PATHOPHYSIOLOGY AND PRESENTING SIGNS AND SYMPTOMS

Of the four heart valves, three are tricuspid (aortic, pulmonic, and tricuspid) and one is bicuspid (mitral). Failure of normal function of these valves is due to lesions that make them incompetent and allow backward flow (regurgitation) or to lesions that decrease orifice size and cause restriction of flow (stenosis). In addition, combinations of these lesions may occur within the same valve, and multiple valves may be involved (commonly the aortic and mitral valves with rheumatic heart disease). Cardiac murmurs result from (1) increased blood flow across a normal or abnormal valve orifice, (2) turbulent flow across a narrow or irregular orifice into a dilated blood vessel or cardiac chamber, or (3) backward flow across an incompetent valve or other cardiac defect. However, most systolic murmurs are related to a physiologic increase in blood velocity and are not pathologic.

Physical examination establishes where the murmur occurs within the cardiac cycle and its location, duration, and intensity. Murmurs are classified as systolic, diastolic, and continuous. Systolic murmurs are further subclassified into holosystolic (pansystolic), midsystolic, early systolic, and late systolic. Clicks can be heard from the snapping shut of diseased valves. Diastolic and continuous murmurs are nearly always pathologic and require investigation, even in the absence of symptoms. Although many systolic murmurs merit investigation, especially those associated with symptoms, the majority of systolic murmurs do not signify valvular disease.

A summary of the typical findings in the major valvular abnormalities is presented in Table 61.1. A heart affected by valvular pathology has the ability to compensate over time, and symptoms are commonly absent for decades. Emergency physicians need to be able to identify when certain valvular lesions have progressed to the point that they are clinically important and responsible for the patient’s symptom complex. Shortness of breath, arrhythmias, and heart failure are common reasons why patients with VHD seek treatment at an ED. Other clinical scenarios include valve infection, myocardial infarction with papillary muscle dysfunction, and failure of a prosthetic device, which can cause rapid heart failure and shock. It should be kept in mind that valvular pathology is in the differential diagnosis of patients with congestive heart failure, shock, and angina. The clinician should perform a careful cardiac examination with attention...
Patients with fever and new heart murmurs may have pain, shortness of breath, arrhythmia, syncope, or shock. Valvular causes must be kept in mind in patients with chest ventricular hypertrophy (LVH) or atrial fibrillation (AF). ECG may reveal signs of left atrial enlargement with tall or biphasic P waves, as well as left ventricular hypertrophy (LVH) or atrial fibrillation (AF).

**Table 61.1** Characteristics of Common Cardiac Murmurs

<table>
<thead>
<tr>
<th>VALVE PATHOLOGY</th>
<th>MURMUR</th>
<th>LOCATION</th>
<th>VALSALVA EFFECT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic stenosis</td>
<td>Systolic crescendo-decrescendo</td>
<td>Base to neck</td>
<td>Decreases</td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td>Diastolic rumble</td>
<td>Apex</td>
<td>Decreases</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td>Diastolic rumble</td>
<td>Apex</td>
<td>Decreases</td>
</tr>
<tr>
<td>Mitral regurgitation</td>
<td>Holosystolic</td>
<td>Apex to axilla</td>
<td>Decreases</td>
</tr>
<tr>
<td>Mitral valve prolapse</td>
<td>Midsystolic click, late crescendo-decrescendo</td>
<td>Apex to axilla</td>
<td>Increases</td>
</tr>
<tr>
<td>Tricuspid regurgitation</td>
<td>Holosystolic</td>
<td>Left sternal border</td>
<td>Decreases</td>
</tr>
</tbody>
</table>

A new diastolic murmur in a patient with chest or back pain should raise suspicion for aortic dissection and requires urgent testing and consultation, as well as control of the heart rate and blood pressure. Patients with valvular heart disease may become hemodynamically unstable with rapid atrial fibrillation and require cardioversion. Infectious endocarditis should be considered in all patients with a prosthetic valve and fever. Blood cultures and empiric antibiotics are indicated. Beside echocardiography should be performed in patients with shortness of breath who have recently undergone prosthetic valve replacement to rule out pericardial tamponade.

**Differential Diagnosis and Medical Decision Making**

Valvular causes must be kept in mind in patients with chest pain, shortness of breath, arrhythmia, syncope, or shock. Patients with fever and new heart murmurs may have infectious endocarditis. An aortic insufficiency (AI) murmur in a patient with chest pain would lead to a diagnostic work-up for aortic dissection. Patients sustaining blunt trauma may rarely have traumatic aortic injury and, though rare, acute traumatic valvular injury as well.

