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Anesthetic Management in Adults with Congenital Heart Disease

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Abstract

Purpose of Review Adults with congenital heart disease (ACHD) are a complex and growing population that presents numerous challenges for anesthetic management. This review summarizes special considerations for anesthetic management in ACHD.

Recent Findings The adult patient with congenital heart disease may require anesthetic care for multiple surgeries and interventions throughout their lifetime. The cardiac and extracardiac manifestations of ACHD have important perioperative implications that affect anesthetic management. Recent American Heart Association/American College of Cardiology and European Society of Cardiology guidelines endorse a multidisciplinary, team-based approach to care. The cardiac anesthesiologist, endorsed as part of this multidisciplinary team, must have a thorough understanding of congenital heart disease pathophysiology and common extra-cardiac manifestations of ACHD.

Summary Safe anesthetic management in adult congenital heart disease should incorporate a multi-disciplinary approach to patient care. Anesthesiologists and centers with special expertise in ACHD care should be utilized or consulted whenever possible.

Keywords Congenital heart disease · Cardiac anesthesia · ACHD · Extra-cardiac manifestations of ACHD

Introduction

Moderate to severe congenital heart disease (CHD) occurs in approximately 1% of live births [1, 2]. Improvements in pediatric cardiology, congenital cardiac surgery, and perioperative intensive and anesthetic care have led to increased longevity in the CHD population, with most patients now living well into adulthood, and it is estimated that the number of adults with congenital heart disease (ACHD) exceeds the pediatric CHD population [2]. The heterogeneity of cardiac defects found in congenital heart disease is significant, and while some congenital cardiac malformations can be surgically corrected during childhood, many cardiac lesions can only be palliated. Thus, adult patients with congenital heart disease may have numerous surgical reoperations, percutaneous interventions, and electrophysiologic procedures related to their respective cardiac lesion. They also frequently present to the operating room for non-cardiac surgery. Heart failure, pulmonary hypertension, and arrhythmias are common sequelae in the ACHD population, but these patients also have other significant extracardiac comorbidities and associated syndromes that warrant special consideration and increase perioperative risk [3, 4]. In addition, adults with congenital heart disease are often lost to follow-up during transition of care from pediatric to adult cardiology specialists, sometimes leaving their burden of cardiac and comorbid disease unmanaged for several years [5, 6]. Given the growing population of adult patients with congenital heart disease and a limited number of specialized ACHD centers and providers [7], physicians and health care providers in non-specialized ACHD centers may encounter these patients in their daily practice. This review will specifically focus on the anesthetic management of ACHD patients.

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Nomenclature and Surgical Risk Stratification

Clinical tools exist to help assess surgical risk and to risk stratify patients for congenital heart surgery, such as the Society of Thoracic Surgeons (STS)-European Association for Cardio-Thoracic Surgery (EACTS) Congenital Heart Surgery Mortality Score and Categories (STAT Mortality Score and Categories) [8]. However, STAT Categories only take into account surgical complexity, and ACHD patients exhibit significant extracardiac comorbidities in addition to the aging process. The 2018 ACC/AHA guidelines introduced a new classification system for defining the severity of ACHD, the ACHD Anatomy and Physiological Classification (ACHD AP) [9••], which considers the patient's native anatomy, surgical repair, and current physiology. The previous guidelines had defined disease severity primarily by anatomy. The ACHD AP was not intended to be used as a surgical risk or mortality score, but at least one center has demonstrated its ability to predict early mortality risk, similar to existing methods [10]. As utilization of the ACHD AP classification system becomes more widespread, this nomenclature may help to standardize and improve perioperative communication among multidisciplinary ACHD care teams.

Extracardiac Manifestations of Congenital Heart Disease and Anesthetic Implications

Patients with ACHD typically have cardiac and non-cardiac sequelae [11], as well as a higher risk of certain types of cancers [12] and metabolic syndromes [13]. Preoperative assessment of ACHD patients requires an individualized approach because of the heterogenous nature of this patient population (see Fig. 1). Baehner and colleagues describe a four-pronged approach to preoperative evaluation in ACHD [14]:

1. Knowledge of the underlying anatomic defect in each individual patient
2. How the patient has been managed, whether surgically corrected or palliated, and what is the expected clinical course of the patient, especially if they have moderate or complex CHD
3. Awareness of the long-term cardiac and non-cardiac consequences of ACHD which could complicate the anesthetic management
4. The type of surgical procedure being performed and the inherent risks of that procedure in addition to the risks conferred by the underlying ACHD sequelae

