Chapter

26 CONGENITAL HEART DISEASE

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Categorization of congenital heart disease (CHD) may be based on distinctive anatomic or physiologic features of the defects (Box 26.1). Sometimes complete comprehension of the anatomic complexities in a patient with CHD may be difficult because of a wide range of anatomic lesions. Fortunately, many lesions share similar pathophysiologic conditions despite their anatomic variations. Understanding these physiologic conditions will lead to successful management of a patient with complex CHD.

FUNDAMENTAL PATHOPHYSIOLOGY IN CONGENITAL HEART DISEASE

Normally, pulmonary blood flow (Qp) and systemic blood flow (Qs) do not mix, and the entire cardiac output flows sequentially from one circulation to the other. All of the systemic venous return is directed to the pulmonary circulation and, likewise, all of the pulmonary venous return is directed to the systemic arterial circulation. Shunting occurs when a portion of the venous return of one circulation is redirected back to the arterial outflow of the same circulation.[1] This redirected flow occurs when there is an abnormal communication or a defect between two otherwise separate structures. The relative downstream blood pressures of the communicating structures dictate the direction of the shunt flow, whereas the size of the defect determines the amount of shunting. Small defects tend to be *restrictive* with limited flow, and large defects tend to be *nonrestrictive* with unimpaired flow.[1]

Left-to-Right Shunts

A left-to-right (L → R) shunt occurs when part of the pulmonary venous return is redirected toward the pulmonary

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arterial system.[1] This defect may occur at numerous locations, including the pulmonary veins (anomalous pulmonary venous return), the atrial septum (atrial septal defect), the ventricular septum (ventricular septal defect [VSD]), and at the great vessels (patent ductus arteriosus [PDA]).

The portion of the pulmonary blood flow (Qp) that is redirected toward the pulmonary artery is *recirculated* pulmonary blood flow. The portion of the pulmonary blood flow that is appropriately directed toward the systemic circulation (Qs) is *effective* pulmonary blood flow. Their sum is the total pulmonary blood flow (Qp) (Fig. 26.1).

Typically, CHD lesions with left-to-right (L → R) shunts remain acyanotic lesions. However, pulmonary overflow can result in pulmonary edema and hypotension. Prolonged hypotension can lead to circulatory shock with multiple organ system failure and lactic acidosis; the long-term effects of pulmonary overflow may be an increase of pulmonary vascular resistance (PVR) and abnormal cardiac chamber dilation. Over time an unrepaired large left-to-right (L → R) shunt can reverse its direction and become a cyanotic lesion. This is known as Eisenmenger physiology.

Box 26.1 Categorization of Congenital Heart Disease

Acyanotic versus cyanotic—VSD versus TOF
Simple versus complex—ASD versus HLHS
Left-to-right shunt versus right-to-left shunt versus mixing lesions—ASD versus TOF versus HLHS

ASD, Atrial septal defect; *HLHS*, hypoplastic left heart syndrome; *TOF*, tetralogy of Fallot; *VSD*, ventricular septal defect.

Right-to-Left Shunts

A right-to-left (R → L) shunt occurs when a portion of the systemic venous return is redirected to the systemic arterial outflow without first circulating through the lungs. The hallmark of lesions producing a right-to-left shunt is arterial oxygen desaturation. CHD lesions with right-to-left (R → L) shunts are cyanotic lesions. The physiologic effect of a right-to-left (R → L) shunt is arterial oxygen desaturation, because the recirculated oxygen-poor systemic venous blood mixes with the oxygen-rich pulmonary venous blood. The degree of desaturation depends

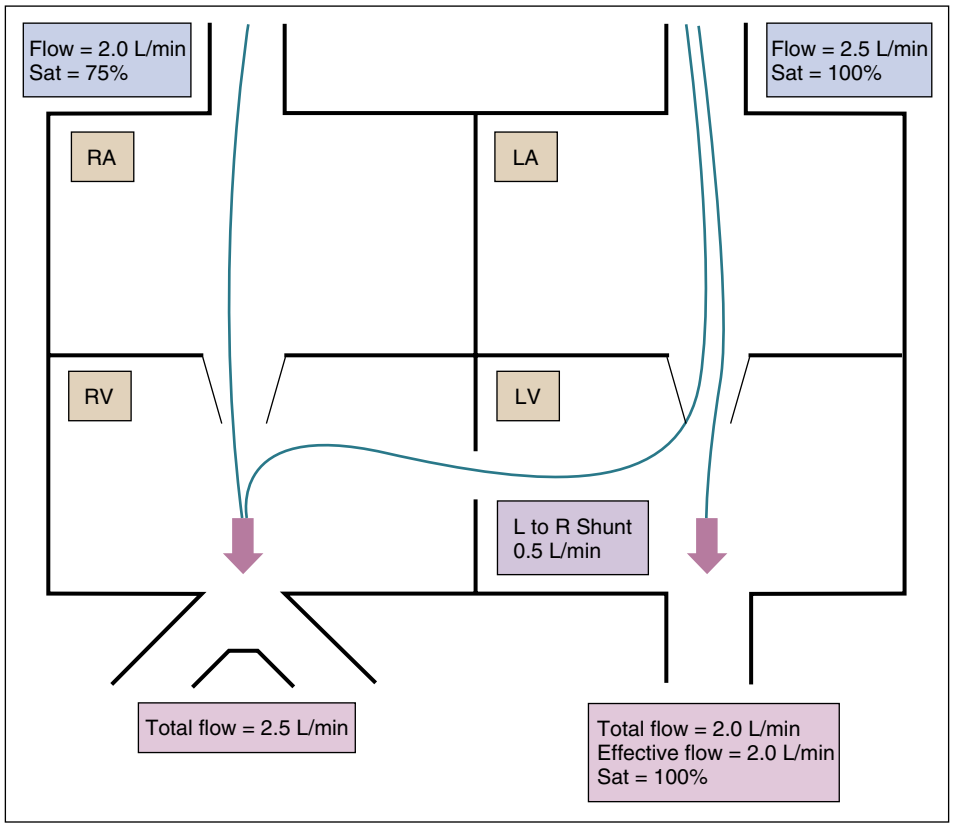

Fig. 26.1 Schematic diagram of a ventricular septal defect. A left-to-right shunt occurs at a septal defect with 0.5 L/min flow. Thus, total pulmonary flow is 2.5 L/min, of which the 2 L/min is the effective pulmonary flow; 2 L/min is also the systemic flow (Qs < Qp). *LA*, Left atrium; *LV*, left ventricle; *Qp*, pulmonary flow; *Qs*, systemic flow; *RA*, right atrium; *RV*, right ventricle.

