patients with PDAs too large for device closure and for distorted anatomy such as a large ductal aneurysm.

Prognosis

Patients with a large PDA who have developed Eisenmenger's syndrome have a prognosis similar to that of other patients with Eisenmenger's syndrome. Patients who underwent PDA repair before the development of pulmonary hypertension have a normal life expectancy without restrictions.

Pulmonary Valve Stenosis

Definition and Epidemiology

Pulmonary valve stenosis occurs in approximately 4 of 1000 live births and constitutes 5% to 8% of congenital cardiac defects. It is one of the most common adult forms of unoperated congenital heart disease. It can occur in isolation or with other congenital heart defects, such as an ASD.

Pathology

In congenital pulmonary valve stenosis, the pulmonary valve leaflets are often fused or thickened, which obstructs blood flow out of the right ventricle. The obstruction elevates RV pressure, and compensatory RV hypertrophy develops. Pulmonary stenosis is often tolerated better than aortic stenosis. Over time, RV dilation and dysfunction may occur.

Clinical Presentation

Most patients with pulmonary valve stenosis are asymptomatic and have a cardiac murmur at presentation. Most unoperated adults with severe stenosis have jugular venous distention, and on palpation, a RV lift at the left lower sternal border and a thrill at the left upper sternal border can be identified. On auscultation, the second heart sound is widely split, and a systolic ejection click may or may not be heard, depending on the mobility of the pulmonary valve leaflets. In most cases, there is a harsh, crescendo-decrescendo systolic ejection murmur, which is heard best at the left upper sternal border; it radiates to the back and varies with inspiration.

Diagnosis

With moderate to severe pulmonary valve stenosis, the ECG demonstrates right axis deviation, RV hypertrophy, and RA enlargement. The ECG is usually normal for patients with mild pulmonary valve stenosis. On the chest radiograph, a prominent main pulmonary artery caused by poststenotic dilatation is a common finding regardless of the degree of stenosis. In patients with severe pulmonary valve stenosis, cardiomegaly due to RA and RV enlargement is often seen.

Echocardiography is the diagnostic method of choice. It allows visualization of the valve anatomy and degree of stenosis and enables estimation of the valve gradient.

Treatment

Survival into adult life and the need for intervention directly correlate with the degree of obstruction. In the Second Natural History Study of Congenital Heart Disease, patients with trivial stenosis (i.e., peak gradient ≤25 mm Hg) who were followed for 25 years remained asymptomatic and had no significant progression of obstruction over time. For those with moderate pulmonary valve stenosis (i.e., peak gradient between 25 and 49 mm Hg), there was an approximately 20% chance of requiring intervention by 25 years of age. Most patients with severe stenosis (i.e., peak gradient of ≥50 mm Hg) require intervention (i.e., surgery or balloon valvuloplasty) by age 25 years. Patients with moderate to severe pulmonary stenosis should be considered for intervention even in the absence of symptoms.

Since 1985, percutaneous balloon valvuloplasty has been the accepted treatment for patients of all ages. Before 1985, surgical valvotomy had been the gold standard. Today, surgical valvotomy is reserved for patients who are unlikely to have successful results from balloon valvuloplasty, such as those with an extremely dysplastic or calcified valve.

Prognosis

After surgical valvotomy for isolated pulmonary stenosis, long-term survival is excellent. However, with longer follow-up the incidence of late complications and the need for reintervention do increase. The most common indication for reintervention is pulmonary valve replacement for severe pulmonary regurgitation. Other long-term complications include recurrent atrial arrhythmias, endocarditis, and residual subpulmonary obstruction.

Aortic Valve Stenosis

Definition and Epidemiology

Aortic valve stenosis is a common abnormality in adults with congenital heart disease. It is usually caused by a bicuspid aortic valve, which occurs in 1% to 2% of adults and is three times more common in males. It typically is an isolated lesion but can be associated with other defects such as coarctation of the aorta or VSD.

Pathology

Aortic valve stenosis results in pressure overload of the left ventricle, which increases wall stress and causes compensatory LV hypertrophy. Diastolic dysfunction and oxygen delivery-demand mismatch ensues. The patient may remain well compensated and asymptomatic for many years, but compensatory mechanisms eventually begin to fail, and LV dysfunction can develop. Patients with a bicuspid aortic valve have abnormal structure of the aortic wall that often leads to ascending aortic dilation.

Clinical Presentation

Most patients with aortic valve stenosis are asymptomatic and are diagnosed after a murmur is detected. The severity of obstruction at the time of diagnosis correlates with the pattern of progression. Symptoms are rare until patients have severe aortic valve stenosis (i.e., mean gradient by echocardiography of ≥40 mm Hg). Symptoms include chest pain, exertional dyspnea, near-syncope, and syncope. With any of these symptoms, the risk of sudden cardiac death is very high, and surgical intervention is mandated.

Patients with moderate to severe stenosis typically have decreased peripheral pulses, an increased apical impulse, and a