

Associated medical conditions in children

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Abstract

An understanding of the risk factors associated with anaesthesia in children with acute or chronic associated medical conditions is important to direct the pre-operative assessment and preparation and to optimize the anaesthetic plan in order to anticipate and prevent perioperative complications. Here, we outline the relevant clinical features and anaesthetic management of some common medical conditions in children. For routine pre-operative assessment, see pages 489–94 (in this issue).

Keywords asthma; autism; children; cerebral palsy; Diabetes mellitus; Down's syndrome; epilepsy; gastro-oesophageal disease; obesity; obstructive sleep apnoea; pre-operative assessment; sickle cell disease

Upper respiratory tract infection

Acute upper respiratory tract infections (URTIs) include rhinosinusitis (common cold), sinusitis, pharyngitis/tonsillitis, ear infections and laryngitis. Children experience 3–8 URTIs per year and, following an URTI, airway hyper-reactivity persists for up to 6 weeks. Children with URTIs are at increased risk of perioperative respiratory adverse events. Whilst the incidence of serious events is low, they have a 2- to 10-fold increased risk of laryngospasm, bronchospasm and hypoxaemia, 11-fold if their trachea is intubated. However, routine cancellation of every child with a URTI is impractical for both families and hospitals.

Factors that increase the risks of adverse events in children with URTI:

URTI related

- Fever (>38 °C)
- Purulent secretions
- Parental view of severity of symptoms
- Malaise, lethargy, decreased appetite
- Lower respiratory tract signs
- Productive cough

Child factors

- Age <1 year
- History of prematurity

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Learning objectives

After reading this article, you should be able to:

- list the clinical features of children with upper respiratory tract infections, asthma, obstructive sleep apnoea, cystic fibrosis, sickle cell disease, anaemia, obesity, diabetes mellitus, renal failure, cerebral palsy, epilepsy, autism, Down's syndrome, oncological diseases, latex allergy, pregnancy, gastro-oesophageal reflux and on long-term steroids
- Focus pre-operative assessment to ascertain the severity and current status of these specific conditions
- describe appropriate pre-operative investigation and optimization of these medical co-morbidities
- outline key issues for subsequent anaesthesia.

- Sickle cell disease
- History of snoring
- History of reactive airway disease (asthma)
- Parental smoking

Anaesthesia and surgery factors

- Airway instrumentation (endotracheal tube (ETT) > laryngeal mask airway (LMA) > face mask)
- Surgery involving the airway
- Major surgery
- Anaesthetist with limited paediatric anaesthesia experience

The best predictor of perioperative complications is the parent's perception of severity of disease. Children with URTI-related risk factors, especially if they have any other risk factor, should have elective surgery deferred for 4–6 weeks. Management decisions in these children should involve a senior anaesthetist.

Anaesthetic management:

- Face mask or LMA should be used in preference to endotracheal tube if appropriate.
- In intubated children, the airway should be suctioned under deep anaesthesia.
- Airway problems must be anticipated on induction and emergence.

Asthma

Asthma is the most common chronic disease among children and its prevalence is rising. Asthma in children causes recurrent episodes of wheezing, cough, difficulty breathing, chest tightness and variable airflow obstruction. Airway hyper-responsiveness and airway inflammation are central components of the disease and may be precipitated by URTI, exercise, emotion and cold air. The severity of asthma can be defined by the level of treatment required to control symptoms (Table 1 shows the 2008 British Thoracic summary of the stepwise management of asthma in children). Brittle asthma is defined as wide Peak Expiratory Flow Rate (PEFR) variability despite intense therapy or sudden severe attacks on a background of apparently well-controlled asthma. It

