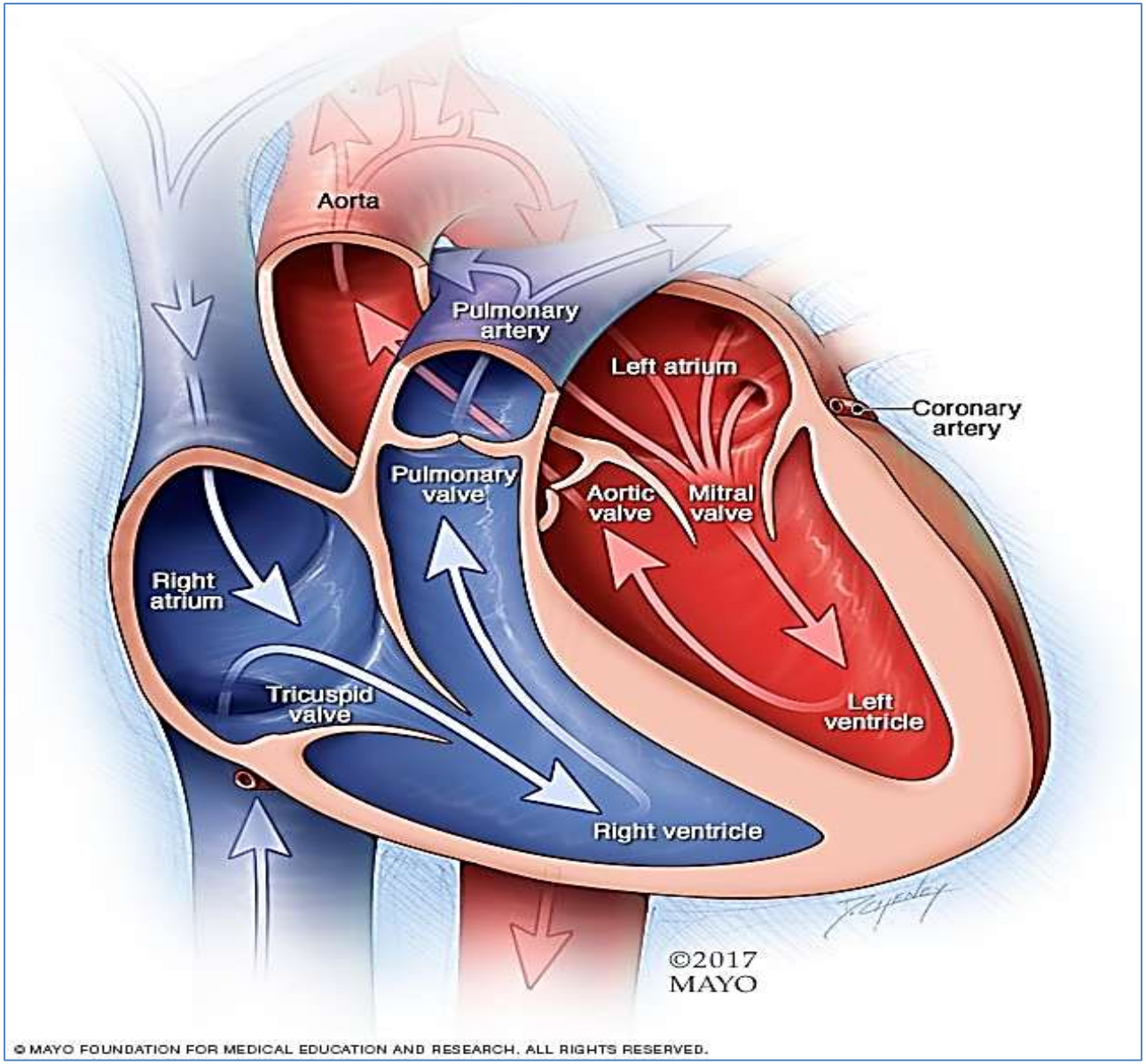
Valvular Heart Disease

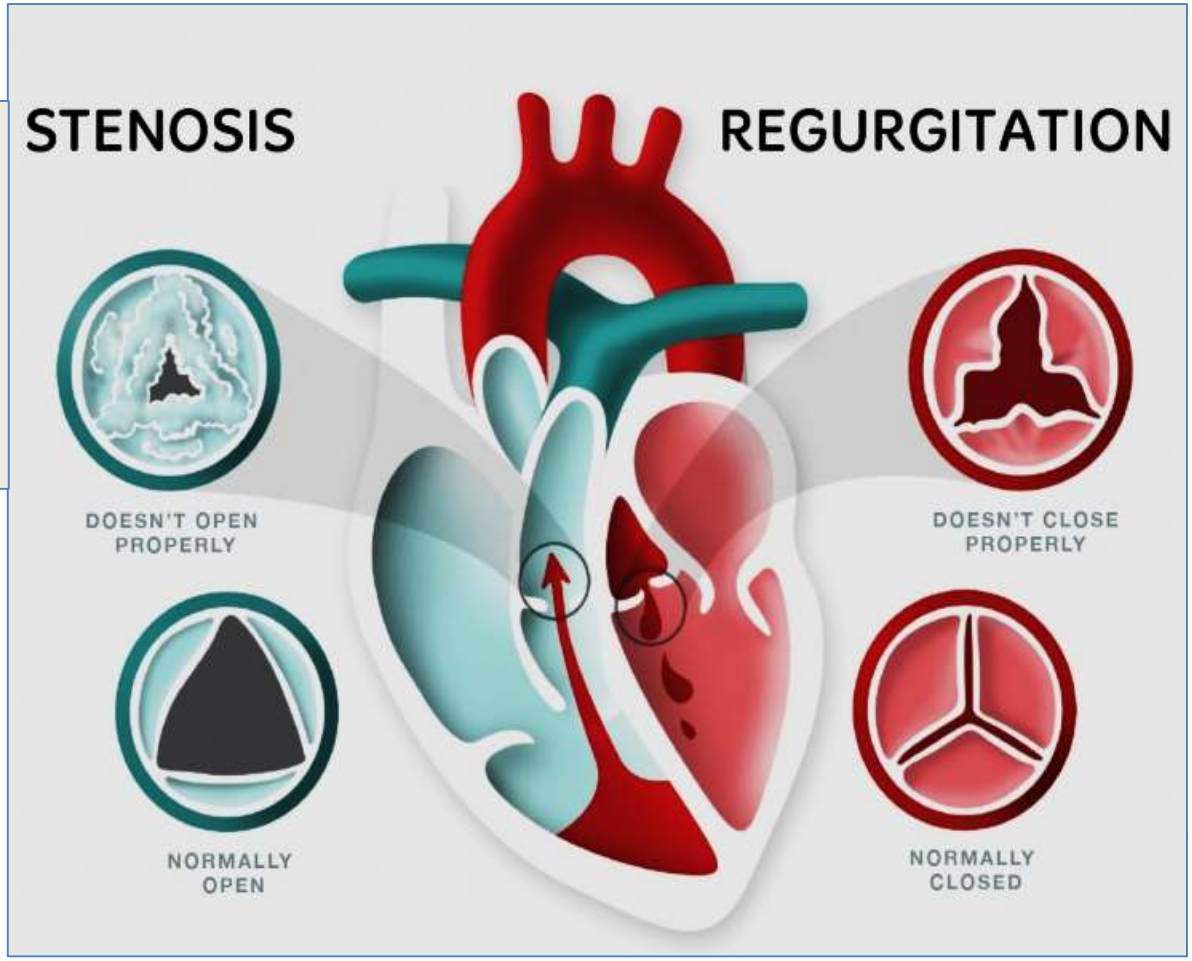
Dr. Nisreen Abu Shahin
Professor of Pathology
Pathology Department
University of Jordan

Normal Heart Valves



VALVULAR HEART DISEASE

Due to a chronic process (e.g. calcification or scarring)

