Διαγνωστική Προσέγγιση Πνευμονικής Υπέρτασης

Αξονική Τομογραφία

ΑΡΓΕΝΤΟΣ Ι. ΣΤΥΛΙΑΝΟΣ ΔΙΕΥΘΥΝΤΗΣ ΕΣΥ
Β΄ ΕΡΓΑΣΤΗΡΙΟ ΑΚΤΙΝΟΛΟΓΙΑΣ ΠΑΝ/ΜΙΟΥ ΑΘΗΝΩΝ
ΑΤΤΙΚΟ ΠΑΝ/ΚΟ ΝΟΣΟΚΟΜΕΙΟ

5TH

WORLD SYMPOSIUM ON PULMONARY HYPERTENSION

NICE ACROPOLIS, Nice February 27-28 / March 1, 2013

European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

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[a]Representing the European Respiratory Society; [b]Representing the Association for European Paediatric and Congenital Cardiology; [c]Representing the International Society for Heart and Lung Transplantation; [d]Representing the European League Against Rheumatism; and [e]Representing the European Society of Radiology.

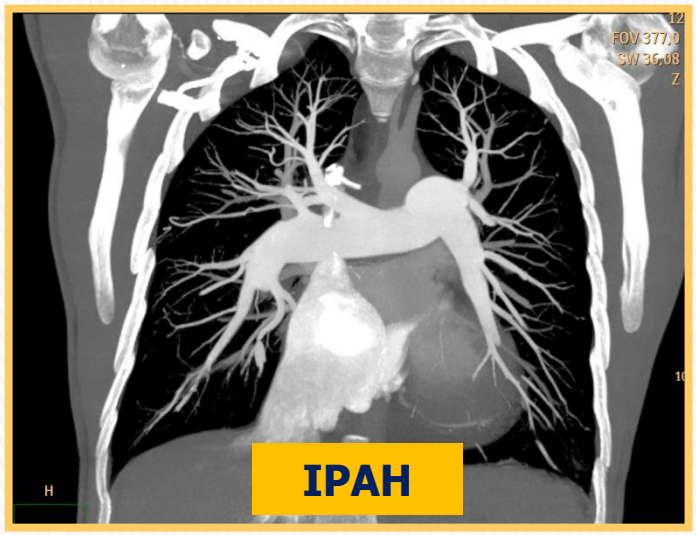
ESC entities having participated in the development of this document:

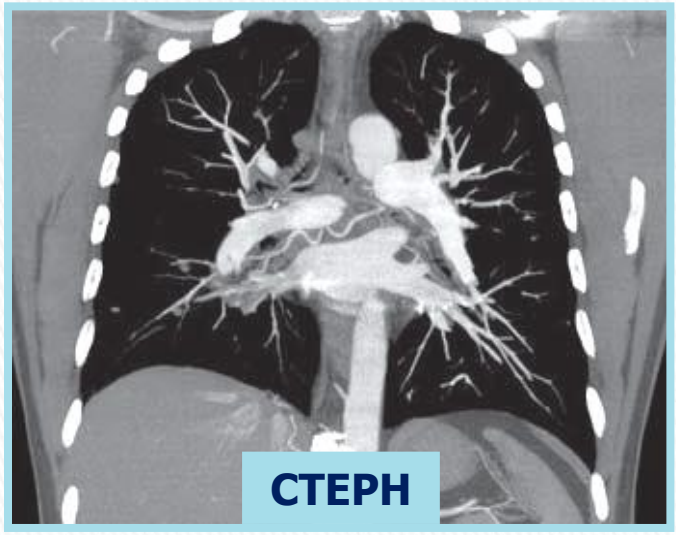
ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

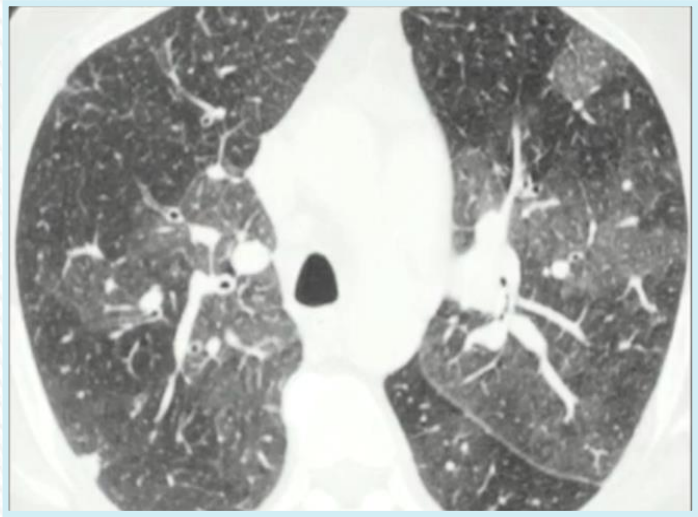
ESC Councils: Council for Cardiology Practice (CCP), Council on Cardiovascular Nursing and Allied Professions (CCNAP), Council on Cardiovascular Primary Care (CCPC).

Pulmonary Hypertension
CT Protocol

- ✓ High Resolution CT : Inspiratory Expiratory
- ✓ Computed Tomography Pulmonary Angiography (CTPA)







- ✓ Γρήγορες σαρώσεις (4-12 sec) μετά IV έγχυση ιωδιούχου σκιαγραφικού
- ✓ Τροποποίηση του πρωτοκόλλου έγχυσης για ταυτόχρονη σκιαγράφηση πνευμονικού και συστηματικού αρτηριακού δικτύου (μεγαλύτερη διάρκεια έγχυσης, ↑ threshold, ↑ delay)
- ✓ Λεπτές τομές submm (0,7mm). Μετεπεξεργασία σε πολλαπλά επίπεδα (MPR, VR, MIP)

Multi Detector CT ADVANCES

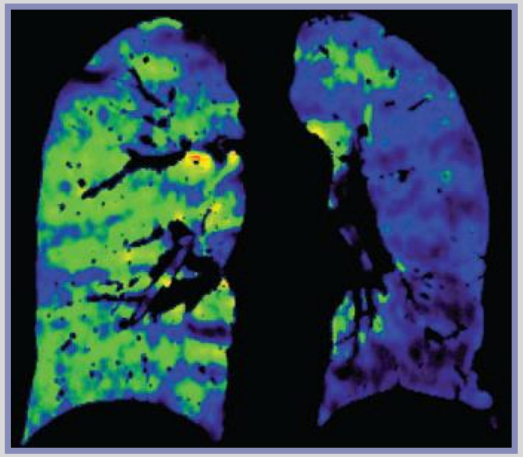
ECG Gated MDCT

- Assessment of RV functional parameters when the patient is unable to undergo MRI
- Increased radiation dose
- No blood flow or pressure data

Iterative Reconstruction Algorithms

Dose reduction up to 50%

Dual Energy CTPA - Pulmonary Perfusion



CT signs suggesting PH

CT signs suggesting the cause of PH

CT signs suggesting PH

CT signs suggesting PH

1. **Vascular Signs (Pulmonary Arteries)**

   Dilatation, loss of distensibility

2. **Cardiac Signs (Right Ventricle)**

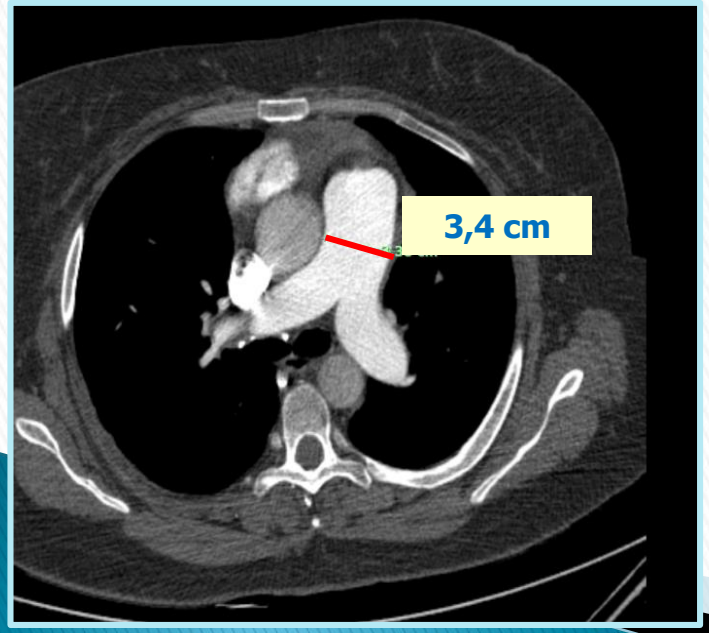
   Hypertrophy, dilatation

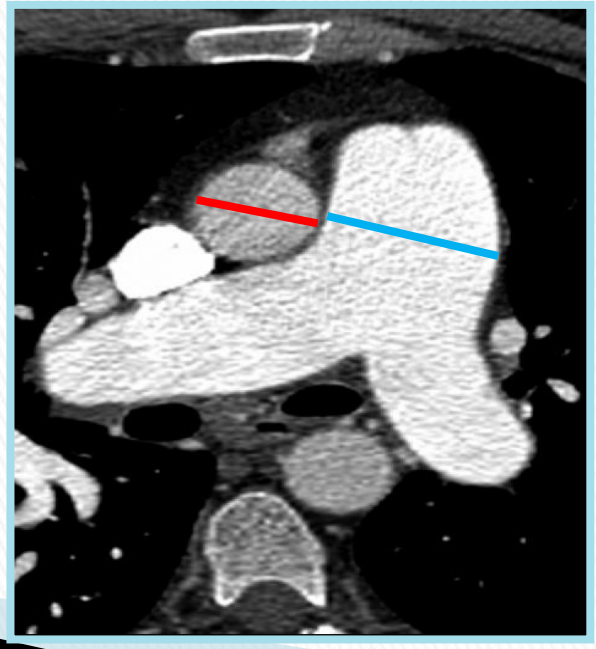
1. Vascular Signs: PA dilatation

1. **Diameter of main pulmonary artery ≥ 29 mm : Sens 79%, spec 83%**

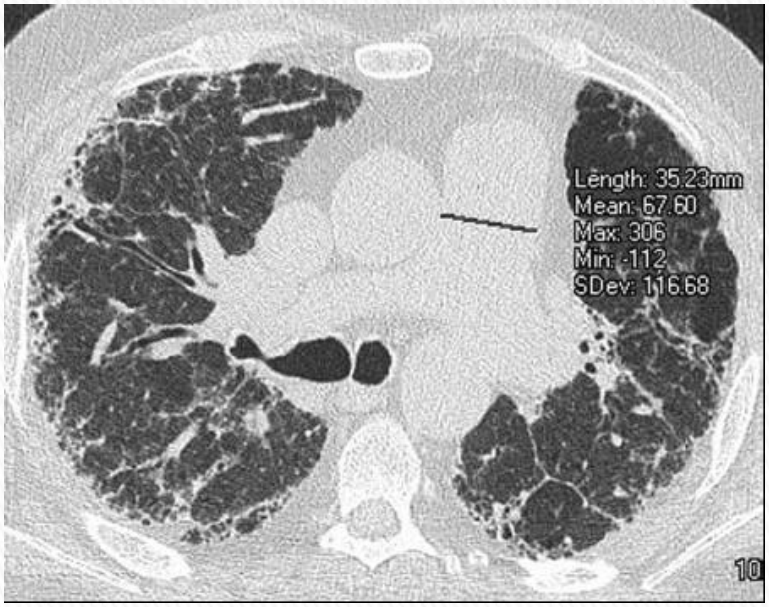
2. **Main PA diameter/ ascending aorta diameter ratio ≥ 1: sens 74%, sp 81%**
   Medicine 93(27):e256

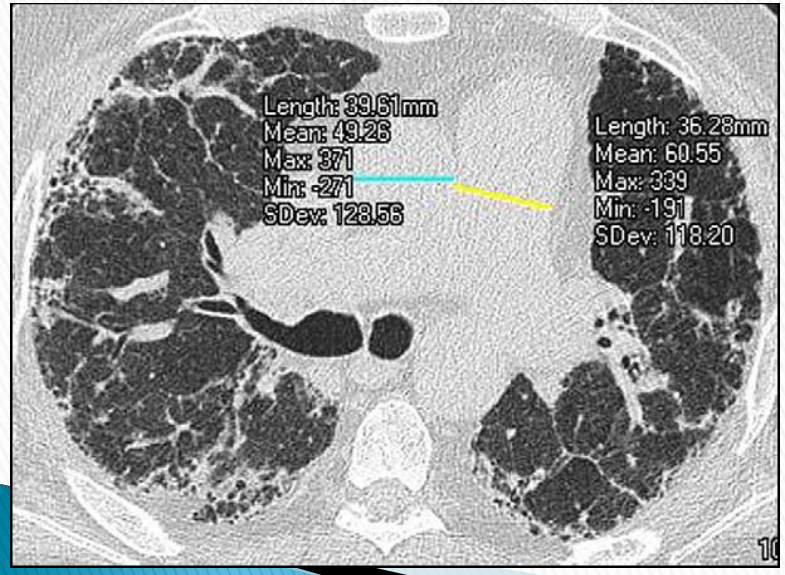
3. **Segmental artery-to-bronchus diameter ratio ≥ 1 in at least three lobes: Spec 100%** *RadioGraphics 2010; 30:1753–1777*





Segmental artery D/ bronchus D Ratio > 1





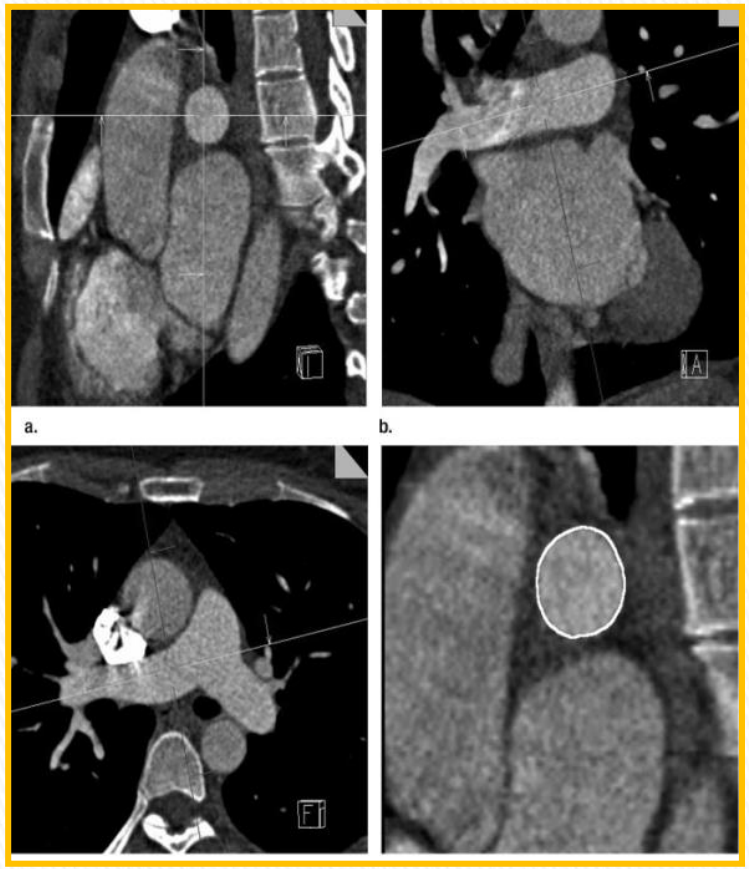
Εξαίρεση

Η διάμετρος της κύριας πνευμονικής αρτηρίας δεν έχει καλή συσχέτιση με την πίεση της πνευμονικής αρτηρίας (mPAP) σε ασθενείς με *πνευμονική ίνωση*. Σε αυτή την περίπτωση, πιο αξιόπιστος δείκτης είναι ο λόγος διαμέτρων ΠΑ/ ΑΑ

Zisman DA, Karlamangla AS, Ross DJ, et al. High resolution chest CT findings do not predict the presence of pulmonary hypertension in advanced idiopathic pulmonary fibrosis. Chest 2007;132:773-9

Devaraj A, Wells AU, Meister MG, et al. The effect of diffuse pulmonary fibrosis on the reliability of CT signs of pulmonary hypertension. Radiology 2008;249:1042-9.

