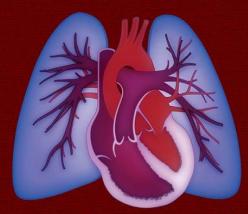


ΕΛΛΗΝΙΚΗ ΕΤΑΙΡΕΙΑ ΜΕΛΕΤΗΣ ΤΗΣ ΠΝΕΥΜΟΝΙΚΗΣ ΥΠΕΡΤΑΣΗΣ

2° ΠΑΝΕΛΛΗΝΙΟ ΣΥΝΕΔΡΙΟ ΠΝΕΥΜΟΝΙΚΗΣ ΥΠΕΡΤΑΣΗΣ



Αθήνα, 15-17 Ιουνίου 2018

Αμφιθέατρο Cotsen Hall (Γεννάδειος Βιβλιοθήκη)

"Χορηγούνται Μόρια Συνεχιζόμενης Ιατρικής Εκπαίδευσης από τον Π.Ι.Σ."



Πνευμονική Υπέρταση Απεικόνιση Ι

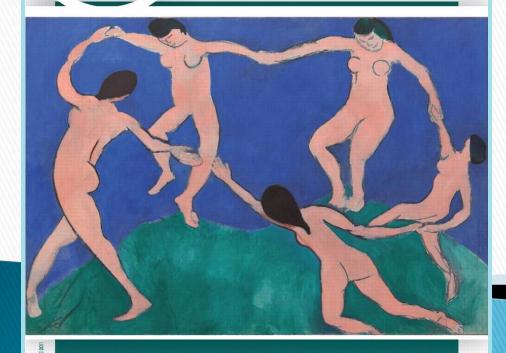
Αξονική Τομογραφία



TH



WORLD SYMPOSIUM ON PULMONARY HYPERTENSION



NICE ACROPOLIS, Nice

February 27-28 / March 1, 2013









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)

Authors/Task Force Members: Nazzareno Galiè* (ESC Chairperson) (Italy), Marc Humbert**a (ERS Chairperson) (France), Jean-Luc Vachieryc (Belgium), Simon Gibbs (UK), Irene Lang (Austria), Adam Torbicki (Poland), Gérald Simonneaua (France), Andrew Peacocka (UK), Anton Vonk Noordegraafa (The Netherlands), Maurice Beghettib (Switzerland), Ardeschir Ghofrania (Germany), Miguel Angel Gomez Sanchez (Spain), Georg Hansmannb (Germany), Walter Klepetkoc (Austria), Patrizio Lancellotti (Belgium), Marco Matuccid (Italy), Theresa McDonagh (UK), Luc A. Pierard (Belgium), Pedro T. Trindade (Switzerland), Maurizio Zompatorie (Italy) and Marius Hoepera (Germany)

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Marc Humbert, Service de Pheumologie, Hópital Bicétre, Université Paris-Sud, Assistance Publique Hópitaux de Paris, 78 rue du Général Leclerc, 94270 Le Kremlin-Bicetre, France, Tel: +33 145217972, Fax: +33 145217971, Emait marc.humbert@apho.fr

ESC Committee for Practice Guidelines (CPG) and National Cardiac Societies document reviewers: listed in Appendix

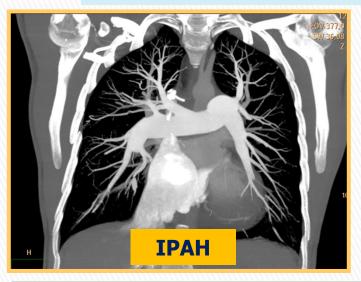
^{*}Representing the European Respiratory Society; *Representing the Association for European Paediatric and Congenital Cardiology; *Representing the International Society for Heart and Lung Transplantation; *Representing the European League Against Rheumatism; and *Representing the European Society of Radiology.

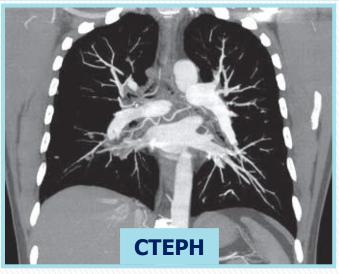
ESC entities having participated in the development of this document:

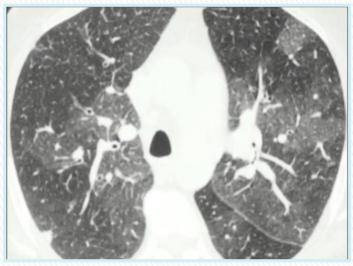
ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutareous Cardiovascular Interventions (EAPCI), European Passociation (EIPRA), Heart Failure Association (HFA), Pest Failure A

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

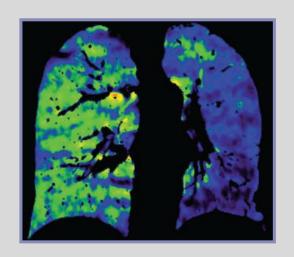


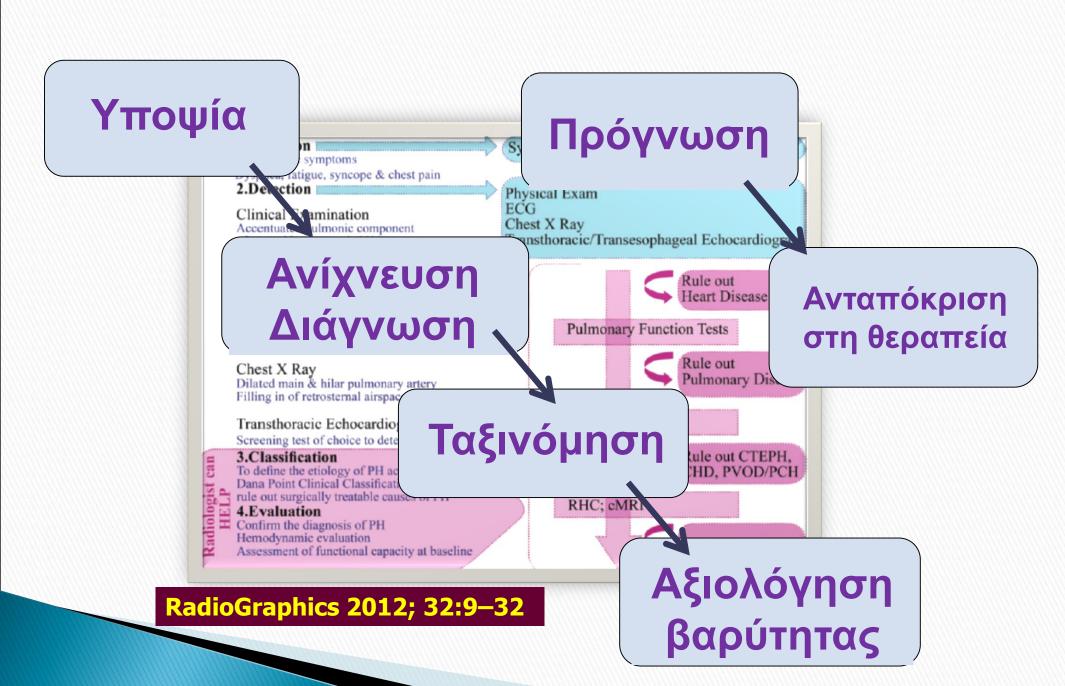
No blood flow or pressure data

Iterative Reconstruction **Algorithms**

Dose reduction up to 50%

Dual Energy CTPA -Pulmonary Perfusion





Role of CT in Pulmonary Hypertension

- ✓ Identify imaging features suggesting PH
 In suspected PH
 In routine practice
- **✓ Identify cause of PH**

