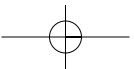
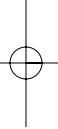
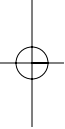
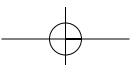
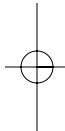
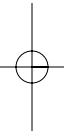
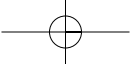


Section one

Valve disease





CHAPTER 1

Mitral stenosis

Kewal Krishan Talwar and Manojkumar Rohit

Introduction

Mitral stenosis (MS) is a progressive disease that can result in serious complications which may be fatal unless an intervention enlarges the mitral valve orifice enough to permit adequate cardiac output. The predominant cause of MS is rheumatic heart disease. Approximately 25% of all patients with rheumatic heart disease have pure MS, and an additional 40% have combined MS and mitral regurgitation.¹

When MS is symptomatic, the anatomic features consist of thickened mitral cusps, fusion of the valve commissures, shortening and fusion of the chordae tendineae, or a combination of these features. Characteristically, mitral valve cusps fuse at their edge, and fusion of the chordae tendineae results in thickening and shortening of these structures. Although the major obstruction in patients with MS is usually caused by fusion of commissures, it may be below the valve itself, secondary to fusion of the chordae, and this assessment is important because significant subvalvular involvement leads to suboptimal results with mitral commissurotomy or balloon dilatation.

Other rare cause of MS include congenital mitral stenosis (e.g. supramitral ring, cor triatriatum), mitral annular calcification, systemic lupus erythematosus, rheumatoid arthritis, and mucopolysaccharidoses.

Although there are multiple clues to the presence of MS by physical examination, they are often subtle and likely to be overlooked during a routine physical examination of an asymptomatic patient. The diagnosis of MS is often made when the patient presents with a complication (e.g. atrial fibrillation, embolism, acute pulmonary edema, or massive hemoptysis).

The various imaging modalities that are useful in confirming the diagnosis and assessing the severity of MS are discussed in this chapter.

Case Presentation

A 25-year-old woman was referred to our Institute with progressive shortness of breath for 6 months, with chest X-ray as shown in Fig. 1.1. This chest X-ray shows straightening of left heart border with pulmonary venous hypertension. How consistent is this with a diagnosis of MS?

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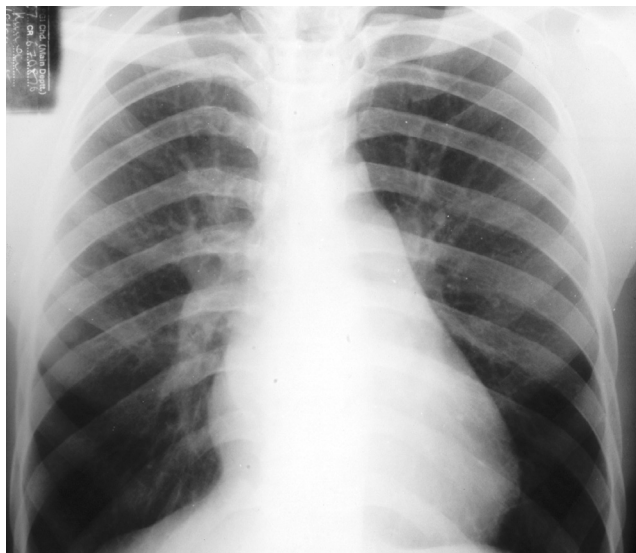


Figure 1.1 Chest X-ray posteroanterior (PA) view showing straightening of left heart border, cephalization of pulmonary veins, and double atrial shadow.

The most frequent roentgenographic findings in MS include left atrial enlargement, redistribution of blood flow to the upper lobes of the lung, Kerley B lines, and enlarged pulmonary artery. Although their cardiac silhouette may be normal in the frontal projection, patients with hemodynamically significant MS almost invariably have evidence of left atrial enlargement.

Left atrial enlargement is one of the earliest signs of mitral stenosis; however, its presentation may be subtle and limited to enlargement of the left atrial appendage, causing a straightening of the left heart border. In more advanced cases, the left atrium is recognized as a double density and elevation of main stem bronchus on the postanterior film. Radiologic changes in the lung fields indirectly reflect the severity of MS. Redistribution of blood flow to the upper lobes correlates best with the degree of mitral valve obstruction. The presence of Kerley B lines is an important finding in patients with MS. These are fine parallel densities in the peripheral lung fields which are perpendicular to a pleural surface and are most frequently seen in the costophrenic sulci. The lines are caused by thickened interlobar septa and signify chronic pulmonary venous hypertension.

