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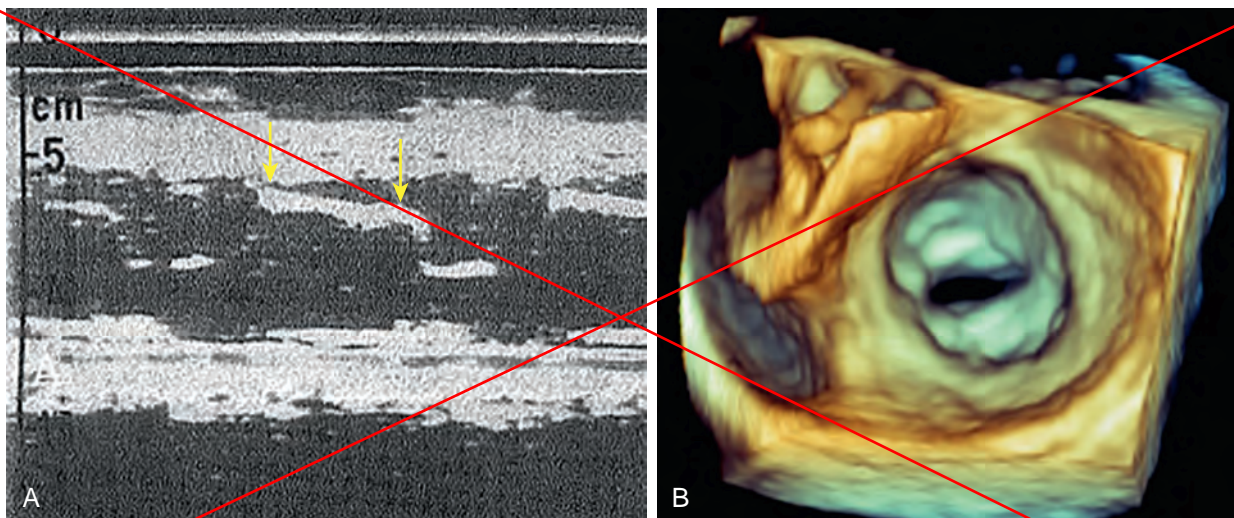
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2 SECTION XVII Mitral Stenosis



f0010 **Figure 107-1.** Mitral stenosis, past and present. **A**, One of the earliest M- mode echocardiograms of the mitral valve recorded by Edler and Hertz in December 1953. Only the anterior mitral valve is vaguely seen (arrows), but note the flat EF slope, which suggests mitral stenosis. **B**, A modern image of three-dimensional, real-time transesophageal echocardiographic view of mitral stenosis (3D zoom modality, left atrial perspective). (A Courtesy of Dr Joseph Roelandt.)

sc0015 **108** Rheumatic Mitral Stenosis

Muhamed Saric, MD, PhD, Roberto M. Lang, MD, and Itzhak Kronzon, MD

s0010 **ETIOLOGY OF MITRAL STENOSIS**

p0210 Rheumatic heart disease is the predominant but not the only cause of mitral stenosis. Mitral stenosis may result from a variety of congenital and acquired conditions. Congenital mitral stenosis is very rare (~1% of all mitral stenosis patients) and may be due to cor triatriatum, a supravulvar mitral ring, or a parachute mitral valve (typically as a part of the Shone syndrome)<sup>1</sup> as discussed in detail in another chapter of this book.

p0215 Nonrheumatic causes of acquired mitral stenosis include lupus erythematosus, carcinoid, rheumatoid arthritis, radiation valvulitis, and age-related degenerative mitral annular calcifications. They are all rare except for mitral annular calcification, which is becoming the leading cause of acquired mitral stenosis in developed countries; this is further discussed in another chapter of this book.

p0220 Worldwide, rheumatic heart disease is by far the most common cause of mitral stenosis. The disease typically starts in childhood with a bout of acute rheumatic fever and is followed by lifelong progressive valvular damage. Acute rheumatic fever is triggered by infection with group A beta-hemolytic *Streptococcus pyogenes*, typically pharyngitis (“strep throat”). The exact mechanism by which the streptococcal infection causes acute rheumatic fever and subsequent progressive, lifelong valvular damage has not been fully elucidated.

p0225 Rheumatic heart disease may be considered an autoimmune disease triggered by streptococcal infection mediated by cross-reactivity between the streptococcal M antigen (mucoïd surface protein) and human epitopes in the heart, skin, and connective and nerve tissues. Streptococci do not invade and are not present in the affected tissues. Only certain strains of group A beta-hemolytic streptococci are rheumatogenic. In general, rheumatogenic strains are highly tropic for the throat and highly contagious via direct person-to-person contact.

p0230 Clinically, acute rheumatic fever is characterized by five major findings of a nonbacterial inflammatory process: (1) pancarditis

(endocarditis, myocarditis, and pericarditis), (2) migrating arthritis of large joints, (3) subcutaneous nodules, (4) skin rash (erythema marginatum), and (5) Sydenham chorea (random rapid dancelike movements of the face and the extremities). There are also minor (nonspecific) signs of inflammation (such as fever, leukocytosis, or elevated erythrocyte sedimentation rate).

The so-called Jones criteria are used to establish the clinical diagnosis of acute rheumatic fever. In addition to evidence of a recent group A beta-hemolytic streptococcal infection (such as anti-streptococcal antibody titers), two major criteria or one major criterion plus two minor criteria are required for the diagnosis. From the historical perspective, the Jones criteria, published in 1944, pioneered the very concept of basing a clinical diagnosis on a set of major and minor criteria.<sup>2</sup>

The index streptococcal infection, as well as subsequent re-infections, sets in motion a pathological process of continued valve damage with lifelong progressive fibrosis and calcifications. Interestingly, clinical history of acute rheumatic fever cannot be elicited in a significant percentage of patients with rheumatic mitral stenosis.

