Navigating the identification, diagnosis and management of pulmonary hypertension using updated ESC / ERS guidelines

Case 2

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76 y old male: past history

14 years	deep vein thrombosis (1990)
1 year	gradual onset breathlessness
	ankle swelling
	Rx angiotensin converting enzyme inhibitor and betablocker
10 months	CTPA showed chronic thromboembolic pulmonary hypertension (CTEPH)
	Rx warfarin since then
	continued to deteriorate

76 y old male: presenting complaint

3 weeks worsening breathlessness
admitted to hospital with gross fluid retention

echocardiogram: peak TR velocity 4.2 m/s
severely dilated right ventricle
moderate pericardial effusion

referred urgently to Hammersmith Hospital

Question: Which one of these statements is correct?

- A significant number of patients with CTEPH do not have any history of pulmonary embolism or deep vein thrombosis
- It is recommended that patients with acute pulmonary embolism should be routinely screened for CTEPH during follow-up.
- CTEPH patients with at least moderate pericardial effusion should have this drained to prevent right heart tamponade
- Patients with CTEPH do not normally require more than 5 years anticoagulation
- All patients with CTEPH should have an inferior vena cava filter inserted

76 y old male: on admission

WHO functional class IV: can transfer bed to chair with difficulty; unable to walk to end of bed

<u>Examination:</u>

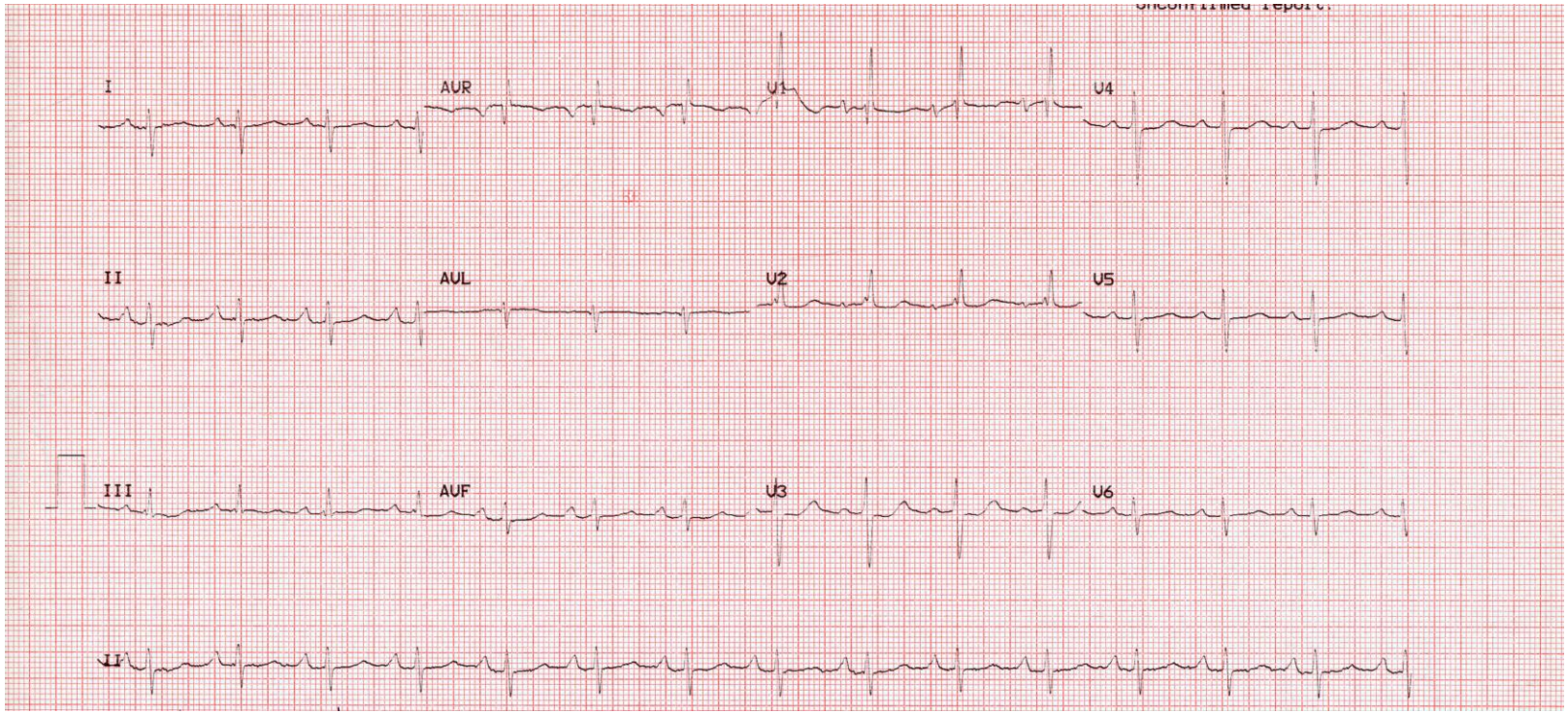
Heart rate 85 bpm, blood pressure 111/72, respiratory rate 18 /min, apyrexial, jugular venous pressure to ear lobes, SpO_2 94% on air

