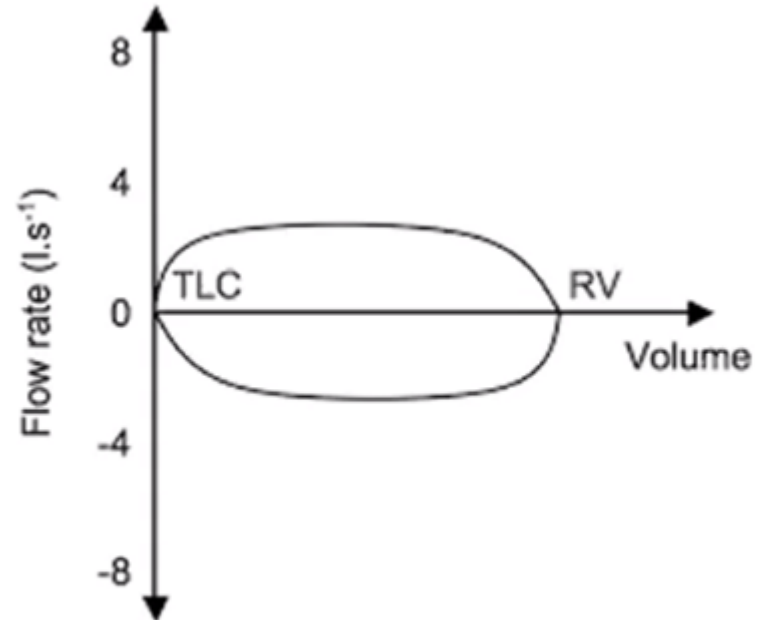


SCE practice Qs

Physiology for SCE

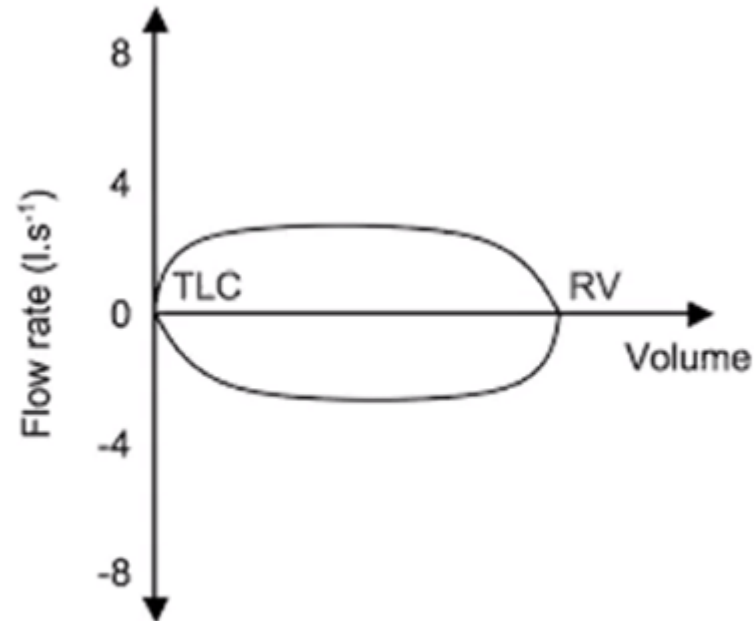
- 33 year old noisy breathing, SOB, with the following flow volume loop

- A. Vocal cord palsy
- B. Tracheomalacia
- C. Tracheal papillomatosis
- D. Tracheal stricture
- E. Asthma



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Fixed airway obstruction

Q

in unilateral diaphragmatic weakness which of the following is normal?

- A. Maximal expiratory pressure
- B. Maximal inspiratory pressure
- C. Sniff nasal inspiratory pressure
- D. VC in the sitting position
- E. VC in the supine position

Q

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- C. Sniff nasal inspiratory pressure
- D. VC in the sitting position
- E. VC in the supine position

Diaphragm is only used for inspiration so doesn't effect expiratory pressure. All the rest will be affected, although VC in sitting is reduced less than VC in supine it is will reduced.

A 55-year-old woman presented with noisy breathing and difficulty walking up stairs. She felt she had been slowing down recently. She had a 75 pack-year smoking history. She had kept a budgerigar since becoming a widow 8 years previously.

Investigations:

| lung function tests: | | | | |
|---------------------------------|--------------|-----------|-------|----------------------|
| | actual value | predicted | SR* | 90% confidence limit |
| FEV ₁ (L) | 2.35 | 2.54 | -0.51 | 1.9–3.2 |
| FVC (L) | 3.25 | 2.99 | +0.61 | 2.3–3.7 |
| FEV ₁ /FVC ratio | 72% | 85% | -0.96 | 68–89 |
| PEFR (L/s) | 3.30 | 6.32 | -3.36 | 4.8–7.8 |
| TL _{CO} (mmol/min/kPa) | 6.50 | 8.06 | -1.34 | 6.1–10.0 |

*SR = standardised residual. Normal range for all parameters is -1.64 to +1.64.

What is the most likely diagnosis?

Answers

- A: COPD
- B: hypersensitivity pneumonitis
- C: idiopathic pulmonary fibrosis
- D: myopathy
- E: upper airway obstruction

- This question shows a markedly reduced PEFr compared to the reduction in FEV₁. The FEV₁/PEFr gives an Empey index >10, in keeping with upper airway obstruction ($=2350 / (3.3 \times 60) = 11.9$).

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What is the most likely diagnosis?

Answers

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- B: hypersensitivity pneumonitis
- C: idiopathic pulmonary fibrosis
- D: myopathy
- E: upper airway obstruction

Correct answer: E

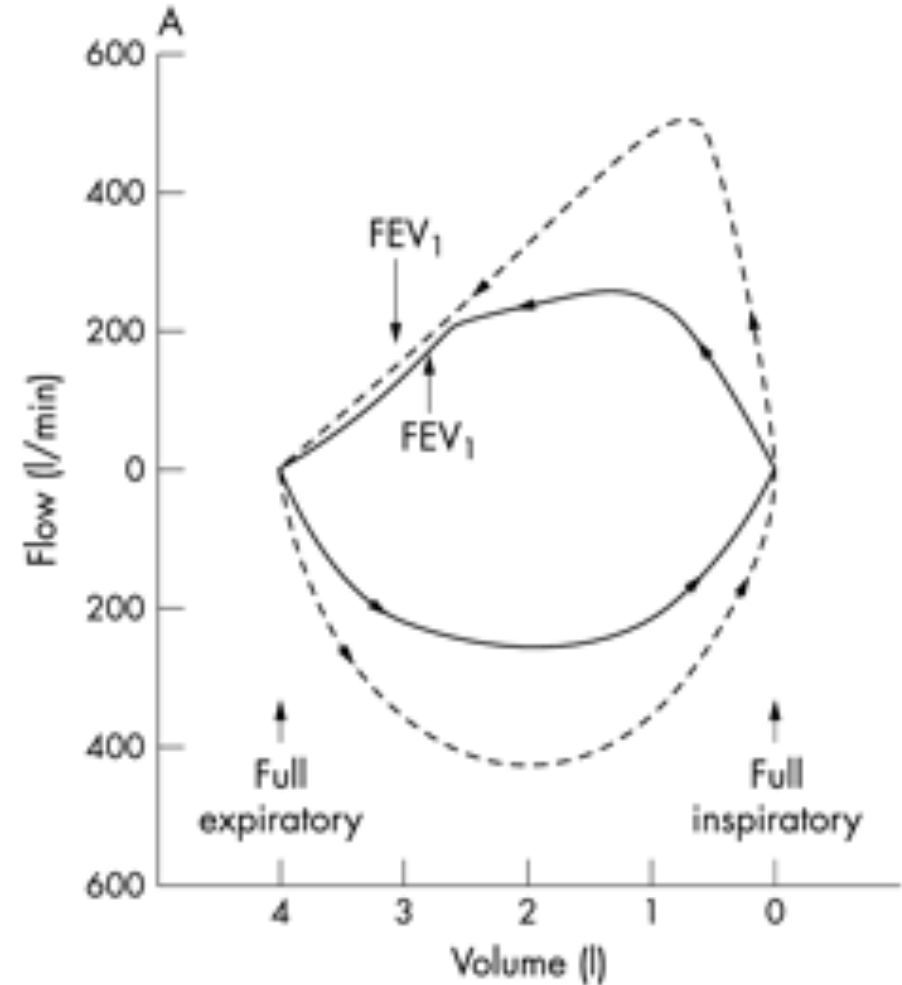
Explanation

This question shows a markedly reduced PEFr compared to the reduction in FEV₁. The FEV₁/PEFr gives an Empey index >10, in keeping with upper airway obstruction ($=2350 / (3.3 \times 60) = 11.9$).

Patient presents with noisy breathing and SOB with WL. They are diagnosed with fixed airflow obstruction and you fear tracheal obstruction.

What is the most accurate Empey index for this patient?

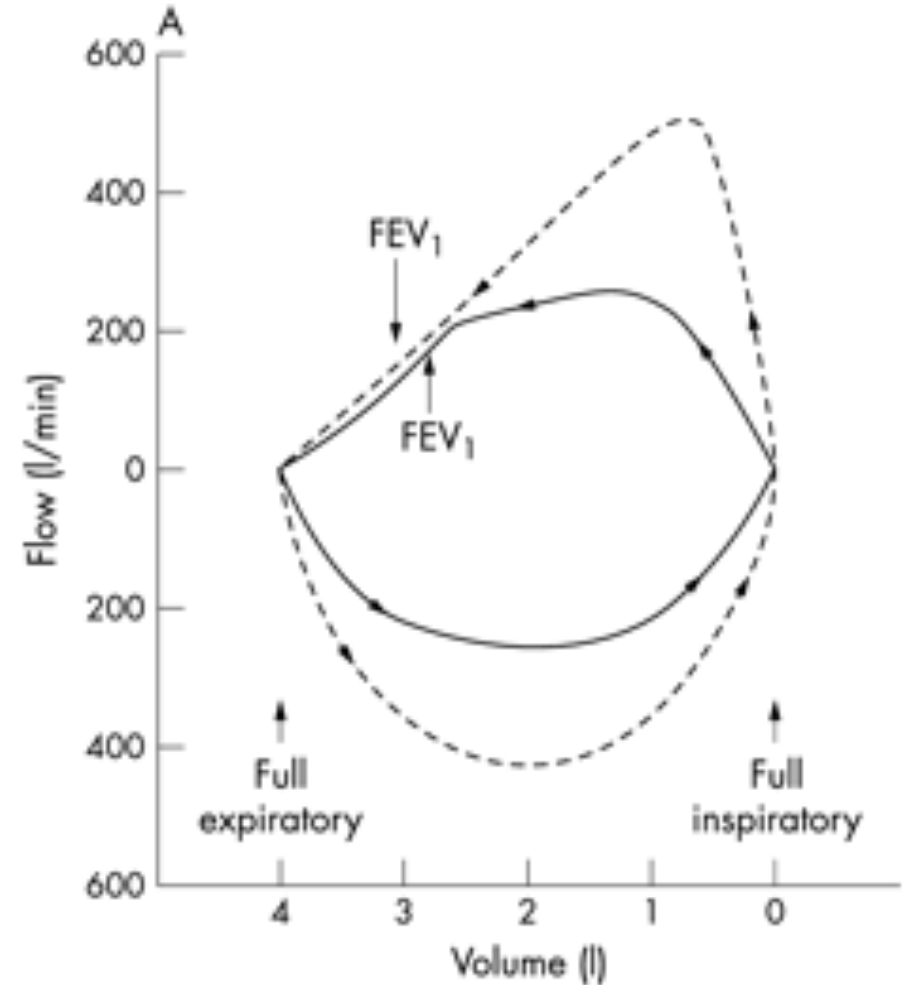
- A. $>10\%$
- B. $>5\%$
- C. $<5\%$
- D. $<10\%$



Patient presents with noisy breathing and SOB with WL. They are diagnosed with fixed airflow obstruction and you fear tracheal obstruction.

What is the most accurate Empey index for this patient?

- A. >10%
- B. >5%
- C. <5%
- D. <10%



Answer explained

- The Empey index, is a way of predicting if a patient has upper airways obstruction and can be performed simply at the bedside. It is the calculation of the ratio of FEV_1 (ml):PEFR (l/min);
 - a normal person will have a ratio of less than 10
 - while a person with upper airways obstruction will have a ratio greater than 10,
 - the higher the index the more severe the obstruction. The index has been well validated on a variety of patients with upper airways obstruction but also in patients with other forms of lung disease including asthma and emphysema.

What is the calculation for the empey index

- An indication of obstruction to the upper airways (trachea and larynx) may be obtained by calculating the ratio of the forced expired volume in one second to the peak expiratory flow rate ($FEV_1/PEFR$).
- This index was found to be usually less than 10 in normal subjects (mean 7.3), and in patients with asthma (mean 6.9), chronic bronchitis (mean 7.7), or interstitial lung disease (mean 6.3).
- A study of simulated upper airways obstruction showed that this index rises as the obstruction becomes more severe. All of 18 patients with proved upper airways obstruction had $FEV_1/PEFR$ indices greater than 10 (mean 14.0).
- This test can be carried out with forced expiratory manoeuvres only, and it does not require the use of complicated equipment.
- An $FEV_1/PEFR$ ratio greater than 10, when upper airways obstruction is suspected, indicates that significant obstruction may be present. High values suggest that the obstruction may be severe, and that further investigations are indicated.

Physiology basics

- Over what time period is PEFR measures?
 - A. 0.2 sec
 - B. 0.02 sec
 - C. 0.002 sec
 - D. 0.5
 - E. 1 sec
- At what volume of lung is expiration most effort dependent?
 - A. Maximum volume a person can manage
 - B. Minimum volume a person can manage
 - C. FEF 75
 - D. FEF 50
 - E. FEF 25

Physiology basics

- Over what time period is PEFR measures? **2 milliseconds**
 - A. 0.2 sec
 - B. 0.02 sec
 - C. 0.002 sec**
 - D. 0.5
 - E. 1 sec
- At what volume of lung is expiration most effort independent?
 - A. Maximum volume a person can manage
 - B. Minimum volume a person can manage**
 - C. FEF 75
 - D. FEF 50
 - E. FEF 25

PEFR is measure in the first two milliseconds of a maximal expiration, that is, at high lung volumes. FEV₁ is in contrast measure over a range of diminishing lung volumes, over 1 second. FEV₁ is therefore less dependent on effort and upper airway resistance than PEFR. That is why in emphysema PEFR can be relatively less reduced compared with the FEV₁. At smaller lung volumes expiration is more and more effort dependent. In asthma and chronic obstructive pulmonary disease both FEV₁ and PEFR are affected

FEV₁ is little affected by the presence of upper airways obstruction. FEV₁ is measured over one second—that is, over a range of diminishing lung volumes, it is less dependent on effort and upper airways resistance. PEFR is measured in the first two milliseconds of a maximal expiration—that is, at high lung volumes. Hence in upper airways obstruction PEFR is much more affected than the FEV₁. In asthma and chronic obstructive pulmonary disease both FEV₁ and PEFR are affected, and in emphysema the peak flow is relatively less reduced compared with the FEV₁.

In a normal adult, what most accurately describes the relationship of forced expiratory flow rate to effort?

Answers

- A: entirely effort-dependent
- B: entirely effort-independent
- C: independent of lung volume at maximal effort
- D: more effort-independent at higher lung volumes
- E: more effort-independent at lower lung volumes

Correct answer: E

Explanation

Expiration depends upon the action of the expiratory muscles coupled with lung compliance. Answers A (entirely effort-dependent) and B (entirely effort-independent) can therefore be excluded. The rate of expiration depends also upon lung volume, so Answer C (independent of lung volume at maximal effort) is incorrect. Although compliance is much less than the force generated by the respiratory musculature, it is greater at lower lung volumes when it will contribute more to the flow rate than at higher lung volumes. Thus, the expiratory flow rate depends less on the musculature (effort independent) at lower lung volumes and E (more effort-independent at lower lung volumes) is the correct answer.

A 72-year-old man presented with breathlessness.

Investigations:

arterial blood gases, breathing air:

| | |
|------------------|---------------------|
| PO ₂ | 5.9 kPa (11.3–12.6) |
| PCO ₂ | 4.0 kPa (4.7–6.0) |
| pH | 7.47 (7.35–7.45) |
| H ⁺ | 34 nmol/L (35–45) |
| bicarbonate | 24 mmol/L (21–29) |

What is his alveolar–arterial oxygen gradient (in kPa)?

Answers

- A: 4.0–6.0
- B: 6.1–8.0
- C: 8.1–10.5
- D: 10.6–12.0
- E: 12.1–14.0

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- D: 10.6–12.0
- E: 12.1–14.0

Correct answer: C

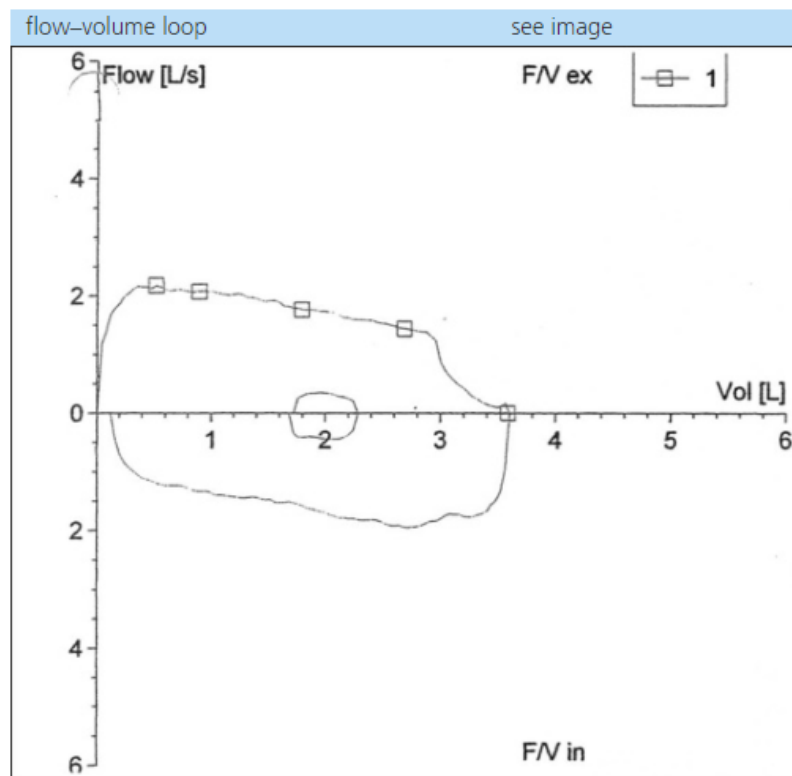
Explanation

A simplified version of the alveolar gas equation is $P_{aO_2} = (P_{iO_2} - (P_aCO_2/0.8)) - P_{aO_2}$, i.e. in this example $21 - 5 - 5.9 = 10.1$.

