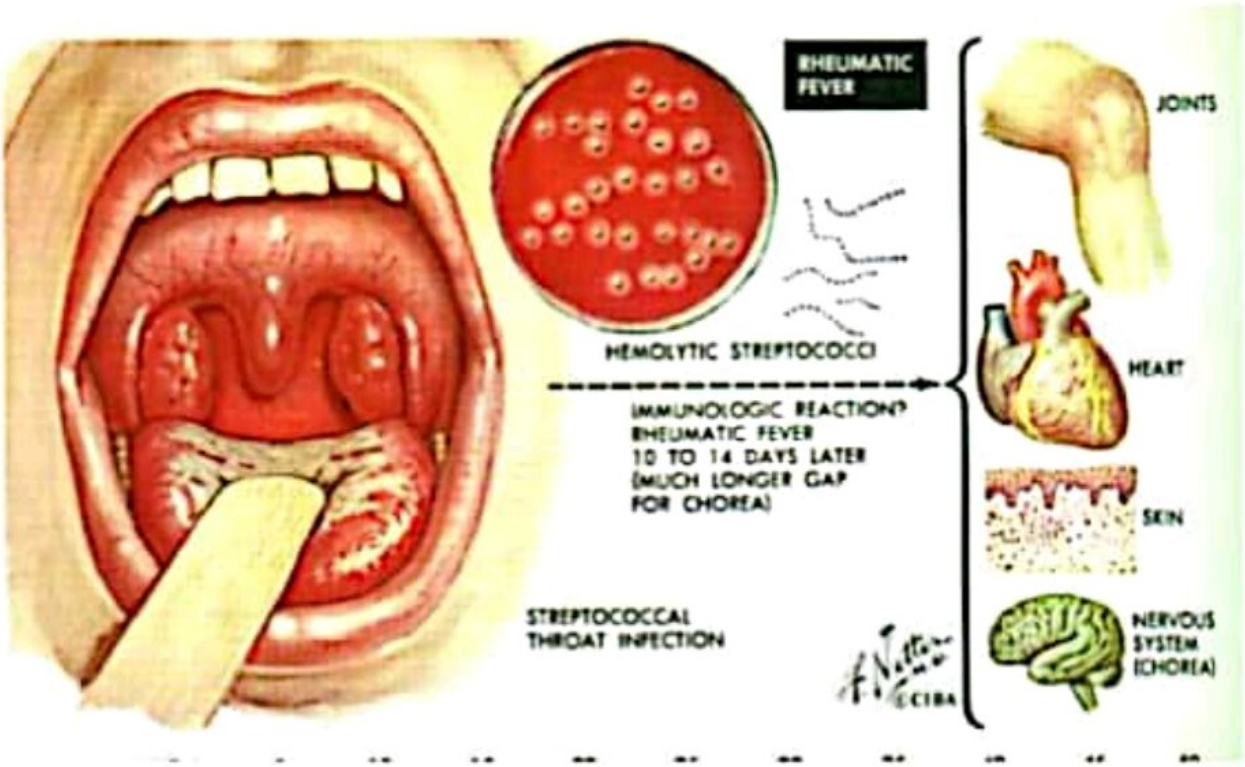
ETIOLOGY

- **Rheumatic fever** is an **immunologically** mediated **inflmmatory** disorder, which occurs as a sequel to group A streptococcal pharyngeal infection.
- **Multisystem** disease affecting the heart, joints, brain, cutaneous and subcutaneous tissues
- acute rheumatic fever remains an important **preventable** cause of cardiac disease.
- It is most common in children 6 to 15 years old.
- A family history of rheumatic fever and lower socioeconomic status are additional factors.

