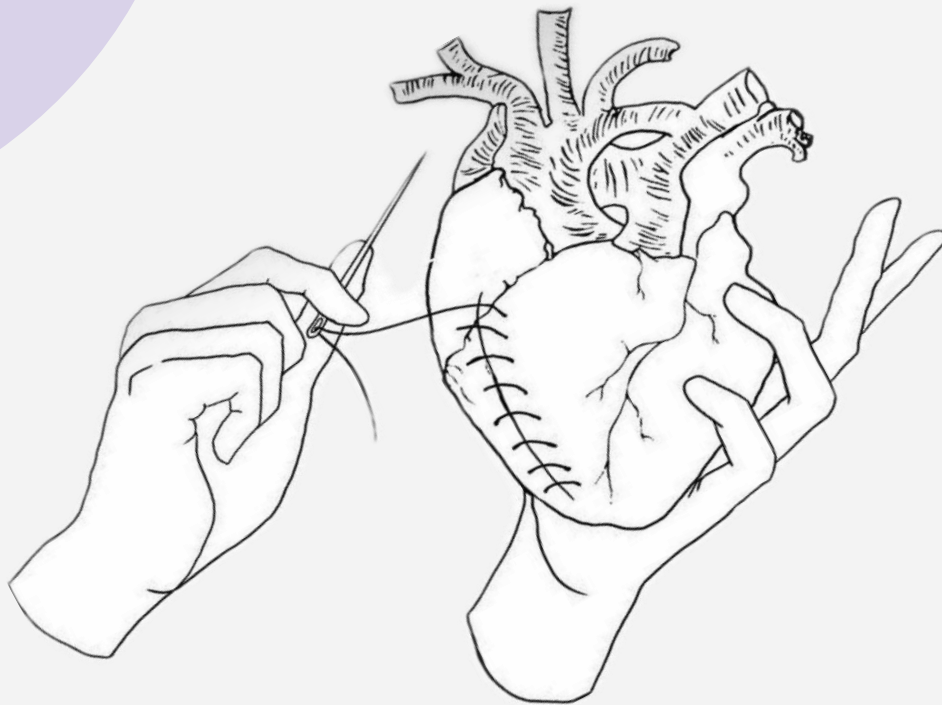




Rheumatic fever, Endocarditis & Heart valves



[Editing file:](#)

COLOR INDEX:

MAIN TEXT (BLACK)

FEMALE SLIDES (PINK)

MALE SLIDES (BLUE)

IMPORTANT (RED)

DR'S NOTE (GREEN)

EXTRA INFO (GREY)

Objective

استعن بالله

Click here for the [objectives file from the male's doctor](#)



Understands the clinicopathological features of rheumatic heart disease which is a major cause of acquired mitral and aortic valve diseases in the Kingdom of Saudi Arabia.



Know the pathological causes and pathophysiological consequences of stenosis and incompetence of all the cardiac valves but particularly the mitral and aortic valves.



Understands the pathology of infective endocarditis so as to be able to identify patients at risk and when appropriate ensure prophylactic treatment is given.

Key principles to be discussed :

1-Pathology and manifestations of rheumatic heart disease as a major cause of valvular diseases in the Middle East and Saudi Arabia.

2-Complications of rheumatic heart disease including atrial fibrillation, valvular and atrial thrombus formation with systemic embolism, cardiac failure and infective endocarditis.

3-Infective endocarditis: predisposing factors, clinical acute and subacute forms, common pathogenic bacteria in IE and complications including valve perforation, thrombosis and septic embolization of the vegetations.

4-Causes and consequences of valvular heart disease with special emphasis on aortic and mitral valve including "floppy or prolapsed" mitral valve.

If you want to read the lecture from Robbins [click here](#)



Acute Rheumatic Fever

Acute Rheumatic Fever

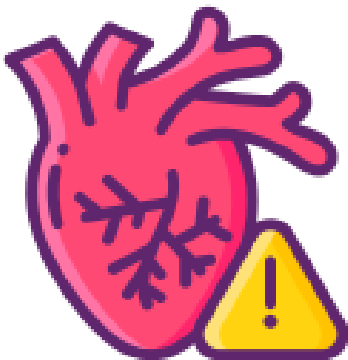
an acute, **immune mediated, multi-system** inflammatory disease that occurs a few weeks **after, group A-beta hemolytic streptococcal pharyngitis infection (with Sore throat)**.
It is an **acute post-streptococcal non-suppurative inflammatory disease (which means there is no pus, ,no abscess.)**with cardiac and extracardiac manifestations.

The inflammation is mainly in the heart, joints, central nervous system and skin.

Occurs in only 3% of patients with group A streptococcal pharyngitis mainly in children, 5 to 15 years of age **but first attacks can occur in adults.**

It's the major health problem in 3rd world countries and in crowded, low socioeconomic urban areas.

