

RHEUMATIC FEVER

The portal of entry is usually the fauces.

- ❑ Sore throat
- ❑ Frank scarlet fever
- ❑ Otitis media
- ❑ Other streptococcal infection precede the onset of the disease by two to three weeks
- ❑ The interval between infection and symptoms suggest a hyper-sensitivity state

RHEUMATIC FEVER

- Group A streptococcus (e.g., M types 1, 3, 5, 6, 18, 24) are more frequently isolated from patients with acute rheumatic fever.
- The attack rate of acute rheumatic fever - Ranges from 0.3 to 3 percent.

+242



RHEUMATIC FEVER

- The incidence of both initial attacks and recurrences of acute rheumatic fever peaks in children aged 5-15 yr.
- Patients who have had one attack of acute rheumatic fever tend to have recurrences
- There is association between the presence of specific HLA markers and susceptibility to acute rheumatic fever.

RHEUMATIC FEVER

Predisposing factors -

- ❑ low socioeconomic status
- ❑ overcrowding
- ❑ poor nutrition
- ❑ poor hygiene
- genetic predisposition.

RHEUMATIC FEVER

Incidence of rheumatic fever is on decline for following reasons-

- ❑ Improvements in living conditions
- ❑ Large measure to the greater availability of medical care.
- ❑ The widespread use of antibiotics.
- ❑ Antibiotic therapy of group A streptococcal pharyngitis has been important in preventing initial attacks.

Studies show a dramatic decline in developed countries

- A- antibiotic coverage has increased
- B- Better housing
- C- Conditions (economic & health) have improved
- D- Decreased bacterial virulence
- E- Easy access to medical care

RHEUMATIC FEVER

A triad of syndromes may result

- *The joints*- Rheumatic fever
(Rheumatic Polyarthritis)
- *Heart*-Rheumatic pancarditis
- *Brain* - Chorea

Poly arthritis is migratory.

Rheumatic fever is one of the cause of migratory poly arthritis

RHEUMATIC FEVER

Pathogenesis

The cytotoxicity theory -

- streptolysin O has a direct cytotoxic effect on mammalian cells in tissue culture.
- Its inability to explain the latent period between an episode of group A streptococcal pharyngitis and the onset of acute rheumatic fever.



3M



RHEUMATIC FEVER

The immunologic theory-

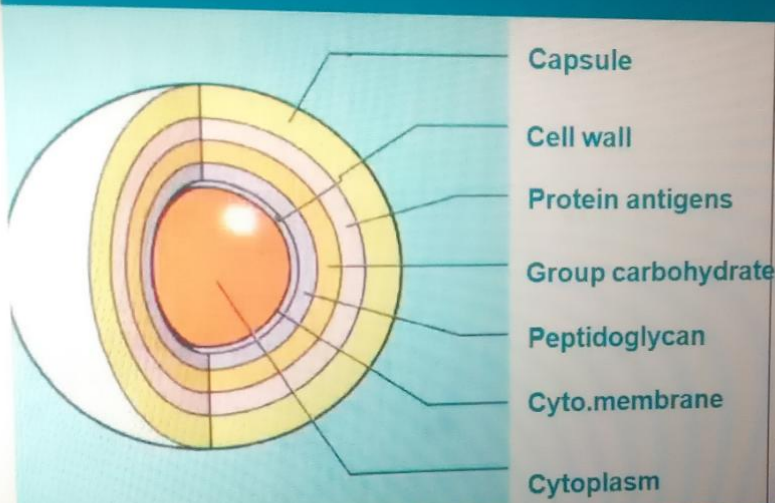
- The latent period between the group A streptococcal infection and the acute rheumatic fever.
- The antigenicity of a large variety of group A streptococcal products and constituents, as well as the immunologic cross-reactivity between group A streptococcal components and mammalian tissues.