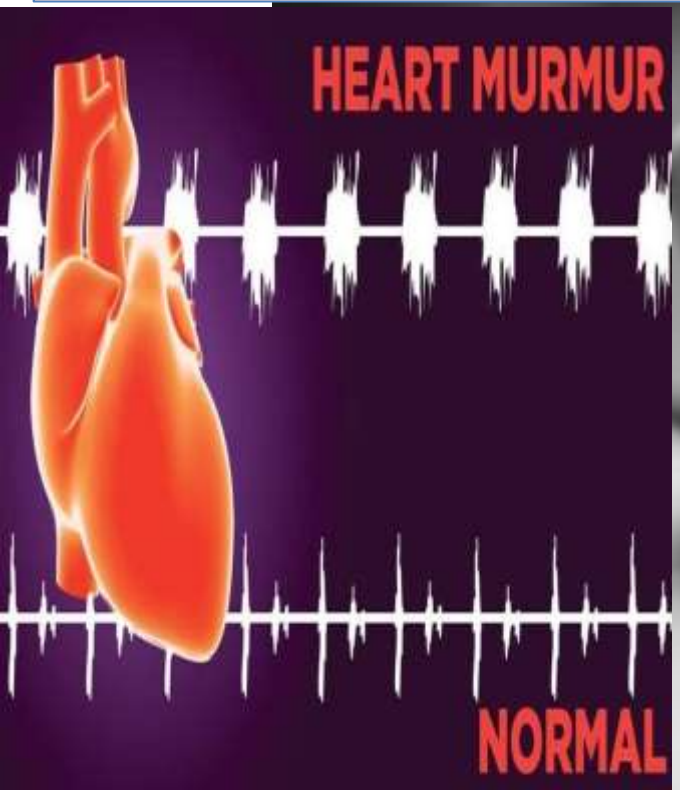


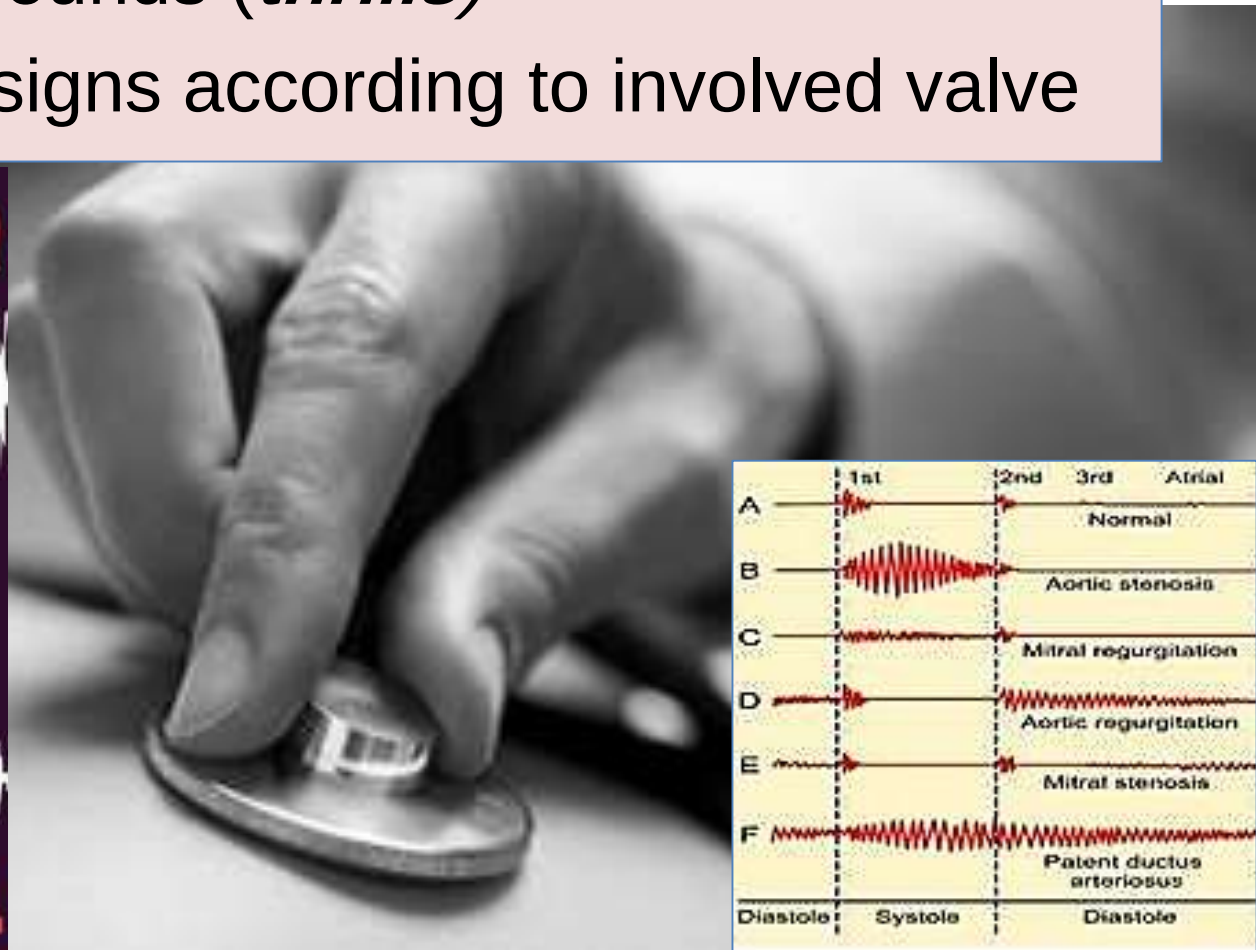
Abnormality:
1-valve cusps
2- supporting structures (e.g. mitral annulus, tendinous cords, papillary muscles)

It can be either acute (e.g. chordal rupture)
or chronic (e.g. scarring)

Clinical Signs of Valve Disease

- Abnormal heart sounds (***murmurs)***
- Palpated heart sounds (***thrills)***
- Specific clinical signs according to involved valve