Although any cardiac valve may be involved, the mitral valve is virtually always affected, and mitral stenosis with or without concomitant mitral regurgitation is the predominant chronic form. Mitral stenosis results primarily from commissural fusion along leaflet edges, with an additional contribution from chordal fusion and shortening. In general, leaflet thickening and calcification proceeds from leaflet tips toward leaflet bases. This is in contrast to mitral stenosis due to age-related mitral annular calcification, in which the process starts at the base of the posterior mitral leaflet.

**EPIDEMIOLOGY**

The prevalence of acute rheumatic fever and thus rheumatic mitral stenosis reflects the overall socioeconomic development as well as the adequacy of access to medical care in a given community.

In developed countries, the incidence of rheumatic fever has declined dramatically since World War II. Although the introduction of antibiotics to treat streptococcal infection has played a major role, the prevalence of rheumatic fever started to decline well before that, at least in part due to elimination of overcrowding in living quarters.<sup>3</sup>

p0255 In the developed countries, new cases of rheumatic fever are rare, with an incidence of less than 1 case per 100,000 people. Thus, most cases of rheumatic mitral stenosis in developed countries are found among immigrants from less developed parts of the world where the prevalence of rheumatic fever is much higher, perhaps as high as 150 cases per 100,000 people.<sup>4</sup>

p0260 Rheumatic mitral stenosis also appears to progress more rapidly in developing countries, where severe symptomatic rheumatic mitral stenosis may be observed even in children and adolescents. In contrast, in developed countries mitral stenosis becomes symptomatic 20 to 30 years after the onset of rheumatic fever. This may be due to more frequent recurrences of streptococcal infections in developing countries.

p0265 Although acute rheumatic fever affects both sexes equally, women are at least twice as likely as men to develop rheumatic mitral stenosis. This may reflect the generally higher prevalence of autoimmune disorders in women compared with men. Rheumatic fever appears to occur exclusively in humans, as there are no known animal reservoirs.

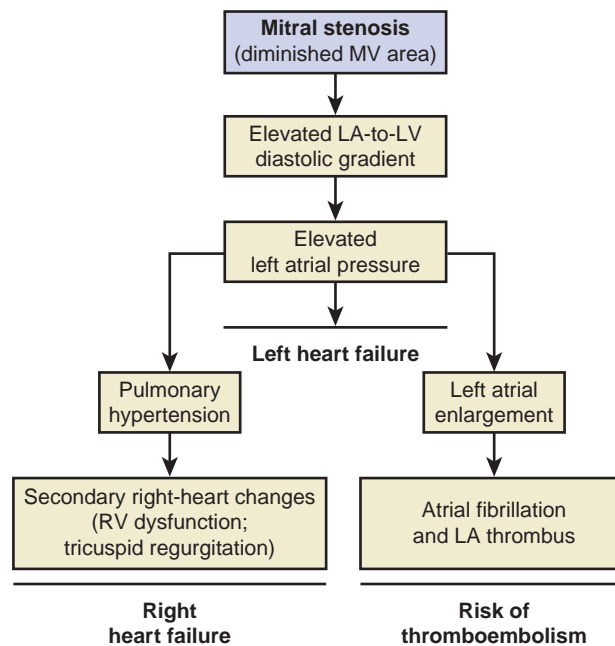


Figure 108-1. Pathophysiology of mitral stenosis. LA, Left atrium; LV, left ventricle; MV, mitral valve; RV, right ventricle. f0015

## s0020 PATHOPHYSIOLOGY

p0270 Clinical manifestations of rheumatic mitral stenosis are due to progressive decrease in mitral valve area, development of atrial fibrillation, and clot formation in the left atrium and left atrial appendage. It appears that the characteristic clinical findings of rheumatic mitral stenosis were first reported in 1668 by the English physician John Mayow (1641-1679).<sup>5</sup> Pathologic changes attributable to narrowing of the mitral orifice include elevation of left atrial pressure, pulmonary edema and other signs of left heart failure, pulmonary hypertension, right ventricular hypertrophy and dilation, secondary tricuspid regurgitation, and right heart failure.

p0275 In the adult, the normal mitral valve area is approximately 4 to 6 cm<sup>2</sup>, and the blood crosses the mitral valve without an appreciable transvalvular pressure gradient during diastole. Once the valve area drops below 2 cm<sup>2</sup>, an abnormal diastolic pressure gradient starts to develop between the left atrium (LA) and the left ventricle (LV). A high transvalvular gradient leads to elevated LA pressure, resulting in symptoms that may range from dyspnea on exertion to frank pulmonary edema at rest. This gradient is inversely proportional to the valve area and directly proportional to transvalvular flow. Conditions known to increase cardiac output, such as exercise, fever, and pregnancy, increase the blood flow and gradient across the valve. In addition, tachycardia (such as sinus tachycardia or atrial fibrillation) shortens the duration of diastole, which interferes with LA emptying and leads to a further increase the LA-LV gradient.

p0280 Elevation of LA pressure in any patient with significant rheumatic mitral stenosis may lead to pulmonary venous hypertension (pulmonary artery pressure elevations with typically normal pulmonary vascular resistance, PVR). For poorly understood reasons, a subset of patients may also develop superimposed arterial pulmonary hypertension because of arteriolar spasm, medial hypertrophy, and intimal thickening (pulmonary hypertension with elevated PVR). In some patients, pulmonary artery pressure may reach or even exceed systemic levels, leading to low cardiac output and symptoms ranging from fatigue to poor organ perfusion and cardiac cachexia.

p0285 Pulmonary hypertension may increase right ventricular (RV) afterload and lead to RV dilation, secondary tricuspid regurgitation, elevation of RV diastolic and right atrial (RA) pressures, and signs

of right heart failure: venous engorgement, leg edema, cardiac cirrhosis, ascites, and protein-losing enteropathy.

