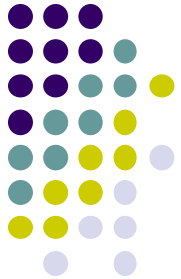
Rheumatic Fever & Rheumatic Heart Disease

Dr. Gauri Metkar
Professor, Pathology

Rheumatic Fever & Rheumatic Heart Disease

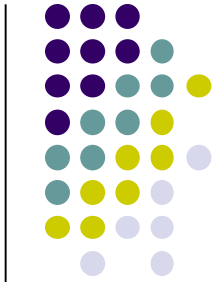


- **Definition: It is an acute immunologically mediated multi-systemic inflammatory disease that occurs a few weeks following an episode of group A Streptococcal pharyngitis.**

- Heart, Joints, CNS, Skin and Subcutaneous tissue affected

- Acute stage: Rheumatic Fever
- Chronic stage: Rheumatic Heart Disease

Important Consequence of Rheumatic Fever



- Chronic valvular deformities
- Permanent dysfunction & many years later – severe, fatal cardiac problems
- Rarely, follows infections by streptococci at other sites such as skin

Incidence:

- Most commonly in children 5-15 yrs
- Slight female preponderance
- Poor socioeconomic strata of society

Aetiopathogenesis:

Acute rheumatic fever

↓

Hyper sensitivity reaction

↓

Induced by group A streptococci β – haemolytic

↓

Antibodies directed against M protein of certain strains of streptococci

→ **<u>cross reaction</u>** with glycoprotein Antigen in heart, joints & other tissues

- Onset of symptoms 2 – 3 weeks after infection, absence of streptococci from lesions - Support concept – Immune response against bacteria.
- Autoimmune – nature of cross reacting antibodies difficult to define
- Only minority of infected patients develop rheumatic fever – Genetic susceptibility
- Chronic sequelae – Due to progressive fibrosis

