

Con il patrocinio di



Associazione Italiana Pneumologi Ospedalieri



# PNEUMOLOGIA 2016

Milano, 16 – 18 giugno 2016 · Centro Congressi Palazzo delle Stelline

**PNEUMOLOGIA 2016**

***NOVITA' IN PNEUMOLOGIA  
L'INDISPENSABILE DA SAPERE***

***La diagnosi di ipertensione  
polmonare post-embolica***

*Dr. Marco Biolo*

*S.C. Pneumologia*

*Azienda Sanitaria Ospedaliero Universitaria Integrata di Trieste*

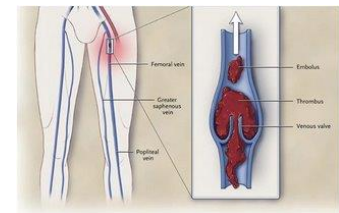
*Direttore Dr. Marco Confalonieri*

# PULMONARY HYPERTENSION CLASSIFICATION

Group 1	Pulmonary arterial hypertension
Group 2	PH owing to left heart disease
Group 3	PH owing to lung diseases and/or hypoxia
<b>Group 4</b>	<b>CTEPH</b>
Group 5	PH with unclear or multifactorial etiologies

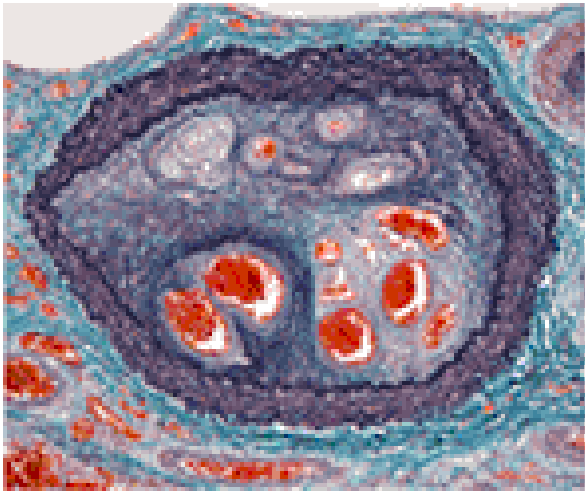
# CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): DEFINITIONS

- Si definisce come un incremento della pressione arteriosa polmonare media maggiore di 25 mmHg persistente dopo 3 mesi dalla diagnosi e di trattamento anticoagulante efficace.
- La tromboembolia cronica, una delle principali cause di grave ipertensione polmonare, si sviluppa dall'ostruzione di rami dell'arteria polmonare in seguito a episodi tromboembolici con incompleta risoluzione del trombo e formazione di fibrosi con rimodellamento della parete vascolare.
- Di conseguenza aumentano le resistenze polmonari (PVR) e la pressione nel piccolo circolo, con progressiva insufficienza cardiaca destra.

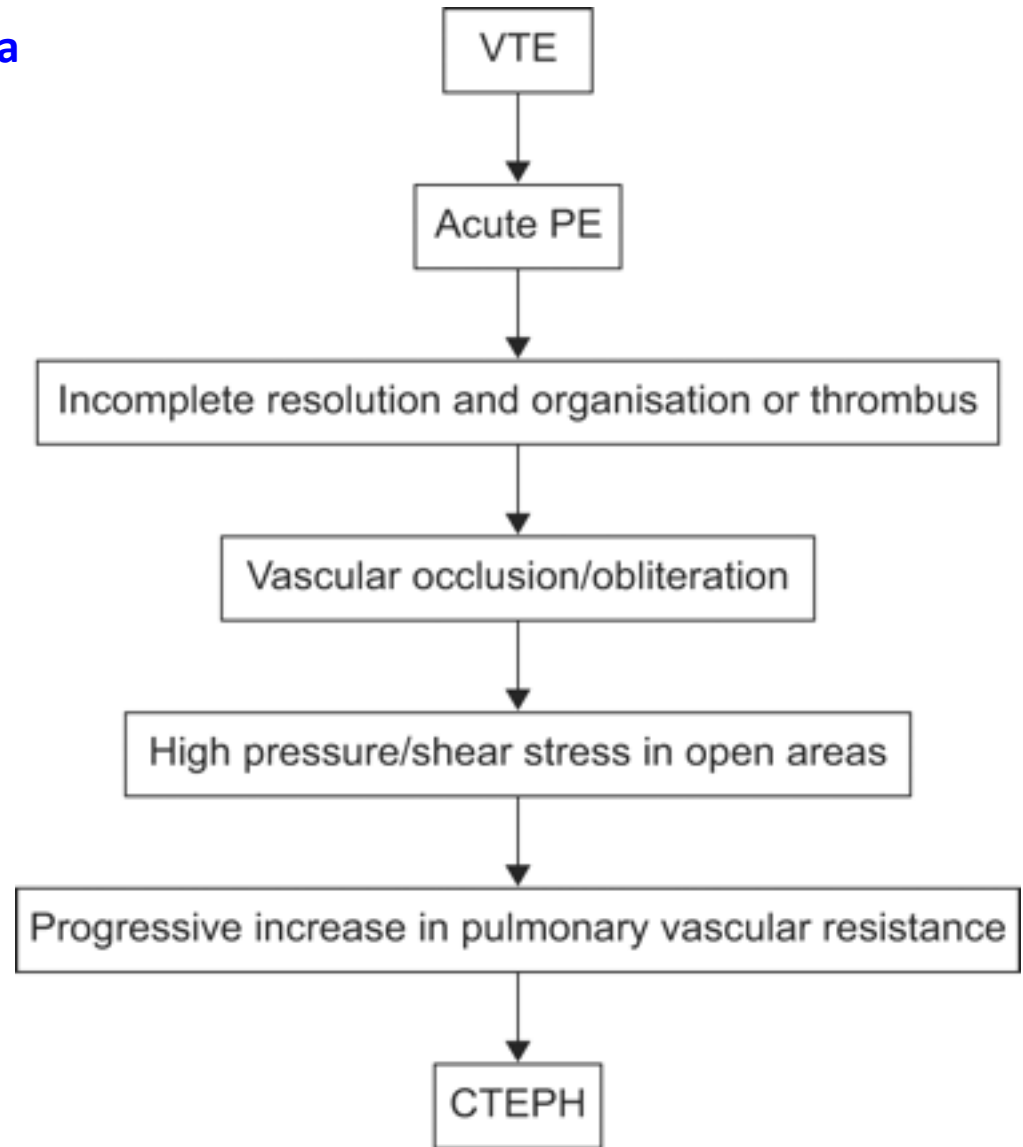


# CTEPH PATHOGENESIS

## Istopatologia della lesione trombotica



**Trombo organizzato in CTEPH.**  
L'anatomia patologica mostra un trombo organizzato con iperplasia fibrosa dell'intima, contenente caratteristici canali all'interno della arteria polmonare (lesione a colino) dovuti a ricanalizzazione del trombo fibrotico all'interno dell'arteria.



**L'ipotesi embolica della patogenesi della CTEPH**

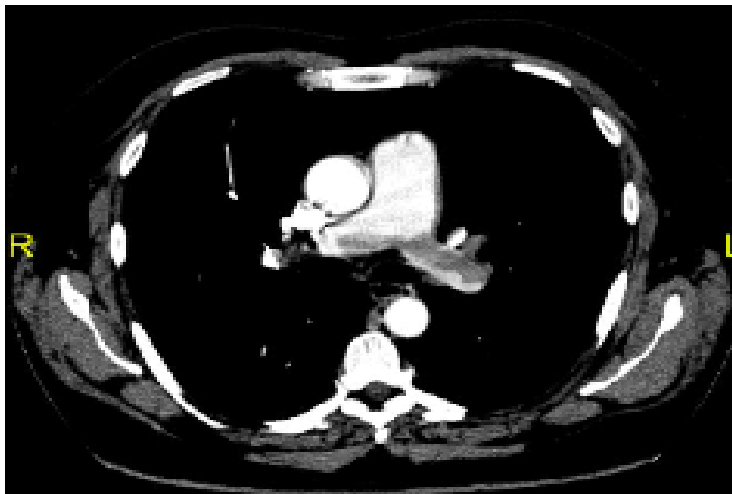
### **Vasculopatia polmonare con trombosi “in situ” delle arterie polmonari:**

- Nei pazienti con CTEPH non è documentato un significativo incremento dei fattori di rischio per embolia polmonare fatta eccezione per LAC e anticardiolipina
- Non è documentata una ridotta attività fibrinolitica
- Spesso non è identificabile in anamnesi un episodio di embolia polmonare

### Silent Pulmonary Embolism in Patients with Deep Venous Thrombosis: A Systematic Review

Paul D. Stein, MD,<sup>a</sup> Fadi Matta, MD,<sup>a</sup> Muzammil H. Musani, MD,<sup>b</sup> Benjamin Diaczok, MD<sup>b</sup>

*The American Journal of Medicine (2010) 123, 426-431*



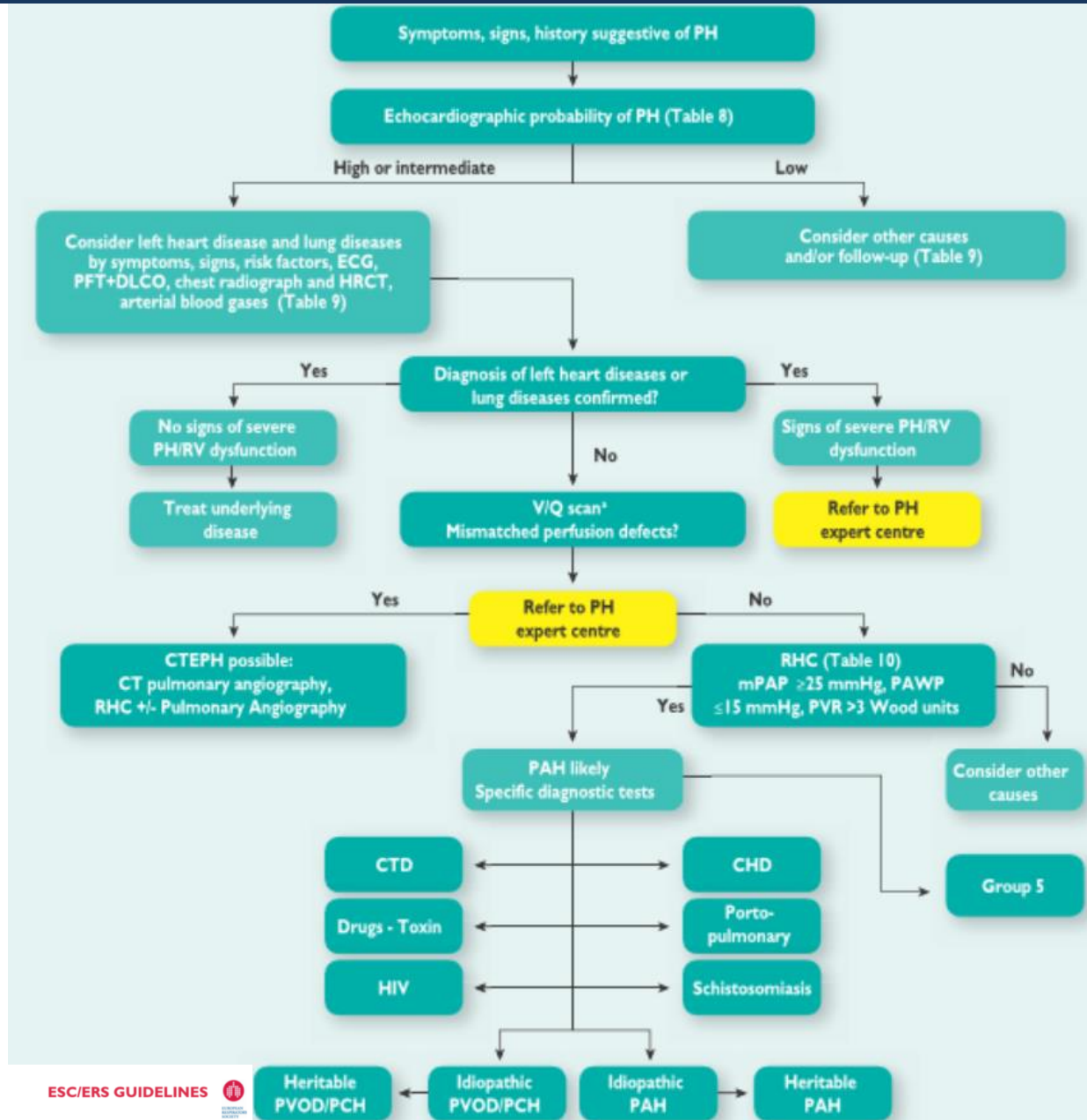
**Figure 1** CT pulmonary angiogram obtained for baseline evaluation in a 46-year-old man with deep venous thrombosis. The patient had no signs or symptoms suggestive of pulmonary embolism. A saddle embolus in the left and right main pulmonary arteries is shown.

#### CLINICAL SIGNIFICANCE

- Silent pulmonary embolism occurs in approximately one third of patients with deep venous thrombosis.
- Silent pulmonary embolism is more frequent in patients with proximal deep venous thrombosis than in those with distal deep venous thrombosis.
- Recurrent pulmonary embolism during treatment for deep venous thrombosis is more frequent in those with silent pulmonary embolism than a first pulmonary embolism in those with no silent pulmonary embolism.
- Silent pulmonary embolism may occur in the central pulmonary arteries.

