Southern African Journal of Anaesthesia and Analgesia 2019; 25(5):S27-30

South Afr J Anaesth Analg
ISSN 2220-1181 EISSN 2220-1173

PACSA SUPPLEMENT

"*There's a child with a heart problem on my orthopaedic list*": An approach to anaesthesia for children with congenital heart disease presenting for non-cardiac surgery

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Congenital heart disease is the most commonly occurring congenital anomaly. 95% of patients with mild to moderate lesions will now survive to adulthood, as will 69% of patients with critical disease. These patients are likely to present for non-cardiac surgery. Assessing risk of anaesthesia in these patients is important as they are at increased risk for perioperative morbidity and mortality. New physiological classifications of congenital heart disease are discussed, as are factors associated with increased risk in these patients. Patients with high and moderate risk should ideally be referred to a unit familiar with congenital heart disease.

Keywords: congenital heart disease, children, non-cardiac surgery, anaesthesia, risk assessment

Introduction

Children with congenital heart lesions are no longer uncommon in clinical anaesthetic practice.

Congenital heart disease (CHD) is the most commonly occurring congenital anomaly, occurring in about eight per 1 000 live births. It is associated with a high incidence (15–30%) of extra-cardiac defects and recognisable genetic syndromes (particularly Trisomy 21).[1] There is a suggestion that the global incidence of CHD is rising; whether this reflects a true increase in the condition or improvement in diagnostic modalities, is uncertain. The reported incidence of CHD in Africa of 2.3 per 1 000 live births is almost certainly a significant under-representation of the situation.[2]

Survival rates for patients with CHD have improved markedly. Where paediatric cardiology and surgery facilities are available, 97% of babies with non-critical CHD will survive to one year, and 95% will survive to adulthood. 69% of babies with critical CHD will also survive to adulthood.[3] For probably the first time in human history, there are now more adults with CHD alive than there are children with CHD.

When presenting for non-cardiac surgery, it is well recognised that these patients have increased morbidity and mortality perioperatively, although the actual incidence is difficult to define accurately. The evidence for risk assessment and the best perioperative management is likewise limited and likely to remain so in the near future because of the enormous variation in pathology seen and the rapidly changing cardiac treatment modalities.[3]

Physiological classifications of congenital heart disease

Anaesthetists often use a physiologically-based classification, as opposed to the anatomically-based classifications used by cardiologists and surgeons, because they provide some guidelines into the perioperative management of these patients. The original physiological classification divided CHD patients into those who were cyanotic and those who were not. This was adequate when very few patients with complex lesions survived but given the variety and complexity of lesions and treatments currently seen, newer classifications have been proposed.

1. Circulation-based physiological classification of CHD

(i) Those patients with normal or 'series' type circulation

In a normal or series type circulation, there is separation of the pulmonary and systemic circulations, which work together in series. Most patients with repaired CHD who have two ventricles will be in this category, as are some types of unrepaired CHD, such as coarctation of the aorta and valvular stenoses or regurgitations.

One sub-group of these patients are those with smaller, unrepaired atrial and ventricular septal defects and patent ductus arteriosus (ASD, VSD, PDA). The blood flow, or shunt, across such a defect depends upon the pressure gradient across the two circulations.

When the pulmonary vascular resistance is lower than the systemic vascular resistance, the shunt is from the systemic (left) to the pulmonary (right) circulation – the 'left-to-right shunt' of acyanotic CHD.

Another subset of these patients have lesions that result from obstruction to the pulmonary circulation, (Tetralogy of Fallot, isolated pulmonary stenosis with a VSD). The resulting shunt is usually from the right side of the heart to the systemic circulation – the 'right-to-left shunt' of cyanotic congenital heart disease.

(ii) Patients with parallel or 'balanced' circulation

In a parallel or balanced circulation, the systemic and pulmonary circulations are in full communication with each other and function in parallel. The blood flow to each circulation depends upon the balance of the systemic vascular resistance to the pulmonary vascular resistance, with the shunt from left-to-right or from right-to-left, depending on alterations of the resistances.

An example of a balanced circulation would be a child with an atrio-ventricular septal defect (AVSD). Patients with artificial defects such as a modified Blalock-Taussig (BT) shunt or an atrial septostomy in uncorrected transposition of the great vessels (TGV) can have balanced circulations.

(iii) Single ventricle circulation

In patients with only one functioning ventricle, palliation by way of conversion to a Fontan circulation usually requires a three stage procedure. Patients may present for non-cardiac surgery after any one of these stages.

