Chapter

31 Anaesthesia for Cardiac Surgery in Children

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Introduction

Most children presenting for paediatric cardiac surgery have congenital heart disease (CHD), the incidence of which is around eight per 1,000 live births (see Table 31.1). Without surgical intervention, around 50% of children with CHD would die during childhood, but with improvements in care almost all now survive to adulthood. Acquired diseases are rare but include endocarditis, intracardiac masses and cardiomyopathies. Children with cardiomyopathy present an increasing need for anaesthesia, for instance, for insertion of mechanical support devices, pacing or defibrillator devices or cardiac transplantation.

Approximately 3,600 paediatric cardiac surgical procedures are carried out annually in the United Kingdom, 20% of which are undertaken in neonates. Most are performed via sternotomy using cardiopulmonary bypass (CPB). Technological advances and miniaturisation of bypass circuits mean it is now feasible to undertake complex cardiac repairs in infants under 2 kg. A smaller proportion of procedures are carried out without bypass via a thoracotomy approach, such as creation of systemic to pulmonary artery (PA) shunts, ligation of patent ductus arteriosus (PDA) or simple coarctation repair.

Paediatric cardiac surgery may aim to be *corrective*, such as ventricular septal defect (VSD) repair, or *palliative*, such as establishment of a stable single-ventricle circulation or valvotomy for valvular stenosis. Staged surgery at different ages is necessary for complex palliative procedures. CPB is also used for non-cardiac surgery, such as complex tracheal repairs or to facilitate removal of tumours adjacent to the heart or major vessels. Owing to greater long-term survival in children with cardiac disorders, there is an increasing need for expertise in the field of adult congenital heart disease (ACHD). This chapter will cover the principles of anaesthesia for cardiac surgery in children.

Table 31.1 Congenital cardiac lesions and their frequency

Lesion	Frequency (%)
Ventricular septal defect	17
Tetralogy of Fallot	12
Transposition of the great arteries	11
Coarctation of the aorta	11
Atrial septal defect	10
Hypoplastic left-heart syndrome	5
Atrioventricular septal defect	3
Pulmonary atresia	1

Overview of Surgery for Congenital Heart Defects

It is essential that the anaesthetist has a clear understanding of the anatomy and haemodynamic consequences of any cardiac lesion. A useful classification of CHD is shown in Table 31.2. Each condition may vary widely in severity, with substantial differences in the physiological and haemodynamic consequences.

Children with inadequate pulmonary blood flow ('right-to-left' shunting) are cyanosed at birth. Tetralogy of Fallot (ToF) is the most common example and may vary widely in severity. Occasionally infants with ToF require a systemic to PA shunt (modified Blalock–Taussig shunt, or mBTS; see Figure 31.1), but more often early complete surgical repair is possible. Conditions such as pulmonary stenosis may be amenable to valvuloplasty by cardiac catheterisation or surgical intervention. In some cases, obstruction to pulmonary flow is so severe that pulmonary blood flow is maintained from the aorta via the arterial duct ('duct-dependent pulmonary circulation'). An infusion of

prostaglandin will be required to keep the duct open prior to surgery or catheter intervention.

Children with increased pulmonary blood flow ('left-to-right' shunting) suffer from cardiac failure. Cardiac failure is managed with diuretics until the child has grown enough to allow full surgical correction. Left untreated, excessive pulmonary flow will lead to irreversible pulmonary hypertension. If cardiac failure cannot be controlled medically, or early surgical repair is not feasible, PA banding may occasionally be required to reduce pulmonary blood flow. PA banding is usually undertaken via sternotomy, without CPB, and involves placing a band around the main PA to restrict pulmonary flow. The band is adjusted to reduce PA pressure distal to the band to about half systemic pressure. Oxygen saturation will generally fall to around 85%. The PA band is removed when definitive surgery is undertaken.

Table 31.2 Classification of cardiac surgical disorders

Category	Specific condition
Inadequate pulmonary blood flow (right-to-left shunting)	Pulmonary atresia Pulmonary stenosis Tetralogy of Fallot
Excessive pulmonary blood flow (left-to-right shunting)	Patent ductus arteriosus Atrial septal defect Ventricular septal defect Atrioventricular septal defect Aortopulmonary window
Abnormal connections (complex shunting)	Tricuspid atresia Transposition of the great arteries Hypoplastic left-heart syndrome Truncus arteriosus Total anomalous pulmonary venous connection
Left-ventricular outflow tract obstruction	Aortic coarctation Hypertrophic obstructive cardiomyopathy Aortic stenosis Interrupted aortic arch

Children with abnormal connections and complex shunting may have a variable combination of cyanosis and heart failure and usually require major surgery involving CPB in the neonatal period. Infants with hypoplastic left-heart syndrome (HLHS) have a single effective ventricle and 'duct-dependent systemic circulation', with blood flow to the distal aorta from the PA via the arterial duct. In transposition of the great arteries (TGA), adequate mixing between systemic and pulmonary circulations may require the duct to be patent. Prostaglandin infusion will be required in both cases prior to surgery or catheter intervention. Infants with total anomalous pulmonary venous drainage (TAPVD) have pulmonary venous return to the right atrium. In 'supracardiac TAPVD', this may be via an abnormal bridging vein in the thorax, and if there is no obstruction to venous return, it may not be

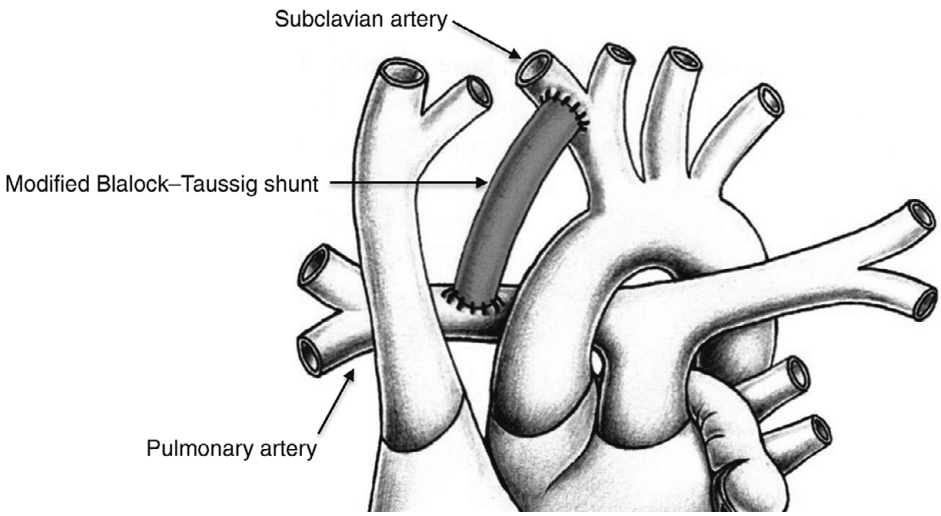

Figure 31.1 Modified Blalock–Tausig shunt.
Source: The PedHeart Resource (www. HeartPassport.com). Courtesy of Scientific Software Solutions, Inc.

