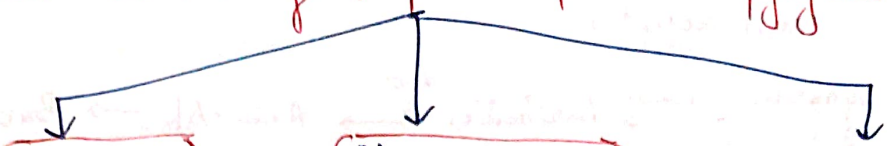
Acute Rheumatic Fever

Diseases caused by Group A β-Hemolytic Streptococcus pyogenes :- → sensitive to bacitracin
→ PYR test +ve.



- Pharyngitis
- Tonsilitis
- Quinsy
- Ludwig's angina
- Impetigo
- Cellulitis
- NF
- TSS
- Scarlet fever

↳ Rheumatic fever : caused by rheumatogenic M types.
→ preceded by sore throat
↳ Glomerulonephritis (GN) caused by nephritogenic M types
→ preceded by sore throat / skin infection

Incidence : M=F

Etiopathogenesis of Rheumatic Fever :-

Immune mediated multisystem disorder characterised by non-suppurative inflammation (joints, heart, skin, s/c, CNS) following an untreated streptococcal throat infection (2-4 weeks)

↓

Inflammation resolves spontaneously (few weeks to months) : no residual damage.
Except : Carditis (valvulitis) → RHD.

* Risk factors :-

① Untreated streptococcus pharyngitis d/t rheumatogenic M types : $M_1, M_3, M_5, M_6, M_{14}, M_{18}, M_{19}, M_{24}$
② Age : 5-15 years
③ Poverty (overcrowding)
④ Resides h/o RF
⑧ Genetic factors : HLA types : HLA DR53
HLA DR7
Alleles of : Toll like receptor
Mannose binding lectin, TNF-α

Pathogenesis: Theories

I) Molecular mimicry : most accepted

Certain Ag of S. pyogenes resemble human Ag → induce Antibodies → Act as → Auto-Ab → Bind to human Ag ↓ Immune complexes (Type II HSN) → Inflammation

- Cross Reactivity b/w Group A carbohydrate cell wall of S. pyogenes c̄ cardiac valves.

II) Tissue injury: d/t toxins of S. pyogenes (SLO & SLS)

III) M protein - Binds to human Ag (collagen type IV)
↓
alters into foreign Ag → induce Ab & form complexes

C/F :- ① Joints: m/c manifestation in ~~75%~~ → foreign patient (earliest) early finding (30-80%) children

- Asymm. migratory polyarthritis - affected joints: knees, ankles, elbows, hip, wrist.

→ m/c feature seen in 90% early within 2 wks most serious

Polyarthralgia / mono

Excellent / dramatic response to Aspirin

② Pan-Carditis :- 1) Pericarditis - fibrinous exudate in pericardial sac (bread & butter pericarditis)

Pt will c/o sharp chest pain which relieves on bending forward.

O/E: Pericardial Rub ⊕

Echo: Pericardial effusion & Cardiomegaly

2) Myocarditis : Characterised by (Aschoff Bodies)
↓
Conduction defects, pt. may present c̄ arrhythmias hypomotility & ↓ pumping function → CHF

child may present c̄ palpitation, dyspnea, orthopnea

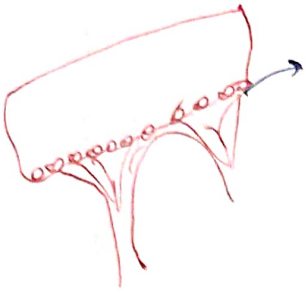
→ Perivascular granuloma : lymphocytes, neutrophils & anitschkow cells / caterpillar cells (condensed chromatin)

→ multiple anitschkow cells → form → multinucleated giant cells (Aschoff giant cells)

⑤ Endocarditis :- 40 % valvulitis heals completely

m/c valve affected : mitral (F>m) > Mitral + Aortic (M>F)

