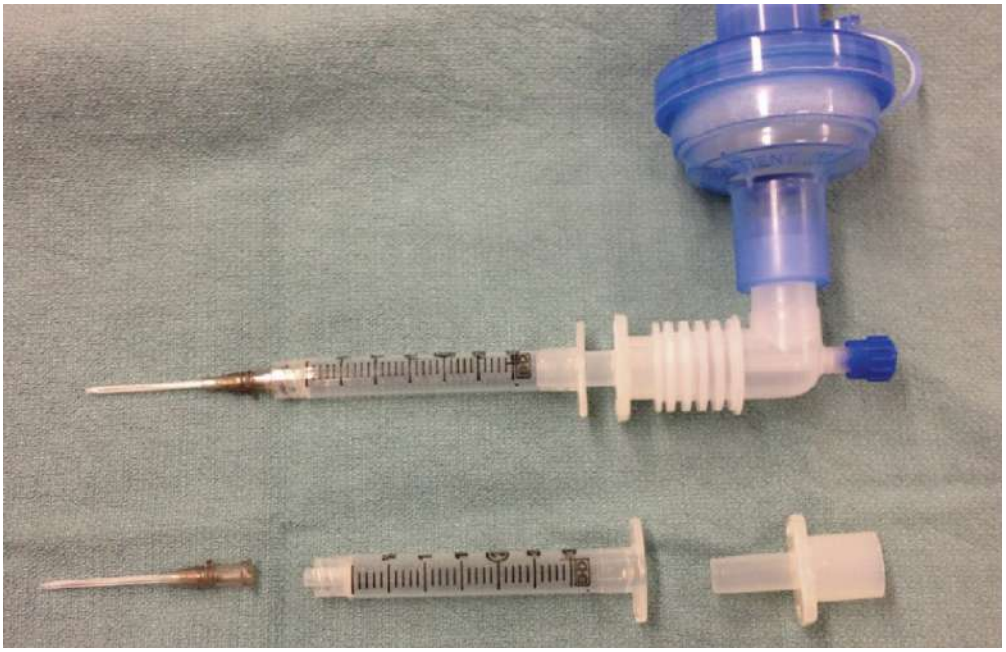

FIG. 24.5.3 Using an 8.0 ETT adaptor and a 3 mL Luer-Lok syringe barrel to attach to an anaesthetic bag. With permission from Patel SA, Meyer TK. Surgical airway. *Int J Crit Illn Inj Sci* [serial online] 2014 (http://www.ijciis.org/text.asp?2014/4/1/71/128016).

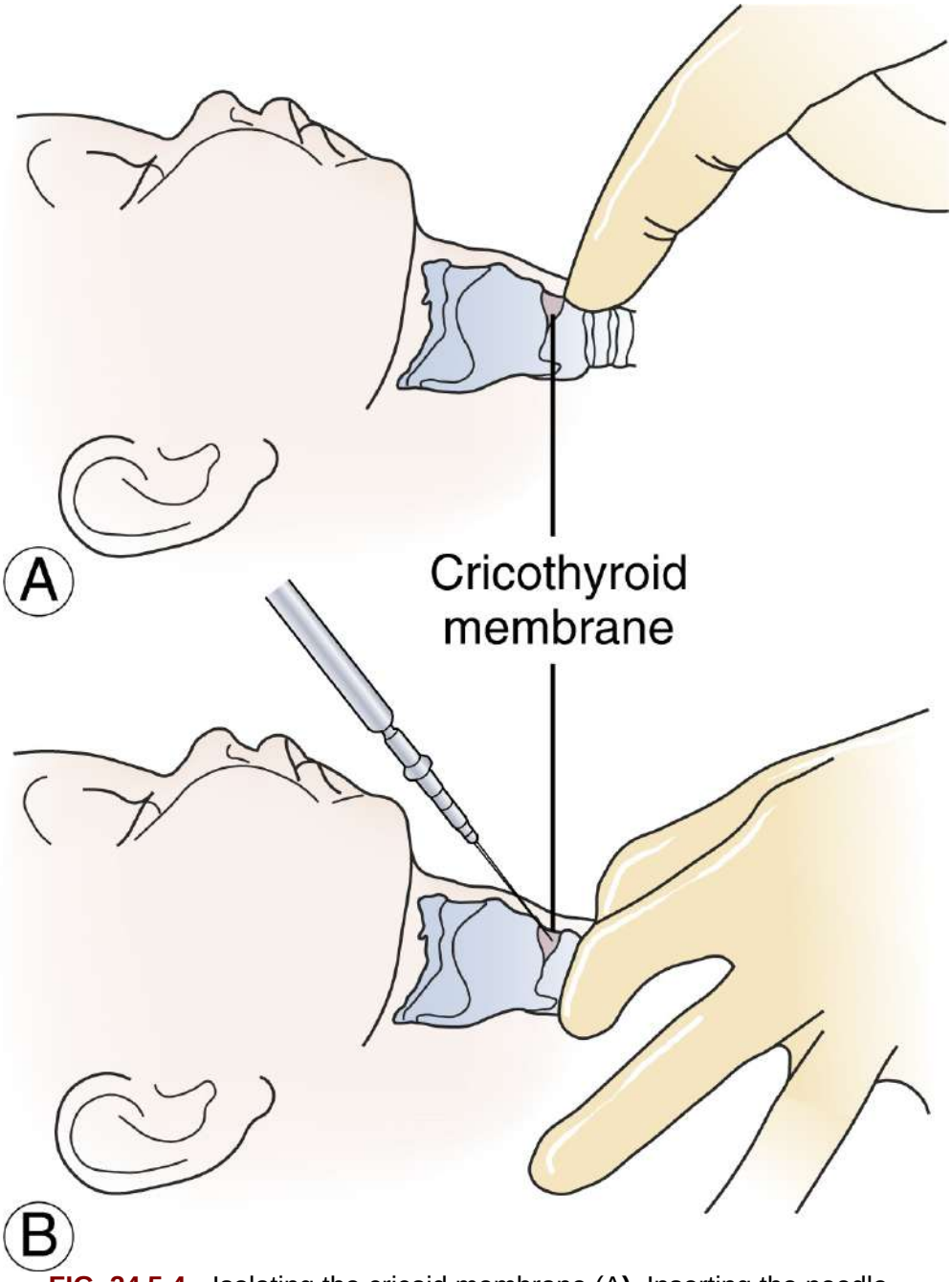

FIG. 24.5.4 Isolating the cricoid membrane (A**).** Inserting the needle through the membrane (B).

Complications

- Bleeding, usually local, rarely large volume
- Inappropriate placement of a needle can cause injury to:
    - the larynx and vocal cords
    - great vessels of the neck
    - nerves
    - oesophagus

  and creat a false tract
- High-pressure oxygen can also cause significant barotrauma resulting in extensive subcutaneous emphysema, pneumothorax and pneumomediastinum.

Tips

- Needle cricothyroidotomy is a temporising measure that may provide enough oxygenation for about 30 minutes. Immediately consider other airway options, such as definitive tracheostomy; know your local referral patterns.
- Remember that the goal through a needle cricothyroidotomy is primarily oxygenation and that great strides are unlikely to be made in ventilation.
- Most exhalation will actually occur through the (obstructed) upper airway. Although it seems counterintuitive, in most cases, the path of least resistance for efflux of air will be via this route rather than through the tiny catheter in the neck. Because of this, any measures to optimise the airway should still be maintained, including positioning and adjuncts.
- A self-inflating bag is not able to generate enough pressure to provide oxygenation through a needle cricothyroidotomy. This is due to the small caliber of the catheter in the neck and extremely high resistance therein. A self-inflating bag is pressure limited at 35 cm H_2O, whereas an anaesthetic bag (T-piece) setup or direct wall oxygen source via a flow meter can generate much higher pressures:
    - Maximum pressure through self-inflating bag = 35 cm water
    - Wall oxygen through flow meter = approximately 4000 cm water (>50 psi)

- Remember to allow adequate time for passive exhalation to prevent barotrauma (minimum exhalation is 4 seconds, may require up to 8 seconds).
- Do *not* let go of the catheter until an alternative airway is established.

Surgical cricothyroidotomy

Equipment

Box 24.5.3 lists the potential equipment for surgical cricothyroidotomy.

Several commercial kits contain all equipment for a cricothyroidotomy tube using the guidewire or Seldinger technique. These will be addressed next.

There are many methods to performing a surgical cricothyroidotomy. A widely accepted method is the knife-finger-bougie technique or some variation thereof.

Preparation

1. Expose the anterior neck as above.
2. Prepare the skin with a surgical cleaning solution.

Procedure

1. Wash hands and don gloves.
2. Palpate the hyoid bone high in the neck, and move caudally to identify the thyroid and cricoid cartilages (Fig. 24.5.4A).
3. Stabilise the larynx by placing thumb and forefinger of the non-dominant hand on either side of the thyroid cartilage.
4. Make a midline vertical incision in the skin from thyroid cartilage to cricoid cartilage (Fig. 24.5.5). Expose the cricothyroid membrane via blunt dissection with a haemostat or a gloved finger.
5. Make a horizontal incision in the cricothyroid membrane, carefully avoiding inserting the scalpel too deeply and lacerating the posterior aspect of the larynx.

Box 24.5.3 Equipment for surgical

cricothyroidotomy

- Suction device (i.e. Yankauer)
- Scalpel
- Haemostat
- Tracheostomy or endotracheal tube (age-appropriate size)
- Oxygen delivery system (same as for
- Endotracheal suction catheter

Optional

- Surgical prep solution: 0.5% chlorhexidine in 70% alcohol or povidone-iodine solution
- 1% Lidocaine (lignocaine) with adrenaline (epinephrine)
- Tracheal hook
- Tracheal dilator

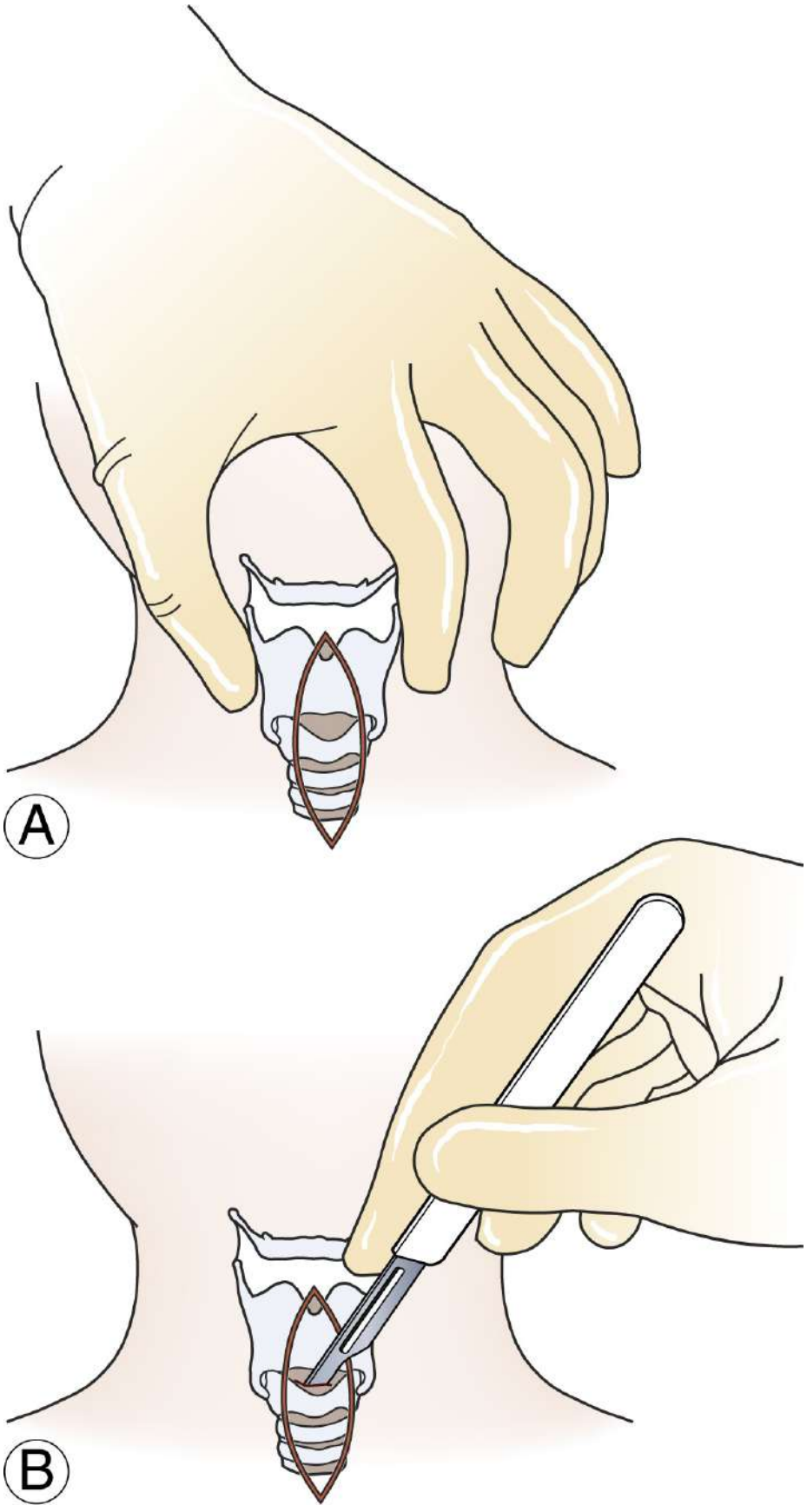
A
B

FIG. 24.5.5 Making the surgical incision in the membrane.

6. Insert your gloved small finger (or the haemostat) into the airway next to the scalpel to hold the space and widen the opening. Remove the scalpel, maintaining the opening with your finger (or haemostat).
7. Insert an appropriate-sized bougie into the tracheal opening, passing it caudally until resistance is met (at the carina). Remove your finger and pass an age-appropriate cuffed endotracheal tube over the bougie into the airway (this should be preloaded). Inflate the cuff and gently remove the bougie (Fig. 24.5.6).
8. Attach a bag-valve device and ventilate the patient. Do not let go of the tube until it has been secured to the patient's neck.

Guidewire or Seldinger technique

The **guidewire or Seldinger technique** is an alternative to the above method of surgical placement of a cricothyroidotomy tube. The procedure begins as with needle cricothyroidotomy yet results in the placement of a larger bore tube. The Cook Melker Cricothyroidotomy Kit™ is a common example. This method can be used primarily by experienced staff or can be used to upgrade an existing needle cricothyroidotomy. Note that primary success rates using Seldinger kits are not as high as for simple needle cricothyroidotomy or for surgical cricothyroidotomy.

Preparation and procedure

1. Position the patient as for surgical cricothyroidotomy.
2. Identify the cricothyroid membrane and prepare the skin with surgical cleaning solution.

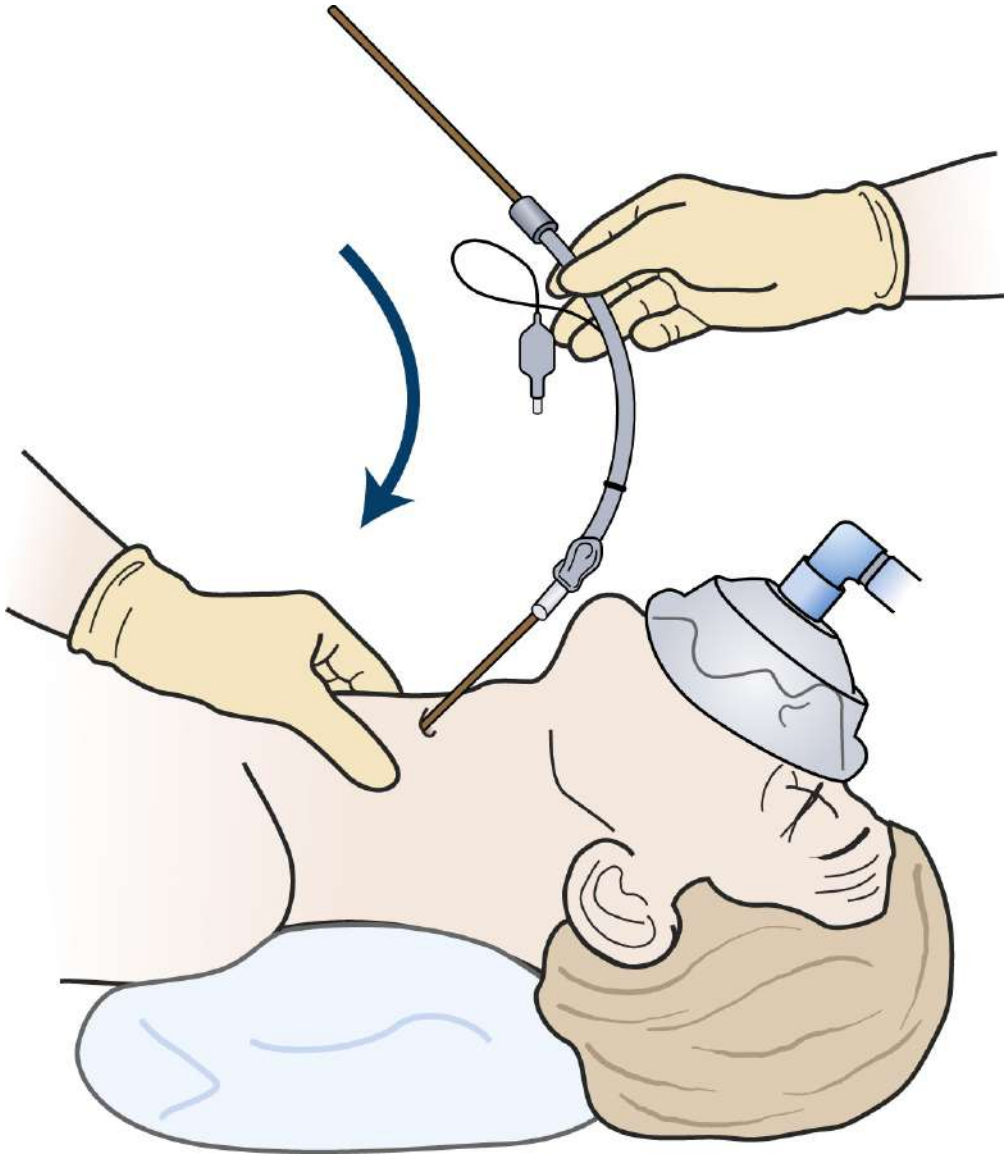

FIG. 24.5.6 Inserting the cricothyroidotomy tube over bougie. Difficult Airway Society 2015 guidelines for management of unanticipated difficult intubation in adults, C. Frerk, V. S. Mitchell, A. F. McNarry, C. Mendonca, R. Bhagrath, A. Patel, E. P. O'Sullivan, N. M. Woodall, I. Ahmad. Difficult Airway Society intubation guidelines working group. *Br J Anaesth* (2015) 115 (6): 827-848.

3. Insert the needle mounted on a syringe through the cricothyroid membrane using the same method as for needle cricothyroidotomy.
4. Angle caudally at 45 degrees to the skin with the bevel of the needle up.
5. Once air rushes into the syringe, remove the syringe and pass the J-tipped guidewire through the needle, then remove the needle. Alternately, the J-tipped guidewire can be passed through the existing needle cricothyroidotomy. Always maintain a hold of the guidewire; this

may require an assistant.

6. Make a 1 cm vertical stab incision along the guidewire through the skin (this step is essential because the skin poses significant resistance to the introducer and tube, increasing the chance of injury and false tract).
7. Insert the tube and dilator unit over the guidewire with a firm screwing motion into the trachea. Follow the curve of the tube and dilator, and hold both the tube and the dilator together as they can slip apart. The flange of the tube should rest against the skin. Have an assistant maintain a secure hold of the guidewire throughout this step. Remove both the wire and the dilator.
8. Provide ventilation through the tube via a bag-valve device.
9. Inflate the cuff if applicable and secure the tube.

Complications

- Bleeding
- Tube misplacement (false tract), resulting in hypoxia and subcutaneous emphysema
- Laryngeal, oesophageal or neurovascular injury
- Barotrauma, which can result in pneumothorax or pneumomediastinum
- Significant late complications include voice change due to vocal cord damage and subglottic stenosis.

Tips

- If the cricothyroid membrane is not visible with the initial vertical incision, extend the incision vertically in both directions.
- Do not cut into the airway blindly as this will increase the rate of complications.
- Have suction readily available for any bleeding into the trachea; however, the amount of bleeding is rarely prohibitive. Bleeding is usually tamponaded once the tube is in place.
- Avoid using too large a tube. This can lead to laryngeal injury. Choose an age-appropriate size as for endotracheal intubation—i.e. use a formula guide or a length-based resuscitation tape.
- If a child requires prolonged ventilation, establish a definitive airway

(tracheal intubation or tracheostomy) as soon as possible.

- Remember that in the CICO situation, more harm is done by NOT proceeding to FONA. In these rare situations, the benefits of action far outweigh the risks. Activate plans for a definitive airway and get help early.

Further reading

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24.6

Chest procedures

Scott Schofield, and Holly Smith

Introduction

Trauma can cause a number of life-threatening chest injuries in children including tension pneumothorax, open pneumothorax, massive haemothorax and pericardial tamponade. It is imperative to have the knowledge and clinical skills required for a number of critical chest procedures for the emergency management of these traumatic pathologies. The same chest procedures are often also required for non-traumatic aetiologies.

Needle Thoracostomy

Background

Normally, the visceral and parietal pleura are closely adherent to one another. However, if either surface is violated, air enters into the potential space between visceral and parietal pleura, creating a simple pneumothorax (Fig. 24.6.1). This typically occurs in the setting of blunt or penetrating trauma. Occasionally, spontaneous pneumothorax occurs, as with excessive air trapping in an asthmatic child. If enough air collects, a tension pneumothorax can develop in which pressure in the space shifts the mediastinum towards the ipsilateral side, impedes venous return to the heart and decreases cardiac output (Fig. 24.6.2). Another pathophysiological mechanism for tension pneumothorax occurs when a patient with a simple pneumothorax is intubated and ventilated. Positive pressure ventilation can force air into the pleural space, resulting in a tension pneumothorax. A tension pneumothorax can progress to shock and cardiopulmonary arrest.

The typical clinical scenario for tension pneumothorax is the child who experiences a penetrating injury to the chest wall or back. The classical clinical

findings of unilateral decreased air entry, hyper-resonant percussion note and tracheal deviation (away from the pathology) may be difficult to appreciate. Other clinical signs are tachypneoa, oxygen desaturation, respiratory distress (retractions, flaring, grunting) and shock. Once tension pneumothorax develops progression to shock and cardiopulmonary arrest can be rapid.

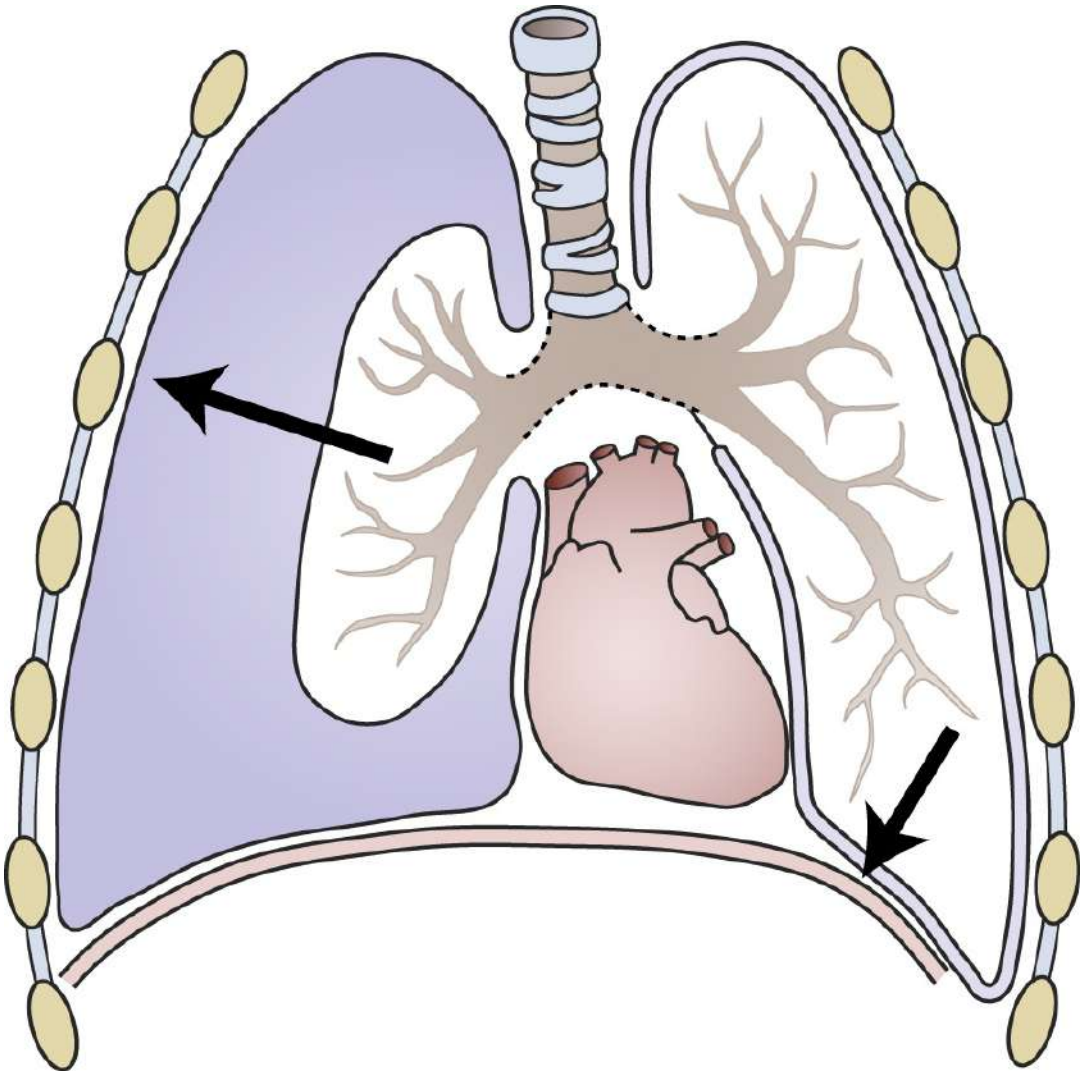

FIG. 24.6.1 Simple pneumothorax.

Evacuation of the air between the visceral and parietal pleura is time critical and paramount. The conversion of the tension pneumothorax to an open pneumothorax with a thoracostomy (needle, finger or tube) will re-expand the lung and improve venous return and cardiac output. Needle thoracostomy is a temporising procedure only. It can be done quickly with minimal equipment and

preparation. As soon as practicable after needle thoracostomy, a formal thoracostomy should be performed with insertion of a chest tube to provide ongoing drainage in a closed system. The risk of failing to perform a needle thoracostomy in a child decompensating from a tension pneumothorax is far greater than the risk of performing the procedure in a child without a pneumothorax. There is no place for hesitation to perform this simple procedure if tension pneumothorax is thought to be present.

Indications

- Blunt or penetrating trauma with clinical signs of tension pneumothorax
- Chest X-ray evidence of a tension pneumothorax in a child in respiratory distress (although clinical diagnosis should preempt and negate the chance of radiological diagnosis)
- Suspected tension pneumothorax in an intubated patient who is rapidly deteriorating (e.g. hypotension, hypoxia)
- Pulseless electrical activity (PEA) in the setting of trauma or asthma.

Contraindications

- A stable child with a simple pneumothorax (i.e. not under tension)
- There is no absolute contraindication to needle thoracostomy in a child with possible tension pneumothorax and shock. While preparing for a formal thoracostomy in the emergency department, performing a needle thoracostomy will not cause further deterioration.

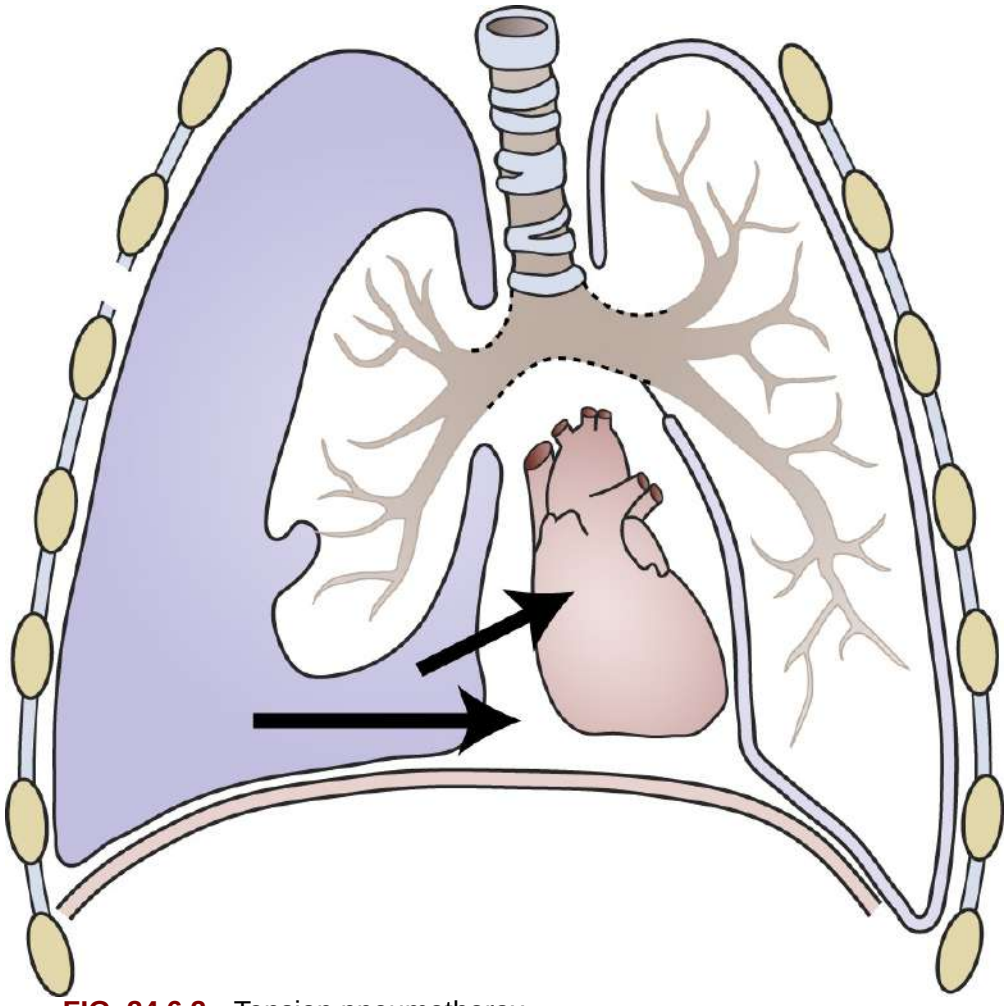

FIG. 24.6.2 Tension pneumothorax.

- If a skin infection is present at the preferred needle site, select an alternative location.

Equipment

1. 14 gauge cannula
2. 10–30 mL syringe
3. Cleaning (alcohol or chlorhexidine) solution/wipe.

Preparation

1. Inspect the anatomy of the ribs, and select an entry site immediately above the rib, avoiding the intercostal neurovascular bundle (Fig.

24.6.3).

2. Position the child supine with the head of the bed angled up at 30 degrees. Have an assistant gently restrain the conscious child.
3. Identify the 2nd intercostal space (above the 3rd rib) at the midclavicular line. This space is ideal since air in the pleural space rises and typically collects towards the top of the lungs. A lateral approach is an alternative but may have more complications, including lung penetration and subsequent adhesions.
4. Prepare the skin of the entry site with cleaning solution or wipe.

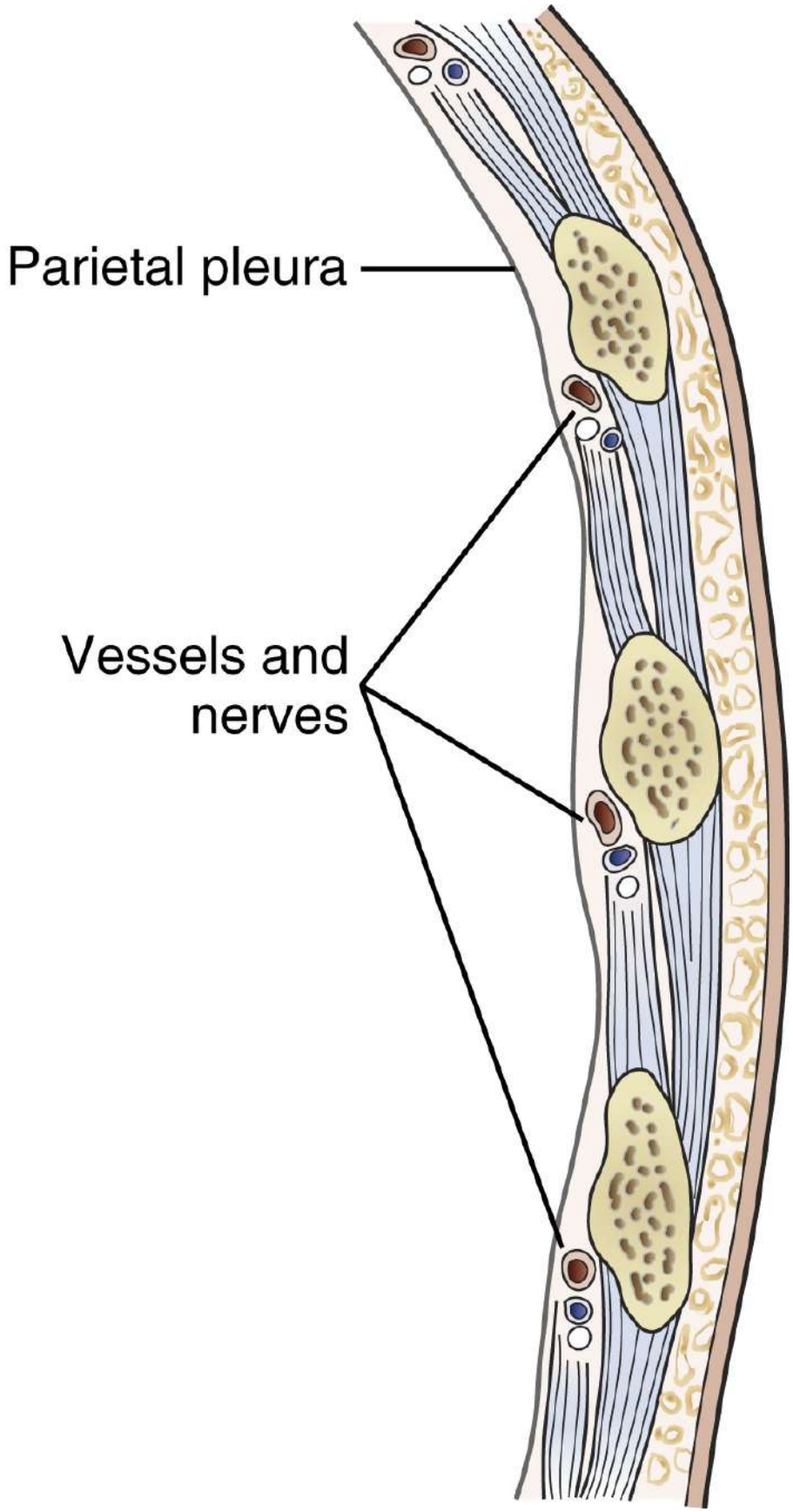
Parietal pleura
Vessels and nerves

FIG. 24.6.3 Anatomy of the ribs and intercostal neurovascular bundle.

5. Use a local and/or intravenous analgesic if time allows and if the child is conscious.

Procedure

1. Attach the cannula to the syringe (some clinicians pre-draw a small amount of sterile water or normal saline in the syringe).
2. Insert the catheter through the chest wall at a 90 degree angle (perpendicular to the chest wall) above the 3rd rib (2nd intercostal space) at the midclavicular line (Fig. 24.6.4A).
3. Provide back pressure on the syringe until air is aspirated (if water is pre-drawn you will see bubbling in the syringe). Once there is a free flow of air into the syringe, continue to pull back on the plunger to evacuate the air.
4. Remove the syringe and needle, leaving the catheter in place (Fig. 24.6.4B).
5. Progress to a formal finger or tube thoracostomy as soon as possible.

Complications

- Performing needle thoracostomy in a patient without a pneumothorax can cause a pneumothorax or injury to the lung parenchyma. Under positive pressure ventilation, this pleural penetration can result in an ongoing air leak, which can then lead to a tension pneumothorax.
- Placing the needle below the rib can cause injury to an intercostal artery and subsequent haemothorax.
- Placing the catheter lower in the chest or closer to the mediastinum can result in diaphragm penetration, bowel penetration, haemopericardium or coronary vessel injury.
- The catheter can easily become occluded and fail to drain air, allowing the pneumothorax to re-accumulate and re-tension.

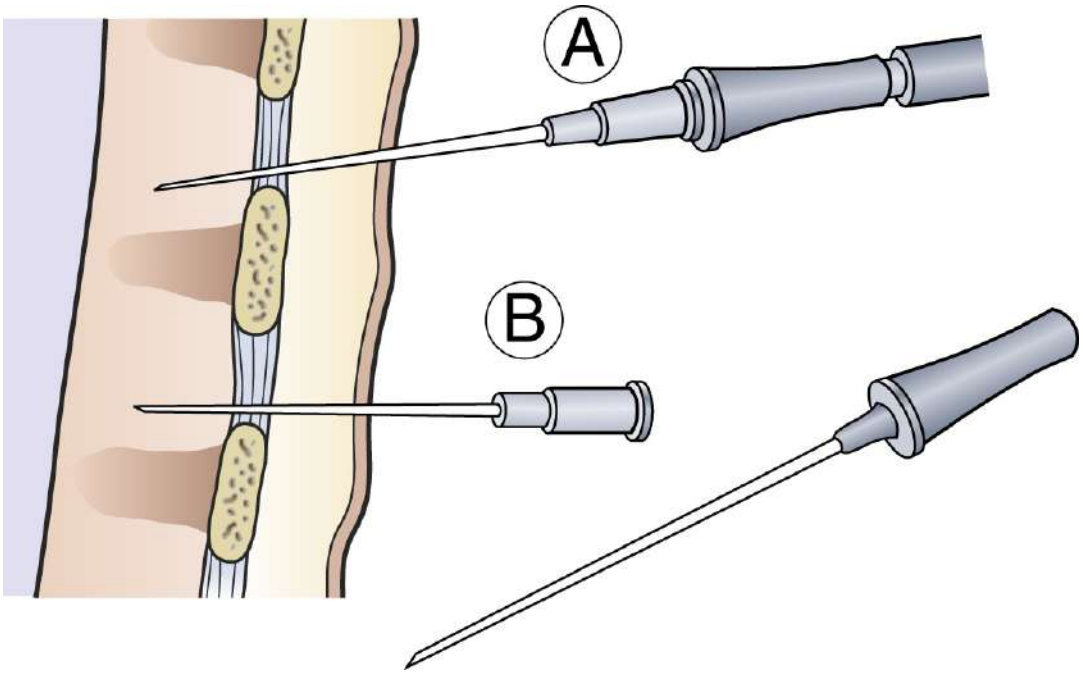

FIG. 24.6.4 Insertion of thoracostomy needle (A) and removing needle, leaving catheter in place (B).

Tips

- Perform tube thoracostomy as soon as possible in the setting of suspected tension pneumothorax. This relieves tension and creates an open pneumothorax.
- Always perform tube thoracostomy after needle thoracostomy to prevent tension pneumothorax from developing.

Tube Thoracostomy

Background

Traumatic pneumothorax and haemothorax are the most frequent paediatric indications for tube thoracostomy. Pneumothorax is a serious sequela of major chest trauma in children and may occur as a result of blunt injury without rib fractures or chest penetration. Sometimes, spontaneous pneumothorax occurs without trauma, from rapid increase in intraluminal pressure, especially with patients who have pre-existing bronchopulmonary disease. Air or fluid in the pleural space can cause significant impediments to oxygenation/ventilation and

drastically reduce cardiac output, usually when the haemothorax is large or the pneumothorax is under tension. Tension pneumothorax and massive haemothorax are life-threatening emergencies that require immediate chest decompression. Occasionally, large pleural fluid collections from non-traumatic aetiologies will require tube thoracostomy for drainage.

Tube thoracostomy is the insertion of an intercostal chest tube to evacuate air and/or fluid from the pleural space. The main differences in the procedure between adults and children are the need for more meticulous patient preparation and smaller catheters required in children. Sometimes, detection of pleural air or fluid is more difficult in children, requiring ultrasound or CT imaging to localise and quantify.

In the setting of tension pneumothorax, needle thoracostomy may precede tube thoracostomy, but tube thoracostomy must always follow needle thoracostomy. Small stable pneumothoraces not under tension (<20–30%) are usually managed with observation alone. There is an increasing tendency to manage even moderate-sized pneumothoraces without tension conservatively. Massive haemothorax occurs when a large vascular structure under systemic pressure bleeds into the pleural space. Surgical assessment for ongoing blood loss is imperative. Red cell transfusion as well as massive transfusion protocol with the addition of platelets and fresh frozen plasma may be required.

Indications

- Large/tension pneumothorax
- Haemothorax
- Rapid pleural fluid accumulation with respiratory distress.

Contraindications

Need for immediate open thoracotomy.

Equipment

- Chest tube tray (Box 24.6.1)
- Appropriate-sized chest tube (Table 24.6.1)
- Pigtail chest tubes (for pneumothorax or pleural effusion only)

- Chest tube drainage device (underwater seal drain, duckbill valve, etc.)
- Wall suction.

Preparation

1. Identify the presence of pneumothorax/haemothorax and the affected side by clinical examination and/or chest X-ray.

Box 24.6.1 Chest tube equipment tray

- Kelly clamps (2)
- Mayo scissors
- Suture scissors
- Needle holder
- Scalpel
- Forceps
- Silk suture
- 10 mL syringe
- 25 gauge needle
- 20 gauge needle
- Sterile towels
- 4 x 4 sterile gauze
- Vacuum device
- Drainage apparatus with water seal
- Plastic connectors (straight and Y type)
- Chest tubes
- Petrolatum
- Lidocaine or bupivacaine for local injection
- Antiseptic prep solution
- Wide cloth tap

Table 24.6.1

Sizing chest tubes

Age	Chest tube size (Fr)
Newborn	8–12
Infant	14–20
Child	20–28
Adolescent	28–36

Formula: chest tube size (in Fr) = 4 × endotracheal tube size (mm).

Use larger tube size for haemothorax.

2. Establish secure vascular access and apply cardiac monitor and pulse oximetry.
3. If the child is conscious, provide parenteral sedation/analgesia.
4. If the child is stable, consider an intercostal nerve block.
5. Explain the procedure to the verbal child.
6. Place a nasogastric tube to decompress the stomach, when there is abdominal distension.
7. Place the child in the supine or upright position with the arm raised on the affected side.
8. Identify the entry site (Fig. 24.6.5), anterior to the mid-axillary line between the fourth and fifth (fourth intercostal space) or fifth and sixth rib (fifth intercostal space), at or above the nipple level.
9. Drape and clean the area widely with povidone-iodine or chlorhexidine solution.
10. Infiltrate the entry site above the rib and into the intercostal space generously with lidocaine in conscious patients.
11. Select a properly sized standard chest tube or pigtail catheter (see Table 24.6.1).

Procedure

Inserting the tube

1. Make a horizontal incision over the rib that is below the selected intercostal space, long enough to easily fit the appropriate-sized chest tube (1–3 cm).
2. Insert a curved Kelly forceps into the incision, and bluntly dissect the

tract by advancing and opening the forceps. The tract should be dissected over the rib (to avoid the neurovascular bundle) into the interspace (Fig. 24.6.6).

3. Continue to blunt dissect through intercostal muscle, and then puncture the parietal pleura, ensuring the forceps are not allowed to penetrate too deep.
4. Widen the hole by opening and removing the opened forceps. Then insert a gloved finger into the hole and sweep it 360 degrees inside the pleura to assure the tract is clear and the lung is not adhered to the chest wall.
5. Insert the appropriately sized intercostal tube into the tract, and direct the tube posterior and inferior for a haemothorax or anterior and superior for a pneumothorax.
6. Facilitate tube entry by grasping the tube with the Kelly to guide tube entry into the pleural space. If using a fenestrated catheter, ensure the tube is inserted beyond the last fenestration.
7. Attach the tube to the drainage system.

Securing the tube

1. Use 2-0 silk to secure the tube.
2. Employ a purse-string suture, wrapping the suture material multiple times in opposite directions around the tube prior to tying off the suture (Fig. 24.6.7). Consider repeating this multiple times. This will ensure secure fixation of the catheter and allow removal of the tube and closure of the incision without additional suturing.
3. Cover the site with a sterile, impermeable dressing.
4. Secure all tube connections.
5. Obtain an X-ray to confirm tube placement and decompression of air or fluid.

Complications

- Haemopericardium
- Tension pneumothorax
- Haemothorax
- Myocardial injury

- Diaphragm perforation
- Solid organ injury
- Bowel injury
- Cardiopulmonary arrest.

Tips

- Sizing a chest tube involves estimation of the size of the child. If an exact size is not known, use the following formula: chest tube size = 2 x nasogastric tube size or 4 x endotracheal tube size.
- Use a smaller pigtail catheter if the pneumothorax needs to be evacuated but is not causing significant tension.
- Use a larger tube for haemothorax.
- Monitor for air leak for evidence of system air leak or tracheobronchial injury.

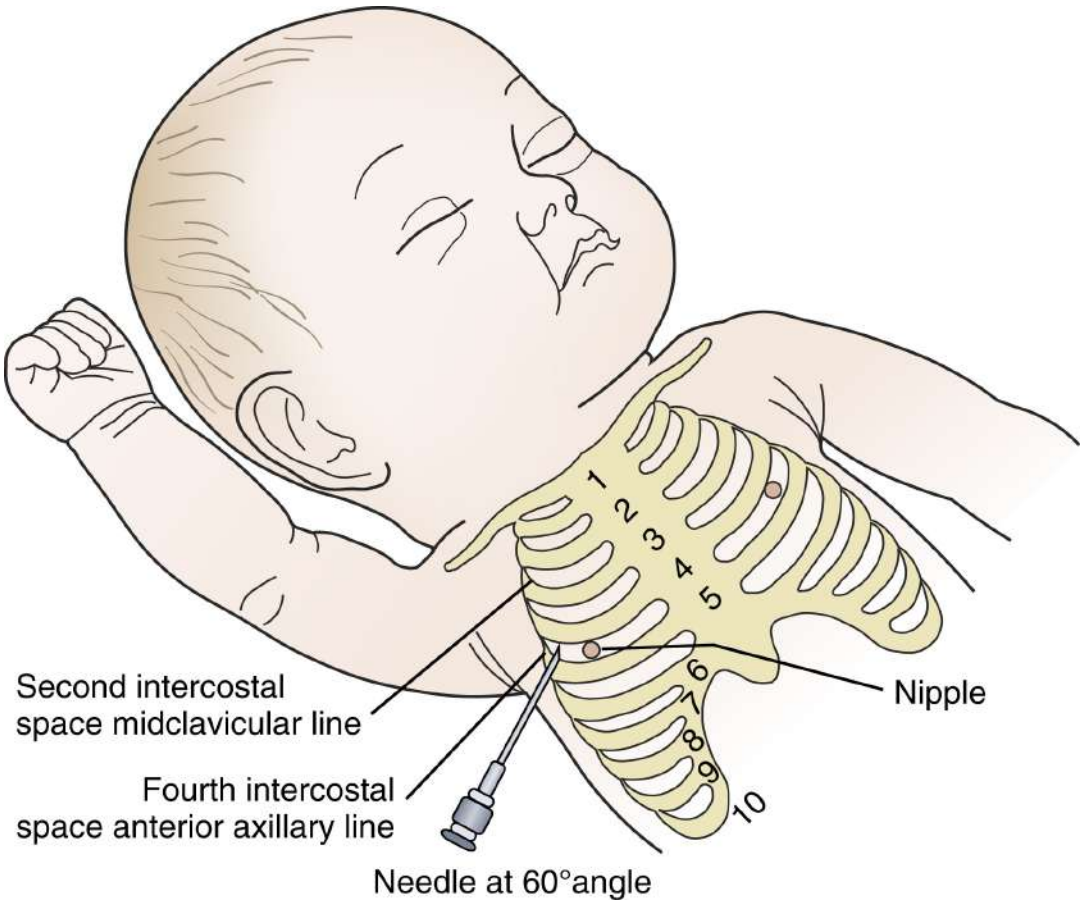

FIG. 24.6.5 Entry site for chest tube.

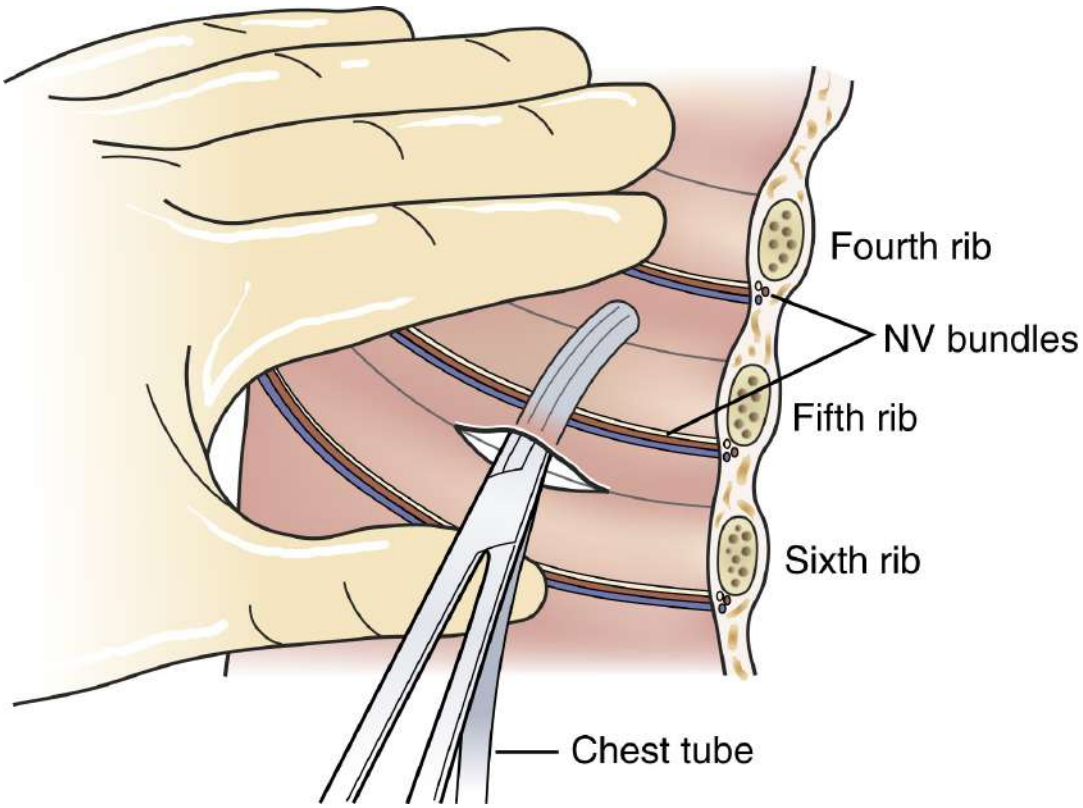

FIG. 24.6.6 Inserting the chest tube.

- Avoid trocars, to minimise lung lacerations.
- If a child with a penetrating chest injury is in shock or respiratory failure, do not wait for a chest X-ray before performing tube thoracostomy.
- Use of CT or video imaging may assist chest-tube placement.

Three-Sided Dressing

Background

Penetrating trauma may cause an open pneumothorax, also known as a sucking chest wound. In this injury air follows the path of least resistance, entering the thoracic cavity through the injured chest wall instead of entering the lungs through the airways during inspiration. Air can become trapped in the thoracic cavity, quickly progressing to a tension pneumothorax.

Emergency treatment of an open pneumothorax involves covering the wound with a three-sided dressing to create a valve that prevents air entering the chest cavity through the chest wall.

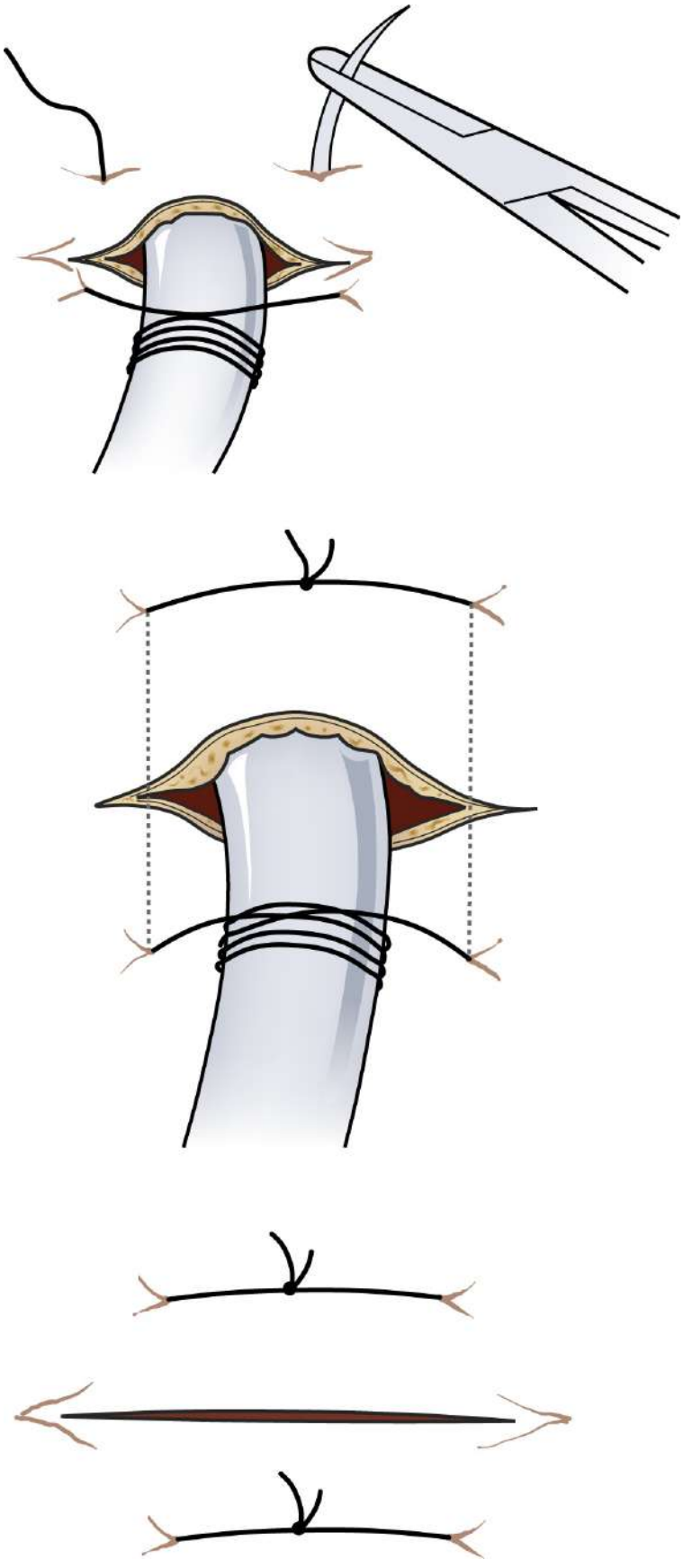

FIG. 24.6.7 Securing the chest tube.

FIG. 24.6.8 Treating a sucking chest wound.

Indications

- Open pneumothorax/sucking chest wound.

Contraindications

- Allergy to tapes/dressings.

Equipment

- Air occlusive dressing
- Adhesive tape.

Preparation

- Clean and dry area of skin around wound to improve adhesion of dressing edges.

Procedure

- Cut air occlusive dressing material to size, allowing extension 6–8 cm from wound edge on all sides.
- Tape three sides of dressing to skin, leaving one side free for egress of air. This allows air to be exhaled, escaping from underneath the dressing. During inspiration negative intrathoracic pressure causes the dressing to occlude the wound and prevents air entering the thoracic cavity (Fig. 24.6.8).

Pericardiocentesis

Background

The heart is extremely sensitive to rapid accumulation of pericardial fluid. Increased pressure from an acute pericardial effusion or haemopericardium may significantly impede venous return and cardiac output. Haemopericardium may occur after a penetrating injury to the torso, including chest and upper abdomen. The high-risk zone for this injury is the triangle formed by the two nipples and the sternal notch.

The classical presentation of paediatric cardiac tamponade is a child with a penetrating anterior chest-wall injury who has distant or muffled heart sounds,

neck vein distension and hypotension (Beck's triad). This classical triad is not easy to appreciate as blood loss from other injuries may concurrently reduce central venous pressures, and audible intensity of heart sounds may be difficult to distinguish in children, especially in the trauma resuscitation scenario. Traumatic cardiac tamponade should be suspected in any child with a penetrating chest wall injury in the high-risk triangle with signs of cardiovascular compromise (tachycardia, poor perfusion, delayed capillary refill, hypothermia, hypotension or diminished pulses).

In contrast to haemopericardium, slow fluid accumulation in the pericardium from other aetiologies, such as infective, immunological or malignant, has a minimal effect on cardiac function. The distensible pericardium readily accommodates slow fluid collections, and the presentation of such patients often does not involve perfusion abnormalities.

Pericardiocentesis is a procedure for either life-saving decompression of acute cardiac tamponade or for diagnostic evaluation and management of a non-emergent pericardial effusion. In the setting of acute cardiac tamponade, decompression of the pericardium will temporarily restore myocardial performance and improve cardiac output, until reaccumulation occurs. Hence, needle pericardiocentesis is a temporising measure that precedes open surgical decompression. In non-emergent settings, an indwelling catheter may be indicated to prevent fluid reaccumulation. The procedure can be done with or without electrocardiographic or echocardiographic guidance, depending on the urgency of the clinical situation. Echocardiographically guided pericardiocentesis has been shown to be safe and effective; however, very few emergency physicians are echocardiography trained. As point of care ultrasound becomes a more prevalent skill in the emergency department, ultrasound-guided pericardiocentesis will likely become the preferred technique for this procedure. It has been suggested that pericardiocentesis is a procedure that very few emergency physicians feel comfortable performing[1], highlighting the importance of familiarity and practice, when possible, with the technique.

Indications

- Cardiac tamponade
- Pericardial effusion of unknown aetiology.

Contraindications

- There are no absolute contraindications in the child presenting emergently with shock or cardiopulmonary arrest and evidence of acute tamponade. If equipment and appropriate personnel are present, open thoracotomy may be preferred over needle pericardiocentesis.
- Uncorrected bleeding disorder in semi-urgent or elective cases.

Equipment

Emergent procedure

1. Cleaning solution (chlorhexidine or similar)
2. 30–50 mL syringe
3. 2.5 or 3.5 inch 18–20 gauge spinal needle
4. Ultrasound machine.

Non-emergent procedure

1. 1% lignocaine
2. 25 gauge needle
3. Two 5 mL syringes
4. Two 6–8 cm (2.5 or 3.5 inch) 18–20 gauge spinal needles
5. Three way stopcock
6. 30–50 mL syringe
7. 18 and 20 gauge over-the-needle catheters
8. Cable with alligator clip at each end
9. Specimen containers
10. Electrocardiograph (ECG) machine
11. Ultrasound (echocardiography desirable) machine.

Indwelling catheter

1. Flexible guidewire or J-wire
2. Plastic over-the-wire catheter.

Standard preparation

1. Place the child in a semi-reclined position (30–45 degrees).
2. Establish secure vascular access and apply cardiac monitoring.
3. Secure the airway if necessary.
4. Administer sedation if time permits in awake patient.
5. Identify the subxiphoid entry site (Fig. 24.6.9), inferior and to the left of the xiphoid process.
6. Prepare the area widely with cleaning solution.
7. Infiltrate the entry site with lignocaine in conscious patient.

Non-emergent procedure

In addition to the preparation:

1. attach the correctly sized spinal needle to a stopcock and 30 mL syringe
2. attach one clip of the cable to the hub of the spinal needle and one clip to the V-lead of the ECG machine (Fig. 24.6.10)
3. turn the ECG to the V-lead position.

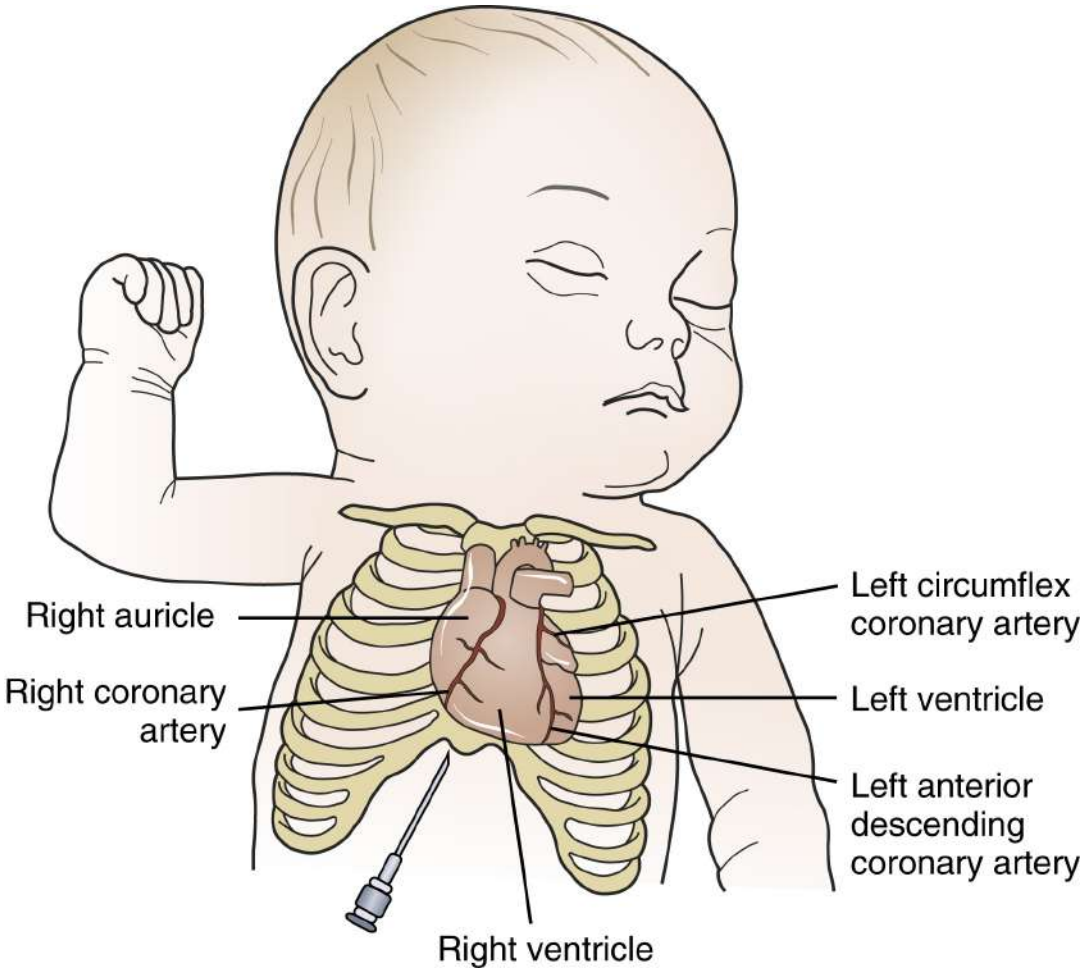

FIG. 25.6.9 Subxiphoid entry site.

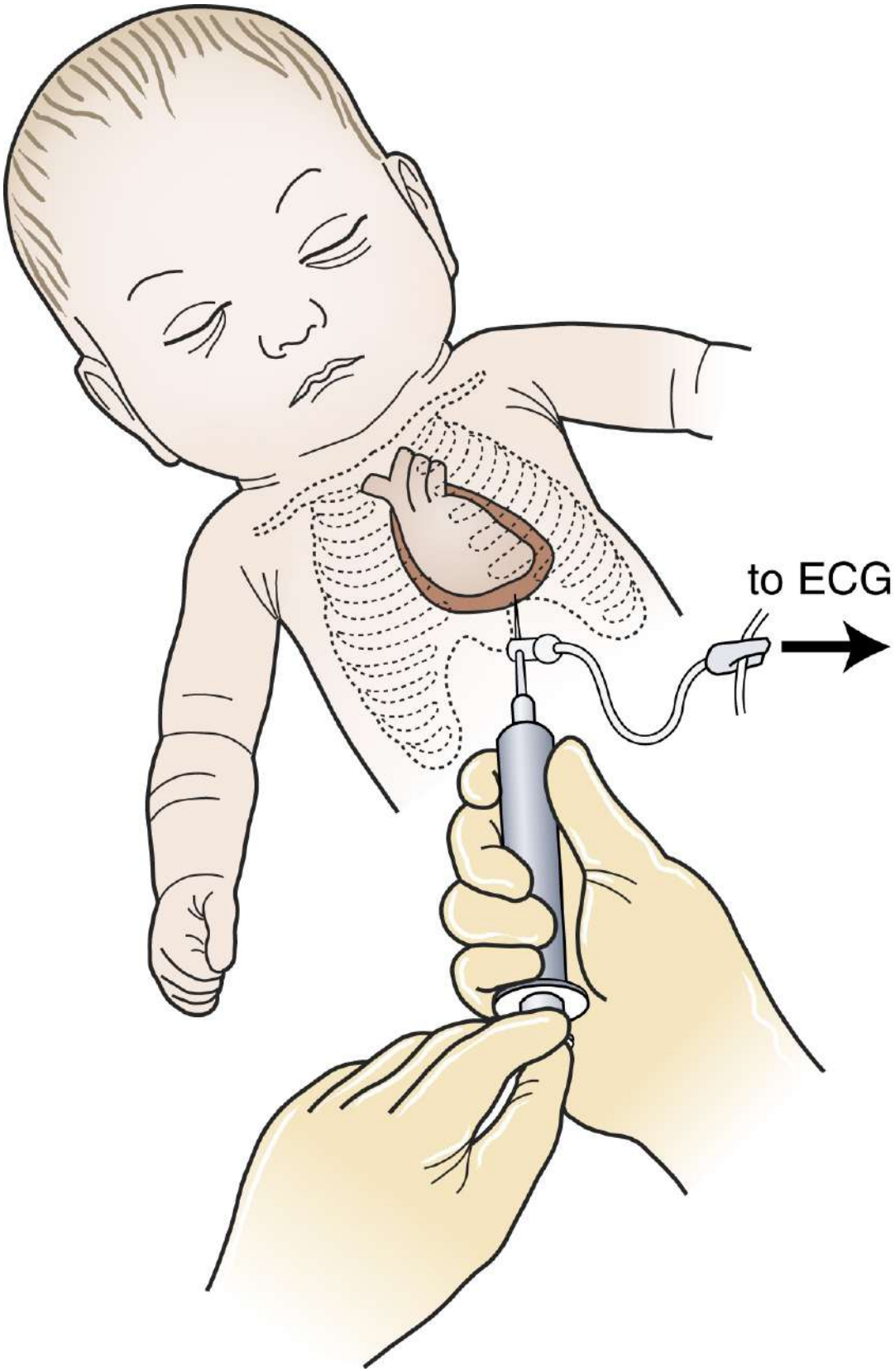

FIG. 24.6.10 Using the ECG for guidance.

Procedure

1. Establish the size, maximal thickness, nearest approach and depth of effusion with ultrasound.
2. Under ultrasound guidance, aim the needle to the left shoulder, and enter at a 30–45 degree angle (see Fig. 24.6.10) towards the anatomic location of the effusion.

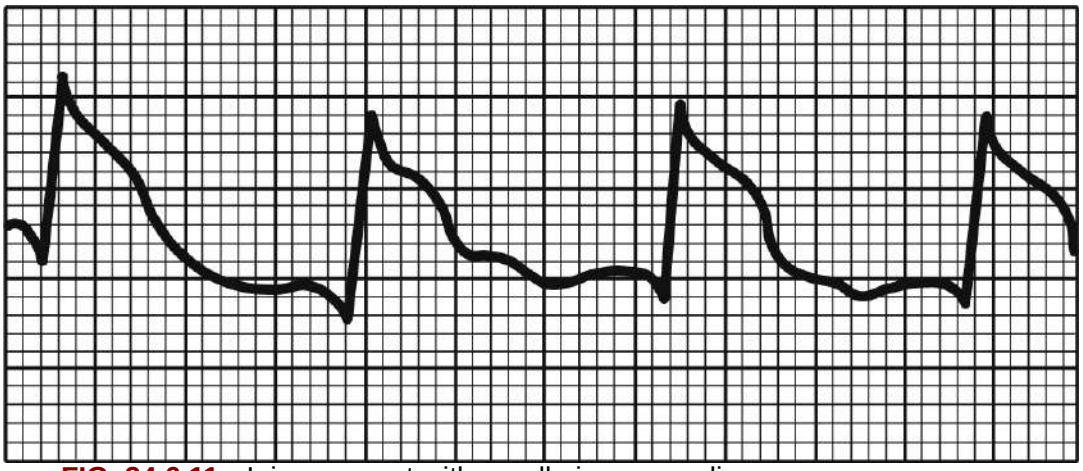

FIG. 24.6.11 Injury current with needle in myocardium.

3. Apply continuous negative pressure whilst advancing the needle until it enters the pericardium.
4. Aspirate until blood or effusion is evacuated from the pericardium.
5. If prolonged drainage is necessary, advance the guidewire or J-wire through the needle, remove the spinal needle, and place the larger catheter with side holes over the wire.

Non-emergent procedure

After the preparation:

1. do constant ECG monitoring
2. if the needle punctures the ventricular epicardium, ST segment elevation or a dysrhythmia may occur (Fig. 24.6.11)

3. if an injury pattern develops, withdraw the needle slightly or reposition more medially
4. always use ultrasound to guide the needle to the area of largest fluid accumulation.

Complications

- Ventricular puncture
- Atrial puncture
- Coronary artery laceration
- Cardiac dysrhythmia
- Haemopericardium
- Pneumothorax
- Haemothorax
- Diaphragm perforation
- Bowel or stomach perforation
- Infection
- Cardiac arrest.

Tips

- If the effusion is not causing tamponade, identify the size and location of the effusion with echocardiography, CT or MRI before doing pericardiocentesis.
- Always use ultrasound guidance.
- If there is a large volume of aspirated blood, the needle may be in the ventricle. The presence or absence of clotting does not reliably indicate needle location.

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24.7

Removing and replacing a tracheostomy tube

Holly Smith, and Scott Schofield

Background

Children with tracheostomy tubes are increasingly common in out-of-hospital and emergency department settings. Congenital airway abnormalities, laryngeal trauma, cervical cord injury, subglottic stenosis and neuromuscular conditions requiring prolonged ventilation often need tracheostomy tube placement. Most of these children live at home and have trained caregivers, often parents. Because the tracheostomy tube may be the primary airway for the child, rapid intervention is required to preserve gas exchange if the tube comes out (decannulation) or becomes obstructed.

