The Respiratory Block

Pneumonia, asthma, COPD, suppurative lung disease, hypercapnic failure, pleural effusion, lung cancer and pulmonary hypertension.

Cases 1–8 · 12,309 words

This block covers the respiratory teaching of both internal medicine courses. Bacterial pneumonia is taught in the infectious diseases block but presents as a respiratory case and is placed here for clinical coherence. The **second course** adds cases 7 and 8.

Case	Lecture it serves
System opener: the respiratory history and examination	History taking in respiratory; physical examination of respiratory
Case 1 — Community-acquired pneumonia	Bacterial pneumonia
Case 2 — Acute severe asthma	Bronchial asthma
Case 3 — Exacerbation of chronic obstructive pulmonary disease	Chronic obstructive pulmonary disease
Case 4 — Bronchiectasis and lung abscess	Suppurative lung diseases
Case 5 — Acute-on-chronic hypercapnic respiratory failure	Respiratory failure
Case 6 — Pleural effusion	Pleural effusion
Case 7 — Lung cancer	Lung cancer
Case 8 — Pulmonary hypertension	Pulmonary arterial hypertension

Pulmonary thromboembolism is taught in the second course and is covered as Seminar 11 in Part Two, where the pre-test probability and imaging decisions can be set out properly. **The approach to obstructive airway disease**, a second-course seminar, is covered by Cases 2 and 3.

Case numbering is provisional and will be reconciled when the blocks are compiled into a single volume.

System Opener · The Respiratory History and Examination

This page is the framework the cases assume. Learn it once and the cases become variations on it rather than separate lists to memorise.

The history

The six cardinal symptoms

Symptom	What to establish
Cough	Duration (under 3 weeks is acute, 3–8 weeks subacute, over 8 weeks chronic), dry or productive, timing across the day, what provokes it, and whether it disturbs sleep.
Sputum	Daily volume — a teaspoon, an eggcup or a cupful. Colour, smell, and whether it settles into layers. Large volumes of purulent sputum every day is bronchiectasis until proved otherwise.
Haemoptysis	Quantify it: streaking, teaspoons or cupfuls. Establish it is truly from the chest and not the nose or stomach. Persistent haemoptysis in a smoker is cancer until proved otherwise.
Breathlessness	Onset over minutes, hours, days or months. Exercise tolerance in metres or flights of stairs against the patient's own baseline. Orthopnoea, paroxysmal nocturnal dyspnoea, and variation through the day and across the week.
Wheeze	Audible or not, when it occurs, what relieves it, and whether it is seasonal or occupational.
Chest pain	Pleuritic (sharp, worse on inspiration, localised) points to the pleura; central and heavy points elsewhere.

Then the four contexts that change the differential

- **Exposures.** Smoking in pack-years, including waterpipe and vaping. Occupation over the whole working life — dust, silica, asbestos with its decades of latency, farming, welding. Birds, mould, air conditioning. Travel and tuberculosis contact.
- **Drugs.** Beta blockers and aspirin provoke bronchospasm; amiodarone, methotrexate, nitrofurantoin and bleomycin cause parenchymal disease; angiotensin-converting enzyme inhibitors cause a dry cough.
- **Atopy and family history.** Eczema, allergic rhinitis, food allergy, and a family history of asthma, tuberculosis, cystic fibrosis or alpha-1 antitrypsin deficiency. Ask about consanguinity where relevant.
- **Disease burden.** Number of exacerbations, courses of oral steroids and admissions in the past year; any previous intensive care admission or intubation; home oxygen or non-invasive ventilation; current inhalers, adherence and technique; vaccination status.

The examination

From the end of the bed

- Respiratory rate counted over a full minute, respiratory distress, posture, ability to complete a sentence, cachexia, audible wheeze or stridor, pursed-lip breathing, the oxygen device and its flow rate, and the sputum pot — always look inside it.
- Chest shape: barrel chest of hyperinflation, kyphoscoliosis, pectus deformity, thoracoplasty or lobectomy scars.

Hands, face and neck

- **Clubbing.** In respiratory disease it means bronchial carcinoma, bronchiectasis, lung abscess or empyema, idiopathic pulmonary fibrosis, cystic fibrosis or mesothelioma. **It is not caused by asthma or**

by chronic obstructive pulmonary disease — if you find it in a patient labelled with either, you have found a second diagnosis.

- Tar staining, fine tremor of beta-agonist overuse, the coarse flapping tremor of carbon dioxide retention, wasting of the small hand muscles in an apical tumour, bounding pulse and warm hands in hypercapnia, pulsus paradoxus in severe asthma.
- Central cyanosis at the tongue, Horner syndrome, jugular venous pressure, supraclavicular and cervical lymph nodes, tracheal position, cricosternal distance (reduced in hyperinflation) and tracheal tug.

The chest, in the order that survives an examiner

- **Inspect** — symmetry of movement, scars, dilated veins, radiotherapy field markings.
- **Palpate** — expansion upper and lower, tactile vocal fremitus, apex beat position, chest wall tenderness.
- **Percuss** — including the clavicles and the axillae, and note the upper level of liver dullness, which is lost in hyperinflation.
- **Auscultate** — breath sound intensity and character, added sounds, vocal resonance. **Always examine the back**, where the lower lobes actually are.
- **Finish** — ankle and sacral oedema, calves, peak expiratory flow, and the observation chart.

Added sound	What it means
Polyphonic wheeze	Many airways of different calibre narrowed together — asthma, chronic obstructive pulmonary disease.
Monophonic, fixed wheeze	One airway narrowed — tumour or inhaled foreign body. Does not move with coughing.
Fine, late inspiratory crackles	Reopening of collapsed alveoli — pulmonary fibrosis, pulmonary oedema. Do not clear with coughing.
Coarse, early inspiratory crackles	Secretions in larger airways — bronchiectasis, chronic bronchitis. Shift or clear with coughing.
Pleural rub	Inflamed pleural surfaces moving on each other — pneumonia, infarction, pleurisy.
Stridor	Upper airway obstruction. Loudest over the neck, inspiratory. This is an emergency, not a sign to characterise at leisure.

Case 1 · Community-Acquired Pneumonia

CLINICAL VIGNETTE

A **68-year-old man** is brought to the emergency department by his son with three days of fever and cough. The cough is productive of thick rust-coloured sputum. He has a sharp right-sided chest pain that is worse on deep inspiration, and has become breathless walking to the bathroom. Since this morning he has been muddled and did not recognise his neighbour.

He has type 2 diabetes mellitus on metformin, and a 40 pack-year smoking history. He has had no vaccinations.

On arrival: temperature 38.9 °C, heart rate 112/min, blood pressure 96/58 mmHg, respiratory rate 28/min, oxygen saturation 88% on room air. He is orientated to person only.

1. Focused History

Characterise the presenting complaint

- **Onset and tempo.** Abrupt onset over hours with a rigor suggests bacterial, classically pneumococcal, disease. A gradual prodrome over one to two weeks with headache and myalgia points to an atypical organism.
- **Cough and sputum.** Ask about volume, colour and blood. Rust-coloured sputum is classically pneumococcal; foul-smelling sputum suggests anaerobic infection or abscess; copious purulent sputum daily for years suggests underlying bronchiectasis.
- **Pleuritic chest pain.** Localises the affected lobe and tells you the pleura is inflamed. Pain that persists or worsens after 48 hours of antibiotics raises the question of empyema.
- **Breathlessness.** Quantify against his own baseline — "how far could you walk last week, and how far today?" — not against an abstract scale.

Red flags to exclude at once

- Confusion or drowsiness, syncope, inability to speak in full sentences, cyanosis, haemoptysis, poor oral intake and reduced urine output. Any of these moves the patient to the resuscitation area before the history is finished.

Questions that discriminate between causes

Ask about	If positive, think
Rigors, single abrupt onset, rusty sputum, herpes labialis	Streptococcus pneumoniae
Young patient, dry cough, headache, arthralgia, rash, recent similar illness in the household	Mycoplasma pneumoniae
Recent hotel or air-conditioning exposure, travel, diarrhoea, confusion out of proportion, low sodium	Legionella pneumophila
Alcohol excess, seizure, stroke or dysphagia, poor dentition, vomiting, foul sputum	Aspiration and anaerobes
Cough over three weeks, night sweats, weight loss, haemoptysis, contact or endemic exposure	Pulmonary tuberculosis
Sudden pleuritic pain and breathlessness with little fever or sputum, calf pain, immobility	Pulmonary embolism
Bird or animal contact, farm or abattoir work, unpasteurised dairy	Psittacosis, Coxiella, Brucella
Known bronchiectasis or cystic fibrosis, recent hospital admission or antibiotics	Pseudomonas or resistant organisms

Risk factors and background

- **Host factors:** age, smoking, chronic obstructive pulmonary disease, diabetes mellitus, chronic kidney or liver disease, heart failure, malnutrition.
- **Immunosuppression:** corticosteroids, chemotherapy, biologic agents, human immunodeficiency virus (HIV) infection, asplenia, transplant. This changes the organism list and therefore the antibiotic.
- **Recent healthcare exposure and antibiotics** within 90 days — the strongest single predictor of resistance.
- **Vaccination status** for pneumococcus and influenza, and **drug allergies**, especially to penicillin.
- **Functional baseline and social circumstances.** Whether he lives alone, and who can observe him, decides admission as often as the physiology does.

2. Physical Examination

General inspection and vital signs

- Respiratory distress: accessory muscle use, nasal flaring, inability to complete a sentence, tripod posture.
- A **complete set of vital signs including the respiratory rate**. The respiratory rate is the earliest and most sensitive marker of deterioration and the one most often left unrecorded. Count it for a full minute.
- Level of consciousness — formally, using an abbreviated mental test or the Glasgow Coma Scale, because "confusion" scores a point on the severity index and must be documented objectively.
- Peripheral perfusion: capillary refill, warm vasodilated peripheries with a bounding pulse in early sepsis, cool and mottled in late shock.

