MITRAL STENOSIS

Causes
1\textsuperscript{st} cause: Rheumatic heart disease (\textit{Streptococcus pyogene} pharyngitis)
- 25\% have isolated MS
- 40\% with mixed MS and MR
- 38\% with multi-valve involvement: 35\% aortic valve, 6\% tricuspid valve (rarely pulmonary valve)
- 2/3 patients are women
- Only half can recall past hx RHD
- Time from rheumatic fever and valve obstruction – few years to > 20 years
Other causes:
- Congenital (rare)
- Rare: carcinoid, LED, PAR, Fabry, \textit{Lutembacher syndrome} (ASD + rheumatic MS)
- Tumor (myxoma), thrombus, large endocarditis, large calcifications

PATHOLOGY

Valve
- Thickening and distortion of valve leaflets and chordae
- Fusion begins at commissures = MS (\textit{fish mouth})
  - Restriction of leaflet motion, especially anterior leaflet = diastolic doming
- Shortening and fusion of chordae = MR
- Stiff leaflets = MS and MR simultaneously
- \textit{Aschoff bodies} present in myocardium, not on valve (2% patients – hallmark of RHD)

Physiopathology
- \textup{LA} = \textup{pulmonary veins} = dyspnea and pulmonary edema
  - Precipitated by: tachycardia, pregnancy, $^{\uparrow}T4$, anemia, infection, AF ($^{\uparrow}\text{LA}$, $\downarrow$ diastolic filling time, $^{\uparrow}$ transmitral gradient, $\downarrow$ forward output)
- LA contraction = \textup{30\% presystolic mitral gradient} – lost in AF = $\downarrow$ CO of 20\% = symptoms (and risk of LAA thrombus)
- \textbf{Pulmonary hypertension} = $^{\uparrow}\text{LA}$ pressure = prominent A wave and small Y descent
- Gradually, $^{\uparrow}$ pulmonary vascular resistance (at exercise, than at rest) = $^{\uparrow}$ pulmonary arterial pressures = RV failure/dilatation + TR
  - Reactive pulmonary hypertension (protective mechanism – $^{\uparrow}$pre-capillary resistance = less pulmonary edema but $\downarrow$ CO)
  - Shunts between pulmonary and bronchial veins = \textbf{hemoptysis}
  - Severe MS = less pulmonary compliance + blood redistribution to the apex
- Normal LV (if pure MS without concomitant disease)

Exercise:
- Moderate MS: pulmonary edema or $\downarrow$ CO
- Severe MS: pulmonary edema and $\downarrow$ CO

LA changes (with carditis):
- LA dilatation
- Fibrosis of LA wall
- Disorganization of atrial muscle bundle
  - All contributes to development of AF

CLINICAL PRESENTATION

- $\leftrightarrow/\downarrow$CO during exercise = \textit{dyspnea, fatigue and decrease exercise tolerance}
- Reduced pulmonary compliance, interstitial edema
- **Hemoptysis** = ruptured of dilated bronchial veins due to LA pressure or ruptured pulmonary capillaries in acute pulmonary edema
  - Pulmonary hemosiderosis
- 15% patients have chest pain (2° RV hypertension)
- Palpitations (with AF – incidence of 40%)
- Ortner syndrome: compression of recurrent laryngeal nerve by dilated LA, lymph nodes or dilated pulmonary artery
- **RV failure and vascular pulmonary resistance** = systemic venous hypertension, edema, ascites, hepatomegaly

**PHYSICAL EXAM**

**GENERAL** - severe – thin, acrocyanosis, peripheral edema, mitral facies (pinkish purple cheeks – chronic low CO and vasoconstriction)

**JVP** – prominent A wave in sinus rhythm, unless AF (tall c-V wave) or PHTN - lost of x decent, RV failure (Kussmaul…)

**CAROTID**
- Normal contour
- Normal or decreased volume (low volume in severe)

**PRECORDIAL**
- S1 palpable at the apex
- P2 and PA lift at LICS 2-3
- OS – palpable b/w LLSB and apex
- Decubitus – may feel diastolic thrill

**LV impulse**
- Not increased
- locate – best place to hear murmur (especially in decubitus)

**RV impulse**
- RV lift – LICS 3-5 (with palpable P2 – confirms pulmonary HTN)

**S1**
- Loud, snapping S1 is hallmark of MS
- Direct relationship b/w audibility and intensity of S1 and OS
  - Both loud when MV mobile enough
  - When stiff, both quieter
- Loudest over apex and LLSB
- **S1 disappears as disease progresses**

