Hôpital Erasme

ULB

Pulmonary Hypertension :

Imaging Findings in Adult Patients

Conflict of Interest

- No conflict of interest relevant to this presentation to disclose.

Pulmonary Hypertension (PH)

- First described by E. von Romberg in 1891 as "*pulmonary vascular sclerosis*".

- After several case reports, only in 1951, after a large study by Dresdale, the term *primary pulmonary hypertension* was introduced.

Pulmonary Hypertension (PH)

- In 1973 in Geneva, a 1st classification of PH was proposed: primary and secondary PH.

- In 1998 in Evian, a new classification was established, aiming to define classes with similar pathologic findings, hemodynamic characteristics, and management.

- In 2013 in Nice, only a few changes were proposed, related to strong new evidence and the classification remained almost the same.

An Orphan Disease with a Fatal Prognosis

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- Depending on the series and causes:
  - Incidence: ~ 2.4 per million/year
  - Prevalence : ~ 15 per million/year

- Survival:
  - Three-year survival rate of idiopathic PAH: 60%

Humbert et al. Am J Respir Crit Care Med 2006
Tueller et al. Swiss Med Wkly 2008
Abenhaim et al. N Engl J Med 1996
Peacock. Eur Respir J 2007
Hachulla. Arthritis Rheum 2009
Condliffe et al. Am J Respir Crit Care Med 2009
Hachulla et al. Arthritis and Rheumatism 2005
Humbert et al. Circulation 2010
Benza et al. Circulation 2010

Key Points

- Definitions
- Classification (Nice 2013): 5 groups
- Imaging findings (X-ray and CT) common to all groups
- Clinical, pathology, and imaging for each group

Definitions

- PH is a haemodynamic and pathophysiological condition defined as an increase in mean pulmonary arterial pressure (PAP) ≥ 25 mmHg at rest as assessed by right heart catheterization.

- PH can be found in multiple clinical conditions: 5 Groups

1. Pulmonary arterial hypertension (PAH)
2. Pulmonary hypertension due to cardiac disease
3. Pulmonary hypertension due to chronic lung disease and/or hypoxia
4. Chronic thromboembolic pulmonary hypertension (CTEPH)
5. Pulmonary hypertension due to unclear multifactorial mechanisms

Simonneau et al. JACC 2013

Hemodynamic Definitions (Nice)

Definition	Characteristics	Clinical group(s)
Pulmonary hypertension (PH)	Mean PAP ≥ 25 mmHg	All
Pre-capillary PH	Mean PAP ≥ 25 mmHg	1. Pulmonary arterial hypertension
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Post-capillary PH	Mean PAP ≥ 25 mmHg	2. PH due to left heart disease
	PWP > 15 mmHg	5. PH with unclear and/or multifactorial mechanisms
	CO normal or reduced	
Passive	TPG ≤ 12 mmHg	
Reactive (out of proportion)	TPG > 12 mmHg	

PAP = pulmonary arterial pressure, PWP = pulmonary wedge pressure, CO = cardiac output,

TPG = Transpulmonary pressure gradient

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Imaging

- Imaging findings common to all groups

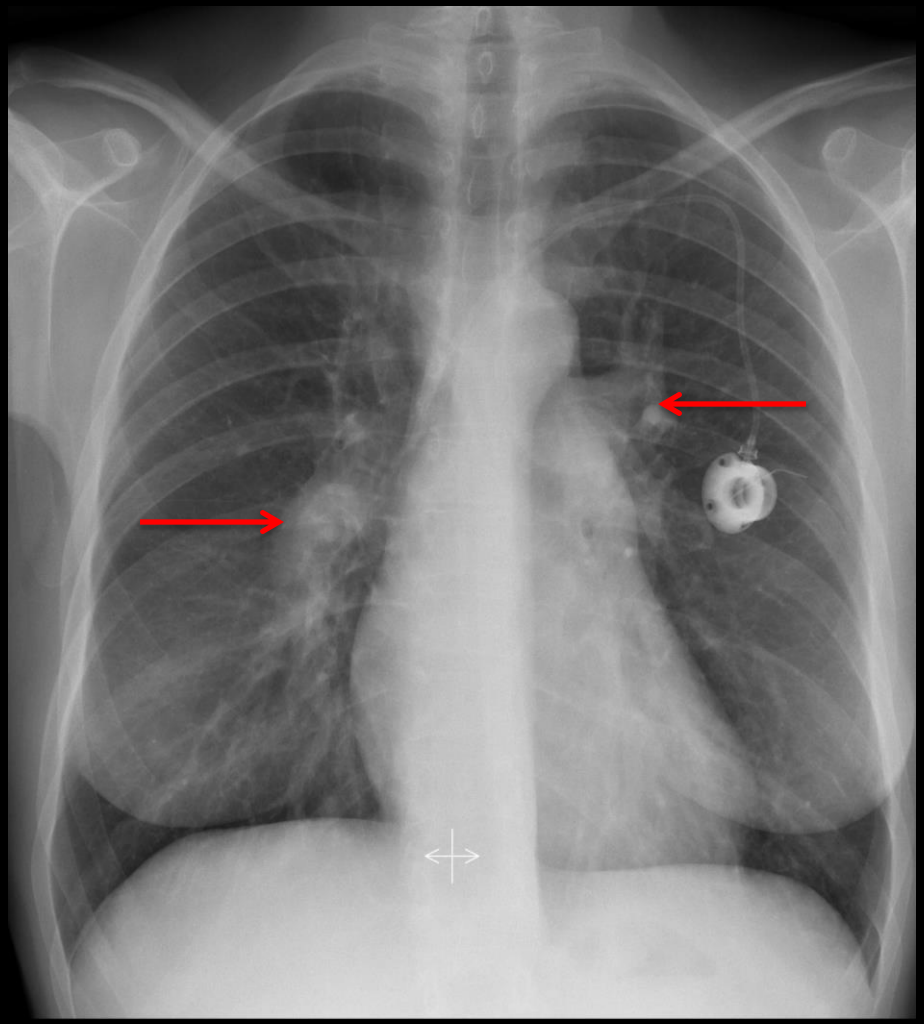
- Specific imaging findings for each group
  - Relevant information about pathology, pathophysiology, and prognosis

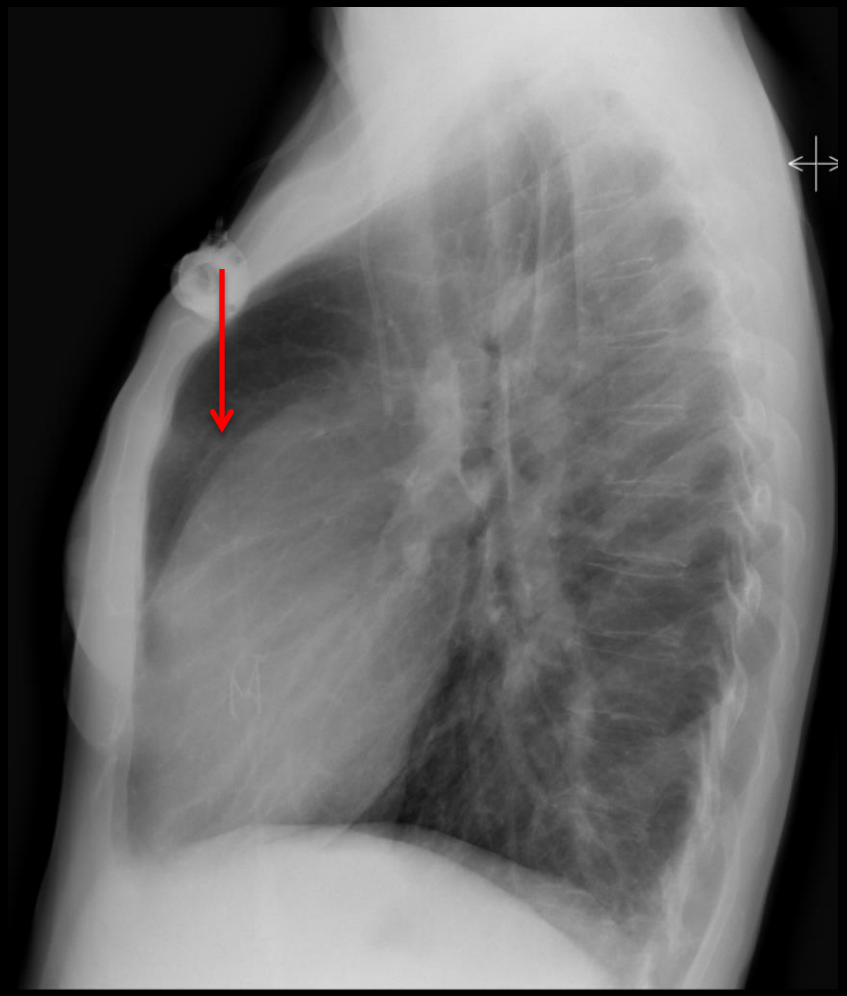
X-Ray Features Common to All Groups

- Normal
- Bulging of the pulmonary artery
- Dilatation of the interlobar artery > 16 mm
- Disparity of proximal-distal vessels: dilatation of central branches of pulmonary arteries with peripheral pruning
- Filling of the retro-sternal space due to RV dilatation
- Increased cardio-thoracic index: usually associated with right heart failure

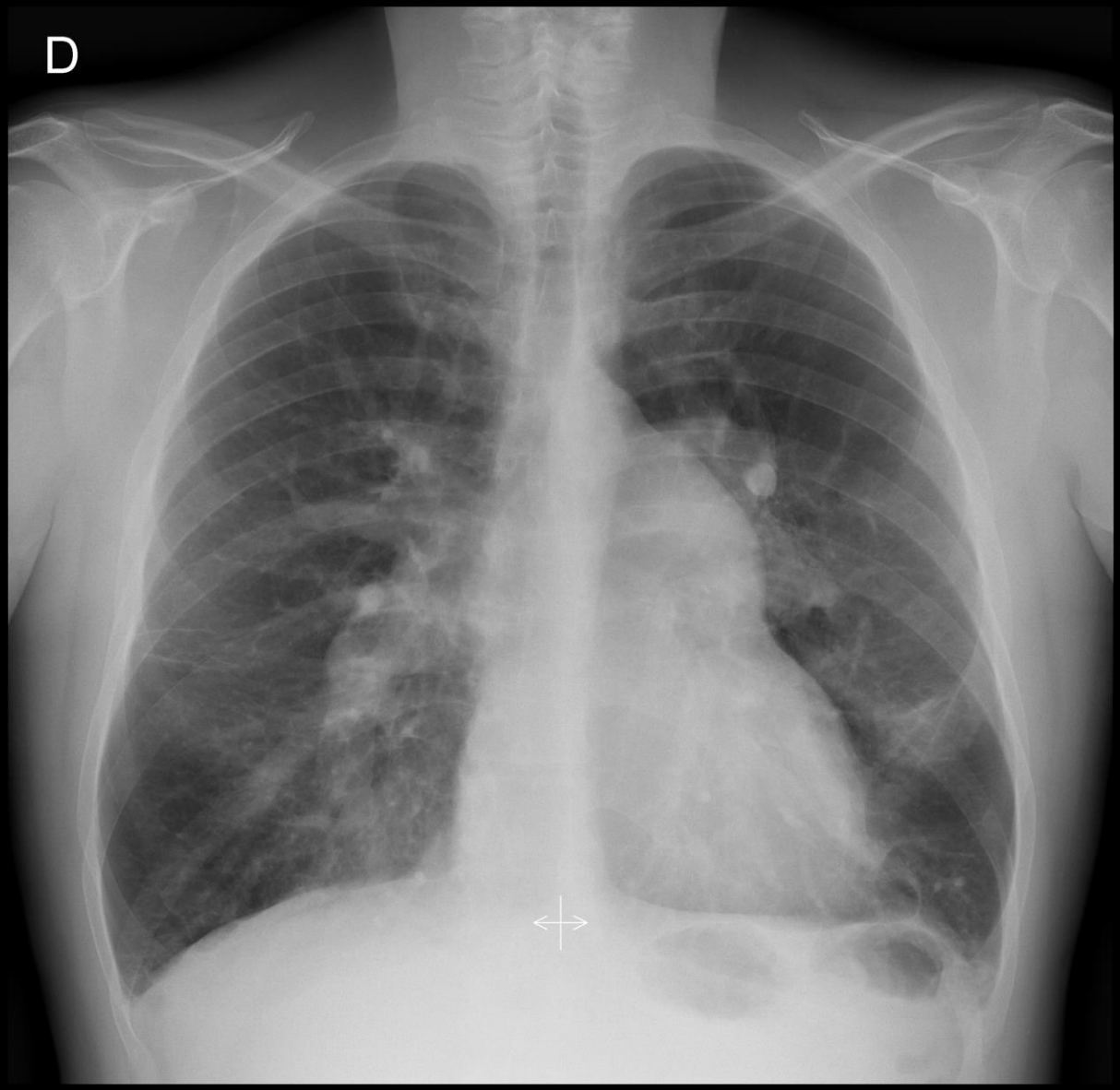
Kanemoto et al. Chest 1979

X-Ray Features Common to All Groups





X-Ray Features Common to All Groups



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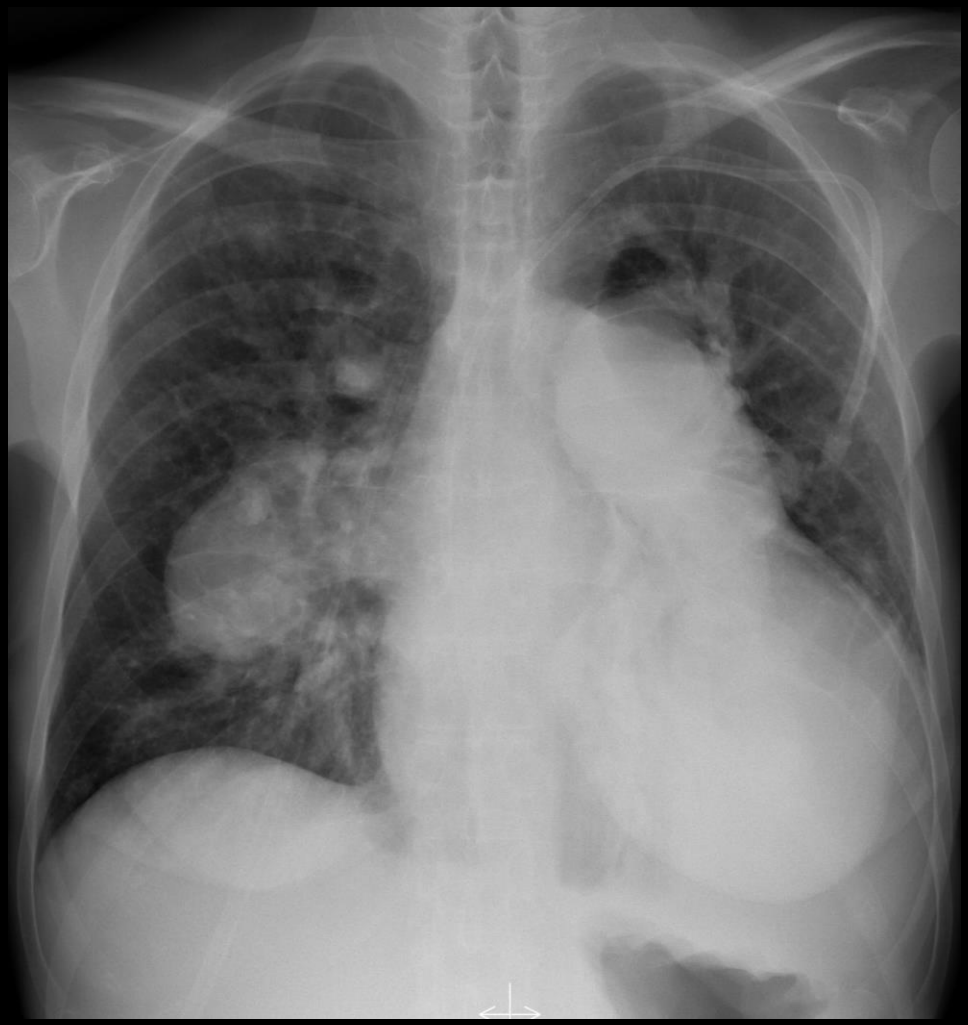
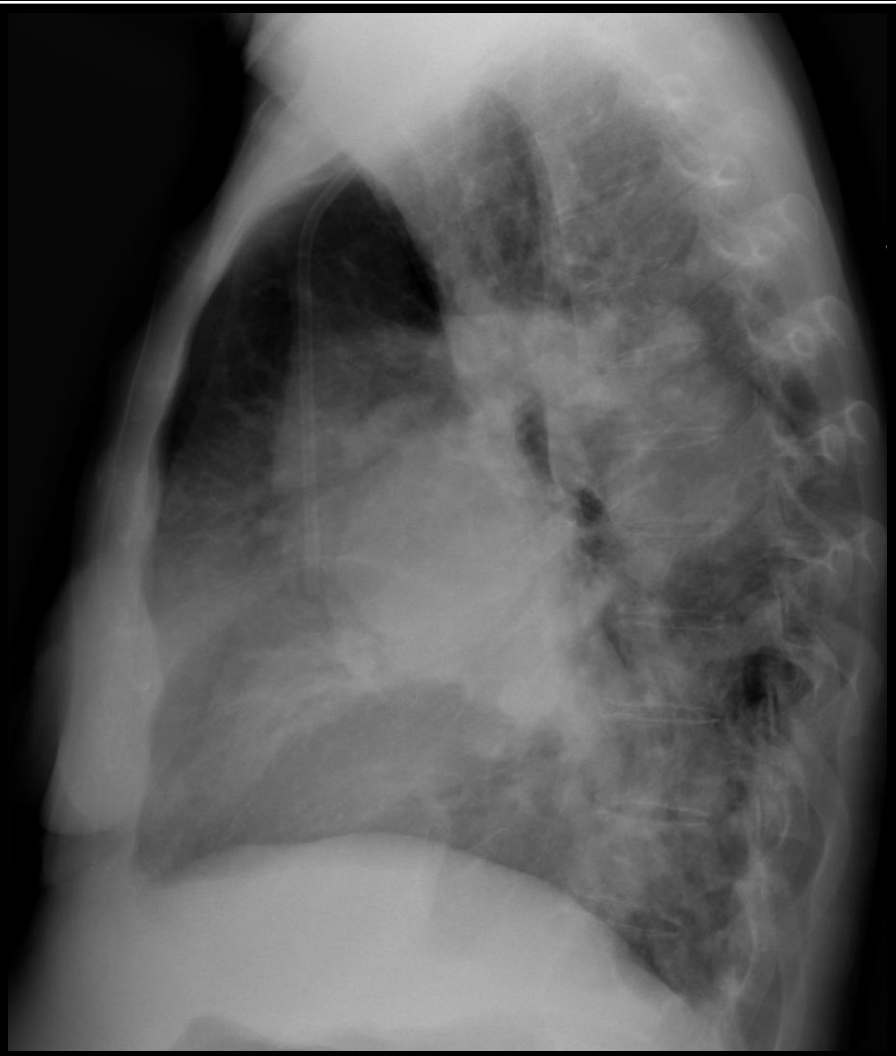


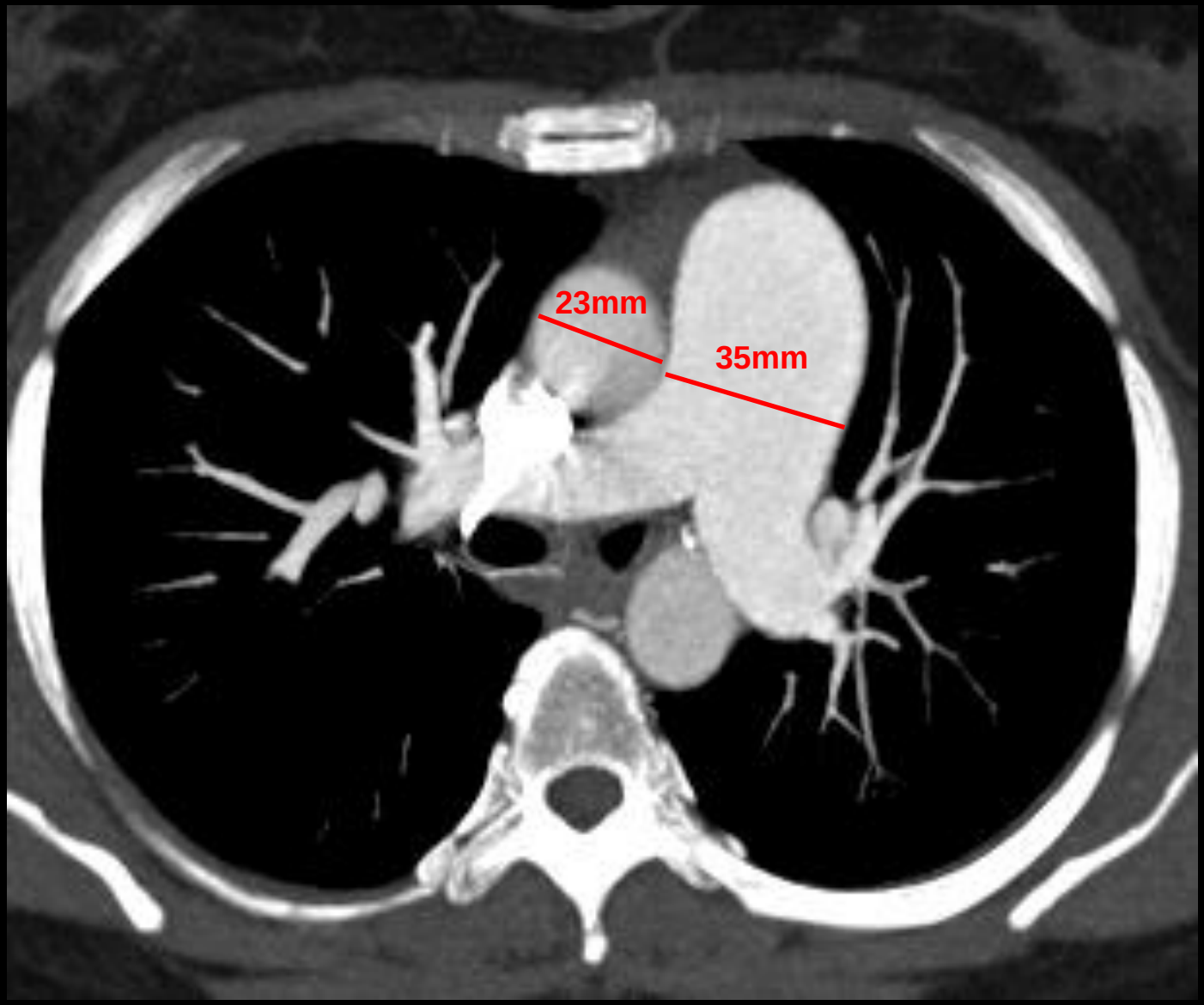


CT Features Common to All Groups

- Dilation of the main pulmonary artery
  - Diameter of the distal main pulmonary artery (within 3cm of its bifurcation) ≥ 29 mm (Se = 87%, Sp = 89%, VPP = 97%)*
  - Diameter of the main pulmonary artery (close to its bifurcation) ≥ 3.32 cm (Se = 58%, Sp = 95%)**
  - Diameter of the distal main pulmonary artery diameter > diameter of the ascending aorta on the same CT section (Sp = 92%, VPP = 96%, VPN = 52%)***

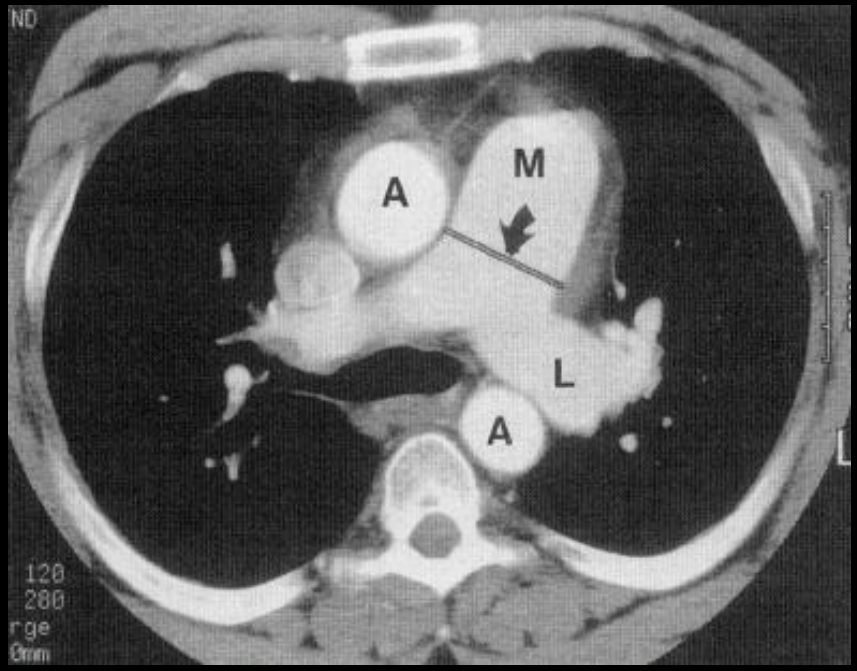
*Tan et al. Chest 1998
**Edwards et al. Br J Radiol 1998
***Ng et al. J Thorac Imaging 1999

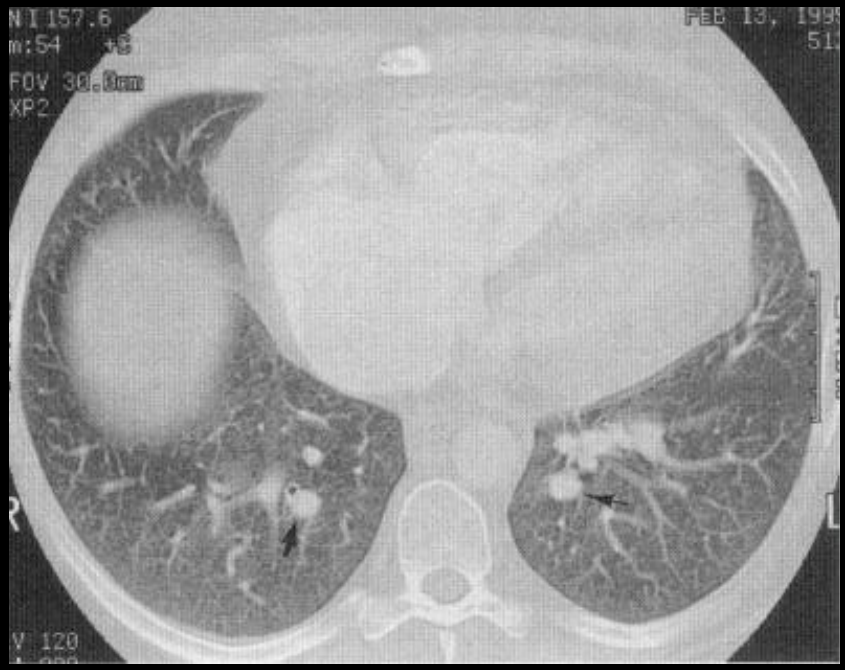
CT Features Common to All Groups



CT Features Common to All Groups

- Dilation of main and segmental pulmonary arteries
  - Diameter of the distal main pulmonary artery ≥ 29 mm and segmental artery-to-bronchus ratio > 1:1 in 3 of 4 pulmonary lobes (Sp = 100%).

