## CVS module – 3

# Valvular Heart Disease.



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### Anatomy





#### Macro and microscopic appearance





#### Valvular Heart Disease

- Valvular heart disease (VHD) comprises a group of conditions that affect the heart valves, leading to two main types of problems:
- Stenosis is the failure of a valve to open completely, obstructing forward flow, usually result from primary cuspal abnormality such as calcification or valve scarring.
- Insufficiency results from failure of a valve to close completely, thereby allowing regurgitation (backflow) of blood, result from either:
- Intrinsic disease of the valve cusps (e.g., endocarditis).
- Disruption of the supporting structures such as : mitral annulus, tendinous cords, papillary muscles, or ventricular free wall.



- Stenosis or regurgitation may occur alone or together in the same valve.
- Valvular disease can involve only one valve (the mitral valve being the than one valve.



#### Causes

- Valvular abnormalities can be congenital or acquired:
- I. Congenital:
- Bicuspid aortic valve:
- The most common congenital valvular lesion.
- Occurs with a frequency of 1% to 2% of all live births.
- Associated with a number of mutations, including those affecting proteins of the Notch signaling pathway.

#### Bicuspid aortic valve. Cont.

- The two cusps are of unequal size, with the larger cusp exhibiting a midline raphe resulting from incomplete cuspal separation.
- Bicuspid aortic valves are generally neither stenotic nor incompetent through early life; however, they are more prone to early and progressive degenerative calcification that gives rise to stenosis



#### II. Acquired valvular diseases

#### Table 11.4 Etiology of Acquired Heart Valve Disease

Mitral Valve Disease	Aortic Valve Disease
Mitral Stenosis	Aortic Stenosis
Postinflammatory scarring (rheumatic heart disease)	Postinflammatory scarring (rheumatic heart disease) Senile calcific aortic stenosis Calcification of congenitally deformed valve
Mitral Regurgitation	Aortic Regurgitation
Abnormalities of leaflets and commissures Postinflammatory scarring Infective endocarditis Mitral valve prolapse "Fen-phen"induced valvular fibrosis Abnormalities of tensor apparatus Rupture of papillary muscle Papillary muscle dysfunction (fibrosis) Rupture of chordae tendineae Abnormalities of left ventricular cavity and/or annulus Left ventricular enlargement (myocarditis, dilated cardiomyopathy) Calcification of mirral ring	Intrinsic valvular disease Postinflammatory scarring (rheumatic heart disease) Infective endocarditis Aortic disease Degenerative aortic dilation Syphilitic aortitis Ankylosing spondylitis Rheumatoid arthritis Marfan syndrome

#### Pathogenesis

At <u>structural</u> level, valvular heart disease result from:

- Degenerative changes.
- Rheumatic fever .
- Infective (bacterial) endocarditis .
- Others .

#### Degenerative Valve Disease

- Degenerative valve disease is a term used to describe changes that affect the <u>integrity of valvular ECM</u>.
- Degenerative changes include the following:
- 1. Calcifications, which can be cuspal (typically in the aortic valve) or annular (in the mitral valve).
- 2 Alterations in the ECM.
- Changes consist of increased proteoglycan and diminished fibrillar collagen and elastin (myxomatous degeneration).
- 3. Aging related: Due to the repetitive mechanical stresses to which valves (40 million beats per year).

### 1. Calcific Aortic Stenosis

- the most common cause of aortic stenosis.
- can be viewed as the valvular counterparts to age-related arteriosclerosis (same risk factors).
- Mostly is asymptomatic and is discovered only incidentally by viewing calcifications on a routine chest radiograph or at autopsy.
- Rarely, it may lead to valvular sclerosis and/or calcification can be severe enough to cause stenosis, necessitating surgical intervention



#### Calcific Aortic Stenosis



Nodular calcifications fill the cusp pockets



Large nodular calcific deposits in the wall

## Clinically



- The hypertrophied myocardium is prone to ischemia, and angina may develop.
- Systolic and diastolic dysfunction collude to cause CHF.
- Cardiac decompensation eventually ensues.

