Valvular Heart Disease

MITRAL STENOSIS

Pathophysiology

❶ **rheumatic fever**.
❷ **calcific degeneration**,
❸ **malignant carcinoid disease**,
❹ **congenital mitral stenosis**.
❺ **SLE** .

The **increased pressure gradient across** the mitral valve ➔**increases left atrial pressure** with resultant ➔**left atrial dilation**, ➔**onset of atrial fibrillation**, and often ➔**formation of left atrial thrombi**, which may embolize. **Elevation in left atrial pressure**, producing ➔pulmonary venous congestion ➔**irreversible pulmonary arteriosclerosis** and ➔pulmonary hypertension and ➔**right ventricular failure**.

presentation

Patients with mitral stenosis may develop symptoms of

- **exertional dyspnea**.
- **dyspnea at rest** or **orthopnea**.
- **easy fatigability** and **cardiac cachexia**.
- **hoarseness**
- **dysphasia**.
- **peripheral edema**, **hepatic congestion**, **cirrhosis**, and **anasarca**.
- **Atrial thrombi**.
- **Angina**.

Diagnosis

On cardiac examination, the **pulse** may be irregular. The **first heart sound** may be accentuated in early mitral stenosis. The **second heart sound** may be accentuated by pulmonary hypertension, and an **opening mitral snap** may be audible in diastole. A **mid-diastolic rumble** may be audible .

The chest radiograph

- **left atrial enlargement** with **prominence of the left atrial appendage.**
- Enlargement of the left atrium is typically seen on the posteroanterior film as a **double contour visible behind the right atrial shadow.**
- **Cephalization of pulmonary blood flow**, and occasionally the presence of **Kerley B lines**, or even **pulmonary edema** .
- **Right atrial and right ventricular enlargement**. The overall cardiac size may be normal, but enlargement of the left atrium and the pulmonary artery may obliterate the normal concavity between the aorta and the left ventricle, producing a **"straight" left border of the heart**.

The electrocardiogram

- **prominent p-wave**,
- **termed p mitrale**.
- **Atrial fibrillation**,
- **right ventricular hypertrophy**, and **right-axis deviation**

Two-dimensional echocardiography

- **mitral leaflet thickening and calcification**,
- **leaflet immobility**, and **thickening of the subvalvular mitral apparatus**.
- **mitral valve gradient**.
- **mitral valve orifice area**.

Left- and right-sided heart catheterization The gold standard for ❶quantifying the severity of mitral stenosis ❷patients older than 35 years or the ❸presence of risk factors for coronary artery disease in patients likely to need an operation

Treatment

I -NON OPERATIVE :- Medical treatment of mitral stenosis includes

- **prophylaxis for bacterial endocarditis**
- **diuretics**
- **antiarrhythmic agents** *and/or* **electrical cardioversion** to convert atrial fibrillation to sinus rhythm
- **digitalis**, **beta blocking agents,** or **calcium channel blocking agents** to control heart rate in atrial fibrillation.

Current **indications for percutaneous or operative treatment of mitral stenosis include**

1. **Moderate mitral stenosis**.
2. New York Heart Association **(NYHA) Class III or Class IV** symptoms.
3. The onset of **atrial fibrillation**.
4. Worsening **pulmonary hypertension**.
5. **Systemic embolization**.
6. **Bacterial endocarditis**.

❤ **Percutaneous balloon mitral valvotomy** is preferred for most patients with significant mitral stenosis.

- *Relative contraindications* to percutaneous balloon mitral valvotomy

1) The presence of left atrial thrombus.
2) 2+ (moderate) or greater mitral regurgitation.
3) Significant leaflet thickening or calcification.
4) Significant scarring of the subvalvular apparatus.

- *Complications* of percutaneous balloon mitral valvotomy include

1) arterial embolism,
2) acute mitral regurgitation, and
3) cardiac perforation,
4) mortality 2%.

II – OPERATIVE :- Several operative techniques are available for treatment of mitral stenosis.

- **Closed mitral commissurotomy without use of cardiopulmonary bypass**
- **Open mitral commissurotomy on cardiopulmonary bypass**
- **mitral valve replacement** is indicated for significant mitral stenosis coincident with ➔2 + or greater mitral regurgitation or with ➔ severe leaflet or subvalvular calcification or fibrosis not amenable to repair.

MITRAL REGURGITATION

Pathophysiology and Natural History

The **causes** of mitral regurgitation are many and include

- rheumatic fever
- mitral valve prolapse (myxomatous valve degeneration, Barlow's syndrome)
- chordal rupture
- bacterial endocarditis
- calcific valvular degeneration
- hypertrophic cardiomyopathy
- myocardial infarction or ischemia.