detected immediately. In 'infracardiac TAPVD', the pulmonary veins drain into the inferior vena cava (IVC), usually via the hepatic veins. This invariably causes significant pulmonary venous obstruction and requires urgent surgery immediately after birth to maintain oxygenation and cardiac output. The presence of engorged pulmonary veins and profound hypoxaemia may occasionally lead to a misdiagnosis of meconium aspiration syndrome or persistent pulmonary hypertension of the newborn. Obstructed TAPVD may be associated with postoperative pulmonary hypertension.

Obstructive lesions of the left ventricle frequently result in left-ventricular hypertrophy and increase in myocardial oxygen demand. Patients are at risk of acute myocardial ischaemia if oxygen demand is further increased by tachycardia or if coronary perfusion pressure falls during anaesthesia. Patients with Williams syndrome and supra-aortic stenosis are particularly challenging and have a high risk of perioperative sudden death. Babies with lesions such as critical aortic stenosis, interrupted aortic arch or severe coarctation may have duct-dependent systemic perfusion and require prostaglandin prior to surgery in the neonatal period.

Preoperative Assessment and Preparation for Anaesthesia

Successful anaesthesia for CHD surgery requires a meticulous, anticipative approach that starts in the preoperative period. Key features of preoperative assessment include:

- Understanding the anatomy, physiology and haemodynamics of the presenting disorder
- Understanding the proposed surgical procedure, and the consequences of any previous and future surgery
- Determining the clinical condition of the child, particularly cardiac function
- Anticipating the effect of anaesthesia on the circulation

The following considerations are of particular importance:

- *Is the circulation 'duct dependent' or shunt dependent?* For infants with duct-dependent systemic or pulmonary circulation or an mBTs, it is important that the shunt remains patent and that flow through the shunt is 'balanced', meaning that systemic and pulmonary artery blood flow are equal. This balance is determined by systemic vascular resistance (SVR) in relation to pulmonary vascular resistance (PVR). It is important to minimise changes to this balance, including during transport. Oxygen is a potent pulmonary vasodilator, and FiO_2 may need to be restricted to room air in some cases.
- *What is the degree of cyanosis?* Children with cyanotic CHD presenting for surgery may be profoundly desaturated, with polycythaemia and coagulopathy. Identification of current anticoagulation therapy, need for perioperative transfusion and expectation of pre- and postoperative haematocrit requirements should be considered.
- *Is myocardial function impaired?* In some lesions, such as cardiomyopathy, there can be severely impaired left-ventricular function. Some patients may already be on inotropic support. The degree of dysfunction is inferred from the ejection fraction (EF) assessed by echocardiography. EF of less than 30% is indicative of poor systolic function. Obstructive lesions may lead to diastolic dysfunction as well as increased metabolic demand.
- *Is there significant pulmonary hypertension?* If so, any hypoxaemia or hypercapnia during induction and intubation will be poorly tolerated. If the right ventricle is significantly hypertrophied or dilated and distorts the left ventricle, systemic hypotension may compromise coronary artery perfusion, rapidly leading to loss of cardiac output. Pulmonary hypertension is a significant cause of anaesthesia-related mortality in children.
- *Is there a septal defect?* If present, it is important to ensure no air bubbles enter the venous system as this can lead to cerebral air emboli, particularly with right-to-left shunting. All infusion lines must be checked meticulously.
- *What is the patient's surgical history?* If there has been a previous sternotomy, there may be fibrous adhesion of the heart to the back of the sternum with the risk of inadvertent injury and significant bleeding during re-sternotomy. This can be a particular problem if there is an

artificial conduit sitting on the front of the heart. During briefing for repeat sternotomy, the anticipated site of emergency cannulation for femoral or jugular bypass should be discussed with the operating surgeon. This has implications for placement of anaesthetic monitoring lines.

- *Is there an existing shunt?* Arterial pressure monitoring can be inaccurate if placed in the same arm as an mBTS.
- *Is there a pacemaker or implantable cardioverter defibrillator (ICD) in place?* It is important to understand the underlying rhythm disturbance. It may be necessary to reprogram a pacemaker to fixed pacing mode or disable the ICD before cautery can be used safely.
- *Is intercurrent illness present?* CHD with high pulmonary blood flow can lead to increased susceptibility to chest infections. Undergoing CPB with an acute respiratory tract infection may result in significant postoperative morbidity and increased ICU stay. It can be difficult to differentiate between acute upper-respiratory tract infection and heart failure in some symptomatic children. The decision to proceed with surgery needs to be considered on an individual basis; delay of elective surgery is usually indicated in the presence of fever or an elevated white cell count, but a mild cough or runny nose is generally not a reason to cancel surgery.
- *Medications.* Some children will be dependent on prostaglandin infusion or inotropic support, and it is important these are not interrupted. Others may be on anti-failure medication such as diuretics or captopril. Some children with ToF who suffer from severe hypercyanotic 'tet' spells may be receiving propranolol. A few may be receiving some form of anticoagulant therapy such as warfarin or anti-platelet drugs. It is advisable to continue most medications as close to the time of anaesthesia as possible, particularly anticoagulation in infants who are shunt dependent, or any therapies for pulmonary hypertension.
- *Laboratory investigations.* Chronic cyanosis can result in polycythaemia and coagulopathy. Thrombocytopenia and clotting factor deficiency may be present. Children taking diuretics or in severe cardiac failure may have metabolic derangements such as hypokalaemia.
- *Fasting time* is of particular importance in infants dependent on a small shunt, particularly if polycythaemic. Prolonged starvation leads to dehydration, hyperviscosity and potential shunt thrombosis. Clear fluids should be given until one hour before surgery, ideally continuously via a nasogastric tube, and IV fluids should be started if there is uncertainty about theatre time.

Preparation should begin well in advance of surgery to allow time for understanding of the condition and to answer questions from the family. Discussion should include details of anaesthesia procedures, including induction technique; invasive monitoring (including potential complications); postoperative analgesia; and intensive care pathways. The last should cover duration of postoperative ICU stay and potential cardiorespiratory support, including mechanical circulatory support where relevant. Sedative premedication and topical analgesia should be prescribed as necessary.