3M



Diagrammatic structure of the group A beta hemolytic streptococcus



Antigen of outer protein cell wall of GABHS induces antibody response in victim which result in autoimmune damage to heart valves, subcutaneous tissue, tendons, joints & basal ganglia of brain



3M



RS

Pathology

□ Inflammatory Lesions in

1. Heart
2. Brain
3. Joints
4. Skin

Aschoff bodies in the atrial myocardium-

- Swelling, fragmentation of collagen fibers
- Alterations in the staining characteristics of connective tissues.



3M



RS



Myocardial Aschoff body – the cells are large, elongated, with large nuclei; some are multinucleate

T. Duckett Jones criteria

- Intended only for the diagnosis of the initial attack of acute rheumatic fever and not for recurrences.
- Five major and four minor criteria
- **When a patient fulfills two major criteria or one major and two minor criteria and meets the absolute requirement for evidence of recent GAS infection.**

3M

○

○

○

○

RS

Guidelines for the diagnosis of Initial attack of Rheumatic fever

Major Manifestations	Minor Manifestations
Carditis	Clinical findings-
Polyarthrititis	Arthralgia
Chorea	Fever
Erythema marginatum	Lab Findings-
Subcutaneous nodules	Elevated acute phase reactants- ESR,CRP
	Prolonged PR interval

Majors

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

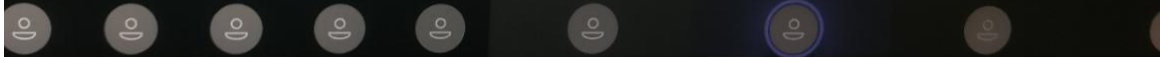
Pnemonic

C²ASE

- C – Carditis
- C – Chorea
- A – Arthritis
- S – Subcutaneous nodules
- E – Erythema marginatum

Three Circumstances – Where ARF diagnosed without strict adherence to Jones criteria

1. Indolent carditis may be sole manifestation
2. Chorea may be the sole manifestation
3. ARF recurrence may not fulfill the Jones criteria



Clinical Features

Arthritis

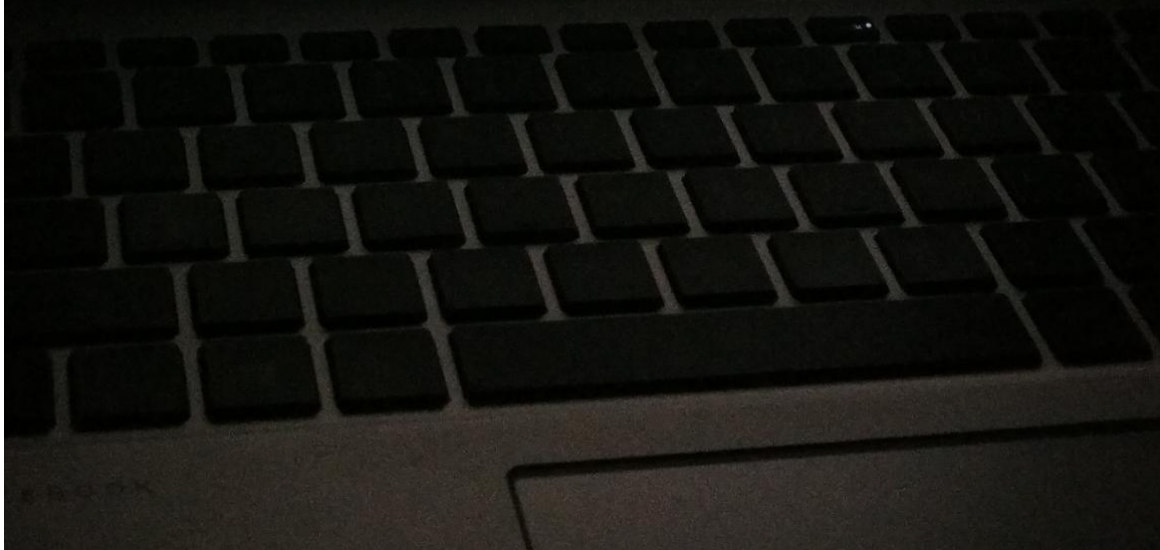
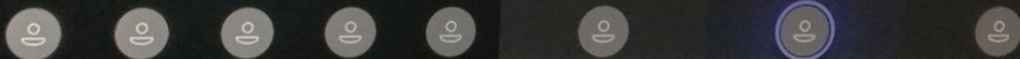
- Flitting & fleeting migratory polyarthritis, involving major joints
- Commonly involved joints-knee, ankle, elbow & wrist
- Occur in 80%,involved joints are exquisitely tender
- *In children below 5 yrs arthritis usually mild but carditis more prominent*
- *Arthritis do not progress to chronic disease*

Migratory Polyarthrititis

- Involves larger joints, particularly the knees, ankles, wrists, and elbows.
- Rheumatic joints are generally hot, red, swollen, and *exquisitely tender*.
- *the pain* can precede *and* can appear to be disproportionate to *the other findings*.
- *The joint involvement is* characteristically migratory in nature

Migratory Polyarthrititis

- A dramatic response to even small doses of salicylates .
- The absence of such a response should suggest an alternative diagnosis.
- Rheumatic arthritis is typically not deforming.
- Arthritis is the earliest manifestation of acute rheumatic fever.



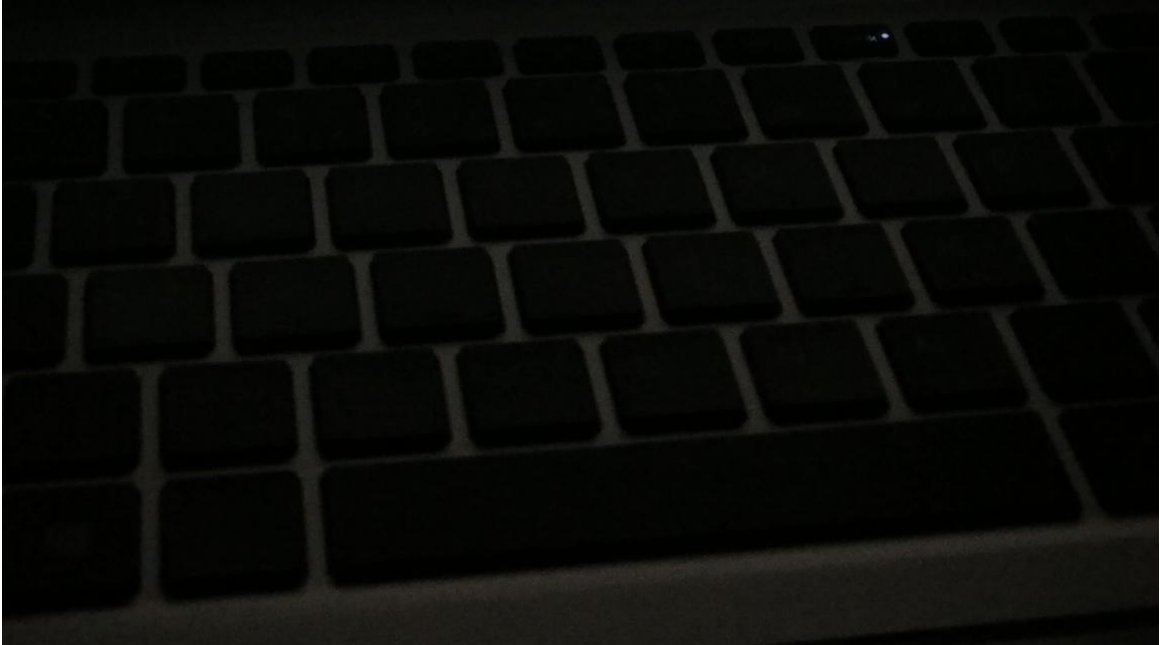
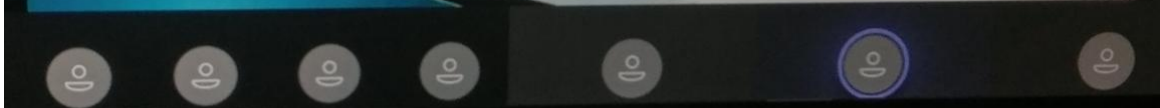
Migratory Polyarthrititis