**Specific Valve Lesions**

**Aortic Stenosis**

Aortic stenosis (AS) is the third most common form of cardiovascular disease in the Western world after hypertension and coronary artery disease. The most common causes of AS are calcification of a normal tricuspid valve and a congenital bicuspid aortic valve. Calcific degeneration of the aortic valve results from an inflammatory process similar to atherosclerotic vascular disease; it begins with intimal injury, such as...
calcification from the base of the cusps to the leaflets, which impairs motion, and progresses through fusion of valve leaflets and stenosis of the aortic valve orifice. When rheumatic heart disease is the culprit, the aortic valve commonly exhibits both stenosis and regurgitation and is usually associated with concomitant mitral valve disease.

The degree of aortic valve disease is based on several factors (Table 61.2). Such assessment may be inaccurate and underestimate valve area and jet velocity in patients with impaired cardiac output. Progression of AS is usually quite slow, with symptoms taking decades to become manifested in most cases. An average 0.1-cm² decrease in valve area occurs per year. The left ventricle is faced with systolic pressure overload and compensates through hypertrophy of the left ventricular (LV) wall. Normal chamber volume is maintained, but with increased wall thickness. The hypertrophic wall may lead to decreased coronary blood flow, even in the absence of stenosis and angina. Diastolic dysfunction and heart failure symptoms may also be present. These patients are dependent on forceful atrial contraction to maintain elevated LV end-diastolic pressure (LVEDP) to overcome the obstruction to outflow. Therefore, individuals who have AF and AS may suffer hemodynamic consequences from loss of the atrial kick.

Auscultation reveals a systolic murmur associated with diminished and delayed carotid pulses (parvus et tardus), a sustained LV impulse on palpation, and a decreased or absent aortic component of the second heart sound (S₂). Parvus et tardus may not be readily apparent in the elderly because of decreased compliance of the aging arterial vessels. Chest radiography may show normal heart size (remember concentric LVH) with rounding of the LV border. ECG can show LVH with a repolarization abnormality, which is seen in 85% of patients with severe AS. Echocardiography is indicated every year for patients with severe AS to assess the severity of the AS, wall thickness, and LV function. Exercise stress testing may lead to complications in patients with symptomatic AS and should not be performed.

Asymptomatic patients with AS have the same life expectancy outcome as age-matched controls. Once symptoms appear, the average survival is 2 to 3 years, with a risk for sudden death (≤1% per year). Angina develops in 35% of patients, syncope in 15%, and congestive heart failure or dyspnea in 50%.

No medical treatment has been shown to decrease progression of disease in the leaflets, although statins are currently being studied. Significant care should be taken when treating these patients with preload-reducing agents (nitrates) because the sudden decrease in preload can cause severe hypotension, which in turn can lead to decreased coronary flow and worsened ischemia and shock. Cardiac catheterization is indicated in patients who have a discrepancy between the clinical complex and the results of echocardiography. Patients in whom cardiac surgery is planned need to undergo cardiac catheterization to assess the need for a concomitant coronary artery bypass grafting (CABG) procedure. The average mortality in patients who undergo aortic valve replacement (AVR) is 3% to 4% and, when associated with a CABG procedure, 5.5% to 6.8%. Percutaneous valvuloplasty is the use of a balloon to fracture calcium deposits in the leaflets. It results in immediate improvement in valve gradients; however, restenosis occurs in 6 to 12 months, and it is therefore not recommended for definitive treatment. This procedure has been used as a temporizing measure in patients who are not initially AVR candidates. Procedures are now available that allow percutaneous AVR in such patients.

### Table 61.2 Aortic Stenosis—Severity of Disease

<table>
<thead>
<tr>
<th>SEVERITY</th>
<th>MEAN GRADIENT (mm Hg)</th>
<th>VALVE AREA (cm²)</th>
<th>JET VELOCITY (m/sec)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>&lt;25</td>
<td>&gt;1.5</td>
<td>&lt;3</td>
</tr>
<tr>
<td>Moderate</td>
<td>25-40</td>
<td>1-1.5</td>
<td>3-4</td>
</tr>
<tr>
<td>Severe</td>
<td>&gt;40</td>
<td>&lt;1</td>
<td>&gt;4</td>
</tr>
</tbody>
</table>

Aortic Regurgitation

Multiple conditions can cause aortic regurgitation (AR); the most important for the emergency department (ED) physician are aortic dissection, trauma, and endocarditis. Patients with congenital bicuspid valves are also at risk for both AI and dissection. Rheumatic heart disease is still the most common cause of AR worldwide. Cardiac examination in patients with AR reveals a murmur during diastole, more low pitched and associated with a widened pulse pressure and displaced LV impulse. Eponyms associated with AR include Corrigan pulse (rapid rising pulse that falls rapidly), Quincke pulse (capillary pulsations at the base of the nail when pressure is applied at the tip), and the Duroziez sign (to-and-fro murmur over the femoral arteries).

