

CVS MODULE - 1

Rheumatic heart disease

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Rheumatic heart disease

- ▶ Rheumatic heart disease is the cardiac manifestation of rheumatic fever.
- ▶ **Rheumatic fever** is an acute, immunologically mediated, multisystem inflammatory disease that occurs after **group A β -hemolytic streptococcal** infections (usually pharyngitis, but also occasionally infections at other sites, such as skin).
- ▶ valvular inflammation and scarring produce the most important clinical features, rheumatic heart disease is essentially the only cause of acquired mitral stenosis.

Rheumatic heart disease

- ▶ The illness is so named because of its similarity in presentation to rheumatism.
- ▶ Multisystem disease affecting the heart, joints, brain, cutaneous and subcutaneous tissues.
- ▶ Acute RF remains an important preventable cause of cardiac disease.

Pathogenesis

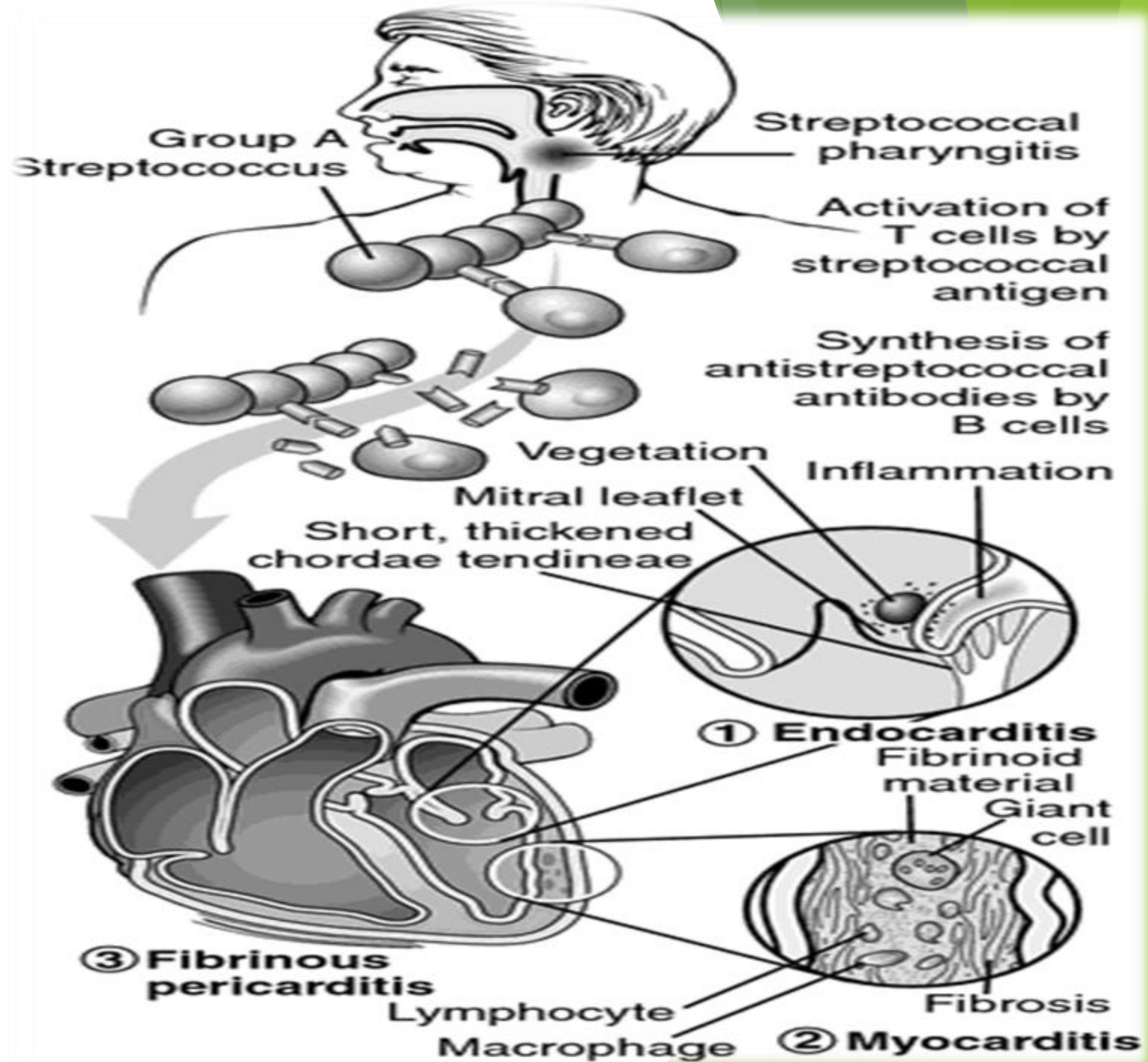
- ▶ The pathogenic mechanism involves **autoantibodies and T cells directed** against group A streptococcal cell wall components that cross-react with heart or brain.
- ▶ Predisposing factors include agent ,genetic and environmental factors such as poverty and overcrowding.

