بسم الله الرحمن الرحيم

Rheumatic Fever

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استاذ طب الاطفال

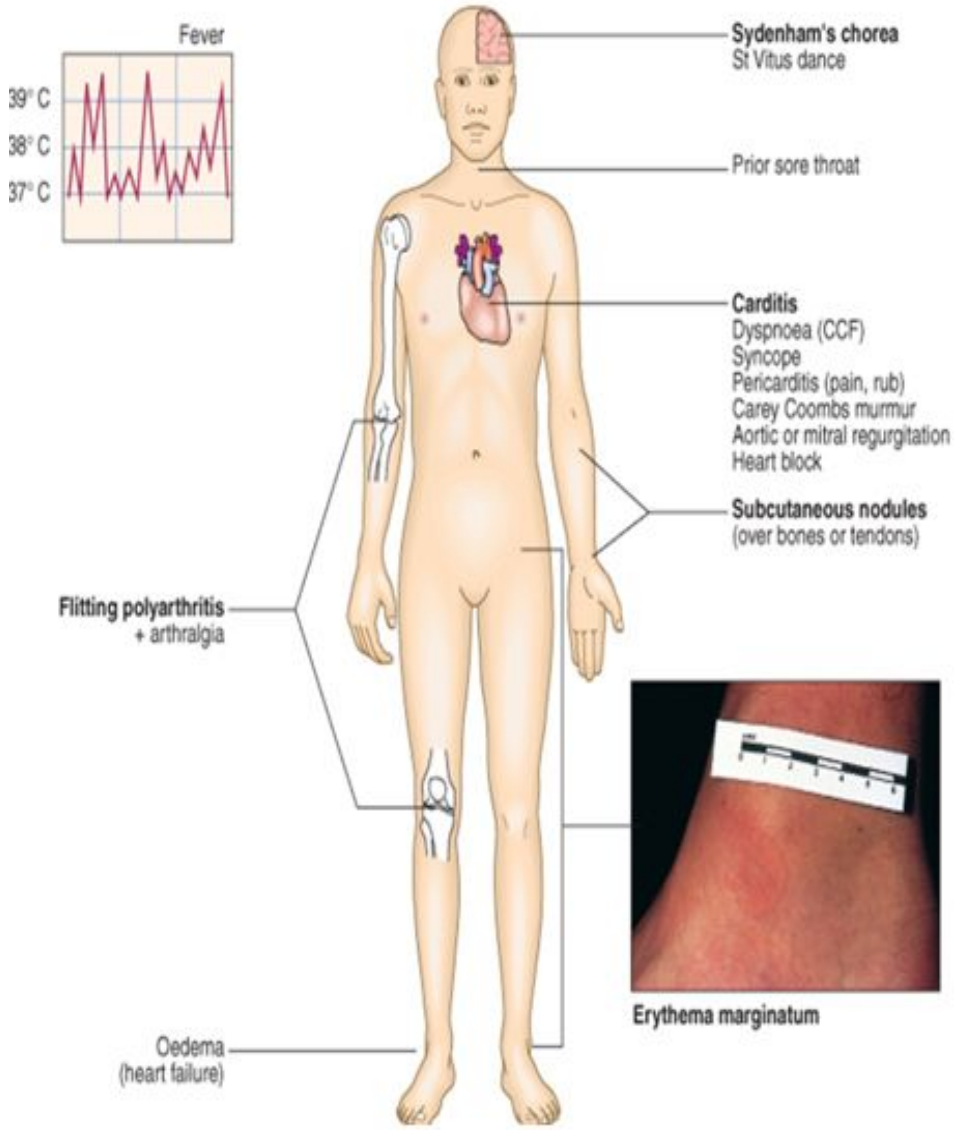
Rheumatic fever

is an inflammatory disease involving the joints, the heart, the CNS, the skin and subcutaneous tissue.

It is:

- **Serious** ⟶ as it leads to permanent cardiac damage (**chronic valvular disease**)

- **Important cause** ⟶ of acquired heart disease in children in developing countries.

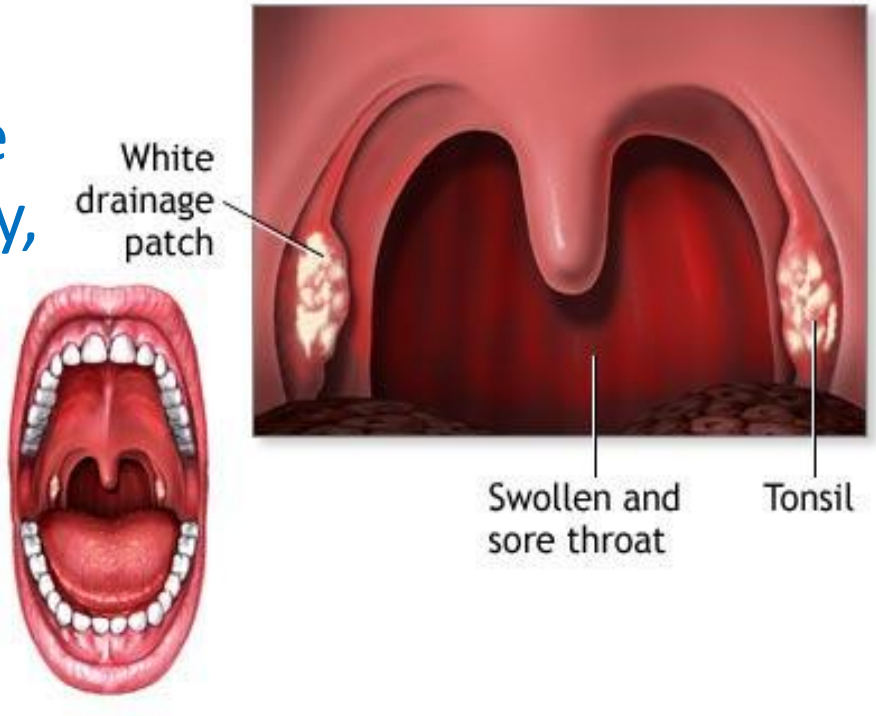


Etiology

Non-suppurative complications of upper respiratory infections by

group A-β hemolytic streptococcal (GAS)

Skin infections by GAS → acute glomerulonephritis but rarely, if ever to acute RF.