Patients with rheumatic mitral stenosis are at high risk for development of valvular atrial fibrillation and systemic thromboembolism. This risk is not directly proportional to the severity of mitral stenosis; one possible explanation is that left atrial enlargement and remodeling reflect not just elevated left atrial pressure but the underlying atrial myocarditis and patient's age.<sup>6</sup> Pathophysiology of mitral stenosis is summarized in Figure 108.1. Quantification of transmitral pressure gradient and mitral valve area is discussed in a separate chapter. p0290

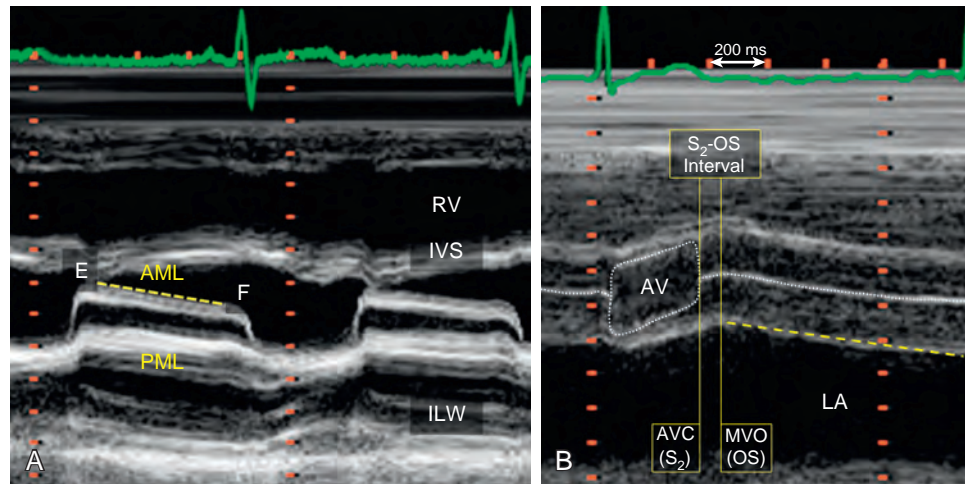
## s0025 PHYSICAL EXAMINATION

By placing the palm of one's hand on the precordium of a patient with rheumatic mitral stenosis, one can feel the diastolic valvular thrill. This palpable valvular thrill ("bruissement") was the first physical exam finding reported in 1806 by the French physician Jean-Nicolas Corvisart (1755-1821). Because of its resemblance to a cat purr, the thrill was later referred to as *frémissement cataire* by the French physician René Laennec (1781-1821). The term was later Latinized to *susurrus felinus*.<sup>7</sup> The invention of the stethoscope by Laennec led gradually to the description of the characteristic heart sounds and murmurs. p0295

Typical auditory finding of rheumatic mitral stenosis obtained by cardiac auscultation and phonocardiography include loud S<sub>1</sub>, an opening snap (OS) following S<sub>2</sub> (originally described in French as *claquement d'ouverture*), and a diastolic rumble. The first heart sound (S<sub>1</sub>) is often loud. An OS is frequently heard after the second heart sound (S<sub>2</sub>). The duration of the interval between the aortic component of the S<sub>2</sub> and the OS (referred to as S<sub>2</sub>-OS interval) is inversely proportional to the severity of mitral stenosis (shorter S<sub>2</sub>-OS interval suggesting severe mitral stenosis). S<sub>2</sub>-OS interval is roughly equivalent to the isovolumic relaxation time.<sup>8</sup> p0300

In the early years of echocardiography, a technique to measure the S<sub>2</sub>-OS interval was developed using M-mode recordings of the aortic valve and the left atrium (Fig. 108.2). In severe mitral stenosis, S<sub>2</sub>-OS interval is typically less than 80 msec. This M-mode technique, though of historic significance, is not a part of the current p0305

4 SECTION XVII Mitral Stenosis



**Figure 108-2.** M-mode echocardiography of rheumatic mitral stenosis. **A**, M-mode recording through the mitral valve. Note marked thickening and increased echogenicity of both mitral leaflets and a very flat EF slope indicative of significant mitral stenosis. **B**, M-mode recording through the aortic valve (AV) and left atrium (LA). Note a very short S<sub>2</sub>-OS interval (about 80 msec) and markedly diminished rate of LA emptying (broken yellow line) after mitral valve opening (MVO). AML, Anterior mitral leaflet; AVO, aortic valve opening; ILW, inferolateral wall; IVS, interventricular septum; OS, opening snap; S<sub>2</sub>, second heart sound; PML, posterior mitral leaflet; RV, right ventricle.

mitral stenosis guidelines.<sup>9</sup> A characteristic diastolic rumble is frequently heard best at the apex. In patients with normal sinus rhythm, there is also an end-diastolic (“presystolic”) accentuation of the rumble. Unfortunately, the auscultatory findings are subtle. They can be frequently missed by an inexperienced ear. When the cardiac output declines, the murmur may become softer. The pulmonic component of S<sub>2</sub> is loud in cases of pulmonary hypertension.

