Chapter 17: Anesthesia for Cardiovascular Procedures

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INTRODUCTION

FOCUS POINTS

1. Fetal cardiac development begins at approximately 22 days of gestation. The fetal circulation allows for preferential shunting of oxygenated blood to the brain and heart.
2. Beginning at birth, the cardiovascular system undergoes drastic physiological changes as it transitions from a parallel to a series circulation. In the transitional circulation, the fetal shunts (ductus arteriosus, ductus venosus, and foramen ovale) close functionally and eventually, anatomically.
3. Normal vital signs value change with age, reaching adult values in adolescence.
4. Congenital heart disease is the most common form of birth defect with an incidence of between 4 and 7 per 1000 live births. Patients with congenital heart disease may have associated extracardiac anomalies and genetic syndromes.
5. Congenital heart disease (CHD) can be classified as cyanotic or acyanotic, depending on the presence or absence of right-to-left shunting.
6. The magnitude of shunting and its hemodynamic significance depends on the location and size of the shunt as well as the pressure gradient across the shunt and the relative compliances of the downstream chambers or resistances of the downstream vessels.
7. Pediatric heart failure can be related to volume- or pressure-overload. It can occur in structurally normal heart with primary cardiomyopathy (dilated, hypertrophic, or restrictive) or secondary cardiomyopathy due to arrythmia, ischemic, toxicity, infection, or infiltrative diseases.
8. The approach to the patient with CHD undergoing noncardiac surgery should be systematic and team-based.

A thorough understanding of cardiovascular physiology and pathophysiology is an unequivocally essential component of the practice of anesthesiology. Pediatric anesthesiologists must be cognizant of the normal physiologic changes that the cardiovascular system undergoes during growth and development, from fetal through adult life. They must be familiar with the broad spectrum of pathophysiology that accompanies congenital, and, to a lesser extent, acquired heart disease and its management. In this chapter, we will review the most salient aspects of this expansive subject and provide a basic framework for the clinical management of the patient with cardiac disease presenting for noncardiac surgery.

DEVELOPMENTAL PHYSIOLOGY

Fetal cardiac development begins at approximately 22 days' gestational age. Several intrauterine shunts form to divert blood away from the fetal lungs, which do not participate in gas exchange. These shunts also help optimize oxygen delivery to the developing brain and heart. Blood that has been oxygenated in the maternal circulation crosses the placenta and enters the fetal circulation via the umbilical vein with an oxygen saturation of 70% to 80%. The umbilical vein enters at the level of the liver, where a portion of the blood provides perfusion to the hepatic circulation and the rest is shunted across the **ductus venosus** toward the heart. This blood travels toward the right atrium (RA) independently from inferior vena cava (IVC) blood. The blood from the ductus venosus enters the RA and is shunted across the **foramen ovale** to the left atrium (LA), where it mixes with the small amount of blood that made it through the lungs and into the pulmonary veins. This mixture of blood (oxygen saturation about 65%) is ejected into the

ascending aorta to preferentially supply oxygenated blood to the brain and heart. Deoxygenated blood from the superior vena cava (SVC) and IVC enters the RA and the right ventricle (RV) and exits via the pulmonary arteries, with a portion of this blood perfusing the lungs but the majority shunted right-to-left across the **ductus arteriosus** to the descending aorta. This blood has an approximate oxygen saturation of 60%.[1–3] In the fetal circulation, the branch pulmonary arteries are small, because the lungs only receive approximately 15% of combined ventricular output. The RV handles 55% of the combined ventricular output, and the LV handles the other 45%; thus, the RV is larger and more dominant than the left ventricle (LV). The pressure in the RV is identical to that in the LV, which is not the case in adult circulation.[4]

The transitional circulation consists of changes to the fetal circulation to accommodate extrauterine life. Once the fetus takes its first breath, the pulmonary vascular resistance (PVR) decreases secondary to the increased oxygen saturation, and ventilation and expansion of the fetal lungs. In addition, the neonate's systemic vascular resistance (SVR) increases due to clamping of the umbilical cord, which disconnects the low-resistance placenta from neonatal circulation. The increased SVR and increased pulmonary blood flow lead to increased left atrial pressures and functional closure of the foramen ovale.[1,3] The decrease in PVR also creates a shift to left-to-right blood flow across the ductus arteriosus with an increase in pulmonary blood flow, which increases left ventricular output and stroke volume. Decreases in intraluminal flow and circulating prostaglandins coupled with increases in vasoactive catecholamines and oxygen tension lead to the functional closure of the ductus arteriosus within the first 10 to 15 minutes of life.[1] Normal anatomic closure via vascular remodeling does not occur until 2 to 3 weeks of age. If PVR exceeds SVR before this occurs, blood flow via the ductus arteriosus can reverse (to become right-to-left) leading to differential cyanosis.

Throughout the first weeks of life, the neonatal heart is noncompliant and functions at close to maximum cardiac output with a fixed stroke volume. For this reason, cardiac output is heart rate–dependent. Increases in afterload result in a decrease in cardiac output.[5] Additionally, the immature heart has a significant parasympathetic tone, so infants and young children are prone to bradycardia with vagal stimulation, such as during tracheal intubation.

As the neonate grows, additional changes take place. The pediatric heart rate reaches a maximum rate at approximately 1 month of life, and then steadily declines to reach adult levels at 10 to 12 years of age. In contrast, there is a decline in respiratory rate from birth to early adolescence, with the steepest decline occurring in infants under 2 years of age. Blood pressure increases through childhood and adult values are usually reached in adolescence.[5] The average normal values for heart rate, respiratory rate, and blood pressure are included in Table 17-1.

Table 17-1

Normal Vital Signs by Age

Age	Heart Rate (beats/min)	Respiratory Rate (breaths/min)	Blood Pressure (mm Hg) (90th percentile BP for 50th percentile height)	
			Boys	Girls
Newborn	120–170	30–80	87/68	76/68
1 year	80–160	20–40	98/53	100/58
3 years	80–120	20–30	105/61	103/62
6 years	75–115	16–22	110/70	107/69
10 years	70–110	16–20	115/75	115/74
17 years	60–110	12–20	133/83	125/80

CONGENITAL HEART DISEASE

Congenital heart disease (CHD) is the most common form of birth defect with a reported incidence of between 4 and 75 per 1000 live births.[6] Since Dr. Gross performed the first pediatric heart surgery in 1938, advances in surgical techniques, anesthesia management, the conduct of cardiopulmonary bypass, and perioperative medical management, especially neonatal critical care, have led to dramatic increases in survival for even the most complex lesions. Pediatric, and increasingly adult, anesthesiologists will encounter patients at various stages of palliation and/or repair. In this section, we present the most commonly encountered lesions and their physiological implications.

