

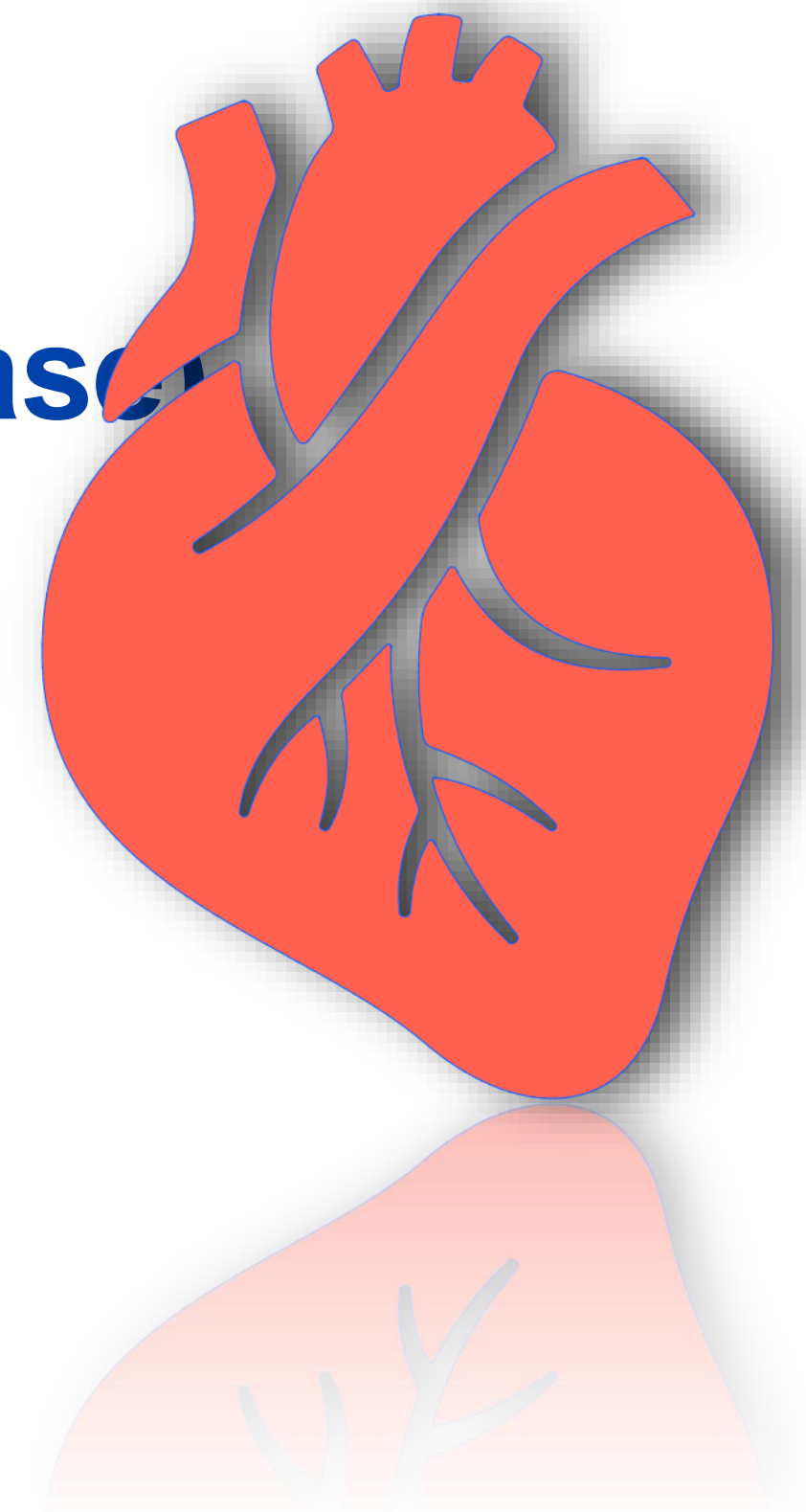
Disease of the heart valves

Mitral valve Disease

Sami Mukhlif Al-obaigy

Interventional Cardiologist and Senior Lecturer

**FIBMS Cardiol , CABM(Med.), D.M(Med) , D.M (Chest Diseases)
,MBCChB , MESC**



Mitral stenosis

- Mitral stenosis is almost always rheumatic in origin, although in older people it can be caused by heavy calcification of the mitral valve. There is also a rare form of congenital mitral stenosis.

Pathogenesis

- In rheumatic mitral stenosis, the mitral valve orifice is slowly diminished by progressive fibrosis, calcification of the valve leaflets, and fusion of the cusps and subvalvular apparatus. The mitral valve orifice is normally about 5cm² in diastole but can be reduced to < 1cm² in severe mitral stenosis. The patient is usually asymptomatic until the orifice is < 2cm². As stenosis progresses, **left ventricular filling becomes more dependent on left atrial contraction**. There is dilatation and hypertrophy of the LA and left atrial pressure rises, leading to **pulmonary venous congestion and breathlessness**. Any increase in heart rate shortens diastole when the mitral valve is open and produces a further rise in left atrial pressure. Situations that demand an increase in cardiac output, such as pregnancy and exercise, also increase left atrial pressure and are poorly tolerated.

