## Chapter 3

# Congenital Heart Disease

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*This chapter will focus on the anesthetic management of children with congenital heart disease for noncardiac procedures, and will focus on the implications of some of the common congenital heart disorders.*

Approximately nine of every 1,000 children are born with some form of congenital heart disease (CHD), which is usually divided into two types: cyanotic (contains a right-to-left shunt) and acyanotic (may contain a left-to-right shunt). The most common cyanotic lesions, in order of decreasing frequency are pulmonary stenosis (PS), transposition of the great arteries (TGA), tetralogy of Fallot (ToF), tricuspid atresia (TA), and pulmonary atresia with intact ventricular septum (PA/IVS). The most common acyanotic lesions, in order of descending frequency, are ventricular septal defect (VSD), atrial septal defect (ASD), aortic stenosis (AS), coarctation of the aorta (CoA), persistent ductus arteriosus (PDA), and complete common atrioventricular canal (CCAVC). A lesion that requires surgery or catheter-based intervention in the first year of life is called ***critical*** CHD.

### Pathophysiology of Congenital Heart Disease

Anesthesiologists that care for children with CHD should fully understand the anatomical components of the lesion and how the blood flows through the heart and lungs. Because of the complexity of the lesions and subsequent repairs, this can often be confusing. Therefore, a structured approach should be used, with a focus on determining the relative ratios of pulmonary and systemic blood flow. These ratios may ultimately determine the important aspects of anesthetic management. This structured approach involves the following steps:

1. Determine if blood flow is obstructed in any part of the heart. Right-sided heart obstructions decrease blood flow to the lungs and result in low $PaO_2$. Left-sided heart obstructions decrease blood flow to the body, and result in decreased tissue perfusion, metabolic acidosis, and shock.
2. Determine if blood is being shunted from one side of the heart to the other. If blood is shunted from the right side to the left side (e.g., ToF), it doesn't have a chance to go through the lungs, resulting in cyanosis. Left-to-right sided shunting (e.g., VSD) will result in volume and pressure overload on either or both ventricles, and may lead to congestive heart failure. In its advanced form, overcirculation of the pulmonary bed leads to pulmonary hypertension, and if untreated, irreversible pulmonary vascular

obstructive disease. This results in a reversal of the shunt (to right to left) and causes hypoxemia and cyanosis (sometimes known as Eisenminger's syndrome). On a basic level, it may seem that the direction of shunting is determined by the location of the defect and obstruction. But, in many cases the resistance in the pulmonary and systemic circuits will determine the direction of shunt. Specialists in CHD like to refer to the ratio of pulmonary vascular resistance (PVR) to systemic vascular resistance (SVR). This ratio will determine whether the patient has a right-to-left shunt, a left-to-right shunt, or both at different times during the cardiac cycle.

3. Determine if there a volume load or pressure load on the heart. When a ventricle is overburdened by volume overload or obstruction to forward flow, the ventricle can begin to fail. In general, the right ventricle responds with dilatation, and the left ventricle develops hypertrophy. In either case, if the load is too much for the ventricle to function correctly, CHF results.

Once these three key points are determined, the anesthesiologist can begin to formulate a plan to safely anesthetize the child with CHD. In reality, the type of lesion will not greatly influence the choice of anesthetic drugs, but rather the approach to ventilation, circulation, and how the anesthesiologist will respond when the child becomes either hypoxic or hypotensive during the case. In the next section, we will take a closer look at the anatomy and physiology of some of the more common causes of CHD, starting with the acyanotic defects.