Induction of Anaesthesia

Haemodynamic stability is the goal in the anaesthetic room. Essential monitoring (ECG, audible pulse oximetry and blood pressure) should be started before induction of anaesthesia. In some children, this may be poorly accepted but should be applied at the earliest opportunity. Ideally intravenous access should be obtained before induction; however, this is not always achievable, in which case it should be established as soon as is feasible after loss of consciousness.

Sevoflurane is widely used for inhalational induction and is generally very safe. Uptake of anaesthetic agents from the lungs is slowed in the presence of a large right-to-left shunt, although this is rarely of practical significance. Importantly, any relative overdose of agent will be equally slow to reverse.

For most patients, there is no definite advantage of one intravenous induction agent over another. All anaesthetic agents have haemodynamic effects and should be used with caution. Propofol is suitable if ventricular function is good but should be avoided where a fall in SVR or diastolic pressure must be avoided.

When ventricular function is poor, intravenous induction can be very challenging:

- *Ketamine* is a direct myocardial depressant, but it usually maintains or increases blood pressure, heart rate and cardiac output via sympathomimetic stimulation. It is a commonly used agent particularly when it is important to avoid a fall in SVR. However, in patients who may be surviving on maximal sympathomimetic drive, such as those with significant impairment of myocardial function, ketamine's unopposed depressant action may lead to a profound fall in cardiac output.
- *Fentanyl* is perceived to be a cardiostable agent but may have detrimental cardiac effects in patients whose cardiac output is maintained by sympathomimetic drive as it suppresses the stress response effectively, which can lead to a fall in blood pressure. Fentanyl also reduces heart rate, an important component in maintaining cardiac output in infants, but may be beneficial where tachycardia is detrimental.
- *Etomidate* has negligible effect on myocardial contractility or blood pressure and is a good agent for patients with very limited myocardial reserve, noting concerns about short-term adrenal suppression and pain on injection.

For patients with very poor myocardial function and low cardiac output, such as those presenting for heart transplantation, it is important to remember:

- Circulation times are slowed, so drug effect may be delayed.
- There is preferential blood supply to key organs such as the brain so smaller induction doses are needed.

The following is the authors' preferred means of induction in high-risk patients: a small dose of fentanyl (0.5–1.0 mcg kg^{-1} IV) is followed by slow increments of midazolam 50 mcg kg^{-1} IV over 10–15 minutes until the patient is having difficulty keeping their eyes open. At this point, ketamine 0.25–0.5 mg kg^{-1} is administered in boluses followed by the muscle relaxant. Blood pressure should be measured at short intervals throughout induction with close attention to any ECG changes.

Pancuronium is commonly used in cardiac anaesthesia because of its long duration of action. Its modest increase in heart rate is beneficial in opposing the reduced heart rate commonly seen with fentanyl and sevoflurane induction in infants, but it should be avoided where a tachycardia may be detrimental, such as infants with poor cardiac function. Vecuronium and rocuronium are good alternatives, with no impact on heart rate.

When inducing anaesthesia in a child with a shunt or duct-dependent circulation, 'balance' of both pulmonary and systemic circulations must be maintained. In patients with pulmonary-to-systemic shunting, injudicious use of anaesthetic agents may drop SVR, favouring systemic perfusion at the expense of pulmonary blood flow, which may precipitate profound hypoxaemia, bradycardia and cardiac arrest. Conversely, administration of high inspired oxygen concentration or over-ventilation must be avoided in patients with duct- or shunt-dependent systemic perfusion. A fall in PVR will increase pulmonary blood flow at the expense of systemic blood flow. This may lead to poor coronary and end-organ perfusion and could lead to cardiac arrest. Maintaining a higher PVR with low FiO_2 will provide adequate flow to the systemic circulation. For many of these patients, oxygen saturation around 80% represents an appropriate balance.

Maintenance of Anaesthesia

Anaesthesia for CPB surgery is patient-specific but can be institutionally guided. It is acceptable to use a combination of moderate dose opioid, typically fentanyl 15–30 mcg kg^{-1}, combined with a volatile agent such as sevoflurane in air or an air/oxygen mix. Nitrous oxide may be used during inhalational induction but should be avoided for maintenance of anaesthesia. In neonates and small infants, volatile agents can cause a profound reduction in blood pressure and cardiac output, and it may be preferable to limit their dose and use higher doses of fentanyl instead. In older children, isoflurane and sevoflurane both cause only a modest fall in SVR and blood pressure at 1–1.5 MAC and are widely used; in these children and those with an anticipated 'fast-track' recovery, excess opioid can and should be avoided. Continuous infusions of propofol or dexmedetomidine are increasingly used and are safe unless cardiac function is very poor.

Regional analgesia in the form of ultrasound-guided paravertebral block (as a single shot or catheter infusion) is opioid-sparing and has gained popularity for its potential in enhanced recovery.

Antibiotic prophylaxis is essential; the choice of antibiotics is guided by institutional protocols and should be administered in the 30-minute period before skin incision.

Monitoring

ECG, pulse oximetry, capnography and invasive arterial and central venous pressure monitoring are required for all cardiac cases. End-tidal CO_2 measurement can be inaccurate in patients with cyanotic CHD and can be up to 2 kPa lower than arterial pCO_2 but provides essential confirmation of tracheal tube placement and a useful trend of ventilation. Non-invasive blood pressure monitoring should be used whilst arterial access is being achieved. A urinary catheter should be placed in all patients undergoing CPB.

Invasive arterial and central venous lines must be secure and reliable and should be placed under full aseptic conditions using ultrasound guidance. For CPB cases, it is customary to insert a multilumen central venous line in addition to a peripheral line. In most cases, this will be via the internal jugular or femoral vein. Thrombosis of central veins is an unfortunate complication of central venous cannulation in small infants. In neonates with univentricular physiology where cavopulmonary connections will be necessary later, such as those with HLHS, it may be preferable to avoid the internal jugular veins if possible, as SVC thrombosis will compromise subsequent successful repair. An umbilical catheter may be in place in newborn infants. Femoral and iliac thrombosis is a common finding after central venous catheters have been placed for even short periods, and the anaesthetist should be mindful of future surgery and catheter interventions when deciding line insertion sites.