- Atrial fibrillation is very common due to progressive dilatation of the LA. Its onset often precipitates pulmonary oedema because the accompanying tachycardia and loss of atrial contraction lead to haemodynamic deterioration and a rapid rise in left atrial pressure. In the absence of AF, a more gradual rise in left atrial pressure may occur. Irrespective of AF, pulmonary hypertension may occur, which can protect the patient from pulmonary oedema. Pulmonary hypertension leads to right ventricular hypertrophy and dilatation, tricuspid regurgitation and right heart failure. Fewer than 20% of patients remain in sinus rhythm but many of these have a small fibrotic LA and severe pulmonary hypertension.

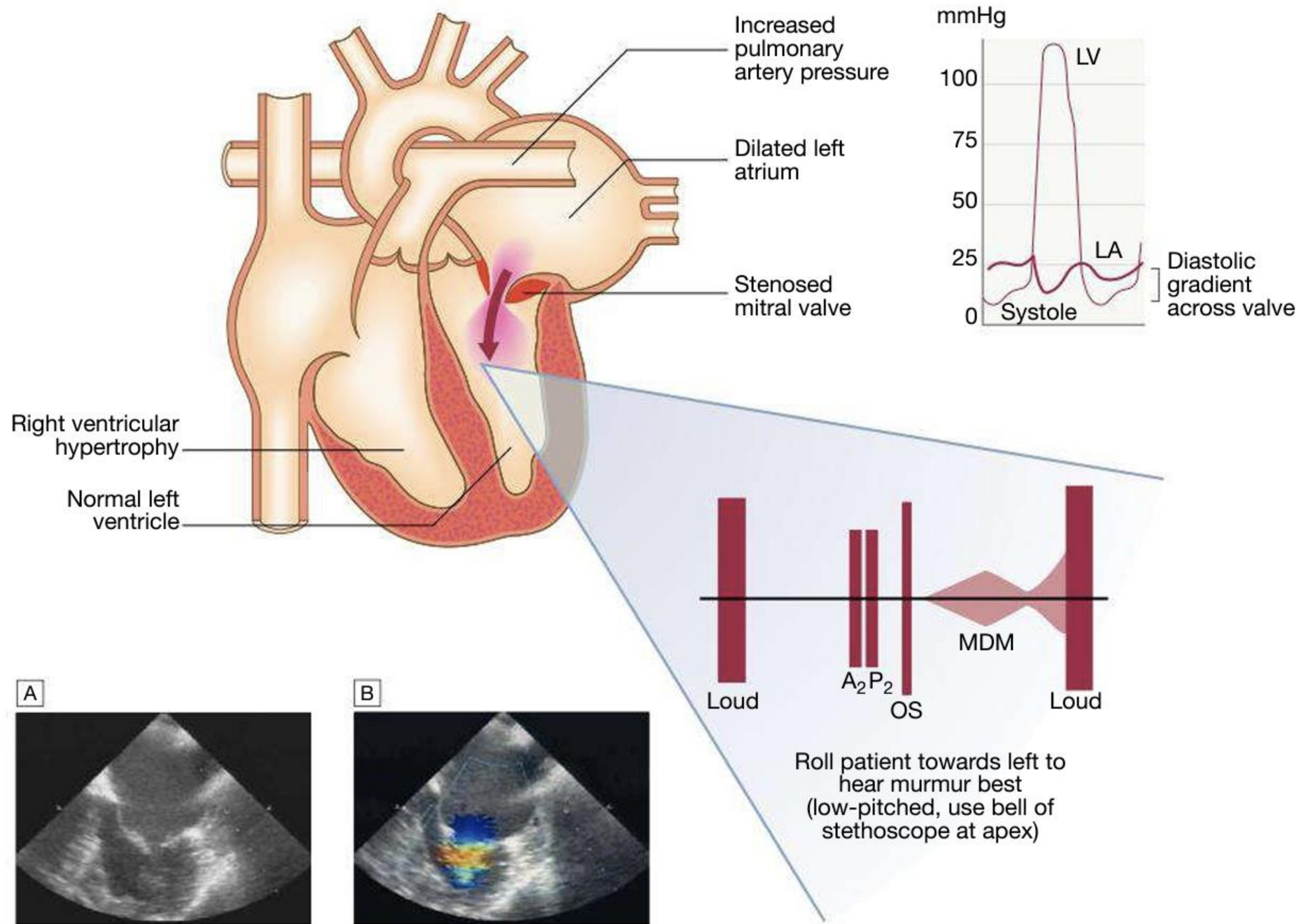


Fig. 16.79 Mitral stenosis: murmur and the diastolic pressure gradient between left atrium (LA) and left ventricle (LV). (Mean gradient is reflected by the area between LA and LV in diastole.) The first heart sound is loud, and there is an opening snap (OS) and mid-diastolic murmur (MDM) with pre-systolic accentuation.

