CHAPTER

25

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Decompensated Valvular Disease in the Cardiac Care Unit

The prevalence of valvular heart disease is strongly influenced by the patient's age. In the United States and other developed countries, significant valvular disease tends to cluster at age extremes; in the young it results primarily from congenital heart disease (CHD) and in the elderly from degenerative wear-and-tear valve changes.

The combined prevalence of moderate and severe CHD is approximately 5 in 1,000 live births.[1] The prevalence of congenital valvular disease is much lower as the bulk of the CHD is accounted for by nonvalvular lesions such as ventricular and atrial septal defects, and patent ductus arteriosus. The prevalence of valvular disease among the elderly ≥75 years of age is approximately 13.3%. This is in contrast to the prevalence rate of 0.7% in the 18- to 44-years age group.[2] Between the age extremes, valvular disease is encountered mostly in the survivors of CHD or in immigrants with rheumatic valvular disease.

Thus a patient in an adult coronary care unit (CCU) in the United States usually falls into one of the following three categories: elderly patients, patients with CHD, and immigrants with rheumatic heart disease. The last two groups often include pregnant women.

Valvular disease presents either as valvular stenosis or valvular regurgitation. When valvular disease is the primary reason for a CCU admission, the patient usually presents with severe degrees of stenosis, regurgitation, or a combination thereof. Among the elderly CCU patients typical valvular disorders include aortic stenosis (AS) and mitral regurgitation; in addition, there is often secondary tricuspid regurgitation. In younger CCU patients, typical valvular disorders include rheumatic mitral stenosis, tricuspid valve endocarditis, and severe pulmonic insufficiency arising as a long-term complication of CHD surgeries performed in childhood.

In general, medical therapy of severe valvular disease is limited to symptom relief and hemodynamic stabilization in preparation for definitive treatment performed by interventional cardiologists and cardiac surgeons. Unfortunately, there is a paucity of solid randomized trial data to support any form of treatment of valvular disease.[3] In the latest joint guidelines of the American Heart Association and the American College of Cardiology, most of the recommendations for the management of valvular disease are based on either nonrandomized trials or expert opinions.[4]

ANTIBIOTIC PROPHYLAXIS OF VALVULAR HEART DISEASE

Routine antibiotic prophylaxis in no longer recommended for acquired valvular disease in a nontransplanted heart unless there is a prior history of endocarditis. For congenital forms of valvular disease in the setting of surgically repaired CHD, antibiotic prophylaxis is recommended only if (1) there is prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure, and (2) there are residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization).[5]

AORTIC STENOSIS

ETIOLOGY AND PATHOPHYSIOLOGY

AS in a typical adult CCU patient arises from calcific degeneration of either a previously normal trileaflet aortic valve (TAV) or a congenitally bicuspid aortic valve (BAV). Calcification process is often enhanced by disorders of perturbed calcium phosphate metabolism such as end-stage renal disease and Paget's disease.[6]

Calcific TAV stenosis is typically encountered among the elderly and calcific BAV stenosis in middle-aged patients (Figure 25.1). It is estimated that AS is present in 2% of all Americans, 65 years of age or older.[2] TAV stenosis may be a manifestation of generalized atherosclerosis. BAV is the commonest congenital heart defect occurring in approximately 1% to 2% of all live births.[7] BAV may present as stenosis or regurgitation and may be associated with aortopathies such as aortic aneurysm, aortic dissection, and coarctation.[8]

The major cardiovascular adaptation to pressure overload caused by AS is left ventricular hypertrophy (LVH). Because of LVH the left ventricular chamber becomes smaller and its wall thicker; both these changes lower the wall stress and allow for preservation of left ventricular ejection fraction (LVEF) for very long periods. When LVEF starts to decrease it is often due to an afterload-LVH mismatch (not enough LVH for the degree of AS) rather than cardiomyopathy.

CLINICAL PRESENTATION

AS is usually asymptomatic unless severe. Patients with severe AS typically present with angina, syncope, sudden death, or heart failure. Physical diagnosis may establish the diagnosis of AS but is often incapable of precisely grading AS. Classic auscultatory findings of AS include a systolic crescendo–decrescendo ejection murmur over the precordium that radiates to the neck, and an occasional absence of the valve closing S2 sound.[9] Carotid upstroke is often weak and delayed (pulsus parvus & tardus).

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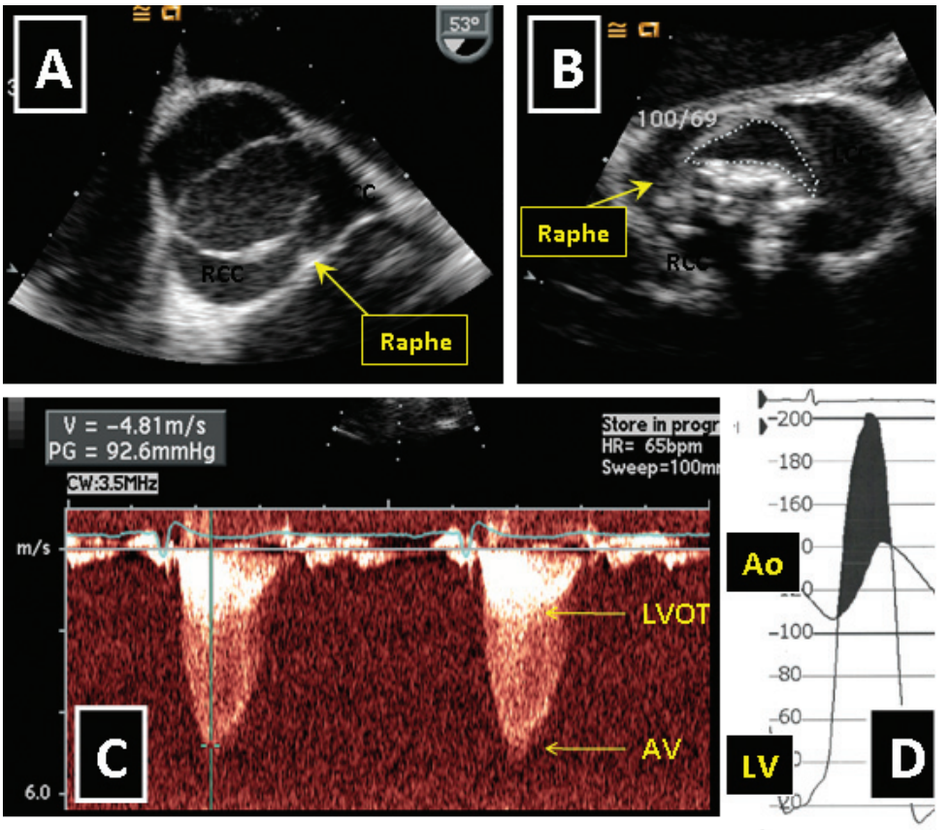

Figure 25.1. Aortic stenosis. **Panel A** and **Panel B** show bicuspid aortic valve on transesophageal echocardiography. In **Panel A**, the bicuspid valve is not significantly stenotic. This is in contrast to **Panel B** which shows a severely calcified and markedly stenotic bicuspid aortic valve. **Panel C** shows continuous wave Doppler recording of blood flow across the aortic valve in systole. Note the wide separation between the normal left ventricular outflow tract (LVOT) velocity and the markedly elevated velocity across the aortic valve (AV). **Panel D** shows simultaneous pressure recordings from the left ventricle (LV) and the aortic root (Ao). Note the wide separation between the aortic pressure and the markedly elevated LV pressure.

