

# Anaesthetic management of children with congenital heart disease for non-cardiac surgery



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## Key points

Children with congenital heart disease undergoing non-cardiac surgery have increased risk of perioperative morbidity and mortality.

Highest risk factors are complex disease, poorly compensated physiology, and the presence of long-term complications.

Intermediate-risk factors are major surgery, emergency surgery, age under 2 yr old, preoperative hospital stay > 10 days, and ASA physical status IV or V.

Transfer high-risk children to a specialist centre. Discuss and consider transfer of intermediate-risk children depending on local skills and facilities.

Sevoflurane or ketamine induction, sevoflurane or isoflurane maintenance, and opioid or regional analgesia are acceptable anaesthetic techniques.

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Congenital heart disease (CHD) is the most common birth defect, occurring in approximately one in 125 live births.<sup>1</sup> Advances in medical care have led to ~90% of these children surviving to adulthood.<sup>2</sup> Children with CHD are subject to the same childhood illnesses as healthy children and therefore present to their local hospital requiring both elective and emergency surgery. In general, children with CHD undergoing non-cardiac surgery have an increased risk of morbidity, perioperative cardiac arrest, and a higher 30 day mortality.<sup>3–6</sup> However, the complexity of cardiac defects and the variety of non-cardiac surgery make it impossible to generalize. A 9-yr-old child with a repaired atrial septal defect (ASD) who requires an emergency appendectomy presents minimal risk; yet, a 12-yr-old with a failing single-ventricle circulation for the same operation presents a significant challenge even to the most experienced paediatric cardiac anaesthetist.

## Aims and limitations

This article aims to discuss the principles underlying the management of children with CHD who present to their local hospital for non-cardiac surgery. We describe the physiology of normal, balanced, and single-ventricle circulations; suggest a classification of high- and low-risk children; and discuss the preoperative assessment, anaesthetic management, and postoperative care of these children in the local hospital setting.

There are three main limitations. First, the evidence for classifying children with CHD who require non-cardiac surgery as high, intermediate, and low risk is very limited. There are no UK data; therefore, the US data are discussed. While this has some limitations in applicability to UK practice, it nonetheless

provides a starting point for risk stratification. Secondly, the evidence base for perioperative management is also limited.<sup>7</sup> This is because few studies exist evaluating anaesthetic techniques in children with heart disease for non-cardiac surgery; studies of complications in a rapidly changing field such as cardiac surgery may reflect outdated treatment strategies; and the variety and complexity of CHD makes a one-size-fits-all approach to evidence-based perioperative management impossible. Thirdly, we do not describe the management of high-risk children, who require specialist paediatric anaesthesia, and intensive care because this review is intended for the district general hospital anaesthetist who may have to manage a child with heart disease either as an emergency or for minor elective surgery.

Therefore, this review presents an applied physiological and evidence-based approach that is general not specific and aimed at the local hospital anaesthetist rather than the specialist.

## Physiology of different types of circulation

### Normal or 'series' circulation

The 'normal' circulation can be considered as separate systemic and pulmonary circulations working together in series. Most types of repaired CHD have this type of circulation. Some forms of unrepaired CHD also have a 'normal' circulation but with one or more holes where mixing of blood can occur [e.g. ASD or ventricular septal defect (VSD)]. Blood flows through the hole down a pressure gradient and creates a shunt. Left-to-right shunts result in increased pulmonary blood flow (PBF) and potentially decreased systemic blood flow; right-to-left shunts cause deoxygenated blood to flow into the systemic circulation, causing

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cyanosis and reduced PBF. The amount of shunting depends on the size of the defect and the pressure gradient. Changes in systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR) as a result of anaesthesia, including the administration of oxygen, have greatest effect on large, unrestrictive defects. Infants with a large unrestricted defect, such as a VSD, may exhibit 'balanced' circulation physiology.

### Parallel or 'balanced' circulation

Instead of the pulmonary and systemic circulations being separate entities, they communicate with each other and function physiologically as being in parallel. The anatomical abnormalities cause the blood flow to the systemic and pulmonary circulation to vary depending on the relative resistance in each circuit. Thus, blood flow to the lungs and body is a 'balance' between SVR and PVR; hence, the term 'balanced' circulation. Excessive PBF causes pulmonary oedema and poor systemic perfusion (which may compromise coronary and splanchnic perfusion); insufficient PBF causes profound cyanosis.