This finding is present in 30% of patients with resting pulmonary arterial wedge pressures less than 20mmHg and in 70% of patients with pressures greater than 20mmHg.

Case Presentation (Continued)

Although the chest X-ray is strongly suggestive of MS, management decisions need to be informed by details of anatomy and severity, so echocardiography remains the examination of choice for evaluating MS. Indeed, the chest X-ray may be unnecessary, and of course should be avoided in a pregnant woman. The patient's M-mode showed a decreased E-F slope and the posterior mitral leaflet moved anteriorly during diastole, indicative of MS (Fig. 1.2). Is the M-mode still useful?

Although a decreased E-F slope is almost always present in severe MS, it is not diagnostic of MS and it is an unreliable indicator of its severity. The specificity of the diagnosis of MS by M-mode echocardiography is greatly improved by visualizing the initial diastolic movement of the posterior mitral leaflet.² In the normal mitral valve, the posterior leaflet moves away from the anterior leaflet during early diastole. In MS, the posterior leaflet moves anteriorly during early diastole.

Case Presentation (Continued)

The patient's two-dimensional (2D) imaging in the short axis view showed a typical "fishmouth" appearance of severely stenotic mitral valve with a mitral valve area of 0.6 cm^2 (Fig. 1.3). Does this confirm the diagnosis of MS on 2D imaging, and how should we evaluate severity of MS and suitability for percutaneous mitral commissurotomy?

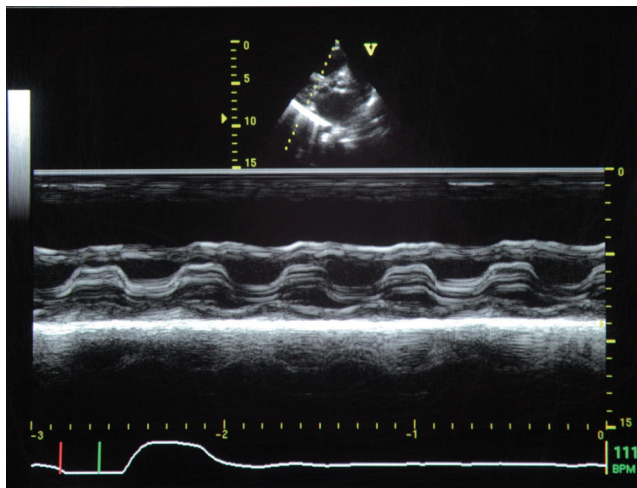


Figure 1.2 M-mode echo showing paradoxical posterior mitral leaflet (PML).

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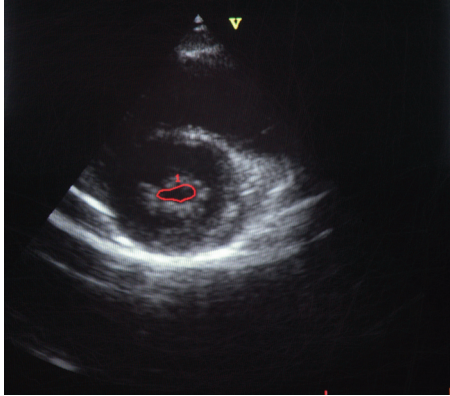


Figure 1.3 Parasternal short axis view showing typical “fishmouth” appearance of severely stenotic mitral valve (mitral valve area [MVA] 0.6 cm^2).

The short axis view allows the mitral valve area (MVA) to be measured by planimetry, although technical factors may compromise the accuracy of this method—not least the difficulty in ensuring that imaging is being performed at the tips of the leaflets. Heavily calcified leaflets may have indistinct borders that are difficult to trace, and there may also be dropout of echoes, leaving gaps in the area to be traced. The hallmark of MS on 2D echocardiography is thickening and restriction of motion of both mitral valve leaflets, with the predominant pathologic process being at the tips of the leaflets and proximal chordae. The abnormal motion of the leaflets is apparent in early diastole. Fusion of the commissures causes restriction in the motion of the tip of the anterior leaflet. The commissural fusion usually causes the posterior leaflet to move anteriorly during diastole with the larger anterior leaflet rather than moving posteriorly.

