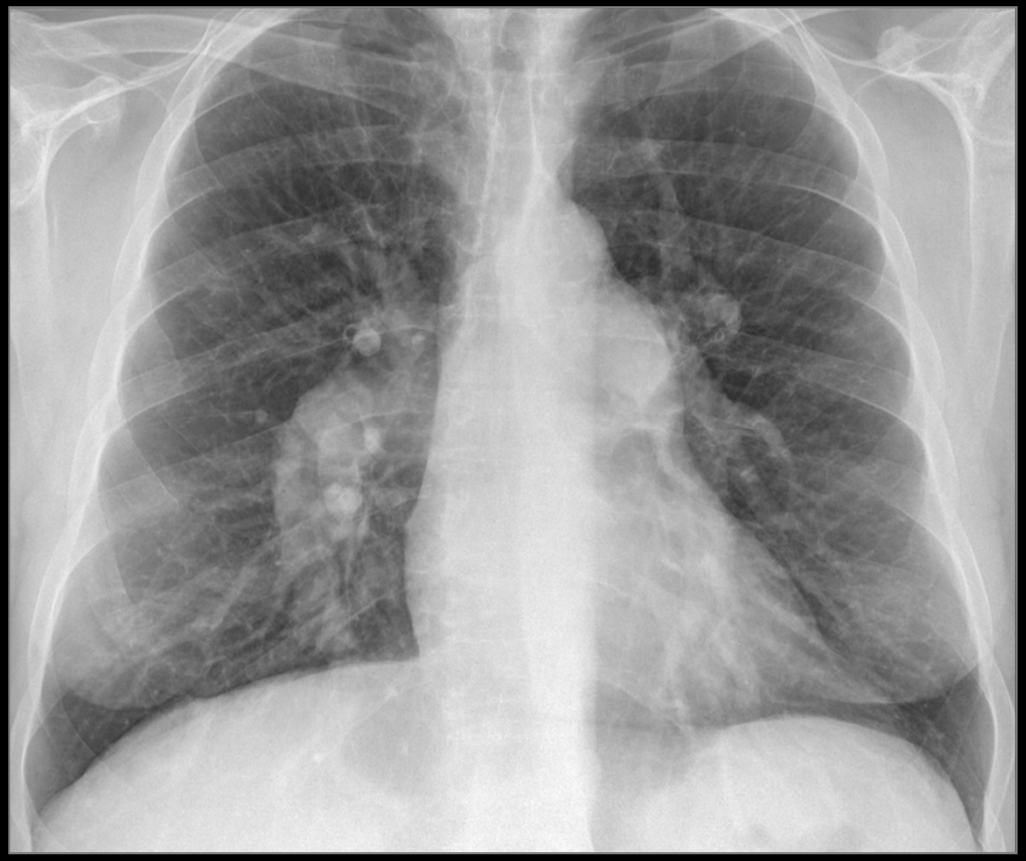
IMAGING OF PULMONARY HYPERTENSION

WITH PARTICULAR EMPHASIS ON CT ANGIOGRAPHY

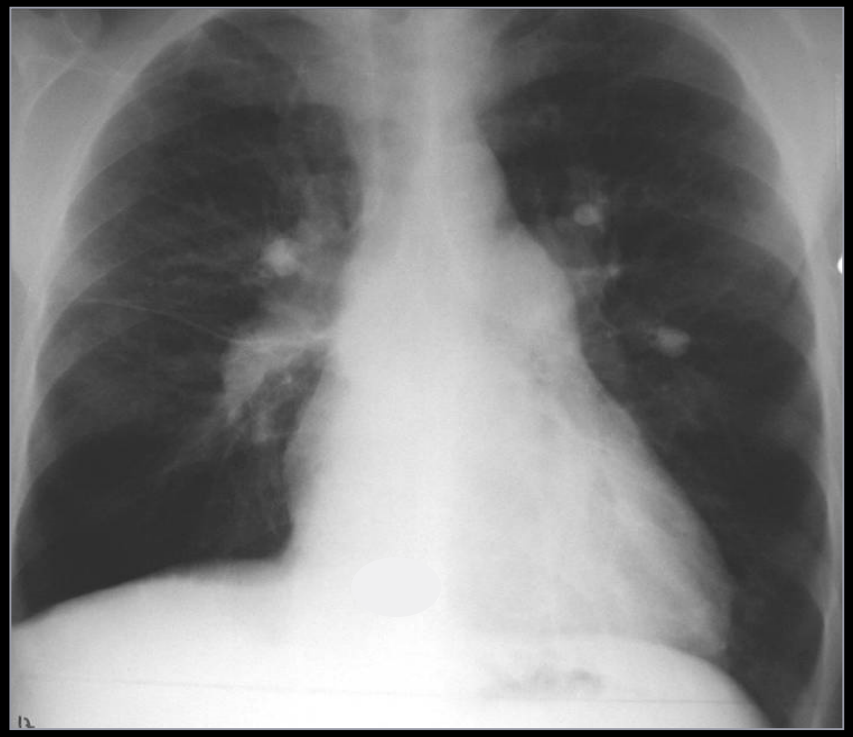
M. ZOMPATORI

DETECTION



Chest X ray is abnormal in 90% of the patients with idiopathic PH at the time of diagnosis, showing:

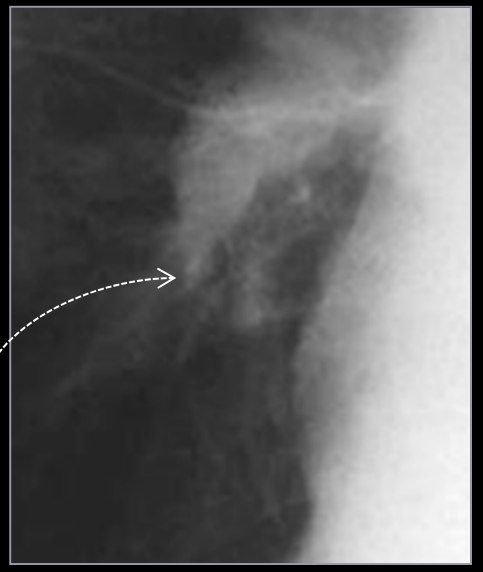
- dilatation of the central pulmonary arteries (60%)
- pruning (barrage) of the peripheral arteries, peripheral oligemia (60%)
- in advanced cases, enlargement of RV and RA (70%) and
- dilatation of the azygos vein (30%)



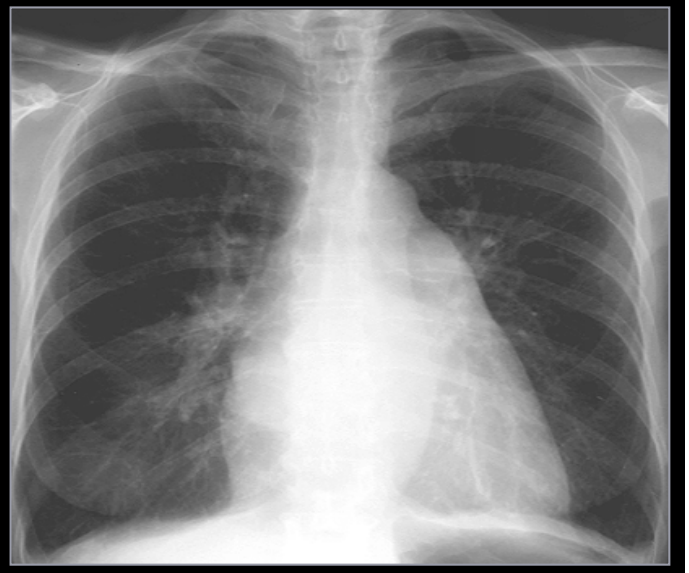
The degree of hypertension in any given patient does not correlate exactly with the extent of radiographic abnormalities and chest X ray is not sensitive for detecting mild cases

Cut-off value: 15 mm

PRUNING

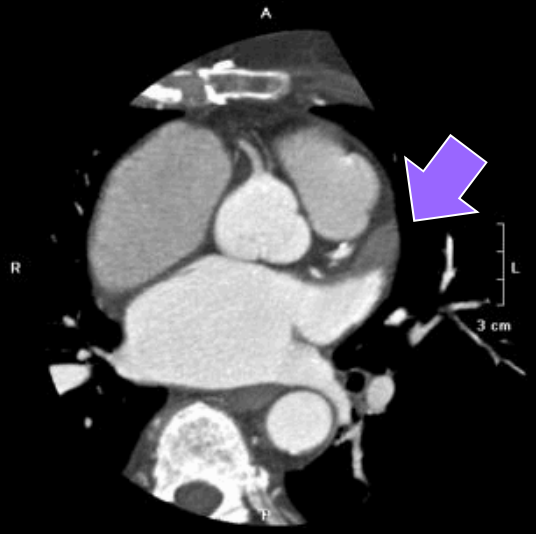


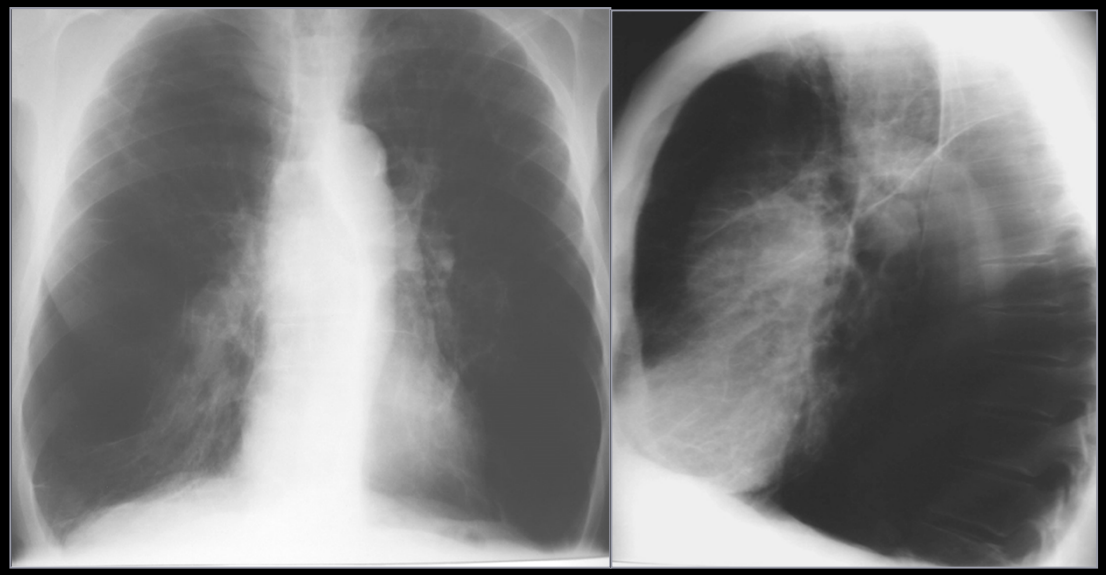
DETECTION

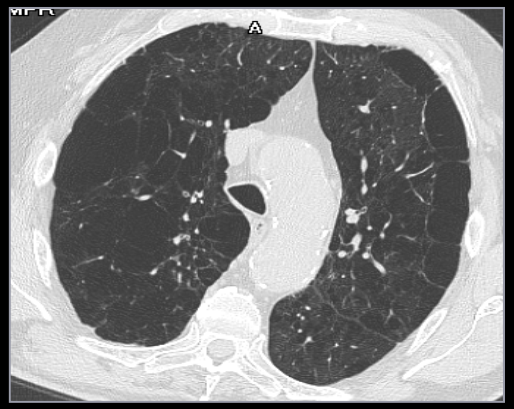


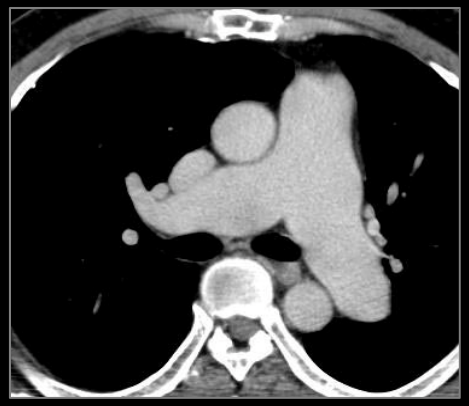
Chest X ray may also exclude or suggest an underlying cause, for ex by showing severe parenchymal changes or pulmonary venous hypertension due to left heart pathology.

Left heart disease is the most common cause of PH (ischemic, valvular, structural). 100% in case of severe left-heart valvular disease. Pay attention to the size of the left atrium.





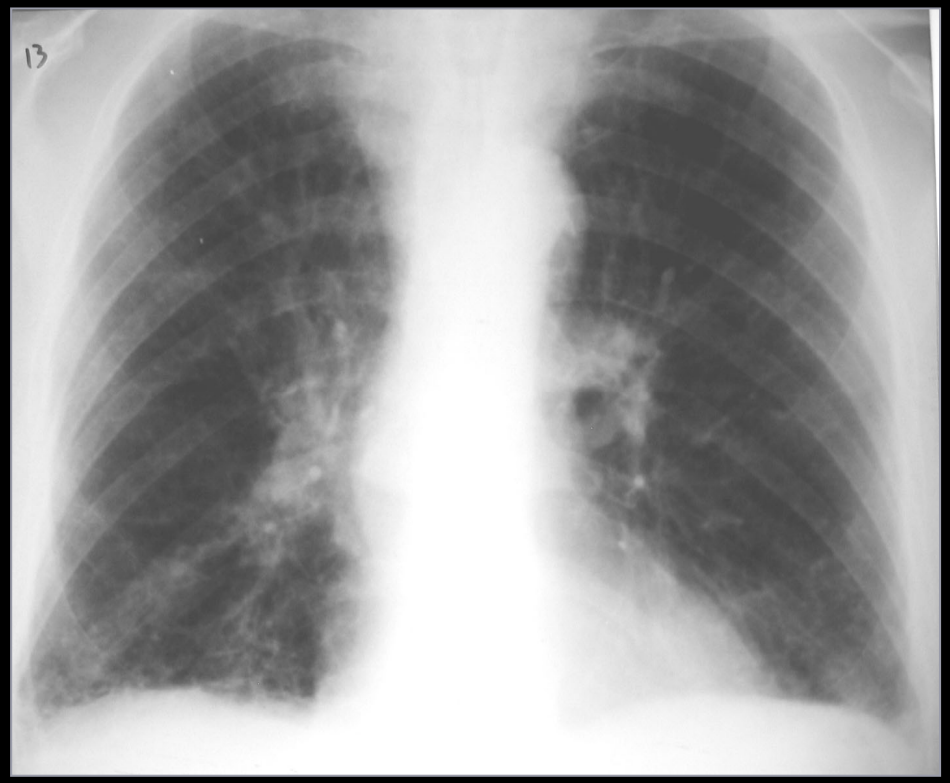


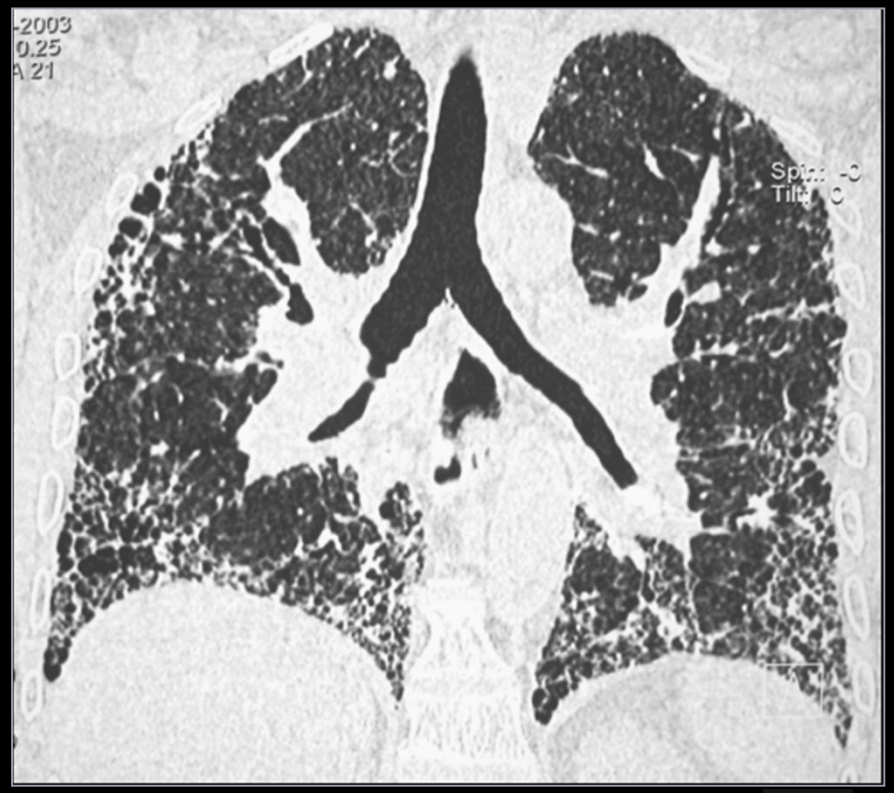


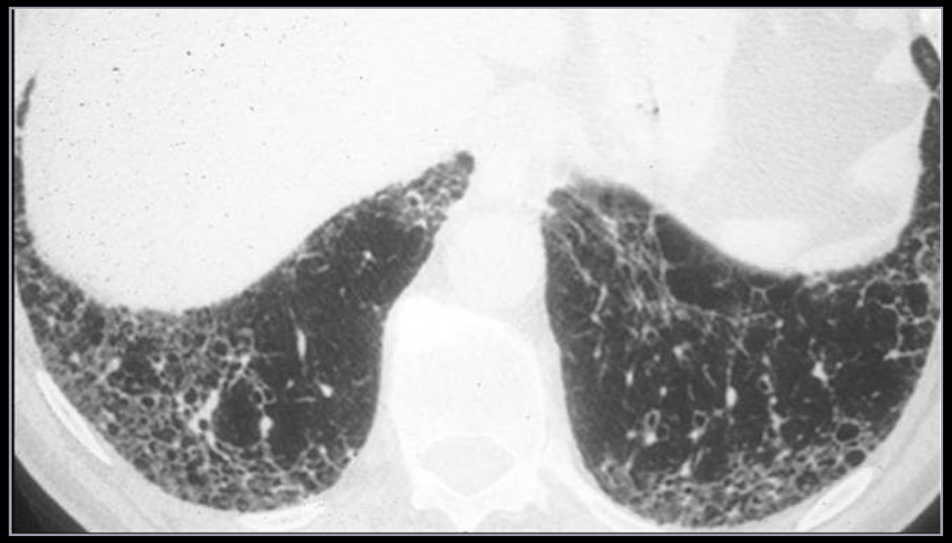
COPD is the 2° most common cause of hypertension. 50% of patients with severe COPD have a (generally mild) hypertension.

Pulmonary artery dilatation is a useful predictor of PH in COPD patients and is an indipendent predictor of mortality.

Recently, a **Vascular Phenotype** has been described, characterized by mild COPD but severe PH and very low DL CO.