- A dramatic response to even small doses of salicylates .
- The absence of such a response should suggest an alternative diagnosis.
- Rheumatic arthritis is typically not deforming.
- Arthritis is the earliest manifestation of acute rheumatic fever.

High dose salicylates

Carditis

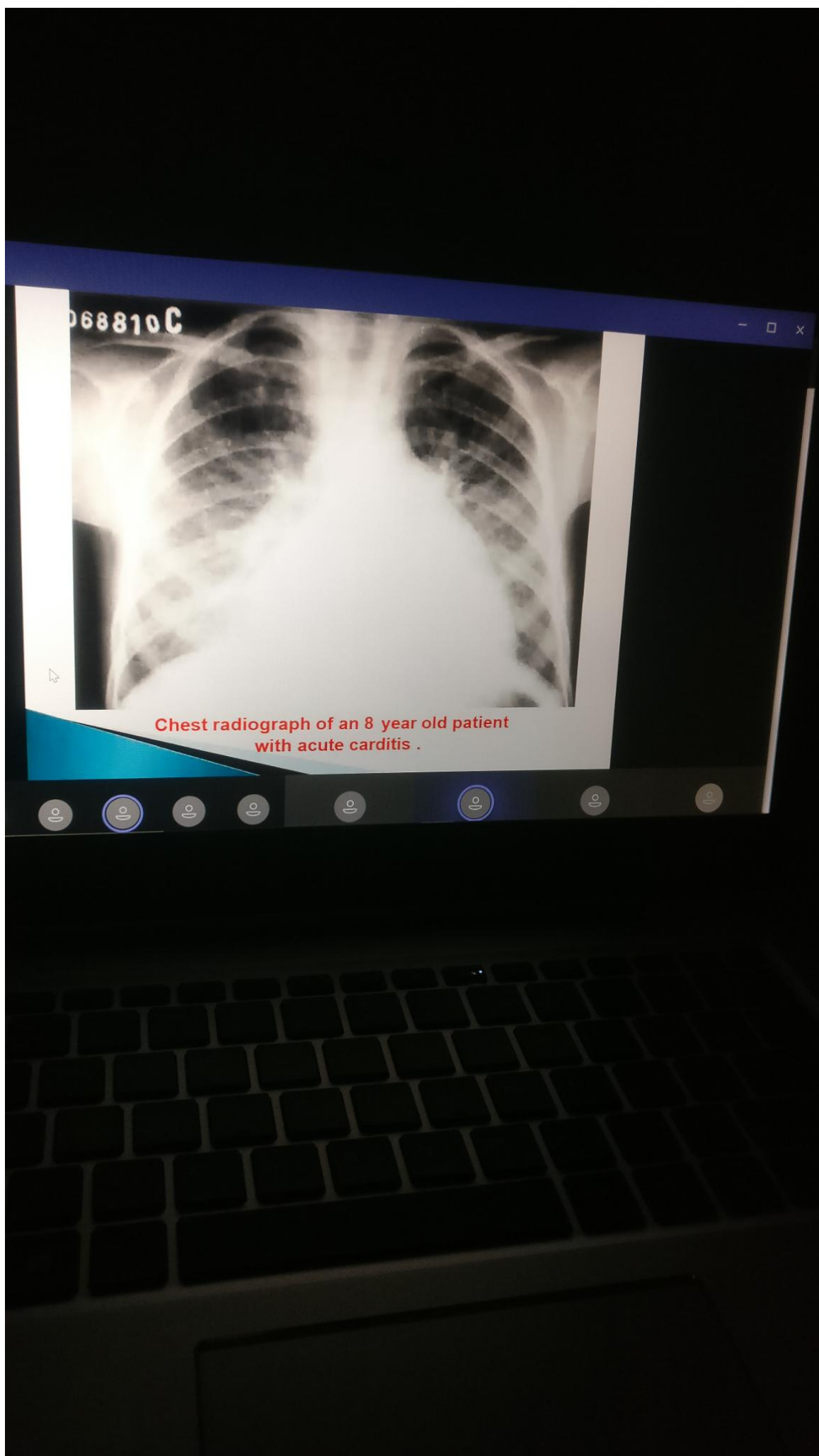
1. Tachycardia
2. A heart murmur of valvulitis
3. Pericarditis
4. Cardiomegaly on X-ray chest
5. Signs of CHF



