The Cardiology Block

Rheumatic fever and mitral stenosis, STEMI, decompensated heart failure, endocarditis, hypertension, valve disease, pericardial disease and the cardiomyopathies.

Cases 9–18 · 13,852 words

This block covers the cardiology teaching of both internal medicine courses. The **first course** supplies cases 9 and 14; the **second course** supplies cases 15 and 18, which extend the block into valvular, pericardial and myocardial disease.

Case	Lecture it serves
System opener: the cardiovascular history and examination	History taking in cardiovascular; physical examination of the cardiovascular system
Case 9 — Acute rheumatic fever	Rheumatic fever
Case 10 — Rheumatic mitral stenosis	Rheumatic heart disease
Case 11 — ST-elevation myocardial infarction	Ischaemic heart disease
Case 12 — Acute decompensated heart failure	Heart failure
Case 13 — Infective endocarditis	Infective endocarditis
Case 14 — Newly diagnosed hypertension	Hypertension
Case 15 — Aortic stenosis	Valvular heart disease, part 1
Case 16 — Chronic regurgitant valve disease	Valvular heart disease, part 2
Case 17 — Pericardial disease	Pericardial disease
Case 18 — The cardiomyopathies	Cardiomyopathies (two lectures)

Hypertensive urgency and emergency is deliberately held back for the seminar case in Part II, so that Case 14 can concentrate on assessment of the newly diagnosed patient. **Pulmonary thromboembolism** is Seminar 11, and **the four pillars of heart failure therapy** are set out in Case 12 — both are cross-referenced from the second-course cases rather than duplicated.

System Opener · The Cardiovascular History and Examination

The history

The five cardinal symptoms

Symptom	What to establish
Chest pain	Site, character, radiation, duration, provoking and relieving factors, and associated autonomic symptoms. Ischaemic pain is heavy or gripping, retrosternal, radiates to arm, neck or jaw, comes on with exertion and settles with rest. Pain that is sharp, positional, or reproduced by pressing on the chest wall is usually not ischaemic — but "atypical" pain in a diabetic or elderly patient still needs an electrocardiogram.
Breathlessness	Exertional tolerance, orthopnoea counted in pillows, paroxysmal nocturnal dyspnoea, and the New York Heart Association class. Ask what changed and over what period.
Palpitations	Ask the patient to tap out the rhythm on the desk. Fast or slow, regular or irregular, abrupt or gradual in onset and offset, and whether accompanied by syncope or chest pain.
Syncope	The circumstances matter more than the event. Syncope during exertion is aortic stenosis or hypertrophic cardiomyopathy until proved otherwise. Cardiac syncope is sudden, without warning, with rapid full recovery. A long prodrome with sweating and nausea suggests a vasovagal cause.
Oedema and weight gain	Ankles by evening, sacral swelling in a bedbound patient, abdominal distension, and rapid weight gain — the most sensitive marker of fluid retention.

Risk and background

- Smoking, diabetes mellitus, hypertension, dyslipidaemia, obesity, physical inactivity, chronic kidney disease, and **premature cardiovascular disease in a first-degree relative** — before 55 in men and 65 in women.
- **Rheumatic history:** recurrent sore throats in childhood, a murmur heard at school or before marriage, previous rheumatic fever, and whether monthly penicillin injections were ever given and for how long.
- Family history of sudden death, cardiomyopathy or inherited arrhythmia, and consanguinity where relevant.
- Full drug history including over-the-counter and herbal preparations, and previous procedures — angiography, stents, bypass grafting, valve surgery, pacemakers.

The examination

General and peripheral

- Breathlessness at rest, cachexia, malar flush, marfanoid habitus, central cyanosis, and the observation chart.
- **Hands:** clubbing (cyanotic congenital heart disease, infective endocarditis), splinter haemorrhages, Osler nodes, Janeway lesions, tendon xanthomata, tar staining, peripheral cyanosis.
- **Pulse:** rate, rhythm and character. A slow-rising pulse indicates aortic stenosis; a collapsing pulse aortic regurgitation; pulsus alternans severe left ventricular dysfunction. Check for **radio-radial and radio-**

femoral delay — the latter is the sign of coarctation, and it is missed by not looking for it.

- **Blood pressure in both arms**, with a postural reading. A wide pulse pressure suggests aortic regurgitation, a narrow one severe aortic stenosis.

The jugular venous pressure

Students lose more marks here than anywhere else in the cardiovascular examination. Position the patient at 45 degrees, turn the head slightly away, and look across rather than at the neck.

How to know it is venous, not arterial	The waveform and what it means
It is not palpable	a wave — atrial contraction. Absent in atrial fibrillation. Large in pulmonary hypertension and tricuspid stenosis. Cannon waves in complete heart block, when the atrium contracts against a closed tricuspid valve.
It has two peaks per cardiac cycle	v wave — atrial filling. Giant systolic v waves are the sign of tricuspid regurgitation, and are accompanied by a pulsatile liver.
It varies with position and respiration	Kussmaul sign — the venous pressure rises rather than falls on inspiration: constrictive pericarditis, and also tamponade and right ventricular infarction.
It is obliterated by gentle pressure at the base of the neck, and rises with pressure over the liver	A raised pressure with a normal waveform means fluid overload or right heart failure.

The praecordium

- **Inspect** for a midline sternotomy scar, a left lateral thoracotomy scar (the old closed mitral valvotomy), a pacemaker or defibrillator box, and visible pulsation.
- **Palpate the apex beat** — its position, and then its character. A **tapping** apex is a palpable first heart sound and means mitral stenosis. A **heaving, sustained** apex is pressure loading: aortic stenosis or hypertension. A **thrusting, displaced** apex is volume loading: mitral or aortic regurgitation. Feel for thrills and for a left parasternal heave of right ventricular hypertrophy.
- **Auscultate** all four areas with diaphragm and bell. Identify the first and second sounds by timing against the carotid pulse before attempting to describe anything else. Listen for added sounds — a third sound, a fourth sound, an opening snap, an ejection click, a metallic prosthetic click, a pericardial rub.
- **Finish** at the lung bases, the liver (pulsatile in tricuspid regurgitation), for ascites, at the sacrum and ankles, at all peripheral pulses, and with the fundi and a urine dipstick where endocarditis is possible.

Murmurs — the table to know

Lesion	Timing and character	Best heard	Accentuating manoeuvre
Aortic stenosis	Ejection systolic, harsh, radiating to the carotids	Right second intercostal space	Sitting forward, expiration
Aortic regurgitation	Early diastolic, soft, high-pitched, decrescendo	Left sternal edge	Sitting forward, breath held in expiration
Mitral stenosis	Mid-diastolic, low-pitched rumble with presystolic accentuation	Apex, with the bell	Left lateral position, expiration
Mitral regurgitation	Pansystolic, blowing, radiating to the axilla	Apex	Left lateral position, expiration
Tricuspid regurgitation	Pansystolic with giant v waves and a pulsatile liver	Left sternal edge	Louder on inspiration
Ventricular septal defect	Harsh pansystolic with a thrill	Left sternal edge	Expiration
Hypertrophic cardiomyopathy	Ejection systolic, no carotid radiation	Left sternal edge	Louder on standing and Valsalva, softer on squatting

The rule that saves you in an examination: right-sided murmurs are louder on **in**spiration, left-sided murmurs on **ex**piration. And a murmur you cannot classify should still be described accurately — timing, site, radiation and what it does with respiration — because an accurate description scores more than a confident wrong label.

Case 9 · Acute Rheumatic Fever

CLINICAL VIGNETTE

A **12-year-old boy** from a rural district is brought in with fever and joint pain. Three weeks ago he had a severe sore throat that was not treated. Six days ago his right knee became hot, swollen and so painful he could not bear weight; that settled over two days and the pain moved to his left ankle, then to his left wrist.

His mother says he tires easily and has become breathless climbing stairs. He is the fourth of seven children in a crowded household.

On examination: temperature 38.4 °C, heart rate 128/min while asleep, respiratory rate 24/min. The left wrist is swollen and exquisitely tender. There is a **soft pansystolic murmur at the apex radiating to the axilla** that was not previously known. Over the trunk there are faint pink rings with clear centres that have changed position since admission.

1. Focused History

- **The preceding infection.** A sore throat two to four weeks earlier — this latent period is what makes the connection easy to miss, because the throat has long recovered. Ask whether antibiotics were given and completed.
- **Characterise the arthritis.** Rheumatic arthritis is **migratory**, affects large joints asymmetrically, is exquisitely painful relative to the swelling, and settles in each joint within days. It responds dramatically to anti-inflammatory drugs — so dramatically that failure to respond should make you question the diagnosis.
- **Symptoms of carditis:** breathlessness, orthopnoea, reduced exercise tolerance, chest pain, palpitations.
- **Symptoms of chorea:** clumsiness, dropping objects, deteriorating handwriting, emotional lability and behavioural change. Chorea may appear months after the infection, long after everything else has settled, and parents often report it as naughtiness.
- **Previous episodes and prophylaxis.** A previous attack is the strongest risk factor for another, and each recurrence damages the valves further. Ask specifically whether monthly injections were given and when they stopped.
- Household crowding, access to healthcare, and other affected family members.

2. Physical Examination

- **Fever and a tachycardia out of proportion to the fever**, persisting during sleep.
- **Arthritis** — hot, swollen, extremely tender large joints, asymmetric and migratory. Document which joints and when, because the migration itself is diagnostic.
- **Carditis** — the feature that determines the patient's future. Listen for a **new or changed murmur**: mitral regurgitation is the commonest, then aortic regurgitation. Listen for a pericardial rub and a third sound, and look for signs of heart failure. A resting tachycardia may be the only sign.
- **Erythema marginatum** — pink macules with a clear centre and a serpiginous margin, on the trunk and proximal limbs, **sparing the face**, evanescent and changing position over hours. Easily missed on darker skin, so look in good light.

- **Subcutaneous nodules** — firm, painless, mobile, over extensor surfaces and the occiput. Uncommon but strongly associated with severe carditis.
- **Sydenham chorea** — purposeless jerky movements, hypotonia, the **milkmaid grip** (irregular squeezing of the examiner's fingers) and the darting tongue that cannot be held protruded.

3. Diagnosis: The Revised Jones Criteria

BEFORE YOU APPLY THE CRITERIA

The criteria differ according to how common rheumatic fever is in the population. **This region is classified as moderate-to-high risk**, so the lower thresholds below apply — using the low-risk criteria here will cause you to miss cases.

Diagnosis requires **evidence of a preceding streptococcal infection**, plus either **two major** criteria or **one major and two minor**.

Major criteria	Minor criteria (moderate-to-high risk population)
Carditis — clinical or subclinical on echocardiography	Monoarthralgia
Arthritis — polyarthritis, or monoarthritis, or polyarthralgia in this risk group	Fever of 38 °C or above
Chorea	Erythrocyte sedimentation rate 30 mm/h or more, or raised C-reactive protein
Erythema marginatum	Prolonged PR interval for age
Subcutaneous nodules	

- **Two exceptions:** chorea alone, and indolent carditis, are sufficient without evidence of preceding streptococcal infection, because both appear so long after the throat infection that serology may already have fallen.
- A recurrence in a patient with established rheumatic heart disease may be diagnosed on **three minor criteria** with evidence of streptococcal infection.

