ACUTE RHEUMATIC FEVER

RHEUMATIC HEART DISEASE

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ACUTE RHEUMATIC FEVER

- A consequence of pharyngeal infection with group A B-hemolytic streptococci which cause sore throat and scarlet fever .
- An autoimmune inflammatory process that develops as a sequel of streptococcal pharyngitis
- Most common in children: 5-15 years
- It usually takes about 1 to 5 weeks to develop if not treated properly
- Mainly in developing countries

RHEUMATIC HEART DISEASE

- The most significant complication of ARF is rheumatic heart disease
- Rheumatic fever is thought to be caused by a response of the body's defense system not a direct effect of bacteria, the immune system response to infection which formed anti bodies against M protein (arises from bacteria) cross react with self antigen (often myosin) – molecular mimic – this causes an autoimmune reaction against native tissues in the heart
- Immune mediated (type II hypersensitivity)

RHEUMATIC HEART DISEASE

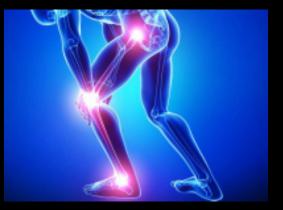
- Exacerbated by incorrect T cell activation
- Rheumatic valves display increased expression of VCAM-1 (vascular cell adhesion protein) which leads to more lymphocyte entry >> more inflammation

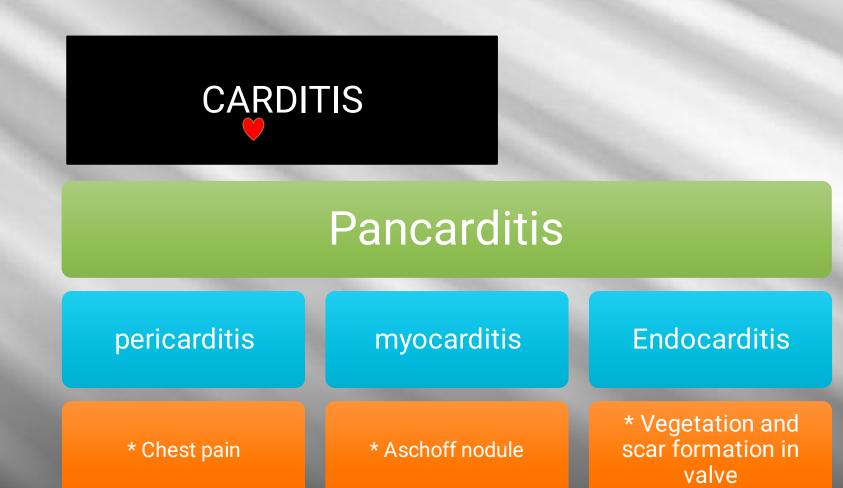
CLINICAL FEATURES

- Major criteria : J[♥]NES
- Joint
- -(carditis)
- -Nodular in skin
- -Erythema marginatum
- -Sydenham chorea

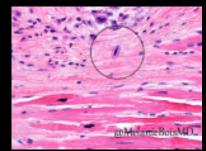


- Large joint
- Migratory polyarthritis (sever pain and swelling)
- 75% of patients
- Improves with analgesics





- Aschoff nodule : granuloma macrophage with giant cell
- Anitschkow cell (enlarged macrophage with ovoid, wavy, rod like nucleus)
- Most common valve affected (rheumatic) M > A > T > P
- Early lesion mitral valve regurgitation
- Late lesion mitral valve stenosis
- Arrhythmia most common cause of death
- 40-60% of patients



NODULAR IN SKIN & ERYTHEMA MARGINATUM

Nodular in skin

- Small (.5-2 cm)
- Extensor surface of bone and tendons
- 10% of patients
- Weeks
- Rare



- Erythema marginatum
 (evanescent rash with ring margin)
 - Concentrated on the trunk , NOT face
 - 10% of patients
 - Rare & Early



SYDENHAM CHOREA

- Rapid Irregular Aimless involuntary movements of the arms, legs, trunk, or facial muscles
- 10% Of patients
- Autoimmune to basal ganglia (minor)
- 8 months to appears



MINOR CRITERIA

1. Fever of 38.2–38.9 °C

2. Arthralgia: Joint pain without swelling LABORATORY:

1. elevated acute phase reactants (ESR,CRP)

2.prolonged PR interval AV block, (Cannot be included if carditis is present as a major symptom).