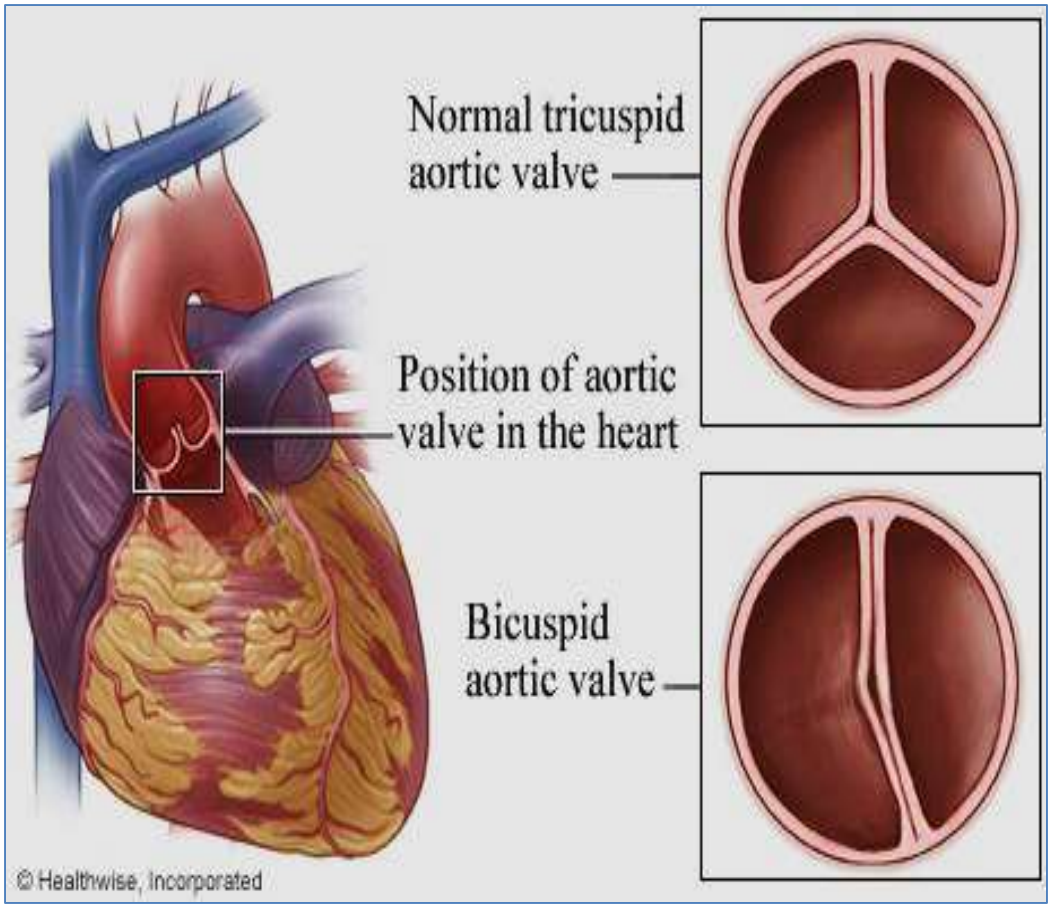




- Valvular abnormalities can be congenital or acquired

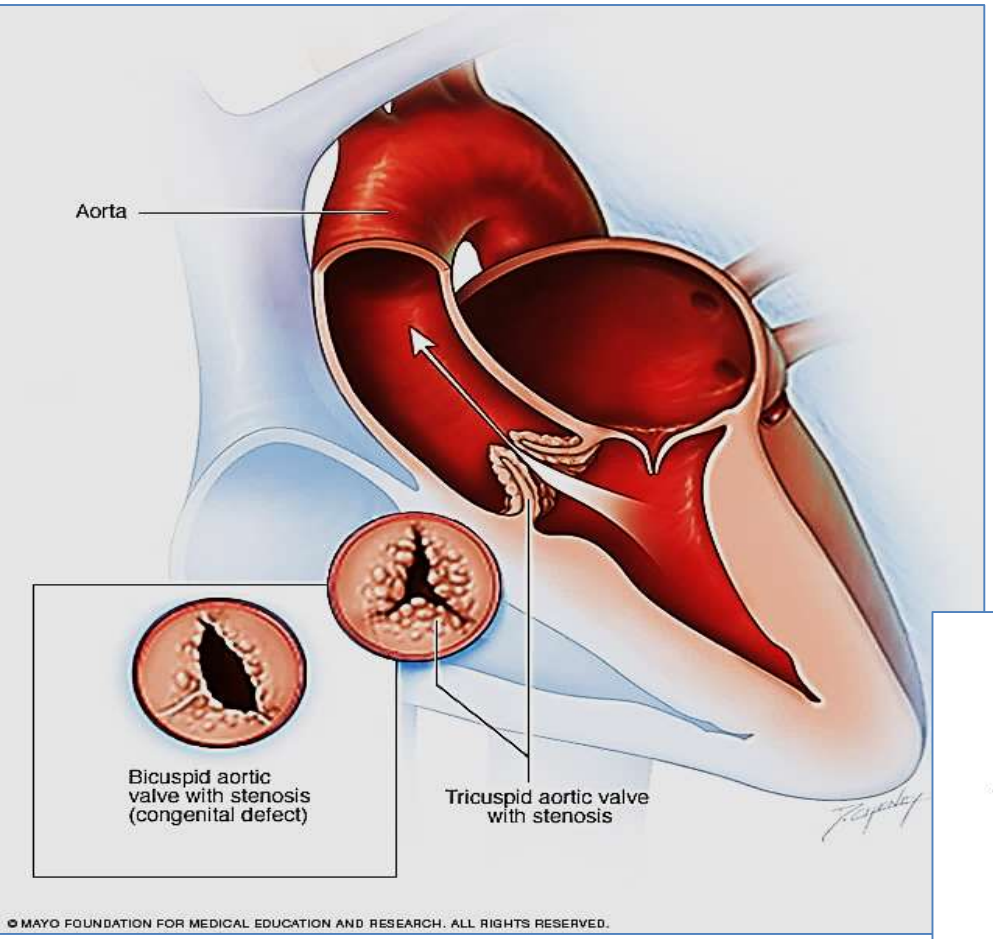
- The most common **congenital** valve lesion is ***bicuspid aortic valve***
- Most important cause of **acquired** valve disease is ***rheumatic fever***

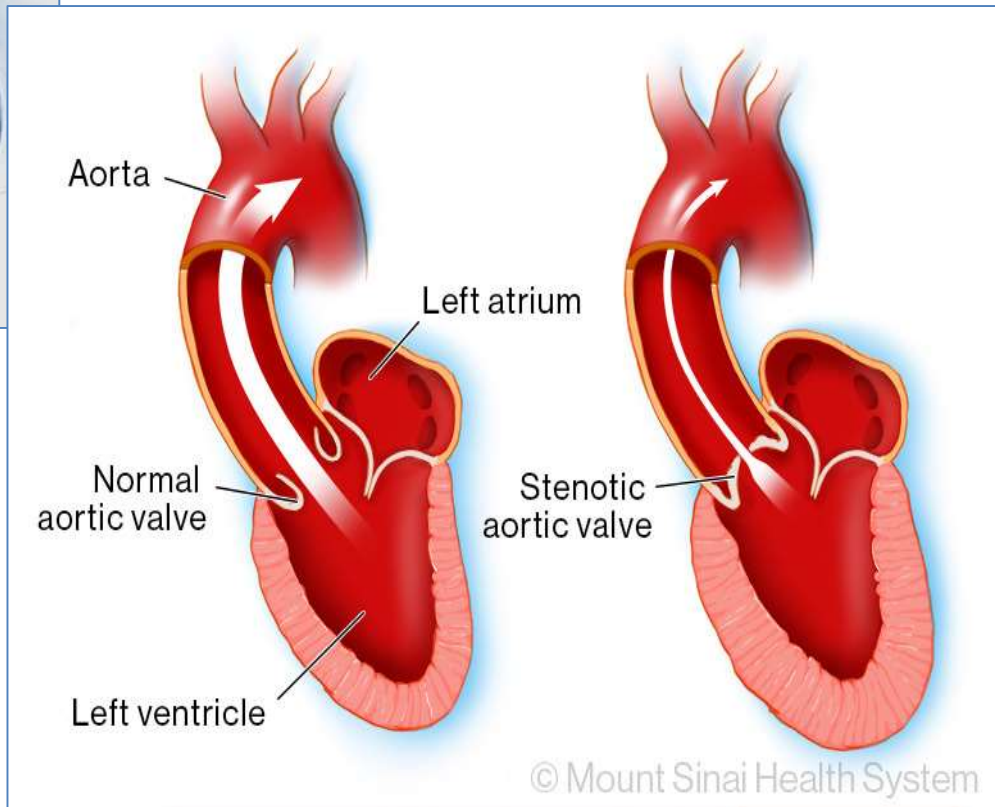
Bicuspid Aortic Valve



-only 2 functional cusps instead of 3
-1% - 2% of live births
-Isolated or associated with genetic mutations
-early life →Asymptomatic
-Later → early & progressive degenerative calcification of aortic valve

