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Anaesthesia for non-cardiac surgery in patients with adult congenital heart disease

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Learning objectives

By reading this article, you should be able to:

- Recognise the increasing prevalence of adult congenital heart disease (ACHD).
- Classify conditions by anatomical complexity and physiology status for risk assessment.
- Interpret key specialist investigations to inform risk assessment.
- Understand anaesthetic considerations for non-cardiac surgery in this cohort of patients.

Key points

- Patients with adult congenital heart disease have diverse conditions and high perioperative risks associated with non-cardiac surgery.
- Classification by anatomical complexity and physiological status aids in perioperative planning.
- Close coordination with multidisciplinary teams from specialist centres improves safety and reduces complications.
- Tailored anaesthetic strategies and postoperative care are crucial for optimal outcomes.

Congenital heart disease (CHD) is the most common group of congenital anomalies, with a prevalence of approximately nine per 1000 live births. Advances in medical and surgical care have significantly improved survival, with 90% of children born with CHD now surviving to adulthood. There are more adults than children living with CHD, and a significant portion are >60 yrs of age.[1–3] As the age of people living with CHD increases, so does the frequency to which they require care for non-cardiac conditions.

Tracking the prevalence of different cardiac lesions across the lifespan of patients with CHD is challenging. The Quebec CHD database, an internally validated registry, contains 35 yrs of longitudinal data. The most common lesions in the database are valve abnormalities and septal defects. Tetralogy of Fallot (ToF) and transposition of the great arteries (TGA) are examples of more frequently occurring complex lesions.[4]

Adult CHD (ACHD) encompasses a spectrum of illness severity. Patients with mild forms may be asymptomatic and have a similar life expectancy to the general population. Many patients undergo complete repair in childhood and require no further intervention. However, patients with severe forms may require palliative procedures and have significant ongoing morbidity.[3] In contrast to paediatric patients, adults with CHD are more likely to have acquired comorbidities (e.g. chronic kidney disease, cardiac cirrhosis) because of the underlying disease and from previous interventions.

Patients with ACHD undergoing non-cardiac surgery are at increased risk of perioperative complications compared with similar patients without ACHD, particularly when the underlying CHD is complex.[5] Consequently, to minimise the risk associated with non-cardiac surgery, it is important that the anaesthetist understands the anatomy, haemodynamic status, and multisystemic impact of CHD. In this article we review the epidemiology and distribution of ACHD. We highlight

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those lesions associated with increased perioperative risk and propose criteria for when to discuss with, or transfer to, specialist centres. In addition, we summarise the key anaesthetic considerations for common congenital lesions.

Management of children with CHD undergoing non-cardiac surgery is the subject of a recent article in *BJA Education*.[6]

Classification

The American Heart Association (AHA) and American College of Cardiology (ACC) Anatomic and Physiological (AP) classification provides a framework for stratifying ACHD and for guiding treatment, follow-up and perioperative management.[7] When first devised, the classification categorised patients solely on anatomical complexity. The 2018 revised guidelines added physiological status, to account for the real-world impact of the disease and the significant variability in symptoms and function. Although not the intended purpose, the validity of the AP classification as a predictive tool has been assessed in at least one study, with encouraging results.[8]

Anatomical complexity

The AP classification groups lesions by anatomical complexity (Table 1).

Simple lesions

Simple lesions are most commonly small, isolated atrial septal defects (ASDs) or ventricular septal defects (VSDs),

Table 1 Classification of ACHD by anatomical complexity. AVSD, atrioventricular septal defect; HCM, hypertrophic cardiomyopathy. Reproduced with permission from the AHA.

I: Simple
Native disease
Isolated small ASD
Isolated small VSD
Mild isolated pulmonic stenosis
Repaired conditions
Previously ligated or occluded ductus arteriosus
Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement
Repaired VSD without significant residual shunt or chamber enlargement
II: Moderate complexity
Aorto-left ventricular fistula
Anomalous pulmonary venous connection, partial or total
Anomalous coronary artery arising from the pulmonary artery
Anomalous aortic origin of a coronary artery from the opposite sinus
AVSD (partial or complete, including primum ASD)
Congenital aortic valve disease
Congenital mitral valve disease
Coarctation of the aorta
Ebstein anomaly (disease spectrum includes mild, moderate and severe variations)
Infundibular right ventricular outflow obstruction
Ostium primum ASD
Moderate and large unrepaired secundum ASD
Moderate and large persistently patent ductus arteriosus
Pulmonary valve regurgitation (moderate or greater)
Pulmonary valve stenosis (moderate or greater)
Peripheral pulmonary stenosis
Sinus of Valsalva fistula/aneurysm
Sinus venosus defect
Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
Supravalvar aortic stenosis
Straddling atrioventricular valve
Repaired tetralogy of Fallot
VSD with associated abnormality, moderate or greater shunt, or both
III: Great complexity (or complex)
Cyanotic congenital heart defect (unrepaired or palliated, all forms)
Double-outlet ventricle
Fontan procedure
Interrupted aortic arch
Mitral atresia
Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)
Pulmonary atresia (all forms)
TGA (classic or d-TGA; CCTGA or l-TGA)
Truncus arteriosus
Other abnormalities of atrioventricular and ventriculoarterial connection (i.e. crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

including those previously repaired without residual shunt. Most small septal defects that are diagnosed in childhood close spontaneously by adulthood. Patients in this category are typically not under regular review.

Moderate lesions

Moderate lesions include larger septal defects, valve abnormalities, ventricular outflow tract obstruction and repaired ToF. Moderate severity lesions develop haemodynamic consequences over time, typically require lifelong monitoring and may require ongoing intervention.