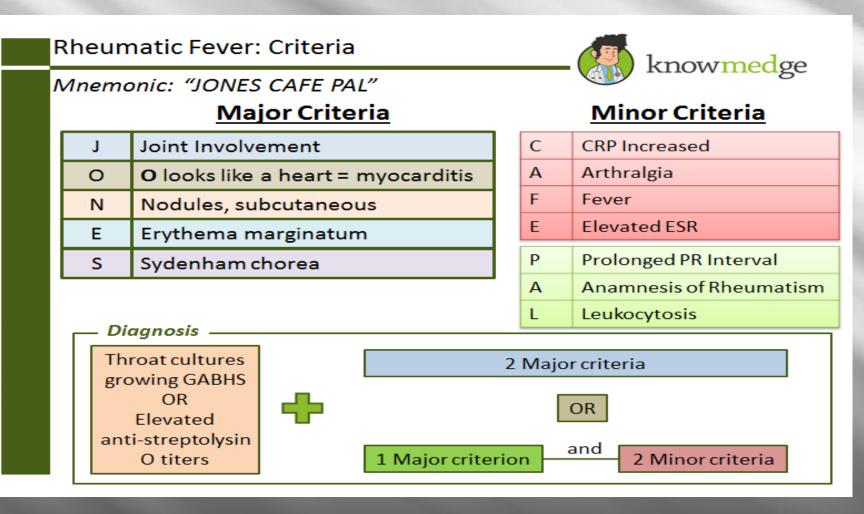
3.Leukocytosis.

4. Previous episode of rheumatic fever or inactive heart disease.

5 increase in anti-streptolysin O (ASO) titers

- If the patient has arthritis, then polyarthralgia cannot be considered as minor criterion.

- If the patient has carditis the prolonged P-R interval cannot be considered as minor criteria



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MANAGEMENT

- 1. Bed rest :
- It lessens joint pain and reduces cardiac workload.
- The duration should be guided by symptoms, along with temperature, leucocyte count and ESR, and should be continued until these have settled.
- Patient can then return to normal physical activity but strenuous exercise should avoided in those who have had carditis.

2. Treatment of cardiac failure :

- If heart failure in these cases does not respond to medical treatment, valve replacement may be necessary and it often associated with a dramatic decline in rheumatic activity .
 - Occasionally, AV block may occur but is seldom progressive and usually resolve spontaneously .
- Rarely, pacemaker insertion may be required .

3. Antibiotic :

- A single dose of benzathine benzylpenicillin (1.2 million U IM) or oral phenoxymethylpenicillin (250 mg 4 times daily for 10 days) should be given to eliminate any residual streptococcal infection.
- If the patient is penicillin-allergic, erythromycin or a cephalosporin can be used .
- For secondary prophylaxis : benzathine benzylpenicillin (1.2 million U IM) or oral phenoxymethylpenicillin (250 mg twice daily), sulfadiazine or erythromycin may be used if the patient is allergic to penicillin.
- Further attacks are unusual after the age of 21, when antibiotic treatment can usually be stopped.

- The duration of prophylaxis should be extended if an attack has occurred in the last 5 years, or if the patient lives in an area of high prevalence and has occupation (such as teaching) with a high risk of exposure to streptococcal infection.
- In those with residual heart disease, prophylaxis should continue until 10 years after the last episode or 40 years of age, whichever is later.

3. Aspirin :

- This usually relieves the symptoms of arthritis rapidly and a response within 24 hours helps confirm the diagnosis.
- A reasonable starting dose is 60 mg/kg body weight/day, divided into six doses .
- An adult, 100mg/kg per day may be needed up to the limits of tolerance or a maximum of 8 g per day.
- Aspirin should be continued until the ESR has fallen and then gradullay tailed off .
- Mild toxicity includes nausea, tinnitus and deafness

vomiting, tachypnoea and acidosis are more serious .

4. Glucocorticoids :

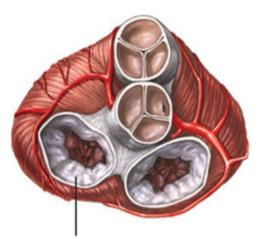
- These produce more rapid symptomatic relief than aspirin and are indicated in case with carditis or sever arthritis .
- There is no evidence that long term steroids are beneficial.
- Prednisolone (1-2 mg/kg per day in divided doses)should be contiued until the ESR is normal and then tailed off.

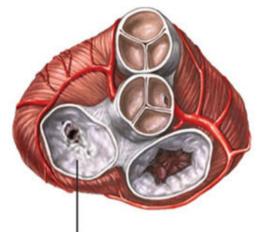
CHRONIC RHEUMATIC HEART DISEASE

- Develops in at least half of those affected by rheumatic fever with carditis.
- Two third of cases occur in women .
- Some episode of rheumatic fever pass unrecognized and it is possible to elicit a history of rheumatic fever or chorea in only about half of all patient with chronic rheumatic heart disease.
- The mitral value is affected in more than 90% of cases, the aortic value is the next most frequently involved, followed by the tricuspid and then the pulmonary value.
- Isolated mitral stenosis accounts for about 25% of all cases, and an additional 40% have mixed mitral stenosis and regurgitation.

PATHOGENESIS

- The main pathological process is progressive fibrosis .
- The heart value are predominantly affected but involvement of the pericardium and myocardium also occurs and may contribute to heart failure and conduction disorders.
- Fusion of the mitral valve commissures and shortening of the chordae tendineae may lead to mitral stenosis with or without regurgitation.
- Similar changes in the aortic and tricuspid valves produce distortion and rigidity of the cusps, leading to stenosis and regurgitation.
- Once a valve has been damaged, the altered haemodynamic stresses perpetuate and extend the damage, even in the absence of a continuing rheumatic process.





Heart with Normal Mitral Valve

Narrowing down of Mitral Valve Rheumatic Mitral Valve (With Stenosis)

