

Acute Rheumatic Fever

By:

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Epidemiology :

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, but remains endemic in the Indian subcontinent, Africa and South America

Pathogenesis

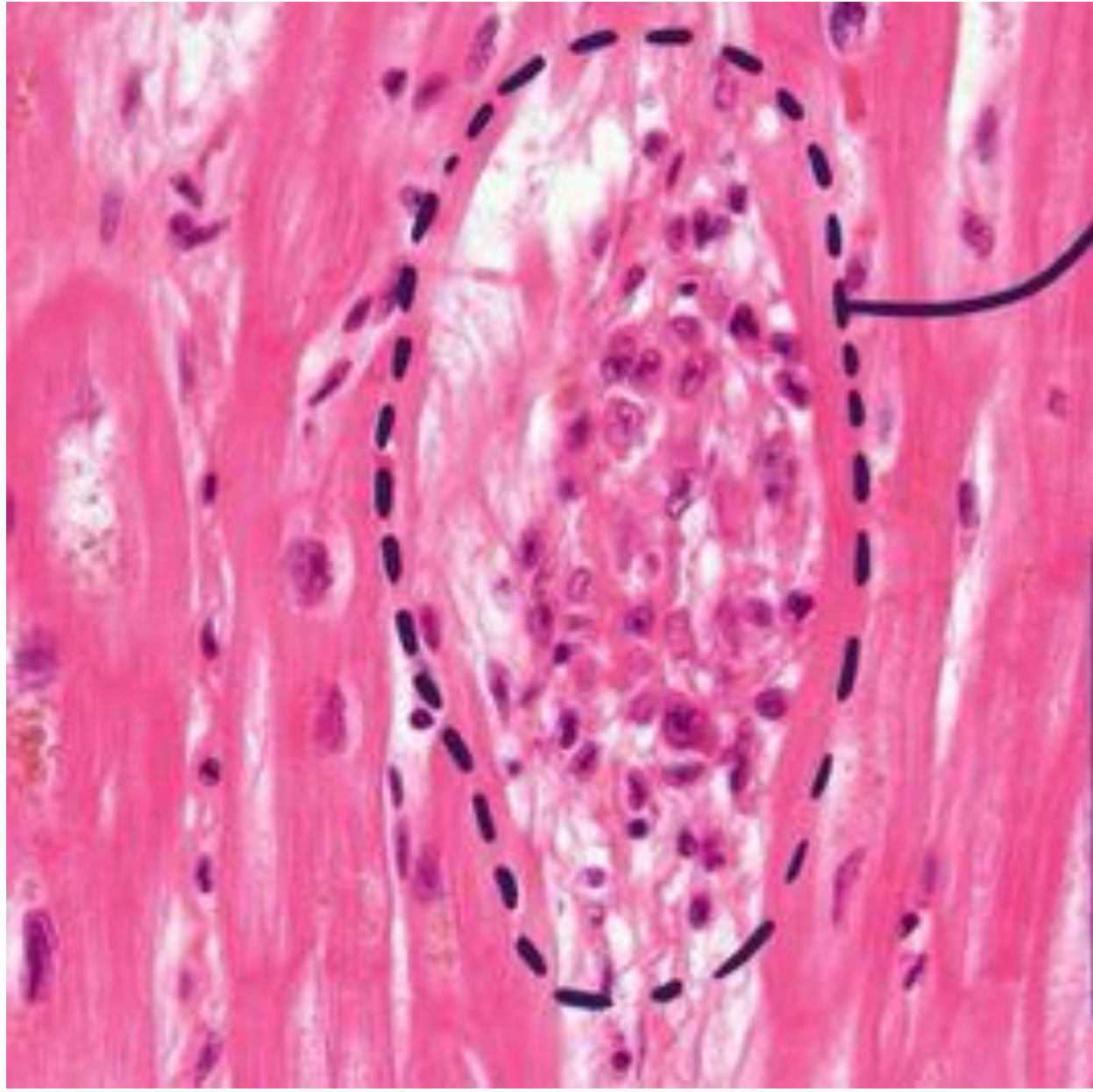
The condition is triggered by an **immune-mediated (type II HSR) delayed** response to infection with specific strains of **group A streptococci**, which have antigens that **cross-react** with cardiac myosin and sarcolemmal membrane proteins.