## Acyanotic CHD

### Ventricular Septal Defect (VSD)

The most common congenital heart defect, (approximately 25% of all congenital cardiac lesions) is the VSD. There are five different variations: a) ***muscular***, which occurs in the muscular portion of the septum, can occur in the posterior, apical or anterior portion, and be single or multiple; b) ***inlet***, which is found in the part of the septum underneath the septal leaflet of the tricuspid valve; c) ***conoseptal***, which is found in the outflow tract of the right ventricle beneath the pulmonary valve; d) ***conoventricular***, which is found in the membranous portion of the septum; and e) ***malalignment***, which results from a malalignment of the infundibular part of the septum. Does the type of VSD influence anesthetic management? Hardly. But we wanted you to at least hear about the different types in case you need to strike up a conversation with a pediatric cardiologist at a bar.

The clinical features of a VSD will be determined by its size and the direction of blood flow through it. If the VSD is relatively small, there are usually no clinical symptoms. When the VSD is large it won't restrict blood flowing through it, and the PVR to SVR ratio will determine the direction of flow. In almost all children, SVR is higher than PVR, and blood flows from left to right through the VSD. If untreated, over time this will result in CHF, but advanced forms result in pulmonary hypertension and reversal of the shunt. CHF can be treated with digoxin, diuretics, and an angiotensin-converting enzyme (ACE) inhibitor while waiting for natural or surgical closure. Small muscular and conoventricular VSDs close naturally (40% by age three years, 75% by age ten years); however, large VSDs

should be closed surgically before pulmonary vascular changes become irreversible. Children with a previous VSD repair may occasionally demonstrate myocardial dysfunction, arrhythmias, or right bundle branch block.

**Atrial Septal Defect (ASD)**

ASD accounts for about 7.5% of CHD. Of course, there are multiple types: a) ***ostium secundum***, which is located in the mid-portion of the atrial septum and is the most common form of ASD; b) ***ostium primum***, which is located low in the atrial septum; c) ***sinus venosus***, which is located at the junction of the right atrium and the SVC or IVC; d) ***coronary sinus***, which results when there is a hole in the wall of the coronary sinus as it traverses the left atrium; and e) ***patent foramen ovale (PFO)***, which results when there is inadequate fusion of the septum secundum and the septum primum. The specific type of ASD doesn't influence anesthetic management unless it causes physiologic abnormalities.

ASDs are usually asymptomatic, but complications of the unrepaired ASD may include paradoxical emboli, decompression sickness, migraine headaches, development of pulmonary hypertension, and an increased risk of meningitis. Nearly all small secundum ASDs close in the first year of life. But with large ASDs, significant left-to-right shunting will require surgical repair or placement of a closure device via cardiac catheterization. Ostium primum, sinus venosus, and coronary sinus ASDs do not close spontaneously and must be closed surgically. PFO's occur in approximately 17–35% of the general population. Children are usually asymptomatic after ASD repair, and there are no unique anesthetic considerations for noncardiac surgery.

**Complete Common Atrioventricular Canal (CCAVC)**

CCAVC (also sometimes known as an endocardial cushion defect) consists of an ostium primum ASD and an inlet VSD and often occurs in children with trisomy 21 (Down syndrome). There is usually a left-to-right shunt at the atrial and ventricular levels, and can result in CHF early in infancy. Pulmonary hypertension may develop from the increase in pulmonary blood flow.

CHF is treated with digoxin, diuretics, and an ACE inhibitor, and surgical repair is usually performed in the first year of life. Complete heart block occurs in 5% of patients undergoing repair, and residual mitral insufficiency may be seen.

**Patent Ductus Arteriosus (PDA)**

Prior to birth, blood bypasses the lungs and travels from the main pulmonary artery to the descending aorta through the ductus arteriosus. Normally, this ductus closes in the first days of life, but in certain conditions (most commonly, prematurity) it remains open as a PDA. PDA represents about 7.5% of congenital heart disease.

A PDA can be caused by a low oxygen level, high blood $PCO_2$, acidosis, and persistent pulmonary hypertension of the newborn. The direction of shunted blood through a relatively large PDA depends on the ratio of PVR to SVR. In a nonrestrictive PDA, a left-to-

right shunt occurs if SVR is greater than PVR. Newborns with a large PDA and left-to-right shunt may show signs of pulmonary overcirculation and CHF, which include a widened pulse pressure, a continuous murmur, and an inability to wean ventilatory parameters.