The incidence and mortality of rheumatic fever has declined over the past 30 years (due to improved socioeconomic condition and rapid diagnosis and treatment **(antibiotics)**of strep. pharyngitis)





Acute Rheumatic Fever

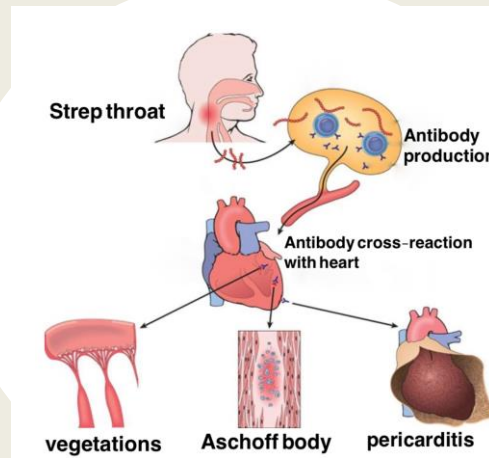


[click here](#)

Etiopathogenesis :

Etiopathogenesis Not yet clear & not completely understood

It is linked to streptococcal infection. Disease occurs 1 to 5 weeks after pharyngeal infection by Group A beta - Hemolytic Streptococcus.



It is most likely an immune mediated process in which--> the causative organisms (streptococci) stimulates in the **formation of antibodies**--> these antibodies cross react with certain antigens present in the heart and joints --> the antigen antibody reaction leads to inflammation.

Repeated attacks or a single severe attack can lead to--> chronic rheumatic heart disease leading to -> cardiac failure.

Rheumatic Heart disease

Rheumatic Heart disease Is A heart disease caused by Rheumatic fever (RF).

Can be acute or chronic.

CARDIAC MANIFESTATIONS OF Acute RHEUMATIC FEVER also called
“acute rheumatic heart disease” or “acute rheumatic
carditis/pancarditis



Patients present with pancarditis inflammation in all 3 layers of heart: endocardium - myocardium - pericardium.

Pericarditis

Inflammation of Pericardium: **fibrinous / serofibrinous** secretion in pericardium between visceral & parietal layers. like butter between two slices bread and therefore also called **Bread & Butter pericarditis**

Myocarditis

inflammation of myocardium many Aschoff bodies. **Can cause sudden death.**

Subendocardial lesions

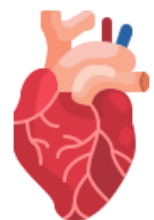
can also be seen, commonly in the left atrium called as **MacCallum plaques.**

Endocarditis

inflammation of the endocardium including the **heart valves (valvulitis) and chordae tendineae**-> results in fibrin deposition on valve leaflets-> forming tiny thrombi along lines of closure called rheumatic vegetations.

Mitral and aortic valve are mainly involved.

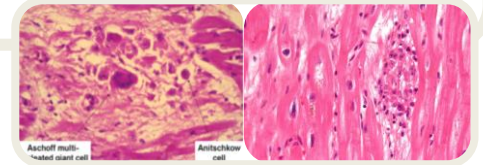
This acute inflammation may either resolve completely or progress to **scarring (with murmur)** with development of chronic fibrotic deformities of the heart valves and chordae tendineae leading to chronic rheumatic heart disease many years later



