

Rheumatic heart disease

4th year lecture

Acute rheumatic fever

- Acute rheumatic fever usually affects children and young adults between the ages of 5 and 15 years.

Pathogenesis

- The condition is triggered by an **immune-mediated delayed response** to infection with specific strains of **group A streptococci**, which have antigens that cross-react with cardiac myosin and sarcolemmal membrane proteins.
- Antibodies produced against the streptococcal antigens cause inflammation in the endocardium, myocardium and pericardium, as well as the joints and skin.
- Histologically, fibrinoid degeneration is seen in the collagen of connective tissues.
- **Aschoff nodules** are pathognomonic and occur only in the heart. They are composed of multinucleated giant cells surrounded by macrophages and T lymphocytes, and are not seen until the subacute or chronic phases of rheumatic carditis.

Clinical features

- multisystem disorder that usually presents with fever, anorexia, lethargy and joint pain, 2–3 weeks after an episode of streptococcal pharyngitis.
- There may be no history of sore throat, however. Arthritis occurs in approximately 75% of patients.
- Other features include rashes, subcutaneous nodules, carditis and neurological changes.

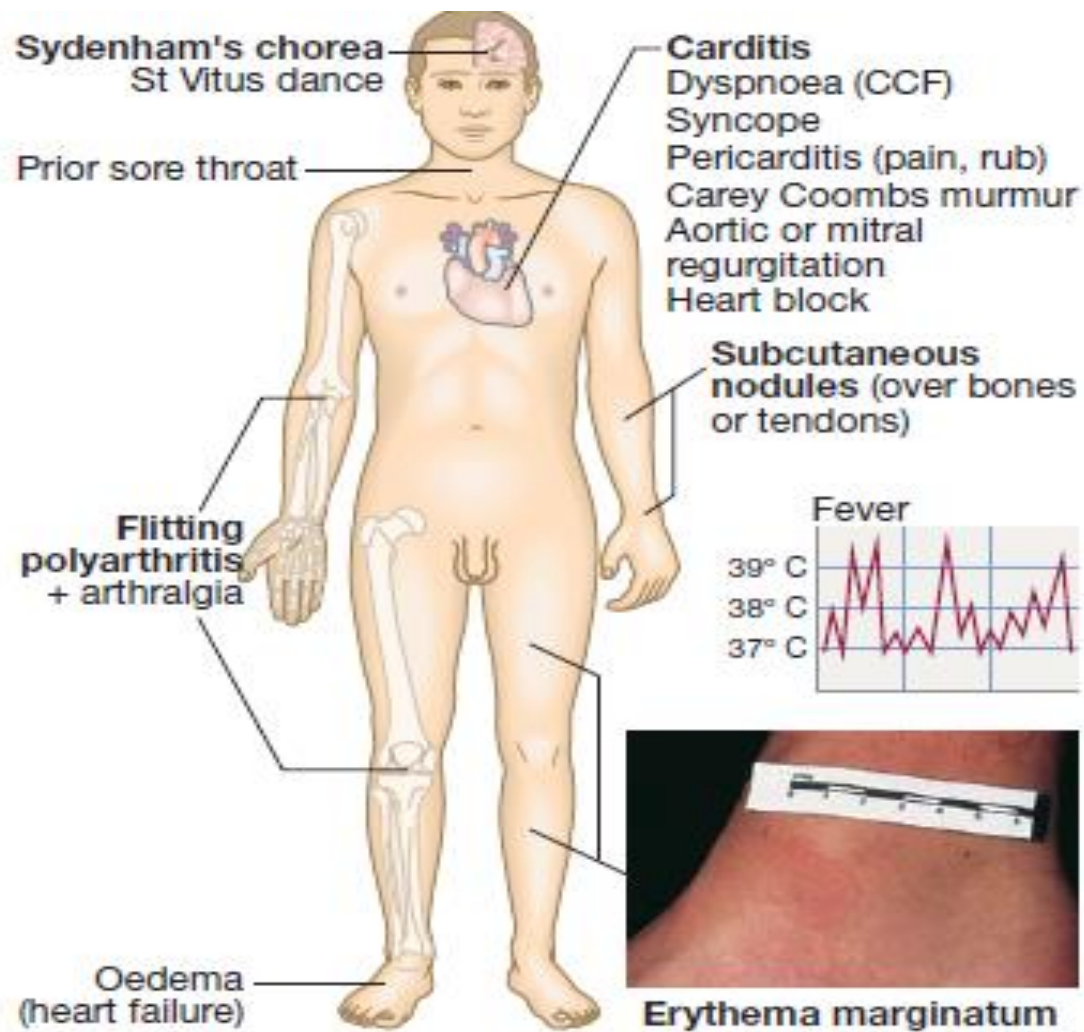


Fig. 16.80 Clinical features of rheumatic fever. Bold labels indicate Jones major criteria. (CCF = congestive cardiac failure) *Inset (Erythema marginatum)* From Savin JA, Hunter JAA, Hepburn NC. *Skin signs in clinical medicine*. London: Mosby-Wolfe, Elsevier; 1997.

The diagnosis, made using the revised Jones criteria:

- is based on two or more major manifestations, or one major and two or more minor manifestations, along with evidence of preceding streptococcal infection.
- A presumptive diagnosis of acute rheumatic fever can be made without evidence of preceding streptococcal infection in cases of isolated chorea or pancarditis, if other causes of these have been excluded.
- In cases of established rheumatic heart disease or prior rheumatic fever, a diagnosis of acute rheumatic fever can be made based only on the presence of multiple minor criteria and evidence of preceding group A streptococcal pharyngitis.



