

Anaesthetic implications of congenital heart disease for children undergoing non-cardiac surgery

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Abstract

Children with congenital heart disease (CHD) are at increased risk of cardiac arrest and 30-day mortality from major and minor surgical procedures compared with healthy children. Therefore, a prerequisite for anaesthetizing these children is a thorough knowledge of the specific cardiac anatomy, cardiorespiratory physiology and the potential risk of complications for each individual case. Anaesthetists must be familiar with not only the normal, series cardiac circulation but also the parallel (or balanced) and single-ventricle circulations. Anaesthetists must also understand the complex interaction between systemic and pulmonary vascular resistance and the many factors that influence these variables, and be aware of the four major complications associated with CHD and know which children are most at risk. Induction and maintenance of anaesthesia should be individualized to the child and tailored to the type of surgery. Whether surgery occurs in the local hospital or tertiary cardiac centre is a matter for debate. Some children require full cardiac anaesthesia, intensive care and cardiology support, making care in the local hospital inappropriate, whereas, for others, care in their local hospital is both safe and convenient.

Keywords anaesthesia; cardiac; children; complications; congenital heart disease; non-cardiac surgery; paediatric

The scope of the problem

Congenital heart disease (CHD) is the commonest birth defect. It occurs in approximately 1 in 125 live births. Of these, 30% have extra-cardiac anomalies (such as tracheoesophageal fistula, anorectal anomalies, cleft lip and palate, and skeletal anomalies), which might require surgery within the first year of life. In addition, advances in perioperative care mean many children now survive to adulthood and are subject to common conditions such as appendicitis, for which they present to anaesthetists at the local hospital. However, children with CHD are at increased risk of perioperative cardiac arrest and 30-day mortality compared with children without CHD.^{1,2} Children most at risk of perioperative morbidity are those with major cardiac anomalies, cyanosis, pulmonary hypertension, congestive cardiac failure,

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Learning objectives

After reading this article, you should be able to:

- describe the physiology of a series, parallel and single-ventricle circulation
- classify congenital heart disease and list four potential major complications of congenital heart disease in children
- describe the key features in preoperative assessment and anaesthetic management.

are <2 years of age and those undergoing emergency surgery. Children with CHD undergoing non-cardiac surgery present a unique set of challenges. The anaesthetic implications of caring for such children are outlined here.

Different types of circulation

Normal 'series' circulation

The normal heart has a pulmonary and systemic circulation that work together in series (Figure 1). Blood flows continually and consistently through the 'series' of chambers, valves and pipes. The blood is pumped under different pressures to both the lungs and the body. There are no holes, so there is no mixing of deoxygenated and oxygenated blood and no option for blood to go anywhere else except the next chamber or pipe in the series. Some forms of CHD, such as atrial septal defect (ASD), have the usual 'series' circulation but one or more 'holes' exist through which mixing of deoxygenated and oxygenated blood can occur. The direction blood flows through the lesion depends on the pressure gradient and is documented in the echocardiography report. Left-to-right shunting of blood results in excessive pulmonary blood flow (PBF), and right-to-left shunting causes reduced PBF and cyanosis. The amount of shunting depends on the size of the defect and the relative pressure gradients, which are a reflection of the resistance to flow in each circuit. Changes in systemic (SVR) and pulmonary (PVR) vascular resistance as a result of anaesthesia have the greatest impact on large, unrestrictive defects.

