

# **Rheumatic Heart Disease**

## Objectives:

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

Team members:Trad Alwakeel + Khalid Aleedan + Yazeed alsuhaibani + Rayan Alqarni

Team leader: Haneen Al Subki

Revised by: Maha AlGhamdi

Designed by: <u>A.Alangari@outlook.sa</u>

Resources:435 team + Davidson + Recall questions step up to medicine

- Editing file
- <u>Feedback</u>

### • Epidemiologic Background:

 $\star$  We are concerned about prevalence and incidence because when we want to diagnose a patient with ARF based on the criteria , we have to know which criteria we should use depending on our country.

Saudi Arabia is classified in moderate risk group.

- Globally RHD is the most common CVD in young people 25 yrs old.
- The overall incidence of ARF is 5-51/100000 people, with a mean of 19 per 100000 people.
- 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 Europe, North America and other developed nations. This decline has been attributed to <u>improved hygiene</u>, reduced <u>household crowding</u>, and improved medical care. Also due to an improve in socio-economical 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).
- A disease of poverty and low socioeconomic status.
- In underdeveloped countries RHD is the leading cause of CV death during the first five decades of life.
- In Saudi Arabia: incidence 30 cases/100000 people/yr and prevalence 310/100000 people in 6-15 yrs age group.

#### • Global burden:

- Total cases with RHD : 20 Millions
- 3 Million have congestive heart failure.
- 1 Million require Valve surgery
- Annual incidence<sup>1</sup> of RF: 0.5 Million, nearly half develop **carditis**
- Estimated deaths from RHD: 230,000/YR.

<sup>&</sup>lt;sup>1</sup> New cases per year

# A. Acute Rheumatic fever: \* ARF leads to RHD after 10-20 years (latent period) . <u>watch video!</u>

| Incidence and<br>pathogenesis | <ul> <li>Acute rheumatic fever usually affects children (most commonly between 5 and 15 years) or young adults .</li> <li>The most common cause of acquired heart disease in childhood and adolescence.</li> <li>The condition is triggered by an immune-mediated <u>delayed</u> response (manifestations appearing after a period of 2-4 weeks) 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 and skin, but the major effect on health is due to damage to heart valves.</li> <li>Histologically, fibrinoid degeneration is seen in the collagen of connective tissues. (VERY IMP) <u>Aschoff nodules</u> 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. Antischkow cells are enlarged macrophages found within granulomas (called aschoff bodies ) associated with the disease . Larger Antischow cells may coalesce to form multinucleted Aschoff bodies . (See the pic)</li> </ul> |  |  |  |  |
|-------------------------------|--|--|--|--|--|
| Clinical<br>features          | <ul> <li>Acute rheumatic fever is a multisystem disorder that usually presents with fever, anorexia, lethargy and joint pain, 2–3 weeks after an episode of streptococcal pharyngitis. There may, however, be no history of sore throat.</li> <li>Arthritis occurs in approximately 75% of patients. Other features include rashes, carditis and neurological changes.</li> <li>Clinical features are not specific. for example : leukemia, sickle cell anemia and viral infections can cause joint pain and swelling. So you should diagnose the patient carefully.</li> <li>As we mentioned, it's a multisystem disorder that will affect:         <ul> <li>CNS : mainly basal ganglia</li> </ul> </li> </ul>  |  |  |  |  |
|                               | <ul> <li>2) Joints and skin : will recover later</li> <li>3) Heart : permanent damage to the valves</li> </ul>   |  |  |  |  |
| Diagnosis                     | <ul> <li>No single test to diagnose ARF.</li> <li>The symptoms and signs are shared by many inflammatory and infectious diseases</li> <li>Accurate diagnosis is important</li> <li>Overdiagnosis will result in individuals receiving unnecessary treatment</li> <li>Underdiagnosis may lead to further episodes of ARF causing damage, and the need for valve surgery, and or premature death</li> <li>Diagnosis is primarily clinical and is based on a constellation of signs and symptoms, which were initially established as the Jones criteria.</li> <li>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.</li> <li>Serological evidence of recent infection with a raised antistreptolysin O (ASO) antibody titre is helpful.</li> <li>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.</li> <li>In cases of established rheumatic heart disease or prior rheumatic fever, a diagnosis of acute rheumatic fever can be made based only on the presence of multiple minor criteria and evidence of preceding</li> </ul>                       |  |  |  |  |

#### group A streptococcal pharyngitis.

#### **1992 Modified Jones Criteria :** (we'll talk the about major criteria in details later on)

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

It is based upon two or more major manifestations, or one major and two or more minor manifestations, <u>along</u> with evidence of preceding streptococcal infection.