Host and environmental factors

- ▶ Age: Adolescents 5-15 years but the initial attack is at younger age.
- ▶ No gender predilection.
- ▶ Common in 3rd world countries.
- ▶ Environmental factors-- over crowding, poor sanitation, poverty, poor housing.
- ▶ A family history and lower socioeconomic status are additional factors.

Pathogenesis

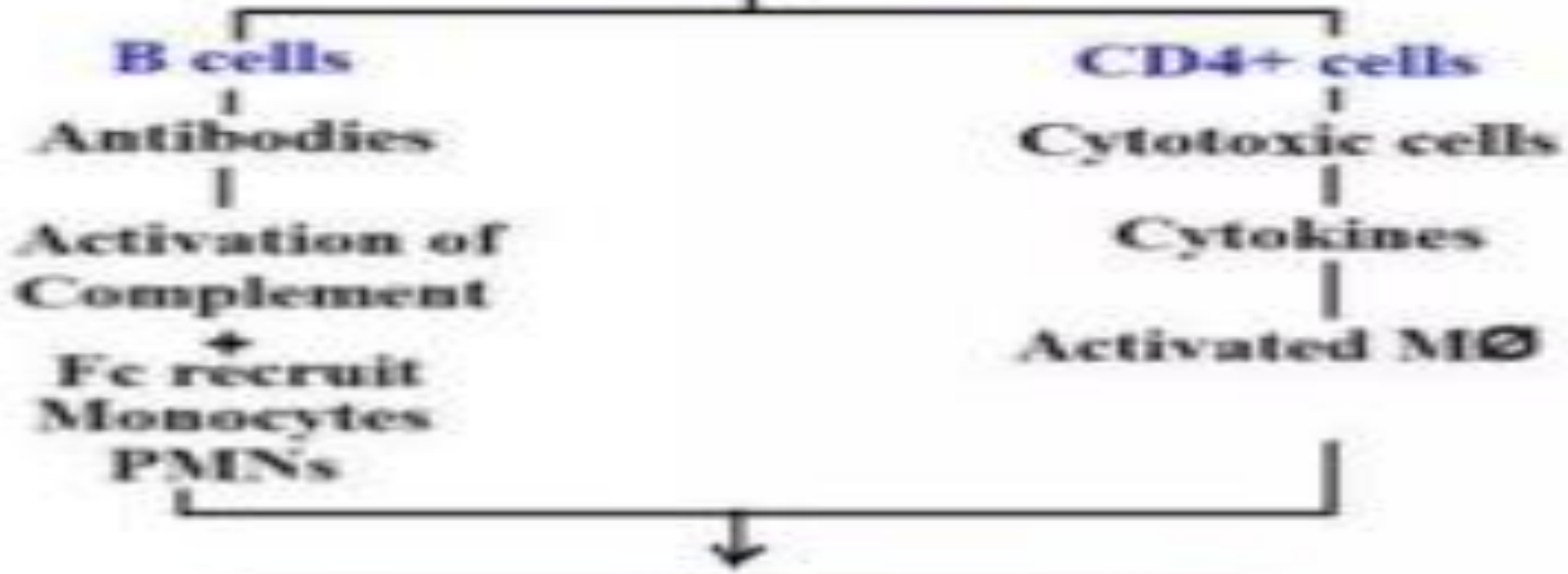
- ▶ There is no direct invasion to the tissue by the microorganisms but its an **autoimmune disease** that involves Ag-Ab interaction.
- ▶ It is due to an immunologic reaction that is a delayed sequela of group A beta-hemolytic streptococcal infections of the pharynx.
- ▶ It must be pharyngeal infection not skin infection.



