# Disease of the heart valves

## Rheumatic Fever

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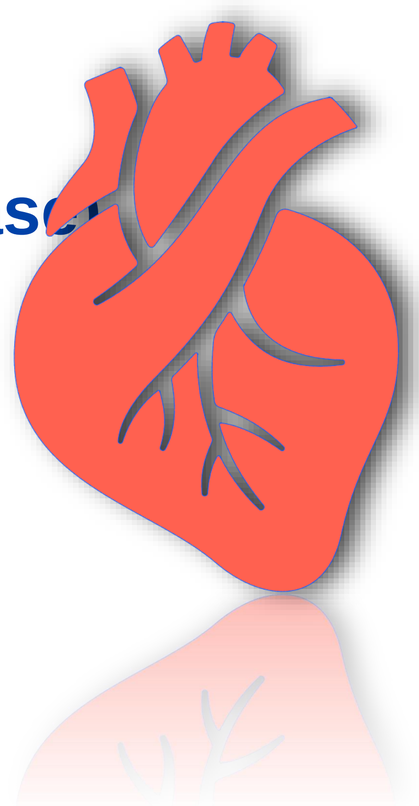

- The heart valves allow forward movement of blood through the cardiac chambers when they are open and prevent backward flow when they are closed. A diseased valve may become narrowed, obstructing forward flow, or become leaky, causing backward flow or regurgitation. Breathlessness is a common symptom of valve disease, and acute severe breathlessness may be a presenting symptom of valve failure.

- Predisposition to valvular disease may be genetically determined, can arise as the result of rheumatic fever or infections, or can occur in association with dilatation of the cardiac chambers in heart failure.

## 16.73 Causes of acute valve failure

### Aortic regurgitation

- Aortic dissection
- Infective endocarditis

### Mitral regurgitation

- Papillary muscle rupture due to acute myocardial infarction
- Infective endocarditis
- Rupture of chordae due to myxomatous degeneration

### Prosthetic valve failure

- Mechanical valves: fracture, jamming, thrombosis, dehiscence
- Biological valves: degeneration with cusp tear

## 16.74 Principal causes of valve disease

### Valve regurgitation

- Congenital
- Acute rheumatic carditis
- Chronic rheumatic carditis
- Infective endocarditis
- Cardiac failure*
- Syphilitic aortitis
- Traumatic valve rupture
- Senile degeneration
- Damage to chordae and papillary muscles

### Valve stenosis

- Congenital
- Rheumatic carditis
- Senile degeneration

*Causes dilatation of the valve ring.

# Rheumatic heart disease

# Acute Rheumatic Fever

- Acute rheumatic fever usually affects children and young adults between the ages of 5 and 15 years. It is now rare in high-income countries in Western Europe and North America, where the incidence is about 0.5 cases per 100 000, but remains endemic in South Asia, Africa and South America. Recent studies indicate that the current incidence of rheumatic heart disease in India ranges between 13 and 150 cases per 100 000 population per year, where it is the commonest cause of acquired heart disease in childhood and adolescence.

- **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**

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

- Using the revised Jones criteria , **the diagnosis 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  of acute rheumatic fever can be made without evidence of preceding streptococcal infection in cases of isolated chorea or pancarditis, if other causes have been excluded. In cases of established rheumatic heart disease or prior rheumatic fever acute rheumatic fever can be diagnosed based only on the presence of multiple minor criteria and evidence of preceding group A streptococcal pharyngitis.

