L25: Rheumatic heart disease

Medicine433@yahoo.com

MEDICINE 433

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.

Definition

- **Acute rheumatic fever:** is an **immunologically mediated** response to infection **(usually pharyngitis)** by **group A beta hemolytic streptococci**, which **cross-react** with cardiac cells component and body will produce **antibodies** against it which cause inflammation.

- **Mostly affect children from 5-15 years old**
- **(it leaves permanent damage on the heart, while other organs develop only acute inflammation and then they heal normally)**

(Antibodies produced against the streptococcal antigens cause inflammation in the endocardium, myocardium and pericardium, as well as the joints and skin).

- **Rheumatic heart disease:** is the **chronic valvular abnormalities (Mitral stenosis most common)** secondary to acute rheumatic fever.

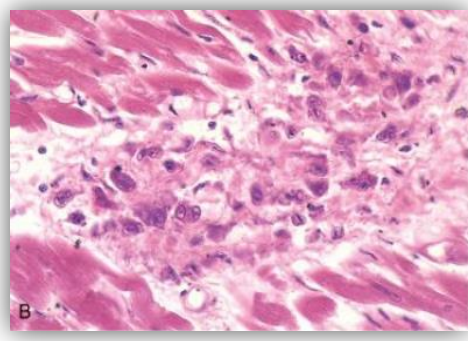
Definition

- A disease of poverty and low socioeconomic status
- Rare in wealthy countries, due to:

1. improved living conditions
2. less overcrowding
3. better hygiene with reduction in transmission of GABHS

❖ 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

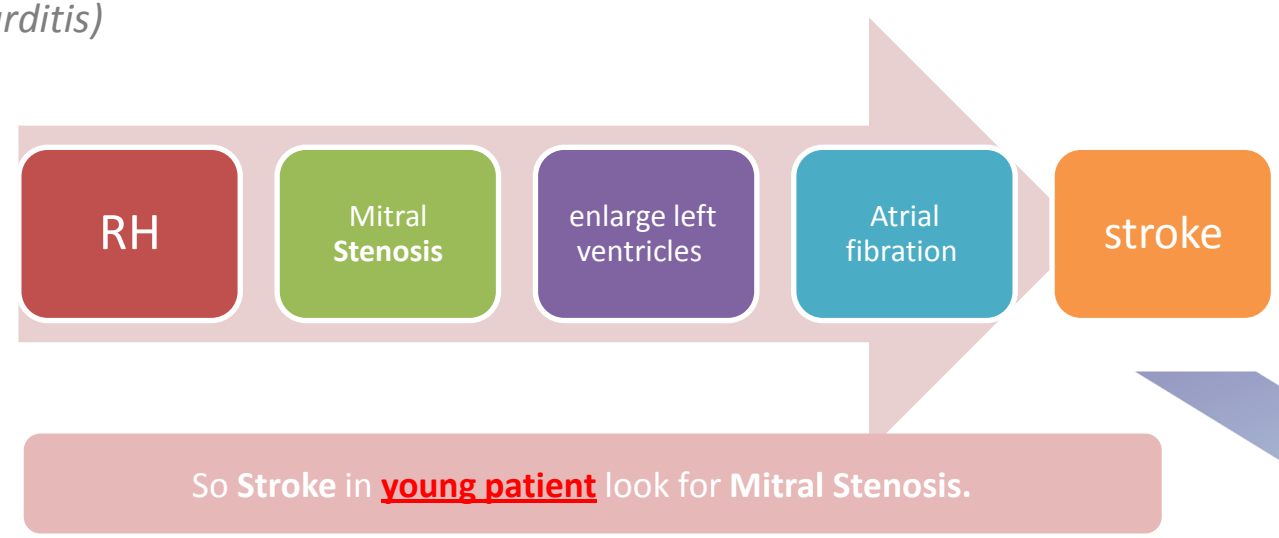
Pathophysiology



1.Fibrinoid degeneration is seen in the collagen of connective tissues.

2.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 subacute or chronic phases of rheumatic carditis)*



RH effect the heart permanently **(specially valves)** , it's the only manifestation that RH leave with a **permanently** damage.

Signs & Symptoms

Acute rheumatic fever is a multisystem disorder that usually presents with:

1. Fever, anorexia, lethargy and joint pain, **2–4 weeks after an episode of streptococcal pharyngitis**.

There may be no history of sore throat.

2. **Arthritis** (swelling and pain) occurs in approximately 75% of patients (most common manifestation).