BTS 2008 summary of the stepwise management of asthma in children

	Children less than 5 years	Children aged 5-12 years
1	Mild intermittent asthma	
	<ul style="list-style-type: none"> Inhaled short-acting β_2-agonist as required 	
2	Regular preventer therapy	
	<ul style="list-style-type: none"> Add inhaled steroid 200-400 $\mu\text{g}/\text{day}$ 	
3	Initial add-on therapy	Initial add-on therapy
	<ul style="list-style-type: none"> Leukotriene receptor antagonist 	<ul style="list-style-type: none"> Inhaled long acting β_2-agonist If inadequate: <ul style="list-style-type: none"> Trial of theophylline or leukotriene receptor antagonist Increase inhaled steroid to 400$\mu\text{g}/\text{day}$
4	Persistent poor control	Persistent poor control
	<ul style="list-style-type: none"> Refer to respiratory paediatrician 	<ul style="list-style-type: none"> Increase inhaled steroid up to 800 $\mu\text{g}/\text{day}$
5		Continuous of frequent use of oral steroids
		<ul style="list-style-type: none"> Maintain high dose inhaled steroids (800 $\mu\text{g}/\text{day}$) Use daily oral steroids at lowest possible dose Refer to respiratory paediatrician

Table 1

can be life threatening. Atopic conditions such as eczema and rhinitis increases the probability of asthma, as does parental smoking. Children with atopy have increased risk of severe allergic reactions. Any child with a history of reactive airways disease is at increased risk of perioperative bronchospasm.

Pre-operative assessment

Establish the severity of asthma and current status from the frequency of symptoms, current medication including steroids, emergency attendances, hospital and intensive care admissions, recent URTI, previous reactions to anaesthesia and tolerance of NSAIDs. Physical examination should assess the presence of tachypnoea, wheeze, crepitations, and increased work of breathing. In children >7 years, PEFr should be compared to baseline (or to that estimated by height) and reversibility with bronchodilators assessed.

Elective surgery should be deferred for 4–6 weeks if wheezing or URTI is present. Premedication with oral midazolam 0.5 mg/kg (maximum 20 mg) reduces anxiety-related bronchospasm. Inhaled or nebulized bronchodilators should be given before induction.

Anaesthetic management:

- For patients with severe asthma, avoid histamine-releasing drugs such as atracurium, thiopentone, morphine and suxamethonium. Propofol, vecuronium, rocuronium and fentanyl are preferable.
- Intubation should be avoided if possible. Ensure adequate depth of anaesthesia.
- Sevoflurane may have more bronchodilatory effects than isoflurane and desflurane.
- If the child has been on regular oral steroids or on high-dose inhaled steroids within the past two months, supplementary intravenous steroids need to be administered perioperatively.
- NSAIDs should only be avoided in children with severe or brittle asthma and in those with history of previous adverse reactions to NSAIDs.

Figure 1 describes the management of intraoperative bronchospasm. Means of giving nebulized/inhaled drugs during anaesthesia should be available. Any systemic involvement should alert to the possibility of anaphylaxis. Spontaneous pneumothorax needs to be excluded with acute deterioration of oxygenation during anaesthesia in asthmatic children.

Obstructive sleep apnoea

Obstructive sleep apnoea (OSA) is characterized by oxygen desaturation and reduced oro-nasal airflow despite preserved thoracic and abdominal respiratory effort. It occurs in 1–2% of children and symptoms include snoring, apnoeas, restless sleep, daytime hyperactivity, urinary incontinence and failure to thrive. The most common predisposing factors are adenotonsillar hypertrophy, neuromuscular disorders and craniofacial abnormalities. Children with OSA have more episodes of airway obstruction, breath holding and oxygen desaturation and they take longer to achieve appropriate depth of anaesthesia. They are at increased risk of post-operative cardiorespiratory complications. Pre-operative history, examination and nocturnal pulse oximetry should identify children at higher risk who will require further investigations such as nocturnal polysomnography and echocardiography to exclude right-heart dysfunction. Polysomnography is the electrographic monitoring of physiological variables during sleep. OSA is diagnosed if polysomnography shows:

- >1 obstructive apnoeas per hour of sleep
 - SpO₂ nadir $<90\%$
 - End-tidal CO₂ >50 mmHg for $>10\%$ total sleep time together with paradoxical respiration or snoring in children without lung disorders.
- Factors indicative of right-heart strain/cor pulmonale:
- ECG: right ventricular hypertrophy (RVH), right-axis deviation
 - CXR: RVH, right atrial enlargement, prominent pulmonary arteries.
 - Echo: right ventricular dilatation, tricuspid regurgitation.

syndrome (ACS), sepsis, aplastic anaemia and stroke. Neonates and infants do not manifest the effects of SCD until they are 4–6 months old due to the protective effects of foetal haemoglobin (HbF).

