Perspective

The natural history of infective endocarditis (IE) has undergone considerable changes in the antibiotic era. The older classifications of acute, subacute, and chronic have become less meaningful; immunologic phenomena, such as Osler’s nodes and glomerulonephritis, are rarely seen; and nosocomial infections from indwelling catheters and devices are increasing. Early diagnosis and treatment play a significant role in the clinical outcomes because IE remains a disease that causes considerable morbidity and mortality.

Principles of Disease

Estimates of the incidence of IE in the United States vary widely, in part because of changing case definitions throughout the years but also because of differences in predisposing conditions among studied populations. For example, the incidence of IE among patients admitted to hospitals in Philadelphia was estimated to be 11.6 cases per 100,000 person-years from 1988 to 1990. In contrast, the incidence of endocarditis in Olmsted County, Minnesota, during the period from 1970 to 2000 was 5.0 to 7.0 cases per 100,000 person-years. Others report significantly lower rates. IE is increasingly a disease of the aged, with more than 50% of cases occurring in individuals older than 60 years. This reflects the pervasiveness of degenerative valve disease in the elderly and the increased prevalence of prosthetic heart valves. Nosocomial endocarditis is increasingly common; the source is often infected intravascular devices, pacemakers, and hemodialysis catheters.

Most patients with bacterial endocarditis have a predisposing valvular abnormality. Among elderly patients, calcific or degenerative disease of the aortic and the mitral valve is the most common predisposing factor. Rheumatic heart disease (RHD), although less prevalent than in prior decades, remains an important predisposing factor for IE among individuals from developing countries. Congenital cardiac lesions involving high pressure gradients (e.g., ventricular septal defects, pulmonary stenosis, and tetralogy of Fallot) also increase risk of IE. A history of previous endocarditis is a major risk factor for recurrence because infected valves heal with irregularities that become a nidus for future vegetations. Mitral valve prolapse (MVP) is now the most common predisposing abnormality for IE in developed countries.

The incidence of IE associated with injection drug use is estimated at 150 to 2000 per 100,000 person-years. Although any valve can be affected, injection drug use is classically associated with right-sided endocarditis.

Pathophysiology

The classic lesion of endocarditis is the vegetation, originating as a sterile thrombus on which microorganisms adhere and colonize. The target is most often the cardiac valve; however, chordae tendineae, septal defects, and the endocardium can be involved. The initial thrombus may form at a site of mechanical damage induced by inflammation, degenerative changes, or abnormal turbulence. In injection drug users, contaminants such as talc can injure the previously normal valve leaflets and encourage bacterial implantation. Theoretically, the onset of bacterial endocarditis is preceded by a period of subclinical bacteremia. Dental procedures, cystoscopy, endoscopy, and other invasive procedures result in transient bacteremia, but more commonly there is no clear precipitant for community-acquired IE.

A number of microorganisms cause IE, with staphylococci and streptococci accounting for the majority of cases (Table 83-1). In a study of nearly 2800 patients with IE from 58 medical centers in 25 countries, staphylococci were the causative agents in 31% (commonly associated with injection drug use) and streptococci in 29%. But a community-based study from Minnesota, where referral bias was minimized, showed that viridans group streptococci continued to be the most common culprit organism. Although many organisms cause IE, a few have such a predilection that they count as a major criterion in the modified Duke score. A blood culture positive for community-acquired enterococcus and S. bovis should suggest IE. In addition, the HACEK group (Haemophilus aphrophilus, Actinobacillus actinomycetemcomitans, Cardiobacterium hominis, Eikenella corrodens, and Kingella kingae), a collection of fastidious gram-negative bacilli, is commonly reported in large studies of IE. Lastly, a positive blood culture or positive antibody titer to Coxiella burnetii (Q fever) is also included in the modified Duke score.
The microbiology of PVE relates to the time of onset: *Staphylococcus aureus* is now the most prevalent pathogen in the first 2 months after valve replacement, followed by coagulase-negative *staphylococci*. One year after surgery, the microbiology mirrors native valve endocarditis. Among injection drug users, the most common infecting organism by far is *S. aureus*, often causing right-sided infections. *Candida* and *Aspergillus* species cause most cases of fungal endocarditis. Predisposing factors include indwelling intravenous catheters, immunocompromise, and injection drug use. Large fungal vegetations can embolize, and analysis of these emboli may suggest fungal endocarditis. Bartonella species are another group of fastidious organisms associated with IE in immunocompromised patients.