upon the magnitude of right-to-left shunt as well as the degree of desaturation of the systemic venous return.[2]

Mixing Lesions

Whereas a shunt connotes a communication between pulmonary and systemic venous circulations with *partial* mixing, many forms of CHD result in a *complete* blending of the two. Mixing lesions therefore are conditions in which oxygen content is equilibrated between the two circulations, yielding identical or nearly identical oxygen saturation at both the pulmonary and systemic arterial level.[2]

As with right-to-left shunts, one of the chief characteristics of mixing lesions is systemic arterial oxygen desaturation. The degree of desaturation depends on the flow volume of the two contributing circulations as well as the difference in the individual oxygen saturation. A decrease in pulmonary venous saturation from apnea or atelectasis will decrease the saturation of the final mixed circulation. A decrease in the systemic venous saturation will also cause the final systemic arterial saturation to decrease. Factors that cause a decrease in systemic venous oxygen saturation include fever (increase of systemic oxygen consumption), low cardiac output states (which cause increased oxygen extraction in the microvasculature), and anemia (decrease in systemic oxygen delivery).

Significance of Qp:Qs Ratio in Mixing Lesions

In CHD with mixing physiology, a Qp:Qs ratio close to 1 will maximize the *effective* component of each circulation and minimize the wasteful *recirculated* component. For Qp:Qs ratio more than 1, preferential flow toward the pulmonary artery increases pulmonary blood flow, resulting in increased oxygen saturation of the mixed blood, but decreases systemic cardiac output and yields less oxygen delivery. For a Qp:Qs ratio less than 1, preferential flow toward the aorta increases systemic blood flow leading to higher systemic perfusion pressure, but the increased output contains blood with lower oxygen saturation and also leads to a decrease in oxygen delivery.

Relative resistance to flow of the two circulations, PVR and systemic vascular resistance (SVR), determines the Qp:Qs ratio. If PVR exceeds SVR, Qs will exceed Qp. Likewise, if SVR exceeds PVR, Qp will exceed Qs. SVR and PVR are both affected by many factors, which are listed in Box 26.2.[3]

Significance of the Patent Ductus Arteriosus

The ductus arteriosus connects the pulmonary artery to the descending aorta and is functionally closed within 4 days after birth in a healthy neonate as a result of an increase in arterial oxygen tension and decrease in placental prostaglandins (also see Chapter 34). However, mixing lesions with one functional ventricle often require a PDA to supply blood flow to the underdeveloped side. Shunting through the PDA in systole is either left to right (e.g., pulmonary atresia with intact ventricular septum) or right to left (e.g., hypoplastic left heart syndrome [HLHS]), depending on which side of the heart is hypoplastic. In other words, systemic blood flow is ductal-dependent in certain lesions such as HLHS. For other lesions such as pulmonary atresia the ductus arteriosus is required for pulmonary blood flow.

Ductal shunting during diastole, however, is usually left to right through the PDA because the aorta has a higher resting tone than the pulmonary artery. This means that a large amount of blood can be diverted to the lungs and away from the coronary arteries during diastole. Consequently the myocardium may become ischemic and infarcted because of coronary ischemia. Maneuvers that decrease PVR will cause more pulmonary runoff and exacerbate coronary ischemia.

Box 26.2 Impact of Anesthetic Management on Peripheral and Systemic Vascular Resistance

Events That Increase Systemic Vascular Resistance
- Light anesthesia
- Sympathetic nervous system activation
- Administration of α-agonists
- Physical manipulations (e.g., compression of the femoral arteries by flexing the hips of infants and small children)

Events That Decrease Systemic Vascular Resistance
- Deep anesthesia
- Administration of vasodilating drugs—nitrates, intravenous and inhaled anesthetics

Events That Increase Pulmonary Vascular Resistance
- Alveolar hypoxemia (e.g., from low inspired oxygen concentrations)
- Hypercapnia
- Acidosis
- High lung volumes or pressures—tend to collapse pulmonary capillaries
- Low lung volumes with atelectasis—tend to collapse larger pulmonary blood vessels
- Light anesthesia
- Sympathetic nervous system stimulation
- Hypothermia

Events That Decrease Pulmonary Vascular Resistance
- Hyperventilation
- Hypocarbia
- Alkalosis
- Oxygenation
- Inhaled nitric oxide
- Warmth
- Bronchodilators (e.g., albuterol)

Eisenmenger Syndrome

CHD may subject the lungs to abnormal blood flow or pulmonary artery pressure (Box 26.3). Over time, the pulmonary vasculature may undergo a process of remodeling with a gradual increase in PVR that results in pulmonary hypertension, even if the underlying hemodynamic

Box 26.3 Defects Resulting in Increased Pulmonary Blood Flow and Pulmonary Artery Pressure Over Time

Increased Pulmonary Blood Flow
Atrial septal defect
Anomalous pulmonary venous return

Increased Pulmonary Artery Pressure
Ventricular septal defect
Atrioventricular canal defect
Aortopulmonary window
Truncus arteriosus
Transposition of the great arteries
Double-inlet left ventricle
Patent ductus arteriosus

problem is corrected.[3] When pulmonary hypertension becomes irreversible and pulmonary pressure becomes supersystemic, blood is preferentially directed toward the systemic circulation and the direction of the shunt is right to left (R → L) even if the original shunting pattern was left to right (L → R). This condition is called Eisenmenger syndrome and often is a contraindication for surgical correction of the shunt.

Obstructive Lesions

Obstructive lesions consist mainly of left ventricular outflow track (LVOT) obstructions and coarctation of the aorta.

Left Ventricular Outflow Track Obstruction

LVOT obstructive lesions account for about 3% to 6% of CHD and can occur at subvalvar, valvar, and supravalvar levels.[4] Valvar aortic stenosis is the most common form of LVOT obstruction in children.[4,5] Bicuspid aortic valve is the most prevalent abnormality in this disease and often leads to clinical manifestations in early adulthood. Subvalvar aortic stenosis encompasses a variety of lesions, which include a thin membrane, thick fibromuscular ridge, diffuse tunnel-like obstruction, and abnormal mitral valve attachments.[6] Supravalvar aortic stenosis (Fig. 26.2) often has an hourglass deformity consisting of a discrete constriction of a thickened ascending aorta at the superior aspect of the sinuses of Valsalva and is often a feature in patients with Williams syndrome.[7]

In utero, critical aortic stenosis can lead to HLHS. Infants with severe aortic stenosis present with heart failure and failure to thrive. Older children with aortic stenosis are rarely symptomatic but will develop left ventricular hypertrophy, premature coronary atherosclerosis, and congestive heart failure over time. Various interventional and surgical approaches are available for aortic stenosis including balloon valvuloplasty, the Ross-Konno procedure, resection of the obstruction, and valve replacement. Timing and type of the surgery depend on the individual case.

