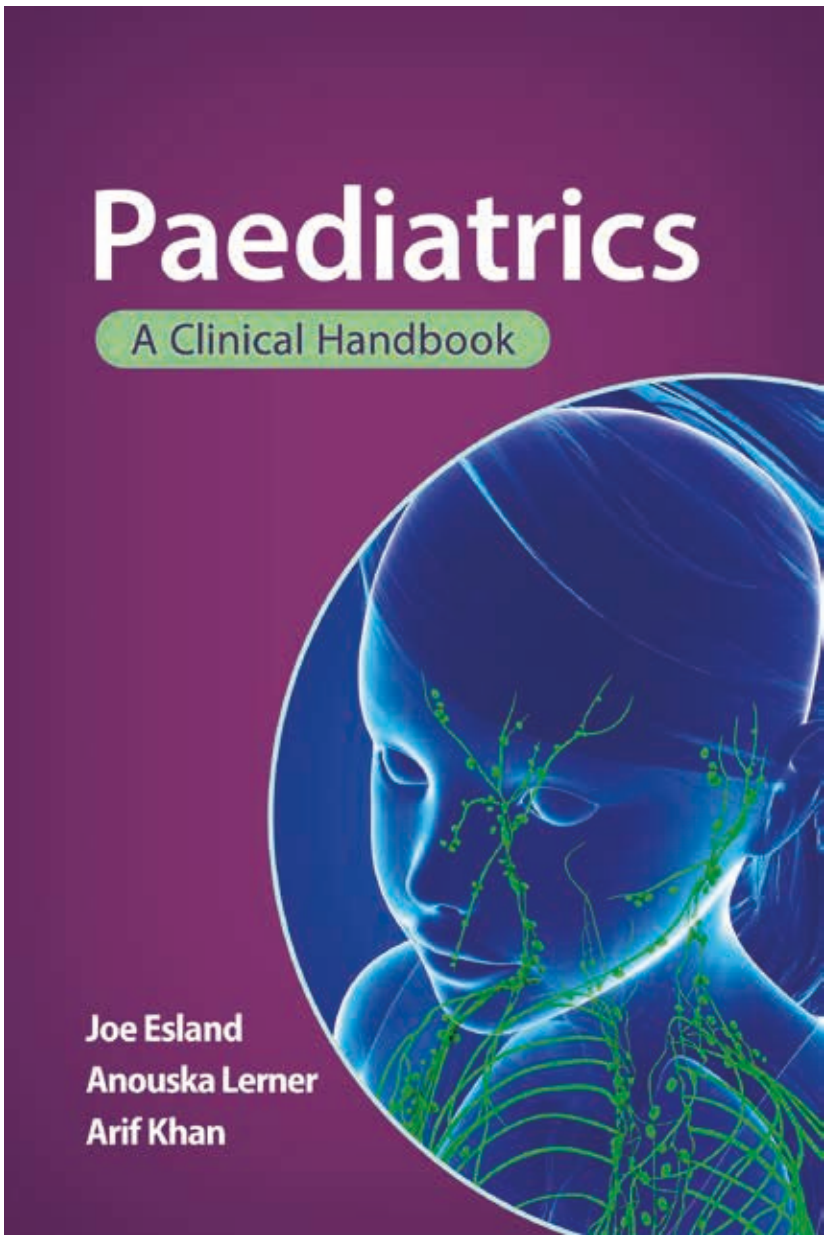
Answers to self-assessment questions



Short Answer Questions

1. An 18-month-old child presents with a 3-day history of diarrhoea and vomiting. They look well from the end of the bed. You diagnose viral gastroenteritis but are concerned that they are dehydrated.

A. Broadly, the clinical features of dehydration can be categorized into which two groups?

Interstitial volume depletion and intravascular volume depletion.

B. How is the severity of dehydration graded?

Hydration is graded based on the percentage loss of body weight as water: mild (<5%), moderate (5–10%) and severe (>10%).

C. What specific clinical signs would you assess?

Begin by assessing their consciousness level from the end of the bed. Their observations are gained, looking for tachycardia, tachypnoea and hypotension. Look at their extremities, assessing for delayed capillary refill time and cool extremities. Assess their face, looking for sunken eyes, a dry tongue and dry mucous membranes. Check for reduced skin turgor by pinching the skin of the abdomen.

D. The child's mucous membranes are dry, but the rest of the clinical examination is unremarkable. You decide they can be managed at home; what important complication must the parents be made aware of?

Hypovolaemic shock.

2. You are called to see a 2-hour-old neonate who, the nurse reports, is 'grunting'. They were delivered at 34/40 to a diabetic mother, but the pregnancy was otherwise uncomplicated. When you arrive, the patient is grunting, tachypnoeic and distressed. You think they have respiratory distress syndrome (RDS).

A. What are the major risk factors for RDS?

The most important risk factor is prematurity. Other risk factors include maternal diabetes and any cause of perinatal asphyxia (e.g. meconium aspiration).

B. Describe the role of surfactant in the lung.

Surfactant reduces alveolar surface tension, so permitting easy ventilation as well as preventing the alveoli from collapsing in expiration.

C. What are the main features of respiratory distress in a neonate?

Head bobbing, nasal flaring, tracheal tug and intercostal recession. Tachypnoea ± hypoxia.

D. What investigations are needed and what would they show?

An urgent blood gas is sent, which would demonstrate hypoxaemia and, if this leads to end organ hypoperfusion, a metabolic (lactic) acidosis. A CXR classically demonstrates bilateral 'ground glass' infiltrates with an air bronchogram.

E. What is the initial treatment for this neonate?

The initial emergent treatment is with oxygen. Thereafter, intratracheal surfactant is usually required, thus necessitating intubation and ventilation.

3. A premature neonate presents at 10 days of age with vomiting, bloody diarrhoea and a distended abdomen. They look unwell and are febrile.

A. What is the most important differential to consider in this patient?

Necrotizing enterocolitis.

B. What investigations are required?

A blood gas would demonstrate a metabolic acidosis. Routine bloods are sent, which should include a CRP. An abdominal X-ray is necessary and would show pneumatosis intestinalis (intramural gas in the bowel wall). If the bowel has perforated, a pneumoperitoneum will be present.