Loud pulmonary second sound with right ventricular third sound

Severe fluid retention

Cold peripheries

Resting ECG on admission



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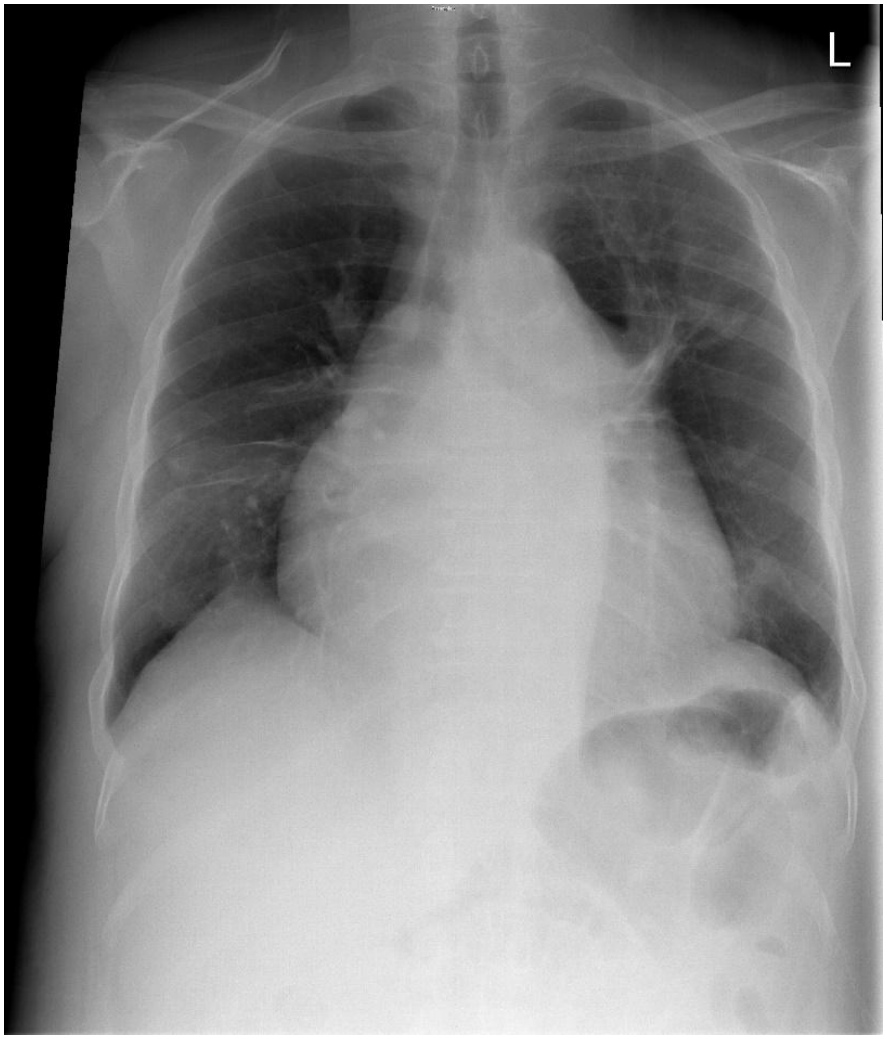
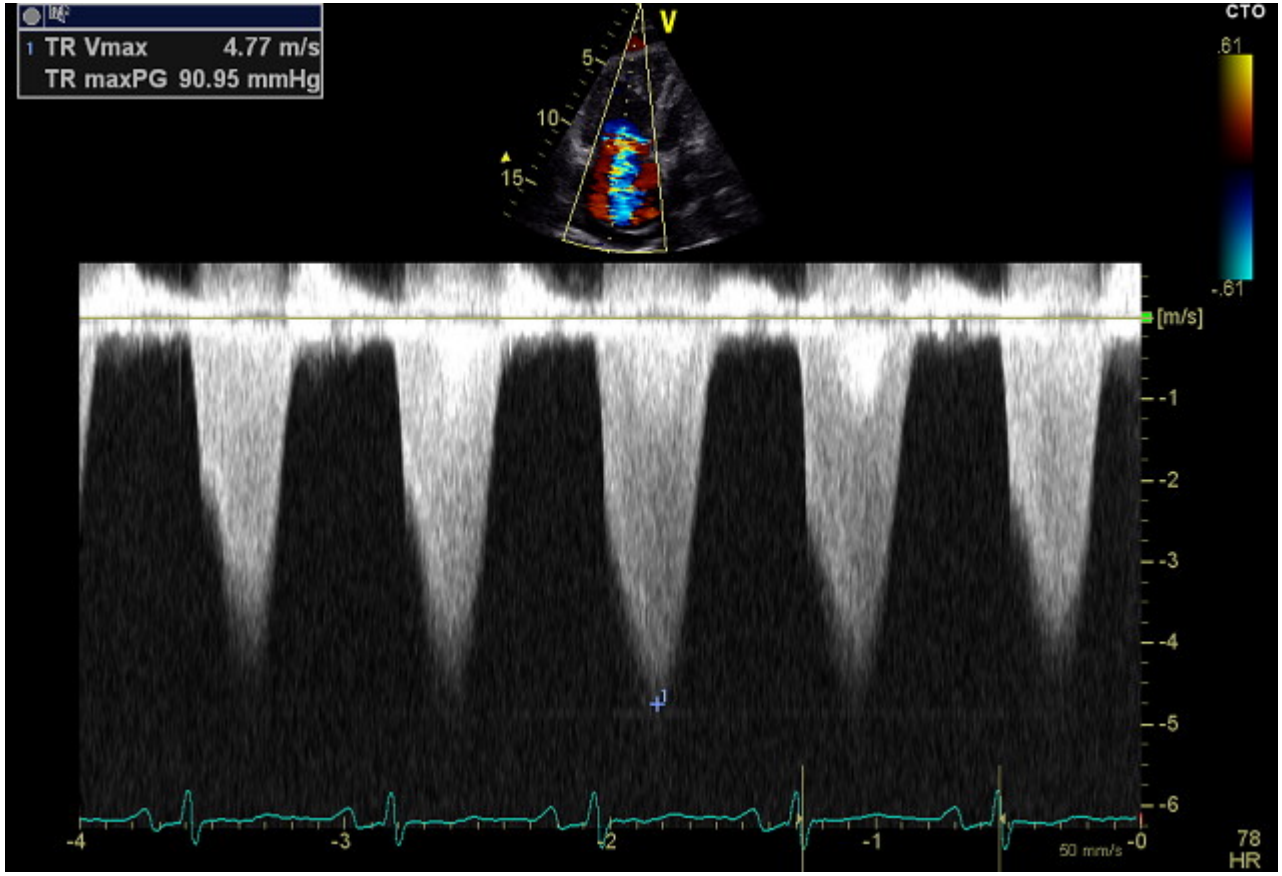
Chest radiograph on admission



