Valvular Heart Disease - 1
Normal Heart Valves
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
• **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 streptococcal antigens (streptolysin O; DNA-ase)
- culture for streptococci is (-) 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)
  - inflammatory lesions in affected tissues
  - *pathognomonic (diagnostic)* for RF
  - collections of T lymphocytes+ plasma cells+ activated macrophages
Diagnosis of Acute Rheumatic Fever

**Major Criteria**
- J: Joint Involvement
- O: O looks like a heart = myocarditis
- N: Nodules, subcutaneous
- E: Erythema marginatum
- S: Sydenham chorea

**Minor Criteria**
- C: CRP Increased
- A: Arthralgia
- F: Fever
- E: Elevated ESR
- P: Prolonged PR Interval
- A: Anamnesis of Rheumatism
- L: Leukocytosis

**Diagnosis**

- In serum: Elevated anti-streptolysin O titers

**Criteria:**
- 2 Major criteria
- OR
- 1 Major criterion and 2 Minor criteria

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*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