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Anesthesia for Noncardiac Surgery in Adults with Congenital Heart Disease

Maxime Cannesson, M.D., Michael G. Earing, M.D.,† Vincent Collange, M.D.,* Judy R. Kersten, M.D., F.A.C.C.‡*

CONGENITAL heart defects are the most common group of birth defects, occurring in approximately 8 in 1,000 live births.[1] Excluding bicuspid aortic valves, the majority of untreated patients born with congenital heart disease die in infancy or childhood, and only 15–25% survive into adulthood.[2] Advances in prenatal diagnosis, interventional cardiology, pediatric cardiac surgery, anesthesia, and critical care have resulted in survival of approximately 90% of these children to adulthood. Now, for the first time in history, estimates suggest that more adults than children are living with congenital heart disease (CHD) in the United States.[3–4] Many of these patients will require additional palliative or curative cardiac surgery and noncardiac surgery at some time during adulthood. Adults with CHD demonstrate specific and complex anatomy and physiology. Perioperative morbidity and mortality are increased in adults with repaired or palliated CHD; however, no major study focusing on this topic has been performed.[5] Few guidelines are available to direct the management of these challenging patients. Nevertheless, a task force of the American College of Cardiology recommended that adult patients with moderate to severe CHD undergoing noncardiac surgery should be referred to an adult CHD center to obtain appropriate consultation with expert cardiologists and anesthesiologists.[6]

The purpose of this article is to provide an overview of the long-term consequences and the preoperative and intraoperative implications of CHD for the anesthesiologist involved in the care of adults with CHD undergoing noncardiac surgery.

Epidemiology of CHD

About 25% of adults with CHD have a mild form of the disease that has allowed them to survive into adulthood without surgical or interventional cardiac catheterization. The most common lesions in this category include mild aortic valve stenosis (usually in setting of bicuspid aortic valve), small restrictive ventricular septal defects, atrial septal defects, mild pulmonary valve stenosis, mitral valve prolapse, and isolated congenitally corrected transposition of the great arteries (table 1). The vast majority of adults with CHD seen in the outpatient setting, however, are patients who have had previous surgical or catheter-based intervention (table 1).

CHD lesions can be functionally classified into those that produce left to right shunts (acyanotic) and those that produce cyanosis (right to left shunting). A left to right shunt exists when oxygenated blood from the left atrium, left ventricle, or aorta transits to the right atrium, right ventricle, or the pulmonary artery. Thus, the lungs receive all the deoxygenated blood from the systemic venous return (effective pulmonary blood flow; amount of deoxygenated blood that is carried to lungs to be oxygenated) plus the volume of fully oxygenated blood that is shunted through the defect. This results in volume overload of one or more cardiovascular chambers or structures depending on the location of the defect. If the defect is large and nonrestrictive, there is both increased flow and transmission of near systemic pressure to the pulmonary vascular bed. Over time, this can lead to irreversible changes in the pulmonary vascular bed, leading to increased pulmonary vascular resistance and associated pulmonary artery hypertension. If the pulmonary artery pressure is at systemic levels, there may be reversed (right to left) or bidirectional shunting at the level of the defect (Eisenmenger syndrome).

Atrial and ventricular septal defects are among the most common congenital abnormalities, constituting 25% of all adult CHD.[7] Other lesions also considered in the acyanotic group that are commonly seen in busy adult congenital outpatient clinics include but are not limited to coarctation of the aorta, congenital aortic

* Assistant Professor of Anesthesiology, Departments of Anesthesiology and Intensive Care, Hospices Civils de Lyon, Louis Pradel Hospital, Claude Bernard Lyon 1 University. † Assistant Professor of Pediatrics, Division of Cardiology, Children's Hospital of Wisconsin Herma Heart Center. ‡ Professor and Vice Chair of Anesthesiology, Departments of Anesthesiology and Pharmacology and Toxicology, Medical College of Wisconsin.

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Address correspondence to Dr. Cannesson: Service d'Anesthésie Réanimation, Hôpital Cardiologique Louis Pradel, 200 avenue du Doyen Lépine, 69500 Bron, France. maxime_cannesson@hotmail.com.