SCD is associated with significant perioperative morbidity and mortality. The heterozygote form sickle cell trait (HbAS) is usually asymptomatic and does not have any anaesthetic implications. Perioperative management must be considered individually by a multidisciplinary team of paediatricians, haematologists, anaesthetists and surgeons.

Anaesthetic management:

- Plan elective surgery weeks in advance
- Admit children before the day of surgery
- Pre-operative assessment – determine symptoms or signs of end-organ damage:
 - Respiratory disease from previous ACS, OSA
 - Cardiomegaly and diastolic dysfunction
 - Neurological deficits from previous CVAs
 - Renal impairment
 - Hepatic dysfunction
 - Chronic pain – analgesia requirements
- Postpone elective surgery if child is unwell
- Check pre-operative percentage of HbS, HbA and HbF
- Crossmatch blood (it might take up to 6 h if antibodies are present)
- Schedule children early in the operating list
- Avoid hyperviscosity (Hb > 12 or Hct > 35%)
- Hydrate aggressively:
 - Start IV maintenance fluids at least 12 h pre-operatively
 - Pay special attention to fluid balance and assessment of blood loss to avoid hypovolaemia
 - Continue IV fluids postoperatively until oral intake is fully established
- Anticipate difficult venous access
- Avoid hypoxia:
 - Perioperative and postoperative pulse oximetry monitoring
 - Give oxygen to maintain SpO₂ >92%
- Avoid acidosis: maintain normocarbica and normothermia
- Prevent infection by giving prophylactic antibiotics
- Provide effective analgesia, they may have high requirements and may be tolerant to opioids
- Avoid tourniquets unless absolutely necessary
- Postoperative nasal CPAP if severe OSA
- Encourage postoperative physiotherapy and early mobilization

Transfusion recommendations for elective surgery in children with sickle cell disease:

- Low-risk surgery in children who have no other risk factors: Hb \geq 7 g/dl
- Intermediate-risk surgery, OSA, recurrent chest problems or other chronic health problems: transfusion to a Hb of 11 g/dl, regardless of HbS levels
- High-risk surgery, previous CVA or ACS, eye surgery and use of tourniquets:
 - Transfuse or exchange transfuse to reduce the HbS <30%
 - Total Hb <11 g/dl

Postoperative serious sickling complications (usually within 48 h of surgery) include painful crisis, cerebro-vascular events and chest crisis. They should be managed with intravenous fluids, oxygen, antibiotics, analgesia, transfusion to Hb >10 g/dl but <12 g/dl in consultation with haematologists. Exchange transfusion is required in severe cases to reduce HbS to <20–30%. Ventilation may be required for acute chest crises.

Anaemia

The incidence in children of haemoglobin <10 g/dL is 0.5%; therefore, routine pre-operative haemoglobin testing should be targeted to children with specific indications such as systemic disease or in children undergoing major surgery. Anaemia is the commonest haematological abnormality in children. Causes vary with age and are often multifactorial.

Causes of anaemia in infants and children:

- Blood loss
 - Occult/internal bleeding
 - Iatrogenic
 - Hiatus hernia
 - Epistaxis
- Increase destruction
 - Rhesus/ABO incompatibility
 - Haemoglobinopathies
- Decreased production
 - Infection
 - Nutritional deficiency
 - Chronic disease
 - Bone marrow depression
- Bleeding disorders
 - Haemorrhagic disease of the newborn
 - Haemophilia
 - Christmas disease

In preterm infants, blood loss from multiple sampling is the most frequent cause and is exacerbated by reduced red cell lifespan and low erythropoietin production. In term newborns, anaemia is usually caused by occult blood loss around the time of delivery (feto-maternal/feto-placental haemorrhage or twin-to-twin transfusion). Neonates and ex-preterm infants (up to 60 weeks post-conceptual age) with haematocrit of <30 are at higher risk of postoperative apnoea. In infants and children, nutritional deficiencies are the commonest cause for anaemia and iron deficiency is the most frequent.

