# Acute Rheumatic Fever: Pathophysiology, Symptoms & Treatment

Acute Rheumatic Fever is an immune-mediated, multisystem inflammatory disease occurring several weeks after an untreated Group A beta-hemolytic Streptococcus (GAS) pharyngitis (sore throat), primarily affecting children aged 3 to 15 years. It is a major cause of acquired heart disease in children worldwide.

## Pathophysiology

- ARF is a **post-infectious autoimmune response** triggered by molecular mimicry.
- Antibodies produced against the **M protein** of GAS cross-react with host tissues such as the heart, joints, brain, and skin.
- This cross-reactivity results in inflammation and tissue damage.
- Only a small percentage of untreated GAS pharyngitis cases develop ARF, but the risk of recurrence increases significantly with subsequent untreated infections.
- The GAS bacteria produce enzymes and toxins (notably **streptolysin O** ), with anti-streptolysin O (ASO) antibody titers serving as a diagnostic marker.

## Clinical Features

### Diagnostic Criteria: Modified Jones Criteria

- Diagnosis requires either:
  - **2 major criteria + 1 minor criterion** , or
  - **1 major criterion + 2 minor criteria** ,
    alongside evidence of preceding GAS infection.

### Major Criteria

- Migratory polyarthritis (large joints)
- Carditis (manifested by signs such as heart failure, tachycardia, pericardial rub, or murmurs)
- Sydenham’s chorea (involuntary movements)
- Erythema marginatum (non-pruritic, serpiginous rash)
- Subcutaneous nodules

### Minor Criteria

- Fever
- Arthralgia (joint pain without inflammation)
- Elevated acute phase reactants (ESR, CRP, leukocytosis)
- Prolonged PR interval on ECG
- History of previous rheumatic fever

## Investigations

- **Anti-streptolysin O titer (ASOT):** Elevated (commonly ? 1:300)
- **Throat culture:** Positive for Group A beta-hemolytic Streptococcus
- **Erythrocyte Sedimentation Rate (ESR):** Raised
- **Chest X-ray:** May show cardiomegaly if carditis is present
- **Electrocardiogram (ECG):** To detect conduction abnormalities (e.g., prolonged PR interval)
- **Echocardiography:** Assess valvular involvement and cardiac function

## Management

### Eradication of Streptococcal Infection

- **Amoxicillin:** 250–500 mg (children 25–50 mg/kg/day in divided doses) orally, TDS for 10 days
- **Erythromycin:** 12.5 mg/kg QDS for 10 days (for penicillin-allergic patients)

### Control of Inflammation and Symptoms

- **Aspirin:** 75–100 mg/kg/day divided into 4–6 doses, gradually tapered over 2 weeks after symptom resolution
- **Chorea management:** Haloperidol 0.025 mg/kg TDS if chorea is present
- Bed rest until symptom resolution is recommended

## Prevention

### Primary Prevention

- Early diagnosis and adequate antibiotic treatment of GAS pharyngitis with:
    - **Benzathine penicillin G:** 1.2 million units IM single dose
    - OR **Phenoxymethyl penicillin:** 125–250 mg orally TDS for 10 days
- Avoid overcrowding to reduce transmission

### Secondary Prevention (Prophylaxis)

- To prevent recurrence and progression to rheumatic heart disease:
    - **Without carditis:** Benzathine penicillin 1.2 million units IM monthly for 5 years or until age 18, whichever is longer.
    - **With carditis:** Lifelong monthly benzathine penicillin or erythromycin for penicillin-allergic patients.

## Patient Education

- Importance of adherence to prophylactic antibiotics and regular follow-up
- Educate about the risk of rheumatic heart disease as a serious complication
- Emphasize early treatment of sore throats to prevent ARF