

VALVULAR HEART DISEASE

- ▶ results in stenosis or insufficiency (regurgitation or incompetence), or both.
- ▶ **Stenosis** : *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).

- ▶ **Insufficiency** : *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

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 congenital or acquired.
- ▶ The most common **congenital** valvular lesion is *bicuspid aortic valve*
- ▶ **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
 - 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 postinflammatory scarring of the mitral valves and aortic valve due to (rheumatic fever) → 2/3 of all 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.
- ▶ ***valvular inflammation and scarring produces the most important clinical features***
- ▶ PATHOGENESIS: a hypersensitivity 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).

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 all over **pancarditis**.
- ▶ Valve involvement → fibrin deposition along the lines of closure → regurgitation

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
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- ▶ 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.
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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
- ▶ 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 polyarthrititis 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.
- ▶ 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% → *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 abuse.
- ▶ Complications:
 - 1- **emboli** (friable nature of the vegetations).
 - 2- **abscesses** at the sites where emboli lodge
 - 3- **septic infarcts**
 - 4- **mycotic aneurysms.**

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%)
- ▶ **Subacute**: nonspecific fatigue, weight loss, and a flulike syndrome; splenomegaly; murmurs

- ▶ **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*)

- ▶ **Diagnosis = (positive blood cultures + echocardiographic (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 → cure rate is 98%
 - enterococci and *Staph. aureus* → cure rate 60% to 90%
 - aerobic gram-negative bacilli or fungi → mortality 50%.
- ▶ IE of prosthetic valves → cure rate is worse than genuine valves