Clinical Review

Update in
Anaesthesia

Anaesthesia in children with congenital heart disease for non-cardiac surgery

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doi:10.1029/WFSA-D-20-00004

Abstract

There is an increasing prevalence of children with congenital heart disease (CHD) presenting for non cardiac surgery due to advances in modern medicine which means more children with CHD are surviving for longer and are thus exposed to the usual illnesses and injuries of childhood. The complex nature of some cardiac lesions, and the frequent association of CHD with other congenital abnormalities means that the perioperative care of these children presents unique challenges for the anaesthetist. This article aims to review the perioperative considerations of children with CHD presenting for non-cardiac surgery. We provide a classification of CHD based on physiology, identify important factors for consideration during risk stratification and discuss an evidence-based approach to perioperative management.

INTRODUCTION

With advances in both diagnostic and interventional medicine, more children with congenital heart disease (CHD) are surviving. In the UK, 1 in every 180 children is born with CHD, and between 2000 and 2010 the number of operations for CHD increased by 80%.[1] This means increasing numbers of children with CHD are presenting for non-cardiac surgery because they are subject to the same range of illness and injuries as other children. Children with CHD present a unique set of challenges and can be a source of anxiety for non-paediatric anaesthetists. A one-size-fits-all approach to anaesthesia is impossible because of the wide spectrum of disorders and varying risk profiles.

This article aims to review the classification of CHD, outline the associated risks, and present an evidence-based approach to the anaesthetic management of children with CHD presenting for non-cardiac surgery.

Limitations: The term CHD is used very broadly and generally includes children born with structurally abnormal hearts, but can also include children born with structurally normal hearts but abnormal electrophysiological pathways causing congenital arrhythmias, for example Wolf-Parkinson White syndrome. Congenital arrhythmias are a separate group of congenital heart diseases with their own unique classifications and treatment pathways, which merit a review of their own. Therefore they are outside the scope of this article and will not be discussed.

CLASSIFICATION OF CHD

Congenital syndromes may also be associated with congenital heart disease and it is important for the anaesthetist to be aware of these syndromes and the cardiac malformations associated with these. For example, children with Down's syndrome commonly present for adenotonsillectomy and 40% of children with Down's syndrome have CHD (most likely ventricular septal defects and atrio-ventricular septal defects).[18] Other examples of congenital syndromes associated with congenital heart disease are discussed in the pre-assessment section of this article.

Children with CHD may be classified anatomically or physiologically. In this article we use a physiological classification based on circulation type. See Table 1.

Normal or 'series' circulation

The normal heart (Figure 1) has two separate circulations: a pulmonary (right sided) and systemic (left side) circulation which work together in series. Examples of CHD with a normal circulation are valvular disorders (for example: aortic stenosis, parachute mitral valve, Ebsteins anomaly) or cardiomyopathies which may be hypertrophic, dilated, restrictive or a combination or one or more types.

Some forms of CHD have a 'normal' circulation but the pulmonary and systemic circulations are not entirely separate, instead mixing of oxygenated and deoxygenated blood occurs through one or more 'holes' (e.g. VSD).

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Table 1: Physiological classification of CHD

Normal Circulation	Balanced/Parallel Circulation	Single Ventricle Circulation
• ASD • VSD • Tetralogy of Fallot • Valvular disorders e.g. AS, Ebstein's anomaly, • Cardiomyopathies • Coarctation of the aorta (CoA)	• Truncus Arteriosus • Transposition of the great arteries • Hypoplastic left heart syndrome • Large unrestricted VSD • Large unrestricted AVSD • Duct dependent lesions such as pulmonary atresia with intact ventricular septum • Children with a BT shunt (if the shunt is large these children will exhibit a balanced circulation)	Children who have had a Glenn or Fontan procedure A Glenn or Fontan procedure may be undertaken for a wide range of underlying conditions but commonly for: • Hypoplastic Left Heart syndrome (HLHS) • Other conditions where there is significant ventricular imbalance such that one of the ventricles would be incapable of maintaining a sufficient output, examples may include: Double Outlet Right Ventricle (DORV) • Pulmonary atresia (PA) with an intact intraventricular septum • Tricuspid Atresia (TA)