**Clinical Features**

Symptoms associated with IE are nonspecific and diverse. The most common symptoms are intermittent fever (85%) and malaise (80%). Other nonspecific symptoms (e.g., weakness, myalgias, back pain, dyspnea, chest pain, cough, headaches, and anorexia) vary widely in their incidence. Many patients seen early during the bacteremic phase of the illness do not have a cardiac murmur and are indistinguishable from the large population of patients who come to the emergency department (ED) with a febrile viral illness. During the initial assessment a careful history should be taken, with attention to any preexisting cardiac pathology or clues suggesting a recent source of bacteremia, such as intravenous drug use, indwelling intravascular catheters, or invasive procedures. In the absence of specific risk factors, the diagnosis of IE may be suspected when infectious symptoms persist or do not follow a typical course for viremia. The classic triad of fever, anemia, and heart murmur is rare.

Some patients will have complications of IE as presenting complaints. The most common and severe are congestive heart failure and neurologic events. Heart failure results from valvular destruction. A patient with strokelike symptoms and a fever should alert one to the possibility of IE. Strokes can be the result of septic emboli or ruptured mycotic aneurysms. Almost all patients with IE have a cardiac murmur at some time during the course of their illness. A murmur, however, may be absent at presentation. For example, fewer than 35% of intravenous drug users with endocarditis have a murmur at presentation. In this population, unexplained fever alone is sufficient to raise concern about possible endocarditis. A substantial minority of patients exhibit some form of vasculitic lesion, including petechiae, splinter hemorrhages, Osler’s nodes, and Janeway lesions. Approximately 30% of patients have splenomegaly. Ocular findings include conjunctival or retinal hemorrhages, the latter of which may have a characteristic pale center surrounded by a red halo (Roth’s spots).

**Diagnostic Strategies**

Laboratory findings in bacterial endocarditis are nonspecific. As with other infectious conditions, leukocytosis is insensitive (occurring in approximately 50% of patients diagnosed with IE) and nonspecific. An elevated erythrocyte sedimentation rate or C-reactive protein level may be present, but these are also nonspecific. Most patients have a mild anemia, and up to 50% have microscopic hematuria as a result of embolic lesions of the kidney. A chest radiograph may show signs of heart failure or embolic disease, and an electrocardiogram (ECG) may display conduction abnormalities if an abscess has formed in the myocardium.

Although not always practical, three blood cultures from three separate venipuncture sites are recommended for patients with a presumptive diagnosis of possible endocarditis, with the first and last cultures drawn at least 1 hour apart. Approximately 90 to 95% of blood cultures are positive unless antibiotics have already been administered. If the patient appears septic, cultures may be obtained more rapidly to permit initiation of early empirical therapy. Cultures need not be timed to the presence of chills or fever because patients with IE typically have a continuous fever. An echocardiogram should be performed in all patients for whom suspicion of endocarditis is moderate to high. Although transthoracic echocardiography (TTE) is highly specific for vegetations in IE, it may be nondiagnostic in up to 20% of patients because of obesity, chronic obstructive pulmonary disease, and chest wall deformities. Overall sensitivity of TTE is at most 60%. Transesophageal echocardiography (TEE), on the other hand, although more invasive and time-consuming, is far superior to TTE in its sensitivity and specificity.

Explicit criteria for the diagnosis of IE are important because underdetection can lead to serious morbidity and death, whereas overdetection can result in weeks of unnecessary antimicrobial therapy. The Duke criteria are the most widely accepted, and they stratify patients with suspected bacterial endocarditis into three distinct categories: definite, possible, and rejected (Table 83-1). The sensitivity and specificity of the Duke criteria are approximately 95% and 99%, respectively.

**Management**

Once the diagnosis of IE is established, whether by clinical, echocardiographic, or microbiologic methods, antimicrobial therapy should be administered. Choice of antibiotics depends on the likely (or known) causative organism but is usually empirical. Guidelines for empirical antibiotic therapy in patients with suspected IE are provided in Box 83-2. In the ED, however, usually without results of an echocardiogram (TTE or TEE), the diagnosis of endocarditis is not confirmed. In addition, there is increasing concern regarding community-acquired methicillin-resistant *S. aureus*. Thus a combination of 15 mg/kg of vancomycin and 2 g of ceftriaxone is a reasonable empirical antibiotic choice in someone with undifferentiated sepsis and suspected endocarditis.

