RHEUMATIC FEVER
&
Rheumatic Heart Disease

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OVERVIEW

- Etiology
- Pathophysiology
- Pathology
- Gross & Microscopy
- Criteria
- Complications

DEFINITION

An acute, immunologically mediated multisystem inflammatory disease occurring a few weeks following an episode of……

GROUP A STREPTOCOCCAL PHARYNGITIS.

Rheumatic Heart Disease

PATHOGENESIS

- Acute Rheumatic Fever is a **HYPERSENSITIVITY** reaction to **GROUP A STREPTOCOCCI. (**The exact pathogenesis is uncertain**)**
- Antibodies against **M Proteins** of certain **strains** of streptococci – cross react with glycoprotein antigen in the **Heart, Joints** and other tissues.
- Onset of symptoms is **2 – 3 weeks after infection.**
- Streptococci are usually **absent** from the lesions. probably due to an **auto immune** response against self – antigens.
- **Genetic Susceptibility** might play a role as play a minority of patients develop R.F.

Rheumatic Heart Disease

- **ONSET**
  Usually 10 days to 6 weeks following pharyngitis in 3% patients.

- **AGE GROUP**
  5 – 15 years (20% of cases seen in middle to later life)

- **CULTURE**
  From the pharynx are sterile at the time of onset of disease.

- Antibodies to streptococcal enzymes are present in the serum (eg. Antistreptolysin O/ ASO.) DNAase B.

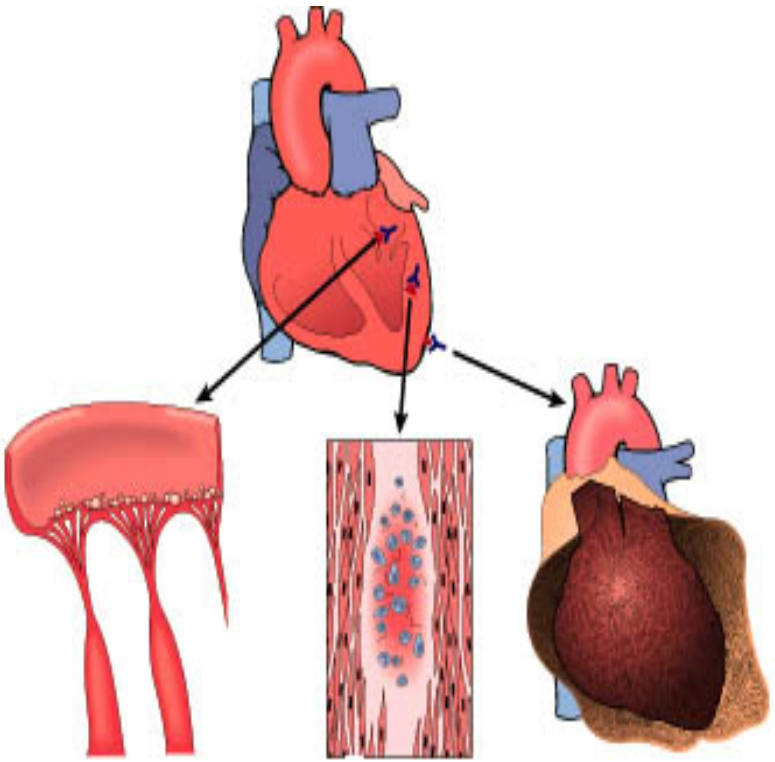
- Main clinical features are **CARDITIS & ARTHRITIS.**

Acute Rheumatic Fever

- Rheumatic fever does not follow infection by streptococci at other sites.

- Reduced incidence over the past 30 years seen due to
    - Improved S.E. conditions.
    - Rapid Diagnosis and Rx of streptococcal pharyngitis
    - ↓ Virulence of group A Streptococci (Unknown reason).

Acute rheumatic fever



- **Endocardium** - formation of vegetations (verrucae) along the line of closure . The mitral valve is commonly affected.
- **Myocardium**- myocarditis with widely disseminated focal inflammatory lesions (Aschoff bodies) scattered about in the interstitium, often perivascular
- **Pericardium** - inflammatory lesions of the epicardial connective tissues may be associated with a fibrinous or serofibrinous exudate. (usually resolves without lesions.)

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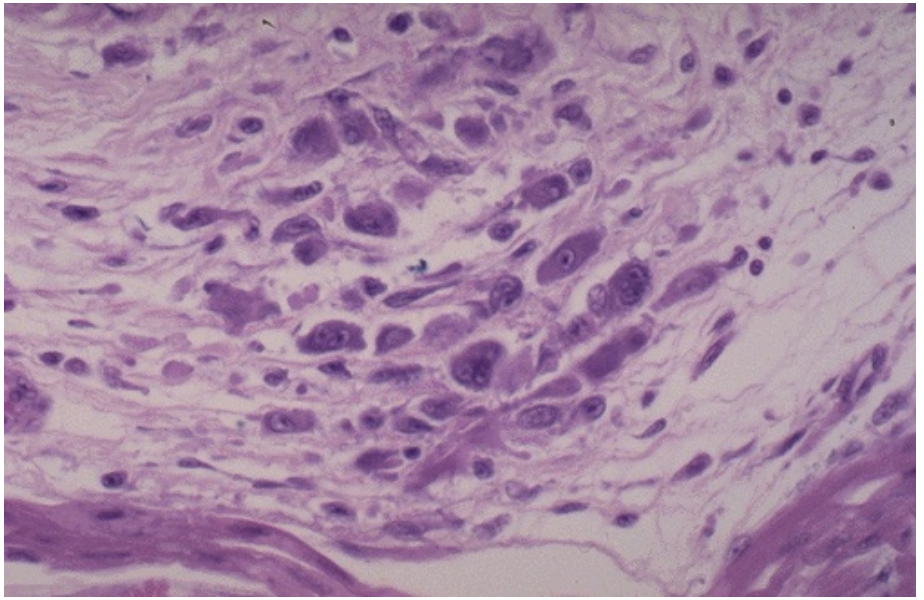
Acute Rheumatic Fever

Morphology

Focal Inflammatory lesions in various tissues.