4. Differential Diagnosis

Diagnosis	The feature that discriminates it
Septic arthritis	One joint, not migratory, systemically unwell, pus on aspiration. Aspirate any single hot joint before assuming rheumatic fever.
Brucellosis	Endemic in this region. Unpasteurised dairy or animal contact, sacroiliac or hip involvement, hepatosplenomegaly, undulant fever, positive serology.
Juvenile idiopathic arthritis	Persistent rather than migratory, symmetric small joints, morning stiffness, and a much longer course.
Infective endocarditis	Also produces fever and a murmur. Blood cultures and echocardiography separate them, and the two can be confused fatally.
Systemic lupus erythematosus	Rash on the face rather than sparing it, cytopenias, renal involvement, positive antinuclear antibody.
Sickle cell vaso-occlusive crisis	Known disease, bone rather than joint pain, anaemia with a raised reticulocyte count.
Reactive and post-streptococcal reactive arthritis	Shorter latent period, non-migratory, poor response to salicylates, and no carditis.

5. Investigations

- **Evidence of streptococcal infection:** throat swab culture and rapid antigen test — often negative by now — and **antistreptolysin O and anti-DNase B titres**, ideally paired two weeks apart to demonstrate a rise. A single raised titre is weaker evidence than a rising one.
- **Inflammatory markers:** erythrocyte sedimentation rate and C-reactive protein, used both for diagnosis and to follow the course.
- **Electrocardiogram** — a prolonged PR interval is a minor criterion. Higher grades of block can occur and are usually transient.
- **Echocardiography in every suspected case, even when no murmur is audible.** Subclinical carditis is a major criterion and changes both the diagnosis and the duration of prophylaxis. This is the single most important investigation.
- **Blood cultures** to exclude endocarditis, complete blood count, and joint aspiration if a single joint is involved.
- Chest radiograph if there is any suspicion of cardiac failure.

6. Management

Treat the streptococcus

- **Benzathine benzylpenicillin** as a single intramuscular dose, or oral penicillin V for ten days. Give it even when the throat swab is negative — you are eradicating carriage, not treating pharyngitis. Use a macrolide if penicillin-allergic.

Treat the inflammation

- Aspirin in anti-inflammatory doses, or naproxen, for the arthritis. The response is usually rapid and complete.
- Corticosteroids are used for severe carditis with heart failure, although the evidence that they alter long-term valve outcome is weak. Treat the heart failure itself on its merits.
- Chorea is usually self-limiting over weeks to months. Reassure the family that it is not behavioural. Carbamazepine or sodium valproate for severe or disabling movements.
- Rest during the acute illness, with gradual return to activity guided by the carditis.

SECONDARY PROPHYLAXIS — THE INTERVENTION THAT ACTUALLY CHANGES THE OUTCOME

The acute attack is rarely what harms the patient. **Recurrences are.** Each one adds valve damage, and the risk is highest in the first five years.

Benzathine benzylpenicillin intramuscularly every four weeks — every three weeks in high-risk patients or where breakthrough occurs. Oral penicillin V twice daily is an inferior alternative reserved for those who refuse injections.

Duration: no carditis — 5 years, or until age 21, whichever is longer · carditis without residual valve disease — 10 years, or until age 21, whichever is longer · **carditis with persistent valve disease — 10 years, or until age 40**, and often lifelong.

Register the patient in a recall system, warn the family that stopping early is the commonest reason young adults present with severe mitral stenosis, and arrange dental review.

7. Teaching Points and Viva Questions

- The arthritis moves; the carditis stays. The joints cause the symptoms, the heart causes the disease.
- Echocardiography in every suspected case — subclinical carditis is invisible at the bedside and alters the prophylaxis by decades.
- An unexplained resting tachycardia during sleep is carditis until proved otherwise.
- Aspirate any single hot joint. Septic arthritis destroys a joint in days and does not wait for serology.
- The most valuable thing you can do for this boy is not what you prescribe today, but ensuring he receives an injection every four weeks for the next ten years.

Questions you should be able to answer:

- State the Jones criteria and explain why they differ between populations.
- Why can chorea be diagnosed without evidence of streptococcal infection?
- What are the two most useful investigations here, and why?
- How long will this boy need prophylaxis, and on what does that depend?
- His antistreptolysin O titre is normal. Does that exclude the diagnosis?

Case 10 · Rheumatic Mitral Stenosis

CLINICAL VIGNETTE

A **28-year-old woman** attends with six months of breathlessness on exertion, now limiting her after one flight of stairs. Over the past two weeks she has felt her heart racing irregularly and has coughed up small amounts of blood twice. She is planning a pregnancy.

She had rheumatic fever at the age of nine and received penicillin injections for four years before they were stopped.

On examination: she has a **malar flush**. The pulse is 116/min and irregularly irregular, of small volume. Blood pressure 104/70 mmHg. The jugular venous pressure is raised. The apex beat is undisplaced and **tapping**. There is a left parasternal heave and a loud pulmonary second sound. On auscultation with the bell in the left lateral position: a **loud first heart sound, an opening snap, and a low-pitched rumbling mid-diastolic murmur** at the apex.

1. Focused History

- **Functional class** — quantify in stairs or metres, and establish over what period it changed. Deterioration over weeks usually means a new arrhythmia rather than progression of the valve lesion.
- **Orthopnoea, paroxysmal nocturnal dyspnoea and haemoptysis.** Haemoptysis in mitral stenosis arises from raised pulmonary venous pressure and rupture of dilated bronchial veins.
- **Palpitations** and their relation to the deterioration. **Atrial fibrillation is the event that unmasks mitral stenosis**, because losing atrial contraction and shortening diastole together collapse left ventricular filling.
- **Embolic events** — transient neurological symptoms, stroke, a cold painful limb, flank pain. The large fibrillating left atrium is an excellent source of thrombus.
- **Compression symptoms** from a dilated left atrium: hoarseness from recurrent laryngeal nerve compression, and dysphagia.
- **Pregnancy plans.** This is not a social question here — it is central to management, and the answer changes the timing of intervention.
- Rheumatic fever history and, specifically, when prophylaxis stopped.

2. Physical Examination

The classical signs, and what produces each

Sign	Mechanism
Malar flush	Low cardiac output with cutaneous vasodilatation
Small-volume, irregularly irregular pulse	Fixed low stroke volume, with atrial fibrillation
Tapping apex beat, undisplaced	A palpable loud first heart sound. The left ventricle is not dilated — it is underfilled.
Loud first heart sound	The stenosed, still-mobile valve closes from a wide-open position
Opening snap	The pliable valve opening abruptly. Its absence suggests a calcified, immobile valve.
Mid-diastolic rumble with presystolic accentuation	Turbulent flow across the narrowed valve; the presystolic component is atrial contraction and is lost once atrial fibrillation develops
Left parasternal heave, loud pulmonary second sound	Pulmonary hypertension and right ventricular hypertrophy
Raised venous pressure, pulsatile liver, ascites, oedema	Right heart failure — the end of the natural history

THREE BEDSIDE SIGNS OF SEVERITY

A shorter interval between the second sound and the opening snap — a high left atrial pressure forces the valve open sooner.

A longer murmur — the gradient persists throughout diastole.

Signs of pulmonary hypertension, and the presence of atrial fibrillation.

Note that the **loudness of the murmur says nothing about severity**. A very tight valve with low flow may be almost silent.

3. Differential Diagnosis

Diagnosis	The feature that discriminates it
Left atrial myxoma	A "tumour plop" rather than an opening snap, positional symptoms, systemic features and weight loss. Excluded by echocardiography.
Austin Flint murmur of aortic regurgitation	A diastolic rumble at the apex, but with a collapsing pulse, wide pulse pressure and an early diastolic murmur at the left sternal edge.
Tricuspid stenosis	Louder on inspiration, with giant a waves in the venous pulse.
Other causes of breathlessness in a young woman	Anaemia, thyrotoxicosis, pulmonary embolism, primary pulmonary hypertension.

4. Investigations

- **Electrocardiogram:** P mitrale — a broad bifid P wave — while sinus rhythm persists, then atrial fibrillation. Right axis deviation and right ventricular hypertrophy indicate pulmonary hypertension.
- **Chest radiograph — reading the film:** a straightened left heart border from the enlarged left atrial appendage, a **double density** behind the right heart border, **splaying of the carina**, upper lobe blood

diversion, Kerley B lines, and sometimes valve calcification. The heart is often not enlarged overall, which surprises students.

- **Echocardiography is the diagnostic and grading test.** It gives the valve area, the mean transmitral gradient, the pulmonary artery pressure, the left atrial size, and the presence of associated regurgitation.
- **Transoesophageal echocardiography** before any intervention, to exclude left atrial appendage thrombus, and to score valve morphology for suitability for balloon valvuloplasty.
- Complete blood count, thyroid function (atrial fibrillation), renal function, and natriuretic peptide where the cause of breathlessness is uncertain.

Valve area	Grade
Above 1.5 cm^2	Mild
1.0–1.5 cm^2	Moderate
Below 1.0 cm^2	Severe (the normal valve area is 4–6 cm^2)

5. Management

Medical

- **Rate control** with a beta blocker, or digoxin in atrial fibrillation. Slowing the heart lengthens diastole and improves filling — the opposite of the instinct to treat breathlessness by increasing output.
- **Anticoagulation with a vitamin K antagonist.** In atrial fibrillation with rheumatic mitral stenosis, **direct oral anticoagulants are contraindicated** and warfarin must be used. This is one of the few remaining absolute indications for warfarin and is a favourite examination question.
- Diuretics and salt restriction for pulmonary congestion.
- **Continue or restart secondary penicillin prophylaxis**, and maintain dental hygiene.

Intervention

- **Percutaneous balloon mitral valvuloplasty** is the treatment of choice for a pliable, non-calcified valve with little regurgitation and no left atrial thrombus. It is highly effective and avoids surgery — particularly valuable in a young woman planning pregnancy.
- **Surgical commissurotomy or valve replacement** when the valve is calcified, when there is significant mitral regurgitation, or when thrombus is present.
- **Indications:** symptomatic severe stenosis; and asymptomatic severe stenosis with pulmonary hypertension, new atrial fibrillation, or a planned pregnancy.

WHY THE PREGNANCY QUESTION MATTERED

Cardiac output rises by up to 50% and heart rate increases through pregnancy. Across a **fixed** stenotic valve, both changes raise left atrial pressure sharply, and women commonly decompensate in the second trimester or at delivery.

Severe mitral stenosis should be treated before conception. Counsel this patient now, not when she is 20 weeks pregnant.