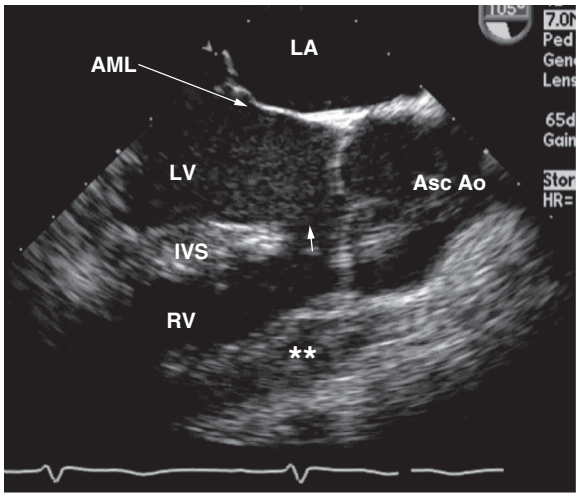

Fig. 26.2 Transesophageal echocardiogram of an adult with unrepaired tetralogy of Fallot. Note that the aorta is straddling over both ventricles, and a ventricular defect (*short arrow*) is seen immediately below the aortic valve. Right ventricle is hypertrophied (**). *AML*, Anterior mitral leaflet; *Asc Ao*, ascending aorta; *IVS*, interventricular septum; *LA*, left atrium; *LV*, left ventricle; *RV*, right ventricle.

Coarctation of the Aorta

Coarctation of the aorta is a discrete narrowing of the thoracic aorta just distal to the left subclavian artery (Fig. 26.3). It can be an isolated lesion or may be found in conjunction with other lesions such as aortic stenosis and VSDs. Infants with critical coarctation are at risk of developing heart failure and death when the ductus arteriosus closes. Those infants whose circulation is ductal dependent need to continue intravenous prostaglandin E_1 infusion to maintain the patency of the ductus arteriosus until surgery. Long-term follow-up of patients with coarctation of the aorta is imperative as there are long-term sequelae in adulthood, including recoarctation, hypertension, aortic aneurysm, coronary artery disease, and stroke.[8] In parturients, preeclampsia can occur in pregnancy, even after successful surgical correction[8] (also see Chapter 33).

PERIOPERATIVE MANAGEMENT

Surgery for CHD is planned with the cooperative input of a multidisciplinary team that includes surgeons, cardiologists, critical care specialists, and anesthesiologists. Patients require optimal medical care prior to surgery. For the anesthesia provider, understanding the physiology of the cardiac lesion and the subsequent effects of the planned surgical palliative or corrective procedure will lead to successful management of the patient (also see Chapter 13).

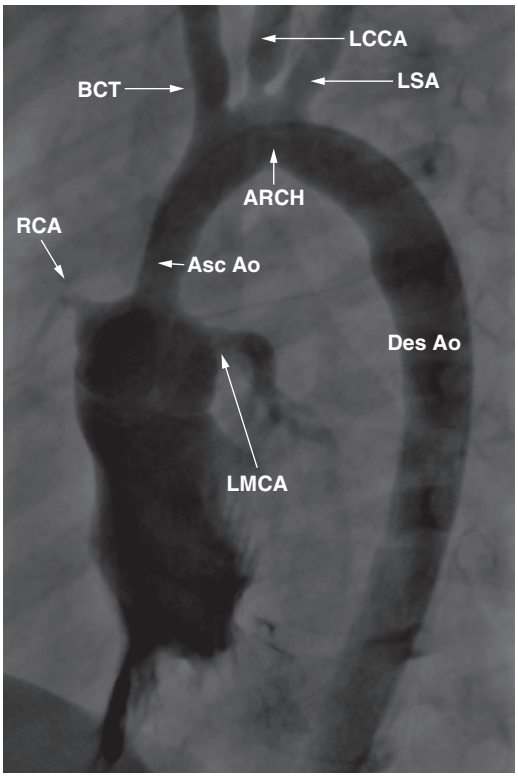

Fig. 26.3 Angiogram of the aorta during systole showing narrowing of the ascending aorta. This is an example of supravalvular aortic stenosis. The left main coronary artery is dilated and the orifice of the left common carotid artery is stenosed. *Asc Ao,* Ascending aorta; *BCT,* brachiocephalic trunk; *Des Ao,* descending aorta; *LCCA,* left common carotid artery; *LMCA,* left main coronary artery; *LSA,* left subclavian artery; *RCA,* right coronary artery.

History and Physical Examination

A review of the patient's history includes attention to details that are of importance to pediatric anesthetic care in general, such as pertinent pregnancy details, prematurity, and postnatal course (also see Chapter 34). Patients with CHD frequently have associated syndromes (trisomy 21, DiGeorge syndrome) or evidence of chronic illness (renal dysfunction, pulmonary edema, imbalances in electrolyte and glucose metabolism). Complete review of the patient's preoperative medications and laboratory studies (i.e., complete blood count, electrolytes, coagulation studies, indices of renal and hepatic function) is mandatory.

The anesthesia provider should review the available diagnostic studies such as the echocardiograms (Fig. 26.4) and cardiac catheterization studies (see Fig. 26.2). Magnetic resonance imaging (MRI) will also provide invaluable anatomic details (see Fig. 26.4). Electrocardiograms and chest radiographs are part of the routine preoperative evaluation. Medical and surgical interventions that

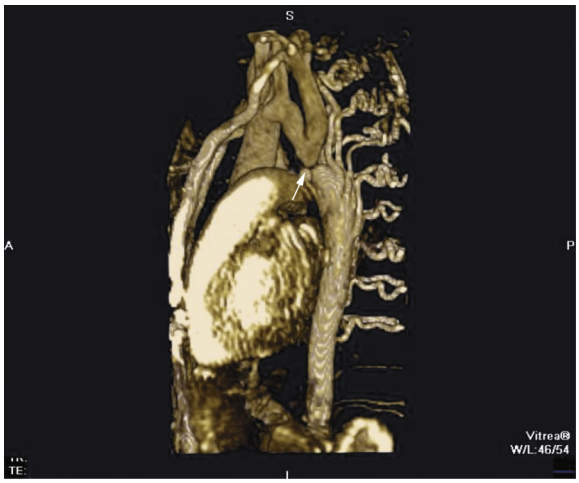

Fig. 26.4 Three-dimensional magnetic resonance image of a coarctation of aorta (*arrow*). Note the numerous collateral arteries to the descending aorta.

have been instituted previously and any interim change or deterioration in the patient's status are evaluated. Previous sternotomy is a risk factor for increased operative blood loss and cardiac trauma during dissection as a result of adhesions to the sternum and chest wall. Many hospitalized neonates require continuous infusions of inotropic drugs or other medications such as prostaglandin E_1 to maintain ductal patency and hemodynamic stability while awaiting surgery.