1. Vascular signs: Decrease in RPA distensibility



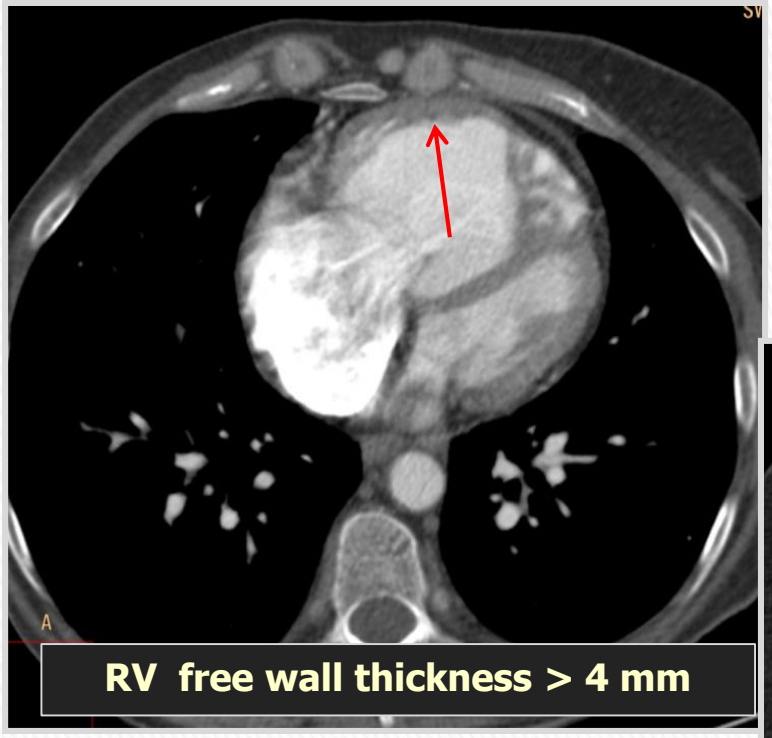
Revel MP et al. Pulmonary hypertension: ECG-gated 64-section CT angiographic evaluation of new functional parameters as diagnostic criteria. *Radiology 2009;250(2):558–566*

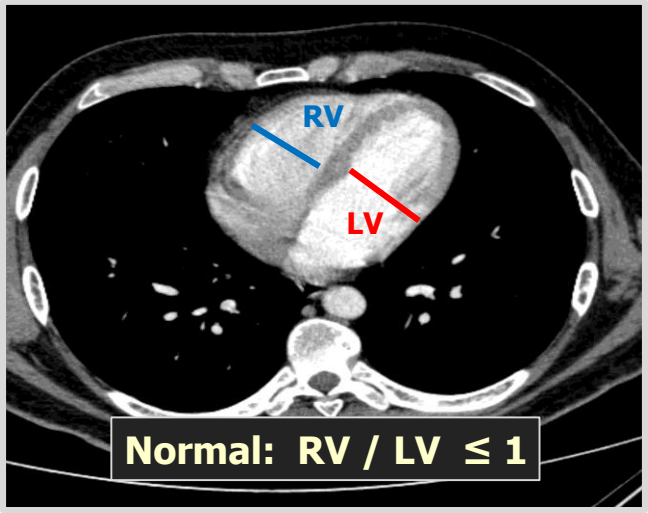
Decrease in right pulmonary artery distensibility

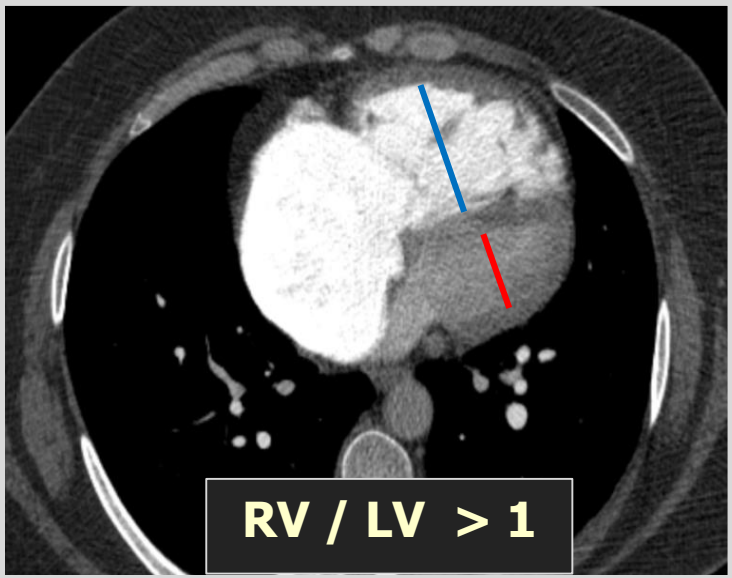
The best pulmonary artery distensibility cutoff for distinguishing between patients with pulmonary hypertension and those without it was 16.5% when a standard method of measurement was used and 13% when a simplified method was used

2. Cardiac Signs

1. **Right Ventricular Hypertrophy**
   RV myocardium more than 4 mm thick
2. **RV dilatation**
   RV/LV diameter ratio ≥ 1
3. **Straightening or leftward bowing of the interventricular septum**



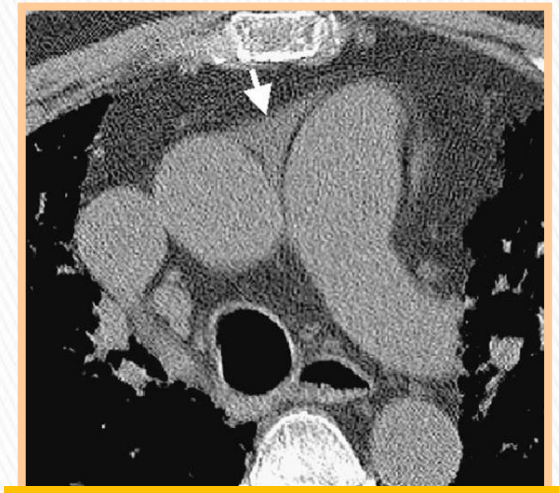




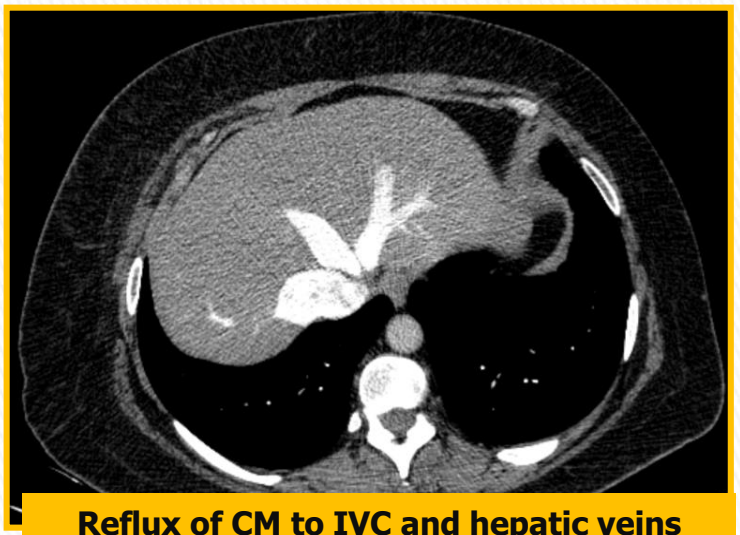
straightening of the IVS, D-shaped LV

… and

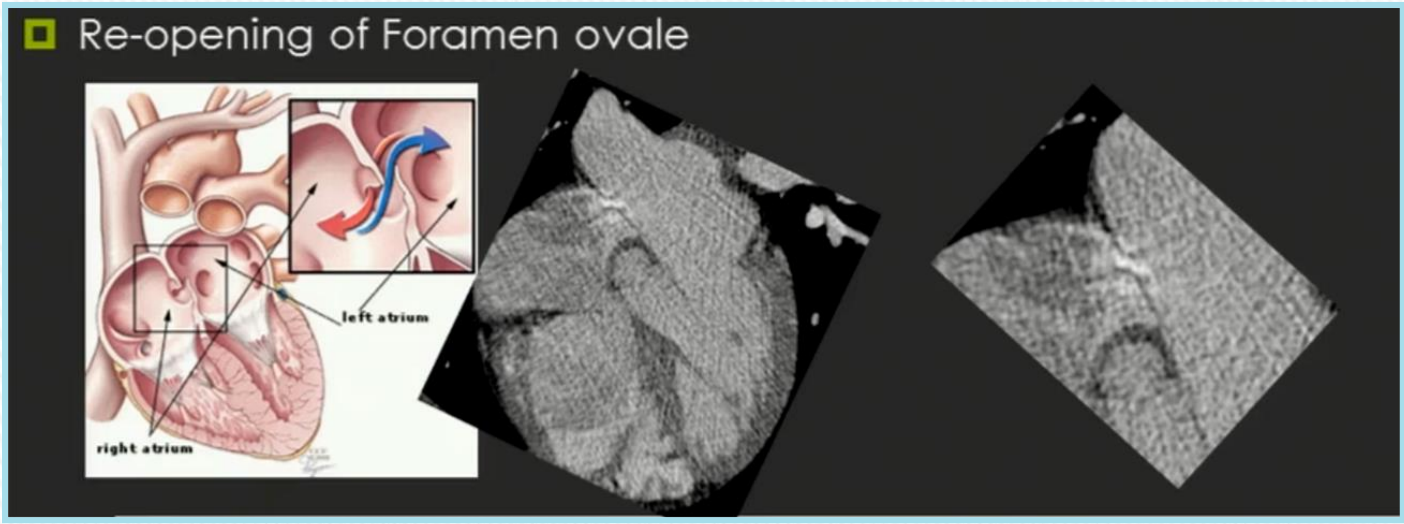
1. Pericardial effusion
2. Reflux of contrast material into IVC and hepatic veins
3. Reopening of foramen ovale



Pericarsial effusion
POOR prognostic sign



Reflux of CM to IVC and hepatic veins

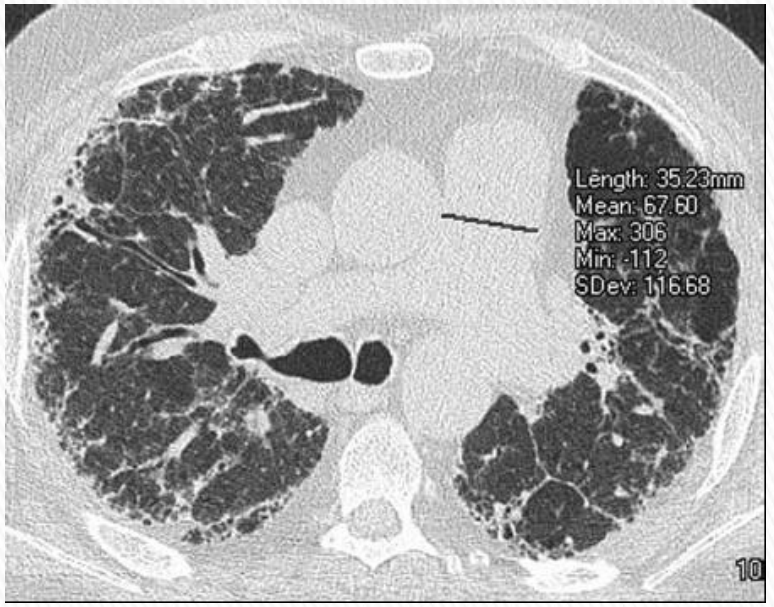


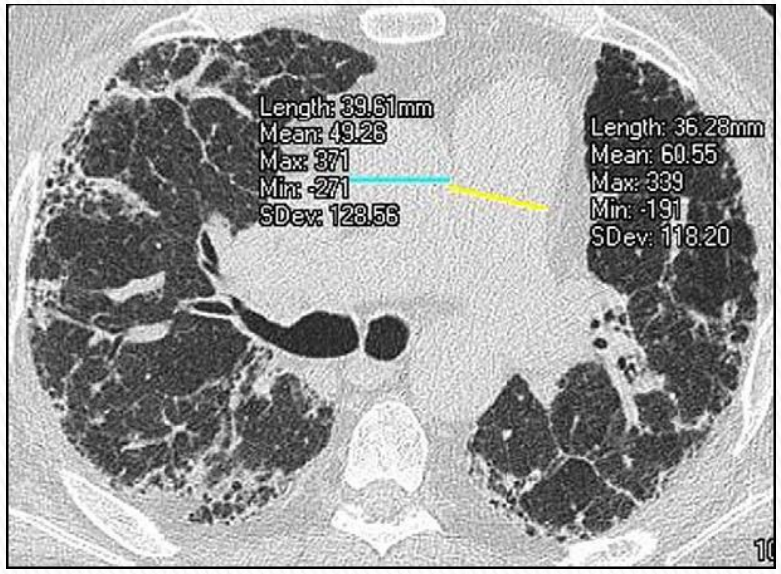
Marie-Pierre LEVEL ECR 2015

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 mild pulmonary hypertension**
- ✓ **Ascending aorta diameter may change with body size and/or age**
- ✓ **PA dilatation may also occur without PH in patients with chronic diffuse pulmonary fibrosis**

Devaraj A et al. Radiology 2008;249:1042-9





Devaraj et al. A Radiology 2008;249:1042-9

CT signs suggesting the cause of PH

GROUP 1' Pulmonary Veno-Occlusive Disease *(PVOD)* Pulmonary Capillary Haemagiomatosis *(PCH)*

- ✓ Rare conditions. Pathological, clinical and radiological overlap
- ✓ Many cases are still misclassified as IPAH. The prognosis appears to be worse
- ✓ PAWP is almost invariably normal
- ✓ Hazardous lung biopsy (the gold standard to confirm a histological diagnosis of PVOD/ PCH), is no longer recommended in most cases
- ✓ The only curative therapy for PVOD/PCH is lung transplantation

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

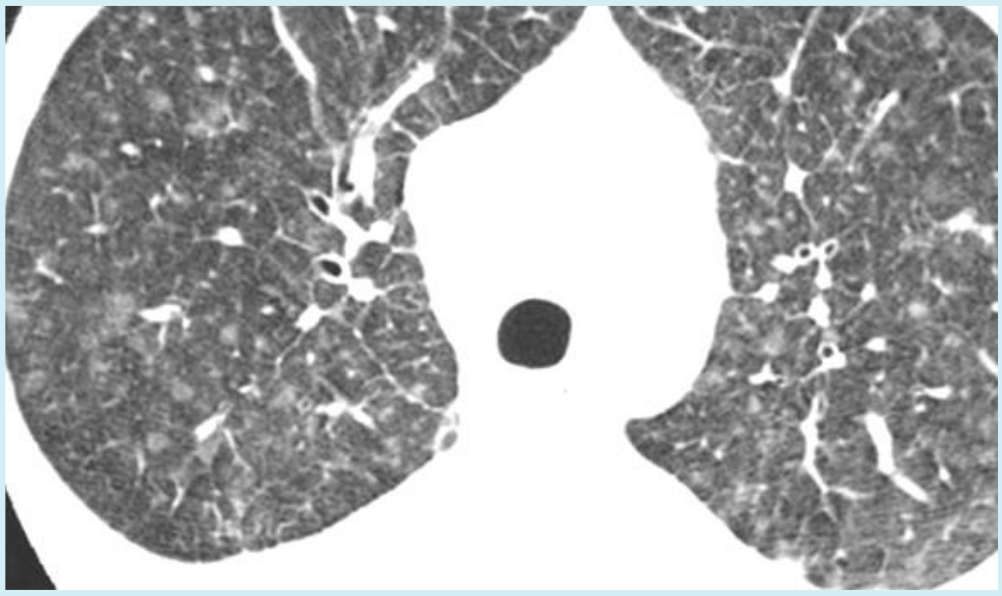
GROUP 1' : PVOD/PCH

High-resolution CT of the chest is the investigation of choice.
V/Q lung scanning is not useful in discriminating PVOD/PCH from IPAH

