Disease of the heart valves

Aortic valve Disease

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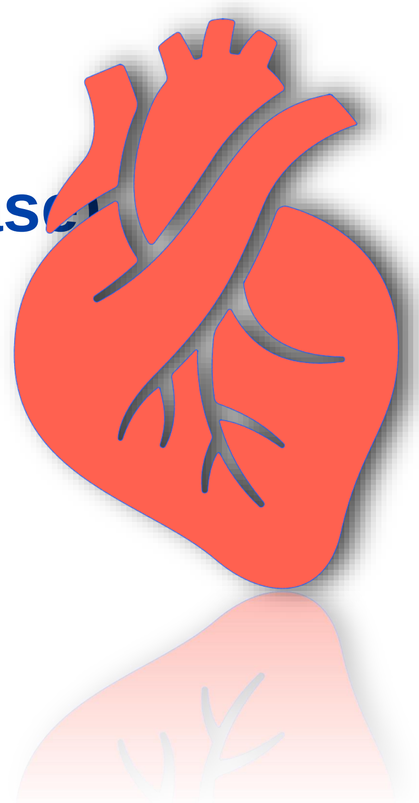


Aortic Stenosis

- In congenital aortic stenosis, obstruction is present from birth or becomes apparent during infancy. With bicuspid aortic valves, obstruction may take years to develop as the valve becomes fibrotic and calcified, and these patients present as young to middle-aged adults. Rheumatic disease of the aortic valve presents at a similar age but is usually accompanied by mitral valve disease. In older people, tricuspid aortic valves may become stenotic as the result of fibrosis and calcification. Stenosis develops slowly, typically occurring at 30–60 years in those with rheumatic disease, 50–60 years in those with bicuspid aortic valves and 70–90 years in those with calcific aortic disease.

16.84 Causes of aortic stenosis

Infants, children, adolescents

- Congenital aortic stenosis
- Congenital subvalvular aortic stenosis
- Congenital supravalvular aortic stenosis

Young to middle-aged adults

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

Middle-aged to older adults

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

- **Pathogenesis**

- Cardiac output is initially maintained in patients with aortic stenosis at the cost of a steadily increasing pressure gradient across the aortic valve. With progression of the stenosis, the LV becomes increasingly hypertrophied and coronary blood flow may be inadequate to supply the myocardium, such that angina can develop even in the absence of coexisting CAD. The fixed outflow obstruction limits the increase in cardiac output required on exercise. Eventually, the LV can no longer overcome the outflow tract obstruction and LV failure results, leading to pulmonary oedema.