Aortic Valve Stenosis





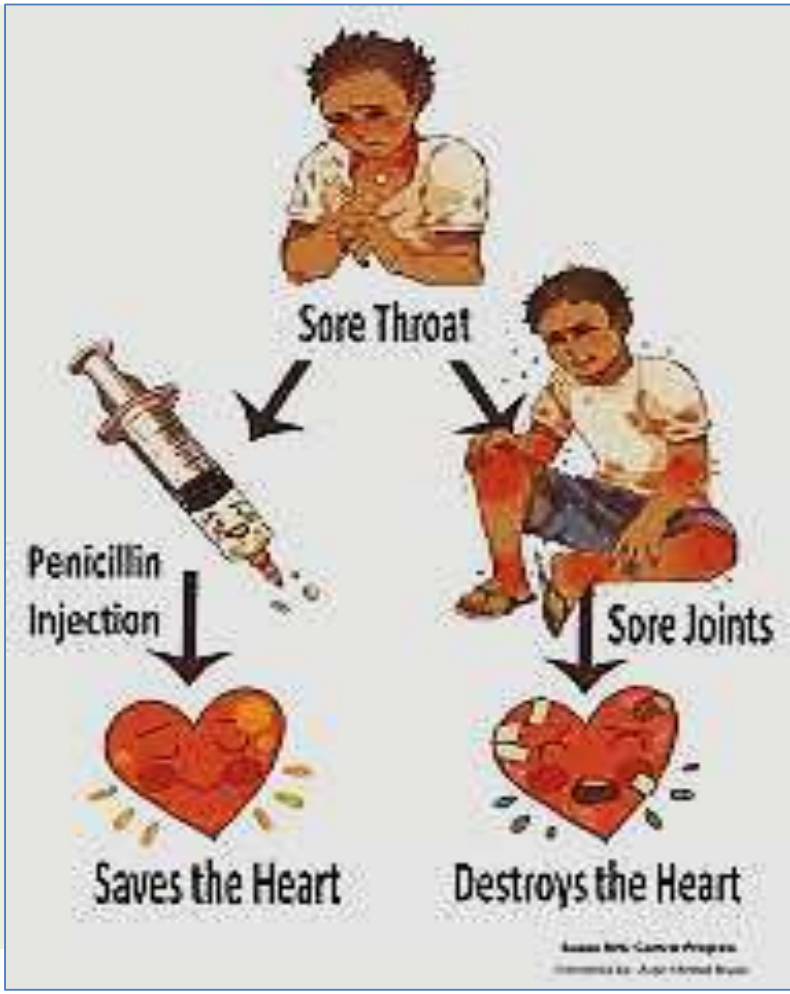
- **Acquired Valve Diseases**

- Mitral valve most common target of acquired valve diseases

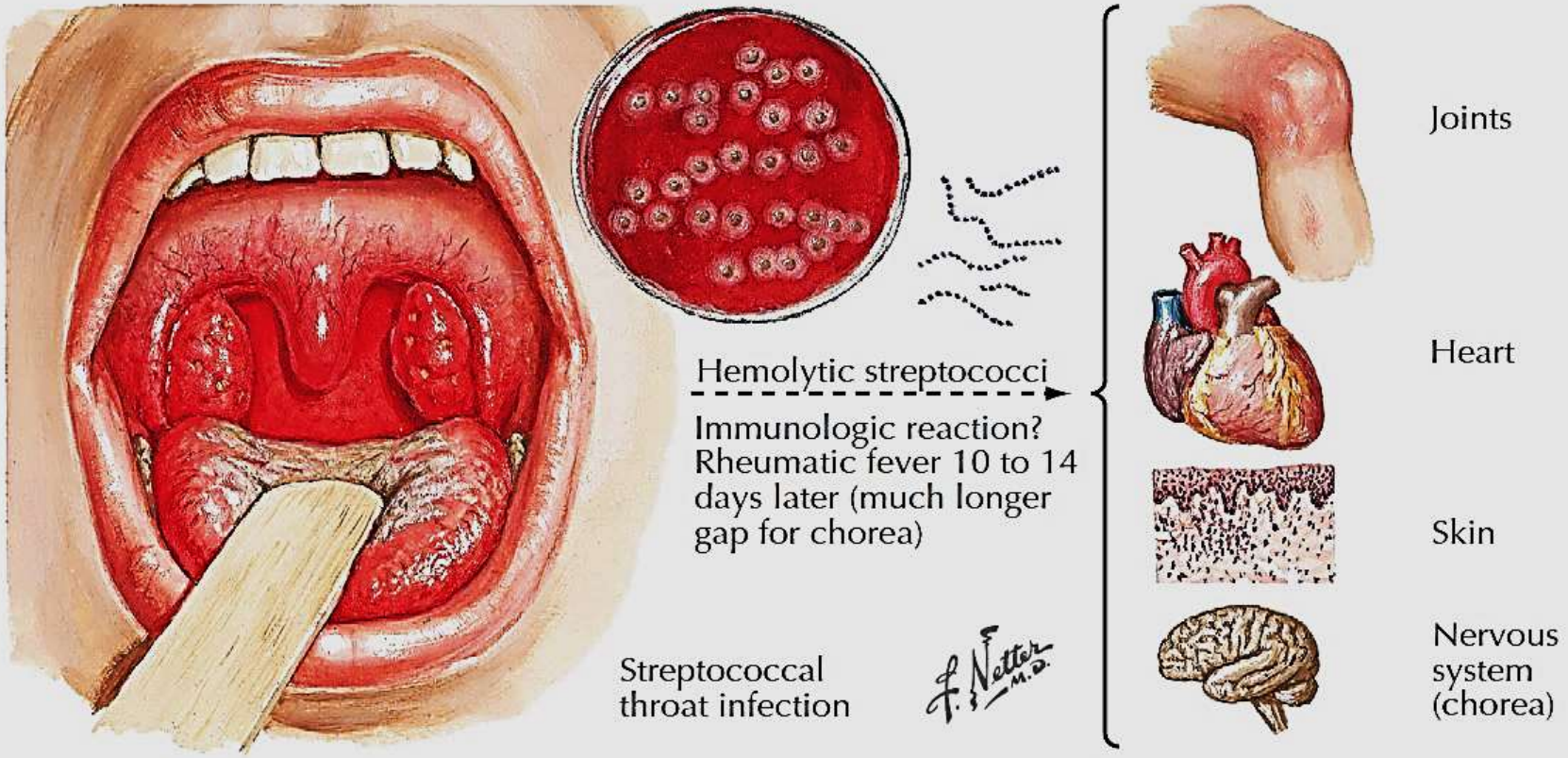
- Most common cause of **acquired** valvular diseases is **post-inflammatory scarring** due to rheumatic fever (2/3)

Rheumatic fever (Rheumatic Valve Disease)

- Immune- mediated inflammatory disease that follows infection by **group A streptococci**
- Incidence↓ in Western world (improved socioeconomics, rapid diagnosis, and Rx of strept. pharyngitis
- Still, important public health problem in developing countries



Rheumatic Fever



PATHOGENESIS:

hypersensitivity reaction due to antibodies against group A streptococcal antigens

These antibodies are cross-reactive with host antigens (heart; brain; joints; skin)

Rheumatic Fever

- Manifestations seen a few weeks after pharyngitis or skin infection
- Major organs involved: heart; joints; skin; and brain
- 2 phases:
- Acute: fever; arthritis; CNS symptoms; carditis
- Chronic: cardiac valve disease
- ……
- **Acute phase:**
- 80% children
- fever; migratory polyarthritis; **carditis**
- Carditis (arrhythmias; myocarditis; cardiac dilation; functional mitral insufficiency and CHF).
- ↑ serum titers of antibody against streptococcal antigens (streptolysin O; DNA-ase)
- **culture for streptococci is usually (-) at time of symptom onset**

Acute Rheumatic Fever- JONES criteria

Signs & Symptoms

Joints (arthritis)

❤ Carditis

Nodules (subcutaneous)

