Chapter

33 Anaesthesia for Children with Cardiac Disease Undergoing Non-cardiac Surgery

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Introduction

Congenital heart disease (CHD) is one of the most common birth defects, occurring in approximately one in 150 live births. Extracardiac anomalies requiring surgical intervention within the first year of life occur in 30% and are often associated with recognised syndromes (Table 33.1). Advances in surgical strategies and medical care mean that around 90% of affected children will survive to adulthood. These children experience the same childhood illnesses and injuries as those without CHD and may present to their local hospital requiring elective or emergency surgery. Overall, children with CHD undergoing non-cardiac surgery (NCS) are at increased risk of perioperative morbidity and mortality and can present a significant challenge to the anaesthetist. However, this increased risk does not extend to all children with CHD, only to defined higher-risk groups, and it is clear that with appropriate planning and expertise even those with significant disease can tolerate major surgery with few complications.

Aims and Limitations

This chapter aims to discuss the principles underlying the management of children with CHD: it includes risk stratification to distinguish those who would benefit from specialist care from those who may be safely managed in their local hospital. There are three main limitations. Firstly, few studies consider children with CHD undergoing NCS with much of the available information coming from cardiac catheterisation. The impact of other surgical procedures on risk is not completely characterised. Surgery involving changes in position (prone or Trendelenburg), major fluid shifts or laparoscopic approaches changing intracavity pressures may produce physiological effects that alter these risks. Secondly, most outcome data from which risk stratification systems are derived from large US databases that do not provide perioperative management details, including where the surgery was performed (large versus small hospitals, teaching versus peripheral) or who delivered the care (cardiac or general paediatric anaesthetist). Finally, children with CHD represent an extraordinarily heterogeneous group of patients. Furthermore, in a rapidly advancing field, observed complications may relate to outdated treatment strategies, making application to current practice difficult. As such, it is not possible to describe an anaesthetic technique suitable for all patients. The approach presented here is consequently a general one, based on applied physiology and informed by the available evidence where possible.

Pathophysiology of Different Circulations

When considering the child with CHD for NCS, a comprehensive understanding of the cardiac lesion is imperative. The likely effects of anaesthesia and surgery can then be anticipated. A simple classification of cardiac disorders is given in Table 33.2, and some common physiological patterns are now discussed.

Left-to-Right Shunts: 'In Series' Circulations

In some cardiac conditions, the dominant path of blood flow follows that of a structurally normal heart: separate systemic and pulmonary circulations operate in series. This is the situation for most types of repaired CHD. Unrepaired conditions that function 'in series' include atrial septal defects (ASD) ventricular septal defects (VSD) and patent ductus arteriosus (PDA). Blood flows through the defect down a pressure gradient, creating a left-to-right shunt, increased pulmonary blood flow (PBF) and volume overload of the right

Table 33.1 Congenital syndromes and associated cardiac lesions

Anomaly & genetic basis	CHD (%)	Typical cardiac lesion	Relevant clinical features
Down syndrome – trisomy 21	40-50	AVSD VSD	Cervical spine instability Prone to pulmonary hypertension Obstructive sleep apnoea
DiGeorge / velocardiofacial syndrome – 22q11 deletion	70-75	Interrupted aortic arch Tetralogy of Fallot Truncus arteriosus	Immune deficiency from absent thymus - irradiated blood, freq. chest infections Hypocalcaemia Cleft lip and/or palate
Turner syndrome – monosomy X	25-40	Coarctation of aorta AS / bicuspid Ao valve	Scoliosis Strabismus
Williams syndrome – 7q11.3 deletion	75-80	Supravalvar AS Pulmonary stenosis	Skeletal/joint abnormalities Dental abnormalities
Noonan syndrome	70-80	Pulmonary stenosis HOCM	Cervical spine: syringomyelia, Arnold Chiari Pectus carinatum/excavatum
VACTERL association	40-80	ASD / VSD Tetralogy of Fallot Truncus arteriosus	Tracheoesophageal fistula +/– oesophageal atresia Anal atresia Renal and vertebral anomalies
CHARGE association	75-80	Tetralogy of Fallot Various others	Choanal atresia Cranial nerve palsy IX/X: swallowing problems Cleft lip and/or palate
Goldenhar syndrome	30-60	Tetralogy of Fallot ASD / VSD	Hemifacial microsomia: difficult intubation Cleft lip and/or palate
Duchenne muscular dystrophy X-linked dystrophin gene	60-75	Cardiomyopathy	Respiratory muscle weakness Scoliosis

Notes: AVSD = atrioventricular septal defect; VSD = ventricular septal defect; AS = aortic stenosis; HOCM = hypertrophic cardiomyopathy; ASD = atrial septal defect.

ventricle. Potential consequences include pulmonary hypertension and cardiac failure. Structurally small defects are termed restrictive: the physiological effects are such that treatment can often be deferred. Larger 'unrestrictive' defects can lead to severe cardiac failure in infancy. The degree of shunt can be quantified using the Qp:Qs ratio (where Qp is pulmonary blood flow and Qs is systemic blood flow), estimated from two-dimensional echocardiography (ECHO). Elevation of the normal 1:1 ratio to 2:1 or above is usually associated with symptoms of heart failure.

