MEDICINE

432 Team

6 Rheumatic Fever AND RHD

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COLOR GUIDE: • Females' Notes • Males' Notes • Important • Additional

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.

Rheumatic Fever from Davidson

Definition:

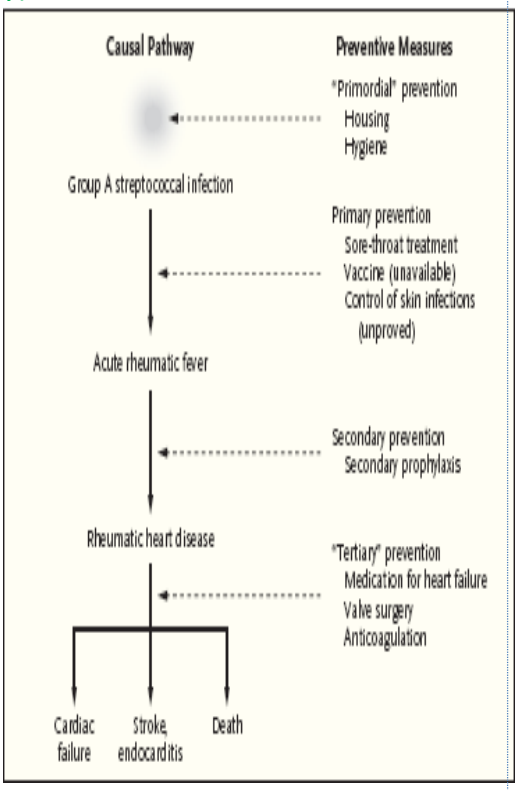
Acute rheumatic fever usually affects children (most commonly between 5 and 15 years) or young adults. The condition is triggered by an immune-mediated delayed response to infection with specific strains of group A beta hemolytic streptococci, which have antigens that may cross-react with cardiac myosin and sarcolemmal membrane protein. Antibodies produced against the streptococcal antigens cause inflammation in the endocardium, myocardium and pericardium, as well as the joints, skin and CNS *(it leaves permanent damage on the heart, while other organs develop only acute inflammation and then they heal normally)*

Note(s):

Studying Rheumatic fever is important because:
** Affects young people in their productive years and may cause permanent heart damage that affect quality of life.*

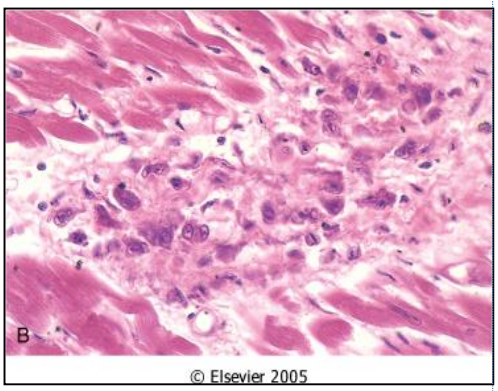
** To know how to prevent it because it is an expensive disease, require surgery, medicine, and tests and follow up.*

- A disease of poverty and low socioeconomic status
- Rare in wealthy countries, due to improved living conditions**, less overcrowding, and better hygiene with reduction in transmission of GABHS**
- Incidence: In Cambodia 2.2/1000 children, Mozambique 2.3/1000, India 0.75/1000
- Total cases with RHD:20 Millions
- CHF: 3 Million
- Valve surgery required in 1 Million
- Annual incidence of RF: 0.5 Million, nearly half develop carditis
- Estimated deaths from RHD: 230,000/YR
- Imposes a substantial burden on health care systems with limited budgets.



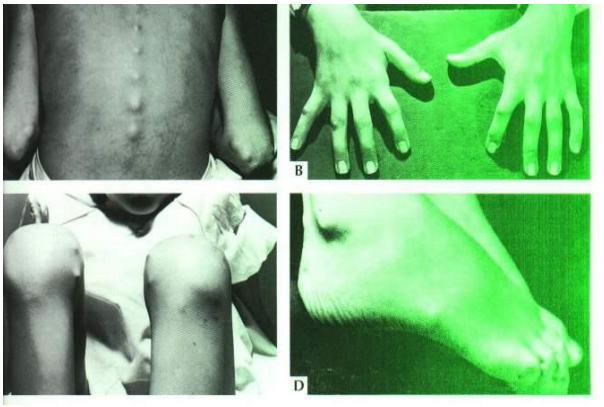
Pathologic Lesions

Histologically, fibrinoid degeneration is seen in the collagen of connective tissues.
Aschoff nodules are pathognomonic and occur only in the heart. They are composed of multinucleated giant cells surrounded by macrophages and T lymphocytes, and are not seen until the sub-acute or chronic phases of rheumatic carditis.