Examples of children with 'balanced' circulation physiology who may present to the local hospital are infants with a large unrepaired atrioventricular septal defect or VSD. These infants have predominantly left-to-right shunt flow. High concentrations of oxygen will increase PBF and reduce systemic perfusion; conversely, large doses of induction agent may reduce SVR so much that shunt flow is reversed causing desaturation. The excessive PBF also makes the child vulnerable to developing pulmonary hypertension (PHT).

Other examples of 'balanced' circulations include children with a modified Blalock–Taussig (BT) shunt, truncus arteriosus, and hypoplastic left heart syndrome. These children can be very difficult to manage, and liaison with a regional paediatric cardiac centre is advised.

### Single-ventricle circulation

Some forms of CHD are not amenable to full anatomical correction, that is, a biventricular repair resulting in a normal 'series' circulation. Therefore, these children will be palliated by creating a circulation based upon a single ventricle. The single-ventricle pumps oxygenated blood around the body, while blood flows passively to the lungs down a pressure gradient from the pulmonary artery (PA) to the left atrium (LA). A BT shunt is usually, but not always, the first stage in the formation of a single-ventricle circulation. The second stage is the formation of a bidirectional (supplying both right and left lungs) cavopulmonary shunt, also known as a Glenn shunt. This procedure is usually performed at 3–5 months of age and connects the superior vena cava to the right PA (RPA). Any residual BT or other shunts are removed or ligated. The child remains cyanosed after this procedure (oxygen saturations 75–85%). The third stage is the formation of a total cavopulmonary connection or the Fontan circulation. The inferior vena cava is

connected to the RPA, thereby separating the pulmonary and systemic circulation and normalizing arterial oxygenation. This is usually performed between 3 and 5 yr of age. The pressure gradient from the PA to the LA is now the sole determinant of PBF.

In the single-ventricle circulation, increases in PVR and intrathoracic pressure can compromise PBF. This has implications for ventilatory strategy. Spontaneous breathing causes negative intrathoracic pressures and augments PBF; however, positive pressure ventilation may allow greater control of oxygenation and minute ventilation, thus avoiding hypoxia and hypercapnia. Positive end-expiratory pressure should be optimized; peak inspiratory pressures and inspiratory times minimized to facilitate PBF.

### Risk classification

Children with heart disease undergoing non-cardiac surgery are at increased risk of mortality and morbidity. However, the range of heart disease and a variety of non-cardiac procedures make risk stratification difficult. Different studies identify a variety of factors associated with a high risk of perioperative complications, such as disease complexity, physiological status, type of surgery, and young age. The most important factors are the physiological status and complexity of heart disease since previous corrective cardiac surgery, whether complete or palliative, does not significantly alter the postoperative outcome.<sup>8</sup> Physiologically well-compensated patients with CHD can undergo elective operations at a low operative risk, whereas poorly compensated patients undergoing urgent or major operations are at high risk.<sup>8</sup> Therefore, in order to provide a practical and structured approach to management, we have classified children into high-, intermediate-, and low-risk groups (Table 1). Individual risk factors are discussed below.

### Physiological status

Physiological status can be divided into four major risk factors: cardiac failure, PHT, arrhythmias, and cyanosis.

#### Cardiac failure

The signs and symptoms of cardiac failure differ with age. Common to all ages include tachypnoea, tachycardia, sweating, and cool peripheries. In infancy, additional signs include poor feeding, failure to gain weight, and hepatomegaly. Cardiac failure can be due to a volume-overloaded heart, pressure-overloaded heart, or both. Volume overload may result from residual shunts or incompetent valves (as is common after tetralogy of Fallot repair) and pressure overload from residual outflow tract obstruction. Children with severe cardiac failure must be identified because they are at very high risk.<sup>5–9</sup> A recent study of children with cardiac failure undergoing non-cardiac surgery or investigations under general anaesthesia showed 10% suffered cardiac arrest and 96% required perioperative inotropic support.<sup>9</sup> Therefore, these children should be transferred to a specialist centre even for minor procedures. In the emergency situation, children may need retrieval

**Table 1** Risk classification of children with heart disease undergoing non-cardiac surgery

High risk	Intermediate risk	Low risk
Physiologically poorly compensated and/or presence of major complications (a) Cardiac failure (b) Pulmonary hypertension (c) Arrhythmias (d) Cyanosis	Physiologically normal or well compensated	Physiologically normal or well compensated
Complex lesions (single-ventricle or balanced circulation physiology, cardiomyopathy, aortic stenosis)	Simple lesions	Simple lesions
Major surgery (intraoperative, intrathoracic, anticipated major blood loss requiring transfusion)	Major surgery (intraoperative, intrathoracic, anticipated major blood loss requiring transfusion)	Minor (or body surface) surgery
Under 2 yr old	Under 2 yr old	Over 2 yr old
Emergency surgery	Emergency surgery	Elective surgery
Preoperative hospital stay more than 10 days	Preoperative hospital stay more than 10 days	Preoperative hospital stay less than 10 days
ASA physical status IV or V	ASA physical status IV or V	ASA physical status I–III

by a specialist transport team. In the rare event of transfer being impossible, advice should be sought from the tertiary centre paediatric cardiology and paediatric cardiac anaesthesia services.