Heart Failure

Heart failure is a common concern in ACHD patients and can involve either the left or right ventricles or both. It is more common in patients with complex CHD compared to simple lesions. Heart failure symptoms are present in 32% of patients with L-TGA, 22% of patients with D-TGA after an atrial switch procedure, and 40% of single-ventricle patients with Fontan palliation [15]. Heart failure is difficult to diagnose in these patients, as they may have shortness of breath and reduced exercise tolerance simply due to their CHD. Conversely, patients may be well-compensated, but may still have underlying ventricular dysfunction. An analysis of cardiac arrests in children with pediatric heart disease showed that patients with heart failure were more likely to arrest from cardiovascular causes than those without heart failure. The most common underlying lesion in those who arrested was single-ventricle physiology, whereas the highest mortality was seen in patients with aortic stenosis and cardiomyopathy [16]. It is imperative to document the patient's ventricular function and exercise capacity preoperatively. An echocardiogram should be obtained and reviewed, as well as measurements of biomarkers such as N-terminal pro brain natriuretic peptide (NT-Pro-BNP), which has a strong correlation with cardiovascular events in patients with heart failure [17, 18]. Advanced imaging and a discussion with the congenital cardiology team may be required. In some instances, identification of the etiology of heart failure and its correction is warranted before elective non-cardiac surgeries. An example would be pulmonary valve replacement in a tetralogy of Fallot (TOF) patient with severe pulmonary regurgitation and consequent right ventricular (RV) dysfunction. In more complex lesions that have been palliated, such as d-transposition treated with an atrial switch procedure, there may be baffle obstruction or dysrhythmias that need to be addressed with interventional or electrophysiology procedures [19].

Heart failure can complicate induction and maintenance of anesthesia. In patients with significant ventricular dysfunction, an arterial catheter should be placed preoperatively. Induction agents such as propofol can reduce systemic vascular resistance and therefore, alter preload, which can then reduce cardiac output and result in hypotension. Patients may require a vasopressor, such as phenylephrine or vasopressin, as a bolus or infusion. If hypotension does not resolve and low cardiac output is suspected, there should be a low threshold to initiate an inotrope infusion such as epinephrine. With increased competency in point-of-care ultrasound (POCUS) among anesthesiologists, it may be appropriate to perform a quick intraoperative assessment of cardiac function for