The first procedure, usually performed as a neonate or infant, is to provide a reliable, low pressure circulation to the lungs by way of a modified BT (aorto-pulmonary) shunt; a Sano (ventriculo-pulmonary) shunt or by banding of the pulmonary artery.

These patients are then palliated to a bi-directional Glenn shunt. This produces a cavo-pulmonary connection supplying the left and right pulmonary artery with blood directly from the superior vena cava (SVC). The BT shunt or other shunts from the heart to the lungs are taken down and the lungs are supplied passively by the SVC.

Full conversion to a Fontan circulation is best done before the age of five years to preserve ventricular function. The inferior vena cava is anastomosed to the pulmonary circulation, resulting in a passive pulmonary circulation with the ventricle providing the systemic cardiac output.

2. Classification of CHD as severe, moderate and mild[4]

Severe CHD presents in the neonatal period or during infancy and usually requires early cardiac surgery. Examples of this type of disease include Tetralogy of Fallot, other cyanotic heart disease, transposition of the great vessels, hypoplasia of the right or left ventricle, critical pulmonary or aortic stenosis, symptomatic coarctation of the aorta, large VSDs, PDAs and AVSDs.

Moderate CHD is amenable to medical treatment until the child grows to a larger size, when corrective cardiac surgery or catheter laboratory corrections may be indicated. Lesions include moderate aortic or pulmonary stenosis or regurgitation, other septal defects and non-critical aortic coarctation.

Patients with *mild CHD* constitute the largest group of patients and often present with minimal symptoms or a cardiac murmur. They may or may not require correction.

Perioperative risk classification of CHD[5,6,7,8]

The following classification of risk is a suggested guideline for assessing children with CHD presenting for non-cardiac surgery.

High risk	Intermediate risk	Low risk
Severity of lesion		
Complex lesions – Tetralogy of Fallot with pulmonary regurgitation, large septal defects, valvular lesions, single ventricles, balanced circulations, cardiomyopathy	Simple lesions – Restrictive ventricular septal defects, small atrial septal defects, repaired defects with normal series circulation	Simple lesions
Physiological compensation		
Poor physiological compensation – cardiac failure, pulmonary hypertension, arrythmias, cyanosis	Good compensation – normal physiology and cardiac function or well-compensated parameters of cardiac functioning – good effort tolerance, satisfactory physical and cognitive development	Good physiological compensation, normal physiology
Type of surgery		
Major surgery – intra-peritoneal, intra-thoracic, major orthopaedic surgery, procedures with significant blood loss likely to require transfusion	Major surgery including procedures that will require blood transfusion	Minor (surface type) surgery
Emergency surgery	Emergency surgery	Elective surgery
Length of hospital stay prior to surgery > 10 days	Length of hospital stay prior to surgery > 10 days	Length of hospital stay prior to surgery < 10 days
Age < 2 years	Age < 2 years	Age > 2 years

Ideally, children with CHD who have a high or moderate risk of complications during anaesthesia should be referred to a unit familiar with CHD.

Physiological compensation of CHD[9]

Physiological compensation in this context refers primarily to good cardio-respiratory function. Correction of CHD does not always result in good cardio-respiratory function, conversely, patients with uncorrected cardiac lesions can be well compensated. An example of a poorly compensated corrected patient could be a child after correction of AVSD with residual pulmonary hypertension and atrio-ventricular valve regurgitation. An example of a well-compensated non-corrected defect would be a child with a restrictive ventricular septal defect and normal pulmonary artery pressures.

Cardiac failure in CHD may be due to volume overload (for example, in defects causing left-to-right shunts); pressure overload (for example, in pulmonary or aortic stenosis) or cardiomyopathy (genetic; due to a single ventricle or acquired,

or after a Jatene switch with poor left ventricular function). Cardiac failure is associated with a high perioperative morbidity and mortality – in one study, there was a 10% risk of cardiac arrest and 96% of the patients required perioperative inotropic support.

Cyanosis is a common feature of patients with right-sided obstructive lesions, such as Tetralogy of Fallot and in those patients who have been palliated with a modified BT or bi-directional Glenn shunt. These patients often have an arterial saturation of between 75 and 85%.

Cyanotic patients are at high risk of complications, because of their cardiac pathology, including 'tet spells' and because of the associated polycythaemia. A low platelet count, the physiological response to polycythaemia, can predispose to coagulation disorders. Hyperviscosity syndromes can be a problem in children under the age of five years, with the risk of cerebral vein and cerebral sinus thrombosis, particularly in the presence of dehydration, fever and starvation.

All children with CHD presenting for surgery should have an electrocardiogram (ECG) as the incidence of arrythmias is high and increases with increasing age. Right-bundle branch block is common but unlikely to lead to heart block. Ventricular ectopic beats in children are more sinister – these patients are at a high risk of sudden death.

