

ANAESTHESIA FOR THE CHILD WITH CONGENITAL HEART DISEASE: POINTERS AND PITFALLS

Congenital heart disease (CHD) is the commonest birth defect.

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You are hoping for a quiet night on call in your regional hospital. You've had a busy but uneventful day in theatre anaesthetising patients on the routine operation list. But, then you receive a call from an orthopaedic surgeon: A 5-year-old boy has been involved in an MVA and has femoral and compound tib-fib fractures. He is unstable and needs to get to theatre soon, and his mom says that he has a 'lekhart' and a Glenn shunt was done at Red Cross Hospital a year ago. Will that be OK? Sigh ...

Congenital heart disease (CHD) is the commonest birth defect, with a reported incidence in developed countries of 1:125 live births. Currently, with multidisciplinary advances in technology, 90% of these children reach adulthood. In South Africa these figures are likely to be different, but the same principles apply.

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Although congenital cardiac anomalies have been recognised for centuries, few beneficial treatments were available until the 20th century. Since 1938, when the first patent ductus arteriosus (PDA) was successfully ligated, there have been remarkable developments in anaesthesia, surgery, bypass technology, interventional cardiology and intensive care to enable even very complicated anatomical variations to be palliated or corrected. In particular, cyanotic congenital heart disease (CCHD) has been the focus of much of this research, with both palliative and corrective surgical and interventional advances having an impact on outcomes and quality of life.

Compared with children without CHD presenting for non-cardiac surgical procedures, those with CHD have an increased perioperative morbidity and mortality. Children at highest risk are those with hypoplastic left heart syndrome (HLHS), aortic stenosis (AS), and cardiomyopathy. Added to this is the age of the infant (those under 2 years are at highest risk), as well as those with additional extracardiac anomalies (see article in this journal).

Pathophysiology of CHD

Understanding the pathophysiology of CHD is key to managing the condition. Many such children may have a trivial or self-limiting anatomical abnormality, e.g. a small ventriculo-septal defect (VSD), which may close spontaneously. Regardless of the lesion, anyone who anaesthetises children regularly will, at some time, be called on to anaesthetise a child with CHD. It is therefore imperative that the anatomical variations and pathophysiology of each patient presenting for surgery are understood, and that the impact of the anaesthetic agents on the patient is known. This problem is further complicated by the medications that each child may be taking, as these may interact

with the anaesthetics or, as in the case of anticoagulants, may require optimisation before surgery.

Although traditionally classified into acyanotic and cyanotic heart disease, CHD is functionally categorised according to anatomical variations and the shunts which consequently develop. The anatomical variations may be divided into 3 groups, but with complex CHD there may be more than one lesion:

- Increased pulmonary blood flow (PBF) causes a volume or pressure overload to the pulmonary circulation (e.g. VSD, atrial septal defect (ASD), PDA).
- Decreased PBF results in a relative inability to oxygenate blood.
- Obstruction to flow causes an increased ventricular work load to overcome the obstruction, as well as resulting in a relatively reduced circulation distal to the obstruction.

Four major consequences of CHD

- Cyanosis is a response to chronic hypoxia because of decreased PBF and/or mixing. It starts when the level of de-oxyhaemoglobin exceeds 2.5 g/dl. When the haemoglobin is high, the patient appears increasingly blue, but when he/she is anaemic this is less obvious.
- Congestive heart failure is due to increased PBF resulting from shunt lesions, obstructive lesions and impaired ventricular contractility. Cardiac failure results in decreased systemic perfusion, increased pulmonary congestion, and increased venous congestion. Clinical features include tachycardia, tachypnoea, a gallop, and hepatomegaly.
- Pulmonary hypertension (PHT) occurs in left to right (L to R) shunts with increased PBF. PHT also develops with prolonged pulmonary venous obstruction (e.g. total anomalous pulmonary venous drainage (TAPVD), high left atrial pressure (e.g. hypoplastic left heart syndrome (HLHS)), and in mixing lesions with increased PBF (e.g. truncus arteriosus). PHT is not usually a feature of the R to L shunts, but should always be kept in mind in case of non-cardiac-related causes such as obstructive sleep apnoea. PHT is dynamic, and may be worsened by hypoxia, hypercarbia, acidosis, and hypothermia. This vasoconstriction may become irreversible from any age – from a couple of months to a few years.
- Arrhythmias. These may be part of the presenting pathology but often occur as a result of surgery. Increasingly more children are surviving surgery, but require cardiac rhythm management devices (pacemakers and implantable cardioverter defibrillators), which necessitate knowledge of their function and optimisation before anaesthesia is administered.