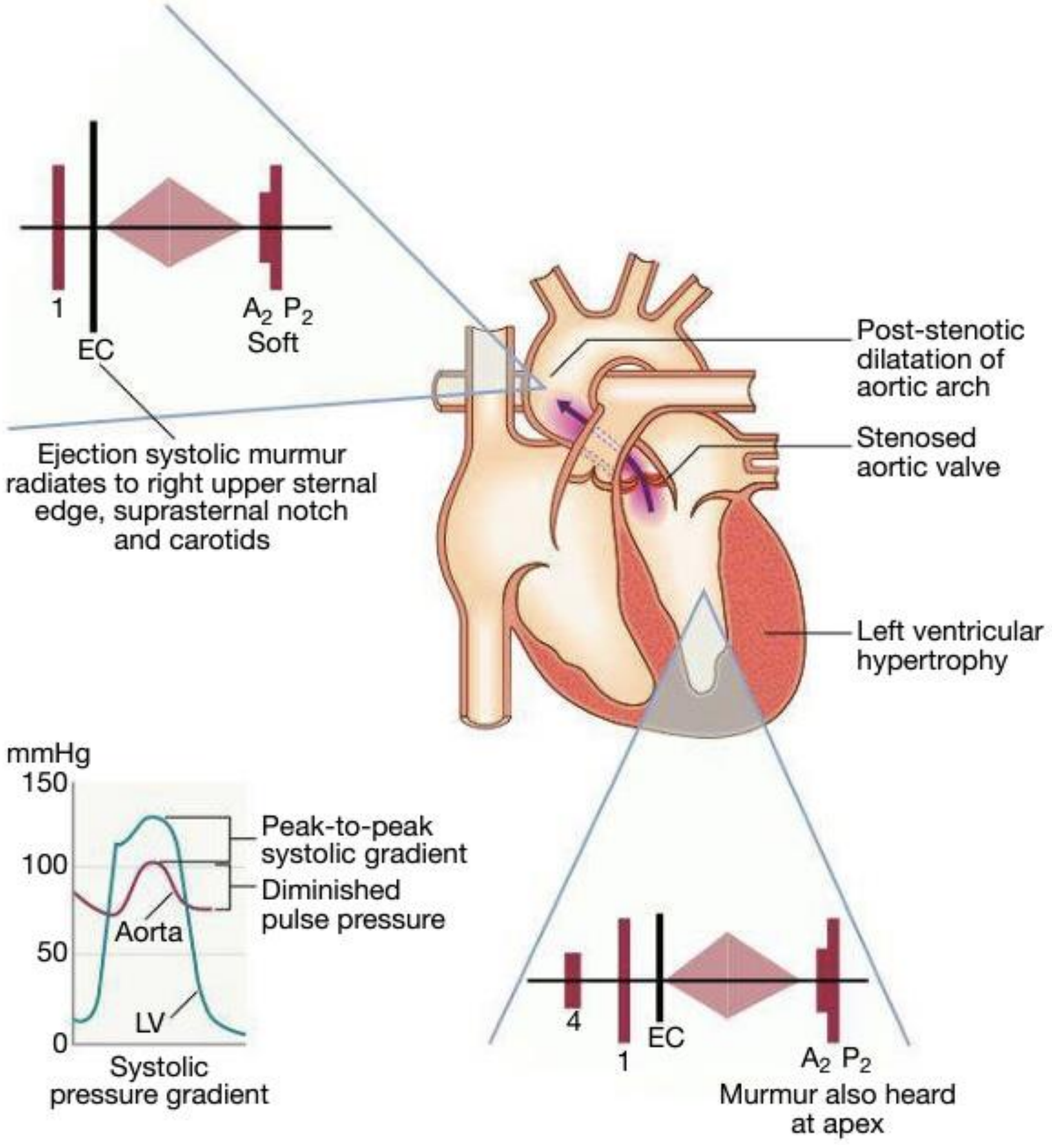


Fig. 16.82 Aortic stenosis. Pressure traces show the systolic gradient between left ventricle (LV) and aorta. The 'diamond-shaped' murmur is heard best with the diaphragm in the aortic outflow and also at the apex. An ejection click (EC) may be present in young patients with a bicuspid aortic valve but not in older patients with calcified valves. Aortic stenosis may lead to left ventricular hypertrophy with a fourth sound at the apex and post-stenotic dilatation of the aortic arch. Fig. 16.11 shows the typical Doppler signal with aortic stenosis.

- **Clinical features**

- Aortic stenosis is commonly picked up in asymptomatic patients at routine clinical examination but the **three cardinal symptoms are angina, breathlessness and syncope** . Angina arises either because of the increased demands of the hypertrophied LV working against the high-pressure outflow tract obstruction, or the presence of coexisting CAD, which affects over 50% of patients. Exertional breathlessness suggests cardiac decompensation as a consequence of the excessive pressure overload placed on the LV. Syncope usually occurs on exertion when cardiac output fails to rise to meet demand, leading to a fall in BP. Sometimes patients with severe aortic stenosis do not complain of symptoms. If, on clinical evaluation, this appears to be due to a sedentary lifestyle, a careful exercise test may reveal symptoms on modest exertion.

