Acute Valvular Heart Disease

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KEYWORDS

• Aortic regurgitation • Aortic stenosis • Mitral stenosis • Mitral regurgitation
• Prosthetic valve dysfunction

KEY POINTS

- A targeted history, physical examination, and basic initial workup can provide early recognition of decompensated valvular heart disease.
- Echocardiography is diagnostic in acute valvular disease in determining etiology and defining severity.
- Early hemodynamic assessment in valvular disease is key to appropriate initiation of medical therapy, which focuses on improving hemodynamics, peripheral perfusion, and relieving vascular congestion.
- Mechanical circulatory support has a role in the treatment of decompensated valvular disease.
- Prosthetic valve dysfunction can cause acute obstruction, regurgitation, and hemolytic anemia; these conditions should be easily recognizable and well-understood.

INTRODUCTION

Valvular heart disease (VHD) is a common phenomenon in the developed world, affecting a large proportion of adults to varying degrees of severity.[1,2] Although the majority of patients have stable valvular disease, with the increasing prevalence in the population, decompensated illness as a result of valvular disease is increasingly recognized. Owing to improved outcomes as the result of cardiac surgery and interventional therapies, the prognosis for VHD has improved over the past several decades. We aim to delineate the initial diagnosis and management of these patients and provide a context for clinicians to provide evidence-based, effective treatment modalities in acutely decompensated VHD.

INITIAL EVALUATION

Medical stabilization of decompensated VHD requires prompt recognition. Early evaluation of hemodynamics and the recognition of an initial appropriate evaluation including a targeted physical examination and history is important. Hypotension, pulmonary congestion, poor peripheral perfusion, altered mentation, and oliguria are signs concerning for cardiogenic shock and heart failure, which may be the result of acutely decompensated VHD. Assessment of respiratory status and anticipating a need for airway management with either intubation or noninvasive positive pressure ventilation is important. A history of intravenous drug use, ischemia, blunt cardiac injury, known VHD, or congenital valvular disease may provide clues regarding etiology. Cardiac auscultation may be helpful; murmurs can provide, in some cases, a clue to the severity of the disease process.

Several noninvasive tests can be undertaken while the patient is being stabilized. Laboratory work, including cardiac troponin and brain natriuretic peptide can provide data regarding volume status and evidence of myocardial ischemia. Chest radiography will confirm pulmonary

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congestion and electrocardiography (ECG) will allow for the evaluation of myocardial ischemia. Early echocardiography can evaluate the presence and severity of disease. For each valvular pathology, further therapy should be tailored as discussed in this article.

AORTIC REGURGITATION

Etiology and Pathophysiology

Acute aortic regurgitation (AR) is among the most dangerous acute valvular pathologies and can result in rapid clinical deterioration. Although AR can have several distinct etiologies (**Table 1**), acute AR is commonly the result of trauma, infective endocarditis, or thoracic aortic dissection extending retrograde toward the aorta.[3,4] Infective endocarditis can cause leaflet perforation and perivalvular abscesses resulting in acute AR. Aortic dissection can cause acute AR, either through direct retrograde extension into the leaflet or through dilation of the sinuses of Valsalva.[5] Direct blunt trauma to the chest may can also cause traumatic leaflet rupture.[5,6]

In chronic severe AR, the left ventricle (LV) has dilated and become compliant to accommodate excess LV end-systolic volume and can maintain a forward stroke volume (SV). However, in acute severe AR, the LV has not adapted to the large regurgitant volume and cannot generate an appropriate forward SV.[7,8] Ultimately, LV end-diastolic pressure exceeds that of the left atrium (LA), resulting in premature closure of the mitral valve during diastole and decreased LV filling, further decreasing SV and increasing left atrial pressure, leading to increased pulmonary pressures, low cardiac output, hypotension, and increased systemic vascular resistance.[7,8]

Clinical Presentation

Severe acute AR is often dramatic in its presentation. Owing to the acute decrease in cardiac output and high LV filling pressures, cardiovascular collapse and acute pulmonary edema are often seen. Preceding fever may be present in patients with endocarditis. Examination findings of profound cardiogenic shock—hypotension, pallor, cool extremities, altered mentation, and pulmonary edema—are common. In contrast, examination findings in chronic severe AR reflect, LV adaptation to the high regurgitant volume and increased SV causes various classic signs of higher output with widened pulse pressure (Duroziez's murmur, waterhammer pulse, widened pulse pressure). In acute severe AR, the pulse pressure will either be normal or decreased owing to rapid equalization of the aortic and LV pressures.[7] Premature mitral valve closure and LV volume overload may cause a soft S1 and S3, respectively. A low-pitched early diastolic murmur can be heard, but the presence of the murmur is not a reliable marker of severity owing to rapid equalization of pressures between the LV and aorta, causing a minimal gradient in diastole and thus minimal murmur. ECG findings usually reflect the underlying disorder, such as acute ST-elevation myocardial infarction (STEMI) from retrograde dissection of a coronary artery, as may occur in acute aortic dissection, or are nonspecific. Chest radiography may show pulmonary edema and pulmonary vascular prominence, along with cardiomegaly or widened mediastinum in patients with aortic dissection.

