Chapter

7

# Preoperative Assessment for Paediatric Anaesthesia

## Considerations for Common Medical Conditions and Risk

Pamela Cupples

## Introduction

Preoperative assessment is an essential part of paediatric anaesthesia. Whilst every child will be seen on the day of their procedure by the responsible anaesthetic team, there is broad acceptance that anaesthetic preoperative assessment (APOA), in advance of the admission, is the optimal standard of care.

The purpose of APOA is to:

- Ensure that the child is medically optimised in advance of their admission
- Identify specific anaesthetic issues, such as difficult airway, previous anaesthetic complications, genetic conditions such as malignant hyperpyrexia or suxamethonium apnoea
- Liaise with the relevant medical teams for those with complex comorbidities to ensure that their expertise and knowledge of the patient is incorporated into the plan for admission, such as endocrine, metabolism and haematology
- Identify high-risk patients, for whom a perioperative multidisciplinary team meeting (MDT) should be convened to consider whether surgery is appropriate and if so to agree on the perioperative plan, including ceilings of care
- Order appropriate investigations – this will be guided by the child's medication, the underlying comorbidities and the type of surgery the child is having
- Prepare the child and the family and explain the specifics of the anaesthetic process, including:
  - Discussion around induction techniques
  - Options for premedication if required, and support for the induction process such as distraction techniques and play therapists
  - Pain management and the options available – regional and opioid-based techniques
  - The recovery process
- Discuss side effects and risks of anaesthesia
- Disseminate information leaflets to support discussion and understanding
- Allow the child and family time to ask questions and explain to the anaesthetic team what is important to them
- Provide contact details should they wish to discuss any part of the anaesthetic again in advance of the admission

APOA services have been shown to:

- Reduce cancellations on the day of surgery and improve theatre efficiency
- Reduce complication rates and mortality
- Improve patient experience and reduce perioperative anxiety

The delivery of an APOA service is reliant on a hybrid model of medical and nursing assessments. Specialist APOA nurses are effective at preoperative screening and are an integral part of the team. Anaesthetic consultants must always support nurse-led clinics. The Association of Paediatric Anaesthetists of Great Britain and Ireland (APAGBI) guidance document on the running of preoperative assessment clinics helps guide the development and running of paediatric APOA services.

Paediatric APOA differs from adults in several key areas:

- The interview will be with both the child and the parent, as young children will be unable to give a detailed history.
- Older children and adolescents should be encouraged to give a history, but this will need

to be supported by a joint discussion with the parent/carer.
- Gillick competent children should ideally make consent decisions for themselves, supported by their parent/carer.
- There is significant anxiety and stress for the family in dealing with children for surgery. You are not only looking after the child but the parent and carer too.
- Comorbidities are more likely due to congenital disorders, prematurity or acute illness.

# APOA Process

## Clinical Evaluation

A detailed review of the medical notes is essential to identify comorbidities, their physiological implications and the indication for surgery. If the child has had previous anaesthetics, these charts should be reviewed if possible, noting the level of anxiety at induction, the need for premedication and induction technique. An assessment of the airway and how it has been managed should be documented, together with any intraoperative complications. The child's current medication and history of allergies should also be reviewed and documented.

The review of the notes should then be combined with an interview to confirm the medical history and allow a physical assessment of the child.

## Interview

The interview process helps to develop a rapport with the child and parent and to explain the anaesthetist's role during the child's admission. A systematic approach to assessment is essential to ensure that nothing is overlooked. A detailed antenatal and neonatal history is important when infants present for surgery. Time spent in NICU, the need for respiratory support and any ongoing respiratory requirements, such as oxygen therapy or non-invasive ventilation, should be documented.

The child's physiological status should be assessed and documented along with objective measurements such as echocardiography (ECHO) and lung function testing where indicated.

If this is the patient's first general anaesthetic (GA), asking about any family history of problems under GA will help to identify those at risk of malignant hyperpyrexia and suxamethonium apnoea. If the child has had previous anaesthetics, it is an opportunity to understand how successful the induction was and what, if anything, can be improved upon, such as distraction techniques, play therapy or premedication.

Medication history should be captured as part of the assessment, including recently stopped drugs that may have anaesthetic relevance, such as steroid therapy or herbal medicines. This should include any allergies the patient may have and their significance, including whether the child carries an adrenaline auto-injector, such as EpiPen®.

Recent laboratory tests should be documented along with the child's vaccination history. Adolescents should also be questioned about smoking and drinking habits and the possibility of pregnancy. Due to the sensitive nature of this questioning, it may be prudent to do this confidentially.

A key part of the interview is a discussion about the anaesthetic process itself, including induction techniques, invasive monitoring if required, approaches to analgesia, need for intraoperative blood transfusion, as well as recovery and postoperative care. This is particularly important for children having major surgery who may require an elective admission to critical care.

## Side Effects and Risks

No assessment is complete without a discussion about side effects and risks of general anaesthesia. Information leaflets can provide a useful adjunct to these discussions; these may outline the admission process, the general anaesthetic, analgesic techniques and side effects and risks. These discussions need to occur well in advance of the admission to allow the family and the patient time to consider the information, ask questions and agree the perioperative plan.

Common side effects such as postoperative nausea and vomiting (PONV), sore throat, behavioural changes and emergence delirium need to be discussed with the family as part of the preoperative assessment. Minor cuts to the lips and tongue and the potential of dental damage may also be mentioned.

New General Medical Council (GMC) guidance has highlighted that communication

with families should be clear and consistent even when the risks of serious harm and death are extremely rare.

There are no validated clinical risk scoring tools that are used in paediatric anaesthesia. Data rely on published audits from different paediatric centres to inform the speciality about the incidence of morbidity and mortality.

The National Audit Projects (NAPs) have helped to inform our discussions around anaphylaxis and accidental awareness. The risk of perioperative anaphylaxis has been quantified as 1:10,000. The risk of accidental awareness during general anaesthesia is quoted as 1:60,000 (NAP 5); whilst rare, when it does occur it is not usually associated with pain or distress.

In due course, the completion of the UK NAP 7 audit into perioperative cardiac arrest will be completed and will inform the specialty of the incidence of cardiac arrest during GA.

The risk of a child dying during a general anaesthetic is rare (1:10,000) and more commonly seen in neonates and those with significant comorbidity, particularly congenital heart disease and pulmonary hypertension. In the fit and healthy child presenting for elective surgery, the risk of dying under GA is quoted as being between 1:100,000 and 1:1,000,000.

## Jehovah's Witnesses (JW)

There are approximately 8.5 million Jehovah's Witnesses worldwide, and 150,000 live in the United Kingdom and Ireland. It is an essential part of the anaesthetic assessment for any child of a JW to have detailed discussions about the surgery and the risks of requiring a blood or blood product transfusion.

The discussions should take the form of an MDT. It should include the responsible surgeon, anaesthetist and senior haematologist with input from the hospital liaison committee and hospital legal team if required. The purpose of the MDT is to:

- Clarify the reason for the proposed surgery and what blood or blood products may be required
- Discuss alternatives to blood transfusion, including cell salvage
- Explain what the risks of blood transfusion are and the consequences of withholding transfusion in the face of life-threatening haemorrhage
- Consult with the family to understand what they will consent to

In the elective setting, if the parents refuse to give consent and it is deemed unsafe to operate without the option to give blood, then a second opinion can be sought. The family may be happy to give consent with some caveats. As long as the team feels it can safely work within these caveats, the case can proceed. It is essential that the family feel that their beliefs are being respected, and that the team will do everything to avoid transfusion where possible. The discussion and the agreed plan need to be documented clearly in the notes.

The team must always work in the best interests of the child, and if blood transfusion is deemed to be lifesaving, it can be done even if parents refuse. Legislation varies between countries, but in England and Wales, a special issue order under Section 8 of The Children's Act 1989 can be obtained from the High Court. They can issue an order providing legal permission for treatment in the face of parental refusal. In an urgent or dynamic situation, blood should be given to minimise the threat to life or to prevent lasting disability whilst such an application is made. Seeking the opinion and agreement of another senior colleague and documenting this in the notes is advised. The hospital's legal team can be helpful in this setting.

## Vaccinations

Immunisation is not a contraindication to GA in healthy children scheduled for elective surgery. Historically, there have been concerns around anaesthesia influencing the efficacy and safety of vaccines or the inflammatory response to vaccines being mistaken for an acute infection. The guidance for non-immunocompromised children is:

- A minimum delay of 48 hours is advised for inactivated vaccines.
- No delay is required for children having a live attenuated vaccine, but there is a small risk of the child developing a fever in the weeks following the vaccination, which may coincide with the admission. This will require assessment at the time to decide if this is vaccine related or due to another cause.
- Children can be safely vaccinated following a GA, providing they have made a full recovery.

## Physical Examination

All children presenting for GA should have the following observations performed:

- Heart rate, oxygen saturation, respiratory rate, temperature
- Blood pressure in children over three and those under three with known hypertensive comorbidity
- Height and weight

In an emergency setting, obtaining an accurate weight may be difficult, and clinicians will need to use a formula to estimate the child's weight. Examples include:

- The Advanced Paediatric Life Support (APLS) formula:
  weight (kg) = 2 × (age + 4)
- Luscombe formula:
  weight (kg) = (3 × age) + 7
- Modified Australian BG method:
  weight (kg) = (2 × age) + 10

All estimates may be inaccurate, but the APLS formula is now felt to be a significant underestimate, and there is a move towards using the Luscombe or the Modified Australian BG method. An accurate weight should be obtained as soon as clinically appropriate.