Guidelines for blood transfusion for infants and children:

- First 24 h and neonates in intensive care if Hb <12 g/dL (Hct = 0.36)
 - Chronic oxygen dependency if Hb <11 g/dL
 - Hb \leq 7 g/dL in patients where there is no chronic anaemia/illness (e.g. renal disease)
 - Symptomatic patients
- Ideally, there should be a two-day wait prior to anaesthesia and surgery post-transfusion.

Obesity

Obesity is defined as body mass index (BMI = weight/height²) >30. Because BMI in childhood changes substantially with age

and gender, international cut-off points have been developed for children between 2 and 18 years of age. Obesity in children is usually due to sedentary life style and poor diet; rarely, it is secondary to specific syndromes such as Prader–Willi and Lawrence–Moon–Biedle. There is evidence that obese children have a higher incidence of difficult mask ventilation, airway obstruction, bronchospasm, major oxygen desaturation and overall critical respiratory events during anaesthesia.

Clinical features

- Associated co-morbidities:
 - Asthma
 - Recurrent respiratory infections
 - OSA
 - Gastro-oesophageal reflux (GOR)
 - Diabetes Mellitus
 - Hypertension
- Pre-operative anxiety
- Difficult venous access
- Altered drug pharmacokinetics
- Increased risk of postoperative thromboembolism (TE)
- Increased incidence of respiratory complications

Anaesthetic considerations

- Anticipate the need for HDU/PICU postoperatively
- If sedative premedication is given, oxygen saturation must be monitored
- If GOR, give pre-operative antacids and consider rapid sequence induction
- Opioids (including PCA), propofol, midazolam and muscle relaxants dosing should be based on ideal bodyweight
- Use total bodyweight for suxamethonium
- Total intravenous anaesthesia (TIVA) based on corrected weight (ideal weight + $[0.4 \times \text{excess weight}]$)
- Use correct blood pressure cuff size
- Care with handling and positioning, induce on operating table
- Use of positive end expiratory pressure (PEEP) to optimize oxygenation
- Use regional and local anaesthetic techniques to minimize opioids side effects
- Monitor neuromuscular blockade
- Extubate awake and sitting up
- Monitor oxygen saturation continuously and give supplemental oxygen for at least 24 h
- If severe OSA consider postoperative CPAP
- Early mobilization and physiotherapy to reduce risk of respiratory complications and TE
- TE prophylaxis:
 - Compression stocking
 - Pneumatic compression devices
 - Subcutaneous heparin

Diabetes mellitus

Diabetes mellitus (DM) is a chronic metabolic disorder that affects 1 in 500 children. 90% of diabetic children have type 1 but there has been an increase in the incidence of type 2 diabetes especially in affluent societies. Childhood diabetes is managed with complex insulin regimes that mimic the natural pancreatic endocrine

function and help to minimize end-organ damage later in life. The aim of safe perioperative management of diabetic children is to maintain blood glucose levels as close to normal as possible (Table 3). The child should be scheduled first on the operating list; routine starvation times must be followed and close liaison with the diabetes team is essential. Emergency surgery is associated with large fluctuations in blood glucose so insulin/glucose infusion should be started early. Fluid resuscitation, correction of hyperglycaemia and electrolyte imbalance are essential before emergency surgery. There is a risk of diabetic ketoacidosis (DKA) and DKA might be present with abdominal symptoms.

