Lecture 1:
Rheumatic heart disease, infective endocarditis and valvular heart disease

- Important
- Explanation
- Addition note

OBJECTIVES

- At the end of this lecture, the students should be able to:

(1)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.

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

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

- The difference between :

Rheumatic Arthritis	Rheumatic fever
systemic disease mainly effects the joints and may effect lung	connective tissue disorder mainly effects the heart

1- Rheumatic fever

Definition : is an acute, immunologically mediated, multi-system inflammatory disease

- It is characterized by inflammatory reaction **involving heart, joints , central nervous system and skin.**
- ~3% of patients with untreated group A streptococcal pharyngitis.
- seen mainly in children, **5 to 15 years of age.**
- Trigger by sore throat infection or tonsillitis

Pathogenesis:

Because previous infection by **group A beta-hemolytic streptococcus** “pharyngitis”.

group A streptococcus produce **M Protein** ”antigen” that is resemble in its structure of protein material (glycoprotein) which is found in heart and joints. **M Protein** triggers immune system to make antibodies that effect bacteria and organs (**Antigen-antibody reaction**)

Morphology:

What is happening in the heart?!

area of focal interstitial **collagen necrosis** surrounded by large cells called (**Anitschkow cells = caterpillar cells**), this lesion called **“aschoff bodies”.** type of macrophages which have slender chromatin and by occasional multinucleated giant cells called **(Aschoff cells)**.

- Effected site

Rheumatic fever effects all three layers of the heart (endocardium, myocardium and pericardium) this called "**pancarditis**" :

1. **Myocarditis**: Can cause sudden death.

2. **Pericarditis**: fibrinous or serofibrinous deposits between visceral and parietal layers of the pericardium. These deposition called "**bread and butter**" because it resembles bread and butter

3. **Endocarditis**: including valves (valvulitis) and chordae tendineae. There is resultant fibrin deposition on valve leaflets forming minute, pale thrombi along lines of closure called rheumatic **vegetations**.

4. **Subendocardial lesions** appear as irregular thick patches commonly in the left atrium called as **MacCallum plaques.**

Clinical presentation

according to **Jones criteria**, which divide into **Major** and **Minor**

Major criteria

- Carditis
- Migratory Polyarthritis
- Sydenham's chorea
  Or Civitus dance
- Subcutaneous nodule
- Erythema marginatum

Minor criteria

- Unexplained fever
- Arthralgia (Joint pain without swelling)
- Raise ESR
- Previous history

**To prove the diagnoses:

1- throat swab, GAS will appear,

2- anti-streptolysin O (ASO)
it will be raised

- **A patient with R.F should have at least one Major, and two Minor**
- The patient come to hospital after 1-5 weeks after infection with jones criteria

CHRONIC RHEUMATIC HEART DISEASE

- In contrast, 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.
- The valve leaflets become permanently thickening, shrinking and dystrophic calcification this gives **"fish mouth appearance = buttonhole stenosis"** this leads to cardiac failure, thromboembolism and infective endocarditis

- Results:
- fibrosis of **valve** leaflets --> **stenosis** (Reduction of diameter)
- fibrosis of **chordae tendineae**--> **regurgitation** (improper closure)
- Ventricular hypertrophy
- Dystrophic calcification .

Histology:

- **No Aschoff bodies**
- **Diffuse fibrosis and neovascularisation**

The most effected site in the heart is valve

(1) The **mitral** valve is **most** involved in RHD

- Mitral stenosis is marked by diastolic pressure higher in the left atrium than in the left ventricle.