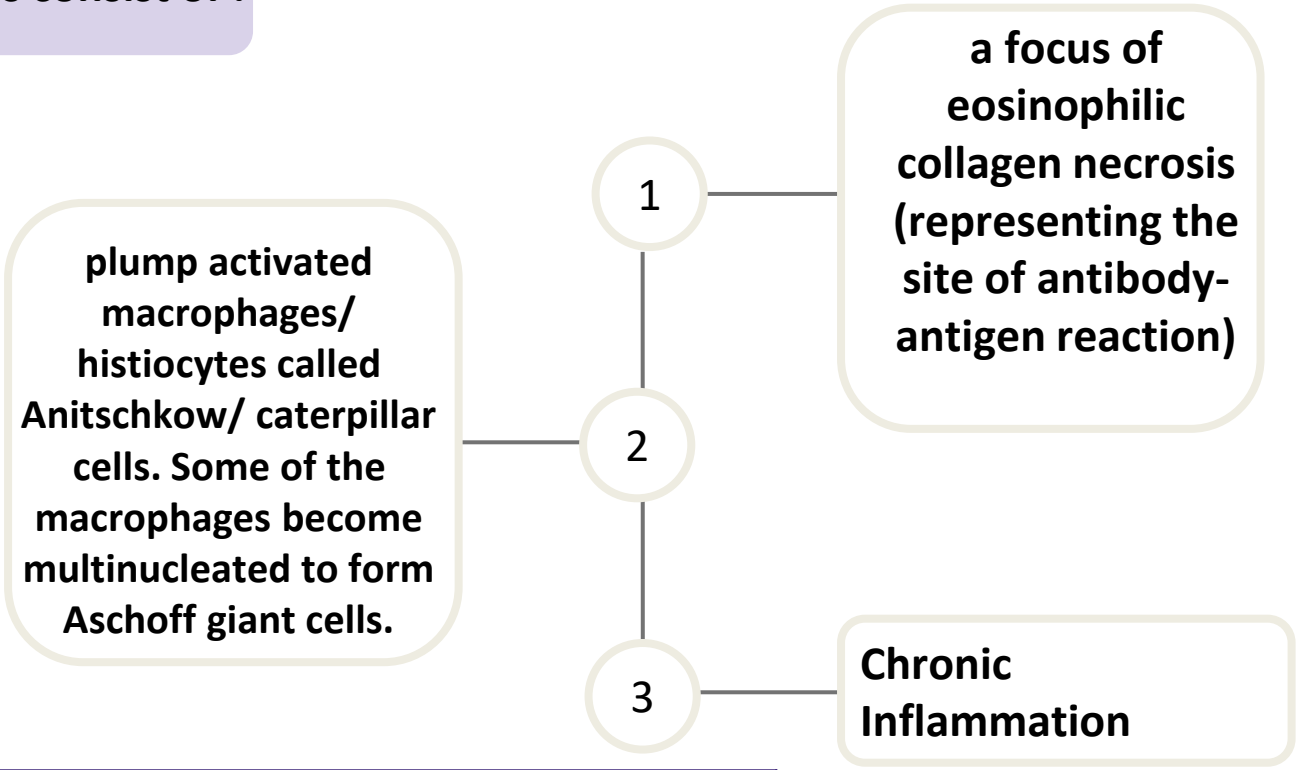


Aschoff Bodies

Aschoff bodies : are multiple **tiny granulomatous** lesions of the heart. They are situated next to small arteries and are characteristically seen in the myocardium (rheumatic myocarditis).



Aschoff Bodies consist of :



Site :	They are found mainly in the myocardium and pericardium. Uncommon in the endocardium and heart valves.
Heal by :	They ultimately heal by fibrosis resulting in a nodule of scar tissue.

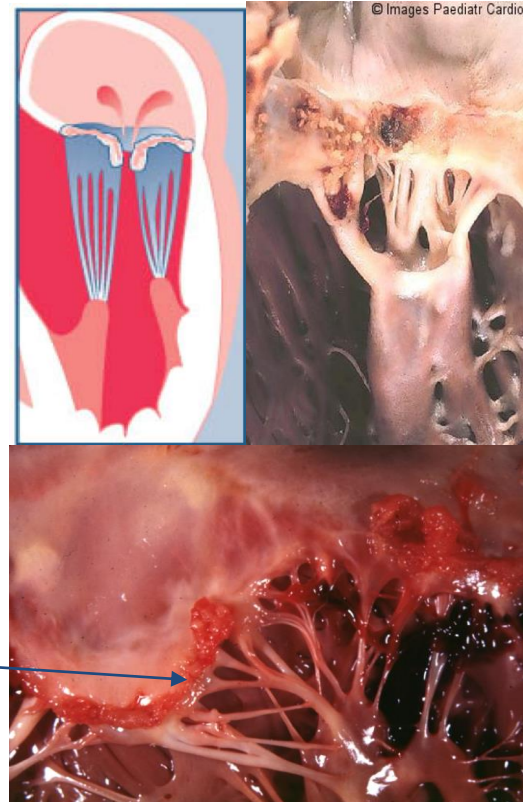
Rheumatic Vegetation

1 Tiny (size of pin's head), sessile (immobile) arranged in a row & firmly with underlying tissue.

2 Situated (located) in the valve cusp, a few millimeters away from free margin (traumatized area).

3 An abnormal growth of tissue around a valve

Heal with fibrosis causes problems in heart valves



Extracardiac Manifestations

1

Joints:

1-Arthralgia

2. Migratory polyarthritis : which is fleeting arthritis in the **large joints** e.g. knee, ankle, elbow wrist etc. It is self limiting with no chronic deformities. Aschoff bodies may be present in the synovial membrane, joint capsule, ligament etc. with joint effusion.

Dr's note :

Arthralgia= pain

arthritis= pain+ swelling+ redness

Migratory =the pain migrates.

(poly)arthritis= more than one joint

And she give an example "so the patient may say yesterday I had pain in my knee and today I feel it in the hip"

Extracardiac Manifestations

2

Skin: skin nodules → erythema marginatum

Subcutaneous tissue: Rheumatic nodules mainly seen over the bony prominences e.g. knuckle, elbow, patella etc.



3

Neurological disorder: uncommon Sydenhem's chorea (St. Vitus' dance) characterized by series of rapid involuntary purposeless movements of the face and arms. This occurs late in the disease.



4

Lung: uncommon, chronic interstitial inflammation and fibrinous pleuritis.



Clinical features of Acute Rheumatic Fever

- Peak incidence: 5-15 years
- History of sore throat: symptoms start 10 days to 6 weeks after by group A Streptococcal pharyngitis
 - By that time the symptoms start the throat/ pharangeal cultures are usually negative.
 - Acute symptoms usually subside within 3 months
 - The mortality from acute rheumatic carditis is low.

Clinical features of Acute Rheumatic Fever

How do we know that patient has a strep infection if the cultures shows negative ? -> Serum antistreptolysin O (ASO titer/ antibodies to group A streptococcal antigens), anti-DNAase B, and antihyaluronidase are raised and provide evidence of a recent infection with group A Streptococcus.

There is no specific test for rheumatic fever
Serologic evidence of a previous streptococcal
 The diagnosis is made based on the clinical findings when either:

**two major or
 one major and two minor**

clinical features / criteria are met. This is called as the **Jones criteria**.



Jones criteria

Acute rheumatic mitral valvulitis superimposed on chronic rheumatic heart disease.

