Extra Topics

objectives:

- It includes important topics not covered in the lecture or the tutorials. this is purely from abu wardah, but it is not enough. read the textbook if possible

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Notes

Important

Book

HEMOLYTIC UREMIC SYNDROME

- is a triad of acute renal failure, microangiopathic haemolytic anaemia, and thrombocytopenia. Typical HUS is secondary to gastrointestinal infection with verocytotoxin-producing E. coli O157:H7, acquired through contact with farm animals or eating uncooked beef, or, less often, Shigella. It follows a prodrome of bloody diarrhoea.

- The toxin from these organisms enters the gastrointestinal mucosa and preferentially localizes to the endothelial cells of the kidney where it causes intravascular thrombogenesis. The coagulation cascade is activated and clotting is normal (unlike in disseminated intravascular coagulation).

- Platelets are consumed in this process and microangiopathic haemolytic anaemia (low serum haemoglobin, high serum lactate dehydrogenase) results from damage to red blood cells as they circulate through the microcirculation, which is occluded. Other organs such as the brain, pancreas, and heart may also be involved.

- With early supportive therapy, including dialysis, the typical diarrhoea-associated HUS usually has a good prognosis, although long-term follow-up is necessary as there may be persistent proteinuria and the development of hypertension and progressive chronic kidney disease. By contrast, atypical HUS has no diarrhoeal prodrome, may be genetic, and may relapse frequently

- A monoclonal anti-terminal complement complex antibody, **eculizumab**, has greatly improved the prognosis of this condition, which previously had a high risk of hypertension, progressive chronic kidney disease and mortality. However, it is very expensive and the duration of treatment is unknown.

sudden unexpected death in infancy

- Deaths that occur suddenly and unexpectedly in infants are termed sudden unexpected death in infancy (SUDI). In some, a previously undiagnosed congenital abnormality, e.g. congenital heart disease, will be found at autopsy or another condition, e.g. inborn error of metabolism, is identified. However, in most instances of sudden death in a previously well infant, no cause is identified even after a detailed autopsy, and the death is classified as sudden infant death syndrome (SIDS).

- There were 200 such deaths in the UK in 2018 (0.3 per 1000 live births). The peak age is 2–4 months, but it occurs throughout the first year; 55% are boys, and the risk is increased in those who were low birthweight (fivefold) and with mothers under 20 years old (fivefold greater than all other age groups).

- The vast majority of such deaths, even when occurring more than once in the same family, are due to natural but unexplained causes. Rarely, the death may be due to suffocation, particularly due to unintentional overlying while bed sharing, or from non-accidental injury.

- The incidence of SIDS has fallen dramatically in the UK since the national ‘Back to Sleep’ campaign advising parents to place their infants on their backs to sleep rather than previous widely given advice to lay them prone

- Further epidemiological studies have identified additional risk factors, and advice to parents to minimize risk of SIDS is summarized in Fig. 6.13. The management following the death of an infant from Sudden Infant Death Syndrome is outlined in Case History 6.1.

- SIDS is the most common cause of death in children aged 1 month to 1 year. The peak age for the occurrence of SIDS is 2–4 months.

Figure 6.13 Advice to parents to reduce the risk of SIDS. (Adapted from: Lullaby Trust. www.lullabytrust.org.uk/wp-content/uploads/Safer-sleep-saving-lives-a-guide-for-professionals-web.pdf.)

Case history 6.1

Sudden infant death syndrome

Max, a previously well 9-week-old baby, is found to be still and lifeless by his mother when trying to wake him for a feed in the morning. The previous night he had fed well at midnight although he had coryzal symptoms the previous day. An ambulance is called and Max is rushed to hospital. Basic life support is initiated by the paramedic ambulance crew at the scene.

The team in the children's emergency department is prepared for his arrival. A member of the nursing team is assigned to accompany his mother to a separate room. After 20 minutes of cardiopulmonary resuscitation, he still has no signs of life, and resuscitation attempts are halted.

Max's father had been called to come directly to the hospital. The paediatrician records a comprehensive account of the resuscitation, the history from the paramedics and findings on complete physical examination, including the absence of signs of external injury. An unexpected death occasions police involvement and a member of the police child protection team accompanies the paediatrician who explains to the parents what has happened and takes a detailed history from each of them.

The parents are offered the opportunity to see and hold their baby and to take photographs and gather mementoes. They are reassured that involvement of the police and social services is standard practice. The local coroner is informed and a multiagency information sharing meeting is convened to discuss the death. A postmortem is performed by a paediatric pathologist, which does not identify a definitive cause of death and a label of SIDS is applied. Follow-up with a paediatrician and bereavement support is arranged.

NEUROLOGICAL ABNORMALITIES

Neurological abnormalities

Table 2.9 Neurological abnormalities which may be identified on observation

Inspection	Appearance	Example
Head and face		
Genetic disorder or syndrome	Dysmorphic features	Down syndrome, Williams syndrome
Myopathic face	Expressionless face, often with ptosis and drooping of mouth	Neuromuscular disease, e.g. myotonic dystrophy
Reduced head circumference	Microcephaly (Fig. 2.11a)	Cerebral palsy, congenital infections, e.g. cytomegalovirus, Zika virus
Ptosis	Drooping of eyelid	Unilateral ptosis – third nerve palsy Bilateral – myasthenia gravis
Facial asymmetry (Fig. 2.11c)	Are movements of forehead preserved or weak?	Preserved: upper motor neurone lesion Weak: Bell palsy (Fig 2.11b)
Tongue fasciculation	Writhing of tongue fibres	Spinal muscular atrophy
Skin		
Neurocutaneous skin lesions	Multiple café-au-lait spots Port-wine stain over trigeminal nerve area	Neurofibromatosis Tuberous sclerosis
Limbs		
Muscle wasting	Reduced muscle bulk	Cerebral palsy, meningomyelocele, muscle disorder
Calf muscle hypertrophy	Muscle replaced by fat and connective tissue	Duchenne muscular dystrophy
Contractures	Shortened muscles from increased tone or hypotonia	Immobility, restricted movements *in utero*
Hypertonic posture	Scissoring, fisting, pronation of arms, extended back and legs	Cerebral palsy
Hypotonic posture	Sitting in frog-like posture of legs	Spinal muscular atrophy
Dystonic posture	Abnormal posturing and extension from fluctuating tone	Dystonic cerebral palsy
Muscle fasciculation	Writhing of muscle fibres	Lower motor neurone lesions
Asymmetry	Unilateral weakness	Hemiplegia

Figure 2.11 Examples of neurological abnormalities identified on observation. (**a**) Microcephaly. (**b**) Asymmetry of the face in Bell palsy. (**c**) Play being used to test muscles of facial expression.

Table 2.12 Different forms of abnormal gait, and some of their causes

Gait	Appearance on affected side	Sample condition
Antalgic (to minimize pain)	Stance phase is briefer than swing	Musculoskeletal pain
Broad-based	Staggering gait, wide-spread feet	Cerebellar ataxia
Choreoform	Irregular, involuntary jerking	Choreoathetoid cerebral palsy
Circumduction (Sweeping)	Hypertonic leg circumducts a plantarflexed foot.	Hemiplegia
Scissoring	Hypertonic legs drag and cross the midline	Diplegia
Waddling	Dropping of the hip girdle on contralateral side to muscle weakness	Proximal muscle weakness
Foot drop	Leg raised to avoid scraping plantarflexed foot on ground	Peripheral motor neuropathy
Stamping	High stamping steps without weakness	Sensory neuropathy

preterm baby

- glucocorticoid therapy before preterm delivery accelerates lung maturity and surfactant production.

- This has been tested in over 15 randomized trials and markedly reduces the incidence of respiratory distress syndrome (relative risk 0.66), intraventricular haemorrhage (relative risk 0.54) and neonatal mortality (relative risk 0.69) in preterm infants.
- For optimal effect, a completed course needs to be given at least 24 hours before delivery and within 1–7 days before birth.
- Preterm infants are particularly liable to hypothermia, and every effort must be made to keep them warm during resuscitation and stabilization.
- Infants of less than 32 weeks' gestation should, with the exception of the face, be placed into a plastic bag or wrapped in clear plastic sheeting without drying to allow the plastic to cling to the skin, and act almost like another layer of skin by avoiding evaporative heat loss. A radiant heat source from the resuscitation table and/or thermal mattress can then warm the baby in the bag/wrap. Using warmed humidified respiratory gases may also help.
- Excessive tissue oxygenation may cause tissue damage to the brain, lungs and eyes from oxygen free radicals

respiratory distress syndrome:

- In respiratory distress syndrome (RDS, also called hyaline membrane disease), there is a deficiency of surfactant, which lowers surface tension.
- Surfactant deficiency leads to widespread alveolar collapse and inadequate gas exchange. The more preterm the infant, the higher the incidence of RDS.
- It is very common in infants born before 28 weeks' gestation and tends to be more severe in boys than girls.
- Surfactant deficiency is rare at term but may occur in infants of diabetic mothers and very rarely from genetic mutations in the surfactant genes
- A proteinaceous exudate (forming a hyaline membrane) may be seen in the airways on histology. Glucocorticoids, given antenatally to the mother, stimulate fetal surfactant production and are given if preterm delivery is anticipated
- The evidence of their benefit is substantial; it significantly reduces RDS, the lung damage of bronchopulmonary dysplasia, intraventricular haemorrhage (IVH) and other causes of neonatal mortality.
- At delivery or within 4 hours of birth, babies with RDS develop clinical signs of: • tachypnoea over 60 breaths/minute • increased work of breathing, with chest wall recession (particularly sternal and subcostal indrawing) and nasal flaring • expiratory grunting in order to try to create positive airway pressure during expiration and maintain functional residual capacity • cyanosis if severe.

preterm baby

respiratory distress syndrome:

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  - increased work of breathing, with chest wall recession (particularly sternal and subcostal indrawing) and nasal flaring
  - expiratory grunting in order to try to create positive airway pressure during expiration and maintain functional residual capacity
  - cyanosis if severe.

The characteristic chest X-ray appearance is shown

- Management is initially with supplemental oxygen and non-invasive respiratory support (continuous positive airway pressure (CPAP) or high-flow nasal cannula therapy).
- Surfactant therapy may be required. It is given by instilling surfactant directly into the lungs via a tracheal tube or a fine catheter inserted directly between the vocal cords into the trachea.
- Mechanical ventilation is initiated if there is inadequate response. Non-invasive respiratory support is used in preference to mechanical ventilation

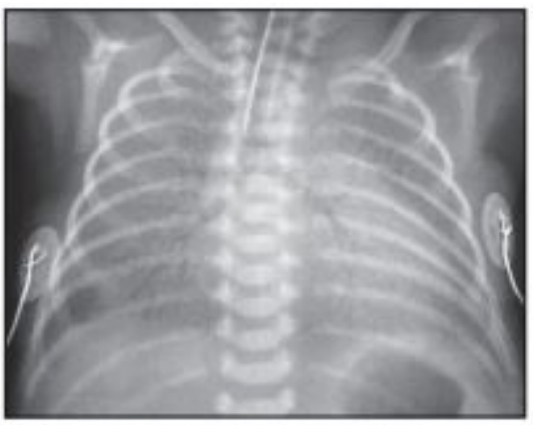


Figure 11.7 Chest X-ray in respiratory distress syndrome showing a diffuse granular or 'ground glass' appearance of the lungs and an air bronchogram, where the larger airways are outlined. The heart border is indistinct. A tracheal tube is present. (From: Lissauer T, Fanaroff AA, Miall L, et al: *Neonatology at a Glance*, ed 3. Oxford, 2015, Wiley Blackwell, with permission.)