Non-cardiac features of CHD

Lungs

- Decreased lung compliance from chronically increased PBF (L to R shunting, pulmonary venous congestion).
- Airway compression by vascular structures.
- Scoliosis, more commonly with cyanotic children, possibly because many of these children have had a thoracotomy and a rib resected. This is not usually severe enough to have an impact on pulmonary function.

- Haemoptysis, phrenic nerve or recurrent laryngeal nerve injury (during previous surgery) may be present. Phrenic nerve damage with diaphragmatic dysfunction will have an impact on respiratory mechanics. Recurrent laryngeal nerve injury may result in chronic aspiration and lung disease.
- CCHD patients have a blunted response to hypoxia, which usually resolves with full correction but, with palliative surgery, may remain life-long.

Children at highest risk are those with hypoplastic left heart syndrome (HLHS), aortic stenosis (AS), and cardiomyopathy.

Haematological

- **Polycythaemia** is a compensatory mechanism to hypoxaemia and cyanosis. Tissue hypoxia results in increased cardiac output and increased oxygen-carrying capacity of the blood by the renal release of erythropoietin, stimulating the bone marrow to increase red cell production and thus increasing blood viscosity. Preoperatively, one may need to consider venesection (when packed cell volume (PCV) is $\geq 60 - 65\%$) to increase stroke volume, improve systemic blood flow, and increase oxygen delivery. These patients may present with embolic cerebral infarction causing neurological symptoms and intracranial abscesses.
- **Hyperviscosity syndrome** results from polycythaemia and is especially problematic with dehydration.
- **Haemostatic abnormalities** correlate with the degree of hypoxaemia and erythrocytosis. Problems include platelet abnormalities of either qualitative or quantitative function, decreased fibrinogen, increased fibrinolysis, and clotting factor deficiencies. These may be problematic before, during, or after surgery.

Neurological

- Due to the hyperviscosity, general features of fatigue, tiredness, muscle weakness, myalgia, and paraesthesia of fingers, toes and lips may occur. Central nervous system (CNS) signs and symptoms include headache, depressed mentation, neurological fallout from transient ischaemic attacks, cerebrovascular accidents with permanent neurological sequelae, dizziness and blurred vision.
- Paradoxical emboli to CNS.
- Brain abscesses.
- Cerebral thrombosis in children with erythrocytosis.
- Nerve compression by vascular structures (recurrent laryngeal nerve).

Clinical presentation

Patients will generally present as pink, blue or grey:

- pink: normal, or L to R shunt (ASD, VSD, PDA)
- blue: R to L shunt or mixing lesions (tetralogy of Fallot (TOF), transposition of the great arteries (TGA))
- grey: haemodynamically unstable, poor cardiac output and poor peripheral perfusion (e.g. co-arctation of aorta, interrupted aortic arch).

Factors associated with the highest peri-operative risk

- age under 2 years
- complex lesions
- major surgery
- emergency surgery
- presence of the long-term consequences of CHD, i.e. arrhythmias, cyanosis, cardiac failure, and pulmonary hypertension
- associated extracardiac anomalies.

Congenital anomalies and clinical syndromes associated with heart lesions include Down's syndrome, Di George's syndrome, mucopolysaccharidoses, Noonan's syndrome, rubella, fetal alcohol syndrome, tracheo-oesophageal fistula, and exomphalos (see article in this journal).