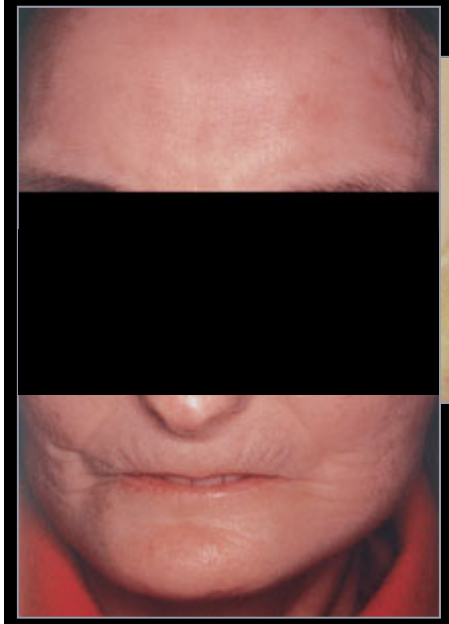


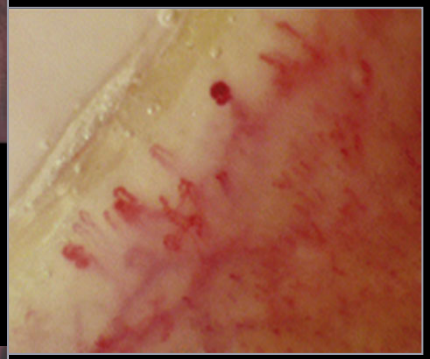


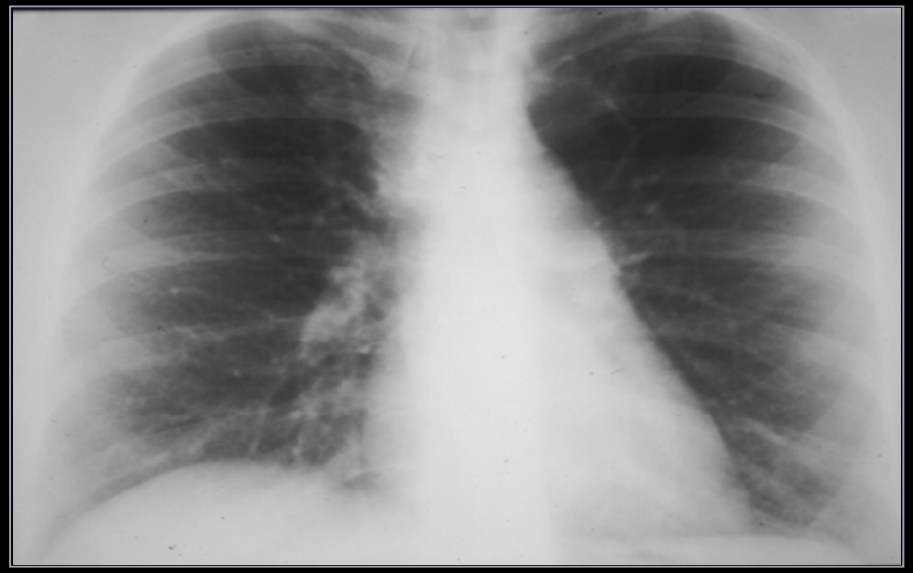


INTERSTITIAL LUNG DISEASES

12% in IPF patients.

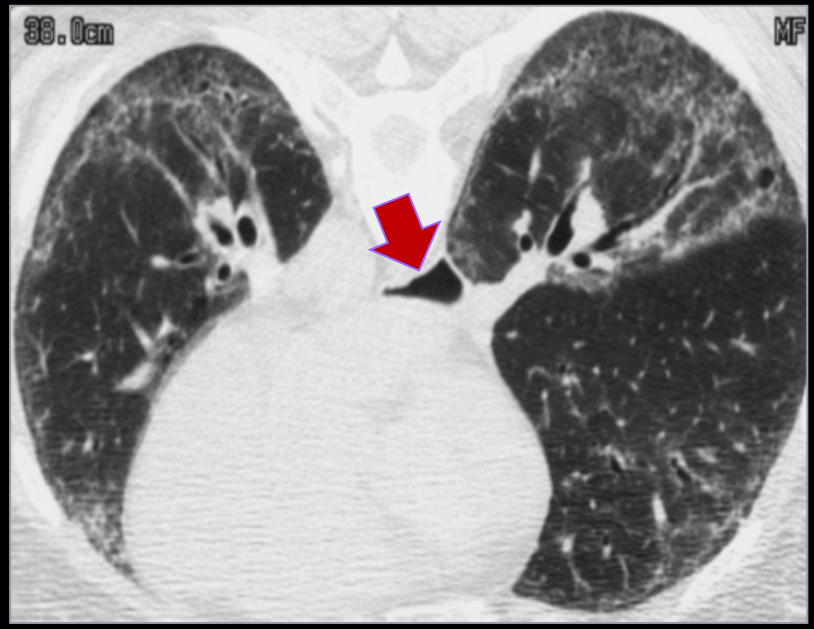




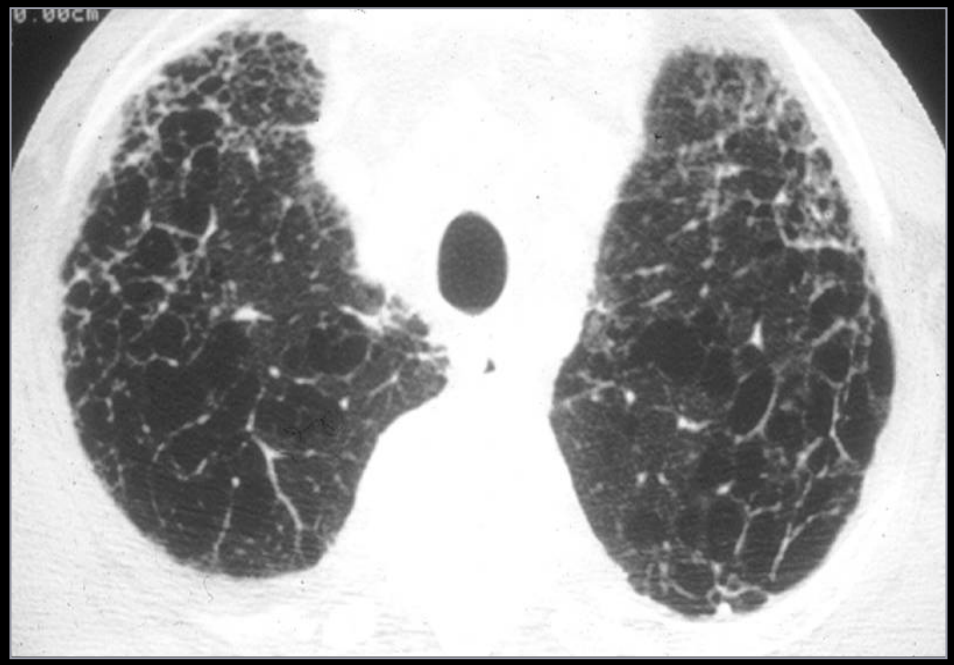


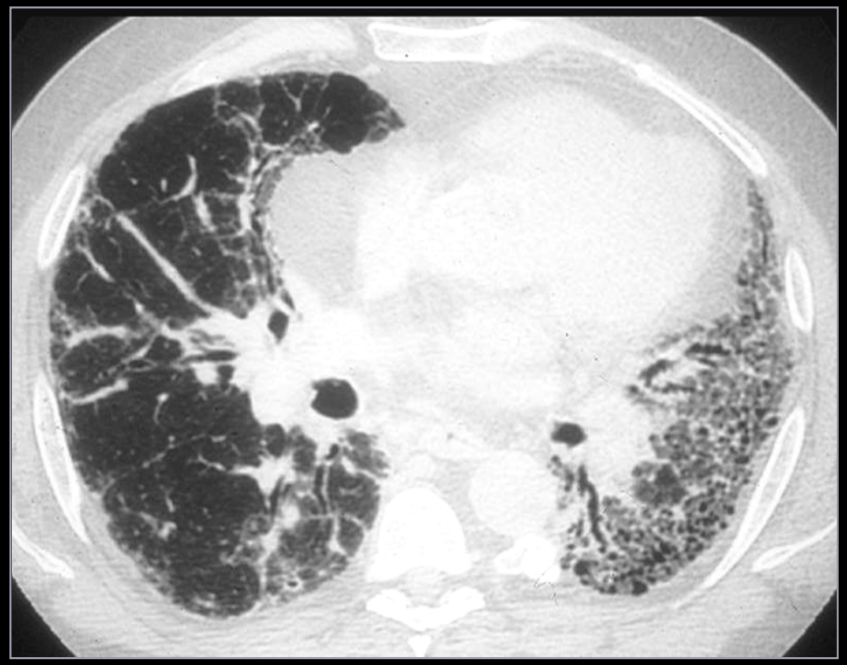
CVD associated PAH represents 25% of PAH cases, with SSc being the most prevalent type. High mortality.

CVD, as well as Sarcoidosis, may show hypertension disproportionate to the severity of lung disease.
Esophageal dilatation is commonly seen in PAH-SSc.
In Sarcoidosis, 50% of patients with PH do not have evidence of pulmonary fibrosis.



Association of fibrosis & emphysema
CPFE





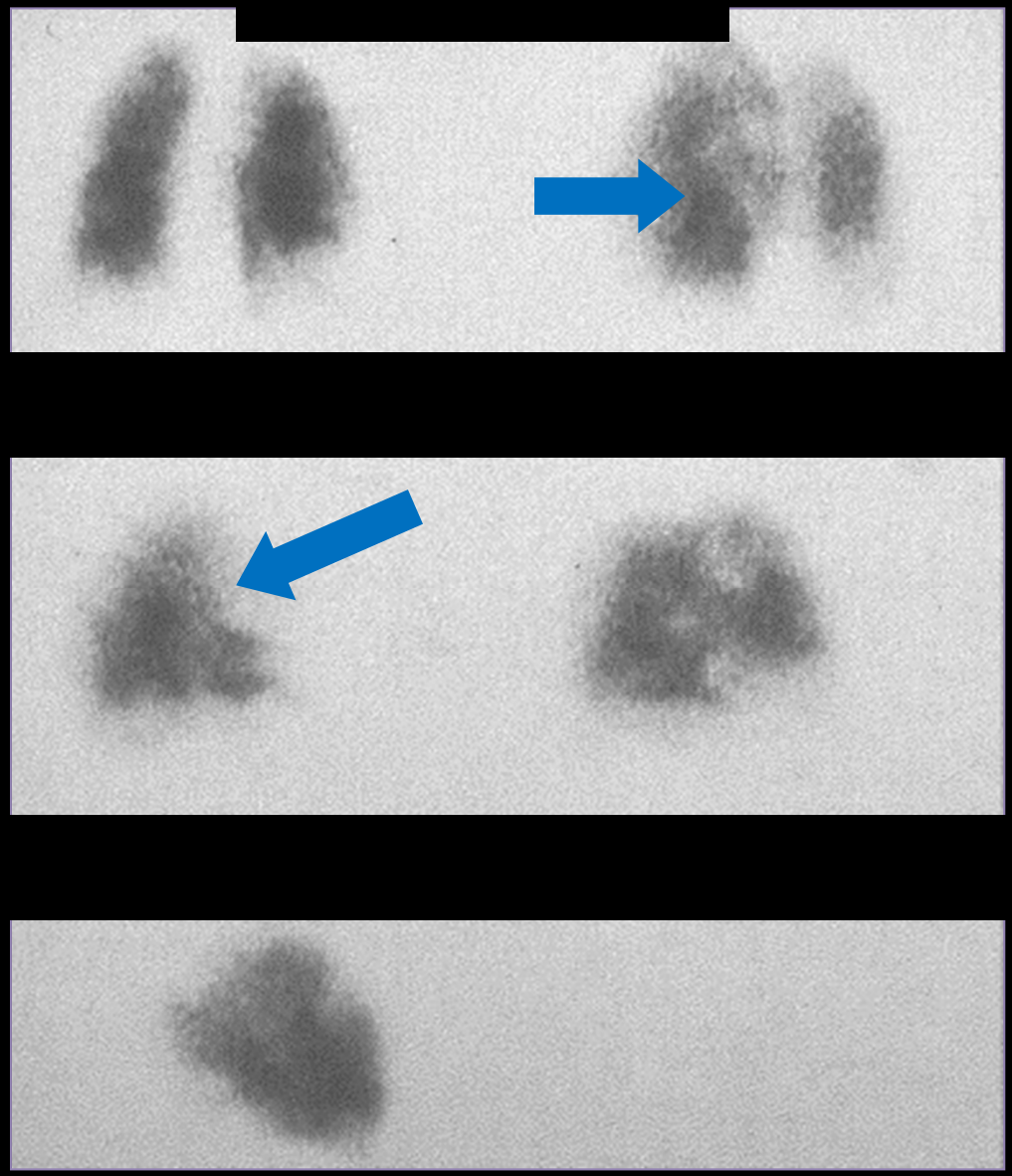
10 % of smokers with a clinical diagnosis of COPD

35% of IPF/UIP patients at presentation

More common (50%) and more severe hypertension, related to the summed baseline extension of ILD & emphysema

High prevalence of lung cancer

CTEPH



V/P lung scan

- PROS: high sensitivity for post-embolic hypertension (CTEPH). High NPV but sometimes in CTEPH scintigraphy demonstrates areas of matched V/Q defects (due to compensatory hypoventilation).

- CONS: false + in a number of different diseases (PA sarcoma, vasculitis, extrinsic compression, mediastinal fibrosis, radiotherapy).
Small peripheral unmatched perfusion defects, with mottled appearance, are also described in idiopathic PAH (iPAH) or PVOD (10%).
The degree of perfusion abnormality can substantially **underestimate** the vascular obstruction and **no DD** is possible between acute and chronic embolism
Cannot determine whether a patient with CTEPH is a good candidate for surgery.

HRCT / multidetector ANGIO-CT WITH CONTRAST INJECTION

CT enables accurate evaluation of central and distal pulmonary arteries down to the 6° order, with high i/o agreement

Sens & spec at the segmental vessels >90%

Can be used instead of scintigraphy for the initial evaluation of suspected CTEPH

PULMONARY HYPERTENSION CT SIGNS

1) VASCULAR SIGNS
   - The main pulmonary artery
   - The other vessels
2) CARDIAC SIGNS
3) PARENCHYMAL SIGNS

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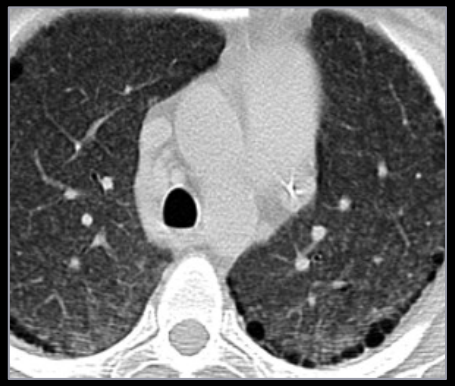
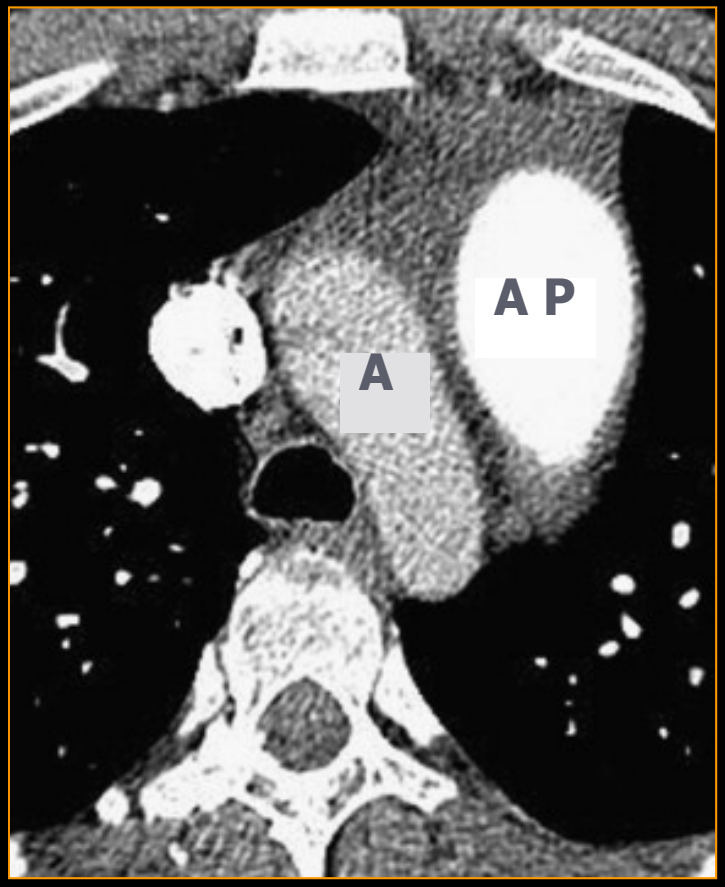
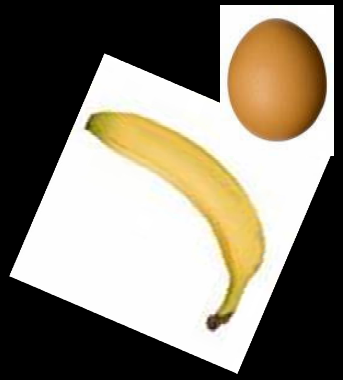






THE EGG & BANANA SIGN

The main PA is visible in the axial view at the level of the aortic arch.

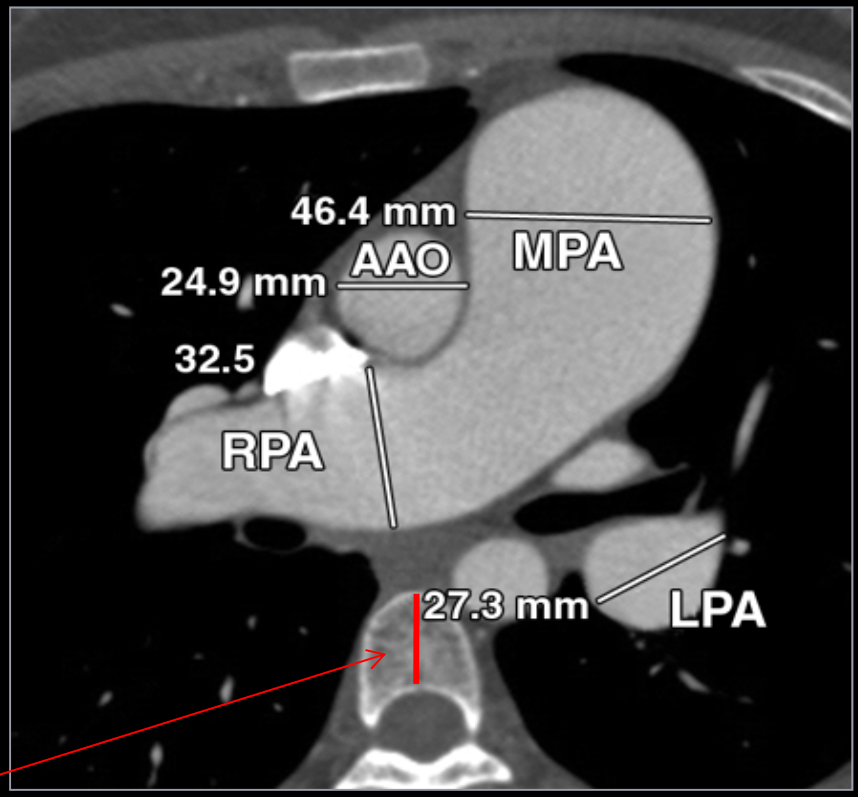
This sign is specific of severe PH.