Major criteria/ clinical features	Minor criteria/clinical features
Migratory polyarthritis of the large joints	Previous rheumatic fever
Erythema marginatum of the skin	Arthralgia
Subcutaneous nodules	Fever
Sydenhem's chorea (St. Vitus' dance)	EKG changes ECG=EKG
Carditis (murmurs, pericardial friction rubs, weak heart sounds, tachycardia and arrhythmias cardiomegaly, pericarditis, and congestive heart failure)	Lab tests indicative of inflammation — elevated acute phase reactants elevated ESR (erythrocyte sedimentation rate), CRP (C-Reactive protein), leukocytosis

a neurologic disorder with involuntary purposeless, rapid movements



CHRONIC RHEUMATIC HEART DISEASE

FEMALES SLIDES

1

Organization of the acute inflammation and subsequent scarring •Aschoff bodies are replaced by fibrous scar, so diagnostic forms of these lesions are rarely seen in chronic RHD

2

The myocarditis and pericarditis components of RF typically resolve **without permanent sequelae**.

3

The acute valvulitis or chordae tendinitis of rheumatic fever **heals by fibrosis (scarring)** and result in **irreversible** deformity of the involved cardiac valve and chordae tendineae.
Severe valvular scarring develops months or years after acute RF.

4

Most harmful effect of rheumatic disease is due to involvement of cardiac valves.
The valve leaflets develop **diffuse fibrosis, become thickened, shrunken and less movable** which can lead to **cardiac failure, thromboembolism and infective endocarditis**.

Valves affected in chronic rheumatic heart disease:
left side of heart is more commonly involved than the right.

Mitral valve alone is most **commonly affected** (70%)

combined mitral/aortic valve
-> after mitral valve damage (25%)

Tricuspid valve is rarely affected
May it caused after IV fluid

Pulmonary valve is **practically never affected**

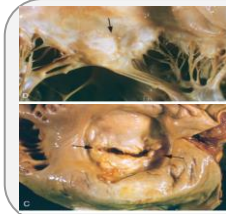
Therefore patient can have **mitral stenosis (most common)**, mitral regurgitation, aortic stenosis and aortic regurgitation

CHRONIC RHEUMATIC HEART DISEASE

The major functional consequence of RHD is:

Type of damage

regurgitation
(improper closure)
fibrosis of chordae
tendineae



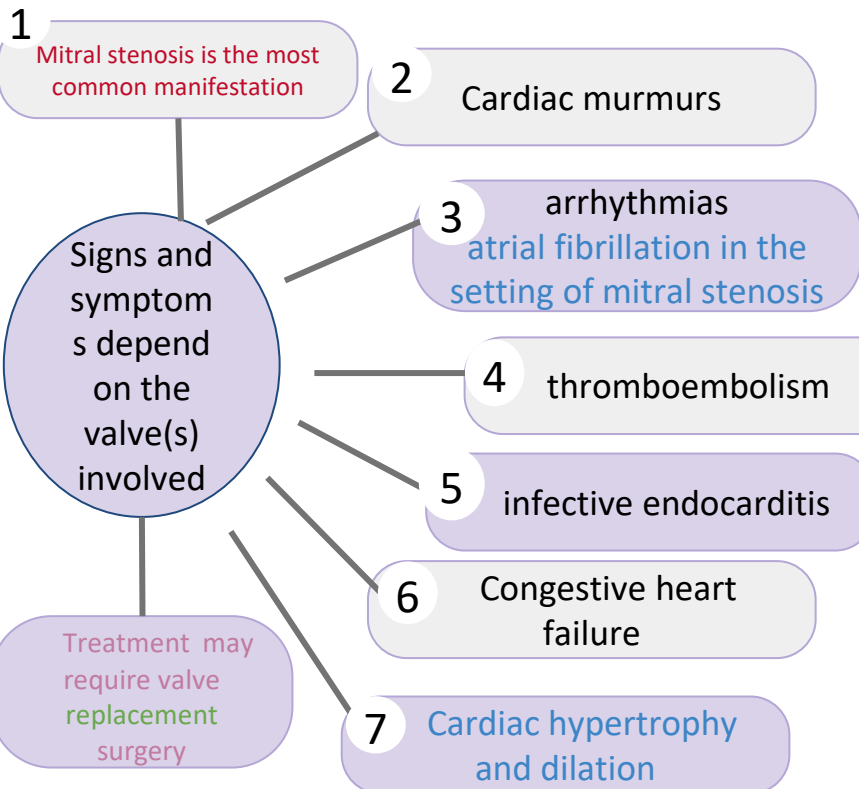
- Mitral stenosis with diffuse fibrous thickening and distortion of the valve leaflets, commissural fusion (arrows)
- Thickening and shortening of the chordae tendineae

stenosis
(Reduction of diameter)
fibrosis of valve leaflets

Clinical features :

- More likely to occur when the first attack:
 - In early childhood
 - Severe
 - Recurrence
- The long term prognosis is highly variable