Doppler echocardiography assesses the severity of the stenotic lesion and color flow imaging is instrumental in determining associated mitral regurgitation. This is important because moderate mitral regurgitation (more than 2+) would be a contraindication to perform a closed procedure. MS produces characteristic changes in the mitral flow velocity pattern, involving an increase in the early diastolic peak velocity of flow and slower than normal rate of fall in velocity. The transvalvular pressure gradient can be measured continuously throughout diastole and correlates well with mean pressure gradient measured by cardiac catheterization. However, the pressure gradient is affected by heart rate, cardiac output, and valvular regurgitation in addition to orifice area, and hence it provides only a rough estimate of severity.

The pressure half-time is the time required for the instantaneous gradient across the valve to fall to half of the peak value (Fig. 1.4). This means of assessing MVA is usually sufficiently accurate for clinical use. The pressure half-time method is not valid for several days after mitral balloon valvuloplasty, probably because of a decrease in left atrial pressure without a commensurate improvement in left atrial compliance.

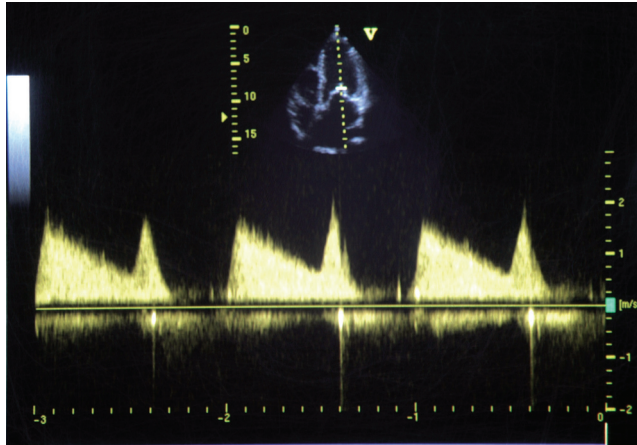


Figure 1.4 Continuous wave Doppler showing gradient across severely stenotic mitral valve.

Case Presentation (Continued)

Pressure half-time measurement in this case showed a mean gradient of 16 mmHg at the heart rate of 66 b/min (Fig. 1.4). Color Doppler performed on our index patient confirmed the stenotic mitral valve and fortunately showed no mitral regurgitation. Is the patient suitable for percutaneous intervention?

An echocardiographic scoring system developed by Wilkins *et al.*³ has been used widely for assessment of suitability for percutaneous mitral commissurotomy. Leaflet rigidity, thickening, valvular calcification, and subvalvular involvement are each scored from 0 to 4. A score of 8 or less is usually associated with excellent immediate and long-term results, whereas scores exceeding 8 are associated with less impressive results. In our experience, slight commissural calcium is not a contraindication for percutaneous mitral commissurotomy, but when the calcium score is more than 2+, the incidence of restenosis is higher and thus surgical repair of the mitral valve is preferable. Significant subvalvular pathology is a more important determinant of suboptimal results following percutaneous mitral commissurotomy.

In addition to determining the presence and severity of MS, it is important to evaluate the heart for secondary effects of MS. These include left atrial enlargement, stasis, thrombus formation, and secondary pulmonary hypertension. The aortic, tricuspid, and pulmonic valves can likewise be directly evaluated for evidence of rheumatic involvement.

8 Chapter 1**Case Presentation** (Continued)

According to the Wilkins scoring system, our patient's mitral valve score was 6. There was no calcium on the mitral valve. This is an ideal candidate for percutaneous transmitral commissurotomy (PTMC) and a good result could be anticipated. What other steps are required before PTMC?

Transesophageal echocardiography

Transesophageal echocardiography (TEE) provides excellent images of the mitral valve leaflets, the left atrium, and the left atrial appendage (Fig. 1.5). Transthoracic imaging is usually diagnostic of MS and can accurately assess the severity of the stenosis. However, in some cases the transthoracic acoustic window may be inadequate. Multiplane TEE visualizes most of this anatomically complex structure and thrombus in the appendage can be accurately diagnosed, although experience is needed to avoid mistaking the pectinate muscle for thrombus.