Where central venous access is technically difficult, the external jugular vein can be used for pressure monitoring and to provide access for administration of drugs. Rarely, it may be necessary to start surgery with peripheral access only; a direct transthoracic right-atrial line can be placed by the surgeon once the chest is open, which can be used for drug infusions whilst the atrium is closed.

Arterial access is usually via the radial, femoral or occasionally the axillary artery. The smaller peripheral arteries are sometimes used but may become 'shutdown' and display an artificially low pressure as the patient is cooled. It is best to avoid the ipsilateral upper limb if an mBTS is present or planned, as it is likely to under-read the true pressure. It is important to place the arterial line in the right arm during non-CPB coarctation repair, as the surgeon will clamp the aortic arch proximal to the origin of the left-subclavian artery during repair. Clinicians and families should be aware of the risks of limb ischaemia, and limbs should carefully monitored after a vessel has been cannulated, especially in neonates.

Core (usually nasopharyngeal) and peripheral temperature monitors should be placed in all cases. Core temperature is essential for the conduct of CPB and is also important to ensure mild hypothermia prior to clamping the aorta during coarctation repair.

Transoesophageal echocardiography (TOE) offers great advantages perioperatively, allowing confirmation of the diagnosis, detection of a small patent foramen ovale (PFO) not seen on transthoracic echo, detection of air within the heart after cardiotomy and assessment of surgical repair and ventricular function. There are now probes suitable for neonates (3–4 kg), although care must be taken to avoid cardiorespiratory compromise due to compression by the probe. TOE may be undertaken by a suitably trained cardiologist or anaesthetist. It is important that an 'anaesthetic' TOE operator is not also solely responsible for the anaesthesia. If TOE cannot be used, epicardial echocardiography is often employed whilst the chest remains open in the immediate post-CPB period.

Near-infrared spectroscopy (NIRS) provides a measure of cerebral and somatic oximetry as a function of regional blood flow. Cerebral NIRS can be useful in monitoring changes in cerebral perfusion related to CPB and to malposition of the CPB cannulae. Use of NIRS has become routine in recent years, although long-term outcome data are not yet available.

The Pre-bypass Period

For sternotomy, the patient is placed in the supine position with the arms by the sides and a small roll under the shoulders to bring the sternum forward. All lines, monitors and the catheter should be secured, labelled and accessible. Passive cooling is allowed before bypass.

Sternotomy is very stimulating, and adequate opioid, regional and/or volatile anaesthetic is

required to blunt a hypertensive response. Following sternotomy, anaesthesia requirements are reduced. Lung compliance usually increases following sternotomy, and ventilatory parameters should be monitored. Haemodynamic instability can occur during mediastinal dissection due to manipulation of the great vessels, impaired venous return, arrhythmias, bleeding or labile pulmonary vascular resistance. Particular vigilance is required if there is a shunt or duct-dependent circulation, as distortion may interfere with pulmonary or systemic perfusion, and it may be necessary to ask the surgeon to pause to allow recovery.

Prior to CPB, heparin is administered (400 IU kg^{-1}) to prevent clotting of the extracorporeal circuit, a bolus (dose dependent on circuit size) is also added into the CPB circuit. Heparin should always be administered into a secure line. The adequacy of heparinisation is checked after two to three minutes, usually by the activated clotting time (ACT), which should be above 400 seconds.

Cardiopulmonary Bypass

It is important for anaesthetists to understand the principles of CPB, although in many centres the responsibility for running bypass during surgery falls to the surgeon and the perfusionist. In some centres, the anaesthetist may also be involved in running the bypass. CPB requires drainage of the systemic venous return from the patient and, following oxygenation, return via a cannula placed in the ascending aorta. The aortic cannula is placed first, and its correct placement within the lumen of the aorta is vital; this is checked by reading the arterial pressure from the CPB whilst testing the line. NIRS may demonstrate a fall in cerebral oxygenation on the rare occasion that the cannula tip is misplaced.

Unlike most adult cardiac cases where a single venous cannula is placed in the right atrium, paediatric CPB usually requires separate venous cannulae in the SVC and IVC to allow intracardiac surgery to be conducted via the right atrium. If there is an additional major source of venous drainage to the heart, such as a left-sided SVC, this may also need to be cannulated. Venous return to the bypass circuit is usually passive and requires appropriately sized cannulae to ensure complete emptying of the heart. CVP should fall to near zero. A vacuum can be added to the CPB if required to facilitate venous drainage. An advantage of measuring CVP in the internal jugular vein is that obstruction to cerebral venous return by the SVC cannula will be apparent. NIRS may also detect a fall in cerebral blood flow.

Once the cannulae are secured, the surgeon instructs the perfusionist to start bypass. Often this may start with just one venous cannula in place. Lung ventilation is continued until both cannulae are in place and the patient is at predicted 'full flow'.

The blood concentration of drugs such as fentanyl will fall substantially on initiation of CPB due to haemodilution and adsorption of the drug to the CPB circuit, so it is usual to give a supplementary bolus of fentanyl immediately before CPB. A volatile agent is administered into the CPB sweep gas by the perfusionist to maintain anaesthesia during CPB. Hypothermia reduces the MAC of volatile agents and metabolism of drugs, so these measures are usually sufficient to maintain adequate anaesthesia. In children in whom CPB is conducted at only mild hypothermia, a hypnotic agent such as midazolam or a propofol infusion may be administered to minimise the risk of awareness.

Blood flow on CPB is non-pulsatile, with flow rates generally around 3 l $min^{-1}m^{-2}$. The perfusionist will use blood gas parameters, venous oxygen saturation levels and NIRS to monitor adequacy of perfusion.

Mean blood pressure on CPB is generally maintained at around 35–60 mmHg. The pressure often falls on institution of CPB due to haemodilution and vasodilation, but this is corrected by the perfusionist within a few minutes. If there is a PDA or an mBTS, this will need to be ligated to prevent flow into the lungs. Other sources of arterial runoff and low pressure, such as aortopulmonary collaterals, can be more difficult to manage. If blood pressure remains low, it may be necessary to give a vasoconstrictor such as phenylephrine. NIRS may be useful in monitoring adequacy of cerebral perfusion on CPB. Haematocrit is normally maintained at around 20–25% on CPB; increasing this may also improve blood pressure and cerebral oxygenation.

Many operations are performed with core temperature between 30°C and 34°C to reduce systemic and cerebral metabolic demands, although there is an increasing trend to undertake surgery at near normal temperature. For longer, more complex procedures, the temperature may be reduced

to 25–30°C. Some surgery requires hypothermia to 18°C to allow total cessation of bypass. This is known as deep hypothermic circulatory arrest (DHCA) and is discussed in the next subsection.