Patients with acute AI have a sudden large regurgitant blood volume. Because the heart has not had time to compensate, large rises in LVEDP and left atrial pressure and a decrease in stroke volume occur. Patients are seen in shock, with hypotension and pulmonary edema. If the LVEDP and aortic pressure gradients are close enough, decreased coronary blood flow and myocardial ischemia can develop. This low gradient may make it impossible to hear a murmur. Emergency bedside echocardiography with color flow Doppler imaging is necessary to establish the diagnosis.

Medical management of acute AI should be directed toward reducing afterload and increasing contractility. Combinations of nitrupresside and dobutamine may augment forward flow and reduce regurgitant volume and LVEDP. Intraaortic balloon pump treatment is contraindicated because it increases regurgitant volume as it inflates during diastole. Ideally, patients with acute AR have a lesion that is surgically amenable and medical management is only a bridge to definitive therapy.

In patients with chronic AI the left ventricle has had time to compensate to the increased volume by increasing both LV end-diastolic volume and LV wall compliance. This allows accommodation of the increased regurgitant volume without significantly increasing chamber pressure. Stroke volume is increased to make up for the backward flow volume.

Echocardiography is essential in the diagnostic management of patients over time. Patients who have AI, normal LV function, and no symptoms do not need any medical treatment. Valve replacement is recommended in patients with severe AR or LV dysfunction, regardless of whether they are
symptomatic. Medical therapy with vasodilating drugs (nifedipine, hydralazine, angiotensin-converting enzyme inhibitors) is reserved for patients with severe AR who are not surgical candidates or as short-term treatment before valve replacement. Beta-blockers will theoretically increase time during diastole and worsen the amount of regurgitant volume. The rate of progression from an asymptomatic patient with normal LV function to an asymptomatic patient with LV dysfunction is 3.5% per year. Once LV dysfunction is detected on ultrasound, most patients will become symptomatic in 2 to 3 years.

MITRAL STENOSIS

The normal mitral valve has an area of 4 to 5 cm², and symptoms usually begin when the valve area becomes less than 2.5 cm². Mitral stenosis (MS) nearly always results from previous rheumatic fever. In fact, a history of rheumatic fever can be elicited in up to 60% of patients with MS. A period of 20 to 40 years may transpire between the occurrence of rheumatic fever and the onset of symptoms, and another 10 years may elapse before these symptoms become disabling. The ratio of women to men with MS is 2:1. MS can also result from severe degenerative calcification, especially in the elderly. Dyspnea is the cardinal symptom of significant MS. Shortness of breath may not develop in patients with milder MS until AF with rapid ventricular response or pregnancy occurs or it is precipitated by exercise or infection.

Because the main problem in MS is obstruction to blood flow, no medical therapy is available that will relieve this obstruction. The principal treatment is surgical repair or valve replacement. Beta-blockers will cause bradycardia and increase diastolic filling time and may benefit patients in whom symptoms develop secondary to tachycardia. Anticoagulation is indicated in patients with MS and AF because systemic embolization occurs in 10% to 20% of patients with MS.

Percutaneous mitral balloon valvotomy in the hands of skilled operators can be the procedure of choice in patients with moderate to severe MS who are symptomatic (significant MR and left atrial thrombus are contraindications to this procedure). Open surgical commissurotomy and mitral valve replacement are other alternatives for mitral valve treatment.

MITRAL REGURGITATION

Causes of mitral regurgitation (MR) can be split into organic and functional categories. Organic causes include mitral valve prolapse (MVP), rheumatic fever, endocarditis, and certain medications (most recently the use of diet supplements). The most common functional cause of MR is secondary LV dilation leading to a dilated annulus. Other functional causes include ruptured chordae tendineae, ruptured papillary muscle, and infective endocarditis. Patients with MR may exhibit a holosystolic murmur in association with a first heart sound (S₁) that is soft or absent and, frequently, a normal second heart sound (S₂).

Patients with acute MR have acute volume overload on the left atrium and left ventricle and, given lack of time for compensation, both decreased cardiac output and increased pulmonary congestion. The role of medical therapy is limited to stabilizing the patient’s hemodynamics before surgery. Diminishing MR, increasing forward flow, and improving pulmonary congestion can all be accomplished by afterload reduction with nitroprusside (if the patient can tolerate it) or an inotrope (dobutamine). An intraaortic balloon pump can also be used as a temporizing measure before definitive treatment. Therapy for acute papillary muscle rupture is emergent valve repair or replacement.