Referral to a specialist centre

In all situations, except life-threatening emergencies, the following children should be referred to a specialist centre or anaesthetised by someone with an in-depth knowledge of anaesthesia for children with cardiac disease:

- cyanotic cardiac disease
- CHD plus a difficult airway
- neonate with CHD
- Eisenmenger syndrome: irreversible PHT with a R to L shunt
- PHT
- AS
- hypoplastic left heart syndrome
- single ventricle pathology (e.g. Fontan circulation)
- cardiomyopathy
- any child requiring intensive or high care where this is not available.

NB: A child who is under 2 years of age with a difficult airway and CHD should only be anaesthetised by an experienced paediatric anaesthetist.

Peri-operative management

Always answer the following questions before committing to the case:

- Do you, as the anaesthetist, understand the pathophysiology of the cardiac lesion(s) and the consequences of palliative or corrective procedures that have been performed on your patient (Table I)?
- What is the severity of the disease: is there cardiac failure?
- Is this an isolated lesion or part of a syndrome?

- Is there a history of previous surgery? If so, can you access the information?

Table I. Important questions to assess risk

- Is there a shunt? A shunt is an abnormal pathway for blood to flow through an intra-cardiac, an extra-cardiac or a combined lesion
- Is there obstruction to or reduction of flow owing to a supra-valvular, sub-valvular or a primary valvular abnormality? Is this fixed (aortic stenosis) or dynamic (tetralogy of Fallot)?
- Is there an increase or decrease in pulmonary or systemic flow?
- Are there any indications of poor ventricular function – clinically or from investigations?
- Is this a single ventricle lesion, or are the ventricles functioning as normal balanced chambers?
- Is there valve regurgitation?
- What will be the impact of drugs and ventilator settings on the lesion (i.e. how will these affect systemic vascular resistance (SVR), pulmonary vascular resistance (PVR) and shunt flow)?
- What is the severity of the lesion?
- Are there non-cardiac features of CHD?
- What medication is prescribed?

If there is any concern about the safety of the patient, do not proceed without discussion with an experienced colleague.

Goals

- Evaluate the airway: need to maintain the airway and optimise oxygenation.
- Improve or optimise oxygenation, but be aware of the unnecessary use of 100%

oxygen with CCHD and the production of unwanted oxygen radicals.

- Preserve myocardial function.
- Optimise balance between systemic and pulmonary vasculature resistances.
- Air bubbles: meticulous attention to remove all air bubbles from intravenous tubing to avoid systemic air emboli (especially via a R to L shunt).
- Plan the anaesthetic, from premedication, preoperative oxygenation, induction, maintenance, monitoring – especially placement of lines, haemodynamic manipulation, and mode of ventilation.
- Be prepared: resuscitation drugs, equipment, analgesia and postoperative care (ensure an ICU bed for ill or unstable patients).

Important preoperative considerations

Anatomical and physiological data review

Review all available data and recent investigations. These may include blood tests, radiographs, echocardiography reports, and cardiac catheterisation findings. A valuable exercise is to draw either a simple diagram or box sketch of the anatomy to clarify blood flow, identify shunts and decide whether or not manipulation of the SVR will benefit the patient.

It is advisable to contact the cardiologist of the tertiary institution to find out as much as possible about the child, the surgery that the child has had, and the medication he/she has been administered. A meticulous, goal-directed anaesthetic and surgical history should be taken, and specific attention paid to the cardiovascular, respiratory and neurological systems. Of particular relevance is a history of cyanotic spells, squatting, failure to thrive, poor feeding and sweating. Measure the blood pressure in all four limbs.

Understanding the pathophysiology is the key to managing CHD.

A drug history is vital, as anti-failure therapy, anticoagulants, anti-hypertensive and anti-arrhythmic agents may all be prescribed, and may affect peri-operative management.

