

Rheumatic Fever

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Introduction and Background

- Rheumatic fever causes chronic progressive damage to the heart and its valves.
- The disease has been known for many centuries! The association between sore throat and rheumatic fever was not made until 1880.
- In 1944, the Jones criteria were formulated to assist disease identification.
- Until 1960, it was a leading cause of death in children and a common cause of structural heart disease.

Introduction and Background

- Dramatic declines in the incidence of rheumatic fever are thought to be largely due to antibiotic treatment of streptococcal infection.

Pathophysiology

- RF is the result of a previous Group A beta Hemolytic streptococcus infection *of the URT*.
 - *Skin infections have not been linked to ARF .*
 - Certain beta-streptococcal serotype (eg, M types 3, 5, 18, 19, 24) is linked directly to acute rheumatic fever.
 - Non-group A streptococci has never been shown to cause this disease
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- It is an autoimmune response secondary to molecular mimicry following group A streptococcal pharyngitis.

RF a Systemic Disease

- It is characterized by a proliferative inflammatory lesion of the tissue, especially that of the heart,
joints,
blood vessels,
and subcutaneous tissue.

Epidemiology

The frequency of streptococcal infection depends on

- The virulence of the bacterial strain, and
- M protein subtypes determine the incidence of rheumatic fever in a population.
- As a result ;acute rheumatic fever occurs during the school-aged years when streptococcal pharyngitis is most prevalent.
- Similarly, prevalence is higher in the colder months when streptococcal pharyngitis is most likely .

Epidemiology

- Rheumatic heart diseases remains the most common cause of acquired HD in all age group.
- Mortality rate has decreased dramatically in the recent decades due to more aggressive antibiotic use

Epidemiology

- Race : more of crowding than race .
- Sex: No sex predilection exists, *but* mitral valve prolapse and Sydenham chorea occur more often in females .
- Age : any age group may be affected but **most cases are reported in persons aged 5-15 years.**

Clinical Picture

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- 1- Migratory polyarthritits 75 %
- 2- Carditis 50-60 %
- 3- Erythema marginatum <3 %
- 4- Subcutaneous nodules <1 %
- 5- Chorea 10-15 %

Carditis

- Approximately 60% of all patients with ARF will develop rheumatic heart disease.
- Characterized by pancarditis:
 - _ myocarditis
 - _ pericarditis
 - _ endocarditis

Varies in severity from mild to fulminant potentially fatal ...

Carditis

- Cardiac involvement is the major cause of long-term morbidity.
- ARF causes inflammation of **valvular** endocardium.
- **One or more valves** (*most commonly the mitral valve*) are involved .
- *left ventricular dilation and congestive heart failure*, occurs sometimes decades later.
- *Vegetations* may develop on damaged valves and become infected leading to endocarditis.

Carditis

- **Myocarditis** is usually present but is not the direct cause of heart failure.
- Patients without carditis during the initial episode have a relatively low risk of developing carditis during recurrences.
- *Those with carditis as part of the initial episode are at greater risk of developing recurrences and of sustaining further cardiac injury.*

Carditis

- Myocarditis and /or pericarditis without endocarditis is rarely due to ARF
- *Mitral Valve disease may be isolated or combined with Aortic Valve disease*
- Right side valvular disease (TV and PV) is uncommon
- Isolated Aortic valve disease is uncommon
- *Valvular insufficiency is characteristic (MR, AR)*
- *Valvular stenosis* needs several years or decades after acute illness to appear
- *Mitral Stenosis and Aortic Stenosis may develop sooner after ARF in younger children in developed countries.*

Carditis

- Other manifestations of carditis include pericardial effusion, pericarditis , and arrhythmia.

Migratory polyarthrititis

- Occurs early in the disease
- larger joints such as the knees, ankles, elbows, and wrists are involved most .
- Joints are red, warm and swollen,
- Migratory arthritis, not symmetric
- Pain is VERY SEVERE
- Does not result in chronic or destructive joint disease

Migratory polyarthrititis

- Disappears in 12-24 hr after anti-inflammatory treatment starts , if not given may last for week
- If no response to treatment suspect ARF dx ?
- An inverse relationship between the severity of arthritis and cardiac involvement

Sydenham Chorea

- Occurs in 10-15 % of patients
- Occurs much later than other manifestations (can be months)
- Can be the only symptom of ARF .
- Usually disappears in few weeks to months but recurrence has been reported .

Sydenham Chorea

- Frequently starts with subtle neurologic behavior disorder.
- Emotional lability
- Incoordination(handwriting)
- Poor school performance
- May be one limb or all 4 extremities