Hands, face and neck

- Tar staining, peripheral and central cyanosis, a carbon dioxide retention flap if there is coexisting chronic obstructive pulmonary disease, herpes labialis around the mouth, dehydrated mucous membranes.
- Tracheal position — central in uncomplicated consolidation. Deviation means collapse (pulled towards) or a large effusion or tension pneumothorax (pushed away).

The chest, in sequence

- **Inspect:** reduced movement on the affected side, tachypnoea, scars, chest wall deformity.
- **Palpate:** chest expansion, comparing sides; tactile vocal fremitus is **increased** over consolidated lung because solid tissue transmits sound better than air.
- **Percuss:** dull over consolidation. **Stony** dullness means fluid, not consolidation — this single distinction changes your next step from antibiotics alone to a diagnostic pleural tap.
- **Auscultate:** bronchial breathing over the consolidated lobe, coarse late inspiratory crackles, increased vocal resonance and whispering pectoriloquy. A pleural rub may be heard where the pain is.
- **Then look for complications:** an area of stony dullness with absent breath sounds at the base (parapneumonic effusion or empyema), a new murmur (endocarditis as a source), neck stiffness (meningitis from the same pneumococcus).

The bedside distinction you must be able to make. Four processes reduce chest expansion and all four appear in examinations. The table below is the answer to the commonest viva question in respiratory medicine.

Sign	Consolidation	Pleural effusion	Pneumothorax	Lobar collapse
Trachea	Central	Central, pushed away if large	Central, pushed away if tension	Pulled towards
Expansion	Reduced	Reduced	Reduced	Reduced
Percussion	Dull	Stony dull	Hyper-resonant	Dull
Breath sounds	Bronchial	Absent or diminished	Diminished	Diminished
Vocal resonance	Increased	Decreased	Decreased	Decreased
Added sounds	Crackles, rub	Rub above the fluid	None	None

3. Differential Diagnosis

Diagnosis	The feature that discriminates it
Pulmonary embolism with infarction	Hypoxia out of proportion to the chest radiograph; risk factors for venous thromboembolism; little or no purulent sputum.
Acute pulmonary oedema	Orthopnoea and paroxysmal nocturnal dyspnoea, raised jugular venous pressure, bilateral basal crackles, third heart sound, no fever.
Exacerbation of chronic obstructive pulmonary disease	Wheeze and prolonged expiration dominate; the radiograph shows no consolidation.
Pulmonary tuberculosis	Weeks rather than days of symptoms, weight loss, night sweats, upper lobe or apical changes with cavitation.
Lung cancer with post-obstructive pneumonia	Smoker, weight loss, haemoptysis, and consolidation that fails to clear on the follow-up radiograph.
Empyema	Persisting fever and pain after 48–72 hours of appropriate antibiotics, with stony dullness at the base.

4. Investigations

At the bedside

- **Pulse oximetry on room air**, documented as such — a saturation on oxygen without the flow rate recorded is uninterpretable.
- **Arterial blood gas** if the saturation is below 94%, if there is shock, or if chronic obstructive pulmonary disease raises the possibility of carbon dioxide retention. Expect type 1 respiratory failure; a rising carbon dioxide in a tiring patient is an ominous sign.
- **Electrocardiogram** — to detect the atrial fibrillation that sepsis commonly precipitates, and because chest pain in a 68-year-old deserves one.
- **Capillary blood glucose** — infection destabilises diabetes and may precipitate a hyperglycaemic emergency.

Laboratory

Test	What you expect, and what it changes
Complete blood count	Neutrophil leucocytosis. A low white cell count (<4 × 10^9/L) is a marker of severe disease, not of mild disease.
Urea and electrolytes	Urea is one of the five severity criteria. Hyponatraemia raises the suspicion of Legionella.
C-reactive protein	Baseline for trend. Failure to fall by more than half by day four predicts treatment failure and should trigger reassessment.
Liver function, lactate	Lactate above 2 mmol/L in the presence of infection defines sepsis and mandates the one-hour bundle.
Blood cultures, two sets	Before the first antibiotic dose, but never delaying it. Yield is low in mild disease — reserve for moderate and severe cases.
Sputum Gram stain and culture	Useful only if a good deep specimen is obtained before antibiotics. Send acid-fast staining if tuberculosis is possible.
Urinary antigens	Pneumococcal and Legionella antigen in severe disease. They remain positive after antibiotics have been started.
Respiratory viral panel	Influenza and severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) — changes isolation and adds antiviral treatment.
Human immunodeficiency virus test	Offer in any severe or recurrent pneumonia, and whenever the organism is unusual.

Imaging

- **Chest radiograph, posteroanterior.** Expect lobar consolidation with an air bronchogram. Use the **silhouette sign** to localise: loss of the right heart border places the disease in the right middle lobe; loss of a hemidiaphragm places it in a lower lobe. Also inspect for effusion, cavitation and a mass.
- **Point-of-care chest ultrasound.** More sensitive than the radiograph for pleural fluid and it guides the needle. It has largely replaced the lateral decubitus film.
- **Computed tomography of the chest** if the pneumonia fails to resolve, if complications are suspected, or if the radiograph is equivocal in a very sick patient.
- **Repeat the chest radiograph at six weeks** in every smoker and in anyone over 50. Persistent consolidation is a bronchial carcinoma until proved otherwise. This is the single follow-up step most often forgotten.

Pleural fluid, if an effusion is present

Any effusion more than 1 cm deep on ultrasound in a febrile patient should be sampled. Send protein, lactate dehydrogenase, glucose, pH, Gram stain, culture and cytology. A **pH below 7.2**, glucose below 2.2 mmol/L, or frank pus means a complicated parapneumonic effusion or empyema, and the treatment is a chest drain, not more antibiotics.

5. Management

Immediate — the first hour

- **Oxygen** titrated to a saturation of 94–98%, or 88–92% if there is a risk of carbon dioxide retention.

- **Intravenous access, fluid resuscitation** with balanced crystalloid for hypotension, and hourly urine output measurement.
- **Antibiotics within one hour** if septic, and within four hours in all other cases. Take the cultures on the way, not instead.
- **Escalate early.** Persistent hypotension after fluid, a rising lactate, or exhaustion means a critical care referral now rather than at the next ward round.

Score the severity — CURB-65

Criterion	Threshold
C onfusion	New disorientation in person, place or time
U rea	Above 7 mmol/L (about 19 mg/dL)
R espiratory rate	30/min or more
B lood pressure	Systolic below 90 mmHg or diastolic 60 mmHg or less
65 — age	65 years or older

- **0–1:** low risk, suitable for treatment at home.
- **2:** moderate risk, admit to a general ward.
- **3–5:** high risk, consider critical care. Our patient scores 4 (confusion, respiratory rate, blood pressure, age) and requires a high-dependency bed.

The score guides but does not replace judgement. A hypoxic patient with a score of 1, or one who cannot manage at home, is still admitted.

Antibiotic therapy

Setting	Regimen	Duration
Low severity, outpatient	Amoxicillin 1 g three times daily orally. Doxycycline or a macrolide if penicillin-allergic.	5 days
Moderate, ward	Amoxicillin (or ceftriaxone) plus a macrolide such as azithromycin; or a respiratory fluoroquinolone as monotherapy.	5–7 days
Severe or critical care	Intravenous ceftriaxone or co-amoxiclav plus intravenous azithromycin.	7 days
Suspected aspiration	Co-amoxiclav, or add metronidazole to cover anaerobes.	7 days
Structural lung disease	Add antipseudomonal cover, for example piperacillin–tazobactam.	Guided by culture
Risk of resistant Staphylococcus aureus	Add vancomycin or linezolid; consider after influenza and in intravenous drug use.	Guided by culture

Switch from intravenous to oral once the patient has been afebrile for 48 hours, is haemodynamically stable and is absorbing enterally. **A macrolide is added, not substituted,** because atypical organisms are not covered by beta-lactams and cannot be excluded clinically at the front door.

Supportive care

- Venous thromboembolism prophylaxis, analgesia adequate to allow deep breathing and coughing, early mobilisation, chest physiotherapy if sputum retention is a problem, attention to nutrition and to glycaemic control.
- Corticosteroids are not given routinely, but there is now evidence of benefit in severe pneumonia requiring critical care. Follow your local protocol.

Complications to anticipate

- Parapneumonic effusion and empyema, lung abscess, acute respiratory distress syndrome, sepsis with acute kidney injury, new atrial fibrillation, and non-resolving pneumonia.

Discharge and follow-up

- Discharge when the patient is afebrile, saturating adequately on room air, eating and drinking, mentally back to baseline, and has no more than one instability criterion.
- At discharge: smoking cessation advice, pneumococcal and influenza vaccination, a clear safety-net for return, and **the six-week chest radiograph booked before he leaves the ward**.

6. Teaching Points and Viva Questions

- The respiratory rate is the most useful and most neglected vital sign. A rate of 30 in a quiet-looking patient is a medical emergency.
- Bronchial breathing requires a patent airway leading to solid lung; absent breath sounds with stony dullness means fluid between the lung and your stethoscope.
- The elderly may present with confusion, a fall or simply "off legs", and may be afebrile. Absence of fever does not exclude pneumonia.
- Urea appears in the severity score because it reflects both volume depletion and the catabolic burden of severe infection.
- Chest signs may be entirely normal in the first 24 hours. A normal examination does not exclude the diagnosis.

Questions you should be able to answer:

- Distinguish consolidation from a pleural effusion at the bedside using four signs.
- This patient is not improving on day three. What are your next four steps?
- When do you drain a parapneumonic effusion, and on what result?
- Why is a macrolide added to a beta-lactam rather than used alone?
- Why does a smoker need a repeat chest radiograph after recovery?

Case 2 · Acute Severe Asthma

CLINICAL VIGNETTE

A **19-year-old student** with asthma since childhood arrives with two days of worsening wheeze and breathlessness following a head cold. She has been using her salbutamol inhaler almost hourly with diminishing benefit and did not sleep last night. She stopped her preventer inhaler some months ago because she felt well.

