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Rheumatic Heart Disease

★ Objectives:

1. Know that RHD is prevalent in our region, and economic burden
2. Know how to diagnose the disease and how to approach a patient with RHD
3. Know the principles of management
4. Know how to prevent RHD; Who needs prophylaxis and for how long
5. Recognize complications and how to manage it

★ Resources Used in This lecture:

Slides, Kumar, Class notes, 433 teamwork.

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Epidemiologic Background

- Globally rheumatic heart disease is the commonest CVD in young people 25 yrs old
- The overall incidence of ARF from 5-51 per 100000 population with a mean of 19 per 100000 population
- In children 5-14 yrs old 0.8-5.7 per 1000 children with a median of 1.3 per 1000
- The incidence of RF and the prevalence of RHD has declined substantially in developed nations.
- This decline has been attributed to:
  - ✓ Improved hygiene.
  - ✓ Reduced household crowding.
  - ✓ Improved medical care.
- A disease of poverty[1] and low socioeconomic status
- In underdeveloped countries RHD is the leading cause of CV death during the first five decades of life.

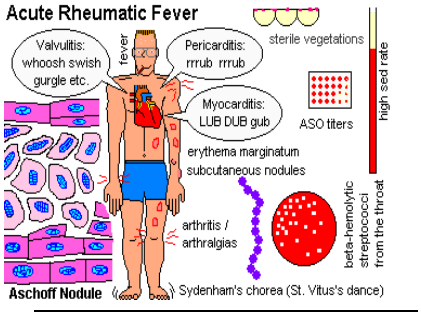
Global Burden of RHD

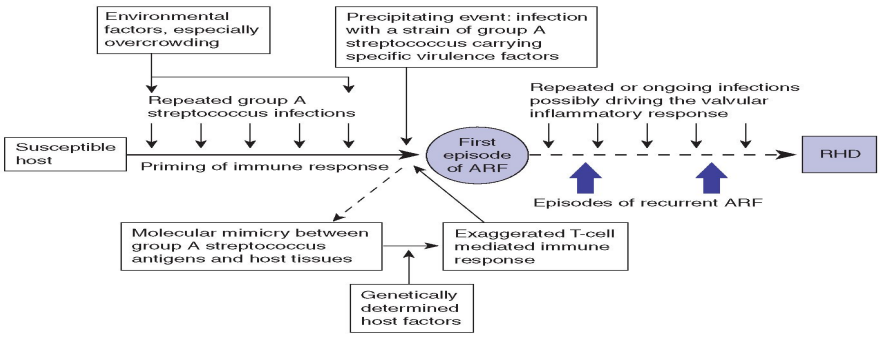
- Total cases with RHD: 20 Millions
- 3 Million have **CHF**.
- 1 Million require **valve surgery**.
- Annual incidence of RF: 0.5 Million, nearly half develop **carditis**
- Estimated **deaths** from RHD: 230,000 per year.
  - Imposes a substantial burden on healthcare systems with limited budgets

ARF VS RHD :

✓ **Acute Rheumatic Fever (ARF):** Is a systemic inflammatory disorder that occurs in **children between 5 and 15 years** of age as a result of **group A beta hemolytic streptococcal throat infection** "Pharynx is the most common site for infection leading to RF". It represents a **delayed immune response** to infection with manifestations appearing after a period of **2-4 weeks.** It affect heart, skin, joints and CNS. the condition is **not due to direct infection of the heart or to the production of a toxin.** (i.e. you won't find bacteria or causative antigen)

✓ **Rheumatic Heart Disease (RHD):** is a long term complication of acute rheumatic fever, that develope later in life (after 10-20 years) with a major effect on health due to damage to heart valves predominantly affecting the mitral and aortic valves.





[1] شد

Pathophysiology of Acute Rheumatic Fever *(from Rapid Review Pathology)*

1. Antibody-mediated disease that follows a group A streptococcal infection of the pharynx.
2. Host develops antibodies against group A streptococcal M proteins.
3. Antibodies that are produced cross-react with similar proteins in human tissue (called mimicry).