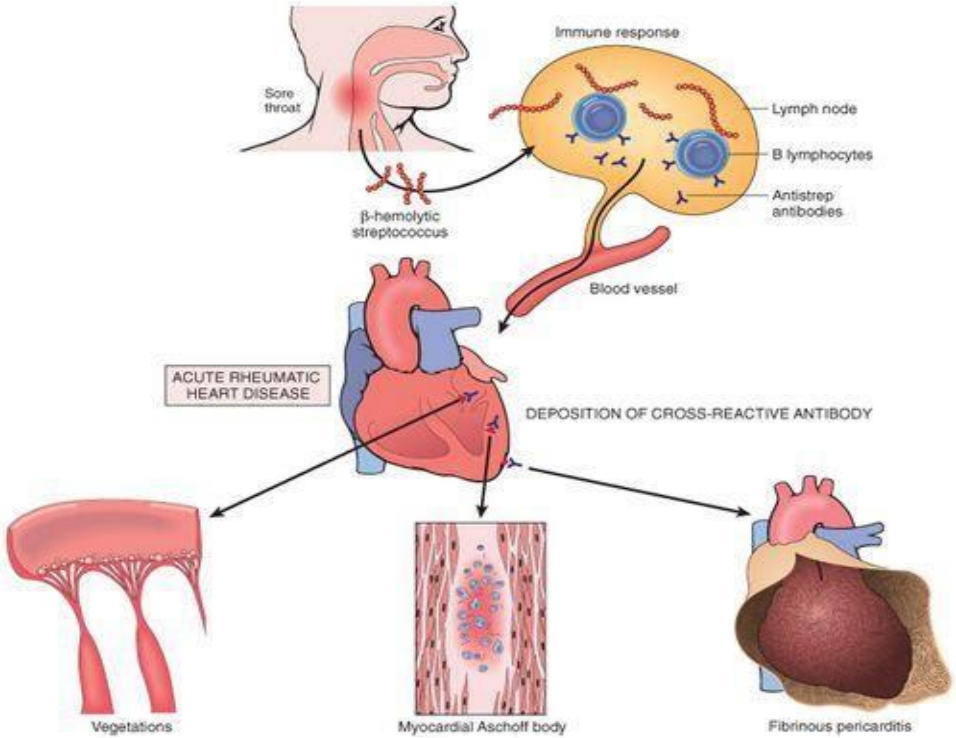


Pathogenesis

*__Autoimmune theory, abnormal immune response__ by __human host__ to some component of __GAS__.

* The resulting __antibodies__ → immunologic damage.

- Latent period is 1-3 weeks

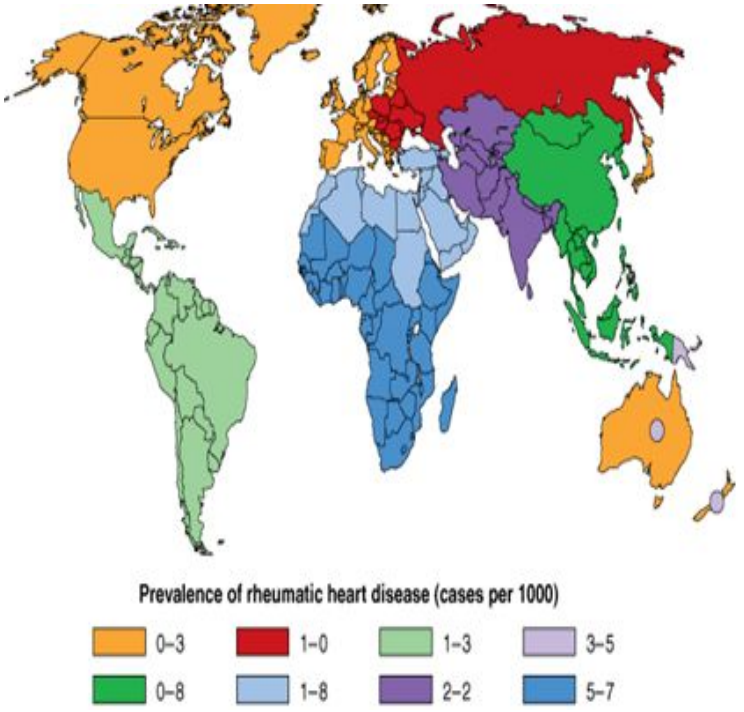


Epidemiology

Rheumatic fever occurs at all ages peaks between **5-15 ys**

A high incidence in **tropical and subtropical climates**.

Over crowding, poor housing, lack of adequate treatment and **genetic** predisposition are predisposing factors



Epidemiology

Low-Risk populations:

Those with incidence ≤2 per 100,000 school-age children per year or all-age rheumatic heart disease

prevalence of ≤1 per thousand population.

Include virtually all of the United States, Canada, and Western Europe.

Epidemiology

High-Risk populations:

Those with incidence >2 per 100,000 school-age children per year or all-age rheumatic heart disease

prevalence of >1 per thousand population.

Include Maoris in New Zealand, aborigines in Australia, Pacific Islanders, and **most developing countries**.

Clinical manifestations and diagnosis
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

A. For all patient populations with evidence of preceding GAS infection

2 Major manifestations or 1 major plus 2 minor manifestations	Diagnosis: **initial ARF**:
2 Major or 1 major and 2 minor or 3 minor manifestations	Diagnosis: **recurrent ARF**:

Clinical manifestations and diagnosis

2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

B. Major criteria

Moderate- and high-risk populations	Low-risk populations
Carditis • Clinical and/or subclinical	Carditis • Clinical and/or subclinical
Arthritis • Monoarthritis or polyarthritis • Polyarthralgia	Arthritis • Polyarthritis only
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules

Clinical manifestations and diagnosis
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

B. Major criteria

Carditis

- Clinical and/or subclinical

Arthritis

- Monoarthritis or polyarthritis
- Polyarthralgia

Chorea

Erythema marginatum

Subcutaneous nodules

Clinical manifestations and diagnosis
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

- Subclinical carditis indicates echocardiographic valvulitis.
- Erythema marginatum and subcutaneous nodules are rarely “standalone” major criteria.
- Joint manifestations can only be considered in either the major or minor categories but not both in the same patient.

Clinical manifestations and diagnosis
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

C. Minor criteria

Moderate & high-risk populations	Low-risk populations
Monoarthralgia	Polyarthralgia
Fever (≥38°C)	Fever (≥38.5°C)
ESR ≥30 mm/h in the first hour and/or CRP ≥3.0 mg/dL§	ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL§
Prolonged PR interval, after accounting for age variability	Prolonged PR interval, after accounting for age variability

Clinical manifestations and diagnosis:
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

C. Minor criteria

Monoarthralgia

Fever (≥38°C)

ESR ≥30 mm/h in the first hour and/or CRP ≥3.0 mg/dL

Prolonged PR interval, after accounting for age variability

Clinical manifestations and diagnosis
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

- CRP value must be greater than upper limit of normal for laboratory.
- Because ESR may evolve during the course of ARF, peak ESR values should be used.

Clinical manifestations and diagnosis
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

- *Prolonged P-R interval* [in the ECG] should not be counted as a minor manifestation in patients in whom *carditis* is counted as a major manifestation.
- A prolonged P-R interval alone does not constitute evidence of carditis or predict long-term cardiac sequelae.

Clinical manifestations and diagnosis

2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

Evidence of a preceding GAS infection:

1- Positive throat culture for streptococci.