TEE is well established as the gold standard for detecting thrombi in the left atrium (LA) and LA appendage, with a sensitivity and specificity of 100% and 99%, respectively.⁴ The semi-invasive nature and safety of the test make it ideal for serial follow-up of thrombi in the LA body and appendage. TEE is also indicated if there is doubt regarding the presence or severity of mitral regurgitation and assessment of subvalvular pathology, if this is unclear on transthoracic echocardiography (Fig. 1.6). In our practice, all patients undergoing balloon valvuloplasty with atrial fibrillation and suspicion of clot on transthoracic echocardiography undergo TEE before the procedure. Finally, TEE may be used to guide the atrial trans-septal puncture during the balloon valvuloplasty procedure,⁵ although we do not use TEE during the procedure at our institution.

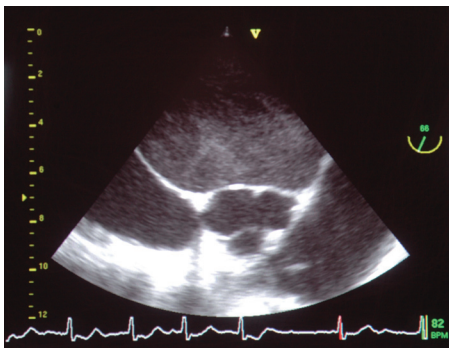


Figure 1.5 Transesophageal echocardiography (TEE) showing large left atrium with dense spontaneous contrast and amputated left atrial appendage.

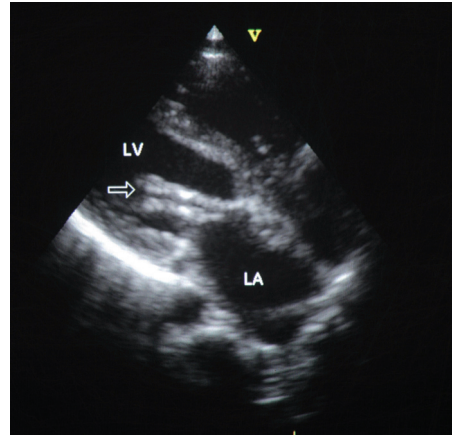


Figure 1.6 Parasternal long axis showing stenotic mitral valve and significant subvalvular thickening.

Case Presentation (Continued)

In this case, TEE was performed before PTMC. TEE showed dense spontaneous contrast but there was no clot in the left atrium (Fig. 1.5).

At cardiac catheterization before PTMC, left heart pressure was measured by retrograde catheterization of the left ventricle. Left atrial pressure was initially measured by pulmonary artery (which is accurate) and later by direct entry into the left atrium through trans-septal puncture. The mean gradient across the mitral valve was 15 mmHg before the PTMC (Fig. 1.7). The mitral valve was successfully dilated with a 26-mm Inoue balloon and the mean gradient across the mitral valve after the procedure was 4 mmHg with mitral valve area of 1.7 cm² and trivial mitral regurgitation.

Recently, there has been much interest in cardiac magnetic resonance imaging (MRI) and three-dimensional echocardiography in the assessment of valve lesions. We do not believe that MRI gives any additional information over echocardiography for MS, and MRI is expensive, time-consuming, and may be compromised by atrial fibrillation, which is not uncommon in MS. Three-dimensional echocardiography is a newer imaging modality and, in selected cases, will be useful, especially to assess the subvalvular apparatus.

Three-dimensional echocardiography

Real-time three-dimensional (R3D) echocardiography is a novel technique that permits visualization of mitral valvular anatomy in any desired plane of orientation. The use of R3D echocardiography in evaluation of mitral stenosis has been studied by Zamorano *et al.*⁶ In 76 patients with significant MS, these

