Chapter 12
Anesthesia for Patients with Congenital Heart Disease

Dean B. Andropoulos, M.D., M.H.C.M.

Introduction

Congenital heart disease (CHD) is the most common birth defect requiring surgical or other invasive intervention. It occurs in approximately eight per 1000 live births throughout the world. Because of this, anesthetists caring for children require a basic understanding of CHD anatomy and pathophysiology. At times, they may have to anesthetize these children for cardiac or non-cardiac surgery. In addition, cardiac repairs are increasingly performed in many parts of the world, especially the complete repair of two-ventricle cardiac lesions where a single surgery can dramatically improve the patient’s quality of life and significantly increase her/his life expectancy. This chapter first presents a classification system for CHD and then discuss an approach to its diagnosis, including history and physical examination, laboratory examinations, and diagnostic imaging (focusing on echocardiography). Next, there is a discussion of the major cardiac lesions with pathophysiology, surgical approach, and anesthetic considerations presented. Following this discussion, there is a general discussion of cardiopulmonary bypass management. Finally, the subject of non-cardiac surgery in CHD is summarized.

Even if an anesthetist will not provide care for patients with CHD, an understanding of the principles discussed in this chapter will improve her/his care of other patients. Pediatric patients with normal hearts still encounter problems with poor cardiac function; fluid overload with cardiomegaly; low blood pressure from infection; hypovolemia; or bleeding; and pulmonary hypertension, especially if they are neonates. Some patients not known to have CHD will become hemodynamically unstable and subsequently be discovered to have heart disease.

Classification of Congenital Heart Disease

A common classification of CHD divides these lesions into seven basic categories: 1) Left-to-right shunts, where an obstruction to right sided outflow causes blood to flow through an intracardiac or extracardiac communication from the left side to the right side of the heart; 2) Right-to-left shunts, where obstruction to right heart outflow causes blood to flow from the right side to the left side of the heart through an intracardiac or extracardiac communication; 3) Left sided
obstructive lesions, where flow out the left side of the heart (at any level) is obstructed; 4) Right sided obstructive lesions, where flow is obstructed through the right side of the heart, usually without cyanosis; 5) Regurgitant lesions, where a cardiac valve is insufficient; 6) Mixing lesions, where there are two cardiac ventricles but extracardiac malformations allow mixing of blood from both sides of the heart; and 7) Single ventricle lesions, where there is a single functional right or left ventricle and complete intracardiac mixing of blood. **Table 12-1** summarizes this classification and presents common examples. It is very important to understand that many patients have more than one type of lesion; for example aortic stenosis with a ventricular septal defect (VSD) is both a left-sided obstructive lesion and a left-to-right shunting lesion.

**Table 12-1: Classification of Congenital Heart Disease**

<table>
<thead>
<tr>
<th>Category</th>
<th>Cyanosis</th>
<th>Pulmonary Blood Flow</th>
<th>Common Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left-to-Right Shunts</td>
<td>No</td>
<td>Increased</td>
<td>VSD, ASD, PDA</td>
</tr>
<tr>
<td>Right-to-Left Shunts</td>
<td>Yes</td>
<td>Decreased</td>
<td>TOF, pulmonary atresia</td>
</tr>
<tr>
<td>Left-Sided Obstructive Lesions</td>
<td>No</td>
<td>Normal</td>
<td>CoA, aortic stenosis, mitral stenosis</td>
</tr>
<tr>
<td>Right-Sided Obstructive Lesions</td>
<td>No*</td>
<td>Normal</td>
<td>Pulmonic stenosis</td>
</tr>
<tr>
<td>Regurgitant Lesions</td>
<td>No</td>
<td>Normal</td>
<td>Ebstein’s anomaly</td>
</tr>
<tr>
<td>Mixing Lesions</td>
<td>Yes</td>
<td>Variable</td>
<td>TGA, truncus arteriosus, TAPVR</td>
</tr>
<tr>
<td>Single Ventricle Lesions</td>
<td>Yes</td>
<td>Variable</td>
<td>Tricuspid atresia, HLHS</td>
</tr>
</tbody>
</table>

*Cyanosis may be present if accompanied by a septal defect. VSD, ventricular septal defect; ASD, atrial septal defect; PDA, patent ductus arteriosus; TOF, tetralogy of Fallot; CoA, coarctation of aorta; TGA, transposition of the great arteries; TAPVR, total anomalous pulmonary venous return; HLHS, hypoplastic left heart syndrome.
Table 12-2 presents the incidence of the most common congenital heart disease lesions in children from birth to 18 years of age in the United States.

Table 12-2: Incidence of Common Lesions in Children With CHD
*Source: Circulation 2012;125:e97.*

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular Septal Defect</td>
<td>20.1</td>
</tr>
<tr>
<td>Atrial Septal Defect</td>
<td>16.8</td>
</tr>
<tr>
<td>Valvar Pulmonic Stenosis</td>
<td>12.6</td>
</tr>
<tr>
<td>Patent Ductus Arteriosus</td>
<td>12.4</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>7.0</td>
</tr>
<tr>
<td>Coarctation of Aorta</td>
<td>6.8</td>
</tr>
<tr>
<td>Valvar Aortic Stenosis</td>
<td>5.5</td>
</tr>
<tr>
<td>Atrioventricular Septal Defects</td>
<td>3.9</td>
</tr>
<tr>
<td>Transposition of the Great Vessels</td>
<td>3.6</td>
</tr>
</tbody>
</table>

Diagnostic Evaluation of Congenital Heart Disease

The fundamentals of diagnostic evaluation of CHD include five basic components: history, physical examination, chest radiograph, electrocardiogram (ECG), and hemoglobin level (Hgb). From these five components, an accurate idea of the cardiac lesion can be discerned in nearly every case. In addition, significant insights can be gained into the pathophysiologic effects of the lesion(s) on individual patients. To these five basic components are added cardiac imaging modalities: echocardiography, cardiac catheterization, cardiac magnetic resonance imaging (MRI), and cardiac computed tomography (CT). Arterial oxygen saturation (SpO₂) completes the diagnostic picture.

History

The patient’s history is crucially important for determining her/his baseline status, duration and severity of symptoms, and previous cardiac diagnostic and therapeutic interventions. For infants, tachypnea and feeding difficulties are very common features of left-to-right shunting lesions. Shunting leads to increased work of breathing and poor ability to breastfeed. Diaphoresis (profuse sweating) is also a common finding with congestive heart failure (CHF). Pallor, listlessness, and cyanosis may be present with more serious cardiac lesions. Poor growth is a common finding in patients with congestive heart failure (CHF). In older children, cyanosis is very common with lesions like tetralogy of Fallot (TOF). Patients with TOF often assume a squatting position to increase their systemic vascular resistance, decrease right-to-left blood flow (shunting), and to increase pulmonary blood flow, which relieves the cyanosis. Older children with CHD often have
dyspnea on exertion and cannot keep up with siblings or peers during play or exercise. Significant left ventricular or aortic obstruction may be accompanied by fainting spells; this is also seen with cardiac arrhythmias, such as supraventricular or ventricular tachycardia. Angina is observed in some older children with coronary artery difficulties or with significant aortic stenosis. Children who had cardiac surgery previously may have sternotomy or thoracotomy scars. If they do this should provoke questioning about the procedure and careful examination of previous medical records to understand what procedure was performed and what (if any) residual lesions remain. Cardiac catheterization procedures may include interventions, such as dilation of aortic and pulmonary valves or closure of a ventricular septal defect. The nature and the result of these interventions should be well understood if possible.

**Physical Examination**

*Physical examination* is best performed when the child is calm. One effective way to calm young children is to have her/him sit in the parent’s lap; distracting them with toys is also effective. Vital signs are measured, including arterial blood pressure (BP) in four extremities, heart rate (HR), respiratory rate (RR), temperature, and the “5th vital sign of CHD”: oxygen saturation (SpO₂) by pulse oximeter. Vital signs outside normal ranges are critically important: HR and RR are often elevated in CHF. The level of cyanosis, as determined by SpO₂, has major implications for anesthesia and surgery. For the physical examination, the first step is *observation* of the child: pallor, tachypnea, listlessness, failure of normal interaction with people, cyanosis, edema, jugular venous distension, and failure to thrive are important findings in patients with significant CHD. *Palpation* is also important: strength and quality of radial and femoral arteries pulses, perfusion (as judged by capillary refill time and temperature of the extremities), edema, hepatosplenomegaly, a cardiac thrill, and position of the cardiac apex all provide important information. *Auscultation* of the heart and lungs is next. Quality and loudness of first and second heart sounds (S₁ and S₂) are important—muffled heart sounds can indicate low cardiac output; a split S₂ accompanies atrial septal defects (ASD), a loud S₂ is heard with pulmonary hypertension, and a single S₂ is heard with pulmonary atresia. Third and fourth heart sounds (S₃ and S₄) are heard with congestive heart failure and cardiomyopathy. Cardiac murmurs are important. Systolic murmurs are heard with restrictive flow, as with aortic or pulmonary stenosis, or a ventricular septal defect (VSD). Diastolic murmurs can be caused by mitral stenosis, and systolic-diastolic or continuous murmurs by patent ductus arteriosus (PDA). Gradation of murmurs from grade I-II/VI (soft-medium intensity murmur), grade III/VI (loud murmur), or grade IV-VI/VI (soft thrill, loud thrill, or audible thrill) gives important clues to the severity and location of the cardiac lesion. Auscultation of the lungs is also very important because it allows the examiner to detect rales (heard in congestive heart failure), wheezing (heard with the “cardiac asthma” that is associated with pulmonary venous obstruction due to left-sided obstructive lesions), or diminished breath sounds (lobar collapse from enlarged cardiac structures).
Chest Radiograph

The chest radiograph (CXR) yields information about both the cardiac lesion itself and the pathophysiology of the individual patient. An anterior-posterior (AP) CXR is often sufficient, but a lateral CXR can add important information. Heart size is an important consideration; a cardiothoracic ratio greater than 55% indicates cardiomegaly. Cardiomegaly is a sign of congestive heart failure, which may be the result of left-to-right shunting or cardiomyopathy. Cardiac silhouette is also important; a “boot shaped heart” is an indication that the main pulmonary artery (PA) is hypoplastic, which occurs in TOF. The narrow mediastinum seen with dextrotransposition of the great vessels (d-TGA) is often referred to as an “egg on a string”. The “snowman” or “figure of eight” appearance of the mediastinum is caused by a distended vertical vein, which is often observed with obstructed supracardiac total anomalous pulmonary venous return (TAPVR). The appearance of the lung fields is crucial: “black” lung fields indicate diminished pulmonary blood flow; normal faint vascular markings indicate a normal pulmonary blood flow; and increased vascular markings and distended circular shadows on the CXR indicate small pulmonary arteries en face, which is indicative of increased pulmonary blood flow. The immediate postoperative CXR is important for assessing position of the tracheal tube, central lines, and evidence of intrathoracic bleeding or intrathoracic air. Figure 12-1 displays examples of common CXR findings.

Figure 12-1: Chest Radiograph in Patients With Congenital Heart Disease
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A: Transposition of the great arteries with ventricular septal defect: narrow mediastinum and increased pulmonary vascular markings; B: Tetralogy of Fallot: “boot-shaped” heart and normal pulmonary vascular markings; C: Cardiomyopathy: cardiomegaly with increased pulmonary vascular markings; D: Total anomalous pulmonary venous return: “Figure of 8” heart with increased pulmonary vascular markings; E: Postoperative CXR after ventricular septal defect repair; endotracheal tube is too low, and catheter (arrow) is in place.

Hemoglobin Level

The baseline hemoglobin (Hgb) level is an important functional test of the degree of cyanosis in CHD. When blood oxygen tensions are lower than normal, the kidney secretes erythropoietin, which induces hematopoiesis and increases red cell mass. This increases the oxygen carrying capacity of blood to compensate for cyanosis (See Chapter 1). In general, the higher the Hgb
level, the lower the resting SpO₂; a normal infant undergoes a physiologic nadir in Hgb to about 10-11g/dl by 3-4 months of age. In cyanotic infants, the increase in Hgb levels is inversely proportionately to the patient’s baseline SpO₂, sometimes reaching 15-16g/dl. Older children and teenagers with chronic, longstanding, severe cyanosis may have Hgb concentrations exceeding 20g/dl, which are often associated with signs and symptoms of hyperviscosity, including headache, visual changes, poor circulation in the extremities, clubbing of the finger and toe nails, and polycythemia-induced coagulopathy. Patients with mixing lesions and mild cyanosis may have near normal Hgb levels.

**Electrocardiogram**

A 12-lead ECG is an important source of preoperative information and for the diagnosis of CHD because it not only provides heart rate and rhythm, it also provides important information about chamber hypertrophy, intraventricular conduction delays, and myocardial strain (ST segment changes). The preoperative ECG also serves as a baseline for comparison if cardiac rhythm changes are suspected postoperatively. **Figure 12-2** is an example of the preoperative ECG in of patient with complete atrioventricular canal.

