Anaesthesia for correction of congenital heart disease (for the specialist or senior trainee)

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Key points
Preoperative assessment includes: assessment of recent infection and need for i.v. hydration (cyanotic children) and antifibrinolytics (neonates and cyanotic children are at increased risk of bleeding).

Before initiation of cardiopulmonary bypass (CPB), heparin 3 mg kg⁻¹ is administered to ensure an activated clotting time of at least 400 s or three times the baseline.

Before separation from CPB, ventilation, heart rate and rhythm, temperature, acid–base, and electrolytes must be satisfactory and inotropes and blood products are nearly always needed in infants.

Haemodynamic instability and haemodilution (anaemia, thrombocytopenia, and reduction in clotting factors) are complications of CPB in children.

Arrhythmias, bleeding, low cardiac output syndrome, pulmonary hypertension, and systemic inflammatory response syndrome are the main immediate postoperative complications.

Congenital heart disease is common occurring in ~1% of live births.¹ The aim of surgery is to achieve a physiologically correct, biventricular repair. However, this is not always possible, and instead, a palliative approach may be required resulting in a single-ventricle circulation. Anaesthesia for correction of congenital heart disease requires an understanding of (i) the principles of neonatal and paediatric anaesthesia, (ii) the anatomy and physiology of congenital heart disease, (iii) the principles of cardiopulmonary bypass (CPB), and (iv) the expected postoperative complications of paediatric cardiac surgery.

Success requires a multidisciplinary team approach. In addition to the anaesthetist, other key members are the paediatric cardiac surgeon, paediatric cardiologist, perfusionist, intensive care staff, and cardiac liaison nurses.

Congenital heart disease can be classified in a number of ways. A simple physiological classification (based on shunt type and outflow tract obstruction) is helpful for anaesthesia as shown in Table 1.

Aims and limitations

This review aims to discuss the principles of preoperative preparation, the management of anaesthesia and CPB, and the common postoperative complications of children undergoing cardiac surgery. Owing to the complexity of congenital heart disease and the variety of surgical procedures, a detailed discussion is beyond the scope of this article. Furthermore, since anaesthesia for paediatric cardiac surgery is regarded as a ‘super-specialist’ consultant post, a detailed knowledge is not required as part of the curriculum for a Certificate of Completion of Training in Anaesthetics.² Therefore, this review is limited to a brief overview of the subject and the discussion of key concepts.

Preoperative considerations

Many children attend a preoperative assessment clinic for both psychological and medical preparation before surgery. Psychological preparation for parents and children is vital. This includes realistic expectations of the day of surgery and duration of procedure, postoperative intensive care, and subsequent rehabilitation. Cardiac liaison nurses play a key role and help provide continuity in this area.

Routine medical investigations include baseline blood tests (full blood count, coagulation screen, and renal and liver function), methicillin-resistant Staphylococcus aureus screening, ECG, chest X-ray, and a recent echocardiogram. Investigations usually occur 1–2 weeks before surgery to allow time for reporting and collating of results.

During the anaesthetic preoperative visit, all investigations including the recent echocardiogram and any angiography must be reviewed to ensure a thorough understanding of the child’s anatomy, physiology, planned procedure, and potential complications. This usually involves discussion with the cardiac surgeon and cardiologist.

Routine history and examination should pay particular attention to the following areas:

(i) Any history of recent upper respiratory tract infection, fever, nocturnal cough, moist cough, or chest wheeze as these predispose to adverse respiratory events.³ Also, ‘acute green runny nose’ or a raised C-reactive protein, liver transaminases, and/or white cell count may be indicative of the presence of infection which could be poorly tolerated after the immunomodulation associated with CPB.

(ii) Evidence of cardiac failure suggested by poor feeding, failure to gain weight, sweating, tachypnoea, tachycardia, and
Anaesthesia for correction of congenital heart disease

Table 1 A physiological classification of congenital heart disease

1. ‘Simple’ left-to-right shunt lesions—these cause an increased pulmonary blood flow (PBF)
   a. Atrial septal defect
   b. Ventricular septal defect (VSD)
   c. Atroventricular septal defect (AVSD)
   d. Patent ductus arteriosus
   e. Aortopulmonary window