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

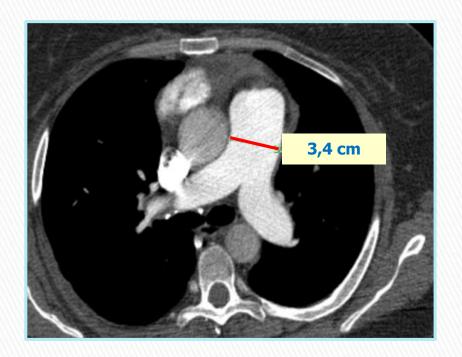
1. Vascular Signs: PA dilatation

- 1. Diameter of main pulmonary artery ≥ 29 mm : Sens 79%, spec 83%
- 2. Diameter of main pulmonary artery ≥ 28 mm : Sens 88%, spec 78%

mPAD: > 29 mm in men, > 27 mm in women

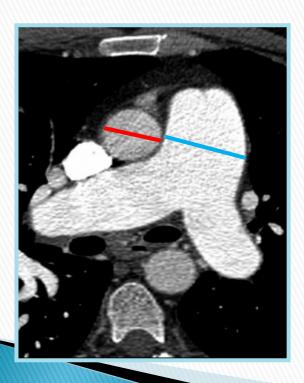
Truong QA, Massaro JM, Rogers IS, et al. Reference values for normal pulmonary artery dimensions by noncontrast cardiac computed tomography: the Framingham Heart Study.

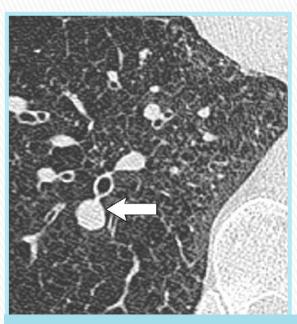
Circ Cardiovasc Imaging 2012;5(1):147–154.



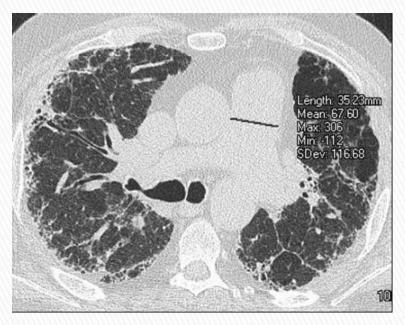
1. Vascular Signs: PA dilatation

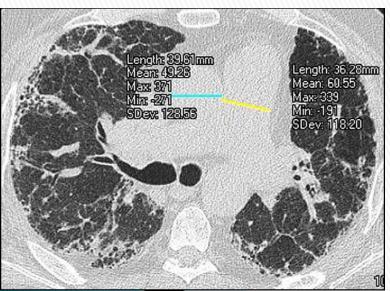
- 1. Main PA diameter/ ascending aorta diameter ratio ≥ 1: sens 92%, sp 72% Boerrigter 2010 Ιδιαίτερα σε άτομα < 50 ετών
- 1. Segmental artery-to-bronchus diameter ratio ≥ 1 in at least three lobes: Sens 87%, spec 68% Devaraj Radiology 2010





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

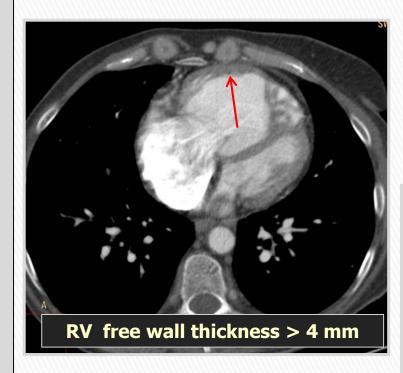


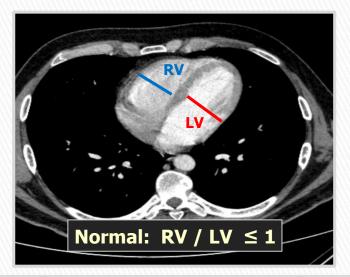
Decrease in right pulmonary artery distensibility

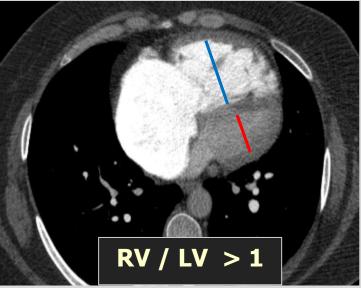
The best pulmonary artery distensibility cutoff for distinguishing between patients with pulmonary hypertension and those without it was 16.5% when a standard method of measurement was used and 13% when a simplified method was used