3. Other features include rashes, carditis and neurological changes.

Investigation

From Daviedsn

Evidence of a systemic illness (non-specific) :

CBC 'Leucocytosis', raised ESR and CRP

Evidence of preceding streptococcal infection (specific) :

- Throat swab culture: group A β-haemolytic streptococci (also from family members and contacts) (positive Throat swab cultures are obtained in only 10-25% of cases)
- Antistreptolysin 0 antibodies (AS0 titres): rising titres, or levels of > 200 U (adults) or > 300 U (children)

Evidence of carditis :

- Chest X-ray: cardiomegaly; pulmonary congestion
- ECG: first- and rarely second-degree AV block; features of pericarditis, T-wave inversion; reduction in QRS voltages
- **Echocardiography**: cardiac dilatation and valve abnormalities (typically shows mitral regurgitation with dilatation of the mitral annulus and prolapse of the anterior mitral leaflet, and may also show aortic regurgitation and pericardial effusion)

Differential diagnosis:

- **Juvenile rheumatiod arthritis**
- **Infective endocarditis**
- **Sickle cell arthropathy**
- **Lupus**
- **Myocarditis**
- **Reactive arthritis**
- **Leukemia**

-ECG in RH – Look for P-R interval (its caused by complication **).**
-CXR in RH – enlargement of Heart Couse of **HF** & **edema**
-ECHO in RH – important (to look to the **valves**)

How to diagnose

A firm diagnosis requires both of the following:

1. Evidence of antecedent GABHS:

a) **Positive** throat culture **OR positive** rapid antigen test for GABHS

b) **Increase Antistreptolysin O antibodies** (ASO titres)

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

2. Using Jones criteria:

Two or more MAJOR OR One MAJOR and Two or more MINOR manifestations along with evidence of preceding streptococcal infection.

Major	Minor
1. Carditis 2. Polyarthritis (red, swollen and tender) 3. Sydenham Chorea 4. Erythema marginatum (skin rash) 5. Subcutaneous nodules 6. Chorea	1. Fever 2. Arthralgia (without swelling) 3. Previous rheumatic fever 4. Raised ESR or CRP 5. Prolonged PR interval on ECG 6. leukocytosis

CON. Jones criteria …

Plus: supporting evidence of preceding streptococcal infection. recent scarlet fever, raised antistreptolysin o or other streptococcal antibody titre, positive throat culture

N.B. Evidence of recent streptococcal infection is particularly important t there is only one major manifestation.

❖ **Arthritis: Most common and earliest** feature of ARG (Occur in 80%)

- **Acute painful asymmetric affect 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)
- **Responds well to Salicylates (Aspirin)**
- Does not progress to chronic disease, no deformity, no swan nick

❖ **Subcutaneous Nodules:** (Occur in 10%)

- **Usually Small (0.5 – 2 cm)**
- **Firm non-tender** painless
- 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 with pale center**, round or irregular
- serpiginous borders, **non-pruritic**, transient rash
- Occurs on trunk, abdomen or proximal limbs but not the face
- Associated with **Carditis**

Other manifestations are rare but includes pleurisy, pleural effusion and pneumonia

❖ **Carditis: (**Occurs in 40- 50% of cases) **Most serious**

- **Pancarditis** (involves the endocardium, myocardium and pericardium) patient is in
- tachycardia (check the sleeping pulse, if high → positive sign),breathlessness, palptations or chest pain (conductions defects sometimes occur and may cause syncope)
- Only manifestation of ARF that leaves **permanent damage.**
- Murmurs of **MR** or **AR** may occur in **acute stage** while **MS** occurs in **late stages**
- Cardiomegaly and CHF may occur

❖ **Sydenham Chorea:** late feature (Occur in 5-10% of cases)

- **In basal ganglia** same as parkinsonism
- May appear even 6 months after the attack of rheumatic fever
- **Purposeless involuntary movements** of muscles of face, neck, trunk, and limbs.
- Clinically manifest: Deterioration of handwriting, emotional liability or **grimacing of face**

Management

1. **Salicylates (Aspirin):** 75-100 mg /kg/day, given as 4 divided doses for 6 -8 weeks, Attain a blood level20-30 mg/dl. **Relieves arthritis rapidly and a response within 24 hours helps confirm diagnosis**(mild toxicity includes: nausea, tinnitus and deafness, more serious are : vomiting, tachypnea and acidosis)
2. **Prednisolone(corticosteroids):** 2mg/kg/day taper over 6 weeks**, given when there is carditis or severe arthritis** (produce more rapid symptomatic relief than aspirin)
3. Bed rest
4. Treat heart failure if present
5. **Valve replacement** later in life once symptoms develop or LV dysfunction occurs from severe valve regurgitation or valve stenosis

No specific treatment for **RH.**
-We give **RH** patient **Steroids** if there is a sever **cardaitis**
-To avoid **recurrent RF** and damege the heart of we give the Pathient long acting penicilin.