Endocarditis is a heterogeneous disease. Although initial treatment is medical, early consultation with a cardiac surgeon is advisable.
**Box 83-1** Duke Criteria (Clinical) for Diagnosis of Infective Endocarditis

**Definite Endocarditis**
Endocarditis is considered definitely present if any one of the following combinations of clinical findings is present:
- Two major clinical criteria
- One major and any three minor clinical criteria
- Five minor clinical criteria

**Possible Endocarditis**
Possible endocarditis is defined as the presence of any one of the following combinations of clinical findings:
- One major and one or two minor clinical criteria
- Three minor clinical criteria

**Rejected Endocarditis**
The diagnosis of endocarditis is considered rejected if any of the following occurs:
- A firm alternate diagnosis is made.
- Resolution of clinical manifestations occurs after 4 days of antibiotic therapy or less.
- Clinical criteria for possible or definite infective endocarditis not met.

**Major Criteria**
Positive blood cultures (of typical pathogens) from at least two separate cultures.
Evidence of endocardial involvement by echocardiography, such as the following:
- Endocardial vegetation
- Paravalvular abscess
- New partial dehiscence of prosthetic valve
- New valvular regurgitation

**Minor Criteria**
*Predisposition*: Predisposing heart condition or intravenous drug use
- Fever: Temperature >38°C
- Vascular phenomena: Arterial emboli, septic pulmonary infarcts, mycotic aneurysm, conjunctival hemorrhages, or Janeway lesions
- Immunologic phenomena: Osler’s nodes, Roth’s spots, and rheumatoid factor
- Microbiologic evidence: Single positive blood culture (except for coagulase-negative Staphylococcus or an organism that does not cause endocarditis)
- Echocardiogram findings: Consistent with endocarditis but do not meet major criteria

**Box 83-2** Initial Empirical Therapy for Bacterial Endocarditis

**Native Valve**
Penicillin G 4 million units IV q4h + nafcillin 2 g IV q4h

Or
Vancomycin 15 mg/kg IV q12h

Plus
Gentamicin 1 mg/kg IV q8h

**Native Valve (+ Injection Drug Use)**
Vancomycin 15 mg/kg IV q12h

**Prosthetic Valve**
Vancomycin 15 mg/kg IV q12h

Plus
Gentamicin 1 mg/kg IV q8h

IV, intravenously.

**Box 83-3** Surgical Therapy for Infective Endocarditis

- Infective endocarditis with acute heart failure
- Fungal endocarditis
- Periannular extension of infection
- Recurrent emboli
- Large mobile vegetations
- Persistent bacteremia

**Box 83-4** High-Risk Conditions for Bacterial Endocarditis

- Prosthetic heart valve
- History of endocarditis
- Unrepaired cyanotic congenital heart disease, including palliative shunts and conduits
- Completely repaired congenital heart defects with prosthesis during the first 6 months after the procedure
- Repaired congenital heart disease with residual defect at or adjacent to the site of the prosthetic device
- Cardiac valvulopathy in a transplanted heart

Advisable when mechanical complications are observed or expected (such as in patients with acute heart failure or those with infections involving prosthetic valves). See Box 83-3. Consultation with an infectious diseases specialist or a cardiologist is also useful.

Febrile patients with suspected IE require admission for definitive diagnosis and initiation of empirical therapy. An exception might be an otherwise well-appearing injection drug user with transient fever that is attributed to an injected contaminant (“cotton fever”).

With appropriate antibiotic therapy, most patients with IE will defervesce within 1 week. The duration of antibiotic therapy must be sufficient to eradicate microorganisms present within the valvular vegetations. This may require 6 weeks, or more, depending on the organism and the type of vegetation. Historically, most patients with IE received the entire course of antimicrobial therapy while in the hospital. The development of home health care, however, allows selected patients with endocarditis to be treated as outpatients during much or all of their therapy. Patients selected for outpatient therapy should be hemodynamically stable, compliant, and capable of managing the technical aspects of intravenous therapy.