Or rapid antigen test.

2 - Elevated and/or rising ASO titer.

3- Raised other streptococcal antibodies: Antideoxyribonculease B.

Clinical manifestations and diagnosis

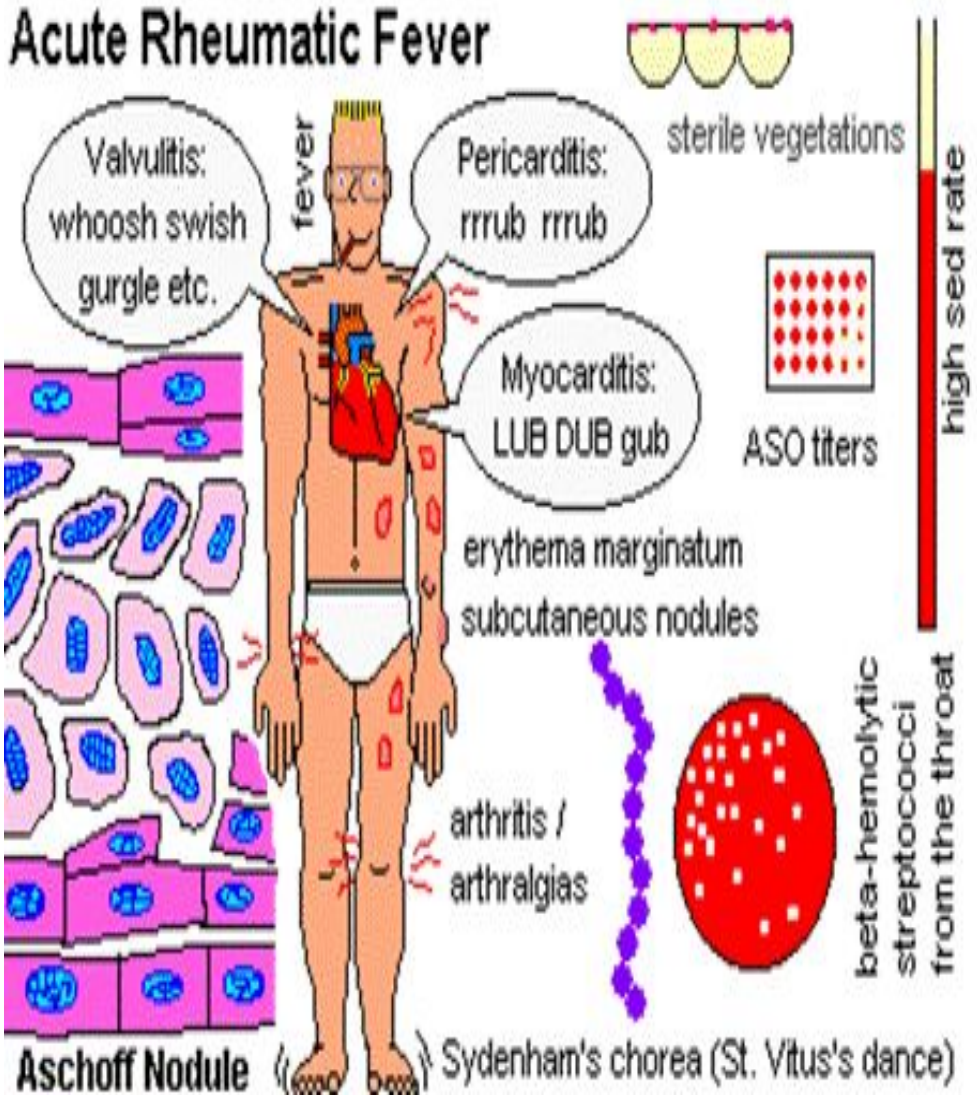
2015 Revised Jones Criteria for diagnosis of Rheumatic Fever

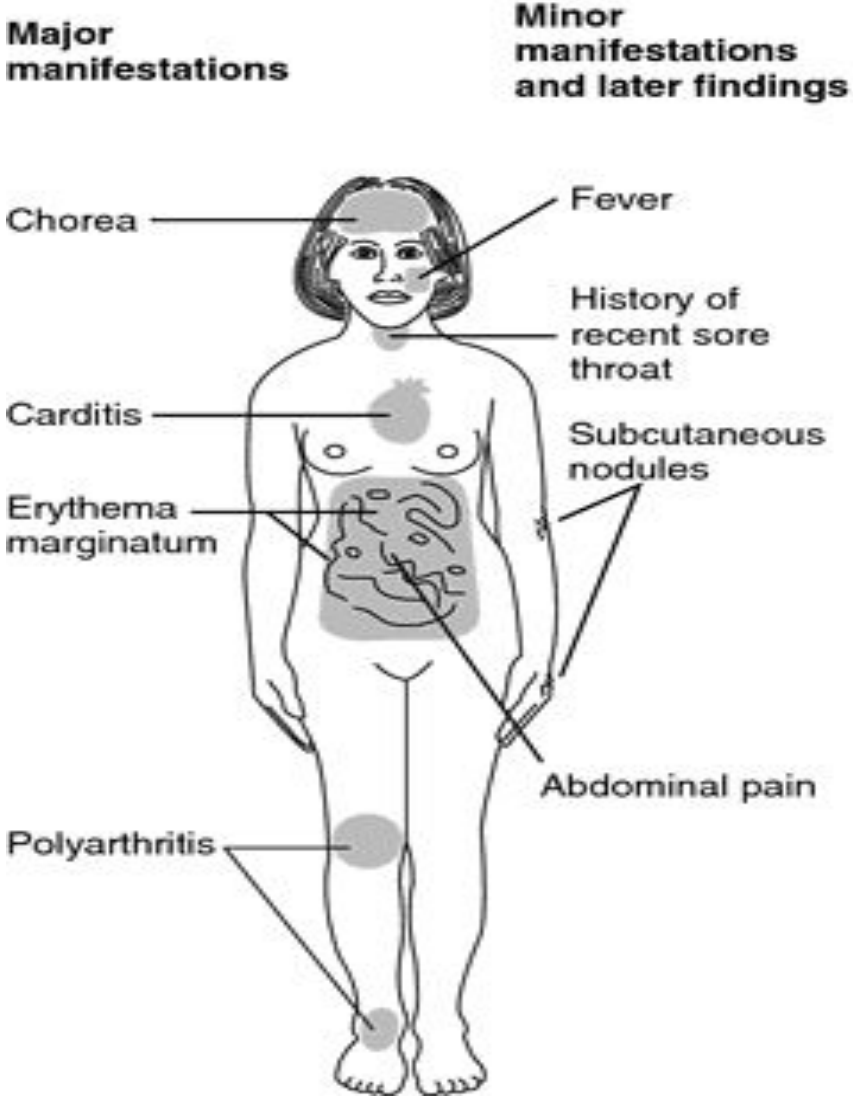
The diagnosis of acute rheumatic fever *should not* be made in those patients with elevated or increasing ASOT who do not fulfill the Jones criteria.