**Figure 12-2: Electrocardiogram of a 4-Month Old Patient with Complete Atrioventricular Canal**
This is a complete, 15-lead ECG obtained in the Cardiology Clinic. Leads I, II, III, aVR, aVL, aVF get data from electrodes on the left and right arms, and left and right legs. Leads V1-V7 obtain data from leads on the chest, starting with V1 just to the left of the sternum near the costal margin. Leads V2-V7 follow a line just above the costal margin, progressively more lateral, until V7 lead is placed at the mid-axillary line. Leads V3r and V4r are placed to the right of the sternum. The long lead II tracing at the bottom of the printout is to diagnose arrhythmias. This ECG shows normal sinus rhythm (P-QRS-T), a rate of 150 beats/min, and the QRS voltages in all of the V leads indicate biventricular enlargement. The electrical axis of the R wave is -70 degrees, the “Northwest Axis” as one would see on a map; this finding is very characteristic of complete atrioventricular canal, or endocardial cushion defect.

**Echocardiography**

Transthoracic echocardiography is the mainstay of imaging diagnosis for congenital heart disease. It is portable, compact, non-invasive, relatively cost effective compared to other imaging methods, and highly accurate for imaging intracardiac anomalies and for cardiac function. These studies can be completed in a short period of time and can be repeated after an intervention. Two-dimensional imaging is the standard for anatomic diagnosis. This information is enhanced by color-flow Doppler interrogation to assess flow across valves, septa, and through shunts. Pulsed wave or continuous wave Doppler interrogation can be used to measure or calculate peak and mean velocities, as well as to calculate pressure gradients across intracardiac communications, valves, or outflow tracts. Ejection fraction and shortening fraction are accurate measures of ventricular function. If possible, the anesthetist should at least have access to the echocardiography reports. However, it is preferable to view the echo images in a presurgery planning conference with a radiologist and surgeon. Transesophageal echocardiography is used in the operating room pre- and postoperatively to assess anatomy, function, and residual defects after surgery. **Figure 12-3** shows examples of echocardiography for common lesions.
Figure 12-3: Echocardiography in Congenital Heart Disease

A: Ventricular septal defect (arrow) in 4-chamber view; B: Complete atrioventricular canal, with primum atrial septal defect (upper arrow) and ventricular septal defect (lower arrow); C: Cor triatriatum (double arrows)—a rare defect consisting of a membrane in the left atrium; D: dextrotransposition of the great vessels. Note the pulmonary artery (PA) arising from left ventricle (LV) and aorta (AO) from right ventricle (RV). Arrow is pointing to a ventricular septal defect. AO = aorta; PA = pulmonary artery; RV = right ventricle; LV = left ventricle; RA = right atrium; LA = left atriumSource: Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 86.

Cardiac Catheterization

Cardiac catheterization is capable of determining the anatomy and physiology of CHD. It is increasingly used for catheter-based interventions, i.e., balloon dilation of pulmonary or aortic valve stenosis, closure of PDA or ASD, or stenting of coarctation of the aorta. Although information from cardiac catheterization may be very useful in preoperative planning, in most settings the echocardiogram will be sufficient for this purpose. An example of information
obtained from a cardiac catheterization is shown on the diagram in Figure 12-4. On a single page, the anatomy, intracardiac and vascular pressures, oxygen saturations in each cardiac chamber, and pulmonary to systemic blood flow ratios are summarized. In addition, the previous surgical history is also listed, making this diagram extremely useful in planning an anesthetic for the cardiac surgery. Patients with simple cardiac lesions almost never undergo cardiac catheterization.

Figure 12-4: Cardiac Catheterization Diagram

This patient has a very complicated CHD lesion; patients with simple cardiac lesions rarely, if ever, undergo cardiac catheterization. \( a \) = a wave pressure in mmHg; \( v \) = v wave pressure in mmHg; \( m \) = mean pressure in mmHg. ABG = arterial blood gas; numbers in white circles are measured oxygen saturations. Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 87.
Cardiac Magnetic Resonance Imaging (MRI) and Computed Tomography (CT)

Cardiac MRI and CT scanning are non-invasive methods of providing accurate images of cardiac anatomy and are particularly suited for extracardiac structures, i.e., the aorta and its branches, pulmonary arteries and pulmonary veins. The equipment is complicated and expensive to maintain; therefore it may not be available in some programs. The advantage of MRI is that no ionizing radiation is involved; however at least 30-60 minutes are required for each study, and breath holding is sometimes required during the study. This means that infants and young children require sedation, or even tracheal intubation and general anesthesia for the study. CT scanning is a much shorter examination, requiring only 3-5 minutes to image cardiac structures; even young children require little or no sedation for the procedure. The major drawback to a CT scan is that radiation exposure can be significant; consequently, many cardiologists prefer MRI. The reader is referred to the bibliography for more information about MRI and CT.

Left-to-Right Shunt Lesions

Left-to-right shunt lesions account for just over 50% of CHD and are the most common lesions requiring surgical correction in many cardiac surgical programs. The pathophysiology of these lesions is highly variable and depends on the size and location of the communication and the relative pressure and resistance on either side of the communication. The pulmonary-to-systemic blood flow ratio, or $Q_p:Q_s$, is highly variable and ranges from just over 1.0 with small shunts, to $>5:1$ with very large shunts. In the latter instance, pulmonary blood flow is five times systemic flow. A shunt is termed restrictive if the communication between cardiac chambers or great vessels is small enough to cause flow acceleration, designated by a peak pressure gradient of $>10-20$ mm Hg, and a Doppler peak velocity of greater than $2-2.5$ m/sec. The smallest shunts tend to be ASDs because the pressure gradient between left and right atria is small, even if the ASD is large. Ventricular level shunts range from small VSDs of $<2-3$ mm in diameter, to very large unrestricted VSDs that allow large amounts of blood to flow from left-to-right. The largest shunts may occur at the level of the great arteries, with a large PDA or aortopulmonary window producing continuous left-to-right shunting from the aorta directly into the pulmonary artery (PA). Besides the pressure gradient across the ventricular septum and the relative compliance of right and left ventricles, the resistance in the systemic and pulmonary circulations influences $Q_p:Q_s$. With relatively high systemic vascular resistance (SVR) and low pulmonary vascular resistance (PVR), the $Q_p:Q_s$ can be $>3:1$. This degree of pulmonary over circulation often causes RV dilation, pulmonary edema, pulmonary venous congestion, and congestive heart failure. Maneuvers that further decrease PVR, i.e., $FiO_2$ of 1.0 and hyperventilation, can increase $Q_p:Q_s$ even further and cause a steal phenomenon (blood is “stolen” from the aorta), decreases aortic pressure, and reduces coronary artery blood flow, which may cause myocardial ischemia. To prevent a “steal” is the rationale for lowering $FiO_2$, even to 0.21, and allowing some degree of hypercarbia, $PaCO_2$ 45-50 mmHg, to increase the PVR and reduce the $Q_p:Q_s$. 
A longstanding unrestrictive left-to-right shunt chronically exposes the pulmonary circulation to high flows and pressures, which results in pulmonary hypertensive changes, including thickening of the muscular medial layer of the pulmonary artery walls and extension of muscularization outward in to smaller pulmonary arterioles. Pulmonary pressure and resistance increase further; at some point they become fixed and unreactive, resulting in right-to-left shunting of blood during systole, and eventually during the entire cardiac cycle. This condition of fixed pulmonary hypertension and right-to-left shunting is termed Eisenmenger Syndrome, which prevents safe surgical closure of the VSD because doing so causes acute right heart failure. The pathophysiology of left-to-right shunts is diagrammed in Figure 12-5.

**Figure 12-5: Pathophysiology of Left-to-Right Shunts**

RVEDP = right ventricular end-distolic pressure; LVEDP = left ventricular end-diastolic pressure. Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 91.
Ventricular Septal Defects

VSDs are the most common form of CHD, occurring in about 20% of patients with CHD and of these perimembranous VSDs (those located in the perimembranous septum) are the most common type. Other types of VSDs include: supracristal or subarterial VSDs, which are located just beneath the pulmonary valve; inlet VSDs are located in the inlet septum just underneath the septal leaflet of the tricuspid valve; and muscular VSDs, which can be located anywhere in the muscular septum (Figure 12-6). Subarterial VSDs can cause aortic valve prolapse and aortic insufficiency from the Venturi effect created by blood flowing at high velocity and causing negative pressure that “sucks” the leaflet toward the VSD. Significant sized VSDs with Qp:Qs >3:1 produce significant right ventricular dilation and may cause pulmonary edema. The infant with a significant VSD has tachypnea, diaphoresis, poor feeding, pulmonary congestion, infections, and failure to thrive. Anticoagulant therapy with diuretics, including furosemide, and afterload reduction with angiotensin converting enzyme (ACE) inhibitors is often required before surgery. Longstanding large VSDs result in fixed pulmonary hypertension and may produce Eisenmenger Syndrome. Findings on physical examination include tachycardia, displaced point of maximal impulse (PMI) downward and lateral at the left 5th or 6th interspace, a grade II-III/VI systolic murmur at the left sternal border, and if Qp:Qs >3:1, a diastolic murmur that is caused by increased blood flow across the tricuspid valve. The patient is acyanotic unless he/she has fixed pulmonary hypertension, in which case the degree of cyanosis parallels the right-to-left shunting of blood. Patients with larger VSDs have cardiomegaly and increased pulmonary vascular markings on CXR. ECG may be normal or reveal evidence of biventricular and/or atrial enlargement. Echocardiography is a very accurate means of diagnosing the location and number of VSDs, as well as their size and the flow velocity across them, which aids in determining if the defect is restrictive.

Large symptomatic VSDs are normally surgically repaired in infancy, which requires cardiopulmonary bypass, aortic cross clamping, and an autologous pericardial patch to close the VSD. Smaller VSDs are often repaired later in childhood. Transesophageal echocardiography (TEE) is very helpful during surgery to ensure that no residual defects remain. With subarterial defects, any aortic valve prolapse may require repair. With repair of the VSD, the ventricular dilation and pulmonary hypertensive changes usually regress over the weeks and months following surgery; the child should have a normal life expectancy.
Figure 12-6: Ventricular Septal Defect Anatomy

Types of VSD: A: supracristal or subarterial; C: perimembranous; F: inlet or canal-type; D,E,G: muscular VSD. B: tricuspid valve cordae. Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 96.

Atrial Septal Defects

Atrial septal defects (ASD) are the second most common CHD defect, comprising about 17% of lesions. The most common of these is a secundum ASD, located in the middle of the atrial septum secundum. The primum ASD, located in the septum primum, is often associated with a partial atroventricular canal (see below). Other types of ASD include the inferior sinus venosus, and the superior sinus venosus ASDs (Figure 12-7). The superior sinus venous ASD is often associated with partial anomalous pulmonary venous return (PAPVR) where two right superior pulmonary veins return abnormally to the superior vena cava (SVC). Some patients have multiple small ASDs, and their patent foramen ovale (PFO) may be large enough to require closure. Because the degree of
left-to-right shunting with ASDs is relatively small, due to the low pressures and low-pressure gradient between the left and right atria, Qp:Qs usually remains at or below 2:1 and symptoms may develop late, sometimes not until the 3rd or 4th decade of life. Dyspnea with exertion is the most common presenting symptom. Pulmonary vascular disease rarely develops, but if it does, it only does so after the 5th decade of life. Physical examination may be normal, except for a soft grade I-II/VI systolic murmur at the left sternal border. The second heart sound is often split and fixed, representing delayed closure of the pulmonary valve. The delay caused by increased right ventricular stroke volume. CXR may reveal cardiomegaly and increased pulmonary vascular markings when ASDs are large. An ECG is often normal, but it can show atrial or ventricular enlargement. Echocardiography provides accurate diagnosis of the lesion and its location.