#### **2015 Revision of Jones Criteria:**

The new criteria divided the population into 2 groups : high and low risk populations.

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

- <u>low risk populations</u> 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
- 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 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 >38.5 c, ESR >60 and or CRP > 3mg/dl for <u>low risk population</u>

fever >38 and ESR >30 and or CRP > 3mg/dl for moderate or high risk population.

<u>A firm diagnosis requires:</u> 2 Major manifestations or 1 Major and 2 Minor manifestations <u>And</u> Evidence of a recent streptococcal infection. Or 3 Minor

**Evidence of Preceding GAS** "Group A strept." Infection: 1) Increased or rising ASO titer or Anti-Dnase B titer.

#### 2) A positive throat culture.

| A. For all patient populations with evidence of preceding GAS infec                                   |  |  |
|---|--|--|
| Diagnosis: initial ARF  | 2 Major manifestations or 1 major plus 2 minor<br>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†<br>• Clinical and/or subclinical  | Carditis <ul> <li>Clinical and/or subclinical</li> </ul>   |  |
| Arthritis<br>← Polyarthritis only   | Arthritis<br>● Monoarthritis or polyarthritis<br>● 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 $\geq$ 30 mm/h and/or CRP $\geq$ 3.0 mg/dL§  |  |
| Prolonged PR interval, after accounting for age variability<br>(unless carditis is a major criterion) | Prolonged PR interval, after accounting for ag<br>variability (unless carditis is a major criterion) |  |

| - | Rheumatic Fever Recurrences:         Reliable past history of ARF: 2 major or 1 major and 2 minor or 3 minor manifestations sufficient for diagnosis         Presence of antecedent (previous) streptococcal infection         When minor manifestations only present exclude other causes. |
|---|---|
|---|---|

| Jones criteria (major criteria) |  |  |  |
|---------------------------------|--|--|--|
| 1-Carditis:                     | <ul> <li>A 'pancarditis' involves the endocardium, myocardium and pericardium to varying degrees. Its incidence <u>declines with aging</u>, ranging from 90% at 3 years to around 30% in adolescence.</li> <li>Occurs in 50-70% of cases</li> <li>The only manifestation of ARF that leaves serious permanent damage</li> <li>May be <i>subclinical</i> (without murmurs so we should use Echo)</li> <li>Cardiomegaly and CHF may occur</li> </ul>   |  |  |
| 2-Arthritis:                    | <ul> <li>This is the most common major manifestation and occurs early when streptococcal antibody titres are high (present in 35-66%, <u>Earliest manifestation of ARF</u>).</li> <li>An acute painful <i>asymmetric and migratory inflammation (Migrating, "Fleeting</i>" polyarthritis) of the large joints typically affects the knees, ankles, shoulders, elbows. The joints are involved in quick succession and are usually red, swollen and tender.</li> <li>Duration short &lt; 1 week.</li> <li>Rapid improvement with salicylates. The pain characteristically responds to aspirin; if not, the diagnosis is in doubt.</li> <li>Does not progress to chronic disease.</li> </ul>   |  |  |
| 3-Sydenham<br>Chorea:           | <ul> <li>Also known as Saint Vitus's Dance, occur in 10-30%, female predominance.</li> <li>This is a late neurological manifestation (extrapyramidal manifestation) that appears at least 3 months after the episode of acute rheumatic fever, when all the other signs may have disappeared (Delayed manifestation of ARF).</li> <li>Emotional lability may be the first feature and is typically followed by purposeless, involuntary, choreiform movements of the hands,face, neck, trunk, and limbs Speech may be explosive and halting (<i>clumsiness, deterioration of handwriting,emotional lability or grimacing of face</i>).</li> <li>Spontaneous recovery usually occurs within a few months. Approximately one-quarter of affected patients will go on to develop chronic rheumatic valve disease</li> </ul> |  |  |
| 4-Subcutaneous<br>Nodules:      | <ul> <li>Subcutaneous nodules occur in 10% of patients.</li> <li>They are small (0.5–2.0 cm), firm non-tender and painless.</li> <li>Best felt over extensor surfaces of bone or tendons (Occur over extensor surfaces of joints, on bony prominences, tendons, spine).</li> <li>They typically appear more than 3 weeks after the onset of other manifestations and therefore help to confirm rather than make the diagnosis</li> <li>Short lived: last for few days.</li> <li>Associated with severe carditis.</li> </ul>  |  |  |