C. What initial treatment is required?

Urgent treatment is required as mortality is high. Stop oral feeding and place an NG tube to decompress the stomach. Switch the child to TPN and, where required, supplement this with additional IV fluids. Prescribe IV antibiotics.

D. When is surgical intervention required?

When conservative measures fail to improve the clinical (or biochemical) state of the child, or if there are features of bowel perforation.

E. What are the most important differential diagnoses?

Hirschsprung disease, volvulus and intestinal perforation (of any cause).

4. A 12-month-old infant is referred for assessment because they are not gaining weight at the expected rate.

A. What is the specific definition of 'failure to thrive' (FTT)?

Failure to thrive is defined as a child whose weight crosses ≥2 centiles on a growth chart, or who is persistently <5th centile.

B. Broadly, the causes of FTT can be divided into 4 groups. Describe these and give example aetiologies.

Poor intake is an important cause, which may be environmental (e.g. inadequate food supply, neglect, abuse) or physical (e.g. cleft palate or an impaired swallow). If intake is adequate it may be that they are unable to absorb this nutrition – for example, in cystic fibrosis. Lastly, their energy requirements may be increased due to pathology (e.g. heart disease or hyperthyroidism) so, despite adequate nutrition, their intake may still be inadequate.

C. Describe how a child's growth is formally assessed.

Height (if >2 y/o) or length (if <2 y/o) is measured. Weight is measured. Head circumference is measured – this is done 3 times, measuring between the brow and the occiput. The largest measurement is recorded.

D. The clinical examination and investigations are unremarkable, so you think the most likely cause is non-organic and simply recommend increasing the child's calorie intake. Which healthcare professional is of high importance in ensuring this is managed well?

A dietitian.

5. A 4-month-old child is brought to the Emergency Department with a deformed left humerus. Whilst examining them you notice bruising behind the ears. The mother states that she thinks the child crawled off a kitchen surface yesterday, although she didn't see it happen.

A. What features of the history are concerning for child abuse?

There are many red flags in this story. There is a delay in presentation and the injury was unwitnessed. This child is non-ambulant, so crawling is unlikely. Additionally, falling from a height would not explain bruising behind the ears.

B. What important differential diagnosis should you consider in a child with multiple bruises?

Coagulopathies are important to exclude in this presentation.

C. What important differential diagnoses should you consider in a child presenting with a fracture?

Osteogenesis imperfecta, malignancy and osteomyelitis can all result in paediatric fractures.

D. What are some essential initial steps that you must take, as the junior doctor, in this situation?

Your role in this case is to recognize the at-risk patient and ensure that you have escalated this in a timely fashion to the lead doctor for child safeguarding. Document your consultation clearly and precisely. Request some initial basic investigations, such as blood tests and radiographs.

6. A 3-year-old child presents to the Emergency Department after her parents notice that she's become less responsive, is feeding poorly and has a temperature of 39.6°C.

A. How should the temperature be measured in children?

Either the tympanic membrane or the axilla.

B. Using a systematic approach, what are the differential diagnoses for children with an acute fever?

The differential can be considered using a body-systems-based approach. Examples include, but are not limited to:

- **CNS: meningitis, encephalitis**
- **ENT: otitis media, sinusitis, URTI, epiglottitis**
- **Respiratory: pneumonia, croup, bronchiolitis**
- **Cardiac: infective endocarditis**
- **GI: gastroenteritis, mesenteric adenitis**
- **Hepatobiliary: hepatitis, cholecystitis**
- **Genitourinary: UTI, pyelonephritis**
- **MSK: septic arthritis**

C. What basic investigations should be considered?

Bloods (including CRP, blood gas and blood cultures), urine dip, CXR, lumbar puncture.

D. Investigations and management are guided by assessing the seriousness of the illness. What system has been devised to assist with this?

Seriousness of illness assessment is guided by using the 'traffic light' system, described by NICE (2013, CG160). This can be used to determine the initial investigations that are required, as well as which patients are safe for outpatient management.

E. When are antipyretics indicated in the management of an acute fever?

They are indicated when the patient is distressed by the pyrexia; however, if the child is not, they can be omitted.

7. A 10-year-old boy attends his GP practice with itchy eyes and sneezing since a cat has been brought into the family home.

A. What is the likely diagnosis?

Allergic rhinitis due to animal dander. Other common causes include pollens and dust mites.

B. What is the pathophysiology of this condition?

This condition is a type 1 hypersensitivity reaction, mediated initially by the acute degranulation of mast cells. Later, eosinophils, basophils and T cells are involved.

C. What are the most important risk factors that should be asked for in the history?

A personal or family history of atopic conditions is important to ascertain (i.e. allergic rhinitis, asthma, eczema).

D. What are the most important initial treatment options for allergic rhinitis?

Limiting exposure to the allergen will greatly improve symptoms. Medical therapy may include antihistamines (e.g. loratadine) and sodium chromoglycate eye drops, for itchy eyes.

8. A 6-year-old child presents to the Emergency Department with a sore throat and an inability to swallow their saliva for the last 3 hours. They look unwell and are febrile. You discover that the child has not received any vaccinations and suspect epiglottitis.

A. Describe the basic pathophysiology of epiglottitis.

Most commonly, epiglottitis is cause by *Haemophilus influenzae B* (Hib), although other bacterial, viral and environmental insults (e.g. heat) can cause it. The epiglottis becomes rapidly swollen which, if left, can lead to pharyngeal obstruction.

B. Describe the early and late symptoms of epiglottitis.

These can be remembered using the '4 Ds'. Early symptoms include dysphagia and drooling. Late symptoms include dyspnoea and dysphonia.

C. What are some important examination findings in epiglottitis?

The child is febrile and typically adopts the 'tripod' position (leaning forward with both arms out and their tongue protruding). They are febrile. There will be signs of respiratory distress (nasal flaring, tracheal tug, intercostal recession) and, when there is impending upper airway compromise, stridor will be heard. It is important that you do not try to directly assess the pharynx if epiglottitis is suspected, as this can cause reflex laryngospasm.

D. Where should this patient be managed?

On the intensive care unit.

E. What treatments are required?

Securing the airway is the first priority, so intubation is typically required. Oxygen and nebulized adrenaline can be used acutely. IV steroids and IV antibiotics are given.