'Parallel' or 'balanced' circulations

Instead of the pulmonary and systemic circulation being in 'series' they can be in 'parallel' (Figure 2). This is also called

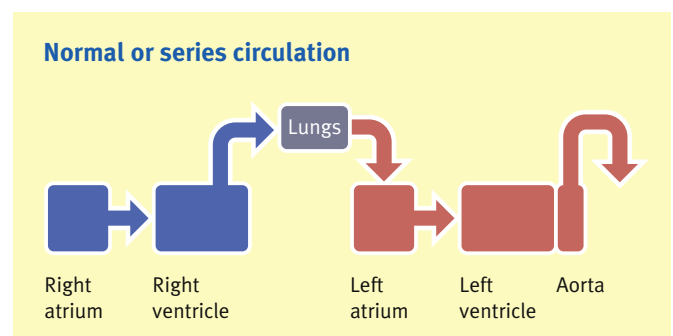


Figure 1

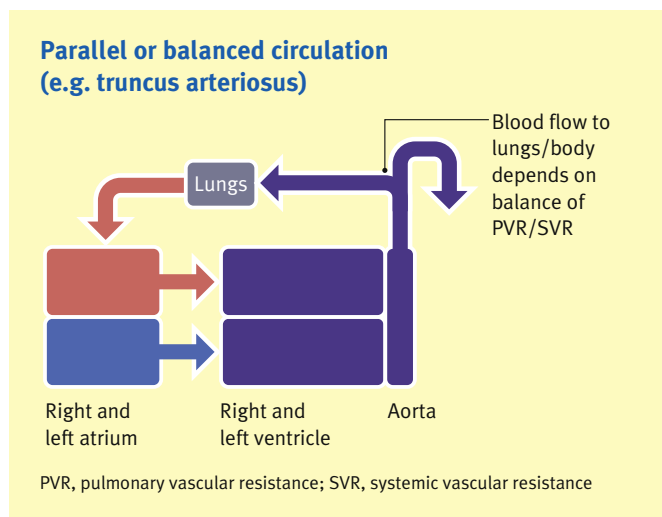


Figure 2

a 'balanced' circulation. In addition to mixing of oxygenated and deoxygenated blood, the anatomical relationship of any intra- and extra-cardiac communications means that blood flow to the systemic and pulmonary circulation varies depending on the relative resistance in each circuit. Thus, how much blood flows to the lungs or to the body is a 'balance' that depends on PVR and SVR. Excessive PBF causes pulmonary oedema and poor systemic perfusion (with associated complications of poor coronary, renal and splanchnic perfusion). Insufficient PBF causes profound cyanosis. Examples of CHD with a 'balanced' circulation include, but are not limited to, truncus arteriosus and hypoplastic left heart syndrome. Because PVR is usually less than SVR, infants with these disorders often have high PBF and poor systemic perfusion that can compromise coronary artery, renal and splanchnic blood flow. Optimization of PVR and SVR in these infants can be very difficult. Liaison with a regional paediatric centre that undertakes cardiac surgery is essential so that specific anaesthetic management can be discussed with a consultant paediatric cardiac anaesthetist.

'Balanced' circulation physiology is also found in children who have had a modified Blalock–Taussig (BT) shunt. A BT shunt is performed to augment PBF in infants whose PBF is otherwise insufficient. The modified BT shunt usually consists of a Gortex tube positioned between the right subclavian artery and the right pulmonary artery. Blood flows down the pressure gradient from the right subclavian artery to the right pulmonary artery. However, an increase in PVR and decrease in SVR (as can occur during a hasty induction of anaesthesia with excessive induction agent, and a mild degree of hypoxia and hypercarbia) can seriously compromise shunt flow, resulting in reduced cardiac output and even cardiac arrest.

Infants with large unrepaired atrioventricular septal defects or ventricular septal defects can also exhibit 'balanced' circulation physiology. These infants have predominantly left-to-right shunt flow. Induction and maintenance of anaesthesia can reduce SVR to the extent that shunt flow is reversed, resulting in sudden and unexpected desaturation. The excessive PBF from the underlying left-to-right shunt also means that these infants are at risk of pulmonary hypertension (see below). Therefore, a pulmonary

hypertensive crisis is another cause of sudden desaturation during anaesthesia.