- A harsh ejection systolic murmur radiates to the neck, with a soft second heart sound, particularly in those with calcific valves. The murmur is often likened to a saw cutting wood and may (especially in older patients) have a musical quality like the ‘mew’ of a seagull . The severity of aortic stenosis may be difficult to gauge clinically, as older patients with a non-compliant ‘stiff’ arterial system may have an apparently normal carotid upstroke in the presence of severe aortic stenosis. Milder degrees of stenosis may be difficult to distinguish from aortic sclerosis, in which the valve is thickened or calcified but not obstructed. A careful examination should be made for other valve lesions, particularly in rheumatic heart disease, when there is frequently concomitant mitral valve disease. In contrast to patients with mitral stenosis, which tends to progress very slowly, patients with aortic stenosis typically remain asymptomatic for many years but deteriorate rapidly when symptoms develop; if otherwise untreated, they usually die within 3–5 years of presentation.

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
- Heaving apex beat (left ventricular pressure overload)
- Narrow pulse pressure
- Signs of pulmonary venous congestion

- **Investigations**

- Echocardiography is a pivotal investigation in patients suspected of having aortic stenosis. It can demonstrate restricted valve opening and Doppler assessment permits calculation of the systolic gradient across the aortic valve, from which the severity of stenosis can be assessed . In patients with impaired left ventricular function, velocities across the aortic valve may be diminished because of a reduced stroke volume; this is called low-flow aortic stenosis.

- When marked aortic regurgitation or elevated cardiac output is present, velocities are increased because of an increased stroke volume and this may overestimate stenosis severity on Doppler echocardiography.

- advanced cases, ECG features of LV hypertrophy are often pronounced , and down-sloping ST segments and T inversion (‘**strain pattern**’) are seen in the lateral leads, reflecting left ventricular fibrosis. Nevertheless, the ECG can be normal, despite severe stenosis. Occasionally, there is evidence of AV block due to the encroachment of the fibrocalcific process on the adjacent AV node and His–Purkinje system; an occasional cause of syncope in these patients. Imaging with CT may be useful in assessing the degree of valve calcification where there is uncertainty of disease severity.

16.86 Investigations in aortic stenosis

ECG

- Left ventricular hypertrophy
- Left bundle branch block

Chest X-ray

- May be normal; sometimes enlarged left ventricle and dilated ascending aorta on postero-anterior view, calcified valve on lateral view

Echo

- Calcified valve with restricted opening, hypertrophied left ventricle

Doppler

- Measurement of severity of stenosis
- Detection of associated aortic regurgitation

Cardiac catheterisation

- Mainly to identify associated coronary artery disease
- May be used to measure gradient between left ventricle and aorta