Physical examination includes an assessment of airway problems (such as might occur in patients with genetic syndromes, e.g., trisomy 21), signs of congestive heart failure (tachypnea, wheezing, dilated neck veins), cyanosis, nutritional status, and any other coexisting conditions. Outpatients who have been scheduled for elective surgery are evaluated the day of surgery for new onset of signs or symptoms of an intercurrent illness such as an upper respiratory tract infection. Inpatients are evaluated for their hospital course along with any developing problems such as an increased white blood cell count, which could indicate the presence of an infectious or inflammatory process.

Preparation for Surgery

All patients must follow the standard American Society of Anesthesiologists (ASA) guidelines for fasting. Outpatients taking cardiac medications are generally advised to continue therapy up to and including the day of surgery, although preference may vary among anesthesia providers regarding diuretics and angiotensin-converting enzyme inhibitors as well as angiotensin receptor blocking (ARB) drugs. Anticoagulants and antiplatelet medications are usually not given several days before surgery.

Operating Room Setup

Preparation of the operating room should include readiness of age-appropriate airway equipment, intravenous equipment, and invasive monitors. All intravenous administration sets should be meticulously de-aired to prevent paradoxical arterial embolization of intravenous air bubbles. A warming blanket or surface cooling equipment is made available, and adjustment of the operating room temperature precedes entry of the patient (27° C for small or premature infants and 24° C for older children). Hemodynamic medications are prepared in weight-appropriate dilutions before surgery. Preparation also includes readiness of specialized equipment such as transesophageal echocardiography (TEE) or nitric oxide (iNO) delivery systems.

Induction of Anesthesia

Induction of anesthesia for patients with CHD may involve either the use of inhaled or intravenous anesthetics (Boxes 26.4 and 26.5). The goal is to execute a smooth anesthetic induction to avoid increased anxiety, crying, coughing, or breath-holding. These events may aggravate unfavorable physiologic effects such as increased right-to-left shunting and dynamic right- or left-sided ventricular outflow tract obstruction in susceptible patients.

The choice of anesthetic and route of delivery are less important than the practitioner's understanding of the hemodynamic effects of a particular drug on the underlying patient physiology. The ability to recognize untoward responses and intervene to correct the problem are hallmarks of the pediatric cardiac anesthesiologist.

Inhaled Induction of Anesthesia

Awake infants and children without intravenous access are frequently amenable to an inhaled induction of anesthesia. This strategy is typically reserved for those with minimal or well-controlled congestive heart failure because a dose-dependent decrease in myocardial contractility occurs with volatile anesthetics. Sevoflurane is probably the preferred volatile anesthetic because of its lack of pungency and airway irritant effect and the absence of cardiac sensitization to catecholamines.[9] Nitrous oxide may hasten the induction of anesthesia and decrease the necessary concentration of sevoflurane. Concerns regarding the propensity of nitrous oxide to increase PVR have not been substantiated in patients with CHD, but the decrease in fraction of inspired oxygen (F_{IO_2}) may impair protection against increased PVR. Because of nitrous oxide's property to expand intravascular air bubbles, its administration is often discontinued shortly after induction of anesthesia.

Placement of a pulse oximetry probe is minimally distressing to an anxious, awake child and provides ample monitoring for the initial stages of an inhaled induction of anesthesia. Once an adequate stage of anesthesia has been achieved, other noninvasive monitors are placed in a timely fashion. After intravenous access is secured, additional intravenous anesthetics, neuromuscular blocking drugs, and possibly anticholinergics may be given before laryngoscopy and tracheal intubation.

Box 26.4 Anesthetics Used for Induction of Anesthesia in Congenital Heart Disease

- Sevoflurane 1.5%-3.5% (ET)
  - Decrease SVR, contractility
  - Decrease dose if N_2O used; may cause myocardial depression
- Fentanyl 20-50 μg/kg
  - Nonsignificant effect on contractility, SVR
  - May cause loss of sympathetic tone, bradycardia
  - May cause chest wall rigidity with rapid administration or large doses
- Sufentanil 5-10 μg/kg
  - Similar to fentanyl
- Ketamine (IV) 1-2 mg/kg
  - Increases HR, increase or no change in SVR, PVR
  - Actually a myocardial depressant with sympathetic stimulant properties; may decrease contractility in patients with depleted SNS
  - May cause bronchorrhea (prevented with atropine, 20 μg/kg)
- Ketamine (IM) 3-5 mg/kg
- Etomidate (IV) 0.2-0.3 mg/kg
  - Preserves HR, SVR, PVR, contractility
  - May inhibit endogenous corticosteroid production; burning pain at the injection site

BP, Blood pressure; *CHF*, congestive heart failure; *CO*, cardiac output; *ET*, end-tidal; *HR*, heart rate; *IM*, intramuscular; *IV*, intravenous; *PVR*, pulmonary vascular resistance; *SNS*, sympathetic nervous system; *SVR*, systemic vascular resistance.