Clinical Features

Acute rheumatic fever is a multisystem disorder that usually presents with fever, anorexia, lethargy and joint pain, 2–4 weeks after an episode of streptococcal pharyngitis. There may, however, be no history of sore throat. Arthritis *(swelling and pain)* occurs in approximately 75% of patients (most common manifestation). Other features include rashes, carditis and neurological changes.



The diagnosis, made using the revised Jones criteria is based upon two or more major manifestations, or one major and two or more minor manifestations, along with evidence of preceding streptococcal infection. However, when chorea *(involuntary movements result from CNS involvement)* or carditis is clearly present and other causes have been excluded, a presumptive diagnosis can be made without evidence of preceding streptococcal infection.

*Diagnosis will be confirmed based on clinical examination **(jones criteria)** + laboratory tests.*

Major manifestations:	Minor manifestations: (in a recurrent attack, 2 minors are enough to confirm)
1. Carditis	1. Fever
2. Polyarthritis (red, swollen and tender)	2. Arthralgia (without swelling)
3. Chorea	3. Previous rheumatic fever
4. Erythema marginatum (skin rash)	4. Raised ESR or CRP
5. Subcutaneous nodules	5. Prolonged PR interval on ECG
	6. leukocytosis

Evidence of antecedent GABHS *(group A beta-hemolytic streptococci)*

1. Positive throat culture *(only 10-25% of cases are positive)* or rapid antigen test positive for group A beta-hemolytic streptococci (GABHS) (manifestations appear after the infection so there may be no bacteria left in the throat → the test is not that diagnostic)
2. ↑ Antistreptolysin O antibodies (ASO titres) *(indicates recent Strep.C Infection)*

Arthritis:

- Most common feature: present in 80%
- Earliest manifestation of ARF
- Major joints: The knees and ankles, shoulders, elbows, and wrists.
- "Migrating", "Fleeting" polyarthritis (Moving from one joint to another)
- Duration: usually short < 1 week (between a day and 4 weeks) RESPONSE QICKLY TO ASPRIN = so not juvenile rheumatic arthritis or not lupus.
- Responds well to Salicylates (Aspirin) no deformity no swan nick
- Does not progress to chronic disease

Note(s):

The difference between arthritis caused by rheumatic fever and other diseases that cause arthritis also is:

- **Migrating** (An arthritis that appears to migrate from one joint to the next)

- **Fleeting** (Passing quickly)

- **Respond** well to aspirin

- **Don't** lead to deformity

- **Affects** large joints

Sydenham Chorea: (St Vitus dance)

- Occur in 5-10% of cases in basal ganglia same as parkinsonism
- Abrupt Purposeless involuntary movements of muscles of face, neck, trunk, and limbs.
- May appear even 6 months after the attack of rheumatic fever, late manifestation.
- Clinically manifest as clumsiness, deterioration of handwriting, emotional liability or grimacing of face

Subcutaneous Nodules:

- Occur in 10%
- Usually Small (0.5 – 2 cm)
- Firm non-tender
- Short lived: last for few days.
- Associated with severe carditis
- Occur over extensor surfaces of joints, on bony prominences, tendons, spine.