A 23-year-old woman, with mild asthma since childhood, complained of shortness of breath on exertion, which had become progressively worse over a few months. She also reported difficulty with her hearing over the previous week. Her usual inhalers had not improved her shortness of breath, but she was able to walk further after a 5-day course of prednisolone. She did not smoke, and worked as a clinical coder – a job she had found increasingly stressful recently.

On examination, she was anxious, with a respiratory rate of 32 breaths/min. When she was not speaking, her breathing sounded noisy. On auscultation of her chest, there were no wheezes or crackles.

Investigations:



What is the most likely diagnosis?

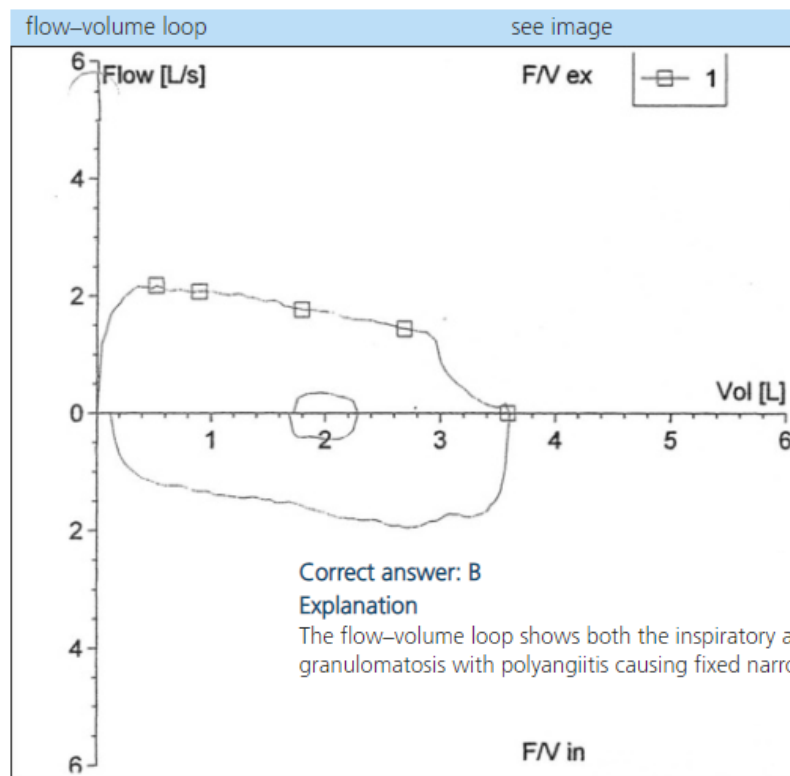
Answers

- A: asthma
- B: granulomatosis with polyangiitis
- C: tracheal tumour
- D: tracheomalacia
- E: vocal cord dysfunction

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Answers

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Q1.

- Patient suffer whiplash, complete right diaphragmatic nerve palsy.
- TLC reduced by 20%,
- What is his Peak flow reduced by?

- A. No change
- B. Increase in 10%
- C. Increase in 20%
- D. Decrease by 10%
- E. Decrease by 20%

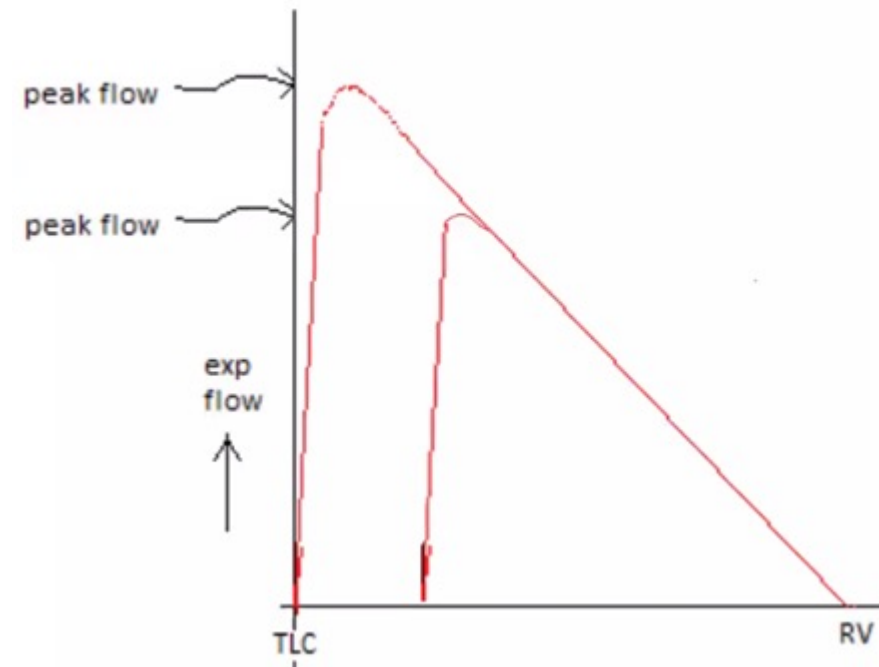
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Peak flow is reduced by approx. 20% as well

As lung volume is reduced the max possible expiratory flow is reduced (in an approx. linear way



SCE Q 2s for physiology

- In a normal adult (normal height and weight) with no underlying lung disease at the end of a normal breath (functional residual capacity), what is the volume of air left in the lungs?

1. Female 1.5L male 2L
2. Female 2.5L, male 3L
3. Female 3L, male 3.5L
4. Female 1L, male 1.5L

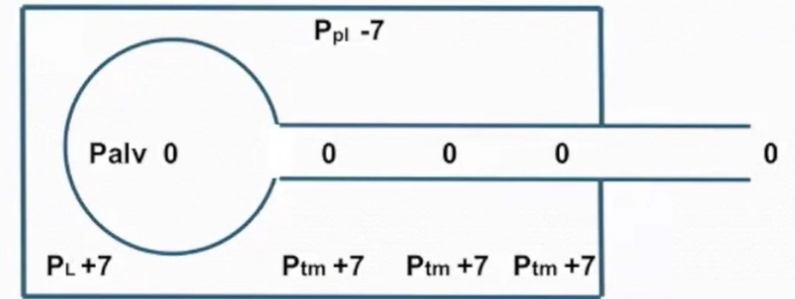
Q2 Answer

- Answer = 2
- Functional residual capacity is the point at the end of normal tidal breathing where the elastic recoil of the lungs wanting to collapse inwards matches the elastic recoil of the chest wall wanting to expand outwards.
- In a female the volume of air at this point is approx 2.5L and in a male is 3L

SCE Q 3 for physiology

- In a normal subject at the end of a tidal breath (functional residual capacity) with their mouth open - what is the normal alveolar pressure (P_{alv}), intrapleural pressure (P_{pl}) and transmural pressure? (P_{tm})
1. $P_{alv} = 0 \text{ cm/H}_2\text{O}$ $P_{pl} = -7 \text{ cm/H}_2\text{O}$ $P_{tm} = +7 \text{ cm/H}_2\text{O}$
 2. $P_{alv} = 7 \text{ cm/H}_2\text{O}$ $P_{pl} = 0 \text{ cm/H}_2\text{O}$ $P_{tm} = +10 \text{ cm/H}_2\text{O}$
 3. $P_{alv} = 10 \text{ cm/H}_2\text{O}$ $P_{pl} = -7 \text{ cm/H}_2\text{O}$ $P_{tm} = 0 \text{ cm/H}_2\text{O}$
 4. $P_{alv} = 0 \text{ cm/H}_2\text{O}$ $P_{pl} = -10 \text{ cm/H}_2\text{O}$ $P_{tm} = -10 \text{ cm/H}_2\text{O}$

Answer Q3



Palv = alveolar pressure

Ppl = pleural pressure

P_L = lung recoil pressure (Palv-Ppl)

Ptm = transmural airway pressure

(all pressures cmH₂O)

- **Answer = 1**
- If you stop breathing at the end of a normal breath (functional residual capacity) with your mouth open, the pressure of air in lungs will be the same as in the atmosphere 0cm/H₂O
- Because your chest wall wants to spring outwards and your lungs want to collapse inwards there will always be a negative pressure between the chest wall and the lung. This is called the interpleural pressure. This is normally -7cm/H₂O. so if you stuck a tube in the pleural space at this point you would see water pushed a tube by 7cm.
- Transmural pressure means the lung recoil pressure. You take alveolar pressure (which is 0 as per atmosphere)- intrapleural pressure which is -7. $0 - (-7) = +7$ cm/H₂O

SCE Q 4s for physiology

- In COPD what benefit does purse lip breathing offer to the patients lung physiology?
 1. No physiological benefit, it purely gives patient a psychological benefit
 2. Purse lip breathing increased elastic recoil and aids expiration.
 3. Purse lip breathing improves ventilation by increasing PEEP and helps to keep the airways open and stops collapse to same degree. Helps create positive pressure in the airways and keeps them open.
 4. Purse lip breathing collapses the small airways and reduces ventilation but increased diffusion capacity.

Q4 Answer

- In COPD what benefit does purse lip breathing offer to the patients lung physiology?

Purse lip breathing improves ventilation by increasing PEEP and helps to keep the airways open and stops collapse to some degree. Helps create positive pressure in the airways and keeps them open.

Back pressure in airway, stops airways collapsing, improves the work of breathing

Q5

Do a Qs with 75 year old pre op spiro and FEV1
FEV1 1.87 (76% predicted; standard residual -1.54)
FVC 2.2.L(80% pred; standard residual -0.24)
FEV1/FVC ratio 0.66

- A. Bronchodilator reversibility testing is required
- B. The result are consistent with a diagnosis of COPD
- C. Patients >75 year old have a degree of airway obstruction
- D. The repeat testing should be arranged
- E. The results are within the normal range.

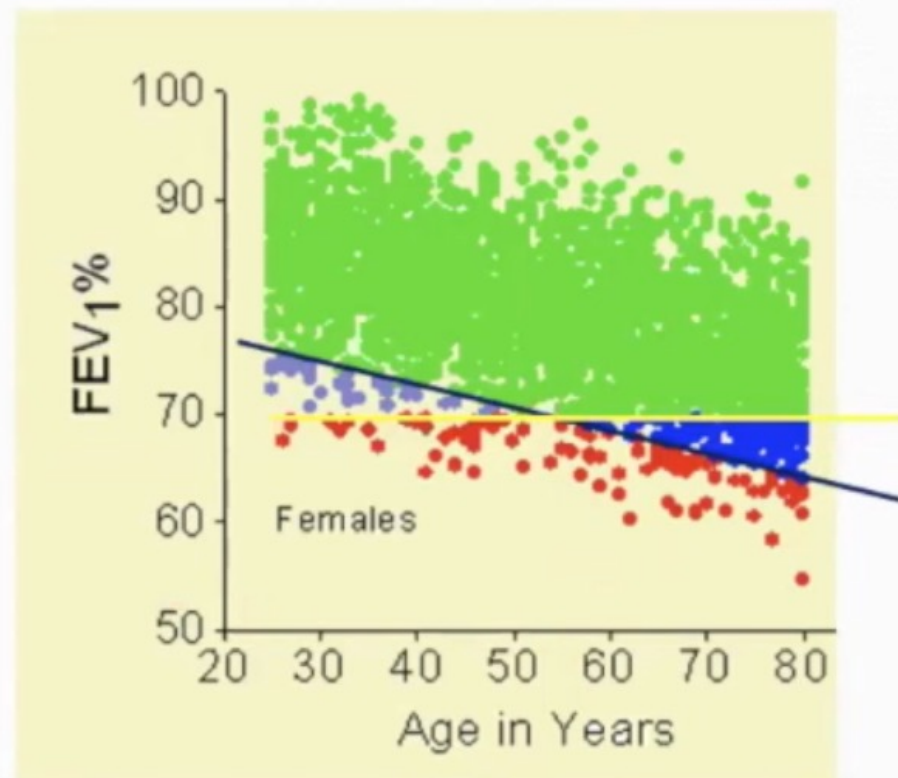
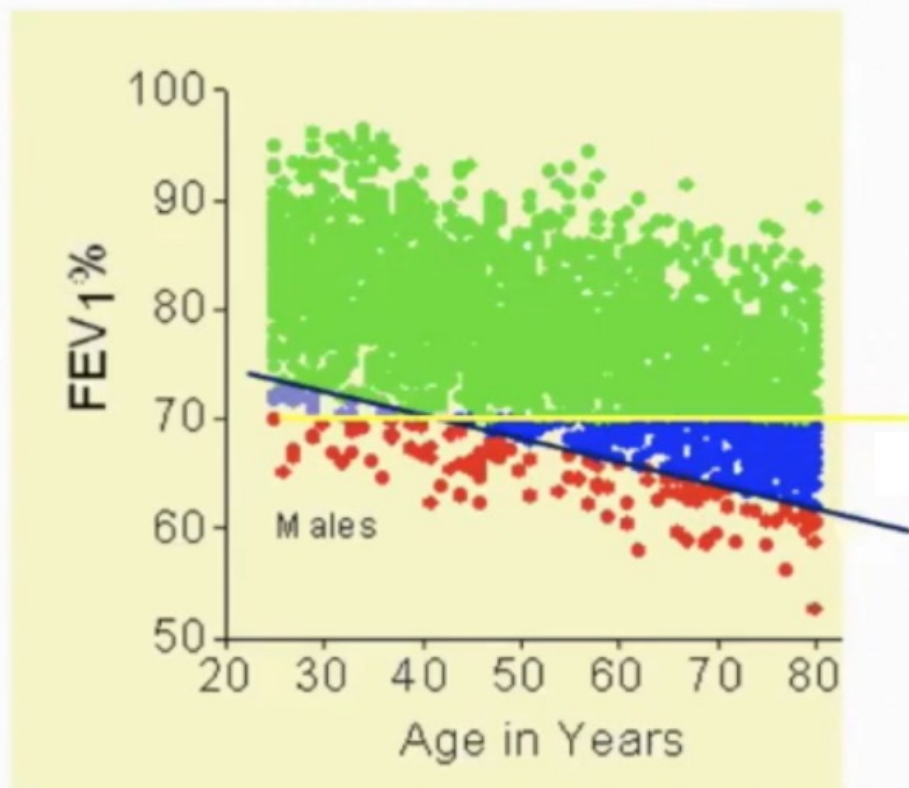
ans

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Problem with using fixed FEV1/FVC ratio of 0.7

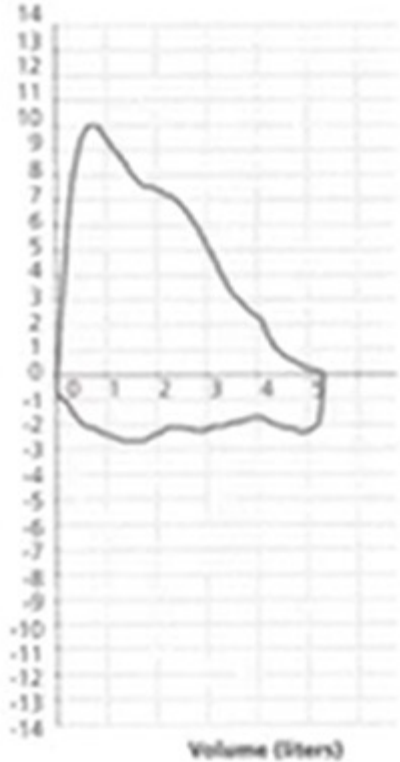
- We used to say that if u could blow 70% of your FVC in 1 sec u don't have airflow obstruction, bit random cut off,
- Problem of 0.7 as cut off in the elderly it will freq over diagnose them as obstructed.
- The LLN is down at 60% often in the elderly
- Also in young you will under diagnose airflow obstruction, so say a 21 year old blows out 71% of FVC this is too little for that age range.



Debating the definition of airflow obstruction: time to move on?
M. R. Miller , O. F. Pedersen, R. Pellegrino and V. Brusasco
ERJ September 1, 2009 vol. 34 no. 3 527-528

Q6

What is the most likely cause of this flow-volume loop?

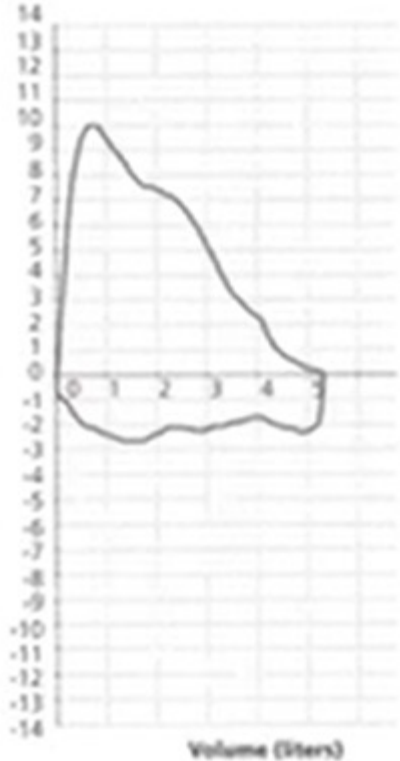


- A. Tracheomalacia
- B. Tumour of main bronchus
- C. Sub-glottic stenosis
- D. Polychondritis
- E. ILO /vocal cord dysfunction

Ans

What is the most likely cause of this flow-volume loop?

- A. Tracheomalacia (variable intrathoracic obstruction)
- B. Tumour of main bronchus (variable intrathoracic obstruction)
- C. Sub-glottic stenosis (fixed large airway obstruction)
- D. Polychondritis (intrathoracic obstruction)
- E. ILO /vocal cord dysfunction = Variable extra thoracic obstruction



Variable extra thoracic obstruction as inspiratory loop reduced

Large Airway Obstruction

A typical shape of the flow-volume loop is seen in cases of obstruction of the large airways.

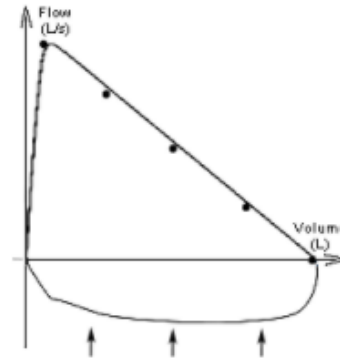
Three different shapes of flow-volume loops can be distinguished.

Variable Extrathoracic Obstruction

Typically the expiratory part of the F/V-loop is normal: the obstruction is pushed outwards by the force of the expiration.

During inspiration the obstruction is sucked into the trachea with partial obstruction and flattening of the inspiratory part of the flow-volume loop.

This is seen in cases of vocal cord paralysis, extrathoracic goiter and laryngeal tumors.



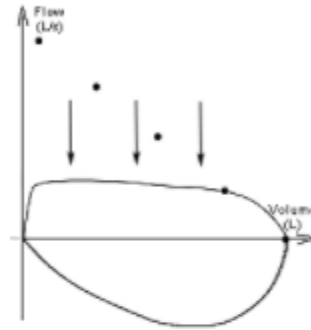
Variable Intrathoracic Obstruction

This is the opposite situation of the extrathoracic obstruction. A tumor located near the intrathoracic part of the trachea is sucked outwards during inspiration with a normal morphology of the inspiratory part of F/V-loop.

