Medical assessment of the paediatric patient

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Careful preoperative assessment is the cornerstone of safe anaesthetic practice as it allows for optimum planning of the child’s anaesthetic and perioperative care. Assessment encompasses evaluation of the child’s present health, past medical and anaesthetic history, and review of relevant investigations. These factors are then integrated with the anticipated effects of surgery to allow planning of an appropriate anaesthetic. The importance of preoperative assessment has been highlighted in the recent publication The Anaesthesia Team which states that all patients should pass through a pre-admission routine and that although many aspects of the assessment can be delegated to members of the team, ultimately the anaesthetist is responsible for deciding that the patient is optimally prepared.2 The medical assessment of the patient is only part of this process.

Anaesthetists are constantly under time pressure to evaluate patients efficiently. There is a trend to expect the anaesthetist to become more of a ‘perioperative medical specialist’ so ensuring that all medical issues which relate to the safe provision of anaesthesia are addressed.20 64 The role of the preoperative assessment clinic is becoming increasingly important, particularly in the preparation of the patient admitted on the day of surgery. While most work has been reported from adult practice, paediatric patients are increasingly being offered the facilities of preoperative assessment clinics. Inadequate preoperative assessment may result in an increase in late cancellation of patients from the theatre list and careful preoperative assessment encourages considered preoperative testing that is worthwhile and effective.2 21 This decreases unnecessary investigations which can be unpleasant for the child and result in unnecessary cost and inconvenience to the hospital service. Routine preoperative investigations, such as measurement of haemoglobin concentration or urinalysis, are not necessary in healthy children.54 58 Focusing investigations at specific patient groups who have particular medical requirements or surgical needs is much more worthwhile.

Review of the child’s present and past medical history, and previous anaesthetic records is essential for a useful medical assessment to be completed. Information on current medication, history of allergies and family history of anaesthetic problems should be collected. A systematic approach is important. Factors such as birth and neonatal histories are very important in the young child. Evaluation of the cardiac and respiratory systems and the airway is initially required, with further examination indicated according to clinical requirements. The majority of children admitted for surgery are healthy, ASA I or II, and are undergoing relatively minor surgery. Complex paediatric syndromes and congenital malformations, although often rare, may have important anaesthetic implications; these are reviewed in the specialist reference books and are outside the remit of this article.37 50 Liaison with specialist paediatric medical teams is frequently essential. It is necessary to evaluate the risks and benefits associated with the procedure requiring general anaesthesia and any potential risk factors associated with anaesthesia so that these are discussed with the parents.

This article discusses selected conditions, some of which occur commonly, such as asthma, diabetes and upper respiratory tract infection. Others we are meeting with increasing frequency because of the successes in paediatric care. This includes children with cardiac disease, those who have had transplants and those with neonatal problems.

Cardiac disease

There are two groups to consider: those known to have cardiac disease and those with symptoms or signs suggesting undiagnosed cardiac disease. Congenital heart disease (CHD) occurs in approximately 0.8% of live births. Fifteen percent of these children also have an extracardiac abnormality and many need investigations or surgery requiring anaesthesia. The possibility of a cardiac lesion should always be considered in any condition which is known to have such an association, for example tracheo-oesophageal fistula, oesophageal atresia, Down’s syndrome and VATER association.37 50 Significant cardiac disease is usually symptomatic in early life with most CHD identified before 3 months of age.

Successful corrective or palliative surgery is now more frequently undertaken early, and this results in increasing numbers of children presenting for subsequent non-cardiac surgery. When cardiac surgery results in a structurally or
functionally normal heart, routine anaesthetic management is appropriate. After more complex repairs, anaesthetic considerations need to be delicately balanced, and some children, such as those with a single ventricle physiology, should be cared for in a centre with cardiological support services. Children who have had corrective or palliative cardiac surgery and are well compensated do not have an increased mortality during subsequent general surgery. There is an increased risk of mortality, particularly after emergency surgery, in babies less than 6 months old who remain physiologically compromised. Mortality related specifically to anaesthesia in children undergoing corrective cardiac surgery is rare and the risks can be related to the ASA status of the child, as with other surgery.

CHD has been classified in many ways and may be very varied and complex. The pathophysiology may fall into several categories: shunts, mixing, obstruction and valvular regurgitation, with complex anomalies having a combination of these components.

The likely effect of a specific cardiac lesion needs to be assessed. Lesions such as atrial and ventricular septal defects (ASD, VSD), and endocardial cushion defects result in normal or overperfusion of the lungs and are therefore usually acyanotic. Excessive blood flow to the lungs, with left to right shunting of blood, results in ventricular volume overload. This produces early signs of congestive cardiac failure as volume load is less well tolerated than pressure load. Unexplained cyanosis always requires further investigation and in a neonate this is urgent. Lesions that result in underperfusion of the lungs because of right to left shunting such as tetralogy of Fallot, result in cyanosis. Conditions with complex shunts, for example transposition of the great arteries, are also usually associated with cyanosis. Obstructive lesions, including coarctation of the aorta, aortic or pulmonary valvular stenosis, or interrupted aortic arch, result in ventricular pressure overload which the paediatric myocardium is more able to accommodate for longer, with fewer symptoms, than an increased volume load.

**Clinical features**

Neonates and young infants with CHD usually present with features of congestive cardiac failure, including tachypnoea, sweating or hepatomegaly. Other features may include cyanosis, increasing polycythaemia, history of recurrent respiratory infections, wheeze or a generalized pattern of the child failing to thrive. Identification of a murmur on routine neonatal review may be the first sign of CHD.

Neonates with coarctation of the aorta, aortic stenosis, hypoplastic left heart syndrome, or interruption of the aortic arch or pulmonary atresia have a circulation which is dependent on maintenance of the patent ductus arteriosus (PDA) to provide pulmonary blood flow. Prostaglandin E1 is used to prevent closure of the duct. These babies may be very sick, with features of multisystem failure and need a period of stabilization in intensive care. Medical assessment focuses on end-organ function, with renal impairment and disseminated intravascular coagulopathy being common.