Clinically, however, most patients will have chronic MR. The heart will compensate by increasing LV size (the usual mechanism for chronic volume overload). This increased size allows a higher volume at lower pressure. The duration of this asymptomatic chronic phase is about 6 to 10 years. Patients with ischemic cardiomyopathy can be treated with beta-blockers and angiotensin-converting enzyme inhibitors. AF rate control is also important. In patients with severe MR who are symptomatic, as well as in asymptomatic patients with severe MR and an ejection fraction of 30% to 60%, surgical therapy is indicated and includes mitral valve repair or replacement.

MITRAL VALVE PROLAPSE

MVP is defined as billowing of the leaflets into the left atrium with or without associated MR. Current estimates of MVP based on newer diagnostic criteria suggest that less than 2.5% of the population is affected. Familial causes may represent a manifestation of connective tissue disease. Patients with MVP may experience chest pain, anxiety, palpitations, and dyspnea. Physical examination reveals a midsystolic click, frequently in association with a late systolic murmur. Patients suspected of having MVP should arrange follow-up with a cardiologist or primary care physician and will probably undergo echocardiography. Patients with MVP who have had a transient ischemic attack or cerebrovascular attack should be treated with daily acetylsalicylic acid.

ENDOCARDITIS AND RHEUMATIC FEVER PROPHYLAXIS

Recently, guidelines for the prophylaxis of infective endocarditis have undergone changes based on working group review. No randomized controlled trial examining the efficacy of prophylaxis before procedures has ever been conducted. In fact, endocarditis is more likely to be related to random bacteremia with daily activities than to specific dental, gastrointestinal, or genitourinary procedures. Therefore, prophylaxis would prevent only a very small number of cases. Because antibiotest have side effects, in many scenarios the risk exceeds any possible prophylactic benefit. High-risk events include dental procedures that involve the gingiva. Prophylaxis would be recommended in (1) patients with prosthetic valves or prosthetic material, (2) patients with a history of endocarditis, (3) patients with valve disease and a transplanted organ, and (4) high-risk patients with congenital heart disease. Antibiotic prophylaxis is not recommended for gastrointestinal or genitourinary procedures or for respiratory procedures.

Patients who have previously had rheumatic fever are at higher risk for the development of another episode of rheumatic fever. Because antibiotics have been shown to prevent recurrent attacks, prophylaxis with penicillin or an antibiotic of the macrolide class is recommended.

PROSTHETIC VALVE DISORDERS

Valve repair and replacement surgery are common procedures that have been significantly refined over the past 40 years, and approximately 100,000 prosthetic valves are implanted in
patients in the United States and 300,000 worldwide. Prosthetic heart valves are divided into two broad classes, mechanical and biologic. Mechanical valves have evolved considerably since their introduction in 1965 with the Starr-Edwards ball valve. Shortly thereafter, in 1969, disk valves were introduced with the Björk-Shiley valve. Problems involving strut malposition led to discontinuation of this valve in the United States in 1986. Bileaflet valves, including the St. Jude valve, which was introduced in 1977, are the most commonly implanted mechanical valves today.

Biologic valves are used in patients in whom long-term systemic anticoagulation is less desirable, and their use must be balanced against the need for reoperation because these valves will degenerate over the years. The most common biologic valve in the United States is the Carpentier-Edwards valve. Valves in the mitral position are at higher risk for thrombus formation than are valves in the aortic position, and some clinicians will treat bioprosthetic valves in the mitral position with warfarin.

A lateral chest radiograph can help in identifying a prosthetic valve. An imaginary line is drawn from the cardiac apex to the carina. Valves above this line are aortic and pulmonic, whereas valves below this line are mitral and tricuspid (Fig. 61.1).

Early complications after valve replacement include pericardial effusion with tamponade (pericardial inflammation and bleeding, which is exacerbated by anticoagulation), perioperative myocardial infarction, Dressler syndrome, AF, and early endocardial infection. Late prosthetic valve complications include AF, thromboembolic phenomena, endocarditis, valve malfunction as a result of thrombus or tissue ingrowth (pannus formation), and mechanical hemolytic anemia. Valve complications should be considered in any patient with a prosthetic valve who has symptoms of dyspnea, syncope, angina, or fever or has neurologic signs or symptoms. Absence of a murmur does not rule out prosthetic valve dysfunction. Cardiology consultation and admission for further evaluation are warranted. In patients with mechanical heart valves and reversal of the international normalized ratio (INR) who have previously had thromboembolism, cessation of warfarin therapy is associated with a 10% to 20% annual risk for recurrent thromboembolism. Therefore, the risk is relatively small for reversal if brief (days), and these risk-benefit decisions must be individualized: Because its onset of action of vitamin K is delayed, its use should be avoided because it will only lead to a prolonged time for achieving a therapeutic INR once warfarin is reinstituted.