ASDs are surgically closed during childhood, usually by age 3-5 years of age if diagnosed early. Older children, teenagers, and adults often have their ASDs diagnosed and closed later in life. ASD closure may be done in the catheterization laboratory via a catheter-deployed “clamshell” device that is inserted through a large sheath in the femoral vein and guided into position by echocardiography. This procedure is only possible if the ASD is a secundum defect in the center of the atrial septum and if there is an adequate rim of tissue surrounding the ASD to which the device can be anchored without impinging on the aortic valve. All other ASDs are closed surgically with an autologous pericardial patch, which requires CPB and a short period of aortic cross clamping or induced ventricular fibrillation. Patients with sinus venosus ASD and PAPVR require an ASD patch that includes the pulmonary veins and directs blood flow from the veins under the patch to the left atrium. Intraoperative TEE is helpful for detecting residual surgical defects. Patients with an ASD usually recover very quickly and have no residual defects. Their life expectancy should be normal.
Patent Ductus Arteriosus

*Patent ductus arteriosus* (PDA) is another common left-to-right shunt lesion, observed in about 12% of patients with CHD (Figure 12-8). The lesion may be isolated but is often part of a constellation of other cardiac lesions. Ductus arteriosus (DA) is part of normal fetal circulation. It functionally closes during the first 24-48 hours of life in most babies and is permanently closed by fibrosis during the first several weeks of life. If the PDA fails to close, it may persist throughout life. After birth a PDA may be essential for providing systemic blood flow for several CHD lesions, including hypoplastic left heart syndrome (HLHS) and moderate to severe coarctation of the aorta. PDA closure in patients with these lesions markedly reduces systemic perfusion, which, for patients with HLHS, is usually rapidly fatal. In patients with severe CoA, PDA closure produces subdiaphragmatic viscera and kidney ischemia, acidosis and shock, cardiac arrest, and death if not reversed. Patients with pulmonary atresia or severe pulmonic stenosis require a PDA with left-to-right shunting to provide enough pulmonary blood flow to sustain oxygenation and maintain organ perfusion and function. Prostaglandin E-1 (PGE-1) infusion 0.025-0.05mcg/kg/min
maintains a PDA in patients with these lesions until surgical palliation or surgical correction of the lesion can be accomplished. Isolated PDAs vary greatly in size and length. Because of this, the magnitude of left-to-right shunt is highly variable, ranging from Qp:Qs of just over 1:1 to >3:1. Symptoms vary from none to tachypnea, tachycardia, and CHF in some infants. Because there is continuous blood flow from aorta to PA, the PVR of large unrestrictive PDAs increases rapidly, causing early pulmonary vascular disease, fixed PVR, and right-to-left shunting. Adults with large untreated PDAs may present with huge, aneurysmal, calcified PDAs and Eisenmenger Syndrome, a situation generally considered untreatable. Physical examination may reveal signs of CHF, with tachypnea, tachycardia, and rales; the pulse pressure is often widened by the continuous diastolic runoff of blood into the PA, which lowers diastolic BP and causes bounding pulses. A long systolic, or continuous murmur is heard at the left sternal border. CXR often reveals cardiomegaly and increased vascular markings. Echocardiography is useful for determining size, length, and the velocity of flow in the PDA. Except for tachycardia, the ECG if often normal; in rare instances ST segment changes are seen on ECG, indicating myocardial ischemia.

**Figure 12-8: Patent Ductus Arteriosus Anatomy**

*Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 92.*
Closure of a PDA in premature infants is often attempted first with indomethacin or ibuprofen. If the PDA remains open, it is surgically closed through a left thoracotomy. This is often done at the bedside, not in the operating room. In premature infants, care must be taken during PDA ligation to monitor lower extremity pulses and perfusion by pulse oximeter to prevent the surgeons from inadvertently ligating the descending thoracic aorta, which may be smaller than the PDA. If the aorta is occluded, hypoxemia rapidly develops in the lower body. If the PDA is of appropriate size and length, patients weighing more than 5kg can have their PDA closed in the catheterization laboratory with occlusion devices placed through a sheath in the femoral artery. However, the most common approach is surgical ligation and division of the PDA via left thoracotomy. A PDA can also be closed thoracoscopically. A PDA or ligamentum arteriosum may be components of a vascular ring encircling the trachea or esophagus. At the end of surgery, the trachea of older infants can be extubated immediately; this is not usually the case for premature infants. Recovery from the procedure and anesthesia is rapid. Pulmonary hypertension may develop in older children or adults with a PDA. If so, they are at greater risk of morbidity or mortality from right heart failure following PDA closure.

Atrioventricular Canal Defects

Atrioventricular canal (AVC) defects, also known as atrioventricular septal defects (AVSD), are relatively common left-to-right shunt lesions that occur in about 4% of patients with CHD. AVCs consist of a primum ASD, an inlet VSD, and abnormalities of the atrioventricular (AV) valves, ranging from a cleft in the mitral valve to a single common AV valve. A partial AVC consists of a primum ASD, cleft mitral valve, and no VSD or a VSD covered by tricuspid valve tissue that prevents left-to-right shunting of blood. An intermediate or transitional AVC has the findings of a partial AVC, but also has a small inlet VSD. A complete AVC (CAVC), also known as an endocardial cushion defect, has a primum ASD, large inlet VSD, and common AV valve (Figure 12-9). Complete AVC is commonly found in patients with Trisomy 21 (Down Syndrome) who often develop irreversible pulmonary hypertension during the first 1-2 years of life if the lesion is not repaired. Symptoms in patients with an AVC depend on the size of the left-to-right shunt. Partial canals tend to be minimally symptomatic, intermediate canals have a higher chance of being associated with heart failure, and CAVC is likely to present with symptoms of heart failure in infancy (tachypnea, poor feeding, poor growth), and frequent respiratory infections. Physical examination may reveal tachycardia and tachypnea with rales, normal S1, fixed split S2, and because the AV valves are often regurgitant, a grade III/VI harsh systolic murmur caused by flow across the VSD and by mitral and tricuspid valve regurgitation. An S3 is often present when the Qp:Qs exceeds 3:1. CXR reveals cardiomegaly and increased pulmonary vascular markings. ECG findings in CAVC are unique because the endocardial cushion defect changes the electrical vector of the heart, placing the R axis in the 0 to -90° quadrant, a “Northwest Axis”, which is essentially pathognomonic for CAVC. The ECG in Figure 12-2 demonstrates this finding. Biventricular and atrial enlargement are also frequently observed in CAVC. Definitive diagnosis is by
echocardiography, which accurately images the ASD, VSD, and the AV valves and their regurgitation. Medical treatment is with oral diuretics, and possibly with ACE inhibitors for afterload reduction. Because of very large left-to-right shunting, the pulmonary circulation of patients with CVAC is exposed to high flows and pressures. If CAVC remains untreated, pulmonary hypertension develops and becomes fixed and non-reactive, which will prevent surgical repair due to the associated high mortality. As their disease worsens, right-to-left shunting of blood causes these patients to become cyanotic. Their CHF symptoms improve, and the CXR now shows a normal sized heart and oligemic lung fields. Growth often improves, giving a false sense of security that the patient has improved and can undergo surgery. Patients with Down Syndrome develop irreversible pulmonary hypertension earlier in life than patients with normal chromosomes; Down syndrome patients with CAVC often become inoperable if their lesion(s) are not repaired in the first 1-2 years of life.

Surgical correction of CAVC requires CPB and involves patch repair of the VSD (if present), patch closure of the ASD, and repair of the AV valves, which usually requires repair of mitral valve cleft and anchoring the AV valve to the patch. CAVC repair is technically challenging for the surgeon. Because of this, varying degrees of residual AV valve regurgitation may remain after surgery. Intraoperative TEE is used to assess the repair at the end of CPB. If abnormalities are found, it may be necessary to return the patient to CPB for further repair. Because of the frequent incidence of pulmonary hypertension, the tracheas of these patients are usually not extubated early. Sedation and mechanical ventilation are required until their pulmonary artery pressures (PAP) decrease. If available, inhaled nitric oxide (iNO) is sometimes used in this setting; however this therapy is a very expensive and may have limited effectiveness. CAVCs are ideally repaired at <6 months of age; transitional AVC is usually repaired at 1-2 years of age, and partial AVC is repaired at about 3-5 years of age. Residual mitral regurgitation or stenosis is a significant problem with AVC repair, and these patients require regular follow-up with a cardiologist because repeat surgeries may be necessary.
Figure 12-9: Complete Atrioventricular Canal Anatomy

Right-to-Left Shunt Lesions

Tetralogy of Fallot

*Tetralogy of Fallot* (TOF), the most common *cyanotic* CHD, occurs in about 7% of patients. The four components of TOF are: 1) Large subarterial VSD with over-riding aorta; 2) RV outflow tract obstruction (RVOTO) at the level of the sub-pulmonary valve infundibulum; 3) RV hypertrophy, and 4) Right-sided aortic arch (about 25% of patients) (*Figure 12-10*). Main and branch pulmonary arteries (PA) may be hypoplastic because reduced pulmonary blood flow reduces/prevents growth of pulmonary vessels. The symptoms of TOF are highly variable and depend on the degree of RVOTO. If RVOTO is mild, TOF can be acyanotic (“pink tet”) and have symptoms similar to those with a large left-to-right shunt, e.g., VSD. With increasing RVOTO, dynamic obstruction to pulmonary blood flow occurs. This increases RV pressure, which causes right-to-left shunting through the VSD and cyanosis. Because RVOTO is a dynamic muscular obstruction, hypercyanotic spells (“Tet Spells”) can occur when anything (e.g., crying, pain, hunger, thirst, etc) increases sympathetic stimulation. The spells may be so severe that the hypoxemia they produce causes the patient to lose consciousness. This lowers endogenous catecholamine secretion and ends the
TOF spell. Treatment of TOF spells outside of the operating room includes oxygen administration, IV volume infusion, and sedation. Morphine effectively treats TOF spells. Older children with unrepaired TOF and cyanotic spells often prevent or treat a spell by assuming a squatting position. Squating “kinks” the iliofemoral arterial system and increases SVR; this decreases the right-to-left shunting. This principle is used intraoperatively as well (see below). Physical examination reveals varying degrees of cyanosis; clubbing of fingers is a late sign. Cardiac examination reveals normal S1, diminished S2, and a harsh grade II-IV/VI long systolic murmur at the left sternal border. CXR often reveals a “boot-shaped heart”, which is due to the hypoplastic main PA. The reduced pulmonary blood flow makes the lung fields of cyanotic patients oligemic. Patients with acyanotic TOF may have normal or even increased pulmonary vascular markings. ECG may reveal RV hypertrophy. The elevated Hgb level (polycythemia) of patients with TOF is proportional to the degree of cyanosis. The worse the cyanosis, the higher the Hgb. Older children with unrepaired lesions may be polycythemic and have Hgb concentrations that exceed 20g/dl. If they do, they can have headache, severe fatigue, and sluggish peripheral circulation. Definitive diagnosis of TOF is by echocardiography, which accurately displays all of its features. Medical therapy for TOF may include a β-adrenergic receptor blocker, such as propranolol, to decrease the force of RV contraction and RVOTO; these drugs often prevent “Tet spells”. Severely cyanotic neonates may require infusion of PGE-1 to maintain the PDA until surgery can be performed.

Figure 12-10: Tetralogy of Fallot Anatomy

Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 111.

The age at which TOFs are repaired varies greatly. A severely cyanotic infant may require early surgery to create a systemic-to-pulmonary artery shunt (Blalock-Taussig, or BT shunt), which is a
3-5 mm Goretex® graft from the right subclavian or innominate artery to the pulmonary artery (Figure 12-11). This surgery is usually performed via right thoracotomy and normally does not require CPB. Placing the shunt allows the infant to survive and grow until he/she is ready for corrective surgery. Corrective surgery is performed at any age from neonate to adult and consists of VSD repair, RVOT muscle resection, and often includes placing a patch across the pulmonary valve annulus onto the proximal PA, which necessitates incising the pulmonary valve annulus. “Tet spells” may occur under anesthesia. If they do, treatment includes IV volume infusion to increase RV stroke volume; deepening anesthesia without lowering the arterial blood pressure, often with an opioid like fentanyl to decrease catecholamine release and reduce RVOTO; increasing SVR with a vasoconstrictor, such as phenylephrine, to produce more left-to-right shunting through the VSD and raise SpO₂. If the sternum is open, the surgeon can increase SVR by partially occluding the aorta. After surgery, varying degrees of either pulmonary insufficiency, or RVOTO may remain. TEE is important for immediate assessment of the repair. Direct RV pressure measurement is often used to determine if the RV pressure following CPB is <50% of systemic, as desired. Older children or teenagers with TOF often require further surgery to place a pulmonary valve. Immediate post-CPB problems include: 1) a non-compliant RV, that necessitates the use of high filling pressures; 2) treatment of arrhythmias, including junctional ectopic tachycardia (JET), which causes low cardiac output. Treatment of JET includes: 1) reducing catecholamine infusion (if being used), 2) cooling the patient, 3) placement of temporary atrial pacing via wires on the RA appendage, and 4) administering procainamide or amiodarone. JET usually resolves in 48-72 hours.
Pulmonary Atresia

