Rheumatic Fever

Introduction

- Reumatic fever is an Autoimmune ,inflammatory , systemic disease of childhood.
- Develop as a complication of a streptococcus infection (group A beta hemolytic streptococcal infection).
- 2 to 3 weeks after the group a streptococcus infection that cause inflammatory lesions in connective tissues primary involves heart,blood vessels,joints, subcut.tissue and CNS. (multisystem inflammatory disease)

Epidemiology

- Ages 5-15 yrs are most susceptible
- Rare <3 yrs
- Girls>boys
- Common in 3rd world countries
- Environmental factors-- *over crowding, poor sanitation, poverty,*
- *Incidence more during RAINfall ,winter & early spring*

Pathogenesis

- This is no direct invasion to the tissue by the micro-organism but its an auto immune disease that involves antigen antibody interaction.

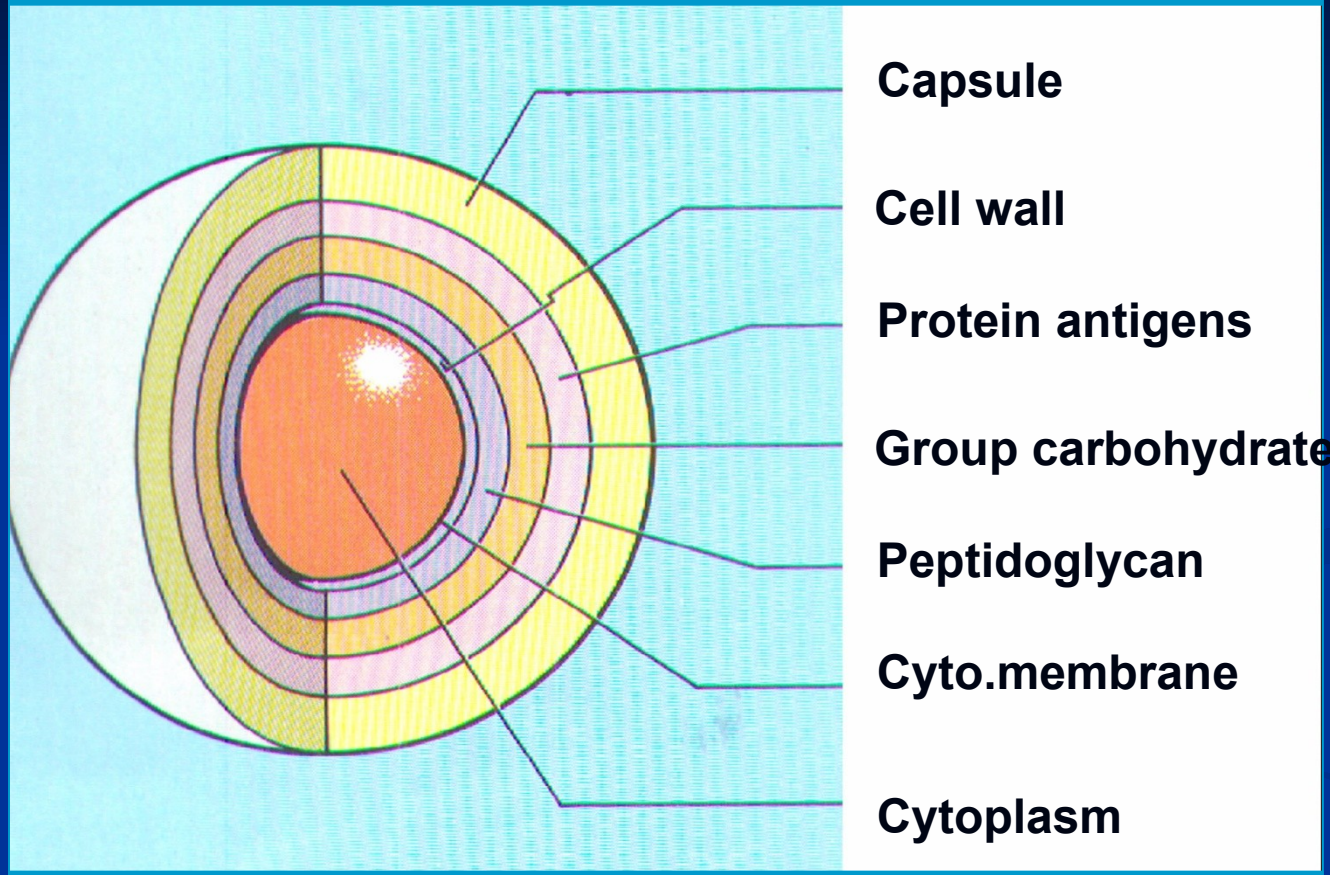
- Delayed immune response to pharyngeal infection with group A beta hemolytic streptococci.

