Rheumatic Fever

ETIOLOGY:

- It is caused by Group A Streptococcus upper respiratory tract infections.
- Two-thirds of patients with an acute episode of RF have history of an URT infection several weeks before, and the peak age and seasonal incidence of acute rheumatic fever closely parallel that of GAS pharyngitis.
- The disease is common in closed communities, such as boarding schools or military bases. Acute RF is associated with poverty and overcrowding.
- certain serotypes of GAS (M types 1, 3, 5, 6, 18, 29) are more frequently isolated from patients with acute rheumatic fever
- The incidence of both initial attacks and recurrences of acute rheumatic fever peaks in children 5-15 yrs. of age, the age of greatest risk for GAS pharyngitis.
- The onset of acute rheumatic fever. (approximately 2-4 wk.) after GAS pharyngitis .

CLINICAL MANIFESTATIONS AND DIAGNOSIS

Because no clinical or laboratory finding is pathognomonic for acute rheumatic fever, the diagnosis depend son Duckett Jones criteria

Diagnosis of acute rheumatic fever can be established when a patient fulfills (2 major) or (1 major and 2 minor) criteria and has evidence of preceding GAS infection.

The 5 Major Criteria are:

1.Migratory Polyarthritis :

Occurs in approximately 75% of patients with acute RF and typically involves larger joints, particularly the knees, ankles, wrists, and elbows. Involvement of the spine, small joints of the hands and feet, or hips is uncommon.

Rheumatic joints are classically hot, red, swollen, and exquisitely tender, with even the friction of bedclothes being uncomfortable.

The joint involvement is characteristically migratory in nature; that is a severely inflamed joint can become normal within 1-3 days without treatment.

Severe arthritis can persist for several weeks in untreated patients. If a child is suspected to have acute RF, it is

frequently useful to withhold salicylates and observe for migratory progression. A dramatic response to even low doses of salicylates is another characteristic feature of the arthritis, and the absence of such a response should suggest an alternative diagnosis. Rheumatic arthritis is almost never deforming. Frequently, arthritis is the earliest manifestation of acute rheumatic fever. There is often an inverse relationship between the severity of arthritis and the severity of cardiac involvement.

2.Carditis :

it occurs in 50-60% of all cases, Carditis and resultant chronic rheumatic heart disease are the most serious manifestations of acute RF and account for essentially all of the associated morbidity and mortality

Rheumatic carditis is characterized by pancarditis, with active inflammation of myocardium, pericardium, and endocardium

Endocarditis (valvulitis) is a universal finding in rheumatic carditis, whereas the presence of pericarditis or myocarditis is variable .Most rheumatic heart disease is isolated mitral valvular disease or combined aortic and mitral valvular disease. Isolated aortic or right-sided valvular involvement is quite uncommon.

Valvular insufficiency is characteristic of both acute and convalescent stages of acute rheumatic fever, whereas mitral and/or aortic valvular stenosis usually appears years or even decades after the acute illness.

Carditis usually presents as tachycardia and cardiac murmurs. Moderate to severe rheumatic carditis can result in cardiomegaly and heart failure with hepatomegaly and peripheral and pulmonary edema.

Mitral regurgitation is characterized typically by a high-pitched apical holosystolic murmur radiating to the axilla. In patients with significant mitral regurgitation, this may be associated with an apical mid-diastolic murmur of relative mitral stenosis. Aortic insufficiency is characterized by a high-pitched decrescendo diastolic murmur at the left sternal border.

A major change in the 2015 revision of the Jones Criteria is the acceptance of subclinical carditis (defined as without a murmur of valvulitis but with echocardiographic evidence of valvulitis) or clinical carditis (with a valvulitis murmur) as fulfilling the major criterion of carditis in all populations

3. Chorea Sydenham:

Occurs in approximately 10-15% of patients with acute rheumatic fever and usually presents as an isolated, subtle, movement disorder. Emotional liability, incoordination, poor school performance, uncontrollable movements, and facial grimacing, all exacerbated by stress and disappearing with sleep.

The latent period from acute GAS infection to chorea is usually substantially longer than for arthritis or carditis and can be months. Onset can be insidious, with symptoms being present for several months before recognition.

Clinical maneuvers to elicit features of chorea include:

(1) demonstration of milkmaid's grip (irregular contractions and relaxations of the muscles of the fingers while squeezing the examiner's fingers)
(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

chorea rarely, if ever, leads to permanent neurologic sequelae.

4.Erythema Marginatum :

Is a rare (approximately 1% of patients with acute rheumatic fever) but 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.

5.Subcutaneous Nodules :

Are a rare (≤1% of patients with acute rheumatic fever) 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:

- ✓ Arthralgia (only if arthritis is not used as a major criterion)
- ✓ Fever
- ✓ elevated acute phase reactants (ESR ,CRP)

- ✓ 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 rheumatic fever.

Evidence of a recent GAS infection. is supporting by:

- ✓ + ve throat culture or rapid streptococcal antigen test(Streptozyme test)
- ✓ elevated or rising serum antistreptococcal antibody titers. ASOT, anti–DNase B, antihyaluronidase.

Differential Diagnosis

ARITHRITIS

i. Juvenile idiopathic ARITHRITIS
ii. Systemic lupus erythematosus
iii. Pyogenic arthritis
iv. Malignancies
v. Reactive arthritis related to gastrointestinal infections (e.g., Shigella, Salmonella)

CARDITIS

i. Viral myocarditis
ii. Viral pericarditis
iii. Kawasaki disease

iv. Infective endocarditis

CHOREA

i. Huntington chorea
ii. Wilson disease

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 typical migratory polyarthritis and those with carditis without cardiomegaly or congestive heart failure 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 congestive heart failure should receive corticosteroids. The usual 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. When prednisone is being tapered, aspirin should be started at 50 mg/kg/day in 4 divided doses for 6 wk. to prevent rebound of inflammation.

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 a few week course of corticosteroids.

PREVENTION

Prevention of both initial and recurrent episodes of acute rheumatic fever depends on controlling GAS infections of the upper respiratory tract.

A. primary prevention

Prevention of initial attacks depends on identification and eradication of GAS causing acute pharyngitis. Appropriate antibiotic therapy instituted before the 9th day of symptoms of acute GAS pharyngitis is highly effective in preventing first attacks of acute rheumatic fever. However, approximately 30% of patients with acute rheumatic fever do not recall a preceding episode of pharyngitis and did not seek therapy. Oral penicillin or erythromycin 50 mg/kg/day or single IM benzathine pencillin 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 particularly susceptible to recurrences of RF with any subsequent GAS upper respiratory tract infection, whether or not they are symptomatic. 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. have elapsed 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 and 1.2 million IU for those weighing >27kg) every 4 wk. . In compliant patients, oral Penicillin V 250 mg twice daily and sulfadiazine or sulfasoxazole 500 - 1,000 mg given once daily are equally effective.

For patient who is allergic to both penicillin and sulfonamides, a macrolide (erythromycin or clarithromycin or azithromycin) may be used.