Diagnosis

Echocardiography is the diagnostic modality of choice in severe AR. The markers of severity include (1) a vena contracta (narrowest neck of the AR jet) width of greater than 6 mm, (2) more than 65% of the LVOT width occupied by the color jet, and (3) holodiastolic flow reversal within the abdominal aorta (indicating flow back through the aortic valve during diastole).[9] Doppler imaging may reveal a dense continuous wave signal with steep diastolic slope of AR velocity indicating rapid equalization of pressures. Premature closure of the mitral valve, suggested with a very short mitral valve deceleration time (<150 ms) may indirectly indicate the severity of the AR jet[9] (**Fig. 1**). Additionally, echocardiography can evaluate LV dysfunction and reveal valvular vegetations. Transesophageal echocardiography (TEE) or computed tomography angiography will reveal the presence of aortic dissection.[9]

Treatment

Acute AR with hemodynamic collapse is a surgical emergency. Stabilization of the patient should be focused on afterload reduction and supporting the volume overloaded ventricle in preparation for surgery. Intravenous vasodilators, such as nitroprusside in normotensive or hypertensive patients and inotropic agents in hypotensive patients to augment SV, are first-line therapies.[10] In general, although beta-blockers are the first line in treatment of patients with acute aortic dissection, they should be avoided in acute AR, because it can decrease SV.[10]

Mechanical circulatory support, including intra-aortic balloon counterpulsation, venoarterial extracorporeal membranous oxygen, or percutaneous ventricular assist devices such as the Impella (AbioMed, Danvers, MA), are contraindicated.[8,9,11]

Table 1
Valvular disease causes, diagnosis, and management

Valvular Pathology	Causes	Physical Examination Findings	Echocardiographic Findings	Medical/Mechanical Stabilization
Acute severe AR	Infective endocarditis Aortic dissection Rupture of congenitally fenestrated cusp Traumatic rupture Iatrogenic	Hypotension Narrow pulse pressure Low pitched, early diastolic murmur Cool, clammy extremities, altered mentation Soft S1 Pulmonary edema	Quantitation AR color Doppler jet >6 mm Jet width/LVOT >65% Dense CW Doppler with steep diastolic slope (PHT <200 ms) Premature mitral valve closure, short MV deceleration time (<150 ms) Supportive signs Holodiastolic flow reversal within the descending aorta	Aggressive afterload reduction with IV vasodilators Inotropes to augment stroke volume MCS, particularly IABP, is contraindicated
Acute severe MR	Ischemia Infective endocarditis Trauma Dynamic LVOT obstruction Iatrogenic	Hypotension Systolic murmur (absent in 50%), low pitched, ending before S2 Large "V" waves Cool, clammy extremities, altered mentation Pulmonary edema	Quantitation Vena contracta >0.7 cm EROA >0.40 cm^2 Regurgitant volume >50% Supportive signs PV flow reversal in systole Flail MV leaflet, ruptured papillary muscle MR color Doppler jet entrainment (Coanda effect)	Aggressive afterload reduction with IV vasodilators Inotropes IABP to decrease mean arterial pressure, improve coronary perfusion in ischemia Advanced MCS (VA-ECMO)

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Table 1
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Valvular Pathology	Causes	Physical Examination Findings	Echocardiographic Findings	Medical/Mechanical Stabilization
Severe aortic stenosis	Calcific Rheumatic Congenital Supravalvular stenosis Metabolic diseases SLE Alkaptonuria	Crescendo–decrescendo systolic murmur, mid-to-late peaking Soft S2 Pulsus parvus et tardus Cool, clammy extremities, altered mentation Pulmonary edema	Quantitation Calculated AVA <1.0 cm^2 by continuity or planimetry Mean gradient >40 mm Hg Peak AV velocity >4.0 m/s Supportive signs Severe calcification of the aortic valve leaflets with reduced excursion in systole Turbulent flow acceleration across the aortic valve in systole LV hypertrophy	Cautious afterload reduction in hypertension Cautious diuresis taking care to avoid preload reduction IABP to reduce afterload and as a bridge to intervention Percutaneous aortic balloon valvuloplasty in refractory hypotension and cardiogenic shock as a bridge to AVR
Severe mitral stenosis	Rheumatic heart disease Accelerated atherosclerotic disease	Rumbling diastolic murmur with "opening snap" Tachycardia Pulmonary edema	Quantitation MVA <1.5 cm^2 Increased mean transmitral gradient >10 mm Hg PHT across MV >220 ms Supportive signs Commissural fusion, rheumatic thickened mitral valve leaflets with reduced motion and "doming" in diastole Increased left atrial size	Diuresis to relieve vascular congestion Beta blockade to increase diastolic filling time Restoration of sinus rhythm Anticoagulation Percutaneous mitral balloon valvotomy in medication refractory patients