A Echocardiogram showing reduced opening of the mitral valve in diastole. **B** Colour Doppler showing turbulent flow.

Clinical features

- Effort-related dyspnoea is usually the dominant symptom . Typically, exercise tolerance diminishes very slowly over many years until symptoms eventually occur at rest. Patients frequently do not appreciate the extent of their disability until the diagnosis is made and their valve disease is treated. Acute pulmonary oedema or pulmonary hypertension can lead to **haemoptysis**. Fatigue is a common symptom due to a low cardiac output. **Thromboembolism** is a common complication, especially in patients with AF. Prior to the advent of anticoagulant therapy, emboli caused one-quarter of all deaths.

- The physical signs of mitral stenosis are often found before symptoms develop and their recognition is of particular importance in pregnancy. The forces that open and close the mitral valve increase as left atrial pressure rises. The first heart sound (S1) is therefore loud and can be palpable (tapping apex beat). An opening snap may be audible and moves closer to the second sound (S2) as the stenosis becomes more severe and left atrial pressure rises. However, the first heart sound and opening snap may be inaudible if the valve is heavily calcified.

- Turbulent flow produces the characteristic low-pitched middiastolic murmur and sometimes a thrill . The murmur is accentuated by exercise and during atrial systole (pre-systolic accentuation). Early in the disease a pre-systolic murmur may be the only auscultatory abnormality, but in patients with symptoms, the murmur extends from the opening snap to the first heart sound. Coexisting mitral regurgitation causes a pansystolic murmur that radiates towards the axilla. Pulmonary hypertension may ultimately lead to right ventricular hypertrophy and dilatation with secondary tricuspid regurgitation, which causes a parasternal lift, and a systolic murmur and giant 'v waves' in the venous pulse.

Clinical feature	Cause
Symptoms	
Breathlessness	Pulmonary congestion, low cardiac output
Fatigue	Low cardiac output
Oedema, ascites	Right heart failure
Palpitation	Atrial fibrillation
Haemoptysis	Pulmonary congestion
Cough	Pulmonary congestion
Chest pain	Pulmonary hypertension
Thromboembolism	Atrial stasis and atrial fibrillation
Signs	
Atrial fibrillation	Atrial dilatation
Mitral facies	Low cardiac output
Auscultation:	
Loud first heart sound, opening snap	Non-compliant, stenotic valve
Mid-diastolic murmur	
Creptitations	Left heart failure
Pulmonary oedema	
Pleural effusions	
Right ventricular heave, loud P ₂	Pulmonary hypertension

- Investigations Doppler echocardiography is the investigation of choice for evaluation of suspected mitral stenosis .
- Cardiac catheterisation may also be required if surgery or valvuloplasty is being considered, to screen for coexisting conditions such as CAD.
- The ECG may show either AF or bifid P waves (P mitrale) associated with left atrial hypertrophy.
- CXR