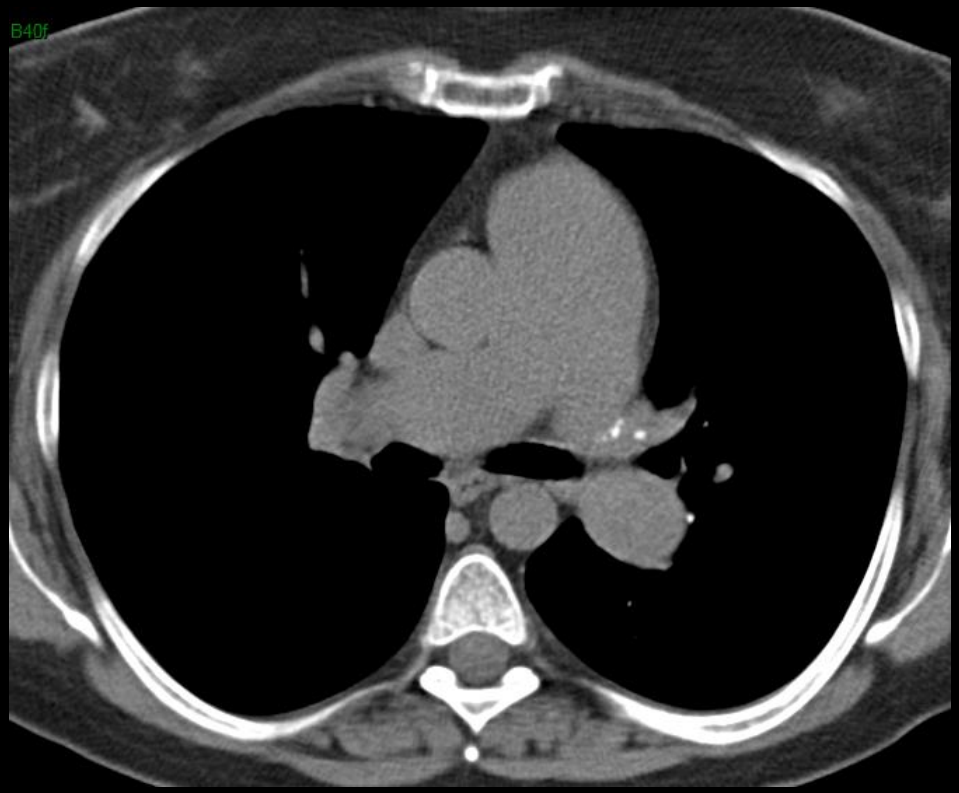


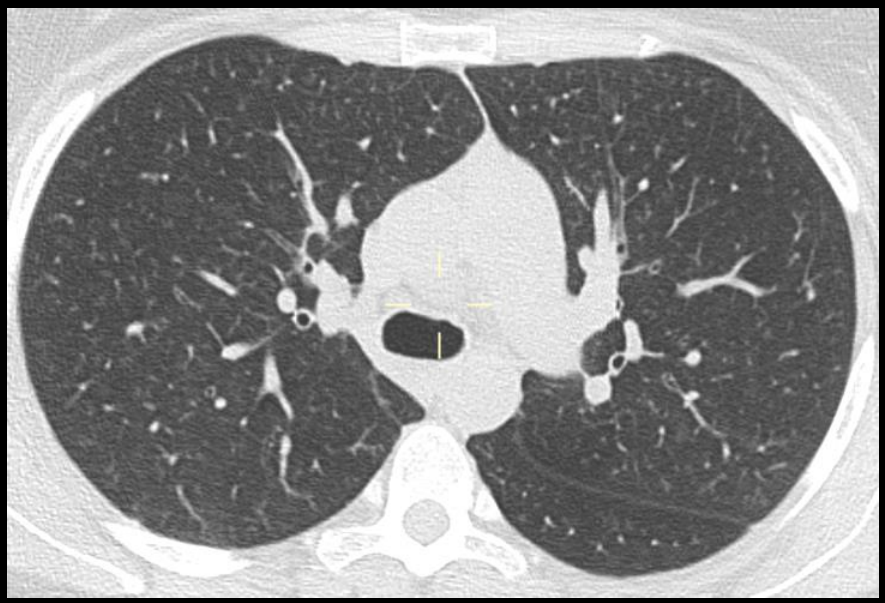


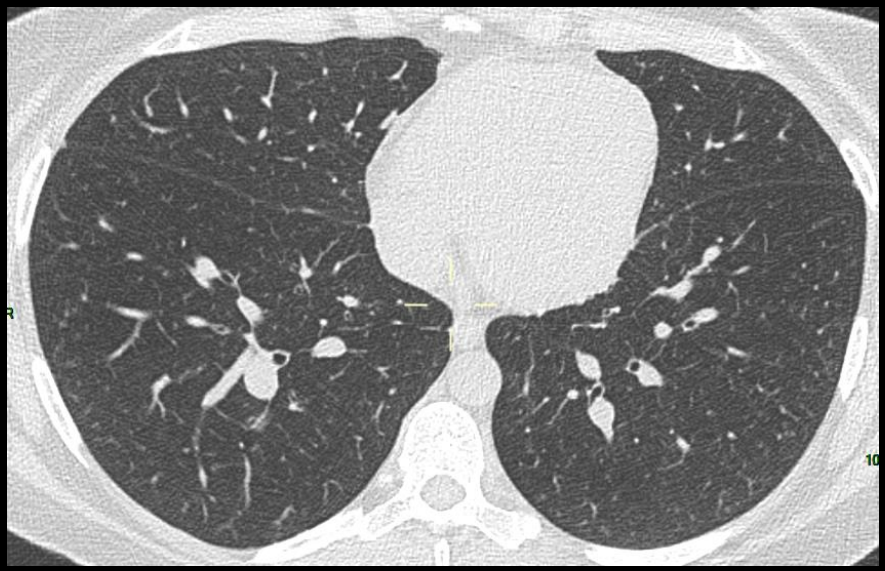
MPAD of 40mm (arrow) in a 35-year-old man with severe primary pulmonary hypertension. A = aorta; M = main pulmonary artery; L = left pulmonary artery. Artery to bronchus ratio (ABR) > 2:1 in the posterior basal segments of the lower lobes in the same patient (arrows)

Tan et al. Chest 1998

CT Features Common to All Groups







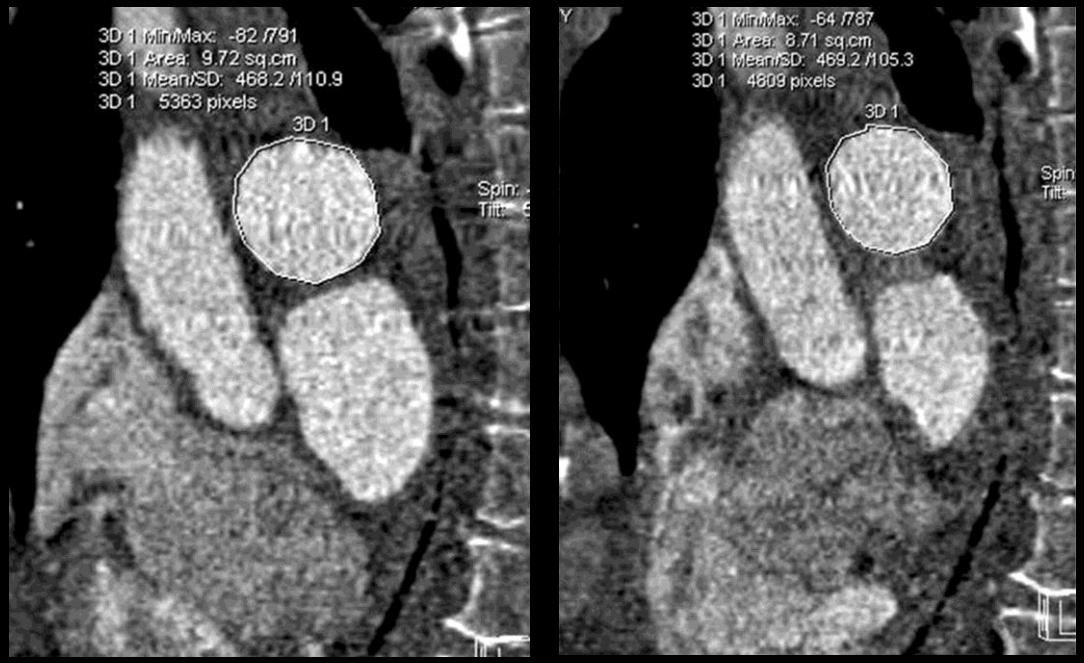
CT Features Common to All Groups

- Dilation of the main pulmonary artery
  - Healthy individuals have occasionally idiopathic dilatation of pulmonary arteries.*
  - A no-dilated main pulmonary artery, by reference to the aorta, does not exclude HP.

*Ring et al Br J Radiol 2002

CT Features Common to All Groups

- Compliance of the main pulmonary artery
  - ECG-gated MDCT study : strong negative correlation (r_s = -.79) between the right pulmonary artery wall distensibility (calculated by the change in cross sectional area between diastole and systole) and the mean pulmonary artery pressure (cutoff = 16.5%; Se= 86%, Sp = 96%).

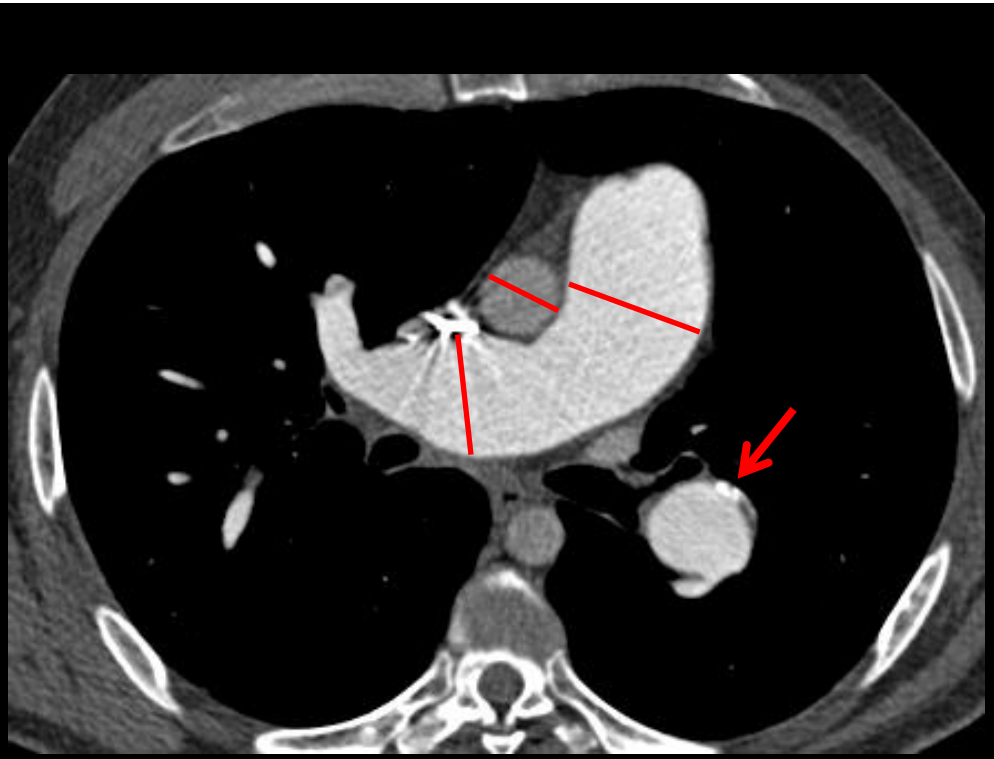


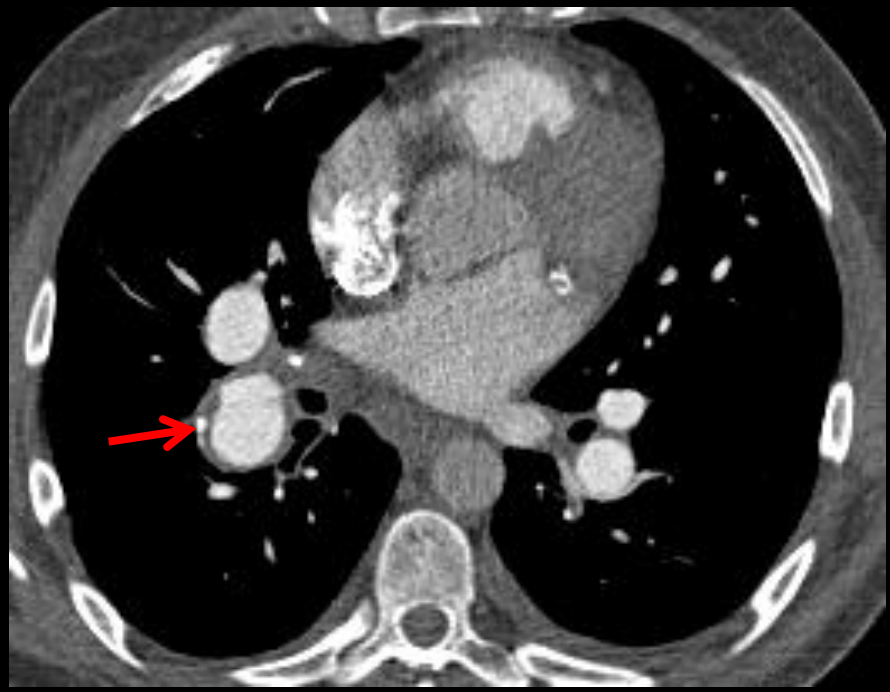
Assessment of RPA distensibility in 65-year-old man with primary PHT (mean PAP, 43mmHg). RPA cross-sectional area was 9.7cm² at **(a)** 20% of the R–R interval and 8.7cm² at **(b)** 80% of the R–R interval. RPA distensibility was only 10%

Revel et al. Radiology 2009

CT Features Common to All Groups

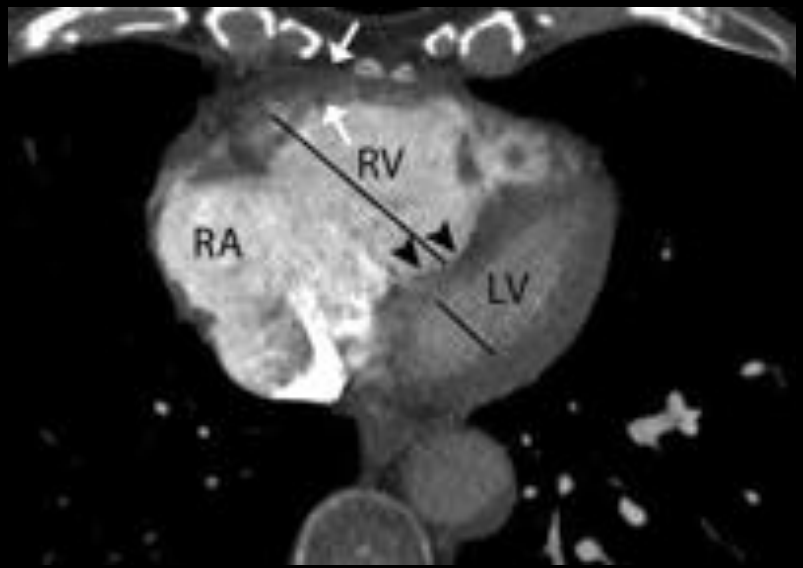
- Dilation of the right and left main pulmonary arteries
- Abrupt narrowing and tapering of the peripheral arteries
- Atherosclerosis of the pulmonary arteries (red arrows)





CT Features Common to All Groups

- Right ventricular hypertrophy
- Right ventricular and atrial enlargement with inversion of the interventricular septa



Right heart abnormalities secondary to chronic thromboembolic pulmonary hypertension in a 47-year-old man. Axial contrast-enhanced CT scan shows dilatation of the right ventricle *(RV)*, with a ratio of more than 1:1 between the right and left ventricle *(LV)* diameters (lines); leftward septal bowing (arrowheads); thickening of the free right ventricular wall (arrows); and dilatation of the right atrium *(RA)*.

Castaner et al. Radiographics 2009

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Simonneau et al. JACC 2013

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Simonneau et al. JACC 2013

1: Pulmonary Arterial Hypertension (PAH)

Hôpital Erasme | ULB

1.1. Idiopathic: Neither family history nor identified risk factor, young adulthood (20-45 years); female predominance (3:1); poor prognosis (5yr-surv.=34%)

1.2. Heritable: BMPR2 mutation in 70% of cases

1.3. Drugs and toxins induced (including appetite suppressants)

1.4. Associated with
- 1.4.1. Connective vascular disease
- 1.4.2. HIV infection
- 1.4.3. Portal hypertension
- 1.4.4. Congenital heart disease
- 1.4.5. Schistosomiasis

1'. Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)

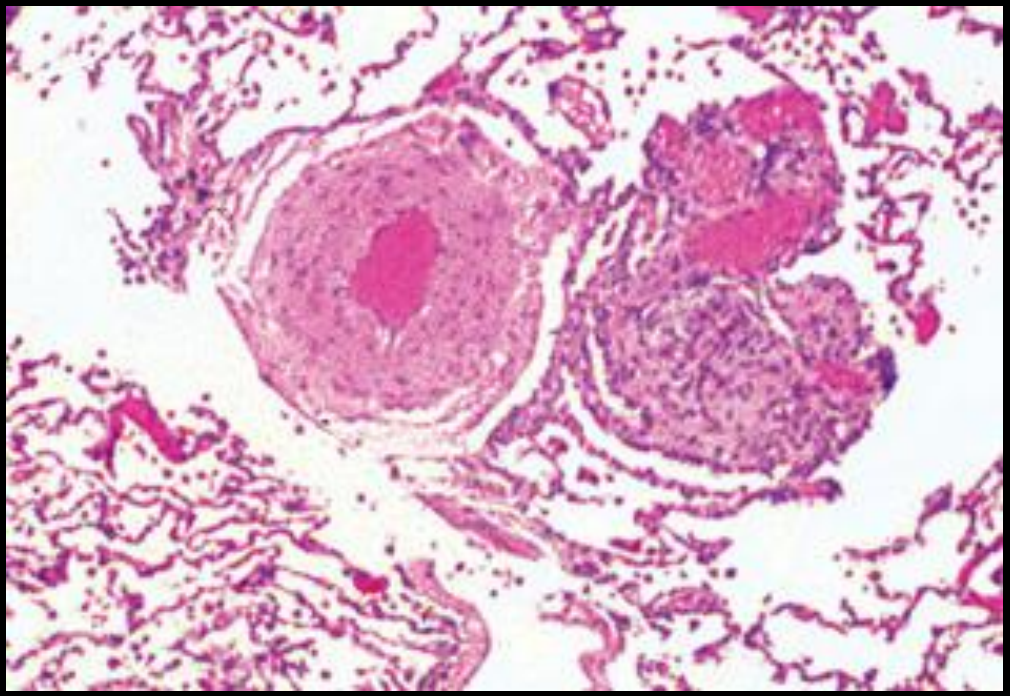
Simonneau et al. JACC 2013

1: PAH: Pathology

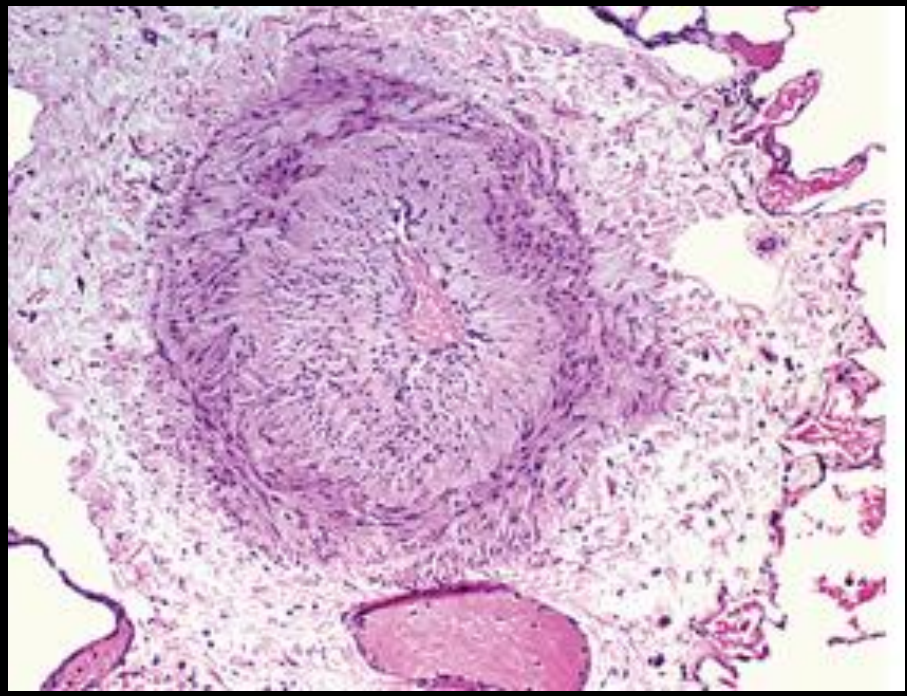
- Distal pulmonary arteries (< 500 microns of diameter)
- Medial hypertrophy, intimal fibrosis
- Plexiform lesions
  - Multiple small vascular channels arising within a small artery
  - Illustrious but not pathognomonic for PAH
  - Focal intimal thickening often beyond branching points, endothelial cells' proliferation leading to capillary-like, sinusoidal channels on a smooth muscle cell and collagen-rich matrix within the lumen resulting in obstruction
  - Draining in dilated veins: hemorrhage can happen due to unstable aneurysm-like vessel wall structure
  - Perivascular inflammation (T-cells, macrophage, mast cells) and rarely classical arteritis

Peacock, Naeije, Rubin. Pulmonary Circulation

1: PAH: Pathology



Small pulmonary artery shows plexiform lesion on the *right* and medial thickening on the *left*



Intimal and medial thickening of a small pulmonary artery

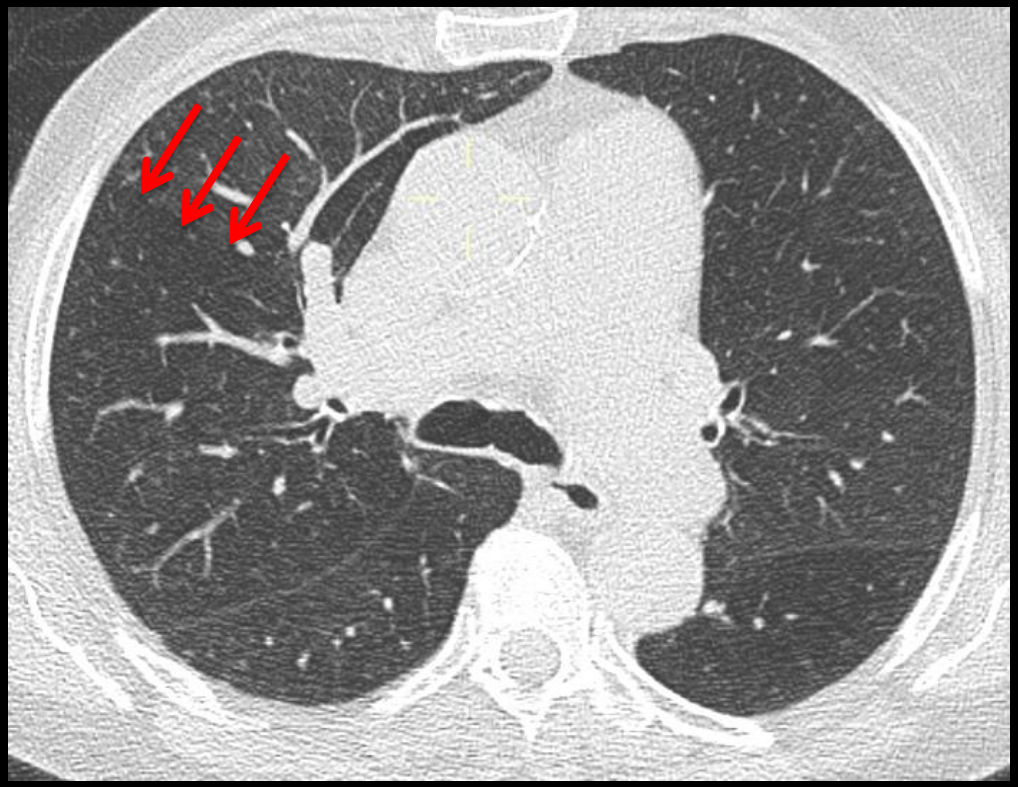
Cagle PT et al. Color Atlas and Text of Pulmonary Pathology. 2007