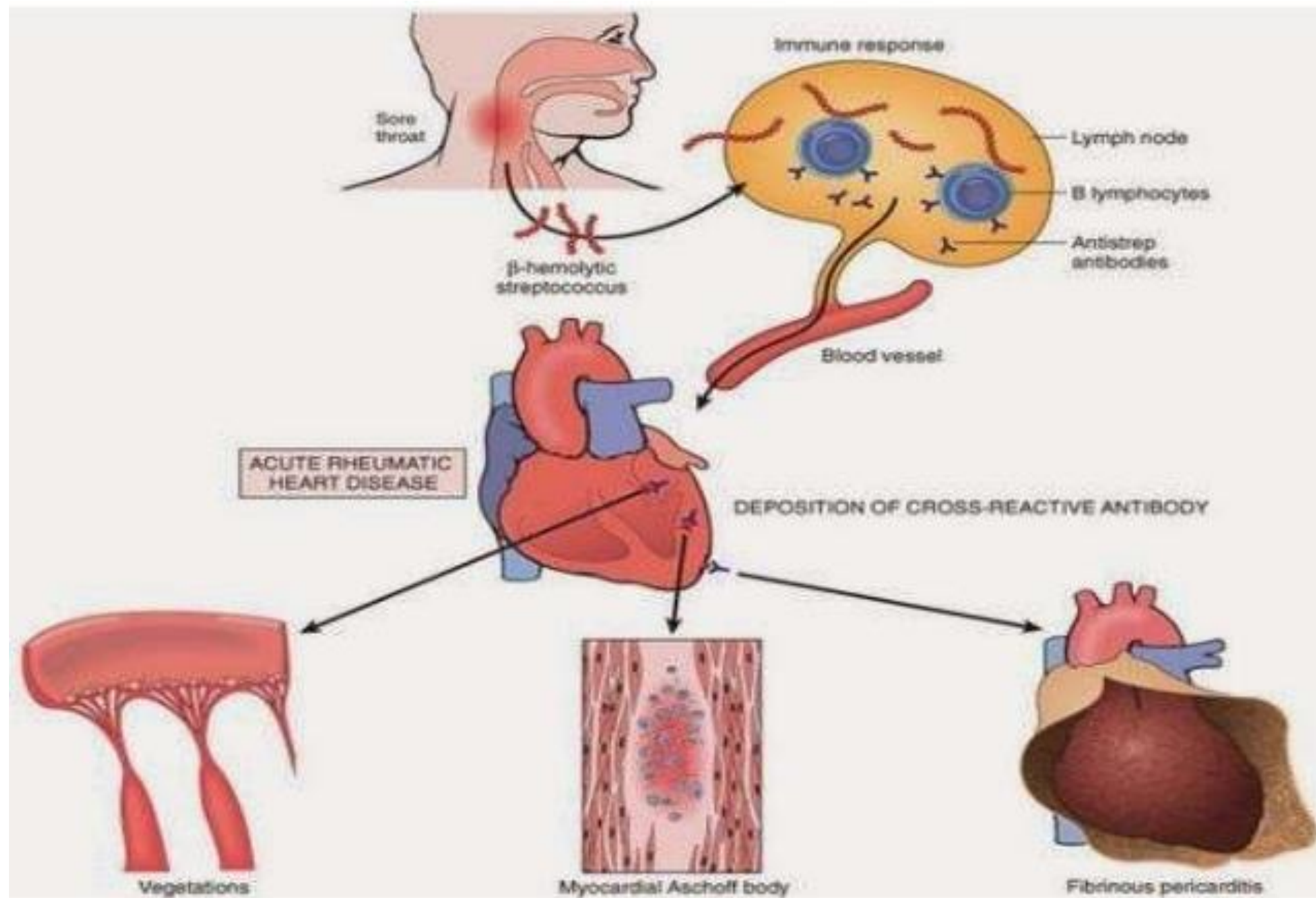
Antibodies produced against the streptococcal antigens cause inflammation in the endocardium, myocardium and pericardium, as well as the joints, skin and nervous system.

