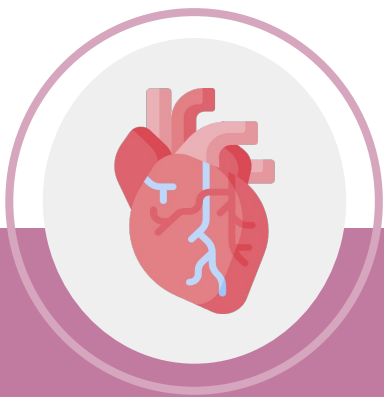


**Editing file**

# Rheumatic Heart Disease

No.3



## Objectives :

- ★ What is ARF And RHD?
- ★ Diagnosis, Jones Criteria & 2015 revision
- ★ Differential Diagnosis
- ★ Investigations
- ★ Management
- ★ Prevention
- ★ Rheumatic Valvular Heart Disease

### Color index

Original text

Females slides

Males slides

Doctor's notes <sup>438</sup>

Doctor's notes <sup>439</sup>

Doctor's notes <sup>442</sup>

New text in slides <sup>442</sup>

Text book

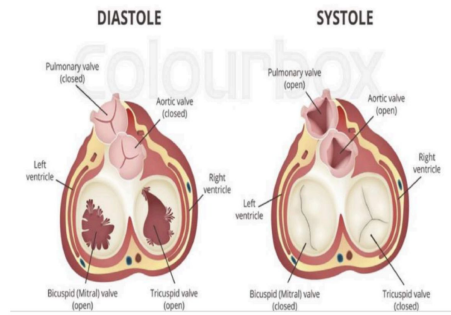
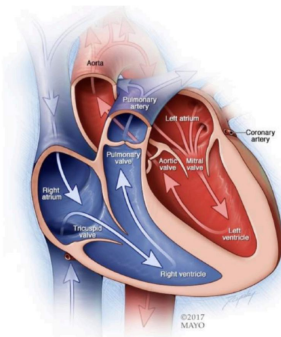
Important

Golden notes

Extra

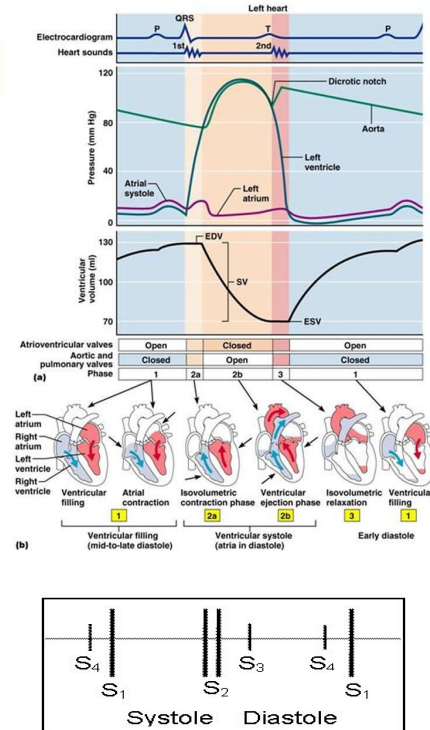
## Characteristics of heart valves

Valve	Structure	Site of auscultation	Open in	Sound
<b>Mitral valve</b>	Bicuspid	Left 5th intercostal space at the midclavicular line (Apex)	Diastole	S1 (LUB)
<b>Tricuspid valve</b>	Tricuspid	Left 5th intercostal space at the sternal border		
<b>Aortic valve</b>	Semilunar	Right 2nd intercostal space at the sternal border	Systole	S2 (DUB)
<b>Pulmonary valve</b>	Semilunar	Left 2nd intercostal space at the sternal border		



## Heart sounds

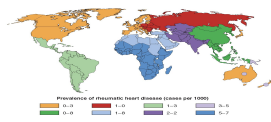
16.8 Normal and abnormal heart sounds				
Sound	Timing	Characteristics	Mechanisms	Variable features
<b>First heart sound (S1)</b>	Onset of systole	Usually single or narrowly split	Closure of mitral and tricuspid valves	Loud: hyperdynamic circulation (anaemia, pregnancy, thyrotoxicosis); mitral stenosis Soft: heart failure; mitral regurgitation
<b>Second heart sound (S2)</b>	End of systole	Split on inspiration Single on expiration (p. 447)	Closure of aortic and pulmonary valve	Fixed wide splitting with atrial septal defect Wide but variable splitting with delayed right heart emptying (right bundle branch block) Reversed splitting due to delayed left heart emptying (left bundle branch block)
<b>Third heart sound (S3)</b>	Early in diastole, just after S2	Low pitch, often heard as 'gallop'	From ventricular wall due to abrupt cessation of rapid filling	Physiological: young people, pregnancy Pathological: heart failure, mitral regurgitation
<b>Fourth heart sound (S4)</b>	End of diastole, just before S1	Low pitch	Ventricular origin (stiff ventricle and augmented atrial contraction) related to atrial filling	Absent in atrial fibrillation A feature of severe left ventricular hypertrophy
<b>Systolic clicks</b>	Early or mid-systole	Brief, high-intensity sound	Valvular aortic stenosis Valvular pulmonary stenosis Floppy mitral valve Prosthetic heart sounds from opening and closing of normally functioning mechanical valves	Click may be lost when stenotic valve becomes thickened or calcified Prosthetic clicks lost when valve obstructed by thrombus or vegetations
<b>Opening snap (OS)</b>	Early in diastole	High pitch, brief duration	Opening of stenosed leaflets of mitral valve Prosthetic heart sounds	Moves closer to S2 as mitral stenosis becomes more severe. May be absent in calcific mitral stenosis