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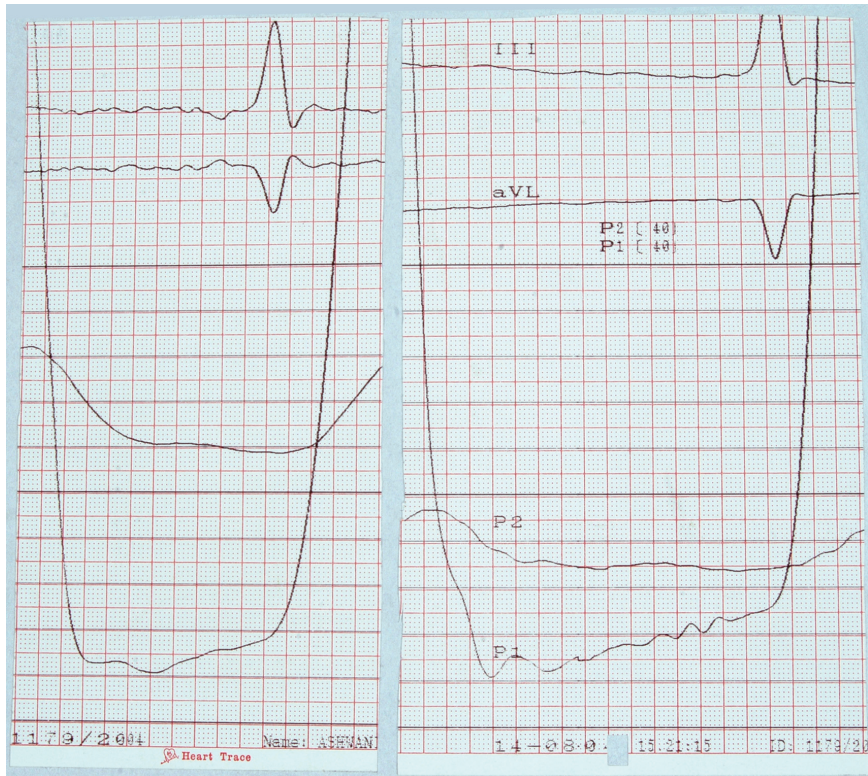


Figure 1.7 Left heart catheterization of left ventricle (LV) and left atrium (LA) tracing showing gradient across mitral valve before and after percutaneous transmittal commissurotomy (PTMC).

authors demonstrated that R3D echocardiography is a feasible, accurate, and highly reproducible technique for assessing MVA. MVA calculation with R3D echocardiography has the best agreement with invasive methods (average difference between both methods: 0.08cm^2). R3D echocardiography may improve the assessment of MS severity in patients with discordant results between different methods and in clinical scenarios where these methods have limitations, particularly after balloon valvoplasty.⁷

Magnetic resonance imaging

Magnetic resonance imaging can be used to identify the presence of valvular stenosis. The high-velocity flow across the narrow valvular orifice may be recognized as a signal void on cine MRI. Imaging may be performed with steady-state free precession (SSFP) imaging for semi-quantitative assessment of valvular dysfunction or with a standard breath-hold segmented gradient-recalled echoplanar imaging sequence (GE-EPI) (Fig. 1.8). A study by

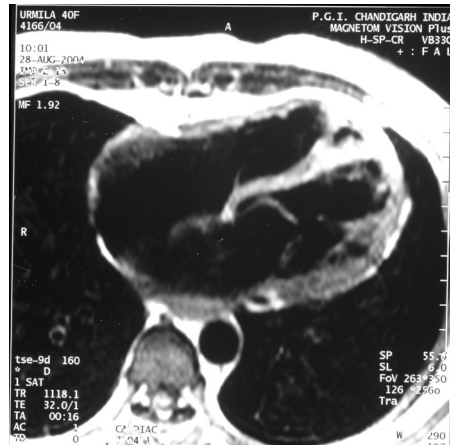


Figure 1.8 Magnetic resonance image (MRI) showing typical thickened doming mitral valve.

Krombach *et al.*⁸ demonstrated both SSFP and GE-EPI sequences to share a high sensitivity (100%), although the image quality of SSFP was rated higher than GE-EPI. Peak flow velocity across MS can be quantified using velocity-encoded cine MRI (VE-MRI), analogous to echocardiography.⁹ Lin *et al.*⁹ reported excellent correlation of MVA by VE-MRI and Doppler echocardiography in 17 patients with MS ($r = 0.86$). An important strength of MRI for evaluation of mitral stenosis is that visualization of the spatial configuration of the mitral valve is excellent and quantification of transvalvular flow jet is unrestricted by echo windows. None the less, the MRI measurement is subject to several potential inaccuracies related to marginal temporal resolution, slice thickness, and signal loss.

Conclusions

In summary, 2D Doppler echocardiography gives sufficient information for imaging of MS. TEE has additional value for the evaluation of LA clot, and in some cases for assessment of mitral regurgitation and subvalvular pathology that is not clear on 2D echo. Of the newer imaging modalities, MRI and 3D echocardiography appear promising, and in selected cases they will avoid invasive (transesophageal or catheterization) studies.

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