Guidelines for the diagnosis of initial or recurrent attack of RF

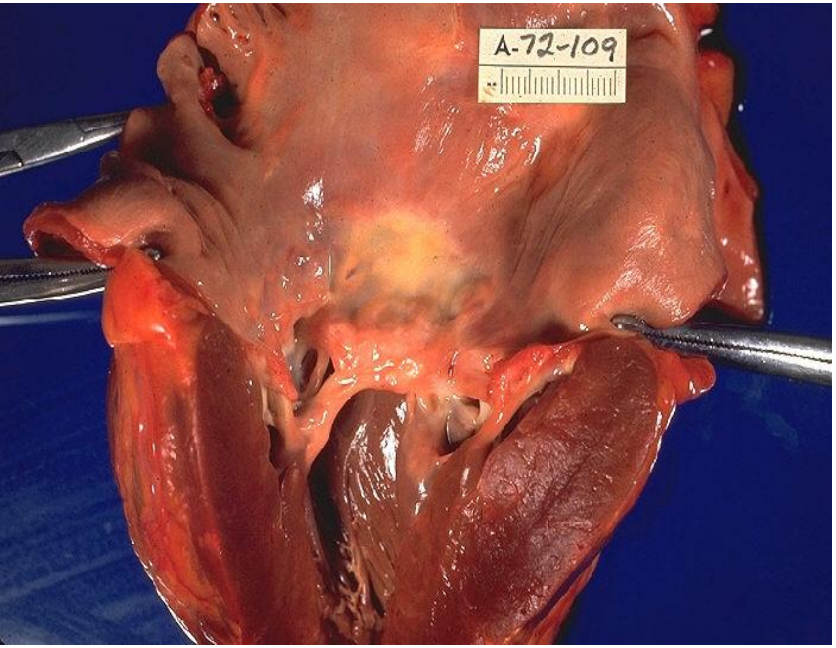
Revised Jones criteria, Updated 2015

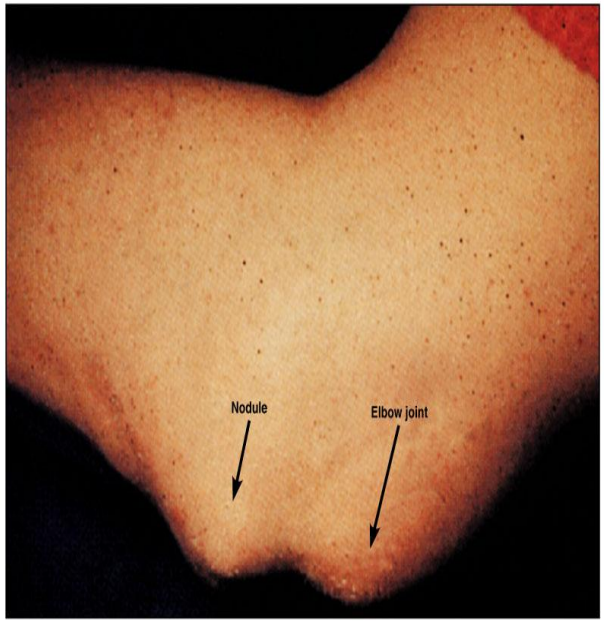
Evidence of preceding GAS	Minor	Major
Positive throat culture for streptococci Or rapid streptococcal antigen test Elevated or increasing ASOT	* Clinical features: Arthralgia Fever * Laboratory features: *elevated* acute phase reactant ↑ ESR ↑ C reactive protein * Prolonged PR interval	- Carditis - Polyarthritis - Erythema marginatum -Subcutaneous nodules - Chorea


Acute Rheumatic Fever
Valvulitis: whoosh swish gurgle etc.
fever
Pericarditis: rrrub rrrub
Myocarditis: LUB DUB gub
sterile vegetations
ASO titers
high sed rate
erythema marginatum
subcutaneous nodules
arthritis / arthralgias
beta-hemolytic streptococci from the throat
Aschoff Nodule
Sydenham's chorea (St. Vitus's dance)

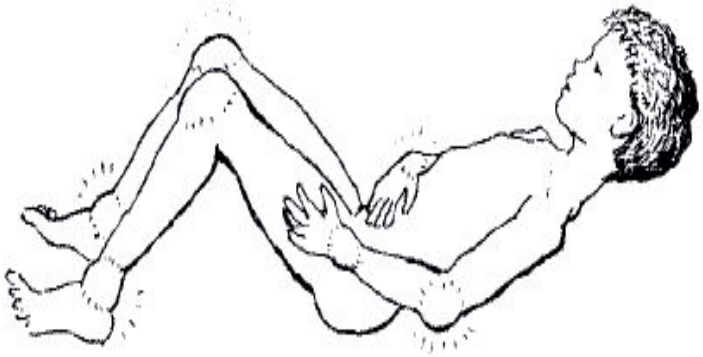

Major manifestations
Minor manifestations and later findings
Chorea
Fever
History of recent sore throat
Carditis
Subcutaneous nodules
Erythema marginatum
Abdominal pain
Polyarthritis