Parallel or 'Balanced' Circulations

More significant anatomical defects allow mixing of oxygenated and deoxygenated blood and lead to systemic and pulmonary circulations operating in parallel. Examples include infants with large unrepaired atrioventricular septal defects (AVSD) and truncus arteriosus. Flow to the systemic or pulmonary vascular beds depends on the relative resistances of each circuit, so flow is dependent on the 'balance' between systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR). High flow to one area compromises flow

Table 33.2 Classification of congenital cardiac lesions

Type of disorder	Common examples
Acyanotic cardiac lesions	
Left-to-right shunts – excessive pulmonary blood flow	
'Restrictive' lesions	Small ASD, VSD, PDA
'Non-restrictive' lesions	Large VSD, PDA AVSD, AP window
Left-ventricular outflow tract obstruction	
	Coarctation of the aorta Interrupted aortic arch Aortic stenosis HOCM
Cyanotic cardiac lesions	
Right-to-left shunts – inadequate pulmonary blood flow	
	Tetralogy of Fallot Tricuspid atresia Pulmonary stenosis/ atresia
Complex shunts with obligatory common mixing	
	Transposition of the great arteries Hypoplastic left heart syndrome Truncus arteriosus TAPVD

Notes: PDA: patent ductus arteriosus, AP: aorto-pulmonary window; TAPVD = total anomalous pulmonary venous drainage.

Table 33.3 Factors affecting pulmonary blood flow

Factors reducing PBF	Factors Increasing PBF
Increased PVR	**Reduced PVR**
Hypoxia	High inspired oxygen
Hypercarbia	Hypocarbia
Acidosis	Alkalosis
Hyperinflation	Normal inflation volumes
↑ Haematocrit	↓ Haematocrit
Decreased SVR	**Increased SVR**
Pyrexia	Hypothermia
Anaesthetic agents:	Vasoconstrictors
- propofol	Sympathetic stimulation
- high dose volatiles	
Sympathetic block	

to the other. Excessive PBF causes pulmonary oedema and poor systemic perfusion and can lead to coronary and splanchnic ischaemia, whereas inadequate PBF causes cyanosis. Anaesthetic interventions can have a significant effect on blood flow in these circumstances. Oxygen is a potent pulmonary vasodilator and will increase PBF at the expense of systemic perfusion. Conversely, large doses of induction agents may reduce SVR sufficiently to reverse a left-to-right shunt and cause profound hypoxia. Factors affecting pulmonary and systemic blood flow are given in Table 33.3.

Duct-Dependent Circulations

The concept of a balanced circulation is particularly important in patients who are duct dependent. In some complex CHD, neonates rely on flow through the ductus arteriosus to maintain either systemic blood flow, such as hypoplastic left-heart syndrome (HLHS), interrupted aortic arch (IAA) or pulmonary blood flow, such as pulmonary atresia. An infusion of prostaglandin E is required to maintain ductal patency until surgery can occur. A well-balanced circulation produces saturations of 75–85%, with higher saturations indicating excessive PBF. Where systemic perfusion depends on the duct, ventilation in room air with moderate hypercapnia and respiratory acidosis may be required to increase PVR, reduce PBF and prevent systemic hypoperfusion, metabolic acidosis and possible cardiac arrest.

Single-Ventricle Circulations

Biventricular repair to create a normal 'in series' circulation is not always possible, so some affected children will be palliated with a circulation based on one functional ventricle. The functional single ventricle (SV) is utilised to pump blood around the body whilst pulmonary blood flow is passive down a pressure gradient from the pulmonary artery (PA) to the left atrium (LA). The archetypal SV circulation is HLHS, but there are a large number of cardiac defects that follow this pathway. Most children require a first-stage surgical intervention in the neonatal period, the nature of which is determined by the underlying lesion. For HLHS, this is the Norwood operation, utilising the right ventricle as the systemic SV, with the formation of a 'neo-aorta' from the anatomical hypoplastic aorta and the main