Complex lesions

Complex lesions include all palliated or unrepaired cyanotic heart disease, single ventricle lesions and other significant malformations such as truncus arteriosus and pulmonary atresia. Complex lesions invariably require multiple surgeries and require lifelong regular monitoring and interventions.

Physiological status

Physiological status is graded A–D, with asymptomatic patients classified as A and those with severe restrictions and life-threatening complications as D (Table 2). Two patients may have the same anatomical classification but be in different physiological grades. For instance, a patient with repaired ToF who is asymptomatic with no complications would be classified as IIA whereas a patient with the same lesion but severe pulmonary regurgitation and exercise intolerance would be classified as IIC.

Risk assessment: (should I be concerned?)

As noted earlier, patients with ACHD undergoing non-cardiac surgery are at increased risk of perioperative morbidity and mortality compared with similar patients without ACHD. Risks are reduced when care is delivered at specialist ACHD centres, particularly for patients with complex lesions.[4,9] Figure 1 details a suggested approach to risk assessment.

Patients with simple complexity or asymptomatic disease (IA) can be safely managed in non-specialist centres. Patients with moderate or greater anatomical complexity or physiological stage ≥B should be discussed early with a specialist centre. The decision to transfer a patient in this grouping depends on their underlying cardiac status, the nature of the required surgery (and its likely cardiovascular impact) and its urgency. Some lesions always confer high perioperative risk (e.g. Eisenmenger syndrome or a failing

Table 2 Classification of ACHD by physiological stage. NYHA, New York Heart Association. Reproduced with permission from the AHA.

A
NYHA FC I symptoms No haemodynamic or anatomic sequelae No arrhythmias Normal exercise capacity Normal renal/hepatic/pulmonary function
B
NYHA FC II symptoms Mild haemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction) Mild valvular disease Trivial or small shunt (not haemodynamically significant) Arrhythmia not requiring treatment Abnormal objective cardiac limitation to exercise
C
NYHA FC III symptoms Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonic, or both) Moderate aortic enlargement Venous or arterial stenosis Mild or moderate hypoxemia/cyanosis Haemodynamically significant shunt Arrhythmias controlled with treatment Pulmonary hypertension (less than severe) End-organ dysfunction responsive to therapy
D
NYHA FC IV symptoms Severe aortic enlargement Arrhythmias refractory to treatment Severe hypoxemia (almost always associated with cyanosis) Severe pulmonary hypertension Eisenmenger syndrome Refractory end-organ dysfunction