Main pulmonary artery dilatation (>2,9 cm in men and 2,7 in women) in the detection of hypertension: sens 87%, spec 89% (100% when threshold is 3,5 cm). RPA and LPA>18 mm are abnormal

THE ROLE OF INTERNAL STANDARDS

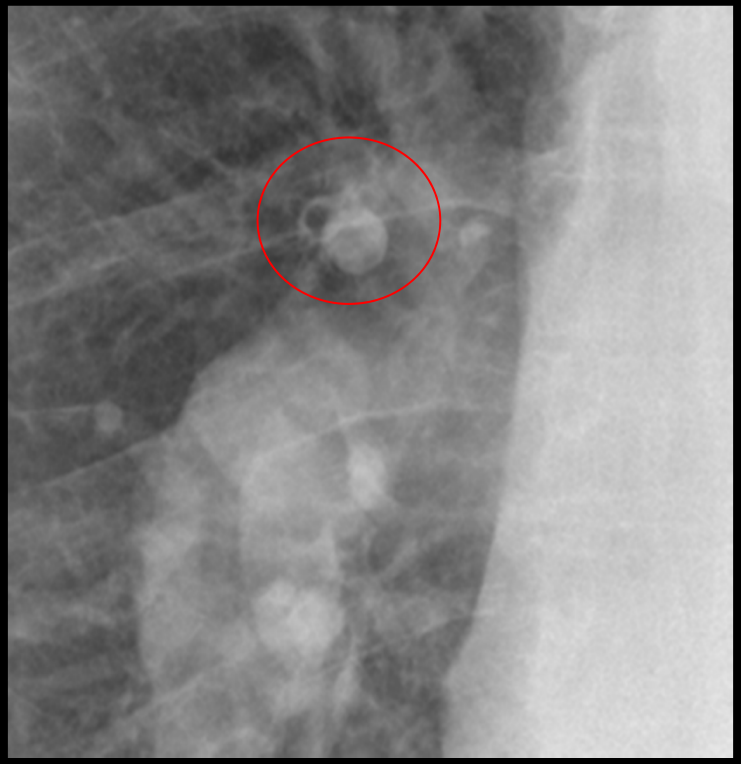
The **PA/AAO ratio > 1**
in individuals less than 50 years old and w/o diffuse pulmonary fibrosis is highly specific for hypertension (spec >90%; PPV 95%; sens 70%). In patients with severe COPD this ratio is more accurate than echocardiography

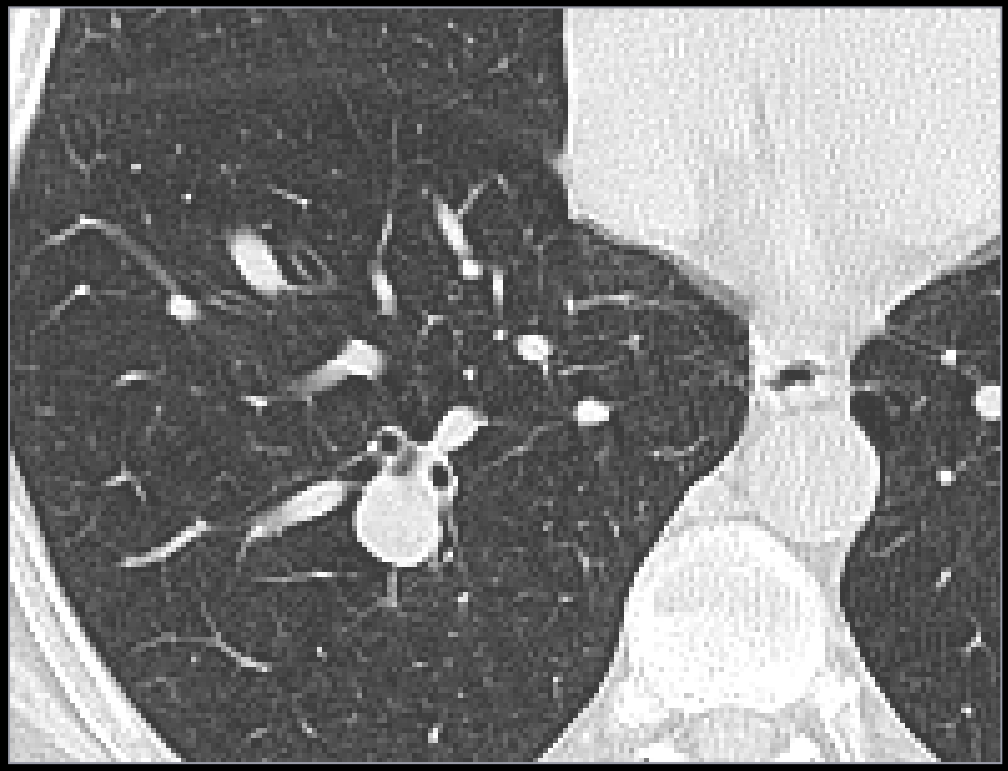
Ratio MPAD/VB >1,5: sens 63%, spec is 93%



When the ascending aorta is dilated, the size of the vertebra can be a useful alternative internal standard

Combination of PA/AAO and echocardiographic measures is more precise than either test alone (composite index)





In the majority of lobes, segmental pulmonary arteries are larger than the associated bronchi

Segmental artery diam/outer bronchus diam ratio > 1,25 indicates pulmonary hypertension with sens 70%, spec >90%. When this finding is associated with MPA dilatation, specificity is 100%.

Notice that in a few normal individuals this ratio may slighly exceed 1 in a single branch, so 3 or more lobes should be evaluated.

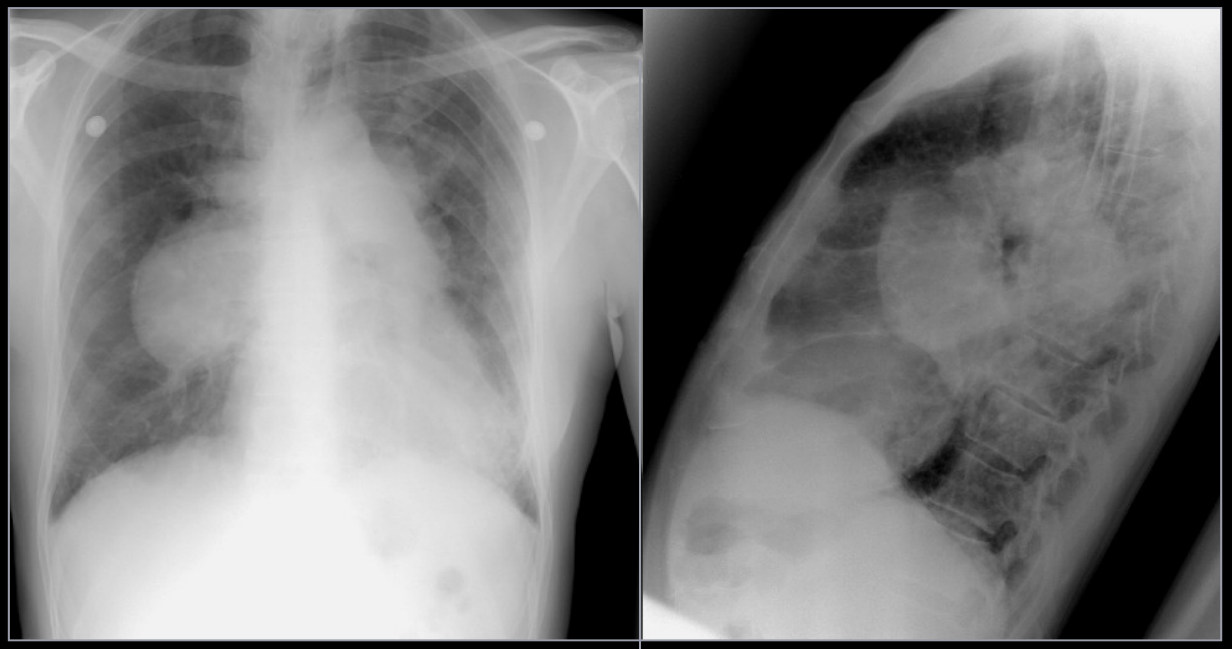
THESE SIGNS ARE MORE SPECIFIC THAN SENSITIVE

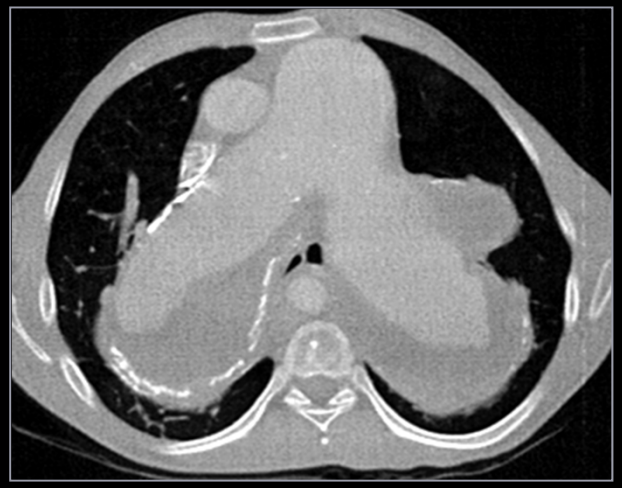
In some cases, CT angiography can demonstrate particular diagnostic features

ex:

Eisenmenger syndrome (congenital defects, atheromatous calcifications, in situ thrombosis, aneurysmal dilatation)

Chronic thromboembolic pulmonary hypertension (CTEPH)

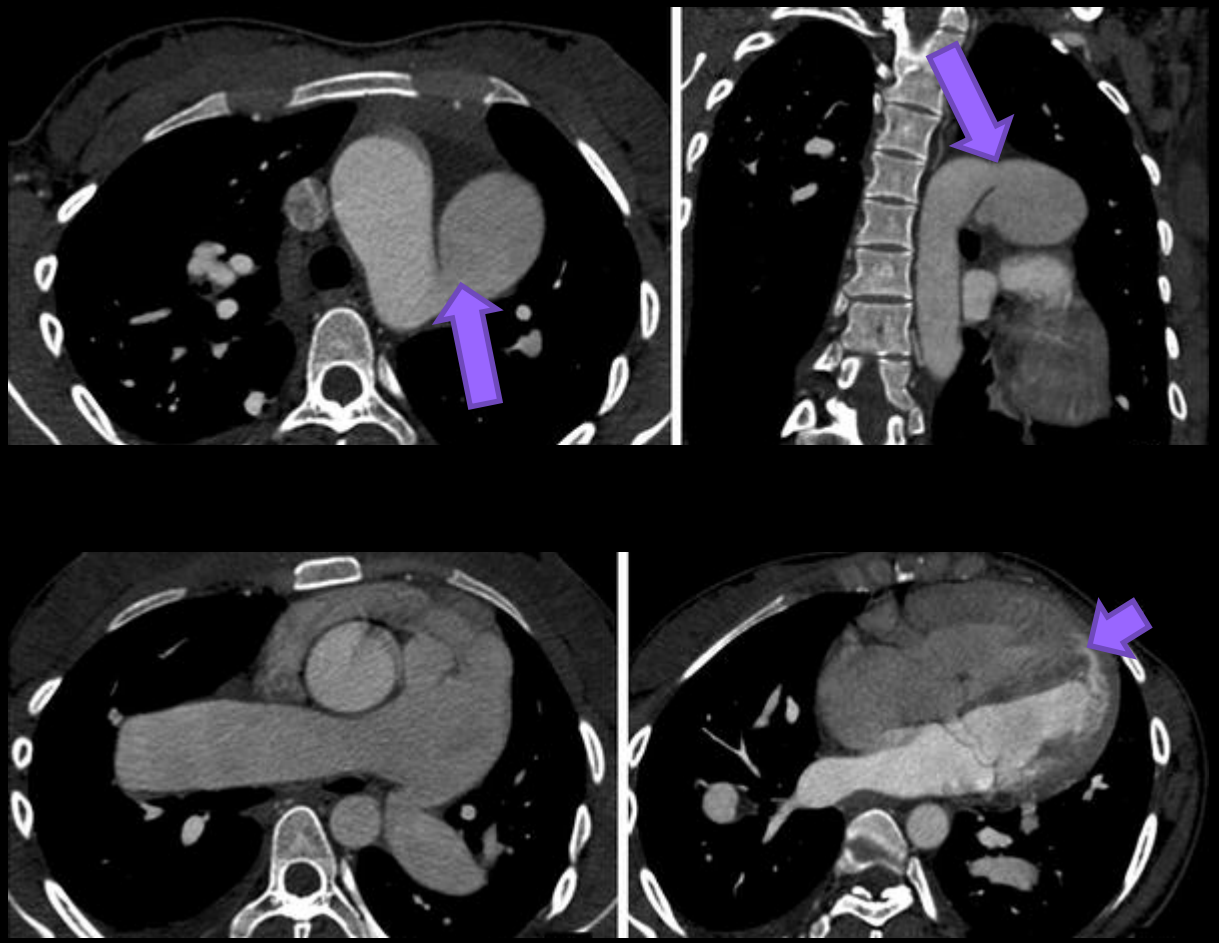




CONGENITAL HEART DISEASES & EISENMENGER SYNDROME

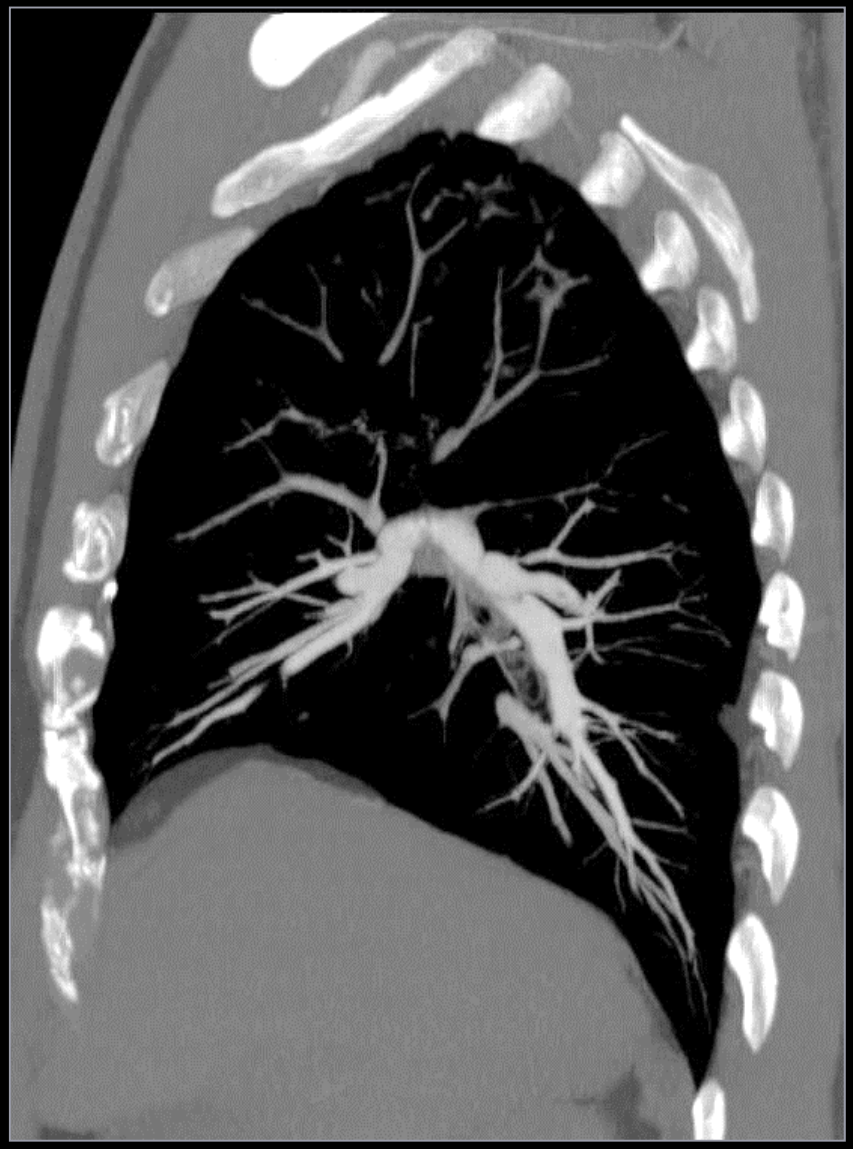
IN 15% ANEURYSMAL DILATATION OF THE CENTRAL PULMONARY ARTERIES

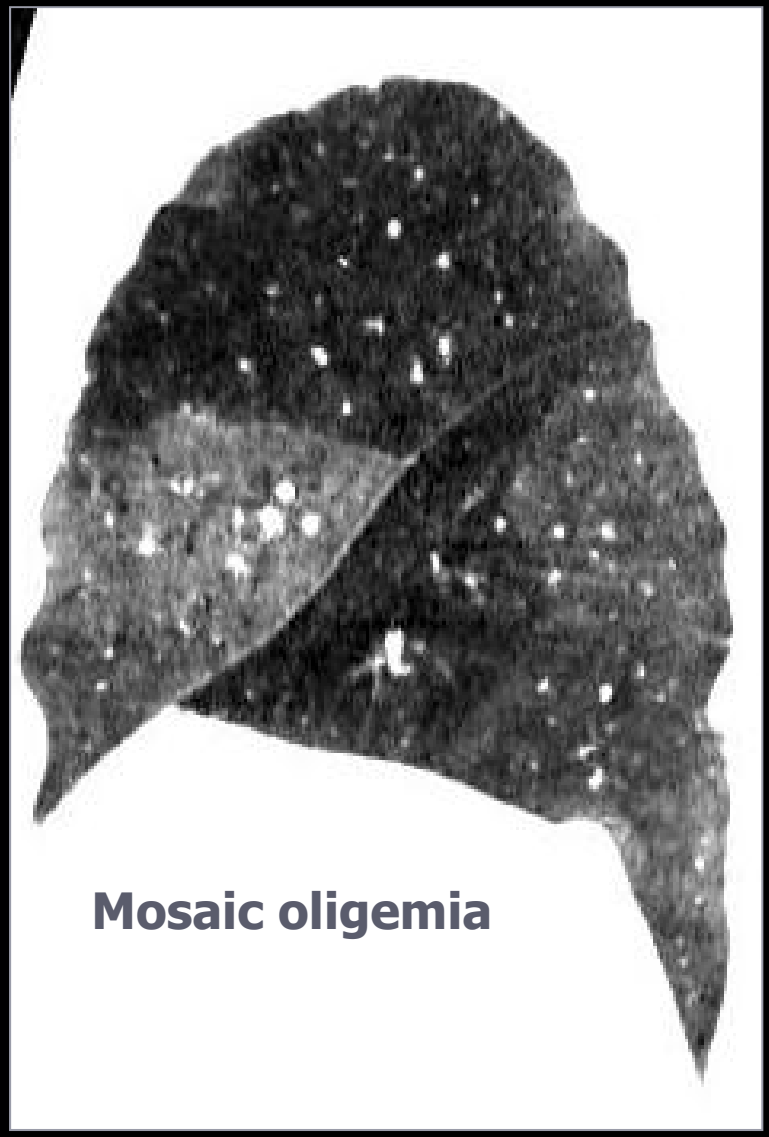
Pulmonary arterial calcifications (15 %) and in situ thrombosis (25%) are seen almost exclusively in PAH-congenital (rare in IPAH). Increased risk of bleeding.
Patients with in situ thrombosis generally do not have segmental perfusion defects on perfusion scintigraphy or mosaic attenuation on HRCT. In situ thrombi: eccentric wall-adherent thrombi with increased vessel size, unlike in cases in chronic embolism where vessel size is reduced. In situ thrombi do not float inside the lumen. No infarcts.