**Prophylaxis**
The American Heart Association has updated guidelines, limiting prophylaxis to conditions with the highest risk of adverse outcome from IE (Box 83-4). Virtually all of the procedures that are routinely performed in the ED, including suturing of lacerations, endotracheal intubation, placement of central venous catheters, vaginal deliveries, and placement of Foley catheters (in the absence of infection), do not require prophylactic antibiotics.

**RHEUMATIC FEVER**

**Perspective**
From 1920 to 1950, acute rheumatic fever (ARF) was the leading cause of death in U.S. children and the most common cause of heart disease in individuals younger than age 40 years. During the 1960s and 1970s, the incidence of ARF in the United States and other developed countries declined dramatically because of widespread antibiotic treatment of streptococcal infections, declining
prevalence of the more virulent strains of group A streptococci, and improved living conditions. Children 4 to 9 years of age remain at greatest risk, with an incidence of ARF of 2 to 14 cases per 100,000.25 In many developing nations, however, ARF continues to be a leading cause of childhood mortality. RHD peaks in adults between the ages of 25 and 34 years and continues to be a leading cause of morbidity and mortality in impoverished areas.26

Principles of Disease

ARF is a delayed, nonsuppurative complication of streptococcal pharyngitis. Although the pathogenesis remains obscure, ARF results from an exaggerated immunologic response to group A beta-hemolytic streptococci that results in antibodies cross-reacting with tissues in the heart, joints, skin, and central nervous system. Patients with a history of ARF are predisposed to recurrent infections, and repeated infections lead to progressive heart damage.

Clinical Features

After the initial pharyngitis, there is a latency period ranging from 1 to 5 weeks (average, 18 days) before symptoms and signs of ARF appear. Up to one third of patients with documented ARF do not remember having had pharyngitis in the preceding month. Fever is present during the acute phase of rheumatic fever. It rarely lasts more than 2 weeks and has no characteristic pattern. Along with fever, manifestations of ARF may include arthritis, carditis, chorea, subcutaneous nodules, and erythema marginatum.

Migratory polyarthritis is the most common manifestation of ARF. Arthritis tends to occur early in the course of ARF and often coincides with a rising titer of streptococcal antibodies. The polyarthritis classically affects larger joints, such as the knees, ankles, elbows, and wrists, and the pain can be more severe than physical findings suggest. Analysis of the synovial fluid generally reveals a sterile inflammatory fluid.

Cardiac manifestations of ARF may be subtle and can include symptoms and signs of pericarditis, myocarditis, and endocarditis. The mitral valve is the most common valve affected in ARF, causing mitral regurgitation accompanied by a new, high-pitched, systolic murmur. Inflammation of the valvular endocardium can result in permanent deformity and impairment of one or more cardiac valves over the course of decades. Stenotic lesions of the mitral or aortic valves are unusual at presentation, but they are common late manifestations of RHD.

Chorea (Sydenham’s chorea and St. Vitus’ dance) is manifested by random, rapid, purposeless movements usually of the upper extremities and face. These can be associated with emotional outbursts. Chorea is relatively rare in ARF and tends to emerge after a longer latency period than some of the other manifestations. Erythema marginatum and subcutaneous nodules are found in fewer than 10% of cases of ARF. Their presence, however, should immediately suggest the diagnosis. Erythema marginatum is a nonpruritic, painless, evanescent “smoke ring” of erythema that commonly appears on the trunk and proximal extremities. Subcutaneous nodules are pea sized and nontender. They typically appear over the extensor surfaces of the wrists, elbows, knees, and occasionally the spine.

Diagnostic Strategies

In 1944, Jones formulated major and minor criteria for the diagnosis of ARF. After multiple revisions, the Jones criteria remain the diagnostic basis for this disease (Box 83-5).27 The diagnosis of ARF requires evidence of an antecedent streptococcal infection plus at least two major, or one major and two minor, manifestations from the Jones criteria. Although throat cultures are usually negative at the time of clinical onset of ARF, antistreptolysin antibody titers remain positive for 4 to 6 weeks from the time of infection. Erythrocyte sedimentation rate and C-reactive protein levels are typically elevated, and a prolonged PR interval is common and suggestive in ARF.

