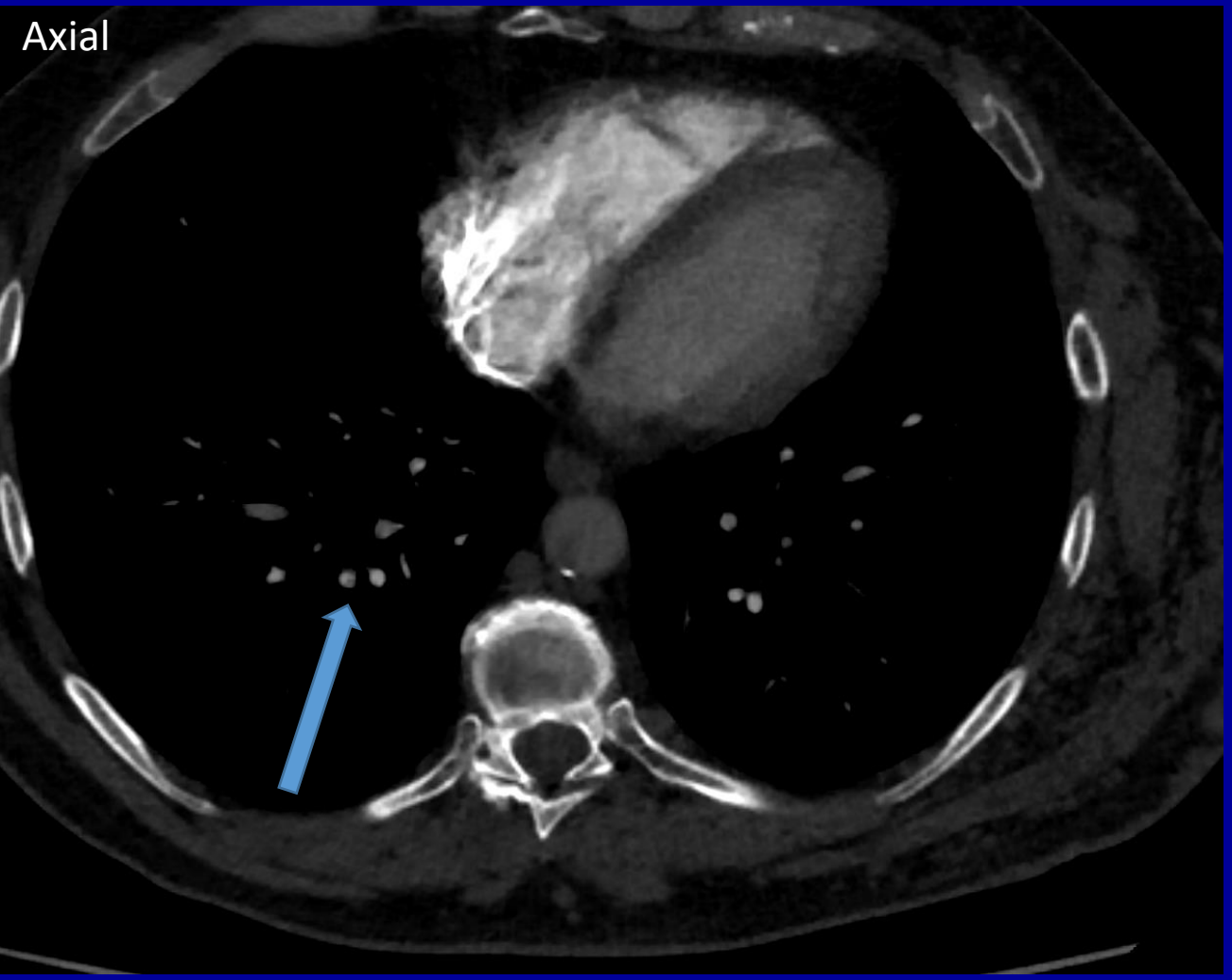
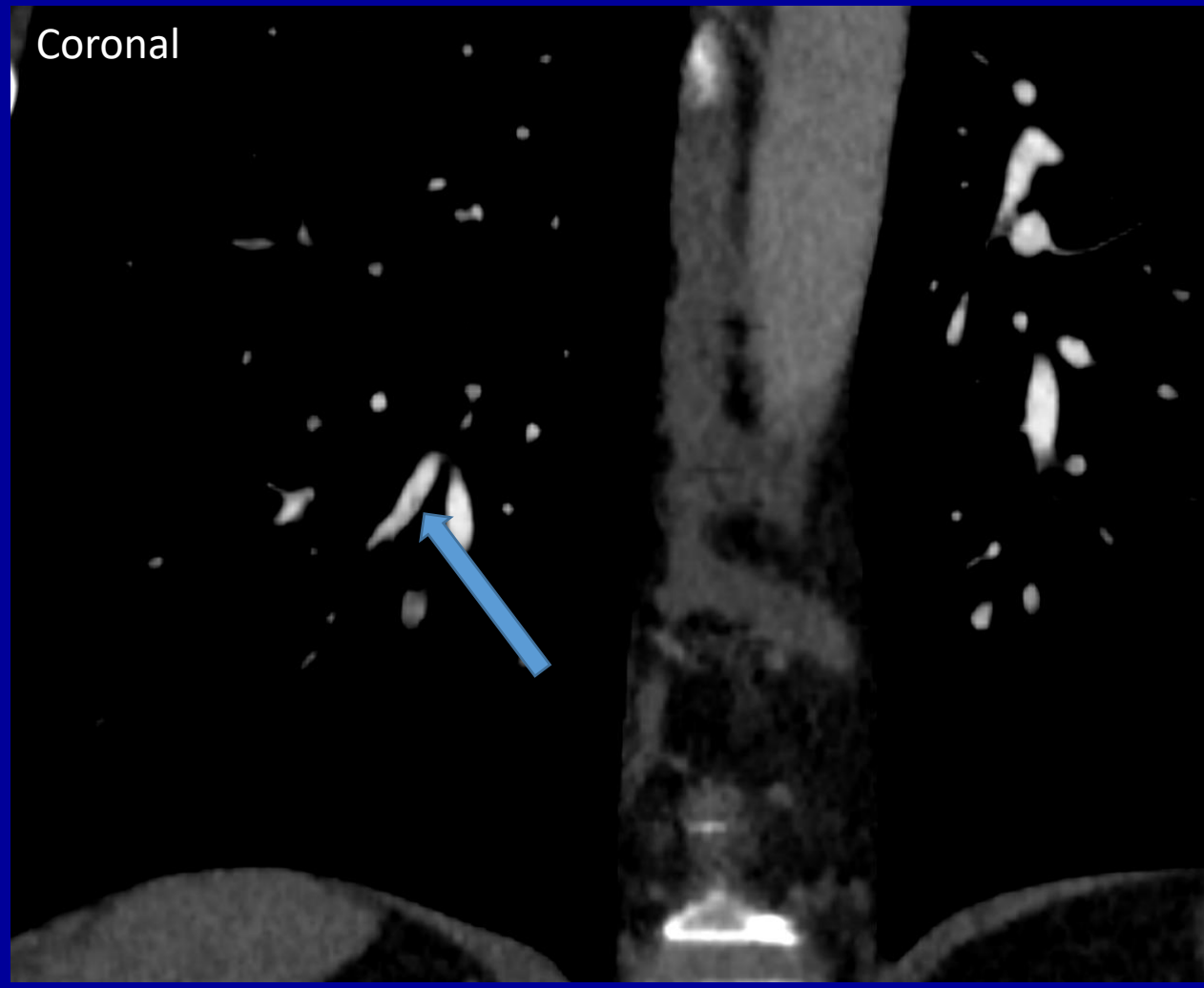
A rare case of pulmonary hypertension: Diagnosis, management and follow up by clinical VIR service.

