

Rheumatic Heart Disease



Objectives :

1. What is ARF And RHD?
2. Diagnosis
3. Jones Criteria & 2015 revision
4. Differential Diagnosis
5. Investigations, Management
6. Rheumatic Valvular Heart Disease
7. Prevention

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Resources :

437 slides, 436 team, Davidson 22nd edition & Kumar 8th edition.

Epidemiologic Background:

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.

- In developed countries the incidence of ARF has declined
- 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).
- **It is 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.
- over past 50 years, incidence ranging 0.2 - 0.64/100,000 (USA).
- 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
- ARF incidence in Kwait 29/100,000, age 5-14 years.
- In Egypt prevalence of RHD, 5/1000 school-children.
- In Yemen 36.5/1000 school-children RHD is A leading cause of CV morbidity & mortality in young people

Global Burden :

| | |
|---------------|--|
| 15.6 Millions | Total cases with RHD |
| 3 Million | Have congestive heart failure. |
| 1 Million | Require Valve surgery |
| 0.5 Million | Annual incidence ¹ of RF, nearly half develop carditis |
| 230,000/YR. | Estimated deaths from RHD: |

3 phases:

- phase 1 : throat infection.
- Phase 2 : RHF (after weeks).
- Phase 3 : RHD (after years).

Why it is important?

- 1- affects young which will cause disability.
- 2- costly → needs cardiac surgery.
- 3- increases mortality.

It imposes a substantial burden on health care systems with limited budgets

A- Acute Rheumatic Fever



It's recommended to watch it before studying this lecture. It is 7:42 minutes.

Incidence

- Acute rheumatic fever usually affects **children (most commonly between 5 and 15 years)** or young adults .
- The **most common cause** of acquired heart disease in childhood and adolescence.

¹New cases per year

Pathogenesis

- The condition is triggered by an **immune-mediated delayed 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.**
- Histologically, fibrinoid degeneration is seen in the collagen of connective tissues.

***(VERY IMP) 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.**



Antischkow cells are enlarged macrophages found within granulomas (called aschoff bodies) associated with the disease. Larger Antischkow cells may coalesce to form multinucleated Aschoff bodies. (See the pic)

* very important to memorize, usually asked about in exams

When the bacteria enter the body it will cause immunological reaction-> the body will produce antibodies against (M protein) which is the component of bacteria and similar structure found in heart, skin, joints and brain.

Clinical Features

- Acute rheumatic fever is a **multisystem disease** 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.
- **Arthritis*** occurs in approximately **75%** of patients. Other features include rashes, **carditis**** and neurological changes.
- 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.

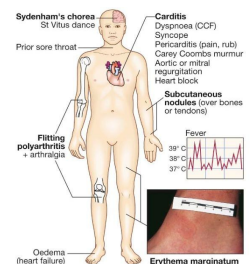
As we mentioned, it's a multisystem disorder that will affect:

- 1) CNS : mainly basal ganglia
- 2) Joints and skin : will recover later
- 3) Heart : permanent damage to the valves.

*not serious because it doesn't leave permanent damage.

**The major problem, we worry about it, because it Destroy the valve and cause permanent damage.

-Murmure in acute attack indicate carditis, HF in acute attack indicate severe reg.



Diagnosis

- **No single test to diagnose ARF.**
- The symptoms and signs are shared by many inflammatory and infectious diseases
- Accurate diagnosis is important, 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
- Diagnosis is **primarily clinical** (it's a syndrome) and is based on a constellation of signs and symptoms, which were initially established as the **Jones criteria.**
- **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 recent infection with a raised **antistreptolysin O (ASO) antibody** titre is helpful.
- 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.

A- Acute Rheumatic Fever

Diagnostic Criteria

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 group A streptococcus pharyngitis.**

1992 Modified Jones criteria Very important

| Major criteria* | Migratory polyarthritis | Carditis | Erythema marginatum | Sydenham chorea | Subcutaneous nodules |
|-----------------|--------------------------------|--------------------------|---------------------|---|----------------------|
| Minor criteria | Arthralgia (Not arthritis!) | First degree heart block | Fever | Elevated inflammatory markers (ESR, CRP) | |

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

* Will be discussed in more detail later on.