A PDA may be treated medically with indomethacin, but will be surgically repaired when it continues to interfere with cardiorespiratory function. Surgical ligation is performed by thoracotomy, video-assisted thoracoscopic surgery, or coil embolization. There are no unique anesthetic considerations in otherwise healthy infants or children that previously underwent PDA repair.

**Aortic Stenosis (AS)**

AS represents up to 5% of CHD. It may be relatively mild or there may be complete atresia as in hypoplastic left heart syndrome (see below). The neonate with critical AS must rely on their PDA for systemic blood flow; if the PDA closes, circulatory shock will occur. Most cases of AS are detected later in childhood by the presence of a murmur.

The clinical manifestations will depend on the degree of stenosis and the ventricular function. Significant stenosis produces a pressure gradient between the left ventricle and the aorta that results in left ventricular hypertrophy and, over time, decreased ventricular compliance and function.

Hemodynamically significant AS will require surgical intervention, which can be accomplished by open surgical valvotomy or balloon valvuloplasty. In some cases, treatment of AS causes aortic regurgitation, which may eventually require aortic valve replacement. In some children, a Ross procedure (pulmonary autograft) is performed, in which the child's own pulmonary valve is moved into the aortic position, a right ventricle to pulmonary artery homograft conduit is placed, and the coronary arteries are reimplanted. If you are reading this book to understand how to anesthetize a child for a Ross procedure, the patient is in grave danger.

**Coarctation of the Aorta (CoA)**

CoA represents about 8% of all congenital heart defects, of which approximately 80% also have a bicuspid aortic valve. It usually occurs distal to the origin of the left subclavian artery at the insertion site of the ductus arteriosus. The coarctation causes an obstruction in the aorta, and increases left ventricular afterload. CHF develops in about 10% of cases in infancy. There is a 15–20% risk of having CoA in girls with Turner syndrome (45, XO).

Neonates with severe CoA need to use their PDA to provide blood to the systemic circulation. If the PDA closes, the infant goes into circulatory shock. Therefore, PGE1 is administered to keep the ductus open until the CoA is repaired. More commonly, CoA presents during childhood. Typically it is diagnosed for investigation of a new heart murmur, accompanied by hypertension of the upper extremities and decreased or absent femoral pulses. Left ventricular hypertrophy and CHF can result from chronic pressure overload.

The CoA can be treated by balloon dilation angioplasty, stent placement, surgical end-to-end anastomosis, subclavian flap repair, patch repair, or graft placement. In many patients hypertension persists throughout childhood; the duration of postoperative hypertension correlates with the duration of hypertension prior to the repair.

## Cyanotic CHD

### Hypoplastic Left Heart Syndrome (HLHS)

HLHS is the second most common type of CHD that presents in the first week of life, and it is the most common cause of death from CHD in the first month of life. HLHS consists of hypoplasia of the left ventricle, aortic valve stenosis or atresia, mitral valve stenosis or atresia, and hypoplasia of the ascending aorta with a discrete CoA. The result is the lack of blood flow through the left heart, causing an obligatory left-to-right shunt at the atrial level and a right-to-left shunt through a PDA. Systemic flow becomes completely dependent on the PDA, and coronary perfusion is retrograde in the presence of aortic atresia or critical aortic stenosis. The diagnosis of HLHS is often made in the first few days of life, when the PDA closes and the infant presents in heart failure and shock. Clinical signs include tachycardia, tachypnea, pulmonary rales (from pulmonary edema), hepatomegaly, and poor peripheral pulses with diminished distal capillary refill. PGE1 is started immediately upon diagnosis (to keep the PDA open) and the infant is prepared for urgent palliative surgery.