Abbreviations: AR, aortic regurgitation; AV, aortic valve; AVA, aortic valve area; AVR, aortic valve replacement; CW, continuous wave; EROA, effective regurgitant orifice area; IABP, intraaortic balloon pump; IV, intravenous; LV, left ventricular; LVOT, left ventricular outflow tract; MCS, mechanical circulatory support; MR, mitral regurgitation; MV, mitral valve; MVA, mitral valve area; PHT, pressure half-time; PV, pulmonary vein; SLE, systemic lupus erythematosus; VA-ECMO, venoarterial corporeal membranous oxygen.

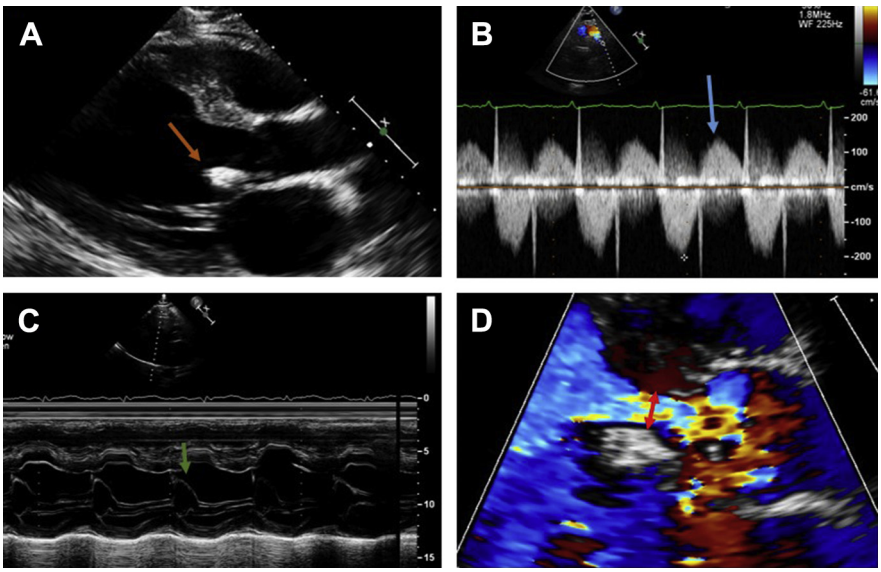

Fig. 1. Aortic regurgitation (AR). (*A*) Large vegetation (*arrow*) on the noncoronary cusp of the aortic valve (AV) causing severe AR. (*B*) Holodiastolic flow reversal in the descending aorta (*arrow*). (*C*) M-mode parasternal long axis showing early diastolic closure of the mitral valve (*arrow*) owing to the large AR jet. (*D*) Large vena contracta of the severely regurgitant AR jet (*arrow*).

The intraaortic balloon pump inflates during diastole, worsening AR severity, whereas both the Impella and venoarterial corporeal membranous oxygen increase LV afterload, also worsening severity of regurgitation and decreasing cardiac output.

Patients with lesser severity of AR who are hemodynamically stable can be treated initially with targeted antibiotic therapy and serial echocardiography to assess for LV dysfunction. In patients with stable acute severe AR, antibiotic therapy for valvular endocarditis may improve the severity of AR, but often valve replacement is still necessary. If LV dysfunction develops with an LV ejection fraction of less than 50% or the LV end-diastolic diameter exceeds 65 mm, then valve replacement should be more urgently considered.[8]

SEVERE MITRAL REGURGITATION

Etiology and Pathophysiology

Acute mitral regurgitation (MR) has numerous etiologies, ranging from endocarditis to myocardial ischemia to iatrogenic injury. Three primary mechanisms mediating the manner in which acute MR manifests are (1) rupture of the chordae tendineae from various causes, (2) tethering or rupture of the papillary muscles, classically owing to myocardial ischemia or "ischemic MR," and (3) MR in the setting of dynamic LV outflow tract obstruction and functional LV dilatation.[11,12]