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:

Secendary patient (already have the disease) give him prophylaxis

- **Rheumatic fever with carditis and residual valvular heart disease** → 10 years since last episode or until age 40y, sometimes lifelong prophylaxis
- **Rheumatic fever with carditis But no residual VHD** → 10 years or until age 21
- **Rheumatic fever without Carditis** → 5 years or until age 21

Chronic Rheumatic Heart Disease

You better revise Valvular heart disease. Here is only summery of lecture:

The mitral valve is affected in more than 90% of cases; the aortic valve is the next most frequently involved, followed by the tricuspid and then the pulmonary valve.

Mitral stenosis: The normal MVA= 4-6 cm

- Most common cause is rheumatic fever and more common in women.
- Clinical symptoms and signs: Dyspnea, Orthopnea, PND, Hemoptysis, Systemic embolism "due to a fib", Hoarseness, dysphagia, Malar flush. Parasternal heave, Cyanosis
- Atrial fibrillation Loud S1, opening snap following S2, and mid-diastolic rumbling murmur.

- Management: diuretic and salt-restricted diet, digitalis and anticoagulant "in a fib", balloon valvulotomy "especially in pregnancy", mitral commissurotomy or valve replacement if balloon dilatation fails.

Fusion of the mitral valve commissures and shortening of the chordae tendineae may lead to mitral stenosis with or without regurgitation. Similar changes in the aortic and tricuspid valves produce distortion and rigidity of the cusps, leading to stenosis and regurgitation.

Mitral regurgitation:-

- Most common causes are ischemia and myxomatous.
- Clinical symptoms and signs: Dyspnea, Orthopnea, PND, Right-sided heart failure, atypical chest pain and palpitation.
- pansystolic murmur radiating to the axilla with thrill, mid-diastolic click , S3 gallop, distended neck veins in sever or acute MR.
- Management: after load reduction, mitral valve repair or replacement.

Aortic stenosis:-

- Calcific aortic valve is the most common cause; bicuspid valve is the most common congenital cause.

Clinical symptoms and signs: **angina, syncope, CHF**. Slow raising carotid pulse, radiate to the carotids. Systolic ejection murmur. Late peaking of murmur, Single S2 : Soft or absent A2, Paradoxical splitting of S2, Arterial Pulse wave form : plateau Small { Parvus } Slow rise { Tardus } , Sustained not displaced PMI, Systolic thrill, S4

- Diagnosis: cardiac cath is the definitive diagnostic; > 0.8 cm2 indicates severe stenosis.
- Management: valve replacement is the treatment of choice.

Aortic regurgitation:-

- Clinical symptoms and signs: angina, dyspnea. Mid-diastolic rumble at the apex, S3 and S4 gallop,
- 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.

- Management: **If the patient stable**: conservative like diurtics, salt restriction, and vasodilator. **Acute**: emergent valve replacement.

MCQs

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

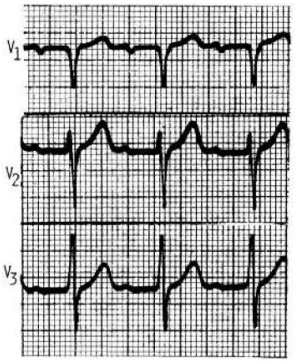
MCQs

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) 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

MCQs

5) A 72-year-old man presents with shortness of breath that awakens him at night. He is unable to walk more than one city-block before stopping to catch his breath.
Physical examination findings include normal blood pressure, bilateral basilar rales, and neck vein distention. T he patient has diabetes and a known history of congestive heart failure. His last echocardiogram revealed a left ventricular ejection fraction of 25%. T he patient has compliant with his medication regimen that includes an ACE inhibitor, beta-blocker, a loop diuretic, metformin, and glipizide. What is the most likely etiology for the patient's heart failure?

a. Metabolic
b. Infiltrative
c. Coronary artery disease
d. Valvular disease
e. Infectious

6) 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

MCQs

7) A 68-year-old man was intubated in the emergency room because of pulmonary edema. Stat echocardiogram reveals an ejection fraction of 45% and severe mitral regurgitation. In spite of aggressive diuresis with furosemide, the patient continues to require mechanical ventilation secondary to pulmonary edema. What is the best next step in treating this patient?

a. Arrange for mitral valve replacement surgery.
b. Begin intravenous milrinone.
c. Begin metoprolol.
d. Begin a second loop diuretic.
e. Begin intravenous enalapril.

8) 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

Answers : 1-D 2- C 3- E 4- B 5- C 6- A 7- E 8-B

DONE BY

Faroq Abdulfattah	Raneem AlOtaibi
Mojahed Otayf	Munira Ahmed
Mubarak Aldosrie	

Medicine is a science of uncertainty and an art of probability

MEDICINE 433