Clinical features of CHD outside infancy may be more apparent. These include the development of cyanosis, clubbing and oedema. Respiratory effects may include wheeze, decreased exercise tolerance or breathlessness. Children with CHD are at increased risk of respiratory infections. Gastrointestinal symptoms, including poor appetite, failure to gain weight and hepatosplenomegaly, may occur. Some children with congestive cardiac failure have a persistent low grade pyrexia without any identifiable infective source. Increasing symptomatology indicates poorly controlled disease.

Antibiotic prophylaxis, for the prevention of endocarditis, is required for all invasive procedures, but local procedures vary. Prophylaxis is not needed routinely for children who have had ligation of a PDA, ASD or routine cardiac catheterization.

**Pulmonary hypertension (PHT)**

PHT is rare and the condition is usually secondary to congenital heart disease, chronic airways disease, upper airway obstruction, adenotonsillar hypertrophy, cystic fibrosis, bronchopulmonary dysplasia or neuromuscular disorders, such as muscular dystrophy. Eisenmenger’s syndrome can result when long-standing left to right intracardiac shunt, such as is present in uncorrected VSD, ASD, PDA and AVSD, results in increasing PHT until the shunt reverses producing a right to left shunt and cyanosis. Symptoms of PHT include breathlessness, particularly on exercise, syncope and later, cyanosis, cough and haemoptysis; sudden death may occur. Signs of right ventricular hypertrophy include characteristic features on chest x-ray and on the ECG. Serial ECG and echocardiography provide a measurement of the progressive nature of the illness. Polycythaemia is a late feature. Children with primary PHT may have few symptoms.

Anaesthetic management requires balancing the anaesthetic techniques and therapeutic agents which have an effect on pulmonary vascular resistance (PVR). The degree of reversibility of PHT is assessed at angiography; this information may be important when planning which agents may be required during anaesthesia.

**The innocent murmur**

Detection of a murmur on routine preoperative assessment is common. Innocent murmurs have been reported in 8–80% of children. It is important to distinguish between innocent and pathological murmurs, but this can be difficult. The respective features are summarized in Table 1.

In the asymptomatic child with a murmur, the two conditions which need to be excluded are hypertrophic obstructive cardiomyopathy and critical aortic stenosis. An ECG shows left ventricular hypertrophy (RV6+SV1 >5 mV) and left axis deviation in both of these conditions.
Table 1 Features of cardiac murmurs

<table>
<thead>
<tr>
<th>Innocent</th>
<th>Pathological</th>
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<tr>
<td>Asymptomatic</td>
<td>Symptomatic</td>
</tr>
<tr>
<td>Soft</td>
<td>Loud</td>
</tr>
<tr>
<td>Early systolic</td>
<td>Pan or late systolic diastolic continuous</td>
</tr>
<tr>
<td>No thrill</td>
<td>Thrill</td>
</tr>
<tr>
<td>Disappears with positioning</td>
<td></td>
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<tr>
<td>May be a venous hum</td>
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Children who have murmurs with features of an innocent nature, who have no signs or symptoms of cardiac disease and in whom an ECG and possibly echocardiography have been reviewed to ensure there are no signs of ventricular hypertrophy, can be anaesthetized safely and referred for cardiological review later (Fig. 1). Some centres use antibiotic prophylaxis routinely in this group. All infants, and those children whose murmurs show pathological features, should be reviewed by a paediatric cardiologist before anaesthesia.52

**Anaesthetic assessment**

Elective non-cardiac surgery must take place when both the child’s general health and their cardiac status are optimal. Assessment initially involves understanding the particular pathophysiology involved. The basic information required includes details of the type of cardiac repair performed and the likely sequelae of the repair. Consideration must be given to the present physical condition of the child and the current medical treatment, particularly anti-failure therapy, β-blockers, anticoagulants, antihypertensive medication, digoxin or aspirin. Blood concentrations of digoxin may need to be checked. If a pacemaker is present, its routine preoperative evaluation is required.

Children with a significant cardiac history should be reviewed before operation by a paediatric cardiologist and those with signs of cardiac failure or who have arrhythmias need to be reviewed by the paediatric team and their medical condition optimized before surgery. If the child is having regular, infrequent reviews by the paediatric team, the most recent report of a stable situation will provide sufficient information.

Children with cardiac disease, undergoing non-cardiac surgery, routinely have an ECG, precordial Doppler echocardiography, chest x-ray, and laboratory tests, including full blood count, urea and electrolytes, creatinine or coagulation studies, as indicated. In addition, some children require review of their angiography results. The results of these investigations taken together allow assessment of their current cardiac status.

The chest x-ray provides an indication of heart size, degree of pulmonary vascularity and the presence of intrapulmonary pathology, such as infection. An ECG is reviewed, noting particularly the presence of arrhythmia, and evidence of ventricular hypertrophy. Some cardiac repairs, such as repair of tetralogy of Fallot, Mustard, Senning or Fontan repairs, are associated with a high incidence of arrhythmias. Echocardiography is one of the most useful investigations in the child with CHD. Echocardiography defines cardiac structure and function.

Transoesophageal echocardiography is being used increasingly in the assessment of paediatric patients, particularly in adolescents in whom a better quality study can be achieved with this method. However, it is invasive, has an incidence of complications and requires general anaesthesia or heavy sedation.35

Angiography is used more selectively in the assessment of the cardiac patient as improvements in echocardiography have allowed sufficient information to be gathered less invasively. It remains particularly useful in determining the coronary anatomy, for assessing multifocal pulmonary blood flow and for measuring specific haemodynamic information, such as pulmonary artery pressures.