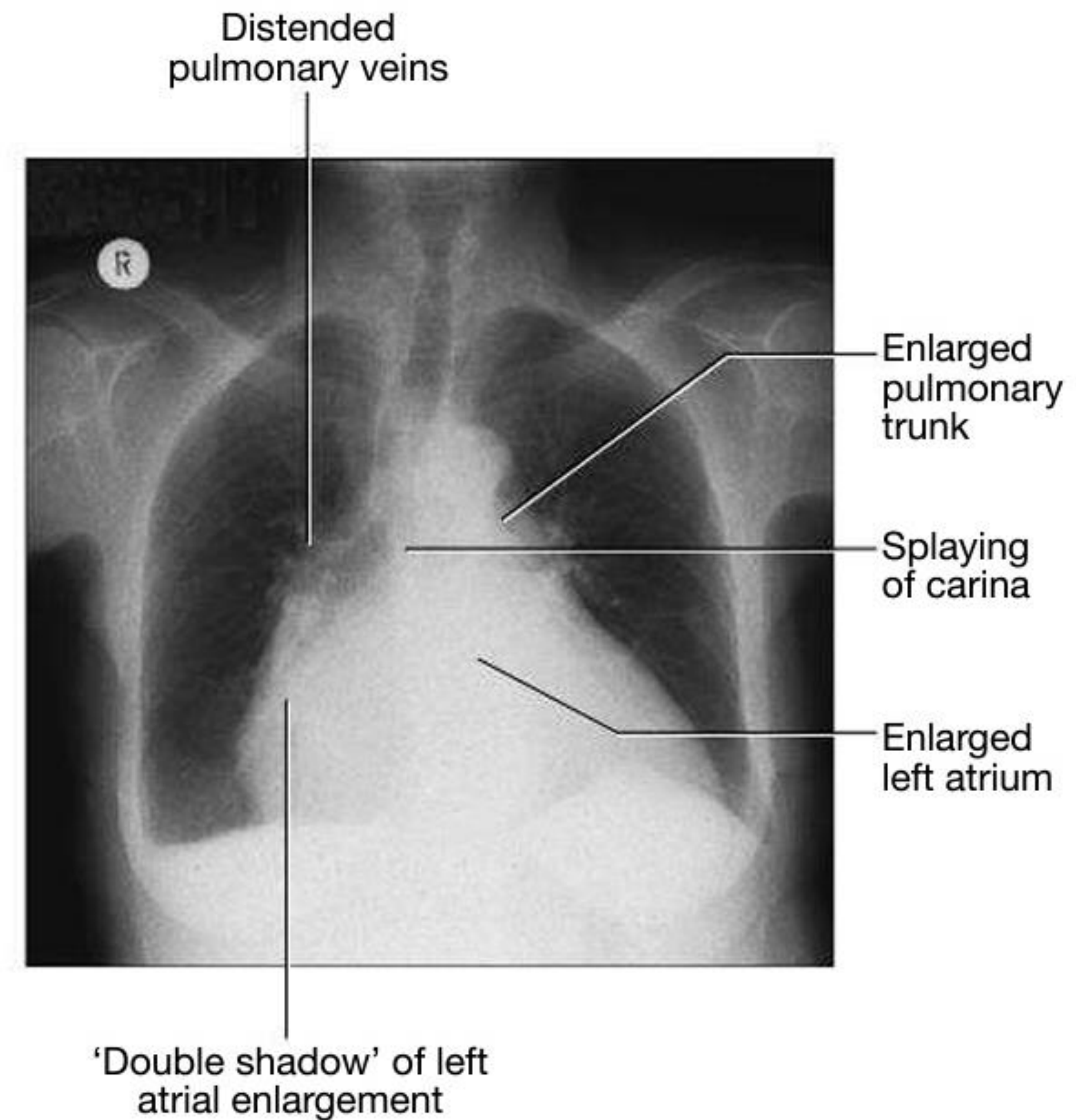


Fig. 16.9 Chest X-ray of a patient with mitral stenosis and regurgitation indicating enlargement of the LA and prominence of the pulmonary artery trunk.

i**16.78 Investigations in mitral stenosis****ECG**

- Right ventricular hypertrophy: tall R waves in V_1-V_3
- P mitrale or atrial fibrillation

Chest X-ray

- Enlarged left atrium and appendage
- Signs of pulmonary venous congestion

Echo

- Thickened immobile cusps
- Reduced valve area
- Enlarged left atrium
- Reduced rate of diastolic filling of left ventricle

Doppler

- Pressure gradient across mitral valve
- Pulmonary artery pressure
- Left ventricular function