Variable Intrathoracic Obstruction

This is the opposite situation of the extrathoracic obstruction. A tumor located near the intrathoracic part of the trachea is sucked outwards during inspiration with a normal morphology of the inspiratory part of F/V-loop.

During expiration the tumor is pushed into the trachea with partial obstruction and flattening of the expiratory part of the F/V loop.

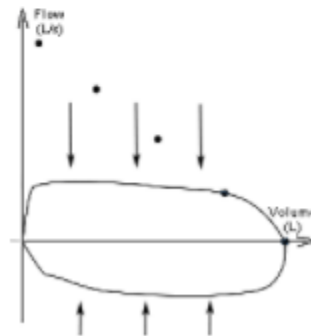


Fixed Large Airway Obstruction

This can be both intrathoracic as extrathoracic.

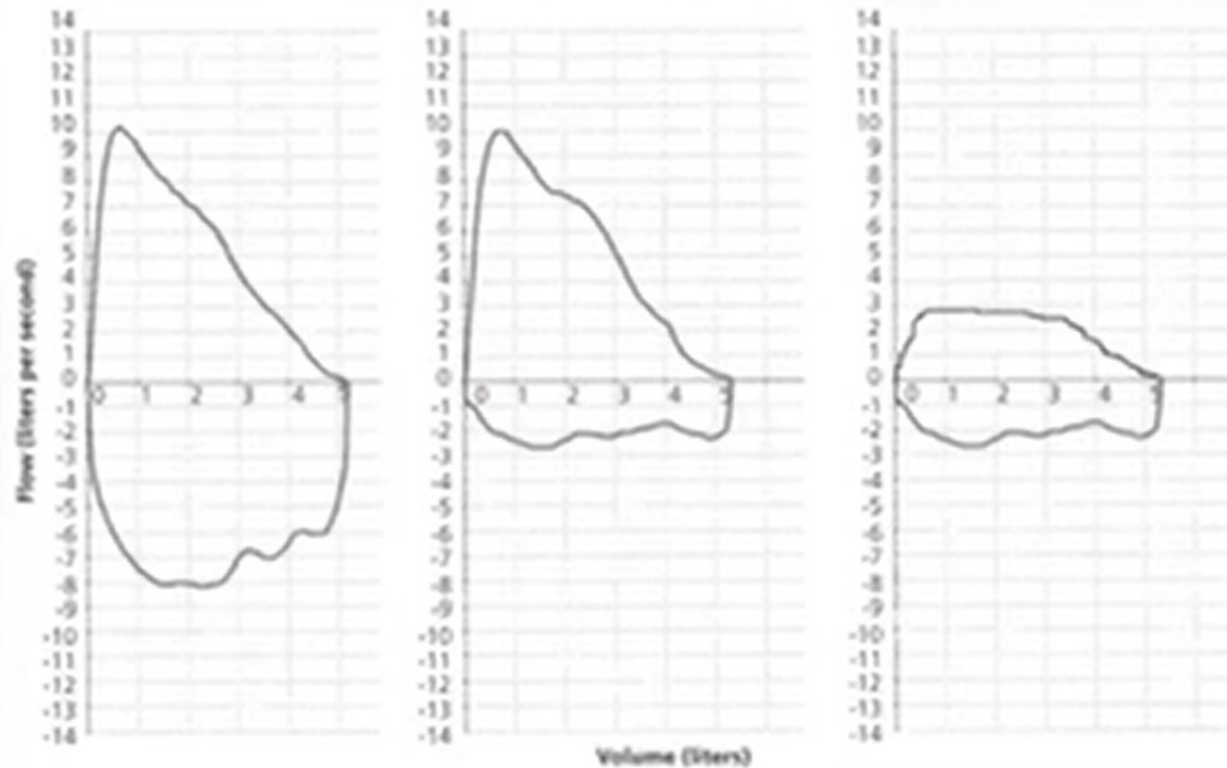
The flow-volume loop is typically flattened during inspiration and expiration.

Examples are tracheal stenosis caused by intubation and a circular tracheal tumor.



Typical flattening of flow-volume loop in fixed airway obstruction

Less common forms of obstruction



Normal

Variable extrathoracic

Fixed extrathoracic

Variable extra thoracic
obstruction – ILO,
tracheomalacia

Variable intrathoracic – lower
tracheobroncho malacia,
polychondritis, tumour of lower
trachea and main bronchus

Fixed extra thoracic obstruction
= goitre , vocal cord narrowing,
subglottic stenosis

Match the histology to the condition

1. Pigmented macrophages in terminal bronchioles

2. Pigmented macrophages in terminal alveoli

a) RBILD

b) DIP

Match the histology to the condition

1. Pigmented macrophages in terminal bronchioles

2. Pigmented macrophages in terminal alveoli

a) RBILD

RB-ILD – 1.

DIP - 2

b) DIP

Qs from SCE RCP

Q 1

A 75-year-old gentleman has a CT scan which shows a nodule of 5mm. For technical reasons, volumetric assessment was not possible on the CT scan. He is an ex-smoker, who stopped 10 years ago, with a 20-pack year history. He also has a background of ischaemic heart disease for which he is on aspirin. He has no respiratory symptoms at present and is living alone and has a WHO performance status of 0. What further follow up is required?

- a) Discharge – no further follow up required Repeat CT at 3 months
- b) Repeat CT at 3 months
- c) Repeat CT at 1 year from baseline
- d) Refer for PET scan
- e) Refer for biopsy

Q 1

A 75-year-old gentleman has a CT scan which shows a nodule of 5mm. For technical reasons, volumetric assessment was not possible on the CT scan. He is an ex-smoker, who stopped 10 years ago, with a 20-pack year history. He also has a background of ischaemic heart disease for which he is on aspirin. He has no respiratory symptoms at present and is living alone and has a WHO performance status of 0. What further follow up is required?

- a) Discharge <5mm
- b) Repeat CT at 3 months ≥6mm
- c) **Repeat CT at 1 year from baseline – 5mm**
- d) Refer for PET scan ≥8mm
- e) Refer for biopsy – Dependent on Herder Risk after PET and other factors

If <5mm = u can discharge

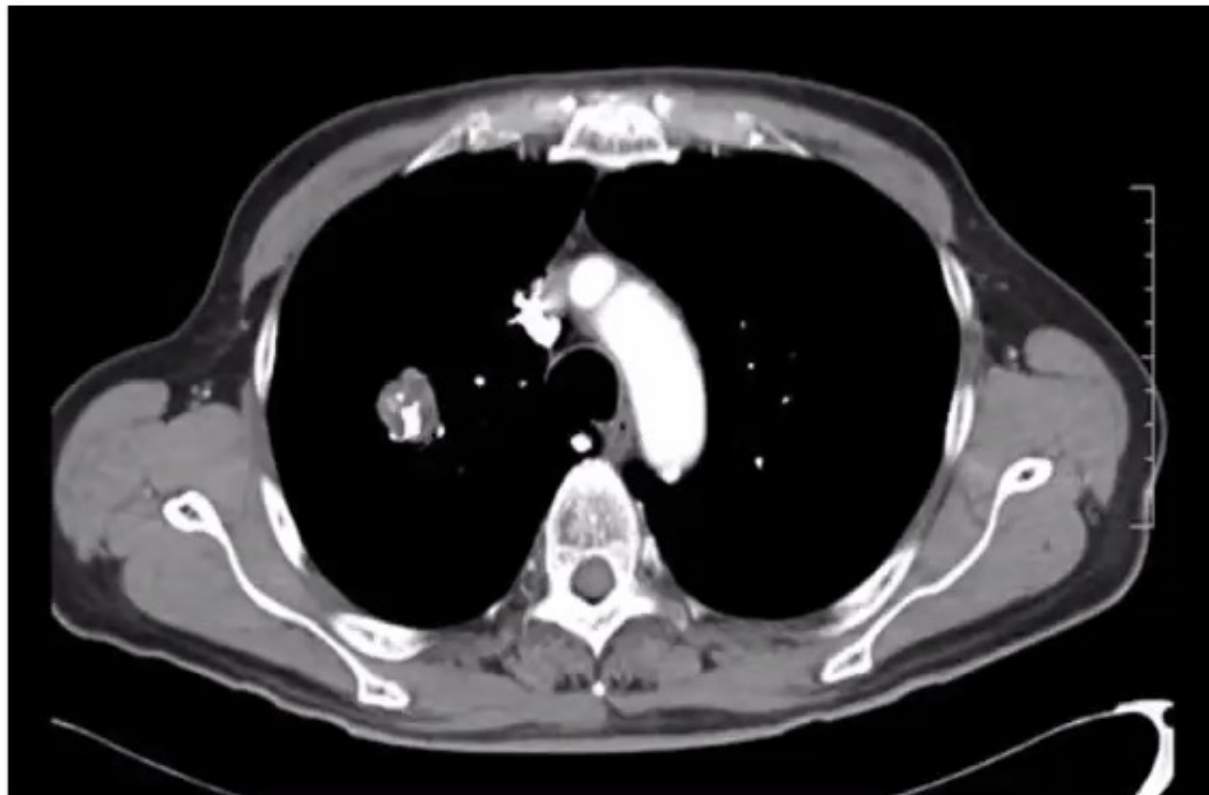
If 5-5.9mm = 1 year CT from baseline

>6mm = 3/12 CT

Q 2

A 55-year-old male undergoes a CT chest to further investigate a chronic cough (see below). What follow up is required?

- a) No follow up required
- b) Repeat CT chest at 3 months
- c) Repeat CT in 1 year
- d) Refer for biopsy
- e) Refer for PET scan

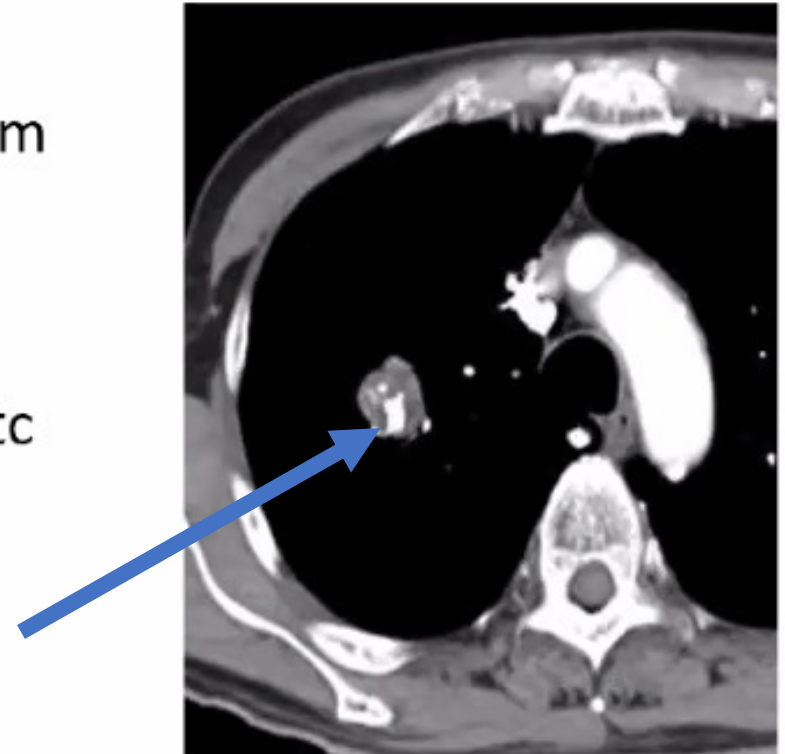


Q 2

A 55-year-old male undergoes a CT chest to further investigate a chronic cough (see below). What follow up is required?

- a) **No follow up required** – Benign features calcium
- b) Repeat CT chest at 3 months – 6-8mm
- c) Repeat CT in 1 year – 5mm
- d) Refer for biopsy – Dependent on Herder Risk etc
- e) Refer for PET scan $\geq 8\text{mm}$

Benign haemartoma



Q 3

A 60-year-old ex-smoker with a 20 pack year history undergoes a CT chest, which shows a solid nodule of 8mm diameter in the RUL with no evidence of spiculation. There is mild centrilobular emphysema on the scan. Brock score calculation shows a 4.1% risk of malignancy. What follow up is required?

- a) No follow up required
- b) CT chest at 3 months from baseline
- c) CT at 1 year from baseline and assess VDT
- d) Refer for biopsy
- e) Refer for excision

Q 3

A 60-year-old ex-smoker with a 20 pack year history undergoes a CT chest, which shows a solid nodule of 8mm diameter in the RUL with no evidence of spiculation. There is mild centrilobular emphysema on the scan. Brock score calculation shows a 4.1% risk of malignancy. What follow up is required?

- a) No follow up required <5mm
- b) CT chest at 3 months from baseline 6-8mm**
- c) CT at 1 year from baseline and assess VDT 5mm
- d) Refer for biopsy – Dependent on Herder Risk after PET and other factors
- e) Refer for excision – Dependent on Herder Risk after PET and other factors

Why not PET? For PET:

- Size >8mm (sensitivity of PET CT)
- Brock Risk $\geq 10\%$

Q 4

A 70-year-old gentleman is found to have a 10mm solid nodule in the RLL with evidence of spiculation. A Brock score is applied, showing a risk of 25.3%. He goes on to have a PET scan which shows moderate FDG avidity with a Herder probability of 70.7%. What is the optimal management?

- a) Biopsy
- b) Repeat CT at 1 year
- c) Repeat PET at 3 months
- d) Reassure and discharge
- e) Repeat CT at 3 months

Q 4

A 70-year-old gentleman is found to have a 10mm solid nodule in the RLL with evidence of spiculation. A Brock score is applied, showing a risk of 25.3%. He goes on to have a PET scan which shows moderate FDG avidity with a Herder probability of 70.7%. What is the optimal management?

- a) **Biopsy** >70% risk (or straight to treatment i.e. Surgery or RT)
- b) Repeat CT at 1 year
- c) Repeat PET at 3 months
- d) Reassure and discharge
- e) Repeat CT at 3 months – 10-70% risk Options = Biopsy, excision biopsy or CT surveillance dependent on individual risk and patient preference

Q 5

A 70-year-old gentleman has a CT chest to investigate left sided chest pain, which reveals a sub-solid nodule of 6mm. He undergoes a repeat CT at 3 months. Brock assessment shows a malignancy risk of 9%. What follow up is required?

- a) CT chest a 1 year, 2 year and 4 year
- b) CT chest at 1 year, then 2 year and then discharge if stable
- c) Image-guided biopsy
- d) Resection
- e) CT chest at 1 year, and discharge if stable

Q 5

A 70-year-old gentleman has a CT chest to investigate left sided chest pain, which reveals a sub-solid nodule of 6mm. He undergoes a repeat CT at 3 months. Brock assessment shows a malignancy risk of 9%. What follow up is required?

- a) **CT chest a 1 year, 2 year and 4 year** – Risk <10%
- b) CT chest at 1 year, then 2 year and then discharge if stable – 4 year F/U for sub-solid nodules
- c) Image-guided biopsy – Option if $\geq 10\%$ risk
- d) Resection – Option if $\geq 10\%$ risk
- e) CT chest at 1 year, and discharge if stable – 4 year F/U for sub-solid nodules

Q 6

An 80-year-old gentleman is referred to the Chest clinic with haemoptysis. He is an ex-smoker, with a 40-pack year history. His comorbidities include GORD and angina. He is on regular bisoprolol 2.5 mg OD and Tiotropium 18mcg OD. A CT chest is performed which shows a right sided 4cm tumour with enlarged right paratracheal lymph nodes. There is also a right sided pleural effusion which shows TTF1 positive adenocarcinoma cells. What stage is his malignancy?

- a) T1a N2 M1
- b) T2a N2 M1a
- c) T2a N3 M1c
- d) T3 N1 M1a
- e) T1 N2 M1b

Q 6

An 80-year-old gentleman is referred to the Chest clinic with haemoptysis. He is an ex-smoker, with a 40-pack year history. His comorbidities include GORD and angina. He is on regular bisoprolol 2.5 mg OD and Tiotropium 18mcg OD. A CT chest is performed which shows a right sided 4cm tumour with enlarged right paratracheal lymph nodes. There is also a right sided pleural effusion which shows TTF1 positive adenocarcinoma cells. What stage is his malignancy?

- a) T1a N2 M1
- b) T2a N2 M1a** – 3.1-4cm = T2a, 4R Node = N2, malignant effusion = M1a
- c) T2a N3 M1c
- d) T3 N1 M1a
- e) T1 N2 M1b

Q 7

A 65-year-old gentleman is referred to Chest clinic with cough and breathlessness. He is an ex-smoker, having stopped 10 years ago, with a 20-pack year history. He is otherwise, systemically well. A CT chest is performed, which shows a 2 cm right lower lobe lesion. An incidental finding of a raised right hemi-diaphragm is noted. There are no pleural or pericardial effusions. There is no evidence of any mediastinal lymphadenopathy. CT abdomen/ pelvis is unremarkable. What is the staging for this lesion?

- a) T3 N0 M0
- b) T2a N1 M1
- c) T2b N0 M0
- d) T1a N0 M0
- e) T1c N0 M1

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- a) **T3 N0 M0** – Raised hemidiaphragm \approx Phrenic nerve invasion = T3 irrespective of size
- b) T2a N1 M1
- c) T2b N0 M0
- d) T1a N0 M0
- e) T1c N0 M1

1.1-2cm = T1b which is not an option here, which helps!

Staging

All size less than or equal to.
T3 = 2cm increment
T4 >7cm

T3/T4 caveats

- T – All $\leq x$ cm
 - X – Cannot be assessed
 - 0 – No primary
 - is – Carcinoma in situ
 - 1a ≤ 1 cm – Can be T1a(min) = Minimally invasive
 - 1b 1.1-2 cm
 - 1c 2.1-3 cm
 - 2a 3.1-4 cm
 - 2b 4.1-5 cm
 - T3 5.1-7 cm
 - Or – Invading any of: parietal pleural, chest wall, phrenic nerve or parietal pericardium
 - Or – Separate tumour nodule(s) in same lobe as primary
 - T4 >7 cm
 - Or – Invading any of diaphragm, mediastinum, heart, great vessels, trachea, recurrent laryngeal nerve, oesophagus, vertebral body, carina
 - Or – Separate tumour nodule(s) in different ipsilateral lobe

- N
 - X – Cannot be assessed
 - 0 = No nodal disease
 - 1 = Ipsilateral peribronchial and/or perihilar lymph nodes and intrapulmonary nodes
 - 2 = Ipsilateral mediastinal and/or subcarinal lymph nodes
 - 3 = Contralateral mediastinal/hilar, ipsilateral or contralateral scalene or supraclavicular nodes
- M
 - 0 = No metastases
 - 1a =
 - Contralateral lung i.e. not T3 or T4 nodules
 - Pericardial or pleural nodules/effusions
 - 1b = Single extra-thoracic metastasis
 - 1c = >1 extra-thoracic metastasis

Stage Groupings

Nodes at least stage 2
M = stage 4

| T/M | Subgroup | N0 | N1 | N2 | N3 |
|-----|----------|------|------|------|------|
| T1 | T1a | IA | IIB | IIIA | IIIB |
| | T1b | IA | IIB | IIIA | IIIB |
| | T1c | IA | IIB | IIIA | IIIB |
| T2 | T2a | IB | IIB | IIIA | IIIB |
| | T2b | IIA | IIB | IIIA | IIIB |
| T3 | T3 | IIB | IIIA | IIIB | IIIC |
| T4 | T4 | IIIA | IIIA | IIIB | IIIC |
| M1 | M1a | IVA | IVA | IVA | IVA |
| | M1b | IVA | IVA | IVA | IVA |
| | M1c | IVB | IVB | IVB | IVB |

Q 8

A 55-year-old gentleman presents with cough and breathlessness. CT chest shows a 2 cm lesion in the right upper lobe, with accompanying subcarinal lymphadenopathy of 11mm and enlarged right hilar LNs of 12mm. What is the most appropriate next investigation?