In most centers the palliative treatment approach consists of three surgical procedures that occur over the first several years of life. The first procedure, which is performed in the first week of life, establishes unobstructed systemic blood flow and allows the majority of neonates to survive infancy. It consists of the creation of a “neoaorta”, which consists of an amalgamation of the pulmonary artery and existing aorta to provide systemic blood flow. In addition an atrial septectomy is performed to create an unobstructed atrial communication, and a modified Blalock–Taussig (BT) shunt (see below) or right ventricle-to-PA conduit to restrict pulmonary blood flow. This prevents pulmonary vasculature obstructive disease that may result from too much pulmonary blood flow.

The second stage is performed at 4–6 months of age and is called a hemi-Fontan or bidirectional Glenn procedure (see below). A connection is created between the SVC and the right PA, so that blood returning from the head bypasses the right ventricle and flows passively into the pulmonary circulation.

The third stage, which is performed at approximately two years of age, is the completion Fontan, in which the IVC is joined into the superior cavopulmonary circulation that was previously created. After this procedure, all venous blood returning to the heart bypasses the right heart and flows passively into the lungs, while the right ventricle now has the responsibility for pumping oxygenated blood returning from the lungs to the body.

The circulation that remains is usually referred to as “Fontan physiology.” Blood flow to the lungs becomes dependent on the transpulmonary gradient, which is the pressure

difference between the Fontan circuit (systemic veins and pulmonary arteries) and the pulmonary venous atrium. Thus, any condition that increases PVR will decrease blood flow through the lungs, and cause hypoxemia. A number of perioperative factors can decrease pulmonary blood flow. After a Fontan procedure, patients may develop atrial arrhythmias or complete heart block. These arrhythmias are poorly tolerated because of the relatively large contribution of atrial contraction to ventricular filling. Adult patients with Fontan physiology may progressively develop myocardial failure, which sometimes manifests as ventricular arrhythmias.

Inhaled anesthetics decrease SVR by dilating arteriolar and venous beds, which results in a decrease in venous return. This may critically limit pulmonary blood flow by decreasing the transpulmonary gradient. In a patient with Fontan physiology, positive-pressure ventilation can also decrease pulmonary blood flow. Positive end-expiratory pressure (PEEP) and elevated mean airway pressures can impede venous return and decrease pulmonary blood flow. Spontaneous ventilation is preferred in these patients because negative intrathoracic pressure increases the gradient between extrathoracic and intrathoracic pressures and results in increased flow through the pulmonary circulation. Preferred anesthetic techniques in the Fontan patient include use of a facemask or laryngeal mask airway (LMA) with spontaneous ventilation, or a regional technique with IV sedation. However, atelectasis is likely in longer cases, in which controlled ventilation may be the most prudent option, with the goal of immediate tracheal extubation at the completion of the procedure.

**Pulmonary Stenosis (PS)**

PS accounts for approximately 8% of all CHD and usually occurs at the level of the valve, although subvalvular and supravalvular stenosis can occur. It can also occur as a component of other heart defects. PS should not be confused with peripheral pulmonic stenosis, which is a benign condition of the newborn that produces a murmur as a result of the acute angle of bifurcation of the main pulmonary artery. The clinical manifestations of PS depend on the degree of valve restriction. Right ventricular hypertrophy occurs as the ventricle attempts to maintain cardiac output. Symptoms of severe PS include CHF and cyanosis.

Moderate or severe PS (gradient ≥50 mmHg) is treated with balloon valvuloplasty. Open surgical repair may be necessary in some cases. Once dilated or repaired, children with isolated PS are relatively healthy and usually present no further anesthetic considerations.

**Tetralogy of Fallot (ToF)**

After the immediate newborn period, ToF is the leading cause of cyanotic CHD. ToF encompasses four defects: an anterior malalignment VSD, a right ventricular outflow tract obstruction, and subsequent right ventricular hypertrophy with an overriding large ascending aorta. Cyanosis occurs by right-to-left shunting across the VSD and decreased pulmonary blood flow. The degree of right ventricular outflow tract obstruction will determine the overall severity of the defect. Right-to-left shunting occurs if the resistance

caused by PVR and the right ventricular outflow tract obstruction exceeds SVR.