Pathogenesis

1. Group A streptococcal (GAS) pharyngeal infection ----→
2. Body produce antibodies against streptococci ----→
3. These antibodies cross react with human tissues because of the **antigenic similarity** between streptococcal components and human connective tissues (molecular mimicry)[there is certain amino acid sequence that is similar between GAS and human tissue, like heart, joint, brain and connective tissues].

**Sterptococcal antigen
M Protein**

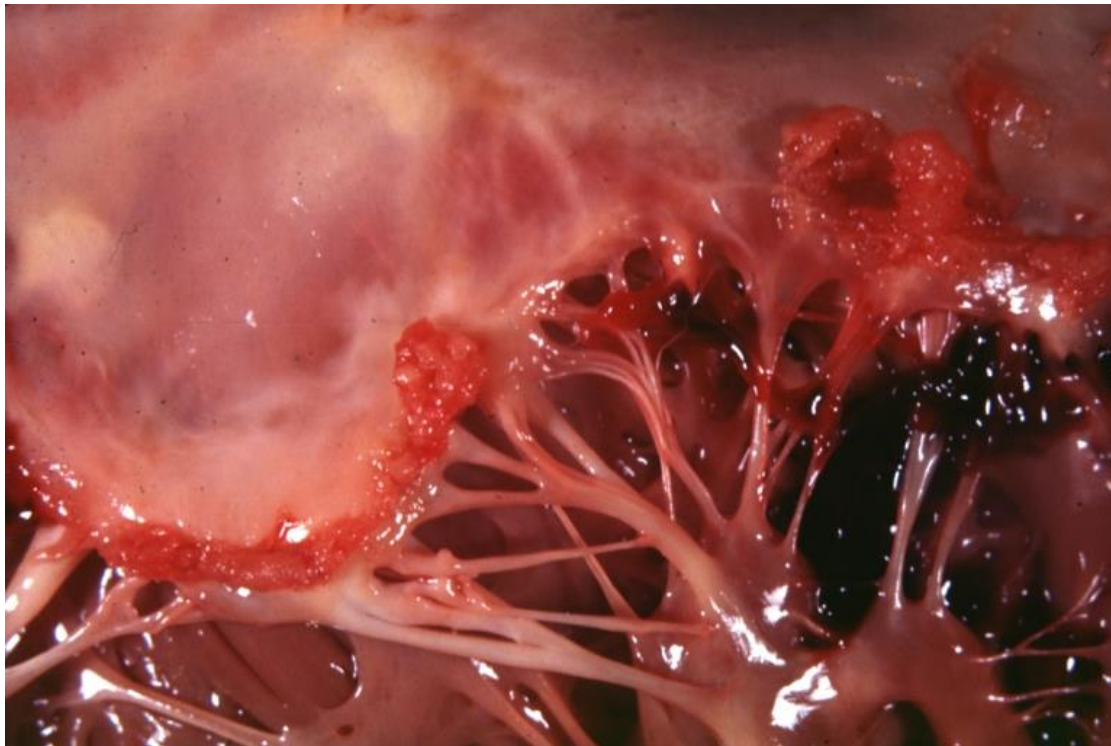


Damage to heart tissue

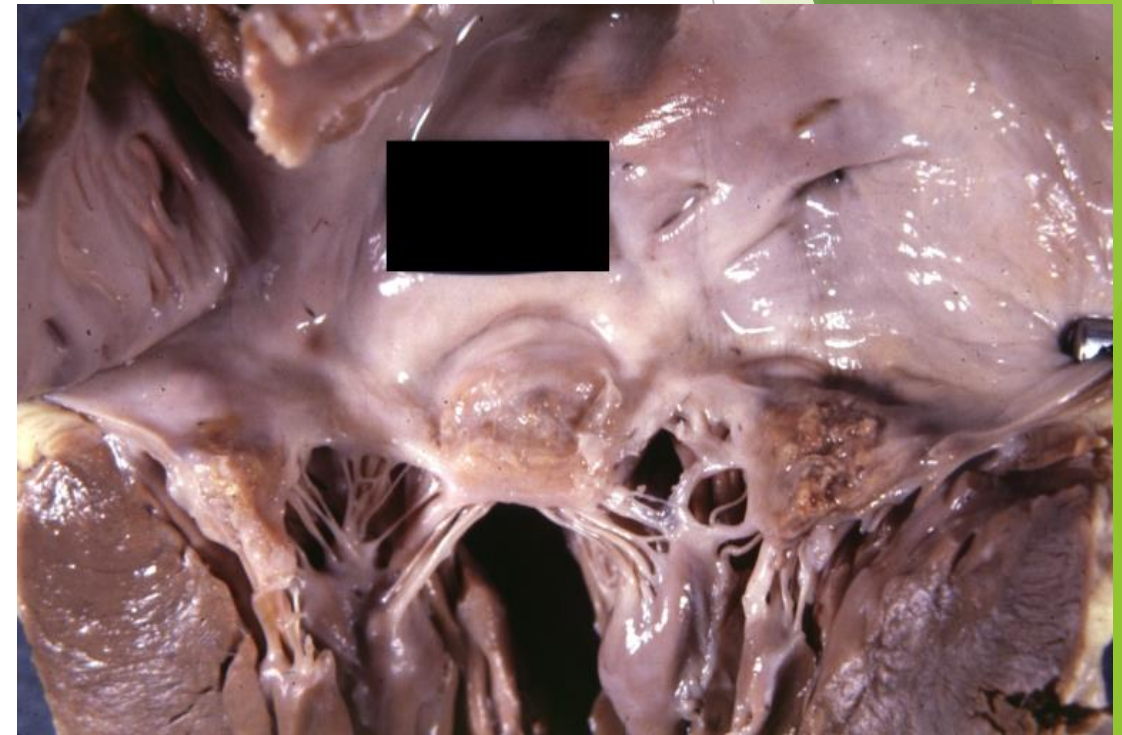
NOTE: M protein and cardiac protein has got structural similarities

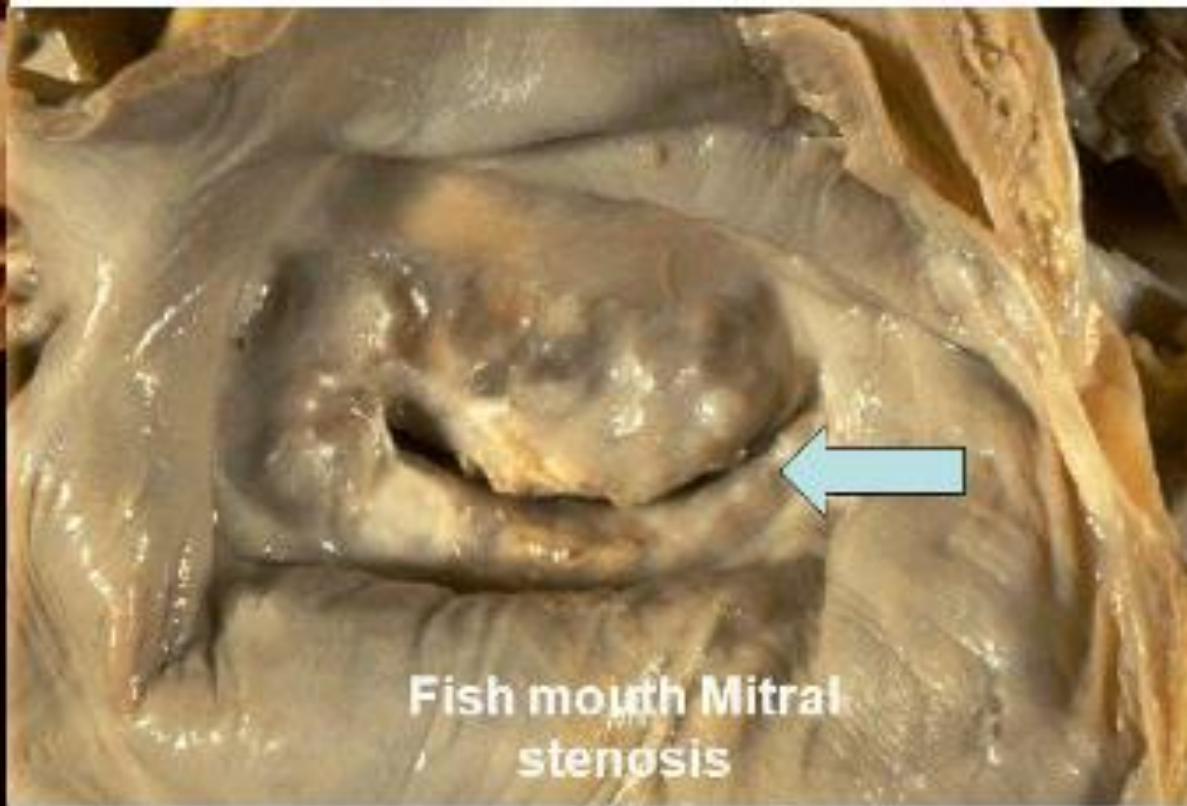
Grossing

MITRAL VALVULITIS



MITRAL SCARRING





The pericardium may exhibit a fibrinous exudate, which generally resolves without sequelae.

