Rheumatic Fever

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Surat
Objectives

- Etiology
- Epidemiology
- Pathogenesis
- Pathologic lesions
- Clinical manifestations & Laboratory findings
- Diagnosis & Differential diagnosis
- Treatment & Prevention
- Prognosis
- References
Etiology

- Acute rheumatic fever is a systemic disease of childhood.
- Group A Beta Hemolytic (GABH) Streptococcal Infection
- It is a delayed non-suppurative sequelae to URTI with GABH streptococci.
- It is a diffuse inflammatory disease of connective tissue,
- Primarily involving
  - Heart
  - Blood vessels
  - Joints
  - Subcutaneous tissue
  - CNS
Epidemiology

- Ages **5-15 yrs** are most susceptible
- Rare **< 3 yrs**
- Girls > boys
- Environmental factors
  - Over crowding
  - Poor sanitation
  - Poverty
- Incidence more during **fall & winter**
Pathogenesis

- Delayed immune response to infection with Group A beta hemolytic streptococci.
- After a latent period of 1-3 weeks, antibody induced immunological damage occur to
  - Heart valves
  - Joints
  - Subcutaneous tissue
  - Basal ganglia of brain
Group A Beta Hemolytic Streptococcus

- **Pharyngitis** – produced by GABHS can lead to:
  - Acute rheumatic fever
  - Rheumatic heart disease
  - Post streptococcal Glomerulonephritis

- **Skin infection** - produced by GABHS leads to:
  - Post streptococcal Glomerulonephritis only.
  - It will not result in Acute Rheumatic Fever nor Carditis
  - As skin lipid cholesterol inhibit antigenicity
Pathologic Lesions

- Fibrinoid degeneration of connective tissue
- Inflammatory edema
- Inflammatory cell infiltration
- Proliferation of specific cells resulting in formation of Ashcoff nodules, resulting in-
  - Pancarditis in the heart
  - Arthritis in the joints
  - Ashcoff nodules in the subcutaneous tissue
  - Basal gangliar lesions resulting in chorea
## Clinical Feature

<table>
<thead>
<tr>
<th>Major Features</th>
<th>Minor Features</th>
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<tbody>
<tr>
<td>1. Arthritis</td>
<td>1. Fever-(upto 101 degree F)</td>
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<tr>
<td>2. Carditis</td>
<td>2. Arthralgia</td>
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<td>3. Chorea</td>
<td>3. Pallor</td>
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<td>4. Subcutaneous nodule</td>
<td>4. Anorexia</td>
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<tr>
<td>5. Erythema Marginatum</td>
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</tbody>
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1. Arthritis

- Flitting
- Migratory Polyarthritis
- Involving major joints
- Commonly involved joints
  - Knee
  - Ankle
  - Elbow
  - Wrist
- Joints are tender
- In children below 5 yrs arthritis usually mild but carditis more prominent
- Arthritis do not progress to chronic disease
2. Carditis

- Pancarditis (endocarditis, myocarditis & pericarditis)
- In 40-50% of cases
- **Carditis is the only manifestation of rheumatic fever that leaves a sequelae & permanent damage to the organ**
- Valvulitis occur in acute phase
- Chronic phase
  - Fibrosis, Calcification & Stenosis of heart valves
  - Fishmouth valve
3. Chorea

- Occur in 5-10% of cases
- Mainly in girls of 1-15 yrs age
- May appear even after the attack of rheumatic fever
- Clumsiness
- Deterioration of handwriting
- Grimacing of face
- Clinical signs
  - Pronator sign
  - Jack in the box sign
  - Milking sign of hands
4. Erythema Marginatum

- Occur in <5%.
- Unique
- Transient
- Serpiginous
- Non-itchy
- More on trunks & limbs
- Lesions of 1-2 inches in size
- Pale center with red irregular margin
- Worsens with application of heat
- Often associated with chronic carditis
5. Subcutaneous nodules

- Occur in 10%
- Painless
- Pea-sized
- Palpable nodules
- Mainly over
  - Extensor surfaces of joints
  - Spine
  - Scapulae
  - Scalp
- Associated with strong seropositivity
- Always associated with severe carditis
Clinical Manifestations