- Body produced antibodies against streptococci.

- These antibodies cross react with Human tissue because of antigenic similarity b/w streptococcus components & human connective tissue (Molecular similarity).

- **Immunologically mediated inflammation & damage to human tissues which have antigenic similarity with streptococcal components like – Heart , Joints , Brain , Skin ,Subcutaneous tissue.**

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, sub cutaneous tissue,tendons, joints & basal ganglia of brain

Symptoms

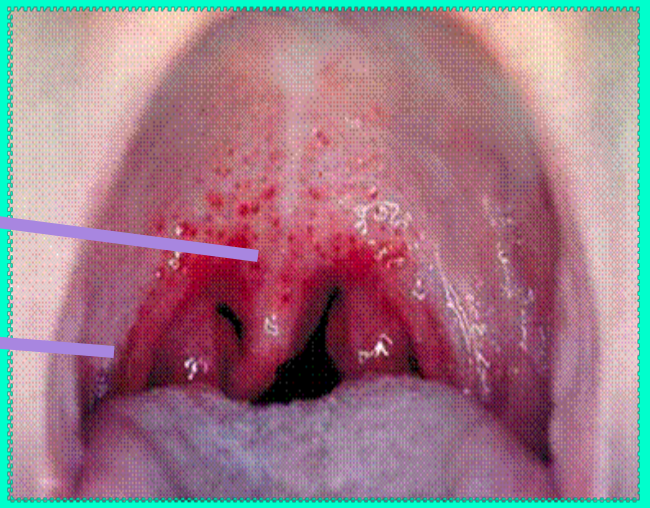
- Usually begins 1 to 6 weeks after a streptococcus ionfection .
- In heart (Corditis)
- In joints (accne arthritis)
- In brain (sydenhum chorea)
- In skin (Erythema marginatum)
- Subcutaneous (subcutaneous nodules)

ACUTE RHEUMATIC FEVER

Redness & swelling of throat & tonsils;