If for some reason the ToF is not corrected during infancy, the child may demonstrate sudden episodes of cyanosis secondary to infundibular spasm that worsens right ventricular outflow tract obstruction. These are commonly known as "Tet" spells and they may last minutes to hours, usually resolve spontaneously, and may lead to syncope, progressive hypoxia, and death. Tet spells can occur at any time in the perioperative period and can be treated by diminishing right-to-left shunting by increasing SVR and decreasing PVR.

ToF is usually repaired within the first six months of life by closing the VSD with a patch and relieving the right ventricular outflow tract obstruction. Some ToF patients are particularly prone to Tet spells at a young age, prior to complete surgical repair. These patients can be managed with the creation of a modified BT shunt as an increased source of pulmonary blood flow, to allow for growth, until a complete surgical repair is possible. Postoperatively, these infants commonly exhibit some degree of residual pulmonary insufficiency and right bundle-branch block. Ventricular arrhythmias may occur in adolescence when there is severe pulmonary insufficiency and right ventricular dilatation or dysfunction.

D-Transposition of the Great Arteries (TGA)

D-transposition of the great arteries (TGA) accounts for approximately 5% of CHD and is the most common form of cyanotic CHD presenting in the neonatal period. In TGA, the great vessels are transposed. In other words, the aorta arises from the right ventricle, and the pulmonary artery rises from the left ventricle. Thus, circulation exists as two separate parallel circuits unless a communication (PDA, VSD, or PFO) can mix the blood. Infants with TGA are cyanotic shortly after birth; as soon as the diagnosis is made, PGE1 is administered to keep the ductus open, and the infant may require emergent balloon atrial septostomy in the catheterization lab.

Surgical treatment for TGA consists of the arterial switch procedure in the early newborn period, for which survival exceeds 95%. Left ventricular function remains good throughout childhood, although supravalvular pulmonary stenosis may remain and require intervention. Occasionally, children will demonstrate atrial and ventricular tachyarrhythmias.

**Tricuspid Valve Atresia (TA)**

TA leads to hypoplasia or absence of the right ventricle. An associated VSD is found in 90% of TA cases. The VSD allows blood to pass from the left ventricle to the right ventricle and into the pulmonary artery. The majority of patients with TA also have pulmonary stenosis. The systemic venous return is shunted from the right atrium to the left atrium through a PFO or ASD and the left atrium and left ventricle handle both systemic and pulmonary venous return. Cyanosis in the neonatal period is correlated with the amount of restriction of pulmonary blood flow.

Newborns with TA manifest cyanosis, poor feeding, and tachypnea within the first two weeks of life. PGE1 is administered to maintain pulmonary flow, and a balloon atrial septostomy is performed if the atrial defect is not adequate. Surgical management involves placing a modified Blalock-Taussig shunt to maintain pulmonary blood flow. Later in infancy, a cavopulmonary anastomosis (hemi-Fontan or bidirectional Glenn) is performed to provide stable pulmonary blood flow. In most centers, a modified Fontan procedure is performed to redirect the inferior vena cava and hepatic vein flow into the pulmonary circulation. As compared to HLHS patients, these children usually benefit from having the left ventricle remain the primary pumping chamber for the systemic circulation.

## Anesthetic Management of Children with CHD

Preoperative Assessment

The extent of the preoperative evaluation will depend on the child's diagnosis and current medical condition. The anatomic and hemodynamic function of the child's heart should be completely understood, and previous anesthetics reviewed. Children that are currently under the care of a cardiologist should have a consult that includes a complete description of the child's cardiac anatomy, and a list of cardiac medications.