Treatment of most tracheostomy problems requires only simple techniques to establish a patent airway, such as suctioning with or without saline. Occasionally, suctioning is not sufficient, and it remains impossible to ventilate a child through an existing tracheostomy tube. In these instances, removal of the old tracheostomy tube and replacement with a new tube of the same size and type are necessary. If this is unsuccessful or a similar tracheostomy tube is not available, then subsequent measures include replacement with a smaller tracheostomy tube if available, or insertion of an endotracheal tube (ETT) through the stoma.

Additionally, many children still have intact upper airways, so provision of oxygen and/or ventilation through the nose and mouth with a bag-valve-mask (BVM) device while occluding the stoma might save the child's life in an emergency.

Some steps may occur concurrently. For example, in the patient with severe respiratory distress or cyanosis, provision of oxygen through the upper airway and preparation for intubation of the upper airway should be occurring while the

existing tracheostomy tube is being changed.

In any tracheostomy emergency, get help early; notify local ear, nose and throat (ENT) and anaesthetic teams if available. Know your local referral patterns.

Indications for emergent replacement

Respiratory distress or failure in the presence of:

- decannulation
- tube obstruction.

Contraindications

No true contraindications exist. However, absence of an appropriately sized tracheostomy tube presents a problem; a larger sized tube should never be used in emergent replacement. In such a case, insert a replacement tube that is smaller in diameter than the original tracheostomy tube.

Equipment

The following equipment should be assembled quickly before tube change, so that everything is at hand in the event of difficulty. Consider having such a list available in your department so that it can be referred to in an emergency:

1. Suction device and sterile suction catheters (diameter less than half of the inner diameter of the tracheostomy tube)
2. Oxygen source
3. Bag-valve-mask device
4. Equipment for upper airway rescue if required: appropriately sized masks for bagging, laryngoscope handle with blades, endotracheal tubes and bougie appropriate for size of patient
5. Round neonatal mask or stoma mask for bagging through stoma if required
6. Tracheostomy cannulas, appropriately sized for patient (Fig. 24.7.1). Use the same type and brand whenever possible, and prepare the same size and one size smaller than the patient already has. Usually the parent has one.

7. ETT (for the stoma as a second line) of the same outer diameter as the original tracheostomy tube and a size smaller
8. Tape or tracheostomy ties
9. Gauze pads
10. Syringes, 5 mL or 10 mL
11. Water-soluble lubricant
12. Scissors
13. Sterile saline
14. Stethoscope
15. $ETCO_2$ monitoring device.

Preparation

- Ask the caregiver if there are any special problems with the child's trachea or tracheostomy and if a replacement tracheostomy tube is available.
- Anticipate difficulties; these can occur at any time and are usually the result of one of the following:
    - False tract
    - Patient agitation or distress
    - Closure of the stoma
    - Spasm of the trachea
    - Stoma is blocked by scar tissue (granuloma)
    - Skin flaps
    - Structural airway abnormalities.
- Always think about your rescue plan; consider the reason the child required the tracheostomy originally. For example, if the child had an obstructed upper airway as the initial indication for tracheostomy placement, rescue breathing with a BVM through the upper airway may be difficult or impossible. Conversely, if the tracheostomy was originally placed for respiratory support in a child with a neuromuscular condition, the upper airway is likely patent and the anatomy favourable.
- Speak directly to the child about what to expect, and attempt to enlist cooperation. Very small children may need to be swaddled to assist the procedure.

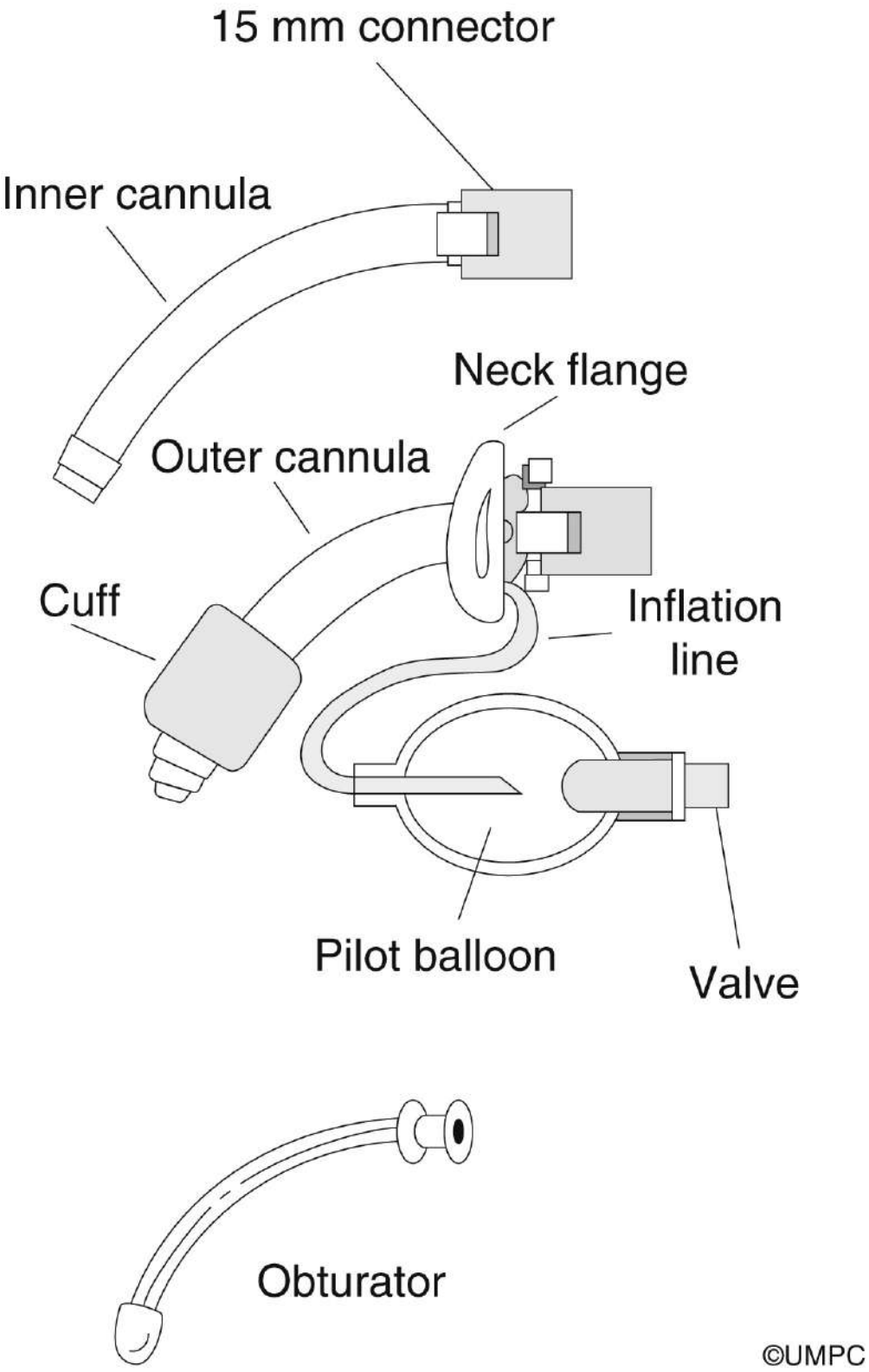

FIG. 24.7.1 Tracheostomy cannulas. From https://www.healthproductsforyou.com/ar-Pediatric-Tracheostomy-Caring-for-Little-Ones.html.

- When possible, have all equipment open and prepared before proceeding; this should include equipment for secondary attempts.
- Provide oxygen through the upper airway passively using mask oxygen or nasal prongs if the child is breathing spontaneously. The unstable or apnoeic child will require a concurrent attempt at positive pressure rescue breaths through the upper airway, with occlusion of the stoma or tracheostomy tube during the preparation phase.
- Enlist a capable assistant.
- Alert appropriate teams as required that a tracheostomy tube is being changed urgently.

Procedure

Removing an old tracheostomy tube

1. Wash hands and don gloves.
2. Position the child with the head and neck hyperextended to expose the tracheostomy site. The child's torso may need to be undressed.
3. If the existing tube has a cuff, deflate it:
    - Connect a 5–10 mL syringe to the valve on the pilot balloon.
    - Draw air out until the balloon collapses.
    - Cutting the balloon will not deflate the cuff.
4. One person should hold the tracheostomy tube in position, while the other cuts or unties the cloth ties that hold the tracheostomy tube in place.
5. Withdraw the tracheostomy tube using a slow, steady, outward and downward motion.
6. Provide oxygen and ventilation through the stoma as needed.

Replacing the tracheostomy tube

Option 1. Using a new tracheostomy tube

1. Insert a tracheostomy tube of the same size and model whenever possible.
2. Insert the obturator inside the tube. If the tube has an inner and outer cannula (i.e. adult type) use the *outer* cannula with the obturator for

insertion.

3. Lubricate the tip of the tracheostomy tube and obturator with a water-soluble lubricant.
4. Hold the device by the flange (wings) or hold the actual tube like a pencil, making sure that the cannula and obturator are held together as a unit.
5. Gently insert the tube with an arching motion posteriorly and downward (follow the curvature of the tube). Slight traction on the skin above or below the stoma may help (Fig. 24.7.2).
6. Once the tube is in place, remove the obturator, attach the bag, and attempt to ventilate. For tubes with an inner and outer cannula, now insert the inner to allow mechanical ventilation with a BVM device.
7. Check placement by observing the patient for bilateral chest rise, listening for equal breath sounds, watching for improving saturations and acceptable capnography. Signs of improper placement include lack of chest rise, resistance to ventilation, air in the surrounding tissues, deterioration in saturations or capnography and patient agitation.
8. If the tube meets with resistance and cannot be inserted, withdraw the tube, administer oxygen through the stoma, and ventilate from above as needed. Do not force a tracheostomy tube that meets resistance.
9. Use a smaller size tracheostomy tube or an ETT for the second attempt.

Option 2. Using an endotracheal tube

1. If a replacement tracheostomy tube is not available, use an ETT of the same outer diameter as the tracheostomy tube. If using an ETT for a second attempt at placement, use a tube of smaller outer diameter.
2. Check the length of the original tracheostomy tube, note the markings on the ETT and plan to advance it to the same depth as the original tube. Avoid cutting the ETT to size, as disrupting the connector to cut the tube may increase the risk of disconnection and loss of the ETT down the stoma.
3. The inserted portion of the ETT will be approximately half the distance needed for oral insertion.
4. Do not advance the tube too far, or it may go into the right main stem bronchus.

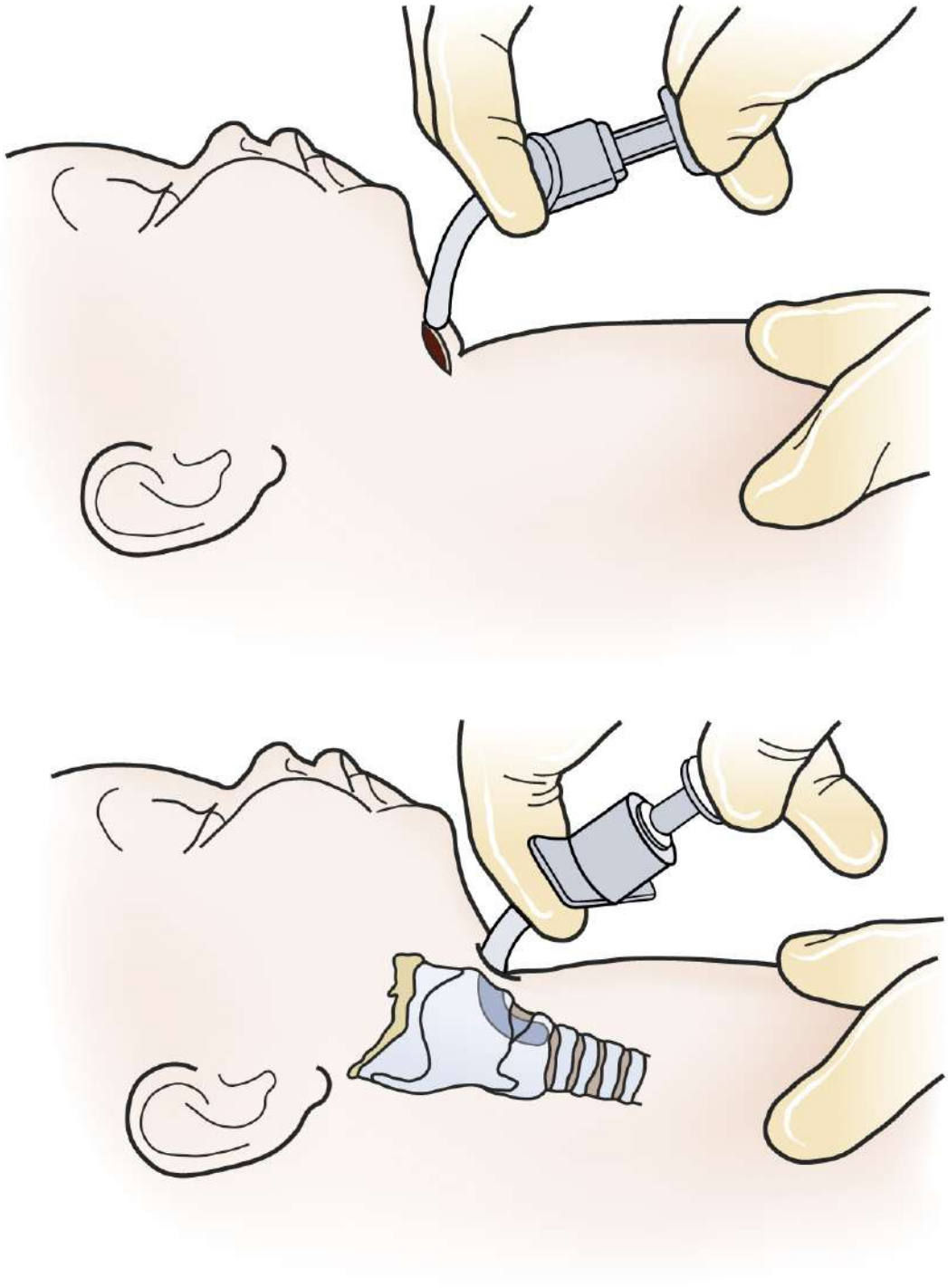

FIG. 24.7.2 Inserting a new tracheostomy tube.

5. A bougie inserted into the ETT may facilitate the procedure.
6. As above, hold the ETT/bougie like a pencil and gently insert the tube with an arching motion posterior and downward (following the

curvature of the tube). Slight traction on the skin above or below the stoma may help.
7. If a bougie is used, remove the bougie carefully while holding the ETT in position.
8. Attempt to ventilate and check adequacy as above.

Option 3. Using a suction catheter as a guide

If unsuccessful with the initial replacement attempts, continue to provide oxygen and ventilation via either the upper airway or stoma, and then use a suction catheter as a guide:

1. Insert a 12 French sterile suction catheter through the tracheostomy tube (i.e. preload the tracheostomy tube onto the suction catheter with the obturator removed).
2. Without applying suction, gently pass the suction catheter into the stoma.
3. Slide the tracheostomy tube along the suction catheter and into the stoma, until it is in the proper position.
4. Remove the suction catheter.
5. Assess ventilation through the tracheostomy tube.

Option 4. Endotracheal intubation and other rescue measures

1. If still unsuccessful, consider either orotracheal intubation or ventilation through the stoma, using a stoma mask or a round newborn mask as available.
2. Alternatively, do BVM over the nose and mouth while covering the stoma with a hand and sterile gauze.
3. Pass a 12 French suction catheter into the stoma, and attach to oxygen source.
4. Ensure help is coming.

Securing the tracheostomy tube

After proper placement, cut the ends of the tracheostomy ties or tape diagonally (allows for easy insertion), pass through eyelets (openings) on the flanges, and tie around the patient’s neck, so that only a little finger can pass between the ties

and the neck.

Complications

- Creation of a false lumen
- Subcutaneous air
- Pneumomediastinum
- Pneumothorax
- Bleeding at insertion site
- Bleeding through tube
- Right main stem intubation with ETT.

Tips

- Provide passive oxygen through the upper airway whenever possible.
- Talk to the caregiver about the size and type of tracheostomy tube and about known problems with the stoma, trachea or tube before proceeding.
- Most children who have lost a tracheostomy tube can be ventilated by BVM over the nose and mouth, with the stoma occluded. Alternately, ventilating through the stoma with a round neonatal or stoma mask can be successful.
- If unable to reinsert a tracheostomy tube, use a similarly sized ETT, or choose a smaller tracheostomy tube.
- Do not force a larger tracheostomy tube through a stoma site.
- New/fresh tracheotomies are especially difficult to manage. Choose a tube of a smaller size, and gently pull the ‘stay sutures’ away from the body so that the trachea is pulled towards the anterior neck.
- Never force a tube that meets with resistance.
- Do not advance an ETT too far into the stoma or it may intubate the right main stem bronchus.
- Always get help early; know your local referral patterns.

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24.8

Central and peripheral intravenous lines

Holly Smith, and Scott Schofield

Background

Most children who present to an emergency department do not need vascular access for drug or fluid therapy. Medications can usually be administered enterally, transmucosally, or intramuscularly or by inhalation. Even fluid administration in dehydrated children does not routinely require vascular access; oral rehydration, performed slowly and methodically, is often successful in children with vomiting and/or diarrhoea. Nasogastric fluids are often preferred over intravenous (IV) if an oral trial has failed. However, when oral and nasogastric rehydration is unsuccessful, or when a child presents critically ill or injured, IV or intraosseous (IO) access (Chapter 24.8) becomes essential.

Finding veins to cannulate in infants and small children can be quite a challenge. The higher ratio of subcutaneous fat and the smaller vessel size in young patients can make venous cannulation difficult. Peripheral venous access (rather than central venous or IO access) offers the highest benefit:risk ratio of any vascular access option. Fig. 24.8.1 illustrates common sites for peripheral IV line insertion. Point-of-care ultrasound can greatly assist in the placement of peripheral IV cannulae, as can other proprietary devices such as the AccuVein™.

When a peripheral site is unavailable, or in an emergent situation, IO access provides a very rapid alternative. The IO route is preferred over central access in initial resuscitation and will tolerate all infusions including hypertonic solutions. If peripheral venous and IO access cannot be obtained, if prolonged hypertonic or vasoactive medications are required, or if central venous pressure monitoring is needed, central venous cannulation is a definitive access method. Central vessels such as the femoral vein, subclavian vein, or internal jugular vein can all be cannulated. Generally, saphenous vein cutdowns are no longer recommended

in children unless all other options have failed, because they are technically difficult to perform and the technique is time consuming in infants and young children, even for experienced operators.

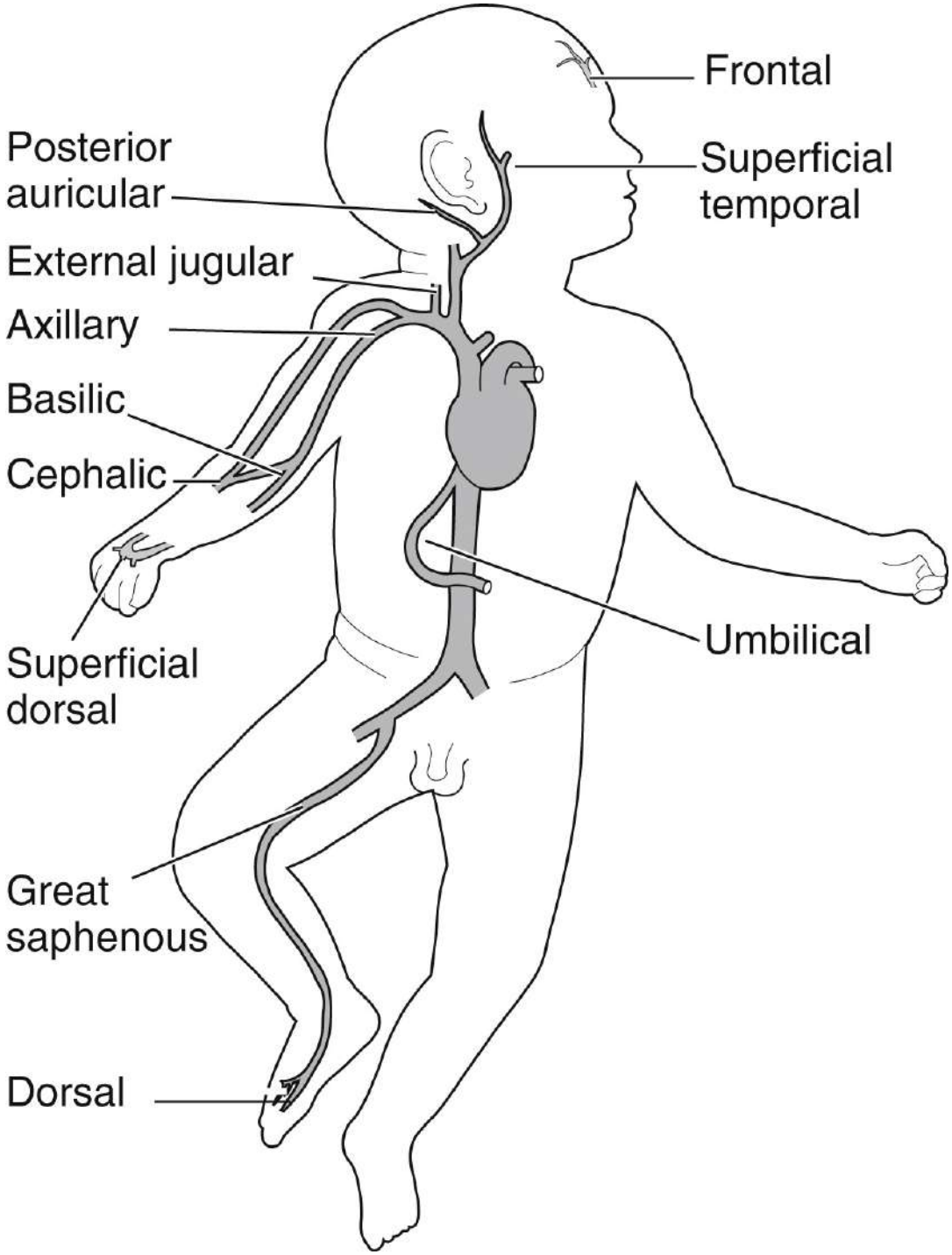

FIG. 24.8.1 Common peripheral intravenous sites.

Make every effort to provide the least invasive form of access indicated by the degree of the child's illness.

Indications

- Peripheral IV access is indicated if enteral, transmucosal, intramuscular, or inhalation routes are not adequate to meet the patient's needs for fluids and/or medications.
- Use a central IV for central venous pressure monitoring and administration of hypertonic solutions, essential access when peripheral and IO attempts have failed.

Contraindications

- Peripheral IV catheters are contraindicated if other less invasive routes can meet the child's needs.
- Central venous catheters are contraindicated if peripheral venous access sites are available and there are no special indications for central venous catheter placement.
- If possible, do not pass peripheral IV catheters:
    - through cellulitic skin
    - in an extremity that has a wound—e.g. burn or laceration
    - distal to unstable fractures or injured veins.
- Avoid placing central lines in non-compressible sites in patients with bleeding diatheses.

Peripheral venous catheter placement

Equipment

Box 24.8.1 lists peripheral IV equipment.

- If time permits, consider numbing the skin with an anaesthetic drug or other means. Age-appropriate distraction or play therapy should also be considered. To anaesthetise skin apply a topical anaesthetic such as the eutectic mixture of local anaesthetics (EMLA™) cream under an occlusive dressing at the selected entry site at least 45 minutes prior to

any IV attempt. Use over-the-needle catheters whenever possible; butterfly needles are far less stable but are acceptable for short-term infusions (i.e. single-dose clotting factor administration). Secure the cannula well with tape, and use arm/legboards to avoid dislodgement of the catheter. Prime any tubing in advance with normal saline, and consider whether three-way tap or a filter is required (prime these as well). Select the smallest sized catheter that will meet the patient's needs for drug and fluid administration.

Preparation

1. Prepare all equipment and place near the child. Avoid using the bed as an equipment tray.
2. Remove any topical anaesthetic at the cannulation site.
3. Have an assistant restrain infants and toddlers; consider wrapping the smaller child with a sheet (Fig. 24.8.2). Your assistant will need to hold the limb to be cannulated. Avoid asking the parent or caregiver to restrain the child; rather ask them to help reassure the child.
4. Attempt to talk an older child through the procedure with a parent or caregiver present.
5. Locate landmarks and determine insertion sites first with an ungloved hand, as this is more sensitive in detecting surface veins and palpating landmarks than the gloved hand.

Box 24.8.1 Equipment for peripheral intravenous insertion

- Gloves
- Arm or leg board
- Tourniquet
- Chlorhexidine pads
- Gauze pads
- 22–24-gauge intravenous (IV) catheters
- Saline flush

- IV tubing, solution and pump
- Protective covering for IV (small plastic cup)
- Tape (enquire about patient sensitivity to tapes)

Procedure

There are a number of possible sites for placement of peripheral IV lines in infants and children. The easiest sites are the dorsum of the hands and feet, the antecubital fossae and, in infants less than 1 year, the scalp. In non-emergent situations, start distally and move proximally if initial attempts fail. Consider using a warm compress to dilate constricted veins and make them more visible. In emergent situations, cannulate the largest vein available and consider less commonly used peripheral sites, such as the external jugular vein or deep brachial vein:

1. Wash hands and don gloves.
2. Clean the insertion site with an appropriate surgical prep solution.
3. Apply a tourniquet just proximal to the insertion site. Use a rubber band around the scalp or an assistant's finger as pressure caudad to the insertion site when cannulating scalp veins.
4. Locate a straight segment of the vein and provide in-line traction away from the direction of catheter insertion.
5. Insert the catheter at a 10–20 degree angle to the skin. Once there is a flash of blood into the hub of the catheter, lower the cannula toward the plane of the skin and advance the catheter with the needle in place another 1–2 mm (this step is crucial). Finally, advance the catheter over the needle into the vein.
6. Draw any blood samples needed, remove the tourniquet and connect the IV line or primed extension tubing and flush. There should be no resistance to flush, no swelling or immediate bruising, no blanching of the skin, and no significant pain at the time of initial flush.
7. Secure the catheter and IV line to the patient with tape and consider placing a clear plastic container over the cannula (Fig. 24.8.3). Stabilise the limb with an arm or leg board, and avoid opaque dressings through which it is impossible to visualise the insertion site.

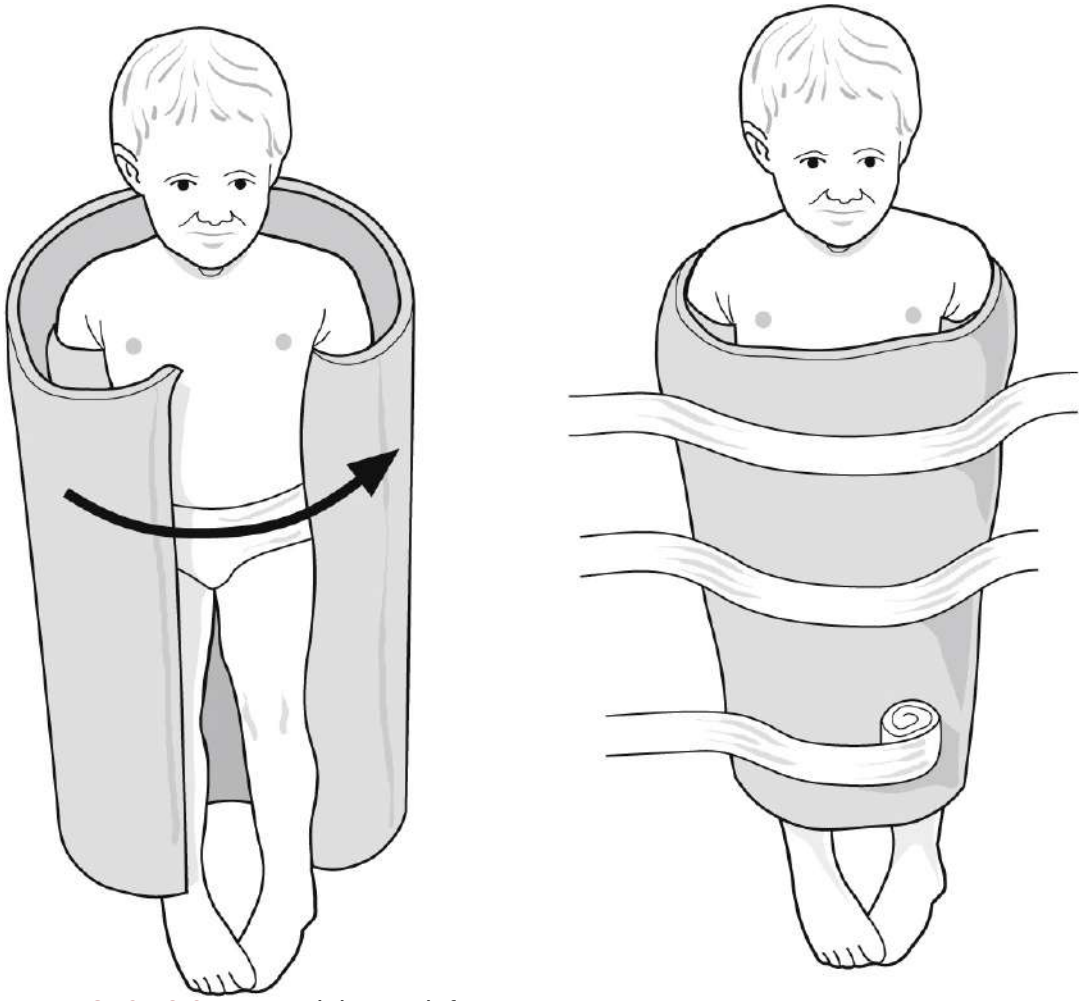

FIG. 24.8.2 Restraining an infant.

If insertion of a peripheral cannula is unsuccessful or the need is emergent, insert an IO catheter. One can also consider cannulating the external jugular vein prior to moving to a central line (if the situation permits). To cannulate the external jugular vein, place the patient in the Trendelenburg position to dilate the vein. Restrain the patient well and turn his/her head slightly away from the cannulation site (Fig. 24.8.4). Clean the area with chlorhexidine or povidone-iodine solution, and use an 18 gauge needle to make a nick in the skin at the catheter site. This will facilitate passage of the catheter through the skin. Use the peripheral insertion technique, but apply firm counter-traction at the entry site to help stabilise the vessel. Secure the cannula carefully to avoid dislodgement and avoid opaque dressings.

Complications

Local complications

- Phlebitis
- Site infection
- Infiltration/extravasation
- Bleeding and significant bruising
- Pressure necrosis due to IV stabilisation methods.

To avoid these complications, clean insertion sites well prior to passing the needle through the skin. Make sure blood can be easily withdrawn from and fluids easily infused through the cannula. Avoid infusing irritant or hyperosmolar solutions through peripheral lines. Tape IV lines securely, but do not create a tourniquet with circumferential tape. Replace peripheral lines every 48–72 hours.

Systemic complications

- Thrombosis
- Air embolism
- Catheter tip embolisation.

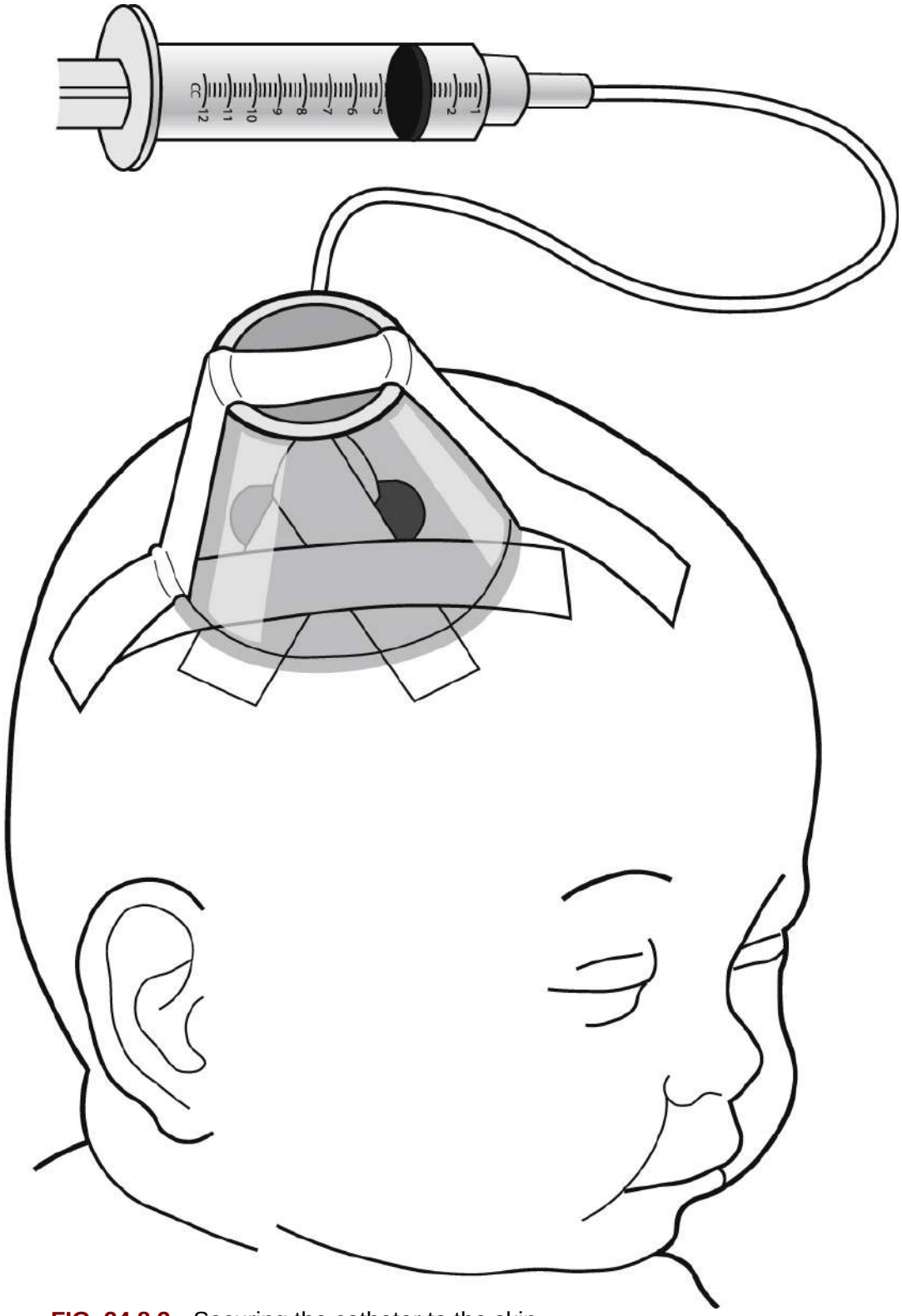

FIG. 24.8.3 Securing the catheter to the skin.

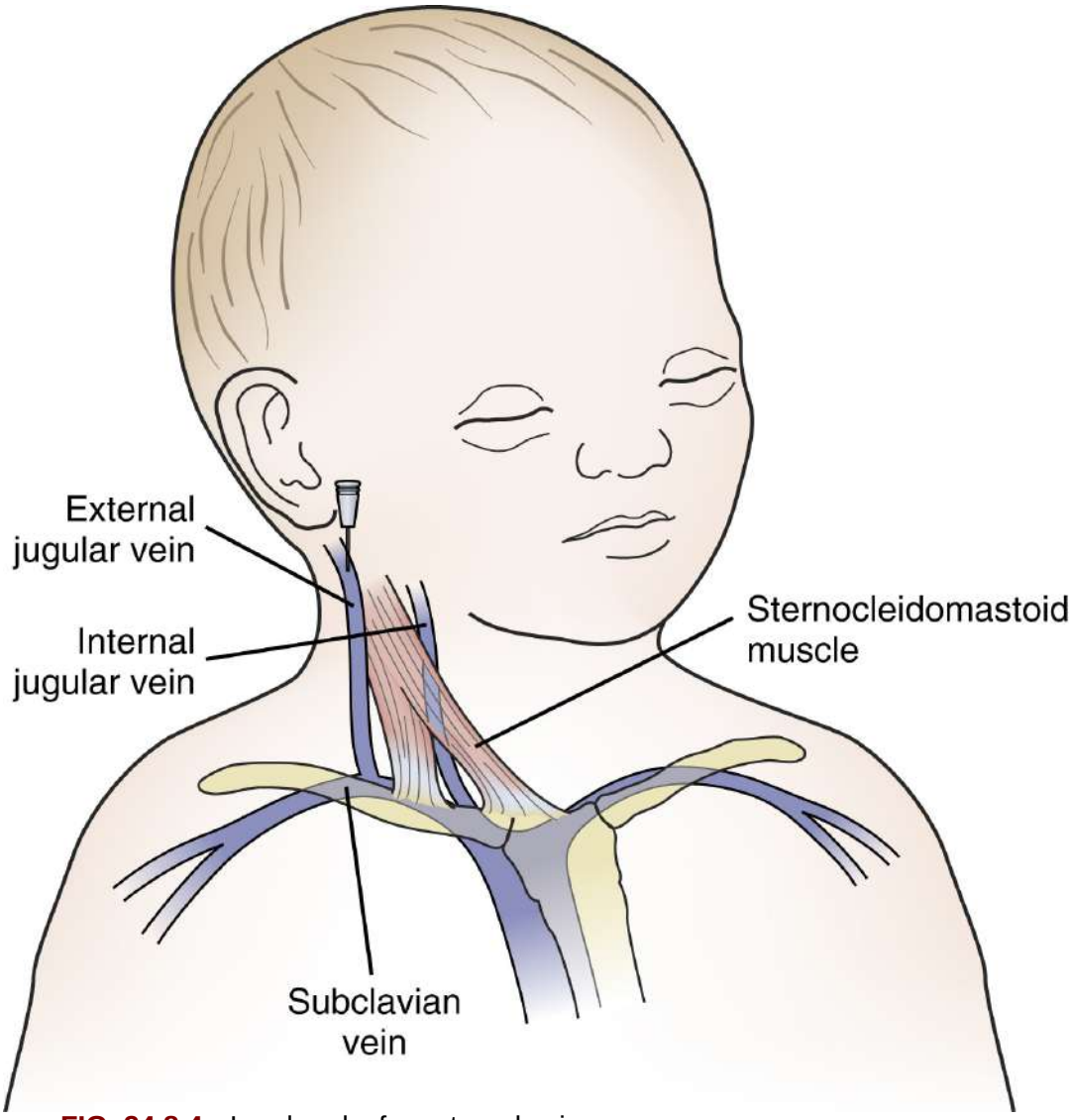

FIG. 24.8.4 Landmarks for external vein puncture: https://www.unboundmedicine.com/harrietlane/view/Harriet_Lane_Handbook/309868/all/Proced taken originally from Dieckmann R, Fiser D, Selbst S. *Pediatric Emergency and Critical Care Procedure*. St. Louis: Mosby; 1997.

Flush catheters regularly but not forcefully. Do not re-insert the needle stylet into the catheter once it has been removed; doing this can shear the catheter and cause catheter fragment embolisation.

Tips

- Whenever possible, use a topical anaesthetic at several possible IV sites at least 45 minutes prior to line placement.
- Avoid joint surfaces and the patient's dominant hand. If a large-bore

catheter is required, use more proximal sites, such as those at the antecubital fossa or external jugular vein.

- Do not let go of recently placed IV catheters until they are well secured.
- Monitor IV catheters and cannulation sites closely, particularly in infants and small children. Persistent crying, extremity swelling, blanching or discoloration of the skin may be signs of extravasation from the catheter, which can result in significant tissue injury.

Central venous line placement

Equipment

The equipment required for placement of a central venous line is usually in a pre-packaged kit. The simplest and most straightforward technique for central-line placement is the guidewire or Seldinger technique. Pre-packaged kits will contain a needle, syringe, J-tipped guidewire, scalpel, dilator and central venous catheter. These should be kept sterile. Determine appropriate catheter size by measuring the child next to a length-based resuscitation tape or by consulting Table 24.8.1. Estimate desired catheter length and insertion distance by measuring from the insertion site externally to the desired tip position.

Table 24.8.1

Sizing central intravenous catheters

Weight (kg)	Age	Catheter size (French)
<5	Newborn to 6 months	3, 4
5–15	6 months to 5 years	5, 7
15	>5 years	5–11

Preparation

1. Central venous catheterisation should be a sterile procedure; every attempt should be made to achieve this, including in resuscitation situations.
2. If time and clinical situation allow, sedate the child prior to insertion of a central venous catheter and/or use a topical anaesthetic such as EMLA™

cream. If the situation is emergent, use a sheet wrapped around the child and an assistant to restrain the child. Excessive patient movement will increase the rate of complication and decrease the likelihood of procedure success.

3. Prepare all equipment on the tray so that items can be easily located. Do not use the child's bed as an instrument table.
4. Flush the catheter with saline solution in advance unless diagnostic blood specimens are required.

Procedure

Sites for central venous line placement in children include the femoral vein, the internal jugular vein, and the subclavian vein. The external jugular vein is also a possible central venous insertion site; however, passage of the catheter centrally via the external jugular vein is difficult because of the acute angle of entry of the external jugular into the subclavian vein. It is, therefore, the least desirable site. Central venous access is optimised by the use of point-of-care ultrasound.

Fig. 24.8.5 illustrates the essential anatomy for placement of a **femoral venous catheter**.

1. Place the patient supine in a mild reverse Trendelenburg position. Maintain the hip in slight external rotation and abduction.
2. Palpate the femoral artery at the inguinal ligament. The insertion site is about a patient's finger breadth medial to the artery and 2–3 cm distal to the inguinal ligament. The right side is easier for a right-handed operator.
3. Measure from insertion site to the umbilicus to estimate catheter insertion distance.

To cannulate the **internal jugular vein**, place the patient in Trendelenburg position with the neck extended and the head turned slightly away from the insertion site. A roll placed under the ipsilateral shoulder will facilitate neck extension, particularly in infants. This should be avoided in major trauma patients where the integrity of the cervical spine is in question. Fig. 24.8.6 illustrates the relevant anatomy and landmarks.

1. Identify the triangle formed by the sternal and clavicular heads of the

sternocleidomastoid muscle and the clavicle.

2. Palpate the carotid artery just medial to the triangle.
3. The insertion site is either at the apex of the triangle or halfway (from mastoid to sternum) along the medial border of the sternal head of the sternocleidomastoid. In either case, aim towards the ipsilateral nipple and away from the carotid artery. The right side is preferable because of the straighter course to the superior vena cava, the lower pleural dome and the absence of the thoracic duct.
4. Place the catheter tip at the junction of the superior vena cava and the right atrium (surface anatomy of this junction is where the third rib attaches to the sternum).

Positioning and anatomy relevant for placement of a **subclavian central venous line** is similar to that for placement of an internal jugular venous line (Fig. 24.8.7).

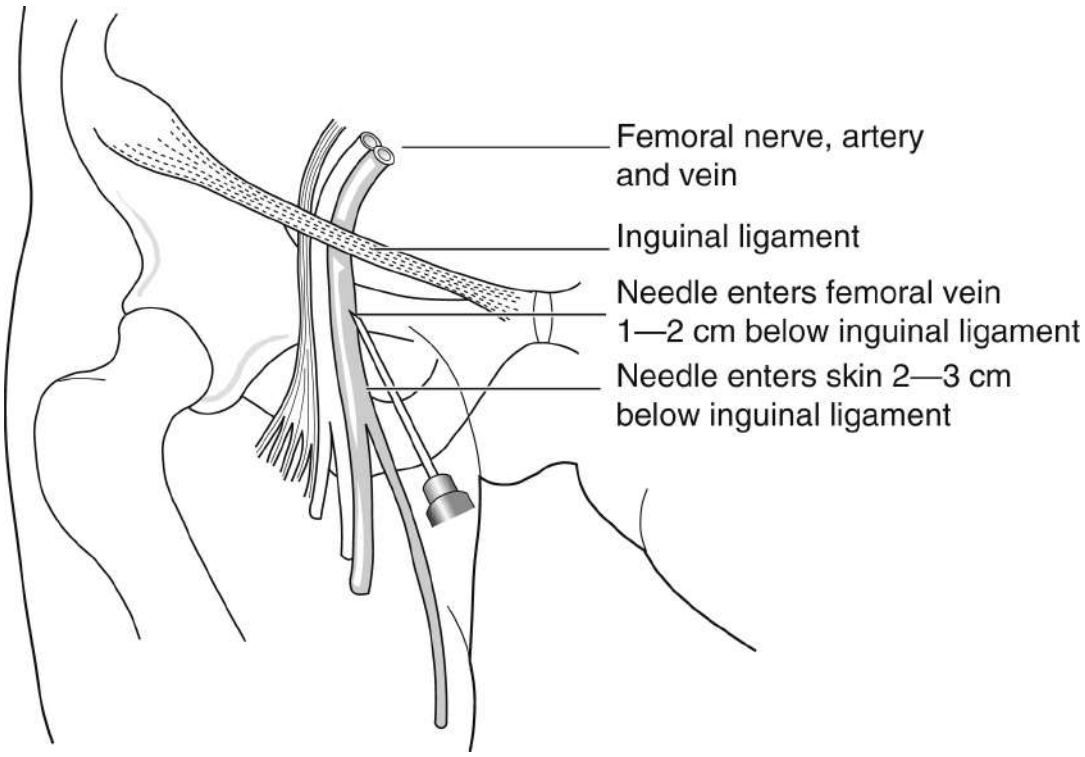

FIG. 24.8.5 Essential anatomy for placement of a femoral venous catheter.

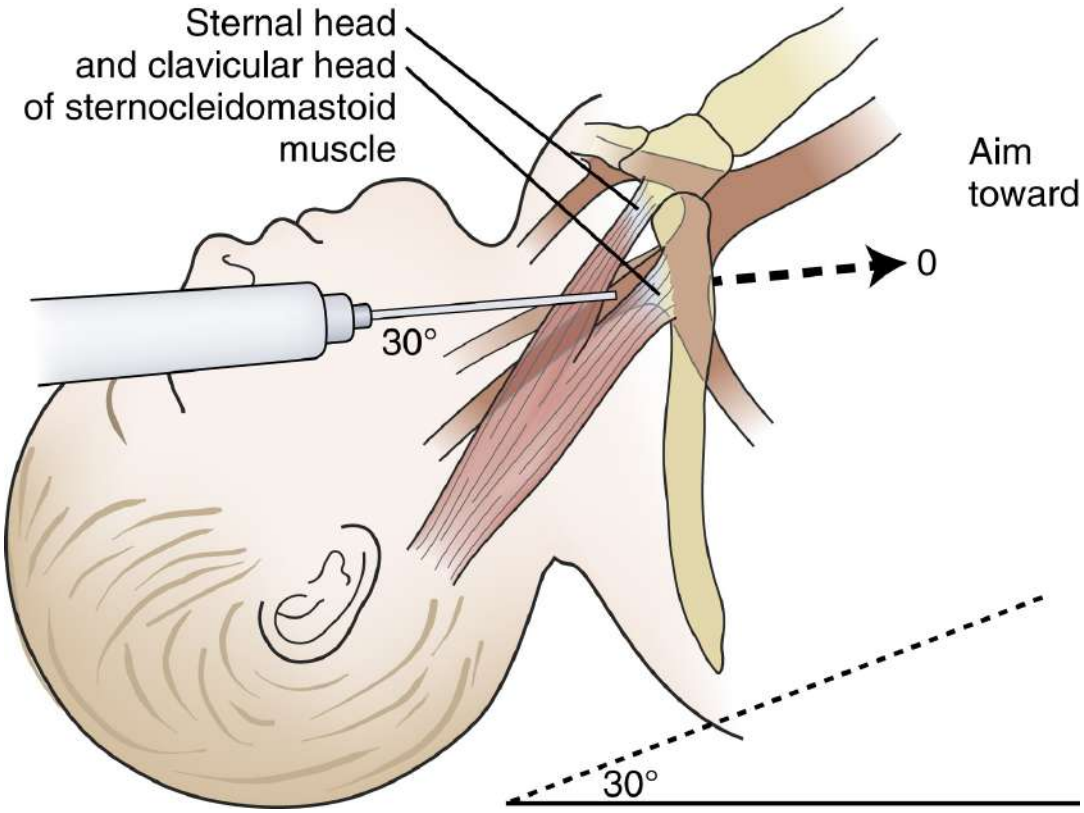

FIG. 24.8.6 Cannulating the internal jugular vein.

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1. Sedate conscious patients before attempting a subclavian line, as patient movement can result in a high rate of complications. Place the patient in Trendelenburg position with a towel roll underneath the ipsilateral shoulder to help keep the neck in extension.
2. Mentally divide the clavicle into thirds.
3. The insertion site is just inferior to the junction of the middle and medial thirds of the clavicle aiming towards the sternal notch. Aim the J curve of the guidewire caudally during insertion. Also, turn the patient's head toward the site of insertion while advancing the wire to avoid passage of the wire up the internal jugular vein.
4. Place the catheter tip at the junction of the superior vena cava and the right atrium as above.

The **general procedure** for placement of a central venous catheter is the same

regardless of anatomic location.

1. Wash hands and don sterile gloves.
2. Prepare all equipment and prime catheter and lumens with saline. Consider the desired insertion distance and observe the guidewire for distance markings.
3. Clean the skin with a povidone-iodine or chlorhexidine solution.
4. Anaesthetise the site with 1% lignocaine if possible.
5. Attach a syringe to the insertion needle so that the bevel lines up with the numbers on the syringe. This allows for identification of bevel direction even when the needle is inside the patient.
6. Insert the needle at the selected insertion site with the bevel pointed anteriorly (femoral), medially (internal jugular), or caudally (subclavian).
7. Angle the needle at approximately 30 degrees to the skin, except when placing a subclavian line. With subclavian line placement, use the shallowest angle possible that allows insertion of the needle underneath the clavicle aiming towards the sternal notch.
8. Maintain constant negative pressure on the syringe while inserting or withdrawing the needle; adequate vessel entry is marked by easy flow of blood into the syringe with gentle negative pressure.
9. Once blood flows freely into the syringe, stabilise the needle by holding it with your non-dominant hand resting on the patient. Remove the syringe carefully, trying not to alter the position of the needle.
10. Insert the guidewire through the needle, J end first (Fig. 24.8.8). Point the J of the guidewire in the direction the catheter is intended to go. The wire should pass easily; do not force it if it does not. Removing the wire and repositioning the needle can often resolve mild resistance. If strong resistance is met, remove the needle and wire together to avoid shearing off the wire into the vein, compress the vein to stop bleeding and then begin again.
11. Once the guidewire is passed to the pre-marked distance, remove the needle while still maintaining hold of the wire.
12. Make a small stab incision with a scalpel over the wire. Insert a dilator over the wire, once again remembering to hold onto the wire at all times (an assistant wearing sterile gloves can help with this).
13. Remove the dilator and insert the catheter over the wire to the pre-

measured distance (Fig. 24.8.9). Remove the wire. Draw off any diagnostic blood specimens. If the catheter has not been flushed in advance, withdraw blood from all lumens of the catheter prior to infusing any fluids or medications to avoid air embolisation.

14. Suture the catheter in place, and apply a clear sterile dressing. Verify location of the catheter tip with an X-ray.

Complications

Box 24.8.2 lists the most common complications of central venous line placement:

- Some bleeding will normally occur after removal of the dilator, this will cease with correct placement of the central line itself. Significant bleeding can occur during traumatic line placement or after arterial puncture. Patients with bleeding diatheses are at great risk for bleeding complications. Compress bleeding sites from femoral or internal jugular sites. These sites are safer than the subclavian route where the internal bleeding site is non-compressible. Avoid subclavian catheterisation in patients who are moving around or who have clotting problems.

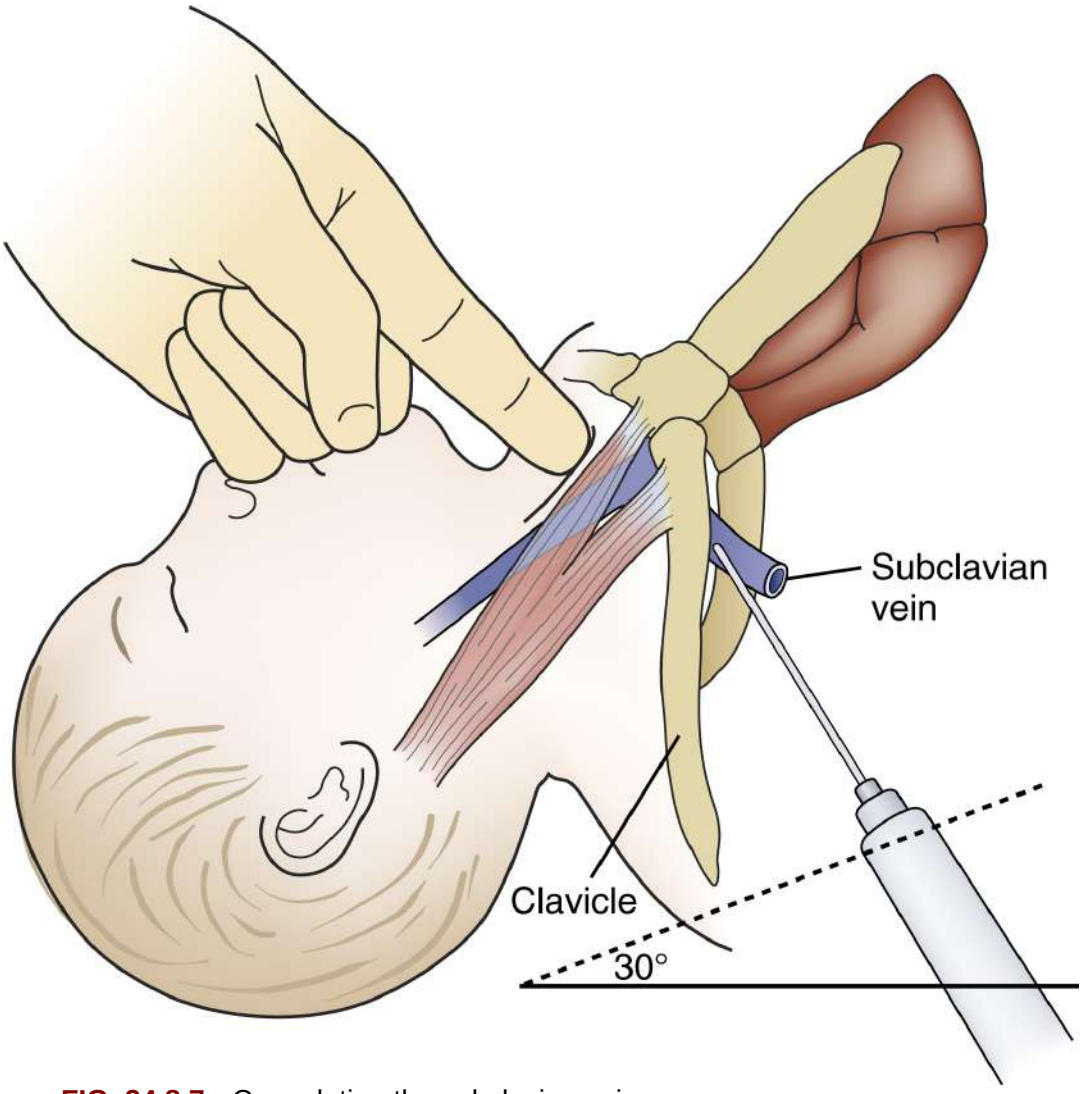

FIG. 24.8.7 Cannulating the subclavian vein.

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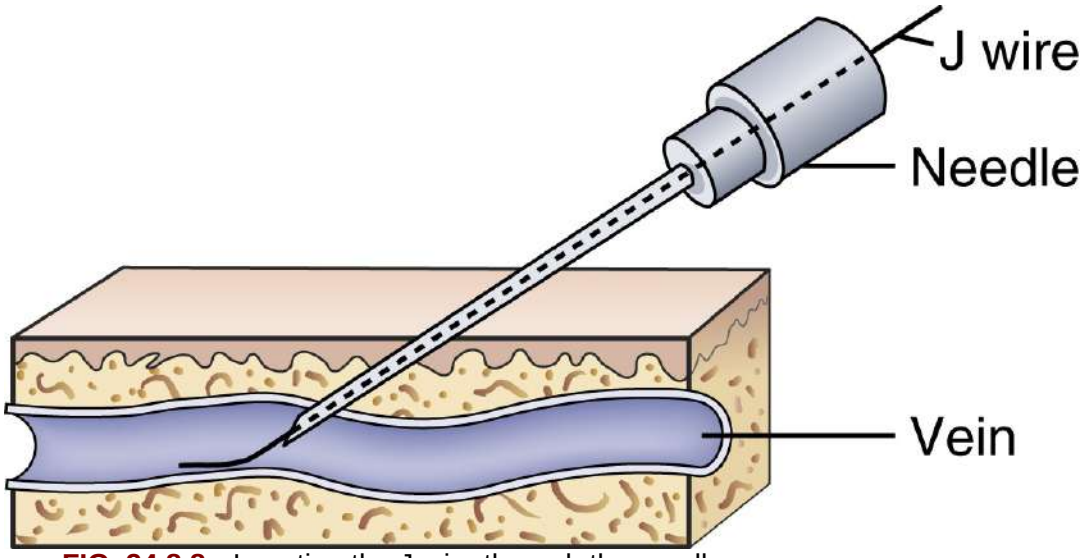

FIG. 24.8.8 Inserting the J-wire through the needle.

Box 24.8.2 Complications of central venous line placement

- Bleeding
- Arterial puncture/cannulation/laceration
- Infection
- Catheter thrombosis
- Pneumothorax
- Haemothorax
- Thoracic duct laceration

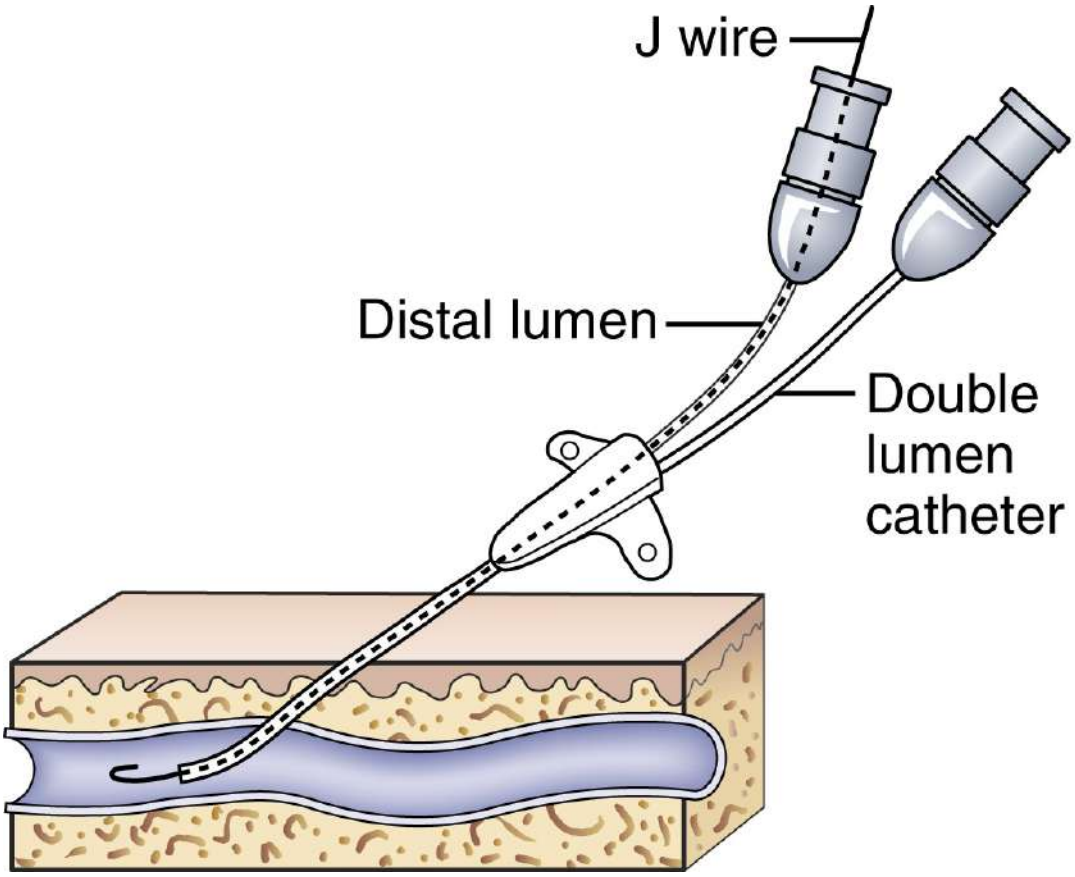

FIG. 24.8.9 Inserting the catheter over the wire to the pre-measured distance.

- Pneumothorax can occur with internal jugular or subclavian line insertion. A shallow angle of insertion with subclavian lines can help prevent this complication. Obtain an X-ray after all line placements, not only to verify catheter position but also to detect a pneumothorax.
- Suspect catheter thrombosis if withdrawal of blood or infusion of fluids becomes difficult. Eventually pain and swelling of the extremity will tip the clinician off to the presence of the clot. Manage thromboses expeditiously with catheter removal.
- Infection is equally likely to occur at any of the central venous sites. Remove infected catheters as soon as possible, and treat aggressively with antibiotics; line sepsis can be fatal. If possible, avoid insertion of a new central catheter for 24–48 hours.

Tips

- In the awake patient, restraint and proper sedation and analgesia will improve procedural success and avoid complications.
- Use of ultrasound is becoming the gold standard to identify vessels prior to line placement.
- If the patient is moving and there is no time for sedation, the femoral site is the safest and most easily compressible vein if bleeding occurs.
- If central venous pressure monitoring is required, use an access site above the diaphragm. While the internal jugular vein is larger, the subclavian site is more comfortable for the patient in the long term.
- If the patient is pulseless, the subclavian vein is the preferred site for central venous catheterisation as it does not require a pulse for localisation and can be used for central venous pressure monitoring.

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24.9

Intraosseous infusions

Holly Smith, and Scott Schofield

Background

Peripheral intravenous (IV) cannulation in critically ill or injured children can be difficult, time consuming and sometimes impossible. Small veins collapse or disappear during shock, and increased body fat in toddlers may camouflage superficial vessels. Central venous access is a difficult procedure that takes too long when treatment is time critical. Surgical cut-down may be risky or impossible in critical situations and is generally no longer advised in small children. Although the endotracheal route is an alternative to vascular access in cardiopulmonary arrest, endotracheal intubation may be delayed, drug absorption may not be reliable, and large-volume fluid administration is contraindicated via this route.

The intraosseous (IO) or intramedullary route for the delivery of resuscitation fluids and medications has been used for over 50 years in children and adults. Many studies have confirmed that the highly vascularised IO space is an excellent route for medications and fluids. The only technical problem is manually piercing the bony cortex in older children. The bones of neonates and infants are usually soft and the IO space is relatively large, so needle insertion is easy in young children. The IO space functions as a non-collapsible vein. The emissary veins of the IO space absorb all parenteral medications, crystalloid fluids, or blood products, which move quickly into the central circulation. Complications are minor and infrequent. Out-of-hospital emergency-care professionals have used IO access with a high rate of success, and it is a recommended access method in the management of cardiac arrest.

There are several possible sites for insertion, but the easiest location in children is the proximal tibia. Good equipment, thoughtful preparation, and effective technique are important for success in IO needle insertion.

Indications

- Cardiopulmonary arrest.
- Any critical emergency in which peripheral cannulation has not been immediately successful, and in which oral, transmucosal, intramuscular or inhalation routes are not adequate to meet the patient's needs for fluids and/or medications.

Contraindications

- Do not use intraosseous access if the child is stable.
- Do not place an IO needle below a fracture site: use the other side.
- Avoid placement of an IO below any open injury on an extremity: use the other side.

Relative contraindications

- Avoid IO insertion in children with osteoporosis and osteogenesis imperfecta due to the high fracture potential.
- Recent prior use of the same bone for IO infusion (potential for extravasation from previous IO sites).

Equipment

1. Commercially available manual IO needles have durable parts intended for penetration of bone. There are several styles (Fig. 24.9.1). A central stylet is universal. There are short, 2.5 cm needles for neonates and infants and longer and 3.0 and 3.5 cm needles for older children. Some needles have stylets with multifaceted cutting edges intended for a rotary insertion, others have bevels and others can be screwed in place. The shaft may have side ports. Once inserted, all function with the same efficacy.
2. The EZ-IO® or IO gun is an excellent alternative to the manual needle and is now standard in most paediatric emergency settings.
3. Other types of styleted needles will work; spinal needles may be

effective in neonates and infants, but these needles bend too easily in the more calcified bones of older children and adolescents.

4. 10 mL syringe
5. Normal saline
6. Stopcock (optional).

Preparation

Identification of the entry site

The best site in children is the anteromedial aspect of the proximal tibia, medial to the tibial tuberosity. Alternative sites are the distal femur 2–3 cm above the patella in the midline, proximal humerus and the medial malleolus at the ankle (Fig. 24.9.2). The sternum is not a recommended insertion site in children.

Positioning the child

For a proximal tibial insertion, place the child supine with the knee slightly flexed and a small towel roll or other bulky material under the popliteal fossa. Make sure the conscious child is well immobilised.

Selecting the correct side

A right-handed operator will more easily insert the needle in the child’s right leg and vice versa for the left-handed operator.

Procedure

1. Wash hands and don gloves. Prepare the access site with chlorhexidine or povidone-iodine solution.
2. Grasp the limb with the non-dominant hand near the insertion site to steady the bone during placement.
3. Introduce the IO needle through the skin until the bony surface is reached, directing slightly away from the growth plate. Avoid starting to twist or drill until the needle has been seated firmly into the periosteum.

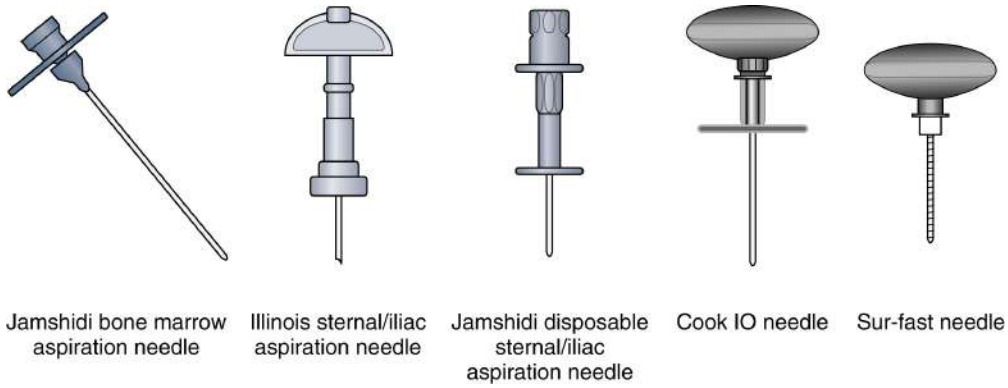

FIG. 24.9.1 Various manual IO needles. Redrawn from "Roberts and Hedges' Clinical Procedures in Emergency Medicine and Acute Care,7th Edition, 2018.

4. For manual insertion of the IO, pierce the bony cortex with a firm, twisting motion from a position directly above the entry site. A 'pop' may be felt as the needle passes through the bony cortex and into the marrow cavity. Do not push too hard on the needle. Too much force may push the needle all the way through the bone and into the soft tissues behind, especially in infants.
5. Remove the stylet and aspirate marrow contents with a 10 mL syringe; these usually bubble up into the syringe. This aspirated bone marrow can be used immediately to reliably check glucose and for other tests as required (see below). Rarely, marrow cannot be aspirated. The IO needle should stand firmly upright in the bone and not feel loose in the tissue. If the needle feels loose in the surrounding tissue, it is not correctly placed and will extravasate into the soft tissues.
6. Confirm correct placement by infusing 10 mL of normal saline without significant resistance. Once the IO needle has been placed, attach a three-way stopcock (if available) to the end of the needle before attaching the IV line.
7. Attach the IV line to the hub and infuse fluids or drugs directly into IO space. Pushing the fluids too forcefully may force the fluids back out of the IO space through the entry point. The rate of fluid infusion may be limited by the fixed size of the marrow space. Fluid must be infused under pressure (i.e. manually or with a pump) and will not flow passively into the marrow space.

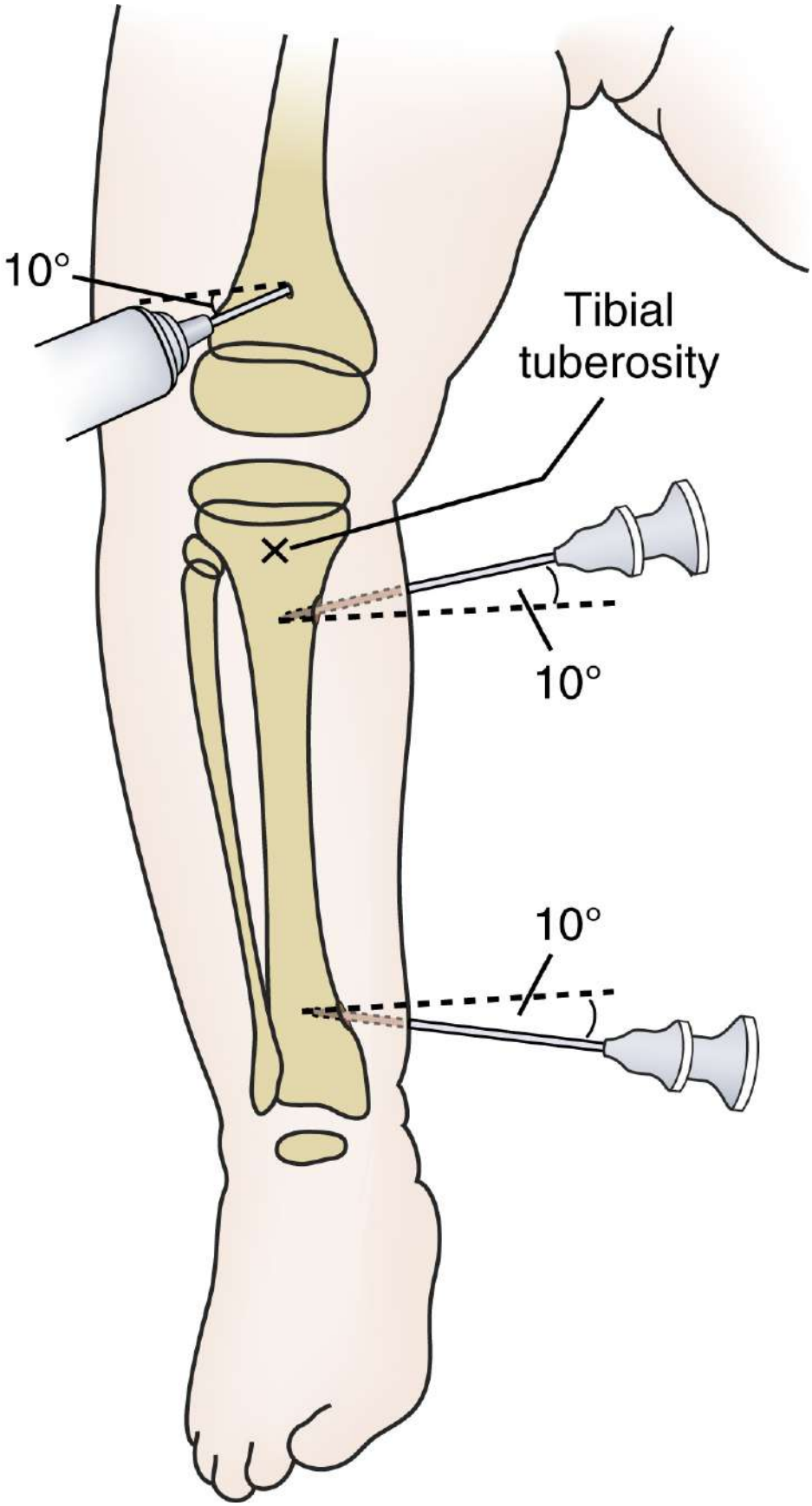
10°
Tibial
tuberosity
10°
10°

FIG. 24.9.2 Potential lower limb intraosseous access sites. From Schexnayder SM. Pediatric septic shock. Pediatrics in Review 1999;20:303–8.