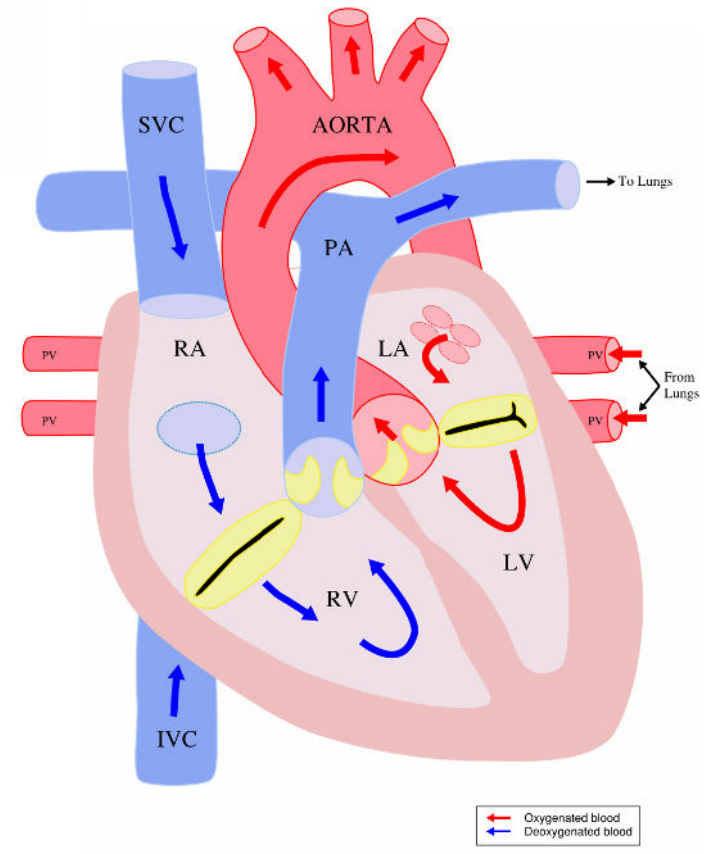

Figure 1: Normal circulation

The direction blood flows through the lesion depends on the pressure gradient and is documented in the echocardiography report as a shunt. Left-to-right shunts result in increased pulmonary blood flow and potentially decreased systemic blood flow; right-to-left shunts cause deoxygenated blood to flow into the systemic circulation, causing cyanosis and reduced pulmonary blood flow.

The direction and amount of shunting depends on the pressure gradient and size of the defect. When the size of the defect is small, this provides a significant resistance and limits the 'shunting' of blood. When the defect is very large, blood may move freely from left-to-right and back again depending on the relative balance between the systemic (SVR) and the pulmonary (PVR) vascular resistance. In this situation infants may exhibit what is known as a 'balanced' circulation physiology – see below. Changes in SVR and PVR as a result of anaesthesia, including the administration of oxygen, have the greatest effect on large, unrestrictive defects i.e. children with 'balanced' circulation (see below).

Parallel or 'balanced' circulation

Instead of the pulmonary and systemic circulations being separate entities working in series, they function physiologically as one entity and behave as parallel circuits. This means that oxygenated and deoxygenated blood mix freely, and the blood flow to the systemic and pulmonary circulation depends on the relative resistance in each circuit. Thus blood flow to the lungs and body is a 'balance' between SVR and PVR, in other words, where the blood goes depends on the ratio of SVR to PVR. This concept is known as Qp:Qs (where 'Q' means flow). A cardiology assessment may report Qp:Qs is 3: 1 which means that 3 times as much blood is flowing to the pulmonary circulation compared with the systemic circulation. Excessive pulmonary blood flow (PBF) causes pulmonary oedema and poor systemic perfusion (which may compromise coronary and splanchnic perfusion) whereas insufficient PBF causes profound cyanosis. The balance is generally high pulmonary flow (resulting in high oxygen saturations, pulmonary oedema, increased work of breathing and risk of high output cardiac failure) and low systemic flow (with associated risks of low diastolic blood pressure and coronary ischemia, and/or poor gastro-intestinal perfusion and necrotising enterocolitis).

Examples of CHD with 'balanced' circulation physiology include large unrepaired unrestricted ventricular septal defects (VSD) (Figure 2), large atrio-ventricular septal defects (AVSD), a modified Blalock-

Taussig (BT) shunt, truncus arteriosus (TA) and hypoplastic left heart syndrome (HLHS). These infants generally have predominantly left-to-right shunt flow because the SVR is higher than the PVR but the circulation is precariously 'balanced' and inducing anaesthesia in these children must be slow and careful to maintain the 'balance': high concentrations of oxygen will increase PBF and reduce systemic perfusion especially coronary artery perfusion; conversely large doses of induction agent reduce SVR causing increased systemic blood flow which compromises PBF and can lead to profound desaturation. These children can be very difficult to manage and liaison with a specialist paediatric cardiac centre is advised.

Single ventricle circulation

Some forms of CHD are not amenable to full anatomical correction i.e. 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, whilst blood flows passively to the lungs down a pressure gradient. A single ventricle circulation is usually created as a two or three staged process:

Stage 1 (some children): If an infant is very cyanotic due to critically low PBF, they will require augmentation of PBF in the first few days of life, commonly via a BT shunt. A BT shunt consists of a small (3-3.5mm) gortex tube usually positioned between the right subclavian artery (RSCA) and right pulmonary artery (RPA).

Stage 2 (all children): formation of a bidirectional (supplying both right and left lungs) cavopulmonary shunt (BCPS), also known as a Glenn shunt. Usually performed at 3 – 5 months of age, this connects the superior vena cava (SVC) to the right pulmonary artery (RPA).

Any residual BT or other shunts are removed or ligated. The child remains cyanosed following this procedure (oxygen saturations 75-85%).

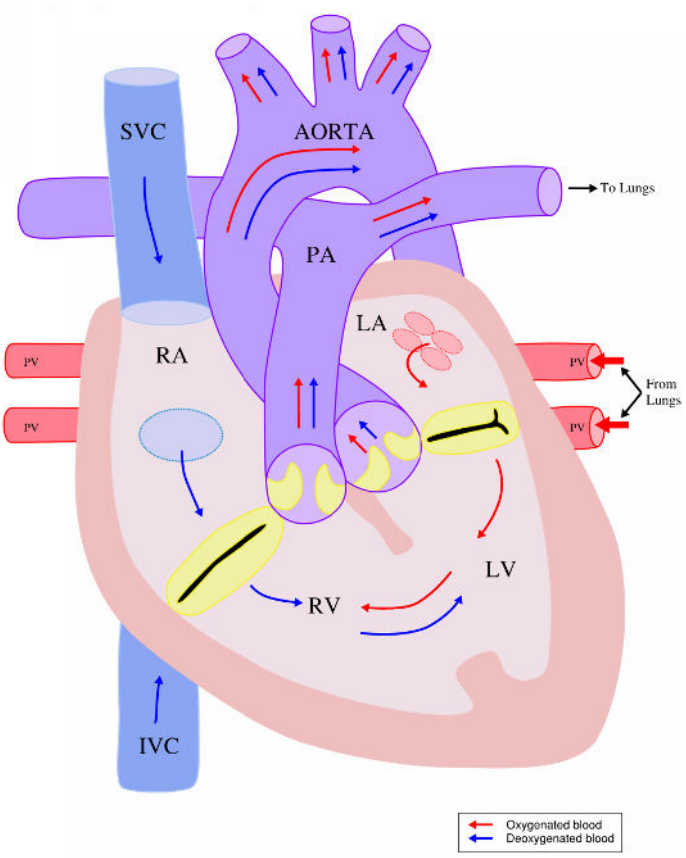

Figure 2: Balanced (or parallel) circulation e.g. large ventricular septal defect

Table 1: Physiological classification of CHD

Balanced/Parallel Circulation	Single Ventricle Circulation
Complexity of CHD	• Single ventricle physiology • Balanced circulation • Cardiomyopathy • Aortic Stenosis
Physiological status of the child e.g. presence of complications of CHD	• Cardiac failure • Pulmonary hypertension • Cyanosis • Arrhythmias
Type of surgery required	• Emergency surgery • Major surgery e.g. intrathoracic, intraperitoneal, vascular reconstructive surgery) • Elective surgery with high risk of major blood loss e.g. (orthopaedic and neurosurgery)
Age of the child	• Under 2 years of age
Hospitalisation of the child before the need for non-cardiac surgery	• Pre-operative hospital stay of 14 days or more

Stage 3 (all suitable children): formation of a total cavopulmonary connection (TCPC) or Fontan circulation (Figure 3). The inferior vena cava (IVC) is connected to the RPA thereby separating the pulmonary and systemic circulation and normalizing arterial oxygenation. This is usually performed between 3 - 5 years of age. The transpulmonary pressure gradient which is from the pulmonary artery (which is under low pressure coming from the superior and inferior vena cava) to the common atrium is now the sole determinant of PBF. The transpulmonary gradient is influenced by volume, pressure, resistance and compliance. Therefore, the single ventricle circulation is very sensitive to increases in volume status of the child, PVR and intrathoracic pressure which can compromise PBF.