Erythema Marginatum:

- Present in 5%
- Reddish border, pale center, round or irregular serpiginous borders, non-pruritic, transient rash
- Occurs on trunk, abdomen or proximal limbs
- Associated with carditis

Carditis:

- Occurs in 40- 50% of cases. Most serious, mainly sever
- Pancarditis (involves the endocardium, myocardium and pericardium) patient is in tachycardia (check the sleeping pulse, if high → positive sign), murmurs, HF, PR elongation, friction rub, febrile.
- Only manifestation of ARF that leaves permanent damage.
- Murmurs of MR or AR may occur in acute stage while mitral stenosis occurs in late stages
- Cardiomegaly and CHF may occur

Differential Diagnosis:

Juvenile rheumatoid arthritis - Infective endocarditis – Sickle-cell arthropathy - Lupus - Myocarditis - Reactive arthritis - Leukemia

Investigations:

1. **CBC, ESR, CRP**
2. **Anti-streptococcal antibodies: ASO titer**
3. **Throat swab for culture**
4. **ECG** *(for PR interval changes)*
5. **CXR** *(for cardiomegaly and HF)*
6. **ECHO** (the best to assess the heart damage. E.g. valvular)

Treatment of ARF:

Note(s):

•Side effects of aspirin: ulceration, gastritis, nausea, vomiting, bleeding, tinnitus, deafness.

- Salicylates: Aspirin 75-100 mg /kg/day, given as 4 divided doses for 6 -8 weeks, Attain a blood level 20-30 mg/dl. Relieves arthritis rapidly and a response within 24 hours helps confirm diagnosis.
- Prednisolone: 2mg/kg/day taper over 6 weeks, given when there is carditis or severe arthritis.
- Bed rest
- Treat heart failure if present
- Valve replacement later in life once symptoms develop or LV dysfunction occurs from severe valve regurgitation or valve stenosis

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(60lb)** **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**

Duration of Secondary Rheumatic Fever Prophylaxis:

- Rheumatic fever with carditis and residual heart disease (persistent valve disease):
  10 y since last episode or until age 40y, (whichever is longer), sometimes lifelong prophylaxis
- Rheumatic fever with carditis But no residual VHD :
  10 yrs or until age 21yrs (whichever is longer)
- Rheumatic fever without carditis
  5 y or until age 21 y, (whichever is longer)

Chronic Rheumatic Heart Disease:

Chronic valvular heart disease develops in at least half of those affected by rheumatic fever with carditis. Two-thirds of cases occur in women. Some episodes of rheumatic fever pass unrecognised and it is only possible to elicit a history of rheumatic fever or chorea in about half of all patients with chronic rheumatic heart disease.

The mitral valve is affected in more than 90% of cases; the aortic valve is the next most frequently affected, followed by the tricuspid and then the pulmonary valve. Isolated mitral stenosis accounts for about 25% of all cases of rheumatic heart disease, and an additional 40% have mixed mitral stenosis and regurgitation. Valve disease may be symptomatic during fulminant forms of acute rheumatic fever but may remain asymptomatic for many years.

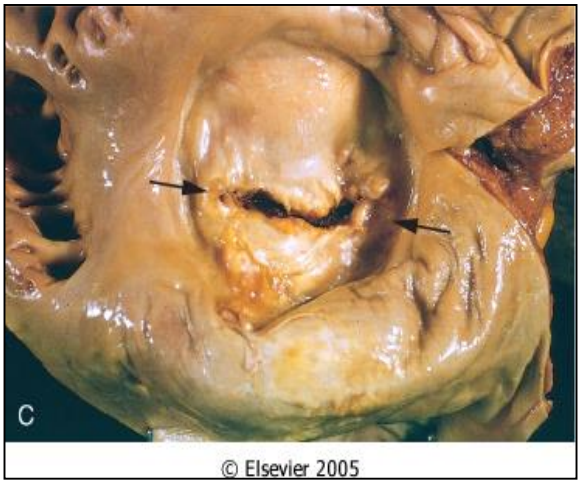


Figure: "fish-mouth"-shaped orifice

Mitral stenosis

- The normal MVA= 4-6 cm
- In severe MS <1
- High LAP
- The rise in LAP causes a similar rise in pulmonary capillaries, veins and artery

Note(s):

Mitral stenosis with diffuse fibrous thickening and distortion of the valve leaflets, commissural fusion (arrows), and thickening and shortening of the chordae tendineae. Marked dilation of the left atrium is noted in the left atrial view

- Long Asymptomatic period after initial attack of RF until onset of class 1 / 2 symptoms: 10 – 30 yrs (latent period)
- Once symptoms develop there is another plateau of 5 –10 yrs before onset of AF
- This followed by a period of 5-10 yrs until onset of class III- IV symptoms

