Rheumatic Fever

➢It is caused by Group A Streptococcus upper respiratory tract infections.

➤The incidence of both initial attacks and recurrences of acute rheumatic fever peaks in children 5-15 yrs. of age.

The onset of acute rheumatic fever.
(approximately **2-4** wk.) after GAS pharyngitis

Rheumatic fever-pathogenesis



Diagnosis of acute rheumatic fever can be established when a patient fulfills

(2 major) or (1 major and 2 minor) criteria evidence of preceding GAS infection.

The 5 Major Criteria are

- 1. Migratory Polyarthritis
- 2. Carditis
- 3. Sydenham Chorea
- 4. Erythema Marginatum
- 5. Subcutaneous Nodules

Migratory Polyarthritis :

- 1) Occurs in 75% of patients
- typically involves larger joints knees, ankles, wrists, and elbows.
 (spine, small joints of the hands and feet, or hips is uncommon).
- Rheumatic joints are classically hot, red, swollen, and exquisitely tender.
- migratory in nature; that is a severely inflamed joint can become normal within 1-3 days without treatment.
- 5) A dramatic response to salicylates is characteristic feature . If a child is suspected to have acute RF, it is useful to withhold salicylates and observe for migratory progression and the absence of such a response should suggest an alternative diagnosis.
- 6) Rheumatic arthritis is almost <u>never deforming</u>

Carditis :

- occurs in 50-60% of all cases,
- pancarditis(myocardium, pericardium, and endocardium)
- Endocarditis (valvulitis) is a universal finding in rheumatic carditis, whereas the presence of pericarditis or myocarditis is variable
- isolated mitral valvular disease or combined aortic and mitral valvular disease. (Isolated aortic or right-sided valvular involvement is uncommon).

- Carditis usually presents as tachycardia , cardiac murmurs, cardiomegaly , heart failure with hepatomegaly , peripheral and pulmonary edema
- Mitral regurgitation is characterized typically by a high-pitched apical holosystolic murmur radiating to the axilla. Aortic insufficiency is characterized by a high-pitched decrescendo diastolic murmur at the left sternal border.
- A change in the 2015 revision of the Jones Criteria is the acceptance of subclinical carditis (defined as echo evidence of valvulitis without a murmur of valvulitis) or clinical carditis (with a murmur)



Normal mitral valve Rheumatic mitral valve (with stenosis)



Sydenham Chorea :

- 10-15% of patients with acute rheumatic fever
- The latent period from acute GAS infection to chorea is usually substantially longer than for arthritis or carditis and can be months
- usually presents as an isolated, movement disorder. Emotional liability, incoordination, poor school performance, uncontrollable movements, and facial grimacing, all exacerbated by stress and disappearing with sleep.
- chorea rarely, if ever, leads to permanent neurologic sequelae.
- Clinical maneuvers to elicit features of chorea include:
- (1) demonstration of milkmaid's grip
- (2) spooning and pronation of the hands when the patient's arms are extended
- (3) wormian movements of the tongue upon protrusion
- (4) examination of handwriting to evaluate fine motor movements



Erythema Marginatum :

- is a rare (approximately 1%)
- characteristic rash of acute rheumatic fever.
- It consists of erythematous, serpiginous, macular lesions with pale centers that are not pruritic. It occurs primarily on the trunk and extremities, but not on the face, and it can be accentuated by warming the skin.



Subcutaneous Nodules

- rare ($\leq 1\%$ of patients)
- finding and consist of firm nodules approximately 1 cm in diameter along the extensor surfaces of tendons near bony prominences.
- There is a correlation between the presence of these nodules and significant rheumatic heart disease.



Minor Criteria are:

- Arthralgia (only if arthritis is not used as a major criterion)
- 2) Fever
- 3) Elevated acute phase reactants (ESR, CRP)
- 4) Prolonged P-R interval on ECG (unless carditis is a major criterion).

Recent Group A Streptococcus Infection

- An absolute requirement for the diagnosis of acute RF.
- + ve throat culture or rapid streptococcal antigen test(Streptozyme test)
- Elevated or rising serum antistreptococcal antibody titers. ASOT, anti–DNase B, antihyaluronidase



TREATMENT

• All patients with acute rheumatic fever should be placed on bed rest and monitored closely for evidence of carditis.

Antibiotic Therapy:

regardless of the throat culture results, the patient should receive 10 days of orally administered penicillin or amoxicillin or erythromycin or a single IM injection of benzathine penicillin to ensure eradication of GAS from the upper respiratory tract.

Anti-inflammatory Therapy

- Patients with polyarthritis and those with carditis without cardiomegaly or CHF should be treated with oral salicylates. The usual dose of aspirin is 50-70 mg/kg/day in 4 divided doses PO for 3-5 days, followed by 50 mg/kg/day in 4 divided doses PO for 3 wk. and half that dose for another 2-4 wk.
- Patients with carditis With cardiomegaly and/or CHF should receive corticosteroids. The dose of prednisone is 2 mg/kg/day in 4 divided doses for 2-3 wk. followed by half the dose for 2-3 wk. and then tapering of the dose by 5 mg/24 hr. every 2-3 days.

 Supportive therapies for patients with moderate to severe carditis include digoxin, fluid and salt restriction, diuretics, and oxygen

 Sydenham Chorea; Sedatives may be helpful early in the course of chorea; phenobarbital ,haloperidol ,chlorpromazine. Some patients may benefit from corticosteroids.

PREVENTION

Prevention of both initial and recurrent episodes of acute RF depends on controlling GAS infections of URT

►A. primary prevention

- Appropriate antibiotic therapy instituted before the 9th day of symptoms of acute GAS pharyngitis is highly effective in preventing first attacks of acute RF.
- Oral pencillin or erythromycin 50 mg/kg/day or single IM benzathine penicillin G 600.000 <27 kg and 1.200.000 for those >27kg

• B. Secondary Prevention

- Individuals who have already suffered an attack of acute RF are susceptible to recurrences of RF with any subsequent GAS URTI. Therefore, these patients should receive continuous antibiotic prophylaxis to prevent recurrences
 - Antibiotic prophylaxis should continue in these patients until the patient reaches 21 yrs. of age or until 5 yrs. since the last RF attack, whichever is longer. (Sometimes lifelong prophylaxis is needed for those with carditis and residual heart disease).

- The regimen of choice for secondary prevention is a single IM injection of benzathine penicillin G (600,000 IU for children weighing ≤27kg ,1.2 million IU for those weighing >27kg) every 4 wk.
- In compliant patients, oral Penicillin V 250 mg twice daily , sulfadiazine or sulfasoxazole are equally effective.
- For patient who is allergic to both penicillin and sulfonamides, a macrolide (erythromycin or clarithromycin or azithromycin) may be used