- a) PET-CT
- b) Lung function tests
- c) EBUS- FNA of nodes
- d) CT head
- e) CT-guided biopsy of lesion

Q 8

A 55-year-old gentleman presents with cough and breathlessness. CT chest shows a 2 cm lesion in the right upper lobe, with accompanying subcarinal lymphadenopathy of 11mm and enlarged right hilar LNs of 12mm. What is the most appropriate next investigation?

- a) **PET-CT**
- b) Lung function tests
- c) EBUS- FNA of nodes
- d) CT head
- e) CT-guided biopsy of lesion

Do A (PET) first to help guide your EBUS



Q 9

A 60-year-old gentleman is referred with a right lower lobe lesion on CT chest. It is 2cm in size, with no local invasion. There is no evidence of any pleural effusions or distant metastases. Right hilar lymphadenopathy with hilar nodes measuring up to 12mm are noted. Biopsy of the right hilar lymph node shows features of adenocarcinoma. He is an ex-smoker with a 10-pack year history. He usually lives alone and is a retired accountant. What would be the most appropriate treatment option?

- a) Chemotherapy alone
- b) Chemo-radiotherapy combination
- c) Surgery alone
- d) Surgery with adjuvant chemotherapy
- e) Neoadjuvant chemotherapy followed by surgery

Q 9

A 60-year-old gentleman is referred with a right lower lobe lesion on CT chest. It is 2cm in size, with no local invasion. There is no evidence of any pleural effusions or distant metastases. Right hilar lymphadenopathy with hilar nodes measuring up to 12mm are noted. Biopsy of the right hilar lymph node shows features of adenocarcinoma. He is an ex-smoker with a 10-pack year history. He usually lives alone and is a retired accountant. What would be the most appropriate treatment option?

- a) Chemotherapy alone
- b) Chemo-radiotherapy combination
- c) Surgery alone
- d) **Surgery with adjuvant chemotherapy –**
 - T1-4 N0-2 (if single zone non-bulky N2) M0 = Surgery
 - N \geq 1 = Adjuvant chemo
- e) Neoadjuvant chemotherapy followed by surgery

T1-T3 = all operable

T4 is odd, T4 isn't inoperable but depends on tumour

N0-1 = always operable

N2 = if single zone and non-bulky N2 = surgery

M0 = for surgery

If N \geq 1 = adjuvant chemo

PET

- Only if for radical treatment
- Oligometastatic disease mets <3 – trial

- Next Qs are from William ricketts

Questions?



- William.Ricketts@nhs.net



Question 1a

A 29-year-old man is referred to the Chest Clinic with recurrent chest infections. He has previously isolated *Haemophilus influenzae* in his sputum. He has no recent medical history of note. As a child, however, he needed multiple courses of antibiotics for ear infections. He is on no regular medications and does not report any gastrointestinal symptoms. He and his partner are trying for a baby but have been unsuccessful for the past 18 months.

Which test would be the most useful initial investigation to look for the cause of his symptoms?

- a) Sweat test
- b) Nasal nitric oxide measurement
- c) Genetic testing
- d) Nasal biopsy
- e) Serum immunoglobulins

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Question 1a

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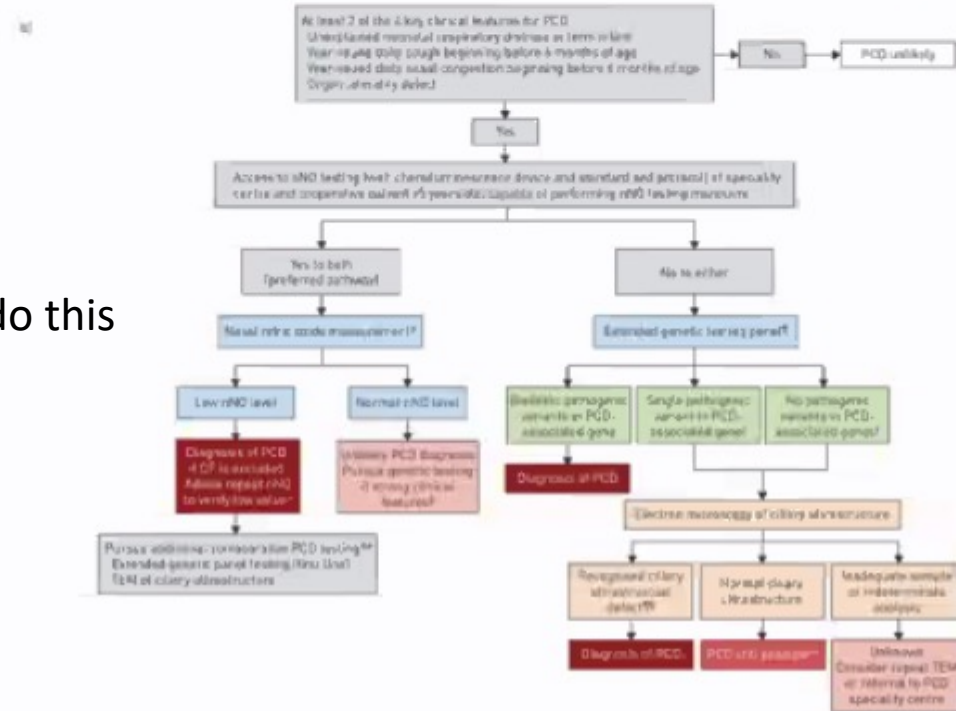
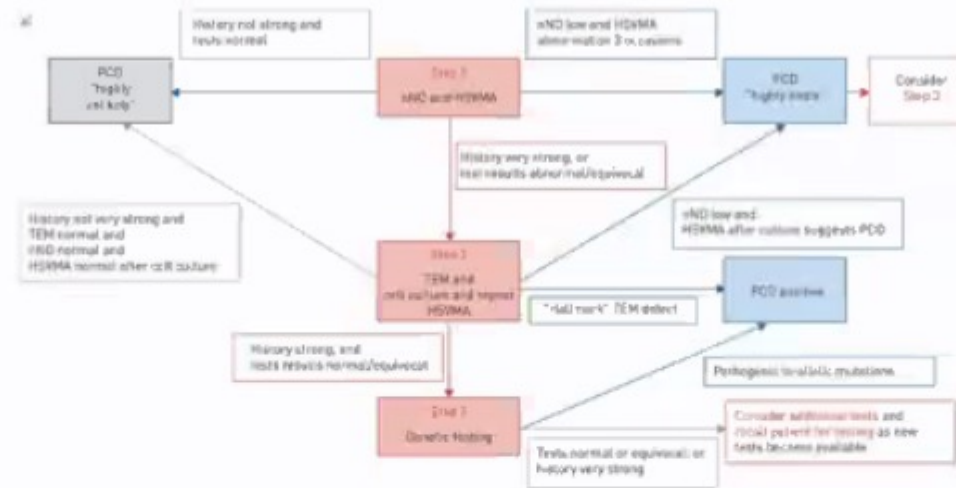
- a) Sweat test
- b) Nasal nitric oxide measurement**
- c) Genetic testing
- d) Nasal biopsy
- e) Serum immunoglobulin

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a) Adapted from the European Respiratory Society primary ciliary dyskinesia (PCD) diagnostic algorithm [6].



Must exclude CVID or CF to do this

Question 1b

A 29-year-old man is referred to the Chest Clinic with recurrent chest infections. He has previously isolated *Haemophilus influenzae* in his sputum. He has no recent medical history of note. As a child, however, he needed multiple courses of antibiotics for ear infections. He is on no regular medications and does not report any gastrointestinal symptoms. He and his partner are trying for a baby but have been unsuccessful for the past 18 months.

What is the mode of inheritance for the most likely diagnosis?

- a) Autosomal recessive
- b) Autosomal dominant
- c) X-linked recessive
- d) X-linked dominant
- e) Mitochondrial

Question 1b

A 29-year-old man is referred to the Chest Clinic with recurrent chest infections. He has previously isolated *Haemophilus influenzae* in his sputum. He has no recent medical history of note. As a child, however, he needed multiple courses of antibiotics for ear infections. He is on no regular medications and does not report any gastrointestinal symptoms. He and his partner are trying for a baby but have been unsuccessful for the past 18 months.

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Question 2

Which of the following parameters is included in the Bronchiectasis Severity Index (BSI) but not in the FACED score for severity assessment of bronchiectasis?

- a) Spirometry
- b) Age
- c) *Pseudomonas aeruginosa* colonization
- d) Exacerbation frequency
- e) Number of lobes involved

Question 2

Which of the following parameters is included in the Bronchiectasis Severity Index (BSI) but **not** in the FACED score for severity assessment of bronchiectasis?

- a) Spirometry
- b) Age
- c) *Pseudomonas aeruginosa* colonization
- d) Exacerbation frequency**
- e) Number of lobes involved

Question 2

| Bronchiectasis Severity Score (BSI) | FACED |
|--|---|
| Age | <u>F</u> EV ₁ %-predicted |
| Body mass index | <u>A</u> ge |
| FEV ₁ %-predicted | <i>P. aeruginosa</i> <u>C</u> olonisation |
| Hospital admissions in last 2 years | Radiological <u>E</u> xtent |
| Exacerbations in last 12 months | MRC <u>D</u> yspnoea score |
| MRC dyspnoea score | |
| <i>P. aeruginosa</i> colonisation | |
| Colonisation with other organisms | |
| Radiological severity | |

Question 3

A 27 year old woman with cystic fibrosis continues to grow *Pseudomonas aeruginosa* from sputum samples despite attempted eradication therapy.

Which of the following treatments is not currently licensed in the UK for treatment of chronic *Pseudomonas aeruginosa* infection in adults with cystic fibrosis?

- a) Nebulised aztreonam
- b) Nebulised tobramycin
- c) Dry powder inhaled colistimethate sodium
- d) Nebulised liposomal amikacin
- e) Nebulised levofloxacin

Question 3

A 27 year old woman with cystic fibrosis continues to grow *Pseudomonas aeruginosa* from sputum samples despite attempted eradication therapy.

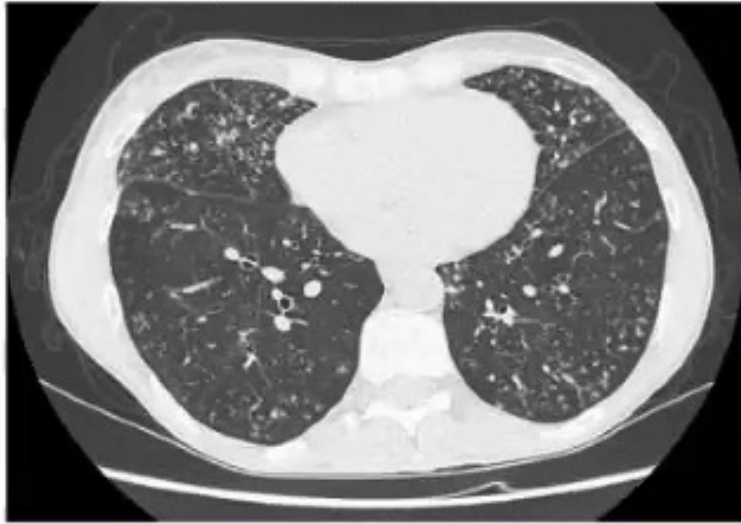
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- c) Dry powder inhaled colistimethate sodium
- d) Nebulised liposomal amikacin**
- e) Nebulised levofloxacin

Question 4

A 45-year-old woman is referred to the chest clinic with a chronic cough. She was previously fit and well and has no significant past medical history of note. She does not report any fevers or weight loss.

A CT chest is performed:



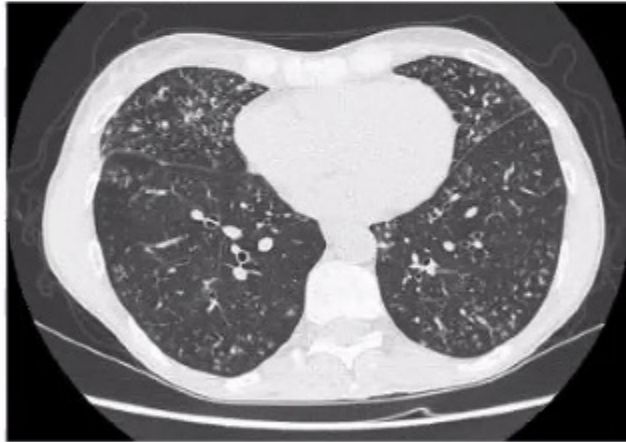
In which of the following scenarios would initiation of NTM treatment be warranted for this patient?

- a) 1 positive sputum sample for *M. avium* complex
- b) 1 positive sputum sample for *M. chimaera* and a negative CT-directed BAL
- c) 1 positive CT-directed BAL for *M. avium* complex
- d) Negative sputum and BAL samples, but typical radiological features
- e) 2 positive sputum samples: one for *M. avium* complex and another for *M. kansasii*

Question 4

A 45-year-old woman is referred to the chest clinic with a chronic cough. She was previously fit and well and has no significant past medical history of note. She does not report any fevers or weight loss.

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- b) 1 positive sputum sample for *M. chimaera* and a negative CT-directed BAL
- c) 1 positive CT-directed BAL for *M. avium* complex**
- d) Negative sputum and BAL samples, but typical radiological features
- e) 2 positive sputum samples: one for *M. avium* complex and another for *M. kansasii*

Appropriate symptoms and radiology findings and one CT directed BAL is enough to start treatment

Question 5

Appropriate regimens for the initial phase of treatment for *Mycobacterium abscessus* pulmonary disease could include all of the following drugs except:

- a) IV imipenem
- b) IV tigecycline
- c) IV amikacin
- d) Oral clarithromycin
- e) Oral minocycline

Question 5

Appropriate regimens for the initial phase of treatment for *Mycobacterium abscessus* pulmonary disease could include all of the following drugs except:

- a) IV imipenem
- b) IV tigecycline
- c) IV amikacin
- d) Oral clarithromycin
- e) Oral minocycline**

Treatment of *M. abscessus* Pulmonary Disease: BTS Guideline 2017

Table 8 Suggested antibiotic regimens for adults with *Mycobacterium abscessus*-pulmonary disease

| <i>M. abscessus</i> | Antibiotic regimen |
|---|---|
| Clarithromycin sensitive isolates or inducible macrolide-resistant isolates | <p>Initial phase: ≥ 1 month† intravenous amikacin 15 mg/kg daily or 3× per week‡ and intravenous tigecycline 50mg twice daily and where tolerated intravenous imipenem 1 g twice daily and where tolerated oral clarithromycin 500 mg twice daily or oral azithromycin 250–500 mg daily</p> <p>Continuation phase: nebulised amikacin‡ and oral clarithromycin 500 mg twice daily or azithromycin 250–500 mg daily and 1–3 of the following antibiotics guided by drug susceptibility results and patient tolerance: oral clofazimine 50–100mg daily§ oral linezolid 600mg daily or twice daily oral minocycline 100mg twice daily oral moxifloxacin 400mg daily oral co-trimoxazole 960 mg twice daily</p> |
| Constitutive macrolide-resistant isolates | <p>Initial phase: ≥ 1 month† intravenous amikacin 15 mg/kg daily or 3× per week‡ and intravenous tigecycline 50mg twice daily and where tolerated intravenous imipenem 1 g twice daily</p> <p>Continuation phase: nebulised amikacin‡ and 2–4 of the following antibiotics guided by drug susceptibility results and patient tolerance: oral clofazimine 50–100mg daily§ oral linezolid 600mg daily or twice daily oral minocycline 100mg twice daily oral moxifloxacin 400mg daily oral co-trimoxazole 960 mg twice daily</p> |

Question 6

A 25-year-old woman is admitted to hospital with fevers, chest pain and a purulent cough. She is alert and orientated with no neurological symptoms. Clinical parameters reveal a heart rate of 100/min, BP 100/60 mmHg, temperature 38°C, oxygen saturations 93% on air and respiratory rate 25/min. On systems enquiry, she developed right sided upper abdominal pain a few days ago. She has recently returned to the UK from a backpacking holiday around SE Asia during the rainy season.

A chest X-ray shows right upper lobe consolidation. Blood tests reveal:

| Full blood Count | Biochemistry |
|---|--|
| Haemoglobin 130 g/L White cell count $15 \times 10^9/L$ Neutrophils $10.6 \times 10^9/L$ Eosinophils $0.24 \times 10^9/L$ Platelets $550 \times 10^9/L$ | Sodium 140 mmol/L Potassium 3.5 mmol/L Urea 8.0 mmol/L Creatinine 80 $\mu\text{mol/L}$ ALT 70 IU/L ALP 230 IU/L Albumin 30 g/L CRP 130 mg/L |

What is the optimal antibiotic therapy?

- a) IV ceftazidime
- b) IV amoxicillin
- c) IV co-amoxiclav + clarithromycin
- d) Oral doxycycline
- e) Oral levofloxacin



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What is the optimal antibiotic therapy?

- a) **IV ceftazidime**
- b) IV amoxicillin
- c) IV co-amoxiclav + clarithromycin
- d) Oral doxycycline
- e) Oral levofloxacin

Burkholderia (not the CF one) – can cause septicaemia or subacute infection, can be incubated for up to 3 weeks. Single agent broad spec abx , extra pulmonary involvement

Question 7

A 60-year-old man presents with a new, mild dry cough. He is an ex-smoker with a 20-pack year history. He denies any systemic symptoms. He has recently been undertaking work on his chimney and spent a significant proportion of his time on the roof. He has no relevant past medical history of note and is on no regular medications. Peripheral saturations are 98% RA with a HR of 60, BP of 130/80 and temp of 36.0°C.

A CT chest shows a 1.3cm solid pulmonary nodules with a pleural base and two further <5mm pulmonary nodules. A CT-guided biopsy of the largest nodule does not show any evidence of malignancy. Blood tests reveal a positive cryptococcal antigen test.

What is the optimal treatment for this patient?

