

# Ravi Vallabhan, MD – Treatment of Mitral Valve Disease



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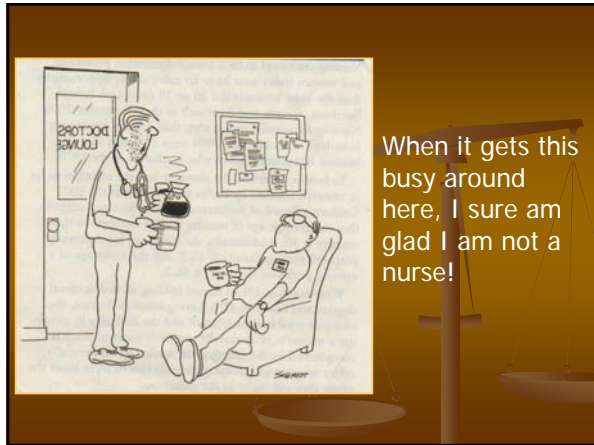
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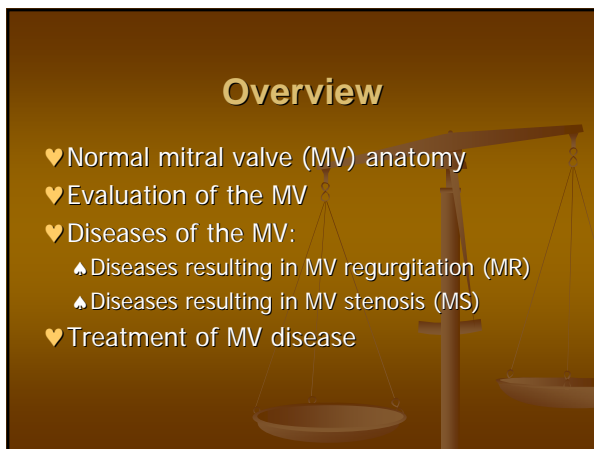
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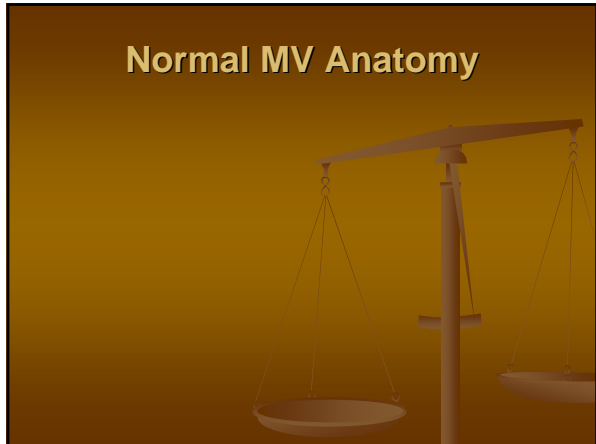
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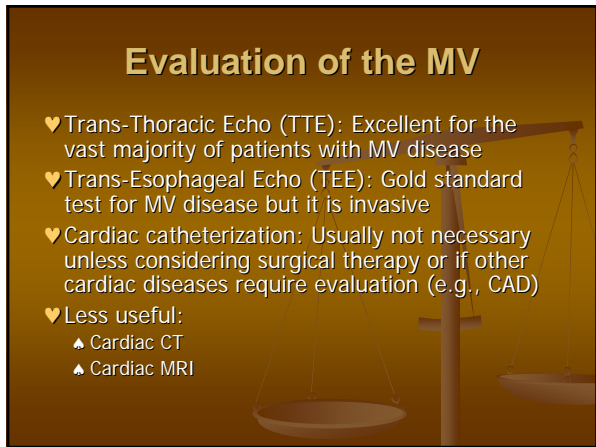
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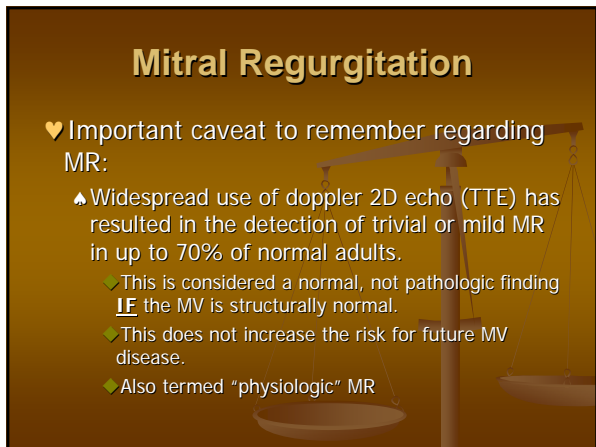
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### Mitral Regurgitation