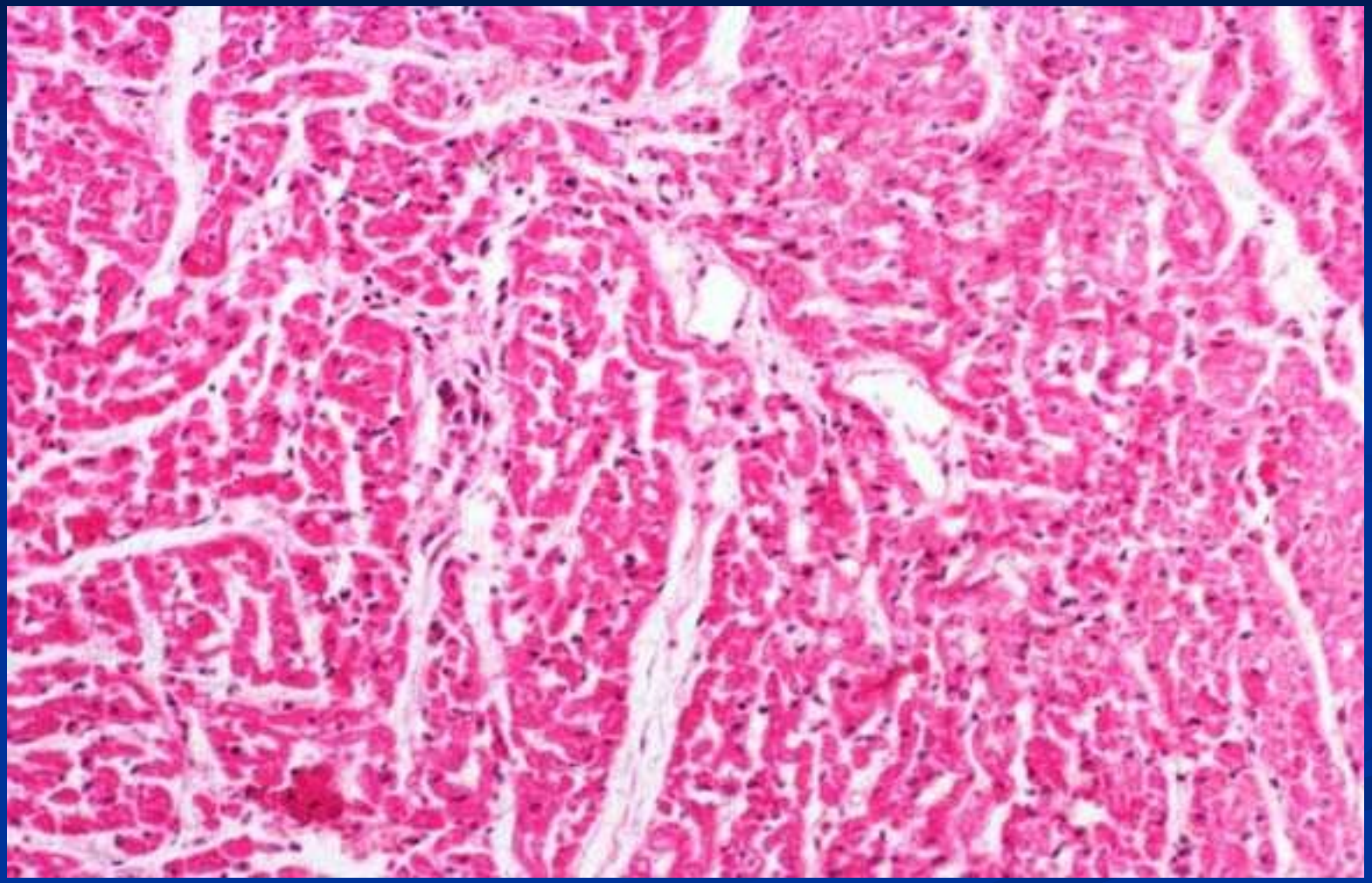
Beefy, swollen, red uvula; Soft palate petechiae ("doughnut lesions")