She was admitted twice in the past year and spent two days in intensive care at the age of sixteen.

On arrival: she speaks in short phrases only. Temperature 37.2 °C, heart rate 128/min, respiratory rate 32/min, blood pressure 118/76 mmHg, oxygen saturation 91% on room air. Peak expiratory flow 38% of predicted. Widespread polyphonic wheeze with prolonged expiration.

1. Focused History

Take it in parallel with treatment. Nebulised salbutamol does not wait for the social history.

This attack

- Speed of onset — over hours suggests a viral or allergen trigger; over minutes raises anaphylaxis or an inhaled foreign body.
- The trigger: upper respiratory infection, allergen, exercise, cold air, cigarette or waterpipe smoke, emotion, occupational exposure, or a drug — **aspirin, other non-steroidal anti-inflammatory drugs, and beta blockers**, including eye drops.
- How much reliever has been used, and whether it is still working. Diminishing response to salbutamol is a severity marker.

Background asthma control

- Night waking, morning dipping, and days per week of symptoms — the practical measure of control.
- Current inhalers, **adherence, and technique**. Ask her to demonstrate rather than to describe. Poor technique is the commonest reason an apparently adequate regimen fails.
- Number of oral corticosteroid courses in the past twelve months and the number of reliever canisters used per month.

RISK FACTORS FOR NEAR-FATAL ASTHMA

Ask directly about every one of these. Their presence changes disposition even if the physiology improves:

Previous intubation or intensive care admission · two or more admissions in the past year · three or more classes of asthma drug · more than one reliever canister per month · repeated attendance at the emergency department · poor adherence or non-attendance · food allergy · psychiatric illness or major social difficulty.

2. Physical Examination

The four bedside measurements that grade the attack

- **Can she complete a sentence?** Ask her name and address and listen to the phrasing, not the answer.
- **Respiratory rate and heart rate**, counted, not estimated.

- **Oxygen saturation on room air.**
- **Peak expiratory flow** against predicted or her own best. It is the single most useful measurement in asthma and takes fifteen seconds.

Signs of danger

- Accessory muscle use, tracheal tug, tripod posture, inability to lie flat.
- **A silent chest.** Air must move to make a wheeze. A chest that has gone quiet in a distressed patient means airflow has fallen too low to generate sound, and is a pre-terminal sign.
- Cyanosis, exhaustion, drowsiness or confusion, bradycardia, hypotension, arrhythmia — the peri-arrest picture.
- Pulsus paradoxus, and subcutaneous emphysema or asymmetric signs suggesting pneumothorax or pneumomediastinum.

3. Grading the Attack

Grade	Features — any one is sufficient
Moderate	Peak flow above 50% predicted, speech normal, no features of a more severe grade.
Acute severe	Peak flow 33–50%, respiratory rate 25/min or more, heart rate 110/min or more, unable to complete a sentence in one breath.
Life-threatening	Peak flow below 33%, oxygen saturation below 92%, silent chest, cyanosis, poor respiratory effort, exhaustion, altered consciousness, arrhythmia or hypotension. A normal carbon dioxide level belongs in this group.
Near-fatal	Raised arterial carbon dioxide, or the need for mechanical ventilation.

4. Differential Diagnosis

Diagnosis	The feature that discriminates it
Anaphylaxis	Minutes from exposure, with urticaria, angio-oedema and hypotension. Treat with adrenaline first.
Chronic obstructive pulmonary disease	Older, heavy smoking history, fixed rather than variable airflow limitation, chronic daily sputum.
Inhaled foreign body	Sudden onset, unilateral monophonic wheeze, and no history of atopy.
Pneumothorax	Sudden pleuritic pain, unilateral hyper-resonance and absent breath sounds. Always suspect it when asthma deteriorates abruptly.
Acute pulmonary oedema	Orthopnoea, raised jugular venous pressure, bilateral basal crackles as well as wheeze, third heart sound.
Pulmonary embolism	Pleuritic pain with hypoxia but little wheeze, and risk factors for venous thromboembolism.
Inducible laryngeal obstruction	Noise loudest over the neck and inspiratory, normal saturations and peak flow, no response to bronchodilator.

5. Investigations

- **Peak expiratory flow** before and after treatment — the measure of response.
- **Pulse oximetry**, aiming to keep saturation at 94–98%.
- **Arterial blood gas** if saturation stays below 92% or there are life-threatening features. Expect a low carbon dioxide from hyperventilation. **A normal or rising carbon dioxide means the patient is tiring** and requires immediate senior and critical care involvement.
- **Chest radiograph** only if pneumothorax or consolidation is suspected, if there is no response to treatment, or if ventilation is being considered. It is not routine.
- **Potassium** — both beta agonists and corticosteroids lower it, and the treatment itself is the commonest cause of hypokalaemia here. Also glucose, complete blood count and C-reactive protein.

No investigation should delay the first nebuliser.

6. Management

Immediate, in this order

Intervention	Detail
Oxygen	Titrate to a saturation of 94–98%. Unlike in chronic obstructive pulmonary disease, there is no reason to restrict it.
Salbutamol	5 mg nebulised, oxygen-driven. Repeat every 15–20 minutes, or give continuously in severe attacks.
Ipratropium bromide	500 micrograms nebulised with the salbutamol, every 4–6 hours, in acute severe and life-threatening attacks.
Corticosteroid	Prednisolone 40–50 mg orally, or hydrocortisone 100 mg intravenously if she cannot swallow. Continue for at least 5 days. No taper is needed after a short course.
Magnesium sulfate	1.2–2 g intravenously over 20 minutes, as a single dose, in acute severe asthma not responding to initial nebulisers.
Escalation	Intravenous salbutamol or aminophylline only with senior input. Involve critical care early for exhaustion, a rising carbon dioxide, or a falling conscious level.

- **Antibiotics are not routine.** Most attacks are triggered by viruses. Give them only for genuine evidence of bacterial infection.
- **Never sedate a breathless asthmatic.** Agitation is hypoxia, not anxiety.

Monitoring

- Peak flow before and after every nebuliser; continuous oximetry; potassium after repeated doses; repeat blood gas if the first was abnormal or the patient is not improving.

Discharge and follow-up

- Discharge when peak flow exceeds 75% of best or predicted, when she has been off nebulisers for 24 hours, and when saturations are stable on air.

- Before she leaves: **inhaler technique observed and corrected**, the inhaled corticosteroid restarted or stepped up, the remainder of the prednisolone course dispensed, and a **written personalised asthma action plan** in her hand.
- Primary care review within 48 hours and a respiratory clinic appointment within four weeks. Every admission is a failure of chronic control and should trigger a review of it.

Complications

- Pneumothorax and pneumomediastinum, hypokalaemia, lactic acidosis from high-dose beta agonists, and respiratory arrest.

7. Teaching Points and Viva Questions

- The silent chest is the most dangerous sign in the room and the easiest to misread as improvement.
- A normal carbon dioxide in a patient who has been hyperventilating for hours is a warning, not a reassurance.
- Peak flow is objective, repeatable and free. Record it before and after every intervention.
- Overuse of reliever inhaler and underuse of preventer is the pattern behind almost every severe attack.
- Clubbing does not occur in asthma. If you see it, look for bronchiectasis or cancer.

Questions you should be able to answer:

- Grade this attack and justify each criterion you use.
- Why is a normal arterial carbon dioxide a life-threatening feature?
- When would you give magnesium, and what is the evidence for it?
- Her potassium is 2.9 mmol/L after four nebulisers. Explain why.
- What must be done before she is allowed home?

Case 3 · Exacerbation of Chronic Obstructive Pulmonary Disease

CLINICAL VIGNETTE

A **66-year-old retired driver** with a 50 pack-year smoking history is brought in with three days of increasing breathlessness. His sputum has increased in volume and turned green. His ankles have been swelling for a fortnight. He normally manages about 100 metres on the flat before stopping.

The ambulance crew found him hypoxic and gave oxygen at 15 litres per minute by reservoir mask. He has become progressively drowsier during the journey.

On arrival: temperature 37.4 °C, heart rate 104/min, respiratory rate 26/min, blood pressure 138/82 mmHg, oxygen saturation 99% on the reservoir mask. He is rousable but confused, with a coarse flapping tremor.

Arterial blood gas on 15 L/min: pH 7.24, carbon dioxide 9.1 kPa, oxygen 22 kPa, bicarbonate 30 mmol/L.

1. Focused History

Characterise the exacerbation

- The three cardinal symptoms: **increased breathlessness, increased sputum volume, and increased sputum purulence**. How many are present decides whether antibiotics are indicated.
- Baseline exercise tolerance, described in metres or stairs, and how far it has fallen.
- Fever, pleuritic pain, haemoptysis, unilateral leg swelling, chest pain — each pointing to a different complication or alternative diagnosis.

Establish the baseline disease

- Exacerbations, courses of steroids and admissions in the past year; **previous non-invasive ventilation or intubation, and whether it helped**; home nebulisers, home oxygen and its prescribed hours.
- Current inhalers, adherence and technique; vaccination status; smoking status now, not only historically.
- Comorbidity that mimics or coexists: ischaemic heart disease, heart failure, atrial fibrillation, anxiety, osteoporosis, malnutrition.
- Alpha-1 antitrypsin deficiency if the disease began young, if he is a light smoker, or if there is liver disease or a family history.
- **Function and wishes.** How he lives, who helps him, and whether escalation to ventilation has ever been discussed. This conversation is far easier now than at three in the morning.

2. Physical Examination

Signs of the underlying disease

- Pursed-lip breathing, accessory muscle use, tripod posture, barrel-shaped chest, reduced cricosternal distance, tracheal tug, indrawing of the lower ribs on inspiration.
- Hyperinflation: reduced expansion, hyper-resonant percussion with **loss of cardiac and liver dullness**, and quiet breath sounds throughout. Wheeze may be present or absent.