Pulmonary atresia is a complex lesion in which the anatomy varies from simple valvar PS with a large VSD and normal sized PAs, to a much more severe form that includes an intact ventricular septum and no true pulmonary arteries. Major aortopulmonary collateral arteries (MAPCAs) supply blood flow to the PAs. For a baby to survive, blood must flow from RA or RV (through a septal) defect to the LA or LV. During neonatal life the PDA supplies pulmonary blood flow. The severe forms of pulmonary atresia will not be discussed further. Pulmonary atresia with VSD and normal sized PAs is often considered a severe form of TOF and is treated in the same way as TOF. Neonates require a BT shunt. When older, the child will undergo complete repair, which consists of closure of the VSD and a transannular patch and RVOT resection, if the pulmonary valve can be used. If the pulmonary valve cannot be used, a conduit (which usually contains a valve) is placed from the RV to PA. Surgical and anesthetic considerations are similar to those for TOF.
Left-Sided Obstructive Lesions

Coarctation of the Aorta (CoA)

*Coarctation of the aorta* (CoA) is the most common left sided obstructive lesion, and is present in almost 7% of patients who have CHD. The aorta is usually narrowed just beyond the isthmus of the descending thoracic aorta, at the level of the ductus arteriosus. In cases of severe CoA, a PDA supplies blood to the lower body. PGE-1 is often required in neonates to keep their PDA open until they undergo definitive surgery. PDA closure may lead to severe LV strain, dilation, and dysfunction, or to cardiovascular collapse and death. Less severe CoA allows the PDA to close, which leads to hypertension of the upper body and upper extremities. LV function is usually preserved. This situation may be tolerated for many years. However, gradual LV hypertrophy develops; collateral arterial blood vessels grow from mammary and thoracic arteries, which are proximal to the CoA. These vessels anastomose with blood vessels distal to the CoA to provide lower body blood flow (Figure 12-12). These enlarged collateral arteries give rise to the characteristic rib notching seen on CXR. Patients may be asymptomatic; but as their upper body hypertension progresses, they may develop headaches. Untreated, adults develop early hypertensive arteriosclerosis. Some patients with severe hypertension have cerebrovascular accidents. Physical examination of a neonate with severe CoA and a closing PDA reveals poor perfusion and pulses in the lower body, delayed capillary refill, acidosis, and eventually cardiovascular collapse. Older infants and children have hypertension in the right arm, diminished femoral and pedal pulses, and a blood pressure gradient from right arm to legs that increases with more severe CoA and in the presence of inadequate collateral arterial circulation. Cardiac auscultation often reveals a soft grade I-II/VI systolic murmur at the left sternal border that radiates to the back. CXR may reveal some cardiomegaly if the patient has left ventricular hypertrophy or left ventricular dilation. Rib notching is observed in older children who develop a collateral blood supply. ECG often reveals LV hypertrophy and possibly a LV strain pattern with ST segment depression in the lateral leads. Definitive diagnosis is by echocardiography, which will delineate position and severity of CoA, whether a PDA is present, flow velocity through the CoA, and LV size, thickness, and function. Collateral arterial supply is not visualized well by echocardiography; CT, MRI, and cardiac catheterization are better tests if available. Medical therapy before surgery may include inotropic support and PGE-1 to keep the ductus arteriosus open in sick neonates who have LV dysfunction, or antihypertensive treatment with β-blocking agents or ACE inhibitors may be required.
Surgical correction of CoA is performed soon after diagnosis is made and is normally done through a left thoracotomy without CPB. Arterial pressure monitoring in the right arm is necessary. In the modern era, the usual repair involves resecting the CoA tissue and remaining ductal tissue and anastomosing the proximal and distal ends of the aorta end-to-end (Figure 12-13). Previous approaches used subclavian flap angioplasty, but this is no longer done because the recurrence rate of CoA was too high. Because the aorta is cross clamped during the repair, there is usually a 15-30 minute period where the only blood flow to the lower body is via collateral circulation. Since the anatomy of collaterals is so variable, there is a small risk of the patient being paraplegic from spinal cord ischemia after CoA surgery. With modern techniques, the risk of this devastating complication is about 0.1%. Risk factors for paraplegia include older children without a well-developed collateral arterial supply and aortic cross clamp times in excess of 30 minutes. For these reasons, the anesthesia and surgery teams use spinal cord protective measures during the repair, including cooling the patient to about 34°C; maintaining a high-normal blood pressure during cross clamping to promote blood flow through collaterals; avoiding hyperventilation, which reduces spinal cord blood flow; and sometimes administering corticosteroids. Limiting aortic cross clamp time is very important. Immediately after surgery, the patient is assessed for her/his ability to move their lower extremities. Balloon angioplasty (via a sheath in the femoral artery) can also be used to treat CoA. Expandable stents can also be placed in the narrowed area, but surgery is normally the preferred treatment in infants and young children. Transcatheter...
treatment is often used for recurrent CoA.

**Figure 12-13: End-to-End Repair of Coarctation of the Aorta**

A: Coarctation of the aorta. The figure shows where the incision will be made (dotted lines). Note that the coarcted area is where the ductus arteriosus enters the aorta. B: Coarcted segment of aorta has been resected and the ductus arteriosus has been ligated. C: The descending aorta has been anastomosed to the arch of the aorta. Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 106.

**Aortic Stenosis**

*Aortic stenosis* (AS) may occur at valvar, supravalvar, or subvalvar areas. There are many causes of AS, including congenital bicuspid aortic valve, Williams Syndrome (chromosome 7 elastin gene defect causing supravalvar AS with coronary artery involvement), and fibromuscular subaortic membrane (*Figure 12-14*). Signs and symptoms of AS range from severe obstruction in a neonate that requires PGE-1 infusion to maintain the PDA and prevent cardiovascular collapse until the AS can be corrected by surgery or in the catheterization laboratory, to completely asymptomatic patients who only have a systolic ejection murmur. After the neonatal period, patients with AS develop LV hypertrophy to maintain stroke volume. If the AS worsens, coronary ischemia may develop, and the patient may experience chest pains or syncope with exertion. This is especially the case for Williams Syndrome, where in addition to supravalvar narrowing at the sinuses of valsalva, the coronary artery ostia may be partially obstructed by abnormal tissue. Ventricular arrhythmias and sudden death may occur in untreated patients. LV dilation and failure are late signs of severe AS. Findings on physical examination include a harsh grade III-IV/VI long systolic murmur at the left sternal border. LV hypertrophy may displace the PMI downward and leftward. CXR reveals normal or increased heart size, and ECG usually indicates LV strain with ST segment depression in lateral leads. Echocardiography makes the definitive diagnosis and will accurately image the obstruction, the velocity of flow across the valve, the degree of LV hypertrophy, and the LV function.
Treatment of critical AS in neonates involves emergent balloon angioplasty in the catheterization laboratory. A balloon catheter is inserted through a sheath in the femoral or umbilical artery. PGE-1 and inotropic support must be maintained, and resuscitation drugs must be available because balloon dilation of the valve obstructs all flow to the coronary arteries for a brief time, which may cause ventricular tachycardia, fibrillation, and cardiac arrest. Older infants and children with AS undergo cardiac surgery with CPB. Many surgical treatments of AS are used, including aortic valvotomy or commissurotomy, aortic valve replacement with a mechanical valve, tissue valve, or autologous pulmonary valve (Ross procedure), pericardial patching with supravalvar AS, and resection of a fibromuscular membrane or LV outflow tract (LVOT) muscle for subaortic stenosis. The risk of coronary ischemia increases with the severity of AS. Patients with peak gradients in excess of 50-60mmHg are at high risk for coronary ischemia. Patients with Williams Syndrome are at even higher risk because of their coronary ostial obstruction. Hypovolemia, tachycardia, non-sinus rhythm, hyperdynamic LVOT contraction, severe depression of LV contractility, and hypotension all significantly increase the risk for cardiac arrest, because all of these conditions decrease stroke volume and coronary perfusion pressure or flow. Because the LV wall is thickened, the LV is at greater risk for inadequate perfusion from low arterial pressures. Low BP increases turbulent flow, which increases the resistance to blood flow across the obstruction. Prolonged fasting, hypovolemia, overdoses of inhaled or IV anesthetics that depress myocardial contractility and lower arterial blood pressure, tachycardia, excessive sympathetic stimulation that increases LVOT force of contraction, must all be avoided during anesthesia care for these patients. TEE is extremely useful after AS repair for detecting any remaining LVOT obstruction or aortic insufficiency (AI) due to the valve repair. Because the obstruction has been
relieved, hypertension is often a problem after CPB. Vasodilators, such as sodium nitroprusside, and β-blocking agents, such as esmolol, are often needed.

**Mitral Stenosis**

*Mitral stenosis* (MS) may be congenital, due to rheumatic heart disease, or residual after repair of AVC. Despite the fact that there is great anatomic variation of the mitral valve in MS, all patients have similar symptoms. As the stenosis worsens, obstruction to flow of blood into the LV causes LA enlargement, LA hypertension, and pulmonary venous congestion and hypertension. Pulmonary capillary engorgement develops, as do interstitial and alveolar edema. With increasing severity of MS, the patient develops pulmonary arterial hypertension over time. Initial symptoms of MS include tachypnea, frequent respiratory infections, and “cardiac asthma”—narrowing of small bronchioles by thickened pulmonary interstitium, and alveolar edema. Atrial arrhythmias, such as atrial flutter, can occur. Late signs include orthopnea and cachexia. Similar to AS, syncopal episodes may occur, due to inadequate systemic blood flow. Findings on physical examination include normal to slow HR and a “diastolic rumble” or a loud S3 caused by turbulent flow across the mitral valve. With significant MS, tachypnea, wheezing and rales are common. CXR reveals an enlarged LA and increased pulmonary vascular markings. ECG is consistent with LA enlargement, and once again echocardiography definitively shows the valve anatomy, the peak and mean flow velocity across the stenotic valve (from which the mean and peak gradient can be calculated), and the degree of LA enlargement. Pulmonary artery pressure can be estimated if the patient has tricuspid or pulmonary regurgitation.

Surgical treatment of MS involves CPB, aortic cross clamping, and comissurotomy or other repair of the mitral valve. Mitral valve replacement with a mechanical valve is possible, but most surgeons avoid this in growing children because it will be necessary to replace the valve as the heart grows and because long-term anticoagulation with warfarin is required. Warfarin therapy is very difficult to manage in children. The physiological considerations for anesthesia for MS surgery are similar to those for AS: HR is best kept normal to low to increase diastolic filling time across the obstruction. BP is kept normal to high to decrease the pressure gradient across the mitral valve, and hypovolemia is avoided so stroke volume can be maintained. Although pulmonary hypertension may be a problem, maneuvers to acutely lower pulmonary artery pressure (PAP) (FiO₂ of 1.0 and hyperventilation) may increase pulmonary blood flow but worsen pulmonary edema and pulmonary mechanics due to the fixed anatomic obstruction at the mitral valve. Non-sinus rhythm is poorly tolerated. TEE is extremely important for assessment of the quality of repair and cardiac function after CPB.

**Right-Sided Obstructive Lesions**

**Pulmonic Stenosis**
Pulmonic stenosis (PS) occurs at the level of the valve and at sub- or supravalvar areas in about 13% of patients with CHD. PS is normally well tolerated. Pressure overload causes compensatory RV hypertrophy. Only in the late stages of the disease, when the RV dilates and fails, does the patient have symptoms of right-sided heart failure, (hepatomegaly, peripheral edema, and ascites). Patients are often asymptomatic until relatively late in their course, when decreased RVOT flow leads to fatigue, dyspnea with exertion, and at times syncope. Physical examination of the patient reveals a harsh systolic murmur, often grade III-IV/VI, at the left upper sternal border. S2 may be soft and single. Since there is no VSD, the patient is often acyanotic. However, if there is right-to-left shunting through a PFO, neonates with severe PS may be cyanotic. A PGE-1 infusion is often needed to maintain the PDA until intervention on the pulmonary valve occurs. CXR may be normal or it may show a degree of right heart hypertrophy. ECG may be normal or show right ventricular hypertrophy. Echocardiography is the best diagnostic test and will precisely image the location and degree of obstruction, flow velocity across the valve, and the degree of RV hypertrophy. Isolated valvar PS is often treated in the catheterization laboratory by balloon dilation of the valve. The balloon is advanced through a sheath placed in a femoral vein. Neonates may require this procedure emergently; but in older infants or children, it is often performed electively. Valvuloplasty leaves varying degrees of pulmonary insufficiency (PI), which is well tolerated. Surgical approaches require CPB and involve incising the valve, with or without resection of infundibular muscle. A patch may be placed across the annulus, similar to TOF. Supravalvar PS is repaired with an autologous pericardial patch of the main PA. TEE is useful for both types of PS to determine the amount of PI and whether the PS has been relieved. Patients usually do very well after surgery. In later years they may require pulmonary valve replacement due to RV dilation and failure from longstanding PI.