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







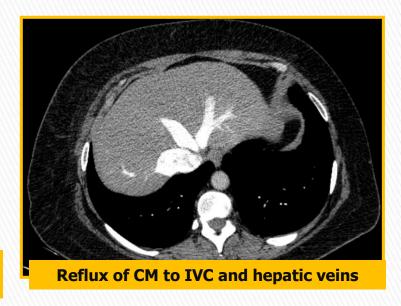
straightening of the IVS, D-shaped LV

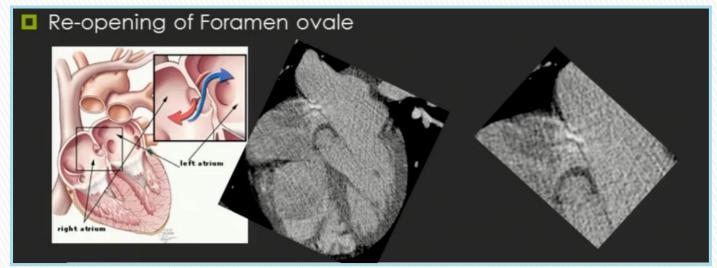
... and

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



Pericarsial effusion POOR prognostic sign





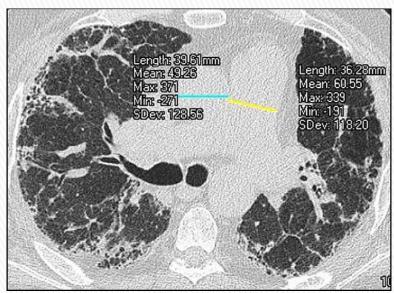
Marie-Pierre REVEL ECR 2015

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

GROUP 1': PVOD/PCH

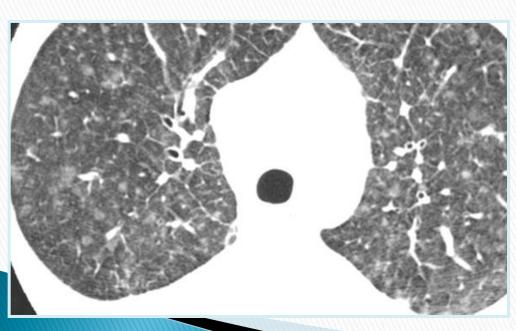
High-resolution CT of the chest is the investigation of choice.

V/Q lung scanning is not useful in discriminating PVOD/PCH from IPAH

Specific CT signs

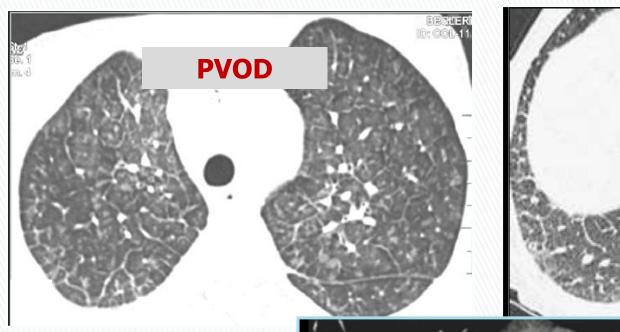
- 1. Smoothly thickened interlobular septa
- 2. Poorly defined centrilobular ground glass opacities
- 3. Lymphadenopathy

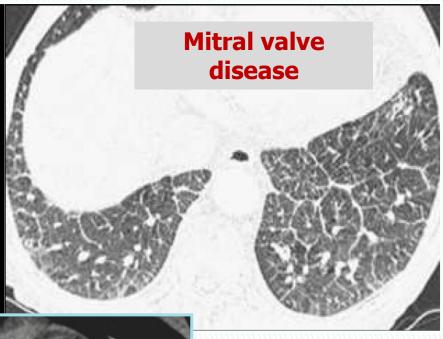
Sens 66% Spec 100% Resten et al. AJR 2004;183:65-70





GROUP 1': PVOD/PCH





In PVOD

... and normal PAWP (< 15 mmHg)

... small central pulmonary veins

GROUP 1': PVOD/PCH

Rare, difficult to diagnose

But, no room for error

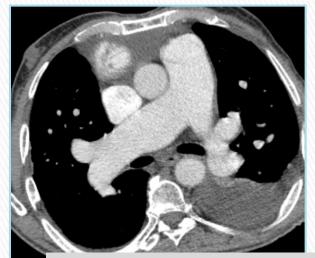
Vasodilators and, in particular, i.v. epoprostenol must be used with great caution because of the high risk of severe drug-induced pulmonary oedema

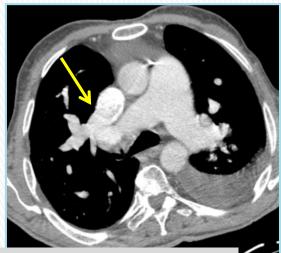




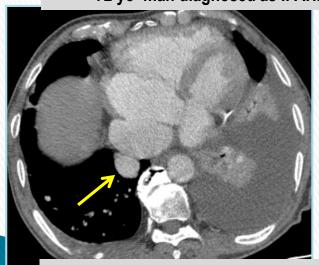
David Montani et al. Medicine 2008;87:220–233

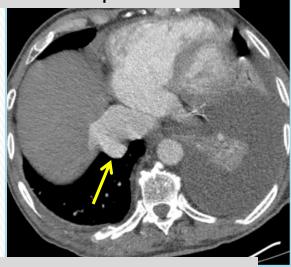
GROUP 1: PAH associated with Congenital Heart Disease (CHD)





72 yo man diagnosed as IPAH. Chest CT for pneumonia



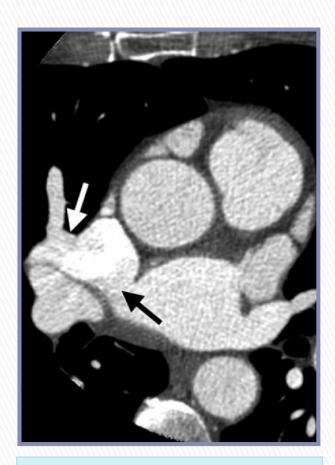


Partial Anomalous Pulmonary Venous Return

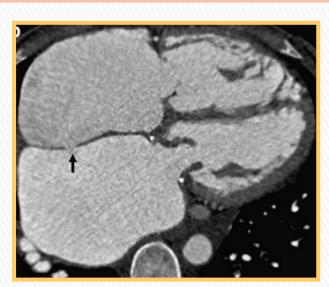
Some malformations, such as patent ductus arteriosus, sinus venosus atrial septal defect or partial anomalous pulmonary venous return, are often concealed and patients are misclassified as suffering from IPAH. Hence, these congenital anomalies should be specifically sought

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

Άλλα ευρήματα που δεν πρέπει να διαφύγουν



Γυναίκα 26 ετών Sinus Venosus





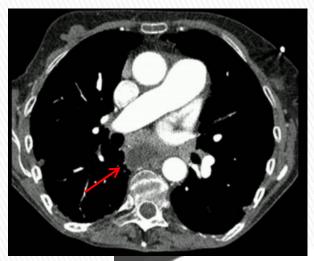




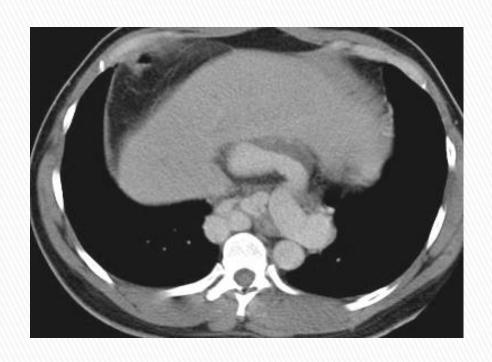
Γυναίκα 44 ετών Patent Ductus Arteriosus