Single-ventricle circulation

A BT shunt can precede a full anatomical correction (e.g. tetralogy of Fallot) or be the first of three stages in the formation of a single-ventricle circulation (Figure 3). The second stage is known as a Glenn shunt or bidirectional cavopulmonary shunt. In this operation, the BT shunt is removed and the superior vena cava (SVC) connected directly to the pulmonary artery, leaving the inferior vena cava (IVC) to continue draining into the single, common atrium and ventricle. The child remains cyanosed after this procedure, with oxygen saturations of 75–85%. The third stage is known as the Fontan procedure or total cavopulmonary connection. The IVC is connected to the pulmonary artery, thereby separating the systemic and pulmonary circulations and preventing mixing. This results in normal arterial oxygenation. Thus, a single-ventricle circulation (Figure 3) consists of one ventricle that pumps blood to the systemic circulation, and a pulmonary circulation that relies on blood flow to the lungs under passive venous pressure from the SVC and IVC. Pulmonary blood flow, therefore, depends entirely on the pressure gradient from the SVC/IVC to the left atrium (usually a common left and right atrium). In the single-ventricle circulation, elevations in PVR and positive intrathoracic pressure can severely reduce PBF. Under anaesthesia, this has implications for ventilatory strategy. For example, spontaneous ventilation improves PBF, but positive-pressure ventilation can give greater control of oxygenation and minute ventilation. Care should be taken to optimize positive end-expiratory pressure, and minimize inspiratory time and peak inspiratory pressure to maximize PBF.

Classification of congenital heart disease

CHD can be classified in a number of ways. Box 1 gives one such classification based on whether there is mixing of blood that is predominately left to right or right to left, whether there is a complex interaction between SVR and PVR or whether there is obstruction to blood flow in the heart. However, CHD is not always straightforward; some children will have more than one

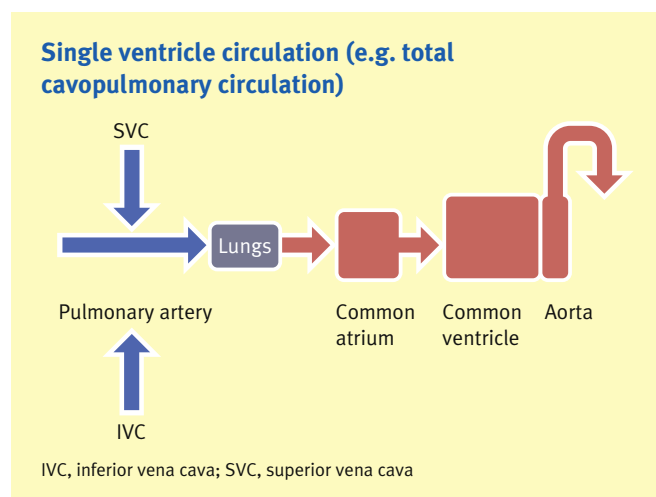


Figure 3

A physiological classification of congenital heart disease

1 'Simple' left-to-right shunt lesions: these cause an increased pulmonary blood flow

- a Atrial septal defect
- b Ventricular septal defect
- c Atrioventricular septal defect
- d Patent ductus arteriosus
- e Aortopulmonary window

NB The effect of the shunt on right ventricular and respiratory physiology differs depending on the level at which shunting occurs.

2 'Simple' right-to-left shunt lesions: these cause a reduction in pulmonary blood flow with cyanosis

- a Tetralogy of Fallot. Consists of right ventricular outflow tract obstruction, right ventricular hypertrophy, ventricular septal defect and an overriding of the aorta
- b Pulmonary atresia
- c Tricuspid atresia
- d Ebstein's anomaly. Consists of downward displacement of an abnormal tricuspid valve into the right ventricular cavity, part of the right ventricle is thus incorporated into the right atrium (atrialized right ventricle), and the remaining right ventricular cavity is malformed

3 Complex shunts: these cause mixing of pulmonary blood flow and systemic blood flow. Cyanosis occurs as a result of complex interactions between systemic vascular resistance and pulmonary vascular resistance

- a Transposition of the great arteries
- b Truncus arteriosus
- c Total anomalous pulmonary venous drainage
- d Double-outlet right ventricle
- e Hypoplastic left heart syndrome

Most of these lesions (except total anomalous pulmonary venous drainage) are examples of a parallel circulation.