The respiratory and cardiovascular systems should be examined and the airway assessed according to the Mallampati classification if possible. Any restriction of neck movement should also be assessed. Examination will depend on the age and cooperation of the child. Cormack and Lehane grade of laryngoscopy may be obtained from previous anaesthetic records. The state of the child's teeth should be noted, including the position of any loose teeth or braces. Known or anticipated difficult venous access should be noted.

## Investigations

Preoperative investigations will depend on the child's comorbidities, current medication, and the surgery they are having. Routine investigations, such as full blood count or urea and electrolytes, are not required in the otherwise healthy child presenting for minor routine surgery. Neonates less than 60 weeks postconceptual age are at increased risk of apnoea following GA, and this risk is increased in those neonates who are also anaemic. In this setting, a full blood count to assess the haemoglobin level is prudent.

Whilst testing for sickle cell anaemia is routine for babies born in the United Kingdom, it is essential to check the status of children from racial groups at increased risk of sickle cell anaemia. The sickle cell solubility test (Sickledex) can rapidly identify the presence of haemoglobin S (HbS) in a blood sample, but it does not distinguish between sickle cell trait (SCT) and disease (SCD). Furthermore, this test is not reliable when HbS levels are below 15–20%, so it is not suited for newborn screening. In the case of a positive test, more detailed haemoglobinopathy testing is required.

Chest X-rays, sleep studies, spirometry and echocardiograms should be ordered based on the child's associated comorbidities.

## Heart Murmurs

Incidental heart murmurs are commonly picked up during a preoperative assessment. The challenge is differentiating innocent murmurs from those that are deemed to be pathological. Less than 1% of children with an incidental murmur have an underlying cardiac condition.

Most children with congenital heart disease (CHD) are diagnosed either antenatally during routine scanning or within the first three months of life. Some conditions can be asymptomatic, so all infants less than one year of age who have a murmur identified should be referred to a cardiologist.

A detailed history should be taken, and children with the following symptoms suggestive of an underlying cardiac condition warrant further investigation:

- Failure to thrive
- Recurrent respiratory tract infection
- Tachypnoea
- Cyanosis
- Pallor/mottling
- Sweating
- Difficulty with feeding
- Syncope or chest pain
- Reduced exercise tolerance
- Family history of sudden death

There should be a high index of suspicion of underlying cardiac disease in the following groups:

- Trisomy 21
- CHARGE syndrome (coloboma of the eye, heart defects, atresia of the choanae,

retardation of growth and/or development, genital and/or urinary abnormalities, ear abnormalities and deafness)
- VACTERL association (vertebral, anal, cardiac, tracheal-oesophageal fistula (TOF), renal, limb)
- Turner syndrome
- DiGeorge syndrome

The challenge is identifying those who require referral to cardiology before anaesthesia. Innocent murmurs are usually:

- Soft murmurs in early systole sometimes described as musical, blowing or vibratory
- Not associated with a pre-cordial thrill
- Variable with changes in posture
- Not associated with increased mortality or morbidity under general anaesthesia

Pathological murmurs are often:

- Pansystolic, late systole or in diastole
- Harsh quality
- Associated with thrills
- Present in clinically symptomatic patients

Children with innocent murmurs should have an electrocardiogram (ECG), looking for signs of ventricular hypertrophy. Children with critical aortic stenosis and hypertrophic obstructive cardiomyopathy (HOCM) can be clinically asymptomatic. ECG features of ventricular hypertrophy include:

- RVH: R-wave in V1 >1.75 mV (<5 yr) or R-wave in V1 >1.25 mV (5–12 yr) or upright T-wave
- LVH: R-wave in V5 or V6 > 4 mV

An asymptomatic child over one year old with an innocent murmur (i.e. venous hum or soft, early systolic with no thrill) and a normal ECG can proceed with surgery and be referred for investigation after the operation. If there is any doubt about the nature of a murmur, transthoracic echocardiography is a non-invasive and straightforward way to resolve the matter in most cases.

The algorithm shown in Figure 7.1 can help in the preoperative assessment of children presenting with incidental murmurs.

## Respiratory Tract Infections

Children will experience between six and eight upper respiratory tract infections (URTI) per year. Of these, 95% will be caused by one of the 200 viruses known to cause coryzal symptoms (cough, nasal congestion and discharge, sore throat and sneezing). Rhinovirus is responsible for 30–40% of these infections.

Due to airway hyperreactivity, there is an increased risk of perioperative complications for two to six weeks following an URTI. This is thought to be due to inflammatory mediators (histamine, interleukin, bradykinin), viral neuraminidases inhibiting the muscarinic M2 receptor and increased tachykinin levels due to viral inhibition of endopeptidases. These postulated mechanisms all promote airway hyperreactivity and bronchoconstriction.

Children who have or are recovering from a recent URTI are thus at increased risk of perioperative respiratory adverse events (PRAE) such as:

- Laryngospasm
- Bronchospasm
- Oxygen desaturation
- Coughing and breath holding
- Atelectasis and pneumonia

In the APOA setting, it is important to take a complete history and to consider other causes for coryzal symptoms such as:

- Croup
- Bronchiolitis
- Pneumonia
- Influenza
- Inhaled foreign body
- Gastro-oesophageal reflux disease (GORD)
- Allergic rhinitis

There are additional independent risk factors for adverse respiratory events in children who have a URTI:

- Ex-premature baby
- <1 year of age
- Asthma or atopy
- Living in a house with a smoker
- Copious secretions
- Airway surgery
- History of snoring

Despite these complications, the evidence would suggest that the majority can be managed safely without any long-term sequelae.

The decision to proceed with or postpone surgery in a child with an active or recent URTI

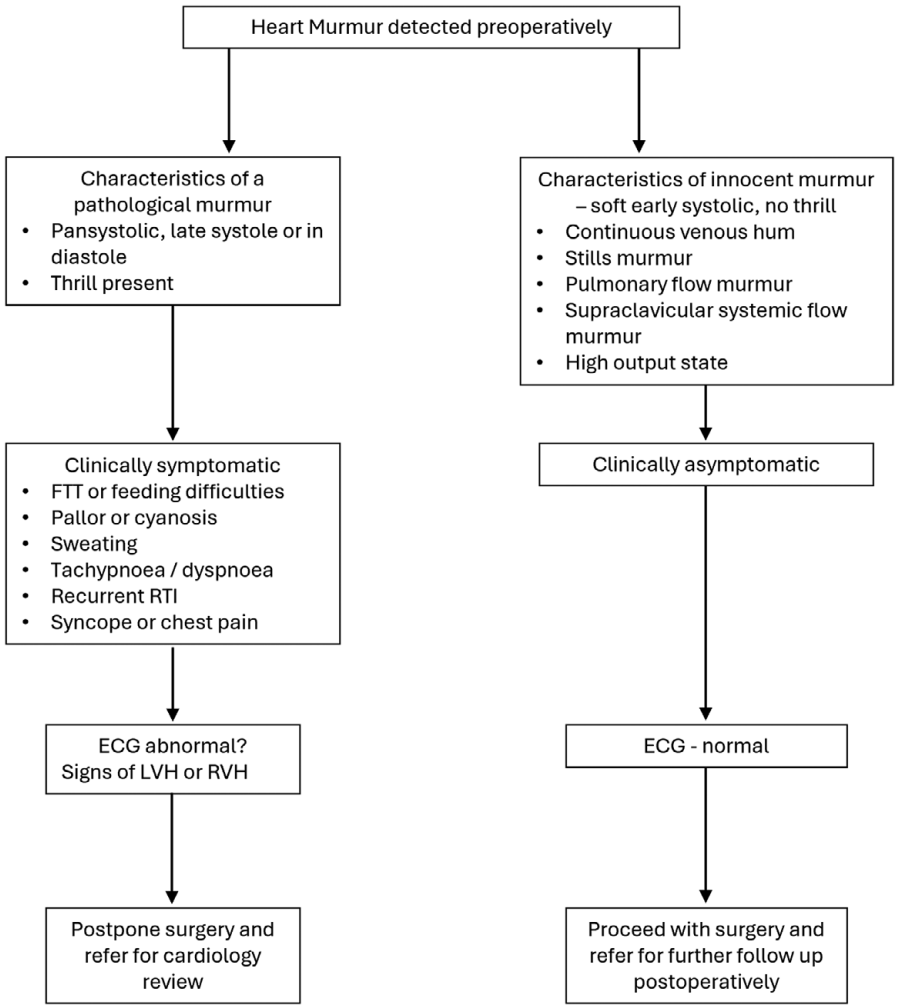


**Figure 7.1** Heart murmur algorithm.

requires an individualised approach, which takes into account:

- Detailed history and examination of the child
- Independent risk factors
- Comorbidities
- Type of surgery

Children who have minimal symptoms, such as isolated clear runny nose or dry cough, who are apyrexial and constitutionally well should proceed with the planned admission. Cancellation should be considered in children who show signs of a more severe URTI:

- A clinically unwell or lethargic child
- Pyrexia, with a temp >38°C
- Copious purulent secretions
- Associated risk factors, as detailed previously
- Complex comorbidities
- Major surgery

Those children with signs of a moderate or severe URTI (green nasal discharge, wet cough and pyrexia >38°C) should be postponed for two weeks and re-evaluated. Providing they have made a full recovery, they can then be re-scheduled.

In the case of croup, bronchiolitis and lower RTI, the procedure should be delayed for six weeks. If the surgery is deemed to be urgent, then a discussion with the surgical team is required to discuss clinical concerns and agree on what time frame would be in the child's best interest.