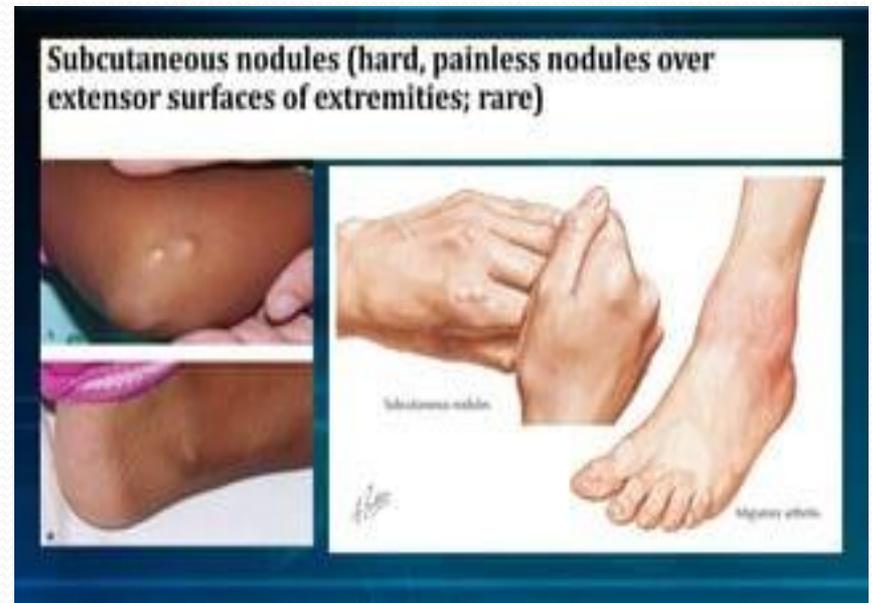
Erythema Marginatum

- **Rare** 3 % of pts
- *Characteristic rash of ARF*
- Erythematous macular lesion with Pale center
- Not pruritic
- Occurs primarily on trunk and extremities ,not on face
- Evanescent rash and can be accentuated by heat



Subcutaneous Nodules

- Very rare
- Mostly in patients with severe Carditis
- Found on the extensor surfaces of the joints (knee , elbow and spine)
- Firm non tender pea size nodules



Diagnosis

Diagnosis

Jones criteria was revised lately in 2015 by American Heart Association.

- Five major criteria
- Four minor criteria

plus

Evidence of preceding group a streptococcus infection

Diagnosis Jones criteria

Major Criteria

- Migratory polyarthrititis
- Carditis clinical or echo
- Erythema marginatum
- Subcutaneous nodules
- Chorea

- In 2015

Can be monoarthrititis in high risk area

Minor Criteria

- Fever
- polyarthralgia
- Elevated ESR /CRP
- Prolonged PR interval on ECG (in about 20 % of cases)

Evidence of Streptococcal infection

- Culture
- Serology
 - ASO titre (elevated in 80% of ARF pt)
 - Anti DNA ase
 - Antihyalurinidase

Differential Diagnoses

Aortic Regurgitation	Scarlet Fever
Atrial Fibrillation	Pericarditis, Acute
Endocarditis	Reactive Arthritis
Huntington Chorea	Rheumatoid Arthritis
Lyme Disease	Systemic Lupus Erythematosus
Mitral Regurgitation	Leukemia
Mitral Stenosis	
Myocarditis	
Kawasaki Disease	

Treatment

Treatment

Acute rheumatic fever (ARF) is usually preventable if antibiotics are initiated within 9 days of the onset of streptococcal infection!

Treatment

Medical therapy for acute rheumatic fever involves :

- Treat group A streptococcal infection.
- Steroids and salicylates in the control of pain and inflammation.
- Heart failure may require digitalis, fluid and sodium restriction, diuretics, and oxygen.
- Phenobarbital and haloperidol may be helpful in controlling chorea.

Treatment

Administer prophylaxis against GABHS infections to patients who have developed ARF.

- Most authorities suggest that prophylaxis be given for 5 years.
- For those who have rheumatic carditis, some authorities suggest lifelong prophylaxis.

Treatment of GABHS

- 10 days of penicillin PO or
- Single shot of 1,200,000 U of Benzathin Penicillin G

Treatment cont.

Patients with arthritis alone or / and mild carditis without myocarditis or heart failure :

- Aspirin, salicylates 100 mg/kg/day (3-5) days
 - then 75 mg/kg/day for 4 wk
 - other NSAIDs can be used and equally effective as salicylates.
- No need for steroids

Treatment cont.

pts with carditis and cardiomegaly:

- should be treated by corticosteroid
 - 1 – prednisone 2 mg/kg/day (Bid) for 2-3 wk
 - 2 – start tapering by 5 mg/24hr every 2-3 days
 - 3 – start salicylates (aspirin) 75 mg/kg/day at the beginning of tapering of prednisone and continue for another 3-4 wk after stopping steroids.
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- treat CHF (Digoxin, Lasix ,fluid restriction , oxygen)

Treatment cont

- Sydenham Chorea :

1 – anti inflammatory agents are usually not indicated

2 – Drugs

_phenobarbital po

is the drug of choice

_haloperidol divided bid po

_chlorpromazine po

Prophylaxis

- **Prophylaxis Antibiotics :**

Benzathine Penicillin G IM monthly shot (1200000 U)

Penicillin V PO 250 mg BID

Sulfadiazine PO 500-1000mg OD

Erythromycin PO 250 mg BID

Sulfadiazine should never be used for acute treatment of GABHS pharyngitis , but it is effective in eradication of colonization of the upper oral tract and is acceptable agent for prophylaxis .

Prophylaxis

- Duration of prophylaxis for ARF :
- No carditis :
prophylaxis for 5 yr or age 21 which ever comes later
- In significant Rheumatic heart disease
Prophylaxis for life (some recommend 10 yr or 40 th birthday)

Prognosis

- 70 % of pts with carditis recover with no residual heart disease
- Pts without carditis during the first attack are unlikely to have carditis with recurrence
- 20 % of pts with chorea can develop RHD within 20yr if they are not put on secondary preventive
- Arthritis and chorea resolve completely without sequelae
- Remember the prophylaxis for endocarditis in those with significant valvular disease



THANK YOU