right-sided pressures, for example, during a bowel movement, coughing, or sneezing, can produce brief periods of right-to-left shunting, with the possibility of paradoxical embolism.

### *Ventricular Septal Defect*

VSDs are incomplete closures of the ventricular septum, allowing free communication of blood between the left to right ventricles; they are the most common form of congenital heart disease (Table 12-2 and Fig. 12-4*B*).

#### MORPHOLOGY

VSDs are classified according to their size and location. Most are about the size of the aortic valve orifice, and about 90% occur in the region of the membranous interventricular septum (**membranous VSD**; Fig. 12-5). The remainder occur below the pulmonary valve **(infundibular VSD)** or within the muscular septum. Although most VSDs are single, those in the muscular septum may be multiple.

***Clinical Features.*** Most VSDs that clinically manifest in the pediatric age group are associated with other congenital cardiac anomalies such as Tetralogy of Fallot; only 20% to 30% are isolated. Conversely, if a VSD is first detected only in an adult, it is usually an isolated defect. The functional consequences of a VSD depend on the size of the defect and whether there are associated right-sided malformations. Thus, large VSDs cause difficulties virtually from birth; smaller lesions are generally well tolerated for years and may not be recognized until much later in life. Moreover, approximately 50% of small muscular VSDs close spontaneously. Large defects are usually membranous or infundibular, and they generally cause significant left-to-right shunting, leading to early right ventricular hypertrophy and pulmonary hypertension. Over time, large unclosed VSDs almost universally lead to irreversible pulmonary vascular disease, ultimately resulting in shunt reversal, cyanosis, and death. Surgical or catheter-based closure of asymptomatic VSD is generally delayed beyond infancy, in hope of spontaneous closure. Early correction, however, must be performed for large defects to prevent the development of irreversible obstructive pulmonary vascular disease.

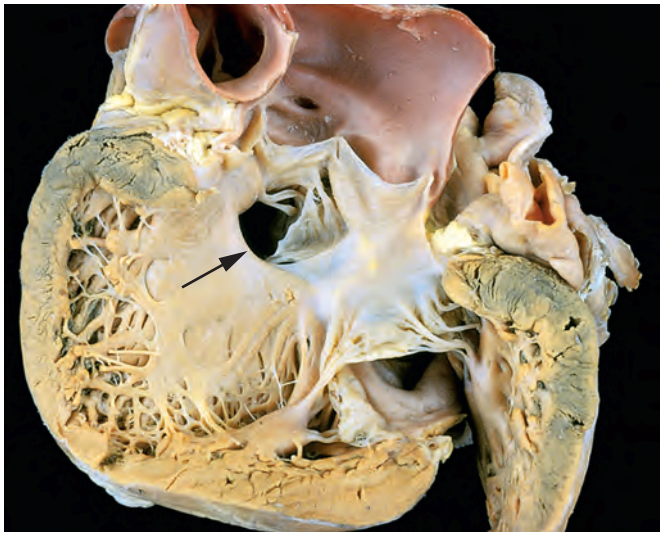

**Figure 12-5** A ventricular septal defect (membranous type), denoted by the arrow. (Courtesy William D. Edwards, MD, Mayo Clinic, Rochester, Minn.)

### *Patent Ductus Arteriosus*

The *ductus arteriosus* arises from the pulmonary artery and joins the aorta just distal to the origin of the left subclavian artery. During intrauterine life, it permits blood flow from the pulmonary artery to the aorta, thereby bypassing the unoxygenated lungs. Shortly after birth in healthy term infants, the ductus constricts and is functionally closed after 1 to 2 days; this occurs in response to increased arterial oxygenation, decreased pulmonary vascular resistance, and declining local levels of prostaglandin $E_2$. Complete structural obliteration occurs within the first few months of extrauterine life to form the *ligamentum arteriosum.* Ductal closure is often delayed (or even absent) in infants with hypoxia (due to respiratory distress or heart disease), or when PDA occurs in association with other congenital defects, particularly VSDs that increase pulmonary vascular pressures. PDAs account for about 7% of cases of congenital heart disease (Table 12-2 and Fig. 12-4*C*), and 90% of these are isolated defects.

PDA produces a characteristic continuous harsh "machinery-like" murmur. The clinical impact of a PDA depends on its diameter and the cardiovascular status of the individual. PDA is usually asymptomatic at birth, and a narrow PDA may have no effect on the child's growth and development. Because the shunt is initially left-to-right, there is no cyanosis. However, with large shunts, the additional volume and pressure overloads eventually produce obstructive changes in small pulmonary arteries, leading to reversal of flow and its associated consequences. In general, isolated PDA should be closed as early in life as is feasible. Conversely, preservation of ductal patency (by administering prostaglandin E) may be life saving for infants with various congenital malformations that obstruct the pulmonary or systemic outflow tracts. In aortic valve atresia, for example, a PDA may provide the entire systemic blood flow.

## Right-to-Left Shunts

The diseases in this group cause cyanosis early in postnatal life (*cyanotic* congenital heart disease). Tetralogy of Fallot, the most common in this group, and transposition of the great arteries are illustrated schematically in Figure 12-6. The others include persistent truncus arteriosus, tricuspid atresia, and total anomalous pulmonary venous connection.

### *Tetralogy of Fallot*

The four cardinal features of TOF are (1) VSD, (2) obstruction of the right ventricular outflow tract (subpulmonary stenosis), (3) an aorta that overrides the VSD, and (4) right ventricular hypertrophy (Fig. 12-6*A*). All of these features result embryologically from anterosuperior displacement of the infundibular septum.

#### MORPHOLOGY

The heart is typically enlarged and is classically "boot-shaped" due to marked right ventricular hypertrophy. The VSD is usually large with the aortic valve at the superior border, thereby