Acute rheumatic fever (ARF)

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Defintion and causes

Acute rheumatic fever (RF) occurs as a result of a complex interaction between group A streptococcus (GAS), a susceptible host, and the environment. An abnormal immune response to GAS infection leads to anacute inflammatory illness that most commonly affects the joints, brain, heart, or skin. Although the other manifestations are self-limiting and resolve without sequelae, carditis may result in significant morbidity and mortality

Causes

- Acute rheumatic fever is believed to be an immunologic response that occurs as a delayed sequelae of group A streptococcal infection of the pharynx but not of the skin. The attack rate of acute rheumatic fever after streptococcal infection varies with the severity of the infection, ranging from 0.3% to 3%
- Important predisposing factors include family history of rheumatic fever, low socioeconomic status (poverty, poor hygiene,medical deprivation), and age between 6 and 15 years (with a peak incidence at 8 years of age)

Pathology

The inflammatory lesion is found in many parts of the body, most notably in the heart, brain, joints, and skin. Rheumatic carditis was considered to be pancarditis, with myocarditis being the most important element. It is now recognized that the valvular component may be as important as or much more important than myocardial and pericardial involvement .

- Valvular damage most frequently and most severely involves the mitral, less commonly the aortic, and rarely the tricuspid and pulmonary valves.
- Aschoff bodies in the atrial myocardium are believed to be characteristic of rheumatic fever
- These consist of inflammatory lesions associated with swelling, fragmentation of collagen fibers, and surrounded by or infiltrated by large multinucleated cells.

Clinical Manifestations

- Acute rheumatic fever is diagnosed by the use of revised Jonescriteria. The criteria are three groups of important clinical and laboratory findings: (1) five major manifestations, (2)four minor manifestations, and (3) supporting evidence of an antecedent group A streptococcal infection

History

- History of streptococcal pharyngitis, 1 to 5 weeks (average, 3weeks) before the onset of symptoms, is common.
- The latent period may be as long as 2 to 6 months (average, 4 months) in cases of isolated chorea

Major criteria	Minor criteria
Carditis	Fever
Chorea	Arthralgia
Polyarthritis	Elevated Acute-Phase Reactants
Erythema marginatum	Erythrocyte sedimentation rate
Subcutaneous nodules	C-reactive protein
	Prolonged PR interval (ECG)

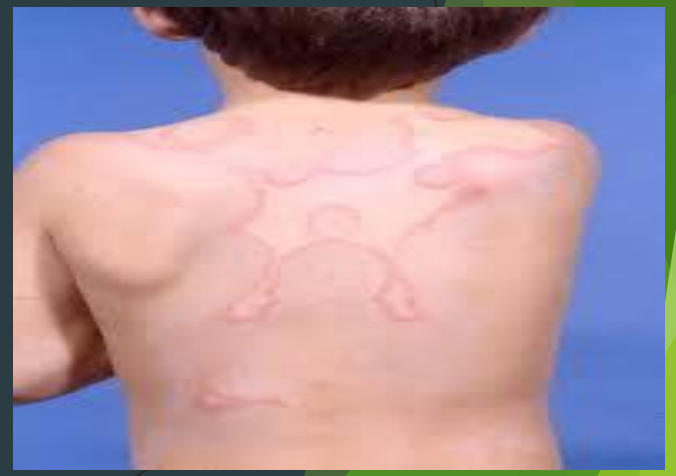
- **Supporting evidence of antecedent group A streptococcal pharyngeal infection**
- Positive throat culture or rapid streptococcal test
- Elevated or rising streptococcal antibody titer(Antistreptolysin O(ASO) titer)

- **Criteria for diagnosis of Primary episode of ARF**
- 2 major *or* 1 major plus 2 minor *plus* 2 evidence of preceding strep infection

- **Arthritis**, the most common manifestation of acute rheumatic fever (70% of cases), usually involves large joints (e.g., knees, ankles, elbows, wrists.
- **Carditis :** occurs in 50% of patients. Signs of carditis include some or all of the following
- Tachycardia (out of proportion to the degree of fever) is common
- A heart murmur of mitral regurgitation (MR) or aortic regurgitation. (AR) (or both) is almost always present.
- pericardial effusion, increased left ventricular (LV) dimension, or impaired LV function
- Pericarditis
- congestive heart failure (CHF)