A baseline $S_{a}O_{2}$ is useful before anaesthesia as significant cyanosis indicates inadequate pulmonary blood flow, right to left shunting or mixing. In a child with tetralogy of Fallot, the presence and frequency of cyanotic spells are checked and their severity and routine management noted. Long-term cyanosis can be complicated by polycythaemia,
hyperviscosity syndrome or coagulopathy. This may result in decreased cardiac function, cerebrovascular occlusive episodes, cerebral abscesses or thromboembolism. Occasionally, if surgery cannot improve pulmonary blood flow, the child may require regular venesection to decrease morbidity from polycythaemia. Venesection just before anaesthesia should be avoided as it may be accompanied by cardiovascular instability. Polycythaemic children must be kept well hydrated and fluid restriction minimized. Children with CHD can safely continue receiving clear fluids orally up until 2–3 h before operation.\textsuperscript{56} If a longer preoperative fast is anticipated, preoperative i.v. fluids should be given.

\textbf{Respiratory disorders}

\textit{Neonates and infants}

The success of neonatal care has meant that increasing numbers of babies, who may have complicated neonatal histories, require surgery for a wide variety of conditions. Some present additional problems during anaesthesia and many have an increased risk of postoperative complications. The group most frequently studied are babies undergoing repair of inguinal hernia, a condition which is much more common in the preterm neonate. All babies require a careful assessment of their medical history with specific attention to their birth history, gestational age at delivery and age at surgery, which is usually expressed as post-conceptual age (PCA). Preterm delivery, defined as delivery at less than 37 weeks’ post-conceptual age, is known to be associated with an additional risk of postoperative complications, particularly apnoea and bradycardia.

The baby’s present physical condition is determined, with particular emphasis on a history of prolonged intensive neonatal care, degree of respiratory reserve, possibility of subglottic stenosis, presence of congenital abnormalities, likelihood of anaemia and information on provision of vitamin K prophylaxis for prevention of haemorrhagic disease of the newborn. A history of apnoea and bradycardic episodes is important as is a history of a continuing or recent additional oxygen requirements, which indicates that the baby may require additional, short-term, respiratory support after operation.

\textit{Apnoea in neonates}

Assessment of respiratory function is reliant on the history and clinical features. Normal newborn babies, born prematurely, are known to have apnoeas of central, obstructive or mixed type. The commonest type of apnoea is that of a central aetiology, but an obstructive or mixed pattern of apnoea is associated with more severe episodes of hypoxaemia.\textsuperscript{46} Apnoeas are frequently accompanied by bradycardia. A sleep study can identify the type of apnoea but it is not a sensitive test nor very useful as a preoperative assessment tool.\textsuperscript{45, 47} Premature babies with no history of apnoea are still at risk of apnoeas after surgery and anaesthesia. Quantifying the risk is difficult and monitoring this group with an apnoea monitor and oxygen saturation monitor is essential after operation. The age at which the ex-preterm baby is no longer at risk has been debated. This question has implications for the management of young babies for minor surgery on a day-case basis. Coté and colleagues attempted to quantify the actual risk of apnoea and reported a combined analysis of eight previously published studies. They concluded there was at least a 5% risk of apnoea in a neonate at 48 weeks PCA if the child was born at 35 weeks and that this same group had a <1% risk of apnoea at a PCA of 54 weeks.\textsuperscript{11} The risk increased as PCA decreased. Identified risk factors included gestational age, PCA, history of apnoeas, occurrence of apnoeas in the recovery room and anaemia.\textsuperscript{11} Administration of caffeine may be successful in preventing apnoeas in this group of patients.\textsuperscript{79} Spinal anaesthesia may be associated with a lower incidence of postoperative complications, including apnoea, hypoxia and bradycardia.\textsuperscript{28, 45} However, it is useful for only a limited number of surgical procedures. Complications occur with spinal anaesthesia, particularly inadequate anaesthesia and high spinal block.\textsuperscript{24} If sedation or even ‘light’ anaesthesia is used in addition to the spinal anaesthetic, the incidence of apnoea is not decreased.\textsuperscript{81} Postoperative apnoea can occur up to 48 h after surgery\textsuperscript{47} and there is no consensus as to when the risk becomes negligible.\textsuperscript{22, 45, 78} Apnoeas have been reported in term babies after anaesthesia but are rare and may be related to other medical factors.\textsuperscript{1, 68}

A full blood count is routinely carried out on all neonates and preterm infants as it is difficult to identify anaemia clinically in this age group and it is associated with an increased risk of postoperative apnoeas in former preterm babies.\textsuperscript{80} Haemoglobin concentration is related to the degree of maternal transfusion at birth. The normal haemoglobin concentration in the premature neonate is 18 g dl\textsuperscript{–1} decreasing to 17 g dl\textsuperscript{–1} in the term baby and to 10 g dl\textsuperscript{–1} by 6 months of age. Babies who have prolonged hospital stays are frequently anaemic because of repeated blood test requirements. Cardiorespiratory reserve is very limited in these babies and preoperative correction of anaemia when present may be required.

\textit{Bronchopulmonary dysplasia (BPD)/chronic lung disease}

BPD occurs as a result of respiratory distress syndrome which is associated with preterm delivery and the effects of mechanical ventilation. Features of this condition include signs of respiratory compromise related to hyperinflation, development of emphysema or bullae, risk of pneumothorax, increased reactivity of the airways and increased risk of respiratory infection (Fig. 2). These babies have poor pulmonary compliance as a result of development of interstitial fibrosis, increased airway resistance and fluid retention. They often remain oxygen dependent, and diuretics...
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Fig 2 X-ray showing chronic lung disease, hyperinflation and patchy infiltration.

and steroids are frequently required. A preoperative chest x-ray is useful and blood-gas analysis may indicate the severity of disease if carbon dioxide retention is present, with or without mild hypoxia. Spinal anaesthesia has been advocated for herna repair in this group of patients. Babies who have been intubated may have subglottic stenosis and this may become apparent during or after anaesthesia, either when a relatively small tracheal tube is used in relation to the size and age of the child or if stridor develops in the postoperative period. The use of atropine as a premedicant is valuable in these babies as it decreases both airway reflexes at laryngoscopy and the likelihood of bradycardia. This group of patients may benefit from i.m. atropine premedication as its effect is more reliable than when administered orally. They may be more likely to have gaseous induction as it is not always easy to achieve vascular access.

