Anaesthetic implications of congenital heart disease for children undergoing non-cardiac surgery

Savini Wijesingha

Michelle White

Abstract

Children with congenital heart disease (CHD) are at increased risk of morbidity, perioperative cardiac arrest and 30-day mortality from major and minor surgical procedures compared to healthy children. Factors associated with greatest risk are the complexity of cardiac disease and the physiological status of the child. Therefore the anaesthetist must understand balanced and single ventricle circulations and be able to assess the effects of four major complications of CHD: cardiac failure, arrhythmias, pulmonary hypertension and cyanosis. Other risk factors include type of surgery, age, ASA physical status and length of *preoperative* hospital stay. Because of the diversity of CHD and the range of surgical procedures a generalized approach to anaesthesia is impossible. Preoperative assessment, induction and maintenance should all be individualized to the child and tailored to the type of surgery. Whether surgery occurs in the local hospital or tertiary cardiac centre depends on risk classification (high, intermediate and low). Some children require full cardiac anaesthesia, intensive care and cardiology support making care in the local hospital inappropriate, whereas for others, care in their local hospital is both safe and convenient.

Keywords Anaesthesia; cardiac; children; congenital heart disease; non-cardiac surgery; paediatric

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Learning objectives

After reading this article, you should be able to:

- list the factors associated with a high and intermediate risk of perioperative complications in children with CHD undergoing non-cardiac surgery
- outline the four categories of 'complex' disease
- list four major complications of CHD
- describe the physiology of balanced and single ventricle circulations
- describe the key features in preoperative assessment

The scope of the problem

The child with congenital heart disease (CHD) presenting for non-cardiac surgery is becoming increasingly common. CHD is the most frequently occurring birth defect (approximately 1:125 live births) and due to advances in medical care, 90% of children with CHD now survive to adulthood. Therefore they present to hospital requiring surgery because they are subject to the same childhood illnesses and injuries as healthy children. However, children with CHD have increased risk of perioperative morbidity, cardiac arrest and a higher 30-day mortality.[1,2]

A one-size-fits-all approach to anaesthesia is impossible due to the wide range of CHD and different types of surgery. Children with CHD for non-cardiac surgery present a unique set of challenges. This article aims to outline a physiological and evidence-based approach to the anaesthetic management of these children.

Risk classification

Several studies have identified factors associated with a high risk of perioperative complications. The most important factors are the complexity of the cardiac lesion and the physiological status of the child at the time of surgery. Previous corrective cardiac surgery, whether complete or palliative, does not significantly alter the postoperative outcome.[3] Physiologically well-compensated patients with CHD can undergo elective operations at minimal risk, whereas poorly compensated patients undergoing urgent or major operations are at high risk.[3] In addition to complexity and physiological status, other risk factors are type of surgery, age, length of hospital stay *before* surgery, and ASA physical status IV or V. On the basis of the available evidence, a pragmatic attempt has been made to classify children as high, intermediate and low risk in order to aid management (Table 1).[4] Individual risk factors are discussed below.

Complex and simple lesions

Children at highest risk of perioperative complications are those with the following complex lesions:[1]

- balanced circulation physiology
- single ventricle physiology
- cardiomyopathy (severe cardiac failure)
- aortic stenosis.

Normal (or series) circulation physiology

The normal heart has a pulmonary and systemic circulation that work together in series (Figure 1). Some forms of CHD, such as atrial septal defect (ASD), have the usual 'series' circulation but one or more 'holes' exist so mixing of deoxygenated and oxygenated blood occurs. The direction blood flows through the lesion depends on the pressure gradient and is documented in the echocardiography report. The amount of shunting depends on the size of the defect and the relative pressure gradients. Changes

Savini Wijesingha *MB ChB FRCA is a Specialist Registrar in Anaesthetics and Intensive Care, training in Edinburgh, UK. Conflict of interests: none declared.*

Michelle White *MB ChB DCH FRCA is a Consultant Anaesthetist and Deputy Chief Medical Officer for the International Charity Mercy Ships and is currently based in Madagascar. Dr White was a Consultant in Paediatric Cardiac Anaesthesia and Paediatric Intensive Care at University of Bristol Children's Hospital from 2007 – 2012, UK. Conflict of interests: none declared.*

Risk classification of children with heart disease undergoing non-cardiac surgery

High risk	Intermediate risk	Low risk
Complex lesions	Major surgery	Simple lesions
Physiologically poorly compensated and/or presence of major complications	Under 2 years old	Physiologically normal or well compensated
	Emergency surgery	Minor (or body surface) surgery
	Preoperative hospital stay more than 10–14 days	Over 2 years old
	ASA physical status IV or V	Elective surgery
		Preoperative hospital stay less than 10 days
		ASA physical status I – III

Table 1

in systemic (SVR) and pulmonary (PVR) vascular resistance as a result of anaesthesia have greatest impact on large, unrestrictive defects.