Botallo & DIV

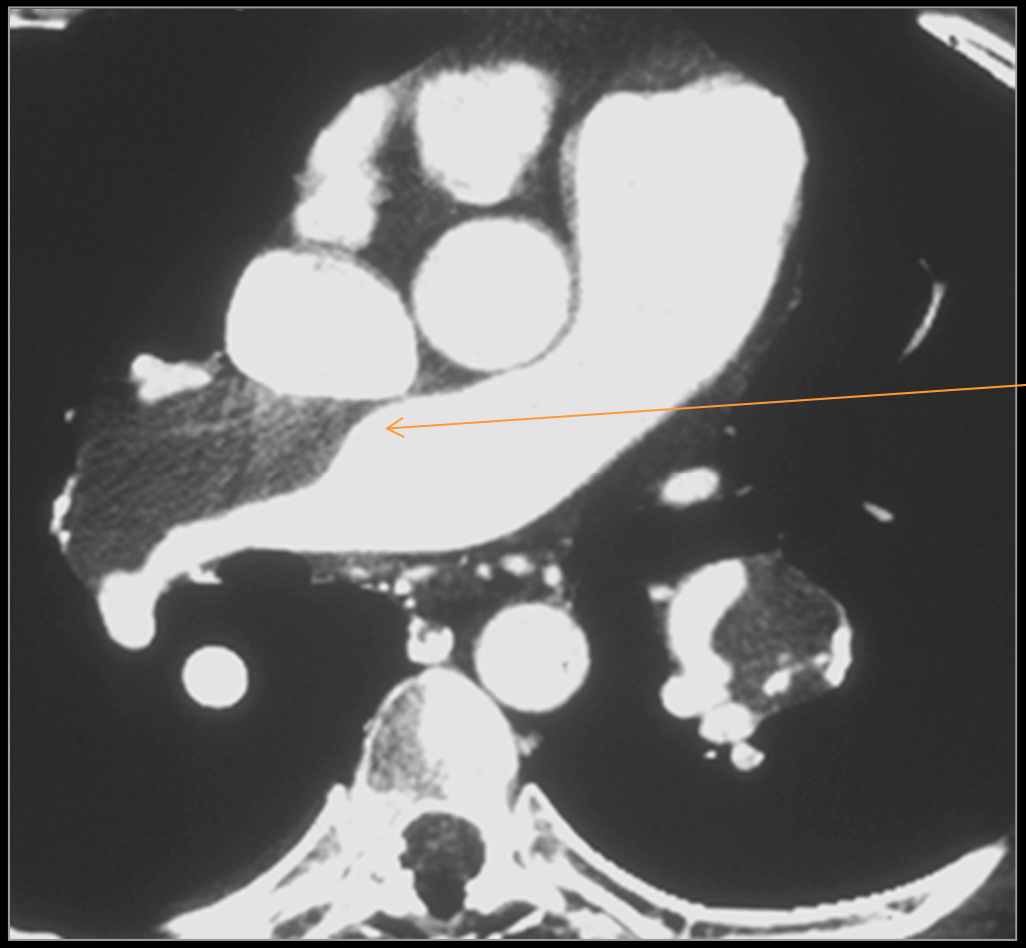
Chronic Thromboembolic Pulmonary Hypertension (CTEPH)





Mosaic oligemia

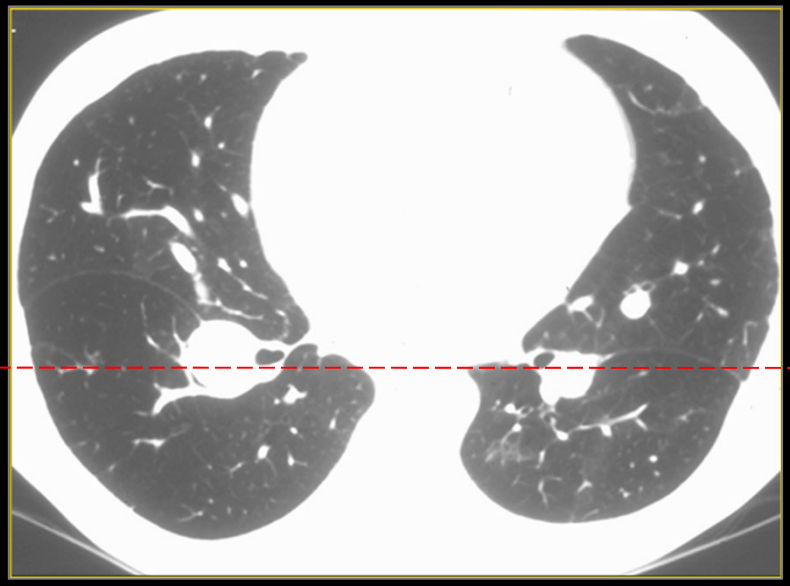
CTEPH



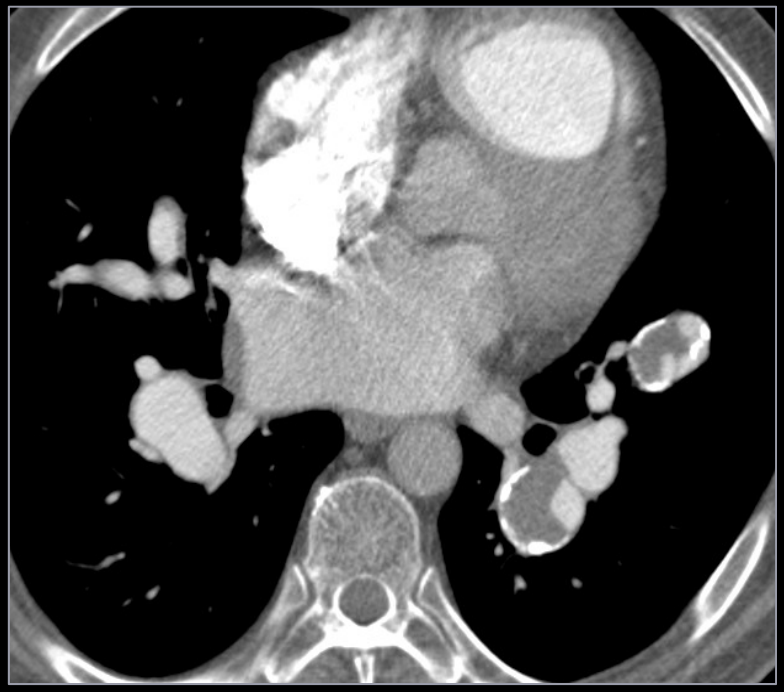
Obtuse angle with the vessel wall

Average density of chronic thrombus 87 HU compared with acute clot (33 HU), due to iron and calcium deposition

CTEPH

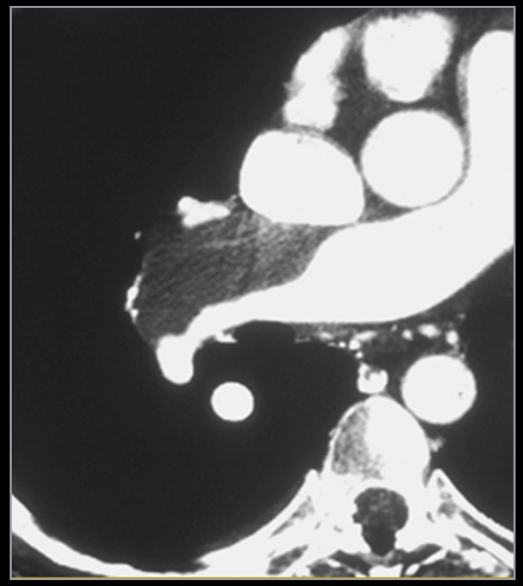


MARKED VARIATION IN SIZE OF THE SEGMENTAL VESSELS
(ASYMMETRIC VESSELS OF THE SAME ORDER)



CALCIFICATION OF THE PULMONARY ARTERIES
is rare in chronic post-embolic hypertension

Chronic thromboembolic pulmonary hypertension (CTEPH)

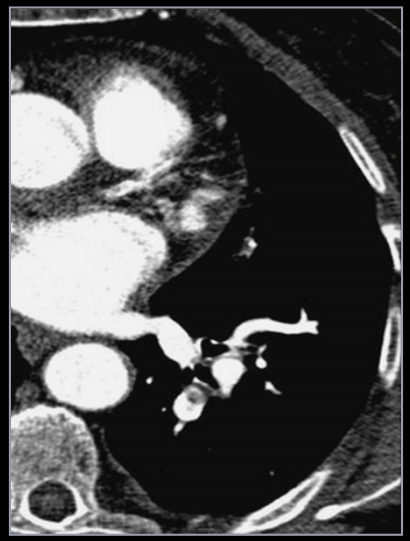


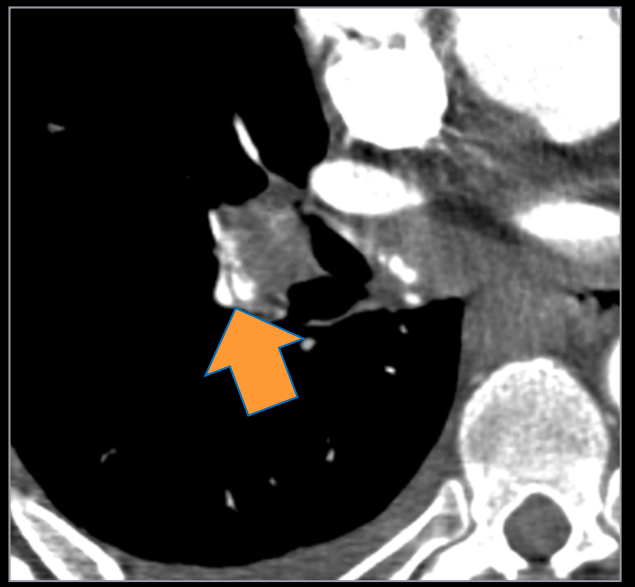
After an episode of acute embolism, lung perfusion scan shows persistent abnormalities in up to 35%, generally with a low degree of vascular obstruction (<15%) but CT is more accurate than scintigraphy.

1 y after an episode of acute pulmonary embolism, >50% of patients have evidence of residual defects on CT study. 2-4% develop PH.

The CT score systems proposed to evaluate the severity of acute PE (Qanadli score, Mastora score etc) cannot evaluate the hemodynamic severity accurately in CTEPH

Chronic thromboembolic pulmonary hypertension (CTEPH)





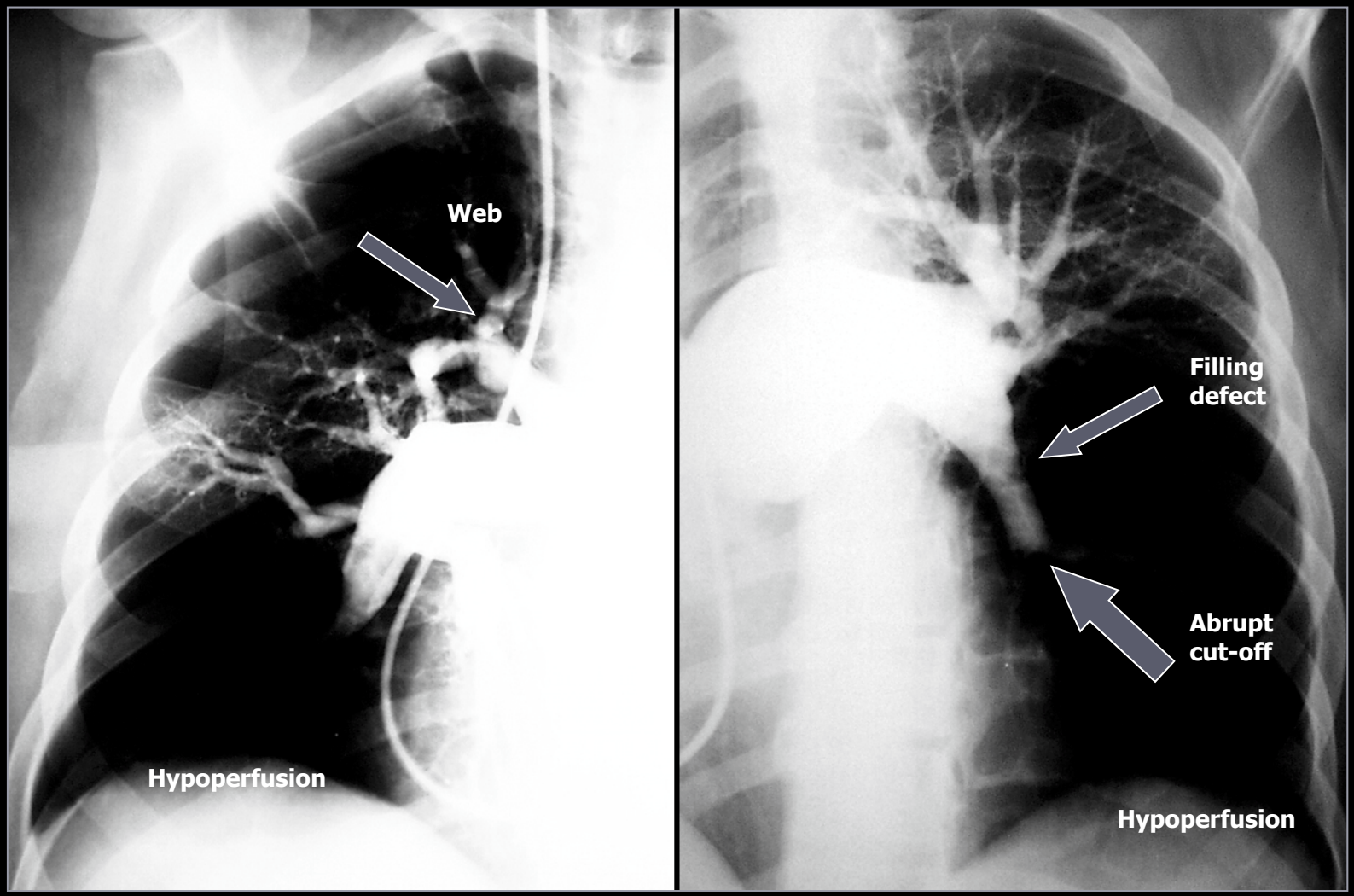
The pulmonary arterial findings for CTEPH are:

organized thrombus, eccentric or circumferential that rarely calcifies; vessel narrowing or obstruction, webs or bands. Also beading of vessels, thready arteries, pouching and intimal irregularities can be observed.

Accuracy of multidetector angio CT versus angiography at the main and lobar arteries: 98%, 94% at the segmental level.

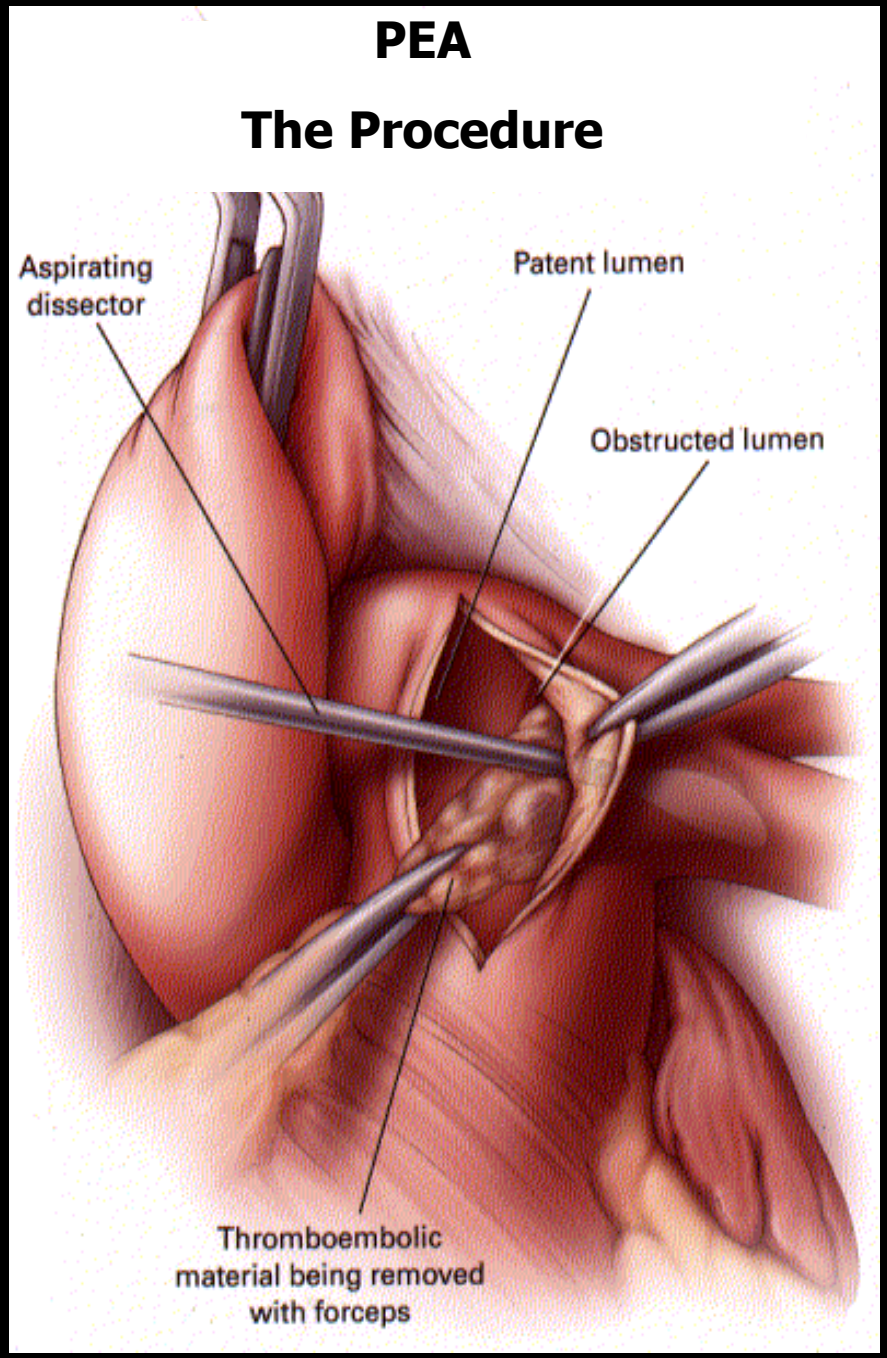
Density of chronic emboli or thrombi (average): 87 HU vs 33 Hu of acute emboli

