ACUTE RHEUMATIC FEVER

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INTRODUCTION

- An abnormal immune response to a GAS infection leads to an acute inflammatory illness that most commonly affects the joints, heart, brain or skin.
- Major public health problem among children and young adults in developing countries.
- It is most important acquired heart disease in children and commonly found in 4 to 15 years of age with incidence rate 5.0 / 1000 approximately.

- ACUTE RHEUMATIC FEVER is an acute autoimmune collagen disease ,occurs as a hypersensitive reaction to Group A beta hemolytic streptococcal(GAS) infection.
- It is characterized by inflammatory lesions of connective tissue and endothelial tissue.
- It effects heart, joint, blood vessels, subcutaneous tissues and brain.

INCIDENCE

- The incidence of rheumatic fever is closely related the incidence of group A STREPTOCOCCAL PHARYNGITIS.
- AGE : 5-15 YEARS ARE MOST SUSCEPTIBLE.
- RARE: Below 3 YEARS
- GIRLS and BOYS : BOTH SEXES ARE EQUALLY AFFECTED.
- COMMON: UNDER DEVELOPED COUNTRIES.
- ENVIRONMENTAL FACTORS: OVERCROWDING, POOR SANITATION, POVERTY.
- INCIDENCE MORE DURING: WINTER SEASON, EARLY SPRING

AHA Scientific Statement March 9, 2015

- ***low risk* ARF incidence <2 per 100 000 school-aged(5–14 years old)** *per year or an all aged prevalence of RHD* **of ≤1 per 1000 population per yea**r *(Class IIa; Level of Evidence C)*

AHA Scientific Statement March 9, 2015

- *Children not clearly from a low-risk population are at moderate to high risk depending on their reference population (Class I; Level of Evidence C*

PATHOPHYSIOLOGY

- It is considered as a sort of hypersensitivity reaction.
- There is an antigen antibody reaction usually following streptococcal sore throat.
- 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.

Key morphologic features of acute rheumatic heart disease.

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

JONES CRITERIA

JONES CRITERIA

18.96 Jones criteria for the diagnosis of rheumatic fever

Major manifestations

- Carditis
- Polyarthritis
- Chorea
- Erythema marginatum
- Subcutaneous nodules

Minor manifestations

- Fever
- Arthralgia
- Previous rheumatic fever
- Raised ESR or CRP
- Leucocytosis
- First-degree AV block

Plus

- Supporting evidence of preceding streptococcal infection: recent scarlet fever, raised antistreptolysin O or other streptococcal antibody titre, positive throat culture

N.B. Evidence of recent streptococcal infection is particularly important if there is only one major manifestation.

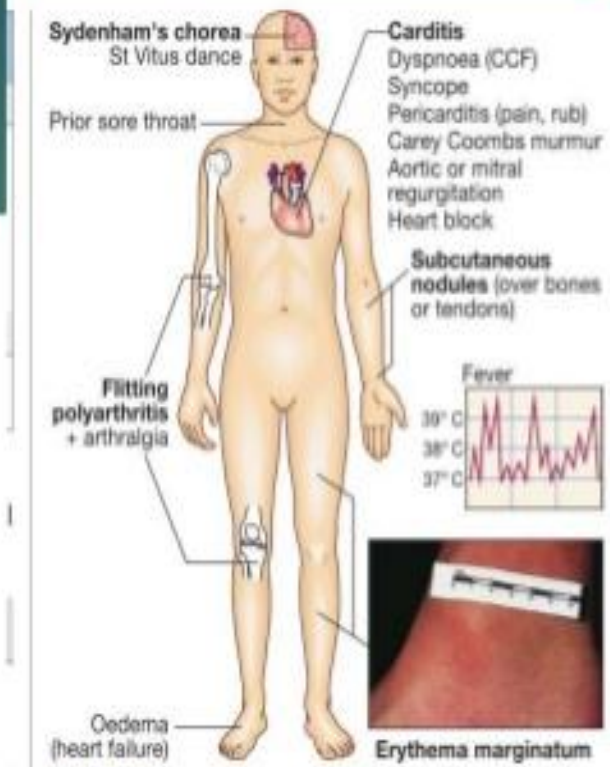


Fig. 18.86 Clinical features of rheumatic fever. Bold labels indicate Jones major criteria (CCF = congestive cardiac failure). *Inset (Erythema marginatum)* From Savin et al. 1997 – see p. 641.