Rheumatic fever-pathogenesis



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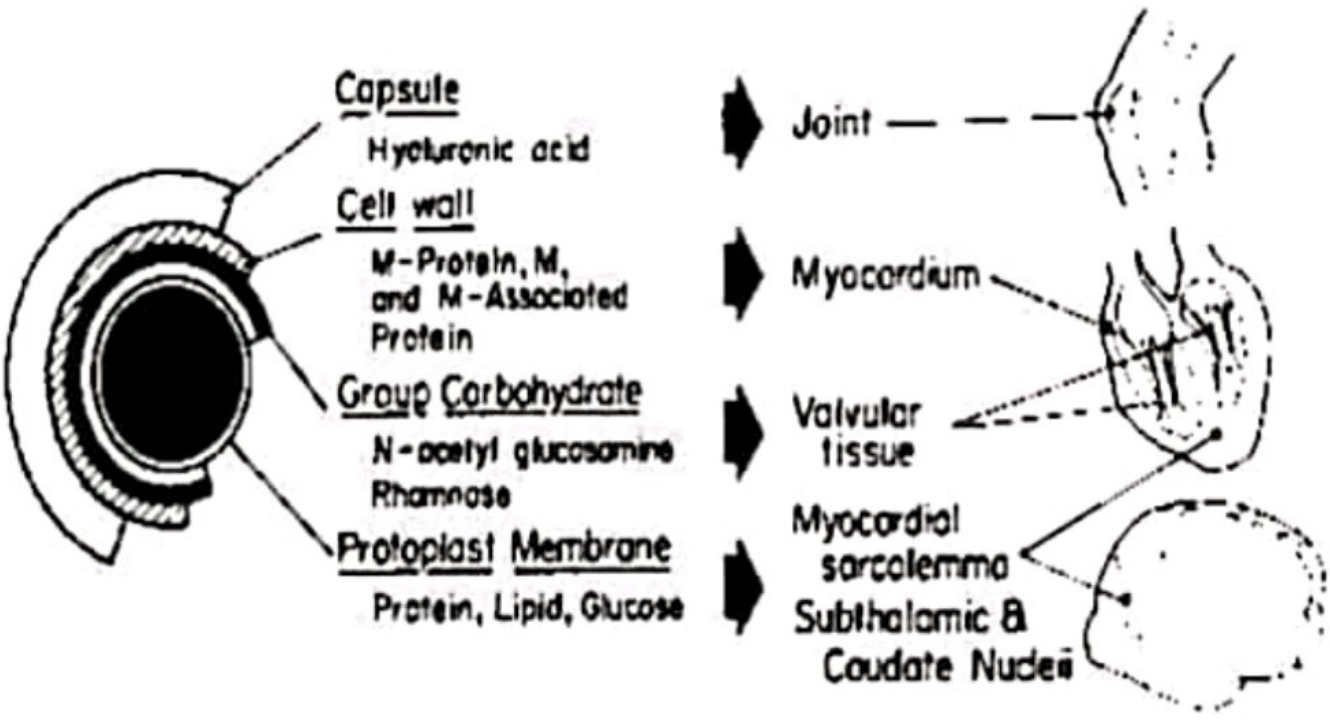


Pathogenesis

- There is no direct invasion to the tissue by the microorganism but its an auotoimmune disease that involves Ag-Ab interaction.
- It is due to an immunologic reaction that is a delayed sequela of group A betahemolytic streptococcal infections of the pharynx.
- It must be pharyngeal infection not skin infection.

- **Group A streptococcal(GAS) pharyngeal infection**
- Body produce **antibodies** against streptococci ->
- These antibodies **cross react with human tissues** because of the **antigenic similarity** between streptococcal components and human connective tissues (molecular mimicry)[there is certain amino acid sequence that is similar between GAS and human tissue]->
- **Immunologically mediated inflamation &**

CLINICAL MANIFESTATIONS

- The infection often precedes the presentation of rheumatic fever by 2 to 6 weeks.
- Acute rheumatic fever is diagnosed using the **revised Jones criteria**, which consist of clinical and laboratory findings.
- **One major and two minor**, or **two major** with **evidence of recent group A streptococcal disease** strongly suggest the diagnosis of acute rheumatic fever.