Long-term steroids

Children on large doses of long-term steroids (including high doses of inhaled steroids) need perioperative steroid supplementation to prevent the effects of suppression of the hypothalamic–pituitary–adrenal axis. Side effects of long-term steroid use include: adrenal suppression; immunosuppression; growth retardation; moon face; sodium and water retention; hypertension; obesity; Diabetes mellitus; hyperlipidemia; potassium loss; gastritis and increased risk of gastric and duodenal ulcers; osteoporosis and avascular necrosis of bone; myopathy; skin thinning and capillary fragility; behavioural changes and psychosis. Children taking regular steroids or those who have stopped steroids within two months should receive hydrocortisone 1 mg/kg (IV) at induction and six-hourly postoperatively until oral steroids are resumed.

Renal failure

Renal dysplasia and obstructive uropathy are the most common causes of renal failure in children under five years, whilst glomerular disorders become more prevalent in older children. Systemic effects of chronic renal failure in children and anaesthetic implications are described in Table 4. Most children are managed medically but dialysis is required for children with end stage renal failure and to treat major problems like fluid overload, hypertension and electrolyte or acid/base abnormalities. Dialysis should be done 12–18 h pre-operatively and U&Es, FBC and coagulation checked prior to surgery.

Indications for pre-operative haemodialysis in children include:

- Uraemia
- Hypervolaemia
- Electrolyte disturbance
- Metabolic acidosis
- Presence of exogenous/endogenous dialyzable toxins

Cerebral palsy

Cerebral palsy (CP) is a spectrum of clinical syndromes caused by pre-, peri- or post-natal central nervous system damage. Abnormal control of motor function by the brain leads to a non-progressive but changing disorder of movement and posture, which may be spastic, ataxic or dyskinetic. In addition to motor deficit, these children often suffer with cognitive impairment, behavioural disturbance and hearing, speech and visual disabilities. Epilepsy is common and bulbar involvement may lead to feeding difficulties, malnutrition, GOR, aspiration and

Perioperative management of diabetic children

Morning list

Minor surgery^a

Pre-operatively

- Omit breakfast
- Omit morning insulin/oral hypoglycaemics
- Avoid prolonged starvation
- Check blood glucose hourly

Intraoperatively

- IV fluids
- Check blood glucose half hourly

Postoperatively

- Give breakfast and short-acting insulin (*1/3rd of total morning dose*)
- Check blood glucose hourly for 4 h
- Discharge if well 2 h post-op
- Admit if nausea and vomiting

Major surgery

Pre-operatively

- Admit the child the day before surgery
- Check blood glucose and electrolytes
- Omit breakfast
- Omit morning insulin
- Check blood glucose hourly

Intraoperative

- IV fluids
- Check blood glucose half hourly
- Start insulin infusion^b

Postoperatively

- Continue insulin infusion^b
- Check blood glucose hourly for 4 h then 2 hourly if stable
- Continue routine maintenance IV fluids^c
- Re-start usual insulin regime when eating and drinking normally

^a Procedure duration <30 min, low risk of post-operative nausea and vomiting, and the child is expected to eat and drink very soon after surgery.

^b Insulin infusion 50 units *Human Actrapid* short-acting insulin in 50 ml 0.9% Saline (insulin 1 unit/ml) infuse at 0.05 units/kg/h via a syringe pump. Adjust according to the blood glucose level. Additional fluids should be given routinely.

^c IV Maintenance fluids usually 5% glucose/0.9% Saline + K⁺ supplements as appropriate. Depending on blood glucose.