- a) No treatment required
- b) Oral fluconazole for 6 months
- c) IV flucytosine and amphotericin for 1 month
- d) Oral itraconazole for 3 months
- e) Oral Posaconazole for 3 months



Question 7

A 60-year-old man presents with a new, mild dry cough. He is an ex-smoker with a 20-pack year history. He denies any systemic symptoms. He has recently been undertaking work on his chimney and spent a significant proportion of his time on the roof. He has no relevant past medical history of note and is on no regular medications. Peripheral saturations are 98% RA with a HR of 60, BP of 130/80 and temp of 36.0°C.

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- c) IV flucytosine and amphotericin for 1 month
- d) Oral itraconazole for 3 months
- e) Oral Posaconazole for 3 months

Cryptococcus in an immune competent, the cryptococcal antigen being positive pushes you towards giving treatment

Q1 Exam

Which of these fulfil the criteria for PAH by right heart catheterisation

- A. RAP 5mmHg, mPAP 44mmHg, PCWP 26mmHg, CO 4l/min
- B. RAP 5mmHg, mPAP 16mmHg, PCWP 8mmHg, CO 4l/min
- C. RAP 5mmHg, mPAP 44mmHg, PCWP 8mmHg, CO 4l/min
- D. RAP 5mmHg, mPAP 44mmHg, PCWP 28mmHg, CO 2l/min
- E. RAP 18mmHg, mPAP 16mmHg, PCWP 14mmHg, CO 7l/min

Q1 Exam

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- E. RAP 18mmHg, mPAP 16mmHg, PCWP 14mmHg, CO 7l/min

Ans = C

Pulmonary hypertension is a haemodynamic diagnosis

- Guidelines state that RHC mandatory for diagnosis and treatment decisions^{1,2}
- PH is defined by current guidelines (ESC 2015) at right heart catheterisation as a mean pulmonary artery pressure $\geq 25\text{mmHg}$
- New definition for PH proposed at the 6th WSPH³
 - The **new upper limit for normal mPAP is 20 mmHg**
 - However, elevated mPAP alone cannot be used to define pulmonary vascular disease: need to also use PVR (mPAP/PCWP/CO)

| Definitions | Characteristics | Clinical groups |
|---|----------------------------|-----------------|
| Pre-capillary PH | mPAP $>20\text{mmHg}$ | 1,3,4 and 5 |
| | PAWP $\leq 15\text{ mmHg}$ | |
| | PVR $\geq 3\text{ WU}$ | |
| Isolated post-capillary PH (IpcPH) | mPAP $>20\text{mmHg}$ | 2 and 5 |
| | PAWP $>15\text{ mmHg}$ | |
| | PVR $<3\text{ WU}$ | |
| Combined pre- and post-capillary PH (CpcPH) | mPAP $>20\text{mmHg}$ | 2 and 5 |
| | PAWP $>15\text{ mmHg}$ | |
| | PVR $\geq 3\text{ WU}$ | |

New definition for PH proposed at the 6th WSPH³

NB includes PVR

Q2 Exam

Which investigation is recommended by the ESC/ERS Guidelines as the first line test when pulmonary hypertension is suspected

- A. Chest X-ray
- B. CT pulmonary angiogram
- C. Echocardiogram
- D. Electrocardiogram
- E. Ventilation perfusion lung scan

Q2 Exam

Which investigation is recommended by the ESC/ERS Guidelines as the first line test when pulmonary hypertension is suspected

- A. Chest X-ray
- B. CT pulmonary angiogram
- C. Echocardiogram
- D. Electrocardiogram
- E. Ventilation perfusion lung scan

C

Imaging is recommended in suspected PH and allows for evaluation of the RV



Chest radiograph is usually abnormal in patients with PH and can be used to assist differential diagnosis¹



Echocardiography is recommended as a first-line, non-invasive diagnostic test in patients with suspected PH¹



Ventilation/ perfusion lung scan is recommended to exclude CTEPH in patients with unexplained PH¹



CTPA +/- perfusion (DECT/LSIM) can provide key information on vascular, cardiac, parenchymal and mediastinal abnormalities^{1,2}
Cardiac magnetic resonance imaging (CMRI) can accurately assess RV size, morphology and function^{1,2}

Right heart catheterization is required to confirm diagnosis of PAH

CTEPH, chronic thromboembolic pulmonary hypertension; CTPA, computed tomography pulmonary angiography; DECT, dual energy computed tomography; LSIM, lung subtraction iodine mapping; PH, pulmonary hypertension; RV, right ventricle/right ventricular.
1. Galiè N, et al. *Eur Heart J* 2016; 37(1):67-119; 2. Kiely DG, et al. *Pulm Circ* 2019; 9(3):2045894019841990.
CT/MRI icon by Freepik available at <https://www.flaticon.com>.

Q3 Exam

Which following medical condition is not associated with pulmonary arterial hypertension?

- A. HIV infection
- B. Portal hypertension
- C. Mitral stenosis
- D. Systemic Lupus Erythematosus (SLE)
- E. Systemic Sclerosis

Q3 Exam

Which following medical condition is not associated with pulmonary arterial hypertension?

- A. HIV infection
- B. Portal hypertension
- C. Mitral stenosis
- D. Systemic Lupus Erythematosus (SLE)
- E. Systemic Sclerosis

C Mitral stenosis is a group 2, the rest are a group 1 causes

Clinical Classification of Pulmonary Hypertension

1. Pulmonary arterial hypertension (PAH)

- 1.1 Idiopathic
- 1.2 Heritable (BMP2, others)
- 1.3 Drugs and toxin-induced PAH
- 1.4 Associated PAH (e.g. CTD, HIV (1.4.2), CHD (1.4.4), schistosomiasis)
- 1.5 PAH long-term responders to calcium channel blockers
- 1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement
- 1.7 Persistent PH of the newborn syndrome

2. Pulmonary hypertension associated with left heart diseases

- LV systolic or LV diastolic dysfunction, valvular disease, cardiomyopathies

3. Pulmonary hypertension associated with lung diseases and/or hypoxia

- COPD, ILD, sleep disordered breathing, alveolar hypoventilation, high altitude

4. Pulmonary hypertension due to chronic thromboembolic disease

- CTEPH...

5. PH with unclear or multi-factorial mechanisms

- SARCOIDOSIS, haematological disorders, renal failure, LAM, segmental PH ...

CTDs and pulmonary hypertension

| | Prevalence | 3 year survival |
|--------------------|------------|-----------------|
| Systemic sclerosis | ≈ 10 % | ≈ 50% |
| SLE | < 1 % | ≈ 75% |
| MCTD / UDCT | ≈ 2-5 % | ? |
| RA | uncommon* | ? |
| Dermatomyositis | uncommon* | ? |
| Polymyositis | uncommon* | ? |

* uncommon in the absence of severe respiratory disease

Q4 Exam

A 60-year-old gentleman presents with progressive breathlessness. ECG shows normal sinus rhythm. He has no history of or risk factors for coronary disease. PMHx includes a splenectomy following a road accident in his 20s. Blood tests reveal a negative troponin, negative D dimer, raised BNP of 500ng/L and normal inflammatory markers. CXR is normal. He is referred to the chest clinic. Lung function tests show a FEV1 85% predicted, FVC 88% predicted, TLC 90% predicted, TLCO 50% predicted, KCO 55% predicted.

HRCT is normal. Echocardiogram shows normal LV function with no suggestion of regional wall motion abnormalities, and a TR velocity of 3.2m/s, with normal RV function.

What is your next best investigation?

- A. Right heart catheterization
- B. V/Q scan
- C. Sleep study
- D. Bronchodilator reversibility
- E. Cardiac MRI

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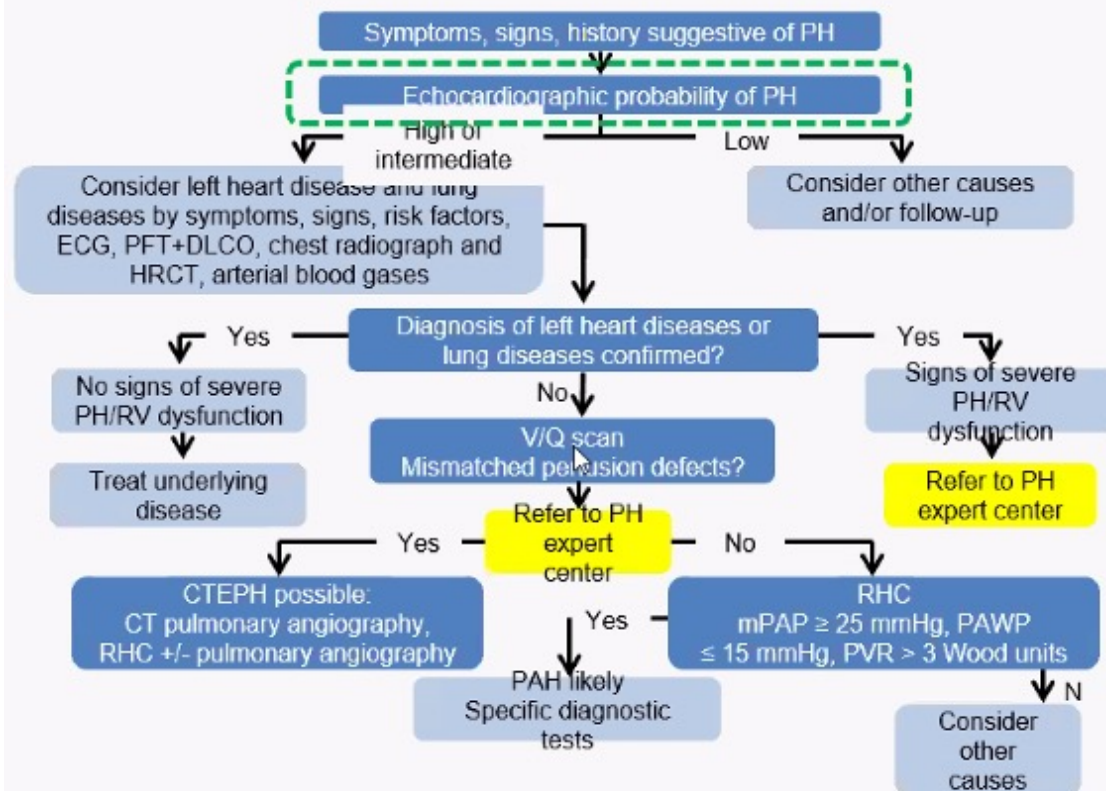
- A. Right heart catheterization
- B. V/Q scan
- C. Sleep study
- D. Bronchodilator reversibility
- E. Cardiac MRI

Ans - B

V/Q before right heart catheter

2015 ESC/ERS Guidelines

Diagnostic algorithm and recommendations



| Recommendations | Class | Level |
|---|-------|-------|
| Echocardiography is recommended as a first-line non-invasive diagnostic investigation in case of suspicion of PH | I | C |
| V/Q or perfusion lung scan is recommended in patients with unexplained PH to exclude CTEPH | I | C |
| Contrast CT angiography of the pulmonary artery is recommended in the workup of patients with CTEPH | I | C |
| Routine biochemistry, hematology, immunology, HIV testing and thyroid function tests are recommended in all patients with PAH to identify the specific associated condition | I | C |
| Abdominal ultrasound is recommended for the screening of portal hypertension | I | C |
| Lung function test with DLCO is recommended in the initial evaluation of patients with PH | I | C |
| High-resolution CT should be considered in all patients with PH | IIa | C |
| Pulmonary angiography should be considered in the workup of patients with CTEPH | IIa | C |
| Open or thoracoscopic lung biopsy is not recommended in patients with PAH | III | C |

CT, computed tomography; CTEPH, chronic thromboembolic pulmonary hypertension; DLCO, diffusing capacity for carbon monoxide; ECG, electrocardiogram; ERS, European Respiratory Society; ESC, European Society of Cardiology; HRCT, high-resolution computed tomography; mmHg, millimeters of mercury; mPAP, mean pulmonary arterial pressure; PAH, pulmonary arterial hypertension; PAWP, pulmonary artery wedge pressure; PFT, pulmonary function test; PH, pulmonary hypertension; RHC, right heart catheterization; RV, right ventricle V/Q, ventilation/perfusion. 1. Galie N, et al. *Eur Heart J* 2016; 37(1):67-119.

Q5 Exam

.
Which of the following statements are true about general measures to consider in the management of patients with PAH:

- A. All patients with PAH should be fully anticoagulated
- B. Patients should be counselled about the mortality risks of pregnancy of up to 20-30%, and the importance of using double contraception from the time of PAH diagnosis
- C. Diuretics, often a combination of a loop and aldosterone antagonist, are rarely useful in later stages of PAH
- D. It is usually fine to proceed with an elective laparoscopic cholecystectomy in a DGH in a patient with moderate risk PAH without discussion with a PH centre
- E. Regular exercise is unlikely to improve exercise performance in PAH

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Ans - B

General Measures in PAH treatment: Anticoagulation, oxygen, exercise, pregnancy

- Non-randomised trials have shown improved survival in PAH patients receiving oral anticoagulants vs. controls
 - 49% vs. 21% (Fuster *et al*, 1984)¹
 - 47% vs. 31% (Rich *et al*, 1992)²
 - COMPERA –recent German registry – **benefit only in IPAH**
 - **Updated World Symposium guidelines – consider on a case by case only**
- Diuretics
 - Clinical benefit RV failure is established¹
 - No randomised trials to establish type / dose
- Keep $pO_2 > 8kPa$ and $SaO_2 > 90\%$ (except Eisenmengers)
- Exercise training improves FC and quality of life
- Avoid surgery / procedures if possible
- Avoid pregnancy: mortality at least 30%

Fuster *et al*, Circulation 1984
Rich *et al*, The New England Journal of Medicine; 1992
Hoeper *et al*, COMPERA Registry
Mereles *et al*, Circulation 2006
Bedard *et al*, EHJ 2012

Q6 Exam

A 28 year-old woman is diagnosed with idiopathic pulmonary arterial hypertension. She is in WHO Functional Class III, has a 6MWT distance of 320 m and a BNP of 230ng/l.

| Right heart catheter | Baseline | Inhaled nitric oxide |
|--|-----------|----------------------|
| mean right atrial pressure | 7 mmHg | 8 mmHg |
| mean pulmonary arterial pressure | 52 mmHg | 28 mmHg |
| mean pulmonary arterial wedge pressure | 8 mmHg | 6 mmHg |
| mean cardiac output | 4.2 L/min | 5.8 L/min |

How would you treat her idiopathic pulmonary arterial hypertension?

- A. Nifedipine, increasing as tolerated
- B. A guanylate cyclase stimulator
- C. A nitric oxide donor
- D. A phosphodiesterase inhibitor
- E. Intravenous prostanoid

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A

Vasoresponder PAH

5-10% of patients with idiopathic, heritable, drug-induced PAH

Respond to inhaled pulmonary vasodilator at RHC (inhaled nitric oxide (NO) best)

Positive response ('Sitbon criteria'):

Drop in mPAP ≥ 10 mmHg to reach mPAP ≤ 40 mmHg
without a fall in cardiac output (1)

Very important to do this test at first RHC

Patients who respond acutely have better survival (2)

High dose CCBs max daily doses:

Nifedipine 120-240mg

Diltiazem 240-720mg daily

Amlodipine up to 20mg

Reassess with RHC at 3-4 months

May need to add PAH therapies later

Q7 Exam

A 21-year-old woman presents with rapidly progressive breathlessness 6 months following a term pregnancy. She describes syncope on minimal exertion, and can only walk a few steps. ECG shows sinus tachycardia with signs of RV hypertrophy. Pulse is 110, BP 90/60mmHg. Blood tests reveal Hb 14g/L, normal liver and renal function, a raised BNP of 500ng/L, lactate 2.8, and normal inflammatory markers. CXR shows normal parenchyma, confirmed on HRCT, with no thromboembolic disease seen on CTPA. Echocardiogram shows a TR velocity of 3.9m/s, reduced RV function with TAPSE 10mm, and a small pericardial effusion. PAH is confirmed on initial RHC in the referring hospital.

How would you risk assess her and/or best plan initial management?

- A. Low risk PAH as she has no oedema to suggest RV failure
- B. Low risk as she survived pregnancy, but she should be seen soon in a PH outpatient clinic
- C. Moderate risk PAH, start PAH therapy as outpatient
- D. High risk features, she should be urgently transferred to a PH centre for RHC with pulmonary vasodilator testing, PAH therapy initiation and consideration of intravenous prostanoids. She is likely to need at least an HDU bed until haemodynamically stable as she has a high risk of cardiac decompensation
- E. High risk PAH, she should be referred now for urgent lung transplantation listing

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2015 ESC / ERS Guidelines recommend regular multiparameter risk assessments

Clinical Evaluation

Exercise Capacity

RV Function

| Determinants of prognosis | Estimated 1-year mortality | | |
|---------------------------------------|---|---|--|
| | Low risk < 5% | Intermediate risk 5-10% | High risk > 10% |
| Clinical signs of right heart failure | Absent | Absent | Present |
| Progression of symptoms | No | Slow | Rapid |
| Syncope | No | Occasional syncope | Repeated syncope |
| FC | I, II | III | IV |
| 6MWD | > 440 m | 165-440 m | < 165 m |
| CPET | Peak $\text{VO}_2 > 15 \text{ ml/min/kg}$ ($> 65\% \text{ pred.}$) $\text{VE/VCO}_2 \text{ slope} < 36$ | Peak $\text{VO}_2 11-15 \text{ ml/min/kg}$ ($35-65\% \text{ pred.}$) $\text{VE/VCO}_2 \text{ slope } 36 - 44.9$ | Peak $\text{VO}_2 < 11 \text{ ml/min/kg}$ ($< 35\% \text{ pred.}$) $\text{VE/VCO}_2 \text{ slope} \geq 45$ |
| NT-proBNP plasma levels | BNP < 50 ng/l NT-proBNP < 300 ng/l | BNP 50-300 ng/l NT-proBNP 300-1400 ng/l | BNP > 300 ng/l NT-proBNP > 1400 ng/l |
| Imaging (echo, CMR) | RA area < 18 cm^2 No pericardial effusion | RA area $18-26 \text{ cm}^2$ No or minimal pericardial effusion | RA area > 26 cm^2 Pericardial effusion |
| Hemodynamics | RAP < 8 mmHg CI $\geq 2.5 \text{ l/min/m}^2$ SvO ₂ > 65% | RAP 8-14 mmHg CI $2.0-2.4 \text{ l/min/m}^2$ SvO ₂ 60-65% | RAP > 14 mmHg CI < 2.0 l/min/m^2 SvO ₂ < 60% |

Galiè N, et al. *Eur Heart J* 2016; 37:67-119; Galiè N, et al. *Eur Respir J* 2019; 53:1801889

Q8 Exam

.

Which of the following statements are true about CTEPH:

- A. Almost all patients with CTEPH can pinpoint an initial acute PE episode
- B. Patients should, in most cases, be referred for pulmonary endarterectomy but there is no need for urgency to pursue this
- C. Risk factors can include splenectomy, infected pacemaker wires, non-O blood groups and VA shunts
- D. It is not usually necessary to screen for antiphospholipid syndrome
- E. Oral PH therapies always improve exercise performance in CTEPH

Q8 Exam

.

