

# Anaesthesia in Children With Congenital Heart Disease for Noncardiac Surgery

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## KEY POINTS

- A physiological approach to understanding congenital heart disease (CHD) is helpful for anaesthetic planning.
- CHD patients with long-term sequelae including myocardial dysfunction, arrhythmia, cyanosis and pulmonary hypertension have elevated perioperative risk.
- High-risk red flags for noncardiac surgery include single-ventricle circulation, parallel circulation, aortic stenosis and cardiomyopathy.
- In patients with CHD, mortality risk is 5 times higher for major versus minor surgery.

## INTRODUCTION

Congenital heart disease (CHD) is the most common congenital defect, with an incidence of 8 in 1000 live births.<sup>1</sup> Advances in medical and surgical treatment have significantly improved survival. In high-income countries, nearly 90% of children born with CHD survive into adulthood. In low- and middle-income countries (LMICs), the lack of robust diagnosing and reporting does not allow for accurate estimates of how many CHD patients survive to adulthood.<sup>2</sup> However, diagnosis and surgical repair of simple CHD lesions are increasing in LMICs.<sup>3</sup> Regardless of setting, with improved survival, more patients with CHD will present for noncardiac surgery.

The anaesthetic management of patients with CHD can be daunting. This tutorial builds on the review article by Raviraj et al. by using case-based discussion to demonstrate how a practical physiological and risk-based framework can be applied to the anaesthetic management of a child with CHD presenting for noncardiac surgery.

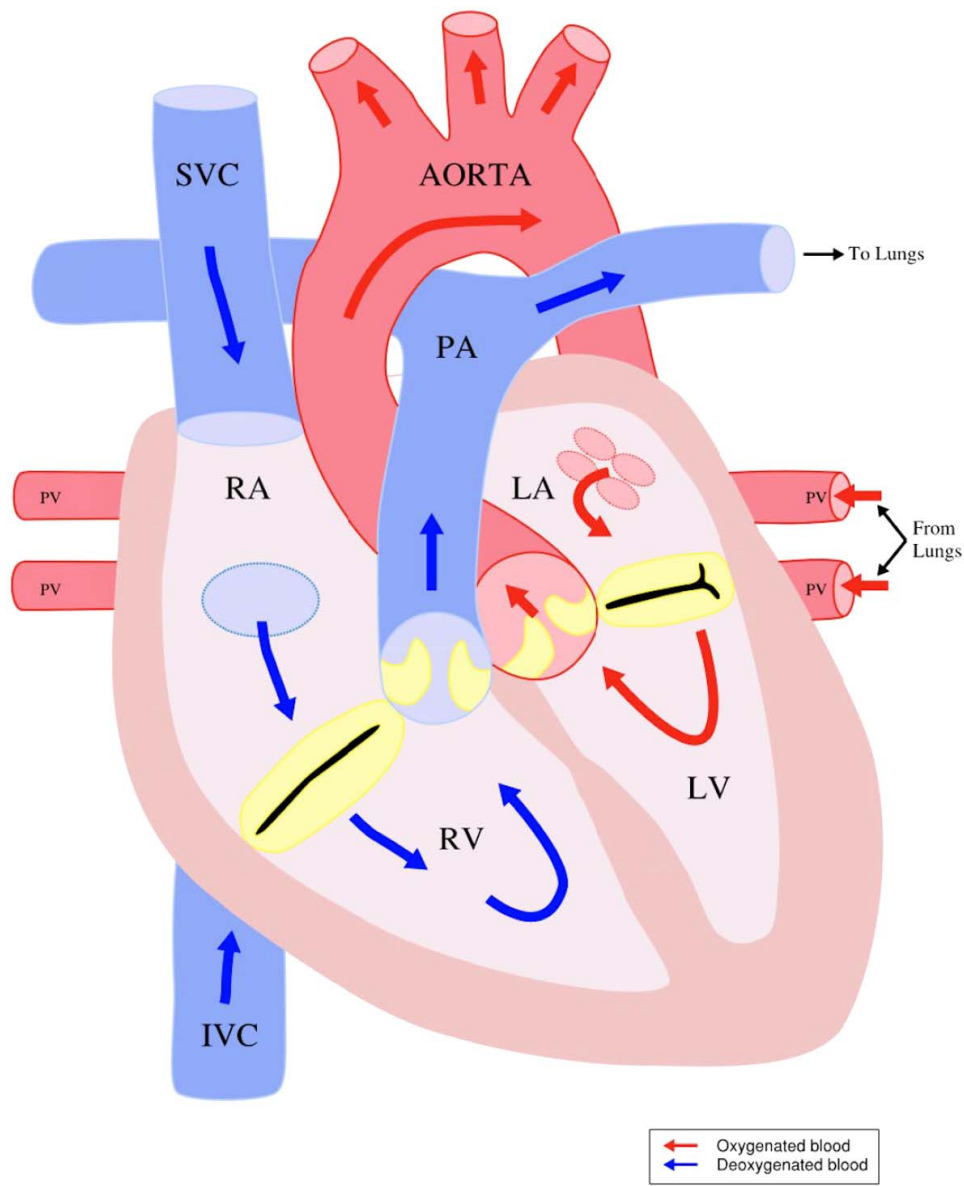
## PHYSIOLOGICAL APPROACH TO CLASSIFICATION OF CHD