Regurgitant Lesions

Aortic Regurgitation

Aortic regurgitation (AR) can be congenital or the result of balloon valvuloplasty for AS, endocarditis, or rheumatic heart disease. Aortic insufficiency (AI) usually presents in late infancy or childhood, not in the neonatal period. As the regurgitant fraction increases, LV diastolic volume and stroke volume increase, and the LV dilates over time, due to slowly developing AR. Acute AR (bacterial endocarditis) can quickly lead to LV failure. Diastolic backflow of blood into the LV decreases diastolic BP and coronary perfusion pressure, causing coronary ischemia. The ischemia leads to more LV dysfunction and ventricular arrhythmias. If not emergently addressed, acute AR may result in cardiac arrest and death. More slowly developing AR produces LV and LA enlargement, pulmonary venous congestion, tachypnea, “cardiac asthma”, dyspnea, and orthopnea. Physical examination reveals tachypnea and a long diastolic murmur at the left sternal border. Signs of low cardiac output, such as pallor, poor peripheral perfusion, and shock, may accompany acute AR. Angina may be present in patients with coronary insufficiency. Because the
diastolic runoff widens the pulse pressure; peripheral pulses are bounding, the so-called “water-hammer pulse”. CXR often shows cardiomegaly with LV and LA enlargement and pulmonary venous congestion. ECG also shows evidence of LA and LV enlargement.

Emergency surgery may be required to correct acute AR. In patients with slowly progressing AR, non-emergent surgery can be required, usually outside the neonatal period. CPB and cross clamping of the aorta allows the surgeon to use one of a number of different techniques to repair the aortic valve. Aortic valve replacement may be required. In older children and teenagers, placement of a mechanical valve is usually well tolerated, which immediately improves the patient’s condition. Infants and young children only undergo aortic valve replacement when their native valve cannot be repaired, or if a cryopreserved homograft, autograft (Ross procedure), or porcine-valved Dacron® conduit can be placed. If the aortic root is replaced, the coronary arteries must be re-implanted. Whenever possible, mechanical valves are put into smaller children. Hemodynamic goals for anesthesia in acute AR are maintenance of coronary perfusion pressures, which may require boluses of vasoconstrictor agents or continuous infusion of these drugs. Tachycardia is best avoided; myocardial contractility should be preserved with inotropes if necessary. For slowly progressive AR, faster HRs reduce the regurgitant fraction. Lower blood pressures reduce afterload and promote forward blood flow into the aorta. Hypovolemia is avoided. TEE is very useful for assessment of the repair and for determining the presence of residual defects. It is also helpful for evaluating LV function. Significant bleeding may be a problem post-CPB; adequate hemostatic blood products should be available.

Mitral Regurgitation

*Mitral regurgitation* (MR) can be congenital or from endocarditis, rheumatic heart disease, or AVC repair. As the stenosis worsens regurgitation of blood from LV into the LA produces LA enlargement, LA hypertension, pulmonary venous congestion and hypertension. The LA enlarges because a high percentage of each stroke volume is ejected backwards into the LA and increases atrial end-diastolic volume. If the mitral valve annulus enlarges, this will make it difficult for the valve leaflets to coapt and further worsens MR. Pulmonary capillary engorgement and interstitial and alveolar edema occur. Over time, severity of MR often increases, and this leads to pulmonary arterial hypertension. Initial symptoms of MR are similar to those with MS and include tachypnea, frequent respiratory infections, and “cardiac asthma”. Atrial arrhythmias, such as atrial flutter, occur. Orthopnea and cachexia are late signs of MR. Physical examination often reveals tachycardia and tachypnea, plus a regurgitant jet-induced loud and long systolic murmur at the left sternal border. The PMI is displaced downward and to the left by the LV enlargement. CXR reveals cardiomegaly and enlargement of both LV and LA. ECG demonstrates LA and LV enlargement and may reveal atrial arrhythmias, such as atrial flutter. Echocardiography demonstrates mitral valve anatomy in detail, along with a qualitative estimate of the regurgitant fraction and the size and function of the LV and LA. During systole color-flow Doppler
echocardiography reveals reversal of blood flow in one or more pulmonary veins in patients with severe MR.

Correction of MR requires repair of the valve and decreasing the size of the annulus. As with MS, mitral valve replacement is avoided in growing children whenever possible. Hemodynamic goals during anesthesia include maintaining a high-normal HR to decrease ejection time and reduce the amount of blood regurgitated into the atrium. Avoiding hypovolemia helps maintain adequate stroke volume. Reducing the arterial blood pressure decreases SVR and promotes forward blood flow. Finally, LV contractility should be maintained at normal levels. Following CPB, it is important to examine the heart for evidence of residual MR or MS and to help decide if further correction is needed.

**Tricuspid Regurgitation**

*Tricuspid regurgitation* is most often caused by Ebstein’s anomaly, a congenital malformation of the tricuspid valve in which the valve annulus and leaflets are improperly placed into the body of the RV. The tricuspid valve itself is also malformed and has shortened chordae, resulting in varying degrees of TR (Figure 12-15). Neonates with severe TR caused by Ebstein’s anomaly have poor forward blood flow into the RV and PA. For perfusion of the LA and LV, there must be an ASD or large PFO, which causes cyanosis. Because PA blood flow is low, a PDA is commonly required for maintenance of oxygenation. The electrical conduction system is also abnormal. In the neonatal period, severe Ebstein’s anomalies are often fatal. Less severe degrees of TR can occur, and these patients may present in later childhood or even as adults. The TR causes RA enlargement and signs and symptoms of right heart failure, such as hepatomegaly, pleural effusions, and peripheral edema. The RV may also enlarge because the regurgitant fraction is high. Atrial dysrhythmias, such as atrial flutter and supraventricular tachycardia, are very common. Physical examination reveals the loud systolic murmur of TR at the right sternal border and RV impulse displacement; an enlarged liver, ascites, peripheral edema, and tachypnea may be evident. CXR reveals cardiomegaly; the pulmonary vascular markings are often normal or slightly increased; and there may be evidence of pleural effusion. ECG frequently demonstrates atrial premature contractions or atrial arrhythmias. Echocardiography accurately diagnoses malformation of the valve, the degree of regurgitation, presence of a PFO or ASD, and RV and LV size and function. Surgery entails a complicated repair of the tricuspid valve and plication of the arterialized portion of the RV. Anesthetic goals include: 1) promoting blood flow into the RV and PA by lowering PVR; 2) using high FiO₂; and 3) mild hyperventilation. Atrial arrhythmias must be diagnosed and aggressively treated; external or internal cardioversion may be required. After CPB, TEE of the heart is very important for assessing the repair. PVR must be kept low to optimize cardiac output.
Mixing Lesions

Mixing lesions are major malformations of the heart and great vessels that allow mixing of pulmonary and systemic blood. With these lesions, there are varying degrees of cyanosis. To oxygenate blood and provide adequate systemic oxygen delivery, both right-to-left and left-to-right shunting of blood must occur. Three of the most common and important mixing lesions will be discussed: dextro-transposition of the great arteries (d-TGA), total anomalous pulmonary venous return (TAPVR), and truncus arteriosus.

Transposition of the Great Arteries

d-TGA, also known as transposition of the great vessels, is a common form of CHD and occurs in about 3.6% of all CHD patients; it presents in the neonatal period. Early surgery is required for survival. The aorta arises abnormally from the RV and the PA from the LV. Approximately 20-25% of these patients also have a perimembranous VSD (Figure 12-16). Oxygenated pulmonary venous blood returns to the left side of the heart, i.e., to the LA and LV, where it is ejected into the PA. Unoxygenated systemic venous blood returns to the RA and RV and is ejected into the aorta. This parallel circulation (rather than the normal series circulation) prevents oxygenated blood from reaching the systemic circulation unless there is a communication at the atrial, ventricular, or PDA
level. These communications let oxygenated blood to enter the RA, RV, and aorta, which is necessary for survival. Without these communications, the patient is profoundly cyanotic and will only survive a few days if this situation is not corrected or palliated. These infants are profoundly cyanotic at birth, and 100% oxygen does not relieve the hypoxemia. Despite the severe hypoxemia, these patients show little evidence of respiratory distress. This observation led to the “hyperoxia test”, which differentiates cardiac from respiratory disease. The test consists of measuring SpO₂ or PaO₂ before and after the patient breathes a FiO₂ of 1.0. Despite the high FiO₂, the SPO₂ of patients with d-TGA does not increase above 85% (PaO₂ below 45mmHg). Patients with pulmonary disease, on the other hand, often increase their oxygenation significantly. Physical examination reveals cyanosis without initial respiratory distress and a soft grade I-II/VI systolic murmur from a PDA or VSD, if present. CXR reveals a narrow mediastinum due to reversal of aorta and PA positions in the mediastinum (“Egg-on-a-String” appearance). The lung fields are black, due to reduced pulmonary blood flow. ECG is non-specific but often shows sinus tachycardia. The echocardiogram makes the definitive diagnosis by showing the PA and its branches arising from the LV and the aorta with its sinuses arising from the RV (Figure 12-3D). Those patients who also have a VSD frequently have sufficient oxygenation to survive the neonatal period without further intervention. Because only 20-25% of patients with d-TGA have a VSD, the remainder requires PGE-1 to maintain the PDA, most of whom will require a balloon atrial septostomy at the bedside under echocardiographic guidance. To do this, a balloon-tipped catheter is introduced through the umbilical or a femoral vein into the LA. The balloon is inflated and vigorously yanked to tear a larger hole in the atrial septum to increase mixing of oxygenated and unoxygenated blood. This immediately increases the SpO₂, usually into the 80-90% range. After a septostomy, it is generally possible to discontinue PGE-1 and allow the patient to recover and feed before surgery.
Surgery for d-TGA is now exclusively the arterial switch operation (ASO). During CPB and aortic cross clamping, the aorta and PA are transected, and their positions switched to restore normal anatomic relationships. In addition, the coronary arteries are translocated with a button of aortic tissue to their proper anatomic location on the aorta, which now arises from the LV. The VSD and ASD are also repaired (Figure 12-17). Coronary artery anatomy is variable, which presents challenges for the surgeon who must translocate these vessels without twisting, kinking, or obstruction them. The ASO is normally performed in the first several weeks of life before the LV becomes deconditioned, which occurs if there is no VSD and the surgery is delayed beyond 4-6 weeks of age. Deconditioning occurs because the pulmonary artery pressure and resistance decrease over time, making it easier for the left (pulmonary) ventricle to eject blood. When deconditioning occurs, the deconditioned LV will have difficulty pumping blood against the higher systemic pressures and resistances when the vessels are switched. Patients who have large VSDs maintain RV and LV pressures equal to systemic pressure, which allows surgery to be delayed until 2-3 months of age. Those with a VSD may develop excessive pulmonary blood flow and early pulmonary vascular disease as PVR decreases. Challenges for the anesthetist include treating low
post bypass cardiac output, which is often the result of coronary obstruction, a deconditioned LV, or inadequate myocardial protection during surgery. LA pressure is often monitored and maintained below 10mmHg to avoid excessive preload to the LV; excessive preloads may be poorly tolerated. Echocardiography, either epicardial or TEE, is an important tool for assessing myocardial function and diagnosing any residual issues from the surgery. Mortality after ASO is low and decreasing. The majority of patients require no further intervention for the remainder of their lives. However, some patients experience coronary artery problems, aortic insufficiency, or supravalvar pulmonic stenosis after surgery.

**Figure 12-17: Anatomy of d-TGA After Arterial Switch Operation**

![Anatomy of d-TGA After Arterial Switch Operation](source)

**Total Anomalous Pulmonary Venous Return**

*Total anomalous pulmonary venous return* (TAPVR) occurs when all four pulmonary veins connect abnormally to the right side of the heart. The three major types are: 1) Supracardiac, where confluence of the pulmonary veins connects via a vertical vein to the innominate vein; 2) Cardiac, where the pulmonary veins connect to the coronary sinus and drain to the RA; and 3) Infracardiac, where the pulmonary veins drain downward through an abnormal vertical vein and connect to the inferior vena cava or hepatic veins (**Figure 12-18**). There must be an atrial level communication (ASD or large PFO) for oxygenated blood to go into the left side of the heart to
maintain systemic cardiac output and systemic oxygen delivery.

The signs and symptoms of TAPVR at presentation depend on the degree of pulmonary vein obstruction and the size of the atrial level communication. Severe pulmonary vein obstruction, with minimal communication at the atrial level, leads to marked pulmonary venous engorgement, profound cyanosis, and death if not treated emergently by surgery. Lesser degrees of pulmonary venous obstruction occur in patients who have adequate atrial communication. These patients have mild to moderate cyanosis and variable amounts of respiratory distress during the neonatal period. In general, the risk of obstruction is high in patients with infracardiac TAPVR; supracardiac TAPVR has an intermediate risk; and cardiac TAPVR has the lowest risk of obstruction. CXR reveals a small heart and pulmonary venous engorgement. The CXR of patients with supracardiac TAPVR often shows a “figure of 8” or “Snowman” appearance due to the abnormal vertical vein on the left side of the mediastinum (Figure 12-1D).