Children with milder degrees of cardiac failure pose less risk. In the local hospital setting, either gaseous or i.v. induction is possible but induction times will be prolonged, so patience is needed to prevent excessive drug administration. Avoid prolonged use of 8% sevoflurane. Propofol may cause a profound decrease in cardiac output and is best avoided. Ketamine is the i.v. agent of choice.<sup>7</sup> Venous access may be difficult so having a second experienced anaesthetist is often helpful.

### Pulmonary hypertension

PHT is defined as having a PA pressure (PAP) >25 mm Hg at rest or 30 mm Hg during exercise. Documented PHT is a clear predictor of perioperative morbidity.<sup>6,10,11</sup> These children are eight times more likely to experience a major complication. Treatment with 100% oxygen, inhaled nitric oxide, i.v. prostacyclin, inotropic support of the right ventricle, and other measures to maintain cardiac output and PBF may all be required. Therefore, if a child is receiving treatment for PHT, they should be transferred to a specialist centre where full paediatric intensive care facilities are available.

Children with PHT also have reduced pulmonary compliance and increased airway resistance causing increased work of breathing. Therefore, respiratory tract infections may be poorly tolerated and have a greater impact on PVR than ordinarily expected.

### Arrhythmias

All children with CHD should have a preoperative ECG. Right bundle branch block is common but unlikely to degenerate into complete heart block. However, ventricular ectopics (VEs) are an ominous sign, as ~30% of children with VEs die suddenly.<sup>12</sup> Furthermore, arrhythmias leading to death arise in ~30% of patients with a single-ventricle circulation. Therefore, children

with VEs on ECG or a single-ventricle circulation should be referred for surgery in a specialized centre with onsite access to paediatric cardiology and paediatric intensive care facilities.

Children who have undergone ventriculotomy or had a right ventricle to PA conduit formed are more at risk of ventricular arrhythmias than other groups of children. If these children have a normal preoperative ECG, they would be classed as low risk (Table 1) and can be anaesthetized in the local hospital.

### Cyanosis

Cyanosis is a common feature of unrepaired or partially palliated CHD. Cyanotic children often have concurrent cardiac failure, PHT, and arrhythmias, making them a very high-risk group. Therefore, most children with cyanotic disease should be transferred to a specialist centre. However, in some situations, it may be acceptable for some cyanotic children without additional complications to undergo minor surgery or investigative procedures in the local setting provided the anaesthetist and other theatre, recovery, and ward staff understand the physiology and perioperative problems relating to cyanosis.

Chronic cyanosis affects most major organ systems. The main problems relating to anaesthesia and surgery are polycythaemia and coagulopathy. Children under 5 yr of age are at risk of hyperviscosity causing cerebral vein and sinus thrombosis. Dehydration, fever, and iron deficiency anaemia increase this risk. Preoperative i.v. fluid therapy may be used to minimize risk. Abnormal laboratory tests of haemostasis are also present in 20% of children with cyanosis. If a child is receiving aspirin, the risk of thrombosis is usually greater than that of bleeding so aspirin therapy should continue. If in doubt contact the child's cardiologist.

### Complexity of heart disease

Children with complex disease are at increased risk. In the context of perioperative complications, complexity has been defined as:<sup>5</sup>

- (i) single-ventricle physiology
- (ii) balanced circulation physiology
- (iii) cardiomyopathy
- (iv) aortic stenosis

The presence of long-term sequelae (cardiac failure, PHT, arrhythmias, and cyanosis) may also be considered as defining ‘complex’ disease.

## Type of surgery

The mortality of children with heart disease undergoing major surgery is 16% compared with 3% for minor surgery.<sup>3</sup> Major surgery is classified as intraperitoneal, intrathoracic, or vascular reconstructive surgery. Furthermore, the most common cause of cardiac arrest in non-cardiac surgery is hypovolaemia including the consequences of massive blood transfusion.<sup>4,13</sup> If cross-matched blood (not simply a ‘group and save’) is required for an operation, this implies a risk of haemorrhage and major blood loss. Therefore, we have cautiously classified this as major surgery (Table 1).