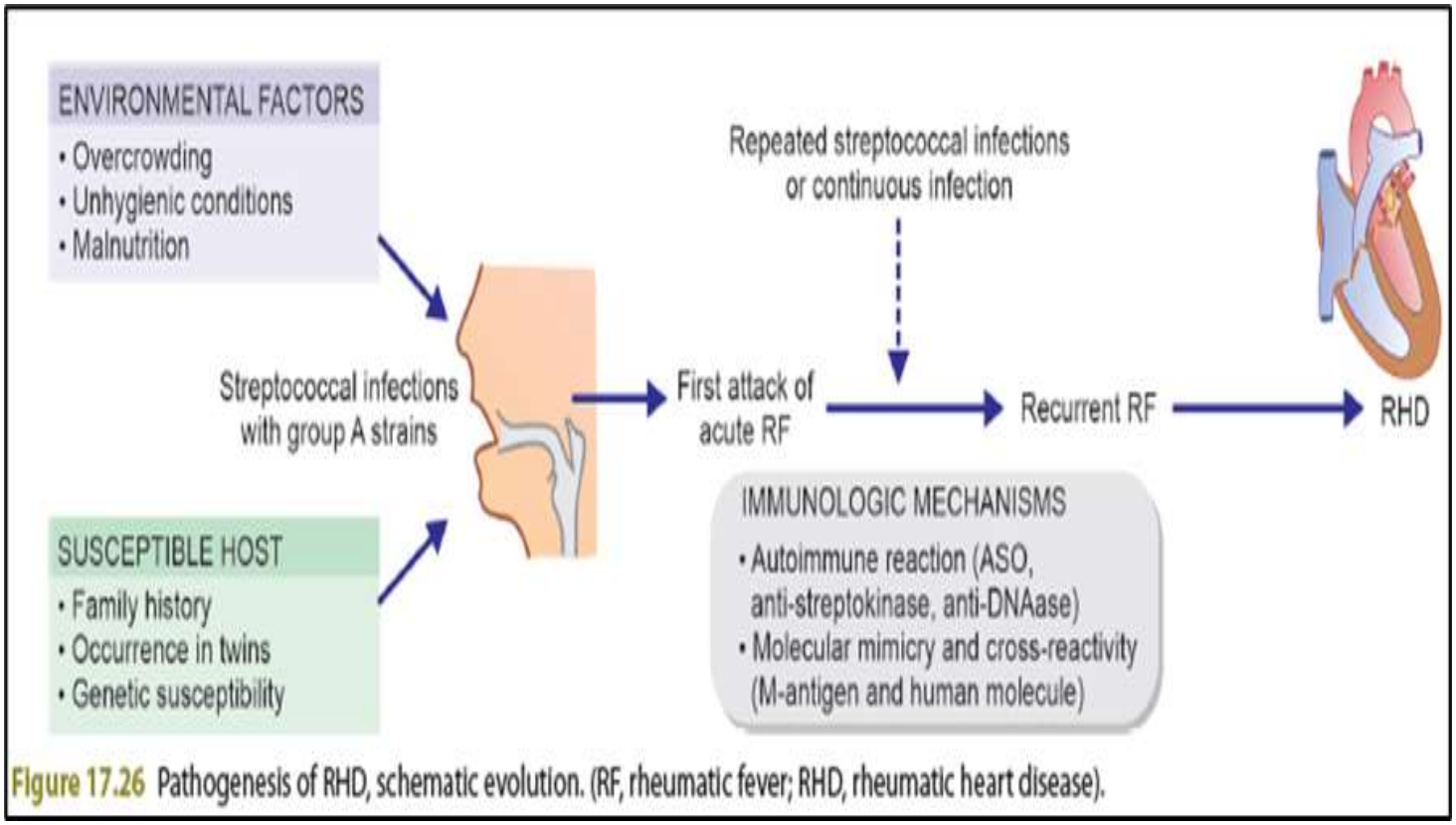


Figure 17.26 Pathogenesis of RHD, schematic evolution. (RF, rheumatic fever; RHD, rheumatic heart disease).

Aschoff nodules / Aschoff bodies

- Spheroidal or fusiform tiny structures
- 1-2 mm in size
- Occurring in interstitium of heart
- Found in vicinity of small blood vessels
- In myocardium, endocardium & pericardium
- Extra cardiac tissues.

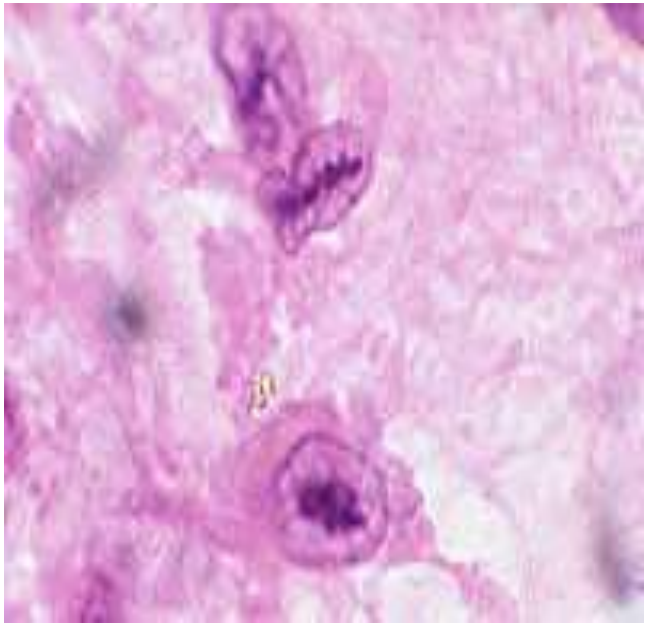
Evolution of Aschoff body:

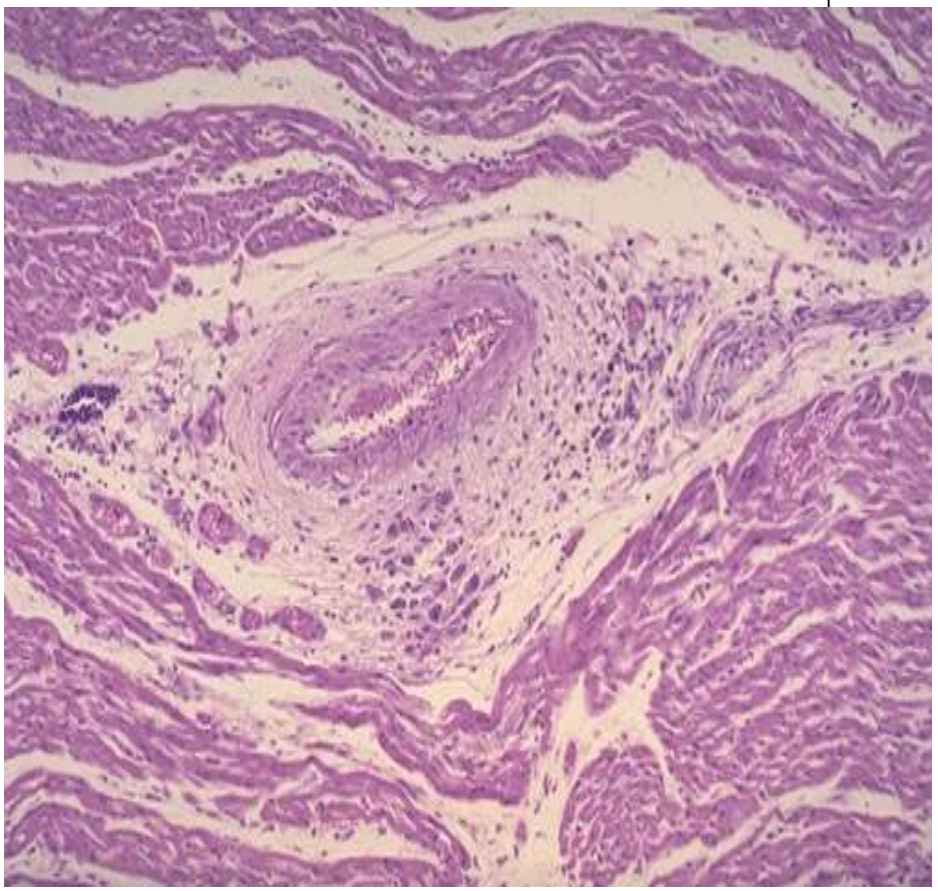
1. **Early (exudative or degenerative) Stage**
   - 4^{th} week of illness
   - Edema and increase in acid mucopolysaccharide in ground substance
   - Separation of collagen fibers
   - Collagen fibers fragmented and disintegrated
   - Focus takes appearance and staining characteristic of fibrin
   - Fibrinoid degeneration