Pulmonary Angiography

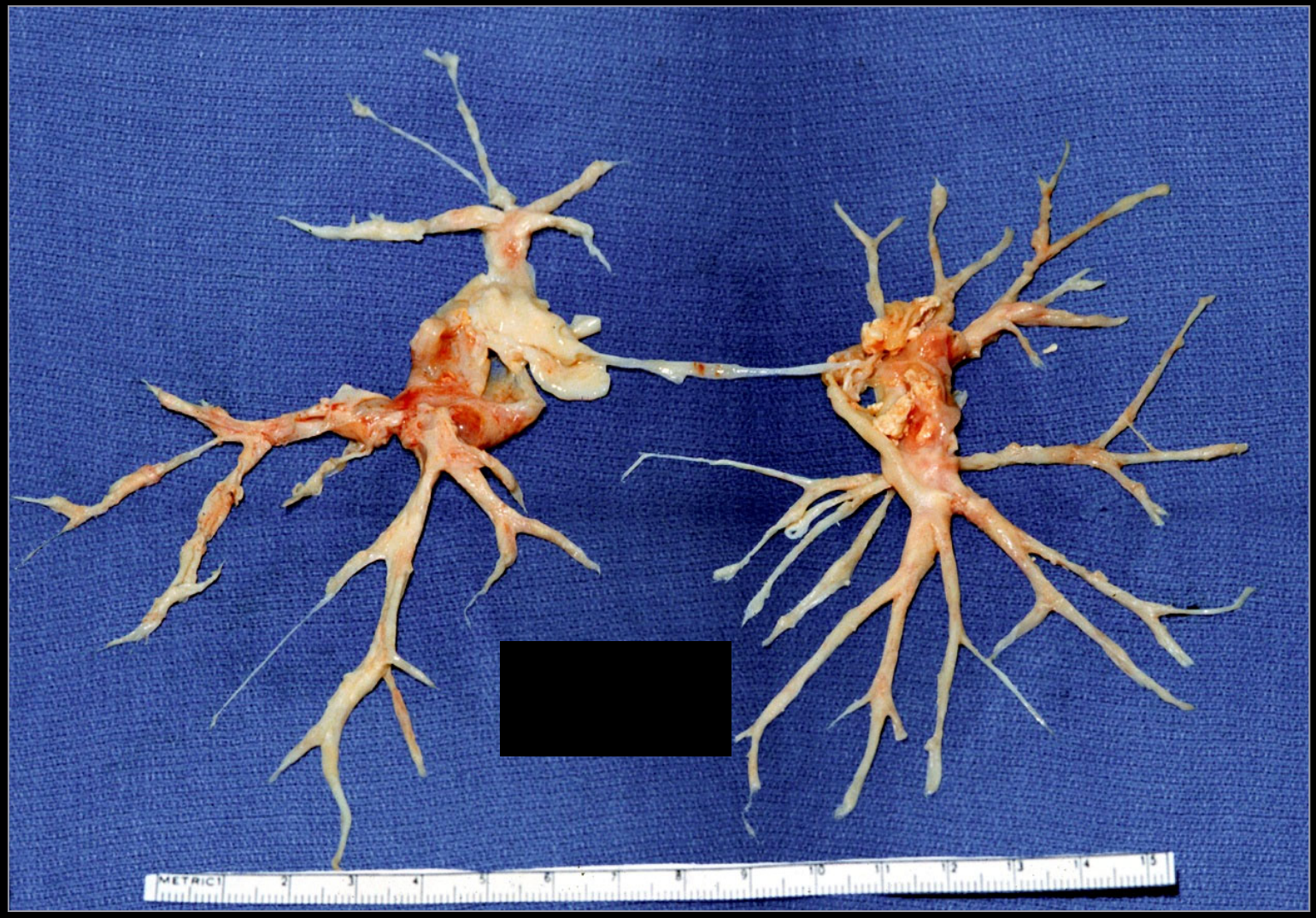


PEA

The Procedure



Specimen



Mortality from PEA: 10%

- In case of CTEPH, if the extent of anatomic disease correlates with the degree of increased PVR, the patient is usually deemed operable but if the disease is mild in the context of a disproportionately elevated PVR, small vessel arteriopathy is likely

- In operable patients with CTEPH, poor subpleural perfusion predicts worse outcome after intervention. In non operable cases, balloon pulmonary angioplasty can be considered. However, also in these cases, a poor subpleural perfusion suggests the presence of small vessels disease and less developed bronchial arteries, predicting failure of treatment

- Post PEA complications: reperfusion pulmonary edema, PA steal syndrome (new areas of V/Q mismatching due to redistribution of flow from normal areas to the newly opened segments), residual thrombi or small vessel arteriopathy, new thromboembolic events

CT SIGNS

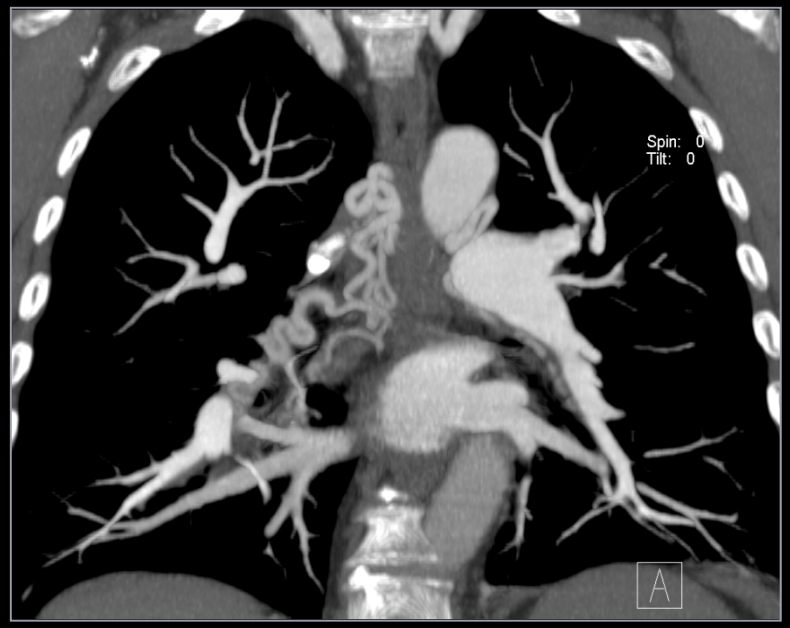
1) VASCULAR SIGNS
   - The main pulmonary artery
   - The other vessels
2) CARDIAC SIGNS
3) PARENCHYMAL SIGNS

CT often demonstrates hypertrophy of the bronchial (diameter > 2 mm, seen along the course of the proximal bronchial tree) and other systemic arteries . This sign is common in CTEPH (70%) and Eisenmenger syndrome; very uncommon in iPAH. Never in portopulmonary hypertension. It can be found also in Takayasu arteritis and Fibrosing mediastinitis.

Non bronchial systemic collaterals in 45%.

In pts with CTEPH, dilatation and tortuosity of the systemic arteries correlate more strongly with the extent of central thrombosis rather than with mPAP. Systemic collateral supply is more extensive in the more severely embolic areas.

This sign predicts a better postsurgical outcome.



MAIN CAUSES OF BRONCHIAL ARTERIES ENLARGEMENT

CTEPH
Bronchiectasis (any cause)
Fibrosing mediastinitis
Takayasu disease
Eisenmenger syndrome
Dieulafoy disease

Frequent hemoptysis
Thin section coronal MIP

CT SIGNS

1) VASCULAR SIGNS
   - The main pulmonary artery
   - The other vessels
2) CARDIAC SIGNS (visible on non gated CT)
3) PARENCHYMAL SIGNS

RV enlargement and <u>hypertrophy</u> <u>(chronic hypertension)</u>

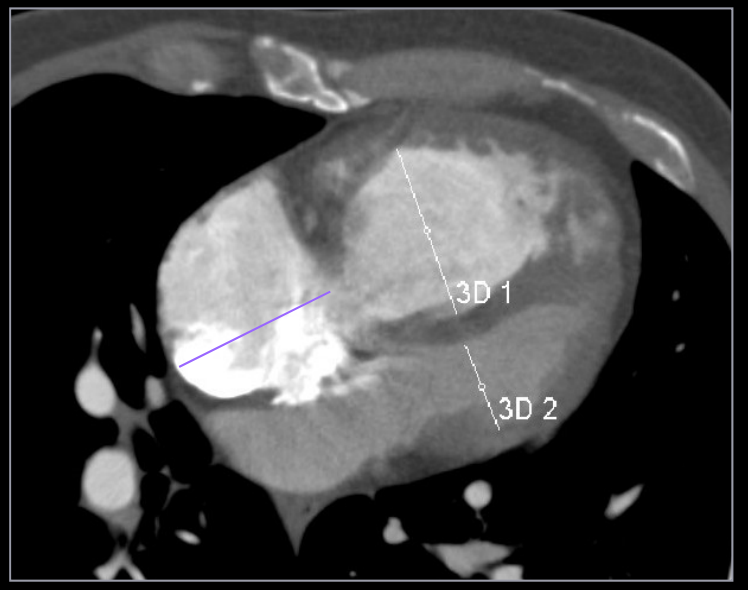
RV myocardial thickness > 6 mm (norm 4)

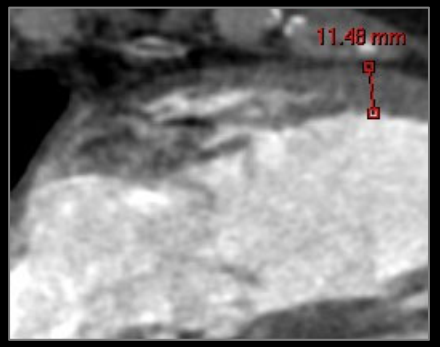
RV / LV ratio > 1,2:1 & bowing of the interventricular septum toward the left side (possible cause of LV diastolic dysfunction).

Dilatation of the right atrium, IVC, sovraepatic veins and coronary sinus.

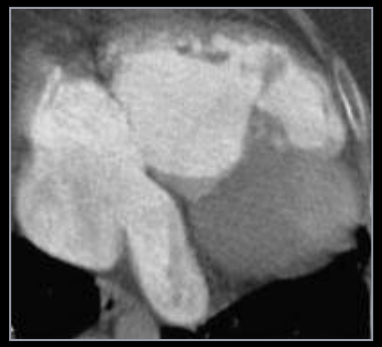
Pleural effusion. In pts with CVD and hypertension, it can be found in >30% and it is often associated with RV failure.

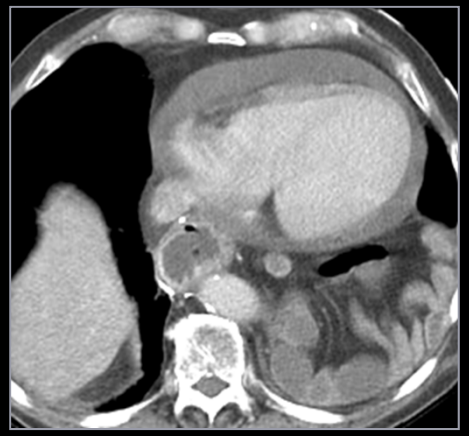
Small pericardial effusion can be found in case of severe hypertension (20% of idiopathic and familial PAH), due to impaired venous & lymphatic drainage. This sign is non specific (ex: connective tissue disorders).

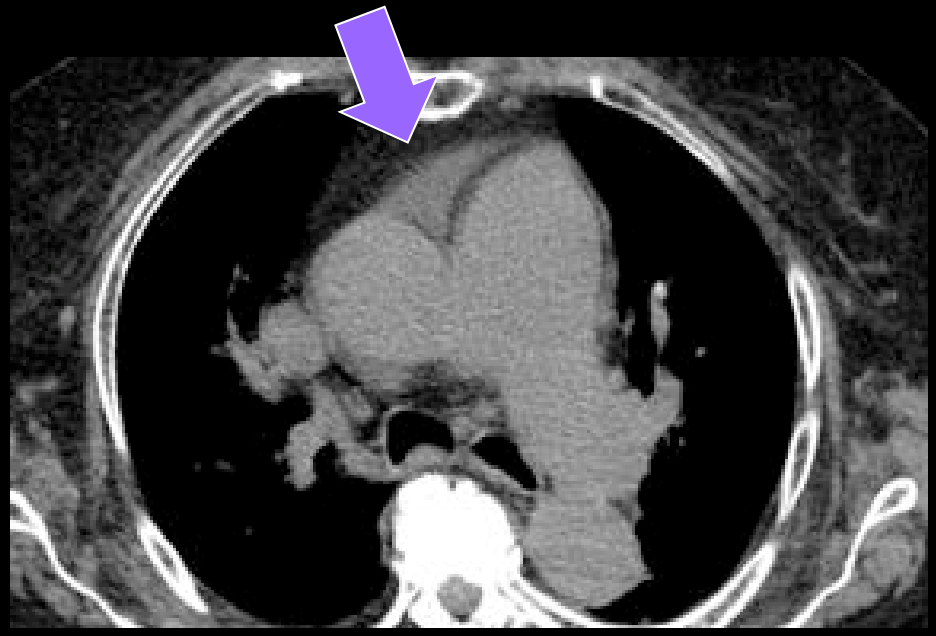




RV diam > 45 mm
RA diam > 35 mm







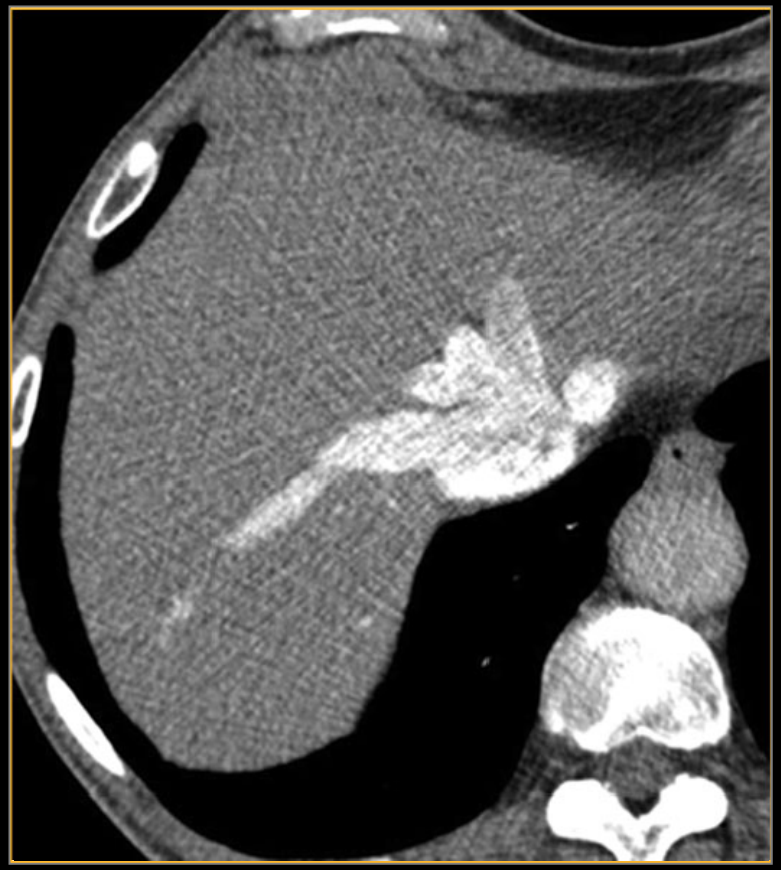
A significant amount of fluid (> 1,5 cm in depth) within the anterior pericardial recess occurs frequently in pts with hypertension (50% of patients with severe IPAH, due to increase in right atrial pressure).

Bikini bottom sign

This sign implies a worse prognosis

Tricuspid valve reflux-regurgitation (sens & spec 90% with an injection rate of up to 4 mL/sec, but sometimes visible in normal subjects when the rate of injection is high).

The degree of reflux correlates with mPAP and represents a strong predictor of outcome.

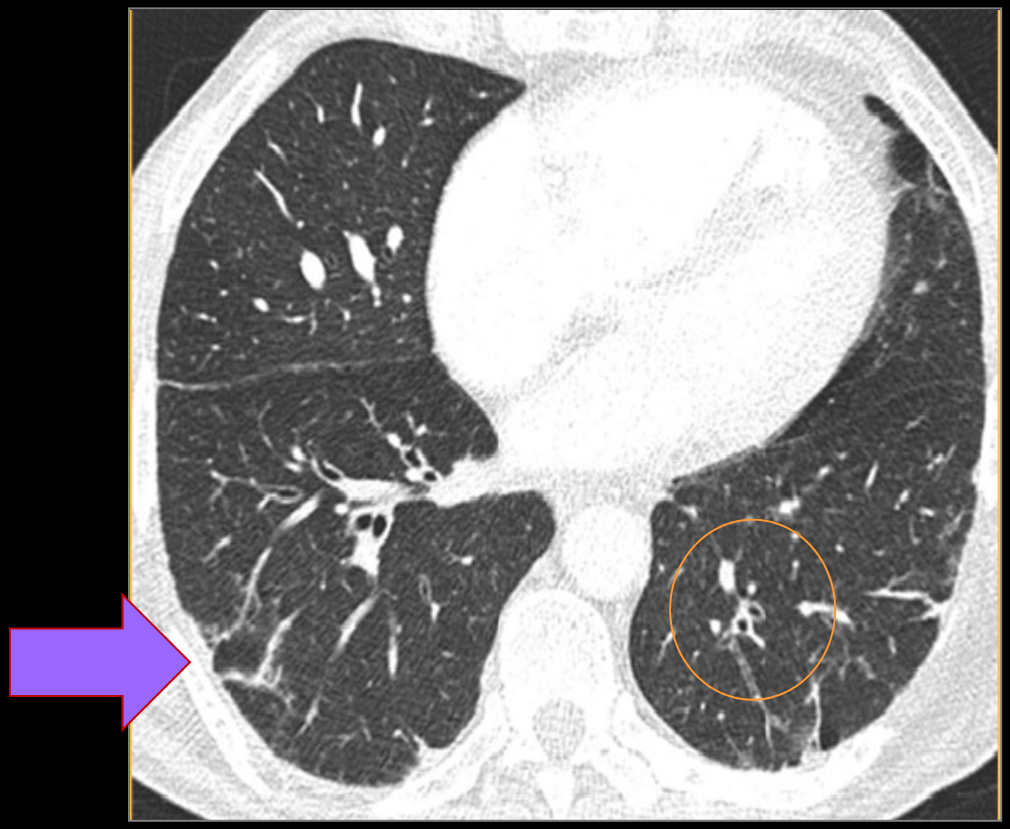


GRADE 3

GRADING:
0= non reflux
1= minimal
2= reflux into IVC but not hepatic veins
3= IVC and proximal hepatic veins
4= IVC, distal hepatic veins; dilatation

CT SIGNS

1) VASCULAR SIGNS
   - The main pulmonary artery
   - The other vessels
2) CARDIAC SIGNS
3) PARENCHYMAL SIGNS



Scars from previous infarctions are common in pts with CTEPH (80%) and generally indicate a poorer outcome after surgery.

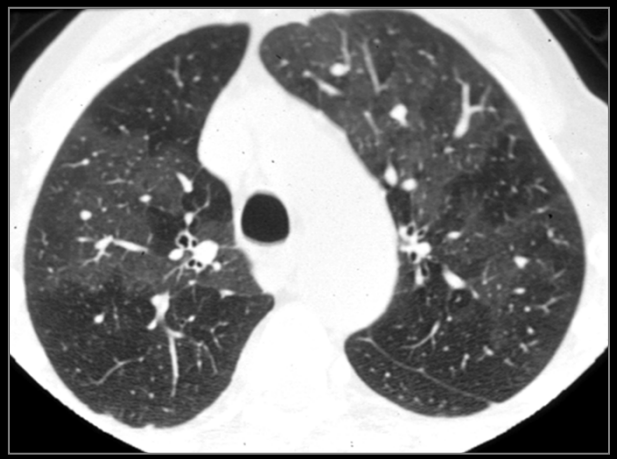
Cylindrical «bronchiectasis» adjacent to occluded or stenotic arteries can occur from repeated bouts of post-thrombotic vascular fibrosis (65% of the cases of post-embolic hypertension) or hypoxic bronchodilatation.Not a feature of Eisenmenger Syndrome.