♥ Causes of mitral regurgitation:

- ♣ Result from abnormalities which involve the MV apparatus:
  - ✔ Leaflets
  - ✔ Annulus
  - ✔ Chordae tendineae
  - ✔ Papillary muscles (and adjacent LV myocardium)
- ♣ Can be divided into 2 basic categories:
  - ✔ Primary MR
  - ✔ Secondary MR

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### Mitral Regurgitation

♥ Diseases which result in MR:

- ♣ **Primary MR:** Diseases which result in MR due to intrinsic abnormalities of the MV apparatus:
  - ✔ MV Prolapse (MVP)
  - ✔ Rheumatic heart disease
  - ✔ Infective endocarditis
  - ✔ Anorectic drugs
  - ✔ Trauma
  - ✔ Congenital (e.g., cleft mitral valve leaflet)

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### Mitral Regurgitation

♥ Diseases which result in MR:

- ♣ **Secondary** (also called **functional MR**):  
The primary abnormality is NOT the MV apparatus but the surrounding structures and MR develops as a secondary problem:
  - ✔ Ischemic heart disease
  - ✔ Dilated Cardiomyopathy (DCM)
  - ✔ Hypertrophic Obstructive Cardiomyopathy (HOCM)

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### MV Prolapse (MVP)

- ♥ Definition: systolic motion of one or both MV leaflets across the plane of the MV annulus into the left atrium (LA)
- ♥ 2 basic types of MVP:
  - ♣ Classic: true abnormality of the MV leaflets and/or chordae tendineae (MV apparatus).
  - ♣ Non-classic: an echocardiographic finding which is a variant of normal MV anatomy and NOT a true disease or condition
    - ◆ Unfortunately, many patients carry this diagnosis

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### MV Prolapse (MVP)

- ♥ Diagnosis of MVP in patients has been made based on:
  - ♣ Symptoms
  - ♣ Physical exam findings
  - ♣ 2D Echocardiography (gold standard)

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### MVP Syndrome

- ♥ Diagnosis of MVP syndrome is based on:
  - ♣ Symptoms (non-specific):
    - ◆ Atypical (non-anginal) chest pain
    - ◆ Palpitations
    - ◆ Dyspnea (shortness of breath [SOB])
    - ◆ Exercise intolerance
    - ◆ Dizziness or syncope
    - ◆ Anxiety or panic attacks
  - ♣ Physical exam findings (usually subtle):
    - ◆ Mid-systolic clicks
    - ◆ MR murmur

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### MVP

- ♥ Several patients have non-specific symptoms and physical exam findings with mild prolapse of the MV leaflets (normal variant) and are diagnosed with "MVP" but do not have a true disease process. This is **Non-classic MVP**
- ♥ A diagnosis of true or **classic MVP** cannot be made without 2D echo confirmation of pathologic MV apparatus abnormalities.

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### Classic MVP

- ♥ Most common congenital cause of MR.
- ♥ Known by several different names:
  - ♣ Click/murmur syndrome
  - ♣ Myxomatous MV disease
  - ♣ Floppy valve syndrome
  - ♣ Barlow's disease

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### Classic MVP

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### Classic MVP

♥ Pathology:

- ▲ Myxomatous degeneration of the MV leaflets and/or chordae tendineae resulting in:
  - ▶ Leaflet thickening and redundancy:
    - ▲ Anterior, posterior, or both leaflets can be involved (most common is P2: middle scallop of the posterior leaflet)
  - ▶ Thinning, stretching, and occasionally rupture of the chordae tendineae
  - ▶ Occasionally, dilatation of the MV annulus

♥ Caveat: the amount of MR does not always correlate to the degree of myxomatous degeneration.