# Introduction to RHD

## Overview on RHD epidemiology

- The incidence of RF and the prevalence of RHD has declined substantially in Europe, North America and other developed nations. This decline has been attributed to improved hygiene, reduced household crowding, and improved medical care. Also due to an improve in socioeconomic status and the usage of antibiotics.
- The major burden is currently found in low and middle income countries (India, middle east), and in selected indigenous populations of certain developed countries (Australia and New Zealand).
- In underdeveloped countries RHD is the **leading cause of CV death during the first five decades of life**<sup>4</sup>.
- **It is a disease of poverty and low socioeconomic status.**
- ARF incidence in Eastern province of SA was 22/100,000, age 5-14 years.
- A study from Western province showed a prevalence of RHD 2.4/1000, age 6-15 years.
- A large study showed an overall prevalence of ARF and RHD in SA 3.1/1000, age 6-15
- 2.8 M new cases and 40.5 M total cases of RHD were identified in 2019 globally, showing a 1.5 and 1.7x increase since 1990 respectively, with highest prevalence in Oceania, South Asia and sub-Saharan Africa.
- Prevalence ranged from 3.4 cases/100,000 in nonendemic regions to > 1000/100,000 in endemic areas



### Global burden

Total RHD cases	Cases with CHF	Cases require valve surgery	Annual incidence of RF, nearly half develop carditis	Deaths from RHD
15.6 million	3 million	1 million	0.5 million	300,000/YR

## 1- Acute Rheumatic Fever (ARF)

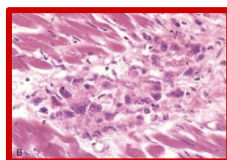
### General characteristics

- Acute rheumatic fever (ARF) is a **sequela of streptococcal infection**—typically **following 2 to 4 weeks after group A streptococcal (strep. pyogenes) pharyngitis** and has rheumatologic, cardiac, and neurologic manifestations.
- Acute rheumatic fever **usually affects children (most commonly 5-15 years)** or young adults.
- The most common cause of acquired heart disease in childhood and adolescence.
- **Rheumatic Heart disease (RHD) is a possible long-term consequence of ARF**<sup>5</sup>.

### Pathophysiology

1. The condition is triggered by an **immune-mediated delayed response** to infection with specific strains of **group A (beta hemolytic) streptococci** → Development of antibodies against streptococcal **M protein** → Cross reaction of antibodies with cardiac myosin and sarcolemmal membrane protein due to **molecular mimicry**.
2. Antibodies produced against the streptococcal antigens cause inflammation in the endocardium<sup>1</sup>, myocardium<sup>2</sup> and pericardium<sup>3</sup>, as well as the joints and skin, **but the major effect on health is due to damage to heart valves**.
3. **Histological findings: Aschoff bodies in myocardium composed of:**
  - a. Central area of fibrinoid necrosis surrounded by characteristic multinucleated giant cells, mononuclear cells (Anitschkow cells) and other inflammatory cells (plasma cells, and T lymphocytes) due to a type IV hypersensitivity reaction<sup>7</sup>

★ Questions about Aschoff nodules are very common, and you will be asked about it forever



1- Endocarditis: Mitral valve is involved more commonly than the aortic valve. It's characterized by small vegetations along lines of closure that lead to regurgitation (While chronic RF will lead to stenosis because of fibrosis)

2- Myocarditis is the most common cause of death during the acute phase.

3- Pericarditis will lead to friction rub and chest pain.

4- Causes of death in RHD : HF, IE, arrhythmia, embolism

5- there is a long gap between the attack of ARF and the RHD ( about 20-30 years )

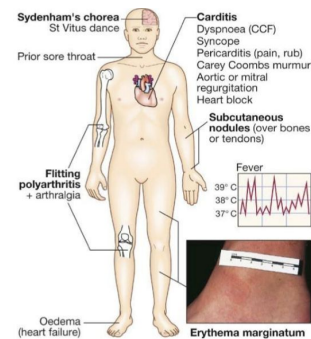
6- ARF is common cause of mortality and morbidity in young people

7- Acute rheumatic fever is due to a type II hypersensitivity reaction, but the chronic sequelae of acute rheumatic fever (i.e., rheumatic heart disease) involve a type IV hypersensitivity reaction that is responsible for non-caseating granuloma formation.

