time, right ventricular hypertrophy becomes prominent, because this chamber functions as the systemic ventricle. Concurrently, the left ventricle becomes thin-walled (atrophic) as it supports the low-resistance pulmonary circulation. Without surgery, most patients die within months. However, improving surgical interventions allow many patients with TGA to survive into adulthood.

Tricuspid Atresia

Tricuspid atresia represents complete occlusion of the tricuspid valve orifice. It results embryologically from unequal division of the AV canal; thus, the mitral valve is larger than normal, and there is right ventricular underdevelopment (hypoplasia). The circulation can be maintained by right-to-left shunting through an interatrial communication (ASD or patent foramen ovale), in addition to a VSD that affords communication between the left ventricle and the pulmonary artery arising from the hypoplastic right ventricle. Cyanosis is present virtually from birth, and there is a high early mortality.

Obstructive Lesions

Congenital obstruction to blood flow can occur at the level of the heart valves or within a great vessel. Common examples include aortic or pulmonary valve stenosis or atresia, and coarctation of the aorta. Obstruction can also occur within a chamber, as with subpulmonary stenosis in TOF.

Coarctation of the Aorta

Coarctation (narrowing, constriction) of the aorta ranks high in frequency among the common structural anomalies. It is twice as common in males as in females; interestingly, females with Turner syndrome are also frequently affected (Chapter 5). There are two classic forms: (1) an "infantile" form—often symptomatic in early childhood—with tubular hypoplasia of the aortic arch proximal to a PDA, and (2) an "adult" form with a discrete ridgelike infolding of the aorta just opposite the closed ductus arteriosus *(ligamentum arteriosum)* distal to the arch vessels (Fig. 12-8). Encroachment on the aortic lumen is variable, sometimes leaving only a small channel and at other times

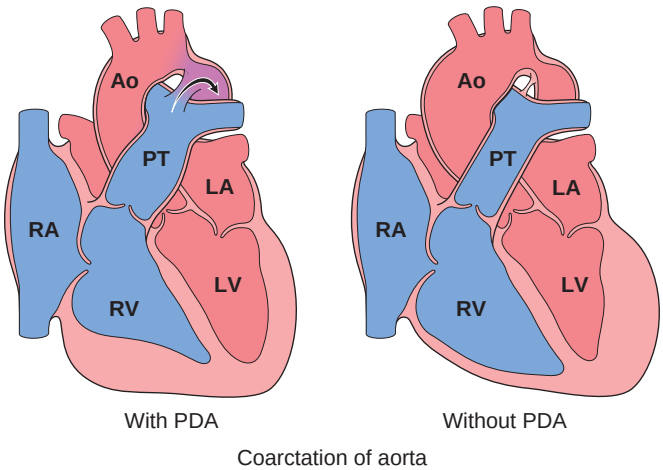

Figure 12-8 Schematic of aortic coarctation with and without patent ductus arteriosus (PDA). Ao, Aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; RA, right atrium; RV, right ventricle; PDA, patent ductus arteriosus. (Courtesy William D. Edwards, MD, Mayo Clinic, Rochester, Minn.)

producing only minimal narrowing. Although coarctation of the aorta may occur as a solitary defect, in 50% of cases it is accompanied by a bicuspid aortic valve and may also be associated with congenital aortic stenosis, ASD, VSD, mitral regurgitation, or berry aneurysms of the circle of Willis.

Clinical manifestations depend on the severity of the narrowing and the patency of the ductus arteriosus. *Coarctation of the aorta with a PDA* usually manifests early in life; indeed, it may cause signs and symptoms immediately after birth. In such cases, the delivery of unsaturated blood through the PDA produces cyanosis localized to the lower half of the body. Many such infants do not survive the neonatal period without surgical or catheter-based intervention to occlude the PDA.

The outlook is different with *coarctation of the aorta without a PDA,* unless the aortic constriction is severe. Most children are asymptomatic, and the disease may go unrecognized until well into adult life. Typically there is hypertension in the upper extremities with weak pulses and hypotension in the lower extremities, associated with manifestations of arterial insufficiency (i.e., claudication and coldness). Particularly characteristic is the development of collateral circulation between the pre-coarctation and post-coarctation arteries through enlarged intercostal and internal mammary arteries, often producing radiographically visible erosions ("notching") of the undersurfaces of the ribs.

With significant coarctations, murmurs are present throughout systole; sometimes a vibratory "thrill" is also present. The long-standing pressure overload leads to concentric left ventricular hypertrophy. With uncomplicated coarctation of the aorta, surgical resection and end-to-end anastomosis or replacement of the affected aortic segment by a prosthetic graft yields excellent results.

Pulmonary Stenosis and Atresia

Pulmonary stenosis or atresia is a relatively frequent malformation leading to obstruction at the level of the pulmonary valve. This can be mild to severe; the lesion can also be isolated or part of a more complex anomaly—either TOF or TGA. Right ventricular hypertrophy typically develops, and there is sometimes poststenotic dilation of the pulmonary artery due to injury of the wall by "jetting" blood. With coexistent subpulmonary stenosis (as in TOF), the pulmonary trunk is not dilated and may in fact be hypoplastic. When the valve is entirely atretic, there is no communication between the right ventricle and lungs. In such cases the anomaly is associated with a hypoplastic right ventricle and an ASD; blood reaches the lungs through a PDA. Mild stenosis may be asymptomatic and compatible with long life, whereas symptomatic cases require surgical correction.

Aortic Stenosis and Atresia

Congenital narrowing and obstruction of the aortic valve can occur at three locations: valvular, subvalvular, and supravalvular. Congenital aortic stenosis is an isolated lesion in 80% of cases. With *valvular aortic stenosis* the cusps may be hypoplastic (small), dysplastic (thickened, nodular), or abnormal in number (usually acommissural or unicommissural). In severe congenital aortic stenosis or atresia, obstruction of the left ventricular outflow tract leads to