During the 1980s and 1990s, there was a determined effort to “correct” congenital heart disease during infancy, usually within the first few days or weeks of life, in an attempt to prevent secondary functional and anatomic changes in the heart and other organs and to avoid the stress of repeated hospitalization and surgery in older children and adults. Despite this, a significant proportion of patients with uncorrected congenital heart disease grow to adulthood because of the relatively “benign” nature of the condition or because they were not offered or they refused surgical treatment. In addition, a number of patients who have undergone palliative or supposedly corrective operations require further surgical treatment in adulthood. The increasing number and the specific nature of these operations have created a new subspecialty of surgery for congenital heart disease. This chapter considers some of the general features and specific conditions of congenital heart surgery in the adult.

General Considerations

Congenital heart defects produce progressive changes in cardiac form and function, as well as secondary effects resulting from chronic systolic or diastolic overload that can involve myocardial cells, the connective tissue framework of the heart, or the microvasculature and endocardium. These abnormalities are qualitatively different for systolic and diastolic overload. Although many of these changes initially have the potential to be reversed, they may become irreversible if left uncorrected. Other cardiac effects may involve progression or development of new obstructive lesions, such as that observed in tetralogy of Fallot, ventricular septal defect (VSD), or aortic outflow obstruction in double-inlet ventricle with malposition of the great arteries. Continued blood turbulence may affect the structure and function of the atrioventricular (AV) or semilunar valves, with thickening or calcification of the cusps or dilatation of the annulus. Poststenotic changes in the great arteries can lead to either dilatation or arterial wall changes as the result of turbulence or failure of development and growth due to chronic reduction in flow. The pulmonary vascular changes caused by an increase in flow and pressure include medial hypertrophy and reactivity of smooth muscle cells, functional and structural intimal changes, and eventual development of irreversible plexogenic lesions. The rate at which such changes occur varies among different conditions and among individual patients. Other changes that may occur are the direct result of previous surgery, which can produce distortion of vessels on intracardiac shunting. Cerebral, hepatic, and renal function may be affected by hypoperfusion, cyanosis, or embolism. Accurate characterization of the extent and reversibility of secondary changes is of special importance in planning the time and type of surgical intervention in adults with congenital heart disease.

Specific Congenital Defects in the Adult

Aortic Valve Stenosis

Congenital bicuspid aortic valve commonly results in progressive stenosis due to thickening and calcification of the cusps [Fig. 13.1]. Accurate measurement of the amount and localization of calcification in the aortic valve can be made using electron beam computed tomography. The amount of calcium in the valve has been shown to be a predictor of progression of the disease as well as an independent prognostic indicator. Valve-conserving surgery is rarely possible in adults, and therefore valve replacement is the most commonly performed surgical procedure. Advances in myocardial protection and other intraoperative techniques have led to a reduction in the perioperative mortality rates for both initial and successive surgery to approximately 1% to 3%. The most important issue is the choice of a valve substitute, which should match the characteristics of the valve to the requirements of the patient.

Although a considerable amount of information is available about the performance and suitability of the different
valve substitutes, this information is often incomplete, and other factors, such as the experience, predictions, and bias of the surgical team, cardiologist, and patient, can influence the choice. There is increasing evidence that the normal aortic valve performs a series of extremely sophisticated functions that depend on the biologic properties of their components. The use of a living valve substitute capable of reproducing most or all of these functions can result in clinical benefit in terms of both survival and quality of life. The only current valve procedures capable of preserving aortic valve viability in the long term are valve repair and the Ross operation. Recent evidence strongly suggests that both survival and exercise capacity are enhanced after the Ross operation when compared to other valve substitutes. Recent progress in tissue engineering holds the promise of making available a fully viable human valve in the not very distant future. The available substitutes include prosthetic valves and stented or unstented xenografts. Prosthetic valves are the most commonly used valve substitutes and have the advantages of ease of insertion and superior durability. However, the need for continuous postoperative anticoagulation can lead to a small but significant morbidity and mortality incidence, particularly in women during the childbearing years. Stented xenografts offer the advantage of predictability, ease of insertion, and low incidence of thromboembolic complications. However, their durability is limited, particularly in patients younger than 35 years. In addition, the stent can become obstructive (Fig. 13.2), which has negative implications for hemodynamic performance and possibly for long-term left ventricular (LV) function. Homograft valves are inserted in the subcoronary position either via a two-suture-line technique or as a root replacement (Fig. 13.3) with reimplantation of the coronary arteries. These valves have excellent hemodynamic performance (Fig. 13.5), are virtually free of thromboembolic complications, and have greater durability than xenografts, particularly in children. However, their insertion is somewhat technically demanding, they are less readily available,