Intravenous Induction of Anesthesia

Patients with poorly controlled congestive heart failure, moderately impaired ventricular function, significant right-to-left shunting, or complete mixing lesions may benefit from the increased stability afforded by intravenous induction of anesthesia. Frequently, these patients come to the operating room from a critical care setting with intravenous access already in place. Traditionally, intravenous opioids have been used in this setting because they produce little or no myocardial depression and also lack vasodilating properties in both the pulmonary and systemic vascular beds.[9] Other intravenously administered anesthetics that are used in patients with CHD include benzodiazepines, etomidate, and ketamine. Propofol can cause hypotension or increased right-to-left shunting in some patients with right ventricular outflow tract obstruction. In other patients with adequate ventricular function, propofol is tolerated when incrementally administered. Ketamine preserves or augments sympathetic nervous system tone and, in so doing, maintains

Box 26.5 Common Congenital Cardiac Lesions: Summary of Anesthetic Goals and Induction Strategies

General Goals and Principles

- Avoid air entrapment in intravenous and pressure tubing; use meticulous clearing techniques.
- Avoid dehydration; give careful orders regarding NPO status (following ASA guidelines).
- Avoid myocardial depression.
- Maintain sinus rhythm whenever possible.
- Well-sedated, cooperative patient is ideal.
- Premedication is indicated for patients older than 1 year (oral midazolam, 0.5-1 mg/kg).
- Close monitoring after sedation.

Lesions Characterized by Excessive Pulmonary Blood Flow

Atrial Septal Defects

- Avoid further decreases in pulmonary vascular resistance (hyperventilation, high F_{IO_2}).
- Consider early tracheal extubation.

Ventricular Septal Defects

- Avoid decreases in pulmonary vascular resistance.
- Avoid excessive myocardial depression, particularly in patients with congestive heart failure—inhaled induction may be rapid.

Atrioventricular Septal Defects

- Avoid decreases in pulmonary vascular resistance before cardiopulmonary bypass.
- Prepare to treat pulmonary hypertension (100% oxygen, hyperventilation, alkalinization, deep sedation).
- Have nitric oxide available and ready.
- Inotropic support frequently is required.

Truncus Arteriosus

- Neonates are critically ill and require close management of systemic and pulmonary vascular resistance to balance systemic and pulmonary blood flow.
- Addition of carbon dioxide or nitrogen may be needed to decrease F_{IO_2} to 17%.

Hypoplastic Left Heart Syndrome

Surgical correction occurs in three stages:

Stage I: Norwood procedure

- Perform ascending aorta and arch reconstruction.
- Perform PDA ligation.
- Construct a reliable pulmonary blood flow source using a Blalock-Taussig shunt or Sano shunt.
- Anesthetic management includes prebypass PGE_1 infusion, maintenance of nearly equal pulmonary and systemic blood flow for adequate systemic perfusion, precautions against air embolism, maintenance of anesthesia with intravenous drugs, and postbypass maintenance of a high hematocrit and probably a need for inotropic support.

Stage II: Glenn procedure

- Create a direct connection between the superior vena cava and pulmonary artery.
- Anesthetic management includes maintenance of a high hematocrit, elevation of the head of the bed to facilitate venous drainage, avoidance of central lines to reduce the risk for pulmonary artery thrombus, and recognition that positive-pressure ventilation of the patient's lungs may decrease pulmonary blood flow and cardiac output.
- Mild hypoventilation may increase oxygen saturation.

Stage III: Fontan procedure

- Reroute blood flow from the inferior vena cava into the pulmonary circulation, usually accomplished using an extracardiac conduit.
- Preload to the heart is completely passive. Management of patient status after Fontan procedure should focus on maintaining reasonable preload, i.e., passive flow from systemic veins to the pulmonary artery and eventually to the common atrium.
- Poor prognostic factors are high pulmonary vascular resistance, tricuspid regurgitation, and decreased ventricular function.

Lesions With Inadequate Pulmonary Blood Flow

Transposition of the Great Arteries (TGA)

- PGE_1 infusion is maintained before cardiopulmonary bypass.
- Patient may need Rashkind procedure (atrial septectomy) if patent ductus does not provide adequate mixing for survival.
- Manipulate pulmonary vascular resistance before cardiopulmonary bypass.
- Use an opioid-based anesthetic.

Tetralogy of Fallot (see Fig. 26.2)

- Adequate preoperative hydration is essential.
- Manipulations are indicated to decrease pulmonary vascular resistance and improve pulmonary blood flow.
- Hypercyanotic episodes are treated by intravenous fluid administration, sedation, and pharmacologically induced increases in systemic vascular resistance (phenylephrine).
- Avoid increases in heart rate, which may worsen infundibular pulmonary stenosis.
- Rate of induction of anesthesia with a volatile anesthetic may be slowed because of a right-to-left shunt.

Tricuspid Atresia or Pulmonary Atresia With Intact Ventricular Septum

- Usually the right ventricle is diminutive or hypoplastic.
- Surgical approach involves an aortopulmonary shunt and subsequent Glenn and Fontan procedures.

Total Anomalous Pulmonary Venous Return

- Severe cyanosis is treated with high F_{IO_2}.
- Avoid systemic acidosis and high hematocrit.

Obstructive Lesions

Coarctation of the Aorta (see Fig. 26.4)

- Use arterial monitoring in the right arm.
- Use cuffed endotracheal tube to provide adequate ventilation to patients requiring the thoracotomy position.
- Avoid acidosis.

Aortic Stenosis

- Avoid tachycardia, dysrhythmias, hypotension.
- Decrease myocardial oxygen demand.
- Maintain preload and afterload.

Subvalvular Aortic Stenosis

- Avoid tachycardia, dysrhythmias, hypotension.
- Decrease myocardial oxygen demand.
- Maintain preload and afterload.

Supravalvular Aortic Stenosis (see Fig. 26.3)

- Note that this is associated with Williams syndrome.
- Patient may have concomitant pulmonary artery stenosis.
- Avoid tachycardia, dysrhythmias, hypotension.
- Coronary abnormalities are common.
- Avoid acute afterload reduction.

ASA, American Society of Anesthesiologists; *F_{IO_2}*, inspired oxygen concentration; *NPO*, nil per os (nothing by mouth); *PDA*, patent ductus arteriosus; *PGE_1*, prostaglandin E_1.

a high degree of circulatory stability. Concern regarding ketamine's propensity to increase PVR has not been substantiated in patients with CHD.[10] Ketamine may be administered intramuscularly to achieve stable induction of anesthesia and allow subsequent vascular cannulation to proceed in an anesthetized patient. Obviously, during the administration of all intravenous drugs, avoidance of air is mandatory. The presence of circulatory mixing or shunting in patients with CHD poses a real risk for paradoxical air embolization should any bubbles reach the central circulation.

Airway Management (Also See Chapter 16)

The size of the endotracheal tube is individualized according to the age and size of the patient. Administration of a neuromuscular blocking drug will facilitate tracheal intubation (also see Chapter 11). The selection of the drug depends on the patient (age, type of lesion, renal function) and the characteristics of the drug (duration of action, hemodynamic properties, and mode of elimination). Vecuronium and rocuronium have an intermediate duration of action but rocuronium has a faster onset of action than vecuronium. Rocuronium also increases heart rate, which is useful in pediatric patients. Succinylcholine is rarely used in pediatric anesthesia.