Less specific findings include malar flush and sign of left and right heart failure as well as signs of low cardiac output. Left atrial enlargement, which may be massive in rheumatic mitral stenosis, occasionally compresses the left recurrent laryngeal nerve to cause hoarseness (Ortner syndrome).<sup>10</sup> A combination of atrial septal defect and rheumatic mitral stenosis is sometimes referred as Lutembacher syndrome.<sup>11</sup>

**ELECTROCARDIOGRAPHY**

Electrocardiography (ECG) may demonstrate signs of LA enlargement (*P mitrale*) with wide, saddle-shaped P wave in leads I and II, as well as late, deep P wave inversion in lead V<sub>1</sub>. In more advanced cases, there is also evidence of RV hypertrophy. Atrial fibrillation is a frequent finding in patients with rheumatic mitral stenosis. In 1749, the French physician Jean-Baptiste de Sénac (1693-1770) was the first to postulate a correlation between rheumatic mitral stenosis and the irregular pulse of “rebellious palpitations” or *delirium cordis*.<sup>12</sup> With the advent of electrocardiography, the British physician Thomas Lewis (1881-1945) recognized, in 1909, this irregular rhythm of rheumatic mitral stenosis as auricular (atrial) fibrillation.<sup>13</sup>

**CHEST RADIOGRAPHY**

Chest x-ray usually demonstrate LA enlargement with straightening of the left cardiac silhouette due to the enlargement of the LA appendage. Some combination of mitral valve calcifications, pulmonary venous engorgement, pulmonary edema, and right ventricular enlargement may also be seen.

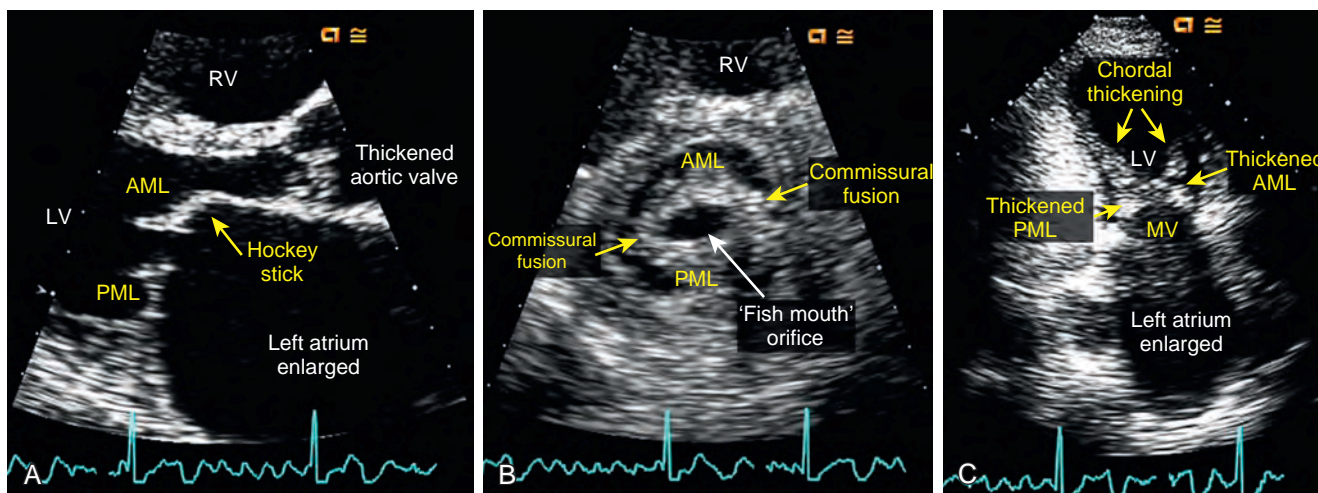
**TRANSTHORACIC ECHOCARDIOGRAPHY**

In the 1950s, rheumatic mitral stenosis was the very first heart disease visualized echocardiographically by the inventors of echocardiography, Inge Edler (1911-2001) and Carl Hertz

(1920-1990).<sup>14</sup> In this section we concentrate on echocardiographic analysis of mitral valve morphology and secondary cardiac changes in rheumatic mitral stenosis (Fig 108.3/Video 108.3, A-D). Quantification of mitral valve area, transvalvular mitral gradient, and right heart pressures is discussed in detail in another chapter of this book.

During bouts of acute rheumatic fever, which are exceedingly rare in developed countries, mitral regurgitation is the primary echocardiographic finding. In contrast, during chronic phase, rheumatic mitral stenosis with or without concomitant mitral regurgitation is the norm. The rheumatic process affects both mitral leaflets and chordae. Leaflet thickening, leaflet calcifications, decreased leaflet mobility, and commissural fusions are the hallmarks of rheumatic mitral stenosis together with chordal fusion and shortening. Leaflet thickening and calcification start at leaflet tips and extends toward leaflet bases over time. Similarly, chordal thickening and fusion starts at leaflet tips and then extends distally toward papillary muscles. This orderly progression of rheumatic disease in the tip-to-base leaflet direction and the tip-to-papillary muscle direction along the chordae is pathognomonic forms the foundation for grading the severity of rheumatic mitral stenosis and eligibility for percutaneous mitral balloon valvuloplasty (see “Treatment,” later). Leaflet thickening can be observed in any two-dimensional (2D) echocardiographic view of the mitral valve, although leaflet thickness is typically measured in the parasternal long-axis view. In this view, one can also observe decreased mobility of the posterior mitral leaflet and the characteristic doming resulting in a hockey-stick appearance of the anterior leaflet (see Fig. 108.3/Video 108.3, A).