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### Classic MVP

♥ Classification:

- ▲ Primary MVP: Idiopathic myxomatous degeneration of the MV apparatus without evidence of any systemic connective tissue disorder. Can occur as a result of:
  - ▶ A spontaneous mutation
  - ▶ Familial MVP:
    - ▲ Autosomal dominant condition with incomplete penetrance
    - ▲ Prevalence in first degree relatives ranges from 30-50%

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### Classic MVP

♥ Classification:

- ▲ Secondary MVP: Myxomatous degeneration of the MV apparatus with evidence of a systemic connective tissue disorder:
  - ▶ Marfan's syndrome
  - ▶ Ehlers-Danlos syndrome
  - ▶ Adult polycystic kidney disease
  - ▶ Osteogenesis imperfecta
  - ▶ Pseudoxanthoma elasticum
  - ▶ Similar condition is seen in some patients with bicuspid aortic valve (BAV) disease

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### Classic MVP

♥ Classification:

- ♣ Functional MVP: Prolapse of a structurally normal MV apparatus:
  - ✓ Papillary muscle dysfunction due to ischemic heart disease
  - ✓ Dilated MV annulus due to dilated cardiomyopathy (DCM)
  - ✓ LV cavity size which is too small for the MV apparatus:
    - ♣ Hypertrophic Cardiomyopathy (HCM)
    - ♣ Dehydration or decreased blood volume

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### Classic MVP

♥ Prognosis and complications:

- ♣ In general, classic MVP has a benign clinical course and good prognosis.
- ♣ However, can be associated with serious complications:
  - ✓ MR and congestive heart failure
  - ✓ Infective endocarditis
  - ✓ Arrhythmias (PACs, PVCs, AFib, SVT, or VT [rare])
  - ✓ Embolic events (e.g., CVA, TIA, etc.)
  - ✓ Sudden Cardiac Death (SCD) ??? True incidence is probably very low except in patients with ischemic heart disease.

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### Classic MVP

♥ Prognosis and complications:

- ♣ Risk for complications can vary depending upon:
  - ✓ Etiology of MVP (e.g., ischemic heart disease → ↑ risk)
  - ✓ Severity of MR (↑ MR → ↑ risk)
  - ✓ Degree of LV systolic dysfunction (↓ LVEF → ↑ risk)

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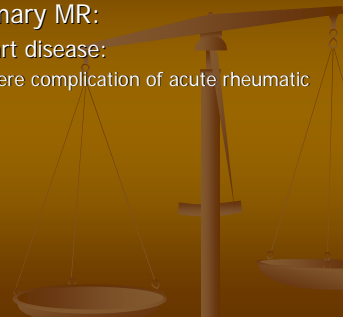
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### Mitral Regurgitation

- ♥ Etiology of primary MR:
  - ♣ Rheumatic heart disease:
    - The most severe complication of acute rheumatic fever (ARF)



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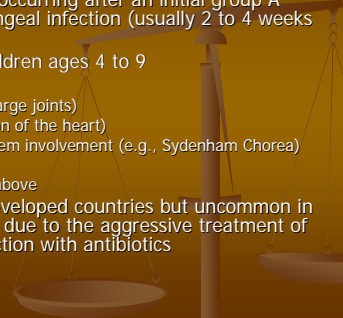
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### Acute Rheumatic Fever (ARF)

- ♥ Acute febrile illness occurring after an initial group A streptococcal pharyngeal infection (usually 2 to 4 weeks later).
- ♥ Most common in children ages 4 to 9
- ♥ Can manifest as:
  - ♣ Migratory arthritis (large joints)
  - ♣ Carditis (inflammation of the heart)
  - ♣ Central nervous system involvement (e.g., Sydenham Chorea)
  - ♣ Rash
  - ♣ Combination of the above
- ♥ Common in underdeveloped countries but uncommon in developed countries due to the aggressive treatment of the initial strep infection with antibiotics



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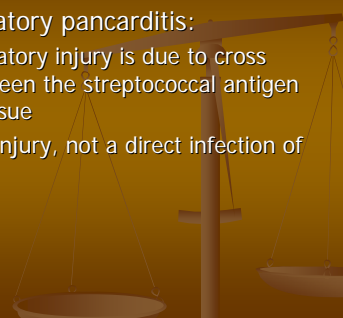
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### Acute Rheumatic Fever (ARF)

- ♥ Acute inflammatory pancarditis:
  - ♣ Acute inflammatory injury is due to cross reactivity between the streptococcal antigen and cardiac tissue
  - ♣ Auto-immune injury, not a direct infection of the heart



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## Acute Rheumatic Fever (ARF)

- ♥ Acute inflammatory pancarditis:
  - ▲ Pericardium: pericarditis
  - ▲ Epicardium
  - ▲ Myocardium: LV systolic dysfunction and congestive heart failure (CHF)
  - ▲ Endocardium
  - ▲ Valves:
    - ◆ Usually involves the MV and AV
    - ◆ Regurgitation is usually the acute abnormality
    - ◆ Stenosis of the MV and AV is rare at this stage and is more common as a late finding

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## Rheumatic Heart Disease

- ♥ Most severe late complication of ARF
- ♥ Usually occurs 10 to 20 years after the initial episode of ARF with carditis
- ♥ In patients who develop carditis with ARF, up to 50% can have valvular heart disease later in life.
- ♥ Most common valve involved is the MV:
  - ▲ Mitral stenosis (MS) is most common
  - ▲ MR is less common

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## Mitral Regurgitation

- ♥ Etiology of primary MR:
  - ▲ Infective Endocarditis:
    - ▲ Infection of a cardiac structure due to seeding
    - Usually a bacterial infection from a remote site.