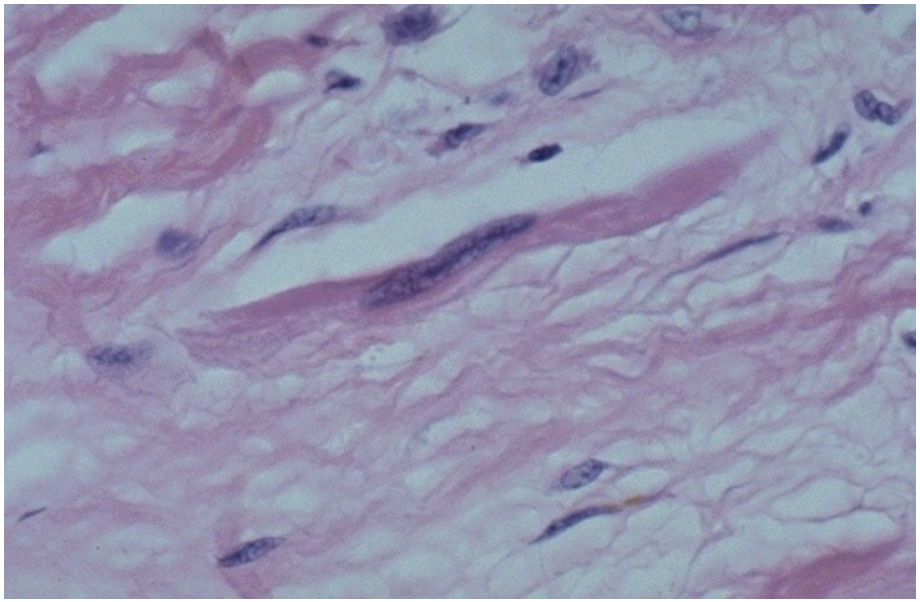


ASCHOFF BODIES

These are distinctive lesion in the heart consisting of swollen eosinophilic collagen, surrounded by lymphocytes and plasma cells.

ANTISCHKOW CELLS / (Plump macrophages)

Cells with abundant cytoplasm and central round to ovoid nucleus in which chromatin is disposed in a slender, wavy ribbon. ***(caterpillar cells.)***

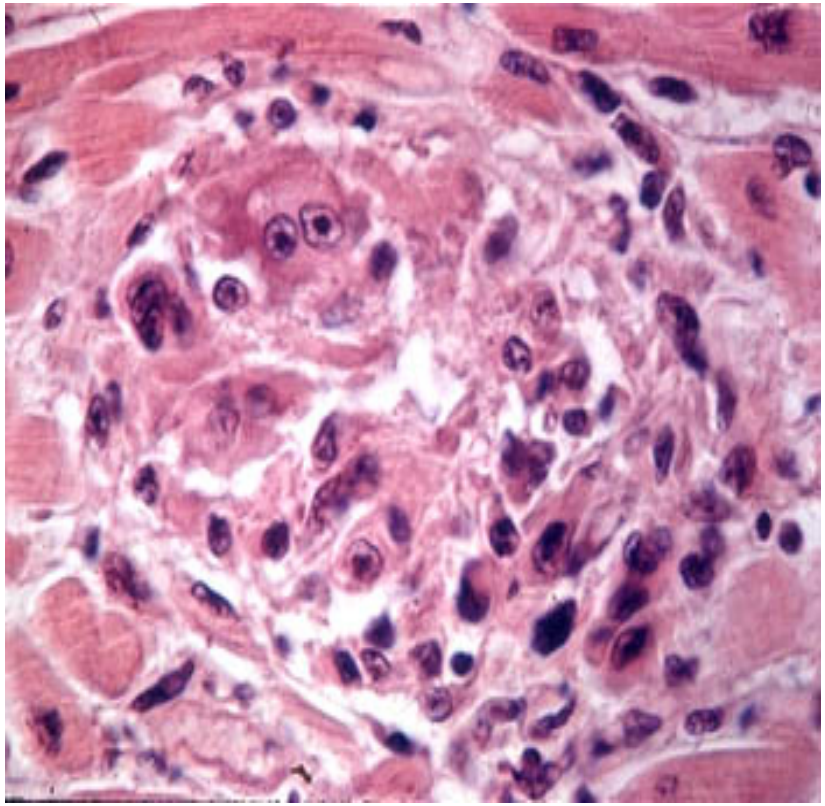


ASCHOFF GIANT CELLS

Multinucleated Macrophages.

Acute Rheumatic Fever

- **ASCHOFF BODIES**



- Inflammatory lesions are found widely scattered in the valves, pericardium, and myocardium of patients who die of acute rheumatic fever (ARF).
- The cellular aggregate seen is typical of the myocardial lesions, is called an Aschoff body (AB).
- The typical AB contains plump histiocytes (Aschoff cells), some of which are multinucleated (Aschoff giant cells).

ASCHOFF BODY (AB).

- The predominance of histiocytic cells, some epithelioid, suggests that the AB is a form of granuloma.
- Aschoff bodies may also contain lymphocytes and plasma cells.
- A central zone of fibrinoid necrosis is seen in Aschoff bodies in early stages of ARF.

ASCHOFF BODY (AB).

- Widespread fibrinoid necrosis and inflammation of the valve substance leads to damage of the overlying endothelial cells.
- With endothelial damage, thrombosis is initiated, and vegetations form along the closing edge of the valve where the leaflets slam together.
- This endocardial damage heals with dense fibrous scars.

Acute Rheumatic Fever

Morphology

PANCARDITIS

Diffuse inflammation and Aschoff bodies in the three layers of heart.