The approach to ventilatory management hinges upon how the circulatory system will be affected by changes in PVR in relationship to the SVR. The ventilation strategy should have minimal impact on blood flow across shunts or on tenuously balanced pulmonary-to-systemic flow ratios. Understanding how changes in PVR will affect the physiology of the cardiac lesion will help govern such parameters as F_{IO_2}, minute ventilation, use of positive end-expiratory pressure, and peak inspiratory airway pressure.

Monitoring (Also See Chapter 20)

Invasive monitoring is usually established after induction of anesthesia. Patients undergoing cardiac surgery generally require arterial line placement, as well as some form of central venous access. The cardiac lesion dictates the site for arterial line placement. For example, in a patient who has a Blalock-Taussig shunt (diversion of the subclavian artery to the ipsilateral pulmonary artery), the arterial line is placed contralaterally. Similarly, patients with coarctation of the aorta may have unreliable pressure measurement in the left upper extremity either because of the location of the coarctation or because of aortic cross-clamp placement at or near the left subclavian artery during surgery. The internal jugular vein is a common choice for central pressure monitoring and infusion of medications intraoperatively. Recently, many centers performing neonatal surgical repairs have moved away from the internal jugular approach owing to the risk of central venous catheter thrombosis. For this reason, many surgeons prefer to directly insert a right atrial catheter intraoperatively before separation from bypass. The catheter is then tunneled through the thorax.

TEE has become an invaluable monitoring tool in the operating room to further delineate anatomy that may not be clearly demonstrated by preoperative transthoracic echocardiography (TTE), to rule out additional defects, and to assess the quality of the repair.

Blood Tranfusions (Also See Chapter 24)

Many operations for CHD will require blood product administration, and the likelihood increases with smaller infants, lower preoperative hematocrit levels, repeat sternotomy incisions, and long cardiopulmonary bypass (CPB) times. Judgment and experience dictate how much and what type of blood products are made available at the start of surgery. Generally, small infants are allocated blood that is as fresh as possible (less than 5 days of storage) because older blood may become significantly hyperkalemic and develop leftward shifting of the oxygen-hemoglobin dissociation curve. Blood is administered with the use of appropriate filters and warming devices because small infants are particularly susceptible to intraoperative hypothermia and to bradydysrhythmias from boluses of hypothermic blood products. Having packed red blood cells (PRBCs) available in the operating room before the skin incision is appropriate in cases of repeat sternotomy inasmuch as these patients are at risk for severe bleeding from unintentional injury to major cardiac structures. Often, other components of the blood, such as fresh frozen plasma (FFP), platelets, and sometimes cryoprecipitate (CRYO), are administered after weaning from CPB, especially for small infants and complex cases with long CPB time.

Antifibrinolytic Drugs

Antifibrinolytic drugs can reduce blood loss and transfusion requirements during surgery for CHD. Aminocaproic acid is the preferred drug in our institution, whereas tranexamic acid is used in others. Aprotinin is no longer available for use.[11,12]

Maintenance of Anesthesia

Typically anesthesia is maintained with a combination of intravenous opioids, benzodiazepines, and volatile anesthetics (doses < 1 minimum alveolar concentration [MAC]), and neuromuscular blocking drugs. Use of a small concentration of a volatile anesthetic minimizes the myocardial depressant effects of the drug while also decreasing the total dose of opioids that would otherwise be necessary to ensure adequate anesthetic depth. Large doses of opioids (fentanyl, 50 to 100 μg/kg intravenously) are often given over the course of an operation.[10,12] These opioids may be administered in divided doses according to judgment of anesthetic depth or in anticipation of noxious surgical stimuli. Alternatively, opioids may be delivered

as a continuous intravenous infusion. Patients who are critically ill or who have complex cardiac anomalies may benefit from high-dose opioid techniques so that the hypotensive and myocardial depressant effects of the volatile anesthetics are minimized. In contrast, limited opioid administration (fentanyl <20 μg/kg) in patients with good cardiac reserve undergoing procedures for simple defects (e.g., atrial septal defect, VSD, PDA, coarctation of the aorta) will facilitate early postoperative tracheal extubation.[10] Dexmedetomidine (Precedex) is sometimes used as an anesthetic adjunct. It is infused at rates between 0.2 and 2.0 μg/kg/h. Dexmedetomidine has a tendency to slow the heart rate in some patients and for this reason has not been widely adopted. Nitrous oxide is generally not used for maintenance of anesthesia because of its propensity to expand unintentional intravascular air emboli.

Monitoring Changes in Shunt Ratios

The potential for significant changes in the circulatory system after anesthetic induction warrants early and possibly repeated analysis of arterial blood gases to allow early correction or refinement of pulmonary ventilation variables, as well as acid-base disorders, before the development of important circulatory derangements. In patients with shunts or mixing lesions, pulse oximetry also provides a continuous monitor of changes in the balance between pulmonary and systemic blood flow or changes in shunt direction or magnitude.

Anticoagulation

Anticoagulation with unfractionated heparin (3 to 4 mg/kg) delivered intravenously is achieved before cannulation for CPB. The subsequent anticoagulation effect is assessed by measuring the activated clotting time (ACT). Target ACT values may vary with institutional preference, but 480 seconds is typical. Additional heparin is administered if target values are not initially obtained. Heparin concentration assays may also be used instead of or as a supplement to the ACT.

