Paediatrics at a Glance

Lawrence Miall
Mary Rudolf
Malcolm Levene

Blackwell Science

Paediatrics at a Glance

This book is dedicated to our children

Charlie, Mollie, Rosie
Aaron, Rebecca
Alysa, Katie, Ilana, Hannah, David
and all those children who enlightened and enlivened us during our working lives.

Paediatrics at a Glance

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Preface

He knew the cause of every maladye,
Were it of hoot or cold or moiste or drye,
And where engendred and of what humour:
He was a verray parfit praktisour.

Geoffrey Chaucer c.1340–1400
A Doctor of Medicine, From Prologue to The Canterbury Tales

Chaucer outlined with some clarity the qualities that a doctor of medicine requires, and emphasized that knowledge about the causes of maladies was required to come to competent diagnosis. We have structured *Paediatrics at a Glance* around children's common symptoms and maladies, and the likely causes for them. We have also attempted to distil for the student not only the knowledge base they require but in addition the competencies they must acquire in order to become 'verray parfit praktisours' when working with children and their parents.

The world has changed since Chaucer's time, and it is now widely acknowledged that the medical curriculum suffers from 'information overload'. We have made great efforts to adhere to the General Medical Council's recommendations in *Tomorrow's Doctors*, and have only included the core knowledge that we consider is required by doctors in training. We have in addition placed great emphasis on the evaluation of the child as he or she presents.

The focus of the book is similar to its parent book *Paediatrics and Child Health.* In both we have attempted to provide a working approach to paediatric problems and child health as they present in primary, community and secondary care. We have now taken the familiar *At a Glance* format and have visually presented each common symptom and led the student through the causes and key components of the evaluation so that a competent diagnosis can be made. Chapters are also devoted to providing the reader with an understanding of children's development and their place in society with additional chapters on nutrition, childcare, education and community services.

Although this book is principally intended for medical students, it may well provide appropriate reading for nurses and other allied professionals who would like to deepen their understanding of children and paediatric management. It is particularly likely to appeal to those who take a visual approach to learning.

Hippocrates wrote in his *Aphorisms for Physicians*, 'Life is short, science is long, opportunity is elusive, experience is dangerous, judgement is difficult'. We have produced this concise volume in the hope that it will help students cope with these hurdles to medical training, and facilitate the development of clinical acumen in their work with children.

Lawrence Miall
Mary Rudolf
Malcolm Levene
July 2002

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List of abbreviations

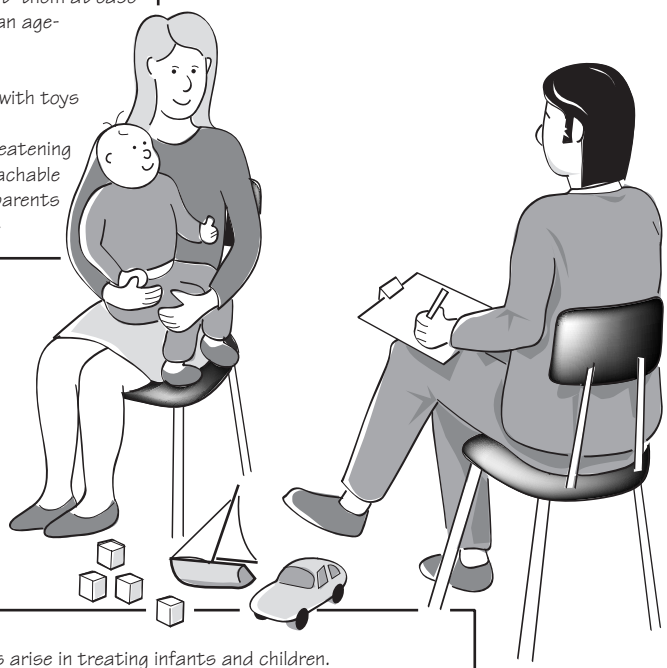
ACTH	adrenocorticotrophic hormone
ADD	attention deficit disorder
AIDS	acquired immunodeficiency syndrome
ALL	acute lymphoblastic leukaemia
ALTE	acute life-threatening event
AML	acute myeloid leukaemia
ANA	antinuclear antibody
APTT	activated partial thromboplatin time
ASD	atrial septal defect
ASO	antistreptolysin O titre
A-V	ateriovenous
AVPU	alert, verbal, painful, unresponsive
AVSD	atrioventricular septal defect
AXR	abdominal X-ray
AZT	zidovudine (azidothymidine)
BCG	bacille Calmette–Guérin
BP	blood pressure
BSER	brainstem evoked responses
CDH	congenital dislocation of the hip
CFTR	cystic fibrosis transmembrane regulator
CHD	congenital heart disease
CMV	cytomegalovirus
CNS	central nervous system
CPAP	continuous positive airway pressure
CPR	cardiopulmonary resuscitation
CRP	C reactive protein
CSF	cerebrospinal fluid
CT	computerized tomography
CXR	chest X-ray
DIC	disseminated intravascular coagulation
DKA	diabetic ketoacidocis
DMD	Duchenne muscular dystrophy
DMSA	dimercaptosuccinic acid
DTPA	diethylenetriamine penta-acetate
EB	Epstein–Barr
ECG	electrocardiogram
EEG	electroencephalogram
ENT	ear, nose and throat
ESR	erythrocyte sedimentation rate
FBC	full blood count
FDP	fibrin degradation product
FTT	failure to thrive
GCS	Glasgow coma scale
GOR	gastro-oesophageal reflux
GP	General Practitioner
G6PD	glucose-6-phosphate dehydrogenase
HbF	fetal haemoglobin
HbS	sickle-cell haemoglobin
HIV	human immunodeficiency virus
HSP	Henoch–Schönlein purpura
HUS	haemolytic uraemic syndrome
IBD	inflammatory bowel disease
ICP	intracranial pressure
IDDM	insulin-dependent diabetes mellitus
Ig	immunoglobulin
IM	intramuscular
INR	international normalized ratio
IRT	immunoreactive trypsin
ITP	idiopathic thrombocytopenic purpura
IUGR	intrauterine growth retardation
IV	intravenous
IVC	inferior vena cava
IVF	*in vitro* fertilization
IVH	intraventricular haemorrhage
IVU	intravenous urogram
JCA	juvenile chronic arthritis
JVP	jugular venous pulse
LMN	lower motor neurone
LP	lumbar puncture
MCH	mean cell haemoglobin
MCUG	micturating cystourethrogram
MCV	mean cell volume
MDI	metered dose inhaler
MLD	mild learning difficulty
MRI	magnetic resonance imaging
NEC	necrotizing enterocolitis
NHL	non-Hodgkin's lymphoma
NICU	Neonatal Intensive Care Unit
NPA	nasopharyngeal aspirate
NSAID	non-steroidal anti-inflammatory drug
OAE	otoautistic emissions
OFC	occipito frontal circumference
P_{CO_2}	partial pressure of carbon dioxide
PCP	*Pneumocystis carinii* pneumonia
PCR	polymerase chain reaction
PCV	packed cell volume
PDA	patent ductus arteriosus
PEFR	peak expiratory flow rate
PMH	past medical history
PT	prothrombin time
PTT	partial thromboplastin time
PUO	pyrexia of unknown origin
PVL	periventricular leucomalacia
RAST	radioallergosorbent test
RDS	respiratory distress syndrome
RNIB	Royal National Institute for the Blind
ROP	retinopathy of prematurity
RSV	respiratory syncitial virus
SCBU	Special Care Baby Unit
SGA	small for gestational age
SIADH	syndrome of inappropriate antidiuretic hormone secretion
SIDS	sudden infant death syndrome
SLD	severe learning difficulty
SSPE	subacute sclerosing encephalitis
STD	sexually transmitted disease

T_4	thyroxine
TB	tuberculosis
TGA	transposition of the great arteries
TSH	thyroid stimulating hormone
U&E	urea and electrolytes
UMN	upper motor neurone
URTI	upper respiratory tract infection
UTI	urinary tract infection
VACTERL	Vertebral anomalies, Anal atresia, Cardiac anomalies, Tracheo-oEsophageal fistula, Renal anomalies, Limb defects
VER	visual evoked response
VSD	ventricular septal defect
WCC	white cell count

1 The paediatric consultation

The doctor–patient relationship

The consultation
- Introduce yourself to the child and their parents. They may be anxious so try to put them at ease
- Use the child's name and talk in an age-appropriate manner
- Explain what is going to happen
- Use a child-friendly atmosphere, with toys available
- Arrange the seating in a non-threatening way that makes you seem approachable
- At the end, thank the child and parents and explain what will happen next



Observations
- While taking the history, try to observe the child and parents
- How do they relate to each other?
- Do the parents seem anxious or depressed?
- Will the child separate from the parent?
- Does the child play and interact normally?
- Is the child distractible or excessively hyperactive?

Ethical issues

A number of difficult ethical issues arise in treating infants and children. These include:
- Deciding whether to provide intensive care to infants born so premature that they are at the threshold of viability (i.e. <24 weeks gestation)
- Deciding whether to continue intensive therapy in an infant or child who has sustained an irreversible severe brain injury and who would be expected to have an extremely poor quality of life
- Deciding whether to use bone marrow cells from one sibling to treat another sibling
- Making a judgement as to when children are in such danger that they should be removed from the parents and taken into care for protection
- Deciding whether to give life-saving treatment, such as a heart transplant, against the apparent wishes of a young child who may not understand all the implications of refusing such treatment
- Respecting the confidentiality of a competent teenager who does not want her parents to know that she is being prescribed the oral contraceptive pill

Consent
- Children have rights as individuals
- Consent for the consultation and examination is usually obtained from the parents
- Older children who are competent may consent to examination and treatment without their parents, but cannot refuse treatment against their parents' wishes
- A child is defined in law as anyone under the age of 18 years

Paediatric medicine is unique in that the way in which we interact with our patients is very dependent on their age and level of understanding. When seeing a child over a period of time this interaction will evolve gradually from a relationship predominantly with the parents to one with the child as an individual making their own decisions.

Paediatrics covers all aspects of medicine relating to children. As the children grow, so the nature of their medical needs changes, until they match those of an adult. The younger the child the greater the difference in physiology and anatomy from an adult, and so the greater the range of health-related issues to be considered. Paediatrics is not just about diagnosing and treating childhood diseases, but also about maintaining normal health and development and preventing illness. This requires an understanding and appreciation of child health and normal development so that we can put the illness into context, and treat both the illness and the child.

The relationship in a paediatric consultation needs to be with both the child and the carers, usually the parents. Whilst obtaining information from the carer it is vitally important to establish and build a relationship with the child. This relationship changes rapidly with age—a newborn baby will be totally reliant on the parent to represent them, whilst a

young child will have their own views and opinions, which need to be recognized. The older child needs to start taking responsibility for their health, and should be fully involved in the consultation. This ability to interact with children as individuals, and with their parents and families at the same time, is one of the great skills and challenges of child health.

History taking

Taking a good history is a vital skill. The history can often lead to the diagnosis without needing to perform extensive examination or investigations. The history can be taken from a parent, a carer or from the child. Record who gave the history and in what context. A typical history should include:

- **Presenting complaint**—record the main problems in the family's own words as they describe them.
- **History of presenting complaint**—try to get an exact chronology from the time the child was last completely well.

Allow the family to describe events themselves; use questions to direct them and probe for specific information. Try to use open questions—'*tell me about the cough*' rather than '*is the cough worse in the mornings?*' Use direct questions to try to confirm or refute possible diagnoses.

- **Past medical history**—in young children and infants this should start from the pregnancy, and include details of the delivery and neonatal period, including any feeding or breathing problems. Ask about all illnesses and hospital attendances, including accidents.
- Ask about **immunizations** and foreign travel.
- **Developmental history**—ask about milestones and school performance. Are there any areas of concern?
- **Family and social history**—who is in the family and who lives at home? Ask about consanguinity as first-cousin marriages increase the risk of genetic disorders. Ask if there are any illnesses that run in the family. Does anyone have special needs and have there been any deaths in childhood?
- **Take a social history**—which school or nursery does the child attend? Ask about jobs, smoking, pets and try to get a feel for the financial situation at home. The social context of illness is very important in paediatrics.
- What **drugs** is the child taking and are there any **allergies**?
- Complete the **systems enquiry**—screening questions for symptoms within systems other than the presenting system.
- Ask if there is anything else that the family thinks should be discussed.
- At the end, try to come up with a **problem list**, which allows further management to be planned and targeted.

Approaching the examination

- Make friends with the child to gain their cooperation. Try to be confident yet non-threatening. It may be best to examine a non-threatening part of the body first before undressing the child, or to do a mock examination on their teddy bear.
- Try to get down to the child's level—kneel on the floor or sit on the bed. Look at the child as you examine them. Use a style and language that is appropriate to their age—'*I'm going to feel your tummy*' is good for a small child but not an adolescent!
- Explain what you are going to do, but be careful of saying '***can I** listen to your chest*' as they may refuse!
- Babies are best examined on a couch with the parent nearby; toddlers may need to be examined on the parent's lap. Older children and adolescents should always be examined with a chaperone—usually a parent but if the child prefers, a nurse. Allow as much privacy as possible when dressing and undressing the child.
- Sometimes you may need to be opportunistic and perform what examination you can, when you can. Always leave unpleasant things until the end—for example, looking in the throat and ears can often cause distress.
- In order to perform a proper examination the child will need to be undressed but this is often best done by the parent and only the region that is being examined needs to be undressed at any one time. Allow them to get dressed before moving on to the next region.
- Hygiene is important, both for the patient and to prevent the spread of infection to yourself and other patients. Always sterilize or dispose of equipment, such as tongue depressors or auroscope tips, that has been in contact with secretions.
- Much information can be gained by careful observation of the child—this can be done whilst talking to the parents or taking the history. Does the child look well, ill, or severely unwell? Is the child well nourished? Are behaviour and responsiveness normal—is the child bright and alert, irritable or lethargic? Is the child clean and well cared for?
- Is there any evidence of cyanosis or pallor? Does the child look shocked (mottled skin, cool peripheries) or dehydrated (sunken eyes, dry mouth)? Is there evidence of respiratory distress? What is the level of consciousness?
- Assess the child's growth—height and weight should be plotted on centile charts. Head circumference should be measured in infants and in those where there is neuro-developmental concern.

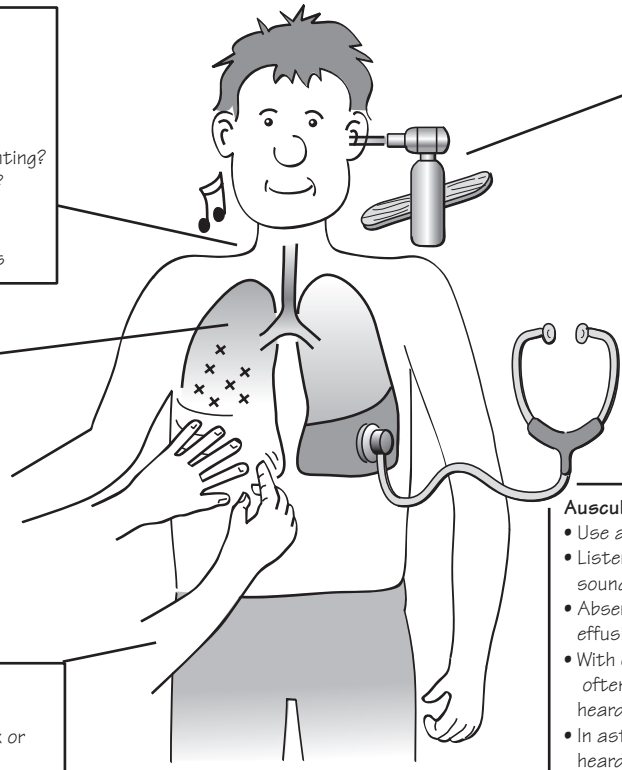
The examination of individual systems is discussed in detail on the following pages.

KEY POINTS

- The consultation is with the child and the carers and both must be involved.
- History taking is a crucial skill.
- Language and approach need to be adapted to the age of the child and the understanding of the family.
- Consent should be obtained for examination, which must be conducted in a child-friendly manner.
- Observation is often more important than hands-on examination when assessing a child.

2 Systems examination

Respiratory system



Observation
- Is there respiratory distress?
  - -nasal flaring, recession
  - -use of accessory muscles
- Count the respiratory rate
- Is there wheeze, stridor or grunting?
- Is the child restless or drowsy?
- Is there cyanosis or pallor?
- Is there finger clubbing?
  - - cystic fibrosis, bronchiectasis

Chest wall palpation
- Assess expansion
- Check trachea is central
- Feel apex beat
- Is there chest deformity?
  - -Harrison's sulcus: asthma
  - -barrel chest: air-trapping
  - -pectus excavatum: normal
  - -pigeon chest: congenital heart disease
- May 'feel' crackles

Percussion
- Resonant: normal
- Hyper-resonant: pneumothorax or air-trapping
- Dull: consolidation (or normal liver in right lower zone)
- Stony dull: pleural effusions

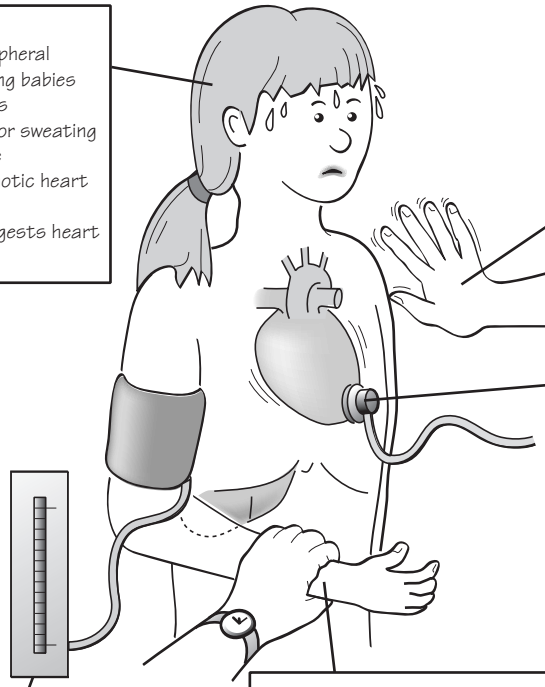
Ear, nose and throat
- Examine eardrums using an auroscope
  - -grey and shiny: normal
  - -red and bulging: suggests otitis media
  - -dull and retracted: chronic secretory otitis media (glue-ear)
- Examine nostrils for inflammation, obstruction and polyps
- Examine pharynx using tongue depressor (leave this until last!)
  - -Are the tonsils acutely inflamed (red +/- pustules or ulcers) or chronically hypertrophied (enlarged but not red)
- Feel for cervical lymphadenopathy

Auscultation
- Use an appropriately sized stethoscope!
- Listen in all areas for air entry, breath sounds and added sounds
- Absent breath sounds in one area suggests pleural effusion, pneumothorax or dense consolidation
- With consolidation (e.g. pneumonia) there is often bronchial breathing with crackles heard just above the consolidated lung
- In asthma and bronchiolitis expiratory wheeze is heard throughout the lung fields
- In young children upper airway sounds are often transmitted over the whole chest. Asking the child to cough may clear them

KEY QUESTIONS FROM THE HISTORY

- Is there a history of cough? Nocturnal cough may suggest asthma.
- Is the child short of breath or wheezy?
- Are the symptoms related to exercise, cold air or any other triggers?
- Has there been a fever, which would suggest infection?
- Has the child coughed up (or vomited) any sputum?
- Is there a family history of respiratory problems (e.g. asthma, cystic fibrosis)?
- Has the child travelled abroad or been in contact with relatives who might have TB?
- Is there any possibility the child may have inhaled a foreign body?
- How limiting is the respiratory problem—how far can the child run, how much school has been missed because of the illness?
- Has the child been pulling at his ears (suggesting an ear infection) or showing difficulty swallowing (tonsillitis or epiglottitis)?

Cardiovascular system



Observation
- Is there central cyanosis? Peripheral cyanosis can be normal in young babies and those with cold peripheries
- If the child is breathless, pale or sweating this may indicate heart failure
- Is there finger clubbing? - cyanotic heart disease
- Is there failure to thrive? -suggests heart failure

Palpation
- Feel apex beat (position and character), reflects left ventricular function
- Feel for right ventricular heave over sternum
- Feel for thrills (palpable murmurs)
- Hepatomegaly suggests heart failure. Peripheral oedema and raised JVP are rarely seen in children

Auscultation
- On the basis of the child's age, pulse, colour and signs of failure try to think what heart lesion may be likely, then confirm this by auscultation
- Listen for murmurs over the valve areas and the back (see p. 94). Diastolic murmurs are always pathological
- Listen to the heart sounds: are they normal, increased (pulmonary hypertension), fixed and split (ASD) or are there added sounds (gallop rhythm in heart failure or ejection click in aortic stenosis)?

Systolic murmur

Circulation
- Measure blood pressure with age-appropriate cuff, which should cover 2/3 of the upper arm
- Check capillary refill time; if more than 2 s, consider shock

Pulse
- Rate: fast, slow or normal?

Age (years)	Normal pulse (beats/min)
<1	110–160
2-5	95–140
5-12	80–120
>12	60–100

- Rhythm: regular or irregular? Occasional ventricular ectopic beats are normal in children
- Volume: full or thready (shock)
- Character: collapsing pulse is most commonly due to patent arterial duct. Slow rising pulse suggests left ventricular outflow tract obstruction
- Always check femoral pulses in infants—coarctation of the aorta leads to reduced or delayed femoral pulses

KEY QUESTIONS FROM THE HISTORY
- Has the child ever been cyanosed?
- Has the child been breathless or tired (may suggest cardiac failure)?
- Has the child been pale and sweaty (may suggest cardiac failure)?
- Ask about the pattern of feeding in babies, as breathlessness may inhibit feeding.
- Review the child's growth—is there evidence of failure to thrive?
- Has there been any unexplained collapse, such as fainting?
- Has the child ever complained of palpitations or of their heart racing?
- Has anyone ever noticed a heart murmur in the past? (Physiological flow murmurs may only be present at times of illness or after exercise.)
- Is there a family history of congenital heart disease?
- If the child has a heart defect, have they been taking prophylactic antibiotics for dental or other invasive treatment? (Consider particularly for valve disorders and ventricular septal defects.)

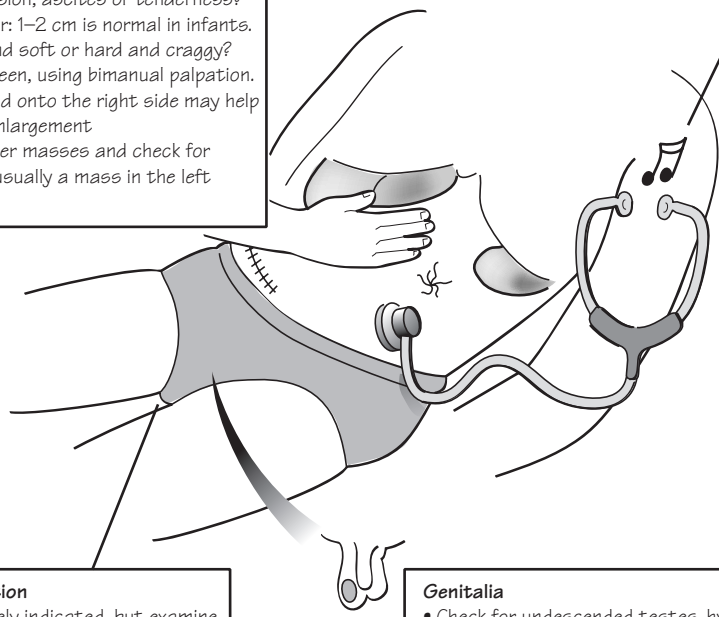
Abdominal system and nutritional status

(See also Chapters 3 and 22)

Palpation
- Use warm hands and ask whether the abdomen is tender before you begin
- Is there distension, ascites or tenderness?
- Palpate the liver: 1–2 cm is normal in infants. Is it smooth and soft or hard and craggy?
- Feel for the spleen, using bimanual palpation. Turning the child onto the right side may help
- Feel for renal enlargement
- Palpate for other masses and check for constipation (usually a mass in the left iliac fossa)

Percussion
- Percuss for ascites (shifting dullness) and to check for gaseous distension

Auscultation
- Listen for normal bowel sounds. 'Tinkling' suggests obstruction



Observation
- Make sure the child is relaxed—small children can be examined on a parent's lap; older children should lie on a couch
- **Jaundice:** look at the sclerae and observe the urine and stool colour (dark urine and pale stools suggests obstructive jaundice)
- Check conjunctivae for anaemia
- **Oedema:** check over tibia and sacrum. Periorbital oedema may be the first thing noticed by parents
- **Skin:** look for spider naevi—suggests liver disease
- **Wasted buttocks:** suggests weight loss and is characteristic of coeliac disease
- Measure the mid upper arm circumference

Rectal examination
- This is very rarely indicated, but examine the anus for fissures or trauma

Genitalia
- Check for undescended testes, hydroceles and hernias. Retractile testes are normal
- In girls examine the external genitalia if there are urinary symptoms

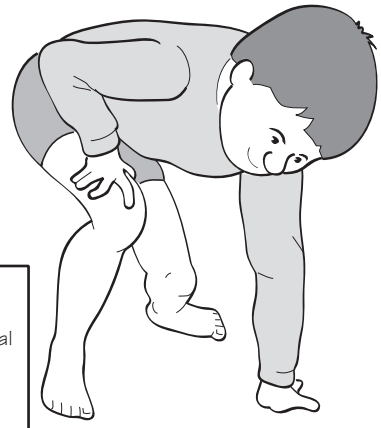
KEY QUESTIONS FROM THE HISTORY

- Review the child's diet. Ask in detail what the child eats. '*Take me through everything you ate yesterday*.'
- Is the quantity of calories sufficient and is the diet well balanced and appropriate for the child's age?
- Ask about the pattern of weight gain. The parent-held record (red book) can provide invaluable information about previous height and weight measurements.
- Does the child have a good appetite?
- Has there been any vomiting?
- In babies ask about posseting (small vomits of milk) and regurgitation of milk into the mouth, which may suggest gastro-oesophageal reflux.
- Has there been any diarrhoea? Always assess what the parents mean by diarrhoea—frequent or loose stools or both?
- Has the child been constipated? Straining, pain on defaecation, poor appetite and a bloated feeling may suggest this is a problem.
- Have there been any urinary symptoms such as frequency, dysuria or enuresis?
- Has the child got any abdominal pain? Ask about the site and nature of the pain.
- Is there a relevant family history (e.g. coeliac disease, inflammatory bowel disease, constipation)?

Neurological assessment

Observation
- Abnormal movements: choreoathetoid 'writhing' movements, jerks in myoclonic epilepsy and infantile spasms
- Gait—this can provide important clues:
  -stiffness: suggests UMN lesion
  -waddling: spastic diplegia, Duchenne muscular dystrophy(DMD) or congenital dislocation of hips
  -weakness on standing, e.g. Gower sign in DMD
  -broad based gait: ataxia
- Muscle bulk/wasting
- Posture: look for evidence of contractures

Cranial nerves
- Examine as in adults



Coordination
- Finger–nose test and heel–shin test, and observe gait. Very important if considering CNS tumours as cerebellar signs are common

Tone
- Hypotonia suggests LMN lesion
- Spasticity suggests UMN lesion and is seen in cerebral palsy, especially in thigh abductors and calf muscles (may cause toe walking)

Power
- Describe in upper and lower limbs, against resistance

Reflexes
- Assess at knee, ankle, biceps, triceps and supinator tendons
- Clonus may be seen in UMN lesions
- Plantar reflex is upwards until 8 months of age, then downwards

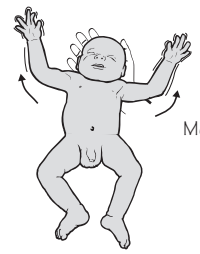
Neurological examination in infants

Young children cannot cooperate with a formal neurological examination so observation becomes more important: watch what the child is doing while you play with them
- How does the infant move spontaneously? Reduced movement suggests muscle weakness
- What position are they lying in? A severely hypotonic baby adopts a 'frog's leg' position (see below)
- Palpate anterior fontanelle to assess intracranial pressure
- Assess tone by posture and handling: a very floppy hypotonic baby tends to slip through your hands like a rag doll. Put your hand under the abdomen and lift the baby up in the ventral position: a hypotonic infant will droop over your hand. Pull the baby to sit by holding the baby's arms: observe the degree of head lag. Hypertonia is suggested by resistance to passive extension of the limbs and by scissoring (crossing-over) of the lower limbs when the infant is lifted up (see below)
- Primitive reflexes are present at birth. Persistence beyond normal period suggests a UMN lesion

Moro reflex	Symmetrical abduction and then adduction of the arms when the baby's head is dropped back quickly into your hand (see below). Disappears by 4 months
Palmar grasp	Touching the palm causes the baby to grip an object. Disappears by 2 months
Asymmetric tonic neck reflex	The arm is extended on the side the baby is facing while the opposite arm is flexed(see below). Disappears by 6 months

Scissoring of the lower limbs

'Frog's leg' position



Moro reflex

Asymmetric tonic neck reflex

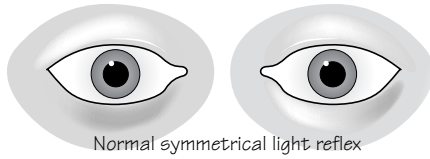
KEY QUESTIONS FROM THE HISTORY
- Has there been any developmental concerns—quickly review major milestones?
- Has there been any concern about hearing or vision?
- Did the child pass the hearing screening check (currently at 7 months)?
- Has the child ever had a convulsion or unexplained collapse?
- Is there a relevant family history (ask specifically about blindness, deafness, learning difficulties and genetic disorders such as muscular dystrophy)?
- Has there been any change in school performance or personality?
- Has the child been clumsy or had a change in gait?
- Has there been any headache or vomiting (may suggest raised intracranial pressure)?
- Ask about function—how is the child limited by their condition, if at all?
- Briefly review the social situation—does the family receive any relevant benefits, e.g. disability living allowance? Are there mobility problems?

The visual system

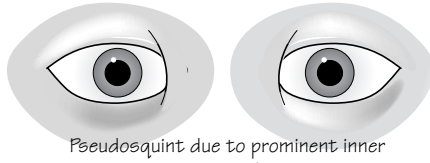
Observation of eyes
- Look at the iris, sclera and pupil
- Check pupils are equal and react to light, both directly and indirectly
- Look for red reflex to exclude cataract, especially in the newborn
- Look at reflection of light on the cornea—is it symmetrical or is one eye squinting? (see box opposite)
- Look at the inner epicanthic folds—if very prominent they may cause a pseudosquint

Visual acuity
- Does the child fix and follow an object through 180 degrees?
- Can they see small objects (e.g. hundreds and thousands, small rolling Stycar balls)
- Older children can perform a modified Snellen chart with objects

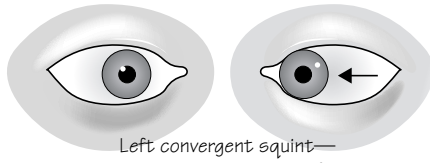
Ocular movements and visual fields
- Test full range of movements, looking for paralytic muscle or nerve lesions
- Look for and describe any nystagmus
- Check visual fields by using a 'wiggling' finger



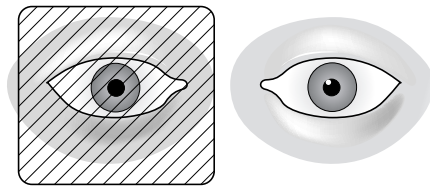
Normal symmetrical light reflex



Pseudosquint due to prominent inner epicanthic folds



Left convergent squint—note assymetric light reflex



When the good eye is covered the squinting eye straightens (fixates)

Assessment of a squint
- Any squint in an infant beyond the age of 6 weeks needs referral to an ophthalmologist. A squinting eye that is left untreated may cause amblyopia (cortical blindness) on that side
- Some 'latent' squints are present only when the eye is tired; the history is important
- Check the corneal light reflex at different angles of gaze
- Check ocular movements—is there a fixed angle between the eyes or a paralytic squint, where the squint increases with eye movement?
- Check visual acuity
- Perform fundoscopy and red reflex
- Perform the **cover test** by asking the child to fix on an object. Cover the 'good' eye and watch the squinting eye flick to fix on the object. Latent squints may also become apparent when that eye is covered

Fundoscopy
- An essential but difficult skill—practise on every child you see!
- Look at the optic disc and retina, the red reflex and the lens

KEY QUESTIONS FROM THE HISTORY

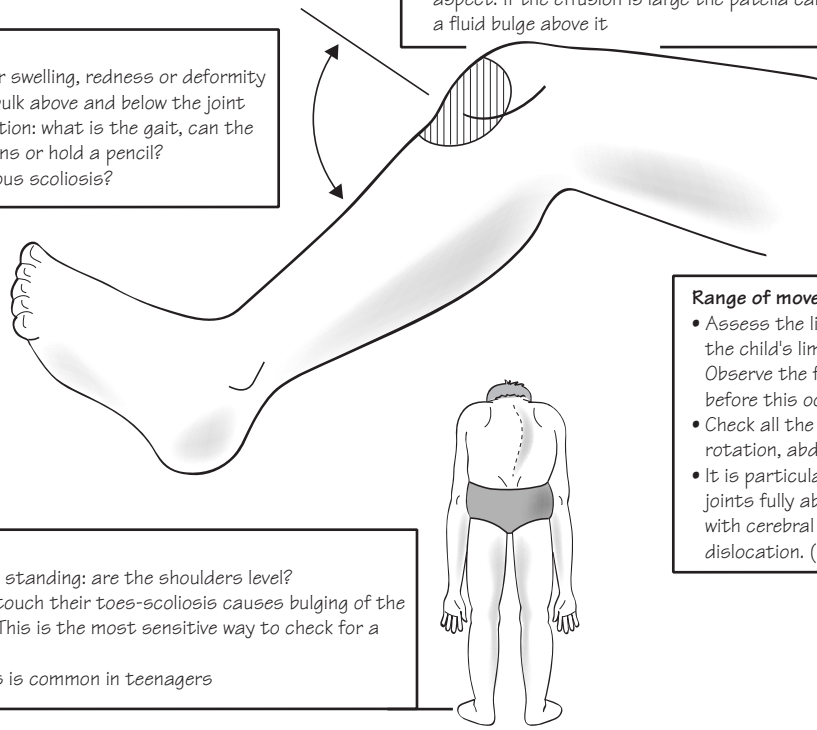
- Have the parents been concerned about the child's vision?
- Has anyone ever noticed a squint?
- Is the child able to see clearly (for example, the board at school)?
- Is there any relevant family history (e.g. retinitis pigmentosa, congenital cataracts)?
- Has the child been complaining of headaches, which may suggest poor visual acuity?
- Has the child seen an optician recently?
- Are there any risk factors for visual problems, such as extreme prematurity, diabetes mellitus or other neurological concerns?

Musculoskeletal system

Individual joint problems are discussed in Chapters 32 and 55.

Palpation
- Feel the temperature of the skin over the joint, and feel for joint tenderness
- Feel for an effusion: in the knee milk fluid down and feel a bulge in the medial aspect. If the effusion is large the patella can be rocked in and out, causing a fluid bulge above it

Observation
- Observe joints for swelling, redness or deformity
- Observe muscle bulk above and below the joint
- Observe the function: what is the gait, can the child do up buttons or hold a pencil?
- Is there any obvious scoliosis?



Range of movements
- Assess the limit of active movements, then move the child's limb to assess passive movements. Observe the face for signs of pain, and stop before this occurs
- Check all the large joints in flexion, extension, rotation, abduction and adduction
- It is particularly important to check that the hip joints fully abduct in newborns and in children with cerebral palsy in order to exclude hip dislocation. (see Chapter 48)

Scoliosis
- Observe the child standing: are the shoulders level?
- Ask the child to touch their toes-scoliosis causes bulging of the ribs on one side. This is the most sensitive way to check for a scoliosis
- Postural scoliosis is common in teenagers

KEY QUESTIONS FROM THE HISTORY

- Has the child had any joint pain or swelling?
- Is the child able to walk and exercise normally?
- Have the parents noticed any change in gait or clumsiness?
- Has there been any unexplained fever (may suggest autoimmune disorders or septic arthritis)?
- What is the level of function like—can the child manage fiddly tasks such as doing up buttons?
- Have the parents noticed any rashes (may suggest rheumatoid disease (see p. 117) or Henoch–Schönlein purpura (see p. 85))?

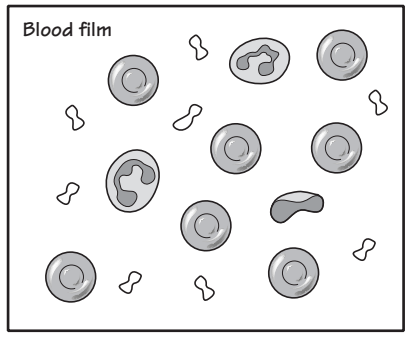
3 Understanding investigations I

Investigations should only be requested to confirm a clinical diagnosis, or if indicated after taking a thorough history and examination. Sometimes investigations are performed to rule out more serious but less likely conditions. Blindly performing investigations as a 'fishing' exercise in the hope of throwing up an abnormality is usually counterproductive, often leading to increased anxiety and further investigations when unexpected results are obtained. These pages describe how to interpret some of the common investigations performed in paediatrics.

Haematology

Haemoglobin
- High at birth (18 g/dl), falling to lowest point at 2 months (range 9.5–14.5 g/dl). Stabilizes by 6 months
- Low haemoglobin indicates anaemia. Further investigation will pinpoint the cause (see below)

Blood film



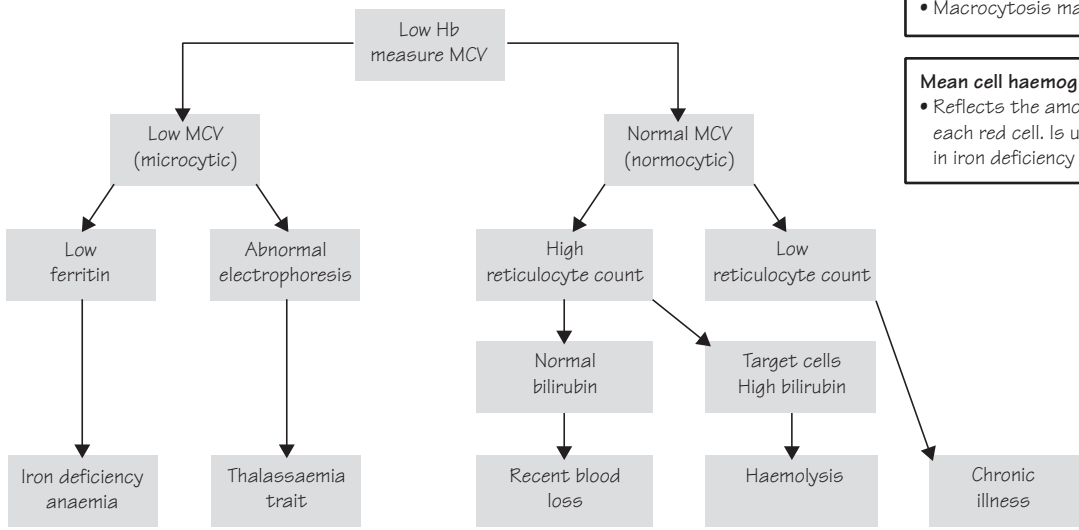
Normal values

Haemoglobin	11 – 14 g/dl
Haematocrit	30 – 45%
White cell count	6 – 15 x 10^9/l
Reticulocytes	0 – 2%
Platelets	150 – 450 x 10^9/l
MCV	76 – 88 fl
MCH	24 – 30 pg
ESR	10 – 20 mm/h

Mean cell volume (MCV)
- Measures the size of RBCs
- Microcytic anaemia (MCV <76 fl) is usually due to iron deficiency, thalassaemia trait or lead poisoning
- Macrocytosis may reflect folate deficiency

Mean cell haemoglobin (MCH)
- Reflects the amount of haemoglobin in each red cell. Is usually low (hypochromic) in iron deficiency

Flow diagram to show the investigation of anaemia



Platelets
- High platelet count usually reflects bleeding or inflammation (e.g. Kawasaki's disease)
- Low platelet count is commonly seen with idiopathic thrombocytopenic purpura (ITP) when there is a risk of spontaneous bruising and bleeding. In the newborn it may be low due to maternal IgG-mediated immune thrombocytopenia

Clotting
- Prothrombin time (PT) compared with a control is used to calculate the INR: normal is 1.0. Principally assesses extrinsic pathway. Prolonged in Vitamin K deficiency, liver disease and disseminated intravascular coagulation (DIC)
- Activated partial thromboplastin time (APTT) reflects the intrinsic pathway. Prolonged in heparin excess, DIC and haemophilia A
- Fibrin degredation products (FDPs) are increased in DIC
- Bleeding time: literally the time a wound bleeds for. Prolonged in von-Willebrand's disease and thrombocytopenia

White blood cells
- Leucocytosis usually reflects infection—neutrophilia and 'left shift' (i.e. immature neutrophils) implies bacterial infection. Lymphocytosis is commoner in viral infections, atypical bacterial infection and whooping cough
- Neutropenia (neutrophils < 1.0 x 10^9/l) can occur in severe infection or due to immunosuppression. There is a high risk of spontaneous infection
- Leukaemia: There is usually a very high (or occasionally low) WCC with blast cells seen. Bone marrow aspirate is required (see Chapter 56)

Interpretation of blood gases

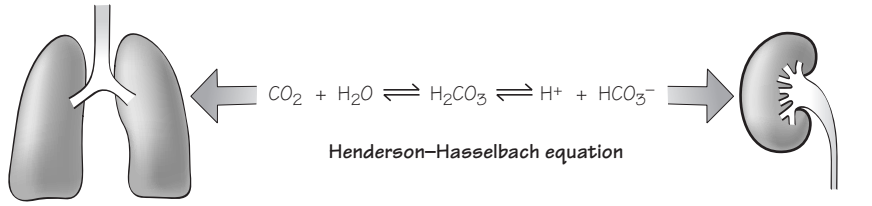
The acidity of the blood is measured by pH. A high pH refers to an alkalosis and a low pH to an acidosis. Once the blood becomes profoundly acidotic (pH<7.0), normal cellular function becomes impossible. There are metabolic and respiratory causes of both acidosis and alkalosis (see below). The pattern of blood gas abnormality (particularly the pH and PCO_2) can be used to determine the type of abnormality.

Metabolic acidosis
- Severe gastroenteritis
- Neonatal asphyxia (build-up of lactic acid)
- Shock
- Diabetic ketoacidosis
- Inborn errors of metabolism
- Loss of bicarbonate (renal tubular acidosis)

Respiratory acidosis
- Respiratory failure and underventilation

Metabolic alkalosis
- Usually due to vomiting, e.g. pyloric stenosis

Respiratory alkalosis
- Hyperventilation (e.g. anxiety)
- Salicylate poisoning: causes initial hyperventilation and then metabolic acidosis due to acid load



Compensation can occur by the kidneys, which can vary the amount of bicarbonate excreted. A persistent respiratory acidosis due to chronic lung disease will lead to retention of bicarbonate ions to buffer the acid produced by CO_2 retention. Hence, a **compensated respiratory acidosis** will have a low–normal pH, a high PCO_2 and a very high bicarbonate level

Normal arterial blood gas values

pH	7.35 – 7.42
PCO_2	4.0 – 5.5 kPa
PO_2	11 – 14 kPa (children)
	8 – 10 (neonatal period)
HCO_3^-	17 – 27 mmol/l

Determining the type of blood gas abnormality

	pH	PCO_2	PO_2	HCO_3^-
Metabolic acidosis	Low	N/low*	N	Low
Respiratory acidosis	Low	High	N/low	N/high*
Metabolic alkalosis	High	N/high*	N	High
Respiratory alkalosis	High	Low	N/high	N/low*

*Refers to the compensated state

Electrolytes and clinical chemistry

The normal values are shown below. Alterations in sodium usually reflect alterations in the level of hydration and total body water content. A sudden fall in sodium can cause fitting. High potassium levels can cause serious cardiac arrhythmias and need to be controlled rapidly. High potassium levels are commonly seen in acute renal failure, or may be artefactual due to haemolysis if venepuncture was difficult. Therapies to reduce a high potassium level include salbutamol, insulin and dextrose (to drive potassium into the cells) and calcium resonium.

Characteristic patterns of serum electrolyte abnormality sometimes suggest particular diagnoses:
- **Pyloric stenosis:**
  There is often a metabolic alkalosis, a low chloride and potassium concentration (due to repeated vomiting and loss of stomach acid) and a low sodium concentration
- **Diabetic ketoacidosis:**
  There is a metabolic acidosis with a very low bicarbonate, a high potassium, high urea and creatinine and a very high glucose concentration
- **Gastroenteritis:**
  Urea concentration is high, but the sodium may be either high or low depending on the sodium content of the diarrhoea, and on the type of rehydration fluid that has been administered

Normal ranges

Sodium	135 – 145 mmol/l
Potassium	3.5 – 5.0 mmol/l
Chloride	96 – 110 mmol/l
Bicarbonate	17 – 27 mmol/l
Creatinine	20 – 80 µmol/l
Urea	2.5 – 6.5 mmol/l
Glucose	3.0 – 6.0 mmol/l
Alkaline phosphatase	150 – 1000 (infants)
	250 – 800 (child)

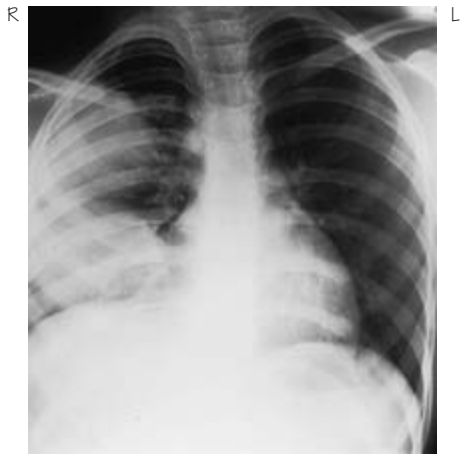
Causes of abnormal sodium balance

Hypernatraemia (Na^+ >145 mmol/l)
- Dehydration
  - fluid deprivation or diarrhoea
- Excessive sodium intake
  - inappropriate formula feed preparation
  - deliberate salt poisoning (very rare)

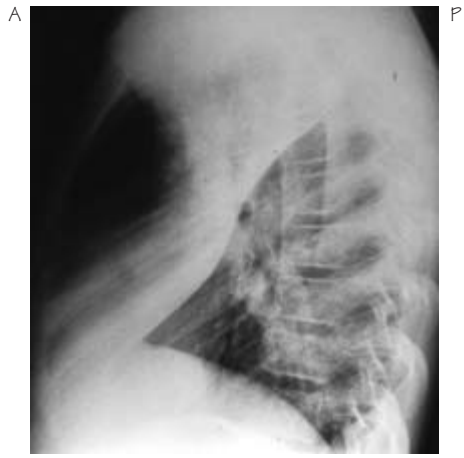
Hyponatraemia (Na <135 mmol/l)
- Sodium loss
  - diarrhoea (especially if replacement fluids hypotonic)
  - renal loss (renal failure)
  - cystic fibrosis (loss in sweat)
- Water excess
  - excessive intravenous fluid administration
  - SIADH (inappropriate antidiuretic hormone secretion)

4 Understanding investigations II

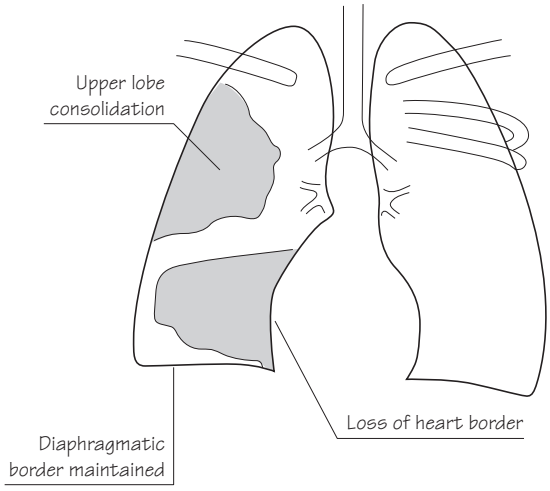
Features to look for on a chest radiograph

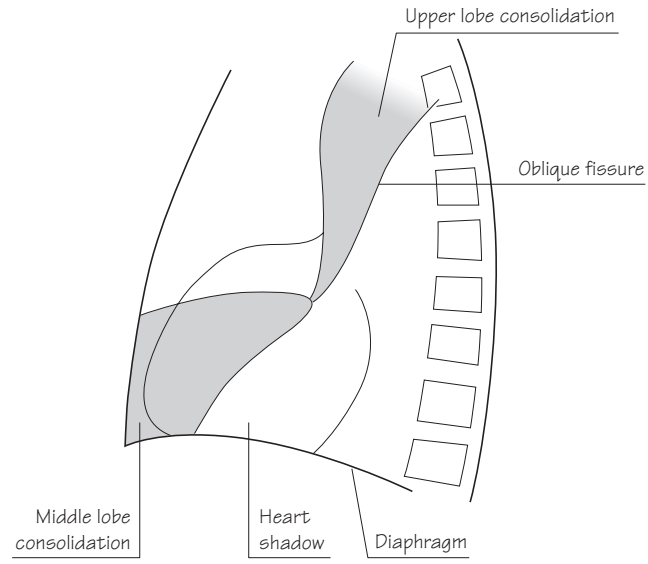


CXR of right middle and upper lobe pneumonia



Lateral X-ray showing right upper and middle lobe pneumonia





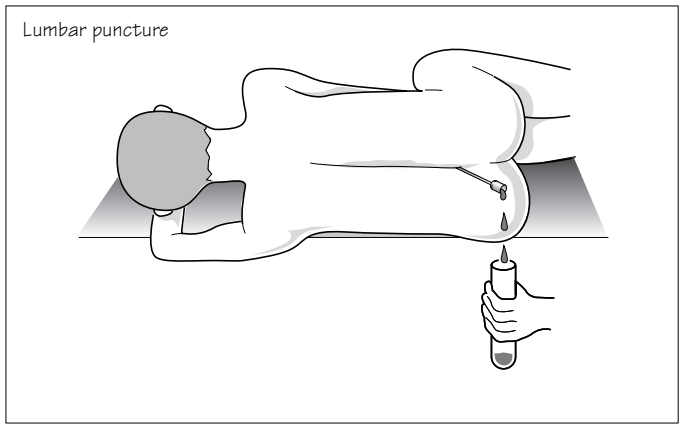
As respiratory disorders are so common in paediatric practice, it is very important to be able to accurately interpret chest radiographs. If there is uncertainty the film should be discussed with an experienced radiologist.

- Identify the patient name, date and orientation (left and right).
- Check the penetration—the vertebrae should just be visible behind the heart shadow.
- Check that the alignment is central by looking at the head of the clavicles and the shape of the ribs on each side.
- Comment on any foreign objects such as central lines.
- Examine the bony structures, looking for fractures, asymmetry and abnormalities (e.g. hemivertebrae). Rib fractures are best seen by placing the X-ray on its side.
- Check both diaphragms and costo-phrenic angles are clear. The right diaphragm is higher than the left because of the liver. Check there is no air beneath the diaphragm (indicates intestinal perforation).
- Look at the cardiac outline. At its widest it should be less than half the width of the ribcage (cardiothoracic ratio <0.5).
- Look at the mediastinum—note that in infants the thymus gland can give a 'sail'-like shadow just above the heart.

• Check lung expansion—if there is air trapping the lung fields will cover more than nine ribs posteriorly, and the heart will look long and thin.
• Examine the lung fields looking for signs of consolidation, vascular markings, abnormal masses or foreign bodies. Check that the lung markings extend right to the edge of the lung—if not, consider a pneumothorax (dark) or a pleural effusion (opaque). Consolidation may be patchy or dense lobar consolidation. A lateral X-ray may be required to determine exactly which lobe is affected. A rule of thumb is that consolidation in the right middle lobe causes loss of the right heart border shadow and right lower lobe consolidation causes loss of the right diaphragmatic shadow. Always look at the area 'behind' the heart shadow for infection in the lingula. If the mediastinum is pulled towards an area of opacity consider collapse rather than consolidation as the pathology.

Lumbar puncture and CSF analysis

Lumbar puncture is usually performed to diagnose or exclude meningitis. It should not be performed if there is evidence of raised intracranial pressure, if the child is haemodynamically unstable (e.g. septic shock) or if there is a low platelet count or coagulopathy. A fine spinal needle with a stylet is passed between the vertebral spines into the cerebrospinal fluid (CSF) space. A few drops of CSF are then collected for microscopy, for culture and for analysis of protein and glucose concentrations. Samples can also be sent for polymerase chain reaction (PCR) analysis to look for evidence of meningococcal or herpes infection if meningitis or encephalitis is suspected. Normal CSF is usually 'crystal clear'. If it is cloudy, this suggests infection. Fresh blood which clears usually indicates a traumatic tap, but a massive intracranial haemorrhage must be considered if the CSF remains blood-stained. Old blood from a previous haemorrhage gives a yellow 'xanthochromic' appearance. A manometer can be used to measure the CSF pressure, though this is not routinely performed.



Analysis of CSF.

	Normal	Bacterial meningitis	Viral meningitis
Appearance	Crystal clear	Turbid, organisms seen	Clear
White cells	<5/mm^3	↑↑↑ (polymorphs)	↑ (lymphocytes)
Protein	0.15–0.4 g/l	↑↑	Normal
Glucose	>50% blood	↓	Normal

Urinalysis

Fresh urine should be collected into a sterile container from a midstream sample if possible. Urine bags placed over the genitalia may be used in infants, but beware of contamination.
• Observe the urine—is it cloudy (suggests infection) or clear?
• What is the colour—pink or red suggests haematuria from the lower urinary tract? Brown 'cola' coloured urine suggests renal haematuria.
• Smell the urine for ketones and for the fishy smell of infection. Unusual smelling urine may suggest an inborn error of metabolism.
• Dipstick test the urine using commercial dipsticks. This may reveal protein (suggests infection, renal damage or nephrotic syndrome), glucose (present in diabetes), ketones (in diabetic ketoacidocis, DKA) or nitrites (suggestive of infection). These sticks are very sensitive to the presence of blood, and may detect haematuria even if the urine looks clear.
• Finally, examine the urine under the microscope for white cells, red cells, casts and the presence of organisms. If suspicious of infection send a sample for culture. A pure growth of >10^5 colony-forming units of a single organism and >50 white cells/mm^3 confirms infection (see p. 69).

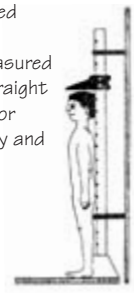
KEY POINTS
• Before ordering an investigation consider how the result might alter the management.
• Try to focus investigations on the differential diagnosis based on clinical assessment.
• Sometimes investigations can be used to quickly rule out important or serious diagnoses.
• If a test is performed you must chase up the result.

5 Growth and puberty

Growth

Accurate measurement of growth is a vital part of the assessment of children. In order to interpret a child's growth, measurements must be plotted on a growth chart. If there is concern about growth, the rate of growth must be assessed by measuring the child on two occasions at least 4–6 months apart.

Height
- Use a properly calibrated standing frame
- The child should be measured barefoot with knees straight and feet flat on the floor
- Stretch the child gently and read the measurement

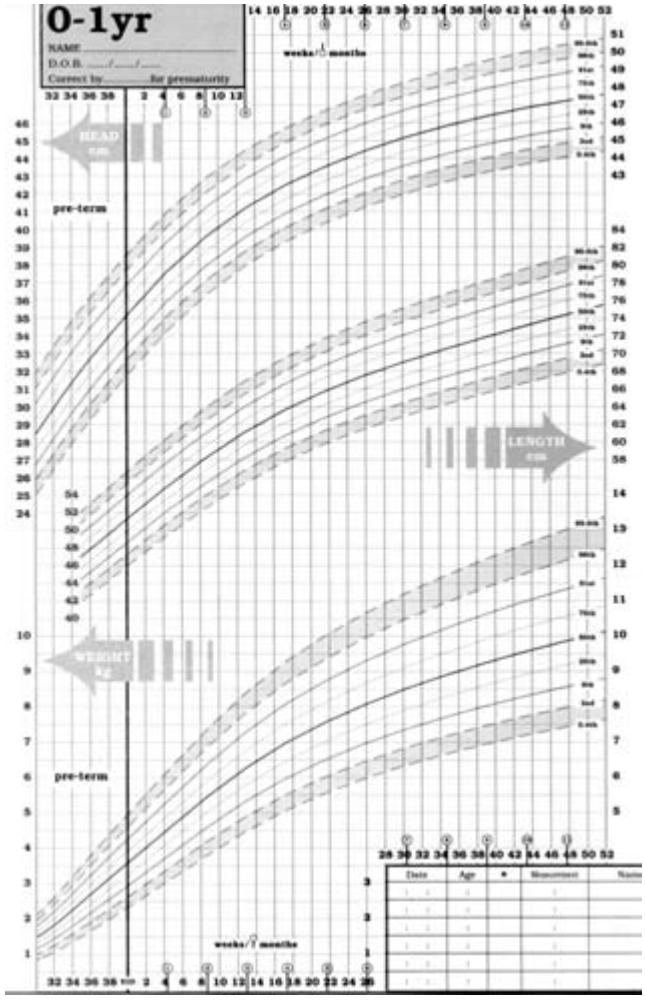


Length
- The child should be measured lying down until 2 years of age
- Measuring the length of infants requires skill
- Use proper equipment and two people to hold the child

Weight
- Scales must be calibrated accurately
- Babies should be weighed naked (no nappy!)
- Older children should be weighed in underwear only

Head circumference
- Use flexible non-stretchable tape
- Obtain three successive measurements and take the largest to be the occipito frontal circumference (OFC)

GROWTH CHART



The y-axis uses kg for weight and cm for OFC and height/length

The x-axis may be divided in to months or decimal age

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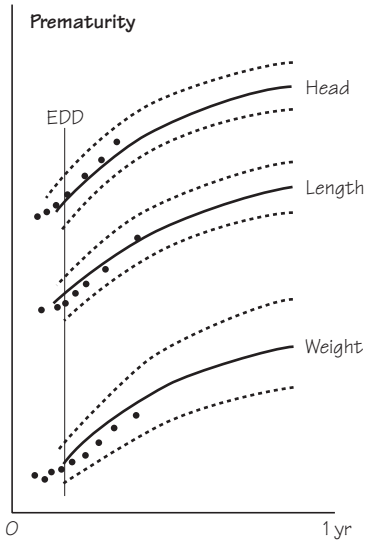
Plot on a growth chart.
In the UK the 1990 UK Growth References are used:
- Nine equidistant centile lines are marked
- For weight, the centiles are splayed and the population is skewed towards being overweight
- The 50th centile is the median for the population
- A measurement on the 98th centile means only 2% of the population are taller or heavier than the child
- A measurement on the 2nd centile means that only 2% of the population are lighter or shorter than the child

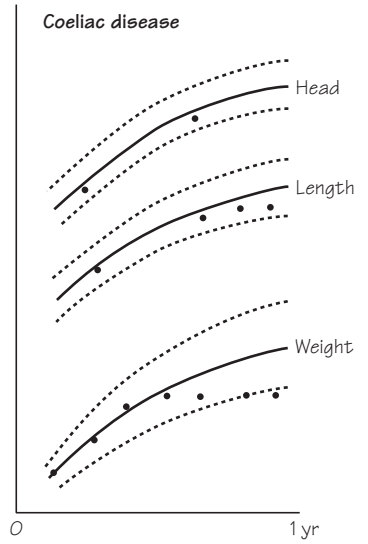
Principles of plotting
- The child's measurement should be marked with a dot (not a cross or circle)
- Correct for prematurity up to the age of 2 years
- Height should follow one centile between 2 years and puberty
- Plateauing of growth and weight, or height less than 0.4% merits an evaluation of the child (see Chapter 41).
- Infants may normally cross centiles in the first year or two, but consider whether failure to thrive is a problem (see Chapter 42).
- A child's final height is expected to fall midway between the parents' centile positions

Examples of growth charts



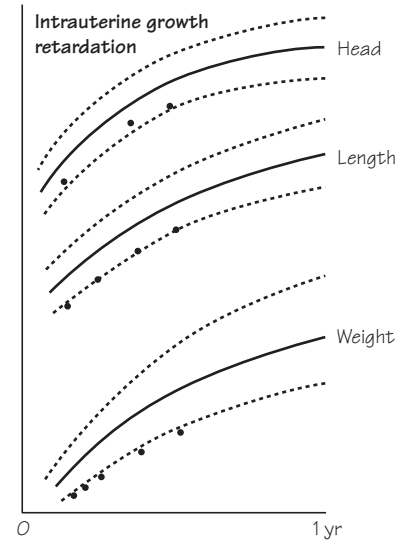
Premature baby

- The baby was born at 30 weeks gestation and is now 26 weeks old
- Corrected age is 16 weeks



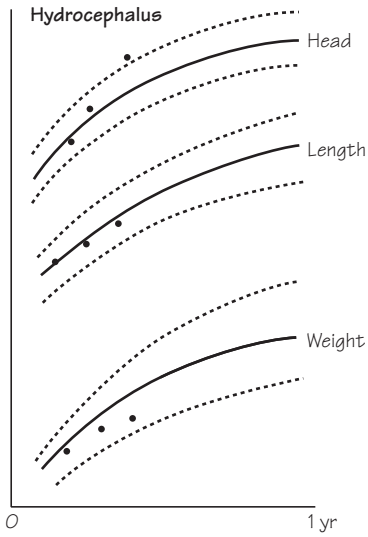
Coeliac disease

- Note fall-off in weight at time of weaning when wheat was introduced
- The fall-off in length occurs later



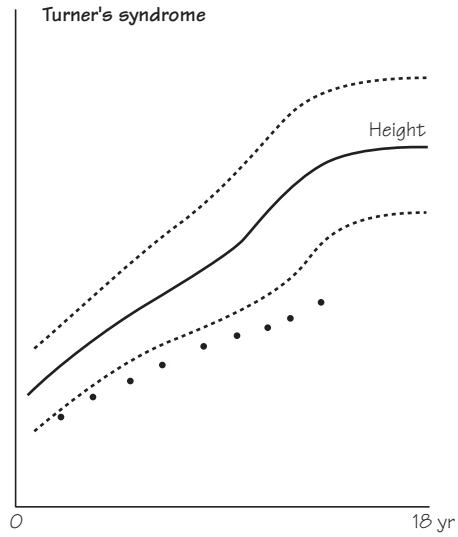
Intrauterine growth retardation (IUGR)

- Low birth-weight baby
- Many IUGR babies show catch-up but this baby clearly has not, and may have reduced growth potential
- The IUGR probably started early in pregnancy because OFC and length are also affected



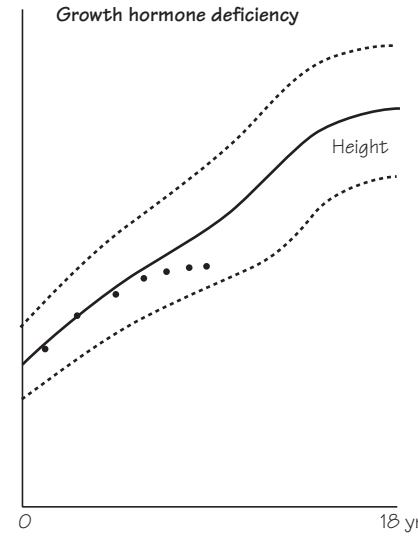
Hydrocephalus

- The head circumference is crossing centile lines upwards
- A normal but large head would grow above but parallel to the centile lines



Turner's syndrome

- Poor growth
- Absence of pubertal growth spurt
- The child should have been referred for growth-promoting treatment when young



Growth hormone deficiency

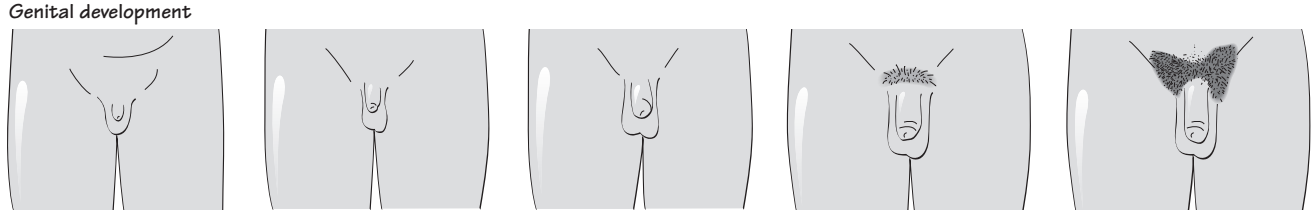
- Note the fall-off in height
- GH deficiency is rare
- It can be congenital, but as growth has plateaued at the age of 6 years, pituitary deficiency due to a brain tumour must be considered

Puberty

Puberty is evaluated by clinical examination of the genitalia, breasts and secondary sexual characteristics. The scale used is known as Tanner staging.