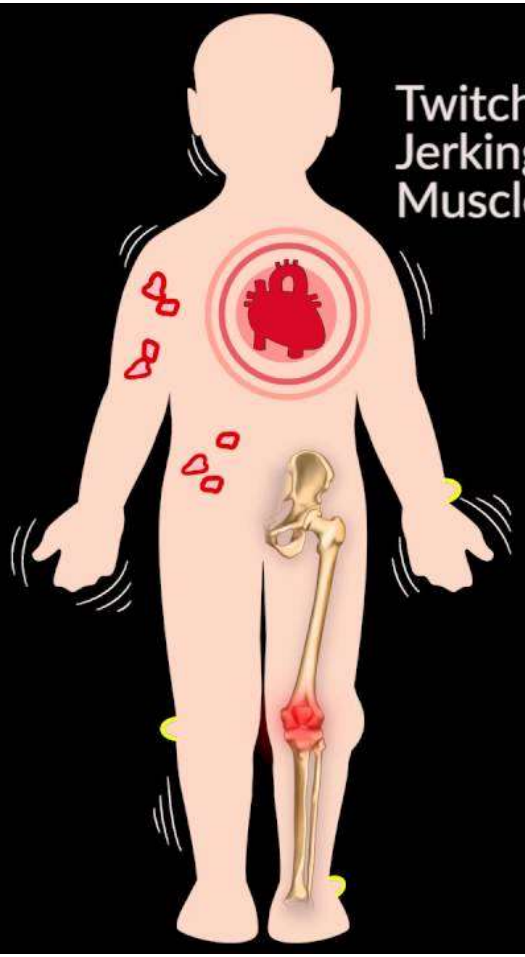
Erythema marginatum

Sydenham's chorea

-can present 3-4 months after GAS infection

-mean duration: 12-15 weeks

-episodes may last 6-12 months



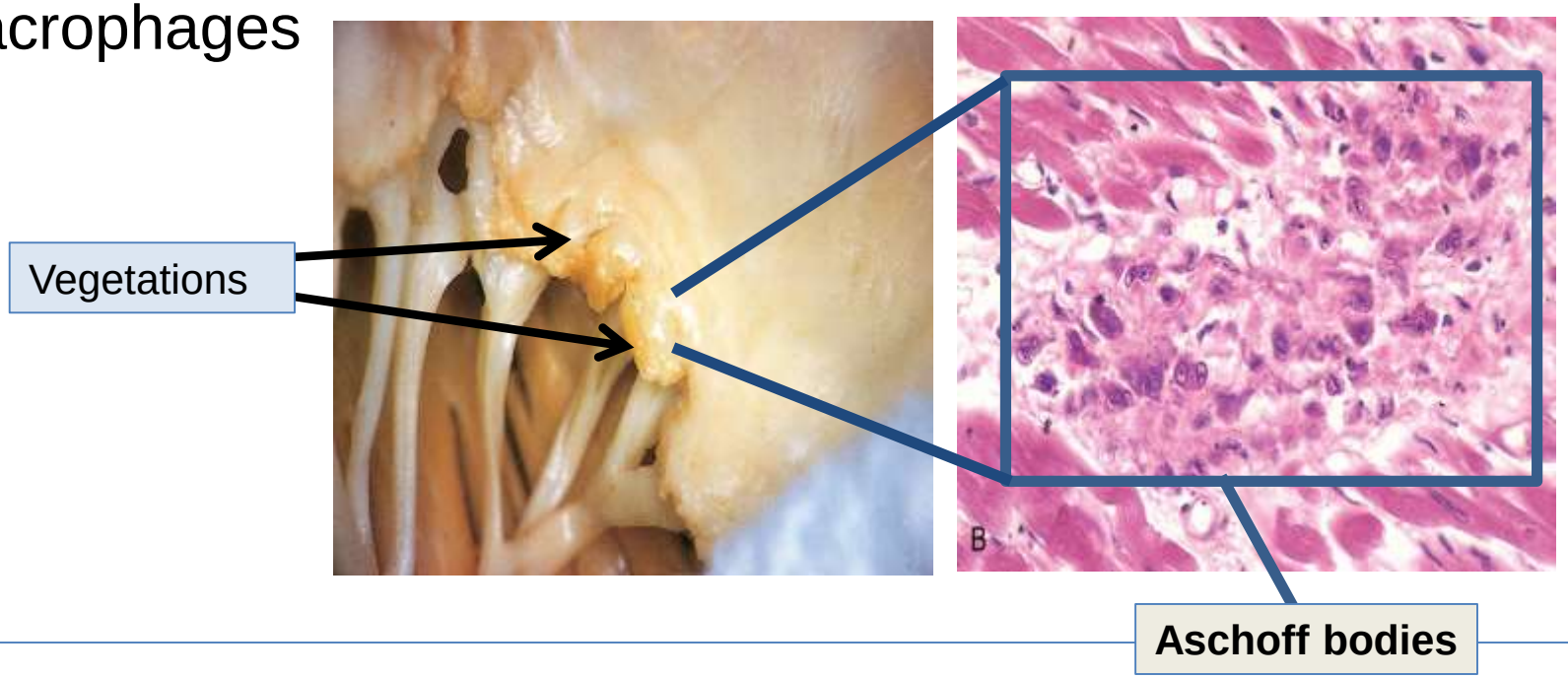
Carditis
Morphology- Acute Phase

- Valve vegetations
- **Aschoff bodies :**

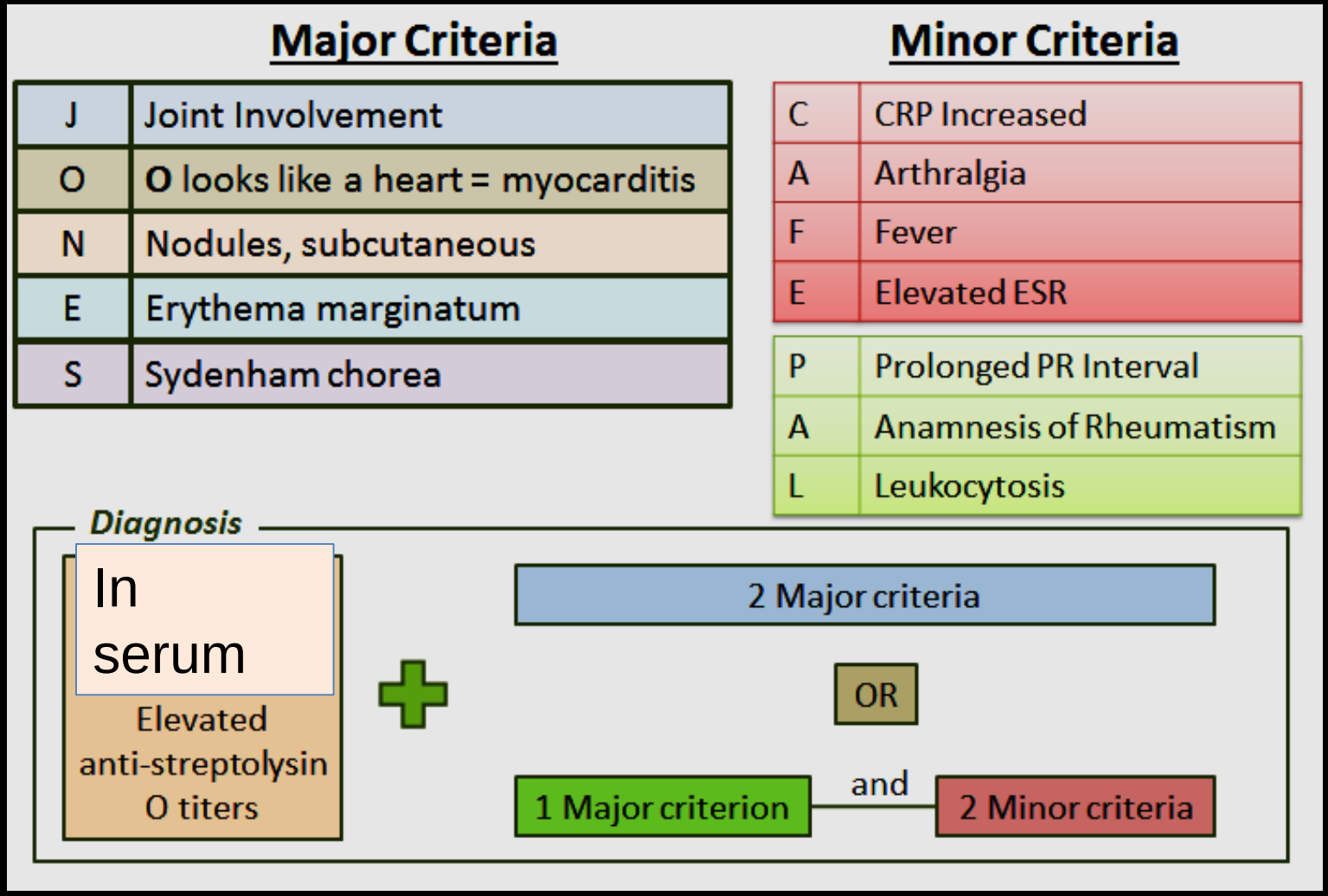
- Are inflammatory lesions in affected tissues