#### 2. Myxomatous Mitral Valve.

- Non-inflammatory progressive disarray of the valve structure and mechanical integrity caused by an altered synthesis and/or remodeling by type VI collagen.
- Myxomatous degeneration of the mitral valve is a common feature of Marfan syndrome and occasionally occurs in other connective tissue disorders.

#### Myxomatous Mitral Valve





voluminous and thickened leaflets

thickening and proliferation of the spongiosa with pooling of glycosaminoglycan that expands to the fibrosa.

### Infective Endocarditis (IE)

Is a microbial infection of the heart valves or the mural endocardium that leads to the formation of vegetations composed of thrombotic debris and organisms, often associated with destruction of the underlying cardiac tissues.

#### Causative organisms

- Streptococci.
- Staphylococci
- fungi, rickettsiae (agents of Q fever), and chlamydial species.
- The aortic and mitral valves are the most common sites of infection, although the tricuspid valve is a frequent target in the setting of intravenous drug abuse.

#### **Risk factors**

- Pre existing structural heart disease: rheumatic heart disease (RHD), hypertrophic cardiomyopathy (HCM), subaortic stenosis, ventricular aneurysm
- Degenerative valve disease: mitral valve prolapse, bicuspid aortic valve, calcific aortic valve disease
- Cardiac interventions: prosthetic valve, pacemaker, defibrillator, IVDA (right sided endocarditis), indwelling catheter
- Congenital heart disease: small ventricular septal defect, bicuspid aortic valve.

Pathogenesis Experimental evidence indicates that 2 factors are needed to establish endocardial infection:

- Endothelial damage over valve cusp with PATHOGENESIS OF INFECTIVE ENDOCARDITIS platelet deposition (thrombus).
- Episode of bacteremia

Underlying valvular or non valvular structural abnormality Blood flow turbulence and endothelial damage Fibrin deposition and thrombus formation (NBTE) Bacterial growth in thrombus and formation of dense microcolonies Microorganisms induce further platelet deposition by eliciting tissue factor Fibrin deposition, platelet aggregation and microorganism proliferation together form infected vegetation

### Classification

- Acute endocarditis:
- refers to destructive infections, frequently involving a <u>highly virulent</u> organism attacking a previously <u>normal valve</u>.
- It is associated with of <u>substantial morbidity and mortality</u>, even with appropriate antibiotic therapy and/or surgery.
- Subacute endocarditis:
- refers to infections by organisms of <u>low virulence</u> affecting a previously <u>abnormal heart</u>, especially scarred or deformed valves.
- The disease typically appears <u>insidiously</u> and—even if untreated— follows a protracted course of weeks to months; most patients recover after appropriate antibiotic therapy.

#### Morphology



Large vegetation on atrial aspect of valve



•Fresh vegetations contain platelets and fibrin with a rich infiltrate of neutrophils

\* In chronic lesions, vegetations may show varying degrees of organization, vascularization and calcification

### **Clinical features**

- Vague constitutional symptoms such as fever, rigors, night sweats, anorexia, weight loss, arthralgia
- Development of new murmur or change in nature of an existing murmur .

#### Neurological:

Confusion, hemiplegia and sensory dysfunction due to emboli.

#### Renal:

Infarction causes loin pain and hematuria.

Splenomegaly and splenic infarction due to emboli

#### **Clinical features. cont.**



**Osler nodes:** tender lesions found on finger pulps and thenar / hypothenar eminences

**Eyes:** Roth spots (boat shaped hemorrhages with pale centers, in retina) .

#### Prognosis

- ▶ IE generally is fatal.
- The outcome of valvular disease depends on the valve involved, the degree of impairment.
- However, with appropriate long-term (6 weeks or more) antibiotic therapy and/or valve replacement, mortality is reduced
- Antibiotics should not be initiated before three sets of blood cultures have been taken

#### Endocarditis in SLE Libman-Sacks Endocarditis

- Is characterized by the presence of <u>sterile vegetations</u> on the valves of patients with systemic lupus erythematosus.
- The lesions probably develop as a consequence of immune complex deposition and thus exhibit associated inflammation, often with fibrinoid necrosis of the valve adjacent to the vegetation; subsequent fibrosis and serious deformity can result in lesions that resemble chronic rheumatic heart disease.
- These can occur anywhere on the valve surface, on the cords, or even on the atrial or ventricular endocardium.