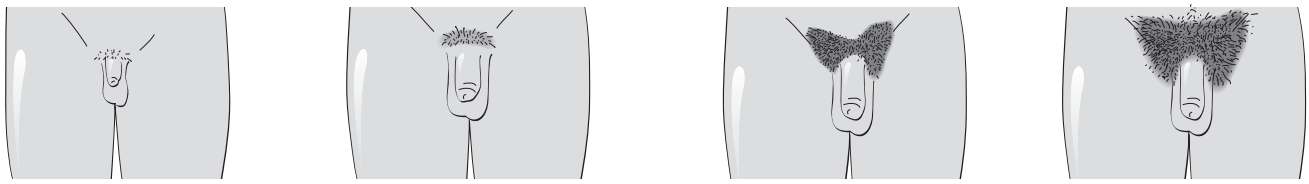
Boys

Genital development



Stage 1 Stage 2 Stage 3 Stage 4 Stage 5

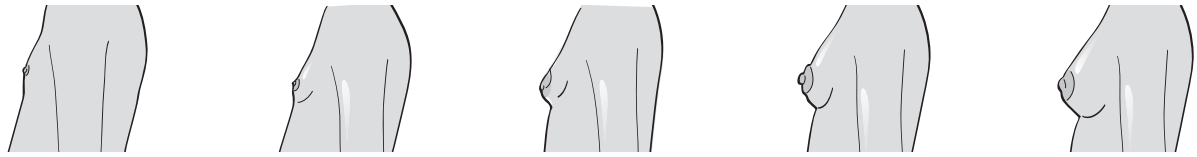
Pubic hair growth



Stage 2 Stage 3 Stage 4 Stage 5

Girls

Breast development



Stage 1 Stage 2 Stage 3 Stage 4 Stage 5

Pubic hair growth

Stage 2 Stage 3 Stage 4 Stage 5

Principles of puberty

- The first signs of puberty are usually testicular enlargement in boys, and breast budding in girls.
- Puberty is precocious if it starts before the age of 8.5 years in girls and 9.5 years in boys.
- Puberty is delayed if onset is after 13 years in girls and 14 years in boys.
- A growth spurt occurs early in puberty for girls, but at the end of puberty for boys.
- Menarche occurs at the end of puberty. Delay is defined as no periods by 16 years of age.

6 Development and developmental assessment

Gross motor development

Prone position

Birth
Generally flexed posture

6 weeks
Pelvis flatter

4 months
Lifts head and shoulders with weight on forearms

6 months
Arms extended supporting chest off couch

Pull to sit

Birth
Complete head lag

6 weeks
Head control developing

4 months
No head lag

Sitting

6 weeks
Curved back, needs support from adult

6–7 months
Sits with self-propping

9 months
Gets into sitting position alone

Standing and walking

6 months
Stands with support

10 months
Pulls to standing and stands holding on

12 months
Stands, and walks with one hand held

15 month
Walks independently and stoops to pick up objects

Fine motor development

Grasping and reaching

4 months
Holds a rattle and shakes purposefully

5 months
Reaches for object

6 months
Transfers object from hand to hand

7 months
Finfinger feeds

Building bricks

12 months
Gives bricks to examiner

15 months
Builds a tower of two cubes

18 months
Builds a tower of three to four cubes

Manipulation

5 months
Whole hand grasp

9 months
Immature pincer grasp

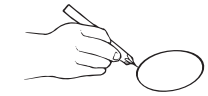
10 months
Points at bead

12 months
Mature pincer grasp

Pencil skills

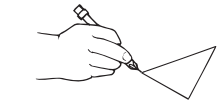
18 months
Scribbles with a pencil

3 years
Draws a circle



4 years
Draws a cross

5 years
Draws a triangle



Speech and language development

Speech

3 months
Vocalizes

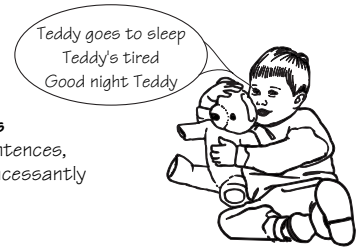
8 months
Double babble

12 months
Two or three words with meaning

18 months
10 words

24 months
Linking two words

3 years
Full sentences, talks incessantly



Social development

6 weeks
Smiles responsively

16 weeks
Laughing out loud

7 months
Stranger anxiety

9 months
Peek a boo, waves bye bye

15 months
Drinks from a cup

18 months
Spoon-feeding self

About $2^1/_2$ years
(very variable)
Toilet trained by day

3 years
Dresses self (except buttons)

Development and developmental assessment

Parents are always interested in their child's developmental progress and are usually concerned if any aspect is delayed. It is an important indicator of a child's wellbeing, and delay or abnormal development may indicate serious limitations for later life. Advanced development of language and fine motor skills may be a sign of intelligence.

An assessment of developmental progress is important at all clinical encounters with children. You firstly need to know the normal progression of development in the early years, and then develop your skills in assessing babies and children of different ages.

How to perform a developmental assessment

- Young children often will not co-operate so make the most of observing them informally. You may have to rely heavily on parental report, especially for language skills.
- Be systematic and evaluate the four developmental areas in turn– gross motor, fine motor/adaptive, speech and language, social and also assess hearing and vision.
- It is hard to remember all the milestones, so make sure you learn the essential milestones given below. Then ensure you know how skills progress. You can always check the age at which skills are acquired after you have finished your evaluation.
- Remember that you need to correct for prematurity until the child is two years old.
- Present the tasks one at a time and try to have as few distractions for the child as possible.
- The most useful equipment to have is bricks and a crayon.

Milestones that are essential to remember.

Age	Milestone
4–6 weeks	Smiles responsively
6–7 months	Sits unsupported
9 months	Gets to a sitting position
10 months	Pincer grasp
12 months	Walks unsupported
	Two or three words
18 months	Tower of three or four cubes
24 months	Two to three word sentences

Developmental warning signs

There is a wide variation in the age at which milestones are met. It is therefore important to be aware when it is abnormal if a child has not yet acquired certain skills.

Developmental warning signs.

At any age	Maternal concern Regression in previously acquired skills
At 10 weeks	No smiling
At 6 months	Persistent primitive reflexes Persistent squint Hand preference Little interest in people, toys, noises
At 10–12 months	No sitting No double syllable babble No pincer grasp
At 18 months	Not walking independently Less than 6 words Persistent mouthing and drooling
At 2½ years	No 2–3-word sentences
At 4 years	Unintelligible speech

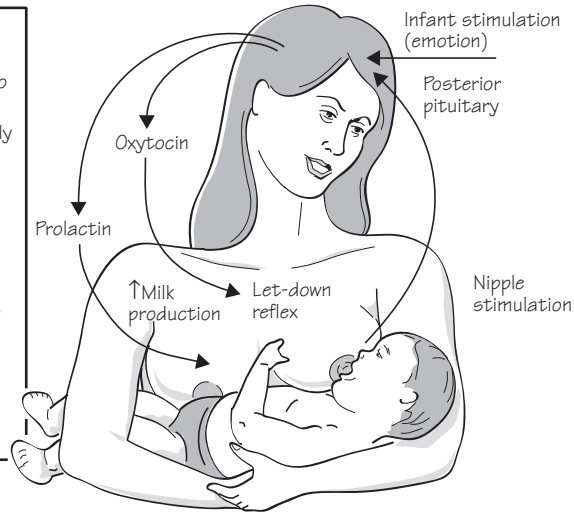
KEY POINTS

- Delay in one area is often not of concern and maybe familial.
- Delay in all areas is a cause for concern (see Chapter 46, global developmental delay).
- Do not forget to correct prematurity.
- See p. 100 for causes of delayed development.

7 Infant nutrition

Breast-feeding

Ways to encourage successful breast-feeding
- Introduce concept of breast-feeding to both parents antenatally
- Put the baby to the breast immediately after delivery
- Allow the baby to feed on demand, especially in the early days
- Avoid offering any formula feeds
- Ensure mother receives good nutrition and plenty of rest
- Provide skilled breast-feeding advisors to help mother through any initial problems with breast-feeding
- Ensure correct 'latching on' with the baby's mouth wide open and good positioning



Lactation
- At birth, prolactin levels rise sharply and this is further stimulated by the infant sucking at the breast. Prolactin determines milk production from the breast alveoli, and is increased by the frequency, duration and intensity of sucking
- The actual flow of milk is aided by the 'let-down' reflex. Rooting at the nipple causes afferent pathways to stimulate the posterior pituitary to secrete oxytocin, which stimulates the smooth muscle around the alveolar ducts to express the milk from the breast. The let-down reflex can be stimulated by hearing the baby cry or by contact with the baby, and can be inhibited by stress or embarrassment
- The majority of the milk is taken from the breast in the first 5 min and this may be followed by non-nutritive sucking

Advantages of breast-feeding
- Perfect balance of milk constituents
- Little risk of bacterial contamination
- Anti-infective properties (IgA, macrophages, etc.)
- Ideal food for brain growth and development
- Convenient
- No expense of purchasing milk
- Psychologically satisfying
- Reduces risk of atopic disorders

Theoretical problems with breast-feeding (rare)
- Can initially be exhausting for the mother
- Can transmit infection (e.g. HIV, although in developing countries the best advice is still to exclusively breast-feed)
- Some drugs can be excreted in breast milk (e.g. warfarin)

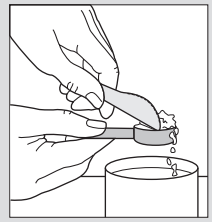
Weaning
- 0–4 months: breast or formula milk only
- 4–6 months: puréed or liquidized foods
- 6–9 months: finger foods, juice in a cup
- 9–12 months: three meals a day, with family
- >1 year: cow's milk in a beaker or cup; adult-type food chopped up

Formula milk feeds

- Formula milks are based on cow's milk, but are carefully adjusted to meet the basic nutritional requirements of growing infants. The fat component is generally replaced with polyunsaturated vegetable oils to provide the correct essential fatty acids. Minerals, vitamins and trace elements are then added. Milks with more casein are produced for 'hungrier' babies aged 4–6 months, and 'follow-on' milks with more iron for babies over 6 months.
- Formula milk is usually made up from a dry powder, by adding one level measure of powder to each 30 ml (1 fl.oz) of cooled boiled water. Great care must be taken to sterilize the bottles and teats carefully to avoid introducing infection. The milk is then re-warmed prior to feeding. This should not be done in a microwave as pockets of milk may be heated to dangerous levels.

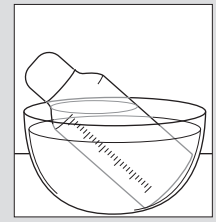

1. Sterilize the feeding bottle


2. Add the appropriate volume of cooled boiled water to the bottle


3. Add 1 level scoop of milk powder to each 30 ml of water


4. Shake bottle well


5. Keep in fridge until ready to feed


6. Rewarm the feed to room temperature or body temperature prior to feeding

Nutrition from birth to school age

Milk provides all the nutrients needed by newborn infants for the first 4–6 months of life. Breast milk is the ideal milk for human babies, but formula milk may be needed as an alternative in some cases. The newborn infant has high calorie and fluid requirements and to achieve optimal growth requires approximately 150 ml/kg/day fluid and 110 kcal/kg/day (462 kJ/kg/day). About 40% of this energy comes from carbohydrate (mostly lactose) and 50% from fat. Milk also contains protein in the form of casein, lactalbumin and lactoferrin. Colostrum is the thin yellow milk produced in the first few days which is high in immunoglobulins.

Infants also require adequate amount of minerals such as calcium and phosphate, as well as vitamins and trace elements. Breast milk is deficient in Vitamin K, and so all newborn infants are given vitamin K at birth to prevent haemorrhagic disease of the newborn. Weaning onto solids usually starts around 4 months, and infants should not have cows milk until they are over a year.

Technique of breast-feeding

Mothers should be encouraged to put their babies to the breast soon after delivery. Little milk is produced but the suckling stimulates lactation. Colostrum is produced in the first days which is rich in energy and anti-infective agents. It is important that the baby is taught to 'latch-on' to the breast properly with a widely open mouth so that the areolar and not just the nipple is within the babies mouth. The majority of the milk is taken by the baby in the first 5 min. Time after this is spent in non-nutritive suckling. Mothers can feel their breast 'emptying'. Babies should not be pulled off the breast, but the suck released by inserting a clean finger at the side of the babies mouth. Each feed should start on the alternate breast. In the first few days the breasts may become painfully engorged with milk and the nipples sore, especially if the baby's position is not optimal. Mothers need a lot of encouragement and advice to get through this time. It is important to try to avoid alternating breast and formula feeds. Formula feeds should only be introduced if breast-feeding is contraindicated or has failed completely. It is not appropriate to 'top-up' with formula or use bottles to give the mother a rest. This may help in the short term but usually leads to production of milk tailing off and breast-feeding failing altogether.

Weaning

Most healthy infants do not require weaning until 4–6 months of age, although some premature infants may want solids earlier. Generally cereals, rusks or rice-based mixtures are introduced first, mixed with expressed breast milk or formula milk. This semi-solid mixture can be given by a spoon before milk feeds. Puréed fruit or vegetables are also suitable. Modern baby cereals are gluten free which may be associated with a fall in the incidence of coeliac disease (see Chapter 25). As the child grows older the feeds can become more solid and are given as three meals a day. From 6 to 9 months they will enjoy finger-feeding themselves and can chew on rusks or toast. From about 9 months they can generally eat a mashed or cut-up version of adult food. Undiluted full-fat pasteurized cows milk can be given from 1 year of age. Earlier introduction of cows milk or persistence of exlusive breast-feeding can lead to iron deficiency. Vitamin supplements may be needed from 6 months in breast-fed babies, until they are on a full mixed diet.

Nutrition in the preschool years

As a toddler the child becomes more adept at holding a spoon and can feed independently, and can drink from a beaker or cup. Milk is no longer the main source of nutrients, although the child should still drink a pint a day. Whole-fat milk should be used until age 5 to provide plenty of calories. A well-balanced diet should include food from the four main groups:

- Meat, fish, poultry and eggs.
- Dairy products (milk, cheese, yoghurt).
- Fruit and vegetables.
- Cereals, grains, potatoes and rice.

In order to avoid dental caries it is important to avoid very frequent snacking on sugary foods or drinks—three meals and two snacks is recommended, although this may be adapted to the individual child. Iron-deficiency anaemia is common at this age, due to high requirements for growth and poor dietary intake, especially in the 'faddy eater'. Vitamin C present in orange juice can enhance iron absorption from the gut.

Nutrition in the school-age child

At school, children have to learn to eat food out of the family setting. They usually have a midday meal, and fruit or milk may be provided at break times. The principles of healthy eating should be maintained, although peer pressure to eat crisps or sugary snacks is high. Schools have an educational role to play in encouraging healthy eating and a healthy lifestyle. During adolescence there is a greater energy requirement to allow for increased growth. This may coincide with a lifestyle that leads to snacking and missing meals, or to restrictive dieting or over-consumption of fast food. Obesity and eating disorders often have their onset around this time.

KEY POINTS

- Breast milk provides ideal nutrition for babies.
- The optimum time for weaning is 4–5 months.
- Formula feeds need to be made up carefully to avoid infection.
- 'Doorstep' cows milk should not be used until 1 year.
- Full-fat milk is recommended until 5 years of age.
- Toddlers need to be allowed to explore food and develop independent eating habits.

8 Common problems for parents

The crying baby
- Wet or dirty nappy
- Too hot or too cold
- Hungry
- Wind
- Colic
- Environmental stress
- Reflux oesophagitis
- Teething

If sudden severe crying, consider:
- Any acute illness
- Otitis media
- Intussusception
- Strangulated inguinal hernia

Temper tantrums
- Normal, peak at 18–36 months
- Screaming
- Hitting
- Biting
- Breath-holding attacks (see p. 74)

Strategies that may help
- Avoid precipitants such as hunger and tiredness
- Divert the tantrum by distraction
- Stay calm to teach control
- Reward good behaviour
- Try to ignore bad behaviour until calm
- Use time-out

Sleeping problems
- Difficulty getting to sleep
- Waking during the night
- Sleeping in parents bed
- Nightmares and night terrors

Eating problems in toddlers
- Food refusal
- Faddy eating—only eating one or two types of food
- Overeating
- Battles over eating and mealtimes
- Snacking
- Excessive drinking of juice

Unwanted habits
- Thumb sucking
- Nail biting
- Masturbation
- Head banging
- Hair pulling
- Bedwetting
- Encopresis (passing faeces in inappropriate places)

Aggressive behaviour
- Temper tantrums
- Hitting and biting other children
- Destroying toys
- Destroying furniture
- Commoner in boys and in larger families
- May reflect aggression within family
- Requires calm, consistent approach
- Avoid countering with aggression
- Use time-out and star charts

What you need from your evaluation

History

- Ask what is troubling the parents most—is it the child or other stresses in their lives, such as tiredness, problems at work or marital problems?
- What are the triggers for difficult or unwanted behaviour? Does it occur when the child is hungry or tired, or at any particular time of day?
- Colic tends to occur in the evenings; tantrums may be commoner if the child is tired
- Does the behaviour happen consistently in all settings or is it specific to one place, e.g. the toddler may behave well at nursery but show difficult behaviour at home?
- Does the behaviour differ with each parent?
- How do the parents deal with the behaviour—do they get angry or aggressive, are they consistent, do they use bribery or do they give in to the toddler eventually?
- What strategies have the parents already tried to deal with the situation?
- Is there any serious risk of harm? Some behaviour, such as encopresis or deliberate self-harm, may reflect serious emotional upset. Most toddlers who are faddy eaters are growing well and do not suffer any long-term nutritional problems

Examination

- Not usually required
  If the parents are concerned about sudden-onset or severe crying in a baby, it is important to exclude serious infection such as meningitis or urinary tract infection, intussusception, hernias and otitis media
- Babies with colic are usually less than 3 months old, go red in the face with a tense abdomen and draw up their legs. The episodes start abruptly and end with the passage of flatus or faeces

Management

- In most cases the parents can be reassured that the behaviour is very common, often normal and that with time and common sense it can be controlled
- With tantrums it can be helpful to use the **ABC** approach:
  - A What **a**ntecedents were there? What happened to trigger the episode?
  - B What was the **b**ehaviour? Could it be modified, diverted or stopped?
  - C What were the **c**onsequences of the behaviour? Was the child told off, shouted at or given a cuddle?
- Generally, it is best to reward good behaviour (catch the child being good) and ignore bad behaviour. Star charts can be very useful: the child gets a star for good behaviour (staying in bed, etc.) and then a reward after several stars
- Parents must try hard not to be angry or aggressive as this may reinforce the bad behaviour in the child

Common emotional and behavioural problems

Such problems are seen so often that many would regard them as normal, although in a small minority the behaviour can be so disruptive as to cause major family upset. The general practitioner or paediatrician should be comfortable giving basic psychological advice on behaviour management to help parents through what can be a stressful, exasperating and exhausting phase of their child's development.

Crying babies and colic

Crying is usually periodic and related to discomfort, stress or temperament. However, it may indicate a serious problem, particularly if of sudden onset. In most instances it is just a case of ensuring that the baby is well fed, warm but not too hot, has a clean nappy, comfortable clothes and a calm and peaceful environment. A persistently crying baby can be very stressful for inexperienced parents. It is important that they recognize when they are no longer coping and are offered support.

Infantile colic is a term used to describe periodic crying affecting infants in the first 3 months of life. The crying is paroxysmal, and may be associated with hunger, swallowed air or discomfort from overfeeding. It often occurs in the evenings. Crying can last for several hours, with a flushed face, distended and tense abdomen and drawn-up legs. In between attacks the child is happy and well. It is important to consider more serious pathology such as intussusception or infection. Colic is managed by giving advice on feeding, winding after feeds and by carrying the baby. Colic is not a reason to stop breast-feeding, although the mother going onto a cows milk-free diet can occasionally help. Various remedies are available but there is little evidence for their effectiveness. Infantile colic usually resolves spontaneously by 3 months.

Feeding problems

Once weaned, infants need to gradually move from being fed with a spoon to finger feeding and feeding themselves. This is a messy time, but the infant needs to be allowed to explore their food and not be either force-fed or reprimanded for making a mess.

Toddler eating habits can be unpredictable—eating huge amounts at one meal and sometimes hardly anything at the next. At this age, mealtimes can easily become a battle and it is important that they are kept relaxed and the child is not pressurized into eating. Small helpings that the child can complete work best, and second helpings can be given. Eating together as a family encourages the child to eat in a social context. Feeding at mealtimes should not become a long protracted battle!

Sleeping problems

Babies and children differ in the amount of sleep they need and parents vary in how they tolerate their child waking at night. In most cases sleeping 'difficulties' are really just habits that have developed by lack of establishing a clear bedtime routine. Difficulty sleeping may also reflect conflict in the family or anxieties, for example about starting school or fear of dying. Successfully tackling sleeping problems requires determination, support and reassurance.

- **Refusal to settle at night**. Difficulty settling may develop if babies are only ever put to bed once they are asleep. A clear bedtime routine is important for older children—for example a bath, a story and a drink.
- **Waking during the night**. This often causes a lot of stress as the parents become exhausted. It is important to reassure the child, then put them back to bed quietly. Sometimes a technique of 'controlled crying' can be helpful—the child is left to cry for a few minutes, then reassured and left again, this time for longer. Taking the child into the parental bed is understandable, but usually stores up problems for later when it is difficult to break the habit.
- **Nightmares**. The child wakes as the result of a bad dream, quickly becomes lucid and can usually remember the content. The child should be reassured and returned to sleep. If particularly severe or persistent, nightmares may reflect stresses and may need psychological help.
- **Night terrors**. Night terrors occur in preschool years. The child wakes up confused, disorientated and frightened and may not recognize their parent. They take several minutes to become orientated and the dream content cannot be recalled. These episodes should not be confused with epilepsy. They are short-lived and just require reassurance, especially for the parents.

Temper tantrums

These are very common in the third year of life (the 'terrible two's') and are part of the child learning the boundaries of acceptable behaviour and parental control. They can, however, be extremely challenging, especially when they occur in public!

The key to dealing with toddler tantrums is to try to avoid getting into the situation in the first place. This does not mean giving in to the child's every demand, but not letting the child get over tired or hungry, setting clear boundaries and enforcing these in a calm, consistent and controlled manner. There will still be times when tantrums occur—where safe to do so they are best ignored until the child calms down. If this fails then 'time-out' can be a useful technique. The child is taken to a safe quiet environment, such as a bedroom, and left for a few minutes until calm. This is usually very effective as it removes the attention the child desires, and allows the parents time to control their own anger.

Unwanted or aggressive behaviour

Young children often have aggressive outbursts which may involve biting, hitting or scratching other children. These require consistent firm management, with use of time-out and star charts for good behaviour. It is important not to respond with more aggression, as this sends conflicting messages to the child. If aggressive behaviour is persistent it is important to explore other tensions or disturbances within the family. At school age, the school will often need to be involved in a behaviour management programme.

Unwanted behaviours such as thumb-sucking, hair-pulling, nail-biting and masturbation are also common in young children. The majority can be ignored and will resolve with time. Masturbation can usually be prevented by distracting the child or dressing them in clothes which make it more difficult. The older child should not be reprimanded but informed that it is not acceptable in public.

KEY POINTS

- Emotional and behavioural problems are extremely common, to the point of being part of normal child development.
- Parents need to be encouraged that they can manage most behaviour with a clear strategy.
- A calm, confident and consistent approach to the child's behaviour is recommended.
- Parents should reward good behaviour and try to minimize attention given to undesirable behaviour.

9 Adolescent issues

Adolescence is the time between childhood and full maturity and is when growing-up occurs. It is a time of great physical, psychological and social change, and can be a time of considerable stress for adolescents and their parents.

Physical changes
- Growth spurt occurs—may feel 'gangly'
- Secondary sex characteristics develop:
  - -pubic hair
  - -facial hair and testicular enlargement in boys
  - -breast enlargement in girls
- Voice deepens in boys
- Girls undergo menarche and become fertile
- Acne may develop
- Gynaecomastia may develop in boys

'Tasks' of adolescence
- Establish sense of identity
- Achieve independence
- Achieve sexual maturity
- Take on adult responsibility
- Develop adult thinking

Psychological problems
- Eating disorders
- Depression
- Self-harm
- Overdosing on medicines
- Suicide

Psychological changes
- Develop insight
- Able to use abstract reasoning
- Develop logical thought
- Able to reason morally, often leading to questioning of parents and awareness of social injustice in the world
- Search for independence
- May be emotional turmoil and conflict
- Experimentation and risk-taking behaviour

Health issues
- Contraception and safe sex
- Acne
- Eating disorders
  - -anorexia
  - -bulimia
  - -obesity
- Chronic illness (diabetes, cystic fibrosis, Crohn's disease, asthma)
- Health promotion
- Issues of consent

Health destructive behaviour
- Alcohol
- Smoking
- Drug use
- Substance abuse
- Accidents
- Unsafe sex
  - -sexually transmitted disease
  - -unwanted pregnancy
  - -teenage pregnancy
- Excessive dieting

Vulnerable adolescents

Certain groups of adolescents are at particular risk of a poor outcome through adolescence and may also have difficulty accessing healthcare. They include:
- those with chronic illness (e.g. diabetes)
- physical disability or learning difficulties
- the homeless and unemployed
- victims of physical, emotional or sexual abuse
- those who are pregnant
- some ethnic minority groups
- those from disrupted homes

Social change
- Still dependent on parents financially and for housing
- Greater freedom and flexibility
- Self-motivation and self-discipline expected by school
- Sexual interest and activity increases; most experience some form of sexual activity
- Face leaving school and moving to higher education, work, financial independence or unemployment

Approach to the adolescent

- Adolescence is generally a time of life when illness is rare
- Partly because of this, healthcare facilities for adolescents are poor, often falling between paediatric and adult care
- The low rate of contact with doctors means health promotion must be delivered to the adolescent
- Adolescents may be concerned about confidentiality when seeing their family doctor
- Drop-in clinics can offer immediate advice on health issues, counselling for emotional and personal problems and contraceptive advice
- The way in which health professionals treat adolescents is important

How to treat adolescents

- Take time to listen
- Show respect for their emerging maturity
- Allow them to express their concerns
- Avoid making judgmental statements
- Assure confidentiality (but make it clear there are times when confidentiality must be broken, e.g. after disclosure of ongoing abuse or if others are at risk)
- Respect the need for privacy—offer to see them without their parents

Destructive health behaviour

• **Alcohol**. For many, drinking is a regular part of their teenage lifestyle: 77% of boys and 66% of girls aged 15 drink 6–10 units of alcohol per week.

• **Drugs and substance abuse**. An increasing proportion of teenagers use drugs. Cannabis and Ecstasy are commonly used and about 5% of teenagers experiment with hard drugs (cocaine, heroin, amphetamines). Solvent abuse is also common, especially amongst disadvantaged teenagers. Signs of drug use may include: mood changes, loss of appetite, loss of interest in schoolwork and leisure interests, drowsiness or sleeplessness, furtive behaviour, stealing, unusual smells on clothing.

• **Smoking**. While the incidence of adult cigarette smoking is decreasing, teenage smoking is increasing, especially amongst teenage girls. Among 11- to 15-year-olds 10% smoke regularly. Those who smoke are more likely to try other drugs and to drink alcohol. There is an increased risk of bronchitis, emphysema, lung cancer and cardiovascular disease in those who smoke from an early age.

• **Accidents**. Road traffic accidents (pedestrian, car and motorbike) are the leading cause of death in this age group. Alcohol and failure to wear seat belts and crash helmets increase the risks. Sports injuries and drowning are also common at this age.

• **Unsafe sex**. 35% of girls and 45% of boys have experienced sex by the age of 16. Most do not use contraception initially. They are at risk of sexually transmitted disease, including human immunodeficiency virus (HIV), and pregnancy. Human papilloma virus is now thought to be a major risk factor for cervical carcinoma.

Sexual health issues

• **Menstrual complaints**

  • Amenorrhoea is often physiological as periods may be very irregular or scanty for months after the onset of menarche. Stress associated with moving schools or exams can disrupt periods and those undergoing intense athletic training may develop amenorrhoea due to disruption of the hypothalamic–pituitary axis. Pregnancy should always be considered as a cause.

  • Menorrhagia (heavy periods) is common in the year after menarche due to anovulatory cycles.

  • Dysmenorrhoea (painful menstrual cramps) is a common symptom and a common cause of missing school. Treatment includes prostaglandin synthetase inhibitors (e.g. mefanamic acid) or the combined oral contraceptive pill, which also offers contraception.

• **Teenage pregnancy**. 40% of sexually active teenagers become pregnant within 2 years of initiating intercourse. Britain has the highest teenage pregnancy rate in Europe. Many teenage girls get pregnant deliberately, and there are increased risks for the mother (pre-eclampsia, preterm labour, postnatal depression) and for the baby (higher infant mortality, sudden infant death syndrome (SIDS), accidental and non-accidental injury and low birthweight).

• **Abortion**. One-third of teenage pregnancies end in abortion, which is still a common form of contraception in this age group. There is a need for careful counselling to prevent long-term psychological trauma. The 'morning-after pill' is available and can be taken up to 72 h after unprotected intercourse.

• **Contraception**. More adolescents are becoming sexually active at younger ages, and less than half use any contraception at the time of the first intercourse. Motivation, information and ready access to contraception are all required if teenage pregnancy is to be prevented. Condoms are cheap and prevent spread of sexually transmitted disease including HIV. They do have a failure rate and require some motivation to be used. The oral contraceptive pill is the most reliable method if taken correctly but requires premeditation. There is a small risk of thrombotic events, especially in cigarette smokers. Intrauterine devices are not usually offered to nulliparous women and carry an increased risk of pelvic infection.

Eating disorders

Eating disorders are commoner in girls than boys, and often start as innocent dieting behaviour. The age of onset is decreasing with 20% occurring before 13 years of age. Eating disorders are characterized by an intense fear of becoming fat and a distorted body image, so that even extremely wasted individuals feel they are fat. There may be preoccupation with food and bizarre eating behaviours.

• **Anorexia nervosa**—extreme dieting with restriction of fat and carbohydrate intake is used to control weight. There may also be excessive physical activity. One definition of anorexia is the loss of more than 20% of weight-for-height. Features include emaciation, amenorrhoea, constipation, hair loss and lanugo hair. Bradycardia, hypothermia and hypotension develop with extreme malnutrition. The mortality rate for anorexia can be up to 10% so it should be taken seriously. Management involves refeeding up to the desired weight, either as an out-patient or by supervised eating in an in-patient facility. Occasionally nasogastric feeding may be required. Psychotherapy and behavioural modification techniques are then used to try to maintain the desired weight. Antidepressants should be used if there is coexisting depression. The overall prognosis is good, although about a quarter develop relapsing episodes of anorexia and 5% commit suicide, often in adult life.

• **Bulimia**—bouts of binge eating, followed by purging with laxatives or by inducing vomiting. Weight is usually normal or increased. Oesophagitis, parotid swelling and staining of the teeth are all signs of chronic vomiting. Psychotherapy and behavioural management can try to encourage a normal self-image and more normal eating behaviour.

KEY POINTS

- Adolescence is a time of great physical, psychological and social change.
- Adolescents often show health-destructive and risk-taking behaviour.
- Eating disorders are not uncommon and should be recognized.
- Health workers need to find novel ways of engaging with adolescents, especially particularly vulnerable groups.

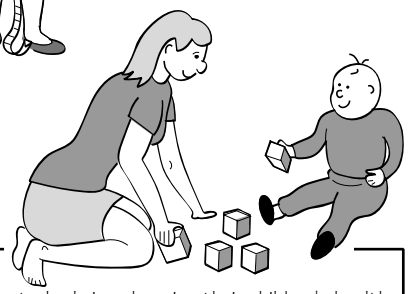
10 The child health service

Who is involved?

Health visitors
are nurses specially trained in childcare and development, and are responsible for most of the child health surveillance and promotion programme for preschool children. They work with general practitioners, and run child health clinics, visit at home and provide support, particularly for families identified as being in need or at risk

School nurses
are specially trained to work with children at school. They are responsible for identifying children with medical needs, facilitating their care at school, liaising between professionals and supplying medical information to school staff. They review every child at school entry

General practitioners
now have responsibility for the routine aspects of the pre-school child health programme, i.e. routine examinations and immunizations (although community paediatricians may still run baby clinics in disadvantaged areas)



Parents
have a central role in enhancing their children's health and should be seen as partners in child health surveillance

Community paediatricians
specialize in working in the community. Some are specialists in child protection, disability or audiology

What does the child health service provide?

- Guidance in areas of child health
- Health promotion and education
- Prevention of disease by immunization
- Measurement and recording of physical growth
- Monitoring of developmental progress
- Detection of abnormalities
- Identification of children in need

What records are kept?

- **Parent-held child health records (the red book)**
  Parents are issued with a child health record at the time of their child's birth. It records child health surveillance checks, growth, parental observations, primary care and dental and hospital visits, and health education and advice
- **Professional records**
  Professionals keep their own records of contact. Computer-based systems are increasingly used, particularly for child health surveillance
- **Special registers**
  Many districts keep registers of children with special needs or chronic illness. These are useful for providing parents with information about services, keeping track of referral and review, anticipating needs and auditing the service. Parental permission is required before placing a child on a register
- **Child protection register**
  Social services departments keep a register of children who have been abused or neglected so that professionals can readily determine if a child or others in the family are known to be at risk

Health education and promotion

Young families are growing up more isolated, without support of extended families. The child health programme provides information and advice to parents, and promotes parenting skills.

Baby care

Advice is given about issues such as clothing, bathing, handling and positioning the baby. Information is also given about normal development, what to expect from the child, how to promote learning and how to recognize developmental difficulties. Guidance is given for common medical problems, and how to manage them.

Nutrition

Addressing nutritional issues forms a major part of a health visitor's work. It includes promotion of breast-feeding, advice about weaning, dealing with eating difficulties commonly encountered in toddlers, and education about healthy diets for the entire family.

Behavioural problems

Behavioural concerns around crying, sleep and temper tantrums are universal. Advice and support in the early stages can avoid them developing into major problems.

Dental care

Information should be provided about dental hygiene, the use of fluoride, and regular dental check-ups.

Passive smoking

Children exposed to passive smoking are at greatly increased risk for respiratory disorders. Avoidance of exposing children to smoke at home is an important health promotion issue.

Accidents

Accidents are the commonest cause of mortality in childhood and an important cause of morbidity. Most accidents occur in the home, so education of parents can have an important impact on their prevention. Areas which should be addressed include:

- Use of car seats and belts.
- Road safety and cycle helmets.
- Gates on stairs, guards on windows.
- Caution in the kitchen.
- Installation of smoke detectors.
- Fire guards, flame proof clothing.
- Covering electric sockets, avoid trailing flexes on kettles and irons.
- Never leaving young children alone in the bath.
- Keep medicines and poisons out of reach with locks on cupboards.
- Keep small toys away from toddlers,and no nuts before age 5 years.

Obesity

Obesity is becoming epidemic in older children and there is serious concern about the impact this will have on their health as adults. There is also occasionally concern that excessive weight gain has a 'medical cause'. Medical causes such as hypothyroidism, Cushing's disease and hypothalamic syndromes are extremely rare and always affect (reduce) a child's growth in height. If an overweight or obese child's height gain is normal then no investigations need be done. Attempts should be made to ensure the child has a healthy balanced diet and an increase in physical activity. Reversing obesity is hard and so preventive strategies are important in the childhood population.

Health promotion in school

School provides an invaluable opportunity to educate the young about healthy living, and hopefully the school years are a time when adjustments in lifestyle can be made more easily than later on in life. Issues of particular importance which are addressed are:

- Nutrition.
- Physical activity.
- Reducing risk factors for obesity.
- Drugs and alcohol abuse.
- Contraception and safe sex.
- Sexually transmitted disease.
- Smoking.
- Healthy relationships.
- Parenting skills.

Child protection

It is the duty of professionals (and indeed anyone) to report concerns regarding the possibility that a child is the victim of neglect, non-accidental injury or emotional or sexual abuse. The role of the child health service includes:

- Reporting suspected victims of abuse and neglect.
- Following children at risk for abuse and neglect—health visitors are particularly well placed for this.
- Providing guidance to reduce the risk of abuse.
- Liaison with social services.
- Monitoring children in care and on the Child Protection Register (see also Chapter 45).

Child protection procedures for an abused child

If there is any suspicion that a child has suffered abuse or neglect, the child is referred immediately to a paediatrician experienced in child protection work. If it is concluded that the child has been abused or is at risk of abuse the social services department is immediately informed.

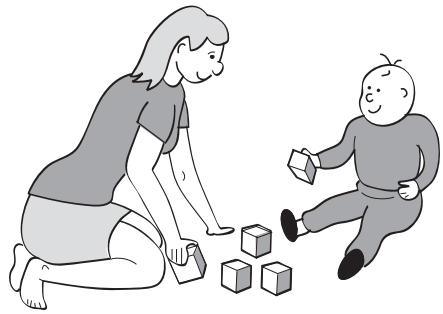
If the child is deemed to be in danger, or further assessment is required, he or she is admitted to a place of safety, usually a hospital ward or social services institution until a fuller enquiry can be made. An emergency care order can be obtained from court if the family resists admission or investigation.

The social work team usually takes the lead in planning the strategy for management. Initial policy is worked out at a case conference, attended by all professionals involved, and the parents. Many children are allowed home, initially under supervision and with appropriate support. Occasionally it is necessary to take the child away from the parents. This is generally a hard decision and requires a court order. The child may be placed with another member of the family, in foster care, or in the case of an older child in a group home.

For the child returned home, support must be provided. This may be in the form of placement in a social service day nursery, or voluntary and self help groups may be available to help the parents overcome their difficulties. Social services departments keep a record, the Child Protection Register, of children who have been abused or neglected, so that professionals can readily determine if a child or others in the family are known to be at risk.

11 Child care and school

Child care
Increasingly, mothers are working outside of the home and need to find care for their children. Options include:
- A nanny or minder
- A childminder who takes other children into their own home and has to be legally approved and registered with the Department of Social Services
- A day nursery staffed by nursery nurses and run either privately, by social services departments, or by voluntary organizations
- Family centres, in disadvantaged areas, which provide care and also parenting guidance



Education
Compulsory education begins at age 5 and continues to age 16. For younger children there are opportunities to meet, play and socialize with others.

Preschool
- Mother and toddler groups for children accompanied by a carer
- Playgroups run by trained and registered leaders
- A limited availability of nursery school places from 3 years of age

School
- Primary school from 5 to 11 years of age
- Secondary school from 11 to 16 years
- Sixth form, sixth form college or college of further education at age 16 years

The child with special educational needs
A number of agencies are involved in providing services for the family of a child with special needs.

Education services are responsible for:
- Assessing learning difficulties
- Providing preschool home teaching
- Nursery schooling
- Education in both mainstream and special schools

Social services are responsible for providing:
- Preschool child care
- Relief care such as babysitting services, respite care and home help
- Advice about benefits-a disabled child is entitled to receive the Disability Living Allowance, with a mobility component from the age of 5 years, and parents may receive the Invalid Care Allowance provided they are not in full-time work
- Assessment for services needed on leaving school
- Child protection

Voluntary organizations provide:
- Support and information to families
- Play facilities
- Educational opportunities
- Sitting services

School: mainstream or special school?

Where possible, children with special needs are educated in mainstream schools, with extra help provided in the classroom as needed. This often involves the employment of a special needs assistant for the child, along with physiotherapy, occupational therapy and speech and language therapy support. Mainstream placement has the advantage of integrating children with special needs into a normal peer group in their own locality, and encouraging their adaptation to normal society at an early age. It is also advantageous for other children to learn to live alongside children with disabilities. However, there may be disadvantages as mainstream schools usually have comparatively large classes, may have inadequate support and the buildings may be poorly adapted for the child with physical difficulties.

Special schools, on the other hand, provide expert teaching in small classes, by staff who have an understanding of handicapping conditions. Transport and health service support is also provided. However the disadvantage lies in the child's limited exposure to 'normal life'. Often, a satisfactory compromise between mainstream and special schools is the establishment of special units for children with disabilities in the mainstream setting.

The statement of special educational needs

According to the 1981 Education Act, the education authority is obliged to assess children who are likely to need additional educational provision because of severe or complex difficulties.

Following this assessment a legally binding document is produced known as the 'statement' of special educational needs. The statement is drawn up on the basis of a formal assessment by an educational psychologist, a medical report and reports from any other involved professionals such as therapists and the child's nursery or school. The child's educational needs and the provision which must be made to meet them is clearly outlined. The statement is reviewed on an annual basis. For children with less severe needs, the school is required to provide appropriate help, but no additional resources are supplied by the Education Authority.

Common problems at school

Hyperactivity

Hyperactivity is characterized by poor ability to attend to a task, motor overactivity, and impulsivity. Boys tend to suffer more than girls, and there is often a family history. It is commoner in children with developmental delay, and those who were temperamentally difficult babies. It is also seen in children who have never been given limits or taught to develop self control, and can occur as a reaction to tensions and problems in the home.

Hyperactive children are restless, impulsive and excitable, and fail to focus on any activity for long. They tend to have little sense of danger, and require great vigilance. They often have difficulties on starting school. Management includes routine and regularity in everyday life, with firm boundaries set for behaviour and consistency in discipline. On starting school, the support of the teacher is essential in helping with adjustment.

Attention deficit disorder

Attention deficit disorder (ADD) refers to a difficulty in generally focusing on tasks or activities. It may or may not occur with hyperactivity and is commoner in boys. The child with ADD is fidgety, has a difficult time remaining in his or her seat at school, is easily distracted and impulsive, has difficulty following instructions, talks excessively and flits from one activity to another. Daydreaming occurs when hyperactivity is not a feature.

Management includes a regular daily routine with simple clear rules, and firm limits enforced fairly and sympathetically. Overstimulation and overfatigue need to be avoided, and a structured school programme is required with good home communication. There is increasing evidence that central nervous system (CNS) stimulants such as methylphenidate are effective in improving concentration. Diets restricting artificial colourings or flavourings remain controversial, but do not help the majority of these children.

Aggressive behaviour

Aggressive behaviour can lead to bullying in school and delinquency beyond. If extreme, the psychiatric term, conduct disorder, is applied. Counteracting aggression with more aggression is unproductive, and consistency is required in the management. Both time-out and star charts are positive methods. Staff at school must be involved in the management, in order to address academic or social problems, and to institute behaviour modification.

Teasing and bullying

Ten per cent of children report being bullied once per week and 7% of children are identified as bullies. Bullying tends to be more common in primary schools, and varies from school to school. The child may react by becoming withdrawn, or aggressive or develop psychosomatic symptoms, and it should be considered as a cause of distress whenever a child is disturbed, or refusing to go to school. In schools where bullying is a problem, a whole school approach is most effective, where both the victims and the bullies are helped. The individual child needs help in handling the situation and increasing self-esteem.

Non-attendance at school

Most absences from school occur as a result of illnesses, which are usually minor, but may be prolonged through parental anxiety. In some circumstances, children may be kept at home to help as carers or at work. Two situations where the doctor may become involved are school refusal and truancy. In school refusal everyone knows where the child is, but in the latter the child's whereabouts are unknown during school hours. School refusal may be due to separation anxiety (common on first starting school) or school phobia, which is usually triggered by distressing events, such as problems with peers or teachers. Truancy is commonest in high school. Persistent truancy is associated with generally antisocial behaviour, poor academic achievement and unsettled family background.

Management of both must involve close collaboration between the parents and the teachers. In most cases of school refusal the child should be returned to school as quickly as possible. Truancy is harder to tackle, and the education welfare officer should become involved if the truancy is persistent.

School failure

Reasons for school failure include the following.

Educational

- Limited intellect.
- ADD.
- Hearing or visual deficit.
- Dyslexia.
- Dyspraxia.

Social

- Problems at home.
- Peer problems.
- Absence from school.

School failure is associated with low self-esteem, behavioural difficulties and psychosomatic disorders, and has profound effects on achievement in adult life and chances of employment. It is important to address causes such as dyslexia, attention deficits and visual or hearing impairments that reduce the child's potential to learn, and can lead to frustration and other negative psychological reactions.

Dyslexia

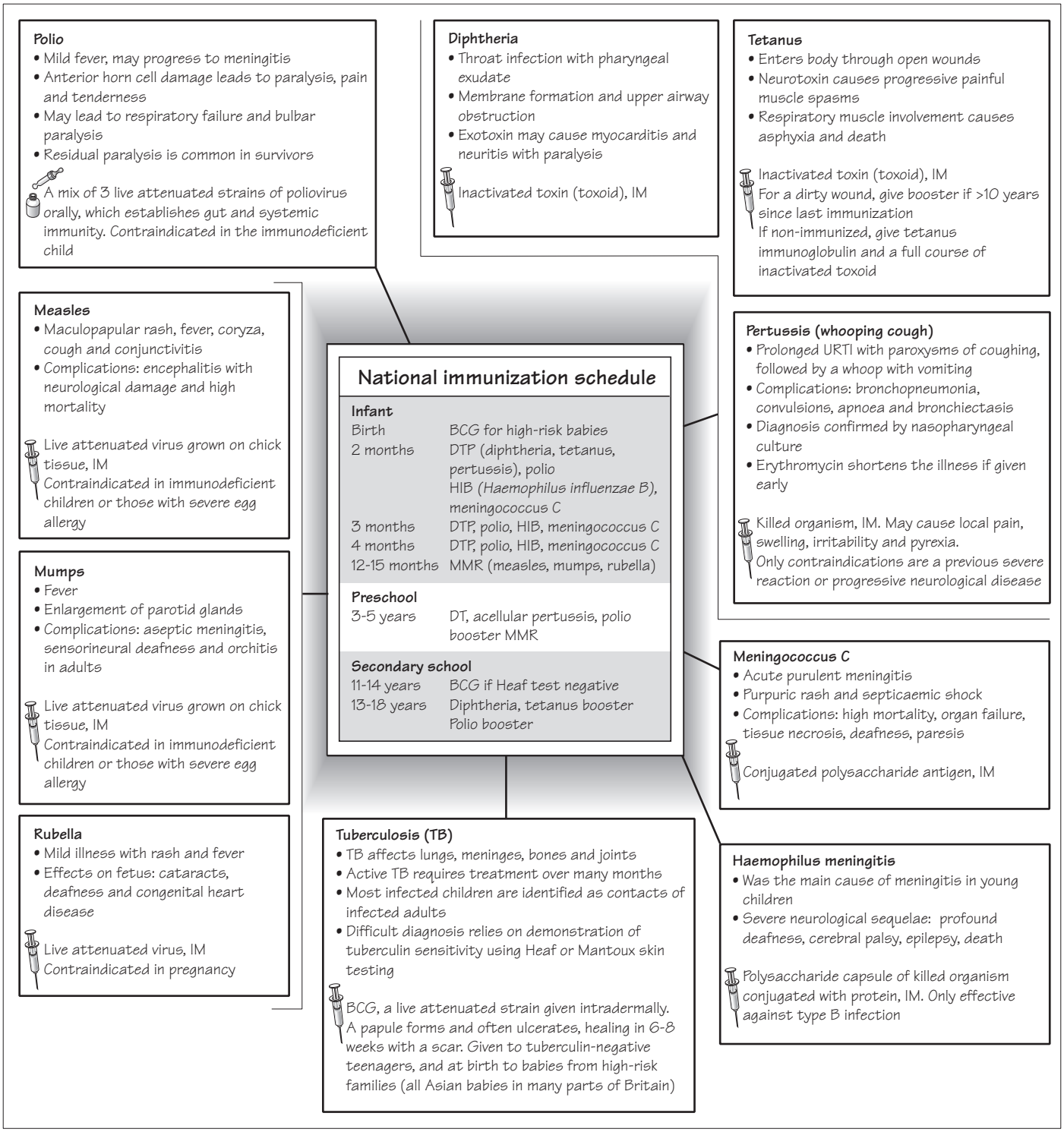
Dyslexia is the commonest type of specific learning difficulty. The dyslexic child is unable to effectively process the information required in order to read. The result is a reading ability below that expected for the child's level of intelligence. Dyslexia must be differentiated from slow reading due to limited intellect or inadequate teaching. It is commoner in boys and there is often a family history.

There may be a history of delay in learning to talk, and spelling is also affected. If unrecognized the child is likely to fail at school, and commonly responds by withdrawing or exhibiting disruptive behaviour. The diagnosis must be confirmed on testing by an educational psychologist, and individual help is required to overcome the difficulties.

Dyspraxia (clumsiness)

Fine motor incoordination leads to untidy writing, gross motor incoordination and difficulty with sports. The academic and social difficulties that ensue can cause considerable unhappiness and behaviour problems if it is not recognized and dealt with helpfully. An occupational therapist can assist in devising a programme to help overcome the difficulties and build self-confidence.

12 Immunization and the diseases they protect against



KEY POINTS

Immunizations should not be given to a child who:

- Is younger than indicated in the schedule.
- Is acutely unwell with fever.
- Has had a serious reaction to a previous dose of the same vaccine.
- Live attenuated vaccines (e.g. polio, measles, mumps, rubella, BCG (bacille Calmette–Guérin)) should not be given to children with immune deficiency states (e.g. cytotoxic therapy or high dose steroids).
- Repeat immunizations should not be given sooner than indicated in the schedule.
- If a child misses an immunization it should be given later. There is no need to restart the course.

13 Screening and surveillance tests

Age	Condition	The test
Neonate	**Congenital hypothyroidism** If treated early with thyroid hormone, the child grows and develops normally. Untreated it results in cretinism with severe learning disability (mental retardation)	Analysis of thyroid (T4) or thyroid stimulating hormone (TSH) from blood on filter paper obtained by heel prick. If positive, urgent referral to a paediatric endocrinologist is required
Neonate after 72 h of age	**Phenylketonuria (PKU)** PKU causes severe learning disability (mental retardation). A low phenylalanine diet prevents the build-up of metabolites which cause brain damage	The Guthrie test is carried out on the same sample as thyroid testing. The baby must be on full milk feeds for 3 days before testing. Referral to a metabolic clinic for dietary advice and long-term follow-up is required
Neonates, 6 weeks	**Congenital cataracts** Early treatment prevents permanent visual impairment	When looking through an ophthalmoscope, if white light instead of red is reflected from the retina, it suggests a cataract or other intraocular pathology. Immediate referral to an ophthalmologist is required
Neonates, 10 days, 6 weeks	**Congenital dislocation of the hips** (p. 105) Early orthopaedic treatment is effective in preventing limp and painful disability from dislocated, subluxed or dysplastic hips. A harness or splint is used initially. If this fails surgery is required	The Ortolani and Barlow procedures are carried out for babies up to the age of 3 months (see p. 105). Limited hip abduction, shortening of the leg and limp (once walking) are sought beyond 3 months. Ultrasound is useful to confirm the diagnosis
Neonates, 6 weeks	**Cryptorchidism** (p. 79) Undescended testes are at risk for infertility and malignancy. Surgery should be performed before the age of 2 years	Examine the baby with warm hands. If on repeat examination the testes are still impalpable, refer to a paediatric surgeon
Weight at routine health visitor contact; length/height at 18 months, 3.5 years and school entry	**Growth** (Chapters 41 and 42) Poor growth can result from organic and inorganic causes	If weight or growth faltering is identified, a paediatric evaluation is required
Test at 6–8 weeks, 7–9 months, 18–24 months, 3–4 years	**Development** (Chapter 46) Delay in one or more areas of development may be indicative of learning difficulties or a neurological problem	All developmental areas are tested and referral to a paediatrician is made if delay or disorder is found
Neonates, 6–8 weeks, preschool	**Congenital heart disease** (Chapter 51) If congenital defects are missed, irreversible cardiopulmonary changes or infective endocarditis may result	Identification of a heart murmur is the commonest presentation
7–9 months	**Hearing impairment** (p. 121) If hearing deficits are not identified before language is acquired, permanent language impairment can result	The distraction hearing test is still commonly used. Neonatal screening by otoacoustic emission is being introduced
School entry, 8, 11 and 14 years	**Vision and hearing at school** Myopia is very common. Hearing impairment identified at this age is due to secretory otitis media as most sensorineural deafness is detected earlier	The Snellen chart or a letter-matching card for younger children is used. Referral to an optician is required if the child can only read letter size 6/12. Colour vision defects are identified using screening cards at age 11. Sweep audiometry tests the child's ability to hear sounds at a set level across the main speech frequencies. Failure requires an ENT and audiological evaluation

KEY POINTS

Screening is the identification of unrecognized disease or defects by using tests. It sorts out apparently well children who may have a problem from those who do not, but is not intended to be diagnostic. Cost must be considered and balanced against that of treatment if the problem presents later, and of medical care as a whole.

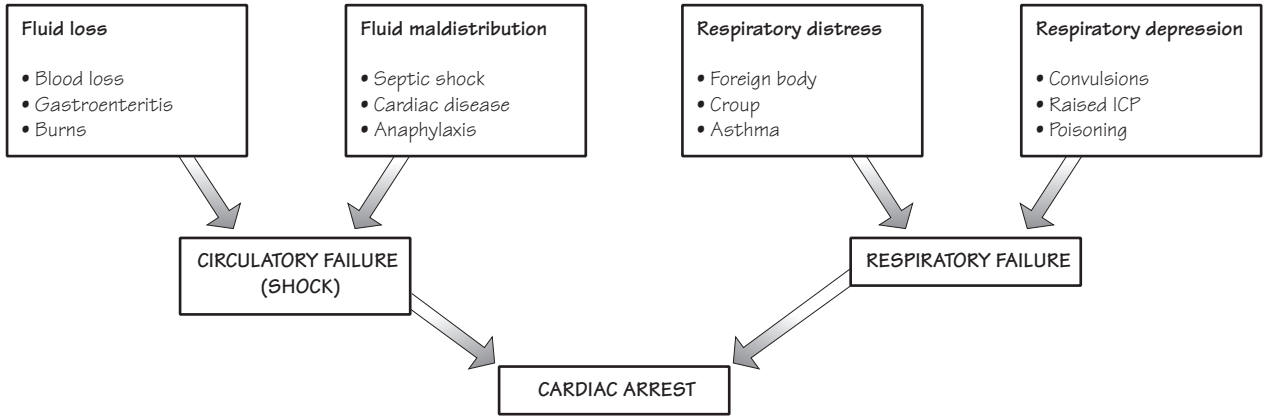
Conditions suitable for screening should:

1. Be identifiable at a latent or early symptomatic stage.
2. Be treatable.
3. Early treatment should influence the course or prognosis.

14 The acutely ill child

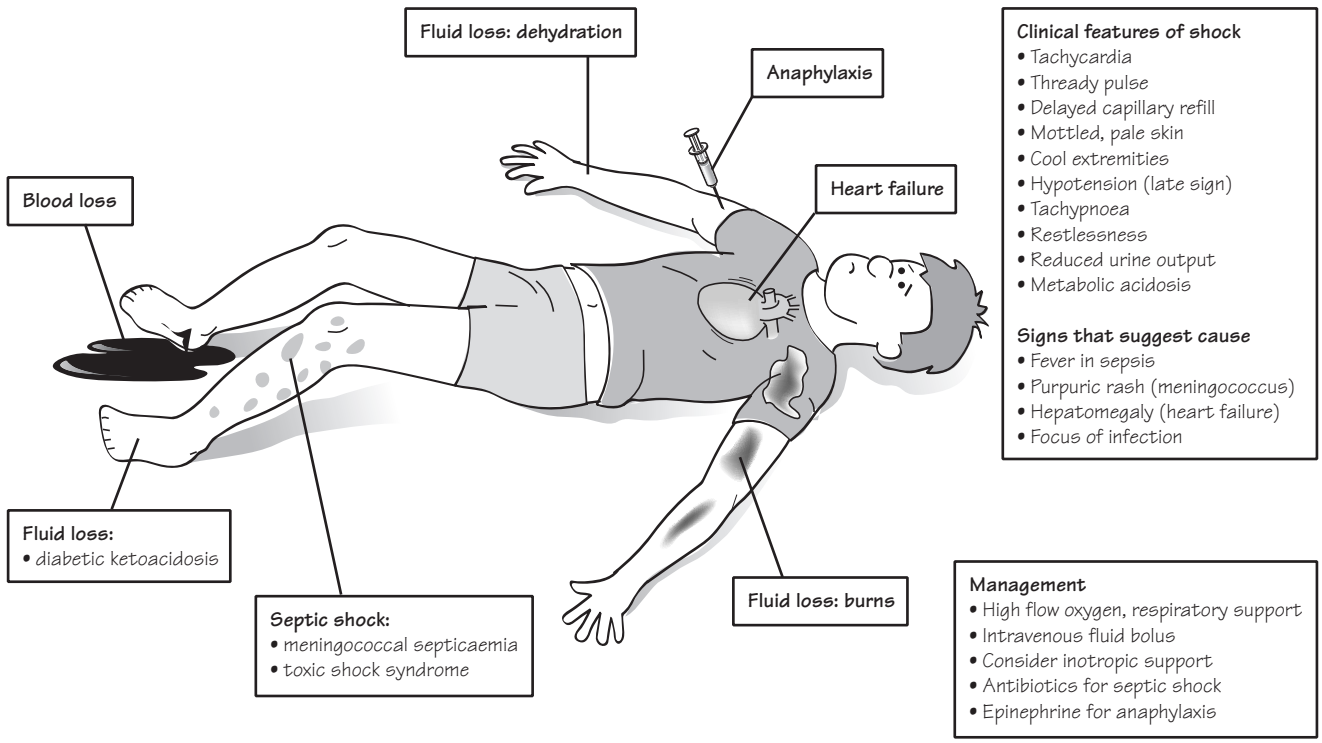
Recognition of acute illness

Children may become critically ill very rapidly, and their survival depends on prompt recognition of the severity of their illness, appropriate life support and rapid treatment. In children the cause of cardiac arrest is almost always due to respiratory or circulatory failure rather than a primary heart problem. It is therefore important to recognize the early signs of respiratory and circulatory failure and to correct these before cardiac arrest occurs.

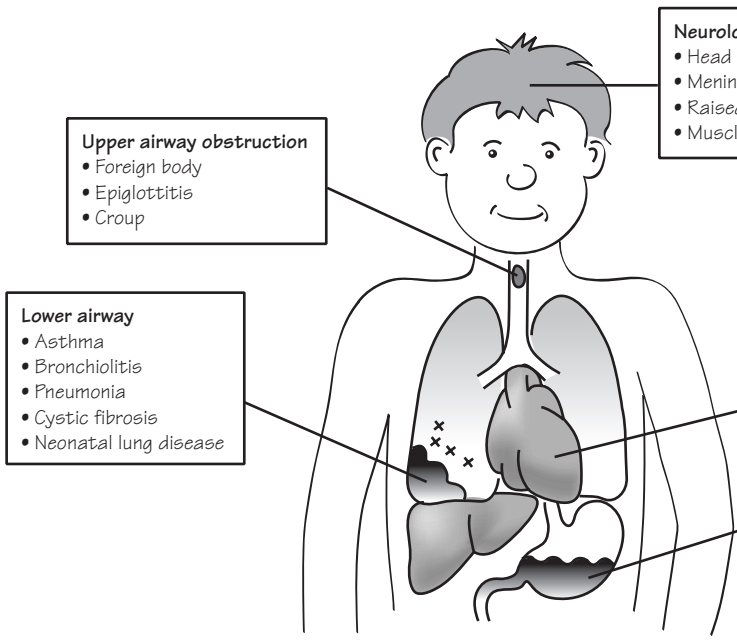


Circulatory failure (shock)

Shock is used to describe a state of inadequate tissue perfusion due to an acute failure of circulation. The body responds by redistributing blood to vital organs such as the brain and the heart, at the expense of the skin, muscles and bowel. Children in shock look pale and have poor skin perfusion. Blood pressure is maintained in children by peripheral vasoconstriction, so that hypotension is a very late sign of shock.