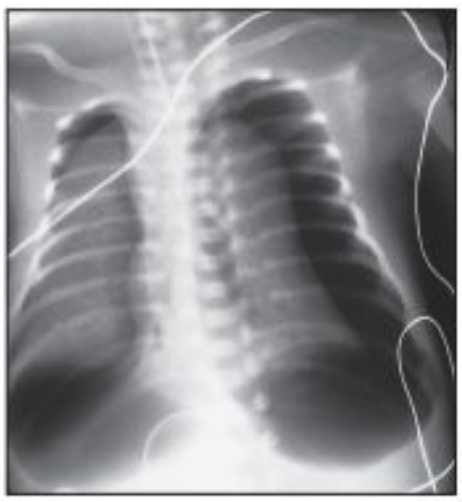


Figure 11.8 Chest X-ray showing bilateral pneumothoraces in a preterm infant with respiratory distress syndrome.

preterm baby

- A preterm infant's fluid requirements will vary with gestational and chronological age. It is adjusted according to the infant's clinical condition, plasma electrolytes, urine output, and weight change.
- first day, about 60–90 ml/kg is usually required, which increases by 20–30ml/kg per day to 150–180ml/kg per day by about day 5.
- Preterm infants have a high nutritional requirement because of their rapid growth. Preterm infants at 28 weeks' gestation double their birth weight in 6 weeks and treble it in 12 weeks, whereas term babies double their weight in only 5 months and treble it in a year. Infants of 35–36 weeks' gestational age are mature enough to suck and swallow milk. Less mature infants usually need to be fed via a nasogastric or orogastric tube.
- Preterm infants are at an increased risk of infection, as IgG is mostly transferred across the placenta in the last trimester and no IgA or IgM is transferred. In addition, infection in or around the cervix is often a reason for preterm labour and may cause infection in the infant shortly after birth.
- Preterm infants are also at particular risk of nosocomial (hospital-derived) infection, often associated with indwelling catheters or mechanical ventilation.

Necrotizing enterocolitis

- is a serious illness and one of the major challenges facing neonatal medicine. The incidence is inversely proportional to gestational age.
- It is typically seen in the first few weeks of life.
- The aetiology of necrotizing enterocolitis is poorly understood, but is thought to be due to ischaemic injury and bacterial invasion of the bowel wall and altered gut microbiome, which is improved with breast milk and possibly prebiotics and probiotics, and adversely affected by formula feeds, unduly rapid increase in enteral feeds and antibiotics.
- Risk factors are intrauterine growth restriction, especially if accompanied by antenatal reversed end diastolic flow on Doppler studies and perinatal asphyxia.
- Early signs of necrotizing enterocolitis include feed intolerance and vomiting, which may be bile stained. The abdomen becomes distended and the stool sometimes contains fresh blood.
- The infant may rapidly become shocked and require mechanical ventilation because of abdominal distension and pain. The characteristic X-ray features are distended loops of bowel and thickening of the bowel wall with intramural gas, and there may be gas in the portal venous tract. The disease may progress to bowel perforation.
- Treatment is to stop oral feeding and give broad spectrum antibiotics to cover both aerobic and anaerobic organisms. Parenteral nutrition is needed and mechanical ventilation and circulatory support are often required. Surgery is performed for bowel perforation, difficulty with mechanical ventilation, or a failure to respond to medical management The disease has significant morbidity and a mortality of about 20%.

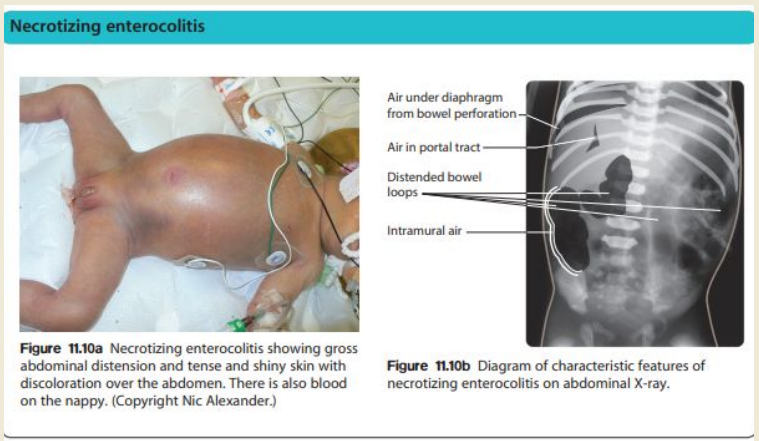


Figure 11.10a Necrotizing enterocolitis showing gross abdominal distension and tense and shiny skin with discoloration over the abdomen. There is also blood on the nappy. (Copyright Nic Alexander.)

Figure 11.10b Diagram of characteristic features of necrotizing enterocolitis on abdominal X-ray.

preterm baby

brain lesions:

- Cranial ultrasound is performed to identify a range of brain lesions to which preterm infants are predisposed.
- Haemorrhages in the brain occur in 25% of very-low birthweight infants and are easily recognized on cranial ultrasound scans. Typically, they occur in the germinal matrix above the caudate nucleus, which contains a fragile network of blood vessels.
- Most intraventricular haemorrhages (IVHs) occur within the first 72 hours of life. They are more common following perinatal asphyxia and in infants with severe RDS. Pneumothorax is a significant risk factor.
- Antenatal glucocorticoids prior to preterm delivery is associated with a reduction in the incidence and severity of RDS, and therefore of IVH.
- Small haemorrhages are confined to the germinal matrix, but larger haemorrhages may extend into the ventricles. The most severe haemorrhage is unilateral hemorrhagic infarction involving the parenchyma of the brain; this usually results in hemiplegia
- A large IVH may impair the drainage and reabsorption of cerebrospinal fluid (CSF), thus allowing CSF to build up under pressure. This dilatation may resolve spontaneously or progress to hydrocephalus, which may cause the cranial sutures to separate, the head circumference to increase rapidly
- Preterm infants are susceptible to white matter injury and abnormal cerebral development following ischaemia and inflammation even in the absence of haemorrhage. It may result in cystic white matter lesions visible on cranial ultrasound. Bilateral multiple cysts, called periventricular leukomalacia (PVL), have an 80%–90% risk of spastic diplegia, often with cognitive impairment

Retinopathy of prematurity:

- affects developing blood vessels at the junction of the vascularized and nonvascularized retina. There is vascular proliferation, which may progress to retinal detachment, fibrosis and blindness.
- It was initially recognized that the risk is increased by uncontrolled use of high concentrations of oxygen. Now, even with careful monitoring of the infant's oxygenation, retinopathy of prematurity is still identified in about 35% of all very-low-birthweight infants, with severe disease requiring treatment in 5%.
- The eyes of susceptible preterm infants (≤1500g birthweight or LESS THAN 32WEEKS are screened by an ophthalmologist.
- Laser therapy reduces visual impairment, and intravitreal anti-VEGF (antivascular endothelial growth factor) therapy is being investigated.
- Severe bilateral visual impairment occurs in about 1% of very-low-birthweight infants, mostly in infants

preterm baby

bronchopulmonary dysplasia:

- ınfants who still have an oxygen requirement at a postmenstrual age of 36 weeks are described as having bronchopulmonary dysplasia (BPD) (also called chronic lung disease).
- The lung damage is now thought to be mainly from delay in lung maturation, but may also be from pressure and volume trauma from artificial ventilation, oxygen toxicity and infection.
- The chest X-ray characteristically shows widespread areas of opacification, sometimes with cystic changes.
- Some infants need prolonged artificial ventilation, but most are weaned onto CPAP or high-flow nasal cannula therapy followed by supplemental oxygen, sometimes over several months

- Corticosteroid therapy may facilitate earlier weaning from the ventilator and often reduces the infant's oxygen requirements in the short term, but concern about increased risk of abnormal neurodevelopment including cerebral palsy limits use to those at highest risk and only short, low-dose courses are given.
- Some babies go home while still receiving additional oxygen. A few infants with severe disease may die of intercurrent infection or pulmonary.

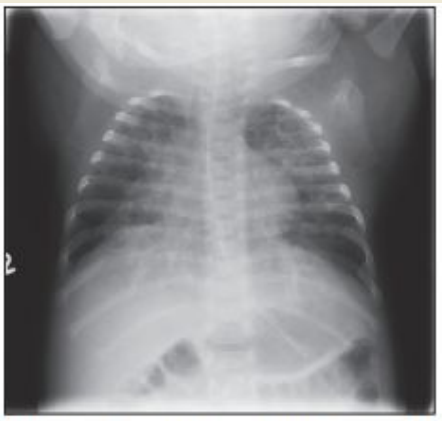


Figure 11.12 Chest X-ray of bronchopulmonary dysplasia (BPD) showing fibrosis and lung collapse, cystic changes and over-distension of the lungs.

Summary of problems of very low birthweight infants (<1.5 kg)

Respiratory

Respiratory distress syndrome (surfactant deficiency) (74%)
- respiratory distress within 4 hours of birth
- antenatal corticosteroids and surfactant therapy reduce morbidity and mortality
- oxygen therapy, but excess may damage the retina
- nasal CPAP (continuous positive airway pressure) (86%) and mechanical ventilation (64%) – often required to expand lungs and prevent lung collapse; high-flow nasal cannula therapy may also be used (75%)

Pneumothorax (4%)
Apnoea and bradycardia and desaturations
Bronchopulmonary dysplasia (BPD) – oxygen requirement at 36 weeks post-menstrual age (25%)

Circulation
Hypotension – may require volume support, inotropes or corticosteroids
Patent ductus arteriosus – needing medical treatment (12%) or surgical ligation (3%)

Nutrition
Nasogastric tube feeding – until 35–36 weeks post-menstrual age
Feeding intolerance - PN (parenteral nutrition) often required

Gastrointestinal
Necrotizing enterocolitis (5%) – serious, management is medical or surgery for bowel necrosis or perforation

Metabolic
Hypoglycaemia – common
Electrolyte disturbances
Osteopenia of prematurity from phosphate deficiency

Hearing
Checked before discharge

Jaundice – common, low treatment threshold

Anaemia
Often need blood transfusions

Eyes
Retinopathy of prematurity – may need laser therapy (5%)

Temperature control
Avoid hypothermia
Nurse in neutral thermal environment
Nurse in incubator or under radiant warmer
Clothe if possible
Humidity reduces evaporative heat loss

Infection
Common and potentially serious (2% early-onset and 11% late-onset infection)
Increased risk of early-onset infection – group B streptococcus
Main problem is nosocomial infection – mainly coagulase-negative staphylococcus, also other infections

Brain injury
Haemorrhage (25%, severe grade III/IV 8%) – germinal layer, intraventricular, parenchymal
Ventricular dilatation – may need ventriculo-peritoneal shunt
Periventricular leukomalacia (3%) – ischaemic white matter injury

Following discharge
Specialist community nursing support helpful, if available
Increased risk of respiratory infection and wheezing – especially from bronchiolitis (caused by respiratory syncytial virus, RSV) and pertussis; may need intensive care
Give routine immunizations according to chronological age
Consider prophylaxis against RSV infection
Increased rehospitalization – respiratory disorders, inguinal hernias
Monitor growth, development (for learning disorders, co-ordination, cerebral palsy), behaviour, attention, vision, hearing – increased risk of impairment, according to corrected age until 2 years old

Diaphragmatic hernia

- Diaphragmatic hernia This occurs in about 1 in 4000 births. Many are now diagnosed on antenatal ultrasound screening. In the newborn period, it usually presents with failure to respond to resuscitation or with severe respiratory distress.
- In most cases, there is left-sided herniation of abdominal contents through the posterolateral foramen of the diaphragm. The apex beat and heart sounds will then be displaced to the right side of the chest, with poor air entry in the left chest.
- Vigorous resuscitation may cause a pneumothorax in the normal lung, thereby aggravating the situation.
- The diagnosis is confirmed by X-ray of the chest and abdomen
- Once the diagnosis is suspected, a large nasogastric tube is passed and suction is applied to prevent distension of the intrathoracic bowel.
- Initial management is focused on stabilizing the infant's ventilation.
- The main problem is pulmonary hypoplasia, as compression by the herniated viscera throughout pregnancy has prevented development of the lung in the fetus. This is often compounded by pulmonary hypertension, Subsequently, the diaphragmatic hernia is repaired surgically. If the lungs are hypoplastic, mortality is high.