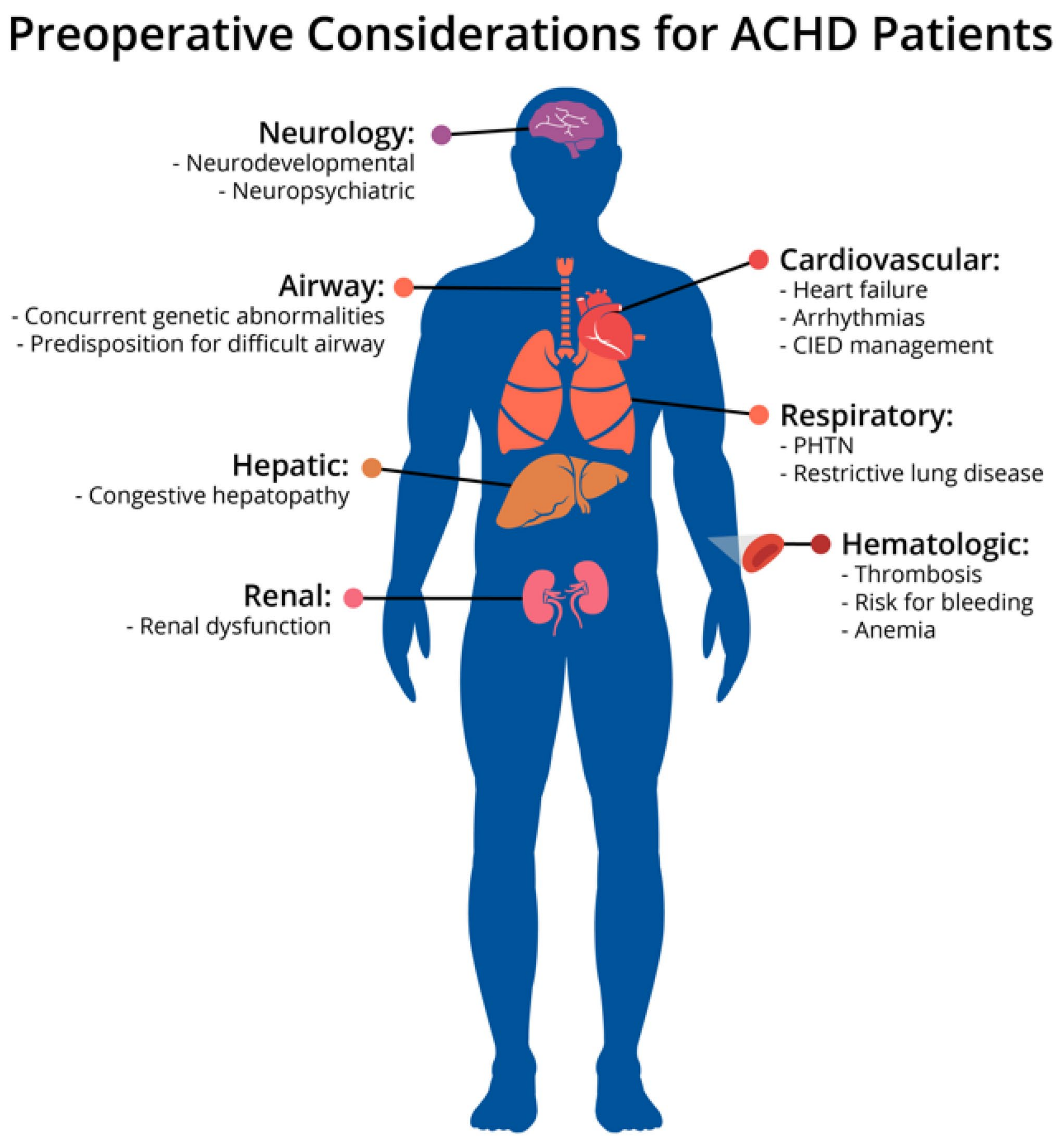

Fig. 1 General preoperative considerations for the ACHD (adult congenital heart disease) patient. (PHTN = pulmonary hypertension, CIED = Cardiovascular implantable electronic device)

refractory hypotension with a low threshold to call for a transesophageal echocardiogram (TEE) by a cardiac anesthesiologist. It is also important to manage intraoperative administration of crystalloids and blood products, since these patients are at a high risk of decompensation and volume overload.

Arrhythmias

Arrhythmias are common in patients with CHD and are prevalent in adults with late intracardiac repairs, palliated or unrepaired lesions [20]. They can range from asymptomatic to hemodynamically significant and poorly tolerated, especially in the intraoperative period. Supraventricular arrhythmias are most common in late ASD closures or patients with atrial switch procedures and Fontan operations, where up to 50% of episodes can be symptomatic [21, 22]. Intra-atrial reentrant tachycardias are the most common etiology of supraventricular tachycardia (SVT) in late ASD closures [23]. Patients may be managed with Class I or Class III antiarrhythmic drugs, atrioventricular nodal blocking agents, antiplatelet agents, and/or anticoagulants. The latter medications have implications for bleeding and risk of thrombosis in the perioperative period. Ventricular arrhythmias are common in those who have had a ventriculotomy, such as TOF patients, or in those patients with reduced LV or RV function [24]. Myocardial fibrosis and scarring from prior ventricular incisions can impair conduction and lead to a reentry circuit that can result in ventricular tachycardia. These patients require implantable cardioverter defibrillator (ICD) placement. Alternatively, patients may require permanent pacemaking for bradyarrhythmia, which can occur due to sinoatrial (SA) or atrioventricular (AV) node injury or ischemia.

Arrhythmias can complicate the perioperative course, especially if they result in hemodynamic instability. It is important to have quick access to emergency medications and antiarrhythmics such as lidocaine and amiodarone, and defibrillator pads should be placed on the patient with the defibrillator system located inside or just outside the operating room for easy access. Preoperative team communication should identify one individual in the operating room responsible for managing the defibrillator at the direction of the anesthesiologist for cardioversion, defibrillation, or pacing during a poorly tolerated arrhythmia. In patients with a cardiac implantable electronic device (CIED), device management should be in accordance with the ASA practice advisory on CIEDs [25]. Important individual details include the brand and type of device the patient has, whether they are pacemaker dependent and the device settings. If electrocautery interference is expected above the umbilicus, the pacing function should be switched to an asynchronous mode and the ICD tachycardia function deactivated. All devices should be interrogated postoperatively to restore baseline settings and ensure proper function.

Pulmonary Hypertension

Pulmonary hypertension (PH) is common in ACHD, likely due to long-standing intracardiac defects, with a prevalence of approximately 10%. It is associated with perioperative morbidity and death. Data from the pediatric literature suggests that the incidence of perioperative cardiac arrest ranges from 0 to 5%, and perioperative death occurs in up to 1.5% [16, 26]. PH can progress to Eisenmenger syndrome, which can limit operability of the intracardiac defect [27]. Eisenmenger syndrome also increases an individual's risk of premature death [28]. Anesthetic management begins with preoperative assessment of severity of PH and the medications being used to manage it. Patients may be managed on phosphodiesterase type-5 (PDE5) inhibitors (sildenafil, tadalafil), endothelin receptor antagonists (bosentan, ambrisentan), prostacyclin analogs (epoprostenol, treprostinil, iloprost), or selective prostacyclin agonists (selexipag) [29], and these medications may result in hypotension requiring vasopressors.

