Rheumatic Heart disease

Objectives:

- Know that RHD is prevalent in our region and has economic burden.
- Know how to diagnose the disease and how to approach a patient with RHD.
- Know the principles of management.
- Know how to prevent RHD ; who needs prophylaxis and for how long.
- Recognize complications and how to manage them.

[Color index : Important | Notes | Extra]

Resources:

- 435 slides

[Editing file | Feedback | Share your notes | Shared notes | Twitter]

- Done by: Lina Alshehri |Kawthar Almousa | Khawla AlAmmari | Fahad AlAbdullatif
- Team sub-leader: Mana AlMuhaideb
- Team leaders: Khawla AlAmmari & Fahad AlAbdullatif
- Revised by: Ahmed Alyahya

"Medicine is an art, nobody can deny it."

RHD Vs. ARF

Epidemiologic Background:

- 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 improved hygiene, reduced household crowding, and improved medical care.
- 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.** (these patients don't seek medical attention when the get pharyngitis)
- **In underdeveloped countries RHD is the leading cause of CV death during the first five decades of life.**

Global burden:

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

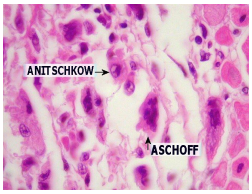
➔ Imposes a substantial burden on healthcare systems with limited budgets.

- What is the difference between rheumatic fever and rheumatic heart disease?

Rheumatic fever is an inflammatory disorder caused by a Group A strep throat infection. It affects the connective tissue of the body, causing temporary, painful arthritis and other symptoms (see JONES criteria below). In some cases rheumatic fever causes long-term damage to the heart and its valves. This is called rheumatic heart disease (the late manifestation). >
RHD is a long-term complication of ARF.

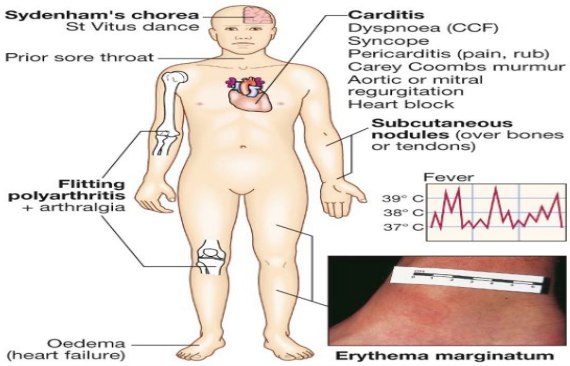
A. Acute Rheumatic fever:

7 : 24 minutes

Incidence and pathogenesis	• Acute rheumatic fever usually affects **children** (most commonly between 5 and 15 years) or young adults but the heart complications don't start to appear until 20 -30yr. • The **most common cause** of acquired heart disease in childhood and adolescence. • 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 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.** • Histologically, fibrinoid degeneration is seen in the collagen of connective tissues. **Aschoff nodules** are pathognomonic and occur only in the heart. They are composed of **multinucleated giant cells** surrounded by macrophages and T lymphocytes, and are **not seen until the subacute or chronic phases of rheumatic carditis.** There is no bacteria  Anitschkow cells are enlarged macrophages found within granulomas (called Aschoff bodies) associated with the disease.Larger Anitschkow cells may coalesce to form multinucleated **Aschoff giant cells**.

Clinical features

- 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.
- **Arthritis** occurs in approximately **75%** of patients. Other features include rashes, carditis and neurological changes.



Diagnosis

- No single test to diagnose ARF.
- The symptoms and signs are shared by many inflammatory and infectious diseases
- Accurate diagnosis is important
- Overdiagnosis will result in individuals receiving unnecessary treatment
- Underdiagnosis may lead to further episodes of ARF causing damage, and the need for valve surgery, and or premature death
- Diagnosis is primarily clinical 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.
- 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 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, along with evidence of preceding streptococcal infection.

2015 Revision of Jones Criteria:

1-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.

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.

