**AORTIC REGURGITATION**

**Background:**

* Primary disease of the aortic valve leaflets, the wall of the aortic root, or both may cause aortic regurgitation (AR).

**Pathophysiology:**

* Chronic AR produces LV volume overload that leads to a series of compensatory changes, including LV enlargement and eccentric hypertrophy.
* The enlarged ventricle is more compliant and is well suited to deliver a larger stroke volume to compensate for the regurgitant aortic flow. It can also accommodate an increased EDV.
* Wall thickness must increase to compensate for the increased ventricular dimensions. These compensatory changes are necessary to minimize or normalize wall stress according to the **LAPLACE LAW**
  + Wall tension/stress is related to the product of intra- ventricular pressure and radius divided by wall thickness).
* Hypertrophy observed in a volume- overload state usually is eccentric, as opposed to concentric hypertrophy observed in a pressure- overload state (i.e., aortic stenosis).
* The increased myocardial mass in a hypertrophic heart enables individual sarcomeres to shorten to a normal degree.
* As long as LV wall stress is maintained in the normal range, the LV preload reserve, contractility, and ejection fraction (EF) remain within the normal range. This is the chronic compensated stage. During this phase of the disease, most patients remain asymptomatic for decades because chronic AR generally is a slow and insidious disease with very low morbidity during a long asymptomatic phase.
* With time, transition from a compensated to a decompensated state marks the progression of the disease. Progressive LV enlargement beyond that required by the valvular regurgitation occurs and is associated with a change of the LV from an elliptical shape to a spherical shape.
* The cause of this pathologic dilatation is not well understood, but loss of the collagen support system that acts as a skeleton for the heart may play a substantial role. These maladaptive changes in the interstitial of the heart are an intricate part of the LV hypertrophy process.
* In addition, diminished coronary flow reserve in this hypertrophied ventricle is thought to result in chronic sub- endocardial ischemia, even in the absence of epi- cardial coronary artery disease (CAD). Eventually, sub- endocardial necrosis and fibrosis occur, along with disruption of the collagen support system, with loss of LV systolic function.
* The neuro- hormonal response complicates the disease state further by its excessive growth stimuli, which are thought to be partially responsible for apoptosis (programmed cell death) of the remaining functional myocytes.
* The vicious cycle continues until the decompensated stage develops over many years. Progressive LV enlargement, spherical LV shape, increased wall stress, decline in the contractility and EF, increased afterload, and decreased diastolic compliance with a rise in end-diastolic pressure characterize this stage. Frequently, development of congestive symptoms heralds this stage, but an insidious deterioration of ventricular function may occur without overt clinical signs.
* In **ACUTE AR**, the normal- sized left ventricle poorly tolerates the sudden large volume imposed on it. The left ventricle poorly accommodates the abrupt increase in end-diastolic volume, and diastolic filling pressure increases rapidly and dramatically. This leads to an acute decrease in forward stroke volume, and, although tachycardia develops as a compensatory mechanism to maintain cardiac output, this often is insufficient. The rise in LV filling pressure is transmitted to the left atrium, pulmonary veins, and pulmonary capillaries, leading to pulmonary edema and congestion. Acute AR usually is severe and rapidly leads to LV decompensation and/or failure and cardiogenic shock