For many paediatric cardiac procedures, it is necessary to stop the heart by administering a cardioplegia solution into the aortic root. The aorta is cross-clamped to ensure the potassium-containing solution is forced into the coronary arteries. Traditionally this solution was cooled to around 4°C to assist myocardial preservation, but warm blood cardioplegia solutions are increasingly used. Cardioplegia is important for myocardial preservation and is usually repeated every 15–20 minutes. Occasionally, if the procedure is likely to be very short, the heart is fibrillated to prevent ejection and the risk of air embolism whilst repair is undertaken.

Deep Hypothermic Circulatory Arrest

If surgery is required on the aortic arch or head and neck vessels, and in some other complex repairs, the patient is cooled to 15–18°C and the circulation stopped to allow the aortic cannula to be removed. At least 20 minutes of cooling should occur before the circulation is stopped. Ice packs should be applied to the head to aid brain cooling. It is difficult to quantify the long-term impact of this technique on neurodevelopmental outcome, but DHCA times of greater than 40 minutes are associated with an increased risk of cerebral injury. Because of this risk, some centres use selective cerebral perfusion during DHCA. After the circulation is restarted, an adequate period of re-perfusion and re-warming is required before ending bypass.

Discontinuing Bypass

Once the surgical repair is completed the patient is rewarmed to 36°C in preparation for coming off CPB. Electrolytes, particularly potassium and calcium, are corrected by the perfusionist. Ultrafiltration may be used to raise the haematocrit above 30%.

The trachea should be suctioned before restarting ventilation and adequacy of ventilation confirmed. Unless there is a physiological contraindication, ventilation is initially with high inspired oxygen and low-dose volatile agent. The volatile can then be increased and inspired oxygen reduced depending upon the haemodynamic and ventilatory parameters after successfully discontinuing CPB.

CPB causes a variable degree of myocardial dysfunction, so it is common to start an inotropic infusion before coming off bypass. Choice of inotrope varies according to local practice and preference and may need to be adjusted post-CPB to address any particular haemodynamic issues. Rhythm disturbances should also be addressed, and it may be necessary to pace the heart to ensure an adequate heart rate, preferably with atrioventricular synchrony. Transoesophageal echocardiography is useful to assess ventricular function and identify any air in the left side of the heart.

If cardiac output or blood pressure is unsatisfactory, it may be necessary to go back on CPB to allow the myocardium time to recover, resolve rhythm disturbances, increase inotropic support or review the surgical repair. Acute rhythm disturbance and ventricular dysfunction are occasionally due to air emboli in the coronary arteries. This is associated with acute ECG changes and usually resolves over a short period. Increasing the blood pressure with an inotropic agent to improve coronary perfusion may be beneficial.

Modified Ultrafiltration

After successful separation from CPB, some patients will undergo modified ultrafiltration (MUF). This reduces total body water, raises haematocrit and mean arterial pressure and removes inflammatory cytokines. MUF returns warm, 'haemoconcentrated' oxygenated blood into the right side of the heart and can lower pulmonary vascular resistance and attenuate pulmonary hypertension.

Protamine Administration and Management of Coagulopathy

Protamine is administered to reverse heparinisation once the patient is stable and further bypass runs are unlikely. It is important for the anaesthetist to check that the perfusionist has discontinued the CPB pump suckers before administering protamine, to avoid clotting the circuit. Protamine is infused slowly at a dose of 3–4 mg kg^{-1}. Protamine is given slowly, as it can cause increased PVR seen as a temporary reduction in lung compliance and possible hypoxaemia, followed by hypotension.

This is usually short-lived but may require treatment with adrenaline and increased ventilatory support.

Heparin reversal is assessed by checking that the ACT has returned to baseline. Persistent prolongation of the ACT may be caused by residual heparinisation or coagulopathy from any cause (e.g. platelet dysfunction, low fibrinogen). Infusions of platelets, cryoprecipitate or plasma may be required, particularly in neonates. Thromboelastography may be useful in guiding management. Coagulopathy may persist, particularly in neonates, cyanotic patients or those receiving anticoagulant medication.

In patients in whom bleeding is anticipated (complex patients, neonates, patients undergoing repeat sternotomy), antifibrinolytic therapy can be used from the start of the case to reduce bleeding. Tranexamic acid is considered safe and effective in neonates and children, although the optimal dosing regimen remains unclear. The use of aprotinin is controversial given an absence of evidence for superiority over tranexamic acid and following a suggestion of increased complication rates in adults; however, it is still used with ongoing drug surveillance in children, particularly for neonates undergoing complex repairs.

For most patients, we would aim to ensure that haemoglobin is above 100 gl^{-1} in the postoperative period. Haemoglobin should be maintained above 120 g l^{-1} in patients with palliated repairs who continue to have oxygen saturations in the 80s.

Further Management Following CPB

Following satisfactory haemostasis, epicardial pacemaker leads are attached, chest drains are inserted and the chest is closed. Most patients are managed postoperatively on the intensive care unit. Adequate sedation and analgesia are required prior to termination of the volatile agent at the end surgery. A propofol infusion can be useful for transfer. A clear and comprehensive handover to the intensive care team is essential.

In some infants, particularly neonates who have undergone long and complex surgery, sternal closure leads to tamponade of the heart and impaired function. In these infants, the sternum may be stented open and the wound protected with a silicone membrane. The chest is closed after two to three days when cardiac function has improved.

Pacemakers in Cardiac Surgery

Rhythm disturbances may occur after paediatric cardiac surgery and epicardial pacing wires are routinely placed. Heart block may occur after repairs involving the ventricular septum, and atrial dysrhythmias may occur because of suture lines in the atrium. Patients undergoing tetralogy of Fallot repair are particularly prone to junctional tachycardia. Dysrhythmias are poorly tolerated in lesions such as univentricular repairs and heterotaxy. Some patients will require pacemaker support after bypass.

It is important for the anaesthetist to understand pacemaker codes and which settings to use. The pacemaker code is a series of letters describing the type of pacing. The first letter represents the chamber being paced, the second the chamber being sensed and the third the response to sensing. The chambers are atria (A), ventricle (V) or both/dual (D). Pacing is inhibited (I), triggered (T) or both (D) on sensing; pacing being delivered regardless of sensing is designated by the letter 'O'. Permanent pacemakers are more sophisticated and often have a fourth and fifth letter in the code.