Additional investigations should only be considered on an individual basis. Routine blood

**Table 7.1** Premedication

| Drug | Dose | Onset | Additional comments |
|---|---|---|---|
| Midazolam | 0.5 mg $kg^{-1}$ oral<br>Max. dose 20 mg<br><br>0.3 mg $kg^{-1}$ buccal<br>Max. dose 10 mg | 30 min | Anxiolytic, amnesic<br>Bitter taste so may need to be disguised in flavoured juice<br>Occasional paradoxical excitation |
| Clonidine | 0.3 mg $kg^{-1}$ oral | 45–60 min | Colourless and tasteless<br>Caution in cardiovascular disease/ instability |
| Ketamine | 3 mg $kg^{-1}$ oral combined with oral midazolam | 10–15 min | |
| Dexmedetomidine | 2 μg $kg^{-1}$ (range 1–4 μg $kg^{-1}$) intranasal or buccal | 25 min | |
| Temazepam | 10 mg used in adolescent age group | 60 min | |

inflammatory markers and chest X-ray (CXR) may be normal in the face of a severe viral URTI.

## Premedication

Perioperative anxiety can be a challenge for the anaesthetic team and cause significant distress for the child and their parent. It is associated with adverse outcomes, including behavioural changes, emergence delirium and enuresis.

The management of perioperative anxiety starts with the APOA. The main concerns are explored and addressed through reassurance and explanation. Several specific strategies can be employed ahead of the admission:

- Information leaflets, videos and the use of hospital and operating theatre tours
- Play therapists
- Psychology in severe cases of perioperative anxiety

Despite a detailed discussion, anxiety on the day of the procedure needs to be anticipated and managed. Play therapists can be extremely helpful, as may:

- Distraction techniques – videos, books, toys, blowing bubbles
- Hypnosis and breathing techniques
- Keeping the anaesthetic room environment calm and quiet, altering the ambient light, playing music and limiting the number of health care professionals present

Even when the aforementioned techniques are used, some children will still require premedication (see Table 7.1). This is more common in:

- Children <4 years of age
- Children requiring multiple GAs
- Children who have had previous traumatic experience
- Children with learning difficulties

Planning for premedication should be discussed with the parent and the child. They should understand the range of responses with the particular drug prescribed and the anticipated time to optimal effect. Once administered, the child should be monitored closely by skilled staff, and resuscitation equipment must be close to hand, such as suction, oxygen, and self-inflating bag-valve-mask.

Sedative premedication may not be appropriate in some clinical conditions:

- Anticipated difficult airway
- Obstructive sleep apnoea (OSA)
- Risk of gastric aspiration
- Obtunded conscious level / raised intracranial pressure
- Acute systemic illness

Other drugs that may be required in the preoperative setting are:

- Topical local anaesthetic agents to facilitate cannulation:
  - 4% tetracaine gel, e.g. Ametop®, effective within 30–45 minutes

  - 2.5% prilocaine and 2.5% lidocaine cream, effective after one hour of application and giving up to 120 minutes of topical anaesthesia, e.g. EMLA® or Nulbia®
  - Ethyl chloride spray, e.g. Cryogesic®, which causes a reduction in skin sensation within a few seconds of application and lasts for approximately one minute
- Analgesics: ibuprofen and paracetamol can be given orally 30 minutes before GA
- Antimuscarinics may be given before airway surgery
- $H_2$ receptor antagonists, which are rarely given in the paediatric setting but can be used in those at risk of aspiration

## Fasting

In 2018 the APAGBI published a consensus statement, endorsed by many national paediatric anaesthetic societies, to support a change in the fasting guidance for clear fluids from two hours to one hour. The evidence supporting this change includes:

- A two-hour fast is usually significantly longer in practice, commonly six to seven hours and in some cases 15 hours.
- There is increased incidence of thirst, hunger and anxiety with longer fasting times.
- Patients exhibit better behaviour when allowed clear fluids up to one hour before surgery.
- There is less behavioural and metabolic derangement.
- Stomach volumes are low with clear fluids given up to one hour before anaesthesia.

The current guidance is:

- Clear fluids (max 3 ml $kg^{-1}$) can be given up to one hour prior to surgery
- Breast milk up to four hours
- Solid foods and formula milk up to six hours

It is important to remember that there is delayed gastric emptying in a trauma patient, which will be compounded by the use of opioids. In the trauma setting, the gastric contents and volume will be determined by the time between the last food taken before the injury took place. It is prudent to consider an induction technique that allows for the full stomach.

## Prevention of Perioperative Deep Venous Thrombosis (DVT)

The incidence of DVT in children is thought to be 0.05–0.14 per 10,000 of the paediatric population. In hospitalised children, the incidence is higher, quoted as 5–8 per 10,000 admissions. DVT is associated with increased morbidity and mortality, mainly due to the development of pulmonary embolism.

There are two peaks in the incidence of DVT; children <2 years of age, usually associated with central venous access, and adolescents over 13 years of age.

All children over 13 years or >40 kg presenting for surgery should have a DVT risk assessment. Eighty per cent of children developing a DVT will have at least one associated risk factor:

- Immobility
- Obesity
- Malignancy
- Infection
- Presence of central venous catheter: risk for femoral > subclavian > internal jugular lines
- Acquired and congenital thrombophilia
- Sickle cell anaemia
- Oestrogen-containing oral contraceptive pill (OCP)

## Prophylaxis

Immobility is a risk factor for DVT, so early mobilisation should be encouraged when appropriate. Dehydration should be avoided, and any reversible risks factors reduced where possible. Central venous lines should be removed at the earliest opportunity.

Anti-embolic stockings and intermittent pneumatic compression devices should be used in children over 13 years of age or those weighing >40 kg having surgery lasting >1 hour.

The use of thromboprophylaxis with low molecular weight heparin (LMWH) will depend on the number of associated risk factors the child has balanced against the bleeding risk during the surgical procedure. LMWH used in the perioperative period include:

- Enoxaparin
  - 0.75 mg $kg^{-1}$ every 12 hours in children <5 kg
  - 0.5 mg $kg^{-1}$ every 12 hours in children >5 kg

- Dalteparin
  - 75 IU $kg^{-1}$ every 12 hours, in children <5 kg
  - 50 IU $kg^{-1}$ every 12 hours, in children >5 kg

The anti-coagulated child must be monitored closely whilst on LMWH using anti-factor Xa levels. The placement, removal or repositioning of an epidural catheter should occur at least 12 hours after administering standard prophylactic LMWH doses. The care of children on LMWH requires coordinated care with surgeons, anaesthetists, the pain team and haematology.

## Common Medical Conditions of Childhood

Anaesthetists involved in the care of children must have comprehensive knowledge of common medical conditions found in childhood and their impact on anaesthesia. Collaboration between local hospitals and tertiary centres supports anaesthetists delivering paediatric anaesthesia in district general hospitals.

### Obstructive Sleep Apnoea (OSA)

Sleep-disordered breathing describes a spectrum of conditions, ranging from primary snoring through upper-airway resistance syndrome to OSA. OSA is the most serious condition, with the highest associated incidence of perioperative morbidity and mortality. OSA has a prevalence of 1–4% compared to snoring, which is 20%.

Patients with OSA have repeated episodes of airway obstruction and arousal from sleep. In contrast, those with primary snoring do not have apnoea, hypopnoea or gas exchange abnormalities due to compensatory neuromuscular mechanisms to maintain airway patency.

Several conditions can be associated with OSA:

- Adenotonsillar hypertrophy
- Craniofacial abnormalities
- Hypotonia
- Obesity
- Retrognathia and micrognathia
- Midface hypoplasia
- Macroglossia and glossoptosis
- Genetic conditions

It is worth noting that in conditions such as Down syndrome, mucopolysaccharidosis and achondroplasia, there may be airway obstruction at multiple levels.

Untreated OSA can lead to chronic hypoxia, hypercapnia and respiratory acidosis, leading to pulmonary hypertension and cor pulmonale. Chronic hypoxia is an independent risk factor for right-ventricular hypertrophy.

A detailed history may identify signs and symptoms of OSA:

- Snoring or gasping
- Paradoxical breathing
- Apnoea or cyanosis
- Laboured breathing during sleep
- Daytime somnolence
- Headaches
- Poor performance at school
- Enuresis
- Behavioural changes

A child with a history suggestive of OSA may need further diagnostic investigations such as a sleep study, for example polysomnography (PSG), which is the gold standard for the diagnosis and quantitative description of OSA. PSG continuously monitors physiological variables during different sleep phases and can differentiate primary snoring from OSA. It will also provide a more complete description of obstructive events occurring during sleep. Some patients, especially those with neuromuscular conditions, may display a mixed picture of both central and obstructive sleep apnoea.

Apnoea is defined as a decrease in flow by more than 90% for two breaths or more. Hypopnoea is defined as a decrease in flow by more than 50% coupled with a 3% decrease in oxygen saturation or electroencephalographic evidence of arousal.

The apnoea/hypopnoea index (AHI) quantifies the number of apnoeas or hypopnoeas occurring during one hour of sleep and helps to classify the grade of OSA.

Mild OSA has one to five episodes per hour, moderate OSA is 6–10 per hour and severe is >10 per hour.

PSG is expensive and not readily available, so that overnight oximetry may be used instead. It has high specificity but low sensitivity for the diagnosis of OSA. The McGill Oximetry Scoring System measures the number of clusters of oxygen desaturations. A child with severe OSA would have three or more desaturations to <90%. In addition, the oxygen saturation nadir and the ETCO2 gives

valuable information about the severity of the OSA.