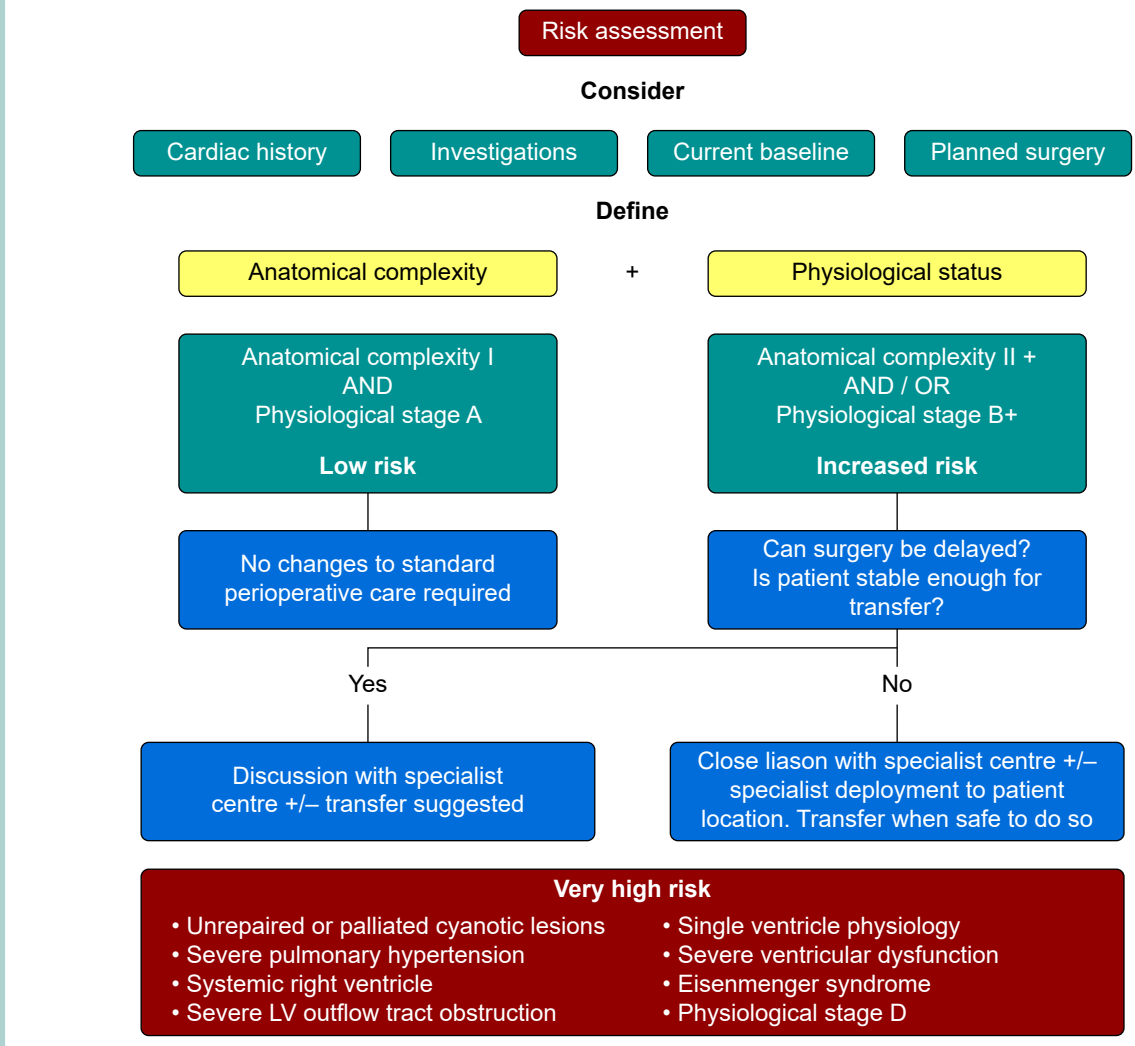

Fig 1 Suggested risk assessment process for patients with adult congenital heart disease presenting for non-cardiac surgery at a non-specialist centre.

Fontan circulation) and should be transferred unless immediate intervention is required. If transfer is not possible, close communication with the specialist centre is essential. Local cardiologists can provide valuable support. Deploying individual specialists (such as an anaesthetist) to the referring centre should be considered. If urgent surgery is required in a high-risk patient, postoperative transfer to a specialist centre may be appropriate.

Telemedicine plays an increasing role in managing patients with ACHD, as it improves access to specialist services in an era of increasing demand. Telemedicine also allows clinicians in the referring centre to connect with members of the multidisciplinary team (MDT) who have immediate access to the patient's records.

Preoperative assessment

In specialist ACHD centres, patients are assessed and optimised by the MDT before elective surgery. In a non-specialist setting, the urgency of the required intervention determines the time that is available for preoperative evaluation and optimisation. If immediate intervention is required, then a focused history and assessment of the patient's functional status may be all that is possible. For patients requiring less urgent intervention, a thorough assessment is appropriate, with the aim of answering the following questions:

- What is the patient's cardiac anatomy and its classification?
- What corrective or palliative procedures have taken place? Are further cardiac procedures planned?
- What is the patient's physiological status?
- What systemic complications are present?
- What are the implications of the proposed non-cardiac surgery on the underlying ACHD diagnosis?

Not all patients have a complete understanding of their underlying condition, and it is common for asymptomatic patients to be lost to follow-up. Where possible, previous medical records should be reviewed.

Investigations

Standard investigations should include a full blood count, along with tests of renal function, electrolytes and coagulation. Increased brain natriuretic peptide is associated with worse perioperative outcomes and may aid in risk assessment.[10] A 12-lead ECG establishes a baseline for later comparison and may identify conduction abnormalities, arrhythmias, or show signs of chamber enlargement. A chest radiograph may identify indicators of severe disease (e.g. cardiomegaly, pulmonary artery enlargement) or show features of heart failure.

Echocardiography

An echocardiogram is vital for understanding the patient's cardiac anatomy and function. Ventricular dysfunction is common, and heart failure remains the leading cause of death in patients with ACHD.[2] Echocardiography is also useful to determine the presence of pulmonary hypertension, which complicates many types of CHD. Features of pulmonary hypertension include a dilated right ventricle, flattening of the interventricular septum (producing a 'D-shaped' left ventricle in short axis), right ventricular (RV) hypertrophy, and enlarged pulmonary arteries. Systolic pulmonary artery pressure can be estimated from the velocity of the tricuspid regurgitation jet, with a regurgitant jet velocity >3 ms^{-1} indicating a systolic pulmonary artery pressure >36 mmHg plus right atrial pressure (i.e. a pulmonary artery systolic pressure >40–45 mmHg). The pulmonary acceleration time is another useful measurement, with a value <105 ms suggesting pulmonary hypertension.[11]

The echocardiogram must be interpreted in the context of the underlying cardiac lesion. If the right ventricle is the systemic pump (e.g. in patients with a Senning or Mustard repair for TGA) then RV failure will have different implications than if the right ventricle is the pulmonary pump. Echocardiograms in patients with complex cardiac anatomy require interpretation by specialists with expertise in CHD.

CT and MRI

Cardiac CTs and MRIs provide detailed anatomical and functional information that may not be available from an echocardiogram, particularly in patients with complex repairs, extracardiac conduits, or vascular anomalies.

Right heart catheterisation

Right heart catheterisation quantifies intracardiac shunt and provides pressure measurements within the right heart. Right heart catheterisation is particularly useful for quantifying the cause and severity of pulmonary hypertension in patients with complex CHD, where derived echocardiographic indices of pressure may be less reliable.

Pulmonary hypertension is confirmed by a mean pulmonary artery pressure ≥20 mmHg. Reporting the pulmonary artery pressure as a ratio of systemic pressure is useful, as it quantifies the pressure burden on the right ventricle and highlights the importance of maintaining the systemic-to-pulmonary pressure gradient (Table 3). The subtype of pulmonary hypertension can be deduced from the pulmonary artery wedge pressure and pulmonary vascular resistance (PVR) (Table 4).

For patients with intracardiac shunts, the ratio of the pulmonary-to-systemic blood flow ($\dot{Q}p:\dot{Q}s$ ratio) is used to assesses severity. $\dot{Q}p:\dot{Q}s$ may be estimated with echocardiography or measured during right heart catheterisation. A $\dot{Q}p:\dot{Q}s$ ratio >1.0 indicates a left-to-right shunt, while a ratio <1.0 indicates a right-to-left shunt. For patients with lesions associated with left-to-right shunting (e.g. a VSD), a $\dot{Q}p:\dot{Q}s$ <1 may indicate the development of Eisenmenger syndrome. Moderate or greater left-to-right shunt causes right heart strain and leads to chronic pulmonary hypertension (Table 5).