Major Criteria in the Jones System for Acute Rheumatic Fever

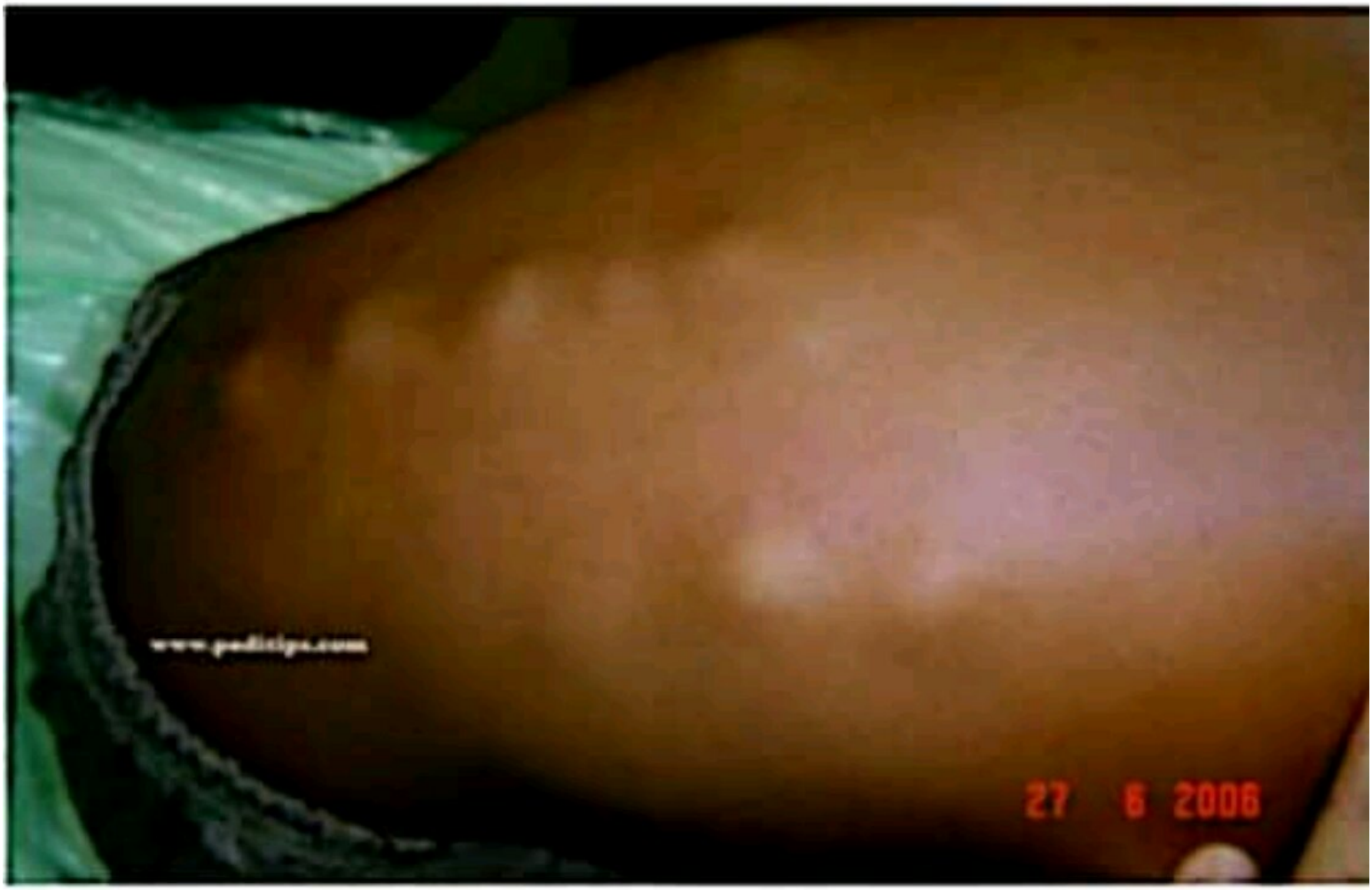
Sign	Comments
Polyarthritis	Common; swelling, limited motion, tender, erythema
	•migratory, large joints, no residual deformity, rapid response to aspirin(if aspirin given,24 to 48hrs joint pain will disappear ;thus used as diagnostic test)
Carditis	Common; pancarditis, valves, pericardium, myocardium
	•Murmur(mitral or aortic regurgitation-endocardium involved) •Heart failure •Cardiac enlargement(myocardium involvement) •Pericardial rub or effusion(pericardium involvement)
Chorea (Sydenham disease)	Uncommon; presents long after infection has resolved; more common in females, Spasmodic, unintentional, jerky choreiform movements, speech affected, fidgety, late manifestation
Erythema marginatum	Uncommon; pink macules, ring or crescent shaped, transient patches over trunk and limbs, elicited by application of local heat; nonpruritic
Subcutaneous nodules	Uncommon; Painless, hard nodules beneath skin, over bony prominence, tendons and joints, present over extensor surface of elbows, knees, knuckles, and ankles or scalp and spine, associated with repeated episodes and severe carditis;