Specific CT signs

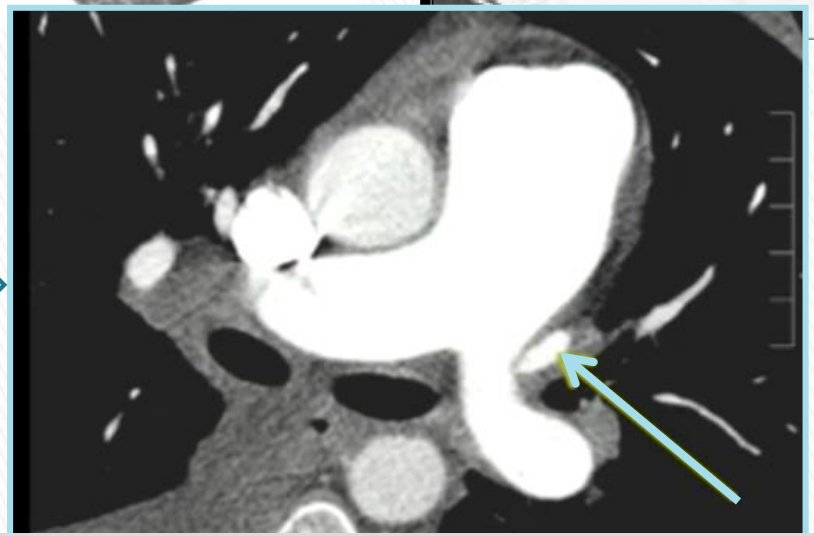
1. **Smoothly thickened interlobular septa**
2. **Poorly defined centrilobular ground glass opacities**
3. **Lymphadenopathy**

Sens 66% Spec 100% Resten et al. AJR 2004;183:65-70



GROUP 1' : PVOD/PCH

In PVOD



… small central pulmonary veins

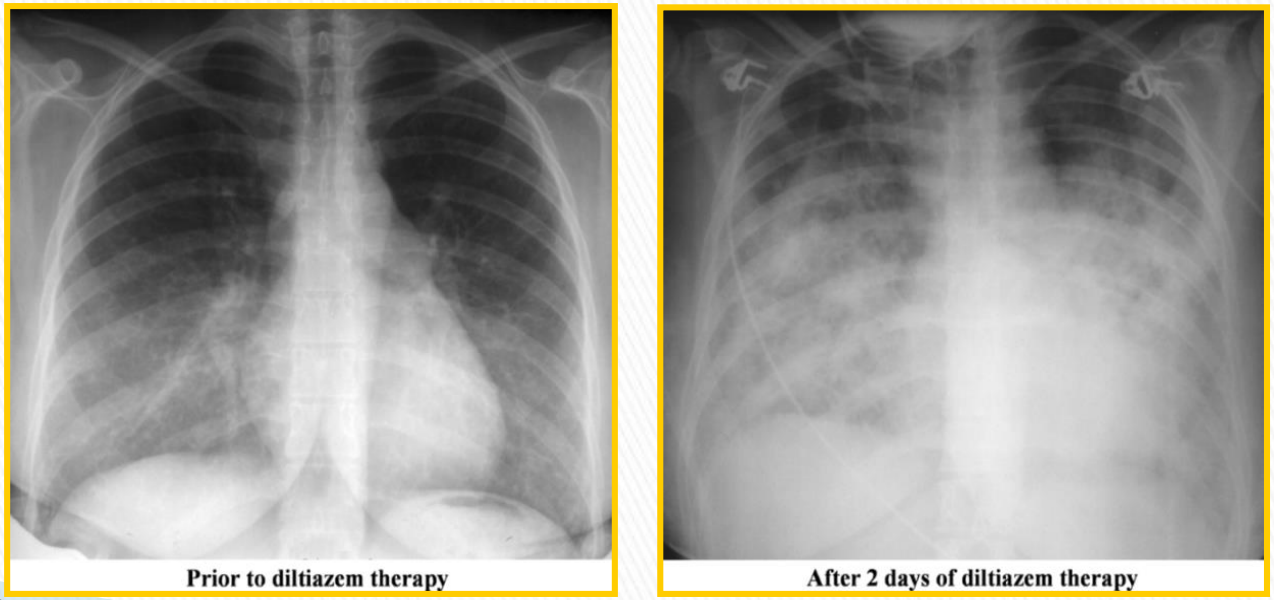
… and normal PAWP (< 15 mmHg)

GROUP 1' : PVOD/PCH

Rare, difficult to diagnose

But, no room for error

Vasodilators and, in particular, i.v. epoprostenol must be used with great caution because of the high risk of severe drug-induced pulmonary oedema

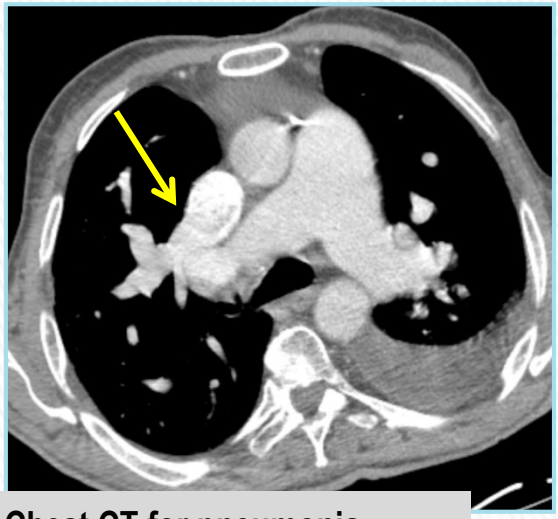


Prior to diltiazem therapy

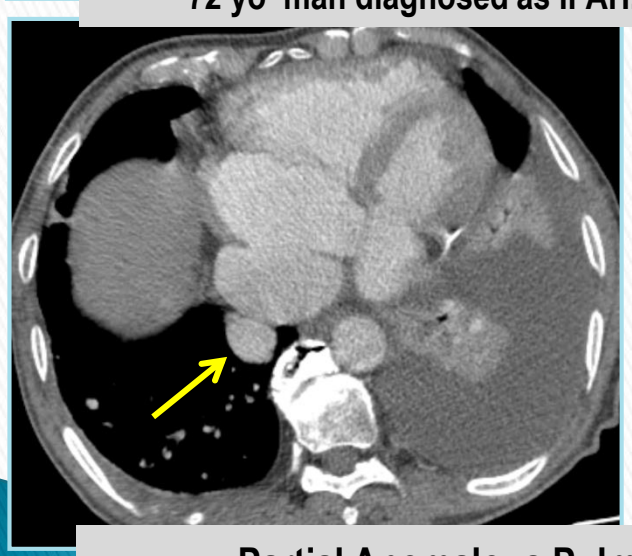
After 2 days of diltiazem therapy

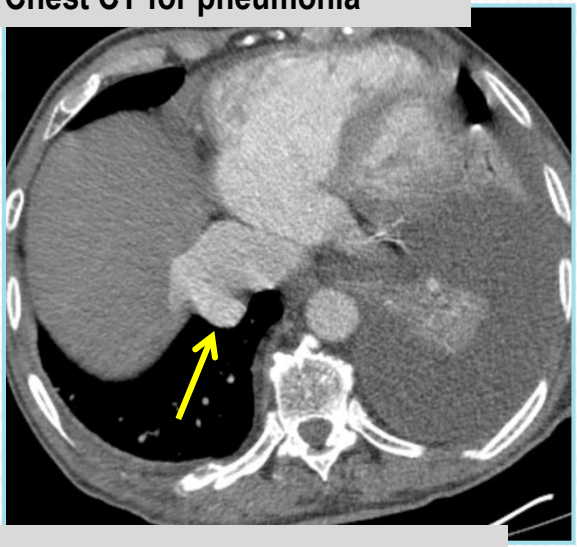
David Montani et al. Medicine 2008;87:220–233

GROUP 1: PAH associated with Congenital Heart Disease (CHD)



72 yo man diagnosed as IPAH. Chest CT for pneumonia





Partial Anomalous Pulmonary Venous Return

Some malformations, such as *patent ductus arteriosus, sinus venosus atrial septal defect or partial anomalous pulmonary venous return*, are often concealed and patients are misclassified as suffering from IPAH. Hence, these congenital anomalies should be specifically sought

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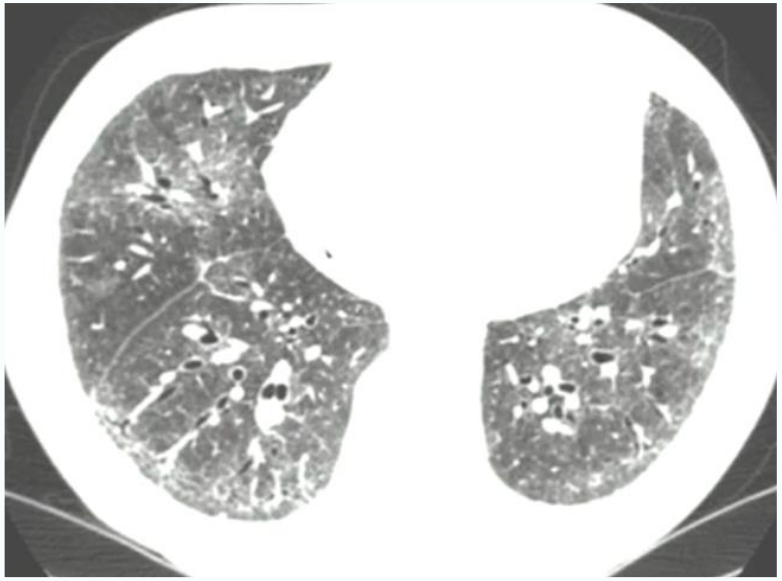
GROUP 1: Associated with CTD (mainly SSc, SLE , mixed CTD)

Specific CT features

Infiltrative lung disease, mainly NSIP in SSc

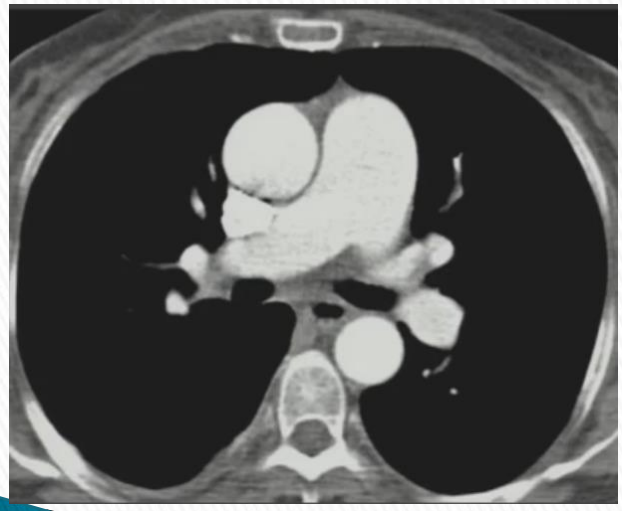


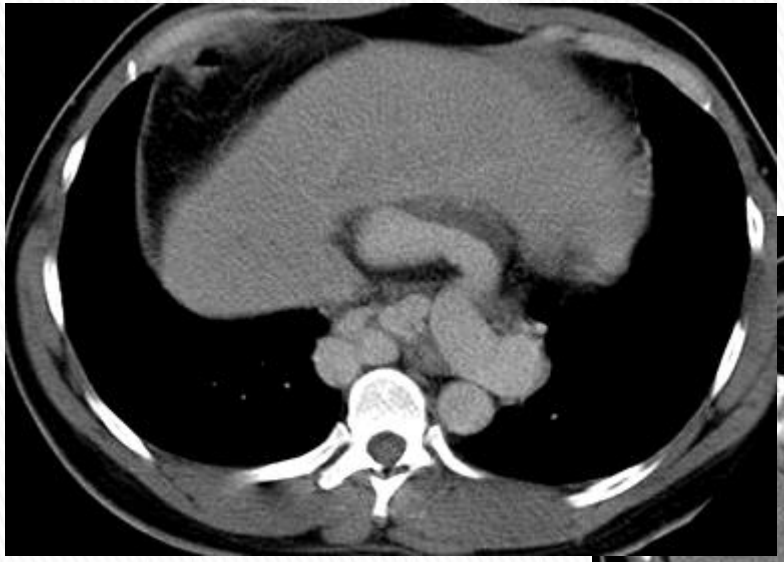
Άνδρας 62 ετών, CREST

GROUP 1: Associated with portal hypertension

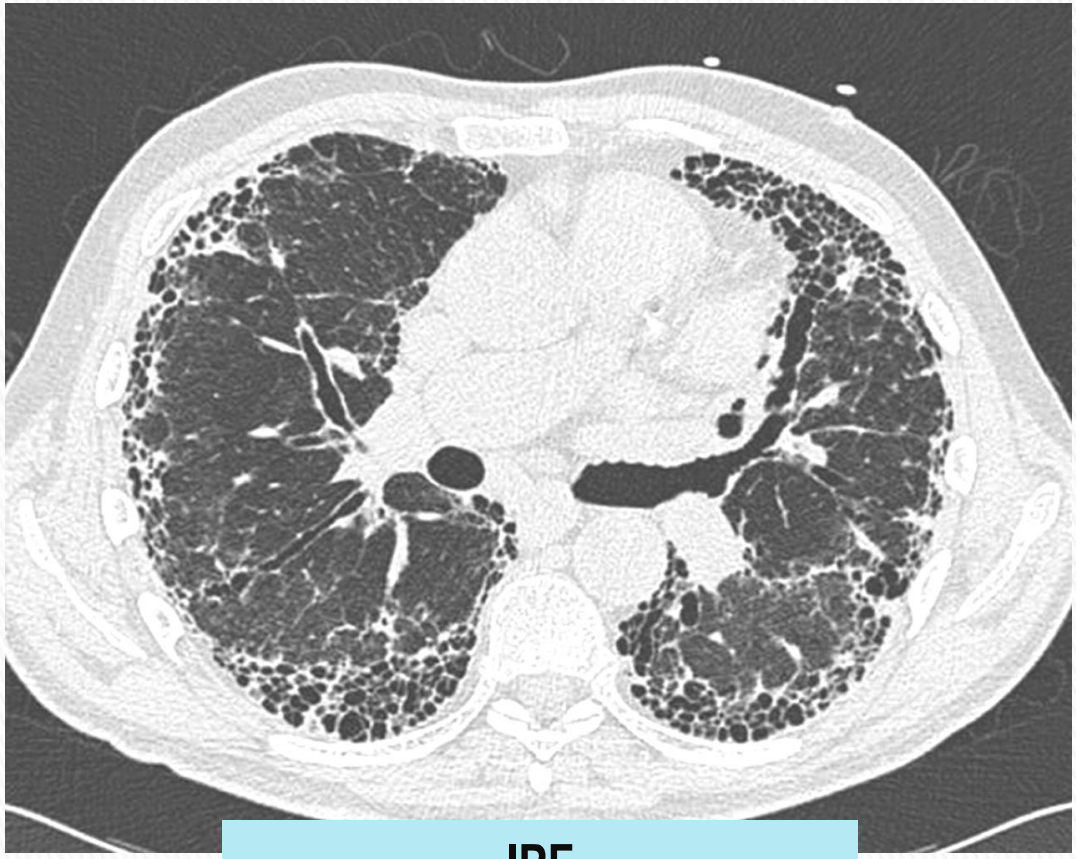
Specific CT features

Those related to portal hypertension (splenomegaly, esophageal varices, ascites …)