Children with severe OSA and chronic hypoxia are at significant risk of PHT, RVH and cor pulmonale, and this will require investigation with ECG and ECHO.

The ideal GA for children with OSA is sedative and opioid sparing; they are sensitive to opioids, so the dose will need to be reduced and titrated carefully. Regional anaesthesia should be used when appropriate. Such children are at increased risk of airway obstruction and oxygen desaturation at induction and emergence. The anaesthetist must be prepared for this and be able to safely intervene with airway adjuncts. Those children with OSA and airway anatomy that would raise concerns of a potential difficult intubation require careful planning and preparation.

The immediate postoperative period is also a critical time where they may show signs of ongoing airway obstruction and desaturation. Those children on continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BIPAP) at night should bring their non-invasive ventilators as they may need to be used in the immediate postoperative period.

Day surgery should be avoided due to the risk of postoperative airway obstruction, and overnight monitoring will be required in a ward environment able to manage children on non-invasive ventilation.

## Asthma

Asthma is characterised by reversible airway obstruction, inflammation and bronchial hyperreactivity; it is associated with an increased risk of perioperative morbidity and mortality due to bronchospasm and hypoxia. Its incidence is increasing worldwide. Despite the additional risk, severe bronchospasm is relatively rare in asthmatics undergoing GA. Most well-controlled asthmatics can be anaesthetised safely.

Asthma is more commonly seen in atopic children, mediated via IgE hypersensitivity. It may also be seen in ex-premature children with a history of intermittent positive pressure ventilation (IPPV), children exposed to passive smoking and other environmental factors.

Not all wheezing is asthmatic; in some children it may be due to post-viral wheeze or tracheobronchomalacia. These groups are at risk of airway hyperreactivity and of developing bronchospasm under GA.

A thorough preoperative assessment is an essential part of preparing an asthmatic child for elective surgery. A detailed history is required to assess how well controlled the child is, focusing on:

- Symptoms: wheeze, usually polyphonic; nocturnal cough causing sleep disturbance; signs of shortness of breath
- Frequency of acute episodes: whether infrequent and short-lived or frequent requiring admission to hospital
- Triggers such as changes in temperature, exercise, animals and URTI
- Effectiveness of current medication
- Compliance with medication
- Who oversees the management: general practitioner (GP) led or under specialist care
- Hospital admissions requiring intubation, ventilation and admission to the paediatric intensive care unit (PICU)
- A recent course of oral steroids to manage exacerbations

Asthmatic children presenting for elective surgery should be medically optimised, and if this is not the case, surgery should be deferred if possible.

### Investigations

Children under five years of age will not have a formal diagnosis of asthma but will be medically managed with inhalers. Some of these children will not have asthma but will have post-viral wheeze, which will get better with time.

A formal diagnosis requires children over the age of five to engage with testing such as spirometry, FeNO and the bronchodilator reversibility test.

### Perioperative Management

There should be meticulous attention to preoperative assessment and medical optimisation. Continuation of medications in the perioperative period is essential with the avoidance of triggers. Despite comprehensive preparation, acute bronchospasm can occur, and the anaesthetist must have a plan to manage this.

Bronchospasm is more likely at induction, during airway management or at extubation. Vagal stimulation during endoscopy, with peritoneal or visceral stretching, or suctioning of the airway can also trigger bronchospasm.

Initial signs include reduced lung compliance with a rising $ETCO_2$ and a characteristic obstructive pattern on the capnograph tracing. Hypoxia may ensue without appropriate treatment.

Management of intraoperative bronchospasm includes:

- Use 100% inspired oxygen.
- Address reversible causes (e.g. endobronchial intubation or carinal irritation).
- Deepen anaesthesia using a volatile agent.
- Administer a short-acting βeta 2 agonist such as salbutamol or ipratropium bromide, either as a metered dose inhaler via an airway adaptor or as a nebuliser.
- Give IV salbutamol 5–15 mcg $kg^{-1}$ or aminophylline 5mg $kg^{-1}$ over 10–15 minutes.
- Use magnesium sulphate IV 40 mg $kg^{-1}$ as a bronchodilator and anti-arrhythmic.
- Administer hydrocortisone 4 mg $kg^{-1}$ IV.
- Resistant bronchospasm may require IV adrenaline 1 mcg $kg^{-1}$.
- Ventilation strategies should include a slow respiratory rate, with a longer expiratory time to allow for increased airway resistance.
- Higher airway pressures will be required, but the balance should be a peak pressure to deliver an adequate tidal volume to allow oxygenation and tolerating moderate hypercapnia.
- Placement of an arterial line may be required as there will be a discrepancy between $ETCO_2$ and $PaCO_2$.

It is important to remember the potential adverse cardiovascular effects of many of these treatments.

Non-asthmatic causes of bronchospasm should also be excluded, such as anaphylaxis or aspiration of gastric contents or foreign body.

## Cystic Fibrosis (CF)

CF is a multi-system autosomal recessive disease with an incidence of 1:2,500, mainly affecting the exocrine glands. It is prevalent in Western Europe, and the UK carrier rate is 1:25. CF is caused by a mutation in a single gene coding for the cystic fibrosis transmembrane regulator (CFTR), which is located on the long arm of chromosome 7. The genetic defect causing CF was first identified in 1989; more than 2,000 mutations of this gene have now been identified.

The CFTR is a chloride channel located at the apical border of the epithelial cells lining most exocrine glands. Disruption in the chloride channel adversely affects the $Na^+/Cl^-$ balance, causing mucus to be viscid and difficult to clear in children with CF.

Since 2007 there has been national neonatal screening for CF, such that most cases are identified early. Despite this, some cases are still not detected until later life. Carrier testing is available as well as antenatal testing.

CF can present in the neonate as bowel obstruction due to meconium ileus. Other presentations include recurrent respiratory tract infections, failure to thrive and malabsorption.

CF is a multi-system disease, but the effects on the respiratory system tend to be the most significant. Progressive lung disease occurs due to reduced clearance of viscid mucus producing patchy atelectasis. Cycles of inflammation and infection follow with associated chronic hypoxia. The lungs become chronically colonised with pathogens, which include Pseudomonas aeruginosa, Staphylococcus aureus, Haemophilus influenzae, Stenotrophomonas maltophilia, Burkholderia cepacia and Aspergillus. CF patients develop chronic airway obstruction and bronchiectasis. End-stage progression leads to pulmonary hypertension, cor pulmonale and death.

The exocrine function of the pancreas is reduced, and pancreatic duct obstruction leads to reduced absorption of essential vitamins. CF patients require lifelong pancreatic enzyme replacement therapy to maintain normal nutrition. Endocrine function is also affected, and CF-related diabetes (CFRD) may occur in adolescence. Other important sequelae of CF include liver disease, including biliary cirrhosis and portal hypertension; infertility; nasal polyps; and osteoporosis.

Treatment aims to minimise respiratory infections, optimise the patient's nutritional status, slow the disease process and offer symptom control.

Children with CF require carefully planned admission for any elective surgery. This should include a recent medical review by the CF team and recent investigations detailing the child's current medical state (full blood count [FBC], coagulation screen, urea and electrolytes [U&Es], liver function tests, blood glucose, CXR, arterial blood gas [ABG] and spirometry).

Before admission, the frequency of chest physio should be increased in anticipation of the admission, and all nebulised medication should be continued in

the immediate preoperative period. Respiratory complications are the biggest concern and should be anticipated. Surgical and anaesthetic decisions should be taken to reduce respiratory complications wherever possible. For example, laparoscopic surgery is preferable to open surgery since it will reduce pain and diaphragmatic splinting. Short surgical procedures are tolerated best.

### Anaesthetic Considerations

A detailed APOA is required ahead of elective surgery. Surgery may need to be postponed if there are concerns about intercurrent respiratory infection. Perioperative physiotherapy is essential to help with mucus clearance and minimise the risks of respiratory tract infection in the perioperative period. Oxygen therapy will be required postoperatively, and this should be humidified to help with mucus clearance. Airway management will depend on the length and type of planned surgery. Intubation is tolerated and allows access to toileting of the airway and tighter control on ventilation during the surgical procedure. Postoperatively, these children will generally require an overnight stay and may require admission to PICU depending on their preoperative respiratory status. Those with CFRD will require a perioperative plan detailing how to manage their diabetes. Glycaemic control can be challenging due to steroid medication and potential intercurrent infection.

## Bronchopulmonary Dysplasia (BPD)

This chronic lung disease is one of the most common long-term sequelae of premature birth. Advances in the care of premature babies have significantly improved the course of this condition, including:

- Surfactant
- Antenatal steroids
- Lung-protective ventilation strategies

Babies at risk of developing BPD are usually born between 24–28 weeks gestation. Due to advances in the care of premature babies, the clinical course of 'new' BPD is one of mild respiratory distress syndrome (RDS) with a continued need for supplemental oxygen. There are consistent signs of arrested lung development with a simplified alveolar structure and dysmorphic capillaries. 'New' BPD is classified as mild, moderate or severe depending on the baby's gestational age, their continued need for oxygen beyond 36 weeks and their need for positive pressure ventilator support.

Babies and children with BPD presenting for GA may have sequelae of long-term ventilation, such as tracheobronchomalacia, subglottic stenosis and airway granulomas, and are at risk of:

- Ongoing airway reactivity and obstruction
- Oxygen desaturation
- High peak airway pressures during ventilation and higher $PaCO_2$

They are also at risk of pulmonary hypertension (PHT); this carries a high morbidity and mortality rate under GA and should be actively excluded. Infants requiring continuing ventilatory support, a high oxygen requirement, high $PaCO_2$, cyanotic episodes and failure to thrive are at particular risk of PHT.

