Malfunction of any of the cardiac valves results in a less efficient circulatory system. Valvular dysfunction causes work overload in one or both ventricles. In extreme cases, resultant congestive heart failure can cause death. More information about etiology, pathogenesis, differential diagnoses, and diagnostic approaches used for evaluation of valvular diseases can be found in chapters 27–33.

BACKGROUND

Before the discovery of penicillin, rheumatic heart disease was commonplace. Physicians recognized that mitral valve stenosis frequently followed rheumatic fever. This obstruction to blood flow through the mitral valve was not medically treatable however. For “stenosed” mitral valves, physicians described the need to relieve the obstruction surgically. The first successful attempt at surgical treatment involved incising the left atrial appendage, placing a finger through the incision into the left atrium, feeling the stenotic mitral valve, and relieving the obstruction by simple finger pressure. Soon after these initial therapeutic approaches, special knives and dilators were developed to relieve mitral valve stenosis. In the early days of cardiovascular surgery, these procedures were all performed on the beating heart.

The notion of using anticoagulant heparin to allow blood to circulate outside a patient’s vasculature without clotting led to the development of cardiac and pulmonary bypass machines in the 1950s. It was then possible to keep the patient alive while stopping the heart for surgical repair. The ability to stop the heart, examine valve pathology, and try to repair it stimulated surgeons’ collaboration with mechanical engineers in developing prosthetic valves to replace those that were too diseased to repair. Initial attempts to duplicate valve leaflets with flexible, nonbiologic materials failed. The leaflets of these valves were too stiff in comparison with normal valve leaflets.

FIRST-GENERATION PROSTHETIC VALVES

Attempts at using nonflexible leaflets by constructing hinged valve leaflets resulted in hinge thrombosis and malfunction. Design engineers then focused on free-floating occluders, such as discs or balls retained in a cage-like housing. This general valve design produced the first clinically useful valves, including the Hufnagel, Starr-Edwards, Smeloff-Cutter, and Beall valves (Fig. 34-1). In 1958, the Starr-Edwards valve was used in the first clinically successful valve replacement.

Although these early designs functioned as intended, the first caged-ball valves had major shortcomings: (1) they were bulky in design and did not fit well into a small ventricle or aorta; (2) they had a small internal orifice, making them relatively stenotic; and (3) they stimulated thrombus formation, which precipitated thromboembolic events, necessitating long-term anticoagulation therapy.
Figure 34-1  First Generation of Synthetic Prosthetic Valves

The first generation of clinically useful synthetic valves had a free-floating ball or disc occluder retained in a cage-like house.

- **Hufnagel valve**
- **Starr-Edwards valve** settled (mitral valve position)
- **Smeloff-Cutter valve**
- **Beall valve**
- Disc elevated by very slight pressure to demonstrate closure
- Movable ball (up and down)
- Movable disc (discoid)
SECOND-GENERATION PROSTHETIC VALVES

The disadvantages of early prosthetic valves led to the development of two divergent lines of valve design using synthetic materials or biological tissue. The caged-ball valves were modified, and pivoting hingeless disc valves, such as the Lillehei-Kaster, Medtronic-Hall, and Björk-Shiley valves, were developed. The St. Jude and Carbomedic valves were the first successful hinged leaflet valves (Fig. 34-2).

Homograft valves harvested at autopsy and preserved in antibiotic solution or frozen were the first nonsynthetic valves to be implanted successfully. Their limited availability prompted the use of porcine valves procured from slaughterhouses. Porcine valves were preserved with glutaraldehyde and mounted on a modified nylon-covered plastic or metal stents. Valves made of pericardium were also developed and used successfully (Fig. 34-3, lower). Many of these valve designs are still in use today.

ETIOLOGY AND PATHOGENESIS

Cardiac valve pathology comprises two broad categories, congenital valve deformity and acquired valvular dysfunction. Congenital deformity can occur in one or more cardiac valves (see section VIII). Patients with severe congenital valvular dysfunction can die if prompt surgical intervention is not undertaken. In patients with normally developed hearts, infection can cause valvular dysfunction at any age. Rheumatic heart disease secondary to untreated streptococcal infection and bacterial endocarditis can destroy a normal heart valve. Generalized inflammatory illnesses, such as lupus erythematosus, rheumatoid arthritis, and eosinophilic endocarditis, as well as carcinoid disease, similarly can cause valvular dysfunction. Connective tissue diseases, such as Ehlers-Danlos syndrome and myxomatous degeneration, can cause valve deformity and dysfunction. Severe myocardial ischemia and injury can cause papillary muscle dysfunction, which can result in mitral valve insufficiency. Finally, just as aging often results in atherosclerotic changes and calcium deposition in arterial walls, so can it affect the aortic valves, sometimes with severe calcification of the leaflets.