Children

Asthma

Mild asthma featuring episodes of wheezing is extremely common in young children and easily managed with bronchodilators. Asthma tends to occur in children with a history of atopy. There is some evidence that the incidence of this condition is increasing, possibly because of environmental factors. The majority of children outgrow this type of mild asthma. Severe symptoms occur in less than 10% of children with asthma and require much more aggressive treatment. Guidelines for the management of childhood asthma have recently been published. Repeated hospital admissions may be required to maximize therapy and intensive care management is occasionally necessary. This pattern of asthma, although much less frequent, can be very difficult to manage.

A careful history is of most value in determining the type, frequency and severity of asthma in an individual child. It is unwise to undertake elective surgery within 4 weeks of a major exacerbation of asthma. Anaesthesia with tracheal intubation, following recent exacerbation of asthma, is potentially associated with respiratory complications, particularly cough, bronchospasm, increased risk of pneumothorax and postoperative ventilation.

When seen for anaesthetic review before operation, the child should be well and the asthma should be satisfactorily controlled with the individual’s medical regimen. If this is not the case the child should be referred to the paediatric team and medical therapy maximized. This may require several days preoperative admission. If the child has symptoms of an upper respiratory tract infection, elective surgery should be postponed as an increase in respiratory complications and exacerbation of asthma caused by increased responsiveness of the airways have been reported under these circumstances.

Assessment of the child’s respiratory function is best achieved by looking for signs of respiratory distress, including tachypnoea, use of accessory muscles of respiration, and the presence of wheeze or focal signs on examination of the chest. More formal testing of pulmonary function can be used successfully in children more than 7 yr of age if they are able to co-operate with the procedure. This allows the degree of reversibility of the airway resistance to be determined and the amount of improvement in pulmonary function tests, with treatment, to be measured. Some children keep regular charts of daily peak flow which allows interpretation of preoperative values in the context of their expected respiratory function. The measured small decrease in respiratory function (FEV₁ and FEFR) after anaesthesia and surgery in children with mild, well-controlled asthma is the same as that for children without asthma. Respiratory mechanics are not different between asthmatic and non-asthmatic children when anaesthetized with either propofol or halothane.

Routine chest x-ray is not required in mild asthma but may be useful in more severe cases to exclude acute infection, the presence of bullae, hyperinflation or pneumothorax. Blood-gas analysis is rarely useful in the assessment of asthmatic children but recording of oxygen saturation in air can provide helpful baseline data.

Before operation, all children should continue receiving their regular medication and use their inhalers before anaesthesia. Additional steroid cover may be required for those receiving regular steroid medication and for those who have been receiving steroids in the preceding 2 months who may potentially have a degree of adrenal suppression. Inhaled steroids alone do not cause suppression of steroid production and additional steroid cover is not required (Table 2). High-dose methylprednisolone is used in some centres in severe asthmatics.

Children receiving theophylline may need to have concentrations checked. The therapeutic range is 10–20 µg ml⁻¹. Occasionally, aminophylline infusions are required in the
peroperative period in poorly controlled asthmatics and careful monitoring of theophylline concentrations is then essential.

Premedication is useful as the stress of surgery may precipitate an attack of bronchospasm. The choice of anaesthetic is affected by the selection of drugs used and those which may predispose to histamine release and bronchoconstriction are avoided. Use of non-steroidal anti-inflammatory drugs (NSAID) is debated. They should be avoided in severe asthma, although in milder forms they may be safe. Techniques which avoid tracheal intubation, when appropriate for surgery, may be less likely to provoke bronchospasm.60

Cystic fibrosis (CF)

This multisystem disease of exocrine glands results in damage to the lung, pancreas and hepatobiliary system. It is inherited in an autosomal recessive manner and is common, occurring in 1:2000 of the population. CF presents as meconium ileus in the neonate or later with cough, wheeze, recurrent chest infections, clubbing and a generalized failure to thrive. Improvements in paediatric care have enhanced both the quality and length of life for patients with CF and most children now reach adulthood. Many children with CF remain relatively well on regimens of regular physiotherapy, prophylactic antibiotics, aggressive management of acute infection, use of pancreatic supplements and nutritional support.

The respiratory disease usually predominates, resulting in a progressive deterioration of lung function caused by excessive, viscous tracheobronchial secretions and impaired mucociliary clearance mechanisms. The pattern of respiratory deficit is of a mixed obstructive and restrictive nature. Long-term bronchiectasis, fibrosis and chronic airway obstruction may develop, resulting in respiratory failure and cyanosis. Hypercapnia occurs late in the disease with increasing ventilation–perfusion mismatch. Chronic hypoxia may lead to pulmonary hypertension, cor pulmonale and eventually cardiac failure. Most of the morbidity and mortality of CF is related to respiratory involvement.18

Liver disease may result in poor function, coagulopathy or varices. Diabetes occurs in approximately 12% of CF patients.

Anaesthesia is required for all common paediatric surgical conditions and also for specific interventions related to the management of CF. These include bronchiolar alveolar lavage for diagnostic purposes, nasal polypectomy, gastrostomy to aid feeding, insertion of indwelling i.v. lines and vascular access ports for supportive therapy, administration of nutritional support and long-term medication.

This group is increasingly being offered heart–lung or lung transplantation as their condition deteriorates and success is reported with this intervention for the treatment of end stage respiratory disease.66 82

Assessment and investigations. Assessment of the child with CF aims to identify the extent of the disease. This is initially from the clinical history, noting particularly the severity of cough and productivity of sputum, frequency of respiratory infections, amount of physiotherapy support required and degree of limitation of exercise. Review by the paediatric team and maximization of medical therapy are the preoperative goals. Signs of respiratory compromise include tachypnoea, hyperinflation of the chest, crackles and wheeze, and a prolonged expiratory phase of respiration. The child should not have any signs of acute respiratory infection. Preoperative SaO2 in air provides a useful baseline. Blood-gas analysis is helpful late in the disease when an increase in carbon dioxide partial pressure is indicative of decompensated respiratory disease and may increase the likelihood of postoperative ventilation. Pulmonary function tests allow documentation of the degree of impairment and extent to which respiratory function can be improved with bronchodilators. Some children with CF are receiving oxygen at home, which indicates that postoperative ventilation may be needed, especially if the proposed surgery causes splinting of the diaphragm. ECG detects any evidence of right heart strain and ventricular hypertrophy. Other features of cardiac impairment such as the presence of peripheral oedema or hepatomegaly may also be present. Diabetes is managed as routine for diabetic patients.