On the 2D parasternal short-axis view at the level of the mitral valve, one appreciates commissural fusions resulting in a fish-mouth appearance of the mitral orifice (see Fig. 108.3/Video 108.3, B). It is in this view that 2D planimetry of the mitral orifice is performed—with the caveat that on 2D, one may not be able to identify true leaflet tips because in rheumatic mitral stenosis, the orifice may be eccentric and separate from the 2D short-axis plane. The eccentricity of the orifice can often be appreciated in 2D apical views of the mitral valve. Three-dimensional (3D) echocardiography, especially multiplane reconstruction techniques, overcomes limitations of 2D echocardiography and allows for precise orifice planimetry exactly at leaflet tips as described in the chapter on quantification of mitral stenosis. Chordal involvement is best still in the apical transthoracic views, especially the apical two-chamber



**Figure 108-3.** Transthoracic echocardiography (TTE) of rheumatic mitral stenosis. **A**, Parasternal long-axis view demonstrates characteristic hockey-stick appearance of the anterior mitral leaflet (AML), decreased mobility of the posterior mitral leaflet (PML), and markedly enlarged left atrium consistent with rheumatic mitral stenosis. Note also the rheumatic aortic valve thickening. **B**, Parasternal short-axis view at the level of the mitral valve. Note the commissural fusion resulting in mitral stenosis with the characteristic fish-mouth appearance of the mitral orifice. **C**, Apical two-chamber view demonstrates marked chordal thickening and chordal fusion as well as thickening of both mitral leaflet and marked left atrial enlargement. AML, Anterior mitral leaflet; LV, left ventricle; MV, mitral valve; PML, posterior mitral leaflet; RV, right ventricle. (See corresponding Video 108-3, A-D.)

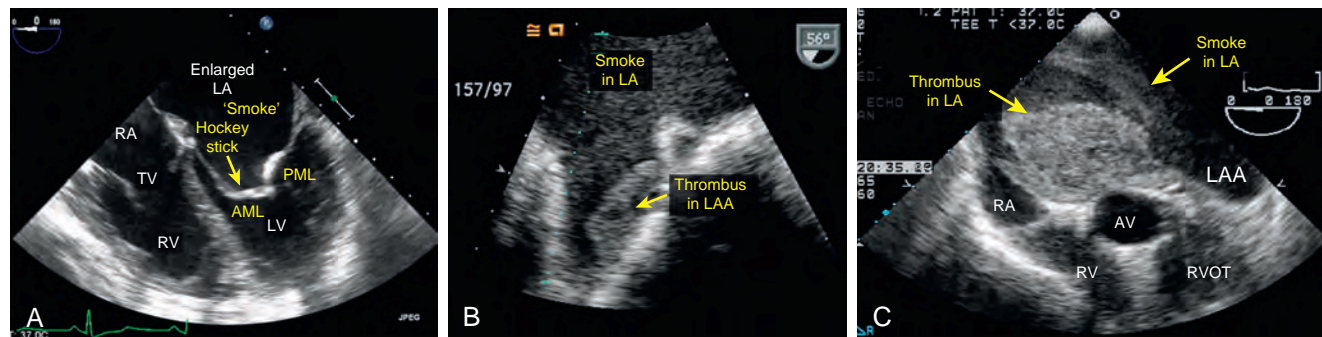
view or its transesophageal echocardiography (TEE) equivalents, such as the transgastric two-chamber view (see Fig. 108.3/Video 108.3, C).

Changes associated with rheumatic mitral stenosis include often a very marked left atrial enlargement (which is not necessarily proportional to the degree of mitral stenosis), aortic and tricuspid regurgitation, right heart enlargement, and elevation of left atrial and right heart pressures. Left ventricle typically has normal size and systolic function.

Many of the previously mentioned changes can also be seen on M-mode echocardiography. The severity of mitral stenosis can be semiquantitatively assessed on M-mode echocardiography by measuring either the EF slope of the mitral valve (see Fig. 108.2, A) or by the rate of left atrial emptying (see Fig. 108.2, B). Normally, EF slope is steep (>8 cm/sec). The flatter the EF slope, the more severe the mitral stenosis. Similarly, the slower the rate of LA emptying during diastole, the more severe the mitral stenosis. Although of historic importance, these M-mode techniques are imprecise and are not part of the current American Society of Echocardiography (ASE) valvular stenosis guidelines.

### TRANSESOPHAGEAL ECHOCARDIOGRAPHY

The routine echocardiographic evaluation of rheumatic mitral stenosis does not require TEE. However, TEE should be considered when the image quality and the Doppler information are suboptimal or do not correlate with the clinical impression (Fig. 108.4/Video 108.4, A). TEE is especially useful in the evaluation of complications of rheumatic mitral stenosis (such as LA clot or endocarditis) and in guiding percutaneous mitral balloon valvuloplasty. Patients with rheumatic mitral stenosis are at a high risk for developing intracardiac thrombus. This is particularly the case in those with atrial fibrillation; however, LA stasis with smoke, sludge, and thrombus formation often occurs in such patients even in sinus rhythm. In rheumatic mitral stenosis, the majority of thrombi are in the left atrial appendage (LAA) (see Fig. 108.4/Video 108.4, B); however, a substantial proportion occurs in the body of the left atrium (see Fig. 108.4/Video 108.4, C). According to one surgical series, 57% of thrombi were in the LAA versus 43% in the LA.<sup>15</sup> This is contrast to nonvalvular atrial fibrillation, in which thrombi outside the LAA are unusual.



**Figure 108-4.** Two-dimensional transesophageal echocardiography (TEE) of rheumatic mitral stenosis. **A**, Midesophageal four-chamber view demonstrates findings typical of rheumatic mitral stenosis: thickening and calcifications of both anterior (AML) and posterior mitral leaflet (PML), hockey-stick appearance of AML, and marked enlargement of the left atrium (LA). **B**, TEE demonstrates a large thrombus in the left atrial appendage (LAA) as well as smoke in the body of the LA. **C**, TEE shows a very large thrombus in the body of the LA as well as smoke in LA and LAA. (See corresponding Videos 108-4, A-C.)