Most local hospitals would not undertake major surgery in children. However, not all teaching hospitals that perform major paediatric surgery have onsite access to paediatric cardiology, cardiac anaesthesia, and intensive care. Whether children with CHD should undergo surgery in this environment is unknown. However, given that major surgery increases perioperative risk from 3% to 16%, we suggest children with physiologically well-compensated disease or simple disease undergoing major surgery present intermediate risk. The presence of additional risk factors such as age under 2 yr or prolonged hospital stay may be additive risk factors. Therefore, we recommend that such children are at the very least discussed with a specialist centre and serious consideration given to transferring the child.

## Other

Other incremental risk factors for mortality in children with CHD undergoing non-cardiac surgery are emergency surgery,

preoperative hospital stay of 10 days or more, ASA physical status IV or greater, birth at a tertiary centre (reflects co-existing morbidity or complexity of cardiac disease), and age under 6 months.<sup>3</sup> Other studies suggest that children even up to 2 yr of age are still at significant risk.<sup>5</sup>

A summary and stratification of the risk factors described above is shown in Table 1.

## Preoperative assessment

A thorough understanding of the underlying CHD, including anatomy, physiology, and identification of risk factors, is vital before anaesthetizing these children. If any high-risk factors are present (Table 1), the child should be transferred to a specialist centre because of the high likelihood of requirement for intensive care and cardiology support. Depending on the locally available facilities and expertise, intermediate-risk children should be discussed with the specialist centre and consideration given to transfer. A child deemed at low risk can be anaesthetized safely in the local hospital setting. Transfer of these children is unnecessary and places undue burdens on the family (e.g. cost associated with travel, lack of social support networks, etc.) and creates extra unnecessary work for the specialist centre.

The specific issues to be addressed when taking a history and examining the child are shown in Table 2.

## Suggested management for elective procedures

High risk: transfer to specialist centre.

Intermediate risk: discuss with specialist centre and consider transfer.

Low risk: manage in local hospital.

## Suggested management for emergency procedures

High and intermediate risk: seek advice from the paediatric intensive care transport team and paediatric surgeons about feasibility of transfer. If transfer is impossible, seek advice from specialist centre cardiologist and paediatric cardiac anaesthetist regarding

**Table 2** Key features in preoperative assessment

1. *Knowledge of the underlying lesion and type of circulation?* Are changes in SVR/PVR likely to be of significant importance? Are the oxygen saturations what is to be expected for the type of lesion?
2. *Evidence of long-term complications* and other features that put children into a high-risk category (Table 1)
3. *Evidence of recent upper or lower respiratory tract infections:* this may cause changes in airway reactivity and PVR which may be poorly tolerated in children with reduced pulmonary compliance or PHT and particularly those with a Glenn or Fontan circulation
4. *Venous access:* may be problematic and alter the choice of anaesthetic technique. Many children have had multiple peripheral and central venous lines in the past
5. *Routine drug therapy:* most cardiac medications should be continued before operation. Some anaesthetists prefer to omit ACE inhibitors based on adult literature, but evidence in children is lacking. Cardiac medications are not associated with electrolyte disturbances so routine preoperative blood testing is unnecessary. Aspirin should be continued to prevent shunt thrombosis, and children on warfarin need admission for monitoring and establishment on i.v. heparin
6. *Sedative premedication:* commonly used to avoid distress, minimize oxygen consumption, and may also reduce the amount of induction agent so minimizing reductions in SVR
7. *Endocarditis prophylaxis:* national guidelines must be followed