CAIR 2022

Residents and Fellows day case submission

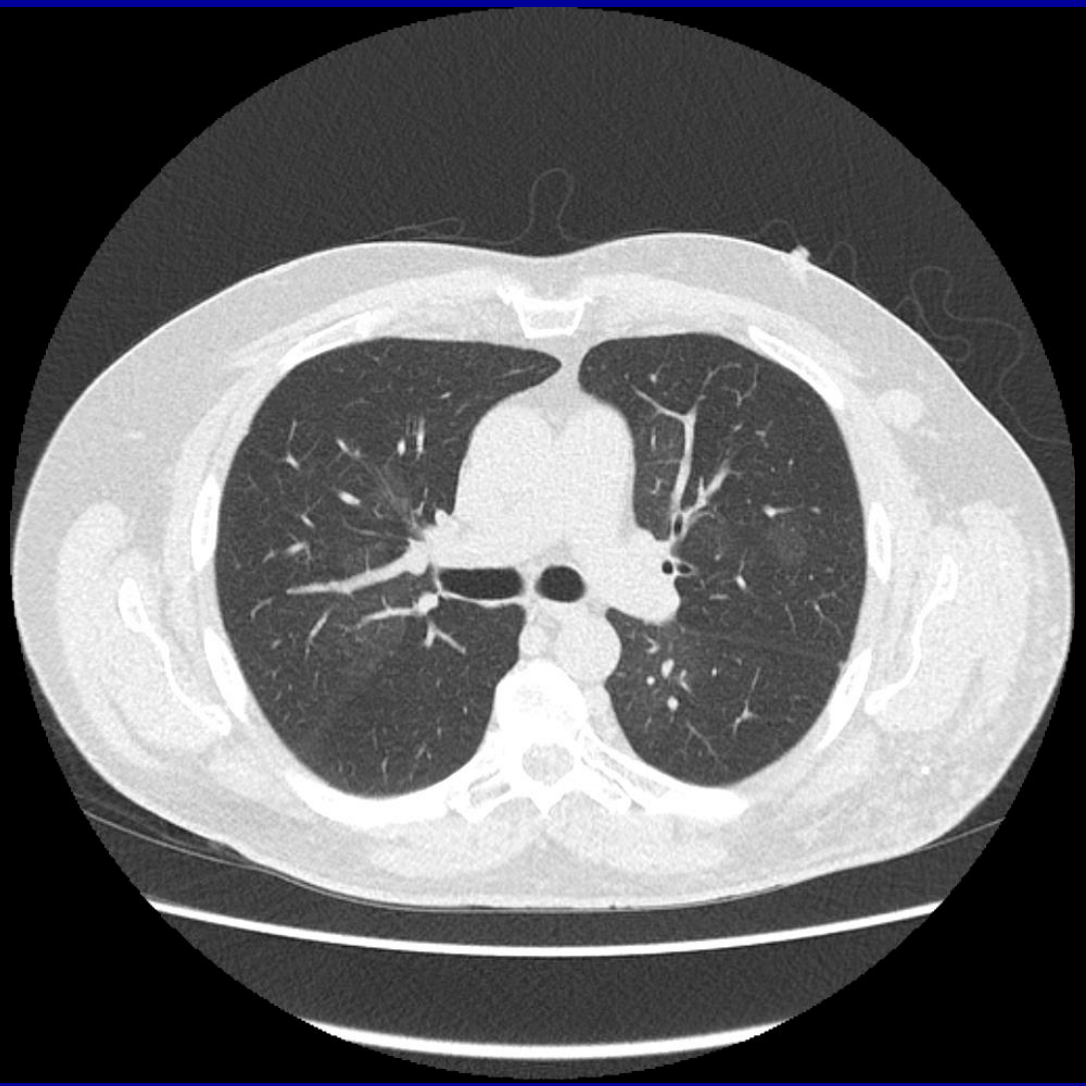
- 58 year-old man.
- 2-3 years of progressive shortness of breath. Limited to 1-2 flights of stairs.
- Desaturation to 80% on walking in room air. Met criteria for home oxygen.
- Left heart failure excluded by cardiology. Right heart cath: Mean pulmonary artery pressure (PAP) 50 mmHg (normal <25). Referred to pulmonary hypertension service.



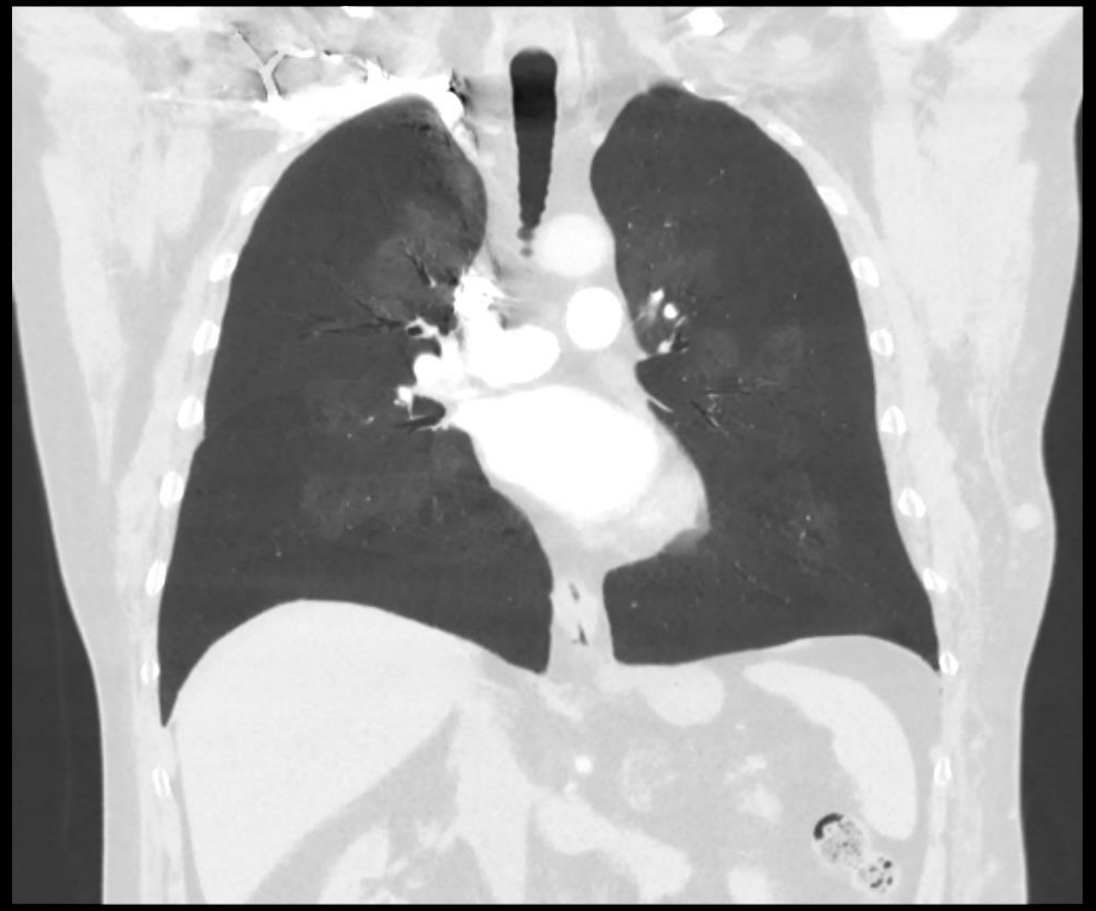


CT Pulmonary Angiogram: Webs / stenoses within multiple pulmonary artery branches (arrows)