Table 1. Common Congenital Heart Defects Seen in Adult Congenital Heart Disease Centers

Conotruncal abnormalities after repair (Tetralogy of Fallot, truncus arteriosus, double outlet right ventricle)
Coarctation of the aorta after repair
Transposition of the great arteries after atrial or arterial switch procedure
Complex single ventricles after the Fontan procedure
Pulmonary valve stenosis
Congenital aortic valve stenosis
Atrioventricular canal defects (complete and partial) after repair
Secundum atrial septal defects
Congenitally corrected transposition of the great arteries
Sinus venosus atrial septal defects with partial anomalous pulmonary venous return

valve disease with associated stenosis, regurgitation or most commonly both, subaortic stenosis, congenital abnormalities of the mitral valve leading to stenosis and/or regurgitation, congenitally corrected transposition of the great arteries, and Ebstein's anomaly of the tricuspid valve that is typically a cyanotic lesion in infancy.

Cyanotic heart disease includes those anatomical heart defects that produce a limitation in pulmonary blood flow or result in mixing of oxygenated and deoxygenated blood. Both conditions lead to decreased blood oxygen content and cyanosis. Unlike the acyanotic forms of congenital heart disease, the majority of patients with cyanotic congenital heart disease will have had at least one and often several previous interventions before adulthood. The most frequent defects seen in the outpatient adult congenital setting include tetralogy of Fallot, complete transposition of the great arteries (also known as D-transposition), and various forms of single ventricles.[8] Other defects include total anomalous pulmonary venous return, truncus arteriosus, and double outlet right ventricle.

Long-term Consequences of CHD and Effect on Anesthesia Management

Adult patients with CHD are now surviving longer than ever before, and it is becoming increasingly apparent that even the most simple lesions can be associated with long-term complications.[9] Long-term cardiac complications include: pulmonary hypertension, ventricular dysfunction, dysrhythmias and conduction defects, residual shunts, valvular lesions (regurgitation and stenosis), hypertension, and aneurysms. Noncardiac sequelae include secondary erythrocytosis, cholelithiasis, nephrolithiasis, developmental abnormalities, central nervous abnormalities, such as seizure disorders from previous thromboembolic events or cerebrovascular accidents, hearing or visual loss, and restrictive and obstructive lung disease. Adult patients with CHD requiring noncardiac surgery can be viewed on a continuum in which some patients have defects that have not been corrected, some have received palliative repairs (*e.g.*, partial or total cavopulmonary shunts), and others have undergone complete anatomic correction. Some patients will present with mild disease and will require less aggressive management, whereas others with complex disease will require care from cardiologists and anesthesiologists with significant expertise in CHD. In nearly all cases, CHD in adults should be viewed as a systemic condition with associated multiorgan dysfunction.[10,11]

Pulmonary Hypertension

Adults with CHD may develop pulmonary hypertension for a variety of reasons. Potential etiologies include pulmonary venous hypertension secondary to elevated ventricular end diastolic pressure, elevated pulmonary venous atrial pressure, or pulmonary vein stenosis. Many of these patients also continue to have decreased oxygen saturation secondary to residual shunts, poor lung function, and persistent decreased pulmonary blood flow. The main etiology for pulmonary hypertension in adults with congenital heart disease, however, is the presence of long-standing large and nonrestrictive defects. This allows for both increased flow and transmission of near systemic pressure to the pulmonary vascular bed, leading to irreversible vascular changes and elevated pulmonary vascular resistance. Manifestations begin in childhood and are progressive. Early on increases in pulmonary vascular resistance may be reversible; over time, however, increases become permanent. Vascular changes include hypertrophy of the media of small muscular arteries and arterioles, intimal cellular proliferation, smooth muscle cell migration into the subendothelium, progressive fibrosis, and obliteration of arterioles and small arteries.[12] Eisenmenger syndrome refers to the development of pulmonary hypertension secondary to long-standing left to right shunting.

Patients with Eisenmenger syndrome represent a particular challenge for the anesthesiologist. Perioperative mortality is increased,[13] and noncardiac surgery should only be performed if absolutely essential in patients with Eisenmenger physiology. Predictors of mortality include syncope, age at presentation or development of symptoms, poor functional class, supraventricular dysrhythmias, elevated right atrial pressures, low oxygen saturation (less than 85%), renal insufficiency, severe right ventricular dysfunction, and trisomy 21.