Causes of respiratory failure



Neurological
- Head injury
- Meningitis
- Raised ICP
- Muscle weakness

Upper airway obstruction
- Foreign body
- Epiglottitis
- Croup

Lower airway
- Asthma
- Bronchiolitis
- Pneumonia
- Cystic fibrosis
- Neonatal lung disease

Severe cardiac failure

Drug ingestion

Clinical features
- Shortness of breath
- Tachypnoea
- Cyanosis
- Nasal flaring
- Grunting
- Intercostal recession
- Restlessness or confusion

Symptoms and signs of the underlying disease
- Expiratory wheeze
- Inspiratory stridor
- Crackles
- Neurological weakness

Investigations
- Oxygen saturation
- Arterial blood gases
- Chest X-ray

Management
- Assess severity by examination, blood gases and oxygen saturation monitor
- Give high flow oxygen
- Intubate and ventilate if rising pCO_2. (The decision to ventilate is based on clinical criteria, not just blood gases)
- Treat the underlying cause: antibiotics (infection), bronchodilators and steroids (asthma), remove foreign body

Respiratory failure

Respiratory failure is defined as inadequate respiration to maintain normal arterial oxygen and carbon dioxide concentrations. Respiratory failure is obvious if the child is apnoeic or deeply cyanosed, but it is important to be able to detect impending respiratory failure and to intervene quickly.

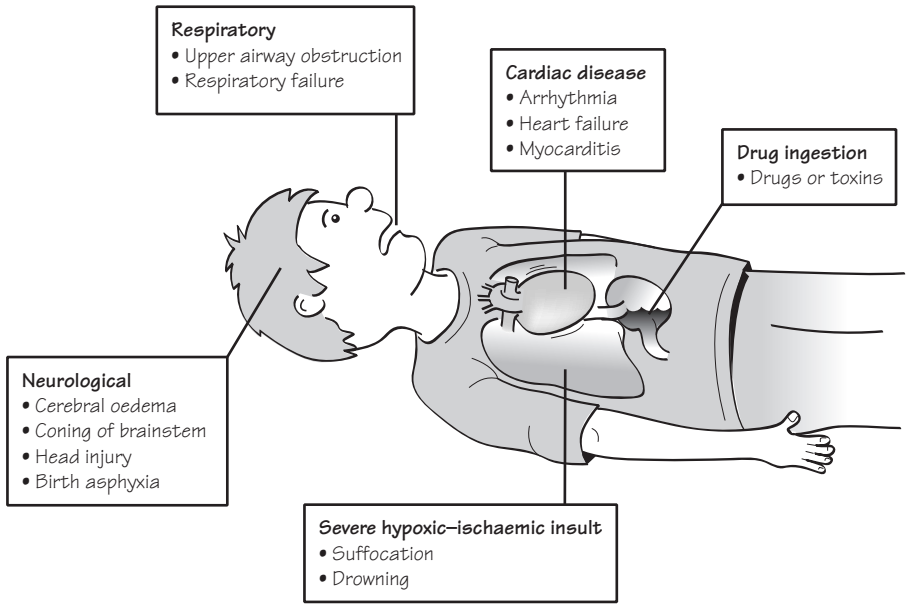
Acute upper airway obstruction

Acute upper airway obstruction is a medical emergency as death may result if it is not effectively treated immediately. It can be due to infection (epiglottitis, croup) or inhalation of a foreign body (especially common in toddlers who put small objects in their mouths). Presentation is with acute sudden onset of choking, coughing and cyanosis, followed by collapse. There may be an inspiratory stridor and marked intercostal recession. If epiglottitis is suspected the doctor must not examine the child's throat and investigations should be delayed until the airway is protected or the obstruction relieved.

Choking should be managed by opening the airway, removing any visible obstruction or if this is impossible performing back-blows or the Heimlich manoeuvre to expel the obstruction from the airway. The Heimlich manoeuvre should not be attempted in infants due to the risk of trauma to the liver and spleen; instead alternate back blows with chest thrusts with the child held in a head-down position. If these measures are unsuccessful artificial ventilation will be required and an emergency tracheostomy or crico-thyroidotomy may be necessary to bypass the obstruction.

The Heimlich manoeuvre

Commoner causes of cardiorespiratory arrest



Management
- Call for help!
- Open the airway
- Check for respiration
- Ventilate using high flow oxygen via bag and mask, or intubation
- Provide mouth-to-mouth if no equipment available
- Check for a central pulse
- Commence external cardiac massage
- Alternate five chest compressions to one breath (15 : 2 in older children)
- Establish intravenous or intraosseus access
- Give appropriate drugs (such as epinephrine)
- Check cardiac rhythm and defibrillate if appropriate
- Always check blood sugar as hypoglycaemia may be an easily treatable cause

Cardiorespiratory arrest

Cardiac arrest is the endpoint of severe respiratory or cardiac failure that has not been adequately treated. The causes are outlined above. Cardiorespiratory arrest outside hospital requires rapid basic life support until skilled help arrives. In hospital cardiorespiratory arrest should be managed by a team of skilled personnel. As most cardiorespiratory arrests in children are secondary to hypoxia rather than due to cardiac disease it is crucial to achieve adequate oxygenation using high flow oxygen and artificial respiration, and to circulate this oxygen by providing appropriate cardiac massage.

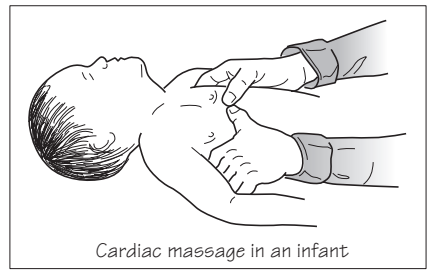
Establishing an airway and artificial ventilation

The airway should be opened by lifting the chin and tilting the head back to a 'sniffing the air' position. In infants the head should be in the neutral position. If there is a possibility of cervical spine injury then the airway should be opened by the 'jaw-thrust' method. The airway should then be cleared by removing any vomit or secretions with suction. Artificial ventilation can be given by mouth to mouth or in infants by mouth to mouth and nose. After five rescue breaths check for a central pulse and commence cardiac massage.

External cardiac massage

In infants cardiac massage can be achieved by encircling the chest with both hands and compressing the sternum with the thumbs, one fingerbreadth below the nipples. In young children the heel of one hand is used one fingerbreadth above the xiphisternum and in older children two hands are used two fingerbreadths above the xiphisternum.

If these measures are not effective then drugs such as epinephrine, bicarbonate and fluid volume may be necessary, depending on the cause of the cardiac arrest. Epinephrine can be given via the intravenous (IV), intraosseous or endotracheal routes. Defibrillation is very rarely required in paediatric cardiac arrests, but is indicated for certain cardiac arrhythmias such as ventricular fibrillation, ventricular tachycardia and supraventricular tachycardia unresponsive to drug therapy.



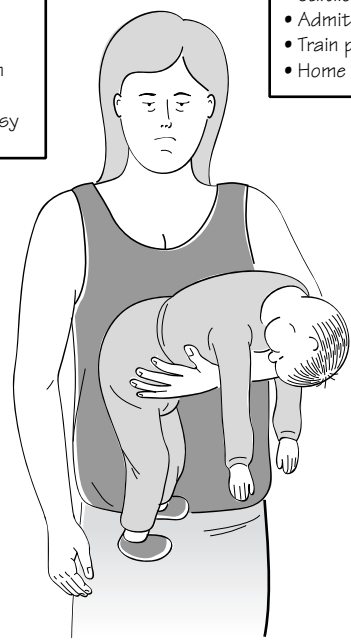
Cardiac massage in an infant

Sudden infant death and acute life-threatening events

Aetiology
- Commonest cause of death in infants >1 week
- Increased risk if prone, overheated or smoky environment
- Normally previously well babies, sometimes with minor cough or cold
- In 20% an unexpected cause is found at autopsy

Examination
- Found collapsed
- Pale and mottled
- Bradycardia
- Hypotension

Investigations
- Blood sugar
- Infection screen
- CXR and barium swallow
- ECG monitoring
- Metabolic screen



Management
- Cardiopulmonary resuscitation (CPR)
- Admit for observation and investigation
- Train parents to administer CPR
- Home apnoea monitor may relieve anxiety but is of no proven benefit

Differential diagnosis
- Infection
- Gastro-oesophageal reflux
- Neurological abnormality
- Hypoglycaemia (rare)
- Cardiac arrhythmia (rare)
- Inborn error of metabolism (rare)
- Suffocation (rare)

'Back to sleep' campaign
- Put babies to sleep on their backs
- Avoid overheating
- Use blankets rather than duvets
- Ideal room temperature is 16–20°C
- Lie the baby at the foot of the cot so that it cannot slip under the covers
- Avoid exposure to cigarette smoke
- Consult a doctor if your baby is ill

Acute life-threatening events and SIDS

SIDS is the 'sudden death of any infant or young child, which is unexpected by history, and in which a thorough postmortem fails to demonstrate an adequate cause for death'. Some children are found in a collapsed state, not breathing and looking grey or mottled, but can be successfully resuscitated. This is referred to as an acute life-threatening event (ALTE) or as 'near-miss cot death'. SIDS is the commonest cause of death in infants after the first week of life, and the peak occurrence is between 2 and 4 months. In the UK the incidence of SIDS has fallen by over 50% due to the 'back to sleep' campaign which advises that babies should be put to sleep on their backs, at the foot of the cot and not overheated or exposed to cigarette smoke.

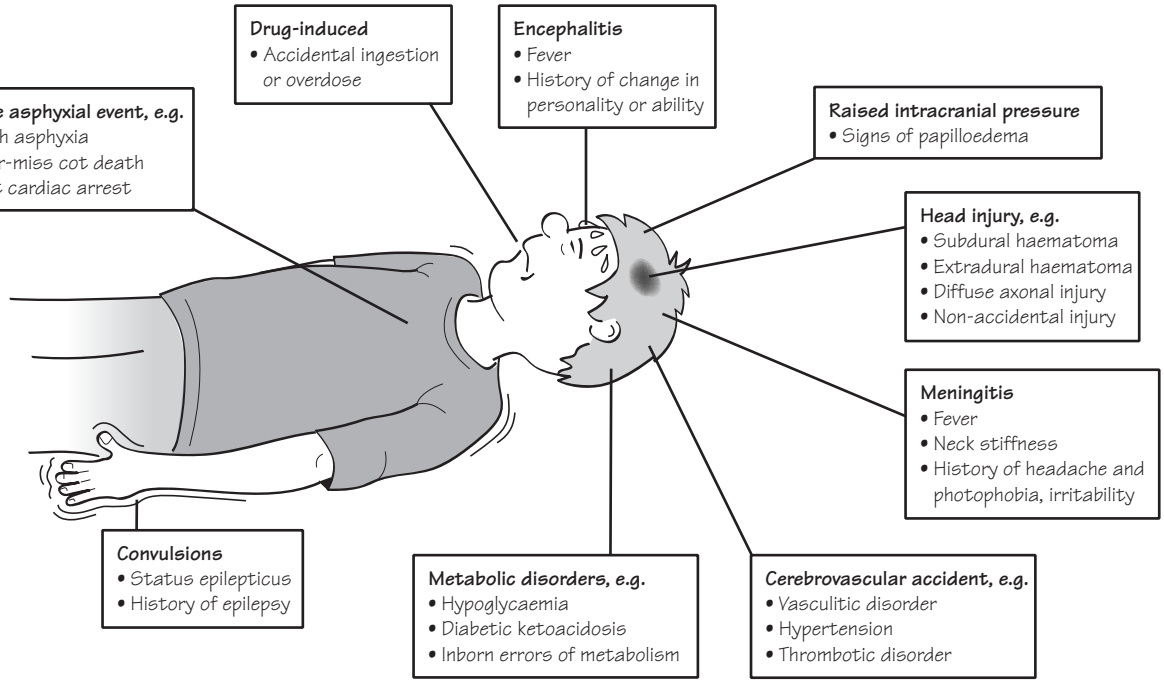
Focal points for the assessment of the acutely ill child

- Call for help immediately.
- Rapidly assess the airway.
- Perform airway opening manoeuvres (but protect the cervical spine if there is any history of trauma).
- Give high-flow oxygen. If the child is not breathing, ventilate with a bag and mask or intubate.
- Check for pulse and assess circulation by capillary refill time.
- Commence chest compressions if a pulse is absent or less than 60 beats/min.
- Establish IV or intraosseus access and give drugs and fluids as appropriate.
- Apply pressure to any active bleeding points.
- Rapidly assess the neurological state by looking at pupils, posture and the level of consciousness.
- Always check blood sugar level.
- Perform appropriate investigations and commence definitive treatment (e.g. infection screen and broad spectrum antibiotics if sepsis is suspected).
- Once the child has been stabilized, transfer to an intensive care unit for definitive care.

KEY POINTS
- Cardiac arrest is usually secondary to respiratory failure or shock.
- Providing adequate oxygenation is critical.
- Shock causes tachycardia and delayed capillary refill time and is treated with boluses of fluids.
- Upper airway obstruction is a common cause of acute respiratory failure in young children.
- The 'back to sleep' campaign has reduced SIDS by 50%.

15 The unconscious child

Causes of coma



What you need from your evaluation

History

- Ask about the possibility of drug ingestion (either deliberate or accidental in young children)
- Was there a prodromal illness or contact with serious infection (e.g. meningitis)
- Assess the possibility of non-accidental injury (p. 99)
- Is there a history of convulsions and for how long did they last?
- Was the child neuro-developmentally normal prior to the onset of coma?

Examination

- **Vital signs:** is there a bradycardia (suggests raised ICP) or tachyarrhythmia (drug ingestion). Deep, sighing respiration (Kussmaul) suggests diabetic ketoacidosis. Ketones may be smelt on the breath
- Look for a focus of infection. Check for rashes and neck stiffness, pneumonia and UTI
- Check pupils: are they symmetrical and do they constrict appropriately to light?
- Check for abnormal posture (decorticate or decerebrate posture)
- Assess the level of consciousness using either the modified Glasgow coma scale or the more rapid AVPU (see opposite)
- Always check the blood glucose. Hypoglycaemia is the commonest metabolic cause of coma

Investigations and their significance

● Blood glucose	Hypo- or hyperglycaemia
● Full blood count	May indicate infection or acute blood loss (Hb and PCV low)
● Blood culture	May identify infective cause
● U&E	High urea in dehydration. Sodium may be high or low
● Blood gases	Metabolic or respiratory acidosis (see Chapter 3 for interpretation)
● Chest X-ray	Infection or cardiac failure, trauma (e.g. rib fracture)
● CT or MRI scan	Focal pathology (tumour, haemorrhage, abscess)
● Lumbar puncture	May show evidence of infection (meningitis, encephalitis) or bleeding (e.g. subarachnoid haemorrhage)

LP should not be attempted in the unconscious child until raised ICP has been excluded, due to the risk of brain herniation (coning)

AVPU coma scale

Alert
responds to **V**oice
responds to **P**ain
Unresponsive

A score of 'P' corresponds to a Glasgow Coma Scale (GCS) of 8, and suggests the airway should be protected by intubation to prevent aspiration

Coma

A child who is deeply unconscious is said to be in coma. Encephalopathy refers to the precomatose state with an altered conscious level. An unconscious child requires urgent and careful evaluation to establish the cause of the coma and to commence appropriate therapy. Whatever the cause of the coma the airway must be protected and adequate ventilation maintained.

Meningitis

Meningitis is caused by either bacterial or viral infection invading the membranes overlying the brain and spinal cord and should be considered in any irritable child with unexplained fever. It is commonest in the neonatal period but can occur at any age. The causes are listed below.

Viral meningitis is preceded by pharyngitis or gastrointestinal (GI) upset. The child then develops fever, headache and neck stiffness. In bacterial meningitis the child is drowsy and may be vacant. Irritability is a common feature, often with a high pitched cry and convulsions may occur. Examination shows an ill child, with a stiff neck and Kernig's sign (pain on extending the legs) may be positive, though this is not reliable in young infants. Tonsillitis and otitis media can also mimic neck stiffness. In infants the fontanelle may be bulging. A petechial or purpuric rash suggests meningococcal meningitis.

Meningitis is confirmed by a lumbar puncture (see p. 20), which will show a leucocytosis, high protein count, low glucose and may show organisms present. The fluid will look cloudy to the naked eye. Culture or PCR analysis will confirm the organism, but treatment should be commenced empirically as soon as the cultures have been taken.

Intravenous cefotaxime or benzyl penicillin is usually given, depending on the age of the child and the likely organism. Steroids may reduce meningeal inflammation in *Haemophilus* meningitis. Meninigococcal menininigitis is associated with pharyngeal carriage and household contacts should receive prophylaxis with rifampicin. Meningococcal septicaemia is discussed in Chapter 37.

Encephalitis

Viral infection sometimes spreads beyond the meninges to infect the brain tissue itself. This is known as meningo-encephalitis. The onset is often more insidious and the child's personality may change and they may become confused or clumsy before the onset of coma. Meningism is less of a feature. The lumbar puncture may reveal a lymphocytosis and specimens should be sent for viral culture and PCR analysis. Herpes simplex virus or *Mycoplasma pneumoniae* may be responsible, and you should always ask about contact with herpetic lesions (coldsores). Treatment with acyclovir, erythromycin and cefotaxime is given until the organism is known.

In herpes encephalitis the electroencephalogram (EEG) and a magnetic resonance imaging (MRI) brain scan may show temporal lobe involvement.

Metabolic causes of coma

In the absence of trauma or infection, a metabolic cause for coma should be considered. By far the commonest metabolic cause is hypoglycaemia, and a blood sugar must be checked immediately at the bedside in every unconscious child. Hypoglycaemia may be due to inadequate carbohydrate intake or excess insulin in children with diabetes mellitus, but it can also be the presenting feature in infants with inborn errors of metabolism or adrenal insufficiency. Hyperglycaemia in uncontrolled diabetes can lead to keto-acidosis with coma, though the onset is often more gradual. Diabetes is discussed in detail in Chapter 53.

Any severe metabolic derangement can cause coma, including severe uraemia (in renal failure) or high ammonia (inborn errors of metabolism such as urea cycle disorders), severe hypernatraemia or hyponatraemia. Coma can also be caused by cerebral oedema from over-rapid correction of electrolyte imbalance in severe dehydration.

Unexplained coma

In unexplained coma the possibility of non-accidental injury such as shaking injury must be considered. A computerized tomography (CT) brain scan and skeletal survey may show evidence of trauma and retinal haemorrhages may be present. Accidental drug ingestion or overdose, or deliberate poisoning may cause coma, and a urine toxicology screen can sometimes identify the drug. Drugs affecting the CNS such as opiate analgaesics, alcohol and antidepressants are often implicated.

Causes of meningitis.

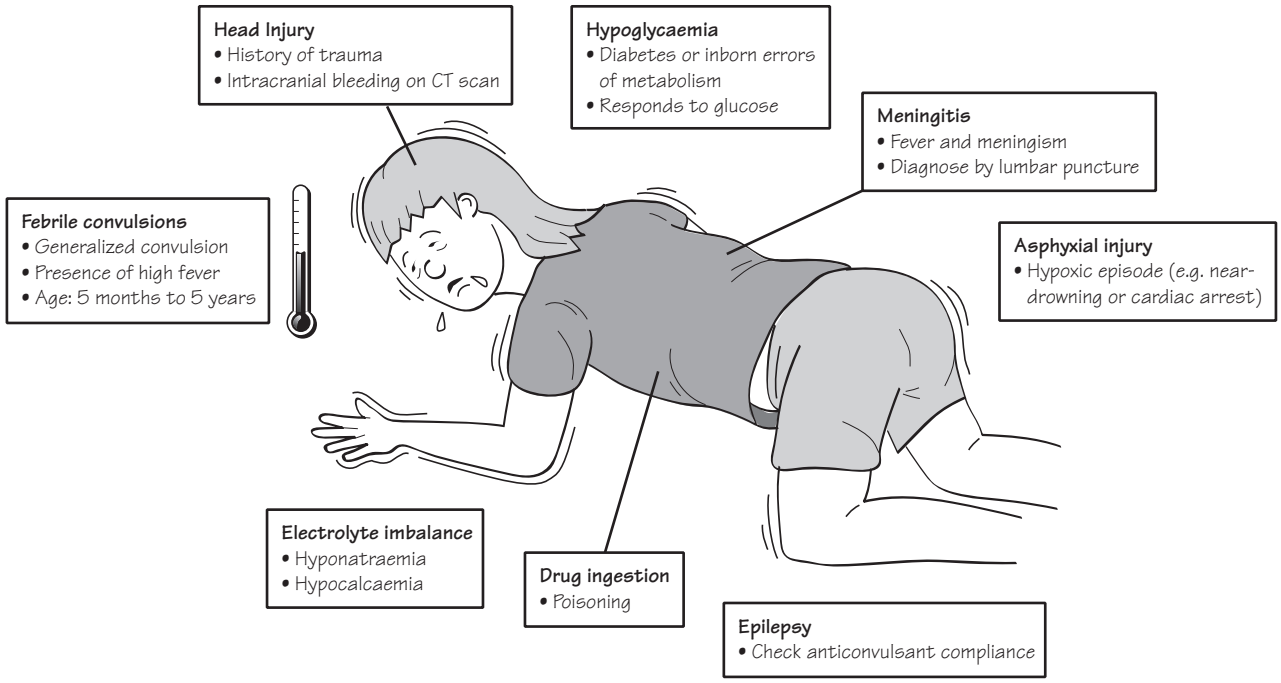
Viral	Mumps virus
	Coxsackie virus
	Echovirus
	Herpes simplex virus
	Poliomyelitis (if unvaccinated)
Bacterial	*Neisseria meningitidis*
	Haemophilus influenzae type B
	Streptococcus pneumoniae
	Group B *Streptococcus* (in newborn)
	Escherichia coli and *Listeria* (in newborn)

KEY POINTS

- Coma can be evaluated rapidly using the AVPU (alert, voice, pain, unresponsive) score.
- Always check the blood glucose in any unconscious child.
- Consider the possibility of poisoning or drug overdose.
- Altered consciousness, fever and irritability suggest meningitis, even in the absence of neck stiffness.
- Never perform a lumbar puncture in an unconscious child until raised intracranial pressure has been excluded.

16 The fitting child

Causes of convulsions



What you need from your evaluation

History

- Is there a history of previous convulsions? The child may have established epilepsy
- How long has the convulsion lasted? Seizures lasting less than 20 min are unlikely to cause brain damage
- Obtain an accurate description of the convulsion-how did it begin, was it focal or generalized? Speak to witnesses. Some parents may have video footage
- Was the child unwell or pyrexial beforehand? Could it be a febrile convulsion or part of a CNS infection?
- Is the child developmentally normal? Non-febrile convulsions are much commoner in children with learning disability or cerebral palsy
- Is drug ingestion or poisoning possible? There may be an organic treatable cause for the fits

Examination

- Make sure the airway is open
- Is the convulsion generalized, affecting all limbs?
- Check the temperature
- Is there an obvious focus of infection?
- Are there signs of trauma or head injury?
- Examine the eyes—are they flickering or rolling?
- Look for signs of meningitis and check the pupils

Treatment

- Give oxygen and maintain a patent airway
- Place the child in the recovery position
- Give rectal diazepam
- Correct any metabolic disturbance
- Give dextrose if hypoglycaemia likely
- Consider intravenous anticonvulsants—lorazepam, phenytoin, paraldehyde
- If in prolonged status epilepticus, thiopentone infusion and ventilation may be needed

Investigations and their significance

● Blood glucose	Must always be checked in any fitting child. Can be done at the bedside
● U&E	Hyponatraemia, hypocalcaemia and hypomagnesaemia can cause fits
● Lumbar puncture	If meningitis suspected, but beware raised ICP in prolonged fit
● CT/MRI scan	If any history of trauma or focal neurological signs suggesting intracranial lesion
● Blood and urine cultures, throat swab, CXR	To look for focus of infection in febrile convulsions
● Urine toxicology	If drug ingestion or overdose suspected

Generalized convulsions

The term convulsion is synonymous with fit or seizure. Convulsions are due to synchronous discharge of electrical activity from a number of neurones, and this manifests itself as loss of consciousness and abnormal movements. In a generalized convulsion all four limbs and the face are affected. Convulsions are common. They occur in 3–5% of children. They do not necessarily mean the child will go on to develop epilepsy, and many children only ever have one convulsion. However, 60% of epilepsy develops in childhood. Children's brains are particularly susceptible to convulsions and the commonest trigger is the rise in temperature during a febrile illness. These are known as febrile convulsions and are described below. Epilepsy is discussed in Chapter 60.

Febrile convulsions

These usually occur between the ages of 5 months and 5 years in neurologically normal children and are triggered by fever, usually as part of a viral upper respiratory tract infection, although they may also be triggered by any febrile illness including acute otitis media, urinary tract infection (UTI) or tonsillitis. The convulsion is usually short lived, lasting only a few minutes. Status epilepticus occurs in less than 1% of febrile convulsions. Febrile convulsions should be managed by identifying and treating the source of the fever, and cooling the child by undressing them and sponging their skin with tepid water. Antipyretics such as paracetamol or ibuprofen should be given. If the convulsion persists for more than 10 min it should be terminated with rectal diazepam. Investigations should be performed to exclude serious infection, and if no obvious focus of infection is found a lumbar puncture is indicated to exclude meningitis.

Advice to parents is very important. About one-third of children will have further febrile convulsions in the future. Parents must be taught how to manage any future febrile illnesses and taught basic first aid management of convulsions. The prognosis, which is good, should be explained. Children with uncomplicated febrile convulsions are at no greater risk of epilepsy, although if the convulsion lasts more than 20 min they may be at increased risk of temporal lobe epilepsy. Overall the risk of epilepsy is 2–3%. Regular anticonvulsant medication is only very rarely prescribed, if the child is having very frequent or prolonged febrile convulsions.

Management of the fitting child

Most parents who witness their child fitting imagine that the child is going to die and seek medical attention urgently. Children may still be fitting when they present to the GP or to hospital. The most important thing is to support the airway and turn the child into the recovery position, semi-prone with the knee flexed under the chest (see box opposite). Objects should not be put into the mouth (except an artificial airway). If oxygen is available this should be given by facemask. If the convulsion is ongoing, rectal diazepam should be given as an anticonvulsant to terminate the seizure. Occasionally some convulsions persist; this is know as status epilepticus and is described below.

It is vital to check the blood glucose immediately in any fitting child, as hypoglycaemia is a common and rapidly treatable cause. Not all children with hypoglycaemic convulsions are diabetic; some may have inborn errors of metabolism and may not tolerate even short periods of starvation. Once the convulsion has terminated the child may remain drowsy or 'postictal' for some time. They should be observed carefully and kept in the recovery position until they are able to maintain their own airway. If it is the child's first convulsion the parents will require much reassurance and will need to be taught how to manage future episodes. Occasionally this will include prescribing rectal diazepam to be administered at home.

Status epilepticus

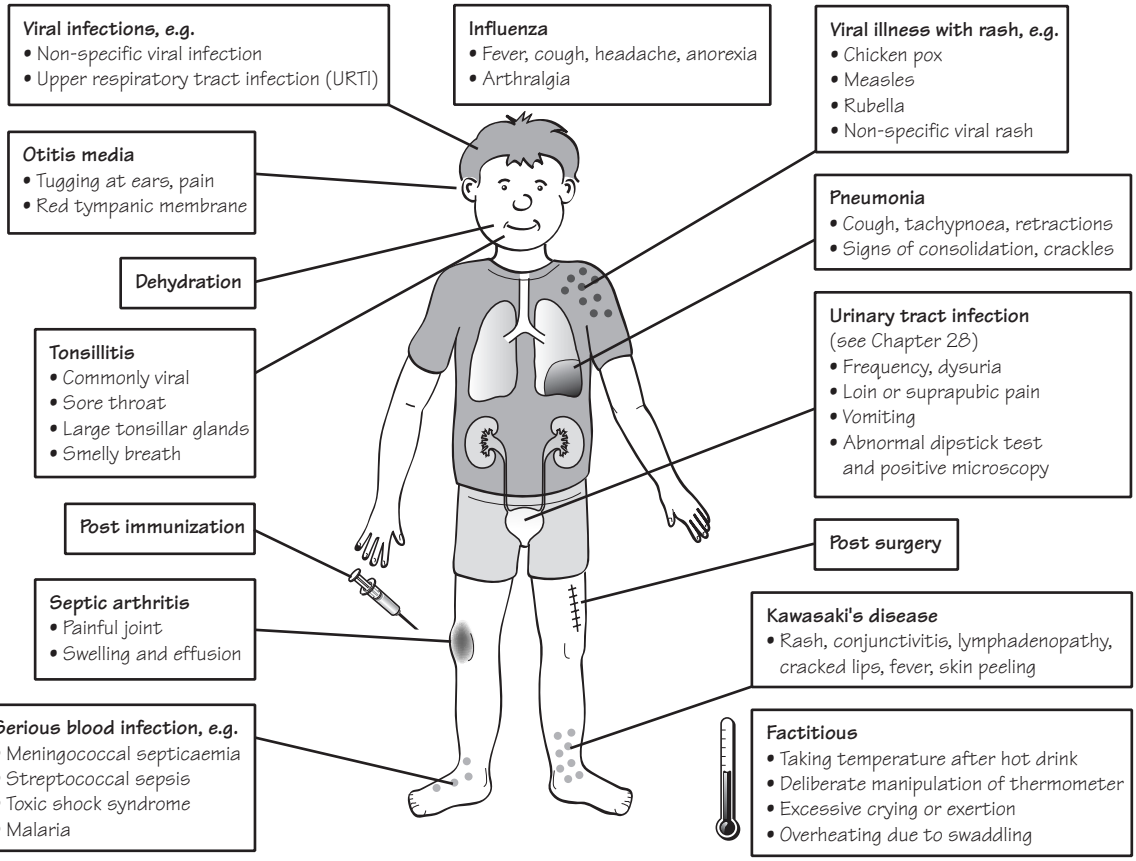
Seizures may be very prolonged and are an important cause of coma. Status epilepticus is defined as continuous seizure activity for more than 30 min or a series of seizures without full recovery in between. Status may occur following febrile convulsions or more commonly in children with known epilepsy, or with other acute causes such as trauma or metabolic disturbance. The child's airway should be maintained, oxygen given and blood glucose checked. If rectal diazepam has been given, IV lorazepam should also be given. Rectal paraldehyde or IV phenytoin may be tried. If all these have failed an anaesthetist should be called and the child should undergo rapid sequence induction of anaesthesia with thiopental. Any child with very prolonged seizures should be monitored carefully on an intensive care unit, and urgent investigations performed to identify the cause.

KEY POINTS

- Febrile convulsions occur in 3% of children between age 5 months and 5 years. The prognosis is usually excellent.
- Children who are fitting must be placed in the recovery position and their airway maintained.
- Always check blood sugar as hypoglycaemia is a common and treatable cause.
- Any convulsion lasting more than 10 min should be terminated with rectal diazepam.
- Status epilepticus is fitting for more than 30 min and requires urgent treatment.

17 The febrile child

Causes of fever



What you need from your evaluation

History

- The parents have normally noticed the fever and may have checked the child's temperature
- Ask about the duration and pattern of the fever—does it occur at particular times of the day?
- Is there pain? Earache, difficulty swallowing, dysuria or frequency may point to the source
- Are there associated features such as malaise, anorexia, vomiting, coryza, cough or rash?
- Has there been contact with other children with infection such as meningitis or chicken pox?
- Has the child just been vaccinated?
- Is the child still drinking adequate amounts of fluid?
- What anti-pyretics and cooling measures have been tried?

Investigations and their significance

• Full blood count	Leucocytosis with neutrophilia suggests bacterial infection
• Throat swab	*Streptococcus* requires treatment with penicillin
• Blood culture	If positive, suggests septicaemia. Treatment may have to commence before result known
• Lumbar puncture	To exclude meningitis and encephalitis. Should be performed in any seriously ill child when no focus of infection can be found, especially in infants <1 year
• Urine analysis	Pure growth of a single organism with leucocytosis confirms infection. Protein and red cells may be present. Dipsticks can be used to test for leucocytes, protein and nitrites
• Chest X-ray	May reveal cause of fever in infants as chest signs may not always be apparent

Examination

- Check the temperature: oral, axilla or rectal
- Does the child look seriously ill? Is there a rash, tachypnoea, tachycardia or dehydration?
- **Chest:** are there signs of respiratory infection—tachypnoea, recession, crackles or grunting?
- **Throat:** feel for cervical lymphadenopathy and look at tonsils. Is there an exudate?
- **Ears:** are the tympanic membranes red or bulging?
- **CNS:** is the child orientated? Is there floppiness or signs of meningism?
- **Urine:** check the urine with dipstick or microscopy

Management of fever in children

Temperature can be measured rectally, orally, or in the axilla using a thermometer. Thermal devices can also give an estimation of temperature directly from the skin or from the ear canal. Fever is defined as an axillary temperature above 37°C. The height of the fever doesn't necessarily correlate with the severity of the illness and fever can commonly occur in children with minor illnesses. Fever is usually a response to infection, and the child often appears flushed as blood vessels in the skin vasodilate in an attempt to lose heat. Fever is an unpleasant symptom and should be treated. Some young children are at increased risk of febrile convulsions if their temperature rises very rapidly (see p. 47).

Fever can be treated by undressing the child and allowing them to lose heat through the skin. Sponging the skin with tepid water can also bring down the temperature by evaporation. The mainstay of treatment is antipyretics such as paracetamol (Calpol) or occasionally ibuprofen. Paracetamol can be used regularly in the correct dosage to keep the child's temperature down. Aspirin should not be used in children under 12 years as it is associated with the development of severe liver failure (Reye's syndrome).

Viral upper respiratory tract infections

Upper respiratory tract infections (URTIs) are extremely common in children, occurring 6–8 times a year on average. They are especially common when toddlers start at nursery or playgroup and when children start school. At these times they are exposed to a large number of viral infections to which they have no immunity. The child often has coryza (runny nose) or acute pharyngitis associated with fever. Young infants may have difficulty breathing and feeding because they are obligate nose breathers. The tympanic membranes are often inflamed. In acute pharyngitis the tonsillar fauces and palate are inflamed and cervical lymph nodes may be enlarged. Treatment is symptomatic, with antipyretics such as paracetamol. Saline drops may improve nasal congestion in infants. The infection usually lasts 3–4 days. Antibiotics are not indicated!

Tonsillitis

Tonsillitis is usually viral in origin. In older children the commonest bacterial organism is the group A beta-haemolytic streptococcus. The child may complain of a sore throat or dysphagia and they usually have a fever. There is often tender cervical lymphadenopathy which may cause neck stiffness. Associated adenitis in the mesenteric nodes may cause abdominal pain. The tonsils will be enlarged and acutely inflamed. They may have a white exudate in bacterial tonsillitis although this is not always a reliable sign. Exudates can also occur with infectious mononucleosis (glandular fever) and with diphtheria (now very rare). The breath may smell offensive in bacterial tonsillitis. Acute tonsillitis should be distinguished from the hypertrophied but non-inflamed tonsils which are common in preschool children. Most children do not require antibiotics and can be managed with saline gargles and paracetamol. If bacterial infection is suspected this should be confirmed by a throat swab. Streptococcal tonsillitis should be treated with benzyl penicillin for 10 days. Complications of tonsillitis are rare but include otitis media, peritonsillar abscess (quinsy) and poststreptococcal glomerulonephritis. Chronically enlarged tonsils can cause upper airway obstruction and obstructive sleep apnoea. This is an indication for tonsillectomy.

Acute otitis media

This is a very common disorder, especially in young children and can occur in babies. The commonest causes are *Streptococcus pneumoniae*, *Haemophilus influenzae* and viruses. Otitis media is especially common if there is Eustachian tube dysfunction, which can be associated with URTIs, obstruction from enlarged adenoids, cleft palate and Down syndrome. Otitis media presents with fever, deafness and pain in the ear. The child may be irritable and may tug or pull at the affected ear, or infection may be aymptomatic. Examination shows a red, inflamed and bulging tympanic membrane, with loss of the light reflex. Treatment with ampicillin shortens the duration of symptoms. Prognosis is generally good even if the tympanic membrane has perforated.

Complications include secretory otitis media (glue ear), conductive deafness and mastoiditis. In secretory otitis media, recurrent acute infections lead to a thick glue-like exudate building up in the middle ear. On examination the tympanic membrane appears thickened and retracted with an absent light reflex. If there is significant hearing loss, ventilation tubes (grommets) may be inserted through the tympanic membrane to allow the middle ear to drain. These are particularly indicated if there is language delay due to the conductive deafness associated with glue ear.

Fever in newborn infants

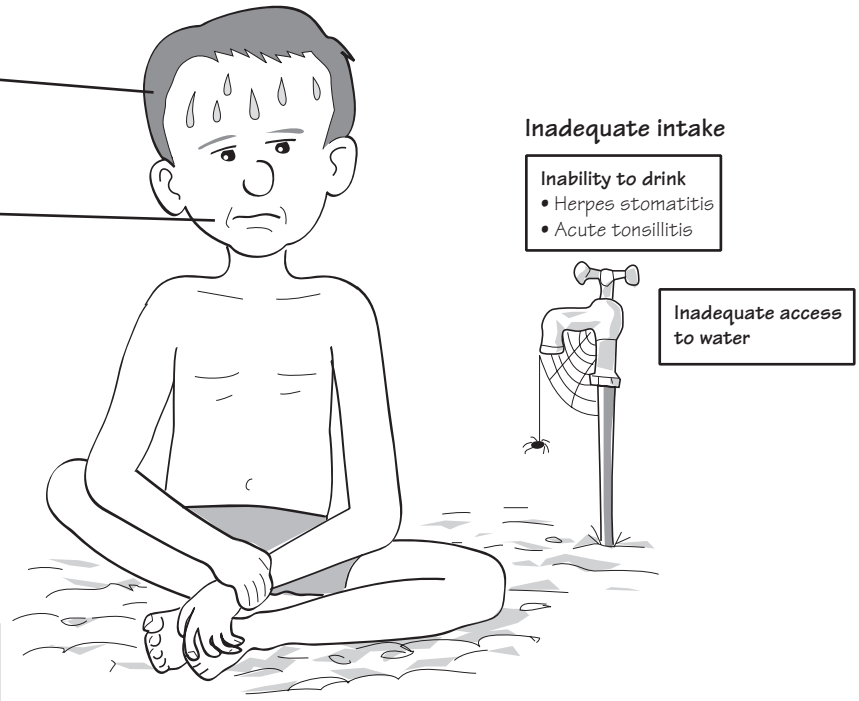
Fever in an infant less than 8 weeks old should always be taken seriously because this may represent late onset congenital infection. Signs of sepsis at this age can be quite non-specific, so a significant fever should always prompt a careful examination and appropriate investigations. If well, the child may just be observed, but if significantly ill will require a full infection screen including urine culture, chest X-ray and possibly lumbar puncture.

KEY POINTS

- Fever is a very common symptom in children, and can usually be managed by simple cooling measures and paracetamol.
- Any ill child with a high fever should be examined carefully to exclude serious infections such as meningitis, UTI or pneumonia.
- Any fever in a baby less than 8 weeks old should be taken seriously.
- Otitis media and tonsillitis are common causes of fever in young children.
- Most fevers are associated with non-specific viral infections or URTIs.

18 Acute diarrhoea and dehydration

Causes of dehydration



What you need from your evaluation

History

- Has there been diarrhoea and/or vomiting?
- Is the vomiting projectile (pyloric stenosis)?
- How many loose stools have there been?
- Is the child passing less urine than normal? Ask when was the last wet nappy?
- How often and for how long has the child been vomiting?
- Does the child have cystic fibrosis or diabetes?

Investigations and their significance

(Investigations are required only in moderate to severe diarrhoea or if the child is very ill)

• U&E	For electrolyte imbalance and renal function
• Blood gas	Metabolic acidosis or alkalosis
• Urinalysis	For osmolality or specific gravity
• Blood sugar	To exclude diabetic ketoacidodis
• Stool culture	In gastroenteritis and food poisoning

Examination

- Weigh the child and compare with previous weight (if known) to assess dehydration
- In young infants feel for a pyloric mass during a test feed (pyloric stenosis)
- Assess the degree of dehydration (mild, moderate or severe) as follows:

	Mild	Moderate	Severe
Mouth and lips	Dry	Dry	Dry
Urine output	Normal	Reduced	None for 12 h
Mental state	Normal	Lethargic	Irritable or coma
Pulse rate	Normal	Tachycardia	Tachycardia
Blood pressure	Normal	Normal	Low
Capillary refill time	Normal	Delayed	Very delayed
Fontanelle	Normal	Sunken	Very sunken
Skin and eye turgor	Normal	Reduced	Very reduced
Dehydration (%)	**<5**	**5–10**	**>10 (shock)**

Treatment

- Use oral rehydration therapy where possible
- Treat shock with boluses of IV fluids
- Rehydrate slowly to replace fluid loss over at least 24 h
- Correct any electrolyte imbalance

Dehydration

Water is the major component of the body, accounting for up to 80% of an infant's body weight. Loss of more than 5% of this water represents significant dehydration. Fluid may be depleted in the intracellular or extracelluar compartments. If a significant amount of fluid is lost acutely from the intravascular part of the extracellular space, then shock may ensue. Normal body fluid is a balance between intake (drinking) and output (urine output, stool volume, sweat and insensible losses such as vapour in expiration). If intake does not keep up with losses, then the child will become dehydrated. The commonest cause of dehydration in children is gastroenteritis due to diarrhoea and vomiting.

Acute diarrhoea

Episodes of acute diarrhoea are not uncommon in children, and are usually due to infection, although not always GI infection. Sadly dehydration due to gastroenteritis is still a major cause of mortality in children in the developing world. Gastroenteritis is usually viral, and rotavirus is the main agent causing winter epidemics. Diarrhoea follows 1–2 days after low-grade fever, vomiting and anorexia. There may be acute abdominal pain and malaise. The diarrhoea resolves within a week and the management is adequate rehydration (see below). Bacterial gastroenteritis has a similar presentation and the commonest pathogens are *Escherichia coli, Shigella*, *Salmonella* and *Campylobacter*. Meningism and febrile convulsions can occur with shigella, whilst bloody diarrhoea occurs in shigella and *Campylobacter* infection. Infection with the 0157 strain of *E. coli* can be followed by haemolytic uraemic syndrome—a life-threatening disease with haemolysis and acute renal failure. Antibiotics should not be prescribed for uncomplicated gastroenteritis. If there is evidence of septicaemia the child should be admitted for IV antibiotics.

Any form of infection or febrile illness can cause diarrhoea, especially in infants. This includes viral URTIs, chest infections, otitis media and UTI. Use of antibiotics may in itself cause diarrhoea due to a disturbance of the normal enteric flora. Recurrence of diarrhoea on refeeding is most likely to be due to lactase deficiency and may require a lactose free diet for a number of weeks.

Management of dehydration

Try to determine the cause of the diarrhoea and the degree of dehydration. Ask about the duration of diarrhoea, whether there has been vomiting and when the child last passed urine.

The degree of dehydration can be assessed by the pulse, blood pressure, mucous membranes, urine output, skin turgor and by feeling the fontanelle (see box opposite). You should be able to decide whether the child has mild (<5%), moderate (5–10%) or severe (>10%) dehydration.

In mild dehydration the only physical sign may be a dry mouth, whilst with severe dehydration the child may be semiconscious or shocked.

The child should be weighed, the difference between the weight at presentation and a recent weight can be used to estimate the volume of body water that has been lost (1 kg approximates to 1 litre). If the child is significantly dehydrated blood should be taken for urea, electrolytes and bicarbonate.

Bicarbonate may be lost in diarrhoea leading to metabolic acidosis, or if there is persistent vomiting (e.g. pyloric stenosis) then loss of H^+ ions may lead to metabolic alkalosis. Sodium may be low in hyponatraemic dehydration or high if more water than sodium has been lost (hypernatraemic dehydration) or if the child has been given over-concentrated formula feeds or excessive salt. In hyponatraemic dehydration (Na^+ <130 mmol/l) the child is lethargic and the skin feels dry and inelastic. In hypernatraemic dehydration (Na^+ >150 mmol/l) the child is very thirsty and the skin may feel doughy.

Mild dehydration (<5%): may be treated at home using oral rehydration therapy, as long as the child is not vomiting excessively. The child should be encouraged to drink a rehydration solution which contains glucose and salt in the correct concentration to aid water absorption and restore electrolyte balance. Breast-feeding may be continued, but if the infant is formula fed, milk can be reintroduced once the diarrhoea has settled.

Moderate and severe dehydration: these children are usually admitted to hospital and may require IV fluid therapy. If shock is present the circulation is restored by boluses of colloid. The volume of fluid necessary to correct the deficit of water and to provide maintenance fluids and cover ongoing losses is then given over 24 h (see box below). The fluid used should be saline or dextrose saline. Too-rapid rehydration can lead to dangerous fluid shifts and hyponatraemia. The electrolytes must be checked frequently and fluids adjusted to normalize the sodium and potassium concentrations. The urine output must be monitored and fluid balance calculated regularly.

CALCULATING THE REPLACEMENT AND MAINTENANCE FLUID REQUIREMENTS

An infant weighing 7.5 kg is thought, on the basis of clinical examination, to be 10% dehydrated.

Fluid deficit (ml) = weight × percentage dehydration × 10 = 750 ml

Maintenance fluids = 100 ml/kg/day for the first 10 kg of body weight
= 100 × 7.5 kg = 750 ml

This child therefore needs 750 + 750 = 1500 ml fluids over the first 24 h to rehydrate and then maintain normal hydration.

Note: maintenance fluids covers essential urine output and insensible losses. If there are significant ongoing losses (e.g. diarrhoea) this volume may need to be increased further. The best initial fluid is usually 0.45% saline with 5% dextrose. The electrolyte content can be adjusted once serum electrolytes are known.

KEY POINTS

- Gastroenteritis is the commonest cause of dehydration. In developing countries it is a major cause of infant mortality.
- It is important to accurately estimate the degree of dehydration by clinical evaluation.
- Wherever possible try to rehydrate the child with oral rehydration therapy. Breast feeds should be continued.
- IV treatment of significant dehydration requires accurate calculation of required fluid volumes and careful correction of electrolyte imbalance.

19 Vomiting

Causes of vomiting

Newborn and infants

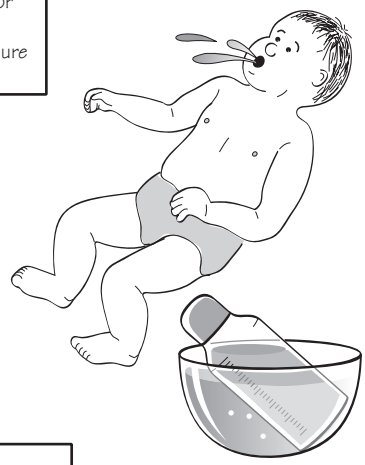
Overfeeding
- Feeding >200 ml/kg/day

Gastro-oesophageal reflux
- Due to lax gastro-oesophageal sphincter: positional vomiting
- May lead to oesophagitis or aspiration pneumonia
- May cause apnoea and failure to thrive

Pyloric stenosis
- 4–6 weeks old
- Projectile vomiting after feed
- Hungry after vomiting
- Constipated
- Palpable pyloric mass

Whooping cough
- Paroxysmal cough

Small bowel obstruction
(congenital atresia or malrotation)
- Bile-stained vomiting
- Presents soon after birth
- May have abdominal distension



Constipation

Older children and adolescents

Gastroenteritis
- Usually with diarrhoea
- History of contact with infection
- Check for dehydration
- Usually self-limiting

Systemic infection
- Meningitis
- UTI (pyelonephritis)

Migraine
- Characteristic headache

Raised intracranial pressure
- Effortless vomiting
- Usually neurological signs
- Papilloedema

Bulimia

Toxic ingestion or medications

Pregnancy

What you need from your evaluation

History

- In infants it is important to differentiate posseting from serious vomiting. With significant vomiting the child will look ill and be failing to gain weight or may even be losing weight
- Take a thorough feeding history, as overfeeding is not uncommon in a thriving baby who seems hungry but vomits the excess milk after a feed
- Always ask about projectile vomiting (pyloric stenosis) and bile-stained vomiting. The latter suggests intestinal obstruction and must be investigated urgently
- The presence of diarrhoea suggests gastroenteritis
- Fever suggests infection, and it is important to look for infection outside the gastrointestinal system; UTI, otitis media and meningitis may all present with vomiting. Vomiting with infection tends not to be projectile
- Paroxysms of coughing followed by turning red or blue and vomiting suggests whooping cough
- Gastro-oesophageal reflux should be suspected in infants and children with disability such as Down syndrome or cerebral palsy

Examination

- Check for dehydration, especially with gastroenteritis
- Feel for a palpable pyloric mass in any young infant
- Check for abdominal distension, which suggests intestinal obstruction
- Check for papilloedema and hypertension in cases of unexplained vomiting to exclude raised ICP as a cause
- Look for signs of meningitis

Investigations and their significance

Investigations are required only in particular cases.

• Plasma urea and electrolytes	To assess electrolyte imbalance in dehydration and in pyloric stenosis
• Plasma chloride, pH and bicarbonate	To assess degree of metabolic alkalosis in pyloric stenosis
• pH monitoring and barium swallow	May show *significant* gastro-oesophageal reflux
• Upper gastrointestinal contrast study	Mandatory in bile-stained vomiting in newborn to exclude malrotation

Regurgitating a small amount of milk, called posseting, is normal in babies. Vomiting refers to more complete emptying of the stomach. Vomiting is one of the commonest symptoms in childhood, and is often due to gastroenteritis. It may be associated with more serious infections such as pyelonephritis, or may be the presenting symptom of life-threatening conditions such as meningitis or pyloric stenosis. In newborn infants bile-stained vomiting suggests a congenital obstruction to the GI system.

Gastro-oesophageal reflux

Gastro-oesophageal reflux (GOR) is a common symptom in babies and in children with cerebral palsy or Down syndrome. It is due to a weak gastro-oesophageal sphincter which allows stomach contents to reflux into the oesophagus. It may present with trivial posseting or significant aspiration, oesophagitis or apnoea. Vomiting is worse after feeds and on lying down, and may cause failure to thrive. Abnormal posturing may occur with acid reflux. GOR is usually diagnosed on the basis of a typical history. Investigations should only be performed if the reflux is significant. These include a barium swallow and monitoring the oesophageal pH for 24 h using a pH probe. The presence of acid in the oesophagus usually represents reflux of stomach acid and the percentage of time that this occurs can be calculated over 24 h. Oesophagoscopy is used to confirm oesophagitis. Simple reflux can be managed by thickening the feeds and nursing the infant in a more upright position. In very severe reflux drugs that affect gastric emptying and gut motiltiy can be used and a small number of children with recurrent aspiration require surgical fundoplication. Most gastro-oesophogeal reflux resolves over time as the infant is weaned onto a more solid diet.

Pyloric stenosis

Pyloric stenosis is caused by hypertrophy of the pylorus muscle. It usually develops in the first 4–6 weeks of life and is said to be most common in first-born male infants. The vomiting is characteristically projectile, occurring immediately after a feed. The vomitus is not bile-stained and the infant is usually hungry. Examination may show weight loss and dehydration and the infant may be irritable due to hunger. Careful palpation after a test feed may reveal a hard mobile mass in the epigastric area. If there is doubt ultrasound examination may show a thickened and elongated pyloric muscle. Blood tests may show a low serum chloride, potassium and sodium, and an alkalosis secondary to protracted vomiting of stomach acid. The infant should be fully rehydrated with careful correction of the electrolyte imbalance before definitive surgery is performed. Rehydration may take at least 24 h. Surgery involves splitting the pylorus muscle without cutting through the mucosa (Ramstedt's pyloromyotomy). Oral feeds can usually be commenced soon after surgery.

Bowel obstruction

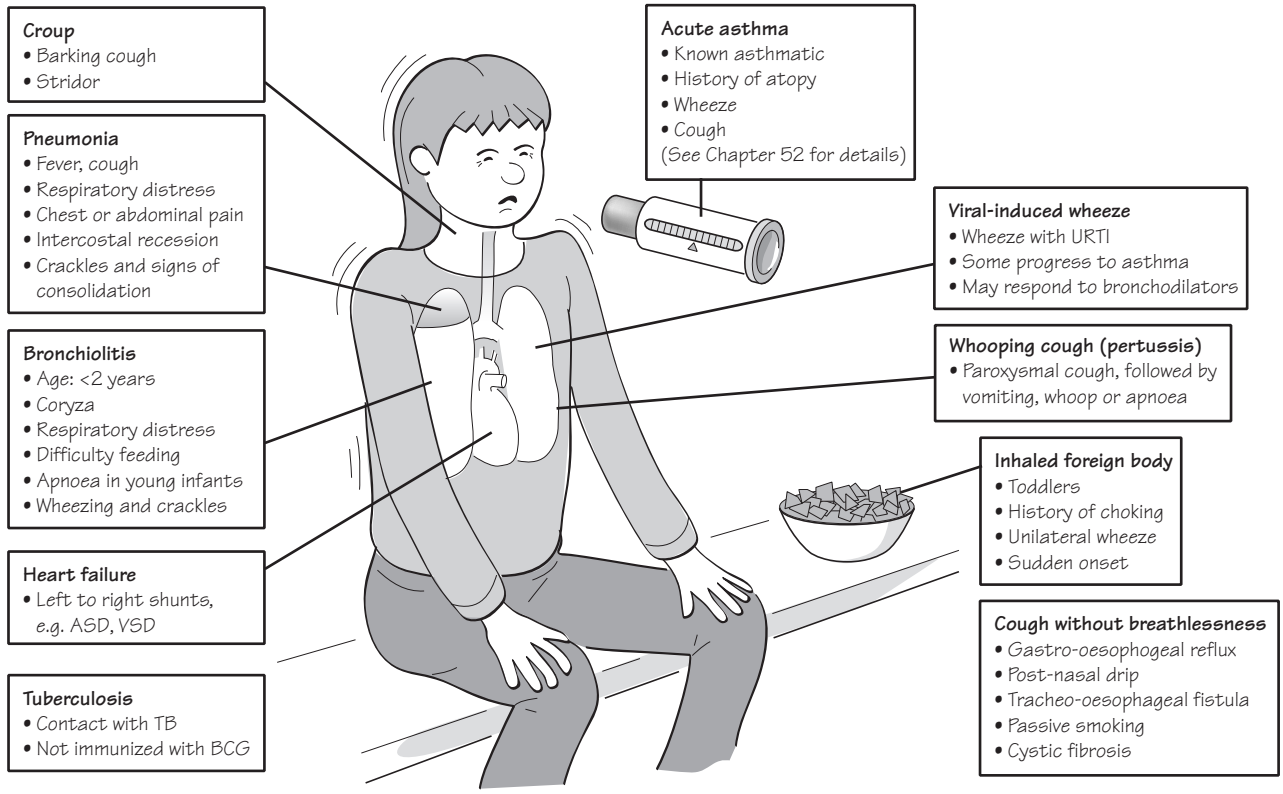
Bile-stained vomiting in the first days of life should always be investigated urgently. It may be due to congenital duodenal or ileal atresia or to a malrotation of the small bowel. Duodenal atresia is more common in Down syndrome. Other causes of bowel obstruction include Hirshprung's disease (colonic aganglionosis) and meconium ileus (in cystic fibrosis). In older infants intussusception should be suspected (see p. 59). All infants with bile-stained vomiting should have a naso-gastric tube passed to aspirate the stomach and feeds should be stopped pending investigation with an upper gastro-intestinal contrast study. In congenital malrotation the small bowel is rotated on its mesentery and a Doppler ultrasound may show malalignment of the mesenteric vessels. Once the cause of the obstruction has been identified and the child has been rehydrated, definitive surgery can take place.

KEY POINTS

- Vomiting is often due to infection or gastroenteritis.
- Pyloric stenosis presents at 4–6 weeks with projectile vomiting.
- Gastro-oesophageal reflux is common and usually responds to simply thickening the feeds.
- Bile-stained vomiting in an infant is a serious symptom which always requires investigation.

20 The chesty child

Causes of cough and breathlessness



What you need from your evaluation

History

- Are there features of infection such as pyrexia or poor appetite?
- Is there a history of previous episodic breathlessness suggesting recurrent asthma?
- Is the child atopic—asthma, hayfever, eczema?
- Is there a relevant family history? e.g. asthma, cystic fibrosis, TB
- Is there an underlying condition, such as congenital heart disease or prematurity, that increases the risk of severe bronchiolitis?

Examination

- Are there signs of respiratory distress—grunting, nasal flaring, intercostal recession, tachypnoea?
- Are there any additional noises—wheeze, stridor, cough?
- Are there signs of consolidation—reduced air entry, crackles, bronchial breathing, dullness on percussion and reduced expansion? (NB: signs are often not focal in young children)
- Are there signs of a chronic respiratory condition, e.g. finger clubbing, chest deformity?
- Is there evidence of congenital heart disease?
- Is there cyanosis?
- Is the child pyrexial?
- Can the child talk in full sentences?
- Is the peak expiratory flow rate (PEFR) normal?

Investigations and their significance

• Chest X-ray	Focal consolidation suggests bacterial infection; diffuse suggests viral or atypical pneumonia. Hyperinflation in asthma and bronchiolitis. May be patchy collapse in bronchiolitis
• Full blood count	Neutrophilia in bacterial pneumonia Lymphocytosis in pertussis
• Sputum culture	To isolate causative organisms. Acid-fast bacilli may be seen in TB
• Naso-pharyngeal aspirate	Viral immunofluorescence for respiratory syncitial virus in bronchiolitis
• Per-nasal swab	To isolate *Bordetella pertussis*
• Viral titres	In atypical pneumonia, e.g. mycoplasma
• Blood cultures	In suspected bacterial pneumonia may isolate *Streptococcus pneumoniae* or *Staphylococcus aureus*
• Mantoux test	In suspected TB
• Bronchoscopy	If inhaled foreign body likely, or for diagnostic bronchio-alveolar lavage

Children very commonly present with respiratory symptoms which can include coryza, breathlessness, cough, wheeze or noisy breathing. Often these symptoms are associated with a viral URTI (see p. 49) or asthma (see Chapter 52). Common infectious causes of 'chestiness' are pneumonia, bronchiolitis and whooping cough. Some children will have a persistent cough without any other chest signs, and some of the causes are listed opposite.

Pneumonia

Pneumonia is the term used to describe an infection of the lower respiratory tract, which can be either bacterial or viral. Viral causes include respiratory syncitial virus (see below), influenza virus, para-influenza, adenovirus and Coxsackie virus. Bacterial causes are *Streptococcus pneumoniae*, which often causes lobar pneumonia, *Haemophilus influenzae*, *Mycoplasma pneumoniae* and group B beta-haemolytic *Streptococcus*, which only occurs in the newborn. It is important to consider whether there are any predisposing factors such as a congenital anomaly of the bronchi, inhaled foreign body, immunosuppression, recurrent aspiration (e.g. with a tracheo-oesophageal fistula) or cystic fibrosis.

Acute pneumonia usually presents with a short history of fever, cough and signs of respiratory distress, including tachypnoea and intercostal recession. Grunting is common in infants. Signs include dullness to percussion, bronchial breathing and crackles, reflecting the underlying consolidation. Clinical signs are often not reliable in infants and the diagnosis should always be confirmed by chest X-ray. This may show a lobar pneumonia or a more widespread bronchopneumonia. With lobar pneumonia a repeat X-ray is usually performed 4–6 weeks later to confirm that the infection has resolved. Blood and sputum cultures may reveal the organism. Antibody titres or cold agglutinins may be useful in diagnosing mycoplasma pneumonia, which often has a more insidious onset and requires treatment with erythromycin. Penicillin is the first-line antibiotic for lobar pneumonia. Complications of pneumonia include pleural effusion, septicaemia, bronchiectasis, empyema (infected pleural effusion) or lung abscess (may follow staphylococcal pneumonia).

Bronchiolitis

Bronchiolitis is an acute viral infection which causes respiratory distress and wheezing in infants less than 18 months, due to obstruction of the small airways. It is usually caused by respiratory syncitial virus (RSV) and occurs in epidemics in the winter months. Adenovirus, influenza and para-influenza virus can also cause bronchiolitis. Coryza is followed by cough, respiratory distress and wheeze. Some infants will have difficulty feeding or may have apnoea. Examination reveals widespread wheeze and fine crackles and over-expansion of the chest. Chest X-ray will show hyperinflation and patchy collapse or consolidation. A nasopharyngeal aspirate (NPA) can be taken to look for RSV in the respiratory secretions using immunofluoresence. Most children with bronchiolitis do not require any specific treatment but indications for admission to hospital include poor feeding, apnoea, increasing respiratory distress or the need for oxygen. The illness usually lasts 7–10 days and most recover fully although bronchiolitis may be associated with recurrent wheezing during infancy. A minority of children, particularly those with chronic lung disease or an underlying congenital heart defect will require mechanical ventilation on an intensive care unit, and bronchiolitis has a mortality of 1–2%. A monoclonal antibody (Palivizumab) against RSV can be given prophylactically to high-risk infants throughout the winter months to provide passive immunity against infection.