8. Secure the needle to the overlying skin with tape. It is good practice to give one staff member the task of safeguarding the IO if possible.
9. Monitor the calf to ensure that there is no swelling to indicate leakage of fluid. If this occurs, stop using the IO and insert another at another site in another bone.

Complications

- Compartment syndrome
- Failed infusion
- Growth-plate injury
- Bone infection
- Skin infection
- Skin necrosis
- Bony fracture.

The EZ-IO® Intraosseous Vascular Access System

Mechanical devices such as the EZ-IO® System have simplified the insertion of an IO needle as they are not dependent on the manual process and not limited by thickness of the bony cortex. The EZ-IO® System (Teleflex Incorporated, Wayne, PA, USA) includes a reusable battery-powered driver that operates like a small drill. If the driver is unavailable or inoperable, manual insertion may be utilized. The EZ-IO® System includes the patented cutting needle tip and powered driver enabling clinician control of the insertion with proven tactile feedback (Fig. 24.9.3).

The procedures for insertion of the EZ-IO® needle are similar to those described above for the manual device. The difference is in the ease of cortex penetration and in the choice of paediatric needle size, which must be considered before insertion (Fig. 24.9.4).

Size choices (all 15 gauge):

- 15-mm-long needle generally used for children weighing between 3 kg and 39 kg who have minimal soft tissue overlying the insertion site.
- 25-mm-long needle for children weighing greater than 40 kg (or smaller children who have thick subcutaneous tissue overlying the bone).
- 45-mm-long needle is available for larger children with significant tissue overlying bone.

Avoid using an undersized needle in well-covered children as this may result in early dislodgement of the needle and/or lead to pressure and tissue necrosis under the flange of the hub.

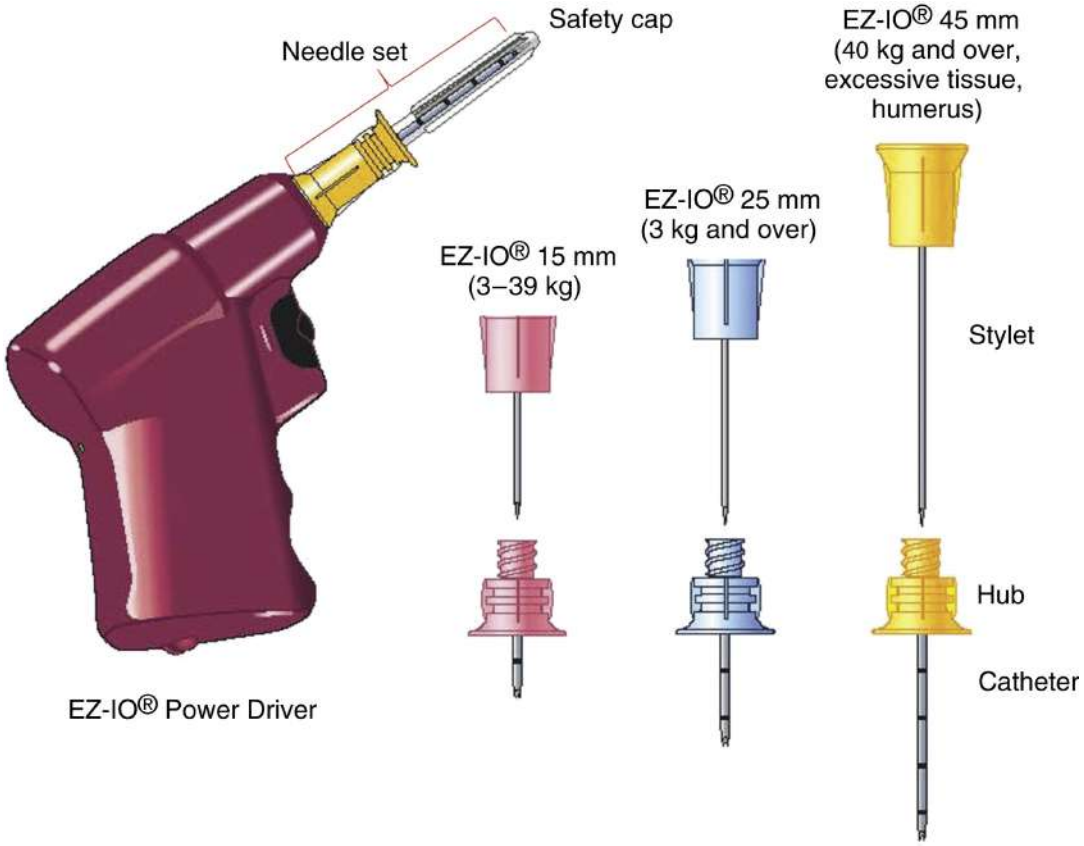

FIG. 24.9.3 EZ-IO® Power Driver and Needle Sets - description and selection. Image courtesy of Teleflex Incorporated. © 2018 Teleflex Incorporated. All rights reserved.

Tips

- Be careful when inserting an IO of any kind in a young infant, because it is quite easy to penetrate through the bone into the soft tissues and cause a compartment syndrome.
- If the child is conscious, provide generous local anaesthesia at the entry site, including the periosteum.
- In addition to serving as a route for drug and fluid administration, blood/marrow samples drawn from the site can be used for culture, haematologic, and biochemical analysis. Emergency blood group and crossmatching is reliable with an adequate sample. A complete blood cell count may be difficult to interpret, as it reflects the marrow cell count rather than that in the peripheral circulation. Remember that blood aspirated from the marrow space clots rapidly, even if placed in a tube containing heparin. Marrow specimens should not be run through point-of-care blood gas machines and should be clearly labelled as marrow specimens when being sent to the lab.
- All commonly used IV fluids and drugs can be administered via the IO route.
- The rate of infection from IO needle placement is low and comparable to that of IV cannulation.
- Remember that the EZ-IO™ has a limited number of uses, so avoid 'spinning' the drill needlessly.

Further reading

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24.10

Umbilical vessel cannulation

Holly Smith, and Scott Schofield

Background

During fetal life, two umbilical arteries transport nutrients and oxygen from the placenta, and one umbilical vein helps dispose of fetal waste. During delivery, these vessels are cut and clamped, and the newborn is separated from the placenta. However, the umbilical vessels can be recannulated and utilised for emergent vascular access in ill neonates for up to 7 days after birth. This is an excellent method of central drug and fluid delivery because peripheral venous access in infants in the first week of life can be quite difficult, particularly in the ill or intravascularly depleted neonate. The other alternative for emergency vascular access in newborns is the insertion of an intraosseous needle (Chapter 24.8).

While any of the umbilical vessels are available for vascular access, the umbilical vein is technically easier to cannulate. Therefore, use the vein in an emergent situation. Consider cannulating one of the umbilical arteries if invasive blood pressure monitoring and arterial blood gas sampling are going to be important for patient management.

Indications

- Emergent vascular access for resuscitation
- Central access for delivery of fluids, medications, or exchange transfusion
- Frequent blood sampling
- Haemodynamic monitoring.

Contraindications

Do not place an umbilical vessel line if a peripheral intravenous access is available and adequate for the infant's needs.

Equipment

Box 24.9.1 lists the equipment for cannulation of either the umbilical artery or vein.

Preparation

1. Place the infant supine under a radiant warmer with all extremities restrained.
2. Use cardiorespiratory and pulse oximetry monitors for the duration of the procedure.

Box 24.10.1 Equipment for cannulation of the umbilical artery or vein

- Sterile gown, gloves and mask
- Povidone-iodine solution
- Sterile gauze
- Sterile drapes
- Umbilical tape
- 3.0 silk suture with needle
- Scalpel
- Haemostats
- Smooth, curved iris forceps
- Iris scissors
- Needle driver
- Umbilical catheter (3.5 or 5.0 French)
- Saline and syringes, consider heparinised saline
- Three-way stopcock

3. Wash hands and don a mask, sterile gown and sterile gloves.
4. Prepare all equipment in advance; select appropriate-size catheter, and consider its markings relative to planned depth of insertion.
5. Flush the umbilical catheter with saline and attach a primed three-way stopcock.

Procedure

Umbilical vein catheterisation

1. Have an assistant hold up the umbilical stump, and and scrub the umbilicus and abdomen broadly with povidone-iodine solution from xiphoid to pubis. Povidone-iodine remains preferred in neonates as it is less irritating to newborn skin.
2. Apply sterile drapes to cover the infant's torso, revealing only the umbilical stump; do not cover the infant's face (monitoring is crucial).
3. To provide haemostasis, gently tie umbilical tape at the base of the umbilical cord at the junction with the skin.
4. Using a scalpel, cleanly cut the cord approximately 1–2 cm away from the skin. Try to avoid a sawing method of cut as this can make vessel identification within the Wharton's jelly difficult.
5. Identify the two thick-walled arteries and the larger, thin-walled umbilical vein (Fig. 24.10.1).
6. Holding the upper edges of the umbilical cord (Wharton's jelly) with fine haemostats, insert the flushed 3.5 F (preterm) or 5 F (term) catheter into the umbilical vein (Fig. 24.10.2).
7. Use gentle but steady pressure while inserting the catheter, and provide gentle upward counter-traction on the cord using the haemostat if resistance is met.

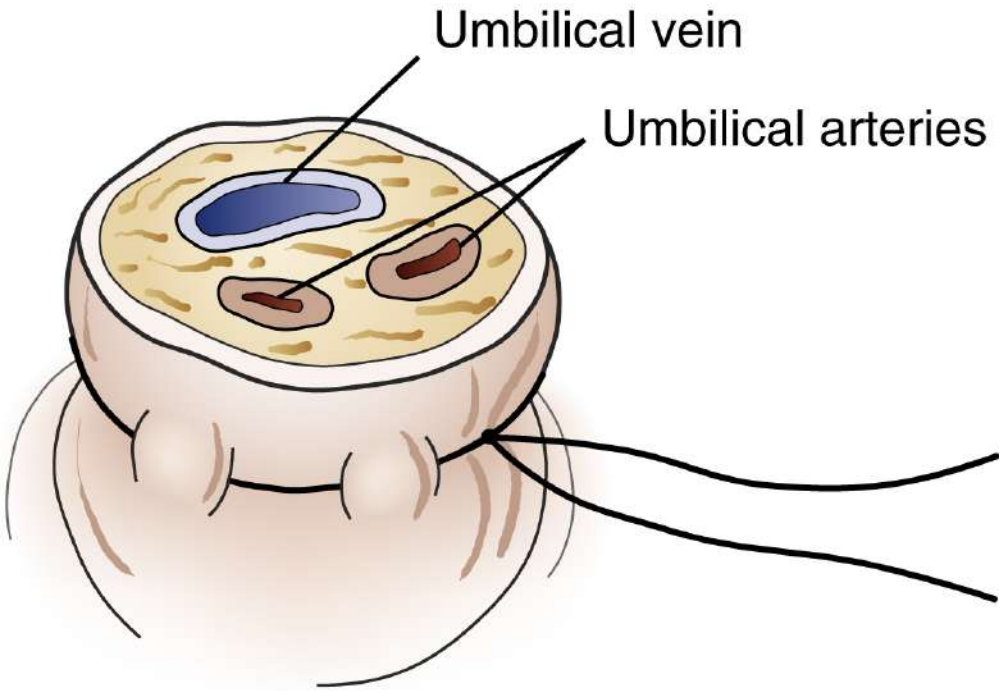

FIG. 24.10.1 Identifying the umbilical arteries and vein.

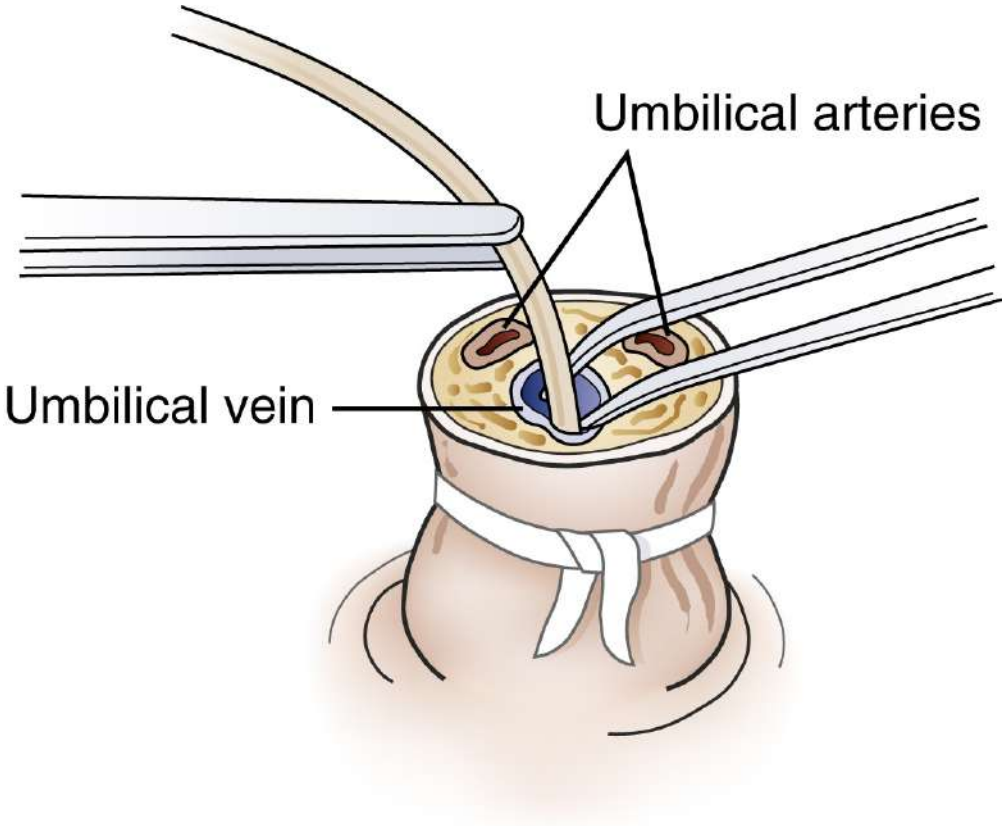

FIG. 24.10.2 Inserting the catheter into the umbilical vein.

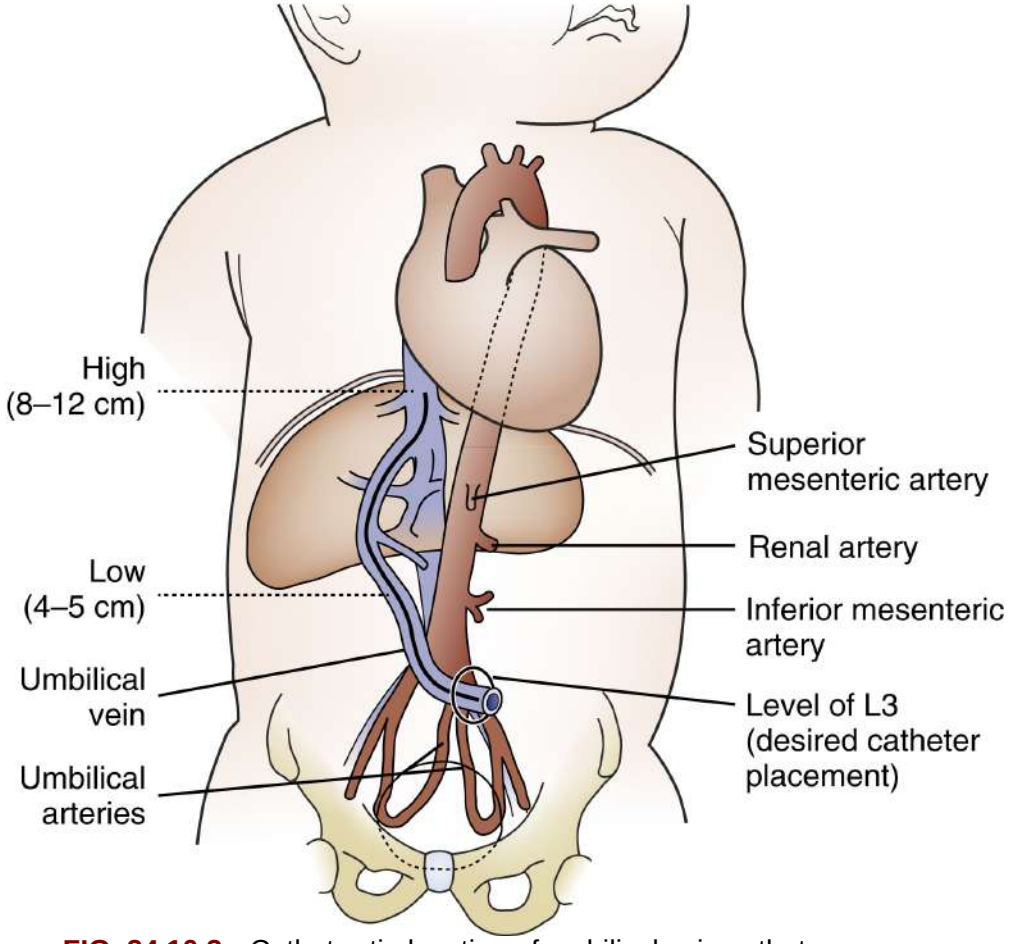

FIG. 24.10.3 Catheter-tip location of umbilical vein catheter.

8. Insertion distance:

In emergency situations, insert a **low umbilical vein catheter (UVC)**, which sits in the subhepatic umbilical vein below the portal vein. This will only be inserted approximately 3–5 cm beneath the abdominal wall (5–7 cm including the umbilical stump). Blood should return readily through the catheter when aspirated (Fig. 24.10.3).

Alternately, for a longer term catheter, placement in the vena cava above the liver is preferred. To aim for a **high UVC** above the diaphragm (via the portal system) insert to a predetermined depth as estimated by:

$$\text{Desired distance(cm)} = \text{weight} \times 1.5 + 5 + \text{stump}$$

The deeper (high) line allows for administration of hyperosmolar and irritating fluids as well as central venous pressure monitoring. Confirm that blood returns freely through the catheter prior to infusion of any fluids or medications. A high line must have its position checked radiographically prior to use.

9. Anchor the catheter to the umbilical cord with a suture, and secure the catheter to the abdomen with tape.
10. Confirm catheter position radiographically:
    - T8–10 high line
    - T12–L1 low line (in resuscitation situation, do not delay therapy for X-ray; if blood aspirates freely then use the line).

Umbilical artery catheterisation

1. Prepare and drape the abdomen and umbilical cord as with umbilical vein catheterisation.
2. Loosely tie umbilical tape at the base of the umbilical cord at the junction with the skin, and cut the cord approximately 1–2 cm from the skin in a similar clean manner as above.
3. Identify the two thick-walled arteries and attach two fine-tipped haemostats to either side of the umbilical cord to stabilise the vessels (Fig. 24.10.4). Do not place the haemostats on the arteries.
4. Insert the smooth curved iris forceps into the selected artery to gently dilate it (Fig. 24.10.5).
5. Continue dilating until the forceps can be inserted about 1 cm, remove the forceps and gently introduce and advance the pre-flushed catheter (3.5 preterm; 5.0 term) towards the patient's feet with mild cephalad traction on the cord (Fig. 24.10.6).
6. Overcome any resistance with gentle steady pressure, but do not force the catheter.

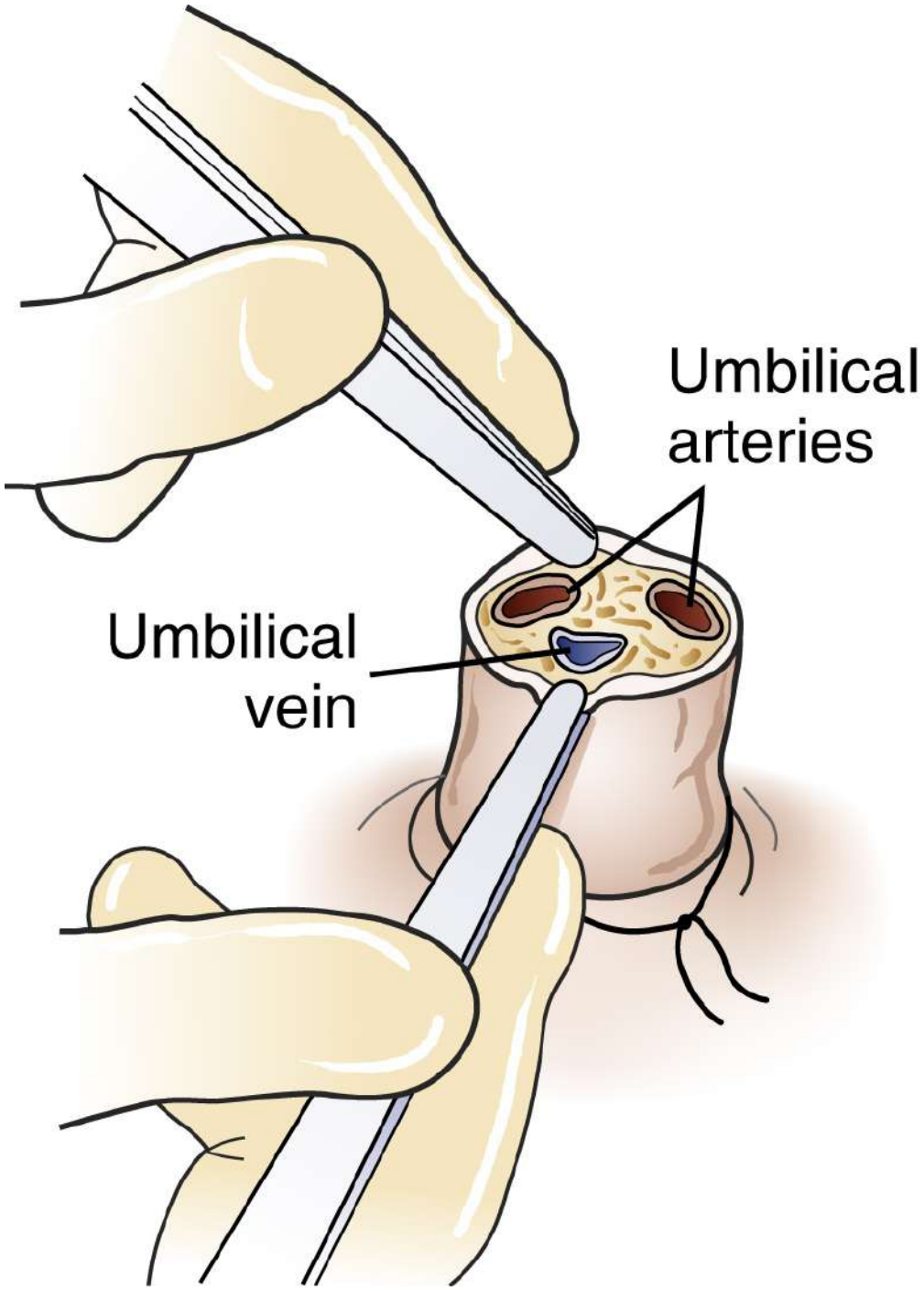

FIG. 24.10.4 Stabilising the vessels.

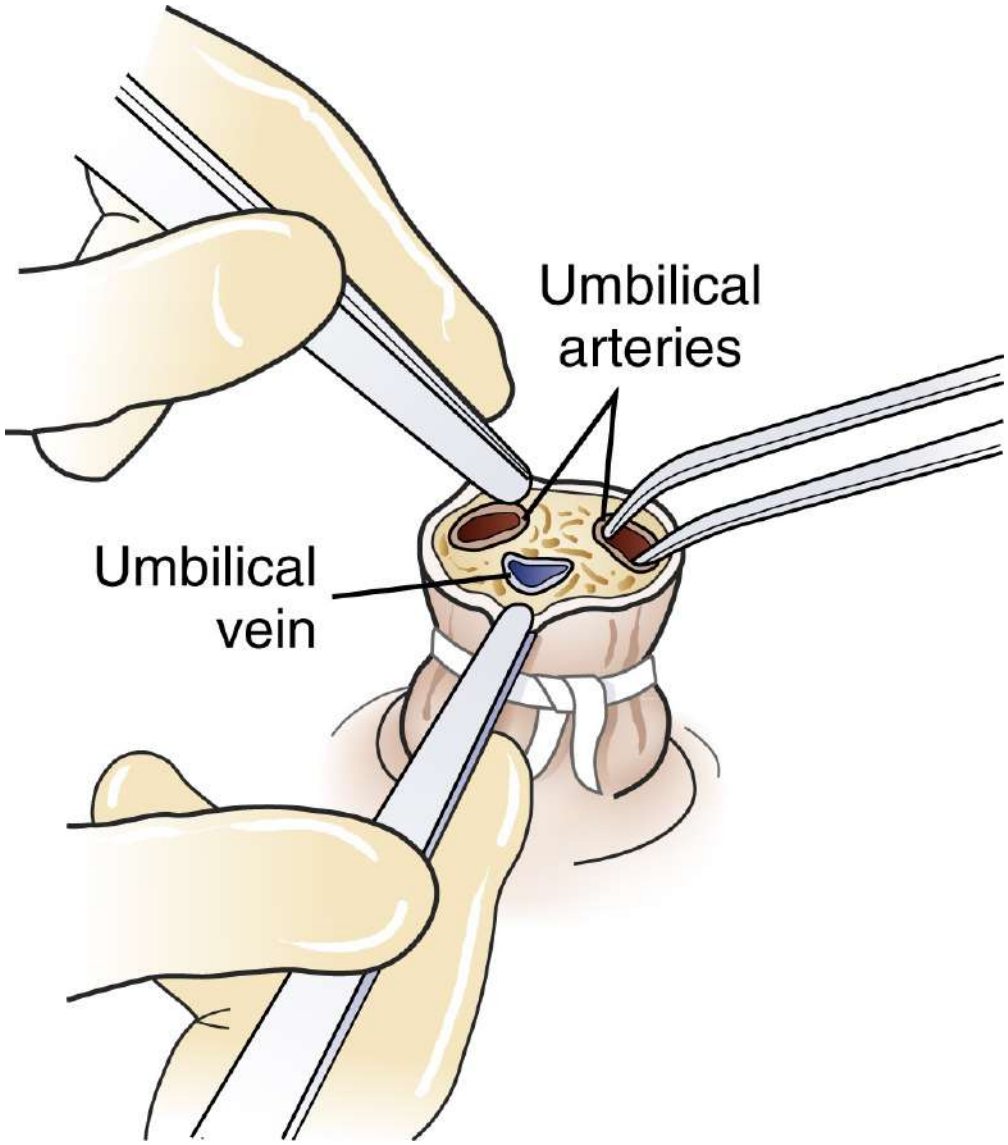

FIG. 24.10.5 Inserting the iris forceps into the artery.

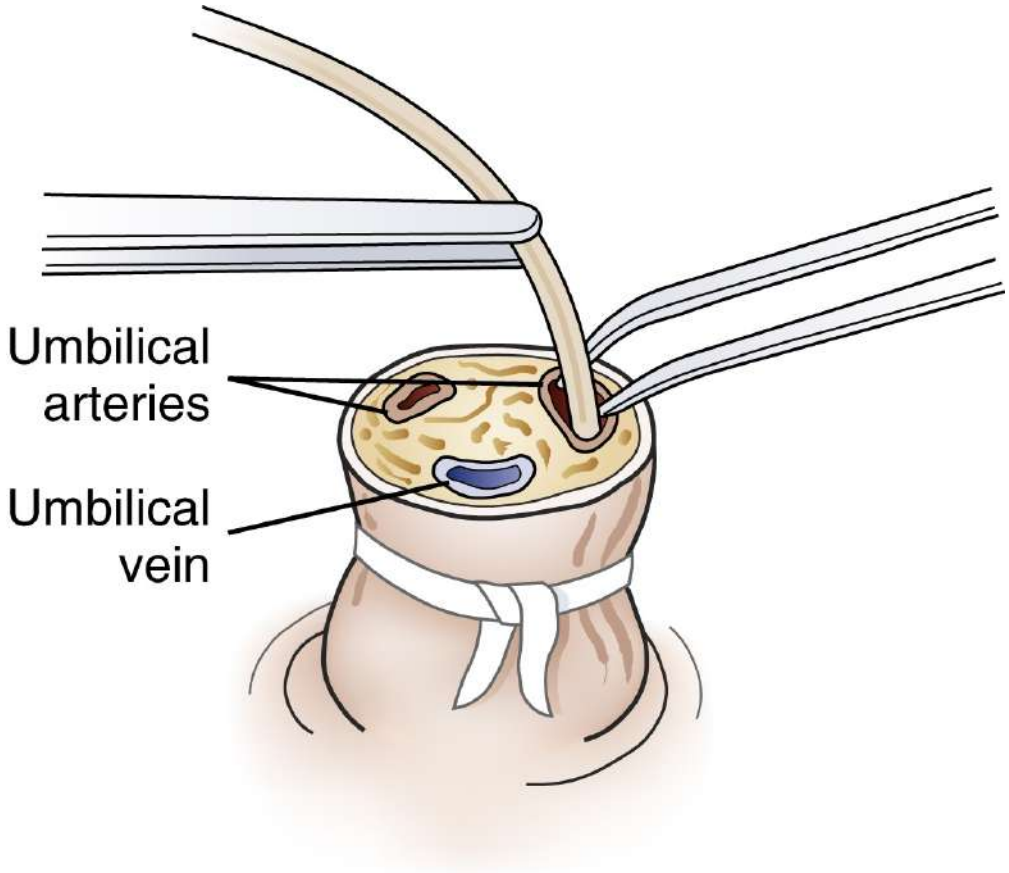

FIG. 24.10.6 Advancing the umbilical artery catheter.

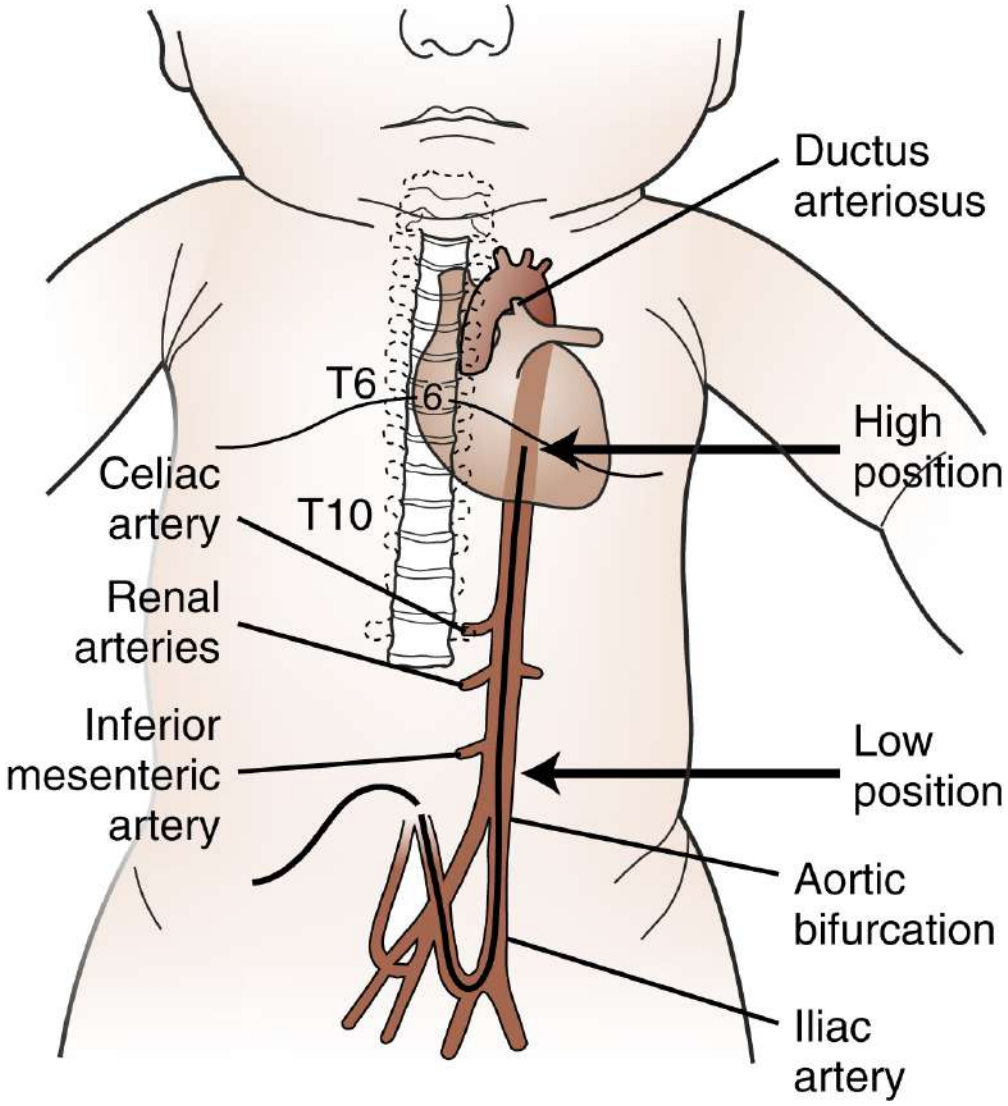

FIG. 24.10.7 Catheter tip location umbilical artery catheter.

7. One would generally always aim for a high umbilical artery catheter (UAC) placement rather than a low UAC (Fig. 24.10.7). Low catheter insertion distances are based on patient weight and are available on nomograms. High UAC insertion distance can be calculated by:

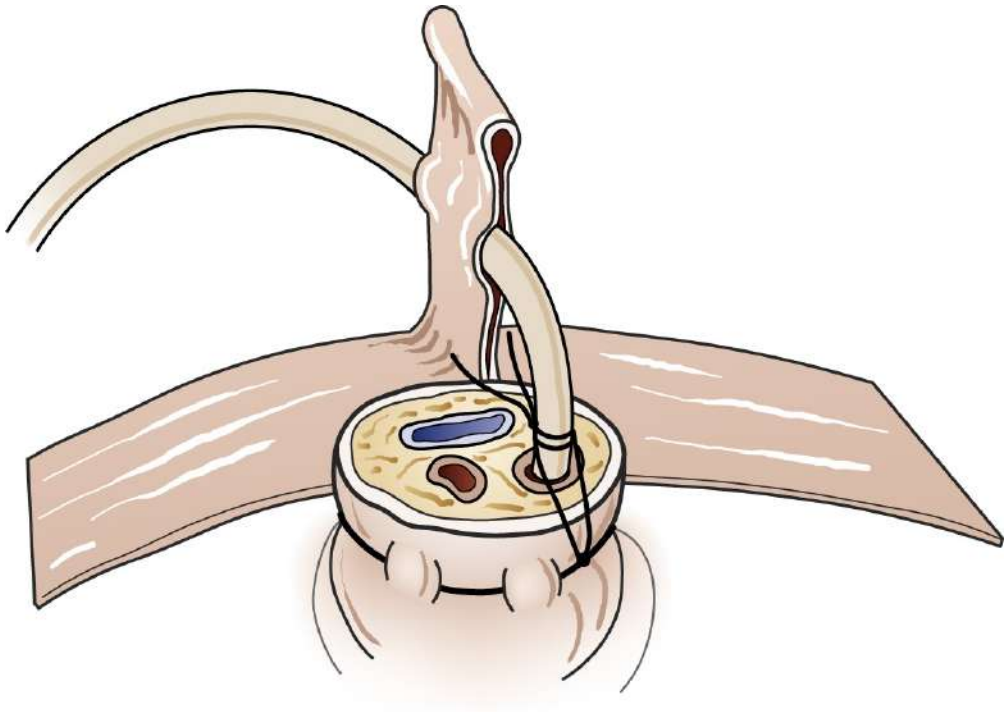

FIG. 24.10.8 Anchoring the catheter to the umbilical cord.

$$\text{UAC} = 3 \times \text{weight} + 9\ \text{cm} + \text{stump}$$

8. Anchor the catheter to the umbilical cord with a suture, and secure the catheter to the abdomen with tape (Fig. 24.10.8).
9. Verify placement of the radio-opaque catheter by thoraco-abdominal X-ray (Fig. 24.10.7):
    - High UAC between the T6 and T9 vertebrae
    - Low UAC at level of L3 vertebra.

Complications

- Haemorrhage
- Infection
- Air embolism
- Vessel dissection

- Perforation
- If umbilical venous catheters are left in the liver, sclerosing substances injected via the catheter can cause hepatic damage.
- Thromboembolic events, particularly to kidneys, intestines and the lower extremities. If blanching or cyanosis of the lower extremities occurs, remove the catheter immediately.

Tips

- Always advance catheters slowly and use only gentle pressure if resistance is encountered.
- Aggressive catheter insertion can result in creation of a false lumen or perforation of a vessel.
- Always confirm catheter position radiographically.
- If an umbilical venous catheter is needed emergently (i.e. prior to radiographic confirmation of position) place it in a low position to avoid accidental injection of sclerosing medications directly into the liver.
- High catheter insertions appear to be associated with fewer complications, but the position MUST be confirmed by X-ray prior to use.
- A catheter can be withdrawn to alter position but should never be pushed in farther once the initial sterile procedure is complete.

Further reading

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24.11

Defibrillation

Holly Smith

Scott Schofield

Background

Resuscitation from pulseless cardiac arrest rhythms in children includes ventilation, oxygenation, effective chest compressions and, in certain instances, delivery of an asynchronous electrical shock to the myocardium (defibrillation) in an effort to reset the heart's intrinsic automaticity. Cardiac arrest rhythms (and hence resuscitation protocols) are typically divided into 'shockable' and 'non-shockable' rhythms. Shockable rhythms include ventricular fibrillation (VF) and pulseless ventricular tachycardia (VT). Ventricular arrhythmias represent approximately 5–10% of presenting paediatric cardiopulmonary arrest rhythms. Non-shockable rhythms of asystole, pulseless electrical activity and bradycardia are not managed using defibrillation. Detailed resuscitation protocols are discussed in another chapter, with this section being dedicated to the steps of delivering safe defibrillation to the arrested child.

Certain non-arrest rhythms can also be treated using a lower energy shock that is delivered synchronously with the QRS complex (i.e. supraventricular tachycardia [SVT], ventricular tachycardia with a pulse). This is called synchronised cardioversion and follows a similar procedure to defibrillation, albeit at a modified dose and with the synchronous function activated on the defibrillator. The procedure herein can be used for such cases.

Current technology for defibrillators

Biphasic devices capable of providing biofeedback on timing and quality of compressions, as well as a record of the underlying rhythm, are available and

advised.

The electrical physiology of the shock given can have a monophasic or biphasic waveform. In older monophasic defibrillators, the shock is delivered in only one direction (vector) from one electrode to the other, with no ability to measure tissue (chest) impedance. With current biphasic defibrillators, the shock is delivered sequentially along two vectors, allowing for measurement of thoracic impedance and modification of their internal resistance to deliver the prescribed electrical energy with less damage to the myocardium. Biphasic waveforms were initially developed for use in implantable cardioverter defibrillators (ICD) and later adapted to external defibrillators; biphasic defibrillators are now considered gold standard.

Pads versus paddles

Current recommendations suggest the use of hands-free defibrillators with pads rather than paddles. Pads are self-adhesive and generally come in two sizes; infant (for patients <10 kg) and standard (for all >10 kg). If only standard pads are available, these can be used in all ages, but care must be taken to avoid overlap of the electrical component of the pads. Placement of the pads in infants is generally anterior and posterior (Fig. 24.11.1), and either antero-lateral or anterior-posterior in children 1–8 years (Fig. 24.11.2)**.** Over 8 years and in adults, the pads should be placed antero-lateral. If using paddles rather than pads, the paddles must be placed firmly on the chest over gel pads. The paddles should only be charged on the chest, and no cardiopulmonary resuscitation (CPR) should be performed during charging. Following delivery of the shock, the paddles should be immediately returned to the defibrillator.

Asynchronous versus synchronous

For certain non-arrest dysrhythmias, synchronised delivery of electricity is indicated. In these instances, the user must activate the synchronous mode (SYNC), which marks the contractions. Charging procedure is unchanged; however, the dose is reduced to 0.5–2 J/kg. Delivery of the shock involves holding the ‘discharge’ button for a longer period of time (up to 5 seconds) to allow the machine to deliver the shock in time with the peak of a recognised QRS complex. If no shock is delivered, then the QRS was not sensed; reassess the patient as well as your equipment, and reconsider options.

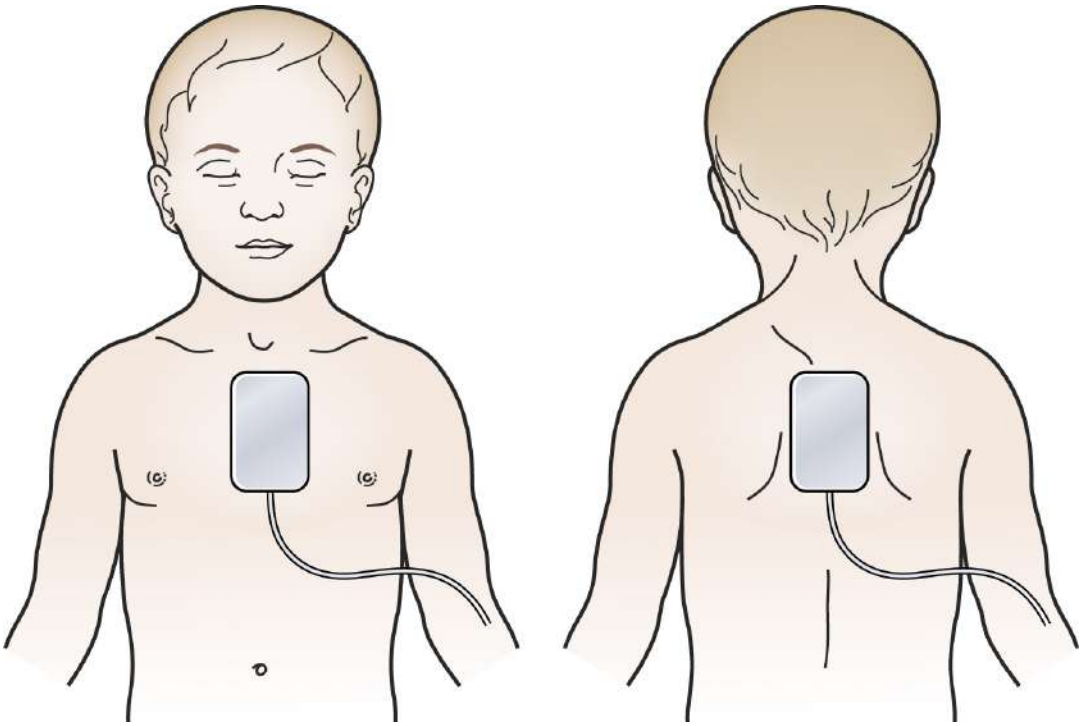

FIG. 24.11.1 Placement of pads on the infant or small child's chest and back (antero-posterior).

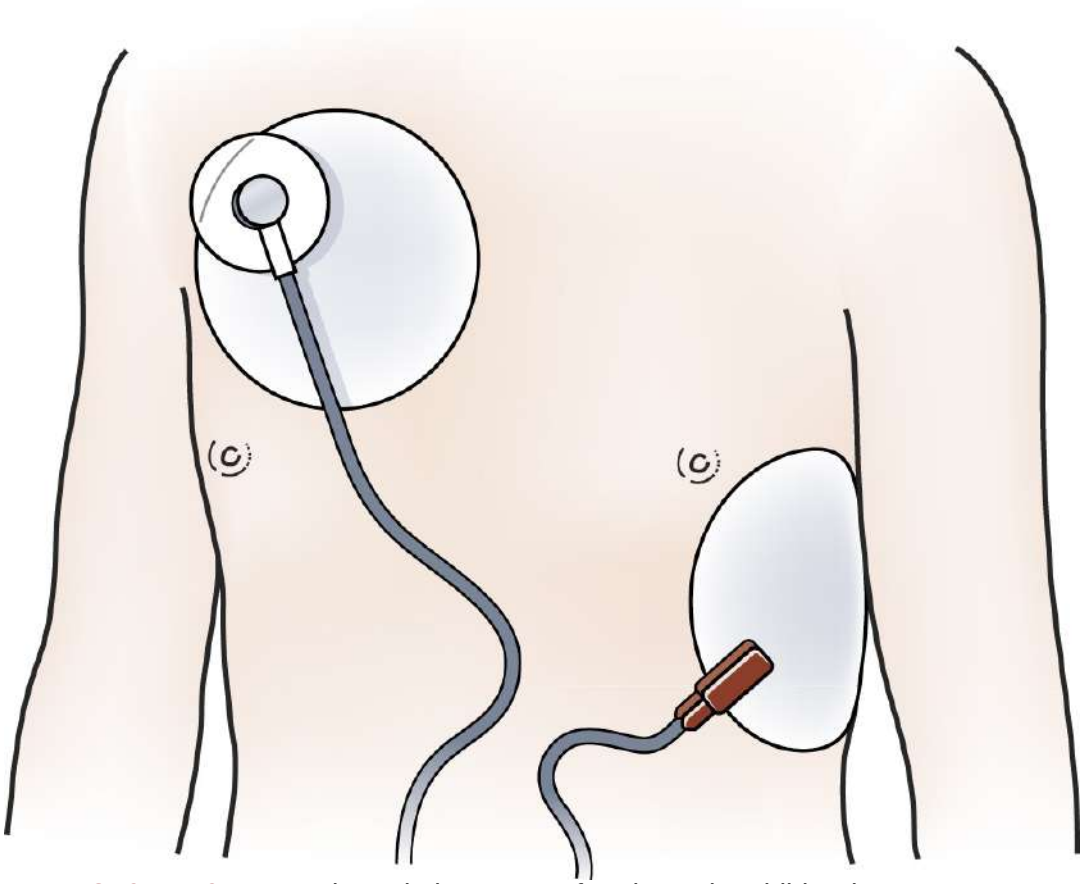

FIG. 24.11.2 Anterolateral placement of pads on the child's chest.

The International Liaison Committee on Resuscitation (ILCOR) compiles guidelines and recommendations on resuscitation from cardiac arrest. Local protocols and guidelines should be adhered to.

Indications for defibrillation (asynchronous)

- VF
- Pulseless VT.

Indications for synchronous cardioversion

- SVT with evidence of shock
- VT with shock and pulse present
- Atrial fibrillation or atrial flutter with shock
- Stable VT or SVT in collaboration with specialist.

Contraindications

- Conscious patient with good perfusion.

Equipment

1. Standard defibrillator (generally biphasic in major centres)
2. Automated external defibrillator (AED):
    - Newer models feature lower power outputs to deliver lower energy shock for children.
3. Defibrillator pads (appropriately sized).

Standard preparation

Basic life support (BLS) and advanced life support (ALS) should be performed according to local published guidelines (ILCOR). Discussion about current resuscitation guidelines can be found elsewhere in this text:

1. Assess for danger. Check for patient's responsiveness. Send for help.
2. Open airway, check for breathing and commence respiratory support using basic and/or advanced airway support measures. Monitor saturations and end-tidal CO_2 if available.
3. Check for pulse and signs of life. Begin chest compressions at a rate of 100 compressions per minute to a depth of one-third to half of the chest. Coordinate ventilations and compressions in accordance with guidelines.
4. Apply available monitoring and defibrillator pads of appropriate size, in the desired positions. Pad placement is generally anteroposterior in infants and anterolateral in children over 10 kg. Pads generally have desired positions pictorially printed on them. Know your local equipment. Generally, a shockable rhythm should be anticipated in a pulseless patient, and defibrillator pads prepared and applied upon recognition of pulselessness (along with effective CPR). Delays to CPR should be minimised.
5. Turn on the defibrillator, and do *not* activate the synchronised mode for shockable cardiac arrest rhythms (asynchronous is the default for most defibrillators). For SVT, VT with a pulse, atrial fibrillation or flutter, use

of the synchronised mode involves turning ON the synchronised function.

Standard procedure

Identification of the shockable rhythm should be made quickly and the appropriate algorithm adhered to. Outcome of shock delivery is best if rescuers minimise the time between the last compression and delivery of the electrical shock. Many jurisdictions have adopted some variation of the 'COACHED' algorithm, which outlines the steps for efficient, effective and safe defibrillation, with minimal interruptions to CPR (Fig. 24.11.3). The COACHED algorithm fits nicely with the shockable rhythm protocol. A single person on the team should be tasked with being in charge of the defibrillator, and the whole team should be aware of who this is.

Coached

1. The person in charge of the defibrillator reminds everyone that '**C**ompressions should continue' until otherwise advised. Uninterrupted high-quality CPR is the goal.
2. The person in charge of the defibrillator says 'remove free flowing **O**xygen'; at this point any free flowing oxygen must be removed from the patient. A brief but accepted period of apnoea will follow.
3. The person in charge of the defibrillator says '**A**ll others stand clear'; at this stage everyone EXCEPT the person providing chest compressions should step back from the patient. Chest compressions should continue.
4. **C**harge the defibrillator to a target dose of 4 J/kg. The calculated energy dose will not likely be available on the defibrillator. In this case round UP to the next nearest dose. For example, dosage for a 14 kg child would be 56 J; the closest available energy selection might be 60 J. This differs between machines. Note the sound your machine makes during charging and when it is ready to be delivered. (For synchronised cardioversion, the dose is 0.5–2 J/kg with the SYNC function activated.)
5. The person in charge of the defibrillator tells the person providing chest compressions '**H**ands off'. At this point the person delivering CPR should stop compressions, raise his or her hands and respond 'I am safe'.

6. A formal and verbal **E**valuation of the patient's rhythm should be made at this point. For example, 'this is ventricular fibrillation, which is a shockable rhythm'.
7. The person in charge of the defibrillator should press the shock/discharge button to **D**efibrillate the patient if he or she is in a shockable rhythm. Alternately, the charge should be dumped and the defibrillator disarmed if the child is in a non-shockable rhythm. Different machines allow for the charge to be dumped in different ways (e.g. by pressing on the dose dial); learn the method to disarm the defibrillator in your setting.

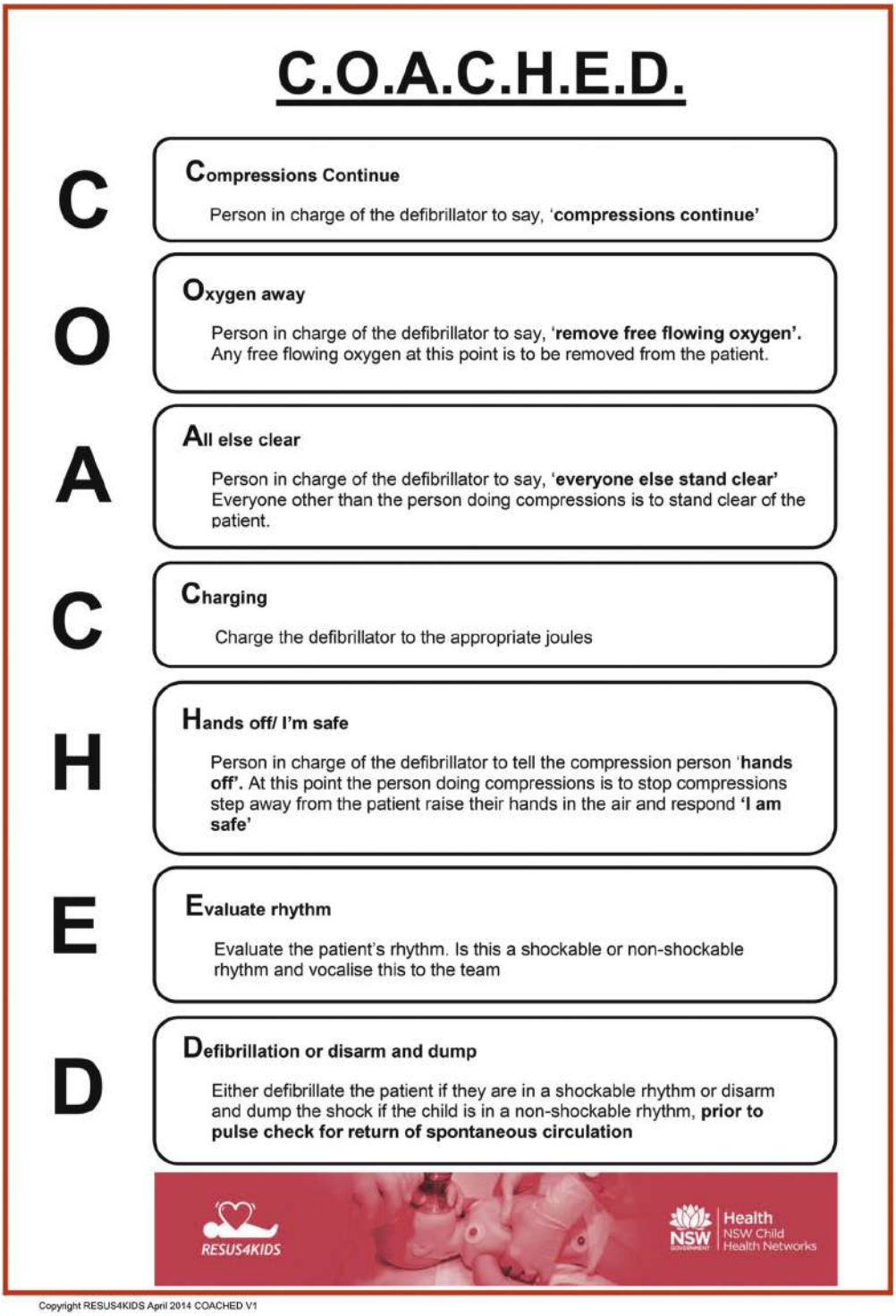

FIG. 24.11.3 COACHED algorithm for safe defibrillation, with permission from Dr Fenton O'Leary, Resus4Kids.

8. CPR should be resumed immediately following delivery of the shock and continued for 2 minutes until repeat of the above steps is indicated. During this time, other measures should be continued as per the

‘shockable rhythm protocol’ (i.e. advanced airway measures, intravenous cannulation or intraosseous cannula insertion, preparation of resuscitation drugs, and consideration and management of cause).

9. Treat reversible causes, remembering the ‘Hs’ and the ‘Ts’: Hypoxia, Hypovolaemia, Hypo/hyperkalaemia, Hypothermia, Tension pneumothorax, Tamponade (cardiac), Toxins, Thromboembolism.
10. Treat other rhythms that develop such as PEA or asystole (i.e. shift to the non-shockable rhythm protocol.

Automated external defibrillators procedure

Automated external defibrillators (AEDs) can be used in arrested children over 1 year of age. Makes and models of AEDs vary with respect to sensitivity in recognising paediatric arrhythmias and ability to attenuate energy output. If an AED is likely to be used for children, teams should check that the performance of the particular model has been tested against paediatric arrhythmias and can recognise them with reproducibility. Specific paediatric pads and/or programmes that attenuate the energy output of the AED are recommended for children aged 1–8 years. However, if these are not available, it is recommended that an unmodified adult AED be used in children over 1 year, rather than delay the delivery of a potentially life-saving intervention.

In children aged less than 1 year, there is little evidence regarding the safety and efficacy of the use of AEDs in general (partly owing to the rarity of shockable rhythms in this age group), and the decision to proceed with AED must be taken in the context of what other equipment is available. In the setting of an arrested infant where an AED is available, the risk/benefit balance will likely favour the use of the AED:

1. Open airway ventilate while preparing equipment.
2. If child is pulseless, begin chest compression.
3. Turn AED on.
4. Attach AED electrodes to chest wall (right clavicle, and one in the left anterior axillary line, under the left nipple).
5. Allow the device to analyse rhythm.
6. If defibrillation is indicated, clear the area and deliver electricity.
7. Do CPR and check airway, breathing and circulation for 2 minutes, then re-analyse rhythm and deliver additional shocks as indicated.

Complications

- Ineffective delivery of defibrillation because of failure to charge, improper positioning on the chest, incorrect pad or paddle size
- Burns on the chest wall
- Failure to ‘clear’ before voltage discharge, leading to electrical shock of a team member or bystanders (rare)

Table 24.11.1

Appropriate electrical charge for countershock

Dysrhythmia	Mode	Charge
Ventricular fibrillation Pulseless ventricular tachycardia (VT)	Asynchronised (defibrillation)	4 J/kg
VT with pulse Supraventricular tachycardia with shock Atrial fibrillation and atrial flutter with shock	Synchronised	0.5–2 J/kg *

* Synchronised dose varies with jurisdiction.

- Tachydysrhythmias
- Bradycardia
- Myocardial damage or necrosis
- Cardiogenic shock
- Embolic phenomena.

Tips

- Biphasic defibrillators have better speed, safety and efficiency on the first shock than older monophasic defibrillators.
- Minimise the interval between stopping compressions and delivering shocks, and always resume CPR immediately after shock delivery. The COACHED algorithm helps to achieve this.
- The correct energy dose for defibrillation (with either a monophasic or biphasic device) in infants and children is unknown but is extrapolated from adult and animal models. When shocks are indicated for VF or pulseless VT, using an initial energy dose of 4 J/kg of either waveform is considered safe. Doses higher than 4 J/kg, especially if delivered with

a biphasic defibrillator, may also be safe and effective.

- When performing chest compressions, push hard, push fast and minimise interruptions of chest compression; allow full chest recoil and don't provide excessive ventilation.
- For a child with VF or pulseless VT, use the asynchronised mode – the defibrillator will not discharge in the synchronised mode.
- Self-adhesive electrode pads are simpler, safer and more efficient than the conventional paddles when defibrillating or cardioverting.
- If the child has SVT or VT with a pulse and is showing sign of cardiogenic shock, proceed to synchronised cardioversion at a dose of 0.5–2 J/kg. This is done by turning ON synchronous mode on the defibrillator, selecting the desired Joules and holding the discharge button until the shock is delivered in time with the peak of the QRS complex.
- Alert patients requiring synchronised cardioversion should have appropriately considered analgaesia and/or sedation prior to delivery of charge. Arrested patients being managed under the shockable rhythm protocol are unconscious and do not require analgaesia/sedation as this would delay definitive treatment.

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24.12

Transurethral catheterisation and suprapubic bladder aspiration

Scott Schofield, and Holly Smith

Background

Urinary tract infections (UTI) are a common cause of fever in children and source for serious bacterial infections in infants. Obtaining an uncontaminated specimen of urine is essential to accurately diagnose a UTI. Older children will be able to provide a mid-stream urine sample for urinalysis, microscopy and culture. Obtaining a clean urine specimen from infants or children who cannot provide mid-stream specimens often requires a procedure to sample directly from the bladder. Attempts to obtain urine specimens via bags attached temporarily to the perineum or by more recently reported bladder stimulation techniques are frequently unsuccessful and produce contaminated specimens.[1–5]

Obtaining an appropriate urine specimen in a child unable to provide a mid-stream sample requires either transurethral catheterisation or suprapubic aspiration. Successful transurethral catheterisation is variable and dependent on procedural experience as well as anatomical variance. When transurethral catheterisation has been or is likely to be unsuccessful, suprapubic aspiration of urine directly from the bladder should be considered. Suprapubic aspiration provides a sterile urine specimen, often with less trauma to the infant than repeated failed attempts at urethral catheterisation.

Transurethral catheterisation and placement of an indwelling catheter (IDC) are also useful in the management of critically ill and injured children, to monitor urine output in the setting of shock or trauma. Urine output is a useful measure of end organ perfusion and can help guide fluid resuscitation of a child in shock.

Indications

Indications for transurethral catheterisation

- Collection of a diagnostic urine specimen
- Intermittent bladder decompression for neurogenic bladder
- Urological imaging
- Measurement of urine output in critically ill or injured child
- Acute urinary retention.

Indications for suprapubic bladder aspiration

Collection of a sterile diagnostic urine specimen when transurethral catheterisation has been or is likely to be unsuccessful.

Contraindications

Urethral catheterisation

- Suspected pelvic trauma
- Blood at the urethral meatus
- Upward displacement of the prostate in males following trauma
- Perineal haematoma.

Suprapubic aspiration

- Coagulopathy
- Thrombocytopenia
- Significant abdominal distension
- Massive organomegaly
- Recent abdominal surgery
- Low bladder volume (<10 mL) identified with bladder scan or point-of-care ultrasound (Fig. 24.12.1).

Transurethral catheterisation

Equipment

The equipment required for transurethral catheterisation is often available in prepackaged trays to which only the appropriately sized catheter need be added. Remember to use lignocaine jelly as a local anaesthetic before inserting a catheter, particularly when catheterising the older male patients:

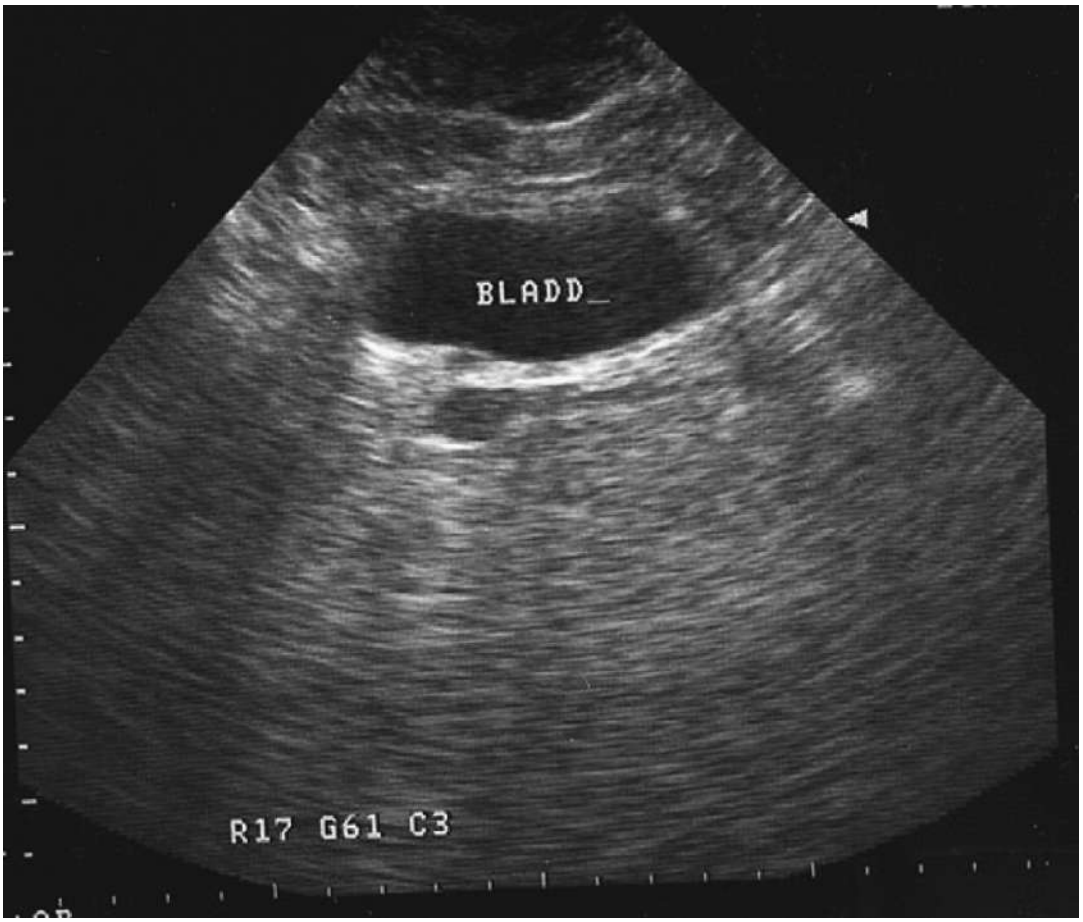

FIG. 24.12.1 Using ultrasound for suprapubic aspiration.

- Dressing trolley
- Catheterisation pack and drapes
- Sterile gloves
- Appropriate-size catheter (Table 24.12.1)
- Xylocaine (lignocaine) jelly syringe, or plain sterile lubricant for infants
- Sterile water for balloon (if inserting IDC)
- 5 mL syringe

- Specimen jar
- Appropriate cleaning solution (e.g. chlorhexidine 0.1%)
- Drainage bag and tape to secure catheter to leg (if inserting IDC).

Preparation

1. Review the male and female anatomy of the perineum
2. Determine the appropriate catheter size by using a length-based resuscitation tape or by using a table listing equipment sizes by age (Table 24.12.1)
3. Have all equipment ready near the child
4. Place the appropriately sized catheter and the syringe with lignocaine jelly on the sterile field
5. Have an assistant gently restrain the child in a frog-leg position to prevent disruption of the sterile field and unnecessary trauma due to patient movement
6. If the child is older, discuss the procedure, clearly relating each step.

Procedure

1. Use sterile technique
2. With a male patient, use the non-dominant hand and a gauze to retract the foreskin and expose the meatus. In the circumcised male, the gauze is still useful to maintain a firm grasp on the penis during catheterisation.
3. With a female patient, use the thumb and forefinger of the non-dominant hand to spread the labia majora and expose the urethra.
4. Clean the urethral meatus three times with cleaning solution, making sure to swab from front to back in females to avoid contamination.
5. Insert a few millilitres of lignocaine jelly, using a syringe device to provide topical anaesthesia, particularly in male patients.
6. Lubricate the catheter with the same jelly, ensuring the end is not occluded by lubricant.

Table 24.12.1

Indwelling urinary catheter size guide

Indwelling urinary catheter size guide

Catheter size guideline
Use the smallest bore that will allow good drainage to minimise bladder and urethral trauma.
Consider silicone catheter if for long-term use.

Age	Weight	Foley
Neonate	< 1200 g	3.5 French umbilical catheter
Neonate	1200–1500 g	5 French umbilical catheter
Neonate	1500–2500 g	5 French umbilical catheter or size 6 Foley
0–6 months	3.5–7 kg	6
1 year	10 kg	6–8
2 years	12 kg	8
3 years	14 kg	8–10
5 years	18 kg	10
6 years	21 kg	10
8 years	27 kg	10–12
12 years	Varies	12–14

Royal Children's Hospital, Melbourne, Australia, Clinical Practice Guideline on Indwelling urinary catheter - insertion and ongoing care [First published March 2012, most recently revised August 2017] Available From http://www.rch.org.au/rchcpg/hospital_clinical_guideline_index/Indwelling_urinary_catheter_isize-guideline.

7. Insert the catheter into the urethra. Do not force the catheter against resistance. Once urine flows through the catheter, stop advancing.
8. Discard the first 1 mL of urine, which may be contaminated, and collect a specimen.
9. If the catheter is being left in, inflate the balloon and gently pull back until resistance is felt. Inflate the balloon with the amount of fluid listed on the catheter.
10. Connect the catheter to a sterile closed drainage system.
11. Secure the catheter. With a balloon-tipped catheter, secure to the inner thigh with tape. With non-balloon catheters, tape the tube to the shaft of the penis in a spiral fashion or to the proximal inner thigh next to the labia majora in females.
12. Remember to return the foreskin to its original position in uncircumcised males to prevent paraphimosis and penile constriction.

Complications

- Urinary tract infection (more common with IDC)
- Urethral or bladder trauma
- Haematuria
- Paraphimosis
- Vaginal catheterisation
- Urethral strictures.

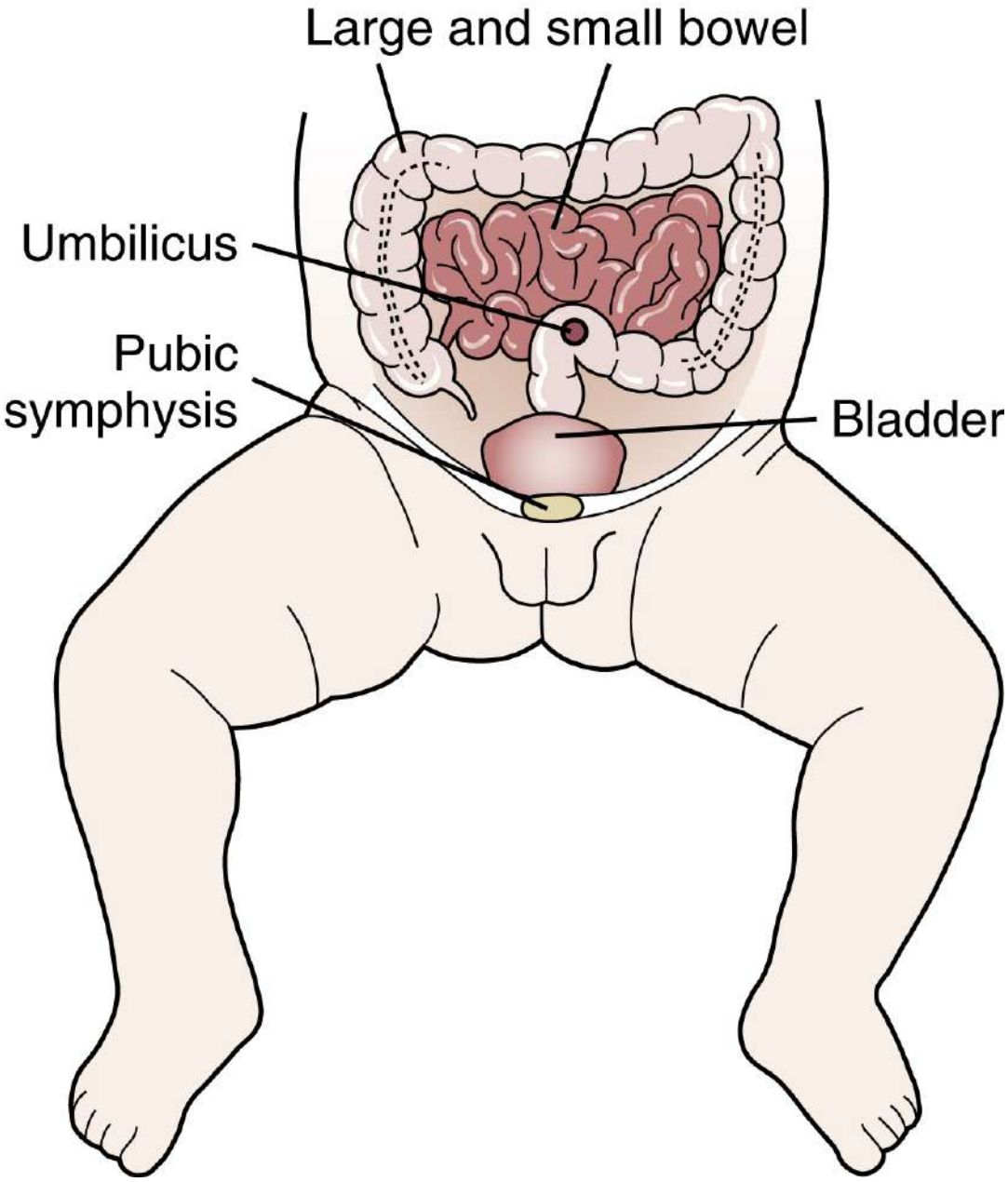

FIG. 24.12.2 Anatomy of the lower abdomen.