- *pathognomonic (diagnostic)* for RF

- collections of T lymphocytes+ plasma cells+ activated macrophages

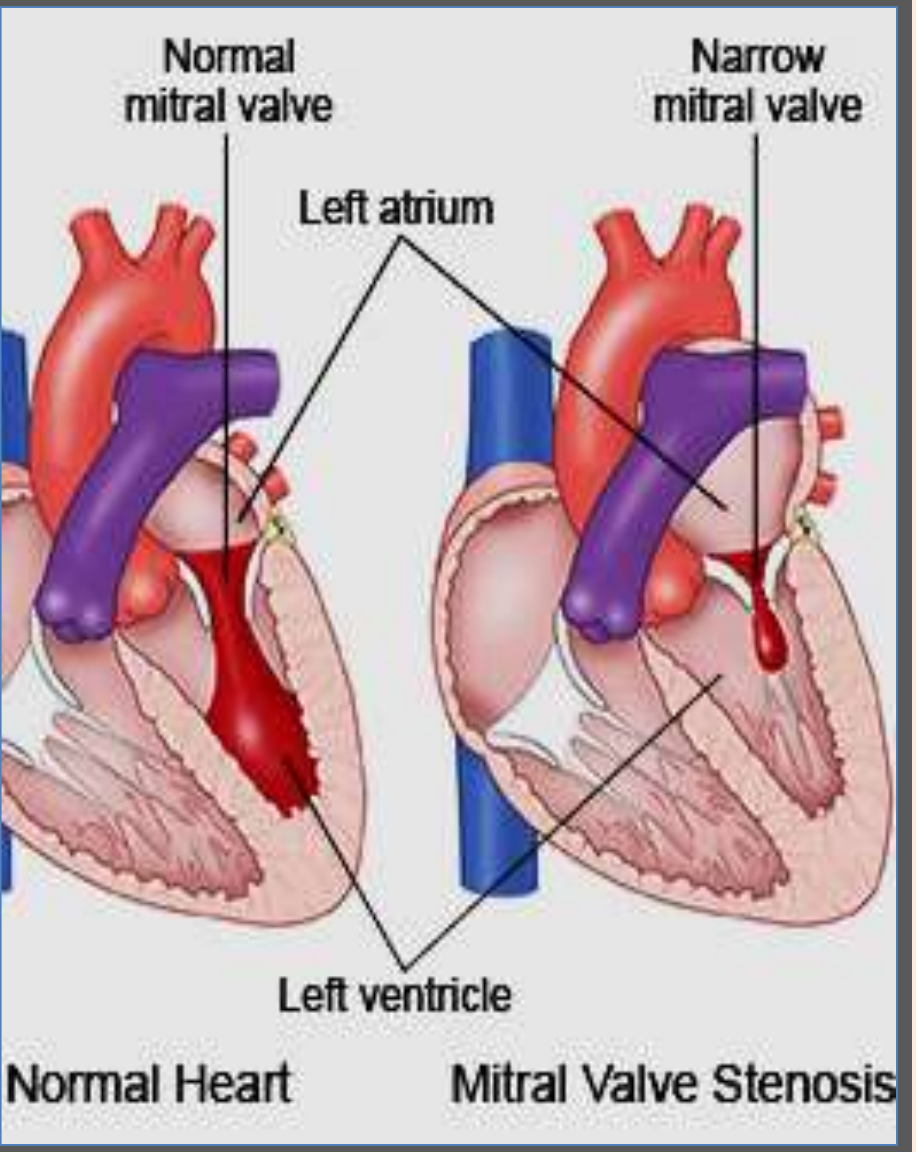


Diagnosis of Acute Rheumatic Fever



* Anamnesis: a preliminary case history of a medical patient

Chronic Rheumatic Carditis- Clinical Picture



Onset: years/ decades after initial acute episode

Chronic inflammation→ scarring → stenosis

murmurs - CHF - arrhythmias- mural thrombi

Prognosis: variable.

Management: Surgical repair or replacement of diseased valve

Chronic Phase - Morphology

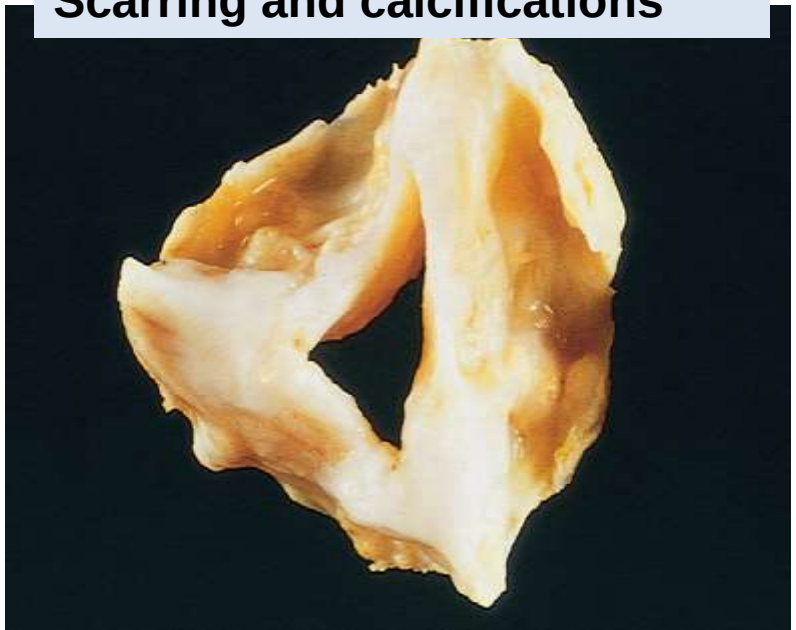
- Inflammation is followed by **scarring**
- Aschoff bodies **rarely** seen now

- **Valve stenosis** (most imp. functional consequence)

mitral valve (m/c)
aortic disease
tricuspid valve
pulmonary valve (rare)

↓

Scarring and calcifications

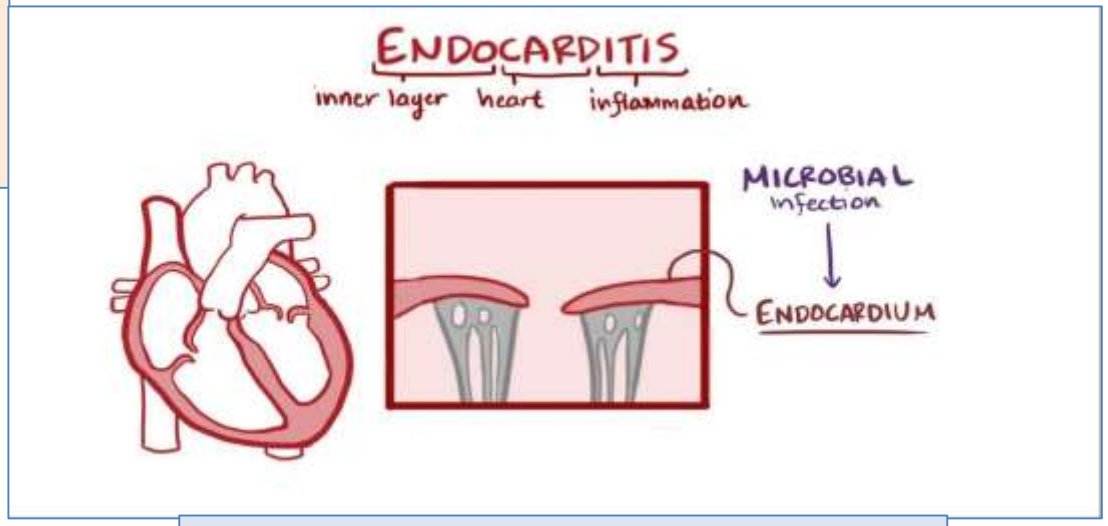


Infective Endocarditis

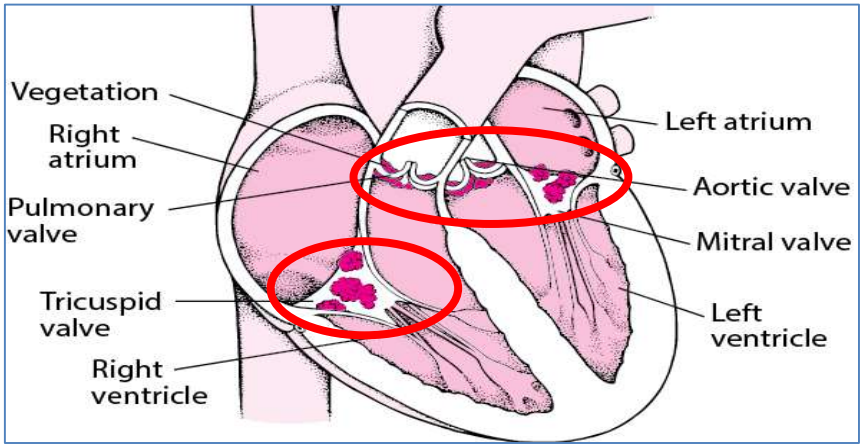
Infective Endocarditis (IE)

- Microbial (mostly bacterial*) invasion of heart valves and endocardium
- bulky, friable ***vegetations*** (necrotic debris+ thrombus+ organisms).