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### Mitral Regurgitation

♥ Etiology of primary MR:

- ▲ Anorectic drugs (used for weight loss by suppressing appetite):
  - ▶ True incidence of drug induced valvulopathy is unknown. Data is relatively weak.
  - ▶ Pathologic features are similar to patients with Carcinoid heart disease (plaque like encasement of the leaflets with leaflet thickening) felt to be due to increased Serotonin release.
  - ▶ Specific drugs:
    - ▲ Fenfluramine (Pondimin)
    - ▲ Dexfenfluramine (Redux)
    - ▲ Phentermine (Adipex, Fastin, Ionamin): No reports of valvulopathy when used alone.
    - ▲ Combination therapy: Fenfluramine and Phentermine (Fen/Phen). Highest incidence of valvulopathy.

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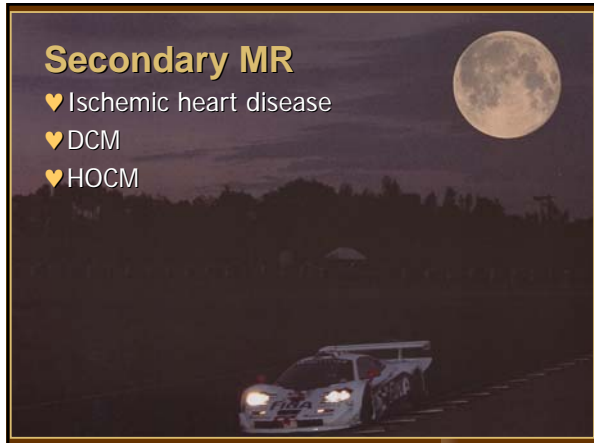
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### Secondary MR

- ♥ Ischemic heart disease
- ♥ DCM
- ♥ HOCM



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### Mitral Regurgitation

♥ Ischemic heart disease resulting in secondary (functional) MR:

- ▲ This results from a myocardial infarction involving the lateral wall of the LV.
- ▲ Posteromedial papillary muscle dysfunction or infarction.
- ▲ Retraction or rupture of posteromedial papillary muscle
- ▲ Retraction of the posterior MV leaflet or flail leaflet resulting in MR

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### Mitral Regurgitation

- ♥ DCM (LV systolic dysfunction) resulting in secondary (functional) MR:
  - ♣ As LV systolic function decreases ( $\downarrow$  LVEF), the LV chamber will dilate as a compensatory mechanism.
  - ♣ This results in enlargement of the MV annulus and incomplete coaptation of the MV leaflets and central MR.

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### Mitral Regurgitation

- ♥ HOCM resulting in secondary (functional) MR:
  - ♣ HOCM is characterized by obstruction of the LV outflow tract due to:
    - ✓ Abnormal LV septal hypertrophy (or asymmetric septal hypertrophy [ASH])
    - ✓ Systolic anterior motion (SAM) of the anterior MV leaflet
  - ♣ Results in eccentric MR toward the inter-atrial septum

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### MR

- ♥ Treatment of MR:
  - ♥ Acute
  - ♥ Chronic

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### Mitral Stenosis

♥ Etiology of MS:

- ▲ Rheumatic heart disease:
  - ▶ Most common cause of MS (> 75-90% depending upon the literature)
  - ▶ Occurs late (several years) after development of ARF. Patients in developed countries usually present in their 40's and 50's
  - ▶ Many patients (30-50%) do not report a prior history of ARF

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### Mitral Stenosis

♥ Other etiologies of MS:

- ▲ Severe mitral annular calcification (Ca<sup>++</sup>), especially in the presence of end-stage renal disease
- ▲ Infective endocarditis (MS occurs late as a result of scarring, Ca<sup>++</sup>, and fibrosis of the MV)
- ▲ Prosthetic valve dysfunction due to pannus formation, leaflet Ca<sup>++</sup>, or thrombus formation. Can be rapid in onset.
- ▲ Other very rare disorders (< 1% of cases):
  - ▶ Congenital MS, carcinoid heart disease, systemic lupus erythematosus, endomyocardial fibrosis, rheumatoid arthritis