A full blood count reveals anaemia, usually caused by poor nutrition, or increase in leucocyte count with chronic infection. The presence of significant liver disease is identified by increased liver enzyme levels, decreased albumin concentrations and abnormalities of coagulation, which may need to be corrected before operation. A chest x-ray may reveal chronic infiltration of the lung, local consolidation, bronchiectasis, cardiac enlargement or pericardial or pulmonary effusions (Fig. 3).

Features indicative of end-stage disease include an FEV1 less than 30%, hypoxic or hypercapnic respiratory failure.

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**Table 2 Steroid cover for surgery**

| On steroids at present | Hydrocortisone 1 mg kg\(^{-1}\) i.v. | 1. At induction 2. Every 6 h i.v. after operation until able to take steroids orally 3. Reduce to maintenance level over next 4 days, as tolerated |
| Off steroids in preceding 2 months | Hydrocortisone 1 mg kg\(^{-1}\) i.v. | 1. With premedication 2. Every 6 h after operation for 24–48 h 3. Review need for steroids |
| Off steroids for longer than 2 months | 1. No cover but hydrocortisone should be available |
with or without cor pulmonale, decreasing exercise tolerance, increasingly frequent and prolonged hospital admissions, and failure to gain weight despite attempts to supplement the diet.41

Poor preoperative nutritional status is an important factor and associated with increased mortality after major surgery, such as transplantation.66 Frequently this situation can be improved with early nutritional support. Gastro-oesophageal reflux may also be present.

Review of the child’s microbiological results may reveal bacterial colonization frequently with multiply resistant organisms. Precautions to avoid cross infection are very important.

Upper respiratory tract infection (URTI)

URTI occur commonly in childhood with a reported frequency of 2–9 episodes per year in the normal child. Chronic nasal discharge, with features very similar to URTI, is also common, particularly in a child who suffers from adenoidal hypertrophy.

Some studies show an increased incidence of complications during anaesthesia for up to 6 weeks after a URTI.73 There are sporadic reports in the literature of severe morbidity and indeed mortality when the presence of a URTI is the only preoperative indication of any disorder. Complete lung collapse, pneumothorax and myocarditis are some of the major, rare events which have been reported.36 43 83 It has been recommended that surgery is postponed for 4–6 weeks after each URTI. This may cause considerable upset to the child and inconvenience to the parent and hospital service if frequent cancellations occur. Schreiner and colleagues, in their study of more than 15 000 children undergoing day-case procedures, estimated that if all children with mild symptoms had been postponed, 2000 children would have been cancelled to prevent 15 episodes of laryngospasm. In practice, only 0.5% of patients in this study actually had their surgery postponed.65

Prudent judgement is required before proceeding with elective anaesthesia when a child is symptomatic of URTI or is recovering from a URTI. The difficulty when assessing the child comes in quantifying the risk. Several retrospective and prospective studies have examined if there is actually an increase in respiratory complications in children with a URTI and the evidence is conflicting. Several points should be noted. First, most studies have included mainly or exclusively day-case patients who are ASA I or II. Second, the diagnostic criteria for URTI vary. Most studies exclude children who are systematically unwell, those with a fever greater than 38°C, and those who have wheeze or other chest signs on auscultation of the chest.

If URTI is a risk factor, what other factors increase that risk? Children aged less than 1 yr appear to have an increased incidence of airway complications as do those anaesthetized by less experienced anaesthetists and those undergoing airway surgery.10 65 Tracheal intubation may also increase the likelihood of an intraoperative respiratory event but the reported incidence of this complications varies considerably.

In Cohen and Cameron’s study, cough, laryngospasm, bronchospasm and decrease in oxygen saturation were reported to be increased 2–7 times in children with URTI undergoing anaesthesia and by 11 times if intubation of the trachea was required.10 However, Tait and Knight’s study showed no increased risk of complications in children who were symptomatic or non-symptomatic of a URTI undergoing minor surgery.71 They subsequently reported that the increase in complications occurred mainly in those children who had a recent infection, particularly if they had been intubated, rather than in those with current URTI, even though 24% had positive viral cultures.72 Children who had received an anaesthetic during a URTI had a shorter duration of illness.71 Some anaesthetic agents are known to inhibit viral growth in vitro and the mechanisms resulting in hypersensitivity of the respiratory system are complex and multiple.34 Children with URTI are also more likely to have more transient decreases in $S_{a}O_{2}$ in the perioperative period.17 42 49

Perhaps the most important factors are present history and parental opinion. Schreiner and colleagues noted that the parents’ interpretation of their child’s symptoms was more accurately associated with risk of laryngospasm than assessment of URTI features.65

On the day of surgery, these factors need to be taken into account when evaluating the child. A chest x-ray and leucocyte count are occasionally indicated. Those with mild symptoms of runny nose, sneeze, mild fever <38°C and mild cough could be considered for surgery after canvassing parental opinion as to whether the child is generally unwell.65 73 Those with moderate to severe URTI who have features of systemic illness, such as myalgia, pyrexia, anorexia, malaise or headache, and those with a productive

Fig 3 X-ray showing cystic fibrosis, hyperinflation, peribronchial thickening and bronchiectasis.
cough and signs of a lower respiratory tract infection should be postponed for 4–6 weeks.73

Sickle cell disease (SCD)

Children with SCD may present for many types of surgery. The commonest procedures linked to their disease are cholecystectomy for gall stones and splenectomy, but incidental surgery includes all frequently undertaken paediatric surgical conditions.44 As always, elective surgery is associated with a lower complication rate than emergency procedures and allows time for careful reasoned management of the child. Preparation of children with SCD needs close liaison with the haematological and surgical teams.