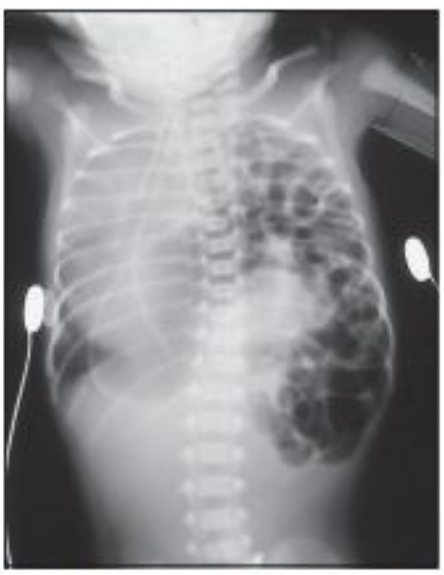


Figure 11.18 Chest X-ray of diaphragmatic hernia showing loops of bowel in the left chest and displacement of the mediastinum.

GIT

Small bowel obstruction:

- This may be recognized antenatally on ultrasound scanning. Otherwise, small bowel obstruction presents with persistent vomiting, which is bile stained unless the obstruction is above the ampulla of Vater.
- Meconium may initially be passed, but subsequently its passage is usually delayed or absent with no transition to normal stool. Abdominal distension becomes increasingly prominent the more distal the bowel obstruction
- High lesions will present soon after birth, but lower obstruction may not present for some days.

GIT

Small bowel obstruction:

- Small bowel obstruction may be caused by:
  - atresia or stenosis of the duodenum – one-third have Down syndrome and it is also associated with other congenital malformations
  - atresia or stenosis of the jejunum or ileum – there may be multiple atretic segments of bowel
  - malrotation with volvulus – a dangerous condition as it may lead to infarction of the entire midgut
  - meconium ileus – thick inspissated meconium, of putty-like consistency, becomes impacted in the lower ileum; almost all affected neonates have cystic fibrosis.
- The diagnosis is made on clinical features and abdominal X-ray showing intestinal obstruction.
- Atresia or stenosis of the bowel and malrotation are treated surgically, after correction of fluid and electrolyte depletion. Meconium ileus may be dislodged using Gastrografin contrast medium but otherwise will require surgery.

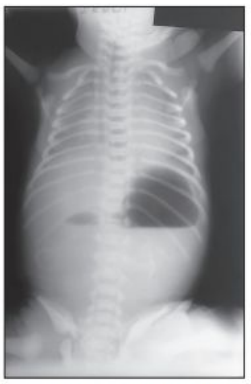


Figure 11.33 Abdominal X-ray in duodenal atresia showing a 'double bubble' from distension of the stomach and duodenal cap. There is absence of air distally.

Intussusception:

- Intussusception describes the invagination of proximal bowel into a distal segment. It most commonly involves ileum passing into the caecum through the ileocaecal valve
- Intussusception is the most common cause of intestinal obstruction in infants after the neonatal period. Although it may occur at any age, the peak age of presentation is 3 months to 2 years of age.
- Clinical features are paroxysmal, colicky pain with pallor, abdominal mass and redcurrant jelly stool. Shock is an important complication and requires urgent treatment.
- Reduction is attempted by rectal air insufflation unless peritonitis is present. Surgery is required if reduction with air insufflation is unsuccessful or for peritonitis.

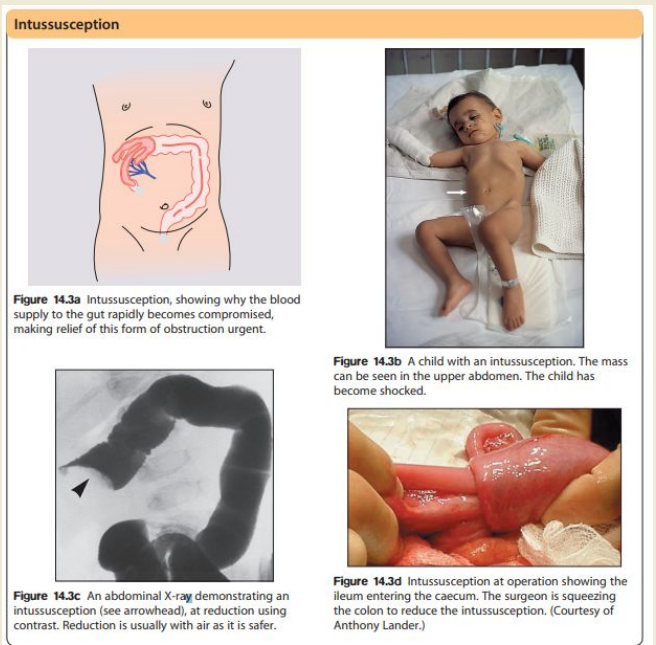


Figure 14.3a Intussusception, showing why the blood supply to the gut rapidly becomes compromised, making relief of this form of obstruction urgent.

Figure 14.3b A child with an intussusception. The mass can be seen in the upper abdomen. The child has become shocked.

Figure 14.3c An abdominal X-ray demonstrating an intussusception (see arrowhead), at reduction using contrast. Reduction is usually with air as it is safer.

Figure 14.3d Intussusception at operation showing the ileum entering the caecum. The surgeon is squeezing the colon to reduce the intussusception. (Courtesy of Anthony Lander.)

GIT

Malrotation and volvulus:

- Malrotation is a congenital abnormality of the midgut, in which the small intestine most commonly lies predominantly on the right-hand side of the abdomen, with the caecum in the right upper quadrant.
- This results from a failure of the intestine to 'rotate' into the correct position during fetal life and secure or 'fix' the mesentery in the correct position. The reason for this developmental failure is unknown.
- Uncommon but important to diagnose. Usually presents in the first 1–3 days of life with intestinal obstruction from Ladd bands obstructing the duodenum or volvulus. May present at any age with volvulus causing obstruction and ischaemic bowel.
- Clinical features are bilious vomiting, abdominal pain and tenderness from peritonitis or ischaemic bowel. An urgent upper gastrointestinal contrast study is indicated if there is bilious vomiting.
- Treatment is urgent surgical correction.

pyloric stenosis:

- there is hypertrophy of the pyloric muscle causing gastric outlet obstruction. It presents at 2–8 weeks of age, irrespective of gestational age. It is more common in boys (4:1), particularly first-born, and there may be a family history, especially on the maternal side.
- Clinical features are: non-bilious vomiting, which increases in frequency and forcefulness over time, ultimately becoming projectile, feeds normally after vomiting until dehydration leads to loss of interest in feeding, weight loss if presentation is delayed. A hypochloremic hypokalemic metabolic alkalosis develops as a result of vomiting stomach contents, Hyponatraemia may also be present
- Diagnosis: Classically, pyloric stenosis has been confirmed by performing a test feed, where the baby is given a milk feed which initially calms the hungry infant, and allows for examination. The diagnosis is made if the pyloric mass, which feels like an olive, is palpable in the right upper quadrant . This has been replaced by ultrasound by visualizing the hypertrophied pylorus
- Management: is surgical (pyloromyotomy), this can only be performed safely after acid-base electrolyte imbalances have been corrected, which may take more than 24hours of intravenous fluid rehydration

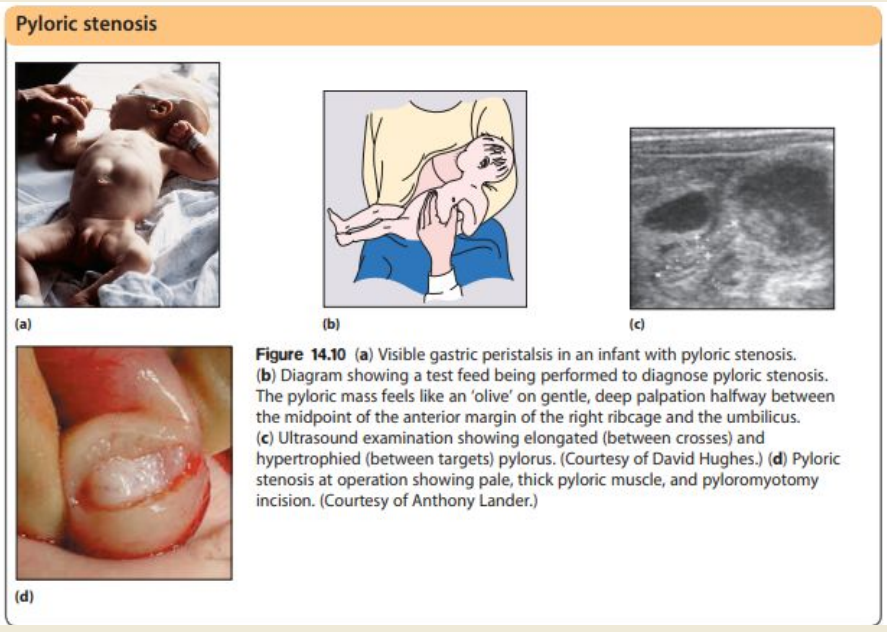


Figure 14.10 (**a**) Visible gastric peristalsis in an infant with pyloric stenosis. (**b**) Diagram showing a test feed being performed to diagnose pyloric stenosis. The pyloric mass feels like an 'olive' on gentle, deep palpation halfway between the midpoint of the anterior margin of the right ribcage and the umbilicus. (**c**) Ultrasound examination showing elongated (between crosses) and hypertrophied (between targets) pylorus. (Courtesy of David Hughes.) (**d**) Pyloric stenosis at operation showing pale, thick pyloric muscle, and pyloromyotomy incision. (Courtesy of Anthony Lander.)

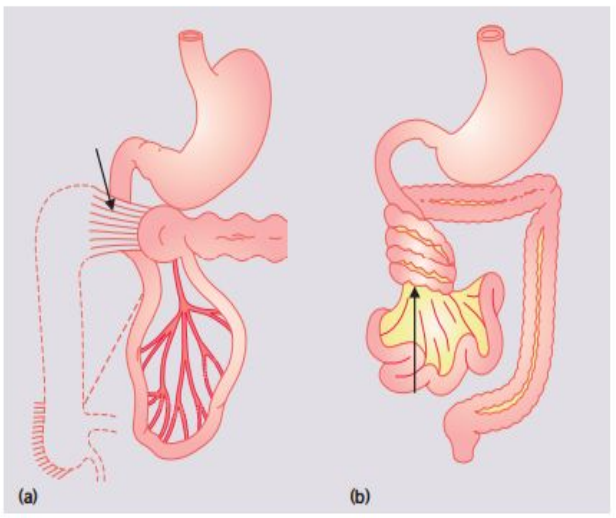


Figure 14.5 (**a**) The most common form of malrotation, with the caecum remaining high and fixed to the posterior abdominal wall. There are Ladd bands (arrow) obstructing the duodenum. Dotted lines show normal anatomy. (**b**) Volvulus from rotation of the bowel (arrow). This will result in ischaemia of the small and proximal large intestine.