This has implications for ventilatory strategy where the pros and cons of spontaneous ventilation versus positive pressure ventilation need to be carefully evaluated. Spontaneous ventilation causes negative intrathoracic pressures thereby improving PBF; positive pressure ventilation may reduce PBF, but does allow greater control of oxygenation and minute ventilation, thus avoiding hypoxia and hypercapnia. Positive end-expiratory pressure should be optimized; and peak inspiratory pressures and inspiratory times minimised to facilitate PBF.

In order to progress to a Fontan circulation certain criteria must be met which include but are not limited to: a low PVR, suitable single ventricular function, and minimal valvular regurgitation. If these criteria are not met then children not suitable for a Fontan may remain 'stuck' at stage 2.

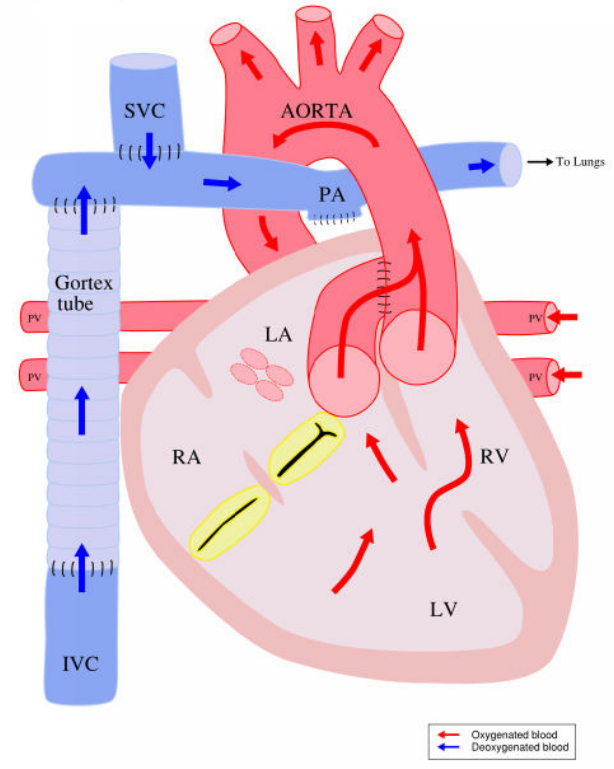

Figure 3: Single ventricle circulation e.g. after total cavopulmonary connection (Fontan) for palliative correction of a double inlet left ventricle lesion with transposition of the great arteries, which is the commonest form of single ventricle.

RISK STRATIFICATION OF CHILDREN WITH CHD UNDERGOING NON-CARDIAC SURGERY

Risk stratification for children with CHD undergoing non-cardiac surgery can be challenging and requires an understanding of the complexity of the cardiac lesion and the functional status of the child at the time of surgery. Some children with corrected CHD lesions have minimal risk of adverse events i.e. similar to children without CHD[2]; while other children with complex CHD are at significant risk of morbidity and mortality when undergoing non-cardiac surgery.[3] The most important risk factors are the complexity of CHD, the physiological status, age, and the type of surgery. The individual risk factors that have been identified are listed in Table 1 and discussed in detail below.

Complexity of heart disease

The following four categories of CHD are associated with a significantly increased risk of adverse perioperative events: single ventricle physiology, balanced circulation physiology, dilated or restrictive cardiomyopathies and aortic stenosis leading to left ventricular outflow tract obstruction. All of these conditions leave the child with significantly limited reserve. Careful risk to benefit analysis should be done prior to proceeding with an anaesthetic/operation in these cases.[2-10, 18]

Physiological Status:

If the child is physiologically well and asymptomatic this conveys a lower risk than children who have developed sequelae leading to poor physiological compensation from their cardiac lesion. The presence of any of the following long-term sequelae of CHD indicate a child is at high risk of perioperative adverse events: cardiac failure, pulmonary hypertension, arrhythmias and cyanosis.