# 1- Acute Rheumatic Fever (ARF) cont.

## Clinical features

- Acute rheumatic fever is a **multisystem disease** that usually presents with fever (due to pyrogenic toxins), anorexia (due to several cytokines), lethargy and joint pain, **2-4 weeks after an episode of streptococcal pharyngitis**. There may be **no history** of sore throat.
- Arthritis** occurs in approximately **75%** of patients. Other features include subcutaneous nodules, erythema marginatum, carditis and neurological changes.



## Diagnosis

- Diagnosis is primarily clinical** and is based on a constellation of signs and symptoms, which were initially established as the **Jones criteria**. There's no single test to diagnose ARF.
- Only about 25%** of patients will **have a positive culture** for group A streptococcus at the time of diagnosis because there is a latent period between infection and presentation.
- Serological evidence of prior infection with GAS e.g.** Elevated ASO (Anti-Streptolysin O) titer or Anti-DNAse B titer
- A presumptive diagnosis of acute rheumatic fever can be made without evidence of preceding streptococcal infection in cases of isolated chorea or pancarditis, if other causes for these have been excluded.
- The symptoms and signs are shared by many inflammatory and infectious diseases.
- Accurate diagnosis is important<sup>1</sup>, because:**
  - 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

## Diagnostic criteria

- A firm diagnosis requires: 2 Major manifestations OR 1 Major and 2 Minor manifestations along with Evidence of a recent streptococcal infection** (Elevated ASO titer or Anti-Dnase B titer and a positive throat culture)

## 1992 Modified Jones criteria

Important as Mcqs



JONES Major criteria <sup>2</sup>	PEACE Minor criteria
<ol style="list-style-type: none"> <li><b>Migratory polyarthritis (Joints)</b></li> <li>♥ <b>Carditis</b></li> <li><b>Subcutaneous Nodules.</b></li> <li><b>Erythema Marginatum</b></li> <li><b>Sydenham chorea</b></li> </ol>	<ol style="list-style-type: none"> <li><b>Previous rheumatic fever</b></li> <li><b>Elevated temperature (fever)</b></li> <li><b>Arthralgia</b></li> <li><b>Elevated inflammatory markers (ESR<sup>3</sup>, CRP<sup>4</sup>)</b></li> <li><b>ECG: First degree heart block</b> (Prolonged PR interval)</li> </ol>

1- accurate diagnosis + prophylaxis administration > to prevent further attack of ARF ( cause every attack will cause more damage)

2- Discussed in details in page 6

3- Erythrocyte Sedimentation Rate: is a test that indirectly measures the degree of inflammation present in the body. The test actually measures the sedimentation of erythrocytes (RBCs) in a sample of blood that has been placed into a tall, thin and vertical tube. RBCs normally settle out relatively slowly, but when they settle at a faster rate, this indicates that there is an increased level of proteins, particularly CRP and fibrinogen, and these proteins increase in the blood in response to inflammation.

4- C-reactive protein: Is a substance produced by the liver in response to inflammation.

# 1- Acute Rheumatic Fever (ARF) cont.

## ◀ Diagnostic criteria cont.

### ◆ 2015 Revision of Jones criteria

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

1

- A. **Low risk populations** have been defined as those with:
- ARF incidence < **2:100000 school-age children** or
  - All age prevalence of RHD of < **1:1000 population per year**
- B. Children not from low risk population have been considered to be at **moderate or high risk**.

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** and **Polyarthralgia** has been included as a **major criteria in moderate or high risk population**.

4

**Fever, ESR & CRP:**

- A. **low risk population:** >38.5°C, ESR >60 and or CRP > 3mg/dl  
 B. **Moderate or high risk population:** >38°C and ESR >30 and or CRP > 3mg/dl.

### ◆ So, let's mix the modified and revised versions of Jones criteria

For all patient populations with evidence of preceding GAS infection:

- Diagnosis of **Initial ARF**: **2 Major OR 1 Major and 2 Minor**. (If it's the first time, **3 minor aren't enough**)
- Diagnosis of **recurrent ARF**: **2 Major OR 1 Major and 2 Minor OR 3 Minor**. - Presence of antecedent streptococcal infection - When minor manifestations only present exclude other causes.