4 Obstructive lesions

- a Coarctation of the aorta
- b Interrupted aortic arch
- c Aortic stenosis
- d Pulmonary stenosis

Box 1

lesion, thereby fitting into more than one category, whereas others might move between categories as complications develop.

Four potential major complications of congenital heart disease in children

Arrhythmias

Some cardiac surgery procedures predispose patients to the development of arrhythmias in later life. Children at high risk are those with extensive atrial sutures or those who have undergone ventriculotomy.

Damage to the sinus node causes atrial arrhythmias, such as sinus bradycardia, sinoatrial block or supraventricular tachycardia. These can be asymptomatic or associated with haemodynamic collapse and sudden death. Permanent damage is commonest after repair of sinus venous defects, ASDs, atrial switch procedures (Mustard or Senning) and total cavopulmonary anastomosis (Fontan procedure). Children who have undergone a Mustard, Senning or Fontan procedure have a risk of atrial dysrhythmias that increases year by year. Perioperative atrial tachycardias are common in children with Ebstein's anomaly, and malignant arrhythmias and sudden deaths during placement of intracardiac lines have been described. Therefore, central venous cannulation should be undertaken cautiously.

Damage to the atrioventricular node and bundle of His causes ventricular arrhythmias. This is commonly seen in patients who have undergone ventriculotomy or had a right ventricle (RV) to pulmonary artery (PA) conduit. Necrosis and progressive fibrosis extending into the conduction system is thought to be the cause of late-onset arrhythmias. Arrhythmias leading to death arise in up to 30% of patients with single-ventricle circulations.³ Children who have undergone a repair of tetralogy of Fallot are at increased risk of sudden death or sustained ventricular tachycardia. Children who experienced transient congenital heart block after corrective tetralogy of Fallot surgery have a 10 times greater risk of developing late-onset congenital heart block. Poor exercise tolerance is suggestive of RV failure and this is a risk factor for ventricular tachycardia and sudden death.

Preoperative assessment includes a careful history, noting exercise tolerance, evaluation of arrhythmia risk (as described above, such as previous ventriculotomy, RV-PA conduit, tetralogy of Fallot repair, Mustard, Senning or Fontan procedure; previous postoperative arrhythmias, even if transient) and a preoperative ECG. Right bundle branch block is common but unlikely to degenerate into congenital heart block. Ventricular ectopics are an ominous sign because 30% of patients with this disorder are at risk of sudden death. In a child at risk of arrhythmias, factors known to lower the threshold for ventricular ectopics should be used with caution; for example, hypercarbia, acidosis, hypoxia and large doses of local anaesthetic with epinephrine.

Cardiac failure

Cardiac failure occurs when the heart cannot pump enough blood to meet the metabolic demands of the body. It is the end result of a continually volume- or pressure-overloaded heart. Cardiac reserve is the difference between cardiac output at rest and that at maximum capacity (i.e. the spare capacity of the heart to undertake more work). Limited cardiac reserve describes the situation in which the heart is functioning at near maximal capacity even when the child is resting.

Children with limited cardiac reserve must be identified because they are susceptible to cardiac failure during anaesthesia. Clinical presentation varies with age. Tachypnoea, tachycardia and cool peripheries are common to all ages. In infants, additional signs include poor feeding, failure to thrive, sweating and hepatomegaly. Older children manifest poor exercise tolerance, poor weight gain and chest crackles.

Gaseous or intravenous induction is possible, but induction times will be prolonged, so patience is required to prevent excessive administration of induction agent. Sevoflurane 4% rather

than 8% is often sufficient; propofol can have a profoundly deleterious effect on blood pressure and cardiac output in these children; and ketamine might be a more preferable drug. If severe cardiac failure is suspected, attention should be given to the use of vasoactive agents such as milrinone or dobutamine before induction, and invasive monitoring might be needed even for minor surgery. Major surgery should be undertaken only with extreme care after liaison with the cardiologist and surgeon, and frank discussion with the child and his or her parents. Full paediatric intensive care facilities should be available.