The mitral valve annulus can also be severely calcified, with or without valvular dysfunction.

CLINICAL PRESENTATION

The presenting symptoms in patients with dysfunctional valves vary considerably, depending on the type and severity of dysfunction and the location of the affected valves. Diseased valves can become incompetent, stenotic, or both. Young patients with moderate aortic valve stenosis are often asymptomatic. Likewise, many patients with moderate mitral valve stenosis or insufficiency may be asymptomatic. In general, patients whose valve dysfunction progresses eventually experience dyspnea on exertion. Syncope or angina pectoris, alone or in association with dyspnea, can develop in patients with aortic stenosis.

DIFFERENTIAL DIAGNOSIS

In patients presenting with dyspnea and fatigue, noncardiac causes, such as anemia, hypertension, pulmonary pathology, and hypothyroidism, should be excluded. Primary cardiac myopathy (see chapters 12–16) should be considered. Coronary artery disease must be ruled out if angina pectoris is a symptom.

DIAGNOSTIC APPROACH

Physical findings such as cardiac murmurs, wide pulse pressure, cardiomegaly, hepatomegaly, ascites, or pedal edema help to confirm abnormal circulatory conditions. Chest radiography and ECG offer supportive evidence of cardiac pathology. The most descriptive and definitive tests pinpointing cardiac valve anomalies are echocardiography in association with hemodynamic data from cardiac catheterization.

MANAGEMENT AND THERAPY

Surgical Therapy

A variety of procedures are available to treat cardiac valvular disease. Replacing diseased valves with prosthetic valves has become a routine procedure and valve repair—particularly mitral and tricuspid valve repair—has evolved dramatically. Techniques routinely used in the repair of mitral and tricuspid insufficiencies include ring annuloplasty, resection of prolapsing portions of leaflets not supported by chor-
Figure 34-2  Second Generation of Synthetic Prosthetic Valves and Biologic Valves

Second-generation synthetic prosthetic valves were hingeless pivoting disk valves and hinged bileaflet valves.

Tissue valves made of porcine aortic valves, pericardium, or cadaver homografts are also important in valve replacement surgical therapy.
Figure 34-3

Chordal Transfer, Sliding Annuloplasty, and Ring Annuloplasty

Torn anterior leaflet chordae

Ascending aorta wall

Anterior leaflet

Posterior leaflet

Portion of the posterior leaflet, with chordae transferred to repair the anterior leaflet (Carpentier quadrangular technique)

Left atrium

Chordae tendineae

Papillary muscles

In these views, the medial aspect of the mitral valve, chordae tendineae, and papillary muscles apparatus has been removed.

Left ventricle

Anterior leaflet reconstructed

Insertion of a flexible annuloplasty ring may be indicated to reestablish the coaptation and strengthen the annular support.

Cut for sliding annuloplasty technique

In these views, the valvar apparatus is shown complete with chordae tendineae and papillary muscles.
Surgical Treatment for Valvular Heart Disease

dae, shortening or using artificial chordae, and increasing or decreasing the leaflet area by sliding annuloplasty (Fig. 34-4). In patients who need aortic valve replacement, some surgeons advocate the Ross procedure, which entails transplanting a patient’s pulmonary valve into the aortic position. This provides the patient with a living, durable, nonthrombogenic, and hemodynamically superior valve. The pulmonary valve is then reconstructed using a tissue homograft valve. The choice of procedure depends on many factors, including the patient’s valve pathology, age, and ability to tolerate and comply with long-term anticoagulation.

Mitral and Tricuspid Valves

Patients with mitral and tricuspid valve pathology should be considered for valve repair rather than replacement because the operative mortality associated with repair of these valves is lower than that associated with their replacement. After surgery for either valve repair or replacement, patients need to receive anticoagulants for 3 to 6 months until the surgical site is endothelialized. Patients with repaired valves or valves replaced with biologic tissue can then discontinue anticoagulation if they remain in sinus rhythm. The long-term incidence of thromboembolic events is generally lower in patients with repaired valves in comparison with patients with replaced valves. This is one of the reasons that valve repair is preferable to valve replacement, when the repair is technically feasible.