Tips

- There are circumstances in which urine specimens can be obtained non-invasively using urinary bags. This collection technique may be appropriate for investigation or diagnosis of conditions other than infection.

Suprapubic aspiration

Equipment

- Topical anaesthetic
- Bladder scanner or point of care ultrasound
- Sterile gloves
- 3–5 mL syringe
- 23 gauge needle (25 g for premature neonate)
- Cleaning solution (alcohol swab or chlorhexidine 0.1%).

Preparation

1. Review the anatomy of the lower abdomen (Fig. 24.12.2)
2. Restrain the child in a frog-leg position
3. Have all equipment nearby.

Procedure

1. Prepare the area with topical anaethetic unless procedure is urgent (e.g. investigation prior to antibiotic administration for sepsis).
2. Be ready to catch an opportunistic midstream specimen during preparation and procedure.
3. Locate a spot approximately 1 cm cephalad of the pubic symphysis in the midline.
4. Use ultrasound to determine if the bladder is full enough. If the bladder diameter is 3.5 cm or if 10 cm^3 of urine is present on a volumetric bladder scanner, the procedure is likely to be successful.
5. Prepare the skin with cleaning solution.
6. Using a 3–5 mL syringe attached to appropriate sized needle, insert the needle at an angle approximately 10–20 degrees cranially (Fig. 24.12.3).
7. Provide constant backpressure on the syringe.
8. Aspirate the urine specimen, remove the needle and place a small bandage on the site.

Complications

- Bowel perforation. This is a rare complication. The only reports of bowel penetration are in children with markedly distended abdomens. Even if needle penetration of the bowel occurs, there is no treatment and infection is extremely unlikely.
- Infections around the puncture site can occur, but are unusual with appropriate cleaning.
- Microscopic haematuria (<10 RBC per high powered field) can occur transiently following aspiration.

Tips

- Ultrasound guidance of suprapubic aspiration improves the success rate of the procedure considerably, from approximately 50% to close to 90%. The ultrasound is performed to determine whether there is enough urine present for a suprapubic tap to be successful, not to direct needle placement. The ultrasound is performed using gel placed on the ultrasound probe head to improve the quality of the image. If the bladder contains greater than 20 mL of urine the suprapubic aspiration should be successful. If the volume is less than this, or the bladder diameter is less than 3.5 cm then give fluids and wait for more urinary volume to be present.

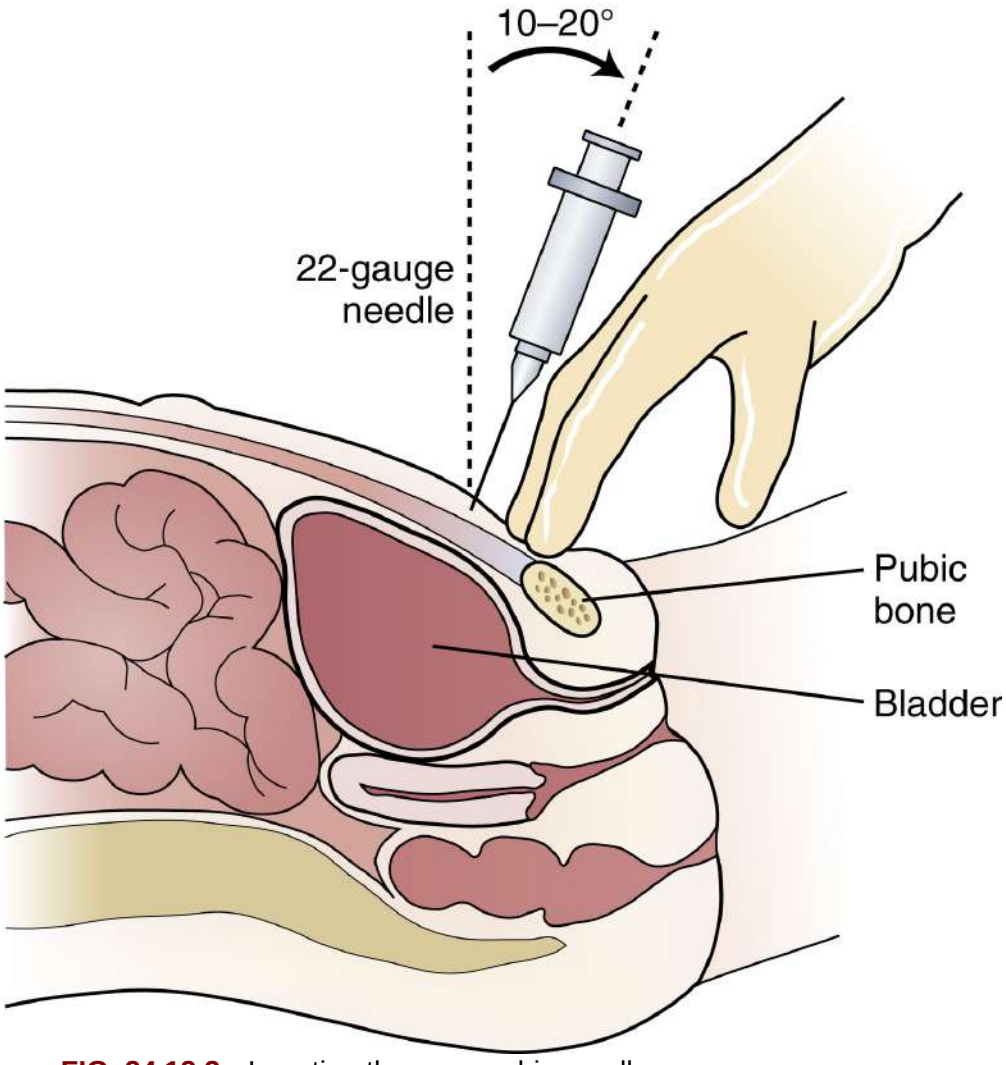

FIG. 24.12.3 Inserting the suprapubic needle.

- A bladder scan device can be used if ultrasound equipment or expertise is not available. This technique is not as accurate, particularly with small bladder volumes (<20 mL).
- It is a good tip to always have a container at the ready to collect a clean catch of urine as undressing an infant, temperature sensitivity or pressure with the probe in the suprapubic area may stimulate urination.

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24.13

Lumbar puncture

Scott Schofield, and Holly Smith

Background

Lumbar puncture (LP) is a time-honoured method for obtaining cerebrospinal fluid (CSF) for diagnostic evaluation of suspected central nervous system (CNS) abnormalities. LP is an essential diagnostic procedure in children with suspected meningitis; it is the only simple method of obtaining fluid for rapid diagnosis and appropriate pathogen analysis to guide specific treatment.

While LP is most commonly performed to diagnose meningitis, it is also a useful procedure to help identify encephalitis, CNS haemorrhage, malignancy, and other rare metabolic and degenerative conditions of childhood. Occasionally, LP is a therapeutic procedure in treatment of such conditions as pseudotumour cerebri, or for administration of intrathecal antibiotics or chemotherapeutic agents.

Lumbar puncture is a relatively simple procedure in infants and children but has a known complication rate, with both minor and major sequelae. Appropriate patient selection, preparation, positioning, and aseptic technique will avoid most complications. Do not perform LP immediately on haemodynamically unstable patients or in patients with evidence of elevated intracranial pressure and/or with focal clinical neurological signs; these children first require meticulous management of airway, breathing and circulation and early administration of antibiotics (following blood culture). LP can be deferred until after stabilisation and brain imaging, as indicated.

Indications

- Clinical symptoms and signs of meningitis or encephalitis in neonate, infant or child.

- Evaluation of sepsis in infant <1–3 months of age (depending on current local guidelines).
- Evaluation of seizure with fever in child <6 months or >5 years of age, multiple or prolonged seizures associated with fever in any child. Note that simple febrile convulsions do not routinely require an LP unless clinical suspicion of CNS infection. Also remember that LP should not be done during the ictal or immediate post-ictal phase (see contraindications).
- Intrathecal drug administration.

Contraindications

- Coma: absent or non-purposeful response to painful stimulus
- Signs of raised intracranial pressure (drowsy, diplopia, abnormal pupillary responses, unilateral or bilateral motor posturing, papilloedema, Cushing's response).
- Cardiovascular compromise/shock
- Respiratory compromise
- Focal neurological signs or seizures
- Active or recent seizures (within 30 minutes or not regained normal conscious level afterwards)
- Coagulopathy/thrombocytopenia.

In children with suspected elevation of intracranial pressure from oedema or space-occupying brain lesions, brain herniation with LP is an important risk. Consider doing a CT scan of the brain prior to LP. Note, however, that a normal CT scan does not completely rule out an elevated intracranial pressure (ICP); if there is ongoing concern about ICP, then LP should be deferred and treatment commenced.

Other relative contraindications include local skin infection at the entry site, and known or suspected cord tethering (i.e. unexplained dimple or pit above the natal cleft).

In general, if there are contraindications to LP and meningitis is suspected, do not defer appropriate steroid and antibiotic therapy.

Equipment

1. Pre-packaged LP trays are available in most hospitals.
2. 18, 20 or 22 gauge styleted spinal needles:
    - 3.5 cm (1.5 inch) for neonates, infants, young children
    - 6 cm (2.5 inch) for older children and adolescents
    - 9 cm (3.5 inch) for large patients
3. Chlorhexidine 0.5% in alcohol 70% prep solution or povidone-iodine solution.
4. 1% lignocaine [lidocaine]
5. 25-gauge needle and a 3-5 ml syringe.
6. Also consider a topical anaesthetic preparation (EMLA™, Angel™)
7. Four capped sterile specimen tubes
8. Manometer and stopcock (for children aged >2 years if indicated)

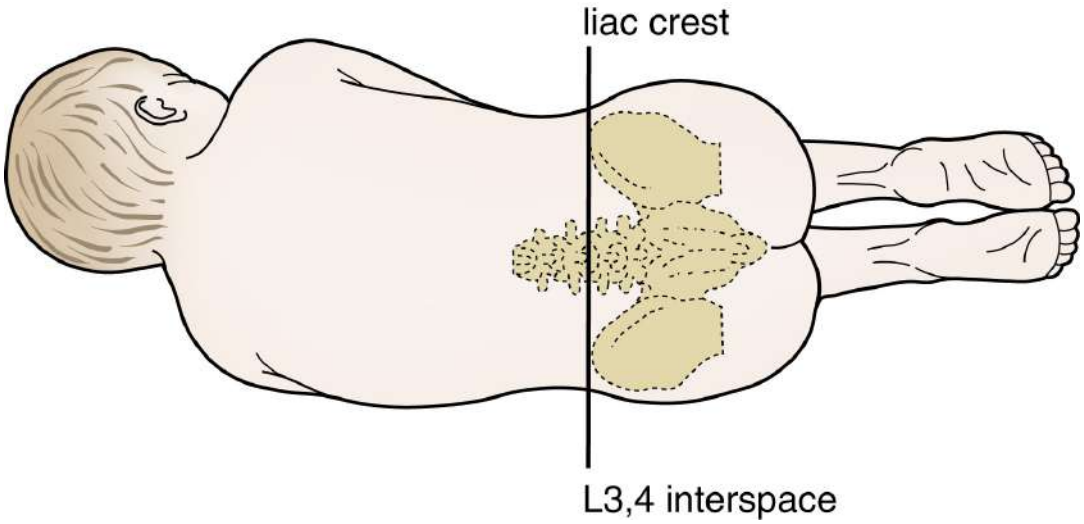

FIG. 24.13.1 Lateral decubitus position with landmarks.

Preparation and positioning

1. Assess and stabilise the ABCs as required. Establish secure vascular access, obtain blood cultures and appropriate blood tests, administer oxygen and begin cardiac monitoring and pulse oximetry.
2. Place the child in a lateral decubitus (Fig. 24.13.1) or sitting position (Fig. 24.13.2) at the edge of the bed.
3. If the child is conscious, consider sedation.
4. Landmarking is done by identifying the iliac crests and considering an

imaginary line connecting the top of the two crests. This line traverses the top of the L4 vertebra in most patients; the L4 spinous process should be palpable in the midline just below this level. The L4–L5 interspace lies below this and the L3–L4 interspace above. Care should be taken when considering the plane of the child's back and the needle entry should be in a sagittal plane in the midline (i.e. reference to the child and not to the plane of the bed).

5. Preference is made to use the L4–L5 interspace in the first instance (especially in small infants), L3–L4 interspace if unsuccessful. This differs from the preference in adults because the conus medullaris terminates near L3 at birth, but at L1–L2 by adulthood.
6. Don sterile gloves and prepare the area widely with chlorhexidine or povidone-iodine solution. If the child is conscious, infiltrate the entry site with 1% lignocaine and a 25-gauge needle, then the deeper tissue to the level of the paraspinal ligaments. Alternately, consider a topical anaesthetic preparation such as EMLA™ on the skin and infiltrate deeper tissues with 1% lignocaine.

Lateral decubitus position

1. Flex the child's knees and torso up towards the chest, but do not overflex the neck and compress the airway.
2. Have an assistant hold the child firmly in this decubitus position.

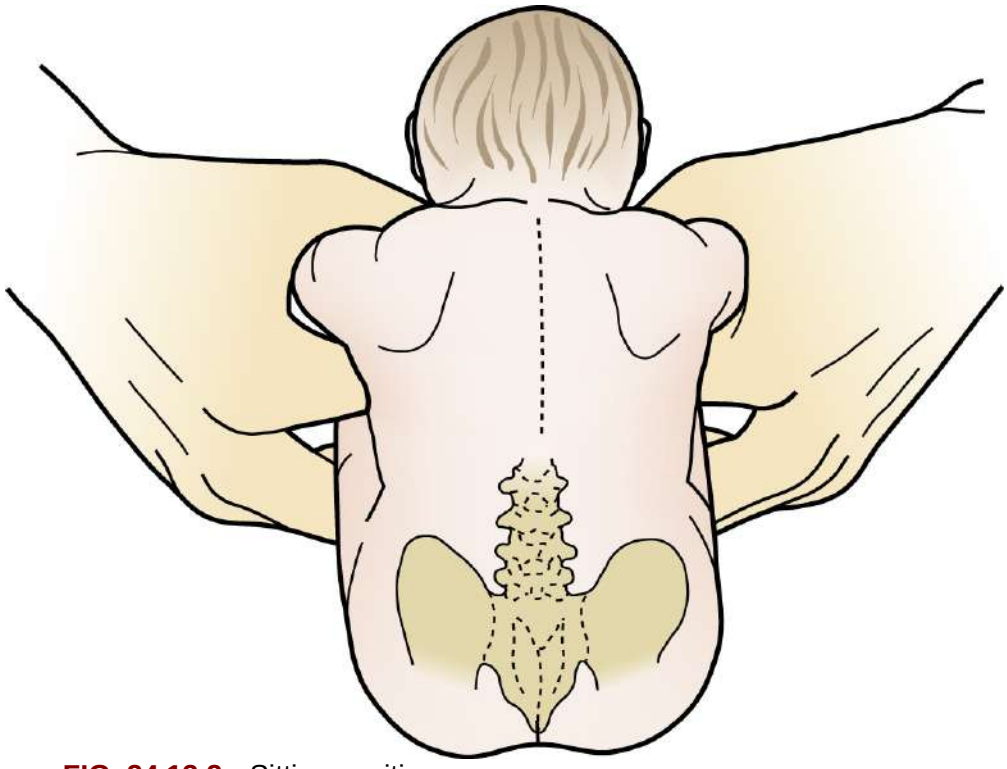

FIG. 24.13.2 Sitting position.

3. Remember that this can be quite frightening and uncomfortable for toddlers and older children.
4. The patient should lie left side down for right-handed operators.

Sitting position

1. Have an assistant hold the child upright with the hips flexed. Hold the child's right elbow and knee with the left hand and the left elbow and knee with the right hand.
2. Put the thighs against the abdomen and flex the trunk.
3. Keep the craniospinal axis perpendicular to the transverse plane of the line connecting the iliac crests.
4. This position may be more comfortable for older children.

Procedure

1. Drape the area.
2. Grasp the needle between the thumb and index finger of your dominant

hand; hold the needle with the bevel upwards

3. Aim the needle into the interspace at 60 degrees, just below the L4 spinous process and advance slowly until there is mild resistance at the paraspinal ligaments. Care should be taken that the needle remains in a sagittal plane with respect to the child, rather than considering the plane of the bed as the reference.

Table 24.13.1

Interpretation of cerebrospinal fluid

	PMN ($x10^6$ /L)	Lymphocytes ($x10^6$ /L)*	Protein (g/L)	Glucose (CSF:blood)	Gram stain	Opening pressure/*tip
Normal (>1 month age)	0	≤ 5	<0.4	>0.6 or ≥2.5 mmol/L	Negative	5–20
Normal term neonate	0	<20	<1.0	≥0.6 or ≥2.5 mmol/L	Negative	Not reliable
Bacterial meningitis	100–10000	Usually <100	>1.0	<0.5	Positive (80% cases)	Elevated
Partially treated meningitis	50–100	Usually <100	>1.0	<0.5 may be normal	Likely to be negative	Elevated/do polymerase chain reaction (PCR)
Viral meningitis	Usually <100	10–1000 may be normal	0.4–1.0 may be normal	>0.6 or ≥2.5 mmol/L (normal)	Negative	Normal or slightly elevated/do PCR
TB meningitis	Usually <100	50–1000 may be normal	-5.0 may be normal	<.4 may be normal	Negative	Elevated/do acid-fast bacillus (AFB)

* Note that some labs report polymorphonuclear leukocytes and monocytes; with monocytes predominating in viral, fungal, amoebic and partially treated bacterial meningitis.

4. Advance the needle slowly until there is a ‘pop’ as the needle enters through the dura into the subarachnoid space. This is felt as a subtle give in infants and younger children.
5. Remove the stylet.
6. If CSF does not appear at the hub, replace the stylet and advance further.
7. If bony resistance is met at any stage, do not force the needle, rather stop and reconsider landmarks and trajectory of the needle. Reposition as required.
8. Once CSF is yielded, proceed with measuring the opening pressure if indicated then collect specimens. Normal opening pressure for a child in the lateral decubitus position is 5–20 cm, but a struggling child may artificially elevate this number. Do not attempt to measure an opening pressure in a sitting child because it is unreliable. CSF collection is done by free drainage rather than aspiration.
9. Limit CSF withdrawal to 2–3 mL in neonates or 3–6 mL in infants and older children, aliquoted into 3–4 separate and sequential tubes, labelled accordingly.
10. Replace the stylet and remove the needle.
11. Dress the entry site and encourage the child to remain lying for a period of 1–4 hours if possible in order to minimise CSF leak.
12. Send CSF for cell count/differential, glucose, protein, Gram stain and

bacterial culture, and other pathogen studies (e.g. polymerase chain reaction [PCR], viral cultures, AFB or fungal cultures). Generally, the most accurate cell count is obtained on the final sample of CSF. If the CSF is bloody, ask for a cell count on both tubes #1 and #4. Clearing of blood suggests a traumatic tap, whereas no clearing suggests subarachnoid haemorrhage.

13. Interpret CSF findings (Table 24.13.1).

Complications

Local

- Puncture-site pain
- Backache
- Headache (less common <10 years of age)
- Traumatic LP (difficult to interpret) or unsuccessful LP (no yield of procedure) – common
- Vomiting
- Temporary paralysis
- Epidermoid tumours
- Discitis
- Epidural haematoma or abscess
- Epidural abscess
- Osteomyelitis.

Central nervous system/systemic

- Subarachnoid haemorrhage
- Subdural haemorrhage
- Brainstem herniation
- Cardiorespiratory arrest due to positioning.

Tips

- Good positioning and restraint of the infant or child greatly facilitates the procedure.

- If no CSF appears in the hub after initial penetration, rotate the needle 90 degrees and withdraw the stylet.
- Use PCR/ antigen tests for partially treated or Gram-stain negative CSF.
- If the child appears ill or if contraindications to LP exist, administer empiric antibiotics capable of crossing the blood–brain barrier after blood cultures are obtained. Proceed to LP only once the child is stable.
- Herpes encephalitis is a diagnostic consideration in young infants: consider with history of contact, elevation of monocyte count in CSF and increased RBC in non-bloody tap. Have high index of suspicion and treat with Aciclovir pending CSF viral culture and herpes PCR. These need to be requested specifically.
- If the LP is traumatic and CSF is bloody, a CSF WBC: RBC ratio can be calculated to assist in interpreting initial results:
    - The WBC:RBC ratio is calculated as a simple ratio on CSF fluid. A WBC:RBC ratio <0.001 (1:1000) makes bacterial infection less likely.
    - 'Observed: predicted' (O:P) CSF WBC ratios are considered in many centres; these are of low sensitivity and should not be used to rule out meningitis.
    - In the case of a traumatic tap, the safest option is to treat as meningitis and await CSF culture and/or PCR results.
- Reference ranges for CSF WBC counts vary in the literature, and acceptable values can be region-specific. The values in Table 24.9.1 are a conservative accepted estimate. Remember the following rules of thumb:
    - Presence of polymorphonuclear cells in the CSF is generally abnormal.
    - Lymphocyte count reference ranges can vary widely from up to 20 at birth down to zero by 1 year. Generally, if a clinical suspicion of meningitis exists, then treatment pending cultures is safe.
    - Protein level in the CSF is similarly variable, with high levels at birth (higher preterm), and falling rapidly thereafter. Protein level can also be raised by a traumatic tap.
    - All features of the CSF should be taken in context, together with clinical suspicion. No single component is diagnostic. Treatment with appropriate antibiotics per local and regional

guidelines, pending culture results is the safest option.

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24.14

Reduction of paediatric inguinal hernias

Damir Ljuhar, and Ramesh Nataraja

ESSENTIALS

1 The diagnosis of an inguinal hernia should be made prior to attempted reduction.

2 Ensure there is no evidence of obstruction, shock or peritonitis prior to reduction.

3 Understand and apply the anatomy of the inguinal canal to the technique.

4 Reduce by taxis using the two hand technique; one funnelling the hernia and the other pushing the hernia back towards the external inguinal ring.

5 Referral to paediatric surgeons for all incarcerated inguinal hernias is essential.

Introduction

Inguinal hernias commonly present as a lump in the groin or scrotum and are usually more evident when the child is crying or straining. As there are many causes of an inguinoscrotal swelling, with testicular torsion in boys being an important differential diagnosis, manual reduction should be attempted only once the diagnosis of an inguinal hernia has been confirmed. In children with an incarcerated inguinal hernia, reduction should be performed only if there is no evidence of obstruction, shock or peritonitis from gangrenous bowel. The aim of

early reduction is to prevent bowel necrosis, decrease the likelihood of testicular atrophy and stabilise the patient.

Preparation

The patient should be placed in an appropriately private setting with adequate time and staffing. The child should be kept warm and usually does not require full exposure. A cold child may be more unsettled, and tenderness may be more difficult to ascertain. As there may have already been previous attempts at reduction, the child should be given adequate analgesia and/or sucrose for procedural pain management. Intravenous analgesia can be given initially or if the first attempt at reduction is unsuccessful. Parents may be present for the procedure; however, they should be made aware that the child may become quite distressed during the procedure.

Procedure

The aim of the procedure is to guide the herniated intraabdominal content back through the external inguinal ring into the peritoneal cavity. This is achieved by guidance through the inguinal canal and back through the internal inguinal ring. In order to successfully do this, it is important to have an understanding of the anatomy of these structures. The external inguinal ring lies more medially and inferior in the inguinal region, while the internal inguinal ring is more lateral and superior, midway between the anterior superior iliac spine and the pubic tubercle (Fig. 24.14.1B).

Firstly, with the thumb and index finger of one hand, a funnel is created over the external inguinal ring that will help to guide the content back into the inguinal canal (Fig. 24.14.1C). Applying upward and lateral traction with this hand will also help keep the external and internal rings open. Using the thumb and index finger of the other hand, the reducing hand, gentle pressure is applied to the hernia, slowly pushing it back along the direction of the inguinal canal (Fig. 24.14.1D). The funnel created should help direct the content through the external inguinal ring and prevent it from spreading around the inguinal region. At this point, it is important to be directing the herniated content laterally into the external inguinal ring rather than applying direct pressure over the hernia, as this compresses the hernia posteriorly. Using a circular motion with the reducing hand as gentle pressure is being applied laterally can help the reduction.[1]

Similarly, the fingers of the reducing hand can be used to walk the bottom of the sac towards the external inguinal ring.

Successful reduction will be noticed with the hernia disappearing, a reduction of the groin swelling, and improvement in the clinical state of the child. Upon reduction, a finger should be placed over the internal inguinal ring to prevent the content from herniating out immediately (Fig. 24.14.1E).

In the event that the hernia is unable to be reduced, prompt referral to the paediatric surgical team should be made. If there is overlying skin changes or oedema and the hernia is unable to be reduced, then the hernia is considered incarcerated. If reduction is unsuccessful after approximately 5 minutes of continuous pressure, trying sedation (if not already used) and repeating the procedure is warranted. The two main factors responsible for being unable to reduce an incarcerated inguinal hernia are young age and duration of symptoms. In the event of an irreducible or strangulated inguinal hernia, urgent surgical reduction and herniotomy is indicated.

Inguinal hernias in girls

The incidence of inguinal hernias in girls is seen up to 10 times *less* than in boys.[2] In girls, ovaries are the most common to herniate into the sac, and sliding hernias are also known to occur where part of the bladder wall, fallopian tube or uterus has been found in the hernia.[3] This is in contrast to boys where the most commonly herniated content is small bowel. In case of tenderness, attempt at reduction should not be made in the emergency department as this suggests torsion of the ovary and urgent surgical intervention is required.[4] When there is no tenderness, reduction again is not recommended and referral to paediatric surgery is indicated. Small bowel can still herniate through the inguinal canal in girls and this will be noticed by a significantly larger swelling. This still requires reduction using the same method described above.

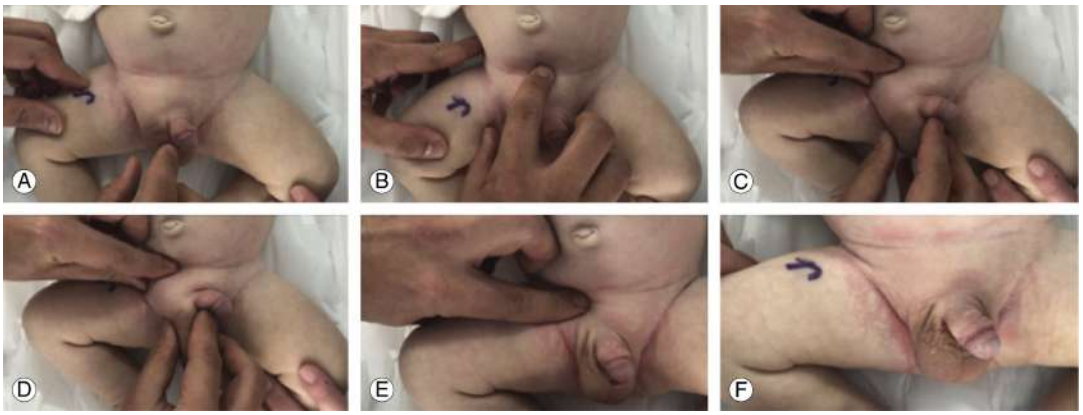

FIG. 24.14.1 Reduction of a paediatric inguinal hernia.

Complications

- Recurrence. In the patient with a reducible inguinal hernia, an outpatient referral to the paediatric surgical team should be made for prompt surgical repair. In the child with an incarcerated hernia that has been reduced, there is a 15% risk of re-incarceration within 5 days. [5] This warrants more urgent intervention; however, surgical repair is usually delayed by 1 or 2 days to allow resolution of the oedema caused by the incarcerated hernia.
- Incomplete reduction. An inguinal hernia not fully reduced past the internal inguinal ring may lead to one wall of the bowel still being incarcerated. If there is concern regarding the completeness of reduction, referral to the paediatric surgical team is recommended.
- Reduction of necrotic bowel. A rare but serious complication, avoided by not reducing an inguinal hernia in a child with symptoms and signs of shock or peritonitis.
- Testicular atrophy due to compromised vascular supply to the testis. Patients at the most risk of this are premature infants and infants younger than 3 months of age.[6]

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24.15

Paraphimosis

Annette Chang, and Ramesh Nataraja

ESSENTIALS

1. Recognition of paraphimosis.
2. Immediate treatment to prevent complications.
3. Adequate analgesia throughout the procedure is essential.

Introduction

Paraphimosis is a painful urological emergency condition which occurs when the foreskin is retracted behind the glans penis despite a constrictive distal preputial ring. This results in venous congestion of the glans. If untreated this condition will result in impaired arterial circulation and may cause necrosis of the glans penis. To prevent this complication from occurring; timely diagnosis and emergency reduction of the retracted foreskin is required. Various methods are documented in literature of which the common techniques are described in this chapter.

Indications

- Paraphimosis.

Contraindications

- Inadequate analgesia.

Procedures

The techniques described below may be employed to reduce paraphimosis, dependent on the individual health professional's experience and training. For all of the techniques described below, adequate procedural analgesia/sedation is essential. Modes of administration include topical application (lignocaine [lidocaine] gel 2%, EMLA cream), a penile block, and/or a systemic route (nitrous oxide, intranasal fentanyl). It is also important to ensure that the patient's privacy and modesty is maintained throughout the procedure.

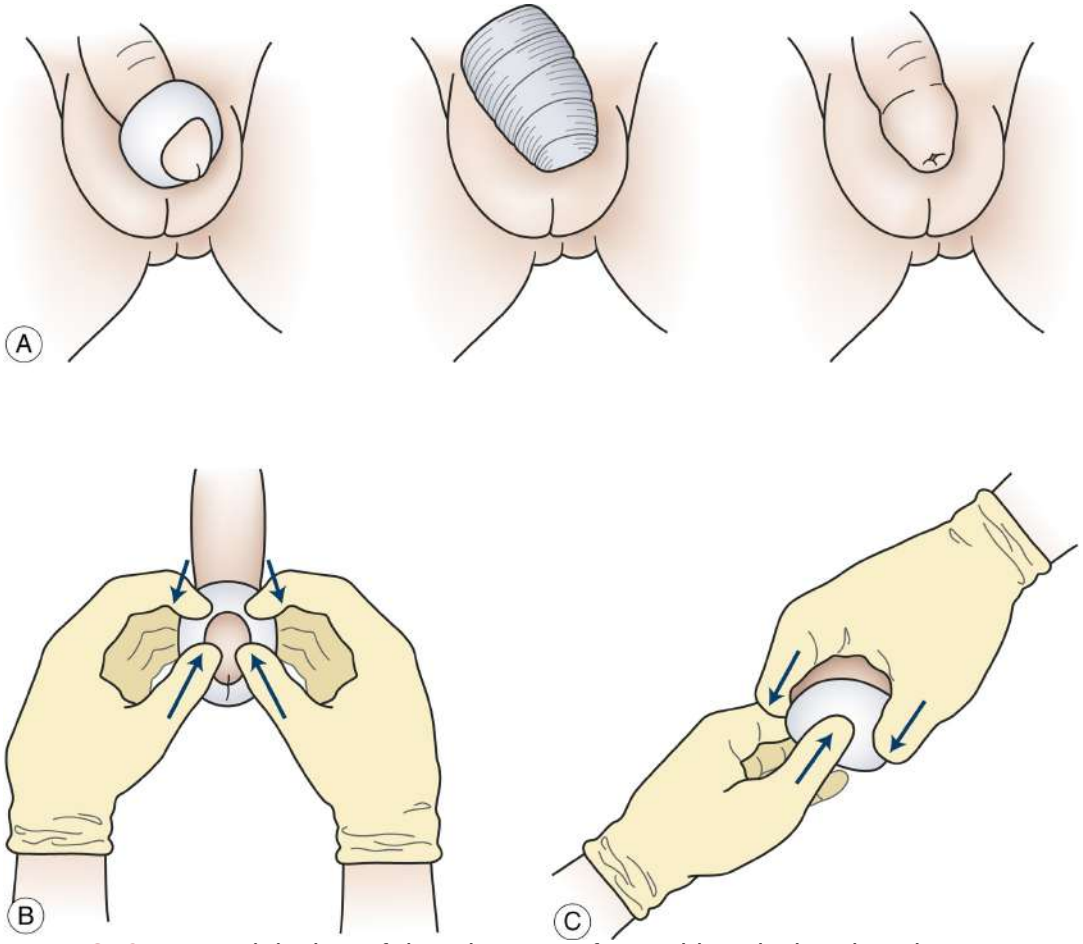

FIG. 24.15.1 (A) View of dorsal aspect of paraphimosis; bandage is wrapped around paraphimosis and along length of the penis; view of penis once the bandage is removed. (B) Manual reduction of paraphimosis: the thumb pads gently press the glans penis back through the narrowed foreskin opening, while the fingers pull the retracted foreskin distally over the glans penis. (C) Manual reduction of paraphimosis: compression and traction by one hand wrapped around the glans penis and paraphimosis. The thumb of the opposite hand is used to apply pressure to the glans

penis to enable reduction.

Non-invasive methods

1. Osmotic agents (e.g. granulated sugar, dextrose 50% solution) are usually the first-line treatment:
    a. The agent of choice is applied topically to glans penis and the paraphimosis and held in place by a cut gloved finger.
    b. This is left in place for 1–2 hours to allow for reduction of swelling and followed by gentle manual reduction of the paraphimosis.[1]
2. The use of ice has also been described in a similar fashion to reduce oedema of the glans penis to allow for manual reduction.[2]
3. The EMLA glove method further adds to these non-invasive techniques:
    a. The thumb of a glove is cut at the base into which a single tube of EMLA is placed (2.5% lidocaine and 2.5% prilocaine).
    b. This is placed over the paraphimosis and oedematous glans and left in-situ for 30 minutes, followed by manual reduction.[3]
4. All of these techniques, however, take a considerable time to be effective and a more active approach may be required.

Invasive methods

1. Manual compression is described as invasive and hence this requires adequate procedural analgesia/sedation. These methods include a bandage or hand compression of the paraphimosis (Fig. 24.15.1).
2. The compression technique was initially described by Ganti et al.:
    a. A flexible self-adhering bandage (2-inch) is wrapped around the penis commencing distally at the glans penis and advancing proximally.
    b. The duration of wrap is approximately 5–7 minutes.[4–6]
    c. This was modified by Pohlman et al. with their usage of a 1-inch CoFlex bandage.[5]

d. This may result in either auto-reduction of the paraphimosis or requires additional manual reduction.

3. Hand compression involves a two-staged procedure:
    a. Initial compression is applied circumferentially by wrapping the fingers and palm around the oedematous glans penis and paraphimosis.
    b. Once the swelling is reduced, a two-handed technique is used.
    c. Further compression is maintained by one hand wrapped around the glans penis and paraphimosis.
    d. This hand provides compression and traction simultaneously.
    e. The thumb of the opposite hand is used to apply pressure to the glans penis to enable reduction.
    f. An additional method is placing bilateral thumbs onto the glans penis and pulling the foreskin over the glans with the fingers.[5–7]

The Dundee technique

The Dundee technique should not be used in the paediatric population. It involves use of a 26G needle to perform multiple punctures in the oedematous prepuce. Firm pressure is applied to reduce oedema by expressing fluid. This is followed by manual reduction of the paraphimosis.[8] It may be utilised under a general anaesthetic or in an older adolescent if it is assessed that they will tolerate it.

Post-reduction management

Once the paraphimosis is successfully reduced, the patient may be discharged home after spontaneous passage of urine. Advice against retraction of the foreskin within a fortnight of the reduction procedure is advocated.[7]

Contraindications to attempted reduction

If the glans penis has a clinical concern of ischaemia or the paraphimosis is unable to be reduced, a surgical referral is required for further management of this condition. Previously, a dorsal slit procedure was employed which may

follow with a formal circumcision. This may still be necessary with the failure of non-operative techniques.

Complications

1. Failure to recognise paraphimosis
2. Incomplete reduction of paraphimosis
3. Vascular compromise to glans penis
4. Acute urinary retention.

Tips

1. Patience
2. Requires adequate procedural analgesia/sedation
3. Unsuccessful reduction will require surgical referral for reattempt at reduction (this may require general anaesthetic).

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24.16

Gastrostomies and other enteral feeding devices – trouble shooting in the emergency department

Rupert Hinds, and Simon Craig

ESSENTIALS

1 Fistula tracts close over quickly. When in doubt secure with the largest Foley catheter that will fit and seek expert advice.

2 If the initial percutaneous endoscopic gastrostomy (PEG) displaces in the first 4–6 weeks the tract may not be established and blind placement of a PEG should not be undertaken.

3 If the external disk is becoming hard to rotate or more protuberant, and/or feeds are getting harder to administer, consider a buried bumper.

Background

Increasing numbers of children in the community are being fed via percutaneous endoscopic gastrostomy (PEG) tubes. They may also be used in conjunction with jejunal extensions. PEG feeding is a safe and efficient[1] long-term feeding device in those who cannot maintain normal nutrition/hydration with oral intake, particularly those who are unable to feed safely or effectively.

PEG placement may be considered in children with:

- neurological disorders such as cerebral palsy

• chronic disorders with increased need for nutrition, e.g. chronic renal failure
• craniofacial abnormalities
• malabsorption states, such as cystic fibrosis, short bowel syndrome
• oncology patients.

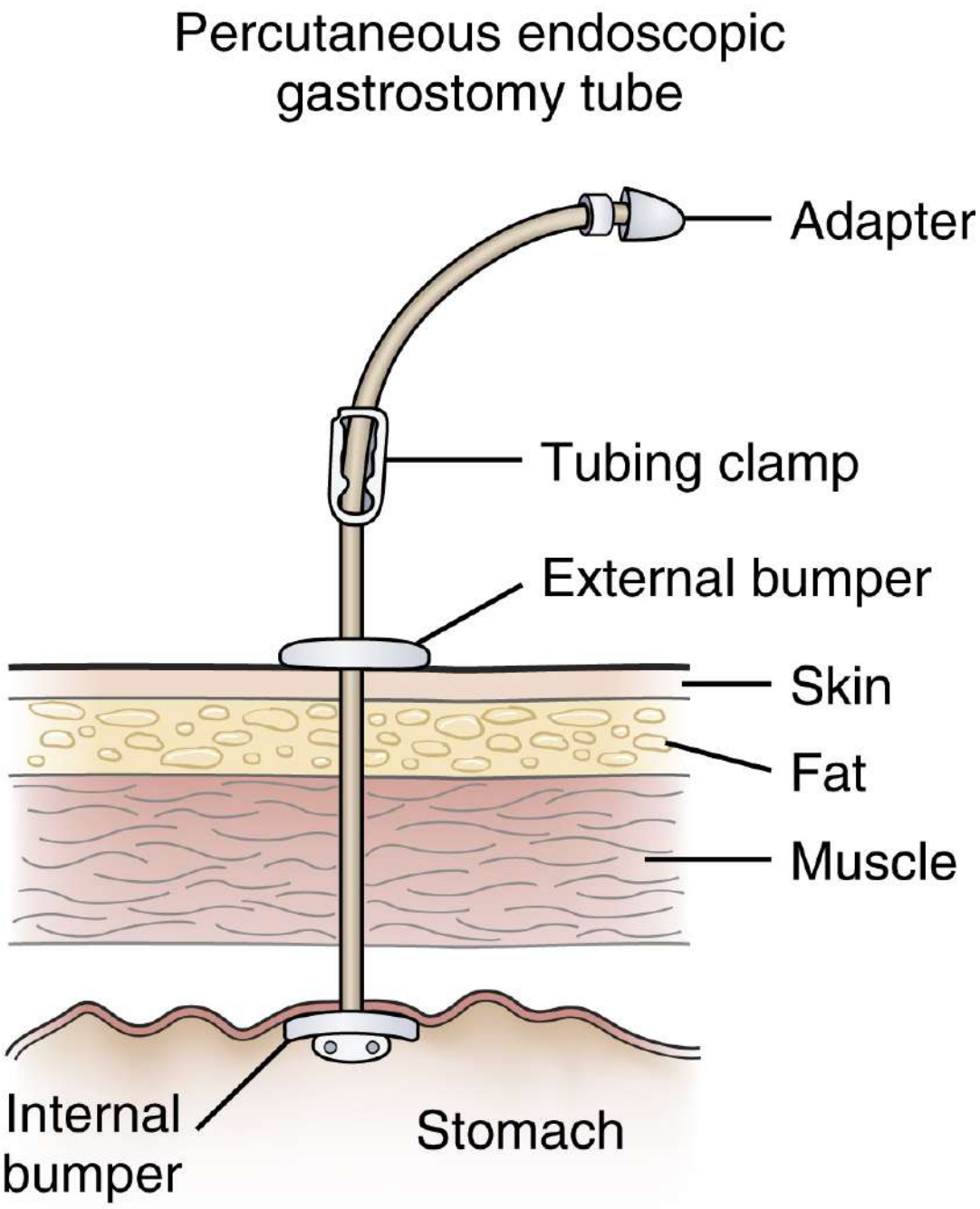

FIG. 24.16.1 Percutaneous endoscopic gastrostomy tube.

Anatomy of a percutaneous endoscopic gastrostomy tube

PEG tubes consist of a feeding tube placed into the stomach, anchored by an internal bumper or balloon, and held against the abdominal wall by an external bumper (Fig. 24.16.1). The external end of the tube connects to feeding tubing, which may be given as intermittent bolus feeds or, occasionally, continuously.

Percutaneous endoscopic gastrostomy insertion method

In most children requiring a gastrostomy, the procedure is carried out endoscopically, although some children may have had their device placed laparoscopically. Children are typically admitted post procedure to ensure tolerance of feeds, recovery from anaesthesia and good pain control.

The initial PEG device may be changed to a low profile device 3–4 months after placement.

Complications

Although a commonly used technique now for more than 20-years, complications are still described with a frequency of 0.5–17%.[2] Furthermore, a return to an emergency department within the first 30 days of procedure is not uncommon.[3] For practical purposes, complications may be usefully divided in to ‘early’, within the first 30-days, and late, outside this period, but typically in the first 2-years.[4]

Percutaneous endoscopic gastrostomy displacement

Gastrostomy tubes may become displaced and the most important management step will always be to ensure that fistula patency is maintained. Whilst it is not always the case, the fistula may close over within 4–6 hours if not secured, necessitating the placement of a new initial PEG if enteral feeding is still required.

Many parents may have been trained to replace a low profile balloon device and may have a spare. This can be safely replaced and used immediately with no confirmatory radiology required. Some low profile devices need endoscopic placement; when in doubt secure the fistula with the largest diameter Foley catheter that will fit and seek expert advice. Whilst not a long-term solution, a Foley catheter can be taped to the skin and used for feeding temporarily.

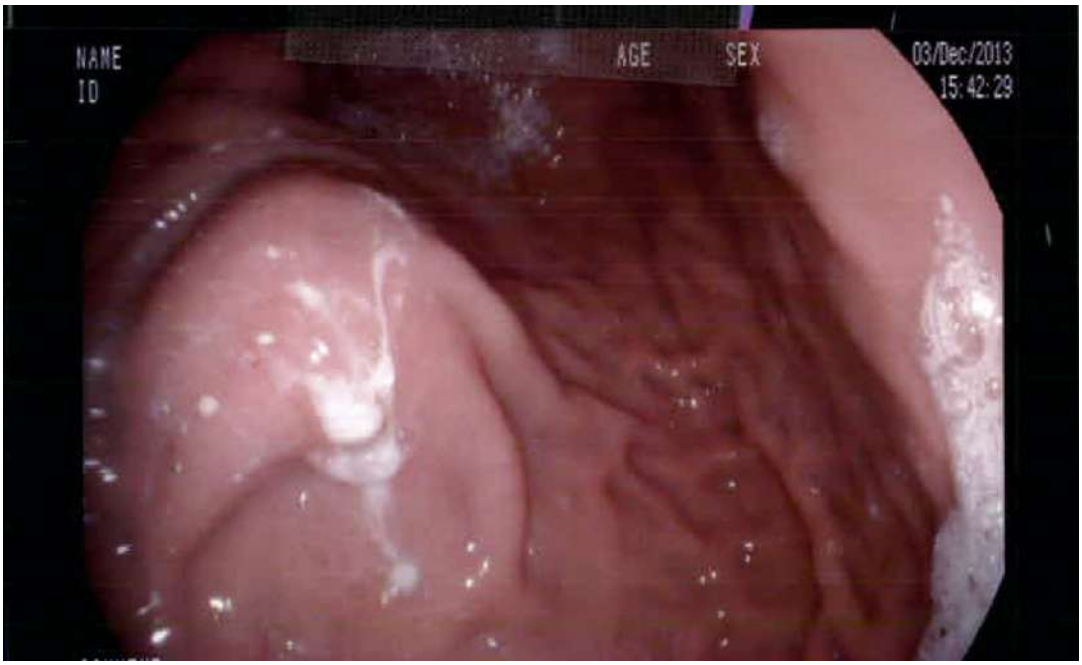

FIG. 24.16.2 Buried bumper with disappearance of percutaneous endoscopic gastrostomy from stomach seen at endoscopy.

If the initial PEG displaces in the first 4–6 weeks, the tract may not be established and blind placement of a PEG should not be undertaken. The tract should be secured with a Foley catheter and the gastrostomy will need to be placed either endoscopically or radiologically, as per local expertise. Liaison with the initial treating team is essential.

Buried bumper

A buried bumper (BB) describes the migration of the internal flange of the PEG into the anterior abdominal wall. The child's carer may have noted that the external disk is hard to rotate or more protuberant, or that feeds are getting harder to administer. If this complication is considered then prompt endoscopy

or contrast study should be organised to try and minimise the risk of peritonitis or abscess formation. If confirmed, endoscopic removal of the BB and placement of a new PEG is most commonly performed (Fig. 24.16.2).

Gastrocolocutaneous fistula

Gastrocolocutaneous fistulas (GCF) are a rare but serious complication, due to inadvertent puncture of the transverse colon during placement. Most commonly they present within the first few days of PEG insertion, but on occasion it can be months later.[4]

Children in general and particularly those with spinal anomalies are at highest risk of this complication. Clinical features of a GCF may include faecal discharge from the PEG, undigested food in the stool, resistance to feeding and lack of stool output. If this diagnosis is considered prompt referral to treating team for possible surgical management is vital. A CT scan may be needed to confirm the presence of a GCF, but discussion with the treating team first is appropriate.

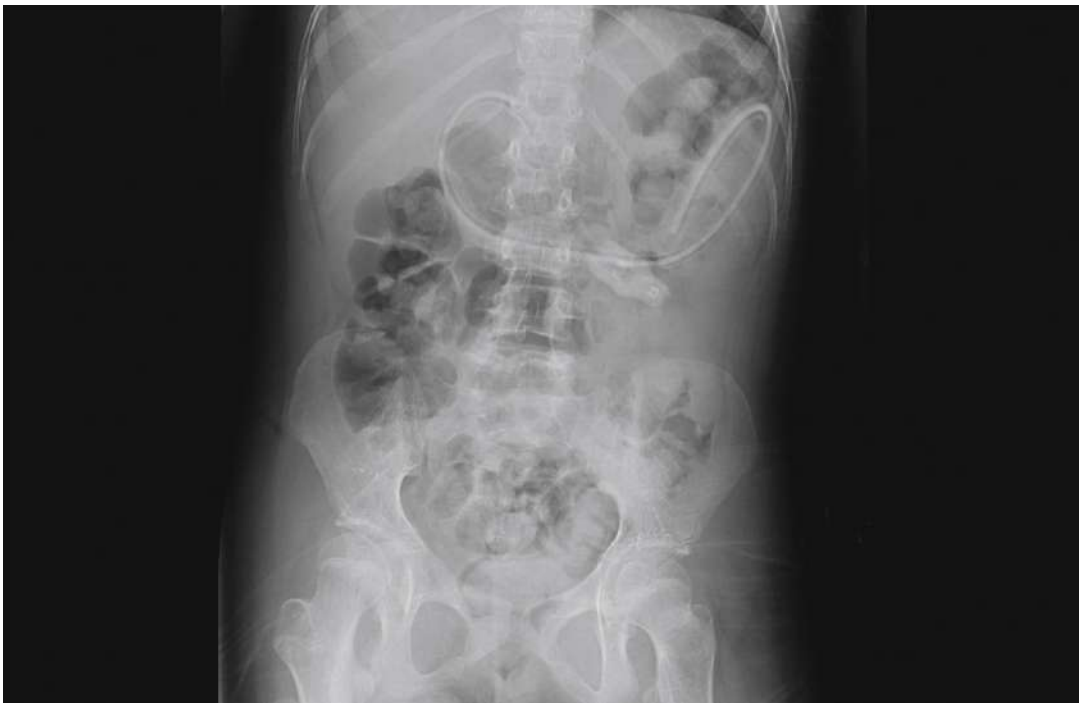

FIG. 24.16.3 Plain abdominal X-ray demonstrating appropriately placed percutaneous gastrostomy with jejunal extension.

Peristomal leak

A small amount of leak of clear fluid is common and does not require treatment. However, if there is significant leak of feeds preventing ascertainment of full calorie/fluid requirements or problematic erythema and burning around the gastrostomy, action should be taken.

There are two main causes, loosening of the device and delayed gastric emptying. Loose PEGs may require the external flange to be rotated and/or tightened. If possible, comparison of the distance to skin on the PEG tube with that at initial placement should be undertaken. Low profile ‘button’ devices may become loose if the child has lost weight.

Children with neurological problems may have delayed gastric emptying. Whilst a gastric emptying study may be considered, in most cases a trial of acid suppression with a proton pump inhibitor to reduce skin burning or a prokinetic such as domperidone or erythromycin should be trialled. Seek advice from the child’s usual paediatrician regarding choice of therapy.

A leak which worsens after feed administration may indicate an excessive rate of feeding. This can be assessed with the help of a dietician.

Peristomal infection

Cellulitis around the PEG site is most commonly seen early. Those children who are immunosuppressed or significantly malnourished are at the most risk. The use of prophylactic antibiotics at the time of placement has significantly reduced this complication.[5] Erythema around the site may represent leak of gastric contents rather than infection. If cellulitis is considered then antibiotics covering skin organisms should be commenced, such as flucloxacillin; the decision on whether this is oral or intravenous will depend on the degree of inflammation and the clinical state of the patient. If the child has presented with cellulitis within the first 2 weeks after insertion of an initial PEG, management should be discussed with the treating team.

Mechanical problems – blockage

Tube occlusion may occur at almost any time after PEG/PEJ placement. Frequency will depend on pattern/type of feeds and medications. Jejunal tubes are particularly at risk. Advice should be given to parents to thoroughly flush PEGs with water after feed and medications. Different institutions will have their

own protocols for obstructed feeding tubes. There is limited evidence to support these strategies; however, common techniques include the use of phosphate-containing soft drinks such as cola or pancreatic enzymes to unblock the tube. If these measures fail, the device will need to be changed.

Jejunal feeding devices

A small number of children will require trans-pyloric feeding. These will include those with delayed gastric emptying, recurrent aspiration in spite of gastrostomy feeds and children with severe gastro-oesophageal reflux not responsive to medical therapy and not suitable for fundoplication. Typically, these devices are an extension through an existing PEG or a purpose-built device (PEG-J). They are prone to blockage and displacement. They can only be replaced endoscopically or by an interventional radiologist under fluoroscopic guidance. If they have completely fallen out, then a Foley catheter can be placed to keep the tract open, pending definitive placement. If there is doubt about the position of the distal tip, consider a plain abdominal X-ray (Fig. 24.16.3).

Table 24.16.1

Early and late complications of percutaneous endoscopic gastrostomy placement

Early complications	Late complications
Aspiration (increased in neurological disorders)	Mechanical problems – blockage
Bleeding	Gastrocolocutaneous fistula (can be early)
Colonic/small bowel injury	Buried bumper syndrome (see Fig. 24.15.2)
Displacement of percutaneous endoscopic gastronomy (PEG)*	Displacement of PEG
Granulation tissue	Granulation tissue
Hepatic/splenic laceration	Persistent (>2 weeks) gastrocutaneous fistula after elective removal, with leak of gastric contents
Peristomal infection	Peristomal leak (may represent loose device or delayed gastric emptying)
Pneumoperitoneum (often seen in uncomplicated PEG placement but can also represent colonic trauma)	

* If the initial PEG displaces in the first 4–6 weeks the tract may not be established and blind placement of a PEG should **not** be undertaken.

Granulation tissue

More than half of all children with gastrostomies will develop granulation tissue and in about 20% it occurs recurrently. Typically, it first appears within the first month post gastrostomy insertion. Granulation tissue is a normal physiological response, and a key part of management is parental reassurance that some granulation tissue is normal. It will often improve with time.

Whilst there is a paucity of informative data, if the tissue bleeds excessively trials of topical steroids or silver nitrate can be considered. Liaison with stomal/PEG nurses where local expertise exists is very helpful.[6]

It is also important to try and prevent formation of excessive granulation tissue in the first place by ensuring that the PEG site stoma is kept dry and clean, and that movement causing friction is minimised. If a low profile (button) device is being used, make sure it has been measured and is well fitting.

Acknowledgements

Special thanks to Megan Murray and Jennifer McDonald, PEG outreach nurses at Monash Health for their input into this chapter and their day-to-day assistance with our patients.

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SECTION 25

Orthopaedics and Rheumatology

OUTLINE

25.1

Orthopaedics and rheumatology

Damien McKay, and Felix Regenfelder

ESSENTIALS

1 Painful dysfunction of limb, posture, or gait may be caused by trauma, infection or, less commonly, inflammatory or neoplastic developments.

2 Careful attention to history and close assessment of posture, joint range of motion and response to palpation are required to identify and accurately diagnose focal pathology.

3 Assessment of the entire musculoskeletal system and a generalised systemic examination is essential not only in children with generalized musculoskeletal pain but also in children with focal musculoskeletal pain without a history of trauma.

Introduction

The child who has bone and joint pathology may present in a variety of ways. Although the differential diagnosis of acute paediatric musculoskeletal pain and dysfunction is wide, the key diagnoses to consider are infection, inflammation, malignancy and trauma resulting in fracture. Fractures are in the top ten paediatric emergency department (ED) presentations in Australia.[1] The potential diagnosis differs depending on whether the presentation is that of focal musculoskeletal pain or generalised/multifocal pain. An awareness of the range of possible conditions and their clinical presentation, together with skilled physical examination and clinical reasoning, is essential for optimal outcomes.

The child with acute musculoskeletal pain or dysfunction

General approach

The spectrum of musculoskeletal pathology occurring in the paediatric and adolescent population is very different from that seen in the adult population.

Infant and child development has a marked influence on the musculoskeletal pathology seen and its manifestations, as well as the techniques used in assessment. Specific patterns of injury are also seen during adolescence. These age-specific differences in musculoskeletal pathology are outlined in Box 25.1.1.

Assessment

History

The most important characteristics of a child with musculoskeletal pain or dysfunction are the following:

- Age
- Trauma history
- Wellbeing
- Pain magnitude
- Pain localisation
- Temporal pattern of pain.

Age

The neonate or infant with focal pain or dysfunction should not be discharged without a specific diagnosis (most of which, with the specific exception of pulled elbow, warrant admission) and should generally be assessed and reevaluated by someone experienced in this area. Adolescents with a limp or knee or hip pain must have slipped upper femoral epiphysis (SUFE) specifically ruled out.

Trauma history

The history is less sensitive or specific than in adult medicine because:

- children are often poor historians.

- minor injuries are very frequent.
- there is always a possibility of abusive injury and inadequate or erroneous history.

A clear history of completely normal function prior to a specific event that precipitated crying and subsequent pain and dysfunction should prompt a search for a fracture. In older children a clear understanding of the mechanism of injury may help to guide examination and investigations.

Wellbeing

The following key points should be sought:

- History of fever/malaise
- Reduced appetite or activity level, which are usually abnormal in sepsis or malignant tumour disease
- Minor illnesses are also common and can co-exist with injury.

Pain magnitude

- Septic arthritis is painful to the slightest movement.
- Subacute arthritis and Perthes' disease may have varying levels of disability and pain.

Pain localisation

This is the foundation of acute musculoskeletal diagnosis and efficient use of investigations. A suggested sequence of examination for young children with musculoskeletal pain is shown in Box 25.1.2.

Temporal pattern of pain

Pain that is worse with activity or worse late in the day is suggestive of a mechanical aetiology. Pain, and more specifically joint stiffness, presents in the morning or after periods of inactivity are suggestive of an inflammatory aetiology. Disruption of night sleep is a red flag for malignancy or bone tumour.

Box 25.1.1 How children differ: impact of development on musculoskeletal pathology

Body proportions

- Large, heavy head: higher fulcrum for spinal disruption
- Low centre of gravity
- Relatively short legs.

Physiology of developing bone

- Zone of calcification: weakest point of muscle/tendon/ligamentous/bony continuum
- Increased plasticity means increased susceptibility to plastic deformation, e.g. torus, bowing fractures
- Physeal vulnerability predisposes to specific pathological responses to subacute or chronic microtrauma, e.g. avascular necrosis of ossification centres, slipped upper femoral epiphysis
- Physeal damage from fracture or ischaemia produces long-term complications
- High blood flow to physeal zones increases risk of vascular dissemination of bacterial disease, e.g. osteomyelitis.

Joints and ligaments

- Increased flexibility means less brittleness and fewer fractures in response to arcuate deformation, e.g. carpal, tarsal, rib and vertebral fractures less common
- Ligaments generally stronger than bones: sprains uncommon and complete ligamentous tears rare.

Exposure to mechanisms

- Infant-abuse mechanisms especially shearing/shaking forces
- Pre-school children fall on average four times a day (same-level falls)
- Highly active pre- and school-age children have frequent, relatively low-force impactions from falls (monkeybars, trees)
- Pedestrian versus car causes classical femoral or tibial/fibula injuries from

fender, plus head injury from secondary impact
- High-force MVA/MBA/high-energy sports injuries increase from adolescence.

Immunology

- Increased incidence of bacterial and viral infection
- More rapid dissemination of, and destruction from, bacterial infection.

Psychology

- Fear and pain make young children difficult to examine and increase importance of observation and gentle handling
- Immature intellectual development means poor verbalisation of symptoms, incomplete self-other differentiation, and other blocks to symptom communication
- Medical management of traumatic experiences, such as injuries, may influence future psychological responses to trauma
- Altered body perception increases susceptibility of adolescents (particularly females) to unconsciously exaggerated dysfunction in response to minor injuries.

Healing

- Rapid healing of most fractures (e.g. femoral fractures: infants 3 weeks!)
- Enormous remodelling potential of deformation within arc of use
- Great ability to compensate, physically and psychologically, for loss of function
- Minimal stiffness after immobilisation.

MBA, motorbike accident; MVA, motor vehicle accident.

Other useful historical features include:

- aggravating factors
- history of previous musculoskeletal pain or dysfunction

- level of participation in sport and physical activity
- bullying or school/home problems
- presence of associated symptoms including:
    1. fever, anorexia, weight loss, intermittent night pain (neoplastic disease)
    2. back pain (discitis, vertebral osteomyelitis)
    3. abnormal bleeding (leukaemia, haematological disorders)
    4. morning stiffness (inflammatory causes)
    5. migratory polyarthralgia, preceding pharyngitis (acute rheumatic fever and post streptococcal reactive arthritis)
    6. preceding viral illness, diarrhoeal illness (reactive arthritis)
    7. associated abdominal pain (psoas abscess, acute abdomen)
    8. insect bite (Lyme disease), travelling to/migrated from an area endemic to Lyme disease.

Examination

- Gait if lower limb pathology
- Posture and symmetry of affected region
- Activity level
- General wellbeing and clinical observations (fever, heart rate).
- Regional musculoskeletal examination including range of motion, palpation
- Skin integrity
- Abdominal exam if indicated, including organomegaly
- Lymphadenopathy
- Focal pathology, e.g. warts, bites and callosities.

Start the examination in the most comfortable position for the child, e.g. on the lap of one of the parents, and begin with an unaffected extremity. Take note of the resting position of the affected joint or joints and any asymmetry, redness, swelling or wasting compared to the contralateral side. The affected joint or joints should be compared in symmetrical posture as position influences external appearance. Isolate joints (e.g. knee) when assessing range of motion so that incidental movement of another joint (e.g. hip) does not cause misleading results.

Examination of the entire musculoskeletal system may be necessary to

exclude abnormalities in other asymptomatic joints. Paediatric Gait Arms Legs and Spine (pGALS) is a simple, quick, standardised, structured assessment tool to distinguish abnormal from normal joint in children and young people with suspected musculoskeletal disorders.[2]

At the conclusion of the history and examination the emergency physician should have established the overall wellbeing of the child, the site and severity of musculoskeletal pain, practical precipitants, and the likelihood of traumatic, infective, or other processes. In patients with minimal physical findings, or findings out of keeping with other aspects of history and examination, the environmental and psychological context of the pain and dysfunction should be explored further. Clues to possible neoplastic illness are discussed later in this chapter.

Investigations

Pathology

Full blood count (FBC), C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and blood culture are indicated in a child with a painful, swollen joint with limited range or in a child with fever and refusal to weight bear.

FBC should also be performed when considering malignancy. In any child with an atraumatic painful or swollen joint the presence of anaemia, leucopoenia or thrombocytopenia even when single cell lines are involved warrants further consideration of malignancy as a potential diagnosis.

Radiology

Plain radiographs

X-rays should be performed if clinical examination suggests localised pathology but are less useful in children under 10 years of age with acute-onset limp (<1 week) without a specific history of trauma due to their lower sensitivity in diagnosing osteoarticular infections. Although plain radiographs need to be performed in all non-weight bearing children, children with localised pathology especially with history of trauma, in all adolescents irrespective of trauma history and persistent unexplained limb pain, initial plain radiological evaluation may not readily identify some diagnoses such as stress fractures, toddler’s fracture, Perthes’ disease and osteomyelitis.

Box 25.1.2 Tips for assessing young children

- The foundation of acute musculoskeletal diagnosis is precise localisation of pain and dysfunction.
- The foundation of a productive paediatric examination is a relaxed, non-fearful child.
- Personality and parental factors aside, a gentle, slow-moving, highly observant examiner whom the child senses has the trust of the parent, will be most successful in gaining meaningful information:

1. Be friendly and establish rapport with the parent.
2. Ask parents for clues about site of pain, e.g. is it worse during nappy changes or being picked up under arm, crying associated with going over bumps in car ride, etc. If the child is old enough, ask him/her to tell or show you exactly where it hurts.
3. Where possible, observe child at free play before attempting palpation.
4. Visually scan child for alterations of posture, symmetry or function.
5. Record any other clues, such as temperature, dysmorphic features, bruising and psychological state.
6. Examine the infant or child in parent's arms (under 6 months and over 5 years they may often be assessed on the couch without concern).
7. Introduce a washable or disposable toy, such as light, name-badge or 'funny face'.
8. Make your first touch very gentle and at a site distant from the target limb! Keep some touch continuous so the rhythm is reassuring. Verbal soothing, such as a hum, may settle anxiety.
9. Palpate the limb in question from one end to the other, watching the child's face continuously.
10. The first sign of discomfort may be an eyebrow flicker (i.e. a pre-frown) or a flinch.
11. If discomfort is sensed, move touch to another area immediately and slowly move back in even more gently for confirmation and further information.
12. Put all joints through their full range of movement unless

discomfort is noted.
13. Examine the spine, groin and abdomen of all lower limb or gait-disturbed children.
14. Areas of focal tenderness or limited range of motion can be further assessed by radiology or ultrasound.

Radiological findings in osteomyelitis are usually not present for up to 10 days from the onset of illness, at which time there may be periosteal elevation outlined by new bone formation and/or lucent areas.[4] Plain radiographs may show initial deep soft tissue swelling in the metaphyseal region and loss of fat planes followed by periosteal reactions and then lytic lesion ‘rat bite’ in the bone after 2–3 weeks in a patient with osteomyelitis.[5] Plain radiographs in patients with transient synovitis are typically normal but may show medial joint space widening. They are, however, useful in excluding other conditions like fractures and SUFE.

Due to the higher burden of pelvic or gonadal irradiation, non-selective or routine pelvic/lower limb radiology should not be encouraged. In spite of negative X-ray findings, always consider other investigative modalities if clinical suspicion persists. An effusion is better diagnosed by ultrasound and deep soft tissue infection, such as osteomyelitis, by MRI.

Ultrasound

The primary role of ultrasound in the investigation of acute musculoskeletal pain is in assisting with the identification of joint effusion. It is particularly useful for specific joints such as the hip where ultrasound is a sensitive, non-invasive assessment tool for evaluation of the irritable hip, its value in this setting is discussed in Chapter 25.2. Ultrasound is of limited benefit in the assessment of other joints over and above careful and structured clinical examination.

Bone scintigraphy

Isotope bone scans with three-phase technetium-99m MDP are sensitive early and well tolerated by children but lack specificity. Confusion may arise particularly in relation to physeal sites, where uptake is already above baseline. ‘Hot’ scans indicate increased osteoblastic uptake and generally relate to a response to infection or injury. A ‘cold’ scan suggests infarction, which in severe cases may be a consequence of osteomyelitis. Although the results may be

challenging to interpret in children because of normal uptake into the growth plate, the ability to image the entire skeleton can be useful for screening patients in whom precise localisation of the site of pain is proving to be difficult. Lower cost, lower risk of radiation and lower rate of needing sedation in younger children may be some of the advantages of this test. Bone scan is of particular value in children in whom multifocal disease is suspected, e.g. neonatal osteomyelitis or chronic recurrent multifocal osteomyelitis (CRMO).[4]

Magnetic resonance imaging

Lack of radiation, superb soft tissue and bony pathology, multiplanar capability and large field-of-view imaging make MRI an excellent mode of imaging for musculoskeletal pathology. Limitations include cost, access limitations, and the need to remain still for a longer time period, necessitating general anaesthetic in children under 5–7 years of age.

MRI is the investigative modality of choice to delineate the site of possible bacterial infection in a child with localised musculoskeletal pain and a septic picture in whom septic arthritis has been ruled out by clinical assessment, ultrasound or arthrocentesis.[6] MRI may also play a role in confirming diagnosis in osteomyelitis, delineating the extent of malignancies, identifying subtle stress fractures and in early recognition of avascular necrosis of the femoral head.[7] It is a useful mode of imaging in identifying the site of possible bacterial infection in a systemically unwell child with localised symptoms in whom septic arthritis has been ruled out by definitive investigations.[8]

Generalised or multifocal bone/joint pain

Introduction

The child presenting with generalised or multifocal acute musculoskeletal pain and dysfunction represents a different diagnostic spectrum. This is outlined in Box 25.1.3. As might be expected, infective and inflammatory disorders predominate and the ‘risk profile’ relates to the possible consequences of the generalised disease process, such as rheumatic fever, nephritis in association with Henoch–Schönlein purpura (HSP), leukaemia or juvenile idiopathic arthritis.

In assessing the child with multifocal joint or bone pain, or with arthralgia or arthritis in a setting of a febrile illness, the following features should be

explored:

History

- Antigen exposure – drugs, infections, insects and animals
- Previous infection or autoimmune dysfunction
- Features suggestive of infection – fever and its pattern and duration, other symptoms
- Features suggestive of metabolic drain – lethargy, weight loss, anorexia and night sweats.

Examination

- Skin and mucosae
- Eyes (visual acuity mandatory)
- Bones (including spine), joints and surrounding soft tissues
- Lymph nodes
- Liver and spleen
- Heart and lungs
- Kidneys and urine.

Investigations

- Urinalysis and microscopy for casts
- Inflammatory markers – FBC and film, CRP and ESR

Box 25.1.3 Causes of acute multifocal limb or joint pain in children presenting to the emergency department

Infection

- Acute viral illness*
- Streptococcal disease

- Other bacterial illness, e.g. *Neisseria*
- Kawasaki disease.

Post-infectious (immune-mediated)

- Serum sickness*
- Reactive arthritis*
- Rheumatic fever.

Inflammatory/vasculitic

- Henoch–Schönlein purpura*
- Juvenile idiopathic arthritis (JIA)
- Associated with SLE/IBD/other systemic inflammatory disorders.

Neoplastic

- Leukaemia*
- Neuroblastoma.

Haematological

- Sickle cell
- Haemophilia.

IBD, inflammatory bowel disease; SLE, systemic lupus erythematosus.

* Common causes.

Box 25.1.4 Features suggestive of malignancy in infants/children presenting with bone/joint pain or limp

- Non-weight-bearing or refusal to walk
- Night-time pain, waking from sleep
- Non-articular bone pain or tenderness
- Back pain
- Abnormal bone swelling
- Systemic illness (fever, rash, weight loss, anorexia, night sweats)
- Bruising
- Abdominal mass or organomegaly
- Abnormal neurology
- Low Hb, WCC, or platelet count
- High ESR or CRP (out of proportion to joint findings).

CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; Hb, haemoglobin; WCC, white cell count.

- Serology, for recent infection due to, e.g. streptococcus (antistreptolysin O test (ASOT), antiDNAse), Epstein–Barr virus (EBV), cytomegalovirus (CMV), parvovirus, mycoplasma, Barmah Forest virus, Ross River fever virus and *Yersinia*
- Rheumatological investigations – antinuclear antibodies, rheumatoid factor and HLA-B27 antigen.

Specific conditions

Juvenile idiopathic arthritis

Juvenile idiopathic arthritis (JIA) is a chronic, autoimmune, inflammatory joint disease. The spectrum of the clinical presentation of this condition is outlined in Table 25.1.1. Although the course of these disorders can be chronic, the onset of arthritis is at times rapid. Presentation with pain is variable, and patients may present to their primary care services or ED. Associated symptoms may include reduced function (e.g. unwillingness to play) or gait abnormalities. Inflammatory symptoms (such as morning stiffness) may be more pronounced after a period of inactivity. Multiple joint involvement usually suggests inflammatory arthropathy; however, the most common subtype of JIA is oligoarthritis, with the

knee as the most frequent presenting joint.[8] For appropriate risk management, septic arthritis should be considered and if necessary excluded in children presenting with single joint involvement. High fevers, refusal to allow examination of the joint, as well as raised leucocyte count or pronounced elevation of inflammatory markers, may suggest septic arthritis.

