Rheumatic Fever

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DEFINITION:

- Acute rheumatic fever is a systemic disease of childhood ,often recurrent that follows group A beta hemolytic streptococcal infection
- It is a diffuse inflammatory disease of connective tissue primarily involving heart, blood vessels, joints, subcut.tissue and CNS

Etiology

- Beta hemolytic streptococci.
- Genetic

Epidemiology

- Ages 5-15 yrs are most susceptible
- Rare <3 yrs
- Girls>boys
- Environmental factors--*over crowding, poor sanitation, poverty,*
- *Incidence more during fall ,winter & early spring*

Pathogenesis

- Delayed immune response to infection with group A beta hemolytic streptococci.
- After a latent period of 1-3 weeks, antibody induced immunological damage occur to *heart valves, joints, subcutaneous tissue & basal ganglia of brain.*

Pathologic Lesions

- Fibrinoid degeneration of connective tissue, inflammatory edema, inflammatory cell infiltration & proliferation of specific cells resulting in formation of **Ashcoff nodules,** resulting in-

  -***Pancarditis*** *in the heart*

  -***Arthritis*** *in the joints*

  -***Ashcoff nodules*** *in the subcutaneous tissue*

  -Basal gangliar lesions resulting in ***chorea***

Clinical Features

1.Arthritis

- Flitting & fleeting migratory polyarthritis, involving major joints
- Commonly involved joints-knee,ankle,elbow & wrist
- Occur in 80%,involved joints are exquisitely tender
- *In children below 5 yrs arthritis usually mild but carditis more prominent*
- *Arthritis do not progress to chronic disease*

Clinical Features (Contd)

2.Carditis

- Manifest as **pancarditis**(endocarditis, myocarditis and pericarditis),occur in 40-50% of cases
- *Carditis is the only manifestation of rheumatic fever that leaves a sequelae & permanent damage to the organ*
- *Valvulitis occur in acute phase*
- *Chronic phase- fibrosis,calcification & stenosis of heart valves(fishmouth valves)*

Clinical Features (Contd)

3.Sydenham Chorea

- Occur in 5-10% of cases
- Mainly in girls of 1-15 yrs age
- May appear even 6/12 after the attack of rheumatic fever
- Clinically manifest as-clumsiness, deterioration of handwriting, emotional lability or grimacing of face
- Clinical signs- pronator sign, jack in the box sign , milking sign of hands

Clinical Features (Contd)

4.Erythema Marginatum

- Occur in <5%.
- Unique,transient,serpiginous-looking lesions of 1-2 inches in size
- Pale center with red irregular margin
- More on trunks & limbs & non-itchy
- Worsens with application of heat

Clinical Features (Contd)

5.Subcutaneous nodules

- Occur in 10%
- Painless, pea-sized, palpable nodules
- Mainly over extensor surfaces of joints,spine,scapulae & scalp
- Associated with strong seropositivity

Clinical Features (Contd)

Other features (Minor features)

- Fever-(upto 101 degree F)
- Arthralgia
- Pallor
- Anorexia
- Loss of weight

Laboratory Findings

- High ESR
- Anemia, leucocytosis
- Elevated C-reactive protien
- ASO titre >200 Todd units.
  (Peak value attained at 3 weeks,then comes down to normal by 6 weeks)
- Throat culture-BHstreptococci

Laboratory Findings (Contd)

- ECG- prolonged PR interval, 2nd or 3rd degree blocks,ST depression, T inversion
- 2D Echo cardiography- valve edema,mitral regurgitation, LA & LV dilatation, and decreased contractility

Diagnosis

- Rheumatic fever is mainly a clinical diagnosis
- *No single diagnostic sign or specific laboratory test available for diagnosis*
- Diagnosis based on **MODIFIED JONES CRITERIA**