Table 3 Pulmonary hypertension severity. PH, pulmonary hypertension; PAP, pulmonary artery pressure.

Severity of PH	Mean pulmonary artery pressure	Relation to systemic BP
Normal	Mean PAP <20 mmHg	Pulmonary artery pressures <1/3 systemic BP
Mild	Mean PAP 21–24 mmHg	Pulmonary artery pressures 1/3–1/2 systemic BP
Moderate	Mean PAP 25–39 mmHg	Pulmonary artery pressures 1/2–2/3 systemic BP
Severe	Mean PAP ≥40 mmHg	Pulmonary artery pressures equal more than systemic BP

Extracardiac manifestations of adult congenital heart disease

Respiratory disease

Respiratory disease is common, with one study demonstrating a restrictive pattern on spirometry in nearly half of patients with ACHD.[12] Patients with ACHD are also at increased risk of pulmonary emboli, pulmonary haemorrhage and pneumonia.[13,14] Patients with more complex disease may have had prolonged periods of tracheal intubation early in life and can develop subglottic stenosis, which may complicate airway management.

Renal dysfunction

The prevalence of renal dysfunction in patients with ACHD is 30–50%.[15] Moderate or severely reduced glomerular filtration rate (GFR) has been associated with a three-fold increase in mortality compared with those with a normal GFR, which most likely reflects the fact that renal dysfunction is a marker of disease severity.[16] Careful monitoring of fluid balance and early institution of renal replacement therapy are appropriate to manage volume overload and acidosis during the perioperative period.

Thrombosis and bleeding

Patients with ACHD are predisposed to coagulation abnormalities. Thrombosis may result from stasis (e.g. attributable to atrial arrhythmias, enlarged cardiac chambers), endothelial disruption (attributable to frequent invasive procedures and the presence of stents and grafts), or haematological abnormalities (e.g. thrombocytopaenia, platelet dysfunction and acquired von Willebrand disease).

Patients with intracardiac shunts who develop venous thromboembolism are at high risk of paradoxical embolism. Patients with chronically increased central venous pressures

Table 4 Categorisation of pulmonary hypertension based on right heart catheterisation data. PH, pulmonary hypertension.

Type of PH	Mean pulmonary artery pressure	Pulmonary artery wedge pressure	Pulmonary vascular resistance	Key characteristics
Normal	<20 mmHg	≤15 mmHg	<2 Wood units	Normal pulmonary haemodynamics
Precapillary PH	≥20 mmHg	≤15 mmHg	>2 Wood units	Pulmonary vascular disease (e.g. pulmonary arterial hypertension, chronic thromboembolic PH, lung diseases)
Postcapillary PH	≥20 mmHg	>15 mmHg	≤2 Wood units	Associated with left heart disease or increased left atrial pressures
Combined pre- and postcapillary PH	≥20 mmHg	>15 mmHg	>2 Wood units	Features of both left heart disease and pulmonary vascular remodelling

Table 5 Left-to-right shunt severity.

$\dot{Q}p:\dot{Q}s$	Shunt severity	Clinical significance
1:1	No shunt	Normal pulmonary—systemic flow
1.0–1.5:1	Mild	Small left-to-right shunt. Often asymptomatic
1.5–2.0:1	Moderate	Significant left-to-right shunt, risk of right heart overload and pulmonary hypertension over time
>2.0:1	Large	Severe left-to-right shunt, high risk of right ventricular failure and pulmonary hypertension

(e.g. as a result of subpulmonic valve stenosis or a Fontan circulation) are at risk of liver congestion, which can progress to 'cardiac' cirrhosis, further contributing to coagulopathy. Polycythaemia is common in in patients with cyanotic heart disease. Consequently, relative anaemia (i.e. a 'normal' haemoglobin concentration) may be missed.

Other preoperative considerations

Arrhythmias

Arrhythmias are common in patients with ACHD and are a frequent cause of death.[2] Arrhythmias can arise as a result of structural abnormalities or occur as a consequence of frequent interventions.

Atrial dilatation predisposes to supraventricular arrhythmias; ventricular dysfunction or previous ventriculotomy predisposes to ventricular arrhythmias. Patients may be taking antiarrhythmic medications. Patients with single ventricle physiology or severe ventricular dysfunction are at particular risk of acutely decompensating if they develop new arrhythmias. Patients with a history of arrythmias or severe ventricular dysfunction should have external defibrillator pads attached before inducing anaesthesia.

Patients with ACHD commonly have implantable cardioverter defibrillators (ICDs) and pacemakers, with a high likelihood of being pacing dependent. Perioperative management should include consultation with a cardiac physiologist to interrogate the device and reprogramme it, if necessary. In emergencies, where monopolar diathermy is necessary, placement of a magnet over the device will set it to asynchronous pacing and disable the defibrillator function.

Perioperative anticoagulation

Because of increased thromboembolic risk, many patients with ACHD require long-term anticoagulation or antiplatelet therapy. The decision to reverse, bridge with i.v. heparin, or continue an oral anticoagulant should ideally involve input from an ACHD cardiologist. The need for anticoagulation will impact upon the decision to use regional anaesthesia.