18.5 New York Heart Association (NYHA) functional classification

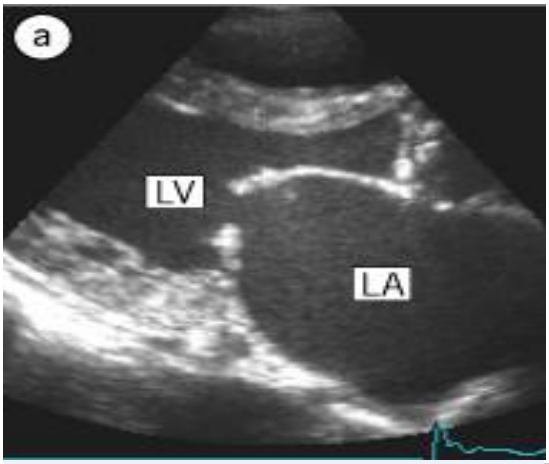
• Class I	No limitation during ordinary activity
• Class II	Slight limitation during ordinary activity
• Class III	Marked limitation of normal activities without symptoms at rest
• Class IV	Unable to undertake physical activity without symptoms; symptoms may be present at rest

Clinical Features:

- Dyspnea (pulmonary congestion)
- Fatigue (low cardiac output)
- Palpitation (atrial fibrillation)
- Hemoptysis (10%)
- Hoarseness: Ortner's syndrome (compression of left recurrent laryngeal nerve by enlarged LA)
- Dysphagia (compression of esophagus)
- Stroke or peripheral embolization
- Cyanosis (Mitral facies, malar flush)
- Tapping apex (S1)
- Parasternal heave
- Diastolic thrill
- Accentuated S1 , accentuated S2
- Opening snap
- Mid-diastolic rumble

Investigations:

- CXR: Straightening of the left heart border; Double density; Kerley B lines, CA in MV.
- ECG: LAE, P Mitrale, RV dominance.
- ECHO: MVA (mitral valve area), PAP (pulmonary artery pressure).



Note(s):

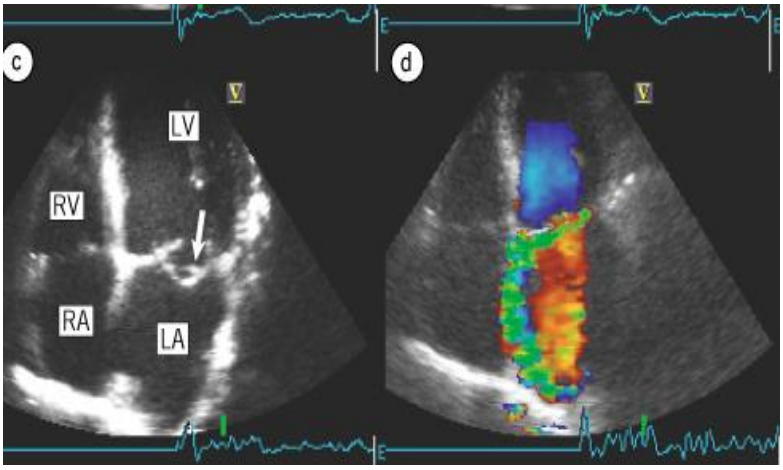
Two-dimensional echocardiography in mitral stenosis. (a) Parasternal long-axis view through a stenotic mitral valve, demonstrating typical diastolic doming of the anterior leaflet.

Management:

- B-Blockers ,calcium channel blockers (CCB)
- Digoxin (AF)
- Warfarin
- Balloon Valvuloplasty
- Mitral valve replacement

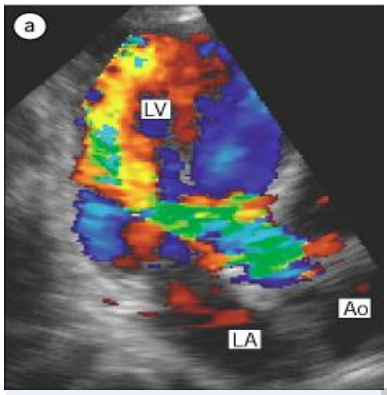
Mitral Regurgitation:

- Asymptomatic
- Dyspnea , orthopnea, PND
- Displaced PMI, Thrill
- Soft S1,
- Pansystolic murmur
- Treatment is surgical



Aortic Regurgitation:

- **Water-hammer / collapsing pulse.**
- **Wide pulse pressure.**
- **Corrigan's sign:** pulsation of the carotid arteries.
- **De Musset sign:** head bobbing.
- **Muller sign:** pulsation of the uvula.
- **Quincke's pulse:** pulsation of the fingernail capillaries.
- **Hill's sign:** BP findings may include popliteal systolic pressure ≥ 60 mm Hg higher than brachial pressure.



