TABLE 282-3 COMPLEX ADULT CONGENITAL HEART DISEASE

Cyanotic congenital heart diseases (all forms)
Eisenmenger's syndrome
Ebstein's anomaly
Tetralogy of Fallot or pulmonary atresia (all forms)
Transposition of the great arteries
Single ventricle; tricuspid or mitral atresia
Double-outlet ventricle
Truncus arteriosus
Fontan or Rastelli procedures

ovale. Anatomic obliteration of the foramen ovale ordinarily follows its functional closure soon after birth, but residual "probe patency" is a common normal variant; ASD denotes a true deficiency of the atrial septum and implies functional and anatomic patency. The magnitude of the left-to-right shunt depends on the ASD size, ventricular diastolic properties, and the relative impedance in the pulmonary and systemic circulations. The left-to-right shunt causes diastolic overloading of the RV and increased pulmonary blood flow. Patients with ASD are usually asymptomatic in early life, although there may be some physical underdevelopment and an increased tendency for respiratory infections; cardiorespiratory symptoms occur in many older patients. Beyond the fourth decade, a significant number of patients develop atrial arrhythmias, pulmonary arterial hypertension, and right heart failure. Patients exposed to the chronic environmental hypoxemia of high altitude tend to develop pulmonary hypertension at younger ages. In older patients, left-to-right shunting across the ASD increases as progressive systemic hypertension and/or coronary artery disease (CAD) result in reduced compliance of the LV.

Physical Examination Examination usually reveals a prominent RV impulse and palpable pulmonary artery pulsation. The first heart sound is normal or split, with accentuation of the tricuspid valve closure sound. Increased flow across the pulmonic valve is responsible for a midsystolic pulmonary outflow murmur. The second heart sound is widely split and is fixed in relation to respiration. A mid-diastolic rumbling murmur, loudest at the fourth intercostal space and along the left sternal border, reflects increased flow across the tricuspid valve. In ostium primum ASD, an apical holosystolic murmur indicates associated mitral or tricuspid regurgitation or a ventricular septal defect (VSD).

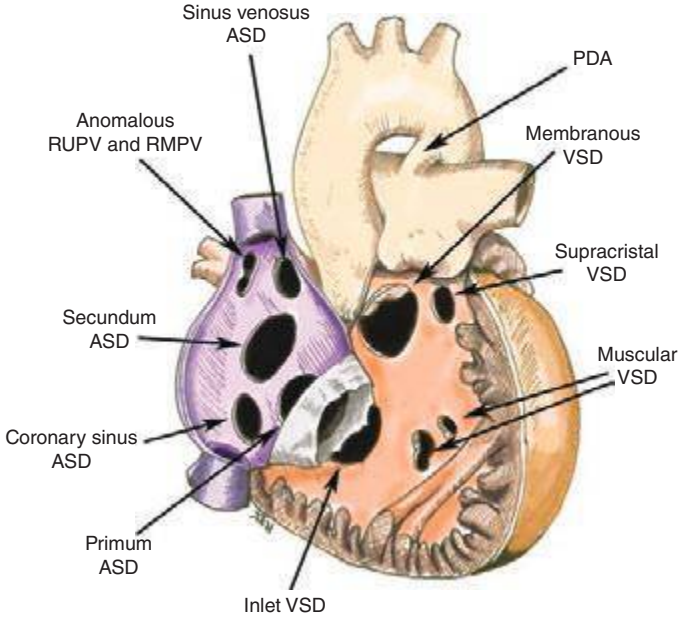

FIGURE 282-1 Types and locations of congenital cardiac defects. ASD, atrial septal defect; PDA, patent ductus arteriosus; RMPV, right middle pulmonary vein; RUPV, right upper pulmonary vein; VSD, ventricular septal defect.

These findings are altered when increased pulmonary vascular resistance causes diminution of the left-to-right shunt. Both the pulmonary outflow and tricuspid inflow murmurs decrease in intensity, the pulmonic component of the second heart sound and a systolic ejection sound are accentuated, the two components of the second heart sound may fuse, and a diastolic murmur of pulmonic regurgitation appears. Cyanosis and clubbing accompany the development of a right-to-left shunt (see "Ventricular Septal Defect" later in this chapter). In adults with an ASD and atrial fibrillation, the physical findings may be confused with mitral stenosis with pulmonary hypertension because the tricuspid diastolic flow murmur and widely split second heart sound may be mistakenly thought to represent the diastolic murmur of mitral stenosis and the mitral "opening snap," respectively.

Electrocardiogram In ostium secundum ASD, electrocardiogram (ECG) usually shows right-axis deviation and an rSr′ pattern in the right precordial leads representing enlargement of the RV outflow tract. An ectopic atrial pacemaker or first-degree heart block may occur with the sinus venous ASD. In ostium primum ASD, the RV conduction defect is accompanied by left superior axis deviation and counterclockwise rotation of the frontal plane QRS loop. Varying degrees of RV and right atrial (RA) enlargement or hypertrophy may occur with each type of defect, depending on the presence and degree of pulmonary hypertension. Chest x-ray shows an enlarged RA and RV, and pulmonary artery and its branches; increased pulmonary vascular markings of left-to-right shunt vascularity will diminish if pulmonary vascular disease develops.

Echocardiogram Echocardiography reveals pulmonary arterial and RV and RA dilatation with abnormal (paradoxical) ventricular septal motion in the presence of a significant right heart volume overload. The ASD may be visualized directly by two-dimensional imaging, color-flow imaging, or echocontrast. Echocardiography and Doppler examination have supplanted cardiac catheterization. Transesophageal echocardiography is indicated if the transthoracic echocardiogram is ambiguous, which is often the case with sinus venosus defects, or for guiding catheter device closure (Fig. 282-2). Cardiac catheterization is performed if inconsistencies exist in the clinical data, if significant pulmonary hypertension or associated malformations are suspected, if CAD is a possibility, or when attempting transcatheter closure of the ASD.

TREATMENT ATRIAL SEPTAL DEFECT

Operative repair, usually with a patch of pericardium or of prosthetic material or percutaneous transcatheter device closure, if the ASD is of an appropriate size and shape, should be advised for all patients with uncomplicated secundum ASD with significant left-to-right shunting, i.e., pulmonary-to-systemic flow ratios ≥1.5:1. Excellent results may be anticipated, at low risk, even in patients >40 years, in the absence of severe pulmonary hypertension. In ostium primum ASD, cleft mitral valves may require repair in addition to patch closure of the ASD. Closure is not usually carried out in patients with small defects and trivial left-to-right shunts or in those with severe pulmonary vascular disease without a significant left-to-right shunt. However, the use of pulmonary vasodilators with resultant reduction in pulmonary artery pressure and resistance may allow closure of ASD in patients with pulmonary vascular disease.

Patients with sinus venosus or ostium secundum ASDs rarely die before the fifth decade. During the fifth and sixth decades, the incidence of progressive symptoms, often leading to severe disability, increases substantially. Medical management should include prompt treatment of respiratory tract infections; antiarrhythmic medications for atrial fibrillation or supraventricular tachycardia; and the usual measures for hypertension, coronary disease, or heart failure (Chap. 279), if these complications occur. The risk of infective endocarditis is low, and antibiotic prophylaxis is not recommended (Chap. 155).