6

Congenital Heart Disease

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INTRODUCTION

Congenital heart defects are the most common group of birth defects, occurring in approximately 9 of 1000 live births. Without treatment, most patients die in infancy or childhood, with only 5% to 15% surviving into adulthood. Advancements in surgical and medical practices have resulted in survival of approximately 90% of these children to adulthood. For the first time in history, estimates suggest that more adults than children are living with congenital heart disease in the United States and that there is a 5% increase every year.

Most adults living with congenital heart disease have had interventions performed (Table 6-1). Although most children who undergo surgical intervention survive to adulthood, total correction usually is not the rule. Adult patients with congenital heart disease are surviving longer than ever before, and it is becoming apparent that even the simplest lesions can be associated with long-term cardiac complications (i.e., arrhythmias and conduction abnormalities, ventricular dysfunction, residual shunts, valvular lesions, hypertension, and aneurysms) and noncardiac complications (i.e., renal dysfunction, restrictive lung disease, anxiety, depression, and liver dysfunction). Most adults with congenital heart disease need lifelong follow-up.

ACYANOTIC HEART DISEASE

Atrial Septal Defects

Definition and Epidemiology

Atrial septal defects (ASDs) are communications between the atria that allow shunting of blood from one atrium to the other. They are among the most common congenital anomalies seen in adolescents and young adults, occurring in 1 of 1500 live births and constituting 6% to 10% of all congenital heart defects.

There are four main types of ASDs. Ostium secundum defects are the most common, accounting for 75% of all ASDs. This defect occurs in the region of the fossa ovalis and results from excessive absorption of the septum primum or insufficient development of the septum secundum, or both.

Ostium primum defects represent about 20% of all ASDs and represent a form of atrioventricular septal defect (i.e., partial or incomplete atrioventricular canal). These defects are located in the inferior aspect of the atrial septum adjacent to the mitral and tricuspid valves. The defects result from lack of closure of the ostium primum by the endocardial cushions, which are embryologic swellings in the heart that form the primum atrial septum, the inlet portion of the ventricular septum, and parts of the mitral and tricuspid valve. The lesions often are associated with clefts in the mitral and tricuspid valves.

Sinus venosus ASDs represent 5% of all ASDs and are located at the entry of the superior vena cava into the right atrium. Frequently, there is associated partial anomalous drainage of the right upper pulmonary vein. This defect results from resorption of the wall between the vena cava and pulmonary veins.

An unroofed coronary sinus is a rare form of ASD, representing less than 1% of all ASDs. The coronary sinus is in apposition to the posterior aspect of the left atrium, but the orifice is in the right atrium. When a defect exists in the roof of the coronary sinus, a communication between the left atrium and right atrium exists, allowing shunting.

TABLE 6-1 MOST COMMON CONGENITAL HEART DEFECTS SURVIVING TO ADULTHOOD WITHOUT SURGERY OR INTERVENTIONAL CATHETERIZATION

Mild pulmonary valve stenosis
Bicuspid aortic valve
Small to moderate size atrial septal defect
Small ventricular septal defect
Small patent ductus arteriosus
Mitral valve prolapse
Partial atrioventricular canal (ostium primum atrial septal defect and cleft mitral valve)
Marfan syndrome
Ebstein's anomaly
Congenitally corrected transposition (atrioventricular and ventriculoarterial discordance)

Pathology

All four types of ASDs allow oxygenated blood to pass from the left atrium into the right atrium, resulting in volume overload of the right atrium and right ventricle (Fig. 6-1). The degree of shunting is determined by the size of the ASD and the compliance of the left and right cardiac chambers. Comorbidities that increase left-sided filling pressures (i.e., left ventricular [LV] diastolic dysfunction, myocardial infarction, and mitral stenosis) may result in an increased left-to-right shunt. Over time, significant left-to-right shunting can cause enlargement of the right atrium and right ventricle, eventually leading to right ventricular (RV) systolic dysfunction and failure. Pulmonary hypertension may occur in approximately 26% of patients with a secundum ASD. However, significant elevation in pulmonary vascular resistance is rare.