Aortic stenosis

Aortic valve structure

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Aortic insufficiency- aortic dissection

- Hypertension
- Connective tissue disorders
  - Marfan’s: 1 in 5000.
    - 70-85% familial - Autosomal dominant
      - *Fibrillin-1* (15q21) ~ 500 distinct mutations

Medial degeneration without atherosclerosis
Mitral stenosis

Acute Rheumatic Fever

Immunologically mediated, following 1-6 weeks after pharyngitis with Group A Streptococcus in 3% of those infected.

Antibodies against streptococcal M proteins cross-react with similar antigenic determinants in joints, heart, skin, CNS.

Acute Rheumatic Fever: Major manifestations (need 2 for diagnosis)

1. Migratory polyarthritis in 75%
2. Carditis in 50%
3. Sydenham’s chorea in 10% (involuntary, rapid, purposeless movements – caudate)
4. Erythema marginatum of skin in 10% (brown/pink with pale center)
5. Subcutaneous nodules (0.5 – 2 cm; extensor surfaces, occur late)
Acute Rheumatic Fever

**Minor Manifestations** (need 2 plus 1 Major): Previous rheumatic fever, arthralgia, fever, increased ESR or CRP, prolonged P-R interval.

**With 2 Major or Major/2Minor**
Supportive evidence of streptococcal infection: ASO Ab, anti-DNAase B, positive throat culture

**Excellent prognosis: 1% mortality - cardiac**

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Acute Rheumatic Fever

**Pancarditis**

Pericarditis

Myocarditis (Aschoff body)

Endocarditis with sterile vegetations

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[Aschoff body of acute rheumatic carditis: a cardiac granuloma]
Chronic Rheumatic Heart Disease
Follows Acute Rheumatic Heart Disease by decades. MV 65-70%
MV & AV 25%
Mitral stenosis, aortic stenosis.
Arrhythmias (A. fib. with left atrial enlargement).
Heart failure.
Predisposed to infective endocarditis.
Chronic Rheumatic Heart Disease: “Fishmouth” deformity of the mitral valve
Mitral Valve Prolapse

- Most are asymptomatic
- Mid-systolic click
- Rarely: chest pain, dyspnea, fatigue, or psychiatric manifestations
- ~3% have complications:
  - Infective endocarditis
  - Mitral insufficiency
  - Thrombus formation with embolization
  - Arrhythmia/sudden death (unknown mechanism)
Endocarditis
Non-bacterial thrombotic endocarditis (NBTE)

- Small (1-5 mm) sterile masses of fibrin on lines of closure.
- Hypercoagulable state: patients are often debilitated (“marantic” endocarditis) or may have malignancy (mucinous adenocarcinoma), renal failure, sepsis.
- May embolize to brain, kidney, heart.
- With bacteremia, may be converted to infectious endocarditis.
Heart Valve Prostheses: Complications

- Mechanical: component failure
- Bioprostheses: degeneration with calcification
- Both: Paravalvular leak, endocarditis.

Thrombosis and tissue overgrowth are less common in bioprostheses than in mechanical valves.
AV prosthesis with thrombus (from aortic side)

Tissue overgrowth of MV prosthesis (from atrial side)
Endocarditis of ball valve prosthesis (Starr-Edwards)