Endocarditis prophylaxis

Guidance from the European Society of Cardiology and AHA suggests endocarditis prophylaxis should be restricted to those undergoing dental procedures who are at high risk (e.g. patients with prosthetic valves, unrepaired cyanotic defects, residual shunts, or patients who have had endocarditis previously).[7]

Cognitive impairment

Congenital heart disease is associated with developmental delay and cognitive impairment. The cause is multifactorial and includes genetic linkages (e.g. Down syndrome, DiGeorge syndrome), the effects of the cardiac lesion (e.g. cerebral hypoxia, particularly during early brain development in patients with cyanotic heart disease), or complications of previous interventions.

Anxiety and depression

Anxiety and depression are common in patients with ACHD, with an estimated prevalence of up to 60% in younger individuals.[17] Chronic anxiety may stem from the lifelong nature of the condition, ongoing medical surveillance and uncertainty about the future. Anxiety can worsen when treatment occurs away from a familiar centre, particularly in an emergency. Frequent invasive procedures may lead to needle phobia. Whenever feasible, we encourage caregivers to be present until anaesthesia is induced. The anaesthetic team should be mindful of the psychological state of the patient and try and be as a flexible as possible.

Intraoperative considerations

Patients with ACHD present with a broad range of altered physiology and the anaesthesia plan must be tailored to the individual.

Induction and management of anaesthesia

The primary goal during the perioperative period is to preserve physiological stability. Irrespective of the technique chosen, induction of anaesthesia should be done in a slow, controlled

fashion. Most agents used for inducing anaesthesia cause systemic and pulmonary vasodilation and must be titrated carefully. Ventricular dysfunction and shunts will delay the onset of anaesthesia. In patients with significant physiological decline (physiological stage ≥C), poor ventricular function, or Eisenmenger syndrome, preemptive use of vasopressor drugs should be considered during induction of anaesthesia.

Vascular access and monitoring

An arterial catheter for pressure monitoring—placed before anaesthesia is induced—should be considered. As a result of previous procedures, many patients have difficult i.v. access. Chronic occlusion of femoral or neck vessels is common. Monitoring must be planned around the anatomic variations and previous interventions. Coarctation of the aorta with residual obstruction results in an increased arterial pressure in the upper limbs compared with the lower limbs. If coarctation of the aorta was repaired using a subclavian flap, pressure readings from the left arm may be inaccurate. A residual Blalock–Taussig shunt (connection between the subclavian or carotid artery and the pulmonary artery) placed in infancy, can result in falsely low blood pressure readings in the upper limb on the affected side. It may be necessary to measure arterial pressure from more than one location to ensure adequate perfusion. A central venous catheter (CVC) is useful in many patients and may be essential during major surgery. However, a CVC carries a small risk of thrombosis or obstruction of the superior vena cava, which can have devastating consequences in patients with a Glenn shunt, described below.

Regional anaesthesia

Regional anaesthesia can reduce sympathetic stimulation during the intraoperative and postoperative periods. Regional techniques are particularly beneficial for patients with complex ACHD, who are more likely to be opioid-tolerant as a result of frequent previous interventions, and be at high risk of chronic pain. The benefits of regional anaesthesia must be weighed against the risk of causing hypotension and the need for anticoagulation.

Laparoscopic *vs* open surgery

Laparoscopic surgery is associated with reduced postoperative pain, lower wound complication rates and faster recovery compared with open surgery, making it an appealing option in patients with ACHD. However, pneumoperitoneum reduces systemic venous return and increases PVR, which can cause a major decrease in cardiac output in patients with a Fontan circulation, who are dependent on passive pulmonary blood flow (see below). Pneumoperitoneum can also transiently reverse a left-to-right flow in patients with intracardiac shunts, causing hypoxaemia and increasing the risk of systemic embolisation. Close communication between the surgeon and the anaesthetist is essential to minimise the risk of complications. Using low insufflation pressures can help prevent catastrophic hypotension or shunt reversal.

The physiological and anaesthetic implications of individual lesions

Categorising ACHD by anatomical complexity and physiological impact is useful for risk stratification. However, organising lesions by their effects on circulation is more practical when planning and managing anaesthesia care.

Shunt lesions

Atrial septal defects, VSDs and patent ductus arteriosus (PDA) are common causes of left-to-right shunting. Large atrial shunts cause right atrial and RV volume overload, predisposing patients to arrhythmias and eventual RV failure. Ventricular shunts increase pulmonary blood flow and lead to left ventricular volume overload. Both atrial and ventricular shunts can lead to pulmonary hypertension. Chronically increased PVR may result in Eisenmenger syndrome, a late complication marked by shunt reversal (i.e. right-to-left) and resting hypoxaemia. Eisenmenger syndrome is associated with high perioperative risk. Atrioventricular septal defects and partial anomalous pulmonary venous connection are examples of more complex left-to-right shunts.

The primary haemodynamic goal in patients with left-to-right shunts is to avoid increasing shunt flow and the associated decrease in systemic cardiac output. Hyperoxia and hypocarbia should be avoided as both reduce PVR. While vasoconstrictors are useful to treat anaesthetic-related vascular capacitance change, they should be used cautiously in patients with left-to-right shunts to avoid an excessive increase in systemic vascular resistance (SVR), which will increase shunt flow. Most anaesthetic agents reduce SVR more than PVR, and therefore, may transiently improve left-to-right shunting.

For patients with Eisenmenger syndrome, a decrease in SVR and an increase in PVR worsens right-to-left shunting, worsening hypoxaemia. Consequently, rapid treatment of anaesthesia-induced systemic vasodilation with a vasopressor and avoiding factors that increase PVR (e.g. hypercarbia, acidosis, high PEEP) are essential.