Cyanosis

Chronic cyanosis is associated with polycythaemia and abnormal haemostasis. Polycythaemia is an adaptive response to hypoxia allowing normal systemic oxygen delivery without a sustained increase in cardiac output. As the haematocrit increases, blood viscosity increases dramatically, which in turn increases SVR and PVR and reduces coronary artery perfusion. In children with cyanotic heart disease, hyperviscosity is associated with thromboses of intracranial veins and sinuses, which can result in stroke. Children under 5 years are at highest risk, especially in the presence of dehydration, fever or iron deficiency. Haematocrit levels greater than 65% can actually reduce oxygen delivery because of increased viscosity, especially if iron deficiency is also present. Hyperviscosity syndrome is characterized by headache, dizziness, blurred vision, general fatigue, muscle weakness, myalgia, paraesthesia of fingers, toes and lips, and reduced mentation.

Children with cyanotic CHD should have their haemoglobin and haematocrit measured. Preoperative intravenous fluid therapy should be considered in children with a haemoglobin greater than 18 g/dl (or haematocrit >60%) to avoid symptoms of hyperviscosity or thrombosis. Preoperative fasting should be kept to a minimum, and these children should be scheduled first on the operating list.

Of those children with cyanotic CHD, 20% have abnormal laboratory tests of haemostasis. Prolongation of prothrombin time or partial thromboplastin time are the commonest abnormalities. Other reported abnormalities include thrombocytopenia, platelet dysfunction, hypofibrinogenaemia and accelerated fibrinolysis.⁴ Abnormal haemostasis seems to correlate with the degree of cyanosis and erythrocytosis, but the cause is unclear. Indeed, even when coagulation tests are normal, the risk of excessive postoperative bleeding remains.

Children might give a history of easy bruising or epistaxis, but lack of signs and symptoms should not provide reassurance. All cyanotic children should be assumed to be at risk, and drugs such as aspirin, non-steroidal anti-inflammatories and heparin can exacerbate the already abnormal haemostasis. If a child is on aspirin to maintain shunt patency, the risk of shunt thrombosis is usually greater than that of bleeding, so aspirin should be continued.

In cyanotic children, the use of premedication is common. Premedication minimizes distress, thereby reducing oxygen consumption and preserving the already limited oxygenation. End-tidal carbon dioxide monitoring underestimates arterial carbon dioxide because of right-to-left shunt. Cyanotic children also have blunted hypoxic ventilatory response. Therefore, profound hypoxia occurs without causing the normal response of increased ventilation, especially when respiratory depressants such as opioids have been given. Therefore, close supervision in

the recovery room and during the first postoperative night is recommended.

Pulmonary hypertension

Pulmonary hypertension (PHT) is defined as having a mean PA pressure above 25 mm Hg at rest or 30 mm Hg on exercise. There are many causes of PHT, which are classified by the World Health Organization into five categories. However, in the context of children with CHD presenting for non-cardiac surgery, two groups should be considered at risk of PHT:

- 1 those with, or a history of, excessive PBF due to left-to-right shunt lesions; these children might have reactive pulmonary vasculature, making them at risk of a PHT crisis
- 2 those with prolonged obstruction to pulmonary venous drainage or exposure to high left atrial pressures.

PHT develops earlier in some lesions (e.g. atrioventricular septal defects) and in certain patient populations, such as children with Down's syndrome. The reversibility of PHT after repair of the defect is also variable. In PHT, the vascular bed is associated with fixed structural changes but variably reactive vascular smooth muscle affected by a number of factors. Acidosis, hypercarbia, hypoxia, hypothermia, increased sympathetic stimulation and increased airway pressure all increase PVR. Appropriate management of these variables can reduce PVR, improve right ventricle function and minimize the degree of right-to-left intracardiac shunting.