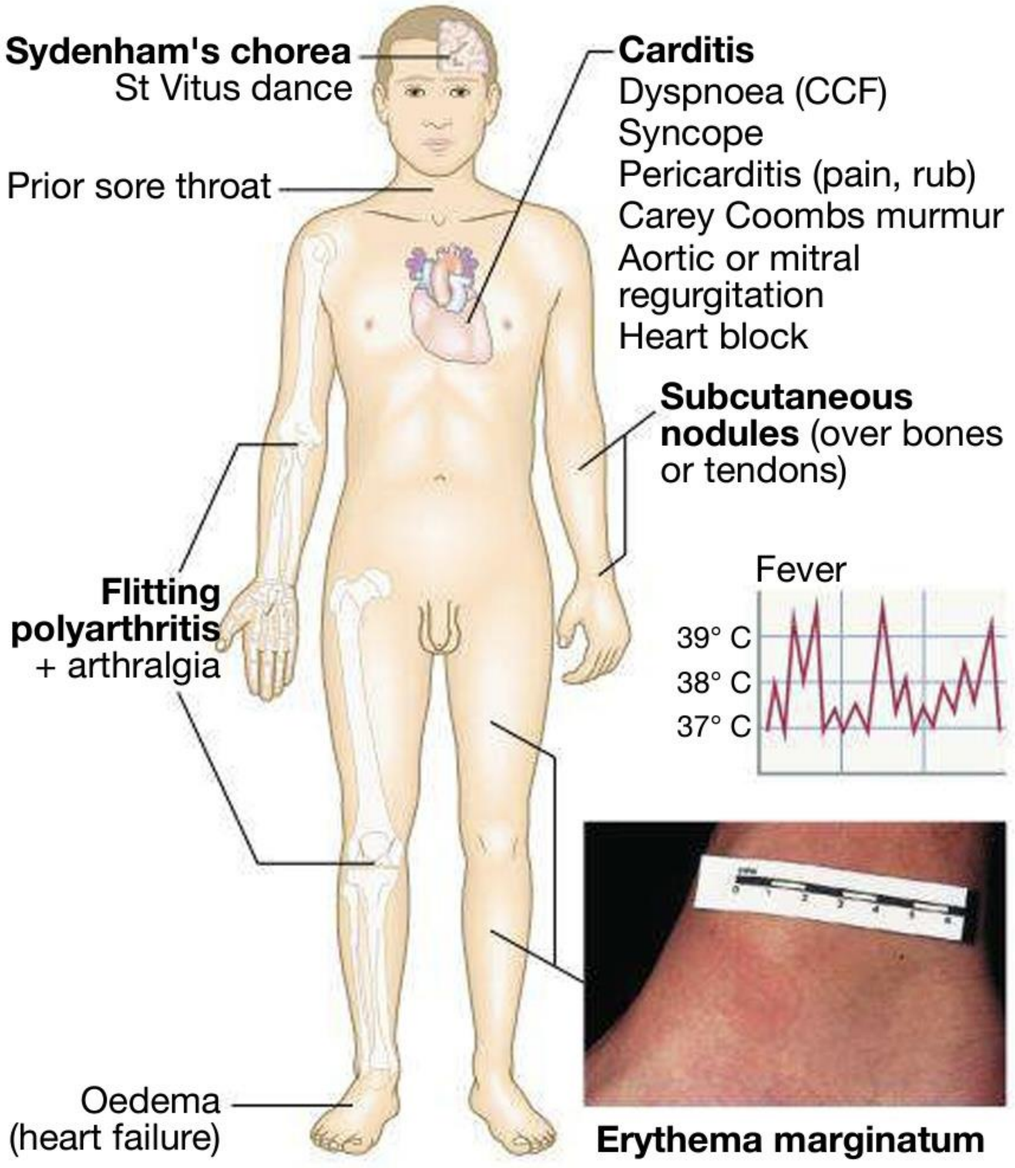


**Fig. 16.78 Clinical features of rheumatic fever.** Bold labels indicate Jones major criteria. (CCF = congestive cardiac failure) *Inset (Erythema marginatum) From Savin JA, Hunter JAA, Hepburn NC. Skin signs in clinical medicine. London: Mosby–Wolfe, Elsevier; 1997.*

## 16.75 Jones criteria for the diagnosis of rheumatic fever

### Major manifestations

- Carditis
- Polyarthritis
- 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.

- **Carditis**

- Rheumatic fever causes a 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 commonest major manifestation and occurs early when streptococcal antibody titres are high. An acute painful, 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 lesions :** Erythema marginatum occurs in less than 5% of patients. The lesions start as red macules that fade in the centre 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’s chorea**, also known as St Vitus dance, is a late neurological manifestation that appears at least 3 months after the episode of acute rheumatic fever, when all the other signs may have disappeared. It occurs in up to one-third of cases and is more common in females. 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. Spontaneous recovery usually occurs within a few months. Approximately one-quarter of affected patients will go on to develop chronic rheumatic valve disease.

- Other features Other systemic manifestations, such as pleurisy, pleural effusion and pneumonia, may occur but are rare.

- **Investigations**

- Blood should be taken for measurement of ESR and CRP since these are 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 and provide supportive evidence for the diagnosis but are normal in one-fifth of adult cases of rheumatic fever and most cases of chorea. Echocardiography should be carried out and typically 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** is important, as it lessens joint pain and reduces cardiac workload. The duration should be guided by symptoms, along with temperature, leucocyte count and ESR, and should be continued until these have settled. Patients can then return to normal physical activity but strenuous exercise should be avoided in those who have had carditis.

- **Treatment of cardiac failure** Some patients, particularly those in early adolescence, can develop a fulminant form of the disease with severe mitral regurgitation and, sometimes, concomitant aortic regurgitation. If heart failure does not respond to medical treatment, valve replacement may be necessary and is often associated with a dramatic decline in rheumatic activity. Occasionally, AV block may occur but is seldom progressive and usually resolves spontaneously. Rarely, pacemaker insertion may be required.

- **Antibiotic therapy**

- A single dose of benzathine benzylpenicillin (1.2millionU 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. Patients are susceptible to further attacks of rheumatic fever if another streptococcal infection occurs, and long-term prophylaxis with penicillin should be given with oral phenoxymethylpenicillin (250 mg twice daily) or as benzathine benzylpenicillin (1.2millionU IM monthly), if adherence is in doubt. Sulfadiazine or erythromycin may be used if the patient is allergic to penicillin; sulphonamides prevent infection but are not effective in the eradication of group A streptococci. 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.

- **Aspirin** This usually relieves the symptoms of arthritis rapidly and a response within 24 hours helps confirm the diagnosis. A reasonable starting dose is 60 mg/kg body weight/day, divided into six doses. In adults, 100 mg/kg per day may be needed up to the limits of tolerance or a maximum of 8 g per day. Mild toxicity includes nausea, tinnitus and deafness; vomiting, tachypnoea and acidosis are more serious. Aspirin should be continued until the ESR has fallen and then gradually tailed off.

- **Glucocorticoid steroids** 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 gradually reduced.

# 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. Some episodes of rheumatic fever pass unrecognised and it is possible to elicit a history of rheumatic fever or chorea in only about half of all patients with chronic rheumatic heart disease. 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 main pathological process in chronic rheumatic heart disease is progressive fibrosis. The heart valves are predominantly affected but involvement of the pericardium and myocardium also occurs and may contribute to heart failure and conduction disorders. Fusion of the mitral valve commissures and shortening of the chordae tendineae may lead to mitral stenosis with or without regurgitation. Similar changes in the aortic and tricuspid valves produce distortion and rigidity of the cusps, leading to stenosis and regurgitation. Once a valve has been damaged, the altered haemodynamic stresses perpetuate and extend the damage, even in the absence of a continuing rheumatic process.