A. **Pericardium (Bread & Butter)**
B. **Myocardium**
C. **Endocardium**

PANCARDITIS

- **PERICARDITIS**
- **MYOCARDITIS**
- **ENDOCARDITIS**
- **MACCALLUM PLAQUES**

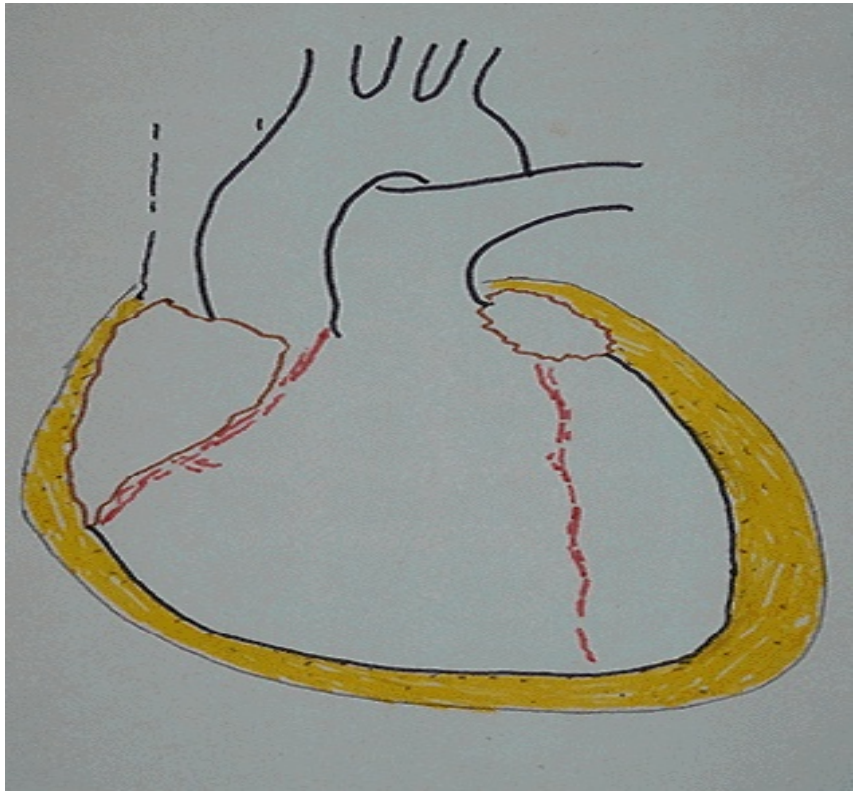
ARhF: PANCARDITIS

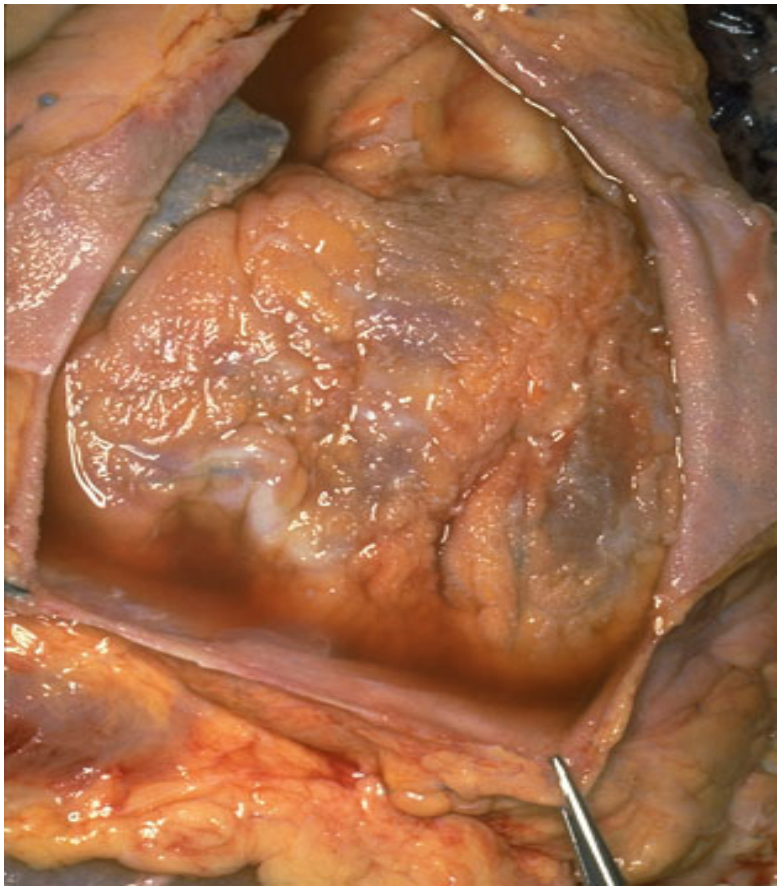
PERICARDITIS

- It is inflammation of the pericardium.
- Fibrinous/ serofibrinous pericardial exudate is present. (**BREAD & BUTTER)**
- Usually resolves without sequel.