DIAGNOSIS

Electrocardiogram (EKG) often shows signs of LVH and left atrial enlargement. Chest X-ray (CXR) rarely provides specific evidence of AS; occasionally aortic valve calcifications are seen on CXR.

Echocardiography is the primary means of diagnosing and grading AS. Aortic valve area (AVA) is the primary criterion of AS. When the flow across the aortic valve is not diminished, the magnitude of the peak and mean gradient across the aortic valve in systole is inversely related to AVA (Table 25.1). Other typical echocardiographic findings of AS include LVH, left atrial enlargement, and often a normal LVEF.

A low transvalvular gradient (mean gradient ≤30 mm Hg) does not exclude severe AS. Patients with AS may have low transvalvular gradients for two reasons: (1) Afterload-hypertrophy mismatch and (2) concomitant cardiomyopathy (such as ischemic heart disease). Differentiating between these two groups is extremely important as the patients in the first group will benefit from aortic valve surgery, whereas those in the second group may not.

Differentiation between the two groups is made after measuring aortic valve parameters (AVA and the gradients) at rest and following intravenous infusion of increasing amounts of dobutamine (starting at 5 μg/kg/minute and escalating up to 20 μg/kg/minute).[10] Aortic valve parameters are usually measured by echocardiography (modified dobutamine stress echo). However, these measurements may also be performed during cardiac catheterization.

Changes in the following three parameters are measured during dobutamine stress testing in patients with low-gradient AS: left ventricular stroke volume, AVA, and the mean gradient. If there is an increase in stroke volume of ≤20%, the patient likely has severe cardiomyopathy and is usually not a candidate for aortic valve surgery. If the stroke volume increases by 20% or more than two scenarios are possible: (1) AVA remains essentially the same but the mean gradient increases above 30 mm Hg, and (2) AVA increases by 0.2 cm^2 or more but the gradient remains essentially unchanged. The patients in the first group have true severe AS and will benefit from aortic valve surgery. The patients in the second group have pseudosevere AS in the setting of left ventricular cardiomyopathy; they are unlikely to benefit from aortic valve surgery (Table 25.2).

For routine diagnosis of AS cardiac catheterization is usually not necessary. However, coronary angiography is commonly performed in patients with AS who are scheduled to undergo

TABLE 25.1 Grades of Aortic Stenosis

	Valve Area	*Valve Area Indexed For Body Surface Area*	*Peak Velocity*	*Peak Gradient*	*Mean Gradient*
	cm^2	*cm^2/m^2*	*m/sec*	*mm Hg*	*mm Hg*
Normal	2.0-4.0		< 2.5	< 25	
Mild Aortic Stenosis	> 1.5		2.5-3.0	25-36	< 25
Moderate Aortic Stenosis	1.0-1.5		3.1-4.0	37-64	25-40
Severe Aortic Stenosis	< 1.0	< 0.6	> 4.0	>64	> 40

aortic valve surgery as coronary artery disease is common in the typical age group of patients with AS. When comparing aortic gradients obtained by cardiac catheterization, it is important to emphasize that it is the mean but not the peak aortic gradient that correlates well with echocardiography.

MEDICAL THERAPY

There are no proven medical therapies for prevention or treatment of AS. Antibiotic prophylaxis of bacterial endocarditis in patients with AS is no longer recommended; for details see section on Antibiotic Prophylaxis of Valvular Heart Disease in this chapter's introduction.

Patients with severe AS often present to CCU with clinical signs of heart failure. Diuretics, angiotensin converting enzyme (ACE) inhibitors, and digitalis may be used with caution. Although historically the use of intravenous vasodilators was contraindicated in patients with severe AS, a small study in CCU patients have demonstrated that intravenous nitroprusside actually relieves symptoms of heart failure in patients with severe AS and severely reduced left ventricular systolic function.[11] In that study, intravenous nitroprusside was started at a mean dose of 14 ± 10 μg per minute, and the dose was increased to a mean of 103 ± 67 μg per minute at 6 hours and 128 ± 96 μg per minute at 24 hours.

SURGICAL AND PERCUTANEOUS INTERVENTIONAL THERAPY

AS is a mechanical problem that requires a mechanical solution. Surgical aortic valve replacement (AVR) is currently the preferred therapeutic choice in patients with AS as it improves symptoms and survival. Major indications for AVR are listed in Table 25.3.

Percutaneous interventions for AS include aortic valve balloon valvuloplasty and percutaneous AV replacement. Balloon valvuloplasty is an effective form of therapy for congenital AS. However, balloon valvuloplasty in calcific TAV or BAV aortic stenosis has been rather ineffective (valve area seldom increases above 1.0 cm^2 and there is a high rate of restenosis), and it has significant morbidity and mortality. At present, typical indications for balloon valvuloplasty in calcific AS include temporary relief of AS in patient undergoing noncardiac surgery (such as hip replacement) or in those with terminal illness (such as cancer).

Percutaneous valve replacement is a new and promising therapy for AS. Although the first percutaneous implantation of an aortic prosthetic valve has been reported in 2002, such a procedure is still considered experimental in the United States and has been performed only within an approved clinical trial.[12]

IMPACT ON PREGNANCY

Patients with calcific AS are seldom of child-bearing age. In patients of child-bearing age, AS is usually caused by noncalcific congenital abnormalities (such as unicuspid aortic valve). Young patients with moderate-to-severe AS should be advised against conception until AS is relieved. Those who nonetheless become pregnant may or may not develop severe symptoms. Pregnant patients with AS and mild symptoms may be managed conservatively during pregnancy with bed rest, oxygen, and β-blockers. Pregnant patients with severe symptoms may need percutaneous or even surgical intervention. These procedures carry significant risk to both the mother and the unborn child.