In patients with acute coronary syndrome, as many as 14% have been shown to have at least mild MR and about 3% had moderate-to-severe MR; in the SHOCK trial registry (Should We Emergently Revascularize Occluded Coronaries for Cardiogenic Shock), 8% of patients with cardiogenic shock as the result of acute myocardial infarction had severe MR.[13] Across the spectrum of acute coronary syndrome, patients with moderate to severe MR have a much higher mortality. Papillary muscle rupture as the result of ischemia is seen across the spectrum of acute coronary syndrome, most commonly owing to dysfunction or rupture of the posterior papillary muscle, because the posterior papillary muscle does not have a dual blood supply.[14,15] Anterior ischemic papillary muscle rupture is exceedingly rare.[16]

In acute MR, the LA has not had time to adapt to the abrupt increase in volume of regurgitant blood, causing a marked increase in LA pressure and pulmonary edema. Owing to the high regurgitant volume, SV and cardiac output may decrease precipitously, leading to global hypoperfusion.[7]

Clinical Presentation

Severe acute MR usually presents as cardiovascular collapse, including signs of poor tissue perfusion and cardiogenic shock, as well as lung auscultation indicating pulmonary edema. With

increased right-sided filling pressures, large "v" waves can be seen in the jugular venous pulsation. A systolic murmur is often heard, but owing to a markedly diminished pressure gradient between the LA and LV, the murmur is often low pitched and ends well before S2. A large proportion of patients may have no murmur on auscultation, or "silent MR."[7,11] The low intensity of the murmur may also be due to low systemic blood pressures in patients with hemodynamic collapse, further decreasing the LV–LA gradient.

ECG findings are usually nonspecific, unless diagnostic of ischemia as the causative etiology. Chest radiography reveals the presence of pulmonary edema. A unique finding can be the presence of unilateral pulmonary edema, which is a function of the eccentricity of the mitral regurgitant jet preferentially through a single pulmonary vein; this finding has been shown to be independently associated with an increased risk of mortality.

Diagnosis

Echocardiography is essential in diagnosing acute MR and can differentiate the etiology. The severity of the MR jet can be graded using color flow, but in acute MR owing to rapid equalization of LA–LV pressures and inadequate color flow visualization, the severity of the jet can actually be underestimated.[17] In this instance, TEE can provide improve visualization of the severity of MR and objective calculation of the effective regurgitant orifice area that, when greater than 0.40 cm^2, suggests severe MR. A jet vena contracta of greater than 0.7 cm is considered severe, as is the presence of an entrainment effect of the MR jet or "Coanda" hugging the wall of the LA in systole.[17] Systolic pulmonary venous flow reversal, evidence of ischemia with regional wall motion abnormalities, and signs of leaflet perforation or vegetation on the mitral valve can also be seen. TEE can localize flail leaflets well as better delineate the various scallops on the anterior and posterior mitral leaflets[17] (**Fig. 2**). This is particularly helpful in planning for surgical intervention.

When suspicion for acute MR is high but not readily seen using echocardiography, left and right heart catheterization may be useful if indicated. The presence of giant "v" waves on the pulmonary capillary wedge pressure tracing may indicate the presence of severe MR; the accuracy and specificity of this finding is not clear, but may be helpful in the appropriate clinical context.[18]

Treatment

Medical stabilization of acute severe MR depends on the patient's hemodynamic status. Hypertensive or normotensive patients benefit from afterload reduction with intravenous vasodilator therapy. The initial goal should be to reduce diastolic blood pressure and mean arterial blood pressure, because this will decrease the regurgitant mitral valve orifice size and increase cardiac output.[19] Hypotensive patients benefit from inotropes, particularly dobutamine and milrinone, because they do not typically vasoconstrict the peripheral vasculature. Diuretics may alleviate pulmonary edema.

An intraaortic balloon pump in patients with acute severe MR may decrease ventricular workload and allow for improved perfusion in cardiogenic shock.[11] Venoarterial extracorporeal membranous oxygen or Impella in acute MR is sparsely described, except in case reports,[20] although theoretically these devices may improve cardiac output and hemodynamics and, in patients with ruptured or ischemic papillary muscles, the impeller in an Impella can further damage the subvalvular apparatus and, thus, is relatively contraindicated.[21]

The timing of coronary angiography is dictated by the urgency of surgical intervention—emergent surgery should not be delayed for coronary