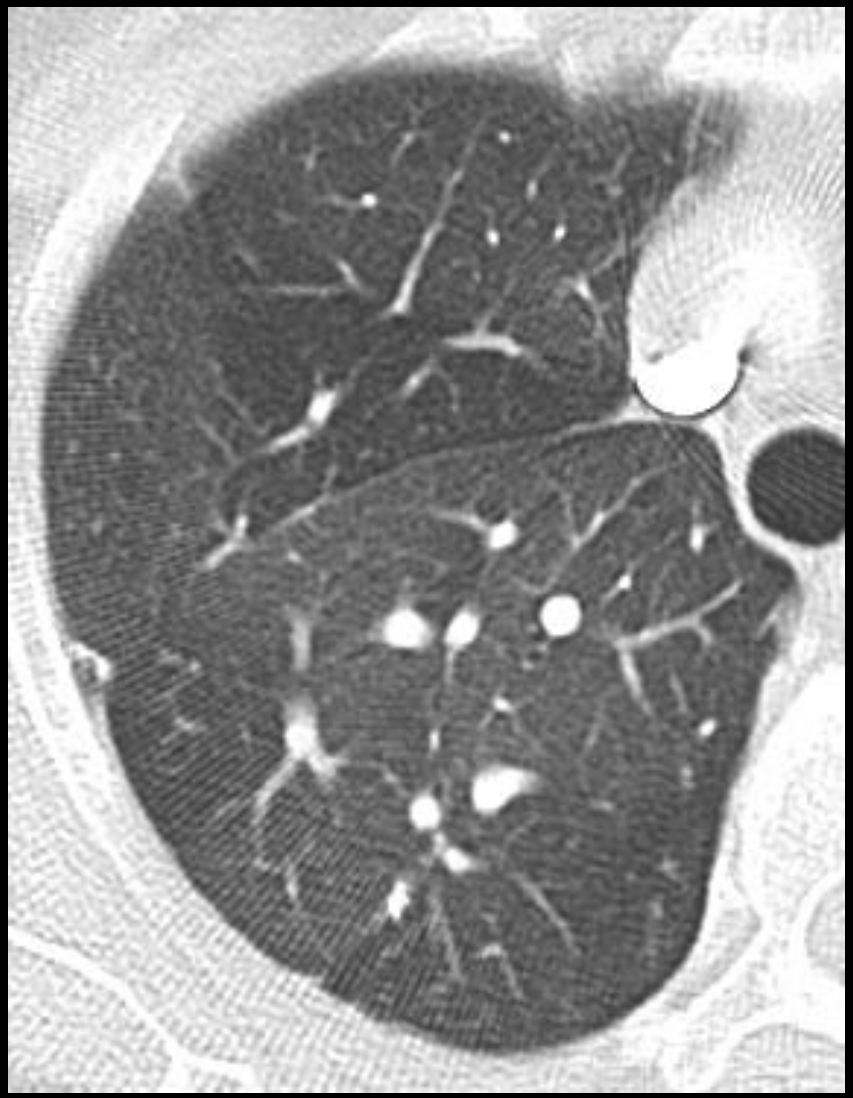
1: PAH: CT Features

- Mosaic attenuation
  - Sharply demarcated areas of heterogeneous attenuation in the lung parenchyma that predominantly conform to the borders of the secondary lobule
  - Mostly observed in PH of vascular etiology
  - 74% of patients with PAH (may be absent)

Sherrick et al. AJR;1997

1: PAH: Mosaic Attenuation



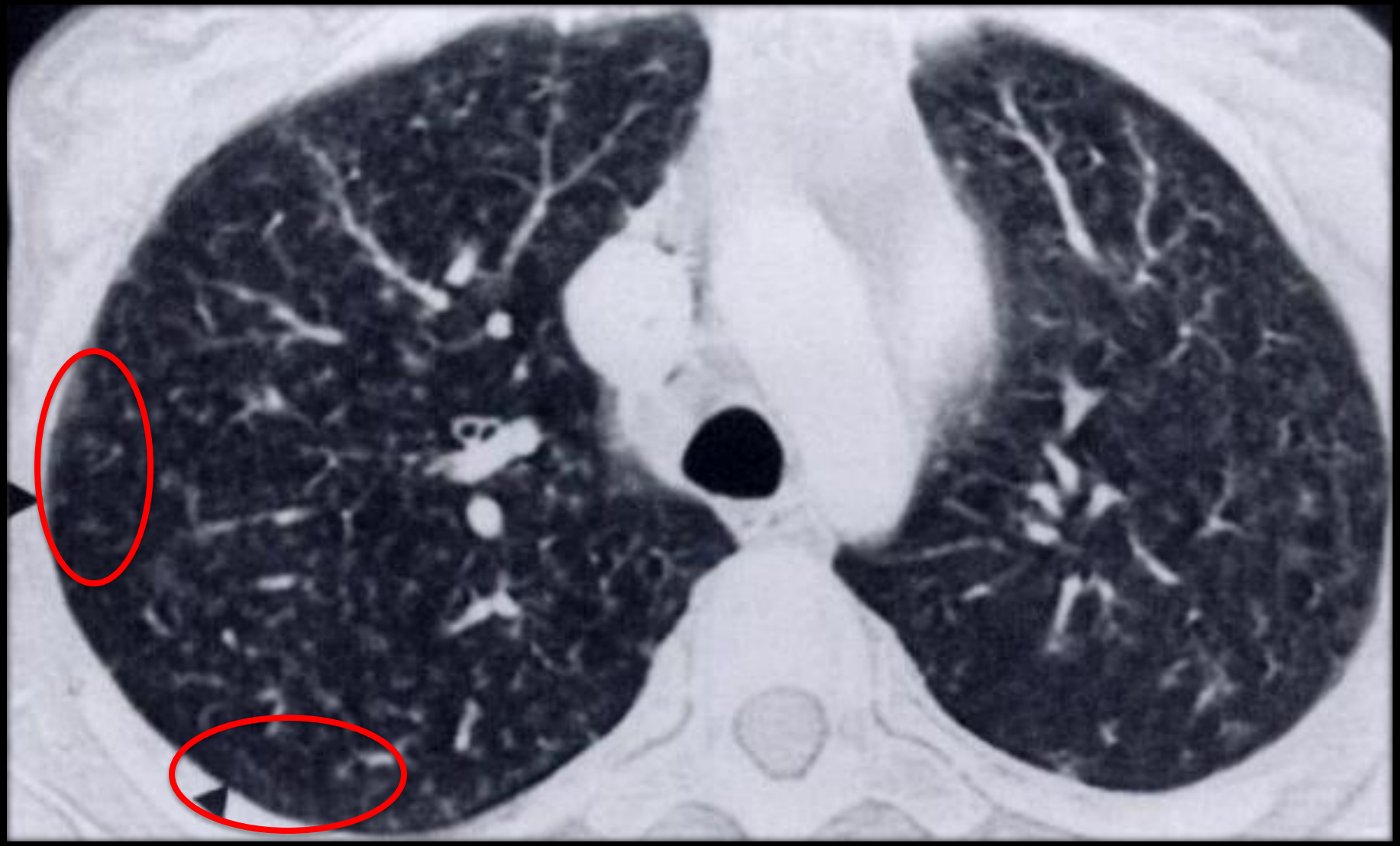


1: PAH: CT Features

- Ground-glass nodules
  - <3 mm in diameter with centro-lobular distribution
  - Interstitial and alveolar stellate fibrotic lesions containing cholesterol clefts and, multinucleated giant cells and lymphocytes
  - Attributed either to degradation of excess surfactant or recurrent pulmonary hemorrhage

Nolan et al. AJR 1999

1: PAH: Ground Glass Nodules

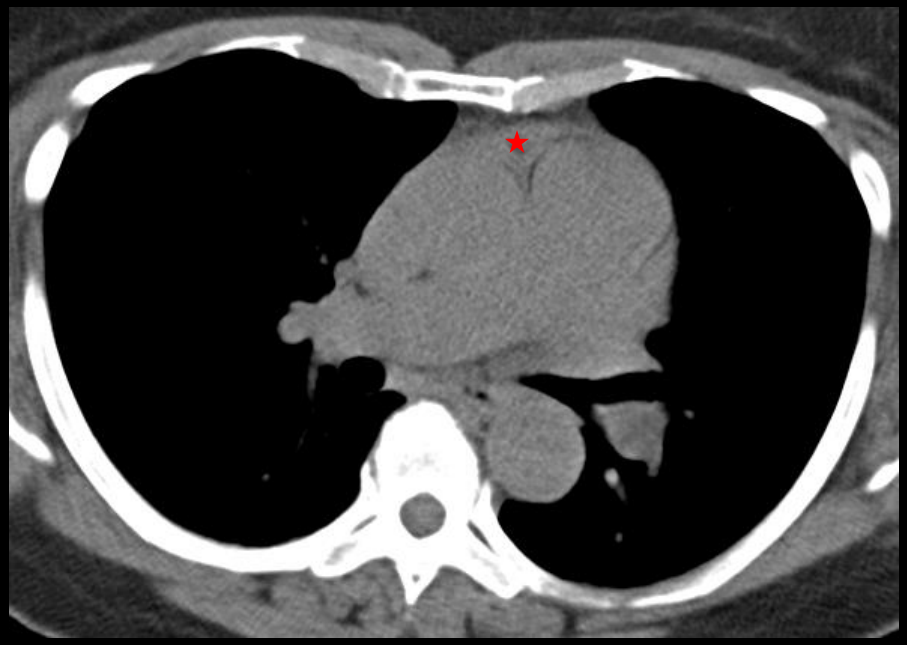


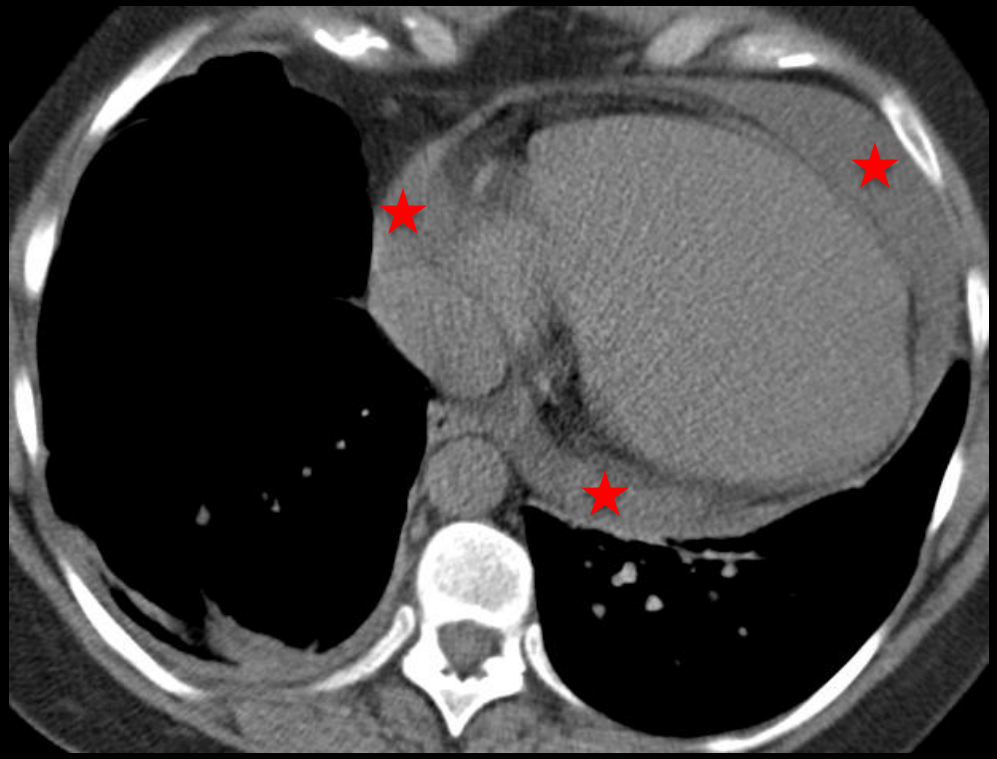
1: PAH: CT Features

- Pericardial effusion
  - Related to poor outcome; triple 1-year mortality
  - Refractory to diuretics
  - Usually located in the anterior pericardial recess
  - Correlated with right atrial size, displacement of IVS and a tendency to greater ventricular size
  - Probably a manifestation of right heart failure: result of impaired lymphatic and venous drainage due to elevated right atrial pressure

Hinderliter et al. 1999

1: PAH: Pericardial Effusion





1: PAH: CT Features

- Mediastinal lymph nodes
  - 20% of patients
  - Usually related to pleural effusion
  - < 20 mm

T Moua et al. Chest 2013

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Hôpital Erasme | ULB

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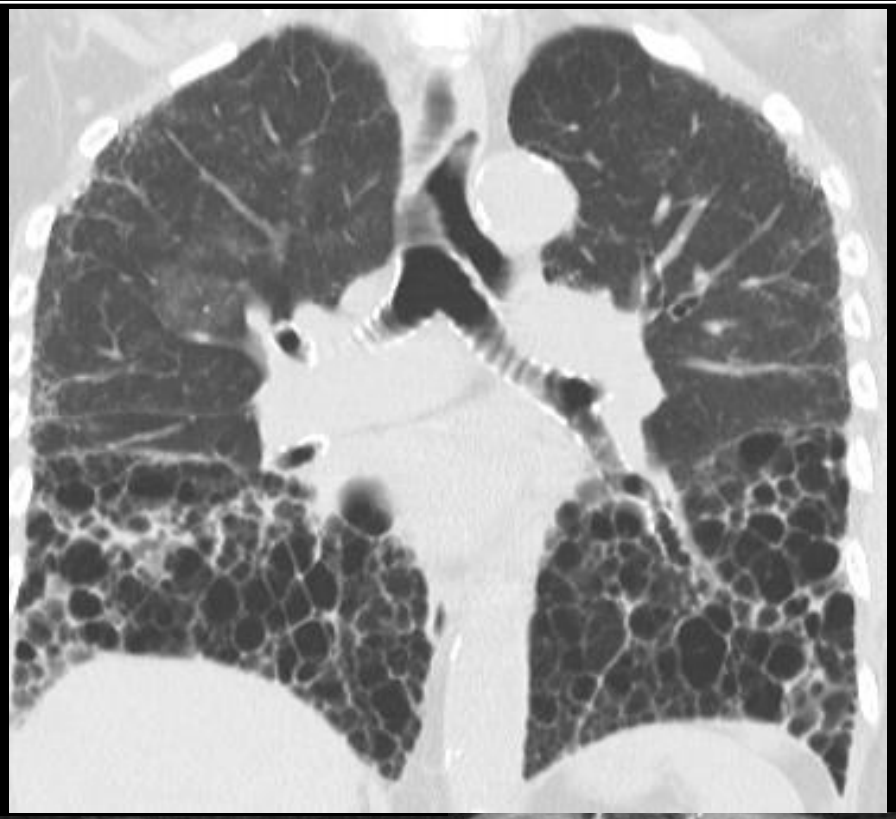
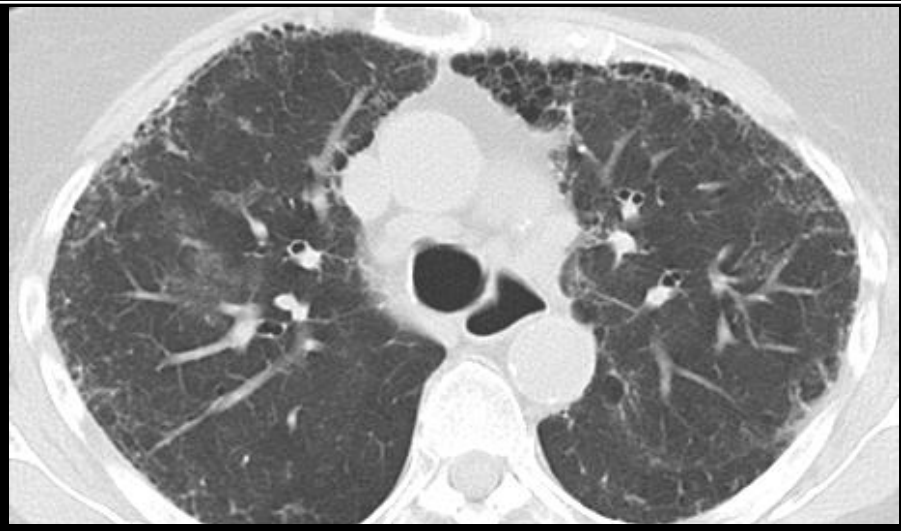
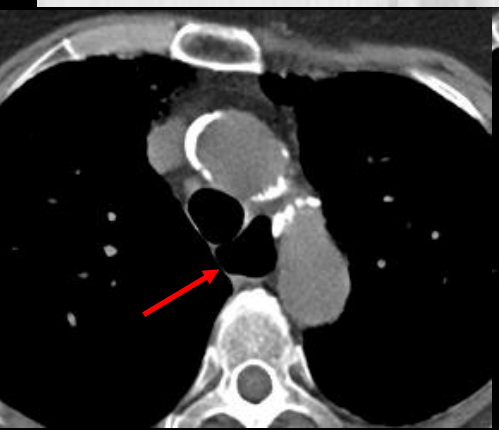
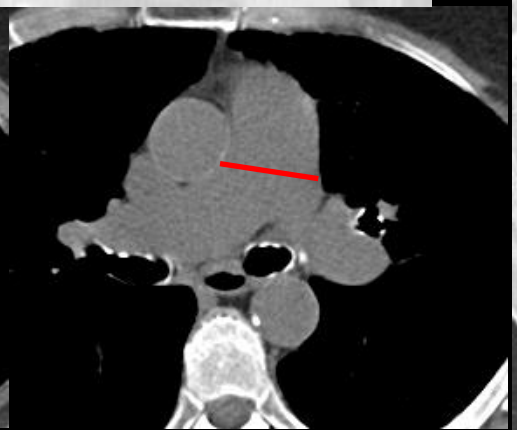
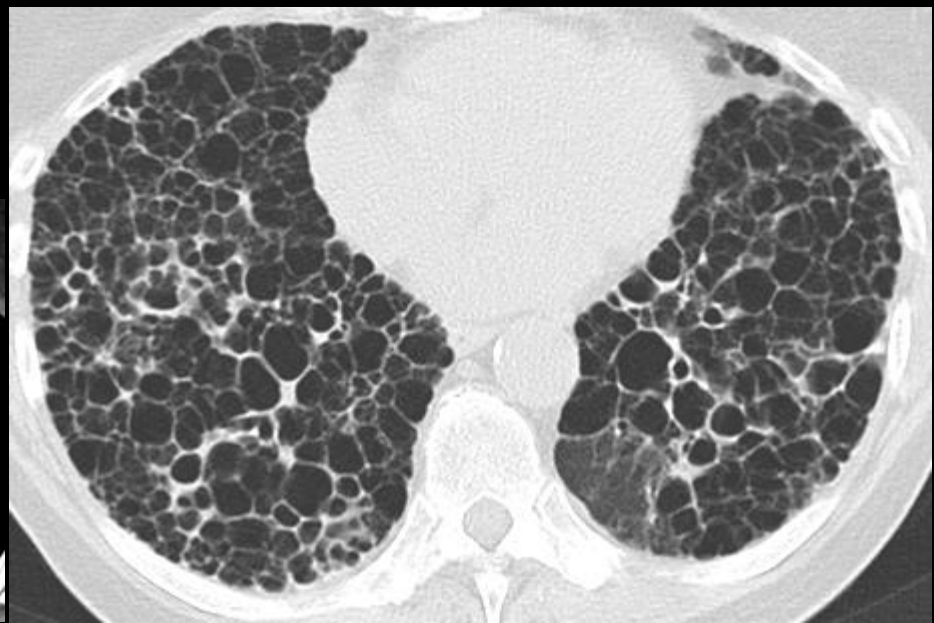




Systemic Sclerosis







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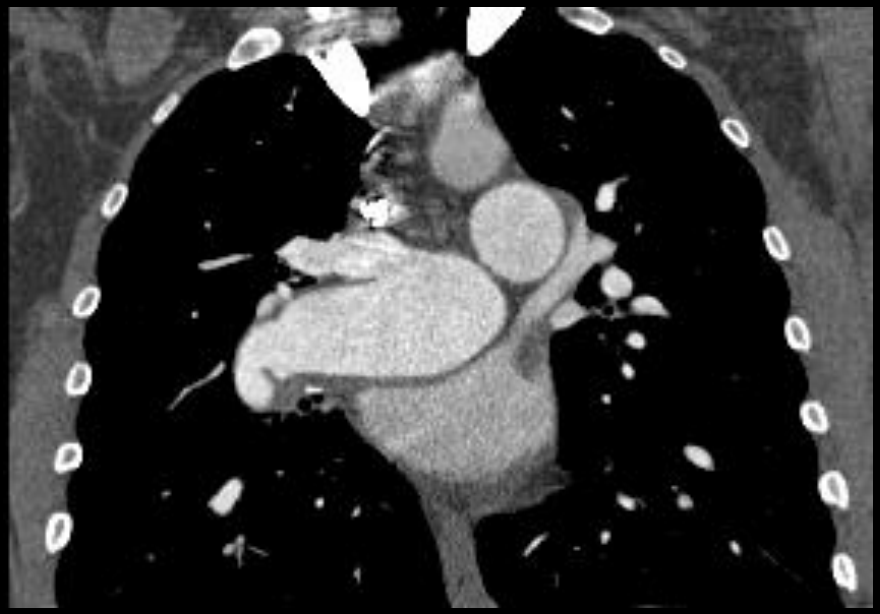
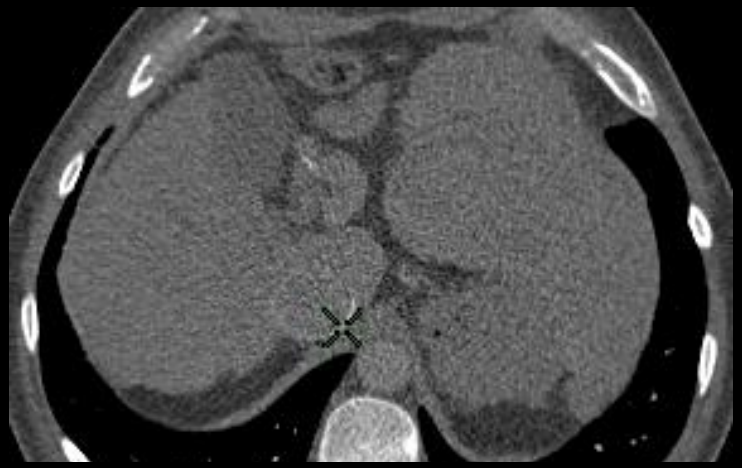
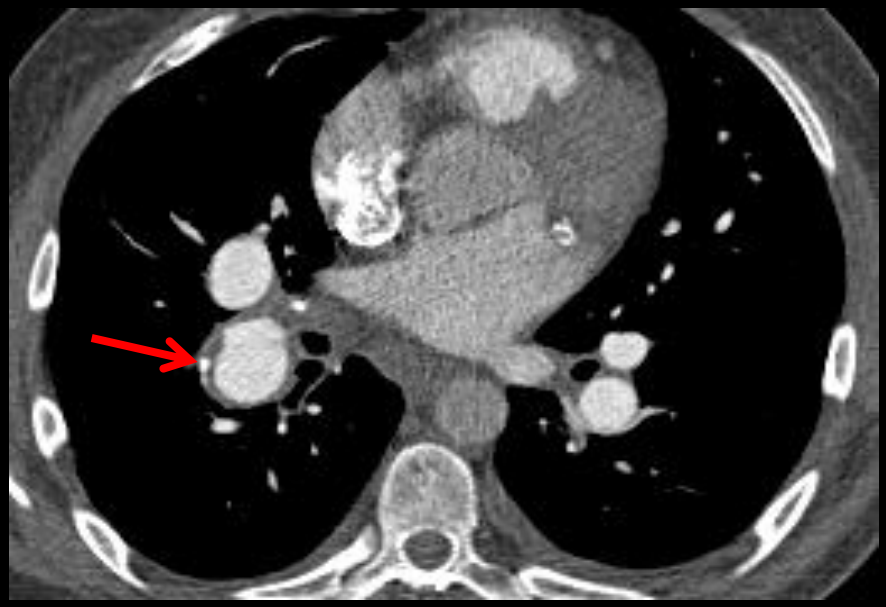
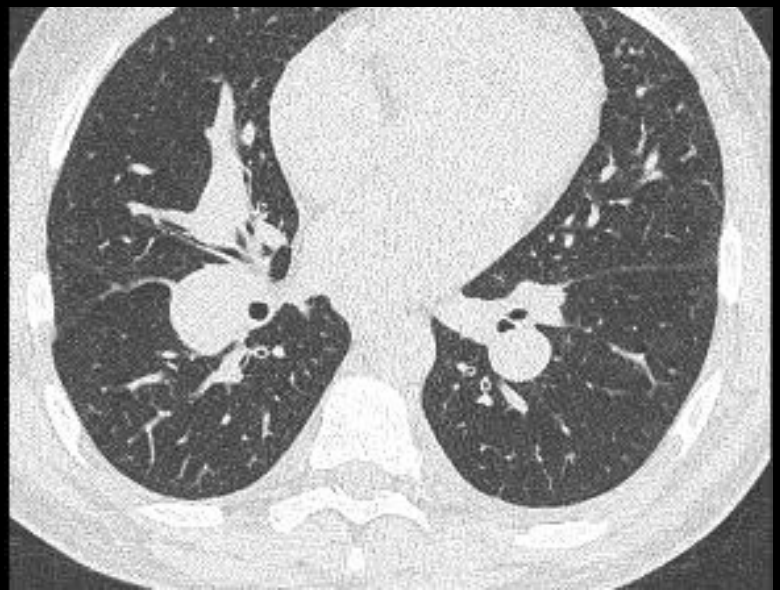




PAH associated with Liver Cirrhosis





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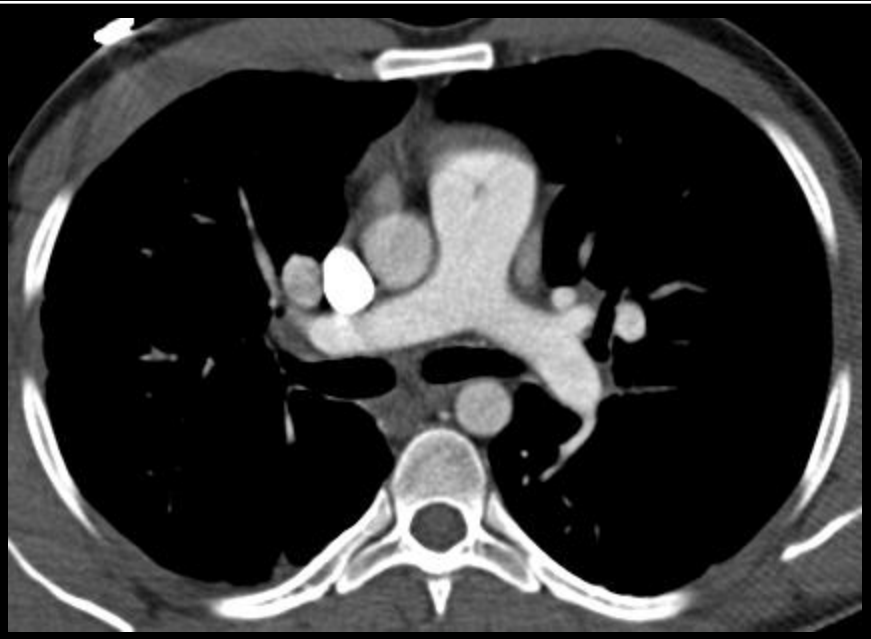

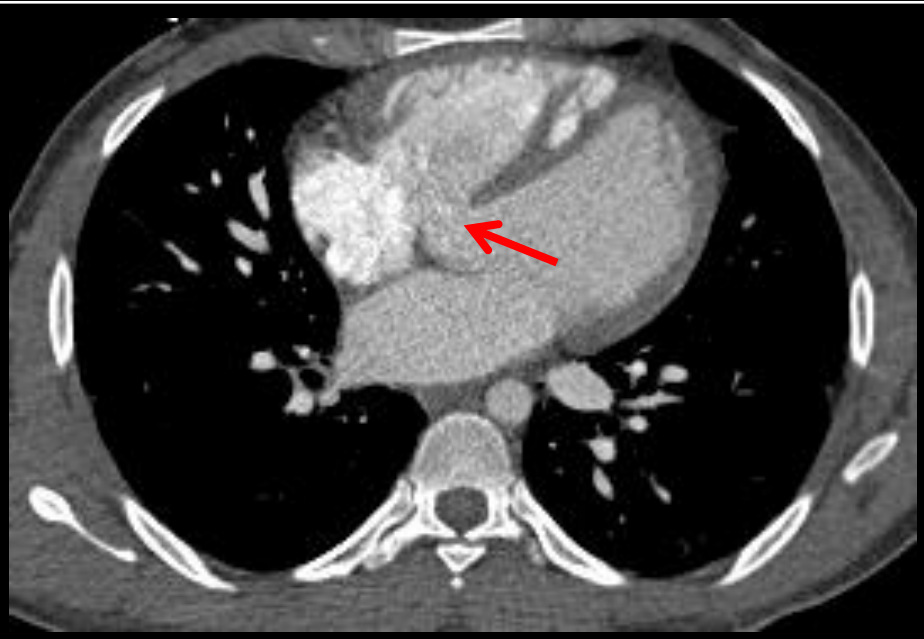

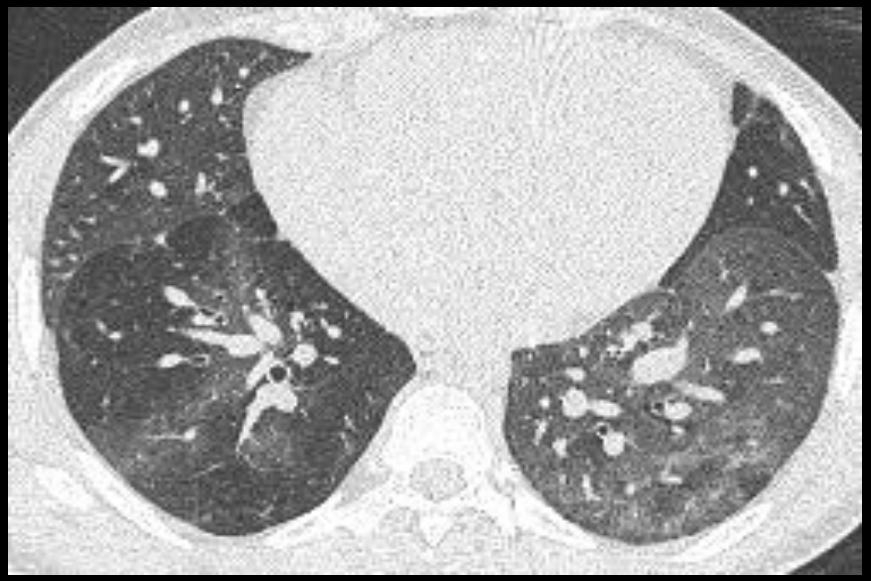

PAH associated with ventricular septal defect

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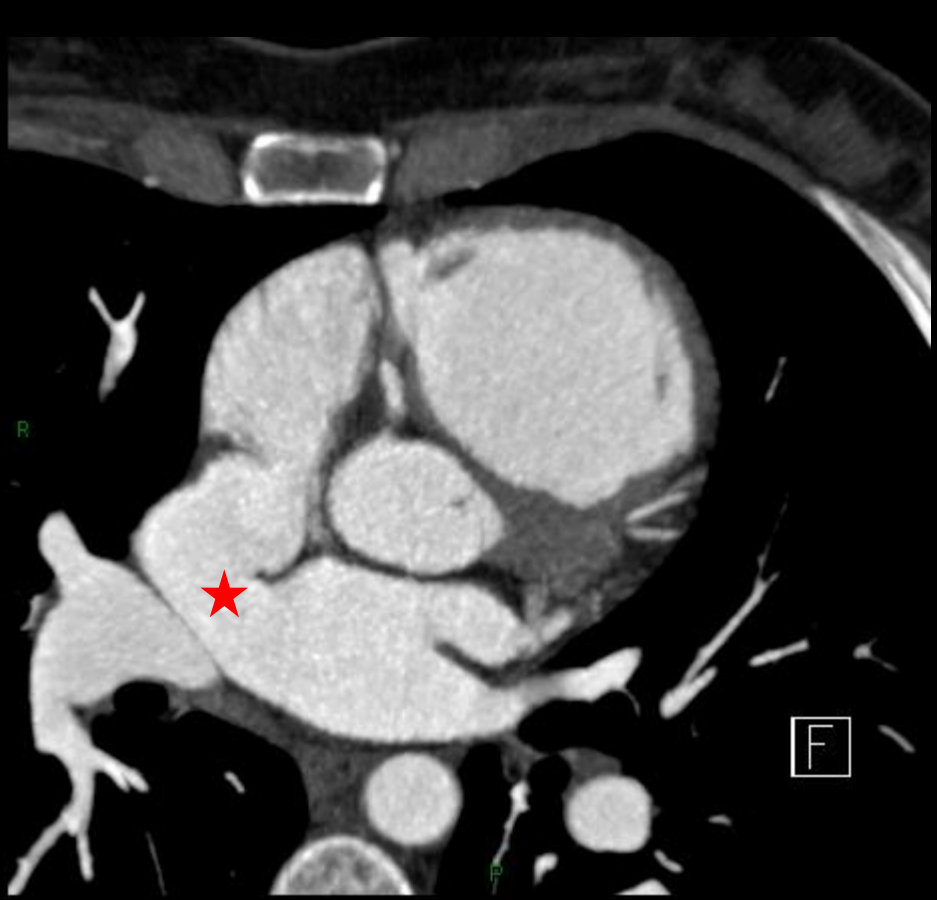

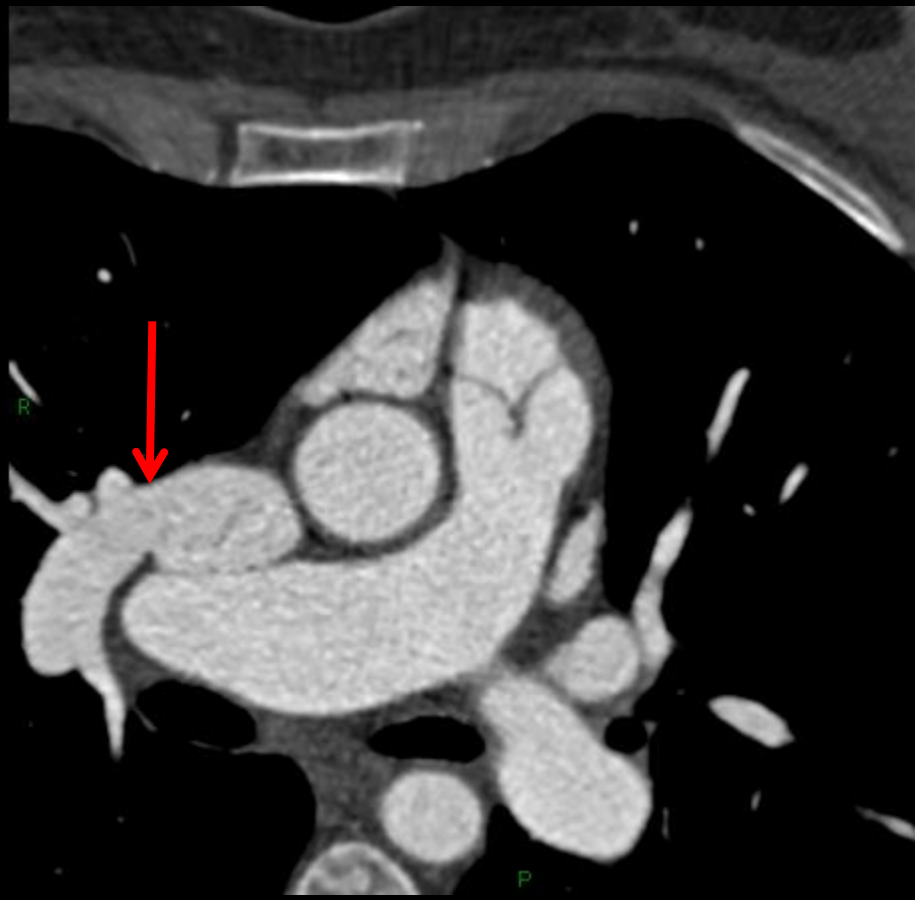

PAH associated with atrial septal defect and abnormal pulmonary venous return

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1.3. Drugs and toxins induced (including appetite suppressants)

1.4. Associated with
- 1.4.1. Connective vascular disease
- 1.4.2. HIV infection
- 1.4.3. Portal hypertension
- 1.4.4. Congenital heart disease
- 1.4.5. Schistosomiasis

1'. Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)

Simonneau et al. JACC 2013

1: Pulmonary Arterial Hypertension (PAH)

1'. Pulmonary veno-occlusive disease (PVOD)
Pulmonary capillary hemangiomatosis (PCH)

- Included in Group 1 because histologic features, clinical presentation, and risk factors similar to PAH
- Overlap between PCH and PVOD, because of the site and the size of the pulmonary vessels involved (capillary and adjoining venules, respectively)

➢ Essential to distinguish PVOD/PCH from other precapillary PAH, because potentially fatal pulmonary edema may be induced by vasodilatator therapy

Simonneau et al. JACC 2013

1': Pulmonary Veno-Occlusive Disease (PVOD)

- Very rare
- Male predominance (2:1)
- Idiopathic or heritable, drug-, toxin- and radiation-induced and associated forms (pregnancy, bone marrow transplantation)
- Clinical presentation indistinguishable from PAH: slowly progressive dyspnea
- Episode of acute pulmonary edema, hemoptysis
- Digital clubbing and bi-basal crackles unusual in other forms of PAH

Frazier et al. Radiographics 2007
Frazier et al. Radiographics 2000
Swensen et al. AJR 1996

1’: PVOD – Pathology

- Post-capillary lesions in septal veins and pre-septal venules consisting of fibrous remodeling of the intima
- Alveolar hemorrhage
- Pulmonary edema

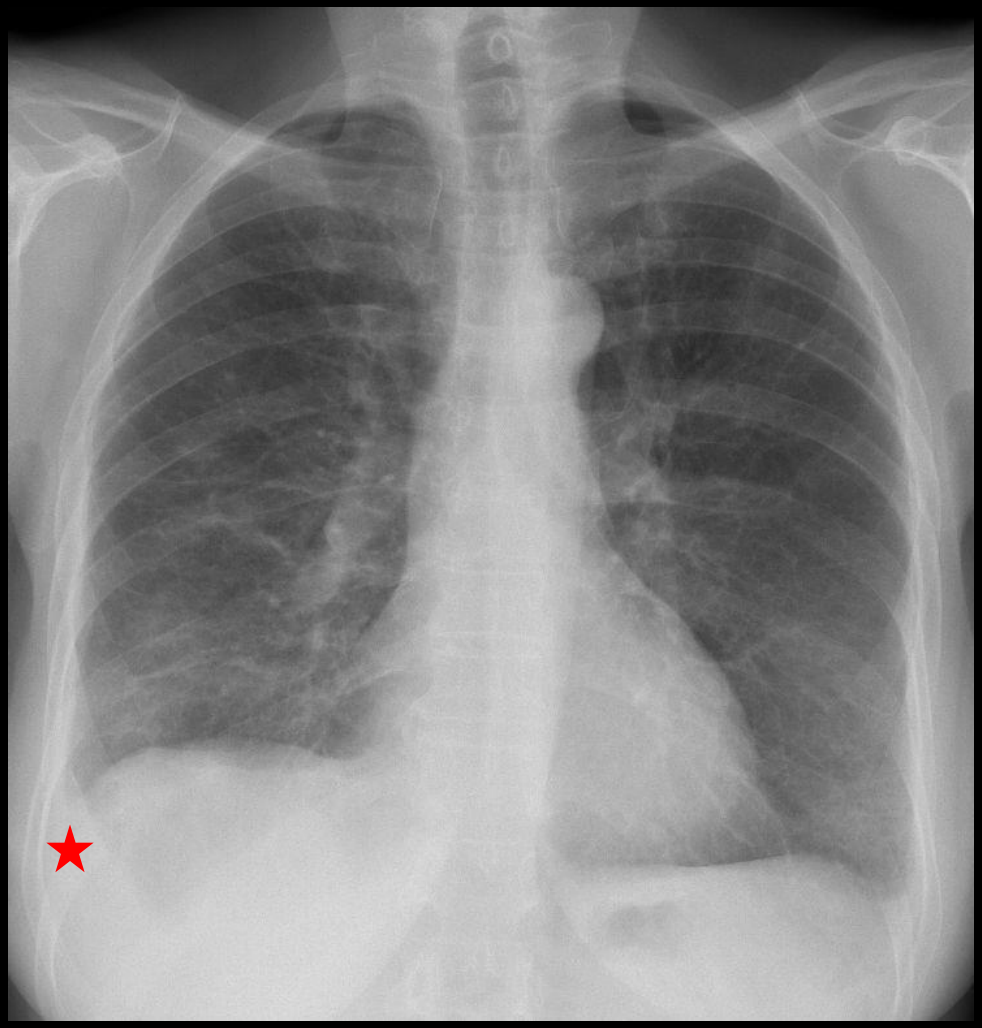
Frazier et al. Radiographics 2007
Frazier et al. Radiographics 2000
Swensen et al. AJR 1996

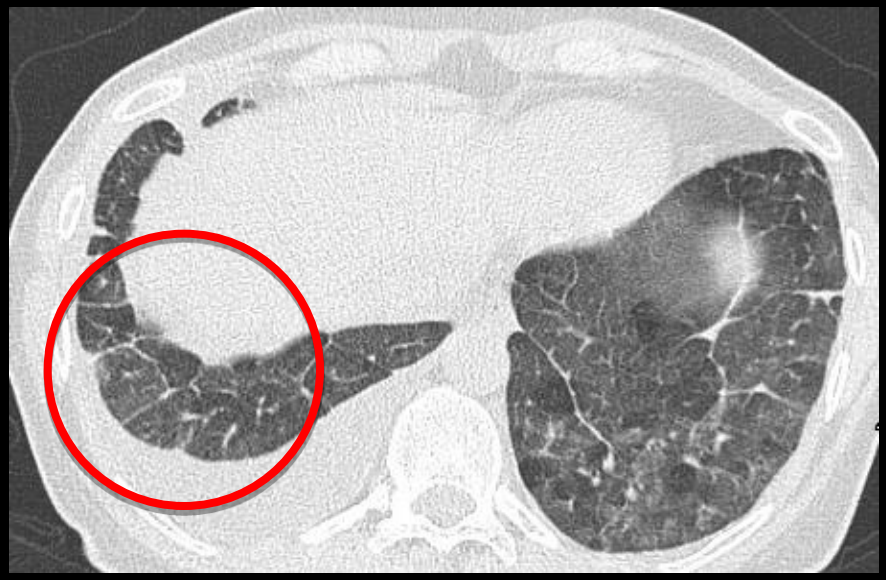
1': PVOD – Particular CT Features

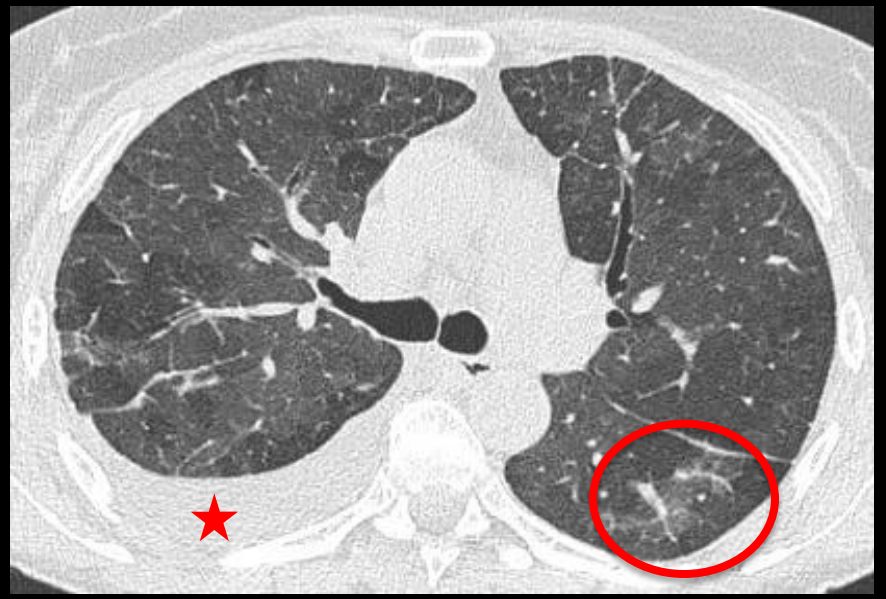
- Combination of PH with pulmonary edema
  - Small central pulmonary veins
  - Interlobular septal thickening
  - Patchy centrilobular ground-glass opacities
  - Mosaic lung attenuation
  - Pleural effusions
  - Mediastinal lymphadenopathy
  - Dilation of the central pulmonary arteries
  - Right ventricular enlargement
  - Normal sized left atrium and ventricle
  - Normal caliber pulmonary veins

Frazier et al. Radiographics 2007
Frazier et al. Radiographics 2000
Swensen et al. AJR 1996
Montani D et al. Eur Respir J 2009
Montani D et al. Medicine 2008

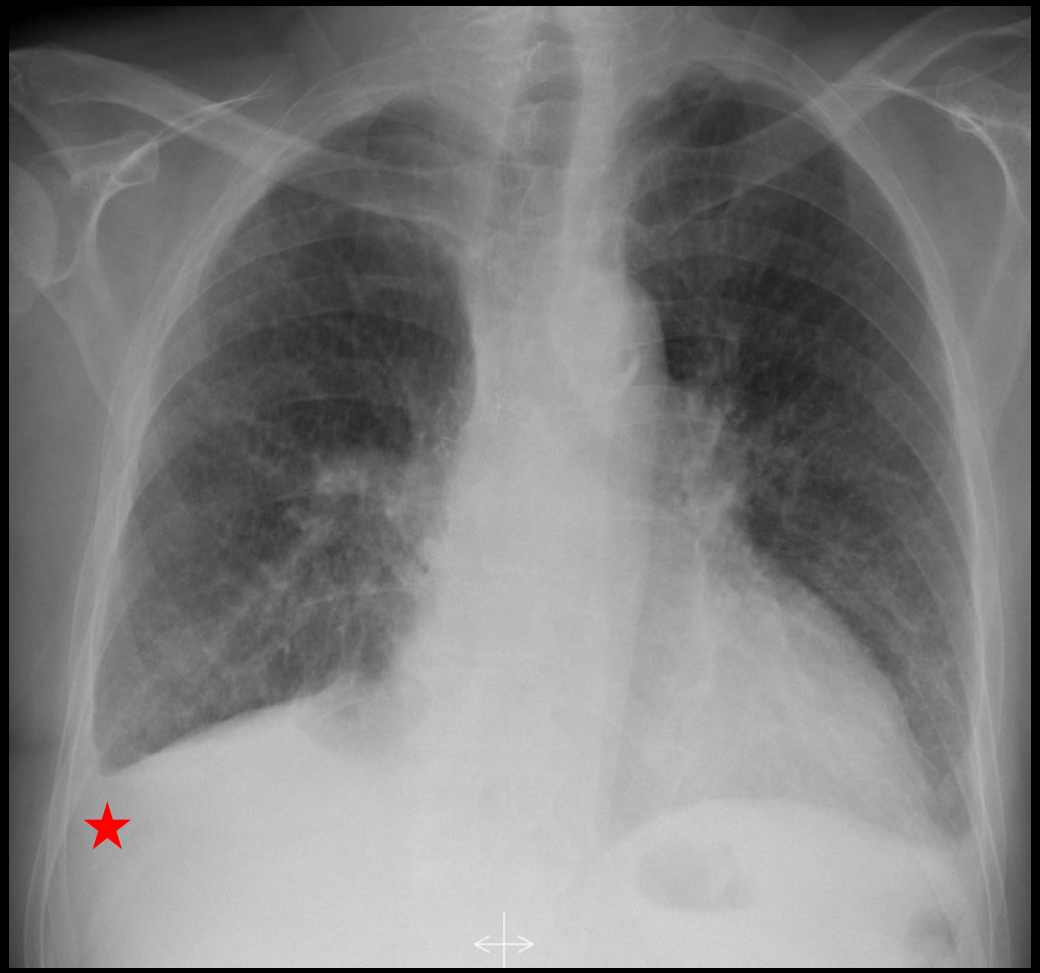
1’: PVOD – Particular CT Features

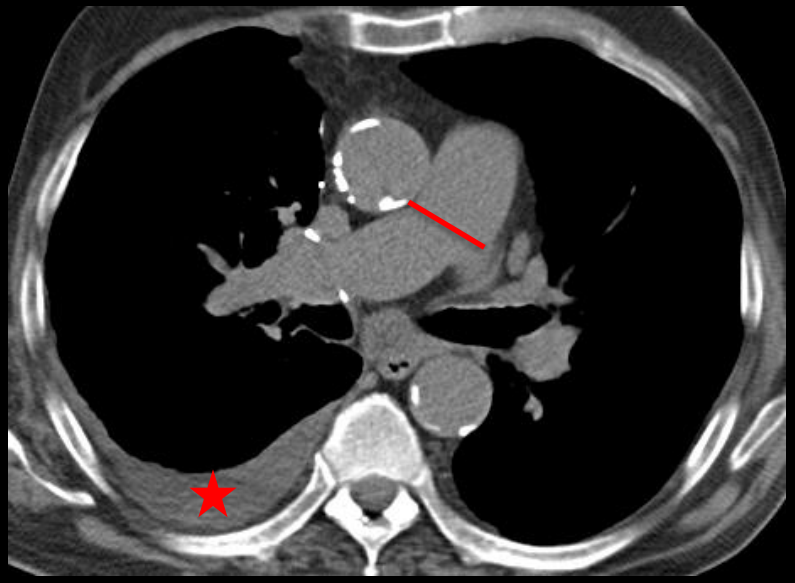


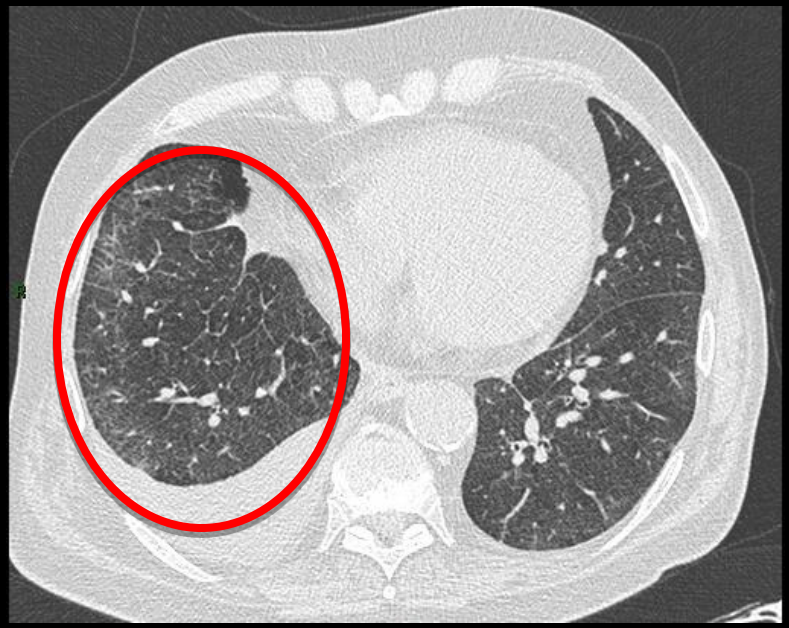




1’: PVOD – Particular CT Features







1': Pulmonary Capillary Hemangiomatosis (PCH)

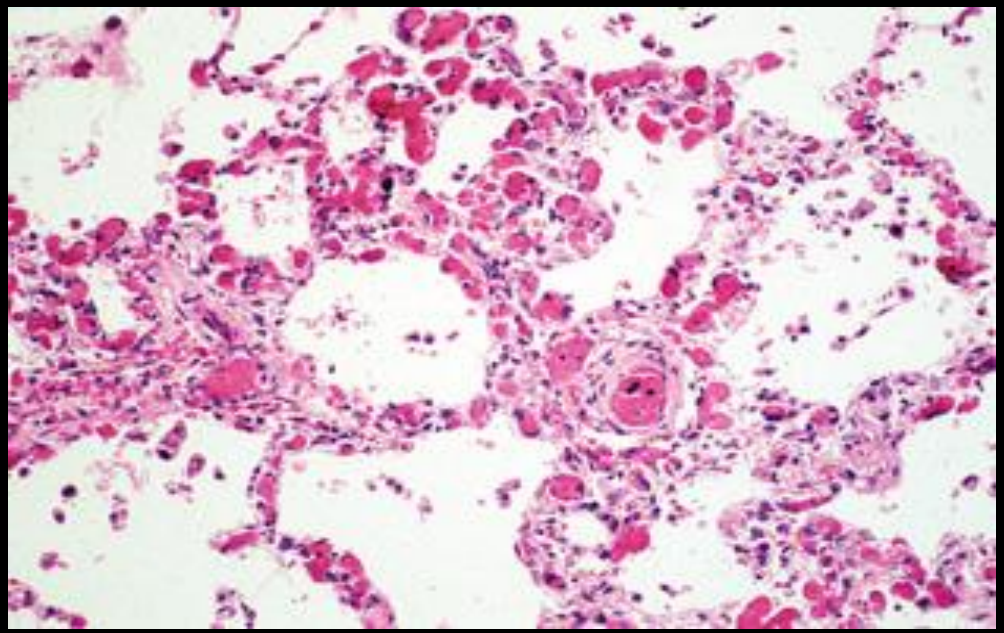
Hôpital Erasme

ULB

- Unknown etiology
- No gender predominance
- 20-40 years
- Poor prognosis
- Clinical presentation indistinguishable from PAH

1': PCH – Pathology

- Proliferation of thin walled capillary channels (abnormal capillaries) invading alveolar septae
- They subsequently compress the walls of veins and venules causing intimal fibrosis and secondary veno-occlusion



Proliferating double layer of alveolar capillaries, with congestion and dilation

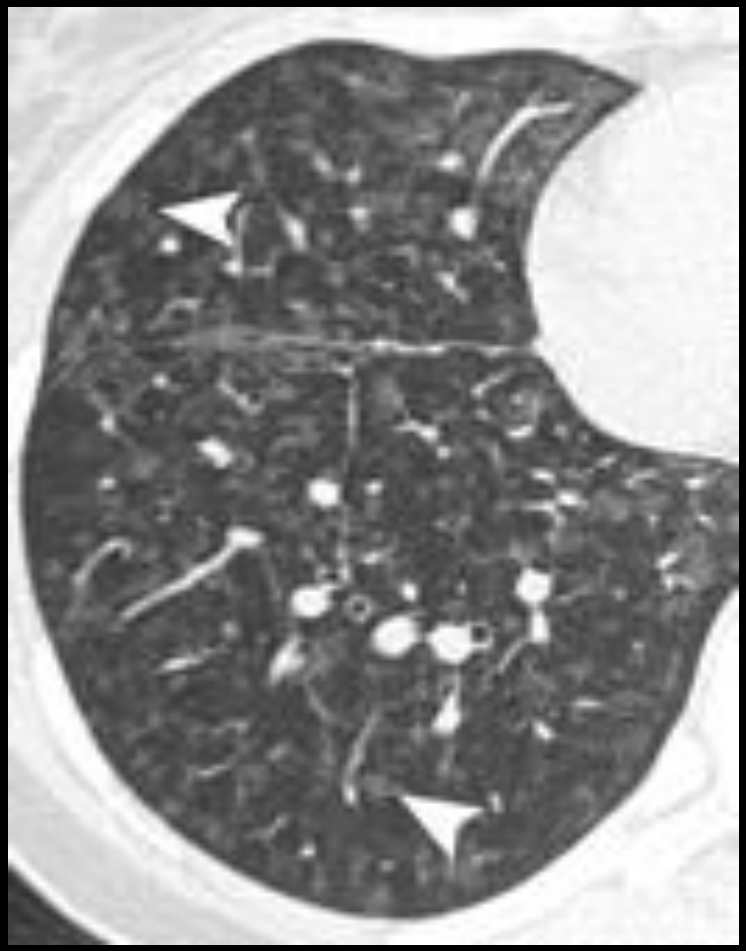
Cagle PT et al. Color Atlas and Text of Pulmonary Pathology. 2nd Edition (2007). Lippincott, Williams and Wilkins

1’: PCH – CT Features

- Widespread ill defined centrolobulaire nodules of GGO
- Lobular GGO
- Interlobular septal thickening
- Lymphadenopathy
- Pleural effusion
- Pericardial effusion
- Dilation of central pulmonary arteries
- Enlargement of rights chambers of heart
- Normal left atrium

Frazier et al. Radiographics 2007
Frazier et al. Radiographics 2000

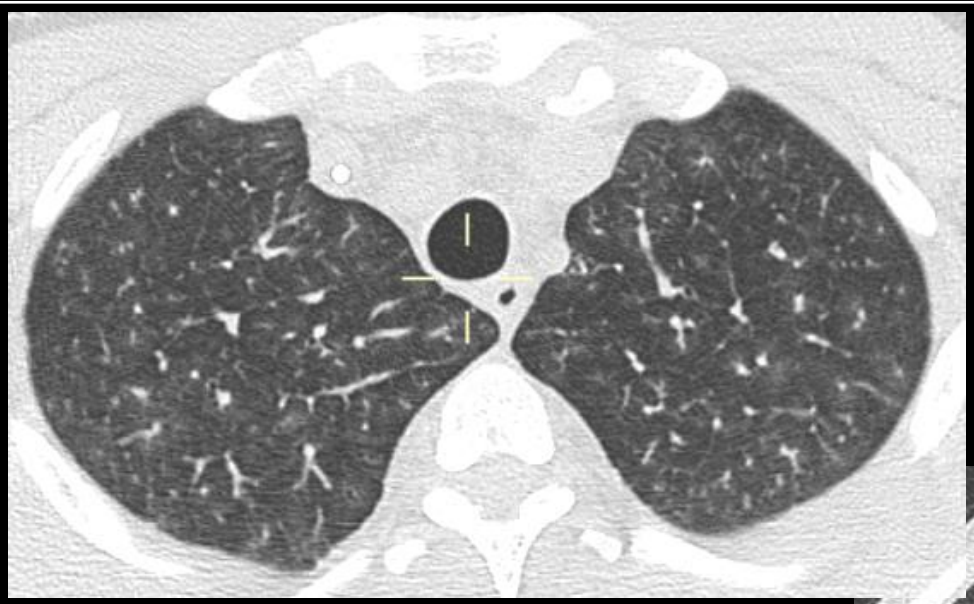
1’: PCH – CT Features

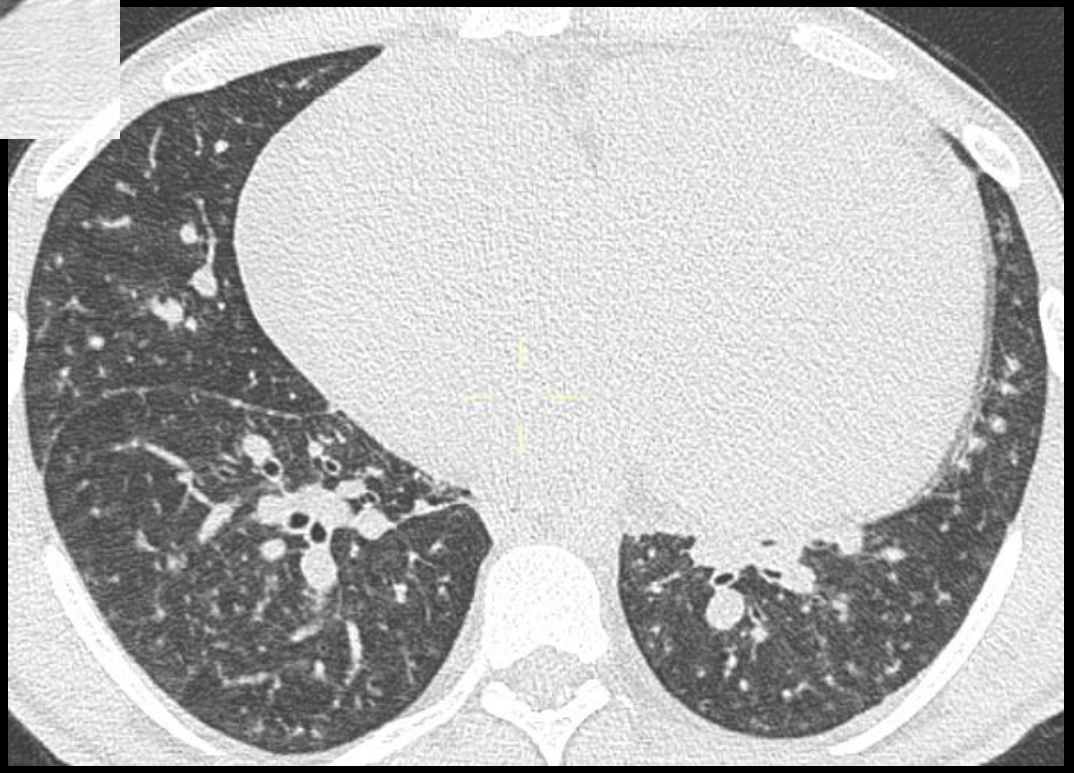


PCH in a 22-year-old woman. Axial CT image (lung window settings) collimated to the right lower lobe shows well-circumscribed ground-glass nodules (arrowheads) and no septal lines.

Frazier et al. Radiographics 2007

1’: PCH – CT Features





Hemodynamic Definitions (Nice)

Definition	Characteristics	Clinical group(s)
Pulmonary hypertension (PH)	Mean PAP ≥ 25 mmHg	All
Pre-capillary PH	Mean PAP ≥ 25 mmHg	1. Pulmonary arterial hypertension
	PWP ≤ 15 mmHg	3. due to lung diseases
	CO normal or reduced	4. Chronic Thromboembolic PH
		5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	Mean PAP ≥ 25 mmHg	2. PH due to left heart disease
	PWP > 15 mmHg	5. PH with unclear and/or multifactorial mechanisms
	CO normal or reduced	
Passive	TPG ≤ 12 mmHg	
Reactive (out of proportion)	TPG > 12 mmHg	

PAP = pulmonary arterial pressure, PWP = pulmonary wedge pressure, CO = cardiac output,

TPG = Transpulmonary pressure gradient

2015 ESC/ERS Guidelines. *Eur Haert J*

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2015 ESC/ERS Guidelines. *Eur Haert J*

Definitions

- PH can be found in multiple clinical conditions: 5 Groups

1. Pulmonary arterial hypertension (PAH) ✓
2. **PH due to cardiac disease**
3. PH due to chronic lung disease and/or hypoxia
4. Chronic thromboembolic PH (CTEPH)
5. PH due to unclear multifactorial mechanisms

Simonneau et al. JACC 2013

2: PH Due to Cardiac Diseases

- Systolic dysfunction
- Diastolic dysfunction
- Valvular disease

- Backward transmission of filling pressures

- PAWP >15 mmHg

Simonneau et al. JACC 2013
Vachiéry et al. JACC 2013

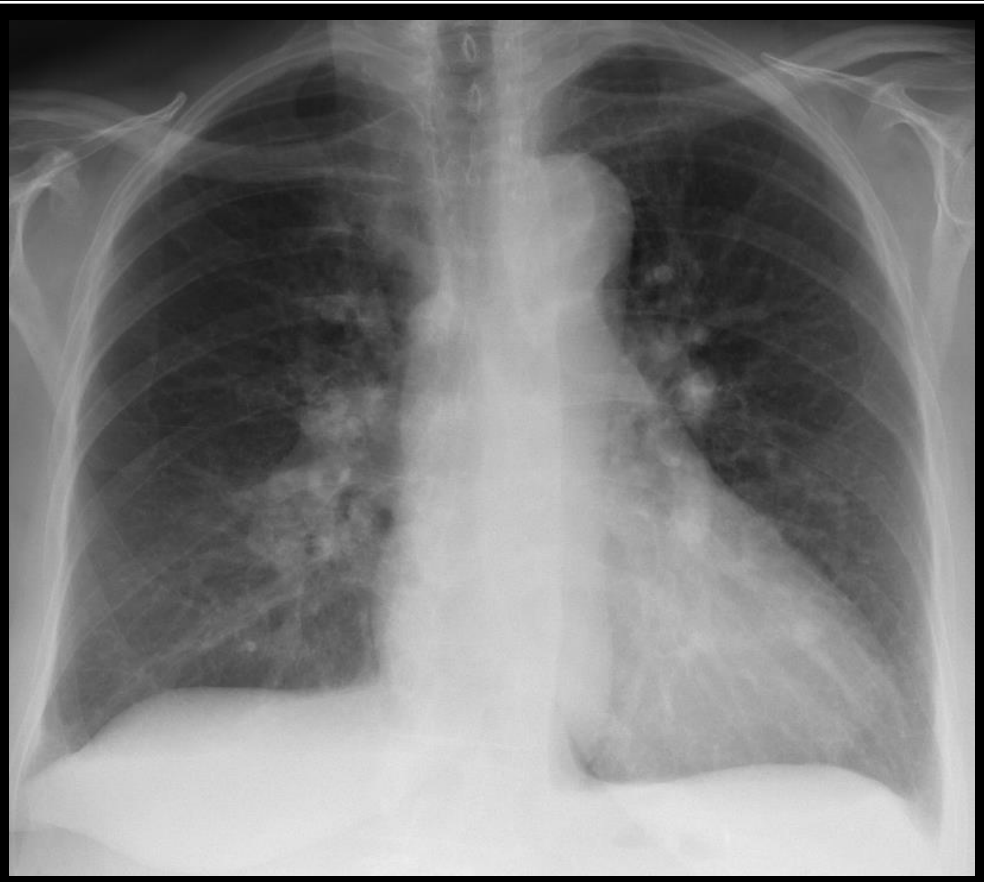
2: PH Due to Cardiac Diseases

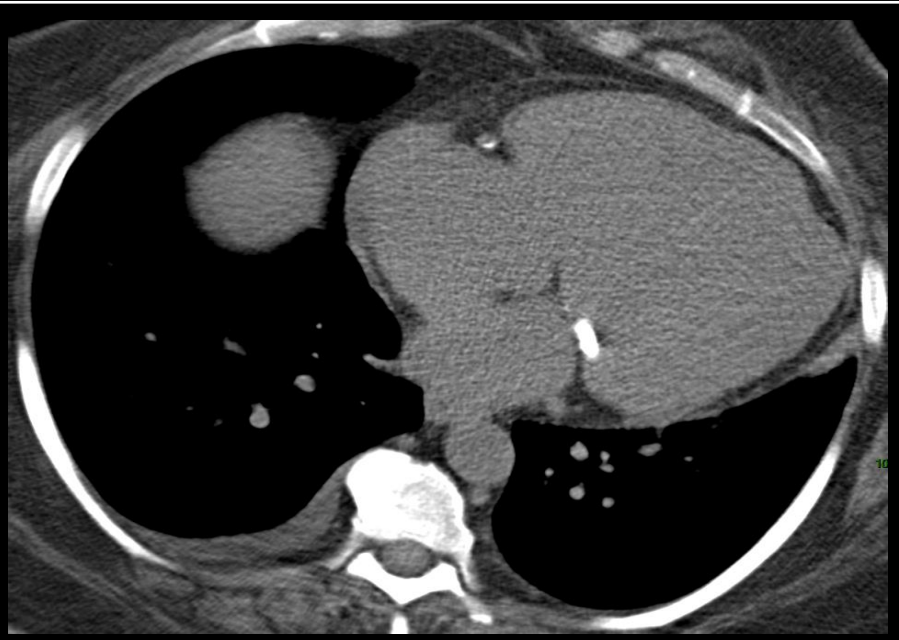
- Imaging findings include the superposition of pulmonary arterial and venous hypertension

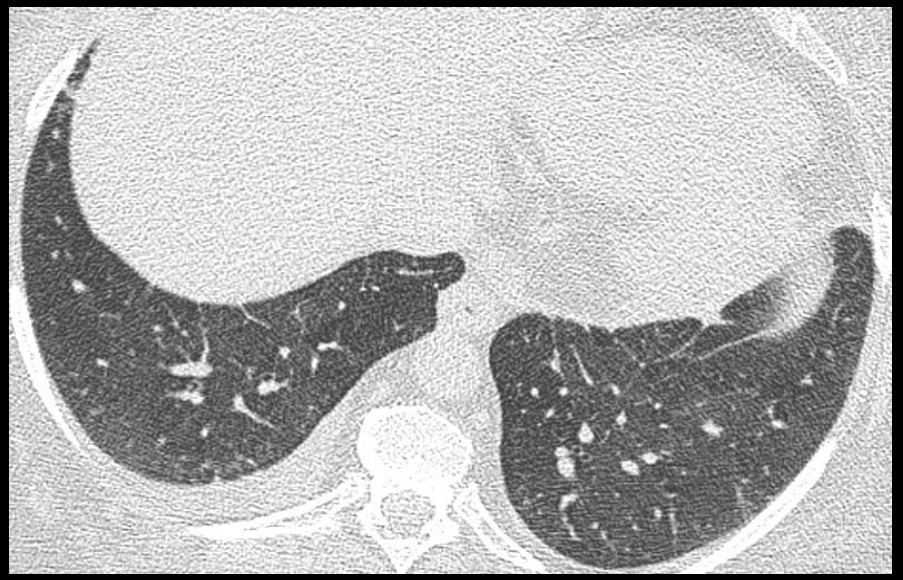
- The image can strongly resemble PVOD; in the latter the left cavities are of normal dimensions

Hansell et al. Imaging of Diseases of the Chest. 5th Edition. Mosby Elsevier

2: PH Due to Cardiac Diseases







Definitions

- PH can be found in multiple clinical conditions: 5 Groups
  1. Pulmonary arterial hypertension (PAH) ✓
  2. Pulmonary hypertension due to cardiac disease ✓
  3. **PH due to chronic lung disease and/or hypoxia**
  4. Chronic thromboembolic PH (CTEPH)
  5. PH due to unclear multifactorial mechanisms

Simonneau et al. JACC 2013

3: PH Due to Chronic Lung Disease and/or Hypoxia

Hôpital Erasme

ULB

A. Chronic Obstructive Pulmonary Disease (COPD)

- Prevalence of PH in advanced COPD > 50%
- Generally mild severity
- Presence of PH associated with shorter survival and frequent exacerbation

Devaraj et al. Radiology 2008

3: PH Due to Chronic Lung Disease and/or Hypoxia

Hôpital Erasme

ULB

B. Idiopathic Pulmonary Fibrosis (IPF)

- PH in up to 46% patients with IPF
- In IPF, the main pulmonary artery diameter is an unreliable indicator of PH (PA dilation occurs in the absence of PH)
- The ratio PA diameter to ascending aorta diameter is a more reliable marker of PH
- Lack of correlation between the mean PAP and the extent of fibrosis

Devaraj et al. Radiology 2008

3: PH Due to Chronic Lung Disease and/or Hypoxia

Hôpital Erasme | ULB

C. Other pulmonary diseases with mixed restrictive and obstructive pattern
   - Chronic bronchiectasis
   - Cystic fibrosis
   - Combination of emphysema (upper zones) and fibrosis (lower zones) : prevalence PH 50%

D. Sleep-disordered breathing
E. Alveolar hypoventilation disorders
F. Chronic exposure to high altitude
G. Developmental abnormalities (thoracic cage deformities, diaphragmatic disorders, neuromuscular diseases, spinal cord injuries)

Definitions

- PH can be found in multiple clinical conditions: 5 Groups

1. Pulmonary arterial hypertension (PAH) ✓
2. PH due to cardiac disease ✓
3. PH due to chronic lung disease and/or hypoxia ✓
4. **Chronic thromboembolic PH (CTEPH)**
5. PH due to unclear multifactorial mechanisms

Simonneau et al. JACC 2013

4: CTEPH

- Result of incomplete resolution and organization of recurrent PE (Occurs in 3.8% of cases of acute symptomatic PE)
- Up to 2/3 of patients have no history of PE

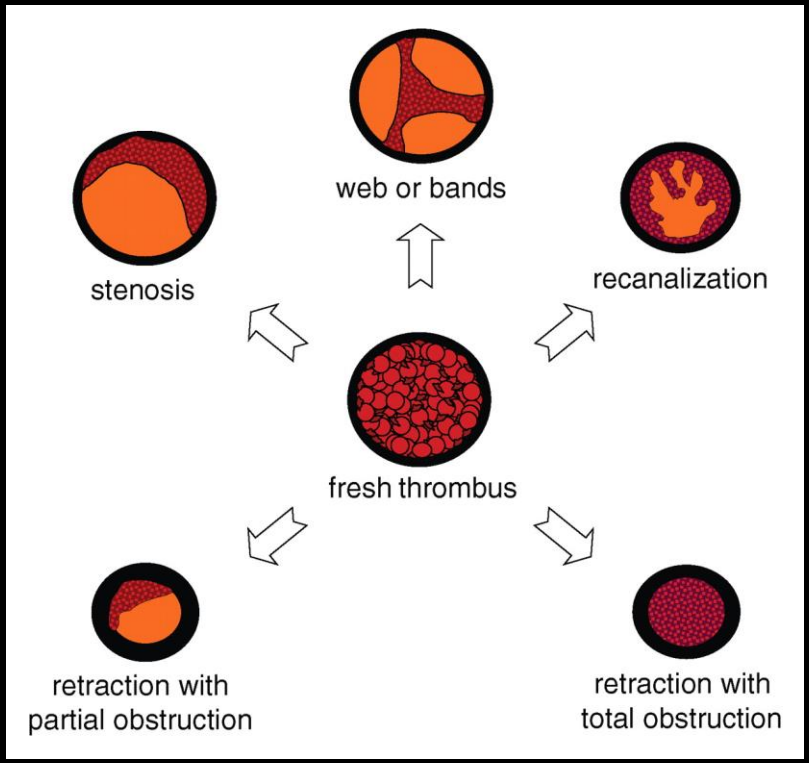


Diagram shows the various possible results of disturbed resolution of a thrombus:

- vascular stenosis
- retraction with total obstruction
- retraction with partial obstruction
- recanalization
- residual fibrous cords (web or bands)

Castaner et al. Radiographics 2009

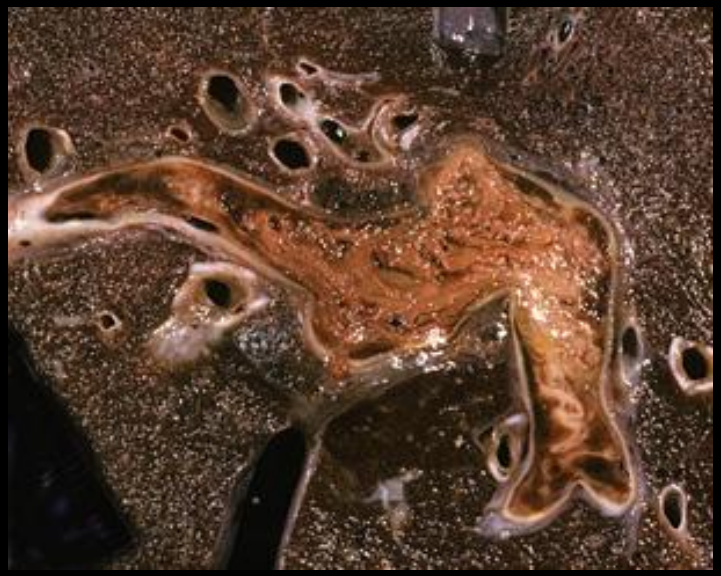
4: CTEPH

- Risk factors
  - Woman
  - Malignant, cardiovascular, or pulmonary disease
  - Lupus anticoagulant, anti-cardiolipin antibodies

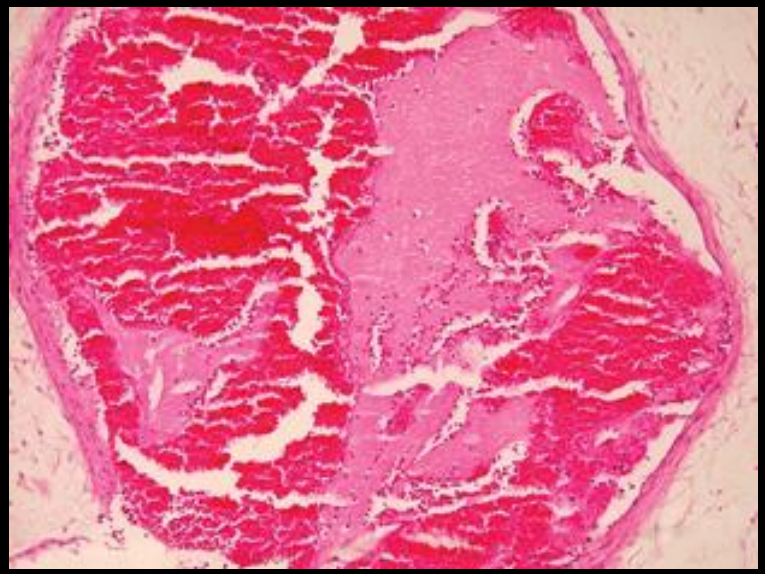
- Conditions that predispose patients to CTEPH and predictive of poor prognosis
  - Splenectomy
  - Ventriculo-atrial shunt (hydrocephalus)
  - Myeloproliferative disorders
  - Chronic inflammatory bowel disease

Castaner et al. Radiographics 2009

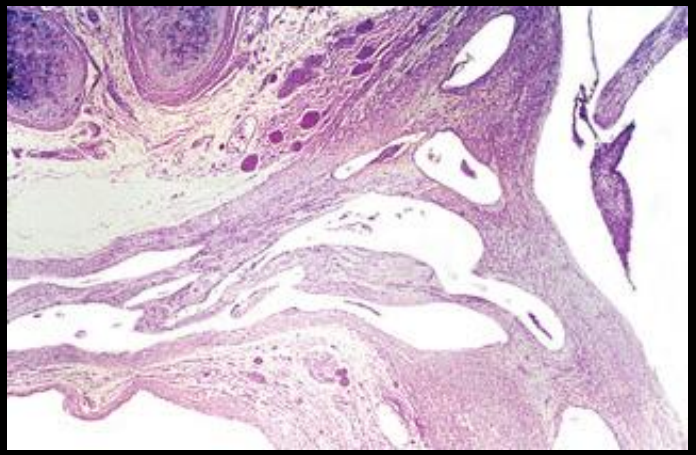
4: CTEPH – Pathology



Pulmonary artery with a laminated, firm, granular thromboembolus



Small pulmonary artery distended with a recent thromboembolus, consisting mostly of fibrin and red blood cells



Large pulmonary artery branch with old organized thrombus replaced by pulmonary webs

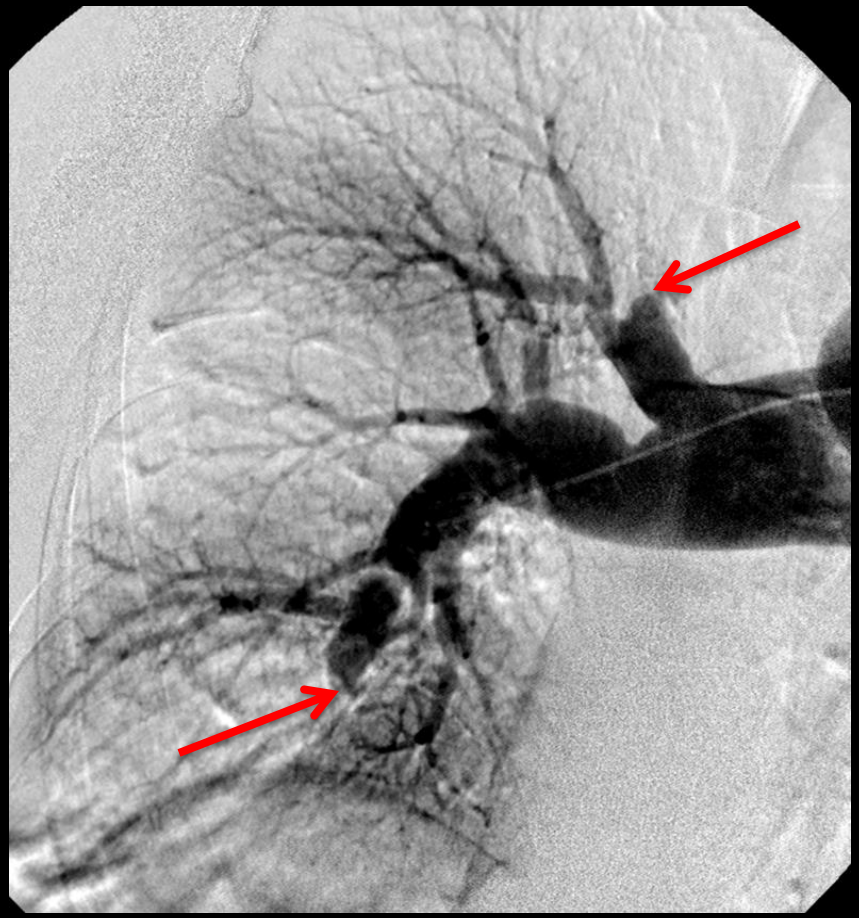
Cagle PT et al. Color Atlas and Text of Pulmonary Pathology. 2nd Edition (2007). Lippincott, Williams and Wilkins

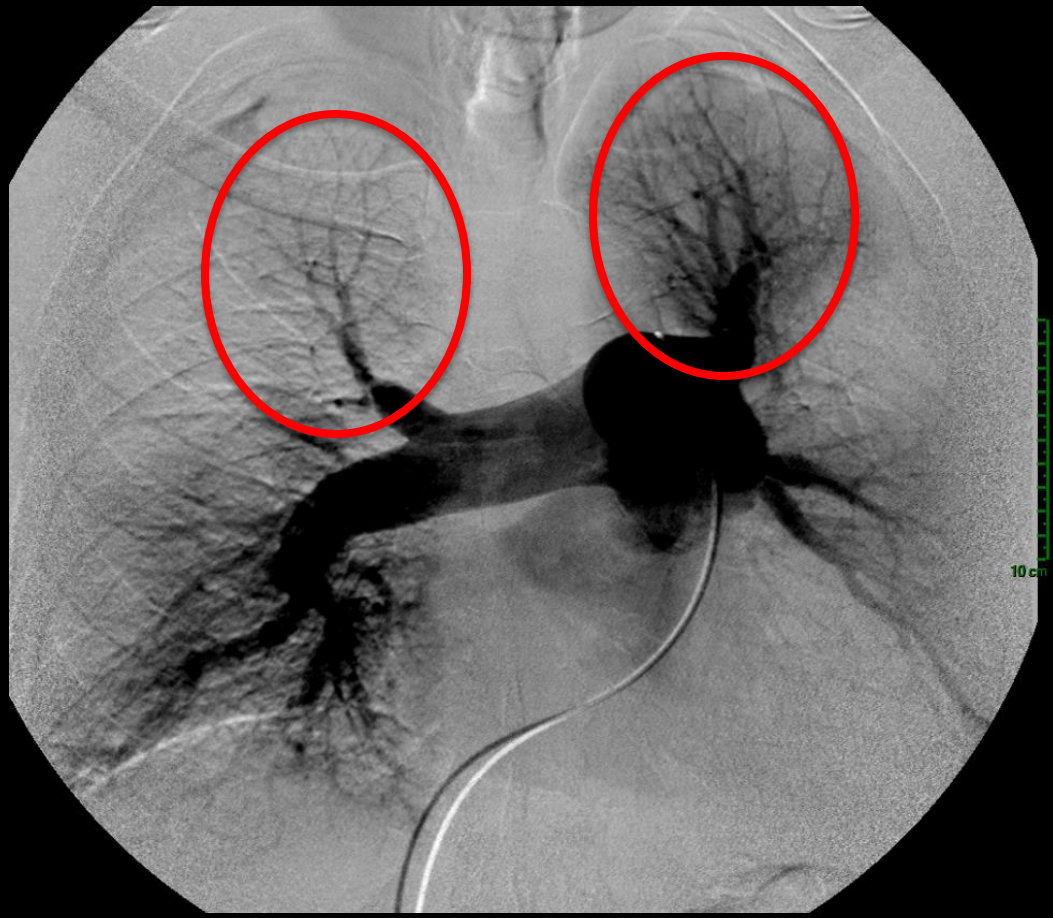
4: CTEPH – Pulmonary Angiography

- Pouching defects: obstructive or partially occlusive chronic emboli organized in a concave configuration
- Webs or bands: decreased attenuation opacities/lines traversing the lumen usually in lobar or segmental branches
- Scalloping appearance due to intimal irregularity
- Abrupt narrowing, lost of gentle tapering
  - Recanalization, eccentric organized thrombus, artery contraction by organized thrombus
- Obstruction of lobar vessels at their point of origin

Auger W et al. Radiology 1992

4: CTEPH – Pulmonary Angiography



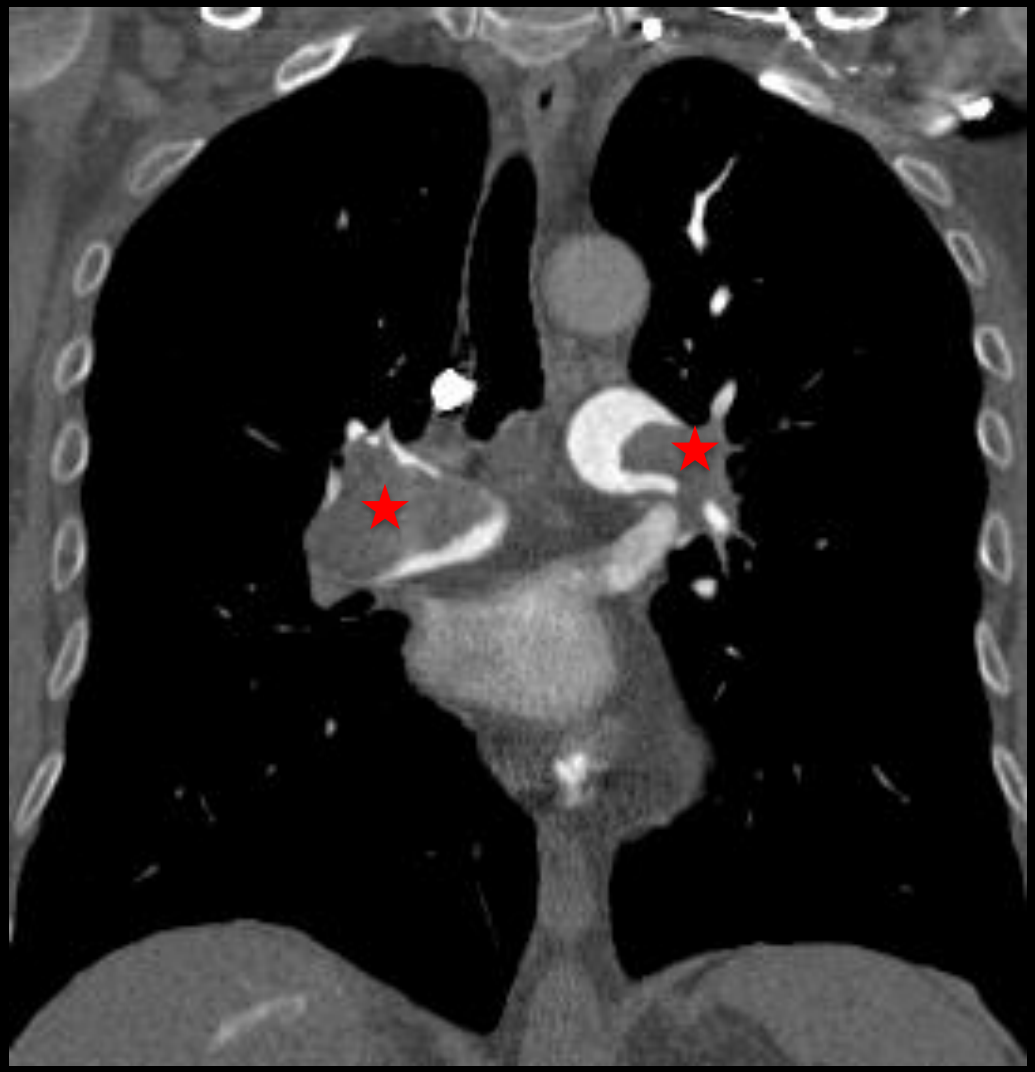


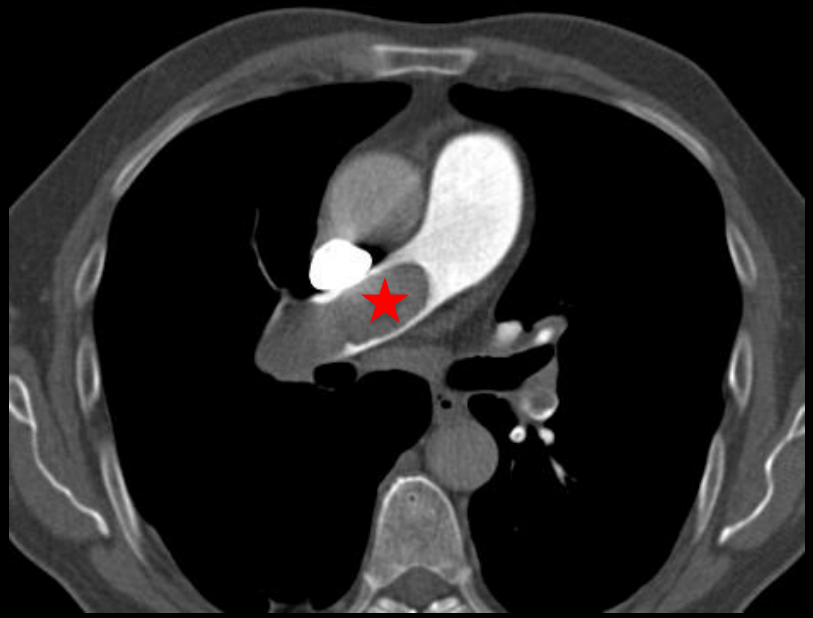
4: CTEPH – CT Features: Pulmonary arteries

- Partial filling defects
- Thickening of the pulmonary arterial wall, irregular contour of the intimal surface
- Bands and webs (multiples bands forming a network) : thin lines surrounded by contrast material, most frequently in lobar or segmental arteries
- Calcifications within chronic thrombi

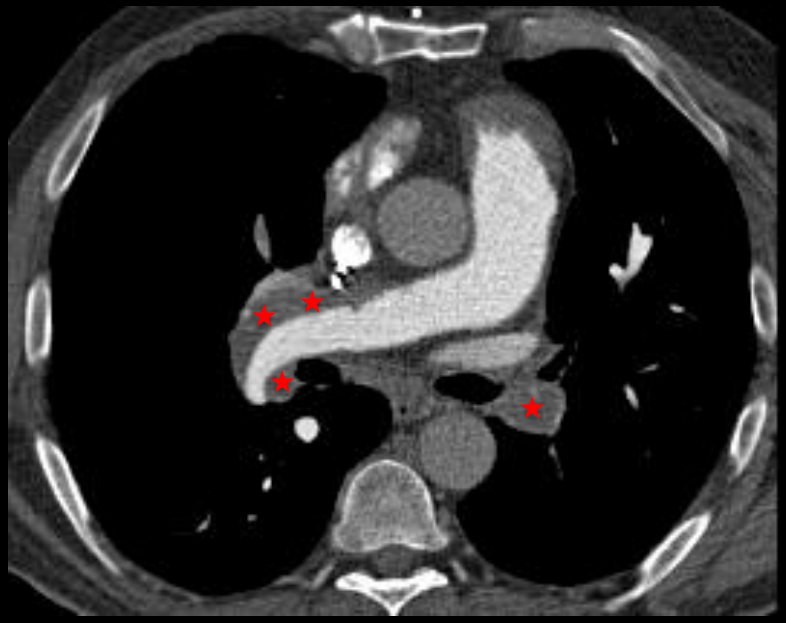
Auger W et al. Radiology 1992

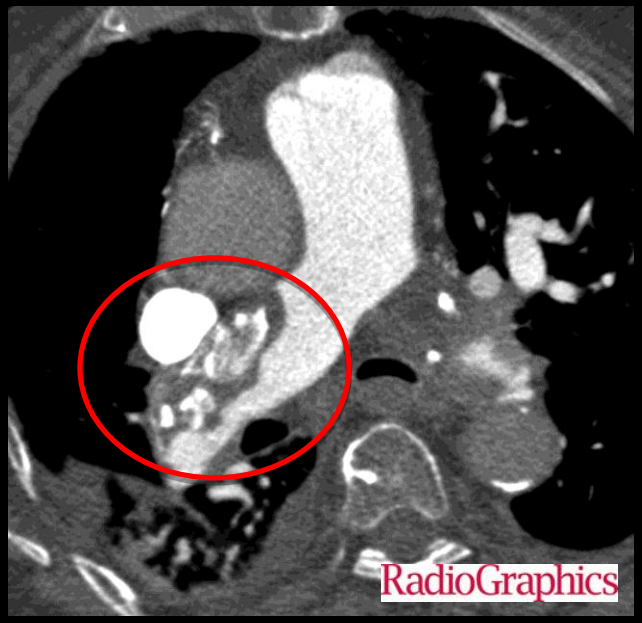
4: CTEPH – CT Features: Pulmonary arteries

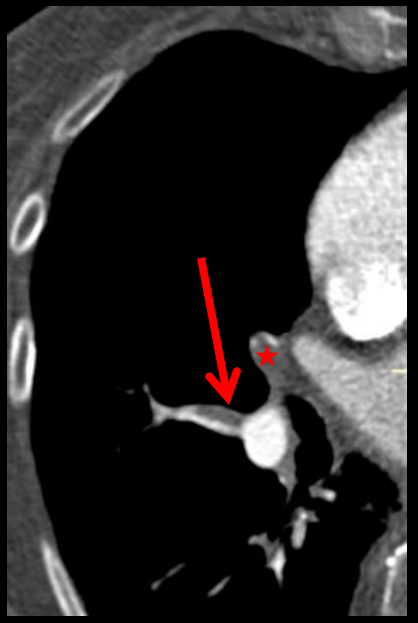


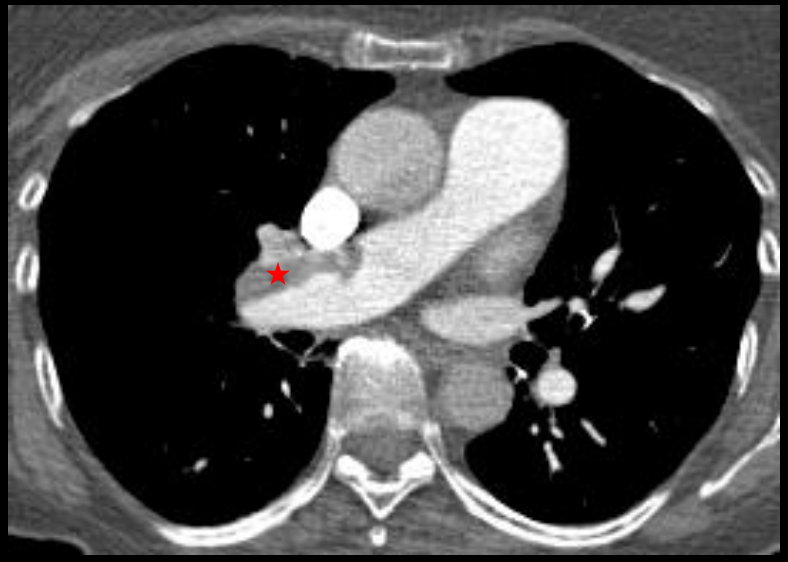


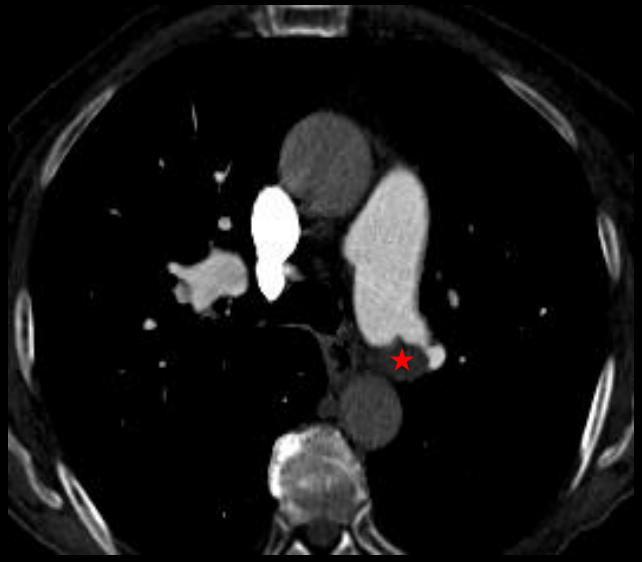
4: CTEPH – CT Features: Pulmonary arteries



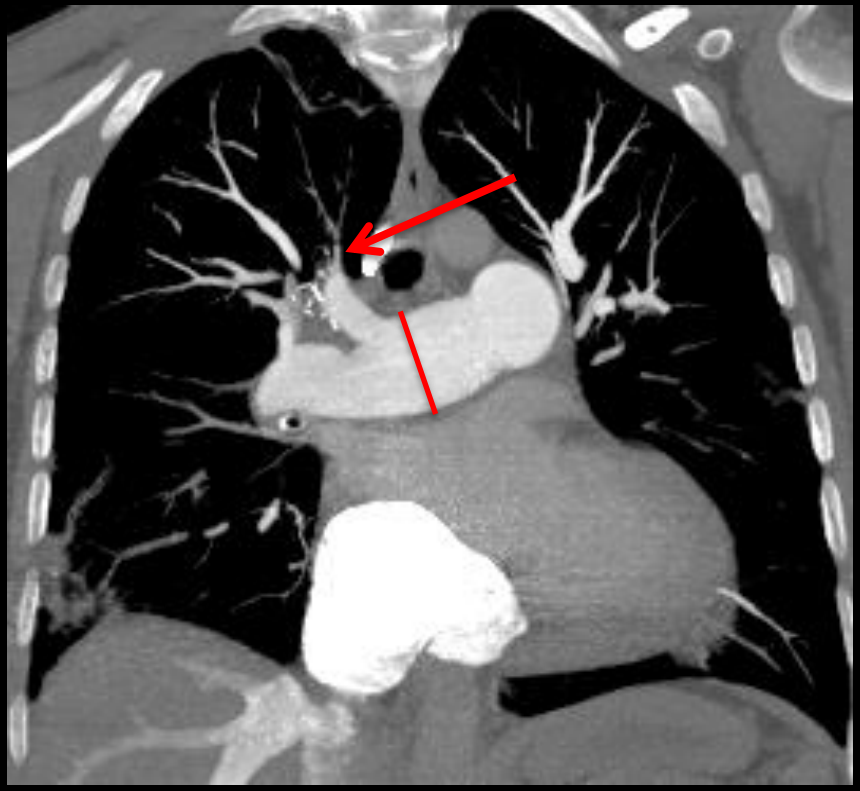


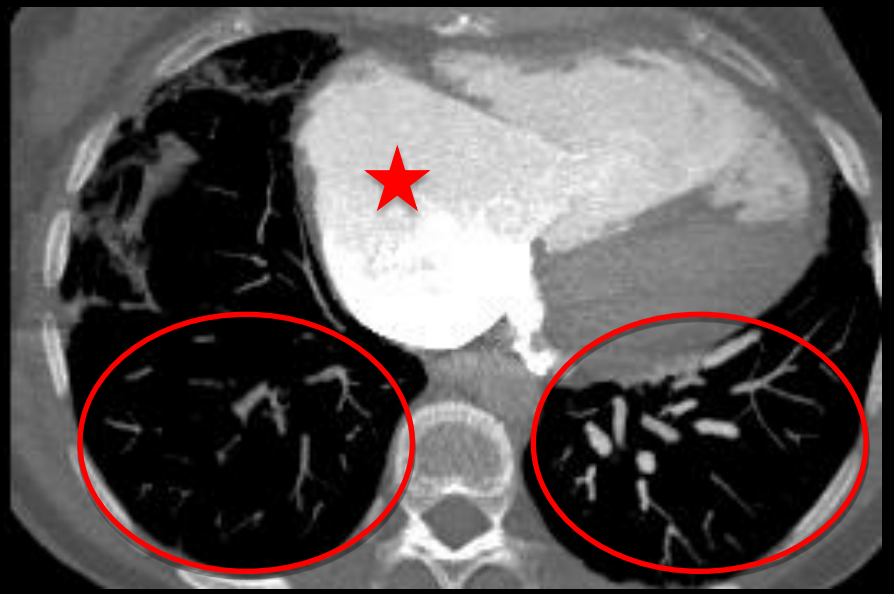


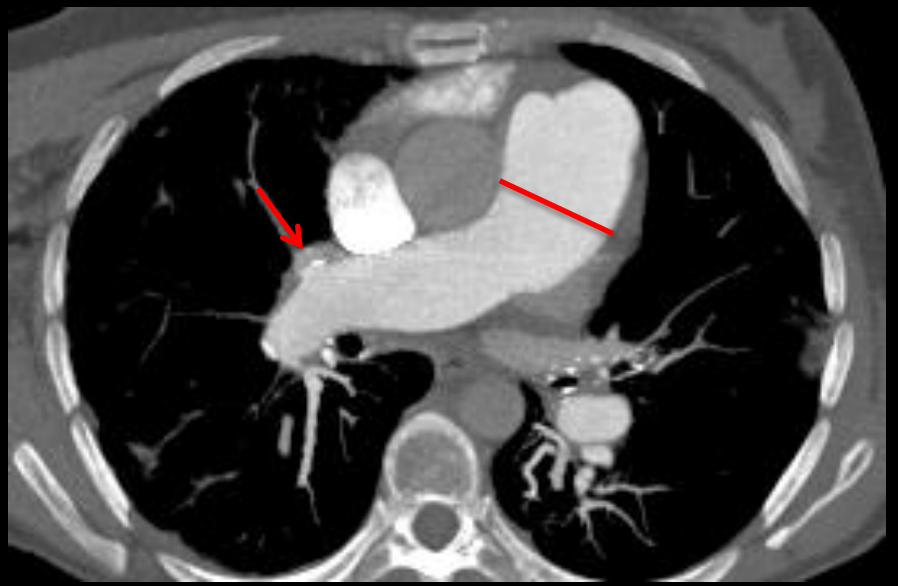




4: CTEPH – CT Features: Pulmonary arteries







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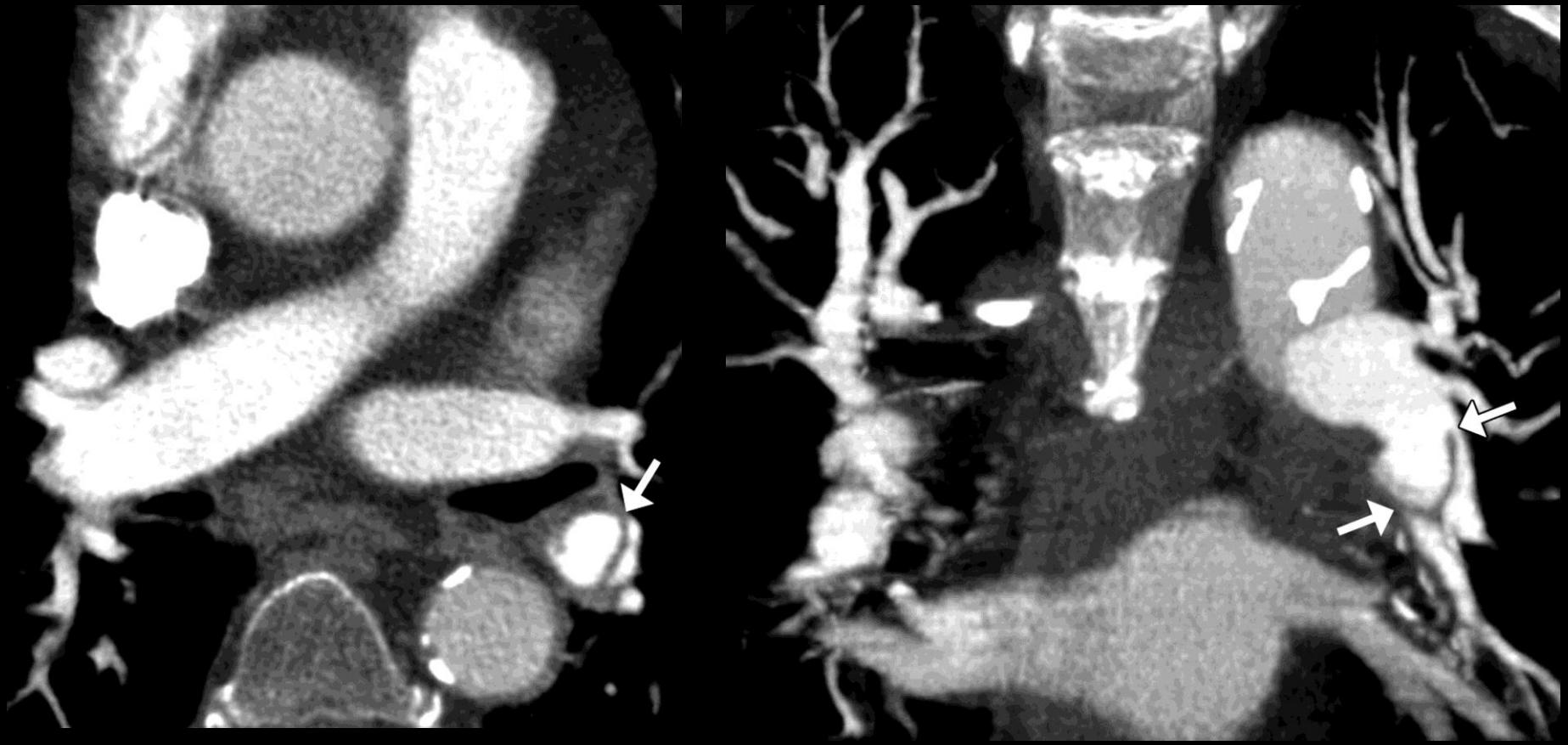
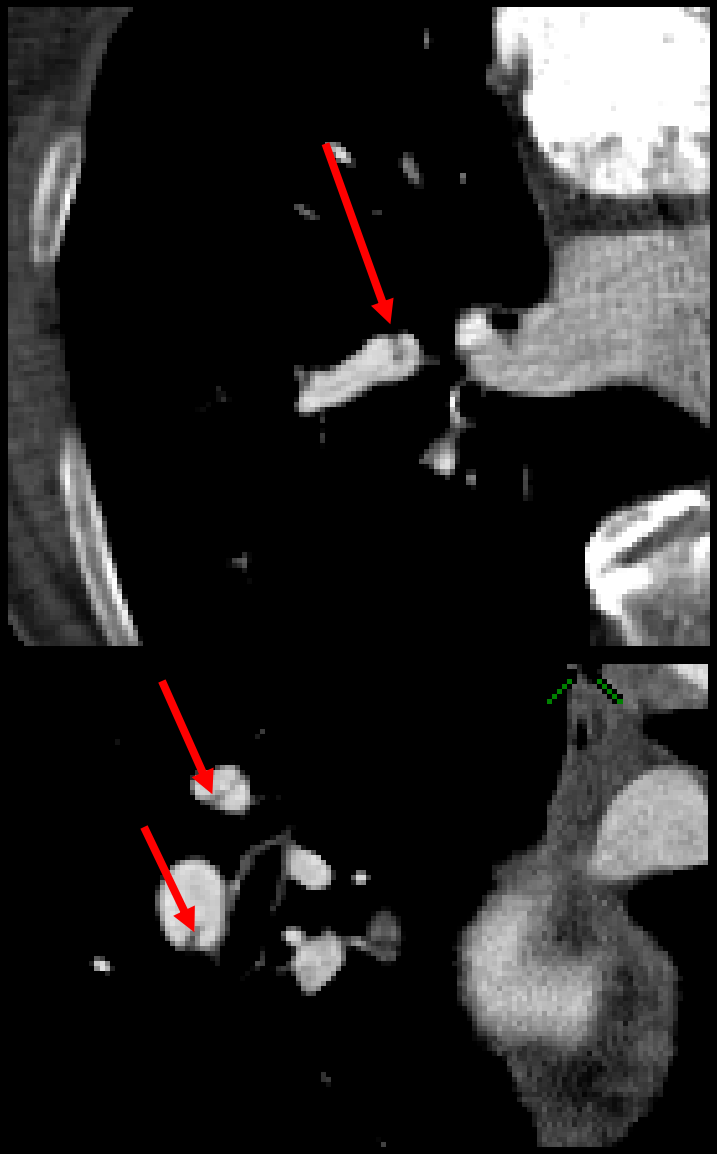
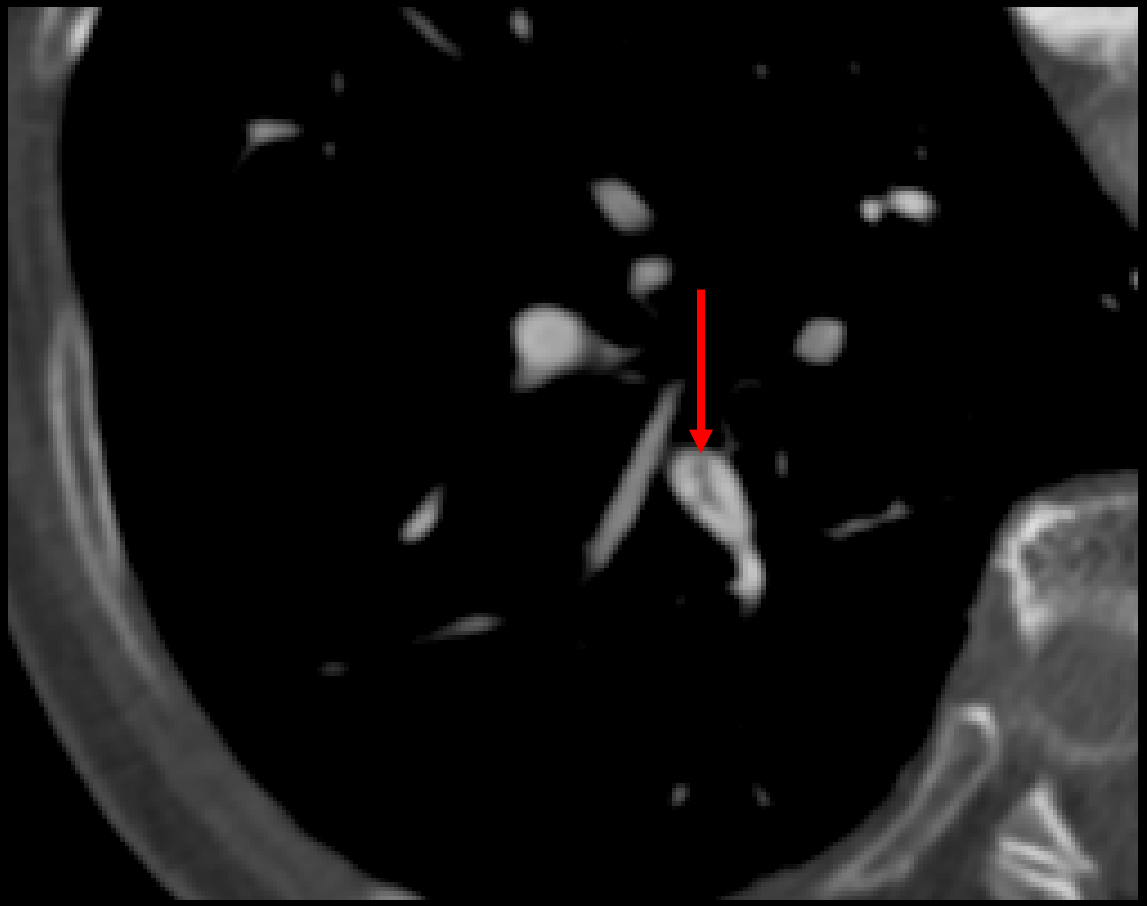


Residual band from a pulmonary thrombus in an 83-year-old woman with dyspnea

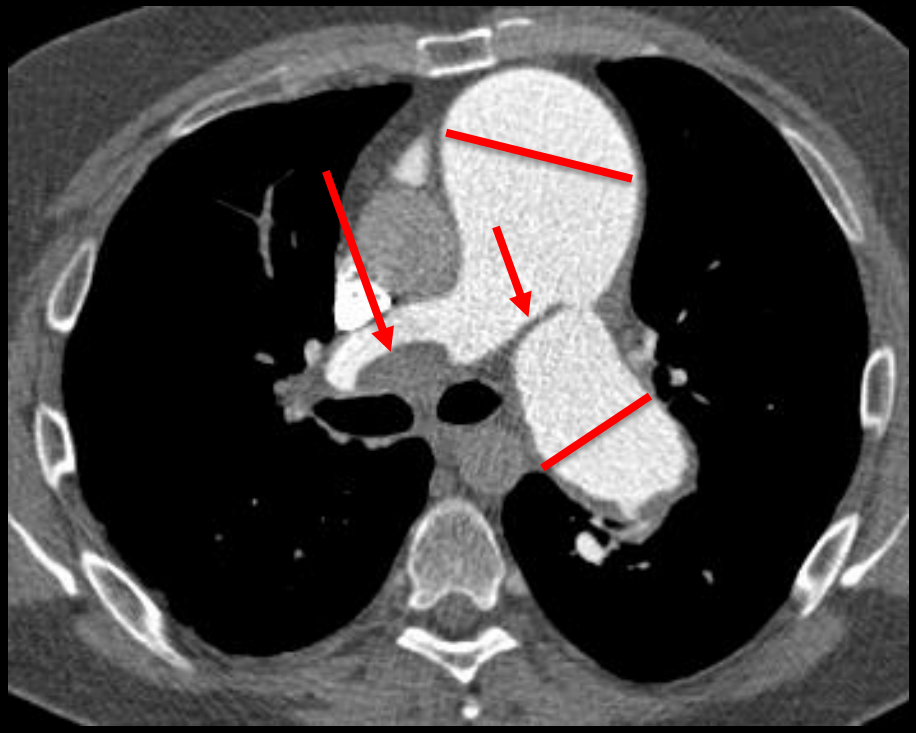
Castaner et al. Radiographics 2009

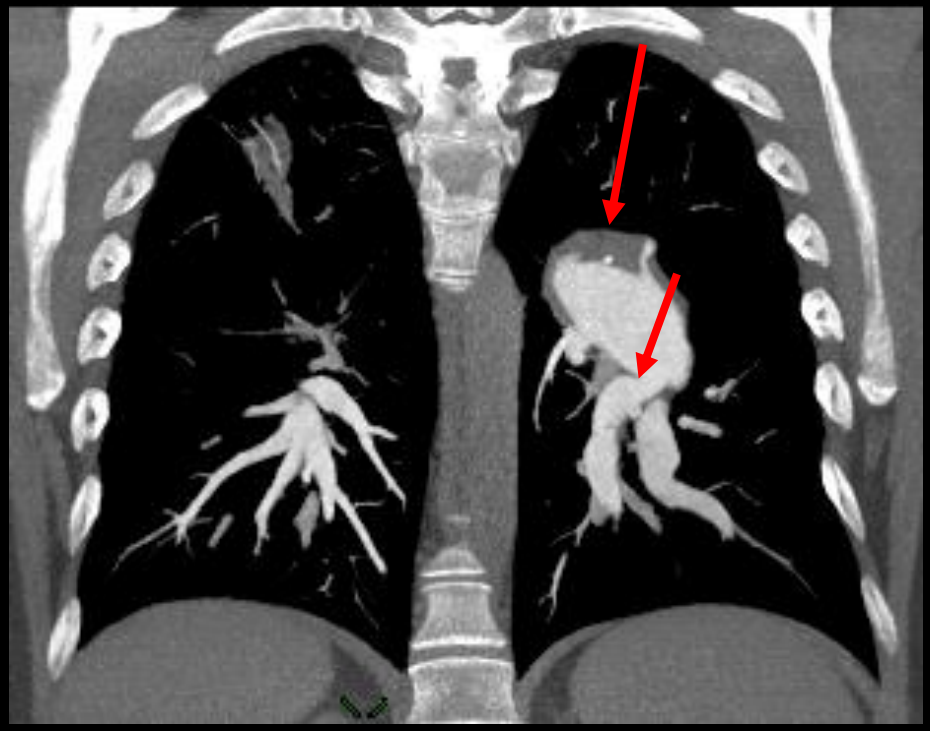
4: CTEPH – CT Features: Pulmonary arteries





4: CTEPH – CT Features: Pulmonary arteries



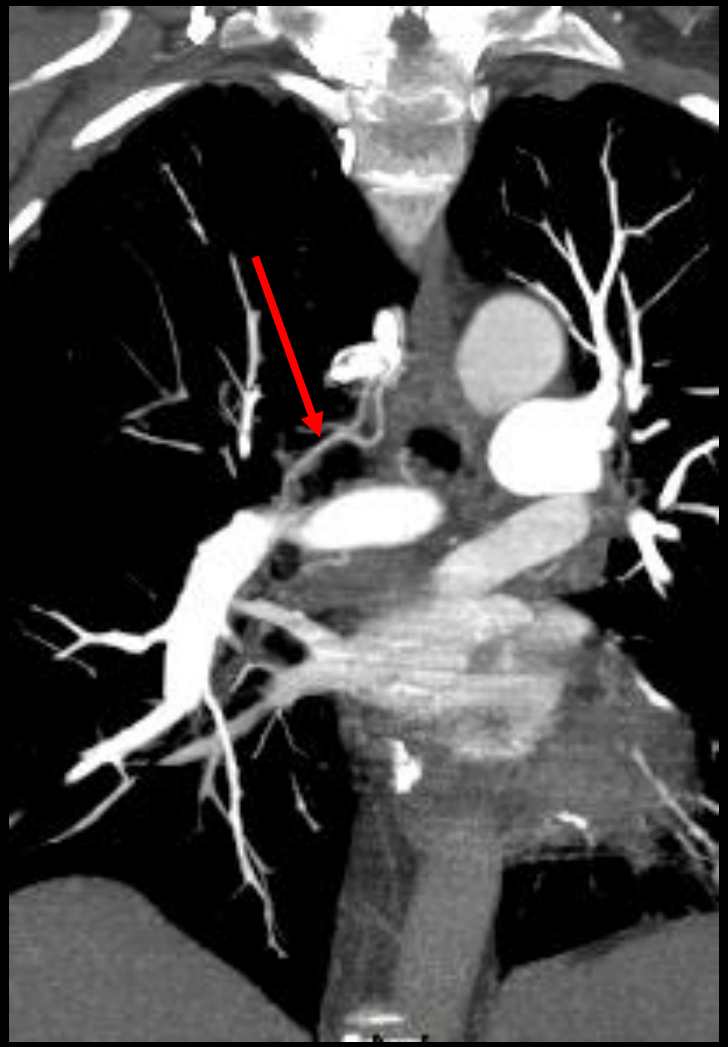


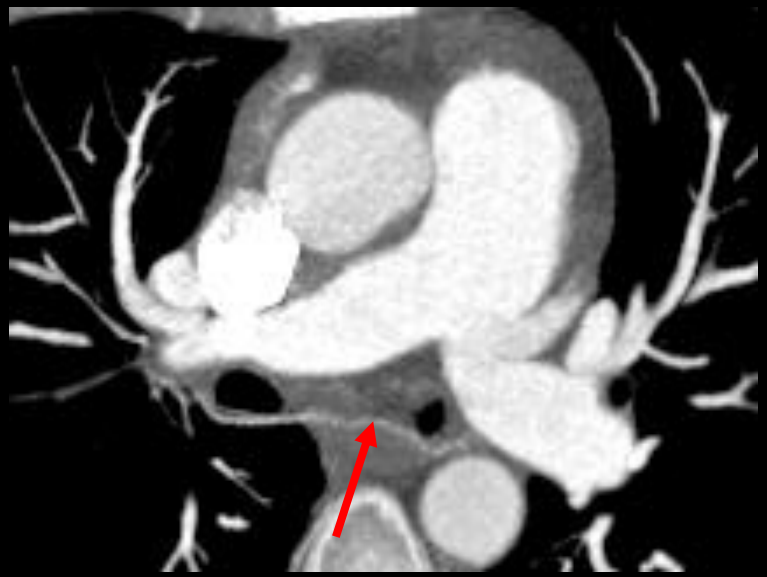
4: CTEPH – CT Features: Systemic Arteries

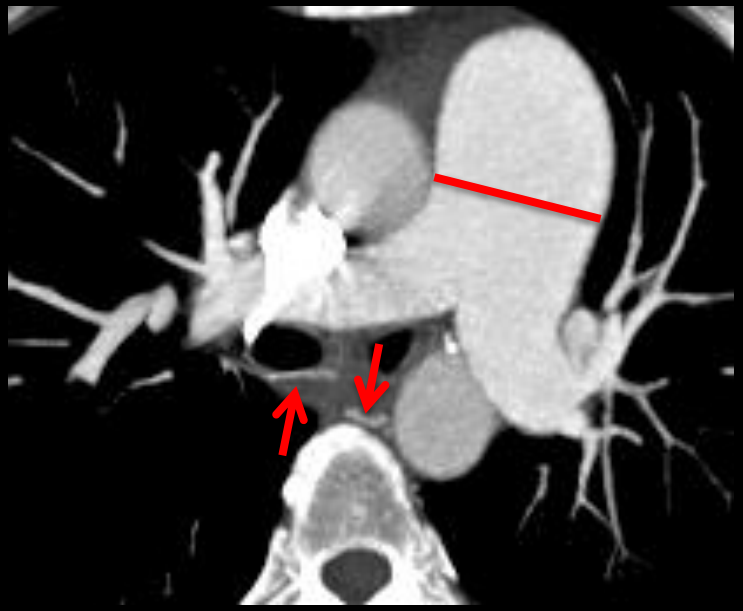
- Systemic collateral supply
  - Bronchial (47-77%) and non bronchial arteries (45%) dilation
  - Non bronchial systemic collateral (inferior phrenic, intercostal, internal mammary arteries)
  - Seen in 73% patients with CTEPH >< 14% with idiopathic PAH:
    - ➢ Could be a useful for discriminating Group 1 *vs*. 4 patients
  - Dilated bronchial arteries are positively correlated with a lower mortality after TEA
  - Responsible for hemoptysis

Grosse et al. Radiographics 2010
Castaner et al. Radiographics 2009
Remy-Jardin et al. Radiology 2005

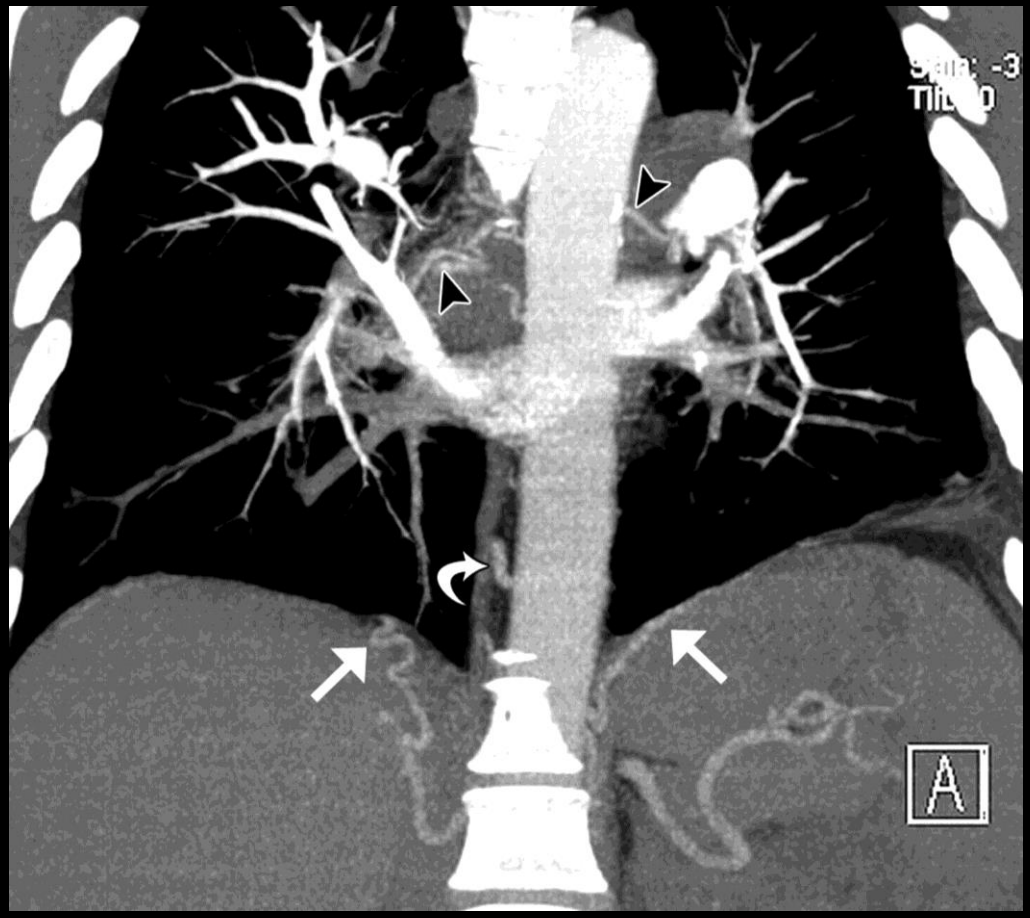
4: CTEPH – CT Features: Systemic Arteries







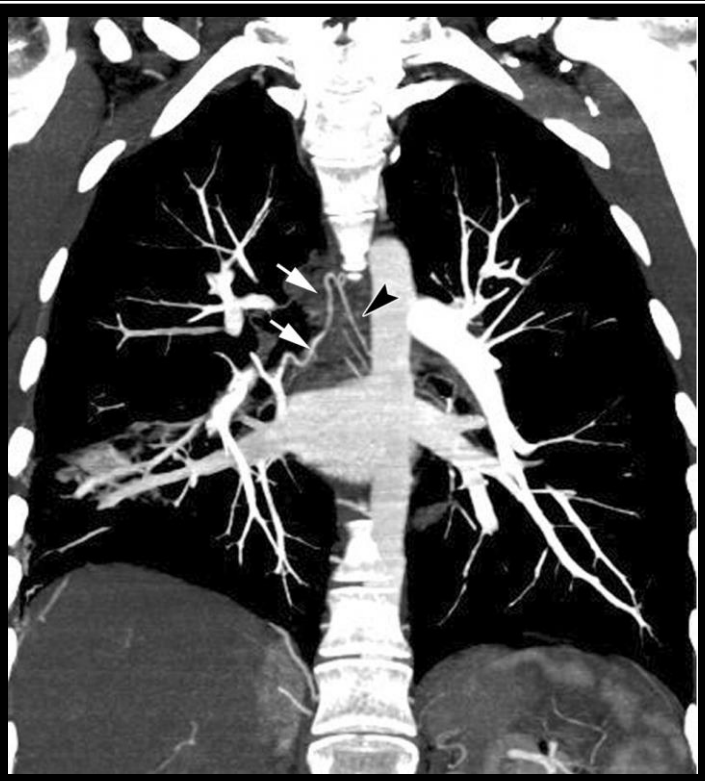
4: CTEPH – CT Features: Systemic Arteries

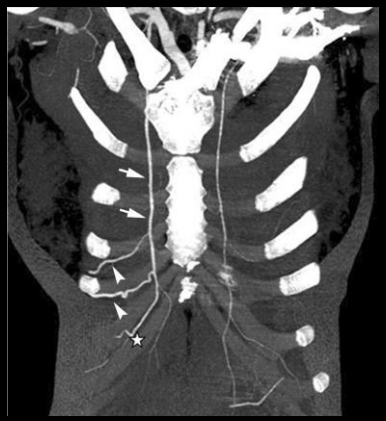


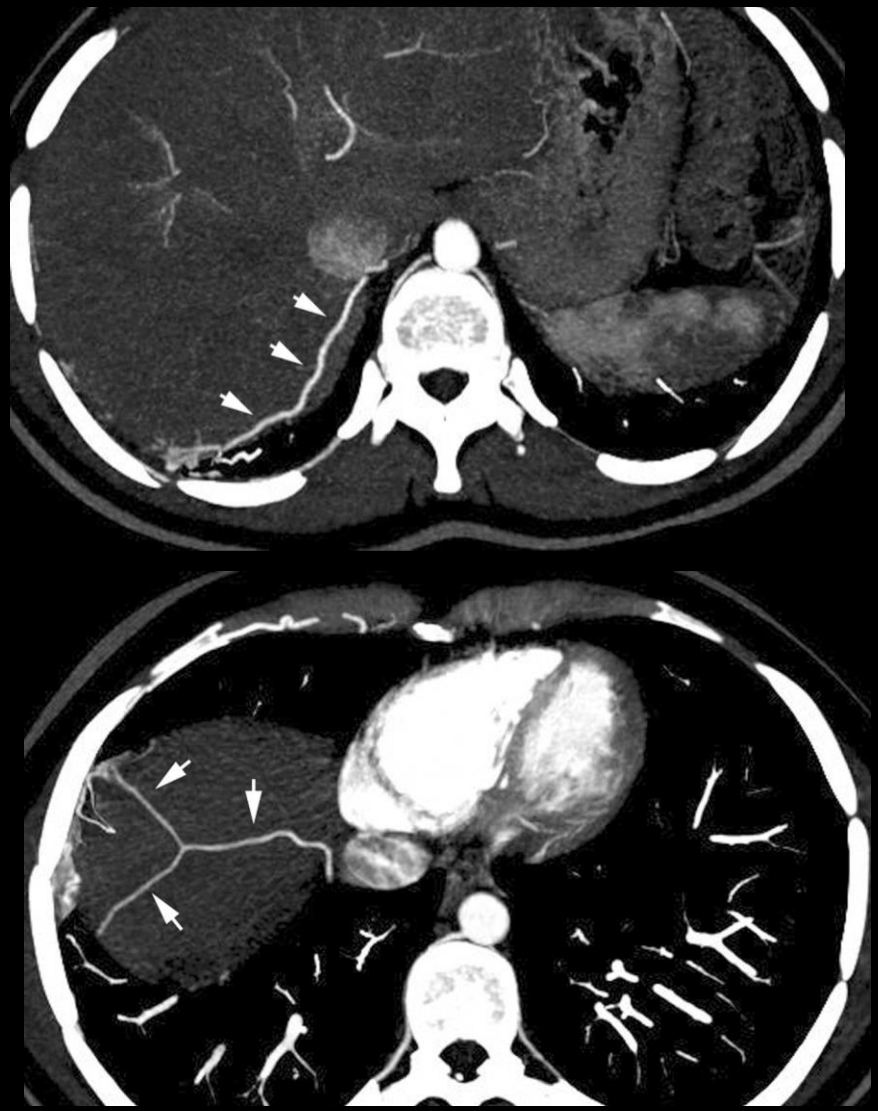
Chronic pulmonary thromboembolism in a 47-year-old man with multiple episodes of acute pulmonary thromboembolism. Coronal 30-mm-thick maximum intensity projection CT image shows marked enlargement of branches of the right and left inferior phrenic arteries (straight arrows), right and left bronchial arteries (arrowheads), and an intercostal artery (curved arrow).

