

30. Rheumatic Heart Disease

Etiology

1. Group A Streptococci
 - a. The organism causes pharyngitis, usually tonsillitis
 - b. Rheumatogenic strains (M-18) are distinct from those which cause glomerulonephritis
 - c. Patients are invariably >3 years old (school age)
2. Major immunologic activity against streptococcal M protein with antibody formation

Acute Rheumatic Fever (Jones' Criteria)

Major Criteria Mnemonic: "**CANES**"

1. **C**arditis - pancarditis (peri-, myo-, endocarditis / vasculitis)
2. **A**rthritis, migratory, polyarticular with fevers, Jaccoud's Arthropathy (swan-neck)
3. **N**odules, Subcutaneous: firm, usually over bony prominences or tendons
4. **E**rythema marginatum - evanescent pink rash, trunk and proximal extremities
5. **S**ydenham's Chorea
 - a. Abrupt and purposeless involuntary movements, usually hands / face
 - b. May include confusion or delirium

Minor Criteria:

Arthralgia, fever, prolonged PR intervals, laboratory abnormalities

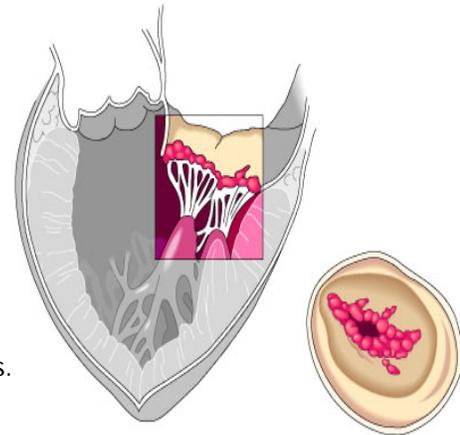
Two major or one major and two minor criteria to make diagnosis of rheumatic fever

Carditis

1. Pancarditis - any or all layers may be affected
 - Pericarditis, Myocarditis, Endocarditis (including vasculitis)
2. Valve Murmurs
 - Mitral > Aortic, Mitral regurgitation is most common acute lesion
 - Mitral stenosis is most commonly seen valve in women with RHD

Histopathology

1. Aschoff bodies are characteristic lesion (unclear etiology)
 - a. These are clear, whitish areas on organ infarction, granulomas
 - b. Damage including the influx of lymphocytes, local strep infiltration???
2. Non-Bacterial Thrombotic Endocarditis (NBTE)
 - a. NBTE lesions extremely common in RHD and cause fusion of commissures.
 - b. Fusion most common in mitral valve, second in aorta; rarely pulmonic.
 - c. Late stages show rolling, thickening and calcification



Diagnosis

1. History
 - a. Highly suspicious history occurs in only ~50% of patients with clear RHD
 - b. Erythematous rash with pharyngitis
2. Murmurs or other findings characteristic of Rheumatic Fever on examination
 - a. Mitral Stenosis most common
 - b. Aortic stenosis (usually with aortic regurgitation) may also occur
3. Carditis with polyarthritis together is most common findings (44%)
4. Complete blood count - WBC may be quite high; anemia often present
5. Elevated ESR, C-reactive protein
6. High Anti-Streptolysin O Titer (ASO) or DNase B level or hyaluronidase
7. Rare in continental USA

Treatment

1. Initial anti-streptococcal therapy (1.2mU Benzathine penicillin IM)
2. Salicylates to control fever; consider glucocorticoids in severe acute disease
3. Prophylaxis: Monthly IM injection of 1.2 mU benzathine penicillin (>5 years)
4. Alternative prophylaxis: 250mg po Pen V bid or Erythromycin 250mg po bid
5. All patients at risk for progressive valve damage should receive prophylaxis