

Rheumatic fever: edited by 429 team

Rheumatic fever is an inflammatory disease that occurs in children and young adult (the first attack usually occurs at between 5 & 15 years of age) as result of group A β -hemolytic streptococci(pharyngeal infection “ URTI”). It affects the heart, skin, joints and central nervous system (basal ganglia) .

➤ Epidemiology:

- RF/RHD remain a major cause of morbidity and mortality in developing countries
- 15.6 millions have RHD worldwide
- 500,000 new cases of RF/yr worldwide nearly half develop carditis
- 230,000 death/yr due to RF/RHD worldwide.
- Girls > boys.
- Imposes a substantial burden on health care systems with limited budgets
- Incidence more during fall ,winter & early spring
- A disease of poverty and low socioeconomic status
- Rare in wealthy countries, due to improved living conditions, less overcrowding, and better hygiene.

➤ Etiology:

- Acute rheumatic fever represents a delayed immune response to **URTI** with **GABH streptococci**.
- Manifestation appear after a latent period of 1-3 weeks .
- It is a diffuse inflammatory disease of connective tissue, primarily involving heart, blood vessels, joints, subcut.tissue and CNS.

Causal pathway	Preventive measure
↓	Primordial prevention: Housing Hygiene
Group A streptococcal infection	
↓	Primary prevention Sore throat treatment Vaccine (unavailable) Control of skin infections (unproved)
Acute rheumatic fever	
↓	Secondary prevention (because every recurrent attack makes it more damage) Secondary prophylaxis
Rheumatic heart disease	
↓	Tertiary prevention: Medication for heart failure, Valve surgery, anticoagulation.
Cardiac failure, stroke endocarditis, death	

➤ Pathogenesis:

- It develops because of an autoimmune reaction triggered by molecular mimicry between the cell wall M proteins of the *strep. pyogenes* and cardiac myosin and laminin (antibody induced immunological damage).
 - The condition not due to direct infection of the heart or to the production of a toxin.
 - All the three layer of heart muscle maybe affected, the characteristic lesion is **aschoff bodies** (granulomatous lesion with central necrotic area).
 - In rheumatic valves, there are **verrucous vegetations** at the leaflet margins in addition to the edematous thickening.
-

➤ Clinical features:

Acute rheumatic fever has two distinct patterns of presentation:

1. Sudden onset: fever, polyarthritis 2-4 weeks after streptococcal pharyngitis.
2. Insidious or Subclinical-mild symptoms joint pain.

⊙ Poly Arthritis

- Most common feature (80%)
- Earliest manifestation of ARF
- Mainly affecting large joints , the knee , ankle , shoulder and elbow
- Leave no permanent damage
- Migrating fleeting polyarthritis
- Duration : short < 1 week
- Respond well to salicylates
- Doesn't progress to chronic disease

⊙ Carditis:

- A chronic condition, Occurs in 40-50% of cases.
- Mainly affect the aortic & mitral valve.
- Pancarditis (all layers of heart).
- Only manifestation of ARF that leaves permanent damage.
- Murmurs of MR or AR may occur in acute stage while mitral stenosis occurs in late stage.
- Cardiomegaly and CHF may occur.
- Appearance of a pericardial effusion and ECG changes of pericarditis (raised ST segment)or myocarditis (inverted or flattened T wave).
- First degree or greater AV block or other Cardiac arrhythmias .

⊙ Sydenham chorea

- 5-10% is involvement of CNS.
- Mainly in girls of 1-15 yrs age.
- Abrupt purposeless involuntary movement of muscles of face, neck, trunk and limbs
- Clinically manifest as-clumsiness, deterioration of handwriting, emotional lability or grimacing of face
- May appear even 6 months after the attack

⊙ **Erythema marginatum:**

- Transient
- Pink Rash with red borders center is pale
- Rare: present in 5%.
- Mostly on the trunk and limbs coalesce into crescent or ring shaped
- non-itchy & Worsens with application of heat.
- Associated with chronic carditis

⊙ **Subcutaneous nodules:**

- 10%, painless, pea sized, hard nodules beneath the skin .
- May also occur, particularly over tendons, joints and bony prominences.
- Usually 0.5-2 cm
- Short lived : last for few days
- Associated with strong seropositivity
- Always Associated with severe carditis

⊙ **Minor Features:**

- Fever
 - Arthralgia
 - Previous RF or RHD
-

➤ **Laboratory Findings:**

- High ESR.
 - Anemia, leukocytosis.
 - Elevated C-reactive protein.
 - ASO titer >200 Todd units. (Peak value attained at 3 weeks, then comes down to normal by 6 weeks).
 - Throat culture-GABH streptococci.
 - ECG- prolonged PR interval, 2nd or 3rd degree blocks, ST depression, and T inversion.
 - 2D Echo cardiography- valve edema, mitral regurgitation, LA & LV dilatation, pericardial effusion, decreased contractility.
-

➤ **Diagnosis:**

• **(MODIFIED JONES CRITERIA)**

2 major criteria

OR *1 major & 2 minor + evidence of group A β hemolytic streptococci.*

⊕ **Major criteria :**

1. Carditis
2. Polyarthrititis
3. Chorea
4. Erythema marginatum
5. Subcutaneous nodules

⊞ **Minor manifestations:**

1. Fever
 2. Arthralgia
 3. Previous rheumatic fever or rheumatic heart disease
 4. Increased concentration of ESR/C-reactive protein (non-specific indicators of inflammation).
 5. Leucocytosis
 6. Prolonged PR interval on ECG
 7. Evidence of antecedent group A streptococcal infection:
 - Positive throat culture (antigen test positive for group A streptococcus).
 - Elevated antistreptolysin O titer or other streptococcal antibodies .
 - History of recent scarlet fever.
-

➤ **Differential diagnosis:**

- Juvenile rheumatoid arthritis
 - infective endocarditis
 - Sickle cell arthropathy
 - Lupus
 - Myocarditis
 - Reactive arthritis
 - Leukemia.
-

➤ **Treatment of ARF:**

- Bed rest
 - Salicylates: aspirin:
 - 75-100 mg/kg/day given as 4 divided doses for 6-8 weeks
 - Attain a blood level 20-30 mg/dl to prevent salicylate toxicity.
 - Prednisolone:
 - 2 mg / kg /day taper over 6 weeks
 - Given when there is carditis to prevent inflammation.
 - Treat heart failure if present
 - Valve replacement later in life once symptoms develop or LV dysfunction occurs from severe valve regurgitation or valve stenosis
 - Secondary prevention of rheumatic fever (prevention of recurrent attacks)
 - Patients with a history of rheumatic fever should receive antibiotic prophylaxis with erythromycin or amoxicillin for dental/GI/genitourinary procedures .
 - Duration of secondary rheumatic fever management:
 - Rheumatic fever with carditis and residual heart disease has to have long duration of treatment at least 10 years since last episode or until age of 40 years or life long.
 - Rheumatic fever without carditis 5 years of treatment until 21 years
-

➤ Prognosis:

- Prognosis is good if recurrence is prevented by continuous antibiotic prophylaxis- particularly if no carditis in the initial attack.
 - If carditis, half of them can develop chronic rheumatic heart disease. Recurrence following streptococcal sore throat is high in patients with previous carditis .
 - For development of RHD, it takes 10-20 years in western world but earlier in underdeveloped world due to malignant nature of the disease.
 - Mitral valve is most commonly affected, followed by aortic and tricuspid valves.
 - So these patients need long term follow up.
-