Anaesthetic Management of Adults with Congenital Heart Disease Presenting for Noncardiac Surgery

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KEY POINTS

- Adult congenital heart disease confers significant morbidity and mortality.
- Lesions can be stratified as simple, moderate, and complex in terms of prognosis and outcomes.
- Comorbidities impact perioperative planning for noncardiac surgery, and specialist input should be sought for moderate and complex lesions.
- A thorough understanding of the underlying physiology is necessary to ensure a good perioperative outcome.
- No single anaesthetic technique is better than another and should be tailored to the patient.

INTRODUCTION

Congenital heart disease (CHD) is the most common birth defect in the United Kingdom, affecting approximately 0.8–0.9% of live births.[1,2] In addition, some conditions of CHD are not diagnosed until later in life, for example, bicuspid aortic valve disease. Before the development of paediatric cardiac surgery, less than 20% of children with CHD survived to adulthood; now, adults account for 66% of the CHD population.[3] Therefore, more of these patients will be presenting for noncardiac surgery in the future. Each patient with adult CHD (ACHD) is unique, but all have an increased risk profile during noncardiac surgery. ACHD patients can be classified as simple, moderate, and complex in terms of outcomes and level of care required (Table). Although stable patients with simple lesions can likely undergo noncardiac surgery in a nonspecialist setting, those with more advanced lesions, especially complex ones, should have specialist input perioperatively. Some schematic depictions of complex lesions are presented in Figure.[4]

Identifying these patients, understanding their cardiac anatomy, and determining their functional capacity is essential before proceeding with surgery. Anaesthetic management must be individualised according to lesion, age, corrective surgery, remaining defects, long-term complications, and previous cardiac history. ACHD patients are at high risk of cardiac arrhythmias in the first 24 hours postoperatively.[5] It is important to meticulously plan their management including need for any pacemakers and implanted defibrillators. Knowing and anticipating the significance of the cardiovascular effects of various anaesthetic medications is essential, especially with heart failure. Ventilatory strategies to minimise changes in blood flow distribution in the presence of cardiac

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TAKE ONLINE TEST

Simple ACHD		Moderate-Complexity ACHD	Severe-Complexity ACHD
Unrepaired lesions	**Repaired lesions**	Aorta to left ventricular fistula	Conduits
Isolated aortic valve disease	Previously ligated or occluded ductus arteriosus	Partial or total anomalous pulmonary venous drainage	Cyanotic heart disease
Isolated mitral valve disease	Repaired sinus venosus or secundum atria septal defect without residual defect	Coarctation of aorta	Any single ventricle circulation
Isolated patent foramen ovale	Repaired ventricular septal defect without residual defect	Ebstein's anomaly	Double-outlet ventricle
Small atrial septal defect or ventricular septal defect	Balloon valvuloplasty	Significant infundibular right ventricular outflow tract obstruction	Fontan procedure
Mild pulmonary stenosis		Unrepaired ductus arteriosus	Eisenmenger syndrome
		Moderate to severe pulmonary stenosis or regurgitation	Mitral, tricuspid, or pulmonary atresia
		Aortic stenosis	Transposition of great vessels
		Tetralogy of Fallot	Truncus arteriosus
		Ventricular septal defect with associated anomaly	Pulmonary hypertension

Table. Examples of Simple, Moderate, and Severe Adult Congenital Heart Disease.[4] ACHD, adult congenital heart disease

shunts is also important. The optimal centre to conduct surgery and perioperative care, with resources including specialist cardiothoracic anaesthesia and critical care, is vital for the best outcomes. This tutorial describes an approach to preoperative assessment, anaesthetic care, and postoperative management.

PREOPERATIVE ASSESSMENT

Understanding the anatomy and physiology of the individual lesion is essential, in addition to associated heart failure, arrhythmias, and comorbidities. Lesions with raised pulmonary vascular resistance (PVR) represent the highest risk and should be discussed with the specialist team before surgery.[4]

For elective surgery, patients should attend a preoperative anaesthesia clinic and have a multidisciplinary team meeting with anaesthesia, critical care, cardiology, and surgery. Baseline electrocardiograms should be captured (many have abnormalities) and echocardiography performed to assess cardiac function.[6]

Common comorbidities include heart failure, arrhythmias, endocarditis, pulmonary hypertension (PHTN), neurological complications, and haematological disease (bleeding/thrombosis/anticoagulation). Approximately one-third of CHD patients have extracardiac anomalies, such as can occur in vertebral defects, anal atresia, cardiac defects, tracheo-oesophageal fistula, renal anomalies, and limb abnormalities and Down's syndrome. Understanding the implications of such extracardiac anomalies for noncardiac surgery is important.[7]

