Ventricular Septal Defects in Adults

David S. Williams, MD

Ventricular septal defects are one of the most common congenital cardiac malformations and can be associated with many types of congenital and acquired heart disease.

First described in 1847, ventricular septal defects account for 20% of congenital cardiovascular malformations and 10% of those are diagnosed in adults. Prevalence has increased due to improved diagnostic testing and is estimated at 1.17/1000 live births and 0.5/1000 adults. They can be associated with atrial septal defect (35%), patent ductus arteriosus (22%), right aortic arch (13%) and less often pulmonary stenosis.

The size of an isolated defect determines the direction and volume of the left-to-right shunt, which results in volume overload of the left atrium, both ventricles, and the pulmonary arteries. It is the volume of the shunt that determines the clinical presentation and natural history of the patient. About 30%–50% of small defects will spontaneously close; those which do not will not become larger. Expectation of life is normal with a small (about 3%) lifetime risk of endocarditis, which is more common in older men. Small defects are usually asymptomatic and may also represent a larger defect that has undergone incomplete closure. Larger defects are uncommon in adults unless associated with protective valvular or subvalvular pulmonary stenosis. Because larger defects will soon cause signs and symptoms of volume overload or be complicated by pulmonary hypertension (Eisenmenger complex), they will present and warrant intervention much earlier in life. Long-term survival in these cases will depend on age, functional class, cardiomegaly, pulmonary artery pressure, and associated anomalies at time of surgery.

Most ventricular septal defects can be diagnosed by auscultation: the murmur produced is a harsh high-frequency holosystolic plateau-shaped sound, best appreciated in the left third and fourth intercostal spaces. The noninvasive method of choice for evaluation is the echocardiogram, which has a sensitivity for detection of 88%–95%. The electrocardiogram is usually normal but may show an intraventricular conduction delay or right bundle-branch block. Likewise, the chest x-ray is usually normal, although enlargement of the cardiac silhouette may result from volume overload in direct relationship to the magnitude of the shunt.

The treatment of an isolated ventricular septal defect depends on the type and size of the defect, shunt severity, pulmonary vascul-
lar resistance, functional capacity, and associated anomalies. Surgical closure reduces the risk of endocarditis by 50%, reduces pulmonary artery pressure, improves functional classification, and increases long-term survival. Studies have shown an operative mortality rate less than 2% in uncomplicated cases, and a 25-year survival rate of 89% after surgical repair. Residual defects occur in up to 34% of operative repairs, regardless of surgical approach, but are usually small and hemodynamically insignificant. The risk for conduction blocks and serious dysrhythmias continues even after surgery and correlates, as one might expect, with higher functional classification, cardiomegaly and increased pulmonary artery pressure.

In the figures, the most common membranous defect (75%–80%) and less common muscular defect (5%–20%) are shown.

REFERENCES