specialty centers that are trained and equipped to manage this challenging population. Ongoing efforts to increase awareness, resources, and advocacy are essential for the necessary growth of this specialty.

CARDIAC DEVELOPMENT

(See also Chap. 265e) CHD is generally the result of aberrant embryonic development of a normal structure or failure of such a structure to progress beyond an early stage of embryonic or fetal development. This brief section serves to introduce the reader to normal development so that defects may be better understood; by necessity, it is not exhaustive. Cardiogenesis is a finely tuned process with transcriptional control of a complex group of regulatory proteins that activate or inhibit their gene targets in a location- and time-dependent manner. At about 3 weeks of embryonic development, two cardiac cords form and become canalized; at that point, the primordial cardiac tube develops from two sources (cardiac crescent or the first heart field, pharyngeal mesoderm or the second heart field); by 21 days, these fuse into a single cardiac tube beginning at the cranial end. The cardiac tube then elongates and develops discrete constrictions with the following segments from caudal to cranial location: sinus venosus receives the umbilical, vitelline, and common cardinal veins: atrium, ventricle, bulbus cordis, truncus arteriosus, aortic sac, and the aortic arches. The cardiac tube is fixed at the sinus venosus and arterial ends.

Subsequently, in the next few weeks, differential growth of cells causes the tube to elongate and loop as an "S" with the bulboventricular portion moving rightward and the atrium and sinus venosus moving posterior to the ventricle. The primitive atrium and ventricle communicate via the atrioventricular canal from which the endocardial cushion develops into two parts (ventrally and dorsally). The cushions fuse and divide the atrioventricular canal into two atrioventricular inlets and also migrate to help form the ventricular septum. The primitive atrium is divided first by a septum primum membrane, which grows down from the superior wall to the cushions; as this fusion occurs, the mid-portion resorbs in the center forming the ostium secundum. Rightward of the septum primum, a second septum secundum membrane grows down from the ventral-cranial wall toward—but not reaching—the cushions, and covering most, but not all, of the ostium secundum, resulting in a flap of the foramen ovale. The primitive ventricle is partitioned by a finely tuned set of events. The interventricular septum grows up toward the cushions, and the cushions form an upper inlet septum; between the two portions is a hole called the interventricular foramen. The left and right ventricles begin to develop side by side, and the atria and their respective inlet valves align over their ventricles. Finally, these two parts of the septum fuse with the bulboventricular ridges, which, once having septated the truncus arteriosus, extend into the ventricle. The bulbocordis divides into a subaortic portion as the muscular conus resorbs, whereas the subpulmonary section has elongation of its muscular conus. Spiral division of the common truncus arteriosus rotates and aligns the pulmonary artery and aortic portions over their respective outflow tracts, the aortic valve moving posterior over the left ventricle (LV) outflow tract and the pulmonary valve moving anterior over the right ventricle (RV) outflow tract, with a wraparound relationship of the two great arteries.

Early on, the venous systems are bilateral and symmetric and enter two horns of the sinus venosus. Ultimately, except for the coronary sinus, most of the left-sided portions and the left sinus–venosus horn regress, and the systemic venous system empties into the right horn via the inferior and superior vena cavae. The pulmonary venous system, initially connecting to the systemic venous system, develops as buds from the developing lungs, which fuse together in the pulmonary venous confluence, at which point the connection to the systemic system regresses. Simultaneously, a projection from the back wall of the left atrium (the common pulmonary vein) grows posteriorly to merge with the confluence, which then becomes a part of the posterior left atrial wall.

The truncus arteriosus and aortic sac initially develop six paired symmetric arches, which curve posteriorly and become the paired dorsal aortae. The detailed description of the selective regression of some of the arches is not presented in this chapter. In brief summary, this process results in the development of arch 3 as the internal carotid arteries, left arch 4 as the aortic arch and right subclavian artery, and part of arch 6 as the patent ductus arteriosus. The two dorsal thoracic aortae fuse in the abdomen with persistence of the left dorsal aorta.

TABLE 282-1 SIMPLE ADULT CONGENITAL HEART DISEASE

Native disease
Uncomplicated congenital aortic valve disease
Mild congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)
Uncomplicated small atrial septal defect
Uncomplicated small ventricular septal defect
Mild pulmonic stenosis
Repaired conditions
Previously ligated or occluded ductus arteriosus
Repaired secundum or sinus venosus atrial septal defect without residua
Repaired ventricular septal defect without residua

SPECIFIC CARDIAC DEFECTS

Tables 282-1, 282-2, and 282-3 list CHD malformations as simple, intermediate, or complex. Simple defects generally are single lesions with a shunt or a valvular malformation. Intermediate defects may have two or more simple defects. Complex defects generally have components of an intermediate defect plus more complex cardiac and vascular anatomy, often with cyanosis, and frequently with transposition complexes. The goal of these tables is to suggest when cardiology consultation or advanced CHD specialty care is needed. Patients with complex CHD (which includes most "named" surgeries that usually involve complex CHD) should virtually always be managed in conjunction with an experienced specialty adult CHD center. Patients with intermediate lesions should have an initial consultation and subsequent occasional intermittent follow-up with an adult CHD specialist. Patients with simple lesions often may be managed by a well-informed internist or general cardiologist, although consultation with a specifically trained adult congenital cardiologist is occasionally advisable.

ATRIAL SEPTAL DEFECT

Atrial septal defect (ASD) is a common cardiac anomaly that may be first encountered in the adult and occurs more frequently in females. Sinus venosus ASD occurs high in the atrial septum near the entry of the superior vena cava into the right atrium and is associated frequently with anomalous pulmonary venous connection from the right lung to the superior vena cava or right atrium (Fig. 282-1). Ostium primum ASDs lie adjacent to the atrioventricular valves, either of which may be deformed and regurgitant. Ostium primum ASDs are common in Down's syndrome, often as part of complex atrioventricular septal defects with a common atrioventricular valve and a posterior defect of the basal portion of the interventricular septum. The most common ostium secundum ASD involves the fossa ovalis and is midseptal in location; this should not be confused with a patent foramen

TABLE 282-2 INTERMEDIATE COMPLEXITY CONGENITAL HEART DISEASE

Ostium primum or sinus venosus atrial septal defect
Anomalous pulmonary venous drainage, partial or total
Atrioventricular canal defects (partial or complete)
Ventricular septal defect, complicated (e.g., absent or abnormal valves or with associated obstructive lesions, aortic regurgitation)
Coarctation of the aorta
Pulmonic valve stenosis (moderate to severe)
Infundibular right ventricular outflow obstruction of significance
Pulmonary valve regurgitation (moderate to severe)
Patent ductus arteriosus (nonclosed)—moderate to large
Sinus of Valsalva fistula/aneurysm
Subvalvular or supravalvular aortic stenosis