Acute hypoxia, hypercarbia, hypothermia, acidosis, and sympathetic stimulation due to pain may cause a sudden increase in pulmonary vascular resistance (PVR) leading to acute RV failure. Principles of anesthetic management include preventing acute increases in PVR while maintaining systemic vascular resistance (SVR). In patients with PH, it is advisable to place a pre-induction arterial line to monitor hemodynamics carefully. Patients are preoxygenated with a FiO_2 of 1.0, respiratory depressants such as opiates are avoided, and a quick intubation is performed. There should be a low threshold to initiate inotropic infusions such as epinephrine (0.02–0.04 mcg/kg/min) prior to induction in cases of significant RV dysfunction. Nitric oxide (20 parts per million) and inhaled epoprostenol (50 ng/kg/min) can be administered via the endotracheal tube or high flow nasal cannula and can promote pulmonary vasodilation intra- and postoperatively [30]. Regional anesthesia is preferred if possible. All patients should be monitored in an intensive care unit postoperatively [31].

Bleeding and Thrombosis

Adults with CHD are prone to coagulation abnormalities, which predispose them to both thrombosis and bleeding. Patients may present with thrombocytopenia and platelet dysfunction, as well as increased peripheral consumption. One hypothesis is that megakaryocytes are introduced to the systemic circulation in patients with right to left shunts bypassing the pulmonary circulation and therefore not fragmenting into platelets [32]. Additionally, acquired von Willebrand syndrome is present in many ACHD patients, especially those with complex CHD, advanced heart failure, and Eisenmenger syndrome [33]. A large subset of the patients may also have preoperative anemia, especially those with cyanotic CHD. Anemia is associated with adverse outcomes including a threefold increased risk of death and warrants aggressive preoperative treatment [34]. It is important for the anesthesiologist to be aware of these defects since intraoperative bleeding in these patients often requires component therapy other than red blood cells. Viscoelastic testing is ideally obtained during bleeding to determine the need for coagulation factors, cryoprecipitate, platelets, and fresh frozen plasma. Desmopressin (DDAVP) may be a useful adjunct to decrease transfusion requirements in cases of platelet dysfunction [35]. Many institutions use an algorithmic approach to bleeding in this patient population during cardiac and non-cardiac surgery [36].

Hypercoagulability is an under-recognized complication of CHD. Adults with CHD have a tenfold higher rate of stroke than matched controls [37], and thromboembolic complications can result in increased mortality. Patients with more complex CHD have the highest risk of thromboembolic events, especially those with transposition of great arteries (TGA), single-ventricle physiology, and cyanotic CHD [38, 39]. Irrespective of the complexity of CHD, the occurrence of atrial arrhythmias predisposes ACHD patients to thromboembolic events, and atrial arrhythmias are three times more prevalent in ACHD [40]. Other important risk factors include diabetes mellitus, hypertension, and recent myocardial infarction. Patients with a Fontan circulation are especially prone to thromboembolic complications, with an estimated prevalence of 10–25% [41, 42]. Their thromboembolic predisposition is multifactorial: atrial arrhythmias are highly prevalent [43], and static blood flow is common, particularly in those Fontans with an atriopulmonary connection [44].

Cyanotic CHD patients have a predisposition for secondary erythrocytosis due to chronic hypoxemia. Hypoxemia stimulates overproduction of erythropoietin, leading to increased red cell mass. The resultant increase in blood viscosity can predispose to tissue ischemia and infarction. Patients may present with symptoms of hyperviscosity syndrome, such as irritability, myalgias, and headaches when the hematocrit exceeds 65% [45]. Prolonged overnight fasting, which is common in the perioperative period, can exacerbate these symptoms. Patients should be questioned about these non-specific symptoms during preoperative workup, and coagulation studies should be performed. Patients may need to be admitted overnight prior to surgery for intravenous hydration. Overall, perioperative strategies to reduce blood loss include preoperative coagulation testing, intraoperative utilization of viscoelastic testing, administration of antifibrinolytics, and the use of heparin management systems during cardiopulmonary bypass.