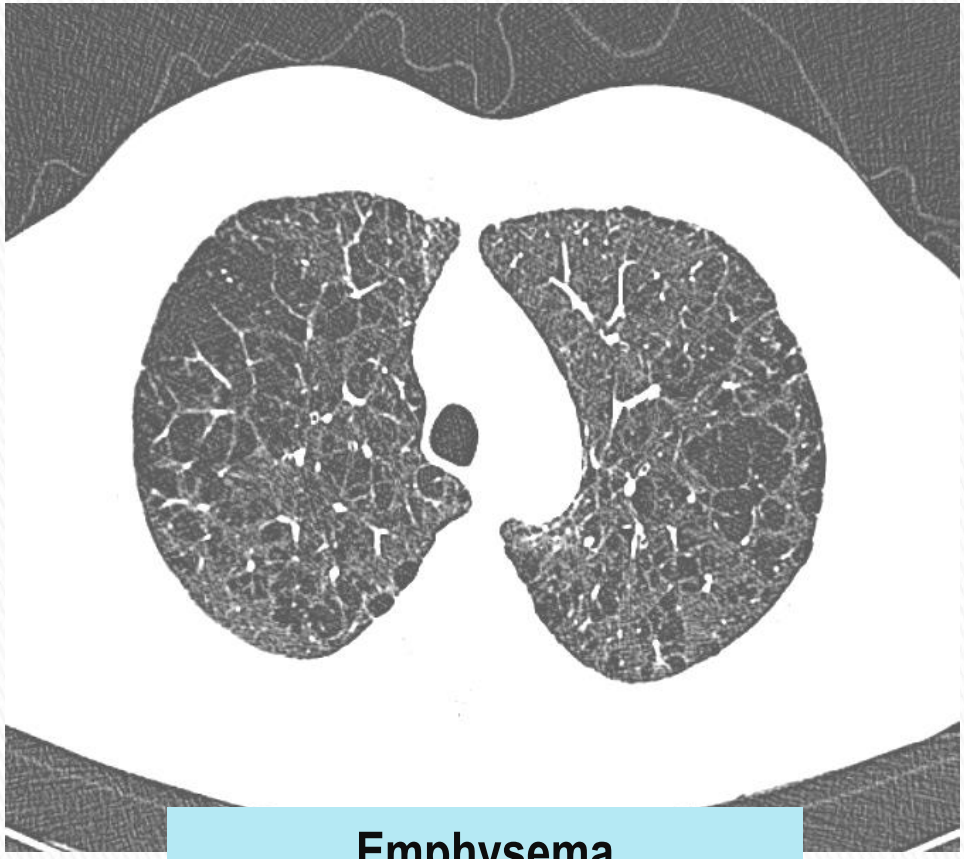




GROUP 3: PH due to lung diseases



IPF



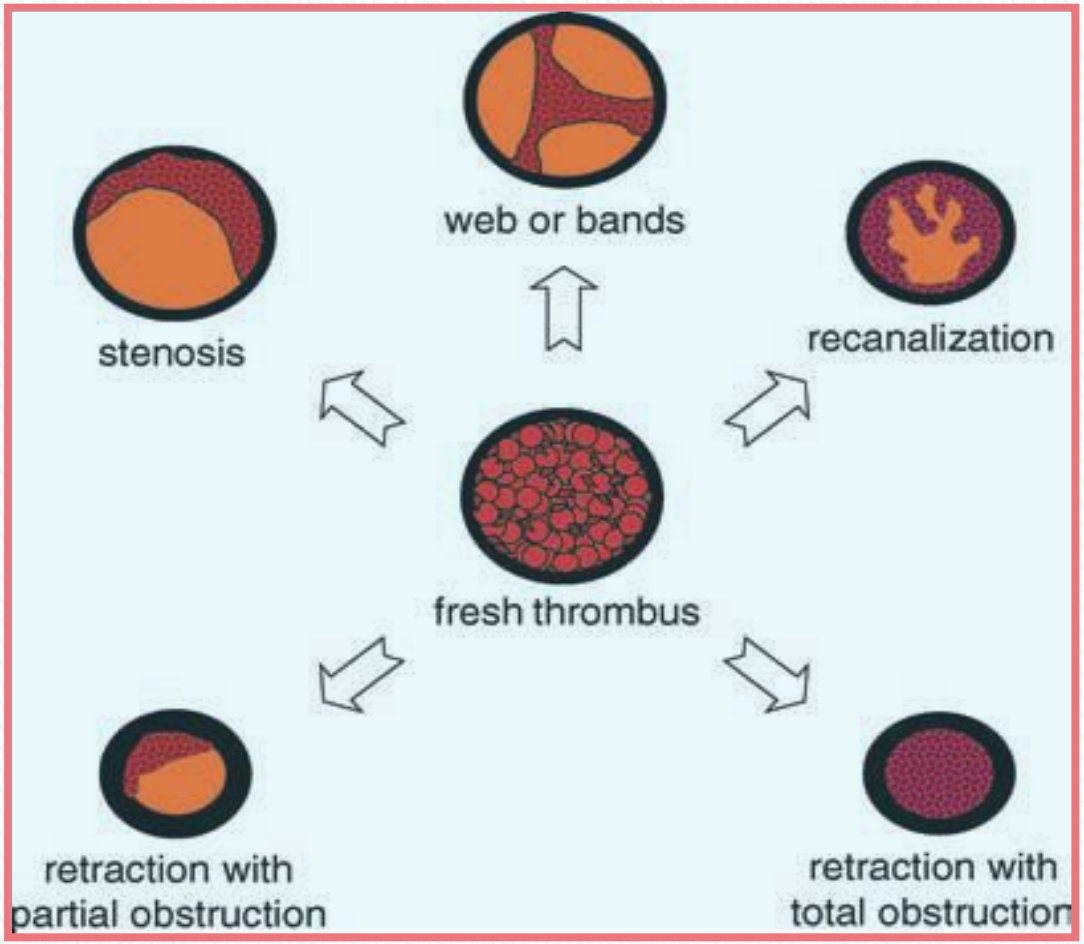
Emphysema

GROUP 4 : Chronic Thrombo Embolic Pulmonary Hypertension (CTEPH)

✓Complicates up to 3,8 % of acute PE

✓ Routine screening for CTEPH after PE is not supported by current evidence

✓It is of the utmost importance to diagnose CTEPH early and, further, to recognize patients eligible for PEA.

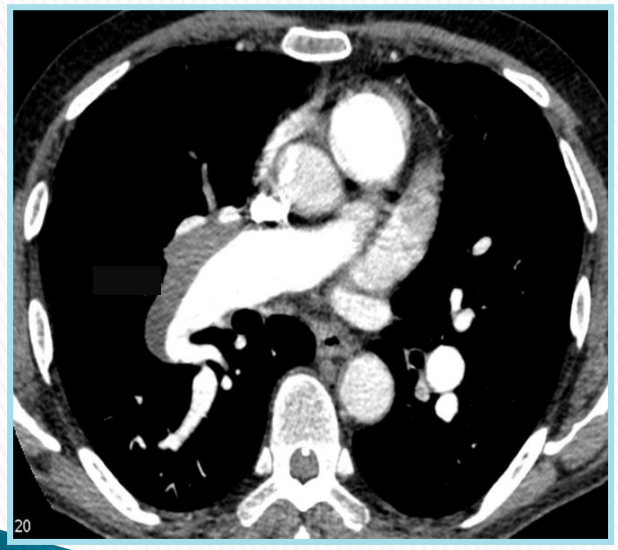


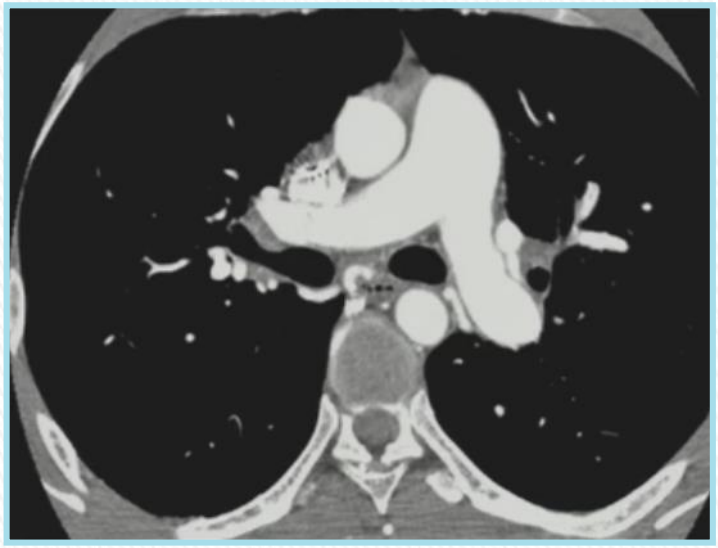
Eva Castañer et al. RadioGraphics 2009; 29:31–53

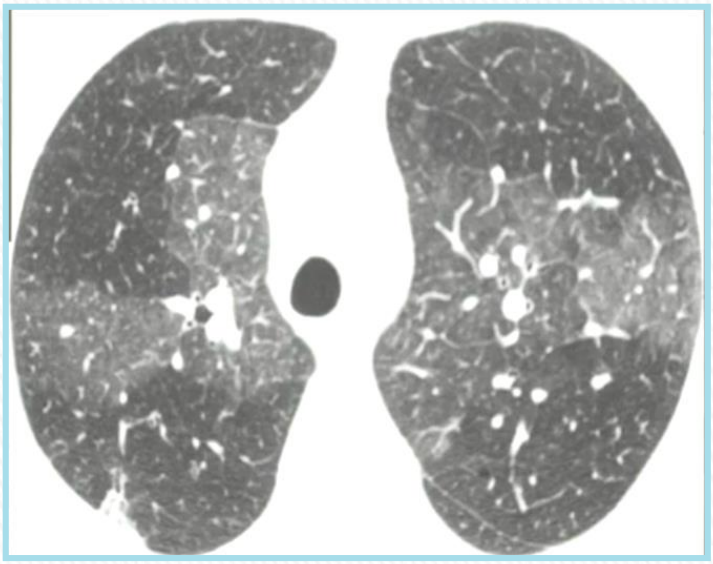
GROUP 4: CTEPH

Specific CT signs

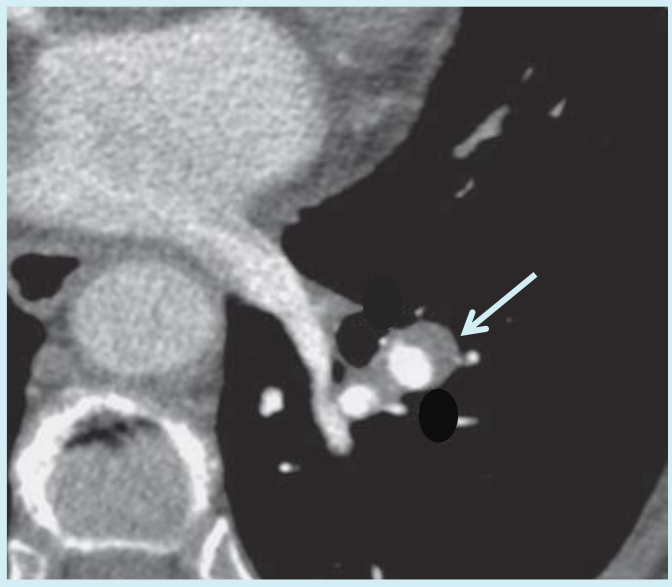
1. **Signs of chronic PE**
2. **Enlarged systemic arteries**
3. **Mosaic perfusion**