| <b>5-Erythema</b><br><b>Marginatum:</b><br>Very few will have<br>this | <ul> <li>Erythema marginatum occurs in less than 6% of patients</li> <li>The lesions start as red macules that fade in the centre(pale center) but remain red at the edges, round or irregular serpiginous (having a wavy margin) borders, non-pruritic, transient rash. The resulting red rings or 'margins' may coalesce or overlap.</li> <li>Occur mainly on the trunk and proximal extremities but not the face.</li> <li>Less common, but highly specific manifestation of ARF.</li> <li>Associated with carditis.</li> </ul> |   |   |  |  |
|---|--|---|---|--|--|
| DDX of ARF :  | Presentation   | Contrit   | Chamber   |  |  |
|   | Polyarthritis and feverSeptic arthritis (including<br>disseminated gonococcal infectiConnective tissue and other<br>autoimmune disease**Viral arthropathy*Reactive arthropathy*Lyme disease*Sickle cell anaemiaInfective endocarditisLeukaemia or lymphomaGout and pseudogout  | CarditisInnocent murmurMitral valve prolapseCongenital heart diseaseInfective endocarditisHypertrophic cardiomyopathyMyocarditis: viral or idiopathicPericarditis: viral or idiopathic  | Chorea<br>Systemic lupus erythematosus<br>Drug intoxication<br>Wilson's disease<br>Tic disorder <sup>‡</sup><br>Choreoathetoid cerebral palsy<br>Encephalitis<br>Familial chorea<br>(including Huntington's)<br>Intracranial tumour<br>Lyme disease <sup>‡</sup><br>Hormonal <sup>§</sup> |  |  |
| Investigations  | <ul> <li>in 2 months, if still abnormal</li> <li>→ Chest X-ray, if clinical or ech</li> <li>→ Echocardiogram Very import</li> <li>→ Throat swab (preferably befo</li> <li>→ Anti-streptococcal serology:<br/>first test not confirmatory)</li> </ul>   | tte (ESR).<br>ged P-R interval or other rhythm abnorm<br>I). To look for Heart block (minor criteri<br>hocardiographic evidence of carditis.<br>tant (consider repeating after 1 month, if<br>ore giving antibiotics): culture for group<br>both ASO and anti-DNase B titres, if av   | f negative)<br>A streptococcus.   |  |  |
|   | echocardiography to confirm<br>or refuse the diagnosis of<br>rheumatic carditis.<br>This table is extra / from Davidson  | <ul> <li>18.97 Investigations in a</li> <li>Evidence of a systemic illness (non</li> <li>Leucocytosis, raised ESR and CR</li> <li>Evidence of preceding streptococca</li> <li>Throat swab culture: group A β-from family members and contad</li> <li>Antistreptolysin 0 antibodies (AS levels of &gt; 200 U (adults) or &gt; 3</li> <li>Evidence of carditis</li> <li>Chest X-ray: cardiomegaly; pulm</li> <li>ECG: first- and rarely second-deg pericarditis: T-wave inversion: restart</li> </ul> | <b>n-specific)</b><br>AP<br><b>al infection (specific)</b><br>haemolytic streptococci (also<br>cts)<br>60 titres): rising titres, or<br>60 U (children)<br>nonary congestion<br>gree AV block; features of  |  |  |