**PHYSICAL SIGNS**

* Hemo- dynamically severe AR causes a **widened pulse pressur**e, often greater than 100 mm Hg, associated with a low diastolic pressure, often less than 60 mm Hg.
* The **de Musset sign** is when patients' heads frequently bob with each heartbeat.
* The **Corrigan pulse** is when patients' pulses are of the water-hammer or collapsing type, with abrupt distention and quick collapse.
* The **Quincke sign** is when light transmitted through the patient's fingertip shows capillary pulsations.
* The **Hill sign** is when popliteal cuff systolic pressure exceeds brachial cuff pressure by more than 60 mm Hg.
* The **Duroziez sign** is when a systolic murmur is heard over the femoral artery when compressed proximally and when a diastolic murmur is heard when the femoral artery is compressed distally.
* The **Müller sign** is systolic pulsations of the uvula.
* The **Traube sign (also called pistol-shot sounds)** refers to booming systolic and diastolic sounds heard over the femoral artery.
* The **apical impulse** in chronic AR is diffuse, hyper- dynamic, and displaced inferiorly and leftward.
* **S3 gallop** correlates with development of LV dysfunction.
* The typical diastolic murmur of AR has a decrescendo shape. A high-frequency early diastolic murmur often occurs in mild AR, whereas a rough holo- diastolic or decrescendo diastolic murmur occurs more commonly in severe AR. The volume and velocity of blood across the incompetent aortic valve tapers off in mid-to-last diastole as the aortic and LV pressures equilibrate. The diastolic murmur of AR is usually best heard adjacent to the sternum in the second to fourth left intercostal space. A concomitant systolic ejection murmur is common in moderate-to-severe AR
* The murmur associated with acute AR may not be impressive. If cardiac decompensation is present, the diastolic murmur of acute AR may be very soft and surprisingly short.
* Antegrade flow across the mitral valve is thought to cause an **Austin Flint murmur, which is a mid- and late-diastolic apical low-frequency murmur** or rumble. The rumble occurs during rapid closure of the mitral valve as flow velocity is increasing across the valve and LV diastolic pressure is rising rapidly because of severe aortic reflux. Its presence indicates severe AR.

**MITRAL REGURGITATION**

**MITRAL REGURGITATION**

Mitral regurgitation (MR) is characterized by an abnormal reversal of blood flow from the left ventricle to the left atrium.

**Pathophysiology:**

-The mitral or bicuspid atrioventricular is composed of the mitral annulus, papillary muscles, chordae tendineae the leaflets.

-Abnormalities in any of these structures can cause MR. The leaflets are continuous with each other at their lines of attachment, called commissures, and are tethered to the left ventricle by the chordae tendineae.

-Chordae tendineae attach to papillary muscles and prevent prolapse of the mitral valve leaflet to prevent reflux of blood into the left atrium.

-MR can be caused by organic disease or a functional lesion (i.e., a normal valve may regurgitate [leak] because of global annular dilatation, focal myocardial dysfunction, or both).

- Congenital MR is rare but is commonly associated with myxomatous mitral valve disease and can be associated with cleft of the mitral valve in persons with Down syndrome.

-In acute mitral valve regurgitation, the incompetent mitral valve allows the ventricular ejection fraction to reflux into the left atrium.

-This volume overload is intensified by the inability of the atrium and ventricle to immediately dilatate, resulting in elevated left atrial and pulmonary venous pressures and acute pulmonary edema.

- The net reduction in forward stroke volume reduces systemic perfusion, can result in hemodynamic deterioration, and can lead to cardiogenic shock.

-In chronic mitral valve regurgitation, the distensibility of the left atrium and ventricle are increased over time.

- This dilatation of the left atrium decreases left atrial pressures, thus increasing preload. The left ventricle dilatates and, via the process of eccentric hypertrophy, generates a larger stroke volume without a significant rise

The LV dilatation may further prohibit the coaptation of the mitral valve leaflets during systolic ejection, leading to progression of LV dilatation and overload.

-Patients with compensated MR may remain asymptomatic for years despite the presence of severe volume overload.

-Ultimately, most people with MR decompensate over the long term.

**Morbidity:** Natural history studies of patients with rheumatic MR have shown 5- and 10-year survival rates of 80% and 60%, respectively.

**Sex:** MR is independently associated with female sex.

**Age:** MR is independently associated advanced age

**Imaging Studies:**

1.Chest radiography

-Evidence of LV enlargement due to volume overload may be observed, although pulmonary congestion, represented by increased pulmonary markings, may not be observed until heart failure has developed.

-Left atrial enlargement also may be observed as a prominence along the right sternal border.

2.Echocardiograph

3.Electrocardiography

-Acute MR is often accompanied by MI

-LV dilatation and hypertrophy

-Left atrial enlargement in chronic mitral valve regurgitation produces a negative P wave in lead V1, but atrial fibrillation may be observed in the late stages.