- **Minor criteria include**
- **Clinical finding:**
- **fever (38.2°C to 38.9°C),**
- **Arthralgia(joint pain without swelling)**
- **previous rheumatic fever**
- **Laboratory finding:**
- **elevated erythrocyte sedimentation rate**
- **C-reactive protein,**
- **ECG: prolonged P-R interval.**

Evidence of recent group A streptococcal disease

- **Supporting evidence for antecedent Group A streptococcal infection**
- **scarlet fever,**
- Positive throat culture *(in 25% of patients)*
- Rapid streptococcal antigen test
- Elevated or rising streptococcal antibody titer – ASO *[anti-streptolysin] or* Anti DNAseB, AH *[anti-hyoluronic acid]*

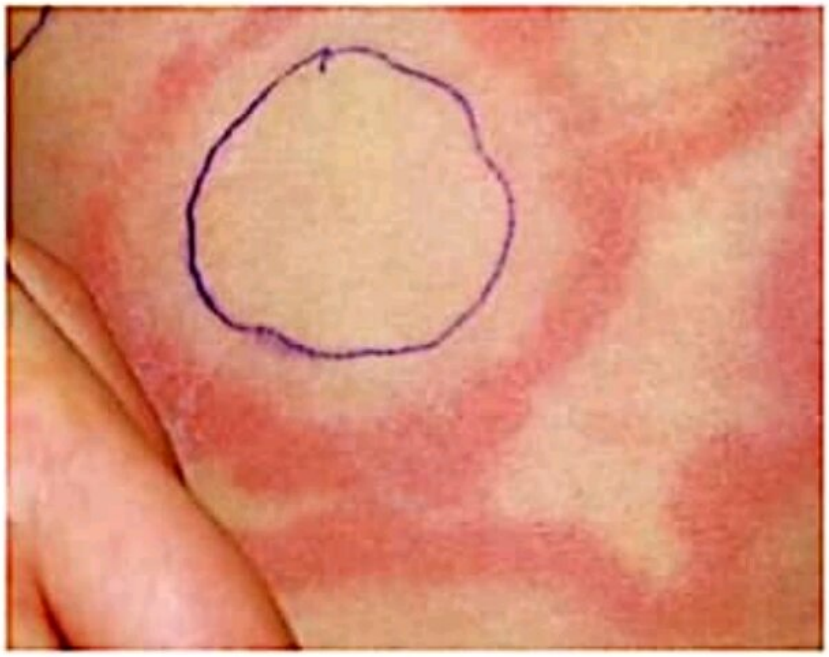
Rheumatic fever-diagnosis



Subcutaneous nodules

(nodules of rheumatoid arthritis are larger)

Rheumatic fever-diagnosis



Erythematous patches with central clearing

Erythema marginatum

Treatment

- **Bed rest** 2-6 weeks(till inflammation subsided)
- **Supportive therapy-** treatment of heart failure
- **Eradication of Organism Anti-streptococcal therapy-** Benzathine penicillin(long acting) 1.2 million units once(IM injection) or oral penicillin 10 days, if allergic to penicillin erythromycin 10 days (antibiotic is given even if throat culture is negative)
- **Anti-inflammatory agents-**

- **a. for Polyarthritis & mild carditis;** anti-inflammatory therapy with salicylates;Aspirin 100 mg/kg per day for arthritis and in the absence of carditis- for 4-6 weeks to be tapered off
- **b.For sever carditis with cardiomegally:** use steroid; Corticosteroids 1-2 mg/kg per day – for 4-6 weeks to be tapered off

Prevention

❑ **Secondary prevention – prevention of recurrent attacks**

- Benzathine penicillin G 1.2 million units IM every 4 weeks
- **Or** Penicillin V 250 mg twice daily orally
- **If allergic to both** – Erythromycin 250 mg twice daily orally

❑ **Duration of secondary rheumatic fever prophylaxis**

- Rheumatic fever + carditis life long.
- Rheumatic fever without carditis- 5 years or until 21 years whichever is longer.
- *(Continous prophylaxis is important since patient may have asymptomatic GAS infection)*

Prognosis

- R.F. may cause permanent damage to the heart but not to the joint(only arthritis) thus its said **"R.F.leaks the joints but bites the heart"**
- The prognosis of acute rheumatic fever depends on the degree of permanent cardiac damage.
- Cardiac involvement may resolve completely, especially if it is the first episode and the prophylactic regimen is followed.
- The severity of cardiac involvement worsens with each recurrence of rheumatic fever.