2. ‘Simple’ right-to-left shunt lesions—these cause a reduction in PBF with cyanosis
   a. Pulmonary atresia
   b. Pulmonary stenosis
c. Tricuspid atresia
d. Ebstein’s anomaly. Consists of downward displacement of an abnormal tricuspid valve into the RV cavity, part of the RV is thus incorporated into the right atrium (atrialized RV), and the remaining RV cavity is malformed

3. Complex shunts—these cause mixing of PBF and SBF. Cyanosis occurs as a result of complex interactions between systemic SVR and PVR
   a. Transposition of the great arteries (TGA)
   b. Truncus arteriosus
c. Total anomalous pulmonary venous drainage (TAPVD)
d. Double outlet RV
   e. Hypoplastic left heart syndrome
   Most of these lesions (except TAPVD) are examples of a parallel circulation

4. Obstructive lesions
   a. Coarctation of the aorta
   b. Interrupted aortic arch
c. Aortic stenosis
d. Pulmonary stenosis

- hepatomegaly. These children have poor cardiac reserve and may be very sensitive to the vasodilatory effects of induction agents. Many will have large left-to-right shunts and so high concentrations of oxygen will increase shunt flow at the expense of systemic perfusion.

(iii) Pulmonary hypertension (PHT) which is a risk in children with increased pulmonary blood flow (left-to-right shunt), obstructed pulmonary venous drainage, or increased left atrial pressure (LAP). PHT develops earlier in some lesions (e.g. atroventricular septal defects, tricuspid atresia) and in certain patient groups (e.g. Trisomy 21). Separation from CPB will require attention to minimizing pulmonary vascular resistance (PVR) and nitric oxide may be required.

(iv) Cyanosis which increases the risk of hyperviscosity before operation and increased bleeding after operation. Hyperviscosity can cause cerebral vein and sinus thrombosis. Risk factors include: age <5 yr, dehydration, fever, and iron deficiency anaemia. Preoperative i.v. fluid therapy may be used to minimize the risk especially in children with a haemoglobin concentration of 18 g dl$^{-1}$ or greater. Abnormal laboratory tests of haemostasis are documented in 20% of children with cyanosis, but all children with cyanosis are at increased risk of postoperative bleeding. Therefore, consideration should be given to the use of antifibrinolytics such as tranexamic acid, and the need for blood products such as cryoprecipitate, platelets, and/or fresh frozen plasma should be anticipated.

(v) Potential sites for peripheral and central venous access. Previous surgery or recent cardiac catheterization may make this difficult.

(vi) Building a rapport with the child and family, and a frank discussion of perioperative risk.

(vii) Presence of congenital syndromes associated with heart defects which have implications for perioperative care. The range of syndromes is large but common and important ones include: Di George syndrome which requires irradiated blood products, CHARGE syndrome where choanal atresia may preclude nasal intubation and midface hypoplasia and micrognathia can make airway management increasingly difficult with age.

Premedication is often unnecessary in infants under a few months of age but is not contraindicated. For other children, sedative premedication is commonly used to avoid distress, minimize oxygen consumption, and may reduce the amount of induction agent so minimizing reductions in systemic vascular resistance (SVR). Benzodiazepines are commonly used, but the use of other agents such as triclofos or clonidine is also reported.

Management of anaesthesia

The usual anaesthetic technique involves a gaseous or i.v. induction, muscle relaxation, opioid analgesia, and maintenance with a volatile agent, although other methods are described.

In small infants, venous access may be difficult so a cautious gas induction with sevoflurane is frequently used. Ketamine has no effect on SVR, increases mean arterial pressure (MAP), and is well tolerated in children with PHT, making it the i.v. agent of choice. Etomidate also provides haemodynamic stability but may be associated with adrenal suppression. Propofol profoundly decreases SVR and MAP, which alters shunt dynamics. In children with a right-to-left shunt, propofol worsens cyanosis by increasing shunt flow. Therefore, propofol is unsuitable for many children with heart disease. Induction times are prolonged in children with cardiac failure so patience is required to prevent excessive drug administration.

