

## VALVULAR HEART DISEASE

There are two basic physiopathologic abnormalities affecting the cardiac valves:

- 1) Stenosis: failure of a valve to open completely due to thickening or fusion of leaflets, partially impeding forward flow.
- 2) Insufficiency or regurgitation: failure of a valve to close completely, resulting in partial backward flow.

For any given valve, these abnormalities may be pure or mixed (stenosis + insufficiency).  
Clinically, stenosis and regurgitation lead to murmurs.

Effects of valvular disease on the heart:

### MITRAL VALVE STENOSIS:

- small left ventricle
- enlarged left atrium
- left sided congestive heart failure
- pulmonary hypertension
- right sided congestive heart failure

### MITRAL VALVE REGURGITATION:

- hypertrophy and dilatation of the left atrium and left ventricle
- left sided congestive heart failure
- pulmonary hypertension
- right sided congestive heart failure

### AORTIC VALVE STENOSIS:

- left ventricular hypertrophy and dilatation
- left sided congestive heart failure
- pulmonary hypertension
- right sided congestive heart failure

### AORTIC VALVE REGURGITATION:

- left ventricular hypertrophy and dilatation
- left sided congestive heart failure
- pulmonary hypertension
- right sided congestive heart failure

Causes of valve disease by valve and type of lesion:

**MITRAL VALVE STENOSIS:**

- Chronic rheumatic heart disease (CRHD)

**MITRAL VALVE REGURGITATION**

- Acute rheumatic fever (ARF)
  - CRHD
  - Infectious endocarditis
  - Mitral valve prolapse
- Rupture of a papillary muscle or chordae tendinae (secondary to myocardial infarction, mitral valve prolapse)
- Left ventricular dilatation
  - Mitral annular calcification

**AORTIC VALVE STENOSIS**

- CRHD
- Senile calcific aortic stenosis (bicuspid or unicuspid aortic valve)
- Calcification of a congenitally deformed valve

**AORTIC VALVE REGURGITATION**

- Intrinsic Valvular disease
  - Infectious endocarditis
  - CRHD
- Aortic disease
  - Syphilitic aortitis
  - Ankylosing spondylitis
  - Rheumatoid arthritis
  - Marfan Syndrome

Discussion of specific entities follows:

**I. Rheumatic Heart Disease:**

Two forms of the disease:

- **Acute rheumatic fever** (ARF)
- **Chronic rheumatic heart disease** (CRHD)

**A. Acute Rheumatic Fever:**

- ARF occurs 2-3 weeks, usually after streptococcal pharyngitis by certain rheumatogenic strains of group A beta-hemolytic streptococcus, mostly in children.
- ARF is a multisystemic disease; in the heart, it causes a pancarditis.
- Jones criteria (using the clinical and laboratory presentations listed in the appendix of these notes) are used to establish the diagnosis of ARF and to differentiate rheumatic fever from other diseases.

**Jones Criteria**

Major criteria:	Minor criteria:
Carditis	Fever
Polyarthritits	Arthralgia
Chorea	Hx of ARF
Subcutaneous nodules	Elevated ESR, C-RP, anti-streptolysin O, anti-hyaluronidase, anti-streptokinase or leukocytosis
Erythema marginatum	EKG changes

Two major or one major and two minor manifestations, with supporting evidence of a recent streptococcal infection, indicate the probable presence of rheumatic fever.

- The pathogenesis of ARF remains a mystery although the host immune response and genetic factors have been implicated.
- Verrucous vegetations may be seen along the line of closure (most commonly on mitral valve alone, followed by both mitral and aortic valves)
- Within the valves are foci of inflammatory reactions consisting of lymphocytes and histiocytes (valvulitis).
- The pathognomonic histologic change in the heart is the Aschoff body (focus of collagen necrosis surrounded by chronic inflammatory cells and Anitschkow cells with owl-eyed appearance in cross section, caterpillar appearance on longitudinal section), which eventually resolves to a fibrous scar. An Aschoff myocyte is a multinucleated Anitschkow cell. These may be seen in the endocardium, myocardium and pericardium.

#### **B. Chronic Rheumatic Heart Disease (CRHD):**

- Heart valves are involved exclusively.
- There is a lag period of 5-30 years before clinical evidence of valvular dysfunction becomes apparent as a result of repeated bouts of ARF.
- In advanced CRHD, the valvular lesions show fibrosis, calcification, cusp retraction, chordal thickening and fusion, and characteristic commissural fusion ("fish-mouth" orifice).
- The mitral valve alone is involved in 2/3 of cases; both mitral and aortic valves are involved in 1/3 of cases (rarely is there isolated aortic valve involvement).
- Both stenosis and regurgitation result from rheumatic valve destruction. Mac Callum's patch is an area of endocardial thickening in the left atrium due to the regurgitation of blood.
- The characteristic manifestation of CRHD is mitral valve stenosis (in fact, other causes of mitral stenosis are so rare that, for all intents and purposes, the cause of mitral stenosis in the adult is almost without exception CRHD).
- The primary complications of mitral stenosis include atrial fibrillation, congestive heart failure leading to pulmonary hypertension, increased risk of endocarditis, mural thrombi.