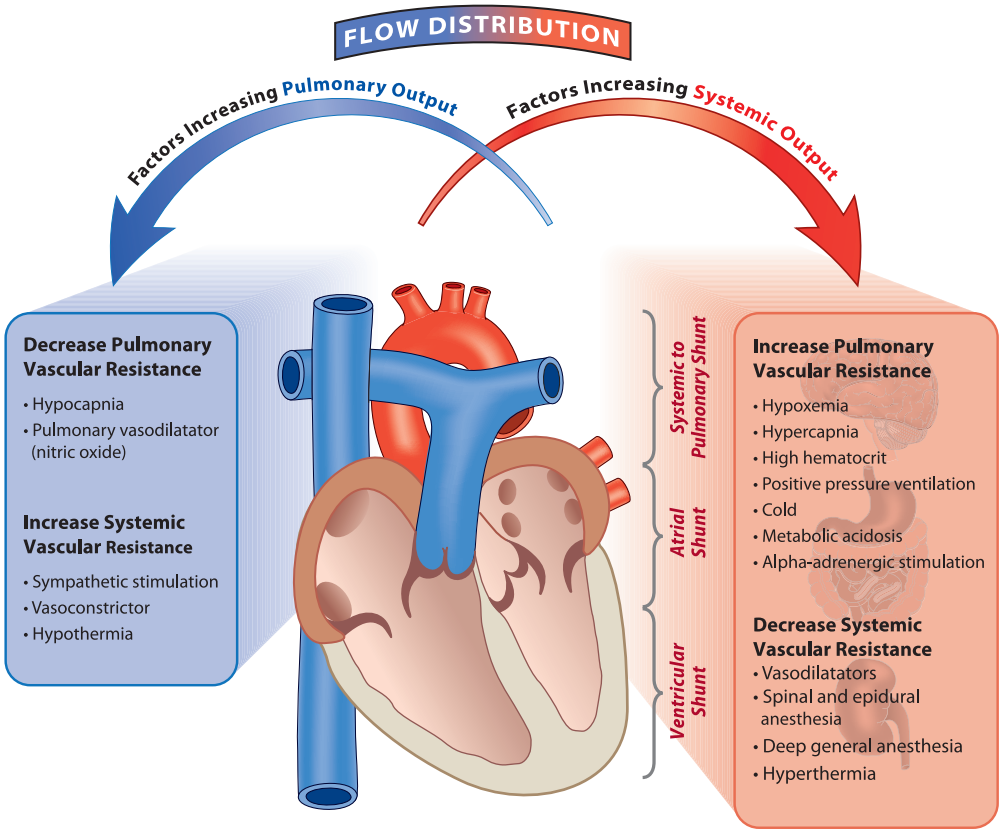

Fig. 1. Factors that influence the distribution of blood flow between the systemic and pulmonary circulations are shown.

A primary goal of anesthetic management in patients with pulmonary hypertension is to minimize increases in pulmonary vascular resistance and to maintain systemic vascular resistance (fig. 1). Abrupt increases in pulmonary vascular resistance may precipitate either acute right ventricular failure and decreased cardiac output in patients without intracardiac shunting or oxygen desaturation followed by decreased cardiac output in patients with intracardiac shunting. In both cases, severe bradycardia may occur with progression to cardiac arrest. Prevention and treatment of pulmonary hypertensive crisis includes hyperventilation (with 1.0 fractional inspired oxygen concentration), correction of acidosis, avoidance of sympathetic nervous system stimulation, maintenance of normothermia, minimization of intrathoracic pressure, and use of inotropic support. Inhaled nitric oxide may be useful to treat sudden increases in pulmonary vascular resistance, and this drug should probably be available in the operating room for use in high-risk patients. Regional anesthesia may be an acceptable alternative to general anesthesia for patients undergoing peripheral procedures. However, spinal or epidural anesthesia may produce unacceptable decreases in systemic vascular resistance in patients with unrestrictive intracardiac shunts, and this action could exacerbate right to left shunting. Conversely, general anesthesia allows for optimal control of ventilation and may be preferable in patients undergoing high-risk surgery.

Bleeding and Thrombosis Risk

Most patients with Eisenmenger syndrome will be severely cyanotic, defined as an oxygen saturation of no more than 85%. Cyanotic patients are at high risk for both perioperative bleeding and thrombosis, even during minor surgery. Both are secondary to abnormalities in platelet number and function as well as complex abnormalities in the coagulation system. Thrombocytopenia is felt to be secondary to shortened platelet survival due to peripheral consumption. The abnormalities in the coagulation pathways are less understood. Patients with severe cyanosis have low levels of circulating vitamin K–dependent clotting factors, factor V, and von Willebrand factor. This leads to an elevated International Normalized Ratio and a prolonged activated partial thromboplastin time. These patients, however, do not have an elevated bleeding time, likely secondary to increased blood viscosity and decreased flow. Further contributing to the bleeding risk is the presence of arteriolar dilation and increased tissue vascularity. This is felt to be secondary to increased release of endothelium-derived nitric oxide and prostaglandins in response to increased wall sheer stress in the setting of increased blood viscosity.