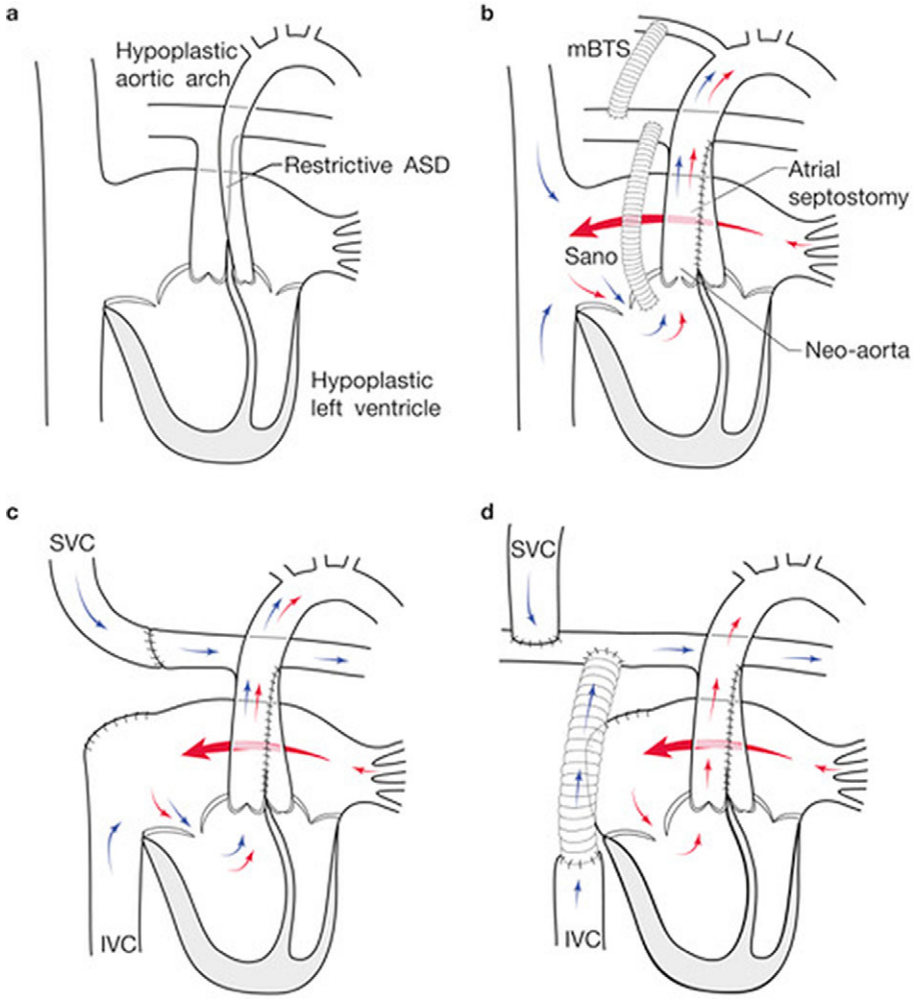

Figure 33.1 (a) Hypoplastic left-heart syndrome with a small left ventricle, hypoplastic aortic valve and arch and atrial septal defect. (b) Norwood procedure: anastomosis of the aorta to the pulmonary artery and atrial septostomy, ligation of PDA and formation of either a modified Blalock–Taussig (BT) shunt or Sano (right ventricle to pulmonary artery) shunt. (c) Bidirectional Glenn: connection of the superior vena cava (SVC) to the right pulmonary artery (RPA) and ligation of either a BT or Sano shunt. (d) Fontan: inferior vena cava (IVC) also connected to the RPA, shown here using the currently favoured extracardiac conduit.

pulmonary artery, and the creation of a modified Blalock–Taussig (BT) shunt or a Sano shunt to establish pulmonary blood flow (Figure 33.1). For other lesions, pulmonary artery banding or formation of a BT shunt may suffice in the first instance (Figure 33.2).

The second stage is common to all and involves the formation of a Glenn shunt at around three to five months (Figure 33.1c). This involves connecting the superior vena cava (SVC) to the right pulmonary artery (RPA), with the removal of any remaining shunts. Postoperatively, the child remains cyanosed (oxygen saturations 75–85%), and the ventricle remains volume loaded, receiving venous return from both inferior vena cava (IVC) and pulmonary veins. As the child grows, the proportion of venous return from the SVC reduces from the 60% seen in infancy towards adult values of around 35%, and pulmonary blood flow becomes inadequate. The third stage of total cavopulmonary circulation (TCPC) or Fontan procedure is performed between three to five years and involves connecting the IVC to the RPA, such that all systemic venous return now flows passively directly into the pulmonary circulation. A 'fenestration' may be created as part of the procedure; this is a surgical connection between the conduit connecting the IVC to the RPA and