| Treatment of | NO specific treatment   |
|--------------|---|
| ARF          | <ul> <li>Now you diagnosed your patient with ARF, what treatment options you can offer him/her?</li> <li>1. Bed Rest : <ul> <li>It's important, as it lessens joint pain and reduces cardiac workload. The duration should be guided by symptoms, along with temperature, leucocyte count and ESR, and should be continued until these have settled.</li> </ul> </li> <li>2. Salicylates : <ul> <li>Like Aspirin, this usually relieves the symptoms of arthritis rapidly and a response within 24 hours helps confirm the diagnosis. We should monitor the patient for toxicity (usually he will have tinnitus and vomiting)</li> <li>3. Penicillin : <ul> <li>Like Procaine Penicillin 4 million units/day x10 days.</li> <li>if the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.</li> </ul> </li> <li>4. Steroids (Prednisolone): <ul> <li>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 (Given when there is severe carditis). There is no evidence that long-term steroids are beneficial.</li> </ul> </li> </ul></li></ul> |
|              | <ul> <li>5. Heart Failure Treatment :         Like diuretics and ACEI.         If heart failure develops, and does not respond to medical treatment, valve replacement may be necessary and is often associated with a dramatic decline in rheumatic activity.     </li> <li><u>Secondary Prevention of Rheumatic Fever (Prevention of Recurrent Attacks) :</u> شرح للصورة اللي تحت : Primary prevention is important by improving socio-economical status         Patients are susceptible to further attacks of rheumatic fever if another streptococcal infection occurs, and long-term prophylaxis with penicillin should be given as benzathine penicillin Long acting penicillin (1.2 million U IM monthly),     </li> </ul>  |
|              | <ul> <li>Further attacks of rheumatic fever are unusual after the age of 21, when treatment may be stopped. However, it should be extended if an attack has occurred in the last 5 years, or if the patient lives in an area of high prevalence or has an occupation (e.g. teaching) with high exposure to streptococcal infection. In those with residual heart disease, prophylaxis should continue until 10 years after the last episode or 40 years of age, whichever is later. Long-term antibiotic prophylaxis prevents another attack of acute rheumatic fever but does not protect against infective endocarditis. "Summarised in the picture"</li> </ul>   |

| Secondary Prevention<br>Attacks)         | of Rheumatic Fever (Preventio                             | n of Recurrent | Duration of Secondary RI   | neumatio   |  |
|--|---|----------------|--|--|--|
| Agent Dose                               |   | Mode           | Prophylaxis  |  |  |
| Benzathine penicillin G                  | 1 200 000 U every 4 weeks*                                | Intramuscular  | Category   | D  |  |
| or<br>Penicillin V<br>or<br>Sulfadiazine | 250 mg twice daily<br>0.5 g once daily for patients 27 kg | Oral           | Rheumatic fever with carditis and residual heart disease (persistent valvular disease) | 10 y since l<br>or until age<br>ever is lon<br>life long pro |  |
|  | 1.0 g once daily for patients >27 kg                      |                | Rheumatic fever with carditis  | 10 yrs or u  |  |
| or individuals aller                     | gic to penicillin and sulfadiaz                           | ine            | But no residual VHD  | (whichever i   |  |
| Erythromycin                             | 250 mg twice daily  | Oral           | Rheumatic fever without carditis   | 5 y or until a<br>( whichever                                |  |
| n high-risk situations, a<br>ecommended  | dministration every 3 weeks is justi                      | fied and       |  | ,  |  |

### B.Chronic Rheumatic heart disease: (In Adults)

- Chronic valvular heart disease develops in at least <u>half</u> of those affected by rheumatic fever with carditis.
- **The mitral valve is affected in more than 70% of cases** (usually Stenosis); the **aortic valve** is the next most frequently involved (40%), followed by the **tricuspid**(10%) and then the pulmonary valve(2%).
- Valve disease may be symptomatic during fulminant forms of acute rheumatic fever but may remain asymptomatic for many years.
- Mitral Stenosis is more common in females (3:1), while males have higher incidence of Aortic Regurgitation.

## **Pathology:**

• The main pathological process in chronic rheumatic heart disease is progressive fibrosis.

**Refresh your memory :** The heart sound is basically "LUB" which is S1 - This sound is produced by closure of Atrioventricular valves- + "DUB" which is S2 - This sound is produced by closure of Aortic and pulmonary valves .