9. A neonate with Down syndrome has not passed meconium within 48 hours of birth. You suspect Hirschsprung disease.

A. Which other diagnosis is an important differential for the delayed passage of meconium?

Meconium ileus, of which 90% have cystic fibrosis.

B. Describe the pathophysiology of Hirschsprung disease.

There is an absence of ganglion cells in the myenteric plexus of the bowel wall, resulting in absent/ineffective peristalsis and muscle spasms of the anal sphincter. This causes a functional obstruction of the bowel. It is important to recognize that, *in utero*, the ganglion cells migrate in a craniocaudal fashion; consequently, the distal bowel is affected most frequently.

C. On examination, the child is febrile, with a distended, tender abdomen. Why are these clinical features of high importance?

Hirschsprung disease can be complicated by enterocolitis, which can lead to bowel perforation and sepsis. If the described clinical features are present, the neonate requires urgent surgical intervention (typically for diversion).

D. What is the definitive investigation for diagnosis?

Rectal biopsy.

E. How is uncomplicated Hirschsprung disease treated?

Initially, decompress the bowel by siting a nasogastric tube. Saline enemas are commenced on a routine basis. Definitively, surgery is required to resect the aganglionic bowel segment.

10. At birth, a male neonate is found to be clinically cyanosed and looks unwell. Their saturations are 82% on room air.

A. Broadly, how can the aetiologies of cyanotic heart disease be grouped?

Cyanosis is seen if sufficient deoxygenated/insufficient oxygenated blood enters the systemic circulation. The broad underlying causes can be grouped as follows:

- **Right-to-left shunts (e.g. tetralogy of Fallot or transposition of the great arteries). In these cases, deoxygenated blood bypasses the lungs and enters the systemic circulation.**
- **Mixed shunt (e.g. atrioventricular septal defect), resulting in intracardiac mixing of oxygenated and deoxygenated blood.**
- **Outflow obstruction defects (e.g. hypoplastic left heart syndrome) are reliant on either/both a septal defect or a patent ductus arteriosus to allow blood to move between the left and right circulations. This permits mixing of oxygenated and deoxygenated blood, leading to cyanosis.**

B. The patient is found to have transposition of the great arteries (TGA). Describe the anatomy found in this disease.

The aorta and pulmonary arteries arise from the incorrect ventricle: the aorta from the right ventricle and the pulmonary artery from the left. This means that the left

and right circulations remain completely separate and, to permit mixing of blood and therefore sustain life, a septal defect and/or a patent ductus arteriosus is required.

C. What are the characteristic features on the CXR?

The heart is described as looking like an 'egg-on-a-string' – the mediastinum is narrowed and the heart is globular.

D. What measures are taken to ensure ongoing mixing of the left and right circulations?

Acutely, a prostaglandin infusion will help to maintain patency of the ductus arteriosus, and a balloon septostomy can be performed to create a ventricular septal defect.

E. What is the definitive treatment required?

An arterial switch is required, typically performed at ~3 weeks of age.

11. A 5-month-old infant presents to the Emergency Department with lethargy and irritability. They are not feeding well and they have a fever. A urine sample is sent and is positive for *E. coli*. This is the 4th episode since birth.

A. How would you describe this urinary tract infection (UTI)?

As the child has now had ≥3 episodes, this is a 'recurrent' infection.

B. What are the important risk factors to consider in this patient?

It is essential to consider an underlying anatomical structural defect (e.g. duplex kidney or horseshoe kidney) and vesicoureteric reflux (VUR).

C. Which investigations are required for this patient?

Initially, a urine sample should be sent for MC&S. Bloods should be sent. Imaging for children <6 months old is directed by the NICE guidelines. In this case, they require an acute ultrasound (USS), a DMSA (dimercaptosuccinic acid) scan and an MCUG (micturating cystourethrogram) to look for structural defects within the urinary tract.

D. What acute treatment is required?

Empirical antibiotics are required until the MC&S results are back. Symptomatic treatment should be prescribed, to treat pain and fever, if distressing the child.

E. What are some preventative measures that can be initiated, to try to reduce recurrence?

Treatment of any underlying structural defect is important. Prophylactic antibiotics should be considered. Treat constipation, if required, and give advice on good oral intake, hygiene and regular voiding.

12. An 8-year-old girl presents to the Emergency Department with a headache and photophobia. She has sickle cell disease, but is otherwise well. You suspect meningitis.

A. What do you think are the likely causal organisms in this case and why?

Children with sickle cell disease are more prone to infection by encapsulated bacteria, such as *Neisseria meningitides* and *Streptococcus pneumoniae*.

B. Describe the clinical tests performed to assess meningeal irritation.

There are two common tests performed:

- **Kernig sign: the child is supine and has the hip and knee passively flexed to 90°. The knee is then passively straightened, which produces pain along the spine.**

- **Brudzinski sign: the child is again supine. This time the neck is passively flexed, which produces a reflex flexion of their hips.**

C. You send a panel of bloods, blood cultures and perform a lumbar puncture. Describe the appearance, white cells present, protein content and glucose content of the CSF in bacterial meningitis.

In bacterial infection, the CSF appears turbid and has neutrophils present. The protein content is high and the glucose content is low. In comparison, in viral disease, the CSF in clear and contains lymphocytes. Protein content is normal/mildly elevated and the glucose content is normal/mildly low.

D. When is treatment for meningitis commenced and what does it include?

Treatment with IV antibiotics is started empirically immediately; you should not wait for the results of diagnostic tests, even if you think the cause is likely to be viral. IV corticosteroids are given in children >3 months old.

E. What are the main complications of meningitis?

These can be remembered using the mnemonic 'SAD REP is Deaf':

- **Sepsis/Shock/SIADH**
- **Ataxia/Abscess**
- **DIC**
- **Retardation**
- **Epilepsy**
- **Paralysis**
- **Deafness**

13. A 7-year-old child presents to the GP practice with a painful limp.

A. You decide to take a history and ask specifically about 'red flags'. What 'red flag' questions do you need to ask, and which broad pathologies do these questions aim to address?