#### Major criteria

Low-risk populations	Moderate- and high-risk populations
<ol style="list-style-type: none"> <li><b>Carditis:</b> <ol style="list-style-type: none"> <li>Clinical and/or subclinical (by echo).</li> </ol> </li> <li><b>Arthritis:</b> <ol style="list-style-type: none"> <li>Polyarthritits <b>only</b></li> </ol> </li> <li><b>Sydenham chorea</b></li> <li><b>Erythema marginatum</b></li> <li><b>Subcutaneous nodules</b></li> </ol>	<ol style="list-style-type: none"> <li><b>Carditis:</b> <ol style="list-style-type: none"> <li>Clinical and/or subclinical (by echo).</li> </ol> </li> <li><b>Arthritis:</b> <ol style="list-style-type: none"> <li>Polyarthritits or <b>monoarthritis</b></li> <li><b>Polyarthralgia</b></li> </ol> </li> <li><b>Chorea</b></li> <li><b>Erythema marginatum</b></li> <li><b>Subcutaneous nodules</b></li> </ol>

#### Minor Criteria

Low-risk populations	Moderate- and high-risk populations
<ol style="list-style-type: none"> <li><b>Polyarthralgia</b></li> <li><b>Fever ( ≥ 38.5°C)</b></li> <li><b>ESR ≥ 60 mm</b> in the first hour or CRP ≥ 3mg/dl</li> <li><b>Prolonged PR interval</b>, after accounting for age variability (Unless carditis is a major criterion)<sup>1</sup></li> </ol>	<ol style="list-style-type: none"> <li><b>Monarthralgia</b></li> <li><b>Fever ( ≥ 38°C)</b></li> <li><b>ESR ≥ 30 mm</b> in the first hour or CRP ≥ 3mg/dl</li> <li><b>Prolonged PR interval</b>, after accounting for age variability (Unless carditis is a major criterion)<sup>1</sup></li> </ol>

<sup>1</sup> If carditis is present as a major manifestation, a prolonged P-R interval cannot be considered an additional minor manifestation.

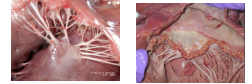
## Diagnostic criteria cont.

### Jones major criteria explanation:

#### Carditis <sup>1</sup>



- A '**pancarditis**', occurs in 50-70% of cases. Its incidence declines with increasing age.
- It may manifest as **breathlessness<sup>2</sup>, palpitations or chest pain**
- **Tachycardia is common; its absence makes the diagnosis of myocarditis unlikely.** (Sometimes only symptom of carditis)
- **The only manifestation of ARF that leaves serious permanent damage.**
- **Valvular lesions<sup>4</sup> :**
  - **Mitral valve: (Most common to be involved)**
    - Early mitral regurgitation (due to vegetation) or prolapse while mitral stenosis in late stage (due to fibrosis)<sup>f</sup>
  - **Aortic valve:**
    - Early Aortic regurgitation while aortic stenosis in late stage
- **A soft Pansystolic murmur due to mitral regurgitation** is very common and/ or a soft mid-diastolic murmur (the Carey Coombs murmur) is typically due to valvulitis.
- May be **subclinical** (without murmurs or other S&S) so we should **use Echo.**
- Cardiomegaly and CHF may occur (due to myocarditis)
- Myocarditis is the most common cause of death during the acute phase
- **Signs of carditis: tachycardia, murmurs of MR/AR, cardiomegaly and signs of CHF**



#### Arthritis



- This is the **most common major presenting manifestation and occurs early** when streptococcal antibody titres are high, occurs in 35-66% of cases. Duration is short < 1 week. and it is the **Earliest Manifestation of ARF**
- An acute **painful asymmetric and fleeting<sup>3</sup> migratory inflammation of the large joints** typically affects the **knees, ankles, shoulders, elbows.** The joints are involved in quick succession and are usually red and swollen.
- **Rapid improvement with salicylates (aspirin) and NSAID.** The pain characteristically responds to aspirin; if not, the diagnosis is in doubt.
- Does not progress to chronic disease.
- Monoarthritis is now accepted as a major criterion



#### Sydenham chorea

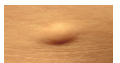


- Also known as **Saint Vitus's Dance**, occur in 10-30%, female predominance.
- This is a **late extrapyramidal neurological manifestation** that appears at least 3 months after the episode of acute rheumatic fever, when all the other signs may have disappeared. (month after GAS infection)
- **Emotional lability may be the first feature** and is typically **followed by Abrupt purposeless, involuntary, Irregular, nonrepetitive choreiform movements 'Fidgety'** of the **hands, face, neck, trunk and limbs.**
- Clinically manifest as clumsiness, deterioration of handwriting, emotional lability or grimacing of face.
- Spontaneous recovery usually occurs within a few months. Approximately one-quarter of affected patients will go on to develop chronic rheumatic valve disease.
- Strongly associated with carditis
- Can be a standalone criterion for diagnosis of ARF

#### Subcutaneous nodules

(Associated with severe carditis)

- They are **small** (0.5–2.0 cm), **firm** and **painless, free mobile (not tender)** and are **best felt over extensor surfaces** of joints, bone prominences, tendons and spine. occurs in 10% of cases.
- Rare but highly specific manifestation of ARF
- Short lived (lasts for few days). They typically appear more than 3 weeks after the onset of other manifestations and therefore **help to confirm** rather than make the diagnosis.