2015 Revision of Jones criteria

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

- In accordance with the degree of prevalence of ARF/RHD in the population:**
 - 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
 - Children not from low risk population have been considered to be at moderate or high risk.
- 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.
- Aseptic monoarthritis** has been included as a major criteria in moderate or high risk population.
- Polyarthralgia** has been recognized as a major manifestation for moderate or high risk population
- Fever:** low risk population: >38.5 c, ESR >60 and or CRP > 3mg/dl, while moderate or high risk population: fever >38 and ESR >30 and or CRP > 3mg/dl for

A firm diagnosis requires: 2 Major manifestations or 1 Major and 2 Minor manifestations And Evidence of a recent streptococcal infection. Or 3 Minor. Evidence of Preceding GAS "Group A strept." Infection:

- *Increased or rising ASO titer or Anti-Dnase B titer.
- *A positive throat culture.

| A. For all patient populations with evidence of preceding GAS infection | |
|--|--|
| Diagnosis: initial ARF | 2 Major manifestations or 1 major plus 2 minor manifestations |
| Diagnosis: recurrent ARF | 2 Major or 1 major and 2 minor or 3 minor |
| B. Major criteria | |
| Low-risk populations* | Moderate- and high-risk populations |
| Carditis† | Carditis |
| • Clinical and/or subclinical | • Clinical and/or subclinical |
| Arthritis | Arthritis |
| • Polyarthritides only | • Monoarthritis or polyarthritides |
| | • 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 ≥10 mm in the first hour and/or CRP ≥3.0 mg/dL§ | ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL§ |
| Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion) | Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion) |

- 1) Because pts with immune deficiency have negative ABO
- 2) If you have suspicion of GAS you have to get a culture

A- Acute Rheumatic Fever

Rheumatic Fever Recurrence

- 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 Major Criteria



| | |
|------------------------|--|
| Carditis | <ul style="list-style-type: none">- A 'pancarditis' involves the endocardium, myocardium and pericardium to varying degrees. Its incidence declines with aging, ranging from 90% at 3 years to around 30% in adolescence.- Occurs in 50-70% of cases.- The only manifestation of ARF that leaves serious permanent damage.- May be <i>subclinical</i> (without murmurs so we should use Echo).- Murmurs of MR or AR may occur in acute stage, while mitral stenosis occur in late stage- Cardiomegaly and CHF may occur. (in the acute phase) |
| Arthritis | <ul style="list-style-type: none">- This is the most common major manifestation and occurs early when streptococcal antibody titres are high (present in 35-66% ,Earliest manifestation of ARF).- An acute painful <i>asymmetric and migratory</i> inflammation (Migrating, "Fleeting" 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. Rarely affects the spine.- Duration short < 1 week.- Rapid improvement with salicylates. (Aspirin) The pain characteristically responds to aspirin; if not, the diagnosis is in doubt.- Does not progress to chronic disease. (Doesn't have any late manifestations as the RHD) |
| Sydenham chorea | <ul style="list-style-type: none">- Also known as Saint Vitus's Dance, occur in 10-30%, female predominance.- 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).- 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>).- Spontaneous recovery usually occurs within a few months. Approximately one-quarter of affected patients will go on to develop chronic rheumatic valve disease.. |

Subcutaneous nodules

- Subcutaneous nodules occur in 10% of patients.
- They are small (0.5–2.0 cm), firm non-tender and **painless**.
- Best felt over extensor surfaces of bone or tendons (*Occur over extensor surfaces of joints, on bony prominences, tendons, spine*). Key words (firm non tender and its on extensor surfaces of joints)
- They typically appear more than 3 weeks after the onset of other manifestations and therefore **help to confirm rather than make the diagnosis**
- Short lived: last for few days.
- **Associated with severe carditis.** (When you have it suspect Carditis)



Erythema marginatum

Very few will have this (means fungal infection)

- Erythema marginatum occurs in less than 6% of patients
- 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.
- **Occur mainly on the trunk and proximal extremities** but not the face.
- **Less common, but highly specific manifestation of ARF.**
- **Associated with carditis.**



DDx of ARF

| Presentation | | |
|---|-----------------------------------|--|
| Polyarthritides and fever | Carditis | Chorea |
| Septic arthritis (including disseminated gonococcal infection) [†] | Innocent murmur | Systemic lupus erythematosus |
| Connective tissue and other autoimmune disease ^{††} | Mitral valve prolapse | Drug intoxication |
| Viral arthropathy [*] | Congenital heart disease | Wilson's disease |
| Reactive arthropathy [*] | Infective endocarditis | Tic disorder [‡] |
| Lyme disease [*] | Hypertrophic cardiomyopathy | Choreoathetoid cerebral palsy |
| Sickle cell anaemia | Myocarditis: viral or idiopathic | Encephalitis |
| Infective endocarditis | Pericarditis: viral or idiopathic | Familial chorea (including Huntington's) |
| Leukaemia or lymphoma | | Intracranial tumour |
| Gout and pseudogout | | Lyme disease [*] |
| | | Hormonal [§] |

Investigations

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

***Remember that if you're suspecting ARF or if you've confirmed it, you should do an echocardiography to confirm or refuse the diagnosis of rheumatic carditis.**

^{*}to detect subclinical carditis, subclinical means murmur we can't hear so we use ECG to detect it.