Management

All patients with ARF should receive antibiotic therapy regardless of the clinical history of pharyngitis. Penicillin can be administered orally (250 mg for children and 500 mg for adults, two or three times daily for 10 days) or intramuscularly (600,000 units of benzathine penicillin in children weighing <25 kg and 1.2 million units in adults as a one-time dose).

Treatment for arthritis consists of anti-inflammatory agents, most commonly aspirin, administered until symptoms are absent and the erythrocyte sedimentation rate and C-reactive protein concentration normalize. Patients with severe carditis are often treated with corticosteroids, but their effects show conflicting results.28 Patients with congestive heart failure should be managed accordingly. Treatment of patients with ARF involves only symptom relief and does not decrease the likelihood of progression to RHD. Primary prevention involves treating those with group A streptococcal pharyngitis within 9 days of the onset of symptoms because this greatly decreases the risk of ARF. Patients with a history of ARF should receive ongoing prophylactic antibiotics (generally penicillin) to prevent recurrences. The recommended duration of secondary prophylaxis varies depending on the presence and severity of cardiac involvement.29

Valvular Heart Disease

Valvular Anatomy

Of the four heart valves, three (tricuspid, pulmonic, and aortic) are composed of three cusps, whereas the mitral valve has only two cusps. Each cusp is a double layer of endocardium that is attached at its base to the fibrous skeleton of the heart. The margins of the cusps are attached to muscular projections from the ventricles (papillary muscles) via tendinous cords (chordae tendineae). Contraction of the ventricle, and consequently the papillary muscle, results in the opening or closing of the valve depending on its location.
Mitral Stenosis

The most common cause of mitral stenosis is RHD. Symptoms of valvular dysfunction typically develop after a latency period of one to three decades. Many patients will not recall a history of ARF. Less common causes of mitral stenosis include congenital mitral stenosis and mitral annular calcification.

Pathophysiology

The normal cross-sectional area of the mitral valve orifice is 4 to 6 cm². Stenosis becomes clinically significant when the area falls below 2 cm². Impeded flow from the left atrium to the left ventricle results in left atrial hypertension, restricted cardiac output, and, ultimately, pulmonary congestion. As the disease progresses, patients may develop pulmonary hypertension and right ventricular failure.

The most common complication of mitral stenosis is atrial fibrillation, which in the absence of rate control is not well tolerated. Patients with underlying mitral stenosis will decompensate under other conditions associated with increased cardiac demand and reduced ventricular filling, such as pregnancy, anemia, infection, and hyperthyroidism.

Clinical Features

Early symptoms of mitral stenosis include reduced exercise tolerance and dyspnea on exertion. Patients with more advanced disease may have orthopnea and, if right ventricular failure is present, peripheral edema. Hemoptyysis, caused by rupture of a bronchial vein, and hoarseness, caused by compression of the recurrent laryngeal nerve, are classic but rare presentations. Aside from the typical signs of heart failure, findings that suggest the presence of mitral stenosis include a loud S1 and an opening snap in early diastole accompanied by a low-pitched, rumbling diastolic apical murmur.

Although the chest radiograph may be normal, in more advanced cases, left atrial enlargement may be suggested by straightening of the left heart border. Common ECG abnormalities in addition to atrial fibrillation include left atrial enlargement and, ultimately, right ventricular hypertrophy. Echocardiography confirms the diagnosis and assesses the severity of disease.

Management

Medical treatment for patients with mitral stenosis comprises diuresis for symptoms of vascular congestion and anticoagulation for atrial fibrillation. Once symptoms have developed, however, median survival without intervention is 7 years. Several surgical options exist, ranging from balloon valvulotomy or open commissurotomy to valve reconstruction or replacement. Management of the patient with mitral stenosis in the ED centers on identification and treatment of underlying precipitants such as atrial fibrillation or anemia, diuresis, and referral for definitive intervention.

Mitral Regurgitation

Acute and chronic mitral regurgitation are two distinct disease entities. Acute mitral regurgitation is a true emergency. It can result from idiopathic rupture of chordae tendineae, papillary muscle dysfunction in the setting of acute ischemia (or rupture 2-7 days postinfarction), or rarely, perforation of a valve leaflet in the setting of infectious endocarditis or trauma. Chronic mitral regurgitation, on the other hand, most commonly occurs either in the setting of dilated cardiomyopathy (caused by enlargement of the mitral annular ring) or in the setting of RHD. This frequently coexists with mitral stenosis. Other causes of chronic mitral regurgitation include MVP and connective tissue disorders, such as Marfan syndrome and Ehlers-Danlos syndrome.