Care to avoid i.v. air entrainment is important in all patients with shunt lesions. However, air entrainment is especially dangerous in patients with Eisenmenger syndrome, as right-to-left shunting risks paradoxical gas embolus and the attendant risk of stroke.

Obstructive lesions

Congenital aortic stenosis accounts for 3–6% of all CHD with bicuspid valves being a relatively common indication for aortic valve replacement in middle age.[18] Coarctation of the aorta typically occurs just distal to the left subclavian artery, classically resulting in upper extremity hypertension and potential malperfusion of the abdominal organs and lower extremities. Coarctation of the aorta is associated with aortic root dilatation and bicuspid aortic valve. Treatment involves surgical repair or a transcatheter procedure (stenting or balloon angioplasty). Transcatheter techniques are associated with a high recurrence rate. Mild pulmonic stenosis is considered a simple disease that is unlikely to progress.[19] Moderate or severe pulmonic stenosis may require intervention to prevent the development of RV hypertrophy and heart failure.

Isolated obstructive lesions result in hypertrophy of the associated ventricle, with the potential for ventricular dysfunction and heart failure. Patients with severe obstructive lesions have a fixed stroke volume and tolerate tachycardia poorly. The principles of haemodynamic management are similar to that of obstructive cardiac lesions in non-ACHD patients. Low-normal heart rate should be maintained to ensure an adequate time for ventricular filling, and active

treatment of hypotension with vasopressors to prevent subendocardial ischaemia.

Ebstein's anomaly

Ebstein's anomaly is characterised by dysplasia and apical displacement of the tricuspid valve, with subsequent 'atrialisation' of the right ventricle. Mild forms of Ebstein's anomaly may be asymptomatic. Severe disease results in significant tricuspid regurgitation, right atrial enlargement and RV impairment. If RV function is adequate, the preferred method of surgical repair is the cone procedure, which involves detaching and reimplanting the tricuspid leaflets to form a more functional valve. If RV function is severely impaired or the right ventricle is hypoplastic, a Glenn procedure may be necessary to reduce the demands of the right ventricle. The Glenn procedure involves attaching the superior vena cava directly to the pulmonary artery so that systemic venous return from the upper body flows passively through the lungs. If RV hypoplasia or dysfunction precludes a biventricular circulation, Fontan palliation may be required (see below). After a cone procedure, patients are at risk of atrial arrhythmias and RV dysfunction. Atrial tachyarrhythmias can remain persistent, with chronic atrial fibrillation a poor prognostic sign.[20]

Cyanotic lesions

Tetralogy of Fallot

Tetralogy of Fallot is the commonest cyanotic heart defect. It is characterised by a VSD, overriding of the aorta, RV outflow tract (RVOT) obstruction, and RV hypertrophy. Repair typically takes place within the first 6 months of life and involves closure of the VSD, resection of RVOT muscle bundles and placement of a transannular patch (Fig. 2). After repair, ToF is considered a moderate complexity lesion.

For patients with repaired ToF undergoing non-cardiac surgery, anaesthetists must be aware of potential residual haemodynamic and electrophysiological sequelae. Persistent shunts increase the risk of paradoxical air embolism. Pulmonary regurgitation typically develops over time, particularly if enlargement of the RVOT was required at the time of initial repair. Right ventricular dysfunction is common and there may be persistent RVOT obstruction. Increases in PVR and afterload should be avoided. Ventricular arrhythmias are common, and many patients have an ICD.

Mixing lesions

Mixing lesions are those in which oxygenated pulmonary blood and deoxygenated systemic blood flow is mixed. Examples include TGA, total anomalous pulmonary venous return and truncus arteriosus.

There are two types of TGA: dextro (d-TGA) and levo (l-TGA, now commonly called congenitally corrected TGA; ccTGA). With the more common d-TGA, the aorta arises from the right ventricle and the pulmonary artery from the left, creating two parallel circulations. Oxygenation is dependent on right-to-left flow through a PDA or a VSD. The physiological and anaesthetic implications of d-TGA depend on the type of repair that was performed in infancy. Before the *arterial* switch operation was introduced in the 1980s, d-TGA was treated with an *atrial* switch operation, either the *Mustard* or *Senning* procedure. Atrial switch procedures (Fig. 3) used an atrial baffle to redirect blood flow to the correct ventricle. Over time, patients are prone to arrhythmias and RV failure, as the morphological RV remains the systemic ventricle.[21]

Nowadays patients with d-TGA undergo an arterial switch procedure shortly after birth (or the Rastelli procedure if a VSD is present). The arterial switch operation involves connecting the aorta and pulmonary artery to the left and right ventricles, respectively, and reimplanting the coronary arteries into the aortic root. Long-term complications after the arterial switch operation include stenosis of the re-implanted coronary arteries, neoaortic root dilatation and pulmonary artery stenosis.