REVISED JONES CRITERIA

Table 10–18. The 2015 revised Jones criteria.[1]

Population	Criteria	
	Major	Minor
Low risk	Carditis (clinical or subclinical)	Polyarthralgia
	Arthritis (polyarthritis only)	Fever (≥ 38.5°C)
	Chorea	ESR ≥ 60 mm/h or CRP ≥ 3.0 mg/dL (or both)
	Erythema marginatum	Prolonged PR interval (unless carditis is major criterion)
	Subcutaneous nodules	
Moderate and high risk	Carditis (clinical or subclinical)	Monoarthralgia
	Arthritis (monoarthritis, polyarthritis, polyarthalgia)	Fever (≥ 38°C)
	Chorea	ESR ≥ 30 mm/h or CRP ≥ 3.0 mg/dL (or both)
	Erythema marginatum	Prolonged PR interval (unless carditis is a major criterion)
	Subcutaneous nodules	

[1]For all patients with evidence of preceding group A streptococcal pharyngitis: initial acute rheumatic fever can be diagnosed when 2 major criteria or 1 major plus 2 minor criteria are met. Recurrent acute rheumatic fever can be diagnosed when 2 major or 1 major plus 2 minor or 3 minor criteria are met.

ESR, erythrocyte sedimentation rate; CRP, C-reactive protein.

Modified, with permission, from Gewitz, MH et al; American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young. Revision of the Jones criteria for the diagnosis of acute rheumatic fever in the era of Doppler echocardiography. A scientific statement from the American Heart Association. Circulation. 2015 May 19;131(20):1806–

CARDITIS

- Incidence varies from 50%-70%.
- The clinical diagnosis of carditis is based on;

  1) Presence of significant murmurs (MR/AR)

  2) Pericardial rub

  3) Unexplained cardiomegaly with CHF.

Common in young . 80% of patients develop it within first 2 weeks of RF.

- **Endocarditis/ Valvulitis** :

  Almost always associated with a murmur of valvulitis

  Valve involvments- 92 – 95% mitral valve involvement (70 – 75 % isolated MV)

  20 – 25% aortic valve involvement(5-8% isolated AV)

Arthritis/ polyarthritis

- Incidence is 35-66 %
- Most common & most earliest manifestation.
- Typically involves larger joints – knee, ankle, wrist, & elbow.
- Involved joints - hot, red, swollen, and tender.
- Migratory in nature, Not deforming .
- A dramatic response to small doses of salicylates.

CHOREA

- Incidence is 10- 30 %
- The most definite manifestation of acute rheumatc fever.
- Involuntary choreoathetoid movements primarily of face, tongue and upper extremities.
- More common in females, rare after 20 yrs.
- Emotional lability, incoordination, poor school performance, uncontrollable movements, and facial grimacing.
- Exacerbated by stress and disappears with sleep.

SUBCUTANEOUS NODULES

- Rare , 0-10%.
- Freely mobile.
- Painless.
- 0.5 - 2 cm.
- Occur in crops over bony prominences or extensor tendons.
- Common locations - elbow, wrist, knee, ankle & achilles tendon.S

ERYTHEMA MARGINATUM

- Less than 6 %.
- Erythematous, macular lesions with pale centers that are not pruritic.
- Multiple lesions primarily on the trunk or proximal extremities,rarely on distal extremities & never on face.
- It occurs early in course of RF.