Any long- or medium-acting neuromuscular blocking agent may be used. Pancuronium is commonly used in neonates because it produces a tachycardia. This is helpful because the cardiac output in neonates is rate-dependent, and it also offsets the bradycardic effect of large doses of opioid. Pancuronium in combination with fentanyl produces very stable cardiovascular conditions. High-dose opioid techniques (fentanyl 25–50 μg kg$^{-1}$) and spinal anaesthesia have been shown to reduce the stress response in infants undergoing cardiac surgery. However, recent years have seen a reduction in the amount of opioid used, in order to facilitate early extubation either immediately in theatre or within a few hours of admission to the intensive care unit (ICU). Spinal anaesthesia is rarely used in UK practice. Either sevoflurane or isoflurane
can be used for maintenance and both have been shown to have no effect on shunt fraction in children undergoing cardiac catheterization. A nasal rather than oral tracheal tube is common practice in infants and small children. Nasal tracheal tubes make ICU nursing care easier and may be more stable. Arterial and central venous access is secured and a urinary catheter inserted. Cerebral monitoring, using near-infrared spectroscopy to give a cerebral tissue oxygenation index, is now considered mandatory by many experts. The same technology can also be used to monitor splanchic/renal and skin perfusion.

Before skin incision, surgical prophylactic antibiotics are administered according to local guidelines. Antifibrinolytic therapy (e.g., tranexamic acid) should be considered in patients at high risk of bleeding (neonates, cyanotic patients, and those undergoing redo or complex surgery requiring prolonged CPB). Steroids (dexamethasone or methylprednisolone) may also be used to reduce the inflammatory response to CPB.

Management of CPB

Good communication between the surgeon, anaesthetist, and perfusionist is required during CPB. In infants, the priming volume of the CPB pump can be more than twice the child’s total blood volume. This causes significant haemodilution with anaemia, thrombocytopenia, and a reduction in clotting factors, thereby contributing to the coagulopathy associated with CPB in children. In infants, blood is usually added to the prime aiming for a haematocrit of 21–24%.

Before the institution of CPB, baseline arterial blood gases and activated clotting time (ACT) should be checked. Vigilance is required during surgical dissection and aortic and venous cannulation because there may be significant haemodynamic instability due to anatomical distortion or arrhythmias. At the request of the surgeon, heparin 3 mg kg\(^{-1}\) is administered and the ACT measured. An ACT of 400 s (or at least three times the baseline) is required before institution of CPB and is monitored throughout the duration of CPB.

During CPB, ventilation is stopped but anaesthesia, analgesia, and muscle relaxation must be maintained. Isoflurane (or sevoflurane) is added to the CPB circuit in a concentration of 0.5–1.0% or a propofol infusion administered. Additional opioid and muscle relaxation is administered either to the child before the commencement of CPB or added to the CPB prime. Further doses of benzodiazepines may also be used. Depending on the type of surgery, surgical technique, and/or surgical preference, the perfusionist will allow the child’s core temperature drift to 34–35°C (so-called ‘warm bypass’) or the child is actively cooled to 32°C or lower (minimum of 15°C). For certain types of surgery involving the ascending aorta and aortic arch, it can be impossible to perfuse the body via the aortic cannula; therefore, the child is cooled to 15–17°C and ice packs applied to the head. At this temperature, the circulation can then be either completely arrested (known as deep hypothermic circulatory arrest) or selective brain perfusion maintained by a carotid artery cannula while the perfusion to the rest of the body is arrested (known as regional low flow perfusion). Perfusion pressure, venous saturations, cerebral oxygenation, haematocrit, and electrolyte and acid–base status are continually monitored. Target perfusion pressure varies with age and ranges from an MAP of 30 to 50 mm Hg (neonates–young adults). At low temperatures, acid–base management becomes more complex and either an α- or pH-stat strategy may be used. Both produce similar results at 37°C but differ significantly below 27–30°C. During hypothermic CPB in children, pH-stat management reduces seizures, ICU length of stay, and mortality. As temperature decreases, H\(^+\) and OH\(^-\) dissociation constants increase so the H\(^+\) concentration decreases and pH rises. Therefore, since electrochemical neutrality must be maintained, at 37°C, the cellular pH is 7.4, whereas at 20°C, the cellular pH is 7.8. As temperature decreases, cellular pH is mediated by carbon dioxide (CO\(_2\)). Solubility of CO\(_2\) increases as temperature decreases, causing a decrease in partial pressure (at 37°C: P\(_{CO_2}\) = 40 mm Hg, at 20°C: P\(_{CO_2}\) = 16 mm Hg). pH-stat management involves addition of CO\(_2\) to the CPB circuit so that the total content of blood CO\(_2\) is increased. Thus, at 20°C, the P\(_{CO_2}\) is maintained at 40 mm Hg rather than being allowed to decrease to expected values of 16 mm Hg (i.e. if the total CO\(_2\) content was kept the same) and the pH is maintained at 7.4 instead of rising to 7.8. Therefore, if blood gases are temperature-corrected, the pH would be more acidic and the CO\(_2\) higher than expected at normothermia.