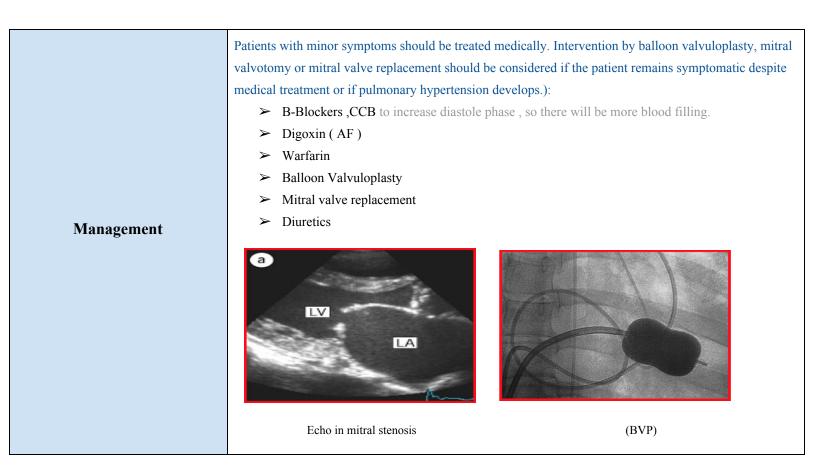
## Watch video!

|       | Mitral Stenosis: The most common valvular abnormality in RHD  |
|-------|---|
| Notes | The flow of blood from LA to LV is <b>restricted</b> and left atrial pressure rises, leading to pulmonary venous<br>congestion and breathlessness. There is dilatation and hypertrophy of the LA, and left ventricular lling<br>becomes more dependent on left atrial contraction.<br>→ The normal mitral valve area (MVA) = 4-6 cm <sup>2</sup><br>→ In severe ms <1.5 cm <sup>2</sup><br>→ High LAP<br>→ The rise in LAP causes a similar rise in pulmonary capillaries, veins and artery.<br>✓ Ventricular diastolic pressure will remain normal because it is protected by the Mitral<br>Stenosis ! |
|       |   |

| Clinical Features: |                | <ul> <li>Dyspnea</li> <li>Fatigue no enough blood bumped to the body to suply it.</li> <li>Palpitation</li> <li>Hemoptysis (10%) due to pulmonary hypertension so the vessels will rupture.</li> <li>Hoarseness (Ortner's syndrome)<sup>2</sup></li> <li>Dysphagia</li> <li>Stroke or peripheral embolization Why ? Left atrium will be enlarged due to stenosis, so the rhythm will be changed from sinus to atrial and the patient usually develops Atrial Fibrillation. Afib will lead to blood clotting that will form a thrombus which travels to the brain causing stroke (and sometimes stroke is the main presentation in a patient with Mitral stenosis).</li> <li>Parasternal heave It's precordial impulse that may be felt in patients with cardiac or respiratory disease.</li> <li>Diastolic thrill A vibration felt over the heart during ventricular diastole. it may be caused due to mitral valve stenosis .</li> <li>Accentuated<sup>5</sup>S1</li> <li>Accentuated S2</li> <li>Opening snap S2 is followed by an opening snap can give an indication as to the severity of the stenosis . the closer the opening snap follows S2 , the worst the stenosis .</li> <li>Mid-diastolic rumble</li> </ul> |
|--------------------|----------------|--|
|                    |                | flush)<br>➤ Tapping apex <sup>4</sup> (S1)   |
|                    | Chest<br>X-ray | <ul> <li>Straightening of the left heart border</li> <li>Double density</li> <li><u>Kerley B lines</u> These are short parallel lines at the lung periphery</li> </ul>   |
| Investigations     | ECG            | LAE (Left Atrial Enlargement), P Mitrale(left atrial enlargement), RV dominance, Atrial Fibrillation   |
|                    | Echodoppler    | <ul> <li>Calcification in MV</li> <li>You can find heart enlargement</li> </ul>  |

 $<sup>^{2}</sup>$  paralysis of the vocal cords, due to the enlargement of pulmonary artery $\rightarrow$  compression of recurrent laryngeal nerve <sup>3</sup> distinctive facial appearance associated with **mitral**stenosis. Someone with **mitral** stenosis may present with rosy cheeks, whilst the rest of the face has a bluish tinge due to cyanosis. ... It is due to low cardiac output, and therefore low perfusion of the facial skin, caused by the stenosis. <sup>4</sup> Tapping apex beat is present only in mitral stenosis where the left ventricular size and filling is less.