**THERAPY**

s0050 p0355 Rheumatic mitral stenosis is a progressive lifelong disease. Without treatment, the 10-year survival is less than 15% once significant symptoms develop, and in those who develop severe pulmonary hypertension, mean survival drops to less than 3 years.

s0055 **Medical Therapy**

p0360 There is no effective medical therapy that alters the progression of valvular disease except for long-term antibiotic prophylaxis of recurrent streptococcal pharyngitis in endemic areas. Medical therapy with diuretics and heart-rate controlling agents (beta blockers, calcium channel blockers, and digoxin) provides symptomatic relief. It appears that the very first clinical use of digoxin was in patients with atrial fibrillation and rheumatic mitral stenosis, as reported in 1785 by the British physician William Withering (1741-1799), who discovered digitalis.<sup>16</sup>

p0365 Management of atrial fibrillation (including chemical and electrical cardioversion) should follow standard guidelines. Chronic anticoagulation to prevent thromboembolism is recommended in patients with atrial fibrillation and may be considered even in patients with sinus rhythm. In contrast to nonvalvular atrial fibrillation, efficacy of anticoagulation in rheumatic mitral stenosis has not been proven in a randomized trial; nonetheless, anticoagulation is recommended based on several retrospective studies.<sup>17</sup> Although patients with rheumatic mitral stenosis are at risk for mitral valve endocarditis, current guidelines do not recommend routine antibiotic prophylaxis for endocarditis. Significant rheumatic mitral stenosis is a mechanical disorder that requires a mechanical treatment for effective symptom relief and improvement in survival.

s0060 **Percutaneous Mitral Balloon Valvuloplasty**

p0370 In eligible patients, percutaneous mitral balloon valvuloplasty (PMBV) is the treatment of choice; surgery is reserved for those who cannot undergo valvuloplasty. Currently, approximately 1500 PMBVs are performed in the United States annually. The PMBV technique was perfected in the 1980s by Kanji Inoue of Japan, who developed an ingenious balloon (Inoue Balloon, Toray Industries Inc, San Mateo, CA), which remains the preferred balloon for PMBV.<sup>18</sup>

p0375 Transthoracic echocardiography allows selection of appropriate candidates for valvuloplasty using four parameters: three related to mitral leaflets (thickening, mobility, and calcification) and one related to the degree of chordal involvement. From these, the so-called Wilkins score is derived.<sup>19</sup> Each parameter is scored on a scale from 0 (normal) to 4 (severe); thus the Wilkins score ranges from 0 (normal valve) to 16 (poorly mobile, severely calcified leaflets with severe chordal fusion and shortening). Good valvuloplasty candidates should have a Wilkins score of 10 or less and preferably 8 or less (Table 108.1).

TEE plays essential role in the refinement of patient selection (Fig. 108.5/Video 108.5, A), guidance of percutaneous valvuloplasty (see Fig. 108.5/Video 108.5, B), and assessment of postvalvuloplasty results (see Fig. 108.5/Video 108.5, C, D). In the absence of contraindications, PMBV is recommended in the following instances: (1) in symptomatic patients with moderate or severe mitral stenosis; and (2) in asymptomatic patients with moderate or severe mitral stenosis when pulmonary artery systolic pressure is greater than 50 mm Hg at rest or greater than 60 mm Hg with exercise, or when there is new-onset atrial fibrillation; and (3) in symptomatic patients with mild stenosis (valve area > 1.5 cm<sup>2</sup>) when pulmonary artery systolic pressure is greater than 60 mm Hg, pulmonary artery wedge pressure is greater than 25 mm Hg, or mean mitral valve gradient is greater than 15 mm Hg during exercise. Contraindications for PMBV include unfavorable mitral valve Wilkins score (≥ 10), more than moderate mitral regurgitation, and the presence of intracardiac thrombus.

TEE plays essential role in guiding PMBV, including guidance of transseptal puncture, visualization of wires and catheters, and proper placement of the valvuloplasty balloon across the mitral orifice, as well as in the assessment of procedure outcomes. A favorable outcome is characterized by leaflet separation along commissural lines with a concomitant increase in mitral valve area and a decrease in transvalvular gradient (see Fig. 108.5/Video 108.5, C). Leaflet tear is the most feared complication of PMBV as it may lead to severe acute mitral regurgitation necessitating urgent mitral valve surgery (see Fig. 108.5/Video 108.5, C).