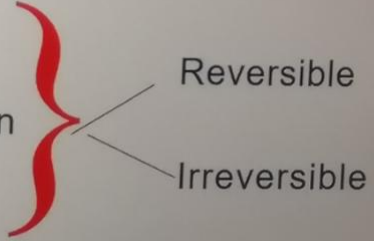
Carditis

1. Tachycardia
2. A heart murmur of valvulitis
3. Pericarditis
4. Cardiomegaly on X-ray chest
5. Signs of CHF

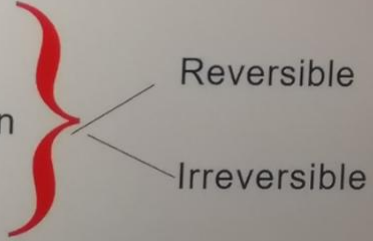
Signs of both RHF and LHF



Carditis presentation

- Tachycardia out of proportion to fever
sleeping pulse rate raised
 - Pericardial rub
 - CCF, gallop rhythm and so on
 - Cardiac enlargement
 - Pulmonary hypertension
 - Cardiac murmur
- 
- Reversible
- Irreversible

Carditis presentation

- Tachycardia out of proportion to fever
sleeping pulse rate raised
 - Pericardial rub
 - CCF, gallop rhythm and so on
 - Cardiac enlargement
 - Pulmonary hypertension
 - Cardiac murmur
- 
- Reversible
- Irreversible

1 degree rise in temp causes 10 bpm Inc in HR

Murmur

- High pitched apical holosystolic murmur radiating to axilla – Mitral regurgitation
- An apical mid diastolic murmur
- A high pitched decrescendo diastolic murmur- upper sternal border- Aortic regurgitation

+291

Carditis sequelae (chronic)