Echocardiogram tricuspid regurgitation velocity



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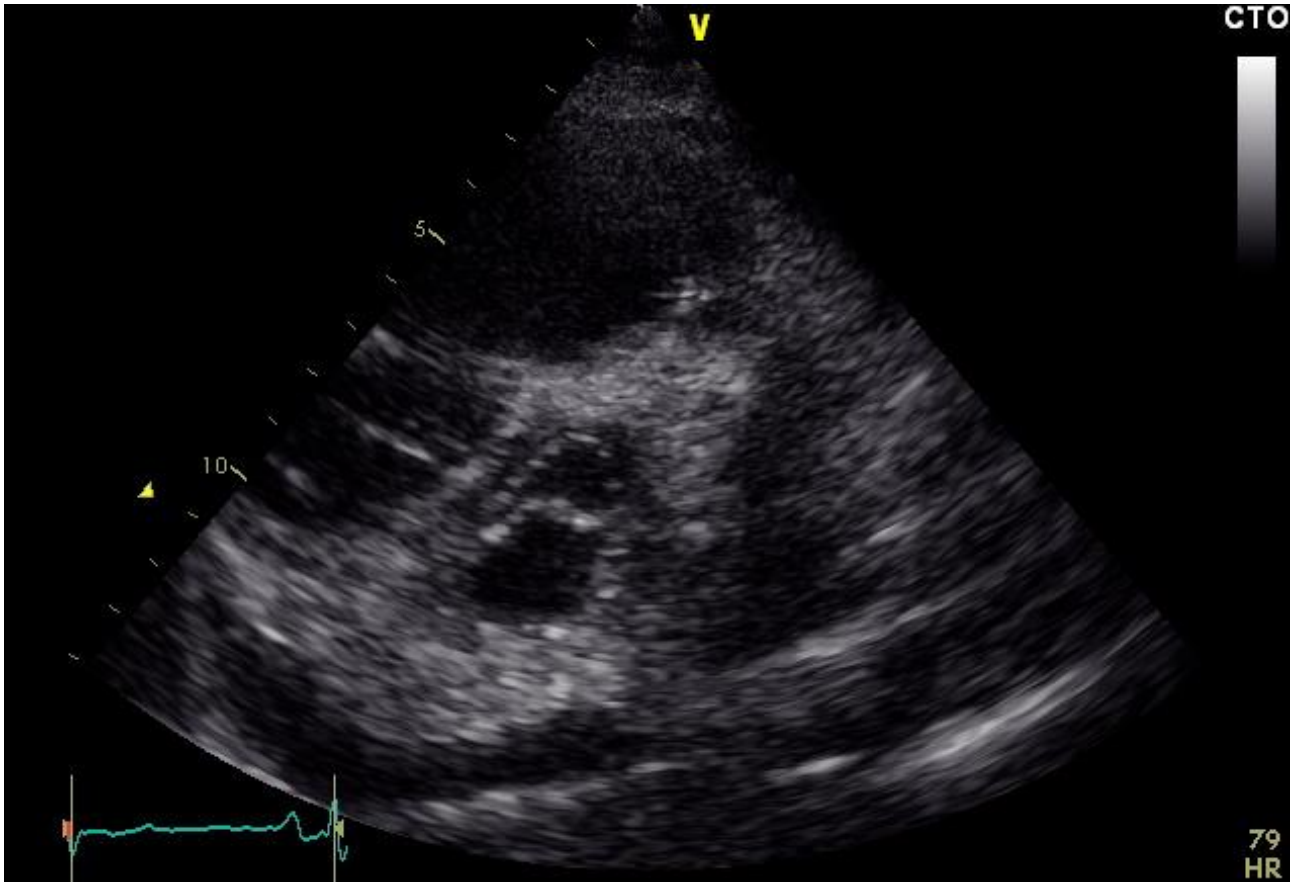
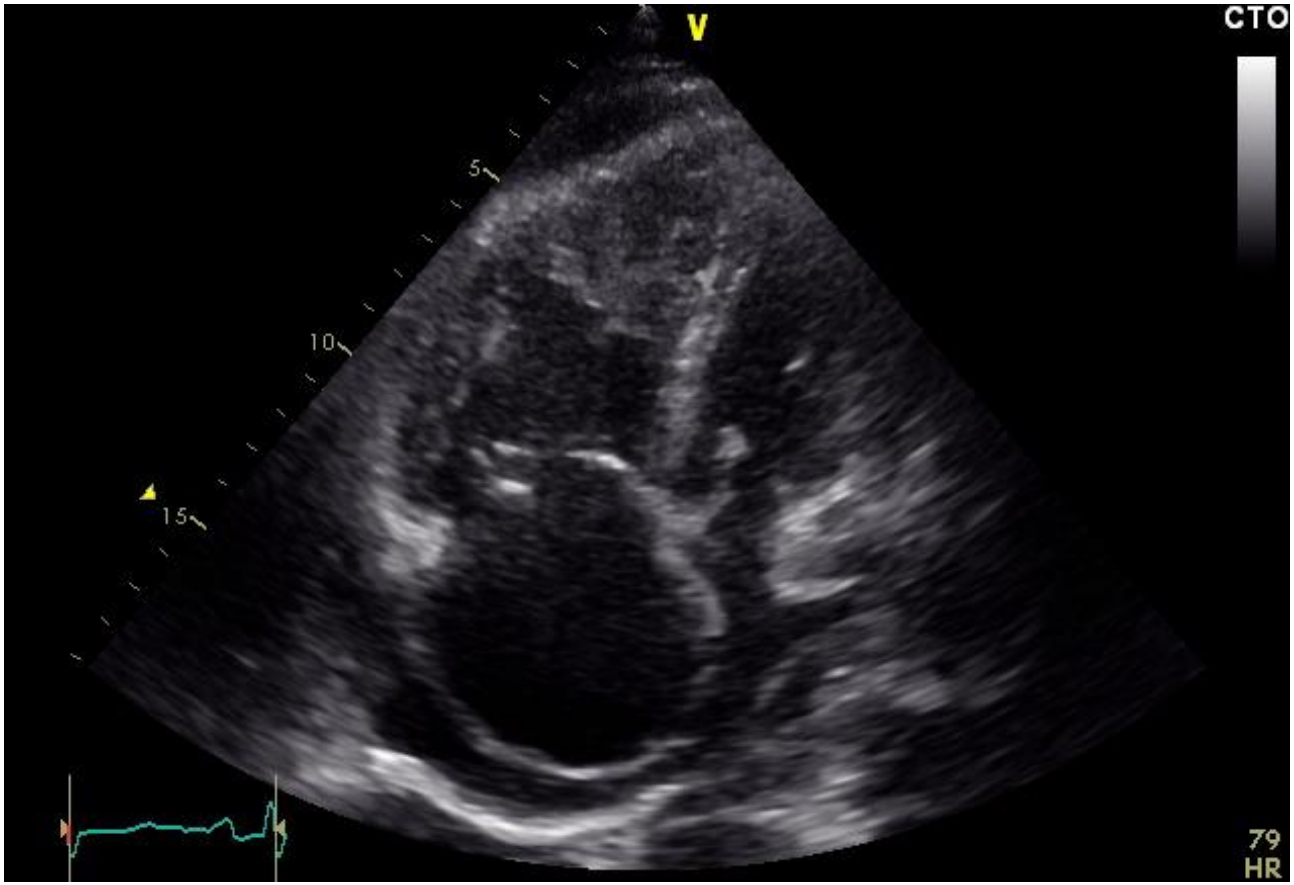
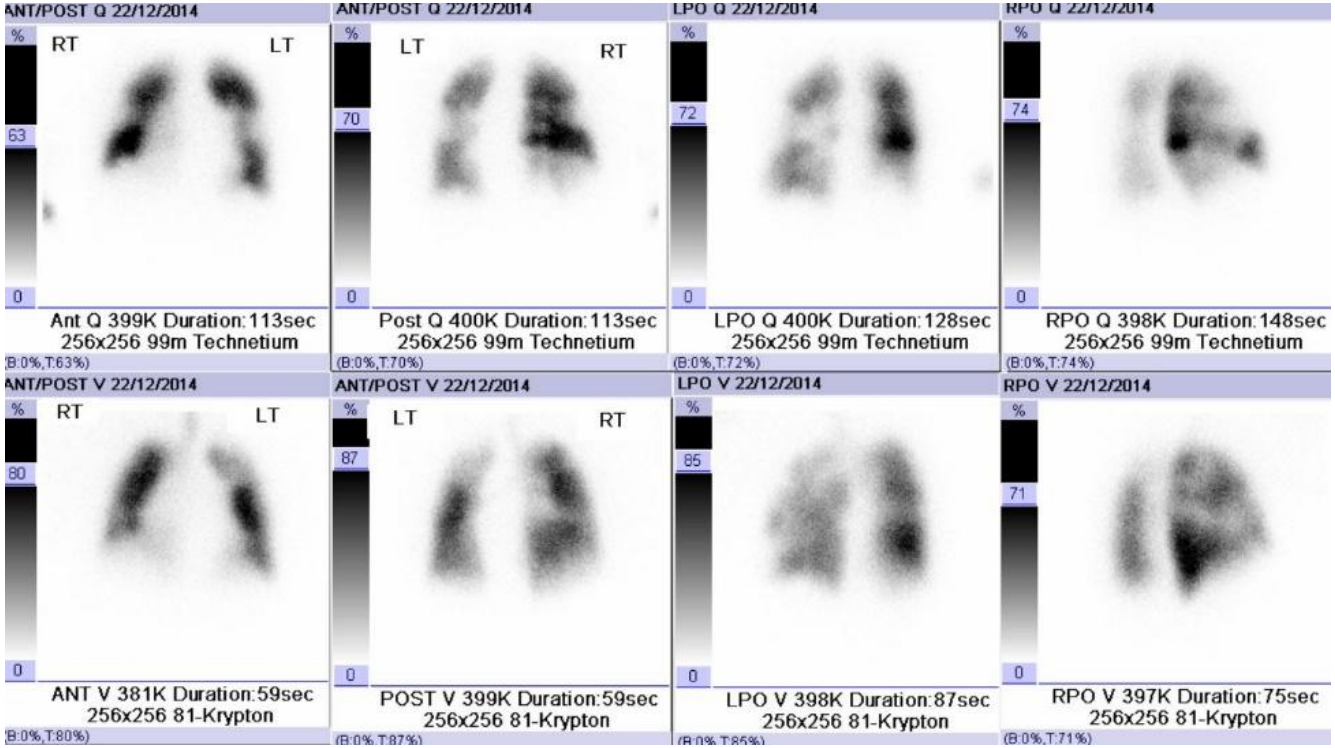
Echocardiogram parasternal short axis