Pathophysiology

Acute mitral regurgitation is associated with low left atrial compliance and thus sharply elevated left atrial pressure that results in acute pulmonary congestion. In contrast, chronic mitral regurgitation is characterized by high left atrial compliance and near-normal left atrial pressures with reduced forward output. Patients with chronic mitral regurgitation typically decompensate in the setting of volume overload.

Clinical Features

The characteristic presentation of acute mitral regurgitation is one of fulminant pulmonary edema. This is accompanied by a unique, harsh, mid-systolic murmur that radiates to the base and not the axilla. Patients typically have no prior history of heart failure. The ECG may display signs of ischemia or infarction.

The presentation of chronic mitral regurgitation is similar to that of chronic systolic heart failure with clinical symptoms and signs of decompensated congestion. The murmur is classically described as holosystolic, heard best at the apex and radiating to the axilla. The ECG often reflects both left atrial and ventricular hypertrophy. Atrial fibrillation is common, and left atrial enlargement may be suggested by the chest radiograph. Echocardiography may demonstrate a normal or above-normal ejection fraction, but much of the diastolic flow is retrograde.

Management

When the diagnosis of acute mitral regurgitation is suspected, emergency echocardiography and cardiac catheterization will assess the degree of regurgitation and urgency for surgery. Initial stabilization should include treatment of pulmonary edema with nitrates and diuretics. In a hypotensive patient, a counterpulsation intra-aortic balloon pump may provide temporary stabilization as a bridge to surgery.

The natural history of chronic mitral regurgitation is generally a very slow progression with a 15-year survival approaching 70% with medical therapy, including diuretics and afterload-reducing agents. Once the ejection fraction falls below 60%, valve repair or replacement is advisable before the development of irreversible left ventricular dysfunction.

Aortic Stenosis

The most common cause of aortic stenosis is calcific degeneration, which is prevalent in the elderly with coronary artery disease. This also occurs in younger individuals with a bicuspid aortic valve. Aortic stenosis can also occur along with mitral stenosis in RHD.

Pathophysiology

The normal aortic valve area is greater than 3 cm². Significant obstruction occurs when the valve area is reduced by more than 50%. Critical aortic stenosis is defined by a valve area of less than 0.8 cm² or a pressure gradient across the valve that exceeds 50 mm Hg. Compensatory left ventricular hypertrophy can maintain cardiac output until the stenosis becomes severe. Further progression of disease is associated with left ventricular dysfunction, left atrial enlargement, and atrial fibrillation. Individuals with severe or critical aortic stenosis are preload dependent and have very little cardiovascular reserve. Disruption of the delicate
balance between myocardial oxygen supply and demand (e.g., ischemia, rapid atrial fibrillation, dehydration, and acute blood loss) can result in precipitous decompensation.

Clinical Features

Classic symptoms of aortic stenosis progress from angina (increased demand resulting from wall stress, and decreased supply resulting from reduction in perfusion pressure) to exertional syncope (fixed cardiac output and vasodepressor response) and congestive heart failure (diastolic and systolic dysfunction). In an older patient with chest pain, particularly if seemingly preload dependent, the possibility of aortic stenosis should be considered.32

The classic auscultatory finding in aortic stenosis is a crescendo-decrescendo systolic murmur heard best at the base (right second intercostal space) that radiates into the carotids and is associated with the presence of an S4 gallop and a soft aortic component of S1. It is important to note that as the severity of disease increases, the murmur peaks later and becomes less apparent. Carotid pulses may be delayed (tardus) and diminished in intensity (parvus). The ECG typically reveals left ventricular hypertrophy. Echocardiography is required for assessment of the severity of stenosis and the presence of left ventricular dysfunction.

Management

The natural history of aortic stenosis is one of slow progression without symptoms for years. Once symptoms develop, survival is markedly reduced unless the valve is replaced, and medical management has a limited role. Management of decompensated aortic stenosis in the acutely setting centers on judicious fluid resuscitation, blood transfusion, restoration of sinus rhythm, and avoidance of vasodilators and diuretics and inotropic agents if possible. When there is no response to medical therapy and the patient is a candidate for valve replacement, an intra-aortic balloon pump may provide a bridge to surgery.