Once surgery is completed, the child can be separated from CPB. However, before this can happen, several criteria must be met. These include:

- Ventilation re-established and both lungs seen to fully inflate.
- Heart rate and rhythm acceptable (if not will need pacing).
- Vasoactive infusions commenced (most infants require inotropic support such as dopamine or low-dose epinephrine after CPB and a lusitrope such as milrinone is also frequently used especially for complex surgery).
- Core temperature > 36°C, and peripheral temperature at least 34°C.
- Normal acid–base and electrolyte status.
- Blood available and blood products if required.

Management after separation from CPB

After separation from CPB, transoesophageal or epicardial echocardiography is performed to evaluate the adequacy of surgical repair. Modified ultrafiltration (MUF) can then begin if required. MUF removes excess body water thereby increasing haematocrit and also removes some inflammatory mediators. MUF has been shown to improve cardiac output and decrease PVR. When MUF
is complete, protamine 3 mg kg\(^{-1}\) is given to reverse the heparinization and the ACT checked to ensure that it has returned to baseline. Blood and clotting factors are administered as required and may be guided by thromboelastography. Once haemostasis is achieved and the surgery has finished, the child is transferred to the ICU. During transfer, full invasive monitoring is continued, and emergency drugs and fluid should be available. A comprehensive handover including the surgical details and echocardiography findings must be given to ICU medical and nursing staff.

**Surgery without CPB**

Some types of congenital heart surgery are performed without the use of CPB, for example, correction of coarctation of the aorta, pulmonary artery (PA) banding, and shunt procedures (modified Blalock–Taussig shunt or central shunt). In general, PA banding is done to limit excess pulmonary blood flow until the child is able to have a definitive procedure. This strategy ‘protects’ the lungs by reducing the risk of developing PHT caused by excessive pulmonary blood flow under high pressure. Shunt procedures are performed to augment pulmonary blood flow in situations where pulmonary blood flow is otherwise inadequate.

**Postoperative care**

Although most postoperative care is undertaken by a specialist paediatric intensivist, the paediatric cardiac anaesthetist must be familiar with common postoperative complications requiring immediate attention. This allows the anaesthetist to anticipate and treat problems early while still in theatre. It also helps in addressing parental anxiety about what to expect perioperatively.

Maintaining adequate cardiac output is an important aspect of postoperative care. Cardiac output is difficult to measure in children so surrogate markers such as MAP, venous saturations, and serum lactate are often used. Close attention is also paid to the key components of cardiac output, that is, heart rate and stroke volume. Slow heart rates are treated with pacing and tachyarrhythmias managed aggressively often by cooling, amiodarone, and overdrive pacing. Stroke volume consists of preload, afterload, and contractility; preload is maintained by the judicious use of fluids to maintain central venous pressure (CVP) and LAP at predetermined values, milrinone is commonly used to reduce afterload, and dopamine or epinephrine is used to improve contractility. Occasionally, sternal reopening is required to improve a low cardiac output state, or in complex neonatal surgery, a delayed sternal closure technique may be used from the outset.

Complications after cardiac surgery can be divided into five general areas:

1. **Arrhythmias**
2. **Bleeding**
3. **Systemic inflammatory response syndrome (SIRS)**
4. **PHT**
5. **Low cardiac output syndrome (LCOS)**

Arrhythmias may be a second- or third-degree heart block which requires pacing, or a tachyarrhythmia such as supraventricular tachycardia or junctional ectopic tachycardia. Management of tachyarrhythmias involves excluding other causes such as pain and seizure activity, correcting hypoxia, acidosis, and electrolyte abnormalities, ensuring adequate sedation and paralysis, cooling to 35°C, and considering amiodarone and overdrive atrial pacing.