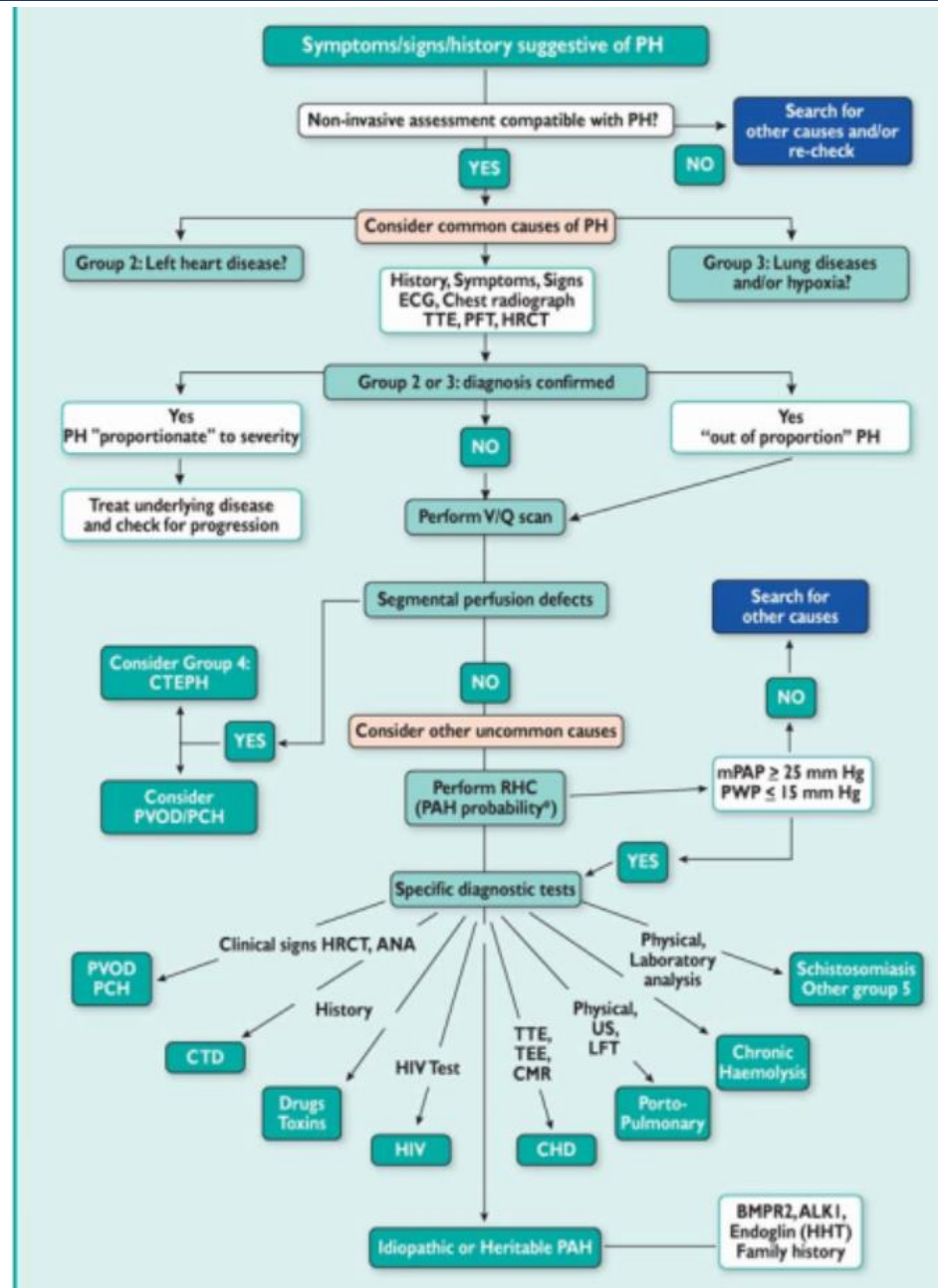


# IP DIAGNOSTIC ALGORITHM ESC-ERS 2015

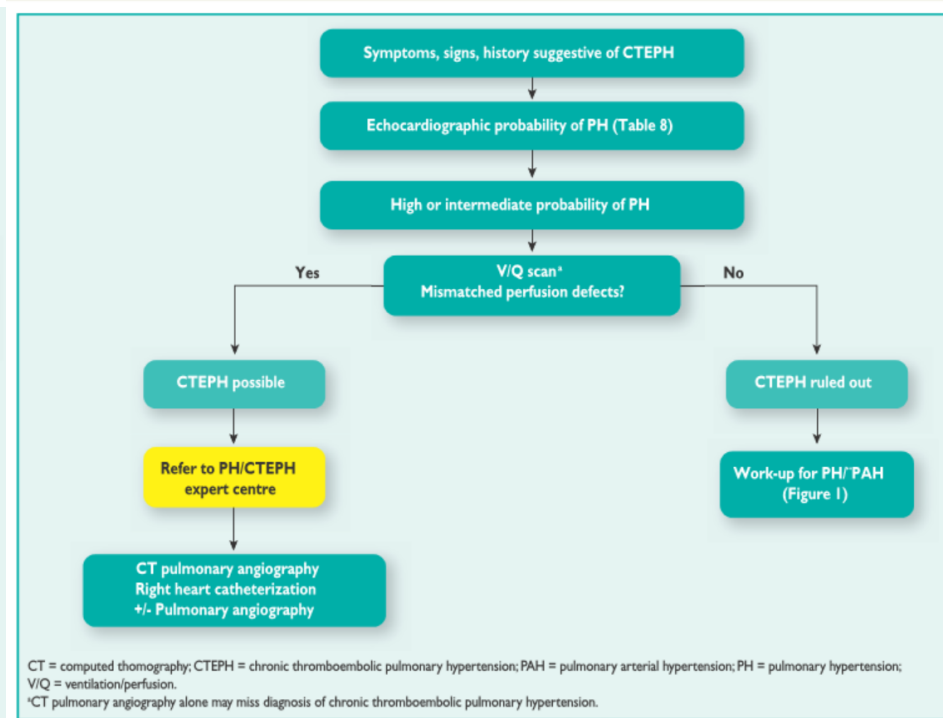
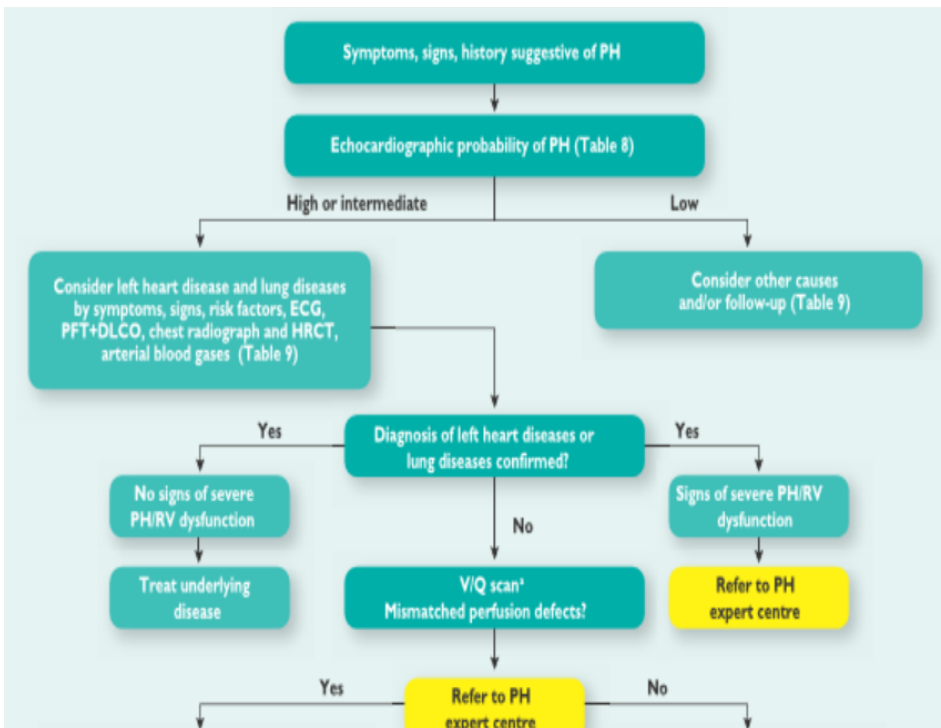




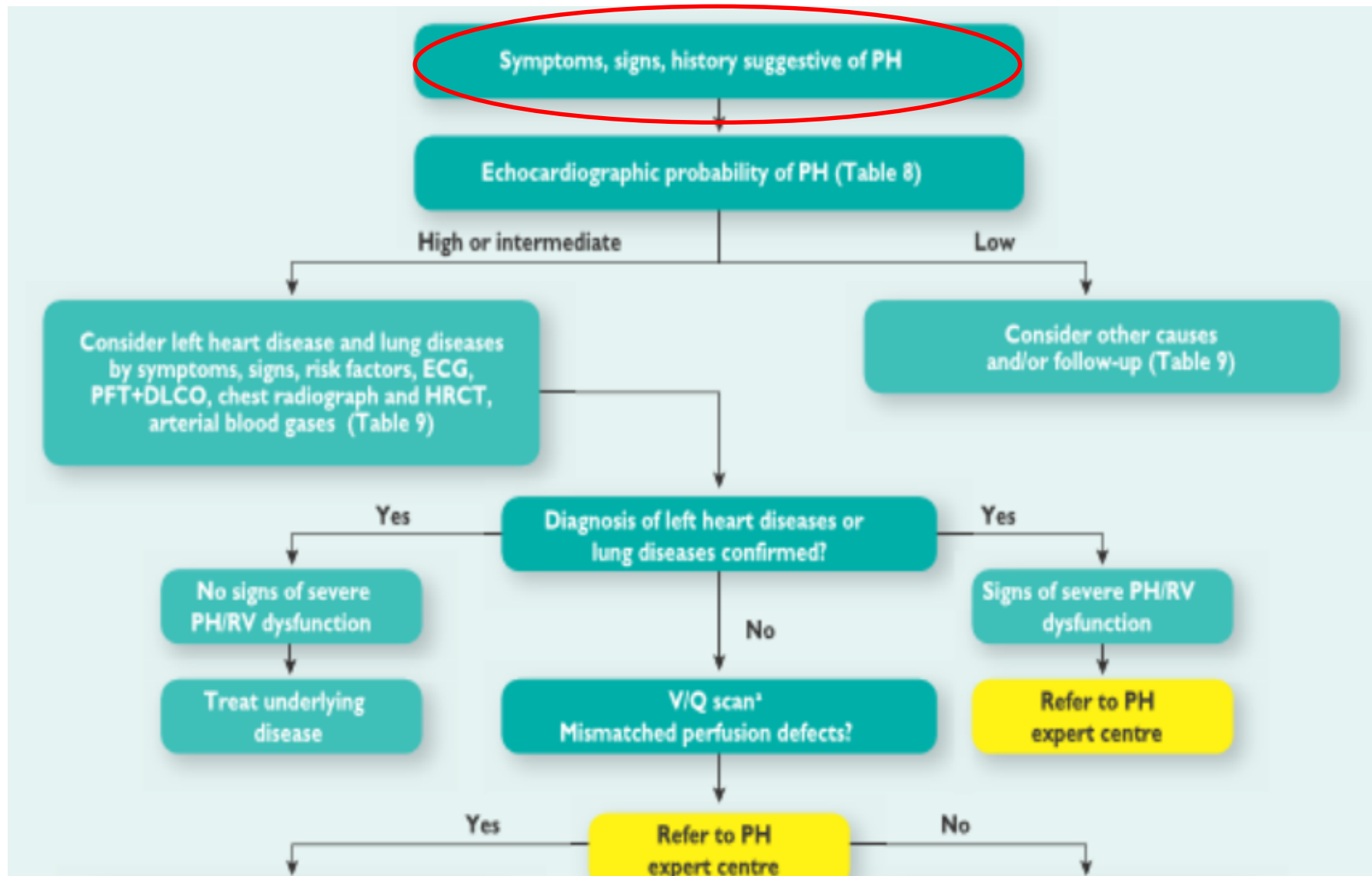
# IP DIAGNOSTIC ALGORITHM ESC-ERS 2009



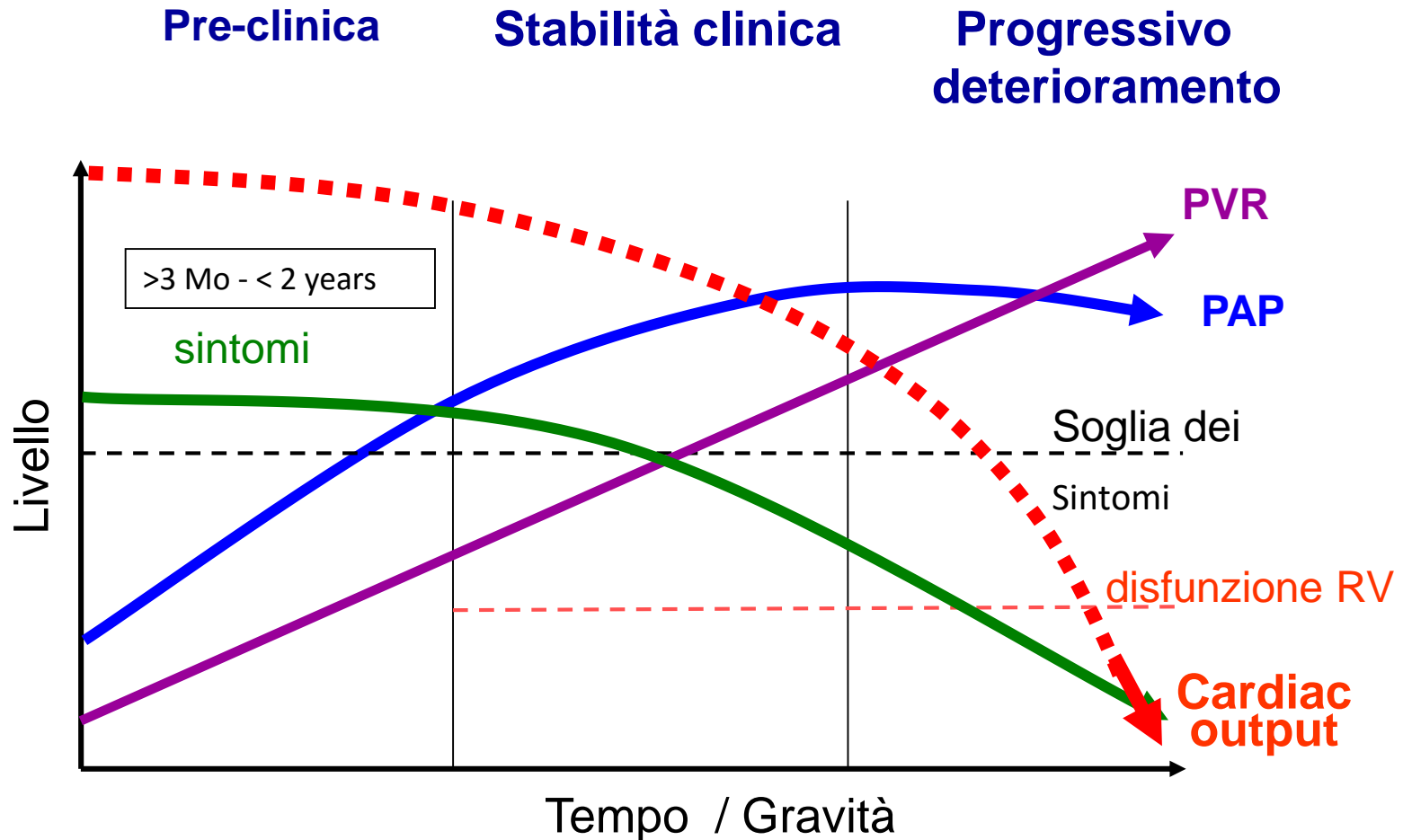
# IP DIAGNOSTIC ALGORITHM



# IP DIAGNOSTIC ALGORITHM



# CTEPH CLINICAL PRESENTATION



<b>Feature</b>	<b>CTEPH</b>	<b>IPAH</b>
<b>Gross pathology</b>	<ul style="list-style-type: none"> <li>• Organized, central thrombi</li> </ul>	<ul style="list-style-type: none"> <li>• Some thrombotic pathology</li> </ul>
<b>Histopathology</b>	<ul style="list-style-type: none"> <li>• Plexogenic arteriopathy</li> </ul>	<ul style="list-style-type: none"> <li>• Plexogenic arteriopathy</li> </ul>
<b>Symptoms</b>	<ul style="list-style-type: none"> <li>• Shortness of breath</li> <li>• Fatigue</li> <li>• Weakness</li> <li>• Haemoptysis (late)</li> </ul>	<ul style="list-style-type: none"> <li>• Shortness of breath</li> <li>• Fatigue</li> <li>• Weakness</li> <li>• Angina (late)</li> <li>• Syncope (late)</li> </ul>
<b>Signs</b>	<ul style="list-style-type: none"> <li>• PH and right heart failure</li> <li>• RV third heart sound</li> <li>• Oedema (late)</li> <li>• Elevated jugular v. p. (late)</li> <li>• Ascites (late)</li> </ul>	<ul style="list-style-type: none"> <li>• PH and right heart failure</li> <li>• RV third heart sound</li> </ul>
<b>Family history</b>	<ul style="list-style-type: none"> <li>• No</li> </ul>	<ul style="list-style-type: none"> <li>• Seen in 6–10% of cases</li> </ul>
<b>Genetic basis</b>	<ul style="list-style-type: none"> <li>• None identified</li> </ul> <p>rare paediatric cases</p>	<ul style="list-style-type: none"> <li>• Genetic basis in up to 30% of sporadic IPAH cases (e.g., BMPR-II)</li> </ul>

# CTEPH RISK FACTORS

- Survivors of acute pulmonary embolism
- Thrombophilic disorders (LAC, Ab antiphospholipid, protein C and S deficiency, prothrombin gene mutation, activated protein C resistance including factor V mutation, antithrombin III deficiency, elevated factor VIII)
- Splenectomy
- History of malignancy
- Have very high blood pressure in their lungs' arteries (greater than 50 mmHg) when their PE is discovered
- Ventriculo-atrial shunt and infected pace-makers
- Thyroid replacement therapy
- Blood groups other than 0

J Thorac Dis. 2015 Nov;7(11):1927-38. doi: 10.3978/j.issn.2072-1439.2015.11.43.