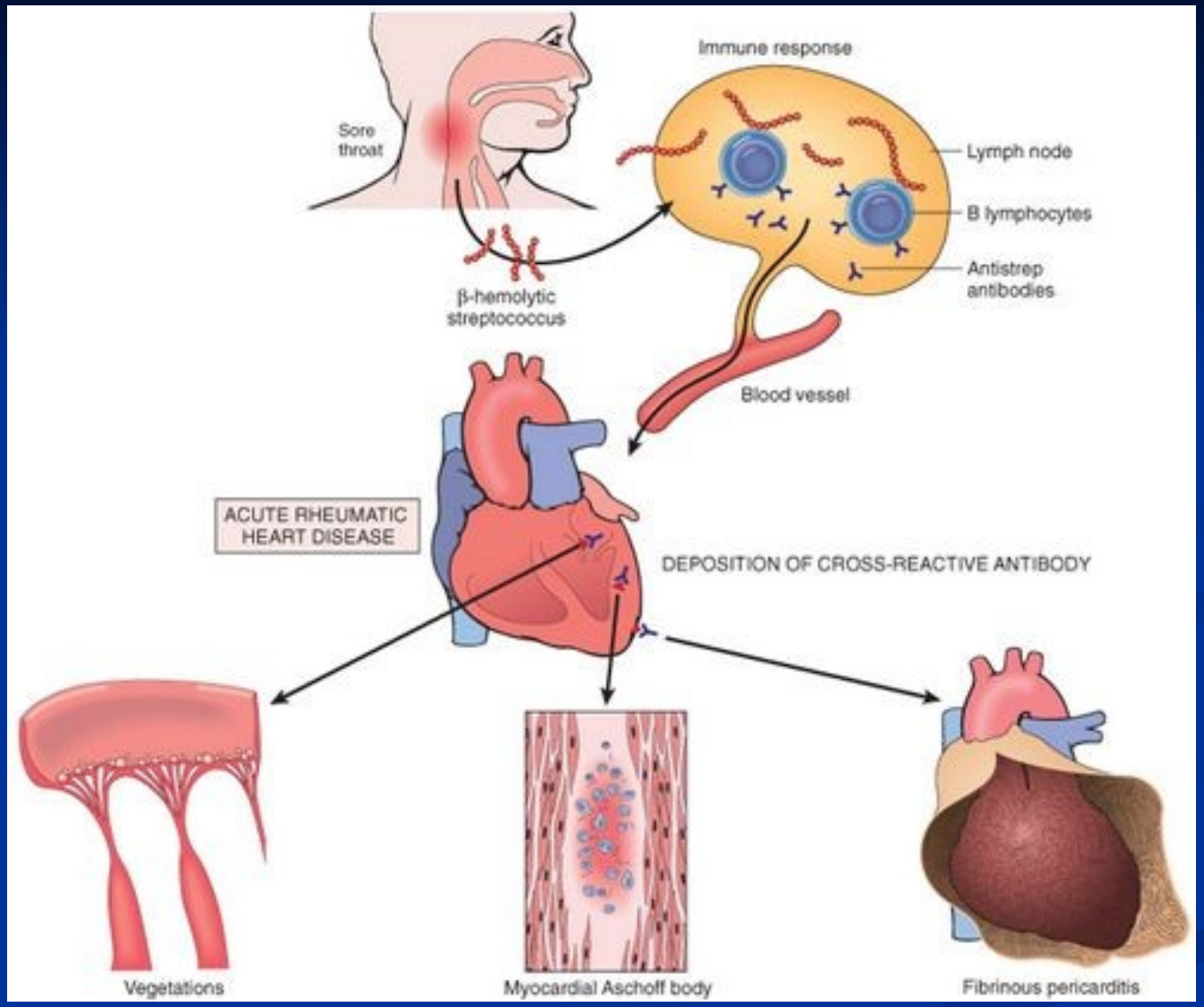
Tonsillopharyngeal erythema & exudates



Sore throat: fever, white draining patches on the throat & swollen or tender lymph glands in the neck

Histology of Myocardium in Rheumatic Carditis (200X)