Jones Criteria (Revised) for Guidance in the Diagnosis of Rheumatic Fever*

Major Manifestation	Minor Manifestations		Supporting Evidence of Streptococal Infection
	Clinical	Laboratory	
Carditis Polyarthritis Chorea Erythema Marginatum Subcutaneous Nodules	Previous rheumatic fever or rheumatic heart disease Arthralgia Fever	Acute phase reactants: Erythrocyte sedimentation rate, C-reactive protein, leukocytosis Prolonged P-R interval	Increased Titer of Anti-Streptococcal Antibodies ASO (anti-streptolysin O), others Positive Throat Culture for Group A Streptococcus Recent Scarlet Fever

*The presence of two major criteria, or of one major and two minor criteria, indicates a high probability of acute rheumatic fever, if supported by evidence of Group A streptococcal nfection.

Recommendations of the American Heart Association

Treatment

- **Step I** - primary prevention (eradication of streptococci)
- **Step II** - anti inflammatory treatment (aspirin,steroids)
- **Step III**- supportive management & management of complications
- **Step IV**- secondary prevention (prevention of recurrent attacks)

STEP I: Primary Prevention of Rheumatic Fever

(Treatment of Streptococcal Tonsillopharyngitis)

Agent	Dose	Mode	Duration
Benzathine penicillin G	600 000 U for patients 27 kg (60 lb) 1 200 000 U for patients >27 kg	Intramuscular	Once
	or		
Penicillin V (phenoxymethyl penicillin)	Children: 250 mg 2-3 times daily Adolescents and adults: 500 mg 2-3 times daily	Oral	10 d
For individuals allergic to penicillin			
Erythromycin: Estolate	20-40 mg/kg/d 2-4 times daily (maximum 1 g/d)	Oral	10 d
	or		
Ethylsuccinate	40 mg/kg/d 2-4 times daily	Oral	10 d

Recommendations of American Heart Association

Step II: Anti inflammatory treatment

Clinical condition	Drugs
Arthritis only	**Aspirin** 75-100 mg/kg/day,give as 4 divided doses for 6 weeks (Attain a blood level 20-30 mg/dl)
Carditis	**Prednisolone** 2-2.5 mg/kg/day, give as two divided doses for 2 weeks Taper over 2 weeks & while tapering add Aspirin 75 mg/kg/day for 2 weeks. Continue aspirin alone 100 mg/kg/day for another 4 weeks

3.Step III: Supportive management & management of complications

- Bed rest
- Treatment of congestive cardiac failure:
  -digitalis,diuretics
- Treatment of chorea:
  -diazepam or haloperidol
- Rest to joints & supportive splinting

STEP IV : Secondary Prevention of Rheumatic Fever (Prevention of Recurrent Attacks)

Agent	Dose	Mode
Benzathine penicillin G	1 200 000 U every 4 weeks*	Intramuscular
or		
Penicillin V	250 mg twice daily	Oral
or		
Sulfadiazine	0.5 g once daily for patients 27 kg (60 lb 1.0 g once daily for patients >27 kg (60 lb)	Oral

For individuals allergic to penicillin and sulfadiazine

Erythromycin	250 mg twice daily	Oral

*In high-risk situations, administration every 3 weeks is justified and recommended

Recommendations of American Heart Association

Duration of Secondary Rheumatic Fever Prophylaxis

Category	Duration
Rheumatic fever with carditis and residual heart disease until (persistent valvar disease*)	At least 10 y since last episode and at least age 40 y, sometimes lifelong prophylaxis
Rheumatic fever with carditis but no residual heart disease (no valvar disease*)	10 y or well into adulthood, whichever is longer
Rheumatic fever without carditis	5 y or until age 21 y, whichever is longer

*Clinical or echocardiographic evidence.

Recommendations of American Heart Association

Prognosis

- Rheumatic fever can recur whenever the individual experience new BH streptococcal infection,if not on prophylactic medicines
- Good prognosis for older age group & if no carditis during the initial attack
- Bad prognosis for younger children & those with carditis with valvar lesions

THANK YOU