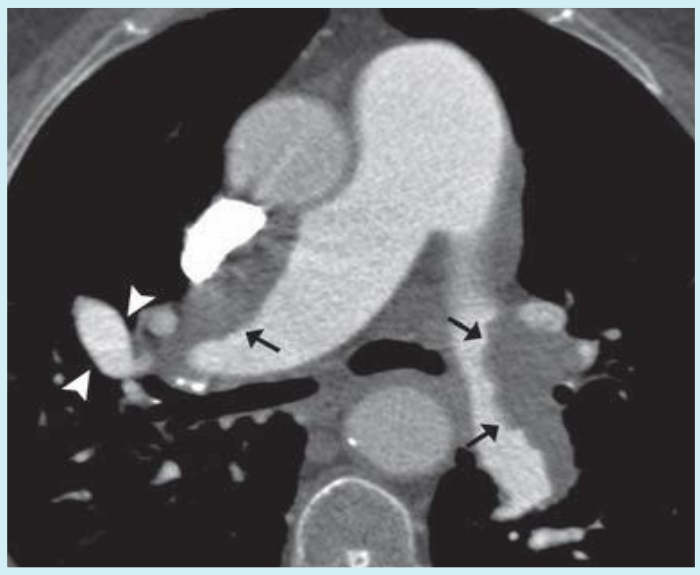




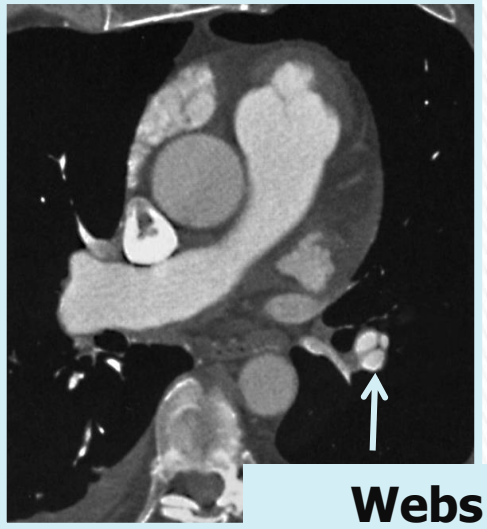


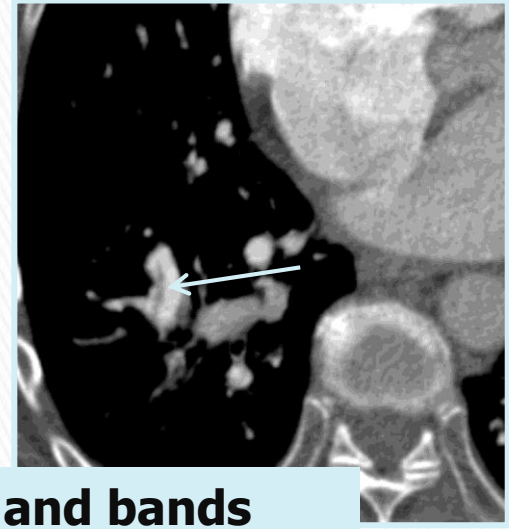
GROUP 4: CTEPH



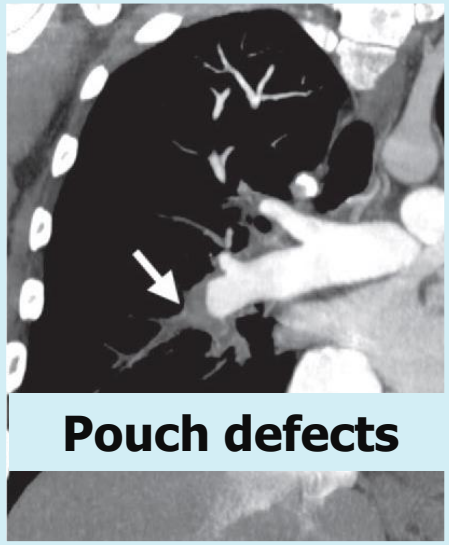


Eccentric thrombi Obtuse angle between thrombus and vessel wall





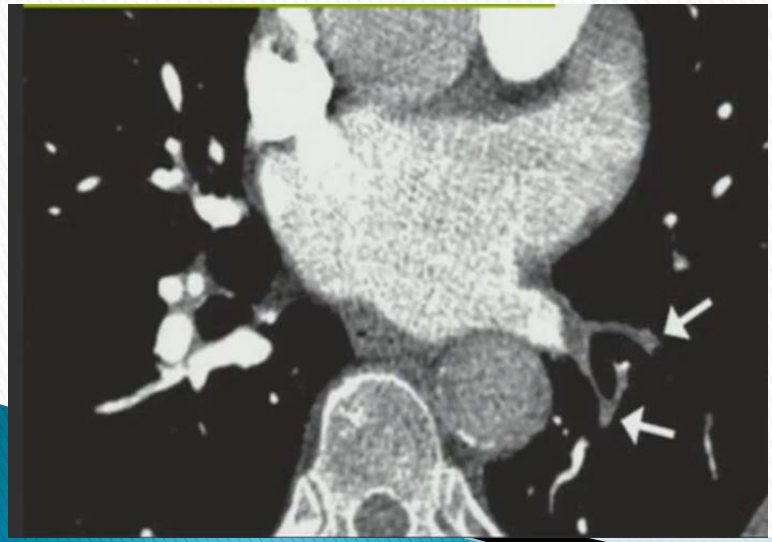
Webs and bands

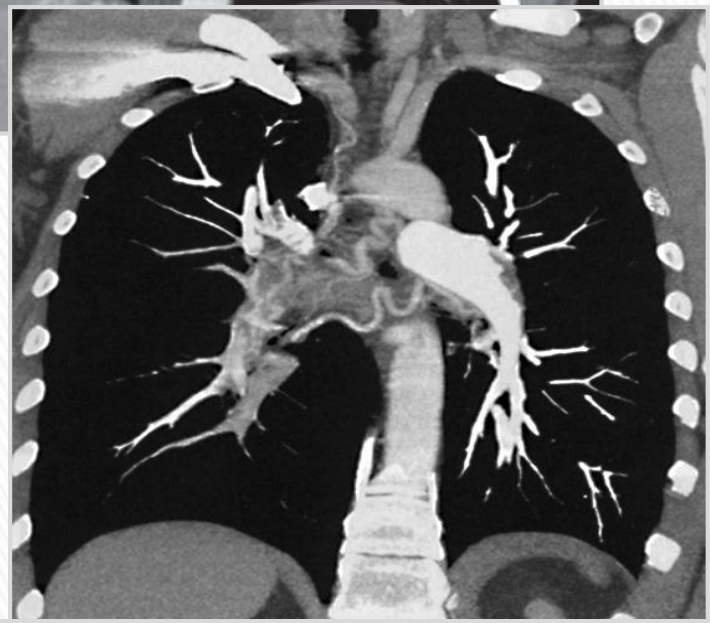


Pouch defects

GROUP 4: CTEPH

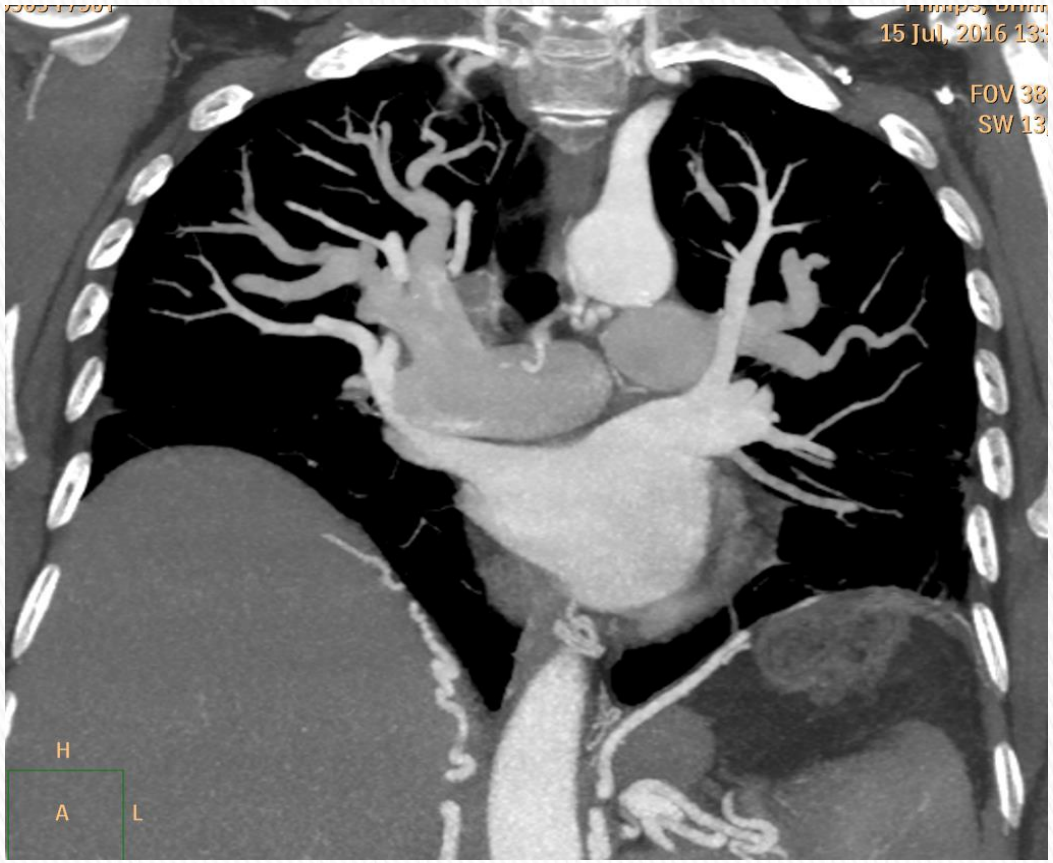
Small size occluded vessels

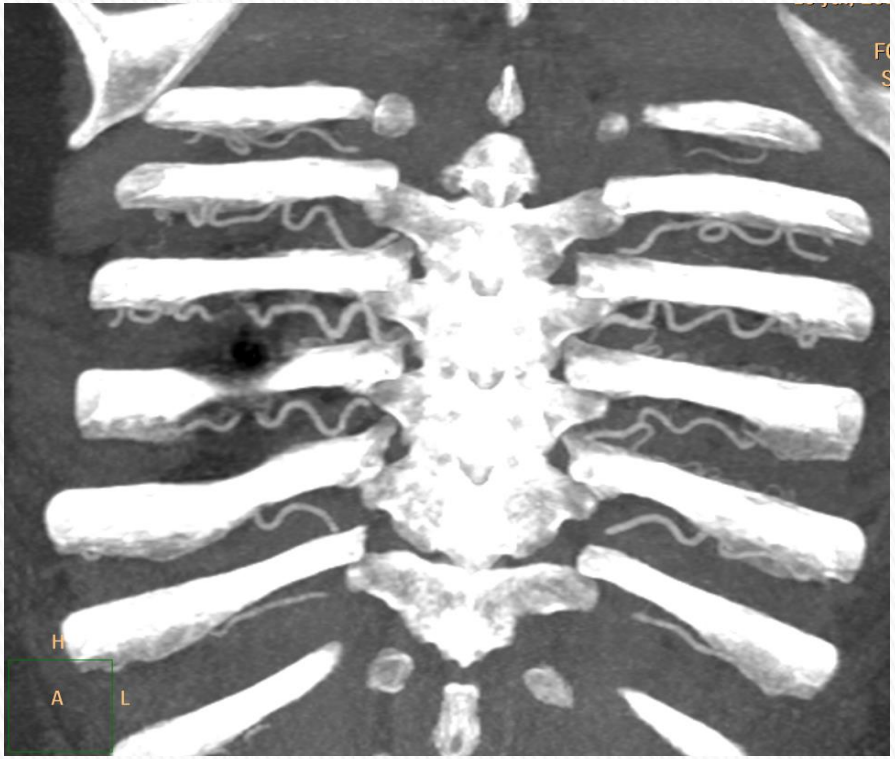




Enlarged bronchial arteries

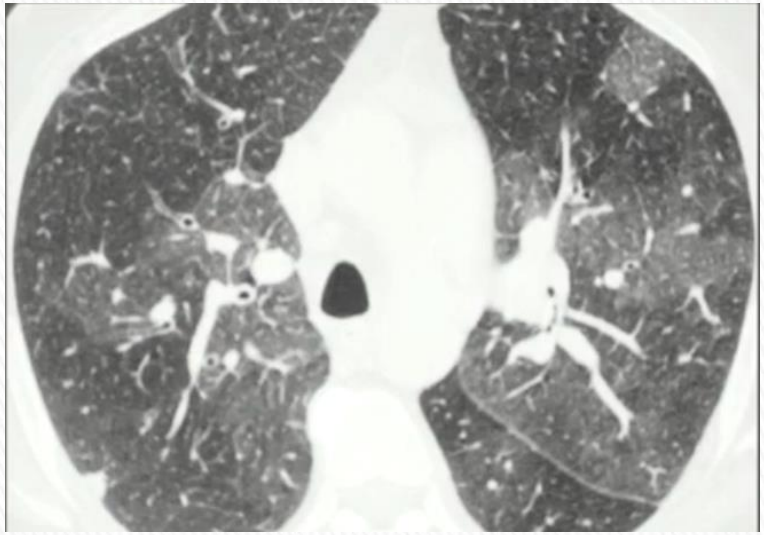
GROUP 4: CTEPH





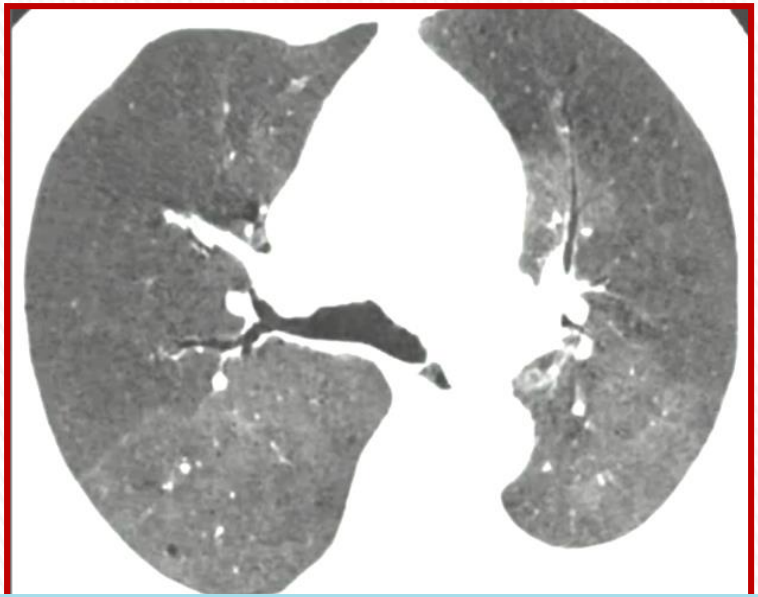
Systemic collaterals

GROUP 4: CTEPH



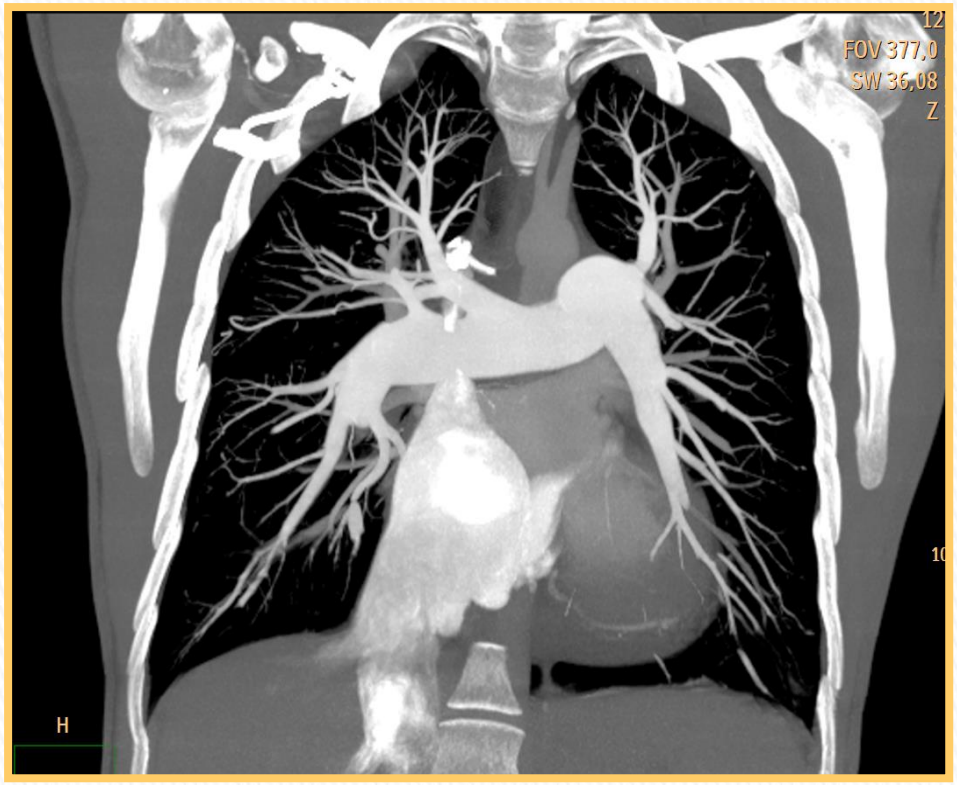
Mosaic Perfusion

Mosaic perfusion can be subtle



Minimum Intensity Projection (MinIP)

CT Pulmonary Artery Angiography: in the work-up for CTEPH



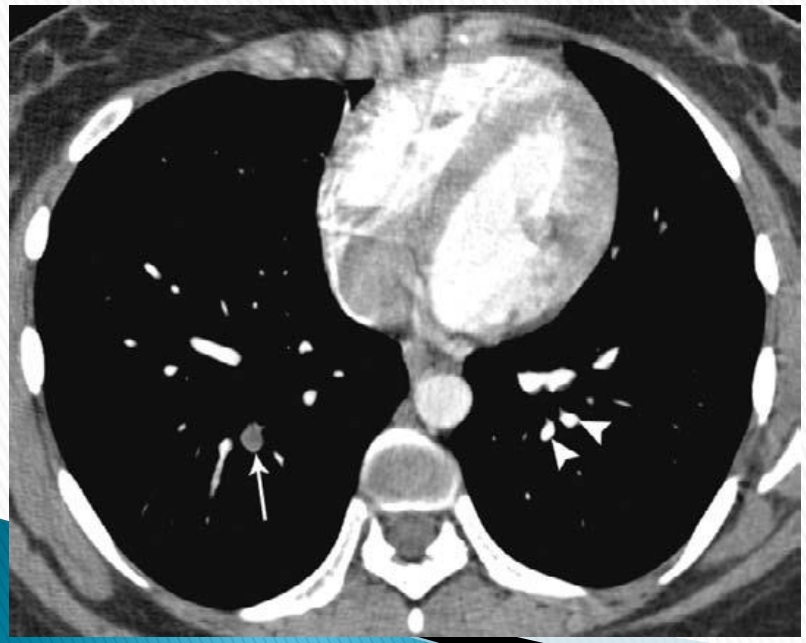
IPAH

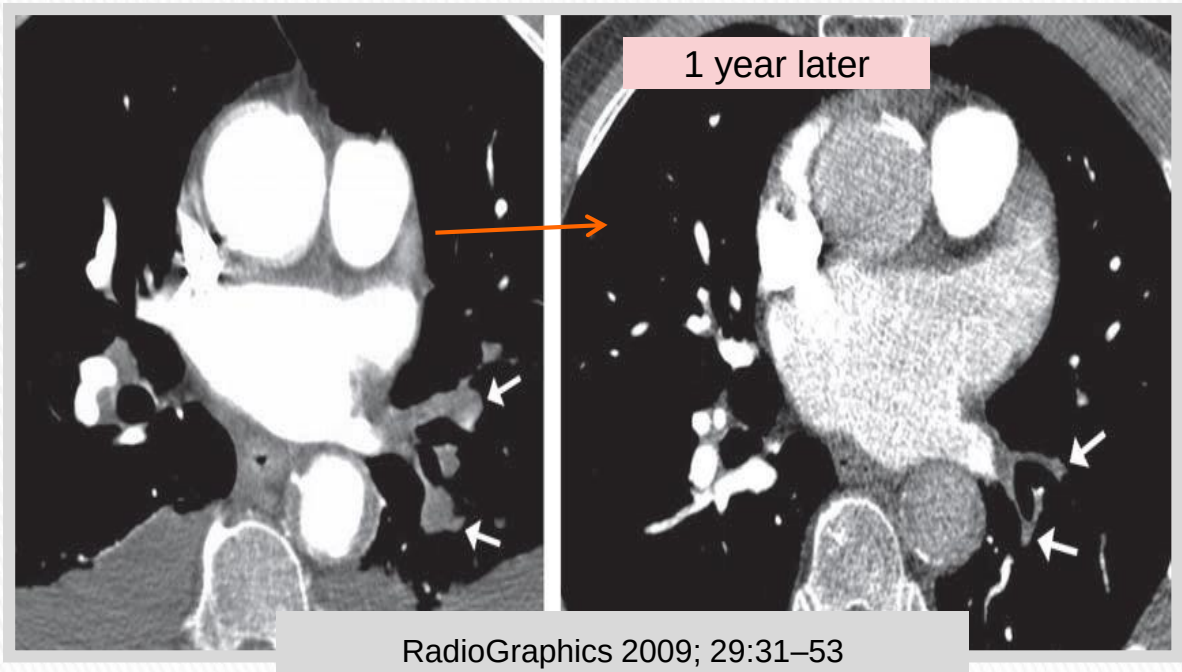
CTEPH

Key clinical question : Dd between PAH and CTEPH

Οξεία vs Χρόνια

ΟΞΕΙΑ ΠΕ	ΧΡΟΝΙΑ ΠΕ
Αύξηση της διαμέτρου των αγγείων	**Ελάττωση** της διαμέτρου των αγγείων