Presence of systemic features such as persistent fevers (more than 2 weeks), especially with quotidian pattern (regular spikes of temperature at a predictable time), evanescent rash, lymphadenopathy and organomegaly, with or without arthritis, should alert the attending clinician to the possibility of systemic arthritis. Serositis may manifest as pleural or pericardial effusion, requiring urgent treatment as cardiovascular emergency. Abdominal pain may be the result of peritoneal irritation by the intraabdominal inflammatory fluid, or non-specific vasculitis affecting abdominal viscera. Arthritis or arthralgia may be minimal or absent in the early stages of disease.[10]

Depending on the duration of fever at the time of ED presentation, and the clinical features present, differential diagnosis for these children may include acute viral or bacterial infection, Kawasaki disease and other vasculitides, malignancy and other autoimmune disorders such as systemic lupus erythematosus. Investigations must include blood culture, full blood examination and inflammatory markers, viral serology, and coagulation studies. Investigations as appropriate to the clinical findings and differential diagnoses, e.g. chest X-ray, electrocardiogram (ECG), echocardiogram, and abdominal ultrasound may be warranted. Anaemia of chronic disease is usual, with elevated inflammatory markers and thrombocytosis, which may exceed 10^6 per cubic millimetre.[11] Liver enzymes may be elevated, whilst coagulation abnormalities may be present and correlate with disease activity.[12]

In order to optimise the outcomes of children with JIA, contact with a paediatric rheumatologist must be made. Non-steroidal anti-inflammatory drugs (NSAIDs) may be used to manage symptoms until specialist review; however, disease-modifying drugs such as systemic steroids, methotrexate and intra-articular corticosteroid joint injections may be required.

Macrophage activation syndrome (haemophagocytic lymphohistiocytosis)

This rare disorder may present as a complication of systemic arthritis and include coagulopathy and disseminated intravascular coagulation,

encephalopathy, with liver and multiorgan failure.

Laboratory investigations reveal hepatic dysfunction, consumption coagulopathy with hypofibrinogenaemia, lowered haematological indices without classical leukaemic features, and a rapidly falling ESR usually inappropriate to the level of severity of the patient's condition. High-dose steroids, cyclosporin and other cytotoxics, with high-dependency support in an intensive care unit can be life-saving.[13]

Urticaria and serum sickness

The young child presenting with low-grade fever and urticarial rash may also have arthralgia or arthritis. Toddlers and preschool children may present with dramatic skin signs of urticaria, with migratory wheals with or without target lesions, which are the hallmark of erythema multiforme. Low-grade temperature may be present and there may be significant soft tissue swelling and arthralgias. Whilst up to 50% of cases may be idiopathic, the most common single trigger identified with this symptom complex in Australia is cephalosporin use.[14] Atypical features such as mucosal lesions, inability to weight-bear, high fevers and 'sick' appearance, haematuria or organomegaly require more detailed investigations and exclusion of more serious aetiologies. If symptoms are mild, this condition may be managed expectantly with removal of any unnecessary medication and rest. Antihistamines and NSAIDs may be useful for itch and joint pains.

Henoch–Schönlein purpura

This leucocytoclastic vasculitic disorder of uncertain aetiology is among the more frequent diagnoses presenting as joint pain and rash in the paediatric population. Characteristically there are four potentially affected systems:

1. Skin – extensor surface petechiae and maculopapules in lower limbs and buttocks
2. Joints – acral arthritis, but more commonly arthralgia and associated limb swelling
3. Kidney – micro- or macroscopic haematuria, with or without proteinuria, may be present or there may be frank nephritis
4. Gastrointestinal system – abdominal pain with submucosal vasculitis

presenting as intussusception, per rectum bleed or melaena, or peritoneal irritation.

Table 25.1.1

Juvenile idiopathic arthritis

	Systemic onset	Polyarticular		Oligo-articular	Psoriatic arthritis	Enthesitis-related arthritis
		RF Negative	RF Positive			
% of JIA	10–20	20–30	5–10	50–60	5–15	15
Gender (F:M)	1:1	9:1	4:1	4:1	3:2	1:9
Usual onset age	Any	2–12 (peak 2–3)	Adolescence	2–12 (peak 2–3)	Mid childhood	Adolescence
Joint pattern	Symmetric Multiple Small and large joints	Often asymmetric Multiple Small andlarge joints	Symmetric Multiple Small andlarge joints	Asymmetric Large joints (knee, ankle, wrist, elbow)	Asymmetric Small and large joints including hips and especially dips	Asymmetric Large joints Axial Skeleton
Extra-articular Involvement	Fever Rash Lymphadenopathy Hepatomegaly Splenomegaly Serositis	Painless uveitis(especially if ANA positive)	Rheumatoid nodules	Uveitis in 20%	Psoriasis Nail pitting Dactylitis Uveitis 10% Enthesitis	Symptomatic uveitis 20% Enthesitis IBD Aortitis

ANA, antinuclear antibodies; IBD, inflammatory bowel disease; JIA, juvenile idiopathic arthritis.

Table 25.1.2

Diagnosing acute rheumatic fever in Australia[16]

	High-risk groups*	All other groups
Initial episode of ARF	Two major or one major and two minor manifestations plus evidence of a preceding GAS infection †	
Recurrent attack of ARF in a patient with known past ARF or RHD	Two major or one major and two minor or three minor manifestations plus evidence of a preceding GAS infection †	
Major manifestations	Carditis (including subclinical evidence of rheumatic valve disease on echocardiogram) Polyarthritis or aseptic monoarthritis or polyarthralgia‡ Chorea¥ Erythema marginatum§ Subcutaneous nodules	Carditis (excluding subclinical evidence of rheumatic valve disease on echocardiogram) Polyarthritis‡ Chorea¥ Erythema marginatum§ Subcutaneous nodules
Minor manifestations	Fever# ESR ≥30 mm/h or CRP ≥30 mg/L Prolonged P–R interval on ECG**	Fever# Polyarthralgia or aseptic mono-arthritis‡ ESR ≥30 mm/h or CRP ≥30 mg/L Prolonged P–R interval on ECG**

All categories assume that other more likely diagnoses have been excluded. Please see text for details about specific manifestations. CRP, C-reactive protein; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate; GAS, group A streptococcus.

* High-risk groups are those living in communities with high rates of ARF (incidence >30 per 100,000 per year in 5–14-year-olds) or RHD (all-age prevalence >2 per 1000). Aboriginal and Torres Strait Islander Australians living in rural or remote settings are known to be at high risk. Data are not available for other populations, but Aboriginal and Torres Strait Islander Australians living in urban settings, Maori and Pacific Islander people, and potentially immigrants from developing countries may also be at high risk.

† Elevated or rising anti-streptolysin O or other streptococcal antibody, or a positive throat culture or rapid antigen test for GAS.

‡ A definite history of arthritis is sufficient to satisfy this manifestation. Other causes of arthritis/arthralgia should be carefully excluded, particularly in the case of monoarthritis (e.g. septic arthritis, including disseminated gonococcal infection), infective or reactive arthritis (e.g. Ross River virus, Barmah Forest virus, influenza, rubella, mycoplasma, cytomegalovirus, Epstein–Barr virus, parvovirus, hepatitis and Yersinia), and auto-immune arthropathy (e.g. juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis,

sarcoidosis). Note that if polyarthritis is present as a major manifestation, polyarthralgia or aseptic monoarthritis cannot be considered an additional minor manifestation in the same person.

[¥] Rheumatic (Sydenham's) chorea does not require other manifestations or evidence of preceding GAS infection, provided other causes of chorea are excluded.

[§] Erythema marginatum is a distinctive rash (see text). Care should be taken not to label other rashes, particularly non-specific viral exanthemas, as erythema marginatum.

[ǂ] Oral, tympanic or rectal temperature ≥38°C on admission or documented during the current illness.

[Θ] Note that, if carditis is present as a major manifestation, prolonged P–R interval cannot be considered an additional minor manifestation in the same person.

Risk management involves ruling out other causes of petechial illness, including leukaemia, idiopathic thrombocytopenic purpura and partially treated meningococcal illness, and watching for gastrointestinal and renal complications. Children must be followed-up with urinalysis for 6 months for delayed renal complications.[15]

Rheumatic fever

Rheumatic fever has become rare amongst Caucasians in Australia and New Zealand but still occurs and causes preventable morbidity and mortality in the Aboriginal and Pacific Islander populations of these countries, particularly in northern and central Australia and the Maori and Pacific Islander population of certain regions of New Zealand.[16] The incidence in indigenous children aged between 5 and 14 years in northern Australia is estimated to be 250–350/1000 children.[16] Infected skin lesions from scabies are more often a source of the *Streptococcus* than throat carriage in this group. Because of the disparity of likelihood in different populations, the Australian Heart Foundation recommends different diagnostic criteria in high- and low-risk Australian sub-populations. These revised, bi-level Modified Jones criteria are shown in Table 25.1.2.

Rheumatic fever in indigenous Australian children classically presents with an extremely painful polyarthritis, particularly affecting knees or ankles. Pain out of proportion to clinical effusion is the rule. The arthritis is slowly migratory but affected joints often overlap in time. Treatment with NSAIDs and/or aspirin (to which the joints in rheumatic arthritis are acutely sensitive) reduces the clinically apparent number of joints involved.

Rheumatic fever classically occurs up to a month after a skin infection, whereas other immune-mediated post-streptococcal disorders (reactive arthritis and glomerulonephritis) tend to occur earlier, e.g. 2 weeks post infection.

Although rheumatic fever and rheumatic heart disease are discussed in more detail in another section (see Chapter 5.7), it is critically important to consider rheumatic fever in any indigenous or Pacific Islander child presenting with acute arthralgias and fever, in view of the serious sequelae and preventable nature of this disease.

Post-streptococcal and other post-infective immune-mediated reactive arthritides

Post-streptococcal and other post-infective reactive arthritides (as distinct from the rheumatic fever complex) need to be considered in the differential diagnosis of any young child presenting with rash and joint swelling. These conditions may occur after viral or bacterial infections, alone or in immunogenic combination with antibiotics. These conditions have generally a favourable joint outcome, although occasionally other system manifestations may be observed.

Children may also have other clinical manifestations, including cutaneous vasculitis (e.g. erythema nodosum) or uveitis. The reactive arthritis pattern of post-streptococcal arthritis is more fixed than the migratory arthritis of classical rheumatic fever; tenosynovitis is often present; and there are no cardiac manifestations. Post-streptococcal arthritis also lacks the characteristic aspirin sensitivity of rheumatic fever.[17] However, the association mandates investigation of these children with FBC, ASOT and antiDNAse, ECG, and referral for follow-up by a paediatrician or paediatric rheumatologist. Other organisms implicated in immune-mediated arthritis include EBV, CMV, parvovirus, mycoplasma, and various gastrointestinal pathogens.[18]

The triad of urethritis, arthritis and conjunctivitis associated with *Chlamydia* infection may present as reactive arthritis in the sexually active adolescent, whilst other arthritogenic bacteria (e.g. *Salmonella* or *Shigella*) may cause arthritis associated with gastroenteritis. In each case, an attempt must be made to identify the causative organism and, if indicated, antimicrobial treatment implemented.

Neoplastic presentations

Bone or joint pain may be among the presenting symptoms in children with leukaemia, neuroblastoma and other bone-marrow-related malignancies. This pain is generally more severe and unremitting, and the debility more extreme,

than in children with juvenile idiopathic arthritis. Blood films must always be carefully analysed for the three cell lines (red cells, white cells and platelets). Minor degrees of anaemia, leucopenia, or thrombocytopaenia are common early features: these must be flagged and followed closely.[19,20] ESR is traditionally elevated out of proportion to the degree of 'arthritic' manifestations.[13]

Plain radiography has been suggested as first-line radiological investigation to differentiate between acute lymphocytic leukaemia (ALL) and JIA, to aid establishment of a correct diagnosis in the child with persistent bone or joint pain.[21] Soft tissue swelling and osteopenia were characteristics of JIA cases, whilst radiolucent metaphyseal bands and coarse trabeculation were almost exclusive to ALL patients.

Malignancies presenting in this fashion include leukaemia and neuroblastoma.[19,20,22]

Bone tumours

Osteosarcoma and osteogenic sarcoma are uncommon but serious causes of recent-onset limb pain or limp in the paediatric population. Characteristic features include:

- osteosarcoma – bone destruction and infiltration
- osteogenic sarcoma (Ewing's tumour) – periosteal new bone formation, 'sunray' and 'onion-skin' appearance.

Differential diagnosis includes neuroblastoma and osteomyelitis. These children should be referred for urgent orthopaedic review and investigation.

Osteoid osteoma is an occasional cause of musculoskeletal pain, with pain more pronounced at night. X-rays show an area of periosteal thickening and new bone formation around a central radiolucency.

Simple and aneurysmal bone cysts may also be seen, either as incidental findings or in the setting of pathological fracture. All of these can be referred to the orthopaedic unit for further evaluation.

Other important subacute paediatric musculoskeletal presentations

Traction apophysitis

These disorders are the result of repetitive traction of the growth cartilage. Commonest sites are the tibial tuberosity (quadriceps via patellar tendon: Osgood–Schlatter disease) or the calcaneal apophysis (calf muscle via Achilles tendon: Sever's disease), but the condition can occur at the site of any apophysis, depending on the biomechanical demands of the activity the child participates in. In Osgood–Schlatter disease, the child or adolescent typically presents during the growth spurt with a story of anterior knee pain getting up from sitting or going up or down stairs, with tenderness and at times swelling localised to the tibial tuberosity. In Sever's disease there is pain on walking or running and heel tenderness. Although the X-ray may show fragmentation of the apophysis and soft tissue swelling, an X-ray is not recommended, as this is primarily a clinical diagnosis (and fragmentation may be a normal radiological variant in developing apophyses). Treatment includes rest and/or activity modification, non-steroidal anti-inflammatory medication, physiotherapy and rheumatology or orthopaedic referral to rule out alternate pathology or to manage persisting pain and disability.

Torticollis

The infant presenting with torticollis, without specific trauma or abnormal neurology, is most likely to have congenital muscular torticollis ('sternomastoid tumour'). Theories of pathogenesis include in utero crowding and compartment syndrome. Characteristic features include:

1. onset from birth, often brought to parents' attention by new observer
2. 'cock-robin' appearance (head tilt on involved side with contralateral chin rotation) with range of motion limited by the affected sternomastoid tightness
3. normal neurology, behaviour and occipitofrontal circumference
4. firm painless swelling or tightness palpable within the sternomastoid muscle opposite the chin
5. mild facial hemihypertrophy due to increased relative blood flow to the dependent side.

Hips should always be checked, as a significant proportion may have developmental dysplasia.[23] Infants with this characteristic constellation of features should be referred for physiotherapy and follow-up.

Other causes of torticollis in the paediatric period include vertebral anomalies such as Klippel–Feil syndrome and neurological disorders including brain and spinal cord tumours and ocular dysfunction. Wide-based gait and frequent falls may be accompanied by cervical spine tumours. Children with JIA may present with abnormalities in head and neck posture caused by cervical spine arthritis as their initial presentation, although symmetrical reduced range of motion is the more common finding.

Causes of acute torticollis in children with previously normal neck posture and motion include atlantoaxial rotary subluxation (see Chapter 24.2), a reaction to other acute head or neck pathologies, such as lymphadenopathy or retropharyngeal abscess, and short-term muscle spasm in association with respiratory tract disorders or minor trauma. Features suggestive of significant underlying pathology and need for radiological and specialist referral include:

- other congenital or orthopaedic anomaly
- abnormality of neurological or ophthalmological assessment
- symptoms suggestive of intracranial pathology, e.g. headache, vomiting, irritability
- atypical musculoskeletal examination, e.g. limited range of motion, visual disorder, or persistent or fixed torticollis.

Vertebrospinal inflammation

While uncommon, vertebrospinal inflammation and infection are characterised by delay in diagnosis and diagnostic confusion.[23] These children may present with back or abdominal pain or with altered gait or neurological dysfunction. In discitis and vertebral osteomyelitis there will be point tenderness over a spinous process, particularly to percussion, and localised scoliosis or muscle spasm. Fever and constitutional symptoms are variable. Inflammatory markers are characteristically elevated. X-ray may show muscle spasm or soft tissue oedema, or an abnormal intervertebral disc space. Periosteal reactions may be present in the child with symptoms for greater than 7–10 days. Bone scans are helpful to localise an abnormality where examination findings are equivocal. Definitive diagnosis and evaluation are best made by MRI. Treatment with bed rest and intravenous antibiotics is usual, although isolated discitis may respond better to steroid treatment.

Conclusion

Musculoskeletal presentations of children and adolescents to emergency rooms have a wide range from acute illnesses requiring immediate treatment to the more indolent conditions with longer time since symptom onset. Such presentations are age dependent and their urgency may also be related to patient characteristics and their respective environment. Eliciting accurate medical history as well as a detailed medical examination is an essential requirement before investigations and ultimately a rational management plan can be improvised. Early communication with relevant specialties will aid the efficient delivery of the above healthcare path.

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25.2

Child with a limp

Shefali Jani, and Damien McKay

ESSENTIALS

1 Trauma and transient synovitis (TS) are the most common causes of limp presenting to the emergency department.

2 Less common but potentially significant causes of atraumatic limp (septic arthritis, osteomyelitis, solid tumour, leukaemia, slipped upper femoral epiphysis [SUFE], inflammatory arthritis or neurological causes) should also be considered and where necessary ruled out.

3 Children often have a history of incidental trauma: clarify the timing and significance of the trauma as it may not be the cause of the child's symptoms.

4 Isolated knee pain may be the only presenting feature of serious hip pathology.

5 Careful attention to history and close assessment of posture, range of motion and response to palpation are required to delineate focal pathology.

Introduction

Limp is a common presentation to the emergency department (ED) and, although frequently related to a single traumatic episode, the differential diagnosis for limp is wide. As such it is important to have a structured approach to the limping child, a good understanding of common ED presentations of limp, as well as knowledge of a wider list of potential differential diagnoses. The key to correct

diagnosis is a thorough history, a skilled physical examination and judicious choice of investigations.

Many of the ED presentations of limp can be broadly considered then approached in the following way:

Traumatic limp:

Confirm the presence of trauma. The mechanism of injury may provide insight into likely injury. Soft tissue injuries are less common in paediatric populations thus X-rays are frequently indicated and orthopaedic referral is indicated for confirmed fractures and when fractures are suspected on the basis of history and physical examination yet the X-ray appears normal.

Atraumatic limp without fever:

Confirm absence of significant trauma or fever. Adequate analgaesia when the child first presents may assist in assessment and disposition. A good physical examination is needed to localise the cause of the limp and to rule out a non-musculoskeletal cause. If there is no response to adequate analgesia then obtain X-rays, full blood count (FBC), C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) and consider orthopaedic consult.

Atraumatic limp with fever:

Osteomyelitis and in particular septic arthritis are the key diagnoses to exclude. A painful, swollen joint with limited range should be considered septic arthritis until proven otherwise. FBC, CRP, ESR and blood culture are always indicated. Early discussion with the ED consultant and the orthopaedic team are necessary.

The common causes of limp in children are outlined in Table 25.2.1. The wide spectrum of causes of acute limp in children is illustrated in Box 25.2.1.

Diagnoses that require specific treatment to avoid further damage or danger are primarily displaced or unstable fractures (including slipped upper femoral epiphysis), abusive injury (as a presenting feature or incidental finding), bone or joint infection, and neoplastic processes, principally bone tumours and leukaemia.

History

- Duration of limp, precipitants and exacerbating factors:
    - Note that a toddler may simply stop walking or go back to crawling rather than limping.
    - History of trauma.
- History presented in many cases (all children fall) may not be the cause.
- If a traumatic injury is present the limp will occur immediately.
- An injury followed by a period of normal movement/use suggests that the injury may not be the cause of the limp.

NB: in non-accidental injury a history of trauma may not be forthcoming.

- Pain location and type: note that hip pathology may give referred knee or thigh pain.
- Night pain, bony lumps
- Presence of systemic symptoms:
    - Fever, pallor, bruising:
        - Leukaemia (acute lymphoblastic leukaemia) can present with limp as the only symptom in a well-looking child; bleeding disorders may present with limp due to haemarthrosis or muscle haematoma.
    - Rashes:
        - Present in herpes simplex virus, some viral infections, serum sickness (i.e. ceclor reactions)

Table 25.2.1

Common causes of acute atraumatic limp

Infants	Young children	Primary school-age children	Adolescents
Occult fracture[a]	Occult fracture[b]	Transient synovitis	Septic arthritis/ osteomyelitis
Septic arthritis/osteomyelitis Inflammatory arthritis	Transient synovitis Inflammatory arthritis	Septic arthritis/osteomyelitis Inflammatory arthritis	SUFE Inflammatory arthritis
	Septic arthritis/osteomyelitis Perthes' disease	Perthes' disease Psychogenic pain	Osteochondroses Tumour

SUFE, slipped upper femoral epiphysis.

[a] Includes pulled elbow, accidental and non-accidental injury.

[b] Includes pulled elbow, toddler fracture.

Box 25.2.1 Causes of acute limp in children

Trauma

- Fractures – accidental and non-accidental

Infection (point focus)

- Septic arthritis
- Osteomyelitis
- Inguinal lymphadenitis
- Muscle abscess, e.g. psoas

Post-infective

- Rheumatic fever/post-streptococcal arthritis
- Post-infectious arthritis, e.g. *Salmonella, Shigella*, or *Campylobacter* enteritis
- Serum sickness
- Post-immunisation inflammation

Inflammatory

- Transient synovitis
- Vasculitis, e.g. Henoch–Schönlein purpura or Kawasaki disease
- Inflammatory arthritis in lower limbs or axial skeleton, e.g. oligoarthritis
- Enthesitis in pelvis or lower limbs, e.g. enthesitis-related arthritis
- Associated with, e.g. systemic lupus erythematosus, dermatomyositis or inflammatory bowel disease

Primary bone disorders

- Slipped upper femoral epiphysis (SUFE)
- Avascular necrosis, e.g. Perthes' disease (hip), Freiberg's disease (metatarsal heads)
- Osteochondroses, e.g. Osgood–Schlatter disease (patellar tendon insertion), Sever's disease (Achilles tendon insertion)
- Unicameral bone cyst/aneurysmal bone cyst/fibrocystic disease/eosinophilic granuloma
- Blount disease (asymmetrical tibial physis closure)
- Tarsal coalitions

Neoplastic

- Leukaemia
- Neuroblastoma
- Bony tumours, e.g. Ewing's sarcoma, osteosarcoma
- Non-malignant tumours, e.g. osteoid osteoma,

enchondroma

Haematological

- Haemarthrosis, e.g. haemophilia A
- Sickle cell disease arthropathy

Physical

- Splinter/foreign body
- Compensatory, e.g. footwear
- Soft tissue and overuse injury
- Joint hypermobility syndrome
- Plantar warts/calcaneal spurs
- Bites and envenomation

Psychological and idiopathic pain syndromes

- Pain amplification syndromes, conversion disorders
- Complex regional pain syndrome (CRPS, previously known as reflex sympathetic dystrophy)

Abdominal

- Appendix
- Infective and inflammatory bowel disease

Spine

- Scoliosis
- Discitis
- Transverse myelitis
- Spondylolisthesis
- Scheuermann’s disease
- Guillain–Barré syndrome

- Prodromal or intercurrent illness (upper-respiratory-tract infections, sore throat or enteric infections)
- Neurological symptoms:
    - Unilateral weakness or asymmetrical reflexes suggesting spinal cord lesions, new bowel or bladder incontinence
- Constitutional symptoms:
    - Weight loss, unexplained fevers with tumours, inflammatory disorders and chronic infections.

Examination

The approach to musculoskeletal examination in the child is discussed in Chapter 25.1 Orthopaedics and rheumatology.

Key features in regards to examining the child with a limp include the following:

Assess if well or sick, observations should be included:

- Fever (absence does not exclude septic arthritis or osteomyelitis)
- General examination:
  - Rashes, lymphadenopathy, hepatosplenomegaly, bruising or pallor
- Observe the gait:
  - Ability to weight bear
  - Antalgic gait
  - Trendelenberg gait (suggests hip pathology)
- Examine the legs and spine:
  - Fixed flexion deformity of hip
  - Bruising, deformity, swelling, foreign bodies
  - Bony tenderness (include spine)
  - Range of joint movement (including spine)
  - Ensure joint is moved through entire range of motion and range is compared to the non-affected side before range of motion can be considered normal.
- Neurological exam:
  - Lower limb tone, power, deep tendon reflexes and sensation
- Abdominal examination:
  - Organomegaly, localised tenderness and testicular exam.

If hip pathology is suspected internal rotation of hip (assessed with the hip flexed to 90 degrees) is important to assess as a subtle difference in range of motion with pain may be the only indication of a hip joint effusion. The ‘flexion adduction’ test 1 may also be helpful: with buttocks flat on the bed, flex the hip

(first the unaffected and then the affected) to 90 degrees while supporting the lower leg. Then gently attempt to fold the knee over the contralateral leg without the child lifting the buttock off the couch. In the normal hip this adduction should allow the ipsilateral knee to be positioned over the opposite leg whereas an inability to adduct past the midline is suggestive of hip joint pathology, such as slipped upper femoral epiphysis (SUFE) and Perthes' disease.

Investigation

In the ED setting, key investigations relevant to the limping child include:

- FBC and inflammatory markers (ESR and CRP) to rule out infection and malignancy
- X-rays, which should be performed when:
    - there is a history of trauma suggestive of fracture
    - localised bony pathology is evident on clinical exam
    - the older child or adolescent presents with hip pain and/or limp.
- Ultrasound is useful in confirming the presence of a hip joint effusion.
- Bone scan or MRI is useful in identifying the site of possible bacterial infection in a child with localised musculoskeletal pain and a septic picture for whom septic arthritis has been ruled out by clinical assessment, ultrasound or arthrocentesis.

A more detailed discussion of investigations useful in the assessment of musculoskeletal pathology is included in Chapter 25.1.

Clinical decision making in a child with a limp

The child whose examination findings suggest an isolated irritable hip is likely to have one of the pathologies outlined in Table 25.2.2. A decision-making algorithm for managing acute irritable hip is illustrated in Fig. 25.2.1. Spinal, abdominal and pelvic pathology can present as a limp and must be considered prior to this narrowed focus. The infant or toddler, because of their increased risk for invasive bacterial illness, also represents a special circumstance and the ill child under 2 years with an abnormal acute musculoskeletal assessment is best admitted for combined paediatric and orthopaedic assessment and investigation.

Table 25.2.2

Comparative features of paediatric hip-joint pathology

Diagnosis	Transient synovitis	Septic arthritis	Femoral neck osteomyelitis	Perthes'	SUFE	Inflammatory arthritis	Bone tumour
Age	3–6	Any	Any	3–12	9–15	Any	Any
Joint	Hip	Hip, knee	Hip	Hip	Hip	Any	Not joint
Pain (0–3)	1	3	1	0–1 early	1–3	0–2	=1
Illness (0–3)	0	1–3	1–3	0	0	0–2	Variable
ROM	>80%	<50%	<80%	60–80%	Flexes into external rotation	Usually some movement possible	Normal unless spasm
Inflammatory markers	Normal	Elevated	Elevated	Normal	Normal	Variable	Variable
Ultrasound	Effusion	Effusion	?Reactive effusion, ?subperiosteal collection	Diagnostic changes	Diagnostic changes	?Effusion	n/a
Plain X-ray	HTDD up	HTDD up	Bony changes delayed	Diagnostic changes eventually	Diagnostic changes, e.g. widened physis in preslip; Trethowan's sign	May be normal	May be diagnostic

HTDD, head-teardrop distance; ROM, range of movement.

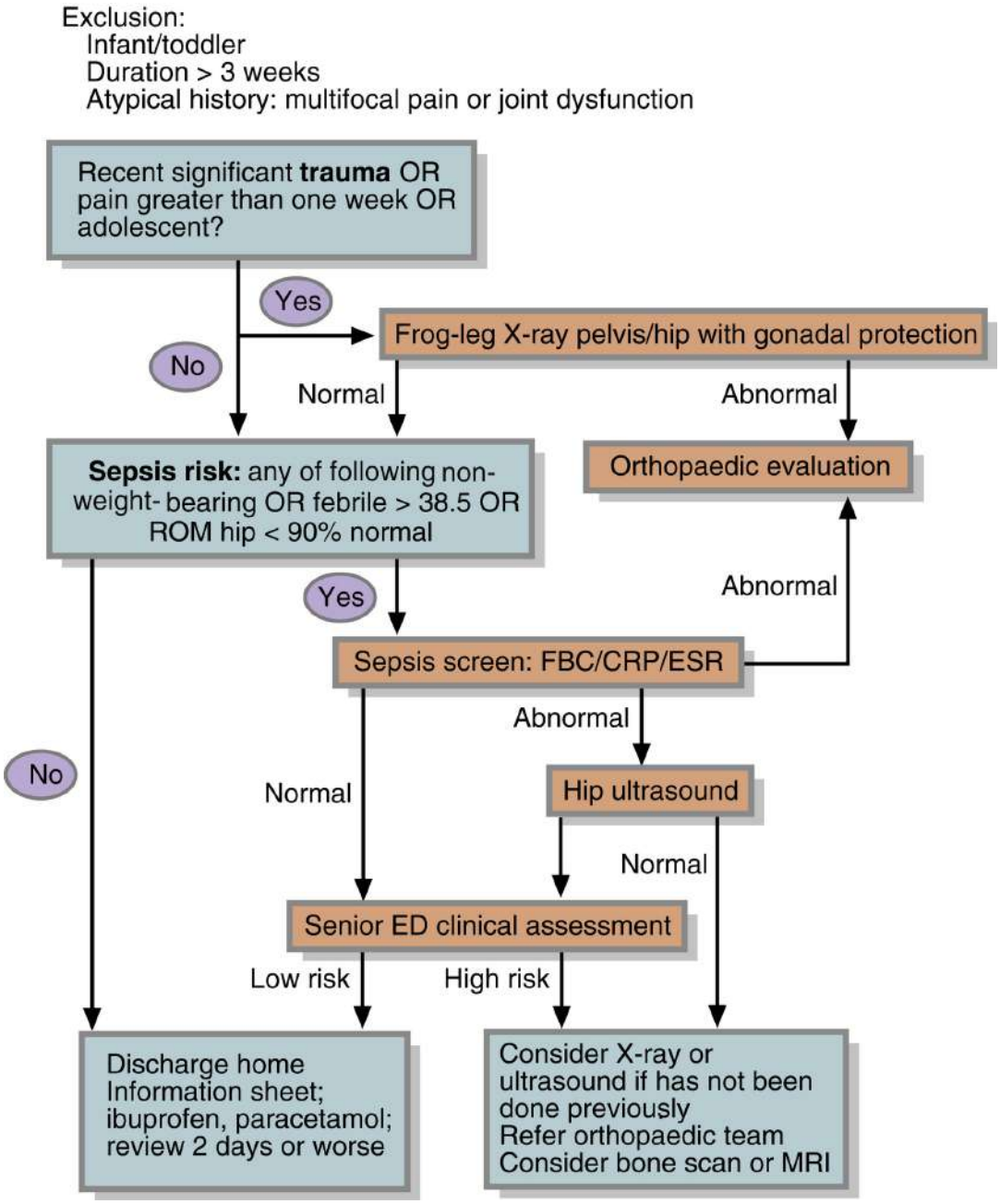


FIG. 25.2.1 Algorithm for management of children >2 years presenting with acute irritable hip.

Box 25.2.2 Risk factors for septic arthritis

Relative immune deficit

- Neonates

- Malnourished
- HIV infection
- Immunosuppressive therapy
- Corticosteroid therapy

Injury mechanism

- Penetrating trauma

Joint disease

- Chronic arthritis
- Sickle cell disease

The child over 2 with acute, atraumatic and localised knee or hip pain and an abnormal hip examination is most likely to have an irritable hip (transient synovitis), and the main issue is to exclude septic arthritis. An ultrasound assessment should confirm the suspicion of a hip effusion as suggested by examination but does not differentiate between transudate and exudate. Box 25.2.2 lists risk factors which increase the risk of septic arthritis; however, the majority of infections occur in children without underlying pathology. Various attempts have been made to create a valid decision rule with a high sensitivity and specificity for bacterial infection;[1–3] however, none has demonstrated sustained power prospectively. The five most important variables to give a predicted probability of septic arthritis in a given child with an acutely irritable hip are shown in Box 25.2.3. Degree of pain and range of motion of the joint are auxiliary clinically important variables in the differentiation.

Box 25.2.3 Features suggestive of deep bacterial infection in a child with an acutely irritable hip

Non-weight-bearing
Febrile >38.5°C
WCC > 12×10^9 L^{-1}
ESR > 40 mm h^{-1}

CRP > 20 mg L^{-1}

Some of the other common acute diagnostic dilemmas include differentiating discitis from vertebral osteomyelitis and psoas abscess from septic arthritis.[4] Due to overlapping clinical features in malignancies and rheumatologic conditions, differentiating these pathologies remains a challenge.

Specific conditions

Transient synovitis

Transient synovitis (TS) is a benign, self-limited inflammatory disorder of uncertain aetiology resulting in synovial inflammation of the hip with associated effusion. It is seen particularly in boys (70%) in the age group 3–10 years. It is often preceded by a viral upper-respiratory-tract infection. Bilateral joint involvement is present in up to 30% of cases.

History

The onset of lower-limb pain is generally gradual and initially may be localised to hip, knee, groin or thigh with a limp or unwillingness to weight bear. The child may be mildly unwell with or without a history of recent non-specific upper respiratory tract illness.

Clinical examination

The affected hip may be positioned in variable degrees of flexion, abduction and external rotation (a position likely associated with lower intracapsular pressure). Range of movement at the hip joint is usually mildly diminished, with discomfort at the end-points of the range.

Differential diagnosis

TS is a diagnosis of exclusion, the major differential being septic arthritis.

Management

Children with localised hip pain should be managed according to the algorithm in Fig. 25.2.1, and non-weight-bearing children with a hip effusion who do not have needle aspiration to rule out septic arthritis should be admitted under the

close observation of the orthopaedic team. Weight-bearing children without unusual or high-risk features can be managed with parental instruction, ibuprofen and ED follow-up in 2 days.[5,6]

Prognosis

Taylor et al. showed a 15% recurrence rate in transient synovitis,[7] and there is some concern about occult Perthes' being an underlying diagnosis, especially in those with delayed bone age at the first imaging.[8] All children discharged with this presumptive diagnosis should have orthopaedic follow-up if pain or limp persists or recurs.

Septic arthritis and osteomyelitis

ESSENTIALS

1 Septic arthritis in children is an orthopaedic emergency with delay or inadequate treatment leading to irreversible joint damage and/or septicaemia.

2 Diagnosis of septic arthritis is suspected on clinical features, confirmed by blood and synovial cell count as well as fluid culture. Isotope scans and MRI have an important role in localising osteomyelitis.

3 *Staphylococcus aureus* and *Streptococcus* species are the most frequent pathogens, via haematogenous spread or direct extension from infected joint to bone. Community-acquired methicillin resistant *S. aureus* (CAMRSA) is increasingly common worldwide and mandates alternative antibiotic treatment.

4 Successful treatment requires parenteral antibiotics and complete drainage of pus from the joint, or surgical clearance of necrotic bone, with long-term follow-up.

Introduction

Septic arthritis is infection of the synovial lining and fluid of a joint. Although haematogenous bacterial spread is the commonest cause of septic arthritis, direct inoculation through penetrating injuries or surgery is known to occur. Direct spread from adjacent bone infection may also occur resulting in osteomyelitis, particularly in joints where the metaphysis is intracapsular as in the hip and shoulder. Phagocytic and neutrophil responses to the bacteria result in proteolytic enzyme release and cytokine production; with synovial abscess formation and cartilage necrosis.[9] Increased intracapsular pressure secondary to pus accumulation may reduce epiphyseal blood flow resulting in growth plate damage and avascular necrosis of the femoral head.

Most infections in children are community acquired and occur in normal joints. Infants and children under 3 years of age are at particular risk of septic arthritis, comprising one-third and one-half, respectively, of a large paediatric series.[10] Along with focal clinical inflammation, children may present with occult infection, pseudo-paralysis or generalised sepsis. Comorbidity or deficient host defences, such as those shown in Box 25.2.2, predispose to infection that may be more rapidly progressive or occur in the older child.

Joints of the lower limb, especially hip and knee, account for two-thirds of the infections in children. Other commonly infected joints include the ankle and shoulder; however, any synovial joint in the body could be affected.

In osteomyelitis, bacteria enter the vascular metaphyseal bone initially then typically extend to the sub-periosteal space forming an abscess. New bone deposition results, with later necrosis of cortical bone. Classically, bone fragments or sequestra are formed over time, which harbour bacteria. Successful treatment must combine eradication of the bacteria and complete removal of any necrotic infected bone. However, in the developed world, earlier diagnosis and aggressive antibiotic therapy have limited the degree of bone destruction present.

The disorder of CRMO (chronic recurrent multifocal osteomyelitis) has been recognised in infants and children. This is an inflammatory process of unknown origin, which demonstrates culture-negative bony inflammation with histological evidence of necrosis and chronic inflammatory cell infiltrates. Biopsy and culture are mandatory to diagnose the disorder, but antibiotics may be discontinued if pathology is consistent with CRMO. In mild cases, treatment of symptoms may be possible with non-steroidal anti-inflammatory drugs (NSAIDs); however, in more severe cases systemic steroids or treatment with intravenous bisphosphonates, and long-term follow-up will be required.[11–15]

Presentation

History

Cardinal features of septic arthritis are recent onset of a painful, red or swollen joint or limb, limp or refusal to bear weight. Pseudoparalysis or refusal to move a limb may occur in neonates and young infants. Infants may present with non-specific symptoms, such as poor feeding, vomiting, lethargy or fever. Infrequently, a vertebral or pelvic infection may be the cause of an abnormal gait or may present with abdominal pain. Relevant past history includes trauma, past history of recurrent staphylococcal infection, such as boils, and immunisation status in respect to *Haemophilus influenzae* and *Pneumococcus*. Consider *Salmonella* osteomyelitis as a potential pathogen in patients with sickle cell disease. Children with methicillin-resistant *S. aureus* (MRSA) osteomyelitis are often more unwell looking than those with methicillin-susceptible *S. aureus* (MSSA), with high temperature, tachycardia and a painful limp .[16]

Examination

Typical findings in septic arthritis include a warm tender joint with marked limitation of passive and active movement due to pain. Children with a painful hip will usually maintain the joint in slight (20 degrees) flexion, abduction and external rotation (in a position of least intracapsular pressure). An effusion is usually clinically evident in peripheral joints except the hip. In general, fever is low grade and most patients will not appear ‘toxic’ or unwell. Osteomyelitis may be suggested by an area of maximal tenderness next to a joint, with a greater range of movement of that joint than would be expected with septic arthritis. Deep or partially treated infections may be clinically subtle, with minimal specific examination findings. Careful examination of the skin may reveal areas of infection or trauma as an entry point for haematogenous seeding. Discitis and spinal osteomyelitis usually present with back pain (focal area of tenderness on examination of the spine).

Investigations

The sensitivity of a raised white cell count (WCC) ($>12 \times 10^9$ L^{-1}) for the identification of septic arthritis is variable (20–75%).[17,18] The absence of an elevated white cell response should never be used to rule out septic arthritis. As such inflammatory markers ESR and CRP should also be performed whenever

infection is considered. ESR has shown higher sensitivity (90–95%) in identifying the subgroup with septic arthritis; however, this may miss early infection.[17] ESR levels fall slowly and hence may not be as useful (when compared to CRP) in monitoring progression of disease. CRP has shown superior sensitivity (up to 80%) [18] and specificity in the early identification of invasive bacterial infection in general and of septic arthritis in particular. CRP rises within 24 hours of acute illness, and also falls rapidly, and can be used to monitor effectiveness of treatment.[19]

A summary of features including investigations suggestive of deep bacterial infection in a child with an acutely irritable hip are shown in Box 25.2.3.

Microbiology

Synovial fluid examination and culture

A cell count of greater than 50,000 per microlitre of synovial fluid aspirate suggests a bacterial cause, although positive cultures can occur with lower counts, and higher counts may occasionally occur in inflammatory conditions.[20] Synovial fluid should also be inoculated directly into culture medium to avoid loss of fastidious organisms such as *Kingella*.[21] Initial Gram stain result (Gram stain may only be positive in up to one-third of cases) and clinical history will guide empiric antibiotic therapy, with definitive therapy based on final cultures.

Organisms implicated in two series involving Australian children[18,22] are shown in Box 25.2.4. An algorithm for initial intravenous therapy of paediatric septic arthritis and osteomyelitis is shown in Fig. 25.2.2.

Box 25.2.4 Organisms cultured in paediatric invasive bone and joint infection in Australia

Most common (80% of most isolates)

Staphylococcus aureus
Streptococcus species

Other organisms

Pneumococcus
Kingella kingae

Fusobacterium
Meningococcus
Pseudomonas
Salmonella

Negative cultures of blood, synovium, or infected bone occur in 40–80% of cases of paediatric septic- and osteomyelitis worldwide.[18,20-22] Increasing availability of polymerase chain reaction (PCR) may increase specific bacteriological identification.[23] Despite increasing CAMRSA and a high negative culture rate, the Australian experience has also been of a low rate in treatment failure, presumably due to early case identification and susceptible organisms. However, the increased prevalence of CAMRSA in the USA has been associated with an increase in severity of osteomyelitis.[24,25] Tuberculosis is still a cause of chronic bone and spine infection in South-East Asia, Papua New Guinea and the Torres Strait Islands and should be considered in atypical cases.[26,27]

There is controversy over the place of ultrasound-guided hip aspiration in the evaluation of the irritable hip with an effusion.[28] The majority of Australian paediatric orthopaedic units favour orthopaedic evaluation of irritable hips with selective aspiration and arthrotomy/washout under general anaesthesia. However, ultrasound-guided diagnostic hip aspiration has a role in some units.[29–31] Ultrasound-guided aspiration of synovial fluid can be useful in diagnosing septic arthritis and may provide immediate but short-term pain relief effect in patients with TS having symptomatic large hip effusions.[32]

The overriding principles are early accurate diagnosis, and minimal delay or exposure of the joint to the chondrolytic enzymes of bacterial joint infection.[33]

Management

Although studies have shown successful conservative treatment,[34] joint drainage with lavage of the joint and empiric parenteral antibiotic therapy are the mainstays of treatment for septic arthritis and should take place without delay, although ideally antibiotics should not be commenced until synovial fluid sample is obtained.

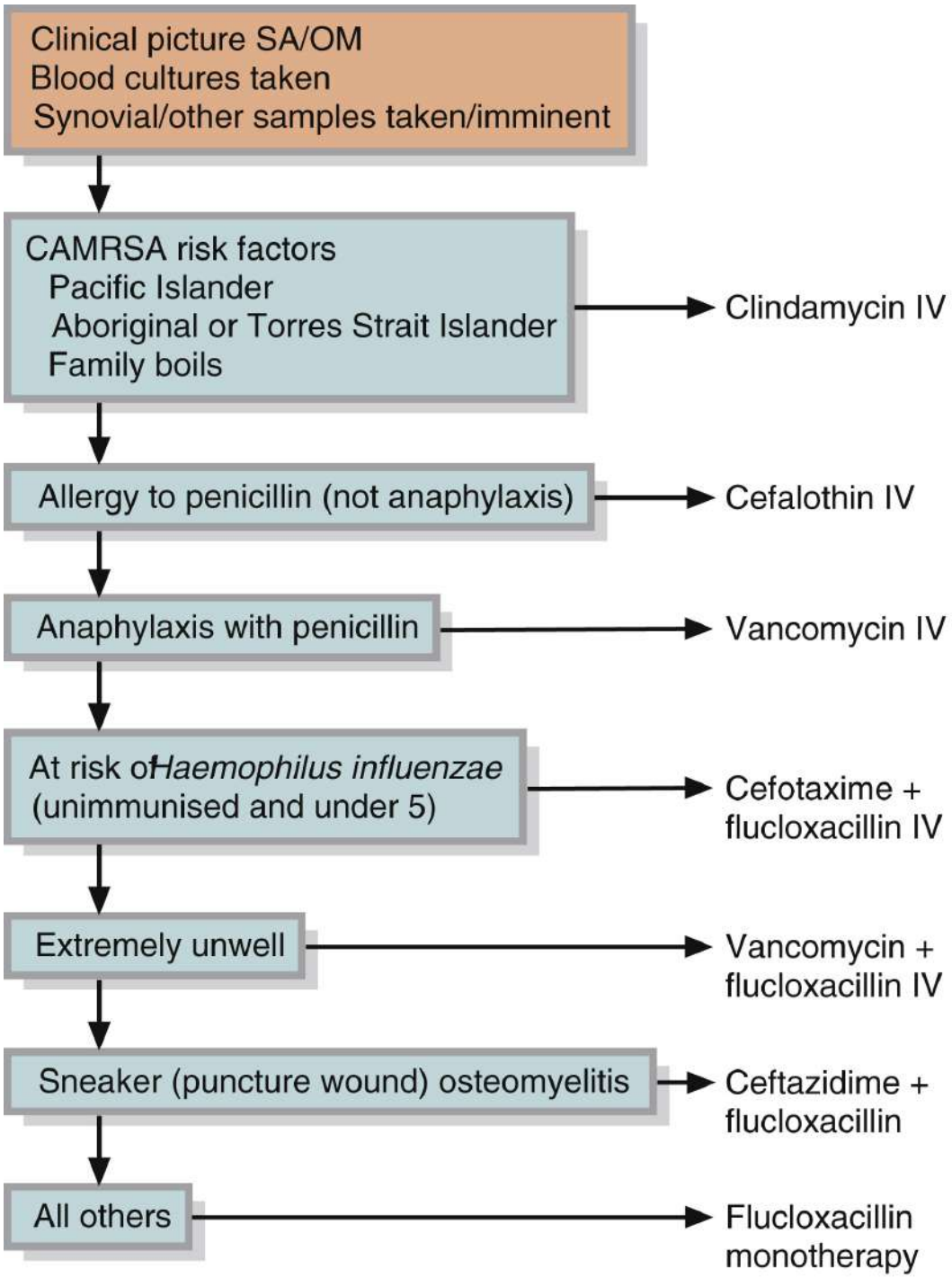


FIG. 25.2.2 Antibiotic therapy for septic arthritis (SA)/osteomyelitis (OM) in Australia.

Suggested antibiotic regimes for bone and joint infection are shown in Fig. 25.2.2 and should be further guided by Gram stain and culture/sensitivity results. A prolonged course of antibiotics may be required in neonates, immunocompromised patients, children with sickle cell disease and osteomyelitis associated with septic arthritis. The duration of antibiotics is variable and depends on the age of the child, organism identified, source of infection and presence of underlying risk factors. Because of earlier diagnosis and other aspects of modern care, in most cases beyond the neonatal period combined in-patient/out-patient treatment can occur with length of treatment guided by clinical and inflammatory markers.[18]

Surgical treatment is undertaken for three main reasons including confirmation of microbiological diagnosis, source control for effective antimicrobial penetration and preservation of joint function to minimise development of long-term complications. Surgical options (aggressive debridement) may be considered in a child with osteomyelitis if medical management is ineffective in difficult-to-treat MRSA cases (lack of sufficient data to support this) and in case of intraosseous abscess development in subacute and chronic osteomyelitis (Brodie's abscesses).[16]

Community-acquired methicillin resistance is becoming an increasing problem world-wide. Specific populations at high risk of CAMRSA include Aboriginal and Torres Strait Islanders in Western and Northern Australia, and Pacific Islanders in Eastern Australia and parts of New Zealand.[35] Children with a personal or family history of recurrent treatment for boils are also at risk.[36]

Neonatal septic arthritis requires special consideration in view of the destructive potential and broader microbiological differential.[37] These children should be reviewed by a team of orthopaedic, neonatal and infectious-disease specialists.

Prognosis

This depends upon the organism, patient comorbidity, age of patient and the adequacy and rapidity of treatment. Remarkable remodelling of bony deformity can occur in the young child provided treatment has been adequate. Out of a 30-year series of 332 infants and children with osteomyelitis, with documented follow-up of 170 cases, complications were described in 19%.[10] These related largely to joint complications; however, 8% demonstrated epiphyseal damage. However, more recent series showed many fewer complication rates.[38] Patients at highest risk of complication include neonates, septic arthritis in which

diagnosis has been delayed, complicated osteomyelitis (involucrae, sequestrum or sinus formation) or epiphyseal involvement, which may cause subsequent limb-length discrepancy or deformity. All cases should have close orthopaedic follow-up.

Prevention

Primary prevention involves minimising skin or muscular trauma or sepsis (e.g. boils, infected scabies), since damaged tissue predisposes to haematogenous bacterial spread of skin micro-organisms. Secondary prevention can be provided by thorough debridement, wound cleansing and antibiotics in the management of open bone trauma.

Perthes' disease

ESSENTIALS

1 Perthes' disease is a chronic disorder of the femoral head in children, causing limp and usually low-grade pain. While most children outgrow their disease, a proportion has permanent structural changes requiring orthopaedic correction.

2 Clinical findings include decreased range of hip motion, particularly internal rotation and adduction, with low-grade discomfort.

3 X-ray changes of established disease include joint effusion, loss of femoral head height and fragmentation of epiphysis.

4 Early X-ray changes may be subtle and children with persisting undiagnosed limp should have MRI examination of their hip and, if necessary, orthopaedic follow-up.

Pathophysiology

Legg–Calve–Perthes' disease is a disorder of unknown aetiology resulting from disruption of blood supply to the capital epiphysis, principally affecting boys (M:F 4:1), commonly in the 4–8-year age group. It is bilateral in approximately

10% of cases. The pathophysiology affects articular cartilage, where synovitis results in oedema, hypermetabolism, hypertrophy and deterioration of the cartilaginous mechanical properties. Cartilaginous ischaemia is thought to be the active mechanism. It is more prevalent in malnourished children,[39] those with delayed bone age, and those exposed to passive smoking.[8,40,41] Other proposed causes include genetic mutations, coagulation abnormalities, traumatic injury to the blood supply and venous congestion.[42] The end result may be subchondral fracturing, anterolateral deformation and joint incongruence, although the majority of children have spontaneous gradual remission of their disease process. Four stages are recognised:[43]

1. Initial – ischaemia causes cartilage hypertrophy and synovitis; X-ray smaller femoral head, increased joint space.
2. Fragmentation – X-ray shows fragmented epiphysis and subchondral radiolucency (‘Caffey’s crescent line’).
3. Reossification – altered shape of femoral head with increased radiopacity.
4. Healing – resolution or persistence of deformity.

Emergency department presentations

When hip pain is present in Perthes’ disease it is usually mild, chronic and dull, increasing with physical activity, often with a history of pain for weeks to months. There are no systemic symptoms. Because of the gradual onset of Perthes’ disease, acute presentations to the ED usually represent either an early (stage 1) Perthes’ and fall within the ‘irritable hip’ differential, as discussed previously, or a flare-up in a child with known Perthes’ disease in whom coincidental comorbidity must be excluded. It is important to remember Perthes’ disease can often present as a painless limp.

Examination

Children with Perthes’ disease have limited range of hip movement, particularly in adduction and internal rotation, from synovial thickening and adductor muscle spasm. Those with longstanding disease may have muscle atrophy, leg-length discrepancy, flexion contractures and a positive Trendelenburg test on the affected side.

Differential diagnosis

In the early stages of disease, X-ray changes will mimic those of TS, and children should be managed according to the irritable hip outline suggested earlier.

Children with more minor hip discomfort and non-specific X-ray or ultrasound evaluations should be referred for MRI examination of hip for detection of early stages of avascular necrosis[44–47] and orthopaedic evaluation if symptoms persist longer than 3 weeks.

Investigation

The plain hip X-ray is diagnostic in established Perthes' disease, although findings may be subtle in early disease. Extensive head involvement, loss of the femoral head 'lateral pillars', subluxation of head beyond the acetabular margin, and late age at presentation are all poor prognostic indicators. MRI examination of the hip joint may be necessary to provide early diagnosis and prognosis (references from above). Perfusion MRI (gadolinium-enhanced subtraction) has been shown to be safe and effective in assisting with classification of the disease, prognosticate outcomes and search for associated abnormalities.[42]

Classification

Currently the Herring lateral pillar classification is most widely used to guide treatment decisions and anticipate the expected outcomes.[42] The lateral pillar is the lateral 15–30% of the femoral epiphysis on an anteroposterior radiograph and the amount (percentage) of collapse of the involved pillar is used to determine the four groups in the classification and is meant to assist in treatment decisions.

Management and prognosis

Preventable treatment strategy is more effective in the early stage than in late stages of disease. The common principle of any surgical intervention used in the management of Perthes' disease is to protect the weak and fragmented head from deforming forces until it is able to reform and reconstitute. Orthopaedic management goals are restoration of range of hip movement and containment of the hip within the acetabulum. These goals are sought through a variety of methods including traction, surgery, orthotics and physiotherapy. In addition to the simple observation, bracing and physiotherapy, conservative management

also includes the use of biological methods such as bisphosphonates, which has been under investigation.

The strongest predictor of long-term outcome is the relationship between the shape of the femoral head and congruency within the acetabulum in addition to the age of presentation.[42] In general, the younger the child the better the outcome; girls do worse than boys of the same age as they are skeletally more mature. Some three-quarters of patients with Perthes' disease are pain-free and active in 10–20-year follow-up studies,[43] but may need hip replacements earlier in life because a high percentage develops osteoarthritis around the 4th and 5th decade of life.[48]

Slipped upper femoral epiphysis

ESSENTIALS

1 All adolescents with undiagnosed acute hip or knee pain should have hip X-rays or ultrasound performed by competent operators even in the absence of history of trauma.

2 Non-weight-bearing adolescents with acute hip pain must not undergo passive hip manipulation (even for X-ray) until diagnosis is established.

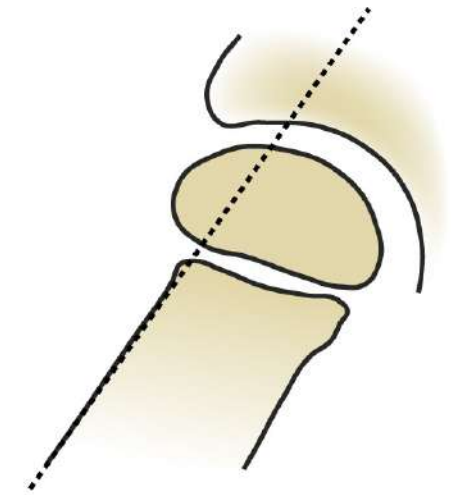
3 Delay in diagnosis and stabilisation of slipped upper femoral epiphysis (SUFE) increases the risks of avascular necrosis, chondrolysis, deformity and long-term poor hip outcomes including degenerative hip arthritis eventually needing hip reconstruction.

4 Early SUFE should not be missed in emergency department (Fig. 25.2.3), and children with suspected SUFE must be immediately referred to orthopaedics.

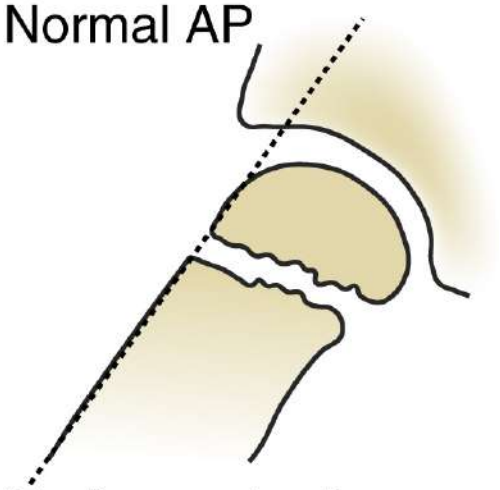
Epidemiology and pathophysiology

SUFE is more common in adolescents, with a peak age of 10–16 years in males and 9–15 years in females, although it has been reported in children as young as

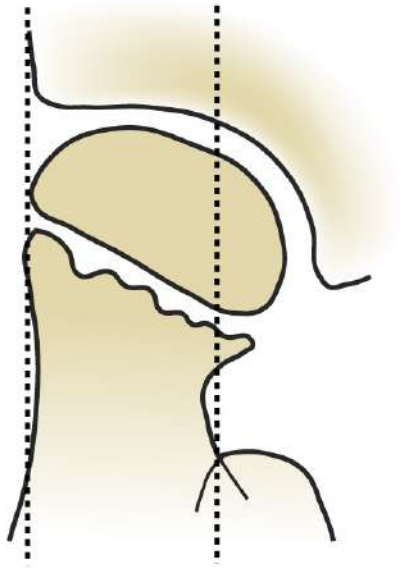
8 years. The condition is defined as posterior and inferior slippage of the proximal femoral epiphysis on the metaphysis (femoral neck) through the epiphyseal plate. SUFE is usually related to puberty (80% occurring during the adolescent growth spurt), and obesity (two-thirds are >90th centile weight-for-height), with genetic and endocrine factors playing a role.[43] Consider an endocrine disorder such as hypothyroidism, hypogonadism, panhypopituitarism and growth hormone supplementation in SUFE patients with unusual presentations, including underweight patients or patients less than 8 years or older than 15 years. Mechanical failure occurs at a widened zone of hypertrophy within the physeal plate. It is important to note that, although traditional Salter–Harris proximal femoral physeal fractures can occasionally occur in adolescence from high-energy injuries, these differ from classical SUFE both in the amount of force involved and in the histological plane of cleavage, i.e. the SUFE can be thought of as a ‘pathological Salter–Harris type 1 fracture’ occurring as a result of minor torsional or low-energy injury in a weakened physeal plate with a high-shear stress load.[43]



Normal AP



Trethowan's sign



Frog leg lateral

FIG. 25.2.3 Slipped upper femoral epiphysis (SUFE). In the normal pelvic anteroposterior (AP), (top) a Klein line (a continuation of the superior border of the femoral neck) should INTERSECT a portion of the femoral head. Trethowan's sign, (middle) in which the Klein line fails to intersect the head in the AP view, is abnormal and represents Grade 1 SUFE. Note the frog-leg view (shown above as in a normal hip) is a better view for diagnosing SUFE in the ambulant child with hip pain, since the posterior slip is usually more clearly shown. However, the hip must not be manipulated into frog-leg position in an unstable (non-weight-bearing) child with possible SUFE, as inadvertent reduction may occur and potentially increase risk of avascular necrosis.

Clinical presentation

A typical presentation of SUFE includes an obese adolescent with pain either in the groin or referred to knee (femoral) or medial thigh (obturator nerve). Leg-length discrepancy may be present with the leg usually held in external rotation. Range of motion depends on degree of slip and chronicity. A classical feature of the chronic slip is obligatory external rotation during hip flexion seen even in mild cases.

Initial presentation of SUFE may take one of four patterns:

1. Preslip. This is usually a retrospective diagnosis, relating to an episode of intermittent pain and evidence of synovitis with widening of the growth plate but without radiological evidence of epiphyseal shift.
2. Chronic slip. Pain longer than 3 weeks in an ambulant child suffering mild to moderate pain, with decreased range of motion. X-ray shows SUFE ± evidence of remodelling.
3. Acute-on-chronic slip. Increased pain and radiological evidence of slip following a period of lower-grade symptoms. Acute slip and evidence of remodelling are present on X-ray.
4. Acute slip (≈10%). Acute, severe hip pain and diminished range of movement with less than 3 weeks' prodrome and no radiological evidence of remodelling.

Thus the most common symptoms of SUFE are limping and poorly localised pain to hip, groin, thigh or knee. The most important clinical sign of SUFE is the limitation in the internal rotation of the hip.

Classification of SUFE is based on the stability of the physis. SUFE is stable

if the patient is able to weight bear with or without crutches (90% of all slips) and is considered unstable if the patient is unable to ambulate even with crutches. This grading is important as unstable SUFE will need immediate attention as they are more likely to result in long-term complications. SUFE is also graded as mild, moderate or severe depending on the percentage of epiphyseal translation.

Plain radiography is used to grade the severity of slip in SUFE using the Wilson's method which measures the relative distance of the epiphysis on the metaphysis in a frog-leg lateral radiograph and the Southwick method which measures the epiphyseal shaft angle on the frog-leg lateral radiograph.[49] The radiological findings in SUFE are outlined in Fig. 25.2.3. It is essential that the emergency physician be aware of the clinical and radiological features of SUFE so that they can recognise at-risk presentations and actively seek Klein's line (a line extending from the superior border of the femoral neck which should intersect the lateral femoral head in the AP view of normal hip/pelvis).

Although the slip is usually radiologically more evident in the frog-leg lateral, manipulation into this position should be avoided in unstable (non-weight-bearing) children with possible SUFE, in case the process of obtaining it may precipitate further shear.

Ultrasound by experienced operators has also been shown to have high sensitivity and specificity for epiphyseal displacement, even in the early stages.[50]

Differential diagnosis

The differential diagnosis of SUFE has been discussed in the section on limp. A normal frog-leg pelvic X-ray in the ambulant child rules out all but the preslip phase of SUFE. In high-risk clinical situations (e.g. past or family history of contralateral SUFE) and persistent unexplained symptoms, further imaging such as MRI should be undertaken.

Treatment

Once an unstable SUFE has been diagnosed, patients should be treated as they are at risk of avascular necrosis. After ensuring that the patient is made non-weight bearing, admission under the orthopaedic team pending surgical fixation should be expedited. Bed rest with toilet privileges is usual, with pain relief as required. The initial goals of treatment include preventing slip progression and avoiding complications. The acutely slipped femoral head is usually fixed by

percutaneous or minimal-incision internal fixation. The pinning of the contralateral side is recommended by some surgeons if the posterior sloping angle is >15°.[51,52]

Complications

Avascular necrosis may occur in up to 50% of unstable slips, with an overall incidence in SUFE of ≈5%. Other complications include chondrolysis and leg-length discrepancy.

Controversies/Developments

1. The necessity or danger of attempted reduction (which may precipitate avascular necrosis) prior to fixation.
2. The value of pinning the contralateral hip (asymptomatic bilateral SUFE evolves in up to 40% of children).

Delays in diagnosis and referral are common and avoidable.[53] Causes of delay include:

- failure to X-ray (particularly with low-grade and/or referred pain)
- failure to recognise knee pain as a presenting symptom of hip pathology
- failure to interpret X-ray correctly, particularly in Grade 1 (ED training should include the X-ray appearance of the normal femoral head)
- failure to refer appropriately (immediate telephone contact with orthopaedic team).

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25.3

Fractures and dislocations

Sarah Martin, and Henry Patrick John Walsh

ESSENTIALS

1 Fractures are common in childhood, due to high-level motor activity, developing coordination and the mechanical properties of the growing skeleton. Compared to adult mechanisms, the majority are relatively low-force injuries.

2 The patterns of bony disruption are completely different from adult fracture patterns and include buckle and greenstick fractures and growth-plate injuries. Bony disruption/deformity is more common than ligamentous disruption: 'sprains' and ruptures are uncommon.

3 Displaced fractures are a common and highly traumatic event for children and rapid attention to physical and psychological distress can minimise the effects of this trauma.

4 Certain missed fractures have a high propensity for serious long-term functional morbidity and must be actively sought. These include elbow injuries, such as lateral condylar and Monteggia-type fractures.

Fracture patterns in childhood

Recent studies in North America and Australia[1–3] demonstrated gaps in knowledge, practice and educational resources relating to acute paediatric fracture management. This is concerning given that fractures in children account for a significant proportion of emergency department (ED) discharge diagnoses (7% at one paediatric institution in 2016)[4] and that mismanagement of fractures

is a common reason for medical litigation.[5]

Fig. 25.3.1 shows the frequency of common fractures presenting to a children's ED during 2016.[4]

Within different age subgroups, the distribution varies thus:

- Infants/toddlers – higher proportions femur and skull fractures
- Primary school – higher proportion of elbow-region fractures, especially supracondylar
- Adolescents – complex ankle fractures; higher force sporting injuries; transition to adult pattern with increasing ligamentous injuries, e.g. elbow dislocations.

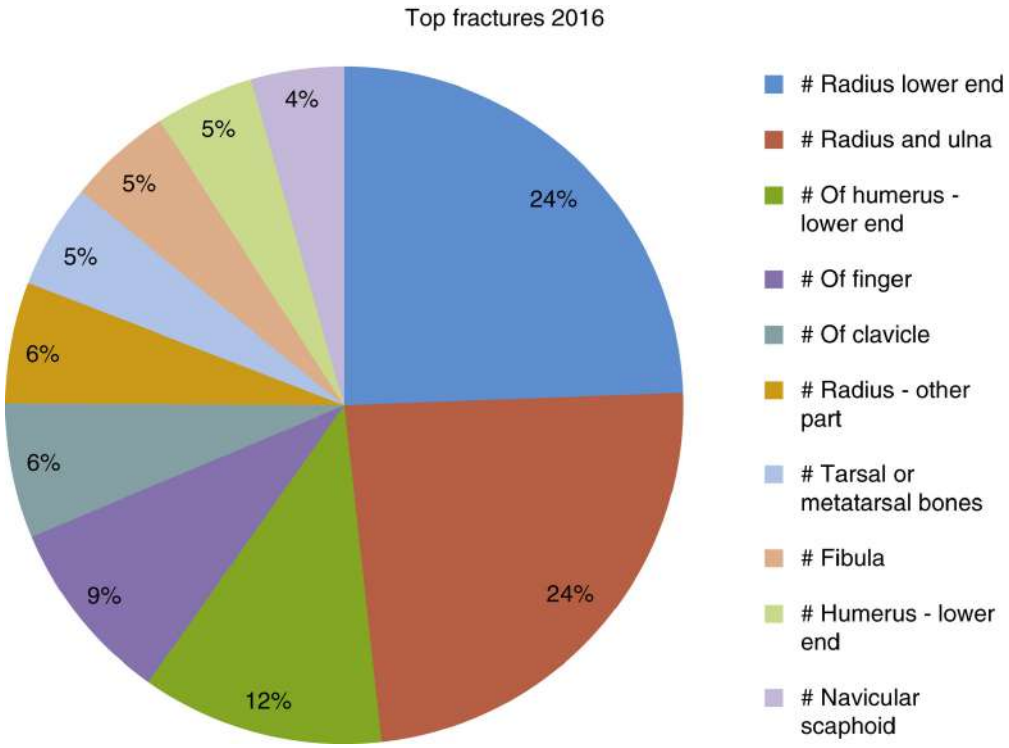


FIG. 25.3.1 Frequency of fracture types at Lady Cilento Children's Hospital emergency department.

The majority of ED paediatric fracture presentations occur at the distal radius and ulna. This is one of the top ten ED diagnoses for children in Australia. Many displaced forearm fractures can be reduced under sedation by emergency staff with appropriate training and follow-up, making this a most valuable area of expertise.[6]

Paediatric limb fractures, depending on the angle of force to which they have been subjected, can occur to the shaft, metaphysis or physeal region. The different quality of developing bone means that even injuries to the shaft and metaphysis tend to have different patterns of deformation from those seen in adults, including ‘torus’ or buckle injuries, bowing, and greenstick fractures. The importance of this awareness for the emergency physician is best illustrated by the Monteggia equivalent injury in which ‘shortening’ from proximal radial dislocation is ‘matched’ by ulnar bowing. The resultant injury has no radiologically obvious ‘fracture’ in the traditional sense but has serious consequences if not recognised and reduced (Fig. 25.3.2).

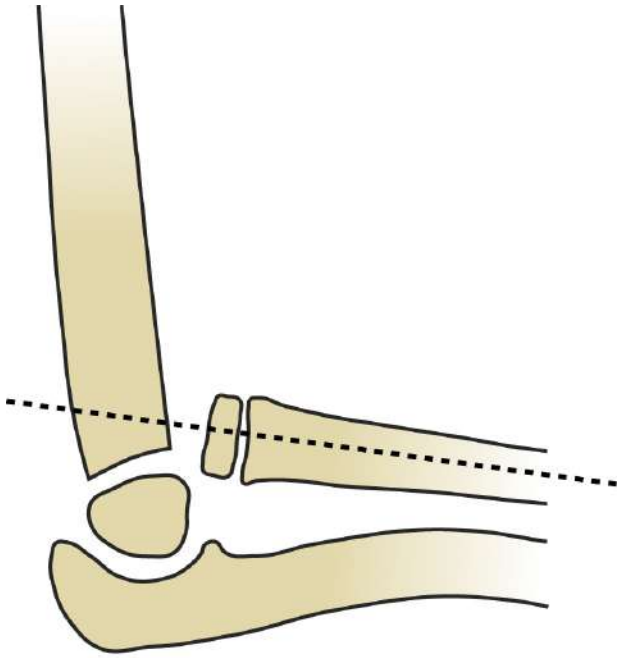
Table 25.3.1 shows some examples of how the same mechanism of injury will cause different outcomes in children and adults. This table illustrates the maxim that children tend to fracture rather than ‘sprain’, as the physis is the weakest point of the musculoskeletal continuum. That is, a ligament will avulse its bony origin or insertion rather than tearing. In some cases, this is to the child’s advantage, as the cellular architecture of developing bone, which contributes to its mechanical weakness, also contributes to rapid healing and extensive remodelling. A midshaft femoral fracture, for example, will heal in 2–3 weeks in an infant, whereas the same disruption will take 12 weeks to union in a teenager.

The Salter–Harris classification (Fig. 25.3.3) remains the most useful way of describing the pattern of cleavage with respect to the physis. In reality, types 1 and 5 represent mechanical force patterns (separation and compression) rather than a radiological pattern as, unless there is lateral translation or adjacent bony or soft tissue deformation, the physis may appear radiologically normal in these injuries.

An example of Salter–Harris type 1 injuries with lateral shift is the so-called ‘slipped distal radial epiphysis’ (Fig.25.3.4). The disorder of slipped upper femoral epiphysis (SUFE) has been discussed in Chapter 25.2 as, although minor trauma may precipitate an acute slippage, the cleavage is due to an abnormal physeal predisposition and should not be looked upon as truly traumatic. Salter–Harris type 2 injuries are the most common physeal injury pattern seen, the metaphyseal corner ranging in size from a barely visible fragment to an extensive triangle. Injuries through the epiphysis itself, Salter–Harris types 3 and 4, are more worrying in their prognosis because they are intra-articular as well as involving the physis. The classic example of a Salter–Harris type 3 injury is the Tillaux fracture (Fig. 25.3.5), while lateral condylar fractures at the elbow are Salter–Harris 4 in type.

Initial assessment and management

The initial assessment of the paediatric isolated limb injury (fracture/dislocation) is shown in Box 25.3.1 and the neurovascular assessment in Box 25.3.2. Limb injury must always be considered in the broader context of trauma. Primary and secondary survey, however brief and targeted, should always be carried out bearing in mind the described injury mechanism and the child's complaints of pain, so that any associated injuries, e.g. to head, abdomen or spine, may be recognised and evaluated early. An efficient early assessment should be able to quickly establish mechanism of injury and predict probable fracture type; identify possible other sites of injury; assess for the presence or absence of compound features and neurovascular impairment; organise pain relief, which may include splinting, before X-rays; consider the need for fasting; and give antibiotics and tetanus immunisation if required.