These patients may have a fragile emotional state as they transition from paediatric to adult services, as this represents a difficult, uncertain time.[8] Anxiety and depression are more common than in the general population.[9] In those who have been managed as children, liaising with the patient's family/primary carer, if appropriate, is vital as they have been strongly influential in shaping their paediatric care and usually clearly understand the anatomy, physiology, and comorbidities. Providing appropriate perioperative psychological support for these patients is beneficial.[10]

A postoperative high dependency or intensive care bed for cardiac monitoring must be available before proceeding with surgery.[3] Cardiac arrhythmias can be detrimental in ACHD and need to be identified and managed immediately.[11]

ANAESTHETIC TECHNIQUE

Patients with ACHD represent a spectrum, from patients with near-normal physiology to those with complex physiology and markedly reduced cardiorespiratory reserve. Conventional anaesthetic management may be appropriate in well patients.[12] In complex cases, the anaesthetic must be modified.[3]

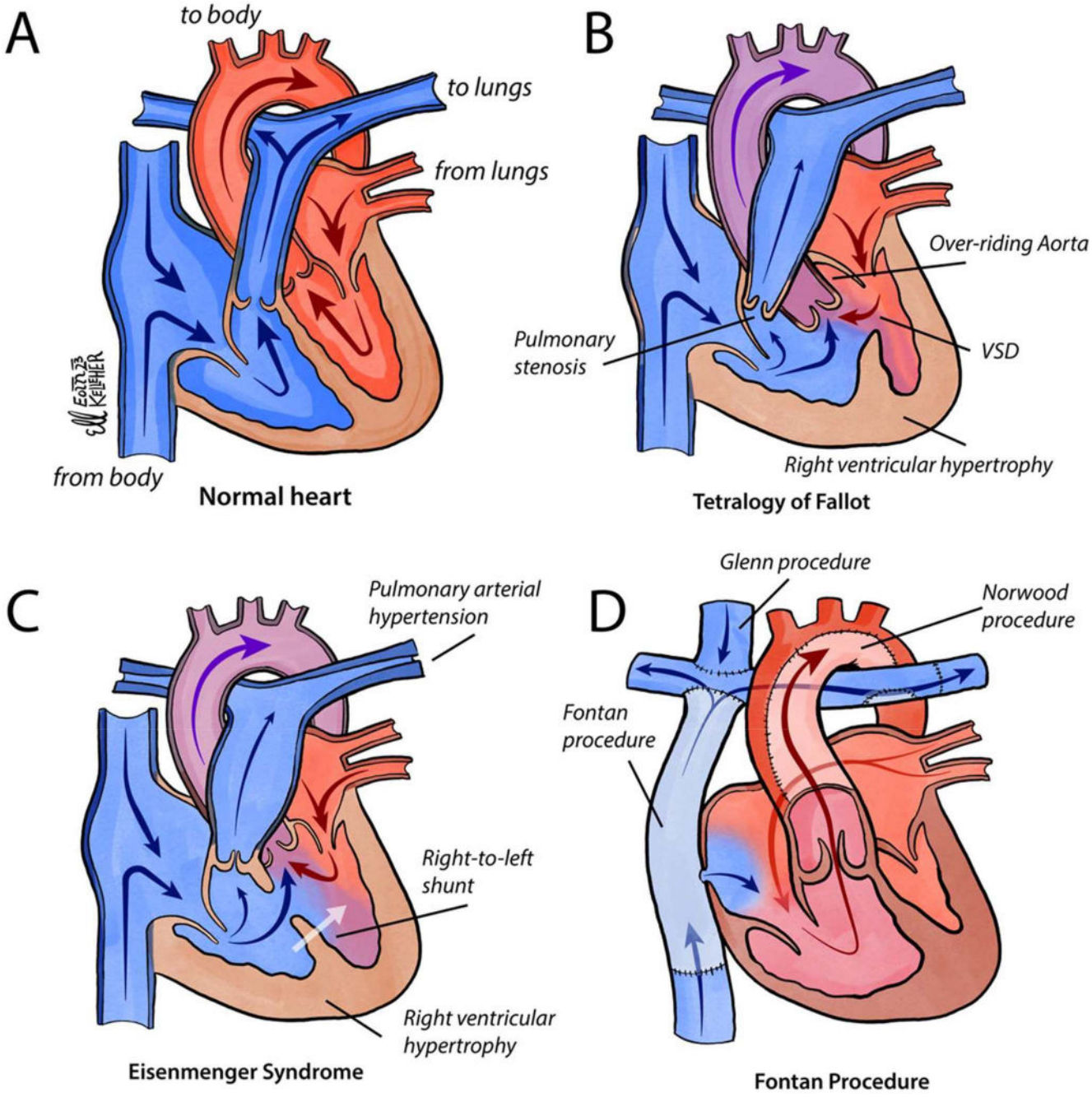

Figure. (A-D) Schematics of normal (A) and common unrepaired (B and C) and repaired (D) adult congenital heart disease cardiac circulations. Abbreviation: VSD, ventricular septal defect. Illustration by Dr Eoin Kelleher

