CHAPTER 123: Valvular Heart Disease

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IMMEDIATE CONCERNS

Critically ill patients with valvular heart disease (VHD) presenting to the intensive care unit (ICU) fall into three primary categories: (a) patients who are critically ill as a result of acute onset, newly acquired VHD; (b) patients with exacerbation or complications of pre-existing VHD; or (c) patients with concomitant VHD who are critically ill from other causes. Most patients present with instability secondary to left heart valvular disease, which, if severe, impairs right heart function, but in patients, right heart valvular lesions can be the predominant problem. Hemodynamic consequences of uncompensated left-sided valvular lesions include diminished cardiac output with tissue hypoperfusion, and pulmonary venous hypertension with pulmonary edema that, if severe, leads to pulmonary arterial hypertension and right heart failure. Isolated right-sided valvular lesions present with reduced cardiac output and systemic venous congestion. Management is determined by the type of lesion and its hemodynamic consequences, and is modified by coexisting derangements. Noninvasive assessment of the hemodynamic derangement by history, physical exam, chest radiography, or transesophageal echocardiography (TEE) is essential, but useful information may also be derived from invasive measurements such as arterial blood pressure, cardiac filling pressures, cardiac output, mixed venous oxygen saturation, and calculated cardiovascular variables such as left ventricular stroke work index, systemic vascular resistance, and pulmonary vascular resistance. Invasive monitoring is particularly useful for guiding and assessing the results of management. Patients with life-threatening valvular disease generally present to the critical care unit with one or more manifestations of congestive heart failure that require immediate sta-


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can be invaluable in revealing pulmonary venous or arterial hypertension, pulmonary edema, pleural effusions, and lung parenchymal abnormalities, and in allowing evaluation of the cardiac contour. Because of the distortion produced by antero-posterior supine radiographs, every effort should be made to obtain sitting 183 cm (72-inch) posteroanterior radiographs as well as lateral radiographs when the patient's condition allows. Early performance of echocardiography is imperative in patients with unexplained heart failure. If the quality of the transthoracic echocardiogram is not optimal, transesophageal echocardiography should be performed.

**CRITICAL ILLNESS IN PATIENTS WITH UNDERLYING VALVULAR HEART DISEASE**

Patients with underlying valvular disease may become critically ill from noncardiac causes. The effect of valvular disease on the management plan is determined by the presence of other cardiac abnormalities, the severity of the physiologic derangements, and the resultant hemodynamic burden. Because these patients present with other illnesses, valvular disease is often detected only by the discovery of cardiac murmurs on physical examination, valve calcification on chest radiographs, or unexplained evidence of left ventricular hypertrophy or atrial abnormality on electrocardiogram (ECG). When abnormalities are suspected, echocardiography is the most useful diagnostic tool for defining the type and extent of the valvular abnormality.

Once the valvular abnormality is defined, its impact on the management plan can be determined by consideration of its severity and specific hemodynamic characteristics (see below). All valve lesions share several common considerations. Antibiotic prophylaxis for endocarditis is important since community-acquired, as well as nosocomial, infections are common in critically ill patients, particularly when invasive procedures are undertaken or indwelling catheters are inserted. Fever and increased work of breathing may increase oxygen demand to a degree not well tolerated, and should be treated vigorously. Sinus tachycardia, atrial fibrillation with rapid ventricular response, and paroxysmal atrial tachycardia reduce left ventricular filling time and may lead to hemodynamic deterioration. They should be aggressively treated, particularly in patients with severely stenotic lesions. Treatment of dysrhythmias includes correction of electrolyte abnormalities, judicious use of digoxin, and intermittent or constant infusion of β-adrenergic blockers, calcium channel blockers, amiodarone, or other antiarrhythmic drugs. Hemodynamically compromised patients who do not promptly respond to the above measures may need urgent cardioversion.

**CRITICAL ILLNESSES WITH SPECIFIC VALVULAR ABNORMALITIES**

**Aortic Stenosis**

**Etiology**

Aortic stenosis (AS) is the most common primary valvular heart disease. Stenosis of the normal tricuspid aortic valve caused by pathology similar to coronary artery disease is the usual cause. A calcific process may involve the bicuspid aortic valve, though the process is slower. Rheumatic aortic valve stenosis, though rare in industrialized societies, may be seen along with mitral valve disease.

**Hemodynamics**

Obstruction to forward blood flow causes compensatory concentric hypertrophy of the left ventricle. Hypertrophy decreases the wall stress, but comes at the price of increased oxygen demand and dependence of ventricular filling on left atrial contractions. As the hypertrophy increases, subendocardial ischemia predisposes these patients to ventricular dysrythmias. Decrease in forward flow with exercise and associated peripheral vasodilatation can cause syncope. Sudden death has been reported in patients with AS. Later in the course, the ventricles dilate and cardiac function is maintained by Frank-Starling mechanisms. The triad of symptoms—syncope, angina, and dyspnea—indicates severe AS and requires surgical intervention. Clinical signs of common valve lesions are noted in Table 123.1.

**Diagnosis**

Common features on ECG include left ventricular hypertrophy with strain pattern, left bundle branch block, and left atrial hypertrophy (biphasic P waves in precordial lead V1). Chest radiography may reveal a boot-shaped heart, calcification of the aortic valve, poststenotic dilatation of the aorta, and pulmonary venous congestion. Echocardiography is the principal modality for confirming the diagnosis of AS (Fig. 123.1); the severity of AS is determined by peak gradients across the aortic valve and calculation of valve area (Table 123.2). It is important to recognize that the gradients will be lower with severe aortic stenosis if the flow across the valve is reduced by hypovolemia or by poor left ventricular function. Echocardiography also provides information about left ventricular function. Patients with AS may have preserved systolic function but with significant diastolic dysfunction. Diastolic dysfunction predisposes these patients to pulmonary edema. Coronary angiography is indicated in patients with aortic stenosis before surgery to rule out associated coronary artery disease (CAD).