Occurs many years after the initial episode of rheumatic fever →

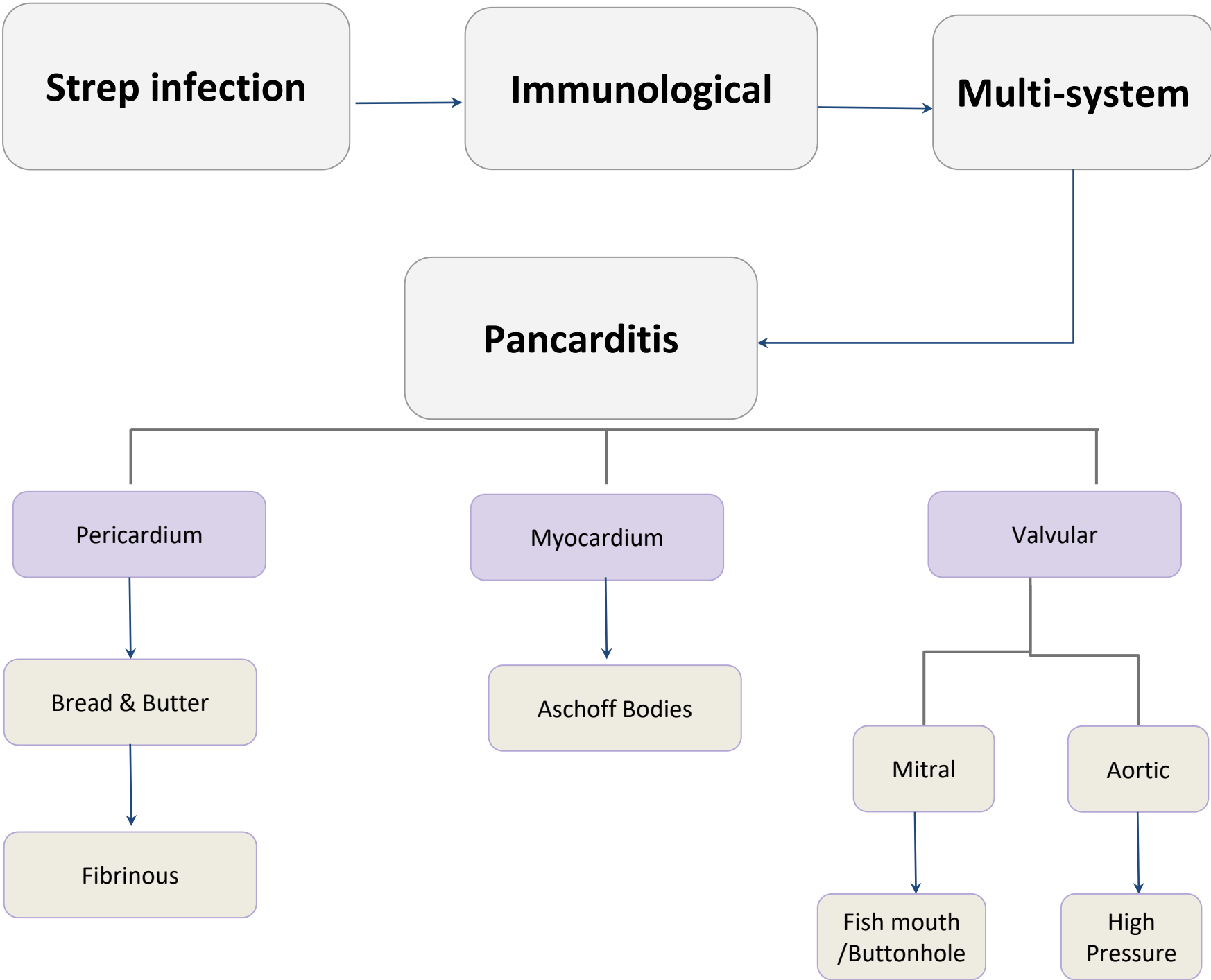


Complications

- 1 **Bacterial infective endocarditis:** the scarred valves of rheumatic heart disease provide an attractive environment for bacteria to grow.
- 2 **Mural thrombi** form in cardiac chambers. They give rise to **thromboemboli**, which can produce infarcts in various organs.
- 3 Congestive heart failure
- 4 Adhesive pericarditis
- 5 Atrial fibrillation

Rheumatic Heart disease

Sequence of Events





Infective Endocarditis

mixed slide between **M** and **F slides**

Definition

infection of the cardiac valves or inner (mural) surface of the endocardium, resulting in the formation of an adherent mass of thrombotic debris that contains microorganisms.

Infective endocarditis is a particularly difficult infection to eradicate because of the avascular nature of the heart valves

A serious infection, characterized by:

- 1 Microbial invasion of heart valves or mural endocardium
- 2 Often with destruction of the underlying cardiac tissues
- 3 **Vegetations on heart valves** are bulky, friable vegetations composed of necrotic debris, thrombus, and organisms

hallmark of IE

Prognosis

depends to some extent on the infecting organism and the stage at which the infection is treated. About 1/3rd of cases of Staph. aureus endocarditis are still fatal

Infective Endocarditis/ Two types



Types	Acute	Subacute
Causative Organism	high virulent staph.aureus	low virulent strept.viridans
Infects what?	Can attack a previously normal valve	Colonize a previously abnormal heart, especially when there are deformed valves
Progression	Rapidly	Slowly
Type of reaction that happens	Has little local host reaction.	It induces a local inflammatory reaction
Prognosis	Death within days to weeks in more than 50% of patients despite antibiotics and surgery	Follows a protracted course of weeks to months Most patients recover after appropriate antibiotic therapy