Before surgery, children with PHT or those who are at risk of PHT should be identified. A recent echocardiography report to estimate PA pressure and presence of intracardiac shunts including direction of flow should be obtained. Any elevation in PA pressure requires referral to a paediatric cardiologist for further investigation. If available, previous cardiac catheterization reports should be reviewed to obtain a measure of PVR and the degree of responsiveness to oxygen and pulmonary vasodilators such as nitric oxide. Children with PHT have a high risk of significant cardiovascular collapse during anaesthesia,⁵ and this must be discussed with the child's family and full paediatric intensive care facilities should be available.

Anaesthetists must understand the specific intracardiac anatomy and the physiological consequences of changes in PVR, SVR and shunt flow because different situations require different strategies. For example, anaesthesia for obstructed hernia repair in a 4-month-old infant with a repaired atrioventricular septal defect and residual PHT requires PVR to be as low as possible to optimize function of the right ventricle and avoid right ventricular failure. However, if the same child had an unrepaired atrioventricular septal defect with predominant left-to-right shunt flow, he or she would have a 'balanced circulation'. In this situation, although the child is at risk of developing PHT, during anaesthesia a slight elevation of PVR can help prevent excess PBF and 'stealing' from the systemic circulation so reducing blood pressure and coronary perfusion. However, a grossly elevated PVR is undesirable because this compromises PBF, causing right-to-left shunting and cyanosis, similar to a PHT crisis. Treatment of a PHT crisis is influenced by the intracardiac anatomy and is beyond the scope of this article. However, generally treatment involves eliminating the stimulating cause by administering opioids/deepening anaesthesia, and if an intracardiac shunt is present, α -agonists (e.g. phenylephrine) should be given. Inotropic support of the right ventricle might also be required.

Application of cardiorespiratory physiology and anatomy in preoperative assessment and risk of complications in anaesthetic management: three specific examples

Cardiac lesion and type of emergency surgery	Preoperative assessment	Risk of four major complications	Induction and maintenance of anaesthesia	Beware of:
2-month-old infant with a large VSD presenting for emergency surgery for scrotal swelling	Unrepaired lesion with left-to-right shunt Normal oxygen saturation ↑ PBF, might need to manipulate PVR/SVR Likely CHF with possibility of poor cardiac function	Heart failure PHT	Slow and cautious because of risk of poor cardiac reserve Gas or intravenous Maintain PVR to avoid excess pulmonary flow Avoid high F_{iO_2} and hyperventilation Might need higher ventilator pressures if pulmonary oedema present	Poorly tolerant of systemic vasodilatory effects of anaesthetic agents Pulmonary vasodilatory effects of oxygen and hypocarbia exacerbating already excessive PBF Avoid air bubbles in venous lines because of the risk of paradoxical emboli
6-year-old child with repaired TOF presenting for tooth extractions for dental caries	Repaired lesion Normal oxygen saturation No need to balance PVR/SVR Assess cardiac function especially with regard to RV function owing to problems with pulmonary valve or residual RVOT obstruction	Arrhythmias: needs ECG Possible RV failure	Gas or intravenous	Caution with local anaesthetic containing high doses of epinephrine
3-year-old with Glenn shunt presenting for reduction and fixation of fractured wrist	Underlying diagnosis: tricuspid atresia Palliated with Glenn shunt Oxygen saturation 80% (as expected) Need to maintain low PVR to aid PBF Cardiac function expected to be adequate	Cyanosis and polycythaemia: keep well hydrated Possibilities of arrhythmias if previous problem	Gas or intravenous Avoid hypoxia, hypercarbia and atelectasis, which increase PVR Avoid high ventilator pressures and high PEEP, which compromise passive PBF	Slight head-up position aids PBF Avoid air bubbles in venous lines because of the risk of paradoxical emboli

CHF, congestive heart failure; F_{iO_2} , fraction of inspired oxygen; PBF, pulmonary blood flow; PEEP, positive end-expiratory pressure; PHT, pulmonary hypertension; PVR, pulmonary vascular resistance; RV, right ventricle; RVOT, right ventricular outflow tract; SVR, systemic vascular resistance; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Table 1

Preoperative assessment

A thorough understanding of the specific anatomy and physiology, detailed history and examination, and assessment of potential complications of CHD is imperative before administering general anaesthesia.