GIT

Box 14.3 'Red flag' clinical features in the vomiting child

Bile-stained vomit	Intestinal obstruction (see Ch. 11, Neonatal medicine)
Haematemesis	Oesophagitis, peptic ulceration, oral/nasal bleeding, and oesophageal variceal bleeding
Projectile vomiting, in first few weeks of life	Pyloric stenosis
Vomiting at the end of paroxysmal coughing	Whooping cough (pertussis)
Abdominal tenderness/abdominal pain on movement	Surgical abdomen
Abdominal distension	Intestinal obstruction, including strangulated inguinal hernia, ascites
Hepatosplenomegaly	Chronic liver disease, inborn error of metabolism
Blood in the stool	Intussusception, bacterial gastroenteritis, inflammatory bowel disease
Severe dehydration, shock	Severe gastroenteritis, systemic infection (urinary tract infection, meningitis), diabetic ketoacidosis
Bulging fontanelle or seizures	Raised intracranial pressure, meningitis
Faltering growth	Gastro-oesophageal reflux disease, coeliac disease and other chronic gastrointestinal conditions

Box 14.6 'Red and amber flag' symptoms or signs in the child with constipation

'Red flag' symptom/signs – urgent referral	**Diagnostic concern**
Failure to pass meconium within 24 hours of life, constipation from or soon after birth, family history of Hirschsprung disease	Hirschsprung disease
Abdominal distension with vomiting	Hirschsprung disease or intestinal obstruction
Ribbon stool pattern	Anal stenosis
Abnormal lower limb neurology or deformity	Neurological or spinal cord abnormality
Abnormality of lumbosacral or gluteal regions, e.g. sacral dimple above natal cleft, or naevus, hairy patch, central pit, or discoloured skin over the spine	Spina bifida occulta
Abnormal appearance / position / patency of anus	Abnormal anorectal anatomy
Perianal bruising or multiple fissures	Sexual abuse
Perianal fistulae, abscesses, or fissures	Perianal Crohn disease
Amber signs – specialist referral; start treating constipation	**Diagnostic concern**
Faltering growth / growth failure	Hypothyroidism, coeliac disease, other causes
Constipation triggered by introduction of cow's milk	Cow's milk protein allergy

(Based on: National Institute for Health and Clinical Excellence (NICE) guideline: Constipation in children, 2019.)

Oral rehydration therapy:

- This is a key component of the management of gastroenteritis.
- It contains both sodium and glucose, which increases active sodium and passive water absorption. This works effectively even in the presence of inflammation of the gut, and is therefore effective in diarrhoeal illness. The oral rehydration solution does not 'stop' the diarrhoea, which often continues, but the absorption of water and solutes exceeds secretion and keeps the child hydrated until the infective organism is eradicated. It should be offered in small amounts given frequently, by nasogastric tube if necessary.

'Classical' coeliac disease

A 2-year-old boy had a history of poor growth from 12 months of age. His parents had noticed that he tended to be irritable and grumpy and had three or four foul-smelling stools a day. On examination he had wasting of the buttocks, a distended abdomen (Fig. 14.13a) and faltering growth (Fig. 14.13b). He had a moderately elevated IgA anti-tTG with normal IgA immunoglobulin levels. As his blood tests were not diagnostic of coeliac disease, a duodenal biopsy was performed. This showed subtotal villous atrophy (Fig. 14.13c,d). He was started on a gluten-free diet and, within a few days, his parents commented that his mood had improved and, within a month, he was a 'different child'. He subsequently exhibited good catch-up growth.

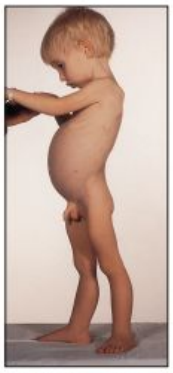


Figure 14.13a Coeliac disease causing wasting of the buttocks and distended abdomen.

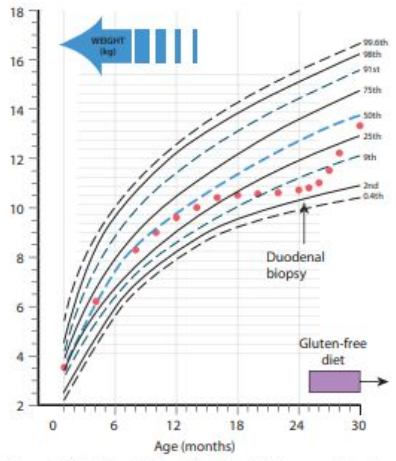


Figure 14.13b Growth chart showing faltering growth and response to a gluten-free diet. (Adapted from: Growth Chart © Royal College of Paediatrics and Child Health.)

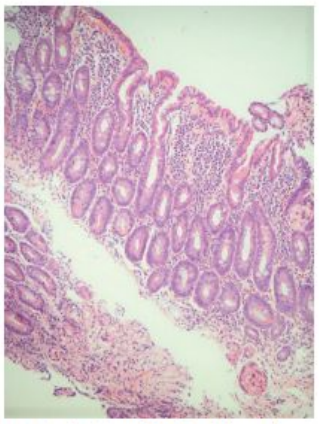


Figure 14.13c Histology of duodenal biopsy showing villous blunting, crypt hyperplasia and intra-epithelial lymphocytosis confirming a diagnosis of coeliac disease. (Courtesy of Professor Marta Cohen.)

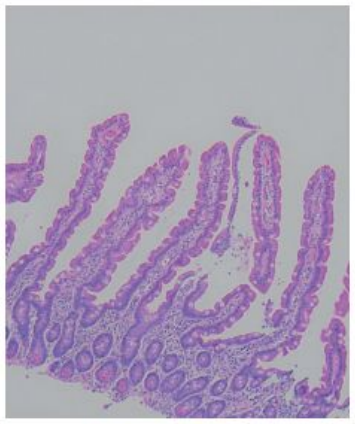


Figure 14.13d Normal duodenal histology shown for comparison. (Courtesy of Professor Marta Cohen.)

Figure 14.14 Causes of nutrient malabsorption. They are uncommon.

Chronic non-specific diarrhoea

- This condition, previously known as toddler diarrhoea, is the most common cause of persistent loose stools in preschool children.
- Characteristically, the stools are of varying consistency, sometimes well formed, sometimes explosive and loose. The presence of undigested vegetables in the stools is common.
- Affected children are well and thriving. In a proportion of children the diarrhoea may result from undiagnosed coeliac disease or excessive ingestion of fruit juice, especially apple juice. Occasionally the cause is temporary cow's milk allergy following gastroenteritis, when a trial of a cow's milk protein free diet may be helpful.
- it almost always improves with age.

INFECTIOUS DISEASES

Scarlet fever:

- This occurs in association with an exotoxin from group A streptococcal pharyngitis.
- It is a diffuse, erythematous maculopapular rash with a sandpaper texture, which appears shortly after the pharyngitis. It usually spreads rapidly to the face, trunk and extremities with increased density around the neck, axillae or groin.
- There is circumoral pallor with the rash sparing the skin around the mouth. The tongue is initially white but desquamates to leave a red strawberry tongue with prominent papillae. There may be desquamation around the fingertips and toes.
- A throat swab may confirm group A streptococcus.
- Antibiotics such as penicillin V or erythromycin may hasten recovery from streptococcal tonsillitis by, on average, only 16 hours. In order to eradicate group A beta-haemolytic streptococci and prevent rheumatic fever, glomerulonephritis and other complications, **10 days of antibiotic treatment is required**. **This is indicated in countries where the risk of rheumatic fever is significant, if the child or young person is returning to a closed institution, e.g. boarding school, or is at increased risk of infection.**

Impetigo:

- This is a localized, highly contagious, staphylococcal or streptococcal skin infection, most commonly occurring in infants and young children.
- It is more common in children with pre-existing skin disease, e.g. atopic eczema. Lesions are usually on the face, neck, and hands and begin as erythematous macules that may become vesicular/pustular or even bullous. Rupture of the vesicles with exudation of fluid leads to the characteristic confluent honey-coloured crusted lesions.
- Infection is readily spread to adjacent areas and other parts of the body by autoinoculation of the infected exudate. Topical antibiotics (e.g. mupirocin) are sometimes effective for mild cases. Narrow-spectrum systemic antibiotics (e.g. flucloxacillin) are generally needed for more severe infections, although more broad-spectrum antibiotics such as co-amoxiclav or cephalexin have better adherence.
- Affected children should not go to nursery or school until the lesions are dry.

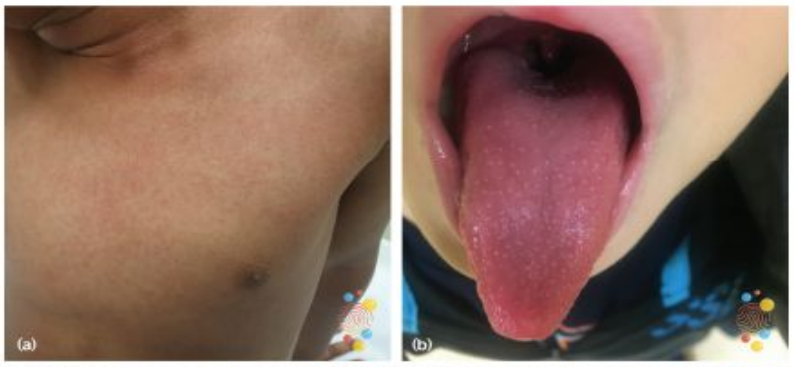


Figure 15.9 Scarlet fever. (**a**) The diffuse, erythematous macular-papular rash with a sandpaper texture, which appears shortly after the pharyngitis. (**b**) Red strawberry tongue. The tongue initially has a white coating, but desquamates to result in a red tongue with prominent papillae. (Courtesy of: Don't Forget the Bubbles.)

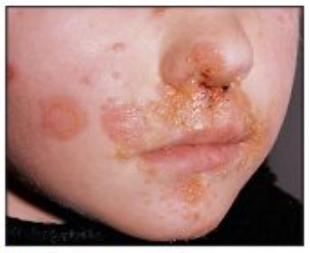


Figure 15.10 Impetigo showing characteristic confluent honey-coloured crusted lesions. (Courtesy of Dr Paul Hutchins.)

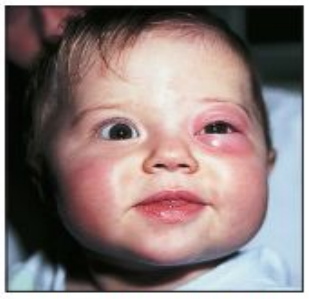


Figure 15.11 Periorbital cellulitis. It should be treated promptly with intravenous antibiotics to prevent spread into the orbit.

INFECTIOUS DISEASES

The human herpesviruses:

- There are currently eight known HHVs that cause infections in humans: HSV-1 and HSV-2, VZV, cytomegalovirus (CMV), EBV, HHV-6, HHV-7, and HHV-8.
- HHV-8 is associated with Kaposi sarcoma in HIV-infected individuals.
- The hallmark of most herpesviruses is that, after primary infection, latency is established and there is long-term persistence of the virus within the host, usually in a dormant state. After certain stimuli, reactivation of infection may occur.