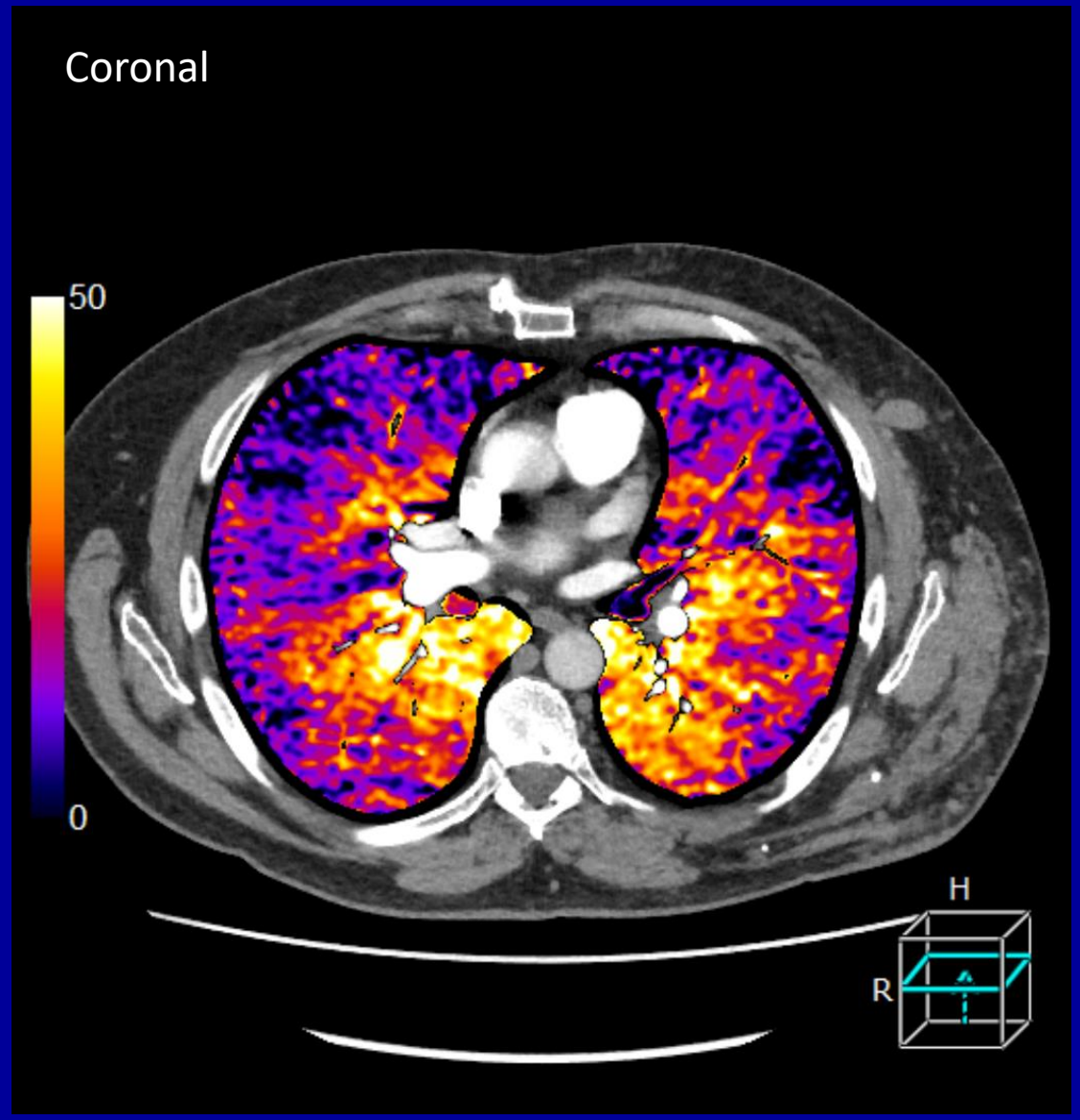
Axial

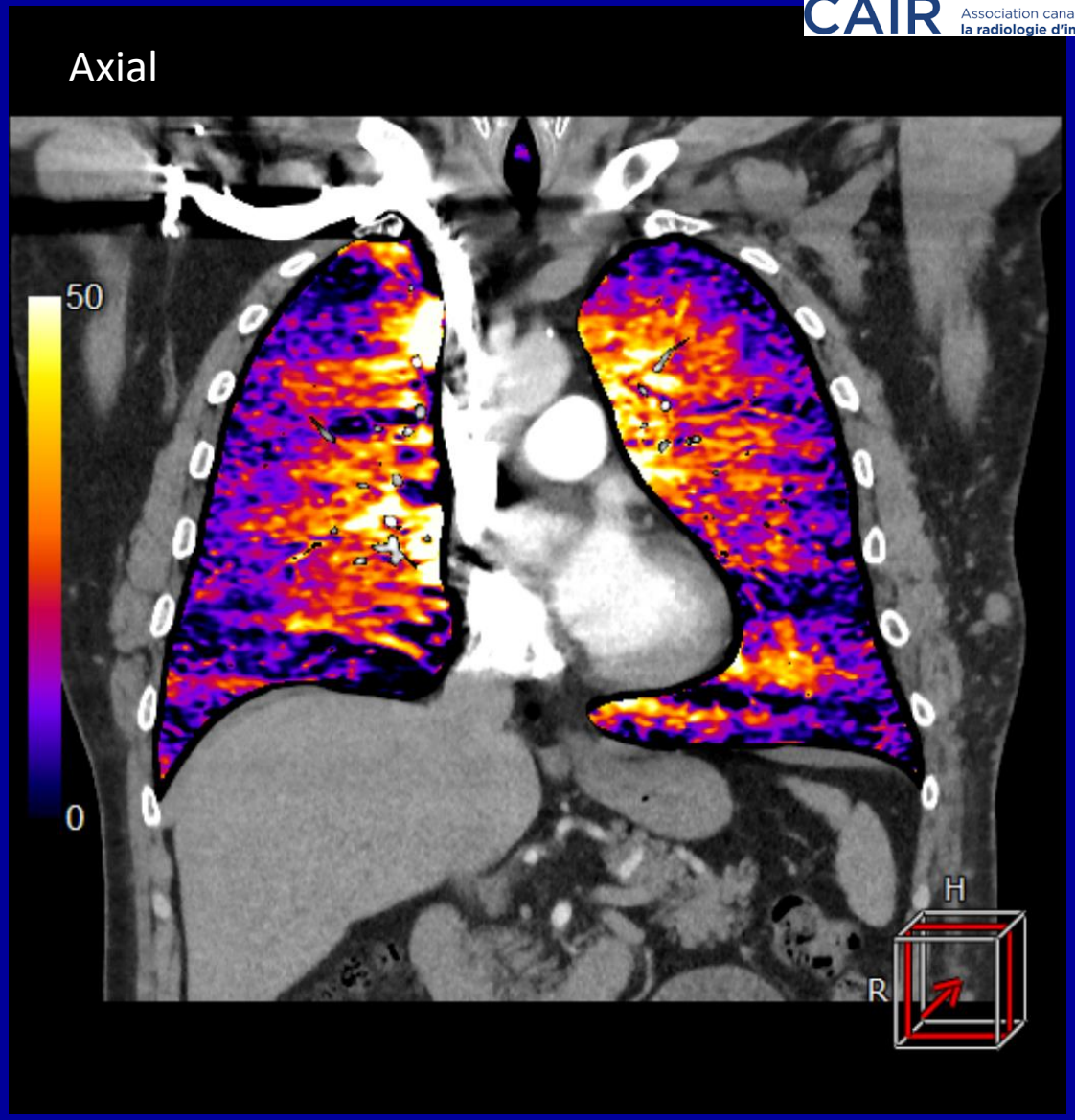


Coronal



Axial and Coronal Minimum intensity projection (MinIP) from chest CT demonstrating mosaic attenuation of lung parenchyma.





Dual Energy CT iodine mapping – reduced perfusion in the periphery of both lungs (darker regions).

Diagnosis: Chronic thromboembolic pulmonary hypertension (CTEPH)

CTEPH = Pulmonary artery stenosis or obstruction secondary to prior embolus ➡ increased pulmonary vascular resistance with progressive vessel remodeling ➡ progressive right heart failure.

CTEPH = precapillary pulmonary hypertension = mean pulmonary arterial pressure (mPAP) ≥ 25 mmHg and a normal pulmonary artery wedge pressure ≤ 15 mmHg during resting right heart cath.

CTEPH: Incidence and Prognosis

- Incidence of CTEPH after acute PE 0.56% - 3.2%

- Prognosis: CTEPH patients with mPAP > 30 mmHg have poor prognosis, if untreated, with a 5-year survival rate of 30% with mPAP > 40 mmHg and 10% with mPAP > 50 mmHg.

World J Clin Cases. 2020 Jul 6; 8(13): 2679–2702.
Respir Med. 2021 Jan 6;177:106293.