<sup>&</sup>lt;sup>5</sup> Very obvious



|                    | Mitral Regurgitation <u>watch video!</u>   |
|--------------------|--|
| Notes              | May also follow mitral valvotomy or valvuloplasty. Chronic mitral regurgitation causes gradual dilatation of the LA with little increase in pressure and therefore relatively few symptoms.  |
| Clinical Features: | <ul> <li>Asymptomatic</li> <li>Dyspnea, orthopnea, and</li> <li>PND(parxosymal nocturnal dyspnia)</li> <li>Displaced PMI<sup>6</sup> تان للتاب بيكون متصنم وبيعمل دسبليسمنت Thrill</li> <li>Soft S1 عكس الستينوسز Pansystolic murmur</li> <li>Mest heard on the lateral side when lying on the side</li> </ul> |

<sup>&</sup>lt;sup>6</sup> The point of maximal impulse (**PMI**) is simply that... the point where there is a maximal impulse against the chest that can be felt. Most often, this is from the apex or tip of the heart: also referred to as the apical impulse. However, in certain conditions, the apex of the heart does not cause the **PMI**.

| Tr | eatm | ient |  |
|----|------|------|--|
|    |      |      |  |

|                       | Aortic Regurgitation Watch video !   |  |  |  |  |  |
|-----------------------|--|--|--|--|--|--|
| Notes                 | This condition is due to disease of the aortic valve cusps or dilatation of the aortic root, The LV dilates and hypertrophies to compensate for the regurgitation. The stroke volume of the LV may eventually be doubled or tripled, and the major arteries are then conspicuously pulsatile. As the disease progresses, left ventricular diastolic pressure rises and breathlessness develops.  |  |  |  |  |  |
| Clinical<br>Features: | <ul> <li>Water-hammer / collapsing pulse</li> <li>Wide pulse pressure (markedly increased systolic BP, with decreased diastolic BP)</li> <li>Corrigan's sign (water-hammer pulse)—rapidly increasing pulse that collapses suddenly as arterial pressure decreases rapidly in late systole and diastole; can be palpated at wrist or femoral arteries)</li> <li>De Musset sign (It's a rhythmic nodding or bobbing of the head in synchrony with the beating of the heart)</li> <li>Muller sign (systolic pulsation of uvula)</li> <li>Quincke's pulse (blanching and flushing, respectively, of the nail bed)</li> <li>Hill's sign (greater systolic BP in the legs than the arms, the difference is &gt;or = 30 mm Hg)</li> <li>★ Early diastolic murmur</li> </ul> |  |  |  |  |  |
|                       | Diastolic murmurs + regurgitation  |  |  |  |  |  |
| Treatment             | Aortic valve Replacement (could be done via Transcathter Aortic Valve Replacement)   |  |  |  |  |  |

| Aortic Stenosis Systolic murmur |   |   |  |  |  |  |  |
|---------------------------------|---|---|--|--|--|--|--|
| Clinical Features:              | <ul> <li>Triad :</li> <li>Angina when the left ventricle enlarges its muscle mass will increase which will increase the oxygen demand and it cant meet the demand because of the stenosis &gt; chest pain . and its the most common presentation.</li> <li>Syncope because there is no enough blood "cardiac output" flow to the brain</li> <li>Dyspnea and PND The worst clinical feature as it indicates HF.</li> </ul> | <ul> <li>Arterial Pulse waveform: Plateau</li> <li>Small (Parvus) diminished carotid upstorke</li> <li>Slow rise (Tardus)delayed carotid upstroke</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 (occurswhenthesplittingis heard during expiration and disappears during inspiration, the opposite of the physiologic split S2)</li> </ul> |  |  |  |  |  |
| Treatment                       | Aortic valve Replacement (could be done via Transcathter Aortic Valve Replacement) why cant we use ballon valvuloplasty like in mitral stenosis ? because the stenosis here is calcification and we cant ballon calcium very well, where as in mitral stenosis it is fibrosis , and we can ballon fibrotic tissue .   |   |  |  |  |  |  |

## **Summary**

AFR : immune-mediated delayed response to infection with specific strains of beta hemolytic streptococci (manifestations after 2-4 weeks), Antibodies produced cause inflammation in the **endocardium, myocardium and pericardium**, as well as the **joints**, **skin**, and **heart valves**.