**Incidence and risk factors of chronic thromboembolic pulmonary hypertension in patients after acute pulmonary embolism.**

Yang S<sup>1</sup>, Yang Y<sup>1</sup>, Zhai Z<sup>1</sup>, Kuang T<sup>1</sup>, Gong J<sup>1</sup>, Zhang S<sup>1</sup>, Zhu J<sup>1</sup>, Liang L<sup>1</sup>, Shen YH<sup>1</sup>, Wang C<sup>1</sup>.

Bonderman D *et al.* *Eur Respir J* 2009;33:325–31.  
Pengo V *et al.* *N Engl J Med* 2004;350:2257–264.  
Abul Y *et al.* *Chron Respir Dis* 2014;11:73–81.



European Heart Journal (2016) 37, 67–119  
doi:10.1093/eurheartj/ehv317

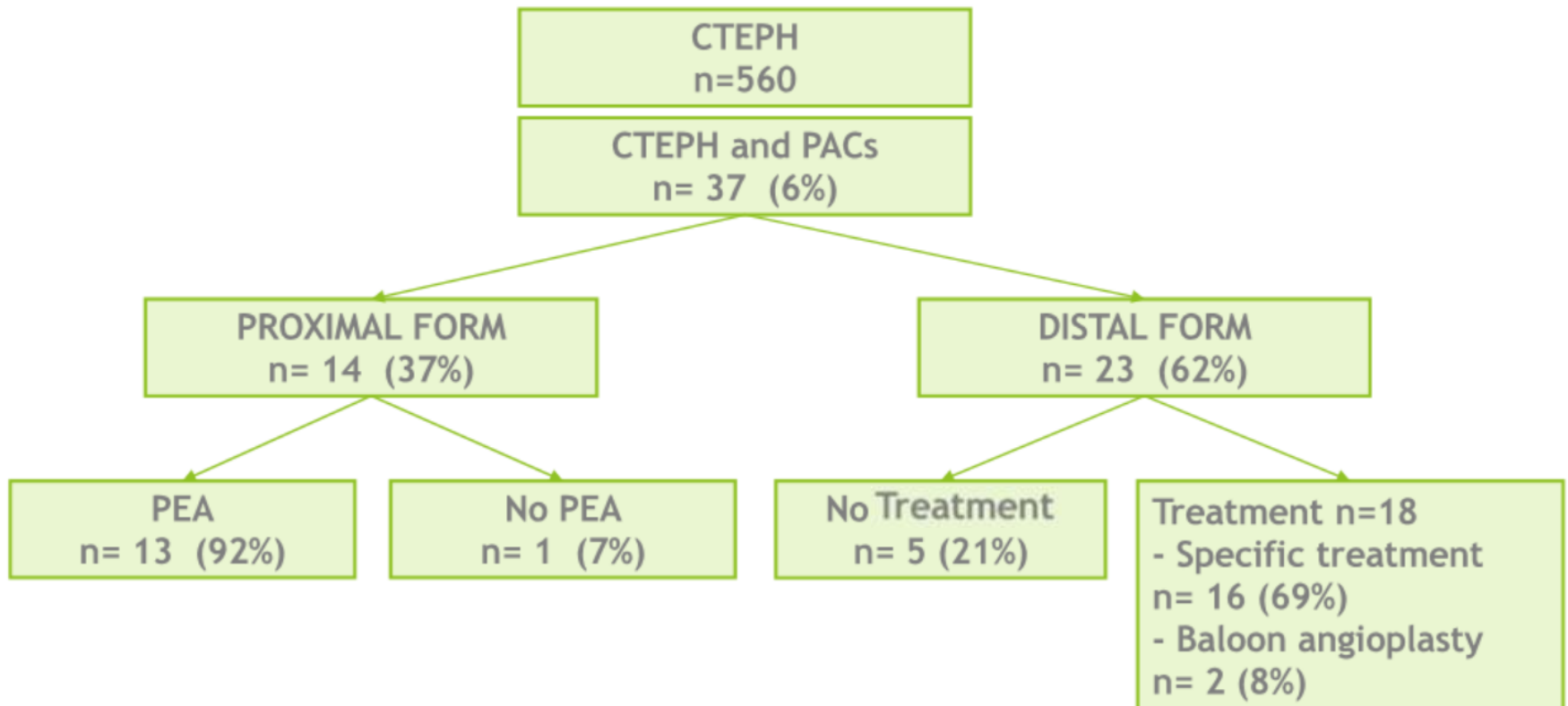
ESC/ERS GUIDELINES





# CTEPH RISK FACTORS

## CTEPH associated with indwelling Port-A-Cath systems (PACs)



# CTEPH RISK FACTORS

- Survivors of acute pulmonary embolism
- Thrombophilic disorders (LAC, Ab antiphospholipid, protein C and S deficiency, prothrombin gene mutation, activated protein C resistance including factor V mutation, antithrombin III deficiency, elevated factor VIII)
- Splenectomy
- History of malignancy
- Have very high blood pressure in their lungs' arteries (greater than 50 mm Hg) when their PE is discovered
- Ventriculo-atrial shunt and infected pace-makers
- Thyroid replacement therapy
- Blood groups other than O
- **Port A Cath presence**

[J Thorac Dis. 2015 Nov;7\(11\):1927-38. doi: 10.3978/j.issn.2072-1439.2015.11.43.](#)

[Incidence and risk factors of chronic thromboembolic pulmonary hypertension in patients after acute pulmonary embolism.](#)

[Yang S<sup>1</sup>, Yang Y<sup>1</sup>, Zhai Z<sup>1</sup>, Kuang T<sup>1</sup>, Gong J<sup>1</sup>, Zhang S<sup>1</sup>, Zhu J<sup>1</sup>, Liang L<sup>1</sup>, Shen YH<sup>1</sup>, Wang C<sup>1</sup>.](#)

[Bonderman D et al. Eur Respir J 2009;33:325–31.](#)  
[Pengo V et al. N Engl J Med 2004;350:2257–264.](#)  
[Abul Y et al. Chron Respir Dis 2014;11:73–81.](#)



European Heart Journal (2016) 37, 67–119  
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES



# ACUTE PE

*Fedullo Clin Chest Med 2001 ; 22 : 561-581*

COMPLETE RESOLUTION

Progression to CTEPH

1% - 4% (2 years)

0.1% - 9.1% (2 years)

*Lang IM, Risk factors and  
basic mechanisms of CTEPH:  
a current understaging. ERJ  
2013, 41:462-468*

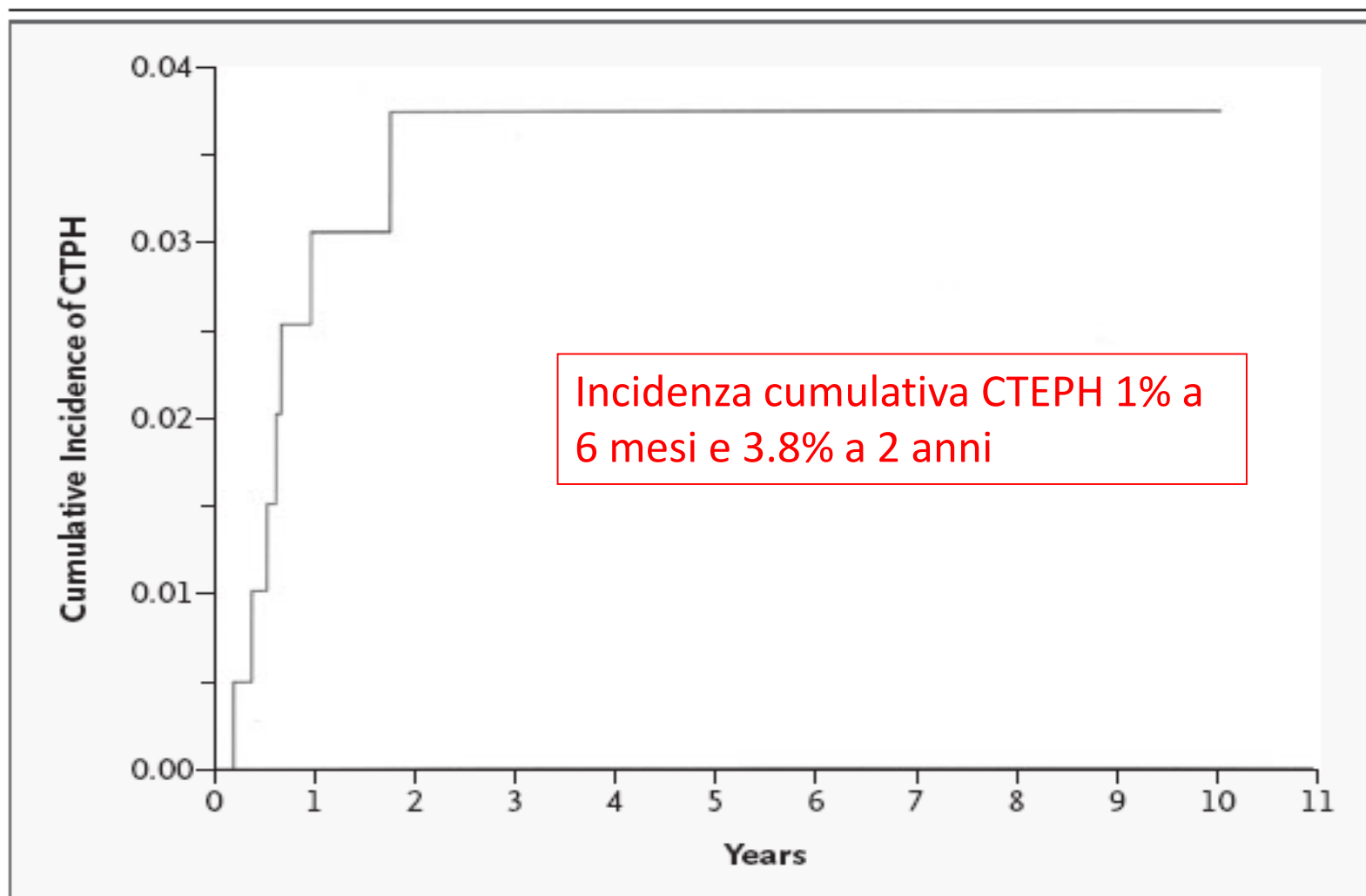
PARTIAL RESOLUTION with normal clinical status

*Focal chronic PE : 13%*

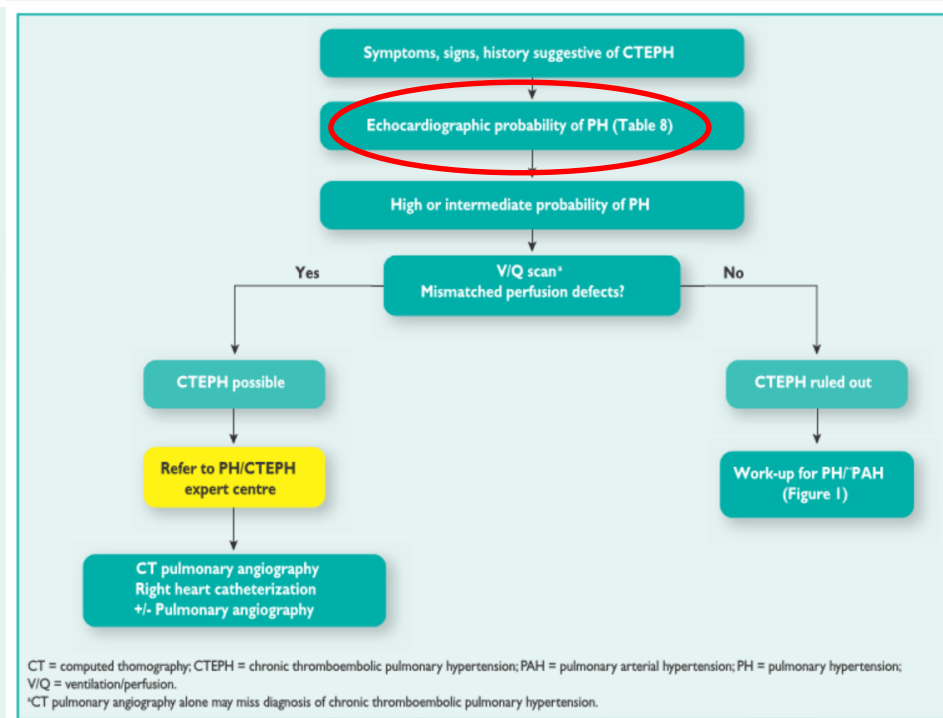
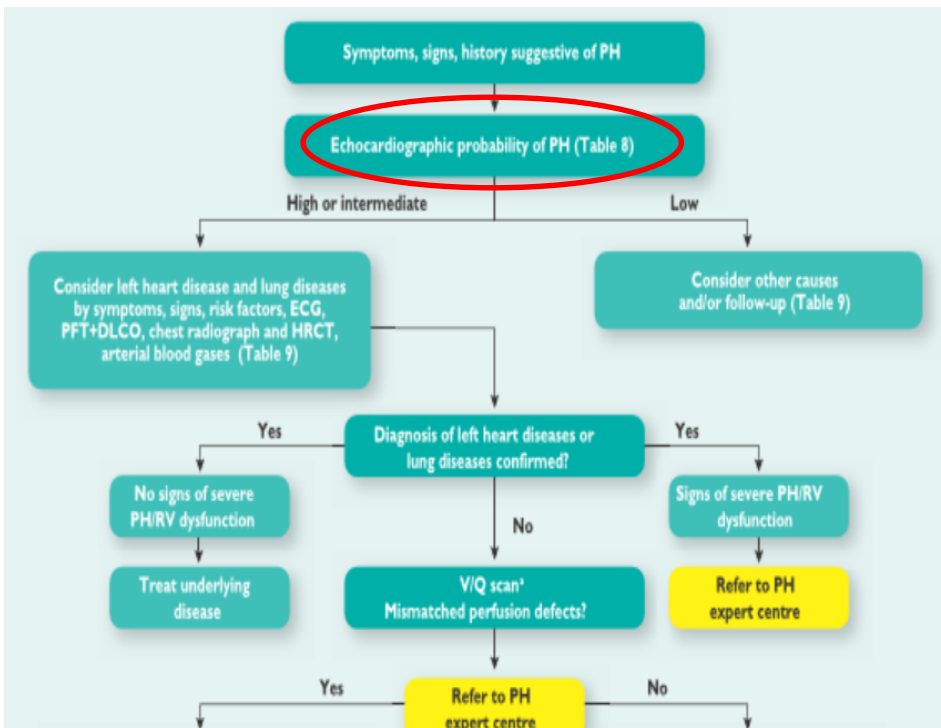
ORIGINAL ARTICLE