**OS**
- Diagnostic of MS
  - Heard when the movement of mitral doming into LV suddenly stops – sudden tensing of valve leaflets after the cups has complete their opening in early diastole
- Med to high frequency
- Intensity doesn’t correlate to severity of MS
- **A2-OS interval**
  - Normal is 0,04-0,12 s
  - Severe MS = narrow A2-OS
- **Best heard medial to LV apex, may be better in decubitus**
- Aids to detection
o Firm pressure with diaphragm
o Mild exercise or handgrip with augment
o Best appreciated in expiration
o Inch from pulmonic area to LLSB to apex

Other findings

Split S2 because of rapid closure P2 by pulmonary HTN – S2 may become single

No B3, unless coexistent MR

Graham-Steel murmur if PR

False decreased OS
- Severe pulmonary HTN
- Large RV
- Extensive MV calcify (esp AMVL)
- CHF or very low CO
- Mixed MR/MS with dominant MR
- Very mild MS
- AS (decreased LV compliance)
- AR

Absence OS - dense MV calcification, big RV

MURMUR
- Mild MS
  o Short early diastolic murmur or only presystolic murmur
  o if both present, there is gap
- Mild to mod
  o Early and little murmurs heard readily
- Mod to severe
  o Pandiastolic rumble
  o Severe MS or fast HR, continuous gradient, long diastolic murmur
  o No presystolic murmur in AF

Contour
- Decrescendo in early diastole, crescendo in late diastole

Frequency – low pitch (rumbling)

Radiation – left axilla and lower left sternal border

Murmur length correlates to severity better than loudness

Louderness depends on velocity and severity - best heard at the apex in left lateral decubitus

Maneuver to make murmur louder
- Apex – pt decubitus – bell, GENTLY pressed, end expiration
- Coughing
- Mild exercise (sit up/ deep knee bends)
- Squat to supine or turn to left lateral
- Handgrip

Quieter diastolic rumble

Low flow - severe MS, severe pulmonary htn, CHF, Afib, especially rapid rates

Associated cardiac lesions
- AS, AR, ASD, pulmonary htn with marked RV enlarged

OS DDx

Prominent S2 split
- P2 best heard in pulm area w/o radiation exc. Phtn.
- A2P2 increase with insp; A2OS constant
- OS as loud at the apex and LSB
- OS decrease with respiration
- A2OS widens with standing position

ECG
- Left atrial enlargement ➔ P mitrale
- Right axis deviation or RV enlargement
- AF

CHEST X-RAY
- Enlarge left atrium
- Enlarged pulmonary arteries
- Mitral valve calcium
- Congestion – Kerley B lines

ECHOCARDIOGRAPHY
- Normal MVA 4-6 cm², MS when MVA 50% (2 cm²)
- Symptoms when MVA < 1,5 cm²
- Progression 0.09 cm²/year
- Restriction of diastolic motion with doming in diastole of the leaflets
  o Anterior leaflet like a hockey stick
- Contraction and fibrosis of mitral apparatus with thickening of the submitral chordae and papillary muscles
- E-F slope on M-MODE

ASE recommendations

Grade 1
- Planimeter (2D short axis) – smallest orifice in diastole
  o Greater accuracy with 3D
- Pressure half-time: time to peak trans-valvular gradient to achieve ½ value and maximum trans-valvular velocity to fall from 71% of it’s initial velocity (V max/√2)
  o PHT = 0,29 * DT ➔ MVA = 220/ PHT (ms)
  o è trans-mitral velocity flow ∝ MVA
  o Not valid in old patients, AR, diastolic dysfunction and non compliant LA
  o PHT > 220 = severe stenosis

Grade 2
- Bernoulli MVA = 0,785 * (LVOT)^2 * VTI_{LVOT} / VTI_{MV}
  o Not valid if significant MR or AR
- PISA

<table>
<thead>
<tr>
<th></th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
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</thead>
<tbody>
<tr>
<td>Valve area (cm²)</td>
<td>&gt; 1.5</td>
<td>1.0-1.5</td>
<td>&lt; 1.0</td>
</tr>
<tr>
<td>Mean gradient (mm Hg)</td>
<td>&lt; 5</td>
<td>5-10</td>
<td>&gt;10</td>
</tr>
<tr>
<td>Pulmonary artery pressure (mm Hg)</td>
<td>&lt; 30</td>
<td>30-50</td>
<td>&gt; 50</td>
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Follow-up
Repeat TTE when changes of symptoms
Repeat TTE
- Very severe MS (< 1.0 cm²): yearly
- Severe MS (< 1.5 cm²): 1-2 years
- Progressive MS (> 1.5 cm²): 3-5 years

CATHETERIZATION
- When TTE or TEE is controversial
- Transseptal approach (ideal)
- R and L catheterization
  - Mean Wedge can substitute mean LA pressure but can overestimate the true gradient due to delays in pressure changes
- Gorlin Formula (Fick or thermodilution)

EXERCISE TESTING
Recommended to evaluate the response of the mean mitral gradient and pulmonary artery pressure in patients with MS when there is a discrepancy between resting Doppler TTE findings and clinical symptoms or signs.
- Exercise can induce pulmonary hypertension and thus, influence valve gradient
- No formal place of pulmonary hypertension in the guidelines, but a rise in RV systolic pressure of more than 60-70 mm Hg should be considered in respect to the patient’s symptoms.