★ Introduction of echo have a major role in changing the criteria of Dx.

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 low risk population

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

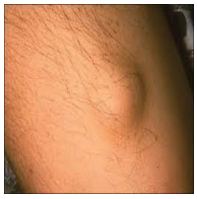
A firm diagnosis requires: 2 Major manifestations or 1 Major and 2 Minor manifestations **And** Evidence of a recent streptococcal infection.

Evidence of Preceding GAS Infection: 1) Increased or rising ASO titer or Anti-Dnase B titer.

2) A positive throat culture.

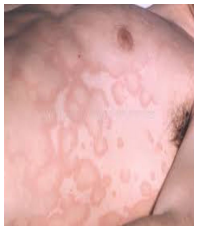
A. For all patient populations with evidence of preceding GAS infection	
Diagnosis: initial ARF	2 Major manifestations or 1 major plus 2 minor manifestations
Diagnosis: recurrent ARF	2 Major or 1 major and 2 minor or 3 minor
B. Major criteria	
Low-risk populations*	Moderate- and high-risk populations
Carditis† • Clinical and/or subclinical	Carditis • Clinical and/or subclinical
Arthritis • Polyarthritis only	Arthritis • Monoarthritis or polyarthritis • 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 ≥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)

- **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 streptococcal infection
- When minor manifestations only present exclude other causes.

Jones criteria (major criteria)	
1-Carditis:	• 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 ***subclinical*** • Cardiomegaly and CHF may occur • Cardiac failure may be due to myocardial dysfunction or valvular regurgitation. • It may manifest as breathlessness (due to heart failure or pericardial effusion), palpitations or chest pain (usually due to pericarditis or pancarditis). ➔ ***Pericarditis may cause chest pain, a pericardial friction rub and precordial tenderness.*** • Other features include tachycardia, cardiac enlargement and new or changed murmurs. A soft systolic murmur due to mitral regurgitation is very common. A soft mid-diastolic murmur (the Carey Coombs murmur) is typically due to valvulitis, with nodules forming on the mitral valve leaflets. • Aortic regurgitation occurs in 50% of cases but the tricuspid and pulmonary valves are rarely involved ➔ ***Murmurs of mitral regurgitation or Aortic regurgitation may occur in acute stage while mitral stenosis occurs in late stages.*** • ECG changes commonly include ST and T wave changes. Conduction defects sometimes occur and may cause syncope.
2-Arthritis:	• **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 *asymmetric and migratory 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. • Duration short < 1 week. • Rapid improvement with salicylates. The pain characteristically responds to aspirin; if not, the diagnosis is in doubt. • **Does not progress to chronic disease.** ★ **Carditis and arthritis are the most common manifestations.** ★ **Although it does not leave serious permanent damage, it may leave deformity.**
3-Sydenham Chorea:	• 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 (*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.. • You have to exclude other diseases eg.huntington's, wilson's, encephalopathies.
4-Subcutaneous Nodules:	• Subcutaneous nodules occur in 5–7% 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).* 

- **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.**

5-Erythema Marginatum:



- Erythema marginatum occurs in less than 5% 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.**

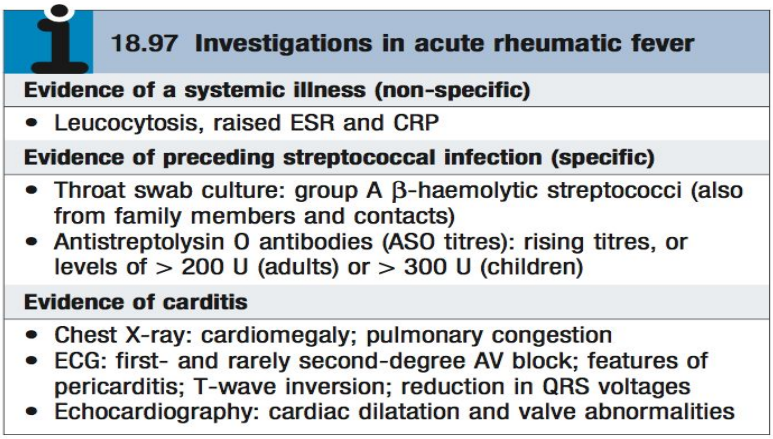
DDX of ARF : (DIFFERENTIAL DIAGNOSIS OF ACUTE RHEUMATIC FEVER)