Infective Endocarditis

Pathogenesis only in M Slides

1

Bacteremia is a pre-requisite, the source could be:

Other organs infection:

- IV drug abuse:
- Usually *Staph aureus*, (right heart side valve affected; Tricuspid)
- Dental or surgical procedure
- Trivial injury, skin, gut, urinary bladder

2

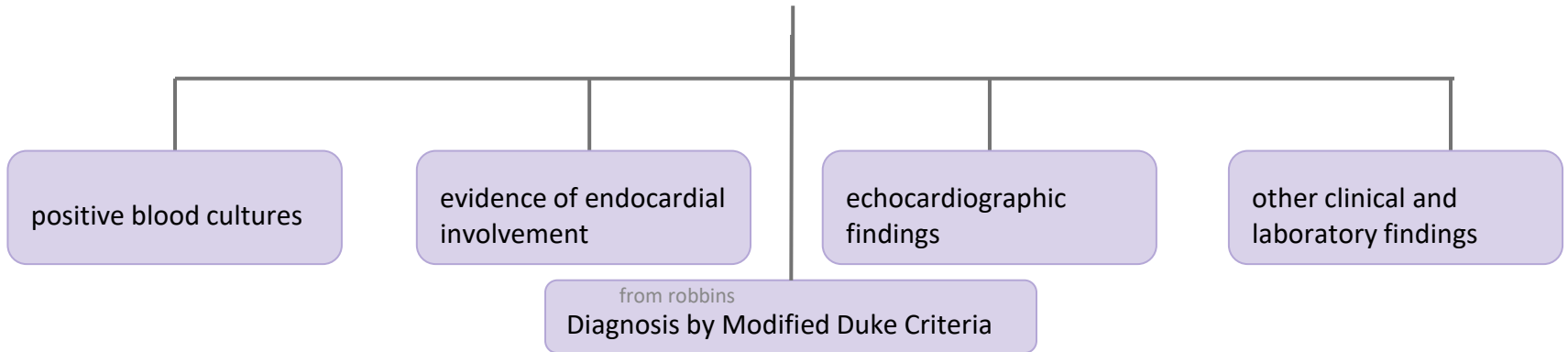
Contributory conditions are immunosuppression and neutropenia

Notes Only in F slides

- Mitral valves are the most common sites of IE followed by aortic valve.
- In IV drug users, the right side valves (tricuspids) are commonly involved.
- Vegetations can be single or multiple, involve one or more valve(s), differ in appearance according to the causative agent.

Infective Endocarditis

Diagnosis



Clinical features / رتبت المعلومات بناءً على المرجع

acute

- fever, chills, weakness, lassitude
- cardiac murmurs (with left sided IE)
- rigor, malaise
- Large vegetation, large emboli

subacute

- Insidious
- Splenomegaly
- Non specific fever, weight loss
- Smaller vegetations, so less embolic

Notes from robbins

- non specific clinical features are : fever, weight loss, flu like syndrome
- vegetations of subacute exhibit granulation tissue in their bases which means healing

Infective Endocarditis

Complications

- Due to large vegetation -> emboli will lead to :Infarction, Metastatic infection, affects distant organs like spleen, brain or heart, **Kidney: Ag-Ab complex -> nephrotic syndrome or Renal failure**
- Congestive heart failure due to valve disease
- It is usually destructive so it can lead to ring abscess and perforation of the aorta and myocardium
- Death up to 60% (high mortality)(acute)
- Low mortality(subacute)
- Ulceration and perforation of valves
- Rupture of chordae tendineae
- Arrhythmias
- Septicemia
- Septic systemic embolization of infected vegetations which travel to multiple sites, causing infarcts or abscesses in many organs (e.g. neurologic deficits due to embolization to the brain or infarcts of the myocardium due to embolization to the coronary artery),
- In IV drug addicts (tricuspid valve/ right sided endocarditis) □ pulmonary emboli.
- Mycotic/ infected aneurysms of vessels.

question might
come in case
scenario including
these risk factors

Cont. Risk factors male and female are mixed

In children: an underlying cardiac lesion (congenital heart disease is most common).

In adults: (note: more than half of adults with bacterial endocarditis have no predisposing cardiac lesion)

1 **Mitral valve prolapse and congenital heart disease** are the common risk factors for bacterial endocarditis in adults.

2 **People with prosthetic valves** are at high risk of developing IE . Prosthetic valve endocarditis is caused commonly by coagulase-negative staphylococci (e.g. *S. epidermidis*).

3 **Intravenous drug abusers** can inject micro-organisms intravenously when taking intravenous drugs, leading to IE. The tricuspid valve is most commonly infected. About 50% of the IE in IV drug abusers are caused by *S. aureus*.

4 Transient bacteremia from any procedure may lead to infective endocarditis e.g. dental procedures, urinary catheterization, **infected indwelling vascular catheters**, gastrointestinal endoscopy, and obstetric procedures.