Classification of CHD can be performed using anatomic or physiologic approaches. An anatomic approach focuses on structural abnormalities, whereas a physiological approach focuses on haemodynamic abnormalities and clinical consequences created by the lesion.<sup>3,4</sup> Using the physiologic approach, CHD can be classified into 1 of 3 physiologic

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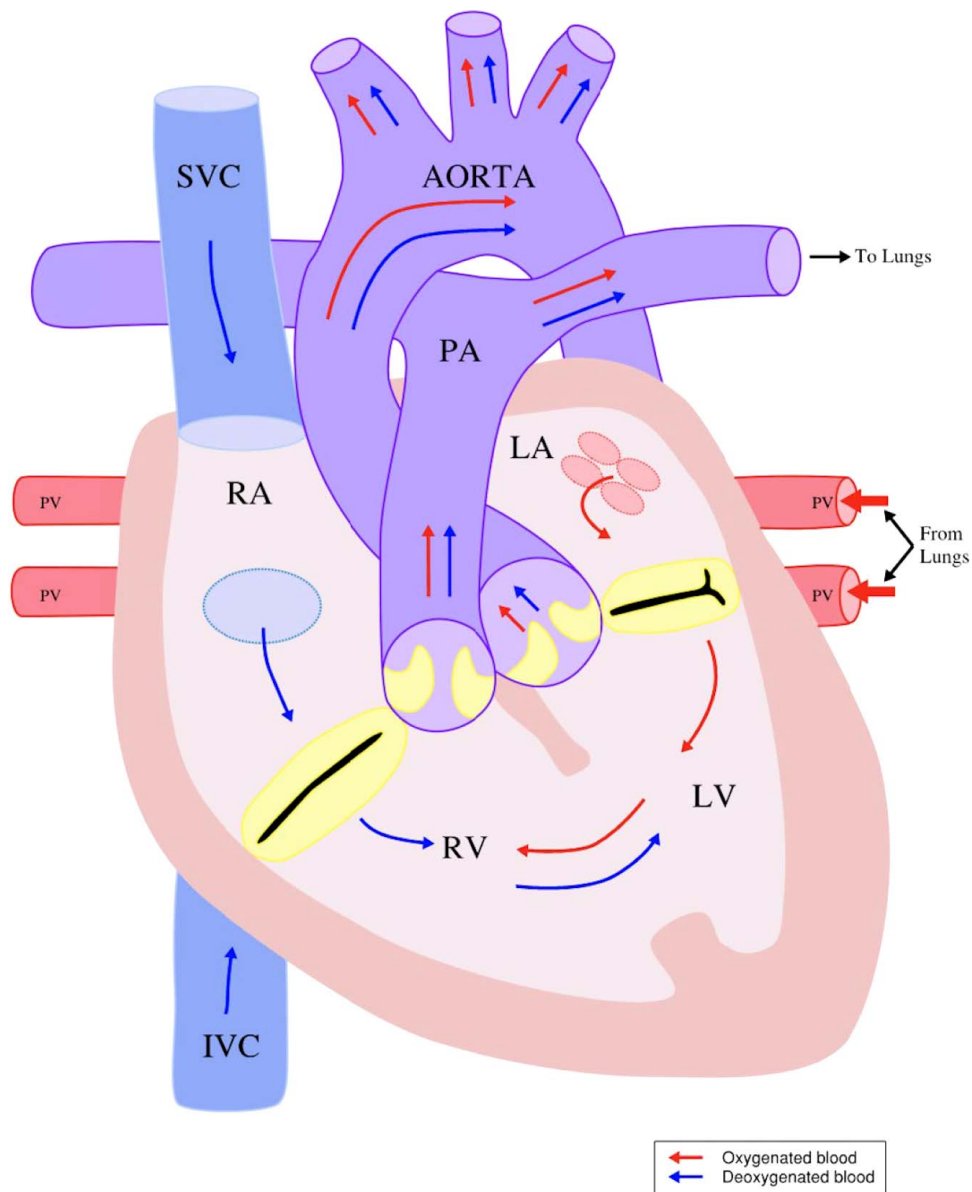
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**Figure 1.** Normal/series circulation. Anaesthesia in children with congenital heart disease for noncardiac surgery. Reprinted from Raviraj et al.<sup>4</sup> with permission of *Update in Anaesthesia*.