6. Teaching Points and Viva Questions

- Mitral stenosis is a disease of diastole. Anything that shortens diastole — tachycardia, atrial fibrillation, fever, pregnancy, exercise — decompensates it.
- The interval from the second sound to the opening snap gauges severity; the loudness of the murmur does not.
- You will not hear the murmur with the diaphragm, or with the patient lying flat. Bell, left lateral, expiration.
- Warfarin, not a direct oral anticoagulant, when atrial fibrillation accompanies rheumatic mitral stenosis.
- An undisplaced apex in a breathless patient with a raised venous pressure should turn your attention to the mitral valve and the right heart.

Questions you should be able to answer:

- Explain each of the classical signs in terms of the underlying haemodynamics.
- Why does atrial fibrillation cause such abrupt deterioration in this condition?
- Why is warfarin required rather than a direct oral anticoagulant?
- Which patients are suitable for balloon valvuloplasty, and what must be excluded first?
- She becomes pregnant before any intervention. What are the risks and how would you manage her?

Case 11 · ST-Elevation Myocardial Infarction

CLINICAL VIGNETTE

A **58-year-old man** presents with 90 minutes of central chest pain, described as a weight on his chest, radiating to the left arm and jaw. He is sweating, nauseated and frightened. The pain began at rest and has not eased.

He smokes 20 cigarettes a day, has type 2 diabetes mellitus and hypertension, and his father died of a heart attack at 52.

On arrival: he is grey and clammy. Heart rate 92/min, blood pressure 148/88 mmHg in the right arm and 144/86 mmHg in the left, respiratory rate 22/min, oxygen saturation 96% on air. Heart sounds are normal with no murmur. Chest is clear.

Electrocardiogram, recorded 6 minutes after arrival: ST elevation of 3 mm in leads V1 to V4, with reciprocal ST depression in leads II, III and aVF.

1. Focused History — Taken While Treatment Proceeds

- **The time of symptom onset.** This is the single most important number in the history, because it determines the reperfusion strategy and the expected benefit. Establish it precisely, and record it.
- Characterise the pain, and ask specifically about features that suggest an alternative: **tearing pain radiating to the back** with maximal intensity at onset (aortic dissection), pleuritic or positional pain relieved by sitting forward (pericarditis), pain after vomiting (oesophageal rupture).
- Previous angina, infarction, angiography, stents or bypass grafting — and whether this pain is the same as before but worse.
- Whether aspirin has already been given, by the patient or by the ambulance crew.
- **Cocaine or amphetamine use**, particularly in a younger patient, which changes management.
- **Bleeding risk and contraindications to fibrinolysis**, in case primary angioplasty is not available within the time window: previous intracranial bleed, recent stroke, surgery, trauma or gastrointestinal bleeding, and anticoagulant use.

2. Physical Examination

Brief and targeted. It should not delay the electrocardiogram or reperfusion.

- **Blood pressure in both arms** and all peripheral pulses — a significant difference raises aortic dissection, which would make antiplatelet and anticoagulant therapy catastrophic.
- **Jugular venous pressure** — raised with clear lung fields in right ventricular infarction.
- **Auscultation** for a new murmur (mitral regurgitation from papillary muscle dysfunction, or a ventricular septal defect), a third or fourth heart sound, and a pericardial rub.
- **Lung bases**, to assign the Killip class, which predicts mortality directly.

Killip class	Findings
I	No crackles, no third heart sound
II	Crackles in less than half the lung fields, or a third heart sound, or a raised venous pressure
III	Frank pulmonary oedema
IV	Cardiogenic shock

3. Investigations

THE PRIORITY

The electrocardiogram must be recorded and interpreted within 10 minutes of arrival. Everything else follows from it.

ST elevation of 1 mm or more in two contiguous limb leads, or 2 mm in two contiguous chest leads (1.5 mm in women), or a new left bundle branch block in a patient with a compatible history.

Do not wait for the troponin. In ST-elevation infarction the troponin adds nothing to the decision and delays reperfusion.

Localise the infarct

Leads with ST elevation	Territory	Artery
V1–V4	Anterior / anteroseptal	Left anterior descending
I, aVL, V5–V6	Lateral	Circumflex
II, III, aVF	Inferior	Right coronary (or circumflex)
Tall R waves and ST depression in V1–V2	Posterior — record posterior leads V7–V9 to confirm	Circumflex or right coronary
ST elevation in V4R	Right ventricular — record right-sided leads in every inferior infarct	Proximal right coronary

- **Bloods:** troponin (for later confirmation and infarct size), complete blood count, urea and electrolytes, glucose and glycated haemoglobin, lipid profile, coagulation screen.
- **Portable chest radiograph** — but never allow it to delay reperfusion. It is looking for pulmonary oedema and for a widened mediastinum.
- **Echocardiography** — regional wall motion abnormalities, ejection fraction, and mechanical complications. Urgently if the diagnosis is uncertain or the patient is shocked.

4. Differential Diagnosis

Diagnosis	The feature that discriminates it
Aortic dissection	Tearing pain radiating to the back, maximal at onset, pulse or blood pressure differential, widened mediastinum. Must be excluded before anticoagulation.
Acute pericarditis	Sharp pleuritic pain relieved by sitting forward, saddle-shaped ST elevation in all territories with PR depression, no reciprocal change.
Pulmonary embolism	Pleuritic pain, hypoxia, sinus tachycardia, right heart strain pattern rather than territorial ST elevation.
Takotsubo cardiomyopathy	Preceded by intense emotional or physical stress, apical ballooning on echocardiography, unobstructed coronary arteries.
Oesophageal spasm or rupture	Related to swallowing or preceded by vomiting; surgical emphysema in rupture.
Early repolarisation	A young, well patient; concave elevation with notched J points, no reciprocal change, and no evolution on serial tracings.

5. Management

Immediate, at the bedside

- **Aspirin 300 mg chewed**, plus a second antiplatelet agent — ticagrelor, prasugrel or clopidogrel — according to the reperfusion pathway.
- **Anticoagulation** as specified by the local pathway.
- **Oxygen only if the saturation is below 90%.** Routine oxygen in the non-hypoxic patient confers no benefit and may cause harm.
- **Nitrate** sublingually or by infusion for pain — **withheld if the patient is hypotensive, has a right ventricular infarct, or has taken a phosphodiesterase inhibitor within 48 hours.**
- **Morphine** with an antiemetic for pain not relieved by nitrate.
- **Continuous cardiac monitoring beside a defibrillator.** Ventricular fibrillation is the commonest cause of death in the first hour and is entirely treatable if witnessed and monitored.

Reperfusion — the decision that determines outcome

Strategy	When, and the target
Primary percutaneous coronary intervention	Preferred whenever it can be delivered within 120 minutes of first medical contact. Target time from arrival at a capable centre to balloon inflation is under 60–90 minutes.
Fibrinolysis	When primary angioplasty cannot be delivered within that window. Give within 30 minutes of arrival, then transfer for angiography within 2–24 hours — or immediately if reperfusion fails.
Timing from onset	Benefit is greatest in the first 2–3 hours and persists to 12 hours. Beyond 12 hours, reperfusion is still indicated if symptoms or ST elevation continue, or the patient is unstable.

- **Absolute contraindications to fibrinolysis:** any previous intracranial haemorrhage; ischaemic stroke within 6 months; cerebral vascular malformation or tumour; major trauma, surgery or head injury within 3 weeks; gastrointestinal bleeding within a month; known bleeding disorder; **suspected aortic dissection**; non-compressible puncture within 24 hours.
- **Right ventricular infarction** — the patient is preload-dependent. **Give fluid, and avoid nitrates, diuretics and opioids**, all of which can precipitate profound hypotension.

Complications to anticipate

Timing	Complication
First hours	Ventricular fibrillation and ventricular tachycardia; bradycardia and atrioventricular block, particularly in inferior infarction; cardiogenic shock.
Days 2–5	Heart failure, atrial fibrillation, pericarditis, mural thrombus.
Days 3–7	Mechanical complications — papillary muscle rupture with acute mitral regurgitation, ventricular septal rupture, free wall rupture with tamponade. Each presents as sudden deterioration with a new murmur and each is a surgical emergency.
Weeks to months	Left ventricular aneurysm, Dressler syndrome, heart failure, ventricular arrhythmia.

Secondary prevention — started before discharge, not at follow-up

- **Dual antiplatelet therapy** for 12 months, then aspirin indefinitely.
- **High-intensity statin**, regardless of the baseline cholesterol.
- **Beta blocker** and **angiotensin-converting enzyme inhibitor**, both started early and titrated.
- **Mineralocorticoid receptor antagonist** if the ejection fraction is 40% or less with heart failure or diabetes.
- **Cardiac rehabilitation** — as effective as several of the drugs and referred to far less often. **Smoking cessation** with pharmacological support. Glycaemic and blood pressure control.
- Echocardiography before discharge; reassessment of the ejection fraction at 6–12 weeks for consideration of an implantable defibrillator if it remains at 35% or less.
- Practical advice the patient will actually ask about: driving restrictions, return to work, and resumption of sexual activity.

6. Teaching Points and Viva Questions

- Electrocardiogram within ten minutes. Time is myocardium, and the clock starts at symptom onset, not at arrival.
- Reciprocal ST depression supports a true infarction and helps distinguish it from pericarditis.
- Every inferior infarct gets right-sided leads before anyone gives a nitrate.

- A new murmur three days after infarction is a mechanical complication until proved otherwise, and needs a surgeon, not a diuretic.
- Check both arms once, at the start. Thrombolysing a dissection is among the worst errors in medicine.

Questions you should be able to answer:

- Localise this infarct and name the artery.
- Primary angioplasty is 3 hours away. What do you do and why?
- The blood pressure falls to 80/50 after a nitrate in an inferior infarct. Explain what happened.
- Distinguish the electrocardiogram of pericarditis from that of infarction.
- Name the four drugs he should leave hospital taking, and the indication for each.

Case 12 · Acute Decompensated Heart Failure

CLINICAL VIGNETTE

A **72-year-old woman** with known heart failure and a previous anterior myocardial infarction presents with four days of worsening breathlessness. She now sleeps upright in a chair and wakes at night gasping. Her ankles are swollen and she has gained 5 kg in ten days.

Two weeks ago she began taking a non-steroidal anti-inflammatory drug bought over the counter for knee pain, and she reduced her own diuretic dose because she disliked the trips to the bathroom.

On examination: she is sitting forward and speaking in short sentences. Heart rate 112/min and irregularly irregular, blood pressure 156/94 mmHg, respiratory rate 28/min, oxygen saturation 89% on air. The jugular venous pressure is 8 cm above the sternal angle. The apex is displaced to the anterior axillary line. There is a **third heart sound** and a pansystolic murmur at the apex. Crackles to the mid-zones bilaterally, and pitting oedema to the thighs.

1. Focused History

- **Quantify the change:** pillow count, distance walked, and the weight gain — which is the most sensitive marker of fluid retention and the easiest for the patient to measure at home.
- **Then find the precipitant.** Decompensation always has a cause, and treating the congestion without finding the cause simply returns the patient to hospital.