5 Bicuspid aorta

6 Chronic Rheumatic heart disease

7 Degenerative Calcific stenosis

Other types of infective endocarditis

1. **Libman-Sacks endocarditis:** Less common, **non-infective**, verrucous endocarditis associated with elevated levels of circulating immune complexes. **Seen in patients with systemic lupus erythematosus(SLE)** characterized by:
 - Unknown etiology
 - Both sides of the valve
 - Deformity to the valve by healing of fibrinoid necrosis and mucoid degeneration
 - Sterile
1. **Endocarditis seen in carcinoid syndrome:** Secretory products of carcinoid syndrome, especially **5-hydroxytryptamine** can cause endocarditis. The endocardial plaques are seen in the right side of heart (**F slides only**)
2. **Nonbacterial thrombotic endocarditis (marantic endocarditis)** (most common in Qs)
 - a. Characterized by sterile vegetations on the leaflets of the cardiac valves. **There is no infective organism. It is aseptic.**
 - b. **Pathogenesis/ association:**
 - i. **Subtle endothelial abnormalities.**
 - ii. Hypercoagulability.
 - iii. Association with malignancy (50%) such as adenocarcinoma
 - iv. **Disseminated intravascular coagulation**
 - Aortic valve most common site.
 - May embolize to different parts of the body including brain, but the emboli are sterile.

ايه خصلنا (IE): VALVULAR HEART DISEASE

Two basic types: **F slides only**

1. Stenosis of valves: failure to open
2. Regurgitation of valves: Insufficiency or failure to close

Both cause murmurs

Causes

1-Congenital

2-Acquired

- post inflammatory scarring e.g. as a late complication of rheumatic fever (most common) or secondary to various other inflammatory processes.
- can occur even with prosthetic cardiac valves
- can be secondary to thrombus formation or infectious endocarditis.

Aortic stenosis

-Commonly caused by calcification and is called as calcific aortic stenosis. Calcific aortic stenosis affects:

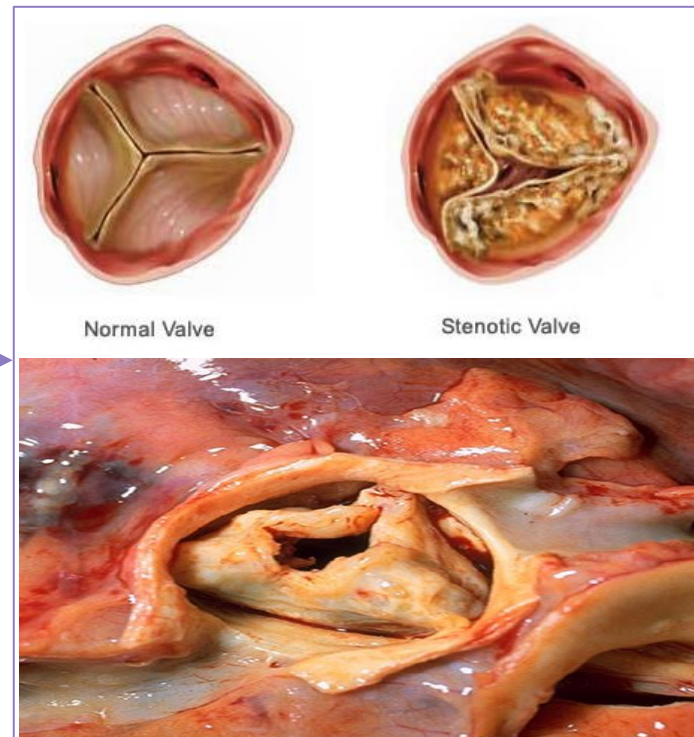
-Normal aortic valve as part of the aging degenerative process in > 60 yrs old.

-Congenital bicuspid aortic valve

-Valves scarred by rheumatic heart disease

Aortic regurgitation/ insufficiency caused by:

- Non-dissecting aortic aneurysm.
- Rheumatic heart disease
- Infective endocarditis
- Syphilitic (luetic) aortitis (rare)



F slides only

VALVULAR HEART DISEASE

Mitral stenosis

- ❖ Mitral stenosis is more common than mitral regurgitation.
- ❖ Most common cause □ rheumatic heart disease.
- ❖ In mitral stenosis (picture):
 - ❖ Leaflets are thickened, fibrotic and fused leading to fish mouth/button hole deformity (stenosed valve looks like fish's mouth or button hole)
 - Increased pressure, dilatation and hypertrophy of left atrium.
 - secondary deposition of Ca^{++}
 - Pulmonary hypertension and lungs are firm and heavy (chronic passive congestion).
 - Right heart may be affected later (right ventricular hypertrophy).



Mitral regurgitation

- Is usually due to rheumatic heart disease.
- can also be due to mitral valve prolapse, infective endocarditis, papillary muscle injury in myocardial infarction etc.
- Leads to left vent. hypertrophy and dilatation.

