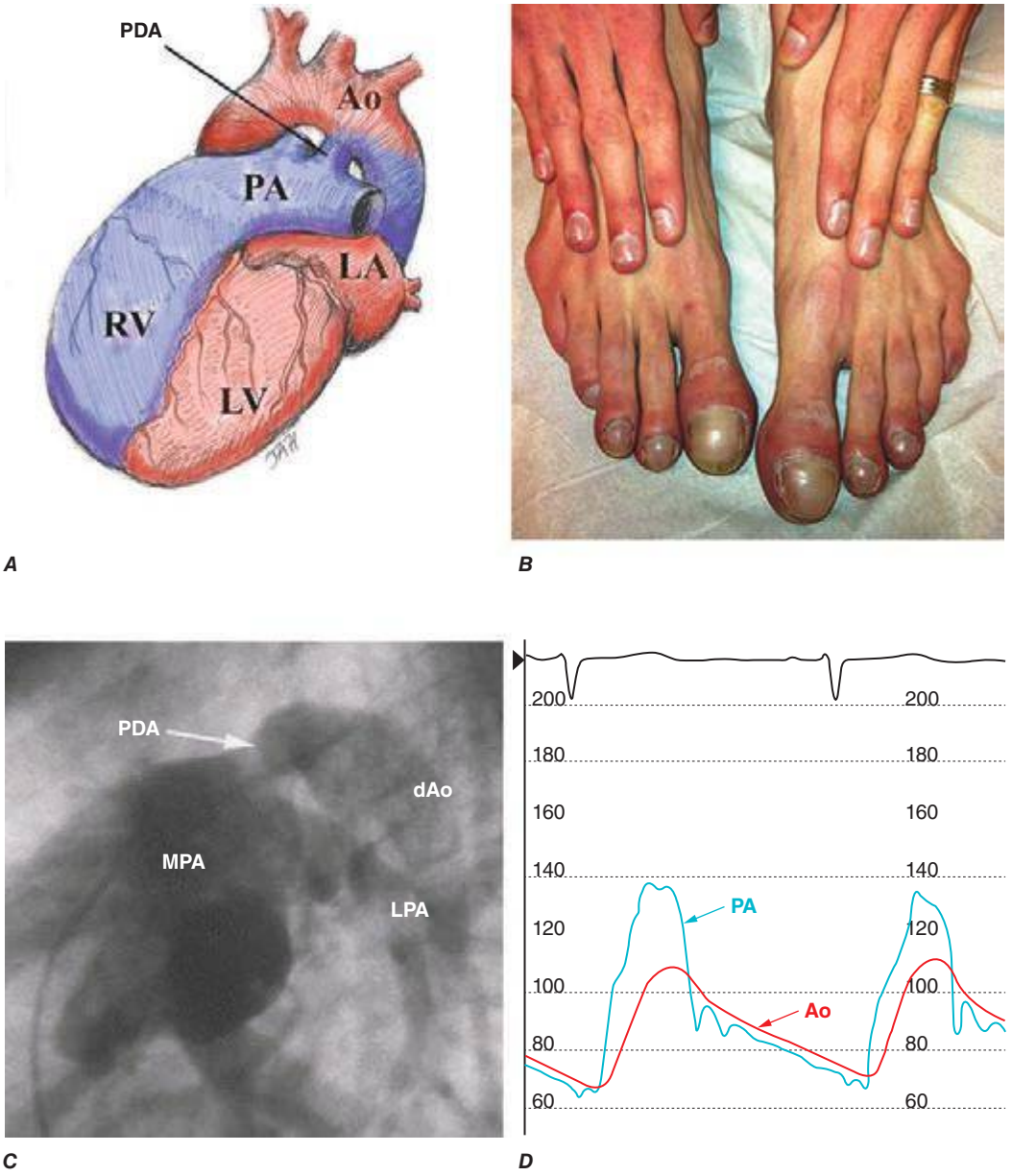

FIGURE 282-3 ***A.*** Patent ductus arteriosus (PDA) in a patient with severe pulmonary hypertension (Eisenmenger's syndrome). Due to the suprasystemic pulmonary arterial resistance, deoxygenated (cyanotic) blood from the right ventricle (RV) and pulmonary artery (PA) is shunted across the PDA to the aorta (Ao). The left atrium (LA) and left ventricle (LV) are labeled. ***B.*** Differential clubbing and cyanosis of the toes due to lower extremity perfusion by the deoxygenated blood crossing the PDA. ***C.*** Angiogram in a dilated main pulmonary artery (MPA) with shunting noted across the PDA to the descending aorta (dAo). The left pulmonary artery (LPA) is labeled. ***D.*** Direct pressure recordings in the Ao and PA demonstrating suprasystemic PA systolic pressure.

fibrosis commonly lead to death within the first year, although up to 20% of patients survive to adolescence and beyond without surgical correction. The diagnosis is supported by the ECG findings of an anterolateral myocardial infarction and left ventricular hypertrophy (LVH). Operative management of adults consists of coronary artery reimplantation, coronary artery bypass with an internal mammary artery graft, or saphenous vein–coronary artery graft.

CONGENITAL AORTIC STENOSIS

Malformations that cause obstruction to LV outflow include congenital valvular aortic stenosis, discrete subaortic stenosis, or supravalvular aortic stenosis. Bicuspid aortic valves are more common in males than in females. The congenital bicuspid aortic valve, which may initially be functionally normal, is one of the most common congenital malformations of the heart and may go undetected in early life. Because bicuspid valves may develop stenosis or regurgitation with time or be the site of infective endocarditis, the lesion may be difficult to distinguish in older adults from acquired rheumatic or degenerative calcific aortic valve disease. The dynamics of blood flow associated with a congenitally deformed, rigid aortic valve commonly lead to thickening of the cusps and, in later life, to calcification. Hemodynamically significant obstruction causes concentric hypertrophy of the LV wall. The ascending aorta is often dilated, misnamed "poststenotic" dilatation; this is due to histologic abnormalities of the aortic media and may result in aortic dissection. Diagnosis is made by echocardiography, which reveals the morphology of the aortic valve and aortic root and quantitates severity of stenosis or regurgitation. **The clinical manifestations and hemodynamic abnormalities are discussed in Chap. 283.**

TREATMENT VALVULAR AORTIC STENOSIS

In patients with diminished cardiac reserve, medical management includes the administration of digoxin and diuretics and sodium restriction while awaiting operation. A dilated aortic root may require beta blockers, angiotensin receptor blockers, or angiotensin-converting enzyme inhibitors. Aortic valve replacement is