The 'red flag' questions aim to identify malignancy, inflammatory arthropathies, musculoskeletal infection, coagulopathy and non-accidental injury. These can be summarized as follows:

- **Worse:**
  - **in the morning (→ ?inflammatory arthropathy)**
  - **at night (→_?malignancy)**
- **Systemically unwell, i.e. night sweats, weight loss (→ ?malignancy, infection, inflammatory)**
- **Redness and swelling over a joint (→ ?infection or inflammatory)**
- **Unexplained rashes or bruises (→ ?coagulopathy or ?NAI)**

B. The child has been feeling non-specifically unwell and feverish. They recently had a short course of antibiotics for an infected wound that they sustained whilst out playing. What diagnosis would you suspect?

With this history, the most likely cause is septic arthritis secondary to haematogenous spread from a soft tissue infection.

C. What clinical signs would you expect to find on examination?

The joint is typically warm, with overlying erythema. It is often held in slight flexion and the child will not let you move it (termed 'pseudoparalysis').

D. How would investigate this case?

Begin with blood tests (including a CRP and ESR), send blood cultures and request radiographs. The diagnosis is confirmed with aspiration of the affected joint and sending the synovial fluid sample for MC&S.

E. What is the treatment for septic arthritis?

The treatment is urgent surgical washout and a prolonged course of antibiotics.

14. A 14-year-old child presents with severe pain in their lower back and left leg. They are known to have sickle cell disease.

A. Describe the underlying pathophysiology of sickle cell disease (SCD).

SCD is a result of an amino acid substitution (glutamine to valine) on the beta-globin chain. Affected units are prone to polymerization in low oxygen tension, resulting in abnormally shaped and mechanically weak red blood cells (RBCs). These RBCs can occlude the microvasculature and undergo haemolysis.

B. What is the cause of this child's presentation and what are the normal precipitants?

This is a typical presentation of a vaso-occlusive crisis, in which the sickled RBCs acutely occlude the microvasculature. Precipitants include infection, cold, hypoxia, dehydration, stress and medications.

C. What investigations might you perform?

Blood tests would show anaemia and a high reticulocyte count. A renal profile should be sent. If you suspect the trigger was infection, ensure you send appropriate bloods (e.g. CRP and ESR) and perform tests to identify the aetiology (e.g. CXR and urine sample).

D. What treatment is required for a vaso-occlusive crisis?

The most important initial steps are to ensure that the child is kept warm, is hydrated (with IV fluid if required) and has ample analgesia (this is typically with an opioid) – these should all be initiated expeditiously.

15. A 6-year-old child presents to the Emergency Department with abdominal pain and a non-blanching rash affecting the buttocks and legs. You suspect that the patient has Henoch–Schönlein purpura (HSP).

A. Describe the basic pathophysiology of HSP.

HSP is an IgA-mediated vasculitis, characterized by IgA deposition in the skin, joints, GI tract and kidneys.

B. What is the 'tetrad' of HSP?

The clinical features can be thought of as a tetrad:

- **Purpuric rash, often affecting the buttocks and extensor surfaces of the lower limbs**
- **Arthralgia, typically affecting the lower limbs**
- **Abdominal pain**
- **Renal disease, ranging from AKI and nephrotic syndrome to haematuria**

C. What are some important differentials for a non-blanching rash?

Meningococcal septicaemia, thrombocytopenia (e.g. ITP, HUS), coagulopathy, acute leukaemia, physical abuse.

D. What investigations are required?

A urine sample to look for renal involvement. Blood tests, to assess renal function, clotting and inflammatory markers.

E. What is the mainstay of treatment?

Treatment is generally supportive, with simple analgesia and anti-inflammatories. Where there is renal involvement, treatment is with corticosteroids ± immunosuppression.

16. A pregnant woman is at her 12-week pregnancy dating scan. She is offered the triple test to screen for Down syndrome.

A. What markers are measured in the triple test and how are they altered in a high-risk pregnancy?

Alpha-fetoprotein (AFP) is low, human chorionic gonadotrophin (hCG) is elevated and unconjugated oestradiol (UE_3) is low.

B. What are some of the common phenotypic features in Down syndrome involving the:

i. face

Brachycephaly, flat nasal bridge, upslanting palpebral fissures, macroglossia

ii. hands and feet

Single palmar crease, large 'sandal-gap' between the 1st and 2nd toes.

C. State some common complications affecting the following body systems in Down syndrome:

i. Gastrointestinal

Duodenal atresia and imperforate anus are common, as is Hirschsprung disease.

ii. Cardiovascular

Septal defects are seen in 30–50% of children with Down syndrome.

iii. Neurological

Early-onset dementia affects ~50% of patients. Learning disability is present.

iv. Endocrine

Hypothyroidism affects ~10% of patients.

17. A 2-year-old child is brought in to the Emergency Department with a persistent cough, which followed an unwitnessed bout of sudden coughing and choking the previous day.

A. What is the likely aetiology for this child's cough?

In children under 3 years of age, a sudden coughing/choking episode should arouse suspicion for an aspirated foreign body.

B. What are the sequelae of an aspirated foreign body?

The foreign body produces an inflammatory reaction, which can lead to airway obstruction – this may cause atelectasis, hyperinflation of the lung or secondary infection. Additionally, the foreign body can cause airway ulceration.

C. If a CXR does not demonstrate a foreign body, what is the gold standard investigation for diagnosis?

Bronchoscopy, during which the foreign body can also be retrieved.

D. What preventative measures can be recommended to reduce the incidence?

Parents should be advised to keep small objects out of reach. Regarding aspirated food, the child should not be allowed to eat easily-aspirated food until they can safely chew, they should sit whilst eating, and they should not be allowed to play whilst eating.

18. A 2-year-old child attends their GP practice with faltering growth, abdominal pain and variable bowel habit. Their mother thinks her child may have coeliac disease, as she suffers from the condition herself.

A. In what common foodstuffs is gluten found?

Wheat, barley and rye.

B. Describe the pathophysiology of coeliac disease.

Coeliac disease is an autoimmune condition characterized by T-cell activation against the bowel wall. This leads to inflammation and subsequent malabsorption of nutrients from the diet.

C. What is the classical habitus of an infant affected by the disease?

Wasting of the buttocks and distension of the abdomen.

D. What investigations are required?

The presence of anti-tTG (tissue transglutaminase) and anti-endomysial antibody (EMA) is diagnostic. Where these are equivocal, a jejunal biopsy is required.