Aortic Insufficiency

Aortic insufficiency, whether acute or chronic, may reflect valvular abnormalities caused by a congenital bicuspid valve, RHD, or infectious endocarditis. In addition, aortic root abnormalities, such as ectasia, aneurysm, or dissection, may be associated with a variety of connective tissue diseases, including Marfan syndrome.

Pathophysiology

With acute aortic insufficiency, left ventricular compliance is low and left ventricular pressure increases rapidly, leading to acute pulmonary congestion. The pressure gradient between the aorta and the left ventricle is minimal. In chronic aortic insufficiency, the left ventricle dilates, allowing the heart to maintain normal or near-normal cardiac output despite significant regurgitation. The enhanced stroke volume results in a wide pulse pressure and the associated clinical signs. Congestion is typically associated with volume overload.

Clinical Features

Patients with acute aortic insufficiency may have a history suggestive of aortic dissection or aneurysm or severe respiratory distress and even frank cardiogenic shock. Acute aortic insufficiency can cause subtle physical findings. The pulse pressure may not be widened, and the short, soft, diastolic murmur may be difficult to detect. Echocardiography, which may be required emergently, is diagnostic.

With chronic aortic insufficiency, a widened pulse pressure may be accompanied by a number of physical findings, such as a rapidly rising and falling carotid pulse (water-hammer or Corrigan’s pulse), spontaneous nail bed pulsations (Quincke’s sign), or a to-and-fro murmur over the femoral artery (Duroziez’s sign). A high-pitched, blowing, diastolic murmur at the left sternal border is characteristic of chronic aortic insufficiency. An Austin-Flint murmur, a soft diastolic rumble caused by the regurgitant stream against the mitral valve, may also be present.

Management

Acute aortic insufficiency is typically a surgical emergency necessitating urgent valve replacement. Medical stabilization entails the cautious use of vasodilators and diuretics. Intra-aortic balloon counterpulsation is contraindicated in the presence of an incompetent aortic valve. In contrast, chronic aortic insufficiency is managed like other types of decompensated heart failure. Valve repair or replacement should be contemplated before the development of left ventricular systolic dysfunction.33

Mitral Valve Prolapse

MVP is defined pathophysiologically as an abnormal movement of one or both of the mitral valve leaflets across the plane of the valve during systole. Although generally a benign condition, it is infrequently associated with more serious cardiac pathology such as mitral regurgitation, endocarditis, and arrhythmias. Echocardiographic studies report a true prevalence of less than 1% in both men and women versus the previously reported 5% with a female predominance.34

Pathophysiology

Structurally, MVP is characterized by myxomatous proliferation of the spongiosa layer within the mitral valve that results in abnormal billowing of the leaflet during systole. MVP may be associated with other connective tissue disorders, such as Marfan syndrome and Ehlers-Danlos syndrome. Although any of the leaflets may prolapse, involvement of the posterior leaflet is more likely to be associated with mitral regurgitation and other cardiovascular complications.

Clinical Features

MVP is associated with a wide variety of clinical symptoms, including chest pain, palpitations, dyspnea, lightheadedness, and fatigue. Appropriately controlled clinical studies, however, such as the Framingham Heart Study, suggest that patients with MVP and control subjects may be equally symptomatic.36 The classic auscultatory features of MVP are a mid-systolic “click” followed by a middiastolic to late systolic murmur over the mitral area. This click results from snapping of the chordae tendineae during the prolapse of the valve.

Diagnostic Strategies

The typical auscultatory findings should suggest MVP that is confirmed by echocardiography. Symptoms attributed to MVP, however, are often not explained by the degree of prolapse or mitral regurgitation. In some of these patients, autonomic or neuroendocrine dysfunction may be a cause of nonspecific symptoms.

Management

Cardioselective beta-blockers may control symptoms such as palpitations, chest pain, and anxiety. Lifestyle modifications, such as
Complications of Prosthetic Valves

Prosthetic heart valves are classified as either mechanical, constructed entirely of synthetic material, or biologic. The latter category includes whole valve transplants (porcine or human) as well as bioprosthetic valves manufactured from bovine pericardium. All prosthetic heart valves are associated with complications, ranging from structural failure and thrombosis to systemic embolization, hemolysis, and endocarditis. The diagnosis of a prosthetic valve complication can be challenging because symptoms and signs are often subtle. The overall incidence of complications is approximately 3% per year.