Extra, from davidson

| 18.97 Investigations in acute rheumatic fever |
|--|
| Evidence of a systemic illness (non-specific) |
| • 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) |
| • Antistreptolysin O antibodies (ASO 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 |

Treatment of ARF

NO specific treatment Now you diagnosed your patient with ARF, what treatment options you can offer him/her?

1. **Bed Rest :**

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.

2. **Salicylates :**

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)

3. **Penicillin :**

Like Procaine Penicillin 4 million units/day x10 days. (Know the dose) if the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.

4. **Steroids (Prednisolone):**

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.

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.

Secondary Prevention of Rheumatic Fever (Prevention of Recurrent Attacks) :

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), if compliance is in doubt, or oral phenoxymethylpenicillin (250 mg twice daily).
- Sulfadiazine or erythromycin may be used if the patient is allergic to penicillin; sulphonamides prevent infection but are not effective in the eradication of group A streptococci.
- 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"

(Imp to memorize and for EXAM)

VERY IMP (Doses too are imp!!)

You have to know in which category the pt is in, in order to know the **duration of prophylaxis** and whether or not he/she needs life long prophylaxis

The key difference is the presence of **VHD (Valvular heart disease)**

No residual VHD means the pt can only have fever and carditis and it goes without damage to the valves

| Duration of Secondary Rheumatic Fever Prophylaxis | |
|---|---|
| Category | Duration |
| Rheumatic fever with carditis and residual heart disease (persistent valvular disease) | 10 y since last episode or until age 40y (whichever is longer), sometimes life long prophylaxis |
| Rheumatic fever with carditis But no residual VHD | 10 yrs or until age 21yrs (whichever is longer) |
| Rheumatic fever without carditis <small>Pts under 20 ys give prophylaxis. Pts >20 if he doesnt have evidence of echo dont give him.</small> | 5 y or until age 21 y, (whichever is longer) |

| 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 (60 lb) 1.0 g once daily for patients >27 kg (60 lb) | Oral |
| For individuals allergic to penicillin and sulfadiazine | | |
| Erythromycin | 250 mg twice daily | Oral |
| *In high-risk situations, administration every 3 weeks is justified and recommended | | |

B- Chronic Rheumatic Heart disease

Sequelae of ARF if not treated

- A leading cause of CV morbidity & mortality in young people
- **Chronic valvular heart disease develops in at least half 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. *Most common MS followed by AR*

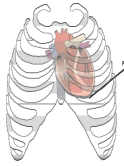
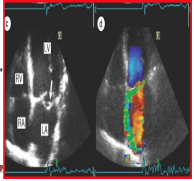
*Frequently asked Q/ Which valve is more commonly involved in chronic rheumatic heart disease?

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 .

Mitral Regurgitation

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 style="list-style-type: none"> - Asymptomatic - Dyspnea, orthopnea, and - PND(paroxysmal nocturnal dyspnea) - Displaced PMI* - Thrill - Soft S1 - Pansystolic murmur Best heard on the lateral side when lying on the side radiating to the axilla |   <p>“Wall hugging”</p> |
| Treatment | Surgically | |

*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.

Mitral Stenosis*



The most common valvular abnormality in RHD

The flow of blood from LA to LV is **restricted** and left atrial pressure rises, leading to pulmonary venous congestion and breathlessness. There is dilatation and hypertrophy of the LA, and left ventricular filling becomes more dependent on left atrial contraction. Ventricular diastolic pressure will remain normal because it is protected by the Mitral Stenosis!

The normal mitral valve area (MVA) = 4-6 cm²

High LAP left atrial pressure

In severe MS <1.5 cm²

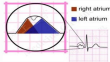
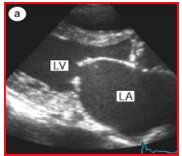
The rise in LAP causes a similar rise in pulmonary capillaries, veins and artery.

*main symptom is SOB.