E. How is coeliac disease treated?

A gluten-free diet is required.

19. A 10-month-old infant attends the Emergency Department with severe abdominal pain, bilious vomiting and 'redcurrant jelly' stools. You suspect intussusception.

A. Where are the most common sites for intussusception in the bowel?

Ileocaecal is most common, but ileo–ileal and colocolic types also exist.

B. Describe the pathological sequence of events that underpin this condition.

There is invagination of the proximal bowel segment into the distal part, causing venous and lymphatic obstruction. This causes narrowing of the bowel (thus leading to symptoms of bowel obstruction, e.g. colicky abdominal pain and vomiting) and further oedema in the bowel wall. Eventually, this will lead to arterial insufficiency and consequent infarction.

C. Describe the classical abdominal examination findings.

A 'sausage-shaped' mass in the RUQ and an empty RLQ (Dance sign)

D. Which imaging types can be used for diagnosis?

An ultrasound shows a 'target' sign. An AXR will demonstrate bowel obstruction and the leading edge of the intussusception may be seen as a 'rounded' shadow on the radiography. The gold standard test is a contrast enema.

E. In the absence of peritonism, how are these children treated?

A barium (or air) enema is the definitive treatment. The bowel should be decompressed by siting a nasogastric tube.

20. A 3-year-old child, who has been suffering with an upper respiratory tract infection, attends the Emergency Department following a first seizure. You think they have likely had a febrile convulsion.

A. What are some of the most pertinent negatives that you must ensure you address in the history and examination?

It is essential that you ensure you specifically ask about sinister causes, such as meningitis, encephalitis, sepsis and non-accidental injury. They must have had a preceding febrile illness and there should be no suggestion of any sinister causes on examination. Their symptoms must have fully resolved within one hour.

B. How is the diagnosis made?

The diagnosis is clinical. Where there is clinical uncertainty, the child should be admitted for observation ± investigation.

C. What is the management of a febrile convulsion?

The management is to treat the underlying diagnosis and control fever with antipyretics.

D. What advice should parents be given about managing seizures in the community?

It is important that the child is not restrained, in order to prevent accidental fracture. Move any hard objects out of the way; you can place protective material (e.g. a pillow). Ensure there is nothing in the child's mouth. After the seizure, place the child in the recovery position.

Single Best Answer Questions

1. An 18-month-old child attends the Emergency Department with a short history of being non-specifically unwell. They have lost 10% of their body weight. Which of the following is NOT a feature of interstitial fluid depletion?

A. Sunken anterior fontanelle

B. Reduced skin turgor

C. Reduced urine output

D. Sunken eyes

E. Dry mucous membranes

Reduced urine output is a feature of intravascular fluid depletion and represents a more significant volume depletion. Other features of intravascular depletion include altered GCS, tachycardia, tachypnoea, delayed capillary refill time and low BP.

2. Which of the following clinical signs is a feature of a narrowed upper airway?

A. Wheeze

B. Stridor

C. Crackles

D. Tracheal tug

E. Nasal flaring

Stridor is a harsh breathing sound produced by breathing against a partially occluded upper airway. Wheeze and crackles are produced in the lower respiratory tract. Nasal flaring and tracheal tug are non-specific signs of respiratory distress.

3. A neonate becomes rapidly cyanosed within the first few days of life. They have a loud ejection systolic murmur that is easily heard with the stethoscope. There is no thrill present. What grade is this murmur on the Levine scale?

A. Grade 1

B. Grade 2

C. Grade 3

D. Grade 4

E. Grade 5

This murmur is Grade 3 – a loud murmur, but no thrill present. Grade 4 is characterized by a loud murmur with a thrill present. Grade 6, the loudest type, has a thrill and is heard with the stethoscope placed near, but not on, the chest wall.

4. You are assessing a 15-year-old boy with diplegic cerebral palsy. On examination, you note that his plantar reflexes are upgoing and his patellar tendon reflexes are exaggerated. What spinal nerve roots does striking the patellar tendon assess?

A. L2/L3

B. L3/L4

C. L4/L5

D. L5/S1

E. S1/S2

Striking the patellar tendon assesses the L3/L4 nerve roots. Striking the calcaneal tendon assesses the S1/S2 nerve roots.

5. You are assessing the growth of a 10-week-old child who was born at 34 weeks' gestation. On the growth chart, what age should the child be plotted as?

A. 4 weeks

B. 5 weeks

C. 7 weeks

D. 9 weeks

E. 10 weeks

Children born prematurely (<37 weeks of gestation) should have their age corrected on the growth chart until they are 2 years old. In this example, the child was born 6 weeks early (as the normal gestation is 40 weeks), so they should be plotted as 4 weeks on the growth chart (i.e. their actual age (10 weeks) minus how many weeks early they were (6 weeks)).

6. You are working in a General Practice and are asked to review a child who the mother thinks has delayed developmental milestones. Regarding these, which of the following is true?

A. The sequence of milestone accrual is variable

B. If not accrued within a defined upper limit, this is sometimes abnormal

C. A 'red flag' is when a trait is gained earlier than expected

D. The time at which a trait is acquired is variable, but always follows a predictable sequence

E. There are 5 groups of skills assessed in the developmental milestones

Milestones are accrued in a predictable, consistent sequence; however, the timing of their acquisition is variable, but within defined upper limits. Failure to have acquired a skill within this upper limit is a 'red flag' and is always abnormal. There are four groups of skills assessed: gross motor, fine motor, speech & language, social skills.

7. Regarding the foetal circulation, which of the following shunts bypasses the foetal liver?

A. Ductus arteriosus

B. Ductus venosus

C. Foramen ovale

D. Placenta

E. Umbilical artery

The ductus venosus bypasses the foetal liver, shunting blood directly from the left umbilical vein into the inferior vena cava (IVC). In adult life, it is found as a remnant called the ligamentum venosum, lying within a fissure on the posterior aspect of the liver.

8. Regarding the heel prick test (Guthrie card), performed on day 5 of life, which of the following conditions is NOT screened for?

A. Congenital adrenal hyperplasia

B. Sickle cell disease

C. Phenylketonuria

D. Cystic fibrosis

E. Congenital hypothyroidism

Congenital adrenal hyperplasia is not screened for in the UK. In addition to the remaining above answers, a number of metabolic defects are included – MCADD, MSUD, IVA, GA1 and HCU.