Precipitant	What to ask or check
Non-adherence, or self-adjusted diuretic	The commonest cause of all. Ask non-judgementally what she is actually taking, not what is on the list.
New arrhythmia, usually atrial fibrillation	Palpitations, an irregular pulse, and the electrocardiogram.
Ischaemia or infarction	Chest pain — which may be absent, particularly in diabetes and in the elderly.
Infection	Fever, cough, dysuria.
Drugs that cause fluid retention or depress the myocardium	Non-steroidal anti-inflammatory drugs, corticosteroids, verapamil and diltiazem, thiazolidinediones, some chemotherapy.
Excess salt or fluid intake	Diet, and the sudden weight rise.
High-output states	Anaemia, thyrotoxicosis, pregnancy, arteriovenous fistula.
Worsening renal function, uncontrolled hypertension, pulmonary embolism	Recent blood results and blood pressure readings.

2. Physical Examination

- **The jugular venous pressure is the single most reliable bedside sign of congestion** and the one most often not attempted. Measure it, record it in centimetres, and repeat it daily.
- Displaced, thrusting apex; a **third heart sound** — the most specific auscultatory sign of decompensation; a functional pansystolic murmur of mitral regurgitation from ventricular dilatation.
- Bibasal crackles that do not clear with coughing, and dullness at the bases from pleural effusions.
- Tender pulsatile hepatomegaly, ascites, and pitting oedema — ankles in the ambulant patient, **sacrum in the bedbound patient**, which is why you must roll them.

- **Assess perfusion as well as congestion:** cool peripheries, narrow pulse pressure, pulsus alternans, confusion and oliguria all indicate low output.

Classify the patient in one of four boxes. It takes seconds and directs treatment better than any score.

	Dry — no congestion	Wet — congested
Warm — adequately perfused	Compensated. Optimise oral therapy.	Warm and wet — the commonest presentation. Diuresis, with vasodilators if hypertensive.
Cold — poorly perfused	Underfilled. Reconsider the diagnosis; consider cautious fluid.	Cold and wet — the dangerous group. Requires inotropic support and critical care input.

3. Differential Diagnosis

Diagnosis	The feature that discriminates it
Pneumonia	Fever, focal consolidation, raised inflammatory markers, and a normal natriuretic peptide.
Exacerbation of chronic obstructive pulmonary disease	Wheeze dominates, sputum purulence, hyperinflated film, and a smoking history.
Pulmonary embolism	Sudden onset, pleuritic pain, hypoxia with a clear film, risk factors.
Renal failure with fluid overload	The same congestion, but with a very high creatinine — and it responds to dialysis, not to more diuretic.
Cirrhosis or nephrotic syndrome	Oedema and effusions with a normal or low jugular venous pressure, plus the stigmata of the underlying disease.

4. Investigations

- **Natriuretic peptide.** Its greatest value is a **negative** result: a normal level in an untreated breathless patient essentially excludes heart failure as the cause.
- **Electrocardiogram** — atrial fibrillation, ischaemia, prior infarction, left ventricular hypertrophy, and QRS duration for later resynchronisation decisions. A completely normal tracing makes heart failure unlikely.
- **Chest radiograph — reading the film:** cardiomegaly with a cardiothoracic ratio above 0.5, upper lobe blood diversion, **Kerley B lines**, perihilar alveolar shadowing in a bat's-wing distribution, pleural effusions, and fluid tracking into the horizontal fissure.
- **Bloods:** complete blood count (anaemia both causes and results from heart failure), urea, electrolytes and creatinine before and during diuresis, liver function (congestive hepatopathy), thyroid function, glucose, troponin, and **iron studies** — iron deficiency is common, symptomatic and treatable even without anaemia.
- **Echocardiography** — essential in every new case, and in decompensation of unclear cause. It gives the ejection fraction, which divides management into two entirely different pathways, and identifies valve disease and regional wall motion abnormality.

5. Management

The first hour

- Sit the patient upright. Oxygen only if hypoxic, targeting 94–98%.
- **Intravenous loop diuretic** — furosemide 40–80 mg, or one to two and a half times the usual home dose in a patient already taking it. Give it intravenously; gut absorption is unreliable in a congested patient.
- **Nitrate infusion** if the patient is hypertensive with pulmonary oedema — it reduces preload and afterload rapidly and is often more immediately effective than the diuretic.
- **Continuous positive airway pressure** for severe pulmonary oedema with persisting hypoxia or exhaustion. It reduces the need for intubation.
- **Inotropes and vasopressors only for hypoperfusion**, in a monitored setting with senior involvement. They are not a treatment for breathlessness.
- **Treat the precipitant:** rate control the atrial fibrillation, stop the non-steroidal anti-inflammatory drug, treat infection or ischaemia.

Monitoring

- **Daily weight before breakfast** is the best single measure of response — aim for 0.5–1 kg loss per day. Strict fluid balance, daily electrolytes and creatinine, and a repeated jugular venous pressure.
- Expect the creatinine to rise modestly during effective decongestion. A small rise is not a reason to stop diuresis in a patient who is still congested.

THE FOUR PILLARS OF CHRONIC THERAPY

These four drug classes each reduce mortality independently, and all four should be started and titrated in every patient with a reduced ejection fraction who tolerates them:

1. An angiotensin receptor–neprilysin inhibitor, or an angiotensin-converting enzyme inhibitor · **2.** A beta blocker · **3.** A mineralocorticoid receptor antagonist · **4.** A sodium–glucose co-transporter 2 inhibitor.

Diuretics relieve symptoms but **do not prolong life** — they are added on top for congestion, not instead. Do not start a beta blocker during acute decompensation, but do not stop an established one unless the patient is in shock.

For preserved ejection fraction: diuretics for congestion, a sodium–glucose co-transporter 2 inhibitor, and rigorous treatment of hypertension, atrial fibrillation, obesity and sleep apnoea.

Before discharge

- Euvolaemic, on oral diuretic for at least 24 hours, with a written **daily weight chart and an action plan** telling her what to do if she gains 2 kg in three days.
- A titration plan for the four pillars, a documented medication review, and an explicit instruction to avoid non-steroidal anti-inflammatory drugs — with an alternative offered for her knee.
- **Follow-up within seven days**, which measurably reduces readmission. Vaccination, cardiac rehabilitation, and consideration of devices at reassessment.

6. Teaching Points and Viva Questions

- The jugular venous pressure and the daily weight will tell you more about this patient than any blood test.
- A normal natriuretic peptide in an untreated breathless patient sends you looking for another diagnosis.
- Always name the precipitant. "Heart failure" is not a complete diagnosis.
- Diuretics treat symptoms; the four pillars treat the disease.
- Oedema with a normal venous pressure is not heart failure — look at the liver, the kidneys and the albumin.

Questions you should be able to answer:

- Name the two precipitants in this history and explain the mechanism of each.
- Why give the diuretic intravenously rather than orally?
- Her creatinine rises from 90 to 120 during diuresis. What do you do?
- Which of her drugs prolong life and which only relieve symptoms?
- How does management differ if the ejection fraction is 55% rather than 30%?

Case 13 · Infective Endocarditis

CLINICAL VIGNETTE

A **42-year-old man** with known rheumatic mitral valve disease presents with five weeks of fever, drenching night sweats, malaise and 7 kg of weight loss. He has been treated twice by his general practitioner with short courses of antibiotics, each of which improved things briefly. He had two teeth extracted seven weeks ago.

On examination: temperature 38.2 °C, pallor, heart rate 96/min. There are **splinter haemorrhages** in two fingernails and tender nodules on the pulps of two fingers. There is a pansystolic murmur at the apex which the notes record as previously soft and which is now loud. The spleen is palpable 3 cm below the costal margin. Urine dipstick shows blood ++ without protein.

1. Focused History

- **Duration and tempo.** A subacute course over weeks with sweats and weight loss suggests a viridans streptococcus on a damaged valve; a fulminant course over days suggests Staphylococcus aureus on a normal one.
- **The predisposing lesion:** rheumatic valve disease, prosthetic valve, previous endocarditis, congenital heart disease, degenerative valve disease, hypertrophic cardiomyopathy.
- **The portal of entry:** dental work and dental hygiene, skin infection, intravenous drug use, indwelling vascular catheters, haemodialysis, recent urological or gastrointestinal procedures, colorectal disease (which is associated with *Streptococcus gallolyticus* and warrants colonoscopy).
- **Regionally important exposures:** unpasteurised milk and dairy, and contact with sheep, goats or camels — **Brucella** and **Coxiella burnetii** are important causes of culture-negative endocarditis here and will be missed unless specifically requested.
- **Recent antibiotics** — the commonest reason blood cultures are sterile, and the reason this man has had five weeks of illness without a diagnosis.
- **Embolic symptoms:** transient neurological events, sudden limb pain, flank or left upper quadrant pain, sudden visual loss, and back pain suggesting spondylodiscitis.

2. Physical Examination

- **Fever and a murmur together is endocarditis until proved otherwise.** A **new or changed** murmur is the finding that matters; a longstanding unchanged murmur is much weaker evidence.
- **Peripheral stigmata** — uncommon now but classical: splinter haemorrhages; **Osler nodes**, which are painful and on the finger pulps; **Janeway lesions**, which are painless and on palms and soles; Roth spots on the retina; conjunctival and palatal petechiae; clubbing in longstanding disease.
- **Splenomegaly**, signs of heart failure, and a full neurological examination for embolic deficits.
- Examine the **teeth and gums**, injection sites, and any indwelling line.
- **Dipstick the urine** — microscopic haematuria from immune complex glomerulonephritis or renal infarction is a minor criterion and takes thirty seconds.

3. Investigations

THE INVESTIGATION THAT DECIDES EVERYTHING

Three sets of blood cultures from three separate venepuncture sites, before any antibiotic is given. In a subacute presentation, space them over at least an hour — there is time, and the diagnosis depends on it.

The bacteraemia of endocarditis is continuous, so cultures need not be timed to fever spikes. If the patient has already received antibiotics and is stable, **it is correct to stop them and re-culture after several days** rather than treat blindly for six weeks without an organism.

- **Serology for culture-negative causes:** Brucella, Coxiella burnetii, Bartonella, Legionella, and fungal cultures. Request these early in this region rather than as a last resort.
- **Echocardiography.** Transthoracic first; **transoesophageal** if that is negative and suspicion persists, if there is a prosthetic valve, or if a complication is suspected. Look for vegetations, abscess, new valve regurgitation and prosthetic dehiscence. **A negative study does not exclude the diagnosis** — repeat it in 5–7 days if suspicion remains.
- **Electrocardiogram, repeated daily. A new atrioventricular block means an aortic root abscess** and is an indication for urgent surgical assessment. This is the single most important electrocardiographic finding in the disease.
- Complete blood count (normocytic anaemia, neutrophilia), C-reactive protein and erythrocyte sedimentation rate for trend, urea and electrolytes, liver function, urinalysis and microscopy, rheumatoid factor and complement.
- Imaging for emboli where clinically indicated: brain, abdomen, and spine. Computed tomography and nuclear imaging have a defined role in prosthetic valve disease.