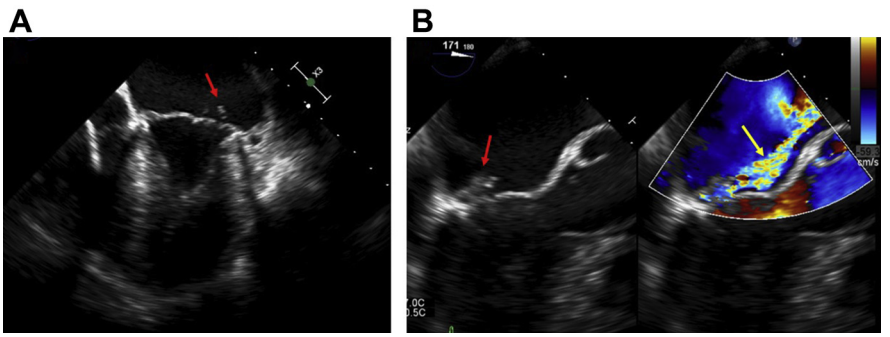

Fig. 2. Transesophageal echocardiogram (TEE). TEE with (*A*) flail P2 scallop of the mitral valve leaflet (*arrows*) with (*B*) resultant severe, eccentric anteriorly directed mitral regurgitation (MR) with entrainment/Coanda effect (*arrow*).

angiography unless concurrent ischemia (particularly STEMI) is thought to be the causative culprit, because revascularization may improve the degree of MR severity in these cases. Otherwise hemodynamically stable patients may benefit from preoperative coronary angiography to delineate the coronary anatomy and need for bypass surgery concurrently with mitral valve replacement.

Surgical Timing

The clinical severity of acute MR differentiates the urgency of surgery. Owing to the acuity of presentation, mortality is high.[22,23] Signs and symptoms of heart failure or echocardiographic evidence of hemodynamic compromise require prompt surgery.[11] Papillary muscle rupture of ischemic MR requires surgical repair and portends a worse outcome, with surgical repair improving mortality to that of a similar patient without rupture.[24,25]

Reperfusion therapy through primary percutaneous coronary intervention of the infarct-related artery is always indicated in patients with STEMI, including those with ischemic MR. Primary percutaneous coronary intervention in patients with STEMI has been found to decrease the incidence of ischemic MR,[26,27] and ischemic MR after STEMI is associated with greater short-term and long-term mortality.[28] In stable patients without papillary muscle rupture, the need for and timing of performing surgical repair for ischemic MR is controversial.[29,30]

Severe MR as the result of endocarditis, in addition to treatment with antibiotics, should be referred for urgent surgical intervention, particularly with persistent hemodynamic compromise or heart failure.[11]

SEVERE DECOMPENSATED AORTIC STENOSIS

Etiology and Pathophysiology

Aortic stenosis (AS) is one of the most common valvular diseases in the population, and the incidence increases with age.[31] Senile calcific AS and congenital AS (including bicuspid aortic valve) are the most common in the United States; rheumatic stenosis is quite uncommon. Calcific AS most commonly presents within the sixth to eighth decades of life in patients with additional comorbidities. Symptomatic severe AS classically presents as either syncope, angina, or heart failure; there is a 50% mortality rate associated with these presentations within 5, 3, and 1 years of onset, respectively.[32]

Severe AS causes the LV to eject against a fixed obstruction increasing LV end-diastolic pressure, leading to compensatory LV hypertrophy. Over time, this impairs LV filling and leads to diastolic dysfunction, decreased exercise tolerance, and worsening dyspnea on exertion owing to an inability of the LV to augment cardiac output during exercise.[7] Systolic dysfunction is rare and often a late finding. Increased myocardial oxygen demand, compression of the coronary arteries, and reduced coronary perfusion time during diastole causes angina.[7,31] Syncope can result from systemic vasodilation with an inability to augment cardiac output during exercise or ventricular arrhythmias.[33] Once symptoms develop, mortality is high and the only cure is valve replacement.[7,32]

Clinical Presentation

Management of decompensated acute severe AS is guided by an assessment of the hemodynamic status. A wide range of presentations from decreased functional capacity to cardiogenic shock can be seen. Cardiac auscultation will reveal a late-peaking crescendo–decrescendo systolic murmur, reduced (or absent) S2 intensity owing to decreased aortic valve leaflet mobility, and a delayed and weak carotid upstroke ("pulsus parvus et tardus").[7]

ECG may show signs of LV hypertrophy, but otherwise findings are often nonspecific and may suggest active myocardial ischemia owing to subendocardial ischemia. Chest radiography will show pulmonary edema in patients with significant heart failure.