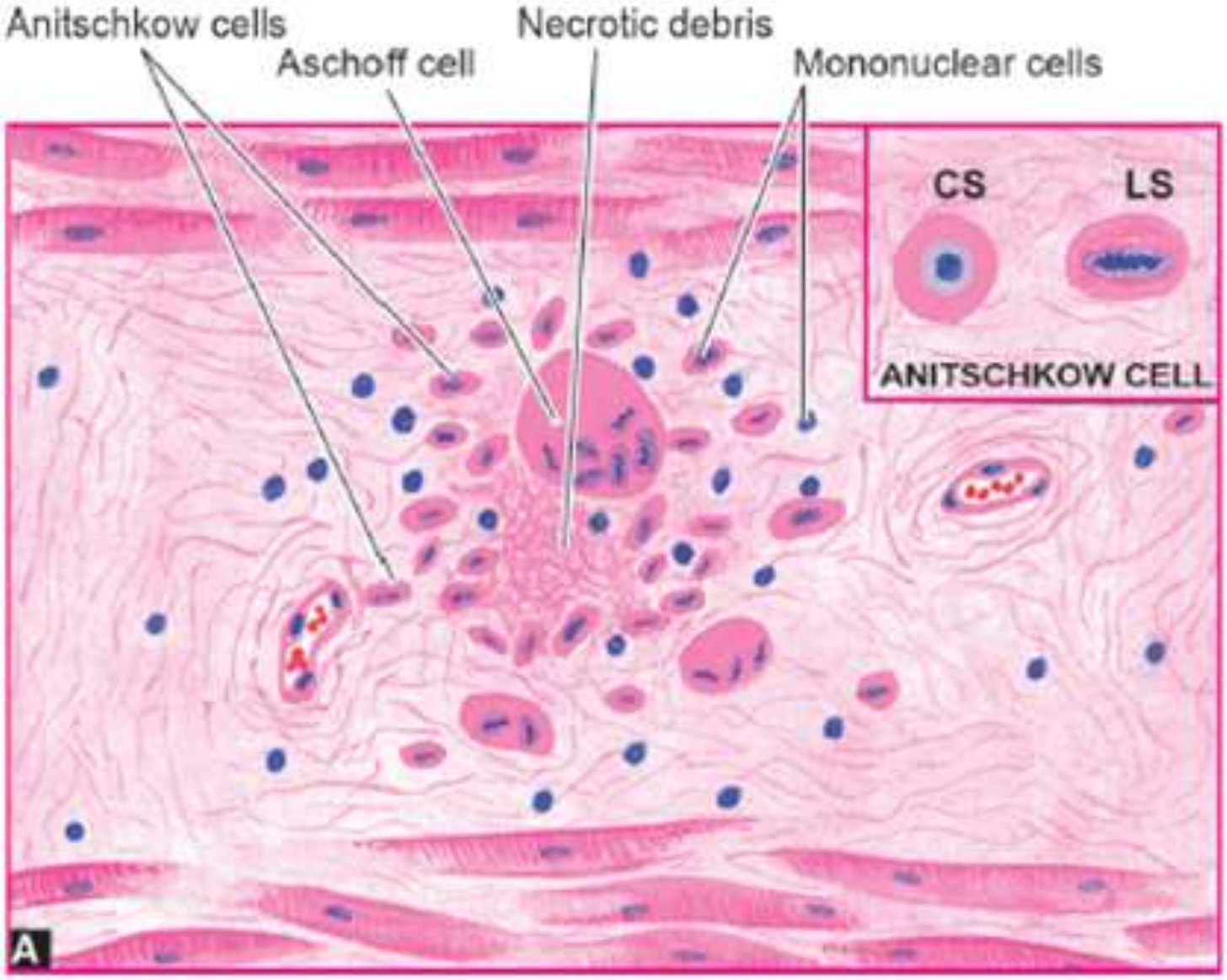
2. Intermediate (proliferative or granulomatous) Stage

- 4^{th} – 13^{th} week of illness
- Fibrinoid change - followed by proliferation of cells
- Lymphocytes, plasma cells, few neutrophils and characteristic cardiac histiocytes (Anitschkow Cells) at margin of lesion

- **Anitschkow Cells:**
  - Present in small numbers in the normal heart but number increases in an Aschoff body.
  - Large mononuclear cells having central round nuclei and contain moderate amount of amphophilic cytoplasm
    - Nucleus – vesicular
    - Chromatin – slender wavy – Caterpillar Cells
  - Some larger macrophages become multinucleated to form Aschoff giant cells






Anitschkow cells
Aschoff cell
Necrotic debris
Mononuclear cells
CS
LS
ANITSCHKOW CELL
A

3. Late (Healing or fibrous) stage:

- 12 – 16 weeks
- Anitschkow cells become spindle shape arrange in palisaded manner
- Aschoff bodies – less cellular and collagenous tissue increased.