In the presence of AV nodal block, dual chamber pacing is required and DDD mode is selected. The atria are paced using AAI or AOO for bradycardia requiring treatment in the absence of AV nodal block.

Fast-track Surgery

Increased pressure on health care resources and scrutiny on outcomes has led to renewed interest in enhanced recovery programmes and early extubation, occasionally in theatre, coupled with short-stay intensive care management following certain cardiac procedures. The spectrum of suitable cases is growing beyond atrial septal defect and simple VSD surgeries. All cases should fulfil certain criteria, including adequate haemostasis, normothermia, minimal inotropic support and minimal ventilatory requirement.

For patients in whom early extubation is planned, multimodal analgesia should be started in the operating theatre. This will include intravenous paracetamol, local analgesic infiltration to the sternal wound or regional analgesia and an intravenous morphine infusion, including nurse-controlled (NCA) or patient-controlled (PCA) analgesia. The historical use of high-dose opioid anaesthesia hinders early extubation and

fast-tracking. An anaesthetic technique using moderate doses of fentanyl (10–15 mcg kg^{-1}) with reversal of short-acting muscle relaxants is more appropriate.

Specific Conditions

Blalock–Taussig Shunt

An mBTS is a GoreTex™ tube from the subclavian artery to the pulmonary artery (see Figure 31.1). Surgery may be undertaken via thoracotomy or sternotomy. Where possible, the monitoring arterial line should not be placed in the ipsilateral arm. Heparin 100 IU kg^{-1} IV is given once the subclavian end of the shunt has been completed. On completion of surgery, it is important to maintain an adequate blood pressure to ensure blood flow through the shunt, and on occasion inotropic support may be necessary. Balancing the circulation is critical and is usually determined by the choice of size of the shunt, occasionally by applying a clip to the shunt if the shunt is too large. Oxygen saturation of 70–80% in air should be expected following completion of the shunt. Excessive shunt flow leads to increased pulmonary flow and reduced systemic flow, high oxygen saturation and signs of low systemic cardiac output. If there is very high runoff from the systemic circulation into the shunt, diastolic pressure can be very low, leading to severe myocardial ischaemia. This can also occur postoperatively as PVR falls during the neonatal period and may require shunt revision.

Tetralogy of Fallot

In this condition, there is right-ventricular outflow tract obstruction (RVOTO) with right-ventricular (RV) hypertrophy, a VSD and an aorta that 'overrides' the VSD (see Figure 31.2). There is a wide spectrum of severity. At one end is the patient with virtually no more than a VSD – the 'pink tetralogy'. At the other end is the very cyanosed patient with total right-ventricular outflow tract obstruction due to pulmonary atresia and right-to-left shunting across the VSD. In some, the main and possibly the branch pulmonary arteries are hypoplastic; it can be very difficult to obtain an acceptable surgical result in these patients. Another variant has an absent pulmonary valve, which results in a hugely dilated pulmonary artery

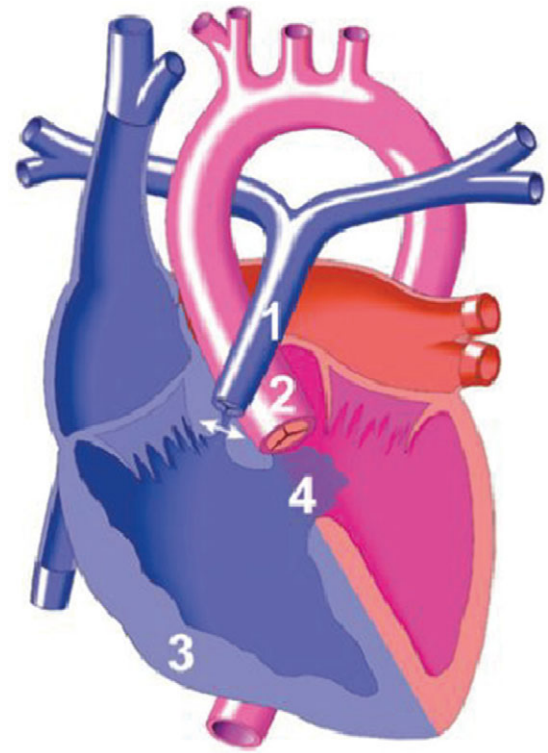

Figure 31.2 Tetralogy of Fallot. Narrowed pulmonary valve and artery (1) with narrowed right ventricular infundibulum (arrows), with aorta (2) over-riding a ventricular septal defect (4) and hypertophied right ventricle (3).
Source: The PedHeart Resource (www.HeartPassport.com). Courtesy of Scientific Software Solutions, Inc.

that can cause significant airway compression, particularly of the left main bronchus.

The classic TOF has a variable degree of RVOTO (and therefore cyanosis) due to a narrowed pulmonary valve annulus, combined with thick infundibular (or sub-valvar) muscle bundles which can spasm to further narrow the outflow. These children have intermittent hypercyanotic spells, known as 'tet' spells, in which spasm of the right-ventricular infundibulum occurs, increasing right-to-left shunting through the VSD. The muscular infundibulum is sensitive to the effects of catecholamines, so spells can be brought on by stress and can be ameliorated using beta blockers. Beta agonists such as adrenaline should be avoided in these patients.

Hypercyanotic spells can be triggered by crying, a fall in SVR at induction of anaesthesia and surgical manipulation. The management of a spell includes administering 100% oxygen, giving a fluid bolus, a dose of opioid such as fentanyl and

physical measures to increase SVR such as pushing the patient's knees up to the chest. If these measures are unsuccessful, a vasoconstrictor such as noradrenaline by infusion or phenylephrine 1–5 mcg kg^{-1} should be administered. Some experts advocate beta blockade with propranolol 50–100 mcg kg^{-1} if the heart rate is high. Esmolol 500 mcg kg^{-1} over one minute followed by an infusion at 50–200 mcg kg^{-1} min^{-1} is an alternative.

Tetralogy of Fallot is repaired using a patch to close the VSD and excising hypertrophied muscle bundles from the right-ventricular outflow tract. It may be necessary to enlarge the outflow tract or the valve annulus with a patch. Widening the pulmonary valve with a transannular patch will produce pulmonary regurgitation. This can make post-bypass management challenging if the hypertrophied right ventricle is poorly compliant. Adequate cardiac output requires a high filling pressure rather than increased inotropic support with catecholamines such as adrenaline. Tachycardia is unhelpful, but maintaining SVR is useful. A phosphodiesterase inhibitor such as milrinone (inodilator/lusitropic effect) combined with noradrenaline is often used. Surgery carries a risk of heart block postoperatively, as the conduction tissue passes close to the borders of the VSD. Occasionally a sinister dysrhythmia known as junctional ectopic tachycardia (JET) occurs which may lead to very poor cardiac output. This is treated with cooling, amiodarone, electrolyte optimisation and minimising inotropic support.