Mitral insufficiency

- Some loss of valvular substance
- Shortening & thickening of Chordae tendinae

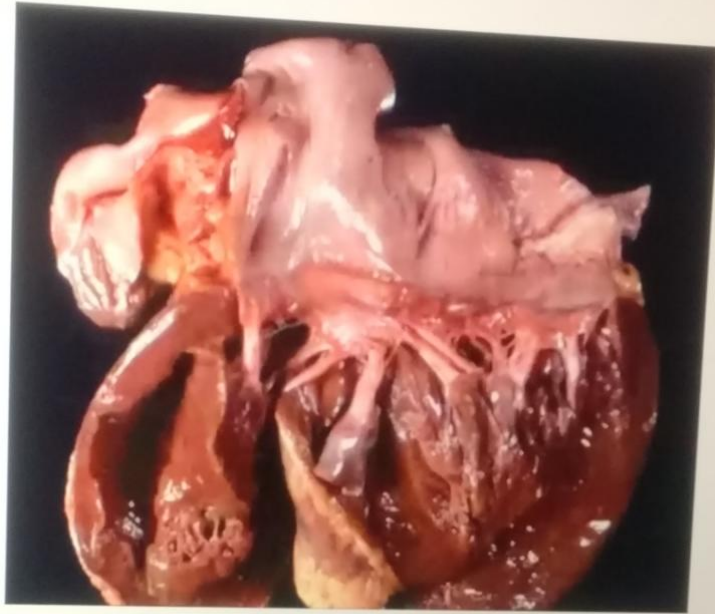
Mitral stenosis

- Takes longer duration to develop after an attack of ARF
- Fibrosis of mitral ring, commissural adhesions
- Contracture of the valve leaflets, chordae & papillary muscles
- Opening snap, low pitched, rumbling mitral diastolic murmur with pre systolic accentuation ending in loud first sound

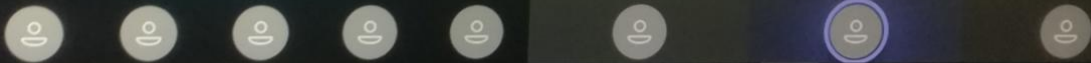
Carditis sequelae (chronic) -

Aortic insufficiency

- Sclerosis of aortic valve- distortion & retraction of the cup
- Characteristic cardiac murmur, early diastolic
- An apical pre systolic murmur (Austin flint)



Rheumatic
heart disease.
Abnormal
mitral valve.
Thick, fused
chordae



Erythema Marginatum

- Nonpruritic serpiginous or annular erythematous rash more prominent on the trunk & inner proximal portions of the extremities.
- Rash disappears on exposure to cold & reappears after hot shower.



Erythema marginatum on the trunk, showing erythematous lesions with pale centers and rounded or serpiginous margins



Subcutaneous Nodules

- Hard, painless, painless, nonpruritic, freely mobile, .2 to 2cm in diameter.
- Found symmetrically, singly or in clusters, on extensor surfaces of both large & small joints, over the scalp or along the spine.
- Lasts for weeks.
- *Always associated with severe carditis*

Sydenham's Chorea

- Neuropsychiatric disorder
- More often in prepubertal girls (8 to 12 yrs) than in boys.
- Neurologic signs – choric movement & hypotonia
- Psychiatric signs- emotional lability, hyperactivity, separation anxiety, obsessions & compulsions

Sydenham's chorea

- 10 – 15 % of patients
- Usually a delayed and often the sole manifestation of acute rheumatic fever
- Characterized by involuntary movements, specially of the face and limbs, muscle weakness, disturbances of speech and gait, poor scholastic performance
- Milk maid grip, spooning and pronation of extended hands, wormian movements of tongue

Sydenham's chorea

- Long latent period
- Uncontrollable movements
- Facial grimacing
- In coordination
- Poor school performance
- Emotional liability
- Exacerbated by stress
- Disappearing sleep
- Rarely leads to permanent neurological sequelae

Minor manifestations

1. Arthralgia without objective changes of arthritis
2. Fever at least 102F
3. Elevated ESR& CRP
4. Prolonged PR interval

Other clinical features

- Abdominal pain
- Rapid sleeping heart rate
- Tachycardia out of proportion to fever
- Epistaxis
- Precordial pain
- Positive family history of rheumatic fever

Diagnosis

- Evidence of recent streptococcal infection can include:
 - Increased antistreptolysin O or other streptococcal antibodies (anti-DNAse B)
 - Positive throat culture for Group A beta-hemolytic streptococci
 - Positive rapid direct Group A strep test
 - Recent scarlet fever

Differential diagnosis

- Arthritis
 - Rheumtoid arthritis (JRA)
 - SLE
 - Reactive arthritis – shigella, Salmonellosis, Yersenia
 - Lyme's disease
- Carditis-
 - viral myocarditis, & Pericarditis
 - Infective endocarditis
 - Congenital heart lesions
- Chorea -
 - Huntington chorea
 - Wilson disease
 - Tics