4.Cardiac catheterization

-Left ventriculography confirms mitral valve regurgitation by demonstrating a flow of contrast into the left atrium.

- LV end-diastolic and end-systolic dimensions can be measured and used to calculate the ejection fraction

--Catheterization can also help detect lesions within the aortic valve, coexistent coronary artery disease through selective coronary artery injection, and other cardiac anomalies such as septal defects

**MANAGEMENT**

Medical therapy

**1.**Afterload-reducing agents, such as nitrates and antihypertensive drugs, are helpful for maintaining the forward-flow state in persons with mitral valve regurgitation.

**2.**If atrial fibrillation is encountered, digitalis therapy is considered.

**3.**Similar to other valvular diseases, prophylactic antibiotics are administered prior to any interventional treatment. However, the current American Heart Association guidelines for endocarditis prophylaxis in patients with mitral prolapse indicate that patients with no murmur and normal leaflets are at low risk; therefore, antibiotic prophylaxis is not necessary.

4.In late-stage mitral valve regurgitation, heart failure develops; diuretics and inotropic agents are administered, and consultation with a specialist in cardiothoracic surgery is arranged.

**5.**The use of balloon counterpulsation should be considered as a preoperative measure

**Surgical Care:**

Indications for surgical interventi

1.Acute mitral regurgitation (MR) with congestive heart failure or cardiogenic shock

2.Class III/IV symptoms (ie, patient symptomatic while at rest or with minimal activity)

3.Class I/II (few or no) symptoms with evidence of deteriorating LV function as evidenced by

* an ejection fraction less than 0.55,
* fractional shortening less than 30%
* either the end-diastolic diameter approaching 75 mm or the end-systolic diameter approaching 50 mm

4.Systemic emboli

5.End-systolic volume index greater than 60 mL/m2 - Most commonly used parameter

**Clinical presentation**

**History:**

-Mitral regurgitation can be tolerated for many years.

-The initial symptoms of dyspnea and fatigue can rapidly progress to orthopnea and paroxysmal nocturnal dyspnea

-Patients with anginal-type pain may have underlying ischemia.

-In those with Mitral Valve Prolapse, palpitations and atypical chest pain are the most frequent complaints. Two thirds are female, often with an underlying panic disorder.

-With underlying coronary artery disease (CAD), regurgitation usually is associated with symptoms of angina pectoris.

-Regurgitation also can develop acutely with myocardial infarction, secondary to papillary muscle rupture..

-When mitral regurgitation is due to left ventricular dilatation and altered valve function, patients often have chronic left-sided heart failure.

-In acute mitral regurgitation from sudden disruption of the mitral valve, the symptoms are due to acute pulmonary edema.

**Physical:**

-With chronic MR, the characteristic holosystolic apical murmur, which radiates to the left axilla and sternal border, may be accompanied by a ventricular gallop (signifying LV dysfunction) followed by an early diastolic rumble caused by the large inflow of blood from a dilatated left atrium.

-If the MR is caused by LV dilatation and depressed ventricular contractile function, this murmur may be mid, late, or holosystolic and may be accompanied by the aforementioned LV (S3) gallop. In this setting, the murmur is usually grade II/VI or less.

-With acute mitral valve regurgitation, a harsh murmur, usually grade III or IV/VI, is heard and is accompanied by a palpable thrill at the apex of the heart.

**Causes:**

**Acute MR**

1.Ruptured chordae or papillary muscle due to acute myocardial infarction or trauma

2.Perforation of the mitral valve leaflet

3.Acute failure of a prosthetic valve

**Chronic MR**

1.Rheumatic heart disease

2.Mitral valve prolapse

3.Coronary artery disease

4.Annular calcification

5.Connective-tissue disorder -Ehlers-Danlos syndrome

Marfan syndrome**,** Osteogenesis imperfecta **,**Systemic lupus erythematosus (SLE)

6.LV dilatation

7.Prosthetic valves

**Surgical options**

-Mitral valve reconstruction with mitral annuloplasty, quadratic segmental resection, shortening of the elongated chordae, or posterior leaflet resection

-Mitral valve replacement with either a mechanical valve (requiring lifelong anticoagulation) or a bioprosthetic porcine valve

**MITRAL STENOSIS**

**Introduction**

-Mitral stenosis (MS) is a narrowing of the inlet valve into the left ventricle that prevents proper opening during diastolic filling.