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### Rheumatic MS

♥ Pathophysiology:

- ▲ Initial injury is formation of tiny nodules along the coapting portions of the leaflets
- ▲ With disease progression (over years [decades]):
  - ▶ Fusion of the leaflet commissures (causes "hockey stick" appearance to leaflets on echo)
  - ▶ Thickening, fusion, and shortening of the chordae tendineae
  - ▶ Thickening, fibrosis, and Ca<sup>++</sup> of the leaflet cusps.

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### Rheumatic MS

♥ Pathophysiology:

- ▲ Net effect of this damage to the MV is a stenotic MV with a symmetric, central oval shaped orifice (fish mouth) and classic “doming” of the leaflets in diastole due to fusion of the leaflet tips at the commissures.
- ▲ Decrease in MV orifice area
- ▲ The degree of leaflet thickening and Ca<sup>++</sup> and chordal involvement is variable.
- ▲ Progression of severity of the valve disease over several years is due to the stress of turbulent flow across a diseased valve, not due to recurrent episodes of ARF.

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### Rheumatic MS

♥ Pathophysiology:

- ▲ Cardiac hemodynamic consequences of MS:
  - ◆ Pressure gradient across MV between LA and LV resulting in ↑ LA pressure → ↑ pulmonary venous, capillary, and arterial pressures → pulmonary edema (congestive heart failure [CHF]) and ↑ pulmonary vascular resistance (PVR).
  - ◆ In isolated MS, LV size and systolic and diastolic pressures are normal with mild MS. However, with progression of MS, all LV parameters above will ↓ resulting in ↓ cardiac output (CO)

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### Rheumatic MS

♥ Severity of MS by MV area:

- ▲ 4 – 6 cm<sup>2</sup> : normal
- ▲ 1.5 – 2.0: mild MS
- ▲ 1.0 – 1.5: moderate MS
- ▲ < 1.0 cm<sup>2</sup>: severe MS
- ▲ Exertional dyspnea begins when MVA < 2.5 cm<sup>2</sup>
- ▲ Resting dyspnea begins when MVA < 1.5 cm<sup>2</sup>

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### Rheumatic MS

♥ Symptoms:

- ▲ With mild MS, many patients are asymptomatic
- ▲ Development of symptoms from episode of ARF was average of 16 years
- ▲ Progression from mild to severe symptoms was average of 7 – 10 years
- ▲ Are exacerbated by:
  - ◆ Tachycardia
  - ◆ ↑ Blood volume
  - ◆ ↑ Cardiac output
  - ◆ Examples: Exercising, pregnancy, AFib

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### Rheumatic MS

♥ Symptoms:

- ▲ Dyspnea
- ▲ Palpitations (usually due to atrial fibrillation [AFib])
- ▲ Hemoptysis
- ▲ Thromboembolic events

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### Rheumatic MS

♥ Complications of MS:

- ▲ AFib
- ▲ Pulmonary HTN (usually reversible)
- ▲ Thromboembolic events
- ▲ Endocarditis
- ▲ Right heart failure with functional TR
- ▲ Hemoptysis
- ▲ Pulmonary hemorrhage

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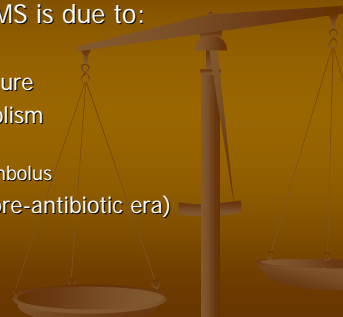
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### Rheumatic MS

♥ Mortality with MS is due to:

- ♣ CHF
- ♣ Right heart failure
- ♣ Systemic embolism
  - ♣ CVA
  - ♣ Pulmonary Embolus
- ♣ Endocarditis (pre-antibiotic era)



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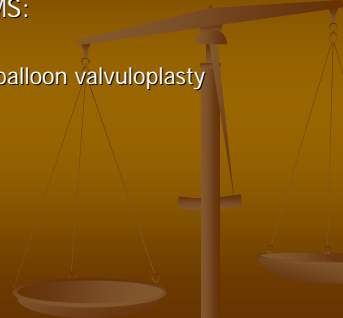
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### Rheumatic MS