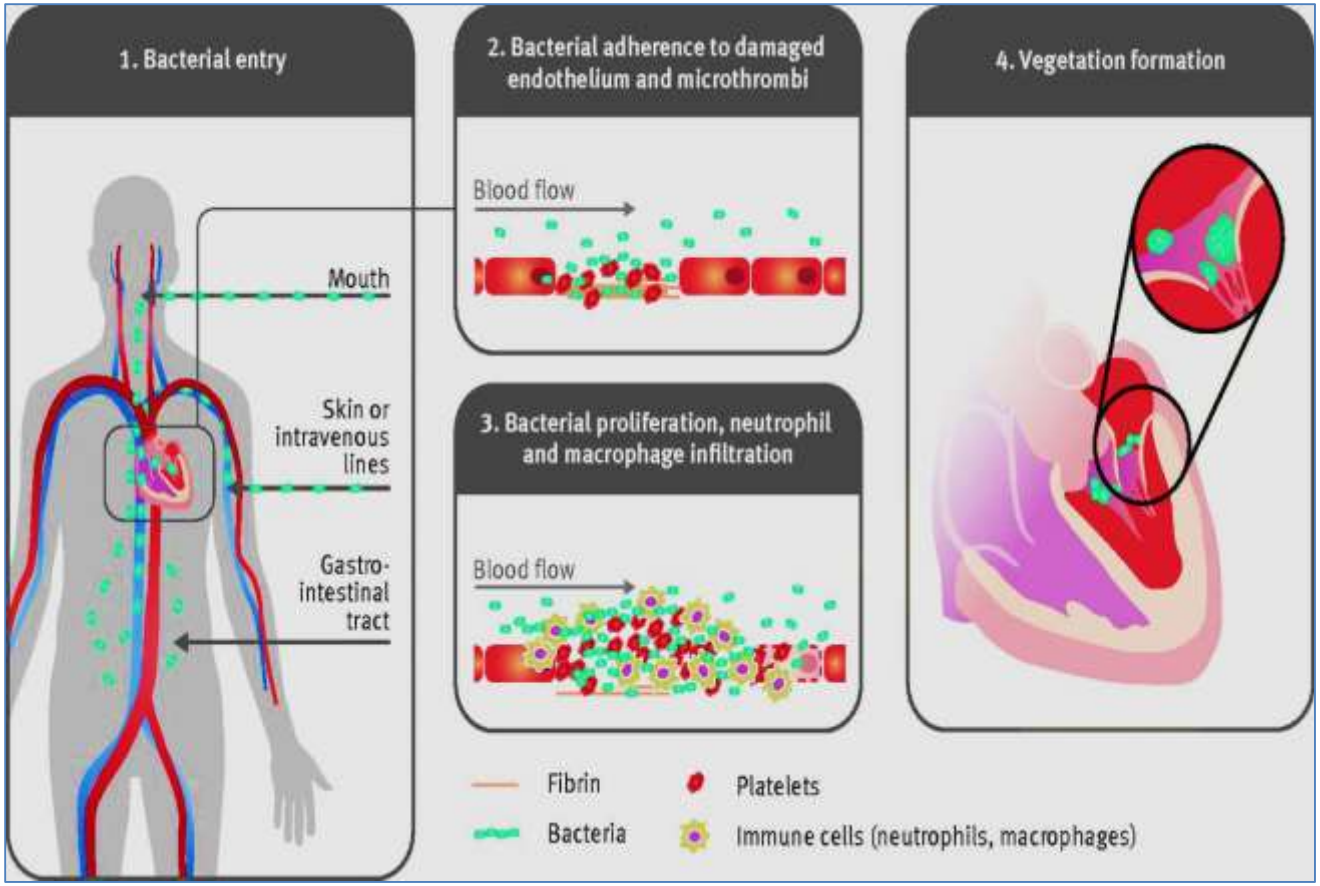
* others include: fungi, rickettsiae; and chlamydia



Infection of heart valves and endocardium



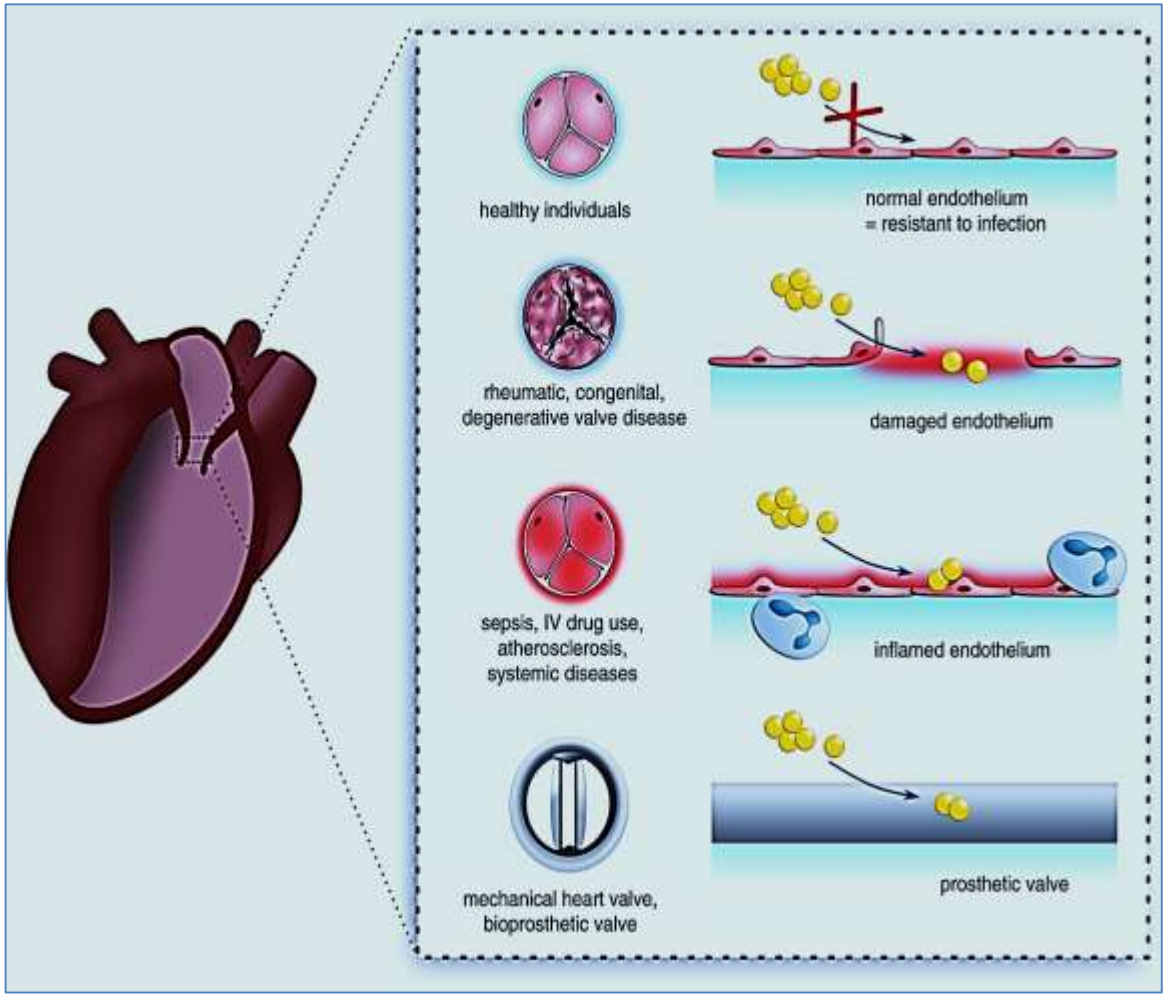
Infective Endocarditis
(Infection of heart valves and endocardium)