The modified Duke criteria

Major criteria	Minor criteria
Typical organism from two separate blood cultures, or persistently positive cultures	A predisposing cardiac lesion, or intravenous drug use
Positive Coxiella burnetii serology	Fever of 38 °C or above
Echocardiographic evidence: vegetation, abscess, or new prosthetic dehiscence	Vascular phenomena — emboli, mycotic aneurysm, intracranial or conjunctival haemorrhage, Janeway lesions
New valvular regurgitation	Immunological phenomena — glomerulonephritis, Osler nodes, Roth spots, rheumatoid factor
	Microbiological evidence not meeting a major criterion

Definite endocarditis: two major, or one major and three minor, or five minor criteria.

4. Management

- **Treatment is by a team** — cardiology, microbiology or infectious diseases, and cardiac surgery involved from the outset, not after complications appear.
- **Empirical therapy after cultures are taken**, guided by whether the valve is native or prosthetic and by local resistance patterns; then narrowed to the organism. **Intravenous therapy for four to six weeks** in

most cases.

- Monitor for response: daily examination for a new murmur or heart failure, daily electrocardiogram for conduction change, repeat cultures to confirm clearance, and monitoring of renal function and antibiotic levels.

Indication for surgery	Detail
Heart failure	Severe regurgitation or valve obstruction causing failure — the commonest and most urgent indication.
Uncontrolled infection	Abscess, false aneurysm, fistula, enlarging vegetation, persisting fever and positive cultures beyond 7–10 days of appropriate therapy, or a fungal or highly resistant organism.
Prevention of embolism	A vegetation greater than 10 mm with an embolic event despite antibiotics, or a very large vegetation greater than 15 mm.

Prevention

- **Dental hygiene and regular dental review** in every patient with a predisposing lesion — of far greater value than antibiotic prophylaxis.
- Antibiotic prophylaxis before dental procedures involving gingival manipulation is now restricted to the **highest-risk group**: prosthetic valves, previous endocarditis, and certain congenital heart disease.

5. Teaching Points and Viva Questions

- Fever plus a murmur means blood cultures before antibiotics. The single commonest reason this diagnosis is delayed is a well-meant prescription.
- A new atrioventricular block is an aortic root abscess until proved otherwise — do a daily electrocardiogram.
- A normal transthoracic echocardiogram does not exclude endocarditis.
- In this region, culture-negative endocarditis should prompt Brucella and Q fever serology early rather than late.
- Half of the peripheral stigmata are named after people; none of them is as useful as three sets of blood cultures.

Questions you should be able to answer:

- He has already had antibiotics. How does that change your approach to the cultures?
- Apply the Duke criteria to this patient and state your level of certainty.
- The PR interval lengthens on day four. What has happened and what do you do?
- List the three broad indications for surgery.
- Which patients still receive antibiotic prophylaxis before dental work, and why was the guidance narrowed?

Case 14 • Newly Diagnosed Hypertension

CLINICAL VIGNETTE

A **46-year-old man** is found to have a blood pressure of 168/104 mmHg at a workplace screening. He feels entirely well. In clinic a week later, after five minutes seated, it is 172/106 mmHg in the right arm and 168/104 mmHg in the left.

He works long hours at a desk, eats most meals outside the home, and does no regular exercise. He smokes 15 cigarettes a day. His father had a stroke at 58. His wife reports that he snores heavily and sometimes seems to stop breathing at night, and he falls asleep in the afternoon.

On examination: body mass index 33 kg/m^2, waist circumference increased. Pulse 78/min regular. No radio-femoral delay. Apex beat is forceful but undisplaced. No bruits. Fundoscopy shows arteriolar narrowing and arteriovenous nipping.

1. Focused History

Establish that it is real, and how long it has been present

- Previous readings — at dental visits, pre-employment checks, pharmacies, or during a previous illness. Patients often have years of unrecognised readings.
- Any home readings, and how the device was used.

Screen for end-organ damage

- Chest pain, exertional breathlessness, orthopnoea, palpitations, transient neurological symptoms, visual disturbance, claudication, and frothy urine or nocturia.

Screen for a secondary cause

Ask about	Cause it suggests
Snoring with witnessed apnoeas, daytime somnolence, morning headache	Obstructive sleep apnoea — the commonest secondary cause, and present in this patient
Muscle weakness, cramps, polyuria, unprovoked hypokalaemia	Primary aldosteronism — far commoner than traditionally taught
Episodic headache with palpitations and sweating, labile pressure	Phaeochromocytoma
Weight gain with thin skin, bruising, proximal weakness, striae	Cushing syndrome
Haematuria, oedema, known renal disease, family history of polycystic kidneys	Renal parenchymal disease
Young onset, abrupt onset, resistant hypertension, flash pulmonary oedema, renal bruit	Renovascular disease
Young patient with leg claudication or radio-femoral delay	Coarctation of the aorta
Drugs: non-steroidal anti-inflammatory drugs, combined oral contraceptive, corticosteroids, decongestants, ciclosporin, erythropoietin, liquorice, cocaine, amphetamines, some herbal preparations	Drug-induced hypertension — always ask, including over-the-counter and traditional remedies

Quantify total cardiovascular risk

- Smoking, diabetes, lipids, family history of premature vascular disease, alcohol, salt intake, physical activity, and body weight. **You are treating the total risk, not the number.**

2. Physical Examination

HOW TO MEASURE THE BLOOD PRESSURE

Everything downstream depends on this being done properly.

Seated and rested for **five minutes**, back supported, legs uncrossed, arm supported at heart level, no talking. A **cuff bladder encircling at least 80% of the arm** — a cuff that is too small is the commonest cause of a falsely high reading in a large arm. **Both arms at the first visit**, then use the higher-reading arm thereafter. Repeat and take the average. A **standing reading** after one and three minutes in the elderly and in diabetes.

- Body mass index and waist circumference; features of Cushing syndrome; **radio-femoral delay**; renal, carotid and femoral bruits; palpable kidneys.
- **Apex beat character** — a sustained, heaving apex suggests left ventricular hypertrophy. Listen for a fourth heart sound.
- **Fundoscopy.** Grade the retinopathy: arteriolar narrowing, arteriovenous nipping, then haemorrhages and exudates, and finally papilloedema — which changes the diagnosis to a hypertensive emergency and the timescale from months to hours.
- Peripheral pulses and a full neurological examination for evidence of previous events.

3. Investigations

- **Confirm the diagnosis out of the office** with ambulatory monitoring, or home readings twice daily for a week. This detects **white-coat hypertension**, which would otherwise commit a well man to lifelong drugs, and **masked hypertension**, which would otherwise be missed entirely. Out-of-office thresholds are lower than clinic thresholds.
- **End-organ assessment:** urinalysis and urine albumin-to-creatinine ratio, urea, electrolytes, creatinine and estimated glomerular filtration rate, and an **electrocardiogram** for left ventricular hypertrophy. Echocardiography if the tracing is abnormal or heart failure is suspected.
- **Cardiovascular risk profile:** fasting glucose or glycated haemoglobin, and a lipid profile.
- **Investigate for a secondary cause** if the patient is under 40, has resistant or abruptly worsening hypertension, has unprovoked hypokalaemia, or has suggestive features. Start with the **aldosterone-to-renin ratio**, plasma or urinary metanephrines, renal imaging, and — in this patient — a **sleep study**.

Grade	Clinic blood pressure
Grade 1	140–159 / 90–99 mmHg
Grade 2	160–179 / 100–109 mmHg
Grade 3	180 mmHg or more / 110 mmHg or more

Be aware that guidelines differ: some define hypertension from 130/80 mmHg while others retain 140/90 mmHg. Know which your institution follows and say so when asked — the examiner is testing whether you know that a threshold is a convention, not a fact of nature.

4. Management

Lifestyle — offered to everyone, and worth more than students assume

- Weight reduction; **salt below 5 g per day**; a diet rich in fruit, vegetables and low-fat dairy; regular aerobic activity of at least 150 minutes weekly; alcohol reduction; and smoking cessation — which does not lower blood pressure but reduces cardiovascular risk more than most drugs.
- Treating the obstructive sleep apnoea in this man may improve his blood pressure and will certainly improve his daytime function.

Drug treatment

- **When to start:** immediately in grade 2 and grade 3; in grade 1 when there is established cardiovascular disease, end-organ damage, diabetes, chronic kidney disease, or high estimated risk. This patient has grade 2 hypertension with retinopathy and starts today.
- **Start with two drugs, ideally in a single combination tablet.** Combination therapy from the outset achieves control faster than sequential single agents and improves adherence.
- **First-line combinations:** an angiotensin-converting enzyme inhibitor or angiotensin receptor blocker, plus either a calcium channel blocker or a thiazide-like diuretic.
- In patients of Black African or Caribbean origin, begin with a calcium channel blocker or thiazide-like diuretic rather than a renin–angiotensin blocker.
- **Beta blockers are not first line** for uncomplicated hypertension; reserve them for angina, previous infarction, heart failure, rate control, or pregnancy.
- In pregnancy: labetalol, nifedipine or methyldopa. **Renin–angiotensin system blockers are absolutely contraindicated.**

Targets and follow-up

- Aim below 140/90 mmHg for everyone, and below 130/80 mmHg if tolerated, particularly with diabetes, chronic kidney disease or established vascular disease. Be less aggressive in the frail elderly, and check for postural drop before intensifying.
- Recheck urea, electrolytes and creatinine **one to two weeks after starting** a renin–angiotensin blocker or diuretic. A creatinine rise beyond about 30% or a falling potassium prompts review.
- Review monthly until controlled, then at least annually with reassessment of risk and end-organ damage.

RESISTANT HYPERTENSION

Blood pressure above target on **three drugs including a diuretic at optimal doses**. Before adding a fourth, check in this order:

Adherence — by far the commonest explanation, and best explored without accusation · **measurement technique and cuff size** · **white-coat effect**, confirmed with ambulatory monitoring · **salt intake, alcohol, and interfering drugs** · **an undiagnosed secondary cause**, especially primary aldosteronism and sleep apnoea.

Once those are excluded, **spironolactone** is the fourth agent of choice.

5. Teaching Points and Viva Questions

- Do not diagnose lifelong hypertension on clinic readings alone. Confirm it outside the clinic.
- Measure it properly. Wrong cuff, unsupported arm and no rest period produce a wrong number, and everything that follows is built on it.
- Both arms once, at the first visit. Thereafter, the higher arm.
- Unprovoked hypokalaemia in a hypertensive patient means primary aldosteronism until proved otherwise.
- Snoring with daytime somnolence is not a lifestyle detail — it is a diagnosis with a treatment.
- You are treating total cardiovascular risk. Stopping the cigarettes will help this man more than any milligram adjustment.