Renal Dysfunction

Renal dysfunction in ACHD patients is present in both simple and complex lesions. Complex CHD patients have an almost 35-fold higher prevalence of renal dysfunction compared to the general population, and patients with Eisenmenger syndrome have the lowest glomerular filtration rates (GFRs) [46]. The implications of renal dysfunction are severe: those with moderate to severe reductions in GFR have a threefold increase in mortality compared to those with normal GFR [46]. The etiology is multifactorial: polycythemia, chronic hypoxemia, exposure to nephrotoxins, and cardiopulmonary bypass have all been implicated [46, 47]. In a recent study of ACHD patients who were predominantly admitted to a cardiac ICU after cardiac surgery, 45% had acute kidney injury (AKI) and of those, 33.7% had persistent renal dysfunction, and 23.6% had AKI at the time of hospital discharge. Both diagnoses were independently associated with mortality at 5 years, and persistent renal dysfunction was associated with a prolonged hospital length of stay [48].

Preoperative identification of renal impairment is helpful for the intraoperative and postoperative care of these patients. Goals of anesthetic management should be to maintain perfusion pressure and cardiac output, as well as to optimize preload. In the postoperative period, these patients may warrant diuresis or early initiation of continuous venovenous hemofiltration (CVVH) to manage volume overload and to improve venous congestion after cardiac surgery complicated by RV dysfunction [49].

Anesthetic Management

The ACHD population is heterogeneous, and patients with similar congenital heart defects can present to the operating room with very different cardiac pathophysiology and extracardiac comorbidities. A multidisciplinary approach should involve close perioperative communication between the cardiologist, surgeon, intensivist, and anesthesiologist. No single anesthetic technique is appropriate for every patient, every CHD lesion, or every surgical procedure. Patients with simple lesions, even corrected ventricular septal defects and atrial septal defects, may have increased risk of developing pulmonary hypertension throughout life [50], and simple defects also appear to be associated with increased morbidity [51]. Thus, it is important for the anesthesiologist to have a thorough understanding of the patient's lesion, previous surgical repairs or interventions, any comorbid conditions, and how anesthetic management and any surgery or intervention could impact the patient's pathophysiology. Preoperative echocardiography, catheterization data, and other pertinent imaging, laboratory tests, and available hemodynamic data should also be reviewed. A thorough airway examination is critical, as patients with congenital heart disease and especially those with coexisting genetic syndromes may have airway abnormalities that could make laryngoscopy or placement of an endotracheal tube difficult [52, 53].

Prior to induction, American Society of Anesthesiologists (ASA) standard monitors are placed. External pacing/defibrillator/cardioversion pads are placed due to higher risk of arrhythmias, which can lead to hemodynamic instability, especially during anesthesia induction and sternotomy. A low dose of anxiolytic medication, typically midazolam, may be utilized in selected patients preoperatively, but it should be noted that use of midazolam is associated with post-operative delirium, particularly in geriatric patients [54]. For vascular access, large bore intravenous (IV) catheters (size 18 gauge or greater) are placed, and arterial line monitoring and central venous access are preferred for major cardiac surgery, with the arterial line placed prior to induction of anesthesia to quickly identify and treat hemodynamic instability. The anesthesiologist must be prepared for difficult central and peripheral intravenous catheter placement, as the ACHD patient may have scarring or thrombosis from previous cannulation sites, and the patient's anatomy may be limited by previous shunts, repairs, or arteriovenous fistulas for hemodialysis access. In most cases requiring general anesthesia, induction of anesthesia can be performed safely with an opioid such as fentanyl, an anesthetic such as propofol or etomidate, and a neuromuscular blocking agent. Propofol must be used cautiously, as it can significantly decrease SVR, leading to hypotension and hemodynamic instability. In especially tenuous ACHD patients with ventricular dysfunction or Eisenmenger syndrome, empiric use of vasopressor or inotrope infusions may lead to a more stable induction [55]. Maintenance of anesthesia is typically accomplished with a balanced approach that includes opioids, inhaled volatile anesthetics such as sevoflurane or isoflurane, and neuromuscular blocking agents. Generally, for ACHD patients with shunt-dependent physiology, anesthetic and hemodynamic management goals should be directed at maintaining the patient's baseline hemodynamics, peripheral oxygen saturation (SpO_2), heart rate, and cardiac output, while minimizing left to right shunting and avoiding right to left shunting. The degree of shunting or pulmonary blood flow to systemic blood flow ratio (Qp:Qs) is often a primary determinant of intervention, with Qp:Qs > 1.5 being abnormally elevated. Anesthetic and hemodynamic management goals for some common ACHD lesions are described in Table 1.