**Therapeutic Considerations**

Patients with AS may require ICU admission because of acute cardiogenic shock, pulmonary edema, severe angina, ventricular dysrythmias, or, less commonly, atrial fibrillation and systemic embolization.

**Drugs**

Drugs commonly used to treat these conditions carry significant risks in patients with AS. β-Blockers, calcium channel blockers, and other antiarrhythmic drugs should be used with caution as patients with AS are sensitive to drugs causing myocardial depression. Pulmonary congestion should be relieved by careful administration of diuretics, digoxin, and nitroglycerin. The use of diuretics and nitroglycerin, however, may result in inadequate preload. Digoxin carries the risk of dysrythmias. Increased left ventricular mass and intracavitary systolic pressure increase oxygen demands, thereby decreasing the patient's tolerance to vasodilatation and tachycardia. Angiotensin-converting enzyme (ACE) inhibitors and other vasodilators such as nitroprusside may precipitate syncope or even sudden death in severe AS, and are relatively contraindicated.
### Table 123.1

**CLINICAL SIGNS IN VALVULAR HEART DISEASE**

<table>
<thead>
<tr>
<th>Aortic stenosis</th>
<th>Aortic regurgitation</th>
<th>Acute mitral regurgitation</th>
<th>Chronic mitral regurgitation</th>
<th>Mitral stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>General signs</td>
<td>Nothing remarkable</td>
<td>Look for Marfan syndrome, ankylosing spondylitis, or seronegative arthropathies</td>
<td>Tachypnea, circulatory shock</td>
<td>Tachypnea, mitral facies (malar flush), tachycardia, peripheral cyanosis</td>
</tr>
<tr>
<td>Pulse</td>
<td>Small volume (parvus) and late peaking (tardus)</td>
<td>Water-hammer pulse, wide pulse pressure</td>
<td>Sinus tachycardia</td>
<td>Irregularly irregular in AF</td>
</tr>
<tr>
<td>Neck and JVP</td>
<td>Prominent a wave</td>
<td>Prominent carotid pulsations (Corrigan sign)</td>
<td>Prominent a wave</td>
<td>Absent a wave in AF</td>
</tr>
<tr>
<td>Precordium</td>
<td>Sustained, nondisplaced or slightly displaced apical impulse, palpable S4, systolic thrill at the base and at carotid</td>
<td>Diffuse, hyperdynamic and displaced apical impulse</td>
<td>Hyperdynamic, infolateral displaced apical impulse, parasternal heave (LAE)</td>
<td>Tapping apical impulse (palpable S2), palpable P2 and parasternal heave in PHT, diastolic thrill rarely</td>
</tr>
</tbody>
</table>

**Auscultation**

<table>
<thead>
<tr>
<th>S1</th>
<th>Soft</th>
<th>Soft in acute AR</th>
<th>Normal/soft</th>
<th>Soft</th>
</tr>
</thead>
<tbody>
<tr>
<td>S2</td>
<td>Narrow split or reverse split S2, absent A2 in severe AS</td>
<td>P2 loud in acute AR</td>
<td>Accented P2/wide paradoxical split</td>
<td>Normal P2/wide paradoxical split</td>
</tr>
<tr>
<td>S3/S4</td>
<td>Prominent S4</td>
<td>S3 heard</td>
<td>Present/present</td>
<td>Present/absent Mid systolic click in MVP</td>
</tr>
<tr>
<td>Clicks and added sounds</td>
<td>Systolic ejection click indicates midsystolic murmur in advanced disease</td>
<td></td>
<td></td>
<td>Opening snap</td>
</tr>
<tr>
<td>Murmur</td>
<td>Systolic soft musical murmur, decreasing intensity in advanced disease</td>
<td>High-pitched, long diastolic murmur in chronic versus low-pitched short diastolic murmur in acute AR, along left sternal border, diastolic murmur of early mitral diastolic closure</td>
<td>Early systolic loud radiating toward base (anterior directed jet) or axilla (posterior directed jet)</td>
<td>Holosystolic, soft or harsh and radiating toward axilla/abdomen, late systolic murmur in MVP</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Mid diastolic murmur with late diastolic accentuation best heard at apex</td>
</tr>
</tbody>
</table>

AF, Atrial fibrillation; PHT, pulmonary hypertension; LAE, left atrial enlargement; MVP, mitral valve prolapse; AR, aortic regurgitation; AS, aortic stenosis.

Undesirable hemodynamic effects of sedative drugs such as propofol, benzodiazepines, and narcotics should also be considered. Propofol can cause significant vasodilatation and hypotension, and therefore should be avoided in patients with fixed cardiac output. narcotics can blunt the hypertensive sympathetic responses without significant myocardial depression and are usually the agents of choice.

**Fluids**

Fluid is administered with extreme caution in patients with AS. Patients with diastolic dysfunction, even when systolic function and ejection fraction are preserved, are extremely sensitive to fluids and can develop pulmonary edema. Filling pressures may need to be monitored with a pulmonary artery catheter (PAC) in such critical situations.

**Monitoring**

Hemodynamic stabilization with drugs and fluids should be carried out with careful monitoring of arterial pressure and cardiac filling pressures. Normal central venous pressure (CVP) does not ensure adequate filling pressures in patients with a stiff left ventricle. Placement of a PAC, while helpful, is not without risks, as it may precipitate malignant dysrhythmias and sudden deterioration. However, the PAC can provide useful
and II should be carried out in all October 25, 2008 15:6
Cardiovascular Disease and Dysfunction

Transesophageal echocardiographic appearance of se-

logic control of the ventricular rate is essential. It is imperative and hypotension. If cardioversion is unsuccessful, pharmaco-
be used, but carries an increased risk of myocardial depression amiodarone and/or cardioversion and initiation of antidysrhythmic therapy with tachycardia should be avoided because severe aortic stenosis results in a fixed stroke volume, therefore potentially reducing cardiac output. Adequate preload should be maintained, and one must recognize that afterload reduction may be hazardous as it can impair coronary perfusion pressure. Severe myocardial dysfunction with low blood pressure and ischemia may require administration of isotropes. Maintenance of adequate coronary perfusion pressure by vasopressors such as phenyl-
ephrine may be necessary in patients with optimized volume and myocardial contractile status. Patients who are refractory to medical management may benefit from insertion of an intra-
aortic balloon counterpulsation pump (IABP) to improve coro-
nary perfusion pressure.