Left-to-Right Shunts

Left-to-right shunts are the result of an aberrant anatomical connection between the systemic and pulmonary circulations. This group represents the most common group of congenital cardiac anomalies and encompasses a number of lesions including patent ductus arteriosus (PDA), atrial septal defect (ASD), ventricular septal defect (VSD), atrioventricular canal (AVC) defects, double-outlet right ventricle (DORV), aortopulmonary (AP) window, and partial anomalous pulmonary venous connection (PAPVC). Despite the various locations of the shunts and potential sidedness of cardiac structures, left-to-right shunts are defined by the physiologic direction of blood flow, that is, from the systemic to the pulmonary circulation. The magnitude of the shunt and its hemodynamic significance are secondary to the location and size of the shunt as well as the pressure gradient across the shunt and the relative compliances of the downstream chambers or resistances of the downstream vessels.

PDA

A PDA occurs when the ductus arteriosus fails to undergo normal anatomical closure. The estimated incidence is 1 per 500 to 2000 live births, which represents roughly 5% to 10% of all CHDs.[7] There is an increasing incidence of PDA with decreasing gestational age, likely reflecting a developmentally immature response to the changing flow dynamics and biochemical milieu of the transitional circulation. In term neonates, most cases of PDA are sporadic; however, genetic disorders and environmental factors such as high altitude, neonatal sepsis, and in utero exposures (eg, rubella, phenytoin, alcohol, amphetamine) have all been implicated as predisposing factors.[7] Because systemic vascular resistance (SVR) is generally greater than

pulmonary vascular resistance (PVR), blood is primarily shunted from the descending aorta through the PDA into the pulmonary circulation with the degree of shunting determined by the dimensions of the duct and the relative difference in PVR and SVR. Increased pulmonary artery blood flow leads to pulmonary "overcirculation." Lung compliance is decreased and over time, microvascular injury leads to small vessel pulmonary vascular occlusive disease (PVOD; intimal proliferation, arteriolar medial hypertrophy) and pulmonary artery hypertension (PAH). As the pulmonary circulation is overperfused, the systemic circulation is underperfused, especially during diastole. The left heart, meanwhile, experiences greater blood return via the pulmonary veins causing left-sided volume overload—left atrial enlargement leads to arrhythmias, increased left ventricular end-diastolic pressure leads to hypertrophy, and increased catecholamine levels lead to tachycardia, increased oxygen demand, decreased diastolic time, and worsened coronary artery steal. Neonates with PDA may present with signs of pulmonary overcirculation (eg, failure to extubate, respiratory distress syndrome, pulmonary hemorrhage) or poor systemic perfusion (eg, necrotizing enterocolitis, renal insufficiency, intraventricular hemorrhage). Infants and children may present with failure to thrive, exercise intolerance, and/or recurrent pneumonias, while adults may present with the same symptoms or with more advanced sequalae such as atrial arrhythmias, infective endocarditis, pulmonary artery hypertension with Eisenmenger physiology, and/or congestive heart failure (CHF).

In the neonate, conservative PDA management consists of protective lung ventilation, permissive hypercapnia, fluid restriction ± diuretics, and inotropic support ± afterload reducing agents. Definitive treatment for PDA is ductal closure approached with pharmacological (nonsteroidal anti-inflammatory drugs [NSAIDs] or acetaminophen), catheter-based devices, and/or surgical intervention. No consensus exists among providers at United States children hospitals regarding the use of indomethacin prophylaxis or NSAID and/or surgical PDA treatment; practice preferences vary between and within hospitals.[8] The relative effectiveness of the various treatment options and their risks, both short- and long-term, has been the subject of numerous studies and Cochrane reviews.[9–18]

ASD

ASDs are common, accounting for 7% to 10% of CHD. There are four types of atrial level shunts:

1. Primum ASD (ASD1), caused by failure of the septum primum to fuse with the developing endocardial cushions;
2. Secundum ASD (ASD2), caused by a relative deficiency of the septum primum following tissue resorption in the region of the ostium secundum;
3. Sinus venosus defect, an unroofed pulmonary vein at the junction of the right atrium (RA) with either the superior vena cava (SVC) or the inferior vena cava (IVC); and
4. Unroofed coronary sinus, the absence of the wall between the coronary sinus and the left ventricle.

ASD2 is the most common type, representing 50% to 70% of atrial-level shunts. Shunt magnitude is determined by the size of the defect and the relative compliances of the ventricles. The left-to-right shunting becomes more prominent with increasing age as the compliance of the left ventricle (LV) decreases. Atrial level shunts cause right-sided volume overload and increased pulmonary blood flow. There is the potential for paradoxical emboli, cerebral abscess formation, and the development of pulmonary artery hypertension.

ASD2 with diameters <3 mm and ≥3 to 8 mm spontaneously close before 18 months of age in 100% and >80% of cases, respectively, while those with diameters ≥8 mm rarely do.[19] ASD1, sinus venosus defects, and unroofed coronary artery sinus do not spontaneously resolve. ASD closure is indicated in symptomatic patients without pulmonary hypertension and in asymptomatic patients when the shunt is large (pulmonary blood flow: systemic blood flow ratio [Q_p:Q_s] ≥ 1.5:1). This can be done in the cardiac catheterization lab provided that the anatomy is favorable and the patient is of adequate size, or in the operating room with cardiopulmonary bypass, generally around 2 years of age. Postoperatively, these patients are at risk for atrial and nodal arrhythmias and more rarely, sick sinus syndrome.

VSD

Isolated VSDs are the most common congenital heart lesions, accounting for 15% to 20% of all CHDs, or up to 50%, if VSD in setting of complex CHD is included. There are four types of VSDs:

1. Membranous (aka perimembranous, subaortic, conoventricular);

2. Subpulmonic (aka infundibular, conoseptal, outlet, supracristal, subarterial);
3. AV canal (aka inlet); and
4. Muscular (aka trabecular).

Membranous VSDs represent the large majority of VSDs and are frequently associated with a PDA and coarctation of the aorta (CoA), whereas subpulmonic VSDs may be associated with aortic regurgitation in the setting of aortic valve leaflet prolapse into the VSD. The amount of shunting is dependent upon the size and pressure gradient across the defect. In small to moderate shunts, the LA and LV experience volume overload while the RV is relatively spared. In large shunts, the RV is subjected to both increased volume and pressure load, while the LA and RV are volume overloaded to an even greater extent. The increase in Q_p is variable and dependent upon the size of the defect and PVR. Small VSDs may remain asymptomatic, while large VSDs may present with failure to thrive (FTT), recurrent lung infections, and signs of CHF. Heart failure and PVOD occur earlier than in patients with ASD. PVOD is accelerated in patients with trisomy 21. The reported rate of spontaneous closure varies widely, from 8% to 83%, depending on the location and size of the defect with the highest rates of spontaneous closure occurring during the first year of life and for small, muscular lesions.[20]

A Q_p:Q_s > 2:1 is an indication for VSD closure. The timing of repair depends on the severity of symptoms. Asymptomatic patients typically undergo surgical repair between 2 and 4 years of age. Larger patients with favorable anatomy may be candidates for transcatheter device closure. During the first year of life, symptomatic VSDs are managed with diuretics, afterload reducers, and/or digoxin. Those that respond to medical management undergo repair at 12 to 24 months, while those that are unresponsive to treatment and those with signs of rising PVR undergo semi-urgent repair regardless of age. If the surgical approach is dependent on a patient's size, pulmonary artery banding may be considered as a temporary palliation allowing for patient growth. Postoperative considerations include residual shunts and conduction abnormalities including right bundle branch block and complete heart block.