Whooping cough

This is a form of pneumonia caused by *Bordetella pertussis*, which tends to occur in young infants or in those who have not been fully vaccinated. The same symptoms can be caused by parapertussis infection, which is not prevented by the pertussis vaccine. In older children whooping cough presents with a coryzal illness followed by paradoxical coughing spasms during expiration, followed by a sharp intake of breath—the whoop. They may turn red or blue in the face and may vomit due to the coughing. In infants it can cause apnoea. Diagnosis is mainly clinical, although a lymphocytosis ($>20\times10^9$/l) is usually present in the blood. The organism may be cultured from a per-nasal swab. Treatment is supportive, although erythromycin can shorten the duration of the illness if it is given early during the coryzal phase.

The paroxysms of coughing can continue for several months after the initial infection. The risks of hypoxic brain injury from acute whooping cough far outweigh the risks of brain damage from the vaccine, and universal vaccination is recommended at 2, 3 and 4 months of age.

KEY POINTS

- The majority of children with 'chestiness' will have a self-limiting viral URTI and do not require antibiotics.
- If a child has recurrent episodes of pneumonia, an underlying cause should be sought and excluded.
- Bronchiolitis is very common in winter and carries a significant mortality, especially amongst infants with chest or cardiac disease.
- Whooping cough can usually be diagnosed by the characteristic paroxysmal cough and associated colour change.

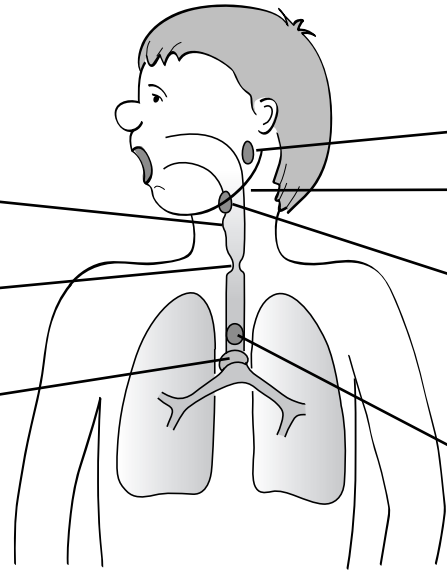
21 Stridor

Chronic stridor

Laryngomalacia
- Present from birth
- Worse on crying
- Improves with age
- Well, thriving child

Subglottic stenosis
- Previous intubation
- Worse with URTI

Vascular ring
- Congenital defect of great vessels
- Worsens over time
- Barium swallow may show indentation



Acute stridor

Croup
- Barking cough
- Coryzal illness

Tonsillar abscess (quinsy)

Anaphylaxis

Epiglottitis
- Sudden onset
- Septic
- Drooling
- Unable to speak
- No Hib vaccination

Inhaled foreign body
- Toddlers
- Sudden onset
- History of choking
- Unilateral signs
- Requires bronchoscopy

What you need from your evaluation

History

- How long has the stridor been present? In a well baby stridor that comes and goes and has been present from birth is usually due to laryngomalacia (floppy larynx), which usually improves with time. Persistent fixed stridor may be due to a vascular ring or, more rarely, vocal cord palsy, or severe micrognathia (e.g. Pierre Robin sequence)
- Does the child look acutely ill? The commonest cause of stridor is croup—it is often worse at night and associated with a barking cough and preceding coryzal symptoms. Always consider epiglottitis, which presents more quickly in a very ill child who cannot swallow or speak
- In any child with sudden onset of stridor, ask about choking as an inhaled foreign body must always be considered
- Is there any history of allergy that would suggest anaphylaxis?

Examination

- Assess the severity by the work of breathing, the presence of intercostal recession and the degree of oxygenation (by colour or by saturation monitoring if available)
- Unilateral wheeze or chest hyperexpansion suggests an inhaled foreign body
- An urticarial rash and angioedema suggest anaphylaxis
- If the child is sitting forwards, unable to swallow and is acutely unwell, consider epiglottitis-in this instance do not try to examine the throat until the airway has been secured. Call for senior anaesthetic help before examining the child
- In chronic stridor assess the shape and size of the jaw. Listen for murmurs which may suggest congenital heart disease, where abnormal great vessels can compress the airways

Investigations and their significance

Investigations will be determined by the likely diagnosis as follows:

• Foreign body	Chest X-ray for unilateral hyperexpansion or radio-opaque objects Rigid bronchoscopy to find and retrieve the object
• Croup	Usually none required
• Epiglottitis	Do not perform investigations until airway secured! Blood culture and FBC
• Persistent stridor	Microlaryngoscopy (if infant not thriving or stridor very severe) to assess larynx and vocal cords Barium swallow (may show indentation of vascular ring)

Stridor is an inspiratory noise usually caused by narrowing of the extrathoracic upper airway. It is a very common symptom in young children and infants, but in a minority of cases can represent severe life-threatening disorders such as inhaled foreign body or epiglottitis. It may be chronic, due to a congenital abnormality, or acute, usually due to infection or obstruction.

Croup (acute laryngotracheobronchitis)

This common condition affects children aged 6 months to 3 years and is due to a parainfluenza infection of all the upper airways. It is commoner in winter and can be recurrent. Croup starts with coryzal symptoms, then proceeds to stridor, wheeze and a barking cough. Children may have a hoarse voice. It is usually self-limiting, lasting only a few days, but can occasionally be very severe requiring intubation and ventilation. Signs of severe croup include increased work of breathing, cyanosis and restlessness. Milder cases can be managed by observation and maintaining good hydration. Nebulized budesonide and oral dexamethasone have been shown to reduce the severity of symptoms and the need for hospital admission. Steam and humidity have not been proven to be beneficial but may provide some symptomatic relief. In those not immunized against *Haemophilus influenzae* it is important to consider epiglottitis as an alternative diagnosis. In toddlers always consider an inhaled foreign body.

Acute epiglottitis

This life-threatening infection is caused by *Haemophilus influenzae* and is now rare thanks to immunization with Hib vaccine. It presents in older children (2–4 years) with signs of sepsis, and an inability to swallow or talk. Children often lean forwards to maintain a patent airway and may drool saliva. If epiglottitis is suspected, examination of the throat is contraindicated as it may precipitate complete airway obstruction. The child should be transferred immediately to an operating theatre for intubation by an experienced anaesthetist. At laryngoscopy a 'cherry red' swollen epiglottis confirms the diagnosis. Occasionally an emergency tracheostomy may be needed. Once the airway is protected, blood cultures can be taken and IV antibiotics (cefotaxime) given. Extubation is usually possible after 48 h. Once the airway has been protected the prognosis is excellent.

Distinguishing croup and epiglottitis

It can sometimes be difficult to distinguish between croup, epiglottitis and bacterial tracheitis (infection of the trachea). Epiglottitis usually affects slightly older children than does croup, has a sudden onset without a preceding coryza and children look acutely 'septic'. They are usually unable to talk, with minimal cough, whereas in croup there is a hoarse voice and a barking cough.

Laryngomalcia

Laryngomalacia (floppy larynx) usually presents with a variable stridor from soon after birth. It is loudest when the infant cries but disappears when they are settled. The stridor is caused by prolapse of the aryo-epiglottic folds which can obstruct the upper larynx at the glottis. This condition usually resolves as the child grows older and a well, thriving baby with a characteristic mild stridor does not necessarily need further investigation.

Chronic or progressive stridor

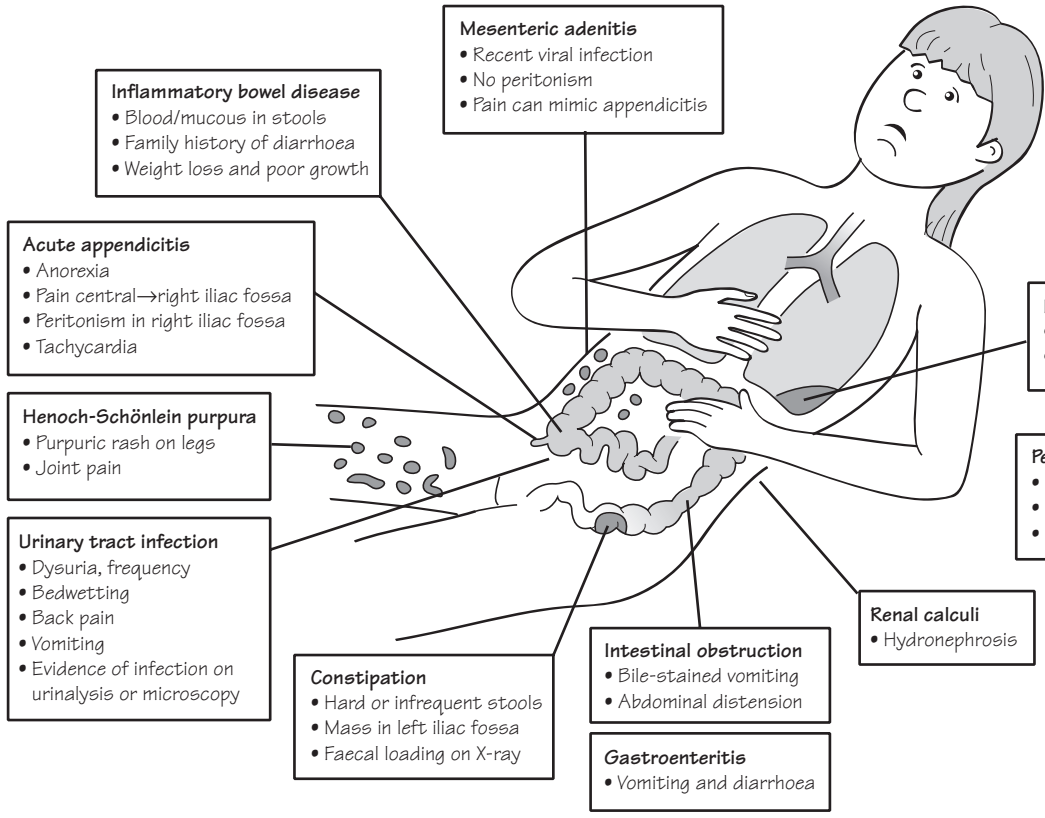
If the infant has signs of respiratory distress or is failing to feed and gain weight adequately, or if the stridor is getting worse, then the child should be investigated with a microlaryngo-bronchoscopy. Rarely this may reveal a more serious cause such as vocal cord palsy, subglottic stenosis or tracheomalacia, which can be associated with a double aortic arch or other vascular ring compressing the trachea. Subglottic stenosis can be congenital but often is due to prolonged tracheal intubation where the trachea has become scarred and narrowed. Papillomata (viral warts) can be acquired vertically from the mother's birth canal—these may grow on the vocal cords and can cause a stridor which gradually progresses over a few weeks.

KEY POINTS

- Stridor suggests upper airway obstruction.
- Always consider an inhaled foreign body.
- Acute epiglottitis is a life-threatening infection.
- Croup responds to corticosteroid therapy.

22 Acute abdominal pain

Causes of acute abdominal pain



What you need from your evaluation

History

- Pain in young children may present with intermittent unexplained screaming. Pallor and screaming are suggestive of intussusception. Older children may point to the site of pain. Pain migrating from the periumbilical area to the right iliac fossa suggests appendicitis. Sometimes children experience referred abdominal pain with lower lobe pneumonia
- Blood in the stool is a serious sign and may indicate intussusception, but also occurs in inflammatory bowel disease, Henoch-Schönlein purpura and some types of gastroenteritis
- It is important to ask about associated features such as vomiting, diarrhoea, recent viral infection, joint or urinary symptoms
- Loss of appetite (anorexia) is a particular feature of appendicitis

Examination

- Examination should include an assessment of how ill the child looks, as well as assessing parameters such as pulse, capillary refill time and temperature
- The abdomen should be palpated very gently at first, while watching the child's face for signs of pain
- Signs of peritonism are a reluctance to move, rebound tenderness, guarding and rigidity
- In mesenteric adenitis there is often palpable lymphadenopathy elsewhere

Investigations and their significance

• Full blood count	Leucocytosis found in acute appendicitis and urinary tract infection
• Urine dipstix test	Nitrite test positive in urinary tract infection Haematuria sometimes seen with HSP
• Urine microscopy and culture	Pyuria and presence of organisms indicate infection
• Abdominal X-ray	Dilated bowel loops: intestinal obstruction Abnormal gas pattern: intussusception Faecal loading: constipation
• Abdominal ultrasound scan	To exclude renal tract abnormality and can be very useful in diagnosis of intussusception
• Barium enema/ air enema	For diagnosis and treatment of intussusception
• CRP/ESR	May be elevated in infection and in inflammatory bowel disease

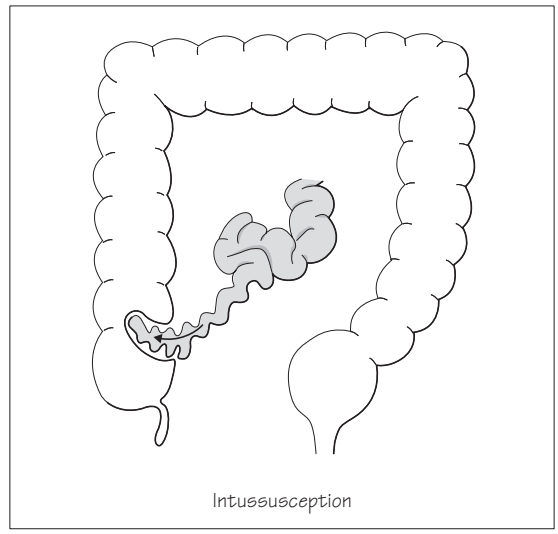
Abdominal pain is a very common symptom in childhood. Acute and chronic abdominal pain are discussed separately as their presentations and causes are quite different. Chronic and recurrent abdominal pain is discussed in Chapter 26. The differential diagnosis of acute abdominal pain includes some important conditions which require surgical intervention. Some of these can present in babies when often there is no clear history of an abdominal problem. These conditions should therefore be considered in any seriously ill child when no other cause can be found. The common causes of acute abdominal pain are described below.

Acute appendicitis

This is the commonest cause of an acute abdomen in childhood and occurs in 3–4 per 1000 children. It can occur at any age but is more common beyond 5 years of age. There is no such condition as the 'grumbling appendix'—the child either has an acute appendicitis or not. Appendicitis is particularly difficult to diagnose in infants and very young children. The presentation in older children is with pain in the umbilical area which moves over a few hours to the right iliac fossa. There is usually anorexia and a reluctance to move. There may be vomiting and a low-grade fever. There may be a leucocytosis and the plasma electrolytes and urine should be checked. Abdominal X-ray is not usually helpful. Once the diagnosis is made the management is an appendicectomy. With skilled surgery the prognosis is excellent. If peritonitis has occurrred there may be severe illness and adhesions may cause later bowel obstruction.

Intussusception

Intussusception is caused by the invagination of one part of the bowel into another; often the terminal ileum into the caecum. It is commonest between the age of 3 months and 2 years. An enlarged Payer's patch (part of the lymphatic system) may form the leading edge of the intussusception and this often follows a viral URTI or gastroenteritis. Classically the child presents with episodic screaming and pallor. Between episodes the child may appear well. Passage of blood and mucous in the stool (so called 'redcurrent jelly' stool) occurs in 75%. A sausage-shaped mass may be palpable in the right side of the abdomen. Abdominal X-ray may show the rounded edge of the intussusception against the gas filled lumen of the distal bowel, with signs of proximal bowel obstruction. Ultrasound can confirm the presence of bowel within bowel—the 'doughnut sign'. The intussusception can often be reduced by an air or barium enema. If this fails or there is evidence of peritonitis then a laparotomy is required for surgical reduction. Unfortunately children still die of intussusception because it can present very non-specifically and the diagnosis is not always considered. If intussusception recurrs the presence of an intestinal polyp should be suspected as the a cause of repeated bowel invagination.


Intussusception

Mesenteric adenitis

This is caused by inflammation of intra-abdominal lymph nodes following an upper or lower respiratory tract infection or gastroenteritis.

The inflamed, enlarged nodes cause acute pain which can mimic appendicitis. With mesenteric adenitis there is no peritonism or guarding and there may be evidence of infection in the throat or chest.

It is usually a diagnosis of exlusion. Treatment is with simple analgesia and the prognosis is excellent.

Other causes of acute abdominal pain

Sometimes acute abdominal pain may be the presenting feature of pathology outside the abdomen. Diabetic ketoacidosis may cause abdominal pain and vomiting (see p. 114). Lower lobe pneumonia may give a referred pain that is described as abdominal pain. In Henoch–Schönlein purpura (HSP) there may be acute abdominal pain as part of a widespread vasculitis (see p. 54). These children are also at risk of intussusception. Urinary tract infection, particularly ascending pyelonephritis causes abdominal pain more than dysuria (see p. 69).

KEY POINTS

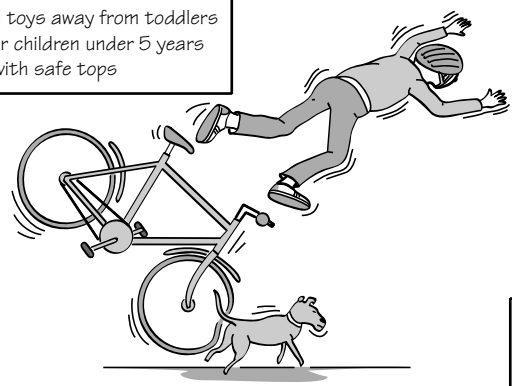
- Intermittent screaming and pallor in an infant may be due to intussusception.
- Appendicitis causes peritonism in the right iliac fossa and anorexia.
- Mesenteric adenitis usually follows an URTI and is self-limiting.
- Lower lobe pneumonia or diabetes can be causes of abdominal pain.
- Urinary tract infection should always be excluded.

23 Accidents and burns

Accidents

Accidents are the commonest cause of death in children after the first year of life and also cause significant morbidity. Each year about 700 children in England and Wales are killed and 10 000 are disabled by accidents. Most accidents are not just chance events but are to some extent predictable, and therefore preventable. As most accidents occur in and around the home, one of the main accident-prevention strategies is parental education and improving the awareness of potential hazards. Some of the common causes of accidents and their prevention strategies are listed below.

Choking
- Keep small toys away from toddlers
- No nuts for children under 5 years
- Use pens with safe tops



Road traffic accidents

This is the commonest cause of accidental death in childhood.
The child is usually a pedestrian or cyclist. Road traffic accidents can be prevented by reducing the speed of traffic and by educating both drivers and children.
- Use child car seats and seatbelts
- Teach road safety to children from a young age
- Traffic calming schemes around schools and playgrounds
- Cycle helmets reduce the number of serious head injuries in cyclists
- Enforce speed limits by use of speed cameras
- Improve access to specialized trauma and neurosurgical centres

Drowning
- Mostly occurs in fresh water (baths, swimming pools, rivers)
- Outcome is better in very cold water due to protective effect of hypothermia
- If the child is resuscitated from a near-drowning, the outcome is usually good

Prevention
- Never leave children unattended in the bath
- Swim only where a lifeguard is present
- Fence off pools and ponds

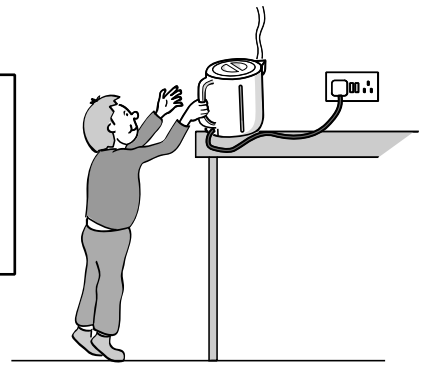
Falls
- Fit stair gates at home
- Fit child-proof window locks
- Soft surfaces in playgrounds

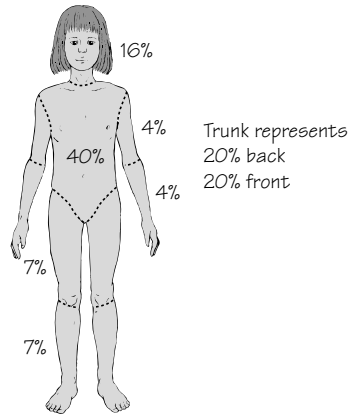
Poisoning (See opposite)

Burns

Every year 50 000 children attend Accident and Emergency departments with burns or scalds. Burns are the second commonest cause of accidental death in childhood after road traffic accidents, and account for about 90 deaths a year. Fatal burns are usually associated with house fires. Half are due to smoke inhalation and half to direct burns. Death from burns arises due to the massive fluid loss through the exposed tissues and due to infection.
The severity of a burn is determined by the temperature and the duration of contact. Most skin burns are due to scalding with hot water or hot drinks

Prevention
- Caution in the kitchen
- Reduce hot water temperature
- Install smoke detectors
- Avoid trailing flexes on kettles and irons
- Use fire guards
- Cover electrical sockets





Management

Remove the heat source and any hot clothing immediately. Cool the skin under a cold tap and wrap the area in a clean sheet or cover with clean cling-film. If there has been smoke inhalation check for wheeze, cyanosis or respiratory distress. There may be soot in the nose and mouth. Check oxygen saturation and carboxy-haemoglobin level (in case of carbon monoxide poisoning). Give high flow oxygen and consider ventilation. The extent of the burns should be assessed and the percentage of the body surface area affecting full-thickness or partial thickness burns should be estimated (palms of hand = 1%). Burns affecting >10% are highly significant and intravenous fluid resuscitation will be required. The fluid management is complicated and depends on the percentage area affected. Give morphine to control pain. Full-thickness burns are less painful than partial ones. Most burns victims are now treated in specialized burns units. Skin grafting may be necessary and psychological support will be needed for the child and the family, especially if there is extensive scarring

24 Poisoning

Accidental ingestion in young children

Accidental poisoning is becoming less common as parents become more aware of the risks and drugs are sold in child-resistant containers. Accidental poisoning most commonly occurs in inquisitive toddlers, especially when they are staying in grandparents' homes where there are likely to be more medicines and household products may be stored less carefully

Common drugs ingested
- Aspirin
- Paracetamol
- Antidepressants

Common household agents
- Disinfectants
- Bleach
- Weedkiller
- Paraffin or white spirit
- Dishwasher tablets

History and evaluation
- Substance ingested
- Time ingested
- Calculate maximum quantity that may have been ingested
- Inspect the product container

Examination
- What is the child's conscious level and are the pupils reacting normally?
- Check pulse and blood pressure and monitor if arrhythmias are likely
- Is there evidence in the mouth of ingestion, *e.g.* ulcers, or clues from the clothing, such as burns or smell?

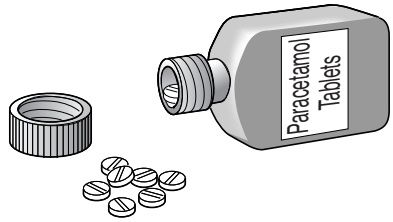
Investigations
- Blood and urine for toxicology if the poison is not known
- Paracetamol, alcohol, salicylate or drug levels, as appropriate
- Blood glucose, especially in alcohol poisoning
- Keep the product and packaging for further analysis

Management
- Discuss with nearest poisons unit
- Where possible remove the poison. Gastric lavage may be necessary, particularly if ingestion was recent (*e.g.* aspirin)
- Activated charcoal can be given to absorb many drugs
- Inducing vomiting with ipecacuanha syrup should only be used if the substance is not absorbed by charcoal and ipecacuanha can be given within 1 h of ingestion of the poison. Avoid if the child is semi-conscious or if the substance is corrosive or likely to be aspirated (*e.g.* paraffin, petrol or turpentine)
- Give specific antidote if available (*e.g.* naloxone for opiates)
- Supportive treatment for respiration. Monitor for cardiac arrhythmias and treat as necessary
- Advice should be given to parents on safety within the home

Intentional overdose in older children and adolescents

Agents used to overdose
- Paracetamol
- Aspirin
- Alcohol
- Drugs of abuse (*e.g.* opiates)
- Sedatives and antidepressants

Risk factors for overdose
- Children in care
- Emotional upset
- Child abuse or bullying
- Psychiatric illness
- Suicidal thoughts (usually rare)
- Other self-harming behaviour



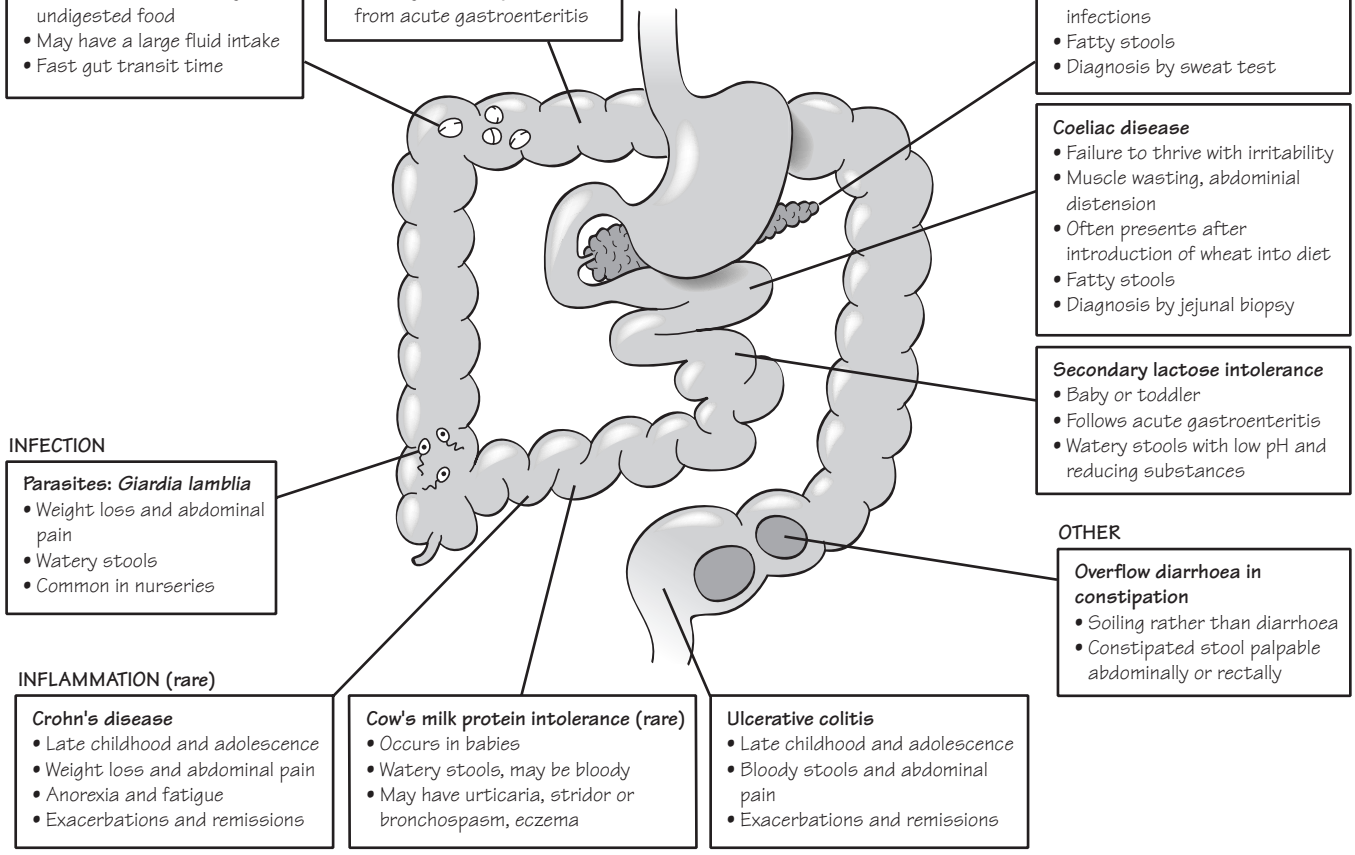
Management
- Evaluation, history and examination, as above
- Removal of poison where possible or administration of charcoal
- Aspirin remains in the stomach for a considerable time and gastric lavage should be considered
- Treatment of the toxic effects of drug, as above
- Admission for assessment by child psychiatrist in all cases
- Consider the possibility of serious risk factors, such as abuse

Paracetamol poisoning
- Rarely severe enough to cause serious problems but liver failure can occur after ingestion of 20–30 tablets and is likely if >150 mg/kg of paracetamol has been ingested
- Activated charcoal should be used where significant quantities have been ingested
- Serum paracetamol levels should be measured 4 h after ingestion and the level plotted on a nomogram. If above the treatment level, an infusion of *N*-acetyl cysteine (Parvolex) should be commenced and continued for at least 24 h. This reduces the risk of liver damage
- The initial symptoms of nausea and vomiting usually settle within 24 h but hepatic necrosis can occur 3–4 days later with the onset of right upper quadrant pain and later encephalopathy
- In significant overdoses, serial measurements of liver enzymes and coagulation times should be made to monitor hepatic function. Serum urea and electrolytes should be used to monitor renal function

25 Chronic diarrhoea

Causes of chronic or recurrent diarrhoea

Frequent stools are often normal in early childhood. Babies have one to seven loose stools per day, which become formed and adult-like in odour and colour after 12 months of age. If the child is thriving and there are no other symptoms or signs, investigations are rarely necessary. Pathological diarrhoeal illnesses can broadly be divided into malabsorption, inflammation and infections.



What you need from your evaluation

History

- **Bowel pattern.** Get an idea of the volume, appearance and consistency of the stools. A diary is helpful in assessing the severity and pattern of symptoms. Is there blood or mucus? NB Odour and 'flushability' are usually not helpful
- **Precipitating factors.** Lactose intolerance is precipitated by acute diarrhoea. Are certain foods troublesome? Are others affected in the family or in childcare?
- **Associated symptoms.** Weight loss or abdominal pain are particularly significant
- **Review of symptoms.** Non-GI diseases often cause diarrhoea and a failure to thrive

Investigations

- These are rarely necessary if a child is thriving and there are no accompanying symptoms or signs

Physical examination

- **Growth.** Obtain height, weight, head circumference and compare with earlier measurements. Chronic disease affects growth. Weight is useful as a baseline if symptoms persist
- **Other features.** Hydration, pallor, abdominal distension, tenderness and finger clubbing are particularly relevant
- **General examination.** Does the child look ill? Look for non-GI diseases that might cause diarrhoea
- **Anorectal examination.** Not routinely indicated

Toddler diarrhoea

Toddlers often experience non-specific diarrhoea, probably due to a rapid gastrocolic reflex. Features are drinking excessive fluids, particularly fruit juices and food particles in the stool. The diagnosis should only be made if the child is thriving. Reassurance is all that is required.

Lactose intolerance

Lactose intolerance is common in babies and young children following gastroenteritis. The superficial mucosal cells containing lactase are stripped off, causing high levels of lactose in the bowel, which prolongs the diarrhoea. Congenital lactose intolerance is rare.

Investigations and their significance

Stool

● Occult blood	Positive in cows milk intolerance, IBD
● Ova and parasites	Parasitic infection
● (3 samples required)	
● Reducing substances	Present in sugar intolerance
● and low pH	(usually lactose)
● Chymotrypsin	Low in pancreatic insufficiency
● Microscopy for fat globules	Globules seen in fat malabsorption (usually pancreatic insufficiency)

Blood

● Full blood count	Anaemia indicates blood loss, malabsorption or poor diet Eosinophilia suggests parasites or atopy
● Plasma viscosity/ESR	High in inflammatory bowel disease
● Coeliac antibodies	A screening test for coeliac disease

Other

● Urine culture	Urinary tract infection
● Sweat test	Cystic fibrosis
● Breath hydrogen test	High H2 in sugar intolerance
● Jejunal biopsy	Flattened villi in coeliac disease
● Barium meal and enema	Characteristic signs in IBD
● Endoscopy	Characteristic lesions in IBD

The diagnosis is suspected if gastroenteritis persists for several days, particularly if the temperature is not raised. A low pH (<6.0) and reducing substances (lactose) are found in the stool. It is rarely necessary to perform lactose challenge or breath hydrogen tests. In bottle-fed babies an empirical change of formula to soy milk (which contains non-lactose sugar) can be tried. The baby should revert to cow's milk once symptoms resolve. The breast-fed baby needs no change of milk.

Coeliac disease

Coeliac disease results from a permanent inability to tolerate gluten, a substance found in wheat and rye. Most children present before the age of 2 years with failure to thrive, along with irritability, anorexia, vomiting and diarrhoea. Signs include abdominal distension, wasted buttocks, irritability and pallor. The stools are pale and foul-smelling. There may also be mouth sores, a smooth tongue, excessive bruising, finger clubbing and peripheral oedema.

Investigations show iron deficiency anaemia and steatorrhoea with fat globules in the stool. Coeliac antibody levels can be used as a screening test, but definitive diagnosis is made on finding subtotal villous atrophy on jejunal biopsy. The treatment is a gluten-free diet, eliminating all wheat and rye products. An improvement in mood, resolution of diarrhoea and good growth occurs promptly. The diet is quite constricting and must be continued indefinitely. The child is often rechallenged with gluten after a period of 2 years (to allow for full villi regeneration) and the biopsy repeated before consigning the child to life-long restriction. Bowel lymphoma may develop as a long-term complication.

Cystic fibrosis

Infants commonly present with diarrhoea and failure to thrive rather than respiratory symptoms. See also Chapter 54.

Inflammatory bowel disease

Inflammatory bowel disease is a cause of chronic diarrhoea in late childhood and adolescence. Both Crohn's disease and ulcerative colitis are characterized by unpredictable exacerbations and remissions. The underlying cause is unknown. The diagnosis is made by barium studies and endoscopy.

Crohn's disease

This presents with recurrent abdominal pain, anorexia, growth failure, fever, diarrhoea, oral and perianal ulcers and arthritis. Remission can be induced by nutritional programmes based on elemental diets. This approach is as effective as steroids and avoids the problem of growth impairment. Immunosuppressant drugs also reduce the need for steroids. Surgical resection may be indicated for localized disease.

Ulcerative colitis

Ulcerative colitis presents with diarrhoea containing blood and mucus. Pain, weight loss, arthritis and liver disturbance may also occur. Treatment is by corticosteroid enemas or suppositories. Sulfasalazine may be given orally, and steroids, immunosuppressive therapy and even colectomy may be required in severe cases. Most cases starting in childhood are severe in terms of activity and extent of involvement. There is a high risk of colonic cancer developing later in life.

Parasites

Giardia lamblia not uncommonly causes outbreaks of diarrhoea in daycare nurseries. It may also be related to travel abroad. The child may be asymptomatic or have diarrhoea, weight loss and abdominal pain. Diagnosis is made on microscopic examination of the stool. Three separate specimens are required as excretion of the cysts can be irregular. A blood count may show eosinophilia and the parasite can also be detected in aspirates obtained at jejunal biopsy for coeliac disease. Treatment is with metronidazole, and in an outbreak asymptomatic carriers should also be treated.

Cow's milk protein intolerance

Allergy to cow's milk protein is rare and often over-diagnosed. The diarrhoea is often bloody, and urticaria, stridor and bronchospasm may occur. Very rarely it can be life-threatening. It is less common in babies who have been breast-fed. The diagnosis is clinical, and symptoms should subside within a week of withdrawing cow's milk. The child should be rechallenged after a period of time (in hospital if original symptoms were severe), and observed for a recurrence of symptoms. Treatment consists of substituting soy milk for cow's milk. In most cases the intolerance resolves in 1–2 years.

Overflow diarrhoea in constipation

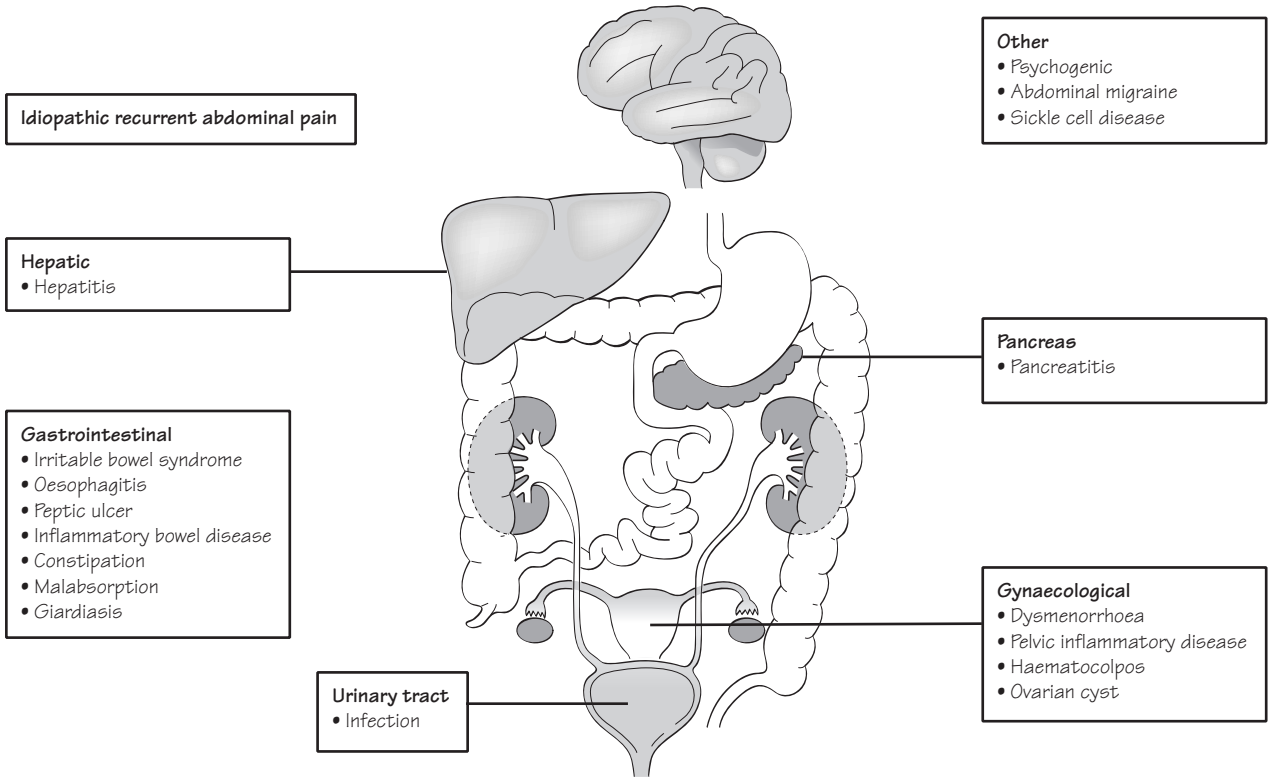
The soiling that results from constipation is sometimes interpreted as diarrhoea. Treatment is directed towards resolving the constipation.

KEY POINTS

- Check that the stool pattern is really abnormal for the age.
- Identify any features suggestive of significant pathology, particularly weight loss or poor weight gain, and abdominal pain.
- Investigations are only required if there are additional symptoms.

26 Recurrent abdominal pain

Causes of recurrent abdominal pain



What you need from your evaluation

History

- Obtain a good description of the pain. Where is it? (non-organic pain is classically periumbilical) Does it affect daily activities?
- Are there constitutional symptoms such as anorexia, weight loss or fever?
- Are there gastrointestinal, urinary or gynaecological symptoms?
- Are there emotional or family problems?

Physical examination

- **Growth:** weight loss or fall-off in growth indicates serious pathology
- **General examination:** look for pallor, jaundice and clubbing
- **Abdominal examination:** is there hepatomegaly, splenomegaly, enlarged kidneys or a distended bladder?
- **Anorectal examination:** not routine in children

Investigations and their significance

Investigations are required only if your evaluation suggests an organic cause

• Full blood count	Anaemia, eosinophilia, infection (leucocytosis)
• ESR or plasma viscosity	Inflammatory bowel disease
• Liver function tests	Liver dysfunction
• Urea and electrolytes	Renal failure
• Amylase	Pancreatitis
• Urinalysis and culture	Urine infection
• Stool for ova and parasites (3 samples)	GI parasites, e.g. giardiasis
• Occult blood	GI blood loss, e.g. inflammatory bowel disease or peptic ulcer
• Abdominal and pelvic ultrasound	Urinary obstruction at all levels, organomegaly, abscesses, pregnancy, ovarian cyst and torsion
• Plain abdominal X-ray	Constipation, renal calculi if radiopaque, lead poisoning
• Barium swallow and follow-through	Oesophagitis and reflux, peptic ulcer, Crohn's disease, congenital malformations of the gut
• Barium enema	Ulcerative colitis
• Endoscopy	Oesophagitis and reflux Peptic ulceration Colitis

Ten to fifteen per cent of school-age children experience recurrent abdominal pain at some point. Only one in ten have an organic problem. A good clinical evaluation is essential as it is rare for organic problems to present with abdominal pain alone, although inflammatory bowel disease, chronic urine infections and parasites may do so.

Idiopathic recurrent abdominal pain

The majority of children presenting with recurrent abdominal pain have no identifiable organic cause. In this circumstance the expression 'recurrent abdominal pain' is often used as a diagnostic term in itself implying that the pain is functional rather than organic. The pain can be very real and severe. The periodicity of the complaint and the intervening good health are characteristic. The children are often described as being sensitive, highly strung and high-achieving individuals, although this is by no means always true. Management must be directed towards reassurance, maximizing a normal lifestyle and minimizing school absence. In the majority of children the pain resolves over time.

An approach to the child with non-organic recurrent pain (abdominal, headaches and 'growing pains'):

- Assure the parents and child that no major illness appears to be present.
- In those children where neither an organic nor psychosomatic cause is found, it can be helpful to use terms such as tension headache, or growing pains, while qualifying this with an explanation that the aetiology is unknown.
- Identify those symptoms and signs which the parents should watch for and which would suggest the need for a re-evaluation.
- Do not communicate to the parents that the child is malingering.
- Liaise with the school to ensure consistent attendance.
- Develop a system of return visits to monitor the symptom. Having the family keep a diary of pain episodes and related symptoms can be helpful.
- During return visits allow time for both the child and parent to uncover stresses and concerns.
- Make every effort to normalize the life of the child, encouraging attendance at school and participation in regular activities.

Other causes

Psychogenic abdominal pain

In some children the abdominal pain is truly psychosomatic and related to stress at home or at school. Obviously these underlying causes must be addressed. In most cases simply indicating the link and explaining that children tend to experience tummy-aches in a similar way to which adults experience headache is enough to reassure the parents and child. An understanding attitude while maintaining that absence from school is unnecessary is a good approach.

Irritable bowel syndrome

This term is sometimes used instead of 'recurrent abdominal pain', particularly if there are minor GI symptoms, and no psychological stresses identified. It has been suggested that the discomfort results from a dysfunction of the autonomic system of the gut. The bowel pattern may be described as varying from pellets to unformed stool. Gas can also be a feature and many of these children give a history of colic as babies. Using the term irritable bowel syndrome often gives families the reassurance that a 'diagnosis' has been made. The symptoms usually resolve over time, but relapses are common

Peptic ulcer

Peptic ulcer is now recognized as an important cause of childhood abdominal pain. The features may be similar to adult ulcer symptoms—epigastric, relieved by food, and there may be a family history. If the diagnosis is suspected a trial of antacids may be used empirically, but if symptoms are persistent confirmation is required by barium studies or endoscopy. Treatment consists of H_2-receptor antagonists and eradication of *Helicobacter pylori* with antibiotics.

Parasitic infestations

The commonest GI parasite in this country is *Giardia lamblia*. Inspection of the stool (three separate samples are required) is merited in all children with recurrent abdominal pain. Threadworms do not cause pain, nor are they detectable on examination of the stool.

Constipation

See Chapter 27.

Inflammatory bowel disease

See p. 63.

Urine infections

See p. 69.

Sickle-cell disease

Abdominal pain is a feature of sickle cell crisis. See p. 97.

KEY POINTS

Nonorganic pain characteristically is:

- Periodic pain with intervening good health.
- Often periumbilical.
- May be related to school hours.

Organic pain—consider this if there is:

- Pain occurring at night.
- Weight loss, reduced appetite, lack of energy, recurrent fever.
- Organ-specific symptoms, e.g. change in bowel habit, polyuria, menstrual problems, vomiting, occult or frank bleeding from any orifice.
- Ill appearance, growth failure, swollen joints.

27 Constipation

Causes of constipation

Acute causes

Fluid depletion
- Caused by fever or hot weather
- May require laxatives
- May lead to chronic constipation

Bowel obstruction
- Rare and due to congenital gut malformations
- Usually presents as acute abdomen, but may present as constipation with vomiting and abdominal pain

Chronic causes

Functional constipation
- Common, particularly in disabled children
- Often stems from withholding from painful defaecation
- May cause megacolon
- Management involves laxatives, bowel training and diet
- Often recurs

Hirschsprung's disease
- Onset in newborn period or infancy
- Failure to thrive and abdominal distension are features
- Diagnosis is by rectal biopsy

What you need from your evaluation

History

- Hardness of the stool, painful defaecation, crampy abdominal pain and blood on the stool or toilet paper are symptoms of constipation. Infrequent but normal stools are not (although very long-standing constipation can be painless). History of an anal fissure is significant
- Onset in infancy suggests Hirschsprung's disease—functional constipation has a later onset
- Precipitating events include mismanagement of toilet training, and fluid depletion caused by hot weather, a febrile illness or vomiting
- Ask about diet as a basis for giving dietary advice on management of constipation

Examination

- **Growth**
  Review the growth chart as Hirschsprung's disease is accompanied by failure to thrive
- **Abdominal examination**
  Hard indentable faeces are often palpated in the left lower quadrant
- **Anorectal examination**
  Rectal examination is not usually indicated, but will reveal hard stools. An anal fissure may be found on inspection of the anus

Investigations

- Plain abdominal X-ray is not usually required, but may show enormous quantities of faeces in the colon
- Hirschsprung's disease is diagnosed by rectal biopsy, indicated if the history goes back to infancy and/or there is poor growth

Management of constipation.

Stage 1: Evacuation of the bowel	
	Diet: in simple cases diet alone is effective
	Laxatives: stool softeners (e.g. lactulose) and/or bowel stimulants (e.g. senokot) may be needed. Increase the dose until the stools become liquid, then reduce
	Enemas: rarely required
	Manual evacuation under general anaesthetic: occasionally required in severe cases
Stage 2: Maintenance	
	Stools should be kept soft by either diet or laxatives for 3–6 months
	Encourage daily bowel movements by sitting the child on the toilet at a fixed time once or twice each day for 5–10 min
Stage 3: Vigilance	
	Start treatment at the first indication of recurrence of hard stools

In normal children the frequency of bowel movements ranges from more than two per day to none for several days. Infrequent bowel movements are common in exclusively breast-fed babies.

Constipation is the passage of hard infrequent stools with painful defaecation. Asymptomatic infrequent bowel movements alone do not constitute constipation.

Soiling refers to faecal staining of the underwear and results from leakage of liquid stool around impacted faeces when a child is constipated. It can be mistaken for diarrhoea. The term is also sometimes used when a child is delayed in gaining bowel control.

Encopresis is the voluntary passage of formed stool in inappropriate places (including underwear) by a child who is mature enough to be continent. It is indicative of severe behavioural problems.

Functional constipation

Constipation often stems from painful passage of a hard stool, causing an anal fisssure. The child withholds further stools to avoid pain. Water is reabsorbed from the colon making the stools harder and more painful to pass. The cycle becomes self perpetuating and the rectum so stretched that colonic dilatation may occur (megacolon). Management is directed at evacuating the bowel, maintenance treatment and good diet. Constipation often recurs, but is controllable with active management.

Foods that can promote good bowel habits.

High fibre foods	Wholewheat bread and flour
	Bran
	High-fibre breakfast cereals
	Fruit (particularly the peel)
	Vegetables
	Beans
	Nuts
Stool softeners	Fluids of any sort
	Orange juice, prune juice
	Fruit

Hirschprung's disease

Hirschprung's disease is caused by the absence of ganglion cells in the bowel wall nerve plexus. It usually presents in the newborn period with delayed passage of meconium and abdominal distension, but may present later with constipation and failure to thrive. Diagnosis is made by rectal biopsy when the abnormal nerve plexus is identified. Management is surgical with resection of the abnormal section of bowel.

KEY POINTS

- Constipation is common and usually functional.
- Constipation from infancy, in conjunction with failure to thrive suggests Hirschsprung's disease.
- Breast-fed babies often have infrequent stools, this is normal.

28 Urinary symptoms

Bedwetting and daytime wetting

Nocturnal enuresis refers to wetting whilst asleep. It usually occurs in normal children and is due to a delay in the development of the normal sphincter control mechanisms. Day and night wetting (diurnal enuresis) may be due to poor bladder sensation or bladder muscle instability. Secondary enuresis refers to wetting in a child who had previously been dry, and is often associated with psychological stress

Primary nocturnal enuresis
- Common: 10% of 6-year-olds and 3% of 12-year-olds wet the bed once a week
- Often familial. Twice as common in boys than girls

Causes
- Delayed maturation (often familial)
- Emotional stress
- Urinary tract infection
- Polyuria due to diabetes or renal disease

Causes of secondary enuresis
- Emotional upset
- Urinary tract infection
- Diabetes mellitus

Causes of diurnal enuresis
- Urinary tract infection
- Neurogenic bladder
- Congenital abnormality (e.g. ectopic ureter)
- Severe constipation
- Psychogenic, due to stress
- Sexual abuse
- Physiological (urgency)

What you need from your evaluation

History

- Has the child ever been dry? If so, at what age? Was there a particular trigger that led to wetting again (e.g. birth of a sibling)?
- Is there a family history of primary nocturnal enuresis? Ask about siblings, parents and grandparents
- Is there anything to suggest stress as a cause? Is there any possibility of sexual abuse?
- Is there any dysuria, frequency or systemic upset to suggest a urinary tract infection?
- Has there been a sudden onset of polyuria, polydipsia, or weight loss to suggest diabetes mellitus or other renal disease?
- How have the parents dealt with the wetting? Have they punished or criticised the child for wetting in the past?
- What methods have they tried, e.g. fluid restriction, lifting onto the toilet at night, star charts?
- What is the pattern of the wetting—nocturnal only, day and night, with urgency or with dribbling incontinence? Are there any features in the history to suggest a neuropathic bladder?

Examination

- Is there any evidence of a neurological or congenital abnormality? Check leg reflexes and perineal sensation.
- Look for evidence of spina bifida occulta such as a lipoma or hairy patch over the sacral area
- Is the child constipated? Is there a palpable faecal mass?
- Is there evidence of renal disease?
- Check for hypertension

Investigations and their significance

Urine microscopy and culture	To exclude UTI
Urine dipstick	To exclude glycosuria
Renal ultrasound and isotope scan, Intravenous urogram (IVU)	If ectopic ureter strongly suspected. (This causes a constant dribbling or incontinence as it connects to the vagina rather than the bladder)

Key points

- Enuresis is common—15% of 5-year-olds wet the bed
- There is rarely an organic cause
- The majority respond to behavioural management
- Psychological stress should be considered in secondary enuresis

Management of primary nocturnal enuresis

- Intervention is not usually advised until the age of 7 years or more, when the child can take some responsibility and normal sphincter control has developed
- Behavioural management with star charts and rewards for dry nights is helpful
- An enuresis alarm, which wakes children when they start to urinate so that they learn to wake up and go to the toilet, is often the most effective treatment and works within a few weeks
- Avoid caffeinated drinks and fruit juice. Do not overly restrict fluid intake. Lifting is best avoided as it trains the child to void whilst half asleep
- Vasopressin (antidiuretic hormone) can be given by nasal spray or tablets at night. This reduces urine output and can be particularly useful for short periods such as going on camp or staying with friends. Oxybutynin reduces detrusor muscle instability in children with a small bladder capacity and urgency

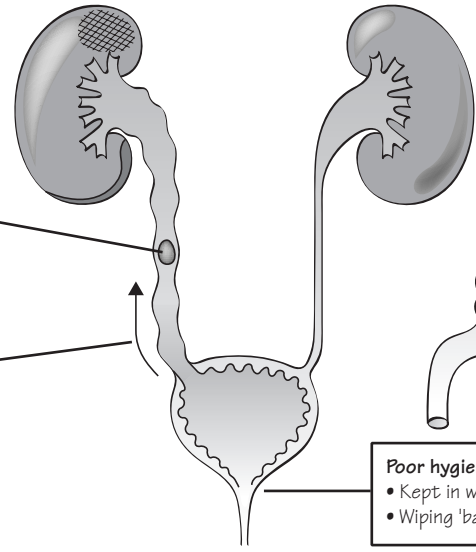
Urinary tract infection

Urinary tract infections (UTIs) are common: they occur in 3% of girls and 1% of boys and 90% are due to infection with *Escherichia coli*. It is important to make a definite diagnosis as a UTI may indicate a congenital renal anomaly or vesicoureteric reflux, which if left untreated may lead to renal failure

Underlying causes of UTI

Obstructed urinary system
- Pelviureteric obstruction
- Urinary stones
- Posterior urethral valves (in boys with poor urinary stream)
- Duplex kidney with obstructed pole
- Horseshoe kidney (associated with Turner's syndrome)

Vesicoureteric reflux
- Retrograde flow of urine from the bladder up into the ureters, renal pelvis or pelvicalyceal system. Can cause hydronephrosis



Idiopathic
- No cause is found for many UTIs

Constipation
- Poor bladder emptying

Poor hygiene
- Kept in wet nappies
- Wiping 'back to front' in girls

What you need from your evaluation

History

- Ask about non-specific fever, irritability and vomiting, especially in infants
- Is there dysuria, frequency or bed-wetting?
- Are there signs of pyelonephritis such as loin pain, vomiting or systemic illness?
- Ask about constipation and assess fluid intake

Examination

- UTI can present with prolonged jaundice, septic shock or failure to thrive in the neonatal period
- Is there any tenderness in the abdomen or over the kidneys?
- Check for palpable kidneys and bladder
- Always check blood pressure to exclude secondary renal impairment
- Examine the spine to exclude neuropathic bladder
- Examine the urine: leucocytes, protein and nitrites on dipstick testing suggest a UTI
- If possible, examine the urine under a microscope, looking for leucocytes, red blood cells and organisms. Abnormal crystals may suggest renal stone disease

Investigations

- A pure culture of $>10^5$ colony-forming units with >50 white cells per high power view on microscopy confirms a UTI. Sterile pyuria (white cells without a growth) can occur in any febrile illness or in renal tuberculosis or inflammation.
- A mixed growth or growth without white cells suggests contamination Any organisms seen in a supra-pubic aspirate sample confirm infection
- Renal ultrasound to look for hydronephrosis, anatomical abnormalities and renal cortical damage. **This should be performed on all children after their first confirmed UTI**
- Abdominal X-ray if any suggestion of renal stones
- DMSA isotope scan is performed in children with recurrent UTIs to look for renal scarring
- DTPA or Mag3 isotope scan can be used to assess obstruction and is used in preference to intravenous urography
- In infants a micturating cystogram (MCUG) is also performed to exclude posterior urethral valves and to look for vesicoureteric reflux

Treatment

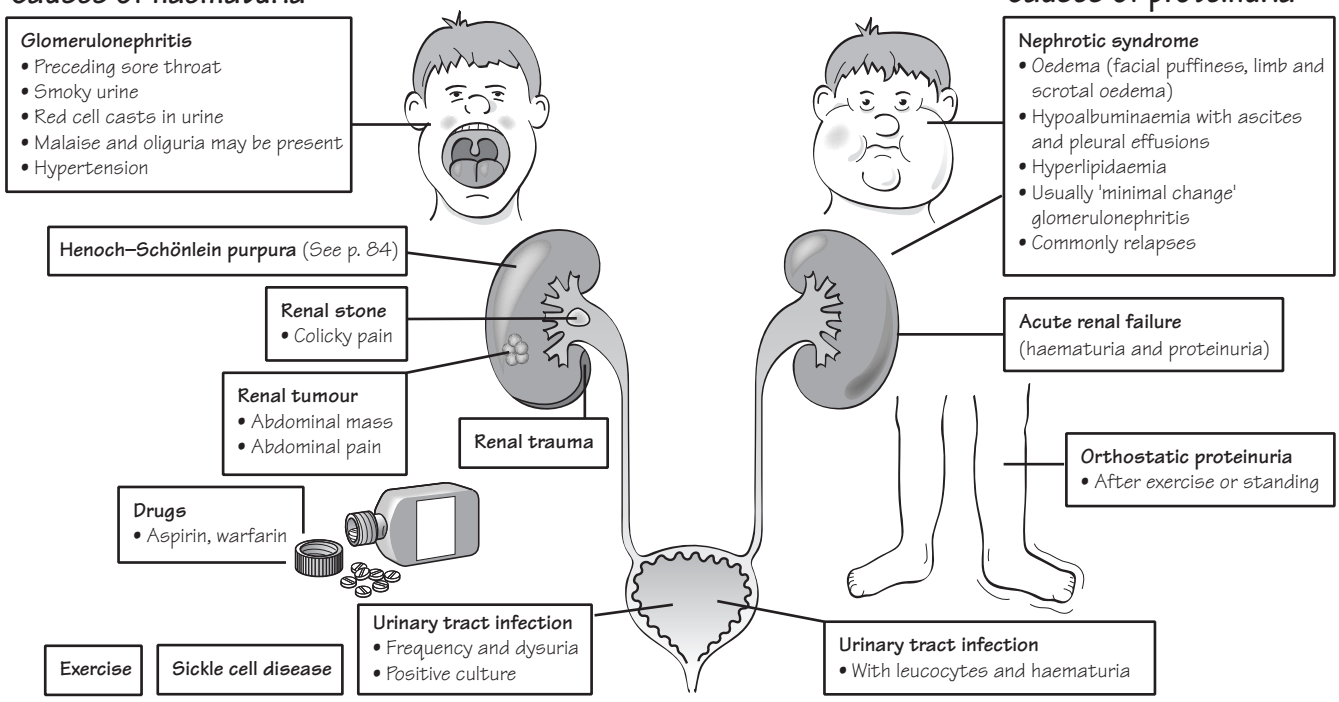
- Trimethoprim is the first-choice antibiotic. Nitrofurantoin, cephradine or amoxil may be effective. A 5-day treatment course is used. A prophylactic dose at night may be recommended for recurrent UTIs or in infants pending investigations
- If there are signs of systemic illness or of pyelonephritis, then intravenous antibiotics are indicated. Gentamicin is the first-line antibiotic in this case
- Analgesia may be required to relieve pain
- Treat any constipation and give advice on good hygiene and maintaining a high fluid intake. 'Double-voiding' of the bladder helps expel residual urine

Key points

- UTIs are common, especially in girls
- Fever may be the only symptom in infants
- Always confirm infection by culture
- Confirmed UTIs require investigation in all children
- In infancy check for obstruction and reflux

Haematuria and proteinuria

Causes of haematuria



Causes of proteinuria

What you need from your evaluation

History

Haematuria

- Make sure you are clear what is being described: is it frank blood, pink urine or a positive dipstick test.(Dipsticks are extremely sensitive to the presence of tiny quantities of blood)
- What colour is the urine? Brown suggests renal origin; fresh red blood or clots suggest bladder origin. Red urine can also be caused by eating beetroot or taking rifampicin
- Are there any other urinary symptoms? Frequency and dysuria suggest a UTI
- Is there severe pain? Renal colic or abdominal pain suggests a calculus (stone) or other obstruction
- Was there a precipitating factor? Enquire about trauma to the kidneys. Throat infections or skin infections may precede acute glomerulonephritis, or nephrotic syndrome and intense exercise may precipitate haematuria
- Is there a family history of renal disease or deafness? (Allport's syndrome causes deafness and nephritis, and is autosomal dominant)?

Nephrotic syndrome

- Has oedema around the eyes been noticed in the morning? Has there been any weight gain?
- What is the urine output? Is the child fluid restricted to a certain volume per day?
- Is this the first presentation or a relapse?
- If the latter, what has the child been treated with in the past?

Investigations and their significance

Urinalysis and culture	For presence of blood, protein, casts or white cells. Pyuria and bactiuria point to a UTI
Full blood count	For anaemia and to exclude HUS
ASO titre/throat swab	For evidence of streptococcal infection
U&E	To assess renal function
Serum C3 complement level	Will be low in some types of glomerulonephritis
Serum albumin level	Low in nephrotic syndrome
Urinary protein/creatinine ratio	High in nephrotic syndrome
Triglycerides and cholesterol level	High in nephrotic syndrome
Renal ultrasound and AXR	May show renal stones
Renal biopsy	If renal function impaired or if there is hypertension, proteinuria and haematuria

Examination

- Blood pressure measurement is mandatory. Hypertension suggests renal disease
- Palpate the abdomen for renal masses (tumour, polycystic kidneys or obstruction) and check for ascites
- Check for pitting oedema over the tibia and sacrum
- Examine for the presence of pleural effusions
- Measure weight and compare with previous values
- Look for any purpuric rash (Henoch–Schönlein purpura (p. 84) or haemolytic uraemic syndrome, HUS)

Acute glomerulonephritis

Acute glomerulonephritis results from immune-mediated damage to the glomerulus. The commonest type in childhood is due to immune complex formation following streptococcal infection. This is common worldwide, although it is now relatively rare in the UK. The presenting complaint is usually haematuria, which is cola coloured and characteristically occurs 1–2 weeks after a throat or skin infection. The child may have malaise, loin pain and headache or be asymptomatic. There may also be mild peri-orbital oedema. Urinalysis shows gross haematuria with granular and red cell casts and sometimes proteinuria. In most children there is mild oliguria (reduced urine output) but in a minority there may be acute renal failure and hypertension. Useful investigations include a throat swab and antistreptolysin O (ASO) titre to look for evidence of streptococcal infection and there may be a low C3 complement level (normal in nephrotic syndrome).

A 10-day course of penicillin is recommended to try to eradicate the streptococcus, although there is no evidence that this alters the course of the disease. The management is similar to that of acute renal failure with strict monitoring of fluid balance and renal function. Salt and fluid restriction may be required and hypertension must be controlled. Very rarely, acute glomerulonephritis leads to acute renal failure when renal dialysis (usually peritoneal dialysis) is required.

Other very rare causes of glomerulonephritis include IgA nephropathy, Alport syndrome (associated with deafness) and Goodpasture syndrome (antiglomerular basement membrane disease, often with associated haemoptysis).