The best way to determine the child's functional status is to assess their limitations of daily activities and exercise. The feeding patterns of infants may provide a clue to cardiac function because of the physical effort involved to suck and swallow. Cardiac reserve is likely reduced if an infant is unable to finish a feed without tiring, or develops cyanosis, diaphoresis, or respiratory distress during feeding. Smaller children with limited cardiac output and increasing oxygen consumption will demonstrate failure to thrive or decreased normal activity. Older children may become more sedentary, spending even more time than usual on Facebook. Syncope, palpitations, and chest pain are additional symptoms of cardiac limitation that should be investigated prior to elective surgery.

Medications administered to children with CHD include diuretics, afterload reduction agents, antiarrhythmics, antiplatelet or anticoagulation drugs, and possibly inotropic or immunosuppressant agents in heart transplant recipients. All scheduled medications should be taken on the day of surgery, except for diuretics, which are usually withheld, depending on the clinical condition of the child.

Preoperative laboratory or diagnostic testing will depend on the nature of the child's illness and any recent manifestations. A hemoglobin level may be indicated for children with cyanotic CHD who compensate for chronic hypoxemia by developing polycythemia. A hematocrit that approaches 65% will increase blood viscosity and interfere with tissue microcirculation, contribute to tissue hypoxia, increase SVR, and predispose to venous thrombosis and strokes. A normal or low hematocrit may indicate relative anemia, and is usually caused by iron deficiency. Iron-deficient red blood cells are less deformable and increase blood viscosity. Anemia or polycythemia should be evaluated and corrected prior to elective surgery. This is often done in consultation with the patient's pediatric cardiologist and/or hematologist.

On physical exam, preoperative vital signs, including room air $SpO_2$, are used as a baseline to determine intraoperative norms. Baseline heart sounds, and the presence of cyanosis or pallor should be noted. The presence of tachypnea or rales on lung auscultation may indicate pneumonia or CHF. An upper respiratory tract infection warrants particularly careful evaluation and possible cancellation, as it may cause significant morbidity in children with CHD, especially when caused by respiratory syncytial virus (RSV).

CHD may be accompanied by tracheobronchial anomalies, such as shortening or stenosis, and may remain unrecognized until endotracheal intubation is required. This is particularly true for children with trisomy 21. A history of prolonged intubation after CHD surgery raises the possibility of an airway abnormality. Inspiratory stridor is an indication of airway narrowing due to subglottic stenosis or a vascular malformation that causes compression of the lower airway.

Neurologic abnormalities are not uncommon in children with CHD. The presence of a right-to-left shunt with polycythemia may predispose to an embolic stroke. Cardiopulmonary bypass is associated with microemboli that travel to the brain and cause vascular occlusion.

Children taking diuretics should have a preoperative evaluation of electrolytes. Depending on the patient's current clinical condition, an ECG or echocardiogram might be indicated.

Preoperative dehydration may be hazardous in children with cyanotic CHD and polycythemia. Attention to preoperative oral or IV hydration is especially important for children with ToF, cyanotic patients with polycythemia, and children with Fontan physiology. Dehydration may cause ToF patients to have a hypercyanotic "Tet spell". Fontan patients are dependent on venous return for pulmonary blood flow, and thus, dehydration may lead to decreased central venous pressure and subsequent decreased pulmonary blood flow and poor cardiac output. These patients may benefit from preoperative admission for overnight hydration. Fasting intervals should be no different for children with CHD than for healthy children and drinking clear liquids two hours prior to planned induction of general anesthesia should be encouraged.

Premedication with oral midazolam is useful to allay preoperative anxiety, and even hemodynamically unstable children can receive titrated IV midazolam. The advantages of preoperative anxiolysis in children with CHD include easy separation from parents, less crying, decreased oxygen consumption, and decreased levels of intraoperative anesthetics. Some anesthesiologists fear that even minimal respiratory depression caused by sedatives may cause significant oxyhemoglobin desaturation in children with cyanotic CHD whose resting oxyhemoglobin saturations lie on the steep portion of the hemoglobin dissociation curve. However, several investigations that assessed this risk demonstrated that preoperative anxiolysis resulted in less oxyhemoglobin desaturation during induction of anesthesia.