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

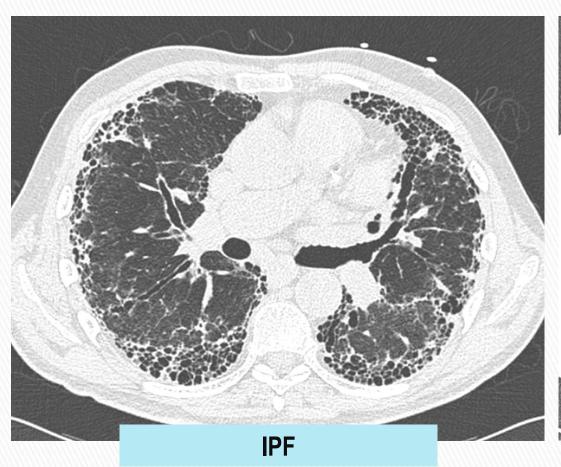
GROUP 1: Associated with portal hypertension

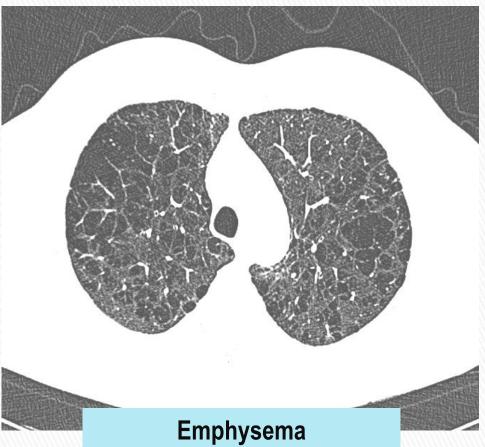






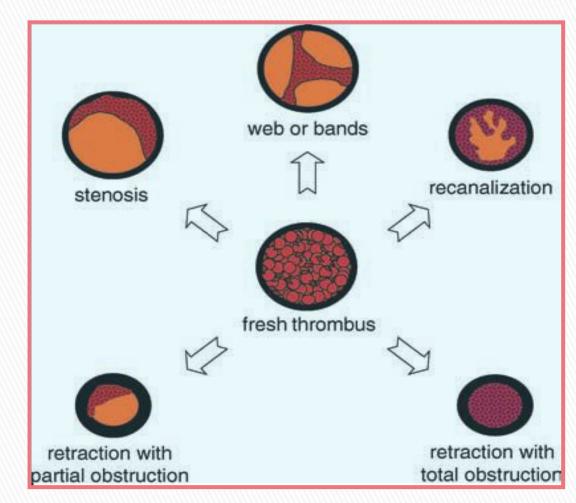
GROUP 3: PH due to lung diseases





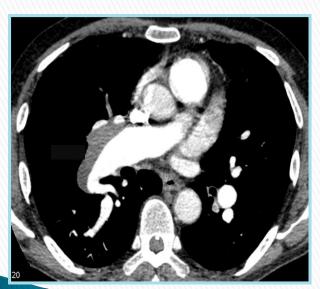
GROUP 4: Chronic Thrombo Embolic Pulmonary Hypertension (CTEPH)

- ✓ 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.

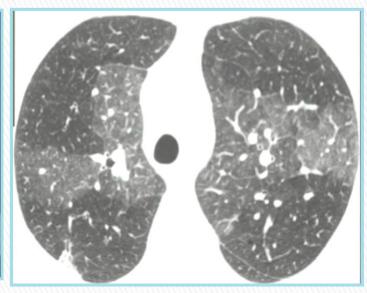


Specific CT signs

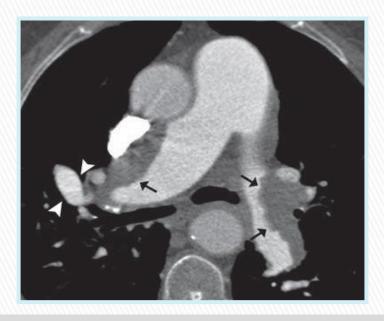
- 1. Signs of chronic PE
- 2. Enlarged systemic arteries
- 3. Mosaic perfusion



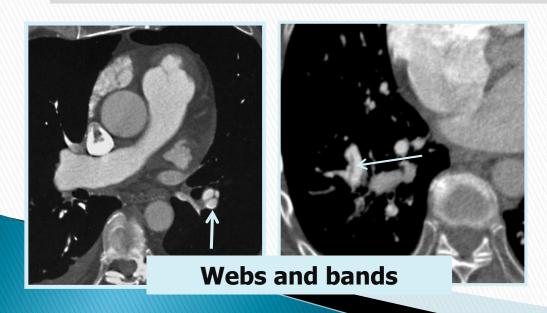


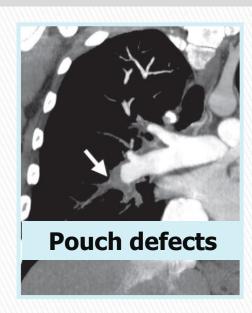






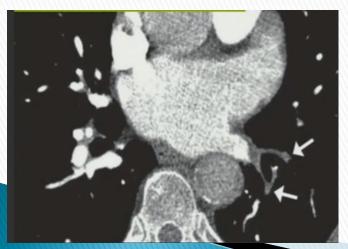
Eccentric thrombi Obtuse angle between thrombus and vessel wall



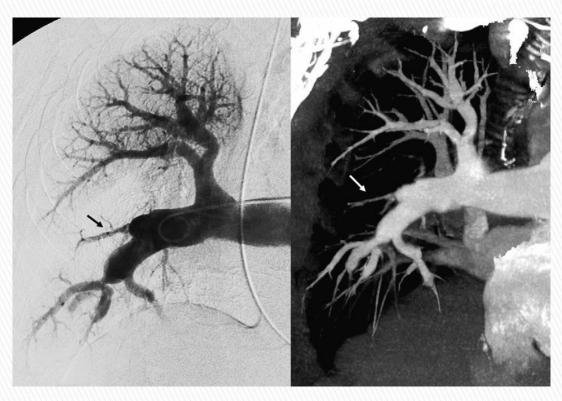


Small size occluded vessels





Post-stenotic dilatation, abrupt narrowing

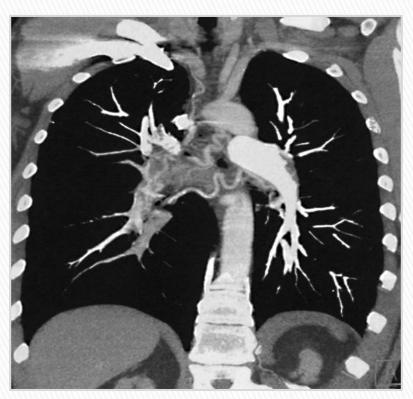


European Journal of Radiology 71 (2009) 49–54

Vascular Remodelling in CTEPH

- Laminated thrombus
- Webs / Bands
- Post-stenotic dilatation
- Pouch
- Occlusion
- Abrupt narrowing
- Intimal irregularity