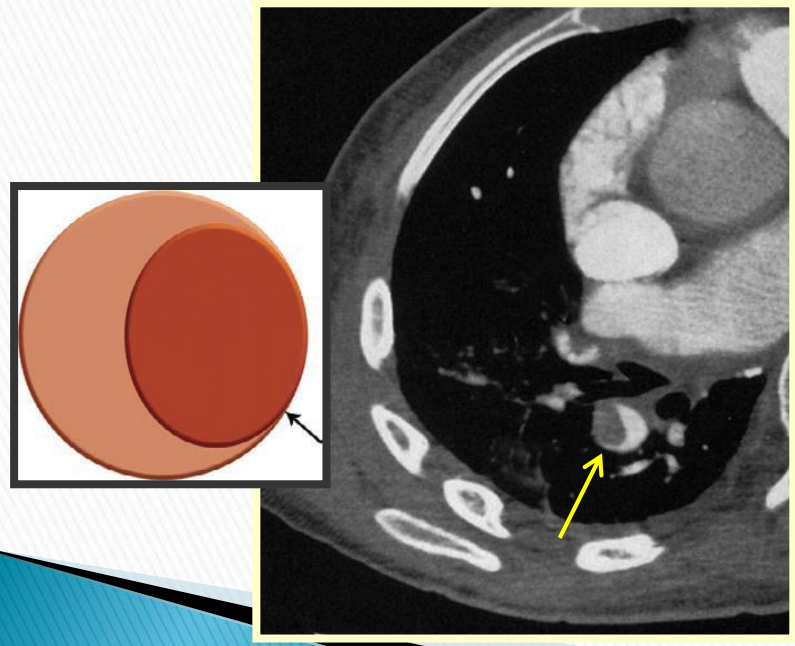


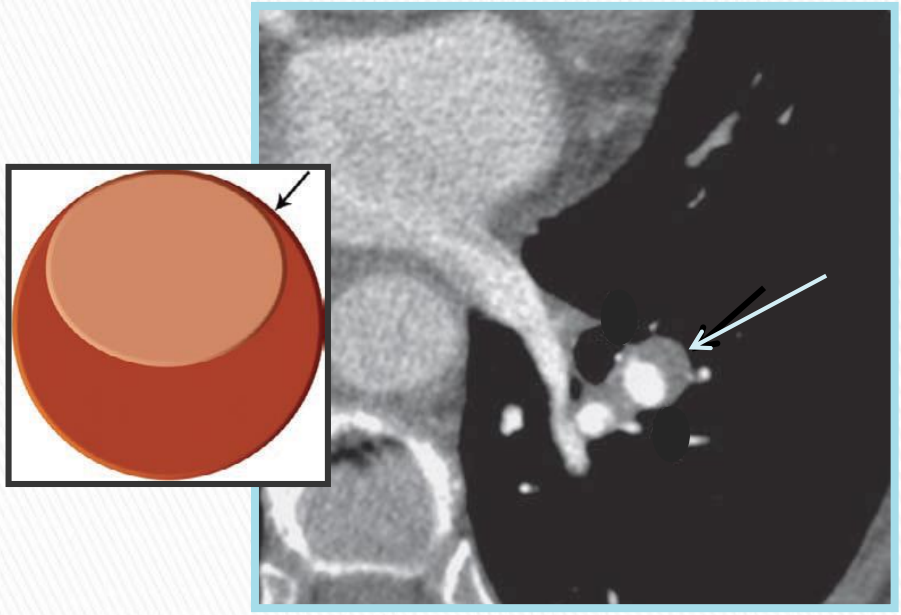


RadioGraphics 2009; 29:31–53

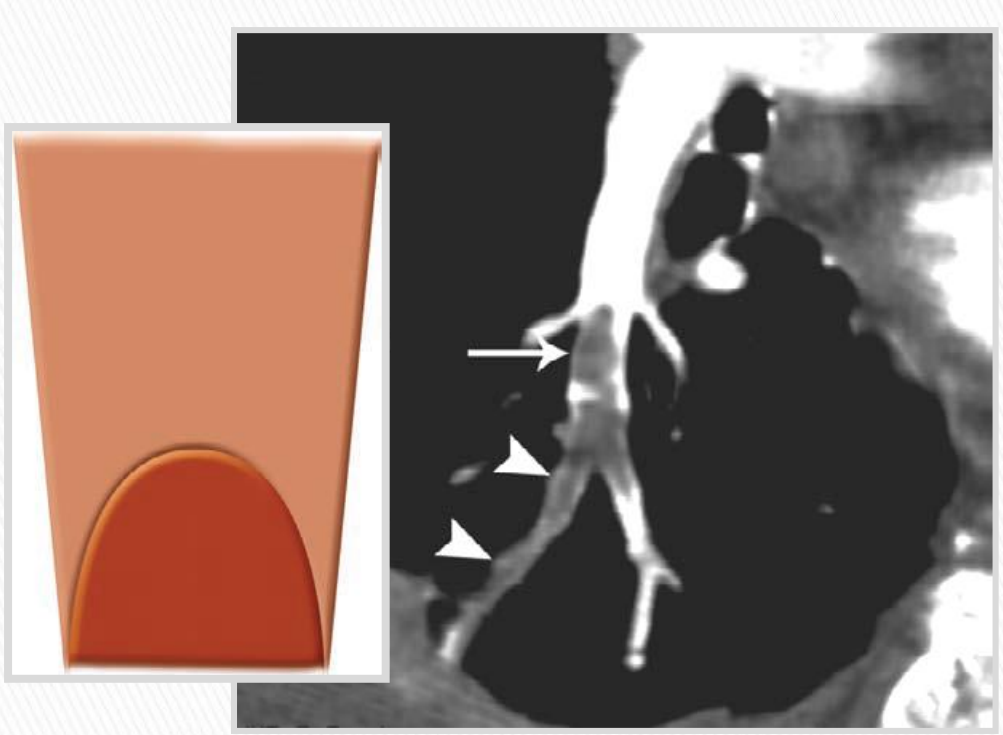
Οξεία vs Χρόνια

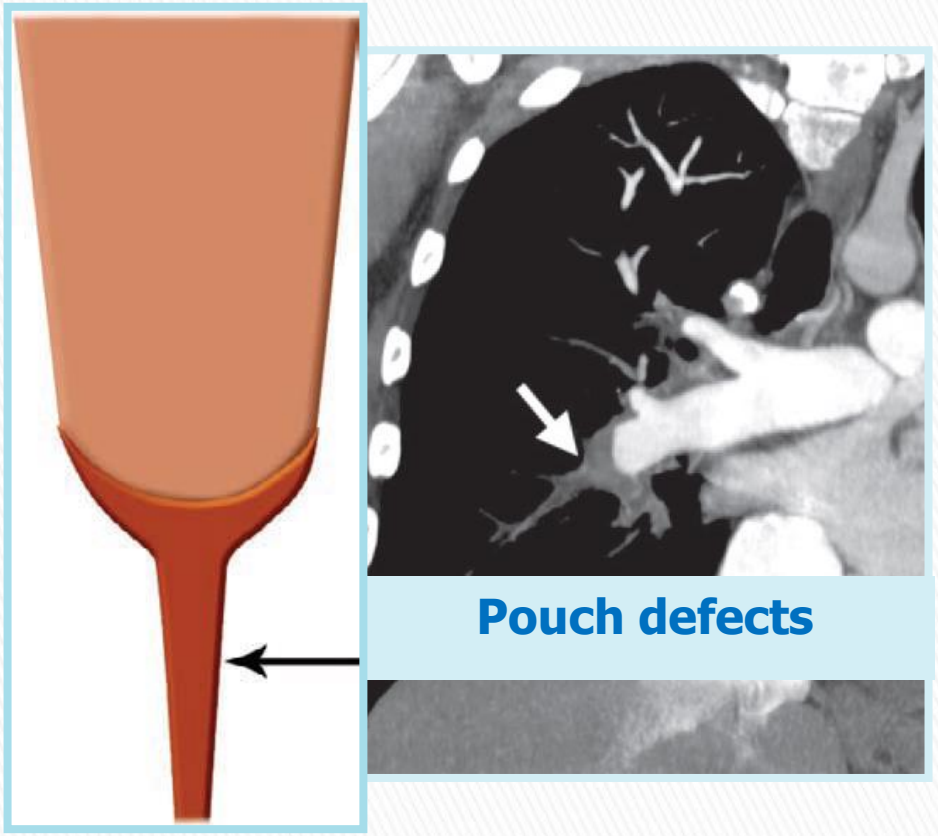
ΟΞΕΙΑ ΠΕ	ΧΡΟΝΙΑ ΠΕ
Οξεία γωνία του θρόμβου με το τοίχωμα του αγγείου	**Αμβλεία γωνία** του θρόμβου με το τοίχωμα του αγγείου





Οξεία vs Χρόνια





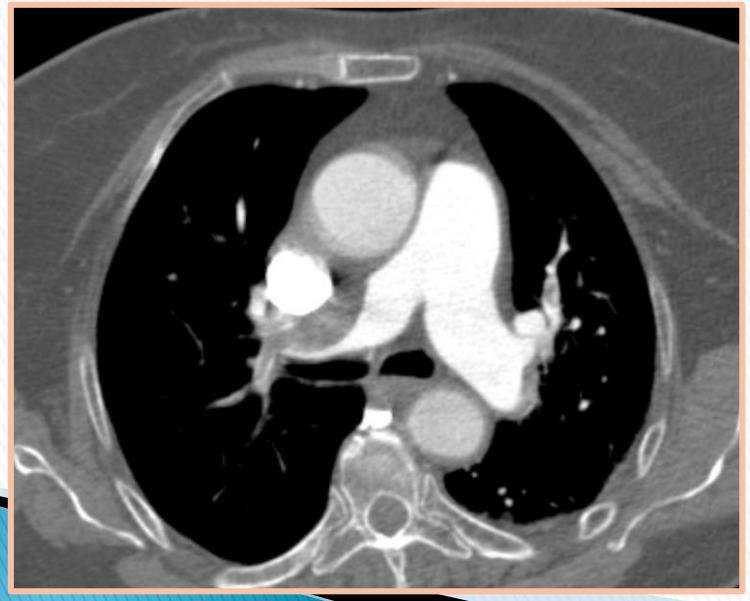
Pouch defects

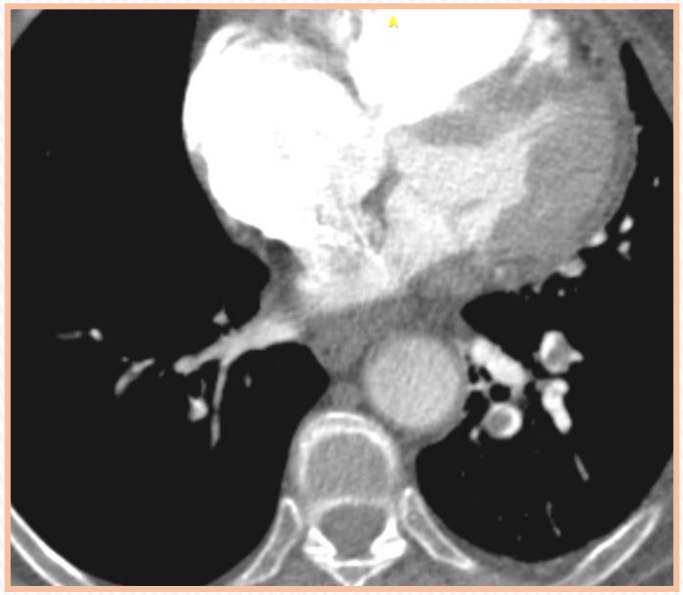
Οξεία vs Χρόνια

ΟΞΕΙΑ ΠΕ	ΧΡΟΝΙΑ ΠΕ
Διάταση (όχι υπερτροφία) της δεξιάς κοιλίας	**Διάταση και υπερτροφία** της δεξιάς κοιλίας
Ασυνήθης η διάταση των βρογχικών αρτηριών	**Συνήθης** η διάταση των βρογχικών αρτηριών

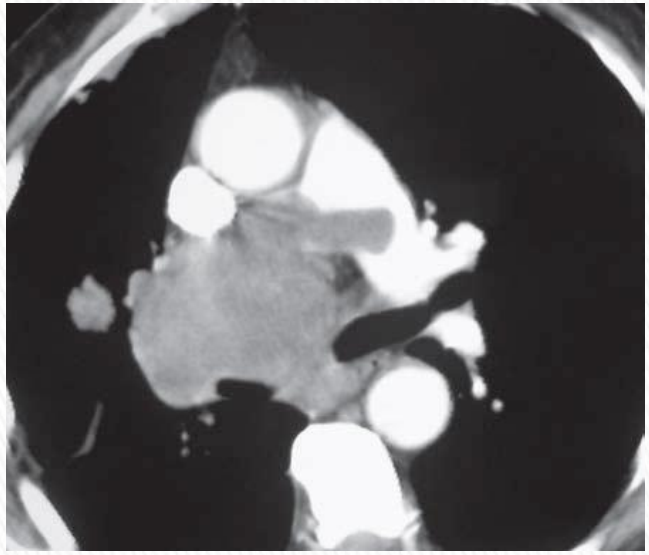
Οξεία vs Χρόνια

- Δεν είναι σπάνιο να συνυπάρχουν σημεία οξείας και χρόνιας εμβολής (Υποτροπιάζουσα πνευμονική εμβολή)
- Η χρόνια πνευμονική εμβολή πολλές φορές αναδεικνύεται σε CTPA που γίνονται για τον αποκλεισμό οξείας πνευμονικής εμβολής
- Η παρουσία διατεταμένων βρογχικών αρτηριών ή υπερτροφίας της δεξιάς κοιλίας είναι ευρήματα που υποστηρίζουν διάγνωση υποτροπιάζουσας ή χρόνιας πνευμονικής εμβολής