9. You are asked to review a child with hypothermia. The below are all ways by which babies lose heat, with the EXCEPTION of:

A. Convection

B. Evaporation

C. Conduction

D. Condensation

E. Radiation

Heat is lost by convection (heat lost to air currents), evaporation (when water evaporates from skin/breath), conduction (heat transferred to a solid surface with which they are in contact) and radiation (heat lost to surrounding surfaces by electromagnetic waves).

10. A 1-week-old premature neonate has become acutely unwell, has started vomiting and has a distended abdomen. They are febrile on examination. Considering the likely diagnosis, what will the AXR show?

A. Pneumatosis intestinalis

B. Small bowel dilatation

C. A 'target sign'

D. A 'double bubble'

E. 'Soap bubbles'

The most likely diagnosis is meconium ileus, which characteristically has pneumatosis intestinalis on the AXR. The 'double bubble' is a sign of duodenal atresia, and represents air in the stomach and the duodenum (proximal to the part that has failed to canalize). The 'soap bubble' appearance is a radiographic characteristic of meconium ileus, representing air between the meconium pellets.

11. An 18-month-old boy presents with acute frank rectal bleeding, but an otherwise unremarkable history. The examination is normal. You suspect that this is a haemorrhagic Meckel diverticulum. Regarding the embryological origin of this, a Meckel diverticulum is a remnant of the:

A. Allantois

B. Vitellointestinal duct

C. Yolk sac

D. Cloaca

E. Mesonephric duct

A Meckel diverticulum is a remnant of the vitellointestinal duct, which connects the midgut with the yolk sac in embryological development. It most commonly presents with per rectum bleeding, due to aberrant secretion of gastric acid.

12. Regarding the features of tetralogy of Fallot, which of the following is NOT present?

A. Ventricular septal defect

B. Overriding aorta

C. Pulmonary valve stenosis

D. Right ventricular hypertrophy

E. Atrioventricular septal defect

An atrioventricular septal defect is not found in TOF; the remaining four form the tetrad.

13. A newborn baby is noted to be cyanosed within hours of birth. Which of the following is the least likely cause?

A. Tetralogy of Fallot

B. Truncus arteriosus

C. Ventricular septal defect

D. Hypoplastic left heart syndrome

E. Transposition of the great vessels

An atrioventricular septal defect is least likely to cause cyanosis, as the shunt is predominantly left-to-right, meaning that blood is oxygenated as it passes through the lungs. The remaining cardiac anomalies are all cyanotic.

14. A 17-year-old boy, who has recently moved to the UK from India, presents to the Emergency Department with breathlessness and a new cyanosis. His fingers are clubbed. The JVP is elevated. He has hepatomegaly and a parasternal heave. There is a loud murmur on auscultation. What is the likely diagnosis?

A. Eisenmenger syndrome

B. Brugada syndrome

C. Takotsubo cardiomyopathy

D. Stiff heart syndrome

E. Williams syndrome

This presentation most likely represents Eisenmenger, as a consequence of a previously undiagnosed cardiac defect causing a left-to-right shunt. In this condition, over time the left-to-right shunt causes pulmonary hypertension and, ultimately, right ventricular hypertrophy. Eventually, the pressure in the right ventricle exceeds that in the left ventricle and leads to reversal of the shunt (now becoming right-to-left), so leading to cyanosis.

15. A 3-year-old infant has had four UTIs in the last few years and is referred for further imaging. This shows a duplex kidney. What is the underlying anatomical defect in this condition?

A. The upper and lower poles of left and right kidneys are fused

B. There is a second, smaller kidney on the same side of the body

C. The lower poles of the kidneys are fused in the midline

D. A kidney with two separate pelvicalyceal systems

E. The kidney has two upper and two lower poles

A duplex kidney has two pelvicalyceal systems; this may unite prior to entering the bladder or enter separately.

A kidney in which the upper and lower poles are fused with the contralateral side is termed a 'pancake kidney'. A kidney where the inferior poles fuse in the midline is a 'horseshoe kidney'. A second, smaller kidney on the same side is a 'supernumerary kidney'.

16. Regarding the sequelae of nephrotic syndrome, the following are all present EXCEPT:

A. Hypertension

B. Hypercoagulability

C. Higher risk of infection

D. Hypovolaemia

E. Hypercholesterolaemia

Hypertension is a feature of nephritic, rather than nephrotic, syndrome. Proteinuria is an important feature of nephrotic syndrome as it:

(i) causes loss of complement and immunoglobulins, so increasing risk of infection
(ii) triggers lipid synthesis in the liver, and
(iii) contributes to hypovolaemia that, in turn, increases hypercoagulability.

17. An 11-year-old boy is seen in his GP practice with enuresis, despite having been continent of urine for many years previously. The following are all examples of secondary causes EXCEPT:

A. Diabetes mellitus

B. Posterior urethral valve

C. UTI

D. Emotional distress

E. Constipation

Secondary enuresis is defined as a child who has previously been continent of urine for >6 months. Children with a posterior urethral valve will not usually have achieved continence, so this is a primary cause.

18. Which of the following neurological motor terms is INCORRECTLY paired with its description:

A. Atonic: sudden loss of muscle tone

B. Clonic: sustained rhythmic jerking of the limbs

C. Grand mal: initial stiffening, followed by rhythmic jerking

D. Tonic: stiffness of the limbs

E. Myoclonic: repetitive, purposeless actions

Myoclonic is a term used for a brief muscle jerk. A repetitive, purposeless action is termed an 'automatism'.

19. An 18-month-old child is brought into the Emergency Department following a 'seizure'. It is reported that the child tripped whilst running, which was followed by an episode of cyanosis, loss of consciousness and jerking of the limbs. This quickly resolved. The examination is unremarkable. Based on this story, what is the most likely diagnosis?

A. Psychogenic non-epileptic seizure

B. Epilepsy

C. Breath-holding spell

D. Head injury

E. Tic disorder

This story is characteristic of a breath-holding spell. The child typically has very minor trauma or emotional distress, which is followed by a period of sustained breath-holding, leading to cyanosis and loss of consciousness. Symptoms rapidly resolve and the examination is normal.