The anesthesiologist must be adept at manipulation of PVR and SVR. The SVR can be increased by pain, surgical stimulus, inadequate or light anesthesia, and administration of vasopressor medications, while PVR can be increased by acidosis, hypoxia, hypercarbia, hypothermia, and use of positive end expiratory pressure ventilation (PEEP). Notably, the use of PEEP can also cause a decrease in PVR after lung recruitment for atelectasis. During anesthesia induction, it is customary to preoxygenate with high inspired concentration of oxygen (FiO_2), but the anesthesiologist must be aware that in certain patients, this can lead to pulmonary vasodilation, pulmonary overcirculation, decreased cardiac output, and hemodynamic instability, a potentially lethal cycle if not quickly acknowledged and treated. After induction, it is appropriate to use the lowest FiO_2 possible to maintain the patient's baseline SpO_2. Intraoperative TEE is performed after induction of anesthesia and post-repair during cardiac surgery, and it is not uncommon to discover additional lesions intraoperatively or residual defects post-repair [56]. Identification and communication of these findings to the operative team is critical. Intravenous inotropes are commonly used to treat ventricular dysfunction in the post-cardiopulmonary bypass period, and milrinone or inhaled pulmonary vasodilators may be beneficial in the setting of RV dysfunction with pulmonary hypertension.

Other anesthetic considerations include surgical bleeding risk and blood product management, as the ACHD patient can have increased bleeding and transfusion requirements from previous sternotomies, preoperative anemia, anticoagulation therapy, hepatic dysfunction, thrombocytopenia, and platelet dysfunction. Intraoperative antibiotic selection is also important, as patients with ACHD are at increased risk for developing infective endocarditis and have high morbidity and mortality when this occurs [57].

Postoperative Considerations: Early Extubation

Patients with CHD present unique challenges in the early postoperative period. There is a continued emphasis to extubate cardiac surgical patients early in the intensive care unit (fast-track extubation) or even in the operating room (ultra-fast-track extubation). Patients who have had extra-cardiac total cavopulmonary connections are uniquely sensitive to the effects of mechanical ventilation. Positive pressure ventilation can alter intrathoracic pressure and reduce venous return and cause hypotension and hemodynamic instability. Therefore, early extubation of Fontan patients in the OR or ICU is a well-accepted strategy. Ono and colleagues compared early extubation (median time 4 h) even in unstable patients, with a slower extubation process (median time 16 h) in 458 patients undergoing extracardiac total cavopulmonary connection. They found significant improvement in hemodynamics after early extubation, both in patients who were stable and unstable [58].

Cardiopulmonary mechanics are also important in adults with CHD following cardiac surgery. In general, the goal should be to achieve early extubation in these patients. Some patients may have impaired RV function, which may be sensitive to the impact of excessive PEEP and positive intrathoracic pressure, which could reduce preload and result in decreased LV filling and cardiac output. If extubation is premature, especially in obese individuals or those with obstructive sleep apnea, hypercarbia can occur and precipitate RV failure leading to hypotension and hemodynamic

Table 1 Pathophysiologic concerns, anesthetic considerations, and hemodynamic management goals for congenital heart defects

CHD lesion	Pathophysiologic concerns	Anesthetic considerations	Hemodynamic goals
Simple Shunts (ASD/VSD/PDA)	Predominantly left-to-right shunts	De-airing of lines and use of air filters	Maintain PVR to avoid pulmonary overcirculation
	Arrhythmias, atrial fibrillation, and heart block	External pacing/cardioversion/defibrillator pads prior to induction	Maintain HR, contractility, preload to maintain CO
	Pulmonary hypertension and Eisenmenger syndrome (rare)	High FiO_2 may cause pulmonary overcirculation	Avoid increase in PVR:SVR ratio causing right to left shunt and hypoxia
	Paradoxical embolus can occur in right-to-left shunting	Inhaled pulmonary vasodilators or milrinone available for PH and RV dysfunction	
		Early extubation in OR (ultra-fast-track) or ICU (fast-track)	
Anomalous coronary arteries	Majority of patients asymptomatic	Monitor for coronary ischemia with TEE and EKG	Maintain coronary perfusion pressure
	Can present as sudden cardiac death during exercise	Use caution with volatile anesthetics and propofol which decrease SVR and CPP	Maintain normovolemia, preload, afterload
	Heterogenous coronary artery anatomy	External pacing/cardioversion/defibrillator pads prior to induction	Avoid increases in contractility and tachycardia
Tetralogy of Fallot	Rarely present for primary repair as adult, often present for PV intervention	Inhaled pulmonary vasodilators and/or milrinone available for PH and RV dysfunction	Avoid increases in PVR and decreases in SVR
	Nature of repair and residual defects, presence of AP collaterals	Use caution with fluid administration in RV failure	Maintain atrioventricular synchrony (NSR)
	RVOTO, pulmonary valve stenosis, regurgitation, or both	External pacing/cardioversion/defibrillator pads prior to induction	Maintain RV contractility
	Arrhythmias, heart block, and sudden cardiac death	Minimal PEEP and optimally spontaneous ventilation	
	Right ventricular dysfunction common		
	Trisomy 21 and other congenital abnormalities common		
Transposition of great arteries	D-TGA vs L-TGA, atrial (Mustard or Senning) vs arterial switch	Monitor for coronary ischemia with TEE and EKG	Maintain coronary perfusion pressure
	L-TGA (ccTGA) may present as adult in heart failure	Inhaled pulmonary vasodilators or milrinone available for PH and RV dysfunction	Maintain preload, atrioventricular synchrony (NSR), and low PVR
	Morphologic right (systemic) ventricular failure in D-TGA	External pacing/cardioversion/defibrillator pads prior to induction	Maintain contractility
	Coronary artery perfusion defects common	Anatomy and previous repairs can impact central venous cannulation	
	Arrhythmias, heart block, and sudden death. Often have PPM/ICD		
	Associated cardiac defects (VSDs common)		
Single ventricle (Fontan/Glenn)	Single ventricle morphology and stage of repair (Can present as Fontan or Glenn)	Perioperative fasting (NPO times) decreases preload. Schedule first case if possible	Maintain preload, atrioventricular synchrony (NSR), and low PVR