Herpes simplex virus infections:

- HSV usually enters the body through the mucous membranes or skin, and the site of the primary infection may be associated with intense local mucosal damage. **HSV-1** is usually associated with lip and skin lesions, and **HSV-2** more commonly with genital lesions, but both viruses can cause both types of disease.

Summary

Herpes simplex virus infections

- Most are asymptomatic.
- Gingivostomatitis – may necessitate intravenous fluids and aciclovir.
- Skin manifestations – mucocutaneous junctions, e.g. lips and damaged skin.
- Eczema herpeticum – may result in secondary bacterial infection and septicaemia.
- Herpetic whitlows – painful pustules on the fingers.
- Eye disease – blepharitis, conjunctivitis, and corneal ulceration.
- CNS – aseptic meningitis, encephalitis.
- Pneumonia and disseminated infection in the immunocompromised.

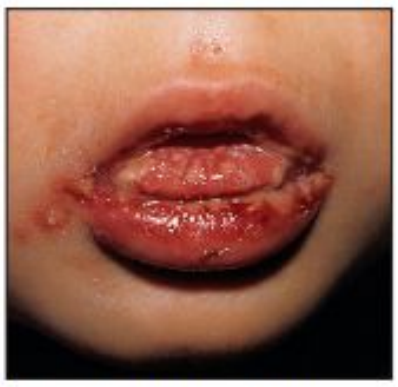


Figure 15.13 Vesicles with ulceration in gingivostomatitis.

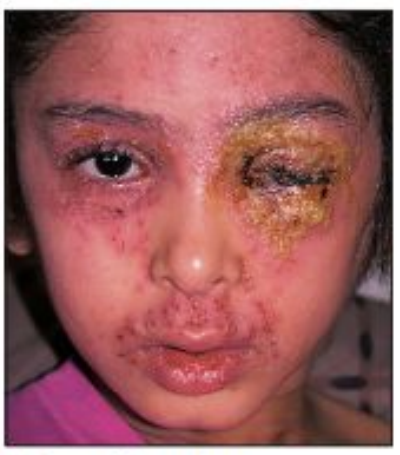


Figure 15.14 Eczema herpeticum.

INFECTIOUS DISEASES

Chickenpox (primary varicella zoster infection):

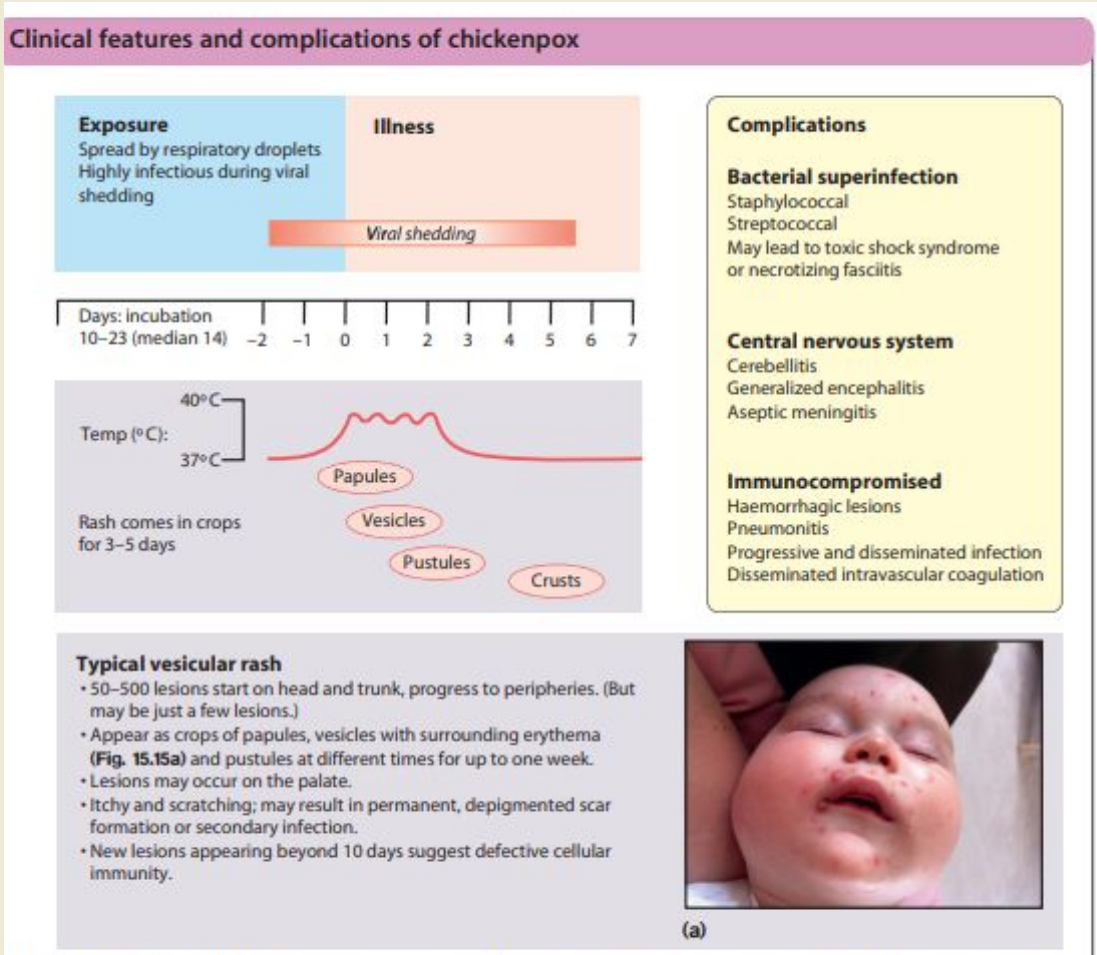


Figure 15.15 Clinical features and complications of chickenpox. (**a**) Vesicles with surrounding erythema appearing in crops are characteristic of chickenpox.

Watch for the child with chickenpox whose fever initially settles, but then recurs a few days later – this is likely to be due to secondary bacterial infection.

Beware of admitting a chickenpox contact to a clinical area with immunocompromised children, in whom it can disseminate and cause potentially fatal disease.

- Oral aciclovir has highly variable absorption and therefore limited benefit, and is not recommended in the UK.
- Immunocompromised children should be treated with intravenous aciclovir initially. Oral valaciclovir can be substituted at a later point if organ dissemination has not occurred.
- Human varicella zoster immunoglobulin is recommended as prevention for high-risk immunocompromised individuals with deficient T-cell function following contact with chickenpox. Protection from infection with human varicella zoster immunoglobulin is not absolute, and depends on how soon after contact with chickenpox it is given.

Shingles:

- is uncommon in children. It is caused by reactivation of latent VZV, causing a vesicular eruption in the dermatomal distribution of sensory nerves

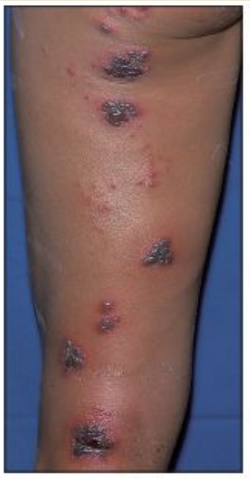


Figure 15.16 Herpes zoster (shingles) in a child. Distribution is along the S1 dermatome. (Courtesy of Dr Sam Walters.)

INFECTIOUS DISEASES

Human parvovirus B19:

- ❖ referred to as ‘slapped-cheek syndrome’. Infections can occur at any time of the year, although outbreaks are most common during the spring. Transmission is via respiratory secretions from affected patients, by vertical transmission from mother to fetus and by transfusion of infected blood products.
- ❖ HPV-B19 infects the erythroblastoid red cell precursors in the bone marrow.

HPV-B19 causes a range of clinical syndromes:

1. asymptomatic infection – common; about 5% to 10% of preschool children and 65% of adults have antibodies
2. erythema infectiosum – the most common illness, with a viraemic phase of fever, malaise, headache, and myalgia followed by a characteristic rash on the face (slapped cheek) a week later, progressing to a maculopapular, ‘lace’-like rash on the trunk and limbs
3. aplastic crisis – the most serious consequence of HPV-B19 infection; it occurs in children with chronic haemolytic anaemias, where there is an increased rate of red cell turnover (e.g. sickle cell disease or thalassaemia); and in immunocompromised children (e.g. with malignancy) who are unable to produce an antibody response to neutralize the infectious agent
4. fetal disease – transmission of maternal HPV-B19 infection may lead to fetal hydrops and death due to severe anaemia, although the majority of infected fetuses will recover

Mumps:

- ❖ Clinical features: The incubation period is 15 days to 24 days.
- ❖ Onset of the illness is with fever, malaise, and parotitis, but in up to 30% of cases, the infection is subclinical.
- ❖ Only one side of the face may be swollen initially, but bilateral parotid involvement may occur over the next few days. The parotitis is uncomfortable and children may complain of earache or pain on eating or drinking.
- ❖ Examination of the parotid duct may show redness and swelling. The fever usually disappears within 3 days to 4 days. Plasma amylase levels are often elevated due to parotid inflammation, and, when associated with abdominal pain, there may be evidence of pancreatic involvement.
- ❖ Infectivity is for up to 7 days after the onset of parotid swelling. The illness is generally mild and self-limiting. Although hearing loss can rarely follow mumps, it is usually unilateral and transient.

INFECTIOUS DISEASES

Measles:

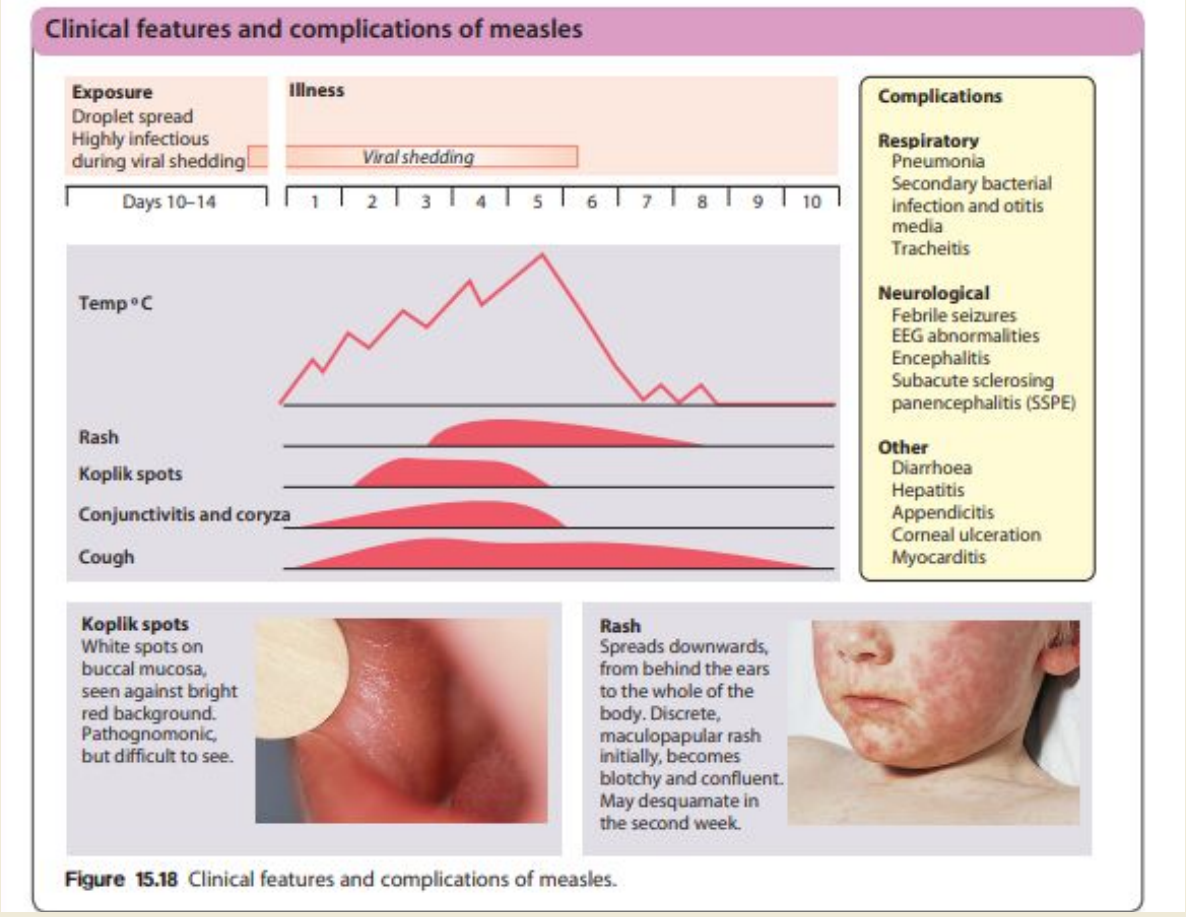


Figure 15.18 Clinical features and complications of measles.