Nephrotic syndrome

Nephrotic syndrome is characterized by proteinuria, low albumin, oedema and high triglycerides. There are a number of causes of nephrotic syndrome, which is due to an increased capillary wall permeability in the glomerulus which allows protein to leak into the urine. The commonest cause by far (85%) is so called 'minimal change' glomerulonephritis, where the histological changes on renal biopsy are very mild. This type is most amenable to therapy. The presenting feature is oedema, which is usually most noticeable in the mornings around the eyelids and as pitting oedema on the legs. There may be history of a recent viral URTI.

With time, weight gain, ascites and pleural effusions develop secondary to the hypoalbuminaemia. Hypertension is rare, but there may be anorexia, abdominal pain, diarrhoea and oliguria. There is an increased risk of infection due to leakage of immunoglobulins, and an increased risk of thrombosis. Urinalysis shows one, two or more 'plusses' of protein and there is a low serum albumin, high triglyceride and cholesterol levels and normal C3 complement.

Treatment of minimal change nephrotic syndrome involves fluid restriction, a low salt diet and corticosteroids (prednisolone). Prednisolone is continued until there is remission of the proteinuria, and then continued at a low dose for 4–6 weeks. Parents should be warned about the immunosuppressive effects of steroids and should avoid live vaccines and chickenpox at this time. Relapses are common, occurring in up to 75% of those who initially respond to steroids. Those who become steroid resistant need a renal biopsy to confirm the pathology and may need treatment with cyclophosphamide. The long-term prognosis is good although relapses may continue for up to 10 years. Other types of nephrotic syndrome (e.g. following HSP) carry a worse prognosis and may progress to chronic renal failure requiring dialysis and eventually transplantation.

Other renal conditions

Acute renal failure

Acute renal failure is defined as a rapid onset of anuria or severe oliguria (<0.5 ml/kg/h). Causes can be divided into prerenal (i.e. poor perfusion), renal or postrenal (due to urinary obstruction). The commonest prerenal cause is hypovolaemia due to gastroenteritis, sepsis or burns. In nephrotic syndrome there may be intravascular hypovolaemia despite extensive extravascular oedema. Intrinsic renal causes include the following:

- Acute tubular necrosis (often secondary to shock).
- Haemolytic uraemic syndrome (HUS).
- Vasculitis and glomerulonephritis.
- Renal vein thrombosis.
- Nephrotoxic drugs (e.g. gentamycin, vancomycin).

Prerenal failure can usually be managed with fluid replacement and inotropic support of the circulation.

Postrenal failure requires relief of the obstruction by catheterization or nephrostomy. Established renal failure requires careful management with fluids restricted to those required to cover insensible losses. No potassium should be added and measures may be needed to control hyperkalaemia. If conservative management is failing, there is severe electrolyte imbalance, progressive acidosis or fluid overload, then renal dialysis is necessary. This can often be achieved using peritoneal dialysis, which uses the peritoneum within the abdomen as a dialysis membrane.

Haemolytic uraemic syndrome

Haemolytic uraemic syndrome is an important cause of renal failure associated with thrombocytopenia, renal failure and haemolytic anaemia due to fragmentation of red blood cells. It often follows an episode of bloody diarrhoea and is associated with a verotoxin producing *E. coli* 0157:H7. The disease can also affect the brain causing an encephalopathy. The prognosis is good if intensive renal support is provided early. Episodic HUS, not associated with diarrhoea, has a worse prognosis with a significant mortality.

Chronic renal failure

Chronic renal failure is unusual in childhood. The commonest cause is a structural renal abnormality such as 'cystic-dysplastic' kidneys or severe obstructive nephropathy. Rarer causes include glomerulonephritis and renal disease as part of systemic disease.

Children with chronic renal failure are usually anaemic, lethargic and have a poor appetite. There may be failure to thrive and renal osteodystrophy due to abnormal vitamin D metabolism. Hypertension and proteinuria are important findings.

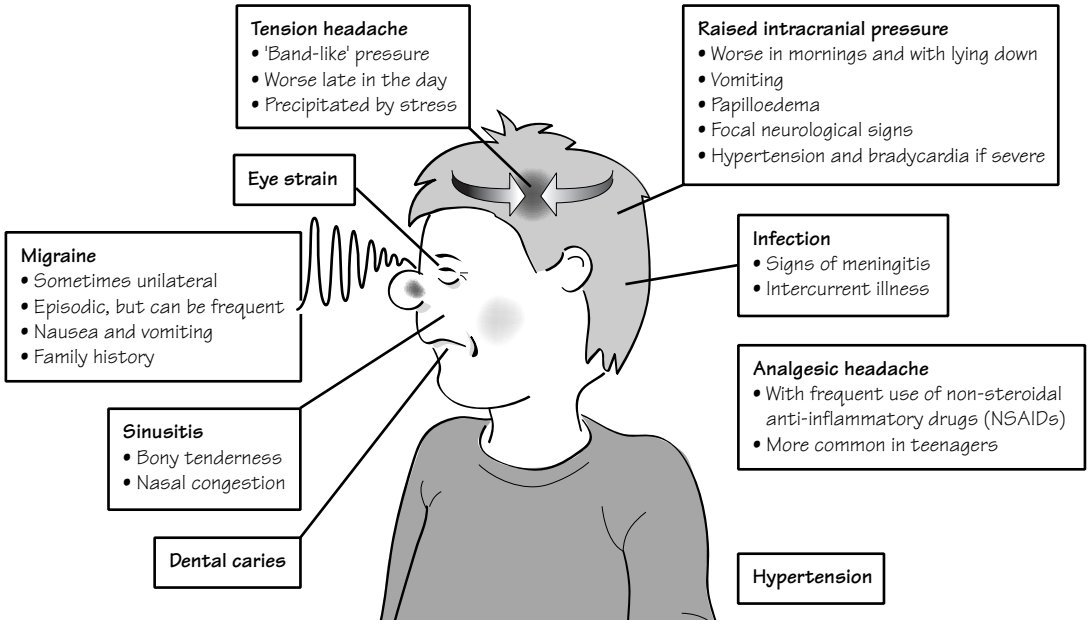
Management involves a high-calorie, low-protein diet that is low in phosphate. Vitamin D supplements are often required and anaemia may be treated by recombinant erythropoietin injections. When the renal disease becomes end stage, children will require dialysis to prevent uraemia. Dialysis can either be haemodialysis or peritoneal dialysis, which can be administered at home. The best long-term treatment is renal transplant from a cadaveric or living related donor.

KEY POINTS

- Haematuria should always be investigated once a UTI has been excluded.
- Post-streptococcal glomerulonephritis and Henoch–Schönlein purpura are the commonest causes of nephritis.
- Nephrotic syndrome usually responds well to steroid treatment, but relapses frequently occur.

29 Headache

Causes of headache



What you need from your evaluation

History

- Is there a family history of migraine? Migraine often tends to be familial
- Obtain a good description of the headaches. Are they unilateral or bilateral? Tension headaches are described as a tight band around the head. Pain in the frontal bones may suggest sinusitis. Migraine is classically throbbing
- Are there associated symptoms? Ask about vomiting and blurred vision, which may be features of raised ICP
- A headache that is worse in the morning or when lying down suggests raised ICP
- Visual auras, such as halos or zigzag lines, are suggestive of migraine
- Photophobia and neck stiffness in addition to headache would suggest meningitis, although they can occur in non-specific viral infections
- Ask about nasal congestion and pain in the teeth or ears as infection around the skull can present as headache

Examination

- Record the blood pressure
  Feel the pulse: is there a relative bradycardia?
- Examine the fundi: look for signs of papilloedema
- Are there any focal neurological signs?

-cerebellar:	nystagmus, ataxia, intention tremor
-infratentorial:	cranial nerve palsies
-cerebral:	focal seizures, spasticity
-pituitary:	endocrine dysfunction, visual field defects

- Look for evidence of dental caries, sinus tenderness, audible cranial bruits (suggests arteriovenous malformation)

Investigations and their significance

• CT or MRI brain scan	Indicated if signs of raised ICP or any focal neurological signs, or if headache is persisting and not responding to normal analgesia. May show hydrocephalus or space-occupying lesion

Headaches are usually benign and serious headaches due to raised intracranial pressure can usually be differentiated on clinical grounds. If a headache is acute and severe, and the child is ill, then serious pathology such as intracranial infection, haemorrhage or tumour must be considered.

Features of concern when evaluating headache.

Acute onset of severe pain
Worse on lying down
Associated vomiting
Developmental regression or personality change
Unilateral pain
Hypertension
Papilloedema
Increasing head circumference
Focal neurological signs

Migraine

This is a common condition in school-age children, and is thought to result from constriction followed by dilatation and pulsation of intracranial arteries. Onset is usually in late childhood or early adolescence. Classically the attack starts with an aura such as zigzaggy vision, followed by a throbbing unilateral headache with nausea and vomiting. Sleep usually ends the attack. In younger children the headache may be bilateral with no preceding aura and no vomiting. Parents often describe the child going very pale. Migraines always cause some reduction in the child's ability to function normally during the attack. There is no diagnostic test and physical examination is normal. The diagnosis is made clinically on the basis of the following.

- Episodic occurrence of headache (rarely every day, but can occur several times a week).
- Aura (often visual), though aura is less common in childhood.
- Nausea in 90% of cases, sometimes vomiting.
- Throbbing headache, sometimes unilateral.
- Positive family history, usually in the mother.
- Impairment of normal function during an attack.

The first line treatment is rest and simple analgesia. Avoiding cheese, chocolate, nuts or caffeine may be helpful. Very frequent or severe attacks may warrant prophylaxis with beta-blockers or pizotifen. Migraine often persists into adulthood, but spontaneous remission does occur.

Tension headache

Tension headaches are common in older school-age children. They are due to contraction of neck or temporal muscles and are felt as a constricting band-like ache, which is usually worse towards the end of the day but does not interfere with sleep. The cause is often difficult to identify, but a proportion of these children will be under some stress, either at home or school. Other family members may suffer similar headaches. Physical examination is normal. Management involves reassurance that there is no serious pathology, rest, sympathy and simple analgesia. Any underlying stress or anxiety in the child's life should be addressed. School absence should be minimized, and the school may need to be involved in developing a management strategy for when the headaches occur. Tension headaches usually become less frequent or resolve spontaneously as the child gets older.

Raised intracranial pressure

Brain tumours, subdural haematomas and abscesses are all rare causes of headache in children. Anxiety about brain tumours is common amongst parents, though these rarely present with headache alone. If a headache is particularly persistent then neuro-imaging may be required to put everyone's mind at rest.

Headaches due to raised intracranial pressure are classically worse on lying down and worse in the mornings, and may wake the child from sleep. There may be associated vomiting, often with surprisingly little nausea. Raised intracranial pressure may also cause blurred vision, high blood pressure and focal nerve palsies (e.g. sudden onset of squint). If papilloedema, hypertension, bradycardia or focal signs are present an urgent CT or MRI brain scan is indicated. The majority of brain tumours are in the posterior fossa or brainstem, so the site of the pain is usually non-specific. They will often have cranial nerve palsies or cerebellar signs. See also p. 118.

Other causes of headache

Headaches are often a feature of minor non-specific viral infections. These should be treated with simple analgesia such as paracetamol. Dental caries, sinusitis and otitis media are all treatable local infections that can cause headache. If headaches seem particularly related to school it is worth checking the child's visual acuity, and considering whether the headaches may be a manifestation of anxiety about school.

KEY POINTS

- Tension headaches are like a constricting band.
- Migraine often has visual symptoms, nausea and there may be a family history.
- Parents are often worried about brain tumours. Raised intracranial pressure, focal neurological signs or unusual features are indications for brain imaging.

30 Fits, faints and funny turns

Types of fits, faints and funny turns

In infants and toddlers

Apnoea and acute life-threatening events (p. 42)
- Found limp or twitching
- Age: <6 months old
- Usually no precipitating event, but consider reflux, sepsis, arrhythmia

Febrile convulsions (p. 47)
- Age: 6 months to 5 years
- Occurs on sudden rise of fever
- Lasts a few minutes

Breath holding spells (cyanotic)
- Older babies and toddlers
- Always precipitated by crying from pain or anger
- Stops breathing, becomes cyanotic and then limp
- No postictal state

Reflex anoxic spells (pallid)
- Precipitated by minor injury
- Turns pale and collapses
- Rapid recovery

Infantile spasms
- A form of myoclonic epilepsy
- Jacknife spasms occurring in clusters
- Developmental regression

Hypoglycaemia and metabolic conditions
- Rare

In school-age children

Epilepsy (see Chapter 60)

Simple absence
- Fleeting vacant look
- 3 per second spike and wave on EEG

Partial epilepsy
- Twitching or jerking of face, arm or leg

Complex partial epilepsy
- Altered or impaired consciousness with strange sensations or semi-purposeful movements such as chewing or sucking
- May have postictal phase

Myoclonic epilepsy
- Shock-like jerks causing sudden falls
- Usually occurs in children with known neurological disability

Syncope (vasovagal)
- Precipitated by pain, emotion or prolonged standing
- Blurred vision, light-headedness, sweating, nausea
- Resolves on lying down

Hyperventilation
- Precipitated by excitement
- Excessive deep breathing, sometimes tetany
- Resolves on breathing into a paper bag

Cardiac arrhythmias (rare)
- Palpitations may occur

What you need from your evaluation

History

- Obtain a description of the episode. Try to visualize the episode and 'replay' back to the witness. What was the child doing at the onset? Were there any precipitating factors? How long did it last? Was there loss or altered consciousness, involuntary movements, or a change in colour (pallor or cyanosis)? How did the child react to the event and was there a postictal phase?
- A developmental history is particularly important if infantile spasms or metabolic conditions are being considered
- Family history: is there anyone in the family with developmental problems, febrile seizures or a metabolic disorder?

Physical examination

- Rarely helpful between episodes
- Undertake a careful cardiac and neurological examination
- Dysmorphic features, micro- or macrocephaly and hepatosplenomegaly suggest a metabolic disorder

Investigations and their significance

The diagnosis is essentially clinical, but investigations must be considered if apnoea, epilepsy or a metabolic problem is suspected

● EEG	-Hypsarrhythmia seen in infantile spasms -3 per second spike and wave activity in absence seizures -Epileptiform activity may be seen in epilepsy (but may be present in normal children)
● ECG	If dysrhythmia is suspected as a cause of syncope or ALTE
● Blood chemistry	Hypoglycaemia, but unhelpful between episodes
● pH monitoring	Apnoea in infants may be due to GOR

Fits, faints and funny turns refer to episodes of transient altered consciousness, which usually present to the doctor after the event is over and may occur recurrently. A good description of the event should allow the different causes to be distinguished from each other, and it can be helpful to ask the family to video the episode. Most of the causes are benign and resolve with age. However, some forms of epilepsy can present in this way and need to be considered in the differential diagnosis. These include simple absence spells, complex partial epilepsy and myoclonic epilepsy, which are covered in more detail on p. 124.

Breath-holding spells

Breath holding spells occur primarily in babies and toddlers.

Cyanotic spells are characteristically precipitated by crying from pain or temper. The child takes a deep breath, stops breathing, becomes deeply cyanotic and the limbs extend. Transient loss of consciousness may occur and even convulsive jerks. The child then becomes limp, resumes breathing and after a few seconds is fully alert. The key to the diagnosis is the typical onset with crying and breathholding and the absence of a postictal phase.

Pallid spells (reflex anoxic seizures) classically follow a bump on the head or other minor injury, which triggers vagal reflex overactivity, causing transient bradycardia and circulatory impairment. The child may or may not cry, but then turns pale and collapses. There is transient apnoea and limpness followed by rapid recovery. The typical history and absence of postictal drowsiness helps distinguish these spells from epilepsy.

For both types of spell reassurance is all that is required, although parents may be quite terrified of recurrences. The attacks are always benign and disappear prior to school age

Syncope (fainting)

Syncope occurs when there is hypotension and decreased cerebral perfusion. It occurs particularly in teenage girls reacting to painful or emotional stimuli, or prolonged standing. Blurring of vision, light headedness, sweating and nausea precede the loss of consciousness which is rapidly regained on lying flat. It is rarely a symptom of cardiac arrythmias or poor cardiac output in childhood. Evaluation includes a cardiac examination, standing and lying blood pressure, and an electrocardiogram (ECG) if there is doubt as to the cause of the faint.

Hyperventilation

Excitement in some children, particularly teenage girls, may precipitate hyperventilaton to the point of losing consciousness. The diagnosis is usually evident in that breathing is excessive and deep, and tetany may also occur. Rebreathing into a paper bag restores the child to normality. If episodes occur frequently, psychological therapy may be required.

Hypoglycaemia and other metabolic conditions

Metabolic disturbance, including hypoglycaemia, may cause loss of consciousness with seizures or a less dramatic alteration in consciousness. An underlying metabolic problem should be suspected in a child if there are features such as developmental delay, dysmorphism, hepatosplenomegaly, or micro- or macrocephaly. Hypoglycaemia may be suspected if there is a temporal relationship of the episode to food.

KEY POINTS

- Most fits, faints and funny turns are benign.
- The history is of paramount importance as the episodes are rarely observed by the doctor.
- The diagnosis is nearly always made on the basis of the history. Physical examination does not often contribute.
- Only carry out investigations if merited by the nature of the episode.

31 Leg pain and limp

Causes of leg pain and limp in childhood

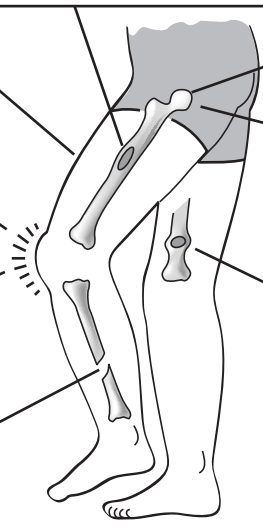
Growing pains
- Preschool children
- Pain often at night, but no limp
- Usually bilateral and located between joints
- Healthy child, no physical signs
- No interference with normal activities

Osteomyelitis
- Fever
- Swelling, erythema, tenderness, decreased movement of the limb
- High ESR, WCC

Diagnosis by X-ray or bone scan

Legg-Perthes disease (avascular necrosis of the femoral head)
- Boys aged 4–10 years
- May follow transient synovitis
- Initially painless
- Pain and limp when fracture occurs
- Diagnosis by X-ray or bone scan

Transient synovitis
- Benign and common in boys aged 2–8 years
- Sudden onset of limp
- No systemic symptoms
- Often preceded by URTI
- Normal investigations and X-ray



Slipped capital femoral epiphysis
- Overweight teenage boys
- Gradual onset of pain in groin or knee
- Diagnosis by X-ray

Septic arthritis (see also p. 77)
- Infant and toddler
- Looks septic
- Swollen hot joint (not obvious in the hip)
- Serious condition

Neoplastic disease
- Benign or malignant
- Pain, tenderness and mass
- Destructive mass on X-ray
- Boring pain in leukaemia

Trauma

What you need from your evaluation

History

- Organic pain tends to be persistent, occurring day and night, interrupts play as well as schooling, is often unilateral or located to a joint in particular
- A limp or refusal to walk is significant
- Weight loss, fever, night sweats, rash or diarrhoea point to organic causes

Physical examination

- Examine the child lying down and walking, and fully examine the leg and groin, not only the knee
- **The limb.** Look for point tenderness, redness, swelling, muscle weakness or atrophy, and limitation of movement for each joint
- ***General examination.*** Look for fever, rash, pallor, lymphadenopathy or organomegaly suggesting infectious or systemic causes

Investigations and their significance

(Only if leg pain is thought to be organic)

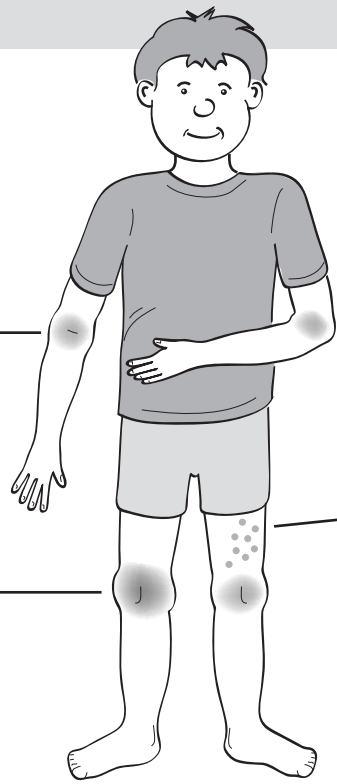
● Full blood count	High white cell count in leukaemia, infections collagen vascular disease
● Plasma viscosity	High in infections, collagen vascular disease Inflammatory bowel disease, tumours
● X-ray	Bone tumours, infection, trauma, avascular necrosis, leukaemia, slipped capital femoral epihphysis
● Bone scan	Osteomyelitis, stress

Key points

- Organic and non-organic causes can be differentiated on clinical grounds. Leg pain alone is usually non-organic
- Important features suggestive of organic disease are a child's refusal to walk, a limp, any physical signs, or constitutional symptoms
- Pain in the hip is referred to the knee, so children presenting with knee pain require a full examination of the leg and groin

32 Swollen joints

Causes of joint swelling



Septic arthritis
- Hot, swollen, acutely tender joint
- Fever may be present
- High white cell count, CRP
- X-ray shows widening of the joint space
- Purulent joint fluid on aspiration

Trauma
- History of injury

Juvenile chronic arthritis (see Chapter 55)
- May be systemic, polyarticular or pauciarticular
- Swelling due to oedema, effusion and synovial thickening
- Child may be irritable with morning stiffness
- Recurrent pattern
- Anaemia and high CRP
- Rheumatoid factor and ANA usually negative

Henoch–Schönlein purpura
- Painful joints +/- swelling
- Purpuric rash on buttocks and thighs

Other causes
- Viral infection
- Leukaemia and other malignancies
- Haemophilia
- Sickle cell disease
- Inflammatory bowel disease
- Psoriasis

What you need from your evaluation

History

- **Joint symptoms:** pain in most conditons is exacerbated by activity, but stiffness alleviated by activity occurs in inflammatory arthropathies
- Are there **systemic symptoms?** Fever, anorexia, weight loss, rash, weakness and fatigue suggest systemic causes
- **Past medical and family history:** important information includes previous arthritis, inflammatory bowel disease, autoimmune conditions, blood dyscrasias and psoriasis

Physical examination

- **Musculoskeletal system:** examine all four limbs and the spine. Look for skin colour changes, heat, tenderness, range of motion and asymmetry
- ***Observe*** the child's gait
- **General examination:** look for anaemia, hepatosplenomegaly, cardiac murmurs and rash

Investigations and their significance

Full blood count	Signs of bacterial infection, anaemia in chronic disease, haemoglobinopathies, malignancy
ESR and plasma viscosity	Elevated in bacterial infection, collagen vascular disease and inflammatory bowel disease
Blood culture	Positive in septic arthritis
ASO titre	High in reactive arthritis or, very rarely, rheumatic fever
Viral titres	Viral arthritis
Rheumatoid factor and antinuclear antibodies	Negative in most forms of juvenile chronic arthritis
X-ray of the joint	Characteristics differ with underlying aetiology
Joint aspiration	Microscopy and culture to exclude or confirm septic arthritis

Key points

- Causes other than traumatic are rare
- If the joint is acutely swollen, rule out septic arthritis as the cause
- Always enquire about systemic symptoms
- Clues to the underlying diagnosis are provided by the history and distribution of the joints involved

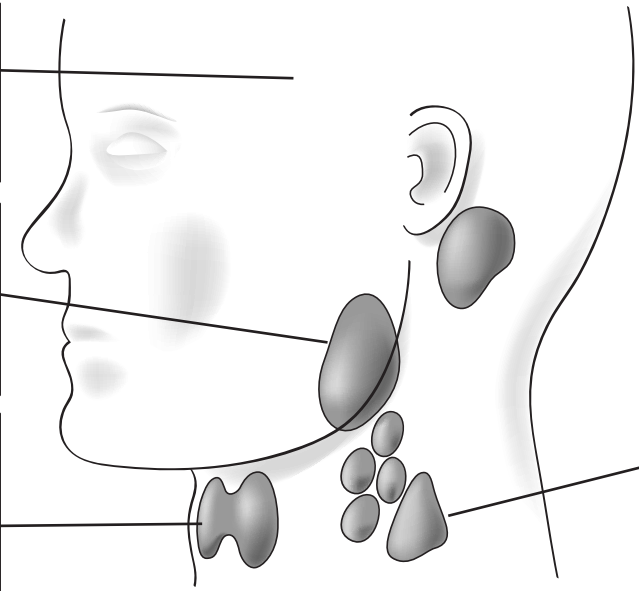
33 Swellings in the neck

Causes of swellings in the neck

Mastoid
Mastoiditis
- Tender inflamed swelling behind ear
- Ear pushed out
- Complication of otitis media
- Medical emergency

Parotid glands
Mumps
- Swelling overlies the angle of the jaw
- Ear displaced up and outward
- Unilateral or bilateral
- Fever and malaise
- Pain on swallowing sweet or sour liquids

Thyroid gland
Thyroiditis
- Anterior midline swelling
- Smooth, diffusely enlarged, non-tender
- Insidious onset
- Signs of hypo-, hyper- or euthyroidism
- Thyroid function tests usually abnormal with elevated thyroid antibodies



Lymph glands
Cervical adenitis
- Tender swollen glands, usually along anterior cervical chain
- Unilateral or bilateral
- Acutely unwell
- Fever, sore throat
- High white cell count

Infectious mononucleosis
- Fever, sore throat
- Large purulent tonsils
- Generalized lymphadenopathy and splenomegaly
- Due to EB virus
- Atypical lymphocytes on blood film

Lymphoma
- Firm, non-tender nodes
- Immobile or matted
- Malaise, night sweats, persistent fever
- Hepatosplenomegaly
- Weight loss

What you need from your evaluation

History
- Ask about malaise and sore throat
- What is the duration of the illness?
- In the case of thyroid swelling, ask about symptoms of hypothyroidism (tiredness, constipation, underachievement at school) or hyperthyroidism (hyperactivity, increased appetite, palpitations, heat intolerance)

Physical examination
- **Identify the site of the swelling:**
  - Lymph nodes usually lie along the anterior cervical chain
  - Parotid glands overlie the angle of the jaw, with displacement of the ear up and out
  - The thyroid is midline anteriorly, and best palpated by standing behind the child
  - The mastoid is behind the ear and pushes the ear out
- **Palpate the gland.** Infected glands are mobile and tender. Malignant glands are fixed and matted
- Look for other sites of infection, e.g. tonsillitis, otitis media
- If the child is acutely unwell, look for signs of dehydration
- If cervical lymphadenopathy is present look for generalized lymphadenopathy and hepatosplenomegaly
- In the case of thyroid swelling, determine if the child is hypothyroid (poor growth, low pulse and BP, delayed tendon reflexes), hyperthyroid (tremor, sweating, fast pulse, high BP, eye signs) or euthyroid

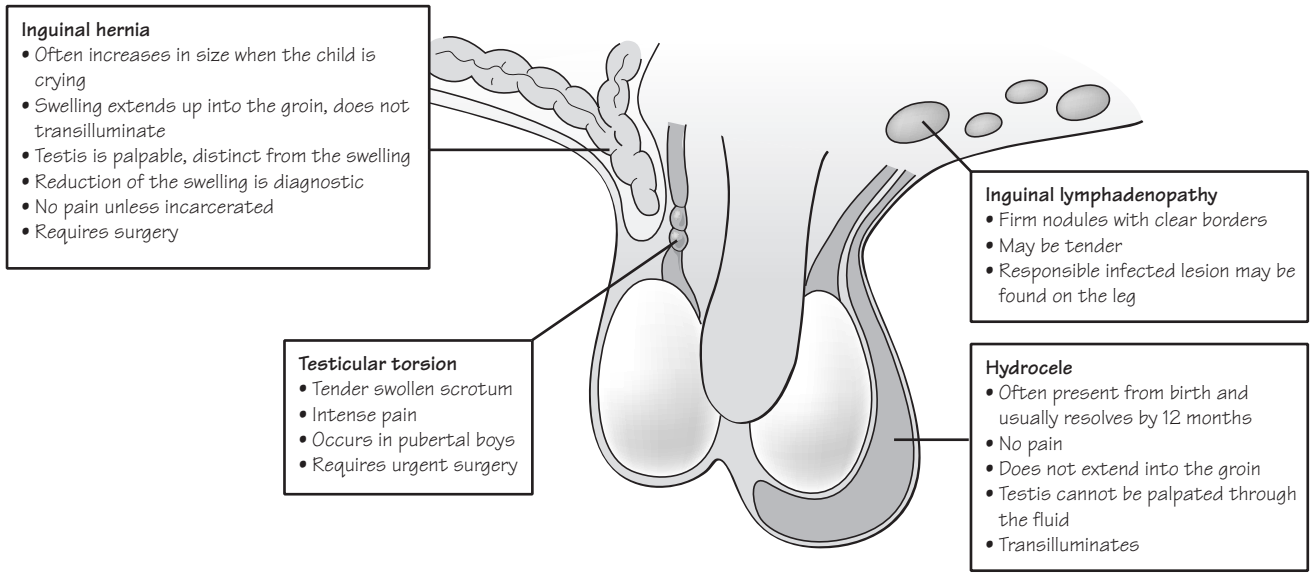
Investigations and their significance

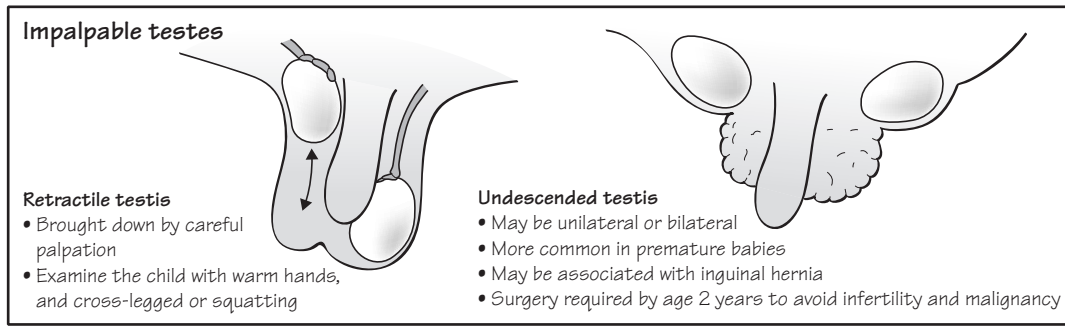
Cervical lymph nodes	FBC	High white cell count in bacterial infection; atypical lymphocytes in infectious mononucleosis
	EB virus screen	Positive in infectious mononucleosis
	Throat culture	Group A haemolytic streptococcal infection needs antibiotics
Parotid glands	Serum or urine amylase	Elevated in mumps, but not usually required for diagnosis
Thyroid gland	Thyroxine TSH	To assess if child is hypo-, hyper- or euthyroid
	Thyroid antibodies	Often positive in thyroiditis
Mastoid process	Tympanocentesis	To identify responsible organism and drain infection

Key points
- Identify the gland involved
- If the process is thought to be infective, assess how sick the child is, and the state of hydration
- If cervical lymphadenopathy is identified, look for generalized lymphadenopathy and hepatosplenomegaly
- If a goitre is found, assess whether the child is hypo-, hyper- or euthyroid
- If mastoiditis is found, admit the child as an emergency

34 Swellings in the groin and scrotum

Swellings in the groin and scrotum and impalpable testes





What you need from your evaluation

History

- **Characteristics of the swelling:** an incarcerated hernia and testicular torsion are both painful. Hernias usually cause intermittent swelling. Hydroceles are often present from birth

Key points

- Incarcerated inguinal hernia and testicular torsion are emergencies
- Hydroceles are present from birth and usually resolve spontaneously
- Undescended testes must be referred by 2 years of age

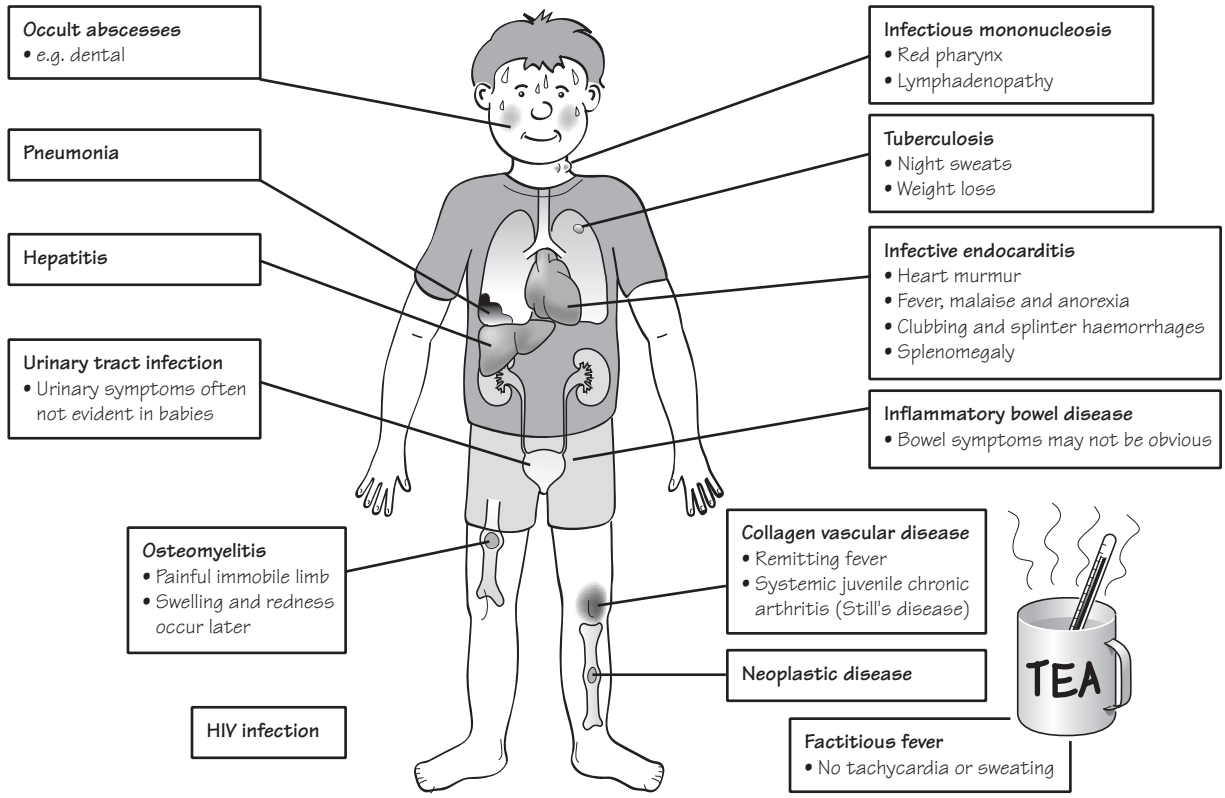
Physical examination

For a swelling

- **Observation:** is the boy in pain? Does the swelling extend into the groin?
- **Palpation:** in an inguinal hernia the swelling extends right up into the groin, and the testis is palpable separate from the swelling. In a hydrocele the testis cannot be palpated through the fluid. Testicular torsion is acutely tender
- Reduction of the swelling by manipulation or spontaneously is diagnostic of a hernia
- **Transillumination:** when a torch is held to the scrotum a hydrocele transilluminates but a hernia does not
- **General examination:** if lymphadenopathy is suspected as a cause, look for an infected lesion on the leg, lymphadenopathy elsewhere, and check for hepatosplenomegaly

35 Pyrexia of unknown origin and serious recurrent infections

Causes of pyrexia of unknown origin



What you need from your evaluation

History

- Review symptoms related to all organ systems
  Ask about:
- -contact with infectious diseases
  -travel
  -exposure to animals

Physical examination

(Repeat physical examinations may be required.)

- **Check the temperature chart.** Repetitive chills (rigors) and temperature spikes suggest septicaemia, abscess, pyelonephritis or endocarditis. There is no tachycardia or sweating in factitious fever
- **Examine the mouth and sinuses.** Oral candida may indicate immune deficiency. A red pharynx may suggest infectious mononucleosis. Tap the sinuses and teeth for tenderness, and transilluminate the sinuses
- **Palpate muscles and bones.** Point tenderness suggests osteomyelitis or neoplastic disease. Generalized muscle tenderness occurs in collagen vascular disease.
- **Heart:** a new murmur or changed murmur may suggest infective endocarditis

Investigations and their significance

• Full blood count	High white cell count in bacterial infection. Very high in leukaemia
• Urinalysis and culture	Occult urinary tract infection
• Examination of blood smear	Parasitic infections, e.g. malaria
• ESR or plasma viscosity	High in bacterial infection Very high in collagen vascular disease, malignancy
• Blood cultures (aerobic and anaerobic)	Bacterial infection Repeat samples needed to diagnose endocarditis, osteomyelitis and occult abscesses
• Liver function tests	Hepatitis
• TB skin test	TB
• X-rays-chest, bones, sinuses, GI tract	Characteristic findings with bacterial infection
• Bone marrow aspirate	Leukaemia, metastatic neoplasms, rare infections
• Serological tests	Infectious mononucleosis, other infections, rarely helpful in collagen vascular disease
• Radioactive scans	Helpful for osteomyelitis, abdominal masses, tumours, abscesses
• Echocardiography	Vegetations seen on heart valves in endocarditis
• Ultrasonography	Identification of intra-abdominal abscesses
• Total body CT or MRI scanning	Detection of neoplasms and abscesses

Pyrexia of unknown origin

Pyrexia of unknown origin (PUO) refers to prolonged fever, defined as more than 1 week in young children and 2–3 weeks in adolescents. In most cases, the diagnosis becomes apparent or the fever resolves within a short period of time. The cause is usually an atypical presentation of a common illness such as urine infection or pneumonia, but more significant causes include endocarditis, collagen vascular diseases, malignancy and inflammatory bowel disease. At times no diagnosis is made, but the fever abates spontaneously.

The child should be hospitalized for careful observation. Antipyretics should not be given as they obscure the pattern of fever, and empirical trials of antibiotics should never be used. Blood cultures (at least three) should be obtained at fever peaks as the yield at that time is higher.

Infective endocarditis

Infective endocarditis occurs as a complication of congenital heart disease, such as ventriculoseptal defect, coarctation, patent ductus arteriosus and aortic stenosis. The commonest causal organism is *Streptococcus viridans* which may be introduced during dental or other surgery: because of this, prophylactic antibiotics are needed to cover any surgery in a child with congenital heart disease.

The child usually presents with fever, malaise and anorexia. Signs include clubbing, splinter haemorrhages in the nails and splenomegaly, and the pre-existing heart murmur may change in character. Microscopic haematuria may be found. The diagnosis is made on blood culture which may need to be repeated on a number of occasions, and echocardiography shows vegetations on the heart valves. Intravenous antibiotics are required for a period of 6 weeks, and serum levels monitored to ensure adequate levels.

Osteomyelitis

Osteomyelitis affects the metaphyses of long bones. The commonest organisms are *Staphylococcus pyogenes*, *Haemophilus influenzae* and *Streptococcus pyogenes*. Although the child may present with PUO, more usually the infected limb is obviously painful and held immobile. Swelling and redness eventually appear, and the adjacent joint may contain a sterile 'sympathetic' effusion. Repeated blood culture determines the causative organism. X-rays are not helpful at presentation, as they take more than 10 days to show changes, but bone scans are useful earlier. The child requires high-dose IV antibiotics for 6 weeks and if there is no immediate response surgical drainage is required. Inadequate treatment leads to bone necrosis, draining sinuses and limb deformity.

Serious recurrent infection

Most children experience recurrent infections. These are commonly respiratory infections which peak when the child starts school or nursery, and despite parental concern they do not require diagnostic exploration. However, recurrent *serious* infections need to be thoroughly evaluated for the underlying cause.

HIV infection and AIDS

Paediatric acquired immunodeficiency syndrome (AIDS) is caused by HIV type 1. The two populations at risk are infants born to infected mothers and adolescents who acquire infection sexually or by IV drugs. Young children usually present by the age of 3 years with features of immunodeficiency: failure to thrive, diarrhoea, candidiasis, hepatosplenomegaly, or severe bacterial infections such as pneumonia, septicaemia, persistent pulmonary infiltrates, *Pneumocystis carinii* pneumonia (PCP), TB and systemic candida.

Diagnosis is made by detection of HIV antibody. Intervention focuses on the use of antiviral drugs, prophylactic antibiotics, viral vaccines and, where necessary, immune serum globulin. The psychosocial and emotional needs of the family must also be addressed. The prognosis is very variable, but in general the earlier and more severe the presentation the worse the prognosis.

Without intervention, 20–30% of babies born to HIV positive mothers become HIV positive themselves, but maternal immunoglobulin G (IgG) may still be measurable up to 18 months in uninfected infants, and so can obscure the diagnosis. Both the administration of zidovudine (azidothymidine, AZT) in pregnancy, and delivery by Caesarian section can reduce transmission to <3%, and the infant should also receive zidovudine for some weeks. In Western countries where the risk of gastroenteritis associated with bottle feeding is low, HIV positive mothers should not breast-feed, as the virus may be transmitted in breast milk.

Splenectomy and hyposplenism

Children who lack an effective spleen are at increased risk of sepsis. Hyposplenism may occur as a result of sickle-cell disease, splenectomy for trauma and some metabolic and haematological conditions. The risk is especially high in children under 5 years old, and penicillin prophylaxis is recommended.

KEY POINTS

- A thorough history and repeat physical examinations are required.

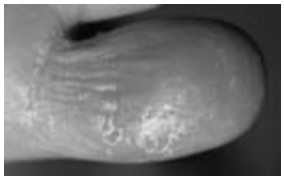
This may save the child from multiple, unpleasant investigations.

- Hospitalization is needed to confirm and observe the pattern of the fever.
- The characteristics of the fever may give a clue to diagnosis.
- Samples for culture should be taken at the peak of the fever.

36 Rashes—types of skin lesions

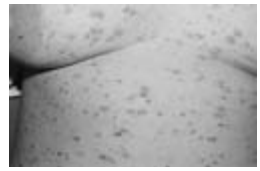
See Plate 1

Desquamation
- Loss of epidermal cells producing scaly eruption
- Examples: post scarlet fever, Kawasaki's disease

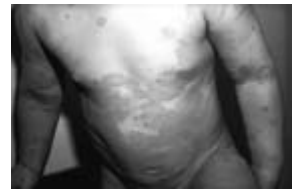


Maculopapular
- Mixture of macules and papules
- Tend to be confluent
- Examples: measles, drug rash

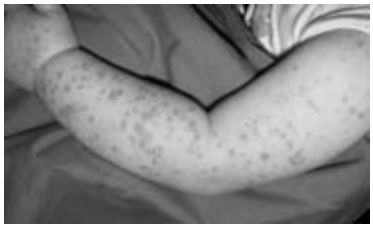
Vesicles
- Raised fluid-filled lesions <0.5 cm in diameter
- Bullae are large vesicles
- Example: chickenpox



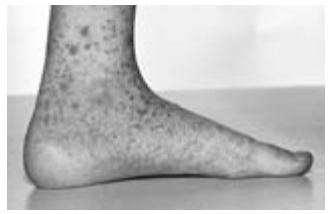
Wheals
- Raised lesions with a flat top and pale centre
- Example: urticaria



Papules
- Solid palpable projections above skin surface
- Example: insect bite



Purpura and petechiae
- Purple lesions caused by small haemorrhages in the skin
- Do not fade on pressure
- Petechiae are tiny purpura
- Examples: meningococcaemia, ITP, HSP, leukaemia



Macules
- Flat pink lesions
- Examples: rubella, roseola, café au lait spot

What you need from your evaluation

History

- Is the child ill or febrile?
- How long has the rash or skin lesion been present?
- Could it be an insect bite or allergic reaction?
- Is it a recurrent problem?
- Is it itchy?
- Has there been contact with anyone else with a rash?

Physical examination

Describe the rash in the following terms:
- Raised or flat
- Crusty or scaly
- Colour
- Blanching on application of pressure
- Size of the lesions
- Distribution (discrete or generalized, or limited to certain sites on the body)

Classify the rash according to the following criteria:
- Acute onset
- Chronic rashes
- Discrete skin lesions
- Birth marks
- Nappy rashes
- Itchy conditions

The following pages will then help you make the diagnosis.

37 Rashes—acute rashes

Rashes of acute onset

Most children presenting with acute onset of a rash and a temperature have a non-specific viral illness. These require only supportive treatment and specific diagnosis is not critical, other than when trying to recognize an epidemic. Maculopapular rashes are often overdiagnosed as measles or rubella, and it is probably preferable to make the diagnosis of viral exanthem if serological testing is not performed. In general, in any of the infectious diseases described below, children are infective during the incubation period—before the rash emerges. If meningococcaemia is suspected intramuscular penicillin must be given as rapid deterioration can occur. The child with idiopathic thrombocytopaenic purpura (ITP) must also be admitted.

What you need from your evaluation

History

- Is the child ill or febrile? Most exanthematous diseases are febrile. In measles and meningococcaemia the child is very ill
- Is the rash itchy? This is a feature of chicken pox and allergic rashes
- Are there associated symptoms? Bleeding occurs in ITP; arthritis and abdominal pain in HSP; and wheezing and stridor rarely with urticaria
- PMH. Exanthems are unlikely to recur (but diagnoses are often inaccurate). Ask about immunizations
- Has there been contact with anyone with an infectious disease?

Physical examination

- Describe the rash: Is it macular, papular, maculopapular, purpuric, petechial, vesicular or wheals? Does it blanch on pressure (purpura and petechiae do not)?
- Distribution: Measles and rubella spread down the body. HSP has a typical distribution
- Is there an enanthem? (rash on mucosal surface) Look for Koplick spots in measles and ulcers in chicken pox
- Carry out a full general examination. Fever and lymphadenopathy are common

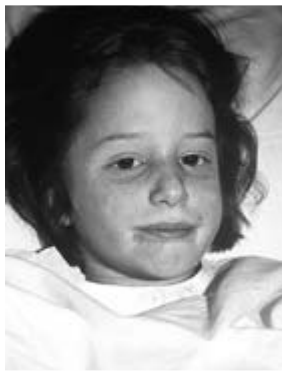
Investigations

- Generally these are not required unless rubella is suspected in a pregnant girl. Cultures are needed in meningococcaemia, and a platelet count in ITP

See Plate 2

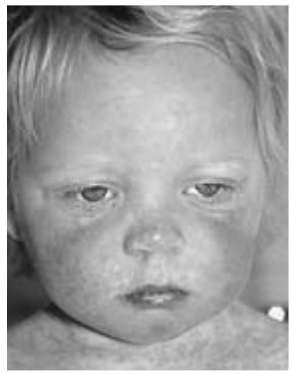
Macular and maculopapular

Scarlet fever



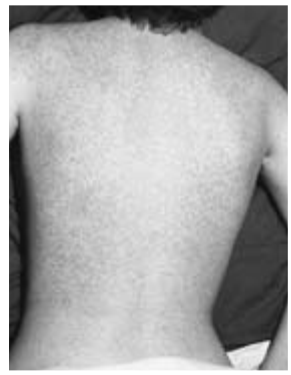
- Fine punctate rash with sandpaper feel, blanches on pressure
- Particularly in neck, axillae and groin
- Fever, headache, sore throat
- Red 'strawberry' appearance of tongue

Measles



- Rash on face and behind ears, spreading down the trunk
- Fever, illness and irritability
- Cough, coryza and conjunctivitis
- Koplick spots in mouth during prodrome
- Lymphadenopathy

Rubella



- Tiny pink macules on face and trunk, rapidly working downwards
- Generally well, with or without fever
- May have lymphadenopathy

Fifth disease

- Mild illness with low-grade fever
- Slapped cheek appearance
- Lace-like rash on body
- Lasts up to 6 weeks

Macular and maculopapular rashes

Measles

Measles is a miserable and very infectious viral illness, characterized by its distinctive rash and the three 'C's'—cough, coryza and conjunctivitis. After an incubation period of 10–14 days there is a prodrome, when Koplick spots appear on the buccal mucosa of the cheeks, looking like grains of salt on a red background, and fever and upper respiratory symptoms develop. The child is ill and irritable. The rash begins on the third or fourth day on the face and behind the ears, and spreads downwards to cover the whole body, beginning to fade after 3–4 days and becomes blotchy. The child is contagious prior to the onset of the rash on the fifth day. Treatment of measles is supportive. Acute otitis media and bronchopneumonia are common complications and require antibiotics. In developing countries there is a high morbidity and mortality, and diarrhoea is also common.

The serious complication of encephalitis occurs rarely, causing drowsiness, vomiting, headache and convulsions. Subacute sclerosing encephalitis (SSPE) is a rare complication which occurs 4–10 years after an attack and is characterized by slow progressive neurological degeneration. High levels of measles antibody are found in the blood and CSF, and the virus antigen has been demonstrated in brain tissue. Immunization with live attenuated vaccine is at age 12–18 months.

Rubella (german measles)

Rubella is usually a mild illness and the rash may not even be noticed. After an incubation period of 14–21 days the rash appears as tiny pink macules on the face and trunk and works its way down the body. The suboccipital lymph nodes are enlarged and there may be generalized lymphadenopathy. Thrombocytopenia, encephalitis and arthritis are rare complications. The rash is non-specific and diagnosis is often erroneously and overconfidently made on clinical grounds.

The importance of rubella lies in the devastating effects it has if contracted during the first trimester of pregnancy. The fetus may die or develop congenital heart disease, mental retardation, deafness and cataracts. For this reason rubella immunization is given in early childhood. If rubella is suspected in pregnancy, titres should be measured immediately and after 10 days to determine if recent infection has occurred.

Scarlet fever

Scarlet fever is the only common childhood maculopapular exanthem caused by bacteria and so requiring antibiotic treatment. It is caused by a strain of Group A haemolytic streptococci.

After an incubation period of 2–4 days fever, headache and tonsillitis appear. The rash develops within 12 h and spreads rapidly over the trunk and neck, with increased density in the neck, axilla and groins. It has a fine punctate erythematous appearance, a 'sandpapery' feel and blanches on pressure. The face is spared, but the cheeks are flushed and there is usually perioral pallor. The tongue initially has a 'white strawberry' appearance, which desquamates leaving a sore 'red strawberry' appearance. The rash lasts about 6 days (less if treated) and is followed by peeling, which is useful in making a retrospective diagnosis.

Penicillin or erythromycin eradicates the organism and may prevent other children from being infected. Sequelae such as rheumatic fever and acute glomerulonephritis are now rare in Western societies.

Vesicular rashes

Chicken pox (varicella)

Chicken pox is a common and highly contagious disease of childhood which is usually mild, except in immunocompromised children. After an incubation period of 14–17 days the rash appears on the trunk and face. The spots occur in crops passing rapidly from macule to papule to vesicle to pustule and then crusting over. Itching is constant and annoying. Vesicles in the mouth rapidly form shallow ulcers. The severity of the disease varies from a few lesions to many hundreds with severe toxicity. The commonest complication is secondary infection of the lesions, and scarring. More severe complications include encephalitis which produces cerebellar signs with ataxia, and thrombocytopenia with haemorrhage. Varicella pneumonia is uncommon in children.

Itching can be reduced by cool baths, calamine lotion and antihistamines. The child is contagious until all the lesions have crusted over. If an immunocompromised child is exposed, prophylaxis with zoster-immune globulin should be considered, and if chicken pox develops, urgent admission for IV acyclovir is indicated. A vaccine against chicken pox may be introduced.

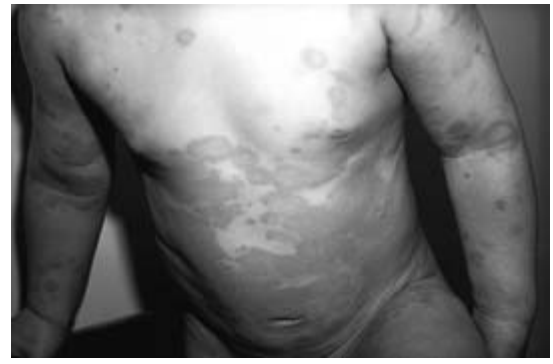
Chicken pox

- Mixture of papules, vesicles, pustules and crusts over trunk and face
- Fever, itching, irritability
- Ulcers in mouth
- May look well or toxic

Wheals

Wheals are a feature of urticarial rashes, usually seen in acute allergic reactions.

Urticaria

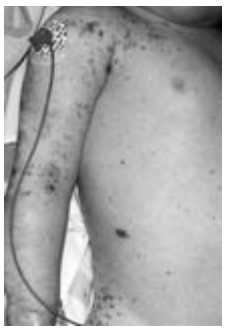


- Well-circumscribed itchy wheals
- Rarely accompanied by wheezing or anaphylactic shock

See Plate 2

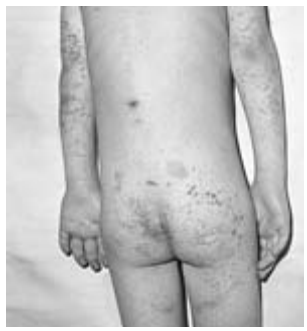
Purpuric rashes

Meningococcal septicaemia



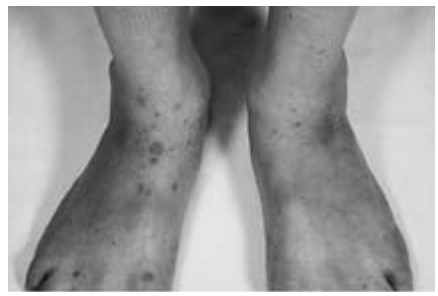
- Petechial, purpuric or morbilliform rash
- May progress rapidly to shock and coma

Henoch–Schönlein purpura (HSP)



- Raised purple lesions, a few millimetres in diameter
- Distributed over thighs, buttocks and legs
- Abdominal pain +/- melaena
- Arthralgia
- Asymptomatic haematuria

Idiopathic thrombocytopenic purpura (ITP)



- Petechial rash over body, with bruising
- Bleeding from other sites, e.g. venepuncture, gums, nose
- Platelet count < 40×10^9

Meningococcal septicaemia

Meningococcal septicaemia is a rapidly life-threatening condition. Within hours of onset of flu-like symptoms, the rash appears with morbilliform, petechial or purpuric characteristics. If the septicaemia is fulminant, the purpura rapidly progress with unrelenting shock and coma. As the consequences of delay are so serious, it must be suspected in any child presenting with purpuria and fever, and intramuscular penicillin given **prior** to transfer to hospital.

Henoch–Schönlein purpura (HSP, anaphylactoid purpura)

This condition is a form of systemic vasculitis, presumed to be due to immune-complex mediated disease. The child presents with a purpuric rash in a typical distribution over the buttocks, thighs and legs. The lesions are purple, raised and a few millimetres in diameter. Arthritis or arthralgia and abdominal pain are commonly experienced and occasionally melaena occurs. The diagnosis is usually made clinically with confirmation of a normal platelet count. Seventy per cent of children develop haematuria and/or proteinuria, but the glomerulonephritis is usually asymptomatic and non-progressive. Treatment of HSP is simply supportive. The rash resolves over a week or two, although microscopic haematuria can persist. Children with renal manifestations need periodic urinary examinations and blood pressure measurements to detect late development of hypertension and renal impairment.

Idiopathic thrombocytopaenic purpura

ITP presents with petechiae and superficial bruising, accompanied at times by bleeding from the gums and nose. It often follows 1–2 weeks after a viral infection. The onset is frequently acute, and the child appears clinically well. The most serious complication is intracranial haemorrhage, which occurs in less than 1% of cases.

Diagnosis is made on finding a platelet count reduced to $<40 \times 10^9$/l and may be below 5×10^9/l. The white cell count is normal and there is no anaemia unless significant blood loss has occurred. As the differential diagnosis includes an aplastic or neoplastic process, bone marrow aspiration is indicated. A normal or increased number of megakaryocytes is seen, reflecting the increased turnover resulting from the peripheral destruction of platelets.

In those who have only mild symptoms no treatment is necessary, but where there is a risk of severe bleeding a short course of steroids may produce a temporary rise in the platelet count. Platelet transfusion is needed if life-threatening haemorrhage occurs. Intravenous gamma globulin causes a sustained rise in the platelet count and may induce remission.

ITP has an excellent prognosis. Severe haemorrhage is usually confined to the initial phase of the disease. In a few children ITP becomes chronic. Splenectomy and immunosuppressive therapy may be required.

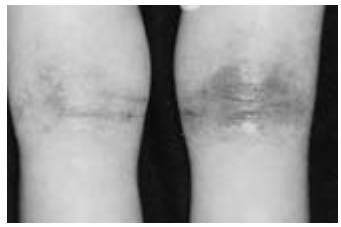
KEY POINTS

- Decide if the rash is macular, papular, maculopapular, purpuric, petechial, vesicular or weals.
- Determine if the child is febrile or ill.
- If the rash is petechial or purpuric and the child is unwell treat with penicillin IM and admit.
- Beware of making a specific diagnosis of measles or rubella clinically. Without serological confirmation 'viral exanthem' should be diagnosed.

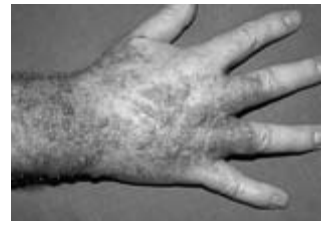
38 Rashes—chronic skin problems

See Plate 3

Atopic dermatitis (eczema)
- Erythema, weepiness and crusting, leading to dry, thickened, scaling skin
- Intense itching
- Often starts in infancy
- Family history of atopy
- Distribution varies with age of the child

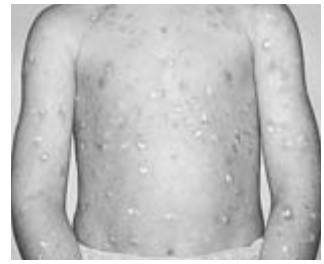


Contact dermatitis
- Erythema and weeping
- Itching
- Caused by irritants such as saliva, detergents and synthetic shoes
- Looks like atopic dermatitis



Seborrhoeic dermatitis
- Dry, scaly and erythematous
- Cradle cap in infancy
- Affects face, neck, axillae and nappy area
- May look like psoriasis

Psoriasis
- Plaques of thick, silvery or white scales with demarcated borders
- On scalp, knees, elbows and genitalia
- Pitting of the nail plate



What you need from your evaluation

History
- Is the rash itchy? This is characteristic of atopic and contact dermatitis
- Are there precipitating factors? Milk, wheat and eggs may precipitate or exacerbate atopic dermatitis. Enquire about irritants causing contact dermatitis
- Is there a family history? Children with eczema often have a family history of atopy. Psoriasis is often familial

Physical examination
- Characteristics of the rash. Fully undress the child to observe the distribution. Note the changing pattern of atopic dermatitis with age
- Other features. Cradle cap and rash behind the ears suggests seborrhoeic dermatitis. Nail pitting and arthritis point towards psoriasis

Most chronic skin conditions in childhood are eczematous. Acute eczema is characterized by erythema, weeping and microvesicle formation within the epidermis. Chronic eczema is characterized by thickened, dry, scaly, coarse skin (lichenification). The commonest type of eczema in children is atopic dermatitis, although contact dermatitis and seborrhoeic dermatitis are also relatively common. Topical corticosteroids form an important part of the management of a variety of chronic skin conditions. They must be used with care as long-term use, particularly of the fluorinated variety, leads to atrophy of the skin and increased hair growth in some. Small amounts of cream applied frequently is more effective than large amounts infrequently. The more potent topical steroids should not be applied to the face, and if applied over the body using occlusive dressings, systemic absorption with adrenal suppression can occur.

Atopic dermatitis

Atopic dermatitis is an inflammatory skin condition characterized by erythema, oedema, intense itching, exudation, crusting and scaling. It often begins at 2–3 months, the onset coinciding with the introduction of cow's milk, wheat and eggs into the diet. There is often a family history of atopy, and allergic rhinitis and asthma often develop later. Genetically susceptible infants may be protected from developing eczema if they are exclusively breast-fed.

In *infancy* the lesions are erythematous, weepy patches on the cheeks which subsequently extend to the rest of the face, neck, wrists, hands and extensor surfaces of the extremities. Pruritus is marked and scratching leads to weeping, crusting, and commonly secondary infection.

By *3–5 years* there is a tendency towards remission, although mild to moderate dermatitis may persist in the popliteal and antecubital fossae, on the wrists, behind the ears and on the face and neck.

During *school years* recurrence tends to occur with antecubital and popliteal involvement, extension to the neck, forehead, eyelids, wrists and dorsa of the hands and feet. The skin becomes dry and thickened and the face can take on a whitish hue. Hyperpigmentation, scaling and lichenification become prominent.

The diagnosis is clinical. Total and allergen-specific serum

immunoglobulin E (IgE) levels are often raised and eosinophilia may be present. Although skin testing is frequently positive it is rarely helpful clinically.

Treatment is directed at trying to interrupt the itch–scratch–itch cycle. During an acute flare-up wet dressings are helpful, with topical steroids applied between dressing changes. Antihistamines can be useful for their sedative and antipruritic effect. Topical or oral antibiotics may be required for secondary infection.

After the acute phase, topical steroids are applied in the form of creams or ointments. The more potent steroid creams must be kept to a minimum and should not be applied to the face. Systemic corticosteroids are only rarely used.

Lubricants are used after application of steroid creams and continued on a prophylactic basis to keep the skin moist. Bath oils can be added to the bath water, so that moisture is sealed into the well-hydrated skin. Dietary restriction is controversial and generally of limited value. Arbitrary exclusion of a number of foods can lead to malnutrition.

The course of atopic dermatitis is fluctuating and fortunately resolves entirely in some 50% of infants by the age of 2 years. A few continue to be problematic beyond childhood. Reasonable control of this chronic condition can usually be achieved in most children.

Contact dermatitis

Clinically, contact dermatitis may be indistinguishable from atopic dermatitis. It can be due to irritants or allergens and results from prolonged or repetitive contact with substances such as saliva, citrus juices, detergents, occlusive synthetic shoes, topical medication and jewellery. In general, contact dermatitis clears on removal of the irritant or allergen and temporary treatment with a topical corticosteroid preparation.

Seborrhoeic dermatitis

Seborrhoeic dermatitis is a chronic inflammatory condition which is commonest during infancy and adolescence. Cradle cap, a diffuse or focal scaling and yellow crusting of the scalp is the commonest manifestation. A dry scaly erythematous dermatitis may also involve the face, neck, axillae, nappy area and behind the ears. If the scaling is prominent it may look like psoriasis, and red scaly plaques may appear. Itching may or may not be present.