Signs of carbon dioxide retention

- Drowsiness and confusion, a coarse flapping tremor, bounding pulse, warm peripheries, headache, and in extreme cases papilloedema.

Signs of cor pulmonale

- Raised jugular venous pressure, right ventricular heave, loud pulmonary second sound, tender pulsatile hepatomegaly, ankle and sacral oedema.

Signs that mean something else is going on

- **Clubbing** — not a feature of chronic obstructive pulmonary disease; look for carcinoma or bronchiectasis. Focal consolidation, stony dullness, unilateral hyper-resonance, or an asymmetric swollen calf.

3. Differential Diagnosis

Diagnosis	The feature that discriminates it
Pneumonia	Focal consolidation on examination and radiograph, higher fever, greater inflammatory response.
Acute heart failure	Orthopnoea, raised jugular venous pressure, third heart sound, bilateral basal crackles, cardiomegaly and upper lobe diversion on the film.
Pulmonary embolism	Hypoxia disproportionate to the film, pleuritic pain, unilateral calf swelling. Consider it in every exacerbation that fails to respond.
Pneumothorax	Sudden deterioration with unilateral hyper-resonance. Emphysematous lungs are prone to it, and it can be subtle on the film.
Lung cancer	Weight loss, haemoptysis, clubbing, a mass or lobar collapse.
Sedative or opioid effect	Small pupils, a low respiratory rate rather than a high one, and the drug chart.

4. Investigations

READING THE BLOOD GAS

pH tells you how acute it is; bicarbonate tells you how chronic it is.

A raised carbon dioxide with a **normal** bicarbonate is acute — the kidney has not had time to compensate.

A raised carbon dioxide with a **raised** bicarbonate and a normal pH is chronic and compensated.

A raised carbon dioxide with a raised bicarbonate **and an acidaemia**, as here, is an acute deterioration on top of chronic retention. That combination is the indication to prepare for non-invasive ventilation.

- **Arterial blood gas** on a documented inspired oxygen concentration, repeated 30–60 minutes after any change in oxygen therapy or treatment.
- **Chest radiograph** — to exclude pneumonia, pneumothorax and a mass, and to look for hyperinflation with flattened hemidiaphragms.
- **Electrocardiogram** — right axis deviation, P pulmonale, right bundle branch block, atrial fibrillation or multifocal atrial tachycardia, and ischaemia.

- **Complete blood count** — secondary polycythaemia reflects chronic hypoxaemia; anaemia worsens breathlessness. **Urea and electrolytes**, **C-reactive protein**, glucose, and theophylline level if he takes it.
- **Sputum culture** if purulent or if he has failed previous antibiotics; **blood cultures** if febrile.
- **Natriuretic peptide, troponin, or computed tomographic pulmonary angiography** when heart failure, ischaemia or embolism cannot be excluded clinically.

Reading the film

- Hyperinflation is more than six anterior ribs above the diaphragm in the midclavicular line, flattened hemidiaphragms, a narrow vertical heart, and increased retrosternal air on the lateral view. Then look specifically at the apices and the lateral edges for a pneumothorax, which is easily missed against emphysematous lung.

5. Management

THE FIRST CORRECTION TO MAKE IN THIS PATIENT

His saturation of 99% is not reassuring — it is the problem. Uncontrolled oxygen has worsened his hypercapnia. **The answer is not to remove oxygen but to control it**: reduce to a 24–28% Venturi mask, target a saturation of 88–92%, and repeat the gas in 30 minutes. Hypoxia kills faster than hypercapnia; the aim is enough oxygen, not none.

Immediate treatment

Intervention	Detail
Controlled oxygen	Venturi mask 24–28%, target saturation 88–92%. Write the target on the observation chart. Reassess with a gas after 30–60 minutes.
Bronchodilators	Nebulised salbutamol 5 mg and ipratropium 500 micrograms. Drive the nebuliser with air, giving oxygen by nasal cannula alongside, in a patient who retains carbon dioxide.
Corticosteroid	Prednisolone 30 mg orally daily for 5 days.
Antibiotics	Indicated when sputum is purulent or when two of the three cardinal symptoms are present. Amoxicillin, doxycycline or a macrolide for 5 days, adjusted to local resistance patterns and previous cultures.
Non-invasive ventilation	For a pH below 7.35 with carbon dioxide above 6.5 kPa persisting after one hour of optimal medical therapy. This patient qualifies now.
Supportive	Venous thromboembolism prophylaxis, cautious diuresis for cor pulmonale, nutrition, early mobilisation. Avoid sedatives and opioids.

Non-invasive ventilation — what a student must know

- It is a **treatment**, not a sign of failure, and it reduces mortality and the need for intubation in this exact situation.
- Contraindications: inability to protect the airway, vomiting, facial trauma or burns, an undrained pneumothorax, haemodynamic instability, and a patient who cannot tolerate the mask.
- **Decide the ceiling of care before starting**, and document it: is non-invasive ventilation a bridge to intubation, or the ceiling itself? Involve the patient and family while he can still participate.

- Reassess with a blood gas one hour after starting. Failure to improve means escalation or a change of plan, not more of the same.

Before discharge, and for the long term

- **Smoking cessation** — the only intervention that alters the decline in lung function. Offer pharmacotherapy, not only advice.
- **Pulmonary rehabilitation** — the intervention with the largest effect on symptoms and quality of life, and the one most often not referred for.
- Inhaler technique reviewed and the regimen rationalised; influenza and pneumococcal vaccination; a rescue pack with a written plan for its use.
- **Long-term oxygen therapy assessment when stable**, at least 8 weeks after the exacerbation: arterial oxygen at or below 7.3 kPa, or at or below 8 kPa with polycythaemia, peripheral oedema or pulmonary hypertension. It must be used at least 15 hours a day to prolong life, and never with continued smoking.
- Nutritional assessment, anxiety and depression screening, and an honest conversation about the trajectory of the disease.

6. Teaching Points and Viva Questions

- Give oxygen, but give it in a measured concentration with a written target. Withholding it altogether is as much an error as flooding the patient with it.
- The bicarbonate is the historian of the blood gas — it tells you how long this has been going on.
- Smoking cessation and pulmonary rehabilitation change outcomes more than any inhaler.
- A patient with chronic obstructive pulmonary disease who deteriorates suddenly has a pneumothorax or a pulmonary embolism until proved otherwise.
- Ceiling-of-care decisions belong in the notes early, made with the patient, not improvised during a crisis.

Questions you should be able to answer:

- Explain the mechanism by which high-concentration oxygen raised this man's carbon dioxide.
- What are your criteria for starting non-invasive ventilation, and what would you do if it fails?
- When do you give antibiotics in an exacerbation, and why not always?
- List the criteria for long-term oxygen therapy and explain why it must be worn 15 hours a day.
- He has clubbing. Does that change your thinking?

Case 4 · Suppurative Lung Disease: Bronchiectasis and Lung Abscess

CLINICAL VIGNETTE

A **34-year-old woman** attends with two weeks of fever, worsening cough and foul-smelling sputum. For as long as she can remember she has coughed every morning and produced sputum daily; over the past two weeks the volume has risen to about half a cup a day and she has coughed up streaks of blood. She has lost 6 kg over three months.

She had severe pneumonia as a child and has had recurrent chest infections since. She has chronic nasal congestion. Her parents are first cousins.

On examination: she is thin. There is **clubbing**. Temperature 38.4 °C, respiratory rate 22/min, oxygen saturation 94% on air. Coarse early inspiratory crackles at both bases that alter after she coughs, with scattered wheeze. The sputum pot contains thick green sputum that has separated into layers.

1. Focused History

Characterise the sputum — this is the diagnostic history

- **Daily volume**, quantified against a household measure. Daily purulent sputum for years is bronchiectasis until proved otherwise, and no other common condition produces it.
- **Smell.** Foul or putrid sputum means anaerobic organisms, and therefore aspiration, abscess or empyema.
- **Haemoptysis**, quantified. Bronchiectasis is one of the commonest causes of massive haemoptysis.
- Whether the symptoms have been lifelong, or began after a specific illness.

Find the underlying cause

Ask about	Cause it points to
Severe childhood pneumonia, measles, whooping cough, previous tuberculosis	Post-infective bronchiectasis — the commonest cause
Chronic sinusitis, infertility, situs inversus	Primary ciliary dyskinesia
Steatorrhoea, poor growth, infertility, diabetes, family history, consanguinity	Cystic fibrosis — may present in adulthood with milder mutations
Recurrent sinopulmonary, skin or gut infections since childhood	Antibody deficiency, especially common variable immunodeficiency
Difficult asthma with brown sputum plugs and fleeting shadows	Allergic bronchopulmonary aspergillosis
Rheumatoid arthritis, inflammatory bowel disease	Associated bronchiectasis
Alcohol excess, seizures, stroke, poor dentition, oesophageal disease, reduced consciousness	Aspiration — the classic setting for lung abscess
Recurrent pneumonia always in the same lobe	Obstruction — inhaled foreign body or tumour

2. Physical Examination

- **Clubbing** — present in established suppurative lung disease and a key discriminator from chronic obstructive pulmonary disease.

- Cachexia and reduced muscle bulk, reflecting the metabolic cost of chronic sepsis.
- **Inspect the sputum pot.** Copious purulent sputum that separates into a frothy upper layer, a clear middle layer and a dense purulent lower layer is characteristic.
- **Coarse early inspiratory crackles that change or clear with coughing** — secretions moving in large airways. Wheeze is common. Signs may be focal or widespread.
- Look for complications: dullness with absent breath sounds (empyema), focal consolidation, and the raised jugular venous pressure, right ventricular heave and oedema of late cor pulmonale.
- Look for the cause: nasal polyps and sinus tenderness, dextrocardia, the teeth and gums, deforming arthropathy of the hands.