TREATMENT

Medical
Anticoagulation if:
  1. MS and AF
  2. MS and prior embolic event
  3. MS and left atrial thrombus

INR between 2.0-3.0
May be considered in patients with sinus rhythm when severe LA enlargement (diameter > 55 mm) or spontaneous contrast on TTE.
Cardioversion may be necessary – multiple factors for rhythm vs. rate control: duration of AF, LA size, a history of embolic event, prior AF events
In rheumatic disease – sinus rhythm difficult to achieve because of fibrosis of interatrial tracts and damage of sinoatrial node
- Penicillin prophylaxis for streptococcal β-hemolytic infections to prevent recurrent rheumatic fever in RAA MS
- Prophylaxis for endocarditis is NOT RECOMMENDED
- Diuretics and restriction sodium intake may improve symptoms.
- Beta-blocking agents and CCB may increase exercise capacity by reducing HR.
- Digitalis – no benefit but can slow HR and is of value to treat RV failure
Mitral valvotomy
Procedure of choice for the treatment of MS in symptomatic patients with severe MS (MVA ≤ 1.5 cm²) so that surgery is reserved to patients who require intervention and not candidate for valvotomy

- Inflation of balloon within orifice (via transseptal approach)
- If ABSENCE of left atrial thrombus or moderate to severe MR
- Results in commissural separation and fracture of nodular calcium
- ↓ gradient 18 to 6 mmHg, ↑ 20% CO, ↑ 2x AVM
- ↓ pulmonary resistances
- Mortality 1-2%
- Complications: stroke, cardiac perforation, severe MR (2%), 15% MR, 5% ASD

Reasonable for symptomatic patients who are at high risk of surgery, restenosis after previous commissurotomy of valvotomy.

Wilkin score: predicts the success and durability of percutaneous balloon valvotomy vs. surgical options

<table>
<thead>
<tr>
<th>Score</th>
<th>Leaflet mobility</th>
<th>Valve thickness</th>
<th>Subvalvular thickening</th>
<th>Valvular calcification</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Highly mobile, little restriction</td>
<td>Normal (4-5 mm)</td>
<td>Minimal chordal thickening</td>
<td>Single area of calcification</td>
</tr>
<tr>
<td>2</td>
<td>Decrease mobility in midportion and base of leaflets</td>
<td>Midleaflet thickening</td>
<td>Thickening 1/3 chordal length</td>
<td>Confined to leaflet margins</td>
</tr>
<tr>
<td>3</td>
<td>Forward movement of leaflets in diastole</td>
<td>Total leaflet thickening (5-8 mm)</td>
<td>2/3 chordal length thickening</td>
<td>Calcification up to mid-leaflet</td>
</tr>
<tr>
<td>4</td>
<td>No or minimal forward movement of leaflets in diastole</td>
<td>Severe thickening (≥ 8 mm)</td>
<td>Complete chordal thickening to papillary muscle</td>
<td>Calcification through all the valve leaflets</td>
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Score ≤ 8 associated with excellent immediate and long-term results – less risk of MR, better survival and less events (Braunwald p. 1498).
Calcium calcification = predictor of poor outcome
TEE should be performed before BVM to exclude LAA thrombus and confirm that MR is not moderate or severe


Surgery
Surgical options: open (direct visualization of the valve) and closed commissurotomy (blinded from LA of LV), MVR (mitral valve replacement)

MVR
- Combined MS and MR
- Extensive commissural calcification
- Severe fibrosis and subvalvular fusion
- Previous valvulotomy
Mortality between 3-8%

Prosthetic valves – associated with valve deterioration and chronic anticoagulation
- Mechanical valves preferred with patients with AF (anticoagulation)
  - Sinus rhythm and < 65 y-o (>> risk of tissue damage leading to 2nd surgery)
- Bioprosthetic valves
  - C-I to warfarin, > 65 y-o
  - Young patients may choose it for lifestyle considerations, despite risk of valve deterioration

**Senile calcified mitral stenosis**
Calcification of annulus and base of leaflets but not commissures
- No place for balloon valvulotomy
- Surgery very challenging with calcified annulus

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