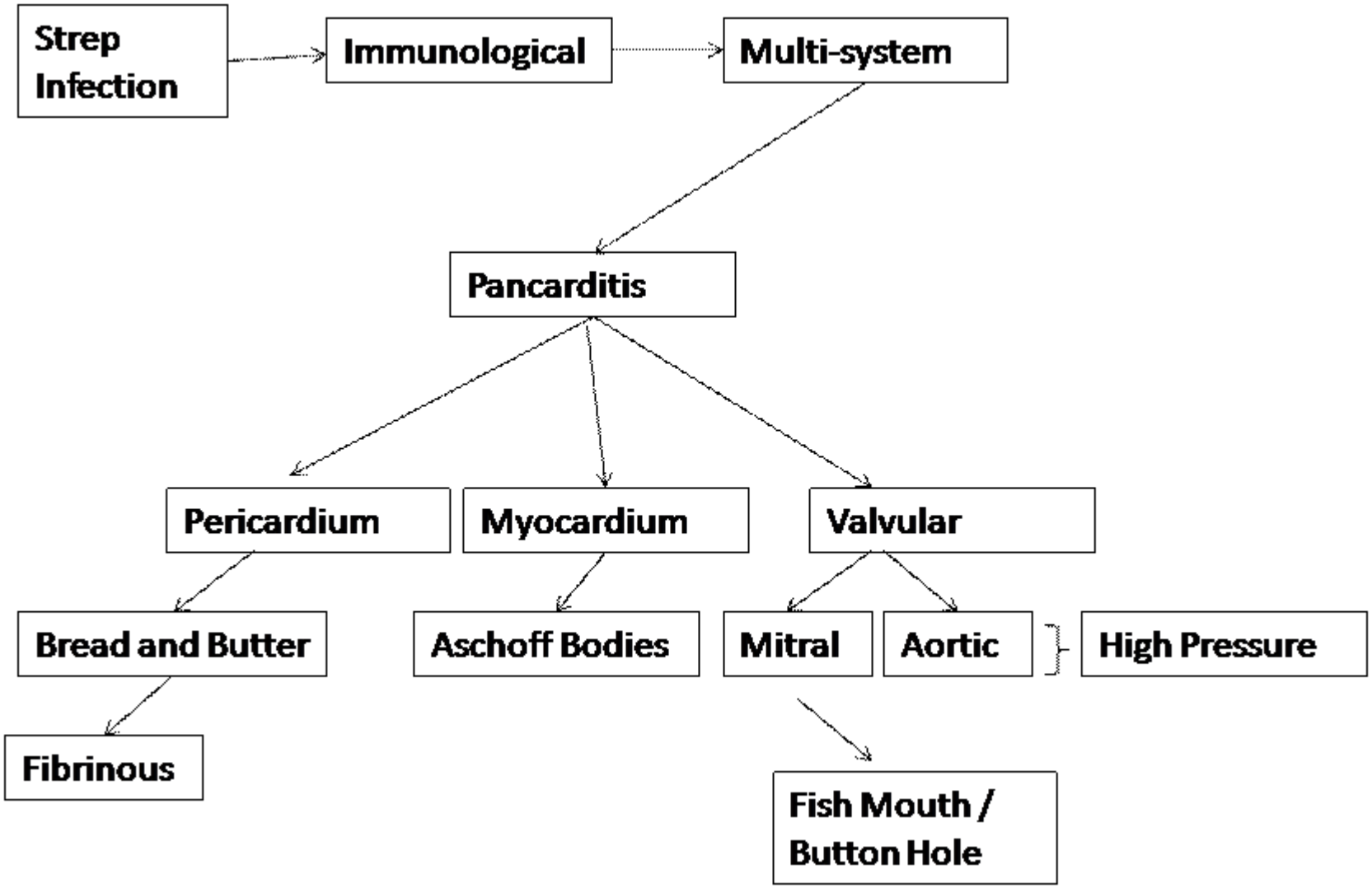
(2) The **aortic** valve is affected most often along with the mitral valve. It can be affected by stenosis or insufficiency.

(3) The tricuspid valve is rarely affected

(4) The pulmonary valve is never involved.

- Manifests: years or decades after the initial episode of rheumatic fever.
- Signs and symptoms+ complications : depend on the valve(s) involved: cardiac murmurs, hypertrophy, dilation, congestive heart failure, arrhythmia, thromboembolism and infective endocarditis.
- Treatment : may require valve surgery.