16.75 Jones criteria for the diagnosis of rheumatic fever

Major manifestations

- Carditis
- Polyarthritits
- Chorea
- Erythema marginatum
- Subcutaneous nodules

Minor manifestations

- Fever
- Arthralgia
- Raised erythrocyte sedimentation rate or C-reactive protein
- Previous rheumatic fever
- Leucocytosis
- First-degree atrioventricular block

Plus

- Supporting evidence of preceding streptococcal infection: recent scarlet fever, raised antistreptolysin O or other streptococcal antibody titre, positive throat culture*

*Evidence of recent streptococcal infection is particularly important if there is only one major manifestation.

Clinical features

- Carditis
- Arthritis
- Skin lesions
- Sydenham's chorea
- Other systemic manifestations, such as pleurisy, pleural effusion
- and pneumonia, may occur but are rare.

Carditis

- pancarditis involving the endocardium, myocardium and pericardium to varying degrees.
- Its incidence declines with increasing age, ranging from 90% at 3 years to around 30% in adolescence.
- It may manifest as breathlessness (due to heart failure or pericardial effusion), palpitations or chest pain (usually due to pericarditis or pancarditis).
- Other features include tachycardia, cardiac enlargement and new or changed murmurs.

- A soft systolic murmur due to mitral regurgitation is very common.
- A soft mid-diastolic murmur (the Carey Coombs murmur) is typically due to valvulitis, with nodules forming on the mitral valve leaflets.
- Aortic regurgitation occurs in 50% of cases.
- but the tricuspid and pulmonary valves are rarely involved.

- Pericarditis may cause chest pain, a pericardial friction rub and precordial tenderness.
- Cardiac failure may be due to myocardial dysfunction or valvular regurgitation.
- ECG evidence commonly includes ST and T wave changes.
- Conduction defects, including AV block, sometimes occur and may cause syncope.

Arthritis

- This is the most common major manifestation and occurs early when streptococcal antibody titres are high.
- asymmetric and migratory inflammation of the large joints typically affects the knees, ankles, elbows and wrists.
- The joints are involved in quick succession and are usually red, swollen and tender for between a day and 4 weeks.