#### Erythema marginatum

(Associated with carditis)

- Erythema marginatum occurs in less than 5-6% of patients (rare)
- The lesions start as round or irregular red macules that fade in the centre (**pale center**) but remain **red at the edges** (wavy/serpiginous margin), non-pruritic (not itchy), transient rash
- The resulting red rings or 'margins' may coalesce or overlap.
- Location: Mainly on the trunk, abdomen and proximal extremities but the face spared.
- Less common, but **highly specific** manifestation of ARF.



1- the heart is the only organ that will left with permanent abnormalities while the other organs will heal without any problems

2- Shortness of breath is caused by the decreased ability of the heart to fill and empty, producing high pressure in the blood vessels around the lung

3- it means that it move from one joint to another e.g. Knee in morning, elbow in noon (**it is important information → to exclude other differential diagnosis**)

4- If you find one valve affected keep in mind may be other valves are affected too , so you have to check it them all.

# 1- Acute Rheumatic Fever (ARF) cont.

## ◀ Differential diagnosis of ARF

important table



Presentation		
Polyarthritis and fever	Carditis	Chorea
<ol style="list-style-type: none"><li>1) Septic arthritis (Including disseminated gonococcal infection)</li><li>2) Connective tissue and other autoimmune disease</li><li>3) Viral arthropathy</li><li>4) Reactive arthropathy</li><li>5) Lyme disease</li><li>6) Sickle cell anemia</li><li>7) Infective endocarditis</li><li>8) Leukemia or lymphoma</li><li>9) Gout or pseudogout</li></ol>	<ol style="list-style-type: none"><li>1) Innocent murmur</li><li>2) Mitral valve prolapse</li><li>3) Congenital heart disease</li><li>4) Infective endocarditis</li><li>5) Hypertrophic cardiomyopathy<sup>3</sup></li><li>6) Myocarditis: Viral or idiopathic<sup>4</sup></li><li>7) Pericarditis: or idiopathic</li></ol>	<ol style="list-style-type: none"><li>1) SLE</li><li>2) Drug intoxication</li><li>3) Wilson's disease</li><li>4) Tic disorder</li><li>5) Choreoathetoid cerebral palsy</li><li>6) Encephalitis</li><li>7) Familial chorea (Including Huntington's)</li><li>8) Intracranial tumor</li><li>9) Lyme disease</li><li>10) Hormonal</li></ol>

## ◀ Investigations

01



### Evidence of a systemic illness:

- a. **CBC: Leukocytosis**
- b. **Raised erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP).**

02



### Evidence of preceding streptococcal infection:

- a. **Throat swab culture (preferably before giving antibiotics)<sup>1</sup>:**  
group A beta-hemolytic streptococci (also from family members and contacts)
- b. **Anti-streptococcal serology:** (Repeat 10-14 days later if first test not confirmatory)
  - i. **Elevated Antistreptolysin O antibodies (ASO titres)<sup>2</sup>:**  
>200 U (adults) or >300 U (children)
  - ii. **Elevated Ant-DNAse B titer.**

03



### Evidence of carditis:

- a. **Chest X-ray:** cardiomegaly; pulmonary congestion
- b. **ECG:**
  - i. **First-degree AV block (Prolonged PR interval);** features of pericarditis; T-wave inversion; reduction in QRS voltages.
  - ii. Arrhythmias
  - iii. Repeat in 2 weeks and again in 2 months, if still abnormal.
- c. **Echocardiography:** (If negative, consider repeating after 1 month)
  - i. Cardiac dilatation
  - ii. Mitral regurgitation with dilatation of the mitral annulus and prolapse of the anterior mitral leaflet; it may also demonstrate aortic regurgitation and pericardial effusion.

1- Throat cultures should be taken but positive results are obtained in only 10–25% of cases since the infection has often resolved by the time of presentation.  
2- Raised levels provide supportive evidence for the diagnosis but are normal in one-fifth of adult cases of rheumatic fever and most cases of chorea.  
3-Great mimicker  
4-very common nowadays

# 1- Acute Rheumatic Fever (ARF) cont.

## Treatment

### Bed Rest:

The duration should be guided by symptoms, along with temperature, leucocyte count and ESR, and should be continued until these have settled.

### Antibiotics:

Like Procaine benzylpenicillin  $G^2$  4 million units/day x10 days. Should be given on diagnosis to **eradicate GAS from throat** infection.

### Heart Failure

**Treatment :**  
Like **diuretics** and **ACEI, Beta blockers.**



### Salicylates:

★ **Aspirin<sup>1</sup>** 60/75-100 mg/kg/day as 4 divided doses for 6-8 weeks, until a blood level of 20-30 mg/dl is attained. This usually **relieves** the symptoms of **arthritis** rapidly and a response within 24 hours helps confirm the diagnosis. ( we give high doses unlike coronary arteries diseases )

### Steroids (Prednisolone):

1-2 mg/kg/day taper over 6 weeks. Produces **more rapid symptomatic relief** than aspirin and is **indicated in cases with carditis or severe arthritis**. Prednisolone should be continued until the ESR is normal and then tailed off.