Clinical features

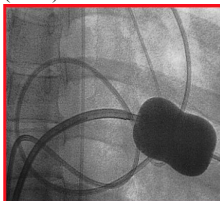
- **Dyspnea** due to pulmonary edema
- **Fatigue** no enough blood pumped to the body to supply it.
- **Palpitation** because of arrhythmias from stretching the LA (A-fib is the most common)
- Hemoptysis (10%) due to pulmonary hypertension so the vessels will rupture. B.c of congested lungs
- Hoarseness (Ortner's syndrome) paralysis of the vocal cords, due to the enlargement of pulmonary artery → compression of recurrent laryngeal nerve
- Dysphagia Pressure of LA
- **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).
- Cyanosis (**Mitral facies²**, **malar flush** (late complication))
- Tapping apex (S1) Tapping apex beat is present only in mitral stenosis where the left ventricular size and filling is less.
- Parasternal heave It's precordial impulse that may be felt in patients with cardiac or respiratory disease. Its either from the pushing of LA or from the hypertrophy of the RV
- Diastolic thrill A vibration felt over the heart during ventricular diastole. it may be caused due to mitral valve stenosis.
- **Accentuated** (very obvious) S1
- Accentuated S2
- **Opening snap** S2 is followed by an opening snap, the distance between S2 and the opening snap can give an indication as to the severity of the stenosis. the closer the opening snap follows S2, the worst the stenosis.
- **Mid-diastolic rumble**

Investigations

| Chest x- ray | ECG  | Echodopler |
|---|---|---|
| <ul style="list-style-type: none"> - Straightening of the left heart border "Mitralization of the left border" - Double density - Kerley B lines These are short parallel lines at the lung periphery | <ul style="list-style-type: none"> - LAE (Left Atrial Enlargement) - P Mitrale (left atrial enlargement) - RV dominance - Atrial Fibrillation | <p>Calcification in MV</p> <p>- GOLD STANDARD</p> <p>- Very specific</p> <p>- "Hockey-stick" appearance</p> <p>You can find heart enlargement</p>  <p>Echo in mitral stenosis</p> |

Management

Patients with minor symptoms should be treated medically. Intervention by balloon valvuloplasty, mitral valvotomy or mitral valve replacement should be considered if the patient remains symptomatic despite medical treatment or if pulmonary hypertension develops.:

- **B-Blockers**, CCB to increase diastole phase, so there will be more blood filling. Slow them down, allow more time for filling of LV (BVP)
 - **Digoxin (AF)** Especially if there is HF and hypotension
 - **Warfarin** to prevent thrombus formation in the LA
 - **Balloon Valvuloplasty** Definitive therapy: you can have up to 3 times but at the end you have to have MV replacement
 - **Mitral valve replacement**
 - **Diuretics** to relieve the congestion
- 

² 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.




Aortic Regurgitation

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.

| | |
|---|--|
| <p>Clinical features (the signs are imp)</p> | <ul style="list-style-type: none"> - Water-hammer / collapsing pulse (very imp) - Wide pulse pressure (markedly increased systolic BP, with decreased diastolic BP) (very imp) - 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) - De Musset sign (It’s a rhythmic nodding or bobbing of the head in synchrony with the beating of the heart) - Muller sign (systolic pulsation of uvula) - Quincke’s pulse (blanching and flushing, respectively, of the nail bed) - Hill’s sign (greater systolic BP in the legs than the arms, the difference is >or = 30 mm Hg) <p>Early diastolic murmur</p> <p style="text-align: center;">Diastolic murmurs + regurgitation</p> |
| <p>Treatment</p> | <p>Aortic valve Replacement (could be done via Transcatheter Aortic Valve Replacement)</p> |

Aortic Stenosis

| | |
|---|--|
| <p>Clinical features</p>  <p>*Thickened and fibrotic</p> | <p>Triad : VERY IMPORTANT</p> <ul style="list-style-type: none"> ❑ 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 > chest pain . and its the most common presentation. ❑ Syncope because there is no enough blood “cardiac output” flow to the brain (low pCO) (exertional) ❑ Dyspnea and PND The worst clinical feature as it indicates HF. <p>Prognosis: is worse with dyspnea, better in syncope and angina Without treatment life expectancy is with:</p> <ul style="list-style-type: none"> - Angina (5 years) - Syncope (3 years) - Dyspnea (1 years) <p>Other clinical features</p> <ul style="list-style-type: none"> - Arterial Pulse waveform: Plateau - Small (Parvus) diminished carotid upstroke Slow rise (Tardus)delayed carotid upstroke Sustained not displaced PMI - Systolic thrill S4 - Late peaking of murmur Single S2 : Soft or absent A2 - Paradoxical splitting of S2 (occurs when the splitting is heard during expiration and disappears during inspiration, the opposite of the physiologic split S2) <p>Aortic stenosis is also seen due to aging calcification.</p> |
| <p>Treatment</p> | <p>Aortic valve Replacement (could be done via Transcatheter Aortic Valve Replacement) why can't we use balloon valvuloplasty like in mitral stenosis ? because the stenosis here is calcification and we can't ballon calcium very well, where as in mitral stenosis it is fibrosis, and we can ballon fibrotic tissue .</p> |