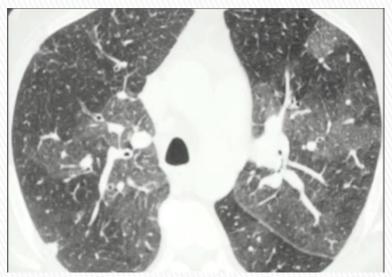


Enlarged bronchial arteries





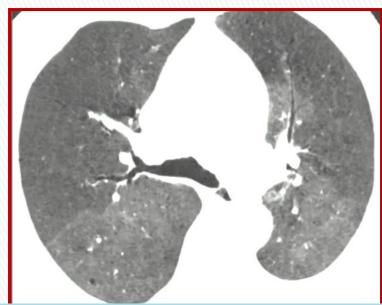
Systemic collaterals



Mosaic Perfusion



Mosaic perfusion can be subtle



Minimum Intensity Projection (MinIP)

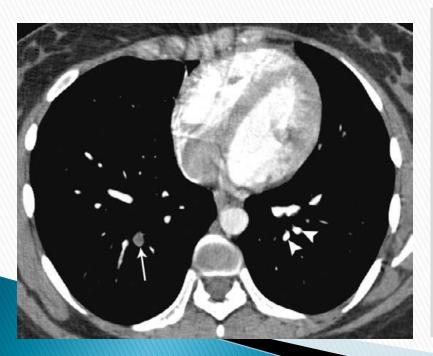
Οξεία vs Χρόνια

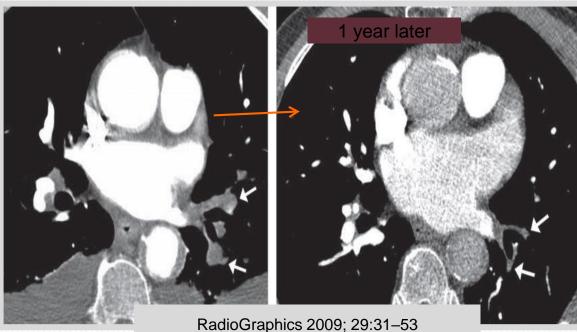
ΟΞΕΙΑ ΠΕ

ΧΡΟΝΙΑ ΠΕ

Αύξηση της διαμέτρου των αγγείων

Ελάττωση της διαμέτρου των αγγείων





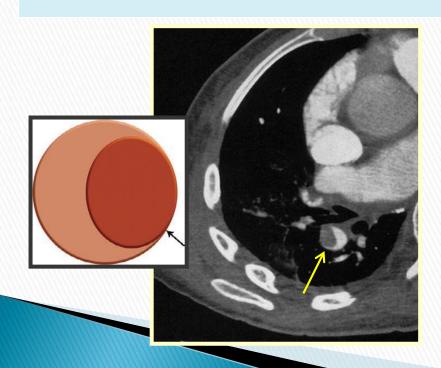
Οξεία vs Χρόνια

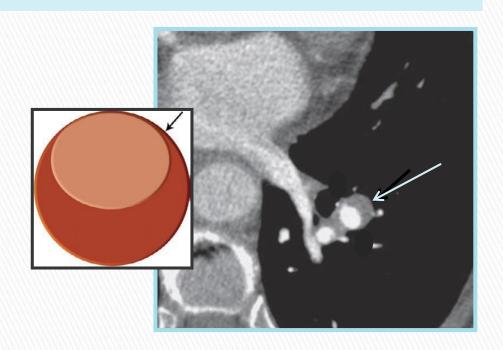
ΟΞΕΙΑ ΠΕ

ΧΡΟΝΙΑ ΠΕ

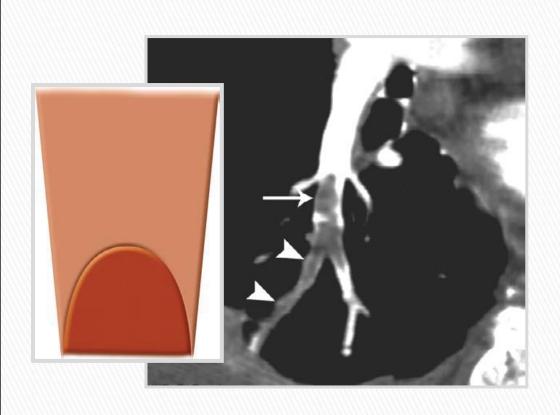
Οξεία γωνία του θρόμβου με το τοίχωμα του αγγείου

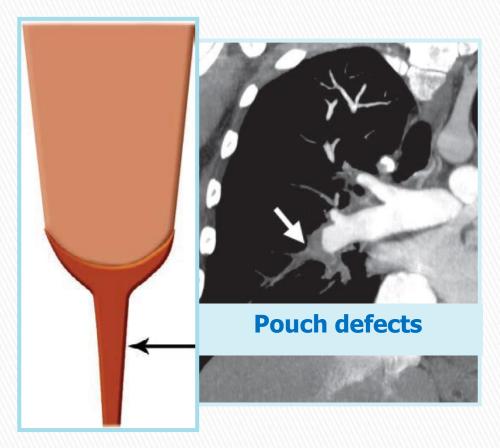
Αμβλεία γωνία του θρόμβου με το τοίχωμα του αγγείου