# Incidence of Chronic Thromboembolic Pulmonary Hypertension after Pulmonary Embolism

Vittorio Pengo, M.D., Anthonie W.A. Lensing, M.D., Martin H. Prins, M.D.,  
Antonio Marchiori, M.D., Bruce L. Davidson, M.D., M.P.H., Francesca Tiozzo, M.D.,  
Paolo Albanese, M.D., Alessandra Biasiolo, D.Sci., Cinzia Pegoraro, M.D.,  
Sabino Iliceto, M.D., and Paolo Prandoni, M.D.,  
for the Thromboembolic Pulmonary Hypertension Study Group\*



# IP DIAGNOSTIC ALGORITHM





# ECHOCARDIOGRAPHY 2009

	Class <sup>a</sup>	Level <sup>b</sup>
<b>Echocardiographic diagnosis: PH unlikely</b>		
Tricuspid regurgitation velocity $\leq 2.8$ m/s, PA systolic pressure $\leq 36$ mmHg, and no additional echocardiographic variables suggestive of PH	I	B
<b>Echocardiographic diagnosis: PH possible</b>		
Tricuspid regurgitation velocity $\leq 2.8$ m/s, PA systolic pressure $\leq 36$ mmHg, but presence of additional echocardiographic variables suggestive of PH	IIa	C
Tricuspid regurgitation velocity 2.9–3.4 m/s, PA systolic pressure 37–50 mmHg with/without additional echocardiographic variables suggestive of PH	IIa	C
<b>Echocardiographic diagnosis: PH likely</b>		
Tricuspid regurgitation velocity $> 3.4$ m/s, PA systolic pressure $> 50$ mmHg, with/without additional echocardiographic variables suggestive of PH	I	B
<b>Exercise Doppler echocardiography is not recommended for screening of PH</b>	III	C

# ECHOCARDIOGRAPHY 2015

**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' <sup>a</sup>	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	

PH = pulmonary hypertension.

<sup>a</sup>See Table 8B.

**Table 8B** Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in Table 8A

A: The ventricles <sup>a</sup>	B: Pulmonary artery <sup>a</sup>	C: Inferior vena cava and right atrium <sup>a</sup>
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 <sup>2</sup>
	PA diameter >25 mm.	

# ECHOCARDIOGRAPHY

**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' <sup>a</sup>	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	

PH = pulmonary hypertension.

<sup>a</sup>See Table 8B.

**Table 8B** Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in Table 8A

A: The ventricles <sup>a</sup>	B: Pulmonary artery <sup>a</sup>	C: Inferior vena cava and right atrium <sup>a</sup>
Right ventricle/left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior vena 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 <sup>2</sup>
	PA diameter >25 mm.	

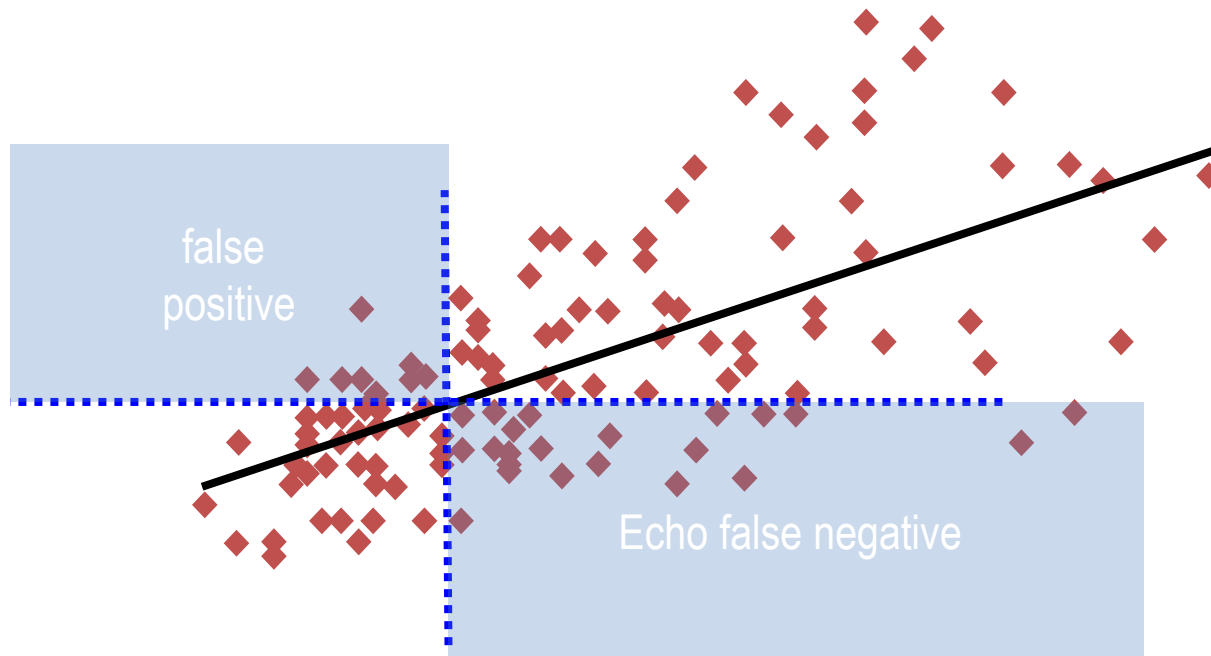
# ECHOCARDIOGRAPHY 2015

**patients with symptoms compatible with pulmonary hypertension, with or without risk factors for pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension**

Echocardiographic probability of PH	Without risk factors or associated condition for PAH or CTEPH <sup>d</sup>	Class <sup>a</sup>	Level <sup>b</sup>	With risk factors or associated conditions for PAH or CTEPH <sup>c</sup>	Class <sup>a</sup>	Level <sup>b</sup>	Ref <sup>c</sup>
Low	Alternative diagnosis should be considered	IIa	C	Echo follow-up should be considered	IIa	C	
Intermediate	Alternative diagnosis, echo follow-up, should be considered	IIa	C	Further assessment of PH including RHC should be considered <sup>e</sup>	IIa	B	45, 46
	Further investigation of PH may be considered <sup>e</sup>	IIb					
High	Further investigation of PH (including RHC <sup>e</sup> ) is recommended	I	C	Further investigation of PH <sup>e</sup> including RHC is recommended	I	C	

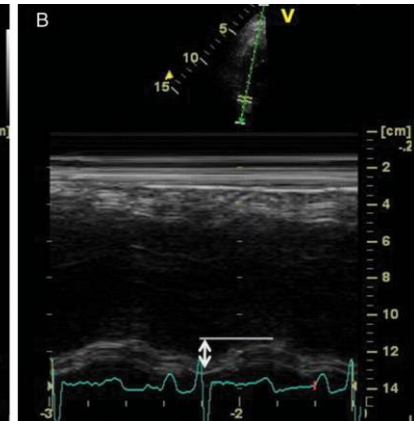
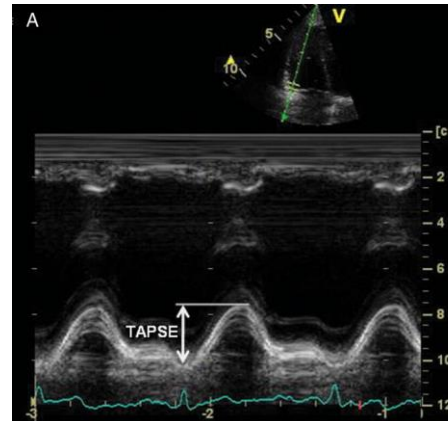
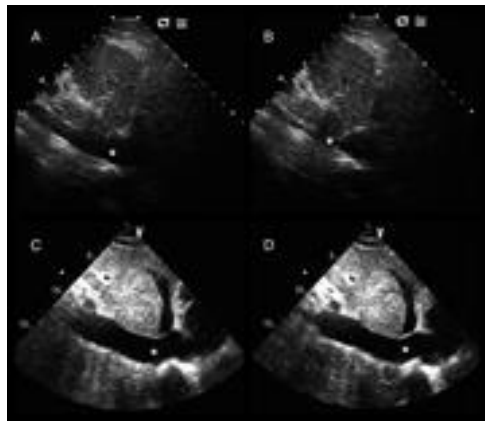
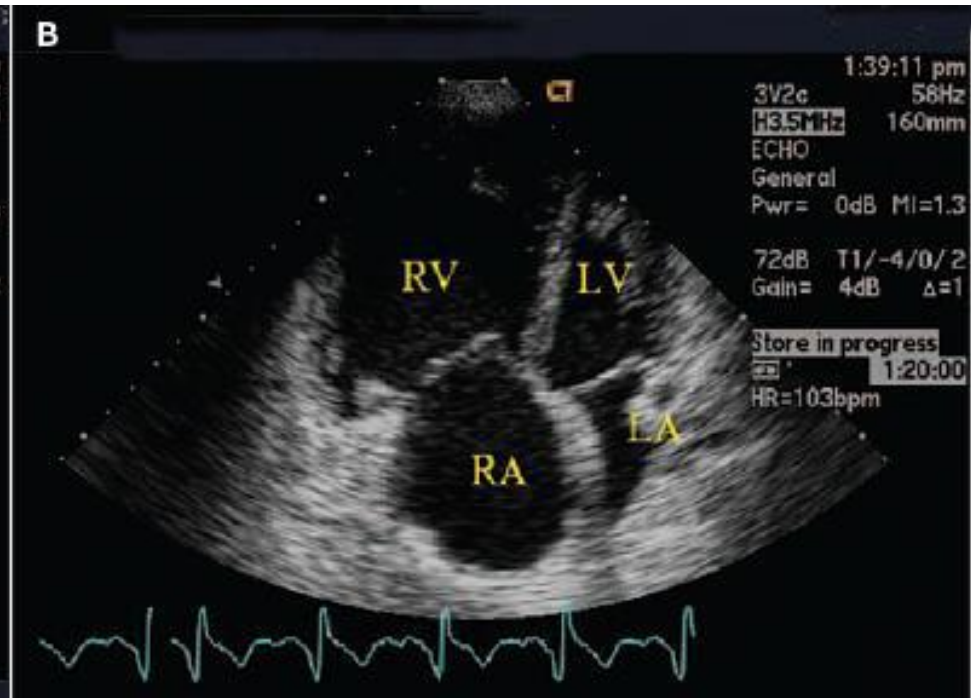
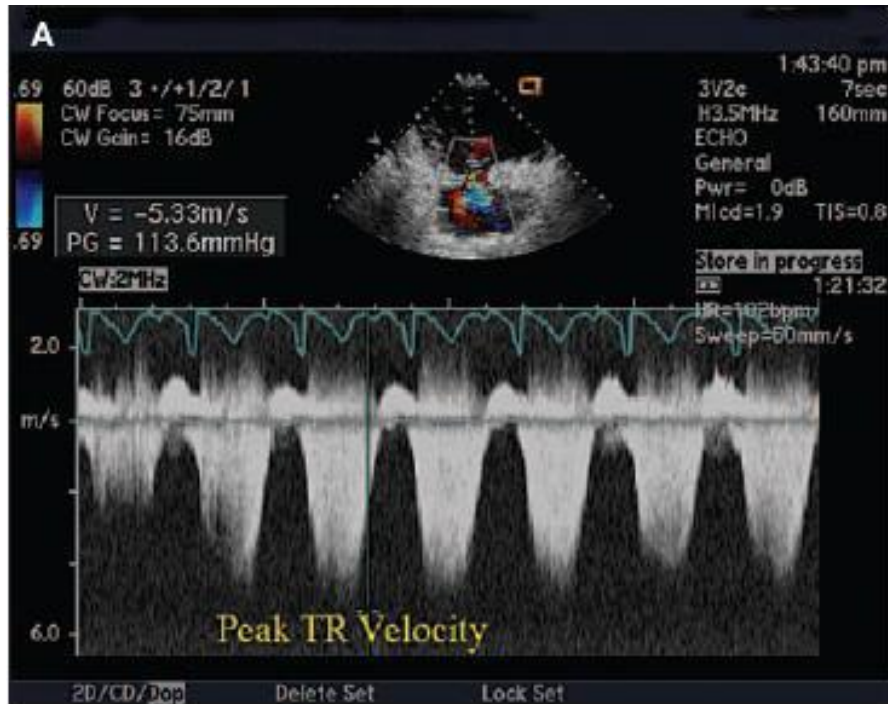
# ECHOCARDIOGRAPHY

La diagnosi definitiva di ipertensione polmonare necessita del cateterismo cardiaco destro perché l'ecocardio stima con variazioni anche  $>20\%$





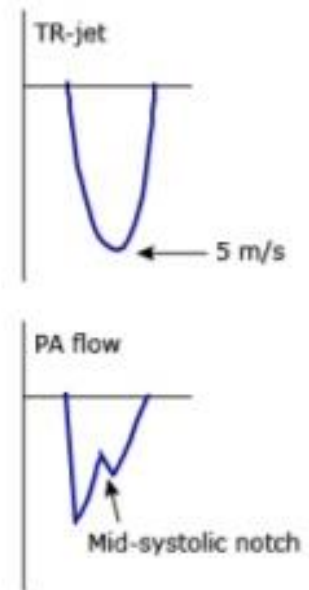
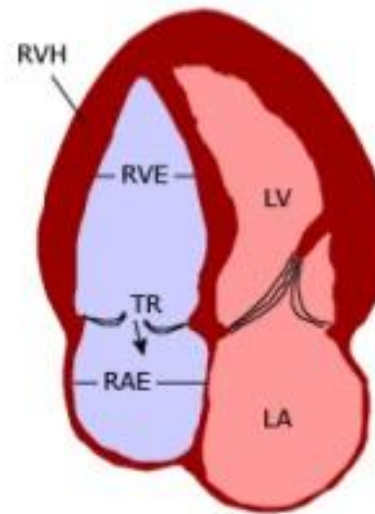
# ECHOCARDIOGRAPHY