- Treatment: for measles is supportive. Children who are admitted to hospital should be isolated. In immunocompromised patients, the antiviral drug ribavirin may be used. Vitamin A, which may modulate the immune response, should be given in low-income countries.

Rubella (German measles):

- The maculopapular rash is often the first sign of infection, appearing initially on the face and then spreading centrifugally to cover the whole body. It fades in 3–5 days.
- Lymphadenopathy, particularly the suboccipital and postauricular nodes, is prominent.
- Complications are rare in childhood but include arthritis, encephalitis, thrombocytopenia, and myocarditis.
- Clinical differentiation from other viral infections (including enteroviruses) is unreliable. The diagnosis should be confirmed serologically if there is any risk of exposure of a non-immune pregnant woman.
- There is no effective antiviral treatment. Prevention therefore lies in immunization.

LYME DISEASE :

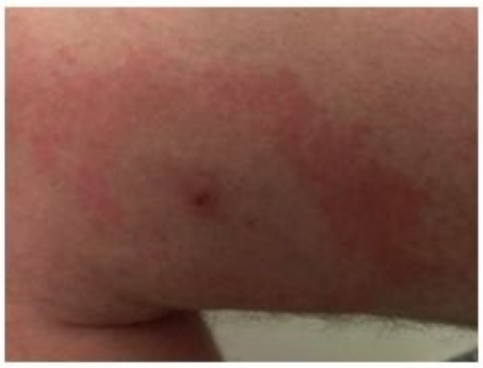


Figure 15.25 Lyme disease. Site of tick bite and red, expanding rash, erythema migrans. Further examples of the rash in Lyme disease can be seen in support of NICE Guideline on Lyme disease at https://www.nice.org.uk/guidance/ng95/resources/lyme-disease-rash-images-pdf-4792273597.

Case history 15.1

Kawasaki disease

This 2-year-old boy developed a high fever of 2 days' duration. Examination showed a miserable child with mild conjunctivitis, a rash, and cervical lymphadenopathy. A viral infection was diagnosed and his mother was reassured. When he presented to hospital 3 days later, he was noted to have cracked red lips (Fig. 15.20a). He was admitted and a full septic screen, including a lumbar puncture, was performed and antibiotics started. The following day, he was still febrile and irritable; his C-reactive protein and erythrocyte sedimentation rate were extremely high. Kawasaki disease was suspected and he was treated with intravenous immunoglobulin and oral aspirin. His clinical condition improved and he became afebrile the following morning. An echocardiogram at this stage showed no aneurysms of the coronary arteries, which are the most serious complication associated with delayed diagnosis and treatment. On the 15th day of the illness there was peeling of the fingers and toes (Fig. 15.20b).

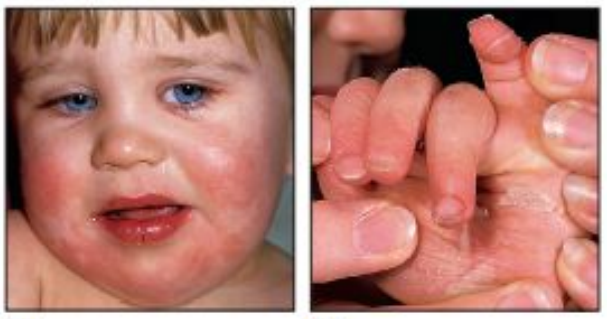


Figure 15.20 (**a**) Red, cracked lips and conjunctival inflammation; and (**b**) peeling of the fingers, which developed on the 15th day of the illness. (Courtesy of Professor Mike Levin.)

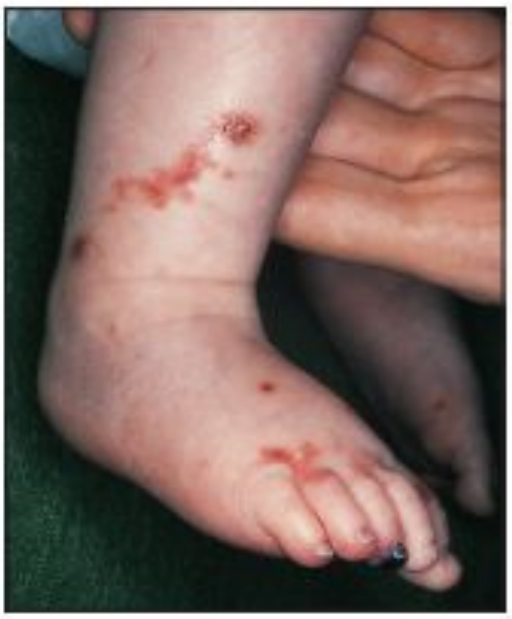


Figure 18.21 Widespread infected emboli and infarcts in a child with bacterial endocarditis. The tip of the third toe is gangrenous.

Table 10.2 The Apgar score

	Score		
	0	1	2
Heart rate	Absent	<100 beats/min	≥100 beats/min
Respiratory effort	Absent	Gasping or irregular	Regular, strong cry
Muscle tone	Flaccid	Some flexion of limbs	Well flexed, active
Reflex irritability	None	Grimace	Cry, cough
Colour	Pale/blue	Body pink, extremities blue	Pink

NEWBORN

Newborn infant physical examination

Box 10.6 Newborn infant physical examination (NIPE)

Birthweight, gestational age, and birthweight centile are noted (Fig. 10.14).

General observation of the baby's appearance, posture, and movements provides valuable information about many abnormalities. The baby must be fully undressed during the examination.

The head circumference is measured with a paper tape measure and its centile noted. Maximum of 3 measurements is recorded. This is a surrogate measure of brain size.

The fontanelles and sutures are palpated. The anterior fontanelle size is very variable. The sagittal suture is often separated and the coronal sutures may be overriding. A tense fontanelle when the baby is not crying may be due to raised intracranial pressure and cranial ultrasound should be performed to check for hydrocephalus.

The face is observed. If abnormal, this may represent a syndrome, particularly if other anomalies are present. Down syndrome is the most common, but there are hundreds of syndromes. When the diagnosis is uncertain, a book or a computer database may be consulted and advice should be sought from a senior paediatrician or clinical geneticist.

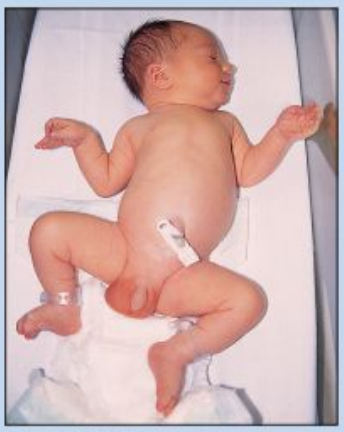


Figure 10.14 Term newborn.

If plethoric or pale, the haematocrit should be checked to identify polycythaemia or anaemia. Central cyanosis, which always needs urgent assessment, is best seen on the tongue.

Jaundice within 24 hours of birth requires further evaluation.

The eyes are checked for red reflex with an ophthalmoscope. If absent, may be from cataracts (see Case History 10.2), retinoblastoma and corneal opacity. This reflex can be hard to illicit in darker-skinned infants but the retinal vessels can be visualized.

The palate needs to be visually inspected, including posteriorly to exclude a posterior cleft palate, and palpated to detect an indentation of the posterior palate from a submucous cleft.

Breathing and chest wall movement are observed for signs of respiratory distress.

On auscultating the heart, the normal rate is 110–160 beats/minute in term babies, but may drop to 85 beats/minute during sleep.

On palpating the abdomen, the liver normally extends 1 cm to 2 cm below the costal margin, the spleen tip may be palpable, as may the kidney on the left side. Any intra-abdominal masses, which are usually renal in origin, need further investigation.

The femoral pulses are palpated. Their pulse pressure is:

- reduced in coarctation of the aorta. This can be confirmed by measuring the blood pressure in the arms and legs
- increased if there is a patent ductus arteriosus.

The genitalia and anus are inspected on removing the nappy. Patency of the anus is confirmed. In boys, the presence of testes in the scrotum is checked by palpation.

Muscle tone – observe for normal, symmetrical movements of all limbs; feel that it is normal when handling the baby; when held prone term babies lift their head to horizontal position.

If abnormal or asymmetrical, **primitive reflexes** may be checked (Fig. 3.4). For the Moro reflex, the head is allowed to extend suddenly, but supported in the examiner's hand. The arms spread in abduction and extension, followed by flexion and adduction. Some parents find it upsetting, and it does not provide additional information if full range of limb movements has been observed.

The whole of the back and spine is observed, looking for any midline defects of the skin.

The hips are checked for DDH. This is left until last as the procedure is uncomfortable.