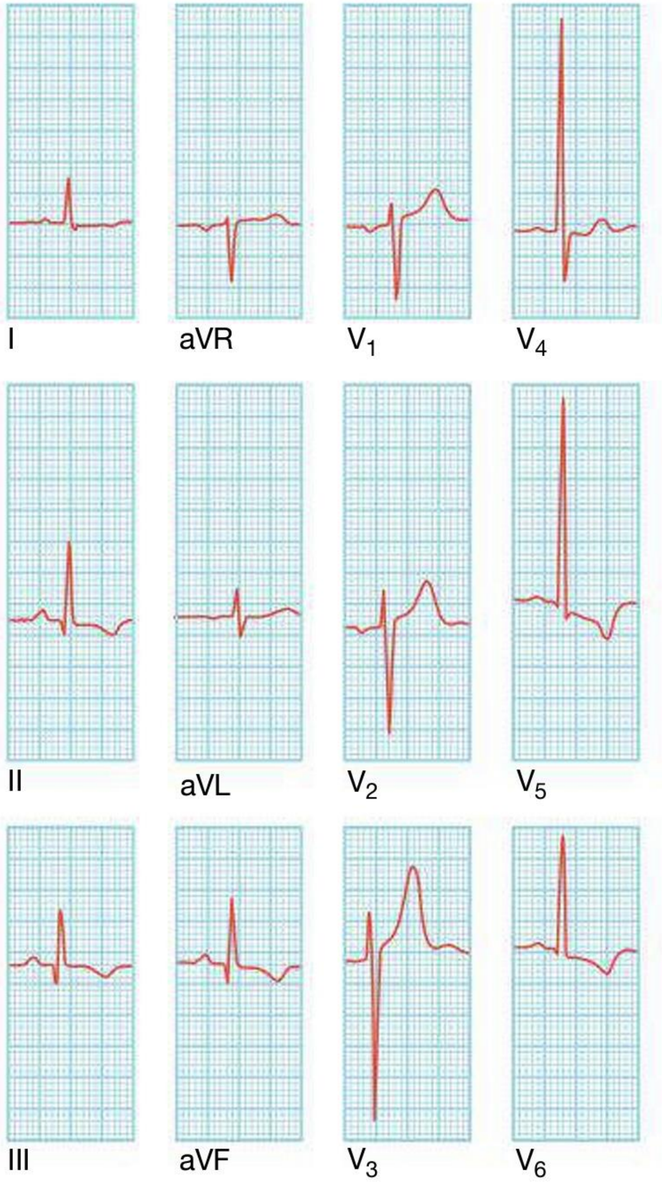
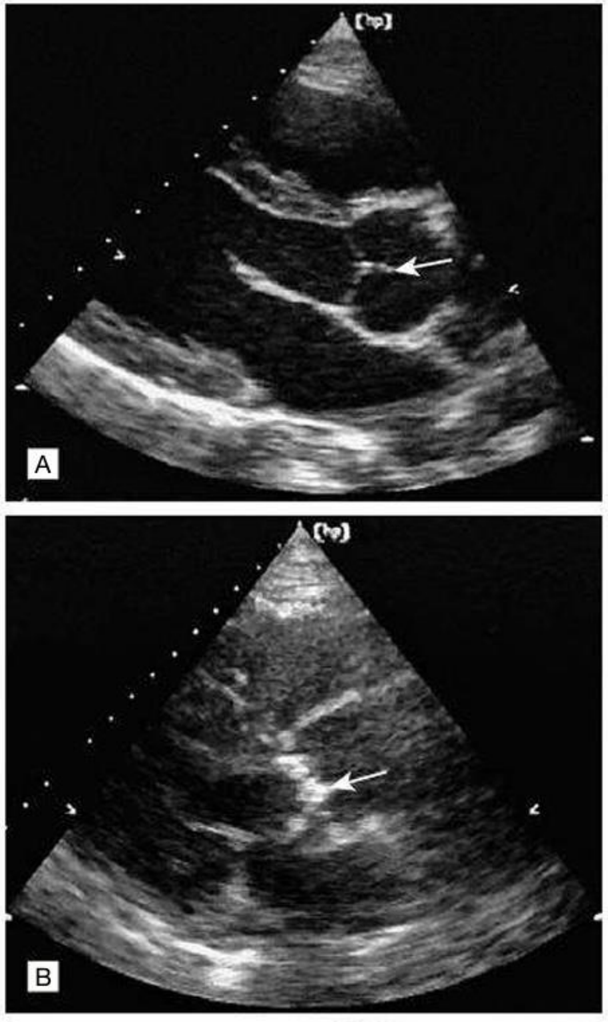


Fig. 16.84 Left ventricular hypertrophy. QRS complexes in limb leads have increased amplitude with very large R waves in V_4 and V_5 and a deep S wave in V_3. There is ST depression and T-wave inversion in leads II, III, aVF, V_5 and V_6: a 'left ventricular strain' pattern.