Case discussed at CTEPH rounds between Respirology, VIR and Thoracic Surgery:

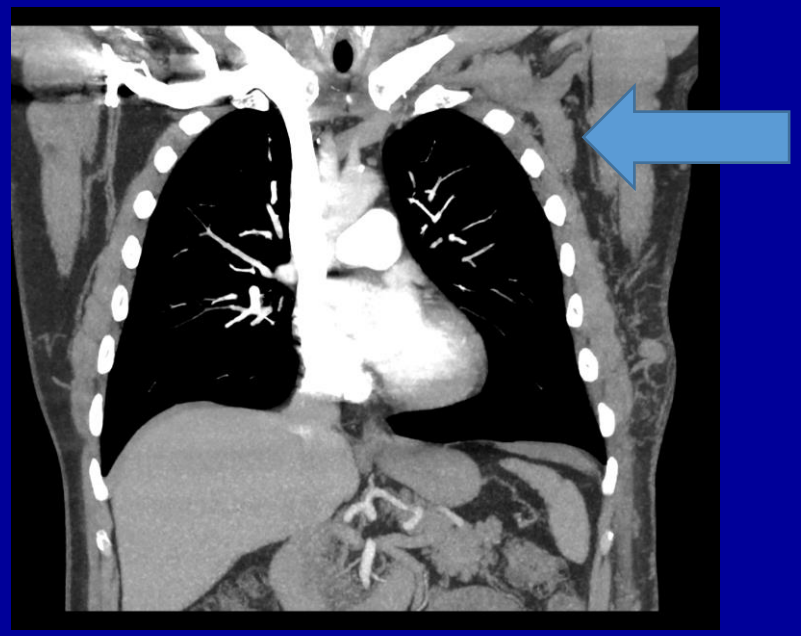
Subsegmental disease – too distal for surgical intervention

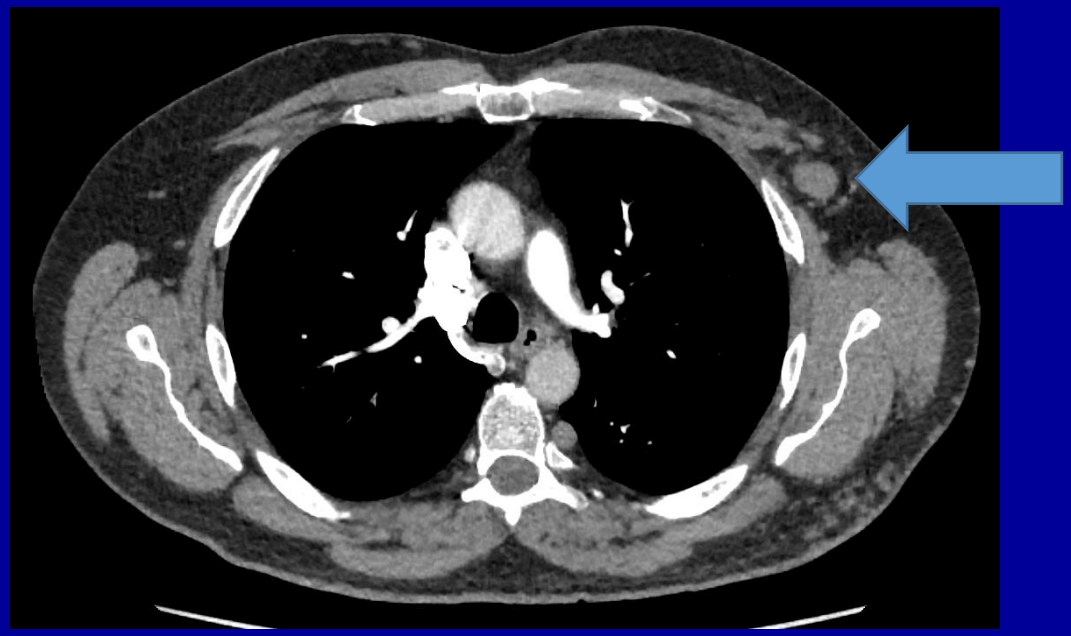
Plan for balloon pulmonary angioplasty.

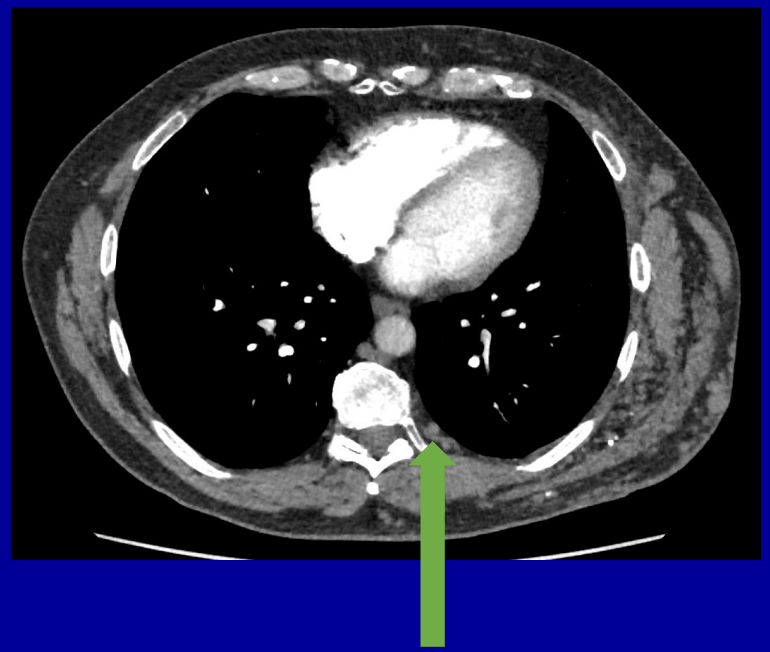
But . . . No known history of pulmonary embolism. What was the cause of this patient's CTEPH?

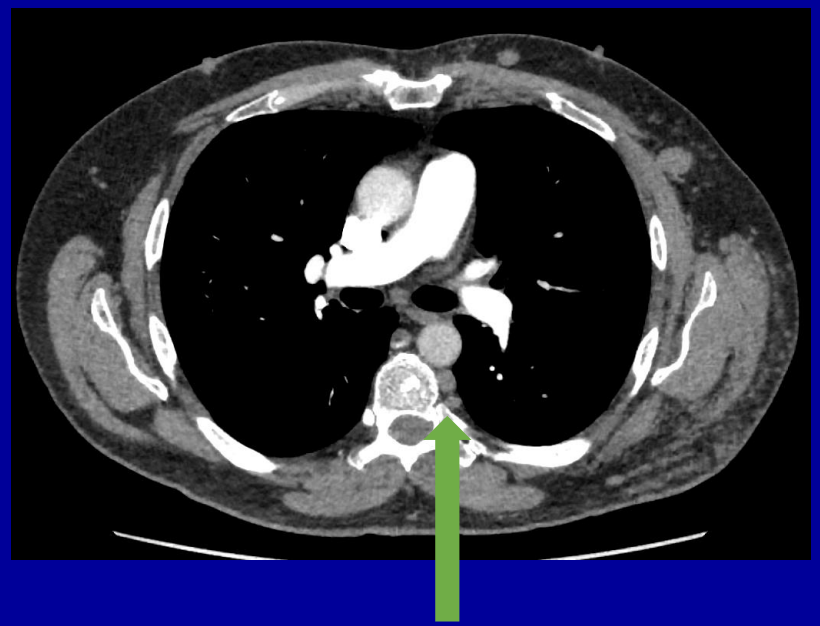
Attended VIR clinic

- Patient reported bluish tinge to the skin of his left chest for many years.
- On examination – distended vessels in left chest wall
- Ultrasound – Non occlusive thrombus in the dilated left chest vessels.









CT Pulmonary embolism study demonstrates:

- No pulmonary embolism
- Large venous collateral in the left chest wall draining to left axillary vein (blue arrows). Findings missed on initial CT Chest.
- Left extrapleural venous collaterals draining into the azygos vein (green arrows).

Diagnosis

- Left chest wall venous malformation (VM), Puig type 3 (enlarged and ectatic draining veins). Likely source of recurrent pulmonary emboli leading to CTEPH.

- Background: The blood flow through VMs is disrupted, turbulent, and (at times) stagnant. Vessel injury and inflammation activate endothelial cells, activate coagulation, consumption of clotting factors, and generate thrombin and fibrin. Recall Virchow's triad:
  - (1) Stasis of blood flow.
  - (2) Injury to vascular wall.
  - (3) Activation of the coagulation cascade.