Normal Development

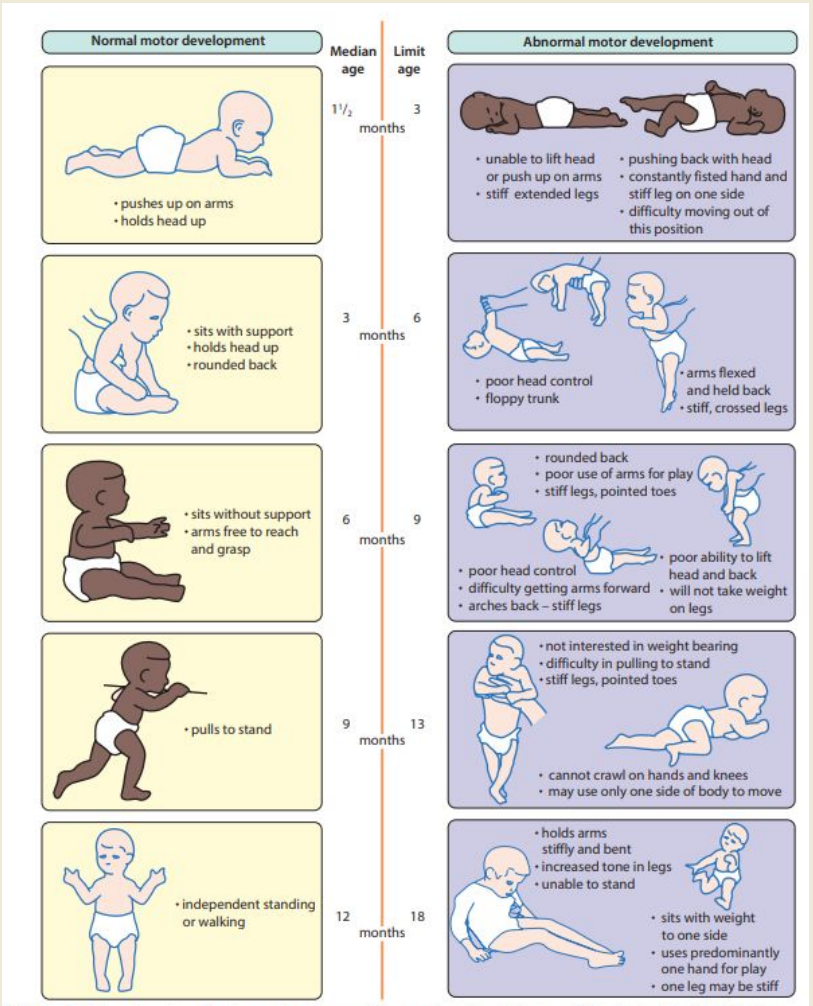


Figure 4.3 Normal motor milestones and patterns of abnormal motor development. Cerebral palsy (hemiplegia or quadriplegia) is the most common cause of developmental problems. (Adapted from: Pathways Awareness Foundatio Chicago, IL; see also pathways.org.)

Neurology

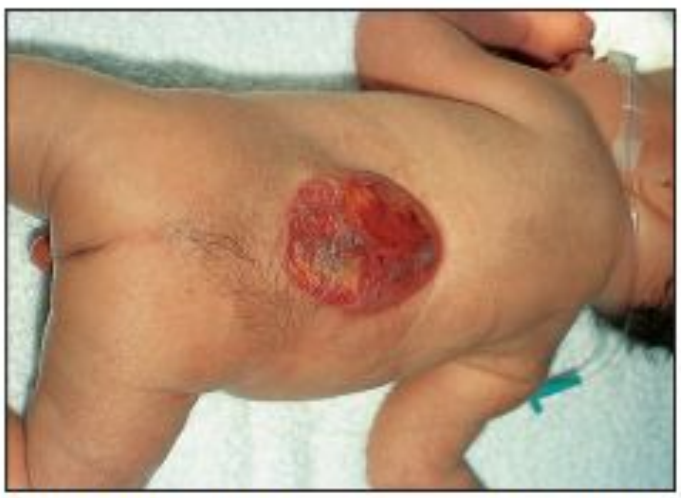


Figure 29.17 Myelomeningocele showing the exposed neural tissue and the patulous anus from neuropathic bowel.

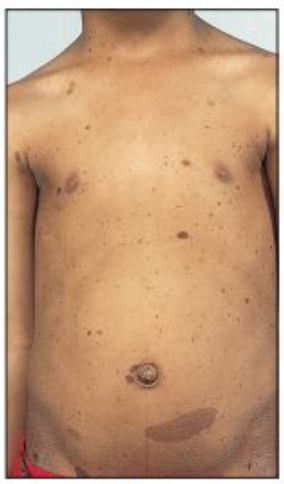


Figure 29.20 Café-au-lait patches and axillary freckling in neurofibromatosis.

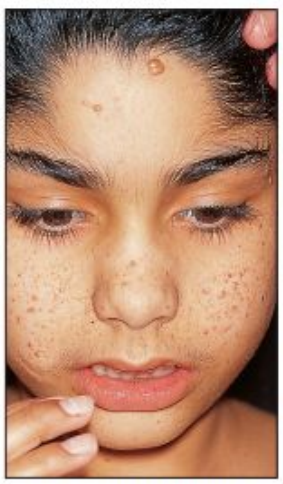


Figure 29.21 Facial angiofibromas in tuberous sclerosis.

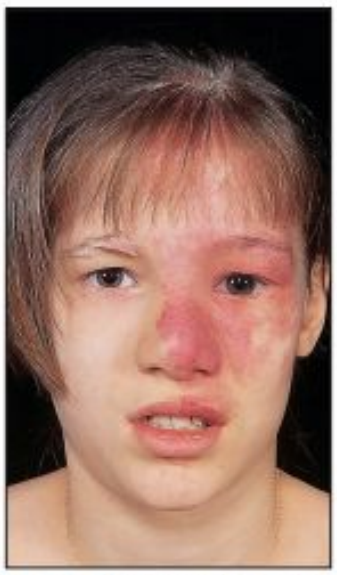


Figure 29.22 Sturge–Weber syndrome. There is a port-wine stain in the distribution of the trigeminal nerve.

Nephrology

Case history 19.3

Nephrotic syndrome

Zakariya developed periorbital oedema (Fig. 19.17) which improved during the day. He was seen by several doctors who diagnosed allergy, conjunctivitis, and hay fever. When he developed ascites and bilateral leg oedema his urine was dipsticked and showed 4+ protein and nephrotic syndrome was diagnosed. Periorbital oedema is often the initial sign of nephrotic syndrome but diagnosis is often delayed until other complications develop. Investigations performed are listed in Box 19.3. He is most likely to have steroid-responsive nephrotic syndrome, and the clinical course is outlined in Fig. 19.18.

Box 19.3 Investigations performed at presentation of nephrotic syndrome

- *Urine protein – on test strips (dipstick)* to confirm heavy proteinuria (≥3+ protein)
- *Full blood count* to assess whether there is an infection. Also a high haemoglobin suggests intravascular fluid depletion
- *Urea, electrolytes, creatinine, albumin* Hyponatraemia is common in presentation of nephrotic syndrome. Intravascular fluid depletion is indicated by high urea and/or creatinine
- *Complement levels – C3, C4* to differentiate from other causes of proteinuria such as postinfectious glomerulonephritis, when C3 will be low, or SLE, when both C3 and C4 are low
- *Antistreptolysin O or anti-DNAse B titres and throat swab* to differentiate from poststreptococcal glomerulonephritis
- *Urinary sodium concentration* which can be helpful to indicate intravascular fluid depletion if low (<10 mmol/L) in a child who is oedematous
- *Hepatitis B and hepatitis C screen* to detect secondary causes of nephrotic syndrome caused by hepatitis B and C viruses. This will also alter the treatment
- *Malaria screen if recent travel abroad* as may cause nephrotic syndrome

Figure 19.17 Facial oedema in nephrotic syndrome which improves during the day and is often misdiagnosed as an allergy.

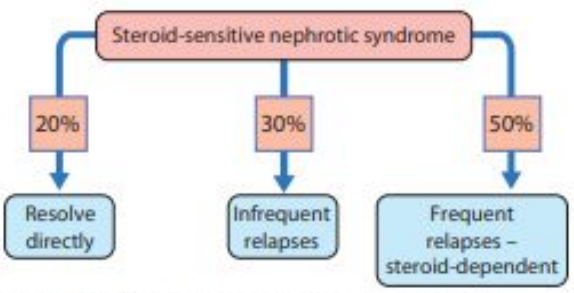


Figure 19.18 Clinical course in steroid-responsive nephrotic syndrome.

Hypernatremic dehydration:

- If intravenous fluids are required, a rapid reduction in plasma sodium concentration and osmolality will lead to a shift of water into cerebral cells and may result in seizures and cerebral oedema. The reduction in plasma sodium should therefore be slow, over at least 48 hours (with 0.9% saline or 0.9% saline with 5% glucose, tailored to response) and the plasma sodium measured regularly, aiming to reduce it at less than 0.5mmol/l per hour.

Physical Abuse

Case history 8.1

Severe physical abuse

A 2-month-old boy was brought into the emergency department by ambulance, with sudden loss of consciousness. His mother accompanying him appeared to have learning difficulties and could not explain what had happened. His father arrived soon after and said that he had been changing the child's nappy on the floor when suddenly he 'went all floppy and asleep'.

The child was unresponsive (U on AVPU) and had shallow breathing. His pupils were dilated. He appeared well nourished and was dressed only in a nappy. There were no obvious injuries seen.

Medical management was rapidly instituted. CT head scan showed subdural haemorrhages (Fig. 8.5). A chest X-ray obtained following intubation showed old posterior rib fractures (Fig. 8.6). Subsequent ophthalmological examination showed bilateral retinal haemorrhages (Fig. 8.7).

The child was transferred to an intensive care unit, where he died. A postmortem skeletal survey showed metaphyseal fractures (Fig. 8.8).

The parents maintained their story, despite the compelling evidence of inflicted head injury and shaking. The case went to the criminal court and both were sentenced on a number of charges.

Severe physical child abuse resulting in death gains considerable attention from the media but is rare, estimated at about 1 child per week in the UK. Many more children suffer permanent injury after serious physical abuse. Most have been seen previously by health professionals. Early recognition and response to child protection concerns could prevent severe injury.

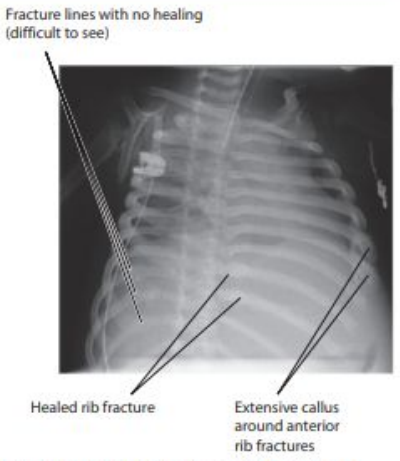


Figure 8.6 Multiple rib fractures of different ages.

Figure 8.7 Retinal haemorrhages from trauma to the head. (Courtesy of Clare Roberts.)

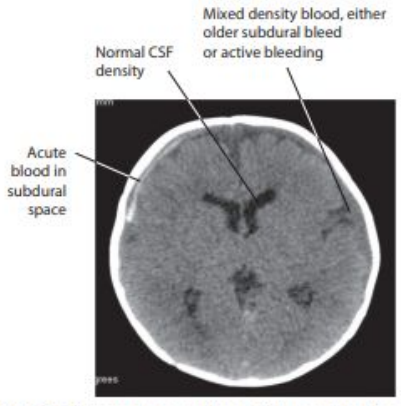


Figure 8.5 Subdural haemorrhage. CSF, cerebrospinal fluid.

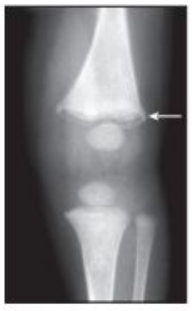


Figure 8.8 Metaphyseal fracture of distal humerus.