ARhF: PANCARDITIS

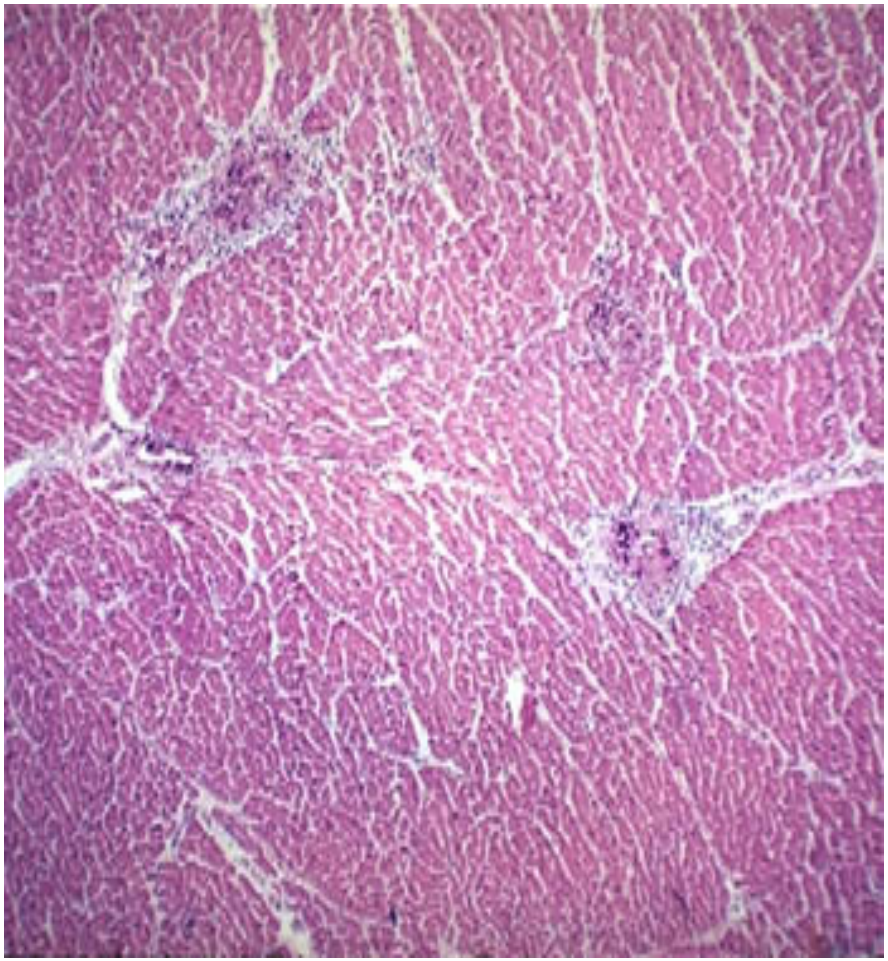
PERICARDITIS





ARhF: PANCARDITIS

Myocarditis

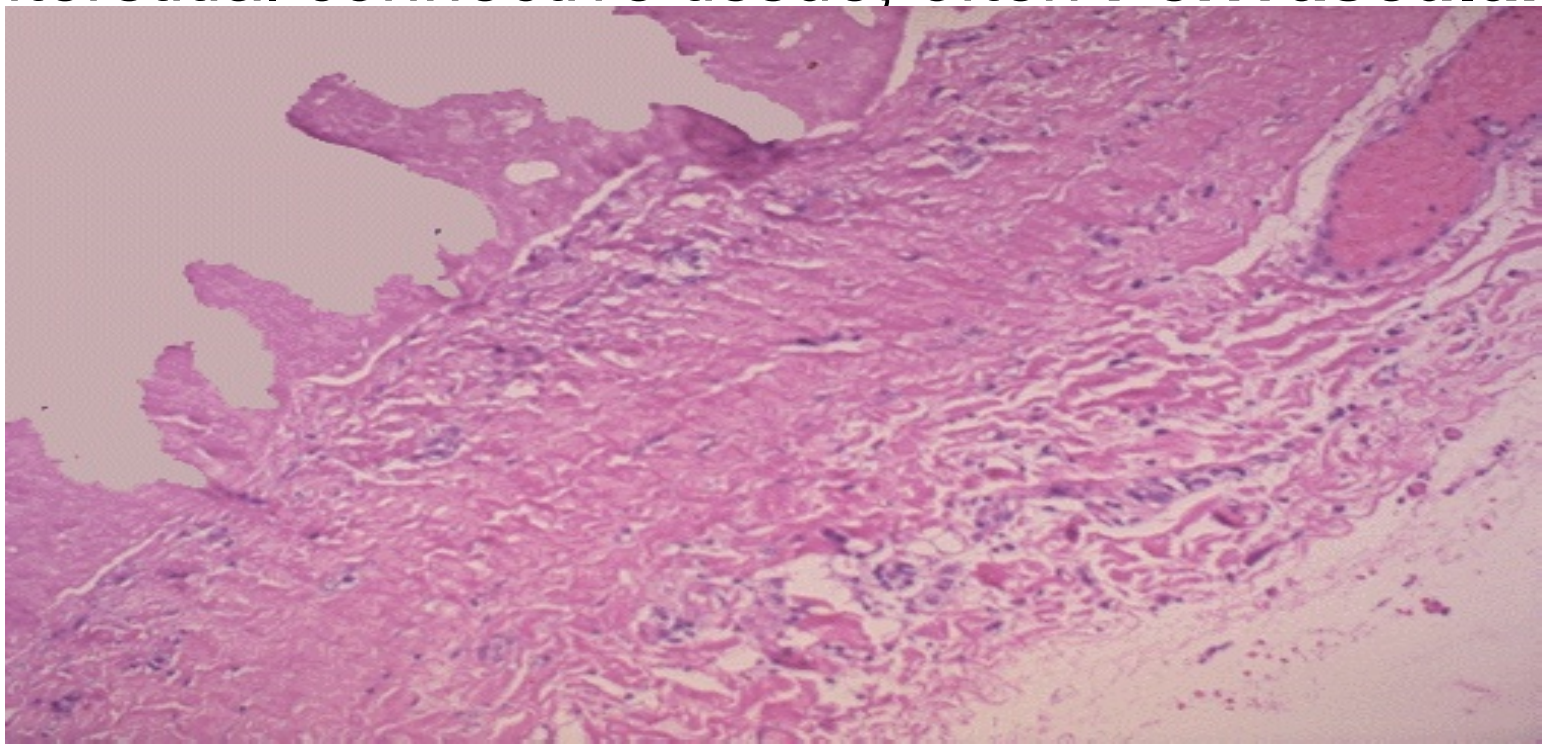


- Section of myocardium from a young patient with severe symptomatic acute rheumatic fever (ARF).
- There are widespread focal inflammatory lesions in the myocardium, predominantly in the interstitium.
- Many lesions are in perivascular connective tissue.
- Myocarditis may be subclinical or may manifest as progressive heart failure.

ARhF: PANCARDITIS

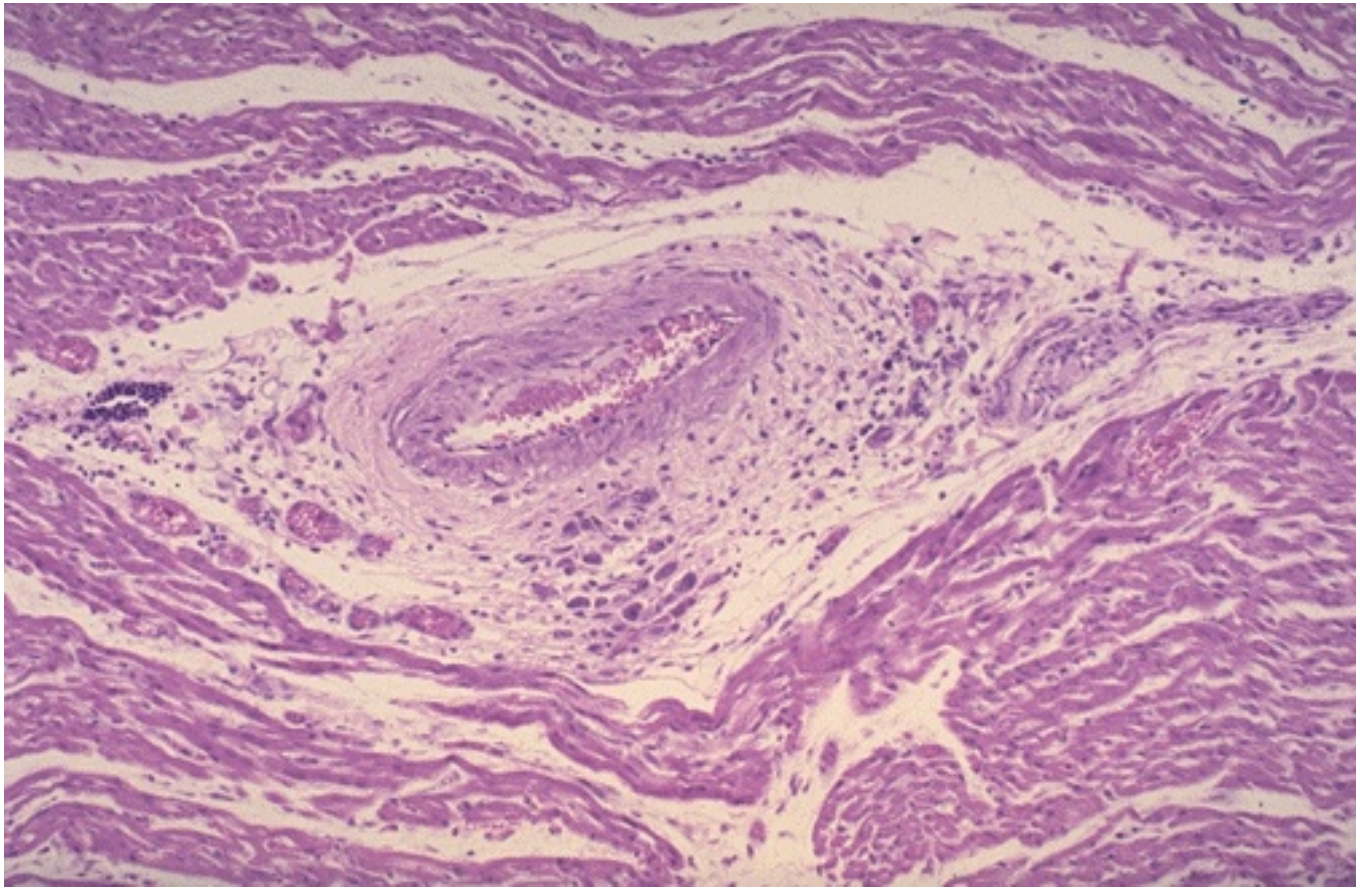
Myocarditis

Consists of scattered Aschoff bodies within the interstitial connective tissue, often **Perivascular.**



ARhF: PANCARDITIS

Myocarditis HP



ARhF: PANCARDITIS

Endocarditis

There is involvement of the endocardium & left sided valves by inflammatory foci.