Although patients with cyanosis have an increased risk for bleeding, this is not protective against thrombosis. In the setting of cyanosis, these patients develop secondary erythrocystosis. Secondary erythrocytosis develops as a compensatory response to chronic hypoxia and results from overproduction of erythropoietin. This results in increased whole blood viscosity resulting from increased red blood cell mass and decreased plasma volume. The end result is decreased flow in the small arterioles and capillaries. This is further exacerbated in the setting of iron deficiency and dehydration. Iron-deficient red blood cells are less deformable and have been found to be one of the strongest independent predictors of thrombosis in the setting of Eisenmenger syndrome.[14]

In the perioperative setting, preoperative fasting might exacerbate symptoms of hyperviscosity and increase the risk of cerebrovascular thrombosis. Thus, adequate hydration with intravenous fluids must be maintained, particularly in fasting patients. Although euvolemic phlebotomy is no longer a routine practice, preoperative phlebotomy to improve surgical hemostasis may be useful when hematocrit levels exceed 65%.[8]

Careful preoperative assessment of the coagulation system is essential, and replacement of coagulation factors and platelets should be considered in patients undergoing moderate or major surgery. In addition, iron deficiency should be corrected preoperatively if the procedure is not urgent. It is important to note, however, that in the setting of secondary erythrocytosis with increased hemoglobin and decreased plasma volume, standard techniques to measure International Normalized Ratio and activated partial thromboplastin time may be unreliable. Because of these changes, the concentration of citrate in the sampling tube must also be adjusted. In most centers, the anticoagulant in the tube can be adjusted by the following formula: anticoagulant in sampling tube (3.8% citrate) in ml = {(100 – hematocrit)/100} + 0.02 for a draw of 10 ml of whole blood.

Heart Failure

Right-sided and left-sided heart failure are common complications of both corrected and uncorrected CHD. Increases in atrial natriuretic peptide, renin, aldosterone, and norepinephrine have been observed in adults with CHD many years after surgical correction, even in asymptomatic patients. Abnormal cardiac autonomic nervous system regulation and altered hemodynamics contribute to the development of heart failure in these patients. Management of left ventricular failure with diuretics, digoxin, angiotensin-converting enzyme inhibitors, and β-blockers is similar to other forms of acquired heart failure, and treatment should be optimized in the perioperative period.[5] In contrast to left ventricular failure, there are no evidence-based guidelines for the management of heart failure in patients with a systemic right ventricle (congenitally corrected transposition of the great arteries, Mustard, or Senning repairs of transposition of the great arteries, and single ventricles).[15] Further clinical trials are warranted.

Dysrhythmias

Atrial and ventricular dysrhythmias are common in adults with CHD. Dysrhythmias arise in patients who have undergone previous curative or palliative surgery as a primary consequence of the underlying congenital defect or secondary to surgical repair.[16,17] For example, supraventricular dysrhythmias occur in 20–45% of patients with previous atrial surgery (late atrial septal defect closure,[18] Mustard, Senning, or Fontan procedures) or in those with atrial distension. The most common form of tachyarrhythmia observed is intraatrial reentrant tachycardia originating from the right atrium. Atrial tachyarrhythmias are often resistant to pharmacological treatment and can result in rapid hemodynamic deterioration. Ventricular dysrhythmias are most frequently encountered in patients who have significantly decreased right or left ventricular function. Other risk factors include previous ventriculotomy, earlier surgical era, or older age at initial surgery. Patients who were repaired late are exposed to longer periods of cyanosis, volume overload, and pressure overload. As a result, they have increased myocardial fibrosis and associated slowing of conduction and an increased risk for dysrhythmias. Acute hypoxemia can provoke ventricular dysrhythmias because subendocardial myocardial perfusion is already impaired in hypertrophied myocardium. Some patients will require a permanent pacemaker to treat bradycardia secondary to postoperative atrioventricular block. The management of patients with pacemakers and intracardiac defibrillators has been reviewed elsewhere.[19]