Definitive Therapy
Considering the unfavorable natural history of AS, any ICU patient with severe AS who continues to deteriorate despite medical therapy should be seen by a cardiologist and a card-
diac surgeon for possible balloon valvotomy or open valve re-
placement. Balloon valvotomy affords temporary improvement in transvalvular gradient—usually with restenosis in about 6 months—and may relieve symptoms in some patients, thus serving as a bridge to definitive surgery. Balloon valvotomy is often very effective for young adults and adolescents with bi-
cuspid valves, although it carries a mortality of 10% in patients with calcific AS (1).

Aortic Regurgitation
Patients with aortic regurgitation (AR) may present to the criti-
cal care physician either because of decompensated chronic AR or due to acute onset of severe regurgitation.

Etiology
Acute AR results from infective endocarditis, with leaflet per-
foration, vegetations, or perivalvular fistula, and aortic dissec-
tion extending into the aortic annulus or aortic root, and from trauma, with avulsions of the annulus and tears of the cusps. Severe, acute hypertension may also cause sudden onset of AR that often reverses after control of hypertension. The causes of chronic AR are diverse; the disease may directly involve the valve or the aortic root. Primary valvular diseases include con-
genital bicuspid valve, prolapse of aortic cusp, rheumatic heart disease, calcific degenerative disease, connective tissue diseases, and subacute bacterial endocarditis. Diseases associated with aortic root dilatation include systemic hypertension, Marfan disease (Fig. 123.2), Ehlers-Danlos disease, granulomatous dis-
ases of the aorta, senile and cystic medial degeneration, annu-
loaortic ectasia, and syphilis.

Hemodynamics
In chronic AR, the left ventricle (LV) dilates and hypertrophies when subjected to volume overload. This keeps wall stresses in check and maintains normal forward stroke volume. As the disease progresses and the compensatory limit is reached, the wall stress begins to rise and systolic function deteriorates, with decreasing forward stroke volume as well as increasing LV end-
diastolic volume (LVEDV) and LVEDP, resulting in symptoms of heart failure. The presence of symptoms, systolic dysfunc-
tion, and an increase in end-systolic dimensions indicate severe

<table>
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<th>TABLE 123.2</th>
<th>ECHOCARDIOGRAPHIC ASSESSMENT OF AORTIC STENOSIS (AS)</th>
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<tbody>
<tr>
<td>Valve area (cm²)</td>
<td>Mild AS</td>
</tr>
<tr>
<td>Mean transvalvular gradient (mm Hg)</td>
<td>Less than 25</td>
</tr>
<tr>
<td>Jet velocity of blood flow across the valve (m/s)</td>
<td>Less than 3</td>
</tr>
</tbody>
</table>
and decompensated AR. During the relatively asymptomatic phase, the patient develops symptoms at a rate of 3.7% per year. In acute AR, the LV is subject to a sudden increase in volume, with no opportunity for a compensatory increase in compliance and eccentric hypertrophy to occur. Without these adaptations, the increase in end-diastolic pressures causes pulmonary venous congestion and pulmonary edema. The severity is dependent on the regurgitant orifice size, duration of diastole—as the degree of AR increases with bradycardia—and the diastolic pressure gradient between aorta and left ventricle. Thus, the patient with acute AR may present with heart failure if the AR is severe, or the initiating event—aortic dissection, trauma, or endocarditis—may dominate if AR is mild.

**Diagnosis**

An ECG is performed to rule out ischemic heart disease in situations of acute AR. The chest radiograph will reveal cardiomegaly in chronic AR or pulmonary congestion with a normal-sized heart in acute AR. TTE is performed to define the mechanism and severity of AR. Transeophageal echocardiography (TEE) is useful in patients with limited TTE windows. TEE defines the nature of perivalvular pathology (e.g., an abscess) better than TTE. An acute aortic dissection may be diagnosed with high sensitivity and specificity with TEE. Other techniques such as computerized tomographic (CT) scanning, magnetic resonance imaging (MRI), and aortography have been used. Each modality has its own advantages and disadvantages (2). TEE is clearly superior to MRI and CT scanning to characterize the valve pathology in cases of acute dissection, and obviates the need for aortography (Fig. 123.3). Coronary angiography can rule out ischemic heart disease before surgery in chronic AR, but is rarely indicated before emergency surgery in acute AR. Echocardiography may also be helpful to rule out the coronary artery involvement in dissections.

**Therapeutic Considerations**

Patients with acute AR are generally ill enough to require ICU admission. While medical therapy may allow patients with mild acute AR to reach a chronic compensated state, emergency aortic valve replacement is almost always indicated in a patient with severe acute AR after medical stabilization.