Οξεία vs Χρόνια



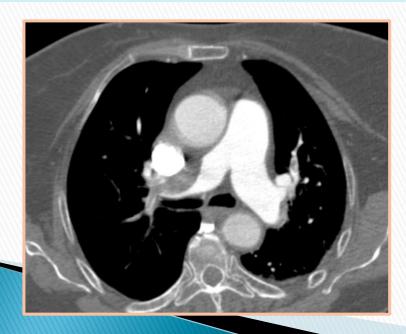


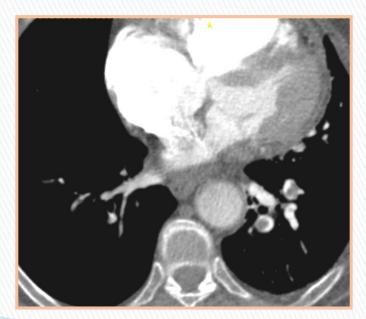
Οξεία vs Χρόνια

ΟΞΕΙΑ ΠΕ	XPONIA 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

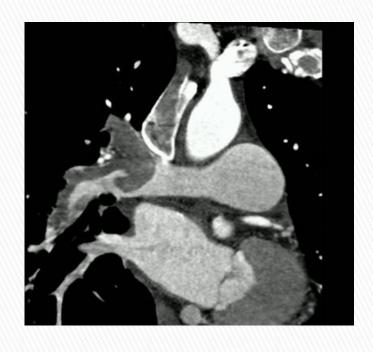
Οξεία vs Χρόνια

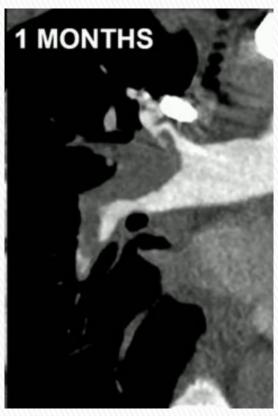
- Δεν είναι σπάνιο να συνυπάρχουν σημεία οξείας και χρόνιας εμβολής (Υποτροπιάζουσα πνευμονική εμβολή)
- Η χρόνια πνευμονική εμβολή πολλές φορές αναδεικνύεται σε CTPA που γίνονται για τον αποκλεισμό οξείας πνευμονικής εμβολής
- Η παρουσία διατεταμένων βρογχικών αρτηριών ή υπερτροφίας της δεξιάς κοιλίας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

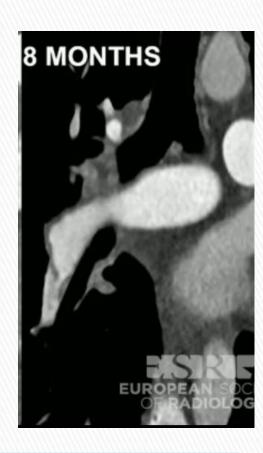




Οξεία vs Χρόνια







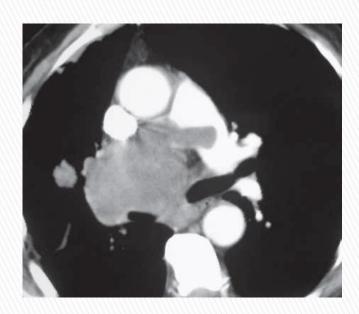
Acute PE



Chronic PE

N. Screaton ECR 2018

Mimics of CTEPH



Pulm. Artery sarcoma



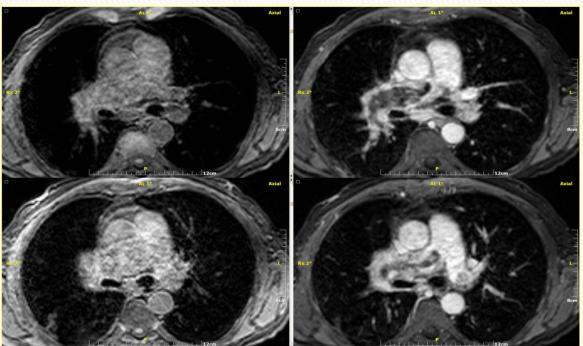
Fibrosing mediastinitis

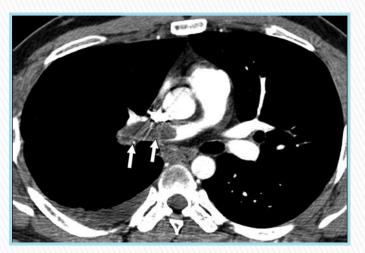


Takayasu

Pulm. Artery sarcoma



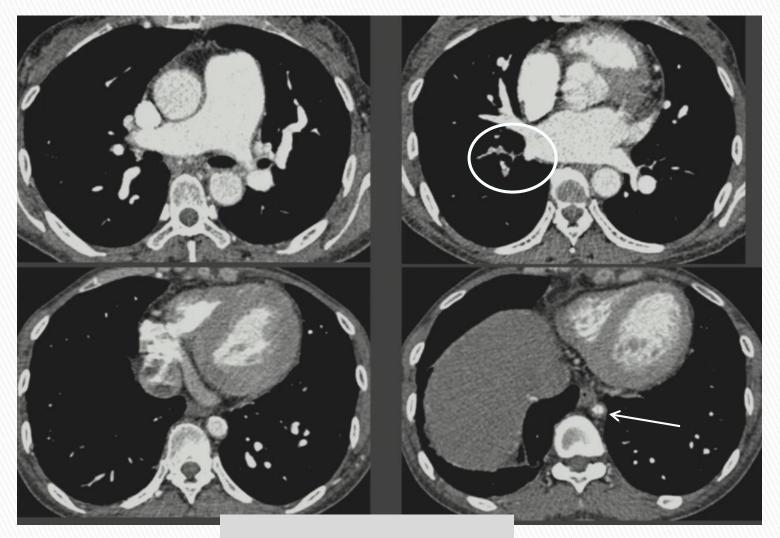






AJR 2007; 188:1691-1693

40 yo woman, severe PH, ASD repair in childhood

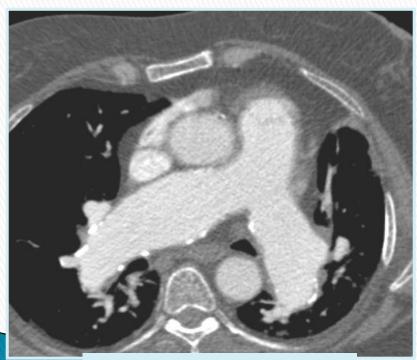


Takayasu

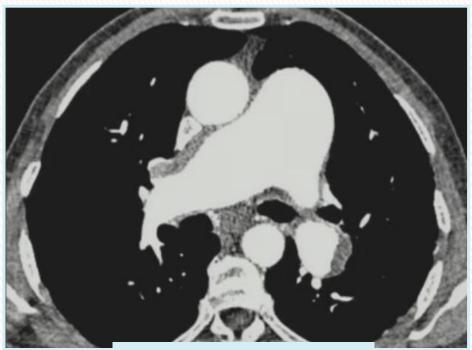
Marie- Pierre REVEL ECR 2015

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

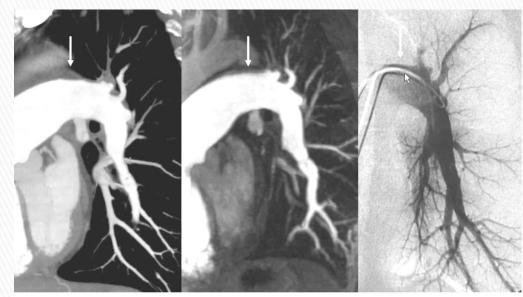


Portal Hypertension

CTPA vs MRA

- ECG-gated MD-CTPA vs. ce-MRA
 - Diagnostic work-up of CTEPH:
 - ✓ Diagnosis and differential diagnosis.
 - ✓ Technical operability.
 - ECG-gated MD-CTPA performs better than ce-MRA (and even DSA?).
 - ➤ Aim: a negative MD-CTPA scan precludes a surgically assessible form of CTEPH.