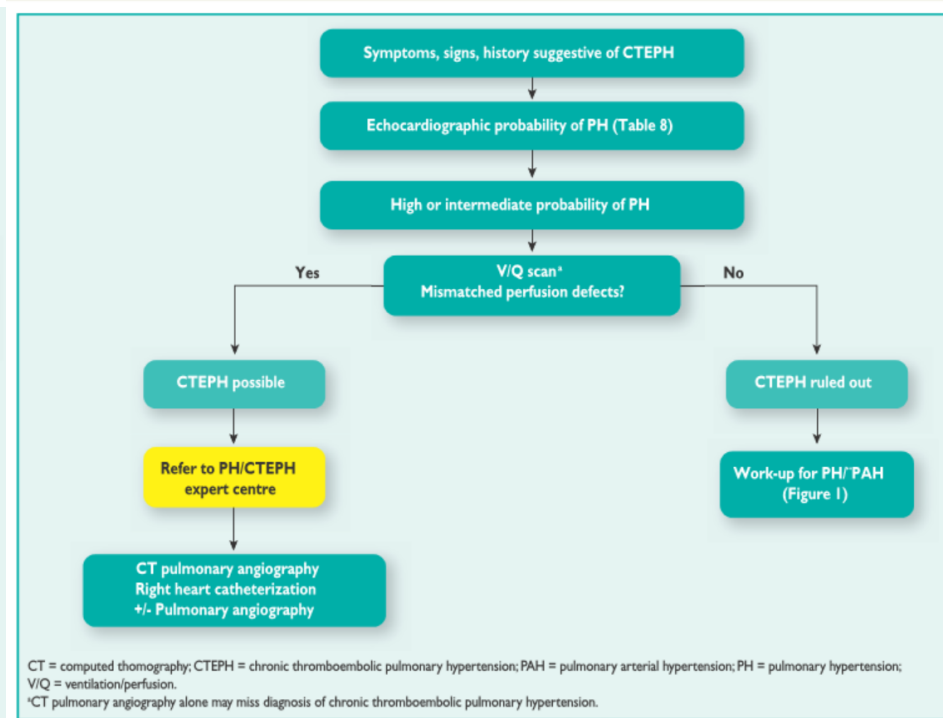
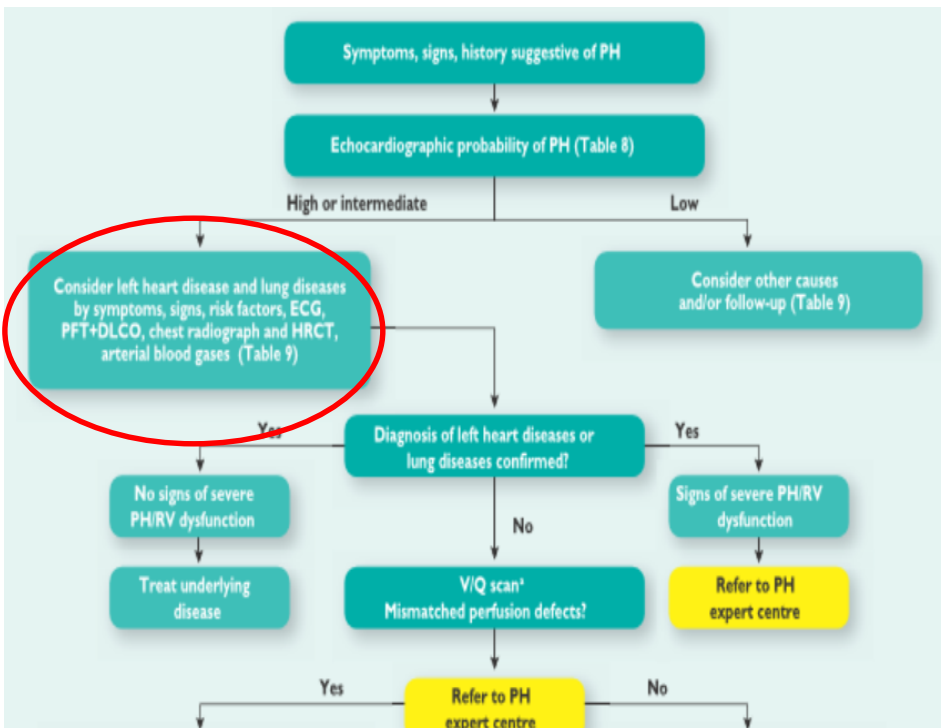


# ECHOCARDIOGRAPHY

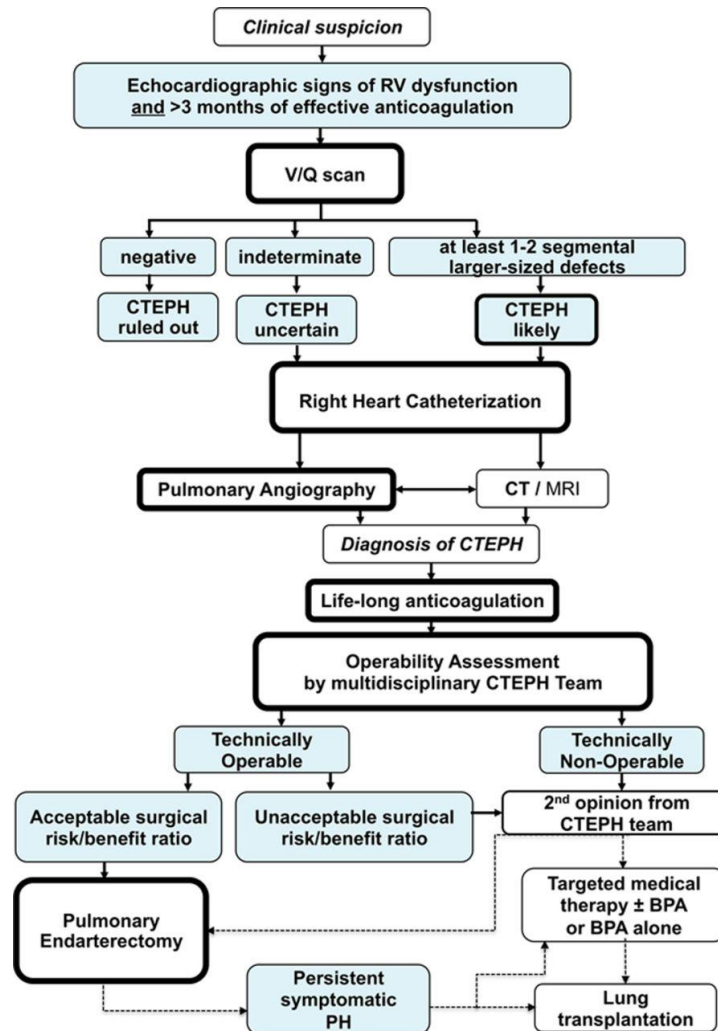
- Main findings are :
  1. Right ventricular enlargement (RVE).
  2. Right ventricular hypertrophy (RVH).
  3. Right atrial enlargement (RAE).
  4. Functional tricuspid regurgitation (TR) with a high velocity regurgitant jet by Doppler (TR jet), and a mid-systolic notch on the pulmonary artery Doppler flow tracing (PA flow).
  5. The interventricular septum is shifted toward the left ventricular cavity.



# IP DIAGNOSTIC ALGORITHM

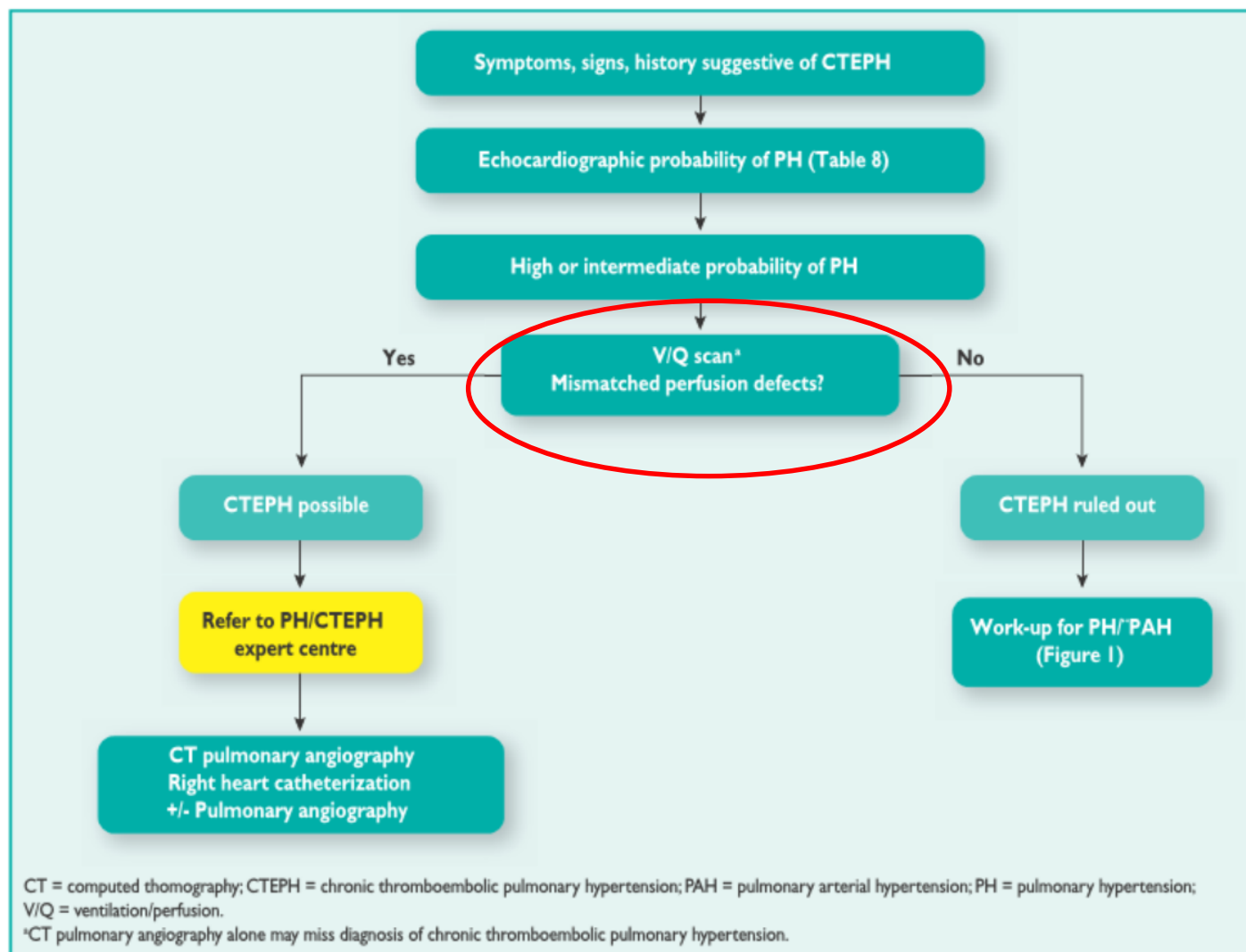


# CONTEMPORARY DIAGNOSTIC AND THERAPEUTIC ALGORITHM



Lang I M , and Madani M Circulation. 2014;130:508-518

# CONTEMPORARY DIAGNOSTIC AND THERAPEUTIC ALGORITHM



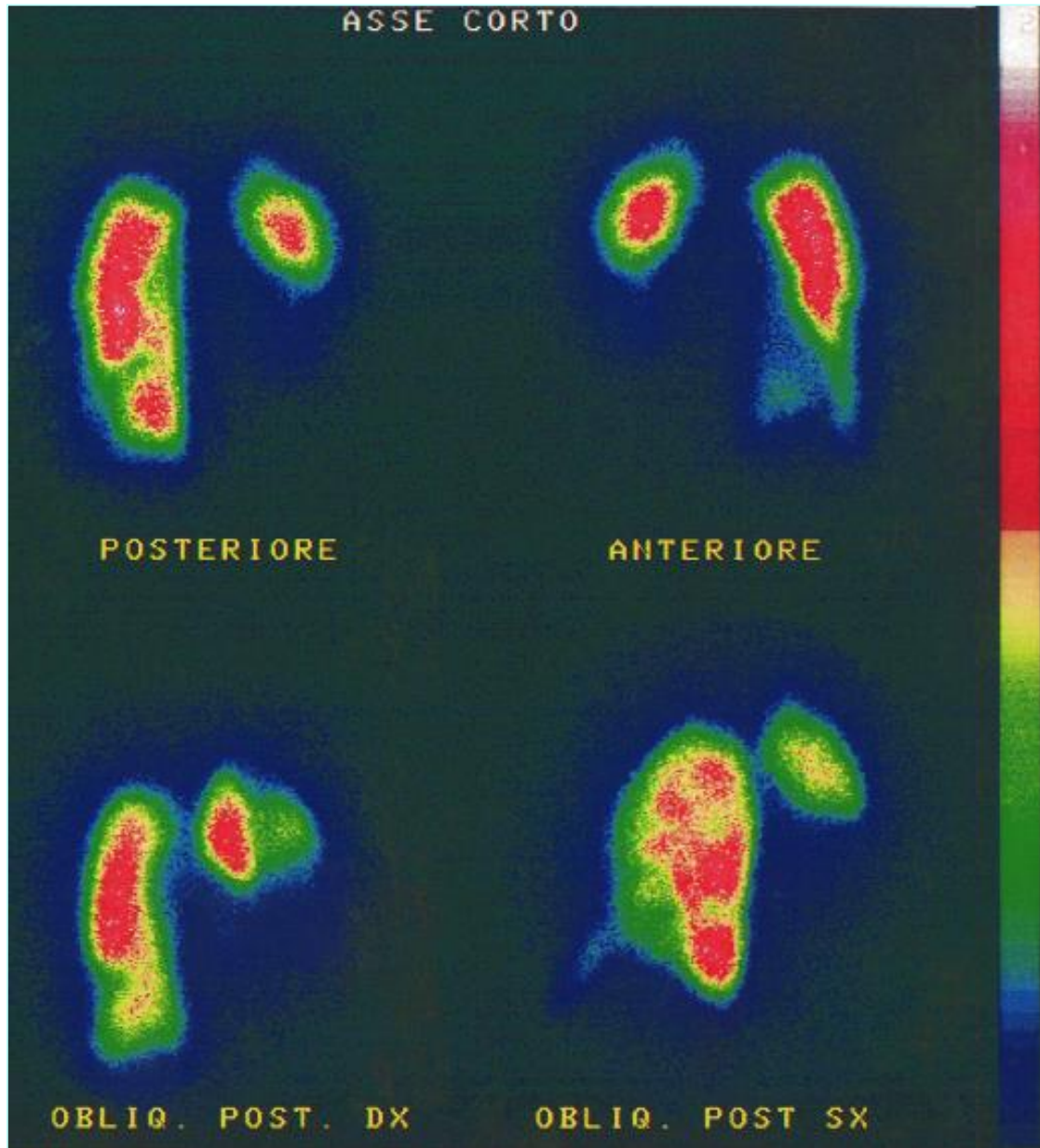
# CTEPH DIAGNOSIS

- **Sospetto clinico**
  - Scintigrafia V/Q
- **Conferma**
  - Cateterismo cardiaco destro
  - AngioTAC