Clinical Presentation

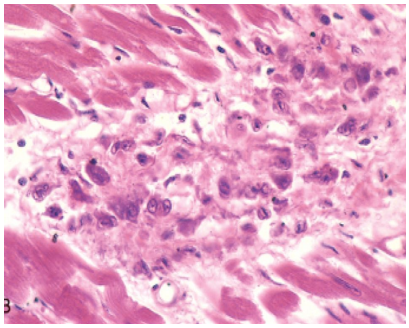
The disease presents suddenly with fever, joints pain, malaise and loss of appetite, **2–4 weeks after an episode of streptococcal pharyngitis.** The clinical features depends on the organs that are involved.

★ Arthritis

- Common, present in 35-66%
- Earliest manifestation of ARF
- Migrating, Fleeting polyarthritis affecting Large joints, such as the knees, ankles, shoulders, elbows and wrists.
- The joints are swollen, red and tener.
- Duration short < 1 week
- Rapid improvement with **salicylates** i.e Aspirin
- Once the acute inflammation disappears, the rheumatic process leaves the joints normal. **Does not progress to chronic disease.**

★ Carditis:

- Occurs in 50-70% of cases
- Only manifestation of ARF that **leaves permanent damage**
- May be subclinical
- Murmurs of MR or AR may occur in **acute stage** while **mitral stenosis occurs in late stages.** i.e with recurrent attacks.
- Transient diastolic mitral (Carey-coombs) murmur due to mitral valvulitis.
- Cardiomegaly and CHF may occur
- Appearance of pericardial effusion & ECG changes of pericarditis (raised ST segments) or myocarditis (inverted or flattened T wave), first-degree or greater AV block or other cardiac arrhythmia.
- Aschoff bodies lesions have a central area of fibrinoid necrosis surrounded by Anitschkow cells (reactive histiocytes).
- Aschoff bodies don't contain any bacteria or virus because it's an immune response.

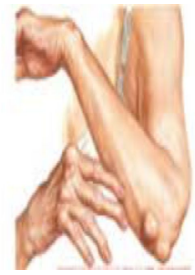


More than 50% of those who suffer from acute rheumatic fever with carditis will later (after 10-20 years) develop **Chronic rheumatic valvular disease.**

★ Sydenham Chorea:

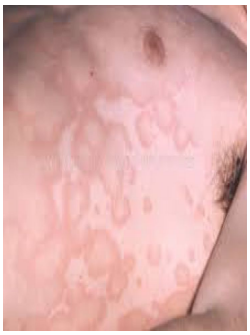
- Also known as **Saint Vitus'dance**.
- Occur in 10-30%, *Female* predominance.
- Extrapyramidal manifestation due to involvement of **basal ganglia**.
- Abrupt **purposeless involuntary movements** of muscles of face, neck, trunk, and limbs.
- *Delayed* manifestation of ARF "appear even 6 months after the attack of RF"
- Clinically manifest as: Clumsiness, deterioration of handwriting, emotional lability or grimacing of face and speech is often affected.

★ Subcutaneous Nodules



- Occur in 10%.
- The nodule is usually 0.5 – 2 cm long.
- Firm nontender "*painless*" hard nodules *under the skin.*
- Occur over extensor surfaces of joints, on bony prominences, tendons, spine
- Short lived: last for few days
- Associated with **severe carditis.**

★ Erythema marginatum



- Present in <6%.
- Less common, but **highly specific** manifestation of ARF.
- Reddish border, pale center, round or irregular serpiginous borders, non-pruritic, transient rash.
- Mostly on the trunk, abdomen and proximal limbs coalesce into crescent or ring-shaped patches.
- Associated with carditis.

Diagnosis of ARF No single test to diagnose ARF

-Accurate diagnosis is important

- **Overdiagnosis** will result in individuals receiving treatment unnecessarily.
- **Underdiagnosis** may lead to further episodes of ARF causing damage, and the need for valve surgery, and or premature death**.**

Diagnosis is **primarily clinical** and is based on a constellation of signs and symptoms, which were initially established as the .