Scalp lesions are usually controlled with antiseborrhoeic shampoo. Inflamed lesions respond to topical corticosteroid therapy. Secondary bacterial infections and superimposed candidiasis are not uncommon.

Psoriasis

Psoriasis is a common chronic skin disorder among adults, one-third of whom become affected during childhood. There is usually a family history. The lesions consist of erythematous papules which coalesce to form plaques of thick silvery or white scales with sharply demarcated borders. They tend to occur on the scalp, knees, elbows, umbilicus and genitalia. Nail involvement, a valuable diagnostic sign, is characterized by pitting of the nail plate. Guttate psoriasis is a variant where multiple small oval or round lesions appear over the body, often following a recent streptococcal infection.

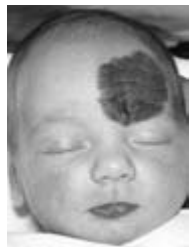
Therapy is mainly palliative. The application of coal tar preparations after a bath is helpful. Salicylic acid ointment is useful in removing scale, but extensive application can result in salicylate poisoning particularly in young children. Topical corticosteroids are effective but must be used with caution. New treatments include UVB light and combinations of steroids and Vitamin D.

39 Rashes—discrete skin lesions

See Plates 3 and 4

Common birthmarks

Pigmented naevus
- Appears at age 2 years
- If large, at risk of malignant change



Strawberry naevus
- Bright red protuberant lesion
- Usually enlarges for 1–2 years, then regresses
- Resolves spontaneously without treatment

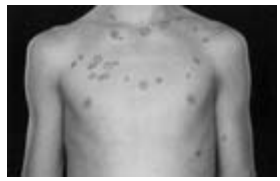
Stork mark (naevus flammeus)
- Occurs on eyelids, neck or forehead
- Fades spontaneously

Portwine stain
- Sharply circumscribed, pink to purple lesion
- Consists of mature, dilated, dermal capillaries
- May be a sign of Sturge–Weber syndrome with an underlying meningeal haemangioma, intracranial calcification and fits

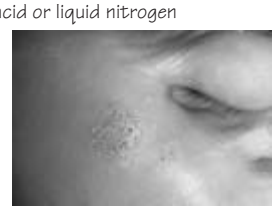
Mongolian spots
- Blue/grey lesions in the sacral area
- Particularly common in black babies
- Fade during the early years

Infectious lesions

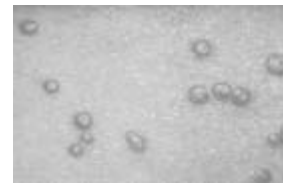
Tinea corporis (ringworm)
- Dry, scaly papule which spreads centrifugally with central clearing
- Diagnosis confirmed microscopically by scrapings in a potassium hydroxide wet mount
- Treat with topical antifungal agents for 2–4 weeks



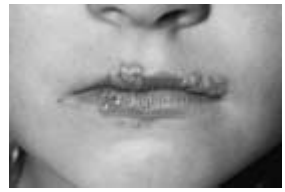
Common warts
- Roughened keratotic lesions with an irregular surface
- Occur on hands, face, knees and elbows
- Called verrucas if present on feet
- Transferred by direct contact
- Disappear spontaneously, but can be treated with salicylic acid or liquid nitrogen



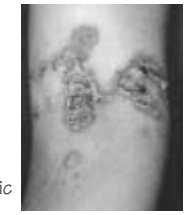
Molluscum contagiosum
- Pearly dome-shaped papules with central umbilicus
- Particularly on face, axillae, neck and thighs
- Self-limited disease



Cold sore
- Single or grouped vesicles or pustules sited periorally
- Recurrent herpes simplex infection
- Recur with colds and stress
- May be treated with acyclovir



Impetigo
- Sticky, heaped-up, honey-coloured crusts
- Group A haemolytic streptococci or staphylococci
- Contagious
- Treat with antibiotics (erythromycin orally, or antibiotic cream if <5 lesions)

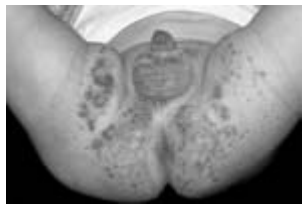


40 Rashes—nappy rashes and itchy lesions

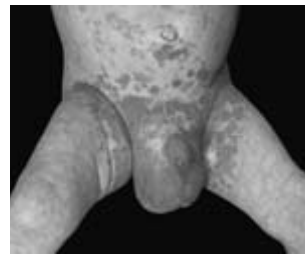
See Plate 4

Nappy rashes

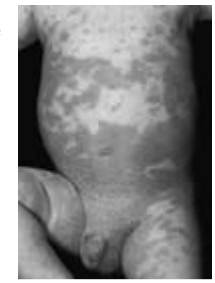
Ammoniacal dermatitis
- Erythematous and/or papulovesicular lesions, fissures and erosions
- Skin folds spared
- Caused by irritation from excretions and chemicals
- Secondary bacterial and candidal infection common, and limited use of hydrocortisone and nystatin cream. Treat by regular washing and changing, exposure to air, and use of protective creams



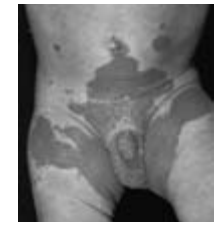
Candidal nappy rash
- Bright red rash with clearly demarcated edge
- Satellite lesions beyond border
- Inguinal folds usually involved
- May have oral thrush (white curds in mouth)
- Treatment with nystatin cream, and orally if necessary



Seborrhoeic nappy rash
- Pink, greasy lesions with yellow scale
- Often in skin folds
- Cradle cap may be present
- Treat with mild topical corticosteriods



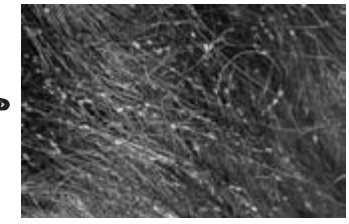
Psoriatic nappy rash
- Appearance similar to seborrhoeic dermatitis
- Family history of psoriasis



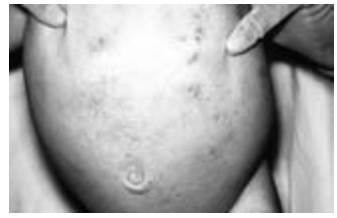
Itchy lesions

Conditions covered elsewhere
- Atopic dermatitis (p. 86)
- Contact dermatitis (p. 87)
- Urticaria (p. 84)
- Chicken pox (p. 84)

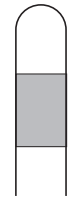
Head lice (Paediculosis capitis)
- Itchy scalp
- Nits (the eggs) are visible as white specks on hair shafts
- Transmitted on clothing, combs or by direct contact
- Treated by combing and use of anti-pediculosis shampoo



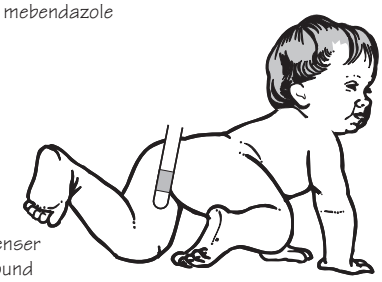
Scabies
- Wheals, papules, vesicles with superimposed eczema
- Intensely pruritic
- Characteristic lesion is the mite burrow between the fingers
- Head, neck, palms and soles are spared in children but not babies
- Mites can be seen on scrapings
- Treat all the household with scabicides and launder bedding



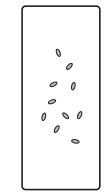
Threadworms
- Anal (and also vulvar) itching
- Diagnosis made by seeing tiny worms or identifying the eggs microscopically using sticky tape
- Treated with mebendazole



Tongue dispenser with tape wound round sticky-side-out



Press against anus on waking



Press against glass slide. Eggs can be visualized microscopically

41 Short stature and poor growth

Steady growth below centiles

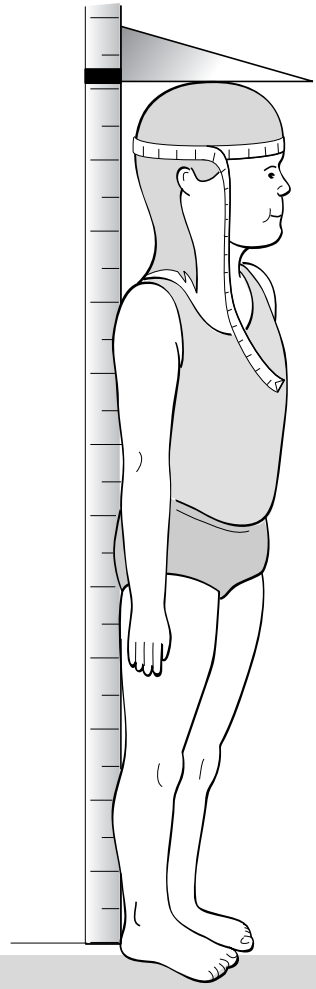
Constitutional (familial) short stature
- Short parents
- Normal history and examination
- No delay in bone age

Maturational delay
- Delayed onset of puberty
- Family history of delay
- Delayed bone age

Turner's syndrome
- Features of Turner's syndrome (not always present)
- XO karyotype
- No pubertal signs
- No delay in bone age

IUGR
- Low birth weight
- The underlying reason for the IUGR may be evident

Skeletal dysplasias (rare)
- Body disproportion with shortened limbs
- Achondroplasia is the commonest form



Fall-off in growth across centiles

Chronic illness
- Usually identifiable on history and physical examination
- Crohn's disease and chronic renal failure may be occult
- Some delay in bone age occurs

Acquired hypothyroidism
- Clinical features of hypothyroidism
- Goitre may be present
- Low T4, high TSH and thyroid antibodies
- Delayed bone age

Cushings' disease (rare)
- Usually iatrogenic due to prescribed steroids
- Cushingoid features
- Delayed bone age

Growth hormone deficiency (rare)
- Congenital or acquired
- May occur with other hormone deficiencies
- Delayed bone age

Psychosocial
- Neglected appearance
- Behavioural problems
- Catch-up growth occurs when child is removed from home

What you need from your evaluation

History

- **Medical history and review of systems.** Identify any chronic condition, such as asthma, arthritis or diabetes, which can affect growth. Ask about symptoms of raised intracranial pressure, malabsorption and hypothyroidism. Long-term steroid administration stunts growth
- **Family history.** Compare the child's growth with parental heights. It normally lies on the centile between parents' height centiles. Late maternal menarche suggests familial maturational delay
- **Birth history.** A child born small for gestational age may have reduced growth potential. Enquire too about perinatal problems
- **Psychosocial history.** Emotional neglect and abuse can stunt growth but also ascertain whether there are social or emotional difficulties resulting from short stature

Physical examination

- **Pattern of growth.** Obtain previous growth measurements from the GP or child health service. A fall-off in growth suggests a medical condition requiring treatment
- **Anthropometric measures.** Obtain accurate measures of length (to 24 months of age) or height, and weight. Plot on a growth chart
- **General examination.** Look for signs of hypothyroidism, body disproportion, stigmata of Turner's syndrome and dysmorphism. Each organ system should be examined for evidence of occult disease

Investigations and their significance

If a decrease in growth velocity has occurred, investigations are always required.

Blood count and plasma viscosity	Inflammatory bowel disease
Urea and electrolytes	Chronic renal failure
Coeliac antibodies	Screening test for coeliac disease
Thyroxine and TSH	Hypothyroidism
Karyotype (in girls)	Turner's syndrome
Growth hormone tests	Hypopituitarism, growth hormone deficiency
X-ray of the wrist for bone age	Delayed bone age suggests maturational delay, hypothyroidism, GH deficiency or corticosteroid excess. A prediction of adult height can be made from it

Short stature usually has a physiological basis and is due to a reduced genetic potential or maturational delay (slow physical development). Fall-off in growth is much more concerning as it suggests an organic problem. Short stature can cause social difficulties, particularly in adolescence for boys, and occasionally psychological counseling is required.

Constitutional or familial short stature

Short parents tend to have short children. In this case the history and physical examination are normal, and the bone age appropriate for age. Reassurance is often all that is required. The use of growth hormone in children with physiological short stature is controversial and probably has little effect on the child's final adult height.

Maturational delay

Children with maturational delay are often called 'late developers' or 'late bloomers'. The child is short and reaches puberty late. The final height depends on the genetic constitution, and may be normal. There is often a family history of delayed puberty and menarche, and the bone age is delayed. Most families simply require reassurance that final height will not be so affected. Sometimes, teenage boys find the social pressures to be so great that it is helpful to trigger puberty early using low doses of testosterone, so causing an early growth spurt. This treatment does not have an effect on final height.

Hypothyroidism

Hypothyroidism may result from Hashimoto's autoimmune thyroiditis which is commoner in girls, or may be congenital. A lack of thyroid hormone has a profound effect on growth, and short stature is often the presenting sign. Other features include a fall-off in school performance, constipation, dry skin and delayed puberty. Low thyroxine (T_4) and high thyroid stimulating hormone (TSH) levels are found on investigation with antithyroid antibodies if the cause is autoimmune. Treatment consists of lifelong replacement with thyroid hormone. Parents are often alarmed when their placid, hypothyroid child is transformed into a normal, active teenager. The prognosis is good.

Rarer hormonal problems

Although Cushing syndrome and disease are extremely rare in childhood, growth suppression from exogenous steroids is not uncommon. In children requiring longterm high steroid therapy, the deleterious effects on growth can often be reduced by giving the steroids on alternate days.

Growth hormone deficiency is a rare cause of short stature. It may occur secondary to lesions of the pituitary such as tumours or cranial irradiation. It can be isolated, or accompanied by deficiency of other pituitary hormones. The diagnosis is confirmed by growth hormone testing. Brain imaging is needed to identify any underlying pathology. Treatment involves daily subcutaneous injections of synthetic growth hormone.

Chronic illness

Any chronic illness can lead to stunting of growth. However, chronic illnesses rarely present as short stature as the features of the illness are usually all-too evident. Chronic conditions that may present with poor growth, in advance of other clinical features, include inflammatory bowel disease, coeliac disease, and chronic renal failure.

Turner's syndrome

Turner's syndrome or gonadal dysgenesis is an important cause of short stature and delayed puberty in girls, caused by the absence of one X chromosome, although mosaicism also occurs. The gonads are merely streaks of fibrous tissue.

As neonates, Turner's babies often have marked webbing of the neck and lymphoedematous hands and feet. In childhood, short stature is marked and the classic features of webbing of the neck, shield shaped chest, wide-spaced nipples and a wide carrying angle may be evident. Some girls are only diagnosed in adolescence when puberty fails to occur. Growth can be promoted by small doses of growth hormone and oestrogen in childhood. Puberty has to be initiated and maintained by oestrogen therapy. Despite treatment, in adult life women with Turner's syndrome are usually short. Recent advances in infertility treatment have resulted in a few women becoming pregnant through *in vitro* fertilization (IVF) with donated ova.

KEY POINTS

- A good history and physical examination will identify most pathological causes of short stature.
- Focus on looking for evidence of intracranial pathology, hormone deficiency, chronic illness and GI symptoms.
- The child's height must be related to the parents' heights.
- Emotional and social consequences of the short stature should be identified.

42 Failure to thrive (weight faltering)

Causes of failure to thrive

(Note that babies may normally cross centiles in the first year)

Environmental/psychosocial (non-organic)
- Commonest cause of failure to thrive (FTT)
- Weight is usually affected first, then height and head growth
- Eating difficulties are common
- Disturbed maternal–infant interaction may be present
- Maternal depression/psychiatric disorder may be present
- Neglect may be a factor

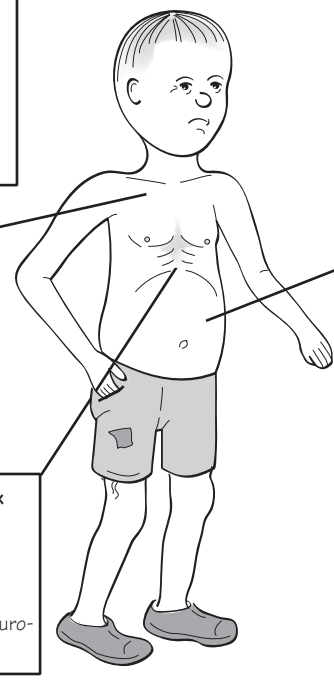
IUGR
- Low birth weight
- If birth length and head size were also small, catch-up is less likely
- May have features of TORCH
- May be due to a genetic syndrome

Immunodeficiency (rare)
- Recurrent infections
- HIV, severe combined immunodeficiency are causes

Cystic fibrosis (see Chapter 54)
- Diarrhoea
- Chest infections

Gastro-oesophageal reflux (see p. 53)
- Pain from oesophagitis
- Apnoea
- Vomiting/possetting
- Common in babies with neurodevelopmental problems

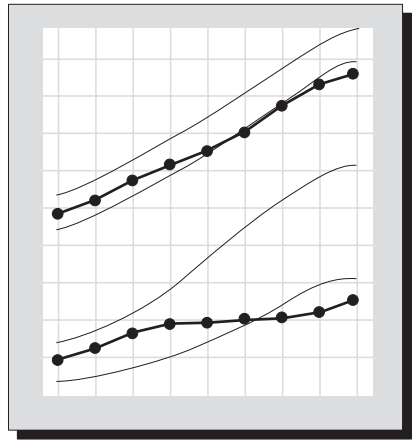
Endocrine dysfunction
- Developmental delay in hypothyroidism
- Growth hormone deficiency very rare



Genetic syndromes
- Low birth weight common
- Dysmorphic features

Chronic illness
- Very rarely an occult cause
- Features depend on the illness

Coeliac disease (see p. 63)
- Weight falls off when gluten is introduced
- Diarrhoea
- Irritability



What you need from your evaluation

History

- **Nutritional history.** Take a dietary history (a food diary can be helpful). Ask about feeding difficulties: did they start at birth, weaning or as a toddler? Consider whether they are a result or cause of FTT
- **Review of symptoms.** A good history identifies most organic conditions. Look for diarrhoea, colic, vomiting, irritability, fatigue or chronic cough
- **Developmental history.** Are there neurodevelopmental problems? Has FTT affected the baby's developmental progress?
- **Past medical history.** Low birth weight and prenatal problems may jeopardize growth potential. Recurrent or chronic illness may affect growth
- **Family history.** Is there a family history of FTT or genetic problems? Are there psychosocial problems?

Examination

- **General observations.** Does the child look neglected, ill or malnourished (thin, wasted buttocks, a protuberant abdomen and sparse hair)? How does the mother relate to the baby?
- **Growth.** Plot growth on a chart (remember to correct for prematurity!)
- **Physical examination.** Look for signs of chronic illness

Investigations

'Fishing' for a diagnosis by carrying out multiple investigations is futile. Obtaining a blood count and ferritin level is useful as iron deficiency is common and affects development and appetite. Otherwise, investigations should be based on clinical findings

Investigations and their significance

• Full blood count, ferritin	Iron deficiency is common in FTT and can cause anorexia
• Urea and electrolytes	Unsuspected renal failure
• Stool for chymotrypsin and fat globules	Low chymotrypsin and the presence of fat globules suggest malabsorption
• Coeliac antibodies, jejunal biopsy, sweat test	Coeliac disease and cystic fibrosis are the most important causes of malabsorption
• Thyroid hormone and TSH	Congenital hypothyroidism causes poor growth and developmental delay
• Karyotype	Chromosomal abnormalities are often associated with short stature and dysmorphism
• Hospitalization	Hospitalization can be a form of investigation. Observation of baby and mother over time can provide clues to the aetiology

The term 'failure to thrive' (FTT) implies not only growth failure, but also failure of emotional and developmental progress, and usually refers to babies or toddlers. The commonest causes of FTT are non-organic.

Concern about growth usually is raised when:

- Weight is under the 2nd centile.
- Height is below the 2nd centile.
- Or, when height or weight cross down two centiles.

However, as normal babies commonly cross centiles in the first 2 years, expertise is needed to differentiate this growth or weight faltering from failure to thrive.

It is very distressing when a young child fails to thrive and the evaluation needs to be carried out sensitively. The purpose of the evaluation is to differentiate the child with a problem from those showing normal growth faltering, and then to identify the contributing factors whether organic or non-organic (which may coexist). It is important that a normal, healthy but small baby is not wrongly labelled as having a problem.

Investigations need to be requested judiciously.

FTT due to environmental or psychosocial causes (non-organic FTT)

Psychosocial problems are the commonest cause of FTT. Problems include difficulties in the home, limitations in the parents, disturbed attachment between mother and child, maternal depression or psychiatric disorder and eating difficulties. Uncommonly, neglect is a factor.

Weight gain is usually first affected, followed in some by a fall in length and head circumference. The child's developmental progress may also be delayed. Children with FTT range across a spectrum. At one end there is the caring home where parents are anxious and concerned. The problem is often one of eating difficulties, where meals are very stressful and parents do their utmost (often counterproductively) to persuade the child to eat. At the other end of the spectrum is the neglected child who shows physical signs of poor care and emotional attachment. In this case the problem is often denied and compliance with intervention poor.

Management must suit the underlying problem. The family health visitor should be involved for nutritional advice and help with eating problems. Occasionally it is helpful to admit the baby to hospital for observation. Practical support can ease the stress, and nursery placement can be very helpful as well as helping to resolve eating difficulties. In those cases where neglect is the cause and the family are not amenable to help, social services must be involved. A minority of children need to be removed from their homes.

Malabsorption

Malabsorption is an important cause of failure to thrive. Symptoms of diarrhoea and colic are usually present as diagnostic clues. The commonest causes of malabsorption are coeliac disease and cystic fibrosis. In the former, the growth curve characteristically shows fall-off in weight coincident with the introduction of gluten to the diet.

Chronic illness

Children and babies with any chronic illness not uncommonly grow poorly. They rarely present as a diagnostic dilemma as the manifestations of the disease are usually evident. However, organic FTT may be compounded by psychosocial difficulties and these need to be addressed. Very rarely, chronic disease can be occult and present as FTT.

Genetic causes

Small parents tend to have small children and the small healthy normal child of short parents should not arouse concern. Usually in this case growth is steady along the lower centiles, but the large baby born to small parents may cross down centile lines before settling onto the destined line.

Genetic syndromes are not uncommonly associated with short stature. If dysmorphic features are present the diagnosis can be suspected. Intrauterine growth retardation results from adverse uterine conditions which may affect infant growth. When this occurs early in gestation, length and head circumference as well as weight may be affected, and growth potential may be jeopardized. The cause of the intrauterine growth retardation should, where possible, be identified.

KEY POINTS

- Be sensitive. It can be very distressing if a baby has FTT.
- Differentiate the baby who is failing to thrive from the normal baby who is crossing centiles.
- Identify any symptoms and signs suggestive of organic conditions.
- Only perform laboratory investigations if there are clinical leads in the history and physical examination.
- Identify psychosocial problems that are affecting the baby's growth and provide appropriate help and support.

43 Heart murmurs

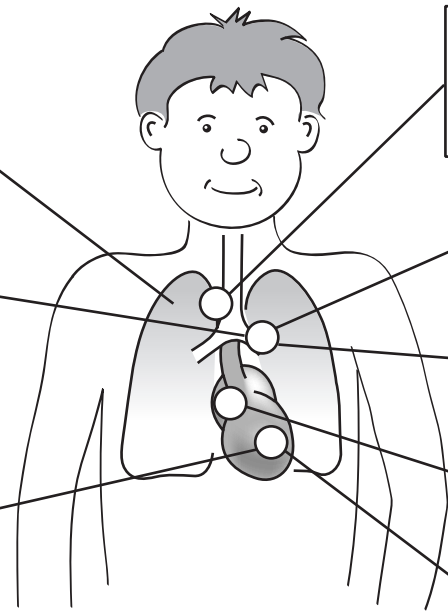
Causes of heart murmers

Innocent murmurs
- Have no clinical significance
- Are systolic and musical
- Do not radiate
- Vary with posture and position

Venous hum
- Blowing continuous murmur in systole and diastole
- Heard below the clavicles
- Disappears on lying down

Pulmonary flow murmur
- Brief high-pitched murmur at second left intercostal space
- Best heard with child lying down

Systolic ejection murmur
- Short systolic murmur at left sternal edge or apex
- Musical sound
- Changes with child's position
- Intensified by fever, exercise and emotion



NB: Patent ductus arteriosus and tetralogy of Fallot are discussed in Chapter 51

Pathological murmurs
- Are pansystolic or diastolic
- Are harsh or long
- May radiate and have a thrill
- May involve cardiac symptoms

Aortic stenosis
- Soft systolic ejection murmur at right upper sternal border
- Radiates to neck and down left sternal border
- Causes dizziness and loss of consciousness in older children

Atrial septal defect
- Soft systolic murmur at second left intercostal space
- Wide fixed splitting of the second sound
- May first be detected at school entry

Pulmonary stenosis
- Short systolic ejection murmur in upper left chest
- Conducted to back
- Preceded by ejection click
- Thrill in the pulmonary area

Ventricular septal defect
- Harsh pansystolic murmur at lower left sternal border
- Radiates all over chest
- Signs of heart failure may be present

Coarctation of the aorta (see p. 111)
- Systolic murmur on left side of chest
- Heard in the back
- Absent or delayed femoral pulses

What you need from your evaluation

History
- Fatigue is the most important symptom of cardiac failure. A baby in cardiac failure can take only small volumes of milk, becomes short of breath on sucking, and often perspires. The older child tires on walking and may become breathless too
- Take a family history. The risk of heart defects is higher in siblings of children with congenital heart disease

Physical examination
- **Murmur.** The quality of the sound and the site where it is heard indicates if it is pathological. Listen for radiation over the precardium, back and neck, with the child both sitting and lying
- **Signs of heart failure:** Look for: failure to thrive and poor growth, tachycardia and tachypnoea, hepatomegaly and crepitations (peripheral oedema is rare in children)
- **Pulse and blood pressure:** Remember that femoral pulses are weak, delayed or absent in coarctation of the aorta. Blood pressure will be higher in the arms than the legs
- **Cyanosis:** is an unlikely finding in children presenting with a heart murmur

Investigations and their significance
These are required only if the murmur is thought to be pathological.
- **Chest X-ray.** Provides information about cardiac size and shape, and pulmonary vascularity
- **ECG.** Provides information about ventricular or atrial hypertrophy
- **Echocardiography.** Evaluates cardiac structure and performance, gradients across stenotic valves and the direction of flow across a shunt
- **Cardiac catheterization.** Rarely required for diagnosis

Heart murmurs are very common in young children. Most are 'functional' or 'innocent' and are not associated with haemodynamic abnormalities. It is important to learn to distinguish these from murmurs associated with cardiac disease. Good management includes the following: Discuss the benign nature of the murmur with the parents. It is helpful to describe it as a simple 'noise' which itself does not indicate a cardiac defect. No investigations are required and they need to be fully reassured.

Defects causing a left-to-right shunt

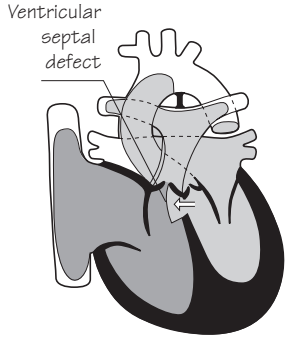
These are the commonest defects. If large, a considerable volume is shunted, causing hypertrophy, dilatation and failure.

Ventricular septal defect

This is the commonest congenital heart lesion. If the defect is small the child is asymptomatic but a large shunt causes breathlessness on feeding and crying, FTT and recurrent chest infections. A harsh pansystolic murmur is heard at the lower left sternal border, and in large defects the heart is enlarged, a thrill is present and the murmur radiates over the whole chest. There may be signs of heart failure. Loudness of the murmur is not related to the size of the shunt.

In large defects cardiomegaly and large pulmonary arteries are seen on X-ray and biventricular hypertrophy on ECG. Echocardiography confirms the diagnosis.

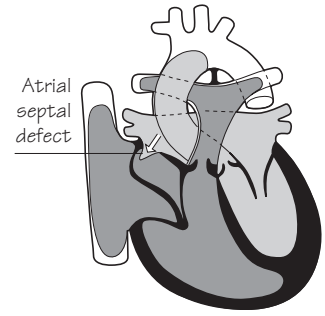
Small defects usually close spontaneously. Large defects with cardiac failure are initially managed medically, but surgical treatment may be required. If uncorrected, pulmonary hypertension can result from the increased blood flow, with reversal of the shunt and cyanosis. Antibiotic prophylaxis is needed in any child with a VSD.



Ventricular septal defect
Large flow causes cardiomegaly and prominent pulmonary arteries

Atrial septal defect

As the murmur is soft, it may not be detected until the child starts school. The systolic murmur is heard in the second left interspace, and is due to high flow across the normal pulmonary valve (and not due to flow across the defect). The second heart sound is widely split and 'fixed' (does not vary with respiration). The child may experience breathlessness, tiredness on exertion or recurrent chest infections. If the defect is moderate or large, closure is carried out at open heart surgery and has a good prognosis. If untreated, cardiac symptoms usually develop in the third decade of life or later.



Atrial septal defect
High flow through pulmonary valve causes a systolic murmur

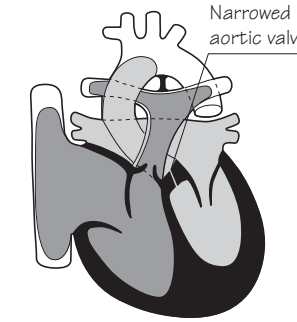
Obstructive lesions

Obstructive lesions can occur at the pulmonary and aortic valves and along the aorta. The chamber of the heart proximal to the lesion hypertrophies, and heart failure may develop.

Aortic stenosis

Aortic stenosis is usually identified at routine examination, but some older children may experience faintness or dizziness on exertion. The systolic ejection murmur is heard at the right upper sternal border and radiates to the neck and down the left sternal border. It may be preceded by an ejection click and the aortic second sound is soft and delayed. The peripheral pulse is of small volume and blood pressure may be low. A thrill may be palpable at the lower left sternal border and over the carotid arteries.

Chest X-ray may show a prominent left ventricle and ascending aorta. Left ventricular hypertrophy is found on ECG. Echocardiography can evaluate the exact site and severity of the obstruction.

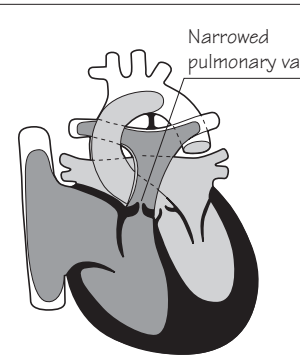


Aortic stenosis
The stenosis causes enlargement of the left ventricle and prominence of the ascending aorta

Severe stenosis is relieved by balloon valvuloplasty—a catheter is passed from the femoral artery and a balloon inflated to widen the stenosis. If unsuccessful, open heart surgery is required. Reoperation is often required later. Children with aortic stenosis are at risk for sudden death and so this is one defect in which strenuous activity should be avoided. Infective endocarditis prophylaxis is required.

Pulmonary stenosis

The pulmonary valve is narrowed and the right ventricle hypertrophied. A short ejection systolic murmur is heard over the upper left anterior chest and is conducted to the back. It is usually preceded by an ejection click. With mild stenosis there are usually no symptoms. In severe stenosis a systolic thrill is palpable in the pulmonary area. On chest X-ray dilatation of the pulmonary artery is seen beyond the stenosis, and if severe an enlarged right atrium and ventricle. The extent of the stenosis can be demonstrated by echocardiography and cardiac catheterization. If severe, balloon valvuloplasty is performed. Surgery is generally successful.



Pulmonary stenosis
Right ventricle hypertrophies to overcome the obstruction

Prophylaxis for infective endocarditis

Any child with congenital heart disease is at risk for developing infective endocarditis. The risk is reduced by surgical repair of the defect but persists with valve replacements. Antibiotic prophylaxis is required for dental treatment, ear, nose and throat (ENT) or GI surgery. It is also important to ensure good dental hygiene and prompt treatment of skin sepsis.

KEY POINTS

- Learn to identify the innocent murmur.
- Look for signs and symptoms of heart failure, including FTT.
- With experience the pathological murmurs can be identified.
- The child with congenital heart disease requires antibiotic prophylaxis.

44 Anaemia and pallor

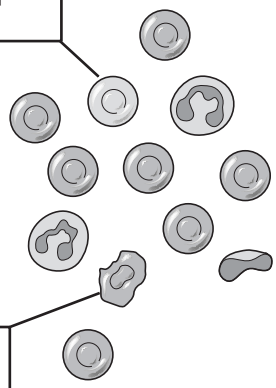
Causes of anaemia and pallor

Hypochromic microcytic anaemias

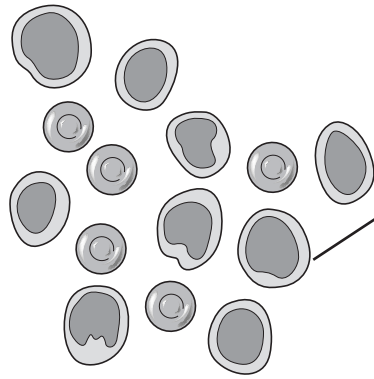
Iron deficiency anaemia
- Asymptomatic
- Common in babies and toddlers
- May be related to poor diet
- Affects intellectual development
- Low ferritin level

Thalassaemia trait
- Asymptomatic
- Affects Asians and Mediterraneans
- High HbA2 and HbF on electrophoresis

Lead poisoning
- Irritability and anorexia
- Colic
- Pica
- High lead level
- Causes acute encephalopathy (now rare)

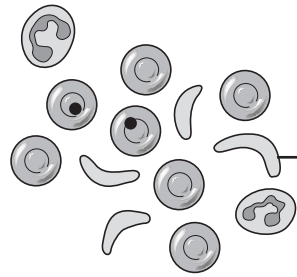


Other anaemias



Leukaemia (see Chapter 56)
- Insidious anorexia, irritability and lethargy
- Pallor
- Bleeding
- Fever
- Low Hb and platelets
- High white cell count with blast cells

Other causes
- Chronic infection
- Chronic renal failure
- Malignancies



Sickle cell anaemia
- Affects black individuals
- Identified by Hb electropheresis

What you need from your evaluation

History

- What is the child's diet like?
  Ask about consumption of milk. Early introduction of 'doorstep milk' causes microscopic bleeding from the gut. Excessive milk intake (>1 pint/day) after 12 months of age can reduce solid, and therefore iron, intake. Is the diet varied? Many young children are faddy about eating iron-rich foods. Ask about pica, a symptom of lead toxicity
- Is there any history of bleeding?
- What is the child's ethnic origin and is there consanguinity? Relevant for haemoglobinopathies
- What are the home conditions like?
  Could there be exposure to fumes or old lead paint?

Physical examination

- Carry out a full physical examination
- Anaemia has to be significant to be clinically apparent. It is best seen in the conjunctivae and nail beds
- Look for evidence of hepatosplenomegaly. This suggests more severe causes of anaemia

Investigations and their significance

As iron deficiency is so common it is usual to give a trial of iron before investigating hypochromic, microcytic iron deficiency.

Full blood count	Severity and type of anaemia (microcytic, hypochromic, etc.) Presence of bizarre cells. Presence of blast cells
Ferritin	Low in iron deficiency
Lead level	High in lead toxicity
Haemoglobin electrophoresis	Abnormal in haemoglobinopathies
Urea and electrolytes	Abnormal in renal failure
Blood and urine culture	Chronic infection
Bone marrow aspiration	Presence of leukaemic cells

Anaemia is usually detected when a blood count is performed routinely or on investigating another problem. It may also be suspected if a child looks pale. Iron deficiency is very common and, in childhood, a trial of iron treatment is usually given before investigating any further. Only if a child fails to respond to treatment are other causes of microcytic hypochromic anaemia considered, namely thalassemia trait or lead poisoning. If a child is ill more serious causes of anaemia are likely. These include chronic infection and chronic renal failure which give a normochromic normocytic picture. The commonest malignancy is leukaemia which can usually be suspected on the peripheral blood count. The haemoglobinopathies have characteristic clinical features.

Iron deficiency anaemia

In early childhood the combination of a high demand for iron to keep up with rapid growth and a poor intake of iron-rich foods makes iron deficiency very common. This can be exacerbated by chronic blood loss induced by early exposure to whole cow's milk. Iron deficiency anaemia can be as high as 50% in some populations, and in many countries young children are screened routinely. Babies beyond 12 months should be limited to one pint of milk daily to reduce blood loss and encourage the consumption of more iron-rich foods. Breast milk is somewhat protective as, although it has a relatively low iron content, the iron is absorbed more efficiently due to the iron binding protein, lactoferrin.

Iron deficiency anaemia is usually asymptomatic, but if the haemoglobin level falls significantly irritability and anorexia occur. Iron deficiency, even without anaemia, can affect attention span, alertness and learning. The initial finding is a low ferritin level reflecting inadequate iron stores. As the deficiency progresses microcytosis, hypochromia and poikilocytosis develop. The treatment is iron salts given orally for 2–3 months. The haemoglobin level starts to rise within 1 week of treatment. Failure to do so suggests non-compliance or an incorrect diagnosis.

Thalassaemia

The thalassaemias are a group of heritable hypochromic anaemias varying in severity, caused by a defect in haemoglobin polypeptide synthesis. Beta thalassemia is the commonest, and affects Asian and Mediterranean individuals. Thalassaemia trait (the heterozygous form) causes a mild hypochromic, microcytic anaemia, which may be confused with iron deficiency. Diagnosis is made by haemoglobin electrophoresis which demonstrates high levels of HbA2 and HbF. It requires no treatment.

Homozygous thalassemia results in a severe haemolytic anaemia, with compensatory bone marrow hyperplasia producing a characteristic overgrowth of the facial and skull bones. Major blood transfusions are required on a regular basis to maintain haemoglobin levels. Haemosiderosis is an inevitable consequence causing cardiomyopathy, diabetes and skin pigmentation, but can be minimized by continuous subcutaneous infusions of the chelating agent desferrioxamine.

Lead poisoning

Lead affects haem synthesis. The main source of lead is car exhaust fumes. Symptoms are usually non-specific consisting of irritability, anorexia and decreased playing. Colic and pica (eating non-nutrient substances such as earth) may occur, but acute encephalopathy with vomiting, ataxia and seizures is now rare. Chronic lead exposure has a detrimental effect on intellectual development. A hypochromic microcytic anaemia is found, and high lead levels confirm the diagnosis. Abdominal X-ray may demonstrate radiopaque flecks, and X-rays of the long bones may show leadlines (bands of increased density at the growing ends). Treatment consists of lead chelating agents which increase lead excretion. The source of lead must be identified and removed.

Sickle-cell anaemia

Sickle-cell anaemia is the commonest haemoglobinopathy, in which one of the amino acid sequences in the globin chain is substituted, causing an unstable haemoglobin (HbS). When deoxygenated, this forms highly structured polymers making brittle spiny red cells which occlude blood vessels, causing ischaemia. In the homozygous condition children experience recurrent acute painful crises which can be precipitated by dehydration, hypoxia or acidosis. Painful swelling of the hands and feet is a common early presentation, and repeated splenic infarctions eventually leave the child asplenic and susceptible to serious infections. Renal damage leads to a reduced ability to concentrate urine, making dehydration a severe problem. Its prevalence is confined to black populations.

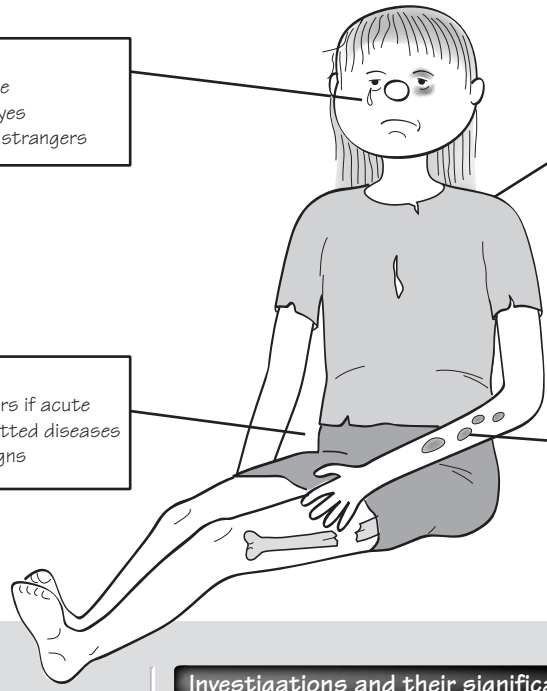
The peripheral blood smear typically shows target cells, poikilocytes and irreversibly sickled cells. Diagnosis is made by haemoglobin electrophoresis, which may also be used for screening in susceptible populations. Treatment is largely symptomatic with analgesics, antibiotics, warmth and adequate fluids during crises. Immunization status must be maintained and daily penicillin given in asplenic individuals. Sickle-cell trait is asymptomatic other than in conditions of low oxygen tensions such as occur at high altitude or under general anaesthesia.

KEY POINTS

- Iron deficiency is common in the childhood years, as it is hard to sustain iron stores in the face of rapid growth and a toddler's borderline intake of iron-rich foods.
- It is usual to give a therapeutic trial of iron for anaemia first, and only to investigate if there is no response.
- Principal causes of microcytic hypochromic anaemia include iron deficiency, lead poisoning and thalassemia trait.
- If a child is ill consider the less common causes of anaemia.

45 Neglect and abuse

Types of abuse and neglect



Emotional abuse
- 'Frozen watchful' appearance
- Expressionless face, wary eyes
- Abnormally affectionate to strangers

Physical neglect
- Unkempt dirty appearance
- Sores
- Uncared for nappy rash
- Failure to thrive

Sexual abuse
- Anogenital bruising and tears if acute
- Pregnancy, sexually transmitted diseases
- There may be no physical signs

Non-accidental injury
- Bruises of suspicious shape or site
- Burns and scalds
- Bites
- Hidden head injuries
- Suspicious fractures

What you need from your evaluation

History

- **How was the injury incurred?** Characteristically, the explanation is unconvincing, does not match the injury and there is a delay on obtaining medical advice. It is particularly suspicious if young not-yet-mobile infants have been injured
- **Past medical history.** Ask about previous injuries
- **Development and behaviour.** Both are affected by neglect and abuse
- **Social history and family history.** Find out who is in the home and who cares for the child. Abuse is more likely where there are changes in partner. Other professionals (e.g. health visitors and nursery nurses) can often provide extra details

Investigations and their significance

As the implications of non-accidental injury are so serious, rare medical causes of excessive bruising or fragile bones must be ruled out

• Photographs	Useful for further consultation and evidence in court
• Full blood count, bleeding time, PT and PTT	To rule out haematological causes of excessive bruising
• Skeletal survey (X-rays)	Certain fractures (of ribs, spiral fractures and metaphyseal chips in the long bones) and fractures at various stages of healing are particularly suspicious
• Pregnancy test and cultures (in sexual abuse)	The finding of sexually transmitted disease is strong corroborative evidence (and requires treatment)

Physical examination

- **General appearance.** Are there signs of neglect? Is the child particularly wary or over-affectionate towards the examiner?
- **Growth.** Plot measurements and weight and compare with previous measurements. Abused and neglected children often fail to thrive
- **Injuries.** Many non-accidental injuries have a characteristic appearance. Multiple injuries are suspicious, particularly if sustained at different times.
  - **Bruises:** Bruises, except on toddlers' legs, may be suspicious. The pattern may indicate how they were acquired. The age (identified from colour) may help in refuting an implausible explanation
  - **Burns and scalds:** Inflicted scalds are classically symmetrical without splash marks. Inflicted cigarette burns cause deep circular ulcers
  - **Bites:** The dental impression can be used forensically to identify the perpetrator
  - **Bony injuries:** Clinical evidence of fractures may be found
- **Neurological examination.** Retinal haemorrhages are a clue to subdural haemorrhage, which can occur when a baby is shaken
- **Signs of sexual abuse.** If sexual abuse is suspected, the genitalia and anus must be examined by an experienced paediatrician. Signs may be overt, such as bruising and tears, or subtle. The absence of signs does not refute the diagnosis

If there is any suspicion that a child is a victim of abuse or neglect, he or she should be seen immediately by a paediatrician experienced in child protection work, and the Social Services Department needs to be informed. The evaluation should be conducted in privacy and the child's trust gained. Comprehensive clear notes must be made and where necessary photographs taken as they may be required for evidence. Helpful information can be obtained from other health professionals such as health visitors, nursery nurses, social workers, the GP and school, who may throw light on the child's circumstances.

If abuse or neglect is confirmed, child protection procedures will be initiated and may include admission to a place of safety, obtaining an emergency care order from court and a case conference. The outcome may involve registration on the Child Protection Register (see p. 35), social services supervision and support at home, or foster care.

Physical abuse (non-accidental injury)

In most cases the abuser is a related carer or male friend of the mother. Most have neither psychotic nor criminal personalities, but tend to be unhappy, lonely, angry adults under stress, who may have experienced child abuse themselves. Injuries may range in severity from minor bruises to fatal subdural haematomas. Abused children are commonly fearful, aggressive and hyperactive, and many go on to become delinquent, violent and the next generation of abusers. Children with repeated injury to the CNS may develop brain damage with learning disabilities or epilepsy. About 5% of abused children who are returned to their parents without intervention are killed and 25% seriously injured.

Subdural effusions and haematomas

A subdural haematoma is a collection of bloody fluid under the dura. It results from rupture of the bridging veins that drain the cerebral cortex. Although any form of head trauma may produce subdural bleeding, the abused infant who is forcibly shaken is particularly susceptible to this injury. Subdural haematomas may be acute, or chronic, in which case they may eventually be replaced by a subdural collection of fluid. They can lead to blockage of CSF flow and hydrocephalus. The infant usually presents with fits, irritability, lethargy, vomiting and FTT. Signs of raised intracranial pressure and retinal haemorrhages are common. The diagnosis is made by radiological imaging, and the treatment neurosurgical. The prognosis for recovery is variable and depends on the associated cerebral insult.

Sexual abuse

Sexual abuse may take the form of inappropriate touching, forced exposure to sexual acts, vaginal, oral or rectal intercourse and sexual assault. Secrecy is often enforced by the offender who is usually male and a family member or acquaintance of the family. It may come to light if disclosure is made, inappropriate sexual behaviour is exhibited or as a result of trauma or genital infections. Signs of trauma may be found in the mouth, anus or genitalia, but absence of signs is common and fewer than half the victims have any substantiating physical evidence. Particularly sensitive and skilled management is required and should only be undertaken by experienced professionals. All victims require psychological support, and without intervention are likely to be seriously disturbed. They often grow up unable to form close relationships, and commonly enter abusive relationships later in life.

Non-organic FTT (see p. 92)

A proportion of young children who fail to thrive do so as a result of neglect, the principal factor being inadequate nutrition. The mother is commonly deprived and unloved herself and often is clinically depressed. The child looks malnourished and uncared for, and immunizations are often not up to date. Delays in development are common, and signs of physical abuse may be seen. When admitted to hospital these babies often show rapid weight gain. With intervention, catch-up growth may occur, but brain growth may be jeopardized and emotional and educational problems are common. Without detection and intervention a small proportion of these children die from starvation.

KEY POINTS

Characteristics of non-accidental injury:

- Injuries in very young children.
- Explanations which do not match the appearance of the injury, and change.
- Multiple types and age of injury.
- Injuries which are 'classic' in site or character.
- Delay in presentation.
- Things the child may communicate during the evaluation.

46 The child with developmental delay

Causes of developmental delay

Central nervous system malformation
- Neural tube defects
- Hydrocephalus

Chromosomal abnormalities
- Down syndrome
- Fragile X

Prenatal injury
- Fetal alcohol syndrome
- Intrauterine infections (TORCH)

Endocrine and metabolic defects
- Congenital hypothyroidism
- Phenylketonuria

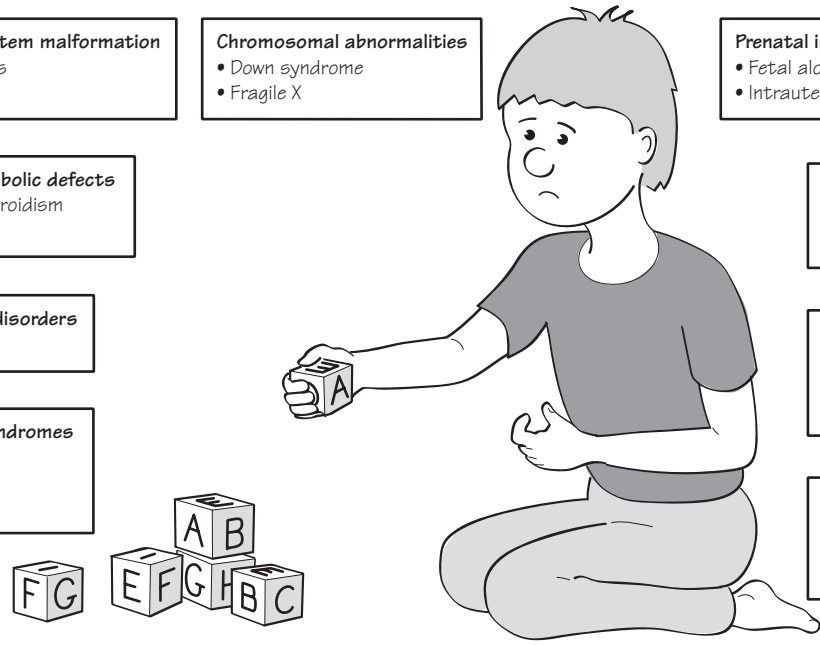
Perinatal injury
- Asphyxia
- Birth trauma

Neurodegenerative disorders
- Leucomalacia

Postnatal injury
- Meningitis
- Non-accidental injury
- Neglect

Neurocutaneous syndromes
- Sturge Weber
- Neurofibromatosis
- Tuberous sclerosis

Idiopathic
- Autism
- Various dysmorphic syndromes



What you need from your evaluation

History

Children are often uncooperative so parental report is particularly important.

Developmental milestones
- Enquire systematically about milestones for the four developmental areas
- Ascertain the extent of delay and which areas are affected
- Remember to allow for prematurity during the first 2 years. Beyond that catch-up in development rarely occurs
- Loss in skills suggests a neurodegenerative condition
- Ask whether there are concerns about vision and hearing

Past medical history
- Enquire into alcohol consumption, medical problems and medication during pregnancy
- Enquire about prematurity and neonatal complications

Family history
- Ask about learning difficulties and consanguinity

Physical examination

Developmental skills
- Assess each developmental area in turn: gross motor, fine motor/adaptive, language and social skills
- Attempt to evaluate vision and hearing
- Assess factors such as alertness, responsiveness, interest in surroundings, determination and concentration; these all have positive influences on a child's attainments

General examination
- Dysmorphic signs suggest a genetic defect, chromosome anomaly or teratogenic effect
- Microcephaly at birth suggests fetal alcohol syndrome or intrauterine infections
- Poor growth is common, but may be due to hypothyroidism or non-organic failure to thrive (look for signs of neglect)
- Look for café-au-lait spots, depigmented patches and portwine stains which are indicative of neurocutaneous syndromes
- Hepatosplenomegaly suggests a metabolic disorder

Neurological examination
- Look for abnormalities in tone, strength and coordination, deep tendon reflexes, clonus, cranial nerves and primitive reflexes, and ocular abnormalities

Investigations

- Chromosome analysis, thyroid function tests and urine screen for metabolic defects are usually obtained in global developmental delay
- More sophisticated metabolic investigations and brain imaging may be indicated for some
- A hearing test is mandatory in language delay

The term *global* developmental delay refers to delay in all milestones, but particularly language, fine motor and social skills and is particularly worrying as it usually indicates significant learning disability (mental retardation). Delay in a single area is much less concerning. Warning signs indicative of significant developmental problems are described in the Table on p. 27.

Repeat assessments may be needed to get an accurate view of a child's difficulties, and referral to an appropriate therapist for further assessment and guidance may be required. When developmental difficulties are complex, the child should be referred to a Child Development Team (p. 120) for assessment and input. Parents concerns must be properly addressed, as ongoing parental anxiety in itself can be damaging to the child.

Severe learning disabilities (mental retardation)

The commonest causes of severe learning disability are Down syndrome (p. 127), fragile X (p. 127) and cerebral palsy (see Chapter 59). With advances in the field of genetics more diagnoses are being made, particularly in children with congenital anomalies and dysmorphic features. This has been facilitated by the development of computerized databases. As chromosomal anomalies are not uncommonly found in children with delay and dysmorphism, it is worth taking blood for a karyotype. However, in about one-third of children with global developmental delay no specific cause is identified.

Intrauterine infections

If infection with organisms such as rubella, cytomegalovirus (CMV) or toxoplasmosis occurs for the first time during pregnancy severe fetal damage can result, leading to multiple handicaps and microcephaly. Visual and hearing deficits are common.

Fetal alcohol syndrome

The fetal alcohol syndrome is a common cause of learning disabilities, and is characterized by a characteristic facial appearance, cardiac defects, poor growth and microcephaly, It is caused by a moderate to high intake of alcohol during pregnancy, with the severity of the problems related to the quantity of alcohol consumed.

Congenital hypothyroidism

Lack of thyroid hormone in the first years of life has a devastating effect on both growth and development. However, since neonatal screening has been introduced, it is now a rare cause of developmental delay. The defect is due to abnormal development of the thyroid or inborn errors of thyroxine metabolism.

Babies usually look normal at birth, but may have features of cretinism, including coarse facies, hypotonia, a large tongue, an umbilical hernia, constipation, prolonged jaundice and a hoarse cry. In the older baby or child delayed development, lethargy and short stature are found. Thyroid function tests reveal low T_4 and high TSH levels.

Congenital hypothyroidism is one of the few treatable causes of learning disabilities. Thyroid replacement is required lifelong and must be monitored carefully as the child grows. If therapy is started in the first few weeks of life and compliance is good, the prognosis for normal growth and development is excellent.

Inborn errors of metabolism

This group of disorders are caused by single gene mutations, inherited in an autosomal recessive manner. Consanguinity is therefore common. They present in a variety of ways of which developmental delay is one, but may also include neonatal seizures, hypoglycaemia, vomiting and coma. Children sometimes have coarse features, microcephaly, FTT and hepatosplenomegaly. Inborn errors of metabolism are rare. Phenylketonuria is the commonest and is routinely screened for in all neonates.

Neurodegenerative disorders

A neurodegenerative disease is characterized by progressive deterioration of neurological function. The causes are heterogeneous and include biochemical defects, chronic viral infections and toxic substances, although many remain of unknown aetiology. Children may have coarse features, fits and intellectual deterioration, and microcephaly. The course for all of these conditions is one of relentless and inevitable neurological deterioration.

Neurocutaneous syndromes

The neurocutaneous syndromes are a heterogeneous group of disorders characterized by neurological dysfunction and skin lesions. In some individuals there may be severe learning disabilities and in others intelligence is normal. Examples include Sturge–Weber syndrome, neurofibromatosis and tuberous sclerosis. The aetiology of these problems is not known, but most are familial.

Abuse and neglect

Emotional abuse and neglect can have serious consequences for a child's developmental progress. The delay is often associated with FTT. On presentation the child may be apathetic, have evidence of physical neglect such as dirty clothing, unkempt hair and nappy rash, and there may be signs of non-accidental injury. If there is any suggestion of regression of developmental skills, chronic subdural haematomas (which can occur as a result of shaking injuries) should be considered.

Intensive input and support is required. Day nursery placement can provide good stimulation, nutrition and care. If the child continues to be at risk for on-going abuse or neglect the child must be removed from the home. The prognosis depends on the degree of the damage incurred and how early the intervention is provided. Children who require removal from the home often have irreversible learning and emotional difficulties.

KEY POINTS

- All developmental areas must be accurately assessed in turn.
- Remember to correct for prematurity in the first 2 years, and carry out a full physical and neurological examination.
- Repeat evaluations may be required over time.
- Attempt to make a diagnosis or identify the aetiology for the difficulties.
- Involve the Child Development Team if difficulties are complex.

47 The newborn baby

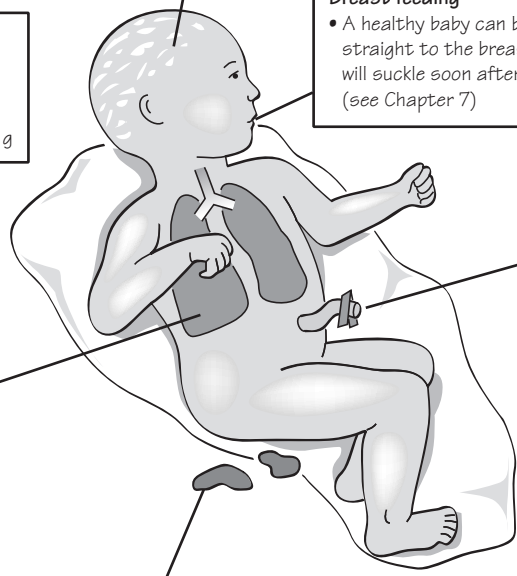
Gestation
- Full-term: born between 37 and 42 weeks gestation
- Preterm: born before 37 completed weeks gestation
- Post-term: born after 42 completed weeks

Vernix
- A white waxy substance that is often present, especially in preterm infants. Thought to provide insulation and lubrication

Mortality statistics
- Stillbirth: a baby (>24 weeks gestation) who shows no signs of life after delivery (including no heart beat)
- Perinatal mortality rate: Stillbirths and deaths within the first week of life per 1000 live-born and stillborn infants; approximately 8 per 1000 in the UK
- Infant mortality rate: Number of deaths in the first year of life per 1000 live-born infants

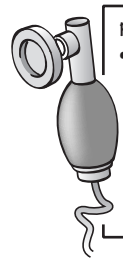
Birthweight
- Small for gestational age: birthweight <10th centile
- Very low birthweight: <1500 g
- Extremely low birthweight: <1000 g

Breast feeding
- A healthy baby can be put straight to the breast and will suckle soon after birth (see Chapter 7)



Umbilical cord
- Normally has two arteries and one vein
- After clamping it dries up and usually separates from the baby within the first week. It can be a site of infection

The first breath
- Clamping the umbilical cord and response to cold stimulates the baby to take its first breath within a minute of birth
- Lung fluid is actively reabsorbed as well as being expelled during delivery
- Surfactant in the alveoli helps them to expand and fill with air
- Most babies can be dried and given straight to the mother. Occasionally, the onset of breathing may be delayed and the baby requires resuscitation



Resuscitation
- Usually, simple stimulation, drying and warming is sufficient but resuscitation may be needed in some babies. Oxygen can be given by bag and mask and the baby placed under a warmer. Intubation may be needed, especially if the baby has aspirated meconium

Meconium
- Green–black stool passed by babies in the first days after birth
- Fresh meconium-stained liquor can be a sign of fetal distress
- The asphyxiated gasping baby can aspirate meconium into the lungs, causing respiratory distress

Vitamin K
- Babies can sometimes be deficient in vitamin K, leading to severe bleeding disorders
- Vitamin K is routinely offered to all babies, either orally or by intramuscular injection. Breast-fed babies require three oral doses as breast milk contains less Vitamin K than formula milks

The Apgar score

Score	0	1	2
Heart rate	Absent	<100	>100
Respiration	Absent	Irregular	Strong cry
Tone	Limp	Some flexion	Good flexion
Reflex to suction	None	Grimace	Cough or sneeze
Colour	White	Blue periphery	Pink

- The Apgar score is often used to describe the infant's condition at birth.
- The score (0–10) is recorded at 1 min and 5 min of life.
- A normal score at 1 min is 7–10.
- Babies with a score of 0–3 at 1 min require rapid resuscitation or they will die

The normal newborn

The vast majority of babies are born at term, in good condition and do not require any medical involvement. Nearly all babies in the UK are born in hospital, where a paediatrician is usually available to attend 'high risk' deliveries where it may be anticipated that some resuscitation will be required. A healthy newborn term infant will cry soon after birth, will be pink with good muscle tone, a normal heart rate and regular respiration. Once the cord has been clamped and cut, these babies can be dried and given straight to their mothers for skin to skin contact and to establish breast-feeding. Newborn babies, especially if they are premature, will be covered in a waxy material called vernix. Post-term infants may have very dry, cracked skin. Babies pass a green/black stool called meconium. This changes to a normal yellow/brown stool after a few days. It is recommended that infants are given Vitamin K at birth to prevent potentially catastrophic bleeding. Newborn infants are routinely examined within the first few days to exclude congenital abnormalities (see Chapter 48) and have blood taken from a heel-prick around day 6 to screen for hypothyroidism and metabolic disorders (see p. 38).

Asphyxia and resuscitation

The perinatal mortality rate, which is currently 8 per 1000 has halved in the UK over the last 20 years, largely due to improvements in obstetric care. The reduction in neonatal mortality rate (now less than 5 per 1000 live births) is due to improvements in the management of babies with complex congenital abnormalities and to improved care of preterm infants. Some babies do still require immediate resuscitation after birth, and personnel attending deliveries must be trained in effective and rapid resuscitation. The need for resuscitation can often be anticipated in advance, and a paediatrician should be in attendance. Such situations include:

- Prematurity.
- Fetal distress.
- Thick meconium staining of the liquor.
- Emergency caesarean section.
- Instrumental delivery.
- Known congenital abnormality multiple births.

The condition of the infant after birth is described by the Apgar score (see opposite). Each of five parameters is scored from 0–2. A total Apgar score of 7–10 at 1 min of age is normal. A score of 4–6 is a moderately ill baby and 0–3 represents a severely compromised infant who may die without urgent resuscitation. Such babies will require intubation and may require cardiac massage. In the most depressed babies IV drugs such as epinephrine and bicarbonate may be necessary to re-establish cardiac output. The outcome for these infants may be poor.