Questions you should be able to answer:

- How would you confirm the diagnosis before committing him to treatment?
- Which secondary cause does this history point to, and how would you test for it?
- What would you start him on today, and why two drugs rather than one?
- His potassium is 3.1 mmol/L and he takes no diuretic. What now?
- At review his pressure is 158/96 on three drugs. Work through your approach.

Case 15 · Aortic Stenosis

CLINICAL VIGNETTE

A **74-year-old man** describes eight months of breathlessness on exertion, now limiting him to about 50 metres on the flat. On three occasions he has felt lightheaded while walking uphill, and last week he **lost consciousness briefly while climbing stairs**, recovering within seconds. He also describes a tightness across the chest on exertion that settles with rest.

On examination: the pulse is **slow-rising and of small volume**. Blood pressure 112/92 mmHg. The apex beat is **heaving but undisplaced**. There is a systolic thrill in the right second intercostal space. **The second heart sound is barely audible.** There is a fourth heart sound and a **harsh, late-peaking ejection systolic murmur radiating to both carotids**.

Electrocardiogram: left ventricular hypertrophy with repolarisation change. **Echocardiography:** peak aortic velocity 4.4 m/s, mean gradient 52 mmHg, valve area 0.7 cm^2, ejection fraction 55%.

WHY THE HISTORY MATTERS MORE THAN THE NUMBERS

Exertional syncope, exertional angina and exertional breathlessness — each of the three is a marker of severe disease, and each arises because a fixed valve cannot increase cardiac output on demand.

The onset of symptoms transforms the prognosis. Asymptomatic severe aortic stenosis is compatible with years of normal life; **untreated symptomatic severe aortic stenosis carries a mortality of roughly 50% at two years.** The symptom, not the gradient, is what triggers referral for intervention.

Which is why the single most important thing you will say to a patient under surveillance is: **tell us the day you become breathless, dizzy or develop chest pain on exertion.**

1. Focused History

- **Characterise the triad**, and quantify the exercise tolerance in metres or flights of stairs against the patient's own baseline.
- **Syncope**: establish that it was exertional, sudden, without warning, and with rapid complete recovery — the pattern of cardiac syncope.
- **Heart failure symptoms:** orthopnoea, paroxysmal nocturnal dyspnoea, oedema.
- **Gastrointestinal bleeding** — the association of aortic stenosis with colonic angiodysplasia and acquired von Willebrand abnormality (**Heyde syndrome**) is real and worth asking about in an anaemic patient with a murmur.
- **Establish the aetiology:** age and calcific degeneration in the elderly; **a bicuspid valve** in a younger patient, which may be familial and is associated with aortic root dilatation and coarctation; and **previous rheumatic fever**, which usually affects the mitral valve too.
- **Comorbidity, frailty and function** — because the whole management decision is which intervention, not whether to intervene, and that depends on the patient rather than the valve.

2. Physical Examination

Sign	Mechanism
Slow-rising, small-volume pulse	Prolonged, obstructed ejection. May be misleadingly normal in the elderly, whose stiff arteries transmit a sharper upstroke
Narrow pulse pressure	Reduced stroke volume
Heaving, undisplaced apex	Pressure-loaded, concentrically hypertrophied but not dilated ventricle
Soft or absent second heart sound	The calcified valve cannot close audibly. This is one of the most reliable bedside markers of severity
Fourth heart sound	Atrial contraction against a stiff hypertrophied ventricle
Late-peaking ejection systolic murmur radiating to the carotids	Turbulent flow across the stenosis; the later the peak, the tighter the valve
Reversed splitting of the second sound	Delayed aortic valve closure

BEDSIDE MARKERS OF SEVERITY

Soft or absent second heart sound · a late-peaking murmur · a slow-rising pulse · a narrow pulse pressure · a fourth heart sound · signs of heart failure.

The loudness of the murmur is not one of them. A very tight valve with a failing ventricle generates little flow and therefore little noise, so the quietest murmurs can accompany the most severe disease.

3. Differential Diagnosis

Diagnosis	The feature that discriminates it
Hypertrophic cardiomyopathy	The murmur does not radiate to the carotids, the pulse is jerky rather than slow-rising, and the murmur is louder on standing and Valsalva and softer on squatting — the reverse of aortic stenosis. See Case 18.
Aortic sclerosis	An ejection systolic murmur with a normal pulse character, normal second sound and no significant gradient. Common in the elderly and not obstructive.
Mitral regurgitation	Pansystolic rather than ejection systolic, radiating to the axilla, with a displaced thrusting apex.
Pulmonary stenosis	Loudest at the left sternal edge and louder on inspiration, with a right ventricular impulse.
Ventricular septal defect	Harsh pansystolic murmur at the left sternal edge with a thrill.

4. Investigations

- **Electrocardiogram:** left ventricular hypertrophy with strain, left bundle branch block, and atrial fibrillation in late disease.
- **Chest radiograph:** often unremarkable. Look for a **calcified valve**, post-stenotic dilatation of the ascending aorta, and cardiomegaly only when the ventricle has failed.
- **Echocardiography is the diagnostic and grading investigation.**

Severe aortic stenosis — any of	Value
Peak aortic velocity	4.0 m/s or above
Mean transvalvular gradient	40 mmHg or above
Valve area	1.0 cm^2 or below (or 0.6 cm^2/m^2 indexed)

- **The trap of low-flow, low-gradient stenosis:** a failing ventricle cannot generate a high gradient across even a very tight valve, so the gradient understates the severity. Where the valve area is small but the gradient low and the ejection fraction reduced, **dobutamine stress echocardiography** distinguishes true severe stenosis from pseudo-severe disease.

- **Before intervention:** coronary angiography or computed tomographic coronary imaging; **computed tomography for transcatheter planning** — annulus size, valve calcification, coronary heights and vascular access; natriuretic peptide; dental assessment; and a formal frailty assessment.

5. Management

WHAT MEDICINE CAN AND CANNOT DO

No drug alters the natural history of aortic stenosis. The treatment is to replace the valve.

Statins do not slow progression. Diuretics relieve congestion but do not help the obstruction.

And be careful with vasodilators. Nitrates, angiotensin-converting enzyme inhibitors and other vasodilators can cause profound hypotension and syncope in severe stenosis, because cardiac output is fixed and cannot rise to compensate for a fall in systemic resistance. Hypertension still needs treating — but cautiously, at low doses, with review.

Indications for intervention

- **Severe stenosis with any symptom** — this patient qualifies on all three counts.

- **Asymptomatic severe stenosis with:** an ejection fraction below 50%; an abnormal exercise test with symptoms or a fall in blood pressure; very severe stenosis with a velocity above 5 m/s; rapid progression; or a markedly raised natriuretic peptide.

- Severe stenosis in a patient undergoing cardiac surgery for another indication.

Which intervention

- **Surgical aortic valve replacement** or **transcatheter aortic valve implantation** — the decision belongs to a multidisciplinary heart team, weighing age, surgical risk, anatomy, frailty, comorbidity and life expectancy. Transcatheter implantation is now used across a wide spectrum of surgical risk, not only in inoperable patients.

- **Balloon valvuloplasty** has no lasting role in calcific adult disease; it is a bridge in extremis, or a treatment for non-calcific congenital stenosis in the young.

- **A mechanical prosthesis requires lifelong warfarin — not a direct oral anticoagulant**, which is contraindicated. A bioprosthesis does not require long-term anticoagulation but degenerates over time, which is the central trade-off to explain to a patient choosing between them.

Surveillance and prevention

- Echocardiographic follow-up at intervals determined by severity — annually in severe asymptomatic disease, less often in mild.
- **Dental hygiene and regular dental review**; antibiotic prophylaxis for the highest-risk group only, as in Case 13.
- **Educate the patient to report the first symptom immediately**, and record that you have done so.

6. Teaching Points and Viva Questions

- Exertional syncope in an older patient is aortic stenosis until proved otherwise.
- A soft second heart sound means severe disease; a loud murmur means nothing.
- Symptoms are the trigger for surgery, and untreated symptomatic severe stenosis kills half of patients within two years.
- Vasodilators in fixed obstruction cause syncope.
- A low gradient with a small valve area and a poor ventricle needs a stress study, not reassurance.
- Mechanical valves need warfarin, never a direct oral anticoagulant.

Questions you should be able to answer:

- Explain each of this man's physical signs in terms of the underlying haemodynamics.
- How would you distinguish this murmur from that of hypertrophic cardiomyopathy at the bedside?
- His valve area is 0.8 cm^2 but the mean gradient is only 25 mmHg and his ejection fraction is 35%. What is going on and what do you do?
- A colleague starts ramipril for his blood pressure of 150/95. What is your concern?
- He is asymptomatic with a velocity of 4.2 m/s and normal function. What do you do?

Case 16 · Chronic Regurgitant Valve Disease

CLINICAL VIGNETTE

A **42-year-old man**, known since childhood to have a **bicuspid aortic valve**, describes a year of gradually worsening breathlessness on exertion and an uncomfortable awareness of his own heartbeat, worst when lying on his left side. He has no chest pain and has never had rheumatic fever.

On examination: the pulse is **collapsing** in character. Blood pressure **160/48 mmHg**. Visible carotid pulsation is present, and the nail beds pulsate on gentle compression. The apex beat is **displaced to the anterior axillary line and thrusting**. There is a **soft early diastolic murmur at the left sternal edge, audible only with the patient sitting forward with the breath held in expiration**, and a separate low-pitched mid-diastolic murmur at the apex.

Echocardiography: severe aortic regurgitation; left ventricular end-diastolic diameter 68 mm; ejection fraction 48%; aortic root 48 mm.

THE GOVERNING PRINCIPLE OF THIS CASE

In stenosis, the patient tells you when to operate. **In regurgitation, the ventricle does.**

A regurgitant ventricle compensates by dilating, and it does so **silently for years**. By the time breathlessness appears, irreversible myocardial damage may already have occurred and the benefit of surgery is reduced.

So the timing of surgery in chronic regurgitation is decided by serial measurement of ventricular size and function on echocardiography, not by waiting for symptoms. This patient is already symptomatic with an ejection fraction of 48% and a markedly dilated ventricle — he has been under-surveilled.

1. Acute or Chronic? — Ask This First

	Chronic regurgitation	Acute severe regurgitation
Presentation	Years of compensation, then gradual breathlessness	Sudden pulmonary oedema and cardiogenic shock
Murmur	Loud and striking, with dramatic peripheral signs	Soft or absent — there is no time for a large gradient or for peripheral signs to develop
Heart size	Dilated	Normal — the ventricle has had no time to remodel
Causes	Bicuspid valve, rheumatic disease, aortic root dilatation, degenerative disease, mitral valve prolapse	Infective endocarditis · aortic dissection · papillary muscle or chordal rupture after infarction · trauma
Management	Surveillance, then elective surgery	Surgical emergency

The quiet murmur with a normal-sized heart in a shocked patient is the dangerous one. Students expect severe valve disease to be loud, and acute severe regurgitation is therefore missed.