Afternoon list

Minor surgery^a

Pre-operatively

- Normal breakfast before 7:30 am
- Short-acting insulin only (*1/3rd of total morning dose*)
- Clear fluids until 2–3 h before surgery
- Check blood glucose hourly

Intraoperatively

- IV fluids
- Check blood glucose half hourly

Postoperatively

- Give lunch and short-acting insulin (*1/3rd of total morning dose*)
- Check blood glucose hourly for 4 h
- Discharge if well 2 h post-op
- Admit if nausea and vomiting

Major surgery^b

Pre-operatively

- Normal breakfast before 7:30 am
- Short-acting insulin only (*1/3rd of total morning dose*)
- Clear fluids until 2–3 h before surgery
- Check blood glucose hourly
- Start IV fluids and insulin infusion at 11 am

Intraoperatively

- IV fluids
- Check blood glucose half hourly

Table 3

pneumonia. Children with spastic CP develop muscle contractures and secondary bony deformities. Physiotherapy, splints, surgery, oral or intrathecal baclofen and botulin toxin injections are useful in treating spasticity. Children with CP commonly undergo repeated orthopaedic procedures. The risk of post-operative respiratory complications is greater in more dependent children in whom anaesthesia and surgery pose a greater stress to their cardiovascular and respiratory systems.

Anaesthetic management

Pre-operatively

- Do not assume cognitive impairment
- Repeat attendees: review previous anaesthetic charts and question children and parents.

- Continue antiepileptic, reflux and antispasticity medication.
- Sedative premedication is useful if anxious or phobic.
- Latex allergy is more common.
- Vascular access is often difficult
- Antacid prophylaxis and rapid sequence induction is indicated with significant GOR.

Intra-operatively

- Resistance to neuromuscular blocking drugs has been reported but sensitivity to suxamethonium does not occur.
- Reduced minimum alveolar concentration of 20% (30% in children taking anticonvulsants)
- Pre-operative fluid deficit commonly underestimated.

Systemic effects and anaesthetic implications in children with chronic renal failure

Systemic effects of chronic renal failure

Cardiovascular

- Left ventricular hypertrophy and diastolic dysfunction
- Hypertension
- Risk of dysrhythmias

Respiratory

- Recurrent infections
- Basal atelectasis and pleural effusion with peritoneal dialysis

Anaemia

- Reduced erythropoietin
- Reduced red cell survival
- Iron and folate deficiency

Coagulopathy

- Thrombocytopenia
- Platelet dysfunction
- Increased capillary fragility

Fluid and electrolyte abnormalities

- Fluid overload
- Hypovolaemia: post dialysis
- Hyperkalaemia
- Hypocalcaemia
- Hyper/Hypo phosphataemia
- Hypo/hypernatraemia

Acid/base abnormalities

- Metabolic acidosis
- Low plasma bicarbonate

Vascular access

- Tunnelled catheters
- Shunts

Neurological

- Peripheral neuropathy
- Encephalopathy
- Seizures

Growth and development delay

Gastrointestinal

- GOR in 70%
- Uraemia results in nausea and vomiting
- Higher prevalence of gastric ulceration

Drug doses

Anaesthetic implications

- Continue normal antihypertensive medication perioperatively and monitor blood pressure.
- Ventilate
- Hb > 6 g/dl is acceptable for surgery associated with minimal blood loss if the child is asymptomatic. For all other surgery keep Hb >8 g/d
- The risks and benefits of central neuroaxial blocks should be weighed carefully
- Avoid NSAIDs
- Dialysis should be scheduled pre and post-op
- K⁺ should be <5.0 mmol/L
- Avoid K⁺ containing solutions
- Know child's daily fluid allowance
- Suxamethonium is safe if K⁺ < 5.5 mmol/L
- Strict asepsis when using tunnelled lines
- Avoid IV access in potential shunt sites
- Avoid NIBP cuffs in same limb as shunts
- Antacids
- Consider rapid sequence induction
- Reduce thiopentone and propofol
- Atracurium, vecuronium and rocuronium safe
- Morphine may accumulate
- Increase PCA lockout periods or consider alternatives (fentanyl or hydromorphone)
- Use regional techniques for analgesia
- Local Anaesthetic duration of action may be reduced
- Avoid Pethidine

Table 4

- Temperature regulation is often abnormal in CP so warming and monitoring of temperature is essential.
- CP children are at increased risk of pressure sores, they should be positioned with care on the operating table and all pressure areas must be padded.
- Multimodal analgesia with paracetamol, NSAIDs and epidural or morphine patient/nurse controlled analgesia (PCA/NCA) infusions is recommended.
- They are at high risk of opioid side effects with more than half experiencing postoperative nausea and vomiting.