Front Pediatr. 2017; 5: 158.
Pediatr Radiol (2003) 33(2):99–103

Evidence: Link between VM and CTEPH

Vascular malformations as underlying cause of chronic thromboembolism and pulmonary hypertension

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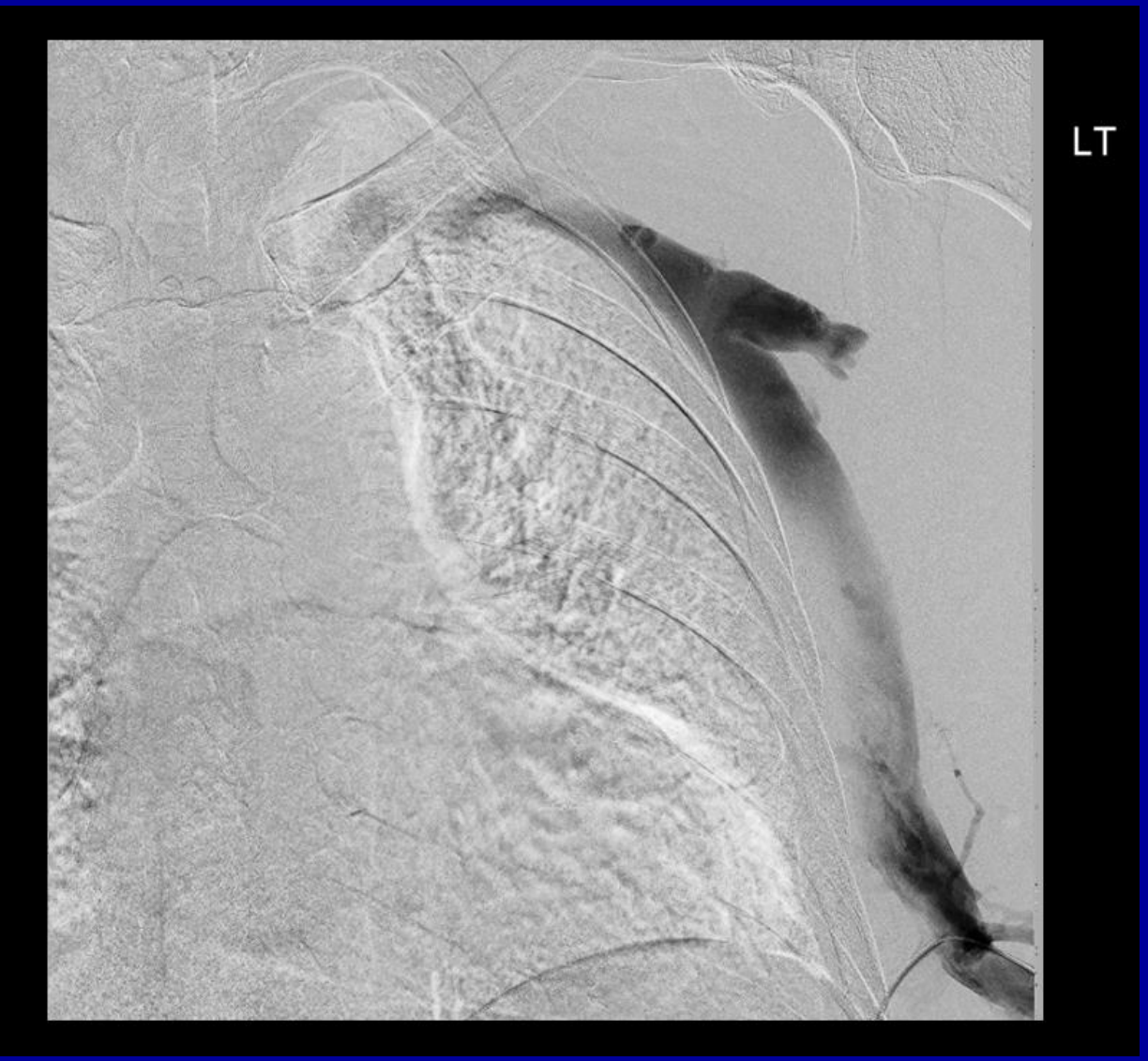
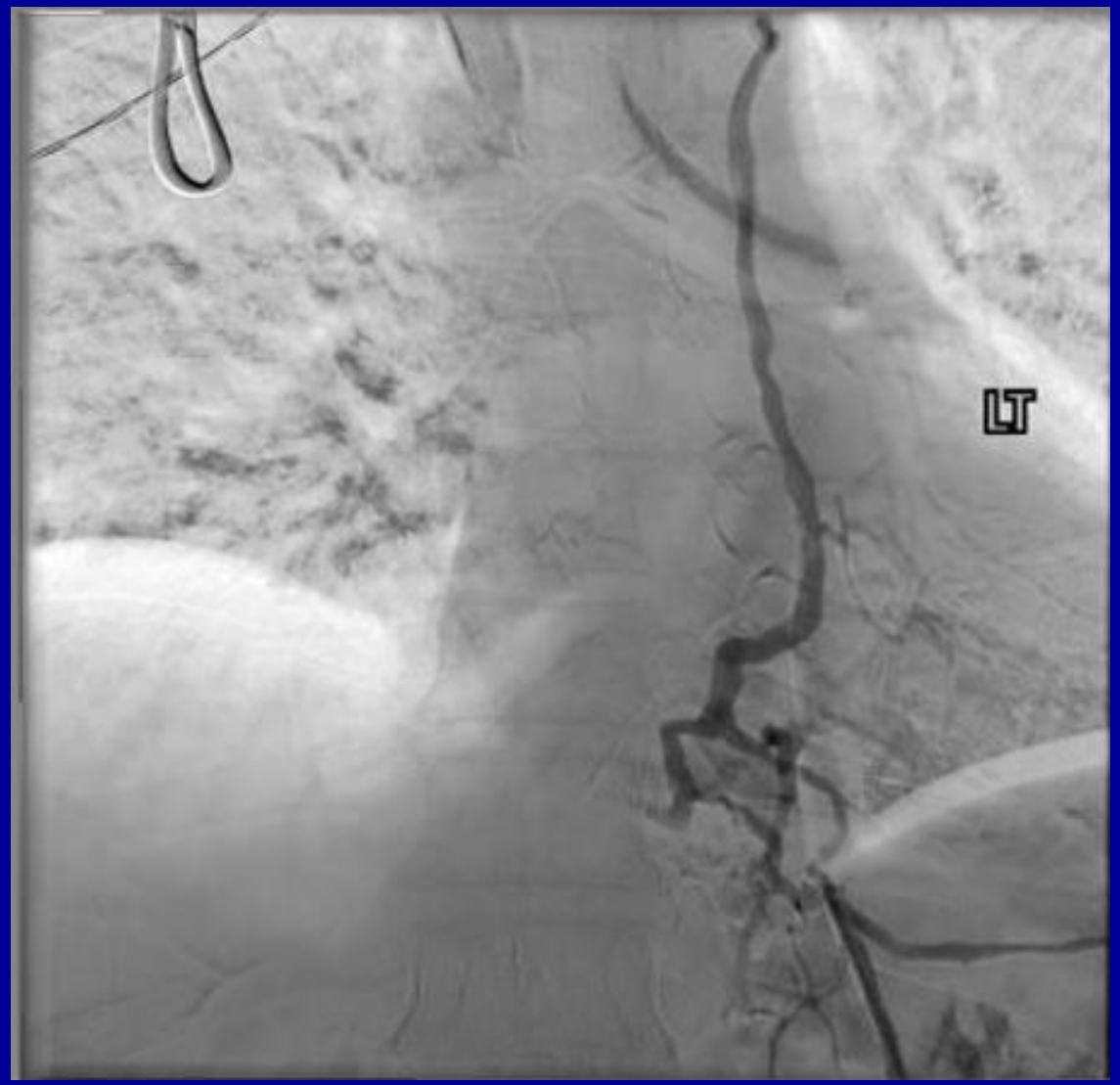
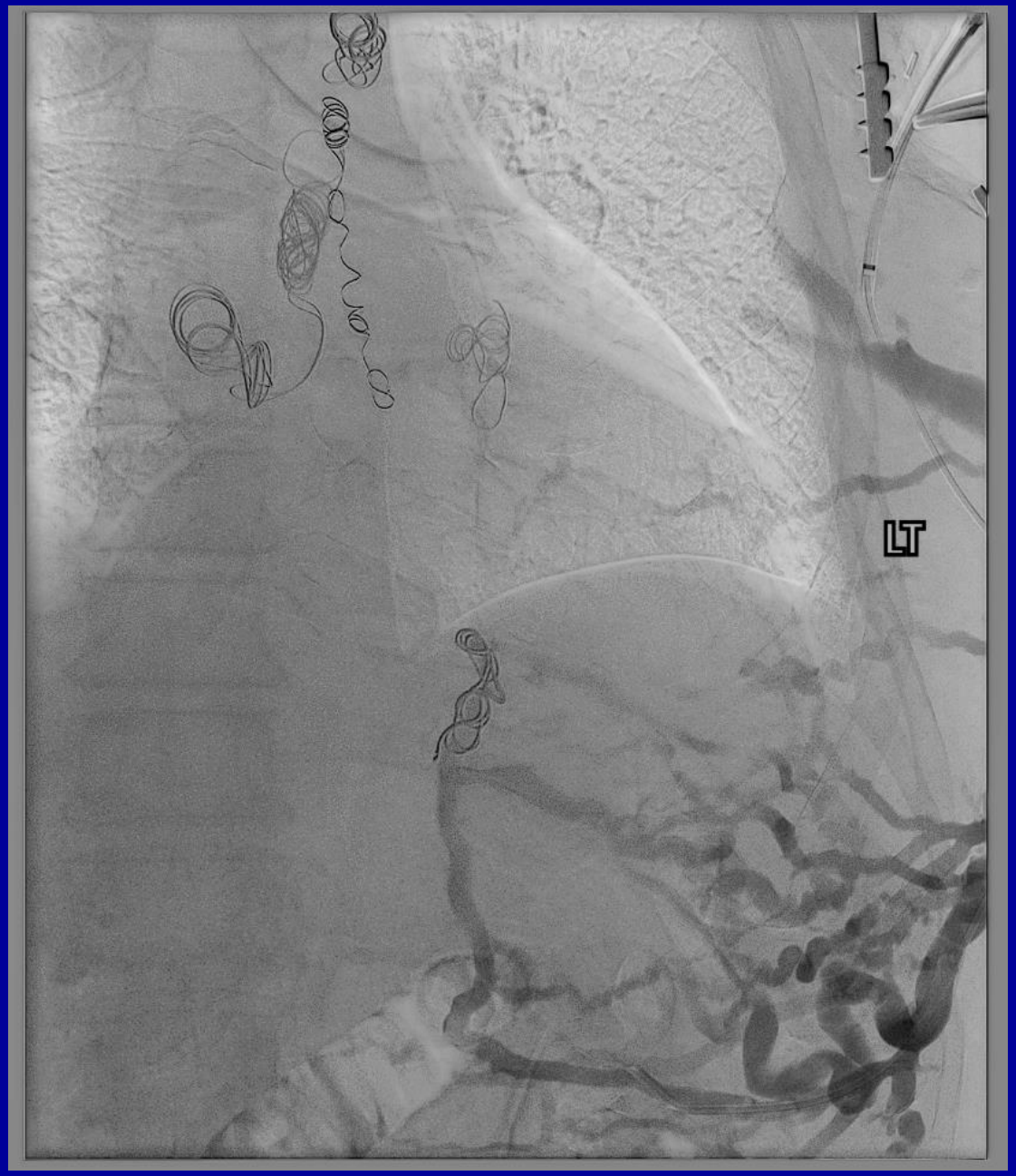
Summary We report four patients with chronic thromboembolic pulmonary hypertension (CTEPH) presumably due to recurrent pulmonary embolism from low-flow vascular malformations, and give a review of the literature.