Cardiac Failure

Cardiac failure results from either pressure or volume overload on the heart. Pressure overload may be caused by residual outflow tract obstruction and volume overload may result from ongoing shunts or incompetent valves.[2] Signs and symptoms of cardiac failure should be sought during pre-operative assessment as they have been shown to be associated with a high risk of adverse events during non-cardiac surgery in several studies.[3-7] Signs include sweating, tachypnoea, tachycardia and cool peripheries. In babies signs also include poor feeding, failure to thrive and hepatomegaly. Older children may complain of poor exercise tolerance, and may have signs of chest crackles and failure to gain weight.

The use of pre-operative angiotensin converting enzyme (ACE) inhibitors has been shown to be associated with haemodynamic instability on induction, while the use of pre-operative digoxin and/or inotropes is associated with haemodynamic instability during maintenance of anaesthesia.[8] A recent study by Lee et al[5] of over 3000 patients with congenital heart disease found that any form of depressed ventricular function was a risk factor for requiring inotropes during non-cardiac surgery. Murphy et al also found that 96% of patients with heart failure required inotrope use and 10% suffered cardiac arrest.[9] Given the severity of risk associated with anaesthesia in children with heart failure, we recommend that they are transferred to a specialist centre for even minor surgery.[10] If transfer is not possible, liaison with specialist paediatric cardiology

and paediatric cardiac anaesthesia services is advised. It should be noted that the time taken for both gaseous and intravenous induction will be prolonged and so considerable patience is necessary during induction to avoid excess drug administration.[2] Prolonged use of 8% sevoflurane and propofol may produce a profound decrease in cardiac output meaning that ketamine is generally a preferable option.[11]

Pulmonary Hypertension

Pulmonary hypertension (PHT) is defined as having a pulmonary artery pressure over 25mmHg at rest or 30mmHg on exercise.[2] PHT is a known risk factor for adverse outcomes following non-cardiac surgery.[2-8,12] In a study by Warner et al of 276 patients with CHD, those with a diagnosis of PHT had a higher complication rate (15%) than those with CHD without PHT (4.7%).[12] The pathophysiology and development of pulmonary hypertension varies with different lesions and management is complex and often includes: 100% oxygen, inhaled nitric oxide, intravenous prostacylin, inotropic support of the right ventricle and other measures to maintain cardiac output and pulmonary blood flow (PBF). Therefore if children who are receiving treatment for pulmonary hypertension require surgery, then they should be transferred to a specialist centre where paediatric cardiac intensive care is available.[10]

Cyanosis

Cyanosis is a common feature of unrepaired or palliated CHD. Chronic cyanosis (hypoxaemia) causes changes in the blood composition, coagulation profile, secondary erythrocytosis and hyperviscosity.[13] Altered coagulation is due to thrombocytopenia, decreased von Willebrand factor and a range of clotting factor deficiencies.[13] These haematological changes increase the risk of both thrombosis and infarction, and in children under 5 there is an increased risk of cerebral vein and sinus thrombosis. These risks are increased by dehydration, fever and iron deficiency. Treatment with intravenous fluids (or at a minimum encouraging oral fluid intake) may be initiated preoperatively to minimize these risks, however further treatment should be based on the advice of specialist paediatric haematology and cardiology services.

Arrhythmias

A pre-operative electrocardiogram (ECG) should be reviewed in all children with congenital heart disease. Previous surgery may have affected conduction pathways causing an increased risk of arrhythmias under anaesthesia. Up to 30% of children with single ventricle physiology will die from arrhythmias.[12] The pre-operative ECG may help to predict risk, for example right bundle branch block is common and unlikely to degenerate into complete heart block, while ventricular ectopics (VE's) are a worrying sign and are associated with an increased risk of sudden cardiac death.[12] Therefore, all arrhythmias, especially VE's should be discussed with a paediatric cardiologist and those deemed to be high risk should be transferred to specialist centres where appropriate paediatric cardiology and intensive care support is available.

Type of surgery

Mortality for children with CHD undergoing major surgery is significantly higher (16%) than for minor surgery (3%).[4] Major surgery is defined as intrathoracic, intraperitoneal or vascular reconstructive surgery. Lee et al also found orthopaedic and neurosurgery cases to be associated with higher risk of adverse events.[5] This may be because the commonest cause of cardiac arrest in non-cardiac surgery was due to hypovolaemia[14,15] and therefore if there is a risk of major haemorrhage from the surgery, this increases the risk of adverse events.