Figure 13.18: Anatomy of Total Anomalous Pulmonary Venous Return

Source: Andropoulos and Gottlieb; Congenital Heart Disease, Anesthesia and Uncommon Diseases, 6th Ed., Fleisher L., (ed.) 2012, p. 103.

Urgent surgery may be required if supportive care (tracheal intubation, pulmonary ventilation, and inotropic support) does not effectively restore adequate oxygen delivery to tissues. CPB with deep hypothermic circulatory arrest (DHCA—see below) is necessary for this surgery. The abnormal vertical vein is ligated, divided, and the pulmonary venous confluence anastomosed to the back of the LA. The ASD is closed. Pulmonary hypertension is commonly encountered in the immediate postoperative period, which usually necessitates a period of mechanical ventilation, sedation, and pulmonary vasodilator therapy with sildenafil, inhaled nitric oxide (iNO), or nebulized prostacyclin, if available. Long-term outcome is usually good if the patient survives the neonatal period and surgery. A few patients have recurrent pulmonary vein obstruction and require further treatment.
**Truncus Arteriosus**

*Truncus arteriosus* consists of a single arterial trunk that arises from the left and right ventricles and gives rise to both the aorta and the PA ([Figure 12-19](#)). A large subarterial VSD is always present, and there is a single truncal valve that opens into the truncus. The truncal valve may be abnormal and may have anywhere from two to six leaflets plus stenosis, regurgitation, or both. The three major anatomic subtypes of truncus arteriosus are: Type I: the truncus arteriosus gives rise to the aorta and a short main PA that branches into left and right PA; Type II: there is no main PA, but the left and right PAs arise from the back of the truncus arteriosus; and Type III: there is no main PA, but the left and right PAs arise from the sides of the truncus arteriosus. Truncus arteriosus results in unique pathophysiology - the systemic, pulmonary, and coronary circulations exist in parallel and have profound interactions that affect cardiac and pulmonary function. At birth, pulmonary vascular resistance is elevated and Qp:Qs is near 1:1, resulting in mild cyanosis from mixing of oxygenated and unoxygenated blood without excessive pulmonary blood flow; aorta and coronary flow are relatively well preserved. As PVR decreases over the first days and weeks of life, Qp:Qs increases to 2-4:1, resulting in pulmonary over-circulation, pulmonary edema, and signs of CHF. The decrease in PVR allows a “steal” of blood flow away from the aorta and coronary arteries toward the pulmonary circulation. Diastolic BP is low, which frequently causes coronary insufficiency, myocardial ischemia, low cardiac output, ventricular arrhythmias, and death, if not treated. Physical examination demonstrates tachypnea, tachycardia, poor peripheral perfusion; cardiac examination reveals a single S2 and a grade II-IV/VI systolic murmur from the VSD or AS. There is often a S3 heart sound from AR or increased diastolic flow into the LV. CXR reveals cardiomegaly with increased pulmonary vascular markings. ECG may reveal sinus tachycardia, LV enlargement, and LV strain with ST segment changes in the lateral leads. Echocardiography produces a very accurate picture of the anatomy of the truncus arteriosus and pulmonary arteries, as well as the anatomy, flow, regurgitation, or stenosis of the truncal valve. The size and configuration of the VSD and the size and function of the ventricles are also imaged. If TA goes untreated and the patient survives, her/his CHF will be followed within weeks to months by pulmonary vascular disease, due to exposing the pulmonary circulation to high flows and pressures. A fixed shunt usually develops within the first year of life and makes the condition inoperable. Right-to-left shunting results in increasing cyanosis.
Correction of truncus arteriosus occurs in the neonatal period using CPB and aorta cross clamping. The repair consists of closing the VSD with an autologous pericardial patch, separating the PA’s from the truncus arteriosus, and inserting a RV-to-PA conduit that contains a valve. Problems with the truncal valve are addressed, and the ASD may be closed. This is a complex operation and major issues for the anesthetist post-CPB include bleeding, pulmonary hypertension, and myocardial dysfunction. If the patient survives the operation and the neonatal period, long-term outcomes are generally good, but subsequent cardiac surgery will be required as the patient grows to replace the RV-to-PA conduit. The truncal valve may also require further corrective procedures.

**Single Ventricle Lesions**

The most common single ventricle lesions occur with tricuspid atresia, which causes hypoplasia of the right heart, and hypoplastic left heart syndrome. Surgical treatment of single ventricle lesions is done in the neonatal period to provide stable pulmonary or systemic blood flow. Then, at age 3-6 months a superior cavopulmonary connection is created (bidirectional Glenn operation). Finally, at 2-4 years of age, a total cavopulmonary connection (Fontan operation) is performed. In general patients with single ventricles require multiple surgical and catheterization procedures and intensive use of resources. They experience much higher mortality and complication rates than patients with two-ventricle. The reader is referred to the Bibliography for extensive information about anesthesia for patients with single ventricles.

**Table 12-3** summarizes the desired hemodynamic goals for the major classes of cardiac lesions, based on the pathophysiological considerations discussed above.
Table 12-3: Desired Hemodynamic Goals for Major Cardiac Lesions

<table>
<thead>
<tr>
<th>Lesion</th>
<th>HR</th>
<th>Contractility</th>
<th>Preload</th>
<th>SVR</th>
<th>PVR</th>
<th>FiO2</th>
<th>PaCO2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left sided obstruction: AS</td>
<td>↓</td>
<td>↓*</td>
<td>↑↑↓</td>
<td>↑↑</td>
<td>---</td>
<td>---</td>
<td>---</td>
</tr>
<tr>
<td>Left sided regurgitation: aortic/mitral</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
<td>↓↓</td>
<td>---</td>
<td>---</td>
<td>---</td>
</tr>
<tr>
<td>Left-to-right shunts: large VSD in infants</td>
<td>↓</td>
<td>↑</td>
<td>↓↓</td>
<td>↓↓</td>
<td>↑</td>
<td>↓</td>
<td>↑</td>
</tr>
<tr>
<td>Right-to-left shunts: TOF</td>
<td>↓↓</td>
<td>↓↓</td>
<td>↑↑</td>
<td>↑↑</td>
<td>---</td>
<td>↑↑</td>
<td>---</td>
</tr>
</tbody>
</table>

HR, heart rate; SVR, systemic vascular resistance; PVR, pulmonary vascular resistance; AS, aortic stenosis; VSD, ventricular septal defect; TOF, tetralogy of Fallot. Changes from baseline: ↓, mild decrease; ↓↓, moderate decrease; ↑, mild increase; ↑↑, moderate increase; ---, no change. *Do not decrease if depressed at baseline; do not increase with depressed baseline contractility with high preload

Cardiopulmonary Bypass

Basic Aspects

The components of a CPB circuit are displayed in Figure 12-20. The main components are a bypass machine with roller pumps for aortic perfusion, cardiotomy suction, and cardioplegia, an oxygenator-heat exchanger, a venous reservoir, tubing for return of venous blood and for aortic perfusion, and cannulae for aortic perfusion and drainage of venous blood. Monitors of flow, pressure, and blood oxygen saturation are incorporated into the circuit. In addition, monitors are incorporated into the venous reservoir and arterial perfusion side of the circuit to detect entrainment of air. An arterial filter is also incorporated to remove thrombi and trap air. An oxygen-air blender and an anesthetic vaporizer are incorporated into the sweep gas line to adjust the FiO2 and add volatile anesthetic. The basic components of pediatric circuits are similar to adult CPB circuits, however, because of the small and variable sizes of pediatric patients, several different sizes of venous reservoirs and tubing are necessary for CHD surgery. Because the blood volume of infants and children is only 75-90ml/kg and the minimum volume of a CPB circuit is 250-350ml, smaller infants and children usually require priming of the CPB circuit with blood [packed red blood cells (PRBC) with or with or without fresh frozen plasma (FFP), or whole blood] to prevent extreme dilution of hematocrit and coagulation factors. Configurations of these
systems vary greatly, but in general the patient must be larger than 10-15 kg before it is possible to prime the circuit without blood (bloodless prime). The composition of fluid used to prime the system in small infants is often blood plus some isotonic balanced salt crystalloid solution, plus heparin, usually 2000-3000 units. The pH may be adjusted to normal with sodium bicarbonate. Larger patients have a crystalloid prime with heparin added, and the pH is adjusted to normal. Modern manufacturing has significantly reduced costs of the CPB circuits, and pre-packaged circuits with appropriate sized tubing, oxygenator, and venous reservoirs are available for infants and children at low cost.

Figure 12-20: Basic Components of a Cardiopulmonary Bypass Circuit

Phases of CPB

The Pre-Bypass Period involves induction of anesthesia, tracheal intubation, insertion of vascular access and monitoring lines (including peripheral IVs, arterial catheter, and central venous catheter, if used). A TEE probe, if used, is inserted and the cardiac anatomy confirmed. Antibiotics and corticosteroids (if used) are administered, and antifibrinolytic agents started (tranexamic acid or aminocaproic acid), if used. Sternotomy is accomplished; a subtotal thymectomy is normally done to increase access to the heart and increase space in the mediastinum; the pericardium is incised; and the heart and great vessels are dissected and prepared for cannulation.

The Cannulation Period starts with the surgeon placing purse string sutures in the root of the aorta to prepare for aortic cannulation. At this point heparin, 300-400 units per kg of body weight, is given to ensure profound anticoagulation during bypass. Heparin must be given through a secure catheter, preferably a central line that has good blood return; alternatively the surgeon can administer the heparin directly into the heart. The activated clotting time (ACT) is commonly used to measure heparin effect; the baseline ACT is normally 120-150 seconds. The target ACT during bypass is 480 seconds. There must be adequate anticoagulation before beginning bypass to prevent clotting of the CPB circuit, which, if it occurs, is a fatal and preventable mistake. The surgeon will then place purse string sutures for insertion of the venous cannulae; most often this is for both superior vena cava (SVC) and inferior vena cava (IVC) cannulae to allow intracardiac access. Then, the aortic cannula is placed, the arterial perfusion line is connected and de-aired, and the correct position of the aortic cannula confirmed by measuring the pressure and by infusing a small amount of volume from the venous reservoir. Next, the IVC and SVC cannulae are placed and connected to the venous return tubing. Cannulation in small infants is often a time of significant hemodynamic instability; some surgeons initiate CPB only after the SVC cannula is placed. Figure 12-21 shows cannulation in an infant for cardiac surgery.

The CPB Initiation Period begins when surgeon, perfusionist, and anesthetist agree that adequate anticoagulation has been achieved, cannulae and tubing are connected correctly, and the pump is ready to assume full flow support. CPB is initiated at low flows; the perfusionist assesses venous drainage by checking venous reservoir volume and the surgeon checks for emptying of the heart. CPB flow is increased over the first minute or so to full flow, normally 100-150ml/kg/min for patients <10kg, and 2.4-2.8l/min/m² for larger patients. Mean arterial pressure (MAP) is assessed, and target MAP ranges are achieved (30-50mmHg for neonates and infants, 40-50mmHg for older children, 50-60mmHg for teenagers and adults). Additional IV anesthetics or opioids are administered into the CPB circuit and volatile anesthetic is added to the sweep gas if indicated. The sweep gas FiO₂ is adjusted to avoid hyperoxia or hypoxia. Normally the FiO₂ is about 0.25-0.30. Blood gases and venous oxygen saturations are determined soon after initiating CPB to ensure adequate delivery of oxygen. Mechanical ventilation is stopped as soon as full CPB is
achieved. The anesthetist assesses the patient’s face, scalp, and fontanelle for signs of superior vena cava (SVC) obstruction, which include bulging fontanelle (in babies), plethora, and cyanosis of the scalp and face.

The CPB Cooling Period starts when the team is satisfied that CPB has been successfully instituted. Cooling is initiated via the heat exchanger of the bypass machine. Hypothermia is frequently used to protect the myocardium, brain, and other organs because hypothermia greatly reduces oxygen consumption and cellular metabolic requirements. Mild hypothermia is 30-34°C and is used for many simple repairs, such as ASD and VSD. Moderate hypothermia is 22-30°C and is used for moderately complex repairs, such as TOF, ASO or CAVC. Deep hypothermia is 17-22°C and is used for complex surgery, such as TAPVR or truncus arteriosus repair that may require DHCA (see below).

Figure 12-21: Aorto-Bicaval Cannulation for Bypass in an Infant

Aorta = aortic cannula; SVC = superior vena cava cannula; IVC = inferior vena cava cannula.