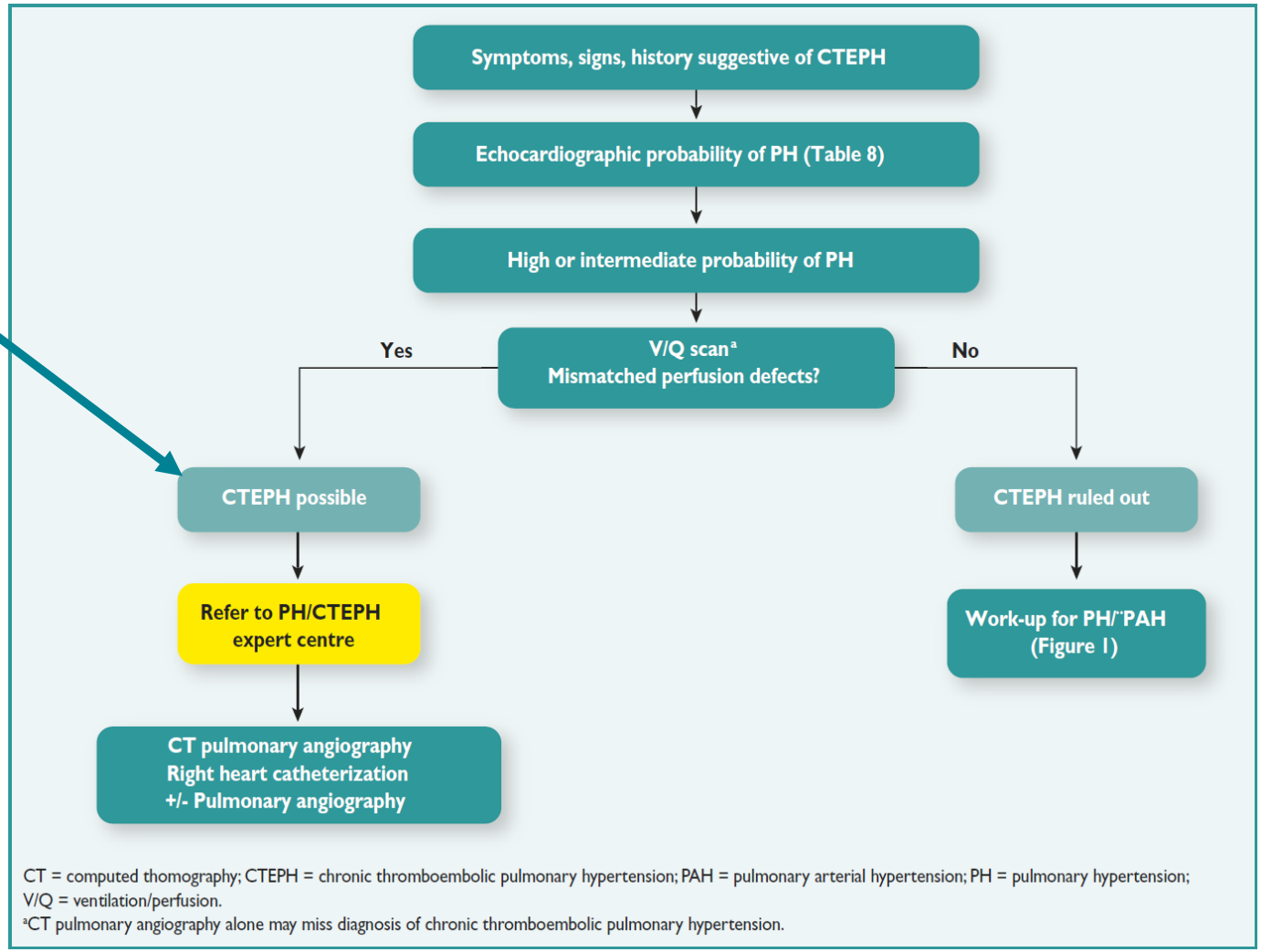
Echocardiogram apical 4 chamber



Ventilation perfusion scan



Diagnostic algorithm for CTEPH



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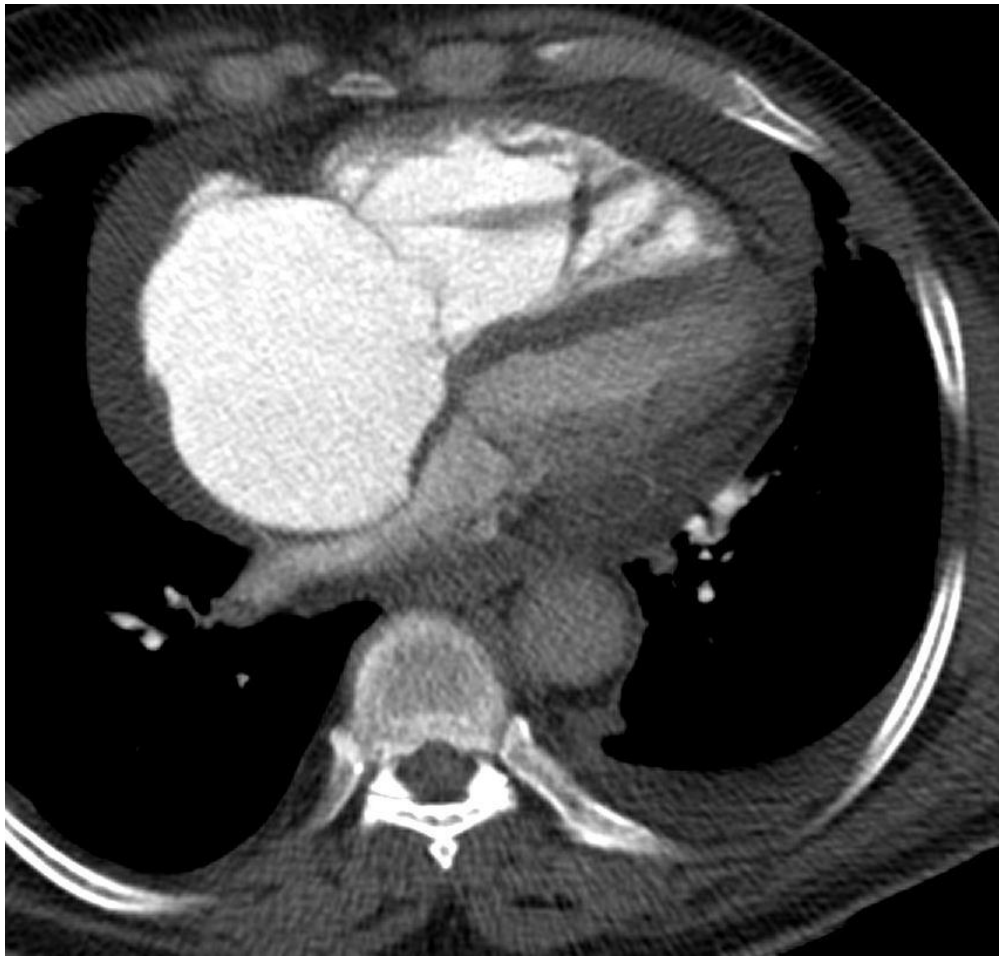
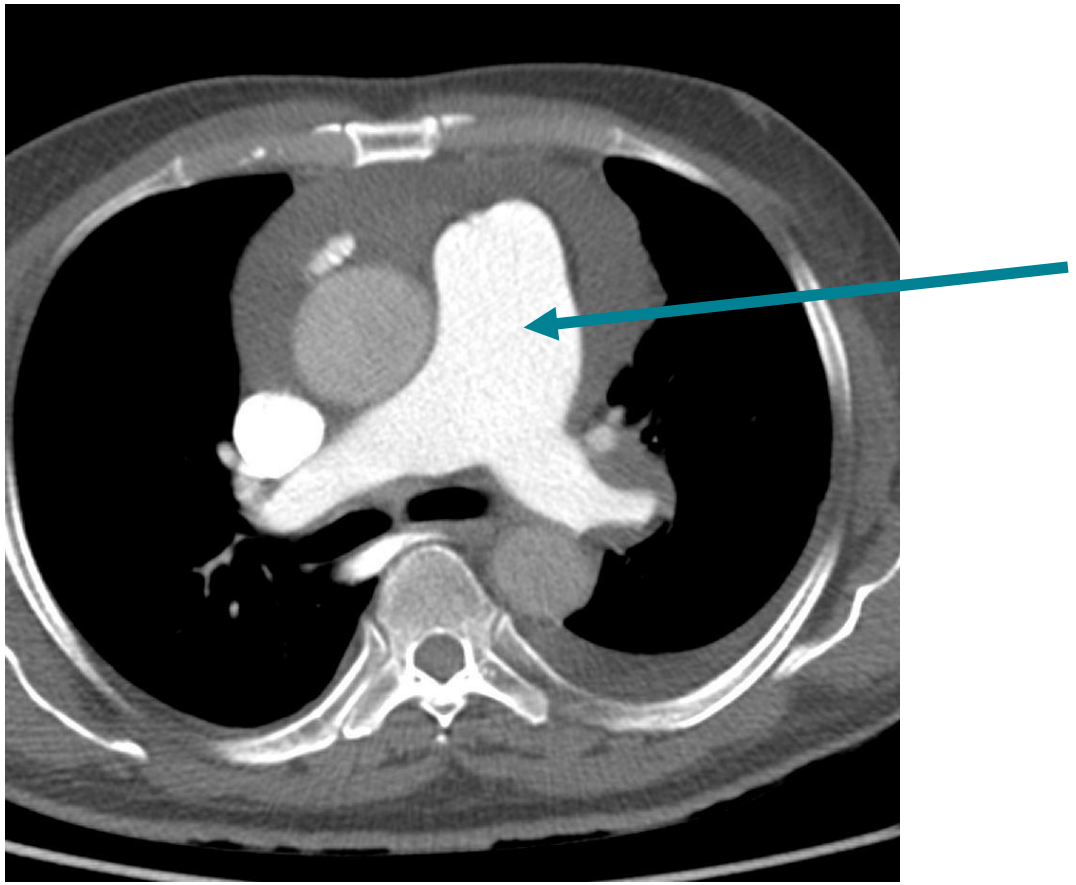
Galie N et al Eur Heart J 2016;37:67-119 & Eur Respir J 46:903-75