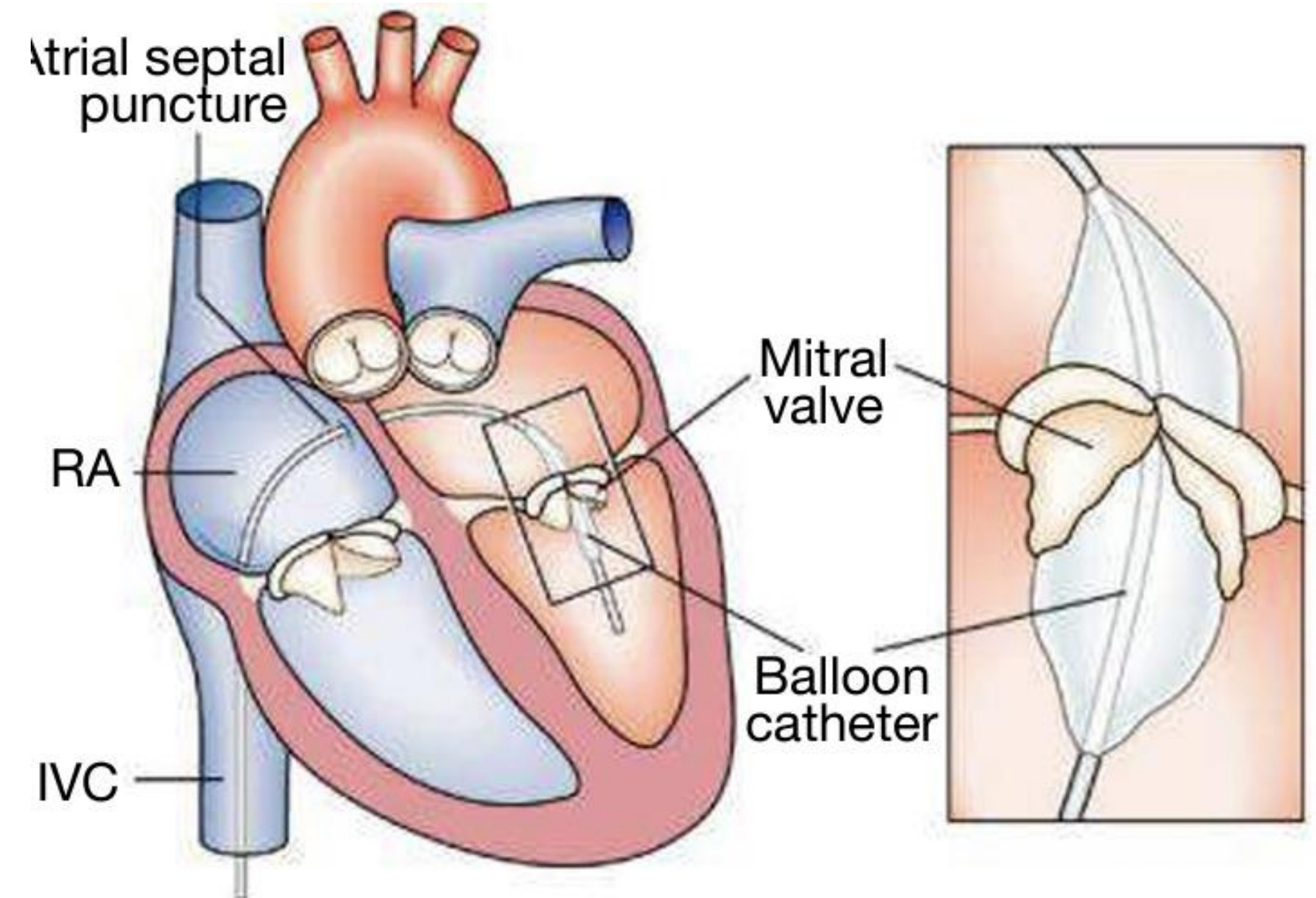
Cardiac catheterisation

- Coronary artery disease
- Pulmonary artery pressure
- Mitral stenosis and regurgitation

Management

- Patients with mild symptoms can be treated medically but intervention by balloon valvuloplasty, mitral valvotomy or mitral valve replacement should be considered if the patient remains symptomatic despite medical treatment or if pulmonary hypertension develops.
- Medical management This consists of **anticoagulation** to reduce the risk of systemic embolism, **ventricular rate control** with digoxin, β -blockers or rate-limiting calcium antagonists for AF, and **diuretic** to control pulmonary congestion.
- **Antibiotic prophylaxis** against infective endocarditis is no longer routinely recommended.

- Mitral balloon valvuloplasty and valve replacement Valvuloplasty is the treatment of choice if specific criteria are fulfilled , although surgical closed or open mitral valvotomy is an acceptable alternative. Patients who have undergone mitral valvuloplasty or valvotomy should be followed up at 1–2-yearly intervals because restenosis may occur. Clinical symptoms and signs are a guide to the severity of mitral restenosis but Doppler echocardiography provides a more accurate assessment. Valve replacement is indicated if there is substantial mitral reflux or if the valve is rigid and calcified



0 Mitral valvuloplasty. A guidewire is introduced into the right atrium the femoral vein and the inferior vena cava (IVC). The interatrial septum , providing access to the left atrium and mitral valve. A balloon catheter nced over the guidewire across the mitral valve and the balloon dilated t e valve and reduce the degree of stenosis.

i**16.79 Criteria for mitral valvuloplasty***

- Significant symptoms
- Isolated mitral stenosis
- No (or trivial) mitral regurgitation
- Mobile, non-calcified valve/subvalve apparatus on echo
- Left atrium free of thrombus

*For comprehensive guidelines on valvular heart disease, see www.acc.org.

Mitral regurgitation

- Rheumatic disease is the principal cause in countries where rheumatic fever is common but elsewhere, including in the UK, other causes are more important . Mitral regurgitation may also follow mitral valvotomy or valvuloplasty.

- **Pathogenesis**
- Chronic mitral regurgitation causes gradual dilatation of the LA with little increase in pressure and relatively few symptoms. Nevertheless, the LV dilates slowly and the left ventricular diastolic and left atrial pressures gradually increase as a result of **chronic volume overload** of the LV.
- In contrast, acute mitral regurgitation causes a rapid rise in left atrial pressure (because left atrial compliance is normal) and marked symptomatic deterioration.