AVC Defects

AVC defects result from failure of the endocardial cushions to fuse with the developing atrial and ventricular septums. In a partial AVC defect, there is a primum ASD and a cleft in the anterior leaflet of the mitral valve (MV). Intermediate AVC defects are characterized by a primum ASD, an unrestrictive inlet VSD, and a divided common atrioventricular valve (AVV) with two distinct orifices. Transitional AVC defects consist of a primum ASD, a restrictive VSD, and a common AVV with single orifice. Finally, complete AVC defects (CAVCs) are composed of a primum ASD, a large unrestrictive VSD, and a common AVV with a single orifice. CAVCs can be further subdivided into three types according to the Rastelli classification based on valve morphology.[21] The presentation and clinical course of AVC defects depend on the magnitude and location of shunting (atrial, ventricular, AVV levels), the degree of ventricular imbalance and AVV regurgitation, as well as the presence of additional cardiac lesions.[22,23] In complete AVC, shunting at the level of the atria and ventricles leads to dilation of all four heart chambers, right greater than left. The degree of pulmonary blood flow is dependent on the relationship between PVR and SVR. Failure to thrive, recurrent pneumonias, and signs of CHF with or without pulmonary hypertension develop early in infancy in patients with CAVC and later with intermediate, transitional, and partial AVCs. Just as with isolated VSDs, PVOD occurs earlier and with greater severity in infants with trisomy 21. Airway obstruction at the level of the left mainstem bronchus may occur in the setting of severe left atrial dilation.

Primary surgical repair of CAVC is generally undertaken between 1 and 6 months of age. When CAVC is associated with other intracardiac lesions precluding repair at this age, pulmonary artery banding may be performed as a temporary palliation in order to limit pulmonary overcirculation and allow for patient growth. Postoperative considerations include residual shunts, AVV stenosis, and/or regurgitation, as well as sinoatrial and/or atrioventricular node conduction abnormalities. A total of 2.4% of patients status post CAVC repair require the placement of a permanent pacemaker.[24]

Obstructive Lesions

There are a number of congenital lesions that cause obstruction of flow to either the pulmonary or systemic circulations. Right-sided lesions include pulmonary stenosis (PS), pulmonary artery stenosis (PAS), tricuspid stenosis (TS), and double-chamber right ventricle (RV). Left-sided lesions include aortic stenosis (AS), coarctation of the aorta (CoA), interrupted aortic arch (IAA), hypoplastic left heart syndrome (HLHS), and mitral stenosis (MS). Given the relative rarity of many of these lesions, only the most common lesions will be discussed here.

Pulmonary Stenosis

Pulmonary stenosis may occur at the valvar, supravalvar, and/or subvalvar (infundibular) levels. Supravalvar PS is often associated with other cardiac and extra-cardiac anomalies and is seen in the setting of congenital rubella as well as several syndromes including Williams-Beuren, Alagille, and DiGeorge syndromes.[25] Subvalvar PS is generally accompanied by a large VSD as in tetralogy of Fallot. In valvar PS, the most common (about 90%) variant, the valve is thickened with fused or absent commisures and there is post-stenotic dilation of the main pulmonary artery (MPA). A variable degree of RV hypertension and hypertrophy with diastolic dysfunction will develop depending on the degree of stenosis present, although the RV may be hypoplastic in neonates with critical PS. Mild stenosis is typically not progressive, however, moderate and severe stenosis generally are. Neonates with critical PS will be maintained on IV prostaglandins in order to maintain pulmonary blood flow via the ductus arteriosus until a balloon valvotomy can be performed in the cardiac catheterization lab. Echocardiographic evidence of moderate or severe disease as well as symptoms of inadequate pulmonary blood flow such as angina, syncope, or pre-syncope regardless of Doppler gradient are also indications for pulmonary valvotomy. Surgery for isolated PS is rarely performed.[26] Long-term considerations in patients who have undergone valvotomy include re-stenosis and/or pulmonary regurgitation.

Aortic Stenosis

Left ventricular outflow tract obstruction (LVOTO) accounts for roughly 10% of CHD. As in PS, AS may occur at the valvar, supravalvar, and/or subvalvar levels. Valvar AS is most frequently caused by a bicuspid aortic valve, although a unicuspid valve is also possible.[27] In critical neonatal AS, the valve is myxomatous with a pinhole opening and the left-sided structures are varyingly hypoplastic.[28] Supravalvar aortic stenosis occurs at the upper margin of the sinuses of Valsalva and is frequently associated with Williams syndrome.[29,30] Subvalvar stenosis may be either tunnel-like or discrete. Discrete lesions such as a simple membrane or fibromuscular ridge are more common, and are in the majority of cases associated with other cardiac defects.[31] The LV is subjected to an increased pressure load leading to LV hypertrophy, the degree of which is dependent upon the severity of stenosis. Aortic root dilation and aortic valve regurgitation may develop in valvar and subvalvar stenosis, respectively. Neonates with critical AS will be maintained on IV prostaglandins in order to maintain systemic blood flow via the ductus arteriosus until a balloon valvotomy can be performed in the cardiac catheterization lab. Balloon valvuloplasty is the primary treatment of choice for isolated valvar AS beyond the neonatal period, provided that there are adequate annular dimensions. Valvar AS with annular hypoplasia, valvar AS with aortic regurgitation (AR) status post balloon dilation, subvalvar AS, and supravalvar AS are all managed surgically. Long-term considerations for these patients include the development of AR, mitral regurgitation (MR), and recurrent AS (especially in subvalvar AS).

CoA

CoA represents 8% to 10% of CHDs, is associated with other lesions (most commonly bicuspid aortic valve), and is commonly seen in association with Turner and Williams syndromes. Less commonly, it may be acquired, as with Takayasu arteritis. Anatomically, there is segmental juxta-ductal stenosis of the descending thoracic aorta. Histologically, there is abnormal smooth muscle cell (SMC) migration and differentiation, such that the subendothelial layer more closely resembles that of the ductus arteriosus than normal aortic tissue.[32] Patients with CoA may be symptomatic or asymptomatic. With ductal closure, neonates and infants with severe CoA will present with signs of CHF (tachypnea, sweating, irritability), signs of poor perfusion (necrotizing enterocolitis [NEC], acute kidney injury [AKI], hepatic failure, seizures), and/or cardiogenic shock. In conjunction with medical optimization, including reopening the ductus arteriosus with prostaglandins, urgent surgical (or less commonly interventional) treatment is indicated in these patients. Less severe coarctation may remain largely asymptomatic throughout childhood and adolescence with patients complaining of rare leg pain and/or mild exercise intolerance. On exam these patients may be hypertensive with an upper-to-lower extremity blood pressure gradient, although the gradient is not a reliable indicator of the severity of stenosis in the setting of collateralization. Their chest x-ray may be notable for the stereotypical findings of a “figure 3 sign” (pre- and post-stenotic dilation of the thoracic aorta) and/or Roesler’s sign (inferior notching of the 3rd-9th posterior ribs). Symptoms unresponsive to medical management, aortic diameter loss of more than 50%, and/or trans-CoA gradient more than 20 to 30 mm Hg are indications for interventional or surgical management. Although interventional management is the gold standard for recoarctation, its use for primary intervention remains controversial as the risk of short-term coarctation is higher.[33,34] Although CoA is anatomically a seemingly simple lesion, patients have life-long cardiovascular sequalae secondary to diastolic dysfunction and abnormal aortic elasticity predisposing them to hypertension, coronary artery disease, cerebral vascular accidents, CHF, and ruptured aortic and/or cerebral aneurysms. Additionally, these patients should be regularly screened for the development of recoarctation, AS (valvar and subvalvar), and MR.