| ARF overview                                      |  |  |  |  |  |  |
|---|--|--|--|--|--|--|
| Clinical features                                 | Fever, lethargy, anorexia, joint pain, arthritis, carditis, neurological changes.  |  |  |  |  |  |
| Diagnosis   | <ul> <li>★ 2 Major manifestations</li> <li>★ 1 Major and 2 Minor manifestations And Evidence of a recent streptococcal infection.</li> <li>★ 3 Minor</li> <li>Evidence of Preceding GAS Infection:</li> <li>1) Increased ASO titer or Anti-Dnase B titer. 2) positive throat culture.</li> </ul>   |  |  |  |  |  |
| Jones criteria<br>(high/lower risk<br>population) | <ul> <li>Major :</li> <li>1. Cardritis</li> <li>2. Migratory polyarthritis</li> <li>3. Sydenham Chorea</li> <li>4. Subcutaneous Nodule</li> <li>5. Erythema Marginatum</li> </ul>  | <ul> <li>Minor :</li> <li>1. Fever</li> <li>2. Elevated ESR</li> <li>3. Polyarthralgia (major in high risk)</li> <li>4. History of rheumatic fever</li> <li>5. First degree heart block</li> </ul> |  |  |  |  |
| Investigation                                     | <ul> <li>WBC.</li> <li>ESR.</li> <li>CRP.</li> <li>Blood cultures, if febrile.</li> <li>ECG to look for Heart block.</li> <li>Chest X-ray, if clinical or echocardiographic evidence of carditis.</li> <li>Echocardiogram .</li> <li>Throat swab: culture for group A streptococcus.</li> <li>Anti-streptococcal serology: both ASO and anti-DNase B titres</li> </ul> |  |  |  |  |  |
| Treatment   | <ul> <li>&gt; Bed Rest</li> <li>&gt; Salicylates</li> <li>&gt; Penicillin</li> <li>&gt; Steroids</li> <li>&gt; Heart Failure Treatment</li> </ul>  |  |  |  |  |  |

#### **Chronic Rheumatic heart disease : progressive fibrosis**

• The mitral valve is affected in more than 70% of cases (usually Stenosis); the aortic valve is the next most frequently involved (40%), followed by the tricuspid(10%) and then the pulmonary valve(2%)

| Mitral<br>Stenosis          | <ul> <li>venous congestion)</li> <li>Fatigue</li> <li>Palpitation</li> <li>Hoarseness</li> <li>Dysphagia</li> <li>Stroke or peripheral<br/>embolization</li> <li>Cyanosis</li> <li>Tapping apex</li> </ul>  | <ul> <li>Parasternal heave</li> <li>Diastolic thrill</li> <li>Accentuated S1&amp; S2</li> <li>Opening snap</li> <li>Mid-diastolic rumble</li> <li>On X-ray : Straightening of the left heart<br/>border, Double density, Kerley B lines or<br/>pulmonary edema.</li> <li>ECG : LAE, P Mitrale, RV dominance, Atrial<br/>Fibrillation.</li> <li>Echodopple : Calcification in MV</li> </ul> | <ul> <li>Management</li> <li>&gt; B-Blockers ,CCB</li> <li>&gt; Digoxin (AF)</li> <li>&gt; Warfarin</li> <li>&gt; Balloon Valvuloplasty</li> <li>&gt; Mitral valve replacement</li> </ul> |
|-----------------------------|---|--|---|
| Mitral                      | Asymptomatic  |  | Manegement  |
| Regurgitation               | Dyspnea, orthopnea, and   |  | Surgical  |
|                             | PND Displaced PML Thrill  |  |   |
|                             | <ul> <li>Displaced PMI, Thrill</li> <li>Soft S1</li> </ul>  |  |   |
|                             |   |  |   |
| Aortic                      | □ Water-hammer / collapsing pulse   |  | Treatment   |
| Regurgitation               | □ Wide pulse pressure   |  | Aortic valve Replacement  |
|                             | • Corrigan's sign (water  |  |   |
|                             | De Musset sign  |  |   |
|                             | Muller sign   |  |   |
|                             | Quincke's pulse   |  |   |
|                             | Hill's sign   |  |   |
| Aortic<br>Stenosis<br>(SAD) | <ul> <li>Syncope</li> <li>Angina</li> <li>Dyspnea and NPD</li> <li>Arterial Pulse waveform:</li> <li>Small (Parvus)</li> <li>Slow rise (Tardus)</li> <li>Sustained not displaced I</li> <li>Systolic thrill</li> <li>S4</li> <li>Late peaking of murmur</li> <li>Single S2 : Soft or absen</li> <li>Paradoxical splitting of S</li> </ul> | PMI<br>t A2  |   |