- **Mitral valve prolapse**

- This is also known as ‘floppy’ mitral valve and is a common cause of mild mitral regurgitation . Some cases are thought to be due to a developmental abnormality of the mitral valve and others due to degenerative myxomatous change in a normal mitral valve. Rarely, mitral valve prolapse may occur in association with Marfan syndrome. In its mildest forms, the valve remains competent but bulges back into the atrium during systole, causing a mid-systolic click but no murmur. In the presence of a regurgitant valve, the click is followed by a late systolic murmur, which lengthens as the regurgitation becomes more severe. A click is not always audible and the physical signs may vary with both posture and respiration. Progressive elongation of the chordae tendineae leads to increasing mitral regurgitation, and if chordal rupture occurs, regurgitation suddenly becomes severe. This is rare before the fifth or sixth decade of life. Mitral valve prolapse is associated with a variety of typically benign arrhythmias, atypical chest pain and a very small risk of embolic stroke or transient ischaemic attack (TIA). Nevertheless, the overall long-term prognosis is good.

- Other causes of mitral regurgitation Mitral valve function depends on the chordae tendineae and their papillary muscles; dilatation of the LV distorts the geometry of these and may cause mitral regurgitation . Dilated cardiomyopathy and heart failure from CAD are common causes of so-called 'functional' mitral regurgitation. Endocarditis is an important cause of acute mitral regurgitation.