Anesthetic Management

Preoperative Evaluation

The preoperative evaluation of patients with CHD undergoing noncardiac surgery should use a multidisciplinary approach that includes the participation of anesthesiologists, cardiologists, intensivists, and surgeons. Guidelines of the task force for the organization of delivery systems for adults with CHD recommend that patients with moderate or complex CHD should be managed in a regional adult congenital heart disease center.[3,6] Perioperative risk is substantially increased in adults with CHD, particularly in those with poor functional class, pulmonary hypertension, congestive heart failure, and cyanosis.[20] Major surgery and procedures that involve one lung ventilation or changes in position (*e.g.*, prone, Trendelenburg) could produce important hemodynamic effects that contribute to increasing risk. Therefore, the anesthesiologist should be familiar with the patient's specific anatomy and physiology as determined from echocardiographic and cardiac catheteriza-

tion results. This knowledge is useful to anticipate intraoperative events that may precipitate acute changes in the magnitude or direction of intracardiac shunts or modulate flow through systemic to pulmonary shunts. If recent examination results are not available, a preoperative echocardiogram may be indicated.[20]

Premedication

Many adults with CHD undergoing noncardiac surgery have undergone previous cardiac surgery and are familiar with anesthesia. Some may present with anxiety, physical limitations, and associated anomalies or syndromes (the most common is trisomy 21). Adults with CHD are more likely to be living with their parents and to develop a variety of psychosocial issues.[20] Consequently, psychological preparation of patients for surgery is important. Premedication with anxiolytics and hypnotics must be undertaken very cautiously because hypoventilation and hypercapnia may produce deleterious increases in pulmonary vascular resistance, particularly, in patients with underlying pulmonary hypertension or systemic to pulmonary shunts. However, patients with chronic hypoxemia retain a normal ventilatory response to hypercarbia as well as to opioid analgesics.

Endocarditis Prophylaxis

The American Heart Association has recently published updated guidelines for the prevention of infective endocarditis.[21] After reviewing the literature over the last 40 yr, an expert panel found that very few cases of endocarditis could have been prevented with antibiotic prophylaxis. As result, only patients with cardiac conditions associated with the highest risk for adverse outcomes should continue following antibiotic prophylaxis before surgery: patients with previous endocarditis, unrepaired cyanotic CHD, including palliative shunts and conduits; completely repaired congenital heart defects with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure; repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization). Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for other forms of CHD.

Intraoperative Management

Adults with CHD who have undergone complete anatomic repair and have no evidence of late functional deterioration can be managed by using conventional approaches. In contrast, patients with more complex CHD and moderate to major surgery will require specific intraoperative management.

Monitoring

In addition to direct examination of the patient, standard conventional noninvasive monitoring including pulse oximetry, electrocardiogram, arterial blood pressure, capnography and temperature are used in all patients. Pulse oximetry is perhaps uniquely important in the management of CHD. For example, decreases in arterial saturation can signify increases in pulmonary vascular resistance, increases in right to left shunting, or decreases in pulmonary blood flow through systemic to pulmonary shunts. In contrast, increases in left to right shunting may not be detected by pulse oximetry and arterial oxygen saturation may be maintained even if systemic cardiac output is severely compromised. The capnogram is altered, and end tidal carbon dioxide concentrations underestimate $Paco_2$ in the case of right to left shunting.

Knowledge of the anatomy and physiology of specific palliative repairs is important for choosing appropriate monitoring. For example, congenital defects that are associated with inadequate pulmonary blood flow (*e.g.*, pulmonary valve atresia or univentricular heart) are palliated with systemic to pulmonary shunts. In patients with a classic Blalock-Taussig shunt (end to side anastomosis of the subclavian and pulmonary arteries) arterial pressure and Spo_2 cannot be measured on the ipsilateral side. A Glenn shunt or bidirectional cavopulmonary anastomosis consists of an end to side anastomosis of the divided superior vena cava to pulmonary artery. Total cavopulmonary connection (Fontan circulation) is established when the pulmonary and systemic circulations are totally separated by diverting all the systemic venous return to the pulmonary artery, usually without interposition of a subpulmonic ventricle. These alterations in intracardiac anatomy complicate the placement of central venous catheters in palliated adults, and the anatomical variations must be considered when interpreting values obtained from central venous monitoring. For example, in patients with a Fontan circulation, central venous pressure reflects mean pulmonary artery pressure. In patients with an intraatrial baffle (*e.g.*, Mustard or Senning procedure), pulmonary artery catheter placement may be difficult or impossible. Vascular access may also be difficult because many of these patients have already undergone previous vessel catheterization. Invasive arterial pressure monitoring can be essential in managing patients with Eisenmenger syndrome, intracardiac or systemic to pulmonary shunts undergoing major surgery who are also sensitive to sudden changes in preload, and systemic and pulmonary vascular resistance. Finally, transesophageal echocardiography might be useful in adults with CHD undergoing noncardiac surgery to monitor intravascular volume status and ventricular function. In the presence of complex CHD, transesophageal echocardiography should be performed by an individual familiar with CHD.