Echocardiography is essential in the diagnosis of severe AS. The severity of AS is best shown by (1) evidence of high pressure gradients across the aortic valve of greater than 40 mm Hg, (2) a calculated aortic valve area of less than 1.0 cm^2 or aortic valve area index to body surface area of less than 0.60 cm^2/m^2, and (3) the anatomy of the aortic valve[34] (**Fig. 3**). A bicuspid aortic valve is seen in 1% to 2% of the population, and patients with bicuspid aortic valve develop sclerosis and stenosis of the aortic valve at a much younger age owing to increased hemodynamic turbulence across the valve.[7,35] The presence of LV hypertrophy can be a clue to long-standing AS.

In patients with discordant echocardiography findings and symptoms or if echocardiography is nondiagnostic, the mean gradient across the aortic valve and the valve area can be directly measured through cardiac catheterization.[31] This assessment is accomplished in the cardiac catheterization laboratory by obtaining arterial access and placing a catheter across the aortic valve, measuring simultaneous pressures within the LV and the aorta. Risks of this procedure include a small stroke risk through dislodgement of calcium on the aorta as well as ventricular arrhythmias.

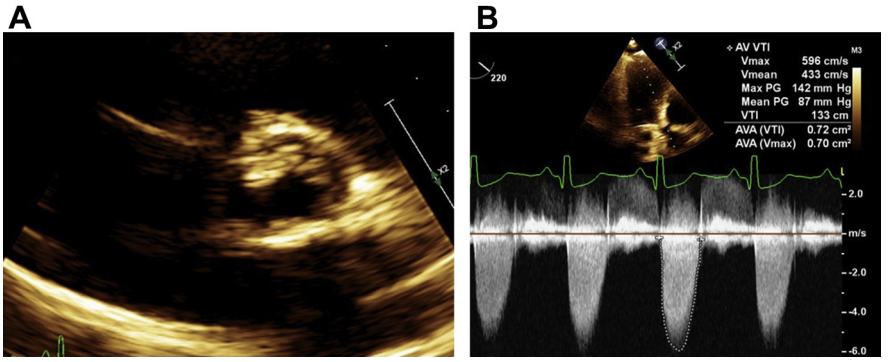

Fig. 3. Severe aortic stenosis (AS). Severe AS with (*A*) Bicuspid, severely calcified aortic valve leaflets. (*B*) Extremely elevated gradients across the valve using continuous wave Doppler with maximum velocity of approximately 6 m/s.

Treatment

Medical stabilization of severe AS is complicated by the presence of a fixed afterload owing to obstruction, and thus patients may be preload dependent. Vasodilators run the risk of decreasing systemic blood pressure and reducing coronary perfusion, diuretics may decrease preload and therefore cardiac output, and inotropes may worsen myocardial ischemia when present. Therefore, medical therapy should be undertaken in a closely monitored clinical setting and is targeted to treating symptoms and stabilizing hemodynamics.[11] Diuretics to reduce pulmonary congestion and vasodilators to treat hypertension, usually low doses of angiotensin-converting enzyme inhibitors, are appropriate.[11]

In patients with atrial arrhythmias, a rhythm control strategy involving restoration of normal sinus rhythm is important as atrial dysrhythmias and loss of "atrial kick" can worsen LV dysfunction.

In critically ill patients with poor perfusion, initial stabilization is preferred from a surgical outcome standpoint, but few medical therapies provide any mortality benefit. In intensive care settings, nitroprusside in patients with decompensated severe AS and low ejection fraction increases cardiac output and decreases systemic vascular resistance, which may improve outcomes.[36,37] Inotropes in patients with hypotension and decreased cardiac output can augment forward flow, but the effect may be limited given the fixed nature of the obstruction in severe AS.

Percutaneous aortic balloon valvuloplasty (PABV) can serve as a "bridge" to surgery or transcatheter aortic valve replacement (TAVR) by reducing the mean aortic valve gradient and increasing the effective aortic valve orifice area in critically ill patients.[38] PABV, however, is not a benign procedure and has a number of feared complications, including stroke from dislodgement of calcium during balloon positioning and the development of acute AR as a result of "cracking" open the valve.[38] Rarely, cases have been described where "rescue TAVR" can be used in development of severe AR. Bicuspid aortic valves are not amenable to PABV and moderate or severe AR is a contraindication to PABV.

Surgery

Severe symptomatic AS is a class I indication for surgical replacement and surgery should not be delayed unnecessarily, but can be complicated by hemodynamic status in AS with heart failure or cardiogenic shock.[11] TAVR is being increasingly used as a modality for valve replacement, is noninferior to surgery in high-risk and intermediate-risk patients, and is superior to medical therapy with respect to mortality, although it is associated with a higher degree of vascular access complications.[39,40]

ACUTELY DECOMPENSATED SEVERE MITRAL STENOSIS

Etiology and Pathophysiology

Mitral stenosis (MS) is a rare disease in the developed world owing to early treatment of *Streptococcus pyogenes* infections, the late sequelae of which can lead to autoantibodies against myocardial tissue resulting in rheumatic fever. Chronic inflammation of the MV leads to commissural adhesion, thickening, and fibrosis causing immobility. Although rare in the United States, it is not uncommon to see clinical MS in areas with high immigrant populations. Owing to streptococcal pharyngitis being a disease of young age, many patients present at a younger age and pregnancy in particular poses a distinct issue.