Note(s):

Severe aortic regurgitation. (a) Apical view showing a wide jet reaching to the apex

Aortic Stenosis

Symptoms: **1- Angina** **2- Syncope** **3- Dyspnea**

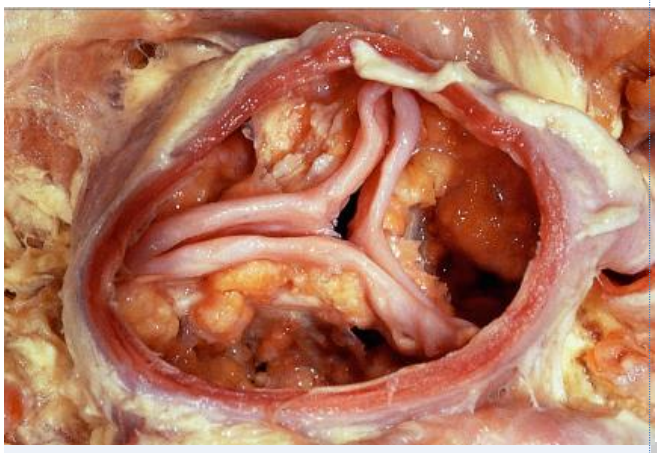
Signs:

- Arterial Pulse wave form : plateau

  Small [Parvus]

  Slow rise [Tardus]

- Sustained not displaced PMI
- Systolic thrill
- S4
- Late peaking of murmur
- Single S2 : Soft or absent A2
- Paradoxical splitting of S2



Note(s):

Fig. Gross pathology of degenerative aortic stenosis. *The figure shows the masses of lipo-calcification on the aortic side of all three leaflets and the absence of commissural fusion.*

Treatment is surgical: Aortic valve Replacement

SUMMARY

1. Rheumatic heart disease results from antistreptococcal antibodies that cross-react with cardiac tissues; it most commonly affects the mitral valve and is responsible for 99% of cases of acquired mitral stenosis.
2. Diagnosis is based on Jones criteria.
3. Treatment includes elimination of any residual streptococcal infection, bed rest, aspirin, and in selected cases corticosteroids, HF management, and surgery.
4. Secondary prevention of further attacks of rheumatic fever is crucial.
5. Antibiotic prophylaxis depends on whether carditis and/or residual damage is present.

EXTERNAL RESOURCES

References	Info
Davidson's Principles and Practices of Medicine, 22nd Edition	Most notes, percentages, and questions
Robbins Basic Pathology, 9th Edition	Summary and pathological descriptions
http://www.merckmanuals.com/professional/cardiovascular_disorders/valvular_disorders/aortic_regurgitation.html	Aortic Regurgitation signs

Questions

1) A 53-year-old lady complains of progressively worsening exertional dyspnoea associated with deterioration in exercise tolerance over the past 2 years. On examination her pulse is irregularly irregular and of small volume. There is a low-pitched mid-diastolic murmur audible at her apex. What is the likeliest cause of her breathlessness?
   a. **Hypertrophic cardiomyopathy**
   b. **Ischaemic heart disease**
   c. **Ventricular septal defect**
   d. **Mitral stenosis**
   e. **Aortic regurgitation**

2) 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

3) 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

4) Which one of the following features is likely to be found in mitral regurgitation but not in mitral stenosis?

a. Irregularly irregular pulse
b. Third heart sound (S3)
c. Right ventricular heave
d. Accentuated pulmonary component of second heart sound
e. Bi-basal crepitation

432 Medicine Team Leaders

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Answers:

1st question: d

2nd question: c

3rd question: e

4th question: b