Some infants with very low Apgar scores may have suffered a hypoxic or ischaemic insult during pregnancy or labour. A healthy fetus can withstand physiological asphyxia for some time, but an already compromised fetus may become exhausted and decompensate with build-up of lactic acid. These infants may develop irreversible organ damage, in particular to the brain. Evidence of severe asphyxia includes a cord blood pH < 7.05, Apgar score of <5 at 10 min, a delay in spontaneous respiration beyond 10 min and development of a characteristic encephalopathy with abnormal neurological signs including convulsions. Death or severe handicap occurs in more than 25% of the most severely asphyxiated term infants.

Intrauterine growth retardation

A baby with a birthweight below the 10th centile is small for gestational age (SGA). This may be normal or may be due to intrauterine growth retardation (IUGR). The pattern of growth retardation gives some indication of the cause. An insult in early pregnancy, such as infection, will cause symmetrical growth retardation with short length and a small head. A later insult, usually placental insufficiency, will cause asymmetric growth retardation with relative sparing of the head (brain) growth due to selective shunting of blood to the developing brain.

Causes of IUGR include multiple pregnancy, placental insufficiency, maternal smoking, congenital infections (e.g. toxoplasmosis, rubella) and genetic syndromes (e.g. Down syndrome). Babies with severe IUGR should be screened for congenital infection ('TORCH' (Toxoplasmosis, Other [syphilis], Rubella, Cytomegalovirus, Hepatitis, HIV) screen).

In the first few days of life, babies with IUGR are at risk of hypoglycaemia and hypothermia due to low glycogen stores and lack of subcutaneous fat. Symptomatic hypoglycaemia carries a poor prognosis for normal intellectual development. If there has been poor head growth during pregnancy intellect may be further impaired. Babies with IUGR usually show catch-up growth during infancy if they are given increased feeds. There is recent evidence that IUGR babies are at increased risk of hypertension, ischaemic heart disease and diabetes in later life.

Vitamin K

Vitamin K deficiency or persistent jaundice can lead to poor synthesis of vitamin K-dependent clotting factors and subsequent bleeding. The bleeding may be minor bruising or significant intracranial haemorrhage, which can cause disability or death. This is known as haemorrhagic disease of the newborn (not to be confused with haemolytic disease of the newborn, due to ABO incompatability). Breast milk is particularly low in vitamin K, unlike formula milk which is supplemented. For this reason vitamin K should be given routinely to all newborn infants. It is either given intramuscularly or orally at birth, 1 and 6 weeks. Babies with persistent jaundice should receive further doses (see Chapter 50).

KEY POINTS

- Most babies are born healthy and do not require any resuscitation.
- Vitamin K is recommended for all babies.
- The Apgar score is used to describe the condition after birth.
- Babies with severe IUGR are at increased risk of asphyxia, hypoglycaemia and hypothermia, and may be at risk of intellectual impairment.

48 Congenital abnormalities

The newborn examination

All newborn babies are carefully examined in the first 24 h of life to check that they are healthy and to detect congenital abnormalities, some of which may not be obvious to the parents. The baby should be fully undressed in a warm room and examined from head to toe. Ask the mother if she has any concerns and whether there is any family history of note, for example of deafness or congenital dislocation of the hips

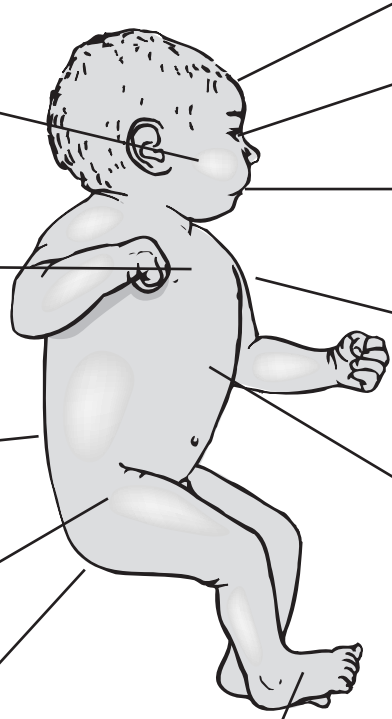
General observation
- Weight, length and head circumference
- Maturity
- Muscle tone
- Reflexes: Moro, grasp, suck and rooting
- Is this a healthy baby who is feeding well?

Skin
- Pallor
- Jaundice
- Cyanosis
- Rashes (erythema toxicum is normal)
- Birthmarks (see p. 88)

Head
- Anterior fontanelle
- Cephalhaematoma (parietal swelling that does not cross suture lines)
- Chignon from Ventouse suction cup

Face—dysmorphic features?
- Low set or simple ears
- Inner epicanthic folds
- Mongolian or anti-Mongolian slant of eyes
- Symmetry of face and mouth
- Accessory auricles and pre-auricular pits
- Micrognathia (small chin)

Eyes
- Red reflex (to exclude cataract)
- Jaundiced sclera
- Colobomata (defect in the pupil)



Mouth
- Cleft lip/palate
- Central cyanosis
- Neonatal teeth

Heart
- Cyanosis
- Heart failure (tachypnoea, hepatomegaly)
- Heart murmur
- Femoral pulses (coarctation)
- Apex beat (dextrocardia)

Chest
- Respiratory rate
- Respiratory distress
- Symmetry of chest movement (pneumothorax, diaphragmatic hernia)

Back and spine
- Spina bifida or posterior encephalocele
- Midline naevus, lipoma or deep sacral pit can suggest an underlying spinal abnormality

Abdomen
- Abdominal distension or bile-stained vomiting suggest bowel obstruction (e.g. atresia)
- Palpable kidneys (hydronephrosis)
- Anterior abdominal wall defects (gastroschisis or exomphalos)
- Three vessels in umbilical cord? (normal)

Hips
- Barlow and Ortolani tests for congenital dislocation of the hips (CDH)
- Ask about risk factors (breech, family history of CDH)

Common syndromes to be aware of:
- Trisomy 21 (Down syndrome)
- Trisomy 13 (Patau syndrome)
- Trisomy 18 (Edward syndrome)
- Turner syndrome (45 XO)
- Noonan syndrome (lymphoedema)
- VATER and VACTERL association
- Pierre-Robin sequence

Genitalia and anus
- Hypospadias (urinary meatus on underside of penis)
- Cryptorchidism (undescended testes)
- Ambiguous genitalia: if both testes are impalpable, consider whether the baby could be a virilized female, due to congenital adrenal hypoplasia
- Imperforate anus (may have fistula to bladder or vagina)

Limbs
- Talipes equinovarus (club foot)
- Polydactyly (extra digits or toes)
- Syndactyly (fused digits or toes)
- Single palmar crease and 'sandal gap' between toes (Down syndrome)
- Contractures (oligohydramnios or congenital muscular disorder)
- Absent radii (VACTERL association)

Overall, the incidence of congenital abnormalities is 10–15 per 1000 births. The commonest abnormality is a congenital heart defect, which occurs in 8 per 1000 births. Congenital heart disease is described in Chapter 51. Abnormalities may be minor such as a small naevus (birthmark) or very severe such as spina bifida. Some babies show characteristic patterns of abnormality which suggest a 'syndrome' diagnosis, such as Down syndrome (trisomy 21). The majority of congenital abnormalities are genetically determined, though in many cases the exact defect has not been determined. Others may be due a combination of genetics and environment, such as spina bifida (see below) or due entirely to environment (e.g. fetal alcohol syndrome).

A *syndrome* is a consistent pattern of dysmorphic features seen together, and suggests a genetic origin. A *sequence* is where one abnormality leads to another—for example the micrognathia in Pierre Robin sequence causes posterior displacement of the tongue, which prevents the palate forming correctly, leading to cleft palate. An *association* is a non-random collection of abnormalities, as in the VACTERL association (see below).

Common syndromes

Down syndrome (trisomy 21) occurs in 1 in 800 live births. There is an association with increased maternal age. Ninety-five per cent are due to non-dysjunction during meiosis and 5% to an unbalanced translocation. Features include hypotonia, brachycephaly, mongolian slant to eyes, a single palmar crease, wide-spaced first toe and congenital heart disease, especially atrioventricular septal defect (AVSD) and VSD. These infants have mild to moderate learning difficulties, an increased risk of hypothyroidism and leukaemia.

Patau syndrome (trisomy 13) occurs in 1 in 4000 to 1 in 10 000 live births. The main features are midline defects including cleft lip and palate, areas of skin loss on the scalp, holoprosencephaly, severe mental retardation, polydactyly, prominent heals and congenital heart defects.

Edward syndrome (trisomy 18) occurs in 1 in 8000 live births and usually leads to death in infancy. These infants have 'rocker-bottom' feet, congenital heart defects, microcephaly with a prominent occiput and overlapping digits.

Turner syndrome (45XO) occurs in females, and leads to short stature and ovarian failure (see p. 91).

Noonan syndrome is phenotypically similar to Turner's but occurs in males and females. There is short stature, neck webbing, lymphoedema and congenital heart disease, especially pulmonary stenosis.

VACTERL association: Vertebral anomalies, Anal atresia, Cardiac anomalies, Tracheo-oEsophageal fistula, Renal anomalies, Limb defects. Infants usually have a tracheo-oesophageal fistula, often with oesophageal atresia and limb abnormalities, particularly small or absent radii leading to shortened, curved forearms.

Cleft lip and palate

Cleft lip is a distressing congenital abnormality because of the initial cosmetic appearance. It occurs in 1 in 1000 infants, and tends to recur in families although there is no autosomal inheritance. There is an associated cleft palate in about 70% of cases. Increasingly cleft lip is diagnosed by antenatal ultrasound scan, which allows the parents to be counselled and to prepare for the distressing appearance. After birth a cleft palate is confirmed by feeling the defect in the palate with a clean finger inserted in the mouth. Some infants have a submucosal cleft which is palpable but not visible. These children are best managed by a multidisciplinary team including plastic surgeon, orthodontist and speech therapist. Surgical repair of the lip is usually performed at 3 months and of the palate at 9 months of age. The cosmetic appearance following plastic surgery is usually excellent. Showing the parents 'before and after' photographs of other infants can help allay their anxieties. Expected difficulties include problems establishing milk feeds, aspiration of milk, speech problems, conductive hearing loss due to Eustachian tube dysfunction and dental problems. Regular audiological assessments are essential.

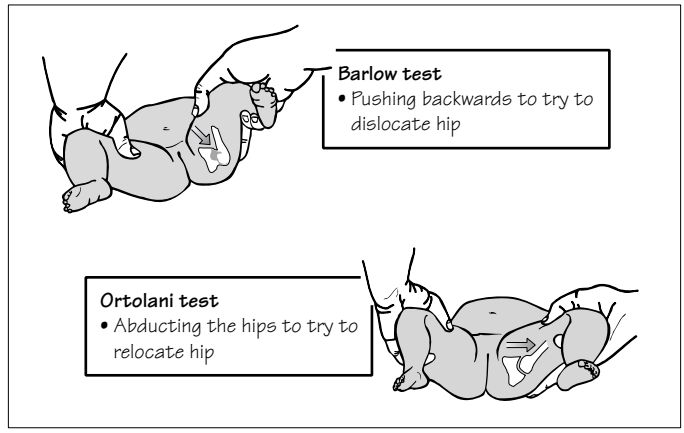
Neural tube defects (spina bifida)

Spina bifida results from the failure of the neural tube to close normally in early pregnancy. It used to be a major cause of severe disability, but the recommendation to take folic acid supplements before conception and in the first 3 months of pregnancy has reduced the incidence by 75%. Routine antenatal ultrasound screening and selective termination of pregnancy has made open spina bifida a rare condition. Neural tube defects are always in the midline. The severity of the lesion depends on the extent to which the neural tube has failed to develop:

- **Anencephaly**. This is the most severe form where the cranial part of the neural tube does not exist and the brain does not develop.
- **Myelomeningocele**. An open lesion where the spinal cord is covered by a thin membrane of meninges. There is severe neurological weakness of the lower limbs with bladder and anal denervation and an associated hydrocephalus. Survivors have severe disability and are at risk of renal failure from recurrent urinary tract infections.
- **Meningocele**. The spinal cord is intact and functionally normal, but there is an exposed sac of meninges which can rupture, with the risk of developing meningitis and hydrocephalus. Surgical closure is required urgently to prevent infection.
- **Spina bifida occulta**. This is a 'hidden' neural tube defect where there is failure of the vertebral bodies to fuse posteriorly. The only clue may be a tuft of hair, naevus, lipoma or deep sacral pit in the midline over the lower back. An ultrasound or MRI scan is indicated to exclude tethering of the spinal cord which can cause neurological dysfunction as the child grows.

Congenital dislocation of the hip

Congenital dislocation of the hip (CDH) occurs in 2 per 1000 infants. Risk factors include breech delivery, a family history, female sex and neurological defects which impair lower limb movement, e.g. spina bifida. Examination includes observation of symmetrical skin creases and leg length, the Ortolani test (a *dislocated* hip will not abduct fully) and the Barlow test (feeling for a clunk as a *dislocatable* hip slips out of the acetabulum). In some areas high-risk babies are routinely referred for ultrasound examination of the hips in the neonatal period. Treatment involves immobilizing the joint in abduction for several months. Surgery is rarely needed.

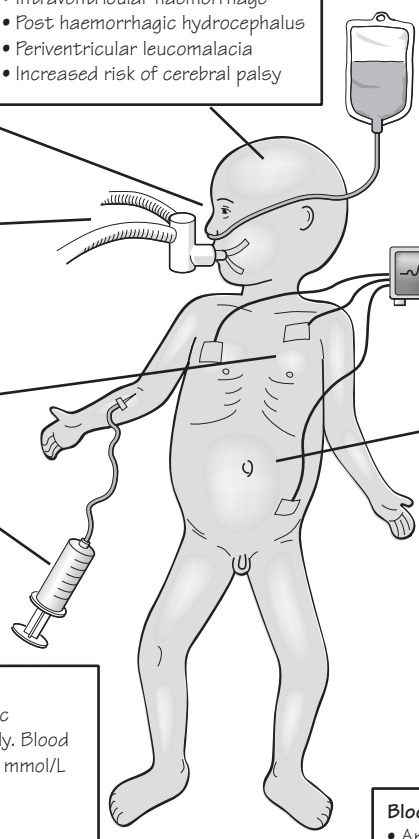


KEY POINTS

- The overall incidence of congenital abnormality is approximately 2%.
- The newborn examination is performed to detect significant abnormalities which may not be obvious.
- Congenital dislocation of the hips is commoner in breech babies.
- Folic acid supplements have dramatically reduced the incidence of spina bifida.

49 Prematurity

Complications of prematurity



Eyes
- Retinopathy of prematurity due to abnormal vascularization of the developing retina
- Requires laser treatment to prevent retinal detachment and blindness

Brain
- Intraventricular haemorrhage
- Post haemorrhagic hydrocephalus
- Periventricular leucomalacia
- Increased risk of cerebral palsy

Temperature control
- Increased surface area to volume ratio leads to loss of heat
- Immature skin cannot retain heat and fluid efficiently
- Reduced subcutaneous fat reduces insulation

Respiratory
- Respiratory distress syndrome (surfactant deficiency)
- Apnoea and bradycardia
- Pneumothorax
- Chronic lung disease

Gastrointestinal
- Necrotizing enterocolitis: a life-threatening inflammation of the bowel wall due to ischaemia and infection and which can lead to bowel perforation
- Gastro-oesophageal reflux
- Inguinal hernias (with high risk of strangulation)

Cardiovascular (see Chapter 51)
- Hypotension
- Patent ductus arteriosus

Nutrition
- May require parenteral nutrition
- Nasogastric feeds until sucking reflex develops at 32–34 weeks
- Difficult to achieve *in-utero* growth rates

Infection
- Increased risk of sepsis, especially group B streptococcus and coliforms
- Pneumonia is common
- Infection is a common complication of central venous lines required for feeding

Metabolic
- Hypoglycaemia is common. Symptomatic hypoglycaemia must be treated promptly. Blood glucose should be maintained above 2.6 mmol/L to prevent neurological damage
- Hypocalcaemia
- Electrolyte imbalance
- Osteopenia of prematurity (with risk of fractures)

Blood
- Anaemia of prematurity
- Neonatal jaundice (see Chapter 50)

What you need from your evaluation

History

- **Risk factors for prematurity:** young maternal age, multiple pregnancy, infection, maternal illness (e.g. pregnancy-induced hypertension), cervical incompetence, antepartum haemorrhage
- Full obstetric history
- **Condition at birth:** Apgar score, resuscitation required
- **Birthweight:** appropriate for gestational age?
- **Gestation:** must be known to give accurate prognosis. Calculate from menstrual period, by early dating ultrasound scan or by assessment of gestation after birth (Dubowitz score)
- Associated problems such as twin pregnancy (much higher risk of poor neurological outcome), congenital abnormalities or infection (chorioamnionitis may have been trigger for preterm labour)
- **Antenatal steroids:** if given, these reduce the incidence of respiratory distress syndrome and intraventricular haemorrhage

Long-term complications

- **Mortality:** only about 20% of infants born at 24 weeks gestation survive. By 27 weeks this rises to 70% and after 32 weeks the chances of survival are excellent
- **Chronic lung disease:** (bronchopulmonary dysplasia). This is a consequence of requiring ventilation and may require long-term oxygen treatment for months or sometimes years
- **Neurological sequelae:** there is a significant risk of hydrocephalus developing secondary to an intraventricular haemorrhage. A shunt may need to be inserted to relieve pressure. Hypotension may have been sustained, leading to peri-ventricular leucomalacia. This carries the risk of cerebral palsy, particularly of the diplegic type
- **Blindness:** as a consequence of severe retinopathy of prematurity (ROP). This is becoming less common with better prevention, detection and treatment of ROP
- **Poor growth:** especially if catch-up growth is not achieved

A baby is premature if born before 37 weeks gestation: 7% of all babies are premature and 1% are extremely premature with birthweight < 1500 g (very low birthweight). Premature babies are now viable from 24 weeks gestation, although mortality is high (only 16% survival at 24 weeks) and about 25% of those that survive will have severe disability. Beyond 32 weeks the prognosis is excellent. Premature infants are at risk of a number of complications, many of which, such as hypothermia, hypoglycaemia and difficulty feeding, are common to both premature babies and babies with IUGR. Some preterm babies will also be light for dates due to IUGR.

Premature babies are cared for on a Special Care Baby Unit (SCBU) or Neonatal Intensive Care Unit (NICU), where they receive specialist medical care. Incubators provide a warm humidified environment to prevent hypothermia and protect the infant's skin, which is often thin, transparent and red, and does not provide an adequate barrier to heat and fluid loss. Feeding problems are common due to immaturity of the GI system. A strong suck reflex does not develop until 35 weeks gestation, so premature babies often need feeding via a naso-gastric tube. Very sick premature babies, or those with concurrent growth retardation or asphyxia, may be at increased risk of necrotizing enterocolitis and are therefore fed intravenously using parenteral nutrition. Premature babies are at risk of infection, either acquired from the mother during delivery or from the hospital environment. Much of the disability caused by prematurity is due to intracranial haemorrhage, and periventricular leucomalacia (PVL). Both may lead to the child developing cerebral palsy.

Respiratory distress syndrome

Respiratory distress occurs very frequently in premature infants. Causes include pneumonia, pneumothorax, meconium aspiration, cardiac failure and diaphragmatic hernia. The commonest cause is respiratory distress syndrome (RDS) due to surfactant deficiency

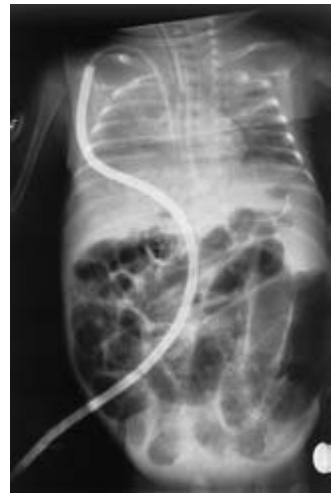
Signs of RDS include tachypnoea, intercostal and sternal recession, cyanosis and expiratory 'grunting'. Diagnosis is confirmed by chest X-ray which shows a 'ground glass' appearance due to alveolar collapse with the presence of an air-bronchogram (radiolucent air in the bronchial tree seen against the airless lung). RDS is caused by deficiency of surfactant, a phospholipid which reduces surface tension in the alveoli. Surfactant is not normally produced until about 34–36 weeks gestation, although the stress of birth usually stimulates production, and RDS is therefore usually self-limiting, lasting up to 7 days. Giving corticosteroids antenatally to mothers at risk of preterm delivery can prevent RDS by stimulating surfactant production. IUGR babies are physiologically 'stressed' and tend to get less RDS because of endogenous corticosteroid release.

Management of RDS involves giving sufficient oxygen and supporting respiration, either with continuous positive airway pressure (CPAP) via the nose, or by mechanical ventilation via an endotracheal tube. Exogenous surfactant can be administered via the endotracheal tube. This treatment has reduced the mortality of RDS by over 40%.

Some babies with RDS develop a complication known as chronic lung disease (broncho-pulmonary dysplasia). This may require long-term oxygen treatment at home.

Necrotizing enterocolitis (NEC)

This is a rare complication due to impaired blood flow to the bowel. Mucosal ischaemia allows gut microorganisms to penetrate the bowel wall causing a severe haemorrhagic colitis. Babies with NEC present with acute collapse, abdominal distension, bile-stained vomiting and bloody diarrhoea. An X-ray may show gas in the bowel wall or portal tract. Management involves stopping milk feeds, supporting the circulation and giving antibiotics. Laparotomy is required if perforation occurs. Complications include intestinal strictures and short bowel syndrome.



Retinopathy of prematurity (ROP)

ROP is common in very premature infants, occurring in up to 50% of very low birthweight (<1500 g) babies. In the majority of cases it requires no treatment and resolves spontaneously. In about 1% of these babies it causes blindness. ROP is caused by proliferation of new blood vessels in an area of relative ischaemia in the developing retina. Oxygen toxicity is one of the causes although the pathophysiology is not yet well understood. At-risk infants should be screened for ROP. If detected, cryotherapy or laser treatment can be used to prevent the risk of retinal detachment and blindness.

Brain injury

Preterm infants are at particular risk of brain injury and this is the most important factor in determining their long-term prognosis. Periventricular leucomalacia, large intraventricular haemorrhage and posthaemorrhagic hydrocephalus are the most important in terms of causing disability. Term infants are also at risk of developing hypoxic–ischaemic encephalopathy after an asphyxial insult (see p. 103).

Intraventricular haemorrhage (IVH) occurs in up to 40% of very low birthweight infants. Haemorrhage develops in the floor of the lateral ventricle and ruptures into the ventricle. In less than 25% of cases the haemorrhage extends into the white matter around the ventricle by a process of venous infarction. This carries a high risk of cerebral palsy (see Chapter 59). IVH may be asymptomatic and is usually diagnosed by cerebral ultrasound scan. The prognosis depends on the extent of IVH.

Post haemorrhagic hydrocephalus occurs in 15% and may require the insertion of a ventriculo-peritoneal shunt.

Periventricular leucomalacia is caused by ischaemic damage to the periventricular white matter. It is less common than IVH, but is the commonest cause of cerebral palsy in surviving infants. PVL is particularly common where there has been perinatal infection, severe hypotension or in monozygotic twins that share a placental circulation. The prognosis is worse if cystic change develops, with 80% developing cerebral palsy.

KEY POINTS

- 7% of infants are born before <37 weeks. 1% are extremely premature.
- Complications are related to organ immaturity and include hypothermia, hypoglycaemia and RDS.
- RDS is due to surfactant deficiency, and can be prevented by giving corticosteroids antenatally.
- Premature babies are at increased risk of cerebral palsy due to intracranial haemorrhage and leucomalacia.

50 Neonatal jaundice

Causes of jaundice in the neonatal period

Unconjugated hyperbilirubinaemia

Prematurity
- Immature liver enzymes

Rhesus incompatibility
- If mother is Rh negative and baby Rh positive, then maternal IgG can cause haemolysis
- Sensitization occurs in earlier pregnancies
- If severe can cause hydrops in utero
- Coomb's test positive

ABO incompatibility
- Usually milder than Rhesus

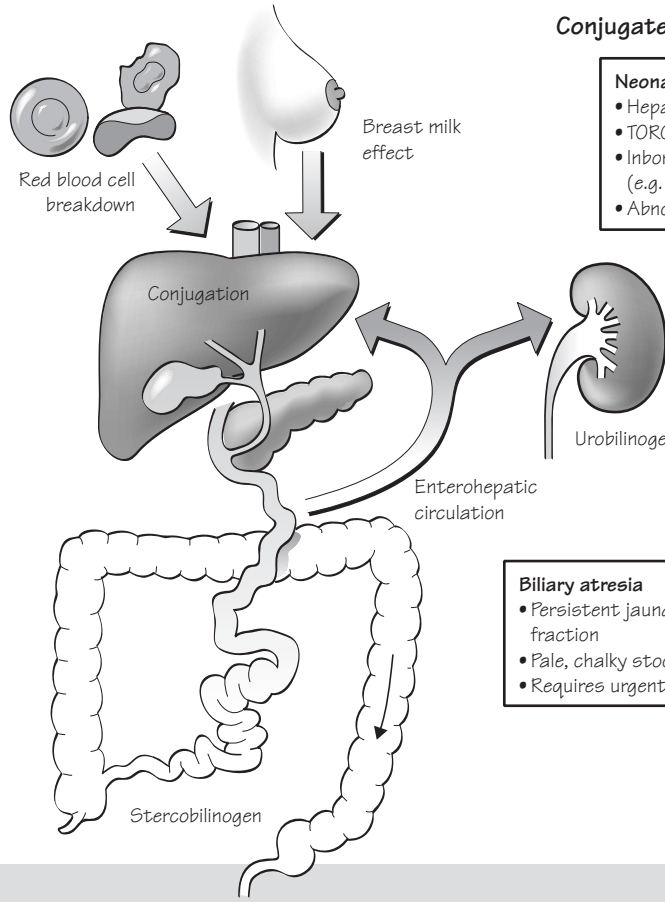
Infection
- Bacterial infection

Bruising
- Skin or scalp bruising from traumatic delivery is broken down into bilirubin

Hypothyroidism
- May be associated with pituitary disease

Breast milk jaundice
- Well baby who is breast-fed
- Jaundice develops in second week

Physiological
- Low liver enzyme activity
- Breakdown of fetal haemoglobin



Conjugated hyperbilirubinaemia

Neonatal hepatitis
- Hepatitis A,B
- TORCH infection
- Inborn errors of metabolism (e.g. galactosaemia)
- Abnormal liver function tests

Cystic fibrosis
- Cholestasis

Choledocal cyst

Biliary atresia
- Persistent jaundice with rising conjugated fraction
- Pale, chalky stools
- Requires urgent referral

What you need from your evaluation

History
- At what age did the jaundice develop? (within 24 h of birth always requires investigation)
- Are there any risk factors for infection?
- Is there a family history (cystic fibrosis, spherocytosis)?
- Is the baby active, alert and feeding well or lethargic and having to be woken for feeds (significant jaundice)

Examination
- What is the extent of the jaundice? (it tends to spread from the head down as it becomes more significant)
- Are there other features of TORCH infection, such as petechiae, anaemia or hepatosplenomegaly?
- Is the baby dehydrated? This exacerbates the jaundice
- Is the baby well or are there signs of infection?

Investigations and their significance

Split bilirubin	Total bilirubin and conjugated fraction (should be <20%)
FBC	Thrombocytopenia suggests TORCH infection Anaemia in haemolytic disease Neutropenia or neutrophilia in infection
Group and Coomb's	ABO and Rhesus incompatibility
Thyroid function	Hypothyroidism
TORCH screen	Hepatitis B, cytomegalovirus infection
Urine metabolic screen	Inborn errors of metabolism
Liver ultrasound	To visualize biliary tree
Infection screen	Urine, blood and cerebrospinal fluid
Liver isotope scan	To rule out biliary atresia in persistent conjugated hyperbilirubinaemia

Management
- Identify the cause and severity of the jaundice
- Use phototherapy to bring down the bilirubin level
- In severe haemolytic disease, multiple exchange transfusions may be required to prevent kernicterus
- Management of conjugated jaundice depends on cause but refer early to hepatologist if biliary atresia is suspected
- Prolonged jaundice can increase the chance of bleeding disorders associated with vitamin K deficiency. Check clotting and give further vitamin K supplements

Jaundice is the yellow pigmentation of the skin that occurs when plasma bilirubin levels are high. The bilirubin is formed from the breakdown of haem in red blood cells and is transported to the liver as unconjugated bilirubin, bound to albumin. In order to be excreted it needs to be made water soluble by conjugation in the liver. Conjugated bilirubin is then excreted in the bile into the duodenum, where some is reabsorbed (the entero-hepatic circulation) and the remainder forms stercobilinogen, which gives the stools their yellow/brown pigment. Some of the reabsorbed conjugated bilirubin is excreted by the kidneys as urobilinogen.

Excessive haemolysis or impaired conjugation leads to a build-up of unconjugated bilirubin, and obstruction to drainage of bile leads to conjugated hyperbilirubinaemia. Unconjugated bilirubin can cross the brain barrier. In high concentrations it is toxic to the basal ganglia and causes 'kernicterus' which used to be a common cause of athetoid cerebral palsy. Kernicterus is now extremely rare due to better obstetric management of Rhesus disease, careful monitoring of bilirubin levels and early treatment with phototherapy.

Treatment

Phototherapy (blue light at 450 nm wavelength) helps convert unconjugated bilirubin to an isomer which can be excreted by the kidneys. In Rhesus or ABO incompatibility, if bilirubin levels rise significantly, despite phototherapy, then an exchange transfusion is required to remove the bilirubin and the maternal IgG antibodies from the circulation.

Physiological jaundice

Jaundice in the neonatal period is very common and is usually due to a physiological immaturity of the liver. It is self-limiting as the liver matures over the first week, and only occasionally needs phototherapy treatment. All premature infants become jaundiced in the first few days of life, due to the immature hepatocytes not being able to adequately conjugate bilirubin. This never requires exchange transfusion but may require phototherapy for a few days.

Haemolytic disease of the newborn

Haemolysis occurs when maternal IgG antibody crosses the placenta and reacts with antigens on the fetal red blood cells. The commonest causes of this are ABO or Rhesus incompatibility. In Rhesus disease the fetus is Rhesus positive and the mother Rhesus negative. The mother will have been sensitized in a previous pregnancy by the passage of fetal red blood cells into her circulation, either at delivery or during a threatened miscarriage. Rhesus negative women are now routinely immunized with anti-D antibody, to 'mop-up' fetal red blood cells before they stimulate maternal IgG production. Haemolysis in the fetus causes anaemia which can lead to hydrops (severe oedema). After birth affected fetuses are anaemic and rapidly develop severe jaundice. The management of Rhesus or severe ABO incompatibility is to deliver the baby before severe haemolysis has occurred and then to wash out the maternal antibodies (and the bilirubin) by performing a series of exchange transfusions, and also by the use of phototherapy.

Biliary atresia

Biliary atresia is a rare but important condition caused by the absence of intra- or extra-hepatic bile ducts. An conjugated hyperbilirubinaemia develops over a period of weeks, and the stools become clay coloured. If undiagnosed the baby will develop liver failure and may die without a transplant. If detected within the first 6 weeks then a hepatoportoenterostomy (Kasai procedure) can achieve adequate biliary drainage. Because of this it is recommended that any baby still jaundiced after 2 weeks has conjugated and unconjugated bilirubin levels checked.

Jaundice in older children

Jaundice is rare in older children. The commonest cause is hepatitis A infection. Other causes include chronic haemolysis due to hereditary spherocytosis or glucose-6-phosphate dehydrogenase (G6PD) deficiency, or liver disease such as autoimmune chronic hepatitis. Reye syndrome, an acute encephalopathy associated with fulminant liver failure, can be induced by aspirin, and this is therefore contraindicated in young children.

KEY POINTS

- Mild jaundice is extremely common, especially in preterm infants.
- Jaundice within the first 24 h or lasting beyond 2 weeks needs investigation.
- Phototherapy and occasionally exchange transfusion are used to treat significant jaundice.
- Biliary atresia causes an obstructive persistent jaundice with pale stools. Early treatment is essential.

51 Congenital heart disease

Presentation of congenital heart defects

Presenting with heart failure

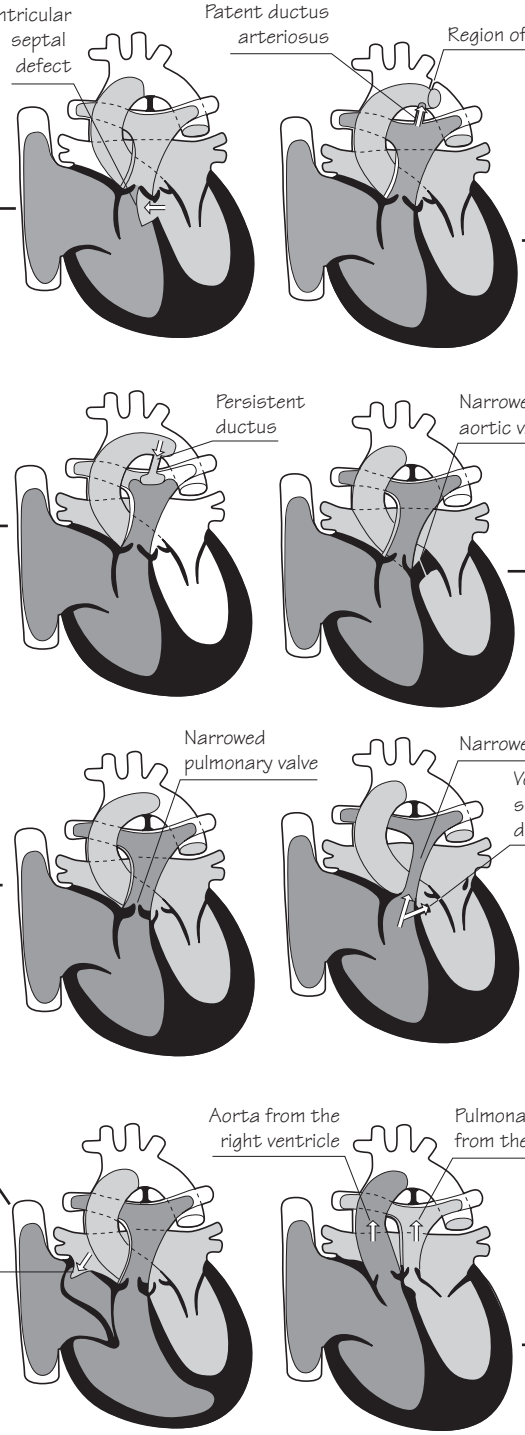
Ventricular septal defect (VSD)
- 32% of congenital heart disease (CHD) (commonest)
- Membranous or muscular
- Can be asymptomatic
- Harsh pansystolic murmur at lower left sternal edge
- Parasternal thrill
- Heart failure at 4–6 weeks

Patent ductus arteriosus (PDA)
- 12% of CHD
- Continuous machinery murmur below left clavicle
- Collapsing pulse
- More common in premature infants

Presenting with a murmur
(see Chapter 43)

Pulmonary valve stenosis
- 8% of CHD
- If critical stenosis, may be cyanosed at birth and duct dependent
- Ejection systolic murmur in pulmonary area radiating to back
- Evidence of right ventricular hypertrophy on ECG

Atrial septal defect (ASD)
- 6% of CHD
- Ostium secundum (defect of foramen ovale and atrial septum)
- Ostium primum (defect of atrioventricular septum)
- Usually asymptomatic at birth
- Fixed split second heart sound
- Pulmonary flow murmur
- Recurrent chest infections
- Heart failure



Presenting with shock

Coarctation of aorta
- 6% of CHD
- If severe, presents as shock when duct closes in first few days of life
- May present with heart failure and a murmur over the back
- Causes hypertension in older children
- Weak or absent femoral pulses
- Associated with Turner's syndrome

Aortic valve stenosis
- 5% of CHD
- If severe, presents as shock or heart failure when duct closes
- May present as asymptomatic murmur
- Older children may have syncope or reduced exercise tolerance
- Ejection systolic murmur radiating to neck

Presenting with cyanosis

Tetralogy of Fallot
- 6% of CHD
- Ejection systolic murmur
- Intermittent cyanosis from birth
- Right ventricular hypertrophy on ECG
- Boot-shaped heart on chest X-ray

Transposition of the great arteries
- 5% of CHD
- Separate pulmonary and systemic circulations, connected only by PDA (or sometimes VSD)
- Severe cyanosis and acidosis at birth
- Requires atrial septostomy urgently followed by definitive 'Switch' operation

Rarer forms of congenital heart disease

- Atrioventricular septal defect (AVSD): especially common in Down syndrome (40%)
- Interrupted aortic arch
- Hypoplastic left heart syndrome
- Peripheral pulmonary stenosis: commoner in Noonan syndrome
- Supravalvular aortic stenosis: with Williams syndrome
- Tricuspid atresia
- Ebstein's anomaly: in infants exposed to lithium
- Total anomalous pulmonary venous drainage (TAPVD): the only lesion to present with cyanosis and heart failure

Investigations and their significance

• Pulse oximetry	To determine degree of cyanosis
• Chest X-ray	Cardiomegaly in heart failure Boot-shaped heart (Fallot's tetralogy) Increased vascular markings with left to right shunts (VSD, ASD, PDA)
• ECG	Right ventricular hypertrophy Superior QRS axis (AVSD, primum ASD)
• Echo	Ultrasound examination of the heart, usually performed by a paediatric cardiologist, can diagnose the vast majority of congenital heart defects
• Fetal Echo	Many defects can be detected antenatally
• Cardiac catheter	To define very complex anatomy

Congenital heart disease (CHD) is the most common malformation, with 7–8 per 1000 liveborn infants having a significant cardiac abnormality. About 8% are associated with chromosomal abnormality (e.g. AVSD in Down syndrome) or more subtle genetic abnormalities, such as a deletion at chromosome 22q11 which is associated with aortic arch defects and hypocalcaemia (diGeorge syndrome). The risk of CHD is higher if there is a family history. Teratogens may cause CHD (e.g. VSD and tetralogy of Fallot in fetal alcohol syndrome).

It is increasingly common for some heart defects to present antenatally, with abnormalities detected on ultrasound. Others present at birth, with cyanosis (e.g. transposition of the great arteries) or shock (hypoplastic left heart syndrome). Some duct-dependent lesions will present when the arterial duct starts to close within the first few days of life (e.g. coarctation of the aorta, critical pulmonary stenosis). Defects with a left to right shunt such as a VSD often present with heart failure and difficulty feeding some weeks after birth. Finally, some lesions may be asymptomatic and are first detected as a heart murmur (e.g. atrial septal defect, aortic stenosis) (see Chapter 43).

Medical management involves the use of diuretics and other drugs to control heart failure, pending definitive surgical repair. Many heart defects can just be monitored for years, and may not need surgical correction. It is important to prevent of bacterial endocarditis.

Coarctation of the aorta

This is a narrowing of the aorta. Severe coarctation will present in the first few days of life when the arterial duct closes and insufficient blood is able to reach the lower limbs and perfuse vital organs such as the kidneys, causing circulatory collapse and acidosis. The key feature is weak or impalpable femoral pulses. The blood pressure may be higher in the arms than the legs and oxygen saturation will be lower in the legs. Milder forms of coarctation will present with heart failure and a murmur or with hypertension in a young adult. The management is IV prostaglandin E2 to keep the duct patent. Once the diagnosis is confirmed, by echocardiography, the narrowed segment is repaired surgically or dilated using a balloon.

Transposition of the great arteries (TGA)

In TGA, the aorta and main pulmonary artery are transposed, so that the aorta comes off the right ventricle and the pulmonary artery off the left ventricle. It always presents soon after birth with profound cyanosis and acidosis. The only way oxygenated blood from the lungs can reach the systemic circulation is across the arterial duct, or a VSD, if present. The emergency management of TGA is to commence a prostaglandin infusion, provide ventilatory and circulatory support and perform an atrial septostomy which allows mixing of oxygenated and deoxygenated blood. The definitive treatment is surgical correction—the Switch operation, where the two great vessels are switched over and the coronary arteries connected to the new aorta.

Tetralogy of Fallot

This is the commonest cyanotic CHD. The 'tetralogy' refers to a large VSD, an aorta that sits over the ventricular septum, pulmonary infundibular stenosis and right ventricular hypertrophy. Some will present with cyanosis at birth but others may be diagnosed by an ejection systolic murmur. Classically these children develop hypercyanotic 'spells' which are relieved by squatting down (to reverse the right to left shunt by increasing left ventricular pressure). Chest X-ray may show a 'boot-shaped' heart. Surgical correction is usually performed at 2–3 months of age.

Patent ductus arteriosus (PDA)

During fetal life the ductus arteriosus (arterial duct) shunts blood from the pulmonary artery to the aorta, bypassing the unexpanded lungs. In normal babies the duct closes within a few days of birth, first by constriction and then by fibrosis. In sick preterm babies, or those exposed to hypoxia, the duct remains open. As the right-sided pressures are now less than the aortic pressure, blood shunts from left to right and cardiac failure and pulmonary oedema may develop. The clinical signs of a PDA are a continuous 'machinery' murmur below the left clavicle and collapsing pulses. A patent arterial duct is particularly common in premature infants receiving mechanical ventilation. Management involves diuretics and fluid restriction, and sometimes administration of prostaglandin synthetase inhibitors. Rarely the duct needs to be closed surgically by ligation or insertion of an occlusion device.

KEY POINTS

- CHD is the commonest congenital abnormality.
- The key to diagnosis is understanding the different modes of presentation.
- Duct-dependent defects will present in the first days of life and need urgent treatment with prostaglandin to keep the duct open.
- With good management and corrective surgery the prognosis for most CHD is excellent.

52 Asthma

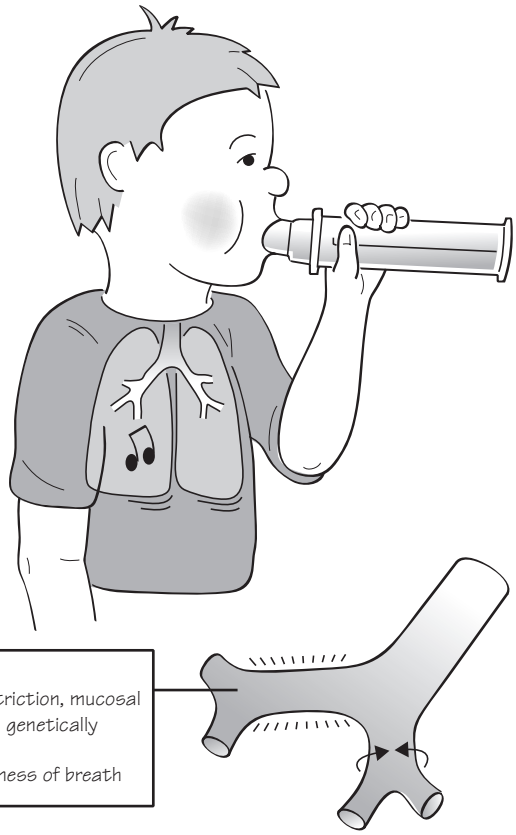
Chronic asthma

Cough
- Recurrent dry cough
- Worse at night
- Worse with exercise

Wheeze
- Expiratory noise due to airway narrowing
- Often triggered by viral infections
- Responds to bronchodilators

Shortness of breath
- Exercise limitation
- Triggers can be exercise, cold, allergens, smoke

Uncontrolled asthma
- Poor growth
- Chronic chest deformity
- Time off school
- Frequent acute exacerbations



Pathology
- Environmental triggers cause bronchoconstriction, mucosal oedema and excess mucous production in a genetically predisposed child
- Airway narrowing causes wheeze and shortness of breath

Acute asthma

Acute asthma attack
- Acutely short of breath
- Cough and wheeze
- Work of breathing increased
- Child often frightened
- May be triggered by viral illness, exposure to allergens, exercise or cold air

Assessing severity

Mild
- Breathless but not distressed
- Peak expiratory flow rate (PEFR) reduced but still >50% of normal

Severe
- Too breathless to talk or feed
- Respiratory rate >50 breaths/min, pulse >140 beats/min
- PEFR <50% of expected

Life-threatening
- PEFR <33% of expected
- 'Silent chest' or cyanosis
- Fatigue, drowsiness

What you need from your evaluation

History

- Ask about the cough and wheeze. What triggers it and at what time of day does it occur?
- How many acute exacerbations have there been? How severe was the worst attack?
- How does the asthma affect the child's life? Does it limit activities; have they missed any school?
- How often has the child had to use reliever treatment? How effective was it?
- Are there other atopic symptoms such as hay fever (allergic rhinitis) or eczema or a family history of atopy?

Investigations and their significance

- PEFR — Peak expiratory flow rate is best recorded in a peak flow diary to monitor change over time
- Chest X-ray — To exclude pneumothorax in severe asthma. Avoid excessive X-rays
- Allergy tests — Skin prick or serum radio-allergosorbent tests (RAST) if history suggests a specific allergic trigger (e.g. cat dander)

Examination

- In well-controlled asthmatics there may be no physical signs between acute exacerbations
- Listen for wheeze. Beware the 'silent chest' of severe asthma when there is almost no air moving
- Look for chronic chest deformity: barrel chest and Harrison's sulcus in severe uncontrolled asthma
- Measure PEFR using handheld peak flow meter
- Check height and weight and plot on centile chart. Poorly controlled asthma will stunt growth, as will overuse of oral corticosteroids
- Check inhaler technique periodically

Management

- **Medication:** 'preventers' (inhaled steroids or sodium chromoglycate) and 'relievers' (bronchodilators)
- **Environmental control:** avoid passive smoking and reduce house dust mite exposure if possible
- **Self-monitoring of disease severity:** PEFR and symptom diary, management plan for each child
- **Education:** of the child, the family and the school on good control of asthma, inhaler technique and emergency treatment of an acute exacerbation

Asthma is the commonest chronic illness of childhood, occurring in up to 15% of all children at some point. The incidence has risen significantly in recent years, partly due to better recognition but possibly also due to environmental pollutants. The symptoms of cough, wheeze and dyspnoea are due to narrowing of the bronchi and bronchioles by bronchoconstriction, mucosal inflammation and thick mucous obstructing the lumen. In a susceptible individual this process is initiated by environmental factors such as dust mite allergens, air pollution, cigarette smoke, cold air, viral infections, stress and exercise.

Presentation

Children with asthma usually present in infancy or early childhood. This diagnosis is clinical, based on recurrent cough or wheeze that responds to bronchodilator treatment. A history of atopy (eczema or hay fever) or a family history of asthma supports the diagnosis. In infancy it is often unclear whether recurrent wheeze is the first manifestation of asthma or merely airway obstruction associated with viral respiratory tract infections such as RSV or adenovirus. As the airways are narrow, mucous and mucosal oedema contribute more to obstruction than bronchoconstriction, and there may be poor response to bronchodilators. If an infant is atopic, or there is a strong family history then it is more likely that these wheezy episodes are an early presentation of asthma.

In older children recurrent episodes of wheeze and cough, especially if triggered by exercise, viral infections or allergens, suggest a diagnosis of asthma. A good response to bronchodilator therapy, either in symptom reduction or improvement in peak expiratory flow rate (PEFR) confirms the diagnosis. A CXR should always be obtained at first presentation to exclude other pathology such as inhaled foreign body. In asthma the CXR may show hyperinflation (due to air-trapping) and areas of collapse (due to mucous plugging). RAST may be helpful in some cases.

Management of chronic asthma

The goal of good asthma management is to relieve the symptoms and allow normal activity, school attendance and growth. A stepwise approach is used—increasing the amount of treatment until control is obtained, then stepping back to the minimum treatment required to maintain good symptom control.

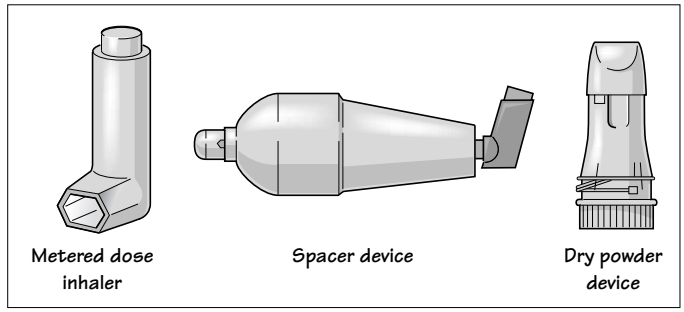
Step 1—Inhaled beta$_2$-agonists (e.g. salbutamol), as required up to three times a week. This is known as 'reliever' medicine. Check the delivery device is appropriate (see below) and the technique is correct.

Step 2—Add in a regular 'preventer' medicine—either sodium chromoglicate or low-dose inhaled corticosteroids. Continue the beta$_2$-agonists as required.

Step 3—Inhaled beta$_2$-agonists as required plus higher dose regular inhaled corticosteroid. Consider a long-acting beta$_2$-agonist (e.g. salmeterol) or modified release theophylline.

Step 4—Inhaled beta$_2$-agonists as required plus regular high-dose inhaled steroid *and* consider a short course of oral steroids.

Step 5 (only in older children)—Inhaled beta$_2$-agonists as required plus regular high-dose inhaled steroid and regular *oral* steroids (but there is a serious risk of affecting growth with systemic steroids).



Inhaler devices

Treatment will only be effective if the drug is delivered in sufficient quantity to the small airways of the lungs. This is best achieved using an aerosolized drug delivery system, via a metered dose inhaler (MDI). One 'puff' of the MDI delivers a known dose of the drug. A high degree of coordination is required to activate the MDI during inhalation, and this method is really only suitable for teenagers. Some MDIs are breath-activated.

For children under 8 years, the MDI can be used in conjunction with a spacer device (e.g. aerochamber, volumatic), so that the child breathes in and out of a chamber containing the aerosolized drug. In infants these should be fitted with a mask to place over the child's mouth and nose.

In children over 8 years, a dry powder device (e.g. Turbohaler, Accuhaler) is used—the child sucks in a fine powder, containing the drug, during inspiration. These devices deliver the required dose more reliably and are easier to use in this age group than MDIs. Nebulizers can be used in infants and for emergency treatment of acute exacerbations although there is evidence that MDIs via a spacer are more effective.

Treatment of severe exacerbation

Acute exacerbations should be treated promptly at home by using more of the reliever medication. If the symptoms continue or worsen then aggressive treatment with high flow oxygen, regular beta$_2$-agonists or ipratroprium bromide via a nebuliser or spacer device (e.g. 15 puffs) and systemic corticosteroids is indicated. In life-threatening asthma (see opposite) an infusion of salbutamol or aminophylline is used. All children should have oxygen saturations measured and be admitted for close observation.

KEY POINTS

- Asthma is the commonest chronic childhood illness, occurring in 10–15% of all children.
- Bronchoconstriction, viscid mucous and mucosal oedema causes airway narrowing with wheeze, cough and dyspnoea.
- Treatment is increased and decreased step-by-step to gain symptom control and maintain a normal lifestyle.
- It is crucial to use an inhaler device suitable for the child's age.

53 Diabetes

Aetiology
- 1 in 500 children
- Destruction of beta islet cells in pancreas leads to insulin deficiency
- Genetic predisposition plus infection

Initial presentation
- Polyuria, polydipsia and weight loss over a few weeks
- Polyuria may cause enuresis
- Diagnosis made by finding high blood sugar and glycosuria (glucose tolerance test rarely needed)

Poor diabetic control
- Polydipsia, polyuria
- Hypoglycaemic episodes
- Poor growth
- Hyperglycaemia, high HbA1c
- Lipodystrophy if inadequate rotation of injection sites

Hypoglycaemia
- The result of excess insulin or inadequate carbohydrate intake
- Feel hungry and shaky
- Pale, sweating, tremors
- Tachycardic
- Drowsy or irritable
- Convulsions or coma
- Hypoglycaemia on testing
- May get rebound hyperglycamia afterwards
- Requires urgent treatment

Diabetic ketoacidosis (DKA)
- May be triggered by infection or poor compliance
- Thirst and polyuria
- Vomiting
- Abdominal pain
- Acetone smell on the breath
- Very high blood sugar level, ketonuria
- Metabolic acidosis on blood gas
- Urea raised, electrolytes disturbed
- Signs of dehydration
- Kussmaul acidotic breathing
- Hypovolaemic shock, drowsiness and coma if not treated urgently
- Requires urgent treatment with fluids and insulin

Ongoing management
- Requires multidisciplinary approach
- Initial correction of metabolic state and education of family
- Treatment with insulin and specific dietary advice
- Monitoring blood sugar levels at home and HbA1c regularly
- Education about diabetic control, injection technique, diet.
- Dealing with emergencies and liaison with school

Prognosis
- No cure. Insulin controls the metabolic disturbance but good control is mandatory to prevent long-term complications in adult life
- Retinopathy, neuropathy, renal impairment and atherosclerosis are the long-term effects of poor control of blood glucose levels

What you need from your evaluation

History

- Ask about polyuria, polydipsia, lethargy and weight loss
- Ask about bedwetting (secondary eneuresis)
- Review the diabetic diary and ask about hypoglycaemic and hyperglycaemic episodes —what triggered them and were they managed appropriately?
- How is the child coping at home and at school? Also ask about siblings
- Is the child managing to eat a healthy diet and modify the diet to certain situations (e.g. snacks before heavy exercise)
- Is insulin being administered correctly with rotation of injection sites?

Examination

- Monitor height and weight as poor growth reflects poor control
- Check for signs of lipodystrophy or lipoatrophy at injection sites
- Check blood pressure annually and fundi in older children (>14 years)
- Check for signs of coexistent coeliac disease or hypothyroidism

Investigations and their significance

• Blood glucose	Monitor regularly at home using finger-prick samples and handheld glucometer
• HbA1c (% of glycosylated haemoglobin)	Reflects control over last 2–3 months
• Urinalysis	For glycosuria, ketones, proteinuria
• Blood gases, U&E	Need to be monitored carefully during acute diabetic ketoacidosis
• Anti-endomysial antibody	Screen for coeliac disease
• Thyroid function tests	Screen for hypothyroidism

Diabetes mellitus

Diabetes affects more than 1 in 500 children. The diagnosis has a major impact on the child and the family in terms of their daily life: the possibility of emergencies such as hypoglycaemia and diabetic ketoacidosis (DKA), and the fear of long-term complications such as retinopathy, renal failure, cardiovascular disease and neuropathy. Diabetes in children is usually insulin-dependent diabetes mellitus (IDDM) due to destruction of the beta cells in the islets of Langerhans in the pancreas, resulting in a lack of insulin. The islet cell destruction is genetically predisposed but probably triggered by a viral illness. The lack of insulin means that glucose cannot be utilized, resulting in hyperglycaemia. The high glucose concentration in the blood spills over into the urine, causing an osmotic diuresis with polyuria and dehydration. This leads to excessive thirst, and weight loss. Because the cells cannot utilize glucose they switch to metabolizing fats, leading to the production of ketones and acidosis. Maintenance treatment of diabetes therefore requires regular administration of insulin to allow normal glucose metabolism.

Initial presentation of diabetes

Children usually present with a short (2–3 week) history of lethargy, weight loss, polyuria and thirst. The polyuria may cause a recurrence of bedwetting. If the symptoms are not recognized the child may develop signs of diabetic ketoacidosis with abdominal pain, vomiting and eventually coma. Newly diagnosed children are usually admitted to hospital to start their insulin treatment and to allow intensive education of the child and family, although more and more this is undertaken by a diabetic nurse specialist who visits the family at home. The family needs to be taught how to inject the insulin, how to monitor blood glucose and test urine for ketones, and how to recognize the signs of hypoglycaemia. They also need advice from a dietician, although the diet should not be over-restrictive, but a healthy balanced diet high in fibre. Children are encouraged to wear a medic-alert bracelet, giving details of their condition.

Growing up with diabetes

The education given to families at the time of diagnosis is crucial to them developing the right approach to their child's diabetes. They need to be careful without being over-restrictive. As the child gets older they can gradually take on more of the responsibility themselves, including injecting insulin and monitoring blood glucose levels. Food intake should not be restricted so that normal growth can occur. Snacks needs to be given as slowly absorbed foods, such as cereal bars or biscuits, rather than chocolate and sweets. Many children go through a 'honeymoon' period soon after diagnosis where they need very little insulin, as they still have some endogenous insulin. More insulin is often required as they go through the pubertal growth spurt.

Insulin therapy

Insulin is usually delivered by an injection pen which injects a predetermined dose of insulin with each activation. The insulin is given as a mixture of short-acting (peak at 2–4 h) and intermediate-acting insulin (peak at 4–12 h). For example, Mixtard-30 or Humulin M3 contain 30% short-acting and 70% long-acting insulin. This is administered subcutaneously in the arms, thighs, buttocks or abdomen, before breakfast and before the evening meal. If injection sites are not rotated regularly then lipodsytrophy and lipoatrophy can occur, leading to erratic absorption of insulin. About two-thirds of the insulin is given in the morning and one-third in the evening, to cover the increased carbohydrate intake during the day and prevent nocturnal hypoglycaemia. Occasionally insulin is given four times a day to achieve tighter control.

Monitoring

Control is assessed by keeping a blood sugar diary and measuring HbA1c levels, which reflect the degree of hyperglycaemia in the preceding few months. The family need to be warned of the symptoms of hypoglycaemia (see opposite) and have a supply of sugary drinks, glucose gel or glucagon injections available at all times. As children become adolescents they should be gradually integrated into the adult diabetic service. At this stage, if not already, they will begin screening for retinopathy and renal disease.

Diabetic ketoacidosis

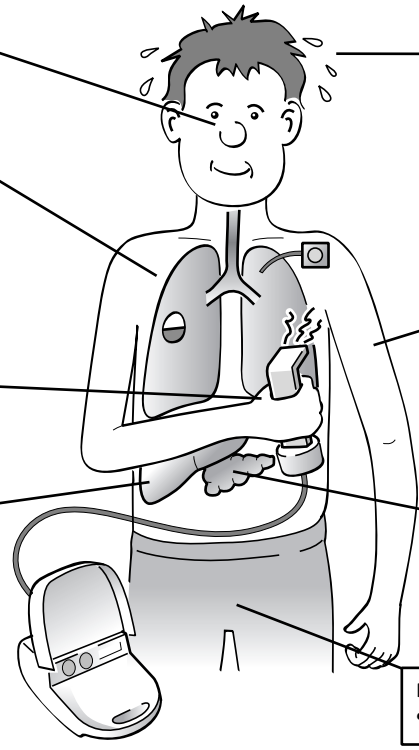
DKA occurs when there is inadequate insulin to meet the metabolic demands. This is due to either poor compliance with insulin treatment, a common problem in adolescence, or to an intercurrent illness which increases insulin demands. A common mistake is to give less insulin because a child is ill, not eating, and vomiting. In fact in this situation they will often need more insulin than usual.

The lack of insulin leads to hyperglycaemia and dehydration due to an osmotic diuresis. A switch to fat metabolism leads to ketone production, resulting in acidosis, vomiting and abdominal pain and gastric stasis. If not treated the child will become comatose. Treatment involves treatment of shock, then gradual rehydration over 24 h. An insulin infusion is used to gradually bring down the blood glucose; at this point some dextrose should be given to allow the cells to switch back to metabolizing glucose. Serum electrolytes and fluid balance must be checked regularly. There is a danger of hypokalaemia as insulin pushes potassium into the intracellular compartment. A naso-gastric tube should be passed to empty the stomach, as there is often gastric paresis. Infection should be sought and treated. Once the child is eating and drinking they can be switched back to their normal insulin.

KEY POINTS

- Diabetes is very common: it occurs in 1 in 500 children.
- Presentation is with a short history of weight loss, polyuria and polydipsia.
- DKA can be life-threatening and needs careful treatment with fluids and insulin.
- Insulin is given subcutaneously as a mixture of short- and long-acting insulin.
- Patients must be able to recognize and treat hypoglycaemia.