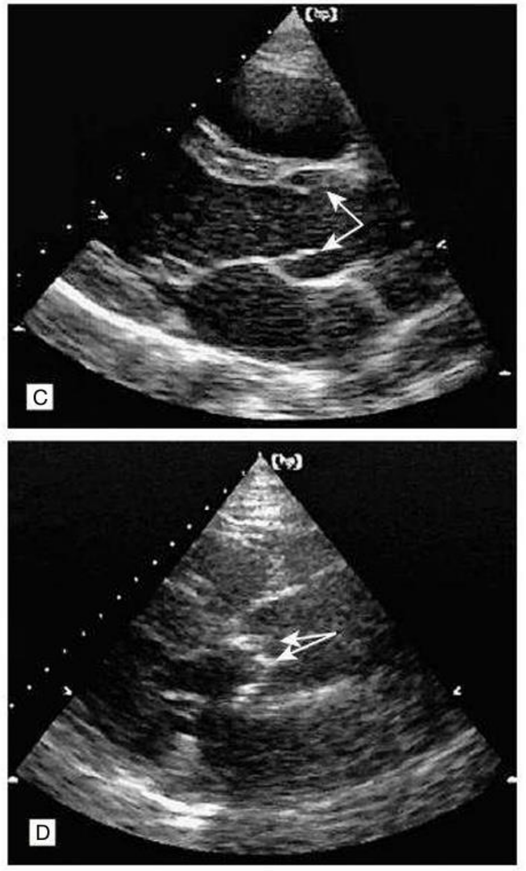


Fig. 16.83 Two-dimensional echocardiogram comparing a normal individual and a patient with calcific aortic stenosis. A Normal individual in diastole; the aortic leaflets are closed and thin, and a point of coaptation is seen (arrow). B Calcific aortic stenosis in diastole; the aortic leaflets are thick and calcified (arrow). C Normal in systole; the aortic leaflets are open (arrows). D Calcific aortic stenosis in systole; the thickened leaflets have barely moved (arrows). *From Newby D, Grubb N. Cardiology: an illustrated colour text. Edinburgh: Churchill Livingstone, Elsevier Ltd; 2005.*

- **Management**

- Irrespective of the severity of valve stenosis, patients with asymptomatic aortic stenosis have a good immediate prognosis and conservative management is appropriate. Such patients should be kept under review, as the development of angina, syncope, symptoms of low cardiac output or heart failure has a poor prognosis and is an indication for prompt surgery. In practice, patients with moderate or severe stenosis should be evaluated every 1–2 years with Doppler echocardiography to detect evidence of progression in severity. The intervals between reviews should be more frequent (typically 3–6-monthly) in older patients with heavily calcified valves.

- Patients with **symptomatic severe aortic stenosis** should have prompt aortic valve replacement. Delay exposes the patient to the risk of sudden death or irreversible deterioration in ventricular function. Old age is not a contraindication to valve replacement and results are very good in experienced centres, even for those in their eighties . This is especially the case with **transcatheter aortic valve implantation (TAVI)**.

- **Aortic balloon valvuloplasty** is useful in congenital aortic stenosis but has limited value in older patients with calcific aortic stenosis.

- **Anticoagulants** are required only in patients who have AF or those who have had a valve replacement with a mechanical prosthesis.

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.
- **Transcatheter 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

- **Pathogenesis**

- This condition can result from either disease of the aortic valve cusps, infection, trauma or dilatation of the aortic root.

- Regurgitation of blood through the aortic valve causes the LV to dilate as cardiac output increases to maintain the demands of the circulation. The stroke volume of the LV may eventually be doubled and the major arteries are then conspicuously pulsatile. As the disease progresses, left ventricular failure develops, leading to a rise in left ventricular end-diastolic pressure and pulmonary oedema.

16.88 Causes of aortic regurgitation

Congenital

- Bicuspid valve or disproportionate cusps

Acquired

- Rheumatic disease
- Infective endocarditis
- Trauma
- Causes of aortic dilatation:
  - Marfan syndrome
  - Aneurysm
  - Aortic dissection
  - Syphilis
  - Ankylosing spondylitis