Subacute Bacterial Endocarditis (SBE) Prophylaxis

The American Heart Association published updated guidelines in 2008 on administration of prophylactic antibiotics to susceptible patients. The mechanism of infective endocarditis

(IE) involves endothelial damage with platelet and fibrin deposition, which allow for bacterial colonization. Perioperative antibiotics are recommended for dental procedures that involve manipulation of gingival tissue or perforation of oral mucosa in "high-risk" patients. These include patients with:

Prosthetic cardiac valves or prosthetic material used for cardiac valve repair

Previous endocarditis

CHD

Unrepaired cyanotic CHD, including palliative shunts and conduits

Completely repaired CHD repaired with prosthetic material or device during the first six months after the procedure

Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device

Cardiac transplant recipients with valve regurgitation due to a structurally abnormal valve

Antibiotic prophylaxis for IE is no longer recommended for patients (without an active infection) undergoing GI and GU procedures, transesophageal echocardiograms, and respiratory tract procedures, unless there is an incision of the mucosa. Respiratory procedures that should be covered include tonsillectomy, adenoidectomy, bronchoscopy, nasotracheal intubation, and any other procedure that involves an incision of the respiratory mucosa.

The current recommendations include administration of oral antibiotics 60 minutes prior to surgery or parenteral antibiotics 30 minutes prior to surgery. Parenteral antibiotic choices include ampicillin 50 mg/kg or cefazolin 50 mg/kg. Patients allergic to penicillin may receive clindamycin 20 mg/kg. Note that the recommended SBE prophylaxis dose is typically two times the normal dose for antibiotic prophylaxis for skin incisions. SBE prophylaxis is not indicated for gastrointestinal or genitourinary procedures.

Although the recommendations have been scaled down significantly since the most previous version, our experience has been that not all pediatric cardiologists will be comfortable with this most recent iteration of the guidelines and will continue to recommend antibiotics in children that do not meet the current high-risk criteria.

## Anesthetic Techniques in Children with CHD

By virtue of their propensity to cause hemodynamic compromise in susceptible CHD patients, there is no anesthetic regimen that is inherently safer than any other. All volatile anesthetics can alter PVR, SVR, myocardial contractility, heart rhythm, heart rate, and shunt flow. In healthy patients, isoflurane produces a drop in SVR by vasodilation, which may decrease mean arterial pressure. In children with CHD, isoflurane slightly increases

heart rate and tends to maintain cardiac index. In normal children and in those with CHD, sevoflurane decreases SVR and can decrease the LV shortening fraction, yet cardiac index and heart rate are maintained. Sevoflurane can also produce diastolic dysfunction. Nitrous oxide produces minimal myocardial depression, and although it is associated with increased PVR in adults, it produces minimal changes in infants with both normal and increased PVR. Nitrous oxide can, however, increase the size of an air embolus.

In children with right-to-left shunts, inhaled induction may result in an increased shunt fraction and cyanosis secondary to a decrease in SVR. In these children, a slow titration of agent is necessary with frequent measurements of blood pressure. The occurrence of hypoxemia that is not due to respiratory causes should be attributed to systemic vasodilation and right-to-left shunting, and should be treated with a direct vasoconstrictor such as phenylephrine.

Intracardiac shunts can affect the rate of anesthetic induction. In the presence of a right-to-left shunt, dilution of anesthetic agent in the left ventricle by venous blood that bypasses the lung results in a decreased concentration of agent reaching the brain. This will, theoretically, slow the rate of induction of anesthesia. Conversely, left-to-right shunts may speed induction of anesthesia by rapidly decreasing the arterial-to-venous difference of agent in the lungs. In clinical practice, these effects are hardly noticeable.