Carpentier classified the mechanical causes of mitral regurgitation as
❶ annular dilation
❷ leaflet prolapsed
❸leaflet restriction .
Type I annular dilatation or leaflet perforation with normal leaflet motion.
Type II leaflet prolapse or ruptured chordae tendinae with increased leaflet motion.
Type III restricted leaflet motion.

Diagnosis

Patients with mitral regurgitation may develop **exertional dyspnea**, **fatigue**, **orthopnea**, **paroxysmal nocturnal dyspnea**, **peripheral edema**, and **angina pectoris**.

On ***physical examination***, **jugular venous distention** and an **irregular pulse** may be present. A **holosystolic murmur** heard best at the apex, with radiation into the axilla. The **first heart sound may be less prominent**, and the **second heart sound may have increased splitting**. An **S_3 gallop** may correlate with depressed left ventricular function.

On the ***chest radiograph***, **left atrial and left ventricular enlargement**, **Kerley B-lines**, and **pulmonary edema** may be present.

The electrocardiogram may demonstrate **p-mitrale**, **left ventricular or biventricular hypertrophy**, or **atrial fibrillation.**

Echocardiography, which may detect mitral valve prolapse, ruptured chordae, vegetations, leaflet thickening, and calcification of the mitral valve leaflet or anulus.

Left- and right-sided heart ***catheterization*** is indicated to evaluate mitral regurgitation, ventricular function, and coronary anatomy in most patients.

Treatment

Medical treatment for mitral regurgitation includes

- **prophylaxis for endocarditis**
- **diuretics**
- **vasodilators**
- **digoxin**, **beta blockade**, or **calcium channel blockade** to control heart rate in atrial fibrillation.

Indications for operation include

1) **3+ or greater mitral regurgitation**.
2) **NYHA Class III or Class IV symptoms of heart failure or angina**.
3) **Recent onset of atrial fibrillation**, **Pulmonary hypertension** or an **abnormal response to exercise testing**.
4) **Evidence of impaired left ventricular function**
5) **Endocarditis**.

In those patients in whom mitral valve pathology is amenable to an effective and long-lasting repair,* mitral valve repair *is the procedure of choice for mitral regurgitation.

A relative indication for* mitral valve replacement *instead of repair is significant fibrosis or calcification of the mitral valve leaflets or subvalvular apparatus.

Mitral valve repair has several **advantages** over mitral valve replacement:

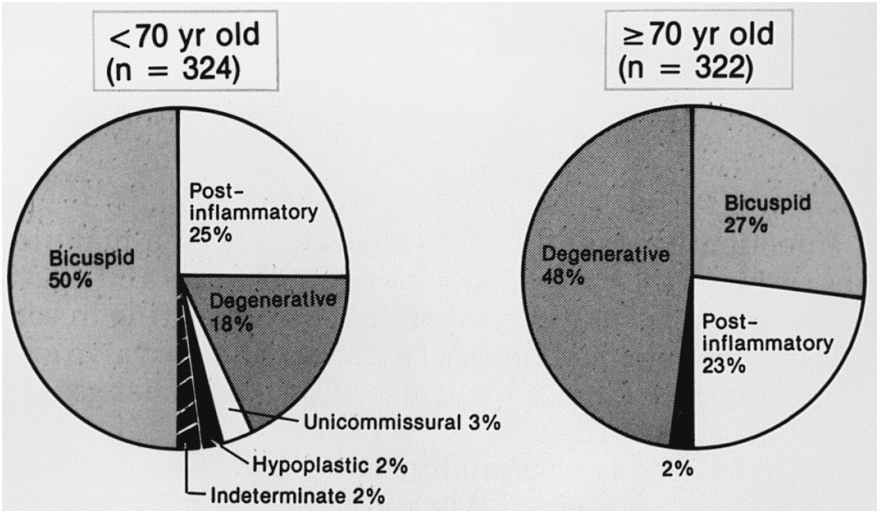
1) **Less impairment of left ventricular function**.
2) **Potentially eliminating the need for chronic anticoagulation**.
3) **Improving durability** relative to biologic prostheses.

AORTIC STENOSIS

Pathologic Anatomy

Aortic stenosis may be **congenital** or **acquired** .

- ***Congenital*** valvular abnormalities may be clinically significant immediately after birth, as with unicuspid and dome-shaped valves. However, congenitally bicuspid valves, usually asymptomatic at birth, generally lead to symptomatic stenosis in the sixth to eighth decades.
- ***Acquired*** aortic stenosis usually results from **rheumatic fever** or from **degeneration** of the valve leaflets.