- The chorea begins with distal movements of the hands
- The chorea usually is generalized
- Muscle weakness
- Milkmaid's grip (milking sign) = the pressure of the patient's grip increases and decreases continuous.
- Hypotonia = can be look like extremities paralysis.
- Speech often is abnormal
  - Sudden changes in pitch and loudness.
- No sensory loss occurs
Investigation Finding

- High ESR
- Anemia,
- Leucocytosis
- Elevated C-reactive Protein
- ASO titre >200 Todd units.
- Anti-DNAse B test
- Throat culture – GABH streptococci
- ECG- Prolonged PR interval, 2nd or 3rd degree blocks, ST depression, T inversion
- 2D Echo cardiography- valve edema, mitral regurgitation, LA & LV dilatation, pericardial effusion, decreased contractility
Diagnosis

- Rheumatic fever is mainly a clinical diagnosis
- *No single diagnostic sign or specific laboratory test available for diagnosis*
- Diagnosis based on **MODIFIED JONES CRITERIA**
# Jones Criteria (Revised) for Guidance in the Diagnosis of Rheumatic Fever*

<table>
<thead>
<tr>
<th>Major Manifestation</th>
<th>Minor Manifestations</th>
<th>Supporting Evidence of Streptococal Infection</th>
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</thead>
<tbody>
<tr>
<td>1. Carditis</td>
<td>Clinical</td>
<td>- Increased Titer of ASO</td>
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<tr>
<td>2. Polyarthritis</td>
<td></td>
<td>- Positive Throat Culture for Group A Streptococcus</td>
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<tr>
<td>3. Chorea</td>
<td>Laboratory</td>
<td>- Recent Scarlet Fever</td>
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<td>4. Erythema Marginatum</td>
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<td>5. Subcutaneous Nodules</td>
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<td>Acute phase reactants:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1 ESR</td>
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<td>2 CRP</td>
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<td>3 Leukocytosis</td>
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<td>4 Prolonged P-R interval</td>
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- Previous Rheumatic fever or Rheumatic Heart Disease - Arthralgia - Fever

- Two major criteria
- One major and Two minor criteria
- Indicates a high probability of acute rheumatic fever, if supported by evidence of Group A streptococcal infection.
Differential Diagnosis

- Juvenile Rheumatiod Arthritis
- Septic Arthritis
- Gouty Arthritis
- Sickle-cell Arthropathy
- Kawasaki Disease
- Myocarditis
- Leukemia
Treatment

Step I
- Primary prevention (Eradication of Streptococci)

Step II
- Anti inflammatory treatment (Aspirin, Steroids)

Step III
- Supportive management & management of complications

Step IV
- Secondary prevention (Prevention of Recurrent Attacks)
STEP I Primary Prevention of Rheumatic Fever (Treatment of Streptococcal pharyngitis)

1. Benzathine penicillin G
   • 600 000 U for patients
   • Intramuscular & Once only

2. Penicillin V
   • Children: 250 mg BD/TDS
   • Adults: 500 mg BD/TDS
   • Orally for 10 days

For individuals Allergic to penicillin

1. Erythromycin
   • 20-40 mg/kg/d QDS daily
   • Oral for 10 days
Step II  Anti inflammatory Treatment

For Only Arthritis

- Aspirin
- 75-100 mg/kg/day in 4 divided doses for 6 weeks

For Carditis

- Prednisolone
- 2-2.5 mg/kg/day in 2 divided doses for 2 weeks
- Taper over 2 weeks & while tapering add Aspirin 75 mg/kg/day for 2 weeks.
- Continue aspirin alone 100 mg/kg/day for another 4 weeks
Step III: Supportive management & management of complications

- Bed rest
- Treatment of congestive cardiac failure:
  - Digitalis, Diuretics
- Treatment of chorea:
  - Diazepam or Haloperidol
- Rest to joints & supportive splinting
STEP IV : Secondary Prevention of Rheumatic Fever (Prevention of Recurrent Attacks)

Benzathine penicillin G = 1 200 000 U
= Intramuscular Every 4 weeks

Penicillin V = 250 mg twice daily Oral

For individuals allergic to penicillin and sulfadiazine

Erythromycin = 250 mg twice daily Oral
Prognosis

- Rheumatic fever can recur whenever the individual experience new GABH streptococcal infection, if not on prophylactic medicines
- Good prognosis for older age group & if no carditis during the initial attack
- Bad prognosis for younger children & those with carditis with valvar lesions