↓

Fibrinoid necrosis within the cusps/ along tendinous cords.

↓

Formation of vegetations **(Verrucae)** 1-2 mm along the ***lines of closure*** of the valve.

ARhF: PANCARDITIS

Endocarditis

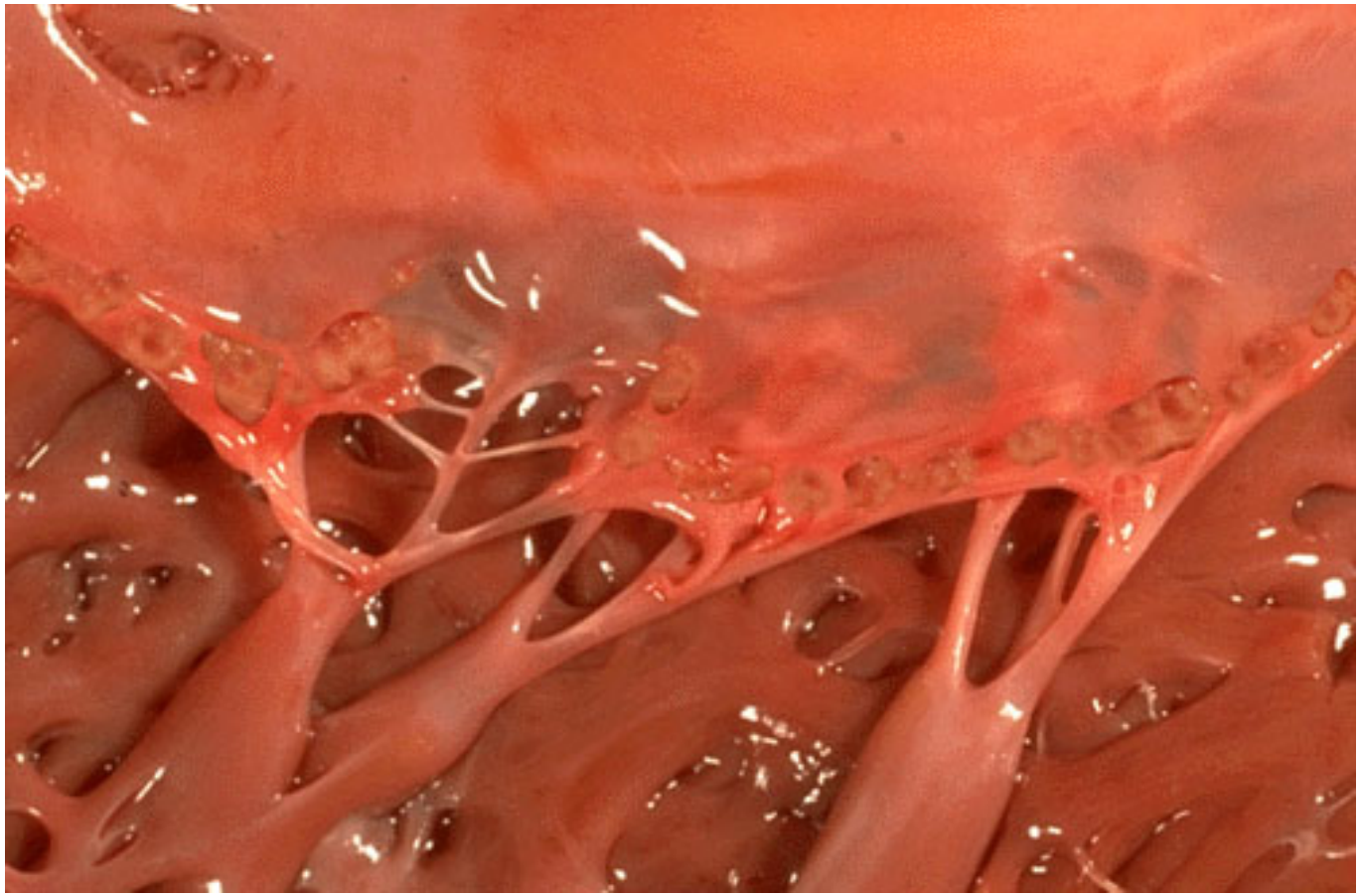
VERRUCAE/ VEGETATIONS IN RF

- 1-2 mm along the lines of closure.
- Irregular warty projections.
- There is little disturbance in cardiac function.
- Arise from precipitation of fibrin at sites of erosion from the underlying inflammation & collagen degeneration.

ARhF: PANCARDITIS

Endocarditis

VERRUCAE/ VEGETATIONS IN RF



ARhF: PANCARDITIS

Endocarditis

MACCALLUM PLAQUES

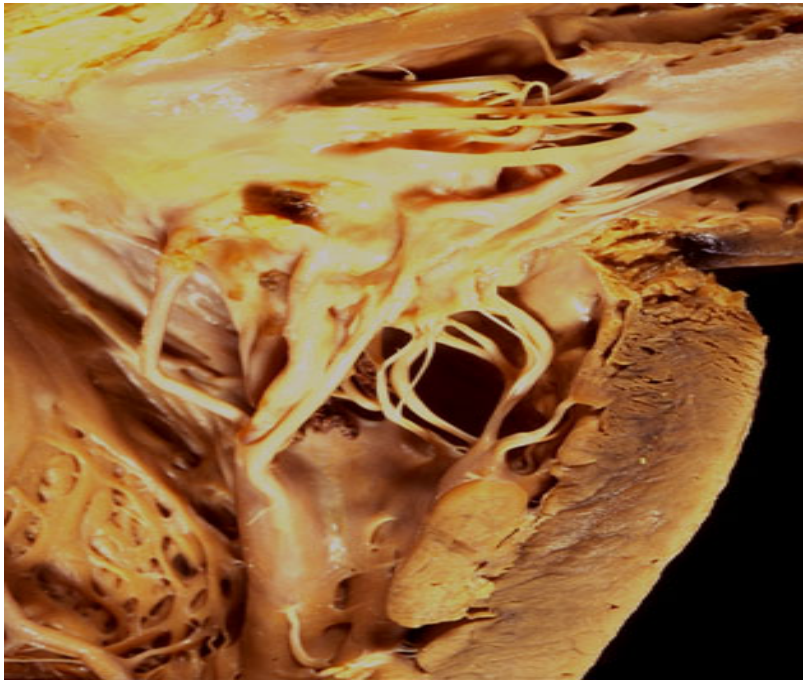
- Subendocardial lesion.
- Present as irregular thickenings in the L.A.
- Exacerbated by regurgitant jets of blood on the atrial wall.