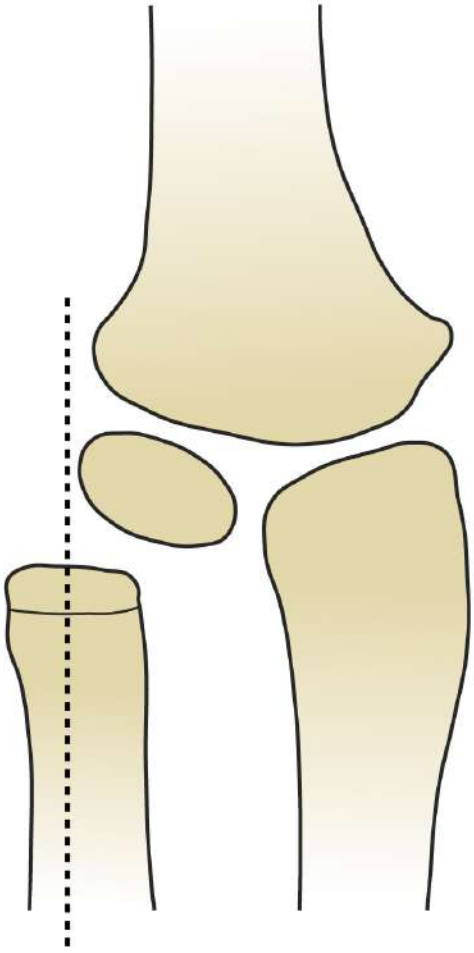


FIG. 25.3.2 Monteggia fracture-dislocation. Demonstration of the abnormal radio-capitellar relationship (see Fig. 25.3.8 for contrast). Drawing by Terry McGuire.

Fracture descriptions to the orthopaedic team should start with the child's age, mechanism of injury, and clinical findings, and proceed to a description of the X-ray, describing the part of the bone affected, type of fracture, extent of angulation and/or displacement and associated findings. Clinical findings must always be kept paramount. Skin breach must be actively sought and described, then photographed and covered with a sterile dressing. Prominently placed photographic displays of common paediatric fractures within the ED may aid accurate description.

Doctors share in the community responsibility for child safety. Within the ED setting this means getting a clear description of the setting and mechanism of injury, particularly with injuries to pre-verbal children. These data are important:

Table 25.3.1

Examples of paediatric versus adult outcomes of common fall mechanisms (different paediatric injuries occur at different ages depending on planes of weakness). The ligaments in children provide greater resistance to shear injury than the growing bone, so avulsion-type injuries occur in place of ligamentous tears or dislocations

Mechanism	Adult injury	Paediatric injury
Fall onto point of shoulder	Acromioclavicular separation	Lateral clavicular fracture
Shoulder extension/compression	Shoulder dislocation	Proximal humeral fracture
Fall on hand, elbow hyperextension	Elbow dislocation	Supracondylar/condylar fractures
Wrist hyperextension/compression	Scaphoid fracture	Distal forearm fracture
Fall onto hand	Colles' fracture	Midshaft, metaphyseal, or epiphyseal fracture
Thumb abduction 1	Bennet's fracture	Metaphyseal fracture base first metacarpal
Thumb abduction 2	Gamekeeper's thumb (ulnar collateral ligament [UCL])	UCL avulsion fracture (Salter–Harris type 3 proximal phalanx thumb)
Rotation of knee on lower leg	Anterior cruciate ligament, cartilage tear	Tibial spine fracture
Valgus/varus knee stress	Ligament, cartilage tear	Distal femoral physeal separation
Forceful jump (quadriceps)	Ligament tear	Patellar tendon avulsion fracture (tibial

		tubercle) fracture
Forceful jump (calf)	Achilles tendon tear	Calcaneal avulsion fracture
Rotation of tibia on calcaneus	Ankle sprains, Pott's fractures	Tibial spiral fracture, Tillaux fracture, triplane fracture
Inversion ankle	Talofibular ligament tear	Salter–Harris type 1 or 2 distal fibula

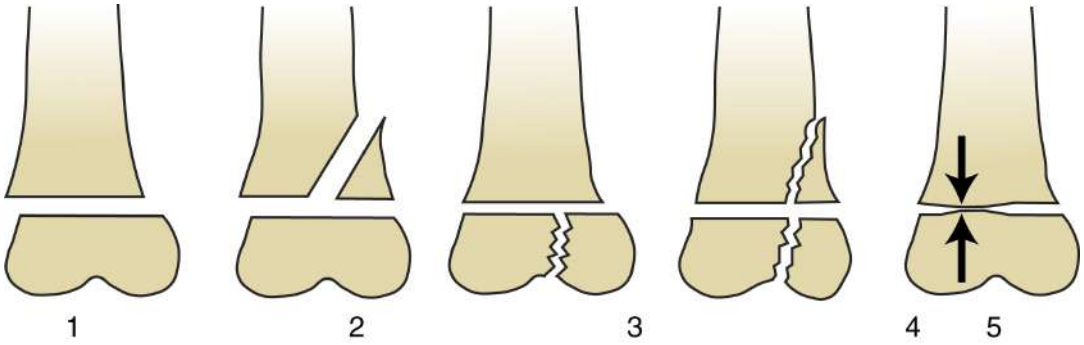


FIG. 25.3.3 Salter–Harris classification of epiphyseal fractures. Drawing by Terry McGuire.

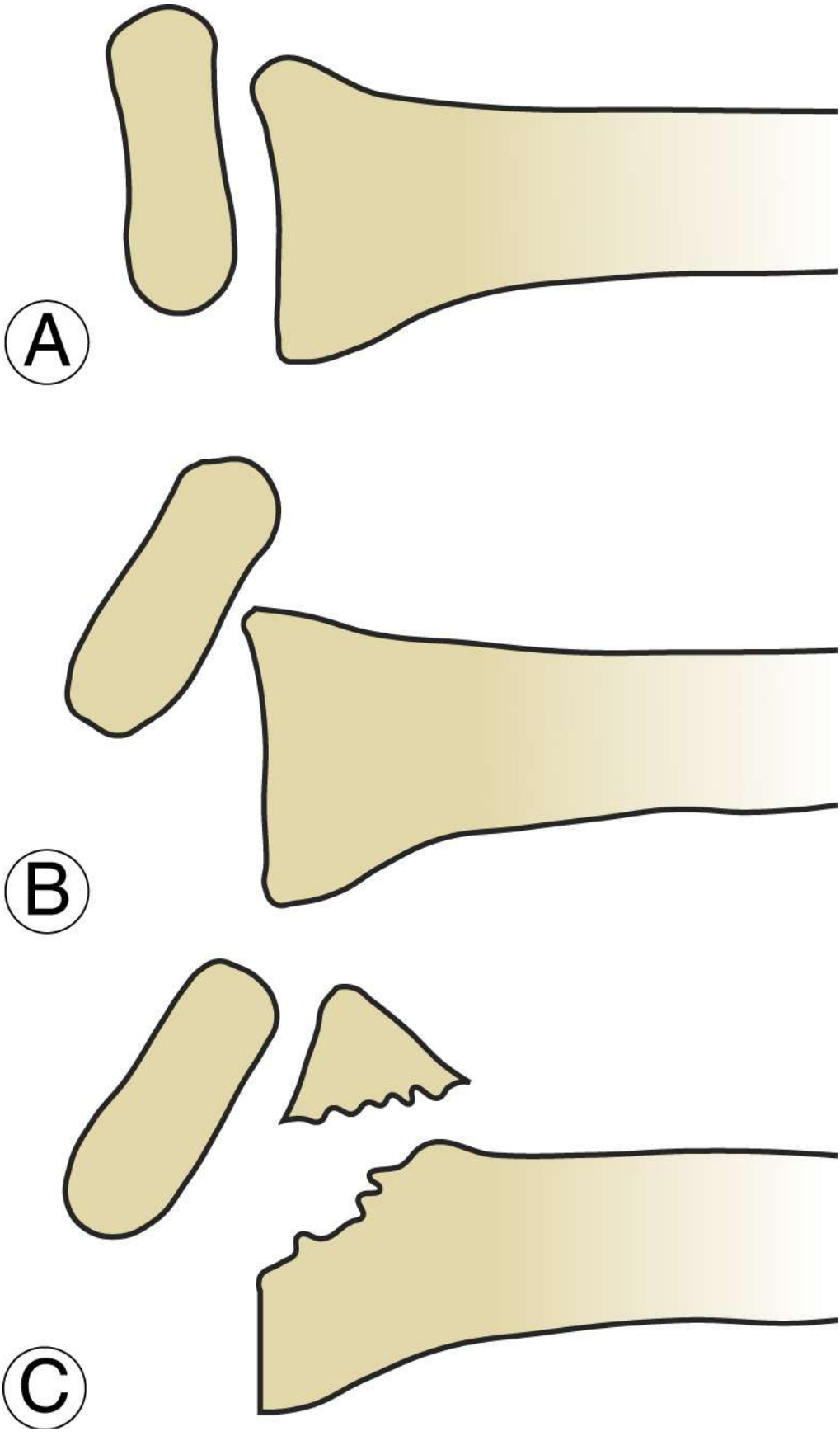

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FIG. 25.3.4 Common distal radial epiphyseal fractures. (A) Partial slipped distal radial epiphysis (Salter–Harris 1) with 10% translation; (B) slipped distal radial epiphysis (Salter–Harris 1) with 50% translation and probable intact dorsal periosteal sleeve; (C) Salter–Harris 2 fracture of distal radius with metaphyseal fragment angulated to 45 degrees and 50% translation of epiphyseal plate. Drawing by Terry McGuire.

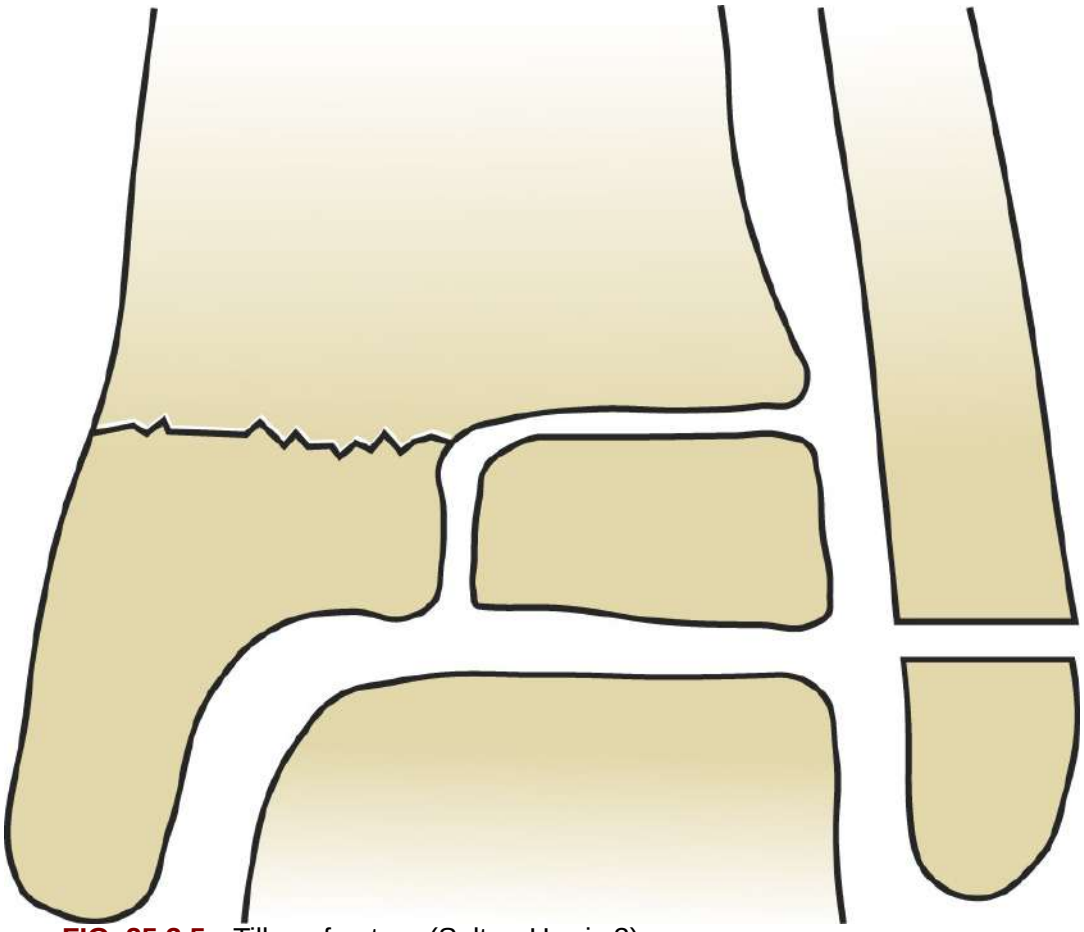

FIG. 25.3.5 Tillaux fracture (Salter–Harris 3).
Early fusion of the medial distal tibial physis and relative superior strength of the distal tibiofibular ligament cause shearing force to separate the central physeal region and travel along the lateral distal tibial physis. Sometimes a metaphyseal fragment is also cleaved (Salter–Harris 4).

- to more clearly anticipate associated injuries (e.g. foreign bodies)
- to gather cumulative injury prevention data to support legislative change (e.g. road access) and public health awareness campaigns

• to flag possible abusive injury, particularly in very young children.

In general, fractures in pre-verbal children without a clear, developmentally appropriate mechanism/history or with other concerning features, will need further assessment. Features suggestive of non-accidental injury are shown in Box 25.3.3, and child abuse is discussed in more detail in Chapter 18.2. Documentation of all findings, both positive and negative, is especially important in these cases. As a minimum, all fractures occurring in children under 12 months should be discussed with a paediatrician or child-protection specialist.[9,10]

Box 25.3.1 Initial assessment and management of traumatic limb deformity

1. ***Rapport:*** establish rapport and explain procedures to both child and parent.
2. ***Mechanism and associated dangers:*** rapidly ascertain mechanism and ensure primary survey stability and allergy potential.
3. ***Pain management:*** rapid pain relief in hospital is provided using intra-nasal fentanyl (1.5 mcg/kg) or inhaled nitrous oxide. These can be used prior to insertion of an intravenous (IV) line and may obviate the need for one in some situations. Providing more enduring analgesia with oral analgesics (paracetamol, ibuprofen and oxycodone) should be considered, even in those that are nil by mouth. Pain is not always managed to an acceptable standard, even in paediatric emergency departments.[1] This should be a focus for audit and education.
4. ***Assess for site of anatomical disruption:*** look at and gently palpate the injured limb to estimate probable anatomical site of disruption, e.g. lateral elbow, mid-shaft forearm, etc. (comparison with other limb is often helpful).
5. ***Check for associated neurovascular dysfunction:*** (see Box 25.3.2), document results of this examination, including when no deficit is found. Notify deficits immediately.
6. ***Check for any evidence of an open wound:*** if this is present, cover with a sterile dressing and commence appropriate antibiotics IV, e.g. cephazolin 50 mg kg^{-1} and notify orthopaedic team immediately. Compound fractures are deemed tetanus prone wounds and therefore

adequacy of tetanus immunisation should be assessed and a booster dose +/- tetanus immunoglobulin should be given if necessary.[7]

7. ***Immobilise limb:*** optimal pain relief for deformed limb fractures includes both splinting and analgesia.[8] Apply a plaster of Paris backslab after initial analgesia is given. This may sometimes obscure the radiological view of the fracture, particularly if the fracture is at the elbow, but achieving satisfactory analgesia is a higher priority.
8. ***Keep stomach empty:*** identify time of last ingestion and inform patient and parent about fasting.
9. ***Organise appropriate radiology.***
10. ***Discuss clinical and radiological findings with orthopaedic team.***

Box 25.3.2 Presence/absence of associated neurovascular injury

- Radial a:
  Pulse compared with other side
- Brachial a:
  Hand perfusion/capillary refill compared with other side
  Beware spasm or intimal shear in supracondylar injuries
- Radial n:
  Sensation = dorsum hand, action = dorsiflex wrist, extend fingers
  Displaced humeral shaft injury
- Median n:
  Sensation thenar eminence, action = opposition, flexion interphalangeal thumb
  Supracondylar # or elbow dislocation
- Ulnar n:
  Sensation hypothenar eminence, action = abduction, adduction fingers
  Elbow dislocation, supracondylar #
- Posterior interosseous n:
  Branch of radial n, action = finger extension

Monteggia #/dislocation

- Popliteal artery:

  Distal pulses compared with other side
  Knee dislocation, complex tibial plateau injuries, supracondylar femur fractures

- Common peroneal nerve:

  Footdrop
  Proximal fibula fractures

The following sections describe the mechanism, recognition and ED treatment of individual fractures in children.

Upper limb and shoulder girdle injuries

Clavicular fractures

These may occur at any age from a fall onto the shoulder or outstretched hand. Older children will present with a history of a fall and pain and swelling overlying the clavicle. Toddlers and infants may be brought for assessment because they are not using the arm so palpation along the clavicle is an important part of the assessment. Clavicle fractures are usually of the middle third, are usually greenstick type fractures, and are treated with a broad arm sling for 4–6 weeks. If the fracture is of the middle third, is undisplaced and the child is less than 12 years old, follow-up and repeat X-ray are generally not required.

Fractures of the lateral third of the clavicle that are undisplaced are still managed in a broad arm sling but should be followed up in a fracture clinic within a week of the injury.[11]

Displaced fractures of the lateral third should be discussed with the nearest on-call orthopaedic service because of the possibility of a lateral clavicle physeal fracture-separation.

Box 25.3.3 Features suggestive of possible non-accidental injury

Fractures

- Proximal humeral or humeral shaft fractures under 3 years
- Corner or ‘bucket-handle’ metaphyseal injuries
- Femoral fractures in infants less than 12 months
- Rib fractures
- Complex skull fractures
- Multiple fractures, especially different ages

Presentation features

- Delayed presentation
- Unwitnessed injury
- Recurrent fractures
- Unexplained soft tissue markings

Assessment

- Draw diagram of injury history as described by witness
- Examine child all over and plot weight
- Ascertain any previous history of burns or fractures
- Is the developmental level compatible with the explanation?
- Is the history adequate to explain the injury?

Refer

- All fractures in children under 12 months, and all fractures in children under 4 years in which there is an inadequate or questionable mechanism, should be discussed with a child-protection specialist. In the interim, supportive and non-judgemental care for child and caregiver must be maintained.

Fractures of the medial third of the clavicle are generally the result of direct trauma and thus may be associated with underlying pulmonary, neurovascular or cardiac injuries. These fractures should be discussed immediately with the nearest orthopaedic service.

Contact sports should be avoided for 8 weeks after a clavicular fracture because of the risk of refracture.

The *neonatal shoulder* may come to medical attention due to asymmetrical arm movement or swelling. Causes include birth injury with clavicular fracture or brachial plexus injury, proximal humeral physeal separation, and joint or bone infection. Senior orthopaedic involvement is essential for the diagnosis, and non-accidental injury must be considered. The prognosis for neonatal clavicular injuries is excellent with conservative management.

Shoulder dislocation

This is uncommon under 10 years. The adolescent anterior dislocation can be reduced by traction in the prone position or by gentle arm traction to a seated child against counter-traction with a sheeted thorax.

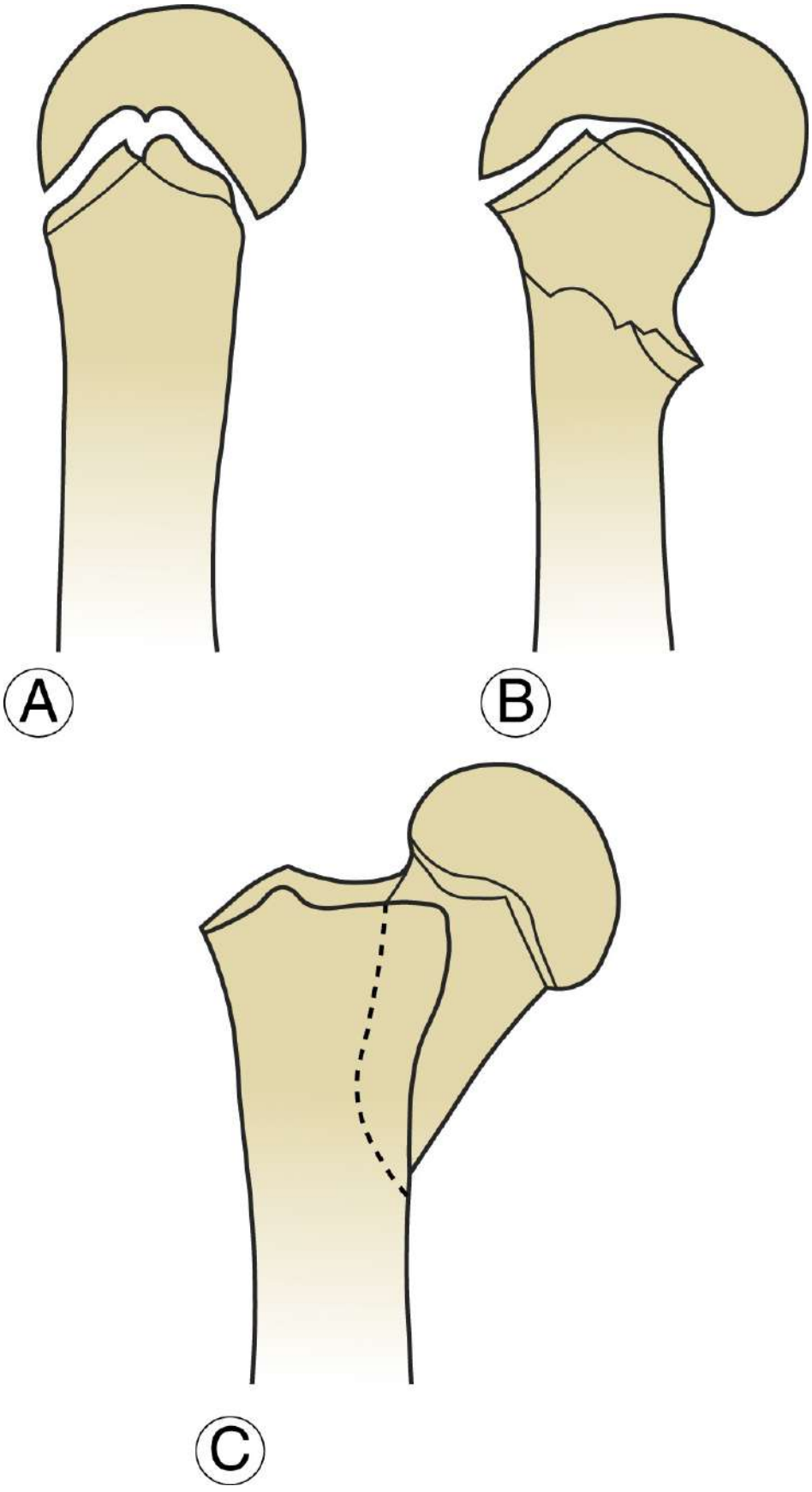
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FIG. 25.3.6 Proximal humeral fractures.
(A) Normal undulating physeal line (not a fracture); (B) greenstick metaphyseal fracture; (C) severely angulated and displaced Salter–Harris 2 fracture at the proximal humeral epiphysis. Due to the universal motion of the shoulder joint, this fracture will still unite and remodel completely with conservative treatment. Drawing by Terry McGuire.

Proximal humerus

These fractures vary from minor buckling at the proximal metaphysis, to proximal humeral epiphyseal Salter–Harris type 2 fracture-separations (Fig. 25.3.6). Because of the universal motion at the glenohumeral joint and the remodelling potential of children, a remarkable range of initial traumatic deformity is acceptable in children prior to physeal closure (age 14–16), including up to 50% displacement of the humeral head relative to the shaft displacement and up to 60 degrees of angulation. This fracture is managed by immobilising the shoulder with a sling or shoulder immobiliser.

Midshaft humeral fractures

These are less common and may be the result of direct blunt or, particularly in the case of spiral fractures of the humerus in toddlers, non-accidental injury. In older children, sometimes these fractures occur with minimal trauma through a bone cyst.

Ensure radial nerve function is checked and documented. Mostly these fractures are managed in a collar and cuff, sometimes with a U-slab plaster of Paris to achieve adequate reduction (axial alignment within 10 degrees).

Injuries to the elbow region

The elbow region accounts for about 10% of all paediatric fractures. Missed or inadequately treated paediatric elbow injuries figure prominently in orthopaedic litigation series[4] and so this topic should be included early on in orthopaedic education sessions to ED registrars and residents.[3]

When teaching the assessment of paediatric elbow injuries, it is useful to start with the examination and radiological findings in the normal elbow, as outlined in Table 25.3.5. The presence of completely normal elbow movement in flexion,

extension, supination and pronation excludes an injury. If elbow movement is not normal, clinical and radiological assessment is aimed at defining which part of the elbow joint is injured. Having an understanding of the order of appearance of ossification centres at the elbow, and the normal capitello-radial head relationship is essential (Figs 25.3.7 and 25.3.8).

Supracondylar fracture

Supracondylar injuries occur in the young school-age child as a result of a fall on the outstretched hand, transmitted through elbow hyperextension to the narrow region between olecranon and coronoid fossae.

Post-traumatic elbow effusion in childhood without a radiologically apparent fracture line most commonly represents an occult supracondylar fracture. These can be managed in a collar and cuff, with the elbow flexed to 90–100 degrees, for 3 weeks. Contact sport should be avoided for a further 3 weeks. No repeat imaging and no orthopaedic follow-up are required unless the elbow remains painful after 3 weeks.

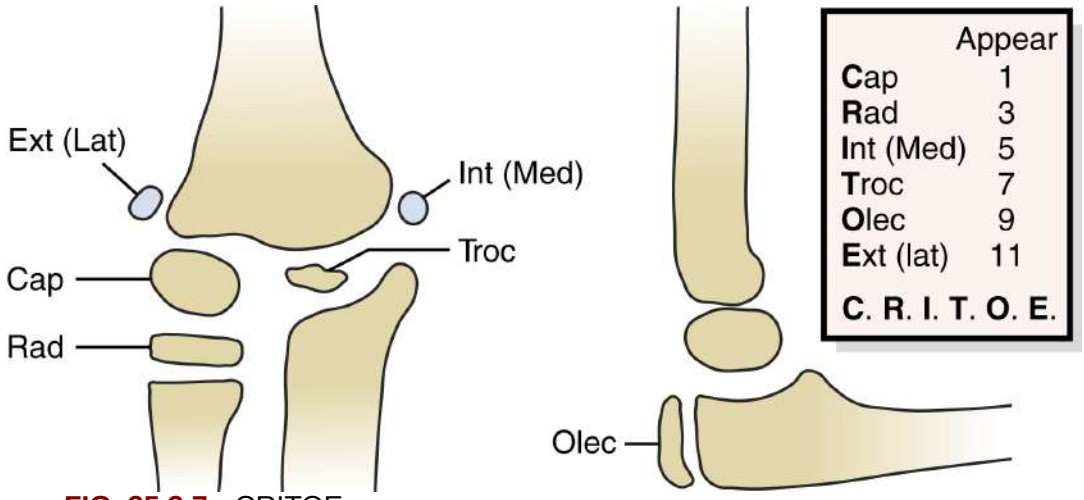

FIG. 25.3.7 CRITOE.
The approximate order of appearance of the six elbow ossification centres according to the CRITOE mnemonic. Drawing by Terry McGuire.

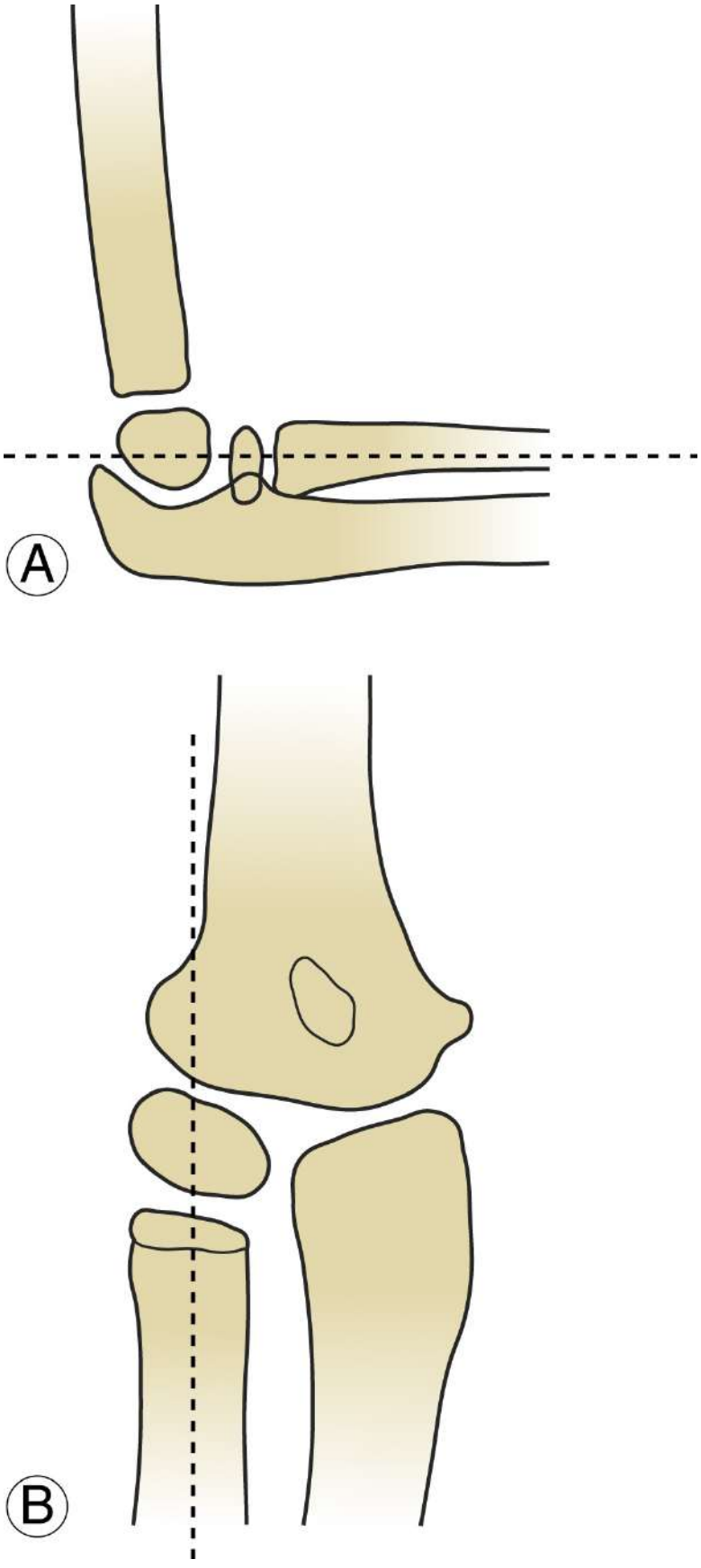
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FIG. 25.3.8 The normal capitello-radial head relationship. (A) the lateral view; (B) the AP view Drawing by Terry McGuire.

The Gartland classification system is used to describe the severity of displacement for extension-type supracondylar fractures, which account for over 98% of paediatric supracondylar fractures. To accurately classify these fractures it is important to obtain a true lateral X-ray of the elbow joint. In a normal elbow, or a non-displaced supracondylar fracture (Gartland grade 1), a line drawn on a lateral view along the anterior surface of the humerus should pass through the middle third of the capitellum. If it passes through the anterior third of the capitellum (Gartland grade 2), or misses the capitellum completely (Gartland grade 3), the fracture is displaced posteriorly.

Management of supracondylar fractures

Gartland type 1 – undisplaced supracondylar fracture

- Immobilisation in an above-elbow (from close to the axilla to the metacarpophalyngeal joints) backslab with 90 degrees elbow flexion for 3 weeks
- Cast removal and review by GP in 3 weeks; re-X-ray only if painful at this stage.

Gartland type 2 – posterior angulation with probable intact periosteal hinge (Fig. 25.3.9A)

It is important to check for associated rotation or varus/valgus injury. Use the ‘anterior humeral line’ as a guide to the degree of posterior angulation (Fig. 25.3.10), and consult orthopaedics about all injuries. Simple (Gartland 2a) fractures with less than 20 degrees of angulation may be managed conservatively in a backslab and collar and cuff, with orthopaedic follow-up. If there is varus/valgus angulation or rotation on the AP view (Gartland type 2b), then orthopaedic consultation is required acutely as surgery may be indicated. Remember that remodelling may correct some loss of flexion or extension but will not correct rotation or varus/valgus deformity.

Gartland type 3 – grossly displaced/rotated (Fig. 25.3.9B)

Check for open injury or neurovascular compromise. Brachial artery spasm or kinking is common with this injury, and the gross associated swelling may predispose to compartment syndrome. Radial pulse and hand perfusion should be continuously reassessed. In cases with extreme swelling, splinting in extension may be safest. Immediate orthopaedic notification is required.

If orthopaedic help is not available within 1 hour of the onset of poor hand perfusion, attempt gentle traction and reduction under procedural sedation, e.g. with ketamine, aiming for the position with best hand perfusion.

Median or radial nerve injury may also occur (usually as praxis) and require orthopaedic evaluation. Ulnar nerve injury is most commonly reported as an iatrogenic injury following internal fixation.

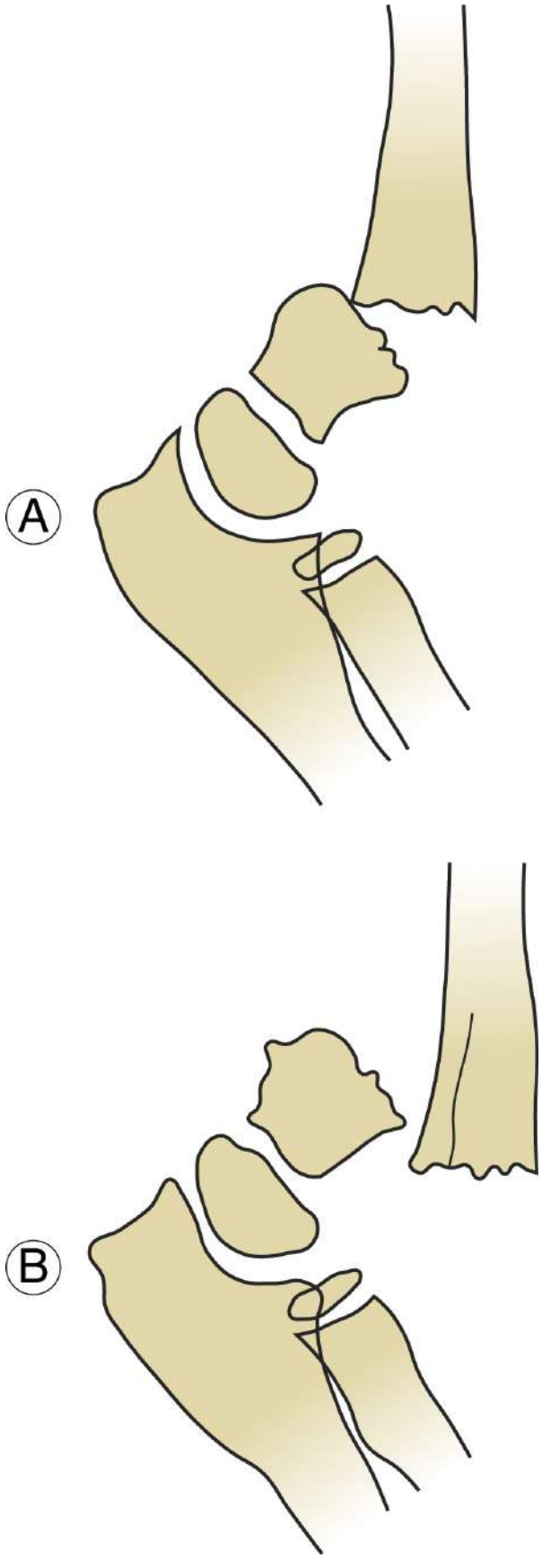
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FIG. 25.3.9 Supracondylar fractures.
(A) Grade Gartland type 2, approximately 45 degrees dorsal angulation but probable intact dorsal periosteal ‘hinge’; (B) grade Gartland type 3, complete displacement, often co-existent rotation and/or neurovascular impairment. Drawing by Terry McGuire.

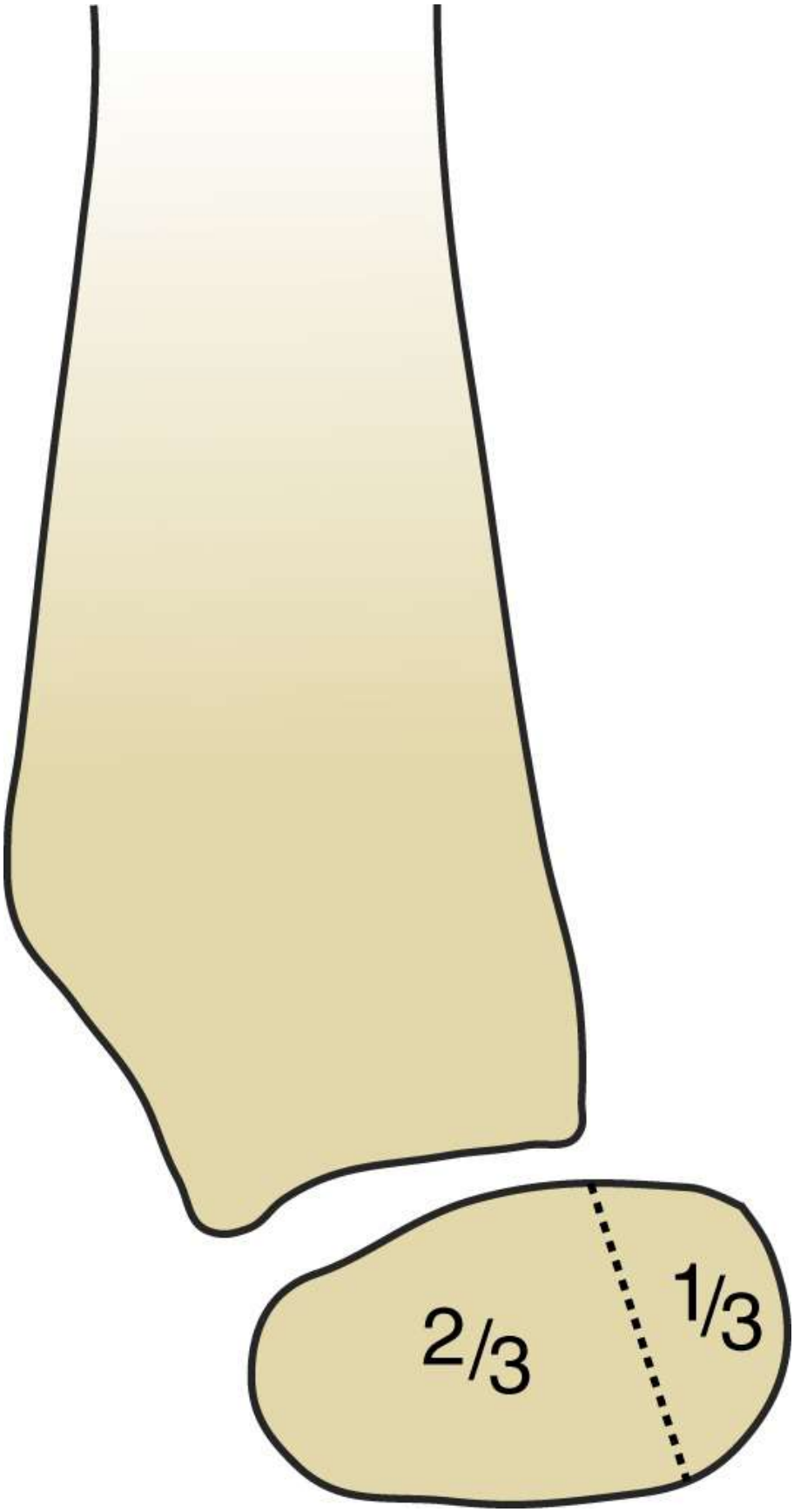
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FIG. 25.3.10 The anterior humeral line rule for subtle supracondylar fractures.
A line passed along the anterior humeral cortex on a lateral elbow radiograph should bisect the anterior and middle thirds of the capitellum. If it passes anterior to the capitellum, there is at least 20 degrees dorsal angulation of the distal humerus. Drawing by Terry McGuire.

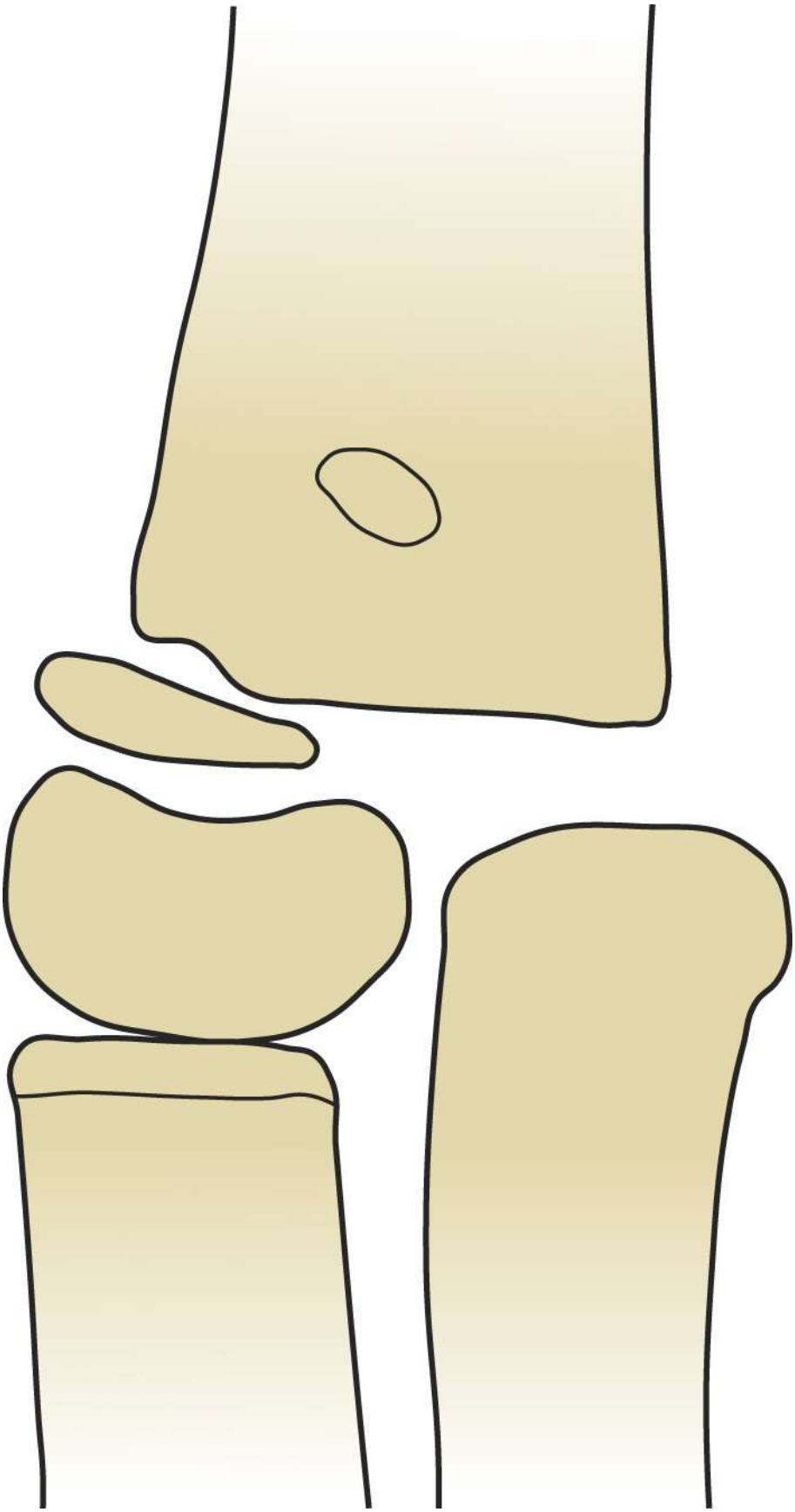


FIG. 25.3.11 Subtle lateral condylar fracture in a toddler. Note only two ossification centres (capitellum and radial head) and thin rim of metaphysis shorn away with capitellum. Greater angulation/displacement of metaphyseal fragment may be shown on lateral view. Gross clinical swelling is the key. Drawing by Terry McGuire.

Supracondylar Gartland types 2b and 3 fractures require admission for manipulation under anaesthesia and K-wiring and occasionally open reduction, and appropriate management of complications.

Intercondylar (T-condylar) fracture

This is a variant of the supracondylar fracture occurring in adolescents. It results from axial impaction and intra-articular separation of capitellum and trochlea, as well as proximal disruption of the medial and lateral distal humeral columns. Treatment is by open reduction with internal fixation.

Lateral condyle

This fracture results from a varus force on the supinated forearm, avulsing the condyle (Figs 25.3.11 and 25.3.12). There is clinical swelling and tenderness, which is maximal over the lateral condyle. It is usually a Salter–Harris type 4 fracture, but the late appearance of the trochlear and lateral epicondylar ossification centres means that the true structural disruption is not demonstrated by radiology, and therefore not appreciated by emergency staff, particularly in the younger child. The varus angulating force characteristically causes disruption commencing above the lateral condyle, passing to a varying extent along the physis, and in complete disruptions exiting either lateral (in the majority of cases; Milch type 1) or medial (Milch type 2) to the capitellar-trochlear groove. If uncorrected, the injury may result in valgus deformity and possible delayed ulnar nerve palsy and degenerative elbow disease.

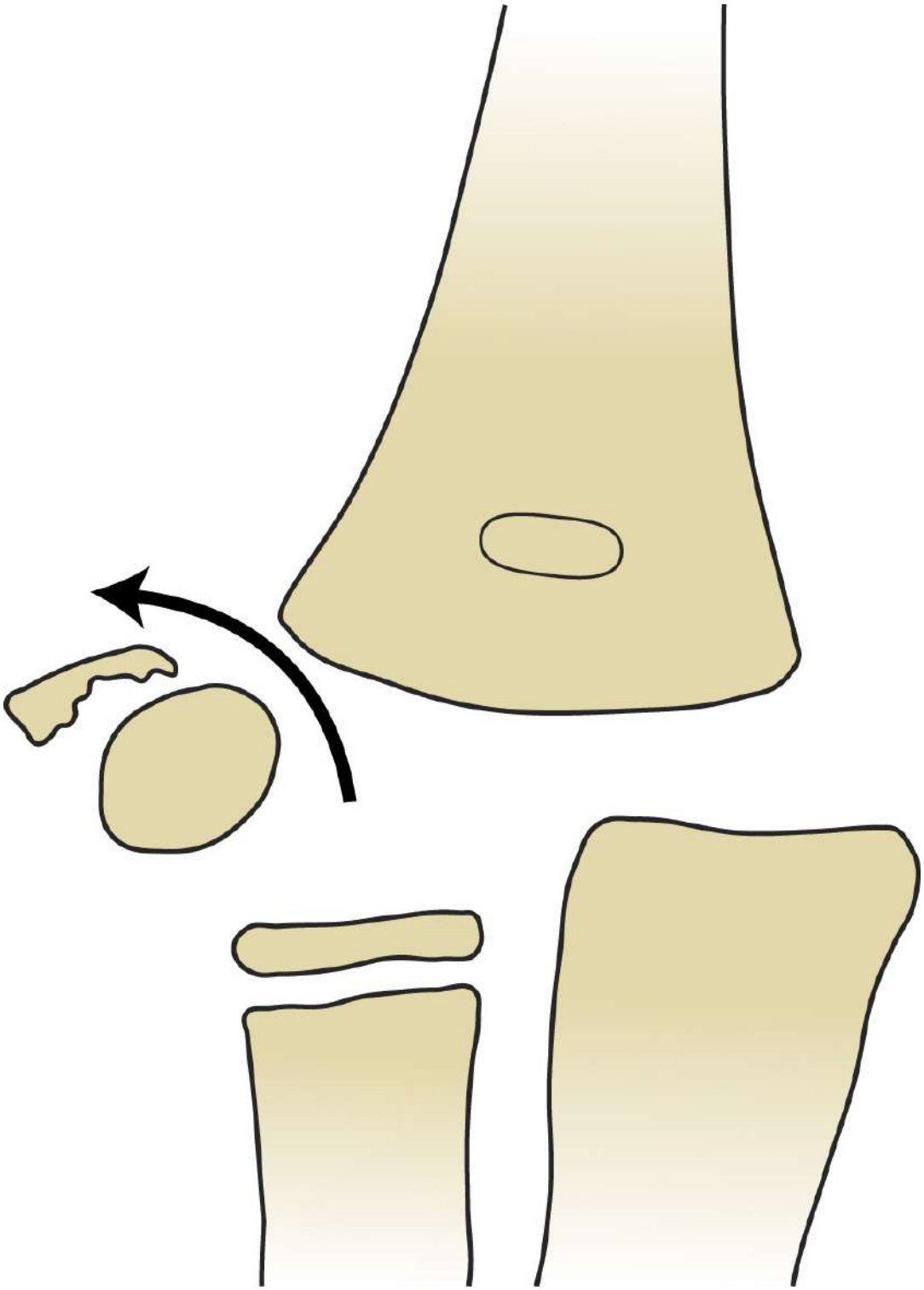


FIG. 25.3.12 Displaced and rotated lateral condylar fracture (Milch type I). Long-term complications may ensue if not surgically fixed. Drawing by Terry McGuire.

Bony displacement is often best seen on the lateral X-ray.

The clinical significance of this fracture means that:

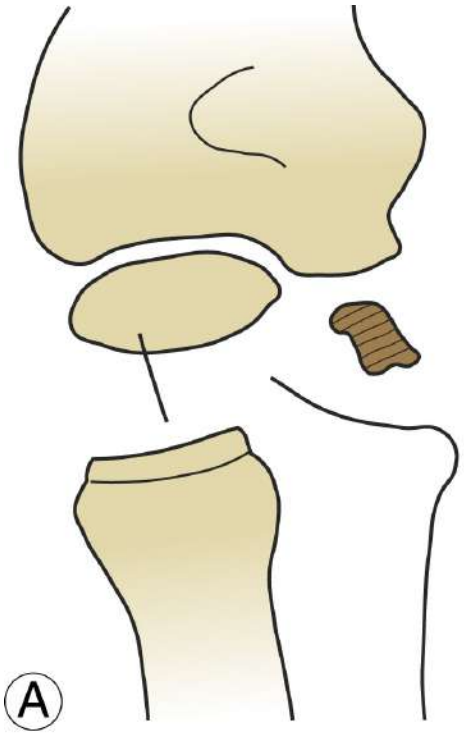
1. Any lateral condyle fracture with a greater than 2 mm separation of fracture segments is likely to require internal fixation, particularly if displacement is proximal.
2. All young children with major elbow deformity/swelling as a result of injury should be assessed early by experienced orthopaedic personnel. Ultrasound, CT and magnetic resonance imaging may all have a role to play in determining the line of injury and consequent best means of fixation. Within the ED, an internal oblique radiograph can be helpful.

The elbow may be supported in a backslab while awaiting orthopaedic review, but X-rays in this radiologically complex region are best performed prior to plaster application, after adequate analgesia.

In infants, lateral humeral condylar separation may occur as a Salter–Harris type 1 fracture and be difficult to diagnose radiologically, although the elbow will be grossly abnormal with maximal swelling laterally. History must explain the varus force and abusive injury should be considered. Ultrasound may be useful diagnostically.

Medial epicondylar avulsion

This may occur in association with other disruptions, e.g. elbow dislocation, or as a discrete event (Fig. 25.3.13). The medial epicondyle is the origin of the common flexor tendon and ossifies at approximately age 6. It will generally reunite readily with the humerus if it lies within 5 mm, unless there is interposing tissue. Occasionally, particularly when the avulsion has occurred in association with a posterior elbow dislocation, the epicondyle and its attachments may become lodged within the elbow joint and may block an attempt at closed reduction. Ulnar nerve injury is a common association. This circumstance is one of the main practical uses of knowledge of elbow ossification centres (see Fig. 25.3.7). These must be systematically reviewed on every elbow X-ray so that missing or misplaced opacities may be identified.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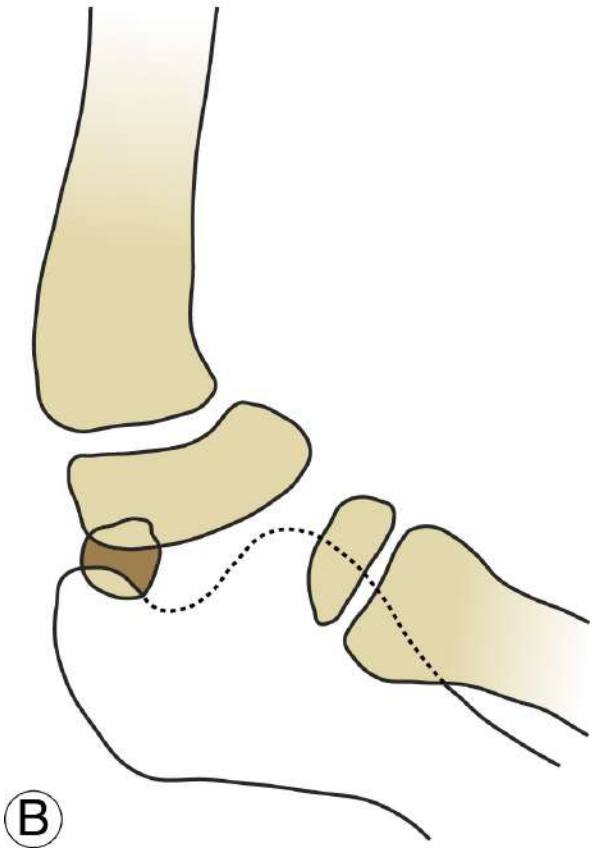
B

FIG. 25.3.13 Medial epicondylar avulsion.
(A) The opacity below the humerus is the avulsed medial epicondyle, which should be sitting more proximal and medial. There should not be a trochlear opacity without a medial epicondylar ossification centre. Developmentally, there should not be an ossification centre in the trochlea position without one in the medial epicondylar position (see Fig. 25.3.7). (B) Lateral view shows opacity 'between' capitellum and olecranon, possibly intra-articular. Drawing by Terry McGuire.

Pulled elbow (radial head subluxation)

Children from age 6 months presenting with acute non-use of one arm, which they hold in a semi-flexed and pronated posture, and a history of traction, can be presumed to have pulled elbow, or radial head subluxation (RHS), if there is point tenderness at the radial head and no palpable elbow effusion, when compared with the other arm. In up to 50% of cases there may not be a history of traction, especially in infants where rolling in bed with one arm fixed under their body appears to be a possible mechanism.

Subluxation occurs because the oval shape of the radial head allows the head to sublux slightly through the annular ligament when the forearm is pulled in pronation. Part of the ligament is 'caught' in the radiocapitellar space preventing normal range of movement at the elbow. In older children the ligament is thicker and more densely attached, and thus subluxation in a child over 6 is unusual.

X-ray and/or ultrasound, seeking alternative diagnoses, should be obtained in any child with other points of focal tenderness, an elbow effusion, a mechanism of greater trauma, an atypical history, e.g. fever, or a failure of the procedure detailed below. The differential diagnoses include septic arthritis/osteomyelitis, fracture or, occasionally, a neurological cause.

Reduction of radial head subluxation

A recent prospective randomised trial has suggested that hyperpronation is more likely than the 'traditional' method of supination and flexion to reduce the pulled elbow on the first occasion and appears to elicit less discomfort.[12]

After a brief parental explanation and oral or intranasal pain relief, these children should be held firmly by a parent while the forearm is hyperpronated. It is helpful for the doctor to cradle the elbow in the outer hand with the thumb over the radial head while their inner arm rotates. Success is usually denoted by

a momentary pain, a palpable click and a return to functional use. If the procedure is not successful, the procedure can be repeated, and if this fails, the traditional method of full, firm supination and flexion can be attempted. This combination of techniques should elicit success in >90% of cases of radial head subluxation. If the above process is unsuccessful, the history and examination should be revisited and imaging sought. Interestingly, while radiographs should be normal (and should *not* show posterior fat pad elevation, which should suggest alternative diagnoses), the radiocapitellar distance is significantly increased in radial head subluxation on ultrasound due to the presence of the interposed ligament. Point-of-care ultrasound may become more widely used to 'rule in' a pulled elbow as the 'hook sign' appears to be a consistent diagnostic finding.[13]

If an alternative diagnosis is not suggested by imaging and careful re-assessment, RHS remains the most likely diagnosis: the child can be allowed home with the arm supported in a sling in a neutral position, and with review at 24–48 hours, at which time many will have spontaneously reduced. Persistent dysfunction beyond 48 hours requires orthopaedic evaluation. Although some children sustain recurrent RHS, it rarely requires operative intervention.

Elbow dislocation

Appearing first in adolescence, this injury, the result of a fall on to the hand with partially flexed elbow, is uncommon in young children (who sustain supracondylar fractures instead). The majority dislocate posteriorly, tearing joint capsule and stretching soft tissues. The displacement may cause fracture to the coronoid process of the ulna or radial neck, or the medial epicondyle may be avulsed and subsequently entrapped in the joint after reduction. Neuropraxis of median or ulnar nerves may occur.

The dislocation should be suspected clinically. After assessing for associated injuries, it should be reduced in the ED, generally under ketamine anaesthesia (Fig. 25.3.14). Gentle downwards traction can be applied to the supinated proximal forearm, with extension of the elbow to about 135 degrees, against countertraction to distal humerus. Full extension should be avoided as it may cause further damage to the ulnar nerve. If there is difficulty in repositioning the olecranon, there may be soft tissue interposition and orthopaedic help should be sought, as a CT scan may be indicated. The reduced elbow should be held in flexion with a posterior splint, a check X-ray performed, and orthopaedic follow-

up arranged.

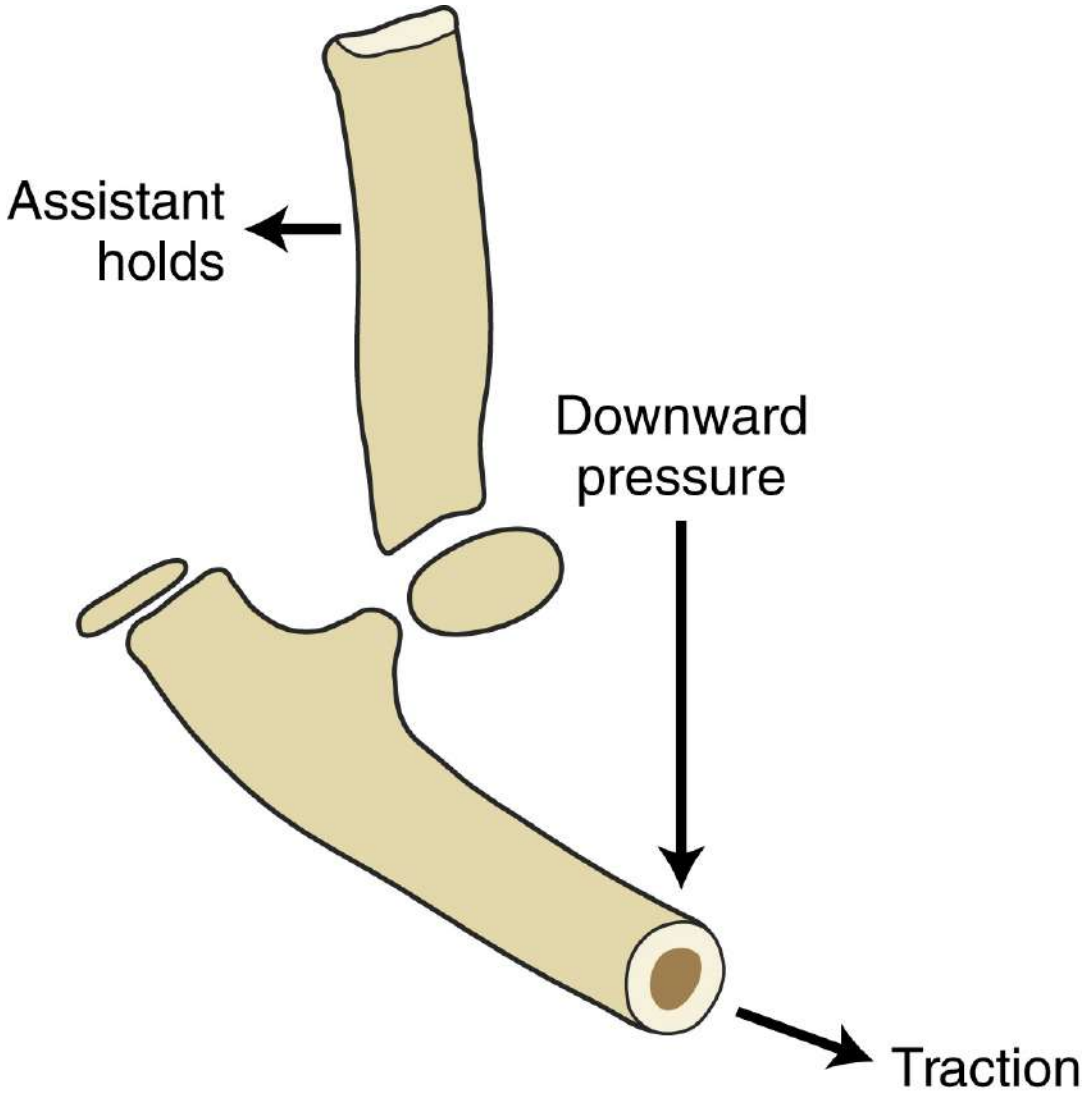

FIG. 25.3.14 Reduction of elbow dislocation.
Assistant anchors humerus to avoid distal/anterior movement. Bring forearm in full supination to approximately 20 degrees from full extension, and provide simultaneous downwards traction and downwards pressure over the proximal forearm to lever the coronoid process back under the distal humerus. Drawing by Terry McGuire.

Proximal radial and ulnar fractures

Olecranon fractures

These may occur in older children as: (1) avulsion (flexion) fractures (generally

requiring internal fixation); (2) extension fractures with intra-articular opening (may be stable in flexion); or (3) comminuted fractures from a direct blow to the elbow. Clinical correlation must be sought in diagnosing olecranon fractures, as the ossification centre, which appears around 10 years of age and may be bipartite or fragmentary in appearance, is easily mistaken for a fracture.

Radial neck fractures

These are relatively common injuries in the paediatric population and usually result from a fall onto an outstretched hand. The radial head itself is largely cartilaginous and rarely injured. Injuries range from subtle torus type ‘beaking’ of the neck, which is best seen on the lateral side on the lateral projection, to displaced Salter–Harris type 1 or 2 fractures. Suspicion of an isolated injury may be made by the presence of localised tenderness at the radial head. Neurovascular injury, particularly to posterior inter-osseous nerve (finger extension), should always be sought. Undisplaced fractures and those with up to 50–60% displacement do well with conservative treatment and may be immobilised in a collar and cuff with orthopaedic follow-up.

Monteggia fracture dislocation

The peak incidence of this injury complex is in the 4–10-year age group. Although, classically, radio-capitellar disruption accompanies an angulated or shortened ulnar fracture, the displacement may occur in association with any displacement to the radioulnar loop. Because of the significant complications of a missed radio-capitellar disruption, this lesion must be actively clinically and radiologically sought in any child with a forearm fracture and elbow swelling. Most units wisely adopt a rule of always X-raying the elbow of any child with forearm or wrist injury, unless the elbow has been specifically clinically cleared (see Box 25.3.4). Junior staff should be made aware of this requirement.

The classical (type 1) Monteggia lesion involves an angulated apex-volar ulnar shaft fracture and anterior displacement of the radial head (see Fig. 25.3.2). Variants include lateral radial head displacement, often in association with proximal ulnar/olecranon fractures (type 3), ‘Monteggia equivalent’ fractures including proximal radial fracture/dislocation, and others. A bowing deformity may be the only evidence of ulnar fracture. Orthopaedic manipulation of the radial head into its articulation is always required.

Midshaft radial and ulnar fractures

These injuries fall into two broad groups: (1) common low-energy greenstick fractures as a common consequence of falls in childhood; and (2) higher-energy complete fractures, which may be difficult to reduce, requiring internal fixation in 5–10% of cases. In the former group, with the exception of bowing fractures (which generally require general anaesthetic/orthopaedic reduction, as prolonged corrective force is necessary), ED reduction may be possible thus:

- apex volar – pronate forearm and apply wrist traction and volar pressure
- apex dorsum – supinate forearm and apply wrist traction and dorsal pressure.

Although these fractures may be simple to relocate using procedural sedation, e.g. with ketamine, their inherent instability makes expert three-point moulding essential. Therefore, manipulation should not be attempted unless such expertise and assistance is available, and follow-up within 1 week is imperative in case of subsequent loss of position. There is ongoing debate within the paediatric emergency medicine community about whether a backslab provides adequate stability post reduction.

Distal radial and ulnar fractures

These common injuries may be metaphyseal or epiphyseal in nature. Dorsal angulation occurs in 80% of cases, but radial or volar angulation/displacement of the distal fragments may also occur. Regarding management, the following guidelines apply:

- Simple *torus (buckle) fractures* with no cortical breach or angulation, which represent plastic deformation only, in children older than 3 years, may be placed in a forearm splint or brace for comfort and referred for follow-up in 1–2 weeks.[14,15]

Box 25.3.4 Requirements for ‘elbow clearance’

The normal paediatric elbow must have

- Full extension, supination and pronation
- No swelling or significant focal bony tenderness
- No abnormal anterior or posterior fat-pad sign on radiographs
- Normally placed and age-appropriate ossification centres (see Fig. 25.3.7)
- An intact radio-capitellar relationship (see Fig. 25.3.8).

- *Undisplaced greenstick fractures*, in which a cortex or periosteum has been breached, have inherent instability and the potential for further deformation. These should be managed in a well-moulded (three-point fixation) plaster in neutral position[16] and reviewed within 1 week.
- *Angulated or displaced greenstick fractures* of the distal forearm generally require reduction if the angulation is greater than 20 degrees, although remodelling potential varies with the age of the child and the distance of the fracture from the physis. Closed reduction may be performed in the ED if staffing and expertise permit.

Again, attention must be paid to ensure that a well-moulded plaster will maintain reduction, and follow-up orthopaedic review should be early enough to detect this and remanipulate if necessary, i.e. within 1 week. Early orthopaedic referral should occur for angulated isolated radial fractures and for fractures of radius and ulna with complete displacement and shortening, as a significant proportion of these manipulations will be problematic or require internal fixation.[16]

Distal radial epiphyseal fractures

These can generally be treated by manipulation and closed reduction in the same way as metaphyseal fractures. There is a very low incidence of subsequent premature physeal closure or other physeal disruption. This risk is greatest following multiple or delayed reduction attempts, or compressive injuries, or distal physeal separation of the ulna. Following manipulation, referral to fracture clinic must be within a week as repeat manipulation of the physis is contra-indicated after 7 days.

Carpal injuries: the scaphoid

Because of the flexibility of the paediatric wrist and the plastic properties of preossified bone, carpal injuries are very rare in children under 10 years. Scaphoid injuries are generally overdiagnosed in children in the ED setting. Of those fractures that do occur, 65% are distal pole and non-union is rare because of the different mechanical and vascular properties of immature bone. As children reach adolescence, their risk of adult-type scaphoid fractures increases. Scaphoid views (AP and oblique with attention to possible obliteration of the navicular fat pad) are suggested in older children if:[17–19]

- adolescent (10 years and over)
- high-velocity injury
- single-point tenderness and swelling over scaphoid both dorsally (in anatomical snuffbox) and on volar surface under base of first metacarpal (more specific finding)
- Kirk–Watson test (pain/clunk in scaphoid/scapholunar ligament on passive radial deviation of wrist)
- pain to compression along first metacarpal ray.

Suspected fractures should be managed with scaphoid plaster and orthopaedic follow-up as usual: conservative treatment usually allows resolution of true injuries but may take up to 6 months.

Metacarpal fractures

Crush injuries to metacarpal bones may occur and adolescence sees an increase in fractures of the head of the fifth metacarpal, as in adults, although the intact distal metacarpal physis will allow some remodelling to correct flexion loss.

Penetrating trauma, tendon damage, open injuries and neurovascular impairment must be identified and referred. Multiple fractures may create an unstable hand plate and finger flexion should be observed for possible associated rotational malalignment, which is not acceptable. Isolated metacarpal fractures will not create malrotation. Isolated metacarpal neck fractures should be treated with neighbour strapping and mobilisation.

Phalangeal fractures

Common paediatric phalangeal fractures include Salter–Harris 2 fractures at the

base of the first phalanx, which may cause radial/ulnar angulation and should be corrected by traction after a ring block. These may be radiologically subtle and require careful clinical evaluation. All open, intra-articular or oblique (unstable) fractures should be referred for orthopaedic evaluation.

Thumb fractures

Forced thumb abduction, e.g. from fall onto the splayed hand, can cause avulsion of part of the proximal thumb physis (a Salter–Harris type 3 injury instead of the adult ulnar collateral ligament tear) or a metaphyseal fracture of the base of the first metacarpal. The intra-articular Salter–Harris type 3 avulsion injury should be internally fixed, so referral is essential. However, in the metaphyseal injury, because of the universal motion of the first carpometacarpal joint, significant angulation and displacement will remodel if the child is under 10 years. The child may have the fracture immobilised in a scaphoid type plaster extending to the tip of the thumb and elevated and be referred for early orthopaedic consultation.

Fingertip injuries

These injuries to young children are extremely common from inadvertent closure in doors or gates, especially in cooler climates. Injuries include partial or complete amputation, nail-plate injuries and distal phalangeal fractures.

The classical crush injury includes a distal phalangeal tuft fracture and a partial amputation anteriorly through the nail bed, with intact volar soft tissue attachments and vascular integrity. Tip salvage is usual. However, without meticulous repair to the nail-bed laceration nail deformities are common, so referral to orthopaedics or plastic services is recommended.

Tip amputations distal to the terminal phalanx may often heal by secondary intent, because of the excellent vascular supply in childhood. More proximal injuries require assessment of nail-bed integrity, the possibility of soft tissue (e.g. nail-bed) interposition within an angulated/displaced fracture, or other indications for reconstructive procedures. Simple procedures may be performed in the ED under ketamine anaesthesia or sedation and ring block.

Lower limb and pelvis injuries

Pelvic fractures

These injuries are less common in paediatric than adult trauma. Avulsion injuries in athletic adolescents may occur. The implications for blood loss and urogenital injury of unstable pelvic fractures in children are similar to those in adulthood. The bladder is an intraabdominal organ in infants.

Hip dislocation

This is uncommon in childhood, occurring either with low force as a result of increased ligamentous laxity, or in the high-force mechanisms more typical of adult hip dislocation. The sequel of avascular necrosis is less common, occurring in only about 5% of cases. Reduction, by gentle closed longitudinal traction against a fixed pelvis, should be performed within 6 hours, ideally under general anaesthetic. Particular care must be taken in the adolescent in whom an occult physeal injury may be displaced. Post manipulation X-rays ± CT are indicated.

Femoral fractures

Although comprising less than 5% of paediatric fracture presentations to the ED, the infant or child presenting with a femoral fracture presents particular challenges to the emergency physician because of:

- the frequent association with major trauma and other unstable injuries
- the high level of pain and emotional distress for the child and the family, particularly in the setting of road trauma
- the need for procedural skills, such as femoral nerve block, and the application of traction splinting, such as the Thomas splint
- the possibility of non-accidental injury.[6]

The simultaneous management of all these challenging priorities is a good test of the mature, multidimensional emergency physician.