AORTIC REGURGITATION

ETIOLOGY AND PATHOPHYSIOLOGY

Aortic regurgitation (AR) may be due to disorders of the aortic root or the aortic valve leaflets. BAV, ascending aortic aneurysm, aortic dissection, and endocarditis are common

TABLE 25.2 Dobutamine Stress Test: Differentiating True From Pseudo Aortic Stenosis

	Stroke Volume Change			
	< 20%		*> 20%*	
	< 0.2 cm^2	**Valve Area Change**	< 0.2 cm^2	> 0.2 cm^2
	Remains < 30 mm	**Mean Gradient**	Becomes > 30 mm Hg	Remains < 30 mm
Conclusion	Severe LV dysfunction		True severe AS	Pseudo-severe AS
AVR Recommendation	No		Yes	No

Abbreviations: AS, aortic stenosis; AVR, aortic valve replacement

TABLE 25.3 Major Indication for Aortic Valve Surgery in Patients with Aortic Stenosis

	Severe AS	*Moderate AS*
Symptomatic	Class I	Patient with moderate AS are usually not symptomatic
Asymptomatic AND one of the following:		
LVEF < 50%	Class I	No AVR
Patient referred for CABG or other heart surgery	Class I	Class IIa
Patient referred for surgery of the aorta	Class I	Class IIa

causes of AR. Conditions that predispose patient to aortopathies and AR include systemic hypertension and connective tissue disorders (such as Marfan syndrome, Ehlers–Danlos syndrome, and ankylosing spondylitis). In the less developed countries, rheumatic heart disease and tertiary syphilis remain important causes of AR.

In AR, a regurgitant volume flows back into the ventricle through the aortic valve during diastole. There it joins the systemic volume that had entered the left ventricle through the mitral valve. During subsequent systole the two volumes leave the aortic valve together; the systemic volume continues into the aorta and its branches, whereas the regurgitant volume flows back into the ventricle. The cycle then repeats itself. Subsequent pathophysiology depends on whether AR is chronic or acute.

In chronic AR, there is a progressive enlargement of the left ventricle to accommodate the combined systemic stroke volume and the regurgitant volume. This remodeling often prevents significant elevation of left heart pressures. It may take years if not decades for patients with chronic AR to develop congestive heart failure owing to progressive left ventricular systolic dysfunction.

This is in sharp contrast to acute AR where there is sudden volume overload of a nondilated left ventricle, marked elevation of left ventricular and left atrial pressures, life-threatening pulmonary edema, and even cardiogenic shock.

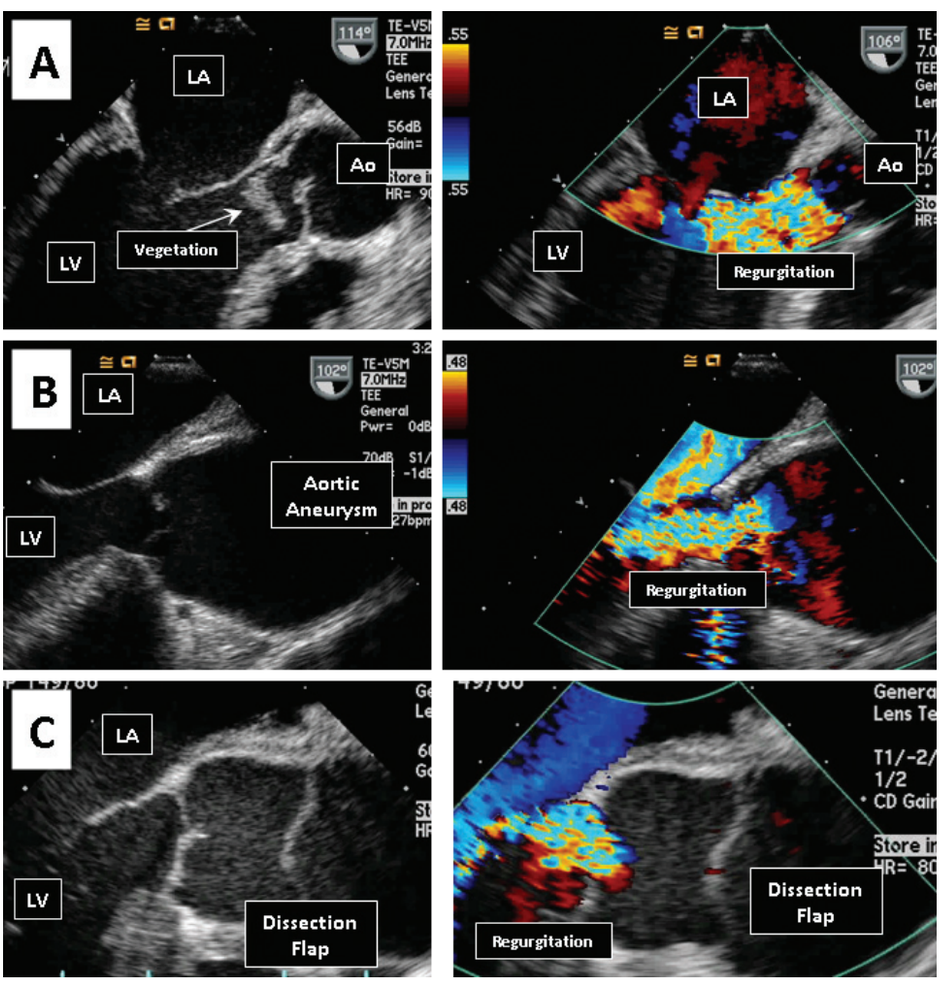

Figure 25.2. Aortic regurgitation. Causes of severe aortic regurgitation include aortic valve endocarditis **(Panel A)**, ascending aortic aneurysm **(Panel B),** and type A aortic dissection **(Panel C)**. LA, left atrium; LV, left ventricle; Ao, ascending aorta.

TABLE 25.4 Grades of Aortic Regurgitation

	Mild	*Moderate*	*Moderate-Severe*	*Severe*
	(1+)	(2+)	(3+)	(4+)
Regurgitant orifice area (cm^2)	<0.1	0.10-0.19	0.20-0.29	>0.3
Regurgitant Fraction	<30%	30-39%	40-49%	>50%
Regurgitant Volume (mL)	30	30-44	45-59	>60
Vena contracta (cm)	<0.3	0.3 - 0.6	>0.6	

Although severe acute AR represents only a minority of AR cases, its often fulminant and life-threatening course necessitates CCU admission. The rest of the AR section will be devoted to severe acute AR.

CLINICAL PRESENTATION

Among the most common causes of severe acute AR are chest trauma, bacterial endocarditis, and the dissection of the ascending aorta (type A aortic dissection). Patients with severe acute AR frequently present with fulminant pulmonary edema and cardiogenic shock. Patient with endocarditis will also have general signs and symptoms of a systemic bacterial illness. Patient with acute type A aortic dissection will usually complain of severe chest pain, often in the setting of uncontrolled systemic hypertension.

