crescendo-decrescendo systolic murmur, is heard at the right upper sternal border. Often, a continuous systolic murmur is heard over the left scapula. It is related to continuous flow across the coarctation of the aorta.

Diagnosis

Patients with significant coarctation of the aorta typically show various degrees of left atrial (LA) and LV enlargement on an ECG. The chest radiograph typically demonstrates normal heart size with dilation of the ascending aorta and kinking or double contouring in the region of the descending aorta in the area of the coarctation, producing the characteristic figure-3 sign.

Most adult patients have rib notching. It is caused by the dilated intercostal collateral arteries eroding the undersurface of the ribs. Echocardiography is used to identify site, structure, and degree of stenosis or restenosis. Echocardiography is valuable for identifying other lesions, LV systolic function, and degree of LV hypertrophy.

Cardiac catheterization remains the gold standard for determining the anatomy and absolute degree of stenosis. In adult patients, cardiac catheterization with balloon dilation and stent placement has become the procedure of choice for the treatment of recoarctation. Newer magnetic resonance imaging (MRI) methods are quite good for imaging coarctation, defining the arch vessel anatomy, and identifying collaterals.

Treatment

Patients with significant native or residual coarctation of the aorta (i.e., symptomatic with a peak gradient across the coarctation of ≥30 mm Hg) should be considered for surgical repair or catheter intervention with balloon angioplasty with or without stent placement. Surgical repair in the adult patient is technically difficult and is associated with high rates of morbidity. As result, catheter based intervention has become the preferred method in most experienced congenital heart disease centers.

Prognosis

After surgical repair, long-term survival is good but directly correlates with the age at repair. Those repaired after 14 years of age have a lower 20-year survival rate than those repaired earlier (79% vs. 91%). Long-term outcome data for catheter-based treatment is limited, but studies suggest that stented patients have lower acute and long-term complications at 60 months (25% for surgery vs. 12.5% for stents). Irrespective of the type of repair, the most common long-term complication is persistent or new systemic hypertension at rest or during exercise. Other long-term complications include aneurysms of the ascending or descending aorta (especially after Dacron patch repair), recoarctation at the site of previous repair, coronary artery disease, aortic stenosis or regurgitation (in the setting of a bicuspid aortic valve), rupture of an intracranial aneurysm, and endocarditis.

Patent Ductus Arteriosus

Definition and Epidemiology

Patent ductus arteriosus (PDA) represents 9% to 12% of congenital heart defects. It is patent in the fetus but normally closes within several days of birth. However, it remains open in about 1 of 2500 to 5000 births. In infants born prematurely, the incidence is even higher, occurring in 8 of 1000 live births. The incidence of PDA is 30 times greater for babies born at high altitudes than for those born at sea level.

Pathology

A PDA allows transit of blood from the aorta into the pulmonary artery and recirculation through the pulmonary vasculature and the left side of the heart. This can result in left-sided chamber enlargement (see Fig. 6-1). As with VSDs, the size of the defect is the primary determinant of the clinical course in the adult patient. PDAs can be clinically categorized as silent PDAs; small, hemodynamically insignificant PDAs; moderate-size PDAs; large PDAs; and previously repaired PDAs.

Clinical Presentation

A silent PDA is a tiny defect that cannot be heard by auscultation and is detected only by other nonclinical means such as echocardiography. Life expectancy is always normal for this population, and the risk of endocarditis is extremely low.

Patients with a small PDA have an audible, long-ejection or continuous murmur that is heard best at the left upper sternal border and radiating to the back. They have normal peripheral pulses. Because there is negligible left-to-right shunting, these patients have normal LA and LV sizes and normal pulmonary artery pressure. Like those with silent PDAs, these patients are asymptomatic and have a normal life expectancy. However, they do have a higher risk of endocarditis.

Patients with moderate-size PDAs may be diagnosed during adulthood. These patients often have wide, bouncy peripheral pulses and an audible, continuous murmur. They have significant volume overload and develop some degree of LA and LV enlargement and some degree of pulmonary hypertension. These patients are symptomatic with dyspnea, palpitations, and heart failure. Patients with large PDAs typically have signs of severe pulmonary hypertension and Eisenmenger's syndrome. By adulthood, the continuous murmur is typically absent, and there is differential cyanosis (i.e., lower extremity saturations are lower than the right arm saturation).

Diagnosis

Patients with silent and small PDAs appear normal by echocardiography and chest radiography. Calcifications may be seen on the posteroanterior and lateral films of an older patient with a PDA. In patients with significant left-to-right shunting, there typically is dilation of the central pulmonary arteries with increased pulmonary vascular markings. On an ECG, broad P waves and tall QRS complexes suggest LA and LV volume overload. A tall R wave in lead V_1 with a right axis deviation suggests significant pulmonary hypertension. Echocardiography is important to estimate the size of the defect, degree of LA or LV enlargement, and degree of pulmonary artery hypertension.

Treatment

All patients with clinical evidence of a PDA are at increased risk for endocarditis. Except for patients with small or silent PDAs and those with severe, irreversible pulmonary hypertension, PDA closure should be considered. Catheter device closure is the preferred method in most centers. Surgical closure is reserved for