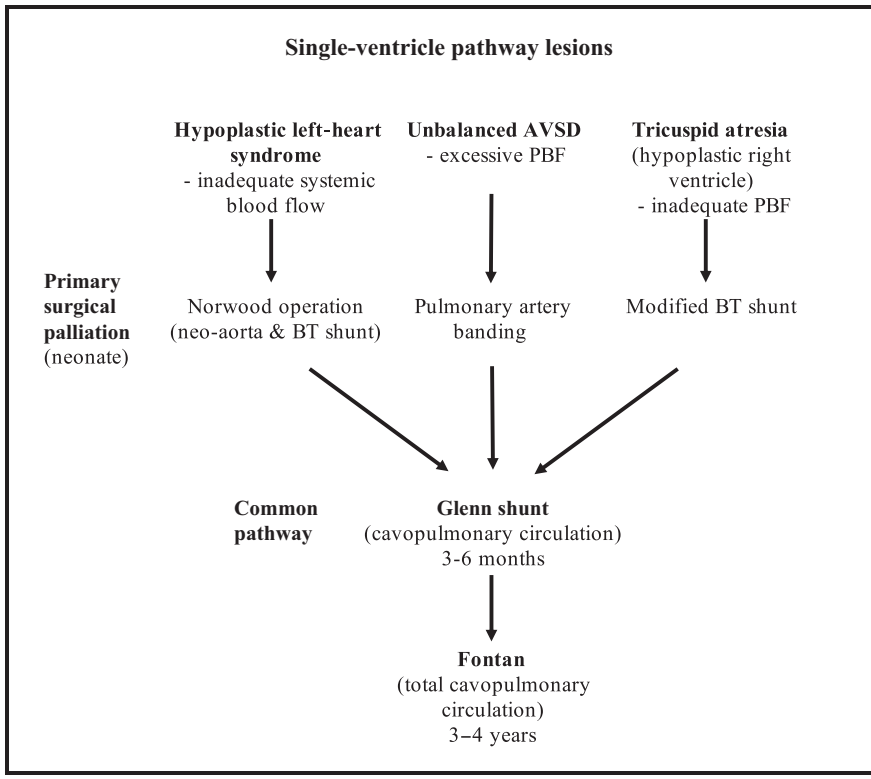

Figure 33.2 Spectrum of congenital cardiac lesions that are ultimately palliated with a single ventricle surgical strategy. AVSD: atrioventricular septal defect; BT: Blalock–Taussig.
Source: Reprinted with permission from Smith S, Walker A. Anaesthetic implications of congenital heart disease for children undergoing non-cardiac surgery. *Anaesthesia and Intensive Care Medicine* 2018; 19(8):414–20. doi: 10.1016/j.mpaic.2018.04.011.

the common atrium and can act as a 'pressure-relief' valve. If PVR is high and PA pressure is elevated, deoxygenated blood can bypass the lungs and enter the heart directly, ensuring that the systemic ventricle remains adequately volume loaded, maintaining cardiac output even in the presence of high PVR, but leading to desaturation. In general, children with an un-fenestrated Fontan should be almost fully saturated, but those with a fenestration may not be.

With single-ventricle physiology (SVP), increases in PVR and intrathoracic pressure associated with intermittent positive pressure ventilation (IPPV) can reduce PBF. The choice of ventilatory strategy represents a compromise. The advantages of spontaneous ventilation, which promotes PBF by generating negative intrathoracic pressure, should be set against the advantages of IPPV with respect to greater control in avoiding hypoxia and hypercapnia. When required, IPPV should be optimised with pressures and inspiratory times minimised to facilitate PBF.

Physiological Consequences of Heart Disease in Children

Cardiac Failure

Cardiac failure in children may be a consequence of structural CHD or due to an intrinsic disease of heart muscle (cardiomyopathy). Regardless of aetiology, the common end result of a volume- or pressure-overloaded ventricle is a failing heart that is unable to meet everyday physiological needs. Children with severely limited cardiac reserve function at or near maximal cardiac capacity even when resting and are reliant on sympathetic tone to maintain their cardiac output. Anaesthetic-induced reductions in sympathetic drive or SVR can produce significant haemodynamic compromise. Left-ventricular systolic dysfunction is defined as an ejection fraction (EF) of less than 55%. There are no formal definitions for degrees of dysfunction, but EF less than 40% should be considered moderate and less than 30% severe. A significant

degree of impairment may exist without symptoms, but when present symptoms of cardiac failure include tachypnoea, tachycardia, sweating episodes (e.g. when feeding), hepatomegaly and failure to thrive. Anaesthetic induction will be prolonged, whether intravenous or by inhalation, so patience is required to avoid excessive drug administration. These children represent a high-risk group and should be managed in a specialist centre wherever possible.

Cyanosis

Children with unrepaired or partially palliated CHD will commonly exhibit cyanosis, and oxygen saturations of 70–80% can be well tolerated for several years. Cyanosis is seen in cardiac conditions with right-to-left shunts and obligatory intracardiac mixing such as BT shunts and Glenn circulations. The main aim during anaesthesia is to maintain oxygenation at a level the child is accustomed to. In the presence of a structural shunt, it will not be possible to achieve 'normal' saturations, although it is relatively common to see an intraoperative rise in oxygen saturations with higher fractions of inspired oxygen and a fall in PVR. End-tidal CO_2 monitoring is inaccurate in cyanosed patients and reads up to 2 kPa less than arterial CO_2; however, the relationship is reasonably reliable, so end-tidal CO_2 can still be used as a proxy for ventilatory efficacy. The hypoxic ventilatory response is blunted in children with cyanosis, so the risks of postoperative respiratory depression are increased.