categories. In normal/series circulation (Figure 1), deoxygenated blood returns to the right side of the heart and travels to the lungs through the pulmonary arteries. This oxygenated blood returns to the left side of the heart and travels via the aorta to the rest of the body. In parallel/balanced circulation (Figure 2), the 2 circulations are not separated. Oxygenated and deoxygenated blood completely mix, and the amount of pulmonary blood flow (PBF) and systemic blood flow (SBF) is determined by the difference between pulmonary vascular resistance (PVR), systemic vascular resistance (SVR) or the presence of obstructive lesions. The volume of PBF to SBF is expressed as the Qp:Qs ratio. A Qp:Qs of 2:1 means there is twice as much PBF as SBF. Increased PBF causes heart failure, pulmonary congestion, respiratory infections, failure to thrive and damage to the pulmonary arteries over time. Low Qp:Qs results in cyanosis. The third category is single-ventricle circulation (Figure 3). In these lesions, there is only 1 functional ventricle, which prevents the child from undergoing a “biventricular” repair to establish a normal/series circulation. Instead, palliative procedures are performed to allow the 1 ventricle to pump blood in series through the aorta, to the body, and the blood then flows down the pressure gradient through the lungs via the pulmonary arteries and back to the heart. PBF is dependent on the transpulmonary gradient. Understanding the physiological classification allows the



**Figure 2.** Parallel/balanced circulation (large VSD). Anaesthesia in children with congenital heart disease for non-cardiac surgery. Reprinted from Raviraj et al.<sup>4</sup> with permission of *Update in Anaesthesia*.