Acute Rheumatic Fever → diffuse inflammation

Aschoff's bodies

Pericardium

Myocardium

Endocardium

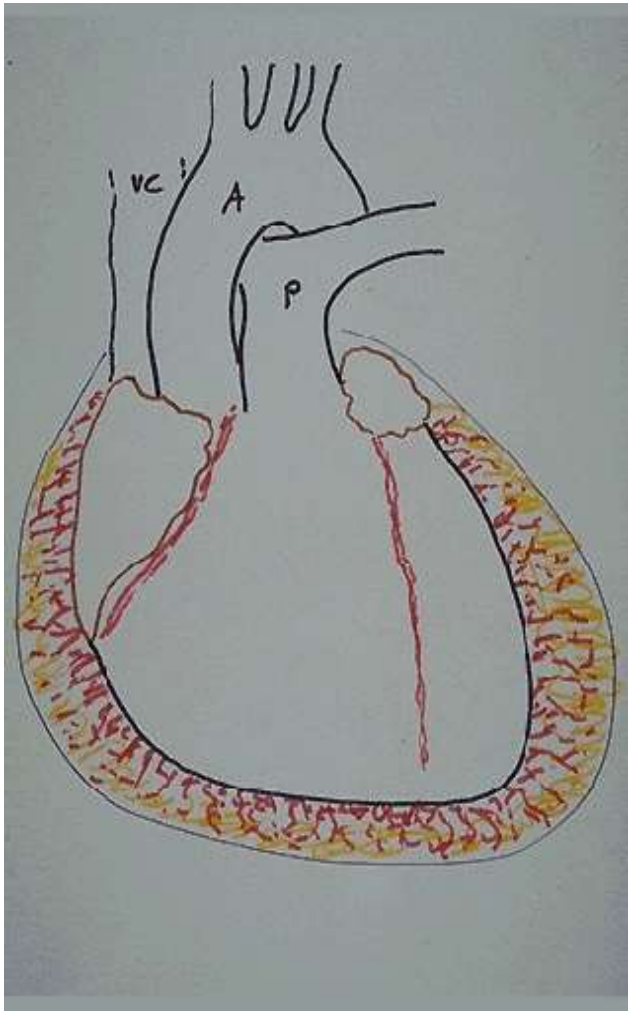
Acute Rheumatic Fever-PANCARDITIS

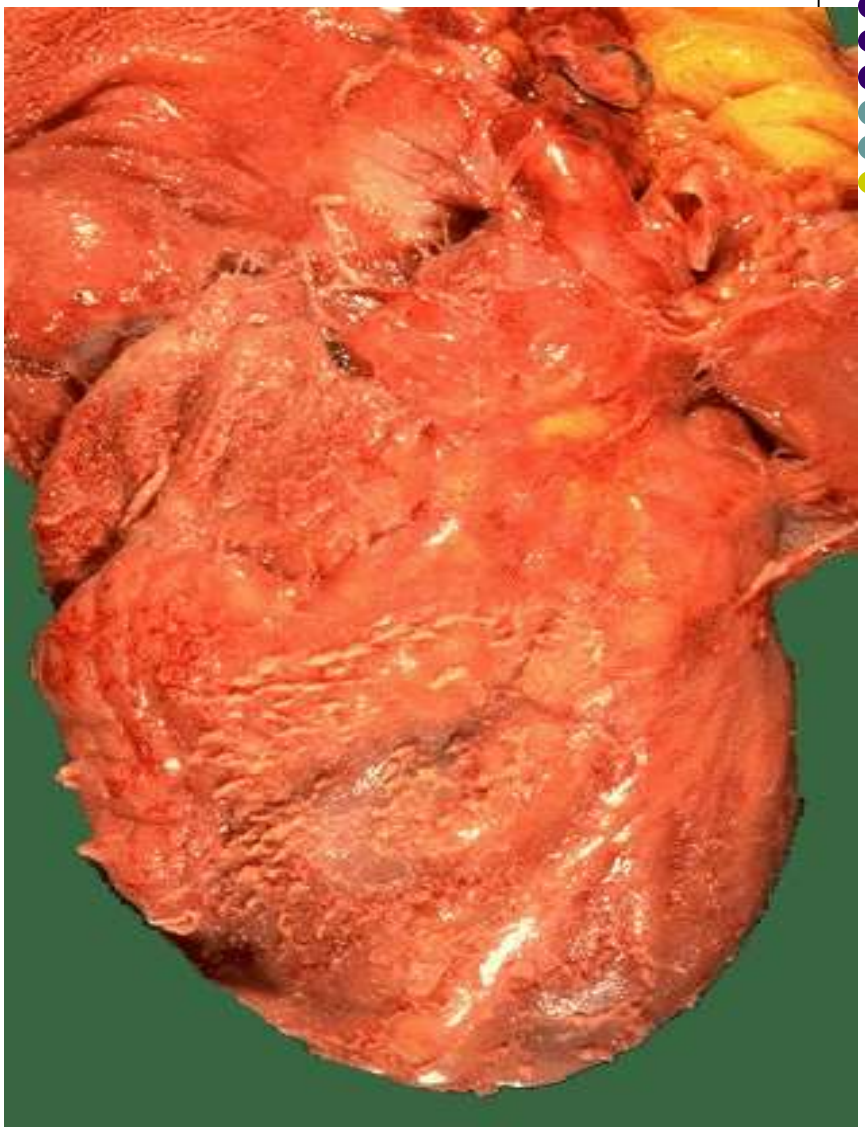
- **Pericardium** – inflammation accompanied by serofibrinous pericardial exudate
  - **Bread & Butter Pericarditis**
  - Generally resolves without sequelae,
  - if fails to resolve - chronic adhesive pericarditis.