Chronic cyanosis affects most major organ systems, but the haematological effects are most pertinent to anaesthesia. Increased erythropoietin production causes increased haemoglobin, haematocrit and viscosity, maintaining oxygen delivery without elevating cardiac output. The haemoglobin level should not be allowed to fall below 110–120 g l^{-1}, with transfusion started early if there is surgical blood loss. However, a haematocrit over 65% can actually reduce oxygen delivery due to increased red cell rigidity. Hyperviscosity is associated with cerebral vein and sinus thrombosis and stroke; the risks are highest in children with fever, dehydration, iron deficiency and those under five years. Preoperative haemoglobin higher than 180 g l^{-1} (haematocrit $>$60%) should prompt consideration of intravenous fluid therapy to reduce this. Cyanosis is associated with an increased risk of bleeding via multiple effects on the clotting pathway, and this remains true even when preoperative clotting tests are normal. In children taking aspirin to maintain shunt patency, the risk of shunt thrombosis is usually greater than the risk of bleeding, so in most cases aspirin therapy should continue.

Pulmonary Hypertension

Pulmonary hypertension (PHT) is defined as a mean pulmonary artery pressure (PAP) above 25 mmHg at rest. Whilst over 50% of cases are idiopathic, PHT may develop secondary to a number of cardiac conditions. Children with CHD at risk include those with left-to-right shunts and excess PBF, such as AVSD, or those with prolonged pulmonary venous obstruction or high left-atrial pressure, for example following repair of total anomalous pulmonary venous drainage (TAPVD). Other causes include chronic lung disease, sickle cell disease or syndromic associations such as Down. There is a wide spectrum of severity, but PHT is a clear predictor of increased perioperative morbidity and mortality. Those with suprasystemic PAP are eight times as likely to experience a major complication as those with subsystemic PAP. The key principle of anaesthesia is to avoid a rise in PVR, which in turn may precipitate acute right-ventricular failure. Cardiac arrest in this setting is difficult to recover from. Factors which elevate PVR include acidosis, elevated $PaCO_2$, hypoxia, hypothermia, increased sympathetic stimulation (from inadequate anaesthesia) and increased airway pressure. Appropriate management of all these factors will reduce PVR, improve RV function and minimise any right-to-left shunting.

Arrhythmias

Children with CHD may be at risk of arrhythmias following surgical intervention, although this risk may not manifest until several years later. Atrial arrhythmias are most common following atrial septal/ sinus venosus defect repairs and atrial switch operations (Mustard and Senning). Conduction defects from damage to the atrioventricular node and bundle of His are commonest following ventriculotomy or right ventricle to pulmonary artery conduit; children with single-ventricle physiology or following tetralogy of Fallot or Ebstein's repairs are at particular risk. In addition to those associated

with structural CHD, there is a heterogeneous group of primary malignant arrhythmias, sometimes referred to as cardiac channelopathies, which predispose children to life-threatening perioperative events. This group includes long QT syndrome (LQTS), Brugada syndrome, arrhythmogenic right-ventricular cardiomyopathy (ARVC) and catecholaminergic polymorphic ventricular tachycardia (CPVT).

All children with CHD should have a preoperative ECG. Whilst right bundle branch block (RBBB) is a common and usually benign finding, ventricular ectopics are an ominous sign as 30% of these patients will eventually die suddenly. The optimal anaesthetic strategy is not well defined. In general, it is recommended that factors known to lower the threshold for ventricular ectopics are avoided; these include hypoxia, acidosis, hypercarbia and large doses of local anaesthetic with adrenaline. Prolongation of the QT interval is associated with polymorphic ventricular tachycardia (Torsades de Pointes) particularly in those with long QT syndromes. Both propofol and sevoflurane appear to have little effect on the QTc (QT interval corrected for heart rate), whereas other drugs prolong it and should be avoided, such as ondansetron, ketamine, suxamethonium and most adrenergic agents. Magnesium is the treatment of choice should a polymorphic tachycardia develop, and prophylactic infusion in those at risk has been advocated.

Risk Stratification

Children with CHD presenting for NCS are at significantly increased risk of perioperative morbidity and mortality when compared with those without. Recent analysis of the US Pediatric Health Information System database that includes some 500,000 surgical cases per year demonstrated that overall mortality rates in children with CHD were almost nine times greater than in those without (1.06% vs 0.12% in 2019). However, the outcomes for children with CHD appear to be improving; between 2015 and 2019, the mortality rate in children with CHD decreased significantly from 1.16% to 1.06%, whilst the rate in those without CHD remained unchanged.

Risk is not evenly distributed across this population, and a summary of identified risk factors taken from a heterogeneous group of both single centre and large database studies is given in Table 33.4. The American College of Surgeons (ACS) National Surgical Quality Improvement Program (NSQIP) database categorises CHD according to residual cardiac lesion and functional status into minor, major or severe categories (Table 33.5). The data demonstrate that children with minor CHD are at no greater risk than the general population for overall mortality (1.2% vs 1.7%), cardiac arrest or adverse events. This group of children may be safely managed in their local hospital according to usual protocols, without needing transfer to specialist centres solely on the basis of their CHD.