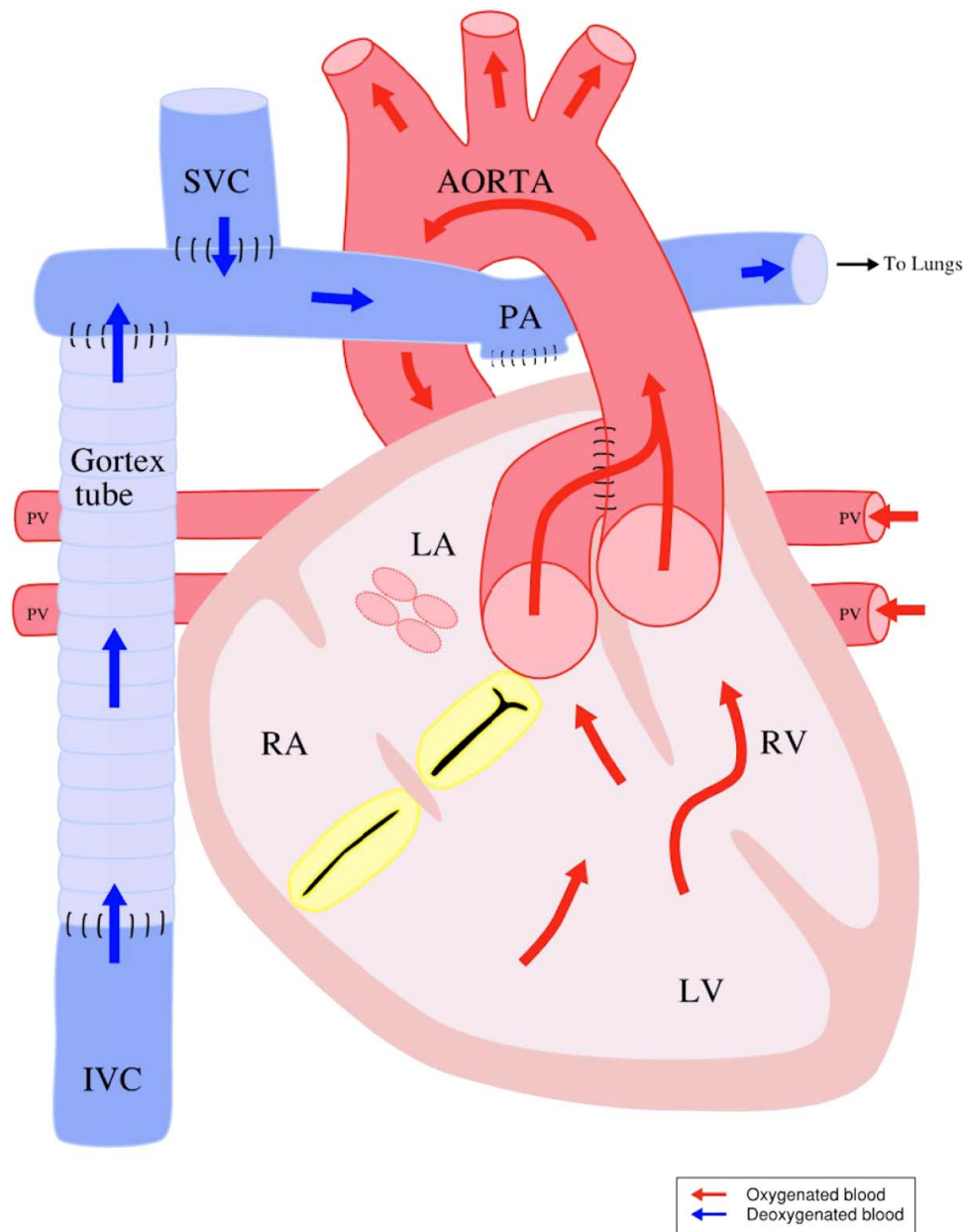
anaesthesia providers to predict the effects of anaesthetic agents and ventilation strategies on SBF and PBF.<sup>5</sup> For additional information regarding the physiologic classification, please refer to the article by Raviraj et al.