2. Focused History

- Exertional breathlessness, orthopnoea, and **awareness of the heartbeat**, which reflects the large stroke volume of chronic aortic regurgitation.
- **Angina** can occur in aortic regurgitation without coronary disease, because a low diastolic pressure reduces coronary perfusion.
- **Aetiology:** bicuspid valve and its family history; rheumatic fever; **connective tissue disease — Marfan and Ehlers–Danlos syndromes**, with a family history of aortic dissection or sudden death; ankylosing spondylitis; hypertension; previous endocarditis; and recent dental or invasive procedures.

- For **mitral regurgitation:** previous rheumatic fever, mitral valve prolapse, previous myocardial infarction, and features of a dilated ventricle causing secondary regurgitation.

3. Physical Examination

Aortic regurgitation — and the eponyms, each with a mechanism

All of these arise from **one thing: a very large stroke volume ejected into a low-resistance circulation, giving a wide pulse pressure**. Learn the mechanism and the list becomes trivial.

Sign	What it is
Collapsing (water-hammer) pulse	Rapid upstroke and rapid collapse; best felt with the arm raised
Wide pulse pressure	High systolic, low diastolic — here 160/48
Corrigan sign	Visible vigorous carotid pulsation
de Musset sign	Head nodding with each beat
Quincke sign	Capillary pulsation visible in the nail bed
Müller sign	Pulsation of the uvula
Duroziez sign	To-and-fro murmur over the femoral artery under light compression
Traube sign	"Pistol-shot" femoral sounds
Displaced thrusting apex	Volume-loaded, dilated ventricle
Early diastolic decrescendo murmur	At the left sternal edge, sitting forward, breath held in expiration — you will not hear it otherwise
Austin Flint murmur	A mid-diastolic rumble at the apex from the regurgitant jet striking the mitral valve. Do not mistake it for mitral stenosis — there is no loud first sound and no opening snap

Mitral regurgitation

- **Pansystolic murmur at the apex radiating to the axilla**, loudest in expiration in the left lateral position; a soft first heart sound; a third heart sound; a displaced thrusting apex; atrial fibrillation; and, in advanced disease, the signs of pulmonary hypertension and right heart failure.
- **Mitral valve prolapse** gives a **mid-systolic click with a late systolic murmur**, and the timing of the click moves with manoeuvres that alter ventricular volume.

4. Investigations

- **Electrocardiogram:** left ventricular hypertrophy; atrial fibrillation in mitral disease.
- **Chest radiograph:** cardiomegaly, and **dilatation of the aortic root and ascending aorta** in aortic regurgitation.
- **Echocardiography, performed serially, with the numbers recorded and compared.** Severity of regurgitation, **left ventricular end-diastolic and end-systolic dimensions, and ejection fraction** — these are the values on which the surgical decision rests. Transoesophageal study to define the mechanism of mitral regurgitation and the feasibility of repair.

- **Cardiac magnetic resonance imaging** for accurate ventricular volumes and regurgitant fraction where echocardiography is equivocal.
- **Computed tomographic or magnetic resonance imaging of the aorta** whenever the root is dilated, with surveillance thereafter.
- Exercise testing to unmask symptoms in a patient who claims to be asymptomatic; natriuretic peptide.

5. Management

Medical

- **Vasodilators help here, in contrast to aortic stenosis.** Treat hypertension with an angiotensin-converting enzyme inhibitor or receptor blocker or a dihydropyridine calcium channel blocker; reducing afterload reduces the regurgitant fraction.
- **Use beta blockers cautiously in aortic regurgitation** — slowing the heart lengthens diastole and therefore increases the time available for regurgitation. They are, however, indicated for aortopathy in Marfan syndrome, alongside an angiotensin receptor blocker and root surveillance.
- Treat heart failure and atrial fibrillation conventionally. **Medical therapy does not substitute for surgery in severe disease** — it manages symptoms while the decision is made.

Indications for surgery — learn these numbers

Lesion	Operate if
Severe aortic regurgitation	Symptoms; or ejection fraction 50% or below; or left ventricular end-diastolic diameter above about 70 mm or end-systolic above about 50 mm (or the indexed equivalents); or aortic root 55 mm or more — with lower thresholds in Marfan syndrome and bicuspid valve disease
Severe primary mitral regurgitation	Symptoms; or ejection fraction 60% or below; or left ventricular end-systolic diameter 40 mm or more; or new atrial fibrillation or pulmonary hypertension

- **In degenerative mitral disease, repair is strongly preferred to replacement** — it preserves ventricular function, avoids a prosthesis and anticoagulation, and has better long-term outcomes. Refer to a centre with a high repair rate.
- **Transcatheter edge-to-edge repair** is an option in selected patients with secondary mitral regurgitation and heart failure, or where surgical risk is prohibitive.
- Endocarditis prevention and dental care; secondary penicillin prophylaxis where the disease is rheumatic (Case 9).

6. Teaching Points and Viva Questions

- In regurgitation the ventricle tells you when to operate, not the patient.
- Every eponymous sign of aortic regurgitation comes from a wide pulse pressure.
- You will not hear the murmur unless the patient sits forward and holds the breath out.
- The Austin Flint murmur is not mitral stenosis.

- Acute severe regurgitation is quiet, has a normal-sized heart, and is a surgical emergency.
- Vasodilators help in regurgitation and harm in stenosis.
- Repair the mitral valve rather than replace it wherever possible.

Questions you should be able to answer:

- Explain his blood pressure of 160/48 and list four signs that follow from it.
- Why does he have a mid-diastolic murmur at the apex, and how do you know it is not mitral stenosis?
- He says he feels well enough. Give three findings that would still make you refer him for surgery.
- A patient with endocarditis becomes acutely breathless with a barely audible murmur and a normal chest radiograph. What has happened?
- Why is a beta blocker a mixed blessing in aortic regurgitation?

Case 17 · Pericardial Disease

CLINICAL VIGNETTE

A **34-year-old man** presents with four days of sharp central chest pain. **It is worse when he lies flat and on deep inspiration, and eases when he sits forward.** It radiates to his left shoulder and up the side of his neck. He had a coryzal illness a week earlier.

On examination: temperature 37.9 °C, heart rate 96/min, blood pressure 124/78 mmHg. There is a **scratchy sound at the left sternal edge**, best heard with the patient leaning forward, which comes and goes.

Electrocardiogram: widespread **concave (saddle-shaped) ST elevation with PR segment depression**, present in both anterior and inferior leads, with ST depression and PR elevation in lead aVR. No Q waves.

C-reactive protein 68 · troponin mildly raised · echocardiography: small pericardial effusion, normal ventricular function.

1. Four Syndromes, One Pericardium

- **Acute pericarditis** — inflammation, with pain and a rub.
- **Pericardial effusion** — fluid, which may be silent.
- **Cardiac tamponade** — fluid under enough pressure to impair filling. A **haemodynamic** diagnosis, not a volume one.
- **Constrictive pericarditis** — a thickened, scarred pericardium restricting filling chronically.

2. Acute Pericarditis

DIAGNOSTIC CRITERIA

Two of the following four:

Characteristic chest pain — sharp, pleuritic, **worse lying flat and relieved by sitting forward**, often radiating to the trapezius ridge, which is close to specific because the phrenic nerve supplies both.

A pericardial rub — scratchy, often triphasic, best at the left sternal edge with the patient leaning forward. It is **evanescent**, so its absence at one examination means little.

Electrocardiographic change — widespread ST elevation with PR depression.

A new or worsening pericardial effusion.

The electrocardiogram — how it differs from infarction

	Pericarditis	ST-elevation myocardial infarction
Distribution	Widespread, crossing territories	Territorial, matching one artery
Shape of ST elevation	Concave, saddle-shaped	Convex, tombstoned
PR segment	Depressed — and elevated in aVR	Normal
Reciprocal ST depression	Absent, except in aVR and V1	Present
Q waves	Absent	Develop
Evolution	ST normalises, then diffuse T inversion	Territorial, with Q wave formation

Causes

- **Viral and idiopathic** — the great majority in young patients.

- **Tuberculous pericarditis** — a leading cause worldwide and an important one in this region. Subacute onset, fever, weight loss, a large effusion, and a high risk of progression to constriction. **Consider it in every patient who is not straightforwardly viral.**
- Bacterial and purulent; **uraemic**; after myocardial infarction — early, or late as **Dressler syndrome**; after cardiac surgery or a percutaneous procedure; malignant infiltration; autoimmune disease including lupus and rheumatoid arthritis; radiotherapy; drugs; hypothyroidism; trauma.

Investigation

- Electrocardiogram, C-reactive protein and erythrocyte sedimentation rate, **troponin**, complete blood count, renal function, thyroid function, chest radiograph.
- **Echocardiography in every patient** — for effusion, for tamponade physiology, and for ventricular function.
- **A raised troponin indicates myopericarditis.** If ventricular function is also impaired, the diagnosis is perimyocarditi, and the patient should be admitted and monitored.
- **Tuberculosis workup** where the picture is subacute or the effusion large: interferon-gamma release assay, chest imaging, and pericardial fluid for adenosine deaminase, acid-fast staining and culture if drained. Autoimmune screen; blood cultures if febrile; cross-sectional imaging in complicated cases.

Treatment

THE REGIMEN

A non-steroidal anti-inflammatory drug — aspirin or ibuprofen — at anti-inflammatory dose with a taper, PLUS colchicine for three months.

Colchicine roughly halves the rate of recurrence and is the single most important addition to the treatment of acute pericarditis in recent decades. It is regularly omitted.

Corticosteroids increase the rate of recurrence and are not first-line. Reserve them for specific indications — autoimmune disease, uraemic pericarditis, pregnancy, or a genuine contraindication to anti-inflammatory drugs.

Restrict exercise until symptoms have resolved and markers normalised — for longer in athletes and in myopericarditis.

- **Admit** if there is fever above 38 °C, a subacute onset, a large effusion or tamponade, myopericarditis, immunosuppression, trauma, anticoagulation, or failure to respond to a week of treatment.
- **Treat the specific cause:** antituberculous therapy for tuberculous disease (with corticosteroids in selected cases to reduce constriction); drainage and antibiotics for purulent pericarditis; intensified dialysis for uraemic disease.
- **Recurrent pericarditis:** colchicine for longer, then corticosteroids, then **interleukin-1 antagonists**, which are highly effective in colchicine-resistant disease.

3. Cardiac Tamponade

EMERGENCY

Tamponade is a clinical diagnosis. The echocardiogram supports it; it does not make it, and waiting for one can kill the patient.

Beck triad: hypotension · raised jugular venous pressure · muffled heart sounds. Add **tachycardia** and **pulsus paradoxus greater than 10 mmHg**, and the diagnosis is made at the bedside.

Electrocardiogram: sinus tachycardia, low voltage, and sometimes **electrical alternans**. **Echocardiogram:** effusion with right atrial and right ventricular diastolic collapse, a dilated inferior vena cava, and respirophasic septal shift.