The principle of therapy is to optimize cardiac output and systemic perfusion, reduce pulmonary venous congestion, and initiate therapy for any underlying disorder. Invasive monitoring is initiated and volume is optimized. Tachycardia is beneficial in maintaining cardiac output, and decreases the regurgitant fraction by decreasing the duration of diastole. β-Blockers are avoided in acute severe AR before surgery, as it inhibits compensatory tachycardia and may precipitate circulatory failure. Hypertension and increased afterload are to be avoided; afterload reduction is indicated with vasodilators such as nitroprusside. Inotropic therapy is advised only in patients with depressed systolic function. The hemodynamic response to therapy is an increased cardiac output with reduction of filling pressures. An IABP is absolutely contraindicated in patients with AR, as it will increase the regurgitant fraction. In contrast to other causes of AR, inotropic therapy is avoided in patients with aortic dissection, as it occurs as a result of long-standing, poorly controlled hypertension or trauma. In both situations, left ventricular contractility is preserved and inotropes are not indicated. β-Adrenergic blockade may be initiated to reduce the velocity of LV ejection and aortic wall stress, therefore preventing extension of the aortic dissection or aortic rupture. In patients with chronic AR who present with an acute decompensation, a search should be made for the precipitating cause, with particular attention to possible infectious endocarditis. Most patients stabilize with medical therapy, but early elective surgery should be considered, as the outlook for medically treated symptomatic patients is poor. Decompensated patients who do not improve with aggressive medical therapy should undergo emergency valve replacement. Mortality with medical therapy alone in this group approaches 100%, while many moribund patients will survive with surgery.

**Mitral Regurgitation**

The mitral valve apparatus is composed of the valve leaflets, mitral annulus, chorda tendineae, papillary muscles, and adjacent...
cardiac chambers, namely the left atrium and left ventricle. Any disruption in the integrity of the mitral valve apparatus may result in regurgitation. One of the major breakthroughs in the management of mitral regurgitation was the functional classification of mitral regurgitation (MR) by Carpentier in early 1980s (3) (Table 123.3).

**Etiology**
An acute presentation of MR to the critical care physician usually results from infective endocarditis with leaflet perforations, vegetations, and perivalvular leaks; connective tissue or myxomatous disorders (e.g., chordal rupture); and ischemic heart disease—infarction and rupture of papillary muscles, or transient papillary muscle dysfunction due to ischemia. Acute rheumatic mitral valvulitis as the cause of mitral regurgitation is less common today.

**Hemodynamics**
Chronic MR leads to adaptation of the left ventricle by dilatation and eccentric hypertrophy. Over many years of increasing regurgitant volume, systolic function may fail, resulting in decreased ejection fraction (EF) and pulmonary hypertension. Left atrial (LA) dilatation leads to atrial fibrillation. In acute MR, on the other hand, the regurgitant volume is mainly ejected into the non-compliant left atrium. As a result, left atrial pressure increases, which further increases the regurgitant fraction. Patients may present with acute onset of fatigue, dyspnea, and chest pain, or can be admitted with pulmonary edema and circulatory shock, depending on the etiology. The differences in clinical signs between acute and chronic forms of MR are given in Table 123.1.

**Diagnosis**
The ECG may show atrial fibrillation, left ventricular hypertrophy, and right ventricular strain, while the chest radiograph may show cardiomegaly, indicating pre-existing heart disease. Pulmonary venous congestion and/or edema with a normalized heart indicate acute MR. An ECG may suggest the etiology in cases of ischemic MR.

Echocardiography remains the standard for the diagnosis of mitral regurgitation. TTE is easy, safe, and quick, and can be performed at the bedside. The mechanism and type of MR should be defined. Qualitative and quantitative assessment of MR can be done by color and spectral Doppler methods. Finally, the suitability for repair and left ventricular function should be assessed. TEE may provide better detail because of superior resolution and, further, has a significant advantage in that it allows definition of anatomic details of the mitral valve apparatus and mechanism of MR; the severity of MR is better evaluated by TEE (Fig. 123.4). Indices of left ventricular function such as EF are unreliable in the presence of severe MR. EF in MR is increased when contractility is normal. Normal EF indicates a significant loss of myocardial function; when EF is reduced to 50% or less, advanced myocardial dysfunction is generally present (4). The American Heart Association/American College of Cardiology (AHA/ACC) guidelines recommend medical treatment when EF is less than 30% and

<table>
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<tr>
<th>Type of MR</th>
<th>Pathology</th>
<th>Disease</th>
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</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Normal motion of leaflets</td>
<td>Endocarditis (leaflet perforation) or various etiologies causing left ventricular dysfunction (annular dilatation)</td>
</tr>
<tr>
<td>Type II</td>
<td>Increased leaflet motion with free edge of the leaflet traveling above the plane of the annulus; this is due to chordal elongation or rupture, papillary muscle rupture</td>
<td>Degenerative myxomatous valve disease</td>
</tr>
<tr>
<td>Type IIIA</td>
<td>Restricted leaflet motion during diastole and systole</td>
<td>Rheumatic heart disease</td>
</tr>
<tr>
<td>Type IIIB</td>
<td>Restricted leaflet motion during systole, papillary muscle displacement</td>
<td>Ischemic or dilated cardiomyopathy</td>
</tr>
</tbody>
</table>

**FIGURE 123.4.** Transesophageal echocardiographic diagnosis of mitral regurgitation (MR) severity. Left atrial wall hugging eccentric regurgitant jet indicates severe MR.
surgical treatment, even in asymptomatic patients, with EF less than 60% to prevent progression of disease (5). Urgent coronary angiography in impacted endocardial or subendocardial ischemia or infarction is a real consideration. Coronary angiography may delineate a culprit lesion, which may be amenable to catheter-based interventions. Severe triple-vessel disease should be referred for surgery. In the case of chordal rupture or infective endocarditis without risk factors for CAD, angiography may be deferred.