DIAGNOSIS

In severe acute AR, there may be few or no auscultatory findings of AR per se; the diastolic murmur of AR is often soft, short, or even absent because there is rapid equilibration of aortic and left ventricular pressures during diastole. There is usually marked tachycardia and S3 gallop. Wide pulse pressure—the hallmark of severe chronic AR—is routinely absent in severe acute AR.

CXR in severe acute AR frequently shows signs of pulmonary congestion. EKG may show tachycardia; there may also be signs of myocardial ischemia (owing to high myocardial demand brought on by very elevated left ventricular pressures or when acute aortic dissection extends into the coronary artery ostia). When endocarditis leads to periaortic valve abscess, EKG may demonstrate varying degrees of atrioventricular conduction block.

Transthoracic and transesophageal echocardiography are the primary means of evaluating AR. Echocardiography may establish the etiology, mechanism, and severity of MR (Figure 25.2). The diagnostic criteria for grading the severity of AR are listed in Table 25.4.

MEDICAL THERAPY

Severe acute AR is a life-threatening medical emergency that necessitates highest levels of CCU care. Endotracheal intubation, oxygen administration, and diuretic therapy are used to treat pulmonary edema. Afterload reduction may be achieved with the use of intravenous vasodilators (such as nitroprusside). Disease-specific therapies, if available, should also be administered (such as antibiotic therapy for endocarditis).

SURGICAL AND PERCUTANEOUS INTERVENTIONAL THERAPY

At present, there are no effective percutaneous interventions for the treatment of AR. Furthermore, intra-aortic balloon pump (IABP) is absolutely contraindicated when significant AR is present. Diastolic counterpulsations—which are the hallmarks of IABP function—worsen AR. In severe acute AR, aortic valve surgery should be performed as soon as possible especially in cases of type A aortic dissection.

IMPACT ON PREGNANCY

Severe AR, whether acute or chronic, is one of the valvular heart lesions that may be associated with high maternal and/or fetal risk during pregnancy. Pregnant women with AR who have New York Heart Association functional class III–IV symptoms, severe pulmonary hypertension (pulmonary pressure >75% of systemic pressures), and/or LV systolic dysfunction are at particular risk for maternal and fetal complications. The same is true for pregnant women having Marfan syndrome with or without AR.

MITRAL STENOSIS

ETIOLOGY AND PATHOPHYSIOLOGY

Rheumatic heart disease is by far the most frequent cause of acquired mitral stenosis (MS). Acquired narrowing of the mitral valve may also be caused by mitral annular calcifications or cardiac tumors such as myxomas (Figure 25.3). However, these nonrheumatic etiologies seldom cause severe MS. Congenital causes of MS are rare (1% of all MS patients) and include obstructive membranes either immediately proximal to the mitral orifice (supravalvular mitral ring) or within the left atrium (cor triatriatum).

Rheumatic heart disease may be conceptualized as a bacteria-triggered autoimmune inflammatory disorder that leads to progressive lifelong valve damage. The rheumatic process starts as acute rheumatic fever usually in childhood following pharyngitis caused by group A β-hemolytic streptococci ("strep throat"). As streptococcal proteins share antigenic properties with certain connective tissue proteins in the human host, the immune response that is mounted against the streptococci also leads to valvular damage.

Rheumatic mitral valve disease is characterized by leaflet thickening, commissural fusion, chordal fusion, and shortening as well as valve calcifications. MS is the primary manifestation of rheumatic heart disease. Women are more affected than men;

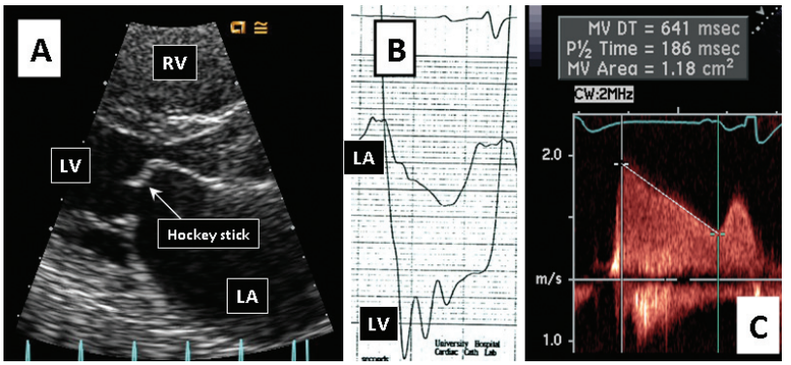

Figure 25.3. Mitral stenosis. **Panel A** shows a 2D transthoracic echocardiogram in the parasternal long-axis view of a patient with rheumatic mitral valve disease. The anterior mitral leaflet has the characteristic hockey stick appearance. **Panel B** represents simultaneous left atrial and left ventricular pressure recordings during cardiac catheterization. Note the marked separation between left ventricular diastolic pressure and the markedly elevated left atrial pressures indicative of severe mitral stenosis. **Panel C** demonstrates the echocardiographic pressure half-time method of determining the mitral valve area in a patient with moderate mitral stenosis. LA, left atrium; LV, left ventricle; RV, right ventricle.

the ratio is approximately 2:1 in favor of women. Rheumatic heart disease is still prevalent worldwide except in the developed countries where most cases are seen in immigrant populations.

Normal mitral valve area in an adult measures approximately 4 to 6 cm^2; this is approximately the area of an American or Canadian 25-cent coin. It takes decades for the valve area decreases to approximately 2.5 cm^2 and for the patient to start developing symptoms. Over the subsequent decades, the mitral valve area diminishes and the diastolic gradient across the mitral valve increases (Table 25.5). Left atrial (LA) enlargement, atrial arrhythmias, and systemic thromboembolism are major complications of rheumatic MS.

CLINICAL PRESENTATION

Dyspnea and other signs and symptoms of congestive heart failure owing to elevated left atrial and pulmonary pressures are the primary manifestations of MS. The degree of symptoms is directly related to the transvalvular mitral gradient in diastole. In turn, the magnitude of the mitral gradient is the result of interplay between the mitral valve area and the blood flow. Doubling the blood flow will quadruple the gradient.

Thus a patient with any degree of MS may become symptomatic if the blood flow is substantially increased such as during exercise, pregnancy, hyperthyroid crisis, fever, or tachyarrhythmia (such as atrial fibrillation). Left atrial enlargement is the primary cardiac adaptation to MS; left ventricular systolic function is typically normal. Atrial fibrillation, left atrial thrombus formation, and systemic thromboembolism (such as strokes) are important contributors to morbidity and mortality of rheumatic MS.

TABLE 25.5 Grades of Mitral Stenosis

	Valve Area	*Mean Gradient*	*Pulmonary Artery Systolic Pressure*
	cm^2	*mm Hg*	*mm Hg*
Normal	4.0-6.0		
Mild Mitral Stenosis	> 1.5	< 5	< 30
Moderate Mitral Stenosis	1.0-1.5	5-10	30-50
Severe Mitral Stenosis	< 1.0	> 10	> 50

These criteria are applicable when the heart rate is between 60 and 90 beats per minute.