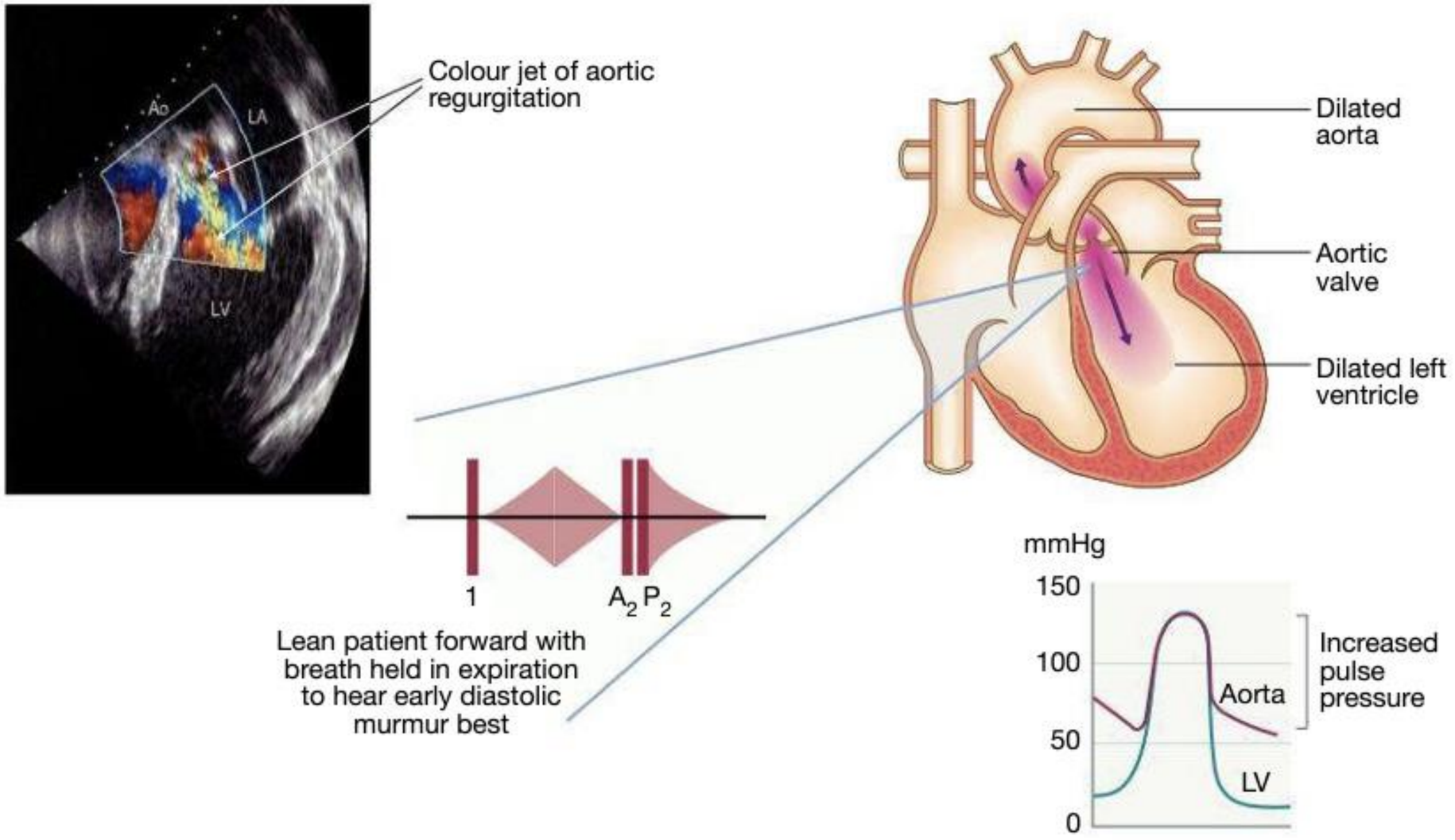


Fig. 16.85 **Aortic regurgitation.** The early diastolic murmur is best heard at the left sternal border and may be accompanied by an ejection systolic ('to-and-fro') murmur. The aortic arch and left ventricle (LV) may become dilated.The inset shows a Doppler echocardiogram with the regurgitant jet (arrows). *Inset (Colour Doppler echo) From Newby D, Grubb N. Cardiology: an illustrated colour text. Edinburgh: Churchill Livingstone, Elsevier Ltd; 2005.*