Primary structural failure is extremely uncommon with modern mechanical valves. When it does occur, the presentation is one of acute severe regurgitation and shock, mandating emergent replacement. With biologic valves, in contrast, structural failure is less dramatic but relatively more common. At 10 years, 20 to 30% of bioprosthetic valves will exhibit evidence of structural failure, and most are replaced electively. Symptoms are characteristically insidious in onset and mimic native valve disease.

Prosthetic valve thrombosis has an incidence of approximately 2% per year, both with biologic valves and with appropriately anticoagulated mechanical valves. Symptoms are of variable duration, are generally subacute, and typically mimic congestive heart failure. The diagnosis is often initially overlooked, and untreated mortality approaches 15%. On physical examination the diagnosis is suggested by a decreased or absent valve click, a new regurgitant murmur, or a louder than expected stenotic murmur. Echocardiography may demonstrate the thrombus or restricted leaflet motion suggesting obstruction. Treatment options include fibrinolytic therapy and surgery.

The incidence of systemic embolization from a prosthetic valve is approximately 1% per year. Compared with patients with aortic valve prostheses, those with mitral valve prostheses have twice the risk of systemic embolization, with an incidence roughly the same in the case of a biologic valve or mechanical valve in an appropriately anticoagulated patient. The target international normalized ratio is 3.0 to 3.5 with a mechanical mitral valve versus 2.5 to 3.0 with an aortic valve. The vast majority of diagnosed embolic events (85%) involve the central nervous system, and roughly half of these result in permanent impairment. In this context, the risk of continued anticoagulation and possible hemorrhagic conversion should be weighed against the risk of a second embolic event.

Hemolytic anemia resulting from sheer forces around a prosthetic valve is usually mild and subclinical, but it may be severe in up to 15% of patients with certain prostheses. Presenting features can be subtle and include dyspnea, fatigue, jaundice, or dark urine. Iron replacement is effective in the majority of patients, although transfusion can be required, and reoperation may be indicated if hemolysis is a result of a periprosthetic leak or other structural failure.

The incidence of PVE is highest during the initial months after surgery and is similar for both mechanical and bioprosthetic valves. Early PVE (within 60 days of surgery) is presumed to be caused by a pathogen acquired perioperatively and is associated with higher morbidity and mortality, whereas late PVE is more likely related to transient bacteremia and is generally associated with a more benign course. As with other forms of endocarditis, fever is by far the most common presenting symptom, and other manifestations are quite variable. Echocardiography can identify vegetations, but a normal study does not rule out endocarditis. In the ED, the diagnosis of PVE is presumptive because the definitive diagnosis requires blood cultures or biopsy. Therefore prosthetic valve patients with no obvious extracardiac source of fever, particularly within 60 days of surgery, should be considered for inpatient admission.

**KEY CONCEPTS**

- Many patients seen early in the bacteremic phase of IE lack a murmur and are indistinguishable from those with viremia.
- Patients for whom suspicion of endocarditis is moderate to high require blood cultures, echocardiography, and admission for definitive diagnosis and initiation of empirical therapy.
- Prophylaxis for IE is rarely if ever indicated for procedures performed in the ED.
- Acute rheumatic fever is a delayed, nonsuppurative complication of streptococcal pharyngitis characterized by arthritis, carditis, chorea, subcutaneous nodules, and erythema marginatum.
- In a patient with severe mitral stenosis, hypovolemia and tachycardia are poorly tolerated. “Slow and full” are appropriate goals.
- In patients with critical aortic stenosis, excessive preload reduction with vasodilators and diuretics is to be avoided.
- In patients with acute aortic insufficiency, classic physical findings may be absent. Medical stabilization entails the cautious use of vasodilators and diuretics. Intra-aortic balloon counterpulsation is contraindicated.
- Complications of prosthetic heart valves range from structural failure and thrombosis to systemic embolization, hemolysis, and endocarditis.

The references for this chapter can be found online by accessing the accompanying Expert Consult website.
References