Table 2. Proposed Management Strategies for Specific Defects

Congenital Heart Disease	Anatomy - Physiology	Potential Issues	Specific Anesthetic Management
Atrial septal defect	L-R shunt	Small to moderate size defects well tolerated Atrial fibrillation (increased risk if repaired after age 40) Risk of paradoxical emboli Large defects lead to arrhythmias, exercise intolerance, and rarely PHT (occurs in less than 5% of patients)	De-air intravenous lines
Ventricular septal defect	L-R shunt May be associated with other defects	Unrepaired: Large defect risk of PHT (50% by age 2) Small to moderate size defects, risk for endocarditis, sub-pulmonic obstruction, subaortic obstruction, and aortic regurgitation Right ventricular failure Repaired: Complete heart block in some patients (rare) Persistent PHT Dysrhythmia	Manage L-R shunt Maintain pulmonary blood flow if R-L shunt present Increased risk of postoperative pulmonary infection Manage pacemaker
Coarctation of the aorta	LV pressure overload and hypertrophy Aortic branch collaterals Associated with bicuspid aortic valve (50-80%) Endothelial dysfunction (diffuse aortopathy)	Blood pressure gradient between upper and lower limbs Risk of bleeding if thoracic surgery LV hypertrophy and LV diastolic dysfunction Systemic hypertension Aneurysms of ascending aorta and descending aorta Premature coronary artery disease Intracranial aneurysms (10%)	Inaccurate blood pressure (left arm) if previous subclavian angioplasty Postoperative hypertension Avoid tachycardia, hypotension
Aortic stenosis	LV pressure overload and hypertrophy	Unrepaired: Pulmonary edema PHT Myocardial ischemia Syncope Post stenotic dilatation Repaired: Aortic regurgitation LV diastolic dysfunction	Avoid tachycardia, hypotension Avoid factors that increase myocardial oxygen consumption
L-(congenitally corrected) transposition of the great arteries	LV is the subpulmonic ventricle RV is the systemic ventricle	Unrepaired: Complete heart block Arrhythmias (atrial and ventricular) Anatomic right ventricular failure Systemic AV valve regurgitation Repaired: Same	Pacemaker management External pacing available Manage dysrhythmias Manage heart failure

(continued)

Anesthetic Technique

There are no evidence-based recommendations to guide the anesthetic management of patients with CHD undergoing noncardiac surgery. Given the large scope of abnormalities encompassed by CHD,[22,23] it is also impossible to propose a single approach for anesthetic management that would address every possible defect. However, a major objective of intraoperative management is to promote tissue oxygen delivery by preventing arterial desaturation, maintaining a balance between pulmonary and systemic flows, and by optimizing hematocrit. Management strategies for specific defects are outlined in table 2.

Anesthetic Agents

There are few studies that have evaluated the hemodynamic effects of anesthetic agents in adult patients with CHD. Most intravenous agents depress myocardial contractility and decrease systemic vascular resistance, and these actions could have an adverse effect on tissue oxygen delivery during induction of anesthesia. Alternatively, some evidence suggests that etomidate may pro-

Table 2. continued

Congenital Heart Disease	Anatomy - Physiology	Potential Issues	Specific Anesthetic Management
Tetralogy of Fallot	Pulmonic stenosis (valvular, subvalvular, and/or supravalvular) RV hypertrophy Overriding aortic root VSD Cyanosis	Unrepaired: Rare, mean age of death 25 yr of age R-L shunt Cyanosis Palliated: Blalock-Taussig shunt Chronic left ventricular volume overload Cyanosis if shunt is too small Pulmonary hypertension Repaired: Sinus and AV node dysfunction Dysrhythmia: atrial and ventricular Ascending aortic aneurysm Residual pulmonary regurgitation or stenosis Residual VSD Left ventricular dysfunction Persistent pulmonary hypertension from previous shunts RV failure from chronic pulmonary insufficiency	Avoid tachycardia, hypovolemia, increased contractility Maintain pulmonary blood flow Maintain sytemic blood pressure Detect and manage dysrhythmias Manage pacemaker External pacing available
D-transposition of the great arteries	Pulmonary artery arises from LV Aorta arises from RV Possible associated lesions: VSD, ASD, PDA, Pulmonary stenosis, coarctation of the aorta Abnormal coronary artery anatomy	Unrepaired: Associated with VSD or ASD or PDA Repaired: Senning or Mustard Atrial dysrhythmia Sinus node dysfunction (by age 40, 50% have pacemaker) Systemic ventricle dysfunction Residual atrial or ventricular level shunts Repaired: Arterial switch Myocardial ischemia (narrowed, occluded coronary arteries, endothelial dysfunction) Ascending aortic aneurysm	Maintain pulmonary blood flow Manage dysrhythmia Manage heart failure Detailed preoperative evaluation, both functional and coronary imaging study
Univentricular heart	Double inlet atrioventricular connections Absence of one atrioventricular connection Single well-developed ventricle	Unrepaired: Rare Dysrhythmias Congestive heart failure Bidirectional shunting Cyanosis PHT Repaired: Blalock-Taussig shunt, Glenn shunt, or Fontan Dysrhythmias Heart failure Hepatic dysfunction Thromboemboli Restrictive lung disease	Manage dysrhythmias Maintain pulmonary blood flow Manage dysrhythmias Maintain low pulmonary vascular resistance Maintain adequate preload Replace coagulation factors

