Valvular diseases

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Mitral Stenosis

**DEFINITION**

- Mitral stenosis is condition of pathological narrowing of mitral valve.
- Normal square area of mitral valve is 4-6 cm$^2$. When it is reduced to half of it, significant hemodynamic changes take place.

![Diagram of Mitral Valve Stenosis](image_url)
Etiology:

- Rheumatic fever - decreased in industrialized countries
- Scarring & fusion of valve apparatus
- Pure or predominant MS occurs in approximately 40% of all patients with rheumatic heart disease
- Two-thirds of all patients with MS are female.
Other, less common etiologies:

- malignant carcinoid disease
- systemic lupus erythematosus
- rheumatoid arthritis
- Fabry disease
- congenital mitral stenosis
- degenerative aortic stenosis (calcific extension - causing mitral annular/leaflet calcification)
Symptoms:

- Fatigue
- Shortness of breath during activities (climbing stairs or even making a bed)
- Sudden awakening from sleep with severe shortness of breath
- Palpitations (irregular or forceful heartbeats)
- Swelling of the ankles
- Long-lasting cough or coughing up blood
**Pathophysiology**

- **Normal valve area:** 4-6 cm\(^2\)
- **Mild mitral stenosis:**
  - MVA 1.5-2.5 cm\(^2\)
  - Minimal symptoms
- **Mod mitral stenosis**
  - MVA 1.0-1.5 cm\(^2\) usually does not produce symptoms at rest
- **Severe mitral stenosis**
  - MVA < 1.0 cm\(^2\)
Mitral stenosis (MS) is characterized by obstruction to left ventricular inflow at the level of mitral valve due to structural abnormality of the mitral valve apparatus typically occurs decades after the episode of acute rheumatic carditis.
acute insult leads to formation of multiple inflammatory foci (Aschoff bodies, perivascular mononuclear infiltrate) in the endocardium and myocardium

small vegetations along the border of the valves may also be observed

the valve apparatus becomes thickened, calcified, and contracted, and commissural adhesion occurs, ultimately resulting in stenosis
Severe mitral stenosis - occurs with a valve area of less than 1 cm$^2$.

As the valve progressively narrows - the resting diastolic mitral valve gradient, left atrial pressure - increases.

This leads to transudation of fluid into the lung interstitium and dyspnea at rest or with minimal exertion.

Hemoptysis may occur if the bronchial veins rupture and left atrial dilatation increases the risk for atrial fibrillation and subsequent thromboembolism.
Pulmonary hypertension may develop as a result of retrograde transmission of left atrial pressure, pulmonary arteriolar constriction, interstitial edema.

As pulmonary arterial pressure increases, right ventricular dilation and tricuspid regurgitation may develop, leading to elevated jugular venous pressure, liver congestion, ascites, and pedal edema.
Physical Examination

- First heart sound (S1) is accentuated and snapping
- Opening snap (OS) after aortic valve closure
- Holodiastolic murmur with presystolic accentuation – low and lasts throughout diastole
- Best murmur is heard in the left lateral position
- Pulse is small - pulsus parvus
Progressive, lifelong disease
Usually slow & stable in the early years.
Progressive acceleration in the later years
20-40 year latency after rheumatic fever to symptom onset
Complications

- Atrial dysrhythmias
- Systemic embolization (10-25%)
  - Risk of embolization is related to age, presence of atrial fibrillation, previous embolic events
- Congestive heart failure
- Pulmonary infarcts
- Hemoptysis
  - Massive - ruptured bronchial veins (pulm HTN)
  - Streaking/pink froth: pulmonary edema, or infection
- Endocarditis
- Pulmonary infections
ECG:
- In mitral stenosis the ECG is often normal.
- Pathological changes in the ECG include:
  - left atrial hypertrophy:
    - causes P mitrale
    - increased voltage in the later part of the P wave gives it a large, bifid appearance in leads II, III and aVF
  - atrial fibrillation is present in 60 to 70%
  - right ventricular hypertrophy:
    - a dominant R wave in V1 and V2 indicates pulmonary hypertension
  - right axis deviation
Echocardiography

- diagnosis of Mitral Stenosis
- hemodynamic severity
  - mean gradient, mitral valve area, pulmonary artery pressure
- right ventricular size and function.
- valve morphology to determine suitability for percutaneous mitral balloon valvuloplasty
- Diagnosis of concomitant valvular lesions
- Reevaluation of patients with known MS with changing symptoms or signs.
- F/U of asymptomatic patients with mod-severe MS
Treatment

- Medical
  - Diuretics for LHF/RHF
  - Digitalis/Beta blockers/CCB: Rate control in A Fib, A flutter
  - Anticoagulation: In A Fib, A flutter
  - Endocarditis prophylaxis

- Balloon valvuloplasty
  - Effective long term improvement

- Surgical
  - Mitral commissurotomy
  - Mitral Valve Replacement
    - Mechanical
    - Bioprosthetic
Mitral regurgitation

second most frequent valve disease

**Etiology:**

**Organic**
- Degenerative
- Rheumatic
- Endocarditis
- Congenital

**Functional**
- Ischemic
- Dilated cardiomyopathy
- Hypertrophic cardiomyopathy
Definition

Mitral regurgitation is defined as an abnormal reversal of blood flow from the left ventricle to the left atrium. It is caused by disruption in any part of the mitral valve apparatus.
Anatomy of the Mitral Apparatus

- Leaflets
- Annulus
- Chordae tendinae
- Papillary muscles
- Left ventricle
**Pathophysiology**

- Blood abnormally flows backward from the left ventricle to the left atrium, both chamber's volume increases.

- Since significant volume of blood is flowing retrograde, forward cardiac output is decreased despite the left ventricular ejection fraction appearing normal.
Thus the ejection fraction, which is usually assumed to represent forward ejection of blood, may be normal even in the presence of forward cardiac failure.

Due to the increased volume, left atrial pressure also increases which leads to compensatory left atrial enlargement and can predispose to atrial arrhythmias.
**Symptoms**

*slowly progressive* - *take years for symptoms to develop asymptomatic patients in the initial stages*

- the first symptoms are usually related to exercise intolerance or dyspnea on exertion.

- Signs of left and right heart failure occur late in disease
left heart failure results in symptoms related to the low cardiac output - with physical activity the heart demands increased cardiac output which is not able to be met in states of heart failure and thus left heart pressures increase significantly causing this transient pulmonary edema.

the most common cause of right heart failure is left heart failure. Right heart failure symptoms include lower extremity dependant edema. When the legs are elevated at night, the fluid redistributes centrally causing pulmonary edema resulting in orthopnea (dyspnea while laying flat) or paroxysmal nocturnal dyspnea (PND). Hepatic congestion can occur causing right upper quadrant abdominal pain.
Patients may occasionally present with the onset of cardiac arrhythmias, most commonly atrial fibrillation. Other less common symptoms include hemoptysis, thromboembolism, or symptoms of infectious endocarditis.

Acute mitral regurgitation presents with dramatic symptoms. Syncope can occur from sudden hypotension related to cardiogenic shock. Marked dyspnea from pulmonary edema may be present.
Physical Examination

Palpation: prominent LV filling wave may be present.

Auscultation:

- $S_1$ may be diminished in acute MR and chronic severe MR with defective valve leaflets
- wide splitting of $S_2$ may occur due to early closure of the aortic valve
- $S_3$ may be present due to LV dysfunction or as a result of increased blood flow across the mitral valve.

murmurs

Quality: Usually high-pitched, blowing

Location: best heard over the apex; usually radiates to the left axilla

Duration: Usually holosystolic
Chest radiography

- Evidence of LV enlargement due to volume overload may be observed (particularly in chronic MR)
- pulmonary
- Left atrial enlargement
- straightening of the left cardiac border due to the large left atrial appendage
Mitrální srdce

- Mitrální: trojúhelníkovitý s napřímenou levou stranou konturou
Color Flow Evaluation MR

- **Mild Central MR**
  - Area: < 4 cm²
  - LA Area: < 10%

- **Severe Central MR**
  - Area: > 8 cm²
  - LA Area: > 40%

- **Severe Eccentric MR**
Aortic stenosis

- is caused by narrowing of the orifice of the aortic valve and leads to obstruction of left ventricular outflow

**Etiology:**

- congenital (unicuspid or bicuspid valve)
- calcific (due to degenerative changes)
- rheumatic
<table>
<thead>
<tr>
<th>Clinical manifestation</th>
<th>Causes</th>
<th>Significance</th>
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<tbody>
<tr>
<td>Decreased exercise tolerance due to exertional dyspnea or fatigue</td>
<td>Diastolic dysfunction</td>
<td>If early indications of aortic stenosis are not recognized, can delay diagnosis and treatment</td>
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<tr>
<td></td>
<td>Decreased cardiac output with exercise</td>
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<tr>
<td>Angina</td>
<td>Increased left ventricular workload and oxygen consumption</td>
<td>May occur with or without coexisting coronary artery disease</td>
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<td></td>
<td></td>
<td>Commonly precipitated by exertion and relieved with rest</td>
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<tr>
<td></td>
<td></td>
<td>Mean survival after symptom onset 5 years if no surgical repair of aortic valve</td>
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<tr>
<td>Syncope</td>
<td>May be precipitated by high left ventricular pressures causing acute baroreceptor-activated vasodilation leading to decreased cardiac output or by an inability to increase stroke volume, when needed, through a narrow, stiff aortic valve</td>
<td>Usually occurs during exercise</td>
</tr>
<tr>
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<td>Mean survival after symptom onset 3 years if no surgical repair of aortic valve</td>
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<tr>
<td>Heart failure</td>
<td>Diastolic dysfunction resulting in pulmonary congestion and dyspnea</td>
<td>Most ominous symptom of aortic stenosis</td>
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<td>Mean survival after symptom onset 2 years if no surgical repair of aortic valve</td>
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*a Based on information from Carabello and Paulus.*
Aortic Stenosis - Physical Exam

- crescendo-decrescendo, midsystolic ejection murmur heard best at the right upper sternal border radiating to the neck and carotid arteries

- S2 heart sound is often paradoxically split in patients with aortic stenosis due to the significantly delayed closure of the aortic valve resulting from the increased time needed to complete LV systole.

- The phenomenon known as "pulsus parvus et tardus" refers to a weak (parvus) and delayed (tardus) carotid upstroke.
<table>
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<th>Study</th>
<th>Purpose</th>
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| Doppler echocardiography          | Estimation of severity of aortic stenosis, left ventricular size, and ejection fraction  
|                                   | Estimation of pulmonary pressures, aortic valve gradient, aortic valve area  
|                                   | Assessment of thickening of aortic valve leaflet, reduced leaflet motion, reduced valve opening |
| Cardiac catheterization           | Assessment of coronary arteries to determine need for simultaneous coronary artery bypass surgery and aortic valve replacement  
|                                   | Direct measurement of left ventricular and ascending aortic pressures to determine aortic valve pressure gradient  
|                                   | Determination of left ventricular systolic pump function quantified by measuring left ventricular end-diastolic and end-systolic volumes, and ejection fraction |
| 12-Lead electrocardiography       | Evidence of left ventricular hypertrophy: Increased R-wave amplitude of the QRS complex in lead V₆, increased S-wave amplitude in lead V₁  
|                                   | ST-segment depression and T-wave inversion in leads facing the left ventricle: I, aVL, V₅, and V₆ |
| Chest radiography                 | Determination of heart size  
|                                   | Detection of calcification in the aortic valve (lateral view)  
|                                   | With heart failure, enlarged heart size from dilatation of left atrium and left ventricle, venous congestion, and pulmonary edema |
| Stress testing                    | Determination of the degree of exercise tolerance  
|                                   | Distinguish between asymptomatic and symptomatic aortic stenosis |
| Brain natriuretic peptide         | Determination of severity of increased left ventricular pressure and volume overload  
|                                   | Distinction between cardiac and noncardiac dyspnea |

*Based on information from Kurtz and Otto,¹ Mookadam et al,²⁷ and Bergler-Klein.²⁸*
<table>
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<tr>
<th>Procedure</th>
<th>Indication</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>Aortic valve replacement</td>
<td>Symptomatic severe aortic stenosis</td>
<td>Aortic valve is removed and a new valve (mechanical or biological) is sewn to the annulus of the native valve</td>
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<tr>
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<td>Severe aortic stenosis with ejection fraction &lt;50%</td>
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<tr>
<td></td>
<td>Severe aortic stenosis and a need for any other heart surgery</td>
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<tr>
<td>Balloon aortic valvuloplasty</td>
<td>Bridge to aortic valve replacement in patients in unstable condition</td>
<td>A balloon is placed across the stenotic valve and inflated and deflated several times per second to widen the valve annulus and reduce degree of stenosis</td>
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<td></td>
<td>Palliative to reduce symptoms when surgery is high risk</td>
<td></td>
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<tr>
<td>Transcatheter aortic valve</td>
<td>Seriously ill patients who are not candidates for conventional aortic valve replacement surgery</td>
<td>Replacement valve inside a stent that is deployed over the diseased aortic valve annulus via a transapical or transcatheter approach</td>
</tr>
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Aortic regurgitation (AR)

- occurs when blood flows backwards across the aortic valve from the aorta into the left ventricle (LV) during diastole.

- This abnormal backflow of blood leads to pathologic changes in the heart in order to compensate for the decreased effective cardiac output that results.
AR occurs from either damage to the aortic valve leaflets or dilation of the aortic annulus.

As the disease progresses, symptoms of congestive heart failure eventually occur. The ultimate treatment is surgical aortic valve replacement, however medical therapy can improve symptoms.
The abnormal backflow of blood that occurs leads to pathologic changes that are highly dependant on how severe the aortic regurgitation is and how quickly it developed. Left ventricular chamber enlargement and hypertrophy take place to help maintain a normal cardiac output in *chronic AR*. In *acute AR* - cardiac pressures increase quickly in this setting and heart failure as well as cardiogenic shock may occur.
Etiology

Aortic leaflet/cusp abnormalities

- **Infectious**: Bacterial endocarditis, rheumatic fever
- **Congenital**: Bicuspid aortic valve calcification
- **Inflammatory**: Systemic Lupus Erythematosis (SLE), Rheumatoid Arthritis (RA), Behcet's syndrome
- **Degenerative**: Myxomatous (floppy valve), senile calcification
- **Others**: Trauma, post aortic valve valvuloplasty, diet drug valvopathy, carcinoid valve disease (requires lung mets or PFO)

Aortic root abnormalities

- **Aortic root dilation**: Marfan's syndrome, syphilitic aortitis, idiopathic aortitis, Ehlers-Danlos syndrome, relapsing polychondritis
- **Loss of commissural support**: Aortic dissection, trauma, supracristal ventricular septal defect (VSD)

Increased afterload

- Uncontrolled systemic hypertension
- **Supravalvular aortic stenosis** (can occur in William's syndrome)
- Coarctation of the aorta
Symptoms

*chronic* AR develops slowly over time, the left ventricle can easily compensate, thus no symptoms occur for a long period of time early in disease. The symptoms of aortic valve regurgitation once they occur are mostly from congestive heart failure.

Left heart failure causes dyspnea from high pressure in the left ventricle transmitting into the pulmonary vasculature.
Right heart failure symptoms include lower extremity dependant edema. When the legs are elevated at night, the fluid redistributes centrally causing pulmonary edema resulting in orthopnea (dyspnea while laying flat) or paroxysmal nocturnal dyspnea (PND). Hepatic congestion can occur causing right upper quadrant abdominal pain.
Physical Examination

auscultation, the typical murmur of aortic regurgitation is a soft, high-pitched, early diastolic decrescendo murmur heard best at the 3rd intercostal space on the left (Erb's point) on end expiration with the patient sitting up and leaning forward.

This murmur is often difficult to distinguish from the Graham-Steele murmur of pulmonic insufficiency
In addition to the above murmur, a systolic ejection murmur may be present at the right upper sternal border simply due to the large stroke volume passing through the aortic valve. An early diastolic rumble may also be heard at the apex due to the regurgitant jet striking the anterior leaflet of the mitral valve causing it to vibrate. This murmur is termed the "Austin-Flint murmur".
In *chronic aortic regurgitation*, visible cardiac and arterial pulsations are common due to the large stroke volume and cause interesting peripheral signs discussed below. The carotid pulse can commonly be seen. The PMI (point of maximal intensity/impulse) is displaced laterally and caudally due to the LV dilation and hypertrophy that occurs.

*Acute aortic regurgitation* will cause a very short, early diastolic decrescendo murmur since the aortic and left ventricular pressure equalized quickly since the left ventricle has not had time to dilate or hypertrophy.
Peripheral Signs

Corrigan's pulse: A rapid and forceful distension of the arterial pulse with a quick collapse.

De Musset's sign: Bobbing of the head with each heartbeat (like a bird walking).

Muller's sign: Visible pulsations of the uvula.

Quincke's sign: Capillary pulsations seen on light compression of the nail bed.
**Diagnosis**

non-specific:

**ECG** - may show left ventricular hypertrophy and left atrial enlargement.

- in acute aortic regurgitation, sinus tachycardia due to the increased sympathetic nervous tone

**Chest radiograph**

- cardiomegaly is present in patients with chronic aortic regurgitation. In acute aortic regurgitation, pulmonary edema is almost universally present. If the aortic regurgitation is due to an aortic dissection, the mediastinum may appear widened.
Echocardiography:

- identify the cause and determine severity
- is almost 100% sensitive and specific for the detection of aortic regurgitation.
- the actual regurgitant jet can be directly visualized
- structural abnormalities such as a bicuspid aortic valve can be seen. Vegetations on the aortic valve may be identified indicating endocarditis. The size of the aortic root can be measured and aortic dissections can be identified.
Treatment

In patients with mild to moderate aortic regurgitation, no specific treatment is required. While reducing afterload with medications can lessen aortic regurgitation and improve symptoms, there is no evidence that this approach delays the need for surgical aortic valve replacement.

The most commonly used medications for this purpose include ACE inhibitors such as lisinopril or ramipril and dihydropyridine calcium channel blockers such as amlodipine and nifedipine.

Surgical replacement (not repair) of the aortic valve is the definitive therapy.
Thank you for your attention