Anatomical features of rheumatic and non-rheumatic mitral stenosis: Potential additional value of three-dimensional echocardiography

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Summary Although mitral stenosis is mostly due to rheumatic fever, other etiologies, such as degenerative, congenital, drug- or radiotherapy-induced mitral stenosis, are emerging and need to be recognized in order to decide the best therapeutic options. This pictorial review describes the echocardiographic features of these different anatomical types and the additional value of three-dimensional echocardiography.

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Abbreviations: 2D, two-dimensional; 3D, three-dimensional; MS, mitral stenosis; MVA, mitral valve area; PMC, percutaneous mitral commissurotomy; TEE, transoesophageal echocardiography; TTE, transthoracic echocardiography.

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Background

Echocardiography plays a major role in the evaluation and decision-making process in patients with mitral stenosis (MS), allowing for confirmation of diagnosis, evaluation of severity and choice of best therapeutic option (feasibility of a percutaneous mitral commissurotomy [PMC] or surgical valve replacement) [1]. MS is mostly due to rheumatic fever but other aetiologies are emerging and should be recognized, as these forms are usually not suitable for PMC. This review aims to present the different aetiologies of MS, their echocardiographic characteristics and the potential value of three-dimensional (3D) echocardiography (Table 1).

Rheumatic mitral stenosis

Rheumatic MS is a frequent cause of valve disease in developing countries and remains a significant problem in Western countries despite the striking decrease in the prevalence of rheumatic fever, as the consequence of immigration from developing countries. Thus, according to the Euro Heart Survey, MS still accounted for 12% of native valvular heart disease [2]. Commisural fusion is the main mechanism of rheumatic MS. Associated lesions are chordal shortening and fusion, and leaflet thickening. Mobility of the posterior valve is almost always reduced whereas mobility of the anterior valve is often preserved. Later in the disease course, superimposed calcification may contribute to the limitation of leaflet motion.

Commisural splitting is the main mechanism by which the mitral valve area (MVA) increases after PMC. Commisural opening should be evaluated in parasternal short-axis and has been shown to be an important predictor of long-term functional outcome after PMC [3]. The achievement of a complete — ideally bilateral — commisural opening is thus a major goal during PMC. Our group has previously shown that 3D echocardiography provides a better assessment of the degree of commissural opening [4]. Fig. 1 presents different forms of rheumatic MS with no, unilateral and bilateral commissural opening in two-dimensional (2D) transthoracic (TTE) and corresponding 3D transoesophageal echocardiography (TEE) views.

| Table 1 | Anatomical characteristic of the different etiological types of mitral stenosis. |
|---------|-------------------------------|----------------|----------------|
| Etiology                                      | Commissures            | Mobility                  | Calcification                    |
| Rheumatic mitral stenosis                     | Fused                | Restrictive motion of the posterior valve | 0 to +++ (Possible)               |
| Native valve                                  | At least partially fused | Restrictive motion of the posterior valve | 0 to +++ (Possible)               |
| Restenosis due to commissural refusion        | At least one commissure completely open | Restrictive motion of the posterior valve | 0 to +++ (Possible)               |
| Restenosis due to valve rigidity              | Both commissures open | Normal mobility of the tip of both leaflets | +   +++ Important calcifications of the mitral annulus and the base of both leaflets |
| Degenerative mitral stenosis                  | Both commissures open | Absence of restrictive motion of the posterior valve | + to +++ Highly suggestive calcifications of the mitral aortic membrane |
| Post-radiation mitral stenosis                | Both commissures open | Absence of restrictive motion of the posterior valve | + to +++ Highly suggestive calcifications of the mitral aortic membrane |
| Congenital mitral stenosis                    | Not applicable (absence of commissure) | Normal                  | Usually not                      |
In addition, real-time 3D-TTE has provided accurate MVA measurements similar to 2D-TTE and, more importantly, has improved the accuracy of planimetry when performed by non-experienced operators [4]. 3D-TEE also provides accurate and reproducible MVA measurements, similar to 2D planimetry performed by experienced operators, and could be considered as a second-line alternative tool for the evaluation of MS severity in patients with poor echocardiographic windows or for teams less accustomed to evaluating MS patients [5].