Post-operatively

- Pain from muscle spasms is common:
 - It is best prevented by using epidural analgesia + clonidine
 - Treat with regular diazepam (0.1 mg/kg PO/IV/PR 6-hourly).
 - Midazolam infusions are an alternative for severe cases but require admission to a high-dependency unit.
- Involve carers at all times to help reduce anxiety and assess pain.

Epilepsy

In children with well-controlled epilepsy having non-neurosurgical operations, seizure control is rarely a problem perioperatively. However, anaesthesia, surgery, starvation and certain drugs can lower the seizure threshold. Pre-operative assessment should include history of the last fit, frequency, precipitants, type of seizures, presence of learning and behavioural difficulties and regular medication. Seizure disorders are sometimes associated with other conditions that may have anaesthetic implications such as CP, Down's syndrome, Angelman syndrome, Sturge–Weber syndrome, Di George syndrome, Tuberous-sclerosis, Neurofibromatosis, Hemihypertrophy and Lesch–Nyhan syndrome. Regular antiepileptics should be continued until and including the day of surgery and re-started early postoperatively, if oral intake is not restored postoperatively, alternative anti-convulsant medications must be prescribed after discussion with the child's neurologist. Some advocate the use of thiopentone for induction to reduce the seizure threshold postoperatively. Common anticonvulsants have liver-enzyme-inducing effects and children on chronic treatment may be resistant to opioids and non-depolarizing muscle relaxants (e.g. carbamazepine and phenytoin produce a 50% reduction in the duration of action of vecuronium).

Autism

Autism is a lifelong developmental disability apparent within the first 2.5 years of life. The unifying symptoms are difficulty with communication and social interaction. For these children, any change in their routine can be very distressing and coming to hospital may cause panic attacks or temper tantrums, which are extremely difficult to manage. Most autistic children are physiologically healthy and tolerate general anaesthesia very well.

Anaesthetic implications:

- Contact the parents in advance to make an individualized plan.
- Arrange for the child to stay in a side room and schedule first on the operating list.
- Consider giving sedative premedication orally in the child's favourite drink (oral or buccal midazolam 0.5 mg/kg maximum 20 mg, ± ketamine 3 mg/kg). Discuss with parents the possibility of using restraint and anticipate the need for help to lift the child on to the trolley.
- Give routine IV fluids and antiemetics to prevent PONV.
- Remove cannula as soon as possible or secure it well.
- Allow the child to wake in the parent's presence and plan for early return home as soon as it is safe.

Trisomy 21 (down syndrome)

Down syndrome is a chromosomal disorder with an incidence of 1–2 per 1000 live births. It involves all body systems and its main clinical features include:

- Microcephaly
- Macroglossia
- Obstructive sleep apnoea (50%)
- Recurrent chest infections
- Congenital subglottic stenosis
- Atlanto-axial instability
- Hypotonia and hyperflexible joints
- Congenital heart disease (50%):
- Developmental delay
- Immunosuppression
- Hypothyroidism

Anaesthetic implications:

- Exclude associated anomalies
- May be uncooperative with induction:
 - Plan with the parents
 - Consider premedication
- Potentially difficult airway although rarely difficult intubation
- Pre-operative cervical X-ray in both flexion and extension only indicated if neurological symptoms but avoid extension of the neck during intubation.
- Post-extubation stridor is common, select smaller size tracheal tube, ensure a leak is present and give Dexamethasone (0.1–0.25 mg/kg) to decrease laryngeal oedema.
- Increased sensitivity to volatile anaesthetic agents and muscle relaxants
- Consider HDU if significant OSA
- Increased risk of postoperative chest infections

Oncological disease

Children with malignant disease require repeated general anaesthesia for diagnostic and therapeutic procedures.