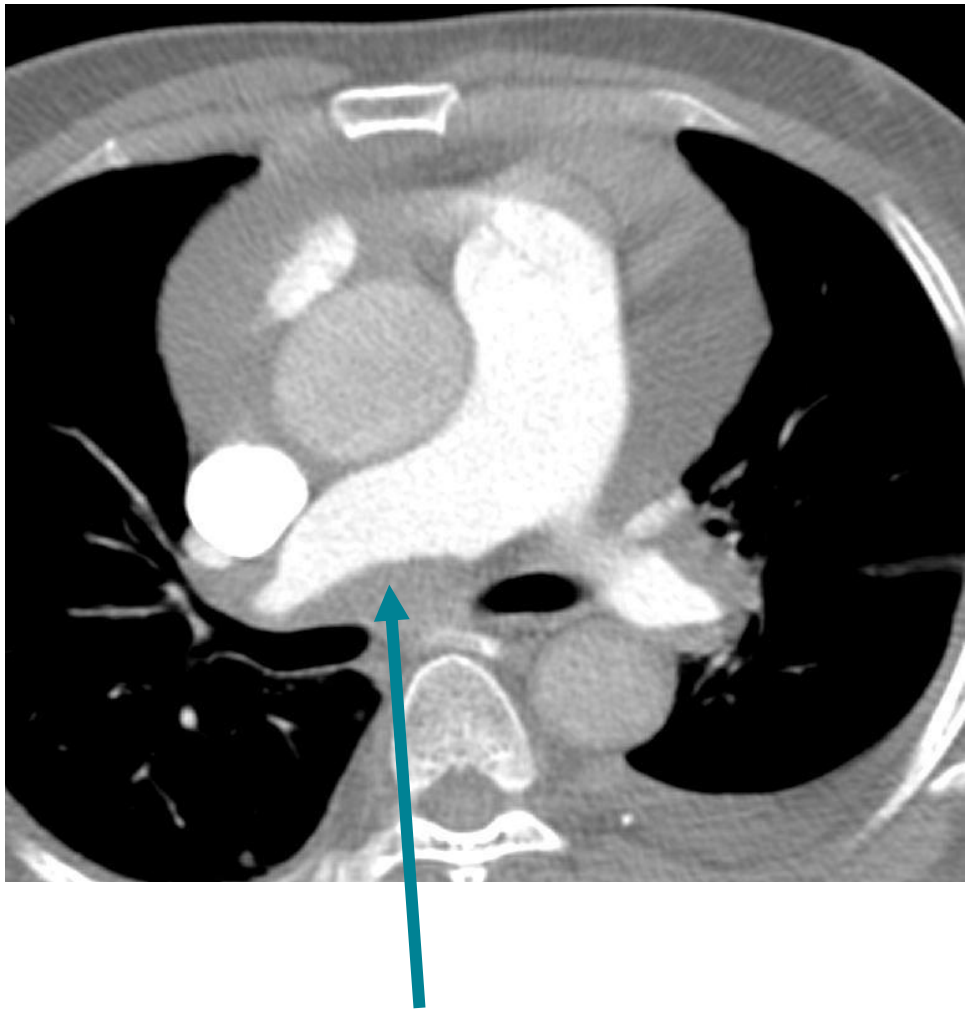
CT pulmonary angiogram showing enlarged right atrium and right ventricle with septal flattening



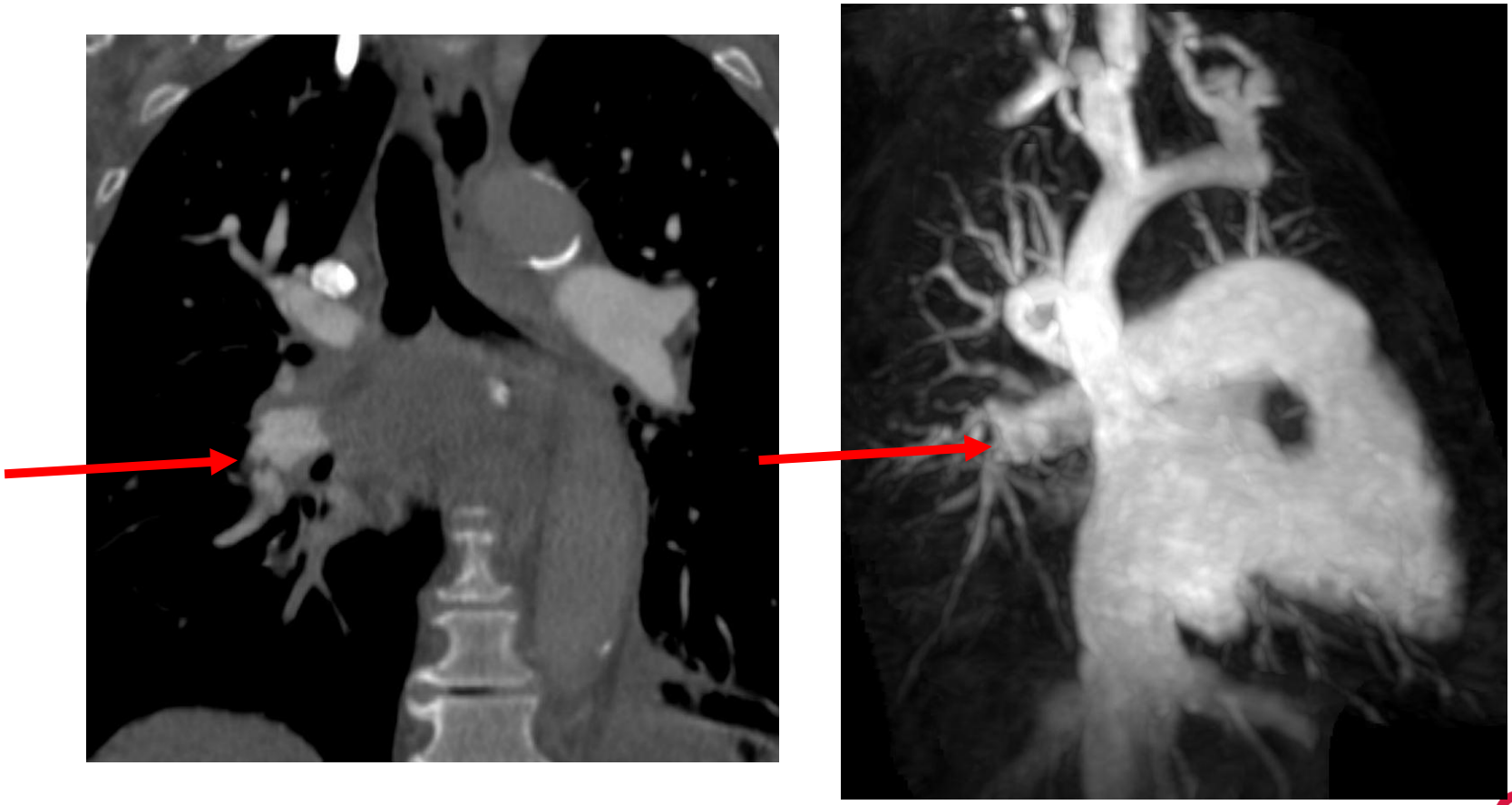
CT pulmonary angiogram showing enlarged main pulmonary artery