Screening

Children who come from an ethnic group which has a high incidence of SCD are screened routinely using the Sickledex test which detects HbS to values of 20–25%. It does not identify the concentration or the accompanying types of haemoglobin. Electrophoresis, by high pressure liquid chromatography, accurately measures concentrations of HbS and identifies the presence of HbA2, F, C, D or others.79 Sickledex screening in children less than 6 months of age is not reliable as screening provides a positive test, when HbS is present, but cannot reliably provide a negative result as the variable concentration of HbF in this age group can, if high, mask the presence of HbS. Electrophoresis in neonates and young infants correctly identifies haemoglobinopathies and some centres use this method for neonatal screening on cord blood. It is important to know whether a baby has SCD as although the presence of HbF offers some protection against a sickle crisis, it can occur particularly in the sick neonate. Clinical review of a blood film can identify the presence of sickle cells in some HbSS patients but detection of HbAS or HbSC variants is not likely by this method.

Children with sickle cell trait (HbAS) have less than 50% sickle haemoglobin, are asymptomatic and their cells sickle only in extreme conditions of acidosis, cold or hypoxia. Sickle cell trait is not associated with increased risk of anaesthesia and no special preparation is required.67 Sickle cell disease HbSS has greater than 75% HbS with the remainder being HbF (SS disease) or HbC (SC disease). SC disease is associated with a higher haemoglobin concentration and less systemic upset, although it also has a significant risk of acute sickle crises.19 44 Children with sickle thalassaemia have 75% HbS and a mixture of HbF and HbA; they are also at risk of sickle crisis.

Assessment

The clinical picture of SCD is variable, with some children showing minimal symptoms or signs. Features of chronic anaemia, presence of mild jaundice, hepatosplenomegaly or the effects of vaso-occlusive disease (VOD) are often present. VOD presents as a painful crisis associated with any of the sickling states. It is a serious and frequently progressive condition. The child’s history identifies the possibility of end-organ damage and those that require additional preoperative investigations. Relatively common problems include acute chest syndrome, cerebrovascular accidents, hepatosplenomegaly, splenic sequestration crisis and aplastic crisis. Chronic lung damage, resulting from multiple pulmonary crises and repeated infection, is manifested as pulmonary fibrosis and restrictive lung disease. Long term, this may result in pulmonary hypertension and the development of cor pulmonale. Routine chest x-ray is useful, and formal assessment using lung function tests may be required.

Cardiomyopathy may develop as a result of chronic anaemia or fibrosis. Serial echocardiography estimates the degree of impaired cardiac function. Pericardial and pleural effusions may be part of this scenario. Major complications after general anaesthesia, including death from myocardial necrosis, have been reported in SCD.63 Renal damage also occurs with VOD, with infarcts in the renal medulla, papillary damage and necrosis. End-stage renal failure may result and renal transplantation has been used in some centres for the management of this complication.26 Occlusive disease of the bone also occurs.

Transfusion considerations

The whole practice of exchange transfusion in sickle disease has been questioned.19 74 In a large collaborative study of patients with SCD undergoing surgery, the incidence of complications was only 0.5%. The majority of children undergoing tonsillectomy or adenoidectomy were transfused before operation, as were approximately 50% of those undergoing myringotomy. It was noted that complications were less in those patients who had undergone transfusions, indicating that transfusion would continue to have a role in the preparation of these patients despite the risks and concerns.44 There is concern that the risks of transfusion may outweigh the risk of development of a sickle crisis during a carefully managed anaesthetic. Complications of transfusion include infection, haemolysis, production of antibodies, and future difficulties with cross matching blood products and haematological treatment, such as bone marrow transplantation, which is used in young patients with severe complications of sickle disease.75 For minor surgery, when there is no risk of abdominal splinting, which may cause limitation to lung function after operation, the potential risks involved with blood transfusion may outweigh the potential advantages.

The decision as to whether a child will need a preoperative blood transfusion has to be discussed with the haematology team responsible for the child’s care. If it is decided that an exchange transfusion will benefit the child, this is usually done to achieve a HbS concentration of less than 40% and a haemoglobin concentration greater than 10 g dl\(^{-1}\). These variables are sufficient for most surgery. However, multiple transfusions over a protracted preoperative preparation period may be required, particularly in children with HbSC
Medical assessment of the paediatric patient

disease who tend to have higher haemoglobin concentrations and therefore take longer to exchange transfuse. A more conservative approach is the use of a blood transfusion to achieve a normal haemoglobin concentration which has been reported to result in a similar incidence of complications in the perioperative period in patients with SC or SCD as the use of conventional exchange transfusion. If a child has had a previous cerebrovascular event or is undergoing cardiac surgery, concentrations of HbS less than 8% are aimed for, but can be difficult to achieve.

Hypovolaemia, dehydration and hyperosmolality, all of which increase blood viscosity, must be avoided in patients with SCD. These children are at increased risk from infection as a result of depressed immunity, decreased splenic function secondary to infarction and poor white cell function. They should receive prophylactic antibiotics.

Tourniquets are avoided whenever possible. Mild metabolic acidosis and an increase in serum lactate occurs during and soon after release of the tourniquet because of the resultant stasis and tissue acidosis. If a tourniquet is essential then the limb must be carefully exsanguinated and the tourniquet used for the minimum time necessary. When used carefully, there is no additional morbidity associated with tourniquet use in patients with sickle cell trait.

**Diabetes mellitus**

The majority of children with diabetes have insulin-dependent type 1 disease. It is the commonest endocrine disorder in children, affecting 1 in 500, with a peak incidence at 7 yr. Children have rapidly changing energy needs, and control of blood sugar can be particularly difficult in older children who may have variable food intake and difficulties in complying with insulin therapy because of the psychological demands of adolescence. Long-term effects of diabetes can be decreased if good metabolic control is achieved. End-organ damage, so important in the adult patient, is not usually a feature of childhood disease. It is essential that children with poorly controlled diabetes are admitted early for stabilization of blood sugar management. Emergency surgery in poorly controlled diabetics, especially those with ketoacidosis, is associated with increased morbidity. Abdominal symptoms may be part of the diabetic ketoacidotic picture rather than a pathology needing surgical intervention. If possible, surgery should be postponed until the child’s condition is improved.