Histopathology

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

It is a multisystem disorder

2–3 weeks after an episode of streptococcal pharyngitis

There may be no history of sore throat

It usually presents with (fever, anorexia, lethargy and joint pain)

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**.

Symptoms of carditis :

Breathlessness (due to heart failure or pericardial effusion)

Palpitations or chest pain (usually due to pericarditis or pancarditis).

Syncope (AV block)

Signs of carditis :

Tachycardia

Cardiac enlargement (mild)

Pericarditis may cause chest pain, a pericardial friction rub and precordial tenderness.

Cardiac failure may be due to myocardial dysfunction or valvular regurgitation

New or changed murmurs:

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

ECG changes in carditis :

It commonly includes ST and T wave changes (myocarditis and pericarditis)

Conduction defects : first degree AV block , and rarely 2nd or 3rd degree AV block

ARTHRITIS

This is the most common and earliest major manifestation (in 75%)

- Acute (1 day and 4 weeks.)
- Inflammation (red , swelling , tender)
- Asymmetric
- migratory (in succession)
- Large joints typically affects the knees, ankles, elbows and wrists.
- Dramatic response to NSAID

Skin lesions

They occurs in about **5%** of patients

They appear more **than 3 weeks after** the onset of other manifestations and therefore help to confirm rather than make the diagnosis.

Erythema marginatum.

- start as red macules that fade in the centre but remained at the edges
- occur mainly on the trunk and proximal extremities but **not the face**.
- The resulting red rings or 'margins' may coalesce or overlap

Subcutaneous nodules

- small (0.5–2.0 cm), firm and painless
- best **felt** over extensor surfaces of bone or tendons

Rheumatic fever-diagnosis



Subcutaneous nodules
(nodules of rheumatoid arthritis are larger)

Erythema marginatum / Subcutaneous nodules



<http://www.hxbenefit.com/erythema-marginatum.htm>

<http://www.doctortipster.com/1789-rheumatic-fever.html>

Sydenham's chorea (St Vitus dance)

- It 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 . It is more common in females.
- Upto **70% of cases have carditis**
- It is a purposeless, involuntary, choreiform movements of the hands, feet or face.
- Emotional lability may be the first feature
- **Prognosis** : Spontaneous recovery usually occurs within a few months.