Chronic Rheumatic Heart Disease

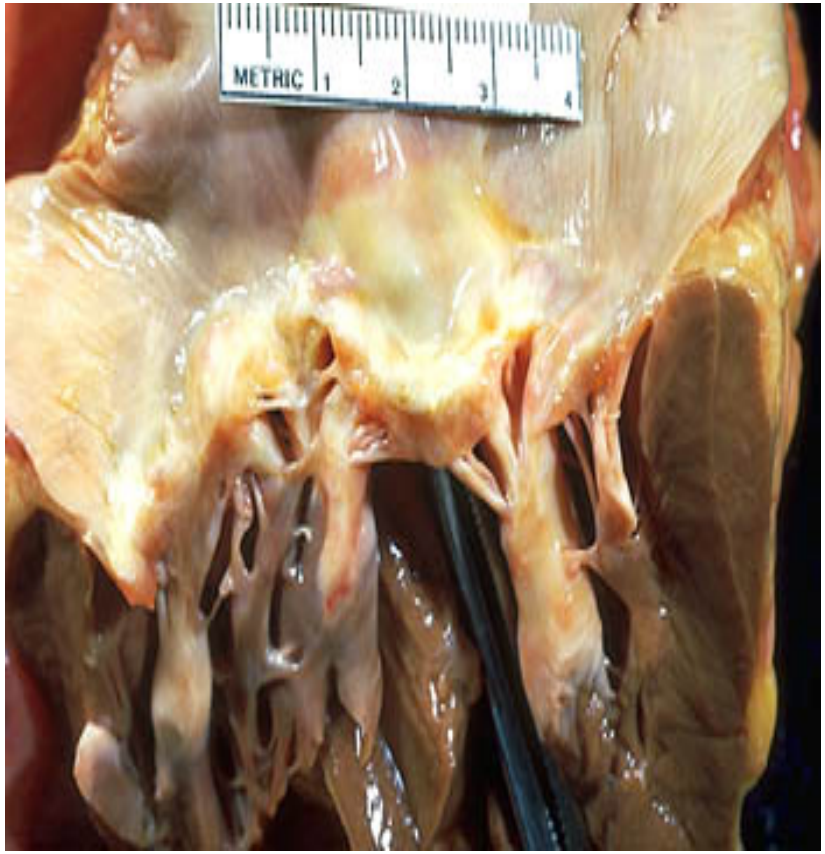
- There is organization of acute inflammation leading to fibrosis.
- **VALULAR LEAFLETS** become thickened and retracted leading to permanent deformity.
- Mainly **Mitral & Tricuspid** valve leaflets are involved with the following anatomic changes.
  - Leaflet Thickening
  - Commisural Fusion
  - Shortening, thickening and fusion of tendinous cords.
- Other valves (eg. Aortic) may be involved in some cases.

Chronic Rheumatic Heart Disease

- Leaflet Thickening
- Shortening, thickening and fusion of tendinous cords.



Chronic Rheumatic Heart Disease

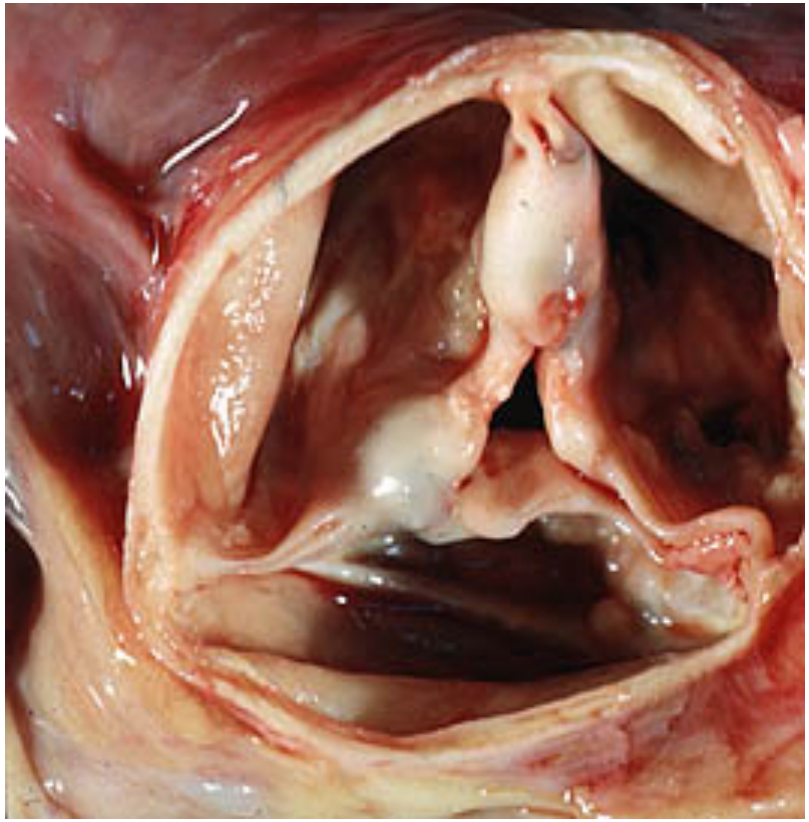


- The fibrotic thickening of the valve leaflets can be seen (patient with postrheumatic mitral stenosis.)