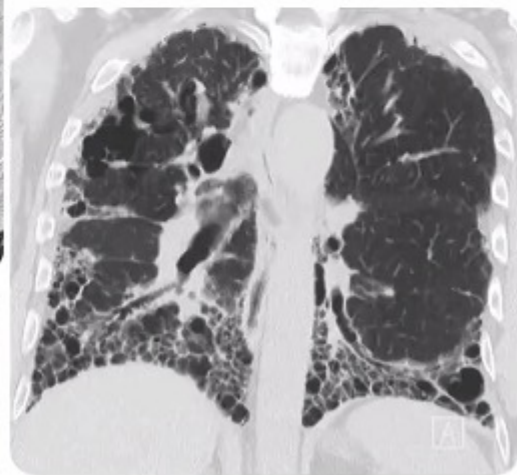
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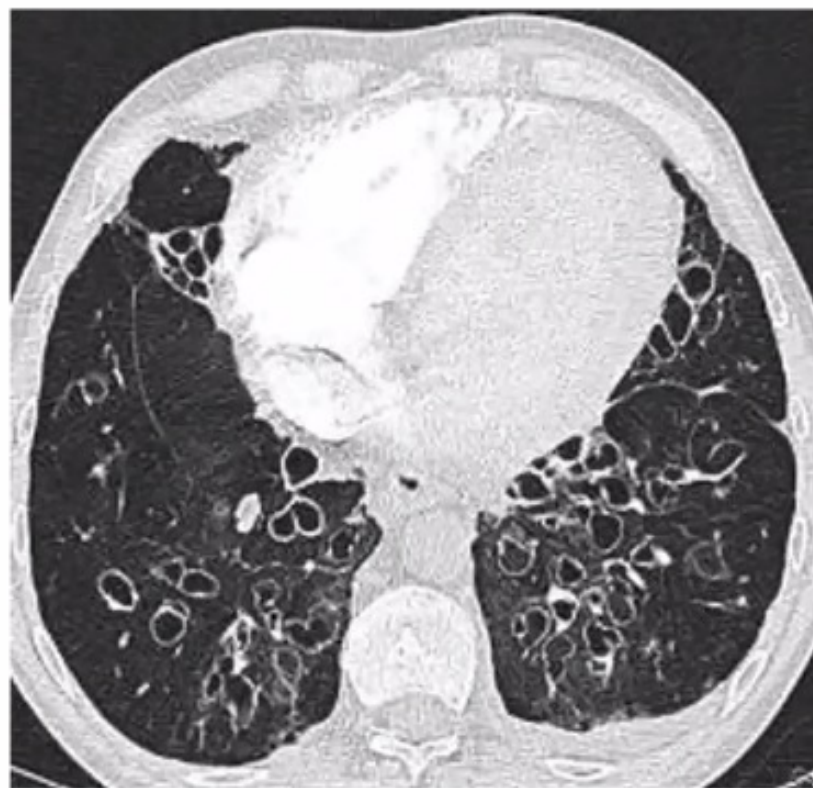
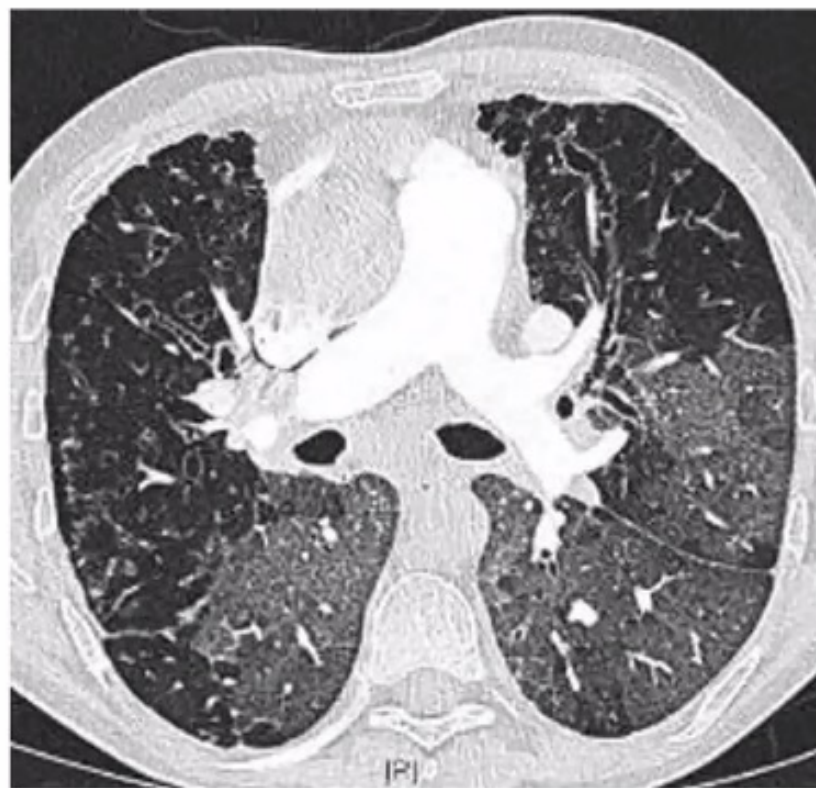
C

Q1 | *What is the most likely radiological diagnosis?*

- **IPF**
- Emphysema
- Bronchiectasis
- Fibrotic HP
- Fibrotic NSIP



Q2 | 40 yo female; Recurrent chest infections. *What is the most likely radiological diagnosis?*



- A) Mosaic attenuation
- B) DIP
- C) Sub-acute HP
- D) Air-trapping
- E) Bronchiectasis

Q2 | 40 yo female; Recurrent chest infections. *What is the most likely radiological diagnosis?*

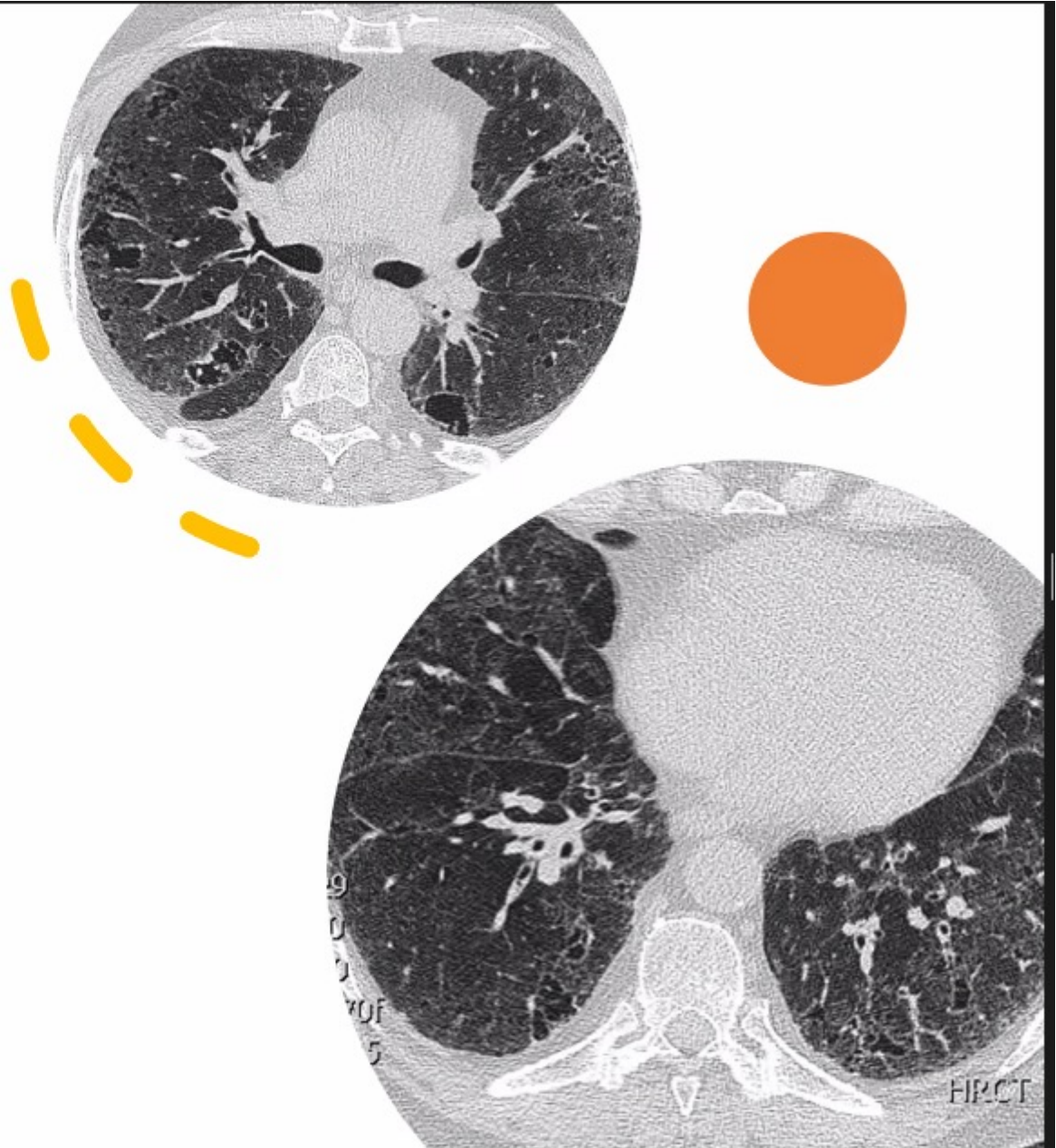


- A) Mosaic attenuation
- B) DIP
- C) Sub-acute HP
- D) Air-trapping
- E) Bronchiectasis

E - You do get mosaic attenuation often in bronchiectasis

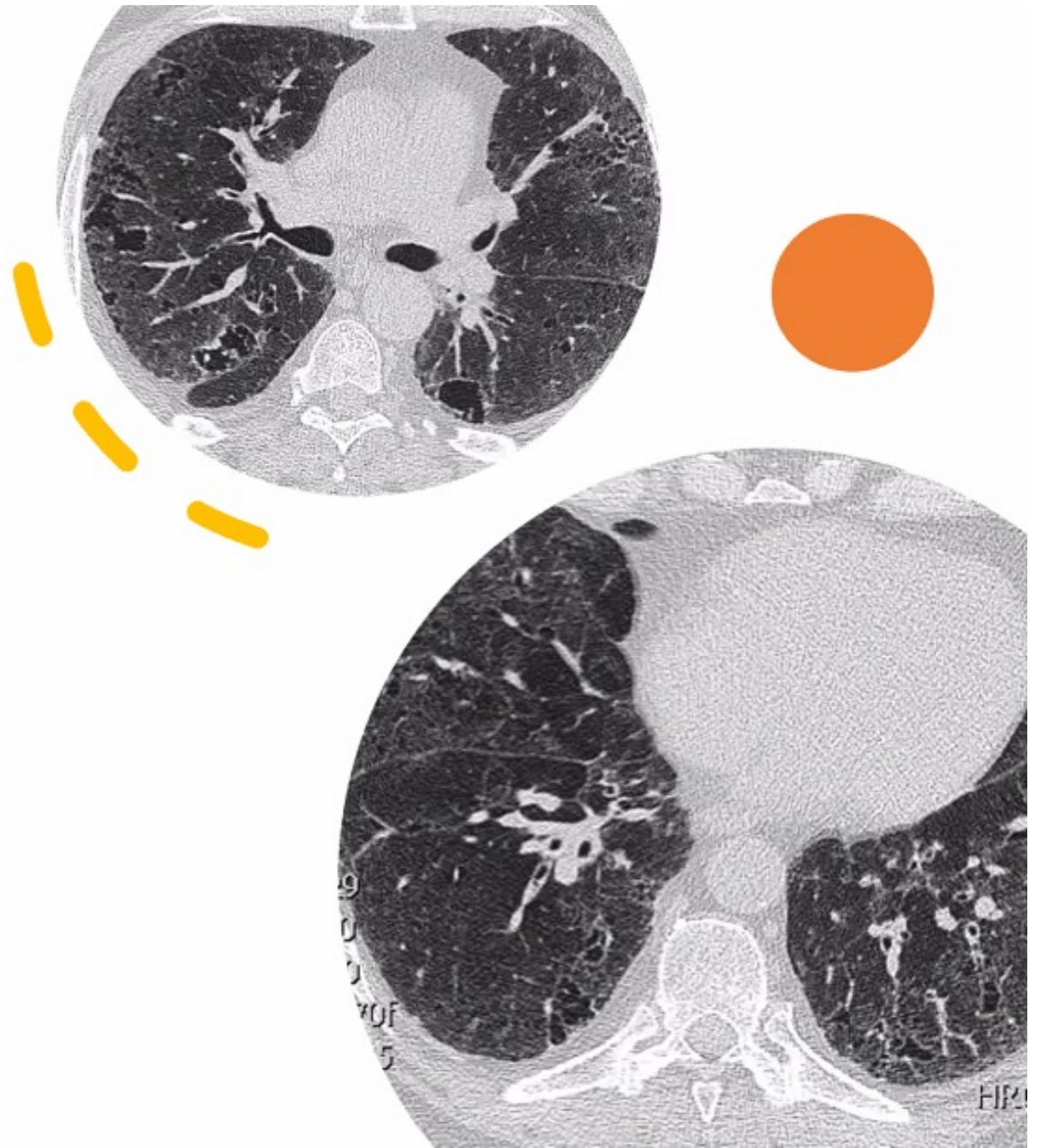
Q3 | 50 yo male; Finger clubbing; Smoker (12-15 roll-ups/day; 35 p-y history). *What is the most likely radiological diagnosis?*

- Emphysema
- DIP
- RBILD
- Smoking-related ILD
- Langerhans' cell histiocytosis



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- RBILD
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Q4 | 45 yo female; progressive SOB (now ~100 yds on flat), dry cough; non-smoker. *What is the most likely radiological diagnosis?*

- Atypical IPF
- DIP
- Fibrotic HP
- Fibrotic NSIP
- Sarcoidosis



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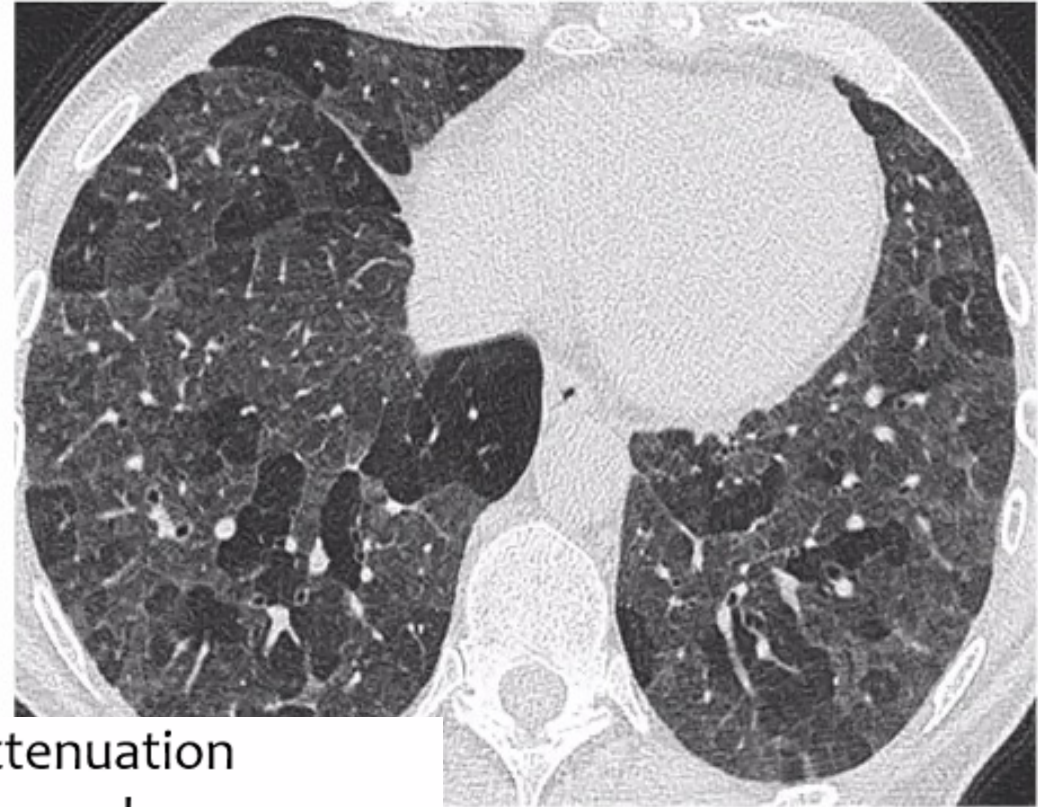
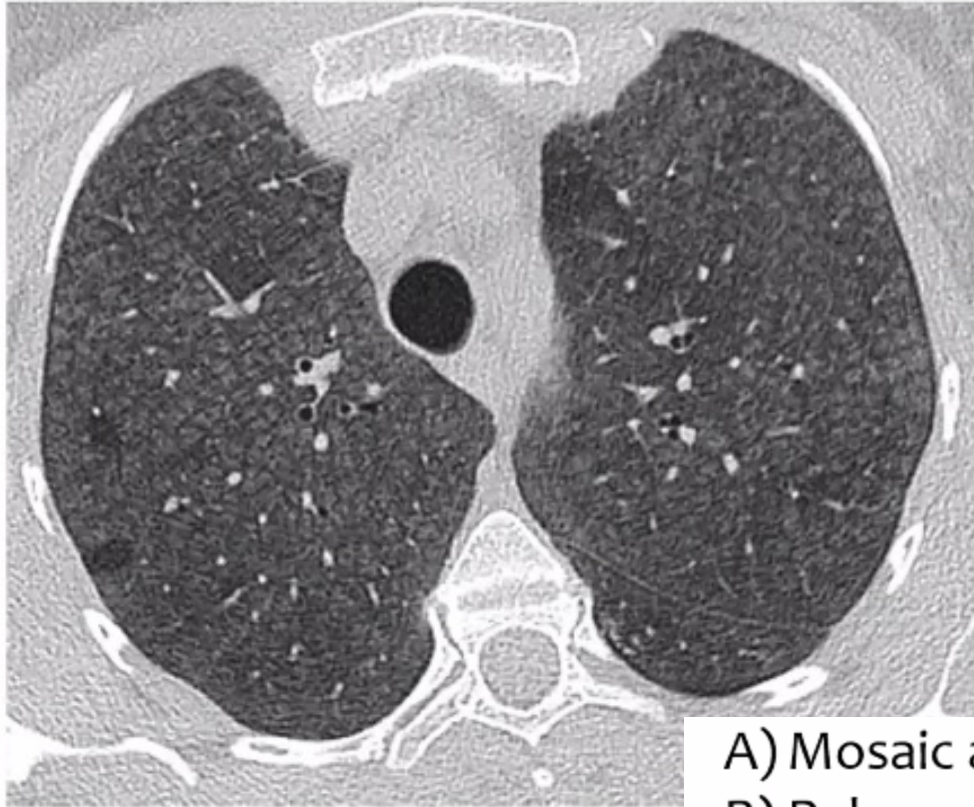
- Atypical IPF
- DIP
- Fibrotic HP
- **Fibrotic NSIP**
- Sarcoidosis



Very rare – fibrotic NSIP = CT disease – u must find it!!