Small amounts of air trapped in IV tubing that enters the circulation can cause complications in children with CHD. Therefore, IV lines must be de-aired prior to connection. With a right-to-left shunt, an injected air bubble can cross into the systemic circulation and cause a stroke if it passes from the aorta to the brain via a carotid or vertebral artery, or result in other end-organ damage. With a left-to-right shunt, air bubbles pass into the lungs and are absorbed.

High oxygen concentrations decrease PVR and increase SVR; hypoxemia increases PVR and decreases SVR. These changes may significantly alter pulmonary blood flow by changing the PVR to SVR ratio in the presence of a large, unrestrictive intracardiac shunt.

Intravenous induction of general anesthesia with propofol can be accomplished by titrating the drug judiciously, depending on the patient's tolerance for changes in heart rate and blood pressure. In theory, a left-to-right shunt will slow IV induction and a right-to-left shunt will speed the time of induction by shunting more anesthetic agent to the brain without passing first through the lungs. But as with inhaled induction, these effects are difficult to appreciate clinically.

Ketamine and etomidate may provide greater hemodynamic stability in children with CHD. The sympathomimetic effects of ketamine tend to maintain heart rate, contractility, and SVR. There are theoretical concerns with ketamine's ability to cause an increase in PVR, especially in patients with Fontan physiology. However, this has not been substantiated in clinical studies performed in children with CHD. For the most part, opioids and benzodiazepines are safe in children with CHD as long as clinically significant bradycardia is avoided.

Regional anesthesia should be encouraged in children with CHD but with several caveats:

1. The child with longstanding CoA and dilated tortuous intercostal arteries is at risk for arterial puncture or excessive absorption of local anesthetic during intercostal blockade.

2. Since the lungs may absorb up to 80% of the local anesthetic on first passage, the risk of local anesthetic toxicity is theoretically increased in a patient with a right-to-left shunt because the brain will be exposed to a higher concentration than usual.

3. Vasodilatation resulting from central axis blockade may be hazardous in patients with left-sided obstructive lesions. Vasodilatation may also cause a decrease in oxyhemoglobin saturation in children with a right-to-left shunt. On the other hand, peripheral vasodilatation in patients with polycythemia may have the benefit of improved microcirculatory flow and decreased venous thrombosis.

4. Children with chronic cyanosis are at risk for coagulation abnormalities and should be adequately evaluated prior to initiation of regional anesthesia.

**Monitoring Children with CHD**

Pulse oximetry reliably predicts oxyhemoglobin saturation in the range that is normally encountered in children with cyanotic CHD ($SpO_2$ 70–90%). However, it may have limited accuracy at oxyhemoglobin saturations below 70%, and should be verified by blood gas analysis when in question.

Intraoperative $P_{ET}CO_2$ monitoring can be unreliable in children with CHD. $P_{ET}CO_2$ will tend to underestimate $PaCO_2$ because abnormal pulmonary ventilation/perfusion ratios result in increased dead space and/or shunt, which alter the arterial to end-tidal carbon dioxide difference.

Blood pressure monitoring accuracy in children with CHD will depend on the presence of arterial tree malformations and anatomical alteration by previous surgical corrections. For example, a modified Blalock-Taussig shunt or left subclavian flap procedure for CoA repair will render the blood pressure reading in the respective extremity inaccurate or difficult to obtain. Prior to CoA repair, lower-extremity blood pressure readings will differ from upper extremity pressures.

**Postoperative Management of Children with CHD**

Hypoventilation or mildly decreased oxyhemoglobin saturation are particularly hazardous in children with CHD. Following tracheal extubation, oxygen should be administered during transport to the PACU (or ICU) and gradually weaned based on the patient's clinical condition. In patients with single ventricle or stage I physiology, oxygen saturation should be titrated to 85% for fear of decreasing PVR, increasing pulmonary blood flow, and decreasing systemic blood flow. Postoperatively, an anesthesiologist or intensivist familiar

with their specific cardiac disease should follow these children closely. Analgesics and commonly used antiemetics are well tolerated in children with CHD.