A review of the child’s current management, including personal records of blood sugar control and insulin requirements, is helpful in assessing their requirements, taking into account the type of surgery planned. A review by the paediatric team is usually undertaken. For minor surgery, diabetic children can be treated as day cases, but surgery should be planned early on the theatre list. Their morning insulin is omitted and their blood sugar and urea and electrolytes checked. Excessive starvation periods are avoided. The BM stix is measured in theatre, but usually during short procedures no additional dextrose or insulin is required. Children, in common with adults, demonstrate an increase in blood sugar associated with the stress response of surgery. BM stix are checked regularly after operation. After surgery, when they are able to eat, children take their usual dose of insulin and normal diet.

Diabetic children who are undergoing more major surgery or who have poor control of blood glucose should be admitted earlier for stabilization of insulin requirements, and monitoring of blood sugar and HbA1 concentrations. A glucose and insulin infusion is set up before surgery. Hourly BM stix evaluation allows adjustments to be made to the regimen, ensuring careful control of blood sugar during and after surgery. The particular regimen is tailored to the child’s needs and may require addition of potassium and higher concentrations of dextrose.

**Latex allergy**

Perioperative anaphylactic reactions to latex have been increasingly reported. These are type I, IgE-mediated hypersensitivity reactions which can be very severe, although there have been no reported deaths during anaesthesia from this condition. Patients who have had a suspected intraoperative anaphylactic reaction should be investigated to identify the cause. Frequently the diagnosis of latex allergy is made retrospectively and latex may now be the commonest cause of intraoperative sensitivity reactions.

Several groups of paediatric patients are at specific risk of this condition. Some children are known to be latex-sensitive as they have had previous documented anaphylaxis to latex. Children who have spina bifida have 500–1000 times the risk of latex allergy, although the reasons for this are not clear. It may be that repeated surgical interventions in these children causes the high incidence in this group, rather than any genetic predisposition to latex allergy. Another identified high-risk group are children who have a history of a sensitivity reaction, such as bronchospasm, urticaria or eye irritation to balloons or other latex-containing toys.

In order to identify children at risk at the preoperative assessment visit, the anaesthetist must ask specifically about sensitivity to latex products and also to certain foodstuffs, including avocado, kiwi fruit, chestnuts or banana, as there is thought to be a cross sensitivity between latex and fruit proteins. A case-matched review of children with spina bifida indicated the additional risk factors for latex anaphylaxis during anaesthesia as a history of a contact allergy to latex, atopy or food allergies, non-white race and a history of greater than nine previous surgical procedures. Healthy children with a history of atopy and asthma are also considered at increased risk.

**Screening for latex allergy**

Preoperative screening to identify latex sensitivity is possible and has been suggested for high-risk groups. RAST and ELISA tests for latex-specific IgE antibodies and skin prick testing identifies children sensitized to latex but these
tests lack specificity for predicting the likely occurrence of an anaphylactic reaction during anaesthesia. The RAST test is no longer considered sufficiently specific, with 20–45% of patients having a negative RAST test, but positive skin tests. A standardized latex skin prick test solution is not widely available and the test itself can precipitate an anaphylactic reaction. It has been suggested that a RAST test is performed first and if negative, a skin prick test is checked. Elevated total serum IgE in combination with a clinical history is a more specific predictor of anaphylactic reactions occurring during surgery in children with spina bifida.

Management
Management requires scrupulous preoperative preparation to decrease the risk of an anaphylactic response during anaesthesia. Regimens vary but usually include provision of a latex-free environment and preoperative administration of steroids, H2 antagonists and antihistamines (Table 3). Latex-free equipment is available and guidelines for the management of anaphylaxis during anaesthesia should be in place at all anaesthetic settings. Some centres feel that pretreatment is of limited value and that it may mask the early signs of anaphylaxis. Instead, they recommend that when providing anaesthesia for a susceptible patient, the environment is kept latex-free. This management may be preferable in that it avoids potentially sensitizing a child who is in one of the high-risk groups. The cost and social implications of pretreatment are relevant when the additional time required for admission, medications used with their potential side effects and need to avoid day care are taken into consideration.

Children who have had major organ transplants
The quality and length of life of many children with end-stage major organ failure have been improved greatly with successful transplantation surgery. These children may then present for surgery for procedures either related or unrelated to their transplant. This is a group that has varied and sometimes complex medical needs. They frequently require additional psychological preparation and support as they have had long, complicated hospital courses during their illness and transplantation surgery. They are very aware of the seriousness of their condition and are often fearful or even morbid in their outlook. Older children, particularly teenagers, may be unwilling to comply with their therapeutic regimens and up to 20% stop their immunosuppressive therapy unilaterally. Parents are usually very well informed on all aspects of care and can often furnish all the up to date investigation results and details of post-transplant requirements.

Issues common to all groups of transplant patients include the constant threat of rejection of the transplanted organ, risk of infection, and effects of immunosuppressive and steroid therapy. Signs of potential rejection must be sought and are dependent on the individual organ transplanted. Infection remains an important cause of morbidity and mortality. The increased risk of infection includes hepatitis, cytomegalovirus (CMV), Epstein Barr and other bacterial, viral or fungal infections. Opportunistic infection, particularly of the lungs, is a frequent problem while the patient remains on immunosuppressive drugs and steroids. Patients who have had transplants, and are CMV-negative, require CMV negative blood. Any patient considered at risk from graft vs host disease from blood transfusion should receive irradiated products.

There is also an increased risk of the child developing tumours, particularly lymphomas. Many children remain on cyclosporin which is potentially hepatotoxic and nephrotoxic, and often results in hypertension. It may also impair marrow function and cause gastrointestinal upset. Cyclosporin concentrations must be in the therapeutic range throughout the perioperative period. Azathioprine and ALG can cause thrombocytopenia. Steroids may produce the typical cushingoid features, hypertension and abnormal glucose tolerance. Supplemental steroids are required (Table 2).