Communication with parents is crucial and should include discussion about associated risks of the procedure and the anaesthetic planned. Good communication between healthcare providers is essential, which should include at the very least a telephonic conversation with the patient's cardiologist, with the planned procedure discussed to evaluate its impact and the anaesthetic on the cardiac pathology, and vice versa. Premedication should be the same as for any child before surgery. Sedative anxiolytic premedication is advised for most children with CCHD (unless there are other reasons

Table II. Good anaesthetic practice points for children with CHD

- Preoperative fasting: minimise starvation times. Give free glucose-containing fluids until 2 hours preoperatively. Alternatively, place an intravenous line for fluid replacement from the time of starvation. CCHD patients have an increased haematocrit and, with their failure to thrive, have limited glycogen reserves; hence this treatment regimen is beneficial from both a physiological and comfort point of view.
- Provide supplemental oxygenation from the time of the administration of premedication sedation.
- Continue cardiac medications until time of surgery. Diuretics may be discontinued on the day of surgery. Depending on the planned surgery, antihypertensive medication may be discontinued on the day of surgery.
- Preoperatively, measure baseline oxygen saturations in the ward with the patient on room air.
- Surgery involving major shifts in fluid compartments, changes in position (prone, Trendelenburg), and alterations in intracavity pressures (laparoscopic surgery) may alter ventilation parameters and haemodynamics and therefore impact on peri-operative risk.
- Endocarditis prophylaxis should be provided in accordance with international guidelines.
- Jugular venous pressure is difficult to use as a reflection of cardiac failure and a sign of venous congestion. Liver size, and a change in the position of the liver palpated in the abdomen, is a much better indicator of central venous pressure. A 1 - 2 cm liver is normal.
- Cardiac failure is diagnosed by an increase in heart and respiratory rates, a gallop rhythm, and hepatomegaly.

for omitting this), and in children with PHT.

Haemodynamic principles of anaesthetic management of shunts

Aim to improve oxygenation and myocardial function by the manipulation of flow through the shunt(s):

Left to right (L→R)

- Avoid increasing SVR: may benefit from afterload reduction.
- Avoid negative inotropes: may need to use inotropes.
- Beware of fluid overload, or worsening congestive cardiac failure (CCF).
- Oxygenate well, ventilate early. Monitor appropriately.

Right to left (R→L)

- Maintain high SVR: adequate fluid administration; pharmacologically – ketamine, phenylephrine; physically – knees to chest, pressure in groins, occlusion of femoral arteries.
- Maintain adequate (or increased) intravascular volume and good blood pressure.
- Avoid increase in PVR.
- Minimise intrathoracic pressure: normal to low ventilatory pressures, low PEEP, complete neuromuscular blockade, especially with suctioning. This should be balanced against the need for improved ventilation with these strategies. If saturations decrease with increased intrathoracic pressures, then decrease the inspiratory pressures and respiratory rate. If the saturations then improve, lower intrathoracic pressures are needed. This presumes an adequate preload.
- Oxygenate well (but avoid administering a high FiO₂ which will not improve the

arterial oxygenation and possibly do harm), monitor appropriately until full recovery.

- Meticulous attention to air bubbles in the intravascular lines is crucial.

Induction

The two induction agents which have been studied most for CHD and non-cardiac surgery are propofol and ketamine. Propofol decreases SVR and mean arterial pressure (MAP). Heart rate, PVR, and pulmonary arterial pressure (PAP) remain unchanged. Shunts: L to R flow decreases, R to L flow increases (therefore reducing PBF and oxygenation). Propofol should not be used in children with AVSDs.

Ketamine increases MAP, but has no effect on SVR, PVR or PAP. It is well tolerated in children with PHT. Etomidate has minimal haemodynamic effects in children with shunt lesions for cardiac catheterisation. Adrenal suppression is well documented in adults and in patients with sepsis.

Sevoflurane, isoflurane, fentanyl and midazolam have all been used, with some reports of reduced cardiac index and contractility in the fentanyl/midazolam group.