Cardiopulmonary Bypass

Most procedures for repair of congenital cardiac defects require use of CPB (also see Chapter 25). As with adults, CPB for infants and children entails diversion of systemic venous return to the CPB machine and return of oxygenated blood to the arterial system. Venous blood is drained passively (by gravity) through two venous cannulas, one for each vena cava. The cannulas converge through a Y-connector to a cardiotomy reservoir, which allows rapid administration of blood products, crystalloid and colloid solutions, medications, and blood suctioned from the field by the surgeon ("pump suction"). The cardiotomy reservoir also provides a temporary buffer in the event that venous return is temporarily interrupted. Blood is next conducted to a pump mechanism, which is generally a centrifugal pump. This adjustable pump permits delivery of a specified rate of blood flow to the patient. Generally, flow rates are adjusted to maintain an age-appropriate mean arterial pressure. Blood is then channeled through a membrane oxygenator, which equilibrates the blood with a supply of fresh gas; in this way, oxygen is added and carbon dioxide is removed. The perfusionist controls oxygenation and ventilation by adjusting the blend (F_{IO_2}) and flow rate (sweep) of the fresh gas. Modern oxygenator circuits also allow rapid adjustment of blood temperature by running cooled or warmed water through a coil in contact with the blood path. Blood is then conducted back to the patient through tubing connected to a cannula positioned in the ascending aorta. An arterial filter is generally used downstream from the oxygenator to prevent microembolization of debris to the arterial tree.[13]

Complete diversion of venous return to the CPB machine followed by aortic cross-clamping and immediate administration of a cardioplegia solution will yield a still and bloodless heart for the surgeon. Because the act of aortic cross-clamping renders the heart ischemic, the cardioplegia solution has a dual purpose of providing both mechanical quiescence and myocardial protection. As with adults, these effects are achieved through the use of a cold (4° C) hyperkalemic crystalloid solution. Hypothermia and electromechanical arrest each contribute to minimizing myocardial oxygen requirements and lengthening the tolerable period of myocardial ischemia.

Calculation of Physiologic Variables

The perfusionist and anesthesiologist take into account the patient's size to calculate the necessary flow rate to maintain metabolic function. Equally important is the patient's estimated blood volume because it determines the degree of hemodilution that results when the patient's blood mixes with the obligatory "priming volume" of fluid that occupies the CPB machine's tubing, oxygenator, and cardiotomy reservoir at the onset of CPB. Whereas adult patients frequently have acceptable degrees of anemia as a result of this hemodilution, infants and small children require smaller, shorter tubing and lower-volume cardiotomy reservoirs to minimize this effect. Most infants require some blood product to be mixed with the circuit prime to preserve adequate oxygen-carrying capacity while on CPB. The amount of blood product required is a function of the patient's starting hematocrit, estimated blood volume, circuit prime volume, and the lowest acceptable limit of anemia (institutional and physician preferences vary but are commonly in the range of a hematocrit of 20% to 30%).

Body Temperature During Cardiopulmonary Bypass

Institutional, surgeon, or anesthesiologist preference also provides the perfusionist with a target patient temperature to be achieved while on CPB. Mild (30° C to 35.5° C) to moderate (25° C to 30° C) systemic hypothermia reduces

metabolic oxygen requirements (7% per degree Celsius) and provides protective effects on both cerebral and myocardial tissue.[13] Hypothermia is usually achieved by active cooling of CPB blood with a heat exchange device incorporated in the membrane oxygenator. Active rewarming is initiated toward the end of CPB. Deleterious effects of post-CPB hypothermia may include myocardial ischemia, cardiac dysrhythmias, elevated PVR, coagulation failure, or renal dysfunction.

Deep Hypothermic Circulatory Arrest

Deep hypothermic circulatory arrest (DHCA) was used in situations in which adequate surgical repair is precluded by CPB cannula placement or by the requirement to repair the aorta at or near the arch.[14-16] Because of adverse outcomes associated with DHCA, many medical centers are using regional low-flow cerebral perfusion via the innominate artery (25 to 50 mL/kg/min) to permit surgical repair in a field unencumbered by CPB cannulas. Cardiac cannulation and active cooling by CPB are required to lower core temperature to approximately 18° C to 20° C. After surgical repair, CPB is reestablished, and the patient is rewarmed and reperfused.

Weaning From Cardiopulmonary Bypass

Successful weaning from CPB requires close communication between the anesthesiologist, cardiac surgeon, and perfusionist. The surgeon requests the perfusionist to start rewarming at an appropriate point during the surgical procedure. The anesthesiologist commences pulmonary ventilation at the surgeon's request after the endotracheal tube is suctioned. With the patient warm and ventilated, weaning from CPB is initiated.

Cardiac Rhythm

Ventricular fibrillation can occur after removal of the aortic cross-clamp and reperfusion of the coronary arteries, especially when the hypothermia has not been fully corrected. It may revert spontaneously to a sinus rhythm but often requires electrical defibrillation. Acid-base or electrolyte disorders (hyperkalemia) may contribute to disturbances in cardiac rhythm. Relative bradycardia or atrioventricular node conduction failure can be corrected by means of temporary cardiac pacing. Many patients with good cardiac reserve who have endured relatively short periods (<1.5 hours) of aortic cross-clamping and the attendant myocardial ischemia may be able to separate from CPB without inotropic assistance. Many others will require infusion of inotropic drugs to achieve adequate cardiac output and systemic blood pressure. In particular, those with preexisting myocardial dysfunction, congestive heart failure, or hemodynamic instability are likely to require pharmacologic assistance for successful separation from CPB (Table 26.1). Inotropic drugs commonly utilized include dopamine, milrinone, epinephrine, and calcium.

Table 26.1 Common Vasoactive Drugs

Drug	Dose Range	Comments
Dopamine	3-20 μg/kg/min	Lower maximum effect than with epinephrine and norepinephrine Tachycardia
Epinephrine	0.02-0.1 μg/kg/min	Drug of choice when maximum inotropic effect is required Strong effect at the medium- to high-dose range Tachycardia
Norepinephrine	0.02-0.1 μg/kg/min	Strong α, β effects, with activity at lower doses than with epinephrine
Milrinone	0.25-1 μg/kg/min	Can lower both PVR and SVR No tachycardia May need α-agonist to prevent hypotension Loading dose typically is 25-50 μg/kg
Dobutamine	1-20 μg/kg/min	Lower maximum effect than with epinephrine and norepinephrine May decrease SVR or BP because of peripheral β_2-vasodilation

BP, Blood pressure; *PVR*, pulmonary vascular resistance; *SVR*, systemic vascular resistance.

Ventilation and Pulmonary Vascular Resistance

The approach to PVR and ventilation of the lungs must be carefully considered before separation from CPB. Patients with simple defects that have been repaired are no longer at risk for shunting and unbalanced Qp:Qs ratios. For this reason, such patients are typically ventilated with 100% inspired oxygen (F_{IO_2} of 1.0) at the time of separation from CPB, with minute ventilation sufficient to avoid respiratory acidosis. Patients with long-standing excessive pulmonary blood flow may have underlying pulmonary hypertension and may be at risk for pulmonary hypertensive crisis at the time of separation from CPB. These patients may benefit from maneuvers that minimize PVR including the application of inhaled nitric oxide (see Box 26.2).