20. A 14-year-old female presents to her GP practice with severe, unilateral headaches. These are described as 'stabbing' and are felt retro-orbitally. The GP thinks that the diagnosis is likely to be cluster headaches, so asks for associated symptoms. Which of the following is NOT a feature of cluster headaches?

A. Ipsilateral red eye

B. Nasal congestion

C. Vomiting

D. Eye watering (epiphora)

E. Facial sweating

Vomiting is not commonly seen in cluster headaches, so may be helpful in helping to differentiate between these and migraines. They are often accompanied by autonomic symptoms, such as redness of the eye, facial sweating, eye watering and nasal congestion.

21. A 13-year-old female returns to her GP with frequent, recurrent migraines. These are severe and interfering with her schooling. Which of the following medicines is most suitable as prophylaxis?

A. Ibuprofen

B. Verapamil

C. Sumatriptan

D. Propranolol

E. Paracetamol

Propranolol and topiramate are recommended by NICE for the prophylaxis of migraine. Sumatriptan is an acute treatment for migraine. Verapamil is recommended for the prophylaxis of cluster headaches.

22. A 7-year-old girl presents to her GP practice with a painless, unilateral reduction in her visual acuity. There is subtle proptosis and a relative afferent pupillary defect on examination. She is referred for an MRI that demonstrates an optic nerve glioma. Her mother has neurofibromatosis-1 (NF1). Regarding the diagnostic features, which of the following is NOT a feature?

A. Café au lait spots

B. Vestibular schwannoma

C. Lisch nodules

D. Axillary/inguinal freckling

E. Anterolateral tibial bowing

Vestibular schwannoma (a benign tumour affecting the vestibulocochlear nerve) is characterizing of NF2 (rather than NF1), a variant of neurofibromatosis that predominantly affects the CNS. All of the other features are characteristic of NF1.

23. Regarding cerebral palsy (CP), which of the following features should alert you to an ALTERNATIVE diagnosis?

A. Loss of attained abilities

B. Motor abnormalities

C. Delayed developmental milestones

D. Persistence of primitive reflexes

E. Learning disability

It is important to recognize that the loss of an ability (regression) is not a feature of CP and should arouse suspicion for an alternative diagnosis. Other features not consistent with CP are the absence of risk factors, development of focal neurology (rather than a static neurological deficit) and a family history of a progressive neurological disorder.

24. A 4-year-old child attends the Emergency Department with a limp, complaining bitterly of left hip pain. They have recently had a sore throat. Their observations are unremarkable. There is a moderately reduced range of motion. The WCC is 9 and CRP is 18. What is the most likely diagnosis?

A. Transient synovitis

B. Juvenile idiopathic arthritis

C. Septic arthritis

D. Malignancy

E. Perthes' disease

This is a typical history of transient synovitis following a probable URTI and with clinical features that mimic, but are less severe than, septic arthritis. The CRP is <20 which is a useful distinguishing biochemical marker. The management is symptomatic, with robust advice about the features of septic arthritis and instruction on when to return to hospital if these develop.

25. Regarding the causes of septic arthritis, which organism is most common in children with sickle cell disease?

A. *Haemophilus influenzae B*

B. *Staphylococcus aureus*

C. *Salmonella* spp.

D. *Neisseria gonorrhoeae*

E. *Streptococcus pneumoniae*

In children with sickle cell disease, *Salmonella* spp. are the most likely causal organisms. *N. gonorrhoeae* tends to affect adolescents, and the remaining organisms affect young children.

26. A 10-year-old female presents with weakness and non-specific joint pains. On examination, you notice that she has a purple discoloration of the eyelids and thickened papules over the extensor surfaces of her hands. Given the likely diagnosis, which of the following blood tests is most useful?

A. Anti-dsDNA

B. RhF

C. Anti-Sm

D. ANA

E. CK

The likely diagnosis is juvenile dermatomyositis. Creatine kinase (CK) is the most useful differentiator of the above tests, although ANA is also positive in ~50% of these patients. Anti-dsDNA and anti-Sm are found in systemic lupus erythematosus (SLE).

27. Regarding achondroplasia, which of the following bones would be unaffected by the underlying pathophysiology?

A. Femur

B. Clavicle

C. Humerus

D. Vertebrae

E. Tibia

Achondroplasia interferes with endochondrial ossification, the process by which most long bones are formed. Of these choices, the clavicle is the only bone not formed by this process; instead, it undergoes membranous ossification, as do the mandible and the bones of the skull.

28. A 14-year-old with Crohn's disease attends their GP practice with increasing fatigability and irritability. Their exercise tolerance has reduced recently. Blood tests demonstrate iron-deficiency anaemia. Which of the following is NOT consistent with this diagnosis?

A. Low MCV (mean corpuscular volume)

B. Low ferritin

C. Low transferrin saturation

D. Low Hb

E. Low TIBC (total iron-binding capacity)

The TIBC is elevated in iron-deficiency anaemia, reflecting the fact that transferrin, the protein which transports iron, is poorly saturated due to iron deficiency (i.e. the transferrin saturation is low); therefore, the capacity to carry iron is high. Ferritin, an intracellular protein that stores iron, is low.

29. A 16-year-old female with sickle cell disease presents to the Emergency Department with chest pain and breathlessness. She is febrile and has chest crepitations on examination. Which of the following investigations is of lowest clinical utility acutely?

A. Reticulocyte count

B. Arterial blood gas

C. Blood film

D. CXR

E. CRP

A blood film is not of high clinical utility acutely, as it does not help with diagnosis or influence management. The remaining investigations should all be performed acutely.

30. An 8-month-old infant is brought to the Emergency Department with excessive bleeding after a small cut following a fall. They have had a number of similar episodes prior to this. Regarding the features of haemophilia, which of the following is true?

A. Haemophilia A affects Factor VII and Haemophilia B affects Factor VIII

B. Spontaneous haemarthroses are a prominent feature of even mild disease

C. The extrinsic coagulation pathway is affected

D. The prothrombin time (PT) is normal, but the activated partial thromboplastin time (APTT) time is prolonged

E. Recombinant clotting factors are given, to keep levels near 100% in affected children

Haemophilia A affects Factor VIII and Haemophilia B affects Factor IX, both components of the intrinsic coagulation pathway. The intrinsic pathway is measured using the APTT and, consequently, the APTT is prolonged; whereas the PT, a measure of the extrinsic pathway, is unaffected.