**Table 3** Anaesthetic management: three specific examples

<p>1. 6 month old with an unrepaired VSD presenting for emergency surgery for a scrotal swelling</p> <p><i>Physiology:</i> compensated (on examination in moderate cardiac failure, on diuretics but feeding well and gaining weight)</p> <p><i>Lesion:</i> simple. Awaiting VSD repair next month</p> <p><i>Risk category:</i> intermediate (physiologically well-compensated, simple lesion, minor surgery, but less than 2 yr old, emergency surgery)</p> <p><i>Management:</i> depends on local anaesthesia and surgical expertise—if local hospital routinely performs surgery on children under 2 yr then safe to undertake locally. Otherwise, consider discussion with and transfer to specialist centre</p> <p><i>Anaesthesia considerations:</i> slow cautious gas or i.v. induction, beware of possibility of poor cardiac reserve. Avoid high inspired oxygen concentration in order to minimize left-to-right shunt and maximize systemic perfusion. May require higher ventilator pressures than expected because of pulmonary oedema. Avoid air bubbles in venous lines because of the risk of paradoxical embolus. Endocarditis prophylaxis unnecessary</p>
<p>2. 6 yr old with a repaired tetralogy of Fallot presenting for dental extractions due to dental caries</p> <p><i>Physiology:</i> compensated (on examination good exercise tolerance, no signs of right heart failure. ECG shows RBBB and no VEs)</p> <p><i>Lesion:</i> simple</p> <p><i>Risk category:</i> low (physiologically well compensated, simple lesion, minor surgery, over 2 yr old, elective surgery)</p> <p><i>Management:</i> perform procedure in local hospital. If VEs present on ECG—consider to be high risk and seek cardiology advice. May need to postpone surgery or transfer to specialist centre as high risk of life threatening arrhythmias</p> <p><i>Anaesthesia considerations:</i> gas or i.v. induction. No need for endocarditis prophylaxis.</p>
<p>3. 8 yr old with a Fontan circulation presenting for emergency surgery for reduction and fixation of a supracondylar fracture with neurovascular compromise</p> <p><i>Physiology:</i> may be well-compensated working single-ventricle circulation</p> <p><i>Lesion:</i> complex single ventricle</p> <p><i>Risk category:</i> high (physiologically well compensated, but complex single-ventricle lesion, emergency surgery)</p> <p><i>Management:</i> discuss with specialist centre paediatric cardiac anaesthetist. Transfer to specialist centre risks limb loss so may not be possible. Needs discussion on individual basis to balance risk of loss of limb vs loss of life if local hospital anaesthetist does not understand complex single-ventricle physiology</p> <p><i>Anaesthesia considerations:</i> gas or i.v. induction, consider spontaneous ventilation as this aids PBF but must avoid hypoxia, hypercarbia and atelectasis which increase PVR and compromise PBF. Full stomach may require rapid sequence intubation and positive pressure ventilation (PPV). If using PPV avoid high pressures, high PEEP, and long inspiratory times all of which compromise PBF. Slight head-up position aids venous return and PBF as does raising the legs. Endocarditis prophylaxis unnecessary</p>

perioperative management. Transfer the child as soon as they are stable enough.

Low risk: manage in local hospital. If concerns seek advice from specialist centre.

### Specific anaesthetic management

Many different anaesthetic techniques have been described, but there is little evidence to recommend one technique over another.<sup>7</sup> Propofol and ketamine are the two most studied agents in children with CHD undergoing non-cardiac surgery. Propofol dramatically reduces SVR and mean arterial pressure (MAP). In children with right-to-left shunt lesions, propofol may cause a clinically significant reduction in oxygen saturation by increasing the shunt flow.<sup>14,15</sup> Ketamine has minimal effect on SVR, MAP, PVR, and PAP, making it the agent of choice when a reduction in SVR is undesirable or in children with PHT. Gaseous induction with sevoflurane is common in paediatric practice and can be used for children with CHD. In high-risk children, avoid prolonged inspired concentrations of 8%, and patience and lower concentrations will often suffice. Consider obtaining i.v. access before induction or having an assistant who is experienced at paediatric i.v. cannulation (either a second consultant anaesthetist or the paediatric senior specialist trainee).

The use of isoflurane and sevoflurane for maintenance of anaesthesia is widely reported. Both agents have minimal effect on myocardial contractility or shunt fraction. The effects of desflurane in children with CHD are less well known. We recommend using the agent one is most familiar with. Propofol infusions are best avoided for reasons outlined above. Opioid infusions and spinal and epidural anaesthesia have also all been used successfully and

their use should depend on the usual practice for the type of non-cardiac surgery being performed.

Table 3 shows three examples of how understanding the physiology, identifying risk factors, and knowledge of anaesthetic pharmacology can be used in planning perioperative management.

### Summary

Children with CHD who present for non-cardiac surgery are at increased risk of perioperative morbidity. High-risk children require transfer to a specialist centre because full paediatric intensive care and cardiology services may be required. Depending on the locally available facilities and expertise, intermediate-risk children should be discussed with the specialist centre and consideration given to transfer. Low-risk children may undergo surgery in the local hospital. All anaesthetists responsible for children with CHD need to understand the anatomy, physiology, and risk factors associated with perioperative morbidity; be able to perform a thorough preoperative assessment; and have a knowledge of anaesthetic pharmacology in relation to the abnormal physiology likely to be encountered when anaesthetizing these children.

### Declaration of interest

None declared.

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Please see multiple choice questions 13–16.