00:05

-00:30

Other rare features

- Pleurisy
- pleural effusion
- pneumonia

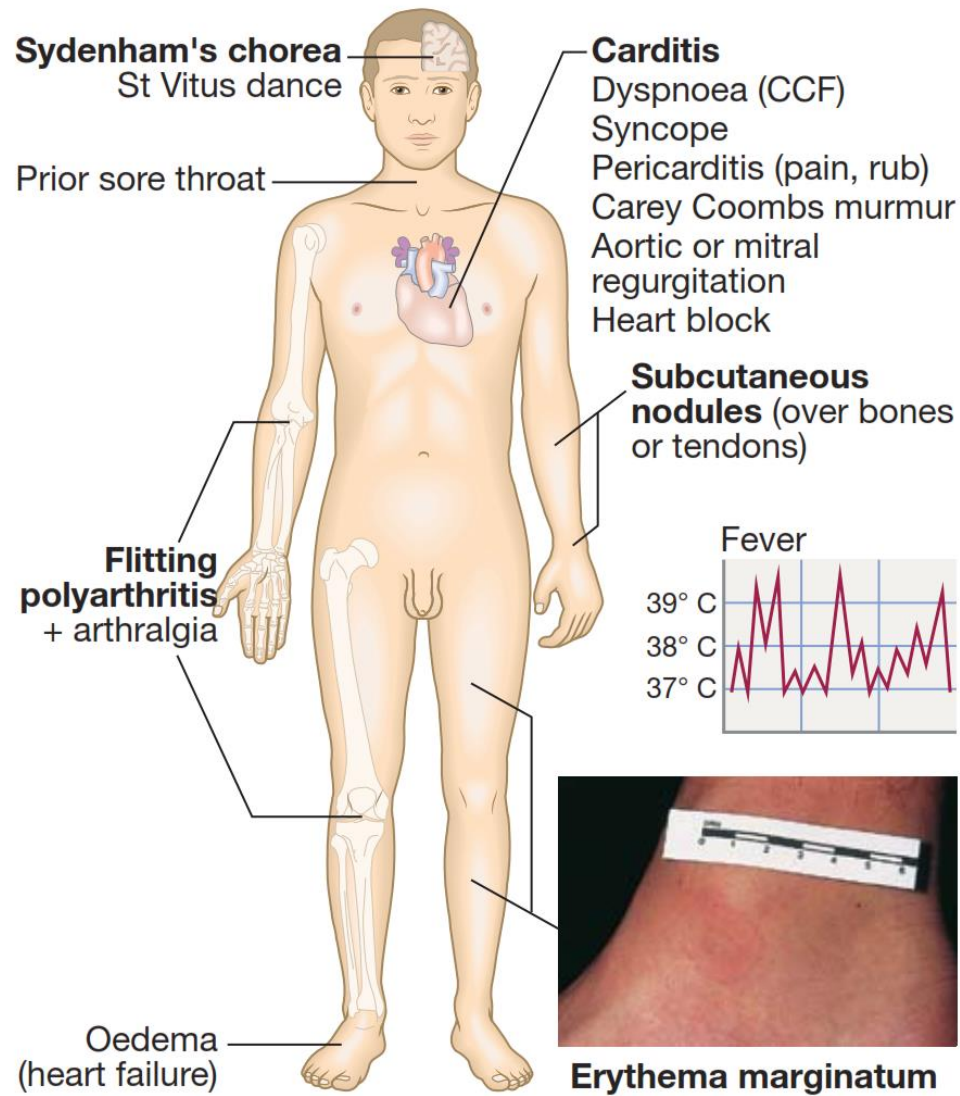