Table 8.3 Examples of injuries and a guide as to how likely it is due to an inflicted injury

Injury	More likely to be inflicted	May be inflicted, accidental or due to an underlying disorder	Less likely or unlikely to be inflicted
Fractures	Any fracture in a non-mobile child (excluding fragile bones) Rib fractures Multiple fractures (unless significant accidental trauma, e.g. road traffic accident) Multiple fractures of different ages	Skull fracture in young child Long bone fractures in a young but mobile child	Fracture in school-age child with witnessed trauma, e.g. fall from swing
Bruises	Bruising in the shape of a hand (Fig. 8.9a) or object Bruises on the neck that look like attempted strangulation Bruises around the wrists or ankles that look like ligature marks Bruise to the buttocks in a child less than 2 years or any age without a good explanation	Bruising to the trunk with a vague history	Bruises on the shins of a mobile child
Burns	Any burn in a child who is not mobile. A burn in the shape of an implement – cigarette, iron A 'glove or stocking' burn consistent with forced immersion (Fig. 8.9b)		A burn to mobile toddler with splash marks, a history of pulling drink onto himself – but may indicate neglect in the form of poor supervision
Bites	Bruising in the shape of a bite thought unlikely to have been caused by a young child (Fig. 8.9c)		A witnessed biting of one toddler by another

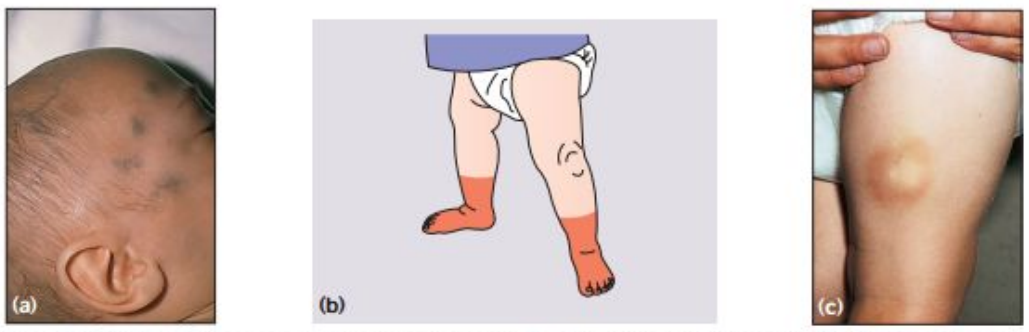


Figure 8.9 (**a**) Bruising from finger trauma to a baby's head. (**b**) Scald with stocking distribution including the soles from forced immersion in hot water. (**c**) A bite mark on an infant's leg. Adult bite marks may be seen in abuse, but bites from other children are not uncommon.

Transient tachypnoea of the newborn:

- This is by far the most common cause of respiratory distress in term infants.
- It is caused by delay in the resorption of lung liquid and is more common after birth by caesarean section.
- The chest X-ray may show fluid in the horizontal fissure. Supplemental oxygen may be required in addition to feeding support with nasogastric feeds or IV fluids if the neonate is unable to feed normally.
- The condition usually settles within the first day of life but can take several days to resolve completely. This is a diagnosis made after consideration and exclusion of other causes such as infection.

Hypoglycaemia

- Hypoglycaemia is particularly likely in the first 24 hours of life in babies with intrauterine growth restriction, who are preterm, born to mothers with diabetes mellitus, are large for-dates, hypothermic, polycythaemic, or ill for any reason.
- Growth-restricted and preterm infants have poor glycogen stores, whereas the infants of a diabetic mother have sufficient glycogen stores, but hyperplasia of the islet cells in the pancreas from exposure to elevated maternal glucose causes increased insulin levels. Symptoms are jitteriness, irritability, apnoea, lethargy, drowsiness and seizures.
- Some studies suggest that blood glucose levels above 2.6mmol/l are desirable for optimal neurodevelopmental outcome
- There is good evidence that prolonged, symptomatic hypoglycaemia can cause permanent neurological disability. Hypoglycaemia can usually be prevented by early and frequent milk feeding. In infants at increased risk of hypoglycaemia, blood glucose is regularly monitored prefeeds at the bedside.
- Management recommend that if the blood glucose concentration is 2–2.6mmol/L, the level is rechecked until satisfactory (at least 3 prefeed levels >2mmol/L), whereas if the level is <2mmol/L the infant is given dextrose gel to the mouth and the level checked (after 30 mins). If the infant has a very low blood glucose (<1.0mmol/L) or <2.0mmol/L and clinical signs or has not responded adequately to two doses of glucose gel, hypoglycaemia should be corrected immediately with an intravenous infusion of dextrose.

Hypoxic–ischaemic encephalopathy

- ❖ HIE may occur if there is:
- ➢ failure of gas exchange across the placenta – excessive or prolonged uterine contractions, placental abruption, ruptured uterus
- ➢ interruption of umbilical blood flow – cord compression including shoulder dystocia, cord prolapse
- ➢ inadequate maternal placental perfusion – maternal hypotension or hypertension • compromised fetus – intrauterine growth restriction, anaemia
- ➢ failure of cardiorespiratory adaptation at birth – failure to breathe.

- ❖ The clinical manifestations start immediately or up to 48 hours afterwards, and can be graded:
- ➢ mild (grade 1) – the infant is irritable, responds excessively to stimulation, may have staring of the eyes, and hyperventilation
- ➢ moderate (grade 2) – the infant shows marked abnormalities of movement, is hypotonic, cannot feed as cannot suck, may have brief apnoeas and may have seizures
- ➢ severe (grade 3) – there are no normal spontaneous movements or response to pain; tone in the limbs is hypotonic; seizures are prolonged and often refractory to treatment; multi-organ failure is present.

- ❖ Randomized clinical trials have shown that **mild hypothermia** (cooling to a rectal temperature of 33°C to 34°C for 72 hours by wrapping the infant in a cooling jacket) for infants 36 weeks' gestation and over with moderate or severe HIE reduces brain damage if started within 6 hours of birth

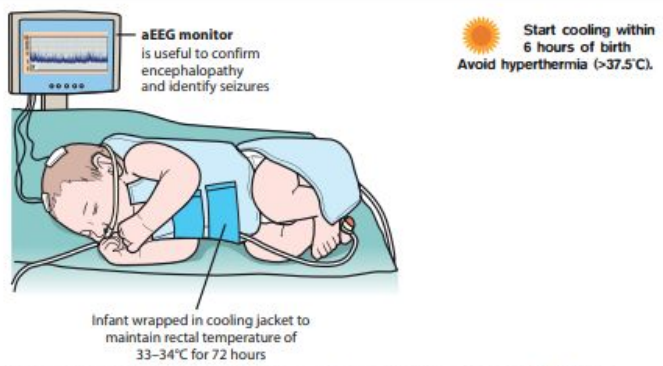


Figure 11.22 Therapeutic hypothermia for moderate or severe hypoxic–ischaemic encephalopathy.

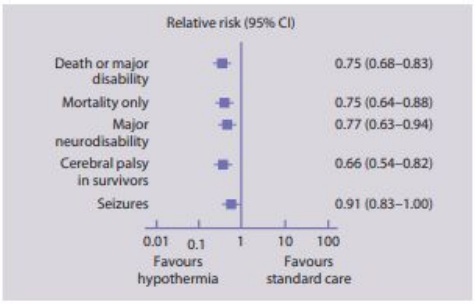


Figure 11.23 Outcomes of therapeutic hypothermia trials compared with standard care for the treatment of term/near-term babies with hypoxic–ischaemic encephalopathy. The figure shows reduction of death or major disability. Reduction in seizures is not demonstrated. (CI, confidence interval.) (Data from: Jacobs SE, Berg M, Hunt R, Tarnow-Mordi WO, Inder TE, et al: Cooling for newborns with hypoxic-ischaemic encephalopathy. *The Cochrane Database of Systemic Reviews* CD003311, 2013.)

Mild hypothermia for moderate and severe HIE reduces death and severe disability and increases the likelihood of survival with normal neurological function.

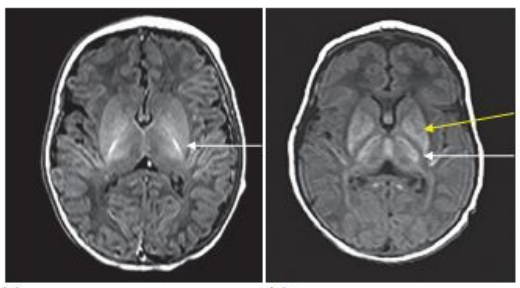


Figure 11.24 Magnetic resonance image (T1 in axial view) of the brain at 14 days in a term infant. (**a**) Normal scan for comparison, showing high signal in the posterior limb of the internal capsule (PLIC) (arrow). (**b**) Following severe HIE, showing loss of the normal high signal from myelin in the internal capsule (PLIC) (white arrow) and abnormal high signal in the adjacent basal ganglia and thalami (yellow arrow). These findings would be associated with a severe motor impairment in the form of cerebral palsy, poor head growth, persistent feeding difficulties, seizures and marked cognitive impairment. (Courtesy of Professor Mary Rutherford.)

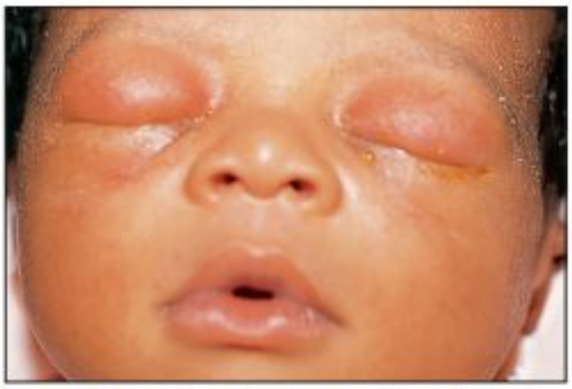


Figure 11.20 Purulent discharge, together with swollen eyelids, in an 8-day-old infant. This is the characteristic presentation of conjunctivitis from *Chlamydia trachomatis*. *Neisseria gonorrhoeae* was absent.

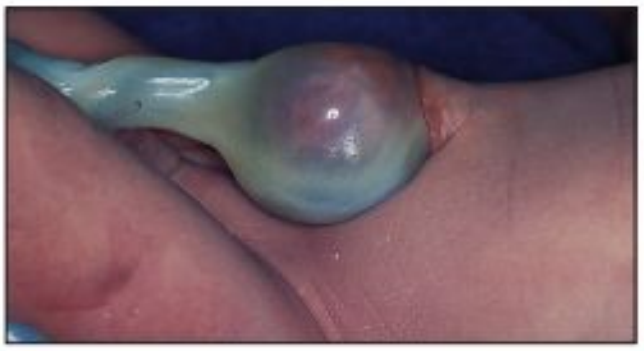
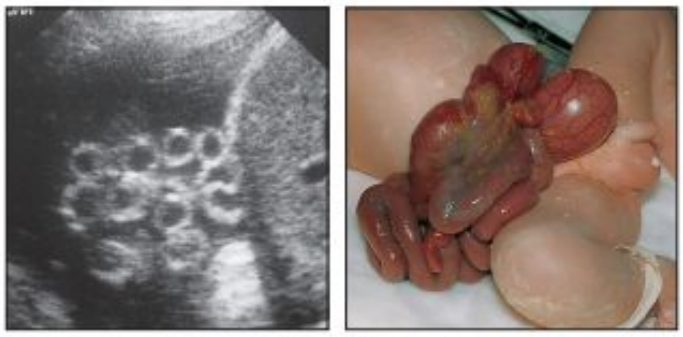


Figure 11.34 Small exomphalos with loops of bowel confined to the umbilicus. Care needs to be taken not to put a cord clamp across these lesions.

Example of antenatal diagnosis – gastroschisis



(a) (b)

Figure 10.2 Gastroschisis on antenatal ultrasound showing free loops of small bowel outside the fetal abdomen, in the amniotic fluid (**a**) and following delivery (**b**). Antenatal diagnosis allowed the baby to be delivered at a paediatric surgical unit and the parents to be counselled antenatally by a paediatric surgeon. Satisfactory surgical repair was achieved. (Courtesy of Karl Murphy.)