**Therapeutic Considerations**

In acute, severe MR, medical therapy has a limited role and is aimed at stabilizing the patient's hemodynamics in preparation for surgical intervention. Early valve surgery is life-saving, and therefore should not be delayed. Since acute MR is often a complication of myocardial infarction, therapy should include the need to maintain coronary perfusion pressure and reduce myocardial oxygen consumption. The utilization of a PAC and invasive arterial pressure monitoring is recommended. The pulmonary artery waveform may show typical V waves of mitral regurgitation. Sinus tachycardia maintains the forward flow and, thus, should not be suppressed. Hypertension and the use of vasopressors should be avoided, as the increased afterload will increase the regurgitant fraction. Diuretics may be needed to reduce pulmonary venous congestion. While in the normotensive patient, vasodilators—such as nitroprusside—can be useful in increasing the forward flow and decreasing the regurgitant fraction, the same cannot be said when the patient is hypertensive. In this case, nitroprusside may further impair coronary perfusion pressure and should not be used except in combination with inotropes. When patients fail to respond adequately to medical support, IABP may be life-saving, as it increases forward flow and blood pressure while diminishing filling pressures. It is effective in increasing coronary perfusion pressure and reducing oxygen consumption, the key factors in maintaining a favorable balance between myocardial oxygen supply and demand. Thus, IABP can be used to stabilize patients while they are prepared for surgery.

The surgical intervention is determined by the nature of the lesion. Chordal rupture or prolapse of the posterior leaflet can be successfully repaired. Revascularization alone may improve ischemic MR in selected patients with transient papillary muscle ischemia without rupture (6). Revascularization can be done using angioplasty, thrombolysis, or bypass surgery, depending on the anatomic characteristics and the severity of the lesions. However, revascularization alone may leave many of these patients with significant degrees of postoperative MR (7). Current lines of evidence suggest that a combination of complete revascularization with mitral valve repair or replacement with posterior chordal sparing is the standard of care for acute ischemic MR refractory to medical management and nonsurgical revascularization techniques (8). Mitral valve annuloplasty is the best approach for ischemic MR due to annular dilatation, diminished systolic contraction of the annulus, or papillary muscle malalignment. Mitral valve replacement should be considered if the repair is unsatisfactory. Papillary muscle rupture carries a high mortality without surgical intervention; surgery does improve the long-term outcome in functional class in these patients.

Infective endocarditis should be treated with antibiotics. However, if surgery is indicated, it should not be delayed for microbiologic clearance. Indications for surgery include congestive heart failure refractory to medical treatment, uncontrolled infection despite antibiotics, recurrent systemic embolism, perivalvular or pericardial extension of infection, fungal endocarditis, and prosthetic valve endocarditis (3).

Mitrval valve prolapse (MVP) deserves special mention because MVP is the most prevalent valvular heart disease and the most common cause of MR. Patients with this condition may present to the ICU because of severe MR, atrial fibrillation with transient ischemic attacks, long QT syndrome and tachyarrhythmias, pulmonary hypertension, cerebral embolism from MVP-related fibrin, infective endocarditis, or even sudden death (9). Medical stabilization and elective surgical repair of the mitral valve are recommended therapies.

**Mitrval Stenosis**

**Etiology**

Mitrval stenosis (MS) is mostly related to rheumatic heart disease. Degenerative calcific stenosis, congenital stenosis, and connective tissue disorders such as systemic lupus erythematosus and Lutembacher syndrome (atrial septal defect with MS) are the other causes for MS. Atrial myxomas and left atrial ball-valve thrombi may present with intermittent obstruction to mitral inflow and mimic MS.

**Hemodynamics**

In rheumatic MS, inflammation of the connective tissue leads to leaflet thickening and calcification, and commissural and chordal fusion. Ultimately, many patients are left with a funnel-shaped mitral apparatus, left atrial hypertension, and elevated left atrial pressure and the transmitral gradient increase, thus maintaining flow. However, the subsequent increase in pulmonary venous pressure leads to hydrostatic pulmonary edema. Pulmonary vasoconstriction increases pulmonary arterial pressure and right ventricular afterload. Over the course of rheumatic heart disease, persistently elevated pulmonary arterial pressure leads to structural changes such as intimal hyperplasia and medial hypertrophy. These structural changes are permanent and result in fixed pulmonary hypertension. Right ventricular dilatation and failure result in tricuspid insufficiency and systemic venous congestion, respectively. The impact of MS on cardiac output is initially determined by the severity of the stenosis itself and the limitation to flow across the narrowed mitral orifice. However, as the disease progresses, right ventricular failure may become severe enough to limit cardiac output. Similarly, progression of rheumatic disease with its associated inflammation, fibrosis, and calcium deposition may impair left ventricular function. Although this effect is not a major determinant of cardiac output in patients with MS, it may become important to consider in some patients after surgical repair. In such situations, the increase in transmural flow may expose the left ventricle to a sudden increase in preload, causing failure.

**Presentation**

Patients with mitral stenosis often present to the ICU with acute cardiogenic pulmonary edema. Precipitating factors such as infective endocarditis, fever, anxiety, pain, atrial fibrillation, and pregnancy should be identified. Occlusion of an enlarging atrial myxoma should be ruled out. Right-sided heart failure with hepatic dysfunction, acute hemoptysis, systemic embolism, and hoarseness of voice may also be present.
Hockey-stick appearance of mitral stenosis.

FIGURE 123.5. Typical appearance of rheumatic mitral stenosis.

Diagnosis

The ECG may show left atrial enlargement, right ventricular hypertrophy, and atrial fibrillation in MS. Chest radiography shows straightening of the left heart border, indicating left atrial and pulmonary artery enlargement. Kerley A and B lines indicate pulmonary venous hypertension. TTE findings include doming of the anterior leaflet, decreased leaflet mobility, increased leaflet calcification and thickness, commissural fusion, calcification of the subvalvular apparatus, increased LA size, and the presence of an LA thrombus (Fig. 123.5). Color flow will show associated mitral regurgitation. Doppler echocardiography allows calculation of pressure gradients, mitral valve area, and estimation of pulmonary artery systolic pressure. Cardiac MRI is increasingly being used in the evaluation of stenotic valvular lesions; it measures valve area by planimetry. Its advantage over echocardiography is the lack of dependence upon good echocardiographic windows.