- 1 Understanding anatomy and physiology can be evaluated by the following questions:
 - a What is the underlying cardiac lesion (this helps to predict complications)? Is the lesion currently unrepaired, repaired (with normal anatomy) or palliated (some form of repair undertaken but not resulting normal anatomy)?
 - b What is the oxygen saturation in air? Is it what would be expected from knowing the child's anatomy and physiology? If not, there is either too much or too little PBF.
 - c Is the cardiac function good, moderate or poor? Review the most recent echocardiogram report and make an assessment of adequacy of cardiac reserve.
 - d Is the PVR or SVR likely to need manipulation to aid systemic or PBF? (The answer to this question will influence both choice of induction agent and ventilatory strategy.)
- 2 History and examination should specifically address the following:
 - a Recent upper or lower respiratory tract infection? This can cause changes in airway reactivity and PVR, which might be poorly tolerated in children with reduced pulmonary compliance or PHT, especially those with a Glenn or Fontan circulation.
 - b ECG/chest radiograph/blood pressure. Children at risk of arrhythmias or outflow tract obstruction should have an ECG recorded for signs of ventricular hypertrophy, ectopics, bundle branch block or other abnormalities. I do not consider a chest radiograph to be mandatory for minor elective surgery, but it should be done before major surgery. Children with previous surgery to the aorta should have four-limb blood pressure recorded.
 - c Venous access. Previous surgery and intensive care admissions with or without cardiac catheterization can result in poor venous access, and central vessels can be hard to recannulate.
 - d Medication. Many children are on numerous drugs such as diuretics, angiotensin-converting enzyme inhibitors, anti-arrhythmics and aspirin. No specific guidelines exist for discontinuation of aspirin before surgery and in children with systemic to PA shunts; however, the risk of shunt thrombosis is greater than the risk of bleeding.
 - e Premedication. Anxiety and distress increase oxygen consumption and myocardial work, which might be poorly tolerated in a child with limited cardiac reserve. Premedication can also facilitate induction of anaesthesia and reduce the amount of induction agent required, so minimizing the drop in SVR.
 - f Endocarditis prophylaxis. Guidelines produced by the National Institute for Health and Clinical Excellence and the American Heart Association should be followed (www.nice.org.uk; CG64). If in doubt, contact the child's cardiologist.
- 3 Assessment of potential major complications of CHD:
 - a Arrhythmias (history of previous arrhythmias including post-cardiac surgery, evidence of ectopics on ECG).
 - b Signs of heart failure.

c Cyanosis and polycythaemia. (Is preoperative hydration required?)

d Pulmonary hypertension. (What is the PA pressure on echocardiography and is further investigation required?)

Anaesthetic management

Because the complexity of CHD and the scope of non-cardiac surgery are so varied, it is impossible to prescribe a formula for anaesthetizing children with CHD for non-cardiac surgery. Induction and maintenance must be individualized to the child according to the issues outlined here and tailored to the type of surgery. Three examples of the application of these principles are given in Table 1.

Children with CHD have a high risk of morbidity and mortality compared with healthy children; therefore, they should be anaesthetized for elective surgery only by a fully trained paediatric anaesthetist who has a thorough understanding of the child's specific cardiorespiratory physiology and potential complications. Whether surgery occurs in the local hospital or tertiary cardiac centre depends on the skill-mix of the team members and the support facilities available. There is no doubt that, for some children, their local hospital provides a safe and convenient environment, yet, for other children, full cardiac anaesthesia, intensive care and cardiology support is essential. Each case requires careful evaluation of the risk factors, thorough forward planning and good communication between all healthcare professionals, including the child and his or her family. ◆

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