Transposition of the Great Arteries (TGA)

In TGA, the aorta arises from the right ventricle and the PA from the left. A VSD may also be present. The condition is incompatible with life unless there is a source of mixing such as an atrial septal defect (ASD), VSD or PDA, without which oxygenated pulmonary venous blood cannot reach the systemic circulation. Patients are usually managed with a prostaglandin infusion preoperatively, and a balloon atrial septostomy is performed if there is inadequate mixing.

The arterial switch operation (ASO) involves disconnecting the great arteries and attaching them to the correct ventricle. The coronary arteries will need to be transposed. Unsatisfactory coronary artery re-anastomosis leading to myocardial ischaemia and poor cardiac output is a major cause of postoperative problems. Following bypass, the left ventricle can be non-compliant, and fluid boluses should be administered cautiously to avoid left-atrial enlargement and coronary distortion. Milrinone and adrenaline are useful in combination following ASO. Measurement of left-atrial pressure (with a line directly placed by the surgeon) can allow early detection of left-ventricular dysfunction and guide fluid and inotrope therapy.

The origin of the coronary arteries lies very close to the PA following repair. Elevation in PA pressure can occlude the coronaries, compromising left-ventricular function, so measures to minimise pulmonary vascular resistance are important.

Single-Ventricle Circulation

Historically, children with defects where the right ventricle was underdeveloped, for example tricuspid atresia, would be managed with a systemic to pulmonary artery shunt but would die in late childhood or early adulthood due to complications of cyanosis. In the 1960s, Fontan described a palliative operation where pulmonary blood flow was achieved by directly connecting the right atrium to the pulmonary arteries. Although this 'single-ventricle' solution was a major advance, complications such as atrial dysrhythmias were significant, and long-term survival remained poor. The Fontan procedure has undergone several modifications and now involves the staged connection of the SVC and the IVC directly to the pulmonary artery, with improved good-quality long-term survival. The total cavopulmonary connection (TCPC) can be used whichever ventricle is underdeveloped, with the developed ventricle becoming the 'systemic' ventricle.

Creating a single-ventricle circulation usually involves three stages. During the neonatal period, PVR is high, and a systemic shunt (mBTS) is performed. The second stage, a Glenn (superior cavopulmonary) shunt involving the anastomosis of the SVC to the right PA is created at around three to five months of age when the PVR has fallen, and sufficient flow at venous pressure can be achieved through the pulmonary arteries. The mBTS is taken down at this time (see Figure 31.3). The operation creates an end-to-side anastomosis from SVC to the right PA so blood perfuses both lungs; this is the bidirectional Glenn (BDG). Oxygen saturation will usually be in the 80s after

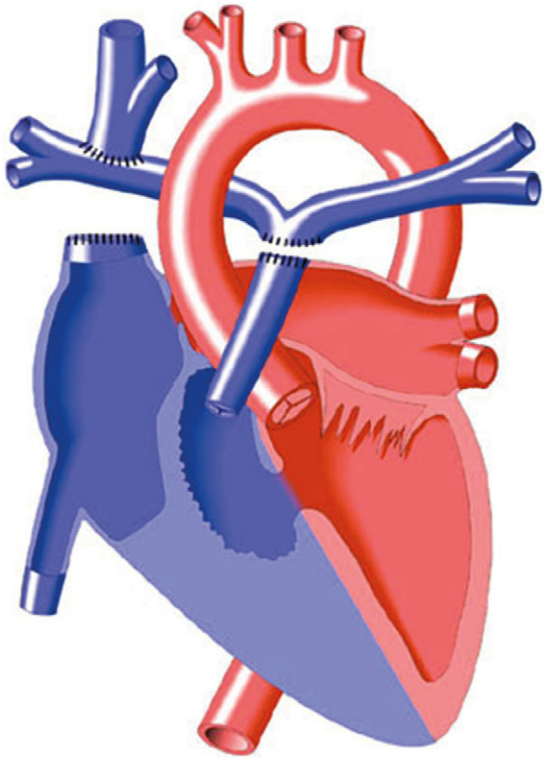

Figure 31.3 Bidirectional Glenn shunt (BDG) in tricuspid atresia, showing anastomosis of the superior vena cava to the right pulmonary artery.
Source: The PedHeart Resource (www.HeartPassport.com). Courtesy of Scientific Software Solutions, Inc.

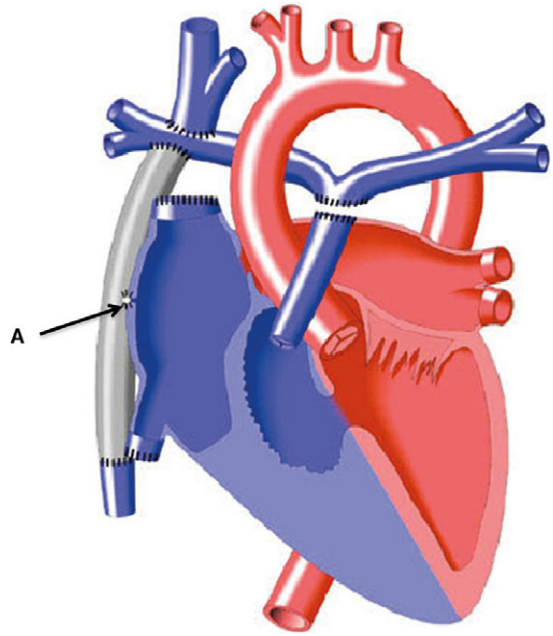

Figure 31.4 TCPC in tricuspid atresia. Both the superior and inferior vena cavae are connected to the right pulmonary artery. There is a fenestration (A) between the IVC to PA conduit and the atrium.
Source: The PedHeart Resource (www.HeartPassport.com). Courtesy of Scientific Software Solutions, Inc.

a BDG. It is important to recognise that pressure measured in the internal jugular vein reflects pulmonary perfusion pressure. This 'venous pressure' must be maintained higher than normal, often 12–15 mmHg, to ensure an adequate transpulmonary pressure gradient. Patients benefit from a low intrathoracic pressure and being in a slightly head-up position after surgery.