Causes of aortic stenosis as a function of age

Pathophysiology

As the valve narrows, the appropriate compensatory **response of the left ventricle is hypertrophy**. As the ventricle hypertrophies, it **becomes more stiff** as its **compliance decreases**; a **higher left ventricular end-diastolic pressure is needed to maintain the same volume of cardiac output** .

Diagnosis

Auscultation of the patient with aortic stenosis reveals

- a **systolic murmur**, best heard at the base of the heart
- a slow, prolonged rise in the arterial pulse, "**pulsus parvus et tardus**." The murmer of severe aortic stenosis is soft and high pitched and is often described as a “sea gull” murmer.

The electrocardiogram is notable for

- left ventricular hypertrophy in 85%
- left atrial enlargement in 80% of patients.
- T-wave inversion and ST-segment depression are common.
- Conduction abnormalities are common some patients may develop complete heart block.
- Atrial fibrillation.

The chest roentgenogram is usually normal but may reveal poststenotic dilation of ascending aorta or calcification of the aortic valve.

The severity of aortic stenosis may be estimated by ***Echocardiography***.

The most accurate measure of aortic stenosis is determined by ***Cardiac catheterization***.

Clinical Course

When the aortic valve area diminishes from the normal 3 to 4 sq. cm. to less than 1 sq. cm., patients are usually symptomatic. Once symptomatic, patient survival is limited. The three principal symptoms of aortic stenosis are

- **Angina**
- **Syncope**
- **congestive heart failure**.

Management

Aortic stenosis is a mechanical obstruction to flow from the left ventricle. ***The only effective therapy is operative intervention***. The existence of symptoms is an indication for valve replacement**.**

Percutaneous aortic balloon valvuloplasty for patients not thought to be surgical candidates.

AORTIC INSUFFICIENCY

Pathologic Anatomy

Aortic insufficiency may result from disease of the valve *leaflets* or of the *aortic root*.

- **Rheumatic fever**
- **Annuloaortic ectasia**
- **Marfan's syndrome**.
- **Congenital bicuspid aortic valves**
- **myxoid degeneration of the aortic valve leaflets**, **as seen in Marfan's syndrome, Ehlers-Danlos syndrome, and cystic medial necrosis**
- **Infective endocarditis**.
- **Trauma or dissection of the aortic wall**.
- ***Acute aortic dissection***
- ***chronic dissection***.
- **Congenital aortic insufficiency** may occur **secondary to subaortic ventricular septal defect**

Pathophysiology

the pathophysiology in aortic insufficiency derives from **left ventricular volume overload.** With *well-compensated* aortic insufficiency, exercise may be tolerated because peripheral vascular resistance declines, lowering left ventricular afterload and increasing effective forward flow. At the same time heart rate increases, which shortens diastolic time, thereby decreasing the regurgitant flow. But as the ventricle ultimately *decompensates*, the left ventricular end-diastolic volume increases even without an increase in aortic regurgitant volume. The end-systolic volume increases as the forward stroke volume declines because ventricular emptying is impaired; the ventricle fails.

Diagnosis

The physical examination of patients with aortic regurgitation SHOW

- **wide pulse pressure**.
- The peripheral pulses rise and fall abruptly (**Corrigan's or water-hammer pulse**),
- the head may bob with each systole (**de Musset's sign**), and
- the capillaries visibly pulsate (**Quincke's sign**).
- a **high-frequency decrescendo diastolic regurgitant murmur**.
- A **mid- to late-diastolic rumble may be heard (Austin-Flint murmur)**
- "**Pistol shot**" sounds may be heard with the stethoscope over peripheral arteries.
- Palpation reveals an enlarged heart and a prominent cardiac impulse described as a "**forceful thrust.**

Doppler echocardiography is the most accurate noninvasive technique to determine aortic insufficiency.

The electrocardiogram is usually nonspecific but may reveal **left ventricular hypertrophy** and **left atrial enlargement**. Atrial fibrillation is common later in the course of the disease.

The chest roentgenogram typically reveals an **enlarged cardiac silhouette** with an **enlarged left atrial shadow** and chronic aortic regurgitation, although the cardiac size may not be enlarged with acute aortic regurgitation.

The severity of the aortic regurgitation may be visualized angiographically at ***cardiac catheterization***.