Sedative premedication is frequently administered to reduce physiological stress and the amount of anaesthetic agents used on induction; this effect can be beneficial in severe disease. However, beware with cyanotic heart disease or PHTN, as hypercarbia associated with hypoventilation will increase PVR and the risk of pulmonary hypertensive crisis.

Maintenance of general anaesthesia involves using either a combination of inhalational and intravenous anaesthesia or total intravenous anaesthesia. There is no evidence supporting one technique over another in ACHD patients undergoing noncardiac surgery.

Regional anaesthesia offers haemodynamic stability but is limited to certain surgeries and by coagulation risk. These patients may be young and anxious, and regional anaesthesia alone may not be appropriate and may need to be combined with anxiolytics.

Neuraxial anaesthesia is well established in ACHD patients, but anticoagulation may limit usefulness. Additionally, the potential decrease in systemic vascular resistance (SVR), commonly more pronounced in spinal anaesthesia, is high risk

in patients with left ventricular outflow obstruction due to fixed stroke volume, which may have a secondary effect to reduce coronary artery perfusion.

MONITORING AND EQUIPMENT

Standard monitoring, according to published guidelines (American Society of Anesthesiologists and Association of Anaesthetists of Great Britain and Ireland), including electrocardiogram, noninvasive blood pressure, oxygen saturation, and ventilation, should be used.[13] Vascular access can be difficult in patients who have had multiple operations and prolonged hospitalisations. Central venous access may be required where peripheral access is unavailable, but a significant thromboembolic risk exists for patients with Fontan circulation. A preoperative ultrasound with doppler can determine which vessels are patent, as these patients may have multiple central vessel thrombosis from prior surgeries. Arterial line insertion depends on lesion complexity and operative risk. In an unrepaired or previous coarctation, a left-sided arterial line may give falsely low blood pressures.

Pulmonary artery catheterization can be difficult because of anatomical abnormalities, and with right-to-left shunting, cardiac output measurement by thermodilution can be misleading. In patients with Glenn- or Fontan-type repairs, superior vena cava pressure should be the same as pulmonary artery pressure.

Transoesophageal echocardiography provides real-time assessment of preload, intracardiac shunting, and ventricular and valve function. For cardiac surgery, the utility of transoesophageal echocardiography is beyond question; in patients undergoing noncardiac surgery, it may be equally valuable. An experienced congenital cardiologist must be available to interpret it.

BLOOD FLOW, VENTILATION, AND OXYGENATION

When caring for ACHD patients, it is important to understand the interplay between pulmonary and systemic blood flow, ventilation, and oxygenation. PVR describes the resistance that must be overcome to flow through the pulmonary vasculature. In ACHD, in patients with PHTN in particular, PVR is chronically increased, and further increases must be avoided perioperatively. In patients with serial circulations (no mixing of systemic and pulmonary blood), increasing PVR can strain the right ventricle and lead to heart failure. In patients with shunt physiology or central mixing (eg, Fontan circulation), changes in PVR can affect the ratio of blood flow between the pulmonary and systemic circulations and must be taken into account. For more information on the ratio of blood flow between the pulmonary and systemic circulations, see ATOTW 511. In all cases, increases in PVR that restrict pulmonary blood flow lead to hypoxaemia.

Hypercapnia increases PVR, and ventilatory strategies should maintain normocapnia. During positive pressure ventilation, intrathoracic pressure increases thereby reducing venous return to the heart and increasing PVR, thus limiting pulmonary blood flow. Inspiratory duration has a greater effect than peak inspiratory pressure on these parameters. Maximal pulmonary blood flow is achieved by decreasing inspiratory duration, which can increase peak inspiratory pressure, so the best compromise has to be achieved to reduce inspiratory duration while limiting the increase in peak inspiratory pressure.

Different lesions may have different oxygen saturation targets. High saturations are usually the goal, but there can be exceptions. In patients with shunt physiology or central mixing, delivery of a high concentrations of oxygen can result in pulmonary vasodilation and alter the ratio of blood flow between the pulmonary and systemic circulations such that systemic hypotension ensues.[3] Therefore, in patients who are stable before admission, it would be prudent to target their baseline oxygen saturations.

