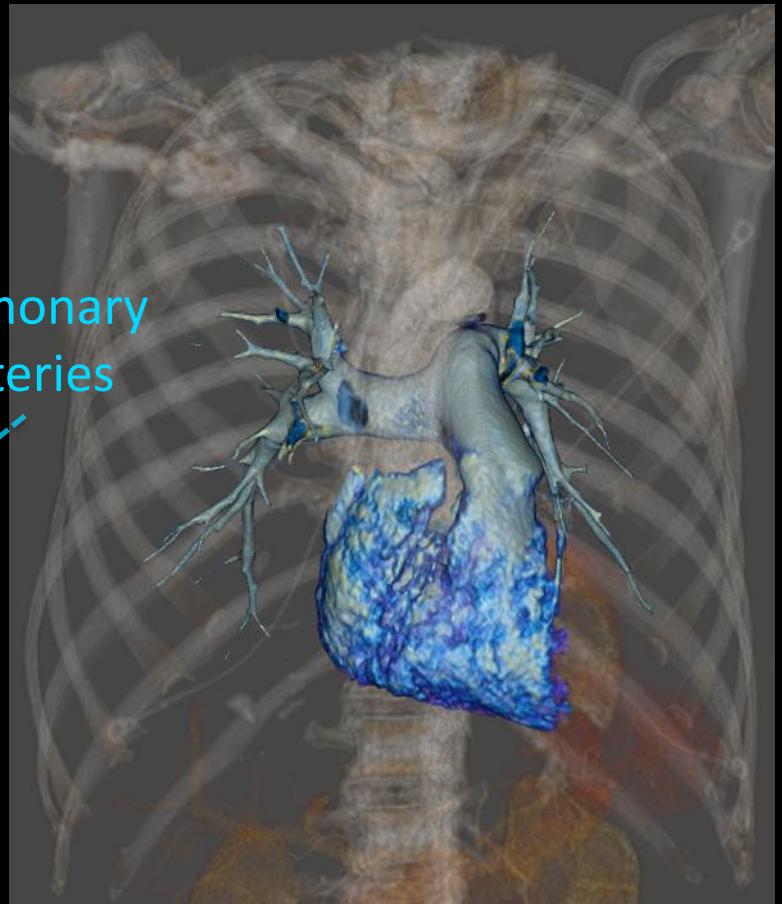
Pulmonary Vascular Disease

We Have No Relevant Disclosures

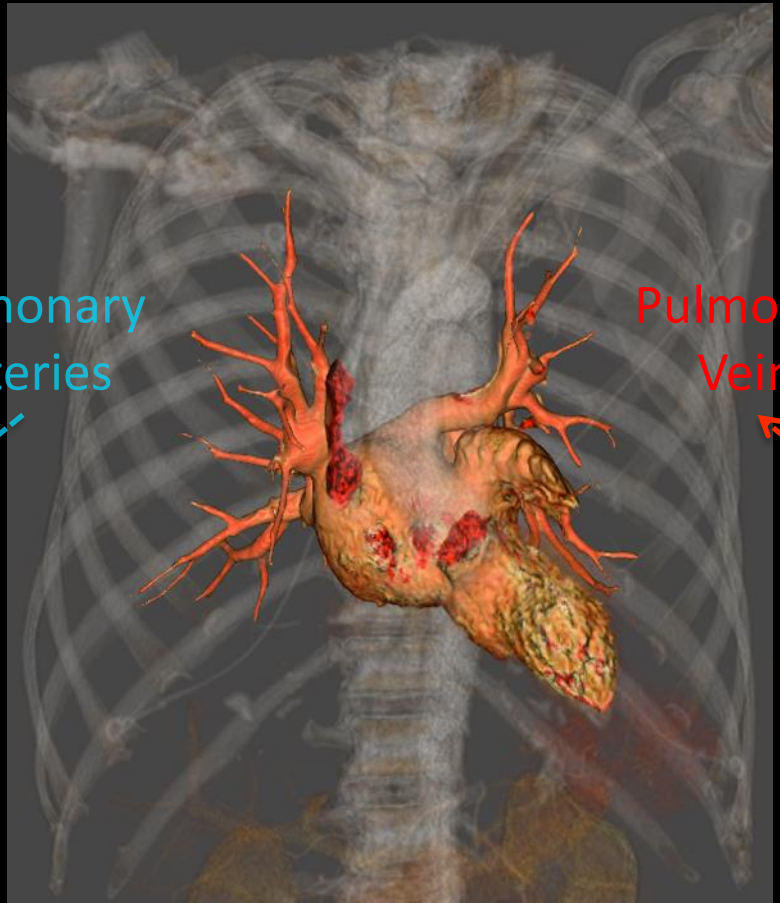
References

- Jaramillo, Felipe Aluja, Fernando R Gutierrez, Federico G Díaz Telli, SebastianYevenes Aravena, Cylen Javidan-Nejad, and Sanjeev Bhalla. “Approach to Pulmonary Hyper Tension: From CT to Clinical Diagnosis” 38, no. 2 (n.d.).
- Simonneau, Gérald. “Haemodynamic Definitions and Updated Clinical Classification of Pulmonary Hypertension,” n.d.

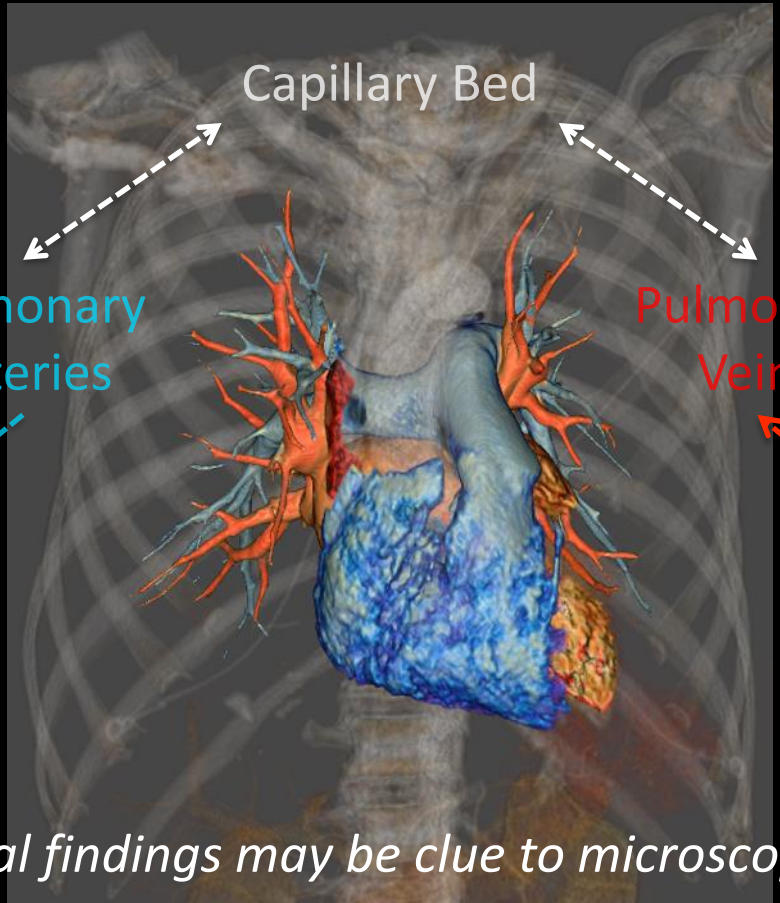
Pulmonary Vascular Disease is a Spectrum



Pulmonary Vascular Disease is a Spectrum



Pulmonary Vascular Disease is a Spectrum



Parenchymal findings may be clue to microscopic disease!

Pulmonary Arteries

- Dilated
  - Pulmonary Hypertension
  - Aneurysm/Pseudoaneurysm
- Narrowed/Occluded
  - Acquired
  - Congenital

Pulmonary Hypertension

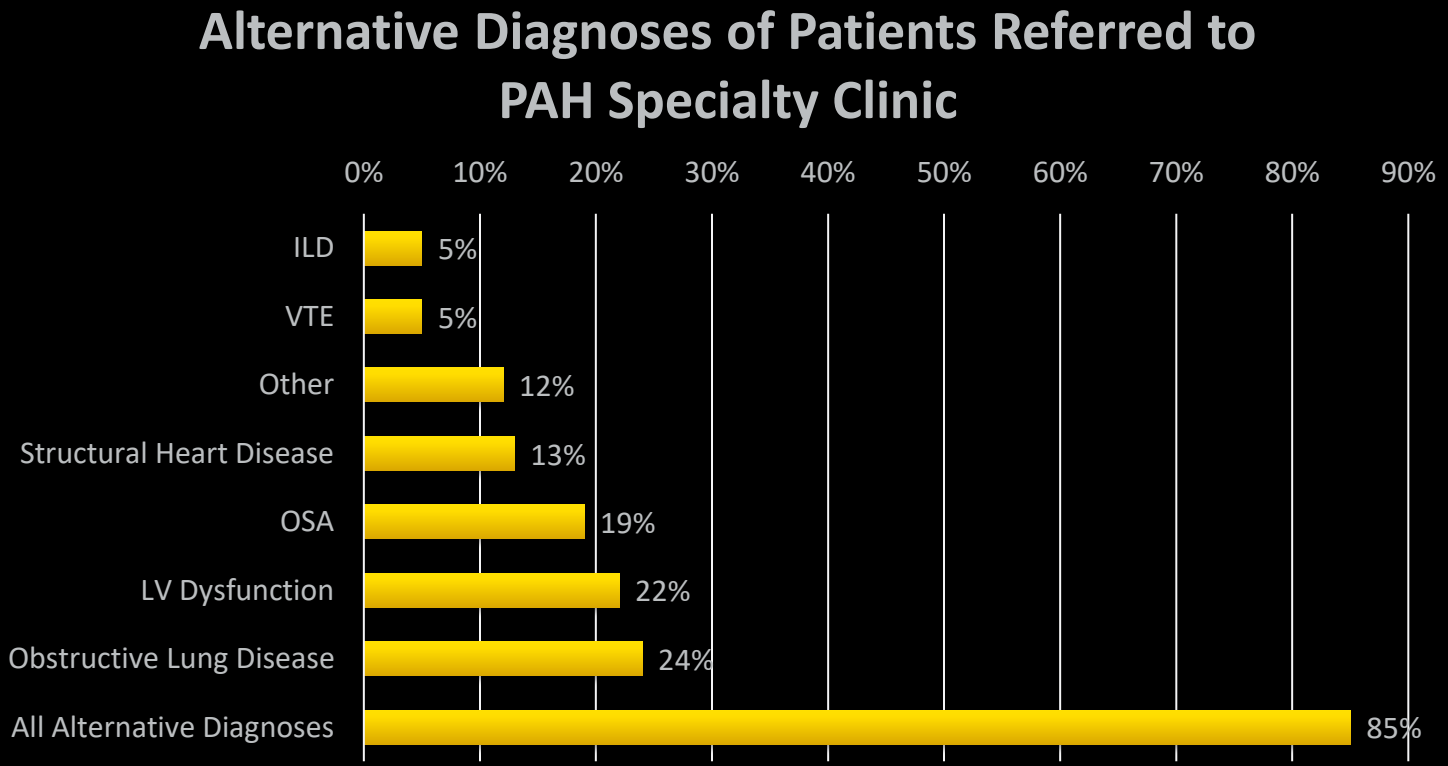
Five Groups*:

1. Pulmonary Arterial Hypertension (PAH)
2. Left heart disease
3. Lung disease/hypoxemia
4. Chronic Thromboembolic Pulmonary Hypertension
5. Unclear/multifactorial

Mean Pulmonary Artery Pressure >20 mmHg

*6th World Symposium on Pulmonary Hypertension 2018

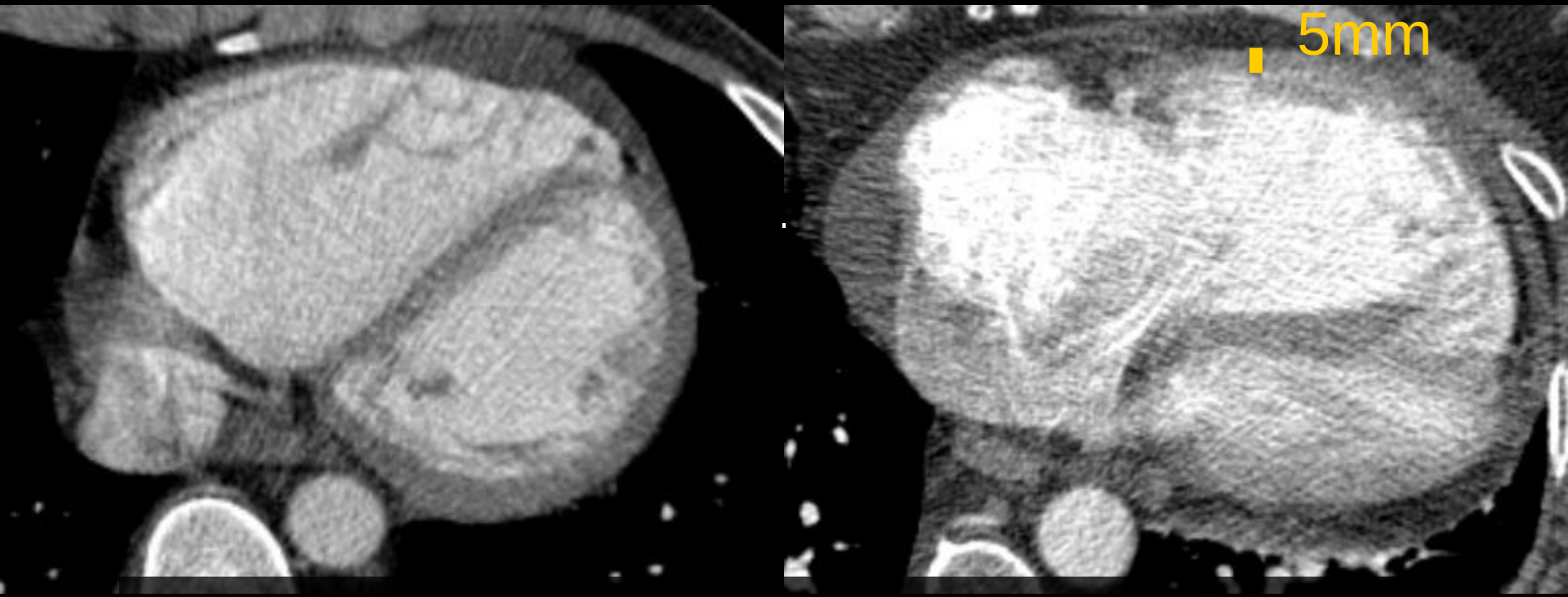
How does the Radiologist help?



N= 268

Moghbelli et al. Am J Respir Crit Care Med 2008; 177:A923.

RV Dilation vs Hypertrophy

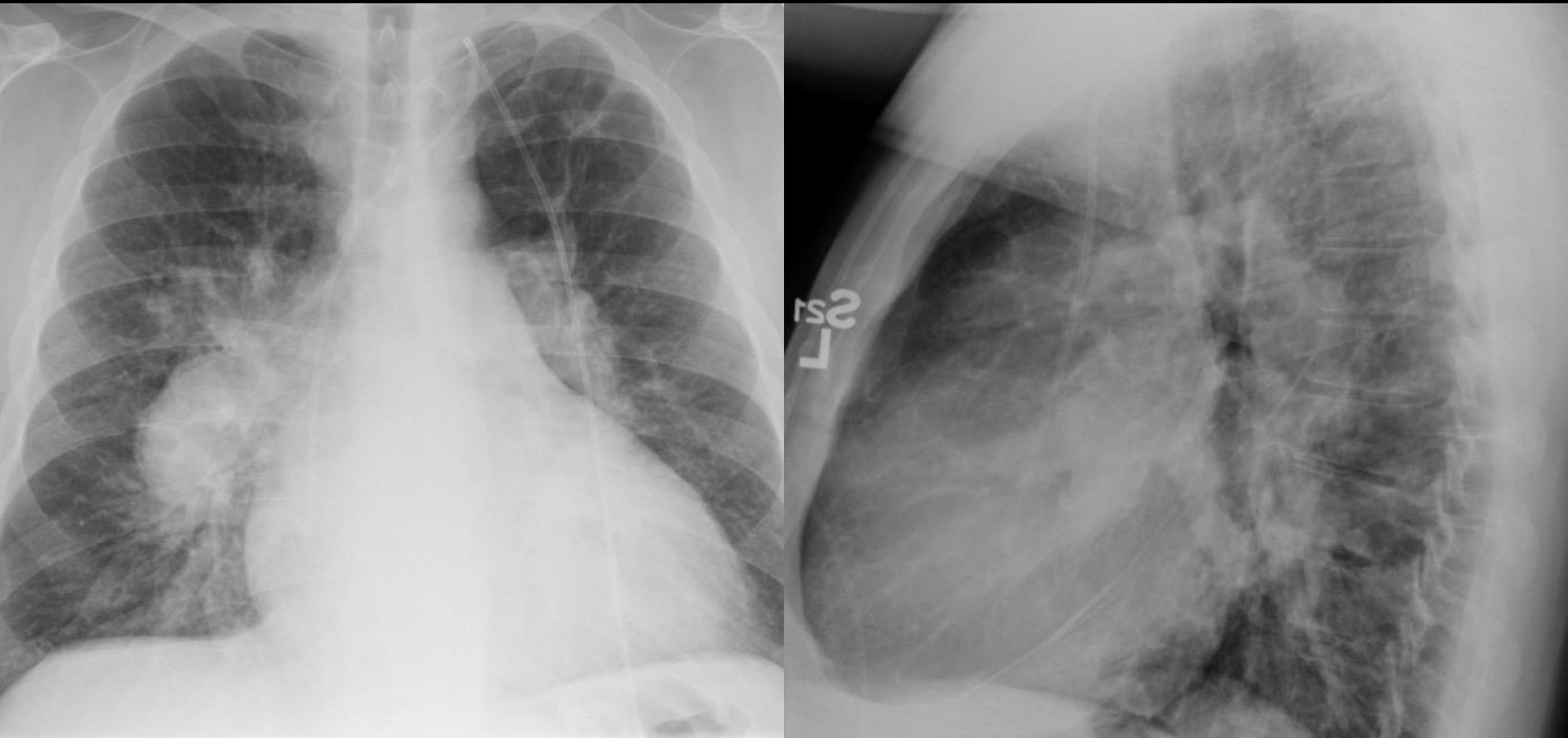


The RV is very sensitive to changes in pressure

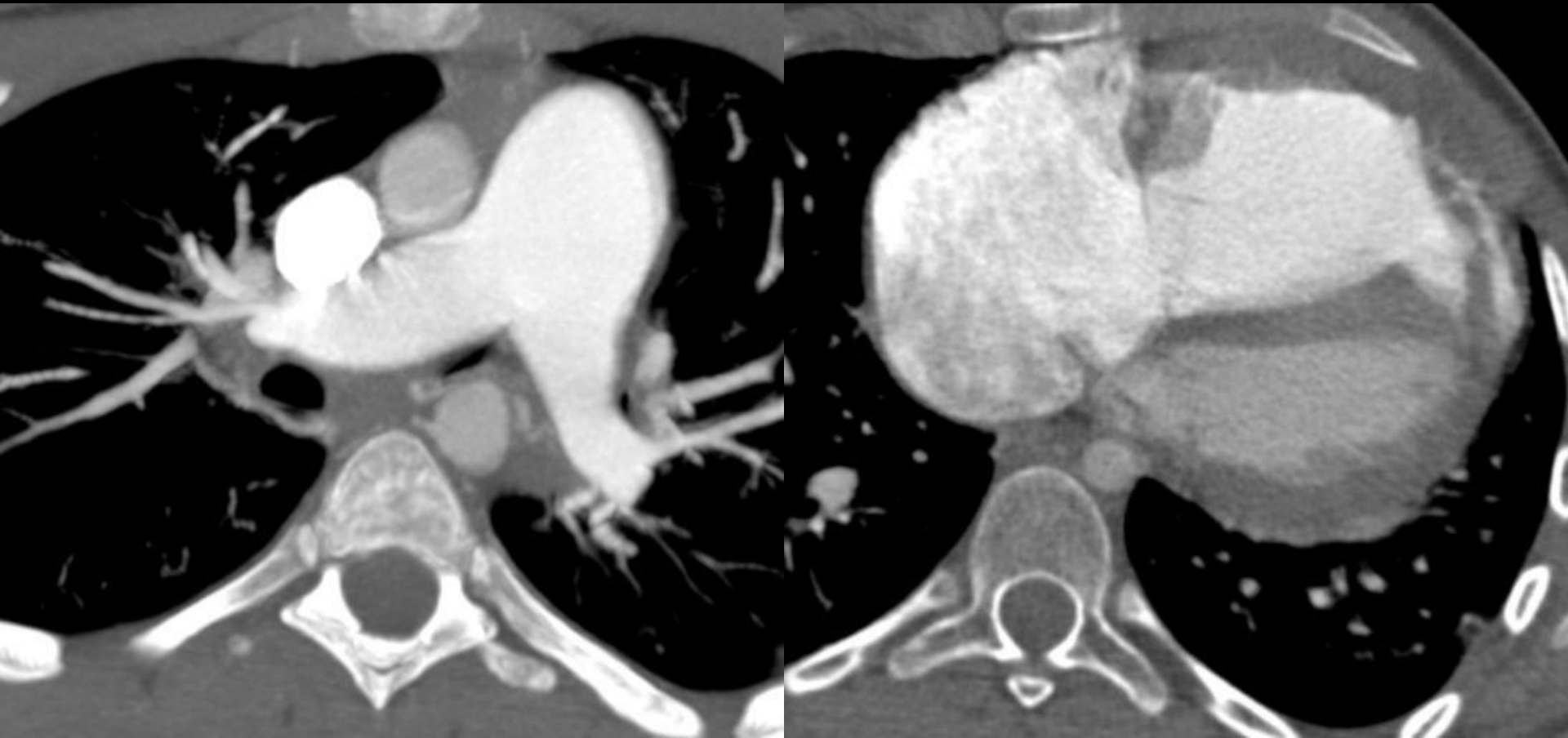
Group 1 - PAH

- Precapillary disease
- Causes:
  - Idiopathic
  - Heritable
  - Drugs
  - CTD
  - Congenital heart disease (shunt)
  - *PAH with overt features of venous/capillaries (PVOD/PCH) involvement*

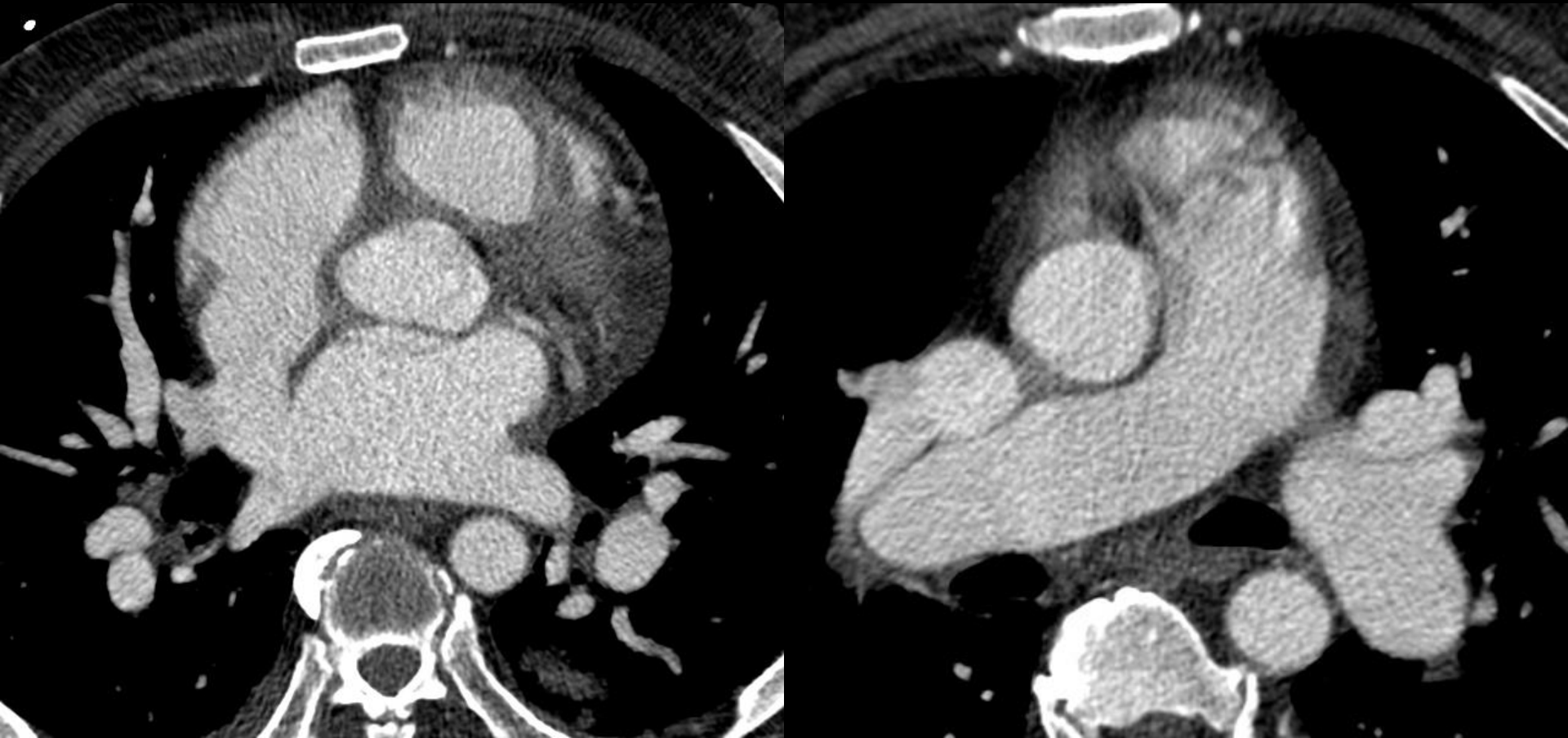
Idiopathic PAH



Idiopathic PAH



PAH from Congenital Heart Disease



Parenchymal Findings in PAH

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- Mosaic attenuation
- Groundglass centrilobular nodules
- Corkscrew vessels

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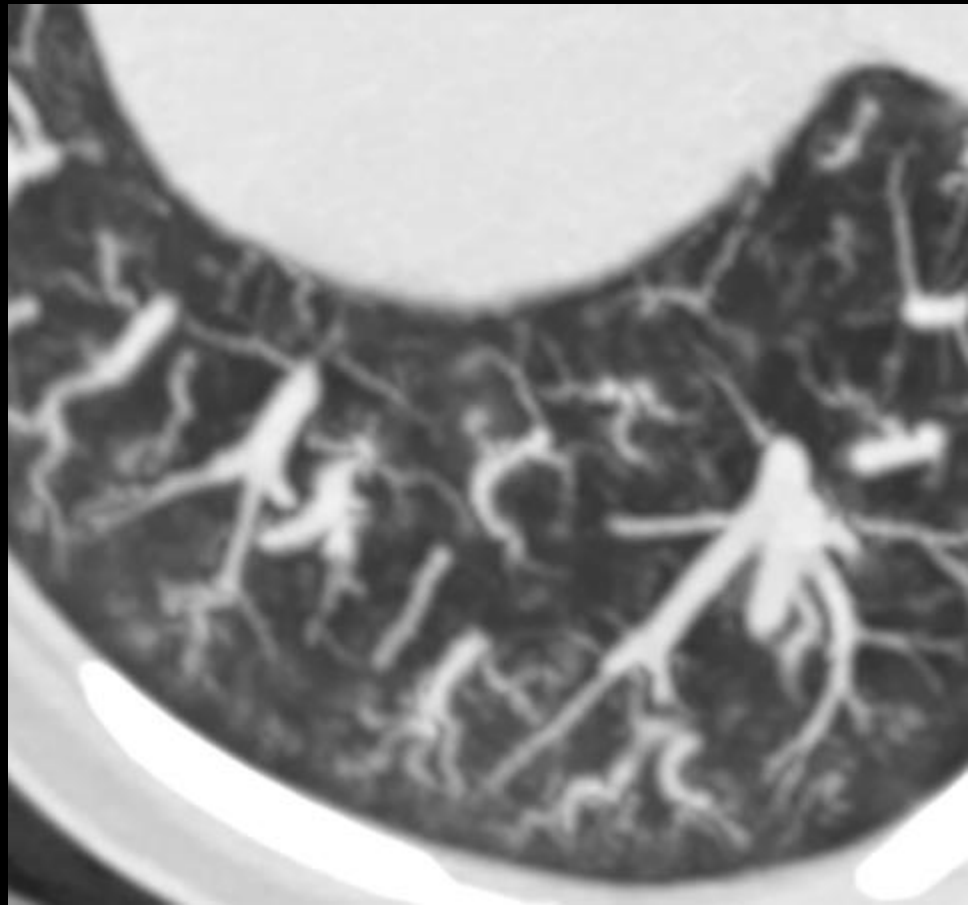
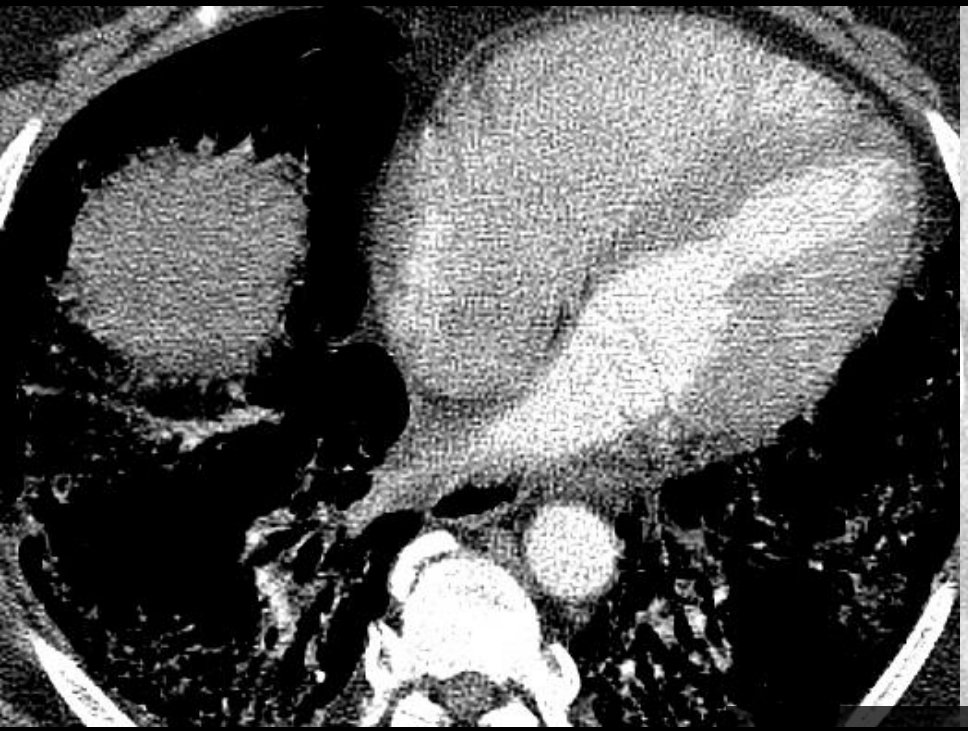
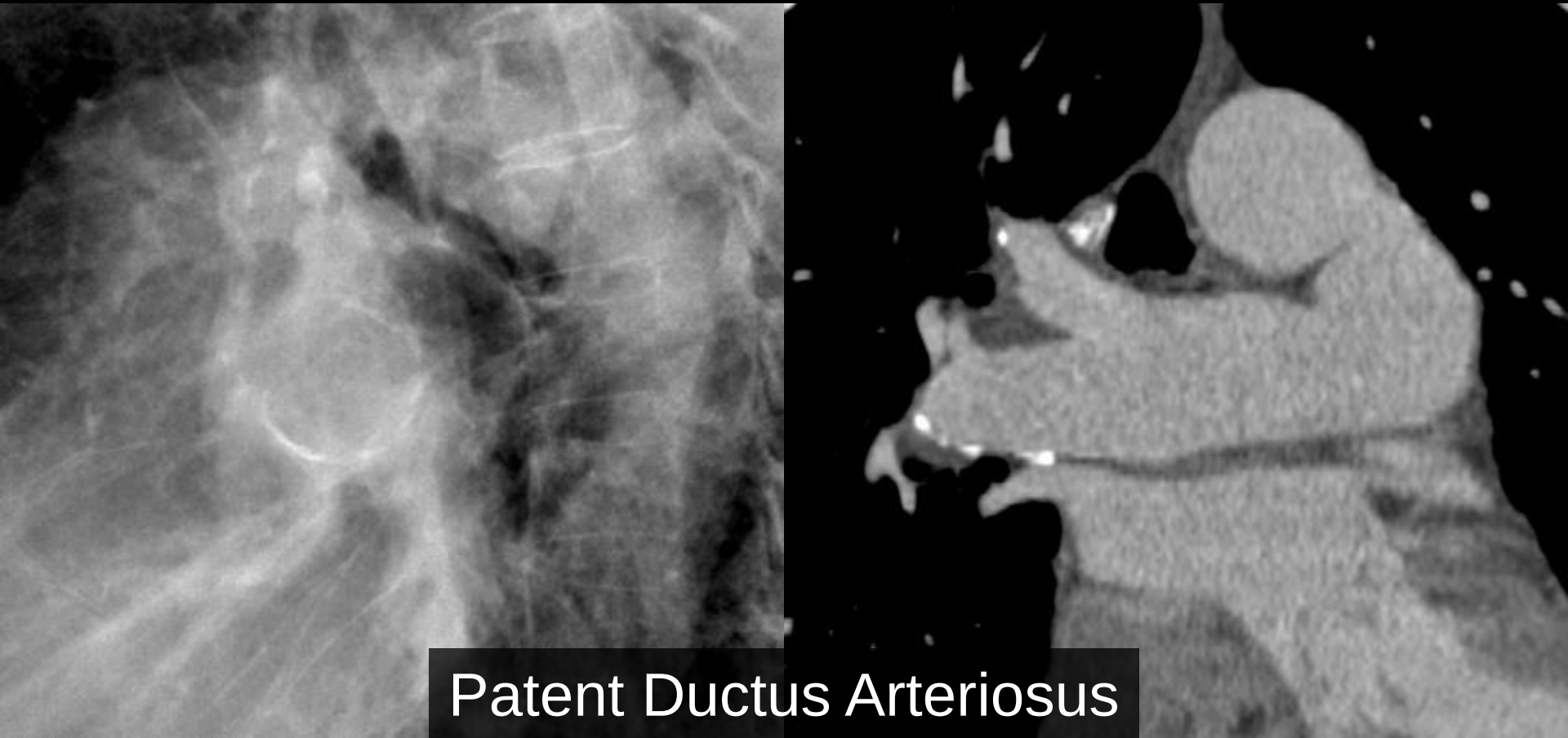


PAH in CTD



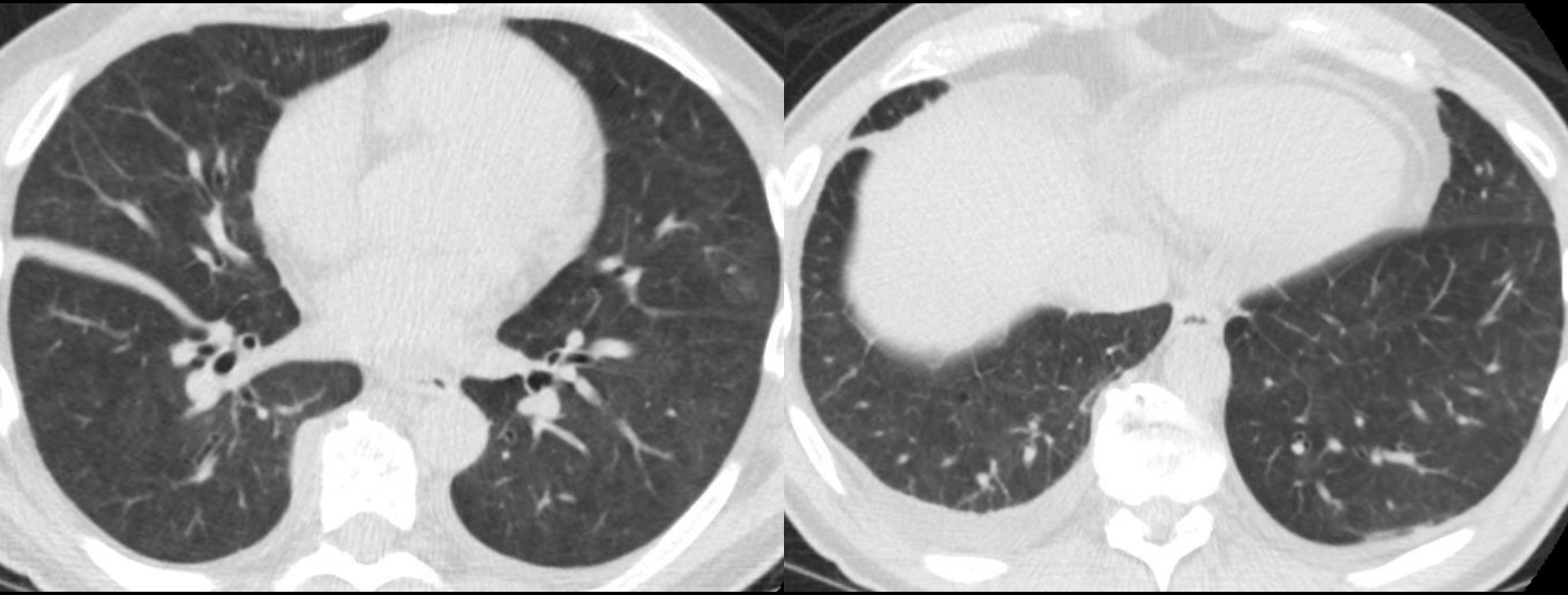
Scelroderma

Atherosclerosis in longstanding PAH



Patent Ductus Arteriosus

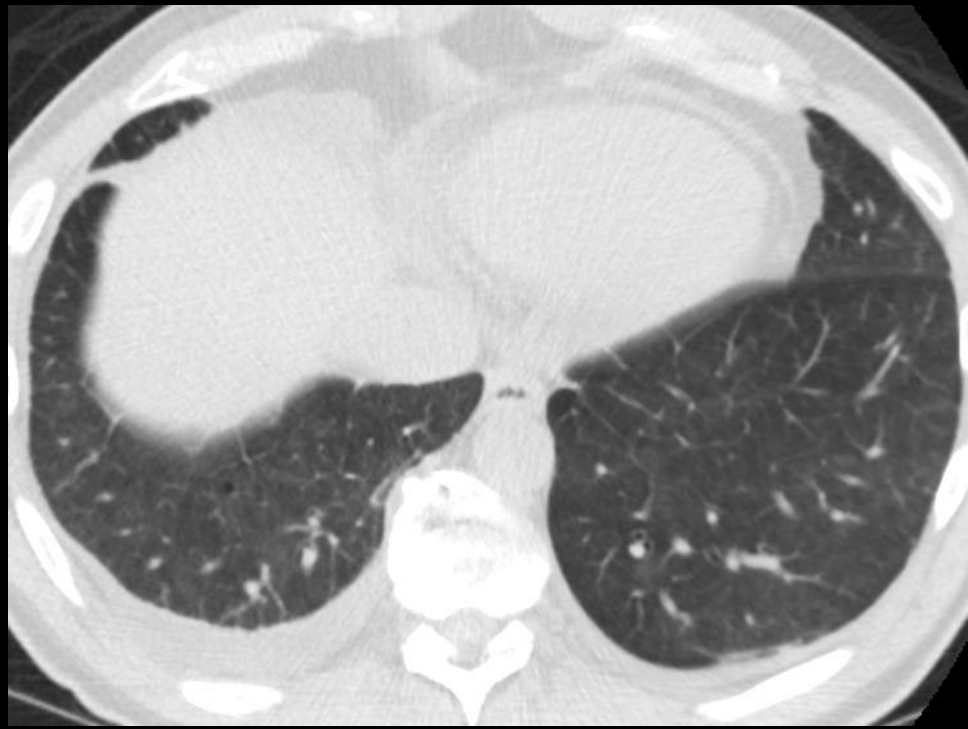
PVOD/PCH



PAH + Pulmonary Edema + Normal Left Heart

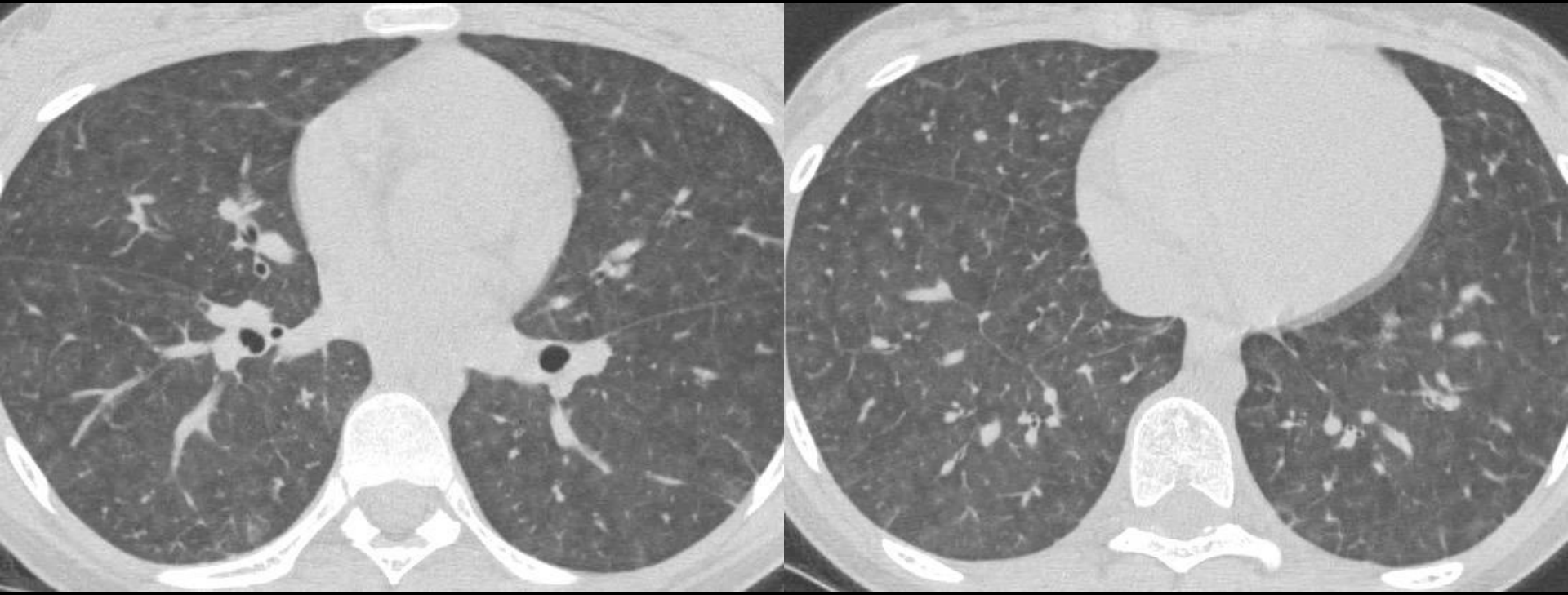
PVOD/PCH

- Diagnostic dilemma (normal PCWP)
- 5-10% of patients with idiopathic PAH
- Incidence ~2/million per year
- Associations
  - Idiopathic
  - HIV, BMT, CTD, sarcoid, PLCH

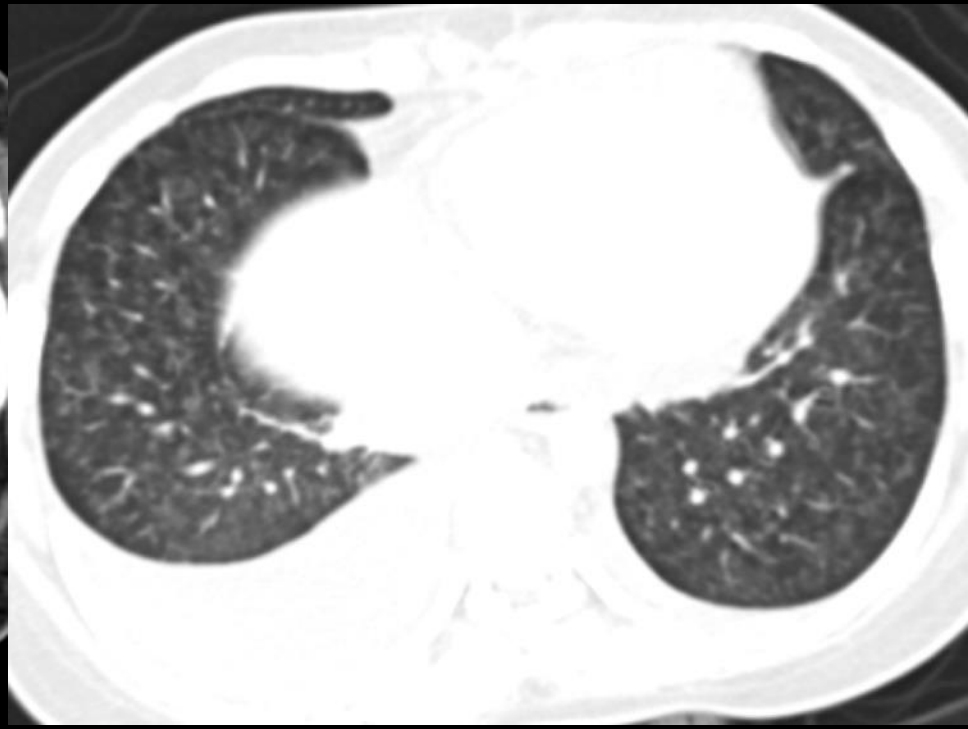


Montani D, Price LC, Dorfmuller P, Achouh L, Jais X, Yaici A, et al. Pulmonary veno-occlusive disease. European Respiratory Journal. 2009 Jan 1;33(1):189–200.

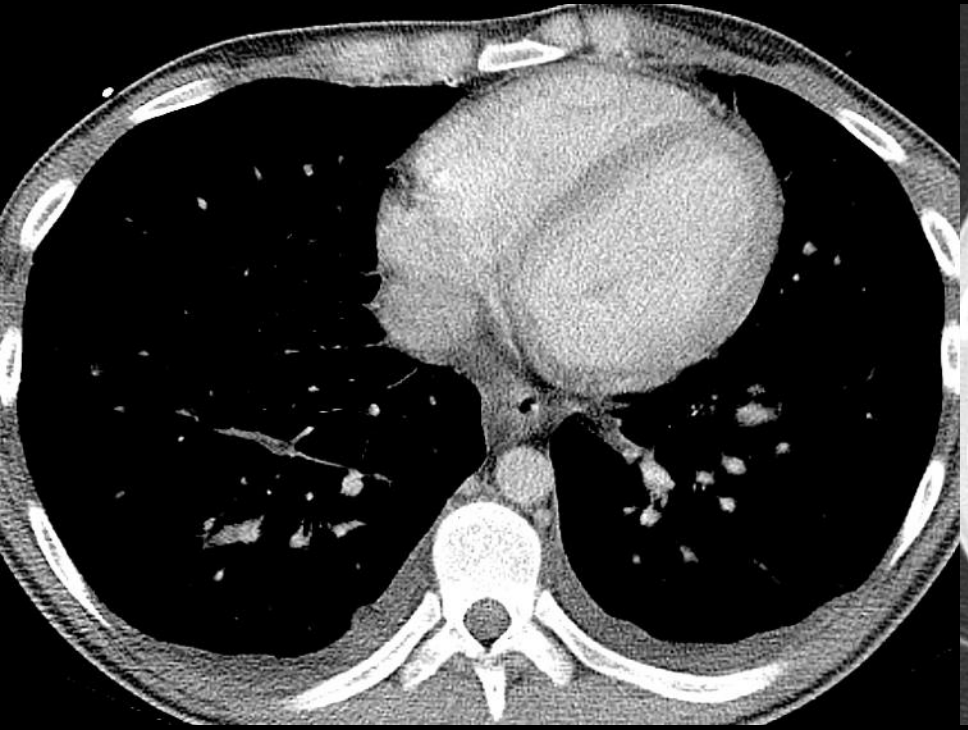
16-year-old – PVOD/PCH

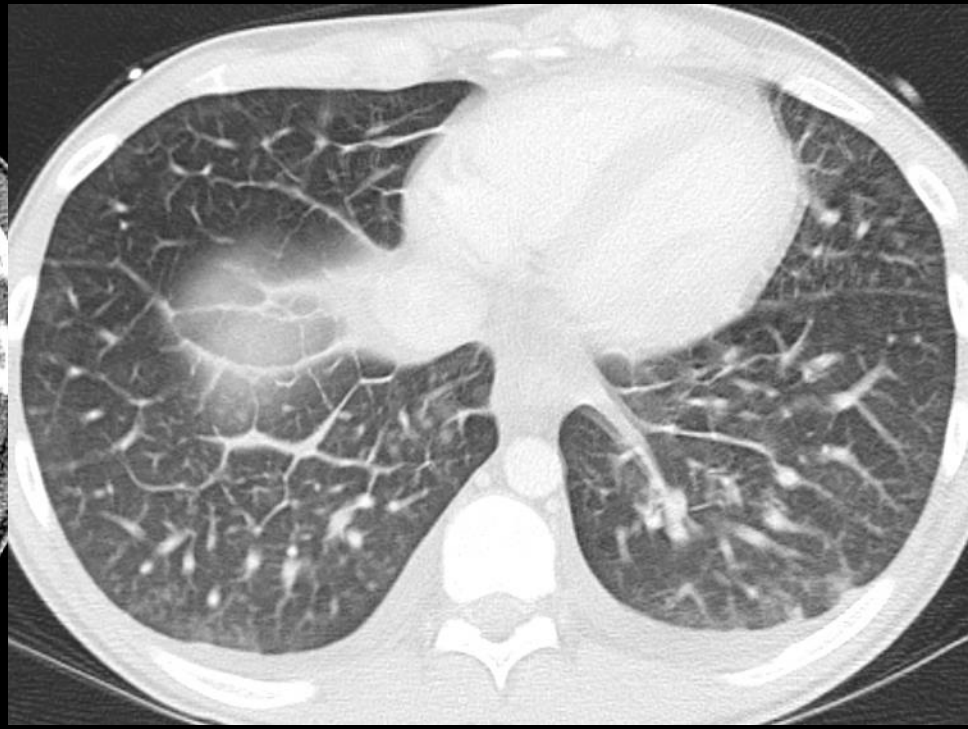


19-year-old – PVOD/PCH



Left Heart Failure (not PVOD/PCH)



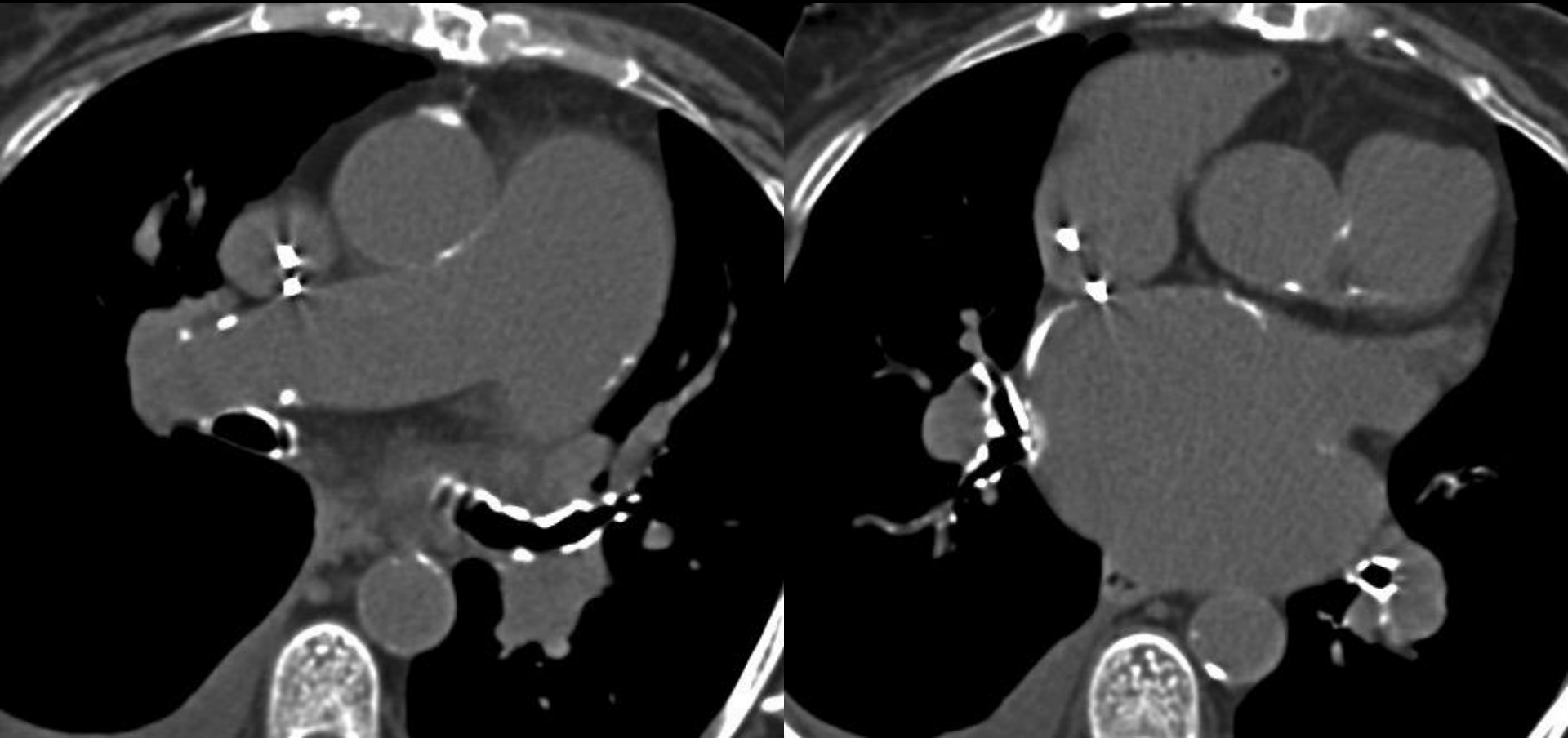


Dilated Left Ventricle

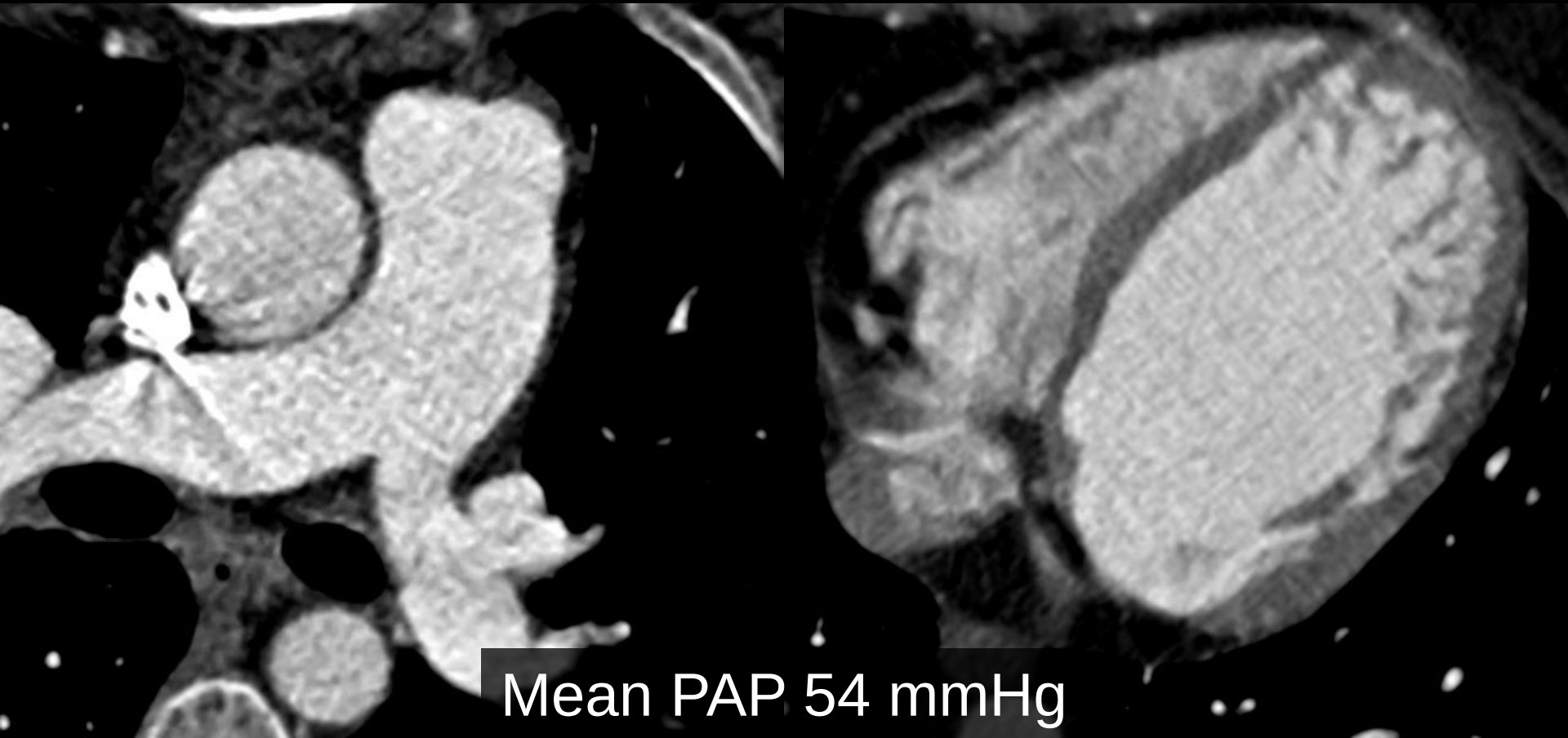
Group 2 – Left Heart Disease

- LV Systolic Dysfunction
- LV Diastolic Dysfunction
- Valvular Disease
- Congenital heart disease
  - Acquired inflow/outflow tract obstruction
  - Congenital cardiomyopathies

Group 2 – Longstanding Mitral Stenosis



Noncompaction cardiomyopathy

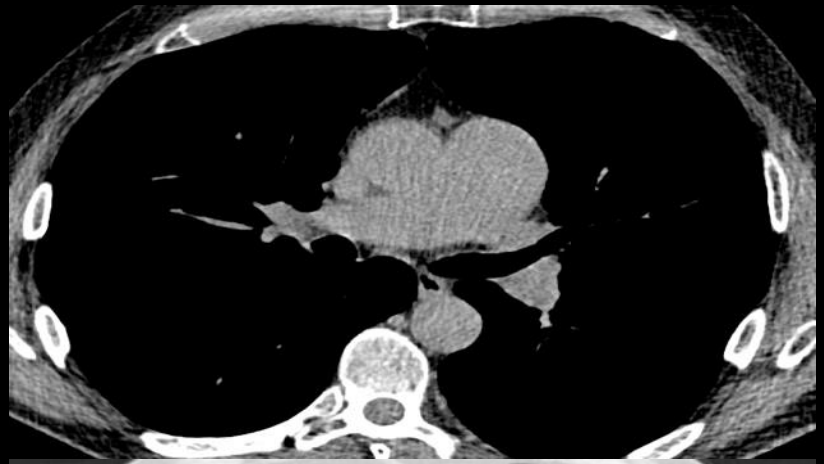


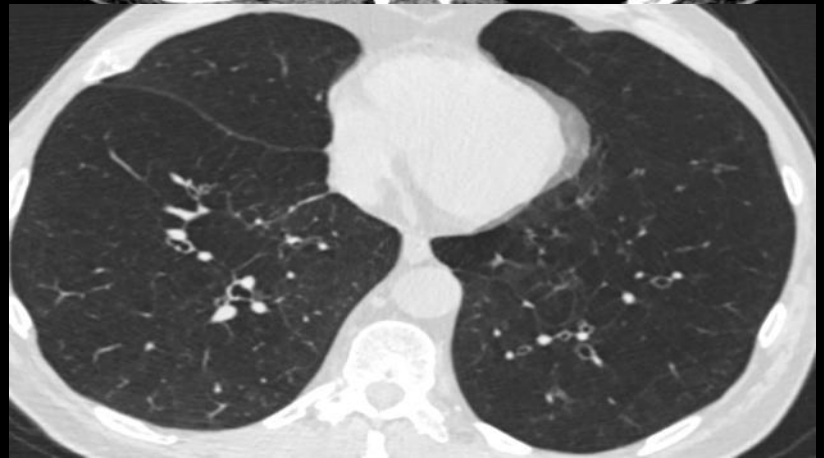
Group 3 – Lung Disease and/or Hypoxia

- COPD
- ILD
- Mixed obstructive and restrictive pattern
- Sleep-disordered breathing
- Alveolar hypoventilation
- Chronic exposure to high altitude
- Developmental lung disease

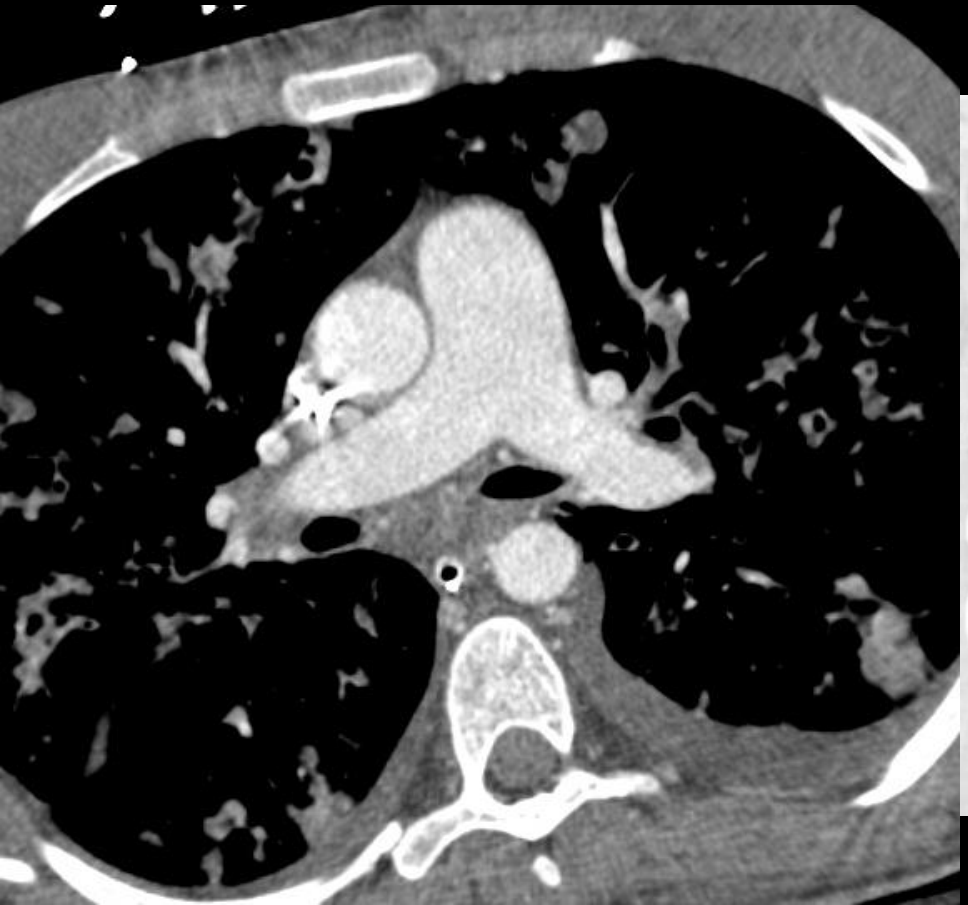
Group 3 – PH in COPD

- Common
- Usually mild but associated with reduced survival
- Lung transplantation is best long-term therapy





Group 3 – Cystic Fibrosis



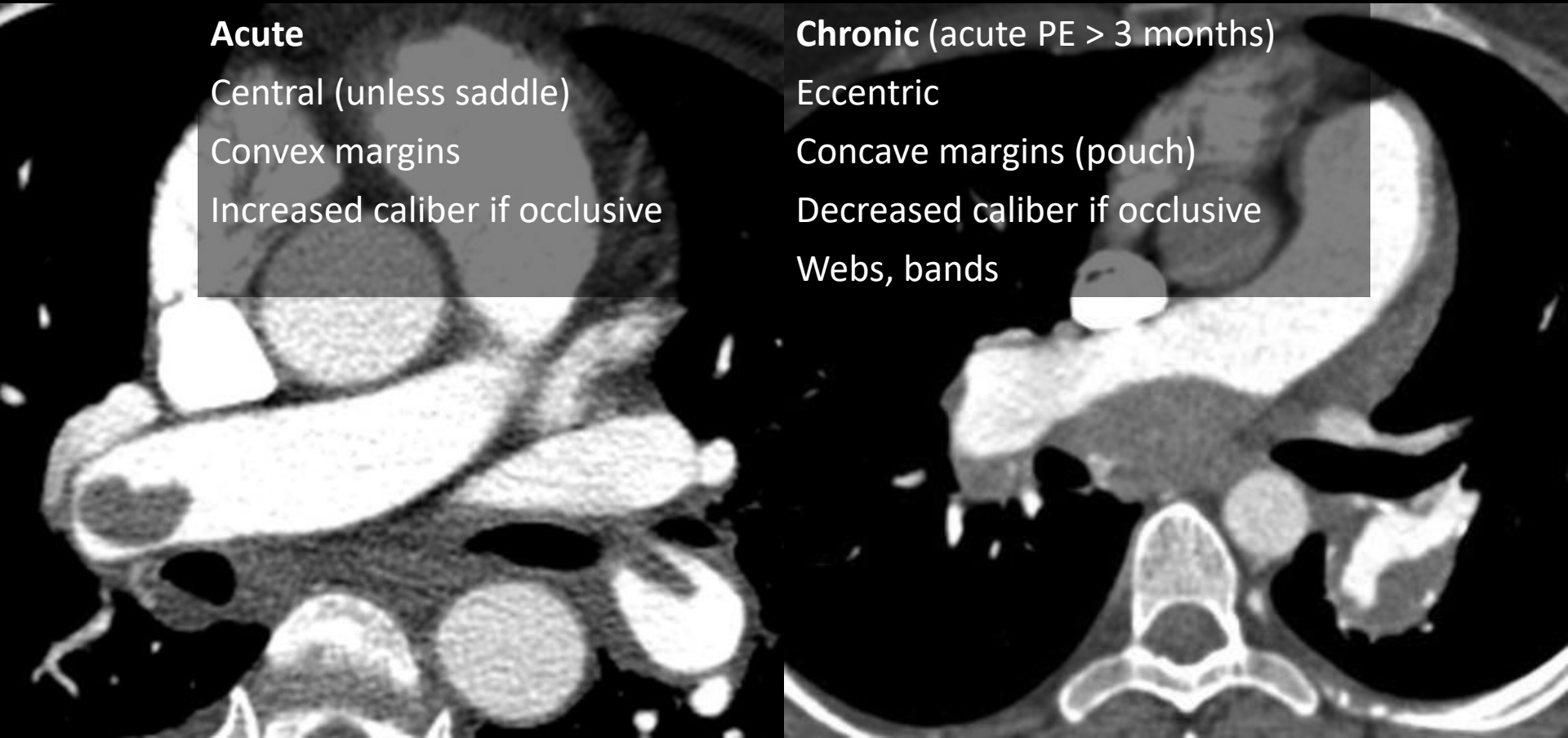
Combined pulmonary fibrosis and emphysema

- AKA CPFE
- PH more frequent/more severe than IPF or COPD alone
- Can occur with IPF or other fibrosis

Group 4 - CTEPH

- Potentially *curable cause of PH*
- Can occur after only ONE episode of acute PE
  - 4-5% of patients with PE develop CTEPH
  - May have no known episode of acute PE

Acute vs Chronic PE



Acute

Central (unless saddle)

Convex margins

Increased caliber if occlusive

Chronic (acute PE > 3 months)

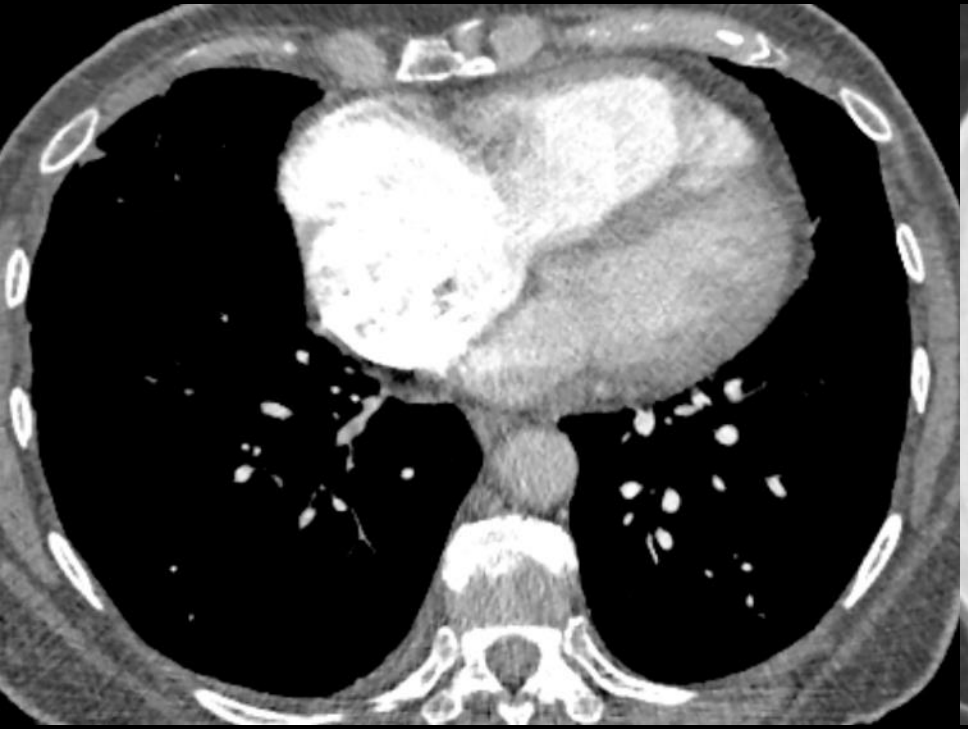
Eccentric

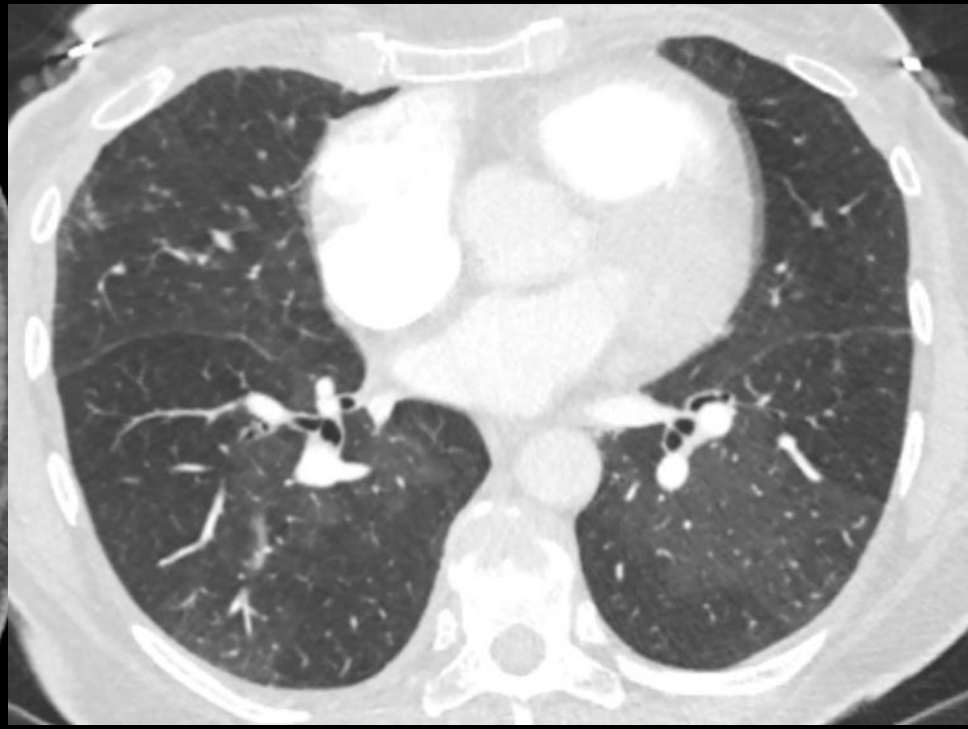
Concave margins (pouch)

Decreased caliber if occlusive

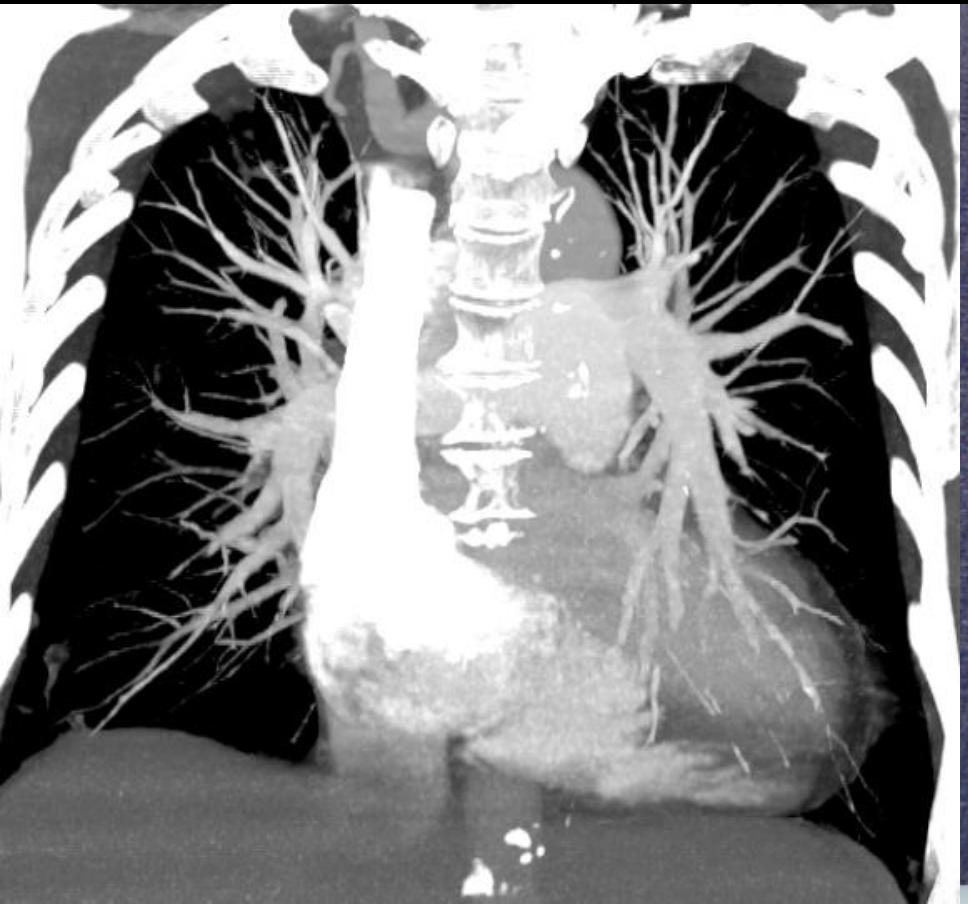
Webs, bands

Worsening Dyspnea 6 months after PE





CTEPH - Thromboendarterectomy

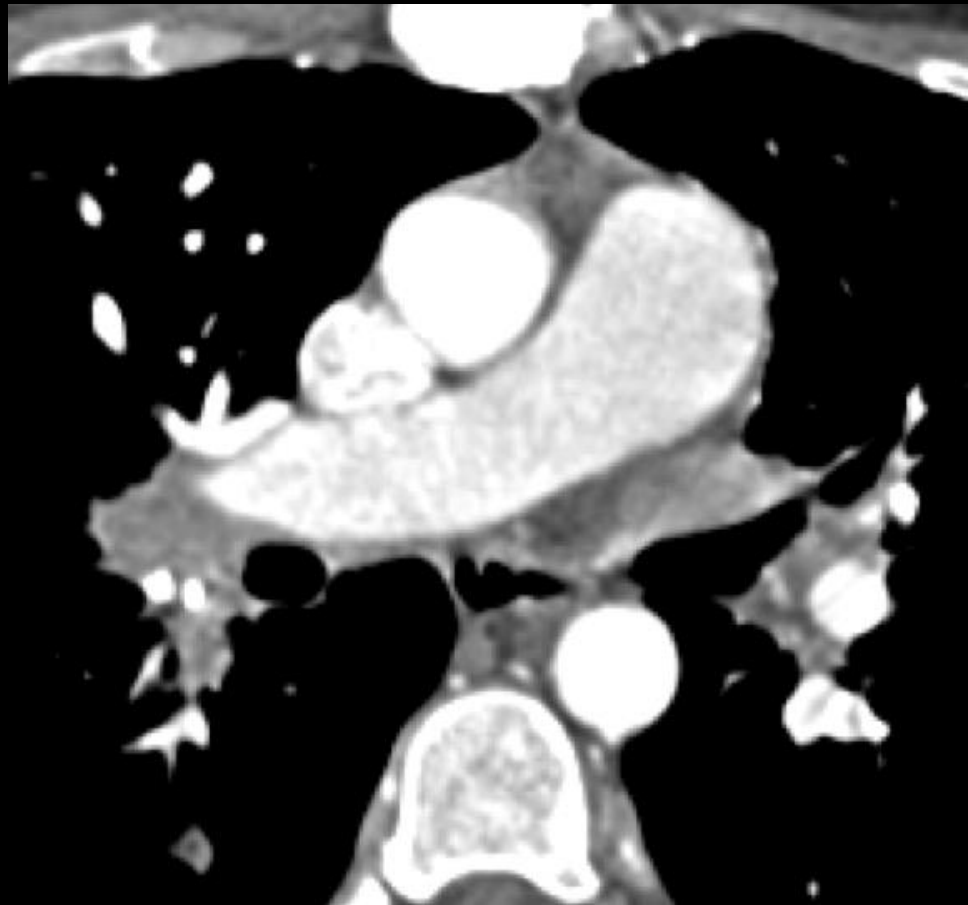


Group 5 – Unclear/Multifactorial

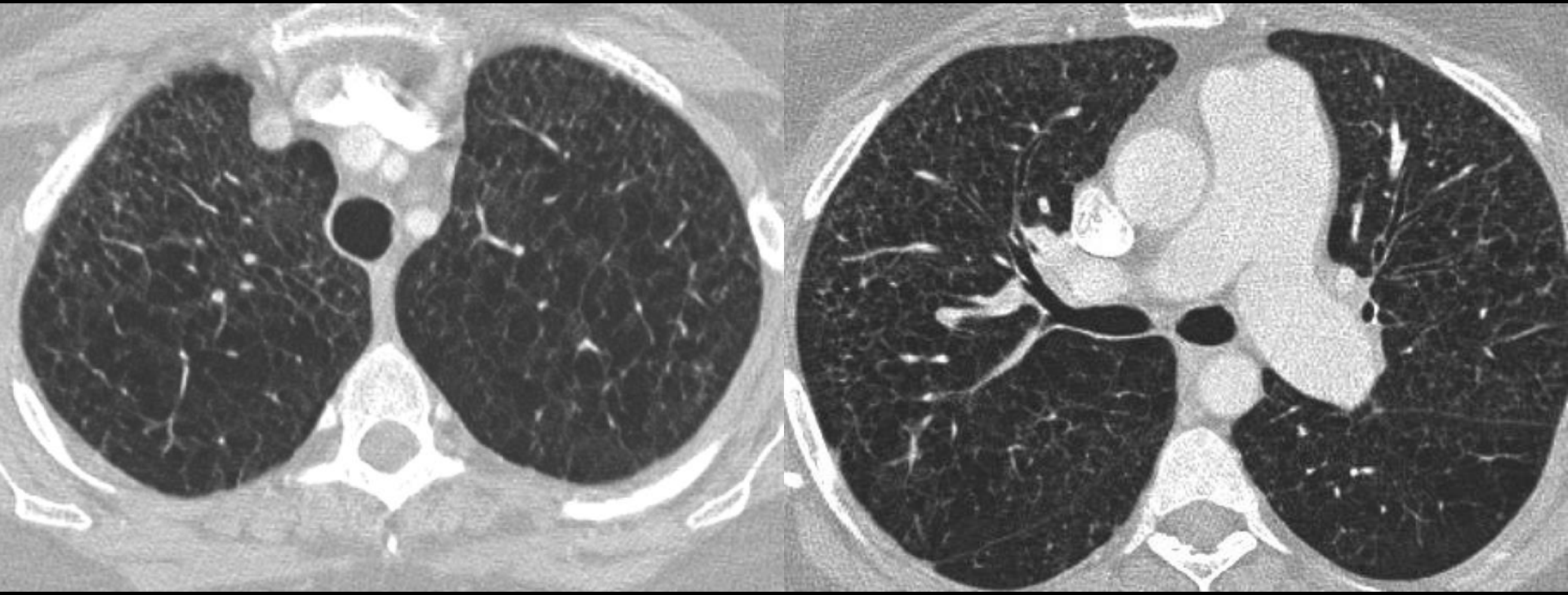
- Hematologic disorders
  - Chronic hemolytic anemia
  - Myeloproliferative disorders
  - Splenectomy
- Systemic disorders
  - Sarcoidosis
  - Pulmonary histiocytosis
  - Lymphangioleiomyomatosis
- Metabolic disorders
- Others
  - Tumoral obstruction
  - Fibrosing mediastinitis
  - Chronic renal failure
  - Segmental PH

Fibrosing mediastinitis

- Confluent soft tissue
- Mass effect on pulmonary vessels
- Enlarged bronchial arteries



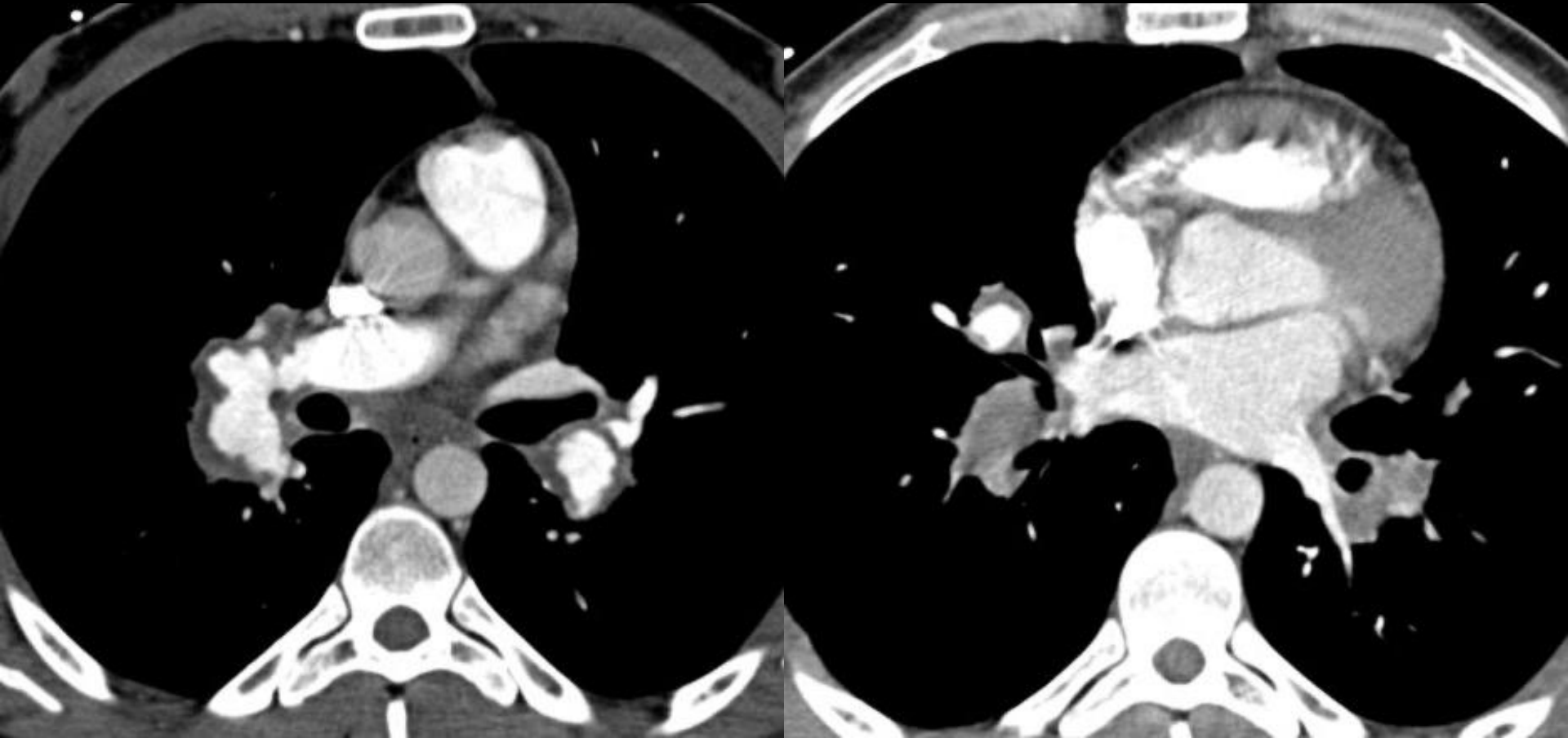
Langerhan cell histiocytosis



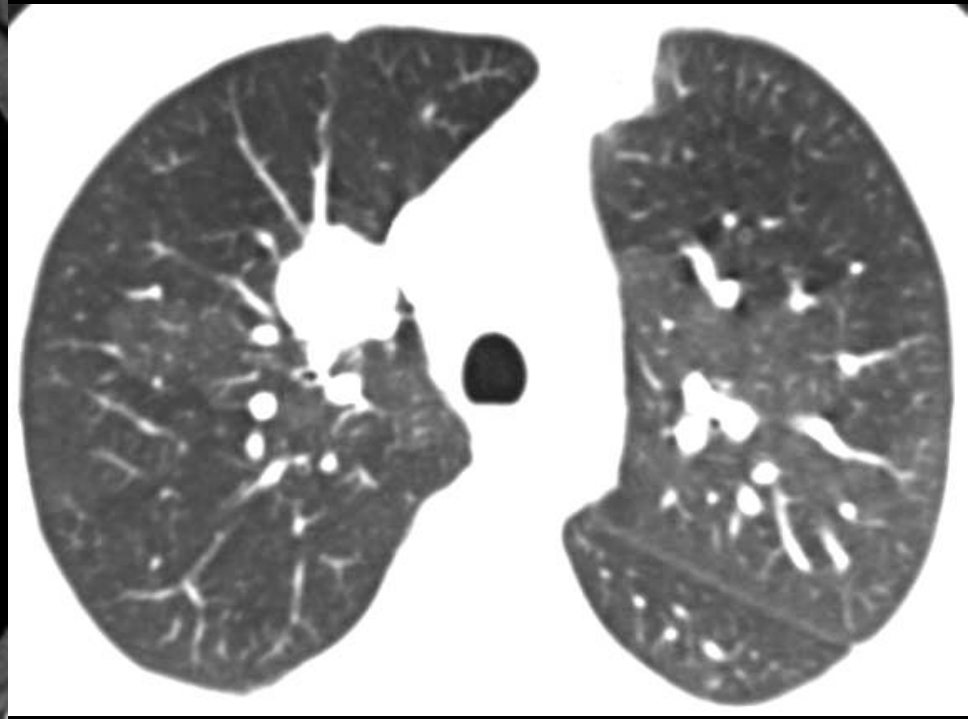
Locally Dilated PA

- Aneurysm or Pseudoaneurysm

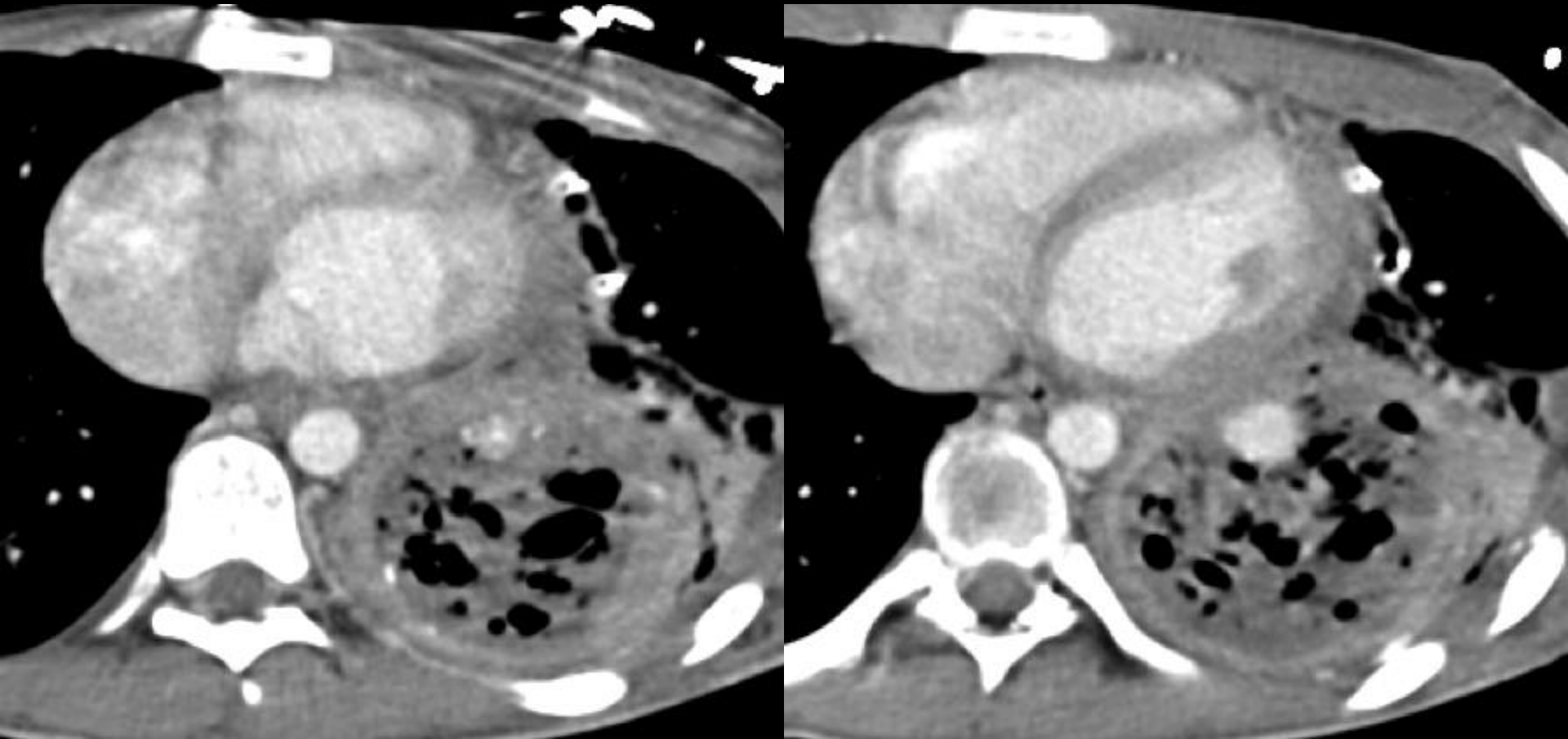
Behçet Disease



Behçet Disease



Mycotic Pseudoaenurysm



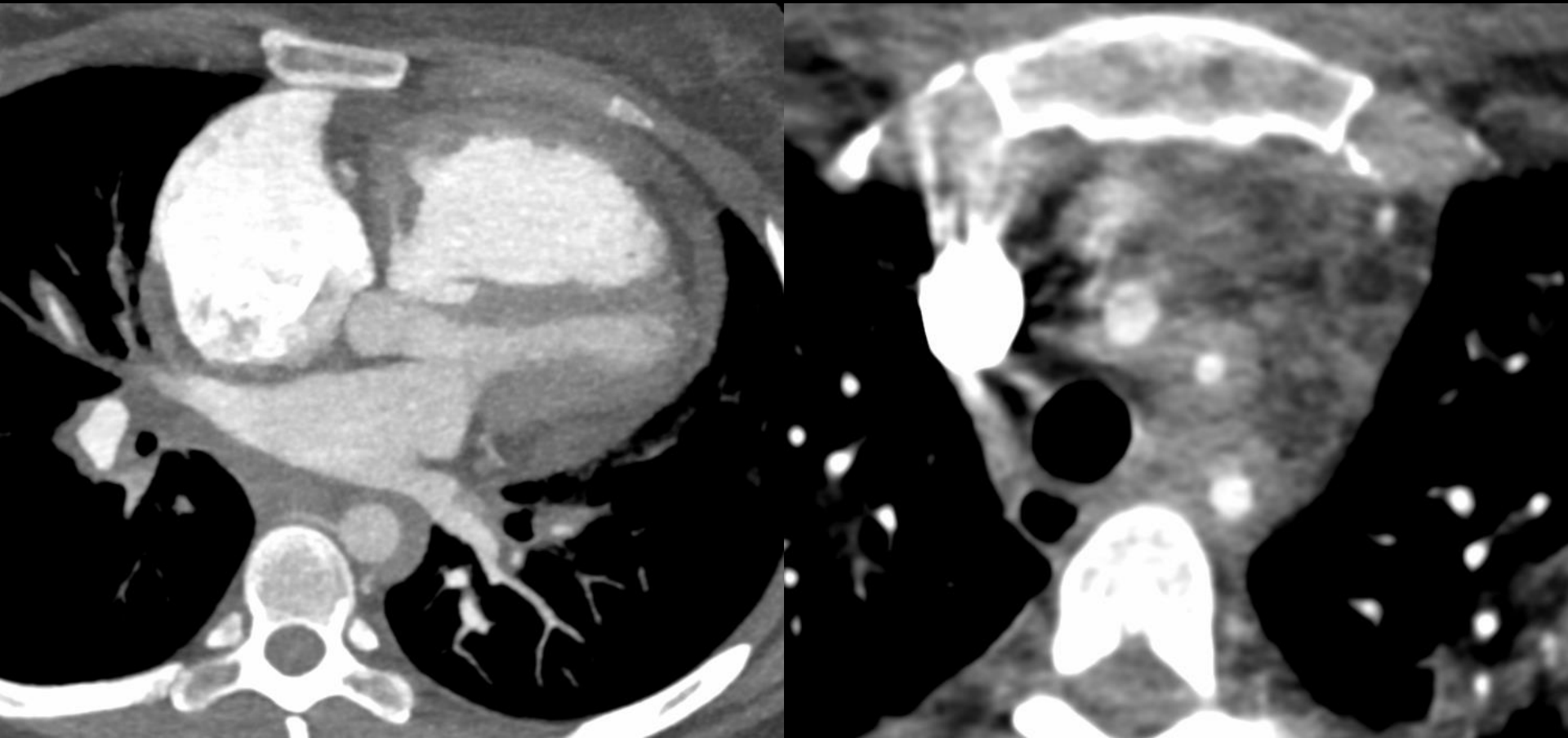
Traumatic PA Pseudoaneurysm

Narrowed/Occluded

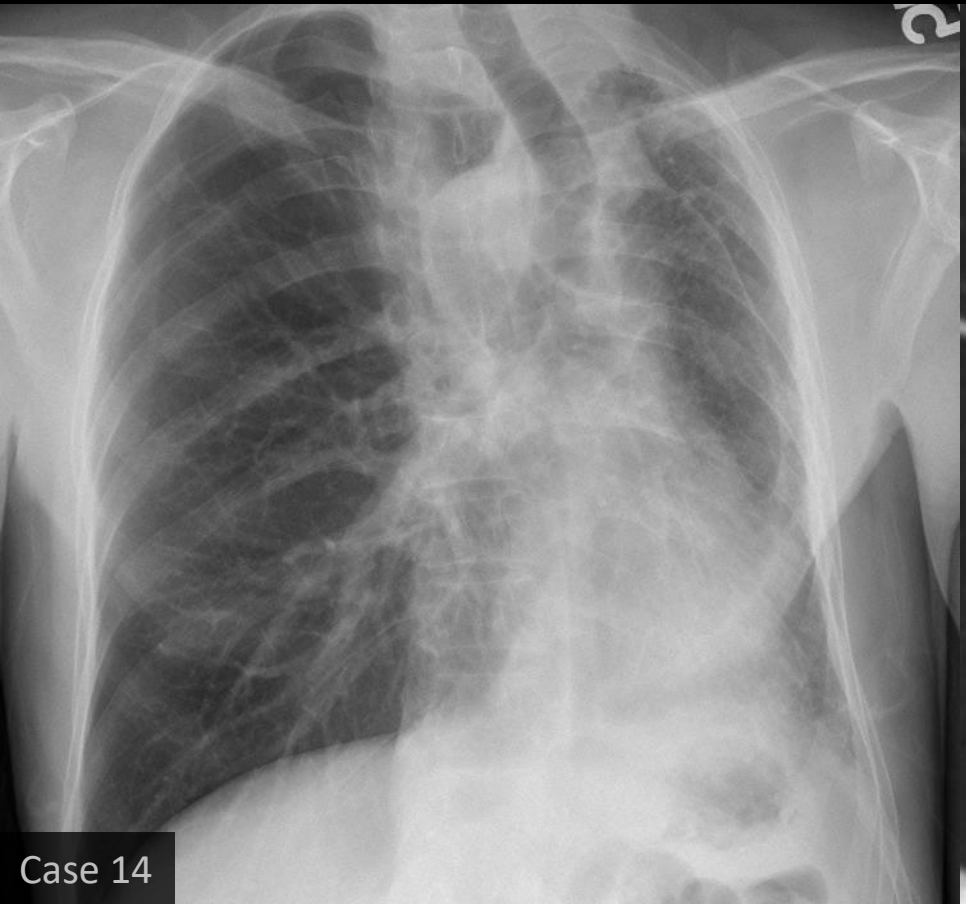
- CTEPH
- Vasculitis
- Congenital

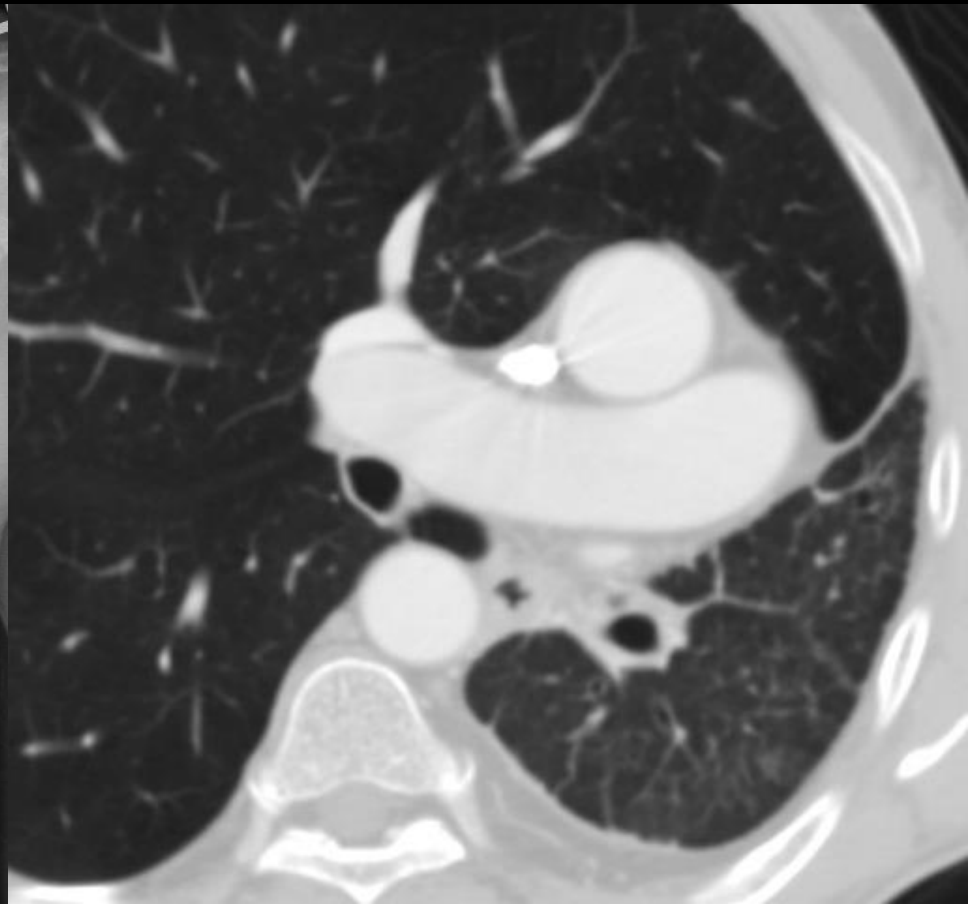
CTEPH

Takayasu Arteritis



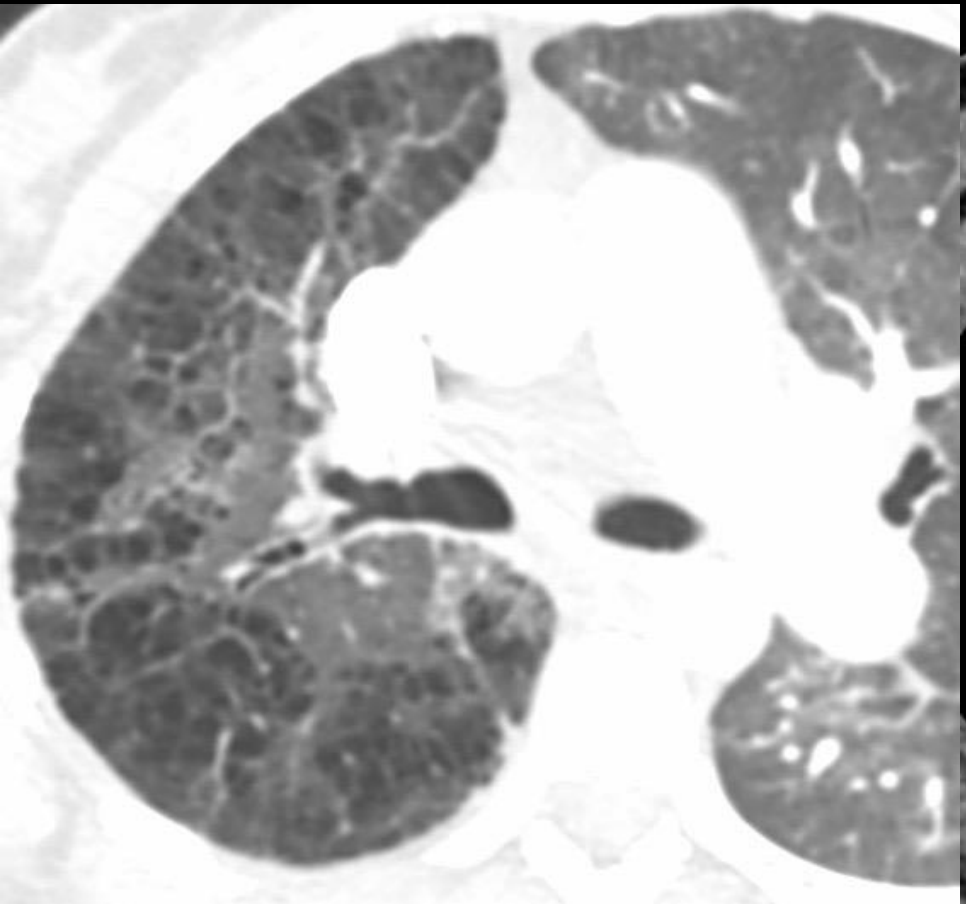
Proximal Interruption of the LPA





Case 14

Proximal Interruption of the RPA

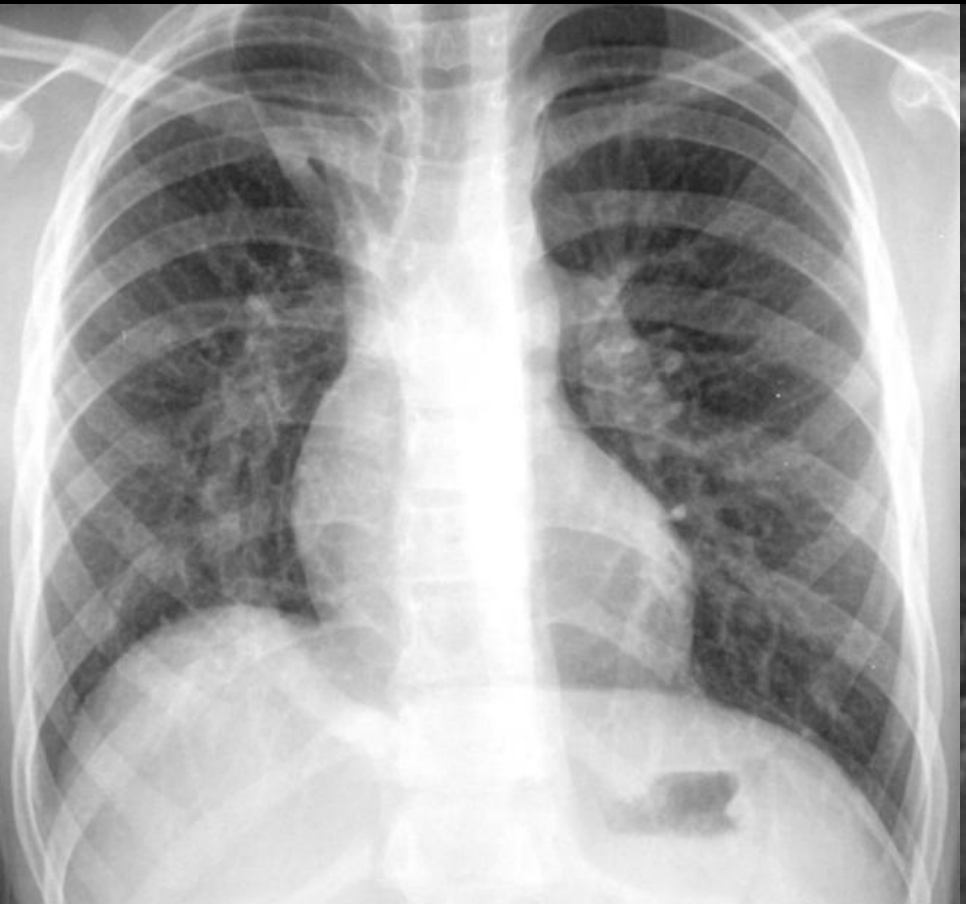


Abnormalities of Vessel Drainage

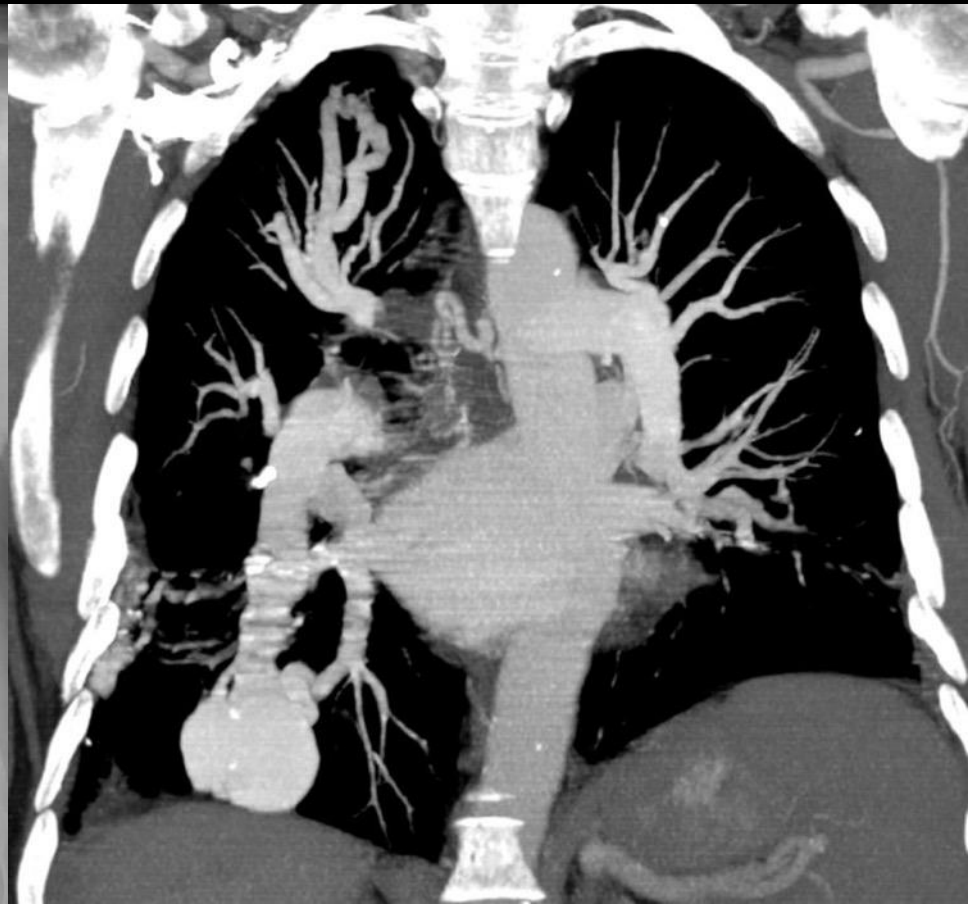
- Anomalous pulmonary venous connection
- Pulmonary arteriovenous malformation
- Meandering Pulmonary Vein

PAPVC

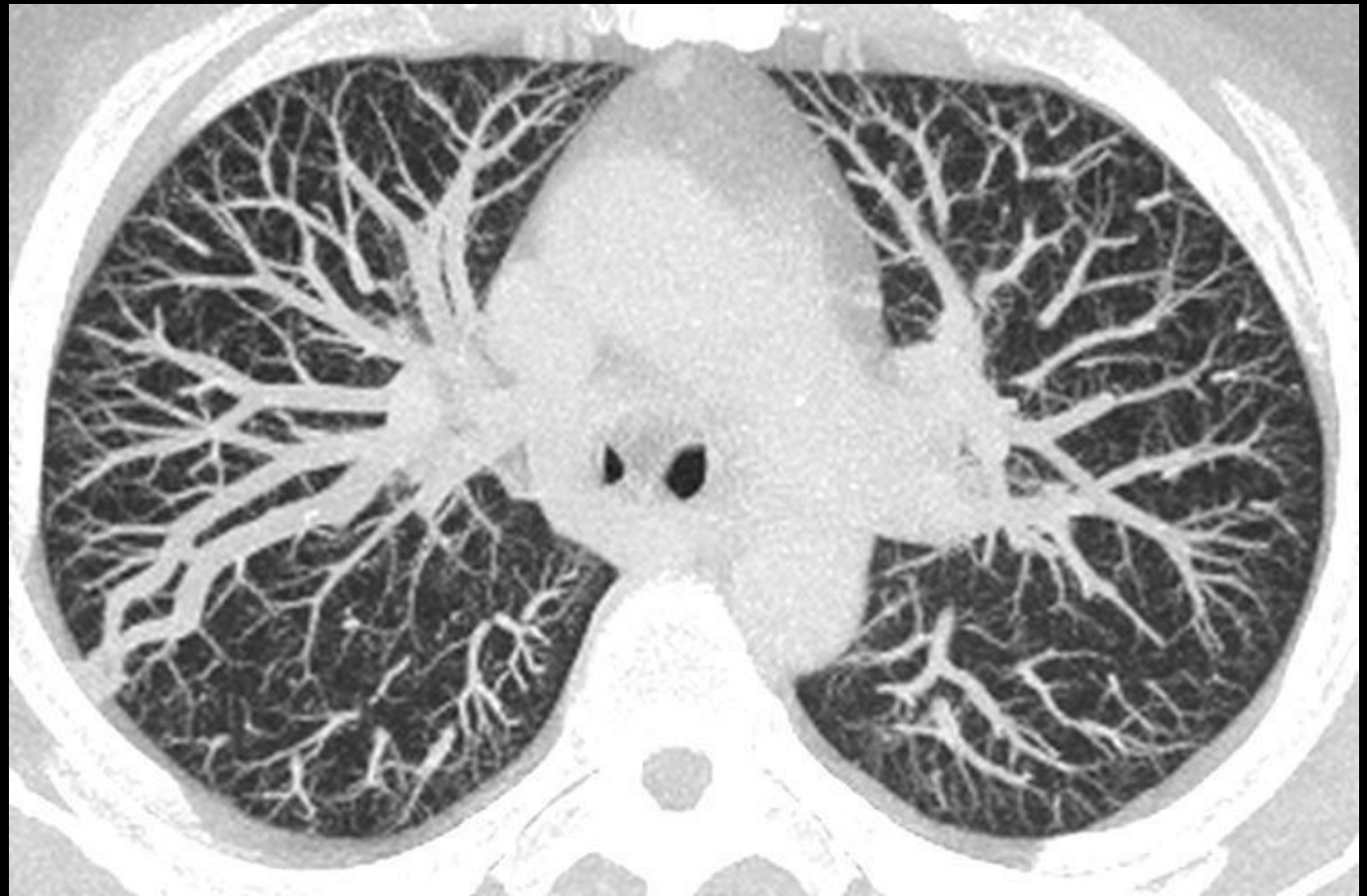
Scimitar Syndrome



Pulmonary AVM

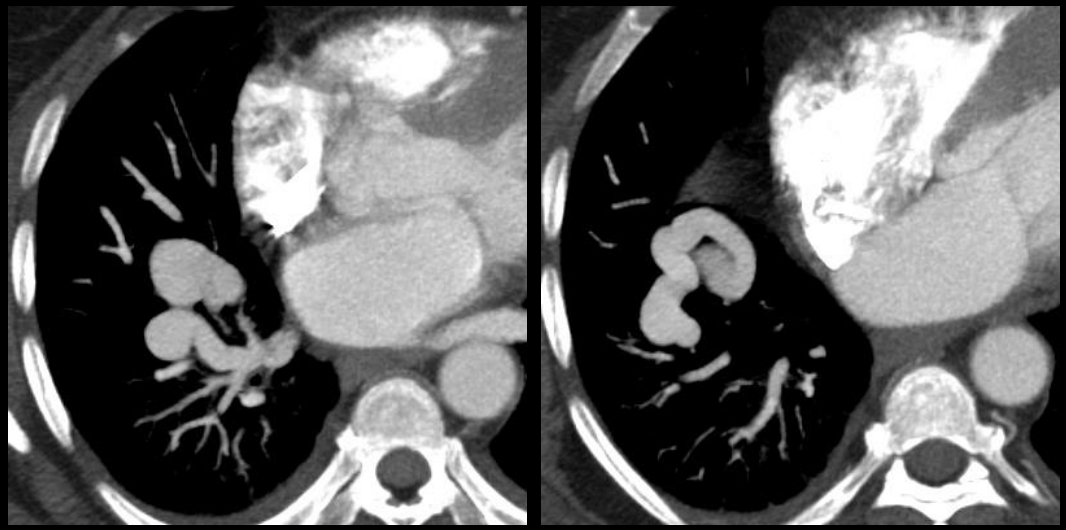


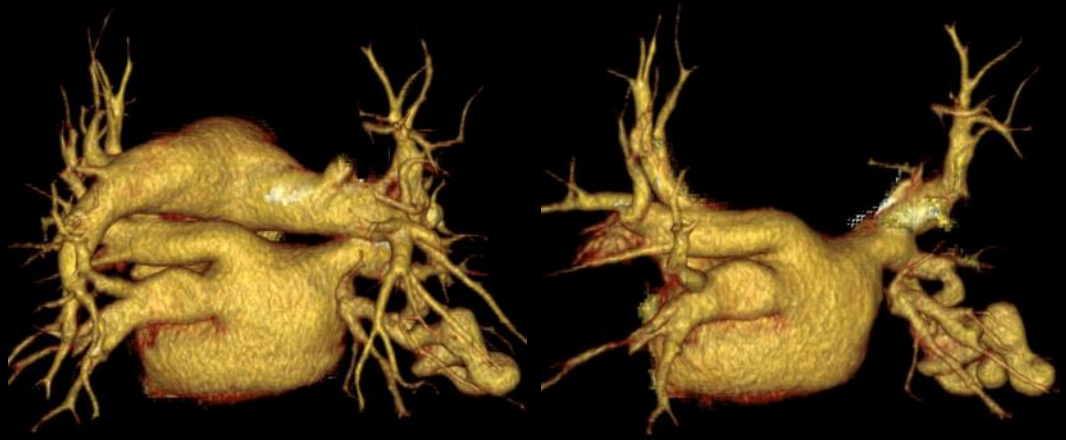
Pulmonary AVM



Meandering Pulmonary Vein

- Rare vascular anomaly
  - Abnormal course
  - Normal drainage
- Not a shunt → negative bubble study
- Little physiologic significance

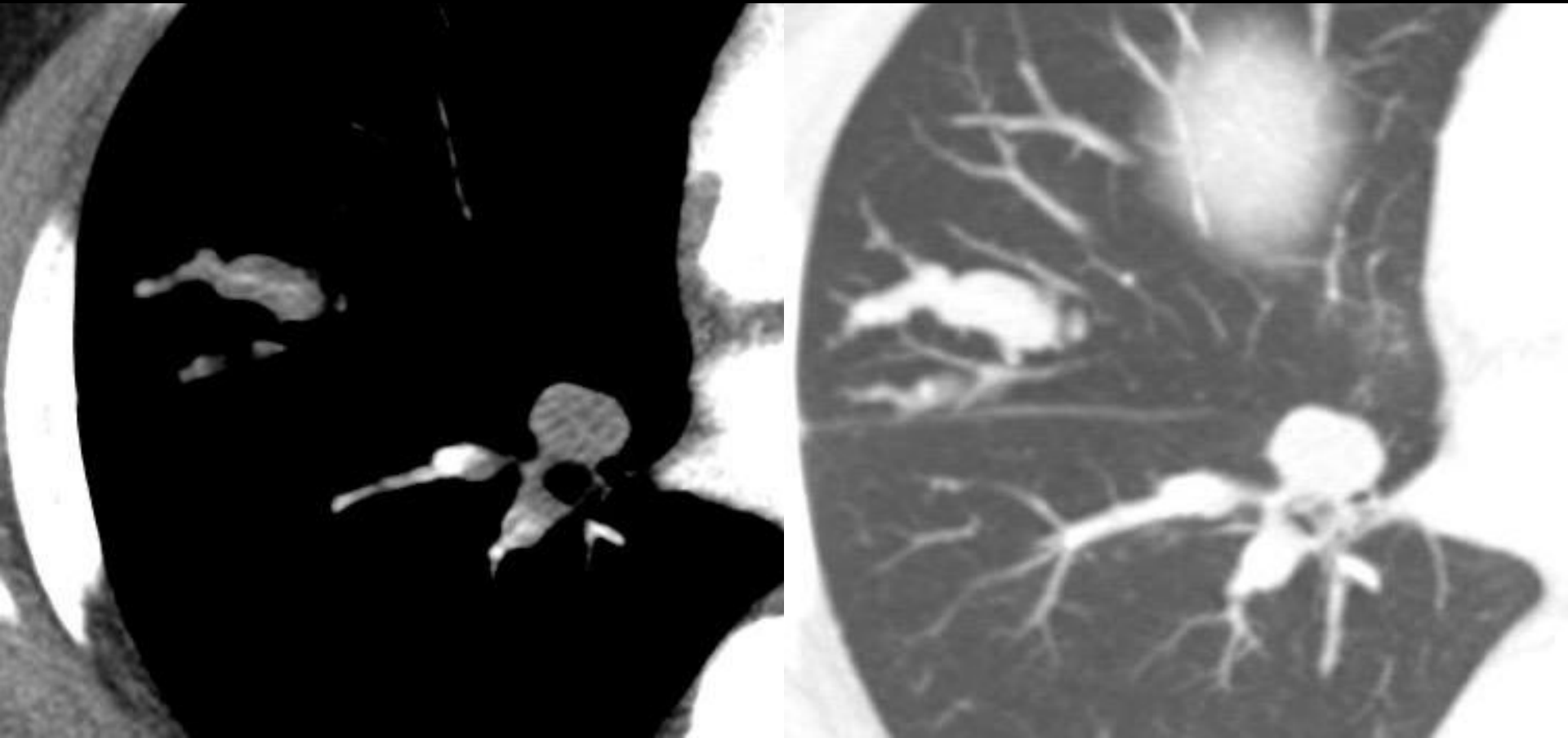




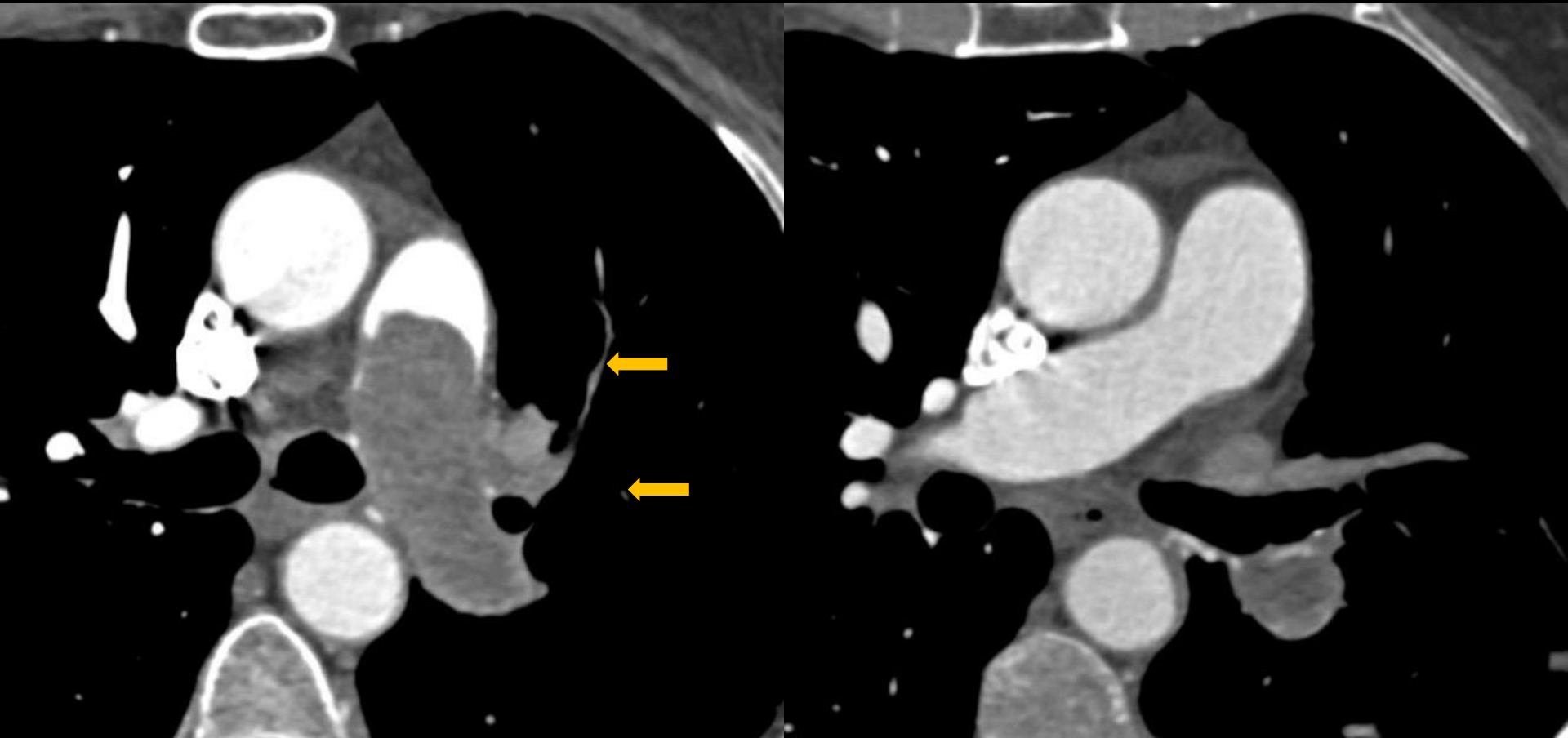
Abnormalities of Vessel Lumen

- Filling defect
  - Pulmonary thromboembolism
  - Tumor embolism
  - Pulmonary artery sarcoma
  - Foreign body

Tumor Embolism



Pulmonary Artery Sarcoma



Nonthrombotic Pulmonary Embolism

Any substance in the venous circulation can embolize to the lungs

Cause mechanical obstruction and/or inflammatory cascade

The right ventricle is *very sensitive* to changes in pressure

Macroscopic vs Microscopic

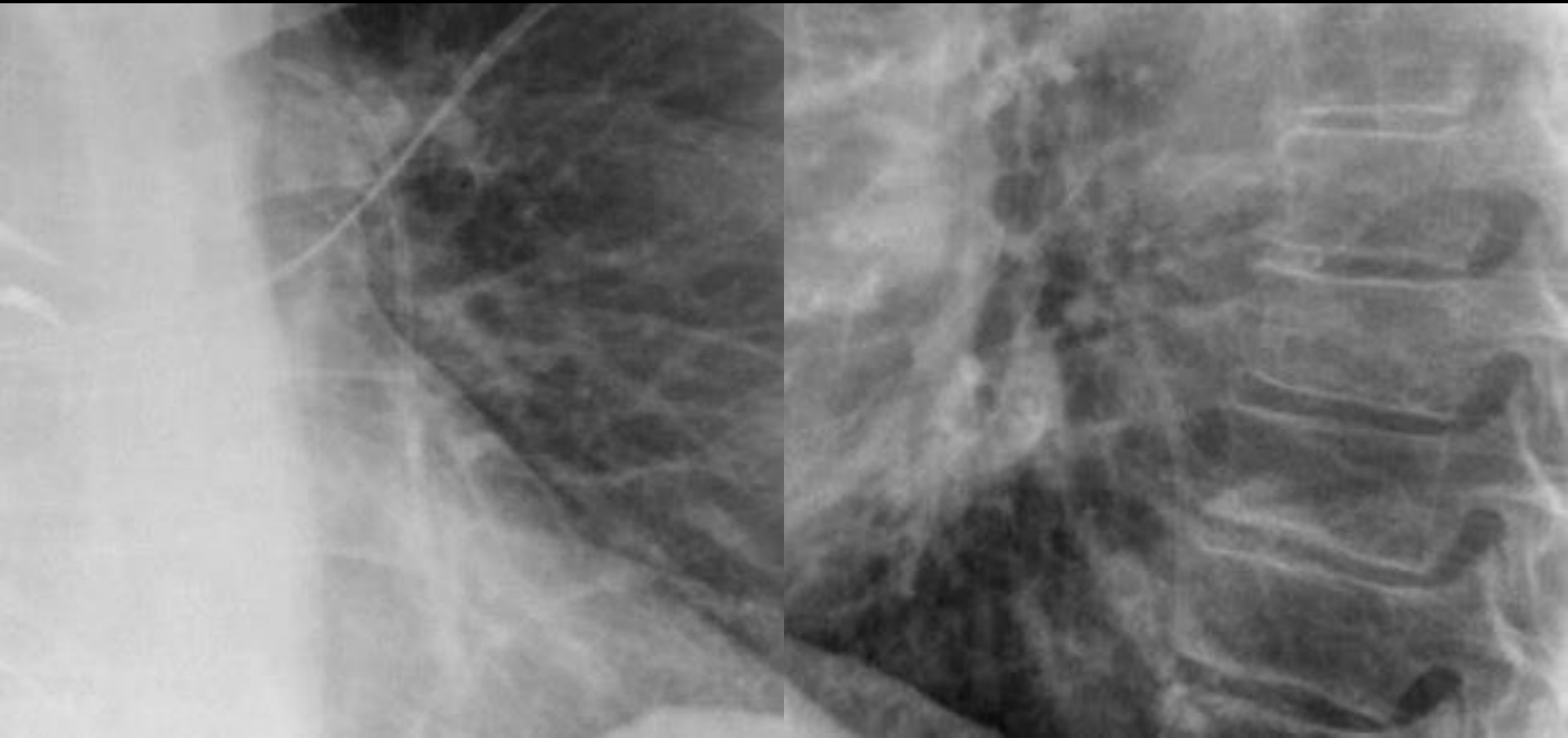
- Bullet
- IVC Filter
- Brachytherapy seed
- Wire
- Methylmethacrylate
- Mercury
- Catheter
- Air

- Excipient
- Amniotic Fluid
- Silicone
- Fat
- Ethiodol
- Chemotherapy

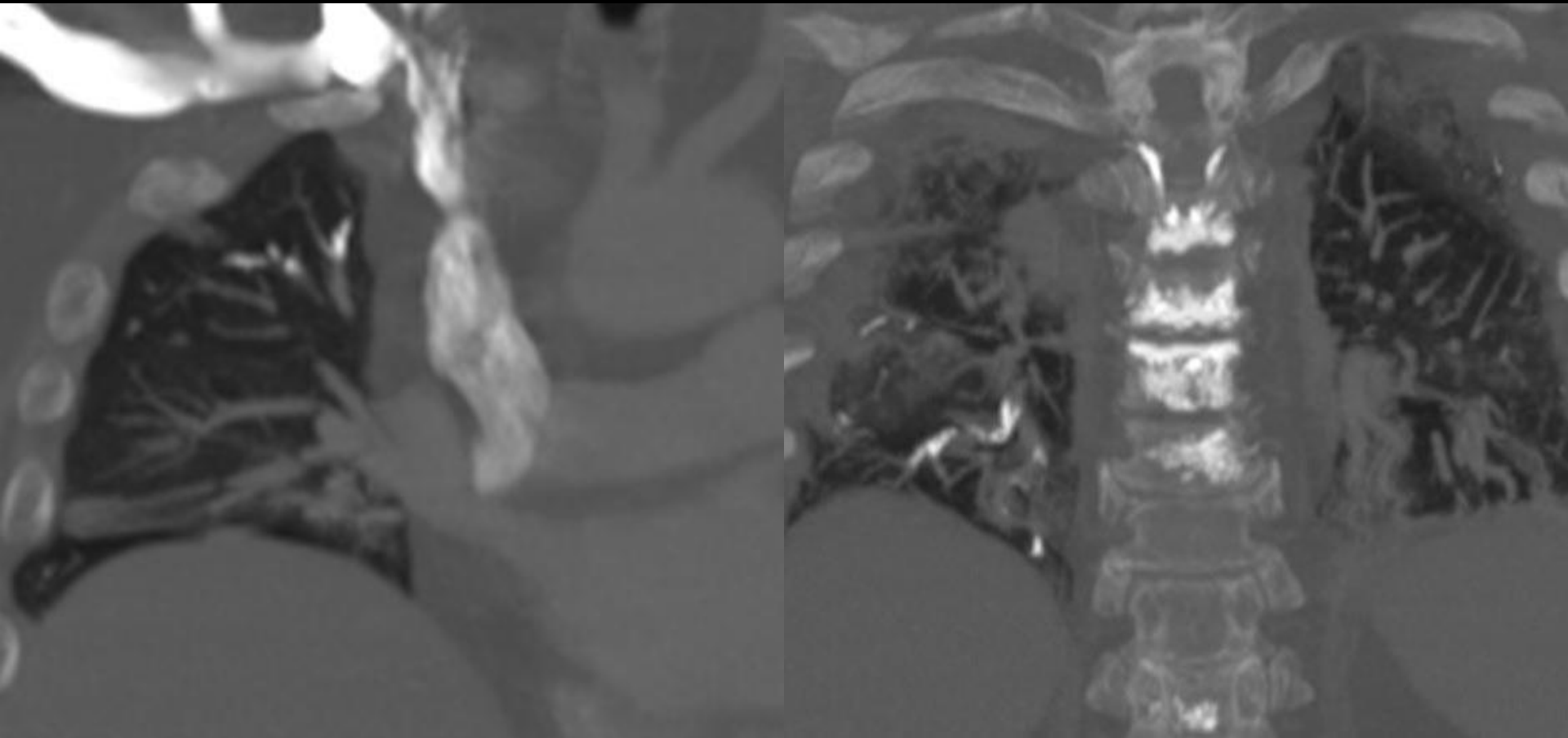
Tumor

Bacteria/Fungi

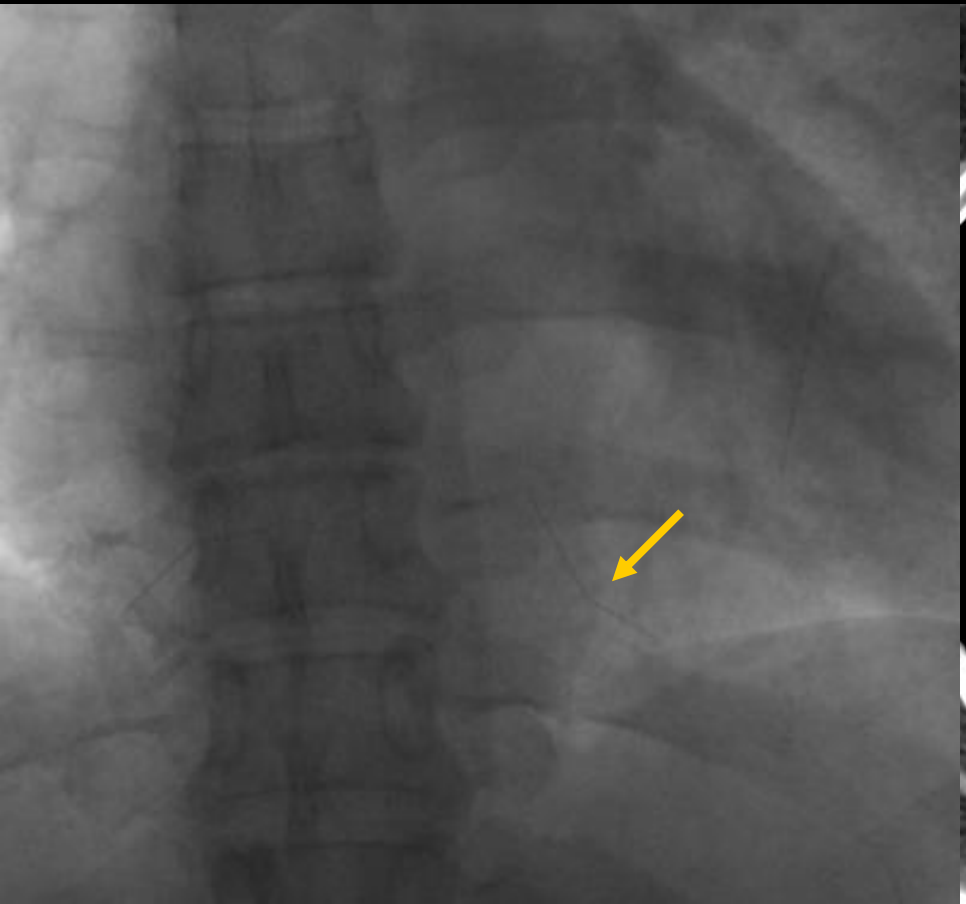
Macroscopic – Catheter Fragment

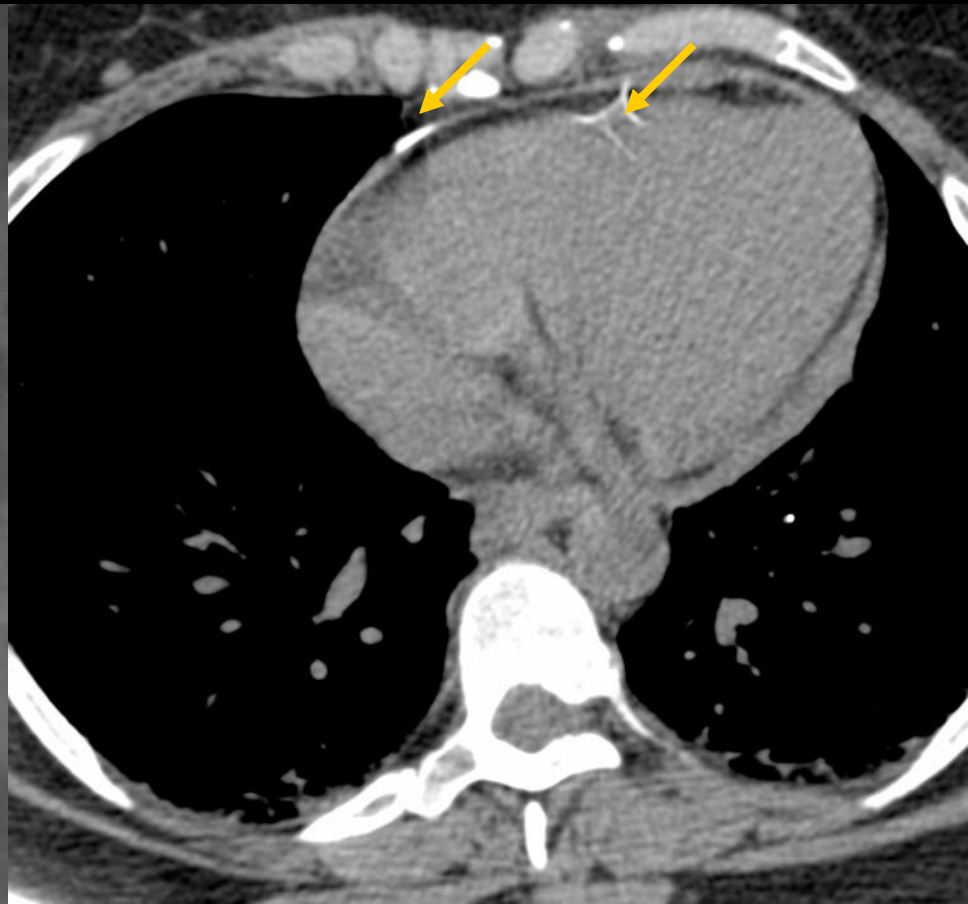


Macroscopic - Methylmethacrylate

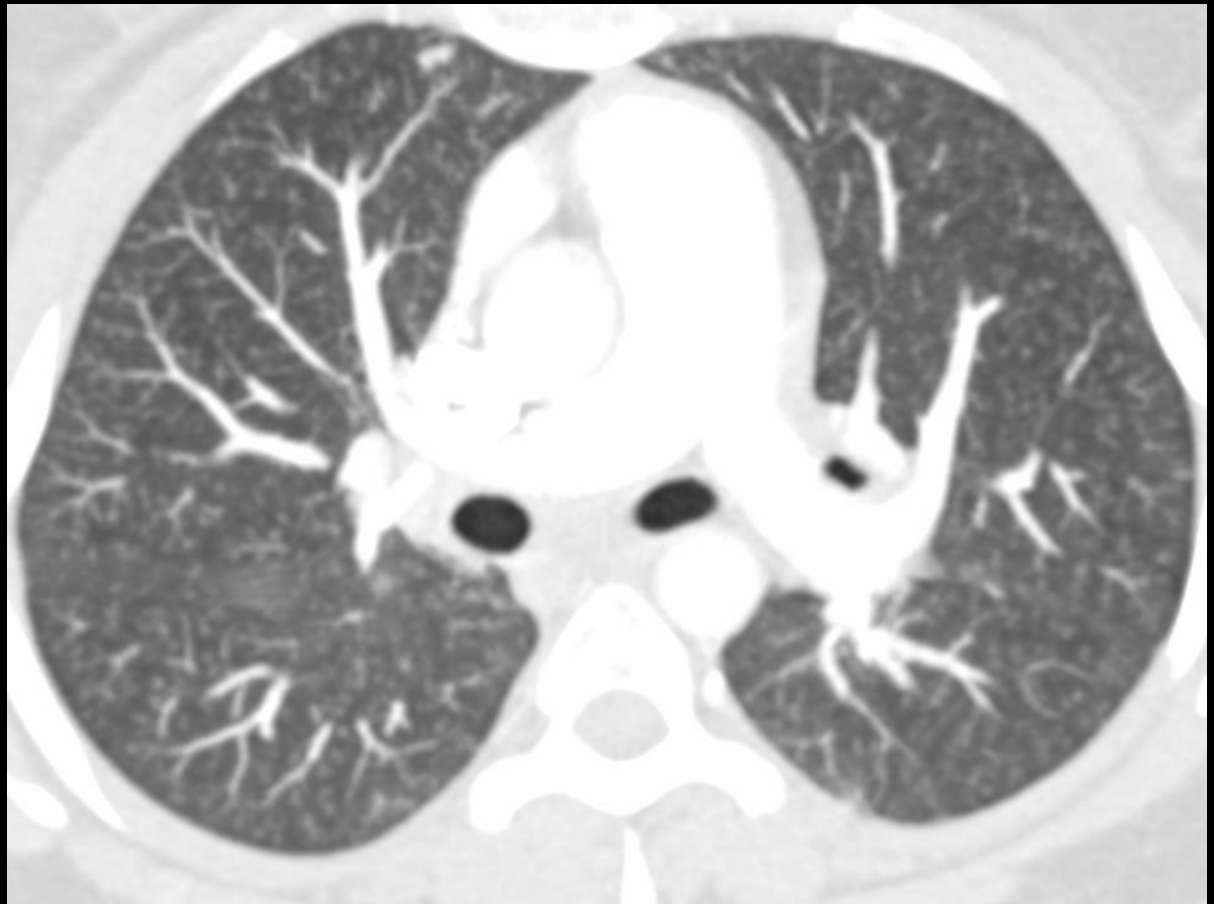


Macroscopic – IVC Filter Prongs



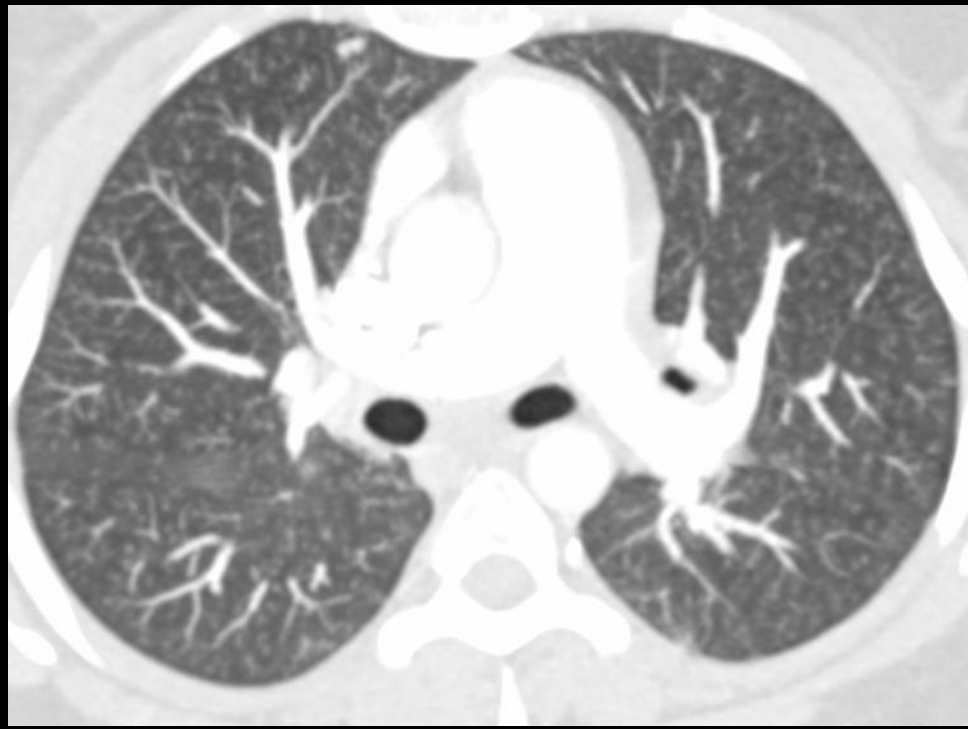


Microscopic – Excipient Lung Disease



Microscopic – Excipient Lung Disease

- Tablets crushed and injected intravenously
- Excipient = inert binder of medication



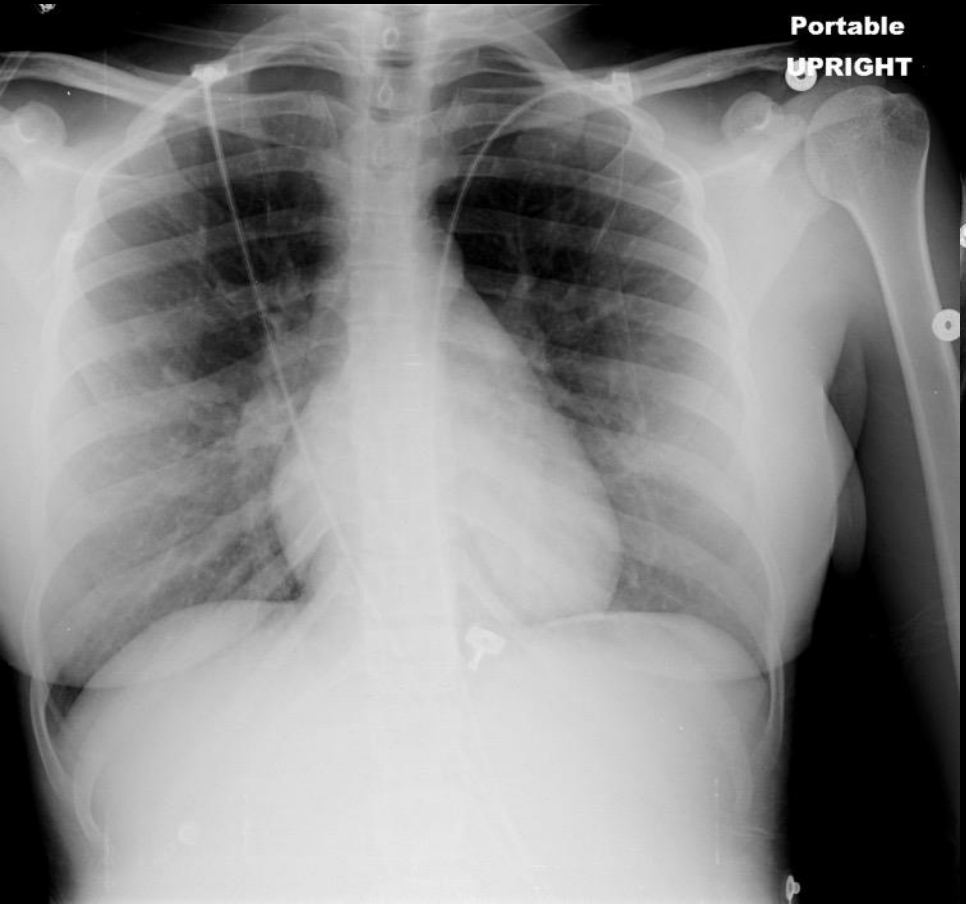
Nguyen et al. Pulmonary effects of i.v. injection of crushed oral tablets: "excipient lung disease". AJR Am J Roentgenol 2014; 203:W506-515

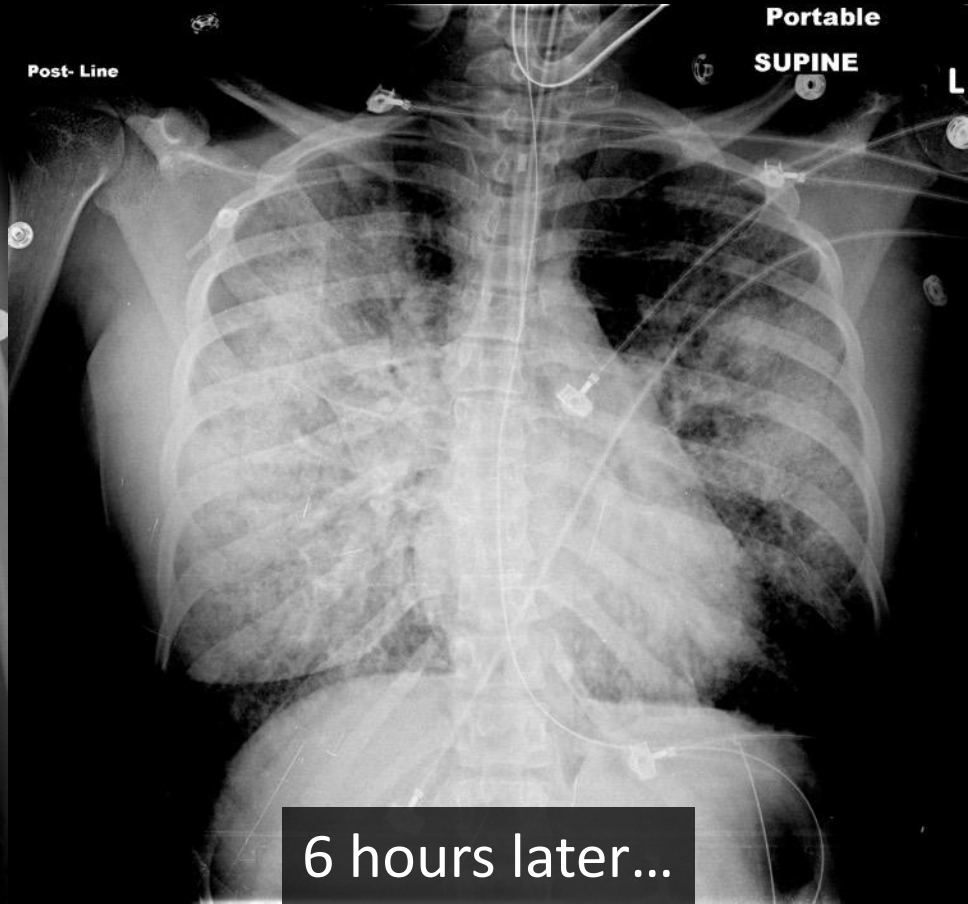
Microscopic – Excipient Lung Disease

- Right heart failure
- Unexplained fever/bacteremia
- Indwelling catheters

Nguyen et al. Pulmonary effects of i.v. injection of crushed oral tablets: "excipient lung disease". AJR Am J Roentgenol 2014; 203:W506-515

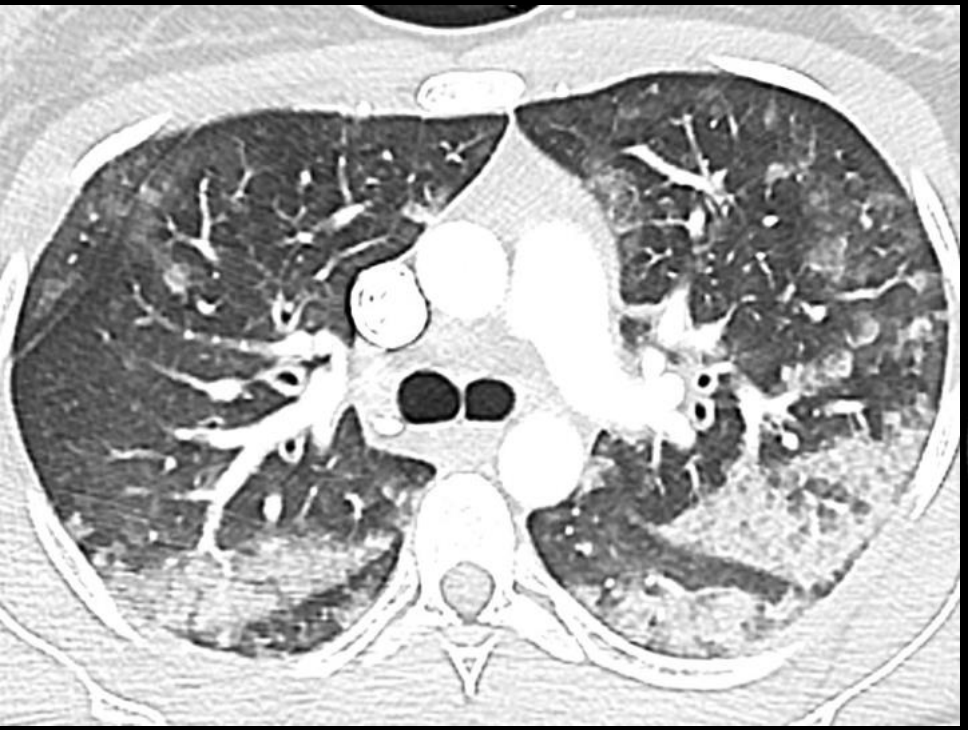
20-year-old after a cosmetic procedure





Microscopic – Silicone Embolization

Microscopic – Silicone Embolization



Microscopic – Silicone Embolization

- Chemical pneumonitis

  → vasoconstriction

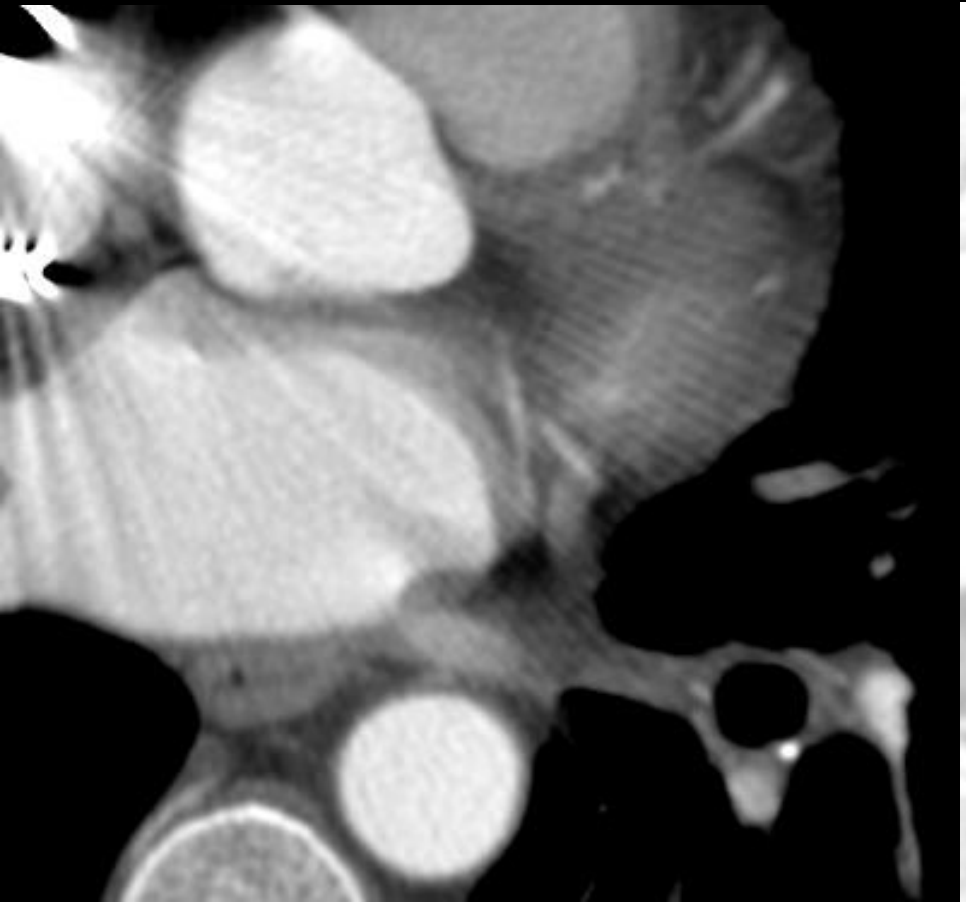
  → acutely elevated right heart pressures

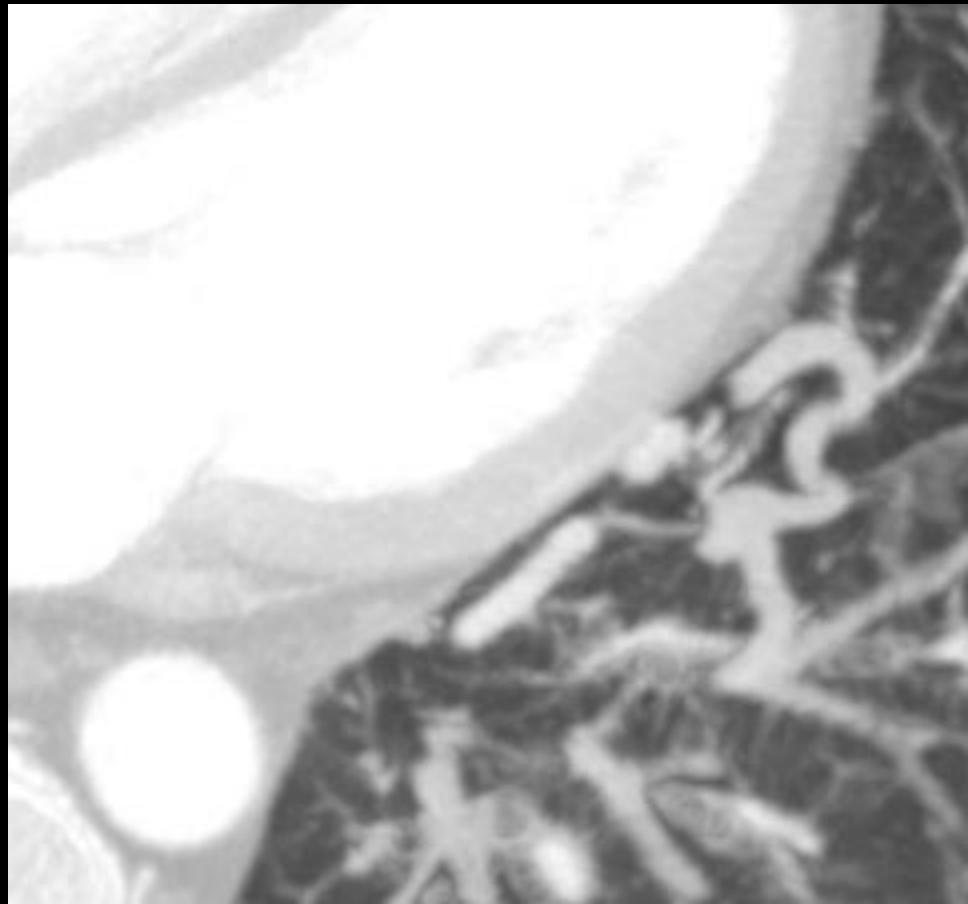
  → right heart failure

Pulmonary Veins

- Stenosis/Occlusion

Pulmonary Vein Stenosis from Ablation





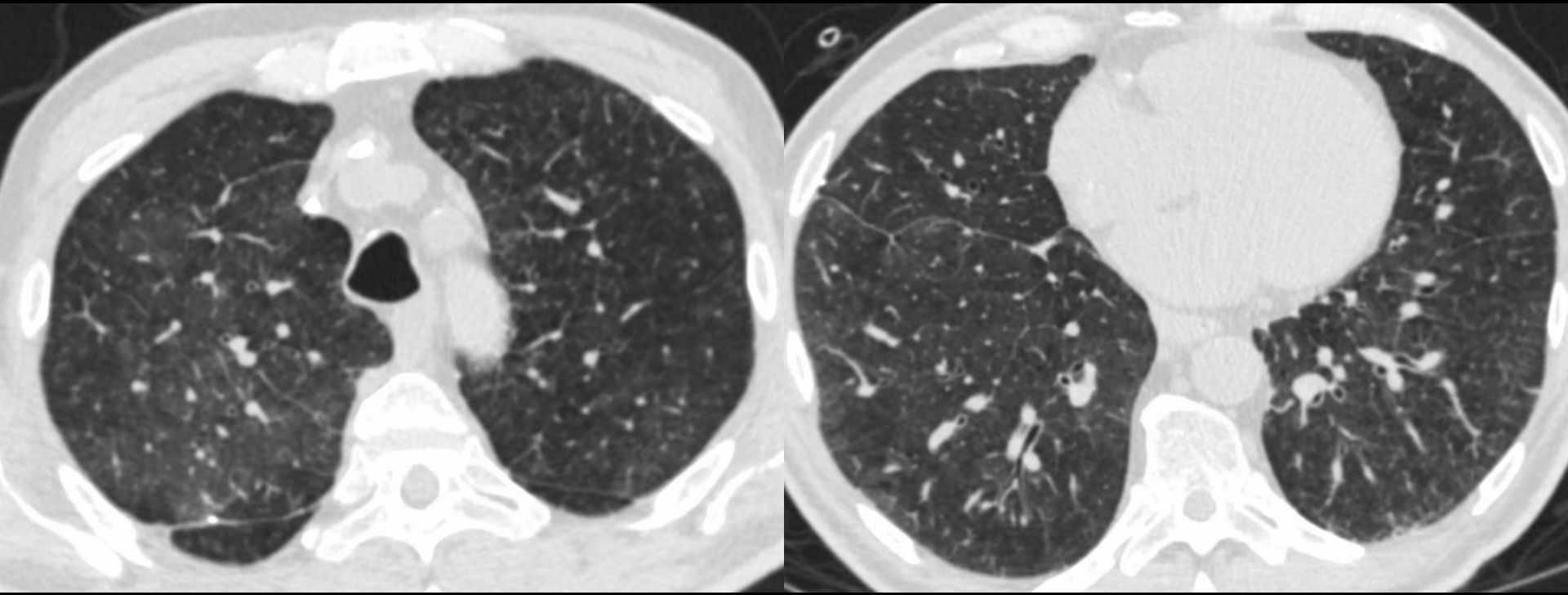
Referred for “ILD”

Pulmonary vein stenosis from ablation

Small Vessels

- PVOD/PCH
- Vasculitis/Capillaritis

PVOD/PCH



59-year-old with PVOD/PCH overlap on explant

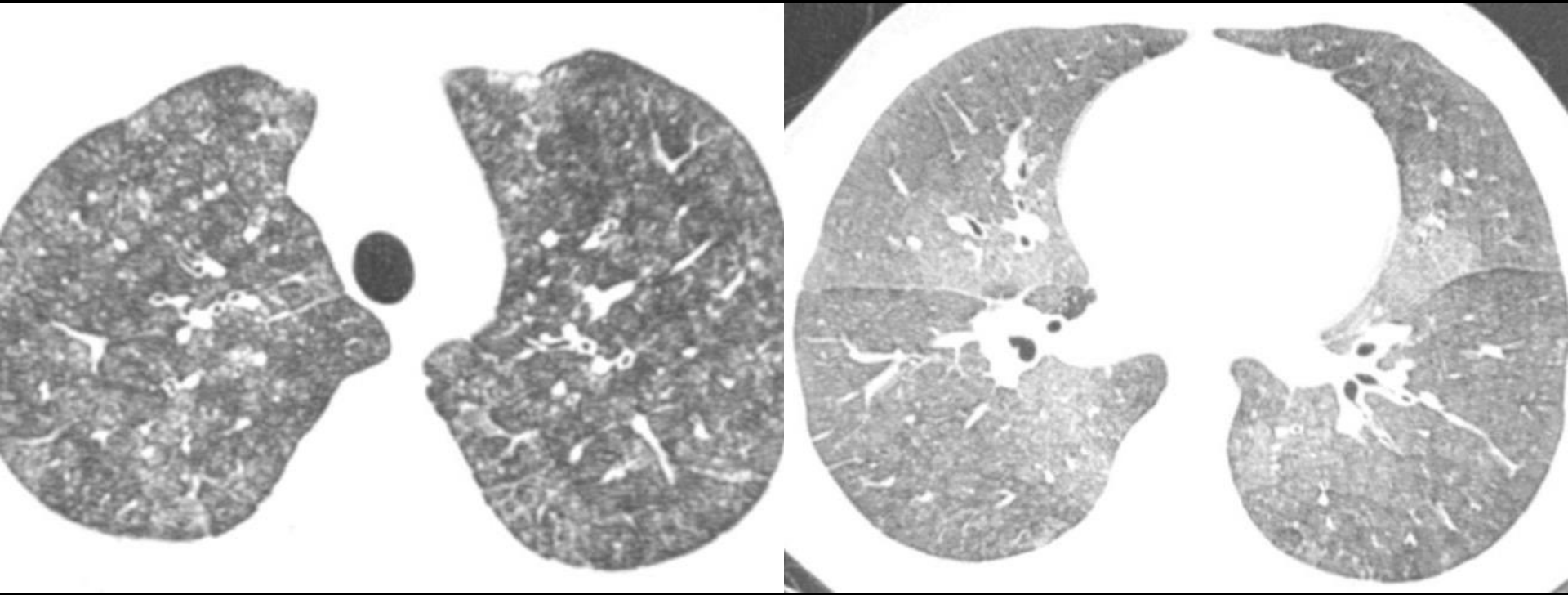
Small Vessels

- Anti-GBM antibody disease (Goodpasture syndrome)
- Primary Vasculitis
  - ANCA-associated
    - Granulomatosis with polyangiitis
    - Microscopic polyangiitis
    - Eosinophilic granulomatosis with polyangiitis (rare)
  - Behçet disease (rare)
  - Henoch-Schönlein purpura (rare)
  - Isolated pauci-immune pulmonary capillaritis

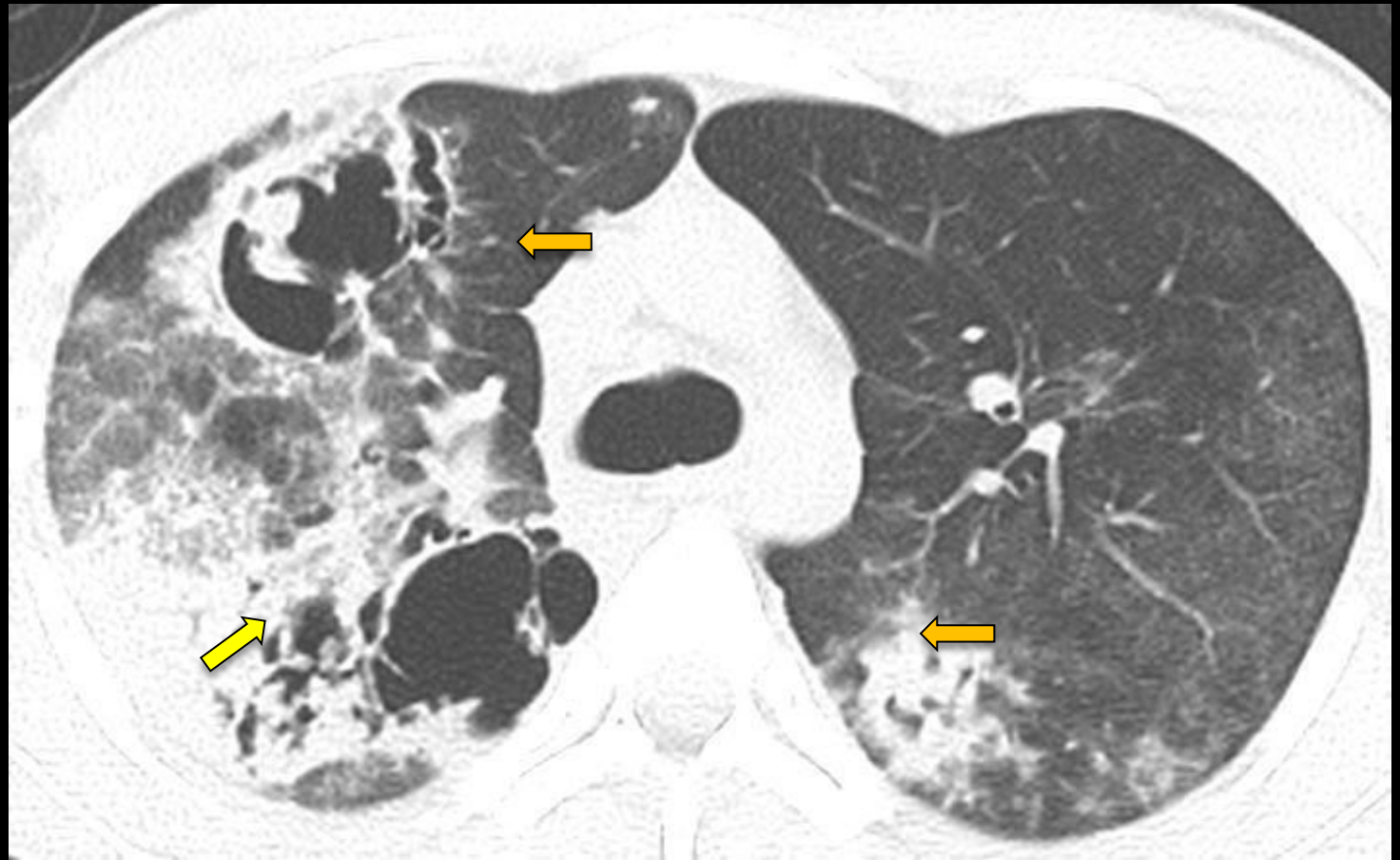
Small Vessels

- Secondary Vasculitis
  - Connective Tissue Disease
    - Systemic lupus erythematosus
    - Rheumatoid arthritis
    - Mixed connective tissue disorder
    - Polymyositis
    - Primary or secondary antiphospholipid antibody syndrome
- Drug-induced hemorrhage

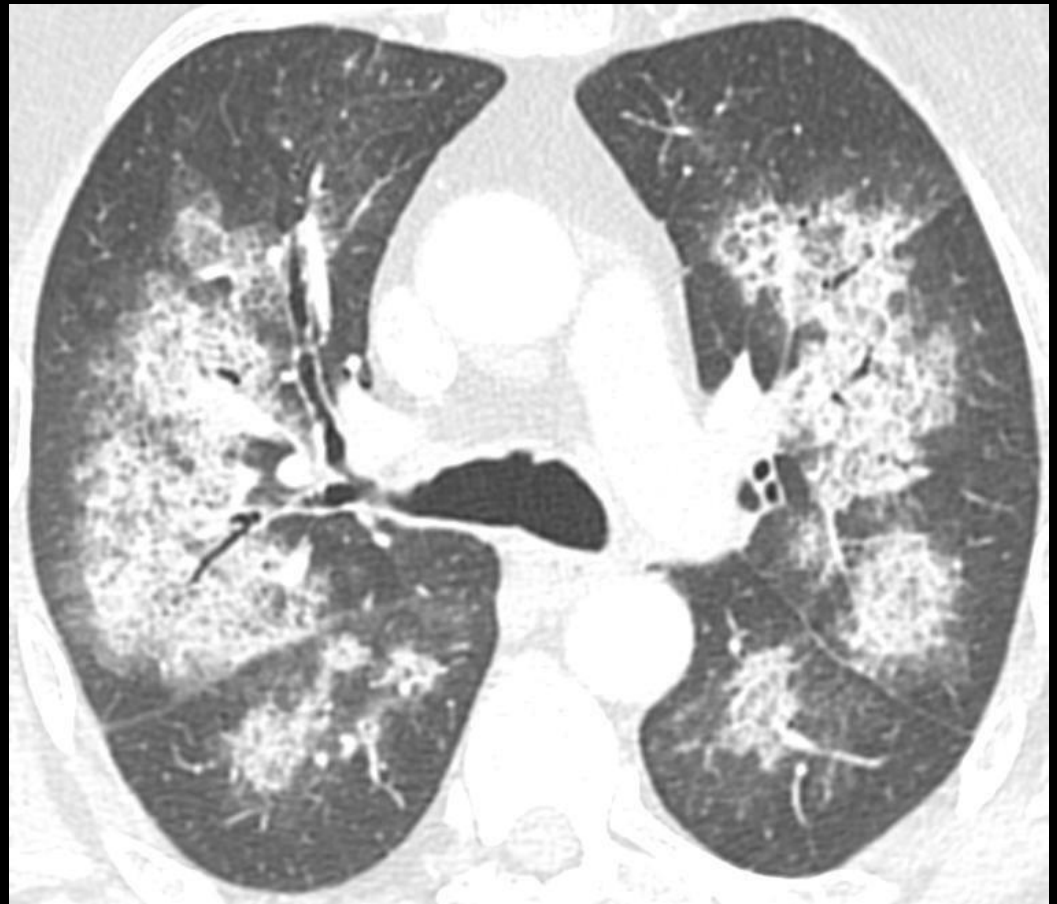
Anti-GBM Antibody Disease



Granulomatosis With Polyangiitis



Granulomatosis With Polyangiitis



Microscopic Polyangiitis

Drug-Induced Hemorrhage

Summary

- Patients with pulmonary vascular disease present with a wide range of imaging abnormalities
- Be familiar with the PH Classification
- Direct findings of large vessel disease usually apparent on CT (or MRI)
- Imaging features of small vessel disease overlap (DAH)