DIAGNOSIS

Classic auscultatory findings of MS include loud first heart sound (S_1), an opening snap (OS) after the second heart sound (S_2), and a diastolic rumble. The duration of the S_2–OS interval is inversely related to the severity of MS (a shorter interval suggests more severe MS). In patients with normal sinus rhythm, there is also an end-diastolic ("presystolic") accentuation of the rumble.

EKG may demonstrate signs of LA enlargement characterized by wide, saddle-shaped P wave in leads I and II (so-called P *mitrale*) as well as late, deep P-wave inversion in lead V1. Atrial fibrillation is frequently present and there may be signs of right ventricular hypertrophy.

Chest X-ray usually demonstrates LA enlargement with straightening of the left cardiac silhouette. Right ventricular enlargement, signs of pulmonary venous congestion, and mitral valve calcification are frequently observed.[13]

Echocardiography is the primary means of diagnosing MS. By echocardiography one can determine the morphology of the mitral valve, its cross-sectional area, and the mean diastolic transvalvular pressure gradient. In addition, the size and function of cardiac chambers as well as the pulmonary artery pressures can be measured. Cardiac catheterization is not necessary for the diagnosis of MS in most instances.

MEDICAL THERAPY

Patients with MS often present to CCU with clinical signs of heart failure, frequently in the setting of atrial fibrillation. The goal of medical therapy in MS is to alleviate symptoms and prevent systemic thromboembolism. Congestion is treated with diuretics and the heart rate is controlled with β-blockers, certain calcium channel blockers (such as verapamil and diltiazem), and digitalis. Patients are initially anticoagulated with heparin then transitioned to warfarin. Specific therapies for atrial fibrillation may also be necessary.

TABLE 25.6 Major Indications For Percutaneous Mitral Balloon Valvuloplasty (PMBV)

General Prerequisites For PMBV
Moderate or severe mitral stenosis
Valve morphology favorable for PMBV
Absence of left atrial thrombus
Absence of moderate or severe mitral regurgitation
Appropriate Candidates
Symptomatic patients
Asymptomatic patients with pulmonary hypertension (PASP > 50 mm Hg at rest; > 60 mm Hg with exertion)
Patients with new-onset atrial fibrillation

When appropriate, patients should receive antibiotics for prevention of rheumatic fever recurrence according to the national guidelines.[4] In contrast, antibiotic prophylaxis of bacterial endocarditis in patients with MS is no longer recommended; for details see section on Antibiotic Prophylaxis of Valvular Heart Disease in this chapter's introduction.

SURGICAL AND PERCUTANEOUS INTERVENTIONAL THERAPY

MS is a mechanical problem that requires a mechanical solution. The 10-year survival of patients with MS receiving only medical therapy is unfavorable (50% to 60% overall and below 15% once significant limiting symptoms develop).[4]

When the morphology of the mitral valve is deemed favorable on echocardiography, percutaneous mitral balloon valvuloplasty (PMBV) is the preferred form of invasive treatment of MS. Major indications for PMBV are given in Table 25.6. If PMBV is unavailable or unfeasible, appropriate patients are referred for surgical intervention that may entail surgical commissurotomy, valve repair, or mitral valve replacement.

IMPACT ON PREGNANCY

Often pregnancy is the first time that a patient with MS becomes symptomatic because intravascular volume, cardiac output, and heart rate increase physiologically during pregnancy. PMBV can be performed during pregnancy as it has low risk of complications to the mother or the fetus.

MITRAL REGURGITATION

ETIOLOGY AND PATHOPHYSIOLOGY

According to the French cardiac surgeon Carpentier, all causes of mitral regurgitation (MR) fall into one of the following three categories: (1) *Annular dilatation* (as seen in dilated cardiomyopathy); (2) *excessive leaflet motion* (as seen in mitral valve prolapse, papillary muscle rupture, or endocarditis); and (3) *restricted leaflet motion* (as seen in rheumatic valve disease and ischemic cardiomyopathy).[14]

In MR, blood exits the left ventricle both antegrade—through the left ventricular outflow tract (systemic stroke volume) and retrograde—through the mitral valve (regurgitant volume). During diastole the regurgitant volume meets in the left atrium the systemic volume returning through the pulmonary veins. The combined volume then enters the left ventricle through the mitral valve. The process leads to volume overload of the left heart.

In chronic MR, there is progressive enlargement of the left atrium and the left ventricle to accommodate the combined systemic stroke volume and regurgitant volume. This remodeling often prevents significant elevation of left heart pressures; consequently it may take years if not decades for the patient to develop congestive heart failure (owing to progressive left ventricular systolic dysfunction) and atrial fibrillation (owing to left atrial enlargement).

This is in sharp contrast to acute MR where there is sudden volume overload of nondilated left heart chambers, marked elevation of left atrial pressures, life-threatening pulmonary edema, and even cardiogenic shock.

It is estimated that there are approximately 2.5 million patients with moderate-to-severe or severe MR in the United States at present.[15] Although acute MR represents only a minority of these cases, every health care professional working in the CCU setting should become proficient in diagnosing and managing this often life-threatening form of MR. The remainder of this section will be devoted to severe acute MR.

CLINICAL PRESENTATION

The leading causes of acute MR are bacterial endocarditis, papillary muscle rupture (traumatic or following myocardial infarction), and chordal rupture in the setting of preexisting myxomatous valve degeneration and mitral valve prolapse.

Irrespective of the cause, patients with severe acute MR frequently present with fulminant pulmonary edema and cardiogenic shock. Patient with endocarditis will present with general signs and symptoms of a systemic bacterial illness. Nontraumatic papillary muscle rupture is a mechanical complication that usually occurs 3 to 5 days after acute myocardial infarction. Posteromedial papillary muscle (which usually has solitary blood supply from either the right coronary or the left circumflex artery) ruptures more frequently that the anterolateral one (which is usually supplied by both the left anterior descending and circumflex arteries).

DIAGNOSIS

In severe acute MR, there may be few or no auscultatory findings of MR per se; the systolic murmur is often soft, short, or even absent because there is rapid equilibration of left ventricular and left atrial pressures during systole. There is frequently tachycardia and S_3 gallop.

CXR in severe acute MR routinely shows signs of pulmonary congestion. EKG may show tachycardia; there may also be signs of myocardial infarction when acute MR is caused by papillary muscle rupture.

Measurement from a pulmonary artery catheter—which is often placed at bedside in the CCU—usually reveals a marked elevation of the systolic V wave in the pulmonary artery wedge pressure tracings (Figure 25.4). Cardiac output is often low.