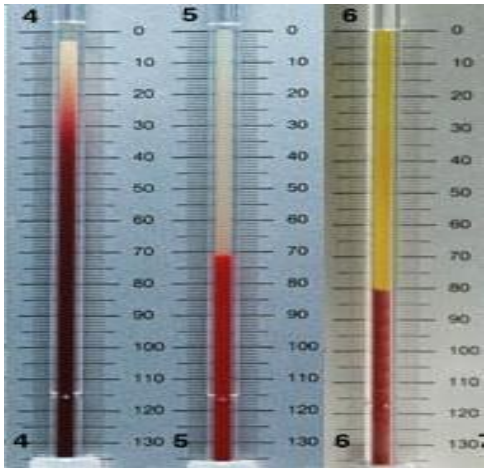
Major Jones criteria

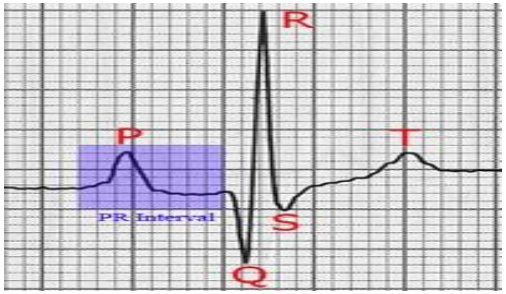




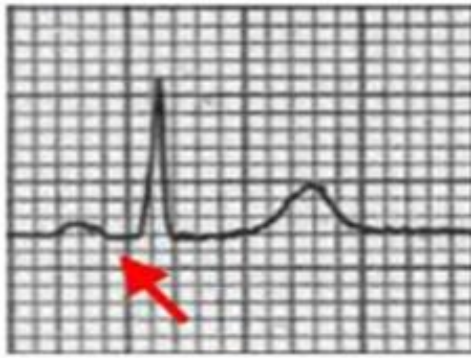
Minor Jones criteria



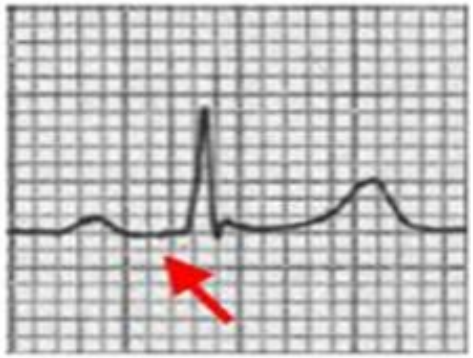




a) Normal PR



b) Prolonged PR



Rheumatic Carditis

- **Most serious manifestations of ARF**
- **Occurs in about 50-60% of all cases of ARF**
- **Pancarditis involves endocardium, myocardium and pericardium**
- **Result in residual chronic valvular lesion**
- **Rheumatic Subclinical Carditis is carditis without a murmur of valvulitis but with echocardiographic evidence of valvulitis.**

Endocarditis (valvulitis)

* Numerous small rheumatic sterile vegetation on the line of closure of the valves

* Mitral valve is commonly affected followed by the Aortic valve

* Valvular regurge is characteristic of ARF

* Valvular stenosis usually appears several years after the ARF

Murmurs in patients with endocarditis

soft - musical - not associated with thrill

1- Apical pansystolic murmur, musical
soft, radiating to axilla, changeable, not associated with thrill,
caused by mitral valvitis → (**MR**)
disappears within 6 months if not associated with chronic MR

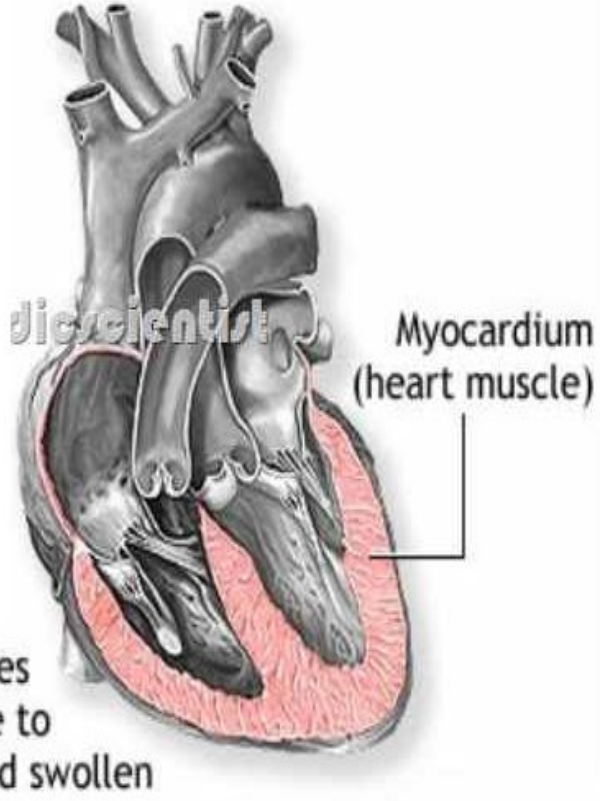
2- Apical low-pitched mid-diastolic murmur → (**MS**)
(**Carey Coombs murmur).**

3- Early diastolic murmur over the aortic area → (**AR**)

4- Appearance of new murmurs.

5- Change in character of previous murmurs.

Myocarditis

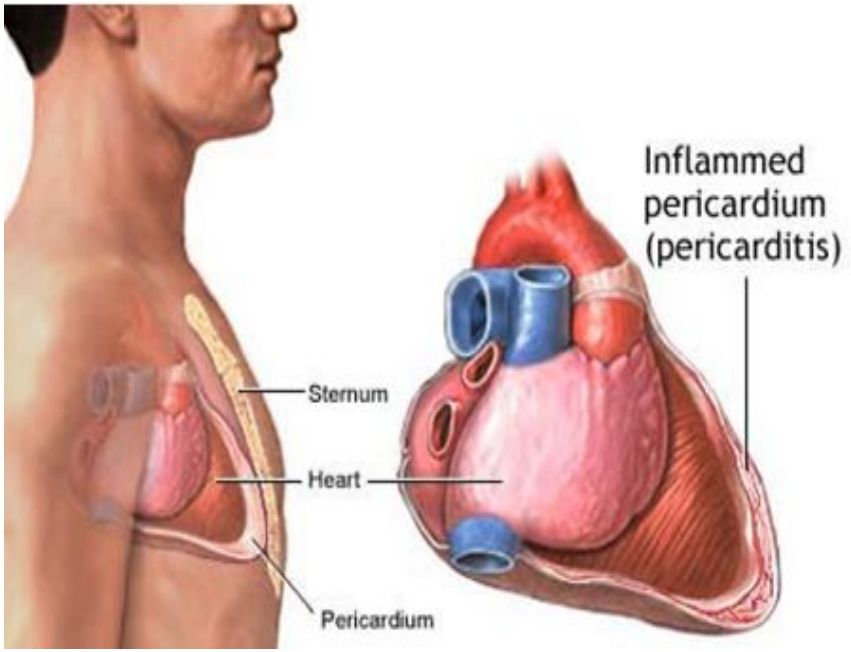


- Cardiac dilatation
- Congestive heart failure
- Tachycardia disproportionate to fever
- Arrhythmias
- Muffled Heart sounds