## Prevention



Very Important

### Primary prevention:

- Aims to prevent ARF from developing as result of GAS infection
- Prompt antibiotic treatment of GAS pharyngitis with **BPG 1.2M units IM**.

### Secondary prevention:

Screening to identify diseases in the earliest stage before the onset of signs and symptoms

- Antibiotics prophylaxis with **IM penicillin G benzathine** 1.2MU every 4 weeks or 3 weeks in high risk situations (alternatively 250mg **oral penicillin V**) ( give IM to increase the patient's compliance on the treatment especially children , because easier to take IM injection once monthly instead of taking the oral drug daily)
- If the patient is penicillin allergic:** Oral **Sulfadiazine** ( 0.5g once daily for patients 27kg (60lb) OR 1g once daily for patients >27kg (60lb)
- If the patient is penicillin and sulfadiazine allergic:** use oral **erythromycin** 250mg twice daily

**Duration depend on the risk and severity of the original episode:**

**Important** ★

**Duration of Prophylaxis depends on :**  
1-age 2-Time since last ARF 3- Presence of rheumatic carditis at presentation 4-Severity of chronic RHD

**1** Rheumatic fever **without carditis:**  
**5 years or until patient reaches 21 years of age.** (whichever is longer)

**2** Rheumatic fever **with carditis:**  
**10 years or until patient reaches 21 years of age.** (whichever is longer)

**3** Rheumatic fever **with carditis and permanent valvular heart defects:**  
**10 years or until patient reaches 40<sup>3</sup> years of age.** (whichever is longer, sometime life long prophylaxis)

1- Aspirin should be continued until the ESR has fallen and then gradually tailed off, If aspirin fails then use Glucocorticoids.

2- If the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.

3- Why 40? Because throat infections after the age of 40 are rare, unless the person is working closely with kids then therapy should be continued



## ◀ General characteristics

- **A sequela of ARF**, chronic valvular heart disease develops in at least half of those affected by rheumatic fever with carditis.
- A leading cause of CV morbidity & mortality in young people.
- **The mitral valve is affected in more than 70% of cases; the aortic valve is the next most frequently involved (40%) , followed by the tricuspid (10%) and then the pulmonary valve (2%).**
- **Mitral Stenosis is more common in females (3:1), while males have higher incidence of Aortic Regurgitation.**
- The main pathological process in chronic rheumatic heart disease is progressive fibrosis.
- 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.

New slides 442



### Chronic Rheumatic Heart Disease:

- RHD typically affects left sided valves mitral and aortic valves
- Right sided valves: affects mainly tricuspid valve as valvulitis or as a result of hemodynamic consequences of mitral valve involvement. Pulmonary involvement is rare
- Acute rheumatic valvulitis manifests as Valvular regurgitation, but overtime chronic inflammation leads to Valvular stenosis from commissural fusion with or without regurgitation

### Clinical Features

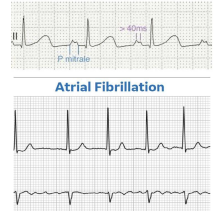
- RHD is usually clinically silent (latent) until it presents later during adulthood
- Palpitations due arrhythmias commonly AF 50% of patients may present without any prior symptoms of ARF.
- Exertional dyspnea which worsens gradually and symptoms of heart failure
- Palpitations due arrhythmias commonly AF . Chest pain and syncope(aortic stenosis)
- Weakness (stroke) or limb ischemia due embolization(Mitral stenosis)
- Hemoptysis (mitral stenosis) .Infective endocarditis

### Investigations: Echo

- Establishing diagnosis of RHD.
- Determining severity of Valvular lesions.
- Determining physiological consequences
- Planning surgical (valve replacement or repair) or interventional therapy (mitral balloon valvuloplasty)
- Screening and active case finding among school-age children in endemic areas to detect subclinical cases; cost-effectiveness needs to be determined

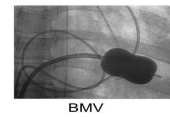
## Investigations: ECG

- LAE, AF (mitral valve disease)
- RVH (mitral stenosis)
- LVH (aortic valve disease)
- CXR: 1- LAE (MS) 2- Cardiomegaly (MR,AR) 3- Pulmonary venous congestion



## Management of RHD

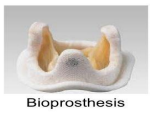
- Medical: management of heart failure
- Interventional: mitral balloon valvuloplasty for mitral stenosis
- Surgical: valve repair or replacement



BMV



Mechanical valve



Bioprosthesis

## Complications of Chronic RHD

- During pregnancy: multidisciplinary team needed
  - 1-Increased risk of poor maternal and fetal outcomes.
  - 2-Stenotic lesions are less tolerated than regurgitation lesions.
  - 3-May require intervention during pregnancy.
  - 4- Pregnant women with prosthetic valves require special care for managing anticoagulation
- Pulmonary hypertension
- Premature death
- Heart failure:- Medical treatment- Catheter based and surgical interventions for severe disease
- Atrial fibrillation:- Stroke & peripheral embolism - Anticoagulation
- Infective endocarditis: a risk before and after surgery - Carries a high mortality = 30% at 1 year IE prophylaxis