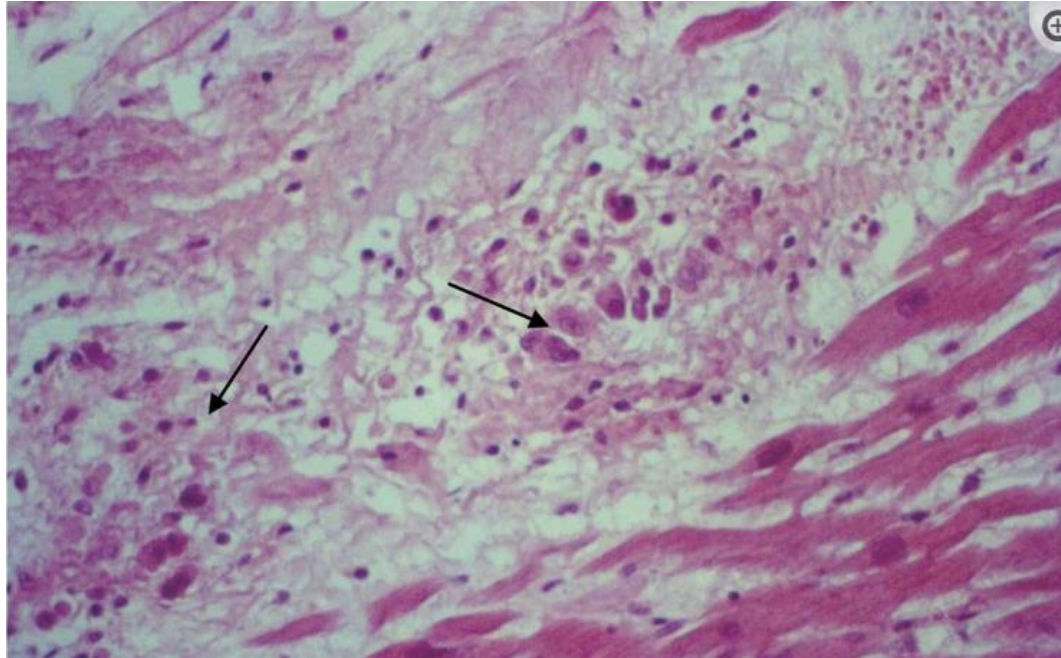


Histology

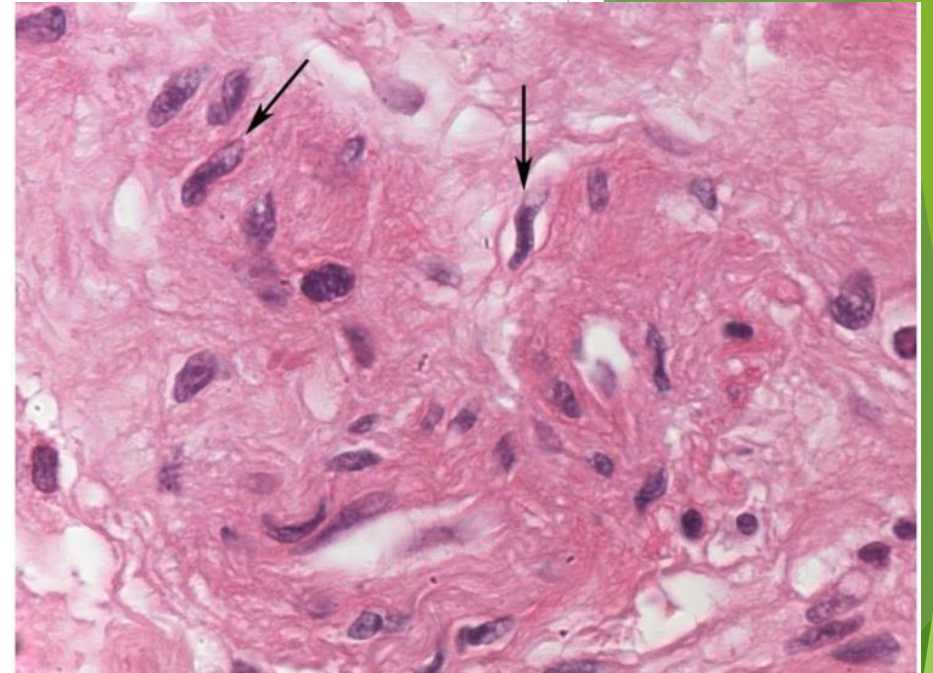
- ▶ Acute rheumatic valvulitis manifests with :
 - ✓ active inflammation characterized by lymphocytic infiltration, **Aschoff bodies**, and **Anitschkow cells** (have abundant cytoplasm and nuclei with chromatin that is centrally condensed).