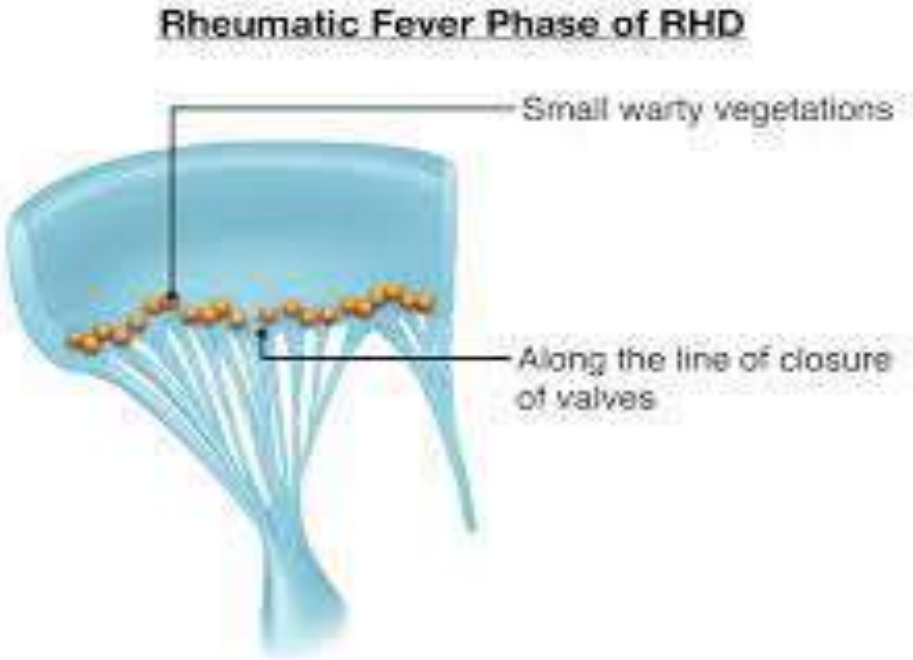
- **Myocardium** – Aschoff bodies
  - Interstitial connective tissue, often perivascular