-Mitral valve leaflets that are typically thickened, commissures that are fused, and/or chordae tendineae that are thickened and shortened.

-The most common cause of mitral stenosis is rheumatic fever (RF).

-After the initial episode of RF, a latency period of 20-40 years occurs until the onset of symptoms. The natural history of mitral stenosis is typically progressive, with a slow and stable course early on, followed by progressive acceleration in the later years.

**Pathophysiology:**

-The serum of patients with RF contains antibodies to the type 5 streptococcal M protein, which cross-reacts with myocardial tissue.

-Pathologic examination of the mitral valve at this time reveals proliferation of fibroblasts and macrophages.

-Subsequent disease may occur as a consequence :

1. Healing of the rheumatic process
2. Sub clinical repetitive rheumatic insults
3. Chronic rheumatic activity
4. Progressive hemodynamic stresses on the traumatized valve

-Some patients experience a chronic stable disease, while others have an accelerated course necessitating early surgical intervention.

-The normal area of the mitral valve orifice is 4-6 cm2, Narrowing of the valve area to less than 2.5 cm2 impedes the free flow of blood and causes a build up of left atrial pressure (LAP) to promote normal transmitral flow volume.

-Critical mitral stenosis occurs when the opening is **reduced to 1 cm2**. At this stage, a LAP of 25 mm Hg is required to maintain a normal cardiac output.

-With progressive stenosis, critical flow restriction reduces left ventricular output.

-The increase in LAP also enlarges the left atrium and raises pulmonary venous and capillary pressures.

- As the disease evolves, chronic elevation of LAP eventually leads to pulmonary hypertension, tricuspid and pulmonary valve incompetence, and secondary right heart failure.

**Prevalence**

Both RF and mitral stenosis remain common in developing nations, and progression of mitral stenosis tends to be more rapid .

**Mortality/Morbidity:**

Overall, the 10-year survival of untreated patients with mitral stenosis is 50-60%, depending on symptoms at presentation.

-In untreated patients, the causes of death are as follows:

* Progressive heart failure in 60-70%
* Systemic embolism in 20-30%
* Pulmonary embolism in 10%
* Infection in 1-5%

**Physical examination**

-Signs of left heart failure-Respiratory distress, evidence of pulmonary edema (eg rales),Digital clubbing

-Low volume pulse

-Pulse may be irregular due to artrial fibrillation.

-Cardiac examination of stenotic mitral valve (best at the apex with the patient in the left lateral recumbent position)

* Observe for active precordium
* Feel the tapping apex-prominent 1st heart sound
* Palpable diastolic thrill
* Prominent 1st heart sound-(An accentuated S1) followed by S2
* Opening snap (OS)
* Characteristic mid-diastolic low-pitched, rumbling murmur best heard at the apex with the patient in left lateral position with the breath held in expiration and with the bell of the stethoscope.

-The duration, and not intensity, of the murmur is a guide to the severity of mitral valve narrowing. However, murmur may diminish in intensity as the stenosis increases.

-The OS and diastolic murmur are often reduced during inspiration and augmented during expiration. Amyl nitrite inhalation, coughing, isometric or isotonic exercise, and sudden squatting all are useful in accentuating a faint or equivocal murmur of mitral stenosis.

Signs of right heart strain/failure eg

-Right ventricular lift may be felt–RV hypertrophy

-A loud pulmonic closure (P2) may be noted in the left parasternal region in patients with pulmonary HTN

-Jugular venous distention, ascites, hepatomegaly, and peripheral edema may be noted.

-Auscultation may reveal a systolic murmur of TR**,** a **Graham Steele** murmur of Pulmonary Regurgitation (a high-pitched, decrescendo, early diastolic murmur of pulmonary insufficiency), and an S4.