Bleeding after cardiac surgery is common and reduced by a meticulous surgical technique and the use of antifibrinolytics such as tranexamic acid. Blood loss into surgical drains is measured regularly. Losses >5 ml kg\(^{-1}\) in the first 2 h or over 1 ml kg\(^{-1}\) thereafter warrants attention. Treatment involves correcting hypothermia and clotting factor abnormalities (including checking the ACT for adequate heparinization reversal with protamine). Any blood loss >10 ml kg\(^{-1}\) demands immediate surgical review.

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**Table 2 Specific complications associated with specific lesions**

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Complication</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVSD</td>
<td>Atrioventricular valve regurgitation</td>
<td>Minimize overload or stretch on the repaired valve using afterload reduction (milrinone, SNP)</td>
</tr>
<tr>
<td></td>
<td>Arrhythmias</td>
<td>Maintain normothermia, correct electrolyte abnormalities</td>
</tr>
<tr>
<td></td>
<td>Pulmonary hypertension</td>
<td>Avoid hypoxia, hypercapnia, acidosis</td>
</tr>
<tr>
<td>TGA</td>
<td>Coronary ischaemia</td>
<td>Avoid overdistension of heart, use small (5 ml kg(^{-1})) fluid bolus’s</td>
</tr>
<tr>
<td></td>
<td>LV dysfunction</td>
<td>Afterload reduction</td>
</tr>
<tr>
<td></td>
<td>Arrhythmias</td>
<td>Maintain normothermia, correct electrolyte abnormalities</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>RV dysfunction, associated with RV failure and LCOS</td>
<td>RV afterload reduction: maintain high CVP, reduce PVR, reduce LAP (improve contractility). Milrinone reduces PVR and improves diastolic function</td>
</tr>
<tr>
<td></td>
<td>Arhythmias</td>
<td>Maintain normothermia, correct electrolyte abnormalities</td>
</tr>
<tr>
<td>Single-ventricle repair, e.g.</td>
<td>Cardiac output is dependent on PBF, where PBF=((CVP – LAP)/PVR)</td>
<td>Keep CVP high (head-up and elevate legs); LAP low (maximize contractility, maintain sinus rhythm, consider milrinone); PVR low (good oxygenation and analgesia; early spontaneous ventilation; avoidatelectasis)</td>
</tr>
<tr>
<td>Fontan</td>
<td>Pleural effusions and liver dysfunction</td>
<td>Maintain normothermia, correct electrolyte abnormalities</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Monitor and treat accordingly</td>
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</tbody>
</table>
sudden decrease in drain losses may imply impending cardiac tamponade and requires urgent attention.

SIRS is common after cardiac surgery, peaks 8–12 h after operation, and may be reduced by the use of steroids and MUF. The fever component of SIRS predisposes to arrhythmias; capillary leak worsens the tissue and lung oedema contributing to poor gas exchange; and reduced cardiac function worsens the already impaired function caused by ischaemia associated with cross-clamping and administering cardioplegia to the heart on CPB.

PHT causes an increase in right ventricle (RV) afterload, decreases RV output which decreases left ventricle (LV) preload, and decreases LV output causing LCOS. PHT also reduces lung compliance and increases airway resistance, thereby increasing the work of breathing. Cardiac lesions with a high pulmonary blood flow or left-sided obstructive lesions (e.g. interrupted aortic arch, mitral stenosis, total anomalous pulmonary venous drainage) are risk factors for developing PHT. Treatment of PHT involves administration of some or all of the following: high inspired concentrations of oxygen, alkalization (aim for pH > 7.45), nitric oxide, milrinone, sildenafil, prostacyclin, and magnesium.

In addition to the general postoperative complications described above, certain defects are associated with specific postoperative complications. For example, repair of a ventricular septal defect is associated with risk of heart block due to surgical interruption of the Bundle of His. Some specific complications associated with common defects are listed in Table 2.

Conclusions

We have described the basic principles of anaesthesia for correction of congenital heart disease including preoperative preparation, anaesthetic and CPB management, and discussion of the common postoperative complications. Understanding cardiac physiology and anaesthetic pharmacology is essential to guide the management strategy and predict complications. Although anaesthesia for paediatric cardiac surgery is considered a ‘super-specialist’ area, the principles we have described provide a starting point for trainees exposed to this type of anaesthesia in a tertiary centre and also form the foundation for adding a more detailed knowledge and understanding if a senior trainee or consultant wishes to develop this area of their practice.

Declaration of interest

None declared.

References


Please see multiple choice questions 17–20.