

Rheumatic Fever

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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

Major Features

1. Arthritis
2. Carditis
3. Chorea
4. Subcutaneous nodule
5. Erythema Marginatum

Minor Features

1. Fever-(upto 101 degree F)
2. Arthralgia
3. Pallor
4. Anorexia

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

Diagnostic Criteria

Jones Criteria (Revised) for Guidance in the Diagnosis of Rheumatic Fever*

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

- 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 Rheumatoid 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