Differential Diagnosis

- Systemic lupus erythematosus - presence of antinuclear antibodies.
- Gonococcal arthritis
- leukemia
- Serum sickness
- Sickle cell disease
- reactive arthritis related to gastrointestinal infections (e.g., *Shigella*, *Salmonella*)

Clinical course

- ❑ Only carditis cause permanent cardiac damage.
- ❑ S/O mild carditis disappear in weeks.
- ❑ Sever carditis may last for 2 to 6 months.
- ❑ Arthritis subside within few days to wks & dose not cause permanent damage .
- ❑ Chorea subsides in 6 to 7 months & dose not cause permanent neurologic damage.

Management

- Bed rest
- Monitor closely for evidence of carditis
- Ambulation as soon as the signs of acute inflammation have subside.
- Patients with carditis require longer periods of bed rest.

Management

▣ ANTIBIOTIC THERAPY

- ▣ 10 days of orally administered penicillin or erythromycin.
- ▣ Single intramuscular injection of benzathine penicillin .
- ▣ After this initial course of antibiotic therapy, the patient should be started on long-term antibiotic prophylaxis.

+279



Supportive therapies

- Digoxin
- Fluid and salt restriction
- Diuretics
- Oxygen.

Management of Chorea

- Physical and emotional stress should be reduced.
- Injection of benzathine penicillin for prophylaxis indicated as in other rheumatic patients.
- Anti-inflammatory drugs not needed in isolated chorea.

For severe cases-

- Phenobarbitone 15 to 30 mg every 6 to 8 hours.
- Haloperidol (0.01-0.03 mg/kg/ 24 hr divided bid PO)
- Chlorpromazine (0.5 mg/kg q 4-6 hr PO)

Prognosis

- The more severe the cardiac involvement at the time the patient is first seen, greater the incidence of residual heart disease.
- The severity of valvular involvement increases with each recurrence.
- Valvular disease resolves more frequently when prophylaxis is followed.

Prevention

Prophylaxis –

- Duration Ideally indefinitely.
- Up to 21 to 25 yrs if no evidence of valvular involvement.
- Chance of recurrence is highest in the first 5 years after the acute rheumatic fever.

Duration of treatment

Rheumatic Fever without carditis	5yrs or until 21yrs – whichever is longer
Rheumatic Fever with carditis but no valvular disease	10yrs or “well into adulthood” – whichever is longer
Rheumatic Fever with carditis and persistent valvular disease	At least 10yrs since last episode and at least until 40yrs; sometimes lifelong

Prevention

Primary prevention –

Possible with a 10 days course of penicilline therapy for streptococcal pharyngitis.

