VALVULAR HEART DISEASE

- **Stenosis**: failure of a valve to open completely, obstructing forward flow.
  - almost always due to a chronic process (e.g., calcification or valve scarring).
• **Insufficiency**: failure of a valve to close completely ➔ regurgitation (backflow) of blood.

• *It* can result from disease of either:
  – valve cusps (e.g., endocarditis)
  
  Or - supporting structures (e.g. mitral annulus, tendinous cords, papillary muscles)

• It can be either:
  – Acute ➔ e.g. chordal rupture
  – chronic ➔ e.g. scarring and retraction
Clinical signs of valve disease:

- abnormal heart sounds called *murmurs*
- palpated heart sound (*thrills*) → severe lesions
- specific clinical signs according to involved valve
• Valvular abnormalities can be congenital or acquired.

• The most common congenital valvular lesion is bicuspid aortic valve

• most important cause of acquired valvular diseases is rheumatic fever
• **bicuspid aortic valve:**
  - only two functional cusps instead of the normal three
  - 1% to 2% of all live births
  - associated with a number of genetic mutations
  - early life ➞ Asymptomatic
  - Later ➞ early and progressive degenerative calcification
• mitral valve is the most common target of acquired valve diseases.

• most important causes of acquired valvular diseases are post-inflammatory scarring of the mitral valves and aortic valve due to rheumatic fever (2/3 of all)
Rheumatic fever (Rheumatic Valvular Disease)

• immune- mediated inflammatory disease
• incidence↓ in Western world (improved socioeconomics, rapid diagnosis, and Rx of strept. Pharyngitis)
• Still, important public health in developing countries

**PATHOGENESIS:** hypersensitivity reaction • due to antibodies against group A streptococcal antigenes that are cross-reactive with host antigens (heart; brain; joints; skin)
Rheumatic fever

- Manifestations are seen a few weeks after the pharyngitis or skin infection.

- Major organs involved: heart; joints; skin; and brain.
Rheumatic fever

- **2 phases:**
  - **Acute:** fever; arthritis; CNS symptoms; carditis
  - **Chronic:** cardiac valve disease
Acute rheumatic fever - clinical picture

- 80% of cases are children
- fever; migratory polyarthritis; **carditis**.
- arrhythmias; myocarditis; cardiac dilation; functional mitral insufficiency and CHF.
- Elevated serum titers of streptococcal antigens (**streptolysin O; DNA-ase**)
- **cultures for streptococci are (-) at the time of symptom onset**
MORPHOLOGY- Acute phase

- discrete inflammatory lesions in affected tissues.
- cardiac lesions = **Aschoff bodies** are *pathognomomic* for RF (collections of T lymphocytes, plasma cells, and activated macrophages)
Acute rheumatic heart disease

Aschoff bodies
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**
- Elevated anti-streptolysin O titers
- 2 Major criteria
  - OR
  - 1 Major criterion and 2 Minor criteria
chronic rheumatic carditis - clinical picture

• **Onset:** years/ decades after initial acute episode

• **Consequence:** valve scarring

• cardiac murmurs - CHF - arrhythmias (esp. A. fib.) - thromboembolism (mural thrombi).

• Prognosis: variable.

• Management: Surgical repair or replacement of diseased valves
Chronic phase- morphology

• Inflammation is replaced by **scarring**
• Aschoff bodies **rarely** seen now

• **Consequence → stenosis** (most important functional consequence of chronic RHD)
  - **mitral** valve (most common)
  - aortic disease
  - tricuspid valve
  - **pulmonary** valve

-↓ frequency
Chronic rheumatic heart disease

Scarring and calcifications
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
Infective endocarditis (IE)

• classified into *acute* and *subacute* based on:

1- the virulence of microorganism
2- presence of underlying cardiac disease
### Table: Comparison of Acute and Subacute Endocarditis

<table>
<thead>
<tr>
<th>Feature</th>
<th>Acute Endocarditis</th>
<th>Subacute Endocarditis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Virulence</strong></td>
<td>a highly virulent organism</td>
<td>low virulent organism</td>
</tr>
<tr>
<td><strong>Most common organism</strong></td>
<td>Staph. aureus</td>
<td>Streptococcus viridans</td>
</tr>
<tr>
<td><strong>underlying cardiac disease</strong></td>
<td>previously normal valve</td>
<td>previously abnormal valve (scarred or deformed)</td>
</tr>
<tr>
<td><strong>Clinical course</strong></td>
<td>rapidly developing</td>
<td>Insidious disease</td>
</tr>
<tr>
<td><strong>Outcome</strong></td>
<td>High morbidity and mortality</td>
<td>most patients recover after appropriate antibiotic therapy</td>
</tr>
</tbody>
</table>
MORPHOLOGY

- friable, bulky, and destructive vegetations on heart valves
- most common: aortic and mitral valves
- tricuspid valve common in I.V. drug abusers.
- Complications of vegetations:
  1. emboli
  2. abscesses
  3. septic infarcts
  4. mycotic aneurysms
Infective endocarditis (IE)
Clinical Features

- fever, chills, weakness, and murmurs
- **Fever** is the most consistent sign of infective endocarditis (almost 100%)

- emboli in different target tissues

- Diagnosis = (positive blood cultures + echocardiographic (echo) findings)

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