Presentation		
Polyarthritis 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.
- ➔ 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).
- ➔ Chest X-ray, if clinical or echocardiographic evidence of carditis.
- ➔ Echocardiogram (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.**



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
- Echocardiography: cardiac dilatation and valve abnormalities

Treatment of ARF

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.
   -75-100 mg /kg/day given as 4 divided doses for 6-8 weeks.
   -Attain a blood level 20-30 mg/dl
3. **Penicillin :**
   Like Procaine Penicillin 4 million units/day x10 days, if the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.
4. **Steroids (Prednisolone):**
   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. 2mg/kg/day tapered over 6 weeks
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. AV block is seldom progressive and pacemaker insertion rarely needed.

We don't have a specific treatment for ARF, so we give salicylates in large doses, and if no response, give steroids).

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

- 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 (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.

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

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 ,(which-ever 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	5 y or until age 21 y, (whichever is longer)

B.Chronic Rheumatic heart disease:

- Chronic valvular heart disease develops in at least half of those affected by rheumatic fever with carditis.
- Two-thirds of cases occur in women. Some episodes of rheumatic fever pass unrecognised and it is only possible to elicit a history of rheumatic fever or chorea in about half of all patients with chronic rheumatic heart disease.
- **The mitral valve is affected in more than 90% of cases**; the **aortic valve** is the next most frequently 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**.
- The heart valves are predominantly affected but involvement of the pericardium and myocardium may contribute to heart failure and conduction disorders.
- 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.
- Once a valve has been damaged, the altered haemodynamic stresses perpetuate and extend the damage, even in the absence of a continuing rheumatic process.

Mitral Stenosis: The most common valvular abnormality in RHD

Notes

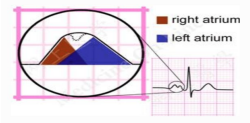
The flow of blood from LA to LV is **restricted** and left atrial pressure rises, leading to pulmonary venous congestion and breathlessness. There is dilatation and hypertrophy of the LA, and left ventricular lling becomes more dependent on left atrial contraction. MS murmurs

- ➔ **The normal mitral valve area (MVA) = 4-6 cm^2**
- ➔ **In severe ms <1.5 cm^2**
- ➔ **High LAP**
- ➔ **The rise in LAP causes a similar rise in pulmonary capillaries, veins and artery.**

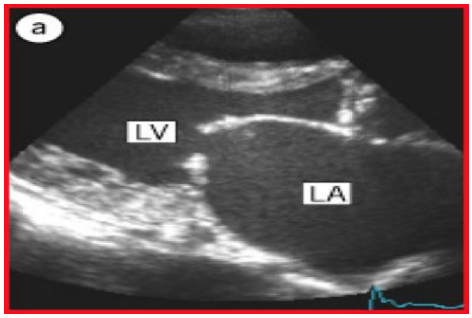
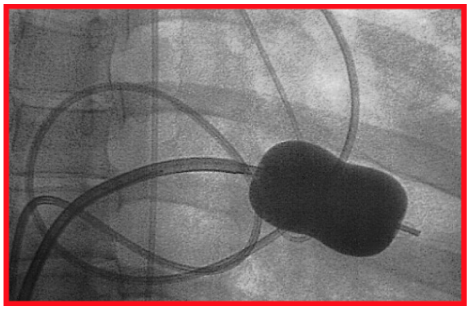
Clinical Features:

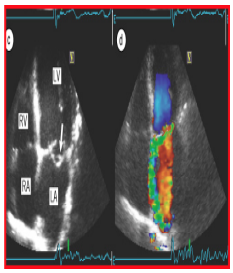
- ❑ **Dyspnea**
- ❑ **Fatigue**
- ❑ **Palpitation**
- ❑ Hemoptysis (10%)
- ❑ Hoarseness (Ortner's syndrome)[1]
- ❑ Dysphagia (Dilated atrium > compressession on the esophagus)
- ❑ **Stroke or peripheral embolization**
- ❑ Cyanosis (Mitral facies, malar flush)
- ❑ Tapping apex (S1)(Tapping apex beat is present only in mitral stenosis where the left ventricular size and filling is less)
- ❑ Parasternal heave(It's a precordial impulse that may be felt in patients with cardiac or respiratory disease.)
- ❑ Diastolic thrill(a vibration felt over the heart during ventricular diastole. It may be caused by mitral valve stenosis)
- ❑ Accentuated[2] S1
- ❑ Accentuated S2 (When pulmonary hypertension develops)
- ❑ 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 worse is the stenosis)
- ❑ Mid-diastolic rumble

Investigations

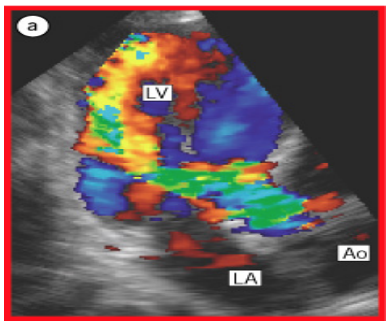
Chest X-ray	• Straightening of the left heart border • Double density • Kerley B lines • Calcification in MV
ECG	LAE (Left Atrial Enlargement), P Mitrale (A notch -broken line- near its peak may or may not be present, see the pic), RV dominance 
Echodoppler	-

[1] paralysis of the vocal cords, due to the enlargement of pulmonary artery→ compression of recurrent laryngeal nerve
[2] Very obvious

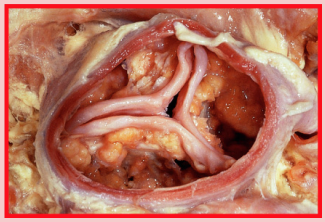
Management	Patients with minor symptoms should be treated medically. Intervention by balloon valvuloplasty, mitral valvotomy or mitral valve replacement should be considered if the patient remains symptomatic despite medical treatment or if pulmonary hypertension develops.): - B-Blockers ,CCB (to increase diastole phase, so more blood filling) - Digoxin (AF) - Warfarin - Balloon Valvuloplasty - Mitral valve replacement  Echo in mitral stenosis  (BVP)

Mitral Regurgitation	
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:	❑ Asymptomatic ❑ Dyspnea, orthopnea, and PND ❑ Displaced PMI, Thrill ❑ Soft S1 ❑ Pansystolic murmur 
Treatment	Surgical

Aortic Regurgitation

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 trebled, and the major arteries are then conspicuously pulsatile. As the disease progresses, left ventricular diastolic pressure rises and breathlessness develops.
Clinical Features:	❏ Water-hammer / collapsing pulse ❏ Wide pulse pressure (markedly increased systolic BP, with decreased diastolic BP) ❏ 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 head in synchrony with the beating of the heart) ❏ Muller sign (uvula pulsating) ❏ Quincke's pulse ❏ Hill's sign (in aortic insufficiency, greater systolic blood pressure in the legs than in the arms; normal arterial systolic pressure in the leg is 10-20 mmHg above that in the arm, whereas in aortic insufficiency the difference may be 60-100 mmHg.) *Displaced PMI (down and to the left) and S3 may also be present. Muller sign Corrigan's sign Quincke's pulse 
Treatment	Aortic valve Replacement (could be done via Transcathter Aortic Valve Replacement)