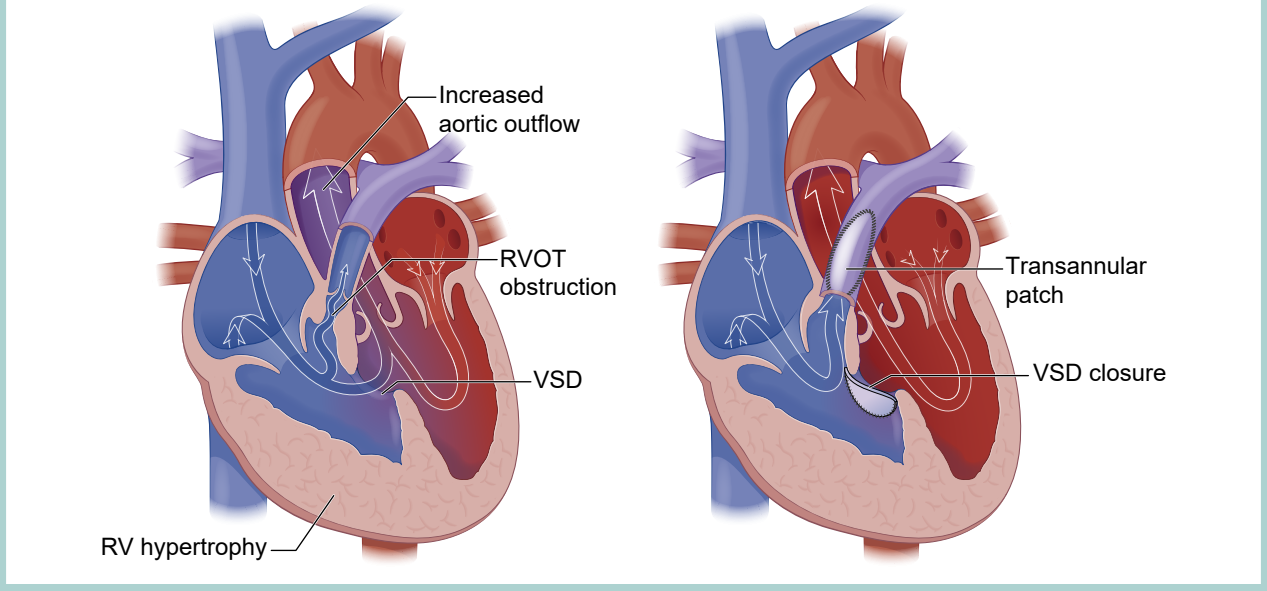

Fig 2 Tetralogy of Fallot, before and after repair.

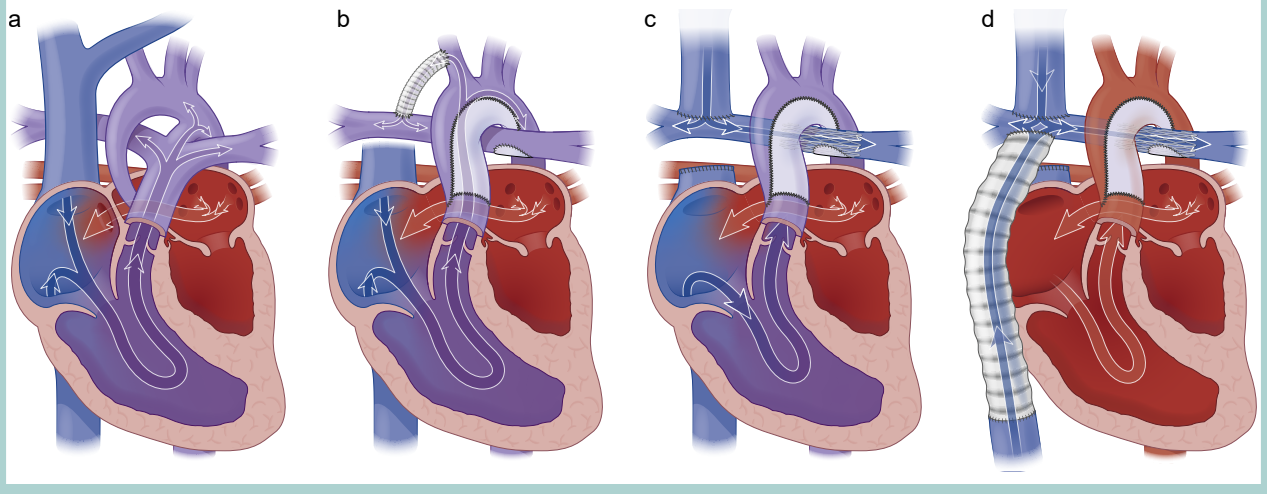

Fig 3 Stages of Fontan palliation for hypoplastic left heart syndrome. (A) Hypoplastic left heart syndrome. (B) Stage I repair, the Norwood procedure. The aorta is reconstructed using the main pulmonary artery and graft material. A modified Blalock–Taussig (BT) shunt is placed, connecting the subclavian artery to the pulmonary artery. An atrial septectomy is performed to allow mixing of oxygenated and deoxygenated blood. (C) Stage 2, the Glenn procedure. The superior vena cava is disconnected and anastomosed to the pulmonary artery and the BT shunt is ligated. (D) Stage 3, the Fontan procedure. The inferior vena cava is disconnected and anastomosed to the pulmonary artery.

Congenitally corrected TGA, is characterised by both atrioventricular and ventriculoarterial discordance, resulting in a physiologically 'normal' circulation with the left- and right-sided circulations in series (in contrast to being in parallel with uncorrected d-TGA) but with a systemic right ventricle. Right ventricular failure and progressive valvular dysfunction are common in adulthood.