The Aortic Cross Clamping Period begins after cooling when a clamp is placed on the aorta above the coronary arteries. Cardioplegia solution is infused into the aortic root through a separate small cannula to create diastolic arrest of the heart. This is accomplished by infusing a 20meq/l potassium (K+) solution into the coronary arteries to cause electrical silence. The ECG should be isoelectric within no more than 1-2 minutes after cross clamping occurs. Ice is often packed around the heart to maintain low myocardial temperatures. Cardioplegia can be re-infused at 20-minute intervals to maintain a quiet ECG; recurrence of ECG should be communicated to the
surgeon and additional cardioplegia given as needed. Aortic cross clamping is required for any surgery within the heart, such as ASD, VSD, TOF, ASO repairs. Aortic cross clamp time is also known as “ischemic time” because the myocardium is not perfused; prolonged periods of cross clamping cause myocardial ischemia, dysfunction, and possibly cardiomyocyte death. “Long” cross clamp times vary by institution and surgeon, but generally if cross clamping lasts longer than 90-120 minutes, the risk for myocardial dysfunction increases.

**Deep Hypothermic Circulatory Arrest** is utilized for some complex neonatal cardiac repairs, such as TAPVR or truncus arteriosus, to reduce blood return to the field and enable the surgeon to visualize and repair tiny structures accurately. At deep hypothermic temperatures, the CPB flow is turned off and all blood is drained from the heart. This leaves a quiet, bloodless field for the critical part of the repair. Safe periods of DHCA are generally thought to be up to 30-40 minutes; longer periods of DHCA are associated with brain injury and problems with long-term neurodevelopmental.

The **CPB Warming Period** begins when the major portion of the intracardiac repair is complete. The aorta is unclamped after the surgeon has performed de-airing maneuvers to reduce/remove air that entered the cardiac chambers while they were open to atmosphere during cross clamping. The heart should start beating spontaneously when the coronary arteries are again perfused with oxygenated blood and the high-K+ cardioplegia solution is washed from the myocardium. The patient is warmed to a nasopharyngeal temperature of about 36°C. The remainder of the cardiac repair is completed during warming. The surgeon places catheters in the RA or LA for pressure monitoring and vascular access; temporary cardiac pacing wires are placed on the atrium and ventricle, if needed. The desired hematocrit is achieved by either hemofiltration, adding blood to the venous reservoir, or both. Cardiac rhythm is assessed. If the rhythm is not normal sinus at the desired rate, temporary cardiac pacing is begun. If needed, inotropic, vasoconstrictor, and vasodilator drug infusions are started during rewarming once the ECG has returned to a near normal configuration. The lungs are inflated gently, the endotracheal tube is suctioned, and the lungs are inspected for bilateral inflation; any atelectatic areas of lung are expanded with several vital capacity breaths, additional suctioning, and possibly nebulized bronchodilators. Desired FiO₂ and ventilator settings are instituted. Calcium chloride is often added to the CPB circuit at this point to achieve normal ionized calcium levels, particularly in neonates.

The **Separation from CPB Period** begins with weaning of CPB flow and gradual impedance of venous return by the perfusionist. During this time the surgeon assesses cardiac function and filling and directs the perfusionist to further reduce flow and venous return until the CPB pump is turned off. Immediately after the patient is separated from CPB, blood pressure, SpO₂, filling pressures (central venous pressure, LA and RA pressures) are assessed, as well as myocardial contractility and ECG rhythm. A TEE examination of the heart is performed to assess the results of
the surgery and determine if there are residual defects, e.g., a residual VSD, that necessitates return to CPB for repair. Cardiac function is assessed. An arterial blood gas is checked to assess oxygenation, ventilation (CO₂), and acid-base status. The most common reasons for being unable to separate from CPB with good cardiac function and output are myocardial ischemia (due to prolonged cross clamp time) or residual cardiac defect, e.g., RVOT obstruction after TOF repair. In addition, air bubbles occasionally lodge in the right coronary artery during weaning, which results in RV dysfunction and ST segment elevation. This usually resolves over a short time with increasing the arterial blood pressure and flow through the coronary arteries. The most common inotropic agents used around the world are milrinone 0.25-0.75mcg/kg/min; epinephrine 0.02-0.1mcg/kg/min, and dopamine 3-10mcg/kg/min. Many patients undergoing simple corrective two ventricle repairs, i.e. VSD and ASD, require no inotropic agents after CPB. Once the team is satisfied with the patient’s hemodynamic status, TEE results, and blood gas, the surgeon requests that protamine be administered to reverse the heparin effects, usually in a dose of 1.0-1.3 mg of protamine per mg of heparin (100 units heparin = 1 mg) given for the original dose. The ACT is checked 3-5 minutes after the protamine is given to ensure that the heparin effect is fully reversed. After protamine, the ACT should be within about 10% of baseline.

The Post-CPB Period extends from the administration of protamine until admission to intensive care unit (ICU) and handoff of care to the ICU team. In many children, bleeding is a primary concern following CPB. The most common cause is bleeding is from suture lines or from small vessels. During surgery the surgeon must meticulously search for and occlude any source of bleeding with an electrocautery, additional sutures, or other hemostatic drugs or techniques that are applied topically to the bleeding area. CPB itself causes problems with coagulation in several ways, the most important being dilution by the CPB prime of coagulation proteins and platelets in <8-10kg patients. Platelets also bind to components of the CPB circuit and tubing, are degranulated, and become inactive. Finally, fibrinolysis occurs by activation of the coagulation cascade, by contact of blood with foreign surfaces, and by the inflammatory response to bypass. In small infants, administration of platelets is the first line of therapy and will resolve most post-CPB bleeding, especially since platelets are suspended in plasma that contains some coagulation proteins. A platelet dose of 1 random donor unit per 5kg patient weight increases the platelet count by about 50,000 per microliter (See Chapter 4). Inadequate fibrinogen is the next most common deficiency in infants, and cryoprecipitate 15-20ml is effective in restoring normal levels of fibrinogen in most infants. Finally, coagulation proteins can be deficient; FFP is often utilized after CPB to correct these deficits. If available, the use of fresh whole blood is far preferable to component therapy for bleeding. Once bleeding is resolved, mediastinal drainage tubes are placed, and the sternum is closed. Many older patients weighing more than 20-30kg have simple cardiac surgery without blood transfusion. If cell salvage is available, residual blood in the CPB circuit at the end of bypass can be washed, filtered, and returned to the patient. However, this blood has few, if any, clotting proteins. After sternal closure, the patient is transported fully
monitored to the ICU and a full report and handoff given to the ICU team. Many patients undergoing simple cardiac repairs have their tracheas extubated in the operating room or shortly after arriving in the ICU.

**Anesthetic Approach to Surgery for Congenital Heart Disease**

**Pre-Surgical Planning**

Ideally a multidisciplinary conference with cardiologists, surgeons, anesthetists, and intensivists is held to present and discuss all cardiac surgery patients. The history and physical examination findings, CXR and laboratory results, and echocardiography and other images (such as cardiac catheterization, MRI, and CT) are presented. A surgical plan is proposed and discussed, and suitability of the patient for surgery is assessed. The group decides upon a final surgical plan. This plan may be altered if the surgeon’s findings at operation differ from the presurgical conference data. The anesthetist plays an important role in this conference and should contribute to the discussion of perioperative risk and planning, particularly where non-cardiac medical problems are concerned.

**Selection of Anesthetic Techniques and Agents**

The preferred approach for designing an anesthetic for cardiac surgery is to devise a set of hemodynamic and ventilatory goals for each patient that are based on the patient’s cardiac anatomy and on the pathophysiology of their cardiac lesion. Only when all of the information has been evaluated can an intelligent anesthetic plan by devised. The information in Table 12-3 summarizes goals for the major categories of CHD.

Next, anesthetic drug selection is considered. The drugs chosen should have hemodynamic and myocardial effects that are most likely to allow the anesthetist to achieve the desired hemodynamic goals. For example, a patient with severe AS presenting for aortic valve replacement should not receive a large dose of propofol for induction of anesthesia because this drug will lower blood pressure, increase the AS gradient, worsen coronary perfusion, and possibly lead to cardiovascular collapse. All volatile anesthetic agents depress myocardial contractility to some extent if their concentrations are high enough, but there are significant differences among the available agents. For example, halothane depresses myocardial contractility more than isoflurane, sevoflurane, and desflurane in pediatric patients, most particularly in infant <6 months of age. If at all possible, halothane is avoided in infants with CHD or in other patients with CHD who have decreased myocardial function. If it must be used, the inspired halothane concentrations should be limited to <1 MAC. Sevoflurane should be used for inhaled induction of anesthesia in these patients when possible because it better preserves myocardial contractility. Isoflurane has even less effect on myocardial contractility and is a good choice of volatile anesthetic for maintenance of anesthesia for most CHD surgery. It is a poor choice for induction
of anesthesia because it causes laryngospasm and bronchospasm. Generally, opioids such as fentanyl have minimal effect on myocardial function and are well tolerated at a variety of doses. Ketamine is a particularly useful drug and may be used for IV or intramuscular induction of anesthesia; its vagolytic and sympathomimetic effect preserves cardiac output in most cases. Table 12-4 summarizes the major hemodynamic effects of common anesthetic agents.

### Table 12-4: Anesthetic Agents and Cardiovascular Effects

<table>
<thead>
<tr>
<th>Agent</th>
<th>Myocardial Contractility</th>
<th>Heart Rate</th>
<th>Arterial Blood Pressure</th>
<th>Cardiac Output</th>
<th>Pulmonary Vascular Resistance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fentanyl</td>
<td>---</td>
<td>↓↓</td>
<td>---</td>
<td>↓</td>
<td>↑</td>
</tr>
<tr>
<td>Midazolam</td>
<td>---</td>
<td>↓</td>
<td>↓</td>
<td>↓</td>
<td>↓</td>
</tr>
<tr>
<td>Ketamine</td>
<td>--- or ↓</td>
<td>↑</td>
<td>↑</td>
<td>↑</td>
<td>---</td>
</tr>
<tr>
<td>Propofol*</td>
<td>↓</td>
<td>↓</td>
<td>↓</td>
<td>↓</td>
<td>---</td>
</tr>
<tr>
<td>Etomidate¶</td>
<td>---</td>
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<td>---</td>
<td>---</td>
<td>---</td>
</tr>
<tr>
<td>Dexmedetomidine</td>
<td>---</td>
<td>↓↓</td>
<td>↓</td>
<td>↓</td>
<td>↓</td>
</tr>
<tr>
<td>Volatile agents</td>
<td>↓</td>
<td>--- or ↑</td>
<td>↓</td>
<td>--- or ↓</td>
<td>↓</td>
</tr>
</tbody>
</table>

Changes from baseline: ↓ = mild decrease; ↓↓ = moderate decrease; ↑ = mild increase; ↑↑ = moderate increase; --- = no change. *Avoid propofol in preload/afterload dependent patients: L side obstruction, dilated cardiomyopathy, decreased LV function; ¶etomidate preferred in these patients

### Vascular Access, Monitoring, and Equipment for Cardiac Surgery

Secure peripheral IV access is essential for CHD surgery. In infants and children the saphenous vein is a reliable relatively large vessel into which at least a 22g catheter can be inserted in neonates and infants. Dorsal hand, wrist, or antecubital veins are large and thick-walled enough to accept large IV catheters. Arterial access is required for all CPB surgery and for most non-CPB CHD surgery, such as repair of CoA. This catheter provides crucial information on beat-to-beat arterial blood pressure and for measuring blood gases and pH. The radial artery is preferred in most cases. However, the femoral artery is large and well collateralized and can be used when the radial artery cannot be accessed. Percutaneous central venous access is utilized for many CHD surgeries, with the right internal jugular vein or femoral veins being the preferred site for placing a double-lumen catheter. Many simple surgeries, such as ASD and VSD, do not require central venous access; two large peripheral IVs suffice for many of these cases. Finally, the cardiac surgeon can place a transthoracic atrial catheter through the chest wall during the warming period on CPB that can be used for several days after surgery. The approach to vascular access varies widely according to the institutional setting, surgeon and anesthetist’s preference and experience, and resources available. Chapter 2 presents a complete discussion of vascular access.
and monitoring.

Monitoring for CHD surgery also includes at least a 3-lead ECG that accurately displays cardiac rhythm and allows rapid diagnosis and treatment of arrhythmias, which are very common during CHD procedures. An oscillometric blood pressure cuff is an important monitor for determining systolic, diastolic, and mean arterial pressures at least every 5 minutes before arterial access is obtained and for monitoring these pressures should the arterial catheter fail during surgery. Pulse oximetry is crucial, not only for routine monitoring of oxygenation, but also to detect the frequent changes in desaturation that occurs with surgical interventions. The plethysmograph function of the pulse oximeter is also important for monitoring peripheral perfusion. While vasoconstriction, low temperature, and low cardiac output will affect this signal, complete loss of the pulse oximeter signal must be addressed immediately. Temperature monitoring is crucial, especially during hypothermic CPB. Two sites: nasopharyngeal, which reflects brain temperature; and rectal, which reflects core organ temperature, are preferred. If it is only possible to measure one temperature, nasopharyngeal is preferred because of the high blood flow to the brain in infants and need to protect this vital organ. Use of a capnograph that work appropriately with an infant’s small tidal volumes is very important, both for standard monitoring of ventilation and for detection of the variable end-tidal to arterial CO₂ gaps commonly seen in cyanotic infants. A large gap (end-tidal CO₂ that is much lower that the arterial CO₂) signifies low pulmonary blood flow, low cardiac output, or both. Anesthetic gas monitoring is also desirable. The anesthesia machine ventilator must be capable of delivering small tidal volumes in the pressure control mode.