♥ Treatment of MS:

- ♣ Medical
- ♣ Percutaneous balloon valvuloplasty
- ♣ Surgical



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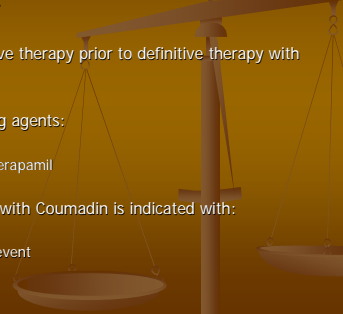
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### Rheumatic MS

♥ Treatment of MS:

- ♣ Medical:
  - ♣ Used as adjunctive therapy prior to definitive therapy with PBMV or surgery
  - ♣ Diuretics
  - ♣ AV nodal blocking agents:
    - ♣ Beta-blockers
    - ♣ Diltiazem or Verapamil
    - ♣ Digoxin
  - ♣ Anti-coagulation with Coumadin is indicated with:
    - ♣ AFib
    - ♣ Prior embolic event



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### Rheumatic MS

♥ Treatment of MS:

- ♣ Percutaneous mitral balloon valvuloplasty (PMBV):
  - ✔ Timing of PMBV or surgery is crucial:
    - ♣ Too early: Risk of the procedure with little or no short or long term benefit
    - ♣ Too late: Risk of irreversible pulmonary HTN
  - ✔ Main indication for PMBV:
    - ♣ Moderate – severe MS (MV area < 1.5 cm<sup>2</sup>)
    - ♣ Symptoms

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### Rheumatic MS

♥ Treatment of MS:

- ♣ Surgery: based on the 2006 ACC/AHA guidelines, surgery is only indicated if:
  - ✔ Patient is not a candidate for PMBV (unfavorable valve morphology)
  - ✔ LA thrombus
  - ✔ MV leaflets are non-pliable and heavily Ca++
  - ✔ Moderate (3+) to severe (4+) MR

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### Rheumatic MS

♥ Treatment of MS:

- ♣ Surgery:
  - ✔ Open commissurotomy
  - ✔ Closed commissurotomy
  - ✔ MV repair (usually with an annuloplasty ring)
  - ✔ MV replacement

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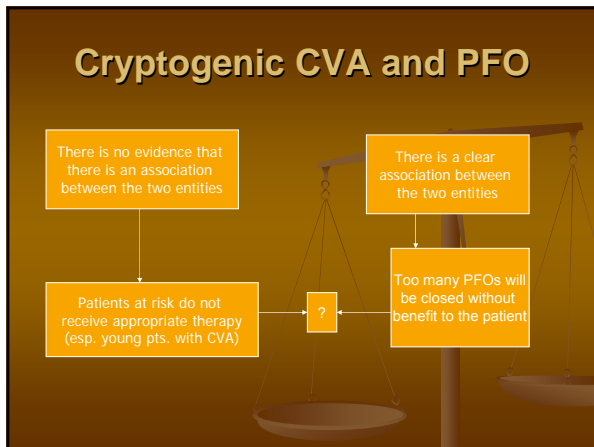
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# Ravi Vallabhan, MD – Treatment of Mitral Valve Disease

What is the problem with the data?

The treatment groups were not studied together (separate registry data) and are therefore subject to a potential selection bias.

Need definitive randomized study → CLOSURE 1 RESPECT

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## ASD

♥ Diagnosed by:

- ♣ 2D Echo
- ♣ TEE
- ♣ Right heart cath
- ♣ Cardiac MRI
- ♣ CT Angiogram

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“Want to hear something funny?”

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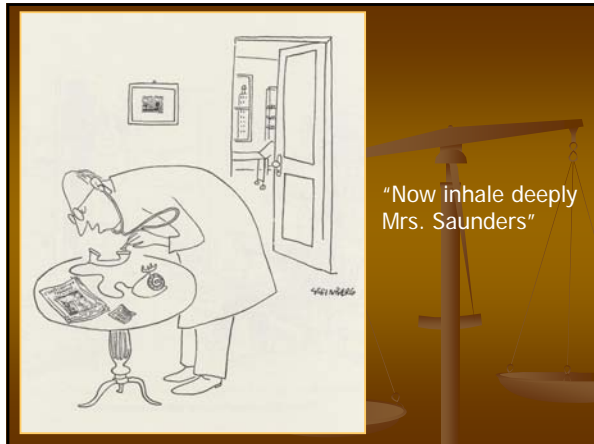
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