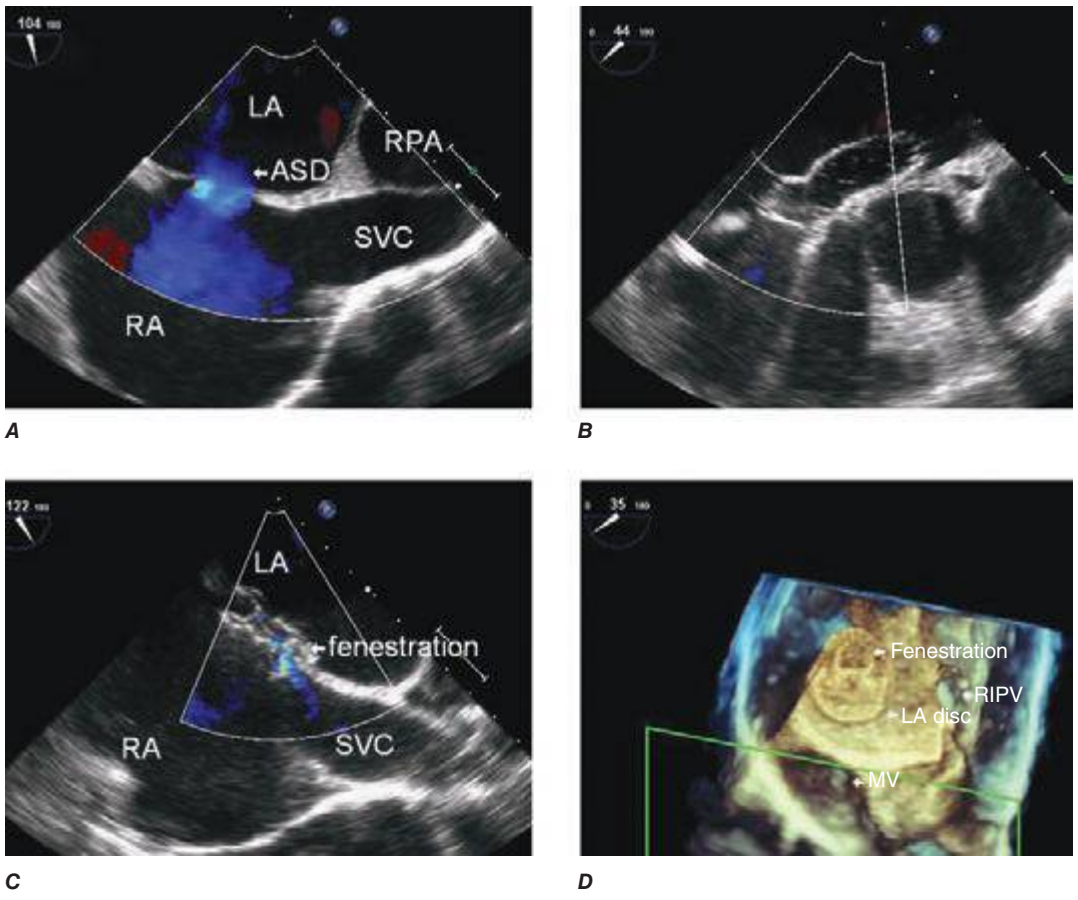

FIGURE 282-2 ***A.*** Transesophageal echocardiogram demonstrating a secundum-type atrial septal defect (ASD) with shunting from the left atrium (LA) to the right atrium (RA). The right pulmonary artery (RPA) and superior vena cava (SVC) are labeled. ***B.*** Transcatheter balloon sizing of the ASD. ***C.*** Atrial septal occluder placement with a small manually created "fenestration" within the device that continues to allow a small amount of flow from the LA to the RA; this is used as a means of preventing left atrial hypertension after ASD closure. Left atrial hypertension may occur in older patients with decreased left ventricular compliance. ***D.*** Three-dimensional image of the septal occluder en-face; note the fenestration in the LA disc. The mitral valve (MV) and right inferior pulmonary vein (RIPV) are labeled.

VENTRICULAR SEPTAL DEFECT

VSD is one of the most common of all cardiac birth defects, either as an isolated defect or as a component of a combination of anomalies (Fig. 282-1). The VSD is usually single and situated in the membranous or midmuscular portion of the septum. The functional disturbance depends on its size and on the status of the pulmonary vascular bed. Only small- or moderate-size VSDs are seen initially in adulthood, as most patients with an isolated large VSD come to medical or surgical attention early in life.

A wide spectrum exists in the natural history of VSD, ranging from spontaneous closure to congestive cardiac failure and death in infancy. Included within this spectrum is the possible development of pulmonary vascular obstruction, RV outflow tract obstruction, aortic regurgitation, or infective endocarditis. Spontaneous closure is more common in patients born with a small VSD, which occurs in early childhood in most. The pulmonary vascular bed is often a principal determinant of the clinical manifestations and course of a given VSD and feasibility of surgical repair. Increased pulmonary arterial pressure results from increased pulmonary blood flow and/or resistance, the latter usually the result of obstructive, obliterative structural changes within the pulmonary vascular bed. It is important to quantitate and compare pulmonary-to-systemic flows and resistances in patients with severe pulmonary hypertension. The term Eisenmenger's syndrome is applied to patients with a large communication between the two circulations at the aortopulmonary, ventricular, or atrial levels and bidirectional or predominantly right-to-left shunts because of high resistance and obstructive pulmonary hypertension.

Patients with large VSDs and pulmonary hypertension are at greatest risk for developing pulmonary vascular disease. Large VSDs should be corrected early in life when pulmonary vascular disease is not severely elevated. In patients with Eisenmenger's syndrome, symptoms in adult life consist of exertional dyspnea, chest pain, syncope, and hemoptysis. The right-to-left shunt leads to cyanosis, clubbing, and erythrocytosis (see below). The degree to which pulmonary vascular resistance is elevated before operation is a critical factor determining prognosis. If the pulmonary vascular resistance is one-third or less of the systemic value, progression of pulmonary vascular disease after operation is unusual; however, if a moderate to severe increase in pulmonary vascular resistance exists preoperatively, either no change or a progression of pulmonary vascular disease is common postoperatively. Pregnancy is contraindicated in Eisenmenger's syndrome. The mother's health is most at risk if she has a cardiovascular lesion associated with pulmonary vascular disease and pulmonary hypertension (e.g., Eisenmenger's physiology or mitral stenosis) or severe LV outflow tract obstruction (e.g., aortic stenosis), but she is also at risk of death with any malformation that may cause heart failure or a hemodynamically important arrhythmia. The fetus is most at risk with maternal cyanosis, heart failure, or pulmonary hypertension.

RV outflow tract obstruction develops in ~5–10% of patients who present in infancy with a moderate to large left-to-right shunt. With time, as subvalvular RV outflow tract obstruction progresses, the findings in these patients whose VSD remains sizable begin to resemble more closely those of the cyanotic tetralogy of Fallot. In ~5% of patients, aortic valve regurgitation results from insufficient cusp tissue or prolapse of the cusp through the interventricular defect; the aortic regurgitation then complicates and dominates the clinical course. Echocardiography with spectral and color Doppler examination defines the number and location of defects in the ventricular septum