Table 1 (continued)

CHD lesion	Pathophysiologic concerns	Anesthetic considerations	Hemodynamic goals
	Type of Fontan repair (extracardiac, lateral tunnel, fenestrated, unfenestrated)	Previous repairs affect vascular access and BP monitoring	Minimize positive pressure ventilation and intrathoracic pressure
	Cardiac output dependent on passive pulmonary blood flow	Positive pressure ventilation and laparoscopy decrease venous return	Ventilation targets low PIP, low RR, minimal PEEP, higher tidal volume
	Hepatic dysfunction, renal failure, malnutrition (PLE), plastic bronchitis	Inhaled pulmonary vasodilators available for PH and RV dysfunction	
	Polycythemic and prothrombotic state	Early extubation in OR (ultra-fast-track) or ICU (fast-track)	
	Arrhythmias, heart block, and sudden death. Often have PPM/ICD	External pacing/cardioversion/defibrillator pads prior to induction	
HOCM	Associated with syncope, angina, dyspnea, and sudden death	Caution with volatile anesthetics and propofol which decrease SVR	Maintain normal to high preload and maintain afterload
	LVOTO worsened by decreased preload and afterload, increased contractility	TEE to examine LVOTO, gradient, and residual VSD after myectomy	Avoid increases in contractility and tachycardia
	Hyperdynamic systolic function and abnormal diastolic function	External pacing/cardioversion/defibrillator pads prior to induction	
	Leading cause of death in young athletes		

Non-exhaustive list of adult congenital heart defects

ASD atrial septal defect, *VSD* ventricular septal defect, *PDA* patent ductus arteriosus, FiO_2 fraction of inspired oxygen, *PH* pulmonary hypertension, *RV* right ventricle, *OR* operating room, *ICU* intensive care unit, *PVR* pulmonary vascular resistance, *HR* heart rate, *CO* cardiac output, *SVR* systemic vascular resistance, *TEE* transesophageal echocardiography, *EKG* electrocardiogram, *CPP* coronary perfusion pressure, *PV* pulmonary valve, *AP* aortopulmonary, *RVOTO* right ventricular outflow tract obstruction, *PEEP* positive end-expiratory pressure, *NSR* normal sinus rhythm, *ccTGA* congenitally corrected transposition of the great arteries, *PPM* permanent pacemaker, *ICD* implantable cardioverter-defibrillator, *NPO* nil per oral, *BP* blood pressure, *PIP* peak inspiratory pressure, *RR* respiratory rate, *HOCM* hypertrophic obstructive cardiomyopathy, *LVOTO* left ventricular outflow tract obstruction collapse. Therefore, while early extubation is preferred, the readiness to extubate should be thoroughly evaluated before discontinuing mechanical ventilation.

Smaller studies have evaluated the feasibility of early extubation of ACHD patients in the operating room. Weismann and colleagues noted that a higher BMI and Risk Adjustment for Congenital Heart Surgery (RACHS) score were strong predictors of deferring extubation in the operating room [59]. Early extubation also results in improved outcomes and lower costs for the patients [60]. Bianchi and colleagues performed a retrospective analysis on 711 procedures and found that patients in the ultra-fast-track extubation cohort required less volume resuscitation, inotropes, and vasopressors and had a shorter ICU length of stay compared to those who underwent conventional extubation. Most importantly, the reintubation rate was low [61•]. Overall, appropriately selected ACHD patients can be great candidates for early extubation, whether it is fast-track in the ICU (within 6 h of ICU admission) or ultra-fast-track (in the operating room).