Transposition of the Great Arteries

In transposition of the great arteries (TGA), there is discordance of the ventriculoarterial (VA) connections. There are two main types, denoted as dextro-TGA (*d*-TGA) and levo-TGA (*l*-TGA), so named for their ventricular looping patterns, with *d*-TGA being the more common anatomic form.

d-TGA

In *d*-TGA, the systemic and pulmonary systems are arranged in parallel—systemic blood returns from the systemic venous circulation and enters the right atrium and then passes through the tricuspid valve into the right ventricle which then pumps it into the aorta. Pulmonary venous blood is returned to the left atrium where it passes through the mitral valve into the left ventricle and then pumped back into the pulmonary circulation. This arrangement is uniformly fatal if there is not a communication (ASD, VSD, PDA) at which intracirculatory mixing can occur. *d*-TGA is the most common cause of cyanotic heart disease in the neonatal period and most neonates with *d*-TGA will present with cyanosis. Those with inadequate mixing will present with severe hypoxemia, progressive acidosis, and ultimately cardiovascular collapse. Prostaglandins are started at birth or at the time of diagnosis in order to maintain ductal patency; pulmonary blood flow is maintained with volume repletion and the maintenance of normal cardiac function. If an unacceptable degree of hypoxemia persists, a balloon atrial septostomy can be performed to create a second, unrestrictive communication.[35] Historically, *d*-TGA was palliated with an atrial level switch (Senning, Mustard); however, the current surgical approach is full anatomic repair via the arterial switch operation (ASO) performed during the neonatal period.[36–38] Long-term outcomes following aortic switch are favorable with neoaortic root dilation, neoaortic valve regurgitation, and coronary artery anomalies being the most common late complications.[39–41]

l-TGA

In *l*-TGA (also known as congenitally corrected or cc-TGA), there is both ventriculoarterial (VA) and atrioventricular (AV) discordance which result in a series circulation, albeit with a systemic RV. Blood returns from the systemic venous circulation and enters the right atrium and then passes through the mitral valve into the left ventricle which then pumps it into the pulmonary artery. Pulmonary venous blood is returned to the left atrium where it passes through the tricuspid valve into the right ventricle and then pumped into the aorta. Patients with isolated *l*-TGA will be fully saturated and may not present until later in life when the systemic RV begins to fail. The overwhelming majority of *l*-TGA patients, however, will have concomitant lesions, most commonly VSD, multilevel LVOTO, tricuspid valve anomalies, and/or conduction abnormalities, which make presentation variable.[42] Traditionally, *l*-TGA was palliated with a physiological repair, that is, repair of the concomitant lesions but with maintenance of VA and AV discordance; however currently, an anatomic repair in which atrial and arterial switches (“double-switch procedure”) convert the LV to the systemic ventricle is favored.[43] Long-term considerations in these patients include progressive AV conduction disturbances, arrhythmias, baffle obstruction, tricuspid valve regurgitation, and residual VSDs.

Cyanotic CHD

Cyanotic CHD involves right-to-left shunting of deoxygenated blood into the systemic circulation. There are three groups of lesions as follows:

1. Those with decreased pulmonary blood flow—tetralogy of Fallot (TOF), pulmonary atresia with intact ventricular septum, critical PS, and tricuspid valve abnormalities (tricuspid atresia, tricuspid stenosis with hypoplastic RV, Ebstein anomaly);
2. Those with increased pulmonary blood flow—truncus arteriosus, *d*-TGA, and total anomalous pulmonary venous return (TAPVR); and
3. Those associated with severe heart failure—HLHS, critical AS, critical CoA, and IAA.

TOF

Tetralogy of Fallot is comprised of the following four lesions:

1. A large, non-restrictive VSD,
2. RV outflow tract obstruction (RVOTO),
3. RV hypertrophy, and

4. An overriding aorta.

The pulmonary arteries may be hypoplastic. TOF accounts for roughly 10% of congenital heart disease and is the most common cause of cyanotic heart disease beyond the neonatal period. Several anatomic variants exist (TOF with PS, TOF with pulmonary atresia and major aortopulmonary collaterals, and TOF with absent pulmonary valve) with TOF with PA being the most common. For the sake of simplicity, only TOF with PS will be discussed further here.

Patients with TOF will have variable presentations depending on the anatomic level (infundibular, valvar, or both), severity and nature (dynamic, fixed, or both) of RVOTO. In patients with minimal RVOTO (acyanotic or "pink" TOF), the shunt across the VSD will be primarily left-to-right and the presentation will be similar to that of a large VSD with signs of congestive heart failure and a normal saturation. In patients with more significant RVOTO (cyanotic or "blue" TOF), the shunt will be bidirectional or primarily right-to-left and there will be some degree of cyanosis at baseline. During infancy, some patients with TOF will experience hypoxic (aka hypercyanotic or "tet") spells that are characterized by uncontrollable irritability, rapid shallow breathing, and severe cyanosis. If left untreated, these spells can progress to syncope, seizures, CVA, or death. They are most commonly seen in the early morning but can also be seen before/after eating and/or be precipitated by noxious stimuli, dehydration, prolonged episodes of agitation, or in relation to things which are known to decrease SVR (bathing, exercise, fever). Pathophysiologically, decreased SVR and/or infundibular spasm lead to increased right-to-left shunting across the VSD, which in turn leads to hypoxemia, hypercarbia, and acidosis. Activation of the respiratory center of the brain leads to hyperpnea. The altered breathing pattern, in turn, increases systemic venous return, further increasing the shunt volume and perpetuating the feed-forward loop. Treatment includes increasing SVR via femoral artery compression or holding the infant in a knee-to-chest position along with the administration of opioids (SC, IM, IV), vasopressors (IV), sodium bicarbonate, and supplemental oxygen. If unsuccessful, ketamine and beta-blockade can also be considered. TOF repair is generally performed in the first year of life, preferably after 3 to 4 months of age. The complete surgical repair entails VSD closure, and RV outflow tract reconstruction (valve-sparing vs transannular patch). Intervention in the neonatal period is reserved for those neonates with severe cyanosis or uncontrollable hypoxic spells and may either be palliative (RVOT stent vs surgical shunt) or a complete repair.[44–46] Post-repair considerations in these patients include residual lesions (VSD, PA stenosis, pulmonary regurgitation), residual RVOTO, arrhythmias, and conduction abnormalities.

