MITRAL VALVE PROLAPSE

CAUSES

- 2,4% population
- **2 women: 1 men**
- Serious MR more frequently in older men (more 50 years old)
- Diverse pathogenic mechanisms of one or more portions of mitral valve apparatus, leaflets, chordae tendineae, papillary muscle and valve annulus
- Systolic click and mid to late systolic murmur = major diagnostic criterion
- TTE: superior displacement of one or both mitral leaflets by more than 2 mm above the plane of the annulus in PSLX
    - Other findings: leaflet thickening and redundancy, excessive chordal length and motion, evidence of ruptured chords, prolapse of leaflet segments
- Familial MVP is transmitted as autosomal trait
- Seen in **MASS** (mitral, aortic, skin and skeletal) phenotype: aortic root enlargement, non specific skin and skeletal changes
    - 90% of Marfan
    - 6% Ehlers-Danlos (may be higher in type IV collagenose syndromes)
    - Also associated with osteogenesis imperfect pseudoxanthoma elasticum, congenital malformations (Ebstein, ASD ostium secundum, Holt-Oram syndrome, HCM)

PATHOLOGY

- Myxomatous proliferation of mitral valve ***leaflets*** (prominent *spongiosa* component – increase of acid mucopolysaccharide)
    - Cups, annulus and chordae tendineae can all be affected by Myxomatous proliferation
    - Myxomatous changes may result in annular dilation and calcification, further contributing to severity MR
- Fibrosis and thickening of MV leaflets, thinning and elongation of the chordae tendineae, ventricular friction lesions
    - Leaflets are grossly abnormal, redundant and prolapsed
- Sites of endothelial disruption are common sites of **endocarditis** or **thrombus formation**
- Severity of MR depends on extent of prolapse
    - Chordae rupture at sites of collagen degenerating and Myxomatous changes – increase MR
- May also affect tricuspid, aortic and pulmonary valves = **Marfan syndrome**
    - **20% MVP have prolapse in tricuspid and aortic valves on TTE**

CLINICAL PRESENTATION

- MVP is the most common cause of isolated MR requiring surgical treatment in US
- 1st cardiac condition predisposing patients to **infective endocarditis**
- Asymptomatic
- *MVP syndrome*:
    - Systolic non ejection click
    - Fatigue, palpitations, autonomic dysfunction, postural orthostatic, anxiety, neuropsychiatric symptoms
- Syncope, presyncope, palpitations, chest discomfort (prolonged, atypical, not related to exertion, brief attacks, severe stabbing at the apex) – may be related to abnormal tension on papillary muscles
- If severe MR: fatigue, dyspnea, exercise limitation
- Symptomatic arrhythmias

PHYSICAL EXAM

- Patients are slim, asthenic
- BP normal or low, orthostatic hypotension may be present
- Straight back syndrome, scoliosis, pectus excavatum
- MR may be absent to severe

Auscultation

- Supine, left decubitus and sitting position
- Diaphragm
- Non ejection systolic click at least **0,14s after S1 - LLSB**
    - Differentiated from aortic ejection click because it occurs **after** the beginning of carotid pulse upstroke
- Click can be *multiple* – produced by sudden tensing of the elongated chordae tendineae and prolapsing leaflets
- Click often followed by a **mid to late crescendo systolic murmur that continues to A_2**
    - **Duration of murmur is function of severity of MR**
    - If murmur confined to latter portion of systole = not severe
    - If MR becomes more severe, murmur commences earlier and becomes *holosystolic*
- DDX midsystolic clicks: tricuspid valve prolapse, atrial septum aneurysm, extra-cardiac factors
- **Dynamic auscultation**
    - Prolapse occurs earlier in systole, click closer to S_1 = ↓ LV volume
        - ↓ impedance of LV outflow
        - ↓venous return
        - Tachycardia
        - ↑ contractility
    - Click and onset of murmur delayed ↑ LV volume
        - ↑ impedance LV outflow
        - ↑venous return
        - ↓ contractility
        - Bradycardia
- When prolapse is severe, click is at onset of systole and may *not be audible*
    - *Straining phase of Valvalva, sudden standing: click and onset of murmur occur earlier in systole*
    - *Supine, leg raising, squatting, maximal isometric exercise, expiration: delay of the click and onset of murmur*
    - *Overshoot phase of Valsalva, prolongation R-R interval, premature contraction of AF: click and onset of murmur is delayed, murmur intensity is reduced*
    - *Manoeuvres that increase BP (exercise): intensify click and murmur*
- HCM vs MVP
    - Valsalva: ↑ intensity HCM murmur, MVP murmur is longer but not louder
    - Premature beat: ↑intensity and duration HCM murmur, MVP murmur remains unchanged or ↓

ECHOCARDIOGRAPHY

- **Prolapse**: one or both leaflets billow by at least 2 mm into LA during systole in PSLX
    - Thickening of the involved leaflet to more than 5 mm support the diagnosis
- Severe myxomatous disease (significant risk of severe MR or IE)
    - ↑ leaflet area
    - Leaflet redundancy
    - Chordal elongation
    - Annular dilation
- TEE details the integrity of the valve apparatus, LV size and function in secondary MR
- Echo findings may be present in absence of click or murmur

- Mild MR not always associated to audible murmur
- Moderate-severe MR in ⅔ **patients with posterior leaflet prolapse** and **25% with anterior leaflet prolapse**

ELECTROCARDIOGRAPHY

- Inverted or biphasic T waves
- Non specific ST changes

ARRHYTHMIA

- VBP, supraventricular and ventricular tachyarrhythmias
- AV block and SSS
- SVT is the most common tachyarrhythmia
- Increase MVP in patients with WPW
- Association between MVP and prolongation of QT interval

ANGIOGRAPHY

- Not recommended

DISEASE COURSE

- Majority remains asymptomatic for years
- Serious complications: 1%/year
- Progressive MR is the most frequent serious complication – 15% patients over 10-15 years
  - Increase LA and LV, CHF, AF, pulmonary hypertension
- Severe MR and endocarditis more frequent if:
  - Murmur and clicks
  - Thickened and redundant mitral valve leaflets (more 5 mm)
  - Men older than 50 y-o
- Endocarditis often aggravates MR and precipitates the need for surgery
- Embolic events more frequent: ½ plegia, TIA, stroke, amaurosis fugax, retinal arteriolar occlusion
  - Thrombus formation on valves (zone of endothelial tearing)
- 2X risk of sudden death (more ventricular arrhythmias)

MANAGEMENT

- TTE
  - Perform exam in 1st degree relatives of patients
- Excellent prognosis in asymptomatic patients
- Follow-up every 3-5 years
  - More frequently in patients with long murmur = evolution of disease (TTE yearly)
- No IE prophylaxis
- ECG, treadmill or holter for palpitations or syncope

TREATMENT

- Aspirin if neurologic event, AF
- MV repair if severe MR (same guidelines as MR) – possible in 90% of patients
- Operative mortality 1,6%

★ **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.

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.
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- Zoghbi W. et al. Recommendations for the evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography. 2003 J Am Society Echocardiography;16:777-802.