The assessment needs to detail the respiratory support required when in NICU (type of ventilatory support, duration, etc.) their current level of ventilatory support and ongoing oxygen requirement. Where there are concerns about PHT, ECG and echocardiography will be necessary.

Patients with BPD may also require regular steroid inhalers and diuretics. Those on diuretics must have hypokalaemia excluded. Those on long-term steroids may require perioperative steroid cover.

## Diabetes Mellitus (DM)

DM is one of the most common metabolic disorders, with approximately 96,000 children under 15 years of age developing type 1 DM annually worldwide. Type 1 DM accounts for 90% of new diagnoses, with type 2 accounting for the remainder. The incidence of DM is increasing annually, and the increase in type 2 DM is associated with rising obesity levels.

The gold standard of care for children with DM is an intensive insulin regime, with differential substitution of basal and prandial insulin aiming to achieve optimal metabolic control. Tight glycaemic control reduces rates of acute and long-term complications.

There are several different diabetic regimes which use a combination of ultra-short-, short- and long-acting insulins to mimic normal physiological insulin release, as closely as possible (see Table 7.2).

**Table 7.2** Types of insulins used in management of type 1 DM

| | | Onset (h) | Peak (h) | Duration (h) |
|---|---|---|---|---|
| Ultra-short | Novorapid®<br>Humalog®<br>Glulisine | <0.5 | 1 | 3–4 |
| Short acting | Actrapid®<br>Humulin S® | 0.5–1 | 2–6 | 3–8 |
| Long acting | Insulatard®<br>Glargine<br>Determine | 3–4<br>2–4<br>2–4 | 4–12<br>None<br>None | 10–20<br>20–24<br>12–18 |
| Mixed | Novomix 30® | 0.5 | 2–6 | 8–10 |

Examples of commonly used insulin regimes include:

- Basal bolus/multiple dose insulin (MDI) regime
- Twice daily insulin – biphasic premixed insulin
- Three times daily dosing
- Continuous subcutaneous insulin infusion (CSII)

All diabetic children need a thorough APOA, which should include a history of their diabetic control, including the frequency of episodes of hypoglycaemia, how they present symptomatically and if diabetic ketoacidosis (DKA) has been a feature of their disease. Preoperative investigations should include FBC, U&E, glucose, HbA1C and urinary glucose/ketones.

Essential perioperative planning includes:

- Multidisciplinary care with preoperative assessment and input from the diabetic team can ensure good perioperative glycaemic control.
- First on the list will help to minimise the risk of excessive fasting.
- Insulin control around the perioperative period will be determined by length of surgery and anticipated time to re-establishing oral intake.
- PONV should be actively managed to give the best chance of returning to normal diet and insulin regime as quickly as possible.
- A multimodal approach to analgesia using regional techniques can minimise the need for opioids, which may increase the chance of PONV.
- Any sign of intercurrent infection should be reviewed and surgery postponed if appropriate.

Each hospital should have a guidance document for the management of the diabetic child presenting for surgery. The principles of the protocol are:

- Avoiding hypoglycaemia and DKA
- Minimising fasting times
- Recognising that children will still require insulin even when fasting
- Ensuring that the child is first on the list if possible
- Maintaining blood glucose at 5–10 mmol $l^{-1}$
- Instigating active treatment if blood glucose is <5 or >14 mmol $l^{-1}$
- Checking blood glucose hourly or more frequently if glycaemic control is difficult
- Checking blood or urine for ketones

### Minor Surgery Guidance

Diabetic children having minor surgery, which is expected to last <90 minutes with no delays to returning to eating and drinking and low risk of PONV, have the following approach to their perioperative insulin:

- Those on MDI take their normal basal dose of insulin.
- Those on an SCII continue on the pump throughout the admission.

Blood glucose must be monitored regularly and a bolus dose of insulin is given if blood sugar is >14 mmol $l^{-1}$. Children are encouraged to eat and drink after surgery and are given their normal dose of rapid-acting insulin with their first postoperative meal. Children who are managed with twice-daily insulin cannot be managed in the same way for minor surgery and are required to follow the guidance for major surgery.

### Major Surgery Guidance

Diabetic children on MDI or twice-daily insulin having planned major surgery stop their normal basal insulin dose before surgery and commence on a variable rate insulin infusion (VRII) along with maintenance fluids (5% dextrose/0.45% saline/0.15% KCl) at the same time (see Table 7.3). Those on an SCII should have the pump stopped two hours prior to surgery and have a VRII started at the same time as the pump is stopped. Blood glucose should be checked hourly and the VRII altered according to the child's blood glucose result.

In an emergency setting, it is important to contact the diabetic team to guide perioperative management. DKA can present with signs of an acute abdomen, so DKA must be excluded; if present, treatment should follow the DKA protocol. Emergency surgery should be delayed until hypovolaemia and electrolyte deficit has been corrected. The management and rehydration of a child with DKA need to be done carefully and slowly with isotonic fluids.

## Adrenal Insufficiency (AI)

Many children presenting for surgery will have adrenal insufficiency due to primary, secondary or tertiary causes, as detailed in Table 7.4.

These children are at increased risk of adrenal crisis in the perioperative period. Any child at risk of adrenal insufficiency needs a detailed history identifying the cause of the AI and their regular management regime when the child is well. If the child has had an adrenal crisis, this should be documented, detailing the background and management. Children with AI will have a rescue regime, which they follow when they are unwell, as in sickness, infection or trauma. Any admission should be made in close consultation with the child's endocrinologist.

The following key points should be adhered to:

- The plan should be medically optimised and discussed with the endocrine team to agree on the perioperative steroid plan.
- Test preoperative FBC, U&Es and blood glucose.
- Regular steroid medication should continue uninterrupted.

**Table 7.3** VRII regime (50 units of actrapid insulin in 50 mls of 0.9% saline to give a concentration of 1 u $ml^{-1}$)[a]

| Blood glucose mmol $l^{-1}$ | Insulin infusion rate units $kg^{-1}$ $h^{-1}$ |
|---|---|
| <4 mmol $l^{-1}$ | 0.01 |
| 4–6.9 mmol $l^{-1}$ | 0.02 |
| 7–8.9 mmol $l^{-1}$ | 0.03 |
| 9–12 mmol $l^{-1}$ | 0.04 |
| >12 mmol $l^{-1}$ | 0.05 |

*Note:* [a] The infusion regime needs to be monitored closely and may need altering in the setting of severely ill children or those on steroid medication. Do not stop the infusion if the blood glucose is <4 mmol $l^{-1}$, as this will cause rebound hyperglycaemia. The insulin rate should be reduced further and the glucose infusion increased to return blood glucose to normal level.

**Table 7.4** Classification of adrenal insufficiency

| Classification of adrenal insufficiency (AI) | Underlying causes |
|---|---|
| Primary AI | Abnormalities of the adrenal gland<br>Most common cause in childhood being congenital adrenal hyperplasia (CAH)<br>Incidence 1:8–10,000 |
| Secondary AI | Abnormalities of the hypothalamus +/– pituitary, e.g. congenital disorders of the hypothalamus +/– pituitary or acquired as a consequence of brain tumour |
| Tertiary AI | Long-term treatment with glucocorticoids with suppression of the hypothalamic–pituitary–adrenal axis – any child taking more than 10–15 mg $m^{-2}$ hydrocortisone equivalent per day for more than a month regardless of the route of administration.<br>Asthma, juvenile chronic arthritis, renal disease, inflammatory bowel disease (IBD), Duchenne muscular dystrophy (DMD) |

- The patient should be first on the list to minimise dehydration.
- Administer IV bolus of hydrocortisone 2 mg $kg^{-1}$ at induction. Major cases can be followed by either a hydrocortisone infusion or four-hourly boluses of hydrocortisone (see 'Further Reading').
- Particular attention should be given to glycaemic control.
- Dexamethasone has no mineralocorticoid action.

CAH is a group of autosomal recessive disorders characterised by abnormal cortisol synthesis. Consequently, children with CAH will have absent cortisol production, elevated levels of ACTH and abnormal production of androgen and mineralocorticoid hormones. Children with CAH require lifelong treatment with glucocorticoids and, depending on the cause of their CAH, mineralocorticoid may also be required. Glucocorticoids replace the deficient cortisol but also help to suppress excess androgen production (see Table 7.5).

The primary goal of treating classical CAH is to reduce the excess androgen production and replace deficient hormones. Proper treatment with the correct dosage of these hormones is crucial in preventing adrenal crisis and virilisation.

Deficiency of 21-hydroxylase is the most common cause of CAH and is responsible for 95% of all CAH cases. The remaining 5% of cases of CAH are caused by other essential enzymes required in cortisol biosynthesis such as 11-beta–hydroxylase. CAH can be classified into classical CAH and non-classical CAH.

Classical CAH due to 21-hydroxylase deficiency can be subdivided into two groups. In the salt-losing form (accounting for 75% of classic CAH), not only is there absent cortisol production but also reduced production of aldosterone, with associated problems with fluid and salt regulation. A concomitant increase in androgen production can present with over-virilisation and ambiguous genitalia in females born with CAH. The simple virilising group accounts for 25% of classic CAH and does not have deficient aldosterone production, so there is normal regulation of fluid and salts.