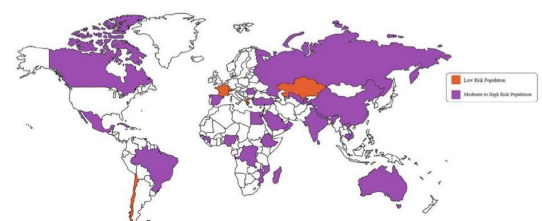
## Barriers to RHD Control


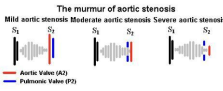
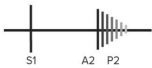
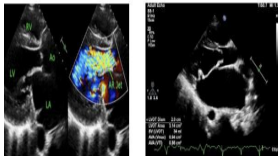
- Poor resources in countries where RHD is endemic
- Access to surgery and catheter-based interventions
- Implementation and adherence to secondary prophylaxis with BPG is a cornerstone in RHD management
- Development of Group A streptococcus vaccine: genomic heterogeneity of Strep A and subsequent sequence variations, limiting effectiveness of vaccine

### Endemic and non-endemic areas countries of RHD



### World Map of RHD



	Mitral Stenosis	Mitral Regurgitation	Aortic Stenosis	Aortic Regurgitation
	<ul style="list-style-type: none"> <li>- The normal MVA=4-5cm<sup>2</sup></li> <li>- In severe ms&lt;1.5cm<sup>2</sup></li> <li>- HighLAP</li> <li>- The rise in LAP causes a similar rise in pulmonary capillaries ,veins and artery</li> </ul>	<ul style="list-style-type: none"> <li>- Asymptomatic</li> <li>-Dyspnea,orthopnea,P ND- Displaced PMI,Thril</li> <li>- SoftS1,</li> <li>- Pansystolicmurmur</li> <li>- Treatment is surgical</li> </ul> 	<p>Aortic stenosis</p> 	<p>Aortic regurgitation</p> 
<b>Clinical features</b>	<ul style="list-style-type: none"> <li>- Dyspnea <b>those are Qs come in physical exams</b></li> <li>- Fatigue</li> <li>- Palpitation</li> <li>- Hemoptysis (10%)</li> <li>- Hoarseness</li> <li>- Dysphagia</li> <li>- Stroke or peripheral embolization</li> <li>- Cyanosis (Mitral facies , malar flush)</li> <li>- tapping apex (S1)</li> <li>- Parasternal heave</li> <li>- Diastolic thrill</li> <li>- Accentuated S1 , accentuated S2</li> <li>- Opening snap</li> <li>- Mid-diastolic rumble</li> </ul>		<p>Symptoms:</p> <ul style="list-style-type: none"> <li>- Angina</li> <li>- Syncope</li> <li>- Dyspnea</li> </ul> <p>Signs:</p> <ul style="list-style-type: none"> <li>- Arterial Pulse wave form :Plateau</li> <li>. Small (Parvus)</li> <li>. Slow rise (Tardus)</li> <li>- Sustained not displaced PMI</li> <li>- Systolic thrill</li> <li>- S4</li> <li>- Late peaking of murmur</li> <li>-Single S2: Soft or absent A2</li> <li>-Paradoxical splitting of S2</li> </ul>	<ul style="list-style-type: none"> <li>-Water-hammer/colap sing pulse</li> <li>- Wide pulse pressure</li> <li>- Corigan's sign</li> <li>- De Muset sign</li> <li>- Muler sign</li> <li>- Quincke's pulse</li> <li>- Hill's sign</li> </ul>
<b>Investigations</b>	<ul style="list-style-type: none"> <li>- CXR: Straightening of left heart border , left atrial enlargement</li> <li>Double density</li> <li>Kerley B lines , CA in MV</li> <li>- ECG: LAE , P Mitrale , RV dominance ,atrial fibrillation</li> <li>- Echodoppler</li> <li>Echo: thickend leaflets with commissural fusion</li> </ul>	<ul style="list-style-type: none"> <li>- Echo</li> </ul>		<ul style="list-style-type: none"> <li>- Echo</li> </ul> 
<b>Management</b>	<ul style="list-style-type: none"> <li>- B-Blockers , CCB</li> <li>- Digoxin (AF)</li> <li>- Warfarin</li> <li>- Balloon Valvuloplasty</li> <li>- Mitral valve replacement</li> </ul>		<ul style="list-style-type: none"> <li>- Aortic valve Replacement</li> <li>- Transcatheter Aortic Valve Replacement</li> </ul>	<ul style="list-style-type: none"> <li>- Aortic valve Replacement</li> <li>- Transcatheter Aortic Valve Replacement</li> </ul>