Castaner et al. Radiographics 2009

4: CTEPH – CT Features: Systemic Arteries







Remy-Jardin et al. Radiology 2005

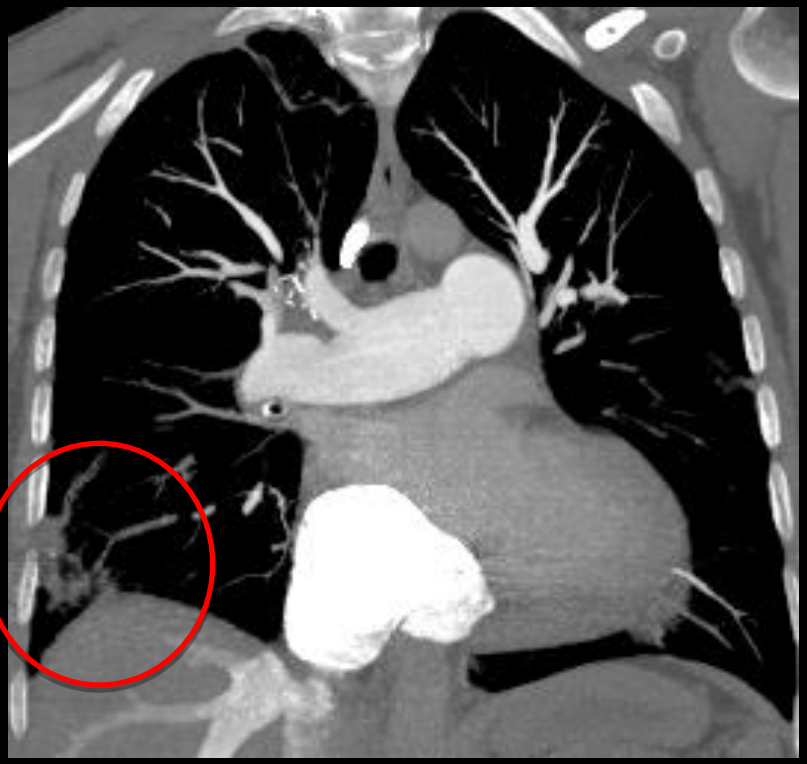
4: CTEPH – CT Features: Lungs

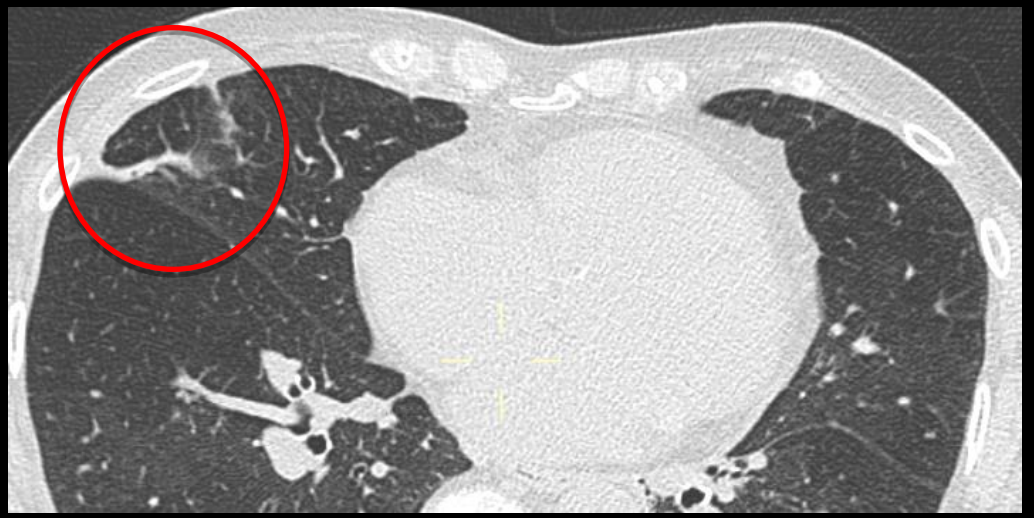
- Scars from prior pulmonary infarctions : often multiples, generally in lower lobes, accompanied by pleural thickening

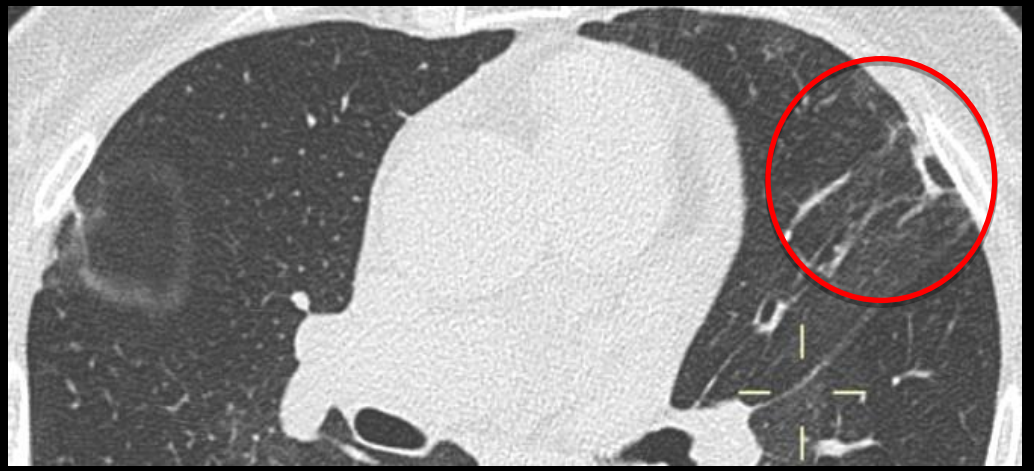
  - Parenchymal bands
  - Wedge-shape pleura-based opacities
  - Peripheral nodules
  - Cavities
  - Irregular peripheral opacities

Grosse et al. Radiographics 2010
Kim et al. AJR 1998
Castaner et al. Radiographics 2009

4: CTEPH – CT Features: Systemic Arteries





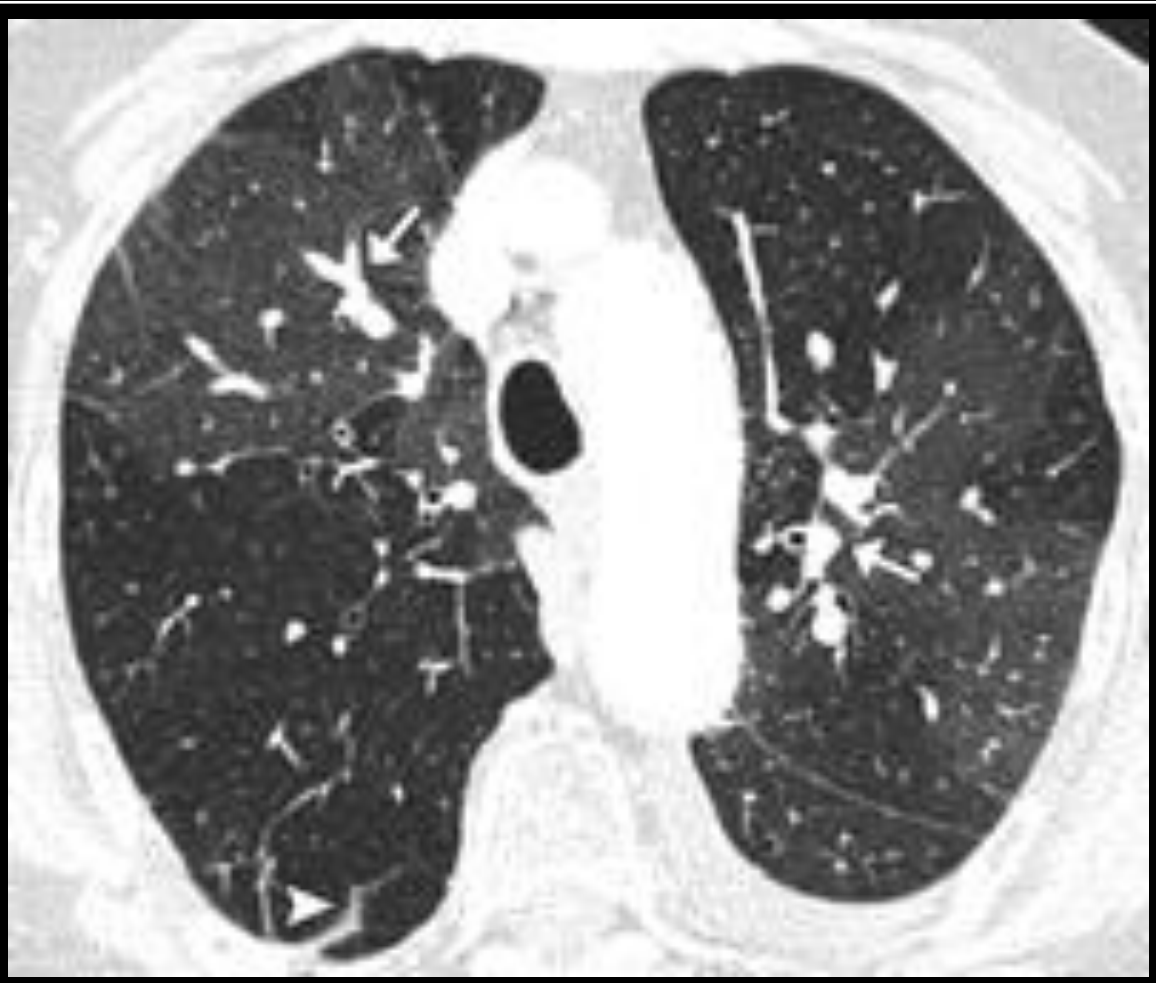


4: CTEPH – CT Features: Lungs

- Mosaic lung attenuation
  - Sharply demarcated areas of hypoattenuation with reduced vessel size, with adjacent areas of normal or relative hyperattenuation
  - Hypoattenuation < hypoperfusion in lung areas distal to occluded vessels or by small-vessel arteriopathy in non obstructed lung areas
  - Hyperattenuation < increased blood flow
  - 77–100% patients with CTEPH
  - Distribution typically segmental or subsegmental

Grosse et al. Radiographics 2010
Castaner et al. Radiographics 2009
Kim et al. AJR 1998
Sherrick et al. AJR 1997

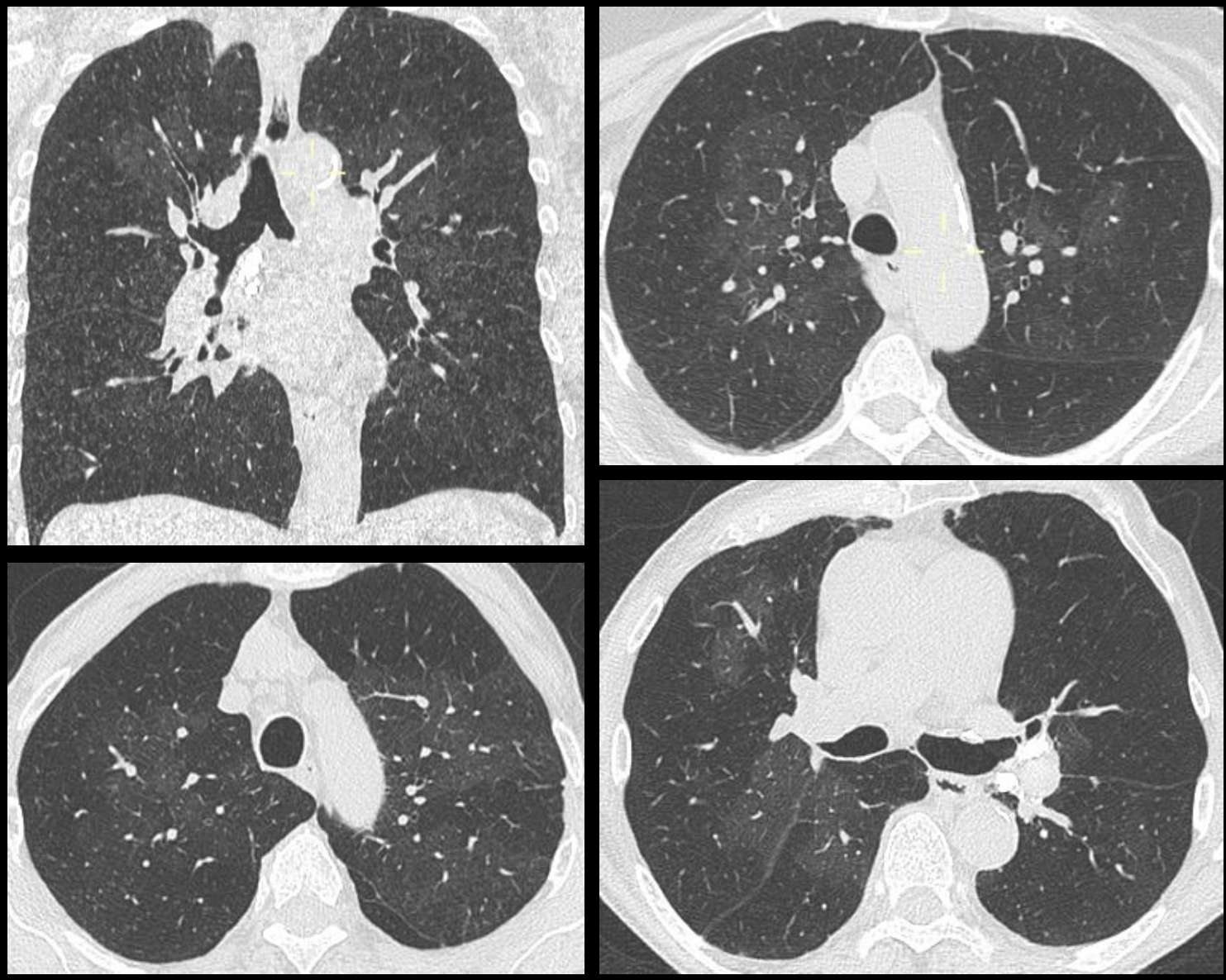
4: CTEPH – CT Features: Lungs



Chronic pulmonary thromboembolism in a 65-year-old man. CT scan (lung window) shows a mosaic perfusion pattern with marked regional variations in attenuation of the lung parenchyma and disparity in the size of the segmental vessels, with larger-diameter vessels in regions of increased attenuation (arrows). A peripheral parenchymal band or scar (arrowhead) from infarction also is depicted.

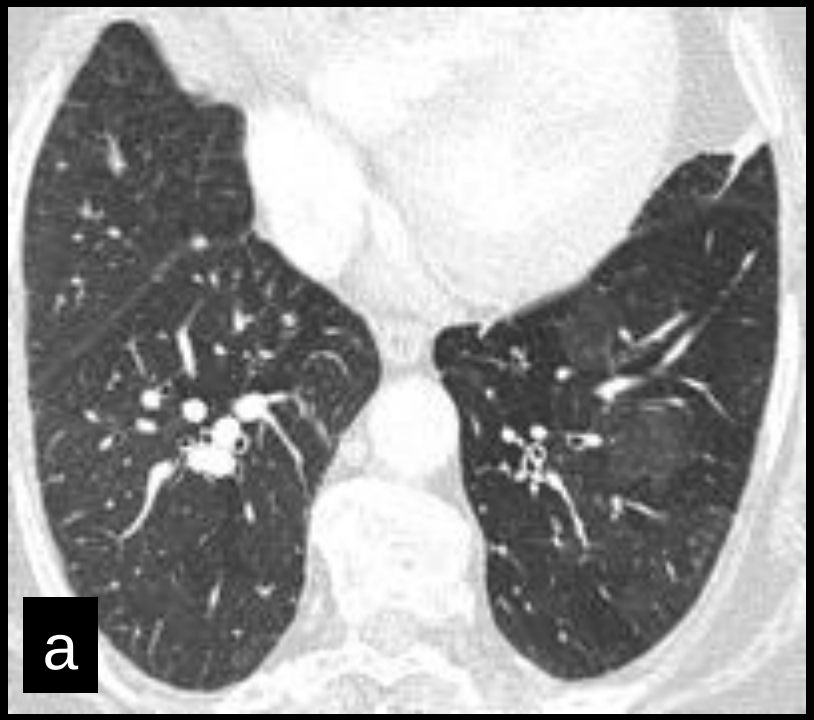
Castaner et al. Radiographics 2009

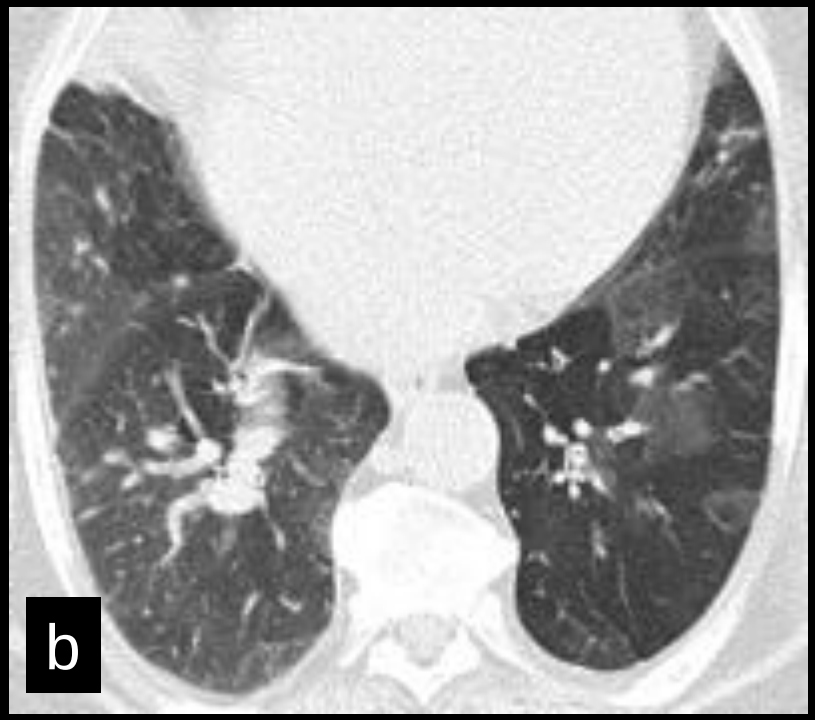
4: CTEPH – CT Features: Lungs



4: CTEPH – CT Features: Airways

- Air trapping on expiratory CT in areas of hypoperfusion



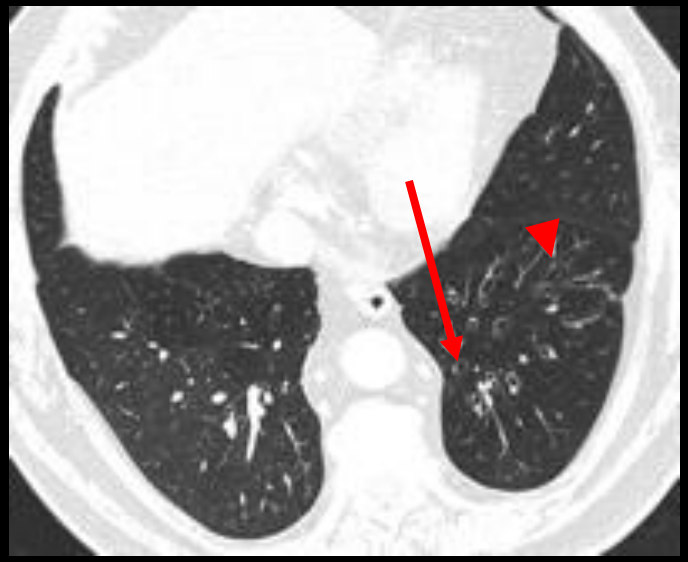


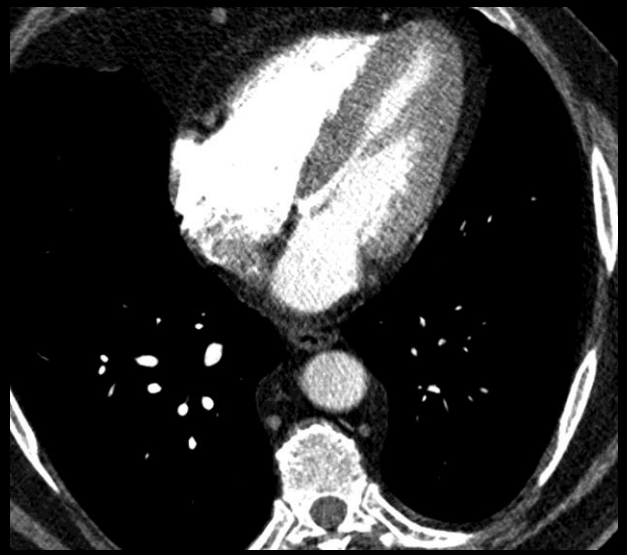
Chronic pulmonary thromboembolism in an 82-year-old woman. **(a)** Axial inspiratory CT scan (lung window) shows chronic thromboembolism of the left lower lobe pulmonary arteries. Note the mosaic perfusion pattern and the diminished size of vessels in this lobe compared with the right lower lobe. **(b)** Axial expiratory CT scan (lung window) at the same level as **a** shows evidence of air trapping in areas of lower attenuation. Air trapping is not specific to airway disease and may be a sign of chronic thromboembolism.

Arakawa et al. J comput Assist Tomogr 2003

4: CTEPH – CT Features: Airways

- Air trapping on expiratory CT in areas of hypoperfusion
- Cylindrical bronchial dilatation: 64% of cases
  - Predominantly lower lung zones
  - Segmental or subsegmental bronchi
  - In areas of severely stenosed or completely occluded pulmonary arteries



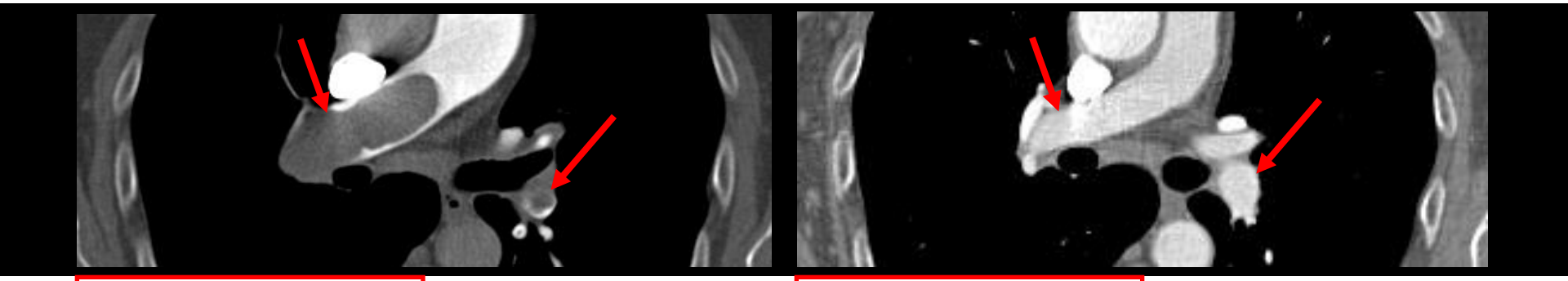


CTEPH in an 82-year-old woman. Axial CT scan (lung window) shows increased bronchial diameters and an absence of normal distal tapering of the segmental and subsegmental bronchi of the left lower lobe (arrow). Note the small arterial segments (arrowhead) at the lateral border of each dilated bronchus.

Castaner et al. Radiographics 2009
Remy-Jardin et al. Radiology 1997

4: CTEPH – Treatment

- Thromboendarterectomy (TEA) is the only curative treatment
- Clinical reference center
- Feasibility of performing TEA depends on
  - Location of obstruction (central vs more distal pulmonary arteries)
  - Correlation between hemodynamic findings and the degree of mechanical obstruction assessed by angiography
  - Comorbidities
  - Willingness of the patient
  - Experience of the surgeon



Pre -TEA

Post -TEA

Simonneau et al. JACC 2009

Hemodynamic Definitions (Nice)

Definition	Characteristics	Clinical group(s)	
Pulmonary hypertension (PH)	Mean PAP ≥ 25 mmHg	All	
Pre-capillary PH	Mean PAP ≥ 25 mmHg	1. Pulmonary arterial hypertension	✓
	PWP ≤ 15 mmHg	3. due to lung diseases	✓
	CO normal or reduced	4. Chronic Thromboembolic PH	✓
		5. PH with unclear and/or multifactorial mechanisms	
Post-capillary PH	Mean PAP ≥ 25 mmHg	2. PH due to left heart disease	✓
	PWP > 15 mmHg	5. PH with unclear and/or multifactorial mechanisms	
	CO normal or reduced		
Passive	TPG ≤ 12 mmHg		
Reactive (out of proportion)	TPG > 12 mmHg		

PAP = pulmonary arterial pressure, PWP = pulmonary wedge pressure, CO = cardiac output,

TPG = Transpulmonary pressure gradient

2015 ESC/ERS Guidelines. *Eur Haert J*

Definitions

- PH can be found in multiple clinical conditions: 5 Groups
  1. Pulmonary arterial hypertension (PAH) ✓
  2. PH due to cardiac disease ✓
  3. PH due to chronic lung disease and/or hypoxia ✓
  4. Chronic thromboembolic PH (CTEPH) ✓
  5. **PH due to unclear multifactorial mechanisms**

Simonneau et al. JACC 2013

5: PH due to unclear multifactorial mechanisms

Hôpital Erasme

ULB

- Hematologic disorders
  - Chronic hemolytic anemia, myeloproliferative disorders, spleenectomy
- Systemic disorders
  - Sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- Metabolic disorders
  - Glycogen storage disease, Gaucher disease, thyroid disorders
- Others
  - Tumor obstruction, fibrosing mediastinitis

Definitions

- PH can be found in multiple clinical conditions: 5 Groups

1. Pulmonary arterial hypertension (PAH) ✓
2. PH due to cardiac disease ✓
3. PH due to chronic lung disease and/or hypoxia ✓
4. Chronic thromboembolic PH (CTEPH) ✓
5. PH due to unclear multifactorial mechanisms ✓

Simonneau et al. JACC 2013

What the Clinician Needs to Know?

Hôpital Erasme

ULB

1. To rule out a pulmonary disease (UIP, severe emphysema, …)
2. To confirm indirect imaging features suggesting PH
3. To rule out CTEPH / extension - location
4. To raise suspicion of PVOD (PH + pulmonary edema)
5. In case of unexplained dyspnea, to suggest the diagnostic directions for PH

Suggested Readings

- Simonneau G. *J Am Coll Cardiol* 2013; 62: D34–41
- Hoeper MH. *J Am Coll Cardiol* 2013; 62: D42–50
- Peña H. *Radiographics* 2012; 32: 9–32
- Frazier AA, Burke AP. *Semin Ultrasound CT MRI* 2012; 33: 535-551
- 2015 ESC/ERS Guidelines. *Eur Haert J* Epud ahaed of print