In chronic MS, the tight MV orifice leads to high left atrial pressures and subsequent high pulmonary arterial pressures and right heart pressures. Overcoming the LA–LV gradient during diastole

causes ventricular dependence on diastolic filling time and atrial contraction. Decompensation of MV disease occurs in volume overloaded states (such as pregnancy) causing high filling pressures or with tachycardia and reduced diastolic filling time. Arrhythmias are common in this population owing to distension and pressure overload of the LA, which serves as a nidus for atrial fibrillation. Loss of atrial systole and an acute increase in left atrial pressure causes pulmonary edema. Likewise, rapid atrial rates reduce diastolic filling time.

Clinical Presentation

Classic physical examination findings include a diastolic murmur, "opening snap" after S2, with increased delay of opening snap indicating worsened severity of MS, implying higher LA pressures required to overcome resistance caused by the stenotic MV. Lung findings and chest radiography with pulmonary edema may also be present. ECG may show left atrial enlargement, but is otherwise usually benign.

Echocardiography will show "hockey stick" doming appearance of the anterior mitral leaflet in the parasternal long-axis view with associated valvular and chordal/subvalvular calcification. Gradients across the mitral valve are acquired with Doppler echocardiography. Generally, a mitral valve area of less than 1.5 cm^2 is considered severe; MS with an area of greater than 1.5 cm^2 is considered not hemodynamically significant enough to cause severe symptoms.

Treatment

Medical therapy in patients with MS is geared toward eventual intervention, usually either percutaneous mitral balloon valvuloplasty (PMBV) or mitral valve replacement if PMBV is not available. Diuretics should be used to reduce pulmonary edema. Prolonging diastole will allow the LV to fill more and reduce left atrial pressure; thus, using beta-blockers to target a lower heart rate and decrease transmitral gradient can lower the mean pulmonary artery pressure. Cardioselective beta-blockers, particularly in pregnant patients, prevent action on the myometrial tissue.[41,42] Ivabradine, a sodium channel blocker with heart rate–lowering effects, may be effective.[43]

The treatment of atrial arrhythmias should be acted on promptly. In nonvalvular atrial fibrillation, a rate control strategy is likely noninferior to rhythm control; however, in severe MS, a rhythm control strategy, much like in AS, will restore atrial systole and improve symptoms. Atrial fibrillation present for less than 24 to 48 hours can undergo prompt cardioversion, but if it has been present longer, TEE should be performed first to rule out left atrial appendage thrombus. Anticoagulation for atrial fibrillation stroke risk in severe MS should always be with warfarin with goal International Normalized Ratio of 2.0 to 3.0, because studies with newer anti-Xa inhibitors (rivaroxaban, edoxaban, apixaban) typically exclude valvular atrial fibrillation.

Percutaneous valvotomy is rarely performed in emergency circumstances, but is useful in patients who have a favorable anatomy and severe MS by valve area (<1.5 cm^2) or very severe MS (<1.0 cm^2) and are on appropriate maximal medical therapy.[11,44] Concomitant significant MR is considered a contraindication to PMBV. In pregnancy, owing to physiologic hypervolemia and tachycardia, there is a lower threshold to pursue PMBV prepartum to reduce the physiologic burden on the heart associated with the stress of labor, and is known to be safe.[45,46]

ACUTE PROSTHETIC VALVE DYSFUNCTION

Owing to the increased incidence of valvular disease and cardiac surgery for valvular pathology, as well as increased longevity of these patients, the incidence of complications in patients with prosthetic valves has also increased over time.

Valve Thrombosis, Thromboembolic Events, and Obstruction

The formation of thrombus on a prosthetic valve can predispose to an increased risk of stroke or peripheral embolic events. Because the risk is highest in the initial first 3 months after valve replacement, empiric anticoagulation is implemented after bioprosthetic valve replacement. Mechanical valves are significantly more prone to thrombosis compared with bioprosthetic valves, necessitating lifelong anticoagulation with warfarin. Although subclinical thrombosis is likely common, clinical thrombosis may present in a variety of manners, from subtle symptoms such as increased fatigue and exertional dyspnea to sudden cardiac death and cardiogenic shock. A subtherapeutic International Normalized Ratio in these patients should necessitate immediate evaluation for possible prosthetic valve thrombosis. Embolic events (acute stroke, peripheral organ emboli) may result from valve leaflet thrombosis and in the correct clinical context this should be further investigated.