ARF : Aleu regurgitation



small wavy vegetations

Rest thickening of valves
Shortening of chordae
fusion of commissures.
↓
Stenosis

CAREY COOMBS MURMUR
Delayed diastolic murmur
Mitral Regurgitation : Soft SI
Pan-systolic murmur
↓
hemodynamic overload
↓
LVF
(morbidity / mortality in ARF)

RHD (stenosis) → CHF, embolism, IE

② Subcutaneous nodules :- mainly in severe carditis
(late) 6wks after onset
seen in 3 - 20 % cases
firm & painless nodules (0.5 - 2mm in size)
bony prominences & extensor aspect of limbs.
elbow, shin, occiput particle

④ Erythema marginatum :- 10 % of patients
(Rare finding)
Early
Pink macules → painless, non-pruritic annular lesions with pale centres & erythematous advancing serpiginous margins (= snake rings)
induced by warmth / heat
- Evanescent
- Limbs & trunk (start)

⑤ Sydenham's Chorea :- St. Vitus dance
15 - 25 % patients, longest latency (last manifestation - 3 months after)
- Rapid jerky, semi-purposeful, uncontrollable movement of hand, feet, face
- Disappear in sleep
- ↓ by sedation
- Emotional lability & muscle weakness.

Abnormal coordination & posture
Alc - hypotonia, emotional lability, jerky speech
Signs :- ① Milkmaid grip sign - Alternate tightening & loosening of grip
② Darting tongue : Uncontrollable writhing in & out of tongue on protrusion

Self-resolving. (2-6 wks)
Rx : Diazepam, Haloperidol.

DIAGNOSIS

MODIFIED JONES CRITERIA : AHA 2015.

Low Risk Population : Incidence of RF ≤ 2/1lakh school going children
Prevalence of RHD ≤ 1/1000 population year

High-risk population

Major Criteria :-	Low Risk	High Risk
Joints	Polyarthritis	Poly / Mono / Polyarthalgia
Carditis	Clinical / Sub-clinical (ECHO)	"
SN	✓	✓
EM	✓	✓
SC	✓	✓

Minor Criteria:		
Arthalgia	Polyarthalgia	≠ monoarthalgia
Fever	≥ 38.5°C	≥ 38°C
Inflam. markers	ESR ≥ 60mm/hr	≥ 30 mm/hr
Prolonged ~~PR~~ PR intv.	CRP ≥ 3mg/dL	"
	✓	✓

Diagnosis of RF :-
Essential Criteria
Prior strept. infect + 2 major or 1 Major + 2 minor

- Throat swab culture +ve
- " " RAT +ve
- Significant ↑↑ ASO

Relapse : 3 minor / 2 major / 2 maj + 1 minor

Treatment :- ① Bed Rest for 2 wks
IM Benzathine penicillin < 27kg → 6 lakh units (or)
>27kg → 1.2 million units
Oral Penicillin V : 250mg QID × 10 days
If allergic to penicillin → Erythromycin 250mg × 10 days.
Mild arthritis / algia → Acetaminophen

Mod - Severe Arthritis ± Carditis (NO CHF) → Aspirin (3-6wks)

Carditis ± CHF ± Arthritis → Steroids x 3weeks

① Oral Prednisolone 2mg/kg/day for 3weeks → Taper over 6weeks

↓

taper steroids x 3weeks

+ Start Aspirin x 1month

↳ 90-120mg/kg/day in 4 divided doses x 10weeks.

↳ taper over 2 weeks.

CHF → SnRx.

Prophylaxis :- IM. Benzathine Penicillin to prevent Strep. Pharyngitis → 3weekly.

Duration: depends on severity.

RF ± no carditis : 5yrs / till 21yrs (whichever is ~~longer~~ later)

RF ± carditis (no RHD) : 10yrs (" ")

RF ± " + RHD : ~~10yrs~~ (till 40yrs) / lifelong

↑ till age 25