ASD = atrial septal defect; AV = atrioventricular; L-R = left to right; LV = left ventricle; PDA = patent ductus arteriosus; PHT = pulmonary hypertension; R-L = right to left; RV = right ventricle; VSD = ventricular septal defect.

vide hemodynamic stability in the setting of CHD[17] similarly to other settings of impaired cardiac function. The potential favorable effects of ketamine in this population have not been adequately investigated. Ketamine has been suggested to increase pulmonary vascular resistance in adults without CHD.[24] However, this agent produces beneficial effects in children with CHD and severe pulmonary hypertension undergoing sevoflurane anesthesia[25] by maintaining systemic vascular resistance and ventricular performance, without increasing pulmonary vascular resistance. As in the case of intravenous anesthetic agents, the choice of a specific volatile anesthetic agent to be used should be based on the patient's physiology and the overall goal of balancing pulmonary and systemic blood flow.

Intracardiac and Systemic to Pulmonary Shunts

Shunting has an important effect on anesthetic management. All intravenous lines must be meticulously deaired to decrease the risk of systemic air embolization in patients with intracardiac shunts. The hemodynamic effects of ventilation strategies, positioning, pharmaco-

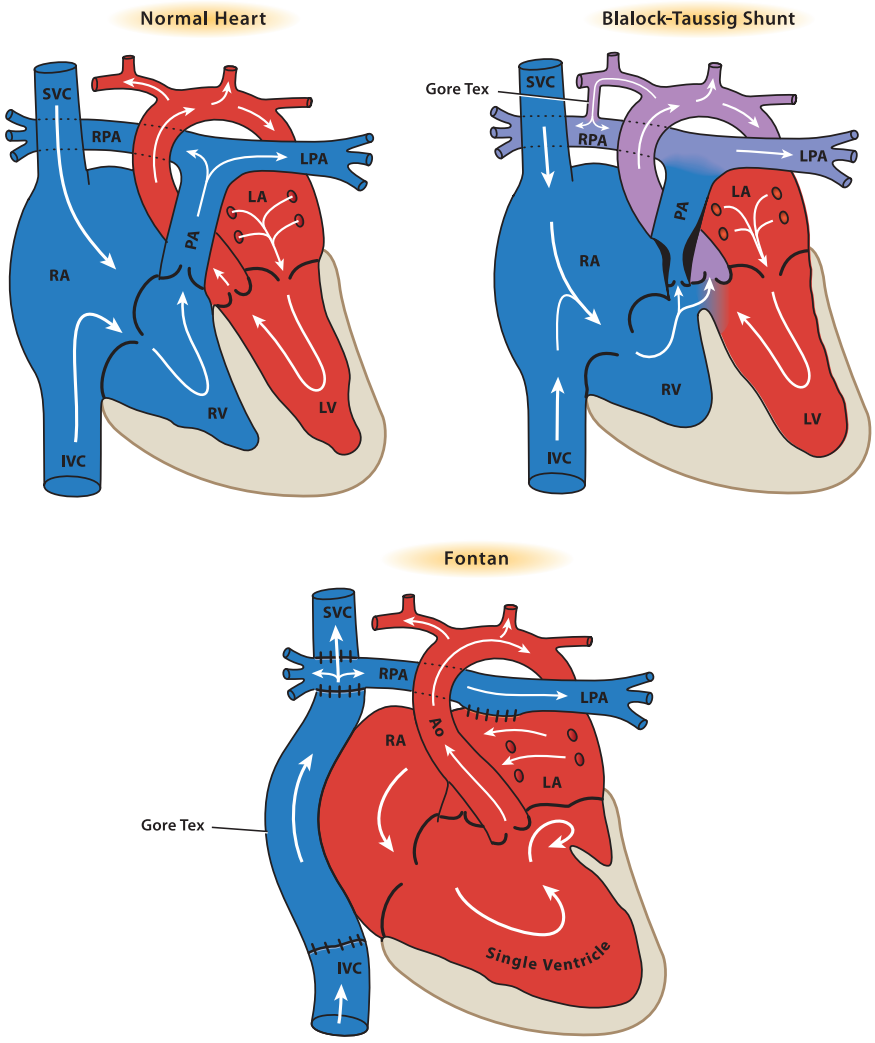

Fig. 2. Schematic representations of normal heart, Blalock Taussig shunt, and Fontan circulation. SVC = superior vena cava; IVC = inferior vena cava; RA = right atrium; RV = right ventricle; PA = pulmonary artery; LPA = left pulmonary artery; RPA = right pulmonary artery; LV = left ventricle; LA = left atrium; Ao = aorta.

logical agents, and blood loss must all be considered to appropriately balance pulmonary and systemic blood flow in patients with intracardiac or systemic to pulmonary shunts (figs. 1 and 2). Ventilation with high airway pressures can compromise venous return, increase pulmonary vascular resistance, and exacerbate right to left shunting in patients with cyanotic heart disease. Inadequate anesthesia and sympathetic nervous system stimulation might increase systemic vascular resistance, exacerbate left to right shunting, and reduce systemic cardiac output in a patient with a large atrial septal defect. Trendelenburg position can increase central venous (superior vena cava) pressure and cause cerebral hypoperfusion in a patient with a Glenn shunt or Fontan. Systemic hypotension can also result in a decrease in pulmonary blood flow, with subsequent arterial desaturation, in a patient with a systemic to pulmonary artery shunt (Blalock-Taussig shunt, or central shunt). These are a few examples of the complex physiology that must be considered in managing adults with CHD for noncardiac surgery.

Univentricular Heart