Pericarditis

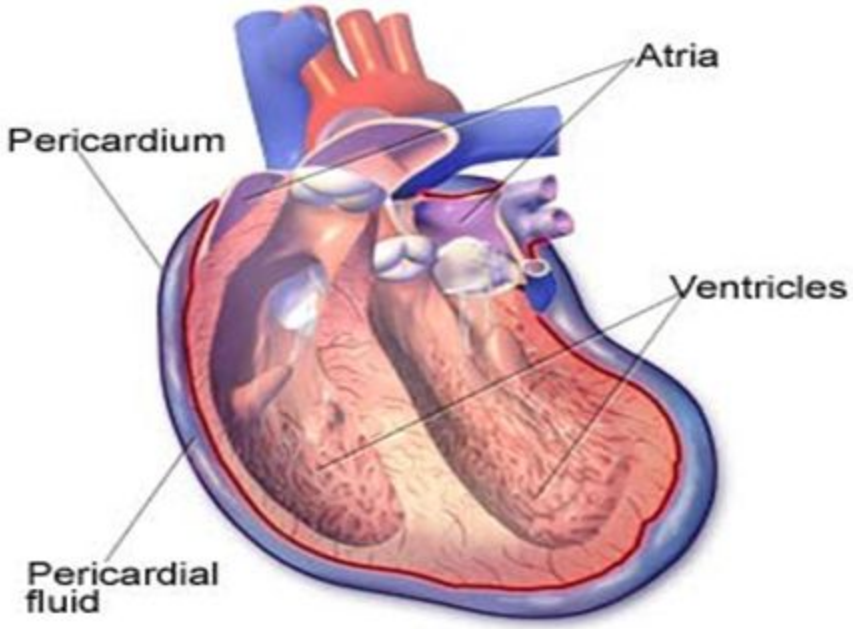
Dry pericarditis

precordial pain and friction rub



Wet Pericarditis

mild to moderate effusion

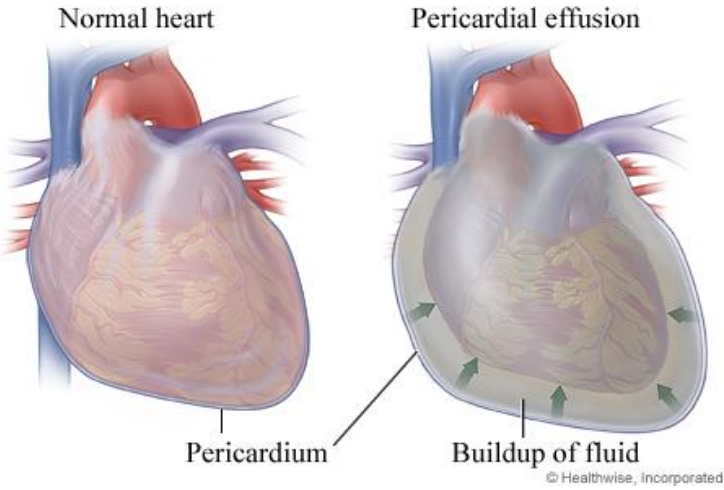


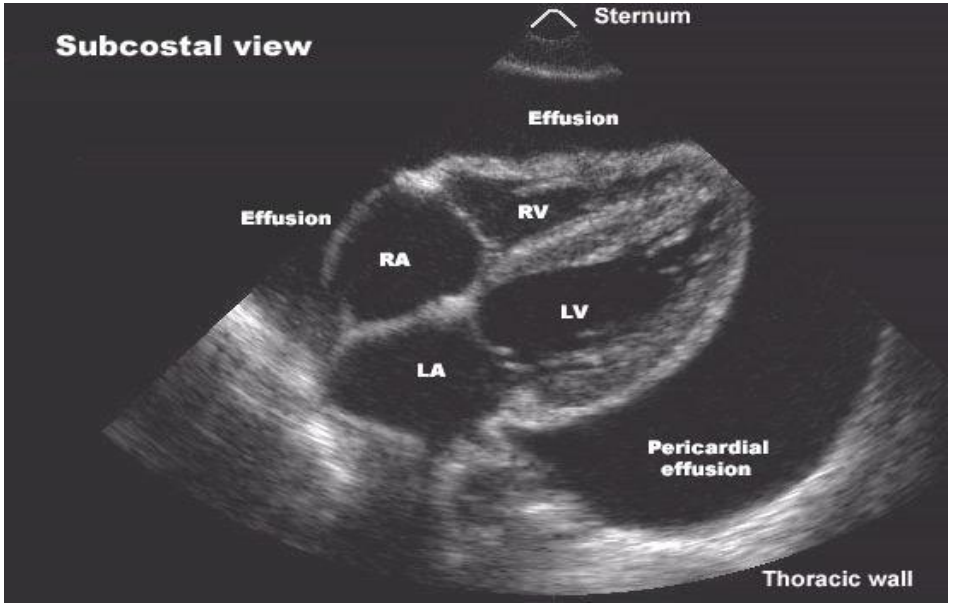
Pericarditis with massive pericardial effusion:

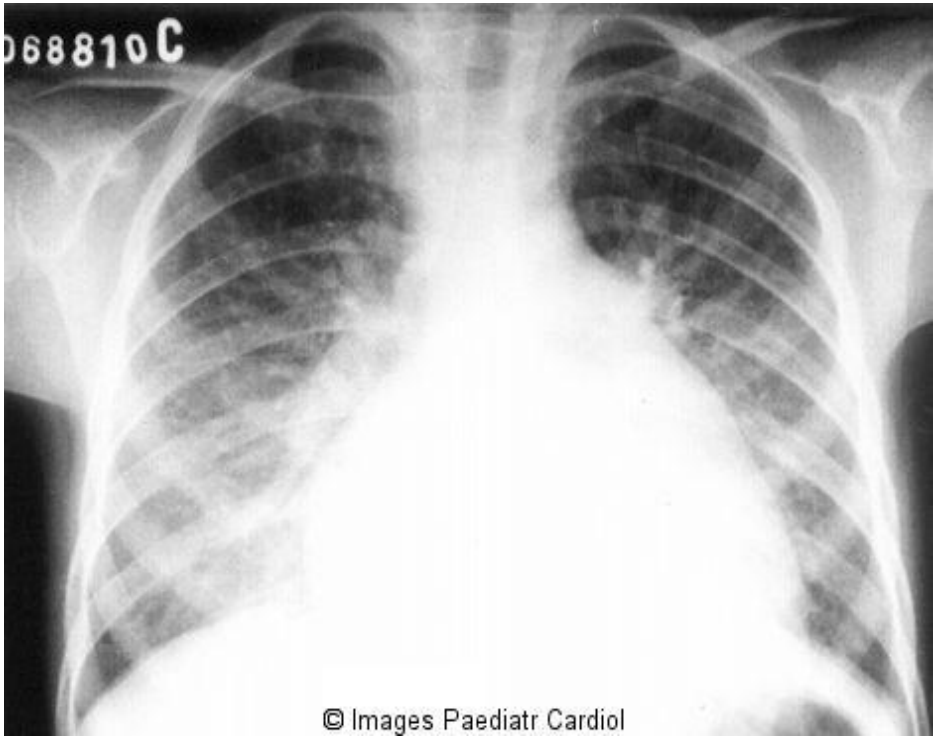
- Weak pulse.
- Pulsus paradoxus

  is an abnormally large decrease in systolic BP during inspiration. The normal fall in systolic BP is <10 mmHg. When the drop is >10mmHg, it is referred to as pulsus paradoxus.
- Congested non pulsating neck veins.
- Weak apical pulsation
- Heart sounds are distant and muffled.
- Dullness outside the apex
- Ewart's sign

compression of the left lung produces dullness and bronchial breathing at the lung base posteriorly.