HLHS

HLHS is a rare form of CHD characterized by a hypoplastic LV, mitral and aortic stenosis or atresia, and a hypoplastic aortic root and ascending aorta. The RV is functionally a single ventricle that must supply both the pulmonary and systemic circulations. Although a small amount of blood may pass anterograde through the left heart in the case of stenotic left-sided valves, the majority of pulmonary venous return will shunt left-to-right across an ASD; the flow to the coronary circulation and the head and neck vessels is supplied in a retrograde fashion by right-to-left shunting across the PDA. Without intervention, ductal closure will lead to metabolic acidosis and cardiovascular collapse. Neonates with HLHS are maintained on prostaglandin infusions in order to prevent this from happening. As PVR drops during the first few days of life, the single RV will provide progressively more blood flow to the lungs and less to the body. Balancing the perfusion to the lungs and the body requires maintenance of cardiac output and avoidance of excessive drops in PVR or increases in SVR.

The complete repair of HLHS is comprised of three stages:

1. There are two main palliative techniques for neonates with HLHS—the Stage 1 palliation (Norwood) and the hybrid approach. The Stage 1 palliation is composed of an atrial septostomy (BAS), main pulmonary artery ligation, PDA ligation, creation of a neoaortic root comprised of the proximal PA and the hypoplastic aortic root and ascending aorta, and establishment of a stable source of pulmonary blood flow with either a modified Blalock-Taussig shunt (mBTS) or an RV-PA conduit (Sano). In the hybrid approach, done without cardiopulmonary bypass, the PDA is stented, bilateral PA bands are placed, and a BAS is performed to prevent left atrial hypertension; the aorta is reconstructed during the Stage 2 palliation. Physiologically, the circulation is relatively unchanged after this palliation, with Q_p:Q_s dependent upon the ratio of PVR to SVR.

2. During the Stage 2 palliation, the bidirectional Glenn (BDG) procedure, the superior vena cava (SVC) is attached end-to-side to the ipsilateral PA and the mBTS or Sano conduit is taken down so that the entirety of PA blood blow is provided by venous return via the SVC. The single ventricle is partially volume off-loaded and the pulmonary blood flow is then dependent upon PVR, intrathoracic pressure, and the dynamics of the cerebral circulation.

3. In the third and final stage, the Fontan procedure, the inferior vena cava (IVC) blood flow is redirected to the ipsilateral PA, creating a single

ventricle series circulation. A small fenestration between the systemic venous blood flow and the common atrium may be created at the time of the surgery in order to serve as a pop-off valve in the setting of systemic venous hypertension, preserving cardiac output at the expense of cyanosis. This fenestration has been shown to decrease immediate postoperative morbidity and shorten intensive care unit (ICU) stay.[47] It may close spontaneously or be closed at a later time with a mechanical device in the catheterization laboratory.

The Fontan circulation is inherently inefficient. It is a single ventricle series circulation with near-normal oxygen levels, but with both inherently decreased ventricular preload and increased systemic vascular resistance (SVR).[48] Ventricular preload is limited by the low-pressure, nonpulsatile driving pressure through the lungs as well as the limited recruitability of the pulmonary vascular bed. The elevation in SVR is due to the fact that the single ventricle must pump blood through three resistance beds (systemic vascular bed, cavopulmonary system, and the pulmonary vascular bed) as compared to the one resistance bed encountered by each ventricle in a biventricular circulation. The altered ventricular loading conditions result in the decreased mechanical efficiency of the Fontan circulation. Cardiac index (CI) and fractional shortening (FS) are lower both at rest and during stress in patients with Fontan physiology as compared with healthy controls.[48] Although the majority of contemporary Fontan patients have a normal ejection fraction (EF), only around a third have normal diastolic function.[49] Fontan patients have obligatory systemic venous hypertension and diminished venous capacitance at baseline. Pulmonary artery flow (ie, systemic venous return) is more dependent upon respiratory mechanics (negative inspiratory pressure and downward displacement of the diaphragm) than the healthy biventricular patient (30% vs 15% contribution) due to the absence of a subpulmonary ventricle and normal interventricular interaction.[48] This lack of interventricular dependence is the reason why Fontan patients do not develop pulsus paradoxus in the setting of cardiac tamponade. The altered hemodynamics of the Fontan circulation predispose these patients to end-organ dysfunction (renal failure, Fontan-associated liver disease, plastic bronchitis, Fontan-associated protein-losing enteropathy), progressive cardiac failure, thrombosis, and increased functional impairment.[50]

HEART FAILURE: ETIOLOGY, PATHOPHYSIOLOGY, AND MANAGEMENT

Heart failure (HF) in the pediatric population is a major public health concern. Although pediatric HF is uncommon compared to the adult population, an increasing number of these patients are reaching adulthood, secondary to the successes in the medical and surgical management of HF. Children whose hospitalization is complicated by HF have a more than 20-fold increase in the risk of death.[51] The most common cause of HF in the United States is congenital heart disease (CHD), while primary cardiomyopathies are the most common cause of HF in children with structurally normal hearts. The incidence of HF in children with CHD is 6% to 24%.[52] The incidence of new-onset HF is 0.87 per 100,000 children under 16 year of age with cardiomyopathies. The highest incidence occurred within the first year of life and more than half were due to dilated cardiomyopathy.[53]

HF is a clinical diagnosis with signs and symptoms that result from a structural or functional impairment of ventricular filling or ejection of blood. This could be secondary to ventricular dysfunction with or without volume or pressure overload. HF may lead to a combination of circulatory, neurohormonal, and molecular abnormalities. In children, these may manifest as poor growth, feeding difficulties, respiratory distress, exercise intolerance, and fatigue.[54,55] The cardiac causes of pediatric HF are listed in Table 17-2.

Table 17-2

Causes of Pediatric Heart Failure

Congenital cardiac malformations	Volume overload	Left to right shunting	Ventricular septal defect
			Patent ductus arteriosus
		AV or semilunar valve insufficiency	Aortic regurgitation in bicommissural aortic valve
			Pulmonary regurgitation after tetralogy of Fallot repair
	Pressure overload	Left-sided obstruction	Severe aortic stenosis
			Aortic coarctation
		Right-sided obstruction	Severe pulmonary stenosis
	Complex CHD	Single ventricle	Hypoplastic left heart syndrome
			Unbalanced AV septal defect
		Systemic right ventricle	L-transposition of the great arteries
Structurally normal heart	Primary cardiomyopathy	Dilated	
		Hypertrophic	
		Restrictive	
	Secondary cardiomyopathy	Arrhythmogenic	-
		Ischemic	
		Toxic	
		Infiltrative	
		Infectious	

Cardiomyopathies in children are characterized by functional abnormalities of cardiac muscle in the absence of coronary, valvular, or congenital heart disease. The most common causes are idiopathic, familial, metabolic, or toxic. Pediatric cardiomyopathy may also be associated with metabolic, myopathic, hematologic, or neurologic diseases. For example, Duchenne's and Becker's muscular dystrophy are associated with high rates of cardiomyopathy.[56]