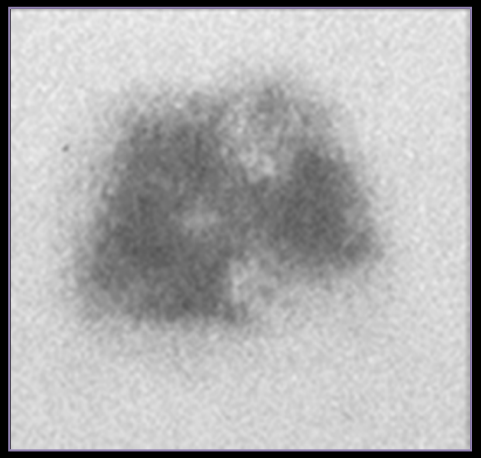
- Mosaic attenuation (areas of under- and over-perfusion, often segmental or sub segmental): 90% of CTEPH versus 70% in cardiac causes of PH and <10% in lung diseases; almost never in pulmonary artery sarcomas

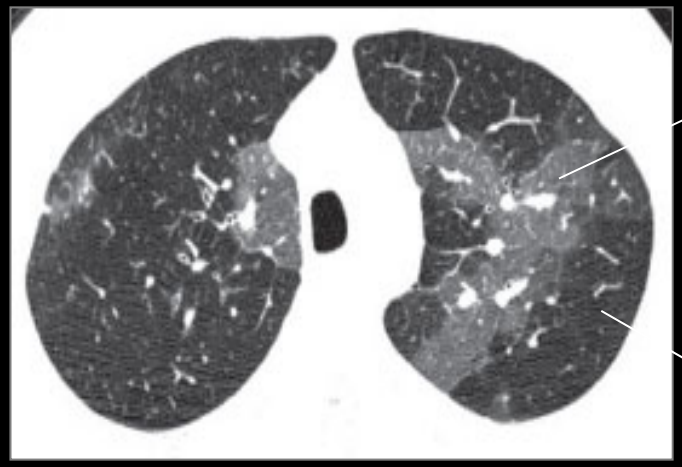
- When using minIP post-processing, sensitivity of CT and scintigraphy are similar. However, small differences can still be found and are related to the different kind of information provided by the two methods. Scintigraphy shows Tc99m MAA trapped in the pre-capillary vessels; angioCT can demonstrate radiologic contrast enhancement in areas perfused only by bronchial collaterals

- Mosaic pattern is significantly less common in patients with idiopathic PAH (perivascular, not segmental)

- Mosaic perfusion can disappear after successful PEA in patients with CTEPH. On the other hand, in CTEPH, diffuse & peripheral perfusion defects are generally a predictor for a poor surgical outcome (severe small vessel obliteration)

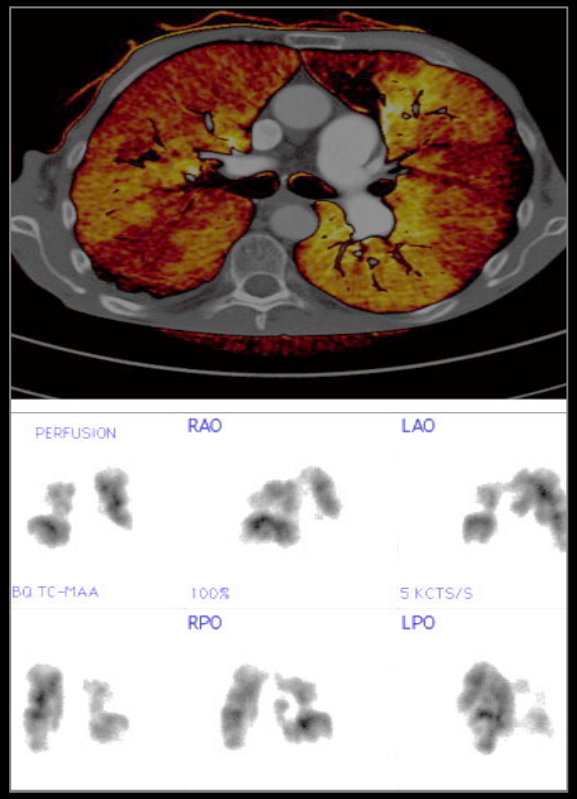






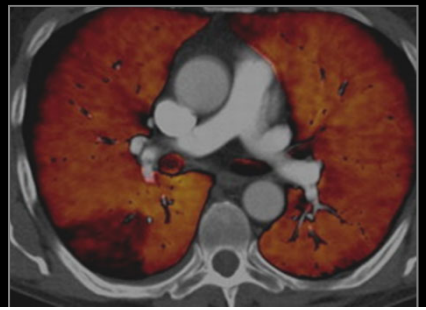
OVER-PERFUSION (not necessarily normal; risk of arteriolar remodelling)

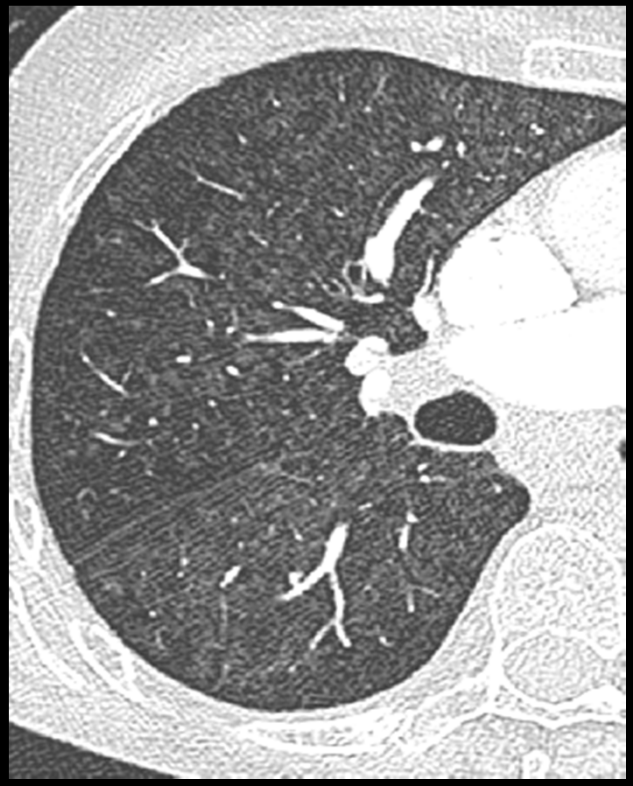
HYPOPERFUSION

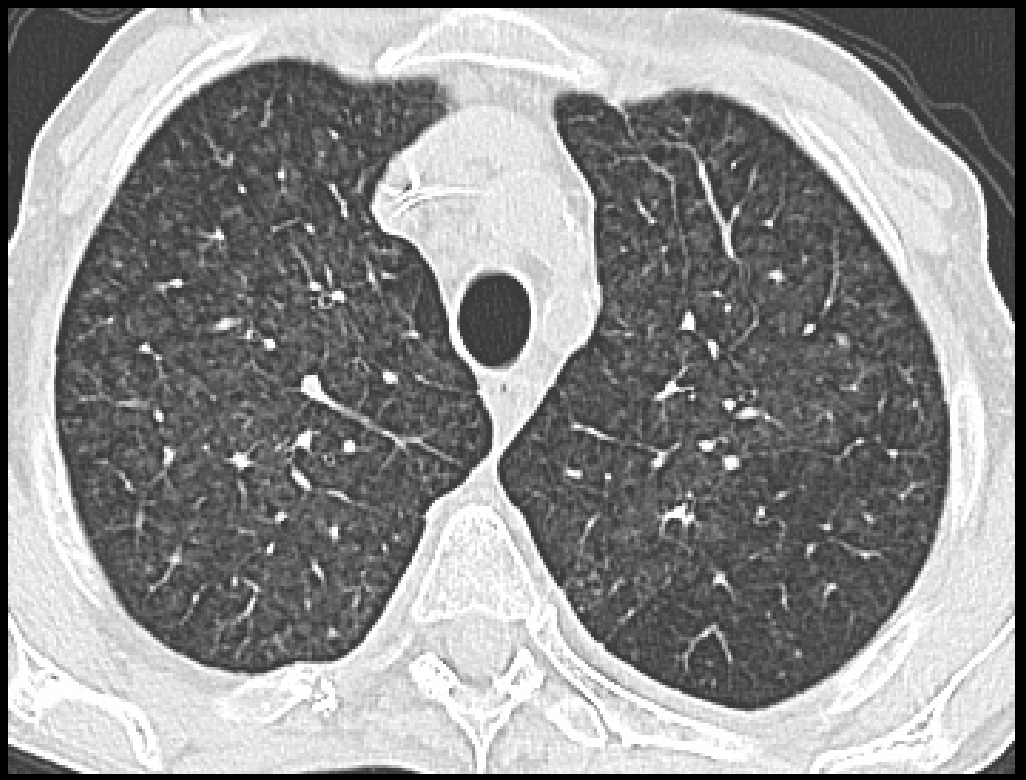


MOSAIC OLIGOEMIA IN A CASE OF CHRONIC EMBOLISM + PRUNING OF THE PERIPHERAL ARTERIES

The mosaic pattern of CTEPH is different from the localized wedge-shaped appearance of acute PE. The difference is better demonstrated by DECT







Areas of GGO have been described in 40% of the patients with severe pulmonary hypertension. Central distribution may be more common in SSc patients, centrilobular pattern is more common in severe iPAH especially in those receiving long term vasodilator therapies, PVOD-PCH and Eisenmenger and predicts poor outcome.

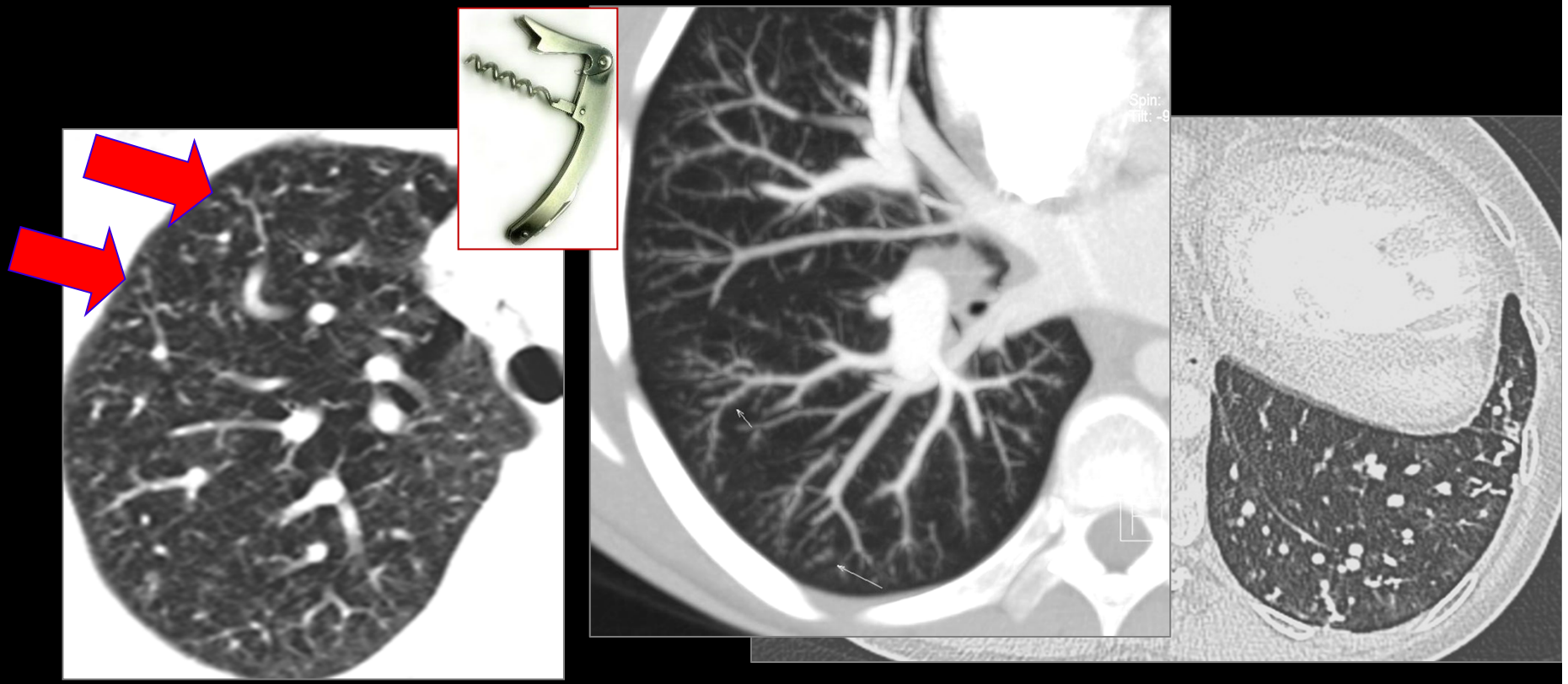
Possible explanations: cholesterol granulomas, pulmonary bleeding, foci of plexogenic arterial lesions, proliferation of capillaries (PCH), areas of the lung perfused by systemic to pulmonary collateral vessels.

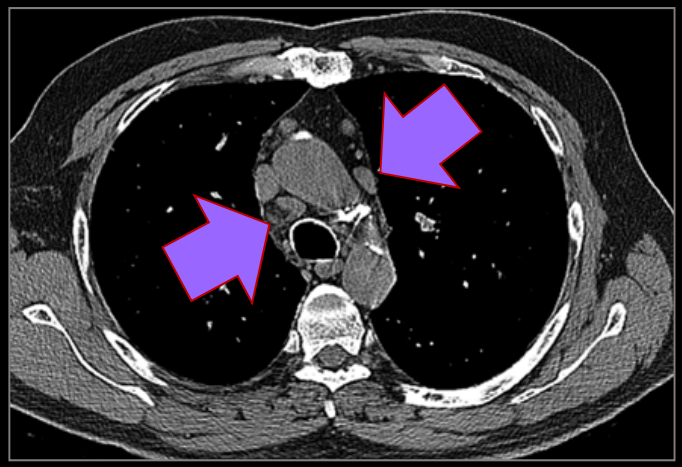
Peripheral neovascularity with a corkscrew morphology (**Sheehan vessels**, probably representing another sign of plexogenic arteriopathy or collateral circulation):

tiny, serpiginous arteries emerging from centrilobular regions, without a connection to pulmonary veins.

Very common in Eisenmenger syndrome (>70%); more rarely found in iPAH or in pulmonary hypertension of any cause.

Their presence & number are a marker of severe hypertension.

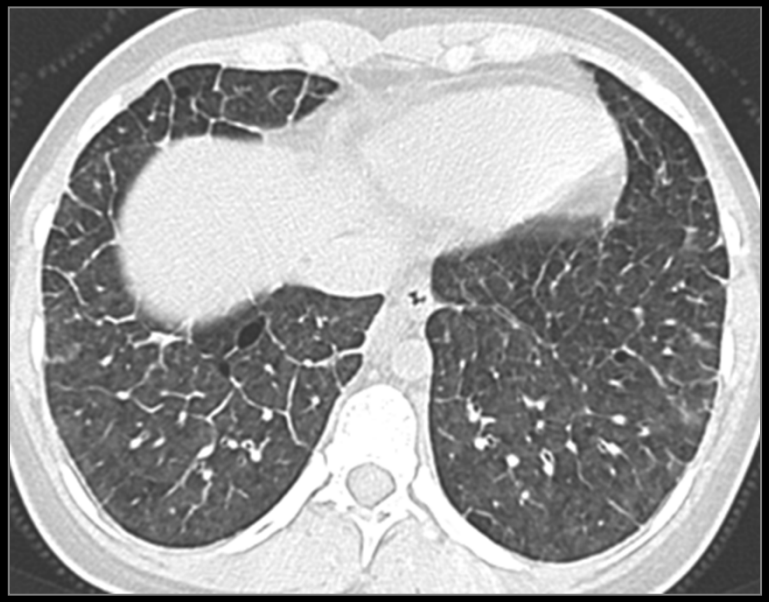


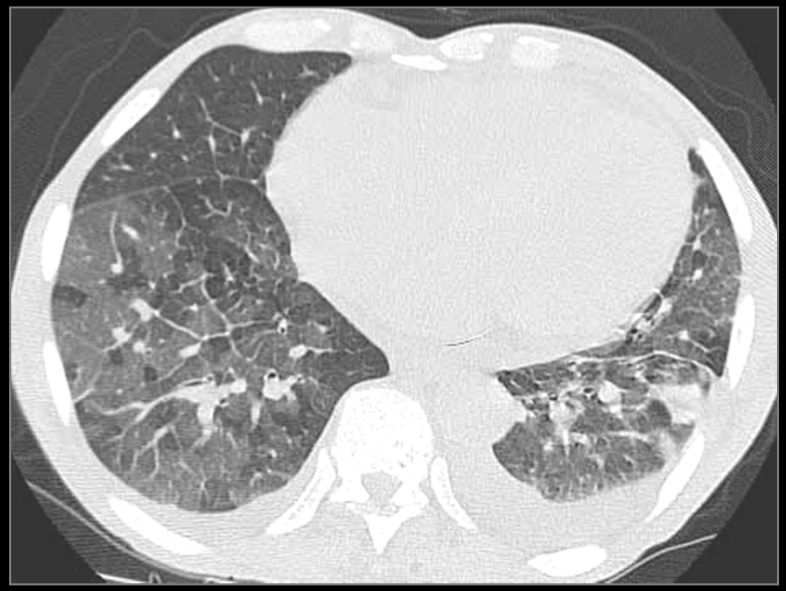


Mediastinal adenopathies can be seen in approximately 20% of patients with idiopathic PAH and are often associated with pleural (not pericardial) effusion.

40% of post-embolic hypertension.

50% of congestive heart failure.





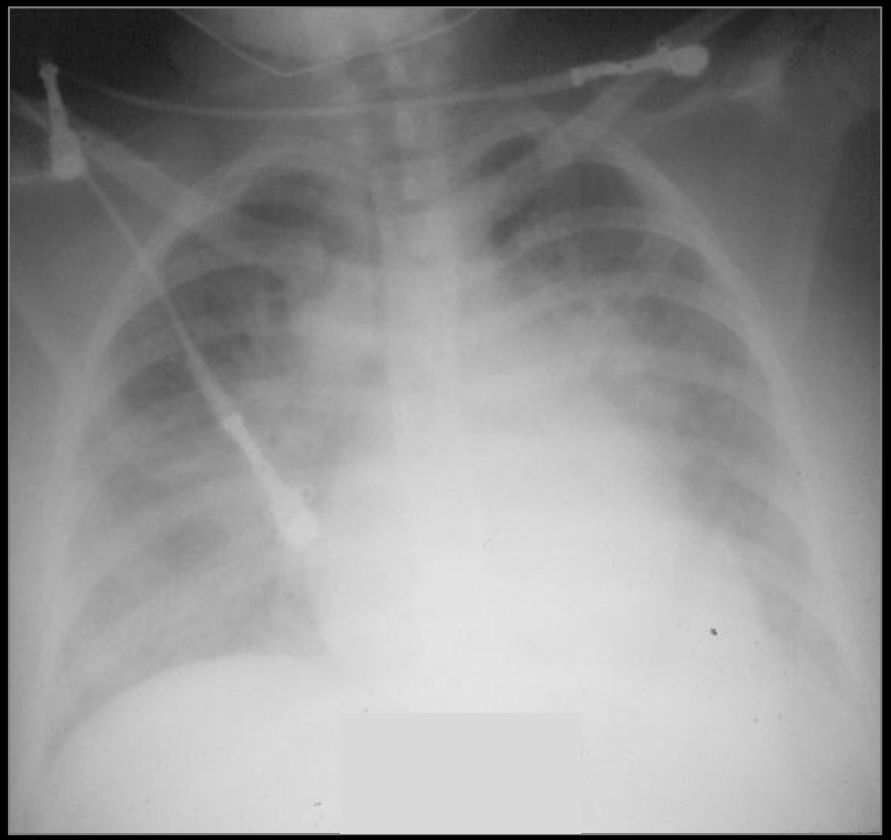
PVOD

Radiological findings, along with a normal PWP

Dilatation of Pulmonary arteries with:
GGO (>80%, often centrilobular, due to alveolar edema and hemorrhage),
interstitial edema with interlobular smooth septal thickening (50%),
enlarged lymph nodes (20%); normal or small pulmonary veins and left atrium,
pleural effusion (not universally present in PVOD; 10% of the pts with PAH; poor outcome),
mosaic perfusion, reflecting regional decreased vascularity with patchy distribution.