As has been mentioned earlier, all limb fractures should be initially approached with rapid primary and secondary survey while the integrity of airways, breathing, circulation, conscious state and spinal column are assessed, mechanism ascertained, and areas of tenderness identified. An intravenous (IV) cannula should be inserted immediately, under nitrous oxide or intra-nasal fentanyl if necessary. Early attention to issues of pain and anxiety has been

shown to reduce the stress and pain of later procedures. Femoral nerve block, preferably ultrasound guided, should be inserted early,[20] with Thomas splint immobilisation following in a timely fashion.

Not all femoral fractures are the result of major trauma. In the newly ambulant child, the torsion resulting from a change of forward momentum with the foot fixed at an angle may produce a spiral midshaft fracture. Mechanisms in abusive injury include forced external rotation or abduction, e.g. from nappy change position or direct blows.

Types of femoral fractures in children, and their orthopaedic implications, are as follows:

- Proximal femoral fractures include transepiphyseal, transcervical, basicervical and intertrochanteric fractures. These are serious injuries. Avascular necrosis and physeal growth arrest are significant potential complications of higher fractures, and early notification and reduction are urgent.
- Slipped upper femoral epiphysis (masquerading as trauma) is discussed in Chapter 25.2.
- In femoral shaft fractures, definitive orthopaedic management depends on the degree of precipitating force, associated injuries, the age of the child, home circumstances and the institution. Low-force injuries in young children may be treated by closed reduction and early hip spica casting. Options in older children usually involve internal or external fixation.

Injuries about the knee

Distal femoral physeal separation

These injuries result from high-force trauma to the knee. Any of the Salter–Harris pattern injuries may occur. Undisplaced injuries may be managed by cast immobilisation. Orthopaedic consultation is imperative because of:

- the possibility of co-existent intra-articular ligament disruption
- the need for perfect articular surface realignment
- the significant possibility of subsequent asymmetrical physeal arrest.

Tibial spine injury

Because of the mechanical properties of developing bone, twisting injuries to the paediatric knee result in avulsion of the tibial attachment of the anterior cruciate ligament (Fig. 25.3.15). The child presents non-weight-bearing, with a large effusion and joint-line tenderness. Although anteroposterior X-ray may be deceptive, lateral projections show a characteristic beak-like appearance of the superior surface of the tibial plateau, with posterior hinging. All children with large effusions should be referred for orthopaedic evaluation. Orthopaedic management involves an assessment, generally under anaesthetic, of the reducibility of the avulsed spine and the likelihood of, e.g. meniscal interposition. Treatment options include immobilisation in extension or partial flexion, and internal fixation. Some degree of subsequent instability and loss of extension may follow.

Avulsion of tibial tubercle

This adolescent injury represents avulsion of the insertion site of the patellar tendon, usually as a result of forceful quadriceps contraction against resistance. Displaced injuries require internal fixation and immobilisation.

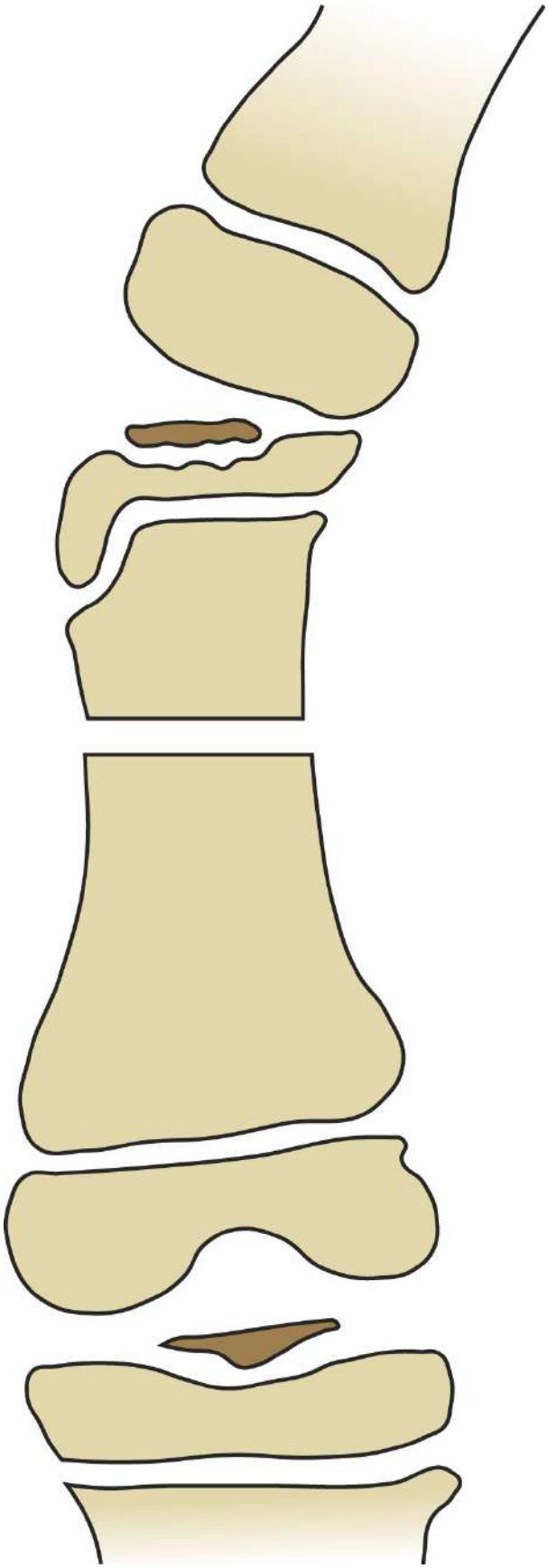

FIG. 25.3.15 Lateral and anteroposterior views of the paediatric knee with an avulsed tibial spine.
Due to the intact anterior cruciate ligament, the tibial spine fracture (shaded) may be hinged posteriorly, opening anteriorly on knee flexion. Drawing by Terry McGuire.

Patellar dislocation

This injury occurs most commonly in female adolescents with an excessive Q-angle (the complementary angle to the vastus lateralis/patellar tendon vectors), torsional anomalies, or hypermobility. The usual pattern is of lateral dislocation following a twist, e.g. during a fall. Most reduce spontaneously or during transport; extension of the knee may be facilitated in the prone position to relax hamstring muscles. A sky-line view should be obtained for best visualisation of osteochondral fragments from the medial patella. The knee should be placed in a Richards-type splint and the child referred for physiotherapy and orthopaedic follow-up. Chronic dislocation occurs in approximately 1:6 cases.

Patellar fractures

These are less common in children than in adults. Osteochondral avulsions may occur with dislocation. Displaced fractures, with tense haemarthroses and lack of full knee extension, should be referred to the orthopaedic team. The patellar sleeve fracture may be radiographically occult as it represents separation of the cartilaginous distal patella from the ossification centre. Findings will include patella alta and excessive anterior tenderness and swelling.

Bipartite patella (secondary ossification centre) is a common radiographic normal variant.

Lower leg fractures

Lower leg fractures are common in childhood. Factors to be considered in their assessment include:

- age
- mechanism (low- or high-force, type of injury, e.g. valgus/rotation)
- degree of angulation or displacement
- associated soft tissue damage or other injuries

- involvement of physes
- integrity of fibula.

Compound injuries and those with physeal involvement or significant angulation or displacement should be referred for inpatient orthopaedic evaluation. Proximal tibial epiphyseal injury may be complicated by vascular compromise, as with adult knee dislocation. Varus or valgus deformity, particularly at the proximal tibia, may progress. Stable, undisplaced or minimally displaced oblique or spiral shaft fractures of the tibia may be placed in a well-moulded above-knee cast with the knee flexed to 90 degrees and the ankle in 15 degrees of plantar flexion. Admission is not required if swelling is minimal, mechanism is clear and parents are sensible.

The so-called ‘toddler fracture’ is an undisplaced tibial shaft fracture that occurs as a result of a rotational shearing force in the newly ambulant child. Presentation is with non-weight-bearing or a limp and the differential includes other pathologies, e.g. irritable or septic hip. Tenderness should be localised to the tibial shaft but initial radiology may be normal. If this diagnosis is suspected in a well child with normal joint examination, plaster of Paris immobilisation and orthopaedic follow-up with repeat X-ray at 10 days scan may be helpful.

Note that some clinicians manage these fractures without immobilisation, given that these fractures are stable and that, in this age-group, the complications of casting are not insignificant.[21]

The other tibial fracture to be aware of is the so-called ‘trampoline fracture’, a transverse fracture of the proximal tibia metaphysis, commonly seen from the impact of a ‘double jump’ on a trampoline.[22] The radiological findings can be subtle and easily overlooked. It is therefore important to take a clear history of activity prior to the injury.

Controversies

1. Controversy exists surrounding boundary issues between emergency medicine and orthopaedics. Who performs which reduction should be determined by consideration of safe, effective resource use between emergency and orthopaedic services. Structured opportunities for interdepartmental teamwork help reduce angst and improve systems and service quality.
2. Length (SA vs. LA POP) and type (circumferential vs. three-quarter

slab) of plaster for unstable forearm fractures post reduction are controversial; however, accuracy of moulding and early review are probably more critical than either other variable.

Ankle fractures

As with adults, inversion, eversion and twisting mechanisms may cause a variety of injury patterns at the ankle depending on age, degree of force and mechanism. Ankle injuries requiring same-day orthopaedic consultation include:

- open injuries
- unstable injuries or ankles with extensive bilateral tenderness and swelling, suggesting possible mortice instability
- displaced or angulated distal tibial fractures
- Salter–Harris types 3 and 4 injuries
- Triplane and Tillaux fractures.

Tillaux fracture

This is a Salter–Harris type 3 fracture at the distal tibial physis, occurring in adolescence after partial closure of the medial growth plate (see Fig. 25.3.5). External rotation of the foot and ankle causes avulsion of the anterolateral portion of the distal tibial physis by its attachment to the fibula (anterior tibiofibular ligament). The diagnosis should be suspected in a non-weight-bearing adolescent with significant anterolateral ankle swelling and tenderness. Oblique ankle views or CT assessment may be required to detail alignment. These fractures must always be referred to an orthopaedic service.

Triplane fracture

This also occurs in the adolescent with external rotation injury, but the fracture line also tears off a section of posterior tibial metaphysis, which is clearly visible on the lateral radiograph.

Both of these transitional fractures may be complicated by subsequent joint incongruity or growth disturbance. Orthopaedic advice should always be sought.

Inversion injuries

Ligamentous rupture is rare in children. Carefully assess point of maximal tenderness. A point of maximal tenderness over the physis may represent a Salter–Harris type 1 separation of the distal fibular physis and should be cast immobilised for 2–3 weeks until stable. Bony injury may also occur at the base of the 5th metatarsal. Radiographically, this is shown by an avulsion fracture line at right angles to the metatarsal bones (the normal apophysis sits parallel and lateral). Although the Ottawa ankle rules have not been specifically validated in children, the requirement for further assessment of an injury involving extensive swelling, bony tenderness, or inability to weight bear seems appropriate. Different ankle rules have been applied in the paediatric setting and have reduced significantly the number of ankle X-rays done.[23]

Controversies

Paediatric fracture management interfaces the emergency practitioner with orthopaedic and physiotherapy teams, amongst whom management of such issues as growth plate injuries (including SUFE), and early mobilisation, may be controversial.

Conclusions

Paediatric fracture patterns are completely different from adult fracture patterns and include buckle and greenstick fractures and growth-plate injuries.

Sprains are uncommon as the physis or ligamentous insertions are the ‘weakest chains’.

Despite a great propensity for remodelling, unrecognised displacement/angulation may have serious and long-term implications.

Future Directions

The increasing trend to outpatient management of injuries will mean increasing numbers of paediatric fractures having definitive treatment in the emergency department. A thorough understanding with these fractures and challenges associated with their management is essential for all emergency physicians working in departments with paediatric patients.

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25.4

Risk management in acute paediatric orthopaedics

Sarah Martin

ESSENTIALS

1 Inadequate identification and management of unstable or displaced injuries such as lateral condylar elbow fractures, Monteggia fracture dislocations and slipped upper femoral epiphysis (SUFE) can cause serious long-term deformity and malfunction.

2 Guidelines and education sessions outlining expected approaches to analgesia (pharmacological and using splints) for acute fractures should be available in all departments as this aspect of fracture management is not always well managed.

3 Fracture immobilisation complications, from loss of position to skin ulceration or compartment syndrome, demand scrupulous departmental risk-management protocols.

4 The importance of adequate initial clinical evaluation and follow-up of patient or clinician concerns cannot be over-stressed.

5 A variety of departmental risk-management strategies provide a safety net for doctor and patient.

Introduction

Failure to diagnose a fracture accounts for up to 80% of emergency department (ED) diagnostic errors[1] and the medico-legal and clinical consequences of missed radiological abnormalities in the ED are well documented.[2] Although

much of the literature relates to adult EDs, the contributing factors, including high staff turnover, variable staff experience, 24-hour practice with multiple hand-overs, wide-ranging procedural demands and varying level of supervision of junior staff according to the time of day, can also be applied to paediatric emergency medicine. The factors that are unique to paediatric fracture identification include clinical assessment in the pre-verbal child and smaller bones with epiphyses meaning ‘normal’ changes with age.[3]

The solutions to these challenges provide exciting opportunities for teaching and training and audit within the ED.

Non-identification or delayed identification of paediatric fractures

There are many articles which discuss fractures missed in EDs because of radiological misinterpretation.[4–7] One study[3] done over a period of 3 years in a paediatric ED found that the most frequently missed fractures were of the hand phalanges, metatarsus, distal radius and tibia.

These articles do not describe the whole problem, however, as fractures can also be missed because of primary assessment failure (failure of thorough history or examination, therefore failure to X-ray). Primary assessment of pre-verbal children has some unique challenges and primary assessment of children will be affected by their developmental age and by the adequacy of analgesia provided to them. Additionally, X-rays may be inadequate in coverage or resolution and therefore not demonstrate the fracture. This tends to be more of an issue in paediatrics where there is a, quite valid, attempt to minimise exposure to radiation from X-rays.

Adverse events in acute paediatric orthopaedics

Adverse events in acute paediatric orthopaedics can be divided into five categories:

1. Missed displaced fractures/dislocations and complications thereof particularly:
    - supracondylar fractures and lateral condyle fractures at the elbow
    - Monteggia fracture-dislocations

- medial epicondyle entrapment after elbow dislocation
- open fractures
- compartment syndrome
- neurovascular injuries
- in multitrauma patients
- in children with complex medical conditions and/or developmental delay.

2. Missed undisplaced fractures especially:
    - buckle fractures, especially at radial neck and distal radius and ulna
    - toddler's fracture of tibia.
3. Delayed identification of less common causes of musculoskeletal pain such as:
    - septic arthritis and osteomyelitis
    - neoplastic infiltration of bone and bone marrow
    - congenital disorders.
4. Adequacy of analgesia:
5. several recent studies[8–11] have shown that pain from acute fractures in children is often under-managed, even in paediatric ED. Complications of fracture treatment such as:
    - plaster-related skin injuries
    - loss of position due to inadequate splinting post manipulation
    - malunion
    - adverse events associated with procedural sedation.

Risk management strategies include:

1. adequate training and supervision of staff:
    - credentialing of new staff for specific procedures, e.g. plastering, fracture reduction and procedural sedation
    - acute orthopaedics as a core part of resident and registrar training, with particular focus on the commonly missed and uniquely paediatric fractures[12]
2. easily accessible and comprehensive departmental information:[13]
    - online guidelines, preferably tailored to the local institution
    - online reference texts
    - 'how-to wall charts' with step-by-step guides to various plasters

    - paediatric fracture identification charts.
3. specific departmental approach to early identification of discordance between radiologist reports and ED diagnosis and treatment, such as mandating that all ED doctors record their X-ray findings in a place that can be seen by the reporting radiologist[14]
4. departmental audits the results of which then inform the content of teaching and training sessions. Audits should focus on:
    - X-ray interpretations
    - adequacy of analgesia provided
    - procedural sedation
    - plaster quality.

As ever, ensuring parents and families feel comfortable to return for review if they have any concerns is a critical aspect of discharge planning. Written information sheets about possible complications related to plaster of Paris casts and splints and about individual fractures optimises communication. And GPs must be included in any discharge communication.

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SECTION 26

Male Genitalia

OUTLINE

26.1

Male genitalia

Colin S. Kikiros

ESSENTIALS

1 A paediatric surgeon or urologist should be contacted to assess an acute scrotum so as not to miss a testicular torsion.

2 The testicles must always be examined when boys present with abdominal pain.

3 Testicular torsion is a condition requiring urgent surgery.

4 Colour Doppler ultrasound examination and nuclear scans of the testicle may be helpful, but should not be relied upon, in the diagnosis of testicular torsion. When in doubt, exploration of the testicle is by far the safest treatment.

The acute scrotum

Introduction

The acute scrotum is defined as a painful and/or enlarged scrotum and may be acute or subacute in onset. The origin of the pathology may be from the testis, the groin or the scrotal skin. In a younger child, the pain will cause the child to be unsettled with crying, and it may be intractable. In an older child, the reluctance to ambulate may be a predominant feature. In addition, referred pain from a sore testis may present as abdominal pain. Therefore in patients presenting with abdominal (especially lower-abdominal) pain one must examine the inguinal scrotal region so as not to overlook torsion of a testicle.

The diffuse vasculitis of Henoch–Schönlein purpura may affect the testis

and/or scrotum. Usually other manifestations of the condition such as a skin rash or haematuria are present. A radionuclide scan may assist in differentiating this condition from torsion of the testicle.

Torsion of a testicular or epididymal appendage

Appendages of the testis and epididymis occur in 90% of testicles. Torsion of these appendages is the most common cause of testicular pain. The most common age of presentation is at the onset of puberty, and this is thought to be due to the release of oestrogens and androgens from the male adrenal gland stimulating the appendages and causing them to enlarge. As the appendages are on a narrow stalk they have a tendency to twist.[1] Oedema of the appendages may also occur following trauma to the testis. Onset of the pain is usually gradual and the child is often able to ambulate without difficulty. Redness and swelling of the scrotum are also mild in the first 24 hours but may increase to an alarming degree in the following days and appearances can then be similar to those of testicular torsion. The scrotal swelling is often due to a small secondary hydrocele. The testis is usually normally aligned and in normal position in the scrotum. Tenderness is maximal at the upper pole of the testis (where the appendage is located), and a blue dot may be seen through the skin at the upper pole consistent with an infarcted appendage.

Torsion of the testis

This is an acute emergency and is due, in most situations, to medial rotation of the spermatic cord. However, in one-third of cases the testis rotates in a lateral direction.[2] The torsion usually occurs spontaneously. However, sometimes it can follow direct trauma to the testis. The pain is likely to be acute and severe and may be associated with nausea or vomiting. The older child is often reluctant to ambulate. Sometimes there is a history of previous short-lived pain in a testis consistent with intermittent episodes of spontaneously resolving torsion. The testis is usually enlarged and in a high position in the scrotum or even in the groin as torsion of the testis results in shortening of the cord. It is not usually in its normal lie, and there is much redness and swelling of the scrotal skin. Usually a secondary hydrocele is present. The contralateral testicle may lie in a bell-clapper fashion owing to the insertion of the epididymis in the central part of the testicle, and this in turn predisposes the testicle to undergo torsion. The child

should be taken to the operating theatre urgently, even if the child is not adequately fasted, as prolonged obstruction of the testicular vessels may lead to partial or complete atrophy of the testicle. Therefore nuclear scans and colour Doppler ultrasound should be avoided if the diagnosis is clear, as these investigations may result in unnecessary delay and they are not reliable. False negatives and false positives have been reported with both modalities. Colour Doppler ultrasonography may be misleading as intratesticular flow may be seen even in testicles that have undergone torsion.[3] Visualisation of a twist in the cord is more reliable.[4] Survival of the testicle will depend on the number of twists that the spermatic cord has undergone, along with the length of time that the cord has been twisted, with the prognosis being excellent for those undergoing surgery within 6 hours of the onset of symptoms.[5,6]

Epididymo-orchitis

Infection or inflammation may affect the epididymis or the testis. Infections in the epididymis may arise from retrograde flow along the vas deferens or lymphatics from urinary tract infections, or from the bloodstream. Inflammation of the testis may arise from conditions such as mumps. The epididymis is tender and swollen, and the testis also may be tender. The testis is of normal lie and in a normal position in the scrotum. A raised interleukin-6 level may be clinically helpful in assisting the diagnosis of epididymitis.[7] Once the diagnosis has been confirmed the child's urine should be sent for analysis and he should then be commenced on antibiotics. Enteric organisms are the usual cause of the urinary tract infection.[8] Once the condition has resolved renal ultrasound and micturating cystourethrogram should be performed as the urine infection may have resulted from an abnormality in the urinary tract, such as posterior urethral valves[9] or vesico-ureteric reflux.

Idiopathic scrotal oedema

The cause of this condition is unknown. The child presents with scrotal discomfort, oedema and erythema of one side of the scrotum, which may spread to affect the entire scrotum, the penis and inguinal and perineal regions. The testis is not swollen and is normally aligned, and the tenderness arises from the palpably thickened scrotal wall. Occasionally, eosinophilia may occur and, characteristically, ultrasound examination shows marked thickening of the

scrotal wall, increased peri-testicular blood flow and a mild reactive hydrocele.[10] The condition resolves in 1–4 days and no treatment, apart from pain relief, is required. Antibiotics and non-steroidal anti-inflammatory drugs are considered unnecessary.

Testicular tumours

Primary or secondary (leukaemia, for example) tumours may cause the testicle to enlarge and become painful. The onset is usually gradual and the pain more chronic. However, testicular tumours may present acutely when they have been subjected to trauma. The testicle may become extremely large, although the scrotal skin is not usually erythematous. Ultrasound and full blood picture may aid in the diagnosis. Paediatric surgeons and oncologists are predominantly required in managing these patients.[11]

Irreducible inguinal hernia

Segments of intestine may on occasion descend into a hernia sac in the scrotum and become irreducible. Acute pain may be felt in the scrotum and also in the groin (see Chapter 7.11 on herniae).

Rupture of the testis

This is usually the result of trauma. The testis becomes enlarged and painful, and there is associated bruising and frequently an associated hydrocele. Ultrasound examination often reveals irregularity of the testicular outline and intratesticular haematoma.[12] The patient should be referred to a surgeon for possible exploration and repair of the testicle.

Acute hydrocele

A patent processus vaginalus may allow intraperitoneal fluid to flow into the space around the testicle in the scrotum. Often the amount of fluid is minimal, and the patient does not present acutely. Occasionally, however, intercurrent illnesses, such as gastroenteritis and upper respiratory tract infections, may result in an increase in volume of the peritoneal fluid. This leads to an increase in the amount of fluid around the testis, and the patient may present to the ED with a large scrotal swelling. The testis in this situation is not painful and one can

determine that the swelling does not extend into the groin. Ultrasound examination is useful in confirming the diagnosis and unnecessary urgent surgery can be avoided.

Acute problems of the penis and foreskin

ESSENTIALS

1. Urinary retention may arise from phimosis or balanitis and requires urgent urinary diversion or circumcision.
2. Priapism is an acute emergency and requires urgent treatment.

Introduction

Currently, approximately 90% of young males in Australia are not circumcised. This compares to a generation ago, when the majority of males were circumcised, often in the neonatal period. As a result, problems with the foreskin are increasing and patients are often referred to the emergency department for treatment.

Parents are often unsure of the correct management of the foreskin. In most cases the foreskin should be left alone until the age of 5 or 6 and then should be retracted gently to clean the under surface of the foreskin and the glans. If the foreskin can be retracted easily at an earlier age, the child should be encouraged to retract it gently in the bath or shower every night to clean the glans and inner surface of the foreskin. If the foreskin cannot be retracted easily, the use of a mild steroid ointment for a short period may correct the phimosis. If this is not successful, circumcision may be required.

Phimosis of the foreskin

In this condition the foreskin cannot be easily retracted, and in the more severe cases the outflow of urine is significantly obstructed. Ballooning of the foreskin may occur with micturition when the urine flow into the foreskin space is greater

than the flow exiting out of the foreskin. Urinary tract infections,[13] dysuria and possibly urinary retention may develop.[14]

In patients with severe phimosis with suspected secondary infection, a sample of urine and swabs from the foreskin should be obtained and the patient should be commenced on intravenous or oral antibiotics. In cases of urinary retention, urinary diversion may be required. For example, a suprapubic catheter may need to be inserted under general anaesthesia. Alternatively, urgent circumcision can treat the condition.[14]

In mild cases of phimosis, which is not a medical emergency, the application of half-strength Betnovate® ointment to the tip of the foreskin for 6 weeks has a reported success of 75–88%.[14,15]

Balanitis

This condition usually arises when phimosis of the foreskin is present and infection has occurred in the space under the foreskin. Bacteria, often enteral or cutaneous in origin, migrate into this space and as they cannot be washed away, infection results. This is frequently a subacute or chronic condition but on occasions can be acute.

A sample of urine and swabs of the foreskin should be obtained, and the patient should be commenced on IV or oral antibiotics. The condition usually subsides within a few days. However, in the long term, circumcision may be required.[16]

Balanitis xerotica obliterans, also called lichen sclerosus et atrophicus, is thought to be an autoimmune condition affecting the foreskin and glans. It leads to irreversible phimosis of the foreskin and presents with inability to retract the foreskin and varying degrees of urinary obstruction. It is best treated with circumcision, as studies using topical steroid treatment have failed to show any permanent improvement.[17,18]

Paraphimosis

On occasions the foreskin may be tight, but the child, or parent, may have attempted to retract it and not returned it to its normal position. Oedema of the foreskin distal to the tight ring can develop and the foreskin can become painfully swollen and difficult to reduce.

The foreskin is more easily reduced the sooner that the patient is treated.[19] The

patient should be given appropriate analgesia and gentle digital pressure should be used on the foreskin through saline-soaked gauze. Once some of the oedema has been dispersed the foreskin should be gently replaced in its normal position. If this is not possible the child will need to be seen by a paediatric surgeon, who may attempt the same procedure. If this again fails, the foreskin should be reduced under a general anaesthetic. Some surgeons favour circumcision whilst the child is anaesthetised so as to avoid another anaesthetic in the future.

Historically alternative methods described to reduce the paraphimosis were the use of hyaluronidase[20] and puncture technique[21] whereby the oedematous foreskin is punctured in several places with an 18-gauge hypodermic needle to evacuate the oedema so as to allow for easy reduction of the foreskin. This treatment is no longer recommended.

Priapism

An erection lasting longer than 4 hours, without sexual stimulation, is defined as priapism. This is an acute emergency. Low-flow (or ischaemic) priapism is due to obstruction of venous outflow from the penis and accounts for 95% of cases. It may occur spontaneously or it can be secondary to medication, sickle cell disease or leukaemia. Usually the corpora cavernosa is painful and fully rigid with minimal or no involvement of the corpus spongiosum and glans penis. Uncontrolled arterial inflow, usually caused by direct trauma to the penis or perineum, results in high-flow priapism.[22] Usually the corpora cavernous are not fully rigid. Both types of priapism can result in a painful, sustained erection. Urgent referral to a paediatric urologist is required for immediate treatment or surgery. Therapeutic aspiration, with or without irrigation of the corpora, is the initial treatment of low-flow priapism. Intracavernous injection of sympathomimetic agents is the next step. Perineal compression should be attempted as treatment for high-flow priapism as it may successfully reverse the condition.[23] If this fails, colour Doppler ultrasonography of the corpora cavernosa can reveal a blood leak. Bilateral internal pudendal arteriography and embolisation can then follow.[24,25] Intracavernous injections of etilefrine can be effective in reducing priapism in children with acute sickle-cell crisis.[26] Failure to resolve the condition in a timely manner may result in permanent inability to have erections and/or penile fibrosis.[27]

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SECTION 27

Transport and Retrieval

OUTLINE

27.1

Paediatric emergency retrieval

Felix Oberender

ESSENTIALS

1 Critical illness in children is comparatively rare and can create unique challenges for clinicians working in healthcare settings that cater predominantly to adults. Paediatric emergency retrieval services provide ready access to specialist expertise and support while also facilitating transport and placement of the child in a tertiary paediatric intensive care unit (PICU). This should allow the referrer to liaise with a single point of contact and otherwise concentrate on managing the critical ill child.

2 The paediatric retrieval team must be skilled, trained and equipped to manage the full range of critical illness in children of all ages.

3 Commonly operating under the remote supervision of a paediatric intensive care retrieval specialist, the paediatric transport team stabilises the child at the referring hospital in collaboration with the local team before transport to the receiving PICU. Stabilisation and readying for transport require a careful balance between necessary interventions and the need to not delay delivery of the child to definitive care.

4 Efficient and effective communication is central to success in retrieval.

Paediatric retrieval

Children constitute a minority of the population in developed countries, and critical illness is rare when compared to adults. As a result, paediatric intensive care and paediatric emergency retrieval are small, specialised fields of practice. In most jurisdictions, paediatric critical care is the remit of a specialised paediatric intensive care unit (PICU) located in a large, tertiary children's hospital. In Australia and New Zealand, there are only seven PICUs. Intensive care for newborns, on the other hand, is a larger field and delivered in 22 neonatal ICUs (NICUs) in Australia and six in New Zealand. For retrieval of critically ill children, a range of operating models exist, which suit the needs, resources and function of different healthcare systems.

Paediatric retrieval may be delivered as part of the regional service of a tertiary PICU. Alternatively, for a large population critical care transport for children may be resourced independently and delivered by a dedicated, stand-alone paediatric transport service. In some jurisdictions, retrieval services operate models combining paediatric, newborn and/or adult retrieval in a single organisation. In any case, the principle of providing a single, regional point of contact for referrers and a specialist retrieval team is based on evidence that this approach to intensive care and emergency transport for children results in better outcomes.[1,2] The paediatric retrieval service consequently plays a pivotal role not only in transferring children from rural, regional and non-paediatric metropolitan hospitals to the PICU or NICU but, importantly, also in facilitating ready access for referrers to the expertise and support available in tertiary centres.

What's different about children?

This general question is addressed in other chapters. From a retrieval point of view, it is worth noting that the management of a critically ill or deteriorating child tends to be a relatively rare event for the emergency physician in a non-paediatric centre. Moreover, the majority of critically ill children are infants or small children younger than 5 years of age, placing many practitioners well outside their comfort zone. In such circumstances, sound general clinical skills combined with over-the-phone advice and support from the tertiary children's hospital or paediatric retrieval service become essential components of high-quality care. Overall, the threshold for transport of a child is often considerably lower than for an adult as the referring hospital's capacity to manage a sick paediatric patient must be taken into account.

Paediatric emergency referrals

In most developed jurisdictions, formal systems for referral of patients, including children, to higher levels of care exist. For emergency referrals this generally involves a telephone call to a selected referral hospital or retrieval service. Clinical and logistical decisions follow from the referral call and the quality of this initial communication is therefore critical. Most retrieval services conduct the referral call as a semi-structured interview, guiding the referrer through essential aspects of the case. The ISBAR communication tool (Introduction, Situation, Background, Assessment and Recommendation)[3] is widely used for this purpose (Box 27.1.1).

Box 27.1.1 ISBAR Referral Tool

Introduction	Referrer's name, clincial role, location and contact number Patient's name, age/date of birth, weight
Situation	The clinical situation (e.g. status epilepticus, child is intermittently apnoeic, will likely require intubation)
Background	1. past medical history 2. presenting problems/history of presenting illness 3. management undertaken, investigations performed 4. review of systems • Airway • • clinical findings • intubation grade • ETT size • ETT position • Breathing • • clinical findings • ventilation parameters • blood gases • CXR • Circulation • • clinical findings, HR, BP, rhythm • • vascular access • • fluids (bolusses for resuscitation and continuous infusions) • • vasoactive medication • D • • clinical neurological findings • sedation • muscle relaxants • • CNS imaging • E • • temperature • blood results incl. blood glucose • microbiology • F • • family/parents
Assessment	The working diagnosis or overall assessment of the situation
Request	Request for advice and/or retrieval with placement in the regional PICU

BP, blood pressure; CNS, central nervous system; CXR, chest X-ray; ETT, endotracheal tube; HR, heart rate; PICU, paediatric intensive care unit.

For the retrieval physician, the process of receiving and triaging a referral requires skill and experience as well as the ability to support the referring colleague looking after a critically ill child far away. The advice offered depends primarily on the patient's age and condition, but the decision to retrieve a sick child is also influenced by several other factors. Location and resources of the referring hospital play an equally important role as does the specific scope of practice of the referral and retrieval service within the healthcare system. Well-developed referral and retrieval systems operate a single point of contact, providing easy access for referrers with centralised advice and allocation of intensive care resources. This streamlines communication as well as decision-making and allows the referrer to focus on the care of the patient.[5,6] The often difficult task of 'bed-finding' together with all other logistics of the case

becomes the duty of the retrieval service. Less centralised systems can be more complex to navigate for referrers and may necessitate several conversations with receiving units and transport providers.

Paediatric referral and retrieval – roles and expectations

The referral and retrieval process usually involves at least four parties:

1. The referrer
2. The coordinator/administrator
3. The paediatric retrieval specialist
4. The transport team.

The referrer

- should only need to make a single phone call for the referral
- uses the ISBAR tool in order to provide details of history, assessment, examination and interventions in a structured fashion.

The coordinator/administrator

- receives the referral call
- documents the referrer's details, contact details and patient demographic data
- notifies the retrieval specialist and transport team who join the referral conference call
- documents the referral
- assists with transport logistics and communications.

The paediatric retrieval specialist

- guides the referrer through a semi-structured, ISBAR-based interview
- advises on patient management
- tasks the transport team with the retrieval and adjusts team composition

and skill mix accordingly
- is available for advice to the referrer and/or transport team
- undertakes bed-finding and liaison with receiving medical and surgical specialists.

The paediatric emergency transport team

- follows and, when indicated, contributes to the referral discussion
- readies for transport (adjusts/updates the retrieval kit to case needs)
- en route to the patient, prepares for the anticipated demands of the case and contingencies
- upon arrival at the referring hospital, assesses the patient and provides situation reports to the retrieval specialist at the operations base
- stabilises the patient
- updates the receiving unit on patient condition and estimated time of arrival.

Paediatric retrieval staff

Paediatric retrieval staff may include doctors, nurses, nurse practitioners and paramedics. Staff undertaking transport of sick children must have an established skill set in paediatric critical care as the retrieval environment does not lend itself to teaching and training of paediatric procedures. With respect to knowledge and expertise, the retrieval team should be guided by a senior specialist with experience in paediatric intensive care as well as in retrieval management who oversees the case remotely from the retrieval service's base of operations.

Both clinical and non-clinical skills are essential for paediatric emergency retrieval:

Clinical skills:

- Advanced paediatric life support
- Anaesthesia with non-invasive and invasive airway management in children of all ages
- Thoracocentesis and chest drain insertion
- Mechanical ventilation of neonates, infants, children and young adults
- Peripheral and central intravenous line and arterial access in

children of all ages.

Non-clinical skills:

- Retrieval communications
- Retrieval crew resource management
- Advanced clinical skills
- Non-invasive and invasive airway management in all paediatric age groups
- Internal jugular and subclavian vein cannulation
- Proficiency in management of unstable children with rapidly progressing life-threatening conditions (e.g. septic shock, cardiogenic shock).

Expert and quaternary subspecialty retrieval skills are infrequently needed and may require deployment of specialist team members for select cases, for example for management of the difficult paediatric airway or extracorporeal life support.

Paediatric retrieval equipment

As not all healthcare environments are guaranteed to provide sufficient equipment and consumables for paediatric patients, the paediatric retrieval team should aim to operate independently from the resources of referring hospital and ambulance service. Most paediatric and newborn retrieval services carry essential contents in ready-to-go retrieval kits that are kept on standby (Box 27.1.2).

Box 27.1.2 Ready-to-go retrieval kit

Airway

Nasal cannulae oxygen prongs
Self-inflating bag/valve/mask
Jackson-Rees modified Ayre's T-piece
Oxygen tubing
Face masks
Guedel airways
Suction unit

Yankauer suction tip
Flexible suction catheters
Laryngoscope handle x2
Laryngoscope blades curved
Laryngoscope blades, straight
Magills forceps
Endotracheal tubes
Introducers (stylets) for endotracheal tubes
Laryngeal mask airways (LMA)

Difficult airway

Bougies
Airway exchange catheters
Video-laryngoscope including blades
Tracheostomy set

Breathing

Paediatric stethoscope
Transport ventilator with paediatric/neonatal capability
Ventilator tubing
CPAP device
Nebuliser kit
Chest drains
Heimlich valves or similar
Oxygen supply and reserve
Portable blood gas analyser

Circulation

IV cannulae
Arm splints
Tourniquet
Transilluminator
Intraosseus needles
Central venous catheter (CVC)

CVC insertion kit
Infusion pumps
Syringes
Minimum-volume infusion extension lines
Infusion/medication labels

Monitoring

Glucometer

The paediatric retrieval kit must also contain a range of medications essential for the treatment of critically ill children (Box 27.1.3).

Criteria for transport

The decision to transport a sick child is very context-specific and depends to a large degree on the referring hospital's capability and paediatric service provision.

Box 27.1.3 Retrieval kit medications for critically ill child

Anaesthestic agents

- Fentanyl
- Ketamine
- Propofol and/or thiopentone

Muscle relaxants

- Rocuronium or suxamethonium
- Vecuronium, atracurium or cisatracurium
- Pancuronium

Cardiovascular medications

- Adrenaline (epinephrine)
- Metaraminol
- Noradrenaline
- Dobutamine
- Atropine
- Adenosine
- Esmolol
- Amiodarone

Sedation and antidote

- Morphine
- Midazolam
- Chloral hydrate
- Naloxone

Antibiotics

- Penicillin
- Third-generation cephalosporin
- Aciclovir

Antiepileptics

- Phenytoin
- Phenobarbitone

Asthma treatment

- Salbutamol
- Ipratropium bromide
- Aminophylline
- Magnesium sulphate
- Methylprednisolone

Other

- 0.9% saline
- Frusemide
- Mannitol
- 3% saline
- Potassium
- Calcium
- Sodium bicarbonate
- Water for injection
- Ondansetron
- Glucagon

Generally, children fall into one of four categories for retrieval:

Intensive care: the child is intubated and ventilated and/or on inotropes.
High-dependency care: the child requires non-invasive ventilation.
At-risk: the child is at risk of requiring intensive care or high-dependency care and is in a centre where this cannot easily or safely be delivered.
Unknown: occasionally, the referral conversation does not yield sufficient information to determine the child's status in a location with limited paediatric skills and expertise. The child is retrieved to a paediatric centre based on precautionary principle.

Box 27.1.4 suggests conditions that should prompt discussion with a paediatric retrieval service and/or paediatric ICU.

Degrees of urgency

Classifying the urgency of a transport is essential for effective clinical decision-making as well as resource allocation:

Time-critical: Imminent threat to life or limb
Urgent: No immediate threat to life or limb. This is the standard scenario for most of paediatric emergency retrieval. An emergency referral

requires urgent but not time-critical action.

Complex: Complex retrievals require a high degree of planning. They can be urgent but cannot usually be executed in a time-critical fashion. Examples include referrals for transport on extracorporeal life-support (ECMO), extreme-distance and international retrievals.

Return/elective: These are non-urgent transports usually facilitated to free capacity in the tertiary centre by returning recovering patients to non-tertiary centres.

Paediatric critical illness can occasionally be difficult to gauge but there are a variety of circumstances that should prompt immediate referral and retrieval without delay. Examples include:

1. Lactate >6 mmol/L or pH <7.0
2. Upper airway obstruction persisting despite more than 2 doses of adrenaline (epinephrine)
3. Upper airway obstruction resulting in hypoxaemia (SpO_2 <90%)
4. Pneumonia with hypoxaemia (SpO_2 <90%)
5. Sepsis or shock requiring intubation
6. Persistent signs of shock despite more than 40 mL/kg fluid resuscitation
7. Ongoing seizures despite 2 doses of midazolam and loading with a long-acting agent (phenytoin, levetiracetam, phenobarbitone)
8. Signs of raised intracranial pressure
9. Unconsciousness with worse than flexion motor response

Box 27.1.4 Conditions that may necessitate retrieval

1. Head injury (symptomatic)
2. Altered level of consciousness (for any reason)
3. Hypoxia despite oxygen therapy
4. High oxygen requirement
5. Respiratory failure (e.g. bronchiolitis, severe asthma, apnoea)
6. Upper airway obstruction
7. Near drowning (especially with neurological depression or respiratory symptoms)
8. Ingestion with risk of circulatory, airway or neurological compromise

9. Envenomation
10. Burns:
    - >10%
    - encircling the neck or involving the airway, face, hands, feet, perineum or inner joint surfaces
    - associated other significant injury
    - electrical or chemical burns.
11. Seizures (with persisting neurological depression).
12. Major trauma (including spinal injury)
13. Metabolic disturbance, e.g.:
    - diabetic ketoacidosis
    - acidaemia
    - severe biochemical abnormality.
14. Heart failure or arrhythmia (symptomatic)
15. Shock (requiring treatment with volume replacement or inotropes), e.g.:
    - blood or fluid loss
    - dehydration
    - septicaemia.
16. Other causes of neurological depression:
    - Central nervous system (CNS) infection
    - Acute life-threatening episode.
17. Any condition with the potential for sudden cardiovascular or neurological deterioration.

Adapting such a list may be helpful in determining the local policy. If in doubt, consult.

10. Any arrhythmia with haemodynamic compromise (shock, hypotension, signs of heart failure)
11. Any child with suspected systemic to pulmonary shunt who is about to be intubated or needing inotropes
12. Any child with suspected cardiomyopathy/myocarditis
13. Cardiac or respiratory arrest.

For time-critical retrievals, it is important to weigh the risk of delay due to time spent waiting for the specialist paediatric retrieval team to reach the patient against the relative risk of immediate transport by a team of local doctor and nurse or paramedic. In a neurosurgical emergency, for example, it is often

preferable to facilitate immediate transport by suitably qualified personnel from the referring hospital to the nearest neurosurgical centre for life-saving intervention.

Definitions

- Mobilisation time: time from decision to retrieve to team ready for departure
- Response time: time from decision to retrieve to arrival at the patient's bedside (= mobilisation time plus travel time)
- Scene time: time spent at the referring hospital for patient stabilisation and readying for transport
- Retrieval time: total time from decision to retrieve to handover to receiving unit
- Mission time: total time from decision to retrieve to return to base.

Transport platforms

The choice of transport platform for retrieval depends on the retrieval system setup, the distance to be travelled and the condition of the patient as well as on a variety of other factors such as weather and momentary overall demand on retrieval resources. Further considerations include traffic, available landing sites, proximity of airports to the referring hospital and the need for transport platform changes.[4]

Road ambulance

For most paediatric retrieval services, road ambulances are the most commonly deployed transport platform for retrievals within a range of approximately 100 km. In well-resourced retrieval services, the transport service may operate its own, custom-fitted road vehicles while in less developed systems, the retrieval team requests transport resources from a local transport provider such as the ambulance service. Compared to rotary and fixed wing platforms, road transport is inexpensive and obviates the need to change transport platforms en route to the receiving unit. The workspace for the retrieval team is comparatively generous and the vehicle can be stopped, should emergency procedures need to take place. However, road ambulances are subject to traffic conditions, delays

and risks of accidents. There has been an increasing awareness in recent years about the hazards of driving with lights and sirens and many ambulances services today operate with strict protocols for the use of emergency signals.

Rotary wing aircraft

Helicopters offer the advantage of fast retrieval over longer distances. If both referring and receiving hospital are equipped with helipads, transport may be point-to-point and highly efficient, if very costly compared to road transport. A more distant landing site at either end, however, necessitates transfer to a road transport platform with the potential to considerably prolong retrieval times and adding the risks associated with inter-platform transfer of the patient. The distance over which a helicopter can travel efficiently and at speed depends heavily on the type of aircraft but also on weather and the required altitude.

From a transport medicine point of view, the working environment inside a rotary wing aircraft is usually quite confined and offers limited ability to assess as well as to deliver treatment to the patient. The stressors of flight to both crew and patient are significant and include noise, vibration and changes in cabin pressure. Most rotary wing aircraft are not pressurised, requiring the retrieval team and aircrew to carefully consider the implications for the patient.

Fixed-wing aircraft

Fixed-wing aircraft are the transport platform of choice for long-distance retrieval as speed and range are greater than those of rotary wing aircraft. Change from and to a road transport platform is required at either end of the air retrieval. Many modern air ambulances operate cabin altitudes within the range of commercial airliners (4000–7000 ft above sea level) and can also be further pressurised to sea level if the patient’s condition demands, e.g. in severe asthma or air-leak syndrome.

As a retrieval environment, the cabin of a fixed wing aircraft is a confined space limiting assessment of and access to the patient. Patient and crew are subjected to the same stressors of flight as in rotary wing aircraft, albeit to a slightly lesser degree. Take-off and landing, however, expose the patient to considerable acceleration and deceleration forces which may exacerbate physiological instability.

Weather

Both rotary and fixed-wing aircraft are subject to weather conditions. In the interest of safety for both patient and crew, the decision-making algorithms of air operators regarding flight clearance are strict and not open to negotiation. Both referrers and retrieval team must refrain from attempting to exert pressure on air crew or operators if clearance to take off or land is not forthcoming despite a patient requiring urgent transport.

While waiting

The input from the retrieval specialist over the phone depends to a large degree on the confidence of the referring doctor. However, it is important to focus on manageable tasks and await their execution. As a general rule, it is good practice for retrieval specialists to avoid giving more than three pieces of clinical advice at a time. Referrers should feel at liberty to point out the limitations of their environment and healthcare team in actioning advice from the tertiary centre. The clinical conversation may be improved by video-conferencing either through a purpose-built system where available or via public-use, online services. The expertise of a range of specialists such as paediatric ICU, paediatric emergency medicine, surgery, burns, neonatology and other paediatric disciplines, as well as toxicology and envenomation experts, is available through regional, state and national networks.

Stabilisation

Stabilisation refers to interventions by the retrieval team after arrival at the referring hospital. It is intended to deliver immediate life-saving treatment and to reduce the risk of deterioration in the transport environment. The approach to stabilisation of the critically ill child has evolved significantly over the past three decades. The quick ‘scoop and run’ of early transport medicine gave way to a ‘stay and play’ philosophy in the 1990s when ‘the PICU came to the patient’.[5] Modern paediatric retrieval medicine takes the best of both these approaches and combines them into ‘safe and swift’ stabilisation and transport. Stabilisation follows standard algorithms such as Airway, Breathing, Circulation, allowing the transport team to follow a rapid, structured approach to the critically ill child. Attention is paid not only to the adequacy of current management but also to safety in transport. For example, airway and vascular access must be not only

adequately in place but also secured to withstand the rigors of travel and potential changes in transport platform. In addition, the retrieval team will plan to manage contingencies of further deterioration in the patient's condition. It is good practice to be ready and able to immediately implement at least one further line of treatment in case of major physiological system changes while en route to the receiving unit. For example, inotrope infusions should be ready for the septic child who stabilised after fluid resuscitation.

Communication and retrieval leadership

Transporting a critically ill child requires the retrieval team to operate across health services, specialties and geographical distances. A professional and streamlined approach to communication and leadership is critical for success and risk mitigation in this context. Senior medical staff with experience in paediatric intensive care as well as retrieval medicine should coordinate and lead the retrieval remotely from the operations centre. The transport team should stay in close contact with the base of operations, have ready access to advice and also deliver mandatory situation reports along the retrieval pathway. In addition, skilful communication with referring and receiving healthcare professionals is required.

Framework for communications during paediatric retrieval

Transport team and coordinating retrieval specialist at base of operations:

Mandatory:

1. Pre-departure briefing
2. Situation report after assessment of the patient at the referring hospital: status, suggested interventions for stabilisation, transport plan, update on logistics
3. Situation report prior to departure from referring hospital
4. Confirmation of arrival in and handover to receiving unit
5. Post-transport debrief.

As required:

- Situation report in case of any major clinical event, e.g. resuscitation and/or increase in level of life support
- Situation report in case of conflict arising between transport team members, with referring hospital or receiving unit staff or with parents.

Retrieval service/transport team and referring hospital:

- Referral conversation and clinical advice
- Retrieval decision, transport platform, estimated time of arrival (ETA), expected interventions for stabilisation including expected demand on local resources, destination
- Ongoing clinical advice as required
- Team introduction on arrival, updates on clinical management, transport logistics and timeframes
- Pre-departure summary
- Notification post arrival in receiving unit
- Post-transport follow-up.

Retrieval service/transport team and receiving unit:

- Notification about the referral and request for patient admission (‘bed-finding’)
- Pre-departure from referring hospital: situation report, anticipated case demand and ETA
- Update on any deterioration of the patient’s condition and/or major interventions en route
- Post-transport follow-up.

Transport team and parents:

- On arrival to the referring hospital: introduction, information about the process of stabilisation and transport, information about ability/inability to accommodate parents on transport, information about communication points with the family
- Pre-departure from referring hospital: update on situation, transport plan and ETA at receiving unit
- Post arrival in and handover to receiving unit: update on completed transport and current situation
- Post-transport follow-up.

Interface with adult retrieval

- In recent years, medical retrieval services for adults have developed and at times surpassed paediatric and newborn transport services with respect to professionalisation of retrieval medicine. Older children and adolescents may be safely and expertly transported by specialised adult retrieval services. Careful patient selection is key and both paediatric and adult retrieval specialists must be careful when making assumption about the other service's role and capability. Good communication between retrieval services, referrers and receiving centres is essential in this space as subtle differences in clinical practice and logistics can lead to delays and adverse events. Responsibility for clinical advice, transport logistics and bed-finding must be clearly assigned in order to avoid loss of effectiveness and efficiency.

Parents

A child becoming critically ill and requiring retrieval is a deeply upsetting event for parents. For the retrieval team, attention to good, if brief, communication with parents is an essential component of paediatric transport.

It is good practice in paediatric emergency retrieval to facilitate at least one parent to accompany their child whenever it is technically possible. Not only does this approach follow most parents' desire to remain close to their child but it also allows families to gain confidence in the transport team's clinical care. This is particularly the case for unstable children where it becomes important for parents to witness the team's efforts to treat their child.

In this context it is critical to discuss with parents the expectations and requirements of the retrieval team, such as conduct in a road or air ambulance or the need to step back during critical procedures. The approach to parents during retrieval should form part of the pre-departure briefing and ensure that all team members are aware about the parameters for parents accompanying their child on the mission.

Table 27.1.1

Examples of quality metrics in paediatric retrieval

Metric	Measure of/confounded by
Average mobilisation time	Service processes, service resources, case load, case mix, weather
Average mobilisation time for time-critical transports	Service processes
Rate of equipment failure	Service resources and processes
Average scene time	Clinical quality (team performance), case mix, referrers
Average scene time for intubated patients requiring no further interventions	Clinical quality (team performance)
Unplanned dislodgement of therapeutic devices	Clinical quality (quality of medical and nursing practice)
Success at first attempt at intubation	Clinical quality (team clinical skill)
Documented verification of endotracheal tube placement	Clinical quality (quality of medical and nursing practice)
Incidence of physiological instability	Clinical quality (medical and nursing expertise and skill), case mix, transport distance
Rate of sentinel events, e.g. cardiac arrest and death en route	Case severity, case mix, clinical quality
Rate of medication administration errors	Clinical quality (quality of medical and nursing practice)
Client satisfaction	Quality of communications, quality of logistics, clinical quality

Paediatric retrieval and end-of-life situations

Death in childhood, fortunately, is a very rare event. As a consequence, however, managing a paediatric end-of-life situation may present a very challenging task for clinicians whose practice does not include looking after dying children. It is not unusual, and is indeed part of the role of the paediatric referral and retrieval service, to support referrers in the management of acute end-of-life situations.

Three main end-of-life scenarios may be encountered in paediatric emergency retrieval:

1. Referral for advice while cardiopulmonary resuscitation (CPR) is in progress
2. Withdrawal of life support or discontinuation of CPR after arrival of the retrieval team at the referring hospital
3. Death during transport.

A referral for advice while CPR is in progress constitutes a time-critical emergency that requires immediate response by a senior specialist in paediatric intensive care and retrieval medicine. In many cases, the referral is a request for a second opinion:

- Have all causes of cardiorespiratory arrest been considered and adequately addressed?
- Have all resuscitative measures been exhausted?
- Do you agree that resuscitation should cease and death be declared?

While outcomes in paediatric cardiac arrest are generally very poor, particularly if cardiac arrest occurred out of hospital, time-critical retrieval should be considered if a reversible cause can be identified (e.g. hyperkalaemia) and CPR is achieving adequate oxygenation and cardiac output.

In rare circumstances, the transport team may arrive at the referring hospital and support the local team in withdrawing life support from the patient. This may be unexpected in a child after profound deterioration or it may be an anticipated mission task with the goal of facilitating acute end-of-life care in a healthcare setting not used to this scenario but close to and/or familiar to the child's family.

The death of a child during transport is an even rarer event. Skilled and professional communication is key in situations where death may occur during retrieval:

- Crew communications and management: discuss the approach to resuscitation including failed resuscitation en route
- Inform parents about the risk of death en route to the receiving unit and facilitate their presence during retrieval whenever possible. This will allow them to be with their child during an extremely life-threating situation and also to witness the efforts of the retrieval team to save their child.

A death during transport constitutes a sentinel event that requires both thorough clinical review and a pro-active approach by the retrieval service to ensure the welfare of staff involved in the case.

Quality

In contrast to hospital-based medical practice, quality measures for paediatric retrieval are few and not universally accepted. This is an area of active research.[6] When trying to measure quality in the complex environment of transport medicine, it is important to have a clear understanding of whether a metric

reflects process, resources or clinical care and how it can be confounded (Table 27.1.1).

Summary

Paediatric intensive care retrieval is a highly specialised area of practice and outcomes are best in centralised transport systems with specialised paediatric retrieval teams. In addition to providing a paediatric skill set, the retrieval team operates within a framework that facilitates ready access to paediatric critical care advice and support for referrers, streamlines communications and provides efficient logistics for optimal outcomes in critically ill children.

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27.2

Sick child in a rural hospital

Murali Narayanan

ESSENTIALS

1 There are significant challenges in translating experience from the urban to rural healthcare settings including differences in the patients, illnesses and health services available.

2 The ‘scoop and run’ model of pre-hospital care is not appropriate in rural/remote medicine – patients must be ‘packaged’ as much as possible before transport.

3 Coordination between rural, central and retrieval services must be optimal.

4 Rural services should be supported through targeted training of personnel tailored to the needs of rural clinicians, easily accessible guidelines, accessible consultation services, rural clinical facilities with necessary resources and a supportive relationship with a major urban centre.

Introduction

Most emergency physicians are trained in urban centres. Their skills and knowledge are most readily applied when trained staff, equipment and hospital facilities are immediately available, and when there is ready access to specialists. This becomes an issue when the sick child presents to the emergency department (ED) in a rural hospital. Protocols and treatment plans that are appropriate to urban hospitals may be difficult to apply directly.

Challenges in the rural setting

One-third of Australians live in areas defined as rural or remote (areas outside major cities). While Aboriginal and Torres Strait Islanders (ATSI) constitute 3% of the total Australian population, they make up 16% of the population in remote areas and 45% of the population in very remote areas. Chronic or recurrent infection, malnutrition, lack of transport, lack of access to health care and educational deficits are all factors which contribute to poorer health status of the people living in these areas.

Income

Although the overall income differences between rich and poor may be smaller, the median household income in rural Australia is 25% lower than that in major cities, and rural families are 50% more likely to be dependent on government pensions and allowances than urban families.

Low income can lead to reluctance to consult a doctor and to reluctance of the doctor to prescribe expensive though appropriate treatments or to commit the family to expensive travel to consult specialists in urban hospitals.

Housing

The quality of housing and of home maintenance is lower in rural areas: maintenance costs are higher and dwellings deteriorate faster in harsh environments and scarce accommodation leads to overcrowding.

Education

Lower rates of secondary school completion in rural areas than in the city affect both the child and his/her parents. Infant mortality is closely related to the level of education completed by the mother.

These issues apply also to children in isolated indigenous communities, where the additional problems of distance, access to health care and a relative scarcity of indigenous health workers lead to relatively late hospital presentation of severe illness.

Culture

A tradition of self-reliance and stoicism, combined with suspicion of medical services or of European society in general and varying issues with language difficulties may lead to reluctance to report illness and to delay in presentation to seek medical attention. Certain cultural issues, for example not speaking the name of a deceased Aboriginal person, or the tendency of Aboriginal women to avoid eye contact with a strange man, become important considerations in the setting of paediatric emergency medicine.

The illnesses

Birth history

Low birth weight and difficult access to antenatal care in rural areas (especially in indigenous communities) can lead to severe illness in the newborn period, with consequent disability and need for hospital care.

Trauma

Age-standardised rates of serious injury in children from road trauma, interpersonal violence and self-harm increase with distance from major cities in Australia: From 50% greater in inner regional areas to 100% greater in very remote areas. Farm accidents occur frequently: two-thirds of accidental child deaths on farms occur in boys.

Farm vehicles, and especially all-terrain vehicles and four-wheel motor bikes, account for many deaths and injuries, especially to farm visitors. Of child deaths on farms, 35–40% are due to drowning in dams or drains, especially in children aged less than 5 years.

Infection and diet

Infective conditions, especially of the skin, respiratory and gastrointestinal systems, occur commonly in children from remote communities, where rheumatic heart disease and post-streptococcal glomerulonephritis may result.

There is lack of easy access to fresh food, fruits and vegetables in remote areas. Dietary deficiencies and substance abuse may also contribute to patterns of illness different from those found in children who present to urban hospitals.

The health services

These include community clinics staffed by nurses and/or GPs; local hospitals (staffed by nurses, remote emergency physicians and GPs) and ambulance services; and regional (base) hospitals, which often have paediatricians, emergency physicians, surgeons, anaesthetists and advanced radiology and pathology facilities. Finally, there are city-based patient-retrieval services and tertiary hospitals with access to paediatric and neonatal intensive care facilities.

Although inner and outer regional and remote/very remote areas in Australia all have more primary care medical practitioners per 100,000 population than urban areas, this is far outweighed by the ready access of city dwellers to urban hospitals and to specialist care.

Rural GPs deal with a wide range of illness in a wide range of patients. The nature of general practice means that any single practitioner may rarely (or never) encounter any one of the critical life-threatening illnesses of childhood. This can potentially contribute to a delayed diagnosis and may lead to dilemmas in management. In many rural and remote areas, GPs may be in solo practice, so that consultation with a colleague is difficult, and many regions lack a regional paediatrician to provide timely consultation.

Regional hospitals frequently offer subspecialty clinics staffed by visiting specialists (e.g. paediatric surgery or paediatric cardiology) but these are relatively infrequent and may not coincide with the child's severe illness. The burden of diagnosis and treatment then falls on the clinician on the spot.

The resources available in rural hospitals vary: for example, pathology and radiology staff may be on-call rather than in-house after hours, and the selection of tests, scans and other investigations that are available may be limited.

Distance and difficulty of access to some rural hospitals mean that delays before arrival of a city-based retrieval team can be protracted, sometimes many hours. During this time, the rural clinician often has to manage a very ill and unstable child within the resources of the local hospital.

Caring for the critically ill child

Decisions to consider when child presents with an emergency

The 'scoop and run' model which may be appropriate in some metropolitan environments is not applicable in the remote setting. In fact, the vast distances over which these patients may need to be transported (usually by the royal flying

doctor service [RFDS] or other state-based retrieval services) mandate that they need to be stabilised *prior* to transfer. It needs to be appreciated that the time taken to adequately prepare a patient for transfer from a remote setting is far in excess of the time taken for the same patient in an urban setting. Effective and ongoing communication between the treating doctor, their specialist colleagues in the tertiary ED or paediatric intensive care unit, and the RFDS/retrieval service is critical to the child's management.

Coordination of a resuscitation team prior to the child's arrival

If the staff in a remote location have prior warning of the arrival of a sick or injured child, they have the opportunity to mobilise any local resources available to them, and also to liaise with tertiary specialist colleagues and ensure that lines of communication are open for when more clinical information is available.

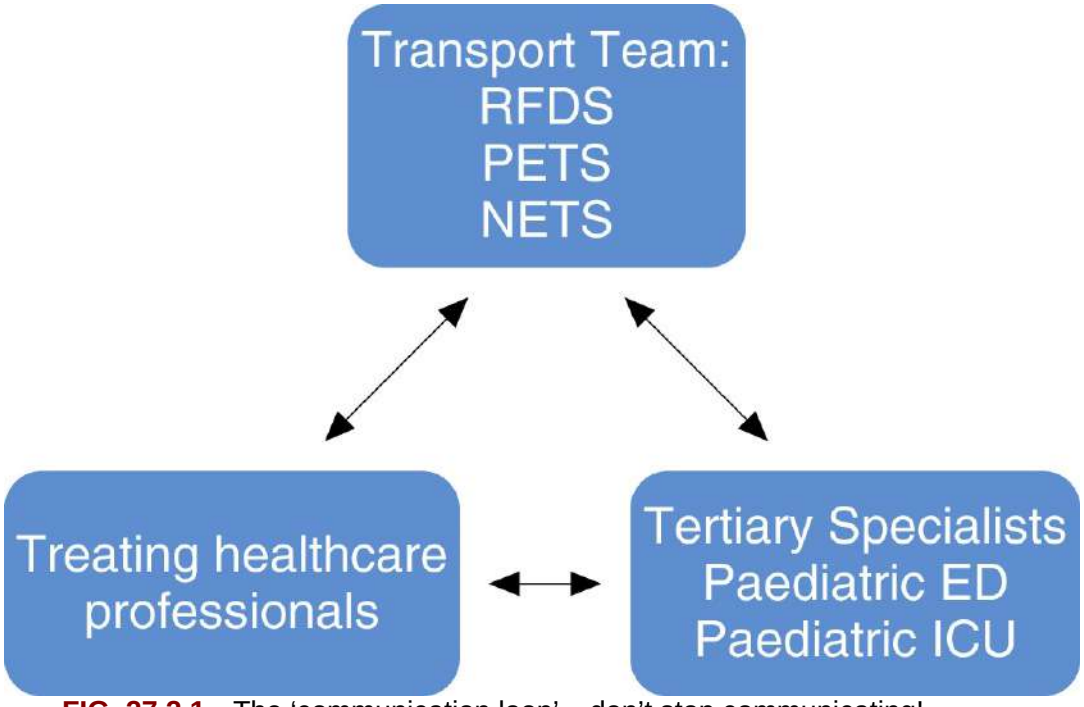

FIG. 27.2.1 The 'communication loop' – don't stop communicating!

Potential problems in the stabilisation of the

child

The unique issues associated with emergency medical transport and retrieval of an unwell child are discussed in Chapter 27.1.

It is important to utilise the expertise of GPs and nurse practitioners, clerical staff, orderlies, RFDS staff and rural medical personnel to share the load when dealing with the multitasking required in the management of the critically ill child. Midwives are very able in caring for sick neonates. One should be mindful of the fact that it is very stressful for the healthcare practitioner dealing with a paediatric emergency in the isolated remote setting. From their perspective they should utilise all available resources to help share the load. Do not underestimate the value of the telephone as an important 'piece of resuscitation equipment' to avail resources in times of need.

Conversely, from the perspective of the receiving tertiary unit, it is important to offer ongoing advice and support and to help plan and facilitate the transfer by liaising supportively with the transferring team (Fig. 27.2.1).

What can be done to assist care in remote environments?

Personnel

The numbers of medical and nursing practitioners available in rural communities may be increased by financial and educational incentives and by imposing conditions on professional registration that require a period of rural service. These may be backed up by enhanced arrangements for in-service education and facilitation of career paths. If these arrangements are to succeed, serious consideration must be given to spouse employment, education of children and provision of incentives such as a house and car.

Education

Regular education sessions to remote environments can be arranged through urban tertiary paediatric hospitals. These should be on-line when possible, with self-testing. They should be relevant to the needs of the local practitioner and should be followed up by face-to-face teaching sessions which are primarily hands-on, using the information given in the on-line tutorials. The hands-on teaching may consist of procedural sessions, as well as paediatric mock scenario

teaching and group discussions of clinical issues which have arisen within the hospital.

This form of regular in-service teaching should be centrally coordinated, with input from the local practitioners' organisation. It must be centrally funded, with adequate time allocation in the local practitioners' calendar.

As tertiary paediatric hospitals have responsibility for medical care of children in their region, they may organise hands-on training programmes (e.g. advanced paediatric life support [APLS] courses), relevant to rural practice and located in the paediatric hospital, in which rural doctors are invited to participate.

Content of training

Illness recognition and resuscitation courses, such as APLS, paediatric advanced life support (PALS), advanced trauma life support (ATLS)/early management of severe trauma (EMST) should be very freely available and the fees government-subsidised for nurses and doctors in rural hospitals. Follow-up training in these areas and on-line plus face-to-face courses on topics relevant to severe childhood illness should also be provided by the staff of the regional tertiary hospital.

Consultation support

A centrally organised and funded framework for provision of 24-hour, 7-day advice on all medical subspecialties (including paediatric subspecialties) would relieve much of the uncertainty of remote rural practice and would remove many of the delays currently seen in diagnosis and management of relatively uncommon conditions.

The use of telemedicine, including teleradiology, as well as the widespread availability of point-of-care biochemical testing can narrow the uncertainty in diagnosis and allow more accurate assessment and monitoring of the response to treatment in the remote setting.

A centrally coordinated telemedicine network requires a terminal at each point of care (rural medical centre or rural hospital emergency department or paediatric ward), capable of high resolution video transmission and reception, and one or more corresponding terminals within the city paediatric hospital (and general teaching hospitals for other medical specialties). Facilities for on-line transmission of medical imaging to a centrally located radiologist are also needed.

A roster of designated subspecialty consultants with access to telemedicine facilities 24 hours per day would complete the consultation support network.

Transport

In some cases, even the above consultation system will not enable the child to be managed at the rural hospital, and specialist care at a paediatric hospital will be needed. A centrally located paediatric triage and retrieval service, staffed by nurses and doctors with training and experience in triage and paediatric retrieval (see Chapter 27.1) can transfer severely ill and unstable patients to an urban tertiary hospital.

When distances to a large city are very great, it may be more appropriate to transfer the child to a smaller base hospital closer to the child's home, if the severity of the child's illness permits.

Management protocols

These should be available for a range of severe childhood illnesses and be immediately available (e.g. on-line) in the ED of rural hospitals. These protocols should be updated regularly to keep abreast of current therapies. They should be modified to be clearly appropriate to rural hospital circumstances, although they may be adapted from those in use in the regional tertiary hospital.

These protocols should be jointly produced by tertiary hospital staff, regional paediatricians and community general practitioners, to ensure suitability for the conditions which prevail in a rural hospital. They should contain advice on when to consult, whom to consult, and how to arrange transfer to a paediatric hospital or to the paediatric ward of a regional base hospital.

Hospital facilities

Apart from telemedicine, teleradiology and point-of-care biochemistry facilities, mentioned above, the emergency department of a country hospital which only occasionally treats severely ill children needs to be equipped adequately and appropriately to deal with the child on those infrequent occasions.

A collection of appropriate resuscitation and paediatric care equipment, common to all rural hospitals, maintained by the central health authority of the state or country, devised and updated by people who are using such equipment

frequently, should be supplied to each hospital. This equipment should be kept in the ED of the rural hospital and checked daily. When an item is used or out of date, it should be replaced promptly from the regional urban store. Such items include a paediatric range of cervical collars; arm splints; intravenous cannulae; laryngoscope blades; endotracheal tubes; oropharyngeal airways and laryngeal mask airways (LMAs).

Relations between rural and urban hospitals

In the interests of better coordination and of uniformly good service provision to children in rural areas, the mechanism by which facilities are extended to rural hospitals and their staff should be administered at the regional paediatric hospital. This mechanism should be sensitive to the needs of staff of rural hospitals, and responsive to their suggestions about changes.

Clearly, it is very important that tertiary hospital staff work cooperatively with practitioners in rural areas and that members of this team are aware of each other's needs. This culture of cooperation, support and mutual respect should be part of the way in which the tertiary hospital sees its role in the health system.

Regular communication between urban and rural practitioners, aided by teaching exchanges, training visits and telemedicine consultations, can promote awareness by each party of the concerns and ideas of the other.

Acknowledgement

The contribution of Robert Henning as author in the previous edition of the textbook is hereby acknowledged.

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SECTION 28

Teaching Paediatric Emergency Medicine

OUTLINE

28.1

Availing web-based resources

Tessa Davis, and Colin Parker

ESSENTIALS

1 Web resources are an integral part of everyday practice.
2 Users can generate web material more easily via media such as blogs, forums, wikis, social networks and podcasts.
3 Clinicians should evaluate the quality of online educational resources and assess individual offerings with a critical eye.

Accessing web-based resources

Over the past decade, the world of paediatric emergency medicine (PEM) has been transformed through the delivery of educational resources via the internet. While books are still a popular medium for doctors with regard to their work and learning needs, almost all junior doctors are using web-based resources daily in their work. It is important for all medical staff to have a working knowledge of how to utilise this evolving technology to its maximum effect. This chapter aims to briefly explore the needs of PEM staff, types of educational tools online and possible future trends.

Needs of paediatric emergency medicine staff

In previous times, resources for staff working in emergency departments (EDs) were scattered around notice boards, bookshelves, folders and in-trays. However, the technology to make these operational, educational and social resources available from a single portal is now mainstream.

Operational materials used in everyday clinical work include work rosters, memos, notice boards and staff directories, as well as clinical resources. Most of these are now delivered online. Clinical guidelines and protocols of local, national and international origin can be accessed via the internet. More significantly, tools for drug doses and other clinical calculations are paving the way for interactive clinical decision support platforms, where disease-management algorithms can be integrated with patient flow, investigation ordering and interpretation, and the electronic health record. Documents are increasingly stored on the web, in the 'cloud'. Many services allow editing and sharing of documents online, as a smarter alternative to emailing multiple versions between collaborators and enabling easy access from any web-accessible device.

Educational materials are used online every day at work and increasingly from home, the requirements varying between individuals and their learning needs. Interactive forms of learning, such as lectures, tutorials and one-to-one supervision, are still relevant but can now be supplemented by newer technologies, to deliver content asynchronously, at a time and place convenient to the learner.

Educational tools available online

Web-based resources are rapidly evolving in terms of the modes of delivery as well as format and flow of content. Medical staff can retrieve such content from the hospital computers, if access allows, or via their own smartphones.

Traditional content such as journal articles or evidence-based guidelines are widely available online. Journal articles can be searched via Pubmed or Google Scholar and then accessed online to read the full content.

Mobile apps such as Read by QXMD (http://www.qxmd.com/apps/read-by-qxmd-app) help doctors keep up to date with the latest journal abstracts in PEM, allowing them to briefly read a feed of the latest publications and select which articles they want to delve into further.

Online journal clubs conduct critical appraisals of recent paediatric articles, for example Archives of Disease in Childhood's Twitter journal club (#ADC_JC). And some websites provide summaries of interesting recent research, for example the Bubble Wrap series on DontForgetTheBubbles.com (DFTB). Institutional guidelines can be readily accessed online, such as NICE (https://www.nice.org.uk/guidance) and the British Thoracic Society

(https://www.brit-thoracic.org.uk/standards-of-care/).