Jones Criteria: In 1944 Dr. TD Jones published a set of guidelines for diagnosis of ARF "Jones Criteria" that has been revised recently -2015 by AHA

A firm diagnosis requires both of the following

1) **2 Major** manifestations or **1 Major and 2 Minor** manifestation of Jones Criteria

2) Evidence of a recent streptococcal infection:

➔ Increased or rising **ASO titer** or **Anti-Dnase B titer**

➔ A positive throat culture

However, when chorea or carditis is clearly present, evidence of an antecedent group A streptococcal infection is not necessary.

★ 1992 Modified Jones Criteria:

Major criteria	Minor criteria
Migratory polyarthritis	Arthralgia
Carditis	Fever
Erythema marginatum	First degree heart block
Syndenham chorea	Elevated inflammatory markers (ESR, CRP)
Subcutaneous nodules	

★ 2015 Revised Jones Criteria:

A. For all patient populations with evidence of preceding GAS infection	
Diagnosis: initial ARF	2 Major manifestations or 1 major plus 2 minor manifestations
Diagnosis: recurrent ARF	2 Major or 1 major and 2 minor or 3 minor
B. Major criteria	
Low-risk populations*	Moderate- and high-risk populations
Carditis† • Clinical and/or subclinical	Carditis • Clinical and/or subclinical
Arthritis • Polyarthritis only	Arthritis • Monoarthritis or polyarthritis • Polyarthralgia‡
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
C. Minor criteria	
Low-risk populations*	Moderate- and high-risk populations
Polyarthralgia	Monoarthralgia
Fever (≥38.5°C)	Fever (≥38°C)
ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL§	ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL§
Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

1. In accordance with the degree of **prevalence** of ARF/RHD in the population:

- **low risk populations**:

defined as those with ARF incidence < 2:100000 school-age children or all age prevalence of RHD of < 1:1000 population per year

- **Moderate or high risk:**

Children not from low risk population have been considered to be at.

2. Advocated the use of Echocardiography in all cases of confirmed or suspected ARF or RHD, to diagnose valvulitis (subclinical carditis) and has been included as a major criterion to diagnose carditis.

3. Aseptic monoarthritis has been included as a major criteria in moderate or high risk population.

4. Polyarthralgia has been recognized as a major manifestation for moderate or high risk population

5. Fever >38.5c, ESR >60 and or CRP > 3 mg/dl for low risk population.
and fever >38 and ESR >30 and or CRP > 3 mg/dl for moderate or high risk population

Investigations

Recommended for all cases
White blood cell count
Erythrocyte sedimentation rate (ESR)
C-reactive protein (CRP)
Blood cultures, if febrile
Electrocardiogram (if prolonged P-R interval or other rhythm abnormality, repeat in 2 weeks and again at 2 months, if still abnormal
Chest X-ray, if clinical or echocardiographic evidence of carditis
Echocardiogram (consider repeating after 1 month, if negative)
Throat swab (preferably before giving antibiotics): culture for group A streptococcus
Antistreptococcal serology: both ASO and anti-DNase B titres, if available (repeat 10–14 days later if first test not confirmatory)

All patients with suspected or confirmed ARF should undergo **Echocardiography** to confirm or refute the diagnosis of rheumatic carditis

Treatment of ARF

- **Bed rest**
- **Treat the streptococcal infection** by antibiotics
    - Penicillin: To avoid recurrent RF and damage the heart.
- **Treat the ARF:**