**Early Tracheal Extubation After Congenital Heart Surgery**

VSD, ASD, and PDA comprise 50% of congenital heart disease. The tracheas of most of these patients can be extubated at the end of surgery. The benefits of early tracheal extubation are also becoming more widely accepted for lesions such as TOF, AS, CoA, and other two-ventricle corrective surgeries. In general, if the patient did not require mechanical ventilation preoperatively, the surgery was uncomplicated, and there are no residual anatomic defects, there is no significant ongoing bleeding, no significant inotropic agent requirement or dysthyemia requiring pacing or drug therapy, and there is no significant pulmonary hypertension, the patient is a candidate for early tracheal extubation, either in the operating room or within the first 2-4 hours after admission to ICU. Limiting the doses of long acting fixed agents, i.e., fentanyl or morphine, and full reversal of neuromuscular blockade are important preparations for tracheal extubation. The anesthetist must also plan for the patient’s analgesic needs following tracheal extubation. Whenever possible, anesthetists must be available in the immediate postoperative period to assist with early airway management of extubated patients.
Anesthesia for Cardiac Catheterization

Cardiac catheterization, as noted above, can be used diagnostically to delineate anatomy and physiology, including Qp:Qs, intracardiac pressures and oxygen saturations, cardiac function, and the degree of pulmonary hypertension and response to pulmonary vasodilators. However, cardiac catheterization is used far less often for diagnosis in the modern era because echocardiography suffices for surgical planning for most patients. Either the cardiologist, or anesthetist provides sedation during diagnostic catheterization. IV ketamine, propofol, dexmedetomidine, opioids, and midazolam, can all be used to sedate these patients. In addition, the anesthetist can provide general anesthesia by endotracheal tube or laryngeal mask airway, if needed. Whatever the sedation or anesthetic technique used, the goals for diagnostic catheterization are the same: provide a steady baseline state from which valid hemodynamic and oxygen saturation data can be obtained. This normally means providing the lightest level of anesthesia possible, a FiO₂ of 0.21, and a blood pressure, HR, and ventilation (end-tidal CO₂) that are as close to the patient’s awake baseline values as possible. Because the most stimulating and painful part of cardiac catheterization is gaining vascular access, usually via the femoral vessels, a brief period of deeper anesthesia plus local anesthetic infiltration of the groin area are sufficient. Once the vessels are catheterized, the anesthetic level is lightened to achieve the hemodynamic and ventilatory goals.

Cardiac catheterization is increasingly used for interventional procedures, such as aortic valve balloon angioplasty in neonates who have critical AS, for ASD and PDA closure, and for dilation and stenting a CoA. For these procedures, a cardiologist may sedate some patients, but in many institutions an anesthetist provides this care. Anesthetic management of patients undergoing interventional cardiac catheterization often requires a general anesthetic and tracheal intubation to provide a stable airway and allow neuromuscular blockade to immobilize the patient during critical phases of the intervention. Diagnostic information is often available before these interventions, and the same principles noted above apply, i.e., providing baseline conditions for hemodynamics and ventilation during the procedure. Before an intervention, e.g., dilation of AS, the anesthetist normally increases the FiO₂ to 1.0 and prepares to resuscitate these high-risk patients if necessary. Drugs needed for resuscitation and a cardiac defibrillator must be immediately available. Thorough pre-procedure discussion and planning with the cardiologist for her/his requirements for the procedure and for untoward events is important. Blood transfusion is frequently needed, and this blood should be immediately available for high-risk procedures. Although the trachea of most patients can be extubated at the end of the procedure, and the patient can be returned to a cardiac ward after recovery, some patients will require ICU care, which must be planned for in advance.
Anesthesia Care of Pediatric Patients (George A. Gregory & Dean B. Andropoulos)

Anesthesia for Non-Cardiac Surgery

Because CHD is so frequent and because survival of CHD patients after surgical or catheter interventions is improving all over the world, patients with CHD will increasingly present for non-cardiac surgery. The types of surgery are generally not different from those required by patients without CHD, circumcision, hernia, myringotomy and tubes, tonsillectomy and adenoidectomy, appendectomy, and incision and drainage of abscesses. The approach to pre-anesthetic evaluation and planning is similar to that for cardiac surgery. A history of previous cardiac surgery and of residual lesions and symptoms is crucially important. The anesthetist must review the latest imaging study, usually echocardiography, the CXR, the last ECG, and the resting SpO2 as part of their search for evidence of residual cardiac lesions. A number of studies over the past decade have provided a much more accurate picture of which CHD patients are at highest risk of cardiac arrest and death when undergoing anesthesia for non-cardiac surgery. The highest risk group appears to be those with pulmonary hypertension, particularly when the PA pressure is at or above systemic levels. Patients with significant LVOT obstruction are also at very high risk and have the greatest risk of death following cardiac arrest. Infants with a single functional ventricle are another group at significant risk for cardiovascular collapse under anesthesia. Finally, patients with cardiomyopathy and decreased myocardial function are at very high risk for less than desired outcomes. Even patients with CHD without PA hypertension are at higher risk for cardiac arrest under anesthesia than patients without cardiac disease. Included in this group are infants with unrepaired large VSDs. Another risk factor for cardiac arrest during non-cardiac surgery is the patient with unrepaired or palliated CHD. These high-risk groups are summarized in Table 12-5.

Table 12-5: Highest Risk Patients for Cardiac Arrest with Anesthesia for Non-Cardiac Surgery

<table>
<thead>
<tr>
<th>High Risk Group</th>
<th>Example</th>
<th>Reasons</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left sided obstruction lesions</td>
<td>Aortic stenosis</td>
<td>Anesthetics lower blood pressure resulting in ↑ stenosis and ↓ coronary perfusion</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>Large untreated VSD in older child</td>
<td>Cathecholamine release from light anesthesia, ↑PaCO₂ and ↓PaO₂ from ventilation problems will PVR and right to left shunt</td>
</tr>
<tr>
<td>Ventricular dysfunction</td>
<td>Dilated cardiomyopathy</td>
<td>Anesthetics lower blood pressure and preload, and decrease myocardial contractility leading to ↓ stroke volume</td>
</tr>
<tr>
<td>Single ventricle infants</td>
<td>Hypoplastic left heart syndrome</td>
<td>↑FiO₂ and ↓PaCO₂ with intubation will ↑Qp:Qs; anesthetics decrease myocardial contractility</td>
</tr>
</tbody>
</table>

VSD = ventricular septal defect; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary-to-systemic blood flow ratio
Infective endocarditis (IE) prophylaxis is an important consideration for patients with CHD who are undergoing non-cardiac surgery. In 2007 the American Heart Association revised its official guidelines, which are very similar to other authorities’ guidelines around the world. To require IE prophylaxis, a patient must have both a cardiac condition and a surgical procedure indication that increases risk for IE. For example, a patient with a repaired VSD >6 months ago who has no residual defect does not need IE prophylaxis for any procedure. A cyanotic patient with an unrepaired TOF would require IE prophylaxis. A surgical procedure with no or very low risk of seeding the blood with bacteria does not require IE prophylaxis, even if the patient has a cardiac condition that might warrant it. For example, simple cystoscopy or endoscopy does not require IE prophylaxis. Dental procedures require IE prophylaxis. If IE prophylaxis is indicated, choice of antibiotics depends on the procedure. A single dose of IV antibiotics 30-60 minutes before the procedure is sufficient. Table 12-6 A, B, and C displays the current recommendations for IE prophylaxis. Antibiotic drugs are recommended for dental and respiratory tract procedures. For other cases, such as infected skin and musculoskeletal tissues, the antibiotic already being given for the infection is sufficient (nafcillin or methicillin, for example, for methicillin-sensitive Staphylococcus aureus infections). For gastrointestinal or genitourinary surgery, an antibiotic is used for prophylaxis against likely bacterial species that would be released into the blood and infect the heart when mucosa are incised. For example, ampicillin, with or without gentamycin, is effective for many urinary tract and gastrointestinal procedures. Clindamycin is often used for gastrointestinal procedures. Discuss the choice of antibiotics with the surgeon. In general, use the same antibiotic that would be used as surgical prophylaxis for patients without heart disease.

**Table 13-6A: Cardiac Conditions Associated with Highest Risk for Infective Endocarditis**

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prosthetic cardiac valve or material</td>
</tr>
<tr>
<td>Previous infective endocarditis</td>
</tr>
<tr>
<td>Congenital Heart Disease*</td>
</tr>
<tr>
<td>Unrepaired CHD, including palliative shunts</td>
</tr>
<tr>
<td>Repaired CHD with prosthetic material; first 6 months after procedure</td>
</tr>
<tr>
<td>Repaired CHD with residual defects</td>
</tr>
<tr>
<td>Cardiac transplant recipients with valvulopathy</td>
</tr>
</tbody>
</table>

CHD, congenital heart disease. * Except for those conditions listed, antibiotic prophylaxis is no longer recommended for any other form of CHD. Modified and reproduced with permission from Circulation. 2007;116:1736-54.
Table 13-6B: Surgical Procedure Indications for Infective Endocarditis Prophylaxis*

<table>
<thead>
<tr>
<th>Indications</th>
<th>Antibiotic Prophylaxis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dental procedures involving manipulation of gingival mucosa or periapical region of teeth</td>
<td>Bacampicillin 50 mg/kg IV</td>
</tr>
<tr>
<td>Respiratory tract procedures: tonsillectomy/adenoidectomy, bronchoscopy, thoracic surgery</td>
<td>Cefuroxime 50 mg/kg IV</td>
</tr>
<tr>
<td>Infected skin, skin structures, or musculoskeletal tissues</td>
<td>Cefazolin 50 mg/kg IV</td>
</tr>
<tr>
<td>Gastrointestinal or genitourinary procedures where mucosa are incised or biopsied (surgery or endoscopy); not needed for simple endoscopy or cystoscopy without biopsy</td>
<td>Clindamycin 20 mg/kg IV</td>
</tr>
</tbody>
</table>

* Except for those conditions listed, in Table 13-6A, antibiotic prophylaxis is no longer recommended for any other form of CHD. Modified and reproduced with permission from Circulation. 2007;116:1736-54.

Table 13-6C Antibiotic Regimens for Endocarditis Prophylaxis for Dental or Respiratory Tract Procedures

<table>
<thead>
<tr>
<th>Setting</th>
<th>Antibiotic</th>
<th>Dose: Single Dose 30-60 Minutes Before Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>No penicillin allergy</td>
<td>Ampicillin</td>
<td>50 mg/kg IV</td>
</tr>
<tr>
<td>Mild penicillin allergy</td>
<td>Cefazolin or Ceftriaxone*</td>
<td>50 mg/kg IV</td>
</tr>
<tr>
<td>Severe penicillin allergy</td>
<td>Clindamycin</td>
<td>20 mg/kg IV</td>
</tr>
</tbody>
</table>

*Do not use cephalosporins with a patient history of anaphylaxis, angioedema, or urticaria with penicillin or ampicillin. Give test dose of cephalosporins with history of mild penicillin allergy: 5 mg/kg IV, wait 5 minutes to assess for any reaction, then give remainder of dose slowly. IV = intravenous. Modified and reproduced with permission from Circulation. 2007;116:1736-54.

Conclusions

This chapter has presented an overview of anesthesia for CHD, focusing on two-ventricle lesions that undergo complete corrective surgery. Because these lesions comprise >50% of patients with CHD, patients with these lesions are increasingly undergoing surgery wide and surviving. Following surgical correction, most of these patients can lead a normal life and have normal life expectancy. Patients with a single ventricle and those with other very complex lesions comprise a smaller proportion of patients with CHD, require a large commitment of resources, and their outcomes in terms of mortality, morbidity, quality of life, and duration of life in general are not as good. The reader is referred to the Bibliography for a more extensive discussion of both the simpler and the more complex CHD patients. Anesthesia for CHD is a complicated field that is constantly changing, but because CHD surgery is increasingly practiced in many settings all over the world, it is hoped that this introduction will be useful to anesthetists who are participating in the care of these patients.
Chapter 12: ANESTHESIA FOR PATIENTS WITH CONGENITAL HEART DISEASE

Bibliography

1. Permission has been obtained from the publishers for the use of all Figures and Tables where necessary.