•

## <u>Questions</u>

# 1. Which one of the following Organisms is the most common cause for Rheumatic Heart Disease ?

A-group A beta hemolytic streptococcus B-group B alpha hemolytic streptococcus C-staphylococcus aureus D- E.coli

## 2. Which one of the following group of people are mostly affected by Rheumatic Heart Disease

? A- Elderly from 40-60 B- Teenagers from 14-18 C-Children from 5-15 D-New born babies

### 3. Which one of the following Valve is mostly affected by Rheumatic Heart Disease ?

- A- Mitral Valve
- B- Tricuspid Valve
- C- Aortic Valve
- D- Pulmonary Valve

#### 4. Which one of the following diseases is Immunologically Mediated ?

A-Hypertension B-DM C-Hyperlipidemia D- Rheumatic Heart Disease

# 5. Which one of the following serological tests is used to detect recent infection by Group A streptococcus?

- A-Antistreptolysin O antibodies
- B-Antinuclear antibodies
- C-Antimitochondorial antibodies
- D-Antigliadin antibodies

### 6. In order to diagnose Rheumatic Heart Disease, which statement is correct ?

A-Migratory polyarthritis only

- B- polyarthralgia + increase ESR only
- C- Subcutaneous nodules + fever only
- D- Sydenham chorea + Prolonged PR interval + Fever

### 7. The only manifestation of ARF that leaves serious permanent damage is ?

A-Arthritis B-Subcutaneous nodules C-Carditis D-Erythema Marginatum

#### 8. Which one of the following is seen under the microscope and confirm the diagnosis of ARF ?

A-Aschoff Nodules

- B- Lewi Body
- C- Caseous Necrosis
- D- Mallory Body

# 9. In order to treat patient with Streptococcal pharyngitis , which of the following is the best for him ?

- A-penicillin
- B-Ciftriaxone
- C- Ciprofloxacin
- D- Gentmycin

# 10. Which one of the following is the main pathological process in Chronic Rheumatic Heart Disease ?

A-Calcification B-Necrosis C-Amyloidosis D- Progressive Fibrosis

### 11. Which one of the following is Clinical feature for Aortic Regurgitation ?

A- Collapsing pulse B- Syncope C-Dyspnea D- Angina

### 12. Which one of the following is Clinical feature for Mitral Regurgitation ?

A-Early diastolic murmur B-Mid systolic click C-Pansystolic murmur D- Systolic Thrill

### 13. The Radiological feature that is seen on Mitral Stenosis is ?

A-Single Density B-Kaerle B line C-Normal left heart boarder D- Coin lesion

### 14. What is the treatment for Aortic Regurgitation ?

A-Warfarin B-Beta Blockers C-Calcium Channel Blockers D-Aortic valve replacement

### 15.Water-Hammer pulse is feature of which one of the following?

A-Aortic Regurgitation B-Aortic Stenosis C-Mitral Stenosis D-Mitral Regurgitation

### 16. Syncope, Angina and Dyspnea are characters for which disease?

A- Aortic Regurgitation B-Aortic Stenosis C-Mitral Stenosis D-Mitral Regurgitation

#### 17. Enlargement of the Heart can be seen in which of the following?

A-Aortic Regurgitation B- Aortic Stenosis C-Mitral Stenosis D-Mitral Regurgitation

### 18. Which one of the following can treat Mitral Stenosis?

A-Digoxin B-Warfarin C-Mitral Valve Replacement D- All the above

## 19. Which one of the following valve is LESS common affected by Rheumatic Heart

#### **Disease?**

A-Aortic valve B- Mitral valve C-Tricuspid D-Pulmonary

### 20. Which one of the following is Clinical feature for Mitral Stenosis?

A-Opening Snap B-Mid diastolic rumble C- Soft S1 D-A & B

Answer key : 1-A 2-C 3-A 4-D 5-A 6-D 7-C 8-A 9-A 10-D 11-A 12-C 13-B 14-D 15-A 16-B 17-B 18-D 19-D 20-D