Therapeutic Considerations

New onset atrial fibrillation with hemodynamic instability should be treated with cardioversion. Cardioversion of a patient with atrial fibrillation of unknown duration or that is known to have persisted for more than 48 hours must be preceded by 3 weeks of anticoagulation or by a TEE to exclude the presence of left atrial thrombus. Anticoagulation should be continued for 4 weeks following cardioversion because the enlarged LA remains “stunned” and does not recover a normal contractile state immediately following cardioversion (10). Anticoagulants should also be used in patients with a prior embolic event and left atrial diameter greater than 55 mm by echocardiography (3). Antidysrhythmics, such as amiodarone, may be used to maintain sinus rhythm but should not be expected to provide indefinite success.

Patients admitted with pulmonary edema should be stabilized with oxygen, morphine, anxiolytics, diuretics, and digoxin; the latter is especially useful in patients with atrial dysrhythmias and congestive heart failure. Sympathetic nervous system activity is increased in patients with mitral stenosis, and sympathetic overactivity worsens the symptom complex; β-blockers are very useful in this situation. Intravenous nesiritide, a synthetic human natriuretic peptide, is also used in the critically ill patients with acute decompensated cardiac failure and pulmonary hypertension. Short-term intravenous infusion of nesiritide is associated with hemodynamic and symptomatic improvements in patients with acutely decompensated congestive heart failure (CHF). Nesiritide may offer tolerability and practical advantages over currently used vasodilators, inodilators, and inotropes in this condition; in particular, nesiritide does not appear to have proarrhythmic effects. Nesiritide also appears to be effective and well tolerated in patients receiving concomitant β-blocker therapy and those with renal insufficiency (11, 12). If the patients are hypotensive, inotropes to improve left ventricular function may not be useful, but may worsen tachycardia and pulmonary edema. Inodilators may be useful in improving right ventricular dysfunction and reducing pulmonary hypertension. Systemic blood pressure may need to be supported with vasopressors, with the caveat that they may adversely impact pulmonary vascular resistance. Assessment of volume status is difficult, as the PCWP does not correlate with LVEDP in patients with MS. However, the PCWP gives useful information about the propensity to develop pulmonary edema.

Emergency invasive intervention is rarely required to relieve MS. If the precipitating events are controlled, intervention by surgery or balloon valvotomy can be scheduled electively after medical optimization. Balloon valvotomy is indicated in patients with suitable anatomy: pliable leaflets, no commissural fusion, and minimal subvalvular calcification. LA thrombus and significant (3+ to 4+) MR should be excluded by echocardiography. Patients who are not candidates for balloon intervention should be referred for surgery. Because of extensive calcifications and marked anatomic distortion, mitral valve repair often is not possible in rheumatic mitral disease, and replacement is necessary in those patients.

Tricuspid Stenosis

Etiology

Tricuspid valve obstruction can be due to anatomic disease of the tricuspid valve or functional, causes secondary to right atrial (RA) tumors and thrombus. Anatomic disease is usually related to rheumatic heart disease, and involvement of mitral valve is common. Other anatomic causes are carcinoid syndrome, infective endocarditis, congenital stenosis or atresia, and methysergide toxicity.

Hemodynamics

Obstruction to right ventricular inflow results in systemic venous congestion: elevated jugular venous pulse (JVP), congestive hepatomegaly, and peripheral edema. Diastole is shortened by increasing heart rate, thus causing dramatic increases in transvalvular gradients. Most patients are symptomatic from coexisting mitral stenosis. The presence of systemic venous congestion out of proportion to pulmonary venous congestion should raise the suspicion for involvement of the tricuspid valve.

Clinical Signs

Physical exam will reveal an elevated JVP, a prominent a wave, and distention of veins of the upper arm and dorsum of
Tricuspid Regurgitation

Etiology
Tricuspid regurgitation (TR) can be classified as structural or functional. Structural diseases of the valve are caused by rheumatic disease, infective endocarditis, carcinoid syndrome, radiation therapy, Marfan syndrome, congenital heart disease, or tricuspid valve prolapse. Functional TR is usually secondary to left-sided pathology, such as left ventricular failure or mitral or--less frequently--aortic valve disease, but may also result from primary pulmonary hypertension, pulmonic stenosis, right ventricular (RV) infarction, and dilated cardiomyopathy; a small number of cases may not have an etiology identified. While most cases of TR are chronic, an acute presentation may occur following penetrating trauma, RV infarction, infective endocarditis, and, more recently, repeated endomyocardial biopsies to diagnose allograft rejection of heart transplantation.

Clinical Symptoms and Signs
Symptoms of low cardiac output and venous congestion present in these patients. Shock and hypotension may develop following acute TR after RV infarction or papillary muscle rupture. Other patients maintain blood pressure but demonstrate signs of right-sided failure. Distended neck veins with a prominent c-v wave, pulsatile hepatomegaly, a precordial bulge, and parasternal heave from RV hypertrophy; soft S, -prominent P, and right-sided S from RV dilatation; pansystolic thrill; and a murmur heard at the left lower sternal border are associated signs. Cases of isolated tricuspid valve endocarditis may disseminate emboli to the lungs, resulting in multiple septic emboli and abscess formation. Peripheral stigmata of infective emboli are usually absent, but may indicate paradoxical embolism or left-sided lesions if present.

Diagnosis
The ECG may show atrial fibrillation, right axis deviation, RV hypertrophy, and a right bundle branch block. Right-sided leads will show ST elevation in RV infarction and may be associated with LV inferior wall infarction. Enlarged RV and cardiomegaly are seen on chest radiography. Echocardiography can yield details about the structural issues with the tricuspid valve—prolapse, vegetations, annular diameter, and rheumatic disease—and helps to rule out thrombus and patent foramen ovale. The diagnosis of a patent foramen ovale is important,
since the elevated right atrial pressure frequently exceeds left atrial pressure, causing a right-to-left shunt that may cause refractory hypoxemia. Severity of TR may be assessed by hepatic venous systolic flow reversal and estimation of regurgitant orifice area (greater than 40 mm² is severe). Also, pulmonary systolic pressure can be estimated by continuous wave Doppler signal of tricuspid regurgitation. Augmenting ultrasound signal with 10% air, 10% patient’s blood, and 80% saline, and estimation of pulmonary artery systolic pressures correlate well with PAC-measured PA systolic pressures (13). Right heart catheterization shows RV pressure greater than 60 mm Hg in functional TR compared to less than 40 mm Hg in structural TR.