Summary 436

ARF : 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**.

| | | |
|--|--|---|
| Clinical features | Fever, lethargy, anorexia, joint pain, arthritis, carditis, neurological changes. | |
| Diagnosis | <ul style="list-style-type: none"> - 2 Major manifestations - 1 Major and 2 Minor manifestations And Evidence of a recent streptococcal infection. - 3 Minor Evidence of Preceding GAS Infection: 1) Increased ASO titer or Anti-Dnase B titer. 2) positive throat culture. | |
| Jones criteria (high/low risk population) | Major : 1. Carditis 2. Migratory polyarthritis 3. Sydenham Chorea 4. Subcutaneous Nodule 5. Erythema Marginatum | Minor : 1. Fever 2. Elevated ESR 3. Polyarthralgia (major in high risk) 4. History of rheumatic fever 5. First degree heart block |
| Investigation | <ul style="list-style-type: none"> ➤ WBC. ➤ ESR. ➤ CRP. ➤ Blood cultures, if febrile. ➤ ECG to look for Heart block. ➤ Chest X-ray, if clinical or echocardiographic evidence of carditis. ➤ Echocardiogram . ➤ Throat swab: culture for group A streptococcus. ➤ Anti-streptococcal serology: both ASO and anti-DNase B titres | |
| Treatment | <ul style="list-style-type: none"> ➤ Bed Rest ➤ Salicylates ➤ Penicillin ➤ Steroids ➤ Heart Failure Treatment | |

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%)

Summary 436

| Condition | Clinical features | | Management and treatment |
|------------------------------|---|---|--|
| Mitral stenosis | <ul style="list-style-type: none"> ➤ Dyspnea (pulmonary venous congestion) ➤ Fatigue ➤ Palpitation ➤ Hoarseness ➤ Dysphagia ➤ Stroke or peripheral embolization ➤ Cyanosis ➤ Tapping apex ➤ Parasternal heave ➤ Diastolic thrill | <ul style="list-style-type: none"> ➤ Accentuated S1 & S2 ➤ Opening snap ➤ Mid-diastolic rumble ➤ On X-ray : Straightening of the left heart border, Double density, Kerley B lines or pulmonary edema. ➤ ECG : LAE, P Mitrale RV dominance Atrial Fibrillation. ➤ Echo Doppler : Calcification in MV | <p>Management:</p> <ul style="list-style-type: none"> ➤ B-Blockers ,CCB ➤ Digoxin (AF) ➤ Warfarin <small>If there is atrial fibrillation to prevent stroke.</small> ➤ Balloon Valvuloplasty ➤ Mitral valve replacement |
| Mitral regurgitation | <ul style="list-style-type: none"> ➤ Asymptotic ➤ Dyspnea, orthpnea and PND ➤ Displaced PMI, thril ➤ Soft S1 | | <p>Management: Surgical</p> |
| Aortic regurgitation | <ul style="list-style-type: none"> ➤ Water hammer / collapsing pulse ➤ Wide pulse pressure ➤ <u>Corrigan's sign</u> (water-hammer pulse) ➤ De Musset sign ➤ <u>Muller sign</u> ➤ <u>Quincke's pulse</u> ➤ Hill's sign | | |
| Aortic stenosis (SAD) | <ul style="list-style-type: none"> ➤ Syncope ➤ Angina ➤ Dyspnea and PND ➤ Arterial Pulse waveform: 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 | | <p>Treatment: Aortic valve replacement</p> |

Examine Yourself !!

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. Antimitochondrial 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

Examine Yourself !!

8. Which one of the following is seen under the microscope and confirm the diagnosis of ARF ?
- A. Aschoff Nodules
 - B. Lewy 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. Ceftriaxone
 - 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 border
 - D. Coin lesion
14. What is the treatment for Aortic Regurgitation ?
- A. Warfarin
 - B. Beta Blockers
 - C. Calcium Channel Blockers
 - D. Aortic valve replacement

Examine Yourself !!

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
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 - 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

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