Transthoracic and transesophageal echocardiography are the primary means of evaluating MR. Echocardiography can

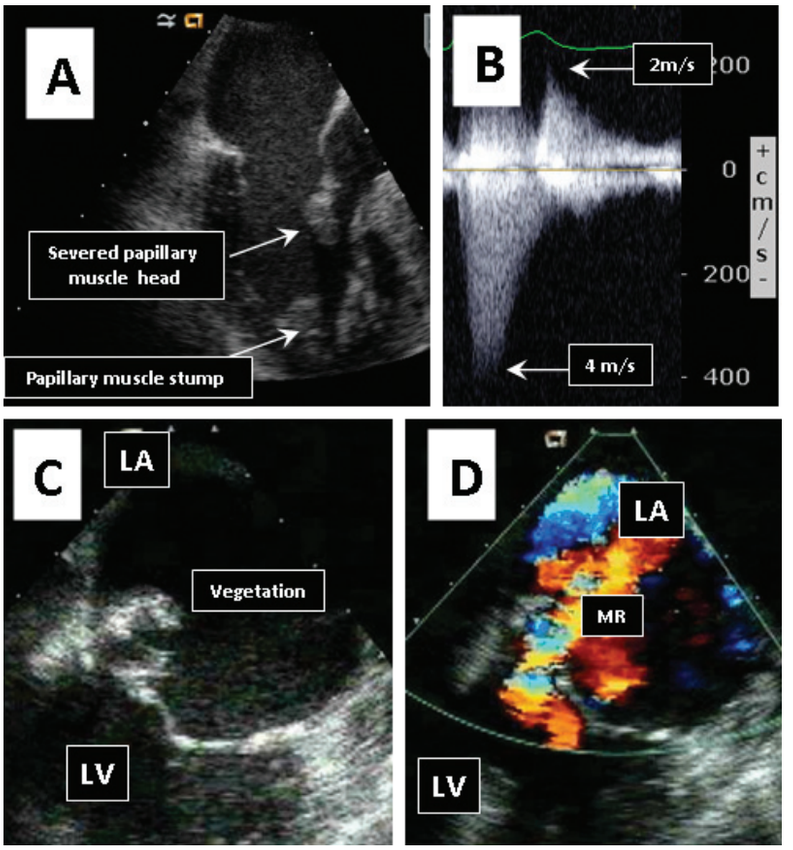

Figure 25.4. Mitral regurgitation. **Panel A** demonstrates anterior papillary muscle rupture in a patient with recent myocardial infarction. **Panel B** shows spectral Doppler tracing in a patient with severe acute mitral regurgitation. Note the relatively low peak velocity of the mitral regurgitant jet (4 m per second in this patient compared with approximately 5 m per second in normal people) indicative of systemic hypotension. The jet also decelerates rapidly towards the baseline indicative of rapid equilibration between LV and LA pressures toward the end of systole. Note also the relatively high antegrade velocity (2 m per second in this patient; normal is approximately 1 m per second) indicative of severe mitral regurgitation and a large regurgitant volume. **Panels C and D** show transesophageal echocardiographic findings in a patient with *Staphylococcus aureus* mitral valve endocarditis. **Panel C** is a 2D image showing large vegetation on the atrial side of the anterior mitral leaflet. **Panel D** demonstrates a very large regurgitant jet originating from the perforated anterior mitral leaflet. LA, left atrium; LV, left ventricle; MR, mitral regurgitation.

establish the etiology, the mechanism, and severity of MR. The diagnostic criteria for grading the severity of MR are listed in Table 25.7.

TABLE 25.7 Grades of Mitral Regurgitation

	Mild	*Moderate*	*Moderate-Severe*	*Severe*
	(1+)	*(2+)*	*(3+)*	*(4+)*
Regurgitant orifice area (cm^2)	<0.2	0.20-0.29	0.30-0.39	>0.4
Regurgitant Fraction	<30%	30-39%	40-49%	>50%
Regurgitant Volume (mL)	30	30-44	45-59	>60
Vena contracta (cm)	<0.3	0.3 - 0.7		>0.7

Note: Vena contracta is the narrowest portion of the regurgitant jet.

MEDICAL THERAPY

Severe acute MR is a life-threatening medical emergency that requires highest levels of care in the CCU. Endotracheal intubation, oxygen administration, and diuretic therapy are used to treat pulmonary edema. Afterload reduction may be achieved with the use of intravenous vasodilators (such as nitroprusside). Disease-specific therapies, if available, should also be administered (such as coronary revascularization and anti-ischemic medical therapy).

SURGICAL AND PERCUTANEOUS INTERVENTIONAL THERAPY

Severe acute MR often requires percutaneous insertion of the IABP, which is threaded through the femoral artery into the descending thoracic aorta with its tip just distal to the origin of the left subclavian artery. Significant coexisting aortic regurgitation is a contraindication for IABP insertion. IABP and the medical therapies described above are usually only palliative; the patient frequently requires urgent surgery to repair or replace the mitral valve.

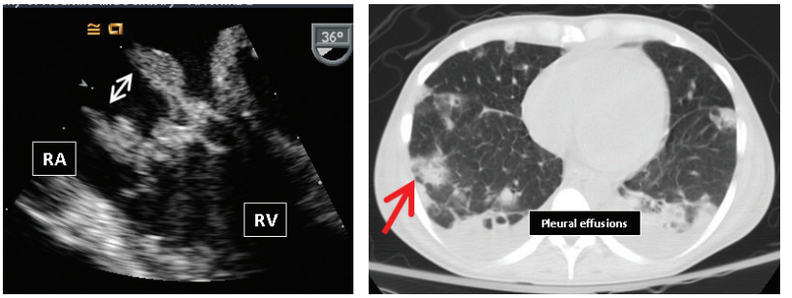

Figure 25.5. Tricuspid regurgitation. **Panel A** shows very large vegetation on the atrial side of the tricuspid valve on transesophageal echocardiogram in an intravenous drug user having *Staphylococcus aureus* bacteremia. **Panel B** shows septic emboli to the lung *(arrow)* in the same patient. RA, right atrium; RV, right ventricle.

IMPACT ON PREGNANCY

Severe MR, whether acute or chronic, is one of the valvular heart lesions that may be associated with high maternal and/or fetal risk during pregnancy. Pregnant women with MR who have New York Heart Association functional class III–IV symptoms, severe pulmonary hypertension (pulmonary pressure >75% of systemic pressures), and/or LV systolic dysfunction are at particular risk for maternal and fetal complications.

TRICUSPID REGURGITATION

ETIOLOGY AND PATHOPHYSIOLOGY

Tricuspid regurgitation (TR) is very prevalent. It is estimated that at present there are approximately 1.6 million individuals in the United States who have at least moderate-to-severe TR.[15] There are numerous causes of severe TR ranging from right ventricular dilatation to congenital leaflet abnormalities (such as Ebstein's anomaly),[16] trauma, carcinoid tumor and endocarditis. TR is frequently a complication of left heart disorders (such as left ventricular dysfunction, mitral and aortic valve disease, etc.).

