CONGENITALLY CORRECTED TRANSPOSITION

The two fundamental anatomic abnormalities in this malformation are transposition of the ascending aorta and pulmonary trunk and inversion of the ventricles. This arrangement results in desaturated systemic venous blood passing from the RA through the mitral valve to the LV and into the pulmonary trunk, whereas oxygenated pulmonary venous blood flows from the left atrium through the tricuspid valve to the RV and into the aorta. Thus, the circulation is corrected functionally. The clinical presentation, course, and prognosis of patients with congenitally corrected transposition vary depending on the nature and severity of any complicating intracardiac anomalies and of development of dysfunction of the systemic subaortic RV. Progressive RV dysfunction and tricuspid regurgitation may also develop in one-third of patients by age 30; Ebstein-type anomalies of the left-side tricuspid atrioventricular valve are common. VSD or PS due to obstruction to outflow from the right-sided subpulmonary (anatomic left) ventricle may coexist. Complete heart block occurs at a rate of 2–10% per decade. The diagnosis of the malformation and associated lesions can be established by comprehensive two-dimensional echocardiography and Doppler examination.

MALPOSITIONS OF THE HEART

Positional anomalies refer to conditions in which the cardiac apex is in the right side of the chest (*dextrocardia*) or at the midline (*mesocardia*), or in which there is a normal location of the heart in the left side of the chest but abnormal position of the viscera (*isolated levocardia*). Knowledge of the position of the abdominal organs and of the branching pattern of the main stem bronchi is important in categorizing these malpositions. When dextrocardia occurs without situs inversus, when the visceral situs is indeterminate, or if isolated levocardia is present, associated, often complex, multiple cardiac anomalies are usually present. In contrast, mirror-image dextrocardia is usually observed with complete situs inversus, which occurs most frequently in individuals whose hearts are otherwise normal.

SURGICALLY MODIFIED CONGENITAL HEART DISEASE

Owing to the enormous strides in cardiovascular surgical techniques that have occurred in the past 70 years, a large number of long-term survivors of palliative or corrective operations in infancy and childhood have reached adulthood. These patients are often challenging because of the diversity of anatomic, hemodynamic, and electrophysiologic residua and sequelae of cardiac operations.

The proper care of the survivor of an operation for CHD requires that the clinician understand the details of the malformation before operation; pay meticulous attention to the details of the operative procedure; and recognize the postoperative residua (conditions left totally or partially uncorrected), the sequelae (conditions caused by surgery), and the complications that may have resulted from the operation. Except for ligation of an uncomplicated patent ductus arteriosus, almost every other surgical repair leaves behind or causes some abnormality of the heart and circulation that may range from trivial to serious. Thus, even with results that are considered clinically to be good to excellent, continued long-term postoperative follow-up is advisable.

Cardiac operations involving the atria, such as closure of ASD, repair of total or partial anomalous pulmonary venous return, or venous switch corrections of complete transposition of the great arteries (the Mustard or Senning operations), may be followed years later by sinus node or atrioventricular node dysfunction and/or by atrial arrhythmias (especially atrial flutter). Intraventricular surgery may also result in electrophysiologic consequences, including complete heart block necessitating pacemaker insertion to avoid sudden death. Valvular problems may arise late after initial cardiac operation. An example is the progressive stenosis of an initially nonobstructive bicuspid aortic valve in the patient who underwent aortic coarctation repair. Such aortic valves may also be the site of infective endocarditis. After repair of the ostium primum ASD, the cleft mitral valve may become progressively regurgitant. Tricuspid regurgitation may also be progressive in the postoperative patient with tetralogy of Fallot if RV outflow tract obstruction was not relieved adequately at initial surgery. In many patients with surgically modified CHD, inadequate relief of an obstructive lesion, a residual regurgitant lesion, or a residual shunt will cause or hasten the onset of clinical signs and symptoms of myocardial dysfunction. Despite a good hemodynamic repair, many patients with a subaortic RV develop RV decompensation and signs of left heart failure. In many patients, particularly those who were cyanotic for many years before operation, a preexisting compromise in ventricular performance is due to the original underlying malformation.

A final category of postoperative problems involves the use of prosthetic valves, patches, or conduits in the operative repair. The special risks include infective endocarditis, thrombus formation, and premature degeneration and calcification of the prosthetic materials. There are many patients in whom extracardiac conduits are required to correct the circulation functionally and often to carry blood to the lungs from the RA or RV. These conduits may develop intraluminal obstruction, and if they include a prosthetic valve, it may show progressive calcification and thickening. Many such patients face reintervention (interventional cardiac catheterization or surgical reoperation) one or more times in their lives. Such care should be directed to centers specializing in adults with complex congenital cardiovascular malformations. The effect of pregnancy in postoperative patients depends on the outcome of the repair, including the presence and severity of residua, sequelae, or complications. Contraception is an important topic with such patients. Tubal ligation should be considered in those in whom pregnancy is strictly contraindicated.

Endocarditis Prophylaxis Two major predisposing causes of infective endocarditis are a susceptible cardiovascular substrate and a source of bacteremia. The clinical and bacteriologic profile of infective endocarditis in patients with CHD has changed with the advent of intracardiac surgery and of prosthetic devices. Prophylaxis includes both antimicrobial and hygienic measures. Meticulous dental and skin care are required. Routine antimicrobial prophylaxis is recommended for bacteremic dental procedures or instrumentation through an infected site in most patients with operated CHD, particularly if foreign material, such as a prosthetic valve, conduit, or surgically constructed shunt, is in place. In the case of patches, in the absence of a high-pressure patch leak, or transcatheter devices, prophylaxis is usually recommended for 6 months until there is endothelialization. Individuals with unrepaired cyanotic heart disease are also generally recommended to receive prophylaxis (Chap. 155).

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GLOBAL BURDEN OF VALVULAR HEART DISEASE

Primary valvular heart disease ranks well below coronary heart disease, stroke, hypertension, obesity, and diabetes as a major threat to the public health. Nevertheless, it is the source of significant morbidity and mortality rates. Rheumatic fever (Chap. 381) is the dominant cause of valvular heart disease in developing and low-income countries. Its prevalence has been estimated to range from as low as 1 per 100,000 school-age children in Costa Rica to as high as 150 per 100,000 in China. Rheumatic heart disease accounts for 12–65% of hospital admissions related to cardiovascular disease and 2–10% of hospital discharges in some developing countries. Prevalence and mortality rates vary among communities even within the same country as a function of overcrowding and the availability of medical resources and population-wide programs for detection and treatment of group A streptococcal pharyngitis. In economically deprived areas, tropical and subtropical climates (particularly on the Indian subcontinent), Central