- Fusion and thickening of the chordae tendineae is seen

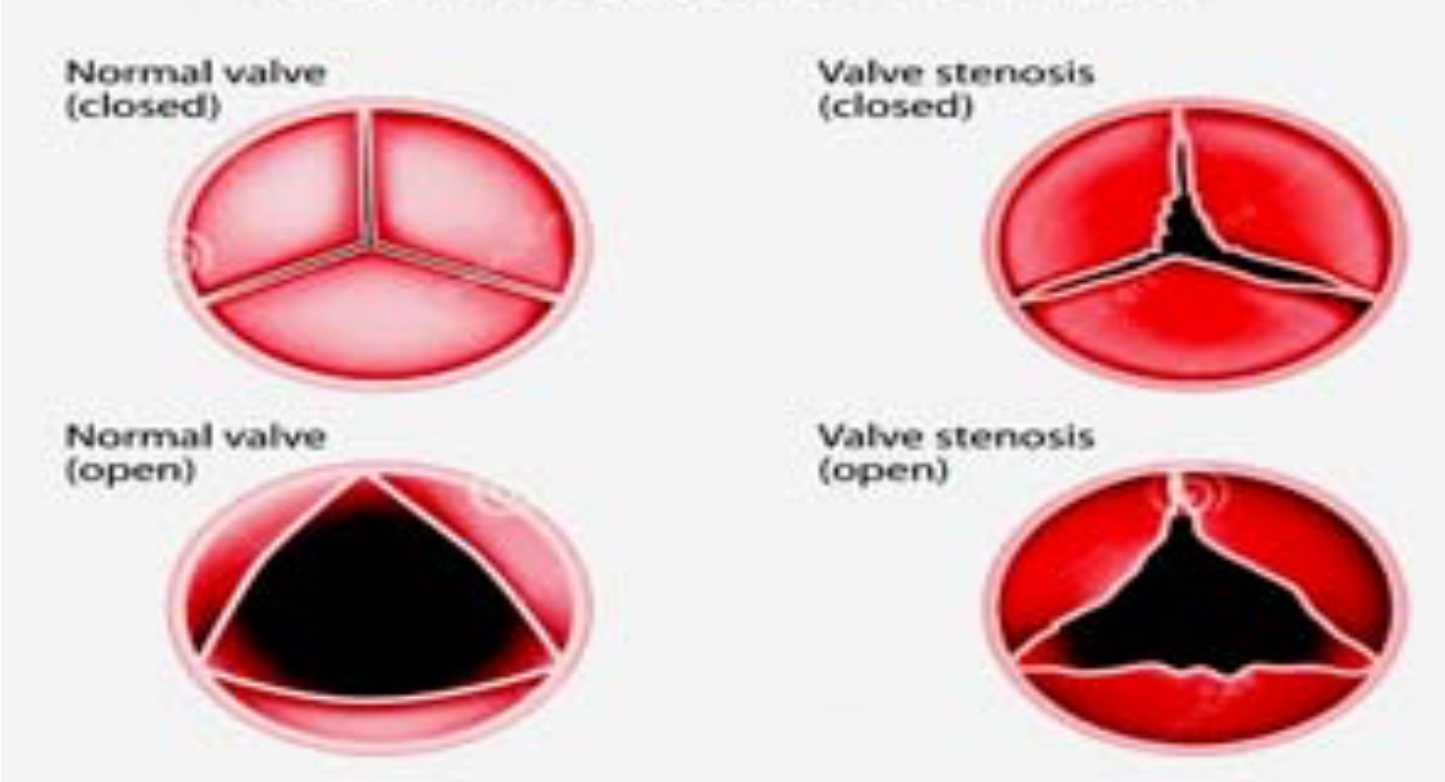
CRHD-Rheumatic Aortic stenosis

- **Commisural Fusion**



- This valve depicts PostRheumatic aortic stenosis.
- Severe cuspal thickening and fusion of the commissures have distorted the three cusps of the valve so that they appear as a single fused unit, fixed in an open but stenotic configuration.
- This valve would generate a complex murmur with components of both aortic stenosis (AS) and aortic insufficiency (AI).

VALVULAR STENOSIS



Chronic Rheumatic Heart Disease

CARDIAC CONSEQUENCES

VALVULAR INVOLVEMENT

- Mitral valve (99%) – most frequently involved
- Only Mitral Valve (65 – 70%)
- Mitral & Arotic Valve (25%)
- Other Valves involved (less severe) are
  - TRICUSPID
  - PULMONARY (VERY RARE)

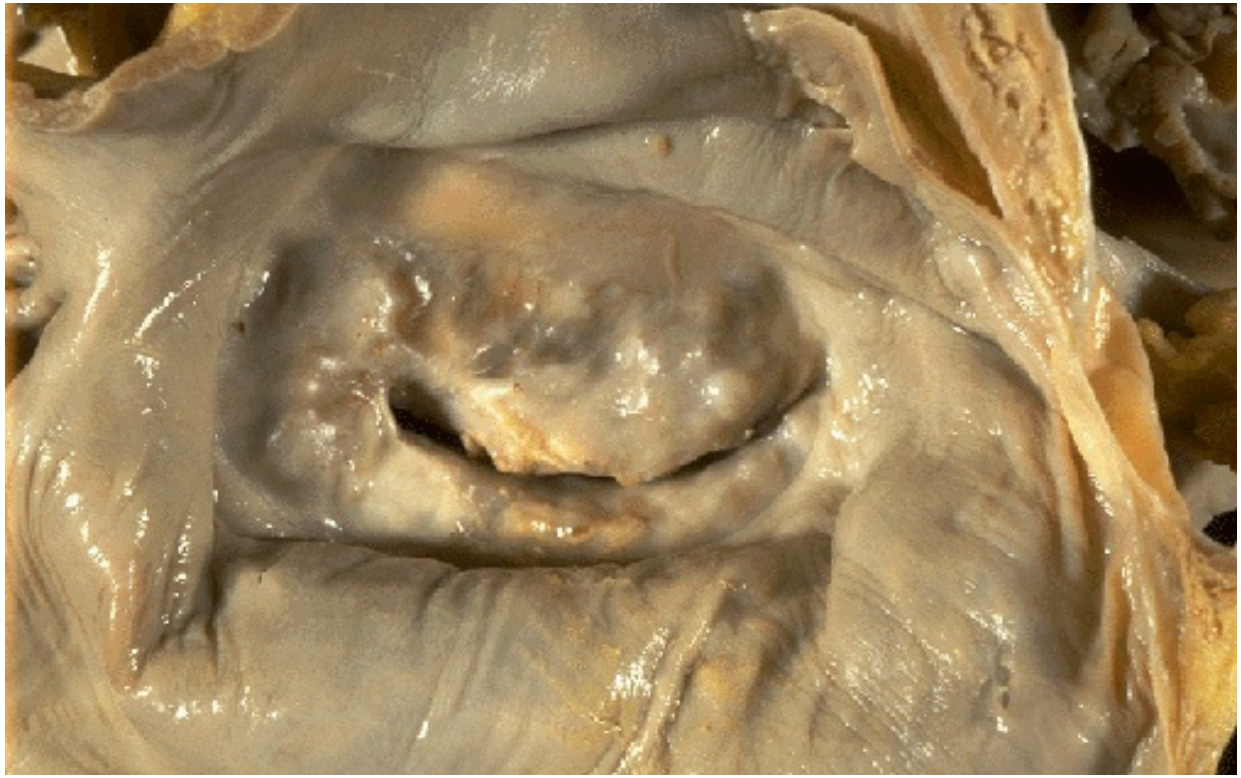
- **FISH MOUTH/ BUTTON HOLE STENOSES** It is seen following fibrous bridging across the valular commissures and calcification.

Chronic Rheumatic Heart Disease

CARDIAC CONSEQUENCES

VALVULAR INVOLVEMENT - Mitral stenoses

FISH MOUTH/ BUTTON HOLE STENOSES



Chronic Rheumatic Heart Disease

MICROSCOPY

- There is diffuse fibrosis and neovascularisation, which obliterates the leaflet architecture (normally avascular)
- Aschoff bodies – Are replaced by fibrous scar.
- Fibrosis from healed inflammation outside the valve is inconsequential.

Chronic Rheumatic Heart Disease

CARDIAC CONSEQUENCES

VALVULAR INVOLVEMENT

CONSEQUENCES OF VALVE INVOLVEMENT

- Mitral stenoses → dilatation of L.A. → Development of mural thrombus in the appendage/ along the wall → congestion in the lungs → pulmonary vascular and parenchymal changes → RVH

- Left Ventricle is usually NORMAL

Rheumatic Heart Disease

CLINICAL FEATURES

Rheumatic Fever is Characterized by a constellation of findings

- **MAJOR**
  - Migratary polyarthritis of large joints.
  - Carditis
  - Subcutaneous nodules
  - Erythema marginatum of skin
  - Sydemham Chorea → characterized by neurologic disorder with involuntary, purposeless, rapid movements.