Hypoxaemia may arise from inadequate pulmonary blood flow or hypoventilation. In patients with poor pulmonary perfusion, avoidance of dehydration, maintenance of SVR, control of PVR, and reduction in oxygen consumption are central to successful outcomes. When hypoventilation is the cause, this can result in hypoxaemic pulmonary vasoconstriction, precipitating or worsening PHTN, and should be avoided by ensuring adequate oxygenation.

Chronic hypoxaemia, which may be present in some lesions, has profound haematological effects on other organ systems. Polycythaemia is a compensatory response that improves oxygen transport at the expense of increased viscosity. Generally, the increased viscosity is well tolerated; however, these patients are at increased risk of thrombosis and stroke. On the other hand, they are also more prone to bleeding as the ratio of red cells to plasma and, hence, clotting factors in these patients is reduced, resulting in more pronounced bleeding.

Finally, chronic lung disease is relatively common in adults with repaired CHD and can affect any of the above factors. Knowing the baseline lung function, including pulmonary function tests, is useful.

INDUCTION DRUGS

The cardiovascular effects of intravenous induction agents are well known. Several are associated with a reduction in SVR, and if right-to-left shunting is present, the shunt will increase.[3] Right-to-left shunting delays uptake of inhalational agents; therefore,

inhalational induction is prolonged. For simple cardiac lesions with left-to-right shunting, sevoflurane does not change the degree of shunting.[14] However, this cannot be extrapolated to patients with complex CHD or right-to-left shunts.

Sevoflurane can be used for inhalation induction and/or maintenance of anaesthesia. High-dose sevoflurane can reduce SVR and myocardial contractility. A slower induction should be expected in patients with myocardial dysfunction or right-to-left shunting. In some parts of the world, halothane is still commonly used, especially in paediatric patients. Compared with sevoflurane, it seems to have a greater negative impact on haemodynamics. Given this and its well-known propensity for causing arrythmias, it seems prudent to avoid its use in patients with CHD.[15]

Propofol reduces SVR and myocardial contractility and causes respiratory depression. Decreased SVR will increase right-to-left shunts and, with rapidly administered doses, may cause shunt reversal in patients with left-to-right shunts. It must be administered slowly and judiciously in high-risk patients.

Ketamine or a single dose of etomidate are alternative anaesthetic agents with minimal haemodynamic effects. In patients with significant myocardial dysfunction, a direct myocardial depressant effect has been reported with ketamine, although it is still widely used in these patients.

Opioids maintain cardiovascular stability even in high doses. SVR and contractility are maintained. There can be a secondary bradycardia that may have implications for cardiac output. In high doses, where respiration is not controlled, use is limited by respiratory depression.

Benzodiazepines can be used as an adjunct to reduce concentration of inhalation or intravenous agents needed to induce and maintain anaesthesia. In patients with severe myocardial dysfunction, the negative inotropic effects must be anticipated.

Dexmedetomidine is a useful adjunct to general anaesthesia as an infusion as it provides analgesia and anxiolysis and can be useful in those at high risk of tachyarrhythmias.[16] It maintains respiratory drive and myocardial contractility, but bradycardia is common. Cautious use is imperative in patients with bradycardia or heart block.

VASOPRESSORS AND INOTROPES

These may be required to support the failing ventricle. These may include milrinone, phenylephrine, adrenaline, noradrenaline, and vasopressin. Each of these have different effects on PVR, contractility, and chronotropy, and these effects should be considered when deciding which to administer.

THROMBOPROPHYLAXIS

These patients may be taking anticoagulants or antiplatelet agents to maintain shunt patency and may have associated abnormalities of the intrinsic and extrinsic clotting systems. Platelet dysfunction may exist despite normal figures.

As above, patients with cyanosis may develop a compensated polycythaemia, which can increase the risk for thrombosis. Using low-molecular-weight heparin thromboprophylaxis, elastic stockings, and early mobilization after surgery are essential. Patients with Fontan circulation or Eisenmenger syndrome (Figure) are at increased risk of thrombosis. Eisenmenger's also carries an increased risk of bleeding, and, therefore, decisions about anticoagulation should be undertaken in consultation with a CHD specialist.[4]

ANTIBIOTICS

ACHD patients are at increased risk of bacterial endocarditis. The risk varies markedly with different lesions. Given the small risk of endocarditis associated with most lesions, combined with the anaphylaxis risk posed by antibiotics and the development of resistant organisms, it is now recommended that antibiotic prophylaxis be reserved only for those patients at highest risk.[13] However, it is important to consult with microbiology locally.