Venous malformations, such as those observed in Klippel- Trenaunay syndrome (KTS) can be associated with hypercoagulability, thrombosis and recurrent pulmonary embolism and ultimately CTEPH. Since many physicians appear unfamiliar with these potential complications, patients may experience a delayed diagnosis of this progressive and potentially life-threatening CTEPH.



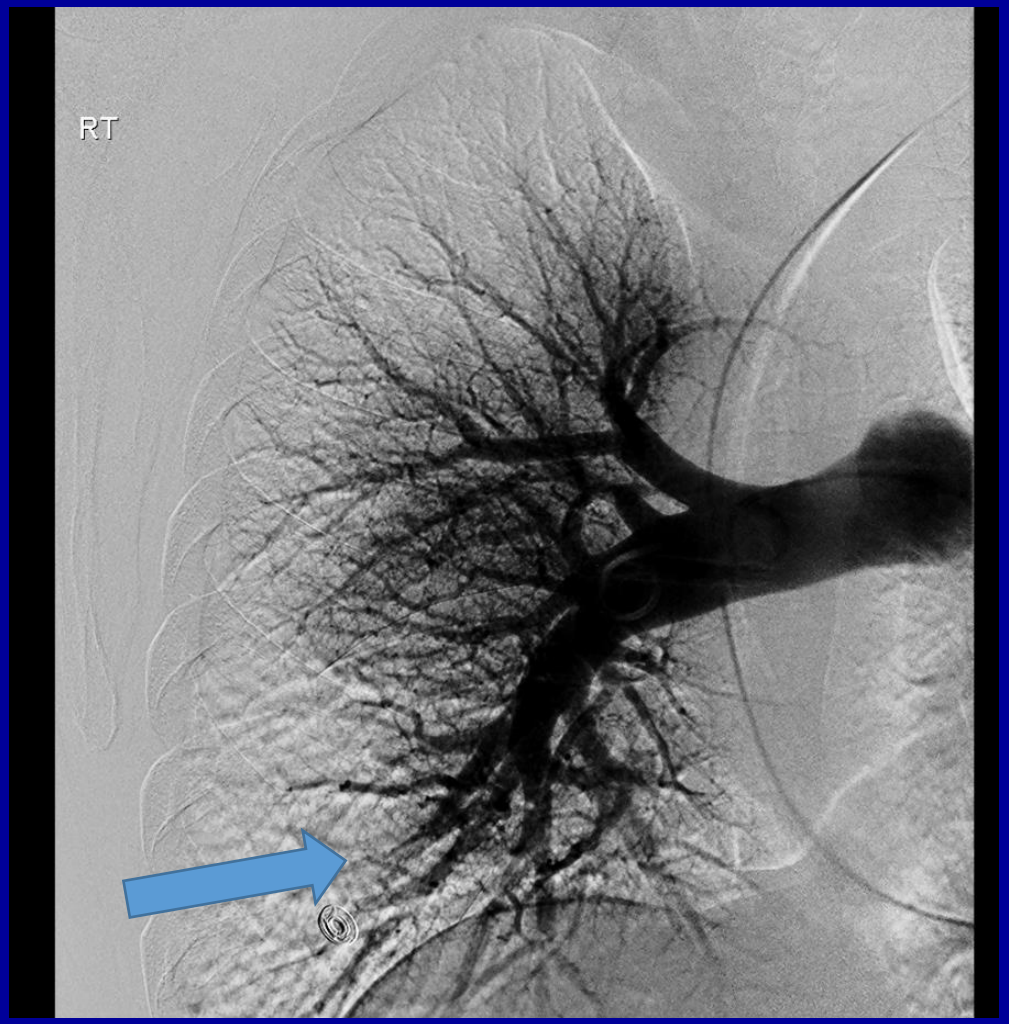


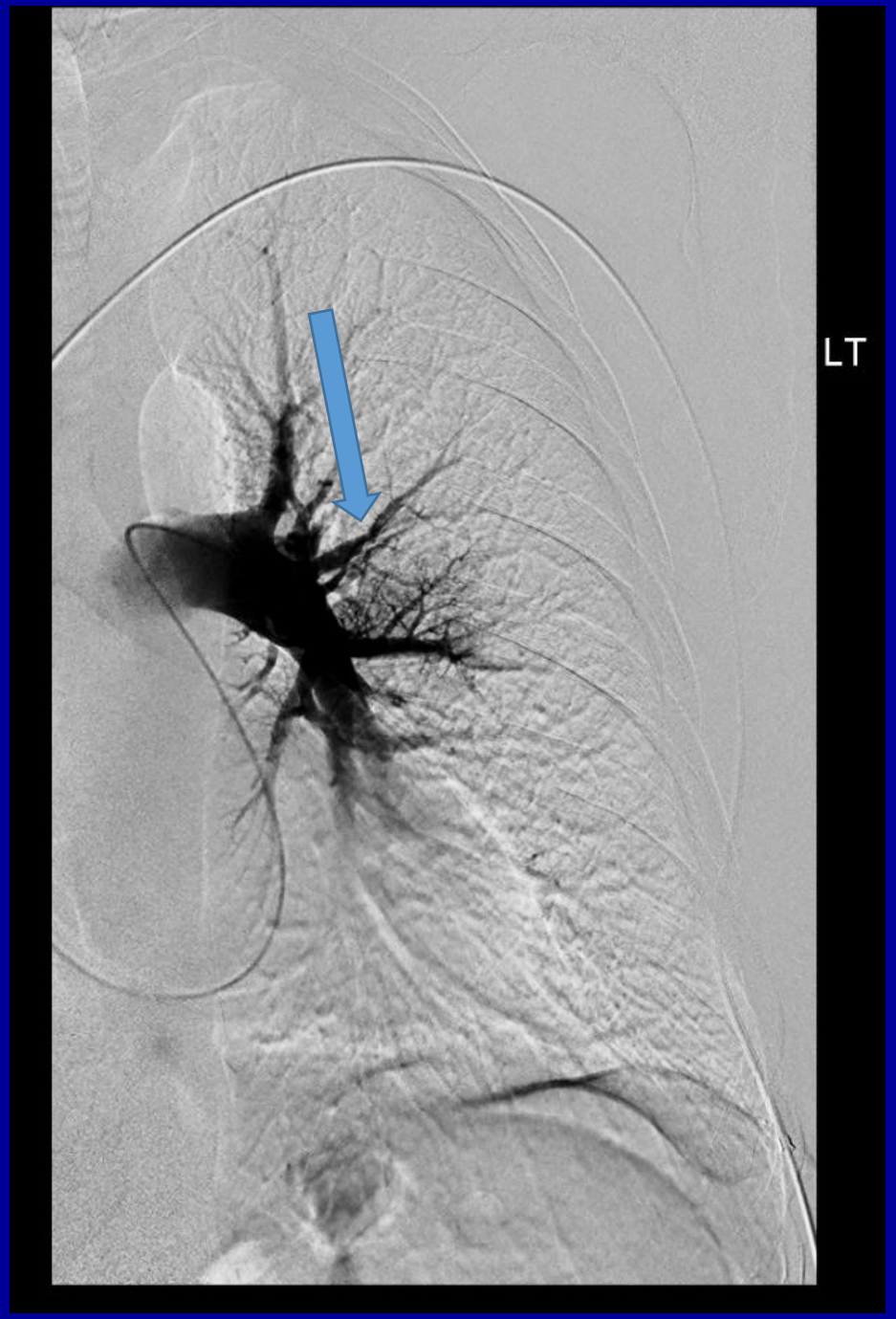
Catheter venography: Complex left chest wall venous malformation draining into the left axillary vein and the azygous system