- ▶ Chronic rheumatic valvulitis is associated with:
 - ✓ Neovascularization, valvular fibrosis and/or dystrophic calcification.

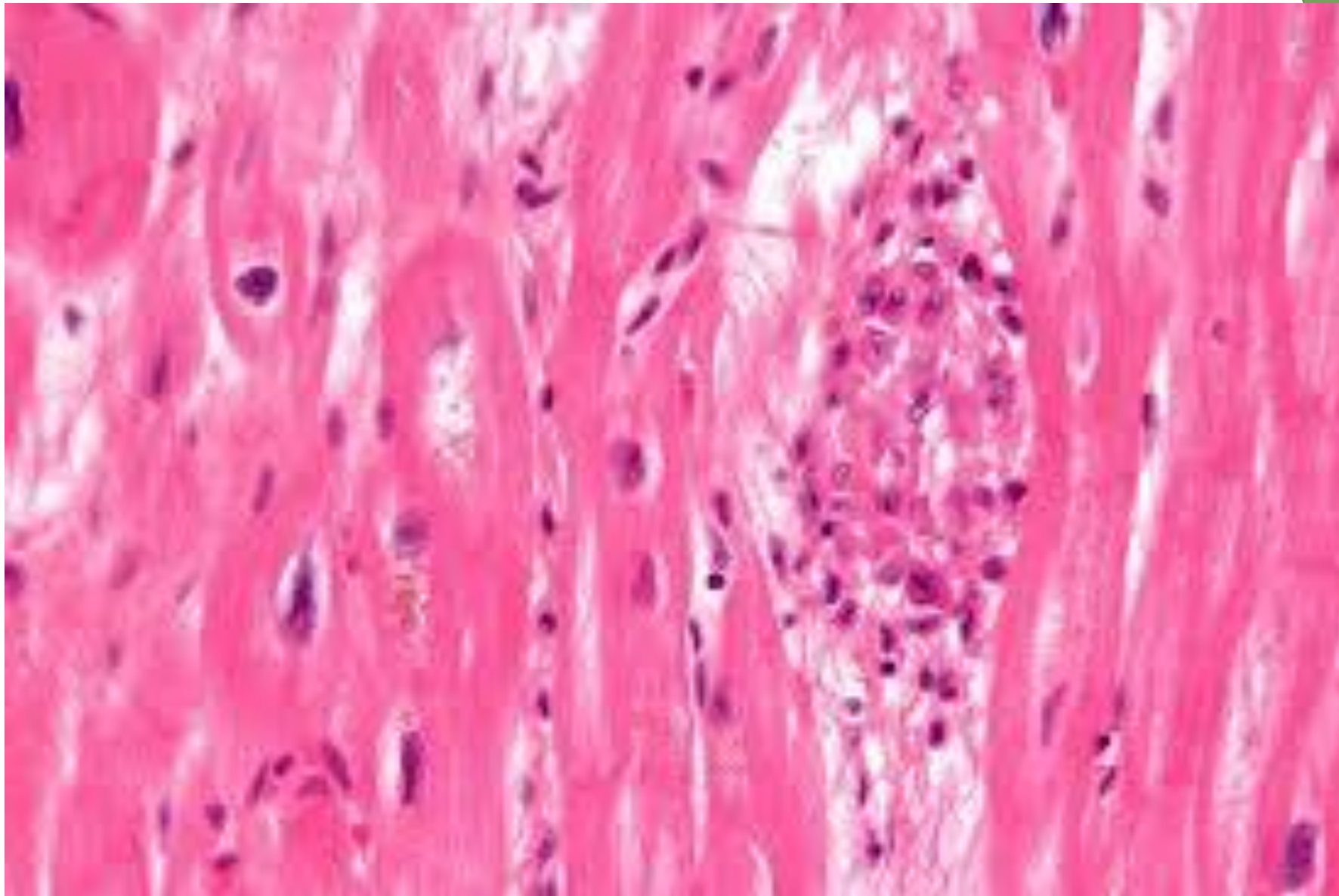
- ▶ During acute rheumatic fever, Aschoff bodies can be found in any of the three layers of the heart—pericardium, myocardium, or endocardium (including valves).