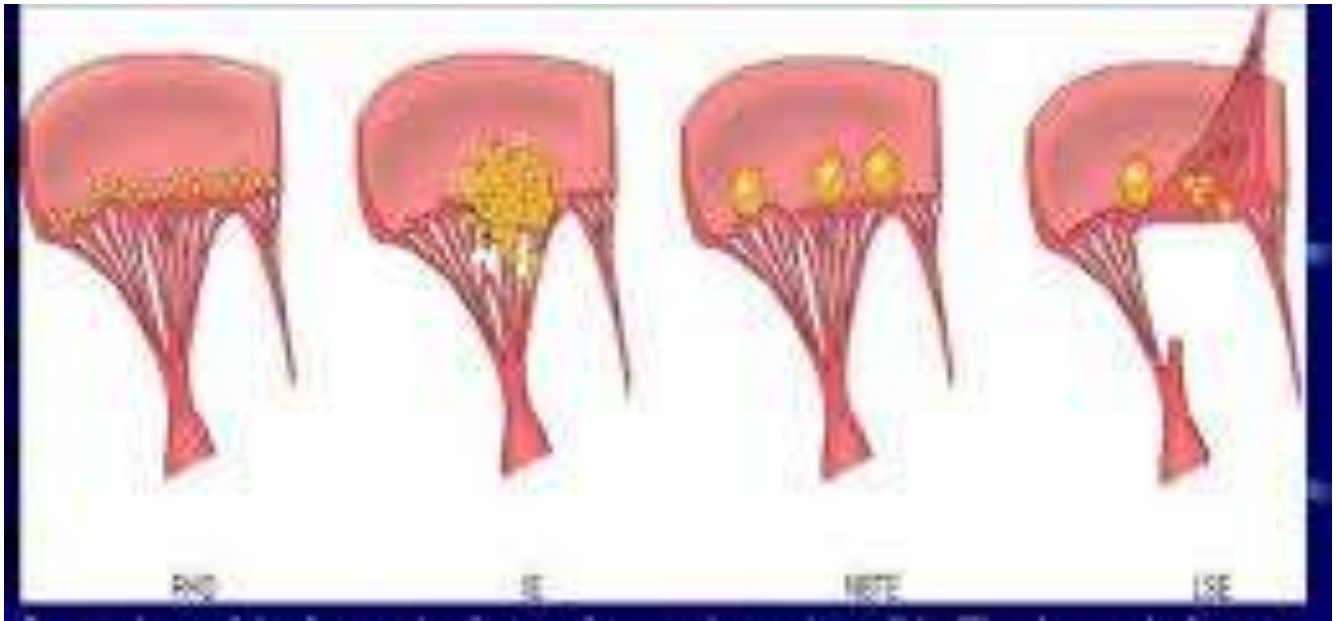
- **Endocardium –**
  - ***Valvulitis***: left sided valves, 1 – 2 mm vegetations – verrucae, along lines of closure,cusps and tendinous cords
  - Fibrinoid necrosis + inflammation

    Erosion ⟶ precipitation of fibrin


VC
A
P




Rheumatic Fever Phase of RHD
Small warty vegetations
Along the line of closure of valves


RHD
IE
NBTE
LSE

- **Mural Endocardium**

Regurgitant jets

↓

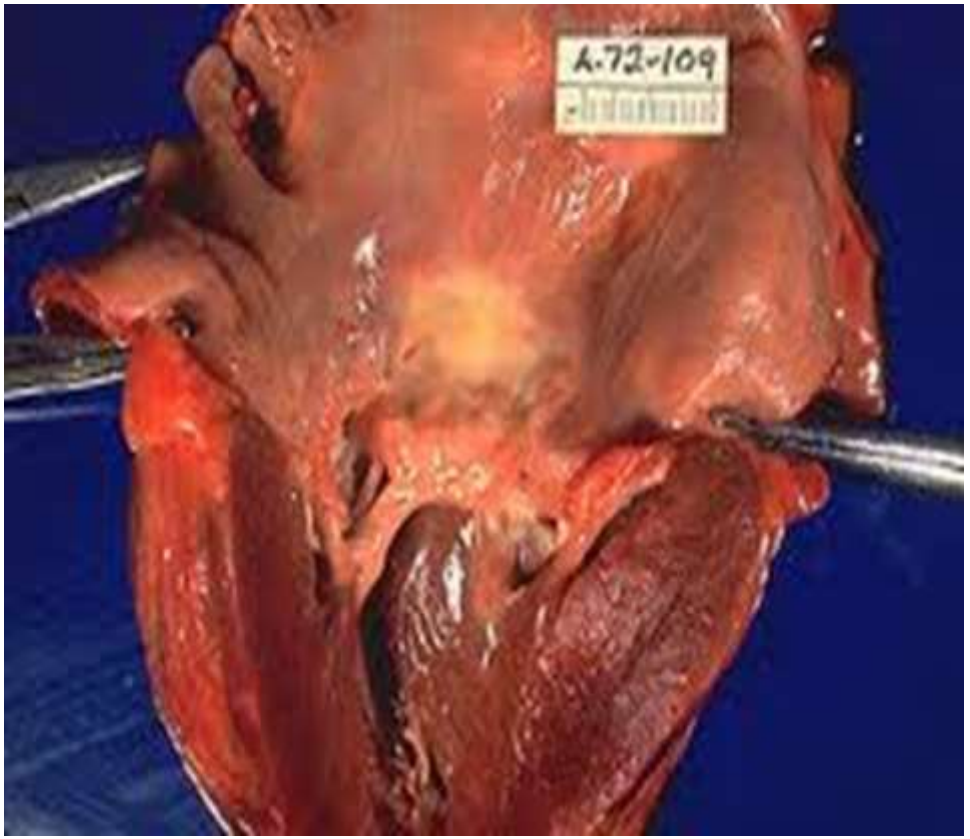
Irregular thickenings & roughened Map like areas

↓

LA

↓

Mac Callum plaques.