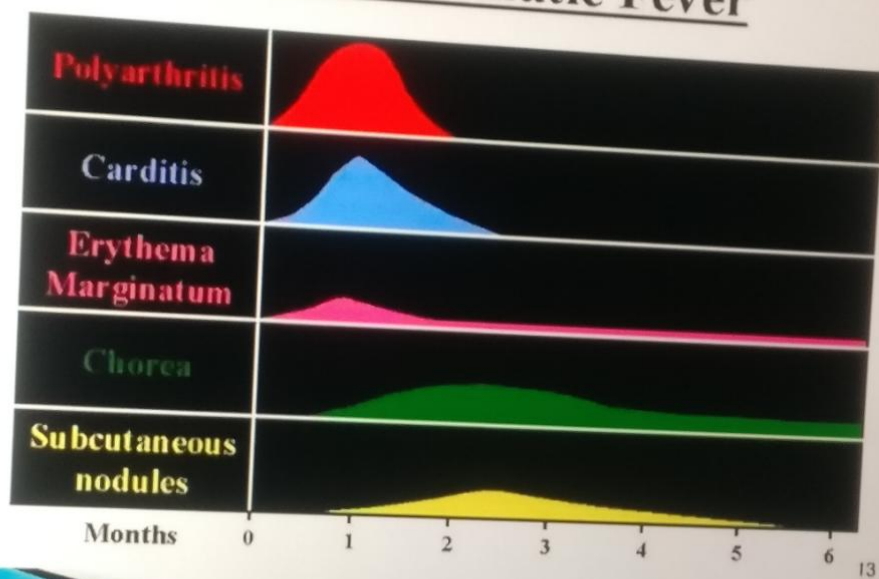
Secondary prevention-

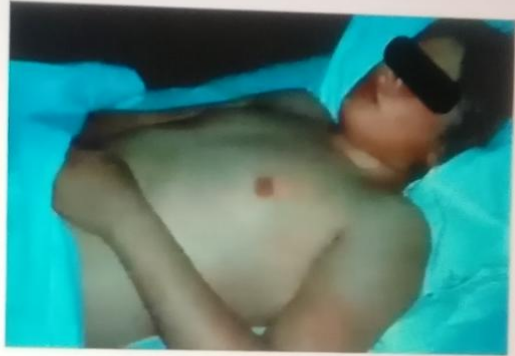
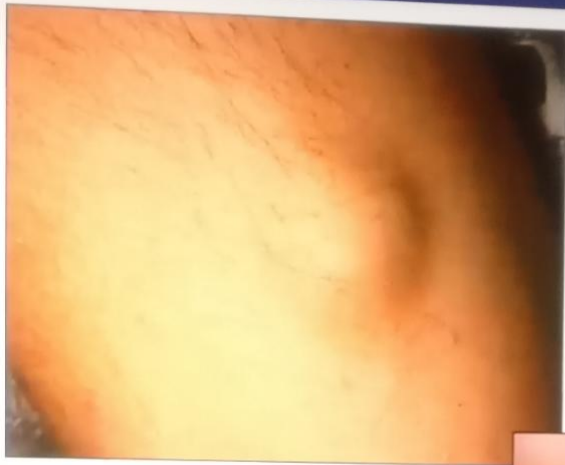
Benzathine penicilline 1.2 million units I.M. every 28 days.

Alternative-

1. Oral penicilline V 250 mg twice daily
2. Oral sulfadiazine, 1 gm once daily.
3. Oral erythromycin 250 mg twice a day.

Clinical Manifestations of Acute Rheumatic Fever





A.S.A.P. Programme for the Control of RHD in Africa:

Focus areas for action

- **Awareness raising:** public, healthcare workers
- **Surveillance:** incidence, prevalence, temporal trends
- **Advocacy:** appropriate funding of the treatment and prevention programmes
- **Prevention:** application of existing knowledge in primary & secondary prevention

ANTI-INFLAMMATORY THERAPY -

Corticosteroids-

- Patients with carditis & cardiomegaly or CCF
- ▢ Prednisone - 2 mg/kg/24 hr in 4 divided doses for 2-3 wk.
- ▢ Followed by a tapering of the dose.
- ▢ At the beginning of the tapering of the prednisone dose, aspirin should be started at 75 mg/kg/24 hr in 4 divided doses for 6 wk.
- ▢ During the full course of anti-inflammatory therapy, antacids are added to overcome irritation to gastric mucosa.

Steroids are always slowly tapered off

Bcz body has stopped producing steroids and if they are suddenly withdrawn

Body will be deficient in steroids

ANTI-INFLAMMATORY THERAPY

Oral salicylates –

Patients with typical migratory polyarthritis and those with carditis without cardiomegaly or CCF.

- Aspirin - 100 mg/kg/24 hr divided qid PO for 3-5 days.
 - followed by 75 mg/kg/24 hr divided qid PO for 4 wk.
 - Salicylate toxicity - tinnitus, hyperventilation.

High dose aspirin has side effects

If aspirin and steroid therapy doesn't treat arthritis

Then the diagnosis of rheumatic disease is probably incorrect