## LONG-TERM SEQUELAE OF CHD

Appreciation of the types of CHD and their potential long-term sequelae and impact on anaesthetic risk is essential when caring for patients with CHD requiring noncardiac surgery. The most important long-term sequelae that increase perioperative risk are the following:

- myocardial dysfunction (including heart failure and cardiomyopathy),
- arrhythmia (including permanent pacing),
- cyanosis,
- pulmonary hypertension.<sup>6–8</sup>

Patients who have had definitive repair of simple lesions (for example, atrial septal defect [ASD], ventricular septal defect [VSD], patent ductus artery [PDA]) are unlikely to have long-term sequelae. Patients with more complex repairs (for example,



**Figure 3.** Single-ventricle circulation. This figure specifically shows a Fontan circulation for a double inlet left ventricle. Staged single-ventricle palliative procedures resulting in Fontan circulation are performed for a variety of lesions, including hypoplastic left heart syndrome and unbalanced complete atrioventricular canals. Anaesthesia in children with congenital heart disease for non-cardiac surgery. Reprinted from Raviraj et al.<sup>4</sup> with permission of *Update in Anaesthesia*.

valve repair and single ventricle palliation) are more likely to have long-term sequelae. For additional information on the 4 key long-term sequelae, please refer to the article by Raviraj et al.

## RISK STRATIFICATION

Risk stratification of patients with CHD for noncardiac surgery is complicated because of the heterogeneity of cardiac lesions and surgical procedures. A pragmatic approach is summarized in Table 1 and involves consideration of 3 factors:

- Assessment of CHD complexity,
- Identification of long-term sequelae and high-risk features associated with poor perioperative outcomes,
- Assessment of the risks associated with the surgical procedure.

|   | High Risk             | Intermediate Risk | Low Risk |
|---|-----------------------|-------------------|----------|
| ACS NSQIP Paediatric CHD Classification               | Severe                | Major             | Minor    |
| Presence of long-term sequelae or a high-risk feature | Presence of 1 or more | None              | None     |
| Surgical risk   | Major                 | Major             | Minor    |

**Table 1.** Noncardiac Surgery Risk Stratification

## Assessment of CHD Complexity

The paediatric database of the American College of Surgeons' National Surgical Quality Improvement Program (ACS NSQIP Paediatric) stratifies CHD patients based on the following definitions:

- Minor: Unrepaired asymptomatic acyanotic CHD (can be on medication) and repaired CHD with no residual hemodynamic abnormalities requiring no medications;
- Major: Unrepaired symptomatic acyanotic CHD and repaired CHD with residual hemodynamic abnormalities;
- Severe: Unrepaired cyanotic heart disease, pulmonary hypertension, ventricular dysfunction requiring medication and patients listed for heart transplant.

When patients in these categories were matched to patients without CHD using age, American Society of Anesthesiologist (ASA) class and surgical complexity, no difference in complications was found for minor category patients. There were increases in morbidity and mortality for patients with major and severe CHD.<sup>9</sup>

## Identification of Long-Term Sequelae and High-Risk Features

The following conditions significantly increase the risk of noncardiac surgery. If time, geography and local resources allow, these patients should be managed at a specialty centre that has on-site cardiology and paediatric cardiac anaesthesia expertise.<sup>6,7,10</sup>

- Single-ventricle circulation
- Parallel circulation
- Aortic stenosis
- Cardiomyopathy
- Heart failure
- Arrhythmias
- Cyanosis
- Pulmonary hypertension
- Age <2 years
- ASA class  $\geq 4$

## Assessment of Surgical Risk

Finally, the surgical procedure should be determined as minor or major. The mortality risk in patients with CHD is 5 times higher for major versus minor surgery.<sup>7</sup> The presence of 1 of the following places the surgery in the major category:

- Emergency surgery
- Intrathoracic surgery
- Intraoperative surgery
- Vascular reconstructive surgery
- Orthopaedic and neurosurgical procedures, with the potential for significant blood loss

## PREOPERATIVE ASSESSMENT

Preoperative assessment of patients with CHD should begin with the physiologic classification and risk assessment as discussed above. From that starting point, additional information should be determined as outlined below.