GGO with areas of abnormality in the airway with traction bronchiectasis, tortuous outline. Surrounding fibrosis.

Q5 | 60 yo male; Dry cough and dyspnoea; recently moved house. **What is the most likely radiological diagnosis?**



- A) Mosaic attenuation
- B) Pulmonary oedema
- C) (non-fibrotic) HP
- D) Air-trapping
- E) Small airways disease

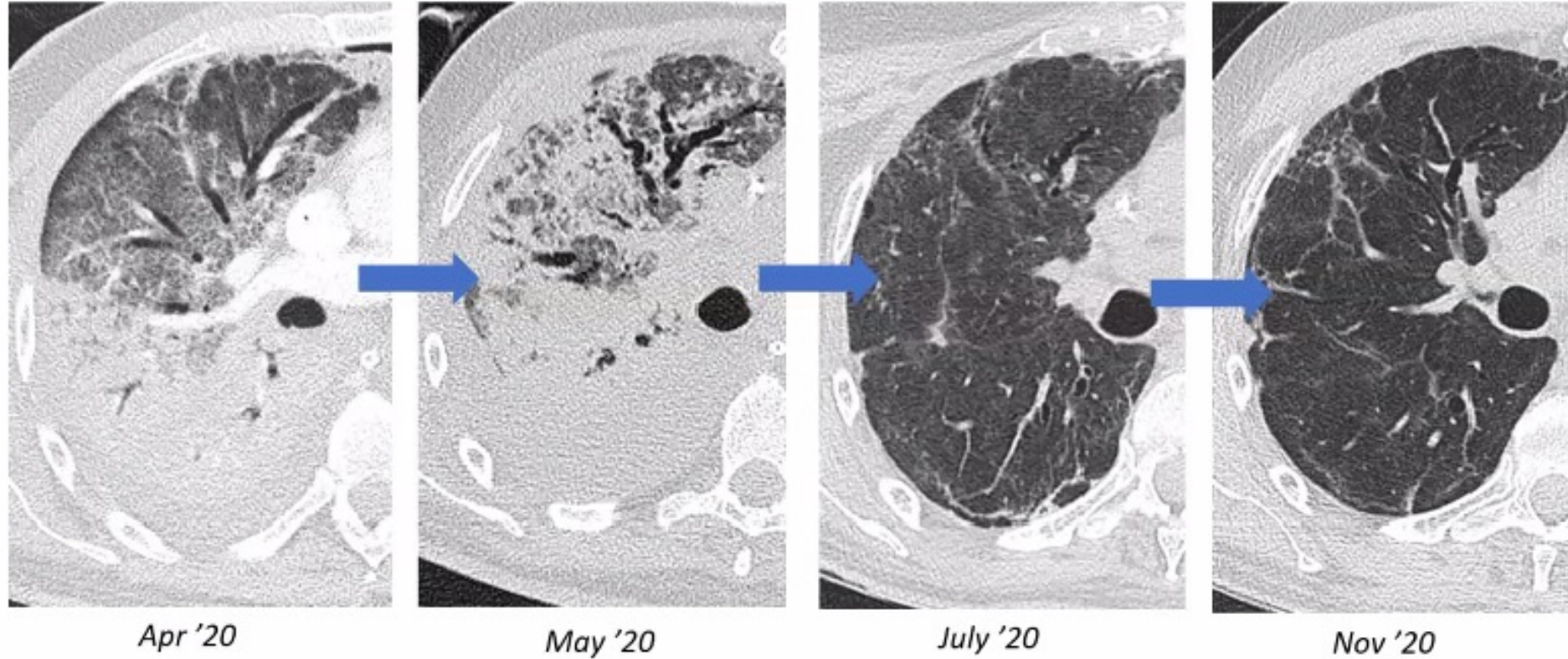
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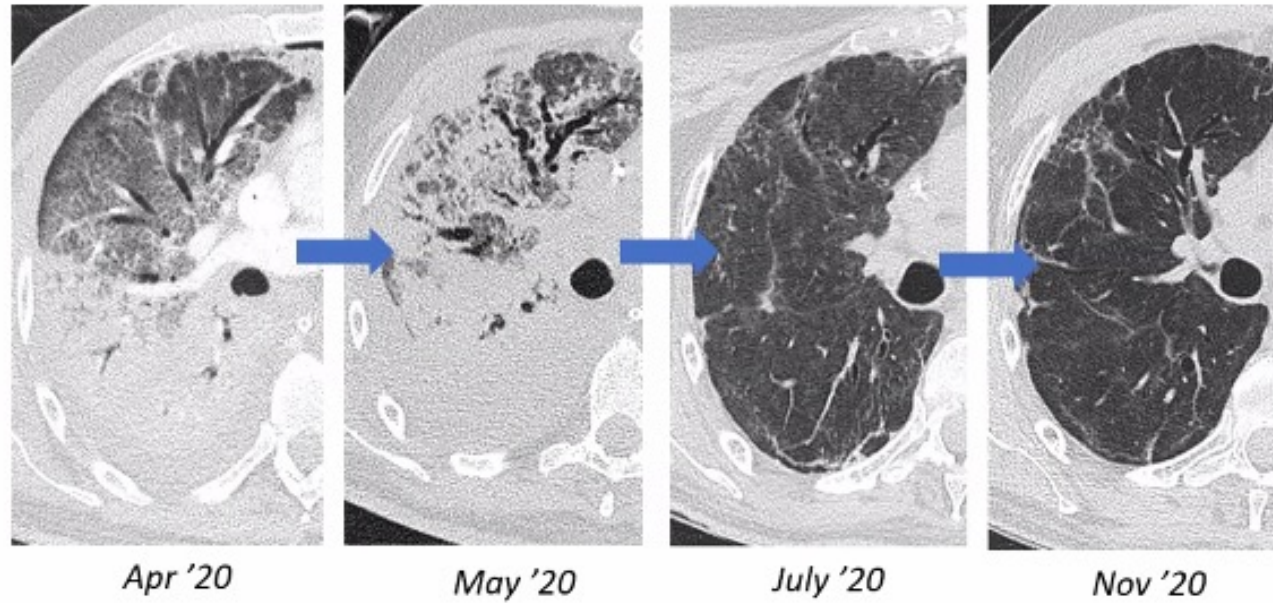
Black and white lung areas in this area are both abnormal, how do I know that?
Look at the vessels, are vessels in black lung small or less than the vessel density in a similar area of lung in the white area.
Centrilobular nodules are the infiltrates and small airways disease also happens in HP which causes the mosaic attenuation

Q10 | 42 yo male; COVID-19 April '20. Prolonged ICU stay on ECMO.



- A) COVID-ILD
- B) Atelectasis
- C) Interstitial lung abnormalities (ILA)
- D) COVID-fibrotic-like changes
- E) Resolving COVID-19-ARDS

Q10 | 42 yo male; COVID-19 April '20. Prolonged ICU stay on ECMO. **Final Diagnosis?**



- A) COVID-ILD
- B) Atelectasis
- C) Interstitial lung abnormalities (ILA)
- D) COVID-fibrotic-like changes
- E) **Resolving COVID-19-ARDS**

Question 1

A 72 year old man presents with a non-productive cough and exertional dyspnoea. He is an ex-smoker (50 PYH) and worked as a builder but does not recall being exposed to asbestos. He has no extra thoracic symptoms. What is the treatment of choice?

- a) NAC
- b) Ambrisentan
- c) Prednisolone
- d) Nintedanib
- e) Sildenafil



Question 1

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- b) Ambrisentan
- c) Prednisolone
- d) **Nintedanib**
- e) Sildenafil



Question 2

With regards to IPF, the following features are associated with a poorer prognosis:

- a) A TLCO <40% at diagnosis
- b) A fall in FVC $\geq 10\%$ over 6-12 months
- c) A fall in TLCO $\geq 15\%$ over 6-12 months
- d) Desaturation on exercise to less than 88%
- e) All of the above

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- c) A fall in TLCO $\geq 15\%$ over 6-12 months
- d) Desaturation on exercise to less than 88%
- e) All of the above

Question 3

Which of the following statements is **true** about nintedanib:

- a) It is recommended in patients with IPF with FVC 60-80% predicted
- b) The commonest side effect is anorexia
- c) The recommended dose is 300mg BD
- d) It may result in fewer exacerbations than pirfenidone
- e) Is an EGFR inhibitor

Question 3

Which of the following statements is **true** about nintedanib:

- a) It is recommended in patients with IPF with FVC 60-80% predicted NICE FVC 50-80%
- b) The commonest side effect is anorexia Diarrhoea
- c) The recommended dose is 300mg BD 150 BD
- d) It may result in fewer exacerbations than pirfenidone Time to first exacerbation is delayed
- e) Is an EGFR inhibitor

Question 5

Mr X has been on Nintedanib for 12 months. At the time of diagnosis his FVC was 2.7L and his TLCO 50%. His exertional symptoms have worsened over the past year. His FVC is now 2.65L with a DLCO of 25%. What is the next step?

1. Echocardiogram
2. Frusemide
3. Stop Nintedanib
4. Start Predniolone
5. Pulmonary Rehabilitation

Question 5

Mr X has been on Nintedanib for 12 months. At the time of diagnosis his FVC was 2.7L and his TLCO 50%. His exertional symptoms have worsened over the past year. His FVC is now 2.65L with a DLCO of 25%. What is the next step?

If u see a disparity between FVC and DLCO think of pul HTN. So echo would be first line

1. **Echocardiogram**

2. Frusemide

Drop in FVC >10%.

3. Stop Nintedanib

4. Start Predniolone

5. Pulmonary Rehabilitation

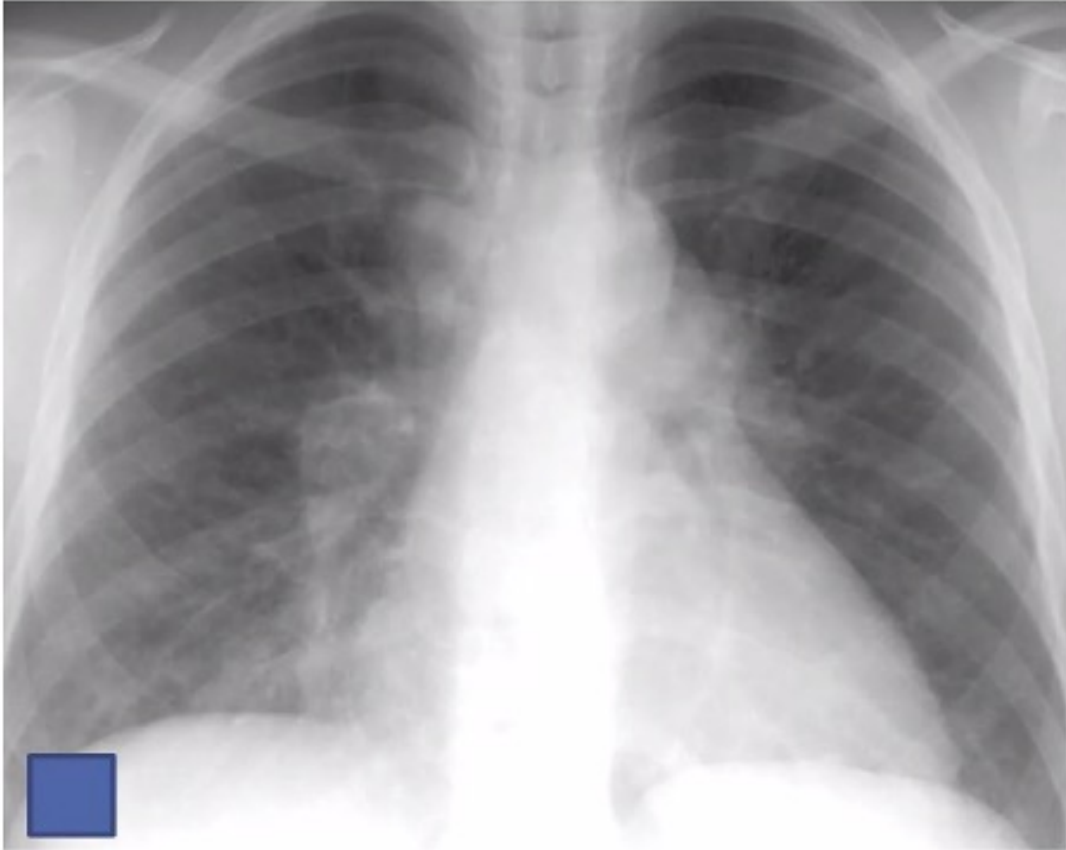
No 1st thing and should have happened already

Question 6

- 76 year-old male, Ex-smoker with established ILDon therapy presented to A&E with a rash. What is the next course of action?
- a) Half the drug dose?
 - b) Switch to a new drug
 - c) Pause the drug temporarily
 - d) Continue the drug
 - e) Start oral steroids?



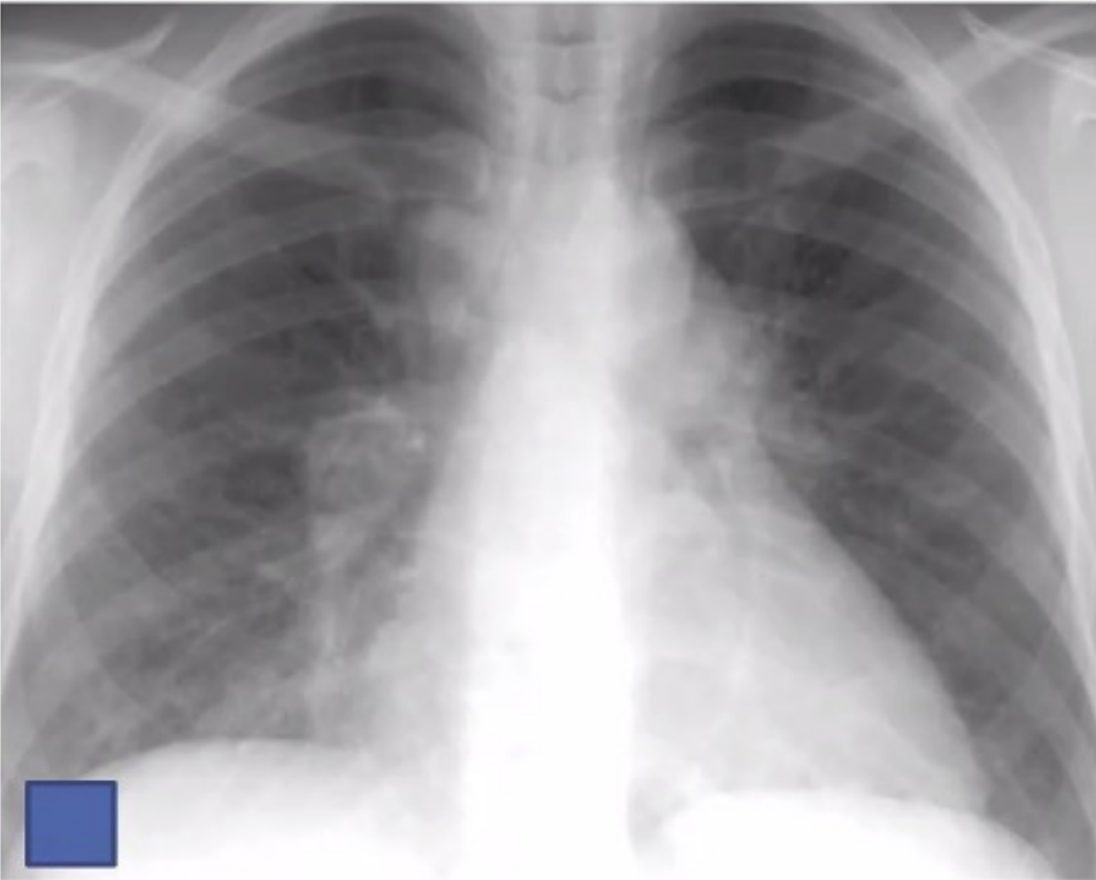
Question 7



A 30-year-old female presents to her GP practice with a dry cough, with no associated breathlessness/ chest pain and no systemic features of fatigue/weight low/ fevers. She has no known past medical history of note and is on no regular medications. A chest radiograph is performed to further investigate her cough. Blood tests reveal a raised calcium and a raised serum ACE. What stage is her disease?

- a) Grade 0
- b) Grade 1
- c) Grade 2
- d) Grade 3
- e) Grade 4

Question 7



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- b) Grade 1
- c) **Grade 2**
- d) Grade 3
- e) Grade 4

NOT CLEAR LUNG FIELDS, interstitial lung fields and BHL therefore stage 2.

Question 8

What would be the next investigation of choice?

- a) CT chest
- b) Echocardiogram
- c) ECG
- d) 24-hour urinary calcium
- e) EBUS + TBNA

Question 8

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- a) CT chest
- b) Echocardiogram
- c) ECG
- d) 24-hour urinary calcium
- e) EBUS + TBNA

ECG yes to check for cardiac sarcoid, Echo if ECG abnormal. CT chest though is first for diagnosis .