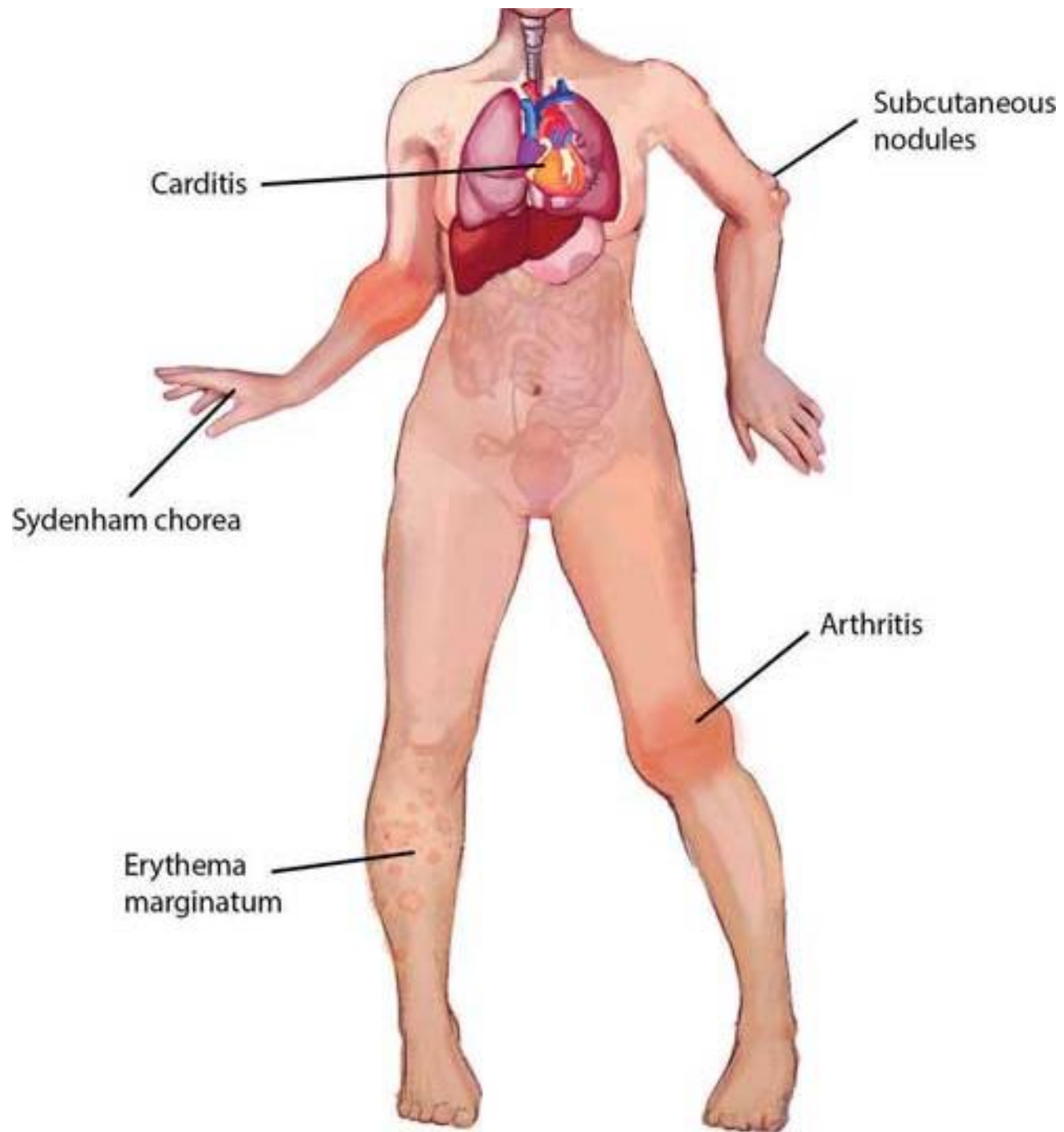
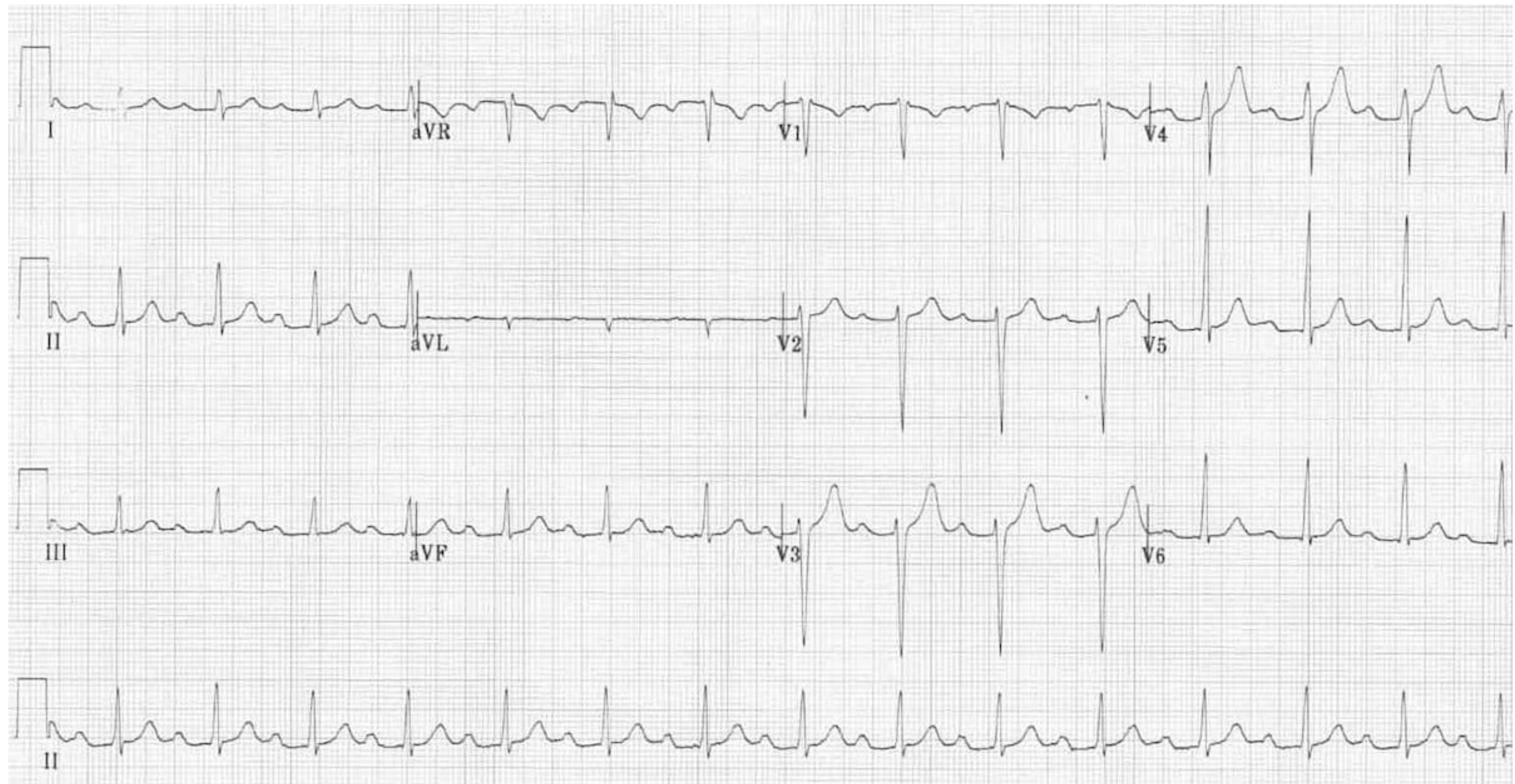


