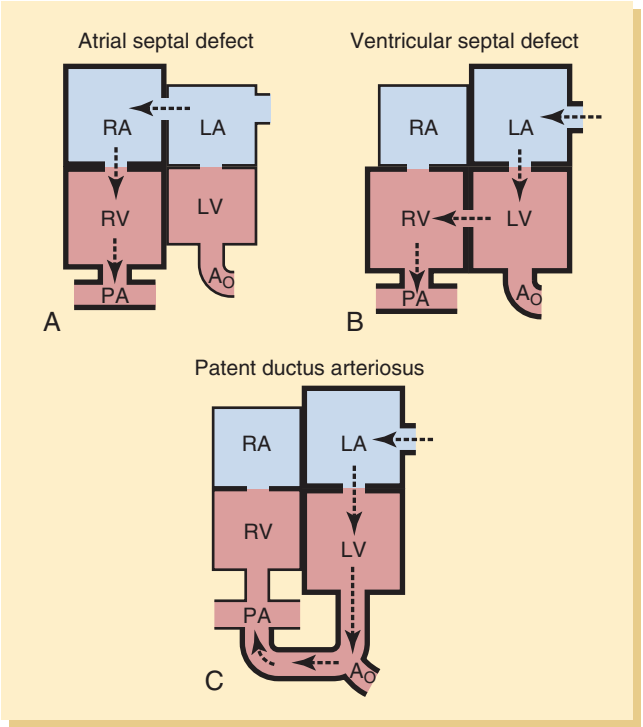

FIGURE 6-1 The diagram shows three types of shunt lesions that commonly survive until adulthood and their effects on chamber size. **A,** Uncomplicated atrial septal defect with left-to-right shunt flow across the interatrial septum, resulting in dilation of the right atrium (RA), right ventricle (RV), and pulmonary artery (PA). **B,** Uncomplicated ventricular septal defect, resulting in dilation of the RV, left atrium (LA), and left ventricle (LV). **C,** Uncomplicated patent ductus arteriosus, resulting in dilation of the LA, LV, and PA. Ao, Aorta. (From Liberthson RR, Walkdman H: Congenital heart disease in the adult. In Kloner RA, editor: Guide to cardiology, ed 3, Greenwich, Conn., 1991, Le Jacq Communications, pp 24–27.)

Clinical Presentation

Although most individuals with an ASD are diagnosed during childhood after a murmur is noticed, a few patients have symptoms for the first time as adults. Most patients are asymptomatic during the first and second decades of life. In the third decade, an increasing numbers of patients develop exercise intolerance, palpitations due to atrial arrhythmias, and cardiac enlargement on the chest radiograph. In patients with ASDs, the RV impulse at the left lower sternal border often has increased force compared with normal. On auscultation, the second heart sound typically is widely split and fixed (i.e., does not vary with inspiration).

All patients have a systolic ejection murmur, which is best heard at the left upper sternal border and is related to increased flow across a usually normal pulmonary valve. When there is a large left-to-right shunt, a mid-diastolic murmur can be heard at the left lower sternal border; it is related to increased flow across a normal tricuspid valve. When a mid-diastolic murmur is identified, the degree of left-to-right shunt is considered to be 1.5 times normal. In the setting of a primum ASD, an additional holosystolic murmur at the apex may be caused by a cleft in the anterior leaflet of the mitral valve, resulting in mitral regurgitation.

Diagnosis

On the electrocardiogram (ECG), the features of ASD depend on the size and type of defect. In the setting of a large ostium secundum, sinus venosus, or unroofed coronary sinus defect, the ECG typically demonstrates evidence of right atrial (RA) enlargement, RV hypertrophy, and right axis deviation. In the setting of an ostium primum ASD, like other forms of atrioventricular defects, there is a superior axis. The chest radiograph is helpful for evaluating the degree of left-to-right shunting. With a small shunt, the radiograph will be normal. As the shunt increases in size, the heart size and pulmonary vascular markings also increase.

The diagnosis of an ASD and its location is confirmed by transthoracic echocardiography in most cases. A sinus venosus ASD is the exception. In this setting, transesophageal echocardiography may be necessary. Cardiac catheterization is rarely performed to diagnose an ASD. However, transcatheter closure has become the preferred treatment option for most ostium secundum defects.

Treatment

The treatment of ASDs involves surgical or transcatheter device closure. For secundum ASD, surgical closure and transcatheter device closure are accepted treatment options. Device closure is the most commonly used technique for closure of secundum defects. This technique, however, requires an adequate rim of septal tissue around the entire defect to allow for device stabilization. For ostium primum, sinus venosus, and unroofed coronary sinus forms of ASDs, surgical closure remains the only option.

Prognosis

Most patients who have undergone early closure of a defect have excellent long-term survival rates with low morbidity rates if repair is undertaken before 25 years of age. Older age at repair is associated with decreased late survival rates and an associated increased risk of atrial arrhythmias, thromboembolic events, and pulmonary hypertension. After the age of 40 years for patients with unrepaired ASDs, the mortality rate increases by 6% per year, and more than 20% of patients develop atrial fibrillation. By 60 years of age, the number of patients with atrial fibrillation increases to more than 60%. Long-term rates of late complications and survival after transcatheter device closure remain unknown.

Ventricular Septal Defects

Definition and Epidemiology

Ventricular septal defects (VSDs) occur in 1.5 to 3.5 of 1000 live births. They constitute 20% of congenital heart defects.

There are four types of VSD: perimembranous, muscular, supracristal, and inlet. Perimembranous VSDs are the most common, comprising 70% of all VSDs. The membranous septum is relatively small and sits directly under the aortic valve. Perimembranous VSDs involve the membranous septum and typically extend into the muscular tissue adjacent to the membranous septum. If not large, these defects may close