Haemarthroses are seen in moderate–severe disease; mild disease tends to present with excessive bleeding after minor trauma.

It is not necessary to keep clotting factor level at 100%, with the exception of during the perioperative period.

31. You are asked to review a 1-week-old neonate with an erythematous, blanching, macular rash on their buttocks. They are otherwise well with normal observations. What is the most likely cause?

A. Erythema toxicum neonatorum (ETN)

B. Scarlet fever

C. Capillary malformation

D. Café au lait spot

E. Infantile haemangioma

The most likely diagnosis is ETN – the rash is blanching, macular, and the child is clinically completely well otherwise. It affects approximately one-third of all neonates.

32. Regarding chickenpox (varicella zoster virus), which of the following is INCORRECT?

A. The child may return to school once the lesions have scabbed over (~day 5)

B. The diagnosis is clinical

C. Aspirin may be given for fever, if it is distressing the child

D. The child should stay away from pregnant women

E. Encephalitis is a rare complication of chickenpox

Aspirin should not be given to any child under 16 due to its association with Reye syndrome, an acute, non-inflammatory encephalopathy with fatty degeneration of the liver.

33. A 5-year-old child attends their GP practice with a rash on their face and arms. They have been feeling non-specifically unwell for the last week. The rash affects the cheeks, but spares the nose, philtrum, mouth and eyes. They have a lacy rash on the extensor surfaces of their arms. What is the likely diagnosis?

A. Rubella

B. Fifth disease

C. Hand, foot and mouth disease

D. Measles

E. Mumps

This is a characteristic description of fifth disease (aka 'slapped cheek'), which has a viral prodrome for ~7–10 days, followed by the typical rash as described above.

34. A 4-year-old child presents to the Emergency Department with a 4-day history of irritability and a fever. On examination, they have a widespread rash, cracked lips with a red, inflamed tongue and red, sore eyes. The skin on their hands is peeling. Considering the most likely diagnosis, what is the most important acute complication of this disease?

A. Cardiac arrhythmia

B. Encephalitis

C. Avascular necrosis of the femoral head

D. Pulmonary infarct

E. Coronary artery aneurysm

The most likely diagnosis is Kawasaki disease, a small/medium-sized vessel vasculitis. The most important complication is coronary artery aneurysm, affecting ~25% of patients, which can cause acute myocardial ischaemia. Myocarditis and pericarditis are also sometimes seen, which may cause cardiac arrhythmia. The remaining options are not features of this disease.

35. An 11-year-old boy is being treated with methylphenidate for attention deficit hyperactivity disorder (ADHD). Which of the following is true regarding its mechanism of action?

A. It inhibits the reuptake of dopamine

B. It promotes the reuptake of both dopamine and noradrenaline

C. It promotes the reuptake of dopamine

D. It inhibits the reuptake of both dopamine and noradrenaline

E. It inhibits the reuptake of serotonin

Methylphenidate blocks dopamine (DA) and noradrenaline (NA) transporters on the synaptic membrane, thus preventing their reuptake. This leads to increased DA and NA in neuronal synapses and therefore increases the neurotransmission of the substances.

36. Regarding common chromosomal abnormalities, which of the following is due to a deletion of genetic material?

A. DiGeorge syndrome

B. Down syndrome

C. Patau syndrome

D. Klinefelter syndrome

E. Edwards syndrome

DiGeorge syndrome is due to a microdeletion of 22q11 on chromosome 22. Its features can be remembered using the acronym 'CATCH-22':

- **C**ardiac anomalies
- **A**bnormal facies
- **T**hymic hypoplasia
- **C**left palate
- **H**ypocalcaemia
- **22**q11.2 deletion

The remaining syndromes are all disorders with excess genetic material.

37. An 11-year-old girl is referred to the allergy clinic, having been recently treated in the Emergency Department for an anaphylactic reaction. The child does not know what the cause of this was, so the decision is taken to perform a skin prick test. Which class of immunoglobulin-mediated response does this test check for?

A. IgA

B. IgD

C. IgE

D. IgG

E. IgM

The skin prick test assesses responses mediated by IgE, the immunoglobulin class that mediates allergy by triggering histamine release from mast cells and basophils.

38. A 7-year-old child attends the Emergency Department in acute respiratory distress. They have a known diagnosis of asthma. Which of the following clinical findings is suggestive of a life-threatening exacerbation?

A. Peak expiratory flow rate (PEFR) 33–50% of predicted

B. Exhaustion

C. Inability to complete sentences in a single breath

D. Tachycardia

E. Oxygen saturations <92%

Exhaustion is a feature of a life-threatening exacerbation of asthma; other clinical features include PEFR <33% predicted, cyanosis, hypotension, confusion, coma and a silent chest on auscultation. It is essential that you are able to assess these, as the presence of one is indicative of life-threatening disease.

All the other stated findings are features of an acute–severe exacerbation.

39. Regarding infertility in male patients with cystic fibrosis, which of the following is true?

A. The contents of the ejaculatory duct are thickened, meaning that they cannot pass down the urethra during ejaculation

B. There is congenital absence of the vas deferens

C. There is impaired motility of the spermatozoa, meaning that they cannot move adequately through the female reproductive tract

D. Venous stasis leads to testicular infarction in early life, resulting in impaired spermatogenesis

E. There is mucus plugging in the reproductive tract, leading to impaired delivery of spermatozoa to the ejaculate

95% of males with cystic fibrosis are infertile due to congenital bilateral absence of the vas deferens (CBAVD).

40. A 15-year-old girl is referred to the first seizure clinic. She has recently moved schools and her parents are in the process of a divorce. Her mother shows you a video of the seizure; what clinical feature is LEAST SUGGESTIVE of a psychogenic non-epileptic seizure (PNES)?

A. Gradual onset

B. Recall of the event

C. A stressor which reliably causes seizure onset

D. Synchronous jerking of the limbs

E. Shaking the head from side to side

Synchronous jerking is suggestive of a true seizure, whereas asynchronous, flailing movements are more often seen in PNES. The remaining clinical features are all suggestive of PNES.