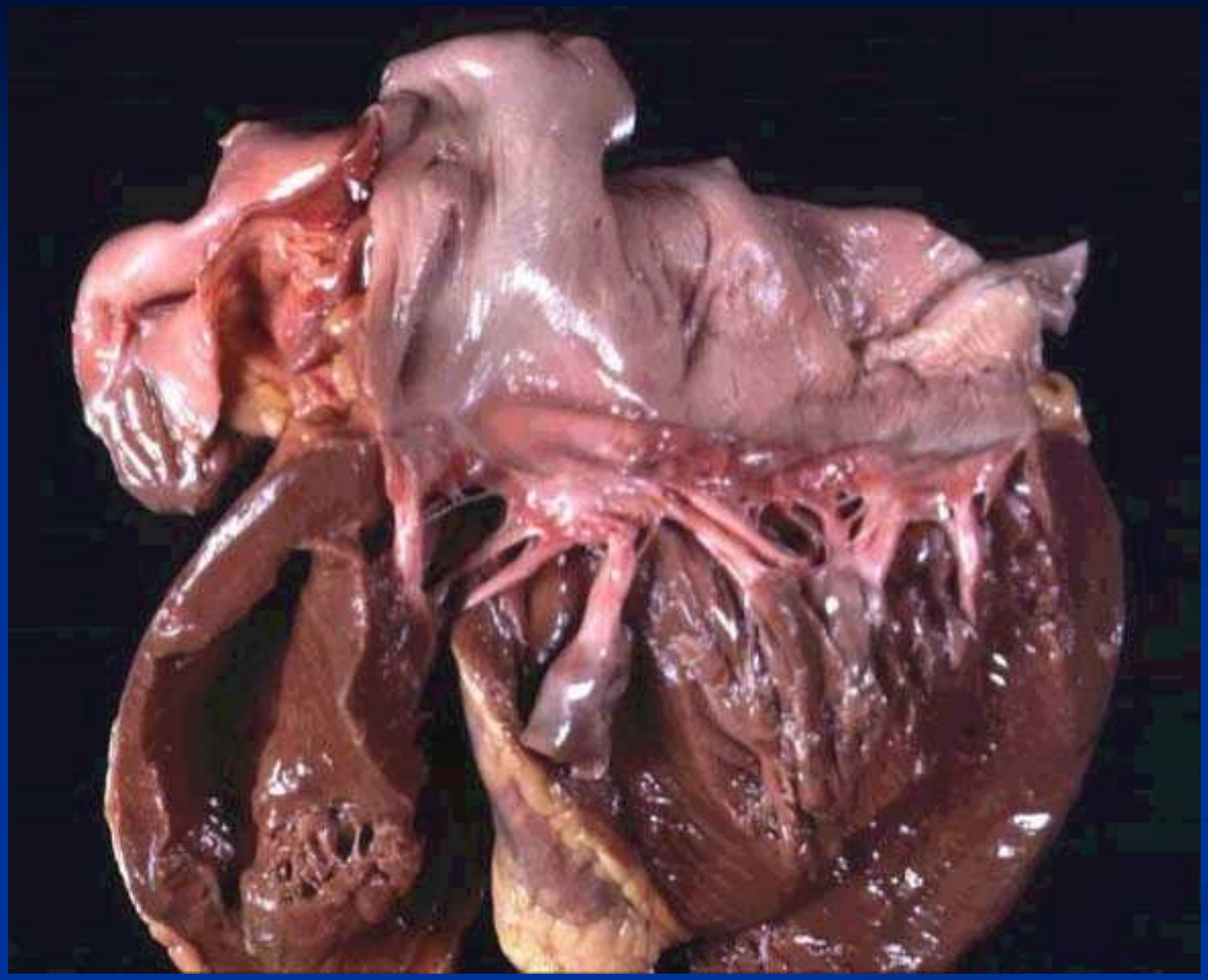

Immune response
Sore throat
Lymph node
B lymphocytes
Antistrep antibodies
β-hemolytic streptococcus
Blood vessel
ACUTE RHEUMATIC HEART DISEASE
DEPOSITION OF CROSS-REACTIVE ANTIBODY
Vegetations
Myocardial Aschoff body
Fibrinous pericarditis

1. ACCNE 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*

2.Carditis

- Manifest as **pancarditis**(endocarditis, myocarditis and pericarditis),occur in 40-50% of cases
- *Carditis is the only manifestation of rheumatic fever that leaves a sequelae & permanent damage to the organ*
- *Valvulitis occur in acute phase*
- *Chronic phase- fibrosis,calcification & stenosis of heart valves(fishmouth valves)*



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

3.Sydenham Chorea

- Occur in 5-10% of cases
- Mainly in girls of 1-15 yrs age
- May appear even 6/12 after the attack of rheumatic fever
- Clinically manifest as-clumsiness, deterioration of hand writing,emotional instability , grimacing movements of face .
- Clinical signs- pronator sign, jack in the box sign , milking sign of hands