Table 33.4 Factors associated with increased risk of perioperative morbidity

Age: less than 2 years
Higher ASA grade
Multiple comorbidities
Emergency surgery
Complex lesions: - Single ventricle physiology, notably: unrepaired; with BT shunt/prior to Glenn formation; failing Fontan - Cardiomyopathy - Left-ventricular outflow tract obstruction, e.g. aortic stenosis
Presence of long-term complications: - Cyanosis - Cardiac failure - Arrhythmia
Pulmonary hypertension particularly with suprasystemic pressures

Source: ASA: American Society of Anesthesiologists physical status classification.

For children in the major and severe ACS NSQIP categories, the same database has been used to develop the Pediatric Risk Assessment (PRAm) score, designed to prognosticate the perioperative risk of mortality. PRAm identifies eight important factors which are weighted to produce a score of 1–10 (Table 33.6), and validation studies have shown good discrimination for prediction of in-hospital mortality. Scores of ≤ 3 are associated with a low risk of mortality (OR 1.54, 95% CI: 0.78–3.04), scores 4–6 with a medium risk (OR 4.19, 95% CI: 2.56–6.87) and scores ≥ 7 with high risk (OR 22.15, 95% CI: 15.06–32.59). This scoring system emphasises the significance of the functional severity of disease as well as markers of

Table 33.5 ACS NSQIP risk stratification for CHD

Classification	Definition
Minor CHD	Cardiac condition with or without medication and maintenance, e.g. ASD, small to moderate VSD without symptoms Repair of CHD with normal cardiovascular function and no medication
Major CHD	Repair of CHD with residual haemodynamic abnormality with or without medications, e.g. tetralogy of Fallot with free pulmonary regurgitation, HLHS including Stage 1 repair
Severe CHD	Uncorrected cyanotic CHD Patients with documented pulmonary hypertension Patients with ventricular dysfunction requiring medication Listed for heart transplant

Table 33.6 PRAm score: multivariate risk score to predict postoperative mortality

Variable	OR	95% CI	Risk score
Emergency procedure	1.7	1.2–2.3	+1
Severe CHD	1.7	1.2–2.4	+1
Single ventricle physiology	1.8	1.1–3.0	+1
Surgery in preceding 30 days	2.0	1.4–2.9	+1
Inotropic support	2.1	1.4–3.0	+1
Preoperative CPR	2.5	1.3–4.6	+2
Acute or chronic kidney injury	4.4	2.0–9.8	+3
Mechanical ventilation	7.8	5.4–11.2	+4

Notes: OR = odds ratio; CI = confidence interval; CPR = cardiopulmonary resuscitation.

critical illness. It also reinforces SV physiology as a major risk factor. Unfortunately, the ACS NSQIP data provides no information about which professionals performed the surgery (general paediatric or cardiac anaesthetist) or what setting the surgery occurred in (large or small, teaching or peripheral hospital). Additionally, some nuance may be lost by the classification of all children with PHT in the severe category. However, PRAm represents the best tool currently available for objective scoring of risk, which may be helpful in determining allocation of resources.

Anaesthetic Management

Preoperative Assessment

A comprehensive understanding of the anatomy, physiology and risk factors of the underlying CHD lesion is essential before proceeding with anaesthesia. Consideration of the appropriate location for surgery is also imperative. Children with minor CHD appear to be at no greater risk than their peers without CHD, so they may be managed locally as expertise and facilities allow. Transfers in this population are unnecessary and place additional burdens on families and specialist centres. However, it is clear that children in higher-risk categories experience increased perioperative morbidity and mortality. An evidence-based approach supports transfer and management where full specialist facilities are available, including cardiology, cardiac anaesthesia and paediatric intensive care support. Early communication and cooperation between specialist and local services are essential to ensure that transfers for high-risk children who *may* require surgery are facilitated in a timely manner. In children who are functionally limited by their disease, serious consideration should be given to whether surgery is truly essential, whether the child is in an optimal state to proceed and whether deferral until after further cardiac intervention is possible.

Preoperative assessment includes a routine anaesthetic history and examination, determination of long-term complications and identifying those features associated with high-risk categorisation (discussed in the previous section). In addition, specific attention should be paid to the following elements:

Echocardiography

A recent ECHO should be acquired and reviewed preoperatively. It provides invaluable information both in confirming the current structural arrangement and providing an assessment of cardiac function. ECHO reports use standard terminology and usually follow a sequential segmental analysis to provide the following information: atrial position and venous drainage; atrioventricular (AV) and

Table 33.7 Common echocardiography terms

Cardiac position:
Levocardia: normal position cardiac mass with apex to left
Dextrocardia: mirror-image cardiac mass with apex to right and right-ventricle anterior
Dextroversion: cardiac mass rotated with apex to right but left-ventricle anterior
Dextroposition: cardiac mass displaced to right but apex remains to the left

Atrial situs – the situation of the atrial chambers:
Situs solitus: normal atria & abdominal organs
Situs inversus: mirror image atria & abdominal organs
Situs ambiguous: no clear lateralisation
Left-/right-atrial isomerism: two morphological left/right atria