**Therapeutic Considerations**

In general, the hemodynamic impact of acute TR is less severe and can be effectively managed with diuretics and inotropes. In rare instances, acute severe TR may require valvular surgery when the process is refractory to medical therapy. Once stabilized, these patients’ long-term prognosis depends on the etiology and severity of the TR. Patients tolerate mild and moderate degrees of chronic TR in the presence of normal LV function. Correction of left-sided heart lesions and treatment of left-sided failure and pulmonary hypertension take priority. If functional TR is severe, surgical intervention may be needed. Ring annuloplasty can improve TR and survive in these patients (14). Cardiac resynchronization therapy with biventricular pacemakers and the Dor procedure (endoventricular circular patch plasty) help patients with functional TR due to stenotic heart failure (15).

Rheumatic TR may be treated with open valvotomy or valve replacement. Valves compromised by infective endocarditis can simply be removed after an aggressive course of antibiotics. Valve replacement can be performed later if there is no recurrence of drug abuse in these patients. When acute TR is the result of RV infarction, the usual management of acute coro-

Diastolic Studies

The ECG is normal in mild pulmonic stenosis. With increasing severity, right ventricular hypertrophy and right atrial enlargement are common. The chest radiograph may reveal poststenotic dilatation of the main and left pulmonary arteries. Echocardiography is particularly useful in assessing the valve morphology, calculating pressure gradients across the pulmonic valve, grading the severity of stenosis, and evaluating right ventricular function. The presence of other congenital abnormalities, tricuspid regurgitation, and right atrial enlargement can also be ruled out by echocardiography. Cardiac catheterization provides confirmation of pressure gradients, full hemodynamic assessment, and identification of associated pulmonary artery branch stenosis. Pulmonary stenosis is graded based on the peak pressure gradient: mild, with a gradient of 25 to 49 mm Hg; moderate, 50 to 75 mm Hg; and severe, greater than 75 mm Hg. Surgical valvuloplasty is recommended for symptomatic patients and those with a peak gradient greater than 50 mm Hg. Surgical valvuloplasty is reserved for severe calcification, dysplasia, endocarditis, and previous valvuloplasty failure. Medical management includes infective endocarditis prophylaxis, treatment of right heart failure and atrial fibrillation, and anticoagulation to prevent thromboembolic complications.
Pathophysiology

In the absence of pulmonary hypertension, volume overload of the right ventricle is well tolerated. Decompensation with resulting right ventricular failure can occur when pulmonary hypertension develops from other causes.

Presentation

PI is usually an incidental auscultatory finding in patients admitted to the ICU for other reasons. Physical findings include the typical decrescendo diastolic murmur along the upper left sternal border. The intensity of the murmur does not correlate well with the severity of regurgitation.

Diagnostic Studies

The ECG is usually normal. The presence of right ventricular hypertrophy suggests pulmonary hypertension. The chest radiograph is normal in mild insufficiency. The pulmonary trunk may be prominent when the insufficiency is moderate to severe. Pulmonary hypertension may be present. Echocardiography with Doppler study can be useful for differentiating pulmonary from aortic insufficiency and for establishing right heart chamber sizes and associated abnormalities.

Therapeutic Considerations

Specific treatment is rarely required. However, therapy should be directed toward control of pulmonary hypertension when present. When the right heart fails, diuretics and sodium restriction are useful, and some clinicians suggest that cardiac glycosides are helpful. Surgical treatment (bioprosthetic valve replacement) is reserved for advanced right heart failure. In patients with a remote repair of tetralogy of Fallot and chronic PI, RV dilation has been linked to sudden death. This has led some clinician-investigators to pursue valve replacement in early stages of RV dilation.

Mixed Valve Lesions

Mitrail Stenosis with Regurgitation

The combination of pressure and volume overload on LA favors early development of symptoms, atrial fibrillation, and congestive heart failure. Because transvalvular gradients may overestimate the degree of stenosis, Doppler measurement of valve area should be considered. Decision making is complex, and intervention is often required before either of the lesions reaches a severe degree. Moderate MR is a contraindication for balloon valvotomy.

Aortic Stenosis and Regurgitation

This combination causes both pressure and volume overload on the LV. The predominant lesion is indicated by the size of the LV: a normal-sized, but hypertrophied, LV signifies predominant AS; a dilated LV suggests dominant AR. As with combined MS and MR, transvalvular gradients may overestimate AM, so planimetry or the continuity equation method should be considered. The threshold for surgery is lowered as compared to single valve-defect patients. Those with severe AS with accompanying AR should be operated on in higher calculated valve areas or in the presence of mild symptoms. Surgery in patients with predominant AR with accompanying AS can be delayed until symptoms develop or asymptomatic LV dysfunction becomes apparent on echocardiography (enlarged ventricular dimensions).

Mitrail Stenosis and Aortic Stenosis

This combination causes serial obstructions resulting in reduced cardiac output and early development of pulmonary venous congestion and hypertension. Low transvalvular gradients characterize this aortic stenosis because of low cardiac output. Mitrail valvotomy is done first, followed by aortic valve replacement as indicated.

Mitrail Stenosis and Aortic Regurgitation

This combination creates a challenge for the physician attempting to make a diagnosis. MS decreases the volume overload of AR, and AR attenuates antegrade mitral valve flow by increasing LV diastolic pressure, thereby decreasing transmitial gradients. Balloon mitral valvotomy followed by aortic valve replacement (AVR), as indicated, is a reasonable approach.