The third stage, TCPC completion, in which the IVC is anastomosed to the PA, is usually undertaken between the age of one and five years. This will only be successful if PVR is low. Ideally the transpulmonary gradient, the difference between systemic venous pressure and pressure in the common atrium, should be less than 10 mmHg after the TCPC.

If PVR is high, there will need to be high systemic venous pressure to drive blood across the lungs, and there may be poor filling of the systemic atrium and low cardiac output. Transpulmonary gradient will be high, and there may be poor oxygenation, pleural effusions, ascites and renal dysfunction. In this situation, the surgeon may 'fenestrate' the TCPC by creating a small hole between the GoreTex™ tube and the systemic atrium, allowing systemic venous blood to shunt through this fenestration to the systemic atrium at times of increased pressure (see Figure 31.4). This can improve preload and cardiac output but will be at the expense of lower oxygen saturation. Intrathoracic pressure should be kept low, and these patients benefit from early transition to spontaneous ventilation and extubation. Dysrhythmias are poorly tolerated, and pacing may be necessary.

Hypoplastic Left-Heart Syndrome

In HLHS, the left ventricle is severely underdeveloped and aortic arch atretic; distal aortic blood flow is initially dependent upon ductal flow from the pulmonary artery. The Norwood operation for HLHS makes use of the right ventricle as the systemic ventricle and creates a neo-aorta using the main pulmonary artery. The branch PAs are disconnected from the main PA and are supplied by either an mBTS or, in the Sano modification, an RV-to-PA conduit.

Table 31.3 Manipulating PVR and SVR

Direction of change of vascular	Intervention resistance
PVR ↑	Hypercapnia Hypoxia Acidosis PEEP High airway pressure Atelectasis
PVR ↓	Hypocapnia Alkalosis High FiO_2 Low haematocrit Nitric oxide Prostacyclin (epoprostenol) Milrinone Low airway pressure
SVR ↑	Vasoconstrictors Hypothermia Raising haematocrit
SVR ↓	Vasodilators Anaesthetic agents Low haematocrit Warming

As both the pulmonary and systemic circulations are supplied by the same ventricle, it is important to balance SVR and PVR to maintain adequate pulmonary and systemic perfusion (see Table 31.3). This usually necessitates low inspired oxygen and a moderately raised $PaCO_2$ to maintain PVR. When appropriately balanced, arterial oxygen saturation will be around 75–80%. These patients can be very challenging perioperatively as they can rapidly develop very poor systemic output if PVR is allowed to fall. Limiting inspired oxygen concentration to FiO_2 0.21 is crucial in this situation. Progression via a Glenn to a TCPC is as described in the previous section.

Coarctation Repair

Coarctation may present in the neonatal period or later in childhood. Neonatal coarctation often presents with severe shock when the PDA closes at around four to seven days of age and can be mistaken for sepsis. Immediate management is to attempt re-opening of the duct with prostaglandin infusion to allow perfusion of the distal aorta from the pulmonary artery. There may be severe left-ventricular dysfunction, and ventilatory and inotropic support may be required. Surgery to repair the coarctation is undertaken when cardiovascular and metabolic stability has been restored. Volatile agents are poorly tolerated in these neonatal patients, and a balanced technique with high-dose fentanyl is routinely used.

Surgery is usually via a left thoracotomy, although occasionally in neonates it can be easier to undertake via sternotomy. During the repair, the surgeon will clamp the aorta, usually for a period of around 15–20 minutes, which places spinal cord perfusion at risk. Moderate central cooling should be achieved before the clamp is applied. In older patients, blood pressure (measured in the right arm) may rise significantly during this period, and it may be necessary to increase the volatile agent or occasionally start a vasodilator. It is important to stop these a few minutes before the aortic clamp is removed to avoid a catastrophic fall in blood pressure and to have volume replacement ready. Dialogue with the surgeon is essential.

Anaesthesia for Ventricular Assist Device Implantation and Cardiac Transplantation

Children requiring a ventricular assist device (VAD) present with poor cardiac function and reserve. Meticulous pre-anaesthetic planning and induction is key. Intravenous, fully monitored induction is preferred, with the theatre team on standby for immediate CPB support. Children presenting for VAD support as a bridge to transplant most commonly have dilated cardiomyopathy but could have restrictive disease or congenital heart disease. They are likely to be supported on inotrope infusions and with poor end-organ perfusion. The important aspects of anaesthesia are as previously described and include a balanced technique with appreciation for slow circulation times and a reduced requirement for most medications. The child may be supported with a left-sided device only or with biventricular support. Once supported by a VAD, the child is anticoagulated to prevent thromboembolic complications. Routine monitoring may be challenging if a non-pulsatile VAD is used, since pulse oximetry and non-invasive BP measurement require pulsatile flow to function. However, once safely supported by a VAD, the child should be more haemodynamically stable. It is important to act on

monitoring, filling pressures and the alarm functions of the support system to maintain cardiac output.

Cardiac transplantation includes a multitude of clinical considerations for the anaesthetist and close collaboration with the transplant team. The presence of mechanical support, an implantable defibrillator or pacemaker, anticoagulation management, pulmonary hypertension, previous sternotomies and vascular access should be assessed. Other issues include management of patient and parental anxiety and support for the family after being called in during inconvenient hours or after a long time on a waiting list. Anaesthesia should be administered with care as described previously, in liaison with the transplant coordination team concerning the timing of organ arrival. The perfusionists and surgeons should be on standby in case of instability on induction of anaesthesia. The anaesthetist is required to start the immunosuppression regimen during surgery and to provide meticulous attention to coagulation and infection control. Providing optimal conditions for surgery and ongoing care can make this a challenging yet rewarding anaesthetic to give. It is not uncommon for patients to require a short period of extracorporeal membrane oxygenation (ECMO) post-transplant, usually for a period of 48–72 hours.

Key Points

- Successful anaesthesia for paediatric cardiac surgery requires a meticulous, anticipative approach that starts in the preoperative period.
- Many infants with congenital heart disease require a 'balanced' circulation in which changes in pulmonary or systemic resistance with anaesthesia can lead to severe adverse haemodynamic disturbances.
- Patients with unrepaired tetralogy of Fallot with severe right-ventricular outflow tract obstruction can develop life-threatening profound hypoxia during induction if systemic resistance falls and may need vasoconstrictor support.
- Children who are very cyanosed and polycythaemic will often have abnormal coagulation.

Further Reading

Andropoulos DB, Stayer SA, Russell IA, Mossad EB, eds. *Anesthesia for Congenital Heart Disease*, 2nd ed. Wiley-Blackwell. 2010.

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