-Large a waves in the jugular pulse indicate the presence of pulmonary hypertension or tricuspid stenosis; with atrial fibrillation, the A wave is absent.

Signs of complications from mitral stenosis

-Endocarditis - Fever, changing murmur, and classically splinter hemorrhage, finger clubbing, petechiae, Roth spots, Osler nodes, or Janeway lesions

-Atrial fibrillation

-Systemic embolization

Other findings

-A holosystolic murmur of mitral regurgitation may accompany the valvular deformity of mitral stenosis.

-“Mitral facies” characterized by pinkish purple patches on the cheeks may be present.

NB. Differentiating an Austin Flint murmur from a mitral stenosis rumble, since the quality of the murmurs is similar.

-The Austin Flint murmur is due to the convergence of the mitral orifice flow and the jet of aortic regurgitation, which results in audible turbulence.

**ECG**

1-Left atrial enlargement: **P mitrale, a wide (<0.12 s)** and notched or bifid P wave in limb lead II.and/or a biphasic P wave in lead V1 with a wide negative deflection greater than 0.04 seconds.

NB.

-P**-Pulmonale is a tall, peaked P wave** (height at least 2.5 mm) in limb lead II, associated with right atrial enlargement from either severe pulmonary hypertension or concomitant tricuspid stenosis or regurgitation.

2-**Atrial fibrillation** usually develops in the presence of preexisting left atrial enlargement.

3-With severe pulmonary hypertension, **right-axis deviation** and **right ventricular hypertrophy** can be seen.

The ECG of right ventricular hypertrophy typically shows tall R waves in the right chest leads, and the R wave may be taller than the S wave in lead V1.

4-In addition, right-axis deviation and right **precordial T-wave inversions** are often present.

**Cardiac catheterization**

-Measures absolute left- and right-sided pressure when pulmonary artery pressure elevation is out of proportion to mean gradient and valve area.

**-**Coronary angiography may be performed in selected patients.

**MANAGEMENT**

Control symptoms

* Treat elevated pulmonary venous pressure
* LV systolic dysfunction
* Heart failure.

General

1-**Bed rest**-avoid strenuous physical activity.

2-**Sodium intake** should be **restricted**

3- Maintenance doses of **oral diuretics?** should be continued.

4-**Beta-blockade** may be useful for patients ONCE oedema cleared

**ACE inhibitors** for those in CCF

6-Manage atrial fibrillation Patients with atrial fibrillation are especially likely to show symptomatic improvement when heart rates are controlled with medication. **Digoxin h**as been popular for rate control in atrial fibrillation

7-Rate control should be maintained with digitalis, a beta-blocker, or a calcium channel blocker. Digitalis works well but has a slow onset of action. A calcium channel blocker may become the preferred agent for its lower tendency to convert the rhythm and risk embolization.

7-**Anticoagulation** is necessary in patients who are unable to maintain normal sinus rhythm.

-Anticoagulation is also beneficial for cases with normal sinus rhythm with a prior embolic event or a left atrial dimension greater than 55 mm noted by echocardiography

- Patients with atrial fibrillation are especially likely to show symptomatic improvement when heart rates are controlled with medication. Digoxin has been popular for rate control in atrial fibrillation

**Sex:**

Although the attack rate for RF is roughly equal among genders, mitral stenosis is 2-3 times more common in women than in men.

**Age:**

Occasionally, patients can become symptomatic before age 20 years. Progression more rapid in the developing countries.

**CLINICAL PRESENTATION**

**History**

-Inquire about the history suggestive of acute RF

* Sore throat
* Abnormal movement
* Skin rash or skin swellings
* Multiple and shifting joint pains

-Also history of repeated streptococcal pharyngitis or scarlet fever in childhood.

-Many patients are asymptomatic.

-Some patients may develop symptoms during physiologic stress such as infection, exercise, fever, or pregnancy.