Rajaram S et al, Eur Radiol 2012; 22: 310-317 Ley S et al, Eur Radiol 2012; 22: 607-616 Sugiura T et al, Chest 2013; 143: 1074-1077 Sugiyama M et al. Jpn J Radiol 2014; 32: 375-382



ECG-gated MD-CTPA

ce-MRA

DSA

Karl-Friedrich Kreitner ESCR 2017

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.

<u>Eur Respir Rev 2017; 26: 160108</u>

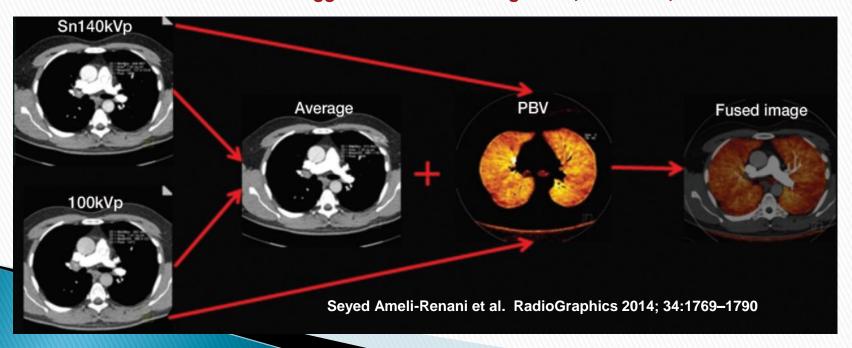
Multidetector CT pulmonary angiography has become an established imaging modality for confirming CTEPH, however, this investigation alone cannot exclude the disease

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

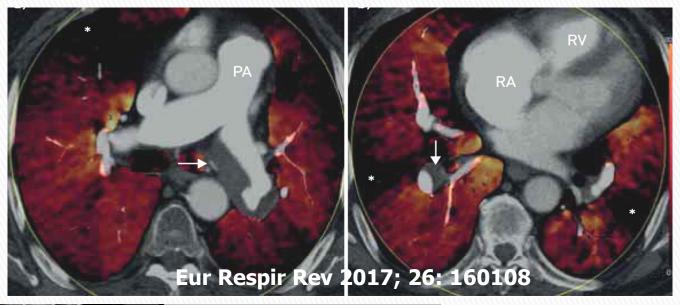
Dual Energy CT- Lung Perfusion

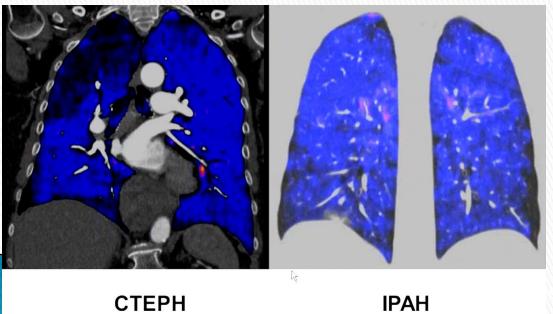
- Dual-energy CT pulmonary angiography allows the generation blood volume (PBV) maps that represent the relative iodine concentration in the lung parenchyma, which is a surrogate marker of perfusion
- Perfusion defects on PBV maps may also result from lung parenchymal disease so careful attention must be paid to avoid a false positive result
- Recent developments of dual-energy CT suggest improved detection of peripheral thromboembolic disease in the future.

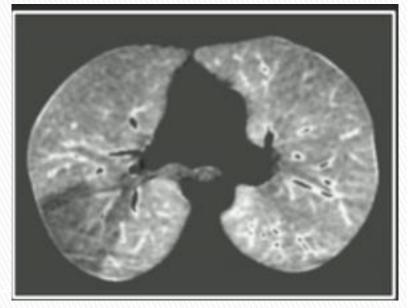
C. E.E. van der Bruggen et al. ERS Monogr 2015; 70: 60-79, 158-170



Dual Energy CT- Lung Perfusion







Χρόνια θρομβοεμβολική πνευμονική υπέρταση Επιλογή ασθενών για θρομβοενδαρτηρεκτομή

Κριτήρια επιτυχούς θρομβοενδαρτηρεκτομής

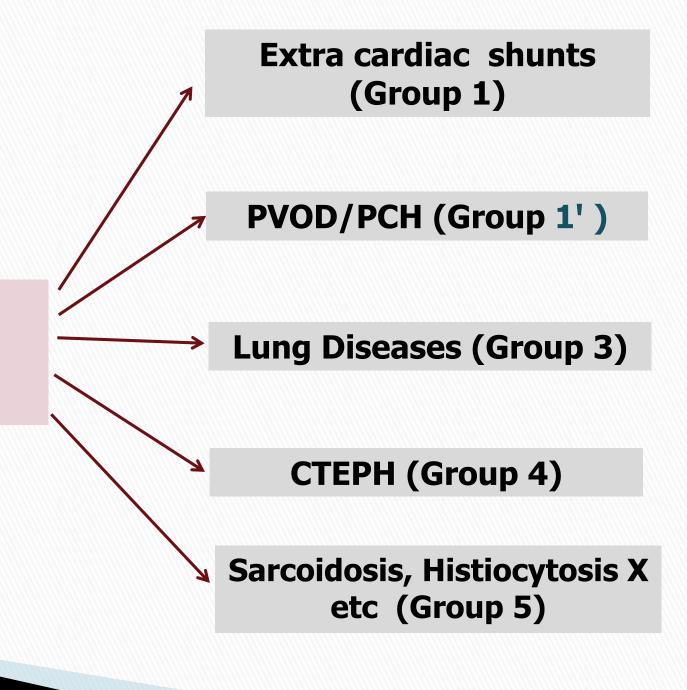
✓ Έκταση προσβολής- εντόπιση. Τουλάχιστον μέχρι τους κεντρικούς τμηματικούς κλάδους
 ✓ Διάταση βρογχικών αρτηριών

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

Αξονική Τομογραφία

- 🕯 Ευρέως διαθέσιμη, όχι ακριβή
- 🕯 Ταχεία, μη επεμβατική
- 🕯 Εξαιρετική διακριτική ικανότητα
- Εκτεταμένη, λεπτομερής αξιολόγηση πνευμονικού παρεγχύματος, αγγειακών δομών, καρδιάς
- **Δυνατότητα σχετικά χαμηλής δόσης (< 5 msV)**