75% of these patients have at least two findings
PVOD CT diagnosis: 70% sens, 90% spec

PVOD AND PCH can be associated in the same patient



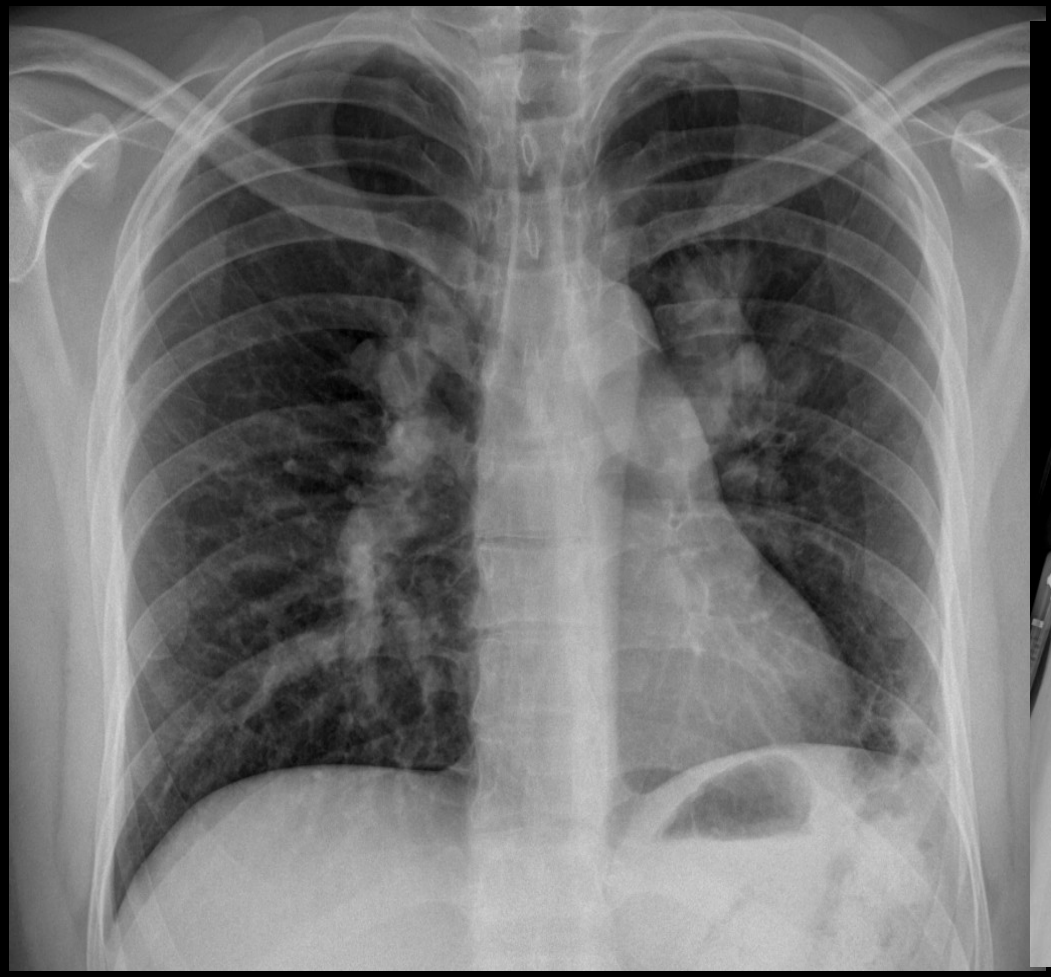
In patients with PVOD and PCH, standard antihypertensive therapy may cause flash edema and death

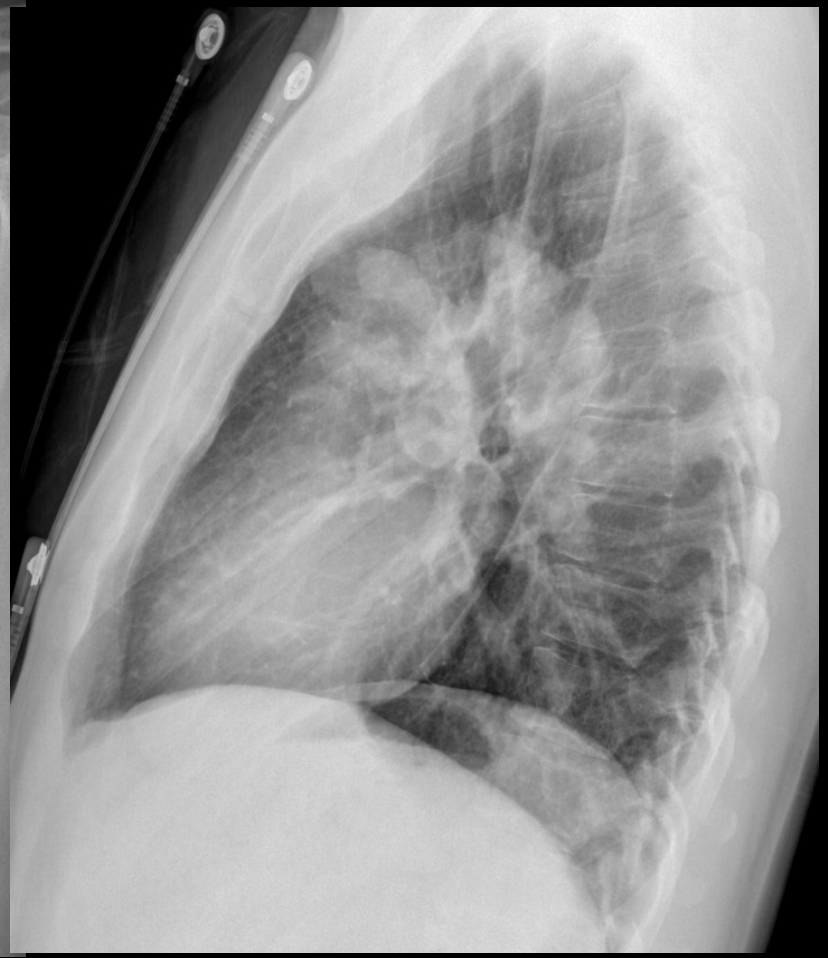
POOR PROGNOSIS

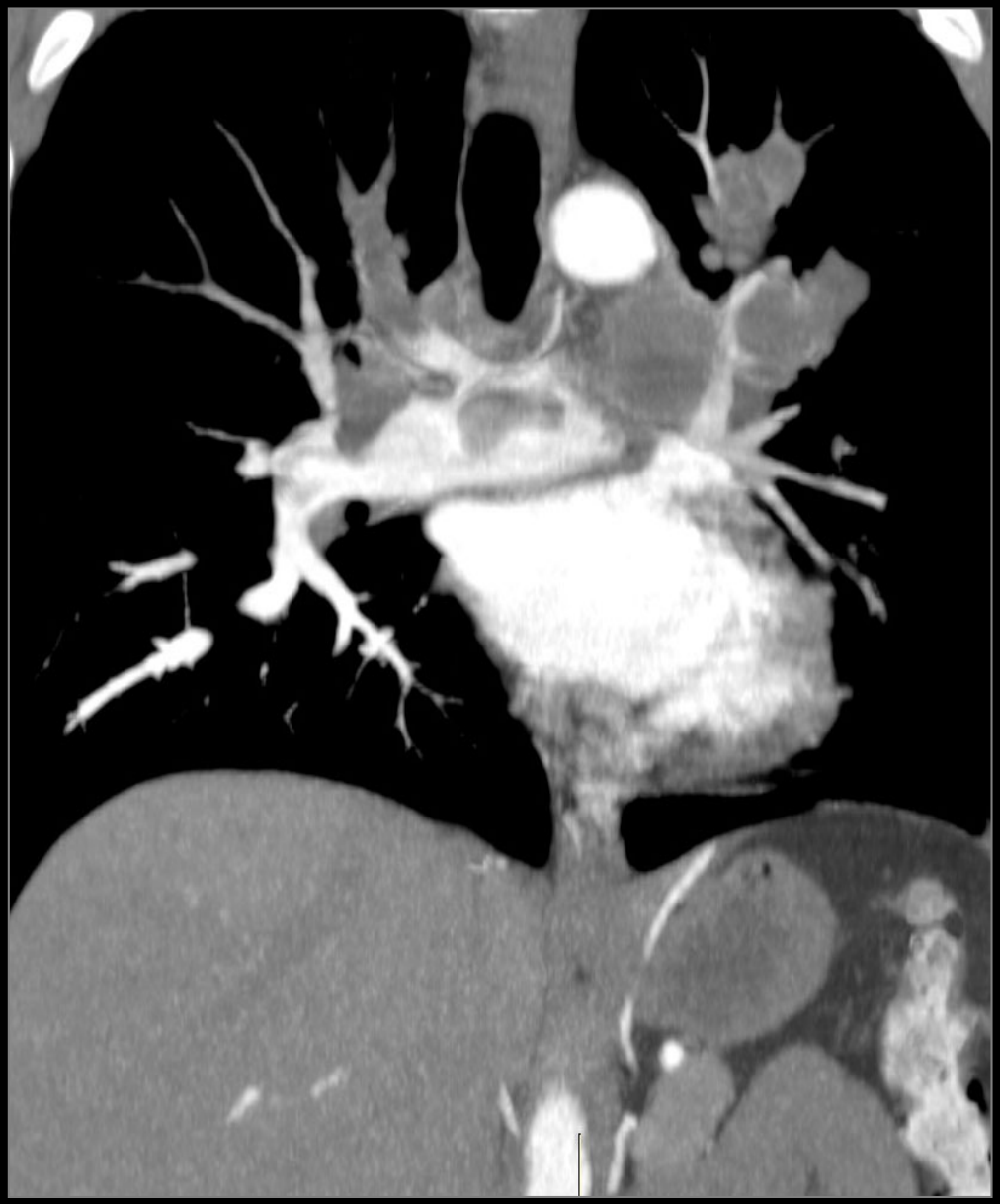
IMAGING FINDINGS INDICATING POOR PROGNOSIS

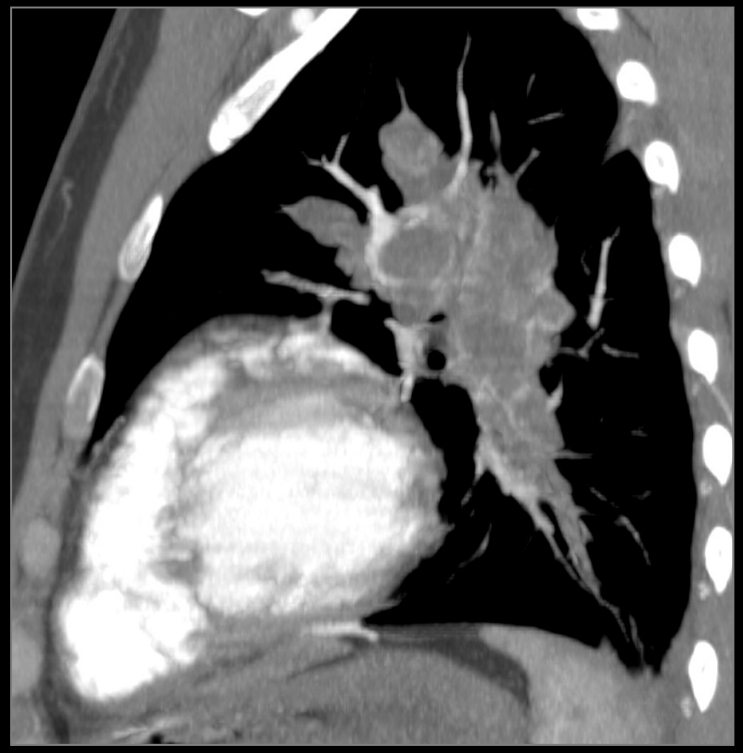
- Pericardial effusion
- Great PA dilatation
- Large right atrial & ventricle size
- Septal shift during early diastole
- Severe tricuspid valve regurgitation
- Mediastinal adenopathies; septal lines
- Myocardial delayed enhancement (MRI; RV ischaemia)