After a successful PMC, late clinical deterioration may eventually occur, with a 40% rate of anatomical deterioration at 10 years [6]. Late anatomical deterioration — or restenosis — may result from commissural refusion or valve rigidity with persistent commissural opening. Distinction between these two features is of crucial importance, as a new PMC may be attempted in case of restenosis with commissural refusion with good mid-term results in selected patients [7], whereas PMC is of no use in case of valve rigidity with persistence of commissural opening. Echocardiography — most specifically 3D echocardiography — plays a major role in the assessment of the degree of commissural opening (Fig. 2).

Degenerative mitral stenosis

Degenerative MS may occur in elderly people. The main lesion is annular calcification, which usually has few or no hemodynamic consequences, unless leaflet thickening and calcification are associated. Valve thickening and calcification predominate at the base of the leaflets, whereas it affects predominantly the tips in rheumatic MS. In addition, there is no commissural fusion. Evaluation of degenerative MS is often challenging using TTE and planimetry is not always feasible or reliable. Three-dimensional TEE is of great help in confirming the absence of commissural fusion and measuring the valve area (Fig. 3). In our experience, measurement of the valve area is often not feasible using 2D echocardiography, whereas it is much easier and reliable using 3D-TEE.

Radiation-induced valvular mitral stenosis

Radiation-induced valvular disease is uncommon and affects approximately 6—15% of patients exposed to mediastinal radiotherapy [8]. On average, valve lesions are diagnosed 11.5 years after radiation therapy and symptoms occur 5 years later [9]. Valvular regurgitations are more common and aortic stenosis is the main reason for surgery. MS is rare and appears many years after the original exposure. Typical echocardiographic features involve thickening and calcification of both aortic and mitral valves. Calcifications of the mitral aortic membrane have been reported to be highly suggestive of radiation-induced lesions [10]. Of importance, there is no commissural fusion (complete bilateral commissural opening) and the posterior leaflet is mobile. Again, 3D echocardiography, which provides direct and intuitive visualization of the mitral valve, clearly shows the absence of commissural fusion, precluding PMC (Fig. 4).
**Figure 2.** Restenosis due to valve rigidity several years after a percutaneous mitral commissurotomy in a patient with a rheumatic mitral stenosis. Both commissures are open (arrows). (A) Two-dimensional transthoracic echocardiography. (B) Three-dimensional transthoracic echocardiography.

**Figure 3.** Degenerative mitral stenosis. The annulus as well as the valve leaflets are thickened and calcified. In contrast, both commissures are open. Transthoracic echocardiography shows poor image definition (A), while three-dimensional transoesophageal echocardiography (B) allows more accurate assessment of the degree of commissural opening and measurement of the mitral area (planimetry) (C).

**Figure 4.** Radiotherapy-induced mitral stenosis. Complete bilateral commissural opening (arrows) in (A) two-dimensional transthoracic echocardiography and (B) three-dimensional transoesophageal echocardiography.

**Congenital mitral stenosis**

Congenital MS is rare, being identified in 0.6% of autopsied hearts with congenital heart disease and in 0.2–0.4% of clinical series. It is usually associated with other cardiac defects. It may result from any or combinations of supravalvular ring (complete or incomplete), annular hypoplasia, leaflet abnormalities (leaflet retraction,
commissural fusion), chordae abnormalities (shortness, absence of chordae) or papillary muscle abnormalities (single papillary muscle inserting into the leaflets, i.e. parachute mitral valve, hypoplastic or arch-shaped papillary muscle) [11,12]. Echocardiography shows the exact function and anatomy of the valve, guiding the therapeutic options (Fig. 5).

Other causes are exceptional, inflammatory diseases, infiltrative diseases, carcinoid disease and drug-induced valve disease (benfluorex) [13]. Leaflet thickening and restriction are common here, while commissures are seldom fused.

Conclusion

A comprehensive assessment of the anatomy of the mitral valve is crucial for the appropriate management of patients with MS. The assessment of the degree of commissural opening is of particular interest to judge the feasibility of PMC. Three-dimensional echocardiography, which provides direct visualization of the mitral valve, is particularly helpful and should be considered as part of the systematic echocardiographic examination, especially in atypical cases. It allows a comprehensive assessment of the degree of commissural opening and an assessment of the MVA in difficult cases or for teams less accustomed to evaluating MS patients.

Disclosure of interest

Éric Brochet and David Messika-Zeitoun have received lecture fees from Phillips.

References