RHD	
<b>Etiology</b>	Occurs two to three weeks after an untreated infection with group A streptococcus (GAS).
<b>Pathology</b>	Molecular mimicry between streptococcal M protein and human cardiac myosin proteins.
<b>S&amp;S</b>	Nonspecific symptoms (e.g., fever, malaise, and fatigue) and <b>JONES</b>
<b>Diagnosis</b>	Primarily clinical, and is based on the Jones criteria. Diagnostic evaluation in acute rheumatic fever typically shows elevated inflammatory markers, positive antistreptococcal antibodies, and valvular damage on echocardiogram.
<b>Treatment</b>	The first-line treatment is penicillin combined with symptomatic anti-inflammatory treatment, typically with salicylates or glucocorticoids (if salicylates are not effective).
<b>Primary Prevention</b>	Prompt antibiotic treatment of GAS pharyngitis with <b>BPG 1.2M units IM</b> .
<b>Secondary Prevention</b>	<ul style="list-style-type: none"> <li>a) Antibiotics prophylaxis with <b>IM penicillin G benzathine 1.2MU</b> every 4 weeks or 3 weeks in high risk situations (alternatively 250mg <b>oral penicillin V</b>)</li> <li>b) <b>If the patient is penicillin allergic:</b> Oral <b>Sulfadiazine</b> ( 0.5g once daily for patients 27kg (60lb) OR 1g once daily for patients &gt;27kg (60lb)</li> <li>c) <b>If the patient is penicillin and sulfadiazine allergic:</b> use oral <b>erythromycin</b> 250mg twice daily</li> </ul>
<b>Duration of Prevention</b>	<ul style="list-style-type: none"> <li>- <b>Rheumatic fever without carditis:</b> 5 years <u>or</u> until patient reaches <b>21 years</b> of age. (whichever is longer)</li> <li>- <b>Rheumatic fever with carditis:</b> 10 years <u>or</u> until patient reaches <b>21 years</b> of age. (whichever is longer)</li> <li>- <b>Rheumatic fever with carditis and permanent valvular heart defects:</b> 10 years <u>or</u> until patient reaches <b>40</b> years of age (whichever is longer, sometime life long prophylaxis)</li> </ul>

# Lecture Quiz

**Q1: A 32-year-old recent immigrant from Haiti presents for her first clinic evaluation. When she was 12 years old, she had rheumatic fever that was treated with antibiotics. She is asymptomatic. Her physical examination shows a heart rate of 70 beats per minute with a blood pressure of 122/77 mm Hg. She has no jugular venous distention. Her lungs are clear. She has a regular rate and rhythm with a loud S1 and physiologically split S2. A soft early diastolic rumble is heard at the apex with an opening snap after S2. There is wide separation between A2 and the opening snap. No peripheral edema is present. Which of the following is the most appropriate next recommendation for this patient?**

- A- Start Anticoagulation
- B- Start oral penicillin V 250 mg twice daily
- C- Start diuretics
- D- Take abx for dental procedure

Hct	42%
WBC	12,000/ $\mu$ L with 80% polymorphonuclear leukocytes, 20% lymphocytes
ESR	60 mm/h

**Q2: 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 central clearing. The following laboratory values are obtained:**

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

**Q3: A 12-year-old girl is brought to the physician by her mother because of high fever and left ankle and knee joint swelling. She had a sore throat 3 weeks ago. There is no family history of serious illness. Her immunizations are up-to-date. She had an episode of breathlessness and generalized rash when she received dicloxacillin for a skin infection 2 years ago. She appears ill. Her temperature is 38.8°C (102.3°F), pulse is 87/min, and blood pressure is 98/62 mm Hg. Examination shows left ankle and knee joint swelling and tenderness; range of motion is limited. Breath sounds over both lungs are normal. A grade 3/6 holosystolic murmur is heard best at the apex. Abdominal examination is normal. Which of the following is the most appropriate pharmacotherapy?**

- A- Vancomycin
- B. Amoxicillin
- C. Clarithromycin
- D. Doxycycline
- E. Ciprofloxacin

# Our Team

This work was originally done by **438 Medicine team.**

Edited by **439 Medicine team:**

## Team Leaders

- *Shaden Alobaid*  
- *Ghada Alabdi*

- *Hamad Almousa*  
- *Naif Alsulais*



**Member :** Abdullah Alanzan  
**Note taker :** Nouf Alsubaie  
Hamad Almousa

Edited for the second time by **442 Medicine team:**

## Team Leaders

- *Mohammed Alrashoud*  
- *Maha Alzahrani*  
- *Shatha Alshabani*

- *Mohammed Alzeer*  
- *Refal Alamry*  
- *Arwa Alghamdi*



**Members:** atheer alahmari  
Bandar almutairi



**[Medicine.med442@gmail.com](mailto:Medicine.med442@gmail.com)**

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