Rheumatic and Degenerative/Calcific MS

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Disclosures

- Speaker’s bureau, Edwards lifesciences
Rheumatic MS

- Most common cause of MS worldwide
- Immune response 2/2 bacterial infection
- Valve inflammation due to cross-reactivity between leaflet tissue and streptococcal antigen
- Begins with formation of tiny nodules along the leaflet coaptation points, then fibrin deposition on leaflets
- Over years to decades: fusion of commissures; thickening, fibrosis and calcification of leaflet cusps; thickening, fusion and shortening of chordae → domed appearance
- Regurgitant process early on, then progresses to stenosis
- Up to 75% of patients with documented recurrences of rheumatic fever have valvular disease at 45 y f/u

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### Table 6: Approaches to evaluation of mitral stenosis

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Units</th>
<th>Formula / Method</th>
<th>Concept</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valve area</td>
<td>cm²</td>
<td>tricuspid mitral orifice using 2D echo</td>
<td>direct measurement of anatomic MVA</td>
<td>- accuracy</td>
<td>- experience required - not always feasible (poor acoustic window, severe valve calcification)</td>
</tr>
<tr>
<td></td>
<td>cm²</td>
<td>pressure half-time</td>
<td>rate of decrease of transvalvular flow is inversely proportional to MVA</td>
<td>- independence from other factors</td>
<td>- dependence on other factors (AR, LA compliance, LV diastolic function) - not valid if significant MR or MR</td>
</tr>
<tr>
<td></td>
<td>cm²</td>
<td>continuity equation</td>
<td>volume flow through mitral and aortic orifices are equal</td>
<td>- independence from flow conditions</td>
<td>- multiple measurements (sources of errors) - technically difficult</td>
</tr>
<tr>
<td></td>
<td>cm²</td>
<td>PISA</td>
<td>MVA assessed by dividing mitral volume flow by the maximum velocity of diastolic mitral flow</td>
<td>- independence from flow conditions</td>
<td>- technically difficult</td>
</tr>
<tr>
<td>Mean gradient</td>
<td>mm Hg</td>
<td>$\Delta P = \frac{\sum \omega}{N}$</td>
<td>pressure gradient calculated from velocity using the Bernoulli equation</td>
<td>- easy to obtain</td>
<td>- dependent on heart rate and flow conditions</td>
</tr>
<tr>
<td>Systolic pulmonary artery pressure</td>
<td>mm Hg</td>
<td>$sPAP = 4V_{max}^{2}/RPA + RA pressure$</td>
<td>addition of RA pressure and maximum gradient between RV and RA</td>
<td>- obtained in most patients with MS</td>
<td>- arbitrary estimation of RA pressure - no estimation of pulmonary vascular resistance</td>
</tr>
<tr>
<td>Mean gradient and systolic pulmonary artery pressure at exercise</td>
<td>mm Hg</td>
<td>$dP_{max} = \frac{4V_{max}^{2}}{N} + \frac{4V_{max}^{2}}{RPA}$</td>
<td>incremental value in assessment of tolerances</td>
<td>- incremental value in assessment of tolerances</td>
<td>- experience required - lack of validation for decision making</td>
</tr>
<tr>
<td>Valve resistance</td>
<td>dyn·sec·cm⁻⁵</td>
<td>$V_{max} = \frac{P_{max}}{(CASA)<em>{L} 	imes (WVT)</em>{M}} / D_{P}$</td>
<td>resistance to flow caused by MS</td>
<td>- initially suggested to be less flow dependent, but not continued</td>
<td>- no prognostic value - no clear threshold for severity - no additional value vs. valve area</td>
</tr>
</tbody>
</table>
Rheumatic MS Evaluation by echo

- MVA planimetry (2D/3D) at leaflet tips
- Pressure ½ time (220/PHT): affected by
- Continuity equation (LV or RV SV/CW MV VTI)
- Mean gradients: affected by HR

Table 13. Stages of MS

<table>
<thead>
<tr>
<th>Stage</th>
<th>Definition</th>
<th>Valve Anatomy</th>
<th>Valve Hemodynamics</th>
<th>Hemodynamic Consequences</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>At risk of MS</td>
<td>Mild valve doming during diastole</td>
<td>Normal transmural flow velocity</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>B</td>
<td>Progressive MS</td>
<td>Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets</td>
<td>Increased transmural flow velocities</td>
<td>Mild to moderate LA enlargement</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Planimetric MVA &gt;1.5 cm²</td>
<td></td>
<td>Normal pulmonary pressure at rest</td>
<td></td>
</tr>
<tr>
<td>C</td>
<td>Asymptomatic severe MS</td>
<td>Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets</td>
<td>MVA ≤1.5 cm²</td>
<td>Severe LA enlargement</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Planimetric MVA ≤1.5 cm²</td>
<td></td>
<td>Elevated PASP &gt;30 mm Hg</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Diastolic pressure half-time ≥220 ms</td>
<td>Decreased exercise</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>with very severe MS</td>
<td>Tolerance</td>
</tr>
<tr>
<td>D</td>
<td>Symptomatic severe MS</td>
<td>Rheumatic valve changes with commissural fusion and diastolic doming of the mitral valve leaflets</td>
<td>MVA ≤1.5 cm²</td>
<td>Severe LA enlargement</td>
<td>Exertional dyspnea</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Planimetric MVA ≤1.5 cm²</td>
<td></td>
<td>Elevated PASP &gt;30 mm Hg</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>with very severe MS</td>
<td></td>
</tr>
</tbody>
</table>

The transmural mean pressure gradient should be obtained to further determine the hemodynamic effect of the MS and is usually >5 mm Hg to 10 mm Hg in severe MS; however, due to the variability of the mean pressure gradient with heart rate and forward flow, it has not been included in the criteria for severity.