Subcostal view
Sternum
Effusion
Effusion
RV
RA
LV
LA
Pericardial effusion
Thoracic wall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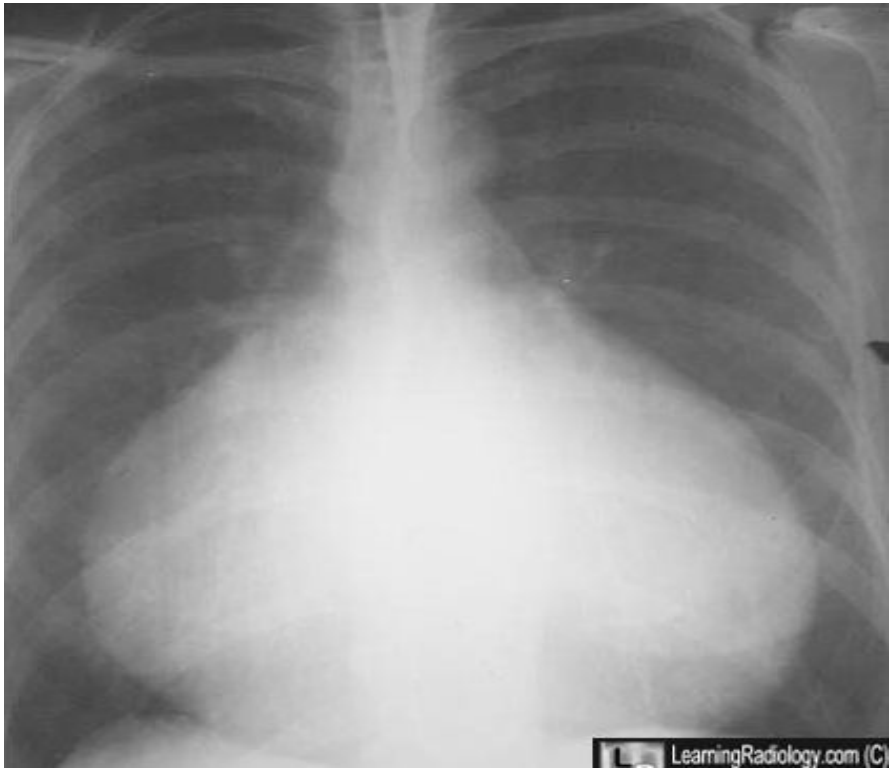
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DD of rheumatic carditis:

1. Other causes of myocarditis such as viral myocarditis.
2. Other causes of pericarditis.
3. Infective endocarditis.
4. Congenital heart disease.

Treatment of carditis

* Mild cases without HF or cardiomegaly

Salicylates **50-70mg/kg/day** orally after meal divided into 4 doses for 3-5 days, then **50 mg/kg/day** divided into 4 doses for 3 weeks, then **25 mg/kg/day** for 3 weeks

* Moderate and severe cases with cardiomegaly and/or HF:

- Bed rest

- **Prednisone** **2 mg/kg/day** divided in 4 doses for 2-3 wk., then **half the dose** for 2-3 weeks

When the patient responds clinically & on lab tests (ESR, CRP), the dose should be tapered by reducing 5 mg/day every 2-3 days.

-**Salicylates** **50mg/kg/day** in 4 divided doses for 6 weeks at the beginning of tapering steroid dose to prevent rebound

-**Supportive therapies** include digoxin, fluid, salt restriction, diuretics and O_2.

Rheumatic Polyarthritis

-Migratory (from one joint to another)& affects several joints.
-Involves large joints.
-Mainly symmetrical.
-The joints are tender, red, warm and swollen.
-Effusion may be present.
-Does not result in chronic joint disease.
-Aspirin or NSAI give dramatic response in 12-24 hours.
-Arthralgia may occur in some joints and frank arthritis in others.

Differential diagnosis:

Other causes of arthritis

- Juvenile rheumatoid arthritis & other collagen diseases.
- Infective endocarditis.
- Arthritis. Of infection
- Malignancy as Leukemia.
- Sickle cell disease.

Treatment of arthritis

Salicylates **50-70mg/kg/day** orally after meal divided into 4 doses for 3-5 days, then **50 mg/kg/day** divided into 4 doses for 3 weeks, and **25 mg/kg/day** for 3 weeks.

Early administration of salicylates to a patient before diagnosis is established may obscure the diagnosis.

Rheumatic Chorea

(Sydenham chorea)

Has long latent period (2-6 months), the onset is usually insidious.

May be the only sign of rheumatic fever (pure chorea) or it may be associated with carditis.

Clinical manifestation:

- Characterized by involuntary movements, emotional liability & hypotonia.
- Purposeless, irregular, rapid, jerking movements of the limbs and grimacing of the face
- Exaggerated by emotional stress and disappear during sleep.
- Drop things, spill from a cup and handwriting deteriorates.
- Speech is commonly slurred.
- May affects one side of the body (hemichorea).

Sydenhams chorea

watch please

Clinical tests for detection of chorea

- **Marked fluctuation in muscle tone**
  (felt by asking the patient to squeeze the examiner's hand).

- **Spooning:**

- **When the tongue is protruded**
  it is rapidly withdrawn to prevent being bitten by involuntary jaw movements.

- **The knee jerk:**
  either of pendulum type(due to hypotonia) or more commonly is sustained or "hung up".

- **Pronation sign:**
  on elevation of the upper limbs above the level of the head with the palms of hands facing each other, there is pronation in the forearms and the limbs fall down gradually.

- **Examination of hand writing** for fine motor movement

In pure chorea the ESR and ASOT are normal. This is attributed to the long latent period (2-6 months), when elevated loog for carditis

DD

Postencephalitic chorea

Cerebral palsy

Prognosis

Chorea is a self limited condition.

Mild cases subside within few weeks - 3 months

Severe cases may progress and require a padded cot.