Pulmonary arterial hypertension is another clear predictor of perioperative morbidity, with an 8% increase in the risk of perioperative complications.

Management of children with CHD

There are no clinically validated regimens for perioperative management of anaesthesia for non-cardiac surgery for patients with CHD. Given the variety of CHD seen and the individual physiological responses to the conditions and to treatment, these will be difficult to produce.[7,10]

Case study

A three-year-old child presents for correction of a club foot.

A history of an uncorrected VSD, diagnosed at the age of nine months after a lower respiratory tract infection, is noted. The child is taking furosemide and digoxin. He has no current symptoms related to his chest or heart. He weighs fifteen kilograms, is active and developing normally. Clinical examination reveals a healthy child with no cyanosis. A loud systolic murmur is heard over the heart. Mild cardiomegaly is present on the chest x-ray. The patient's ECG is normal.

Assessment

This patient has one of the commonest types of CHD (32% of all CHD). He is classified as having a normal, series circulation with a left-to-right shunt. The lesion is in the moderate category, not having required early correction.

There are no signs of cardiac failure (failure to thrive, tachycardia, tachypnoea, repeated lower respiratory tract infections) or pulmonary hypertension, so the child is physiologically well compensated.

There is a possibility of pulmonary hypertension associated with this type of lesion and therefore an echocardiogram would be useful prior to surgery. In this case, the echo reveals a restrictive ventricular septal defect with a left-to-right shunt, mild right ventricular hypertrophy and dilation, and normal pulmonary arterial pressures.

The patient is older than two years of age. The surgery is elective and superficial.

The risk assessment therefore suggests that this patient is at low risk for complications perioperatively.

Suggested management of a patient with well-compensated moderate or mild CHD and a left-to-right shunt

Preoperative considerations

1. Avoid prolonged starvation in children with cardiomegaly who may require higher filling volumes to maintain cardiac output.
2. Sympathetic stimulation caused by crying can increase pulmonary arterial pressure, as can hypoxia or hypercarbia caused by excessive sedation. Judicious use of premedication under close supervision may be warranted.

Intraoperative considerations

1. Standard 'ASA' monitoring is sufficient for minor procedures.
2. There is no evidence to suggest that intravenous induction of anaesthesia is superior or inferior to gas induction. It is acceptable to gain venous access after gas induction in these patients. Avoid using prolonged, high doses of sevoflurane which are vasodilatory and negatively inotropic. Propofol reduces systemic vascular resistance but has little effect on pulmonary vascular resistance. Ketamine is widely used and considered safe.
3. There is a risk of paradoxical air embolism from the right ventricle to the systemic circulation so that meticulous attention must be paid to de-airing intravenous lines and when injecting drugs.
4. Good analgesia is required to avoid sympathetic stimulation. Regional techniques work well.
5. Antibiotic prophylaxis for bacterial endocarditis is not required in this case.

   Antibiotic prophylaxis is required for patients with the following[7]:

   - Gums, teeth, oral mucosal surgery.
   - Patients who have prosthetic material inserted as part of the repair (patches for VSD repairs, BT shunts) within six months of their placement.

- Unrepaired congenital cyanotic conditions and repairs with residual defects.
- Prosthetic heart valves.
- Previous episodes of infective endocarditis.

6. Haemodynamic goals of anaesthesia management:
   - Maintain sinus rhythm.
   - For patients with left-to-right shunts, maintain the ratio of pulmonary to systemic blood flow.
   - Increasing pulmonary blood flow by reducing pulmonary vascular resistance, (high inspired oxygen concentrations, hyperventilation) will result in increased shunt, decreased lung compliance, increased work of breathing and systemic hypotension.
   - Increasing pulmonary vascular resistance (hypoxia, hypercarbia, acidosis, hyperthermia) can result in sudden increases in pulmonary artery pressure and acute right ventricular failure – a pulmonary hypertensive crisis.
   - Vaso-active drugs tend to act on both the systemic and pulmonary circulation.
   - Patients with large hearts and increased resting cardiac output tolerate hypovolaemia poorly but are also susceptible to fluid overload.

Postoperative concerns

Most patients with CHD should be nursed in a high-care environment postoperatively, particularly if they have reactive pulmonary arteries. Avoidance of hypoxaemia, hypercarbia, acidosis, hyperthermia, hypothermia and sympathetic stimulation will avoid cardiac complications.

Summary

Children with CHD are at increased perioperative risk when presenting for non-cardiac surgery. A risk assessment may be useful in deciding whether the child should be referred to a specialised facility.

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