and associated anomalies and the hemodynamic physiology of the defect(s). Hemodynamic and angiographic study may be occasionally required to assess the status of the pulmonary vascular bed and clarify details of the altered anatomy. Cross-sectional imaging modalities such as computed tomography (CT) or magnetic resonance imaging (MRI) are useful in delineating complex anatomy and assessing extracardiac structures.

TREATMENT VENTRICULAR SEPTAL DEFECT

Closure is not recommended for patients with normal pulmonary arterial pressures with small shunts (pulmonary-to-systemic flow ratios of <1.5:1). Operative correction or transcatheter closure is indicated when there is a moderate to large left-to-right shunt with a pulmonary-to-systemic flow ratio >1.5:1, in the absence of prohibitively high levels of pulmonary vascular resistance (pulmonary arterial resistance is less than two-thirds of systemic arterial resistance).

In patients with Eisenmenger's VSD, pulmonary arterial vasodilators and both single- or double-lung transplantation with intracardiac defect repair or heart/lung transplantation show promise for improvement in symptoms **(Chaps. 281 and 320e)**. Chronic hypoxemia in cyanotic CHD results in secondary erythrocytosis due to increased erythropoietin production **(Chap. 49)**. The term polycythemia is a misnomer; white cell counts are normal, and platelet counts are normal to decreased. Compensated erythrocytosis with iron-replete equilibrium hematocrits rarely results in symptoms of hyperviscosity at hematocrits <65% and occasionally not even with hematocrits ≥70%. For this reason, therapeutic phlebotomy is rarely required in compensated erythrocytosis. In contrast, patients with decompensated erythrocytosis fail to establish equilibrium with unstable, rising hematocrits and recurrent hyperviscosity symptoms. Therapeutic phlebotomy, a two-edged sword, allows temporary relief of symptoms but limits oxygen delivery, begets instability of the hematocrit, and compounds the problem by iron depletion. Iron-deficiency symptoms are usually indistinguishable from those of hyperviscosity; progressive symptoms after recurrent phlebotomy are usually due to iron depletion with hypochromic microcytosis. Iron depletion results in a larger number of smaller (microcytic) hypochromic red cells that are less capable of carrying oxygen and less deformable in the microcirculation; with more of them relative to plasma volume, viscosity is greater than for an equivalent hematocrit with fewer, larger, iron-replete, deformable cells. As such, iron-depleted erythrocytosis results in increasing symptoms due to decreased oxygen delivery to the tissues.

Hemostasis is abnormal in cyanotic CHD, due, in part, to the increased blood volume and engorged capillaries, abnormalities in platelet function, and sensitivity to aspirin or nonsteroidal anti-inflammatory agents, as well as abnormalities of the extrinsic and intrinsic coagulation system. Oral contraceptives are often contraindicated for cyanotic women because of the enhanced risk of vascular thrombosis. Symptoms of hyperviscosity can be produced in any cyanotic patient with erythrocytosis if dehydration reduces plasma volume. Phlebotomy for symptoms of hyperviscosity not due to dehydration or iron deficiency is a simple outpatient removal of 500 mL of blood over 45 min with isovolumetric replacement with isotonic saline. Acute phlebotomy without volume replacement is contraindicated. Iron repletion in decompensated iron-depleted erythrocytosis reduces iron-deficiency symptoms, but must be done gradually to avoid an excessive rise in hematocrit and resulting hyperviscosity.

PATENT DUCTUS ARTERIOSUS

The ductus arteriosus is a vessel leading from the bifurcation of the pulmonary artery to the aorta just distal to the left subclavian artery (Fig. 282-1). Normally, the vascular channel is open in the fetus but closes immediately after birth. The flow across the ductus is determined by the pressure and resistance relationships between the systemic and pulmonary circulations and by the cross-sectional area and length of the ductus. In most adults with this anomaly, pulmonary pressures are normal, and a gradient and shunt from aorta to pulmonary artery persist throughout the cardiac cycle, resulting in a characteristic thrill and a continuous "machinery" murmur with late systolic accentuation at the upper left sternal edge. In adults who were born with a large left-to-right shunt through the ductus arteriosus, pulmonary vascular obstruction (Eisenmenger's syndrome) with pulmonary hypertension, right-to-left shunting, and cyanosis have usually developed. Severe pulmonary vascular disease results in reversal of flow through the ductus; unoxygenated blood is shunted to the descending aorta; and the toes—but not the fingers—become cyanotic and clubbed, a finding termed differential cyanosis **(Fig. 282-3)**. The leading causes of death in adults with patent ductus arteriosus are cardiac failure and infective endocarditis; occasionally, severe pulmonary vascular obstruction may cause aneurysmal dilatation, calcification, and rupture of the ductus.

TREATMENT PATENT DUCTUS ARTERIOSUS

In the absence of severe pulmonary vascular disease and predominant left-to-right shunting of blood, the patent ductus should be surgically ligated or divided. Transcatheter closure has become common for appropriately shaped defects. Operation should be deferred for several months in patients treated successfully for infective endocarditis because the ductus may remain somewhat edematous and friable.

AORTIC ROOT–TO–RIGHT-HEART SHUNTS

The three most common causes of aortic root–to–right-heart shunts are congenital aneurysm of an aortic sinus of Valsalva with fistula, coronary arteriovenous fistula, and anomalous origin of the left coronary artery from the pulmonary trunk. Aneurysm of an aortic sinus of Valsalva consists of a separation or lack of fusion between the media of the aorta and the annulus of the aortic valve. Rupture usually occurs in the third or fourth decade of life; most often, the aorticocardiac fistula is between the right coronary cusp and the RV; but occasionally, when the noncoronary cusp is involved, the fistula drains into the RA. Abrupt rupture causes chest pain, bounding pulses, a continuous murmur accentuated in diastole, and volume overload of the heart. Diagnosis is confirmed by two-dimensional and Doppler echocardiographic studies; cardiac catheterization quantitates the left-to-right shunt, and thoracic aortography visualizes the fistula. Medical management is directed at cardiac failure, arrhythmias, or endocarditis. At operation, the aneurysm is closed and amputated, and the aortic wall is reunited with the heart, either by direct suture or with a patch or prosthesis. Transcatheter device closure is a less invasive and effective alternative to surgery.

Coronary arteriovenous fistula, an unusual anomaly, consists of a communication between a coronary artery and another cardiac chamber, usually the coronary sinus, RA, or RV. The shunt is usually of small magnitude, and myocardial blood flow is not usually compromised; if the shunt is large, there may be a coronary "steal" syndrome with myocardial ischemia and possible angina or ventricular arrhythmias. Potential complications include infective endocarditis; thrombus formation with occlusion or distal embolization with myocardial infarction; rupture of an aneurysmal fistula; and, rarely, pulmonary hypertension and congestive failure. A loud, superficial, continuous murmur at the lower or midsternal border usually prompts a further evaluation of asymptomatic patients. Doppler echocardiography demonstrates the site of drainage; if the site of origin is proximal, it may be detectable by two-dimensional echocardiography. Angiography (classic catheterization, CT, or magnetic resonance angiography) permits identification of the size and anatomic features of the fistulous tract, which may be closed by suture or transcatheter obliteration.

The third anomaly causing a shunt from the aortic root to the right heart is anomalous origin of the left coronary artery from the pulmonary artery. In this condition, oxygenated blood from the aortic root flows via a dilated right coronary artery and collaterals to the left coronary artery and retrograde to the lower pressure pulmonary artery circulation via the anomalous left main coronary artery (which emerges from the pulmonary artery). Myocardial infarction and