3. Differential Diagnosis

Diagnosis	The feature that discriminates it
Chronic obstructive pulmonary disease	Smoker, no clubbing, smaller sputum volumes, and a computed tomogram without dilated airways.
Pulmonary tuberculosis	Night sweats, upper zone disease with cavitation, positive acid-fast staining. It also causes bronchiectasis, so the two coexist.
Lung abscess	A single cavity with an air-fluid level, usually in a dependent segment, with an aspiration risk factor.
Empyema	Stony dullness with a pleural collection rather than a parenchymal cavity; pus on aspiration.
Lung cancer with post-obstructive infection	Older smoker, recurrent infection in the same lobe, a mass or collapse.
Cystic fibrosis	Upper lobe predominance, pancreatic insufficiency, raised sweat chloride.

4. Investigations

Establish the diagnosis

- **High-resolution computed tomography of the chest is the diagnostic test.** Look for a bronchoarterial ratio greater than one — the **signet ring sign** — failure of the airways to taper towards the periphery, and visible airways within one centimetre of the pleura.
- **Chest radiograph** — tramline and ring shadows, crowded markings, and in abscess a **cavity with an air-fluid level**. A normal film does not exclude bronchiectasis.
- **The distribution is a clue to the cause:** upper lobes in cystic fibrosis, tuberculosis and allergic bronchopulmonary aspergillosis; lower lobes in post-infective and aspiration disease; the right middle lobe and lingula in non-tuberculous mycobacterial infection.

Establish the organism

- Sputum for Gram stain and culture, **three specimens for acid-fast bacilli**, and fungal culture. Know the colonising organism: Haemophilus influenzae early, and **Pseudomonas aeruginosa** later, which marks more severe disease and changes every future antibiotic choice.

Establish the underlying cause

- Complete blood count with differential, immunoglobulins G, A and M, and specific antibody responses; human immunodeficiency virus test; total and aspergillus-specific immunoglobulin E and aspergillus precipitins; sweat chloride and cystic fibrosis genotyping where suggestive; rheumatoid factor and anti-cyclic citrullinated peptide antibodies; alpha-1 antitrypsin level.
- **Spirometry** — usually an obstructive pattern; used to follow progression.
- **Bronchoscopy** if disease is confined to one lobe, if a foreign body or tumour is possible, or for persistent haemoptysis.

5. Management

Treat the exacerbation

- Send sputum, then start antibiotics guided by previous isolates. **Treat for 14 days**, considerably longer than for pneumonia.
- If colonised with Pseudomonas: oral ciprofloxacin, or intravenous piperacillin–tazobactam or ceftazidime for severe episodes.
- For a lung abscess: **4–6 weeks of antibiotics with anaerobic cover** — co-amoxiclav or clindamycin — with postural drainage. Most resolve medically; percutaneous or surgical drainage is reserved for failure. **Arrange dental assessment**, since the mouth is usually the source.

The long-term programme — where the real benefit lies

- **Daily airway clearance physiotherapy** is the cornerstone of treatment. Active cycle of breathing, postural drainage, and oscillating positive expiratory pressure devices. Teach it, and check she is still doing it at every visit.
- Nebulised isotonic or hypertonic saline before physiotherapy to aid clearance.
- **Long-term azithromycin** three times weekly reduces exacerbation frequency. Exclude non-tuberculous mycobacterial infection and check the corrected QT interval and liver function before starting.
- Treat the underlying cause — immunoglobulin replacement in antibody deficiency, corticosteroids in allergic bronchopulmonary aspergillosis, modulator therapy in cystic fibrosis.
- Bronchodilators if there is demonstrable reversibility, vaccination, nutritional support, pulmonary rehabilitation, and surgical resection for localised disease that remains refractory.

EMERGENCY: MASSIVE HAEMOPTYSIS

Bronchiectasis is a common cause of massive haemoptysis, and it kills by asphyxiation rather than by exsanguination.

Protect the airway. Lie the patient with the bleeding side downwards to keep blood out of the good lung. Give oxygen, obtain large-bore access, correct clotting, and give tranexamic acid. Call for urgent bronchoscopy and **bronchial artery embolisation**, which is the definitive treatment; surgery if that fails.

6. Teaching Points and Viva Questions

- Clubbing with crackles is never simple chronic obstructive pulmonary disease. Think suppurative lung disease, fibrosis or cancer.
- The volume of daily sputum is the single most discriminating point in the history.
- High-resolution computed tomography makes the diagnosis; the plain film may be normal.
- Foul-smelling sputum means anaerobes, which means aspiration — so examine the teeth and ask why she aspirated.
- Physiotherapy is not an adjunct to treatment. In bronchiectasis it *is* the treatment, and antibiotics are the adjunct.

Questions you should be able to answer:

- How do you distinguish bronchiectasis from chronic obstructive pulmonary disease at the bedside?
- Name six causes of bronchiectasis and the single question that screens for each.
- Why does the distribution of disease on the computed tomogram matter?
- Why is Pseudomonas colonisation significant?
- She coughs up 400 mL of blood in an hour. What are your first five actions?

Case 5 · Acute-on-Chronic Hypercapnic Respiratory Failure

CLINICAL VIGNETTE

A **58-year-old man** is brought in by his wife because he has become increasingly drowsy over two days, following a week of cough and fever. For the past year she has noticed loud snoring with pauses in his breathing at night, and he has been falling asleep during the day and waking with headaches. His ankles have been swelling.

He takes no sedatives. He has never smoked.

On arrival: he is rousable to voice but falls asleep between questions. Temperature 37.8 °C, heart rate 96/min, respiratory rate 14/min and shallow, blood pressure 146/88 mmHg, oxygen saturation 86% on room air. Jugular venous pressure is raised and there is pitting oedema to mid-shin. A flapping tremor is present.

Arterial blood gas on air: pH 7.30, carbon dioxide 8.8 kPa, oxygen 7.4 kPa, bicarbonate 33 mmol/L.

1. The Framework

Respiratory failure is not a diagnosis. It is a physiological state with two patterns, and the whole of the assessment follows from deciding which one is in front of you.

	Type 1 — hypoxaemic	Type 2 — hypercapnic
Definition	Oxygen below 8 kPa with a normal or low carbon dioxide	Carbon dioxide above 6.0 kPa, usually with hypoxaemia
Mechanism	The lung fails as a gas exchanger — ventilation–perfusion mismatch or shunt	The pump fails — alveolar ventilation is inadequate
Alveolar–arterial gradient	Raised	Normal in pure hypoventilation; raised if there is lung disease as well
Typical causes	Pneumonia, pulmonary oedema, pulmonary embolism, acute respiratory distress syndrome, pneumothorax, interstitial lung disease	See the three questions below

For type 2 failure, ask three questions in order. They generate the entire differential.

- **Will he not breathe?** Central depression — opioids, benzodiazepines, alcohol, brainstem stroke, raised intracranial pressure, hypothyroidism.
- **Can he not breathe?** Pump failure — neuromuscular disease (Guillain–Barré syndrome, myasthenia gravis, motor neurone disease, muscular dystrophy), phrenic nerve palsy, or chest wall restriction from kyphoscoliosis, circumferential burns or obesity hypoventilation.
- **Is the load too great?** Airway and lung disease — chronic obstructive pulmonary disease, severe asthma late in an attack, bronchiectasis, advanced fibrosis.

2. Focused History

- Speed of onset. Minutes suggests drugs, a neurological event or a pneumothorax; days suggests infection on a chronic background; months suggests progressive neuromuscular or chest wall disease.
- **Symptoms of chronic hypoventilation:** morning headache, daytime somnolence, poor concentration, disturbed sleep, snoring with witnessed apnoeas, ankle swelling.

- **Neuromuscular screening questions:** difficulty rising from a chair or climbing stairs, double vision, slurred speech, choking on food, breathlessness on lying flat (a diaphragmatic sign), and recent gastrointestinal or respiratory infection preceding weakness.
- Every sedative, opioid, alcohol and recreational drug taken. Any oxygen given before arrival, and at what rate.
- Previous non-invasive ventilation or continuous positive airway pressure at home, and adherence to it.

3. Physical Examination

- **Conscious level**, scored and repeated — the most important single observation, and the one that decides whether non-invasive ventilation is safe.
- **Respiratory rate and pattern.** A high rate that falls as the patient tires is ominous. **Paradoxical inward abdominal movement on inspiration** indicates diaphragmatic fatigue or paralysis.
- **Bedside tests of respiratory muscle strength:** ask him to count aloud from one on a single breath, and test neck flexion strength. Both correlate with vital capacity and can be repeated hourly.
- Signs of retention — flapping tremor, bounding pulse, warm peripheries, papilloedema. Central cyanosis. Signs of cor pulmonale.
- Pupils and injection sites if opioids are possible; the chest wall for deformity; a full neurological examination when weakness is suspected.

4. Investigations

- **Arterial blood gas is the definitive investigation** and must be repeated. Record the inspired oxygen concentration every time or the result is uninterpretable.
- **Calculate the alveolar–arterial oxygen gradient.** A normal gradient with hypercapnia means pure hypoventilation — the lungs are healthy and the pump has failed. A raised gradient means there is lung disease as well. This single calculation separates the two mechanisms.
- Chest radiograph, electrocardiogram, complete blood count (polycythaemia in chronic hypoxaemia), urea and electrolytes, C-reactive protein, thyroid function, creatine kinase.
- **In suspected neuromuscular disease, measure the forced vital capacity serially and act on it.** A value below 15–20 mL/kg, or a fall of more than 30% from baseline, indicates the need for ventilatory support. **Do not wait for the carbon dioxide to rise — in neuromuscular failure hypercapnia is a late and pre-terminal finding.**
- Later, when the acute episode has settled: sleep studies, spirometry with lung volumes, and echocardiography for pulmonary hypertension.