- Physiologic classification of CHD
- Risk assessment
- Review of most recent cardiac evaluations: recent electrocardiogram (EKG), echocardiogram, cardiac catheterization and cardiology visit
- Physical examination: vital signs, work of breathing, growth curve, perfusion, fluid status
- Determining whether the patient has comorbidities
- Determining whether the patient requires endocarditis prophylaxis<sup>11–13</sup>

The guidelines vary by region. Please refer to the guidelines followed in your country of practice (guideline websites are listed in references 11-13). All regions have decreased the use of prophylaxis over time. Infective endocarditis is a rare but potentially catastrophic disease. Evidence suggests it is more likely to develop from low-level repeated bacteraemia during daily life than from 1 high-level exposure with a procedure. Good dental hygiene is most important in reducing endocarditis risk.

The American Heart Association (AHA) limits prophylaxis to patients at the highest risk for adverse outcomes from endocarditis who are undergoing procedures at high risk for bacteraemia. The European Society of Cardiology (ESC) also limits prophylaxis to high-risk patients, although the categories are slightly different. The UK National Institute for Health and Care Excellence (NICE) guidelines no longer recommend endocarditis prophylaxis. The following need to be addressed:

- Preoperative logistical planning:
- Timing of the procedure
- Location of the procedure
- Recovery plan
- Whether the patient should be transferred to a higher level of care hospital. For urgent and emergent surgeries, is there time?

oFor elective surgeries, high-risk patients should be transferred to a specialty centre. Low-risk patients can likely be managed anywhere. For intermediate-risk patients, there should be preoperative discussion with a specialty centre to determine the appropriate location for care.<sup>14</sup>

oFor emergency surgeries, low-risk patients should be managed where they are. For intermediate- and high-risk patients, discussion should occur with a specialty centre if time allows, and transfer should be discussed.<sup>14</sup>

## ANESTHETIC CONSIDERATIONS

Patients with CHD have been safely managed with general anaesthesia, neuraxial anaesthesia, regional anaesthesia and sedation. The available research does not recommend a specific technique or medication. In designing an anaesthetic plan, begin with your preferred anaesthetic plan for the specific procedure. Consider how that plan would affect the hemodynamic goals of your patient. Will the patient be able to maintain appropriate cardiac output and oxygen delivery with that plan? In patients with parallel or balanced circulation, how will the anaesthetic plan change the Qp:Qs? If cardiac output is not sufficient during the procedure, what will be the best initial treatment (ie, fluid administration, vasoconstrictor, inotrope)?<sup>7,8</sup> Table 2 provides additional hemodynamic details and considerations of select anaesthetic agents. Table 3 reviews factors that affect Qp:Qs.

Important anaesthetic considerations in patients with CHD include “bubble precautions” and vascular access. Patients with residual pulmonary to systemic connections (for example, VSD, ASD, PDA) are at risk for systemic embolization of air; therefore, careful “de-airing” of fluids and medications is required. Vascular access may be difficult in patients with CHD due to diuresis and/or prior procedures. Review the patient’s record for any known vascular occlusions due to prior interventional cardiology procedures or invasive monitoring lines. If access is suspected to be challenging, have resources available, such as ultrasound and additional providers. In patients at high risk for haemodynamic collapse with induction (for example, severe aortic stenosis, severe systolic dysfunction), consider obtaining intravenous access prior to induction.<sup>8</sup>

## LET’S PRACTICE

A 2-year-old boy with history of tetralogy of Fallot (TOF) who underwent repair at 4 months presents for dental rehabilitation. A recent echocardiogram reveals normal valve and cardiac function and no residual VSD. The patient is active and keeps up with his peers. His recent EKG shows sinus rhythm with a right bundle branch block (RBBB) and no ventricular ectopy. The RBBB is a result of the VSD patch and is not problematic. Patients with repaired TOF are at risk for ventricular arrhythmias and sudden cardiac death as they age. If ventricular ectopy is seen on a preoperative EKG, consultation with the patient’s cardiologist should occur.