Aschoff nodules



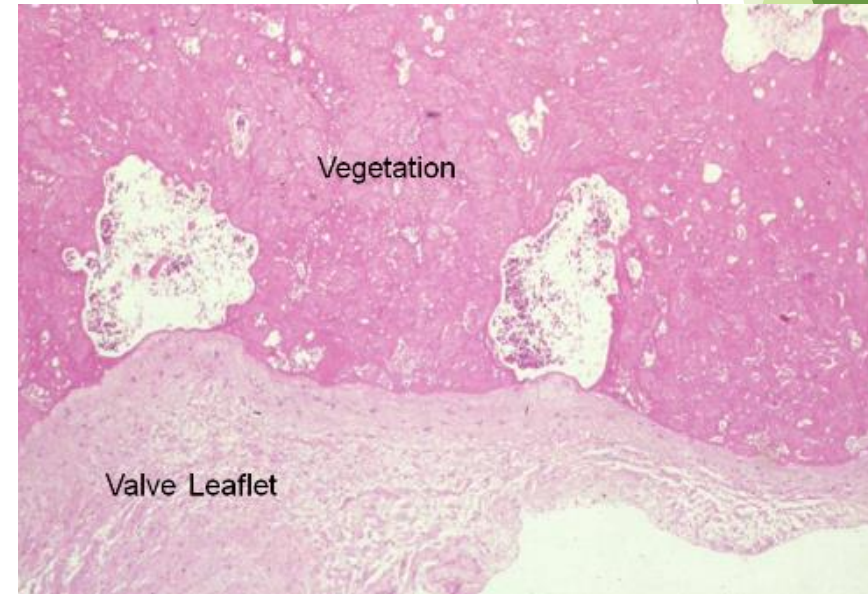
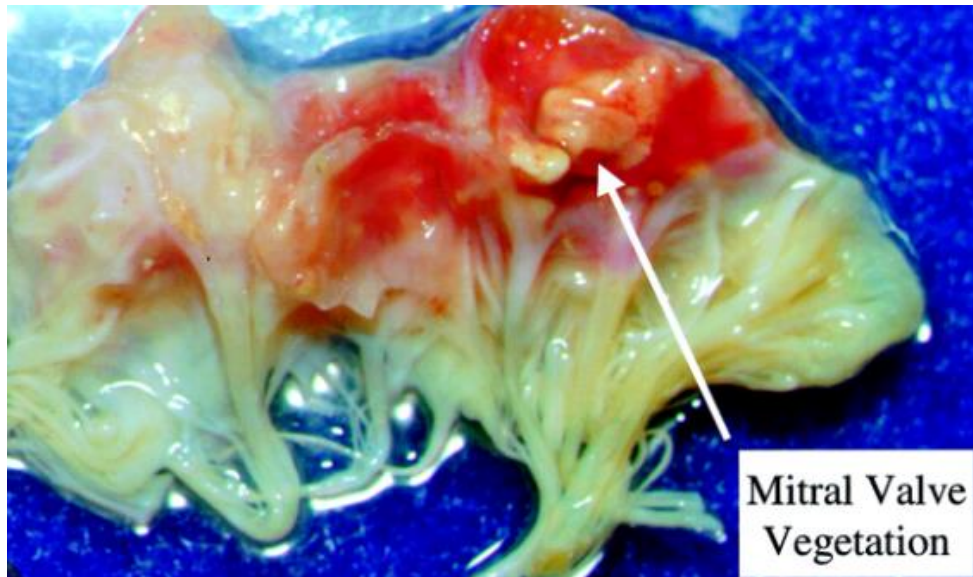
Anitschkow cells.