Salicylates: Aspirin

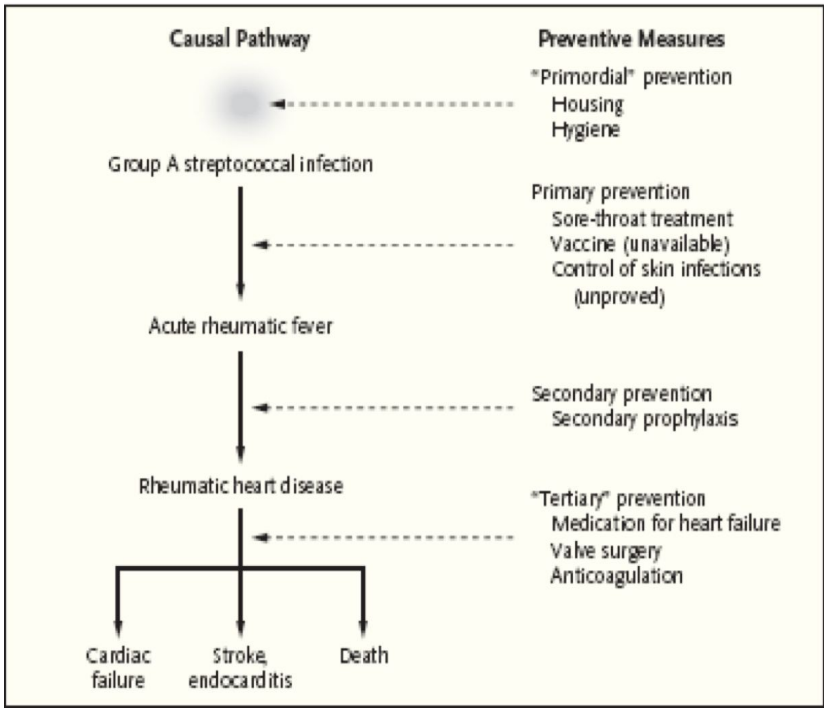
- 75-100 mg /kg/day given as 4 divided doses for 6 -8 weeks
- Attain a blood level 20-30 mg/d
- Relieves arthritis rapidly and response within 24 hours helps confirm diagnosis.

Prednisolone "corticosteroids"

- 2mg/kg/day taper over 6 weeks, **Given when there is severe carditis.**

- **Treat the complications:**
    - Heart Failure = *Diuretics, ACEI*
    - Symptoms develop or LV dysfunction (valvular problems) = *Valve replacement.*

Prevention of RF



1) Primordial Prevention:
Social; housing, hygiene, overcrowding

2) Primary Prevention:
Treatment of Sore Throat.

3) Secondary Prevention:
Monthly Penicillin

4) Tertiary Prophylaxis:
Medications, Balloon Valvuloplasty, Valve Replacement.

Secondary Prevention of Rheumatic Fever (Prevention of Recurrent Attacks)

Agent	Dose	Mode
Benzathine penicillin G Or	1 200 000 U every 4 weeks*	Intramuscular
Penicillin V Or	250 mg twice daily	Oral
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.

Duration of Secondary Rheumatic Fever Prophylaxis

Determined by:

1) The duration since the last episode of ARF (recurrences become less likely with increasing time)
2) Age (recurrences become less likely with increasing age)
3) Severity of RHD

- Should be continued for at least 5 years after initial attack (without carditis) or for 10 years. (with carditis)
- Should be given till age 40 years, sometimes life long for high risk patients or those frequently exposed to Strept infection

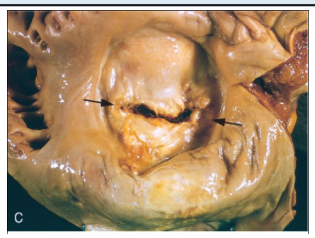
Rheumatic fever + carditis + residual valvular heart disease	10 years since last episode or until age 40 Sometimes lifelong prophylaxis
Rheumatic fever + carditis - no residual valvular heart disease	10 years or until age 21
Rheumatic fever - without Carditis	5 years or until age 21

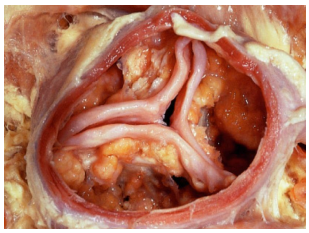
✓ Rheumatic Heart Disease

- Most commonly in Mitral-70%
- Frequently in Aortic-40%
- Less frequently Tricuspid-10%
- Rarely pulmonary valve-2%

