

VALVULAR HEART DISEASE

• results in stenosis or insufficiency (regurgitation or incompetence), or both.

• <u>Stenosis</u> : failure of a valve to open completely, obstructing forward flow.

- almost always due to a primary cuspal abnormality and is virtually always a chronic process (e.g., calcification or valve scarring).

- <u>Insufficiency</u> : failure of a valve to close completely → regurgitation (backflow) of blood.
- *It* can result from either:
 - intrinsic disease of valve cusps (e.g., endocarditis)
 - disruption of supporting structures (e.g., the aorta, mitral annulus, tendinous cords, papillary muscles, or ventricular free wall) without primary cuspal injury.
 - It can be either: Abrupt→ e.g. due to chordal rupture
 - Insidious→ e.g. due to leaflet scarring and retraction

Acquired Valve disease

• The mitral valve is the most common target of acquired valve diseases.

- Clinical signs of valve disease:
 - abnormal heart sounds called *murmurs*
 - palpated heart sound (*thrills*) → severe lesions
 - clinical signs according to the involved valve

- Valvular abnormalities can be <u>congenital</u> or <u>acquired</u>.
- The most common **congenital** valvular lesion is *bicuspid aortic valve*
- <u>bicuspid aortic valve</u>:
- only two functional cusps instead of the normal three
- 1% to 2% of all live births
- associated with a number of genetic mutations
- Asymptomatic in early life; however, the valve is more prone to early and progressive degenerative calcification

• The most important causes of acquired valvular diseases are post-inflammatory scarring of the mitral valves and aortic valve due to (<u>rheumatic fever</u>)→ 2/3 of all <u>valve disease</u>.

Degenerative Valve Disease

- Degenerative changes include
- Calcifications
- Alterations in the ECM
- Calcific aortic degeneration is the most common cause of aortic stenosis. it typically begins to manifest when patients reach their 70s and 80s; onset with bicuspid aortic valves is at a much earlier age (often 40 to 50 years).

Degenerative Valve Disease



Rheumatic Valvular Disease

- is an acute, immunologically mediated, multisystem inflammatory disease that occurs after group A β-hemolytic streptococcal infections (usually pharyngitis, rarely skin infection).
- Rheumatic heart disease is the cardiac manifestation of rheumatic fever.

Rheumatic Valvular Disease

- valvular inflammation and scarring produces the most important clinical features
- PATHOGENESIS: a <u>hypersensitivity</u> reaction due to antibodies directed against group A streptococcal molecules that also are cross-reactive with host antigens

MORPHOLOGY- acute rheumatic fever

- characterized by discrete inflammatory foci within a variety of tissues.
- Myocardial inflammatory lesions = Aschoff bodies are *pathognomonic* for rheumatic fever ((collections of lymphocytes (T cells), plasma cells, and activated macrophages called **Anitschkow cells** with rare zones of fibrinoid necrosis))
- Anitschkow cells: macrophages with abundant cytoplasm and central nuclei with chromatin condensed to form a slender, wavy ribbon (so-called caterpillar cells).

Acute rheumatic heart disease



MORPHOLOGY- acute rheumatic fever

- acute rheumatic fever→ Aschoff bodies found in any of the three layers of the heart-pericardium, myocardium, or endocardium (including valves), or allover pancarditis.
- Valve involvement \rightarrow fibrin deposition along the lines of closure \rightarrow regurgitation

Chronic rheumatic heart disease



Chronic rheumatic heart disease

- characterized by organization of inflammation and scarring.
- Aschoff bodies are **rarely** seen in **chronic** RHD since they are replaced by fibrous scar
- mitral valves is most commonly affected → fishmouth" or "buttonhole" stenoses
- Microscopic: neovascularization and diffuse fibrosis that obliterates the normal leaflet architecture

- The most important functional consequence of chronic RHD is valvular stenosis (most common) and regurgitation (less common)
- **mitral** valve alone: 70% of cases (most common)
- combined mitral and aortic disease: 25%
- tricuspid valve: less frequent, less severe
- **pulmonic** valve: almost always **escapes** injury.
- Complications of mitral stenosis:
- dilated left atrium atrial fibrillation
- mural thrombi.
- Complications of aortic valve disease:
- left-sided heart failure
- right ventricular hypertrophy and failure.

Acute rheumatic fever- clinical picture

- occurs most often in children 80%
- (20%→ adults; arthritis is the predominant feature)
- principal clinical manifestation is carditis.
- symptoms begin 2- 3 weeks after streptococcal infection:

fever; migratory polyarthritis (one large joint after another followed by spontaneous resolution with no residual disability).