Severe TR may be chronic or acute. The impact of severe chronic TR on the right heart is equivalent to the impact of severe MR on the left heart (see above). Among the most common causes of severe acute TR are chest trauma and infective endocarditis (often in the setting of intravenous drug use, central venous catheterization, or pacemaker and defibrillator lead placement).

CLINICAL PRESENTATION

Severe TR presents with jugular venous distension, hepatomegaly, ascites, and peripheral edema. Unlike severe acute MR or AR, severe acute TR is usually not a medical emergency.

DIAGNOSIS

In severe chronic TR there is a systolic murmur that augments with inspiration. Such a murmur is often soft or absent in severe acute TR owing to rapid equalization of right ventricular and right atrial pressures. EKG frequently reveals tachycardia.

CXR may reveal coin lesions in the lungs indicative of pulmonary septic emboli. Transthoracic and transesophageal echocardiography are the primary means of evaluating TR. Echocardiography can establish the etiology, mechanism, and severity of TR (Figure 25.5).

MEDICAL THERAPY

Appropriate antibiotic therapy should be administered to patients with tricuspid valve endocarditis. Disease-specific therapies should be administered whenever possible.

SURGICAL AND PERCUTANEOUS INTERVENTIONAL THERAPY

Tricuspid valve is not essential for life; in the past it was a frequent practice to treat tricuspid endocarditis with surgical excision of the tricuspid leaflet. Nowadays, the tricuspid valve should be surgically repaired whenever possible. If valve replacement is required, a bioprosthesis is the preferred option because of high rate of thromboembolic complications with mechanical tricuspid valve prostheses.

IMPACT ON PREGNANCY

Isolated severe TR regurgitation, unless acute, usually does not present a significant problem during pregnancy.

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PATIENTS AND FAMILY INFORMATION FOR:

Decompensated Valvular Disease

GENERAL CONCEPTS OF VALVULAR HEART DISEASE

The heart has four valves: two on the left side (called mitral and aortic valves) and two on the right side (called tricuspid and pulmonic valves). Normally, the blood flows across the valves without problem and only in forward direction. Normal valves prevent the backflow of the blood through the heart.

Valve disease occurs when the valves become either narrowed or leaky. When a valve is narrowed, the blood has trouble crossing it. When the valve is narrowed the doctors called that condition stenosis. When a valve is leaky, the blood flows backwards, something that should not happen in a healthy heart. When the valve is leaky the doctors call that condition regurgitation or insufficiency.

AORTIC STENOSIS

WHAT IS MY ILLNESS?

One of the valves in your heart called the aortic valve is narrowed; the doctors call this condition aortic stenosis. The aortic valve connects the main pumping chamber of the heart to the main blood vessel that leaves the heart called the aorta. Because of the narrowing, the blood has trouble crossing the aortic valve; this exerts strain on the main pumping chamber of the heart. This is similar to what happens when a highway lane is closed and all the cars now have to go through the remaining lanes; a traffic jam develops.

The narrowing gets worse over time; it usually takes many years for the narrowing to become severe. As the narrowing of the aortic valve gets worse, the walls of the main pumping chamber get thicker but the pumping action of the heart chamber usually remains normal. When the narrowing is very severe, you may experience the following symptoms: chest pain, shortness of breath, or you may pass out. In rare instances you may die suddenly.

HOW WILL I BE TREATED?

Medications may help relieve your symptoms but they cannot cure the narrowing. Once the aortic valve is severely narrowed and you experience above symptoms, the only solution is to have the valve replaced. This requires open heart surgery. Depending on your age and other factors, the surgeon will replace the narrowed valve with either a metal prosthesis or a tissue prosthesis. The advantage of a metal prosthesis is that can last for the rest of your life. The downside of a metal prosthesis is that you have to take a pill for blood thinning for the rest of your life. A tissue prosthesis is made of either pig or cow tissue and looks quite like your valve. If you receive a tissue prosthesis you will not need to take blood thinners. However, a tissue prosthesis usually lasts about only 10 years or so.

WHAT IF I AM PREGNANT OR THINKING OF BECOMING PREGNANT?

If you have severe narrowing of the aortic valve you may need to delay pregnancy until the narrowing is relieved—usually through the use of balloon catheter inserted through you groin, threaded to your heart, and blown up inside the valve to make it larger. If you are already pregnant and you have symptoms (such as shortness of breath or chest pain) you may need to take certain medications that are effective for you but not harmful to the child. If your symptoms during the pregnancy are severe you may need the balloon catheter treatment or even open heart surgery. These procedures carry significant risk to both you and your unborn child.

AORTIC REGURGITATION

WHAT IS MY ILLNESS?

One of the valves in your heart called the aortic valve is leaky; the doctors call this condition aortic regurgitation or aortic insufficiency. The aortic valve connects the main pumping chamber of the heart to the main blood vessel that leaves the heart called the aorta. Because of the leaky valve, with each heartbeat some amount of blood goes from the heart into the aorta (as it should). Unfortunately some amount of blood in you also leaks back from the aorta into the heart. This puts strain on the main pumping chamber of the heart that has to pump much harder. A heart with a leaky valve is like a leaky pail of water. If you carry a leaky pail you will not be able to carry as much water as if you were carrying a pail with no hole in it.

The valve leak may develop over many years or may develop suddenly. If the leak develops over time, you heart has time to adapt. It gets bigger and bigger but over time may weaken. It is more dangerous if the leak develops suddenly. When the leak is sudden and severe, you experience shortness of breath, swelling of your legs, and other symptoms. Sudden and severe leakage is life-threatening and in that instance you will most likely need to be in an intensive care unit.

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HOW WILL I BE TREATED?

Medications may help relieve your symptoms but they cannot cure the leak. If your leak is sudden and severe, you will most likely need to undergo open heart surgery to have the valve repaired or replaced by the surgeon. If the surgeon cannot repair the valve, he or she will replace the valve with a prosthesis.

Depending on your age and other factors, the surgeon will use either a metal prosthesis or a tissue prosthesis. The advantage of a metal prosthesis is that can last for the rest of your life. The downside of a metal prosthesis is that you have to take a pill for blood thinning for the rest of your life. A tissue prosthesis is made of either pig or cow tissue and looks a lot like your valve. If you receive a tissue prosthesis you will not need to take blood thinners. However, a tissue prosthesis usually lasts about only 10 years or so.

WHAT IF I AM PREGNANT OR THINKING OF BECOMING PREGNANT?

Severe leakage of the aortic valve may not be well tolerated during pregnancy and the condition may harm the unborn child. Women with severe leakage who are considering pregnancy may need to delay it until the condition is treated. If you are already pregnant, you may experience significant heart problems during your pregnancy.