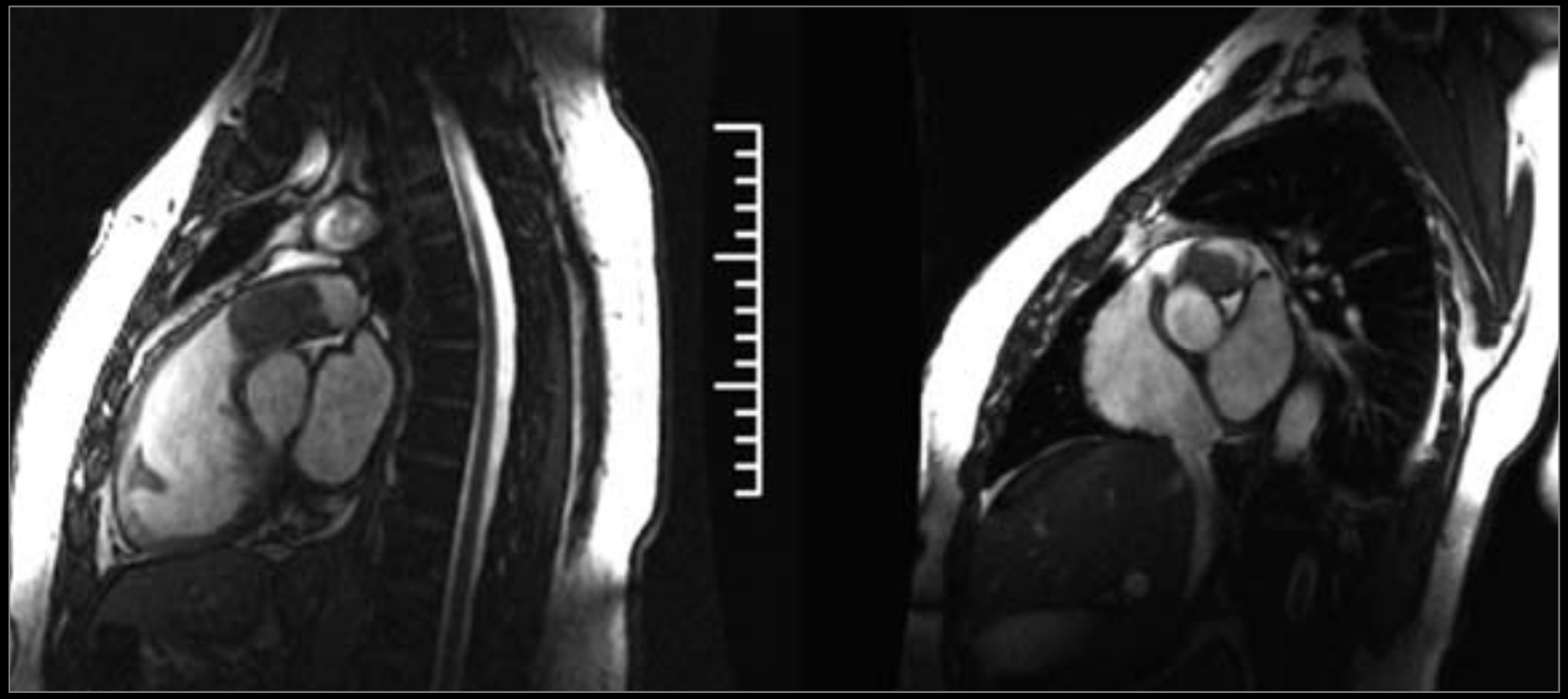
AND, OF COURSE

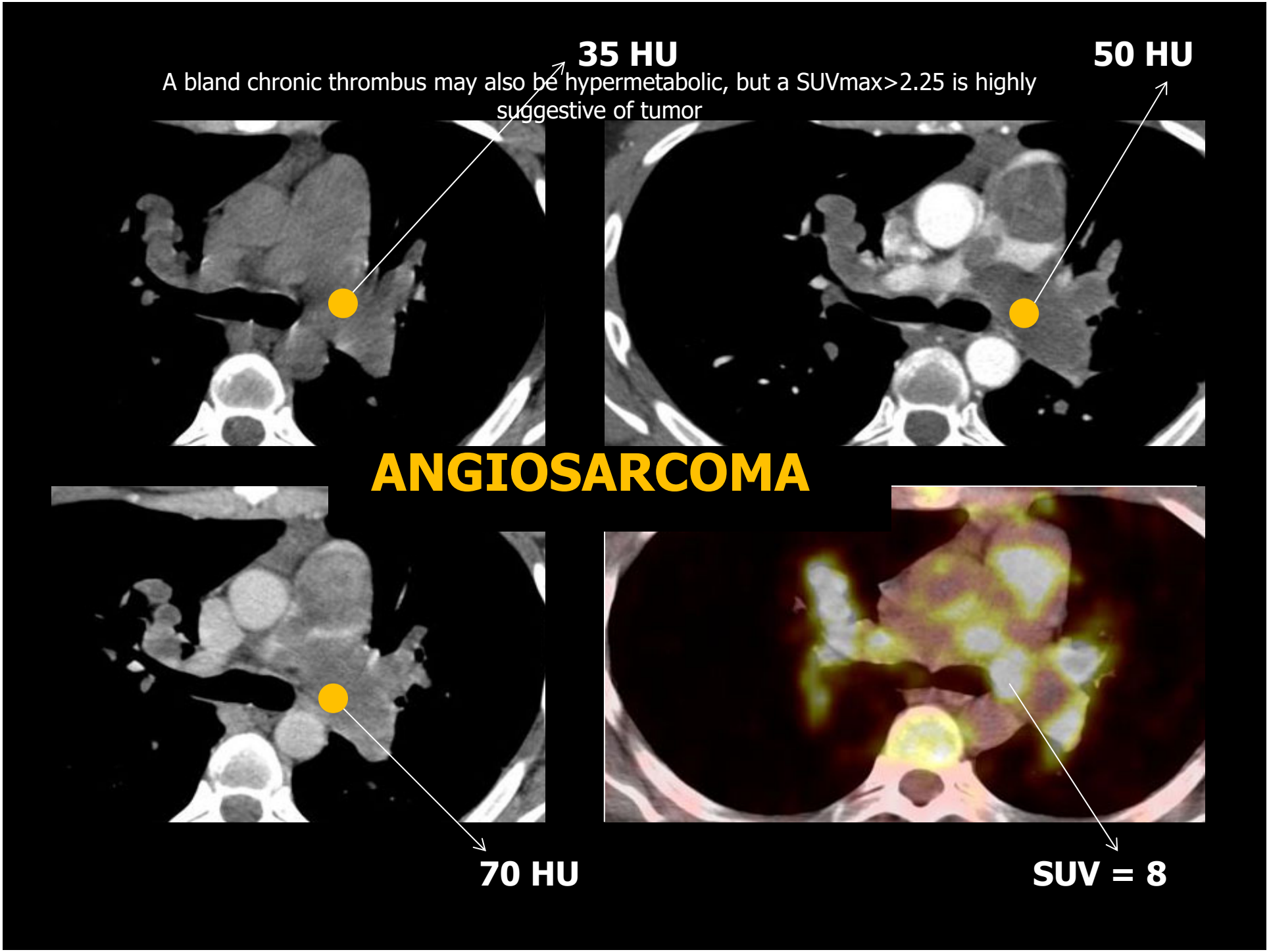






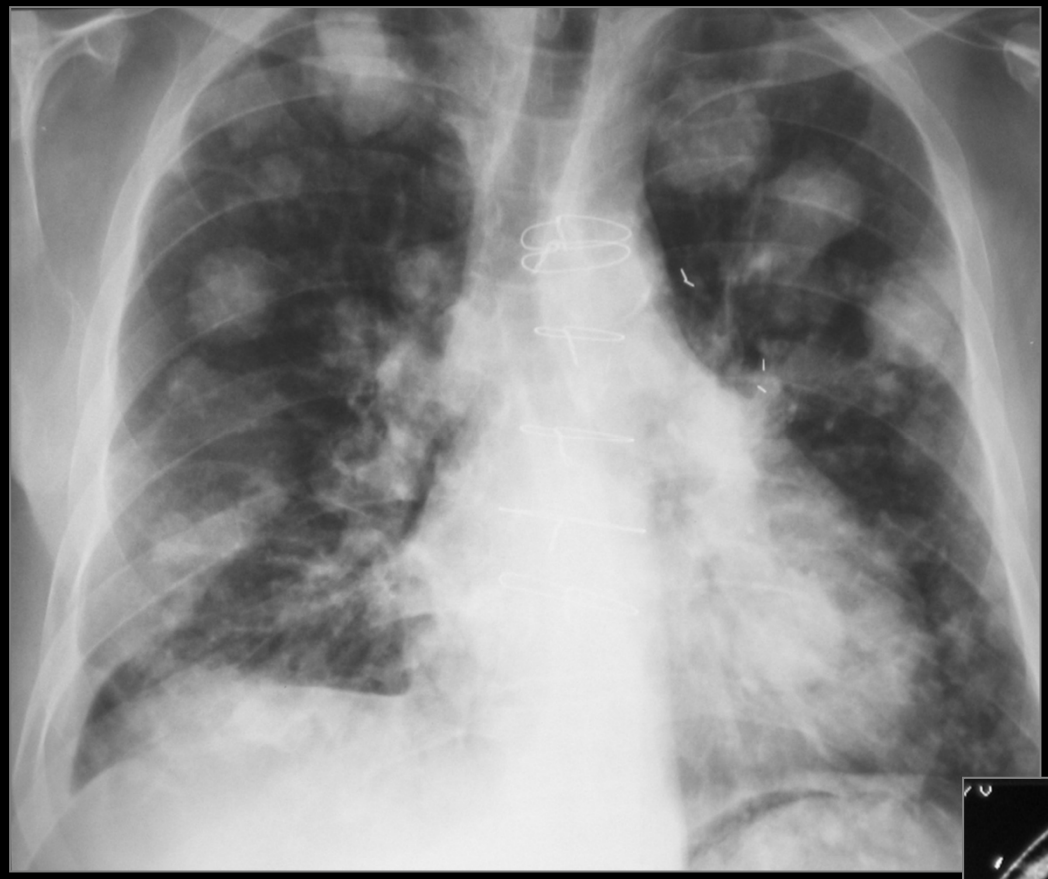


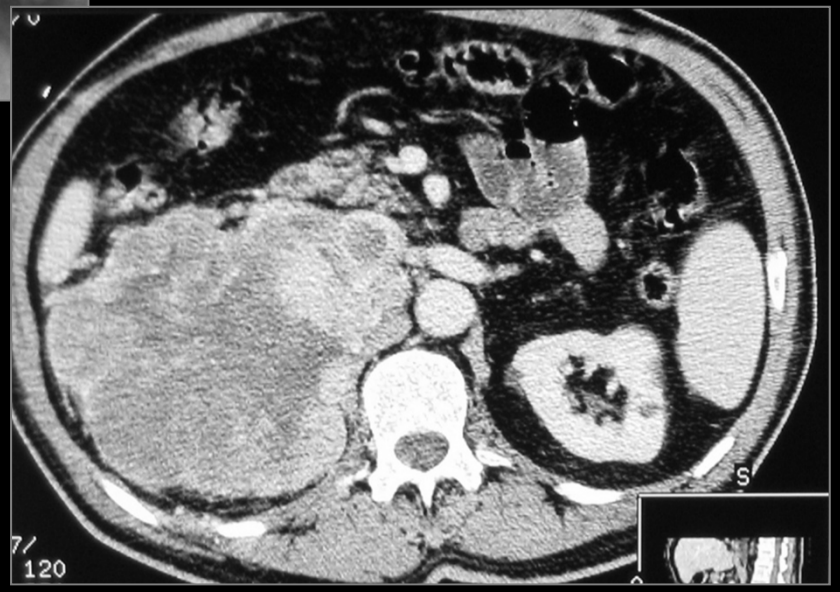


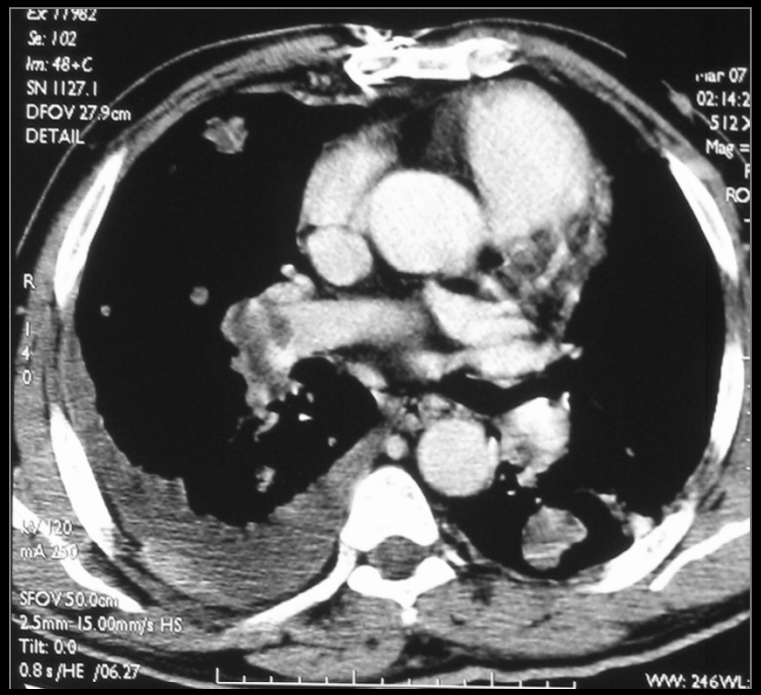

35 HU
50 HU
A bland chronic thrombus may also be hypermetabolic, but a SUVmax>2.25 is highly suggestive of tumor
ANGIOSARCOMA
70 HU
SUV = 8

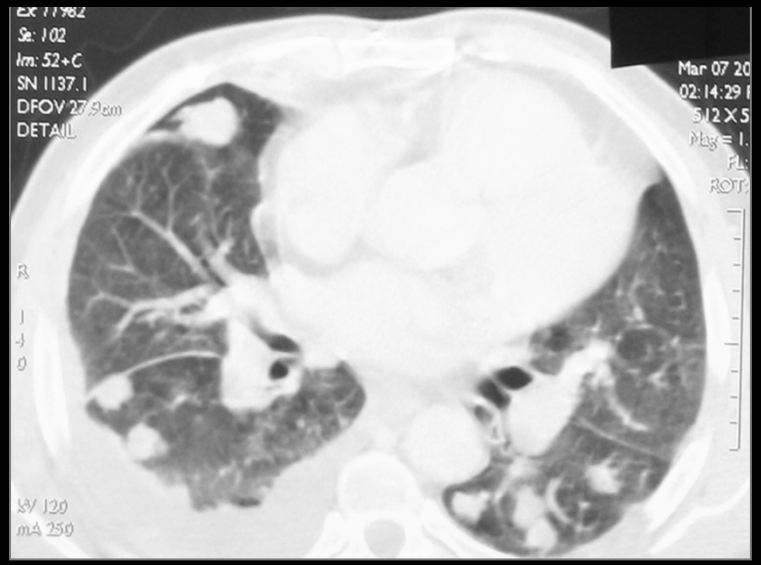
NEOPLASTIC EMBOLISM IN THE LUNG

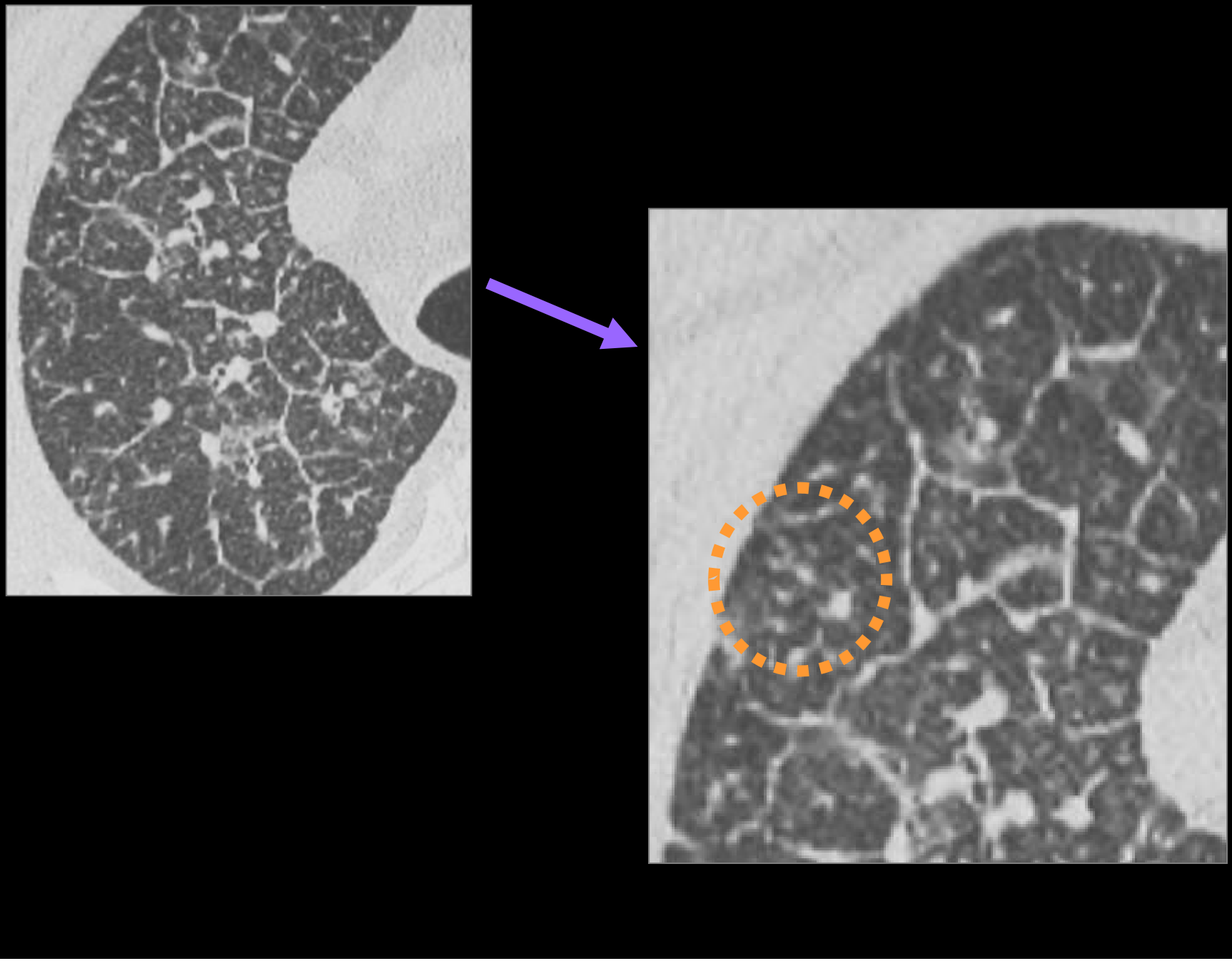
- CENTRAL PULMONARY ARTERIES
- PERIPHERAL ARTERIES (beaded vessel sign)
- INTRALOBULAR ARTERIES (thrombotic microangiopathy +/- typical mets or Carcinomatous Lymphangitis)

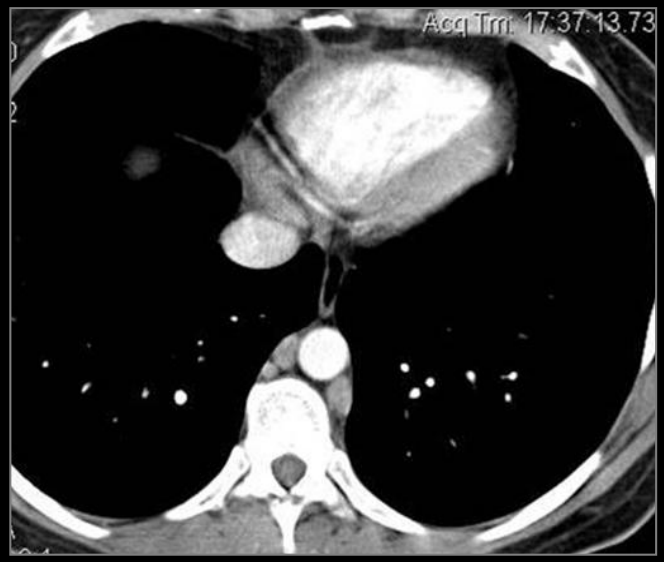


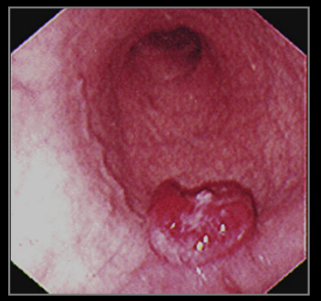

S
7/
120

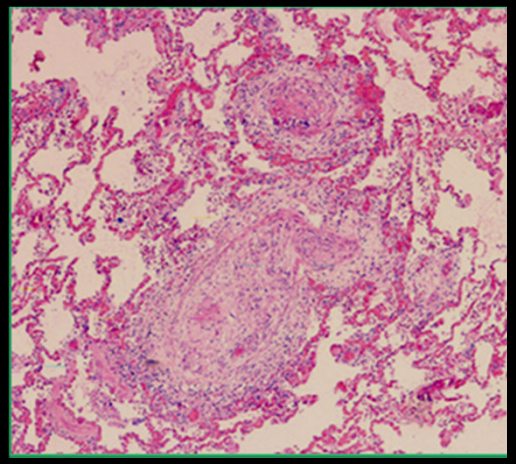

Se: 102
Im: 48+C
SN 1127.1
DFOV 27.9cm
DETAIL
kV 120
mA 250
SFOV 50.0cm
2.5mm 15.00mm/s HS
Tilt: 0.0
0.8 s/HE /06.27

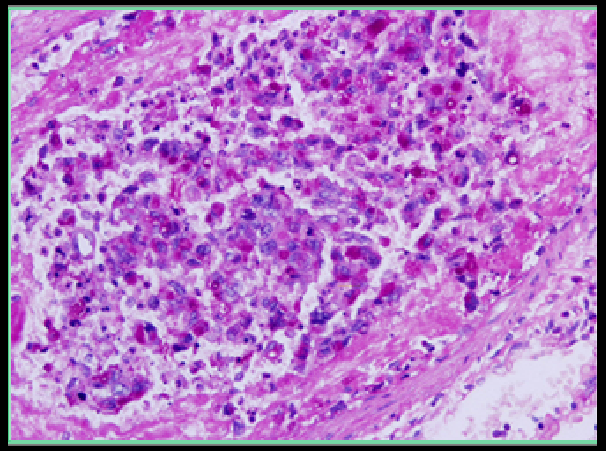

Se: 102
Im: 52+C
SN 1137.1
DFOV 27.9cm
DETAIL
kV 120
mA 250

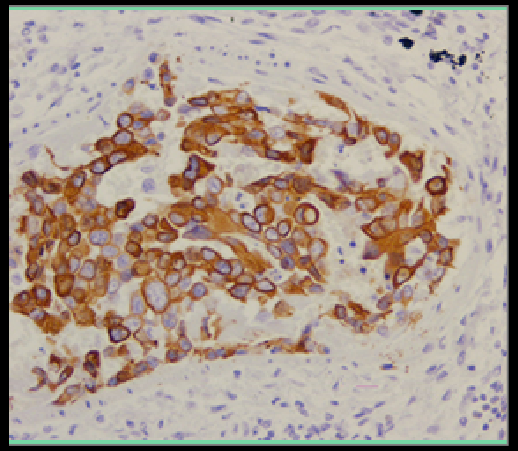












THROMBOTIC MICROANGIOPATHY (FROM METASTATIC **GASTRIC CANCER**)
(also from breast, liver, kidney, lung, prostate tumors and choriocarcinoma), sometimes associated with lymphangitic carcinomatosis and pleural effusion. 2% of the patients who died of a cancer.
DD: PVOD. Evolution more rapid than in Lymphangitic carcinomatosis.

CLINICAL TAKEAWAY

- Although V/Q scanning continues to be used as the initial screening test, CT has emerged as the definitive imaging test of choice in depicting the structural and vascular abnormalities in CTEPH

- A combination of CT signs improves the accuracy in the diagnosis of pulmonary hypertension, providing diagnostic & prognostic informations

CLINICAL TAKEAWAY

- Informations from CT can be used with other tests to stratify patients, establish the etiology and in monitoring progression & response to treatment

- Definitive diagnosis of a new case of pulmonary hypertension requires correlation with clinical, angiographic, hemodynamic findings and sometimes with histopathology

- High suspicion index is required in pts with unexplained dyspnea

- The Dx of Idiopathic PH is a DX of exclusion

- Substantial advances have occurred over the past quarter century in the approach to pulmonary hypertension

- Imaging techniques contribution represents an important aspect of the integrated evaluation of these patients, even if unanswered questions still remain

- Further progress can be expected, related to the development of DECT and MR angiography. MR and prospective gated CT studies have the potential to provide both anatomic and functional data

- Excellent correlation between the perfusion maps obtained using DECT and those obtained using V/Q scans and SPECT-CT exams

- MR plays a complementary role. Stiffness of the proximal PA is a strong indipendent predictor of mortality

CT LIMITATIONS

- CT does not provide pulmonary pressure and resistances (severity of PH cannot be precisely estimated by CT)

- A normal appearance of the pulmonary arteries does not eliminate the possibility of a mild pulmonary hypertension

- Ascending aorta diameters may change with body size and/or age

- PA dilatation may also occur without hypertension in patients with chronic diffuse pulmonary fibrosis

ACR APPROPRIATENESS CRITERIA (2013)

Imaging procedure	Rating
US transthoracic	9
Right heart catheterization	9
Chest X ray	8
CTA with contrast (non coronary)	8
Tc-99m V/Q scan	7
MR heart and P Arteries	6
Arteriography	6
US transesophageal	5

1-3 usually not appropriate; 4-6 may be appropriate; 7-9 usually appropriate

A Systematic Approach

Systematic review of a CTPA includes determination of the quality of study and repeating it if the scan is of poor quality.

First step in the assessment of a CTPA should be to look at the various parameters and assess the presence of PAH. This is then followed by the evaluation of shunts (intra and extra cardiac), left heart morphology (signs of infarction, mitral stenosis, etc.) and right heart morphology.

Pulmonary vasculature and systemic arteries are then evaluated to exclude underlying PTEmbolic disease.
Lung parenchyma is then reviewed to look for ILD-, COPD-, PCH-, and PVOD-related changes.
Lastly, liver and portal vein should be assessed and features of portal hypertension.

NOT SO EASY, BUT DEFINITELY WORTH A TRY.

JUST GIVE IT