Conditions precluding satisfactory repair of the mitral and tricuspid valves include severe scarring and deformation by a disease process such as advanced rheumatic heart disease, advanced lupus, or another inflammatory process involving the valve leaflets and destruction of valve leaflets and annuli by endocarditis. Under these circumstances, the valve should be replaced. Mitral valve replacement should include preservation of a portion of the subvalvular chordae and papillary muscles to aid in preserving normal ventricular contractility.

Aortic Valves

Adult patients with aortic valve pathology are seldom candidates for valve repair; valve replacement is usually necessary for significant aortic stenosis or regurgitation. The patient’s age, the patient’s lifestyle, and the preferences of the surgeon and the patient dictate the type of prosthetic valve replacement.

Patients with prosthetic valves made of biologic tissue have a lower incidence of bleeding because long-term anticoagulation is not required in patients in sinus rhythm. Unfortunately, all tissue valves eventually deteriorate and become insufficient. Deterioration of tissue valves occurs at an accelerated rate in younger patients and in patients with end-stage renal disease on hemodialysis. For older patients, particularly those with a risk of falling, a tissue valve may be the most appropriate choice. Younger patients, with a natural life expectancy exceeding 15 to 20 years, should have prosthetic valves made of durable synthetic materials, such as pyrolytic carbon, titanium, stainless steel, or a combination of these.

Postoperatively, all patients with prosthetic heart valves must be anticoagulated until endothelialization of the sewing ring is complete, as discussed previously herein. Use of non-tissue valves necessitates indefinite anticoagulation.

Issues With Prosthetic Valve Replacement

Nontissue valves must have an appropriate sewing ring sutured to the annulus of the patient’s valve after the leaflets are excised. Sewing rings are usually circular and rigid and vary in thickness. The rigid sewing rings change the natural shape of the valve annulus and, depending on thickness, decrease the size of the internal orifice of the prosthetic valve. Implanting a valve with a circular sewing ring into a noncircular annulus can generate unnatural tension between the valve annulus and sewing ring, which can lead to paravalvular leaks; the surgical approach in these instances must take this possibility into account.

The use of rigid circular sewing rings is unnecessary in biologic tissue valves implanted in the aortic position. Freehand suturing is used to insert autograft pulmonary valves into the aortic position (the Ross procedure). It is also used in homograft cadaver valve implantation and with nonstented freestyle porcine valves.
Exposing the mitral valve through the interatrial septum and an extension of the incision through the roof of the left atrium is common. This surgical exposure allows excellent visualization of the mitral and tricuspid valves and can be performed through a standard sternotomy, as well as through a variety of partial sternotomy and right thoracotomy incisions.
Minimally Invasive Techniques

Minimally invasive coronary artery revascularization surgery uses small incisions and therefore is performed on a beating heart, obviating the use of cardiopulmonary bypass (CPB). In valve repair and replacement procedures, the use of smaller incisions is possible, but eliminating CPB is not feasible with current techniques and prosthetic valves.

Good visualization of the operative field is a prerequisite for proper valve repair or replacement. Smaller incisions limit visualization, although the use of miniature video cameras improves the view of the operative field. The mitral valve is generally the most difficult to visualize, so many surgeons approach it through the intra-atrial septum, sometimes extending the incision to the roof of the left atrium (see Fig. 34-4).

FUTURE DIRECTIONS

Refinements in manufacturing synthetic prosthetic valves and their sewing rings will continue to decrease thromboembolic complications while improving their hemodynamic characteristics. Better chemical preservation of tissue valves will improve their longevity and resistance to deterioration and make tissue valves a more attractive choice for younger patients.

The teaching of valve repair techniques to surgical trainees is already becoming more standardized. The appropriate surgical repair technique will be more predictable from the preoperative, noninvasive echocardiographic examination and hemodynamic evaluation. Freehand valve implantation techniques will find increased use in selected patients, particularly for patients in whom the annulus is small and the valve sewing rings make the prosthetic valves too stenotic. Finally, with clinical acceptance of genetic engineering, farms of genetically altered pigs and baboons might provide viable biologic leaflets, valves, and entire hearts for implantation.

REFERENCES
