MITRAL STENOSIS

Causes

1st cause: Rheumatic heart disease (*Streptocoque 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)

- ⅔ 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, **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 (fish mouth)**
  - Restriction of leaflet motion, especially anterior leaflet = **diastolic doming**
- Shortening and fusion of chordae = **MR**
- Stiff leaflets = MS and MR simultaneously
- Aschoff bodies present in myocardium, not on valve (2% patients – hallmark of RHD)

Physiopathology

- ↑ LA = ↑ pulmonary veins = dyspnea and pulmonary edema
  - Precipitated by: tachycardia, pregnancy, ↓T4, anemia, infection, AF (↑ LA, ↓ diastolic filling time, ↑ transmitral gradient, ↓ forward output)
- LA contraction = ↑ 30% presystolic mitral gradient – lost in AF = ↓ CO of 20% = symptoms (and risk of LAA thrombus)
- **Pulmonary hypertension** = ↑ LA pressure = **prominent A wave and small Y descent**
- Gradually, ↑ pulmonary vascular resistance (at exercise, than at rest) = ↑ pulmonary arterial pressures = RV failure/dilatation + TR
  - Reactive pulmonary hypertension (protective mechanism – ↑pre-capillary resistance = less pulmonary edema but ↓ CO)
  - Shunts between pulmonary and bronchial veins = **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 ↓ CO
- Severe MS: pulmonary edema and ↓ CO

LA changes (with carditis):

- LA dilatation
- Fibrosis of LA wall
- Disorganization of atrial muscle bundle
  - All contributes to development of **AF**

CLINICAL PRESENTATION

- ←→/↓CO during exercise = ***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^o 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

- Firm pressure with diaphragm
- Mild exercise or handgrip with augment
- Best appreciated in expiration
- 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

Absent OS - dense MV calcification, big RV

MURMUR
- mild MS
  - Short early diastolic murmur or only presystolic murmur
  - if both present, there is gap
- mild to mod
  - early and little murmurs heard readily
- mod to severe
  - Pandiastolic rumble
  - Severe MS or fast HR, continuous gradient, **long diastolic murmur**
  - 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
Loudness 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 cm^2, MS when MVA 50% (2 cm^2)
- Symptoms when MVA < 1,5 cm^2
- Progression **0.09 cm^2/year**
- Restriction of diastolic motion with **doming in diastole** of the leaflets
    - 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 recommandations

★ **Suggested reference:** Baumgartner H. et al. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. 2009 J Am Society Echocardiography;22:1-23.

Grade 1

- Planimeter (2D short axis) – smallest orifice in diastole
    - 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 ($V_{max}/\sqrt{2}$)
    - **PHT = 0,29 * DT → MVA = 220/ PHT (ms)**
    - ê trans-mitral velocity flow ∝ MVA
    - Not valid in old patients, AR, diastolic dysfunction and non compliant LA
    - **PHT > 220 = severe stenosis**

Grade 2

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

	Mild	Moderate	Severe
Valve area (cm^2)	> 1.5	1.0-1.5	< 1.0
Mean gradient (mm Hg)	< 5	5-10	>10
Pulmonary artery pressure (mm Hg)	< 30	30-50	> 50

★ **Suggested reference:** Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185 – **Table 13**

Follow-up
Repeat TTE when changes of symptoms
Repeat TTE

- Very severe MS (< 1.0 cm^2): yearly
- Severe MS (< 1.5 cm^2): 1-2 years
- Progressive MS (> 1.5 cm^2): 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)

$$\text{Mitral Valve Area} = \frac{\text{Cardiac Output}}{(\text{Diastolic Filling Period})(\text{Heart Rate})(37.9)(\sqrt{\text{Pressure Gradient}})}$$

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

★ **Suggested reference:** Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185.

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 RECOMMANDED
- 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 cm^2) 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 commisurotomy of valvotomy.

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

Score	Leaflet mobility	Valve thickness	Subvalvular thickening	Valvular calcification
1	Highly mobile, little restriction	Normal (4-5 mm)	Minimal chordal thickening	Single area of calcification
2	Decrease mobility in midportion and base of leaflets	Midleaflet thickening	Thickening 1/3 chordal length	Confined to leaflet margins
3	Forward movement of leaflets in diastole	Total leaflet thickening (5-8 mm)	2/3 chordal length thickening	Calcification up to mid-leaflet
4	No or minimal forward movement of leaflets in diastole	Severe thickening (≥ 8 mm)	Complete chordal thickening to papillary muscle	Calcification through all the valve leaflets

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**

★ **Suggested reference:** Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185 – **Figure 3, table 14**

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

Content of this summary from these references:

- Otto C & Bonow R. Valvular Heart Disease. (2012) In Bonow R. *et al*. Braunwald's Heart Disease, 9th edition, pp. 1468-1539. Philadelphia, PA: Elsevier.
- Nishimura R. A. et al. 2014 ACC/AHA guidelines for the management of patients with valvular heart disease. J Am Coll Cardiol 2014;63:e57-185.
- Baumgartner H. et al. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. 2009 J Am Society Echocardiography;22:1-23.