VALVULAR HEART DISEASE

Mitral valve prolapse

- It is the most frequent valvular lesion in developed countries
- More common in **younger** women
- There is myxoid/mucoid degeneration of the valve which causes ballooning of mitral valves (floppy cusp)
- It results in stretching of the mitral valve, producing a parachute deformity of the cusp with prolapse of the cusp into the atrium during systole. These changes produce a characteristic systolic murmur
- Pathogenesis unknown
- Can be associated with **Marfan syndrome**
- Most patients asymptomatic but can occasionally lead to mitral insufficiency and arrhythmias
- Patients are predisposed to infective endocarditis



Pericarditis all the next is M slide only

Uremia is the most common systemic disorder associated with pericarditis

Serous

Non bacterial causes:
Rheumatic fever
SLE
Tumor
Uremia
Primary viral

Fibrinous, serofibrinous

Same causes of serous
Most commonly due to MI

Purulent

Bacteria (staph, strept, pneumococcus)
Fungi
Parasite
Can lead to mediastinopericarditis or congestive pericarditis

Hemorrhagic

Surgery
TB
Tumor

Caseous

TB
Fungi
Fibrocalcific constrictive pericarditis

Healing

Resolution
Fibrosis:
Epicardial plaque
Thin
Thick massive adhesion

- Adhesive mediastinopericarditis:
 - Heart contracts against the surrounding structures
- Constrictive pericarditis:
 - 1 cm thick dense fibrous obliteration
 - Limit diastolic expansion and cardiac output

Pericarditis all the next is M slide only

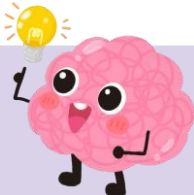
Chronic pericarditis/morphology

Male dr skipped it

- Ranges from delicate adhesions to dense fibrotic scars that obliterate the pericardial space.
- In extreme cases the heart can't expand during diastole : constrictive pericarditis

Clinical picture

- Atypical chest pain (worse on reclining)
- High pitch friction rub
- Significant exudate? cardiac tamponade? faint distant heart sounds, distended neck veins, declining cardiac output and shock
- Chronic constrictive pericarditis? venous distension and low cardiac output



KEYWORDS

Acute Rheumatic Fever	<ul style="list-style-type: none"> ● Group A- beta hemolytic Streptococcus <ul style="list-style-type: none"> ● Aschoff bodies ● Serum antistreptolysin O <ul style="list-style-type: none"> ● Elevated ESR
Chronic Rheumatic heart disease	mitral valve infection
Infective Endocarditis	Vegetation on heart valve / mitral valve most common site of IE
Acute IE	staph aureus- infects normal or damaged valve- IV drug abusers= Affect tricuspid valve
Subacute	Strep viridans, infect damaged valve
Libman sack endocarditis	SLE , non infectious
Marantic endocarditis	With malignancy such as adeno / sterile vegetation
aortic stenosis	Calcification of valve caused by IE or RHF
Mitral stenosis	Fish mouth or button hole appearance / caused by RHF
Mitral valve prolapse	in developed countries / Marfan syndrome

MCQ

C-5-D-4-B-3-A-2-D-1

1- Which one of these is a minor criteria of acute rheumatic fever?

[Slide 9](#)

A)Arthralgia

B)EKG changes

C)Subcutaneous nodules

D) A&B

2- which one of these valves is the most common infected in chronic rheumatic heart disease?

[Slide 10](#)

A) mitral valve (mitral stenosis)

B) tricuspid valve

C)pulmonary valve

D) all of above

3- Fish-mouth mitral valve stenosis is a complication of?

[Slide 12](#)

A) Mitral valve prolapse

B)Rheumatic heart disease

C)Acute endocarditis

D)Carcinoid heart disease

4- IV drug abuser affects which valve and which organism causes it (most common)

[Slide 15](#)

A)mitral, Staph aureus

B) Tricuspid,strep viridans

C) Aortic , Staph aureus

D) Tricuspid ,staph aureus

5-Mitral valve prolapse is associated with which one of these

[Slide 22](#)

A)Carcinoid syndrome

B)Hypercoagulability

C) Marfan syndrome

D) Kidney failure

Cases

6-C 7- C 8-A 9- B

6- A 10-year-old boy previously got infected with group A beta hemolytic streptococci and sore throat and fever , 6 weeks later he developed cardiac symptoms and skin nodules . Which of the following pathologic features correlate with his condition?

A) Acute inflammation	B) Collection of neutrophils and coagulative necrosis	C) Eosinophilic collagen, activated macrophages and chronic inflammation	D) none of the above
-----------------------	---	--	----------------------

7- A 23-year old women present with fever , and ECG changes and she has a new cardiac murmur , after doing some investigation it shows that she has a mitral stenosis which one of the following is usually combined with her condition ?

A)Tricuspid stenosis	B) Pulmonary stenosis	C)aortic stenosis	D) I don't know
----------------------	-----------------------	-------------------	-----------------

8-A person from cartel in mexico at midnight always has a habit of injecting himself with cocaine which of the following he will most commonly get in case of cardiovascular disease

A) acute IE	B) Subacute IE	C) Marantic endocarditis	D) NONE just euphoria
-------------	----------------	--------------------------	-----------------------

9- Non smoker female was diagnosed by a malignant neoplasm one of the complication she had is a sterile vegetation on the aortic valve which of the following is characterized by that

A) Libman-sacks endocarditis	B)Marantic endocarditis	C) Infective Endocarditis	D) Chronic RHF
------------------------------	-------------------------	---------------------------	----------------

Pathology team

Shahad Alshehri

Hessa Alamer

Muhannad Alotaibi

Joud alahmari



Abdulaziz Nasser



Norah albahily

Abdulaziz Alobathani

Shoug Albattah

Faris Alturaiki

Marwah Ahmed

Tariq Alshamrani

Ritaj alsubaie

Waleed Alanazi

Elaaf Albadi

Abdulaziz Alanazi