Mitral Stenosis is more common in females(3:1), while males have higher incidence of Aortic Regurgitation

TYPE	Mitral Stenosis	Mitral Regurgitation	Aortic Stenosis	Aortic Regurgitation
Patho-Physiology	Cause ↑ LA pressure > Pulmonary congestions.	• **Acute**: Sudden pressure ↑ in normal LA Pulmonary congestion. • **Chronic:** Gradual pressure ↑ in a dilated LA > Pulmonary HTN.	Obstruction to LV outflow > LV hypertrophy > CO fails to ↑.	Inadequate closure of AV > ↑ LVEDV > LV dilation and hypertrophy > ↑ Pulmonary pressure.
Clinical Presentations (Significant)	• Hoarseness (Ortner's syndrome) • Dysphagia • Opening snap • Load S1	• Dyspnea • Orthopnea • PND • Soft S1, • Pansystolic murmur	• Angina • Syncope • Parvus et darus • Harsh crescedno-decresendo (systolic murmur) • Thrill • S4 • Paradoxical splitting of S2	• Wide pulse pressure • Water-hammer pulse • Austin Flint murmur
Management	Balloon Valvuloplasty	Afterload reduction / Surgical	Aortic valve Replacement Transcathter Aortic Valve Replacemen	Diuretics





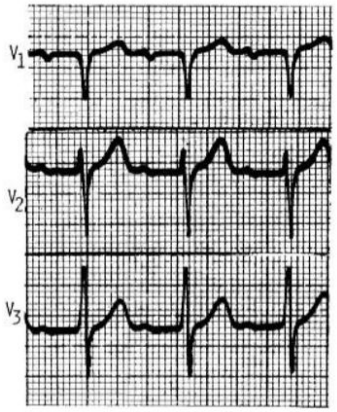
MCQs:

1. A 72-year-old man comes to the office with intermittent symptoms of dyspnea on exertion, palpitations, and cough occasionally productive of blood. On cardiac auscultation, a low-pitched diastolic rumbling murmur is faintly heard at the apex. What is the most likely cause of the murmur?

A. Rheumatic fever as a youth
B. Long-standing hypertension
C. A silent MI within the past year
D. A congenital anomaly
E. Anemia from chronic blood loss

2. An 18-year-old man complains of fever and transient pain in both knees and elbows. The right knee was red and swollen for 1 day during the week prior to presentation. On physical examination, the patient has a low-grade fever. He has a III/VI, high-pitched, apical systolic murmur with radiation to the axilla, as well as a soft, mid-diastolic murmur heard at the base. A tender nodule is palpated over an extensor tendon of the hand. There are pink erythematous lesions over the abdomen, some with centralclearing. The following laboratory values are obtained:

Hct: 42% WBC: 12,000/µL with 80% polymorphonuclear leukocytes, 20% lymphocytes ESR: 60 mm/h The patient's ECG is shown below. Which of the following tests is most critical to diagnosis?



A. Blood cultures
B. Antistreptolysin O antibody
C. Echocardiogram
D. Antinuclear antibodies
E. Creatine kinase

3. A 19-year-old man presents with recent onset of breathlessness and sharp, central chest pain exacerbated by movement and coughing. His heart rate is 110 bpm. On auscultation there is a soft pansystolic murmur and a pericardial friction rub. Echocardiography demonstrates mitral regurgitation. Antistreptolysin O antibody titres (ASOT) are 500 U/mL (normal range < 200). What is the likeliest diagnosis?

A. Infective endocarditis
B. Viral myocarditis
C. Acute rheumatic fever
D. Viral pericarditis
E. Dressler's syndrome

4.In the patient from the previous question, which one of the following features would clinch the diagnosis of rheumatic fever?

A. Temperature > 38oC
B. Positive throat swab culture
C. Cardiac dilatation on echocardiography
D. First-degree block on ECG
E. Flitting polyarthritis

Answers:
1.A
2.B
3.C
4.E