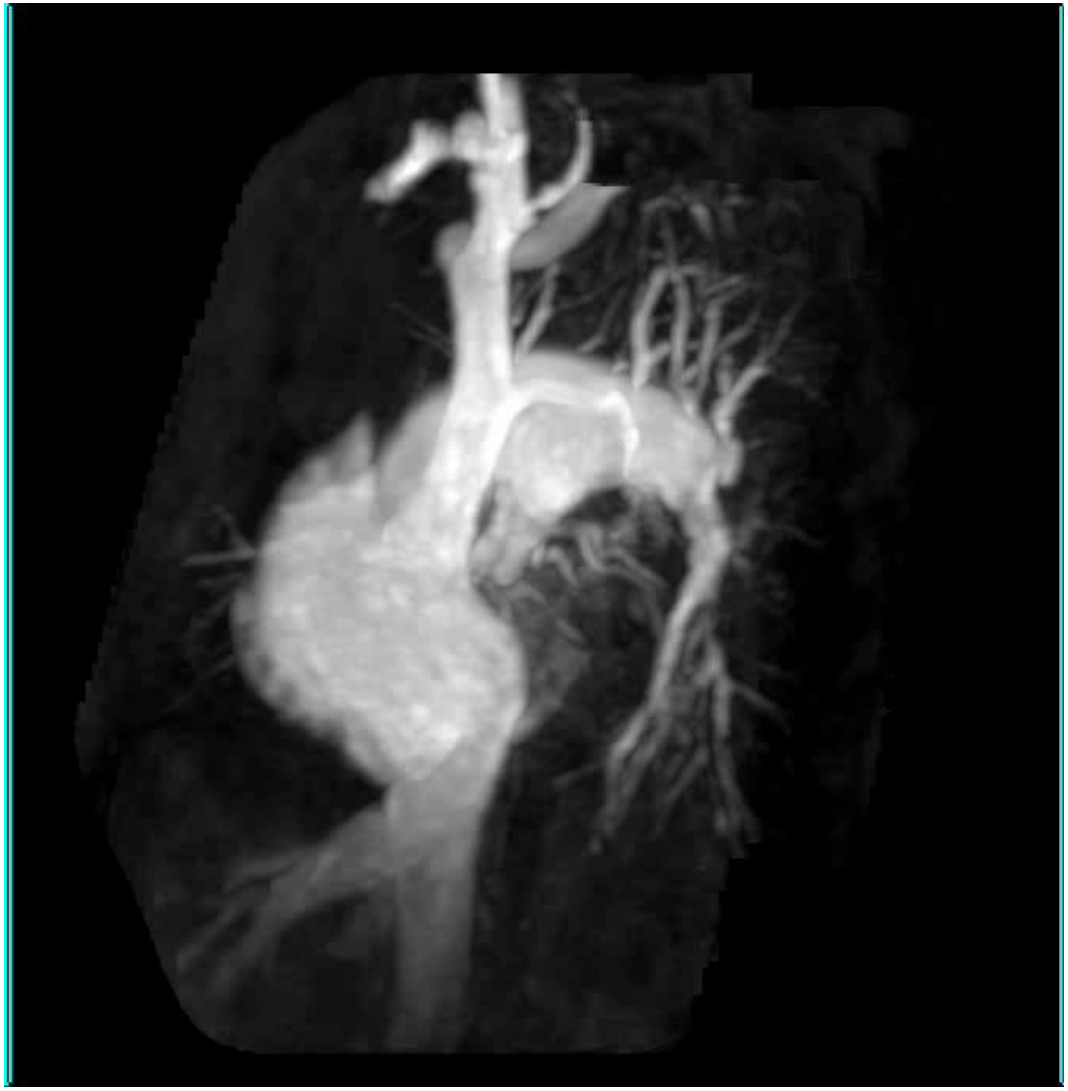
CT pulmonary angiogram showing chronic eccentric thrombus in right pulmonary artery



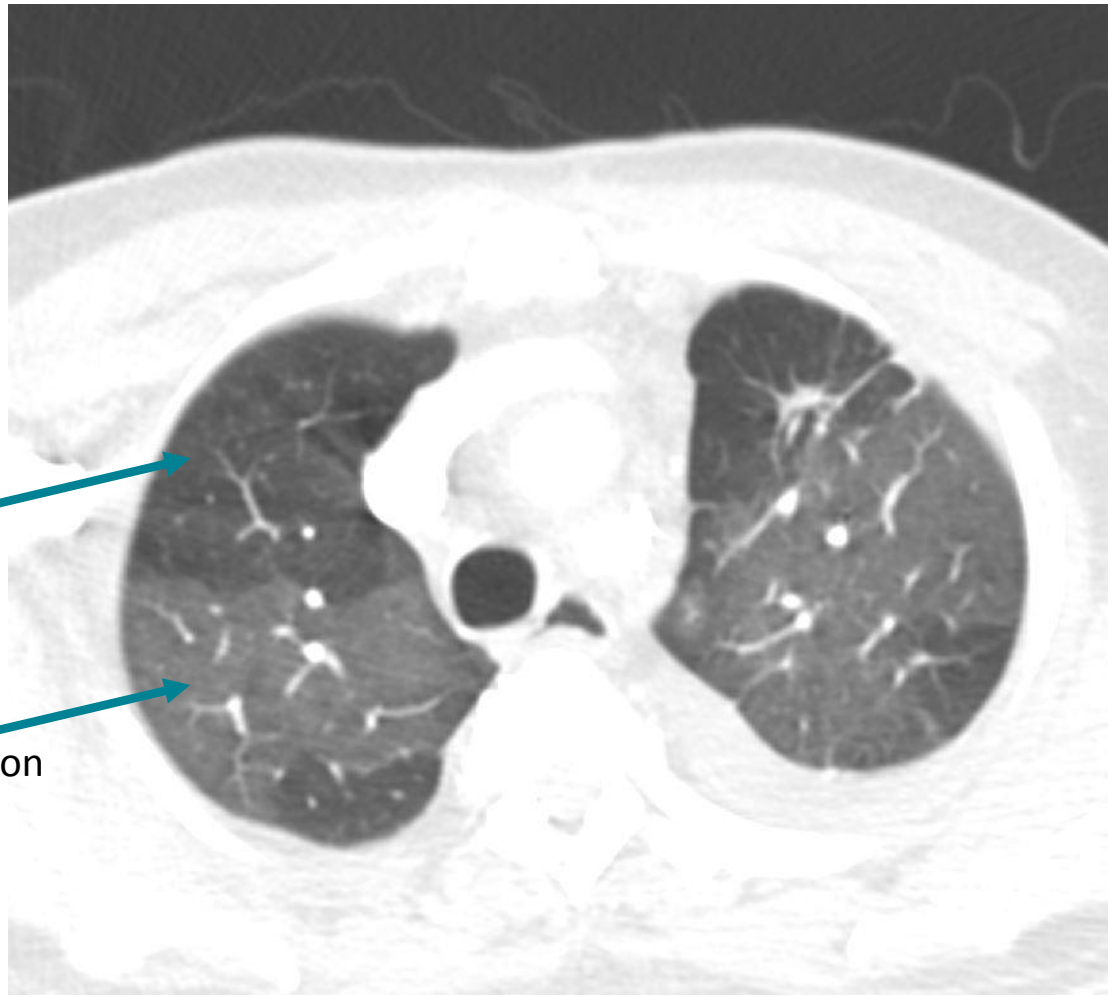
Coronal CT and magnetic resonance images of right lower lobe web