Management: urgent pericardiocentesis, echo-guided where possible, or surgical drainage.

Give fluid. Avoid diuretics, avoid vasodilators, and avoid positive pressure ventilation if you can — this is a preload-dependent state and every one of those reduces preload. The instinct to treat a raised venous pressure with a diuretic is the error to avoid.

4. Constrictive Pericarditis

- Presents as **right heart failure with preserved ventricular systolic function** — progressive oedema, ascites often out of proportion to peripheral oedema, hepatomegaly, fatigue and breathlessness.
- **Tuberculosis is a leading cause worldwide**, along with previous cardiac surgery, radiotherapy, and any cause of chronic pericarditis.
- Signs: raised venous pressure with a **prominent y descent**, **Kussmaul sign** (venous pressure rises on inspiration), a **pericardial knock**, atrial fibrillation, ascites and hepatomegaly.

	Tamponade	Constriction
Jugular venous waveform	Absent y descent	Prominent y descent
Kussmaul sign	Absent	Present
Pulsus paradoxus	Prominent	Usually absent or mild
Onset	Acute or subacute	Chronic, over months
Treatment	Drainage	Pericardiectomy

- **Distinguishing constriction from restrictive cardiomyopathy** is the hard part, and it matters because one is surgically curable: look for pericardial thickening or calcification on computed tomography, septal bounce and respirophasic variation in filling on echocardiography, and characteristic changes on cardiac magnetic resonance imaging. Natriuretic peptide is typically **lower** in constriction than in restrictive cardiomyopathy.
- **Treatment is pericardiectomy**, with diuretics for symptomatic relief in the interim and antituberculous therapy where relevant.

5. Teaching Points and Viva Questions

- Pain relieved by sitting forward and radiating to the trapezius ridge is pericardial.
- Widespread saddle ST elevation with PR depression and no reciprocal change is not an infarct.
- Colchicine for three months, for everybody. Steroids make recurrence more likely.

- A raised troponin means the myocardium is involved and changes the disposition.
- Tamponade is diagnosed at the bedside; give fluid, not furosemide.
- Absent y descent means tamponade; prominent y descent and Kussmaul sign mean constriction.
- Think of tuberculosis in any pericardial disease here that is not clearly viral.

Questions you should be able to answer:

- Give four ways this electrocardiogram differs from that of an anterior infarction.
- What will you prescribe, for how long, and what will you deliberately not prescribe?
- His troponin is raised. Does that change your management?
- Two days later his blood pressure is 84/60 with a raised venous pressure and quiet heart sounds. What is happening and what do you do?
- How would you distinguish constrictive pericarditis from restrictive cardiomyopathy?

Case 18 · The Cardiomyopathies

CLINICAL VIGNETTE

A **28-year-old man** is referred after **losing consciousness while playing football**. He has noticed breathlessness and chest tightness on hard exertion for a year and had assumed he was unfit. **His brother died suddenly at the age of 32, also while playing football.** His parents are first cousins.

On examination: the pulse is **jerky** — a brisk upstroke with a mid-systolic dip. Blood pressure 118/76 mmHg. The apex beat has a **double impulse**. There is a fourth heart sound and an **ejection systolic murmur at the left sternal edge that does not radiate to the carotids**. **The murmur becomes louder when he stands and on Valsalva, and quieter when he squats.**

Electrocardiogram: left ventricular hypertrophy with deep T wave inversion in the lateral leads. **Echocardiography:** asymmetric septal hypertrophy of 19 mm, systolic anterior motion of the mitral valve, and a resting left ventricular outflow tract gradient of 60 mmHg.

1. Three Phenotypes

	Dilated	Hypertrophic	Restrictive
Problem	Systolic failure of a dilated ventricle	Hypertrophy with dynamic outflow obstruction and arrhythmia	Impaired filling with a non-dilated ventricle and preserved systolic function
Causes	Idiopathic and genetic · alcohol · peripartum · myocarditis · anthracyclines and trastuzumab · iron overload, including transfused thalassaemia · thyroid disease · sustained tachycardia · sarcoidosis · cocaine · thiamine deficiency	Autosomal dominant sarcomeric protein mutations. The commonest cause of sudden cardiac death in young athletes	Amyloidosis · sarcoidosis · haemochromatosis · endomyocardial fibrosis · radiation · storage disorders
Treatment	The four pillars of heart failure therapy (Case 12), devices, and treating the cause	Beta blocker, avoidance of vasodilators, risk stratification for implantable defibrillator, septal reduction	Treat the cause — and amyloidosis is now treatable, so it must be actively sought

2. Hypertrophic Cardiomyopathy

THE MANOEUVRE THAT MAKES THE DIAGNOSIS

The murmur of hypertrophic cardiomyopathy gets louder when you reduce ventricular filling and quieter when you increase it.

Louder: standing, Valsalva manoeuvre, nitrates, dehydration, exercise — a smaller cavity brings the hypertrophied septum and the mitral valve closer together and worsens obstruction.

Quieter: squatting, passive leg raise, handgrip — all of which increase preload or afterload.

In aortic stenosis the reverse is true, because the obstruction is fixed and depends on flow. This one manoeuvre, performed at the bedside in fifteen seconds, separates the two diagnoses.

Focused history

- **Exertional syncope or presyncope** — the red flag, and the reason this patient was referred.
- Exertional breathlessness, chest pain, palpitations.
- **A detailed family history across three generations: sudden death, unexplained drowning or road death in a young relative, cardiomyopathy, defibrillators, and consanguinity.** This is a family disease,

and the consultation concerns everybody in it.

- Competitive and recreational sport, and the intensity of it.

Examination

- **Jerky pulse** with a rapid upstroke and mid-systolic decline; **double apical impulse**; fourth heart sound; **ejection systolic murmur at the left sternal edge that does not radiate to the carotids**, with the dynamic behaviour above; and often a pansystolic murmur of mitral regurgitation from systolic anterior motion of the valve.

Investigations

- **Electrocardiogram — abnormal in over 90%**: left ventricular hypertrophy, deep lateral T wave inversion, pathological Q waves, and **giant negative T waves in the apical variant**. A completely normal tracing makes the diagnosis unlikely.

- **Echocardiography:** asymmetric hypertrophy of 15 mm or more, or 13 mm with a family history; systolic anterior motion of the mitral valve; and the **outflow tract gradient measured at rest and with provocation**, since obstruction is dynamic and may be absent at rest.

- **Cardiac magnetic resonance imaging** for apical and unusual variants and to quantify **myocardial fibrosis**, which contributes to arrhythmic risk.

- **Ambulatory electrocardiographic monitoring** for non-sustained ventricular tachycardia, and **exercise testing with blood pressure response**.

- **Genetic testing, and cascade screening of first-degree relatives.**

Management

- **Avoid dehydration, vasodilators, nitrates and excessive diuresis** — all reduce preload and worsen obstruction. **Avoid digoxin and inotropes**, which increase contractility and therefore obstruction.

- **Beta blocker first line**; verapamil or disopyramide as alternatives or additions. **Cardiac myosin inhibitors such as mavacamten** are a targeted treatment for obstructive disease.

- **Septal reduction therapy** — surgical myectomy or alcohol septal ablation — for symptoms refractory to drugs.

- **Risk stratification for sudden death and consideration of an implantable defibrillator**, using a validated risk score. The features that raise risk: **family history of sudden death, unexplained syncope, massive hypertrophy, non-sustained ventricular tachycardia, apical aneurysm, extensive fibrosis on imaging, and an abnormal blood pressure response to exercise.** This patient has two of them.

- **Exercise advice, individualised** rather than blanket prohibition, though high-intensity competitive sport is generally discouraged in higher-risk patients.

THE MOST IMPORTANT THING YOU WILL DO IN THIS CONSULTATION

Every first-degree relative needs an electrocardiogram and an echocardiogram, repeated periodically through adolescence and early adulthood, because the phenotype develops with growth. Where a causative mutation is identified, **genetic cascade testing lets you discharge the relatives who did not inherit it** — which is as valuable as identifying those who did.

Given the consanguinity in this family, the pedigree should be drawn out properly and the extended family offered assessment. **His brother's death was probably preventable, and his cousins' may be.**

3. Dilated Cardiomyopathy

- Management is the management of heart failure with reduced ejection fraction — **the four pillars set out in Case 12**, with device therapy and, ultimately, transplantation.
- **But look for the reversible causes, because several are entirely correctable:** alcohol (abstinence can restore function), **iron overload from transfusion — relevant in thalassaemia, where chelation and T2* monitoring are the treatment**, thyroid disease, sustained tachyarrhythmia, thiamine deficiency, and cardiotoxic chemotherapy.
- **Peripartum cardiomyopathy** presents in late pregnancy or the early puerperium; function often recovers, and a subsequent pregnancy carries significant risk if it does not. Specialist joint care is required.

4. Restrictive Cardiomyopathy — and the Diagnosis Not to Miss

CARDIAC AMYLOIDOSIS

Cardiac amyloidosis is now treatable, which makes it a diagnosis worth chasing rather than a post-mortem finding.

Suspect it in heart failure with preserved ejection fraction plus any of: a **low-voltage electrocardiogram despite thickened walls** — a striking discordance · bilateral **carpal tunnel syndrome**, often years earlier · lumbar spinal stenosis · macroglossia · periorbital purpura · proteinuria · autonomic neuropathy · intolerance of standard heart failure drugs.

Investigate with serum free light chains and immunofixation (for the AL type), **bone scintigraphy** (for transthyretin amyloidosis, which it can diagnose non-invasively), cardiac magnetic resonance imaging, and biopsy where needed.

Treat: tafamidis and other stabilisers for transthyretin disease; chemotherapy for AL amyloidosis. **Use diuretics carefully, and expect beta blockers and angiotensin-converting enzyme inhibitors to be poorly tolerated.** Anticoagulate for atrial fibrillation at a low threshold.

5. Teaching Points and Viva Questions

- Stand the patient up. The murmur that gets louder is hypertrophic cardiomyopathy; the one that gets quieter is aortic stenosis.
- Exertional syncope in a young person with a family history of sudden death is an emergency referral, not a routine one.
- The electrocardiogram is abnormal in almost every case of hypertrophic cardiomyopathy.
- Avoid vasodilators, nitrates, digoxin and dehydration.
- Screen the family — that is where the lives are saved.
- Alcohol and iron are reversible causes of a dilated cardiomyopathy.

- A low-voltage electrocardiogram with thick walls and a history of carpal tunnel syndrome is amyloidosis.

Questions you should be able to answer:

- Which single bedside manoeuvre separates this from aortic stenosis, and what happens to the murmur?
- Name four features that increase his risk of sudden death, and say which he has.
- What do you do about his parents, his sister and his cousins?
- A colleague prescribes glyceryl trinitrate for his chest pain. What is your concern?
- A 68-year-old has heart failure with a normal ejection fraction, thick ventricular walls and a low-voltage electrocardiogram. What is your diagnosis and how do you confirm it?