**Mitral Valve Surgery**

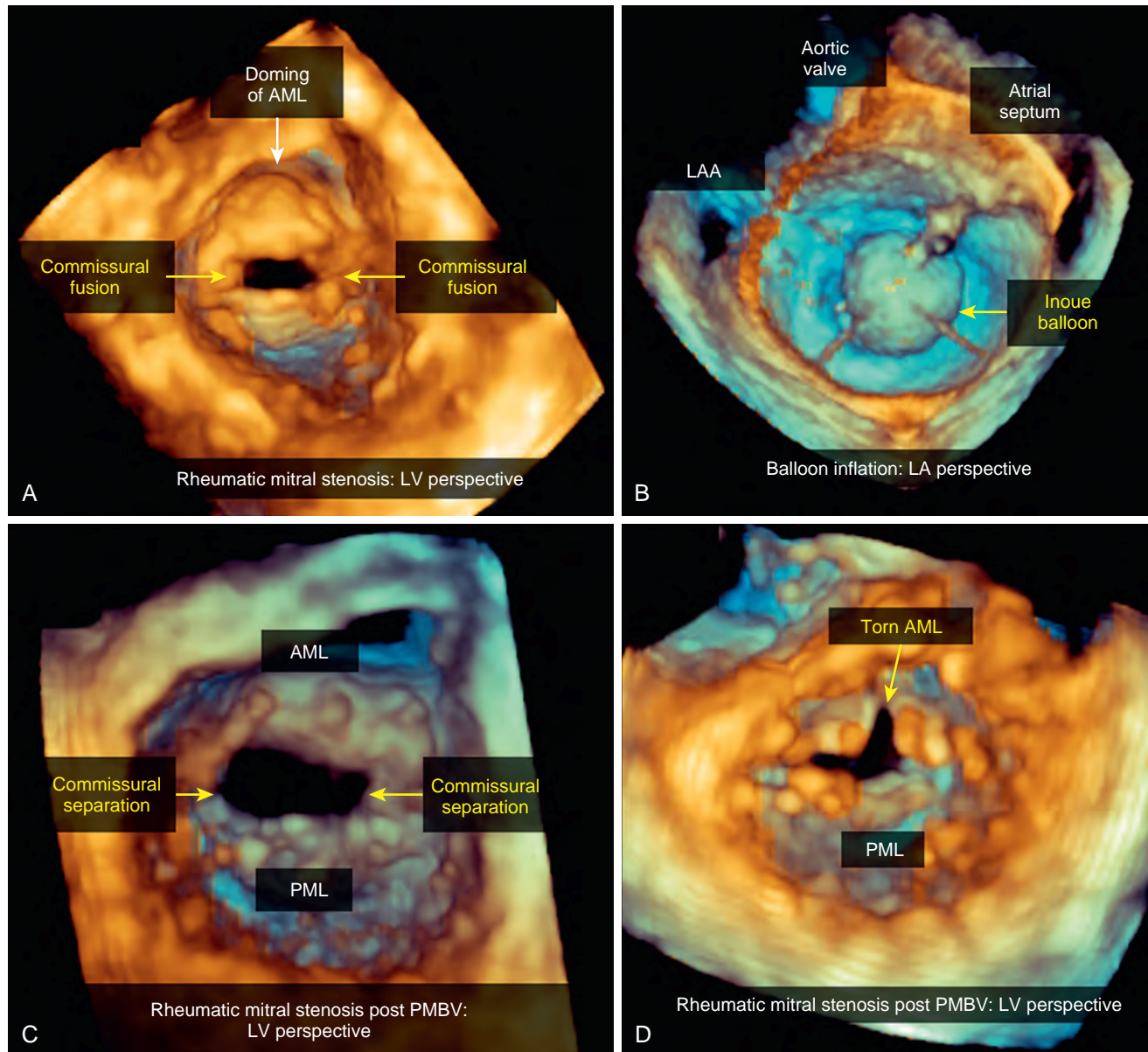
Surgical options include closed or open commissurotomy, mitral valve replacement, and occasionally mitral valve repair. In the 1920s, Henry Souttar (1875-1964) in England and Elliot Cutler (1888-1947) in the United States led the first attempts to perform surgical relief of rheumatic mitral stenosis using what would later be termed closed commissurotomy. Their technique was improved soon after World War II by the American surgeons Charles Bailey (1910-1993) and Dwight Harken (1910-1993); Bailey called the procedure “mitral commissurotomy,” whereas Harken called it “mitral valvuloplasty.”<sup>20</sup> In developed countries, open commissurotomy requiring cardiac arrest and cardiopulmonary bypass is preferred over closed commissurotomy, which is performed on a beating heart. Because of its simplicity, closed commissurotomy is still widely performed in developing countries. In the 1960s, rheumatic mitral stenosis was the first valvular disease to be treated with a prosthetic valve replacement using the mechanical mitral valve developed by Albert Starr (born 1926) and Lowell Edwards (1898-1982).<sup>21</sup> For further discussion of surgical options in rheumatic mitral stenosis, the reader is referred to appropriate treatment guidelines.<sup>22</sup>

Please access ExpertConsult to view the corresponding videos for this chapter.

TABLE 108-1 Wilkins Criteria for Assessment of Mitral Valve Anatomy Prior to Percutaneous Balloon Valvuloplasty

Grade	0	1	2	3	4
Leaflet mobility	Normal	Highly mobile with only leaflet tip restricted	Leaflet mid and base portions have normal mobility	Valve continues to move in diastole, mainly from the base	No or minimal forward leaflet motion in diastole
Valve thickness	Normal	Near normal thickness (4-5 mm)	Mid-leaflets normal; thickened leaflet tips (5-8 mm)	Entire leaflet thickened (5-8 mm)	Marked thickening of entire leaflet (>8-10 mm)
Leaflet calcification	None	Single area of echo brightness	Scattered areas of brightness at leaflet margins	Brightness extends to mid leaflets	Extensive brightness throughout leaflets
Subvalvular thickening	None	Minimal chordal thickening just below leaflet tips	Thickening from leaflet tips to up to 1/3 of chordal length	Thickening extending to the distal third of chordae	Extensive chordal thickening down to papillary muscle

The Wilkins score ranges from 0 (normal valve) to 16 (poorly mobile, severely calcified leaflets with severe chordal fusion and shortening). Good valvuloplasty candidates should have a Wilkins score of 10 or less and preferably 8 or less.



**Figure 108-5.** Three-dimensional transesophageal echocardiography (3D-TEE) in guidance of percutaneous mitral balloon valvuloplasty (PMBV) of rheumatic mitral stenosis. **A**, Before PMBV, there is typical appearance of rheumatic mitral stenosis from left ventricular (LV) perspective. Note the commissural fusion, doming of the anterior mitral leaflet (AML), and the fish mouth appearance of the mitral orifice. **B**, During PMBV, 3D-TEE is used to properly place the balloon across the mitral orifice. **C**, In this patient, PMBV had a favorable outcome: mitral orifice increased in size as a result of commissural separation. **D**, This patient suffered a complication of PMBV, namely tearing of the AML (arrow), which resulted in severe acute mitral regurgitation. AML, Anterior mitral leaflet; LAA, left atrial appendage; PML, posterior mitral leaflet. (See corresponding Videos 108-5, A-D.)