Balanced (or parallel) circulation physiology

Instead of the pulmonary and systemic circulation being in 'series', they may be in 'parallel' (Figure 2). Large intra- and/or extra-cardiac communications mean that not only does mixing of oxygenated and deoxygenated blood occur, but also that blood flow to the systemic and pulmonary circulation varies depending on the relative resistances in each circuit. Thus blood flow to the lungs or to the body is a 'balance' which depends on PVR and SVR, hence the term 'balanced' circulation. Excessive pulmonary blood flow (PBF) results in pulmonary oedema and poor systemic perfusion (which may compromise coronary and splanchnic perfusion); insufficient PBF causes profound cyanosis. These children can be very difficult to manage and liaison with a regional paediatric cardiac centre is advised.

Infants with large unrepaired AVSD or VSD lesions can exhibit 'balanced' circulation physiology. These infants have predominantly left-to-right shunt flow. High concentrations of oxygen will increase PBF and reduce systemic perfusion; conversely large doses of induction agent may reduce SVR so much that shunt flow is reversed causing desaturation. The excessive PBF also makes the child vulnerable to developing pulmonary hypertension. Therefore pulmonary hypertensive crisis is another cause of sudden desaturation during anaesthesia.

Other examples of 'balanced' circulations include children with truncus arteriosus (TA), hypoplastic left heart syndrome (HLHS) or a modified Blalock-Taussig (BT) shunt. A BT shunt is performed to augment PBF in infants whose PBF is otherwise insufficient. The modified BT shunt usually consists of a gortex tube positioned between the right subclavian artery (RSCA) and right pulmonary artery (RPA). Blood flows down the pressure gradient from RSCA to RPA. However, an increase in PVR and decrease in SVR (as may occur during a hasty induction of anaesthesia with excessive induction agent, and a mild degree of hypoxia and hypercarbia) may seriously compromise shunt flow resulting in reduced cardiac output and even cardiac arrest.

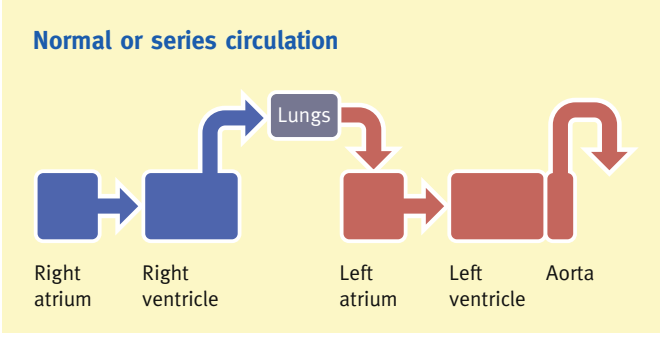

Figure 1

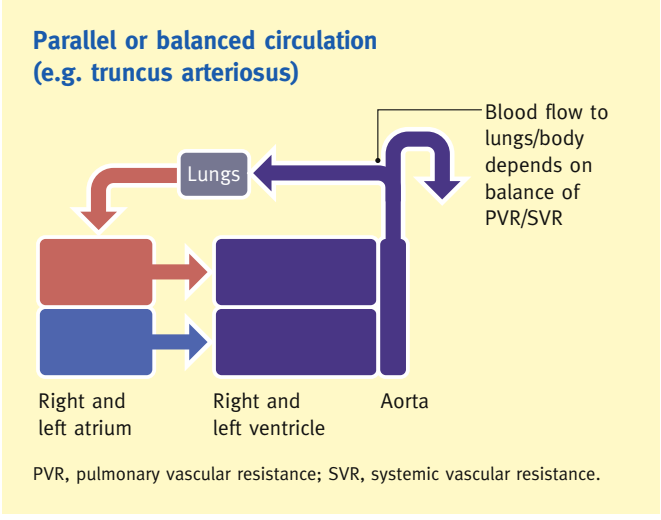

Figure 2

Single ventricle physiology

Some forms of CHD are not amenable to full anatomical correction (i.e. a biventricular repair resulting in a normal 'series' circulation). Therefore these children will be palliated by creating a single ventricle circulation (Figure 3). This is created as a staged process. The first stage known as a Glenn shunt or bidirectional cavopulmonary shunt (BCPS) is usually performed at 3–5 months of age. The superior vena cava (SVC) is connected directly to the right pulmonary artery (RPA) and any previous BT shunt ligated or removed. The child remains cyanosed after this procedure (oxygen saturations 75–85%). The second stage connects the inferior vena cava (IVC) to the RPA thereby