Children with CAH are at risk of adrenal crisis, electrolyte abnormalities and altered glycaemic control. Stressors such as intercurrent infection or surgery can increase their requirements of these essential hormones. All children with CAH will have a rescue regime for steroid replacement during periods of increased demand and should be reviewed preoperatively by their endocrine team, which can advise on the best perioperative management of their steroid requirements. Normal steroid medication should not be interrupted preoperatively, and they will require additional steroid cover at induction, usually 2 mg $kg^{-1}$ of IV hydrocortisone, repeated every four hours. Some children will require a hydrocortisone infusion. All children with classical CAH should be reviewed preoperatively to ensure that their endocrine team is aware of the planned surgery and can advise on the best perioperative management of their steroid requirements.

The AAGBI has published comprehensive guidance on the management of adults and children requiring glucocorticoids in the perioperative period. See 'Further Reading'.

**Table 7.5** Glucocorticoids and their equivalent doses

| 1 mg of prednisolone is equivalent to: | |
|---|---|
| Dexamethasone | 150 mcg |
| Betamethasone | 150 mcg |
| Methylprednisolone | 800 mcg |
| Triamcinolone | 800 mcg |
| Deflazacort | 1.2 mg |
| Hydrocortisone | 4 mg |

## Obesity

Childhood obesity is a major health concern, with one in three children in the United Kingdom being obese by the age of nine. The WHO has estimated that in 2019 38 million children under 5 and more than 340 million children and adolescents aged between 5 and 19 were overweight or obese.

In most cases, obesity is preventable, stemming from excessive calorie intake and physical inactivity. Secondary causes, linked to genetic syndromes such as Prader–Willi, endocrine or neurological disorders or medication related, are rare but important to exclude.

### Defining Obesity in Childhood

The simple application of BMI (kg $m^{-2}$) in isolation to define obesity in children can be misleading as growth is highly variable, particularly in young children. WHO gender-specific body mass index (BMI) growth charts are considered the gold standard for defining obesity in childhood. In children under five, obesity is defined as having a BMI >3 standard deviation (SD) above the median; in older children, obesity is defined as having a BMI >2 SD above the median. The WHO growth charts may also be used to determine the ideal body weight (IBW) for that child, which may be helpful for perioperative drug dosing.

Children who are obese have an increased perioperative risk due to a higher incidence of:

- Hypertension
- OSA and PHT
- Asthma
- Insulin dependent type 2 DM
- GORD

Preoperative assessment should identify and investigate these associated risk factors. Investigations should include:

- Blood pressure (BP) and ECG
- FBC, U&Es, cholesterol and blood glucose
- Sleep study
- ECHO
- IBW calculation

If GORD is a significant problem, appropriate medications should be commenced in advance.

Preoperative consideration must be given to the theatre environment, including the weight limit of the theatre table and having manual handling equipment available. Careful positioning is very important to minimise the risk of pressure sores and nerve injury.

It is well recognised that obese children are at increased risk of perioperative respiratory complications, including laryngospasm, bronchospasm and oxygen desaturation due to reduced FRC, forced vital capacity (FVC) and lung compliance. The abnormal respiratory mechanics are accentuated in the supine position when the abdominal contents exert greater pressure on the diaphragm. Mask ventilation can be difficult, and an oral or nasal airway adjunct may be necessary. Where possible, all obese children should be preoxygenated, and it is generally considered safer to intubate obese children, even for short surgical procedures.

Drug administration can be problematic, as obesity results in variations in the pharmacokinetics and pharmacodynamics of many drugs. Obesity increases the volume of distribution, particularly for lipophilic drugs, and can alter drug clearance and elimination half-life. Renal clearance is higher in obesity, increasing linearly with lean body weight (LBW) rather than total body weight (TBW).

Consequently, there are recommendations for some drugs to be administered according to TBW, such as succinylcholine, whilst others such as propofol, rocuronium and morphine may be more appropriately based on IBW or LBW:

$$LBW = IBW + 0.3\ x\ (TBW - IBW)$$

However, very few studies have been undertaken on obesity-related pharmacokinetic changes in children, and these recommendations are largely extrapolated from adult studies.

In practice, most anaesthetic drugs can be administered titrated to effect, which will usually be less than a TBW dose. In an emergency, succinylcholine should be administered at a dose of 1mg $kg^{-1}$.

## Haemoglobinopathies

### Sickle Cell Anaemia

SCD is a congenital haemoglobinopathy caused by a mutation on chromosome 11. This mutation results in an amino acid substitution on the beta-globin subunit leading to the formation of HbS. HbS is unstable and can precipitate out of solution when in the deoxygenated state forming the classical sickle or curved red cell. SCD is most commonly found in Africa, Southeast Asia, the Middle East and parts of the Mediterranean. The heterozygous state is protective against *Plasmodium falciparum*, and the distribution of SCD closely matches the distribution of malaria worldwide.

Since 2006, all newborns in the United Kingdom have been tested for SCD as part of their neonatal blood spot screening. The incidence of SCD in England in 2021 was 1:2,000 live births.

SCD has an autosomal recessive pattern of inheritance. Those that are heterozygous have SCT and produce both HbA and HbS. The proportion of HbS is approximately 30–40%. Those with SCT have a more benign disease and only show signs

of sickling when under extreme physiological conditions, for example when their oxygen saturations are approximately 40% or during cooling for cardiopulmonary bypass surgery.

The homozygous state (HbSS) has almost 100% HbS, which is a serious life-limiting condition associated with:

- Chronic haemolytic anaemia, due to a reduced red cell life span in SCD of 12 days (versus 120 days for normal red cells)
- Vaso-occlusive disease affecting particularly the bones, extremities, bowel and lungs
- End organ failure
- Severe pain
- Premature death

Sickling in patients with HbSS can be triggered by hypoxia, hypothermia, infection and dehydration.

Surgery in patients with SCD requires meticulous planning, optimisation and input from haematology from the beginning. Clinical history should include the number of acute crises, their triggers and how they were managed, treatment to date and a detailed transfusion history. End organ disease should be investigated appropriately and documented (see Table 7.6).

Patients with SCD will be on folic acid due to the high cell turnover rate. They should be vaccinated against *Strep pneumoniae, Neisseria meningitides,* Hepatitis B, *Haemophilus influenza* Type B; they may be on a penicillin to reduce the chance of infection with pneumococcus. Hydroxycarbamide is used to increase the levels of HbF, which reduces the episodes of sickling and the need for transfusion. Those on hydroxycarbamide may be at risk of neutropenia, so this should be considered as part of the preoperative investigations.

Baseline investigations should include:

- FBC, U&Es and liver function tests (LFTs)
- Hb electrophoresis to quantify the percentage of HbS
- Crossmatch and antibody screening
- CXR
- Echocardiogram
- Spirometry to assess lung function
- Neurological assessment if there is a history of stroke – transcranial Doppler or MRI may be recommended

Patients with SCD are at risk of sickling in the perioperative period. Preoperative exchange transfusion may be indicated, depending on the type of surgery and Hb level. If undertaken, it should be done well in advance, overseen by haematology, aiming for a Hb of 100 g $l^{-1}$ and a HbSS of <30%. To avoid hyperviscosity, the Hb should not increase by more than 40 g $l^{-1}$ in a single transfusion.

Principles of perioperative care for those with SCD include the following:

- Avoid hypovolaemia – consider admitting the night before for IV fluids, encourage oral fluids until one hour before surgery.

**Table 7.6** Clinical impact of SCD

| End organ | Impact of SCD |
|---|---|
| Cardiac | Cardiomegaly and congestive cardiac failure due to anaemia and pulmonary hypertension |
| Respiratory | Multiple pulmonary infarcts leading to pulmonary hypertension<br>Acute chest syndrome<br>OSA |
| Gastrointestinal | Splenomegaly with splenic sequestration and acute drops on haematocrit and platelets<br>Functional asplenism with risk of infection<br>Gallstones<br>Hepatic enlargement but usually with maintained hepatic function |
| Central nervous system (CNS) | Increased incidence of transient ischemic attack (TIA), thrombotic and haemorrhagic stroke |
| Renal | Renal impairment and painful priapism |
| Infections | Prone to infections<br>Parvovirus leading to hypoplastic crisis |
| Bones | Avascular necrosis<br>Extramedullary haematopoiesis – frontal bossing, enlarged maxilla with potential for difficulty in managing the airway |
| Haematology | Bone marrow failure<br>Haemolytic anaemia, hypoplastic anaemia |

- Maintain oxygenation – this may require supplemental oxygen to maintain $SpO_2 >$ 95% postoperatively.
- Maintain normocarbia and avoid respiratory acidosis.
- Avoid hypothermia and acidosis.
- Avoid using vasoconstrictors.
- Minimise use of tourniquets (as explained in the following text).

The use of tourniquets is controversial, with concern around ischaemia of the limb distal to the tourniquet and sickling due to acidosis and release of vaso-active substrates when the tourniquet is released. The decision to not use a tourniquet needs to be weighed against the risk of sickling triggered by significant blood loss from major surgery. If a tourniquet is used, the limb must be fully exsanguinated, and there needs to be meticulous attention to the patient's acid/basis status and volume status. As always, the tourniquet time should be minimised.

Analgesia should be multimodal, using regional techniques where appropriate to minimise opioid use. If using opioids, the patient must be monitored to ensure they are well oxygenated, receiving supplemental oxygen if required. Patients with SCD having major or emergency surgery or those with significant end organ disease, such as significant OSA or PHT, are best looked after in a critical care setting.

## Thalassaemia

This is a group of hereditary anaemias, which are caused by defective synthesis of the alpha chain (alpha thalassaemia) or the beta chain (beta thalassaemia) of haemoglobin.