Other risk factors

Age: In a review of 372 cases with a background of CHD who had a cardiac arrest, 47% occurred in those less than 6 months of age and 70% occurred in those less than 2 years.[16] Other studies also identified young age as a risk factor for adverse events in those with CHD undergoing non-cardiac surgery.[6-7,14]

Pre-op Length of Stay: In a study by Watkins et al, in 145 patients with a preoperative stay >14 days (without intubation) 12.4% required mechanical ventilation post operatively, postoperative length of stay was also longer in those with a preoperative length of stay >14 days.[8]

Other: higher ASA physical status, birth at a tertiary centre (indicating co-existing morbidity or complexity of CHD), and emergency surgery are also independent risk factors for adverse perioperative events in children with CHD undergoing non-cardiac surgery.[2-9]

ANAESTHETIC MANAGEMENT

Pre-operative Assessment

Meticulous preparation and good communication with the multi-disciplinary team including the surgeon and the child's paediatric cardiologist are crucial to ensuring safe delivery of care. If the child presents to a non-specialist centre this may involve communication with several different hospitals. We suggest careful attention is paid to the following factors during pre-operative assessment:

a) Review the cardiac lesion and type of circulation. This should be done by looking at the child's latest cardiology letter, echocardiography (ECHO) report and ECG. Decide if the child is at high risk of adverse events during surgery and therefore requires transfer to a specialist centre.

Based on the evidence-based risk classification described above (Table 1), we suggest the following guidelines for consideration of transfer to a specialist centre. However, the availability of local services and expertise means that decisions will ultimately need to be made on a case-by-case basis:

- Elective Surgery:

High risk: Transfer to a specialist centre

Intermediate risk: Consider transfer to specialist centre: discuss between local team and specialist centre and if required the transfer team

Low risk: Manage in local hospital.

- Emergency Surgery

High and Intermediate risk: Discuss with specialist centre and transfer team whether a time critical transfer is possible. If this is not an option, seek advice from the specialist centres' surgical, anaesthetic and cardiology team regarding perioperative management.

Table 1: Factors affecting pulmonary and systemic vascular resistance

Increased PVR	**Decreased SVR**
Hypoxia, hypercarbia, acidosis, hyperinflation, high PEEP, increased haematocrit	Pyrexia, some induction agents e.g. propofol, general anaesthesia, sympathetic blocks e.g. epidural, spinal
Decreased PVR	**Increased SVR**
High FiO_2, hypocarbia, alkalosis, decreased haematocrit	hypothermia, vasoconstrictors, sympathetic stimulation e.g. pain

Low risk: Manage in local centre, if concerned contact specialist centre for advice.[17]

b) Review the child's latest ECHO report and:

i. Assess if there are any 'holes' in the heart through which shunting can occur. If so, which way is the blood shunting? Think through the physiology of the shunt: will changes to the SVR or PVR have haemodynamic consequences under anaesthesia? Are the oxygen saturations measured appropriate for the type of lesion present?

Also if there are any 'holes' through which shunting can occur, be very careful with air bubbles when injecting fluid / medication because of the risk of air emboli passing from the venous to systemic circulation through the 'hole' and causing arterial cerebrovascular air emboli.

ii. Assess if there is any left or right ventricular outflow tract obstruction. If so, think through the physiological response of the obstruction to changes in SVR and PVR under anaesthesia.

c) Does the child have chronic cyanosis? In these children it is important to avoid dehydration and consideration should be given to minimizing starvation times and the use of intravenous preoperative hydration.

d) Are there any signs of recent upper or lower respiratory tract infections: this may affect airway reactivity and PVR, as management of PVR is central to the perioperative management of children with CHD especially those with single ventricle physiology as mentioned below.[17]

e) Does the child have another associated abnormality/dysmorphic features? Recognisable chromosomal abnormalities are seen in 25% of children with CHD. Children with Trisomy 21 (Down's syndrome) may also have an AVSD or VSD, those with DiGeorge Syndrome (22q11 deletion) may show aortic arch abnormalities, VSD, those with Marfan's syndrome may suffer from aortic root dilatation and dissection, those with Goldenhar syndrome or VACTERL (a syndrome consisting of Verterbral, Ano-rectal, Cardiac, Trachea-Eosophageal, Radial and Limb abnormalities) association may have a VSD or tetralogy of Fallot and those with Apert syndrome may have pulmonary stenosis or a VSD.[18]

f) Premedication: sedative premedication may be beneficial as this can help with anxiolysis, therefore avoiding catecholamine release, which cause tachycardia (thereby worsening any right or left sided stenotic lesion), increase oxygen consumption (which can worsen cyanosis). Premedication may also reduce the amount of induction agent required therefore minimizing the reduction in SVR associated with induction of anaesthesia.