#### **II. Calcific Aortic Stenosis:**

- Causes include
  - (1) senile calcific aortic sclerosis: degenerative process commonly seen in elderly patients (calcification occurs before 6<sup>th</sup> - 8<sup>th</sup> decades of life)
  - (2) calcification of a congenital bicuspid aortic valve (calcification occurs before 5<sup>th</sup> - 6<sup>th</sup> decades of life)
  - (3) calcification of a congenital unicuspid aortic valve (calcification occurs before 5<sup>th</sup> decade)
- Calcific aortic stenosis must be separated from aortic stenosis secondary to rheumatic heart disease; in the former there is no commissural fusion, only fibrosis and thickening; there is no accompanying mitral valve abnormality.

- Complications include dizziness, syncope, angina pectoris, arrhythmia, systolic ejection murmur, left ventricular hypertrophy, increased risk of endocarditis and sudden death.

### III. Mitral Annular Calcification (MAC):

- Commonly seen in elderly patients (age > 70) as a result of degenerative process.
- F::M ratio of 4::1; increased prevalence in renal failure (abnormal calcium metabolism).
- Calcification begins in the mitral valve annulus of the posterior leaflet and may extend into the myocardium, the base or body of the valve leaflets, displacing the valve leaflets upward. May be seen radiographically.
- Complications include mitral regurgitation, arrhythmia, and increased risk of endocarditis.

### IV. Mitral Valve Prolapse (MVP):

- A.K.A. midsystolic click-murmur syndrome, Barlow's syndrome, floppy valve syndrome, billowing mitral valve syndrome.
- MVP affects 0.5-3% of the population; higher prevalence in females and Marfan syndrome. Many patients have an associated peculiar "anxiety neurosis".
- The cause appears to be a congenital defect that leads to excessive accumulation of mucopolysaccharide and proteoglycans (myxomatous degeneration) resulting in weakening of valve leaflets. The valve leaflets billow (also described as ballooning or hooding) into the left atrium during systole.
- Most of the patients are asymptomatic; 10-15% have progressive regurgitation over time and may require surgical replacement of the valve.
- The chordae tendinae are also affected; rupture of chordae can lead to acute mitral regurgitation.
- A characteristic feature of this syndrome is the midsystolic click as a result of tensing and stretching of the chordae tendinae and the valve tissue itself. If the valve is incompetent, a late systolic murmur may also be present.
- complications include arrhythmia, atypical angina, sudden death, increased risk of endocarditis, and rupture of a chordae tendinae.

### V. Non-Bacterial Thrombotic Endocarditis (NBTE):

- A.K.A. marantic endocarditis (marantic is from the word wasted).
- Usually occurs in the setting of an underlying malignant (as a paraneoplastic syndrome) or a condition that leads to progressive "wasting" (inflammatory disease, infections, DIC, etc).
- Small sterile vegetations composed of fibrin are present; the underlying valve is normal.
- Vegetations may embolize to distant organs.

### VI. Infective Endocarditis (IE):

- Risk factors are underlying valvular abnormality, prosthetic valve, drug/alcohol abuse, and immunosuppression.
- Mitral valve and/or aortic valves are affected in most of cases resulting in regurgitation.
- Tricuspid valve involvement is often seen in IV drug user or patients with indwelling venous catheter; they often present with pneumonia or septic pulmonary emboli.
- Common microorganisms associated with infective endocarditis are:
  - S. aureus* - IV drug abuse, previously normal valves
  - S. epidermidis* - prosthetic valves
  - S. bovis* - often associated with colon carcinoma
  - S. pneumoniae* - alcoholics, sepsis

Gram-negatives - IV drug abuse, prosthetic valves, diabetes mellitus

Fungi (Candida spp.) - immunosuppressed patients, IV drug abuse

- **Acute infective endocarditis** (20% of cases): most often caused by *S. aureus*, less frequently *S. pyogenes*; usually in a previously normal valve; the course is more rapid with high fever, shaking chills, rapid valvular destruction, septic emboli, septic shock, and high mortality.
- **Subacute infective (bacterial) endocarditis** (SBE) is caused by less virulent microorganisms such as *S. viridans* and *S. epidermidis* (80% of cases); patients usually have history of abnormal valves and often present with vague, nonspecific symptoms over weeks to months (fever, malaise, anorexia, weight loss, arthralgia).

	ACUTE	SUBACUTE
duration of symptoms	< 6 weeks	> 6 weeks
most common organisms	<i>S. aureus</i> and <i>S. pyogenes</i>	<i>S. epidermidis</i> and <i>S. viridans</i>
virulence of organism	high	low
condition of valve before endocarditis	normal valve	diseased valve
type of valvular lesion caused by the infections	vegetations and perforation	vegetations (perforation rare)

- Diagnostic criteria of IE include murmurs, low grade fever, positive blood cultures, and signs of embolization of infective vegetations to distant organs (the famed Osler nodes, Janeway lesions, Roth spots, splinter hemorrhages).
- Complications include congestive heart failure, infarction of organs with infected thromboemboli, glomerulonephritis secondary to immune complex deposits.
- Treatment requires a prolonged course of antibiotic therapy; the regimens are numerous and complex due to the plethora of etiologic agents and susceptibility.
- Routes of penetration of microorganisms include dental procedures, urinary catheterization, obstetric procedures, surgery.
- All patients with underlying valvular abnormalities must have antimicrobial prophylaxis before dental or surgical procedures.

#### **VII. Libman-Sacks endocarditis**

- Vegetations that occur on valves in systemic lupus erythematosus.
- Most common in the mitral valve.
- Usually not clinically relevant.
- Other cardiac lesions in SLE include pericarditis and myocarditis.