**TREATMENT**

In all patients with VHD, the goals of the emergency physician are to (1) determine whether valvular abnormality is the cause of the signs and symptoms, (2) optimize the hemodynamic status of patients who have suffered acute decompensation, (3) remember that certain subgroups of patients may need prophylaxis for endocarditis, and (4) make the appropriate consultations with cardiology and surgery.

Patients with new heart murmurs should undergo focused physical examination, ECG, and chest radiography. The outpatient arena is usually the venue for asymptomatic patients. Innocent murmurs will not need any further evaluation, whereas other murmurs will require outpatient echocardiography; communication with the patient’s primary care physician or cardiologist (or both) is essential.

It is critical to consider VHD as a cause of the symptoms in all patients with shock, dyspnea, chest pain, syncope, or other symptoms that are consistent with worsening heart failure or arrhythmia. Echocardiography is recommended for all asymptomatic patients in whom the findings on evaluation are consistent with valvular abnormality, as are hospital admission and cardiology consultation. It may be difficult to establish a valvular cause of cardiogenic shock based on physical examination only, and these patients will require emergency echocardiography.

Management of patients with acute valvular decompensation requiring stabilization and hemodynamic support is similar to that for any critically ill patient and begins with airway management. Assessment of circulation includes examination of the pulse, blood pressure, and systemic oxygen delivery. Skin color, temperature, capillary refill time, and measurement of serum lactate and B-type natriuretic peptide should be performed. Maneuvers should then be undertaken to improve cardiac output to meet systemic oxygen demand. Treating shock in patients with acute valvular disorders requires a thorough understanding that maximizing cardiac output and systemic oxygen delivery does not necessarily mean increasing blood pressure. Similarly, patients with pulmonary congestion may not all benefit from a significant reduction in circulating volume or preload. In general, patients who are hypotensive should benefit from volume infusion unless they exhibit overt signs or symptoms of pulmonary edema. However, in patients with MR, greater preload or volume may result in an increase in mitral orifice size and thus in regurgitant volume. In patients in whom an acute coronary
syndrome is a possibility, emergency diagnostic cardiac catheterization may be appropriate. An intraaortic balloon pump may also be useful to a failing heart. A bedside ED echocardiogram demonstrating a pericardial effusion will hasten the work-up for aortic arch dissection.

**DISPOSITION**

Medical and surgical management of patients with VHD is based on symptomatology and echocardiographic measurements. Therefore, patients seen in the ED with symptoms and findings on physical examination, ECG, and chest radiography consistent with valvular dysfunction need an expedited work-up and should be admitted to the hospital for echocardiography and cardiology evaluation. Patients in cardiogenic shock will need intensive care unit management, and if a valvular cause is strongly being considered, such management should occur at an institution with the capability of cardiac surgery because emergency valve surgery may be needed. Prosthetic valve endocarditis should be considered in all patients with artificial valves who exhibit fever, congestive heart failure, or thromboembolic phenomena. Antibiotics need to be instituted empirically after adequate blood is obtained for culture.

**PATIENT TEACHING TIPS**

Murmurs are the sound that blood makes as it moves through the heart, and most of these sounds are normal and do not represent disease.

An ultrasound of your heart is the way that the doctor determines whether the heart valve is leaking (regurgitation) or whether its opening is narrowed (stenosis).

In most situations, people with heart murmurs and no other symptoms do not need any treatment. Some categories of heart murmurs will require monitoring with ultrasound to evaluate for progression of disease.

Chest pain, fainting, shortness of breath, and palpitations are symptoms that require evaluation by a physician and possible cardiology referral.

**DOCUMENTATION**

Intensity (grade 1-6 out of 6), timing during the cardiac cycle, location, and radiation are the principal ways to characterize a cardiac murmur.

Comment on heart size and chamber enlargement on the chest radiograph and whether left ventricular hypertrophy is present on the electrocardiogram.

Document the rationale if transfer is required for urgent cardiology or cardiothoracic surgery consultation.

Document risk-benefit discussions with patients and consultants when reversing anticoagulation in patients with mechanical prosthetic valves.

**REFERENCES**

References can be found on Expert Consult @ www.expertconsult.com.
REFERENCES