Some pain management strategies, such as epidurals, may be relatively contraindicated in children receiving immunosuppressive medication because of the potential risk of infection and clotting abnormalities, but this remains debatable.

Renal transplantation
Assessment focuses on the child’s general physical health and current renal function. It is important to determine if children with successful renal transplants have normal renal function or whether they continue to have some limitation of function. This requires review of urea, electrolyte and creatinine concentrations, full blood count and coagulation studies. Glomerular filtration rate (GFR) is often approximately 50% of normal values and creatinine concentration may remain slightly increased even with a well functioning transplanted kidney. If creatinine concentration if increased by more than 10% above their usual value, the possibility of acute rejection should be considered. The function of the transplanted kidney tends to decline gradually, as shown

Table 3 Latex allergy: preoperative preparation. Patients who have not had a reaction but are in a high-risk group (e.g. history of spina bifida, genitourinary abnormalities or multiple surgical procedures) are not routinely pretreated nor are there any special precautions used. A high index of suspicion is maintained during and after the case

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dose</th>
<th>Administration</th>
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<tbody>
<tr>
<td>Methylprednisolone</td>
<td>1 mg kg⁻¹ (maximum 50 mg)</td>
<td>6 hourly i.v.</td>
</tr>
<tr>
<td>Ranitidine</td>
<td>1 mg kg⁻¹</td>
<td>6 hourly i.v. over 20 min</td>
</tr>
<tr>
<td>Chlorpheniramine</td>
<td>1 month–1 yr</td>
<td>250 µg kg⁻¹</td>
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<tr>
<td></td>
<td>1–5 yr</td>
<td>2.5–5 mg</td>
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<tr>
<td></td>
<td>6–12 yr</td>
<td>5–10 mg</td>
</tr>
<tr>
<td></td>
<td>all 6 hourly i.v. slowly</td>
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All given 6 hourly i.v.– 2 doses before operation and continued for 24 h after operation
by increasing creatinine concentration and decreasing GFR. Renal excretion of drugs is usually normal but drugs which could potentially damage the kidney, such as NSAID, are avoided. The chronic anaemia of renal failure is usually resolved after transplantation but some children remain on erythropoietin. Assessment of the limbs for planned vascular access is important so that vessels which may be useful for vascular shunts later in life are preserved. Prolonged periods of fasting are avoided and an i.v. infusion may be required before operation to ensure maintenance of intravascular volume. Hypertension is a frequent feature and children are usually receiving many drugs, including hydralazine, nifedipine, labetalol and diuretics. Prophylactic antibiotics are usually required.

**Intrathoracic organ transplantation**

After successful transplantation surgery, children usually have multiple general anaesthetics for routine post-transplant investigations. They also require anaesthesia for other childhood surgical procedures. When assessing such patients, recent results of all follow-up investigations must be available. Liaison with the transplant team is essential.

**Heart transplantation**

The long-term outcome after paediatric heart transplantation is better than transplantation including the lungs. Children with heart transplants tolerate subsequent general surgical interventions well. As with other transplants, the common problems are rejection and infection. Other major complications after cardiac transplantation include pulmonary hypertension, coronary artery disease and lymphoproliferative disease. As part of routine post-transplant care, children need investigations, including endomyocardial biopsy and cardiac angiography, which require general anaesthesia.

Preoperative assessment includes reviewing the child’s history and present condition. Potential features of rejection include decreased appetite, general malaise, pyrexia or irritability. Other features include fluid retention and signs of cardiac failure. Anaesthesia during an episode of rejection is associated with increased morbidity and should be avoided whenever possible.

An ECG, echocardiography and chest x-ray are part of the routine preoperative assessment. The ECG is reviewed, looking for arrhythmias, change in axis or decrease in total voltages. Echocardiography provides an indication of present function, with decrease in shortening fraction being indicative of failing cardiac function.

One of the causes of death in children after cardiac transplant is coronary artery disease; this may be asymptomatic as these children do not feel ischaemic pain. Evidence of coronary artery disease was reported in 15% of children in the Stamford series and in 3% of the Harefield series. Additional complications include decreased renal function from immunosuppressive therapy, hypertension and the complications of steroid therapy. Rarely a permanent pacemaker is present and if so a routine pacemaker check is required.

**Lung or heart–lung transplant**

The commonest group of children who have a lung or heart–lung transplant are those with cystic fibrosis. Unfortunately, the long-term survival for lung transplantation is less than that for heart transplantation as many patients develop obliterative bronchiolitis which is part of the rejection process and appears to be difficult to prevent. Recurrent chest infection is common. Routine clinical assessment of present general health, exercise tolerance and identification of any features indicative of rejection or infection is undertaken. Preoperative investigations include full blood count, urea, electrolyte and creatinine concentrations, lung function tests, measurement of arterial pressure and urinalysis. ECG, chest x-ray and pulmonary function tests are performed. If FEV1 and FVC measurements decrease by more than 15% of the child’s usual values, further investigation with bronchoscopy, bronchial alveolar lavage and transbronchial biopsy may be required to determine if rejection or infection is present.

**Liver transplantation**

Children can be very well after successful liver transplantation and hepatic metabolism may be returned to normal. Most children return to normal activities. The majority of patients remain on cyclosporin and steroids after transplant, although some need antihypertensive, diuretic or anti-convulsant medications also. Assessment includes identifying the features of impaired liver function, in particular evidence of residual portal hypertension. This may include ascites, oedema or a history indicative of the presence of oesophageal varices, impairment of respiratory function caused by ventilation–perfusion mismatch and alveolar hypoventilation. In addition to routine preoperative evaluation of renal function and full blood count, a coagulation screen and liver function tests are important, although most are in the normal range after successful transplantation. Acute rejection is shown by the presence of cholestatic jaundice reflected in abnormal liver function tests, prolonged prothrombin time, lymphocytosis and eosinophilia. Prothrombin time is the earliest indicator of impaired liver function although it may remain within the normal range until 70% of liver function is lost. An INR greater than 1.4 is an indication that significant impairment of liver function is present.

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