Aortic Stenosis

Clinical Features: 	❏ Angina (Hypertrophy of left ventricle, and no enough blood → ischemia) ❏ Syncope (usually exertional, cause blood is going to the muscles more than brain) ❏ Dyspnea	❏ Arterial Pulse waveform: Plateau ❏ Small (Parvus)(diminished carotid upstrokes) ❏ Slow rise (Tardus)(delayed carotid upstrokes) ❏ Sustained not displaced PMI ❏ Systolic thrill ❏ S4 (stiff hypertrophy of ventricles) ❏ 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)
Treatment	Aortic valve Replacement (could be done via Transcathter Aortic Valve Replacement)	

Questions

1. A 72-year-old man comes to the office with intermittent symptoms of dyspnea on exertion, palpitations, and cough occasionally productive of blood. On cardiac auscultation, a low-pitched diastolic rumbling murmur is faintly heard at the apex. What is the most likely cause of the murmur?

A. Rheumatic fever as a youth
B. Long-standing hypertension
C. A silent MI within the past year
D. A congenital anomaly
E. Anemia from chronic blood loss

2. 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:

- ➔ **Hct: 42%**
- ➔ **WBC: 12,000/µL with 80% polymorphonuclear leukocytes, 20% lymphocytes**
- ➔ **ESR: 60 mm/h**
- ➔ **ECG : Shows PR prolongation**

Which of the following tests is most critical to diagnosis?

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

3. What is the typical splitting pattern of the S2 heart sound in a patient with aortic valve stenosis?

A. Physiologic splitting
B. Paradoxical splitting
C. Fixed splitting
D. Widened splitting

4. How does the murmur of mild aortic regurgitation differ from that of severe aortic regurgitation (AR)?

A. Mild aortic regurgitation is high pitched and severe aortic regurgitation is low pitched
B. Mild aortic regurgitation is crescendo and severe aortic regurgitation is decrescendo
C. The intensity of the murmur is louder in severe aortic regurgitation
D. Severe aortic regurgitation is shorter in duration compared to mild aortic regurgitation

5. What is the most common cause of mitral stenosis?

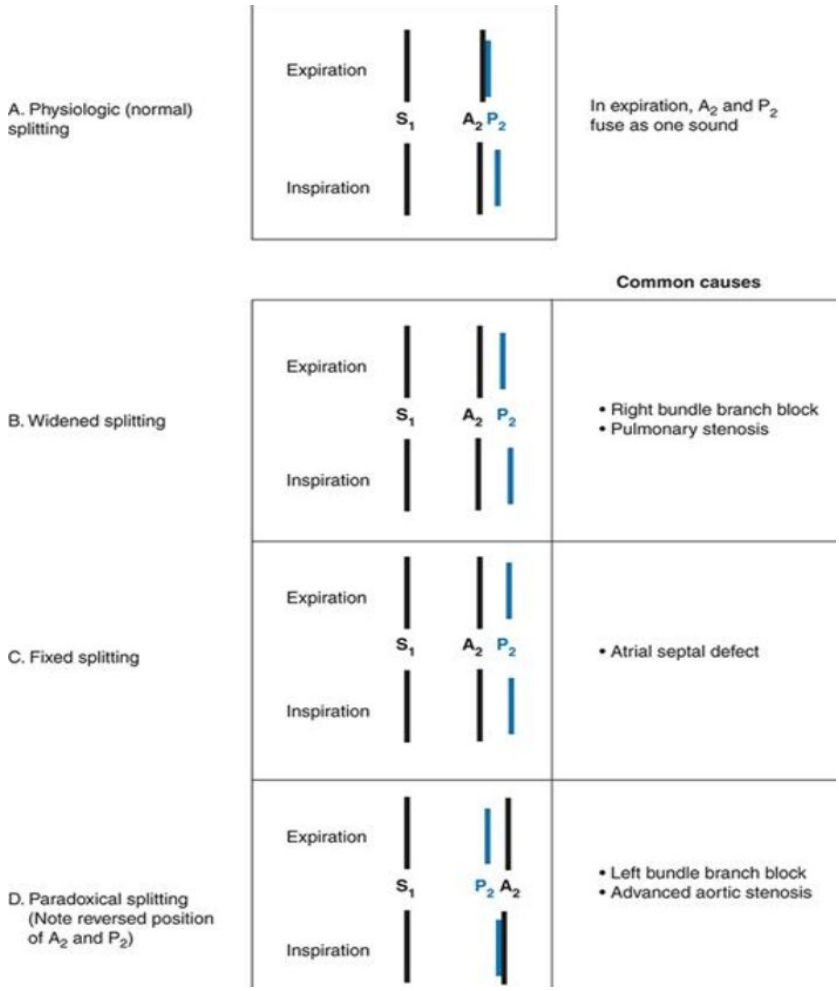
A. Congenital defects
B. Pulmonary Hypertension
C. Rheumatic fever
D. Calcium deposits