Atrioventricular connections (AVC):
Concordant AVC: usual connection of atria to ventricles
Discordant AVC: left atrium connected to right ventricle, right atrium connected to left ventricle
Double inlet left/right ventricle: two ventricle heart but >50% override of atrial to ventricular connection due to malaligned septa
Absent left/right AVC: mitral or tricuspid atresia

Ventriculo-arterial connections (VAC):
Concordant VAC: usual connection of ventricles to aorta and pulmonary artery
Discordant VAC: left ventricle connected to pulmonary artery and right ventricle to aorta (TGA)
Single-outlet VAC: aortic or pulmonary atresia
Double-outlet right ventricle: two ventricle heart but >50% override of aorta from right ventricle

Valve descriptions and sizes:
The z-score describes the number of standard deviations from the mean of the normal size, e.g. a z score of −3 shows the valve is 3 standard deviations smaller than expected

Doppler flow & pressure gradients:
Velocity of flow between chambers may be used to estimate the pressure gradient using the formula:
Pressure (mmHg) = 4 × velocity (m s^{-1})^2

ventriculo-arterial (VA) connections; abnormal shunts and direction of flow; ventricular dimensions and function; and coronary and arch anatomy. Table 33.7 provides a summary of common terms.

A small amount of tricuspid regurgitation (TR) is considered normal, and the velocity of the regurgitant jet is indicative of the pressure difference between right atrium and ventricle. In PHT, this can give an indication of PAP using the simplified Bernoulli equation $\Delta P = 4V^2$ (P = pressure difference (mmHg), V = peak velocity (ms^{-2})): a TR jet of >4 ms^{-1} indicates a high probability of severe pulmonary hypertension.

Intercurrent Respiratory Tract Infection

Children with increased PBF are more susceptible to respiratory tract infections, and distinguishing infection from cardiac failure can be difficult. In children with PHT or Glenn circulation, infection has a greater than expected effect on PVR. Intercurrent infection should be considered a contraindication to elective surgery, as the perioperative risk of hypoxaemia is increased.

Medication

In general, cardiac medications should be continued as usual. Cardiac medications are not associated with clinically important preoperative electrolyte disturbances in children presenting for cardiac surgery, so routine testing is unnecessary. Some anaesthetists omit angiotensin converting enzyme (ACE) inhibitors on the morning of surgery to avoid hypotension on induction. Data on this phenomenon in adults are conflicting and in children is non-existent. Aspirin is commonly used to prevent shunt thrombosis and should be continued if possible. Children taking warfarin will need admission for conversion to intravenous heparin therapy and appropriate monitoring. In children taking the newer direct inhibitors of activated factor X, there should be liaison with haematology specialists.

Endocarditis Prophylaxis

Some children with CHD are generally at increased risk of developing infective endocarditis (IE). However, the evidence that prophylactic antibiotics are effective at preventing perioperative IE is limited, and current NICE guidelines limit their use significantly (see Table 33.8).

Pacemakers and Defibrillators

The indication for insertion and the features of the device should be ascertained and the current status of the device interrogated by a cardiac physiologist. Remember that in smaller children the box will be implanted in the sub-costal region rather than below the clavicle, and that pacing leads will be epicardial

Table 33.8 Guideline for infective endocarditis prophylaxis

Conditions at risk for infective endocarditis:
Acquired valve disease with stenosis or regurgitation
Previous infective endocarditis
Hypertrophic cardiomyopathy
Valve replacement
Structural congenital heart disease including surgically corrected/palliated lesions but excluding those listed next
Conditions NOT at risk for infective endocarditis:
Isolated atrial septal defect
Fully repaired ventricular septal defect
Fully repaired patent ductus arteriosus
Endothelialised closure devices
Procedures NOT requiring antibiotic prophylaxis even for at risk conditions:
Dental procedures
Upper and lower GI tract procedures
GU tract, including urology, gynaecology and obstetrics
Upper and lower respiratory tract including ENT and bronchoscopy

rather than endovascular. In general, sensing functionality should be switched off for the duration of the procedure to avoid electromagnetic interference. If the underlying rhythm is insufficient to maintain adequate haemodynamics, then temporary programming to an asynchronous, non-sensing mode may be appropriate. Implantable cardiac defibrillators (ICD) should have their shock delivery capacity disabled and external defibrillator pads applied remote from the ICD site. The response of pacemakers to magnets is not predictable without knowledge of the device, so they should be avoided. However, in a true emergency all commonly available ICDs may have their shock function disabled by taping a medical grade magnet to the skin. Some devices will emit a tone whilst so disabled. The device should revert to its usual settings once the magnet is removed, although a formal check should be performed as soon as possible.