Presence of a Residual Mixing Lesion

Difficulty arises when a palliative procedure has left the patient with a mixing lesion. This situation is exemplified by surgical treatment of HLHS (Norwood procedure) that results in a single ventricle supplying blood flow to both the pulmonary and systemic circulation (see Box 26.5). In such circumstances, which circulation will be likely to receive

Box 26.6 Causes of Difficulty in Separation From Cardiopulmonary Bypass

- Inadequate pulmonary blood flow (associated with arterial hypoxemia)
- Inadequate systemic blood flow (associated with hypotension and metabolic acidosis)
- Valvular dysfunction
- Dynamic outflow obstruction (decreases in cardiac output related to hyperdynamic or hypovolemic states)
- Decreased systemic vascular resistance (associated with long cardiopulmonary bypass times)
- Cardiac rhythm disturbances
- Hypovolemia

most of the cardiac output must be anticipated and adjusted so that PVR and SVR tend to yield a balanced circulation. Pulse oximetry is an invaluable tool in this particular situation because a patient with a complete mixing lesion will tend to have systemic oxygen saturation near 80% when the systemic and pulmonary circulations are balanced. Systemic saturation greater than 85% to 90% indicates excessive pulmonary blood flow (possibly with resultant systemic hypoperfusion or hypotension), whereas saturation less than 75% indicates inadequate pulmonary blood flow. The best possible milieu in the setting of the underlying defect must be provided in order to promote satisfactory cardiac output, adequate oxygenation, and a balanced circulation.

Difficulty in Separation of the Patient From Cardiopulmonary Bypass

Difficulty in separation from CPB may reflect multiple physiologic derangements but most often is due to inadequate pulmonary blood flow or inadequate systemic blood flow (Box 26.6). After separation from CPB, systemic arterial blood pressure, systemic oxygenation, and acid-base status must be closely monitored. Data derived from a central venous or pulmonary artery catheter may be helpful in diagnosing hemodynamic problems. TEE is useful in evaluating the surgical repair of CHD and cardiac function in the period after separation from CPB. In the event that pharmacologic support of cardiac contractility, vascular tone, and management of ventilation fails to achieve circulatory stability, patients may require resumption of CPB support. In some cases, a period of "rest" on CPB allows resolution of cross-clamp–related ischemic ventricular dysfunction, whereas in other situations, revision of the surgical repair may be indicated. If the patient cannot be separated from CPB despite surgical revision and maximal inotropic support, then extracorporeal life support may be instituted and continued until adequate cardiac and pulmonary function is regained.

Reversal of Heparin-Induced Anticoagulation

After successful weaning from CPB, protamine reverses the anticoagulation effect of heparin and is administered by means of slow intravenous infusion (over at least a 10-minute period). Pediatric patients, although susceptible to some detrimental complications of protamine administration, including anaphylactic, anaphylactoid, hypotensive, or severe pulmonary hypertensive reactions, are often spared these untoward effects more commonly observed in adults.

Coagulopathy

Although return of the ACT to baseline indicates successful reversal of heparin, there can be residual clinical coagulopathy from coagulation factor or platelet deficiency. Hypothermia and hypocalcemia may contribute to in vivo coagulopathy but will not be reflected in the ACT or other laboratory tests of coagulation. Early measurement of the platelet count, prothrombin time, and partial thromboplastin time will facilitate appropriate blood product therapy in the event that hemostasis is not achieved with protamine. Thromboelastography (TEG) is a test of hemostasis performed on whole blood that examines platelet function and the coagulation pathway (also see Chapter 42). However, its use has not been widely accepted owing to the lack of evidence supporting TEG-guided transfusion therapy as yielding better outcomes than the current transfusion practices.[17] Often the degree of clinical coagulopathy necessitates empirical administration of platelets, FFP, or other factor preparations before the results of any laboratory study become available.

Blood Component and Intravascular Volume Therapy (Also See Chapter 24)

Blood component and intravascular volume therapy must be administered very carefully to infants because their total intravascular volume is small in comparison to adults. Unless critically hypovolemic, blood product or volume therapy should proceed in aliquots of approximately 5 mL/kg to prevent excessive intravascular volume and possible ventricular dysfunction. Citrated blood products may cause important degrees of hypocalcemia, and calcium replacement may thus be necessary. Dilutional anemia can occur when administering platelet or plasma preparations. Fluid-warming devices prevent the delivery of cold fluid boluses to cardiac conduction tissue, as well as the development of systemic hypothermia.

Recombinant Activated Factor VII

Recombinant activated factor VII (rFVIIa) is approved by the Food and Drug Administration (FDA) for use in the prevention and treatment of bleeding in patients with hemophilia A or B with inhibitors to factor VIII or IX, factor VII deficiency, or Glanzmann thrombasthenia (also see Chapter 22).[18] Its role in pediatric cardiac surgery is evolving. rFVIIa is appropriate as rescue therapy when conventional hemostatic measures have failed to stop the bleeding after separation from bypass despite conventional therapy. The dose is typically 90 to 120 μg/kg every 2 hours for excessive bleeding. rFVIIa must be used cautiously as there

is risk of thromboembolic complications, particularly in patients who have undergone the arterial switch operation for transposition of the great arteries.[19,20]

POSTOPERATIVE CARE

Children undergoing surgery for CHD are managed in an intensive care setting, where continuous invasive monitoring is possible along with one-to-one nursing care. Mechanical ventilation of the patient's lungs is continued for variable intervals, depending on the type of surgery performed and the overall status of the patient. Sedation is maintained throughout the period of ongoing tracheal intubation. Critical care management entails the continuation of hemodynamic drug infusions and possibly electrical pacing of cardiac rhythm. Early postoperative management frequently involves correction of various electrolyte, glucose, and hematologic parameters. Mediastinal bleeding is assessed frequently. An intense index of suspicion is always maintained for the possible requirement for revision of the surgical repair, and bedside echocardiography is frequently undertaken in the intensive care unit to clarify hemodynamic problems or abnormal convalescence.

QUESTIONS OF THE DAY

1. What factors affect the degree of hypoxemia in a patient with a right-to-left shunt?
2. What is the physiologic effect of patent ductus arteriosus (PDA)? What is the significance of the ductus arteriosus in a patient with one functional ventricle?
3. What is Eisenmenger syndrome? What congenital heart defects are associated with its development?
4. A patient with congestive heart disease (CHD) requires arterial line monitoring. What factors influence the choice of monitoring site?
5. In a patient receiving general anesthesia for repair of CHD, what events can increase or decrease pulmonary vascular resistance (PVR)?

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