5. Management

The oxygen ladder — and why the device matters

Device	When to use it
Nasal cannula, 1–4 L/min	Mild hypoxaemia. Delivered concentration varies with the patient's own breathing pattern.
Simple face mask, 5–10 L/min	Moderate hypoxaemia without a risk of retention. Never run below 5 L/min — the patient rebreathes carbon dioxide.
Venturi mask, 24–60%	The correct choice whenever the delivered concentration matters, because it provides a fixed concentration independent of the breathing pattern.
Reservoir mask, 15 L/min	Critical illness and trauma while the cause is being addressed. Not for a patient at risk of retention.
High-flow nasal oxygen	Type 1 failure needing high concentrations; provides humidification and some positive pressure.
Non-invasive ventilation	Type 2 failure with acidaemia. Bilevel support for hypercapnia; continuous positive airway pressure for cardiogenic pulmonary oedema and obstructive sleep apnoea.
Invasive ventilation	Failure of non-invasive support, inability to protect the airway, or a decision that full escalation is appropriate.

Sequence for this patient

- **Controlled oxygen** by Venturi mask, targeting a saturation of 88–92%, with a gas repeated in 30 minutes.
- **Reverse what is reversible:** treat the chest infection, give bronchodilators if there is any airflow obstruction, and give naloxone if opioids are conceivable.
- **Non-invasive ventilation** for the persisting acidaemia, since the pH is below 7.35 with a carbon dioxide above 6.5 kPa. His drowsiness makes this a decision for a senior clinician in a monitored area, because the same drowsiness raises the risk of aspiration.
- **Document the ceiling of care** and discuss it with the patient and family before he deteriorates further.
- Repeat the gas at one hour. Improvement in pH is the marker of success. No improvement means intubation or an explicit change of plan.

Complications and longer-term care

- Aspiration, barotrauma and pneumothorax, pressure injury from the mask, hypotension from positive pressure, and oxygen-induced hypercapnia.
- After recovery: sleep-disordered breathing assessment and home nocturnal ventilation, weight management referral, avoidance of sedatives, treatment of cor pulmonale, and long-term oxygen assessment when stable.

6. Teaching Points and Viva Questions

- Hypercapnia is a pump problem. Ask in order: will he not breathe, can he not breathe, or is the load too great?
- The alveolar–arterial gradient separates a failing lung from a failing pump, and costs nothing to calculate.
- In neuromuscular weakness, measure the vital capacity. By the time the carbon dioxide rises, the patient is close to arrest.
- A falling respiratory rate in a previously tachypnoeic patient means exhaustion, not recovery.
- Write the target saturation range on the chart. An unqualified instruction to "give oxygen" is how patients like this one become drowsy.

Questions you should be able to answer:

- Classify this man's respiratory failure and justify it from the gas.
- His bicarbonate is 33. What does that tell you about the timescale?
- Give four contraindications to non-invasive ventilation.
- Why is a Venturi mask preferred to nasal cannulae here?
- A patient with Guillain–Barré syndrome has a normal blood gas but a vital capacity of 1.1 litres. What do you do?

Case 6 · Pleural Effusion

CLINICAL VIGNETTE

A **62-year-old man** presents with six weeks of progressive breathlessness and a dull ache in the right side of his chest. He has lost 8 kg without trying and sweats at night. He worked for twenty years in construction and demolition. He is an ex-smoker of 30 pack-years.

On examination: respiratory rate 22/min, oxygen saturation 93% on air. The trachea is central. Expansion is reduced on the right. Percussion is **stony dull** to the mid-zone on the right, with absent breath sounds and reduced vocal resonance below, and bronchial breathing at the upper level of the dullness. There is no clubbing and no palpable lymphadenopathy.

1. Focused History

The two questions that shape everything

- **How quickly did it come on?** Days suggests infection, embolism or trauma. Weeks to months suggests malignancy, tuberculosis or heart failure.
- **Is it painful?** Pleuritic pain means the parietal pleura is inflamed and favours an exudate — infection, infarction, malignant invasion. Painless effusions are more often transudates.

Then screen for each cause directly

Ask about	Cause it points to
Orthopnoea, paroxysmal nocturnal dyspnoea, ankle swelling	Heart failure — the commonest cause overall, and usually bilateral
Jaundice, alcohol, known liver disease, abdominal swelling	Cirrhosis with hepatic hydrothorax
Frothy urine, facial and leg oedema	Nephrotic syndrome
Weight loss, night sweats, haemoptysis, smoking	Malignancy
Occupational asbestos exposure, with a latency of 20–40 years	Mesothelioma or benign asbestos-related effusion — ask about the whole working life, including construction, shipyards, insulation and brake linings
Tuberculosis contact, endemic exposure, prior treatment	Tuberculous pleurisy — typically a young patient with a lymphocytic exudate
Recent pneumonia with persisting or returning fever	Parapneumonic effusion or empyema
Immobility, surgery, malignancy, unilateral leg swelling	Pulmonary embolism with infarction
Joint pain, rash, photosensitivity, dry eyes	Rheumatoid arthritis or systemic lupus erythematosus
Severe epigastric pain, alcohol, gallstones	Pancreatitis

2. Physical Examination

Confirm the effusion

- Reduced expansion, **stony dullness**, absent or greatly reduced breath sounds, reduced vocal resonance and fremitus, with **bronchial breathing at the upper border** where compressed lung lies beneath.
- **Tracheal position:** central in a moderate effusion; pushed **away** by a massive one. If the trachea is pulled **towards** a white-out, the diagnosis is collapse, not effusion — and the management is entirely different.

Then find the cause

- Cervical and supraclavicular lymph nodes, breast examination, clubbing, chest wall masses or tenderness (mesothelioma), radiotherapy marks or surgical scars.
- Raised jugular venous pressure, third heart sound, bilateral oedema (heart failure); jaundice, spider naevi, ascites (cirrhosis); periorbital oedema (nephrotic syndrome); synovitis and deformity (rheumatoid disease); an abdominal or pelvic mass.

3. Investigations

Imaging — reading the film

- On the **posteroanterior radiograph**, about 200 mL is needed before the costophrenic angle blunts; on the **lateral** film as little as 50 mL is visible. Expect a homogeneous opacity with a concave upper border — the **meniscus sign**.
- A complete white-out: **look at the trachea**. Pushed away means effusion or a large mass; pulled towards means collapse or pneumonectomy; central suggests consolidation.
- **Thoracic ultrasound before every pleural procedure.** It confirms fluid, estimates depth, identifies septations, and marks the site. It has substantially reduced the complication rate of pleural aspiration and its use is now the standard of care.
- **Contrast-enhanced computed tomography** for any undiagnosed exudate, performed **before** complete drainage so the pleural surfaces can be assessed. Nodular or circumferential pleural thickening, thickening greater than one centimetre, or mediastinal pleural involvement all suggest malignancy.

Diagnostic aspiration

Note the appearance first: straw-coloured, turbid, frank pus, blood-stained, or milky. Then send the fluid for protein, lactate dehydrogenase, glucose, pH, Gram stain and culture, acid-fast staining and culture, adenosine deaminase, cytology, and triglycerides if it looks milky. Send **paired serum protein and lactate dehydrogenase at the same time** — without them the fluid results cannot be interpreted.

LIGHT'S CRITERIA

The fluid is an **exudate** if any one of the following is true:

pleural protein divided by serum protein is greater than 0.5 · pleural lactate dehydrogenase divided by serum lactate dehydrogenase is greater than 0.6 · pleural lactate dehydrogenase is greater than two-thirds of the upper limit of the normal serum range.

The known trap: in a patient on diuretics, a transudate concentrates and is misclassified as an exudate. When the clinical picture is heart failure but the criteria say exudate, calculate the **serum-to-fluid albumin gradient**; a value above 12 g/L confirms a transudate.

Transudate — the pressures are wrong	Exudate — the pleura is diseased
Heart failure (usually bilateral, right greater than left)	Parapneumonic effusion and empyema
Cirrhosis with hepatic hydrothorax	Malignancy — lung, breast, lymphoma, mesothelioma
Nephrotic syndrome and hypoalbuminaemia	Tuberculosis
Peritoneal dialysis	Pulmonary embolism with infarction
Hypothyroidism, constrictive pericarditis	Connective tissue disease, pancreatitis, drugs, chylothorax

Interpreting the fluid

Finding	What it means
pH below 7.2 in an infected effusion	Complicated parapneumonic effusion — insert a chest drain. This single number decides the management.
Low glucose (below 3.3 mmol/L)	Empyema, rheumatoid disease, tuberculosis, or malignancy.
Frank pus, or organisms on Gram stain	Empyema — drain regardless of the pH.
Lymphocyte predominance	Tuberculosis, malignancy, lymphoma, chronic disease.
Neutrophil predominance	Acute process — parapneumonic, pulmonary embolism, pancreatitis.
Adenosine deaminase above 40 U/L with a lymphocytic exudate	Strongly supports tuberculosis, particularly in a younger patient in an endemic setting.
Heavily blood-stained fluid	Malignancy, pulmonary infarction, or trauma.
Milky fluid with triglycerides above 1.24 mmol/L	Chylothorax — thoracic duct disruption from lymphoma, surgery or trauma.

- **Cytology is positive in only about six in ten malignant effusions.** A negative result does not exclude cancer. If two aspirates are non-diagnostic and suspicion persists, proceed to **local anaesthetic thoracoscopy with pleural biopsy**, which has the highest yield in both malignancy and tuberculosis.
- **Do not aspirate bilateral effusions in a patient with clear heart failure.** Treat the failure and reassess; tap only if the effusions are asymmetric, painful, febrile, or fail to resolve.

4. Management

- **Treat the cause.** Transudates resolve when the underlying disorder is treated — diuresis for heart failure, albumin and sodium restriction in cirrhosis. Drainage alone achieves nothing lasting.
- **Therapeutic drainage** for symptomatic relief. **Remove no more than 1–1.5 litres at one sitting**, and stop immediately for chest tightness, pain or persistent coughing, because of the risk of **re-expansion**

pulmonary oedema.