- **Sydenham's Chorea** is found in 15% of patients with acute rheumatic fever. It is now known that the clinical manifestations of Sydenham chorea occur because of neuropathologic changes and inflammation in the basal ganglia, cerebral cortex, and the cerebellum It occurs more often in prepubertal girls (8–12 years) than in boys. It is a neuropsychiatric disorder consisting of both neurologic signs (choretic movement involuntary, purposeless movements and hypotonia) and psychiatric signs (e.g., emotional lability, hyperactivity, separation anxiety, obsessions, and compulsions)

- **Erythema Marginatum** occurs in fewer than 10% of patients with acute rheumatic fever. The characteristic nonpruritic serpiginous or annular erythematous rashes are most prominent on the trunk and the inner proximal portions of the extremities; they are never seen on the face.



Management

- When acute rheumatic fever is suggested by history and physicalexamination, one should obtain the following laboratory studies:
- complete blood count, acute-phase reactants (ESR and CRP), throat culture, ASO titer , chest radiographs, and ECG
- Cardiology consultation is indicated to clarify whether there is cardiac involvement; two-dimensional echocardiographic and Doppler studies are usually performed at that time.

- Benzathine penicillin G, 0.6 to 1.2 million units intramuscularly, is given to eradicate streptococci
- In patients who are allergic to penicillin,erythromycin, 40 mg/kg per day in two to four doses for 10 days, may be substituted for penicillin
- Bed rest of varying duration is recommended. The duration depends on the type and severity of the manifestations and may range from 1 week (for isolated arthritis) to several weeks for severe carditis.
- Therapy with antiinflammatory agents should be started as soon as acute rheumatic fever has been diagnosed

a. For mild to moderate carditis, aspirin alone is recommended in adose of 90 to 100 mg/kg per day in four to six divided doses. This dose is continued for 4 to 8 weeks, depending on the clinical response. After improvement, the therapy is withdrawn gradually over 4 to 6 weeks

b.For arthritis, aspirin therapy is continued for 2 weeks and gradually withdrawn over the following 2 to 3 weeks. Rapid resolution of joint symptoms with aspirin within 24 to 36 hours is supportive evidence of the arthritis of acute rheumatic fever

c.Prednisone (2 mg/kg per day in four divided doses for 2 to 6 weeks) is indicated only in cases of severe carditis

Treatment of CHF includes the following:

- Complete bed rest with orthopneic position and moist, cool oxygen
- Prednisone for severe carditis of recent onset
- Digoxin is used with caution, beginning with half the usual recommended dose
- Furosemide, 1 mg/kg every 6 to 12 hours

PREVENTION

Primary Prevention

- Primary prevention of rheumatic fever is possible with a 10-day course of penicillin therapy for streptococcal pharyngitis.
- However, primary prevention is not possible in all patients because about 30% of the patients develop subclinical pharyngitis and therefore do not seek medical treatment

Secondary Prevention

- Patients with documented histories of rheumatic fever, including those with isolated chorea and those without evidence of rheumatic heart disease, must receive prophylaxis
- Ideally, patients should receive prophylaxis indefinitely. For patients who had acute rheumatic fever without carditis, the prophylaxis should continue for at least 5 years or until the person is 21 years Of age
- If the patient had rheumatic carditis or residual valvular disease as a result of rheumatic fever, the duration of prophylaxis should be longer

Method of prophylaxis:

- The method of choice for secondary prevention is benzathine penicillin G, 1.2million units given intramuscularly every 28 days
- Alternative methods, although not as effective, are the following
    a. Oral penicillin V, 250 mg, twice daily
    b. Oral sulfadiazine 1 g or sulfisoxazole 0.5 g once daily
    c. Oral erythromycin ethyl succinate, 250 mg, twice daily

Complications

- Valve stenosis. This narrowing of the valve decrease blood flow
- Valve regurgitation
- Damage to heart muscle
- atrial fibrillation
- heart failure