**VQ scan**: maggior sensibilità e specificità, minor radio-esposizione, minori costi

**AngioTAC**: falsi positivi (sarcoma a.polmonare, trombi prossimali associati a PAH e cardiopatie cogenite), non sensibile per lesioni a carico dei vasi subsegmentari (inoperabili ma passibili di trattamento medico), può mostrare reperti associati (oligoemia a mosaico, ipertrofia delle arterie bronchiali, infarti polmonari)

# VENTILATION/PERFUSION LUNG SCAN



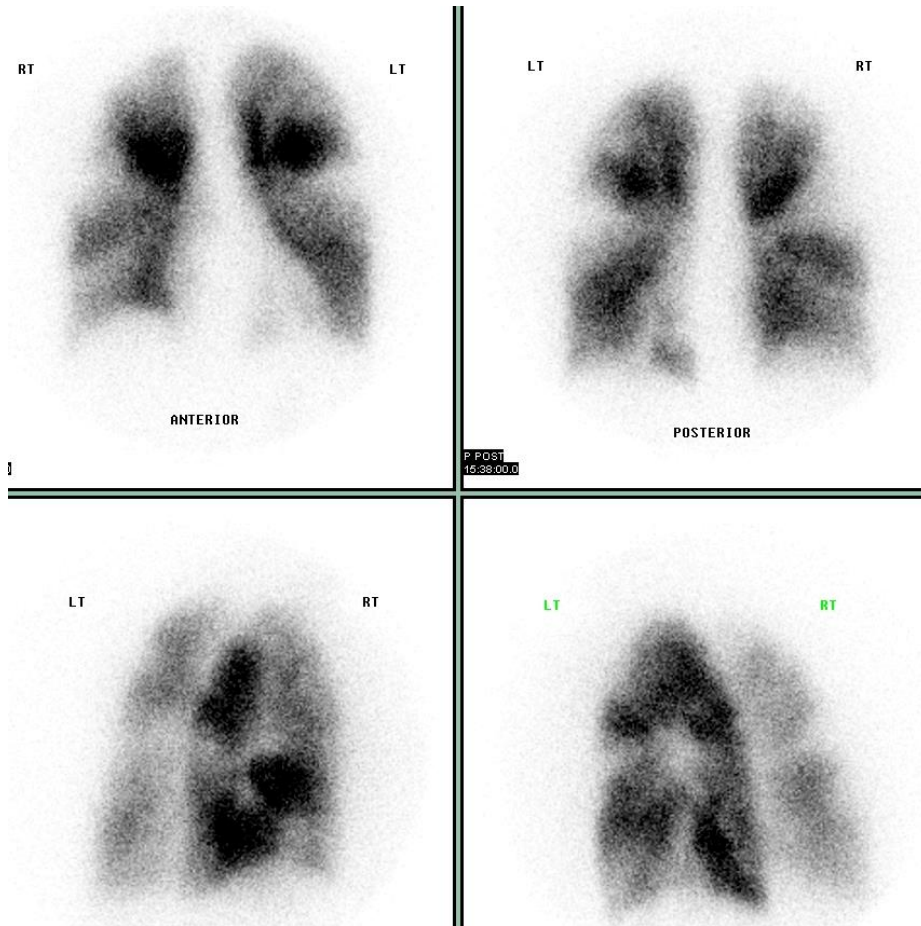
## Scintigrafia polmonare perfusionale

- Una scintigrafia ventilatoria normale e difetti cuneiformi a quella perfusoria è caratteristico per CTEPH
- Il registro internazionale ha evidenziato come nel 9,7% sia alterata la scintigrafia perfusoria mentre quella ventilatoria nel 19%

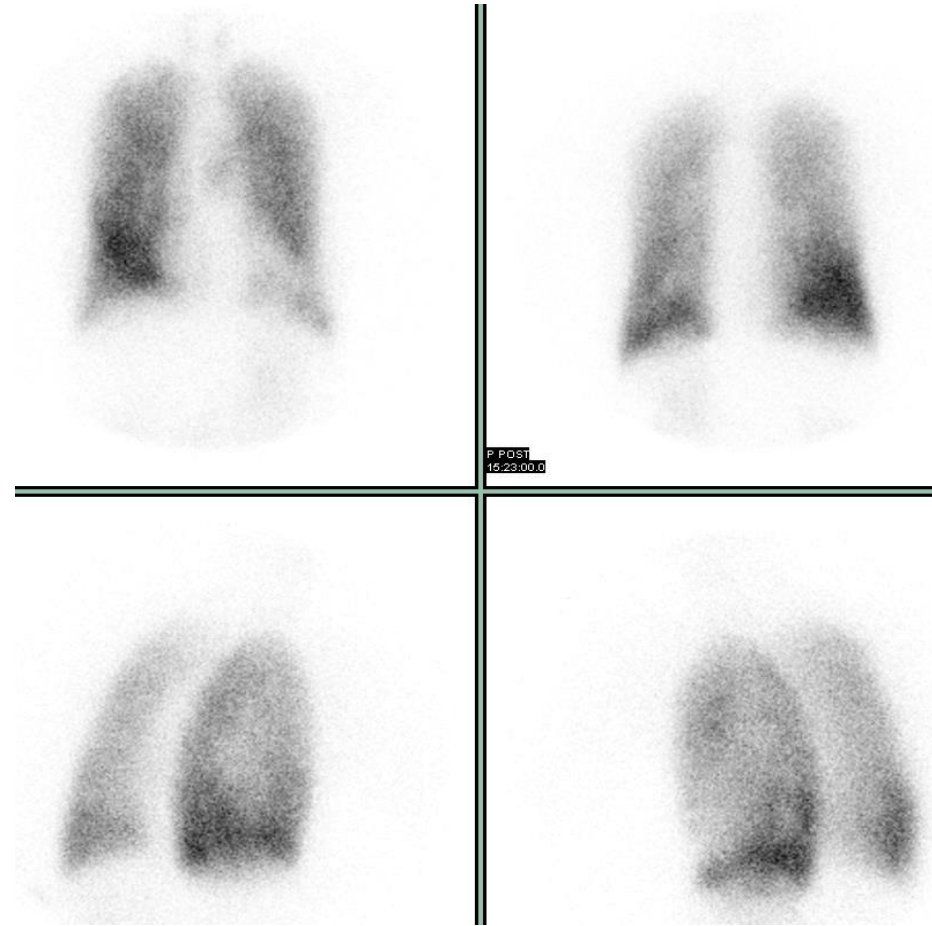


# VENTILATION/PERFUSION LUNG SCAN

## Thromboembolic Disease



## IPAH



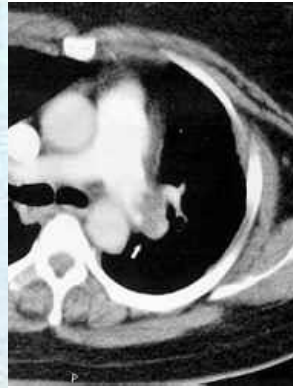
# VENTILATION/PERFUSION LUNG SCAN

Diagnosi Differenziale:  
CTEPH distale vs Ipertensione polmonare idiopatica

Scintigrafia  
Perfusionale

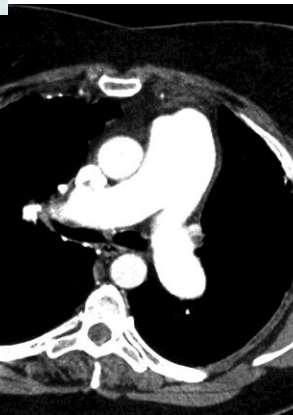
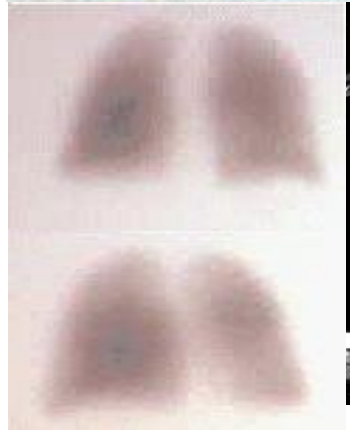
TC  
Spirale

circolo polmonare:  
difetti segmentari  
multipli



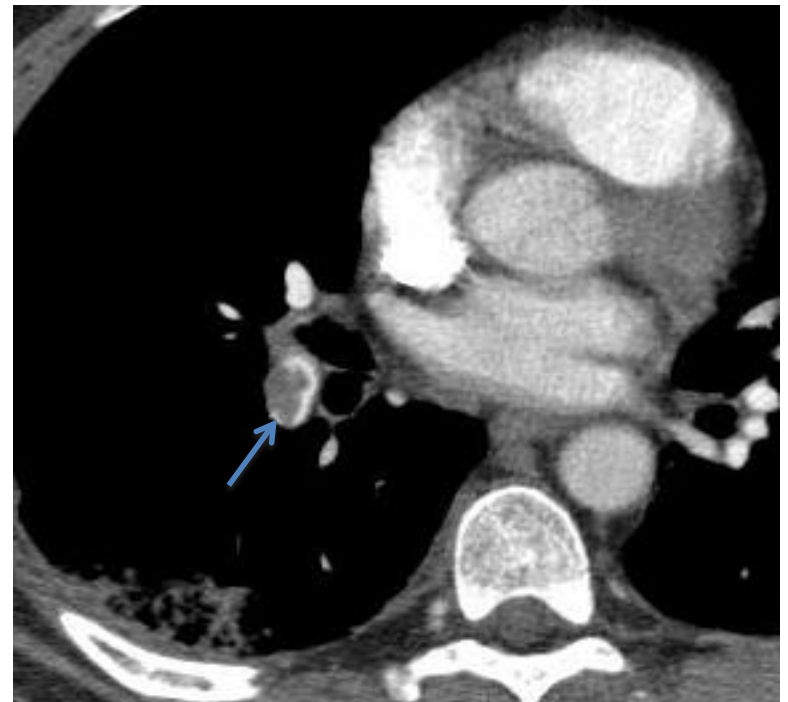
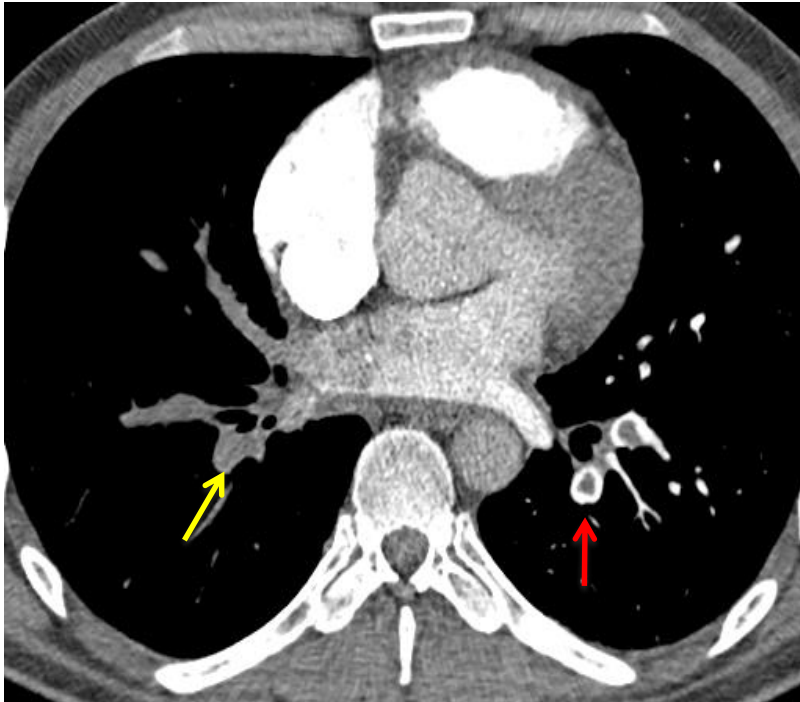
circolo polmonare: difetti  
riempimento rami lobari  
e/o segmentari, piccoli  
infarti polmonari

Tronco comune  
polmonare: normale o  
difetti "sfumati"



Tronco comune  
polmonare: normale o  
trombi stratificati

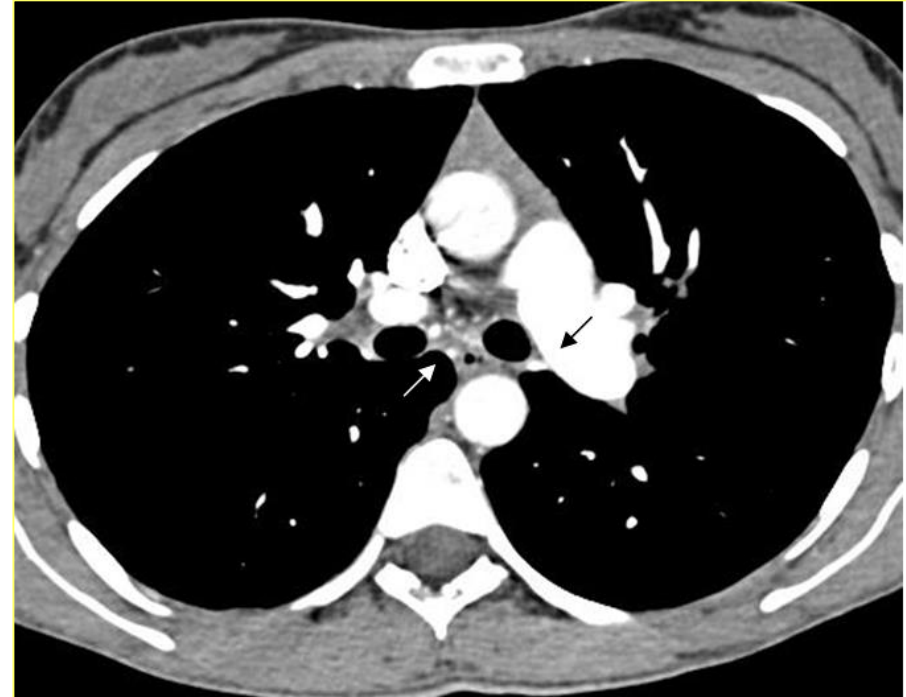
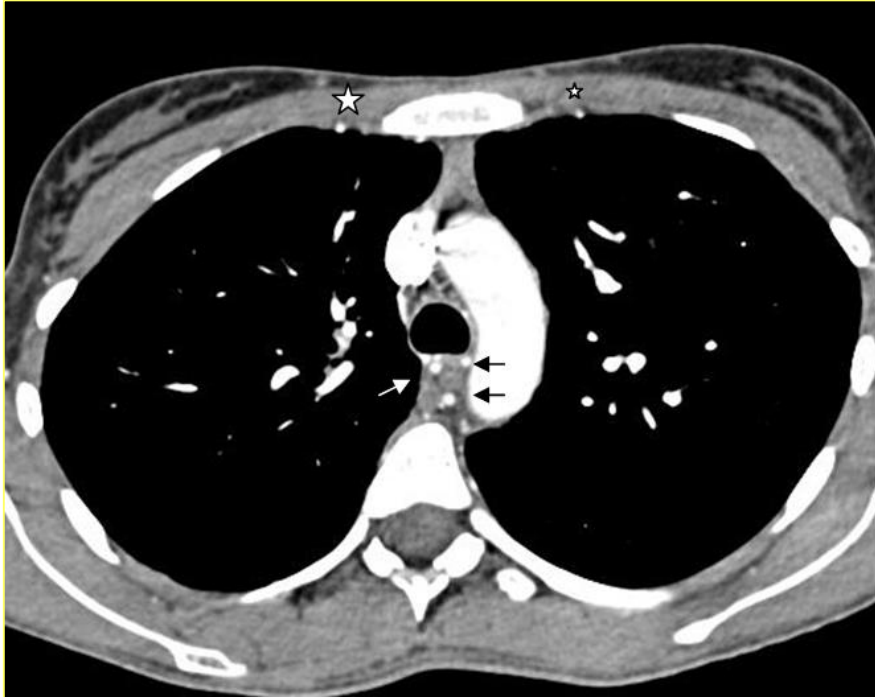
# HRCT – CT ANGIOGRAPHY - MRI