Infective Endocarditis- Risk Factors

- Congenital heart disease
- Acquired heart disease (including rheumatic fever)
- Indwelling vascular catheters
- Intra-cardiac devices & prostheses
- Immunodeficiency
- I.V. drug use/ abuse
- Septicemia

- ? Dental procedures (in patients with risk factors)

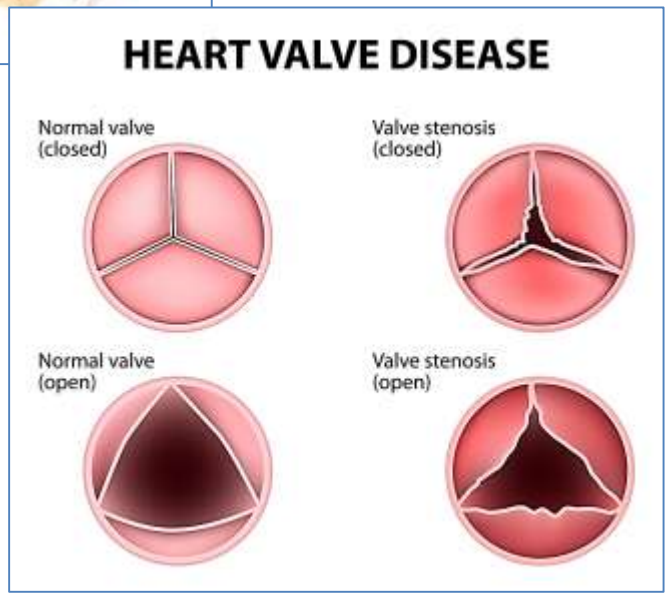


Infective Endocarditis (IE)

Classified into ***acute*** and ***subacute*** based on:

1- the virulence of microorganism

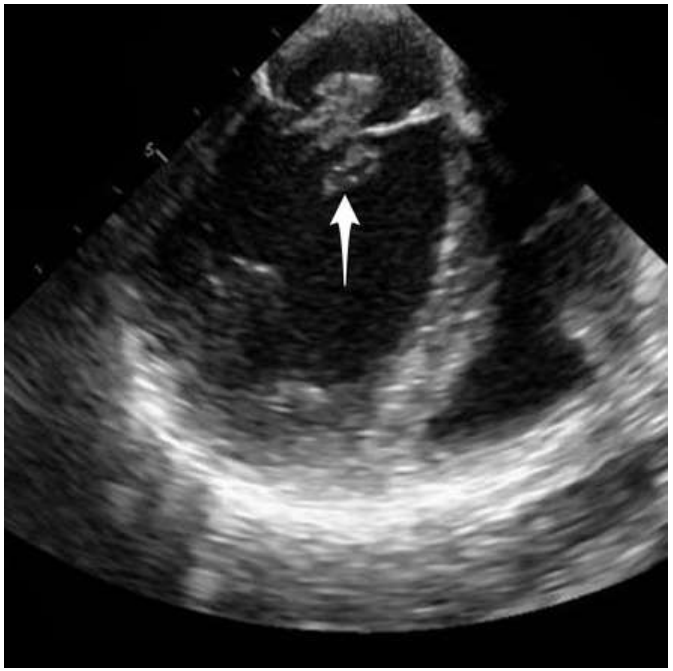
2- presence of underlying cardiac disease



Feature	Acute endocarditis	Subacute endocarditis
Virulence	highly virulent organism	low virulent organism
Most common organism	Staph. aureus	Streptococcus viridans
Underlying cardiac disease	previously normal valve	previously abnormal valve (scarred or deformed)
Clinical course	rapidly developing	Insidious disease
Outcome	High morbidity and mortality	most patients recover after appropriate antibiotic therapy

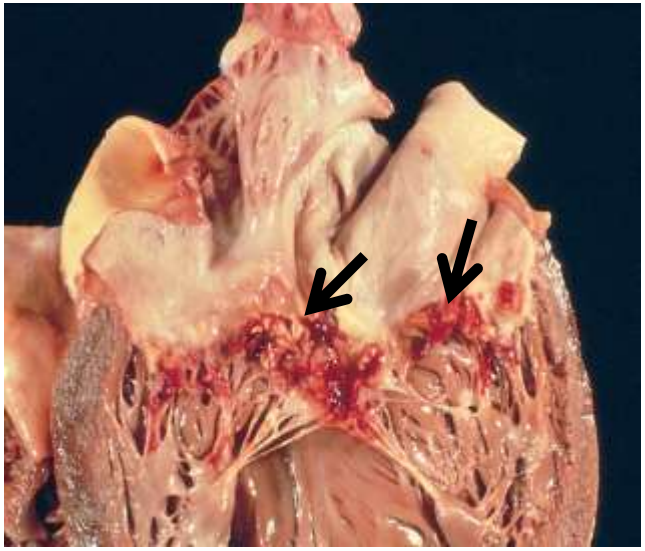
Infective Endocarditis- Clinical Features

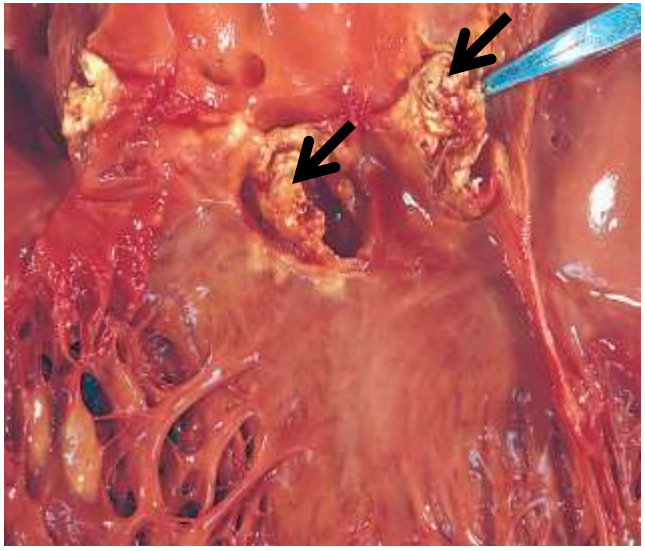
- Fever, chills, weakness, and murmurs
- **Valve vegetations can cause emboli** in different target tissues
- **Diagnosis* = (positive blood cultures + echocardiographic (echo) findings)**
- * depends on certain criteria….



Infective Endocarditis- Morphology

- **Friable, bulky, and destructive vegetations** on heart valves
- Most common: aortic and mitral valves
- Tricuspid valve common in I.V. drug abusers





Clinical Features

- **<u>Complications of IE vegetations:</u>**

1- emboli

2- abscesses

3- septic infarcts

4- mycotic aneurysms

- **Treatment**: long-term (≥ 6 weeks) I.V. antibiotic therapy and/or valve replacement

Infective Endocarditis: Diagnosis

Duke Criteria

- 1994 a group at Duke University standardised criteria for assessing patients with suspected endocarditis
- **Definite**
  - 2 major criteria
  - 1 major and 3 minor criteria
  - 5 minor criteria
  - pathology/histology findings
- **Possible**
  - 1 major and 1 minor criteria
  - 3 minor criteria
- **Rejected**
  - firm alternate diagnosis
  - resolution of manifestations of IE with 4 days antimicrobial therapy or less

Modified Dukes' criteria

- **Major-**
  - 2 positive blood cultures, for an organism known to cause IE
    or
    persistent bacteremia- 2 +ve 12 hours apart or 3 of 4 +ve drawn over 1 hour
  - ECHO evidence- oscillating mass on valve or supporting structures
    or abscess
    or new valvular regurgitation or partial dehiscence of prosthetic valve
- **Minor-**
  - Predisposing factor- cardiac lesion, IVDU
  - Fever >38 °C
  - Vascular phenomenon
  - Immunologic phenomenon
  - +ve blood culture
  - +ve ECHO

Let’s find out?

- Are all people with streptococcal pharyngitis exposed to risk of rheumatic fever?
- In what ways are rheumatic fever and infective endocarditis similar?
- What is different between rheumatic fever and infective endocarditis ?