Skin lesion

- **Erythema marginatum** occurs in less than 5% of patients.
 - The lesions start as red macules that fade in the center but remain red at the edges, and occur mainly on the trunk and proximal extremities but not the face.
 - The resulting red rings or 'margins' may coalesce or overlap
- **Subcutaneous nodules** occur in 5–7% of patients.
 - They are small (0.5–2.0 cm), firm and painless, and are best felt over extensor surfaces of bone or tendons.
 - They typically appear more than 3 weeks after the onset of other manifestations and therefore help to confirm rather than make the diagnosis.

Sydenham chorea

- Late neurological manifestation that appears at least 3 months after the episode of acute rheumatic fever, when all the other signs may have disappeared.
 - Emotional lability may be the first feature and is typically followed by purposeless, involuntary, choreiform movements of the hands, feet or face.
 - Speech may be explosive and halting.
- It occurs in up to one-third of cases and is more common in females.
- Spontaneous recovery usually occurs within a few months.
 - Approximately one-quarter of affected patients will go on to develop chronic rheumatic valve disease.

Investigations

- ESR and CRP.... useful for monitoring progress of the disease.
- Throat cultures should be taken but positive results are obtained in only 10–25% of cases since the infection has often resolved by the time of presentation.
- Serology for antistreptolysin O antibodies (ASO) should be performed.

(Raised levels provide supportive evidence for the diagnosis but are normal in one-fifth of adult cases of rheumatic fever and most cases of chorea.)

- Echocardiography... shows mitral regurgitation with dilatation of the mitral annulus and prolapse of the anterior mitral leaflet; it may also demonstrate aortic regurgitation and pericardial effusion.



16.76 Investigations in acute rheumatic fever

Evidence of a systemic illness

- Leucocytosis, raised erythrocyte sedimentation rate and C-reactive protein

Evidence of preceding streptococcal infection

- Throat swab culture: group A β -haemolytic streptococci (also from family members and contacts)
- Antistreptolysin O antibodies (ASO titres): rising titres, or levels of > 200 U (adults) or > 300 U (children)

Evidence of carditis

- Chest X-ray: cardiomegaly; pulmonary congestion
- ECG: first- and, rarely, second-degree atrioventricular block; features of pericarditis; T-wave inversion; reduction in QRS voltages
- Echocardiography: cardiac dilatation and valve abnormalities

Management

- The aims of management are to limit cardiac damage and relieve symptoms.
- Bed rest
- Treatment of cardiac failure
- Antibiotics
- Aspirin
- Glucocorticoids

- A single dose of benzathine benzylpenicillin (1.2 million U IM) or oral phenoxymethylpenicillin (250 mg 4 times daily for 10 days) should be given on diagnosis to eliminate any residual streptococcal infection.
- If the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.

Penicillin also used for prophylaxis

- Further attacks of rheumatic fever are unusual after the age of 21, when antibiotic treatment can usually be stopped.
- The duration of prophylaxis should be extended if an attack has occurred in the last 5 years, or if the patient lives in an area of high prevalence and has an occupation (such as teaching) with a high risk of exposure to streptococcal infection.
- In those with residual heart disease, prophylaxis should continue until 10 years after the last episode or 40 years of age, whichever is later.
- While long-term antibiotic prophylaxis prevents further attacks of acute rheumatic fever, it does not protect against infective endocarditis.

glucocorticoid

- These produce more rapid symptomatic relief than aspirin and are indicated in cases with carditis or severe arthritis.
- There is no evidence that long-term steroids are beneficial.\
- Prednisolone 1.0–2.0 mg/kg per day in divided doses) should be continued until the ESR is normal and then tailed off.

Chronic rheumatic heart disease

- Chronic valvular heart disease develops in at least half of those affected by rheumatic fever with carditis.
- Two-thirds of cases occur in women.
- The main pathological process in chronic rheumatic heart disease is progressive fibrosis.
- The mitral valve is affected in more than 90% of cases; the aortic valve is the next most frequently involved, followed by the tricuspid and then the pulmonary valve.
- Isolated mitral stenosis accounts for about 25% of all cases, and an additional 40% have mixed mitral stenosis and regurgitation.

The End