Clinical Course

Because of the compensatory mechanisms discussed earlier, patients with chronic aortic regurgitation may be asymptomatic for long periods of time. However, once these compensatory mechanisms begin to fail, left ventricular dysfunction becomes manifest. Symptoms include **:-**

- **Dyspnea on exertion**
- **Orthopnea**
- **Paroxysmal nocturnal dyspnea**
- **Nocturnal angina**.

Management

Patients with symptomatic aortic insufficiency require surgical therapy, because their prognosis, if treated medically, is only a few years. The surgical procedures include;-

- **Aortic Valve Replacement or Aortic valve repair**
- **Ross Procedure** The Ross procedure involves replacement of the, aortic valve with an autograft from the patient's native pulmonary valve. The resected pulmonary valve is then replaced with a pulmonary homograft

HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY or IHSS

(Idiopathic Hypertrophic Subaortic Stenosis)

Pathologic Anatomy and Physiology

The hypertrophy is asymmetrical and most pronounced in the basilar portion of the interventricular septum, creating a muscular prominence in the subaortic left ventricular outflow tract. Left ventricular ejection into the aorta is impeded, but the exact mechanism of this obstruction to flow is disputed. *Three factors contributing to this impediment include* :-

- the ***mechanical obstruction*** to flow created by the subaortic muscular hypertrophy.
- the increased systolic function of the left ventricle, which creates a ***dynamic obstruction*** .
- ***systolic anterior motion of the mitral valve*** (SAM).

Diagnosis

HOCM is usually identified in patients in the **third and fourth decades of life**, although the majority of patients are **asymptomatic**. When symptoms are present, they are typically

- **Angina**
- **dyspnea on exertion**
- **syncope**.
- Unfortunately, the first clinical manifestation may be **sudden death**, which has been associated with exercise in patients with HOCM.

On ***physical examination***, a **harsh crescendo-decrescendo systolic murmur** is present.

The ***electrocardiogram*** is abnormal and reveals **left ventricular hypertrophy** and ST**-segment and T-wave abnormalities.**

The chest roentgenogram may reveal **cardiac enlargement but is more often normal**.

The echocardiogram is diagnostic .

Management

Medical management

- **beta blockade and/or calcium antagonists**.

- **Disopyramide and amiodarone**.
- **Exercise should be avoided** because of the risk of sudden death.

Surgical intervention

❖ **myotomy-myectomy**,
❖ **mitral valve replacement** .

TRICUSPID VALVE DISEASE.

Pathophysiology and Natural History

Tricuspid regurgitation may be caused by either

- **Functional tricuspid regurgitation** is by far the **most common** form of tricuspid valve disease and generally **results from tricuspid anular dilation due to right-sided heart failure and long-standing pulmonary hypertension.**
- **Organic tricuspid regurgitation** is more **unusual** but may occur after **rheumatic fever**, **carcinoid heart disease**, **bacterial endocarditis**, and **trauma**.

Tricuspid stenosis is **uncommon** but may be caused by **rheumatic heart disease**, **carcinoid heart disease**, **congenital defects**, or **cardiac tumors**.

Diagnosis

On **physical examination**, jugular venous distention may be noted along with hepatic enlargement, ascites, and pedal edema.

The **electrocardiogram** may demonstrate

- **enlarged right atrial a wave**
- **right ventricular hypertrophy**
- **atrial fibrillation**.

Chest radiograph➔ Right atrial and/or right ventricular enlargement and occasional **bilateral pleural effusions** may be present.

Echocardiography can readily visualize thickening or deformity of the tricuspid valve leaflets

Right-sided heart catheterization may confirm the diagnosis of tricuspid valve dysfunction

Treatment

Medical management of tricuspid stenosis or regurgitation includes

- **endocarditis prophylaxis**
- **diuretics**.

Indications for operation for tricuspid valve disease include

1. Moderate tricuspid stenosis or regurgitation concurrent with other valvular disease requiring operation,
2. Severe symptomatic tricuspid stenosis or regurgitation
3. Moderate tricuspid disease with onset of atrial fibrillation.

Surgical procedures include

❤ **tricuspid anuloplasty** for functional tricuspid regurgitation
❤ **tricuspid valve repair** for tricuspid stenosis
❤ **tricuspid valve replacement**
❤ **tricuspid valvectomy** for patients with isolated tricuspid valve endocarditis and a high likelihood of returning to intravenous drug abuse.

PULMONARY VALVE DISEASES

Acquired pulmonary valve disease is rare and usually results from the carcinoid syndrome or rheumatic fever. In both diseases, other valve lesions usually dominate the clinical picture. Pulmonary valvotomy is usually feasible for stenosis. Replacement is almost never performed.