# HRCT – CT ANGIOGRAPHY - MRI

## BRONCHIAL ARTERY DILATATION IN CTEPH

*Hasegawa et al. AJR 2004; 182: 67-72*



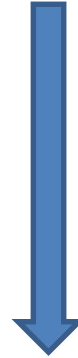
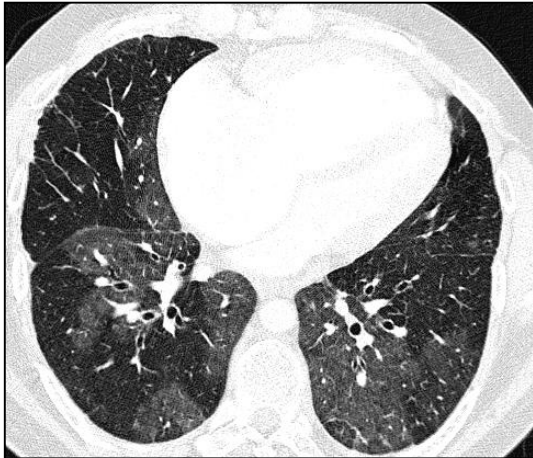
***Acute pulmonary embolism : 7%*** (2 out of 27 patients)

***VS***

***Chronic or recurrent pulmonary embolism : 50%*** (7 out of 14 patients)

# HRCT – CT ANGIOGRAPHY - MRI

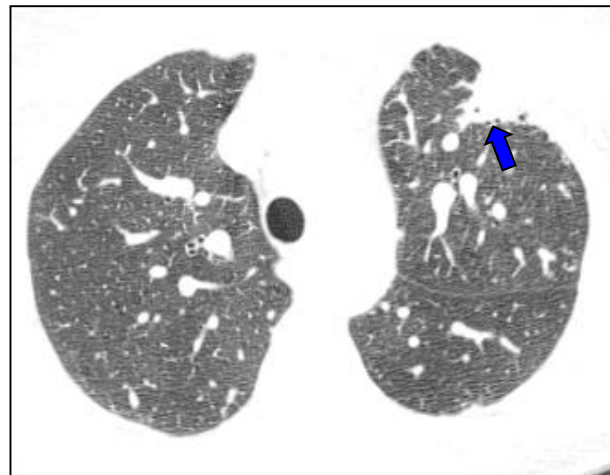
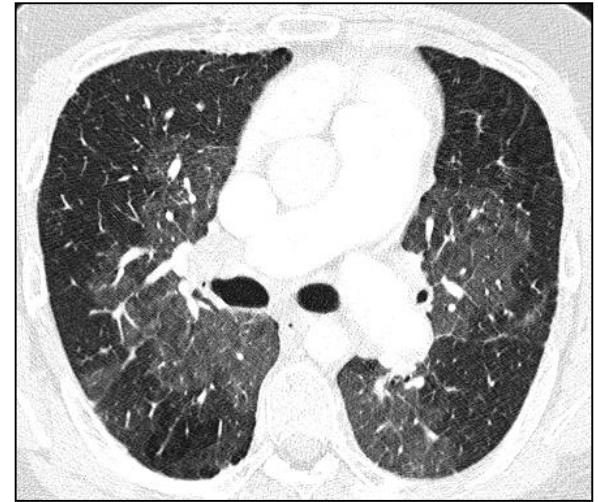
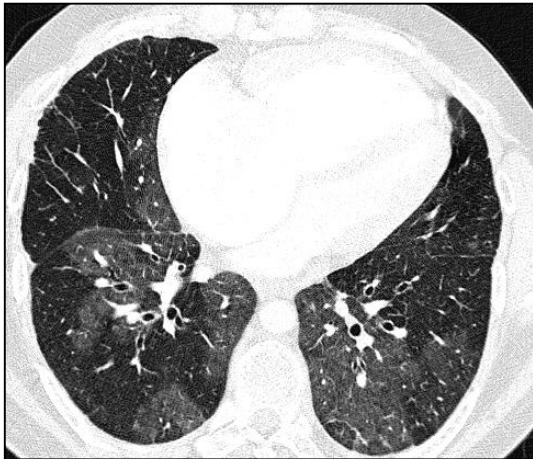
*Oligoemia a mosaic: CTEPH OR AIRTRAPPING?*



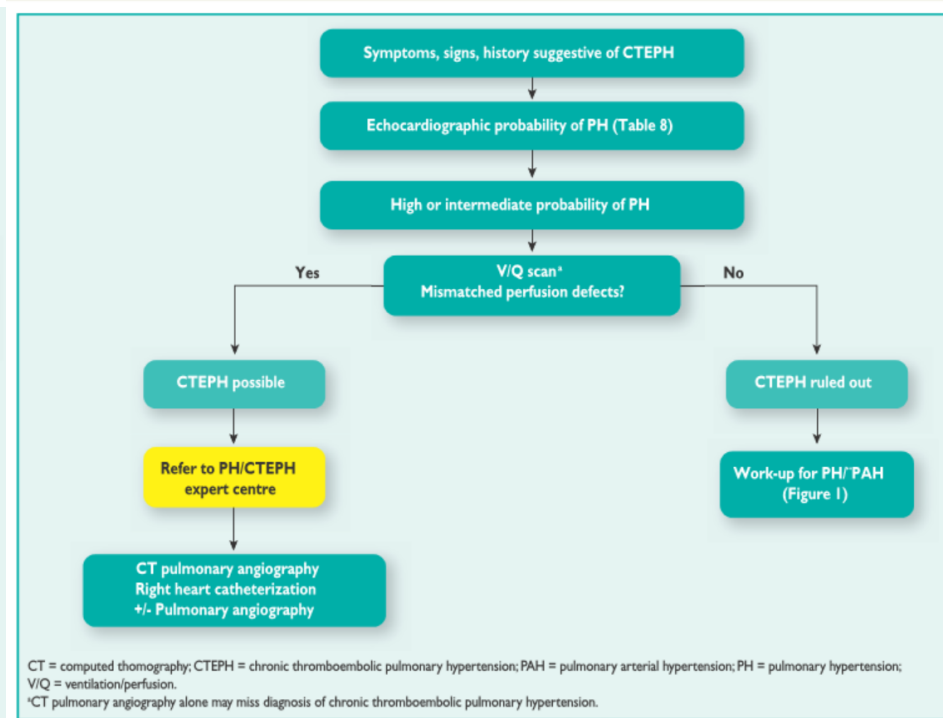
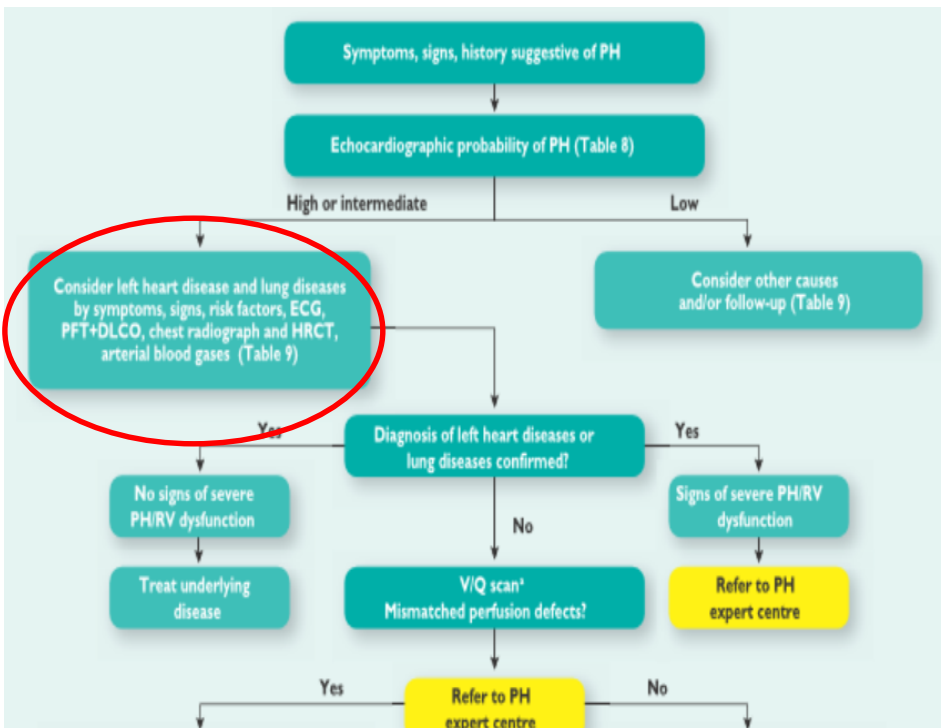


# HRCT – CT ANGIOGRAPHY - MRI

## *Oligoemia a mosaico e Infarto polmonare*



# IP DIAGNOSTIC ALGORITHM



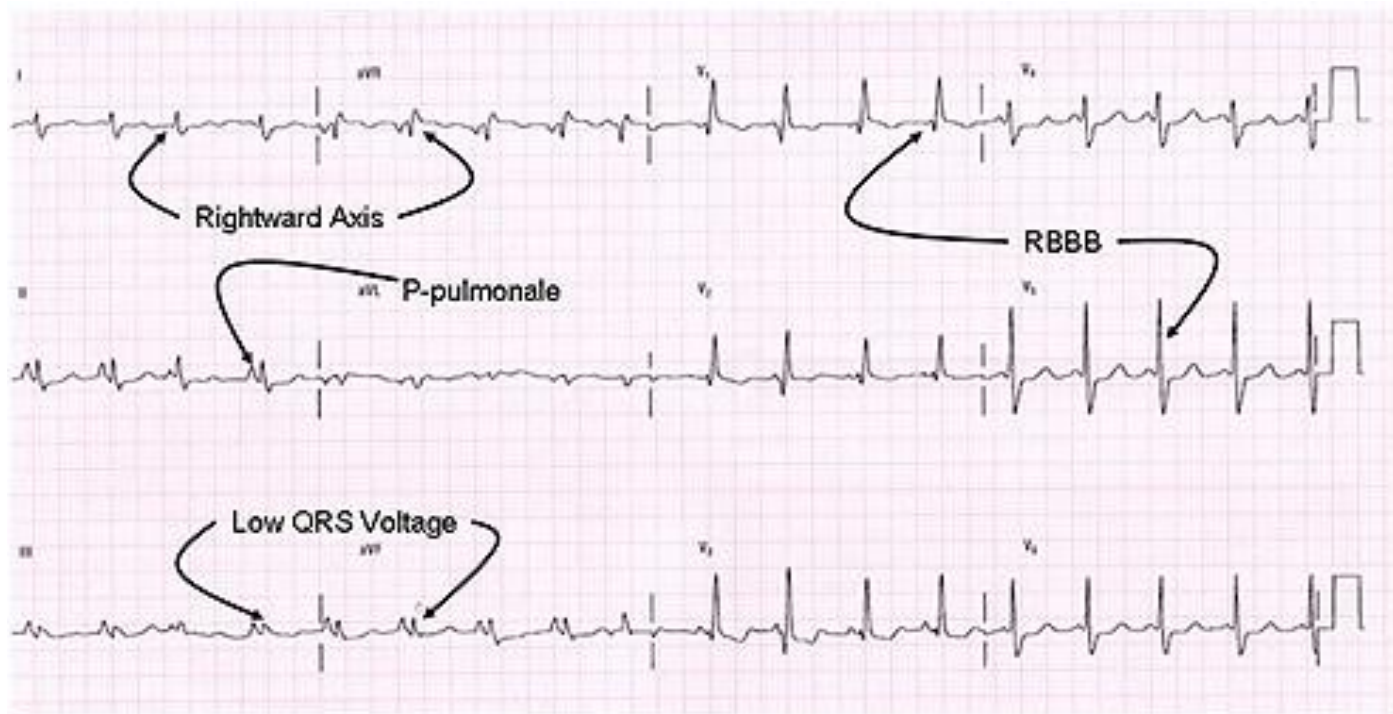
CT = computed tomography; CTEPH = chronic thromboembolic pulmonary hypertension; PAH = pulmonary arterial hypertension; PH = pulmonary hypertension; V/Q = ventilation/perfusion.

\*CT pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.




