DEFINITION

1. **Rheumatic fever** is a multisystem inflammatory disease that occurs as a sequel to pharyngeal infection with the bacterium, Group A Streptococcus pyogenes. Major features are arthritis and carditis.

CLINICAL MANIFESTATIONS

2. These are varied. They include fever, pain (with or without swelling) in one or more joints, endocarditis, pericarditis, myocarditis, pleurisy, subcutaneous nodules, a characteristic skin lesion (erythema marginatum) and an even more characteristic disturbance of central nervous system function, Sydenham's chorea. Carditis is more frequent in the youngest age groups, and the majority of rheumatic fever attacks occurring in adults are manifested primarily by arthritis.

3. Rheumatic fever is rare in children under 4 years of age, most cases occurring in the 6-15 year age group. Initial attacks affecting adults are rare. About two thirds of patients give a history of sore throat one to three weeks before the onset of rheumatic symptoms. Presentation may be abrupt or gradual, and in some cases a specific bout of rheumatic fever is not recognised at all, the patient presenting with established rheumatic heart disease.

4. **Arthritis** usually affects larger joints, particularly wrists, elbows, knees and ankles. Hips are less often affected, small joints of hands and feet rarely, and spine almost never. Characteristically, one or two joints are affected for 2 to 3 days, and the process then affects another joint. Objective signs are usually mild and often arthralgia alone occurs. Untreated, the joint pains settle within 1-4 weeks. The arthritis of rheumatic fever does not lead to permanent damage to the joints affected.

5. **Carditis** is the most important clinical manifestation of rheumatic fever, being the one with permanent effects. It occurs in about 55% of primary attacks and is commoner in young patients. Any heart structure may be affected by the rheumatic process; endocarditis is detected clinically by new or changing murmurs, and pericarditis by a friction rub. Heart failure is the most serious presentation, occurring in younger patients.

6. **Chorea,** once occurring in 50% of patients, is now rare. The latent period is longer than arthritis or carditis, being 1 to 6 months. It features jerky, purposeless movements, exaggerated by tension but disappearing in sleep. It occurs predominantly in females, unlike the other manifestations of rheumatic fever.

7. Other less common manifestations include erythema marginatum, an evanescent macular eruption (which is not pathognomonic, being seen in acute glomerulonephritis and drug reactions) and subcutaneous nodules, which may appear as painless lumps in people with long-standing carditis; these are seen less frequently in recent times.

8. The major **complication** of rheumatic fever is **valvular disease of the heart.** In the USA and Western Europe, rheumatic valvular heart disease has become mainly a disorder of people aged over 50, whereas it remains an important disability and cause of death in children and young adults in Eastern Europe, Asia, Africa, and Central and South America.

AETIOLOGY

9. The **incidence of rheumatic fever** has fallen dramatically in Western Europe, the United States and Japan over the past century. The disease continues to be a major health problem in the developing countries of Asia, Africa, the Middle East and South America.

10. Rheumatic fever is **a sequel to infection of the throat with Streptococcus pyogenes.** The particular type is Lancefield group A beta-haemolytic streptococcus. Human mucous membrane and skin serve as natural reservoirs of the organism, and spread of throat infection from person to person is usually by droplet inhalation. Epidemics of sore throat occasionally result from contaminated milk or food.

11. Streptococcal throat infection has its highest incidence in children aged from 5 to 15 years. It is estimated that 15-20% of infections are asymptomatic.

12. The latent period from an attack of streptococcal throat infection to the onset of rheumatic fever is one to five weeks, with an average of 19 days. Rheumatic fever is most likely to follow clinically severe pharyngitis, but one third or more of cases occur after asymptomatic or very mild streptococcal infection.

13. The mechanism by which streptococcal infection leads to rheumatic fever remains uncertain. There is no evidence of a direct toxic effect of streptococci or cell products. Most authorities currently favour the theory that it is **a form of autoimmune disorder** in which tissue damage is mediated by the host's own immunological response to the antecedent streptococcal infection. Only certain serotypes of Group A streptococci have the potential to cause rheumatic fever. The organism must be able to produce a significant antibody response, and the stronger the antibody response, the greater is the attack rate of rheumatic fever.

14. Frequently, susceptibility to rheumatic fever is familial, although there is no good evidence of a genetic basis. The search for genetic factors has not been conclusive. There is a tendency for rheumatic fever to affect more than one member of a given family.

15. Certain races appear susceptible, including Asians in Britain and Polynesians in New Zealand. This may be due to socio-economic factors rather than genetic ones, because specific racial predilection has never substantiated.

16. **Social factors**, such as crowding, poor housing, poor hygiene and inadequate medical care all contribute to raising the incidence of rheumatic fever in a community. Of these, **overcrowding** is the most significant as regards the spread of streptococcal infection.

Course and prognosis

17. There has been a significant decrease in the incidence and mortality of acute rheumatic fever in the Western world; the disease itself has become milder, and recurrences have been reduced by antibiotics. Improved living standards, better housing and less overcrowding have limited person to person contact, thereby minimising the spread of streptococcal infection. More than 90% of attacks of rheumatic fever abate in 12 weeks, and the prognosis for patients without carditis during the acute attack is excellent. The severity of residual heart disease is related to that of carditis during the acute attack and to the severity and number of recurrences.

Recurrence

18. An attack of rheumatic fever greatly increases the chances that a **subsequent streptococcal throat infection** will be followed by another attack of rheumatic fever. The risk of recurrence is greater in children, in patients with pre-existing rheumatic heart disease, and in those experiencing symptomatic throat infection. The risk declines with advancing age, but nevertheless rheumatic patients remain at increased risk well into adult life.

19. The latent period is the same in recurrences as in initial attacks. The streptococcal throat infection that triggers a recurrence of rheumatic fever need not be symptomatic. In outbreaks of sore throat caused by serotypes of streptococcus associated with rheumatic fever, up to 50% of previous acute rheumatic fever sufferers will have a recurrence. Carditis with a recurrence is more common in those patients in whom it was present in the first attack, but it may occur in any patient.

20. The rate of recurrence may be reduced by long term administration of penicillin by mouth or by injection, and this treatment should be continued for at least 5 years. It is particularly advisable in patients who have had cardiac involvement in the initial attack.

21. **Rheumatic fever** is an inflammatory disease comprising a loosely associated group of clinical manifestations which may occur in almost any combination. While the exact aetiology is unclear, the condition is generally held to result from an **immune reaction to a streptococcal throat infection.** The causative organism infects the throat by **droplet spread** from another person. The disease has a strong tendency to recur following a further streptococcal throat infection.

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