Mimics of CTEPH



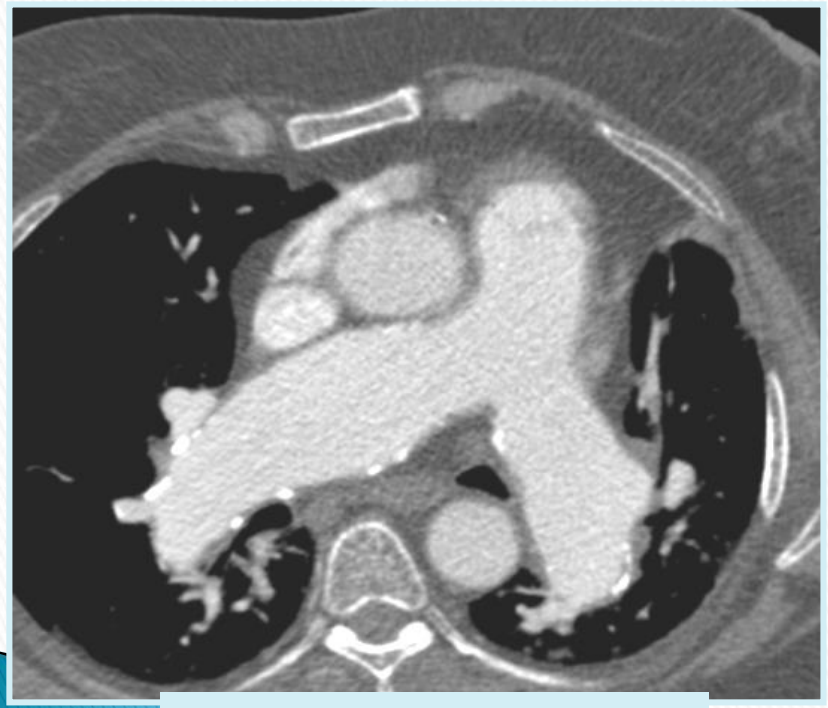
Pulm. Artery sarcoma

Fibrosing mediastinitis

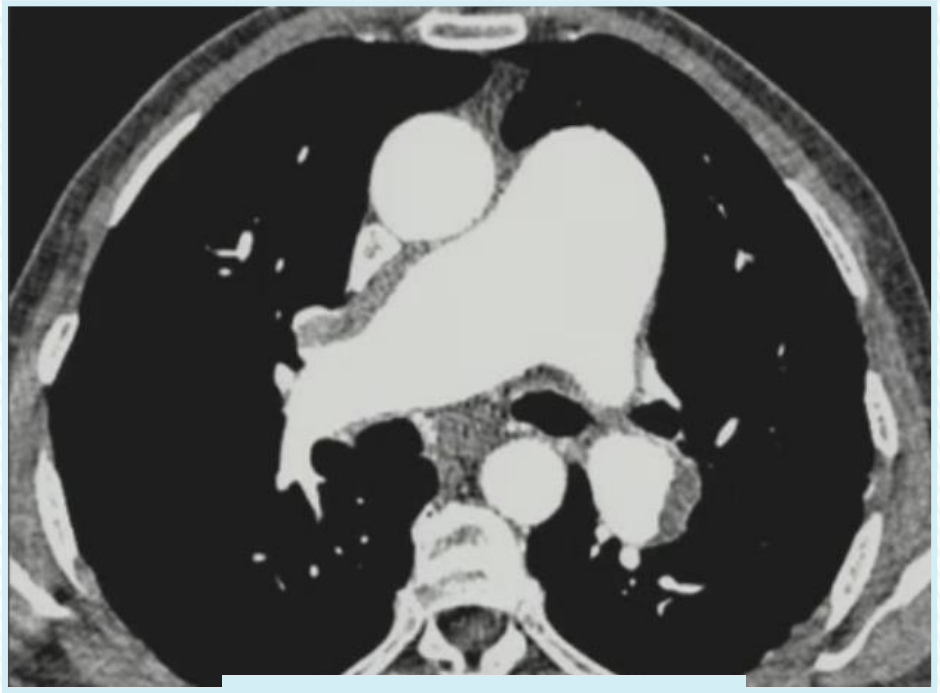
Takayasu

Mimics of CTEPH

Eisenmenger's syndrome and PH due to portal hypertension can be associated with aneurysmal dilatation of the central pulmonary arteries , in situ thrombosis (long standing PH) and mural calcifications. Confusion with eccentric thrombi in CTEPH must be avoided



Eisenmenger



Portal Hypertension

Χρόνια Θρομβοεμβολική Πνευμονική Υπέρταση (CTEPH)

Tunariu N, Gibbs SJ, Win Z, et al. Ventilation-perfusion scintigraphy is more sensitive than multidetector CTPA in detecting chronic thromboembolic pulmonary disease as a treatable cause of pulmonary hypertension. J Nucl Med 2007;48:680–4

They reported a sensitivity rate of detecting chronic thromboembolic disease of just 51% with CTPA versus >96% with a VQ scan.

It is important to note that while CTPA has been proven to be noninferior to V'/Q' in diagnosing CTEPH, a negative CTPA does not exclude CTEPH, as subsegmental disease can be missed if reliance is placed only on CT.
Eur Respir Rev 2017; 26: 160108

Multidetector CT pulmonary angiography has become an established imaging modality for confirming CTEPH, however, this investigation alone cannot exclude the disease

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension European Heart Journal (2016) 37, 67–119

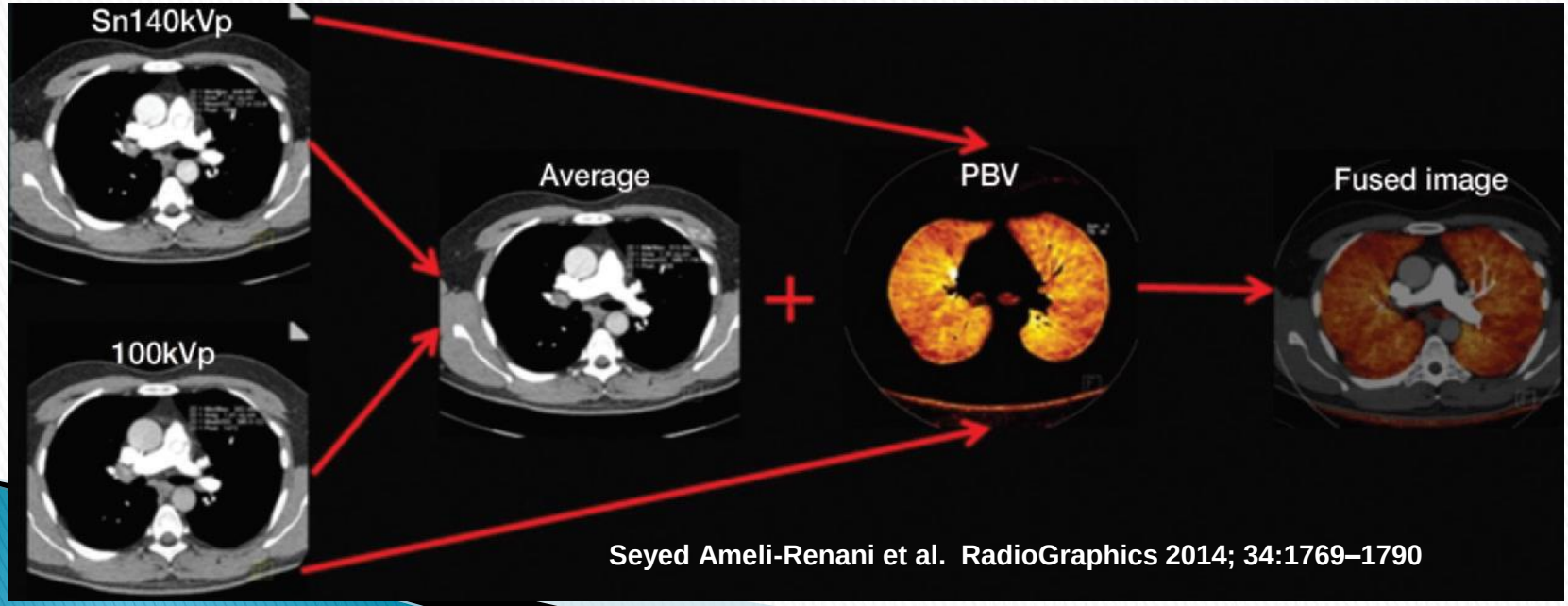
Dual Energy CT- Lung Perfusion

• Dual-energy CT pulmonary angiography allows the generation blood volume (PBV) maps that represent the relative iodine concentration in the lung parenchyma, which is a surrogate marker of perfusion

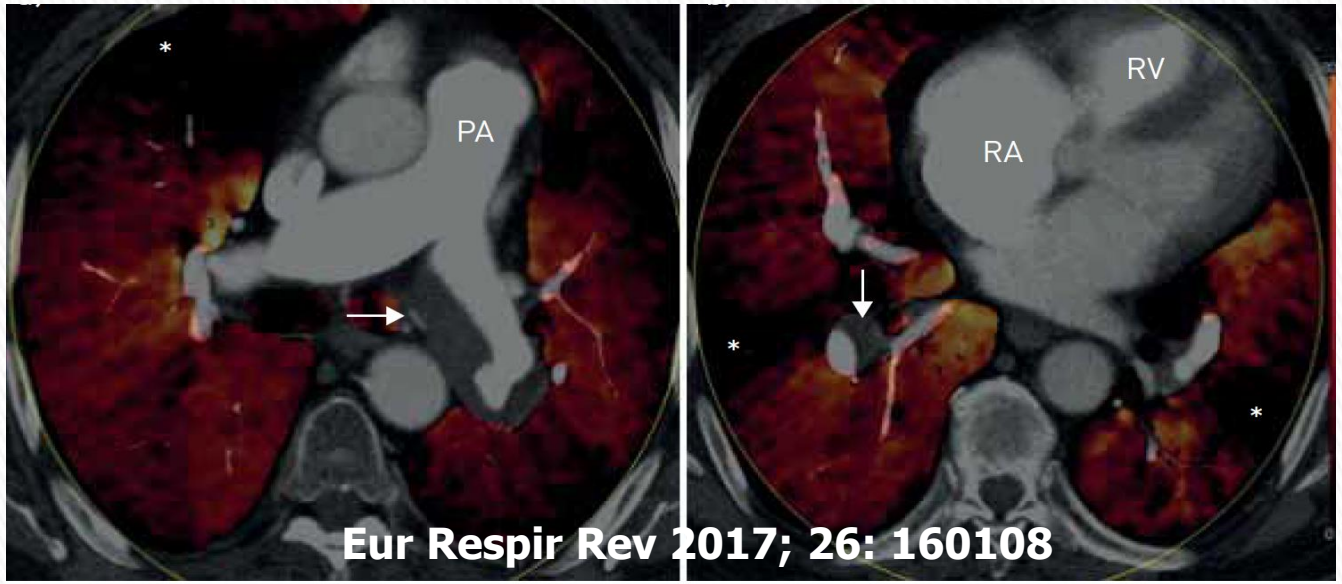
• Perfusion defects on PBV maps may also result from lung parenchymal disease so careful attention must be paid to avoid a false positive result

• Recent developments of dual-energy CT suggest improved detection of peripheral thromboembolic disease in the future.

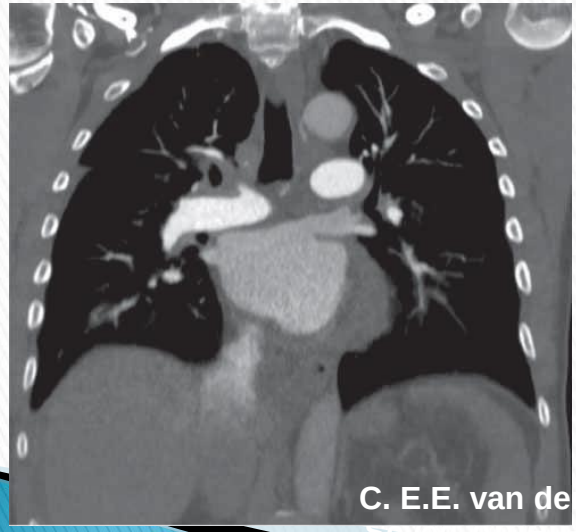
C. E.E. van der Bruggen et al. ERS Monogr 2015; 70: 60–79, 158-170

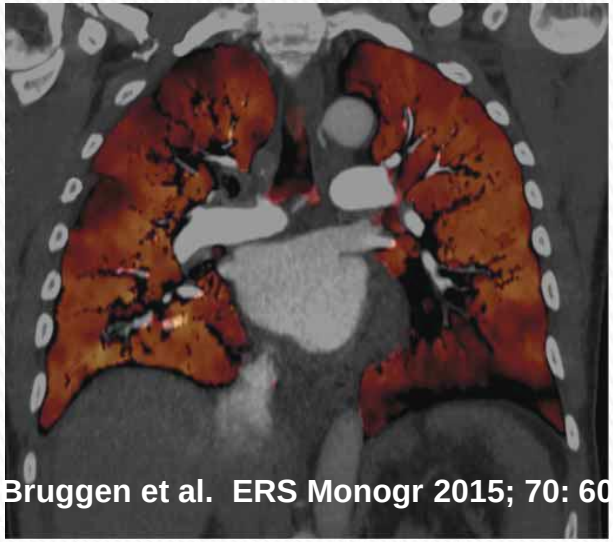


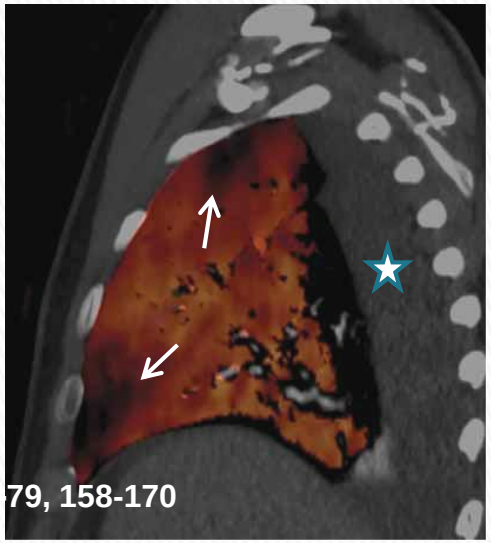
Dual Energy CT- Lung Perfusion



Eur Respir Rev 2017; 26: 160108







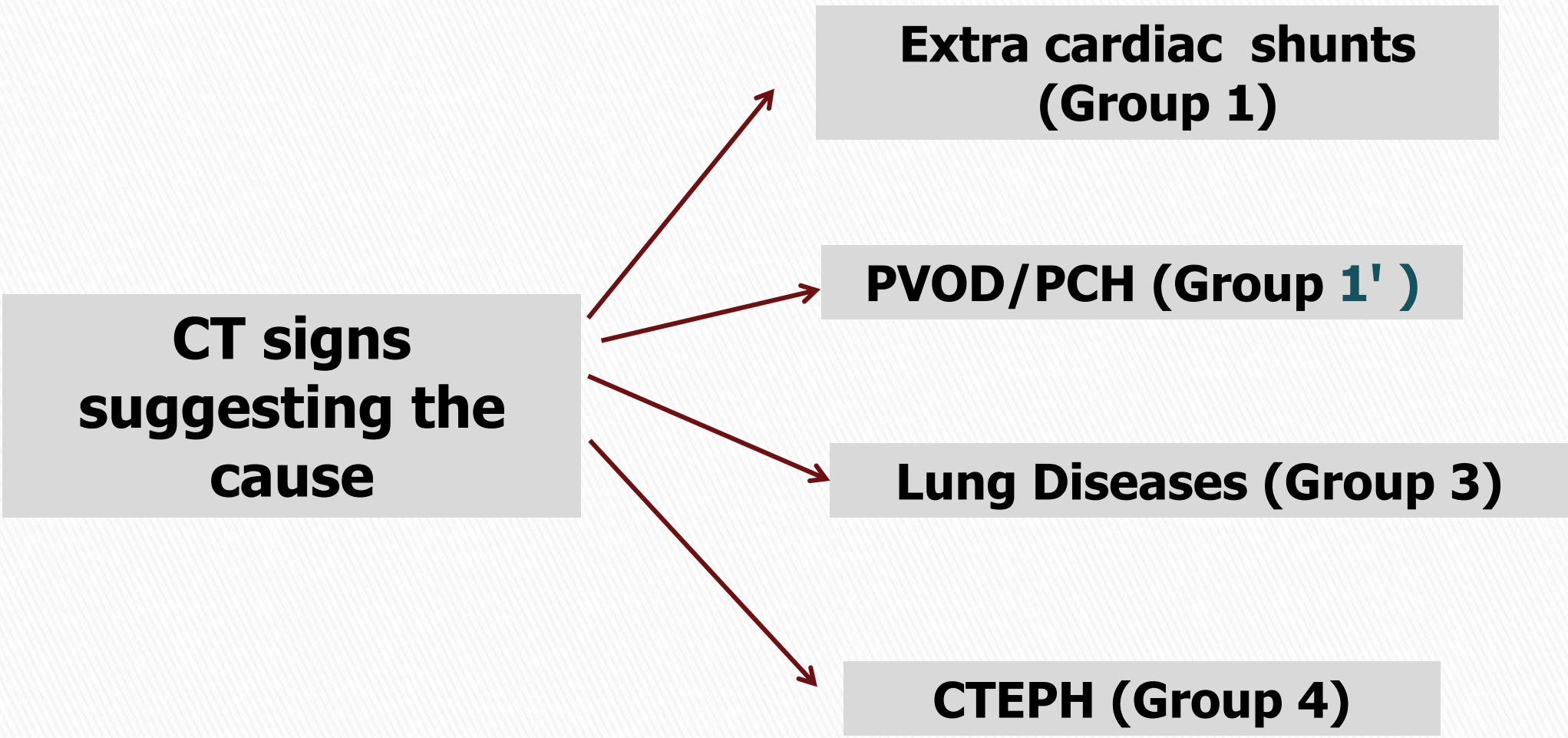
C. E.E. van de Bruggen et al. ERS Monogr 2015; 70: 60-79, 158-170