Chronic Rheumatic Heart Disease

Organization of acute inflammation and subsequent fibrosis

Valves:

- **Permanent deformity of one or more valves.**
- **Mainly mitral; followed by mitral and aortic valve.**

Gross:

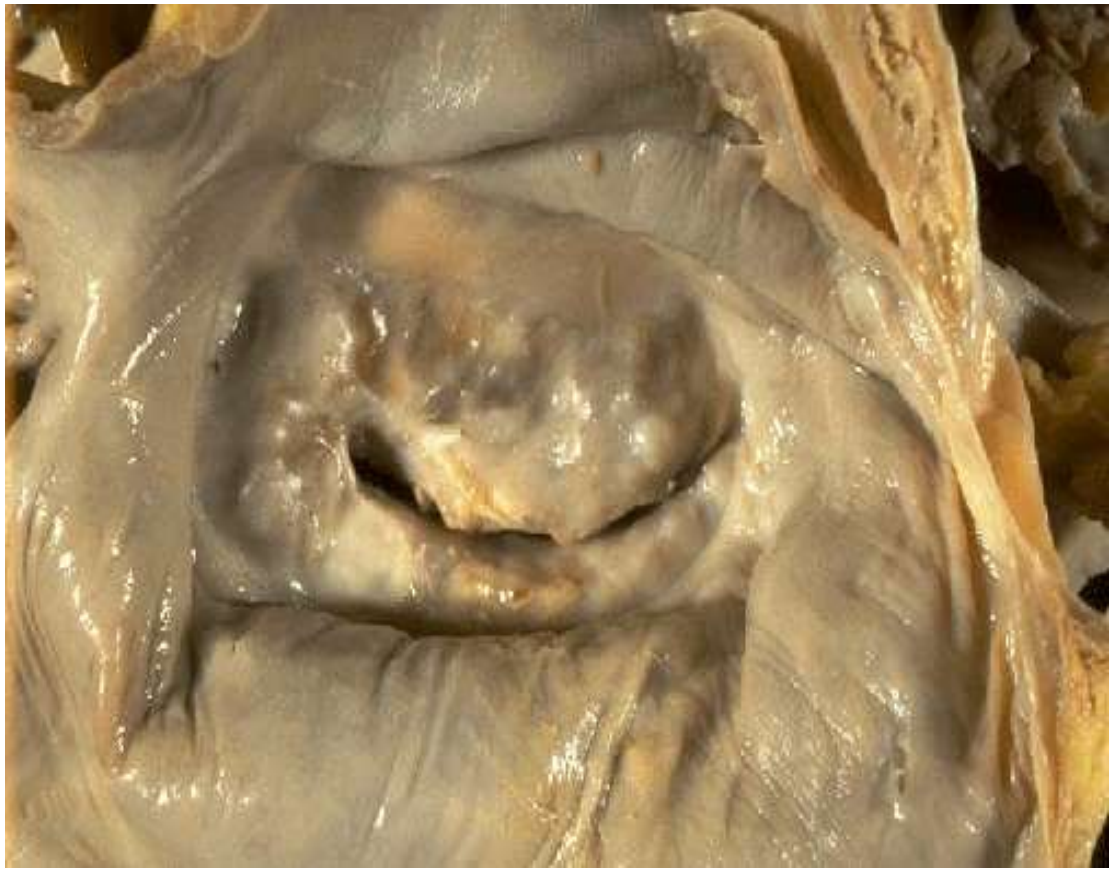
- **Leaflet thickening**
- **Commissural fusion & Calcification,**
- **Shortening, thickening and fusion of tendinous cords – FISH MOUTH/ BUTTON HOLE STENOSIS**

M/E: Diffuse fibrosis & neovascularization
Fibrin & superimposed platelet thrombi