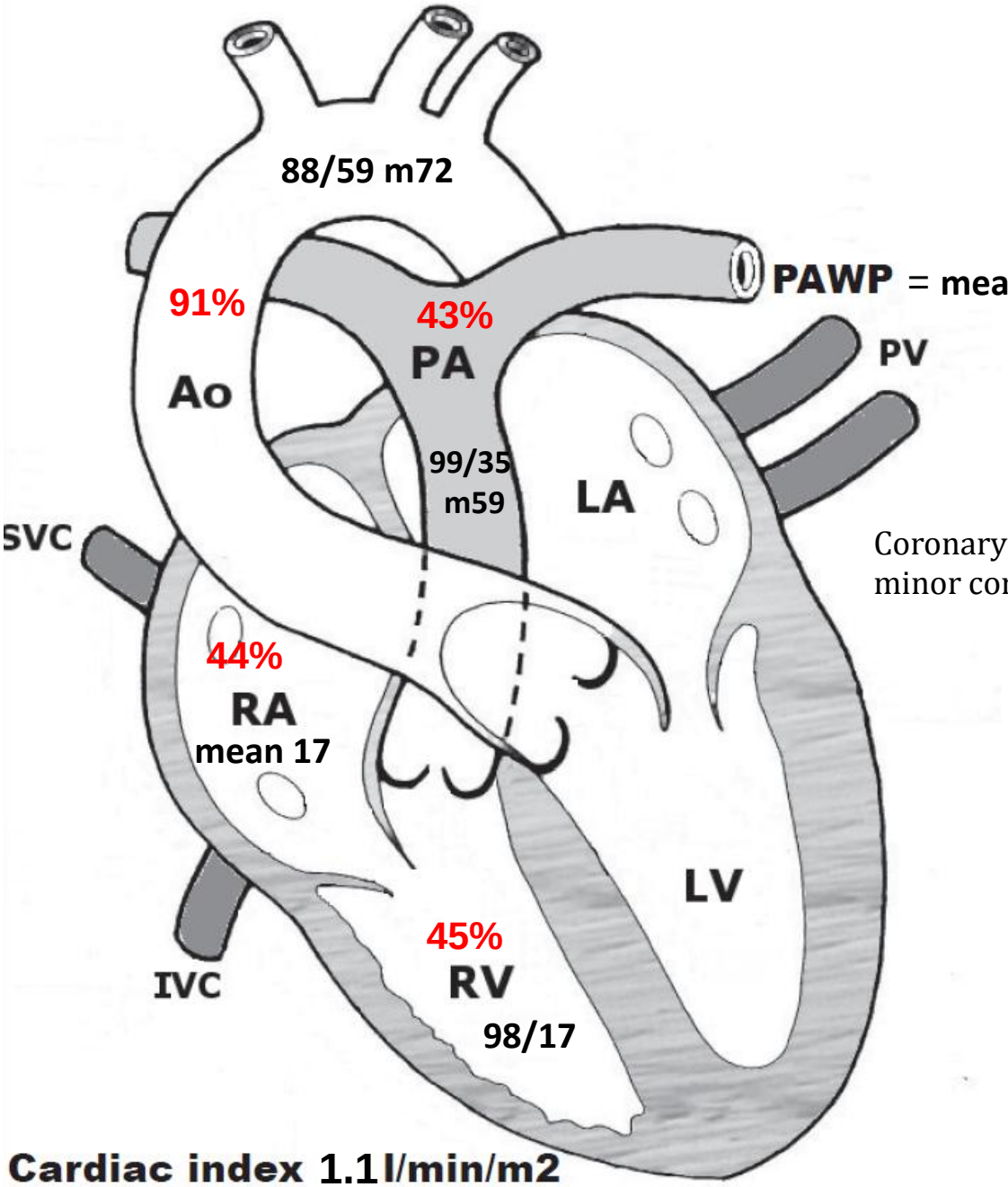


Magnetic resonance pulmonary angiogram (maximum intensity projection)



CT mosaic attenuation of lung parenchyma





Coronary angiography:
minor coronary artery disease

On 2 l/min oxygen
pH 7.47
pCO_2 4.4 kPa
pO_2 6.9 kPa

76 y old male

Diagnosis: CTEPH deteriorating in WHO functional class IV with severe pulmonary hypertension, severely elevated pulmonary vascular resistance and poor right ventricular function

Management:

Furosemide infusion

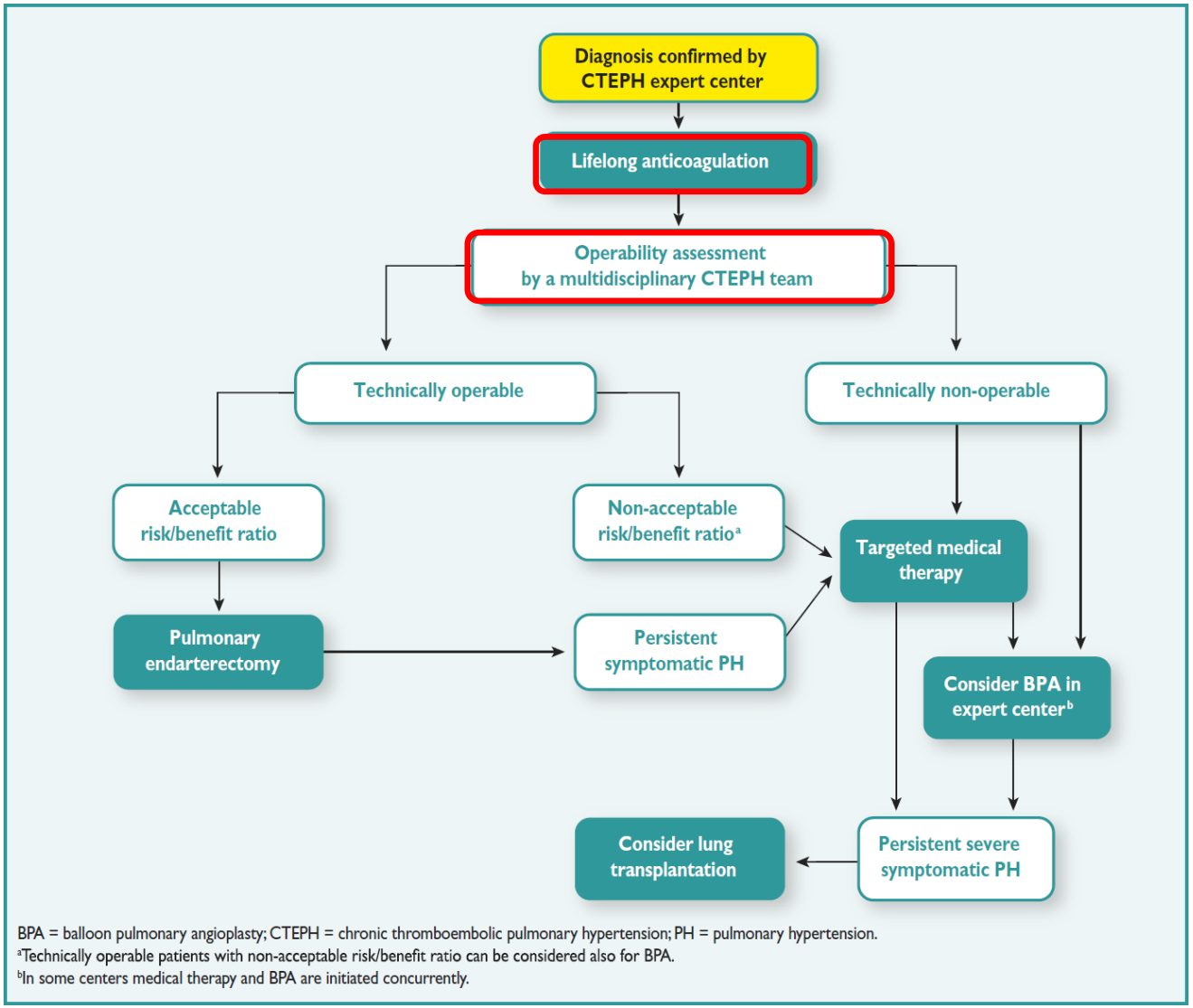
Dopamine infusion

Continue anticoagulation

Question: What further treatment would you recommend next?

- Pulmonary endarterectomy
- Balloon pulmonary angioplasty
- Riociguat for long-term treatment
- Riociguat to stabilize him, then pulmonary endarterectomy
- Intravenous epoprostenol infusion