Perioperative Care

Many different anaesthetic techniques have been described and utilised successfully in children with CHD undergoing NCS, but no single approach can be recommended for all scenarios. In general, prolonged spontaneous ventilation with high-dose volatile agent is potentially hazardous for many: a balanced technique including controlled ventilation and the use of opioids to reduce the dose of volatile agent is usually preferable. Fluid depletion is tolerated poorly in a number of conditions, notably children with shunts and single-ventricle circulations, so preoperative fasting should be kept to a minimum. If fluid replacement is desired but intravenous access is difficult, then preoperative administration of isotonic fluids via nasogastric tube should be considered.

Premedication

For the majority of children with CHD, the decision to administer premedication may be made according to the anaesthetist's usual assessment of probable compliance. Children with CHD may require multiple procedures under general anaesthesia, so ensuring a calm induction is important from a long-term psychological perspective. Oral midazolam has a long history of successful use, and more recently oral dexmedetomidine has been shown to be equally safe and effective. In some high-risk sub-groups where stress and anxiety can worsen haemodynamics, sedative premedication is strongly recommended. This includes children with dynamic left-ventricular outflow tract obstruction such as hypertrophic cardiomyopathy, where agitation may worsen the degree of obstruction, and those with significant pulmonary hypertension, where agitation may precipitate a pulmonary hypertensive crisis.

Induction and Maintenance

All the volatile agents are associated with dose-dependent reductions in SVR (and probably PVR) and myocardial contractility. There are variable effects on heart rate and rhythm. Gas induction with sevoflurane is widely utilised, but particular care should be taken in children with trisomy 21 (Down syndrome), who appear susceptible to bradyarrhythmias with this technique. Even when gas induction is planned, preoperative application of topical local anaesthetic to appropriate venous access sites can be helpful. It allows intravenous access to be obtained at lighter planes of anaesthesia and can therefore help reduce the length of time the child is exposed to high concentrations of volatile agent. At maintenance concentrations, isoflurane and sevoflurane have minimal effect on contractility or shunt fraction, although animal studies suggest that myocardial depression with both agents

is greater in the presence of a pulmonary artery band. Their use has been widely described in several large case-series of high-risk children for NCS, and the available evidence cannot recommend one agent over the other.

The two most studied intravenous agents for induction in children with CHD are propofol and ketamine. In children undergoing cardiac catheterisation, propofol decreases SVR and mean arterial pressure, whilst PAP and PVR remain unaltered. In the presence of a shunt, propofol leads to a reduction in left-to-right shunt and an increase in right-to-left shunt, thus reducing PBF, which can lead to clinically relevant reductions in oxygen saturations. Judicious use of propofol for induction may be appropriate, but it is best avoided in those with impaired myocardial function and pulmonary hypertension and in patients with right-to-left shunts. Total intravenous anaesthesia (TIVA) with propofol may be considered as an option for maintenance for some surgeries in children with some forms of CHD, but the literature suggests that this approach is not routine. Ketamine has minimal effects on cardiac output, SVR, heart rate and blood pressure. It is a direct myocardial depressant, but haemodynamic stability is maintained by central sympathetic stimulation. It is well tolerated in children with CHD, including those with pulmonary hypertension, and represents the agent of choice in this group and in those where a reduction in SVR is undesirable. However, it should be used with caution in children who are already catecholamine depleted, such as those with severe cardiomyopathy, where the direct myocardial depressant effects may become apparent with significant haemodynamic consequences. Ketamine infusions may be used for maintenance where preserving SVR is important, such as a severe left-ventricular outflow tract obstruction, but should be accompanied by additional sedative medication such as benzodiazepines to avoid unwanted neuropsychiatric side effects, particularly in older children.

Analgesia

Opioids, including morphine, fentanyl and remifentanil, have minimal direct effects on either myocardial contractility or systemic and pulmonary vascular resistance. Intraoperatively they are effective at blocking responses to noxious stimuli that may precipitate a pulmonary hypertensive crisis in those at risk. In general, they may be used in a routine manner, including as postoperative infusions as the surgery requires. However, in children with cardiac failure whose cardiac output is dependent on sympathetic stimulation, administration of opioids may obtund the stress response and sympathetic drive and should be titrated with great caution. Regional anaesthetic techniques have been described and used effectively, although reports are rare and include only small numbers. Epidurals with general anaesthesia have been reported in neonates with complex, uncorrected cardiac disease with no complications, and there are case reports of spinal anaesthesia being used alone in infants. Care should be taken in those children taking anticoagulants for their cardiac condition.

Postoperative Monitoring

Identifying the appropriate postoperative location for a child should be part of preoperative planning. The decision will be influenced by many factors, including the nature and severity of the cardiac lesion, the extent of the surgery and the situation of appropriate beds in the operating institution. A cardiology ward in a large teaching hospital with an extensive cardiac surgical practice will be able to support more significant non-cardiac interventions than a general paediatric ward in a district general hospital, where involvement with a high-dependency unit (HDU) or even a paediatric intensive care unit (PICU) may be indicated. Children with some conditions, such as Williams and Brugada syndrome, require prolonged postoperative monitoring even after relatively minor surgery, as the increased risk appears to extend further than usually expected into the postoperative period.

Further Reading

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