Mitrail Regurgitation and Aortic Stenosis

Aortic stenosis aggravates MR by increasing the afterload. MR, by its pressure release effect, obscures even severe AS. Systolic function remains normal with low transaortic gradients. If both lesions are severe, AVR with mitral valve repair or replacement is necessary. Moderate or mild MR may improve after AVR for AS, especially if there is no anatomic lesion in the mitral valve. Intraoperative TEE plays an important role in this decision.

Mitrail Regurgitation and Aortic Regurgitation

These lesions create additive volume loads on the LV; consequently, the sequela of dyspnea and LV dysfunction appear sooner.

Prosthetic Valve Dysfunction

Prosthetic valves in common use are broadly divided into mechanical, bioprosthetic, and homograft valves. St. Jude bileaflet valves are commonly used mechanical valves which need lifelong anticoagulation. Carpentier-Edwards and Hancock bioprosthetic valves are common tissue valves in use. They do not require long-term anticoagulation, but have a short life span and are prone to degenerative changes and failure. Acute valvular complications may result from infective endocarditis, paravalvular leak, valve ring abscess, thrombosis, pannus formation, degenerative calcification, lipid infiltration, dehiscence of the valve, and strut fracture.

Progressive congestive cardiac failure is a common presentation with stenosis and regurgitation. Acute, complete valvular obstruction may lead to sudden death in the absence of surgical intervention. Embolic phenomenon, hemolytic anemia—indicating a paravalvular leak, and a new aortoventricular block—indicating a valve ring abscess may be other presenting symptoms. Prosthetic valve thrombosis may present with non-specific cardiac symptoms. Normally functioning prosthetic valves are associated with clicks and murmurs; hence, disappearance of clicks or a new or changing murmur is important in making the diagnosis.

Echocardiography is essential to make a diagnosis in these patients. As previously noted, TEE is more sensitive and specific in the evaluation of prosthetic valve pathologies than TTE; echocardiography has replaced cardiac catheterization in these cases. Fluoroscopy may be needed in some cases to identify
the nature of the disease and assess the effects of thromboly-
sis. Excessive rocking motion of the valve ring or limited mo-
tion of the valve components due to thrombus or vegetation;
calcification; thickening around the valve due to an abscess; and
a pseudoaneurysm can be identified with two-dimensional
echocardiography. The color Doppler technique may show a
paravalvular leak, pseudoaneurysms, and/or fistula formation.
Calculation of transvalvular gradients help in the diagnosis of
prosthetic valve stenosis. Gradients depend on the type and
size of the valve and dynamic conditions such as cardiac out-
put, blood volume, heart rate, and contractility. Therefore,
it is recommended that the measurements be compared to the
control values obtained immediately after valve replace-
ment. It has also been suggested that it may be more appro-
priate to calculate the prosthetic valve area using the continuity
equation:

\[ \text{Area}_1 \times \text{velocity time integral}_1 = \text{Area}_2 \times \text{velocity time integral}_2 \]

Medical therapy is directed toward treatment of conges-
tive heart failure—diuretics, vasodilators, and inotropes; initi-
ation of antibiotics for infective endocarditis after obtaining
blood cultures; and thrombolysis for certain cases of pros-
thetic valve thrombosis. Staphylococcal organisms predom-
ninate in early (less than 60 days postplacement) prosthetic
valve endocarditis, whereas in late (greater than 60 days post-
placement) endocarditis, there are equal proportions of in-
fec tion caused by streptococcal and staphylococcal organ-
isms (16). Empiric antibiotics are started until culture results
and sensitivities are available. Fibrinolytic therapy is recom-
manded for right-sided thrombosis with a large, clot burden
or New York Heart Association class III to IV symptoms
(see http://www.abouthf.org/questions_articles.htm). Fibrino-
lysis for left-sided lesions is reserved for patients in whom emer-
surgery is high risk or contraindicated because this is
associated with a 12% to 15% risk of cerebral embolism (17).
Ultimately, all prosthetic valve lesions require valve replace-
ment surgery. Reoperative mortality is high in this patient
population.

**IMPORTANT CONSIDERATIONS IN THE TREATMENT OF RIGHT VENTRICULAR FAILURE SECONDARY TO VALVULAR HEART DISEASE**

Chronic RV failure secondary to VHD presents major thera-
pic challenges to the intensive care physian. These patients
are usually debilitated with low cardiac output and pulmonary,
hepatic, and renal dysfunction. Patients develop hepatic fail-
ure secondary to congestive hepatic cirrhosis. Ascites, malnu-
trition, reduced systemic vascular resistance, jaundice, coag-
ulopathy, and renal failure—the hepaticorenal syndrome—are
the manifestations of hepatic dysfunction. The management of
hepatoportal syndrome is challenging and will require invasive
monitoring and, often, renal replacement therapy.

Treatment of pulmonary hypertension in the critically ill
case with VHD decreases RV afterload and helps prevent
and decrease RV failure. This approach, combined with main-
tenance of adequate coronary perfusion pressure, forms the
mainstay of treatment of acute RV failure. Exacerbating fac-
tors of pulmonary hypertension, such as hypoxemia, hyper-
carbia, acidosis, hypothermia, and increased intrathoracic
pressure, should be corrected aggressively. Recent advances in
pharmacologic management of pulmonary hypertension with a wide va-
riety of options for selective pulmonary vasodilatation, with
studies favoring the use of inhaled prostaglandins and nitric
oxide (18). Inhaled pulmonary vasodilators are preferred over
intravenous agents because they do not decrease systemic blood
pressure and also do not increase shunt fraction. Many newer
drugs—including nitric oxide donors and phosphodiesterase
inhibitors—are promising and are under investigation.

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