- **Empyema or complicated parapneumonic effusion:** prompt chest drain plus antibiotics covering anaerobes, continued for two to six weeks. Intrapleural fibrinolytic therapy for loculated collections, and surgical decortication when drainage fails.
- **Malignant effusion:** treat the underlying cancer. For recurrent symptomatic effusions, offer either an **indwelling pleural catheter** for drainage at home or **talc pleurodesis**; the choice depends on performance status, lung re-expansion and patient preference.
- **Tuberculous effusion:** standard antituberculous chemotherapy; most resolve without drainage.
- **Procedural complications:** pneumothorax, bleeding, infection, injury to liver or spleen, and re-expansion oedema. Ultrasound guidance, a properly positioned patient and a limited volume prevent most of them.

5. Teaching Points and Viva Questions

- Stony dullness with absent breath sounds is fluid. Dullness with bronchial breathing is consolidation. This distinction changes the next step from a prescription to a procedure.
- In a complete white-out, the trachea tells you the answer before any test does.
- Never perform a pleural procedure without ultrasound, and never on a patient who is sitting unsupported.
- Light's criteria misclassify diuretic-treated heart failure. Remember the albumin gradient.
- A lymphocytic exudate with a high adenosine deaminase in a young patient in this region is tuberculosis until proved otherwise.
- An occupational history taken properly can be the single most valuable part of this consultation, and it must cover the whole working life.

Questions you should be able to answer:

- State Light's criteria and explain the one situation in which they mislead.
- The pleural fluid pH is 7.05 after a pneumonia. What do you do, and why does the pH matter?
- List five causes of a lymphocytic exudative effusion.
- You drain 2.5 litres and the patient becomes acutely breathless. What has happened?
- Cytology is negative twice but you still suspect mesothelioma. What is your next investigation?

Case 7 · Lung Cancer

CLINICAL VIGNETTE

A **66-year-old man** with a 50 pack-year smoking history says his cough has **changed** over three months — it is now dry, harsher, and worse at night. He has coughed up streaks of blood on four occasions. He has lost 9 kg without trying, has a persistent dull ache between the right shoulder blade and the spine, and **his voice has been hoarse for six weeks**.

On examination: cachectic and hoarse. There is **finger clubbing**. A firm 2 cm node is palpable in the left supraclavicular fossa. Percussion is dull over the right upper zone with reduced breath sounds. No stridor.

Sodium 122 mmol/L · corrected calcium normal · haemoglobin 106 g/L. Chest radiograph: right hilar mass with right upper lobe collapse.

THE HISTORY THAT MATTERS

A smoker who has coughed for years does not present because he has a cough. He presents — if anyone asks — because **the cough has changed**.

The features that should trigger urgent investigation: a new cough or a change in a chronic cough lasting more than three weeks · **haemoptysis** · unexplained weight loss · chest or shoulder pain · breathlessness · **hoarseness** · a chest infection that fails to resolve · **new finger clubbing** · cervical or supraclavicular lymphadenopathy.

And a normal chest radiograph does not exclude lung cancer. In a patient with haemoptysis and weight loss, a normal film means you need a computed tomogram, not reassurance.

1. Focused History

Local invasion — each symptom localises the tumour

Symptom or sign	Structure involved
Hoarseness	Recurrent laryngeal nerve — not laryngitis. In a smoker with weight loss this is a red flag, and it also implies mediastinal involvement
Dysphagia	Oesophageal compression
Facial and arm swelling with distended chest wall veins, worse on bending forward	Superior vena caval obstruction
Shoulder and inner arm pain, wasting of the small hand muscles, Horner syndrome	Pancoast tumour at the apex, involving the brachial plexus and sympathetic chain
Breathlessness with stony dullness	Pleural effusion — or pericardial effusion with tamponade
Elevated hemidiaphragm	Phrenic nerve palsy

Metastatic and paraneoplastic disease

- **Metastases:** bone pain, headache, seizure or focal neurological deficit, jaundice or right upper quadrant pain, skin nodules.

Paraneoplastic syndrome	Usual histology
Syndrome of inappropriate antidiuresis — hyponatraemia, as here	Small cell
Ectopic adrenocorticotrophic hormone — rapid-onset Cushing syndrome with severe hypokalaemia	Small cell
Hypercalcaemia from parathyroid hormone-related peptide	Squamous cell
Lambert–Eaton myasthenic syndrome, cerebellar degeneration, limbic encephalitis	Small cell
Clubbing and hypertrophic pulmonary osteoarthropathy	Adenocarcinoma and squamous
Dermatomyositis; migratory thrombophlebitis and hypercoagulability; anaemia	Any

So the biochemistry hints at the histology before the biopsy does. Hyponatraemia points towards small cell disease; hypercalcaemia towards squamous.

Risk and context

- **Smoking quantified in pack-years, including waterpipe**, and second-hand exposure. **Asbestos** with its decades of latency, radon, silica, arsenic, chromium, diesel exhaust, and **biomass cooking smoke and heavy incense use**, which are relevant exposures here.
- Previous thoracic radiotherapy, chronic obstructive pulmonary disease, pulmonary fibrosis, human immunodeficiency virus, and family history.
- **Performance status, comorbidity and current lung function** — these determine what treatment is possible at least as much as the stage does. And ask, early and gently, what the patient understands and wants to know.

2. Physical Examination

- Cachexia; **clubbing and hypertrophic pulmonary osteoarthropathy**; tar staining; hoarseness; **Horner syndrome and wasting of the thenar and interosseous muscles**.
- **Cervical and supraclavicular nodes** — a palpable supraclavicular node is often the easiest route to tissue and should be sought before anything invasive is planned.
- Tracheal position; signs of collapse, consolidation or effusion; signs of superior vena caval obstruction.
- Liver edge, bone tenderness, skin nodules, and a full neurological examination.

3. Investigations

- **Chest radiograph:** mass, hilar enlargement, lobar collapse, effusion, bony lesion, elevated hemidiaphragm. Then **contrast-enhanced computed tomography of the chest, abdomen and pelvis**, which must include the adrenals and liver.
- **Positron emission tomography with computed tomography** for staging in anyone being considered for treatment with curative intent, and **magnetic resonance imaging of the brain** where indicated.
- **Tissue is essential, and the route depends on the location:** bronchoscopy with endobronchial ultrasound and mediastinal nodal sampling for central lesions; computed tomography-guided biopsy for

peripheral ones; pleural aspiration and biopsy; or biopsy of that supraclavicular node.

THE MOST IMPORTANT PRACTICAL CHANGE IN MODERN LUNG CANCER CARE

Take enough tissue for molecular testing, and say so on the request.

Treatment of advanced non-squamous non-small cell lung cancer now depends on **EGFR, ALK, ROS1, BRAF and KRAS status and on PD-L1 expression**, because a patient with a targetable mutation may respond to an oral tyrosine kinase inhibitor for years.

An inadequate sample means a second biopsy, or a patient given chemotherapy when a tablet would have served them better. This is a real change from the way lung cancer was managed a decade ago, and it is the commonest reason a diagnostic procedure has to be repeated.

- **Bloods:** complete blood count, **sodium**, corrected calcium, liver function with alkaline phosphatase, lactate dehydrogenase, renal function.
- **Before surgery:** spirometry with gas transfer, and cardiopulmonary exercise assessment where lung function is marginal.

4. The Division That Determines Everything

	Non-small cell (around 85%)	Small cell (around 15%)
Types	Adenocarcinoma, squamous cell, large cell	Small cell carcinoma
Behaviour	Grows and spreads more slowly; often localised at diagnosis	Aggressive, and effectively systemic at diagnosis even when imaging looks limited
Role of surgery	Yes — curative resection for early stage disease	Essentially no
Mainstay	Surgery, stereotactic radiotherapy, chemoradiotherapy, and molecularly guided systemic therapy or immunotherapy in advanced disease	Platinum-based chemotherapy with radiotherapy, plus immunotherapy in extensive stage; prophylactic cranial irradiation in selected patients
Course	Variable	Marked initial response, then early relapse

5. Management

- **Every case goes to a multidisciplinary team**, and the decision turns on histology, molecular profile, stage, lung function and performance status together.
- **Non-small cell:** lobectomy for early disease with adequate reserve; stereotactic ablative radiotherapy for the medically inoperable; concurrent chemoradiotherapy for locally advanced disease; and for advanced disease, **a targeted inhibitor where a driver mutation is present, or immunotherapy guided by PD-L1**, with or without chemotherapy.
- **Small cell:** chemotherapy with thoracic radiotherapy in limited stage, chemoimmunotherapy in extensive stage.

Emergency	Action
Malignant spinal cord compression	Back pain with leg weakness, a sensory level or sphincter disturbance. Urgent magnetic resonance imaging of the whole spine, high-dose dexamethasone, and same-day oncology and neurosurgical discussion. Hours, not days.
Superior vena caval obstruction	Rarely an immediate airway threat. Obtain tissue where possible before corticosteroids, then treat with radiotherapy, chemotherapy or stenting.
Hypercalcaemia	Fluids then a bisphosphonate — Case 23.
Massive haemoptysis	Airway protection, bleeding side down, bronchial artery embolisation — Case 4.
Pericardial tamponade; symptomatic brain metastases	Drainage; dexamethasone and urgent imaging.

THE REFERRAL MADE TOO LATE

Refer to palliative care at diagnosis in advanced disease, not when treatment options run out.

Early specialist palliative care alongside oncological treatment **improves quality of life, reduces depression, and in advanced lung cancer has been shown to improve survival**. It is not a signal that treatment has been abandoned, and it should not be presented to the patient as one.

Practical measures that make a real difference: opioids for breathlessness and pain, pleurodesis or an indwelling pleural catheter for recurrent effusion, endobronchial stenting or debulking for large airway obstruction, palliative radiotherapy for haemoptysis and bone pain, nutritional support, and **smoking cessation — which still improves outcomes and tolerance of treatment even after diagnosis**.