• cultures are (-) for streptococci at the time of symptom onset

Acute rheumatic fever- clinical picture

- serum titers to streptococcal antigens (e.g., streptolysin O or DNAase) are elevated.
- clinical signs of carditis→ pericardial friction rubs; arrhythmias; myocarditis; cardiac dilation; functional mitral insufficiency and CHF.
- less than 1% of patients die of acute rheumatic fever.

The diagnosis of acute rheumatic fever

- = (serologic evidence of previous streptococcal infection + two or more of the so-called *Jones criteria*).
- Jones criteria:
 - (1) Carditis
 - (2) migratory polyarthritis of large joints
 - (3) subcutaneous nodules
 - (4) erythema marginatum skin rashes
 - (5) Sydenham chorea, a neurologic disorder characterized by involuntary purposeless, rapid movements.
- Minor criteria such as fever, arthralgias, ECG changes, or elevated acute phase reactants also can help support the diagnosis.

chronic rheumatic carditis- long-term prognosis

- manifest itself clinically **years or decades** after initial episode of rheumatic fever.
- signs and symptoms depend on which cardiac valve(s) are involved: -cardiac murmurs cardiac hypertrophy
- CHF arrhythmias (esp. A. fib.) thromboembolism (mural thrombi).
- scarred and deformed valves are more susceptible to infective endocarditis (IE).
- prognosis is highly variable.
- Management: Surgical repair or replacement of diseased valves

Infective endocarditis (IE)

- Microbial invasion of heart valves or endocardium, with destruction of underlying cardiac tissues→ cause bulky, friable *vegetations* (necrotic debris+ thrombus+ organisms).
- Common sites of infection: valves, endocardium, aorta, aneurysms; prosthetic devices.
- The vast majority of cases → caused by bacteria.

Infective endocarditis (IE)

- Other cases: fungi, rickettsiae (agents of Q fever), and chlamydial species
- classified into *acute* and *subacute*, based on pace and severity of clinical course
- How? 1- the virulence of the responsible microbe

2- whether underlying cardiac disease is present.

Acute versus subacute

- Acute endocarditis
- a highly **virulent** organism (*S. aureus* is most common)
- attack a previously **normal** valve
- substantial **morbidity** and **mortality** even with appropriate antibiotic therapy and/or surgery.

• Subacute endocarditis

- organisms of **low** virulence ($60\% \rightarrow Streptococcus viridans$)
- a previously **abnormal** valve (e.g. scarred or deformed)
- **Insidious** disease; follows a protracted course of weeks to months; most patients **recover** after appropriate antibiotic therapy

MORPHOLOGY

- both acute and subacute disease → friable, bulky, and potentially destructive vegetations (fibrin, inflammatory cells, and microorganisms) on heart valves
- aortic and mitral valves are the most common sites
- tricuspid valve is a frequent target in I.V. drug abusers.
- Complications:
- 1- emboli (friable nature of the vegetations).
- 2- abscesses at the sites where emboli lodge
- **3- septic infarcts**
- 4- mycotic aneurysms.

Infective endocarditis (IE)



Clinical Features

- Acute →a stormy onset including rapidly developing fever, chills, weakness, and lassitude; murmurs
- Fever is the most consistent sign of infective endocarditis (almost 100%)
- **microemboli** in different target tissues:
- Petechia (skin)
- nail bed (*splinter* hemorrhages)
- retinal hemorrhages (*Roth spots*)

painless palm or sole erythematous lesions (Janeway -

lesions)

- painful fingertip nodules (Osler nodes)

Clinical Features

- **Subacute**: nonspecific fatigue, weight loss, and a flulike syndrome; splenomegaly; murmurs
- Diagnosis = (positive blood cultures + <u>echocardiographic</u> (echo) findings)

Prognosis

- depends on the infecting organism and on whether or not complications develop.
- untreated, infective endocarditis generally is fatal.
- **Treatment**: appropriate long-term (6 weeks or more) antibiotic therapy and/or valve replacement
- Mortality :
- low-virulence organisms \rightarrow cure rate is 98%
- enterococci and *Staph. aureus* \rightarrow *cure rate* 60% to 90%
- aerobic gram-negative bacilli or fungi \rightarrow mortality 50%.
- IE of prosthetic valves → cure rate is worse than genuine valves