*CHD physiologic classification:* This patient has been repaired and has a normal/series circulation

*NSQIP category:* Minor, repaired with no residual hemodynamic abnormalities and no medications

*Presence of long-term sequelae or high-risk feature:* None

*Surgical risk:* Minor

*Overall risk:* Minor

*Anaesthetic plan:* This elective procedure could be performed safely at any centre able to manage paediatric patients. Induction of anaesthesia could be accomplished in whichever manner is desired. Endocarditis prophylaxis is not required for this patient under any of the major guidelines (AHA, ESC or NICE).

A 3-month-old boy with unrepaired large-sized VSD presents for emergency repair of testicular torsion. He has a history of sweating while feeding and failure to gain weight and is now fed via a nasogastric tube and is on maximal diuretic therapy. A recent echocardiogram shows left-to-right shunt with normal cardiac function.

*CHD physiologic classification:* This patient with a large VSD has parallel or balanced circulation.

*NSQIP category:* Major, unrepaired acyanotic CHD with symptoms

| Agent           | Effects  | Considerations  |
|-----------------|--|---|
| Sevoflurane     | High dose: ↓ SVR, ↓ contractility<br>Low dose: ↔ SVR, ↔ contractility                                      | Avoid prolonged exposure to a high concentration. Expect a slower induction in patients with myocardial dysfunction and/or right-to-left shunt.   |
| Propofol        | ↓↓↓ SVR, ↔ contractility, respiratory depression   | In patients with shunts, the decreased SVR will increase right-to-left shunt and in high, rapid doses may cause shunt reversal in patients with left-to-right shunts. Must be used slowly and carefully in high-risk patients.  |
| Ketamine        | ↔ SVR, ↔ PVR, ↔ contractility, ↔ respiratory rate, ↑ heart rate (HR), analgesia                            | Effective anaesthetic with virtually no haemodynamic effects. In patients with significant myocardial dysfunction, a direct myocardial depressant effect has been reported, although ketamine is still widely used in this group of patients. Especially useful in patients with right-to-left shunt. |
| Etomidate       | ↔ SVR, ↔ PVR, ↔ contractility, adrenal suppression   | Effective anaesthetic with virtually no haemodynamic effects but adrenal suppression limits use. Consider use in patients with significant myocardial dysfunction if concerned about the use of ketamine.   |
| Opioids         | ↔ SVR, ↔ contractility, ↓ HR, ↓ respiratory rate   | Cardiovascularly very stable, even in high doses, but in nonintensive care patients, using high doses is limited by respiratory depression.   |
| Benzodiazepines | ↔ SVR, ↓ contractility, ↔ HR   | Adjuvant to reduce concentration of inhalation or intravenous agents needed to induce and maintain anaesthesia. Use caution in patients with severe myocardial dysfunction due to negative effects on myocardial contractility.   |
| Dexmedetomidine | ↔ contractility, ↓ HR, ↓ sinus and AV node conduction, ↔ respiratory rate, can cause hypo- or hypertension | Useful as an adjuvant to reduce concentration of inhalational or intravenous agents needed to induce or maintain anaesthesia. Use caution in patients with bradycardia or heart block.  |

**Table 2.** Anaesthetic Agent Considerations in CHD<sup>7,8</sup>

| Resistance    | Factor  |
|---------------|---|
| Increased PVR | Hypoxia   |
|               | Hypercarbia   |
|               | Acidosis  |
|               | Increased intrathoracic pressure  |
|               | Atelectasis   |
|               | Increased haematocrit   |
|               | Hypothermia   |
| Decreased PVR | Sympathetic stimulation (pain)  |
|               | Supplemental oxygen administration  |
|               | Hypocarbida   |
| Increased SVR | Alkalosis   |
|               | Decreased haematocrit   |
|               | Hypothermia   |
|               | Sympathetic stimulation (pain)  |
| Decreased SVR | Vasoconstrictors  |
|               | Hyperthermia  |
|               | Sepsis  |
|               | Volatile anesthetics  |
|               | Propofol  |
|               | Decreased haematocrit   |
|               | Spinal or epidural anaesthesia (more pronounced in older children or adults than in younger children) |