Determining the severity of HF is important in monitoring disease progression. This presents challenges in the pediatric population. The New York Heart Association (NYHA) Heart Failure Classification, which is widely used in adults, uses functional limitation to quantify severity and may only be useful in adolescents. Thus, the Ross Classification was created and recently modified for the assessment of infants in HF. It incorporates feeding difficulties, growth problems, and symptoms of exercise intolerance into a numeric score comparable with the NYHA classification (Table 17-3).[52]

Table 17-3

Modified Ross Heart Failure Classification for Children

Class I	Asymptomatic
Class II	Mild tachypnea or diaphoresis with feeding in infants
	Dyspnea on exertion in older children
Class III	Marked tachypnea or diaphoresis with feeding in infants
	Marked dyspnea on exertion
	Prolonged feeding times with growth failure
Class IV	Symptoms such as tachypnea, retractions, grunting, or diaphoresis at rest

The diagnosis of HF in children is based on a combination of clinical, radiographic, echocardiographic, and laboratory findings. These are discussed in Table 17-4.[56,57]

Table 17-4

Modalities Used in the Preoperative Assessment of Children with Heart Failure

Modality	Findings	Additional Notes
Chest X-ray	Cardiomegaly, pulmonary edema/effusions	-
Electrocardiogram	ST segment or T wave changes, ventricular hypertrophy, axis deviations, bundle branch or AV block, arrhythmias	-
Echocardiography	Ventricular dilation, systolic dysfunction, AV valve regurgitation, pericardial effusion	-
Point of care ultrasound (U/S)	B-lines on lung U/S, diminished EF	
Laboratory studies	Elevated BNP, NT-BNP	May be useful to monitor trends
	Elevated troponin	May be elevated in myocarditis
	Elevated transaminases, creatinine	May indicate poor end-organ perfusion
Endomyocardial biopsy	Determines etiology of myocarditis	Examples: infectious, immune-mediated, storage diseases etc.

Abbreviations: AV:, atrioventricular; BNP:, B-type natriuretic peptide; EF, ejection fraction; NT-BNP, N-terminal probrain natriuretic peptide.

HF patients are managed based on the etiology of HF and the disease severity. It is important to understand the implications of the different modalities used in the management of these patients prior to providing anesthetic care. Many patients will be on several pharmacologic agents to provide

symptomatic relief of HF symptoms. These include diuretics, digoxin, angiotensin-converting enzyme (ACE) inhibitors, and angiotensin II receptor blockers (ARBs). Patients with severe, decompensated HF that are in an inpatient setting may present to the operating room on intravenous diuretics and/or inotropes such as dopamine and epinephrine, which are used to improve cardiac output. These patients may also present on intravenous milrinone, a phosphodiesterase III inhibitor, which increases contractility and reduces afterload. Patients in severe heart failure may no longer respond to pharmacologic therapies and may require positive pressure ventilation, mechanical circulatory support or ultimately, heart transplantation. Positive pressure ventilation can be effective in relieving respiratory distress from cardiogenic pulmonary edema as well as providing alveolar recruitment, improved lung compliance, and decreased left ventricular preload and afterload. This improves cardiac output. Some patients that do not respond to medical therapy will benefit from cardiac resynchronization therapy (CRT). These are usually patients with reduced EF (ie, <35%) and a left bundle branch block (LBBB) pattern on electrocardiogram (ECG). The intraventricular conduction delay or LBBB may worsen HF by causing ventricular dyssynchrony. CRT uses biventricular pacing to decrease ventricular dyssynchrony.[58] HF patients may also suffer from arrhythmias and sudden cardiac death. These patients may require an implantable cardioverter defibrillator (ICD) and management of these devices will be required with the assistance of an electrophysiology specialist in the perioperative period.

Children with decompensated HF and low cardiac output unresponsive to medical therapy could require mechanical circulatory support (MCS) to maintain end-organ function. MCS devices include extracorporeal membrane oxygenation (ECMO) or a ventricular assist device (VAD). MCS may be used as short-term support for reversible causes of HF, for long-term support as a bridge to cardiac transplantation, or less commonly in this population, as destination therapy. The choice of device depends on the size of the patient and the type of support required (cardiopulmonary vs cardiac). ECMO is capable of providing full cardiopulmonary support, while VADs are indicated for isolated ventricular dysfunction. ECMO may be used in the setting of imminent or actual cardiac arrest, during low cardiac output states, or electively in the setting of high-risk procedures.[59] Cannulation can be performed percutaneously, and ECMO can provide full cardiopulmonary support for days to weeks. Venous blood is drained from the patient and circulated through a membrane for gas exchange. Oxygenated blood is then returned to the patient through a large vein (venovenous or VV ECMO) or artery (venoarterial or VA ECMO). VV ECMO is used in patients that only require respiratory support, while VA ECMO can provide both respiratory and cardiac support. The goal of ECMO is to provide adequate gas exchange and oxygen delivery to allow time for the underlying disease process to resolve. The parameters that are monitored include the hemodynamics, acidosis, lactate, venous saturations, urine output, perfusion, and end-organ function. These patients commonly experience hypertension. The decrease in cardiac filling pressures from decompression of the atria by the ECMO circuit causes tachycardia and thus, hypertension. These patients may be treated with antihypertensives, but with caution as they can exacerbate the hypertension/tachycardia by further decreasing preload and filling pressures. Ventilation strategies should be aimed at lung protection and minimizing ventilator-associated lung injury.

VADs can offer either univentricular or biventricular support. Typically, for left ventricular support, blood is drained from the left atrium or left ventricular apex to the pump and returned to the ascending aorta. For right ventricular (RV) support, blood is drained from the right atrium or RV to the pump and returned to the main pulmonary artery. The choice of device depends on the patient's size, anticipated duration, goal of support, device availability, and diagnosis. Devices differ by flow design (pulsatile, centrifugal, or axial), pump location relative to patient (implantable, paracorporeal, or extracorporeal), and delivery system (percutaneous or central). The Berlin Heart EXCOR is the most popular pediatric long-term support device and is the only FDA-approved device for neonates and infants.[60]

There are a number of perioperative physiologic considerations for the patient managed with MCS. First, they are very sensitive to changes in venous and arterial capacitance and may require fluid and/or vasopressor support in the setting of anesthesia-induced vasodilation. Second, given the risk for thromboembolism, these patients are chronically anticoagulated. In the case of ECMO, unfractionated heparin is the most commonly used anticoagulant drug, although direct thrombin inhibitors such as bivalirudin or argatroban are used in the setting of heparin-induced thrombocytopenia (HIT) or heparin resistance. Patients with VADs are usually anticoagulated using heparin and transitioned to warfarin with the addition of antiplatelet agents for long-term anticoagulation. The decision to lower the level of anticoagulation prior to surgery should be a discussion between the anesthesiologist, surgeon, and physician managing the MCS. Third is pharmacokinetics, which is concerned with how the patient's body processes drugs—absorption, distribution, metabolism, and excretion. Fourth, MSC support may alter the pharmacodynamics of administered drugs due to altered clearance, sequestration of drugs in the circuitry components, and an increase in volume of distribution. In patients supported with ECMO, lipophilic and protein-bound drugs are more likely to be sequestered.[61–66] Lastly, the patient with a VAD should be considered a full stomach because of the placement of the device (upper abdomen) even when appropriately nil per os (NPO).