Summary :


Strep Infection
Immunological
Multi-system
Pancarditis
Pericardium
Myocardium
Valvular
Bread and Butter
Aschoff Bodies
Mitral
Aortic
High Pressure
Fibrinous
Fish Mouth / Button Hole

2- Endocarditis

Inflammation of the endocardium is marked by prominent involvement of the valvular surfaces. Usually bacterial or rarely fungal

General considerations

- Characteristics include large, soft, friable, easily detached vegetations, consisting of fibrin and intermeshed inflammatory cells and bacteria.

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

Endocarditis			
	Acute endocarditis	caused by Staphylococcus aureus	is often secondary to infection occurring elsewhere in the body. Like:Bronchiectasis, brain abscess
	Sub-acute endocarditis	caused by Streptococcus viridians	occur in patients with - Congenital heart disease - Valvular heart disease - Rheumatic origin - Drug abuser - Artificial valves

Clinical features

Valvular involvement

mitral valve is most frequently involved.

Tricuspid valve is involved in most cases of intravenous drug abuse.

Aortic valve is involved in most of cases with mitral valve.

Complications

Stenosis.

myocardial, brain, or lung abscess

Microembolization can give raise to petechia, nail bed homorrhage (splinter), retinal hemorrhage (roth spots), painless palm or sole eryrthrematous lsion (janeway lesion), painful fingertip nodules (osler nodes) and septic infarcts in the brain or in other organs

The renal glomeruli may be site of focal glomerulonephritis (focal necrotizing glomerulitis) caused by immune complex disease or by septic emboli.

Which type III hypersensitivity

** We protect the patient from endocarditis by long-term penicillin especially in surgery procedure

** investigation: blood culture

** Vegetations may be single or multiple

3- Valvular Heart Disease

- **Causes**
- occurs as a result of rheumatic fever
- secondary to various other inflammatory processes.
- congenital.
- prosthetic cardiac valves
- secondary to thrombus formation or infectious endocarditis

Mitral valve

Is the most frequent valvular lesion in developed countries

Ballooning of mitral valves (floppy cusp), parachute deformity with prolapse of the cusp into the atrium with systolic murmur.

Component of Marfan syndrome

Aortic valve

Caused by calcification and iscaused by calcification and is called Calcificaortic stenosis

In older people

May congenital

Valve affected and scarred by rheumatic heart disease

Regurgitation or insufficient can be caused by sypilitic

TRICUSPID VALVE

In rheumatic heart disease it is rarely involved together with the mitral and aortic valves

PULMONARY VALVE

Commonly affected by congenital malformations such as in the tetralogy of Fallot.

-Endocarditis of the carcinoid syndrome

- Secretory products of carcinoid syndrome can cause endocarditis
- The valves on the left side of the heart are rarely involved, because serotonin and other carcinoid secretory products are detoxified in the lung.
- thickened endocardial plaques characteristically involving the mural endocardium or the valvular cusps of the right side of the heart.

- Non-bacterial thrombotic endocarditis (marantic endocarditis) (aseptic)

- Characterized by the deposition of small masses of fibrin, platelets on the leaflets of the cardiac valves (sterile). There is no infective organism.

- Pathogenesis/ association:
  - Chronic diseases for long time.
  - Malignancy

  Aortic valve most common site.
  the emboli are sterile

-LIBMAN-SACKS ENDOCARDITIS

- occur in patients with systemic lupus erythematosus

MCQs

1- One of the Major jones criteria:

A. Unexplained fever
B. Arthralgia
C. Erythema marginatum
D. Raise ESR.

2- R.F usually follows an infection with:

A. Streptococcus group A
B. Staphylococcus aureus
C. Streptococcus viridans
D. H.influenza

3- To diagnose a patient with R.F, The patient should have at least:

A. One major, two minor
B. Two major , one minor
C. One major , one minor
D. Two major , two minor

4- One of the tests that is done to confirm diagnosis of R.F:

a) Alpha 1 antitrypsin
b) Rheumatoid factor
c) Anti-streptolysin O
d) Creatine kinase

Answers:
1-C
2-A
3-A
4-C

MCQs

5- Which valves are commonly affected by R.F:

a) Tricuspid , mitral
b) Aortic , mitral
c) Tricuspid , pulmonary
d) Aortic , pulmonary

6- Acute endocarditis usually follows an infection with:

a) Streptococcus group A
b) Staphylococcus aureus
c) Streptococcus viridans
d) H.influenza

7- Sub Acute endocarditis usually follows an infection with:

a) Streptococcus group A
b) Staphylococcus aureus
c) Streptococcus viridans
d) H.influenza

8- A patient withR.F comes with dancing movement, how can you explain this movement:

a) Myopathy
b) Neuromuscular junction disease
c) Sydenham's chorea
d) Psychosis

Answers:
5-B
6-B
7-C
8-C

MCQs

9- One of the complications of Endocarditis:

a) lung abscess
b) Asthma
c) Chronic bronchitis
d) Cancer

Answers:
9-A
10-A
11-B

10- An abnormal narrowing in a valve is called:

a) Stenosis
b) Incompetence
c) Regurgitation
d) Constriction

11- If someone came with previous history of RF , according to jones criteria

A. major
B. minor
C. None of them

MCQs

12- Jones criteria appears after infection ?

A. 1-5 day
B. 1-5 weeks
C. 1-5 month
D. 1-5 year

Answers:
12- B
13-B
14-C

13- What is the main pathological lesion occurs in the heart in acute R.F ??

A. Osler nodes
B. Aschoff body
C. Buttonhole stenosis
D. Ring abscess

14- Libman–Sacks endocarditis associated with ??

A. Carcinoid SYNDROME
B. Drug abusers
C. SLE
D. Elderly

Questions

1- What is Rheumatic fever?

- It’s an Immune mediated disease, common in people from 5 – 15 years old.

2- What Rheumatic fever is usually triggered by?

- By an infection that occurred by Group A beta hemolytic streptococci.

3- What is the protein that’s produced by the Group A hemolytic streptococci?

- Protein M.

4- Mention the minor John’s criteria.

- Unexplained fever
- Arthralgia (Joint pain without swelling)
- Raise ESR
- Previous history

Questions

5- Mention three of the microscopic features of Rheumatoid fever.

- Anitschkow cells.
- Necrotic connective tissue.
- Proliferating blood vessels.

6- In R.F Subendocardial lesions appear as irregular thick patches commonly in the left atrium

" Maccallum patch "

7- Mention the usual causes of Endocarditis.

- Rheumatic fever.
- Artificial valves.
- Congenital heart diseases.
- Drug abuser.

Questions

8- Mention the complications of Sub-acute Endocarditis.

- Brain infarcts and abscess.
- Petechia (Area of hemorrhage).
- Lung abscess.
- Osler nodes (Painful areas on the tips of the fingers).

9- In the Endocarditis of the carcinoid syndrome, Why does it effect the only valvular cusps of the right side of the heart not left?

- The valves on the left side of the heart are rarely involved Because Serotonin (5-Hydroxytrptamine) and other products of carcinoid syndrome detoxified in the lung before the blood going to left side of heart

9- what is the most common site for infective endocarditis ??

mitral followed by aortic vs except iv drugs users right sided like tricuspid valve is the most common

10 - where can we see fish mouth (Buttonhole) deformity ?

in mitral valve stenosis (leaflet is thickened and fibrotic)

Questions

11- what is maranitic endocarditis ?

its non bacterial thrombus endocarditis , usually effect sick people (malignancy 50% , chronic disease), the vegetation and the embolus is sterile

12 - mitral valve prolapse (floppy mitral valve) can be component of ?

marfan syndrome

13- the cause of aortic valve stenosis is

Dystrophic calcification

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