COMBINED PROCEDURE WITH VASCULAR SURGERY:

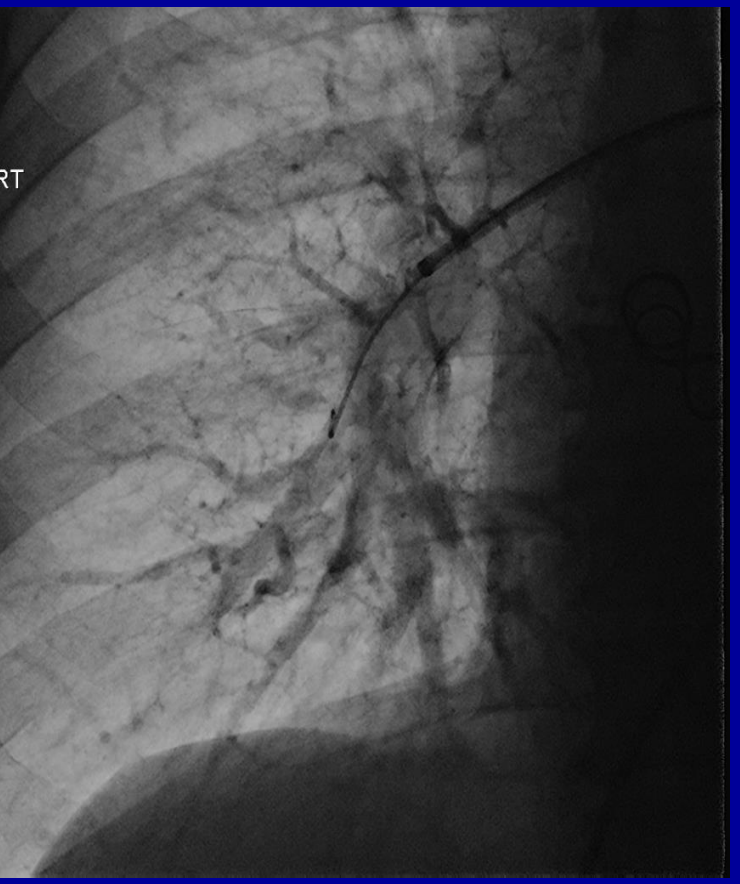
- Surgical ligation of the venous malformation outflow vein which drains to the left subclavian vein.
- Coil embolization of left internal mammary vein.
- Coil embolization of venous malformation outflow vein draining to azygous system.
- Sclerotherapy of left chest wall venous malformation using STS foam.



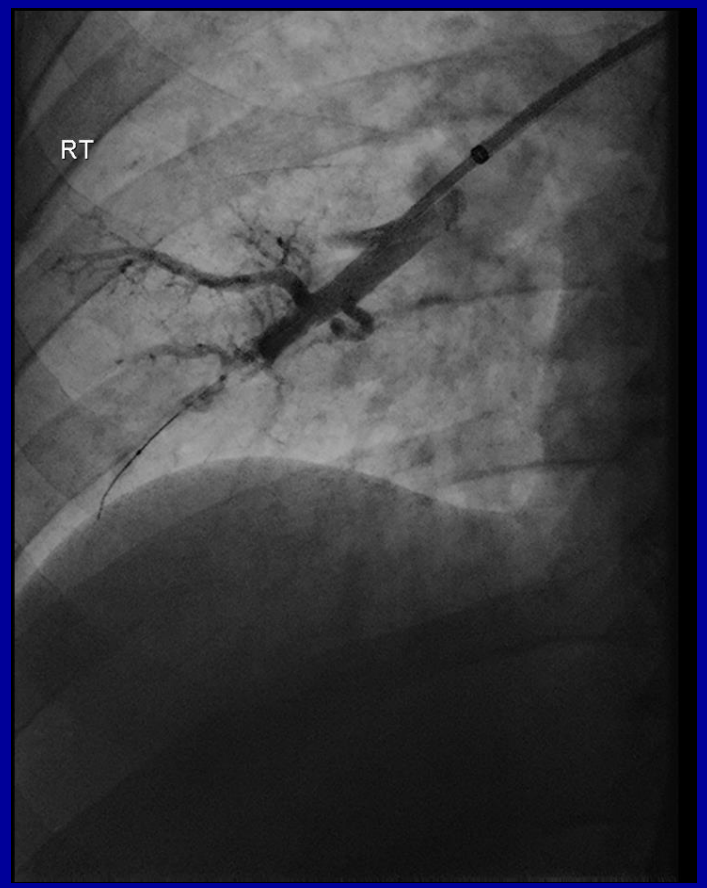


Bilateral pulmonary angiogram via femoral access: Multiple focal arterial webs / stenotic segments. Examples right lower lobe and left upper lobe (arrows).

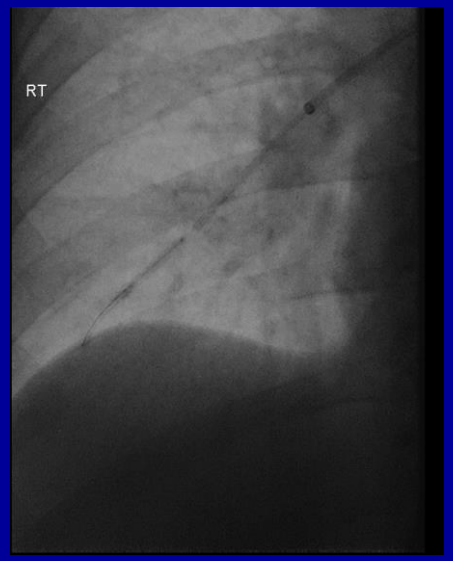
Balloon pulmonary angioplasty



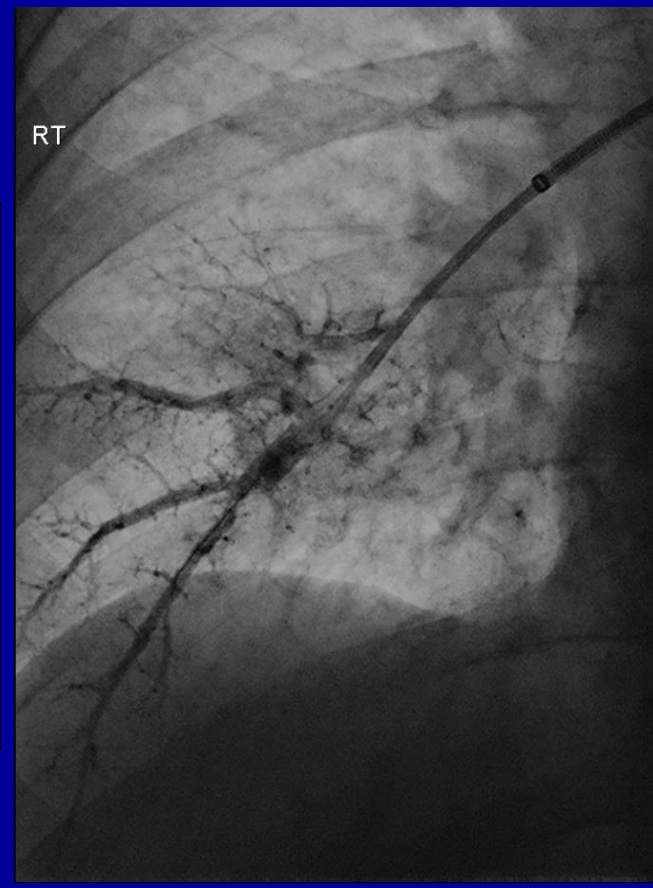
Attenuation of right lower lobe basal segment artery.



Stenosis/web crossed with 0.014” wire



2mm monorail balloon angioplasty



Post angioplasty.

Treatment course

- Anticoagulated. Underwent four BPA sessions
- Pulmonary pressure before treatment 3: 65/34 (mean 41) from baseline 81/35 (50).
- Consultation 2 months after 4th treatment:
  - Saturations improved; 94-95% range on room air. Now able to get up to 97-98% with ease; previously needed O2 to get this level.
  - Very active - skiing, snowmobiling and walking.
  - Able to undergo strenuous activity such as exerting himself for 3 hours to get a 4 wheeler off the ice.
  - Quality of life has improved, no longer concerns about exerting himself.

  Plan: Monitor symptoms and consider 5TH BPA session if deterioration.

Discussion:
BPA for CTEPH

Management of CTEPH

- Pulmonary artery endarterectomy (PEA) is standard of care for operable CTEPH.

- 37% of patients are deemed unsuitable for PEA surgery due to comorbidities and other factors, and nearly half of the operated patients have residual or recurrent pulmonary hypertension.

- Balloon pulmonary angioplasty is an approach for patients who are not good candidates for surgery or have residual pulmonary hypertension after PEA. Combined with medications.

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Evidence for BPA is mounting:

- Khan *et al* meta-analysis 17 studies (2019)
  - - Mean PA pressure decrease 14.2 mmHg.
  - - Increase in 6 minute walk distance 67.3m.
  - - 1.9% mortality within one month.

- Darocha *et al* Prospective multi-center registry (2022)
  - 156 patients.
  - Mean PA pressure decreased 45.1 to 30.2 mmHg.
  - 6 minute walk test increased from 341 to 423 m.
  - Pulmonary artery injury 6.4%.
  - 1.7% mortality within one month.

Int J Cardiol. 2019 Sep 15;291:134-139.
EuroIntervention. 2022 Jan 28;17(13):1104-1111.
Pulm Circ. 2021 Apr-Jun; 11(2):

BPA

- Major complications : Vascular injury, which can be accompanied by hemoptysis and/or hypoxemia. Mortality 1.8 to 3%.
- Most patients will require more than one session, as attempting to treat too many lesions in one session can increase the risk of complications, esp if PA pressure > 40 mmHg.
- Should be performed only at experienced CTEPH centers for patients who are ineligible for PEA

Respir Med. 2021 Jan 6;177:106293.
Eur Respir J, 53 (2019), p. 1802095

International guidelines:

BPA is recommended by **ESC/ERS**, alone or in combination with medications, as treatment for patients with CTEPH who are technically inoperable or carry an unfavorable risk/benefit ratio for PEA (recommendation class IIb, level of evidence C)

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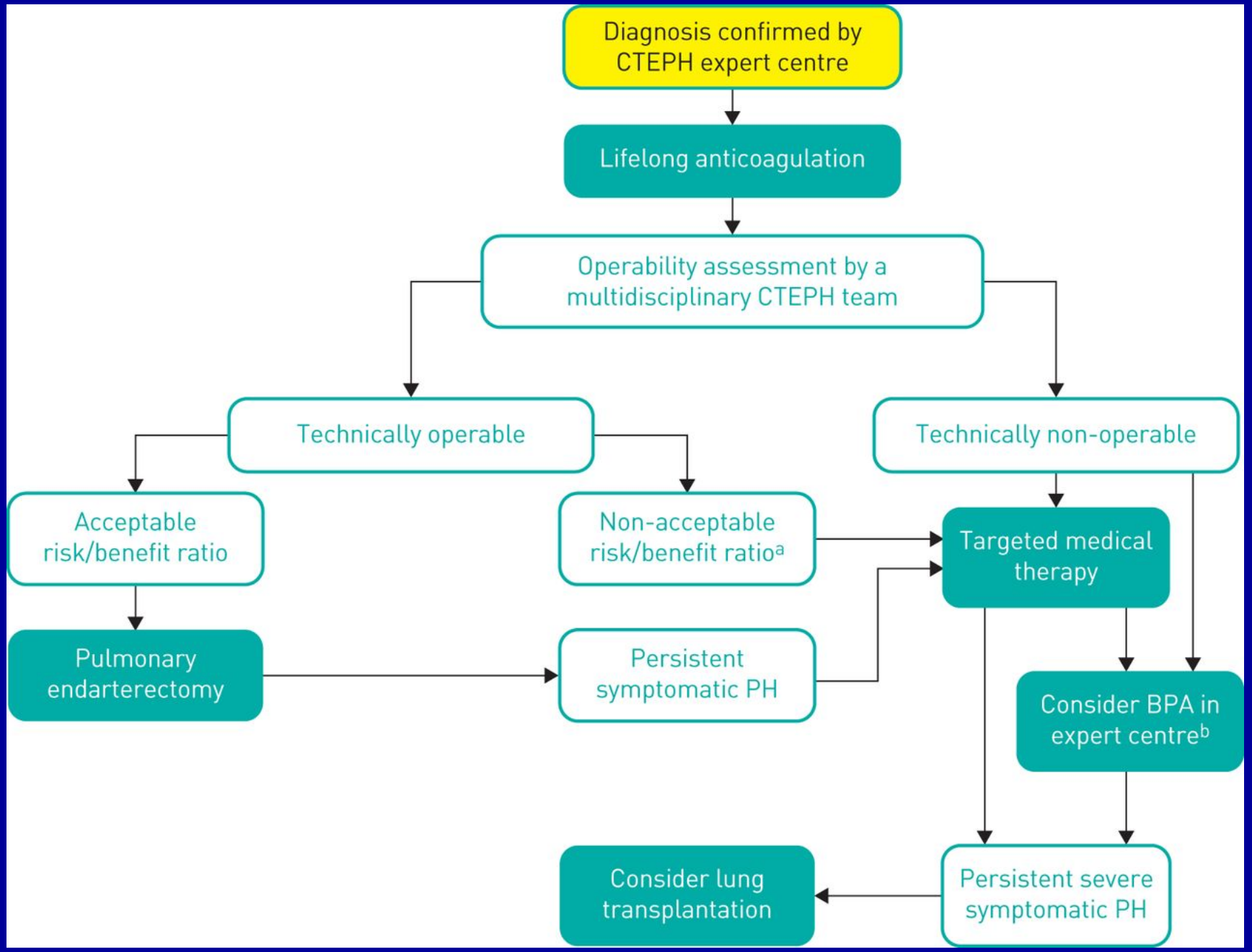
Eur. Respir. J., 46 (4) (2015)



Eur. Respir. J., 46 (4) (2015)

Summary

- Clinical Interventional Radiology service was integral to the diagnosis, treatment and follow up of this patient.
- CTEPH is a diagnosis which is often missed. Considered it in your vascular malformation practice and your diagnostic reporting.
- Evidence is mounting for benefit of BPA, with mean improvement in PA pressure of 15mm Hg.
- The future is likely combination treatment of Surgery / BPA with medications.