The complications related to MCS devices include bleeding, thromboembolism, seizures, and infection. Bleeding can occur at the site of the cannulas, at the surgical site, from gastrointestinal hemorrhage, in the pericardium, or in the brain. In addition, there are potential sources of mechanical

complications leading to pump failure. Due to these serious complications, weaning from support should be considered once the underlying disease process is reversed. However, prolonged support may be a bridge to transplantation.[67]

THE CARDIAC PATIENT FOR NONCARDIAC SURGERY: ANESTHETIC MANAGEMENT

A systematic approach to patients with CHD undergoing noncardiac procedures begins with an understanding of the anatomical and physiological implications of the patient's lesion and prior palliations/repairs as well as an appreciation of the patient's current functional status. It allows for the formulation of intraoperative hemodynamic and respiratory management goals, and directs planning for postoperative disposition. One such approach includes the following seven steps: preoperative assessment, endocarditis prophylaxis, prevention of paradoxical embolization, monitoring devices and intravenous access special considerations, fluid management, hemodynamic and respiratory management, and preoperative planning and postoperative disposition.

Preoperative Assessment

A thorough preoperative assessment includes an understanding of the anatomy and pathophysiology of the underlying defect or disease as well as the evaluation of the current functional status and medications (Table 17-5).[68] Pediatric heart disease is often associated with genetic syndromes that have anesthetic implications of their own, as is shown in Table 17-6.[69] Performance of age-appropriate activities is a surrogate of patient's cardiac function and reserve. Physical examination may reveal cyanosis, clubbing, and signs of congestive heart failure (tachypnea, hepatomegaly, ascites, and edema). Access and airway need to be carefully evaluated. Patients with CHD may have congenital and/or acquired airway abnormalities. They may also have limited venous and arterial access due to multiple palliative procedures. While evaluating a pediatric CHD patient, the anesthesiologist must be mindful of the age of the patient and the impact of multiple previous surgical and anesthetic experiences on the patient and the family.

Table 17-5

Preoperative Evaluation

History	Signs and symptoms of congestive heart failure (eg, failure to thrive) Palpitations or syncope Additional congenital anomalies (eg, airway, genitourinary) Recent and current medications (eg, diuretics, digoxin, ACE inhibitors) Past surgical and intervwentional history Last follow-up
Physical examination	Heart murmur, thrill, arrhythmias Tachypnea, increased work of breathing, rales Poor peripheral perfusion, delayed capillary refill, bounding or diminished pulses Cool extremities, mottled skin, sweating. Hepatomegaly Edema.
Laboratory studies	CBC: erythrocytosis (secondary to cyanosis), anemia (secondary to malnutrition, iron deficiency) Electrolytes: hypokalemia, hyponatremia (secondary to diuretic therapy) Coagulation profile
Additional tests	Echocardiography: function and anatomy ECG: rhythm, signs of atrial or ventricular hypertrophy Chest X-ray: cardiomegaly, pulmonary edema, or infiltrates
Specific studies	Cardiac catheterization: anatomy, pressure gradients, saturations, shunts, resistances Cardiac MRI: anatomy, pulmonary blood flow, RV and LV function

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Table 17-6

Common Syndromes, Associated Congenital Heart Diseases, and Anesthetic Implications

Syndrome	Commonly Associated CHD	Other Anesthetic Implications
CHARGE association (**C**oloboma, congenital **H**eart defects, choanal **A**tresia, **r**enal abnormalities, **G**enital hypoplasia, **E**ar deformities)	65% conotruncal anomalies, aortic arch anomalies	Difficult airway and intubation, renal dysfunction
DiGeorge syndrome (chromosome 22 deletion, catch 22)	Interrupted aortic arch, truncus arteriosus, VSD, PDA, TOF	Hypocalcemia, immunodeficiency, need for irradiated blood products
Duchenne muscular dystrophy	Cardiomyopathy	Hyperkalemic cardiac arrest with succinylcholine,

		rhabdomyolysis with inhalational agents
Ehler-Danlos syndrome	Aneurysm of aorta and carotid artery	Difficult intravenous access, increased bleeding risk
Ellis-van Creveld syndrome (chondroectodermal dysplasia)	50% common atrium	Possible difficult intubation
Fetal alcohol syndrome	25%–30% VSD, PDA, ASD, TOF	Difficult airway, renal disease
Friedreich's ataxia	Cardiomyopathy	Progressive neurological degeneration, glucose intolerance
Glycogen storage disease II (Pompe)	Cardiomyopathy	
Holt-Oram syndrome	ASD, VSD	Upper limb abnormalities
Leopard syndrome (cardiocutaneous syndrome)	PS, long PR interval, cardiomyopathy	Growth retardation, possible difficult intubation
Long QT syndromes: • Jervell and Lange Nielsen • Romano-Ward	Long QT interval, ventricular tachyarrhythmia	Congenital deafness
Marfan syndrome	Aortic aneurysm, AR, and/or MR	Spontaneous pneumothorax, cervical spine instability
Mucopolysaccharidosis: • Hurler (type I) • Hunter (type II) • Morquio (type III)	AR and/or MR, coronary artery disease, cardiomyopathy	Difficult airway, atlantoaxial instability, kyphoscoliosis
Noonan syndrome	PS (dystrophic valve), LVH, septal hypertrophy	Possible difficult intubation, platelet dysfunction, renal dysfunction
Tuberous sclerosis	Myocardial rhabdomyoma	Seizure disorder, renal dysfunction
Shprintzen syndrome (velocardiofacial, 22q deletion)	Conotruncal anomalies, TOF	Difficult airway
VACTERL association	C=CHD, VSD, conotruncal anomalies (TOF, truncus arteriosus)	**V**ertebral anomalies, **A**nal atresia, **T**racheoesophageal fistula, **E**sophageal atresia, **R**enal anomalies, **L**imb defects
Williams syndrome	Supravalvular AS, PA stenosis	Developmental delay, difficult airway, renal dysfunction
Zellweger syndrome (cerebrohepatorenal syndrome)	PDA, VSD, ASD	Neonatal jaundice, kidney and liver dysfunction, coagulopathy, hypotonia

Endocarditis Prophylaxis

Based on the 2007 AHA guidelines, for all dental procedures involving manipulations of gingival tissue or oral mucosa, endocarditis prophylaxis is recommended for patients with the following conditions:[70]

- Prosthetic cardiac valve or prosthetic material used for cardiac valve repair.
- Previous endocarditis.
- Congenital heart disease (CHD):
  - Unrepaired cyanotic CHD, including palliative shunts and conduits.
  - Completely repaired CHD with prosthetic material or device, whether placed by surgery or catheter intervention, during the first 6 months after the repair.
  - Repaired CHD with residual defects in the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibits endothelialization).
- Cardiac transplantation recipients who develop cardiac valvulopathy.