Aschoff nodules

Histology cont.:

- Valve involvement results in fibrinoid necrosis and fibrin deposition along the lines of closure forming 1- to 2-mm vegetations—verrucae—that cause little disturbance in cardiac function.



Note the numerous small rheumatic vegetation on the line of closure of mitral valve These are evanescent in most . In recurrent rheumatic fever the same lesions recur with vigor and become sticky fibrotic and chronic degeneration take place to result in mitral stenosis



Clinical Features

- ▶ Acute rheumatic fever occurs most often in children; the principal clinical manifestation is carditis.
- ▶ However, about 20% of first attacks occur in adults, with arthritis being the predominant feature.
- ▶ Symptoms in all age groups typically begin 2 to 3 weeks after streptococcal infection.
- ▶ The clinical signs of carditis include pericardial friction rubs and arrhythmias; myocarditis may be sufficiently severe to cause cardiac dilation and resultant functional mitral insufficiency and CHF.

RHEUMATIC FEVER

DUCKETT-JONES DIAGNOSTIC CRITERIA

MAJOR CRITERIA

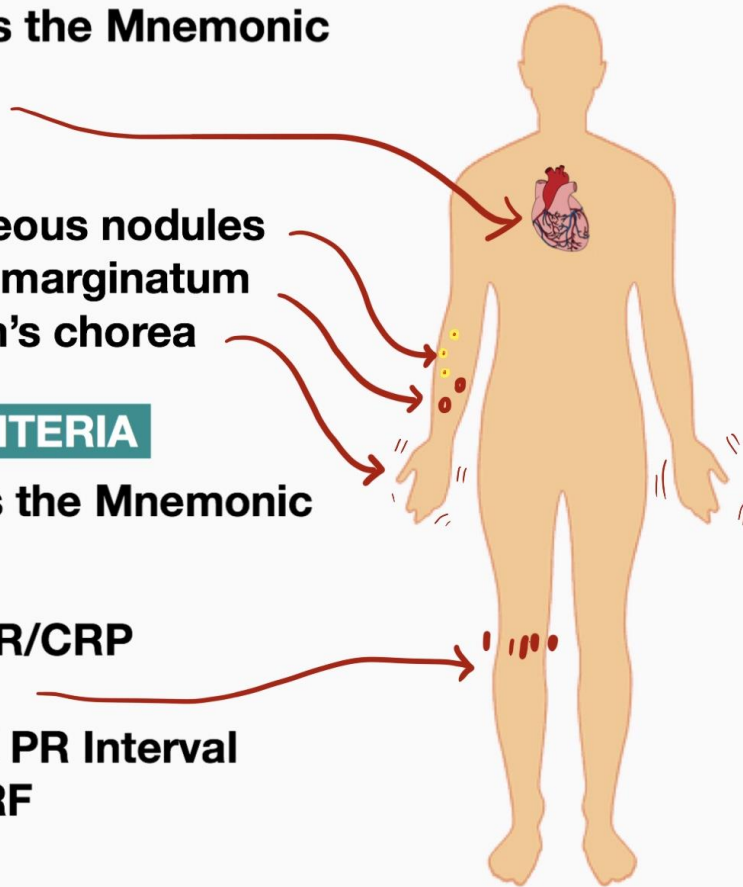
“CASES” is the Mnemonic

- C**arditis
- A**rthritis
- S**ubcutaneous nodules
- E**rythema marginatum
- S**ydenham's chorea

MINOR CRITERIA

“FRAPP” is the Mnemonic

- F**ever
- R**aised ESR/CRP
- A**rthralgia
- P**rolonged PR Interval
- P**revious RF



There must be evidence of streptococcal infection plus:

2 major or 1 major + 2 minor

Erythema Marginatum



Rheumatic fever-diagnosis



Subcutaneous nodules

(nodules of rheumatoid arthritis are larger)

lab



- ▶ ESR, CRP.
- ▶ Elevated serum titers of antibodies against one or more streptococcal antigens (e.g., streptolysin O or DNAase) .
- ▶ Cultures are negative for streptococci at the time of symptom onset

Treatment

- ▶ **Treatment strategies for acute rheumatic fever (ARF) can be divided into the following:**
 - ✓ **Management of the acute attack**
 - ✓ **Management of the current infection**
 - ✓ **Prevention of further infection and attacks**

GOOD LUCK

THANK YOU