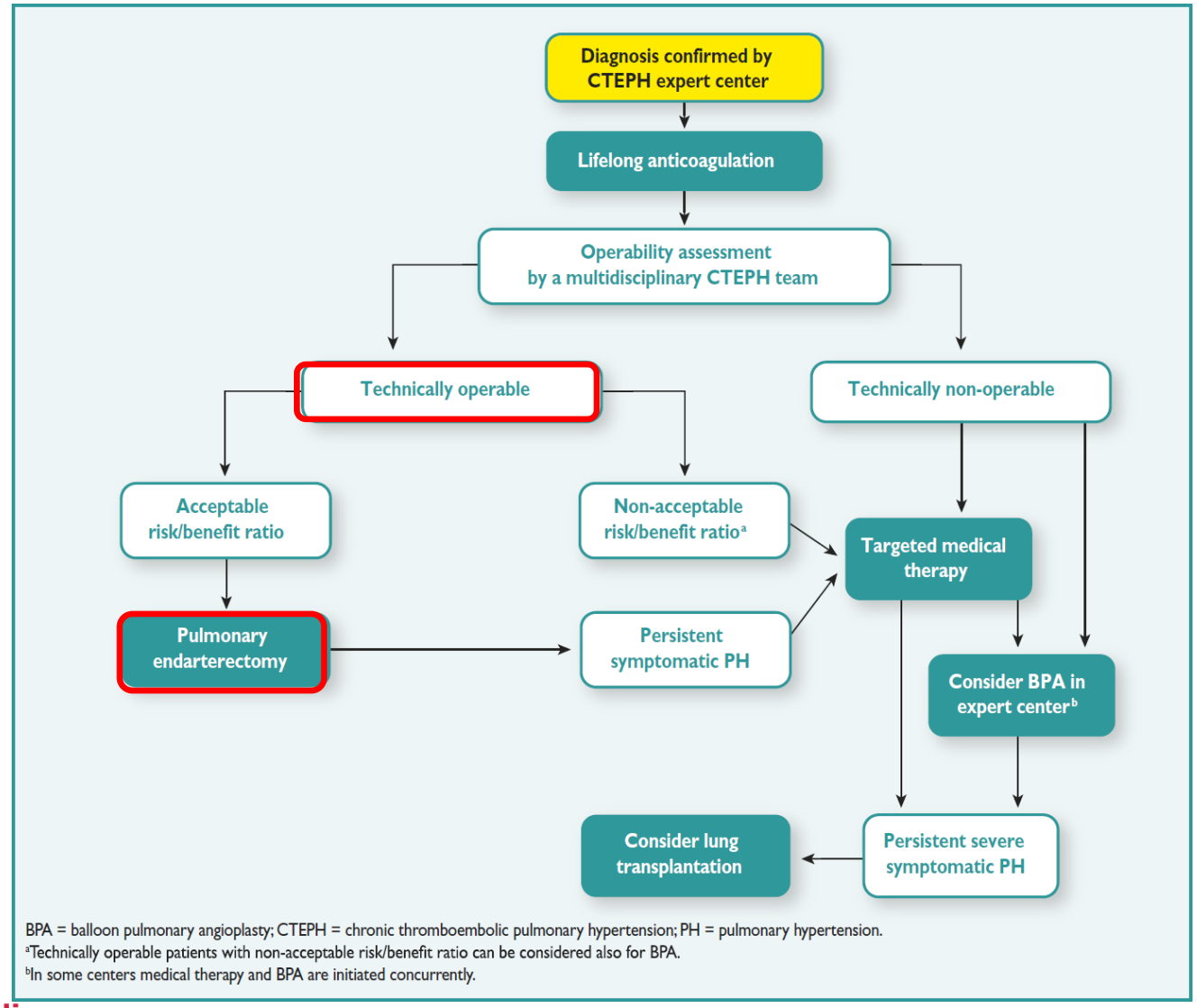
Management algorithm for CTEPH



BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic pulmonary hypertension; PH = pulmonary hypertension.
[a]Technically operable patients with non-acceptable risk/benefit ratio can be considered also for BPA.
[b]In some centers medical therapy and BPA are initiated concurrently.

Galie N et al Eur Heart J 2016;37:67-119 & Eur Respir J 46:903-75

Management algorithm for CTEPH



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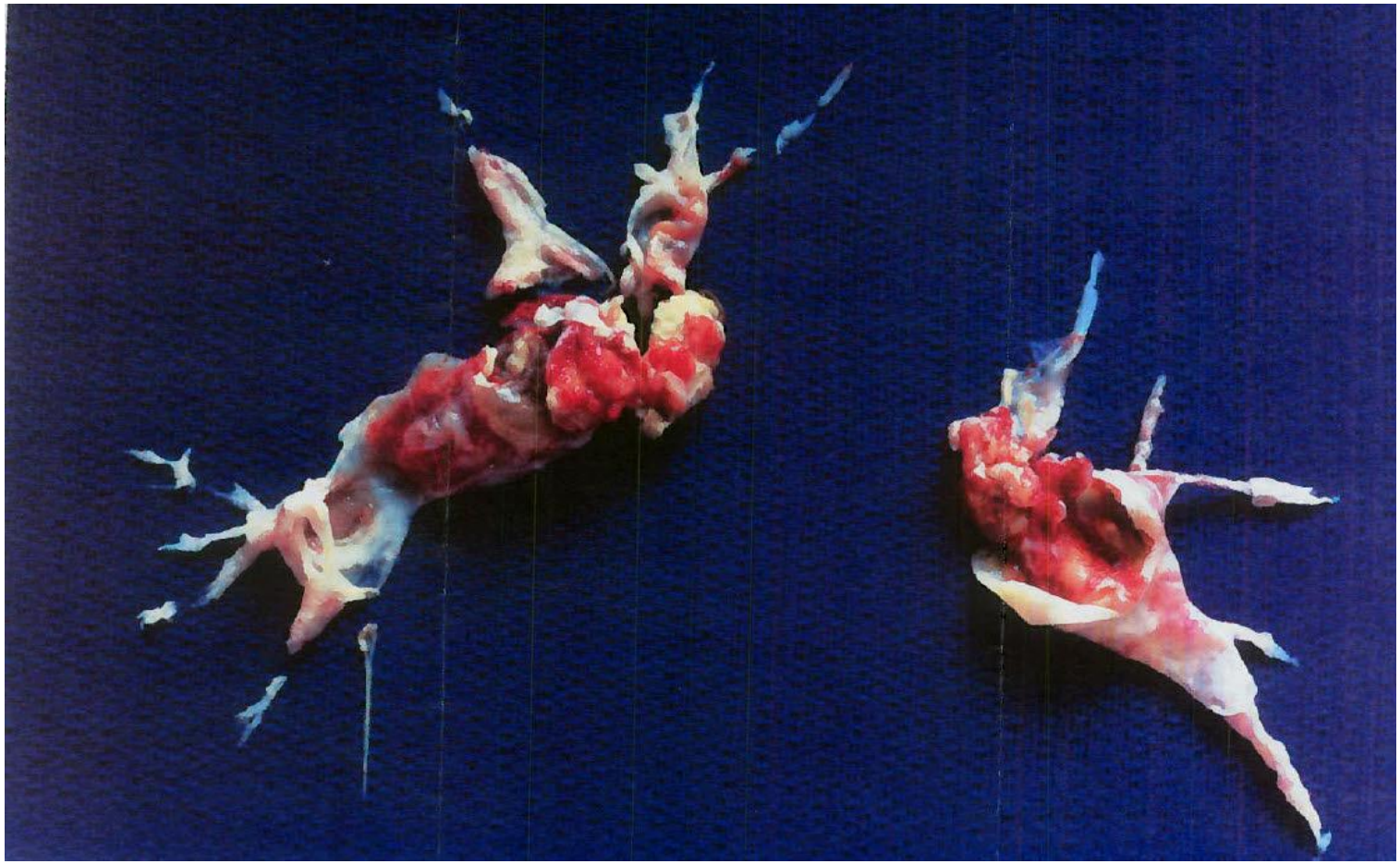
Pulmonary endarterectomy

Pulmonary artery pressure in the operating theatre:

pre-endarterectomy 121/40 mean 65 mm Hg

post-endarterectomy 51/10 mean 26 mm Hg



76 y old male: post operative outcome

Discharged home post surgery after 10 days

Progressive improvement in breathlessness and exercise capacity

By 3 months post operatively:

- **WHO functional class II**
- **Walking 2 miles per day**
- **mean pulmonary artery pressure 25 mm Hg**
- **cardiac index 3.1 l/min/m^2**
- **pulmonary vascular resistance 2.4 Wood units**

By 1 year his echocardiogram was within normal limits

Pulmonary endarterectomy is the treatment of choice for CTEPH

- Can be performed in 60% of CTEPH patients
- In hospital mortality 4.7%
- 3 year survival 89% (n=346, age 60 y)
- Bridging therapy with PAH drugs increases risk of death

Delcroix M et al Circulation 2016;133:859-871

Summary

A 76 year old male presented with gradual onset of breathlessness and heart failure over 1 year, presented in WHO functional class IV with severe oedema.

Investigations confirmed a diagnosis of operable CTEPH with severe right ventricular impairment.

Three months after pulmonary endarterectomy he had near normal haemodynamics and right ventricular function.

Key messages

- **CTEPH is treatable: always consider pulmonary endarterectomy!**
- **Always consider CTEPH if a patient with a previous pulmonary embolism presents with exercise induced breathlessness**
- **CT pulmonary angiography cannot exclude CTEPH: perform a nuclear ventilation perfusion scan**
- **Refer all patients with CTEPH to a multidisciplinary CTEPH team**

Thank you

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