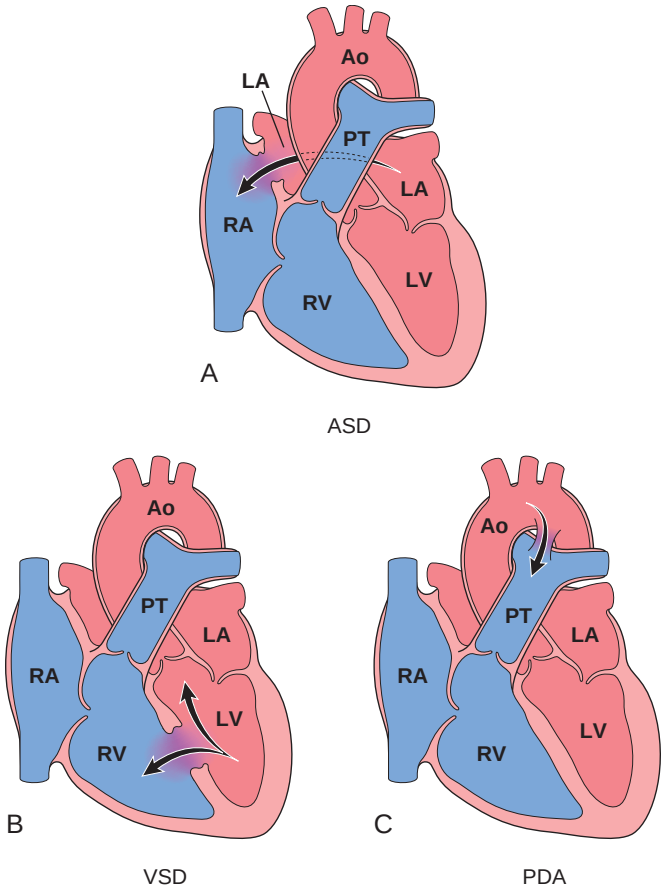

Figure 12-4 Common congenital left-to-right shunts (*arrows* indicate the direction of blood flow). **A,** Atrial septal defect (ASD). **B,** Ventricular septal defect (VSD). With VSD the shunt is left-to-right, and the pressures are the same in both ventricles. Pressure hypertrophy of the right ventricle and volume hypertrophy of the left ventricle are generally present. **C,** Patent ductus arteriosus (PDA). Ao, Aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; RA, right atrium; RV, right ventricle.

Figure 12-4. ASD typically increases only right ventricular and pulmonary outflow volumes, while VSD and PDA cause both increased pulmonary blood flow and pressure. Depending on their size and location, manifestations of these shunts range in severity from no symptoms at all to fulminant heart failure.

Atrial Septal Defect

ASDs are abnormal, fixed openings in the atrial septum caused by incomplete tissue formation that allows communication of blood between the left and right atria; ASDs are usually asymptomatic until adulthood (Table 12-2 and Fig. 12-4*A*). ASD should not be confused with *patent foramen ovale* (*PFO*; see later), which represents the failure to close a foramen (hole) that is part of normal development. Both ASD and PFO result from defects in the formation of the interatrial septum; a brief summary of the developmental stages of this structure follows:

- The *septum primum* is a crescent-shaped membranous ingrowth that sits posteriorly between the right and left atria and partially separates them; the remaining anterior opening, called the *ostium primum*, allows movement of blood from the right to left atrium during fetal development.
- Before the growing septum primum completely obliterates the ostium primum, it develops a second posterior opening called the *ostium secundum*.
- The *septum secundum* is a subsequent membranous ingrowth located to the right and anterior of the septum primum.
- As the septum secundum grows, it also leaves a small opening called the *foramen ovale* that is continuous with the ostium secundum—the foramen ovale/ostium secundum permits continued right-to-left shunting of blood during intrauterine development.
- The septum secundum continues to enlarge until it forms a flap of tissue that covers the foramen ovale on its left side.

This flap of tissue opens and closes in response to pressure gradients between the left and right atria; the valve opens only when the pressure is greater in the right atrium. In fetal life, the lungs are nonfunctional, and the pressure in the *pulmonary circulation* is greater than that of the *systemic circulation*; thus, the right atrium is under higher pressures than the left atrium, and the valve of the foramen ovale is normally open. At birth, with lung expansion, the pulmonary vascular pressures drop, and the right atrial pressures fall below those in the left atrium. As a result, the valve of the foramen ovale closes—and usually permanently seals (see later).

MORPHOLOGY

ASDs are classified according to their location. **Secundum ASD** (90% of all ASD) result from a deficient septum secundum formation near the center of the atrial septum. These are usually not associated with other anomalies, may be of any size, and can be multiple or fenestrated. **Primum anomalies** (5% of ASD) occur adjacent to the AV valves and are often associated with AV valve abnormalities and/or a VSD. **Sinus venosus defects** (5%) are located near the entrance of the superior vena cava and can be associated with anomalous pulmonary venous return to the right atrium.

Clinical Features. ASDs result in a left-to-right shunt, largely because pulmonary vascular resistance is considerably less than systemic vascular resistance and because the compliance (distensibility) of the right ventricle is much greater than that of the left. Pulmonary blood flow may be two to eight times normal. A murmur is often present as a result of excessive flow through the pulmonary valve and/or through the ASD. Despite the right-sided volume overload, ASDs are generally well tolerated and usually do not become symptomatic before age 30; irreversible pulmonary hypertension is unusual. ASD closure (surgical or catheter-based) reverses the hemodynamic abnormalities and prevents the complications, including heart failure, paradoxical embolization, and irreversible pulmonary vascular disease. Mortality is low, and long-term survival is comparable to that of the normal population.

Patent Foramen Ovale. The foramen ovale closes permanently in approximately 80% of people by 2 years of age. However, in the remaining 20%, the unsealed flap can open if right-sided pressures become elevated. Thus, sustained pulmonary hypertension or even transient increases in