Single ventricle anatomy and physiology is probably, along with Eisenmenger syndrome, the most challenging CHD for the anesthesiologist to manage. The Fontan procedure was first described in 1971 and was applied initially to patients with tricuspid atresia. Today, indications for this procedure have been extended to all forms of single ventricles. Since its initial description, at least 10 different variations have been performed. In all forms, this operation bypasses the right ventricle, leaving passive, nonpulsatile flow from both inferior and superior vena cava to the pulmonary artery.[26] Any factor that increases pulmonary vascular resistance will decrease pulmonary blood flow and cause arterial desaturation. Patients with a Fontan circulation frequently present with cardiac and noncardiac complications that include supraventricular dysrhythmias, restrictive lung disease, thromboembolic complications,[27] and hepatic dysfunction. Both procoagulant and anticoagulant effects[28] are observed in patients with a Fontan as a result of liver dysfunction and/or factor loss in patients with protein-losing enteropathy, and these abnormalities substantially

increase the risk for intraoperative bleeding. Patients with a Fontan circulation should maintain an arterial saturation above 90 to 95%. Arterial saturation below 90% in these patients should be considered abnormal and should provoke further evaluation for the presence of venovenous collaterals, arteriovenous malformations, or a residual shunt.

Postoperative Management

Patients presenting with severe CHD and/or high risk surgery should be managed if possible in a postoperative intensive care unit experienced with caring for adults with congenital heart disease. The major risks during the postoperative period are similar to the risks described above. These risks include bleeding, dysrhythmias, and thromboembolic events. In cases of pulmonary hypertension, oral pulmonary vasodilatators such as sildenafil and inhaled nitric oxide may be beneficial.

Conclusions

The number of adult patients with CHD is rapidly increasing, and these patients will be presenting with greater frequency for noncardiac surgery. The cardiovascular anatomy and physiology of CHD are complex and require specific knowledge of the defect and its anesthetic implications. Adults with moderate or severe CHD requiring noncardiac surgery are at high risk, particularly those with poor functional class, pulmonary hypertension, congestive heart failure, and cyanosis; and these patients should receive care in a regional adult CHD center with multidisciplinary collaboration among anesthesiologists, cardiologists, surgeons, and intensivists. There are no evidence-based guidelines for the perioperative management of adults with CHD. Large-scale clinical trials are required to elucidate the optimal anesthetic management of these challenging patients.

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