LAPAROSCOPIC SURGERY

Laparoscopic procedures generally have a lower morbidity and mortality risk than open surgeries, but may present special problems in ACHD. Carbon dioxide insufflation during laparoscopy can produce haemodynamic and ventilatory instability due to increased intra-abdominal pressure and hypercarbia. The high intra-abdominal pressure can reduce preload and increase afterload, ultimately decreasing cardiac output. The pressure can also impair diaphragmatic excursion, increasing intrathoracic pressure and impairing ventilation. Ventilatory consequences include increased airway pressures and decreased pulmonary compliance. These effects are accentuated in patients with PHTN and CHD.[17] Additionally, the carbon dioxide insufflation can diffuse into the blood, causing hypercarbia and, as above, increase PVR. Cautious, meticulous management of ventilation and

abdominal CO_2 insufflation with clear communication between the anaesthetist and surgeon is vital, with a low threshold for conversion to open surgery.[18]

ELECTROPHYSIOLOGY

Arrhythmias are a common complication of ACHD, especially in those with ventricular scarring or right-sided dilatation. Patients with pacemakers and/or implanted defibrillators should have these evaluated pre- and postoperatively. The loss of sinus rhythm in ACHD patients may lead to significant haemodynamic decompensation, stasis, and thromboembolism. Antiarrhythmic drugs are often poorly tolerated due to their negative inotropic effects. Ablation results, although improving, carry poorer prognosis than in patients with structurally normal hearts.[4]

Electrical cardioversion is the primary choice of treatment for atrial fibrillation and SVTs.[4] Supraventricular arrhythmias are common in patients who have undergone atrial surgery. Changes in surgical technique, such as the development of the total cavopulmonary connection instead of the classical Fontan, are associated with a reduced incidence.[19] Antiarrhythmic therapy may effectively suppress ventricular arrhythmias but has not been associated with improved survival.[20]

Drugs, including magnesium and lidocaine, should be available should an arrhythmia occur. It is often appropriate to discuss the anti-arrhythmia drugs in advance of the surgery, so as to ensure their availability during the perioperative period.

SPECIAL SITUATIONS AND POPULATIONS

Pulmonary Hypertension

The aim for these patients is to avoid factors that predispose to PHTN and to reverse any reversible factors while also maintaining SVR and preload, where there is a shunt present. Where practical, peripheral regional anaesthetic techniques are preferred. If general anaesthesia is required, controlled ventilation is almost mandatory with adoption of techniques discussed earlier to minimise rises in PVR. Goals of management include avoiding hypercapnia, optimising oxygenation, avoiding acidosis, and minimising positive inspiratory pressure.

Fontan circulation

The physiological implications of a Fontan-type of circulation are extreme.[21] These patients have only 1 ventricle, which pumps blood only to the systemic circulation. Blood return from the systemic venous circulation into the pulmonary circulation is passive, driven only by the difference in venous pressure and atrial pressure (the transpulmonary gradient). Therefore, factors that increase PVR reduce pulmonary blood flow, resulting in increased central venous pressure and inadequate filling of the systemic ventricle, leading to a reduced cardiac output. Reduced pulmonary blood flow also results in hypoxaemia, causing organ dysfunction. Systemic venous hypertension is an inevitable consequence of the absence of a right-sided pumping chamber and is commonly associated with peripheral oedema. The Fontan circulation is also associated with a high incidence of atrial arrhythmias and thromboembolic complications.[22]

Hypovolaemia or any other decrease in SVR is poorly tolerated in these patients. Additionally, any disturbance in respiratory mechanics may lead to adverse effects. Maintaining spontaneous respiration lowers pulmonary arterial pressure and provides a higher cardiac index.[23] For further information on Fontan physiology and considerations, please see ATOTW 511.

Pregnancy

Parturients with ACHD are now reaching reproductive age and requiring anaesthetic management during labour and delivery. Pregnancy induces cardiovascular changes to meet the increased metabolic requirements of both mother and foetus. Some lesions are well tolerated throughout pregnancy; others can decompensate quickly. Cyanotic lesions are associated with more than 50% deterioration, whereas only 15% of acyanotic patients deteriorate. Please refer to ATOTW 111 and 118 for more information.

SUMMARY

Patients with ACHD who present for noncardiac surgery are at increased risk of perioperative morbidity and mortality, even for small uncomplicated surgeries. Anaesthesiologists must understand the specific cardiopulmonary physiology of ACHD and the associated physiological changes induced by anaesthesia as well as other perioperative needs of this population. More complex patients are best served at specialist centres.

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