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 A2
  - 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 S1 = ↓ 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
  - Maneouvres 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 \( \frac{3}{5} \) 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 1\textsuperscript{st} 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%

Content of this summary from these references: