

Aortic valve disease

AORTIC STENOSIS

Aetiology

- The three common causes of aortic stenosis (AS) are congenital, rheumatic and degenerative.
- It is seen in patients who are 35 years or older and results from calcification of a congenital or rheumatic valve or of a normal valve that has undergone 'degenerative' changes.
- Rare causes of AS include obstructive infective vegetations, homozygous type II hyperlipoproteinaemia, Paget's disease of bone, rheumatoid disease, ochronosis, and irradiation.



16.84 Causes of aortic stenosis

Infants, children, adolescents

- Congenital aortic stenosis
- Congenital subvalvular aortic stenosis
- Congenital supra-ventricular aortic stenosis

Young adults to middle-aged

- Calcification and fibrosis of congenitally bicuspid aortic valve
- Rheumatic aortic stenosis

Middle-aged to elderly

- Senile degenerative aortic stenosis
- Calcification of bicuspid valve
- Rheumatic aortic stenosis

Pathophysiology

- AS can be mild, moderate or severe depending on the constriction and calcification of aortic valve.
- This causes pressure overload on the left ventricle leading to concentric LV hypertrophy, keeping the cardiac output in the normal range

Table 1: Severity of Aortic Stenosis According to Valve Area and Consequent Gradient Across Valve and Echo Doppler Jet Velocity

	Valve Area (cm ²)	Gradient (mm Hg)	Doppler Jet Velocity (m/sec)
Mild	>1.5	<25	<3
Moderate	1 to 1.5	25 to 40	3 to 4
Severe	<1.0	>40	>4

- As the severity of AS increases, cardiac output remains within the normal range at rest but fails to increase in proportion to the amount of exercise performed or does not increase at all.
- Eventually left ventricular failure occurs giving rise to pulmonary hypertension and right-sided heart failure

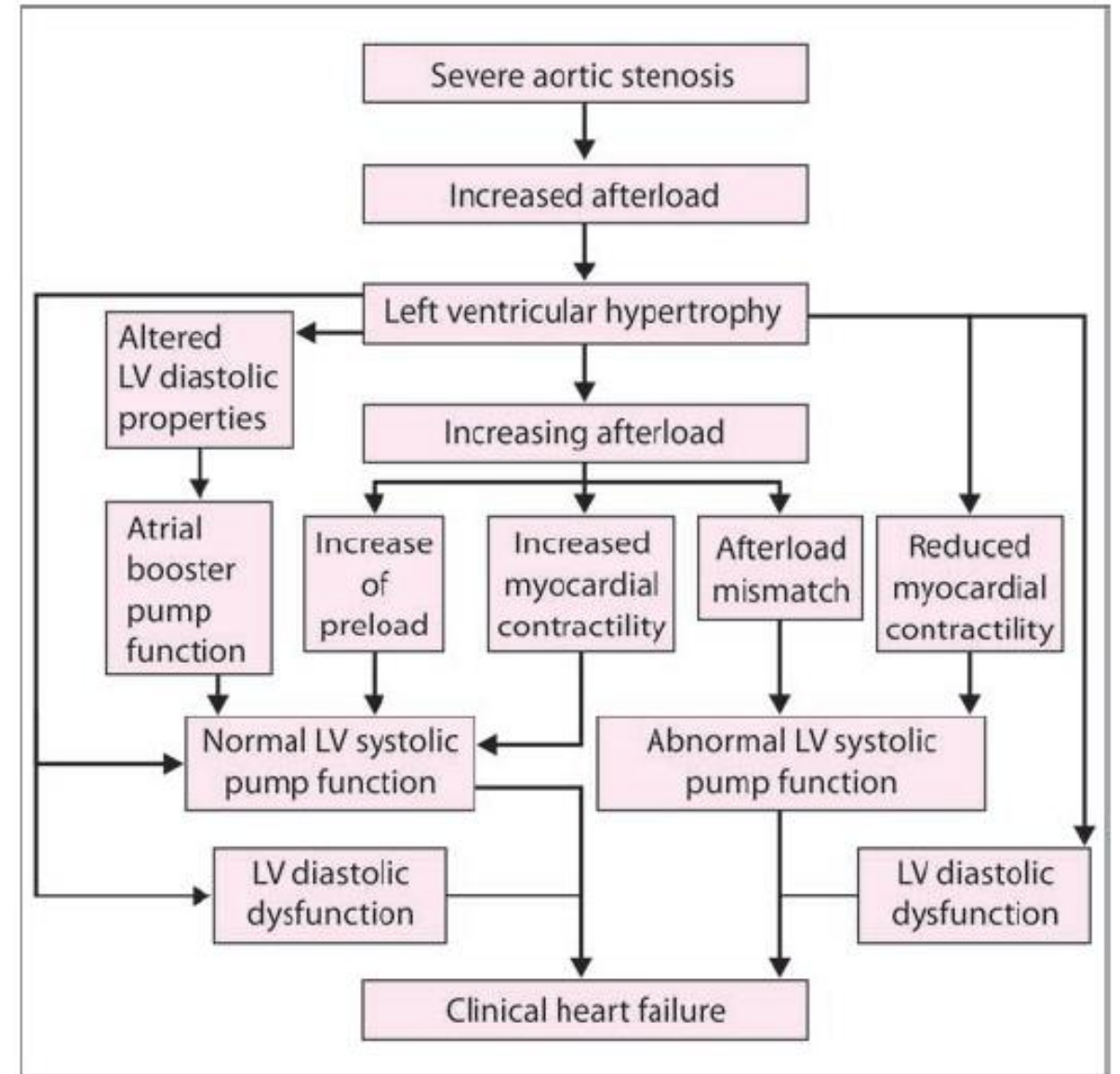


Figure 1: Pathophysiology of aortic stenosis.

Clinical Features

History

- Most patients with valvular AS are asymptomatic.
- The symptoms of AS are angina pectoris, syncope, exertional presyncope, dyspnoea (on exertion, orthopnoea, paroxysmal nocturnal dyspnoea, pulmonary oedema), and the symptoms of heart failure.
- Sudden cardiac death is stated to occur in 5% of patients with AS. It occurs only in those with severe valve stenosis.
- Typical angina pectoris occurs with or without associated CAD.

- Exertional syncope occurring on effort is caused by either systemic vasodilatation in the presence of a fixed or inadequate cardiac output, an arrhythmia, or both.
- Dyspnoea on exertion, orthopnoea, paroxysmal nocturnal dyspnoea, and pulmonary oedema result from varying degrees of pulmonary venous hypertension.
- Systemic venous congestion is rather a late phenomenon with enlargement of the liver and peripheral oedema results from increased systemic venous pressure and salt and water retention.

- There is an increased incidence of gastrointestinal arteriovenous malformations, also known as Heyde syndrome.
- As a result, these patients are susceptible to gastrointestinal haemorrhage and anaemia.
- Bleeding can be caused by an acquired defect in the structure of von Willebrand factor. Calcific systemic embolism may occur.

Physical examination

- The arterial pulse is slow rising and late peaking (**parvus et tardus**).
- A systolic thrill may be felt in the carotid arteries.
- The jugular venous pulse and the heart size are normal in absence of heart failure.
- The cardiac impulse is heaving and sustained in character with the presence of palpable S4.
- The S2 may be single since A2 and P2 are superimposed or A2 is absent or soft.
- It may be paradoxically split because of late A2.
- A systolic ejection sound is often present in young patients with valvular AS and patients with bicuspid AV.

- The turbulent flow across stenotic aortic valve causes a mid systolic ejection murmur that peaks late in systole.
- In older patients, the systolic ejection murmur may be heard only at the apex of the heart (**Gallavardin phenomenon**).
- The intensity of the systolic murmur varies from beat to beat. This characteristic is helpful in differentiating AS from MR, in which the murmur is usually unaffected.
- The murmur of valvular AS is augmented by squatting, which increases stroke volume. It is reduced in intensity during the strain of the Valsalva manoeuvre and when standing.

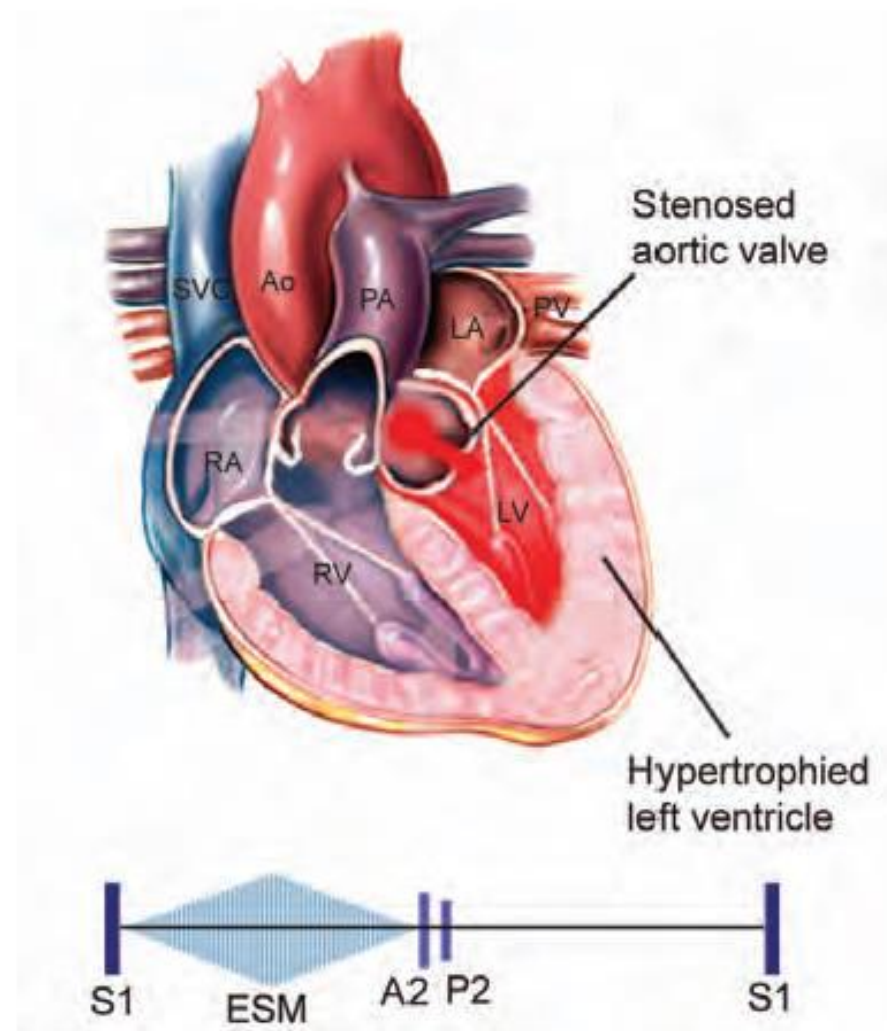


Fig. 3.126: Aortic stenosis

Table 2: Physical Findings with Varying Severity of Aortic Valve Stenosis

	Mild AS	Moderate AS	Severe AS + Normal LV Function	Severe AS + LV Dysfunction	Severe AS + Heart Failure ^a
Arterial pulse	Normal	Slowly rising	<i>Parvus et tardus</i>	<i>Parvus et tardus</i>	Small volume
Jugular venous pulse	Normal	Normal	Normal	Normal	±
Carotid thrill	±	±	±	±	±
Cardiac impulse	Normal	Heaving	Heaving, sustained palpable 'a' wave	Heaving	Heaving or reduced
Precordial thrill	±	±	Usually ++	±	–
Auscultation					
S4	–	±	+	+	–
S3	–	–	–	±	+
ESS	+	±	–	–	–
Peak of ESM	Early systole	Mid systole	Late systole	Late to midsystole, soft	Mid systole, soft or absent
S2	Normal	Normal or single	Single or paradoxical	Single	Single



16.85 Clinical features of aortic stenosis

Symptoms

- Mild or moderate stenosis: usually asymptomatic
- Exertional dyspnoea
- Angina
- Exertional syncope
- Sudden death
- Episodes of acute pulmonary oedema

Signs

- Ejection systolic murmur
- Slow-rising carotid pulse
- Thrusting apex beat (left ventricular pressure overload)
- Narrow pulse pressure
- Signs of pulmonary venous congestion

Investigations

Table 3: ECG Findings

Left ventricular hypertrophy with or without secondary ST-T wave changes. (Pressure overload pattern – ST depression and T wave inversion in lateral leads)

LA enlargement/hypertrophy

Sometimes shows conduction delay, left bundle branch block.

Heart blocks (calcification) can occur rarely

Atrial fibrillation indicates associated mitral disease or heart failure

Table 4: Radiological Abnormalities

Normal sized heart

Dilated proximal ascending aorta (post-stenotic dilation)

Calcification of aortic valve (better seen on fluoroscopy). Calcification does not mean severe AS

Late stages heart size large with pulmonary venous hypertension

Table 5: Echocardiography

Valve thickening and doming on 2D and gradient across valve on CW Doppler

All severe AS in adults will be calcified

Allows definition of valve anatomy, cause of AS, calcification and assessment of severity of AS

Left ventricular hypertrophy, systolic functions (ejection fraction) can be evaluated

Assessment of aortic root dilatation and associated mitral valve disease

Cardiac catheterisation and angiography

- The main indication for cardiac catheterisation and angiography is to look for associated CAD in these patients before surgery.

Natural History

- The natural history of AS in the adult consists of a prolonged latent period.
- Once moderate stenosis is present (jet velocity >3 m/s), the average rate of progression is an increase in jet velocity of 0.3 m/s per year, an increase in mean pressure gradient of 7 mm Hg per year, and a decrease in valve area of 0.1 cm² per year.
- Eventually, symptoms of angina, syncope, or heart failure develop and the outlook changes dramatically.
- After the onset of symptoms, average survival is 2 to 3 years, with a high-risk of sudden death.

Management

Medical treatment

- Patients with severe AS should be cautioned to avoid vigorous athletic and physical activity.
- Exercise stress testing should be absolutely avoided in symptomatic patients.
- Symptomatic patients with severe AS are usually operative candidates because medical therapy has little to offer.
- However, medical therapy may be necessary in patients who are considered to be inoperable (usually because of comorbid conditions that preclude surgery).

- Diuretics must be used with caution because hypovolaemia may reduce the elevated LV end-diastolic pressure, lower cardiac output, and produce orthostatic hypotension.
- ACE inhibitors should be used with caution but are beneficial in treating patients with symptomatic LV systolic dysfunction who are not candidates for surgery.
- Beta-adrenergic blockers can depress myocardial function and induce LV failure and should be avoided in patients with AS. AF should be treated promptly

Table 6: Medical Treatment of Patients with Aortic Valve Stenosis

Antibiotic prophylaxis

- Infective endocarditis
- Recurrent rheumatic carditis

Restriction of activities

- Severe exercise
- Competitive sports

Arrhythmias

- Prevent and/or control
- Restore sinus rhythm, if possible

Cardiac medications (only if essential)

- Avoid negative inotropic and proarrhythmic agents if possible
- Diuretics – use cautiously
- Arteriolar and venodilators – use cautiously

Follow-up of asymptomatic patients

- Mild aortic stenosis: every 2 to 5 years
- Moderate aortic stenosis: every 6 to 12 months
- Develop symptoms: immediate

Surgical treatment

- *Children:* In the adolescent or young adult with severe congenital AS, balloon aortic valvotomy is recommended in all symptomatic patients and in asymptomatic patients with a transvalvular gradient greater than 60 mm Hg or in those with ECG showing ST changes at rest or with exercise.
- The same indications are appropriate for surgical intervention, although balloon valvotomy is probably preferable at experienced centres.

- *Adults:* Aortic valve replacement (AVR) is the treatment of choice for patients of severe AS.
- AVR is indicated for symptomatic patients with severe AS, patients with severe AS undergoing coronary artery bypass graft surgery (CABG), patients with severe AS undergoing surgery on the aorta or other heart valves, and patients with severe AS and LV systolic dysfunction (ejection fraction less than 0.50).



16.87 Aortic stenosis in old age

- **Incidence:** the most common form of valve disease affecting the very old.
- **Symptoms:** a common cause of syncope, angina and heart failure in the very old.
- **Signs:** because of increasing stiffening in the central arteries, low pulse pressure and a slow rising pulse may not be present.
- **Trans-catheter aortic valve implantation (TAVI):** a good option in older individuals because less invasive than surgery.
- **Surgery:** can be successful in those aged 80 years or more in the absence of comorbidity, but with a higher operative mortality. The prognosis without surgery is poor once symptoms have developed.
- **Valve replacement type:** a biological valve is often preferable to a mechanical one because this obviates the need for anticoagulation, and the durability of biological valves usually exceeds the patient's anticipated life expectancy.

AORTIC REGURGITATION

Aetiology

- Aortic regurgitation (AR) can result from disease of the aortic valve (aortic cusp) and due to dilatation of aortic root.
- Physical findings can help differentiate between these although echocardiography is required for this.
- Generally, AR is chronic in most conditions. However, acute AR can be seen in infective endocarditis, aortic dissection and during acute rheumatic fever

Table 7: Aetiology of Chronic Aortic Valve Regurgitation

Aortic root dilatation

Hypertension with aortic root dilatation, atherosclerosis

Various forms of aortitis and arteritis, e.g. giant cell arteritis and Takayasu disease, syphilis

Connective tissue disease, e.g. Marfan syndrome, osteogenesis imperfecta, and Ehlers-Danlos syndrome

Autoimmune diseases, e.g. ankylosing spondylitis, rheumatoid arthritis, and systemic lupus erythematosus

Congenital lesions, e.g. supraaortic or discrete subaortic aortic stenosis, ventricular septal defect, and aneurysm of the sinus of Valsalva

Aortic valve diseases

Congenital bicuspid valve

Rheumatic heart disease

Previous infective endocarditis

Pathophysiology

- Acute AR results in severe volume overload of left ventricle with resultant pulmonary congestion as LV chamber is small and LV and LA pressures are increased markedly.
- There is a fall in cardiac output due to decrease in stroke volume.
- In contrast, in cases of chronic aortic regurgitation eccentric cardiac hypertrophy with the increased preload allows a large increase in LV end-diastolic volume.

- The large total stroke volume allows forward stroke volume to be normal despite a large regurgitant volume.
- The large total stroke volume along with the wide pulse pressure produces most of the physical signs of aortic regurgitation.
- The combined pressure and volume overload stimulates the development of both concentric and eccentric hypertrophy. After many years, LV dysfunction eventually develops, with further LV dilation, elevation of LV filling pressure, decreased EF, and reduced total and forward stroke volume.

Natural History

- The rate of progression to symptoms and/or LV systolic dysfunction is about 4.3% per year in asymptomatic patients of severe AR.
- Sudden death occurs at the rate of less than 0.2% per year in this subgroup of patients.
- The average rate of symptom onset in patients with depressed LV function is greater than 25% per year.
- Mortality rates of greater than 10% per year have been reported in patients with angina pectoris and greater than 20% per year in those with heart failure.

Clinical Features

History

- Patients with aortic regurgitation in the chronic compensated phase are often entirely asymptomatic.
- As LV decompensation proceeds, symptoms of dyspnoea, palpitations, orthopnoea, and in advanced cases, paroxysmal nocturnal dyspnoea and peripheral oedema occur.
- Nocturnal angina occurs due to bradycardia and increased AR with reduced aortic diastolic pressure leading to diminished coronary perfusion.

Physical examination

- In patients with chronic, severe AR, the head frequently bobs with each heartbeat (**de Musset sign**), and the pulses are of the 'water-hammer' or collapsing type with abrupt distention and quick collapse (**Corrigan pulse**).
- A bisferiens pulse may be present and is more readily recognised in the brachial and femoral arteries than in the carotid arteries.
- **Traube sign** (also known as 'pistol shot sounds') refers to booming systolic and diastolic sounds heard over the femoral artery

- **Müller sign** consists of systolic pulsations of the uvula; and **Duroziez sign** consists of a systolic murmur heard over the femoral artery when it is compressed proximally and a diastolic murmur when it is compressed distally.
- Capillary pulsations (i.e. **Quincke sign**) can be detected by pressing a glass slide on the patient's lip, by transmitting a light through the patient's fingertips, or by exerting gentle pressure on the tip of a fingernail.
- Systolic arterial pressure is elevated, and diastolic pressure is abnormally low.
- **Hill sign** refers to popliteal cuff systolic pressure exceeding brachial cuff pressure by more than 60 mm Hg.

- Korotkoff sounds often persist to zero even though intraarterial pressure rarely falls below 30 mm Hg. The point of change in Korotkoff sounds (i.e. the muffling of these sounds in phase IV) correlates with the diastolic pressure.
- The apical impulse is diffuse and hyperdynamic and is displaced laterally and inferiorly.
- The augmented stroke volume may create a systolic thrill at the base of the heart or suprasternal notch, and over the carotid arteries.
- The PR interval may be prolonged, causing a soft S1. A2 may be normal or accentuated when AR is due to disease of the aortic root but is soft or absent when the valve is causing AR.
- P2 may be obscured by the early diastolic murmur. Thus, S2 may be absent or single or exhibit narrow or paradoxical splitting.

- A systolic ejection sound, presumably related to abrupt distention of the aorta by the augmented stroke volume, is frequently audible. S3 gallop correlates with an increased LV end-diastolic volume.
 - Its development may be a sign of impaired LV function, which is useful in identifying patients with severe AR who are candidates for surgical treatment.
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- The aortic regurgitant murmur, the principal physical finding of AR, is one of high frequency that begins immediately after A2.
 - The murmur is heard best with the diaphragm of the stethoscope while the patient is sitting up and leaning forward, with the breath held in deep exhalation. The severity of AR correlates better with the duration than with the intensity of the murmur. When the murmur is musical ('cooing dove' murmur), it usually signifies eversion or perforation of an aortic cusp.

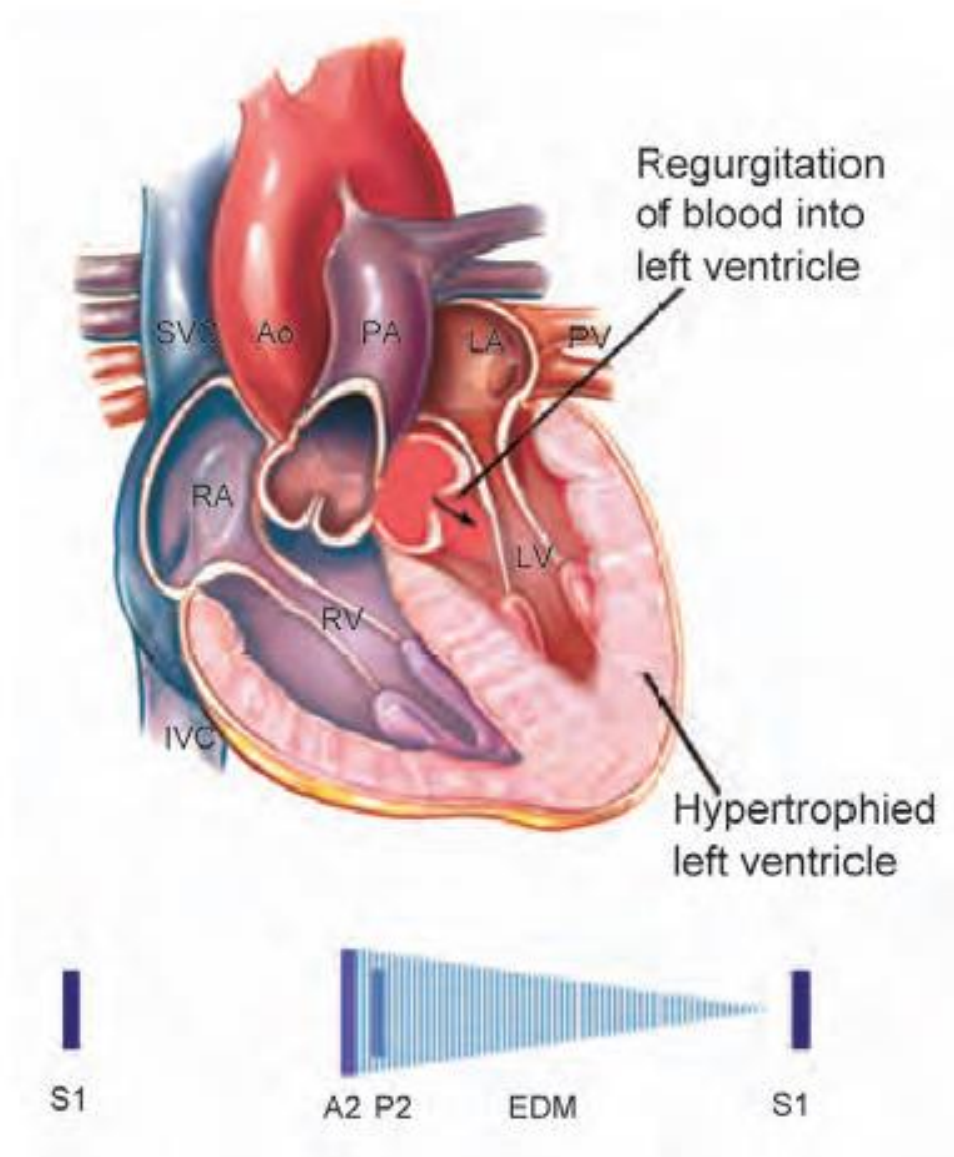


Fig. 3.127: Aortic regurgitation

- When regurgitation is caused by primary valvular disease, the diastolic murmur is heard best along the left sternal border in the 3rd and 4th intercostal spaces.
- However, when it is caused mainly by dilation of the ascending aorta, the murmur is often more readily audible along the right sternal border.
- Many patients with chronic AR have a harsh systolic outflow murmur caused by the increased total LV stroke volume and ejection rate.
- A mid-diastolic and late diastolic apical rumble, **the Austin Flint murmur**, is common in severe AR and may occur in the presence of a normal mitral valve

- The Austin Flint murmur may be difficult to differentiate from that caused by mitral stenosis (MS), but the presence of an opening snap and a loud S1 in MS and the absence of these findings in AR are helpful clues.
- As the LV end-diastolic pressure rises, the Austin Flint murmur commences and terminates earlier.
- The Austin Flint murmur, like the murmur of AR, is augmented by isometric exercise and administration of vasopressors and is reduced by amyl nitrite inhalation.

Signs of Wide Pulse Pressure

- 1. ***Light house sign*** (alternate flushing and blanching of forehead)
- 2. ***Landolfi's sign*** (Change in pupillary size in accordance with cardiac cycle and not related to light).
- 3. Retinal artery pulsations (***Becker's sign***).
- 4. ***de Musset's sign*** (head bobbing with each heart beat). It is due to ballistic effect of severe AR.
- 5. ***Muller's sign***—Systolic pulsations of uvula.

- 6. **Quincke's sign** (capillary pulsations) can be detected by pressing a glass slide on patients lip or nail bed.
- 7. Dancing carotids (**Corrigan's sign**).
- 8. Locomotor brachii.
- 9. Collapsing or water-hammer pulse (↑ systolic pressure and ↓ diastolic pressure); usually pulse pressure is more than the diastolic pressure.
- 10. Bisferiens pulse.
- 11. Pistol shot femorals (**Traube's sign**) arteries ; Pistol shot sound over the femoral vein is heard in severe TR.

- 12. ***Duroziez's sign***—systolic murmur heard over femoral artery when it is compressed proximally and a diastolic murmur when it is compressed distally using the 'bell' of the stethoscope.
- *Duroziez's murmur*—diastolic murmur heard with the diaphragm of the stethoscope when distal pressure is applied.
- 13. ***Hill's sign***—popliteal cuff systolic pressure exceeds brachial cuff pressure by > 20 mm Hg.
- Mild AR 20–40 mm Hg
- Moderate AR 40–60 mm Hg
- Severe AR > 60 mm Hg.
- 14. ***Rosenbach's sign***—pulsations of liver.
- 15. ***Gerhardt's sign***—pulsations over enlarged spleen.

Other Signs

- Apical impulse is displaced downwards and outwards and hyperdynamic in nature.
- Soft S1 (only in acute AR)
- S3 may be heard
- A high frequency decrescendo early diastolic murmur immediately after A2, best heard in left 3rd or 4th spaces with the diaphragm of the stethoscope with the patient leaning forwards, in expiration.
- Flow ESM across aortic valve heard at heart base, conducted to carotids.
- Flow MDM across mitral valve (*Austin-Flint murmur*).
- In MS, there is loud S1 and there is opening snap.
- AR murmur in aortic area – Aortic root dilatation
- AR murmur in 2nd aortic area – Valvular lesion
- AR murmur is increased by hand grip

Severity

The presence of the following indicate severe AR:

- a. Duration of murmur ($> 2/3$ of diastole) is directly proportional to the severity. In moderate to severe AR, murmur becomes holo diastolic and may have a rough quality.
- b. Bisferiens pulse
- c. Hill's sign > 60 mm Hg.
- d. Apical impulse (down and out)
- e. Austin-Flint murmur
- f. Marked peripheral signs.-They are absent in depressed myocardial function.

Table 8: Physical Findings with Varying Severity of Chronic Aortic Valve Regurgitation

	Mild AR	Moderate AR	Severe AR	Severe AR + Left Ventricular Systolic Dysfunction	Severe AR + Heart Failure + Left Ventricular Systolic Dysfunction
Arterial pulse	Normal	Corrigan + to ++	Corrigan +++	Corrigan ++	Corrigan +
Arterial pressure					
Systolic	Normal	Increased + to ++	Increased +++	Increased ++	Normal +
Diastolic	Normal	Decreased + to ++	Decreased +++ to ++++	Decreased ++ to +++	Decreased +
Pulse pressure	Often normal	Increased + to ++	Increased +++ to ++++	Increased ++ to +++	Increased +
Cardiac impulse	Often normal	Hyperdynamic	Very hyperdynamic visible $\pm$ chest may rock	Hyperdynamic	May be hypodynamic
Precordial thrill					
Systolic	–	$\pm$	$\pm$	$\pm$	–
Diastolic	–	–	$\pm$	$\pm$	–
Auscultation					
S4	–	–	–	–	–
S1	Normal	Often soft	Soft	Soft	Soft
S2	Normal	Normal or single	Often single	Often single	Often single
S3	–	+	++ to +++	+++	+++
ESM	$\pm$	+	+ to ++	+ to ++	+
AoDM	+	++	+++ to ++++	++ to +++	+ to ++
Austin Flint murmur	–	–	$\pm$	–	–

Investigation

Table 9: ECG Findings

Left axis deviation with left ventricular hypertrophy of volume overload, pattern (prominent Q waves in leads I, aVL, and V3 through V6)

Tall and upright T-waves. Later on inverted T-wave, with ST-segment depressions

Intraventricular conduction defects occur late in the course and are usually associated with LV dysfunction

Table 10: Radiological Abnormalities

Marked enlargement of cardiac shadow

Calcification of the aortic valve uncommon

Dilation of the ascending aorta (more than in AS)

Severe aneurysmal dilation of the aorta suggests aortic root disease (Marfan syndrome, cystic medial necrosis, or annuloaortic ectasia)

Linear calcifications in the wall of the ascending aorta – syphilitic aortitis

Acute AR: less cardiac dilatation and more pulmonary venous hypertension

Table 11: Echocardiography

Transthoracic echocardiography: measurement of LV end-diastolic and end-systolic dimensions and volumes, ejection fraction, and mass. Serial measurements for follow-up

High-frequency fluttering of the anterior leaflet of the mitral valve during diastole

Doppler echocardiography and colour flow Doppler imaging are the most sensitive and accurate non-invasive techniques in the diagnosis and evaluation of AR

AR can be quantified into mild, moderate, and severe AR on above basis

LV size and ejection fraction are normal

Cardiac catheterisation and angiography

- Cardiac catheterisation is undertaken when the severity of AR is not clear on non-invasive evaluation. The indications for selective coronary angiography are the same as for aortic stenosis.

Management

Medical treatment

- Majority of patients with AR are not candidates for prophylaxis.
- Patients with mild or moderate AR should be followed clinically and by echocardiography.
- Asymptomatic patients with chronic, severe AR and normal LV function should be examined at intervals of approximately 6 months.
- Patients with AR who have limitations of cardiac reserve and/or evidence of declining LV function should avoid heavy exertion.

- Systemic arterial diastolic hypertension should be treated because it increases the regurgitant flow; vasodilating agents such as nifedipine or ACE-inhibitors are preferred and betablocking agents should be used with great caution.
- AF and bradyarrhythmias are poorly tolerated and should be prevented, if possible.

Vasodilator therapy

- There is considerable uncertainty regarding vasodilator therapy.
- Short-term studies spanning 6 months to 2 years have demonstrated beneficial haemodynamic effects of oral hydralazine, nifedipine, felodipine, and ACE-inhibitors.
- But, in view of conflicting data, definitive recommendations regarding the indications for long-acting nifedipine or ACE-inhibitors are not possible.

Surgery: Indications

- AVR is the treatment of choice in symptomatic patients.
- Chronic medical therapy may be necessary in some patients who refuse surgery or are considered to be inoperable because of co-morbid conditions.
- These patients should receive an aggressive heart failure regimen with ACE-inhibitors (and perhaps nitrates), digoxin, diuretics, and salt restriction, but beta-blockers should be avoided.
- In patients who are candidates for surgery but who have severely decompensated LV function, vasodilator therapy may be particularly helpful in stabilising patients while preparing for operation.
- Such patients also respond, at least temporarily, to treatment with digitalis glycosides, salt restriction, and diuretics.

AVR is indicated for following patients with AR:

- 1. Symptomatic patients with severe AR.
- 2. Asymptomatic patients with chronic severe AR and LV systolic dysfunction (ejection fraction 0.50 or less) at rest.
- 3. Patients with chronic severe AR while undergoing CABG or surgery on the aorta or other heart valves.
- 4. Asymptomatic patients with severe AR with normal LV systolic function (ejection fraction greater than 0.50) but with severe LV dilatation (end-diastolic dimension greater than 75 mm Hg or end-systolic dimension greater than 55 mm Hg).

- Thank you