Endocarditis prophylaxis is not recommended for routine gastrointestinal or genitourinary procedures.[70]

Prevention of Paradoxical Embolization

Multiple CHD lesions including simple (ASD, VSD, CAVC) or complex (single ventricle lesions) have an intracardiac communication at the atrial or ventricular levels and repaired cardiac lesions may have a fenestrated patch closure. These intracardiac communications can allow air or thrombotic material to migrate from the venous circulation to the arterial circulation to the brain, gastrointestinal tract, or kidneys. Hence, careful administration of fluid and medications and the use of in-line air filters to prevent air emboli are important to decrease the risk of paradoxical embolization.

Monitoring and Intravenous Access

Standard ASA monitoring including noninvasive blood pressure, electrocardiogram (ECG), and pulse oximetry is recommended for all patients. A 5-lead ECG is recommended in patients at risk of arrhythmia or coronary ischemia such as patients with aortopulmonary shunts or long QT syndrome. Invasive monitoring of blood pressure (arterial line) and central venous pressure should be considered based on the type of procedure (eg, spinal fusion) and patient's cardiac condition and functional status. The location of blood pressure monitoring also depends on the cardiac lesion and prior/planned interventions. Patients with aortic coarctation will have higher blood pressure in the upper extremities compared to lower extremities. Patients with history of a classic BT shunt (subclavian to PA end-to-side anastomosis) may not have a measurable blood pressure in the ipsilateral arm while those with a current modified Blalock-Taussig (BT) shunt may have lower blood pressure on the ipsilateral side of the BT shunt due to steal. It is important to note that peripheral intravenous (IV) access may be difficult.

Management of Fluid Status

The NPO times are similar to other cases with 2 hours for clear fluids, 4 hours for breast milk, 6 hours for formula, and 6 to 8 hours for solid food. Some patients do not tolerate prolonged NPO times and preoperative hydration is required to prevent hemodynamic instability at induction. These patients include patients with Williams-Beuren syndrome, patients on chronic diuretic therapy, and preload-dependent patients such as those with bidirectional Glenn or Fontan physiology. In addition, dehydration in cyanotic patients may worsen blood hyperviscosity leading to thrombosis. If prehydration is not possible, a fluid bolus 5 to 10 mL/kg following induction and IV placement is recommended. Fluid administration should be judicious in patients with pulmonary vein stenosis, mitral stenosis, or diastolic dysfunction to avoid the development of pulmonary edema.

Hemodynamic and Respiratory Management

The anesthesiologist needs to understand the underlying pathophysiology, the hemodynamic goals, and potential risks of hemodynamic instability inherent to a patient's underlying CHD. Beta-blockers are usually continued prior to the procedure while ACE inhibitors are withheld for 24 hours prior to the procedure, and diuretics are either held or continued depending on the nature of the procedure and its intended fluid requirements.

Premedication is individualized based on the patient's and the family's prior experience. Midazolam 1 mg/kg (maximum 20 mg) combined with oral ketamine (3–5 mg/kg for infants and 5–10 mg/kg for children) can be used as an oral premedication. In children who cannot take oral premedication, intramuscular premedication is an option.

In addition to the routine anesthesia setup, it is important to be ready for hemodynamic instability and discuss the intraoperative management with the patient's cardiologist and surgeon. If the patient is at risk of arrhythmia, appropriate antiarrhythmic medications and equipment such as a defibrillator and pacing equipment need to be available. Pacemakers and ICDs should be interrogated prior to the procedure to ensure that they are properly functioning and to inform intraoperative device management. For example, pacemaker-dependent patients undergoing procedures where electrocautery will be used will likely require a setting change to an asynchronous pacing mode in order to ensure continuous pacing, while those with an ICD will require the ICD functionality to be suspended in order to prevent inappropriate shocks. If the patient's functional status requires inotropic support during the procedure, dopamine and epinephrine should be readily available. Table 17-7 lists the most common inotropic and antiarrhythmic drugs.

Table 17-7

Commonly Used Vasoactive Drugs and Antiarrhythmics

Drugs	Bolus	Infusion Rate	Comments
Adenosine 6 mg in 2 mL (3 mg mL^{-1})	100 µg kg^{-1} rapid IV bolus and flush (maximum 6 mg); second dose 200 µg kg^{-1} (max. 12 mg)		Reduce by half for patients who have had a heart transplant. Give as close to IV site followed by a flush
Atropine 8 mg in 20 mL (0.4 mg mL^{-1})	20 µcg kg^{-1} IV		Maximum dose 1 mg for child and 3 mg for adolescent
Amiodarone 150 mg in 3 mL (50 mg mL^{-1})	5 mg kg^{-1} IV slowly over 15–30 min	5–15 µg kg–1 min–1	Adult max. bolus 300 mg for Vfib and/or VTach
Calcium gluconate 100 mg mL^{-1}	30–60 mg kg^{-1} IV		
Dopamine 400 mg in 10 mL (40 mg mL^{-1})		3–10 µg kg–1 min–1	Titrate to effect
Epinephrine 1 mg mL^{-1}	1 µg kg^{-1} to treat hypotension IV; 10 µg kg^{-1} IV for cardiac arrest; repeat every 3–5 min as	0.02–0.1 µg kg–1	Titrate to effect

	needed	min−1	
Isoproterenol 1 mg in 5 mL (0.2 mg mL^{-1})		0.01–0.1 µg kg^{-1} min^{-1}	Tachycardia, palpitations, angina, pulmonary edema, hypertension, hypotension, ventricular arrhythmias, tachyarrhythmias
Lidocaine 20 mg in 2 mL (10 mg mL^{-1})	1 mg kg^{-1} IV	20–50 µg kg^{-1} min^{-1}	
Magnesium 50% 1 in 2 mL (0.5 g mL^{-1})	25–50 mg kg^{-1} IV		For torsades de pointes (max. 2 g)
Norepinephrine 4 mg in 4 mL (1 mg mL^{-1})		0.05 µg kg^{-1} min^{-1}	Titrate to effect
Phenylephrine 10 mg mL^{-1} vial	0.5 µg kg^{-1}	0.1 µg kg^{-1} min^{-1}	Titrate to effect
Procainamide 500 mg mL^{-1} 100 mg mL^{-1}	5–15 mg kg^{-1} IV loading dose over 30–60 min	20–80 µg kg^{-1} min^{-1}	ECG monitoring required Caution: hypotension and prolonged QT, widening of QRS
Vasopressin 20 units mL^{-1}	In children 0.1 unit In adults 1 unit	6–30 mU kg^{-1} min^{-1}	Titrate to effect
Ephedrine 5 mg mL^{-1}	0.05–0.1 mg kg^{-1}		

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Preoperative Planning and Postoperative Disposition

Planning on the timing, location (surgery center, satellite hospital, tertiary main hospital campus), and staffing is an important component of preoperative planning. The patient should be medically optimized, NPO times should be minimized, and the location and staff capable of providing the necessary resources and expertise are required. The ability to escalate care and obtain cardiology support should always be considered even for seemingly simple procedures. Based on the patient's cardiac condition and planned procedure (diagnostic imaging, spine, dental rehabilitation), the postoperative course can range from recovery in the post-anesthesia care unit with discharge home, observation for 24 hours, admission as inpatient to the cardiac floor, to admission to the cardiac intensive care unit.

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