Where Should ACHD Care Take Place, and by Which Anesthesiologist?

Patients with ACHD, especially severe forms of disease, have decreased mortality when cared for in specialized ACHD centers [62]. This is likely due to the higher level of ACHD experience and expertise found in these centers. It follows that the perioperative care of the ACHD patient should occur at specialized ACHD centers with the relevant clinical ACHD expertise and resources to provide optimal patient care. The Adult Congenital Heart Association (ACHA), in efforts to standardize and advance the quality of ACHD care, outline specific staffing and services requirements for its ACHD Comprehensive Care Center Accreditation Program, including cardiac anesthesiologists with ACHD training or experience [63]. Both the 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease and the 2020 European Society of Cardiology (ESC) guidelines for the management of adult congenital heart disease emphasize the importance of a collaborative, multidisciplinary ACHD care team, identifying cardiac anesthesiologists with CHD training and expertise as key members [9••, 64]. The 2018 AHA/ACC Guidelines recommend that in all but the simplest cases of ACHD (ACHD AP classification IA), administration of anesthesia for invasive procedures should be performed by, or in collaboration with, an anesthesiologist with expertise in the management of patients with ACHD. In cases where clinical urgency precludes transfer of the patient, the 2018 AHA/ACC guidelines recommend that consultation with an anesthesiologist with ACHD expertise would be of benefit to on-site providers without ACHD expertise. Thus, it seems reasonable that clinicians with specific expertise in ACHD and CHD should be utilized whenever indicated.

It remains unclear who is best suited to provide anesthetic care for adult patients with CHD. Patients with ACHD can receive perioperative care at both pediatric and adult heart centers, so it stands to reason that those anesthesiologists with backgrounds in either adult cardiac anesthesia and/or pediatric cardiac anesthesia should provide their anesthetic care. However, most anesthesiologists in practice have very limited training or experience with CHD, and a retrospective closed claims analysis of adult patients with CHD found that intraoperative anesthetic care and lack of CHD knowledge were factors contributing to adverse events or patient injury [65]. Furthermore, a survey of anesthesiologists at a large academic anesthesia department found that overall knowledge of CHD was highly variable, and that fellowship-trained cardiac and pediatric anesthesiologists possessed the highest levels of CHD knowledge [66]. This demonstrates not only the importance of ACHD patients receiving appropriate perioperative care at specialized ACHD centers, but it also illustrates the need for further specialized ACHD training for those anesthesiologists who are charged with providing perioperative care for patients with ACHD. Filling this need for further ACHD training by anesthesiologists will be difficult given the shortage of specialized ACHD centers and board-certified ACHD cardiologists and other specialists [67, 68]. Currently, there are no specific ACHD training pathways or ACHD certifications for the cardiac or pediatric anesthesiologist. In practice, a myriad of different combinations of cardiac, pediatric, and pediatric cardiac anesthesia fellowship training and years of clinical experience comprise the teams of anesthesiologists who provide anesthetic care for ACHD patients. However, a proposal by the Congenital Cardiac Anesthesia Society to standardize pediatric cardiac anesthesia fellowship training has been accepted by the Accreditation Council for Graduate Medical Education [69], emphasizing the current need to augment CHD anesthesia education. As the population of adults with CHD grows, there will also be a need to address the deficiency of anesthesia training specific to ACHD.

Conclusion

Improvements in clinical care of ACHD have ensured that more patients are living into adulthood than ever before. As these patients grow older, they will require numerous surgical operations, percutaneous interventions, electrophysiology procedures, and non-cardiac surgeries, most of which necessitate exposure to anesthesia. Many of these patients are critically ill, so a multidisciplinary approach to their perioperative care is essential. The cardiac anesthesiologist must possess a thorough understanding of the cardiac pathophysiology and extracardiac manifestations relevant to each individual patient to ensure optimal care is provided. Recent ACHD guidelines emphasize the need for perioperative care to occur at specialized ACHD centers with involvement of cardiac anesthesiologists who have expertise in the anesthetic management of patients with ACHD. Given a growing ACHD population and a shortage of specialized ACHD centers and board-certified ACHD physicians, new efforts should be made to train and educate anesthesiologists with the expertise needed to provide optimal and safe anesthetic care for these patients.

Declarations

Conflict of Interest The authors declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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