🦃 Όχι πληροφορίες ροών, πιέσεων



CT signs suggesting the cause

Value of MD-CT and MRT in PH imaging

	ECG-gated MD-CT	MR-Imaging
Presence of PH	+++	+ + (+)
Assessment of lung parenchyma	+++	++
Assessment of macro- and microcirculation	+++	+ +(+)
Right heart impairment Pulmonary hemodynamics	+ +/ +	+++
Monitoring of therapy	+	+++
Prognostic significance	-	+++



Symptoms, signs, history suggestive of PH

Echocardiographic probability of PH (Table 8)

High or intermediate

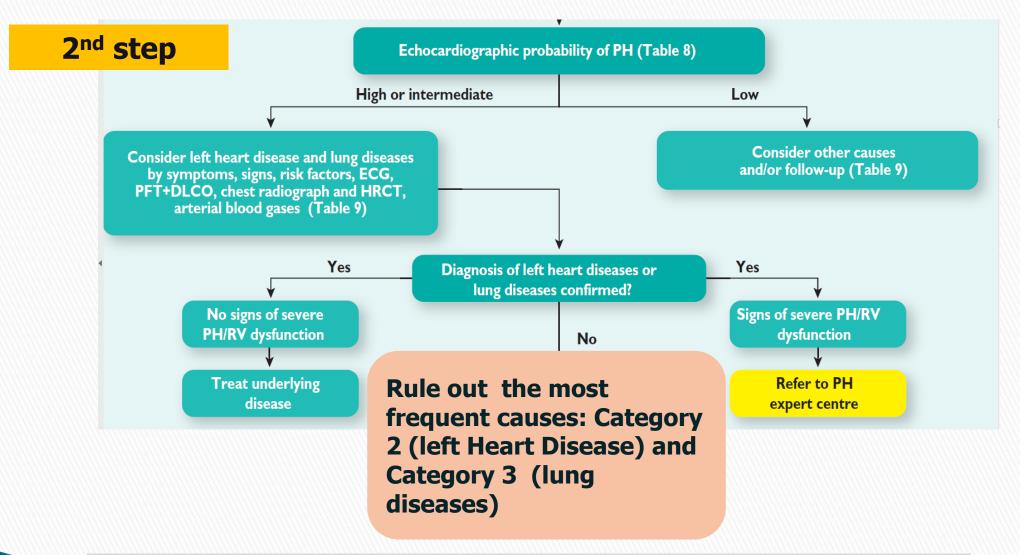
Low

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

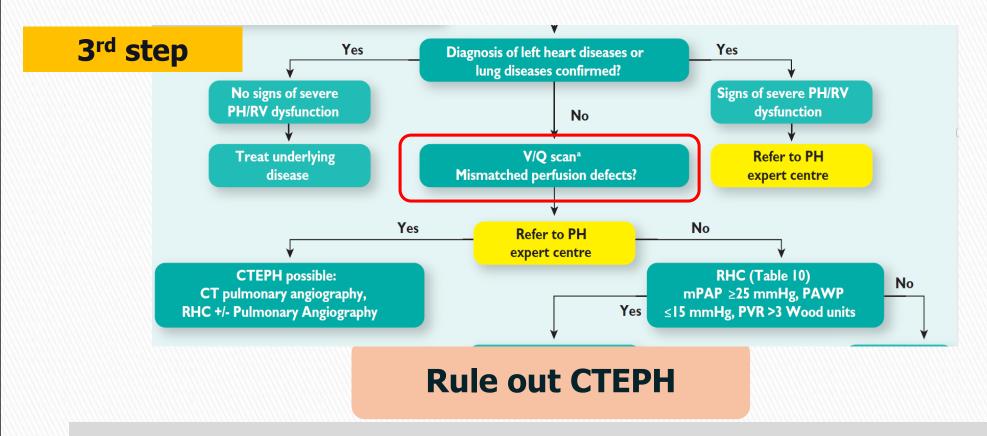
Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'a	Echocardiographic probability of pulmonary hypertension	
≤2.8 or not measurable	No	Low	
≤2.8 or not measurable	Yes	Intermediate	
2.9–3.4	No		
2.9–3.4	Yes	High	
>3.4	Not required		

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 cm ²
	PA diameter >25 mm.	

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



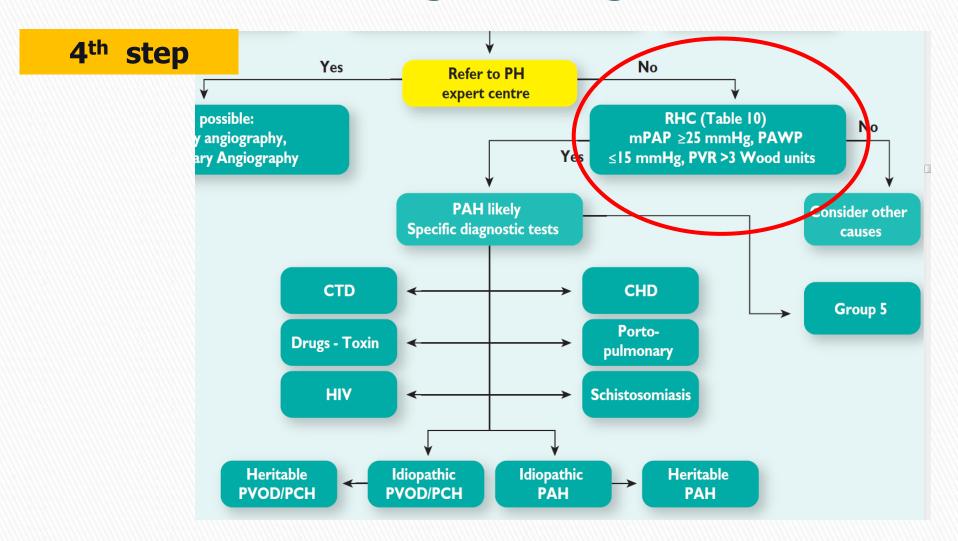
High Resolution CT (HRCT) of the pulmonary parenchyma practically in all patients with high or intermediate probability of PH



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



Take Home Messages

- √ Κομβικός ο ρόλος της μη επεμβατικής απεικόνισης
- ✓ Απαραίτητη η γνώση των σημείων που θέτουν υπόνοια πνευμονικής υπέρτασης
- √ CTPA, ECG-gated MDCTPA: Στην εκτίμηση της CTEPH
- **✓ DECT: Lung Perfusion**
- ✓ PVOD/PCH: HRCT → Υποψία της νόσου
- **✓** Make sure there is not extracardiac shunt
- ✓ CT: Η καλύτερη μέθοδος στην αναζήτηση της αιτίας
 - √ Όχι πληροφορίες ροών, πιέσεων