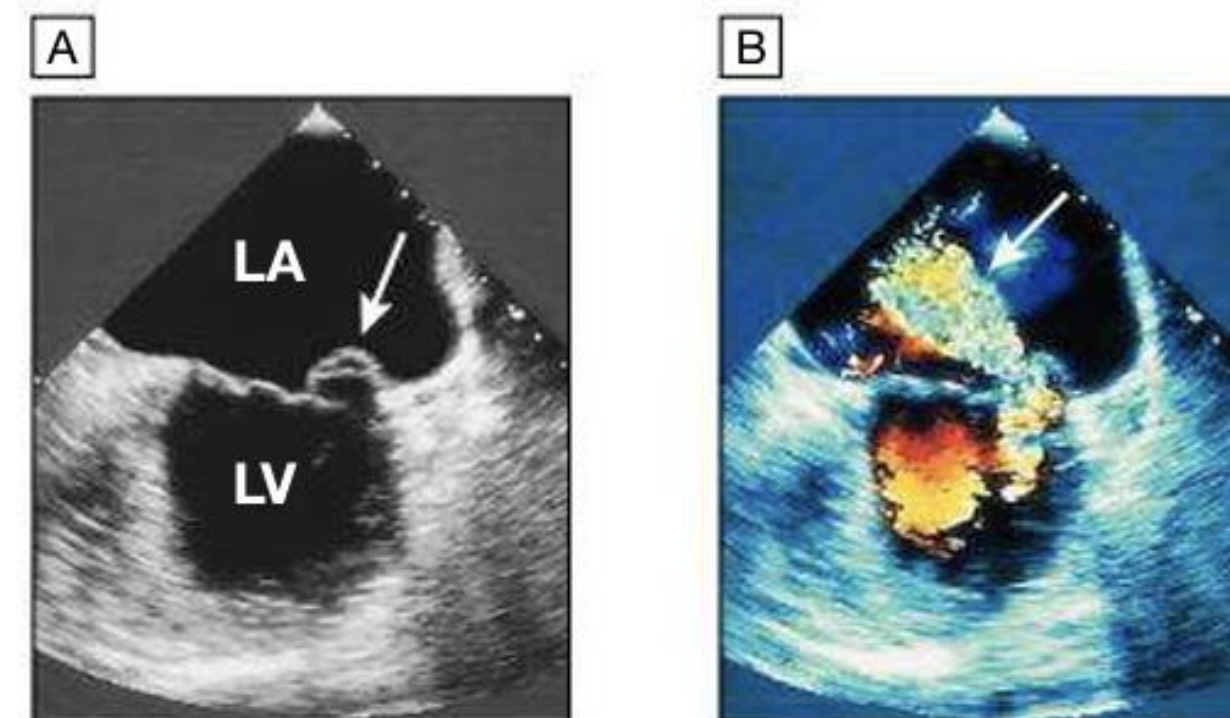
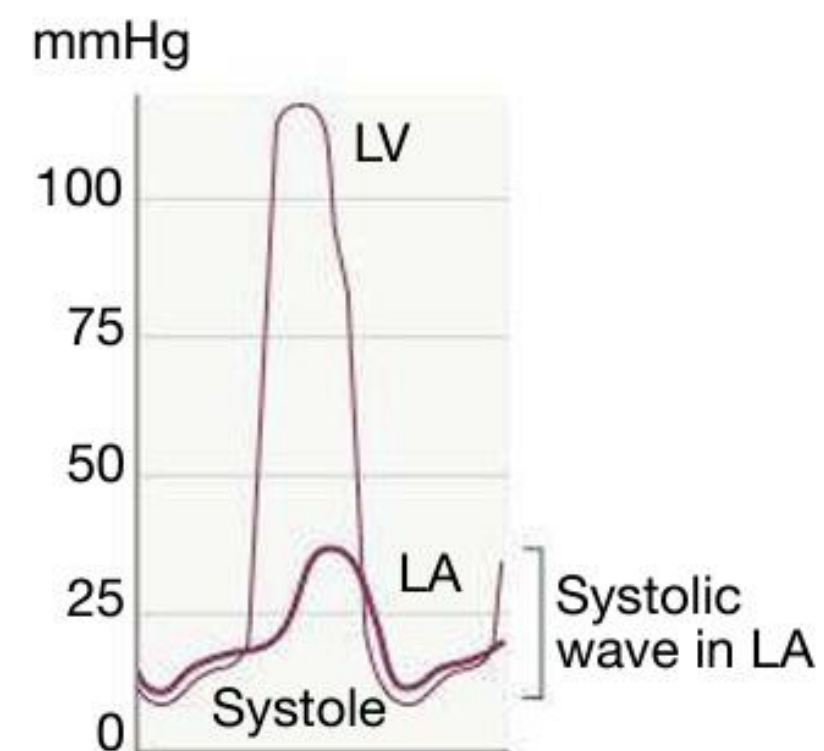
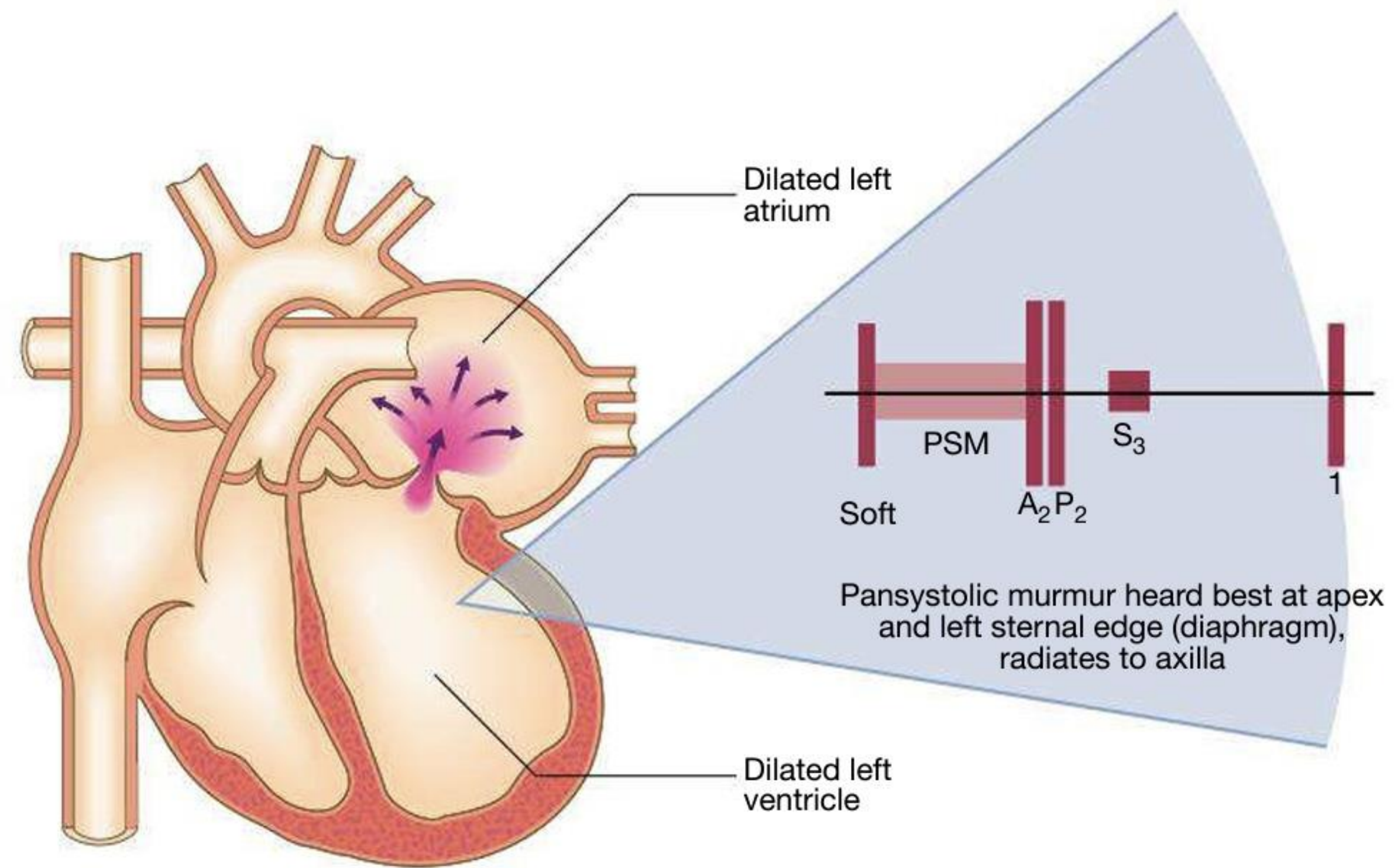