4.Erythema Marginatum

- Occur in <5%.
- Unique,transient,serpiginous-looking lesions of 1-2 inches in size
- Pale center with red irregular margin
- More on trunks & limbs & non-itchy
- Worsens with application of heat
- *Often associated with chronic carditis*

ERYTHEMA MARGINATUM

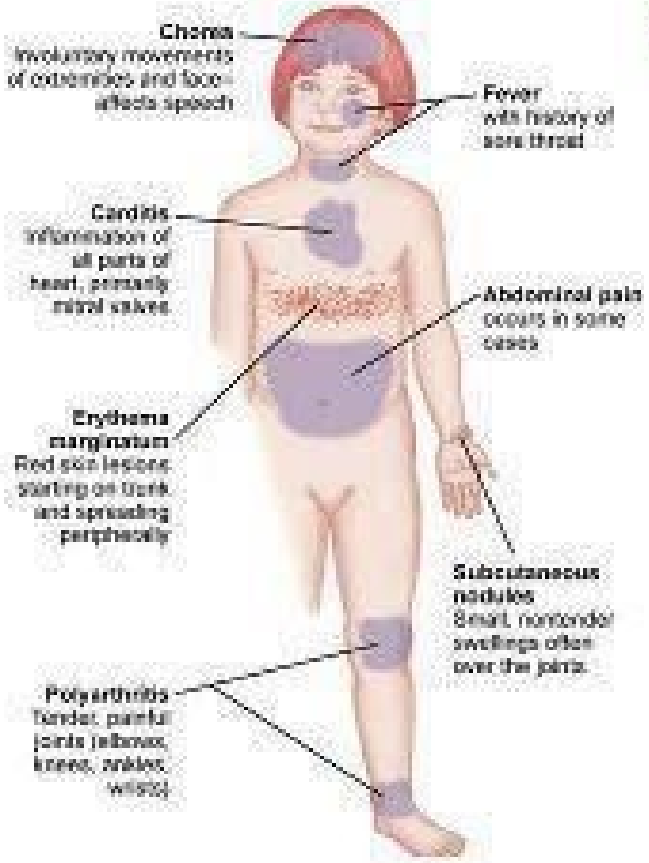
5.Subcutaneous nodules

- Occur in 10%
- Painless,pea-sized,palpable nodules
- Mainly over extensor surfaces of joints,spine,scapulae & scalp
- Associated with strong seropositivity
- *Always associated with severe carditis*

SUBCUTANEOUS NODULES

Rheumatic Fever - Assessment

- **Major**
  - **Carditis**
  - **Polyarthritis**
  - **Chorea**
  - **Erythema marginatum**
  - **Subcutaneous nodules**



- **Minor**
  - **Arthralgia**
  - **Fever**
  - **Laboratory Findings:**
    - ↑ **Erythrocyte sedimentation rate**
    - ↑ **C-reactive protein**
    - **Prolonged PR interval**

Jones Criteria

Other features (Minor features)