g) Venous Access: may be challenging in children who are on diuretics, or who have had multiple procedures, involving peripheral and central venous catheterisation and arterial catheterisation.

h) Does the child require endocarditis prophylaxis for the procedure? The most recent recommendations from the NICE 2008 guidelines 'Prophylaxis against infective endocarditis' recommends that those at risk of infective endocarditis include those with

- valve replacements,
- structural congenital heart disease, including surgically corrected or palliated structural conditions, but excluding isolated ASD, Fully repaired VSD or fully repaired PDA, and endothelialised closure devices.
- Previous infective endocarditis
- Hypertrophic cardiomyopathy[19]

Specific Anaesthetic Management

There are several techniques described for anaesthetising children with CHD, however, there is no strong evidence to recommend one technique over another. It is advisable to consider which factors affect pulmonary and systemic blood flow (Table 3) and consider the cardiac lesion present. Due to the complexity of some children with congenital heart disease, it may be advisable to have a second consultant anaesthetist or an experienced senior trainee present for induction.

Gaseous induction with sevoflurane can be used safely in children with CHD and may be necessary due to challenging venous access. However, it is important to avoid prolonged exposure to 8% sevoflurane as this will cause a drop in SVR leading to decreased myocardial perfusion and contractility.[17] In those with trisomy[21] gaseous induction should be used with caution as bradyarrhythmias may occur.[20]

Propofol and ketamine are the most studied intravenous induction agents. Propofol produces a significant drop in SVR and mean arterial pressure (MAP). In children with right to left shunts, even a normal induction dose of propofol may increase the right to left shunt fraction causing reduced pulmonary blood flow and therefore decreased oxygen saturations.[11] Ketamine has minimal effect of SVR, PVR, MAP and PAP which makes it an ideal choice for many children with complex CHD and especially in those for whom the anaesthetist must avoid dropping the SVR.

Both isoflurane and sevoflurane are commonly used for maintenance of anaesthesia as they have little effect on shunt fraction[21] or cardiac contractility.[22] The effects of desflurane in children with CHD are less well known. Propofol infusions are likely to cause a reduction

in SVR and risk propofol related infusion syndrome, therefore they may be better avoided.

Opioid infusions and regional anaesthesia have all been successfully used[23,24] and therefore we suggest that the anaesthetist uses their preferred method of analgesia for the type of procedure being undertaken. As is the case in all children, spinal and epidural techniques may cause hypotension and the use of small doses of a vasopressor such as phenylephrine may be needed. In cases where reductions in SVR may be deleterious, the anaesthetist should be ready to treat hypotension promptly.

The use of invasive monitoring (central venous pressure monitoring and invasive arterial blood pressure monitoring) should be guided by the risk stratification: the higher the risk, the greater the need for invasive monitoring to allow inotrope delivery, intracardiac measurements and continuous blood pressure monitoring. However, less invasive monitoring such a near-infrared spectroscopy (NIRS), urinary catheters, intermittent capillary blood gas and lactate measurements also provide useful information of end organ perfusion and may be used as surrogates for tissue perfusion and adequate cardiac output either alone or in combination with invasive monitoring depending on the risk stratification.

SUMMARY

Anaesthesia for children with congenital heart disease for non-cardiac surgery requires a comprehensive understanding of the child's cardiac lesion and pathophysiology. Children with complex CHD (balanced or single ventricle circulation, aortic stenosis and cardiomyopathy) and poor physiological status are at the highest risk of perioperative morbidity and mortality. Poor physiological status is indicated by the presence of pulmonary hypertension, cardiac failure, arrhythmias and cyanosis. Other groups at high risk are children who are less than 2 years old, and those who require emergency surgery, or have been in hospital for over 12 days pre-operatively. High risk children for non-cardiac surgery should be transferred to a specialist centre whenever possible. Children at low to intermediate risk undergoing elective surgery may, depending on the locally available facilities and expertise, be operated on at the local hospital. Thorough pre-operative assessment, good communication with the multi-disciplinary team, both locally and at the specialist centre, and an awareness of the complications which may occur is essential for anaesthetising children with CHD for non-cardiac surgery.

ACKNOWLEDGEMENTS

The authors would like to thank Iain Moore for help drawing the figures.

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