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sc0020

# 109 Quantification of Mitral Stenosis

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p0400 Echocardiography is the modality of choice for the diagnosis of mitral stenosis. The joint American Society of Echocardiography and European Association of Echocardiography guidelines for native valvular stenosis feature an exhaustive review of echocardiographic methods for quantitative assessment of mitral stenosis.<sup>1</sup> Full echocardiographic evaluation of mitral stenosis includes the following three sets of parameters: (1) mean diastolic transmitral pressure gradient; (2) mitral valve area (MVA); and (3) secondary changes including measurements of relevant chamber sizes and estimation of right heart pressures. Most modern ultrasound systems contain built-in software packages for determining these parameters. Major methods for quantification of mitral stenosis are presented in Figures 109-1 and 109-2. In most instances, evaluation of mitral stenosis by invasive methods of cardiac catheterization is not necessary unless there is a discrepancy between clinical and echocardiographic findings.

of the exact direction of the transmitral flow. The angle between the interrogating beam and the transmitral jet should be 0 degrees.

Gradient can be assessed by pulsed-wave Doppler with the sample volume at the tips of the leaflets or by continuous-wave Doppler. By tracing the spectral Doppler-derived diastolic transmitral flow velocity envelope and with the use of built-in algorithms available in most modern ultrasound imaging systems, one can obtain the mean MV gradient (see Fig. 109-1, A). To calculate the mean gradient, the system first calculates instantaneous pressure gradients using the simplified Bernoulli equation (see Eq. 109-4, later) and then averages them out:

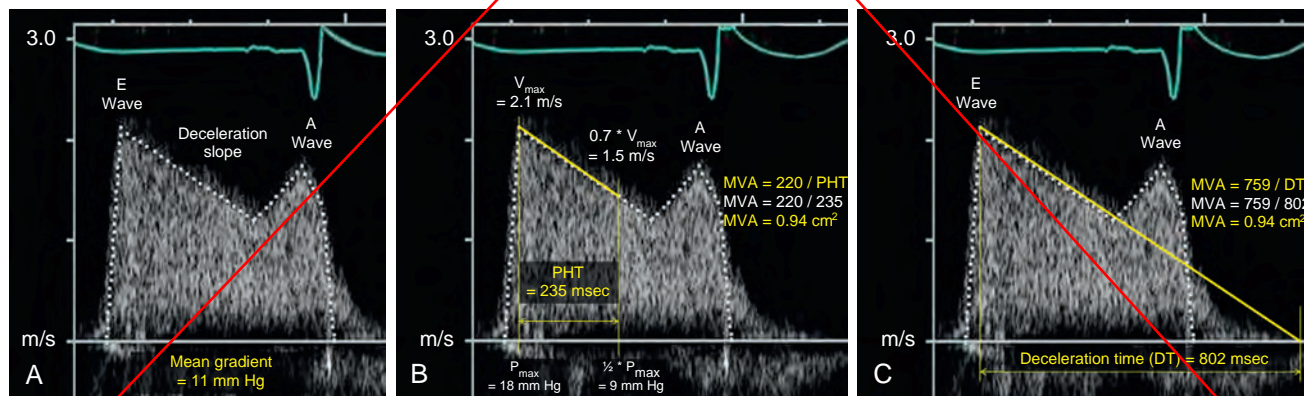
$$\Delta P = \frac{\sum_{i=0}^n V_i^2}{n} \quad (109-1)$$

where  $\Delta P$  is the mean diastolic transmitral pressure gradient,  $V$  is the instantaneous transmitral velocity, and  $n$  is the number of instantaneous gradients measured.

In the presence of atrial fibrillation, mean diastolic gradient should be averaged from multiple (typically five) cardiac cycles. When the mitral valve is normal, there is no significant diastolic transmitral pressure gradient. In severe mitral stenosis, the mean gradient is typically greater than 10 mm Hg; in moderate stenosis it is between 5 and 10 mm Hg, and in mild stenosis it is less than 5 mm Hg. It is important to emphasize that these cutoff values assume a normal transmitral flow (a normal stroke volume and a

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p0405 Mean diastolic pressure gradient is inversely related to MVA; that is, the more severe the mitral stenosis, the higher the mean diastolic pressure gradient across the mitral valve. This gradient can be easily measured by pulsed- and continuous-wave Doppler.<sup>2</sup> The best approach for transmitral flow evaluation and gradient determination should be with the transducer at the apex, imaging in four-chamber or two-chamber views. Color flow imaging can be helpful for the assessment



p0400 **Figure 109-1.** Quantitative assessment of mitral stenosis by spectral Doppler. **A**, Assessment of the mean diastolic mitral gradient. This patient has severe mitral stenosis (mean gradient 11 mm Hg at a heart rate of 70 beats per minute). **B**, Mitral valve area by pressure half-time (PHT). This patient has severe mitral stenosis (mitral valve area 0.94 cm<sup>2</sup>). Note that a 50% drop in initial pressure (from 18 to 9 mm Hg) corresponds to a 70% drop in initial velocity (from 2.1 m/sec to 1.5 m/sec). (See Video 109-1, B, which conceptually corresponds to this panel, albeit in a different patient.) **C**, Mitral valve area by deceleration time (DT). Because DT is approximately 0.29 × PHT, the formula MVA = 220/PHT may be expressed as MVA = 759/DT.