- Fever-(upto 101 degree F)
- Arthralgia
- Pallor
- Anorexia
- Loss of weight

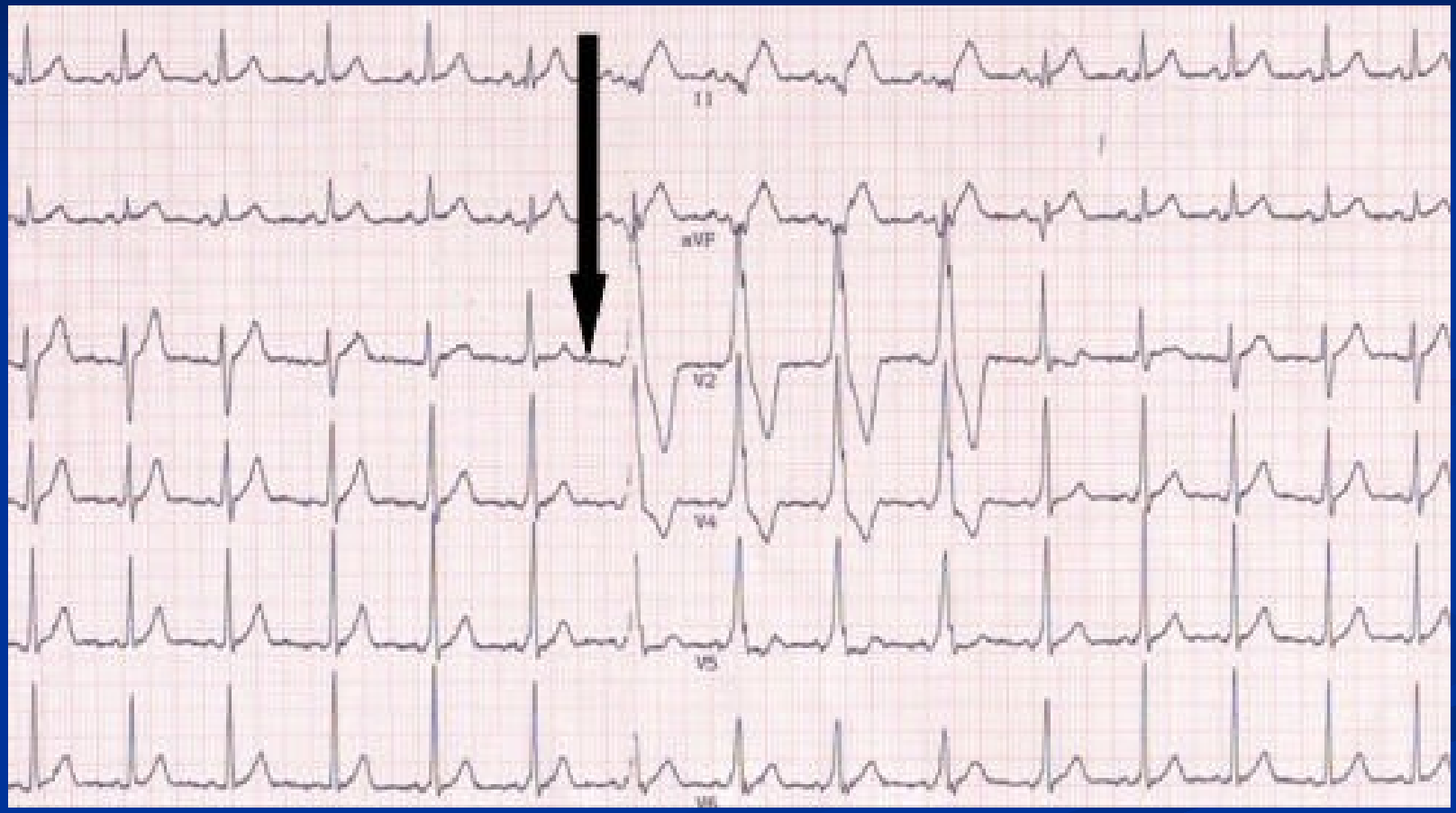
Laboratory Findings

- High ESR
- Anemia, leucocytosis
- Elevated C-reactive protien
- ASO titre >200 Todd units.
  (Peak value attained at 3 weeks,then comes down to normal by 6 weeks)
- Anti-DNAse B test
- Throat culture-GABHstreptococci

Laboratory Findings (Contd)

- ECG- prolonged PR interval, 2nd or 3rd degree blocks,ST depression, T inversion
- 2D Echo cardiography- valve edema,mitral regurgitation, LA & LV dilatation,pericardial effusion,decreased contractility

primarily prolonged PR interval



Cardiomegaly

Diagnosis

- Rheumatic fever is mainly a clinical diagnosis
- *No single diagnostic sign or specific laboratory test available for diagnosis*
- Diagnosis based on ***MODIFIED JONES CRITERIA***

Differential Diagnosis

- Juvenile rheumatiod arthritis
- Septic arthritis
- Sickle-cell arthropathy

- Myocarditis

- Leukemia

Thank You