Fig. 16.81 Mitral regurgitation: murmur and systolic wave in left atrial pressure. The first sound is normal or soft and merges with a pansystolic murmur (PSM) extending to the second heart sound. A third heart sound occurs with severe regurgitation. **A** A transoesophageal echocardiogram shows mitral valve prolapse, with one leaflet bulging towards the left atrium (LA, arrow). **B** This results in a jet of mitral regurgitation on colour Doppler (arrow). (LV = left ventricle)

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16.80 Causes of mitral regurgitation

- Mitral valve prolapse
- Dilatation of the left ventricle and mitral valve ring (coronary artery disease, cardiomyopathy)
- Damage to valve cusps and chordae (rheumatic heart disease, endocarditis)
- Ischaemia or infarction of the papillary muscle
- Myocardial infarction

- Clinical features Symptoms and signs depend on the underlying cause and how suddenly the regurgitation develops . Chronic mitral regurgitation produces a symptom complex that is similar to that of mitral stenosis but sudden-onset mitral regurgitation usually presents with acute pulmonary oedema. The regurgitant jet causes an apical systolic murmur , which radiates into the axilla and may be accompanied by a thrill. Increased forward flow through the mitral valve causes a loud third heart sound and even a short mid-diastolic murmur. The apex beat feels active and rocking due to left ventricular volume overload and is usually displaced to the left as a result of left ventricular dilatation.



16.81 Clinical features of mitral regurgitation

Clinical feature	Cause
Symptoms	
Breathlessness	Pulmonary congestion
Fatigue	Low cardiac output
Oedema, ascites	Right heart failure
Palpitation	Atrial fibrillation
Signs	
Atrial fibrillation	Atrial dilatation
Displaced apex beat	Cardiomegaly
Auscultation:	
Apical pansystolic murmur	Regurgitation of blood from left ventricle to left atrium
Soft S1	Valve does not close properly
Apical S3	Rapid flow of blood into left ventricle
Crepitations	} Left heart failure
Pulmonary oedema	
Pleural effusions	
Right ventricular heave	Pulmonary hypertension
Raised jugular venous pressure	Right heart failure
Oedema	Right heart failure

- **Investigations**

- Echocardiography is a pivotal investigation. The severity of regurgitation can be assessed by Doppler and information may also be gained on papillary muscle function and valve prolapse. An ECG should be performed and commonly shows AF, as a consequence of atrial dilatation.
- Cardiac catheterisation is indicated when surgery is being considered . During catheterisation, the severity of mitral regurgitation can be assessed by left ventriculography and by the size of the v (systolic) waves in the left atrial or pulmonary artery wedge pressure trace.

i**16.82 Investigations in mitral regurgitation****ECG**

- P-mitrale
- Atrial fibrillation

Chest X-ray

- Enlarged left atrium
- Enlarged left ventricle
- Pulmonary venous congestion
- Pulmonary oedema (if acute)

Echo

- Dilated left atrium, left ventricle
- Dynamic left ventricle (unless myocardial dysfunction predominates)
- Structural abnormalities of mitral valve

Doppler

- Detects and quantifies regurgitation

Cardiac catheterisation

- Dilated left atrium, dilated left ventricle, mitral regurgitation
- Pulmonary hypertension
- Coexisting coronary artery disease

- **Management**

- Mitral regurgitation of moderate severity can be treated medically with diuretics and vasodilators. Digoxin and anticoagulants should be given if AF is present . If systemic hypertension is present, it should be treated with vasodilators such as ACE inhibitors or ARBs, since high afterload may worsen the degree of regurgitation. All patients should be reviewed at regular intervals, both clinically and by echocardiography. Worsening symptoms, progressive cardiomegaly or echocardiographic evidence of deteriorating left ventricular function are indications for mitral valve replacement or repair. Mitral valve repair is now the treatment of choice for severe mitral regurgitation, because early repair appears to prevent irreversible left ventricular damage.

- Mitral regurgitation often accompanies left ventricular failure associated with CAD. If such patients are to undergo CABG surgery, it is common practice to repair the valve and restore mitral valve function by inserting an annuloplasty ring to overcome annular dilatation and to bring the valve leaflets closer together. Unfortunately, it can be difficult to determine whether it is the ventricular dilatation or the mitral regurgitation that is the predominant problem. If ventricular dilatation is the underlying cause of mitral regurgitation, then mitral valve repair or replacement may actually worsen ventricular function, as the ventricle can no longer empty into the low-pressure LA.

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16.83 Medical management of mitral regurgitation

- Diuretics
- Vasodilators if hypertension is present
- Digoxin if atrial fibrillation is present
- Anticoagulants if atrial fibrillation is present