Treatment of rheumatic chorea

- Anti inflamatory

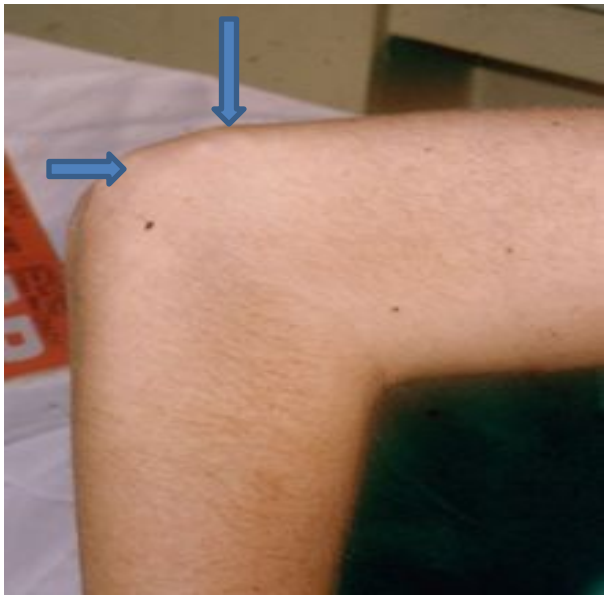
- Phenobarbital

- Haloperidol

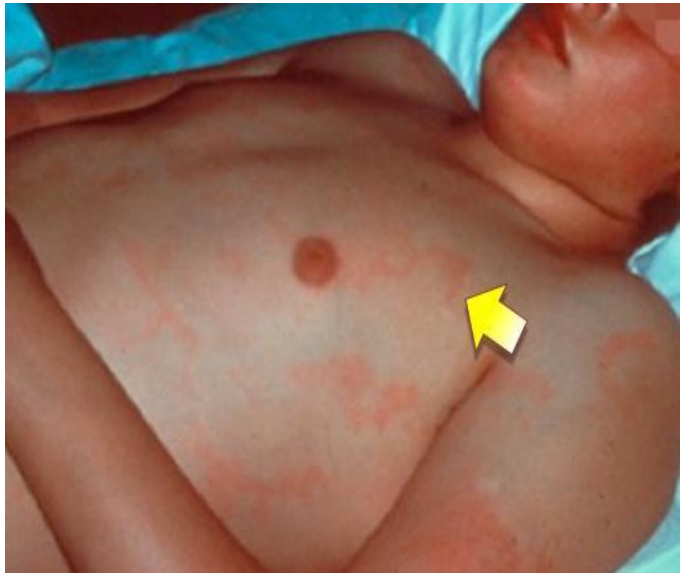
- Chlorpromazine

Subcutaneous nodules

- Bilaterally symmetrical firm nodules varying in diameter from few millimeters to centimeter.
- Movable painless and not tender.
- Occur over the bony prominences.
- Best demonstrated by fully flexing the joint and stretching the skin over the extensor surface.
- When occurs, usually severe carditis is present.



Erythema marginatum



- Red, raised, non pruritic macules extend to form wavy lines or rings with pale centers .
- Coalesce forming irregular patterns, which vary in shape, and site from hour to hour.
- Usually seen over the trunk.

Complications of Acute Rheumatic fever

- Chronic valvular heart disease **(RHD)** after an attack of rheumatic carditis.

- Severe acute carditis is the commonest cause of **death** of rheumatic fever.

Prevention of rheumatic fever

can be divided into three approaches

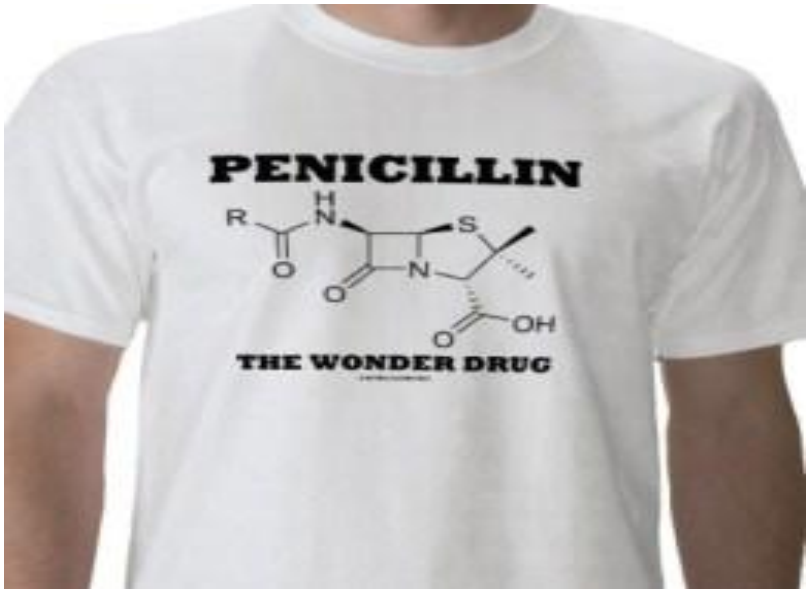
1. General measures
2. Primary prevention
3. Secondary orevention

1. Treatment (eradication) of *GAS* infection

Treatment of streptococcal upper respiratory tract infection **must be within 9 days** ***to prevent an initial attack of rheumatic fever.***

Primary prevention eradication of GAS

all patients with ARF should receive



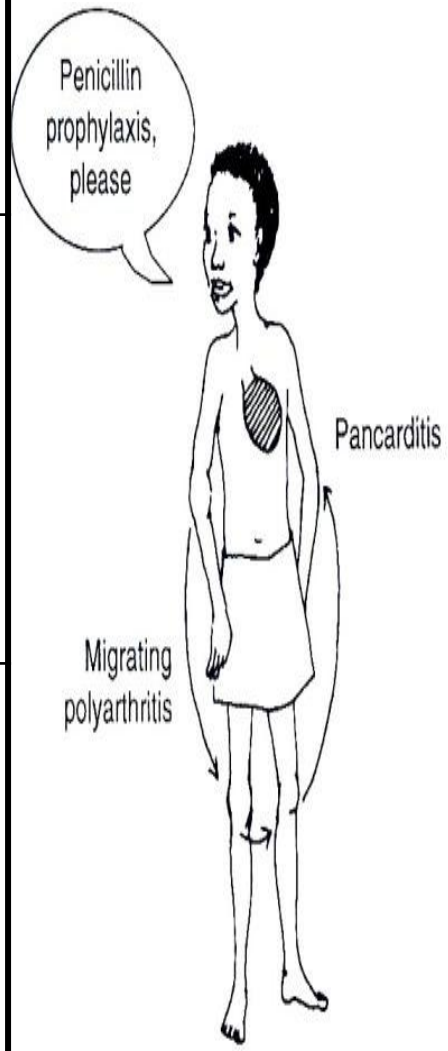
Frequency	Route of administration	Dose	Antibiotic
Once	IM	1.2 million units (weight > 27 kg) 600.000 units (weight < 27 kg)	**Benzathine penicillin G**
Twice daily for 10 days	Oral	250 mg/dose	**Penicillin V (oral penicillin)**
3 doses for 10 days	Oral	40mg / kg/ 24 h	**Erythromycin**

Secondary prevention
(for recurrences of acute rheumatic fever)

Frequency	Route of administration	Dose	Antibiotic
Every 2-3 weeks	IM	1.2 million units (weight > 27 kg) 600.000 units (weight < 27 kg)	Benzathine penicillin G
Twice daily	Oral	250mg	Penicillin V
Once daily	Oral	0.5-1 gm	Sulfadiazine
Twice daily	Oral	250 mg	Erythromycin for allergic patients

Duration of Secondary prevention

5 yr or until 21 yr of age, whichever is longer	RF without carditis
10 yr or well into adulthood, whichever is longer	RF with carditis but without residual heart disease (no valvular disease clinicaly or by echo)
At least 10 yr since the last episode and at least until 40 yr of age, or lifelong prophylaxis	RF with carditis and residual heart disease (persistent valvular disease)



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