54 Cystic fibrosis



Ear, nose and throat
- Nasal polyps

Recurrent chest infections
- Cough
- Purulent sputum
- Pneumonia
- Chronic pseudomonas infection
- Bronchiectasis
- Chest deformity
- Eventual respiratory failure

Finger clubbing
- Seen with chronic lung infection

Liver disease
- Obstructive jaundice in neonatal period (rare)
- Biliary stasis may require treatment with ursodeoxycholic acid
- Eventually liver cirrhosis may develop

High salt losses in sweat
- Salty taste to skin
- Risk of salt-losing crisis during very hot weather

Poor growth
- Require 40% extra energy intake compared with normal child
- Poor weight gain
- Short stature
- Normal growth is achievable with pancreatic replacement, physiotherapy and aggressive treatment of chest infections

Pancreatic insufficiency
- Poor fat absorption
- Steatorrhoea (fatty stools)
- Distended abdomen
- Rectal prolapse
- Meconium ileus at birth (15%)
- Meconium ileus equivalent—can mimic acute appendicitis
- Need to take pancreatic enzymes with meals
- May develop diabetes

Male infertility
- Congenital absence of the vas deferens

Genetics and aetiology

- Commonest serious autosomal recessive condition in Caucasian population
- Affects 1 in 2500 children; 1 in 25 are carriers
- CFTR (cystic fibrosis transmembrane regulator) protein is coded for on chromosome 7. Commonest CFTR mutation (78%) is ΔF508
- Abnormal CFTR protein leads to poor function of epithelial membrane chloride channel with viscid secretions that obstruct pancreatic exocrine ducts and bronchioles
- Abnormal sweat gland function leads to high concentrations of sodium and chloride in the sweat

Investigations and their significance

- **DNA:** Analysis of the CFTR gene may reveal a common mutation (screens for only 30 of 800 mutations)
- **Immunoreactive trypsin (IRT):** Level is high in neonates with cystic fibrosis and is used for newborn screening of 'Guthrie' cards
- **Sweat test:** High concentrations of sodium (>60 mmol/l) and chloride can be measured. This is the basis of the diagnostic test for cystic fibrosis. A minimum of 100 mg sweat is needed
- **Faecal chymotrypsin** and **faecal elastase** levels are low in cystic fibrosis. Faecal fat concentrations can be used to monitor pancreatic enzyme supplementation
- **Lung function tests:** Will show a restrictive and obstructive pattern.
- **CXR:** May show signs of consolidation. With chronic pseudomonas infection cystic changes become evident

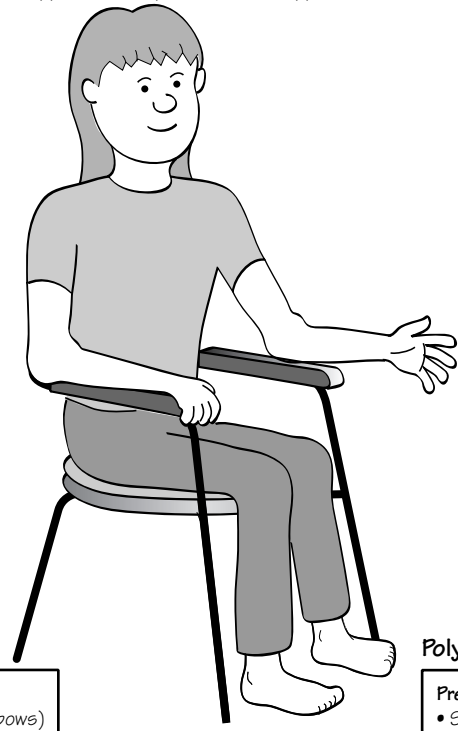
Management

- There is currently no cure for cystic fibrosis. Gene therapy trials are ongoing but results have been disappointing
- Management should involve a specialist multidisciplinary team of respiratory paediatrician, physiotherapist, dietician, specialist nurses and psycho-social support. Treatment should be delivered at home whenever possible
- Parents are taught chest physiotherapy, which must be performed regularly to clear secretions
- Antibiotics are often given prophylactically to prevent chest infections. When infections occur they are treated aggressively, and once colonized with *Pseudomonas aeruginosa* the child may require regular courses of intravenous and nebulized antibiotics. These are often given at home via an indwelling central venous catheter (e.g. portacath)
- Pancreatic enzyme capsules containing lipases, amylases and proteases are taken with meals to aid absorption
- Dietary supplements may be needed to provide sufficient calorie intake. Energy requirements can be 140% of normal due to recurrent infection, coughing and malabsorption. Supplements of fat-soluble vitamins are required
- Heart–lung transplant is offered when the disease has progressed to the end stage. The heart from the cystic fibrosis patient can be used in another recipient (the domino transplant)
- Life expectancy has improved considerably but the median survival is still only 32 years

55 Juvenile chronic arthritis

Juvenile chronic arthritis is a group of conditions that present in childhood with joint swelling lasting more than 6 months. The classification has recently been revised (see box below) but is based on the way the condition initially presents. Treatment is aimed at reducing symptoms of pain and preserving joint function and mobility. In the majority of cases the symptoms resolve but in about 25% of those with the systemic form, the disease continues into adulthood and can be disabling. A multidisciplinary approach with psychological support is necessary for these children.

Classification
- Systemic (Still's disease): 9%
- Polyarticular: 19%
- Oligoarticular (″4 joints): 49%
- Enthesitis associated (HLA B27): 7%
- Juvenile psoriatic arthritis: 7%
- Other



Systemic onset (Still's disease)

Presentation
- Spiking fever, severe malaise
- Salmon-pink rash
- Anaemia, weight loss
- Hepatosplenomegaly
- Arthralgia and myalgia but may have minimal joint symptoms
- May resemble malignancy

Features
- Both large and small joints affected
- 25% have severe arthritis
- Rheumatoid factor negative
- Tempero-mandibular joint may be involved, causing micrognathia

Prognosis
- In 25%, arthritis persists into adulthood with disability
- May require joint replacements

Pauciarticular (up to four joints)

Presentation
- Usually affects large joints (knees, ankles, elbows)
- Commonly affects girls under the age of 4 years
- Minimal systemic symptoms

Features and prognosis
- Rheumatoid factor negative, antinuclear antibody (ANA) may be positive
- High risk of chronic iridocyclitis (inflammation of the anterior structures of the eye). Regular examination with slit lamp is required to screen for this
- Arthritis usually resolves completely

Polyarticular (five or more joints)

Presentation
- Symmetrical involvement of large and small joints
- There may be poor weight gain and mild anaemia
- Morning stiffness
- Irritability in young children

Features and prognosis
- Rheumatoid factor negative in 97%, ANA may be positive
- No eye involvement
- 12% develop severe arthritis but prognosis is generally good

Management

The aims of management are to preserve joint function, to minimize complications, including complications of the treatment, and to aid the psychological adjustment to what can be a chronic disabling condition in some.

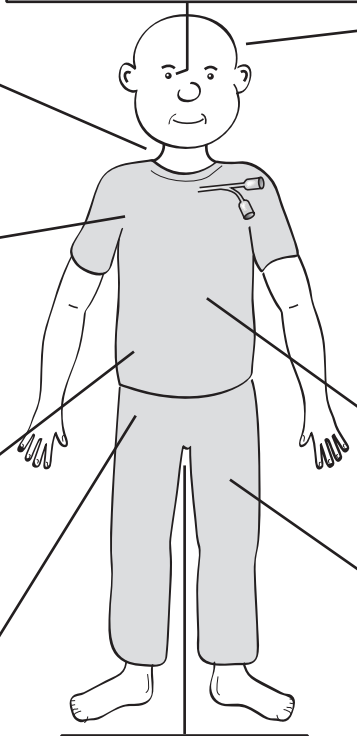
The aim is to reduce joint inflammation using non-steroidal anti-inflammatory drugs (NSAIDs). Steroids may be injected into affected joints, but cannot be used too frequently. Systemic steroids may be required for severe systemic disease. Other steroid-sparing therapies include hydroxychloroquine, penicillamine, gold injections, methotrexate and immune regulator drugs, which can be used in severe disease. Physiotherapy, hydrotherapy and wearing splints can help maintain joint function and mobility. Occupational therapy can help with aids to improve function. The family needs psychological support. Children with residual handicap may require help in planning a suitable career

Complications

Flexion contractures of the joint may develop without regular therapy and splinting. Joint destruction may require eventual joint replacement (e.g. knees, hips) in some children. Growth failure can occur due to the chronic illness, anorexia and the growth suppression effect of corticosteroid therapy. Patients may develop Cushingoid features. Chronic anterior uveitis (iridocyclitis) is asymptomatic but if missed can lead to visual impairment. Whilst the prognosis differs between subgroups, overall the prognosis is good and most children recover completely

56 Childhood cancer

Types of childhood cancer



Leukaemia
- Commonest childhood malignancy (35%)
- 80% acute lymphoblastic leukaemia
- Presents with
  - -malaise
  - -anaemia
  - -bruising
  - -bone pain
  - -lymphadenopathy
- Chemotherapy used to induce remission and prevent relapse
- Overall prognosis good (up to 80% survival)

Retinoblastoma
- Rare but important cause of blindness
- Presents within the early years
- White pupillary reflex or squint

Brain tumours
- Second commonest presentation of childhood cancer (25%)
- Usually primary brain tumour
- Present with raised intracranial pressure or neurological signs:
  - -headache
  - -nausea and vomiting
  - -blurred vision
  - -squint (VI nerve palsy)
  - -ataxia, clumsiness
  - -nystagmus
  - -endocrine dysfunction
- Most tumours occur in the brain stem or cerebellum
- Treatment involves neurosurgical resection and radiotherapy
- Long-term sequelae include endocrine and growth problems

Lymphoma
- Hodgkin's disease and non-Hodgkin's lymphoma (NHL)
- Usually present with lymphadenopathy
- Mediastinal lymph node involvement is common
- Diagnostic excision biopsy followed by chemotherapy

Neuroblastoma
- Usually in children under 5 years
- Arises in neural crest tissue (adrenal medulla and sympathetic nervous system)
- Presents with abdominal mass. Peri-orbital bruising or skin nodules can occur
- Increase in urinary catecholamines

Wilm's tumour (nephroblastoma)
- Presents with large renal mass
- Occasionally causes haematuria or hypertension
- Metastasize via IVC to the lungs
- Associated with aniridia (absent iris) coded for on chromosome 11

Bone tumours
- Usually occur in older children
- Present with bone pain or mass, usually in the long bones
- Ewing's sarcoma
- Osteosarcoma
- Treatment involves chemotherapy and surgery with bone prostheses

Rhabdomyosarcoma
- Arises in mesenchymal tissue
- Head and neck and genitourinary tract are the commonest sites
- 15% present with metastases

Germ cell tumours
- Sacrococcygeal tumour
- Gonadal tumours

Management of childhood cancer

- **Diagnosis**

  The diagnosis of cancer in childhood is devastating. Children should be referred to specialized paediatric oncology centres for optimum management, although shared care with the local hospital is often possible later
- **Treatment**

  The aim of treatment is eradication of the cancer, whilst minimizing damage to the normal tissues. Cancer therapy is toxic and the child requires intensive support treatment including prophylactic antibiotics and good nutritional support
  - **Surgery** is often required for diagnostic biopsy and excision of solid tumours, and for inserting indwelling central venous catheters necessary for chemotherapy
  - **Radiotherapy** is used to treat local disease and for total body irradiation in conjunction with bone marrow transplantation. Adjacent tissues are often damaged and there may be long-term effects on growth if the spine or pituitary gland is irradiated
  - **Chemotherapy** acts by killing cells during cell division. The aim is to kill the rapidly dividing malignant cells without killing normal cells. The drugs are usually given in combination at regular intervals. Side effects include hair loss, nausea, immunosuppression and bone marrow suppression. There is a particular risk of sepsis if the child becomes neutropenic, and any febrile episodes while the child is neutropenic should be treated aggressively with broad-spectrum intravenous antibiotics pending the results of blood and other cultures
  - **Bone marrow transplantation** involves either harvesting bone marrow or using compatible donated bone marrow to replace the patient's suppressed marrow; this allows more intensive chemotherapy to be used. This technique is being used more commonly and earlier in the disease. Side effects include severe immunosuppression and graft-versus-host disease

The incidence of malignant disease in childhood is about 1 per 10 000 children per year and causes 14% of all childhood deaths. The commonest malignancies are acute leukaemia, brain tumours and lymphoma. Overall, there has been a significant improvement in prognosis over recent years due to the use of well-researched and standardized chemotherapy regimes delivered in specialized paediatric oncology centres. The prognosis still depends largely on the particular type of malignancy and on the progression of the disease at the time of diagnosis.

Acute leukaemia

Leukaemia is due to the malignant proliferation of white cell precursors within the bone marrow. These 'blast' cells escape into the circulation and may be deposited in lymphoid tissue. The commonest type of leukaemia in childhood is acute lymphoblastic leukaemia (ALL), where the blast cells are precursors of lymphocytes. Acute myeloid leukaemia (AML) is commoner in Down syndrome. Chronic leukaemias are very rare in childhood.

ALL can occur at any age, but the peak is around 5 years. The prognosis is worse for those presenting under the age of 2 or over 10 years old. The onset is insidious with malaise, anorexia and then pallor, bruising or bleeding. Lymphadenopathy and splenomegaly may be present, and bone pain may occur. Peripheral blood usually shows anaemia, thrombocytopenia and a raised white cell count. Those with an extremely high white count ($>50\times10^9$/L) carry a worse prognosis. Blast cells may be seen on the peripheral blood film. The diagnosis is confirmed by a bone marrow aspirate, which shows the marrow infiltrated with blast cells. Cells are examined for immunological phenotyping and for cytogenetic analysis as these give important prognostic information. ALL can be subdivided into common (75%), T-cell (15%), null (10%) and B-cell (1%).

Treatment of ALL involves combination chemotherapy to *induce* remission (i.e. remove all blast cells from the circulation and restoration of normal marrow function). *Intensification* chemotherapy maintains remission, and methotrexate or cranial irradiation protects the CNS from involvement. Monthly cycles of *maintenance* chemotherapy are then given for up to 2 years. Prophylactic antibiotics (cotrimoxazole) are given to prevent opportunistic infections such as *Pneumocystis carinii*. Care must be taken to avoid live vaccines and contact with infections such as chicken pox. Children who relapse are often offered high-dose chemotherapy and bone marrow transplantation. The overall prognosis for acute leukaemia is good with up to 65% cured and 5-year survival approaching 80%.

Short-term side-effects of treatment

Tumour lysis syndrome. The breakdown of large numbers of malignant cells either before or during treatment can lead to very high serum urate, phosphate and potassium levels and urate crystals can precipitate in the kidneys causing renal failure. Tumour lysis syndrome can be prevented by good hydration and the use of allopurinol (a xanthine oxidase inhibitor).

Bone marrow suppression and febrile neutropenia. Bone marrow suppression may be due to invasion by tumour cells or the effect of chemotherapy. Anaemia and thrombocytopenia can be treated with infusions of red cells and platelets. Neutropenia (neutrophil count $<1.0\times10^9$) is difficult to treat and means the patient is at risk of serious infection. Consequently any significant fever (>38ºC) while neutropenic should be investigated and treated aggressively with broad-spectrum IV antibiotics until culture results are known.

Immunosuppression. Severe immunosuppression may result from treatment. This leaves the child at risk from normally trivial infections. Patients should not be given live vaccines and if exposed to varicella (chickenpox) should be given specific immunoglobulin. If the patient goes on to develop chicken pox they should be treated with acyclovir and immunoglobulin.

Inflammation. Inflammation of gut mucosa and mouth ulcers as well as anorexia can lead to poor calorie intake. Nutritional support with food supplements may be necessary.

Late sequelae of treatment

Short stature or asymmetrical growth may be caused by radiotherapy to the spine or pituitary fossa. The latter may also cause delayed puberty and other endocrine dysfunction including growth hormone deficiency, hypothyroidism and gonadal failure. Cranial irradiation, especially in very young children, can lead to neuro-cognitive effects such as memory loss and poor attention. Chemotherapy can lead to male sterility, nephrotoxicity, deafness, pulmonary fibrosis and cardiomyopathy. There is a significant risk (about 12%) of second cancers due to the carcinogenic effect of chemo- and radiotherapy and an increased genetic tendency. Chronic ill health and poor school attendance may have long-term effects on educational achievement, although this may be minimized by good liaison with school and specialist staff.

KEY POINTS

- Acute lymhoblastic leukaemia is the commonest childhood malignancy, but with effective treatment the 5-year survival is above 75%.
- Immunosuppression and neutropenia increase the risk of infection. Suspected infection must be treated aggressively.
- Survivors of childhood cancer may suffer long-term effects including poor growth and endocrine dysfunction.

57 The child with a disability

Prevalence of disability

10-20 per 1000 children have a significant disability

- **Physical and multiple disabilities**

Cerebral palsy	2.5 per 1000
Spina bifida	0.3 per 1000
Muscular dystrophy	0.2 per 1000

- **Severe learning difficulties**

Chromosomal abnormalities	4.0 per 1000
CNS abnormality	0.2 per 1000
Idiopathic	0.5 per 1000

- **Special senses**

Severe visual handicap	1 per 2500
Severe hearing loss	1 per 1000

How disability presents

- Antenatally or at birth if anomalies are present
- In the first year for motor handicaps and severe learning disabilities
- In the second or third year for moderate learning disabilities, language disorder and autism
- After cranial insults

Assessment of disability

This involves:

- A detailed assessment of the child's abilities
- Recognition of any underlying medical problem
- An assessment of the likely long-term difficulties

A Child Development Team is involved for complex difficulties

Professional	Role
Developmental paediatrician	Diagnosis of medical problems Advice on medical issues
Physiotherapist	Assessment and management of gross motor difficulties, abnormal tone and prevention of deformities in cerebral palsy Provision of special equipment
Occupational therapist	Assessment and management of fine motor difficulties Advice on toys, play and appliances to aid daily living
Speech and language therapist	Advice on feeding Assessment and management of speech, language and all aspects of communication
Psychologist	Support and counselling of family and team
Special needs teacher	Advice on special educational needs
Social worker	Support for the family Advice on social service benefits, respite care, etc.
Liaison health visitor	Support for the family Liaison with local health visitor

Management of disability

- **Giving the diagnosis:** this must be carried out in a skilled way by a senior professional
- **Medical management:** therapeutic input should be provided initially at home or the child development centre, and then in nursery and school
- **Genetic counselling:** required by many families even if no obvious genetic cause is identified
- **Education** (see p. 36): A Statement of Special Educational Needs describes the educational provision that must be made for a child with disabilities. Where possible the child should be integrated into mainstream school
- **Provision of services:** Social Services are responsible for preschool childcare, respite care, home help, advice about benefits and assessment for services on leaving school. Voluntary agencies may provide support and information

Living with a disability

- Parents' initial reaction to news of their child's disability is similar to bereavement; they may feel shock, fear and loss, anger and guilt. Each stage of childhood then requires further adaptation, and independence is an ongoing issue.
- Schools need to be prepared for any anticipated difficulties and to accommodate physical disabilities. Staff must work with therapists to implement their recommendations. Young adult disability teams can advise about options beyond high school

Key points

- The child requires a detailed assessment of their difficulties and abilities
  Parents need an explanation of the nature and causes of the child's disability
- A coordinated programme must be developed to cover the child's and family's needs
- Support is important to help the family cope practically and emotionally
  Educational needs and schooling must be met

58 The child with visual and hearing impairment

Visual impairment

A child is defined as blind if education can only be provided by methods such as Braille that do not involve sight. A child is partially sighted if educational methods such as large print books can be used.

Prevalence

1 in 2500 children are registered blind or partially sighted and 50% have additional handicaps.

Aetiology

The commonest causes are optic atrophy, congenital cataracts and choroidoretinal degeneration.

Clinical features

The eyes may look abnormal or have unusual movements. If the deficit is congenital, psychomotor development is altered. Early smiling is inconsistent and there is no turning towards sound. Reaching for objects and the pincer grip is delayed. Early language may be normal, but complex language may be delayed. 'Blindisms' (eye poking, eye rubbing and rocking) may occur. Hearing deficit or severe learning difficulties are commonly associated.

How visual impairment presents

In neonates the diagnosis is suspected if cataracts, nystagmus or purposeless eye movements are present. Otherwise it may be identified by parents or child health surveillance. Ophthalmological examination (and often visual evoked response (VER) testing is required.

Management

Early intervention focuses on developmental progress, reducing blindisms and increasing parental confidence. A peripatetic teacher from the Royal National Institute for the Blind (RNIB) is provided for preschool children, and advises on school, mobility training and supportive services.

Growing up with visual impairment

Parents need advice on caring for their child, non-visual stimulation and adaptations for the home. Mainstream preschool is usually appropriate with support, but beyond this, placement at mainstream school, a partially sighted unit or school for the blind in part depends on learning abilities. Mobility training is important.

Hearing impairment

Prevalence

Four per cent of children have hearing deficits. Most are mild but 2 per 1000 need a hearing aid and 1 per 1000 needs special education.

Aetiology

Most mild to moderate hearing loss is conductive, resulting from secondary otis media. Sensorineural deafness may be genetic, result from pre- or perinatal problems or follow a cerebral insult.

Factors for risk of deafness

- History of meningitis.
- Children with cleft palate.
- History of recurrent otitis media.
- Children with cerebral palsy.
- Family history of deafness.

How hearing impairment presents

Universal neonatal screening with otoautistic emissions (OAE) is now being introduced to identify congenital sensorineural loss. Audiological testing is required for any child with significantly delayed or unclear speech, children at risk (see above) or where there is parental suspicion of deafness. Testing includes brainstem evoked responses (BSER) if the child cannot cooperate.

Clinical features

Lack of response to sound.
Delayed speech.
Behavioural problems.
Associated problems: learning disabilities, neurological disorders, visual deficits.

Management

Grommets are inserted in children with conductive hearing loss. Hearing aids are fitted for sensorineural deafness, and early speech therapy is needed to develop communication. Genetic counselling may be needed.

Growing up with hearing impairment

Parents need to learn to communicate with the child, which may include sign language. Moderately deaf children can attend a normal school. The severely deaf child requires specialist education at a school for the deaf or a hearing unit attached to a normal school.

59 The child with cerebral palsy

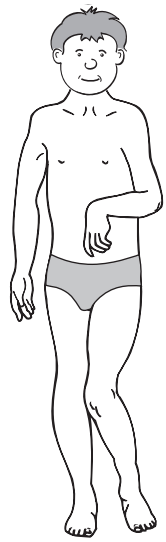
Cerebral palsy is a disorder of movement caused by a permanent, non-progressive lesion in the developing brain. Spastic cerebral palsy is the commonest form

Athetoid cerebral palsy
- Due to basal ganglia damage
- Walking movements
- Intelligence often normal

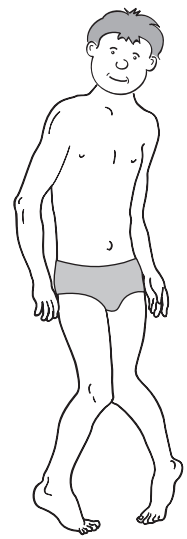
Hemiplegia
- One side of the body
- Arm often more involved than the leg
- Delayed walking
- Tiptoe gait, with arm in a dystonic posture when running

Diplegia
- Both legs involved with arms less affected or unaffected
- Excessive hip adduction (hard to put on a nappy)
- Scissoring of legs
- Characteristic gait: feet in equinovarus and walking on tiptoe

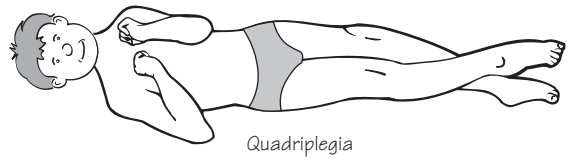
Quadriplegia
- Most severe form
- All extremities involved
- High association with severe learning disabilities and fits
- Swallowing difficulties and gastro-oesophageal reflux common
- Flexion contractures of the knees and elbows often present by late childhood



Hemiplegia



Diplegia



Quadriplegia

Prevalence

- 2–3 per 1000 children

Aetiology

- **Prenatal** — Cerebral malformations; Congenital infection; Metabolic defects
- **Perinatal** — Complications of prematurity; Intrapartum trauma; Hypoxic–ischaemic insult
- **Postnatal** (incurred before 2 yrs of age) — Non-accidental injury; Head trauma; Meningitis/encephalitis; Cardiopulmonary arrest

Diagnosis

Diagnosis is clinical, based on the findings of abnormalities of tone, delays in motor development, abnormal movement patterns and persistent primitive reflexes. Diagnosis may be suspected in neonates but can only be made months later

Associated problems

Children with cerebral palsy may commonly have additional problems (especially if they are quadriplegic or severe hemiplegic):
- Learning difficulties
- Epilepsy
- Visual impairment
- Squint
- Hearing loss
- Speech disorders
- Behaviour disorders
- Undernutrition and poor growth
- Respiratory problems

Prognosis

Depends on the degree and type of cerebral palsy, level of learning diasability and presence of other associated problems. The degree of independent living achieved relates to:
- Type and extent of cerebral palsy
- Degree of learning disability
- Presence of associated problems, e.g. visual impairment, epilepsy

Cerebral palsy is a disorder of movement and posture caused by a permanent and non-progressive cerebral lesion acquired early in brain development. It is often complicated by other neurological and learning difficulties. Although the brain lesion itself in cerebral palsy is non-progressive, the clinical picture changes as the child grows and develops. The underlying brain lesion may result from different insults occurring at various times in the developing brain.

In the neonatal period the diagnosis may be suspected if a baby has difficulty sucking, irritability, convulsions, or an abnormal neurological examination. The diagnosis is usually made later in the first year when the following features emerge.

- **Abnormalities of tone**. Initially the tone may be reduced, but eventually spasticity develops.
- **Delays in motor development:** e.g. marked head lag, delays in sitting and rolling over.
- **Abnormal patterns of development**. Movements are not only delayed, but also abnormal in quality.
- **Persistence of primitive reflexes**: such as the Moro, grasp and asymmetric tonic neck reflex.

The diagnosis is made on clinical grounds, with repeated examinations often required to establish the diagnosis. Once made, a multidisciplinary assessment is needed to define the extent of the difficulties. CT or MRI scan may be useful in demonstrating cerebral malformations, delineating their extent and ruling out very rare progressive or treatable causes such as tumours.

Management of cerebral palsy

Most children with cerebral palsy have multiple difficulties and require a multidisciplinary input. This is best provided by a Child Development Team, who should structure a coordinated programme of treatment to meet all the child's needs, and ensure good liaison between professionals and parents.

Therapy

Physiotherapy

Physiotherapists advise on handling and mobilization, and their role is crucial. The family must be taught how to handle the child in daily activities such as feeding, carrying, dressing and bathing in ways that limit the effects of abnormal muscle tone. They are also taught a series of exercises to prevent the development of deforming contractures. The physiotherapist may also provide a variety of aids, such as firm boots, lightweight splints and walking frames for the child who is beginning to walk.

Occupational therapy

The role of the occupational therapist overlaps with that of the physiotherapist. The occupational therapist is trained to advise on equipment such as wheelchairs and seating, and on play materials and activities that best encourage the child's hand function.

Speech therapy

The speech and language therapist is involved on two accounts—feeding and language. In the early months advice may be required for feeding and swallowing difficulties. Later, a thorough assessment of the child's developing speech and language is required and help given on all aspects of communication, including non-verbal systems when necessary.

Paediatric management

The paediatrician's key role is supportive, along with liaison with other professionals, including at school. In the long term the child needs to be monitored for developmental progress, medical problems, development of contractures or dislocation, behavioural difficulties and nutritional status. Drugs, other than anticonvulsants for epilepsy, have a limited role in cerebral palsy. If spasticity is severe and causing pain, medication to reduce muscle spasm is sometimes prescribed.

Orthopaedic surgery

Even with adequate physiotherapy, orthopaedic deformities may develop as a result of long-standing muscle weakness or spasticity. Dislocation of the hips may occur as a result of spasticity in the thigh adductors and fixed equinus deformity of the ankle as a result of calf muscle spasticity. These may require orthopaedic surgery.

Nutrition

Undernutrition commonly occurs in children with cerebral palsy, and can reduce the child's chances of achieving his or her physical and intellectual potential. Food must be given in a form appropriate to the child's ability to chew and swallow. Energy-rich supplements and medical treatment for reflux may be required. If the child is unable to eat adequate amounts, a gastrostomy may need to be placed.

Growing up with cerebral palsy

The family has to cope with all the difficulties facing any family with a disabled child. However, cerebral palsy, if severe, places particularly heavy demands in terms of time and input. Everyday tasks such as dressing and bathing take time, and feeding, in particular, may take hours each day. The child also needs regular physiotherapy at home, and needs to attend appointments, both for medical follow-up and therapy. In view of this the family needs support, often beyond what family and friends can supply. Voluntary and social service agencies can provide babysitting, respite care and benefits.

Children with milder forms of cerebral palsy can cope at mainstream school, provided minor learning difficulties and physical access are addressed. Children with more severe cerebral palsy need special schooling in a school for the physically or severely learning disabled, depending on the degree of their difficulties.

KEY POINTS

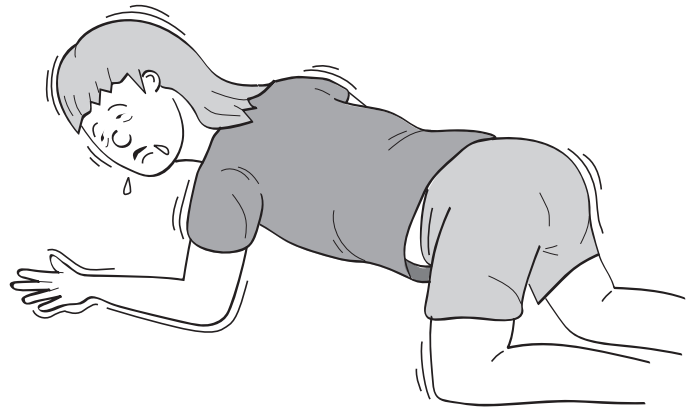
- Physiotherapy is needed to minimize the effects of spasticity and prevent contractures.
- Associated problems must be identified and managed.
- Any special educational needs must be met.
- The family needs adequate financial, practical and emotional support.
- The child's integration into society should be maximized.

60 Epilepsy

Generalized seizures

Generalized tonic–clonic seizures (grand mal)

- Tonic phase: sudden loss of consciousness
  limbs extend, back arches
  teeth clench, breathing stops
  tongue may be bitten
- Clonic phase: intermittent jerking movements
  irregular breathing
  may urinate and salivate
- Postictal phase: child sleepy and disorientated



Simple absence seizures (petit mal)

- Fleeting (5–20 seconds) impairment of consciousness (daydreaming)
- No falling or involuntary movements
- EEG: characteristic bursts of 3/second spike and wave activity

Myoclonic seizures

- Shock-like jerks, often causing sudden falls
- Usually occur in children with a structural neurological/cerebral degenerative condition

Infantile spasms

- Onset usually at 3–8 months of age
- Flexion spasms ('jacknife' or 'salaam')
- Last a few seconds, in clusters lasting up to half an hour
- Regression of developmental skills
- May have a history of perinatal asphyxia or meningitis
- EEG—characteristic hypsarrhythmic pattern

Partial seizures

Simple partial seizures

- Twitching or jerking of face, arm or leg
- Consciousness usually retained
- May start focally and spread (Jacksonian pattern)
- Involved part of the body may be temporarily weak after attack
- May progress to full-blown tonic–clonic attacks

Complex partial seizures (temporal lobe seizures)

- Altered or impaired consciousness associated with strange sensations, hallucinations or semi-purposeful movements
- May show chewing, sucking or swallowing movements
- Postictal phase with amnesia
- EEG may show discharges arising from the temporal lobe

Prevalence

- Approximately 8 per 1000 schoolchildren
- Learning difficulties are a common association

Pathophysiology

- Paroxysmal involuntary disturbances of brain function result in recurrent fits

How the diagnosis is made

- The diagnosis is largely clinical, based on the description of the attacks. EEG has a limited value in the diagnostic process

Paediatric follow-up

Monitor:

- frequency of fits
- side effects of drugs
- psychosocial and educational problems
- anticonvulsant levels if uncontrolled

Prognosis

- Generally good, with resolution of fits in 60% of children with idiopathic epilepsy. Poor prognosis for those with infantile spasms

Seizures, *convulsions* or *fits* are non-specific terms describing an impairment of consciousness, abnormal motor activity, sensory disturbance or autonomic dysfunction. *Epilepsy* is defined as a condition of recurrent fits resulting from paroxysmal involuntary disturbances of brain function and unrelated to fever or acute cerebral insult. Seizures may be *generalized* from the onset, or *partial*, beginning in a localized or focal area of the brain. Partial seizures are defined as *complex* if there is an associated impairment of consciousness. Epilepsy is usually idiopathic, but may result from severe cerebral insult. In some children, particularly those with neurological signs or learning difficulties, there is an underlying anatomical lesion.

The diagnosis of epilepsy is clinical, the key being a good detailed history. Physical examination is usually normal, but the finding of neurological signs suggests possible underlying pathology. Investigations are not usually helpful as 50% of children with epilepsy have normal EEGs on first testing, and 3% of normal children have abnormal EEGs. EEG is useful in diagnosing simple absences and infantile spasms as these have characteristic findings. Twenty-four hour and video EEG

Seizure type and drug therapy.

Type of epilepsy	Drug of first choice	Other drugs
Generalized		
Tonic–clonic (grand mal)	Valproate	Carbamazepine
Simple absences (petit mal)	Valproate	Ethosuxamide
Myoclonic	Clonazepam	Valproate
Infantile spasms	Vigabatrin	ACTH
Partial	Carbamazepine	Phenytoin

recordings are sometimes helpful. CT or MRI is indicated in children with focal neurological deficits, focal EEG changes, increased intracranial pressure, progressive CNS disease or intractable seizures as there is a high chance of a brain lesion or pathology.

Medical management of epilepsy

The goal is to achieve the greatest control of fits while producing the least degree of side-effects. This is best achieved through a monotherapy approach.

- Treatment is started with the most effective drug for the type of fit.
- The dose is gradually increased to maximum recommended levels.
- A second drug is added if the first is ineffective, and the dose increased.
- The first drug, where possible, is gradually discontinued.
- Drugs should be given at intervals no longer than one half-life. Drugs with sedative effects should be given at bedtime and if there is a pattern, the peak level should be timed to coincide with the seizures.

If medical treatment fails, surgery may rarely be tried in children with intractable fits and clinical and electrographic evidence of a discrete epileptic focus. For most children with epilepsy restriction of physical activity is unnecessary, other than attendance by a responsible adult while bathing and swimming. Avoiding cycling in traffic and climbing high gymnastic equipment is prudent.

Management of a tonic–clonic seizure (see p. 46)

In a tonic–clonic seizure, the child should be placed in the recovery position. Parents should be instructed to end the fit by giving diazepam rectally if it lasts more than 10 min. Intravenous drugs should only be given in hospital where facilities are available in the event of respiratory arrest. Children do not need to be hospitalized each time a fit occurs. Emergency treatment is not required for other forms of epileptic fits.

Monitoring a child with epilepsy

The family should be encouraged to keep a diary recording any fits along with medications received, side-effects and behavioural changes. This allows accurate review of the child's condition and the effect of drugs. Physical examination is only required if there is a deterioration in control. Monitoring of anticonvulsant blood levels is only required if fits remain uncontrolled or drug toxicity is suspected. Levels below the therapeutic range can result from inadequate dosage, poor absorption, rapid drug metabolism, drug interactions and deliberate or accidental non-compliance.

Living with epilepsy

Epilepsy is a difficult condition for children to live with as it periodically and unpredictably places them in embarrassing situations. They may suffer from stigmatization and social difficulties, and their integration into school may become affected. Too often physical activities are limited for fear that a fit will place them in danger.

Most parents are initially frightened by the diagnosis of epilepsy and require support and accurate information about the condition. They need to know how to safely manage an acute fit including using rectal diazepam, about side-effects of drugs, the dangers of sudden withdrawal of medication, and social and academic repercussions. There are often concerns about genetic implications, and the teratogenic effects of anticonvulsants must be discussed with the teenage girl.

The family should be encouraged to treat the child as normally as possible and not to thwart the child's independence. This becomes a particular issue in adolescence when compliance too can be a problem. Career guidance is important as some occupations are closed to individuals with epilepsy. Application for a driving licence can only be made after 3 fit-free years whether the person is on or off medication.

Staff at school must be taught the correct management of tonic–clonic fits, although most will not take the responsibility for administering rectal diazepam. Teachers need to be aware of other types of fit such as absence spells, as well as side-effects of drugs, and report these to the parents or school nurse. When epilepsy is associated with learning difficulties, appropriate help needs to be provided.

KEY POINTS

- Ensure the diagnosis is correct.
- Only treat if fits are recurrent.
- Use monotherapy when possible.
- Check plasma levels if control is inadequate and, if low, consider non-compliance.
- For tonic–clonic epilepsy, rectal diazepam should be prescribed for home use.
- Ensure any learning difficulties are addressed.
- Help the child live a normal life with full participation at school and home.

61 Learning disability

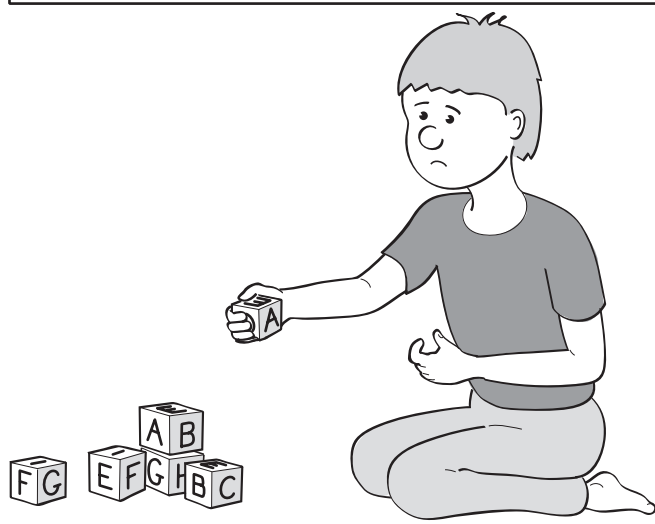
Prevalence
- 4 per 1000 children

Aetiology/pathophysiology

• Chromosome disorders	30%
• Identifiable disorders or syndromes	20%
• Associated with cerebral palsy, microcephaly, infantile spasms, postnatal cerebral insults	20%
• Metabolic or degenerative disease	<1%
• Idiopathic	25%

Clinical features
- Reduced intellectual functioning
- Delay in reaching developmental milestones, particularly language and social skills, in early childhood
- Often associated with:
  Epilepsy
  Vision and hearing deficits
  Communication problems
  Attention deficit/hyperactivity
  Feeding problems and failure to thrive

How learning disability presents
- Dysmorphic features may be evident at birth
- SLD presents as developmental delay before 12 months
- MLD presents with delayed language in toddlers
- The diagnostic process is discussed in Chapter 46 (global developmental delay)



Management (needs to be multidisciplinary)

- Diagnosis of underlying cause
- Early intervention and educational programmes to stimulate cognitive, language and motor development
- School: statementing is required with placement in mainstream or special school
- Behavioural difficulties must be addressed
- Family support and benefits should be provided
- General paediatric care must not be neglected

Paediatric follow-up

- Developmental progress and physical growth require review
- Screening for specific associated problems in some conditions
- Behaviour is often an issue
- Liaison with other professionals is important, particularly regarding education
- The family needs support

Prognosis

- Depends on the underlying cause
- Degree of independent living achieved relates to the level of learning disability and the underlying aetiology

Learning disability (or difficulty) has replaced the terms 'mental retardation' and 'mental handicap'. The disability is classified as mild, moderate or severe according to the intellectual limitation and degree of independence anticipated or achieved. Individuals with *severe* learning difficulties can learn minimal self-care and simple conversation skills, and need much supervision throughout their lives. Those with *profound* learning disability require total supervision, few become toilet-trained and language development is generally minimal.

Children with severe learning disabilities are spread throughout the social classes, and usually have an organic basis for their problem. This contrasts with children with milder learning disabilities, where mostly no organic cause is found and there is a predominance of children from lower socioeconomic classes.

Paediatric management of children with learning disability

The role of the paediatrician is to support and help the family in coming to terms with their child's limitations, to attempt to diagnose the underlying cause, and to manage medical problems. A diagnosis is often not possible, but is of great importance to the family and allows for more accurate genetic counselling. Other aspects of management involve advising on appropriate educational and therapeutic input. Liaison with other professionals is an important aspect of the work.

Growing up with learning disability

The diagnosis of severe learning disability is devastating and families require particularly sensitive support at diagnosis and beyond. Each stage of childhood brings its own issues. Adolescence is usually a par-

ticularly difficult time when issues related to sexuality, vocational training and community living must be addressed.

It is important to begin input early to stimulate cognitive, language and motor development. Therapists from the Child Development Team should provide advice on play activities and suitable toys, give guidance in the development of skills such as feeding, washing and dressing, and instruct parents on the principles of language development, introducing alternative communication systems where appropriate. Attendance at special nurseries, such as Mencap, can be stimulating for the child while providing contact with other families.

Many children with learning disabilities can cope with and benefit from mainstream nursery and primary school, with appropriate help provided. Others, particularly if they have additional disabilities, may be better placed in a special school. A statement of special educational needs (see p. 36) is required. Education must be realistic, and should include teaching skills such as personal care, hygiene and safety, development of acceptable social behaviour, and maximizing independence. On leaving school, facilities should be available for the young adult, including an adult training centre, special hostels, and vocational training schemes.

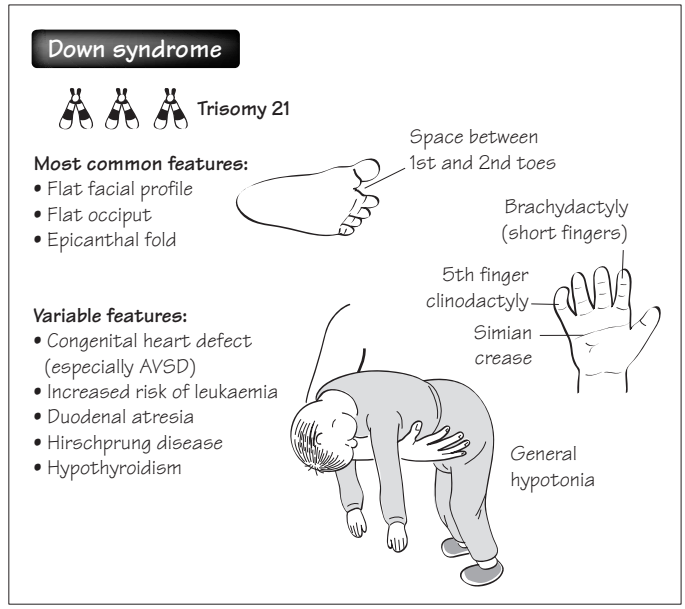
Behaviour problems occur with greater frequency in children with developmental disabilities. This may include attention difficulties or hyperactivity (see p. 37) or stereotypic or self-injurious behaviour. Psychological help is then needed, and occasionally medication too. Genetic counselling is important, whether there is a clearly inherited disorder or not, as the family will want to know the chances of having another affected child. In children with no identified cause, the risk of another sibling being affected is about 1 in 25. However, if multiple congenital anomalies are also present the risk falls to 1 in 40.

Down syndrome (trisomy 21)

Down syndrome is the commonest congenital anomaly causing learning disability. The extra chromosome is usually maternal, and the incidence of Down syndrome increases with maternal age (2% at age 40 years).

Features include upward sloping palpable fissures, epicanthal folds, Brushfield spots (speckled iris), a protruding tongue, flat occiput, single palmar creases, and mild to moderate developmental delay with social skills often exceeding other milestones. One-third are born with GI problems, most commonly duodenal atresia, and one-third have cardiac anomalies (most commonly atrioventricular canal defects). Secretory otitis media, strabismus, hypothyroidism, atlanto-axial instability and leukaemia are common problems. Children with Down syndrome can usually be integrated into mainstream primary school with additional support.

A cardiac evaluation should be performed at birth, routine audiological and thyroid tests are needed throughout childhood and ophthalmological assessment if there is any evidence of a squint. The child's growth needs to be followed on special Down growth charts. The family requires genetic counselling.



Fragile X

Fragile X is an important genetic cause of learning disabilities amongst boys. The chromosomal anomaly consists of a 'fragile site' at the end of one of the long arms of the X chromosome. The diagnosis should be sought in any boy who has unexplained moderate or severe learning disability. Some girls carrying the chromosome have mild learning disabilities.

Autism

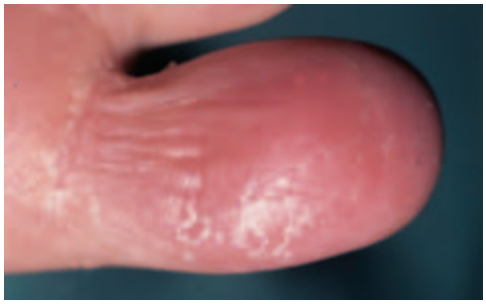
Autism is a condition where there is an inability to relate to others, and language development is very delayed. It often occurs in conjunction with learning disabilities. Characteristically the autistic child fails to develop social relationships, has little non-verbal communication and often has ritualistic behaviour patterns. Special education is often required and the family need good support as it is extremely stressful to have an autistic child in the family.

KEY POINTS

- Where possible the underlying condition should be diagnosed.
- The child's developmental progress should be monitored.
- Appropriate input should be provided in the preschool years and appropriate school placement made.
- The child and parents need a supportive framework.
- Good general paediatric care must be provided.

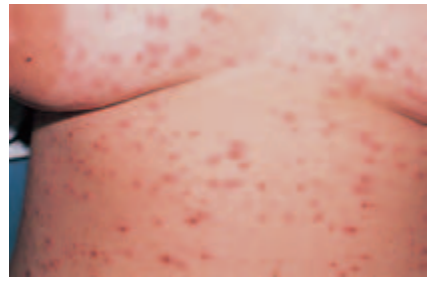
Index

Plate 1: Rashes—types of skin lesions

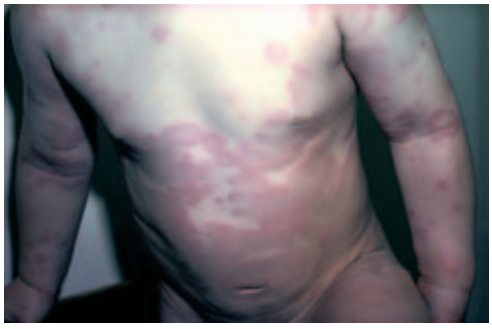


Desquamation

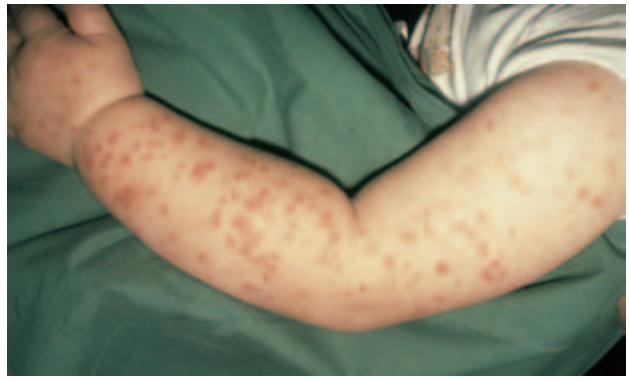
Maculopapular



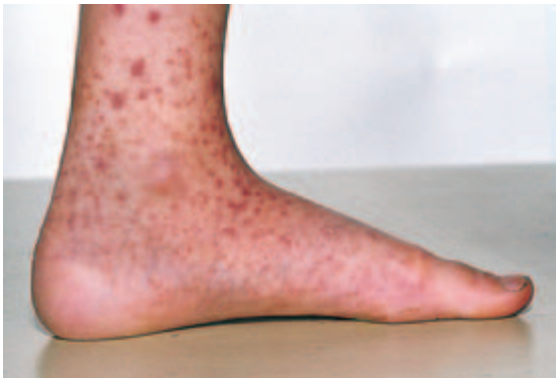
Vesicles



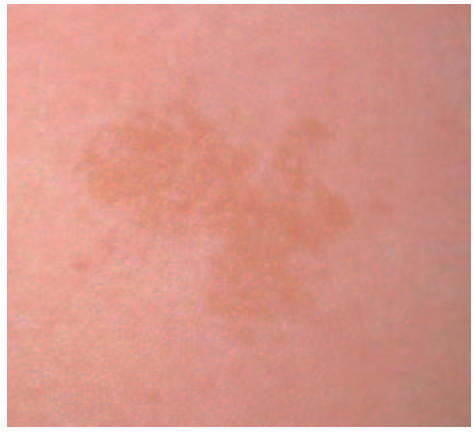
Wheals



Papules

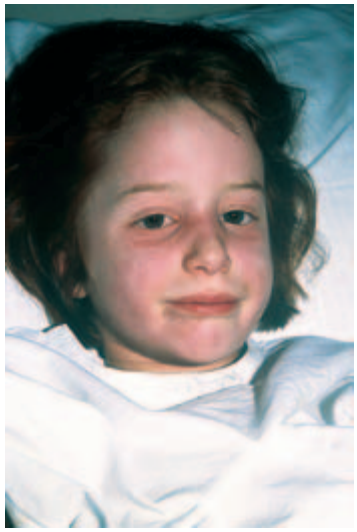


Purpura and petechiae

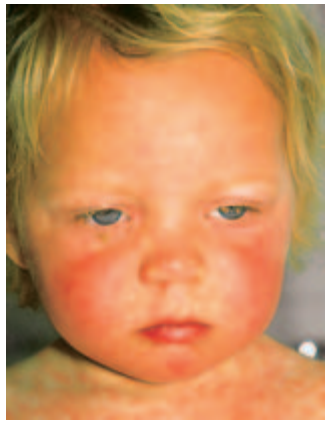


Macules

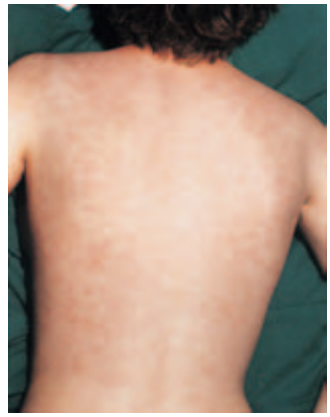
Plate 2: Rashes—acute rashes



Scarlet fever

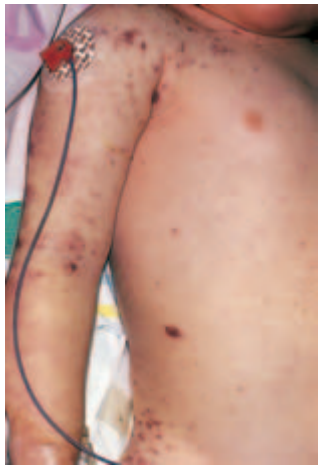


Measles

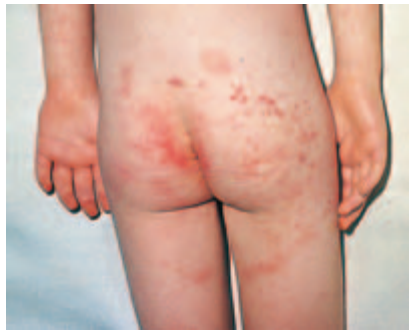


Rubella

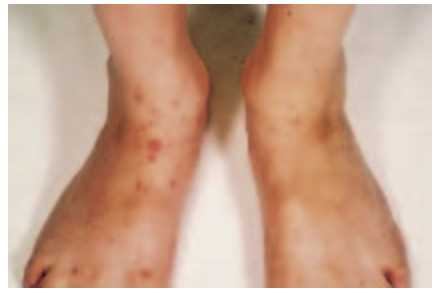
Fifth disease



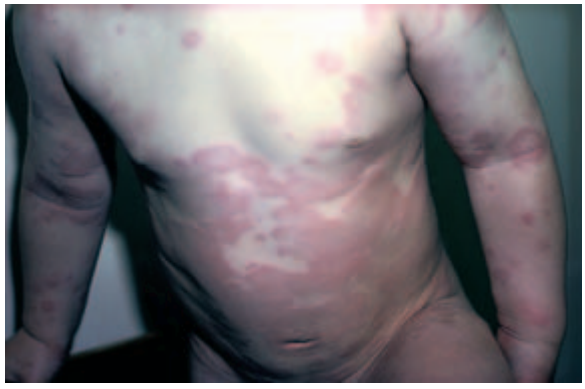
Meningococcal septicaemia



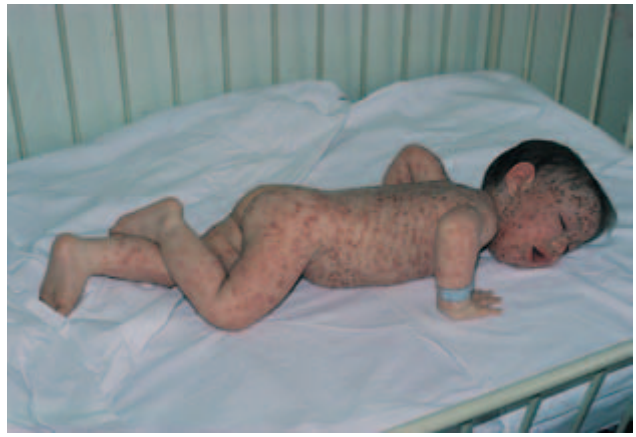
Henoch–Schönlein purpura



Idiopathic thrombocytopenic purpura

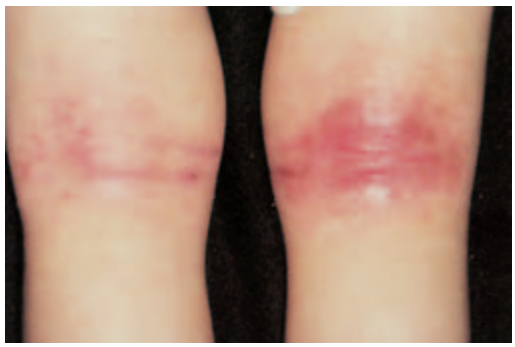


Urticaria

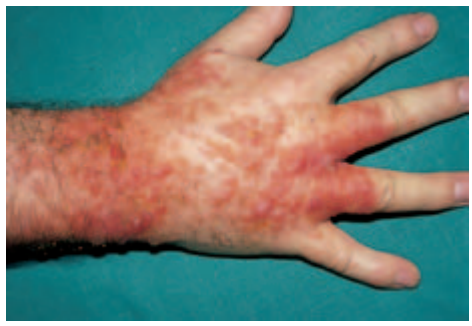


Chicken pox

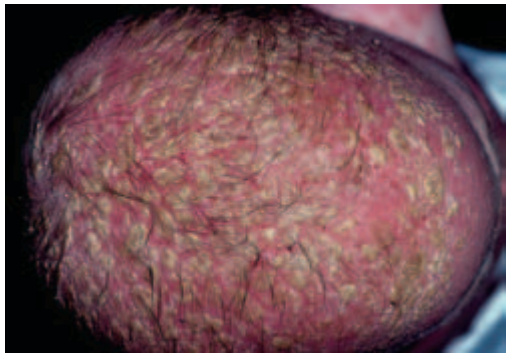
Plate 3: Rashes—chronic skin problems



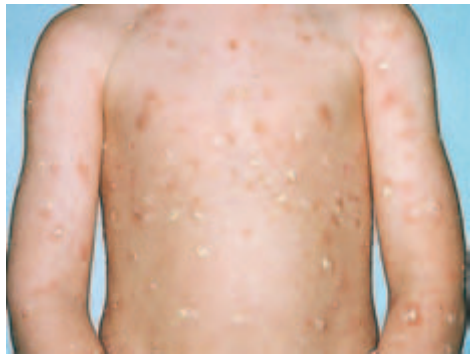
Atopic dermatitis



Contact dermatitis

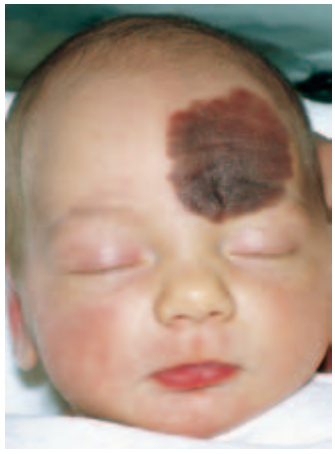


Seborrhoeic dermatitis

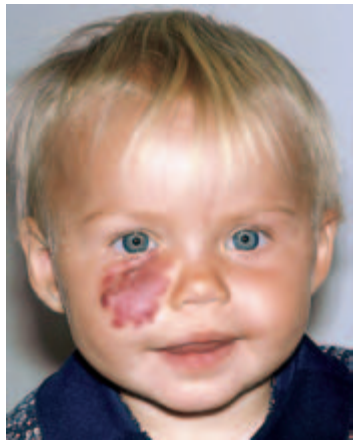


Psoriasis

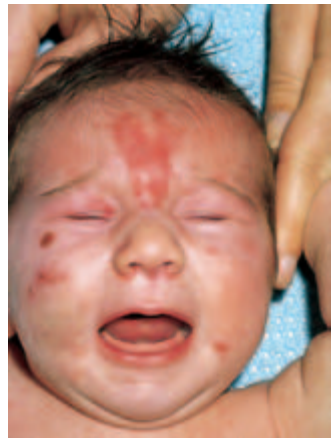
Common birthmarks



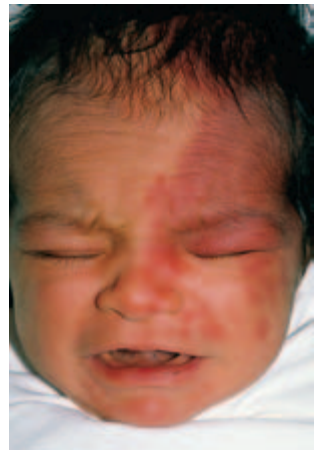
Pigmented naevus



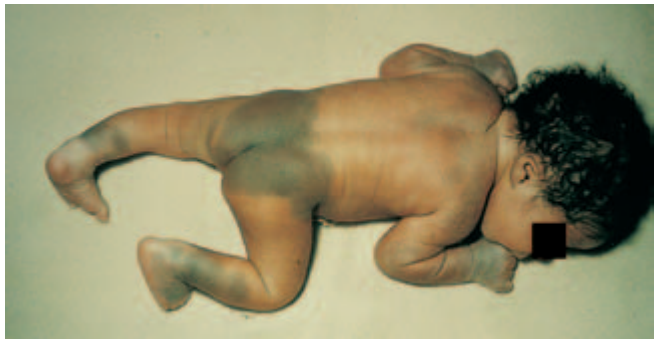
Strawberry naevus



Stork mark

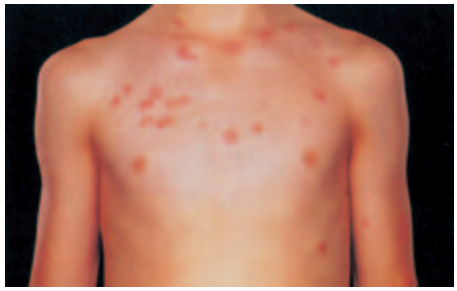


Portwine stain

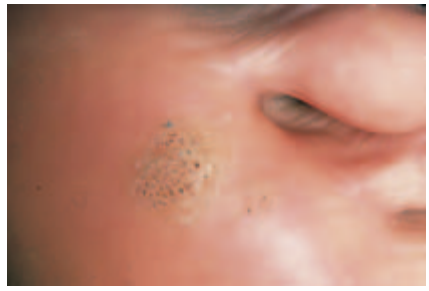


Mongolian spots

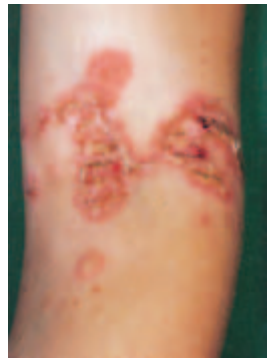
Plate 4: Infectious lesions



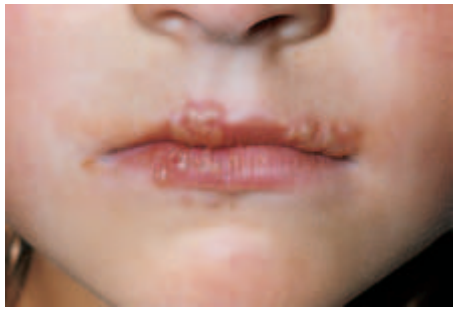
Tinea corporis



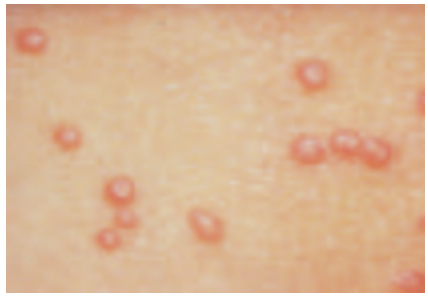
Common warts



Impetigo

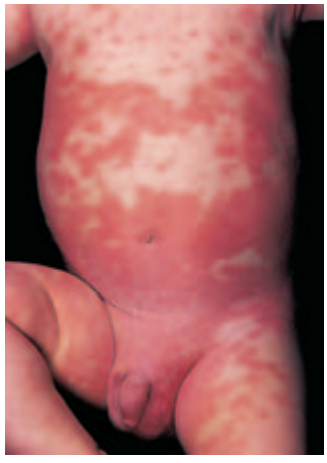


Cold sore

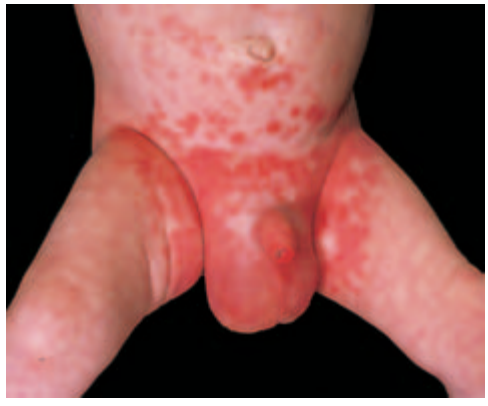


Molluscum contagiosum

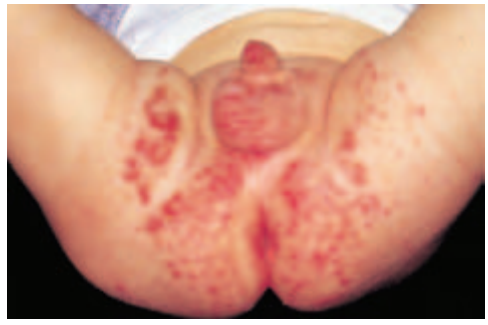
Nappy rashes



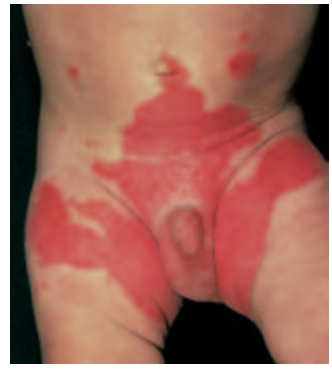
seborrhoeic nappy rash



Candidal nappy rash

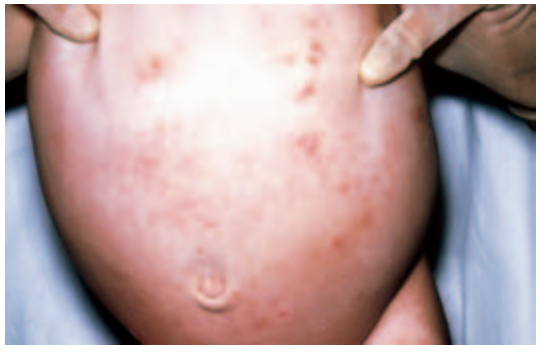


Ammoniacal dermatitis

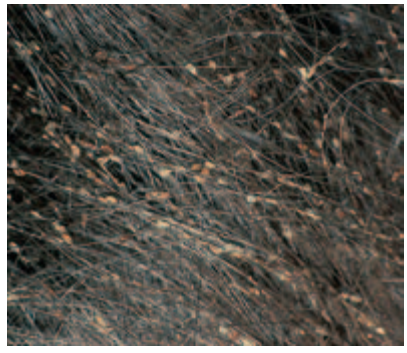


Psoriatic nappy rash

Itchy lesions



Scabies



Head lice