Fig. 16.80 Clinical features of rheumatic fever. Bold labels indicate Jones major criteria. (CCF = congestive cardiac failure) *Inset (Erythema marginatum)* From Savin JA, Hunter JAA, Hepburn NC. *Skin signs in clinical medicine*. London: Mosby-Wolfe, Elsevier; 1997.



Investigations

- WBC, ESR and CRP : for monitoring progress of the disease
- Throat cultures : positive results are obtained in only **10–25%**
- Antistreptolysin O antibodies (ASO) : raised titre in **80%** of adult cases of acute rheumatic fever and **normal most cases of chorea.**
(> 200 in adults , > 300 in children)
- ECG :
- Echocardiography :
- CXR : cardiomegaly , pulmonary oedema



i 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.

i 16.75 Jones criteria for the diagnosis of rheumatic fever

Major manifestations

- Carditis
- Polyarthritits
- Chorea **Late**
- Erythema marginatum
- Subcutaneous nodules **Late ; 5%**

Minor manifestations

- Fever
- Arthralgia
- Raised erythrocyte sedimentation rate or C-reactive protein
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A presumptive diagnosis can be made **without evidence of preceding streptococcal infection** in cases of isolated **chorea (Late manifestation) or carditis (especially chronic)** , if other causes of these have been excluded (Dx by exclusion)

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For all patient populations with evidence of preceding GAS infection

Diagnosis: Initial ARF	2 major manifestations or 1 major plus 2 minor manifestations
Diagnosis: Recurrent ARF	2 major or 1 major and 2 minor or 3 minor

For all patient populations with evidence of preceding GAS infection

Diagnosis: Initial ARF	2 major manifestations or 1 major plus 2 minor manifestations
Diagnosis: Recurrent ARF	2 major or 1 major and 2 minor or 3 minor

Major criteria

Low-risk populations*	Moderate- and high-risk populations*
Carditis Clinical and/or subclinical	Carditis Clinical and/or subclinical
Arthritis Polyarthritits only	Arthritis Monoarthritis or polyarthritits Polyarthralgia
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules

Minor criteria

Low-risk populations	Moderate- and high-risk populations
Polyarthralgia	Monoarthralgia
Fever ($\geq 38.5^{\circ}\text{C}$)	Fever ($\geq 38^{\circ}\text{C}$)
ESR ≥ 60 mm in the 1st hour and/or CRP ≥ 5.0 mg/dL	ESR ≥ 30 mm in the 1st hour and/or CRP ≥ 5.0 mg/dL
Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

*Low-risk populations are those with ARF incidence $\leq 2/100,000$ school-aged children or all-age rheumatic heart disease prevalence of $\leq 1/1000$ population per year. ESR: Erythrocyte sedimentation rate, CRP: C-reactive protein, ARF: Acute rheumatic fever, GAS: Group A *Streptococcus*

Management

The aims

- **Limit cardiac damage**
- Relieve symptoms

.

Bed rest

- The duration should be guided by symptoms, along with temperature, **leucocyte count and ESR**
- 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 in these cases **does not respond to medical** treatment, valve replacement may be necessary

Occasionally, AV block may occur but is seldom progressive and **usually resolves spontaneously**. Rarely, pacemaker insertion may be required.

Acute Antibiotics treatment

A **single dose** of benzathine benzylpenicillin (1.2 million U IM)
or
oral phenoxymethylpenicillin (250 mg **4 times** daily for 10 days)

If the patient is penicillin-allergic, **erythromycin or a cephalosporin** can be used.

long-term prophylaxis

oral phenoxymethylpenicillin (250 mg **twice daily**)

or

benzathine benzylpenicillin (1.2 million U IM **monthly**), if adherence is in doubt.

Sulfadiazine or erythromycin may be used if the patient is allergic to penicillin

Antibiotic prophylaxis can usually be stopped :

Without residual heart disease :

until **5 years after the last** episode or **21 years of age**, whichever is later.

With residual heart disease:

until **10 years after the last** episode or **40 years of age**, whichever is later.

But the duration of prophylaxis should be extended 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.

NOTE: While long-term antibiotic prophylaxis prevents further attacks of acute rheumatic fever, it does **not protect against infective endocarditis**

Aspirin

- 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**. (PPI is recommended for gastric protection)

e.g : 36 kg adolescence : 3600 mg = 600 mg (2 tablets 300mg) x 6

- Aspirin should be continued until the **ESR** has fallen and then **gradually** tailed off.

Glucocorticoids

- These produce more rapid symptomatic relief than aspirin and
- 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**.

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

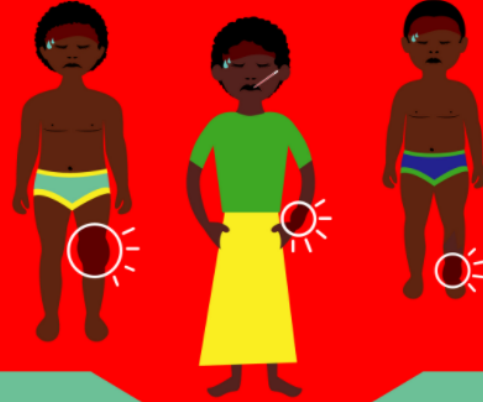
an additional 40% have mixed mitral stenosis and regurgitation.

JOINT PAIN + FEVER CAN BE: RHEUMATIC FEVER

Malaria is not the only cause of joint pain and fever.

Joint pain and fever can indicate

Acute Rheumatic Fever.



Two atipa pee n keken aye kelo arem me wang mwot kede lyeto ento twero bedo anyut me **two adunu nyo two itao.**

Acute Rheumatic Fever can damage the heart.



Two adunu nyo two itao twero balo adunu ni woko.

If your child **(3-17 yrs)** has joint pain and fever take them to the nearest health center for evaluation.



Cwal atini **(me mwaka 3 nio 18)** matye kede arem me wang mwot kede lyeto l ot yat ma cok kedi wek opim.

FREE EVALUATIONS IN LIRA REGIONAL REFERRAL HOSPITAL

Dakatal adwong me Lira
TYE ME NONO