- **Clinical features**

- Until the onset of breathlessness, the only symptom may be an awareness of the heart beat particularly when lying on the left side, which results from the increased stroke volume. Paroxysmal nocturnal dyspnoea is sometimes the first symptom, and peripheral oedema or angina may occur. The characteristic murmur is best heard to the left of the sternum during held expiration ; a thrill is rare. A systolic murmur due to the increased stroke volume is common and does not necessarily indicate stenosis. The regurgitant jet causes fluttering of the mitral valve and, if severe, causes partial closure of the anterior mitral leaflet, leading to **functional mitral stenosis and a soft mid-diastolic (Austin Flint) murmur.**

- Acute severe regurgitation may occur as the result of perforation or tear of an aortic cusp in endocarditis. In this circumstance there may be no time for compensatory left ventricular hypertrophy and dilatation to develop and the features of heart failure may predominate. The classical signs of aortic regurgitation in such patients may be masked by tachycardia and an abrupt rise in left ventricular end-diastolic pressure. The pulse pressure may also be normal or near-normal and the diastolic murmur may be short or even absent.

16.89 Clinical features of aortic regurgitation

Symptoms

Mild to moderate aortic regurgitation

- Often asymptomatic
- Palpitations

Severe aortic regurgitation

- Breathlessness
- Angina

Signs

Pulses

- Large-volume or 'collapsing' pulse
- Low diastolic and increased pulse pressure
- Bounding peripheral pulses
- Capillary pulsation in nail beds: Quincke's sign
- Femoral bruit ('pistol shot'): Duroziez's sign
- Head nodding with pulse: de Musset's sign

Murmurs

- Early diastolic murmur
- Systolic murmur (increased stroke volume)
- Austin Flint murmur (soft mid-diastolic)

Other signs

- Displaced, thrusting apex beat (volume overload)
- Pre-systolic impulse
- Third heart sound
- Fourth heart sound
- Crepitations (pulmonary venous congestion)

- **Investigations**

- Doppler echocardiography is the investigation of first choice for detecting regurgitation . In severe acute aortic regurgitation the rapid rise in left ventricular diastolic pressure may cause premature mitral valve closure.

- Cardiac catheterisation and aortography are usually performed to assess the severity of regurgitation, to determine if there is dilatation of the aorta and to screen for the presence of coexisting CAD.

- MRI can also be useful in assessing the degree and extent of aortic dilatation if this is suspected on chest X-ray or echocardiography.

16.90 Investigations in aortic regurgitation

ECG

- Initially normal, later left ventricular hypertrophy and T-wave inversion

Chest X-ray

- Cardiac dilatation, maybe aortic dilatation
- Features of left heart failure

Echo

- Dilated left ventricle
- Hyperdynamic left ventricle
- Doppler detects reflux
- Fluttering anterior mitral leaflet

Cardiac catheterisation*

- Dilated left ventricle
- Aortic regurgitation
- Dilated aortic root

*Not always required.

- Management

- Treatment may be required for underlying conditions, such as endocarditis or syphilis.

- Aortic valve replacement is indicated if aortic regurgitation causes symptoms, and this may need to be combined with aortic root replacement and coronary bypass surgery.

- Those with chronic aortic regurgitation can remain asymptomatic for many years because compensatory ventricular dilatation and hypertrophy occur, but should be advised to report the development of any symptoms of breathlessness or angina. Asymptomatic patients should also be followed up annually with echocardiography for evidence of increasing ventricular size. If this occurs or if the end-systolic dimension increases to 55 mm or more, then aortic valve replacement should be undertaken.

- If systemic hypertension is present, non-rate-limiting vasodilators, such as nifedipine, should be used to control systolic BP. There is conflicting evidence regarding the need for aortic valve replacement in asymptomatic patients with severe aortic regurgitation. When aortic root dilatation is the cause of aortic regurgitation, as can occur in Marfan syndrome, aortic root replacement is usually necessary.