Single ventricle physiology

A functionally univentricular heart that is not amenable to biventricular repair may be palliated with a Fontan type procedure by creating a total cavopulmonary connection, whereby all of the systemic venous return passes passively to the pulmonary artery. Originally performed for tricuspid atresia, the most common indication for a Fontan procedure is

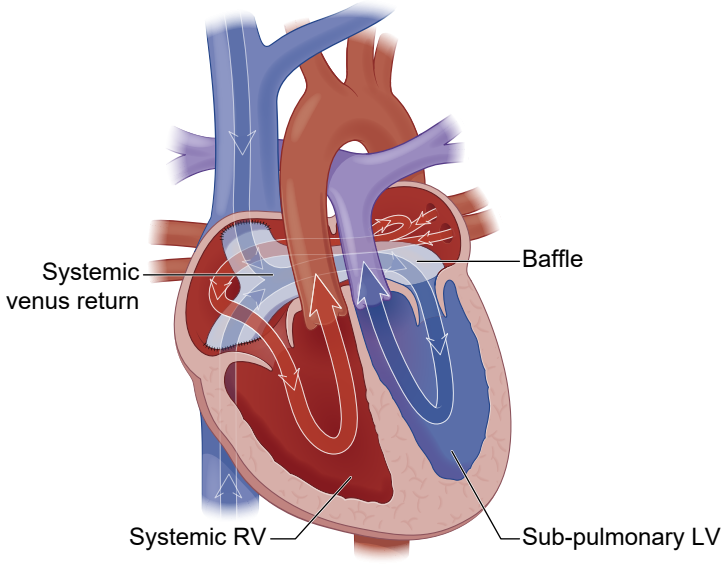

Fig 4 Atrial baffle procedures for treating d-transposition of the great arteries. In the Mustard procedure, synthetic material was used to create the baffle, whereas the in the Senning procedure, the patient's own tissue was used to create the baffle.

now hypoplastic left heart syndrome. A Fontan procedure is usually the final stage of a three-stage repair. Patients have typically already undergone a Glenn procedure, described above. The Fontan procedure involves directing blood from the inferior vena cava directly to the pulmonary artery via a conduit (Fig. 4).

The long-term survival after Fontan palliation has improved over time, being ~80% at 20 yrs, but the associated morbidity is high.[22] Progressive systolic dysfunction of the single ventricle leads to ventricular dilatation, valvular regurgitation and heart failure. The majority of patients experience arrhythmias and are at high risk of thromboembolic events, exacerbated in the perioperative period by reduced mobility, dehydration and interruption of anticoagulation.[23]

The haemodynamic goals in patients with Fontan physiology are focused on maintaining forward flow through the passive pulmonary circulation and supporting the single ventricle. Pulmonary blood return is the central determinant of cardiac output, which itself is dependent on the transpulmonary gradient (i.e. the difference in pressure between the pulmonary artery and the common atrium). The single ventricle cannot compensate for restrictions in flow through the pulmonary bed, and inotropes make little difference to cardiac output when preload is low or PVR is high. Hypoxaemia, hypercarbia and acidosis increase PVR and, therefore, reduce cardiac output. If a CVC is used, the pressure displayed will represent the Fontan circuit (i.e. pulmonary artery) pressure. Fontan circuit pressure should be kept as close to baseline as possible, typically between 10 and 15 mmHg. The arterial oxygen saturation should also be maintained close to baseline, which is usually slightly lower (90–95%) than normal. This can be caused by right-to-left shunting from Fontan baffle leaks, intrapulmonary shunt or venovenous collaterals. A Fontan circulation is particularly sensitive to shortened diastole (adequate filling time is required from passive pulmonary flow) and thus tachycardia can reduce cardiac output. Loss of atrioventricular synchrony increases the transpulmonary gradient, further decreasing preload and cardiac output.

Pulmonary hypertension

Pulmonary hypertension, present in 3–10% of patients with ACHD, is associated with increased perioperative morbidity and mortality.[24,25] The mechanism of pulmonary hypertension is often multifactorial. The most common cause is attributable to shunt-related excess pulmonary blood flow. Another common cause is left-sided heart disease (e.g. valve disease, ventricular dysfunction). Chronic pulmonary hypertension can lead to RV failure. While not caused by pulmonary hypertension *per se*, pulmonary stenosis leads to similar consequences. For single ventricle circulations, ventricular dysfunction and maladaptive pulmonary vascular remodelling can lead to pulmonary hypertension, low cardiac output and heart failure.

As noted above, acidosis, hypercarbia, hypoxaemia, and high PEEP, all worsen pulmonary hypertension. Ensuring patients receive adequate analgesia helps avoid increases in PVR. In patients with RV dysfunction, i.v. fluids should be given cautiously to avoid precipitating RV failure.

Postoperative considerations

All patients with moderate or greater anatomical complexity or physiological stage ≥B should be admitted to a high-dependency unit or ICU for at least 48 h after surgery, particularly if haemodynamic stability or arrhythmias are a concern. Patients with RV dysfunction or a Fontan circulation should be extubated as early as feasible, to avoid the adverse effects of mechanical ventilation, but not in the presence of acidosis and hypercarbia or when pain is not adequately controlled.

Conclusions

Safe perioperative management of adults with CHD who present for non-cardiac surgery demands the anaesthetist has a comprehensive understanding of the patient's anatomy, prior interventions, illness trajectory and medications. It is important that the anaesthetist appreciates the particular risks and haemodynamic goals for individual cardiac lesions. Given the variability and potential complexity of ACHD, a multidisciplinary approach is necessary with close collaboration with specialist centres. When transfer to a specialist centre is not feasible, the anaesthetist should prioritise maintaining haemodynamic stability during and after the non-cardiac procedure. After the procedure, further discussion re management or transfer to a specialist centre may be appropriate.

Declaration of interests

JW has patent #Circulation.2019;139:e698-e800 issued to the American Heart Association. Both authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

MCQs statement

The associated MCQs (to support CME/CPD activity) will be accessible at www.bjaed.org/cme/home by subscribers to *BJA Education*.

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