

# 30. Rheumatic Heart Disease

## Etiology

1. Group A Streptococci most common
  - a. The organism causes pharyngitis, usually tonsillitis
  - b. Rheumatogenic strains 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" (Carditis, arthritis, nodules, erythema, Sydenham's Chorea)

1. Carditis - pancarditis (peri-, myo-, endocarditis / vasculitis)
2. Arthritis, migratory, polyarticular with fevers, Jaccoud's Arthropathy (swan-neck)
3. Subcutaneous Nodules: firm, usually over bony prominences or tendons
4. Erythema marginatum - evanescent pink rash, trunk and proximal extremities
5. Chorea (Sydenham's)
  - a. Abrupt and purposeless involuntary movements, usually hands / face
  - b. May include confusion or delirium

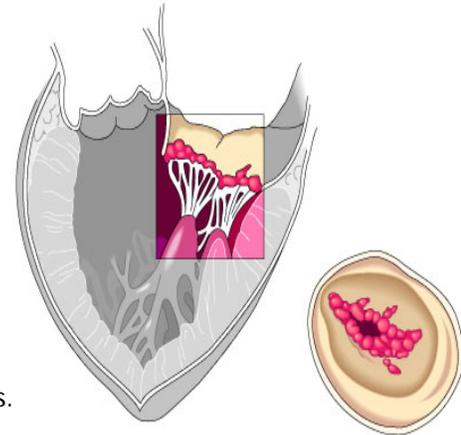
Minor Criteria:

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 are 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

## 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