Χρόνια θρομβοεμβολική πνευμονική υπέρταση
Επιλογή ασθενών για θρομβοενδαρτηρεκτομή

Κριτήρια επιτυχούς θρομβοενδαρτηρεκτομής

- ✓ **Έκταση προσβολής- εντόπιση. Τουλάχιστον μέχρι τους κεντρικούς τμηματικούς κλάδους**
- ✓ **Διάταση βρογχικών αρτηριών**

Heinrich M et al. Chest 2005;127: 1606–1613


CT signs suggesting the cause
Extra cardiac shunts (Group 1)
PVOD/PCH (Group 1')
Lung Diseases (Group 3)
CTEPH (Group 4)

PH Diagnostic algorithm

1st step

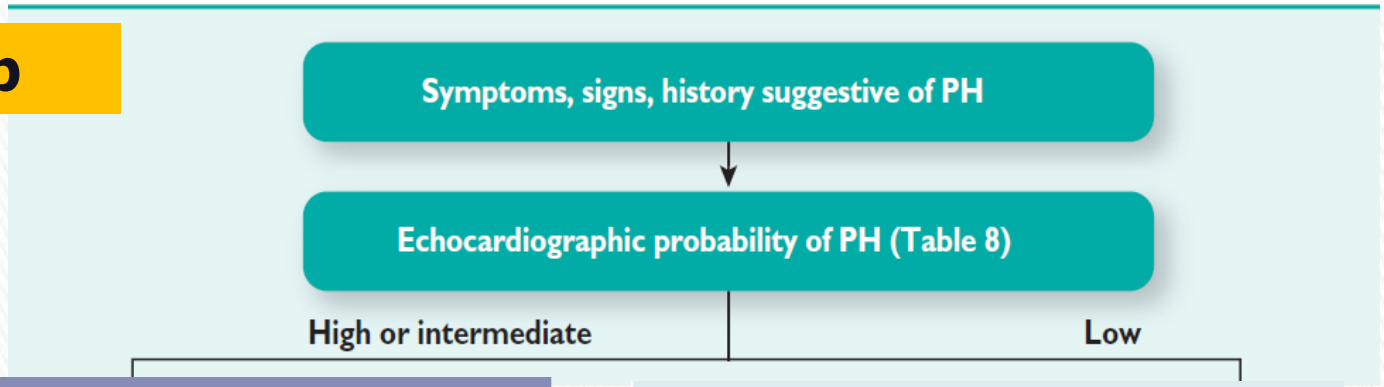


Table 8A Echocardiographic probability of pulmonary hypertension in symptomatic patients with a suspicion of pulmonary hypertension

Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'[a]	Echocardiographic probability of pulmonary hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

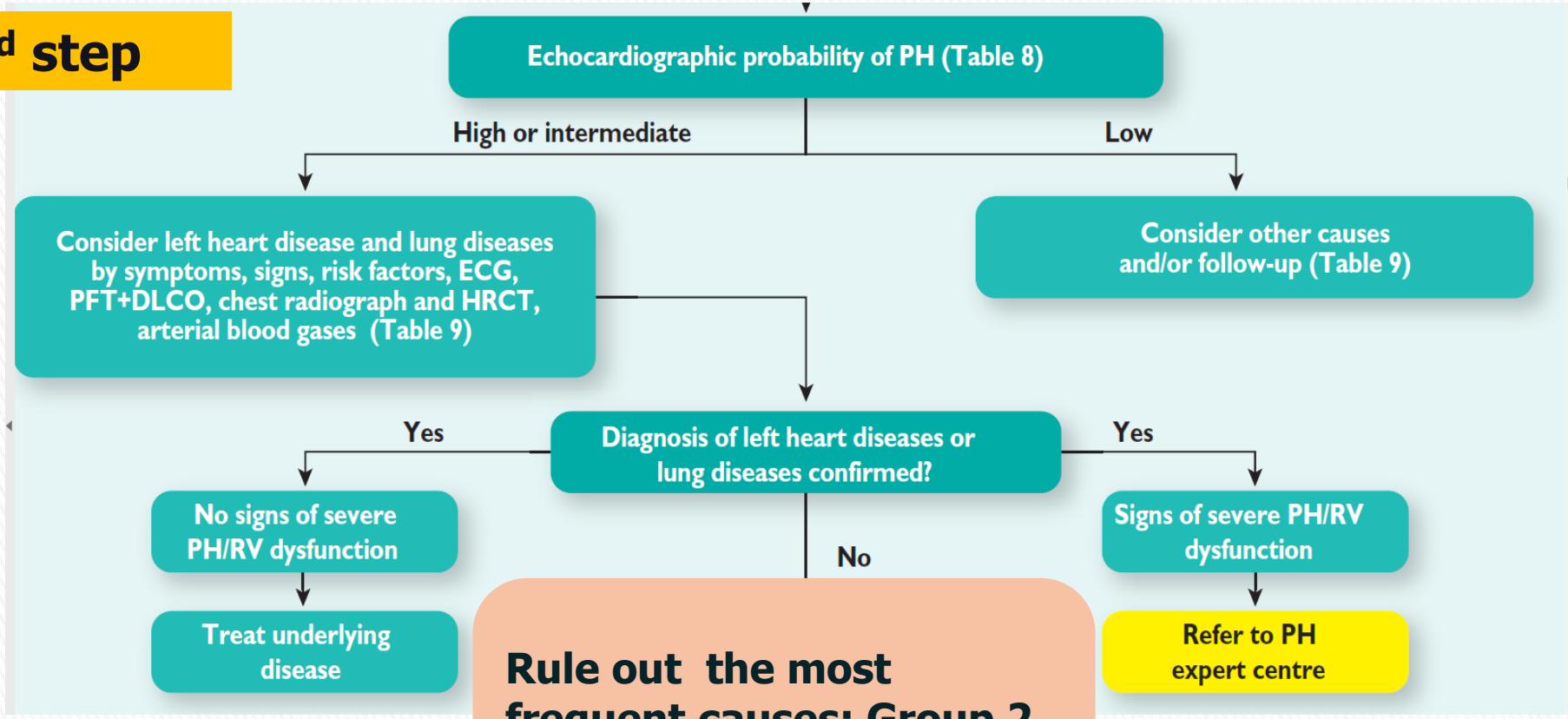
A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm²
	PA diameter >25 mm.	

Echocardiography : ALSO rule out cardiac cause, rule out intracardiac shunt, assess RV function

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

PH Diagnostic algorithm

2nd step

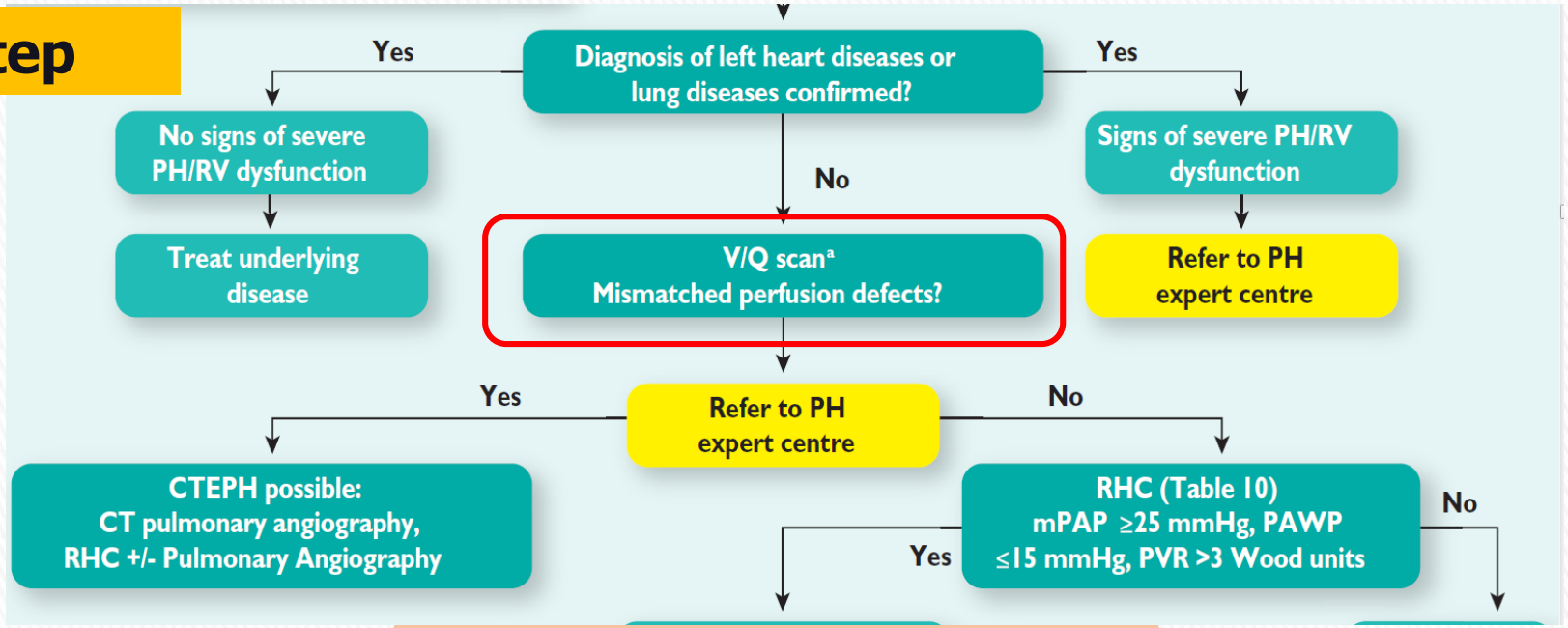


High Resolution CT (HRCT) of the pulmonary parenchyma
practically in all patients with high or intermediate probability of PH

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

PH Diagnostic algorithm

3rd step



Rule out CTEPH

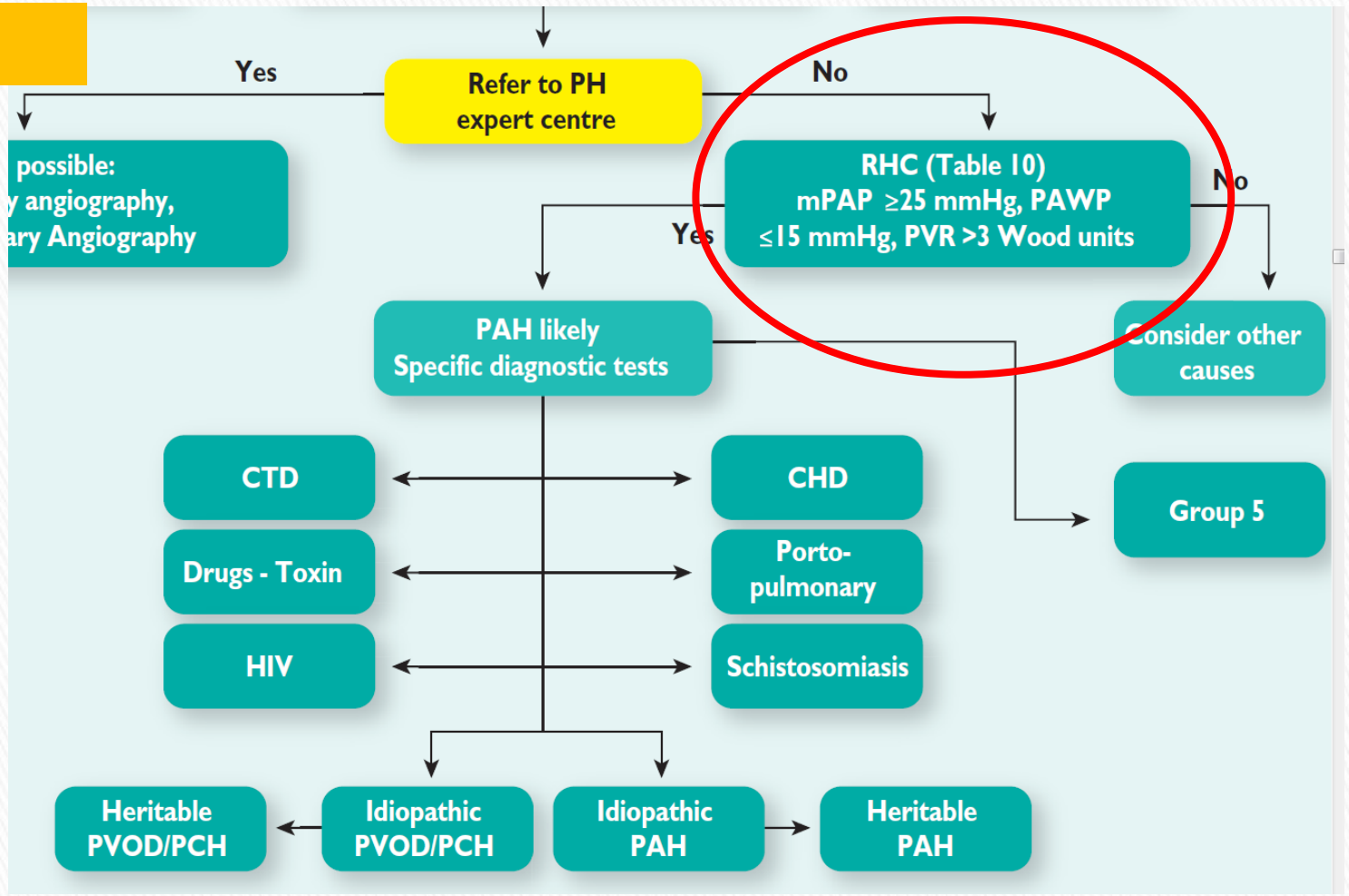
V/Q scan: high sensitivity, high NPV, lower dose than CT

But false positives (PVOD, IPAH), underestimation of vascular obstruction and no dd is possible between acute and chronic embolism

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

PH Diagnostic algorithm

4th step



2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

Take Home Messages

✓ **Κομβικός ο ρόλος της απεικόνισης: Υπόδειξη της διάγνωσης της ΠΥ, αναγνώριση του αιτίου, εκτίμηση της λειτουργικής κατάστασης της δεξιάς κοιλίας, παρακολούθηση της πορείας**

✓ **MDCT: HRCT, CTPA**

- **Διάγνωση της ΠΥ**
- **Ταυτοποίηση αιτίου (PVOD, CTEPH, shunts, Lung diseases)**
- ***Όχι πληροφορίες πιέσεων ή αγγειακών αντιστάσεων***

✓ **DECT: Perfusion imaging**

✓ **ECG Gating**

✓ **Iterative Reconstruction Algorithms: Μείωση της δόσης**

Refer to PH expert centre