Answers: 1.A 2.B 3.B 4.D 5.C

1.A . The history and physical examination findings suggest mitral stenosis. Dyspnea may be present secondary to pulmonary edema; palpitations are often related to atrial arrhythmias (PACs, SVT, atrial flutter, or fibrillation); hemoptysis may occur as a consequence of pulmonary hypertension with rupture of bronchial veins. A diastolic rumbling apical murmur is characteristic. If the patient is in sinus rhythm, a late diastolic accentuation of the murmur occurs because of increased flow across the mitral valve with atrial contraction. A loud first heart sound and early diastolic opening snap may also be present. The etiology of mitral stenosis is usually rheumatic, rarely congenital. Hypertension may cause an S4 gallop but not a diastolic murmur. Myocardial infarction may cause mitral regurgitation because of papillary muscle dysfunction and anemia may cause a pulmonic flow murmur; both of these are systolic murmurs.

2.B. This 18-year-old presents with features of rheumatic fever. Rheumatic fever is diagnosed according to the Jones criteria. Evidence of recent streptococcal infection plus two major manifestations or one major and two minor manifestations satisfy the Jones criteria for diagnosis of acute rheumatic fever. Major criteria include carditis, polyarthritis, chorea, erythema marginatum, and subcutaneous nodules. Minor manifestations include fever, polyarthralgia, elevated erythrocyte sedimentation rate, and PR prolongation on ECG. This patient's clinical manifestations include arthritis, fever, and murmur (consistent with mitral regurgitation). The rash suggests erythema marginatum, and a subcutaneous nodule is noted. Rheumatic subcutaneous nodules are pea sized and usually overlie extensor tendons. The rash is usually pink with clear centers and serpiginous margins. Laboratory data include an elevated erythrocyte sedimentation rate. The ECG shows evidence of first-degree AV block. An antistreptolysin O antibody is necessary to document prior streptococcal infection. Endocarditis (for which blood cultures and an echocardiogram would be ordered) might cause fever, joint symptoms, and the tender nodule but would not account for the diastolic murmur or the characteristic skin lesion. There is no evidence of lupus or myocardial infarction.

3.B



4.D.Mild aortic regurgitation causes a long, soft early diastolic decrescendo murmur. As the regurgitation becomes more severe, the pressure between the aorta and left ventricle equalizes more quickly and the murmur becomes significantly shorter, although the intensity can increase slightly.
The murmur of aortic regurgitation (mild or severe) is a high-pitched, early diastolic decrescendo murmur usually heard best at the 3rd intercostal space on the left (Erb's point) at end expiration with the patient sitting up and leaning forward. If the AR is due to aortic root disease, the murmur will be best heard at the right upper sternal border and not at Erb's point.

In patients with AR, an early diastolic rumble may also be heard at the apex due to the regurgitant jet striking the anterior leaflet of the mitral valve causing it to vibrate. This murmur is termed the Austin-Flint murmur.

In addition to the above two murmurs, a systolic ejection murmur may be present in people with severe AR at the right upper sternal border simply due to the large stroke volume passing through the aortic valve with each systolic contraction of the LV.

5.C. The most common cause of mitral stenosis is rheumatic fever.