Echocardiography and Doppler interrogation of the affected prosthetic valve can diagnose obstruction.[47] Significantly increased gradients from baseline should prompt concern for valve obstruction, particularly over a shorter time period.

High SV states can cause increased transvalvular gradients, particularly across the aortic valve, and should be taken into account. Minimally symptomatic patients still require further workup if there is continued concern; mitral prosthetic valve thrombosis may be better visualized with TEE, as well as monoleaflet or Bjork-Shiley aortic valves. Multidetector computed tomography better characterizes prosthetic valve leaflet motion and can differentiate thrombus from pannus formation.[48] Fluoroscopy in the cardiac catheterization laboratory is an alternative way to visualize the leaflet motion of the valve.

Patients with left-sided valve thrombosis or obstruction causing significant heart failure symptoms (New York Heart Association functional class III or IV) should be referred for surgical intervention, urgently if the thrombus is mobile or large.[11] Smaller thrombi can be treated medically with high-dose anticoagulation, and should thrombus persist despite this measure, fibrinolytic therapy is recommended, followed by warfarin and aspirin therapy. Should this treatment fail, repeat surgical intervention is recommended.[11] Although overall outcomes between fibrinolytic therapy and surgery are comparable,[49] fewer thromboembolic and bleeding complications are seen in surgery, as well as lower rates of rethrombosis.[50]

Pannus formation, calcification, endocarditis, and leaflet fibrosis of bioprosthetic valves can also cause valvular obstruction. TEE is particularly helpful in the evaluation of bioprosthetic valves and can differentiate thrombus and pannus based on the degree of echodensity.[47]

Acute Valvular Regurgitation

Prosthetic valve regurgitation may be either "paravalvular," around the struts of implantation, or central or transvalvular, within the orifice of the valve. Normally functioning bioprosthetic prosthetic valves may have mild transvalvular regurgitation early on, which resolves with time, but paravalvular leak is uncommon and can indicate dehiscence.[47] Mechanical valves often have builtin physiologic regurgitation as "backflow" to prevent the formation of thrombi and can be seen as multiple regurgitant jets[51]; as with bioprosthetic valves, paravalvular regurgitation is abnormal.[47] Percutaneously implanted valves often have paravalvular regurgitation; an increased severity of paravalvular leak can portend worse outcomes.[52] Clinically, patients typically present with heart failure symptoms and a change in the prosthetic valve sound. Jaundice as the result of hemolytic anemia may be present.[7]

Echocardiography can localize and quantify regurgitation severity. TEE and 3-dimensional echocardiography can better delineate the severity of regurgitation.[47,53] Dehiscence of the prosthetic valve causes a "rocking" motion and can be visualized with both TTE and TEE. Multidetector computed tomography can also be used to evaluate these patients.[47,54]

Initial medical therapy for heart failure symptoms should occur in preparation for surgery for severe regurgitation. Asymptomatic and symptomatic patients with severe bioprosthetic or mechanical valve regurgitation should be referred for cardiac surgery. In high-risk patients with severely regurgitant bioprosthetic aortic valves, transcatheter aortic valve implantation or "valve-in-valve" TAVR can be considered, because this modality has been shown to have fairly high procedural success.[55,56]

Hemolytic Anemia

In general, macroangiopathic hemolytic anemia as the result of prosthetic valves is rare, especially with newer generations of prosthetic valves. Older bileaflet and ball valves undergo structural deterioration over time that may result in rough surfaces leading to hemolytic anemia.[57] Clinical signs of heart failure, dark urine, and jaundice are often seen in these patients, as well as a high-intensity murmur. Dark urine, high serum lactate dehydrogenase, and schistocytes on peripheral smear can be indicative of the microangiopathic process. These patients are medically managed with red cell transfusions, iron, and erythropoietin if necessary, but in extreme cases reoperation for the affected valve may be necessary.[11]

SUMMARY

Numerous acute valvular pathologies, owing to the high prevalence of VHD and patients who have had prior valve surgery, are seen in clinical practice. Clinicians should be able to quickly recognize when valvular disease is present and recognize the various potential etiologies, in particular ischemia, trauma, and infection. Initial management of the hemodynamically decompensated patient should target stability before surgical or interventional treatment. Echocardiography, both transthoracic and transesophageal, is the initial diagnostic modality for these patients. Other imaging modalities are used as clinically indicated.

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