MINOR CRITERIA

- 1. Polyarthralgia
- 2. Fever (>38.5 F)
- 3. Raised ESR/ CRP
- 4. Proonged PR Interval

18.97 Investigations in acute rheumatic fever

Evidence of a systemic illness (non-specific)

- Leucocytosis, raised ESR and CRP

Evidence of preceding streptococcal infection (specific)

- Throat swab culture: group A β-haemolytic streptococci (also from family members and contacts)
- Antistreptolysin O antibodies (ASO titres): rising titres, or levels of > 200 U (adults) or > 300 U (children)

Evidence of carditis

- Chest X-ray: cardiomegaly; pulmonary congestion
- ECG: first- and rarely second-degree AV block; features of pericarditis; T-wave inversion; reduction in QRS voltages
- Echocardiography: cardiac dilatation and valve abnormalities

INVESTIGATION

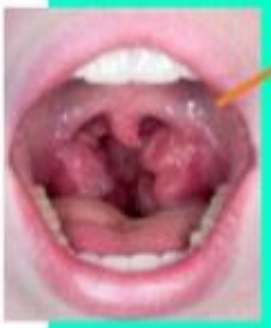
- **Evidence of Preceding Streptococcal Infection**

1.Increased or rising anti-streptolysin O titer or other streptococcal antibodies (anti-DNASE B). A rise in titer is better evidence than a single titer result.

2. A positive throat culture for group A β-hemolytic streptococci.

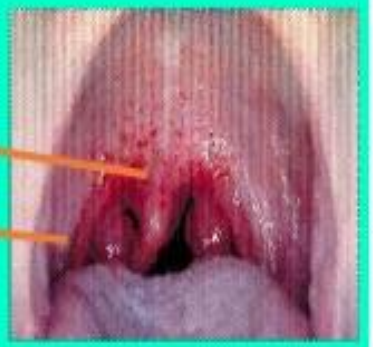
3. A positive rapid group A streptococcal carbohydrate antigen test in a child.

Features suggestive of GABHS infection

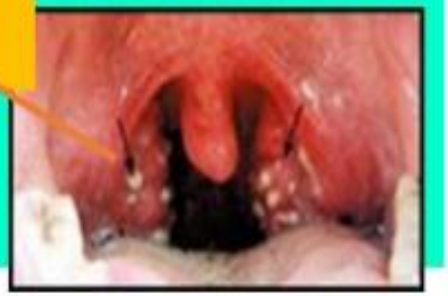


Redness & swelling of throat & tonsils;

Beefy, swollen, red uvula; Soft palate petechiae ("doughnut lesions")



Tonsillopharyngeal erythema & exudates



Sore throat: fever, white draining patches on the throat & swollen or tender lymph glands in the neck

TREATMENT

- **GENERAL MEASURES :**

  Strict bedrest

- **MEDICAL TREATMENT :**
- **Salicylats :**
- Aspirin

  Reduces fever and relieve joint pain and swelling.

  0.6- 0.9 gram 4 hourly.

- **PENICILIN :**
- Benzathine penicillin 1.2 million units, IM, once.
- Procaine penicillin 600,000units daily for 10 days.
- Erythromycin : 40mg/kg/day.

- **Corticosteroids:**
- Rapid improvement of joint symptoms.
- When response to salicylates is inadequate.

SECONDARY PREVENTION

Who should receive prophylaxis?

Patients with documented history of rheumatic fever, Including those with isolated chorea & those without evidence of rheumatic heart disease MUST receive prophylaxis.

Why should receive prophylaxis ?

1. Prevent GABHS infection sequele .
2. Prevent the repeated development of ARF.
3. Prevent the development of RHD.
4. Reduce the severity of RHD.
5. Reduce the risk of death from severe RHD.

How long ?

CATEGORY	Duration after last attack
Rheumatic fever without carditis	At least for 5 yr or until age 21 year, whichever is longer.
Rheumatic fever with carditis but without residual heart disease (no valvular disease)	At least 10 yr or until age 21 year , whichever is longer.
Rheumatic fever with carditis & residual heart disease (persistent valvular disease)	At least 10 yr or until age 40 yr, whichever is longer; sometime lifelong.