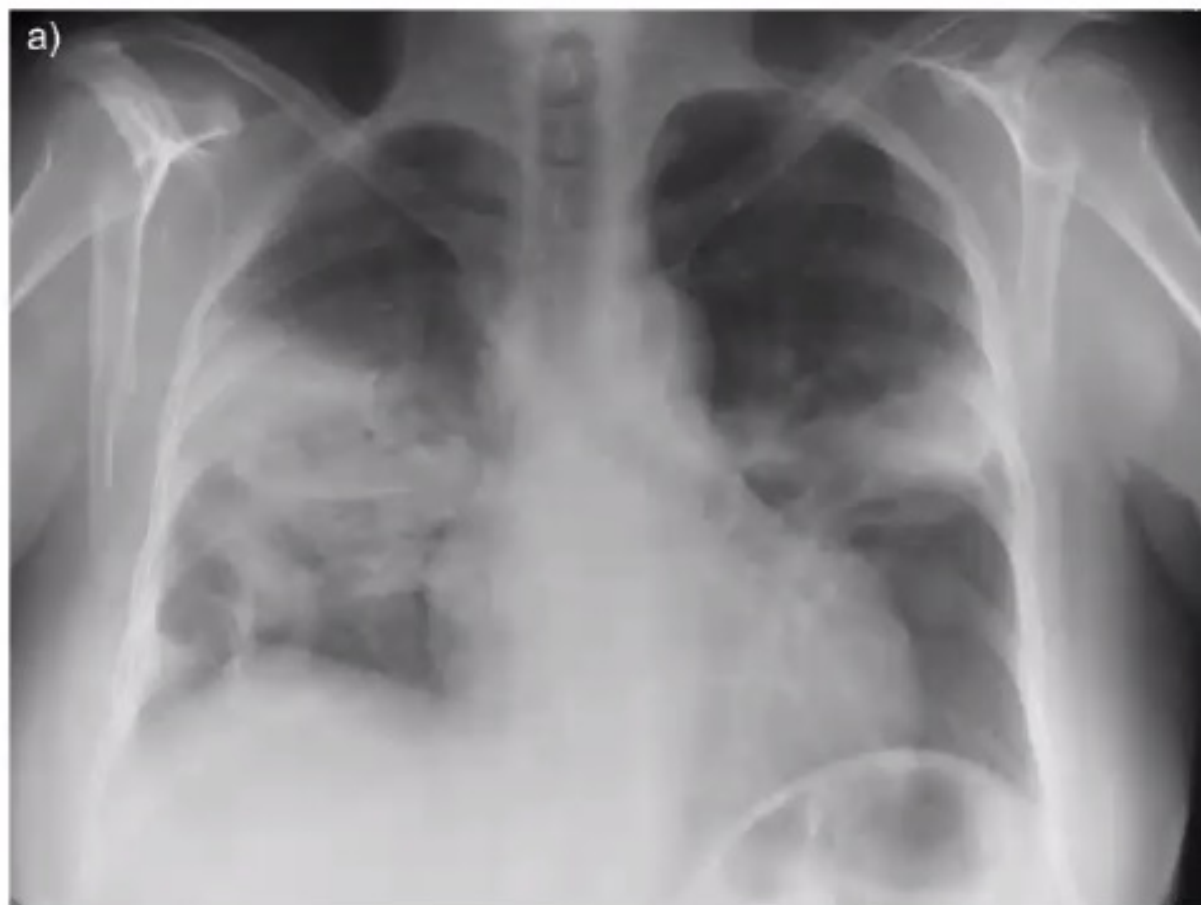
Question 9

- Which of the following medical conditions has the highest prevalence of pulmonary arterial hypertension?
 - a) HIV infection
 - b) Systemic sclerosis
 - c) Portal hypertension
 - d) Systemic Lupus Erythematosus
 - e) Sarcoidosis

Question 9

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 - e) Sarcoidosis

Question 10



A 40-year-old female and life long non smoker presented to her GP with a cough and dyspnea. A CXR showed RUL consolidation and she was treated with Augmentin and the cough resolved. Six weeks later she returns for a follow up CXR (shown) and now reports lethargy. Her FBC is normal and ERS raised at 70. What is the most likely diagnosis?

- a) Tuberculosis
- b) Alveolar proteinosis
- c) Hypersensitivity pneumonitis
- d) Organising Pneumonia
- e) Aspiration pneumonia

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- d) **Organising Pneumonia**
- e) Aspiration pneumonia

Question 11

- A 55 year old female with a 1-year history of cough and breathlessness has investigations which reveal ILD. A thoracoscopic lung biopsy shows lymphoid interstitial pneumonia. What is the most likely cause of her lung disease?
 - a) HIV
 - b) Idiopathic
 - c) Smoking
 - d) Occult neoplasia
 - e) Sjogren's syndrome

Question 11

- A 55 year old female with a 1-year history of cough and breathlessness has investigations which reveal ILD. A thoracoscopic lung biopsy shows lymphoid interstitial pneumonia. What is the most likely cause of her lung disease?
 - a) HIV
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Question 12

- A 29 year old man under the care of the ophthalmologists with low grade uveitis is referred by the medical opthamologists. He is a non smoker and works on a farm. His lung function is normal as are his bloods, except a mild lymphopaenia. What is the most likely diagnosis?

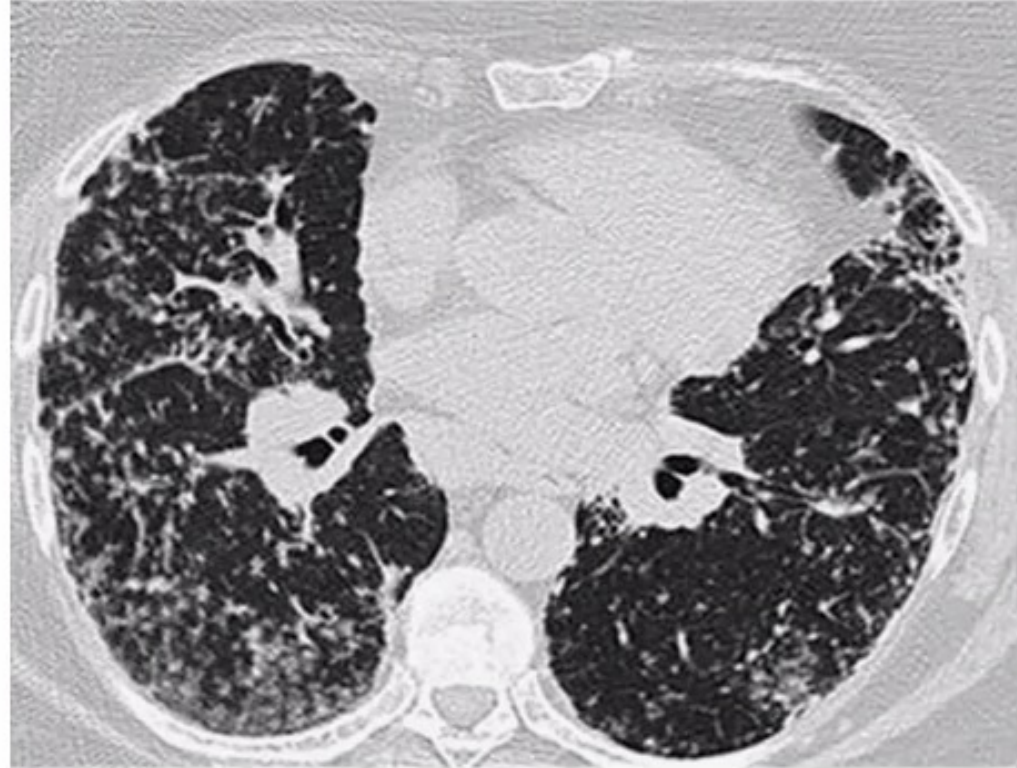
- a) Hypersensitivity Pneumonitis
- b) Idiopathic Pulmonary Fibrosis
- c) Miliary TB
- d) Sarcoidosis
- e) NSIP



Question 12

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- a) Hypersensitivity Pneumonitis
- b) Idiopathic Pulmonary Fibrosis
- c) Miliary TB
- d) **Sarcoidosis**
- e) NSIP



Lymphopaedia, uveitis, young

Miliary TB – would consider this, but no constitutional symptoms and radiologically the beading is along the bronchovascular bundles which is more like sarcoid

Question 13

- A 89 year old man was referred to chest clinic from the lung cancer screening program with an abnormal CT. He is a never smoker, worked in an office based job and is asymptomatic with normal spirometry. What is the most likely diagnosis?
 - a) Hypersensitivity Pneumonitis
 - b) Idiopathic Pulmonary Fibrosis
 - c) Interstitial Lung Abnormalities
 - d) Sarcoidosis
 - e) NSIP



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- d) Sarcoidosis
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Very subtle not widespread, screening project, no symptoms, incidental, minimal changes , basal disease in elderly people

Q1

A 65-year-old gentleman with COPD diagnosed 20 years ago attends the Chest clinic. He is a smoker of 20/day for the past 40 years. His current medications are Spiriva 18mcg OD, Keppra 250mg BD and sertraline 50mg OD. He was diagnosed with epilepsy as a teenager and has been seizure-free for at least 20 years. He enquires about smoking cessation. Which statement is **true**?

- a) He should use e-cigarettes as they are a safe and effective alternative to smoking
- b) Bupropion 150mg daily would be optimal treatment for him .
- c) Varenicline is a good option for him
- d) Pharmacotherapy and behavioural support in combination offers the best chance for continued smoking cessation
- e) Providing brief advice such as risks of smoking, assessing current and previous smoking behaviours is unlikely to help.

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Q2

A 70-year-old gentleman with COPD is referred to Chest clinic with progressive breathlessness. He is an ex-smoker, having stopped 10 years ago, with a 40-pack year history. His exercise tolerance 50 metres on a good day and has had no exacerbations in the past 12 months. His current medications are: Fostair 100/6 2 puffs BD and Braltus 10mcg OD.

Recent lung function tests are as below:

Post bronchodilator FEV1 35% predicted

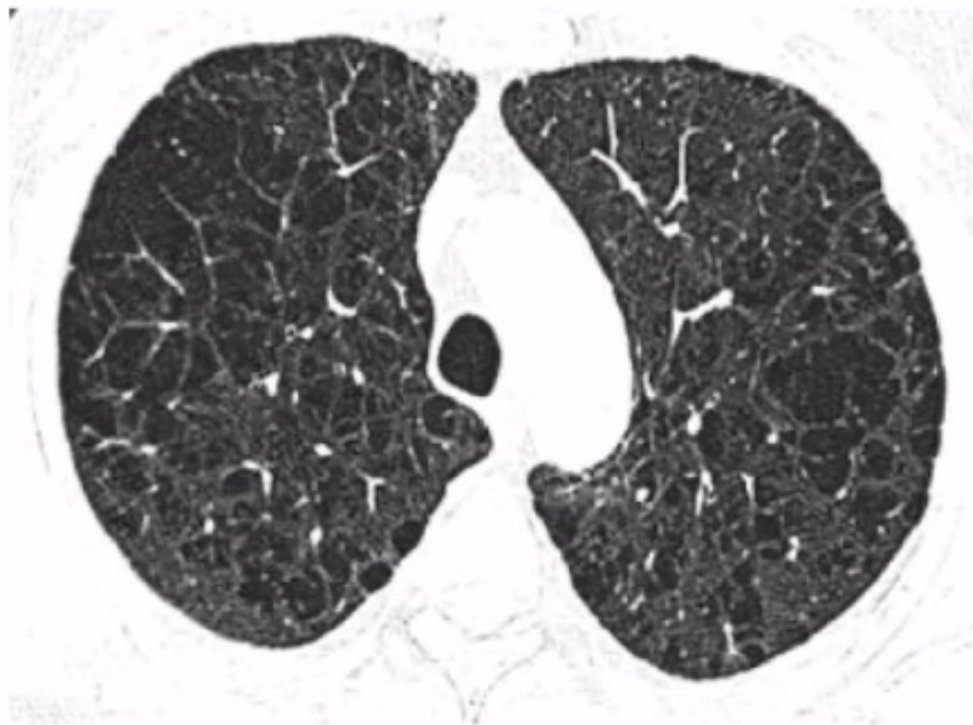
FVC 80% predicted

TLC 115% predicted

TLCO 60% predicted

RV 120% predicted

KCO 50% predicted



Q2

A recent echocardiogram shows normal LVEF, dilated RV with mildly impaired RV function. PASP is predicted at 45mmHg. ABG on room air, shows PH 7.43 pCO₂ 6.5 pO₂ 7.5 HCO₃⁻ 30, BE 5.

What intervention is most likely to improve his prognosis?

- a) Referral for lung volume reduction surgery
- b) Referral for LTOT
- c) Referral for home NIV
- d) Commencement of oral furosemide
- e) Commencement of Trelegy inhalers

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Q3

In a patient with COPD, in which of the following scenarios is hypoxic challenge testing not required when assessing fitness to fly?

- a) Previous hypoxic challenge testing performed last year is normal, 2 exacerbations in the past 12 months, including one hospital admission
- b) Saturations of 95% on RA with exercise induced desaturation to 85% RA
- c) Patient on LTOT at 2L/minute, no recent exacerbations
- d) Saturations of 94% RA and exercise desaturation to 83% RA
- e) Previous admission to hospital with acute decompensated T2RF and on regular co-codamol for lower back pain

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Q 4

A patient with COPD undergoes hypoxic challenge testing with FiO₂ 15% oxygen to assess fitness to fly to South America. ABG after testing is as follows: pO₂ 7.0 pCO₂ 5.0 pH 7.43 HCO₃⁻ 28 BE 4. Corresponding peripheral saturations are 85% RA .

What would you recommend with regards to need for oxygen?

- a) In-flight oxygen required at 2L/ minute
- b) In-flight oxygen not required
- c) Repeat testing as borderline results
- d) Needs LTOT
- e) In-flight oxygen to be prescribed as a PRN basis, to be used only if patient feels breathless

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Q 5

A 55-year-old gentleman with very severe COPD is admitted to hospital with acute decompensated T2RF secondary to IE COPD. He has been steadily deteriorating over the past year, with increased breathlessness on exertion and recurrent exacerbations. His BMI is 19kg/m². His 6MWD is 100m with desaturation to 85% on 6MWD performed 6 months ago. ABG on room air after treatment with BIPAP shows a pH of 7.4 paO₂ of 6.5, pCO₂ 6.0 HCO₃ 30 BE 4. He has been referred for LTOT assessment. Echo shows a dilated RV, but normal LV function. PASP pressures cannot be calculated. Lung function tests performed 6 months ago show :

Post bronchodilator FEV₁ 25% predicted, FVC 70% predicted, TLC 120% predicted, RV 100% predicted, TLCO 50% predicted, KCO 55% predicted

CT chest shows widespread centrilobular emphysema. He is currently on Trelegy inhalers and azithromycin 500mg MWF.

Which of the following is the **least** helpful management strategy for him?

- a) Switch Trelegy to Trimbrow
- b) Referral for lung transplantation
- c) Referral for lung volume reduction surgery
- d) Referral for consideration of home NIV
- ☒ e) Referral for pulmonary rehabilitation

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- c) **Referral for lung volume reduction surgery**
- d) Referral for consideration of home NIV
- e) Referral for pulmonary rehabilitation

FEV₁ <30% and widespread emphysema

Q6a

A 60-year-old gentleman who is an ex-smoker of 20 years is referred to the Respiratory clinic with 1 year history of worsening breathlessness. His FEV1 is 30% predicted, FVC 60% predicted, TLCO 85% predicted, KCO 90% predicted. He is now too breathless to leave the house and has had one hospital admission with IE COPD in the past year. His GP has ruled out other causes of breathlessness. He is on Braltus 10mcg OD and has recently completed pulmonary rehabilitation about 4 months ago.

What is his GOLD grade?

- a) A
- b) B
- c) C
- d) D

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What is his GOLD grade?

- a) A
- b) B
- c) C
- d) D

D

Q6b

Blood tests are performed in clinic for completeness which show the following:
Hb 170 WCC 5.3 Eosinophil count 0.3 Plt 450 U+Es normal.

What would be the next step in management for this gentleman?

- a) Add in Fostair 100/6 2 puffs BD
- b) Replace LAMA with Fostair 100/6 2 puffs BD
- c) Start triple therapy
- d) Switch Braltus to Anoro Ellipta
- e) Add in azithromycin MWF

Q6b

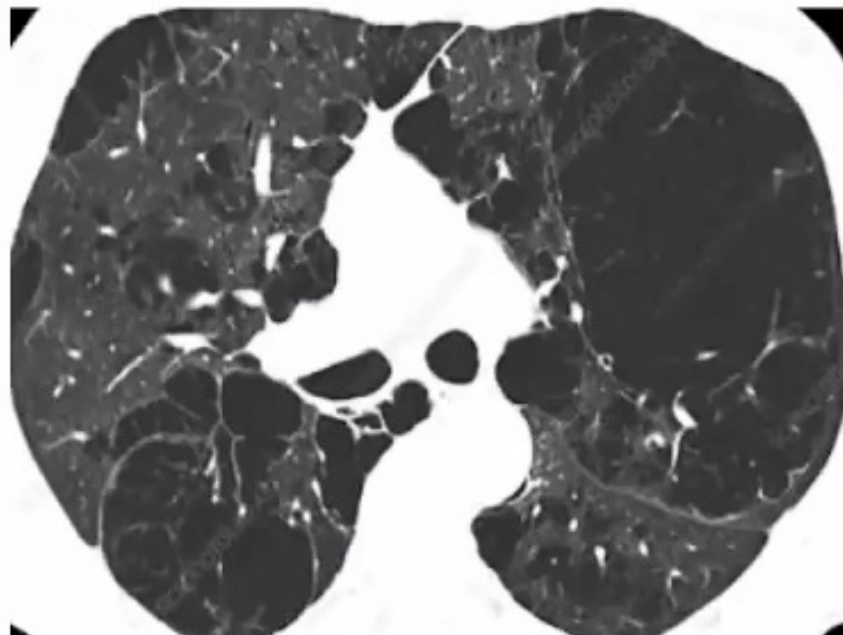
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- c) Start triple therapy
- d) Switch Braltus to Anoro Ellipta
- e) Add in azithromycin MWF

Q 7

A 75-year-old gentleman with COPD patient is admitted to hospital with drowsiness, and fever. He is barely rousable. ABG on room air shows a pH of 7.2 $p\text{CO}_2$ 8.0, $p\text{O}_2$ 6.5 HCO_3^- 25 BE 5. Blood tests show a WCC 6.5 Hb 130 Neut 7.8 CRP 200 Na 147 K 5.5 Urea 12.1 Cr 150 . He has had no previous hospital admission.



Q 7

What is the optimal therapy for him?

- a) Controlled oxygen, antibiotics, nebulizers, steroids and commencement of BIPAP, as ceiling of care
- b) Referral to intensive care for consideration of I+V If deteriorates
- c) Controlled oxygen, antibiotics, nebulizers and steroids and CPAP
- d) Palliation and end of life care
- e) Controlled oxygen, antibiotics, nebulizers and steroids alone

A

Q8

A 50-year-old gentleman is referred to the Respiratory clinic with progressive breathlessness. He is an ex-smoker, with a 10-pack year history. He has no childhood respiratory illnesses of note and has no atopic tendencies that he is aware of. He was given a trial of salbutamol inhalers with no improvement in his symptoms. CXR is normal and CTPA has ruled out pulmonary embolism/ significant parenchymal disease.

Recent blood tests are as follows: Total IgE mildly raised. Aspergillus IgE normal. Eosinophil count 0.4. RAST testing to cat/ dog/ tree pollen normal. RAST to HDM raised. His BMI is 30kg/m².

Lung function tests show: FEV1 50% predicted, FVC 90% predicted, TLC 70% predicted, TLCO 80% predicted, KCO 90% predicted. Bronchodilator reversibility: Improvement in FEV1 by 150 mls, 10% change. FENO 24

What is the most likely cause for his breathlessness?

- a) COPD
- b) Asthma
- c) Hypersensitivity pneumonitis
- d) Allergy induced breathlessness
- e) Raised BMI

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- c) Hypersensitivity pneumonitis
- d) Allergy induced breathlessness
- e) Raised BMI

Might be a case alpha-1 antitrypsin

Q9

A patient with COPD whom you see in clinic is suffering from recurrent exacerbations and you counsel him on starting azithromycin treatment. Which of the following offers optimal practice when commencing this treatment?

- a) ECG to assess QTC interval at baseline, and 1 month post starting treatment
- b) Liver function tests at baseline, 1 month post starting treatment and 6 months thereafter
- c) Counselling regarding potential GI side effects
- d) Sputum for AFB at baseline
- e) ECG and liver function tests at baseline and then 6 months into treatment

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