No Bacteria

Mitral Stenosis ___LA dilatation_____

___Mural thrombus _____long standing congestive changes in lungs

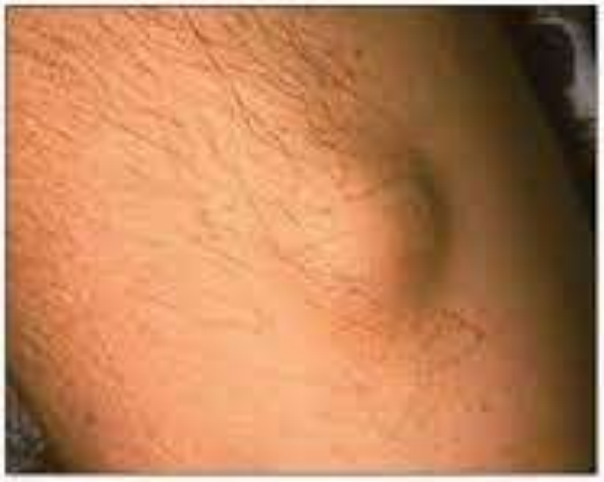


- **Clinical Features*: Rheumatic Fever***

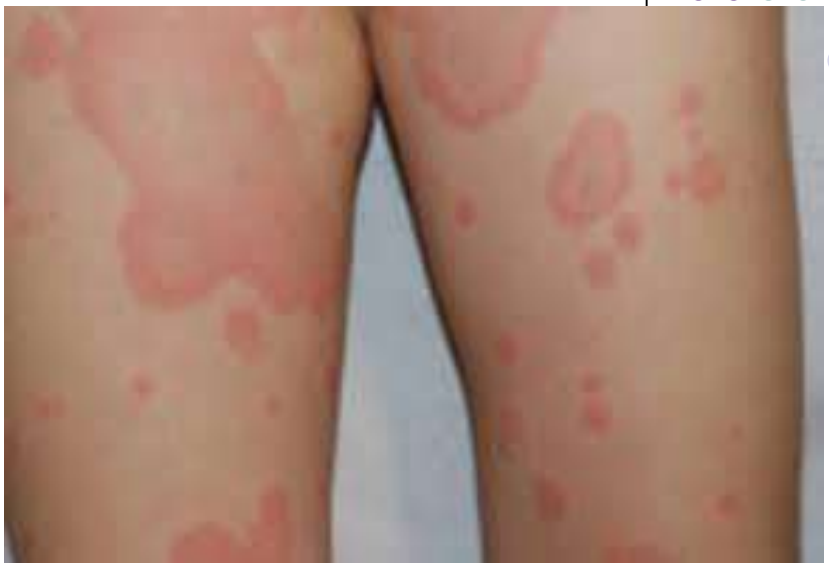
Major Criteria:

1) Migratory poly arthritis of large joints.
2) Carditis
3) Subcutaneous nodules – extensor surfaces wrist, elbow ankle & knee
4) Erythema marginatum of skin - trunk, proximal part of extremities
5) Syndenham Chorea – involuntary, purposeless, rapid movements

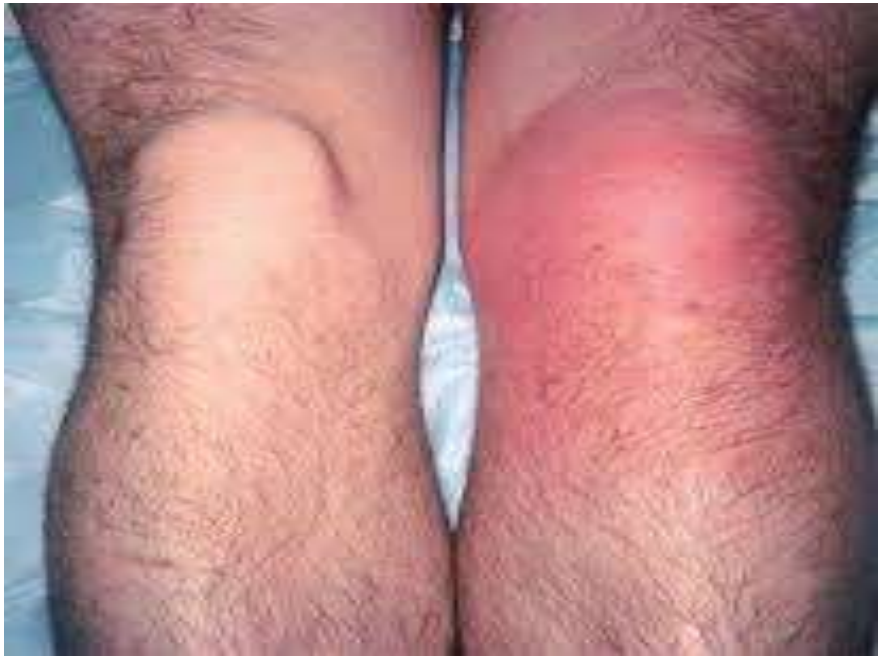
Subcutaneous Nodule



Erythema Marginatum



Polyarthritis



- **Minor Criteria:**

1) Fever
2) Arthralgia
3) Elevated blood levels of acute phase reactants (↑ESR,↑CRP, leucocytosis)
4) Previous history of RF
5) Prolonged PR interval on ECG

- **Supportive evidence:**

Antibodies to streptococcal enzyme (ASO titre) such as streptolysin O & DNA se B are present (+ve throat culture)

2 major OR 1 major & 2 minor should be present for diagnosis

- **Acute Carditis**
  - Pericardial friction rub
  - Weak heart sounds
  - Tachycardia
  - Arrythmias
  - Myocarditis – Cardiac dilation
    - Function mitral valve insufficiency
- **Prognosis for primary attack – Good**
- **1%-fulminant RF**
- **After initial attack**
- Increase vulnerability of reactivation of disease with subsequent pharyngeal infections
- Carditis worsens with each recurrence

- **Chronic Rheumatic Heart Disese**

- Cardiac murmurs
- Cardiac hypertrophy & Dilation
- Heart failure

- A **pericardial friction rub** is a grating, to-and-fro sound produced by friction of the heart against the pericardium.
- The **diastolic murmur of mitral stenosis** is of low pitch, rumbling in character, and best heard at the apex with the patient in the left lateral position. It commences after the opening snap of the mitral valve, and the duration of the murmur correlates with the severity of the stenosis.

- **Complication:**
- **Arrythmias – AF**
- **Thromboembolic complication**
- **Infective Endocarditis**
- **Cardiac failure**
- **Sudden death**

Surgical Repair: Improved outlook