LA indicates left atrial, LV, left ventricular, MS, mitral stenosis, MVA, mitral valve area, and PASP, pulmonary artery systolic pressure.

2014 ACC/AHA guidelines
Wilkins Score

- The degree of leaflet rigidity (0-4)
- The severity of leaflet thickening (0-4)
- The amount of leaflet calcification (0-4)
- The extent of subvalvular thickening (0-4)
- Better outcomes with PBMV with score <= 8 (no severe MR)


Score >= 10 independently predicts severe MR after PBMV

Table 1. Echocardiographic Score for Severe Mitral Regurgitation After Percutaneous Mitral Valvuloplasty

<table>
<thead>
<tr>
<th>IV. Subvalvular disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Normall thickness of chordae structures just below the valve</td>
</tr>
<tr>
<td>2. Thickening of chordae extending up to one-third of chordal length</td>
</tr>
<tr>
<td>3. Thickening to the distal third of the chordae</td>
</tr>
<tr>
<td>4. Extensive thickening and shortening of all chordae extending down to the papillary muscle</td>
</tr>
</tbody>
</table>

The total score is the sum of these echocardiographic features (minimum score 16).

Padial et al.
JACC Vol. 27, No. 5 1225
April 1996:1225-31
ACC/AHA Class I recs

1. Percutaneous mitral balloon commissurotomy is recommended for symptomatic patients with severe MS (mitral valve area <= 1.5 cm², stage D) and favorable valve morphology in the absence of left atrial thrombus or moderate-to-severe MR (280–284, 286, 328). (Level of Evidence: A)

Mitral valve surgery (repair, commissurotomy, or valve replacement) is indicated in severely symptomatic patients (NYHA class III to IV) with severe MS (mitral valve area <= 1.5 cm², stage D) who are not high risk for surgery and who are not candidates for or who have failed previous percutaneous mitral balloon commissurotomy (319–324). (Level of Evidence: B)

Concomitant mitral valve surgery is indicated for patients with severe MS (mitral valve area <= 1.5 cm², stage C or D) undergoing cardiac surgery for other indications. (Level of Evidence: C)
Clinical history

- 48 f
- USOH until she had a stroke at age 37, found to have AF
- Eventually discovered to have rheumatic MS
- Now with progressive symptoms

Borderline Wilkins score

The degree of leaflet rigidity (0-4): 2-3
- The severity of leaflet thickening (0-4): 2
- The amount of leaflet calcification (0-4): 2
- The extent of subvalvular thickening (0-4): 2
Total: 8-9
MVA = 0.98 cm² by 3D planimetry, continuity = 0.96 cm², PHT = 1.2 cm²

MR EROA = 14 mm²
postMVA = 1.3 cm²
No more inflations performed 2/2
moderate MR
Degenerative/calcific MS

ASA/EAE 2009 guidelines - DMS

- “It [degenerative MS] is frequently observed in the elderly and associated with hypertension, atherosclerotic disease, and sometimes AS. However, calcification of the mitral annulus has few or no haemodynamic consequences when isolated and causes more often MR than MS. In rare cases, degenerative MS has haemodynamic consequences when leaflet thickening and/or calcification are associated. This is required to cause restriction of leaflet motion since”
Degenerative/Calcific MS

- Mitral annular calcification extending onto the leaflet apparatus creating mitral stenosis
- Not much data or info on pathophysiology
- Associated with AS and CAD (atherosclerosis), age
- More common cause of MS in developed world vs developing (RHD)
- ~12-16% of MS cases are due to DMS

Transcatheter Mitral Valve Replacement in Native Mitral Valve Disease With Severe Mitral Annular Calcification

Results From the First Multicenter Global Registry
Sick population

Challenges with echo diagnosis

- Many patients in low flow states (concomitance with severe AS) so that transvalvular gradients are lower than expected for MVA
- Variable degrees of MS with severe MAC
- Difficult to see leaflets on TEE due to acoustic shadowing from MAC
- Planimetry: maximum stenosis may not be at tips of leaflets
- PHT does not work
Echo Assessment DMS

- MVA by planimetry (2D/3D reconstruction)
  - CT planimetry
- MVA by continuity (significant MR can cause underestimation)
- Gradients (low flow)

Case

- 74 f
- DM2, OSA, AF, HTN, HLD, Severe AS and CAD s/p CABG/AVR, pHTN
- During prior surgery (6 years prior), MV could not be replaced due to excessive annular calcification
- Increasing symptoms over past several months (exertional dyspnea, edema)
- Admitted for CHF exacerbation and further evaluation
Moderate MR

MVA = 0.99 cm² by Xplane
Mean gradient = 11 mm Hg

Plan

- Percutaneous mitral valve-in-MAC via MITRAL trial (investigational)
- Antegrade (transseptal) access
CT planning (annulus area 396-447 mm²)

Assess risk of LVOT obstruction (neo LVOT)

NeoLVOT area = 247 mm² (low risk)
Transmitral MG = 3 mm Hg

Mean LVOT gradient = 4 mm Hg
Thanks