Beta thalassaemia is due to deletion or mutations in the HBB gene located on chromosome 11. The worldwide prevalence of symptomatic beta thalassaemia is thought to be 1:100,000. It is most commonly seen in the Middle East, Southeast Asia, India and China.

There are three forms of beta thalassaemia, depending on the inheritance: beta thalassaemia minor (or trait), intermedia and major. Children who are heterozygous for beta thalassaemia are said to have beta thalassaemia minor or trait and are either asymptomatic or show very mild anaemia. Those with homozygous inheritance have beta thalassaemia major, also known as Cooley's anaemia. In the most severe form, children present with microcytic, hypochromic anaemia and signs of extramedullary haematopoiesis. Treatment depends on the severity of the anaemia but may include oral supplementation with folic acid or regular blood transfusions combined with oral chelating agents.

Alpha thalassaemia is prevalent in Southeast Asia with an incidence of 1:10,000 compared to an incidence in Northern Europe of 1:1,000,000. Those who are heterozygous for the condition have alpha thalassaemia trait and can be asymptomatic or have very mild anaemia. Those who are homozygous exhibit a more severe phenotype, featuring a microcytic, hypochromic anaemia that may require blood transfusions and other supportive measures.

Children with thalassaemia require preoperative investigations to assess the extent of the disease. Those with severe phenotypes who require regular blood transfusions are at risk of iron overload, so an assessment of their cardiac status, including an ECHO, will be required preoperatively. Extramedullary hematopoiesis may lead to facial changes due to medullary hyperplasia, and this may make laryngoscopy more difficult.

## Glucose-6-Phosphate Dehydrogenase (G6PD) Deficiency

Glucose-6-phosphate dehydrogenase (G6PD) is a housekeeping enzyme expressed in all cells where it catalyses the first step in the pentose phosphate pathway. In red blood cells, this is the sole pathway for the production of NADPH, which is required to maintain glutathione in a reduced state. Failure of this process impairs the ability of the red cell to deal with oxidative stress, which may lead to haemolytic episodes and anaemia that can be severe and, in some cases, fatal.

G6PDd is the most common red blood cell (RBC) enzymopathy in humans, with a global prevalence of 400 million people. There are nearly 200 mutations identified, but a few mutations account for most cases. The most common is the Mediterranean variant, followed by the African (G6PDd – A), Indian and Southeast Asian variants.

The magnitude of enzyme deficiency determines the severity of disease: Class I variants have severe enzyme deficiency with chronic non-spherocytic haemolytic anaemia (<10% residual

enzyme activity), Class II variants also have severe enzyme deficiency (<10% residual enzyme activity) but with intermittent acute haemolysis and Class III variants have moderate enzyme deficiency (10–60% residual enzyme activity) with intermittent acute haemolysis. Class IV and V variants are of no clinical significance – Class IV has no enzyme deficiency, and Class V has increased enzyme activity.

Perioperative care is determined by the type of deficiency the patient has. Patients with G6PD deficiency should not have elective surgery during a haemolytic episode or in the presence of intercurrent infection, which is known to be a potential trigger. Preoperative blood tests should include FBC, LFTs, reticulocyte count, lactate dehydrogenase (LDH) and haptoglobins.

Potential triggers of oxidative stress should be minimised. Measures include avoiding dehydration due to excessive fasting and addressing perioperative anxiety. Intraoperative temperature and glycaemic control should be monitored closely. It is important to be aware of other triggers in this patient group, such as:

- Fava beans
- Dapsones
- Antimalarials, such as primaquine
- Methylene blue
- Nitrofurantoin
- Quinolones, such as ciprofloxacin
- Sulphonamides, such as co-trimoxazole

## Coagulation Disorders

Whilst relatively rare, congenital disorders of coagulation may pose challenges in the perioperative period. Haematology advice should be sought where appropriate.

Von Willebrand disease (VWD) is the most common congenital coagulation disorder. It is caused by a qualitative or quantitative deficiency of von Willebrand factor (VWF), required for platelet aggregation and adhesion. VWF also helps maintain the levels of factor VIII as it acts as a carrier for this factor and in doing so reduces factor VIII clearance.

The incidence is 1:100, with the vast majority of those afflicted being asymptomatic. It is only clinically significant in 1:10,000 patients. VWD has variable autosomal inheritance (dominant or recessive) depending on the subtype. Type 1 VWD is a mild form of the disease due to qualitative deficiency of VWD, type 2 has several subtypes and there is both qualitative and quantitative reduction in VWF. Type 3 is the most severe form as there is no VWF.

Perioperative management depends on the type of VWD. Desmopressin (1-desamino-8-D-arginine vasopressin, DDAVP) works by stimulating the release of VWF and can be used in VWD type 1. Type 3 VWD requires replacement of VWF and factor VIII, with the addition of platelet transfusion if bleeding persists despite replacement of these factors.

Haemophilia can be classified as haemophilia type A, B or C depending on the deficiency of factors VIII, IX or XI, respectively. Haemophilia type A and B are inherited as X-linked recessive disorders, so males are affected, whilst females are carriers. The incidence of haemophilia A is 1:5,000–10,000. Haemophilia B is rarer with an incidence of 1:35,000–50,000.

In the perioperative period, children with haemophilia require replacement of the deficient factor VIII or IX. They should also be assessed for the presence of inhibitors to factor VIII, and if they are present, treatment with recombinant activated factor VII (rFVIIa) or factor VIII inhibitor bypassing activity (FEIBA) may be required.

Preoperatively, patients with inherited coagulation disorders need:

- Detailed history of the underlying condition, type of coagulation disorder and its severity
- History of previous bleeding episodes and their management
- Response to DDAVP, factors VIII and IX
- baseline bloods – FBC, coagulation screen, factor assay, fibrinogen level
- An MDT approach with involvement from haematology, surgery and anaesthesia
- A perioperative plan detailing the management of the coagulation disorder – use of DDAVP, tranexamic acid, recombinant factors or plasma

## Epilepsy

Epilepsy is the most common neurological condition of childhood, with an incidence of 0.5–1% of the paediatric population. The incidence is highest in the first year of life and 1:150 children will be diagnosed before the age of 10. The incidence appears to be declining in high-income countries.

Primary or idiopathic epilepsy has a genetic predisposition with a 1.5–3% risk with paternal inheritance and a 3–9% risk with maternal inheritance. Secondary epilepsy is due to pre-, peri- and postnatal causes. Prenatal causes of epilepsy include inborn errors of metabolism and chromosomal abnormalities, such as trisomy 21. Prenatal infections and congenital malformations such as tuberous sclerosis and neurofibromatosis can also be a cause.

Perinatal causes include hypoglycaemia, hypocalcaemia, hypoxic ischaemia events and intraventricular haemorrhage, which are more commonly found in premature babies. Postnatal events such as infections, including meningitis, cerebral abscess, trauma due to non-accidental injury, or malignancy, can all cause the development of epilepsy.

Seizures can be either partial (focal) or generalised. Partial seizures can show motor, sensory or autonomic characteristics depending on the region of the cerebral cortex from which they originate. In a generalised seizure, there is bilateral symmetrical electrical activity with associated generalised muscle contractions. Seizures are defined as simple when there is no loss of consciousness and complex when consciousness is lost. Infantile spasm, myoclonic seizures and benign partial seizures are very specific to the paediatric population.

Children with epilepsy will be on anti-epileptic drugs (AEDs). The type of drug will depend on the type of seizure, frequency, age of child and side effect profile of the AED. AEDs have significant drug interactions and side effects. Commonly used AEDs such as phenytoin and carbamazepine are hepatic enzyme inducers. Some are also hepatotoxic and can cause thrombocytopenia and platelet abnormalities, such as sodium valproate and carbamazepine. It is therefore important to understand the side effect profile of the AEDs the child is on and check this preoperatively.

Several anaesthetic agents produce epileptiform EEG activity (sevoflurane) or are proconvulsant (ketamine and alfentanil). Ketamine and alfentanil should be avoided, but sevoflurane can be used despite the epileptiform activity that is part of the pharmacological profile.

Children with epilepsy should have a detailed history and examination focusing on:

- Type of seizures, triggers, frequency and duration
- How well controlled the seizures are
- What AEDs the child is currently on – what side effects, if any, has the child had
- What rescue medication the family has and how often is this being given to the child

In the elective setting, the child's seizures should be optimally controlled, but this may not be possible in the child presenting with multiple drug-resistant seizures. Continuing medication in the perioperative period is essential. If there is any anticipated delay to returning to oral intake, then a parenteral form of the AED should be started with the guidance of the neurology team.

A ketogenic diet which is high in fat and low in protein and carbohydrate is used in children with intractable epilepsy or those who struggle with the side effects of AEDs. There are several important issues to consider:

- Avoid IV fluid containing glucose as this may precipitate a relapse in seizure control.
- Neonates and ex-prems are at risk of hypoglycaemia – in this setting, IV glucose should be used and closely monitored to maintain blood glucose within normal limits.
- Monitor the blood sugar perioperatively and aim to keep the glucose level between 3–4 mmol $l^{-1}$.
- Avoid Ringer's lactate as this can compound a metabolic acidosis.
- Urinary ketones should be monitored and kept in the range of 8–16 mmol $l^{-1}$.
- Acid-base status should be monitored regularly along with the blood glucose level.

## Cerebral Palsy (CP)

CP is the most common motor disorder in children. It describes a diverse group of permanent neurological disorders characterised by varying degrees of sensory, motor or intellectual impairment.