- **MINOR**
  - Fever
  - Arthralgia
  - Elevated blood levels of acute phase reactants.

Rheumatic Heart Disease

Diagnosis

JONES CRITERIA

Evidence of preceding group A Streptococcal infection along with –

- **2 Major Manifestations**
- **1 Major + 2 Minor manifestations.**

Rheumatic Heart Disease

Clinical Features

ARTHRITIS

- More common in adults.
- Migratory polyarthritis accompanied by fever.

ACUTE CARDITIS

- Clinically present with following features
  - Pericardial friction rubs
  - Weakened heart sounds
  - Tachycaria
  - Arrythmias

Myocarditis

- Leads to cardiac dilatation → MI → CHF

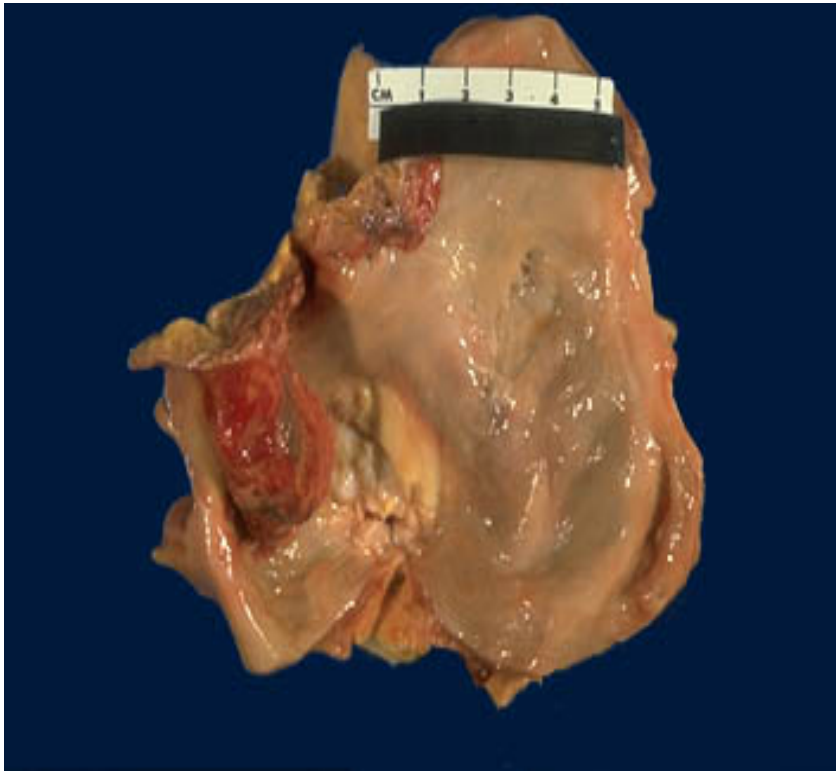
Chronic Rheumatic Heart Disease

Complications

- There is an increased risk with each recurrence.
- Embolisation from mural thrombi (from atria/ appendages)
- Infective Endocarditis on deformed valves.

Chronic Rheumatic Heart Disease

Complications



- The mitral valve has been reduced to a narrow orifice.
- The left atrium is markedly dilated.
- Thrombi have formed on the atrial wall.(atrial endocardial surface)

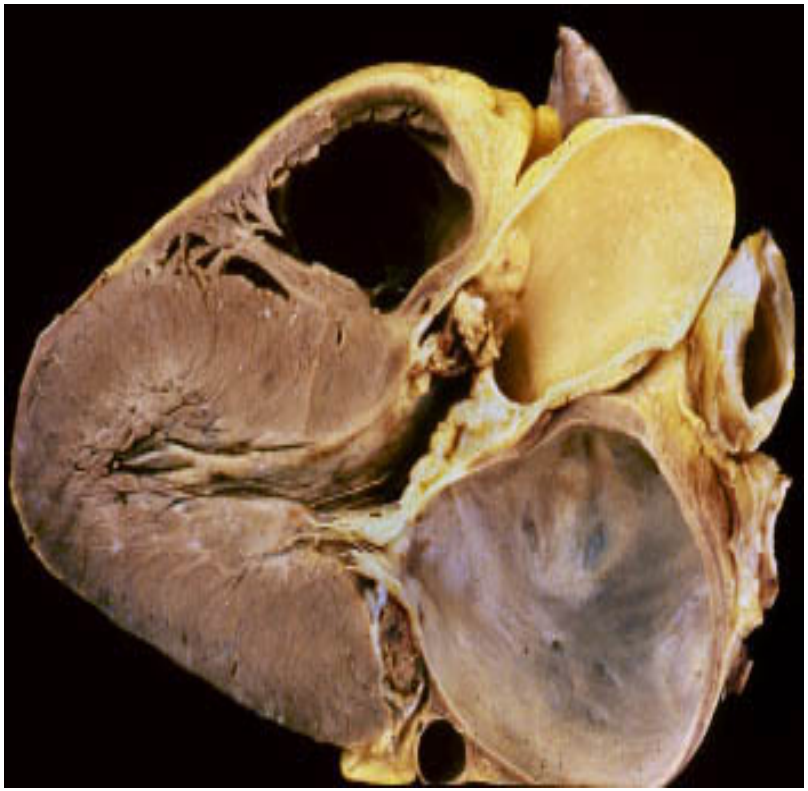
Chronic Rheumatic Heart Disease

Complications

- Usually there are no manifestations for years to decades after initial episode.
- S/s relating to valvular disease are present (depending on the valves involved).
  - eg. cardiac murmurs, cardiac hypertrophy/dilatation, Heart failure.
- Arrythmias
- Thromboembolism
- Infective Endocarditis.

Chronic Rheumatic Heart Disease

Cardiac hypertrophy/dilatation



- Both the mitral and the aortic valves are affected.
- Severe, long-standing, aortic stenosis has resulted in left ventricular hypertrophy.
- Severe left atrial dilation is a result of mitral stenosis.

Rheumatic Heart Disease

SURGICAL TREATMENT

Repair of the diseased valves done by incising the fused commisures and replacement with prosthetic devices.

Rheumatic Heart Disease

CONSEQUENCES

Chronic valvular deformities (Valvulitis)

characterized by -

Deforming fibrotic valvular disease

↓

Mitral Stenosis

Permanent Valvular dysfunction

↓

Severe/ fatal cardiac problems. (decades later)

Rheumatic Heart Disease

PROGNOSIS

- **Good for primary attack (1% mortality).**
- **Increased vulnerability to reactivation of disease is present with subsequent pharyngeal infections.**

THANK YOU