MITRAL STENOSIS

WHAT IS MY ILLNESS?

When the mitral valve is narrowed, the doctors call this condition mitral stenosis. The mitral valve connects the left upper chamber of the heart (called the left atrium) to the main pumping chamber of the heart (called the left ventricle). The upper chamber is like an entryway into the main room.

Because of the narrowing, the blood has trouble crossing the mitral valve and filling the main pumping chamber of the heart. This exerts strain on the left upper chamber of the heart. This is similar to what happens when a highway lane is closed and all the cars now have to go through the remaining lanes; a traffic jam develops.

The narrowing gets worse over time; it usually takes many years for the narrowing to become severe. As the narrowing of the mitral valve gets worse, the left upper chamber gets bigger and the pressures in the lungs get higher. As the upper chamber gets larger, it loses its ability to conduct electricity correctly. This results in abnormal heartbeat called atrial fibrillation. This abnormal heartbeat makes the heart work inefficiently and also allows for blood clots to form inside the heart (something that does not happens in healthy hearts). The clot may shatter and send pieces travelling through the blood stream. If the pieces are big enough they may clog smaller blood vessels throughout the body; it is particularly bad when they end up in the brain where they will cause a stroke. If you develop this abnormal heartbeat you might need to take blood thinners for the rest of your life.

When the narrowing is very severe, you may experience the following symptoms: shortness of breath, difficulty in walking upstairs and uphill as well as swelling of your legs.

HOW WILL I BE TREATED?

Medications may help relieve your symptoms but they cannot cure the narrowing. Once the mitral valve is significantly narrowed and you experience above symptoms, and the only solution is to open or replace the valve. In many patients, the narrowed mitral valve can be opened by small balloons mounted on plastic tubes called catheters. You will first undergo ultrasound imaging of the heart. If the ultrasound doctor thinks that you are a good candidate, you will be sent to the cath lab. There a special cardiologist will insert the catheter into your veins, thread it to you heart, make a small hole between the right and the left upper chambers of your heart to bring the catheter above the narrowed valve. In the next step, the balloon will be passed across the mitral valve and blown up. This procedure, when successful, can make the narrowed orifice much bigger if not normal.

If you are not a candidate for the balloon procedure, you may need open heart surgery. Depending on your age and other factors, the surgeon will replace the narrowed valve with either a metal prosthesis or a tissue prosthesis.

WHAT IF I AM PREGNANT OR THINKING OF BECOMING PREGNANT?

If you have severe narrowing of the mitral valve you may need to delay pregnancy until the narrowing is relieved—usually through the use of balloon catheter inserted through you groin, threaded to your heart, and blown up inside the valve to make it larger. If you are already pregnant and you have symptoms (such as shortness of breath or chest pain) you may need to take certain medications that are effective for you but not harmful to the child. If your symptoms during the pregnancy are severe you may need the balloon catheter treatment or even open heart surgery. These procedures carry significant risk to both you and your unborn child.

MITRAL REGURGITATION

WHAT IS MY ILLNESS?

When the mitral valve is leaky, the doctors call this condition mitral regurgitation or mitral insufficiency. The mitral valve connects the left atrium to the left ventricle. The atrium is like an entryway into the main room.

As the atrium gets larger, it loses its ability to conduct electricity correctly. This may result in abnormal heartbeat called atrial fibrillation. This abnormal heartbeat makes the heart work inefficiently and also allows for blood clots to form inside the heart (something that does not happens in healthy hearts). The clot may shatter and send pieces travelling through the blood stream. If the pieces are big enough they may clog smaller blood vessels throughout the body causing strokes. If you develop this abnormal heartbeat you might need to take blood thinners for the rest of your life.

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Because of the leaky valve, the blood flows backwards into the atrium of the heart. This puts strain on the main pumping chamber of the heart which has to pump much harder. A heart with a leaky valve is like a leaky pail of water. If you carry a leaky pail you will not be able to carry as much water as if you were carrying a pail with no hole in it.

The valve leak may develop over many years or may develop suddenly. If the leak develops over time, you heart has time to adapt. It grows bigger and over time may weaken. It is much more dangerous if the leak develops suddenly. When the leak is sudden and severe, you experience shortness of breath, swelling of your legs, and other symptoms.

HOW WILL I BE TREATED?

Medications may help relieve your symptoms but they cannot cure the leak. If your leak is sudden and severe, you will most likely need to undergo open heart surgery to have the valve repaired or replaced by the surgeon. If the surgeon cannot repair the valve, he or she will replace the valve with a prosthesis.

While you are waiting for your open heart surgery, your intensive care doctors may place a special balloon pump through your groin into aorta. After the balloon is securely placed in the aorta, you will have to be lying of your back all the time until the balloon pump is removed.

Depending on your age and other factors, the surgeon will use either a metal prosthesis or a tissue prosthesis.

WHAT IF I AM PREGNANT OR THINKING OF BECOMING PREGNANT?

Severe leakage of the mitral valve may not be well tolerated during pregnancy and the condition may harm the unborn child. Women with severe leakage who are considering becoming pregnant may need to delay their pregnancy until the condition is treaded. If you are already pregnant, you may experience significant heart problems during your pregnancy.

TRICUSPID REGURGITATION

WHAT IS MY ILLNESS?

One of the valves in your heart called the tricuspid valve is leaky; the doctors call this condition tricuspid regurgitation or tricuspid insufficiency. The tricuspid valve connects the right upper chamber of the heart called the right atrium to a pumping chamber on the right side of the heart called the right ventricle. The upper chamber is like an entryway into the main room.

Because of the leak, you may notice that your neck veins have gotten big and that you have pain under the right rib cage where the liver is. You may also have swelling in your legs.

A very common cause of a leaky tricuspid valve is infection of the valve when germs in the bloodstream eat up the valve tissue. The germs can get into your blood stream if you shoot drugs or have catheters or pacemakers placed in your veins by your doctors.

HOW WILL I BE TREATED?

Medications cannot repair the leak but antibiotics can stop further damage if the leakage is caused by an infection. Even with severe leakage of the tricuspid valve you can live for many years before the heart muscle gives up.

If your leak is severe, you may need to undergo open heart surgery to have the valve repaired or replaced by the surgeon. If the surgeon cannot repair the valve, he or she will replace the valve with a prosthesis. Most likely the surgeon will place a tissue rather than a metal prosthesis. A tissue prosthesis is made of either pig or cow tissue and looks a lot like your own valve. If you receive a tissue prosthesis you will not need to take blood thinners. However, a tissue prosthesis usually lasts about only 10 years or so.

WHAT IF I AM PREGNANT OR THINKING OF BECOMING PREGNANT?

Severe leakage of the tricuspid valve is usually well tolerated during pregnancy unless there are other heart problems.

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