6. Teaching Points and Viva Questions

- It is the change in the cough, not the cough, that matters.
- A normal chest radiograph does not exclude lung cancer.
- Hoarseness in a smoker with weight loss is nerve involvement, not laryngitis.
- Hyponatraemia suggests small cell; hypercalcaemia suggests squamous.
- Small cell versus non-small cell decides whether surgery is even on the table.
- Take enough tissue for molecular testing, or the patient may be denied the best treatment.
- Palliative care belongs at diagnosis, and it improves survival.

Questions you should be able to answer:

- What does his hoarseness tell you anatomically, and what does it imply about stage?
- His sodium is 122. What is the mechanism and which histology does it suggest?
- Which single physical finding offers the easiest route to a diagnosis, and why does that matter?
- The biopsy confirms adenocarcinoma but there is insufficient tissue for molecular studies. What are the consequences?
- He develops mid-thoracic back pain and difficulty walking. What is your sequence of actions?

Case 8 · Pulmonary Hypertension

CLINICAL VIGNETTE

A **34-year-old woman** describes eighteen months of breathlessness on exertion which has progressed from hills to walking on the flat. She becomes dizzy on exertion and fainted once while climbing stairs. Over the past two months her ankles have swollen and her abdomen has become distended.

She has been treated for asthma with inhalers that made no difference, and was told last year that the problem was probably anxiety.

On examination: oxygen saturation 94% on air. **The jugular venous pressure is raised with a prominent a wave.** There is a **left parasternal heave and a loud pulmonary second sound**, with a pansystolic murmur at the left sternal edge that is **louder on inspiration**, giant systolic venous waves and a **pulsatile liver**. There is ascites and pitting oedema to the knees. **The chest is clear.**

Electrocardiogram: right axis deviation, right ventricular hypertrophy, P pulmonale. **Echocardiography:** dilated hypertrophied right ventricle, high estimated pulmonary artery pressure, severe tricuspid regurgitation, normal left ventricle. **Right heart catheterisation:** mean pulmonary artery pressure 48 mmHg, raised pulmonary vascular resistance, normal wedge pressure.

WHY THIS DIAGNOSIS IS MISSED FOR YEARS

The median delay from first symptom to diagnosis in pulmonary arterial hypertension remains **more than two years**, and this history shows exactly why: a young woman with breathlessness and a clear chest is labelled asthmatic, unfit, overweight or anxious.

Progressive unexplained exertional breathlessness with a clear chest and a loud pulmonary second sound needs an echocardiogram. That is the whole lesson of this case, and it costs one investigation.

Exertional dizziness and syncope are not vague symptoms here — they mean the right ventricle cannot increase output on demand, and they carry a poor prognosis.

1. Think First — Five Groups, and the Group Decides the Treatment

Group	Causes
1 — Pulmonary arterial hypertension	Idiopathic · heritable · drug-induced (appetite suppressants, some stimulants) · connective tissue disease, especially systemic sclerosis · human immunodeficiency virus · portal hypertension · congenital heart disease · schistosomiasis, which is one of the commonest causes worldwide
2 — Left heart disease	The commonest cause overall — heart failure with reduced or preserved ejection fraction, valve disease. The wedge pressure is raised
3 — Lung disease and hypoxia	Chronic obstructive pulmonary disease, pulmonary fibrosis, obstructive sleep apnoea, hypoventilation, and chronic residence at high altitude
4 — Chronic thromboembolic disease	The group that may be surgically curable — which is why every patient must be asked about previous venous thromboembolism and screened for it
5 — Multifactorial	Sarcoidosis · haematological disease including myeloproliferative neoplasms and chronic haemolysis in sickle cell disease and thalassaemia · metabolic and renal disease

2. Focused History

- **Breathlessness on exertion is the earliest and often the only symptom.** Then fatigue, exertional chest pain, **presyncope and syncope**, palpitations, and finally ankle swelling and abdominal distension as the right ventricle fails.

- Hoarseness from compression of the left recurrent laryngeal nerve by a dilated pulmonary artery.
- **Screen for each group by history:** cardiac symptoms and hypertension; known lung disease, smoking, snoring and daytime somnolence, **altitude of residence**; **previous deep vein thrombosis or pulmonary embolism**; Raynaud phenomenon, skin tightening and reflux for systemic sclerosis; liver disease; congenital heart disease and murmurs in childhood; appetite suppressant or stimulant use; **freshwater exposure in a schistosomiasis-endemic area**; sickle cell disease or thalassaemia.
- Family history — heritable disease is autosomal dominant with incomplete penetrance.
- **Pregnancy plans and contraception**, which must be discussed at the first consultation for the reason given below.

3. Physical Examination

THE SIGNS

Every physical sign in pulmonary hypertension is on the right side of the heart, and the lungs are clear. That combination is the diagnosis.

Raised jugular venous pressure with a prominent a wave (a hypertrophied right atrium contracting hard) · **left parasternal heave** · **loud pulmonary component of the second sound** · right ventricular third or fourth sound · pulmonary regurgitation · **tricuspid regurgitation with giant v waves and a pulsatile liver** · ascites and peripheral oedema.

- **Then examine for the cause:** sclerodactyly, telangiectasia, calcinosis and Raynaud changes; fine crackles of fibrosis; clubbing; stigmata of chronic liver disease; central cyanosis and shunt murmurs; body habitus and neck circumference.

4. Investigations

- **Echocardiography is the screening test** — right ventricular size and function, estimated pulmonary artery pressure, tricuspid regurgitation, and exclusion of left heart disease and intracardiac shunts.
- **Right heart catheterisation is required to confirm the diagnosis and is mandatory before any targeted therapy is started.** Pre-capillary disease is defined by a **mean pulmonary artery pressure above 20 mmHg with a raised pulmonary vascular resistance and a normal wedge pressure** — the wedge is what separates group 1 from group 2, and it cannot be inferred from an echocardiogram.
- Electrocardiogram — right axis deviation, right ventricular hypertrophy, P pulmonale, right bundle branch block. Chest radiograph — enlarged central pulmonary arteries with pruned peripheral vessels and right ventricular enlargement.

THE ONE TEST NOT TO GET WRONG

Use a ventilation–perfusion scan, not computed tomographic pulmonary angiography, to screen for chronic thromboembolic disease.

Chronic organised thrombus is flat, wall-adherent and easily missed on a computed tomogram, whereas the perfusion defects it causes are conspicuous on a ventilation–perfusion scan.

This matters more than any other investigation in the workup, because chronic thromboembolic pulmonary hypertension is potentially cured by pulmonary endarterectomy — and a missed diagnosis means a curable patient is treated palliatively with vasodilators for years.

- **The rest of the workup:** pulmonary function tests with gas transfer; high-resolution computed tomography; overnight oximetry or sleep study; autoimmune screen; human immunodeficiency virus test; liver function and abdominal ultrasound; **schistosomiasis serology where exposure is plausible**; and genetic testing where the history suggests heritable disease.

- **For risk stratification and follow-up:** functional class, **six-minute walk distance**, and N-terminal pro-B-type natriuretic peptide. Risk category, reassessed at every visit, drives escalation of therapy.

5. Management

- **Refer to a specialist pulmonary hypertension centre.** The diagnosis requires catheterisation and the targeted drugs require specialist prescribing and monitoring; this is not a condition to manage alone.

THE PRESCRIBING ERROR TO AVOID

Targeted pulmonary vasodilators are for group 1 and group 4. They are harmful in group 2 and generally not indicated in group 3.

In pulmonary hypertension due to left heart disease, vasodilating the pulmonary circulation increases flow into a left ventricle that cannot accept it, and precipitates pulmonary oedema. In lung disease, they worsen ventilation–perfusion matching and hypoxaemia.

So the group must be established before a drug is prescribed — which is another reason the wedge pressure matters. Treat groups 2 and 3 by treating the heart or the lung disease.

- **Group 4: refer for assessment for pulmonary endarterectomy, which may be curative**; lifelong anticoagulation; balloon pulmonary angioplasty and riociguat for inoperable disease.

- **Group 1 targeted therapy:** endothelin receptor antagonists, phosphodiesterase-5 inhibitors, riociguat and prostacyclin pathway agents. **Most patients now start on combination therapy**, escalated according to risk. **Calcium channel blockers are used only in the small minority shown to be vasoreactive at catheterisation** — given empirically they cause harm.

- **Supportive care:** diuretics for right heart failure, oxygen for hypoxaemia, iron replacement, supervised exercise rehabilitation, vaccination, and caution with anaesthesia — these patients tolerate hypotension and hypoxia very badly, and the anaesthetist must be told.

- **Lung transplantation** for disease refractory to maximal therapy, referred before the patient becomes too unwell to be a candidate.

THE CONVERSATION THAT MUST HAPPEN EARLY

Pregnancy in pulmonary arterial hypertension carries a very high maternal mortality, because the circulatory changes of pregnancy and delivery cannot be met by a failing right ventricle.

Discuss effective contraception at the first consultation — bearing in mind that combined oral contraceptives raise thrombotic risk and that some targeted drugs, notably endothelin receptor antagonists, are teratogenic and reduce hormonal contraceptive efficacy.

A woman who becomes pregnant needs immediate specialist multidisciplinary care, not a routine antenatal clinic.

6. Teaching Points and Viva Questions

- Progressive breathlessness with a clear chest and a loud pulmonary second sound: request an echocardiogram.

- All the signs are right-sided, which is why the chest sounds normal.
- Exertional syncope means the right ventricle cannot increase output — an ominous sign.
- Catheterisation confirms the diagnosis, and the wedge pressure assigns the group.
- Ventilation–perfusion scan, not computed tomography, to find the curable group.
- Do not give pulmonary vasodilators for left heart disease.
- Raise contraception at the first visit.

Questions you should be able to answer:

- Why is her chest clear, and why does that make the diagnosis rather than exclude it?
- What does the normal wedge pressure tell you, and what would a raised one have meant?
- Which imaging do you use to exclude chronic thromboembolic disease, and why not the other?
- A patient with heart failure and a preserved ejection fraction has pulmonary hypertension on echocardiography. Should you start sildenafil?
- She tells you she is hoping to start a family. What do you say?