**Table 3.** Factors Affecting Pulmonary and Systemic Vascular Resistance

*Presence of long-term sequelae or high-risk feature:* Yes, parallel circulation and age <2 years

*Surgical risk:* Major, due to it being an emergency

*Overall risk:* High, given emergency surgery, parallel circulation and age <2 years

*Anaesthetic plan:* If the patient presented to a local centre, discussion should occur with the nearest specialty centre to determine if there is time to transfer. If there is not time to transfer, consultation should occur to assist the local centre with anaesthetic management. Slow inhalational or intravenous induction should be used. Limit oxygen concentration as it is a potent pulmonary vasodilator that can lead to increased PBF and heart failure. Perform careful “de-airing” to avoid bubbles. Pulmonary compliance may be reduced because of pulmonary oedema. Minimize fluids. Endocarditis prophylaxis is not required under any of the major guidelines, as this is not a high-risk procedure for endocarditis.

An 8-year-old girl with history of hypoplastic left heart syndrome palliated to Fontan circulation presents with severe abdominal pain, nausea and vomiting. She requires an emergency laparoscopic appendectomy. Prior to this illness, she was active and only tired with strenuous activity. A recent echocardiogram showed a patent Fontan pathway and normal ventricular function. Her EKG in the emergency department shows sinus tachycardia.

*CHD physiologic classification:* Single-ventricle circulation. In this case, the right ventricle pumps oxygenated blood to the body, via the aorta. Deoxygenated blood returns from the head and body via the superior and inferior vena cava, which are connected directly to the pulmonary arteries. PBF is driven by the difference between systemic venous pressure and end-diastolic pressure of the single ventricle, termed the *transpulmonary gradient*. If the systemic venous pressure decreases due to hypovolemia or anaesthetic medications or the end-diastolic pressure increases due to decreased cardiac function, flow through the lungs will decrease.

*NSQIP category:* Severe

*Presence of long-term sequelae or high-risk feature:* Yes, single-ventricle circulation

*Surgical risk:* Major due to it being an emergency, intraperitoneal surgery

*Overall risk:* High given single-ventricle circulation and emergency, intraperitoneal surgery

*Anaesthetic plan:* If the patient presented to a local centre, discussion should occur with the nearest specialty centre to determine if there is time to transfer. If there is not time to transfer, consultation should occur to assist the local centre with anaesthetic management. This patient has an acute abdomen and is vomiting and therefore should undergo an intravenous rapid sequence induction and intubation to reduce the risk of aspiration. The profound decrease in SVR and resultant decrease in preload caused by bolus dosing of propofol is unlikely to be tolerated. Etomidate or ketamine would be reasonable. This patient is likely hypovolemic, and a fluid bolus prior to induction is advised. Increased intrathoracic pressure with initiation of positive-pressure ventilation will negatively affect flow through the Fontan pathway. Measures should be taken to minimize mean airway pressure while optimizing oxygenation and ventilation and minimizing atelectasis. The laparoscopic approach can decrease pain and recovery time but has significant haemodynamic effects on the Fontan circulation. Insufflation of the abdomen decreases preload and increases intrathoracic pressure, which have negative effects on the transpulmonary gradient and reduce PBF. Hypercarbia from insufflation with CO<sub>2</sub> will increase PVR and decrease PBF. Use the lowest pressures that allow for surgical visualization and closely monitor haemodynamics with a plan to convert to an open approach if insufflation is not tolerated.<sup>15</sup>

## SUMMARY

As CHD survival continues to improve, more patients with CHD will present for noncardiac surgery. Classifying CHD using a physiological approach facilitates anaesthetic planning. By understanding the physiology, anaesthesia providers can choose the most appropriate anaesthetic agents to optimize haemodynamics. Risk stratifying patients allows for early identification of high-risk patients.

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