Each case requires individualised evaluation and the anaesthetic techniques should be planned accordingly. The choice of an intravenous or inhalational technique will depend on the anaesthetist, and may be determined by the ease or otherwise of vascular access.

The effects of CHD on anaesthetic uptake are determined by intracardiac shunts, cardiac output, cerebral blood flow, functional residual capacity, minute ventilation and anaesthetic blood gas solubility. In theory, R to L shunts have de-oxygenated blood bypassing the lungs, thus diluting the inhalational agent that has been taken up by the PBF, resulting in slow induction with inhalational agents. If a surgical systemic to pulmonary shunt has been placed, the PBF increases and the speed of induction is increased. Theoretically, intravenous induction is more rapid because the induction agent would reach the brain quicker after bypassing the lungs.

In cases of CCHD where referral is not immediately possible, SVR needs to be maintained; hence high-dose inhalational techniques are not advised. However, low-dose volatile (0.5% halothane or 2-3% sevoflurane) may be beneficial for TOF to reduce the RV outflow tract obstruction.

Emergency drugs that should be available are the following: fluids, phenylephrine, adrenaline, sodium bicarbonate. The advantages of ketamine are that it maintains SVR and therefore improves PBF, and also provides analgesia. The theoretical concern of catecholamine effects on the RV outflow tract infundibulum does not pose a clinical problem, but if this is perceived to complicate induction the use of esmolol, at a dose of 0.5 mg/kg by slow intravenous injection over a minute, is beneficial.

Maintenance of anaesthesia and analgesia

The choices remain those of the anaesthetist. Options will depend on the lesion, the ability to alter flow (when a shunt is present),

and the surgery planned. Techniques include total intravenous anaesthesia using ketamine, with or without midazolam, high-dose opioids (when postoperative ventilation is planned), inhalational agents (sevoflurane shows suitable cardiovascular stability) and combinations of the above. Muscle relaxation may be required and antibiotic prophylaxis should be provided according to international recommendations. Nitric oxide should be avoided in patients when there is heart failure (HF) and when PHT is a concern. Desflurane and propofol infusions are not routinely used.

Regional anaesthetic techniques have been used but reports are scarce. The risk:benefit ratio must be considered, as many of these children have coagulation abnormalities, either before and/or after surgery.

Capnography: In the presence of a R to L shunt, the end-expiratory CO₂ consistently underestimates the true arterial CO₂ level.

Postoperative care

Except for children with fully repaired ASDs, PDA and VSDs with no residual PHT, most children with cardiac lesions should be cared for in a high care or intensive care facility. Good pain control and maintenance of normothermia are essential for a stable recovery. Patients who have a Glenn or Fontan circulation (i.e. with a low pressure shunt) benefit from spontaneous ventilation and specific positioning (head up and feet up) and a good preload to facilitate pulmonary perfusion and good oxygenation. Hypovolaemia, especially with CCHD, should be corrected immediately.

Conclusion

There is a definite group of patients who should not be anaesthetised at secondary level hospitals and who, but for exceptional circumstances, should be referred to a specialist centre or to a facility where an anaesthetist with these skills is available. Professional responsibility dictates that any health professional anaesthetising children with CHD must understand the cardiac anatomy and physiology of that particular patient. Children with heart disease are at greater risk of peri-operative critical adverse events than those with normal hearts. The anatomical variations, with the potential consequent development of shunts, require meticulous preoperative evaluation, examination of all available investigations, discussion with the cardiologist and surgeon involved, and goal-directed planning of the anaesthetic technique.

Recommended reading available at www.cmej.org.za

IN A NUTSHELL

- Understanding the pathophysiology, and communication with colleagues, is essential to good outcome.
- Congenital heart disease (CHD) is the commonest birth defect.
- Highest peri-operative risk: aortic stenosis, hypoplastic left heart syndrome, cardiomyopathy, associated extracardiac anomalies.
- Four major consequences of CHD: cyanosis, congestive heart failure, pulmonary hypertension, and arrhythmias.
- Prolonged starvation times are detrimental.
- Know your limitations.