Anaesthetic implications:

- These children commonly have tunnelled central venous catheters for long-term IV access.
 - Careful aseptic technique or non-touch technique should be used to prevent infection.
 - Any channel that is used should be flushed with 10 ml of saline and locked with 10 ml of heparin-saline.
- Chemotherapy side effects include nephrotoxicity (cisplatin), liver damage (especially asparaginase and nitrosureas, methotrexate, cytarabine, cisplatin, cyclophosphamide in high doses), cardiomyopathy (doxorubicin and daunorubicin), pulmonary toxicity and bone marrow suppression.
- Bleomycin can cause pulmonary fibrosis and impaired lung function.
- Massive hepato-splenomegaly can compromise breathing.
- Assess and correct pancytopenia and clotting abnormalities
- Blood products must be irradiated/leucocyte depleted to prevent graft-versus-host disease and CMV negative if bone marrow transplant.

- Paracetamol and NSAIDs mask pyrexia of infection and are often avoided.
- NSAIDs avoided due to potential nephrotoxicity.
- Avoid PR and IM medications

Dexamethasone and tumour lysis syndrome: Dexamethasone must not be given as an antiemetic as it can cause massive tumour lysis syndrome particularly in children with lymphoma. It requires urgent treatment with allopurinol and hyperhydration to prevent life-threatening complications such as renal failure.

Mediastinal masses: tumour masses in the anterior mediastinum can cause airway or superior vena cava obstruction.

Careful pre-operative assessment must include:

- Search for stridor or other signs of respiratory compromise, including any worsening of symptoms on changing of position.
- Review of X-rays/MRI/CT to ascertain extent of mass.

Anaesthetic management:

- Inhalational induction sitting up with maintenance of spontaneous ventilation is the technique of choice.
- Airway obstruction after induction of anaesthesia is due to compression of the trachea or main bronchi with the loss of compensating muscle tone:
 - A change in position might be helpful
 - Rigid bronchoscope should be immediately available to provide a temporary airway.

Latex allergy

Exposure to latex can cause life-threatening anaphylaxis from type I IgE mediated hypersensitivity reactions or more commonly an allergic contact dermatitis (type IV). Testing for type I natural rubber latex allergy is through blood testing, such as RAST (radioallergosorbent test) which identifies what types of IgE proteins trigger allergic reactions. Currently the only effective preventative strategy is latex avoidance.

Children at high risk for serious reactions:

- History of anaphylaxis to latex
- History of allergy to latex or rubber (Urticaria, dermatitis, eye swelling, bronchospasm...)
- History of multiple surgical procedures:
 - Spina bifida.
 - Genitourinary anomalies.

Children at risk for developing latex allergy

- Allergy to bananas, kiwifruits, avocado, stone fruits, raw potato, tomato, papaya, chestnuts or peanuts
- History of a latex glove-associated contact dermatitis

Premedication

There is no evidence that premedication with an anti-histamine or steroids is useful.

Management in Theatre

- Patient should be first on the list.
- Latex free theatre for at least two hours (ideally overnight) prior to case.
- All staff involved in patient care must be made aware of latex allergy status and warning signs should be posted outside the operating theatre.
- A latex free trolley/box should be present with every case which follows the patient from pre-assessment to post-operative care.
- Develop completely latex free surgical areas.

Pregnancy

Adolescent girls may be pregnant when presenting for surgery. Pre-operative testing should be offered to all post-menarche girls. Elective surgery should not be performed at all during pregnancy. Emergency surgery must proceed regardless of gestational age as the primary goal is to preserve the life of the mother.

Gastro-oesophageal reflux (GOR)

Gastro-oesophageal (GOR) may occur in children with hiatus hernia, diabetes mellitus, renal failure, sepsis, and is common in children with neurological impairment (e.g. CP). It may lead to recurrent aspiration and chest infections, weight loss and anaemia. The risk from reflux related to paediatric anaesthesia is uncertain. All anti-reflux medication should be continued up to, and including the day of surgery. Rapid sequence induction should be performed in the presence of GOR symptoms. ◆

FURTHER READING

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