-In late disease- symptoms of left heart failure) are most common

* Exertional dyspnea
* Orthopnea
* Paroxysmal nocturnal dyspnea
* Cough and wheezing. Attacks of frank pulmonary edema may occur.
* Hemoptysis from pulmonary venous hypertension which results in rupture of anastomosis between bronchial vein

-Chest pain due to right ventricular ischemia. And later with right sided failure then oedema,distended neck veins and tender hepatomegaly. If unrecognized may lead to cirrhosis of the liver and ascites.

-**Ortner syndrome** may occur if an enlarged left atrium impinges on the left recurrent laryngeal nerve, causing hoarseness.

-Patients may present with complications of mitral stenosis.

-i) New-onset atrial fibrillation-present as palpitations. Atrial fibrillation is common –about 40% of those with significant stenosis, and it increases in frequency with age and with the increasing size of the left atrium

ii)Systemic embolism- from left atrial thrombi Up to 70% of clinically evident embolic events involve the central nervous system, resulting in stroke, transient ischemia, or death.

-An age greater than 35 and the presence of atrial fibrillation are the most significant associated risk factors for embolism in mitral stenosis patients;

iii)Infective endocarditis

-Precipitant of the heart failure if already on treatment

FAILURE- **F-F**orgotten medication,

**A**-**A**rrythmia/Anaemia,

**I**-**I**schaemia/Infarct/Infection

**L-L**ifestyle (Na and fluid uptake)

**U**-**U**pregulation (increased CO in pregnancy, hyperthyroidism)

**R-R**enal failure ,**E-E**mbolus

-The administration of amyl nitrite causes a marked transient fall in systemic vascular resistance, with an.

increase in heart rate and stroke volume.

-A diastolic rumble caused by mitral stenosis will increase in intensity, whereas the Austin Flint murmur will decrease as the degree of aortic regurgitation is reduced by a fall in systemic blood pressure.

-It should not be given to patients with significant aortic stenosis, unstable angina pectoris, or other conditions in which even brief hypotension could exacerbate the condition.

**Etiology**

**1.**Rheumatic fever (most common, all others are rare)

**2.**Congenital mitral stenosis

3.Degenerative conditions in the elderly.(calcification of the mitral valves)

**4.**Systemic lupus erythematosus (SLE) **and** Rheumatoid arthritis (RA)

**5.**Malignant carcinoid

**6.**Mucopolysaccharidoses (of the Hunter-Hurler phenotype)

**7.Others-**Fabry disease**,**Whipple disease**,**Methysergide

**Imaging Studies:**

**CXR**

-Cardiomegaly with

1)Straightening of the aortic knuckle due to LA enlargement

2)Splaying of the carina-LA enlargement

3) Double shadow by the LA and RA on the right border of the heart.

4) Rarely, calcification of the mitral valve may be seen.

5) Redistribution of blood flow to the upper lung fields-vascular markings

6) Kerley B-lines (short peripheral lines, perpendicular to the pleura) are found in most patients with pulmonary venous hypertension.

Kerley A-lines (long, dense lines radiating from the hilum) are seen in patients with severe, chronic mitral stenosis)

7) Long-standing mitral obstruction may lead pulmonary hemosiderosis with military appearance of lung fields

8) Indentation of the esophagus on Barium swallow

**Echocardiography**

4-modes includeM-mode (motion-mode), 2-dimensional Doppler, and transesophageal echocardiography

-Morphology of the mitral valve

-Measuring orifice size and chamber sizes

-Detailing leaflet mobility

-Thickness of the wall

-Calcification, fusion, and appearance of the commissures

-Ventricular wall motion and ejection fraction

-It provides anatomic and functional information on cardiac chambers and facilitates recognition of other structural abnormalities.

-Transvalvular pressure gradient and pulmonary arterial pressure and determines whether mitral regurgitation, aortic regurgitation, and other valvular abnormalities coexist

5-Prevent recurrent acute rheumatic disease.

Monthly Bezathine Penicillin

**Surgical management**

Definitive management is valve replacement.

Other options

* Percutaneous Mitral Balloon Valvuloplasty

Closed or open mitral commissurotomy