The incidence of cerebral palsy (2:1000 live births) has changed little over the past 30–40 years despite improvements in antenatal care. This has been attributed to increased survival rates in premature babies who have a significantly higher incidence (10–50 times higher) of CP. Eighty per cent of cases develop antenatally, and 20% develop postnatally in the first two years of life. In 30% of cases, no significant abnormality can be identified.

The classification is based on a description of the resting muscle tone – spastic, (accounting for

**Table 7.7** Summary of clinical issues with the child with CP

| Affected system | Clinical Issues |
|---|---|
| Respiratory | Scoliosis with restrictive lung disease – pulmonary hypertension and cor pulmonale at the extreme<br>Respiratory impairment due to chronic aspiration and weak respiratory muscles<br>Chronic lung disease (CLD) due to neonatal RDS<br>Reduced cough |
| Cardiovascular | Difficult to assess exercise tolerance due to limited mobility |
| Gastrointestinal | Poor nutrition due to reduced ability to chew and swallow – combination of oro-motor dysfunction and pseudobulbar palsies<br>Unsafe swallow<br>GORD<br>Malnutrition with anaemia and electrolyte abnormalities |
| Temperature control | Large surface area to body mass, underweight, reduced subcutaneous fat, thin skin and atrophic muscles |
| Musculoskeletal | Spasticity, contractures leading to deformity and dislocation<br>Pain and issues with mobilising and or sitting and positioning are commonplace |

80% of children with CP) dyskinetic, ataxic and mixed. The Gross Motor Function Classification System (GMFCS) is used to characterise different levels of function with the emphasis on the child's ability rather than their disability. A child with GMFCS level 1 can walk unaided and has gross motor skills but will have issues with speed, balance and coordination. At the other end of the scale, a child with GMFCS level 5 will be wheelchair dependent and have issues with anti-gravity control of the head and trunk.

It is important to remember that in the most severe form, CP can have a wide-reaching impact on the patient's neurological, musculoskeletal, gastrointestinal, respiratory and urological function. The patient needs to be medically optimised with careful consideration of the child's physiological status.

## Preoperative Considerations

Assessing the child's intellectual and cognitive level is an essential part of the preparation. This ensures that they have as much input to the process as possible. The child's parents or carers will be invaluable in the perioperative period as they usually have expert knowledge of the child's chronic medical conditions and how best to communicate with them. Children with CP can have significant deformities such as scoliosis and limb deformities, and the parents or carers will be able to guide positioning and support of the child.

Table 7.7 summarises the key clinical issues that should be reviewed and considered. Many children with CP will be on several drugs, including anti-epileptics, antispasmodics, antisialogues, analgesics, proton pump inhibitors/antacids and antibiotics. It is important that these medications are continued in the perioperative period to avoid acute withdrawal, which can present with worsening seizure control or muscle spasms.

Anxiety in the perioperative period can be an issue due to the child being unable to communicate easily or because of previous bad experiences. Sedative premedication needs to be used with caution in CP patients with significant hypotonia.

Preoperative investigations need to be considered as many CP patients have significant comorbidity. Recent blood tests including FBC and U&E, are usually required. Other investigations, such as CXR and more detailed assessment of respiratory status, should be considered where appropriate.

Analgesia is a key consideration. A multimodal approach including regional techniques should be used where appropriate. The impact of opioids on the respiratory system should be considered, incorporating appropriate monitoring and titration to effect. Postoperative spasms can be an issue in children with CP and require anti-spasmodics such as diazepam or midazolam to avoid the cycles of spasm precipitating pain. A careful balance is required to reduce pain and muscle spasms without compromising respiratory function.

## Renal Failure

The UK Renal Registry (UKRR), in its 22nd annual report, quotes the incidence of children requiring long-term treatment for kidney failure as nine in every 1 million of the UK's child population. The majority are aged between 12 and 16 years. In 50% of cases, there is an abnormality of the structure of the kidneys and urinary tract that has been there since birth, such as renal agenesis. Other causes include glomerular disorders and inherited/familial conditions, such as Alport syndrome or polycystic kidney disease (PKD).

There are five stages of chronic kidney disease (CKD). Children in stage 1 have a normal glomerular filtration rate (GFR; normal $>90$ ml $min^{-1}$ per 1.73 $m^2$) but have evidence of kidney damage, such as proteinuria. Those in stage 5 are in established renal failure and have a GFR of $<15$ or are on a form of dialysis.

CKD is a multisystem disease. Children in CKD stages I–III are generally asymptomatic, with symptoms evolving as renal function declines.

Children presenting with CKD or chronic renal failure (CRF) require a detailed history and assessment, specifically addressing:

- The underlying cause of CRF
- Whether the child is anuric and at risk of fluid overload or oliguric and at risk of hypovolaemia
- Their normal fluid balance – 'dry' and 'wet' weight
- Current management, such as peritoneal or haemodialysis and any complications of these treatments to date
- Suitability for transplantation
- Cardiovascular complications – hypertension, left-ventricular hypertrophy (LVH), uraemic pericarditis, uraemic cardiomyopathy, congestive cardiac failure (CCF) and pulmonary oedema
- Pleural effusions and diaphragmatic splinting in those on peritoneal dialysis
- Peripheral and autonomic neuropathy
- Altered biochemistry – hyperkalaemia and risk of dysrhythmia and arrest, hypocalcaemia
- Metabolic acidosis
- Gastrointestinal disease – uraemic induced anorexia and gastroparesis and delayed gastric emptying
- Normochromic, normocytic anaemia due to reduced erythropoietin production and uraemic bone marrow suppression
- Whether the condition is part of a syndrome, in which case other aspects of the disease need to be considered
- The psychological impact of chronic disease

Preoperative investigations should include FBC, coagulation screen, U&Es, GFR, ECG and ECHO +/– CXR.

## Perioperative Considerations

Nephrologists are central to the perioperative care of these children. All normal medication should be continued. The fluid status of the child and the timing of the last haemodialysis (HD) sessions is important. Ideally, surgery should wait for four to six hours after dialysis to allow fluid shifts to settle and take into consideration the use of heparin during HD. In an emergency setting, HD can be done with minimal or no heparin to expedite surgery postdialysis. Premedication with antacids should be considered as well as anxiolytics.

Induction should consider the potential of delayed gastric emptying, so a modified rapid sequence induction may be required. Alterations in pharmacokinetics need to be recognised and drug dosing altered accordingly. Vascular access can be challenging, and central venous access may be required. It is prudent to avoid, if possible, vascular access at the wrist or elbow of the non-dominant hand as arteriovenous fistulas may be required for HD.

**Key Points**

- APOA in advance of admission is the standard of care.
- Side effects and risks need to be discussed in advance and supported by written or electronic information leaflets.
- Perioperative MDTs should be held to discuss high-risk patients and agree on ceilings of care.
- A new murmur which is heard in systole, pansystolic or early diastole, associated with clinical signs or ventricular hypertrophy confirmed on ECG, requires a cardiology review prior to a general anaesthetic.
- Children can drink water up to one hour before GA.

- Poorly controlled asthmatics who have had a recent URTI are at increased risk of bronchospasm, and elective surgery should be deferred.
- Infants with BPD are at increased risk of airway hyperreactivity and PHT in extreme cases.
- Acute illness can precipitate DKA and can have a similar presentation to an acute abdomen. Ketoacidosis should be excluded.
- Coagulation disorders require specific management of the underlying condition.
- The prevalence of obesity is increasing, and comorbidities associated with obesity can present in adolescence.
- Epileptics on a ketogenic diet should avoid dextrose containing IV fluids and should have close monitoring of the blood glucose, which should be kept between 3–4 mmol $l^{-1}$.

## Further Reading


Association of Paediatric Anaesthetists of Great Britain and Ireland (APAGBI). Guidelines on the peri-operative management of patients with sickle cell disease. *Anaesthesia* 2021; 76(6):805–17. https://doi.org/10.1111/anae.15349.

AAGBI. Preassessment services for children undergoing surgery or procedures. Spring 2022. Available at: www.apagbi.org.uk/sites/default/files/2022-05/Best%20Practice_Preassessment%20standards%20in%20Children%20%202022%20-%20Published.pdf. Accessed 21 April 2024.

Haché M, Sun LS, Gadi G et al. Outcomes from Wake Up Safe, the pediatric anesthesia quality improvement initiative. *Paediatric Anaesthesia* 2020; 30 (12):1348–54.

International Society for Pediatric and Adolescent Diabetes. 2018. ISPAD clinical practice consensus guidelines. Available at: www.ispad.org/general/custom.asp?page=ISPADGuidelines2018. Accessed 21 April 2024.

Masaracchia MM, Lee M, Dalesio NM. Obesity in childhood. *BJA Education* 2022; 22(5):168–75. https://doi.org/10.1016/j.bjae.2021.12.003.

Morgan J, Checketts M, Arana A et al. Prevention of perioperative venous thromboembolism in pediatric patients: guidelines from the Association of Paediatric Anaesthetists of Great Britain and Ireland (APAGBI). *Paediatric Anaesthesia* 2018; 28(**5**):382–91.

National Institute for Health and Care Excellence Asthma. 2021. Diagnosis, monitoring and chronic asthma management. Available at: Asthma: diagnosis, monitoring and chronic asthma management - PubMed (nih.gov). Accessed 21 April 2024.

Thomas M, Morrison C, Newton R, Schindler E. Consensus statement on clear fluids fasting for elective pediatric general anesthesia. *Paediatric Anaesthesia* 2018; 28(5):411–4. https://doi.org/10.1111/pan.13370.