Blogs (web logs) are also a great source of clinical information. Blogs are seen as a less traditional method of delivering education but are written by people working in the clinical world, on relevant topics and most will have a good peer-review process prior to publication. Users can leave comments, effectively supporting online discussions. Podcasts deliver this online content via audio and there are several excellent paediatric emergency medicine podcasts (http://www.pemed.org/, http://empem.org) that can be accessed via a podcasting app. The major advantage for busy doctors and nurses is being able to enjoy these offerings while travelling, exercising, or relaxing at home.

Rich site summary (RSS) feeds (e.g. Feedly – https://feedly.com) can be used to collate all the vast swathes of online information available to allow for one easy-to-read feed of relevant content. RSS is a way for information from multiple sources (feeds) to be pushed (rather than having to be actively fetched or pulled) to the user, and collated in one place, the RSS reader or feed aggregator. This allows the user to receive information in a much more efficient way than browsing journals or websites, and content can be viewed at a time and place of the clinician's choice.

Mobile apps are used as quick reference for doctors, particularly as they can be accessed in the absence of a wifi connection or mobile reception. Some staple paediatric emergency apps such as PalmEM, Traumapedia, Rapid ECG, and NICE BNFC can allow instant access to essential information.

Social media

Social media is simply a platform for the delivery, dissemination and discussion of educational content. Social networks, such as Facebook and Twitter, are increasingly being used by medical professionals to broaden their social and professional networks and to share knowledge. Paediatric emergency doctors on Twitter can have thousands of followers and tweet daily about relevant paediatric emergency medicine issues.

See *http://dontforgetthebubbles.com/twitter-people-to-follow/* for an up-to-date list of PEM Twitter people to follow.

Social networks allow for peer-to-peer networking, for using colleagues online to answer and debate clinical questions, and for keeping up to date. Blogs, a type of social media, facilitate the discussion of published educational material through post-publication comments 'below the line' on the blog itself or on other

social networks. This post-publication peer review brings advantages not currently afforded to traditional journals, which rely mainly on accurate pre-publication peer review.

Wikis are web pages that can be edited by any user, thus harnessing the power of collective knowledge, accelerating the editorial process and keeping content current.

Forums are a dedicated online space for discussion topics, usually highly specific to a defined area of interest. Both forums and other networks such as Twitter have discussion threads that are effectively an online record of a written conversation between two or more users. As such, sensible use of social media needs to be borne in mind – anything published online can be seen by anyone at the time of publication or at any point in the future and will be very difficult to retract.

Pitfalls of online content

The challenge for healthcare professionals involves assessing the quality and trustworthiness of each of these sources of information. While most online free open-access meducation (FOAM) content goes through a peer-review system not dissimilar to that of traditional journal articles, the same critical eye should be used with online content as when reading a journal article.

Assess whether the content is reliable by looking at the source, the authors, the references and whether it makes sense in the context of one's own clinical practice. Chan et al. have published scoring tools to evaluate online educational resources.[1,2] The majority of journal articles one reads will not change one's clinical practice and neither will the majority of online content viewed. Both should be treated with the same degree of analysis.

When sharing content online or using online content for other purposes, be mindful of copyright issues. The trend is towards sharing more information for free and greater use of flexible copyright licensing of intellectual property such as the Creative Commons license. Many blogs and podcasts are FOAM,[3,4] which means their content can be used, adapted and reused freely. Always check the copyright terms before using information from a website.

And, of course, patients are at the centre of all we do, and their confidentiality must be preserved. No patient information, stories from work, photographs from workplaces, or identifiable photographs should be used without the express consent of the patient for use including online publication.

Future Directions

As medicine catches up with technological advances, we can look forward to unrestricted, high-speed wireless internet access in our emergency departments, using hand-held and bedside devices which integrate with decision-support software and the electronic health record. Information flow to patients could be facilitated by these new systems, including the electronic provision of summaries of clinical information, test results, discharge instructions and medication lists. Educational interactive spaces incorporating rich site summary feeds, blogs, vodcasts, editable wikis and reference documents will continue to embed themselves further into the foundations of paediatric emergency medicine practice.[5]

Links

Useful web resources for paediatric emergency medicine:

- DontForgetTheBubbles.com
- EMPEM.org
- PedEMMorsels.com
- PediatricEducation.org
- StEmlynsBlog.org
- PEMBlog.com
- PEMSource.org
- RolobotRambles.com
- PEMPlaybook.org
- PEMAcademy.com.

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Conflict of interest

Tessa Davis is a co-founder of DontForgetTheBubbles.com. Colin Parker is the founder of EMPEM.org.

28.2

Teaching paediatric emergency medicine

Colin Parker

ESSENTIALS

1 Effective communication skills and healthy attitudes are prerequisites for good paediatric emergency medicine practice, and their teaching should take precedence over teaching knowledge and skills.

2 Self-preservation skills should also be taught.

3 Clinical experience is essential in gaining perspective on clinical knowledge.

4 There are unlimited methods for acquiring new knowledge; part of teaching new trainees is educating them about how to use available media and methods to best effect.

Introduction

Despite the massive explosion in the availability of information, doctors will never be replaced by computers. Paediatric emergency medicine (PEM) has elements of science, particularly with the steady growth of evidence-based medicine but also relies heavily on the practical application of experience. Perhaps the emergent aspects of PEM, such as trauma resuscitation, have a more formulaic basis; however, clear and effective communication from the team leader is of paramount importance in a challenging resuscitation.

The bulk of PEM does not consist of emergent interventions, but rather risk stratification, and trading information with parents of children who may or may

not be very unwell. Parents bring their children for reassurance, explanation and occasionally some form of treatment. They want to feel that they have been taken seriously and that someone cares about the wellbeing of their child. In short, they want to feel better after seeing the doctor.

The practice of PEM requires a set of skills, knowledge and attitudes broadly similar to those required for the practice of clinical medicine generally but with the added requirement of having to communicate effectively under the pressures of time and emotional stress. Because doctors' attitudes are so enmeshed with ability to communicate, it is important to teach the correct attitudes before teaching skills and knowledge. Doctors with healthy attitudes will be driven to continually expand their knowledge and skills and will seek out any available resources to improve the way they care for their patients.

Desirable attitudes in paediatric emergency medicine

With the success of immunisation programmes and, to a lesser extent, injury prevention strategies, there seems to be a changing spectrum of illness in children presenting to first world emergency departments (EDs), with fewer critically unwell children, medicalisation of behavioural issues, and children with a functional component to their 'dis-ease'. In addition, the public perception of the wonders of modern medicine results in high expectations, which are sometimes difficult for clinicians to meet. The best protection is to approach with healthy attitudes:

- Humility
- Caring
- Empathy and compassion
- Non-judgemental approach
- Honesty and integrity
- Advocacy and healthy paternalism
- Self-monitoring and awareness of cognitive errors in medicine.

Humility

Growth in the acquisition of medical knowledge can be likened to exploring a forest: initially it is hard to see the wood for the trees; as one gets to know the

landscape, one can develop an appreciation for the large valley in which the forest sits. By the third postgraduate year doctors are familiar with the whole valley. Most then travel up to the crest and realise that the valley is but a small part of the countryside, and there is in fact a vast body of knowledge which they may never know and never discover. While knowledge itself is a valuable asset, a high knowledge-to-confidence ratio is perhaps more desirable.

Caring

Many communication difficulties can be overcome by having and projecting a caring attitude. With children especially, parents may not care what the doctor knows, until they know that the doctor cares. Doctors' own biases and personality differences can sometimes make this caring seem a difficult task. Projecting a caring attitude from the start of the consultation usually results in positive feedback which sets the scene for genuine caring to supervene.

Empathy and compassion

Doctors who are themselves parents find it easier to imagine the emotional strain of having a sick child. This tension may sometimes be expressed in fiercely protective terms by a worried parent, in the same way that adult patients often forego the social graces when they are unwell. It is important not to take this personally and try to imagine how the parents are feeling.

Non-judgemental approach

There is a perception among some ED staff that some children should not have been brought to the hospital because 'it's not an emergency'. They may blame the referring clinician, the parent or the patient. Everyone who brings their child to an ED has crossed some threshold of anxiety about the perceived illness, and every one of them has considered not coming to the ED. Most of them do not have significant knowledge of health-related matters. There is no place for a judgemental approach, which will not help the child, or prevent a future ED visit.

Honesty and integrity

It is important to teach our colleagues humility. Doctors are often humbled by

questions they cannot answer and mistakes they may have made. Appropriate teaching to junior colleagues should suggest that answers to parents should be ‘I don’t know but I will try to find out’ and to colleagues ‘that’s a good idea, I forgot to check that’.

Advocacy and healthy paternalism

Planning a course of action requires collaboration between the clinician and the patient or parent. This collaboration needs to walk the fine line between patient/parent autonomy and the benefit of the unique knowledge and perspective of the health professional. In this regard, doctors need to act as advocates for the child and gently steer the collaborative decision towards one that leads to the best outcome for the patient and family.

Self-monitoring and awareness of cognitive errors

Most clinical presentations have a differential diagnosis. Healthy self-doubt and consideration of potentially bad outcomes are useful safeguards. Doctors process and synthesise available information in different ways, subject to a number of errors of cognition. These include diagnosis momentum, where doctors accept the perceptions of other clinicians and allow a diagnostic label to stick without making an independent assessment. Doctors may also be affected by anchoring, where they decide on a particular diagnosis relatively early on in the assessment process, and reject subsequent information that does not fit their (premature) diagnosis. When there is some diagnostic uncertainty, the simple act of discussing the case with a colleague can often provide clarity from this cognitive fog. The main stumbling block is the clinical maturity required to recognise feelings of uncertainty and to act on those feelings.

Skill set for paedicatric emergency

Doctors in PEM need to develop a set of practical skills, including the art of communication, clinical examination, procedural skills, teaching and self-preservation. The most important of these is communication, a skill which is intimately entwined with attitudes and which improves with practice.

Effective communication

The groundwork for an effective consultation starts before meeting the parents. Even in busy, seemingly chaotic EDs, parents are watching and listening to those around them, especially members of staff. Individual doctors may be seen in a good light by the way they interact with staff, patients or parents, or because of subtle clues in the way they are introduced by their co-workers. Non-verbal aspects of communication play an important role in the initial impression; the demeanor of the doctor is probably more important than the way he/she is dressed, although reasonable standards in dress code assist in engendering trust.

The essential communication tasks which need to be achieved in a PEM consultation include establishing rapport, gathering information (taking a clinical history), providing information, and demonstrating the attitudes discussed previously.

An important teaching point is that gaining the trust of children starts with getting their parents or caregivers on side. The child is assessing the way their parent relates to this stranger, relying heavily on non-verbal cues to decide whether to trust the doctor. When talking to the child, an approach that illustrates the doctor's interest in them, at an age-appropriate level, is preferred. Rather than trying to playfully examine a child, time spent exclusively in play or developing rapport for the first minute or two, is time well invested. All family members and supporting visitors should be acknowledged individually. In particular, siblings of pre-school age may become disruptive later in the consultation if they do not feel involved, and it may be helpful to give them a minor task or distraction: 'could you please hold my torch until I need it'.

History-taking may be facilitated by clinical pathways, pro formas, or checklists, but diverting attention to the paperwork is a barrier to forming an effective clinical relationship. These checklists are best incorporated into practice by referring to them after the consultation, and going back to fill in the detail later; this is certainly an incentive to remember the information next time.

Several types of information may need to be provided to the patient and parents. These include imparting medical facts, explaining the natural history of a condition, describing or clarifying risk, and providing insight or perspective to the clinical situation. It may help to impart the doctor's own feelings about the clinical risk in an honest and caring way, for example, 'I'm a bit concerned about some aspects, but my feeling is that your child will be completely better in a day or two'. This often goes a long way towards managing unrealistic expectations

or unreasonable demands, provided that a therapeutic relationship has been established first, such that the parents trust the doctor as someone who genuinely cares about their child.

This is perhaps the most important aspect to teach: establishing a trusting relationship built on mutual respect, by exercising the attitudes previously outlined: humility, caring, empathy, compassion, non-judgemental approach, honesty, advocacy and a healthy awareness of one's own limitations. A useful conclusion to any consultation is to invite questions. Giving the parent or patient permission to clarify any areas of concern provides the doctor with the opportunity to deal with any concealed dissatisfaction. It also can result in positive feedback and reassurance that all aspects have been well explained.

Skilful clinical examination

A good physical evaluation helps to illustrate the special skills and knowledge that have been acquired by the clinician, even if these particular skills are not absolutely essential to the examination. For example, testing the tendon reflexes in a child with a minor head injury seldom changes the clinical impression, but adds value to the relationship by demonstrating thoroughness. Just as it is important to acknowledge all family members and friends at the bedside, one may need to ensure that a parent or grandparent in the waiting room is in attendance for the examination and subsequent explanation of findings. This is more time effective than dealing with queries later.

It is important to teach that the examination of infants and toddlers should usually be more opportunistic than systematic, but also that a confident, gentle and structured examination usually inspires confidence and trust.

Mastering procedural skills

A handful of life-saving emergency procedures and a few time-important urgent practical skills need to be addressed in the training of doctors working in a paediatric emergency setting. These include basic and advanced airway and ventilation skills, intravenous and intraosseous access, and screening tests for infection such as blood cultures, lumbar puncture and urgent urine sampling by suprapubic aspiration or urethral catheterisation.

Learning to teach

With increasing competence it is expected that most doctors will adopt the roles of supervising and teaching others. Teaching the teacher poses a new set of challenges but also brings new rewards because of the variety of learning and teaching styles amongst different individuals. The advantage to those doctors acquiring new knowledge, skills and attitudes is that they can pick and choose from a variety of teachers and thereby develop their own style of clinical practice and teaching. Supervising colleagues involves the constant appraisal and reappraisal of clinical risk, if one considers that every clinical interaction puts three people at risk: the patient, the doctor, and the doctor in charge. Inevitably there is an element of trust, based on intuition, based on previous experiences, and based on the answers to a few pertinent questions. The safest alternative for all is for the reviewing doctor to adopt a hands-on approach and personally meet the child–parent unit, until that collegiate trust is well established and well founded.

Self-preservation

Working as a doctor in PEM is emotionally tiring, because it involves caring communication under pressure. Doctors in this environment need to be taught to be careful to avoid burnout. They should think about providing the greatest good to the greatest number of children over their professional lifetimes. A doctor who is impaired by ill health or emotional exhaustion cannot provide good clinical care.

Strategies for avoiding burnout include learning to say no to extra work (even if it brings extra rewards), learning effective time management and prioritising tasks appropriately. Physical exercise and pursuit of non-work enjoyments, and learning to build mutually supportive working relationships with colleagues also helps. Doing meaningful work as a valued member of a supportive team is the goal to strive for.

Putting knowledge into perspective

There is no real limit to the breadth and depth of the knowledge base of PEM. A continuous thirst for knowledge, inspired by the desirable attitudes previously mentioned, can motivate doctors in this exploration, but it is impossible for us to acquire all the medical facts, theories and controversies. The need for doctors to carry around encyclopedic volumes of knowledge in their heads is slowly

diminishing, as the information age starts to live up to its promise of instant availability of highly specific information. Therefore, learning how to access information, knowing where to look for high-quality, trustworthy content, and being able to critically appraise and assess the relative value of the information are becoming vitally important skills.

The value for the diagnostician in exploring a large body of knowledge such as PEM is not so much in being able to instantly recall specific facts, but rather to acquire a low-resolution background of finer forgotten details. A specific part of the clinical picture may then trigger a diagnostic thought process leading to an appropriate search.

PEM is unpredictable in terms of what challenges might come through the door at any moment, yet there are a few recurring themes which make up a large volume of the clinical work. If one excludes minor injuries and serious trauma, more than 80% of the remaining medical presentations are encompassed by six clinical scenarios: fever, breathing difficulty, vomiting with or without diarrhoea, abdominal pain, skin rash and possible seizure. Therefore there is a relatively well-defined scope of learning for those who need to get comfortable with the majority of childhood clinical conditions presenting to an ED, and a much larger range of conditions for those who need to know more.

While experience on its own can be relatively uninformative, reflecting on experiences enables doctors to increase knowledge and gain perspective. Thus, by seeing many patients and learning a small amount from each clinical encounter, doctors can become experienced, mature clinicians. A voluminous clinical workload without reflection and learning without patients are both sub-optimal routes to this desired outcome.

Helping others acquire knowledge, skills and attitudes: modes of learning

A customised local solution for teaching PEM in a particular ED or institution depends on the spectrum of learners (students, nurses, doctors of varying experience and qualifications), incentives to encourage learning, and relative availability of different information resources: people, books and other media.

The requirements of learners vary depending on whether they are nursing students, nurses of various grades, medical students, prevocational doctors, specialists-in-training from either a paediatric or an emergency medicine background, senior trainees in PEM, or consultants with different backgrounds

and strengths. Learners themselves should be asked what they perceive as their general and specific goals and learning objectives, and ideally these goals should be documented and reviewed with their supervisor during the term.

Incentives to encourage learning include the unavoidable performance appraisal that accompanies employment as a hospital doctor, coupled with the concept of gaining a positive job reference for the next rotation. This incentive is relatively subjective, low-impact, and diffuse, lacks immediate reward for good performance, and does not generally identify the acquired abilities of candidates with any degree of detail. Formal testing in the form of tests or assignments adds a dimension of anxiety and workload which may be perceived as a nuisance for both learners and teachers. For this reason, most testing is ad hoc and informal, being conducted during clinical supervision, shift handover and small-group tutorials, usually as an opportunistic, subconscious activity by a range of assessors. The traditional old-fashioned incentive of accountability for the wellbeing of one's patients, and the awkwardness of not knowing what to do in a given clinical situation, is perhaps being overtaken by the trend towards increasing seniority of supervision at the clinical coalface. Staff are additionally motivated to learn by their caring outlook and pride in their work. The option of a safety net should be considered, whereby there are minimum requirements of documented attendance at learning activities. Unfortunately this does not guarantee the acquisition of a minimum standard of attitudes, skills and knowledge.

Learning resources

People

The centuries-old tradition of mentorship and a clinical apprenticeship is no longer available as a one-to-one model, but this can be approximated by arranging for learners and teachers to do clinical work in the same physical space, within sight and earshot of each other. The opportunity exists in many EDs for doctors from a paediatric background to engage in a two-way exchange of ideas with those from an emergency medicine background. We learn a great deal from teaching our team-mates.

Didactic lectures are a way of sharing information with large numbers of learners but are limited by the relative lack of interactivity and are likely to be superseded by technological alternatives which allow the learners to choose the

time and the environment most convenient for themselves. Small-group tutorials offer more interactivity but are limited by availability of protected teaching time away from clinical duties for both learners and facilitators.

Books

Textbooks, handbooks and journals are still a convenient, reliable, low-tech learning medium. The limitations of portability, cost and infrequent updates are still outweighed by the longstanding trusting relationship doctors have with the distributors of high-quality, peer-reviewed content from experts in their respective fields. The challenge for these distributors is to adapt their resources to newer media and models of delivery, rather than duplicate their books onto a computer screen.

Other media

Exciting new modes of learning are constantly being developed and the current generation of learners awaits these new media with anticipation. While there is an abundance of free content available, a fair proportion of high-quality content is by paid subscription or membership. Delivery modes include audio, video, simulation with high- or low-fidelity training models, dedicated software solutions and an ever-expanding suite of web-based resources.

Audio and video reproductions of lectures, discussions, opinions and procedures may be delivered via physical media or via web technology such as streaming, mp3s, podcasts and vodcasts.

Software solutions such as clinical decision support systems and interactive multiple choice question programmes may eventually be replaced by internet-based interactive spaces or platforms combining traditional text, audio and video content with the interactivity of blogs, forums and wikis so that users can contribute to the body of knowledge while experiencing the content in a direct and meaningful way.

Conclusions

Engaging learners in the task of acquiring the skills and knowledge for the safe and rewarding clinical practice of high-quality PEM is difficult. It starts with the challenge of instilling the appropriate attitudes required to propagate these skills

and knowledge in ourselves and in our colleagues.

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SECTION 29

Paediatric Research in the Emergency Department

OUTLINE

29.1

Research in children in the emergency department

Franz E. Babl, Jeremy Furyk, and Emma Tavender

ESSENTIALS

1 Research is essential to establish an evidence base in paediatric emergency medicine and optimise individual treatment.

2 At the core of any research project is the development of a viable research question which drives the study design.

3 A biostatistician should be involved early in the study design.

4 Agreement on key data elements and outcomes, including definitions, feasibility of collection and validity of observations must happen early in the study design.

5 Studies should be appropriately powered to answer clinically meaningful outcomes.

6 Apart from a lack of subjects, the major problem in clinical research projects is poor design.

7 Good clinical research practice is important in all studies.

8 Consent and risk are key ethical issues in all studies involving children.

9 Implementation of research findings should be considered in all studies.

10 Multicentre research can address the difficulty of recruiting adequate numbers of paediatric patients.

Introduction

Research is an important part of emergency medicine as it provides the scientific basis for optimal patient care. Key elements for conducting high-quality, ethical research are the development of a good research question, use of an appropriate study design, adherence to good clinical research practice and an understanding of the ethical basis for research. Study design, the quality of the conduct of the study and its ethics are all intricately linked.

Increasing numbers of emergency physicians, nurses and allied health personnel are involved in research, and trainees may be required to complete research projects during training. Yet few emergency clinicians have formal research training. In addition, over the past years both national and local regulatory requirements have become more complex. Research funding is often difficult to obtain in both the emergency and paediatric setting. Hospitals and departments often have to focus on clinical care with limited resources for dedicated staff, time for research, research assistants or infrastructure for studies. However, ideally research and audit of care are embedded within routine medical care.

In addition, in paediatric emergency medicine the low frequencies of major outcomes in children can lead to underpowered studies and often require a multicentre approach to achieve an adequate sample size. Informed consent is more difficult to obtain in the emergency setting and the ethics of research in children, as a particularly vulnerable group, creates an additional degree of complexity. Further, research findings alone will not change practice, and findings from tertiary centres may be difficult to implement or not be applicable at non-tertiary acute care settings.

However, a number of strategies can be used to overcome the perceived barriers and achieve quality research in children in the emergency department (ED).

Research science

All research should have a sound scientific basis. Getting the science right is essential before starting any project.

Research question

The most crucial component of every project is the research question. The

question defines all the other elements of the research design, including the hypothesis and objective of the project.

A good study question should be:

- original or add significantly to what is known
- relevant and important
- feasible, based on resources, skills, time, subject availability
- ethical
- plausible, that is there should be a scientific basis for the question
- clearly defined.

Determining these factors requires clinical perspective and a detailed literature search. In clinical research the most relevant or important questions are those that actually change clinical practice.

A question can be defined in terms of the acronym PICOT:

- Population (who should be in the study?)
- Intervention/indicator (what is the intervention or exposure of interest?)
- Comparator (what is the reference to compare the intervention to? what is the gold standard? comparison vs. baseline vs. control group?)
- Outcome (what is the outcome of interest?)
- Time frame (over what time period?).

The outcome measures are often the most difficult to determine and must be clearly defined, ideally with a single primary outcome to answer the study question and further secondary outcomes, which may be multiple, to expand on this question. These should be determined in detail in advance prior to submission of the protocol for ethics review.

Literature review

A literature review should provide a sense of what research has already been undertaken and how the planned research will contribute to the field, and should be undertaken prior to commencing any study. Literature databases like Pubmed, Medline, Google scholar internet search engines, EMBASE and CINAHL (Cumulative Index to Nursing and Allied Health Literature) are useful but may initially be overwhelming. Helpful starting points can be standard textbooks, the

Cochrane Library (www.cochranelibrary.com), BestBets (www.bestbets.org), BMJ Clinical Evidence (http://clinicalevidence.bmj.com) and the assistance of a medical librarian. There are a number of systems to grade the strength of evidence of the literature. A good starting point for grading the quality of evidence and the strength of recommendations that can be applied across a wide range of interventions and contexts is provided by the Grading of Recommendations Assessment, Development and Evaluation (GRADE) working group GRADE.[1] If a literature review determines that the research question has already been answered, to a suitable standard, in a population generalisable to your own, then undertaking the planned research may not be necessary.

Types of studies

Studies can be grouped in a number of ways. Studies can be descriptive, analytical or interventional. Intervention studies can either assess the efficacy of the intervention (does it work in a study?) or the effectiveness (does it work in the everyday ED situation?) (Table 29.1.1).

The ethics of medical research

Following atrocities during the World War II, written codes of medical ethics were developed, such as the Nuremberg Code,[2] the Declaration of Helsinki[3] and the Belmont Report.[4] The key principles developed in these documents still underpin most ethical guidelines.

Key principles

The key principles of research ethics include respect for persons, beneficence and justice as well as ‘research merit and integrity’.

Respect for persons recognises the value of autonomy to an individual. Participants must have the power to make their own decisions, if possible. Having respect for persons requires due regard for beliefs, customs and cultural heritage of individuals as well as respecting privacy and confidentiality. Consent is the key element of respect. The consent to participate in a research project must be free and informed. Free consent implies a voluntary choice not influenced by external coercion, pressure or inducement. Informed consent

implies that the participant has sufficient information and adequate understanding of the proposed research, and, particularly in the paediatric context, is sufficiently mature to understand the consequences of the decision to take part in the study.

The principle of beneficence includes the concept of maximising possible benefits and minimising possible harm while avoiding unnecessary burdens. Non-therapeutic research or research where the risk of harm is possibly greater than the risk of benefit is not beneficent.

The principle of justice implies there should be no inequality in sharing the burden or risks and the benefits of research. The most obvious examples of injustice are where research benefits or risks are unevenly distributed amongst the wealthy and poor, or conducting research exclusively in minority populations to benefit non-minority populations.

Balancing the principles of respect for persons, and justice is not always straightforward. While rightfully considered a core principle of clinical research, it is increasingly recognised that prospective informed consent may not be possible in some circumstances in emergency situations. In this circumstance the ethical principle of respect for persons must be balanced with the ethical principle of justice, as excluding patients on the basis of inability to consent is denying these individuals the opportunity to participate in research from which they or others may benefit.

Ethics of research involving children

Due to the exploitation of children prior to the Nuremberg Code there is added complexity relating to research in regard to children. This relates to the key component of informed consent. The age at which a child develops the maturity to understand, give informed consent or accept risk for altruistic reasons is not predefined by chronological age. There is variability in this, based on complexity of the proposed intervention. It is important to note that ethical standards and criteria for study participation evolve over time, and what is acceptable now may be controversial or unacceptable in the future, just as previous standards may now be considered questionable. This is particularly so in relation to children.

In emergency research there are circumstances where obtaining informed consent prior to enrolment in research is not possible.[5] Examples of this may include interventions for life-threatening emergencies, such as cardiac arrest,

major trauma, status epilepticus or status asthmaticus, particularly when the interventions have a narrow therapeutic window and treatment delays may be detrimental to the patient. There is a community expectation that emergency management of children is evidence based, and guided by quality clinical trials. Even if a parent is immediately available, there may be insufficient time, or the parent may not be in the appropriate state of mind to provide valid informed consent.[5] In these circumstances it may in fact be unethical to seek informed consent, risking delaying potentially life-saving interventions, or producing an unnecessary burden or anxiety on parents or caregivers, in a vulnerable position.[6] Potential for coercion in this circumstance should not be underestimated.

Ethics committees approve research without consent only in exceptional circumstances. In Australia, research without consent is permissible, providing the following nine conditions are met: [7]

- Involvement in the research carries no more than low risk to participants.
- The benefits from the research justify any risks of harm associated with not seeking consent.
- It is impracticable to obtain consent.
- There is no known or likely reason for thinking that participants would not have consented if they had been asked.
- There is sufficient protection of their privacy.
- There is an adequate plan to protect the confidentiality of data.
- There is no known or likely reason for thinking that participants would not have consented if they had been asked, in case the results have significance for the participants' welfare there is, where practicable, a plan for making information arising from the research available to them.
- The possibility of commercial exploitation of derivatives of the data or tissue will not deprive the participants of any financial benefits to which they would be entitled.
- The waiver is not prohibited by state, federal, or international law.

Similar regulatory provisions exist in the USA, UK and New Zealand.

Table 29.1.1

Types of studies

Study groups and question	Study type	Description	Advantages	Limitations	Level of evidence
Descriptive					
Describes the distribution of a certain variable such as an exposure/disease/symptom, e.g. 'How many children vomit after intranasal fentanyl?'	Case series	Case reports without control of interventions for individuals with the same exposure.	May develop hypothesis for later prospective trials.	At risk of selection bias. Data often obtained retrospectively, inaccurate, incomplete data or measurement bias.	IV
	Cohort studies	Prospective or retrospective longer-term follow-up of patients. Can be both observational and/or case-controlled.	Allows examination of multiple exposures on a single condition and study of rare conditions.	At risk of selection bias. Interventions occur in a non-experimental way.	III-2
	Cross-sectional studies	At given time point population studied to determine prevalence (not incidence) of disease.	Quick, relatively inexpensive.	Causal relationship cannot be established.	IV
Analytical					
Evaluate associations to discover cause and effect relations, e.g. 'What factors are associated with vomiting after intranasal fentanyl?'	Randomised controlled trials (RCT)	Randomly allocates participants to intervention or control group.	Definitive method to assess effect of intervention – gold standard. Randomisation minimises selection bias.	Difficult to conduct and large numbers may be required. Usually only tests one intervention at a time.	II
	Case-controlled studies	Recruit disease vs. non-disease and assess exposure status between groups.	Used for rare diseases.	Prone to selection bias. No information on risk of disease is able to be calculated.	III-3
	Crossover studies	In stable disease can alternate treatments in same patients.	Reduces sample size. Patients act as own control.	Each participant needs to be in the study for longer. Can have high drop-out rates, which significantly reduces study power.	III-2
Other					
	Pilot study	Small scale preliminary study conducted prior to a definitive study.	Can be used to assess feasibility, cost, time, adverse events, and to identify problems with study protocol. Often used to derive mean and standard deviation for power calculations.	May delay starting of definitive study. Effort required to undertake pilot may be similar to that of the definitive study.	
Literature review					
	Review	Summary of available evidence from literature.	Should occur prior to undertaking any interventional study.	May be incomplete due to inadequate search strategies andvariable use of 'formal' critique of evidence.	
	Meta-analysis	Combines single studies and conducts a combined analysis.	Used in an attempt to find relationships not apparent in original studies, often due to inadequate power of single studies for uncommon clinically important outcomes.	Subject to limitations in design of the original studies. Different studies have different methodology, a large amount of heterogeneity makes it impossible to combine studies in a formal meta-analysis. Meta-analysis is at risk of publication bias as positive studies are published more than negative studies.	Depends on studies contained. If studies are level II then the meta-analysis is assigned level I.

In situations when prior informed consent is deemed not possible, options include a waiver of consent, or retrospective or deferred consent. Deferred or retrospective consent involves discussing the research with the parent or guardian as 'soon as possible' after enrolment, often after the intervention has been instituted. Consent in this instance is for continued participation in the study and use of data. Advantages are that there is minimal disruption to clinical care, while still allowing for detailed explanation of the purposes of the trial and the opportunity to ask questions.

Assent by the child is a requirement used in some countries (e.g. USA), although formal consent is still required from the legal guardians.

When consent is considered possible, there are in general four recruitment scenarios for children presenting to the ED who may be enrolled in research studies:[7]

1. Infants/toddlers without the capacity to understand or take part in discussions regarding a research project and whose parents/guardians are approached for consent.
2. Children able to understand some relevant information and take part in limited discussion about the research, but whose consent is not required.

Only parent/guardian consent is required for these children.

3. Young people of developing maturity, able to understand the relevant information but whose relative immaturity means they remain vulnerable. The consent of these young people is required but is not sufficient to authorise research, therefore requiring additional parent/guardian consent.
4. Young people mature enough to understand and consent and not vulnerable through immaturity in ways that warrant additional consent from a parent or guardian.

As children can be considered a vulnerable population and have limited capacity to consent, the degree of risk they can be exposed to needs careful consideration with the potential benefits of the research and societal benefit. In general, risk is classified in degrees, and the degree of risk acceptable depends on the importance of the study, and the likelihood of any direct benefit to the child.

In order that some important research involving children may legitimately be carried out, most jurisdictions deem that it is acceptable to have some degree of risk for the child who is unable to consent where there is no direct benefit accruing to that child, provided certain limitations are adhered to.

In addition, in the ED setting there is the added complexity relating to gaining consent for a study at the time of an acute injury or illness. This makes it more difficult to fully assess the child's maturity level and understanding and to appreciate the potential for researchers to apply covert pressure to a reluctant child.

Ethics review process

The implementation of national guidelines and principles to individual projects is left to the discretion of human research and ethics committees (HRECs). HRECs are administered locally by research institutions or hospitals, or in some countries via a national independent structure. Research other than low or negligible risk research must be reviewed by a local ethics committee. Ethics committees are usually composed of representatives from clinical (doctors, nurses, etc.), legal, research and community (laypeople and clergy) groups. All research must be approved in writing by the sponsoring institution before it can commence. Research requiring HREC approval may not start until there is clear

written approval from the committee.

The primary objectives of an HREC are to assess the ethical principles by which research projects in humans are proposed and conducted, protecting the welfare and rights of the research participants and to facilitate research that is or will be of benefit to the researcher's community or to humankind. HRECs may also consider all matters relating to project design, technical feasibility and any other ethical implications associated with each project. Any request for waiver or deferred consent is obviously considered very carefully by HRECs, requiring full and detailed justification, as protection of participants' rights and welfare rests solely with this ethical review process.

Low ethical risk projects, such as clinical audits, may be regarded by the community as an essential part of clinical practice, for example an analysis of the types of ED presentations that did not wait to be seen. These projects should still be presented to an HREC; indeed, journals require documentation of this for publication, or a statement as to why such approval was not required locally. However, in most jurisdictions such projects would undergo an expedited review, without the need for a full ethics submission and the consequent delay. As with all research, in these circumstances it is still important to plan the research carefully, as the project will not be of benefit if a researcher has not collected and analysed the data in a systematic and rigorous manner.

HREC approval is often contingent on other governance processes. The hospital lawyer and hospital insurer may have to sign off on high-risk projects and trials will need prospective registration and notification to national pharmaceutical regulatory bodies may be required in clinical drug or device studies. A study can only be commenced once HREC approval has been obtained and only HREC-approved study documents may be used during a study.

The practice and governance of research

Well-conducted clinical research is more likely to produce results that are closer to the truth and can be trusted to inform clinical practice. There are basic professional standards in the conduct of research which must be met during this process.

Research documents

Good documentation is a key to a successful study. Documents should be kept together, complete, dated and secured. There should be a clear trail of all data from the point of data collection, collation into a database through to publication. This trail may be scrutinised if there is any question about the veracity of a research finding. Clear documentation is also essential to reduce the chance of error and to keep the project running smoothly.

Research protocol

The key document for any study is the research protocol. It sets out why a trial should be run, acts as an operations manual and is the scientific design document for the trial. It is submitted to the HREC at the time of approval application along with the case report forms (CRFs), patient information statement and consent form. The research protocol outlines in detail the research question being asked, background and rationale for the study, the design and methodology by which the question will be addressed, secondary objectives, primary and secondary outcomes, statistical considerations including sample size and power calculations. There should also be a discussion of any ethical implications of the study being undertaken. Templates for protocols can be found on the website of the Standard Protocol Items: Recommendations for Interventional Trials (SPIRIT) group.[8]

Case report forms

The CRF is designed to record all of the required information as defined in the protocol for accurate analysis. Badly designed CRFs negatively affect the quality of data analysis and the ability of the project to answer the research question. CRFs should be designed concurrently with the protocol, preferably with input from a biostatistician and should be piloted prior to study commencement. All data collected for the study should be recorded directly, promptly, accurately and legibly.

Patient information statement and consent form

In order to gain informed consent from a potential research participant, research projects require patient information statements in plain language and consent forms that must be approved by the ethics committee prior to their use. These are intended to outline the proposed study in language laypeople can understand. Good patient information statements are difficult to write and poorly written

ones may cause delays in obtaining ethics approval.[9]

Study document file

The study document file should be kept in the ED administrative area and contain copies of all documentation relevant to the study in an orderly and systematic fashion. Good data handling and record keeping ensures a ‘paper and electronic trail’ that testifies to the accuracy of the reported data, as a study may be audited even years after it has finished.

The research team

Research is a team effort, even for a small project. Good research practice involves having an effective team of researchers and collaborators (such as nurses, junior doctors and allied health workers) with a clear understanding of who is responsible for what and who reports to whom. Even in small studies, biostatisticians should be involved early in the design stage. They will help guide researchers to determine study design and sample size in addition to their role in database creation and data analysis.

Depending on the size and type of study, teams may include, in addition to clinicians, expert researchers, statisticians, pharmacists and/or technicians, health economists, trial coordinators, data managers, data entry staff and research nurses or assistants.

Larger studies may require several groups or committees such as a trial management group (to manage the trial on a day-to-day basis), trial steering committee, data monitoring and/or safety committee and data management committee. These committees should have regular minuted meetings.

Databases and analysis

Good data management should be carefully planned from before the first data are collected through to the analysis of data; otherwise implausible values and inconsistencies are carried through to the analysis phase, where it is very difficult to correct errors. A database is a computer software program where data are entered and stored in a table or file, which is called the data file. Each study patient should have a unique identifier different from the patient’s hospital number to ensure confidentiality. Even for simple databases, a ‘coding manual’ should be created to explain variable names, describe the variables and their

units, specify the number of decimals, etc. Studies are often performed over a period of time and what seems obvious at the beginning of the process may become cryptic at the analysis stage. Study data must be stored securely and saved electronically with password protection.

Reporting guidelines

The reporting of clinical trials has a recommended, standardised approach, outlined by the CONSORT Statement, an evidence-based format aimed at improving the quality and integrity of reporting for RCTs. CONSORT consists of a checklist and flow diagram for reporting an RCT.[10] The flow diagram provides readers with a clear picture of the progress of all participants in the trial, from the time they are randomised until the end of their involvement. In addition to the SPIRIT documents listed above,[8] these documents are also very helpful at the design stage of the trial and ensure that the research protocol is comprehensive and logical.

There are also a number of reporting guidelines for other types of studies such as observational studies, systematic reviews or quality improvement studies. Information about reporting guidelines, including key checklists and flow diagrams, is listed at the EQUATOR website.[11]

Key regulatory documents

International Conference on Harmonisation of Good Clinical Practice guidelines (ICH-GCP) [12]

ICH-GCP, also known as GCP or GCRP (good clinical research practice), is an overarching ethical and quality standard for the design, conduct, performance, recording, analysis, monitoring, auditing and reporting of clinical research. It also includes statements on protection of human rights as a subject in clinical trials. It was developed as an international quality standard which individual jurisdictions can transpose into regulations for clinical trials involving human subjects; this has occurred widely.

National- and state-based regulations

Within jurisdictions, either national or state based, GCP principles have generally been embedded into statements and regulations within which all research conducted must be in accordance. Researchers, and their institutions,

should be aware of these and have a working knowledge of them prior to undertaking clinical research.

The aim of these documents is not only to increase the scientific quality and hence the veracity of findings but also to ensure research is conducted in an ethically responsible manner and that the findings are verifiable. Being compliant with these documents assures the public that the data and reported results are credible and accurate and that the rights, integrity and confidentiality of human research subjects have been protected.

Project registration

Since July 2005 all clinical trials must be registered on a Clinical Trials Register before the enrolment of the first participant. The guidelines of the International Committee of Medical Journal Editors (ICMJE) state that any trial must be registered in order to be published in any of their comprehensive list of journals.[13] The purpose of trial registration is a greater efficiency by reducing unnecessary duplication of research effort, better compliance, avoiding selective reporting of results and a greater assurance that all clinical trials' reports are presented, including those with negative results. There are several clinical trial registers worldwide including ClinicalTrials.gov and the Australian and New Zealand Clinical Trial Registry (ANZCTR).[14]

Privacy and confidentiality

Clinical research almost invariably involves the use of personal information. Patients provide information with the expectation that it will be treated confidentially and the unauthorised disclosure of such information may be a risk to the subject which should be considered in any research project. Similar to medical records in general, patient privacy laws apply to research records.

Implementation research: an emerging research field

Despite the increasing availability of high-quality research evidence, it is common for this evidence to be inconsistently used in decision making across many clinical groups.[15] It can take on average 17 years for research to be incorporated into practice and in some cases the research evidence may never be

completely implemented.[16] Unwanted variations in the care received by children in EDs include antibiotic use and management of asthma, fever and bronchiolitis .[17]

Implementation research is an emerging field of health services research and is the scientific study of methods used to promote the uptake of research findings into practice and policy and to reduce inappropriate care.[18] It can contribute to reducing variations in practice and improving health and efficiency outcomes by studying the determinants of practice change (barriers and enablers), processes and effects of implementation efforts and providing evidence about designing, selecting and improving implementation interventions.[19] The Cochrane Effective Practice and Organisation of Care (EPOC) Group conduct systematic reviews of implementation interventions to improve health professional practice and the organisation of health care services.[20] Interventions such as audit and feedback,[21] local opinion leaders[22] and computer-generated reminders[23] have been shown to be effective at improving professional practice and patient outcomes. However, no single implementation strategy is effective in all healthcare settings and the context and factors known to influence practice change need to be addressed for the intervention to be successful.[24] Interventions tailored to prospectively identified barriers or enablers to change are more likely to improve professional practice.[25]

Implementation science promotes a systematic approach to designing strategies or interventions to improve evidence-based practices including: (1) identifying behaviours contributing to the evidence–practice gap, (2) identifying key determinants of these behaviours and the desired behaviour change using theoretical frameworks, (3) selecting evidence-based intervention components that target these key determinants and (4) evaluating the effectiveness of the intervention including an assessment of mediators of change (whether the intervention components modified the determinants) and a thorough process evaluation to determine how well the intervention was implemented.[26,27]

The ED environment is complex and has unique characteristics that can have an impact on its responsiveness to change and capacity to implement evidence into routine care. There are further challenges in paediatric emergency medicine as most children are cared for in mixed EDs focused largely on adult care with many having limited paediatric emergency expertise.[28] Implementation research in the paediatric emergency medicine setting is currently limited and recommendations for future research include the following:

1. A focus on areas with important evidence–practice gaps
2. An assessment of barriers and enablers, using theory, to inform the choice and design of implementation interventions
3. Evaluation of interventions using rigorous study designs, e.g. cluster randomised controlled trials
4. An explicit description of the nature and content of these interventions undergoing evaluation to enable replication in future studies and to further the science of implementation research in this setting.[29]

Multicentre research

Some of the difficulties in emergency research in children, such as the low frequency of major outcomes or limited applicability of findings, can be overcome by cooperating with other institutions. This recognition has led to a number of cooperative research networks, initially in North America (Pediatric Emergency Research Canada [PERC www.perc-canada.ca] and Pediatric Emergency Care Applied Research Network [PECARN www.pecarn.org]), in Australia and New Zealand (Paediatric Research in Emergency Departments International Collaborative [PREDICT www.predict.org.au] and in Europe (Research in European Paediatric Emergency Medicine [REPEM www.sites.google.com/site/repemnetwork/] and Paediatric Emergency Research in the United Kingdom and Ireland [PERUKI www.peruki.org]). Recently, these networks have started to collaborate in the Pediatric Emergency Research Networks (PERN www.pern-global.com) which allows the conduct of studies across a large number of EDs and across many countries.

Funding research

Finding funding for research is a challenge. Obtaining funding is often a stepwise process from small local grants through to large grants from international bodies, or for multicentre research. Obtaining such funding requires a robust research question and study design and a strong track record. Evidence of seed funding and pilot data is also useful. Seed funding may come from professional societies, research institutions and private practice funds. Infrastructure support may come from the hospitals, universities or research institutes. Leveraging infrastructure support, or smaller grants, off larger grants is a useful strategy. Philanthropic funds are also available, though these are

increasingly competitive. Collaboration with other successful groups and researchers is another key to funding success.

Controversies and Future Directions

1. Trainees are often expected to undertake research during their training. Due to time constraints in training this encourages low-level studies and biases against randomised controlled trials (RCTs) or systematic reviews. Supervisors should offer research training for trainees. Research education material exists to help both supervisors and trainees.[30]
2. Most public hospital EDs are poorly resourced in terms of funding, staffing and infrastructure for research in paediatric emergencies. There is a pressing need to create more academic infrastructure based either within EDs or via affiliated universities or research institutes.
3. The development of cross linkages between EDs and with researchers outside of paediatric emergency medicine through national research networks has helped build capacity in EDs. It has also provided increased exposure of paediatric emergency research to funding agencies which may be the basis for developing high level research studies with dissemination of both research projects and their results into more EDs.
4. Expansion of cross-linkages between international research networks is being developed and should further enhance the dissemination of evidence-based emergency medicine.

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SECTION 30

Adolescent Medicine in the Emergency Department

OUTLINE

30.1

Adolescent medicine in the emergency department

Katherine Barton, and Donald Payne

ESSENTIALS

1 Establishing rapport is essential for a successful adolescent health consultation.

2 Interviewing adolescents in a quiet, private setting, removed from the main emergency department, is of great advantage.

3 It is important to see adolescents on their own for part of the consultation.

4 Explanation of confidentiality is essential.

5 Discussion should be professional and in language the young person is able to understand.

6 Using the HEADSSS framework will help exploration of relevant psychosocial issues.

Introduction

Many health professionals find working with adolescents challenging. Communication can be difficult, the priorities of adolescents are different from those of adults, and issues of consent, confidentiality and privacy take on particular significance.[1,2] Providing care for adolescents also takes time, which is not always readily available in the emergency department (ED). The perceived discomfort that some professionals experience working with adolescents usually

reflects a lack of training in this area.[3] However, as with any area of medicine, specific training can lead to an increase in clinicians' competence and confidence in dealing with adolescents.[4] In the last few years the Royal Australasian College of Physicians (RACP) has made significant progress in developing specific training resources focusing on the health of young people, which are available through the RACP website (https://www.racp.edu.au/trainees/advanced-training/advanced-training-programs/adolescent-and-young-adult-medicine).[5]

Keys to working effectively with adolescents include developing confidence in talking to adolescents and understanding adolescent development.

Although different age ranges have been proposed, it is more appropriate to think of adolescence as a process – during which an individual moves from being a dependent child to an independent adult.[2] The rapid developmental changes that occur during adolescence include the obvious physical changes of growth and puberty as well as the less well-recognised cognitive, emotional and social changes. A key task of adolescence is for individuals to establish their own identity and self-image. This involves developing independence from parents, forming relationships outside the family, challenging authority and experimenting with different behaviours, some of which can pose a health risk. Health professionals working with adolescents need to acknowledge this normal process and be aware of the impact of emerging adolescent behaviours on health outcomes, as well as the effect of illness on normal adolescent development.[2,6]

Adolescent health problems in the emergency department

Whether the setting is an adult, mixed or paediatric ED, all emergency doctors are likely to see adolescents in their daily practice. The leading causes of death among adolescents and young adults are motor vehicle collisions and suicide.[7] The most common causes of morbidity include injuries (both intentional and non-intentional), mental health problems, drug and alcohol misuse and sexual health problems (Box 30.1.1). In addition, the number of adolescents and young adults growing up with chronic diseases of childhood is increasing as a result of improved treatment of these conditions.

Many presentations to ED for primary physical problems are linked with psychosocial issues or with health risk behaviours, such as drug and alcohol use, unsafe sex and physical risk-taking. Clinicians working in ED must therefore be aware of these underlying risk factors and feel confident in being able to discuss

them and liaise with local youth health services as needed.

Mental health issues, particularly anxiety, depression, deliberate self-harm and suicide attempts have been increasing in adolescents.[7] Presentations to the ED of young people in crisis are therefore also increasing. Taking a psychosocial history and performing a suicide risk assessment is now an essential skill for the ED physician. Knowledge of local hospital and community mental health clinics, telephone help lines and websites (Box 30.1.2) allow the emergency physician to provide useful information to adolescent patients and their families.

Box 30.1.1 Health issues for adolescents presenting to medical care[1]

***Injuries*:** motor vehicle, bicycle and pedestrian injuries, work-related injuries, falls, assaults, poisonings
***Mental health issues*:** anxiety, depression, self-harm, suicide attempts, psychosis, personality disorders, eating disorders
***Drug related*:** alcohol, illicit drug use
***Infectious diseases*:** e.g. influenza, meningococcal infection, pneumonia
***Chronic disease*:** asthma, cystic fibrosis, diabetes mellitus, inflammatory bowel disease, chronic pain/fatigue syndromes, allergies, overweight/obesity
***Sexual health*:** pregnancy, sexually transmitted infections

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Australian Institute of Health and Welfare (AIHW). Young Australians: their health and wellbeing 2011. Cat. No PHE 140. Canberra: AIHW, 2011. www.aihw.gov.au/publications

Box 30.1.2 Useful websites

Kids Helpline www.kidshelp.com.au
Lifeline www.lifeline.org.au
Dr Yes www.dryes.com.au
Headspace www.headspace.org.au
Beyond Blue www.beyondblue.org.au and www.youthbeyondblue.com
Reach Out www.reachout.com.au
Australian Drug Foundation www.adin.com.au
Family Drug Support www.fds.org.au

Where's Your Head At? www.drugs.health.gov.au
Sexual Health www.redaware.org.au
Legal Information www.lawstuff.org.au

Presentations to the ED of adolescents relating to substance misuse are also increasing such as intoxication, overdose (deliberate or accidental), and related injuries. Alcohol is commonly involved, however other illicit drugs like marijuana and amphetamines should be considered, and their toxicological features assessed.

Presentations for sexual health issues may not always be clear (e.g. females with abdominal or pelvic pain) but should be considered, particularly as the incidence of sexually transmitted infections (STIs), particularly chlamydia, has been increasing in adolescents.[7] Taking a sexual history, including enquiring about contraceptive use and performing sexually transmitted infection screens are now an important part of the ED consultation.

Somatic complaints in adolescents such as fatigue or chronic pain are common and may present to the ED.[8] Adolescents commonly manifest psychological distress with physical symptoms. Younger adolescents more commonly present with chronic abdominal pain without a known cause, whereas older adolescents may present with headaches or fatigue. Although not primarily an emergency, adolescents may present to the ED in crisis due to acute exacerbations of pain or the family's frustration about a lack of diagnosis and appropriate management.

Ensuring that a thorough physical and psychosocial history has been taken, and that appropriate investigations have been performed (or arranging them) can complete the organic screen and is very useful before referring. If there are associated mental health issues, or significant social impairment (e.g. school non-attendance), a multi-disciplinary adolescent medicine team is most appropriate for these patients, who need a rehabilitation model of management. Sending the adolescent home after a thorough assessment, with a plan for future investigations and referral, as well as an acute pain management plan, can help to prevent future re-attendances and allay patient and family anxiety.

The approach to the adolescent in the emergency department

Establishing rapport

Establishing rapport is essential for a successful adolescent health consultation. Empathy, trust and respect are important keys in any doctor–patient relationship but especially so for adolescents. Although establishing rapport in ED can be challenging, it is definitely possible. Adolescents may present to an adult department, where they could find themselves surrounded by much older patients, or to a paediatric department, full of infants, toddlers and understandably anxious parents. The setting may therefore have a significant impact on the consultation.

The ability to see adolescents in a quiet, private setting, removed from the loud, sometimes chaotic environment of the main ED, is of great advantage. Doctors should be professional, open, honest and themselves. Introduce yourself to the adolescent first, and ask him/her to introduce the other family members present. This puts the adolescent at the centre of the health consultation and emphasises that they are an individual, rather than a medical problem.[9,10]

Seeing adolescents alone

It is important to see adolescents on their own, separate from parents, for at least part of the consultation. This helps to establish rapport and trust and optimises the chances that the young person will talk openly about sensitive issues. Adolescents often contribute little to the history when parents are present. However, on their own, they are much more likely to open up. Parents may find it difficult to separate from the adolescent for the health consultation. This process is helped by explaining that it is routine practice to see adolescents on their own and emphasising the importance of the young person beginning to take responsibility for his/her own health. Spending time with parents on their own afterwards (after discussing with the young person what you will tell them) may alleviate their anxieties.[11]

Unless the urgency of the situation dictates otherwise, it is helpful to begin the consultation by asking adolescents one or two general questions rather than immediately focusing on the medical presentation. This helps to put them at ease and shows that you see them as a person first who happens to have a medical problem.[9]

Psychosocial screening

Adolescence is a critical time when health behaviours are established, and doctors can have an important role in reinforcing positive health behaviours. Presentations to ED should be seen as an opportunity for health promotion, especially considering many young people do not have a local GP.[12] The ED may be the only place where an adolescent (particularly vulnerable youth) receives health care.[13,14]

Although more pertaining to a physician consultation in an outpatient clinic setting, psychosocial screening can be a very useful tool in the ED consultation. Due to the fact that many young people present with mental health issues, it is important to assess the psychosocial factors that may be contributing to the current presentation. This is especially important when young people present to the ED with mental health issues, alcohol and drug issues and chronic unexplained problems such as pain and fatigue. Screening for mental health issues, suicide risk, sexual health issues and substance use in the ED has been shown to be feasible – it is not disruptive to workflow, and is highly supported by adolescents and their families, and may prevent future re-presentations.[15,16]

It is helpful to summarise in advance the types of questions you plan to ask and to explain their relevance to health outcomes. Normalising the process by explaining that you ask all adolescents these same questions can help make the young person feel more at ease. Beginning with an explanation about confidentiality is essential.[6] Sensitive questions can be more easily asked using a third person approach, such as ‘Do any of your friends use marijuana?’ before progressing to asking about the patient themselves.[9]

Using the HEADSSS framework

HEADSSS is used widely around the world; it acts as a helpful guide for clinicians when interviewing adolescents and has been shown to be accurate and reliable and maximise communication.[9,14] HEADSSS begins with relatively unthreatening questions about home, school and activities. These have the dual purpose of gathering information and allowing time to develop rapport. However, difficulties in these areas (e.g. prolonged school absence, no hobbies or interests) may be a reflection of problems in other areas. As mentioned earlier, mental health problems, such as anxiety or depression, and health-risk behaviours, such as smoking, alcohol and other drug use, are common in adolescents and should always be considered. The HEADSSS screen has been updated several times since it was first published, with the latest version,

incorporating screening for problematic internet and social media use. It is available online at http://contemporarypediatrics.modernmedicine.com/contemporary-pediatrics/content/tags/adolescent-medicine/heeadsss-30-psychosocial-interview-adolesce?page=full.[9,17]

Doctors may find it difficult to communicate with adolescents with regard to health-risk behaviours. It is important to remain empathic and non-judgemental, and to convey concern about risk taking and its potentially harmful consequences. It is also important to frame discussions in language which the young person is likely to understand, and to avoid using medical jargon.[2]

Tailoring your approach depending on the developmental stage of the adolescent allows you to take a thorough history more easily.[2,9] For example, younger adolescents (with more concrete thinking) respond best to simple, closed questions. Older adolescents are able to understand more abstract concepts and can more easily answer open-ended questions and contemplate the future. This becomes important when discussing adherence to treatment and health-risk behaviours. Younger adolescents are only able to comprehend short-term consequences, whereas older adolescents may be able to contemplate the longer-term implications of their behaviour.

At times, adolescent patients will be difficult to engage. This is especially so considering that many presentations to ED will be in relation to sensitive issues such as psychosocial problems or substance abuse. It is often a situation of high stress for both the young person and their family. Acknowledging this and demonstrating patience are important.

Confidentiality

One of the barriers to adolescents seeking medical care is a perceived lack of confidentiality. Establishing confidentiality is thus essential at the beginning of the adolescent consultation. It is important to explain that, whilst you may discuss aspects of their case with colleagues and write in patient notes, you will not discuss things with their parents without their permission. It is also essential to explain the limitations of confidentiality. The disclosure of any activity that puts the patient (or others) at serious risk of significant harm (such as suicidal thoughts or physical/sexual abuse) cannot remain confidential. Clearly establishing confidentiality at the beginning of a consultation fosters honest and open communication between the doctor and the young person. Adolescents who

are assured of some degree of confidentiality are more likely to speak frankly.[18] In practice, this increases the chances of being able to address a range of health-related issues, thus opening up the possibility of providing effective health care.

The mature minor principle

The legal age for consent to treatment varies between states within Australia.[19] However, when working with adolescents, regardless of their age, the mature minor principle can always be employed.[10,19] This states that a young person can consent to treatment without parental knowledge if they are considered to be competent to do so by their treating clinician.

The clinician must be confident that the young person understands the proposed treatment, its benefits and risks, and is able to make an informed decision. An adolescent's competence to consent to treatment will clearly depend on the complexity of the treatment proposed. In practice it is best to try to encourage adolescents to involve their parents, or another adult whom they trust, in any treatment decision.

However, sometimes adolescents will choose not to involve their parents. In these circumstances it is sensible for clinicians to consult with another colleague rather than taking sole responsibility for difficult decisions. Clinicians have a duty of care to provide the best treatment for their patients at the time of presentation. In addition, for adolescents, it is particularly important that their experience of health services is a positive one, thereby optimising the chances that they will feel confident to access appropriate health services in the future.[12]

Physical examination

The examination of the adolescent patient is essentially the same as the examination of an adult patient, from general inspection and observation to a systematic approach to each of the major systems. However, clinicians must be sensitive to the developmental stage of the young person. Priority should be placed on privacy and making the young person feel comfortable. Simply explaining what you are about to do can ease embarrassment. It is important to find a chaperone to be present during an examination. Accurate staging of puberty may be required in certain cases. At the end of the examination, the findings should be explained in language the young person is able to understand. If the examination is normal, a simple explanation of this fact will be very

reassuring for a young person.[2]

Linking adolescents with community follow-up

Given that mental health problems, health-risk behaviours and sexual-health problems are common among adolescents and young adults presenting to ED, it is useful for clinicians to have access to information about youth-friendly services available in the local community. These include sexual-health clinics, drug and alcohol centres, hospital and community mental-health services, and drop-in centres. Some may require a referral, other may accept self-referrals. The Medicare Better Access programme currently allows Australian GPs to refer patients to a private psychologist for up to 18 sessions a year with a Medicare rebate. Clinicians should be able to provide adolescents with relevant written information and website addresses (Box 30.1.2).

Summary

Adolescents require clinicians to have a different approach compared to paediatric or adult patients. Priorities should include privacy, confidentiality and adequate time spent with the young person. The reasons for presenting to ED are often complex. Performing a psychosocial screen can be very rewarding. Many young people have no GP and therefore ED visits should be seen as an opportunity for preventive health screening, in addition to acute management. Training in adolescent health for all staff working in ED will be of benefit to both patients and staff.

Controversies and future directions

Where should adolescents and young adults be seen?

There is no consistent policy between different hospitals regarding the upper age limit for attendance at a paediatric emergency department (ED). Some departments will not treat adolescents once they reach the age of 14, while others continue to see young adults into their early twenties. Regardless of the age of the patient, it is important that each ED has the staff and facilities to provide developmentally appropriate care for adolescents and young adults.

Facilities for managing aggressive, intoxicated or distressed adolescents

Presentations of aggressive or distressed adolescents (with or without mental health or substance-use issues) are becoming more frequent in the ED.[20] ED staff need to be able to manage these patients with access to appropriate facilities, such as secure safe rooms that are equipped to allow adolescents to recover without the risk of harming themselves or others.

ED staff require specific training to ensure that they are both competent and confident in managing these patients, and the safety of everyone involved is the highest priority. A team approach is vital with early recognition and verbal de-escalation a first priority before physical and or chemical restraint.[20] Actively listening to the adolescent, treating them with empathy and respect, acknowledging their distress about the current situation (e.g. being brought to ED against their will) and speaking and behaving in a calm, assertive but non-threatening way is essential for effective communication.[21]

Opportunistic health screening

Attendance at ED can highlight the presence of underlying health problems (e.g. anxiety, depression) or health-risk behaviours (e.g. alcohol or other drug use; unprotected sex). Attempting to address these issues is not straightforward and may be considered to be an inappropriate use of health professionals' time in ED. However, addressing these issues has the potential to prevent subsequent problems and re-attendance at ED and is recommended by the Royal Australasian College of Physicians.[22,23] In addition, many adolescents do not have a GP and utilise the ED as their primary source of health care – so the consultation should be seen not just as an opportunity for treatment but also for preventative medical care.[24]

Computer technology may help to overcome some of the barriers to psychosocial screening in the ED. Studies using web-based behavioural questionnaires, which the patient completes prior to medical assessment have shown to be cost effective, provide privacy and confidentiality, are time efficient and are highly acceptable (and even preferable to an interview) to adolescents and their families.[14]

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30.2

Eating disorders and anorexia

Robert Seith

ESSENTIALS

1 Patients can present as medically unstable with cardiovascular complications, electrolyte imbalances and dehydration requiring medical admission.

2 Patients can present as mentally unstable with suicidal ideation, eating disorder compensatory behaviours out of control, or comorbid psychiatric disorders requiring urgent mental health assessment.

3 Early intervention is the best way to assist with successful recovery, therefore it is important to recognise warning signs and refer to the appropriate service.

4 For patients with an eating disorder the following examinations should be carried out: weight, body mass index, postural heart rate and blood pressure, an electrocardiogram and initial blood testing.

5 Patients are at risk of refeeding syndrome and medically unstable patients should not commence feeding until blood results have been reviewed and a meal plan decided with the inpatient team.

Introduction

An eating disorder is a serious psychiatric illness, characterised by eating, exercise and body weight or shape becoming an unhealthy preoccupation of someone's life. Eating disorders result in impaired quality of life and severe psychological distress and have a high morbidity and in the case of anorexia

nervosa (AN) the highest mortality of any psychiatric disorder.[1] It is estimated that close to one million Australians have an eating disorder, and this number is increasing.[2]

The *Diagnostic and Statistical Manual of Mental Disorders* (DSM 5)[3] classifies eating disorders into a number of subtypes; these include AN, bulimia nervosa (BN), binge eating disorder (BED), night eating syndrome, purging disorder and avoidant-restrictive food intake disorder.

History

Consider eating disorders in any patients with significant weight loss (i.e. 10–15% in 3–6 months) even if they are in the normal weight range or overweight. Find out their current weight, pre-morbid weight, percentage weight loss and timing of weight loss. Remember it is abnormal for children to lose weight or fail to gain weight. If a parent is worried about their child's weight it is important to take their concerns very seriously.

Establish if the cause of the weight loss is consistent with an eating disorder by asking specific questions about their weight, body image, exercise and purging (Box 30.2.1).

If they have an established eating disorder then the key questions are aimed at identifying medical and mental stability.

It is important to assess suicide risk and ask about self-harm. A broader pyschosocial assessment may include a HEADSSS screen (Chapter 30.1).

Examination

Patients with suspected eating disorders need weight, height and body mass index (BMI) calculated and plotted on a growth chart appropriate for their sex and age. Postural (lying and standing) heart rate (HR) and blood pressure (BP) measurements are essential and a temperature should also be measured.

A general physical exam is needed to assess level of hydration, to look for other complications (see below), and signs of self-harm. Remember to consider other causes of weight loss while examining the patient.

Investigations

All patients should have an electrocardiogram (ECG). An ECG may show sinus

bradycardia, ST-segment elevation, T-wave flattening, low voltage and rightward QRS axis. A finding of QT-interval prolongation may indicate that the patient is at risk for cardiac arrhythmias and sudden death.

Box 30.2.1 Specific questions for assessing eating disorders

Weight profile

Percentage loss of weight over what time?
Highest weight and when?
Lowest weight and when?
Ideal weight and clothing size?

Restriction

Do you diet?
Do you skip meals?

Diet profile

Tell me what you eat for breakfast/lunch/dinner/snacks?

Associated behaviors with eating

Eating slowly, eating in isolation, rituals around mealtimes, bingeing

Dysmorphic concerns

What do you think about your weight now?

Concern over weight

Do you worry about your weight or body shape?
Do you worry about gaining weight?

Weight control

Do you do other things to control your weight, e.g. exercise, purging, laxatives?

If there are concerns about medical instability, suggested blood tests include full blood examination; urea, electrolytes and creatinine; calcium; magnesium; phosphate; glucose and liver function tests. If the reason for the weight loss is unclear consider adding on an erythrocyte sedimentation rate, thyroid function tests and coeliac screen. If there is sufficient blood to do a nutritional screen, and it has not been done in the last 3 months, iron studies, vitamin B12, red cell folate, vitamin D and zinc can be added on. Table 30.2.1 lists potential investigations and the rationale for requesting them.

Complications

Complications are the result of low weight, malnutrition, vomiting, laxative abuse and water loading:

- Cardiovascular effects: bradycardia, postural tachycardia, orthostatic hypotension, prolonged QT syndrome, potentially congestive cardiac failure, arrhythmias and sudden death

Table 30.2.1

Investigations for eating disorders

	Investigation	Purpose
Acute complications	ECG	Bradycardia; prolonged QTc
	UEC	Hypokalaemia if purging Hyponatraemia if water loading Impaired renal function if dehydrated Rarely may reveal other medical cause of weight loss, e.g. Addison's disease
	LFT	May have biliary sludging with increased bilirubin
	Ca, Mg, PO_4	Abnormalities with refeeding Low calcium if low albumin
	Blood glucose	Hypoglycaemia Excludes diabetes as a cause of weight loss
	FBC	Nutritional anaemia (micro- or macrocytic) Neutropenia Low platelets may occur with malnutrition
To detect other possible causes or comorbidity	ESR	Inflammatory bowel disease Rheumatological disease Chronic infection
	TFT	Abnormalities may reflect sick euthyroid or hyperthyroid as rare cause of weight loss
	Coeliac screen	May present with weight loss
Determine chronic complications of malnutrition	B12 Folate Iron studies Vitamin D FSH/LH/oestradiol	Low levels reflect malnourishment If amenorrhoea post puberty
	Bone density (not urgent)	Determine effect of malnourishment on growing bones or after period of amenorrhoea (PBS subsidy if >6 months) Bone age X-ray necessary in conjunction with bone density for <18 year old

Ca, calcium; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate; FBC, full blood count; FSH, follicle-stimulating hormone; LFT, liver function test; LH, luteinising hormone; Mg, magnesium; PBS, pharmaceutical benefits scheme; PO_4, phosphate; TFT, thyroid

function test; UEC, urea, electrolytes and creatinine.

- Electrolyte disturbances: hyponatraemia (water loading), hypernatraemia (dehydration), hypokalaemia, hypochloraemic alkalosis, hypophosphataemia, hypocalcaemia, hypomagnesaemia and refeeding syndrome
- Renal: renal failure
- Hypothermia
- Gastrointestinal (GI): constipation, abnormal liver function, upper GI inflammation ulceration and rupture, and pancreatitis
- Haematological: anaemia
- Gynaecology: amenorrhoea
- Mental health: suicidal ideation, depression, anxiety and psychosis.

Differential diagnosis

There are various medical causes of weight loss, including endocrine (diabetes, hyperthyroidism and adrenal insufficiency), gastrointestinal (coeliac disease, inflammatory bowel disease and peptic ulcer disease), malignancy and chronic infection, such as tuberculosis.[4] It is imperative that these are ruled out, although managing acute malnutrition should not be delayed while investigations take place.

AN and obsessive compulsive disorder (OCD) may coexist. When obsessions and compulsions primarily focus on eating and weight, the diagnosis is AN. Depression, anxiety and rumination disorder can cause significant weight loss but concerns about weight, shape and fear of weight gain will not be present. Those with rumination disorder will regurgitate food effortlessly after eating rather than it being a deliberate act.

Management

Early intervention is the best way to assist with successful recovery.[5] It is important to be aware of the warning signs that may indicate someone is developing or experiencing an eating disorder. Many people with an eating disorder do not realise they have a problem, or if they do, they may go to extraordinary lengths to hide their behaviour.

Goals of management in emergency department

1. Identify patients who are medically unstable and initiate management.
2. Identify patients who are psychologically unstable and are a risk to themselves or others and refer appropriately.
3. Recognise warning signs that may indicate someone is developing or experiencing an eating disorder and refer to the appropriate service.

Consider medical admission for patients with:

- significant electrolyte disturbance ($K^+ < 3.0$ mmol L^{-1})
- HR < 50 beats $minute^{-1}$
- postural HR increase ≥ 30 beats $minute^{-1}$
- systolic BP < 80 mmHg
- postural hypotension ≥ 20 mmHg
- hypothermia < 35.5°C
- dehydration
- arrhythmia or prolonged QTc (>0.45 s)
- weight <75% of their expected body weight or rapid weight loss
- eating disorder compensatory behaviours out of control, e.g. prolonged fasting, inability to eat at home, or uncontrolled purging and exercising
- social admission – supports failing in the community (only appropriate in rare circumstances).

Feeding

- Medical complications of eating disorders are due to a lack of food. Therefore the mainstay of treatment is ensuring adequate nutritional intake.
- Feeding should commence as soon as possible after blood tests have been taken and results confirmed to be normal. Ideally this is initiated in the ED after discussion with inpatient team.
- Options for feeding are oral intake of food via a meal plan, use of oral nutritional supplements, or nasogastric feeds. Patients should be offered and supported to have an appropriate meal. The nasogastric route is rarely required.
- Decisions about meals and meal plans should be made with the admitting inpatient team and/or dietician, taking into account the risk of refeeding syndrome.

Refeeding syndrome

Refeeding syndrome can be defined as the potentially fatal shifts in fluids and electrolytes that may occur in malnourished patients being refed (whether enterally or parenterally). These shifts result from hormonal and metabolic changes and may cause serious clinical complications.

The hallmark biochemical feature of refeeding syndrome is hypophosphataemia. However, the syndrome is complex and may also feature abnormal sodium and fluid balance; changes in glucose, protein, and fat metabolism; thiamine deficiency; hypokalaemia and hypomagnesaemia; along with neurologic, pulmonary, cardiac, neuromuscular and hematologic complications. Refeeding syndrome usually occurs within 4 days of starting to refeed.

Prognosis

Patients with anorexia have an 18-fold higher mortality[6,7] and around half have co-existing psychiatric conditions which may impact prognosis.[8,9] However, 50–75% receiving a family-based treatment approach reach a healthy weight. At follow-up, 60–90% will have recovered or partially recovered and relapse rates are generally low.[4] Emphasis on early, intensive, outpatient, family-based treatment approaches, with preservation of home and school life where possible, appears to improve outcome; thus demonstrating the importance of early diagnosis and referral.

BN is by nature episodic, acute or chronic. Around 40% recover after 6–12 months' treatment with high relapse rate at follow-up. Remission rates at 5 years are estimated at around 50%.[10] Rates of improvement and remission are better for BED, but the condition shows a similarly chronic course to BN. A small proportion of people with BN will develop AN, but commonly revert back to BN, whereas individuals with BED tend not to develop another eating disorder. Finally, both conditions are associated with obesity in later life[10] with treatment rarely having an effect on overall weight. Data are not yet available to determine whether early intervention significantly improves prognosis, although this seems likely since, as in AN, the duration of the untreated illness is an important predictor of outcome.

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