# ECG

- P pulmonale
- Right axis deviation
- Right bundle branch block
- QTc prolongation (late)
- Supraventricular arrhythmias (late)

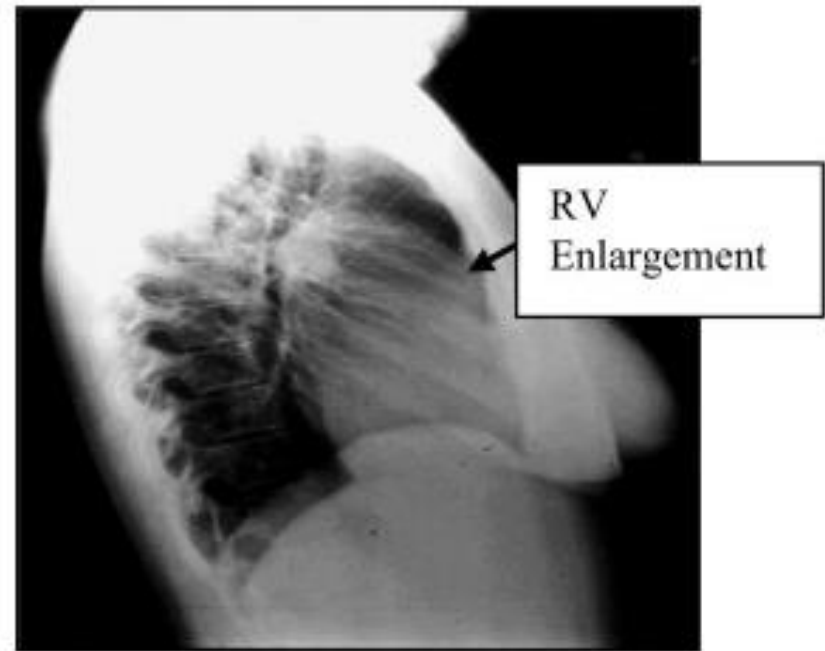
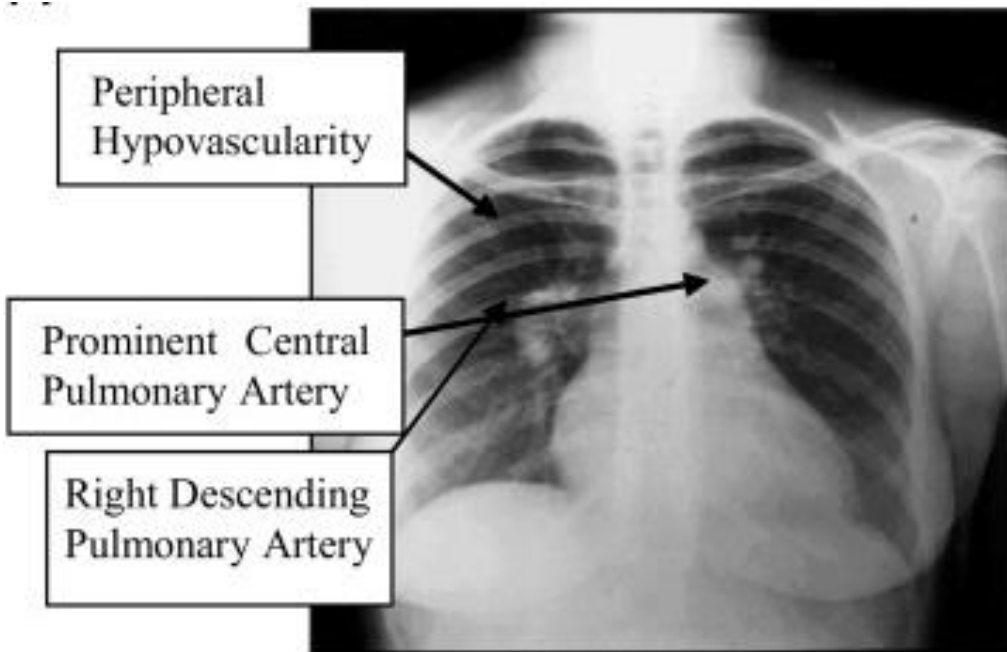


# ABG –BT

- PaO<sub>2</sub> normal or decreased at rest
  - PaCO<sub>2</sub> normal or decreased at rest
  - pH increased (late-severe)
- 
- Routine biochemistry-haematology
  - Thyroid function
  - Thrombophilia screening
  - Antiphospholipid antibodies, anticardiolipin antibodies, lupus anticoagulant.
  - BNP/proBNP
- **Antinuclear antibodies** (anti-centromere, dsDNA, anti-Ro, U3-RNP, B23, Th/To and U1-RNP, U3-RNP, anticardiolipin antibodies..)
  - HIV, Hepatitis, ...
  - .....

# CHEST RADIOGRAPH

- Normal (90%)
- Central pulmonary arterial dilatation
- RA and RV enlargement
- Peripheral hypovascularity



# PULMONARY FUNCTION TEST

- Decreased lung diffusion capacity for carbon monoxide (DLCO) and KCO
- Normal or Mild reduction of lung volumes related to disease severity



# PULMONARY FUNCTION TEST

PFT	CTEPH	PH group 3
Lung diffusion capacity for carbon monoxide	• Decreased DLCO and KCO	• Decreased DLCO and $\geq$ KCO
Lung volumes	• Normal or Mild reduction related to disease severity	• Increased RV /FRC related to specific disease
Lung airflow	• Normal	• Normal / obstruction related to specific disease

# HRCT – CT ANGIOGRAPHY - MRI

- Still considered inferior to CT
- May be complimentary and used in according to local experience practice

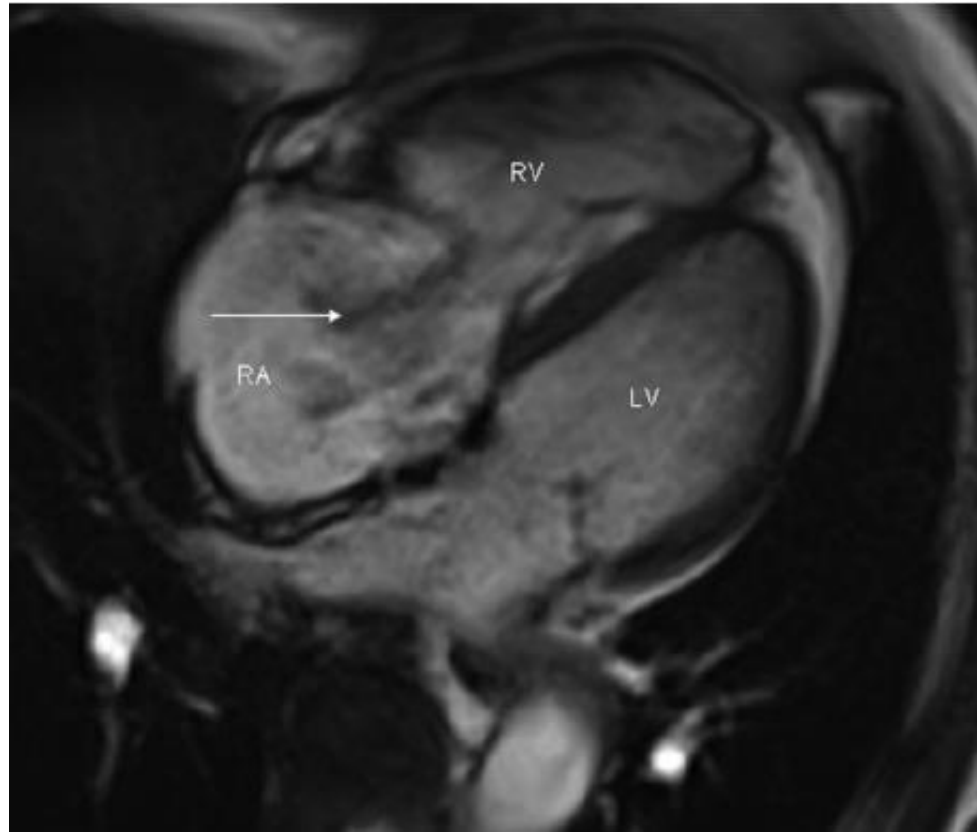


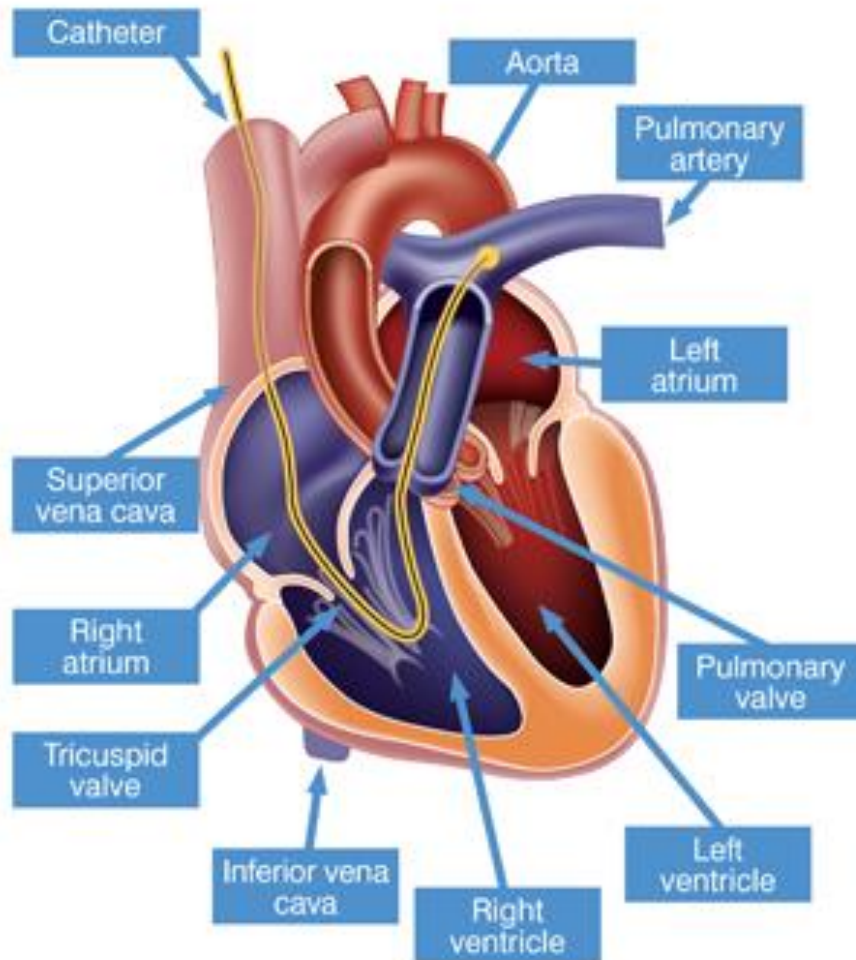
TABLE 1 Summary of techniques used in the diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH)

Technique	Advantages	Potential disadvantages
<b>Ventilation/perfusion (<math>V/Q'</math>) scan</b>	Essential for diagnosing CTEPH Sensitivity >96% Negative result rules out CTEPH Can distinguish between large-vessel occlusive and small-vessel pulmonary vascular disease Less radiation exposure than CTPA Less likely than other techniques to detect incidental findings Avoids potential problems with intravenous contrast	Can give intermediate probability result, which cannot rule out CTEPH May underestimate the burden of vascular obstruction
<b>Conventional pulmonary angiography (CPA)</b>	Can be combined with RHC to provide imaging and haemodynamic information	Invasive
<b>Right heart catheterisation (RHC)</b>	Mandatory in diagnosing CTEPH Essential measurements of mean arterial pressure and pulmonary capillary wedge pressure Information on disease severity, right heart function, mixed venous oxygen saturation	Invasive
<b>Computed tomography pulmonary angiography (CTPA)</b>	Noninvasive High-resolution images New scanners can provide multi-planar and three-dimensional reconstructions of the vascular tree More anatomical detail than MRI: information about vascular wall thickness and surrounding structures not appreciable with CPA No need for direct catheter access Can reveal associated findings, e.g. bronchial artery collaterals, mosaic perfusion patterns Lower cost than CPA Rapid acquisition, even in breathless patients DECTA has improved the detection of distal CTEPH	Sensitivity 51% (lower than $V/Q'$ scan) Chronic disease looks different to acute PE: additional training required Risk of false positives (e.g. pulmonary artery sarcoma) May miss disease in distal segmental or subsegmental vessels May miss inoperable patients who could begin medical therapy May underestimate clot burden
<b>Cardiac magnetic resonance imaging (MRI)</b>	Noninvasive, no radiation exposure Morphological, functional and anatomical assessment of heart and pulmonary circulation Useful for repeat studies, e.g. pre- and post-operative monitoring Phase-contrast MRI can quantify blood flow and peak velocity in the main pulmonary artery Contrast-enhanced magnetic resonance angiography has similar sensitivity to CTPA	Limited availability, expensive, time consuming

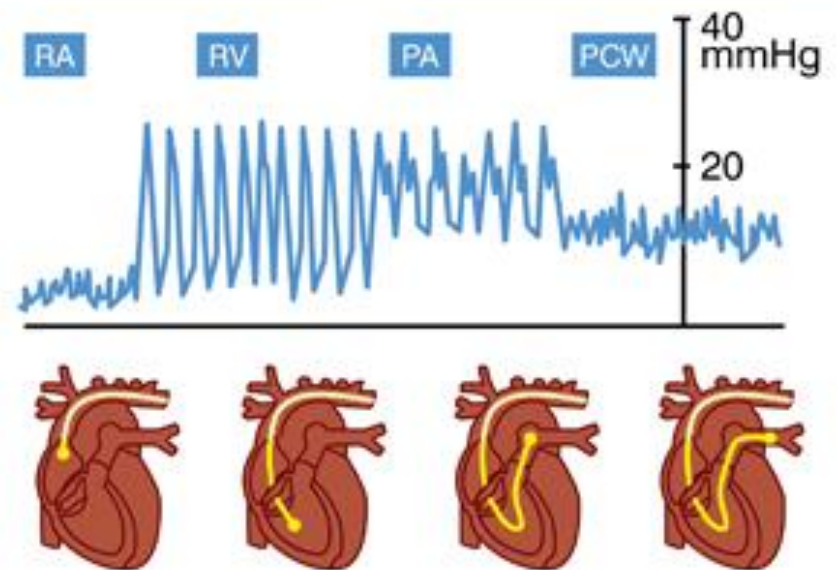
PE: pulmonary embolism; DECTA: dual-energy computed tomography angiography. Information from [14, 17, 18].



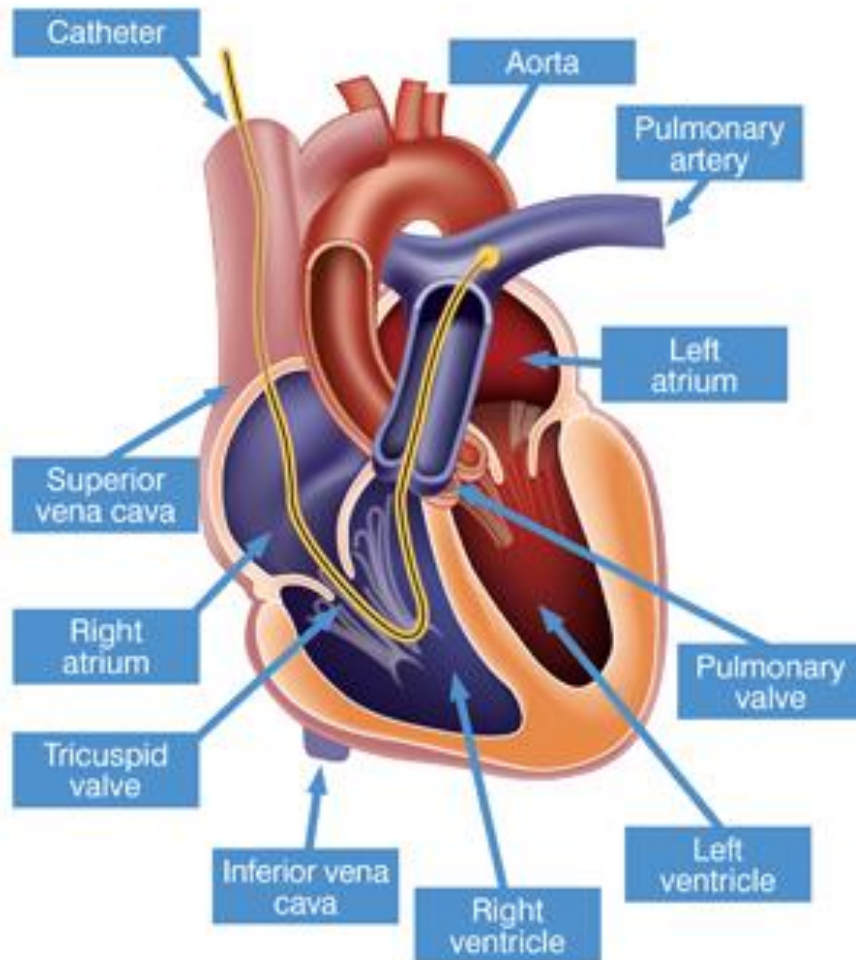
# RIGHT HEART CATHETERIZATION



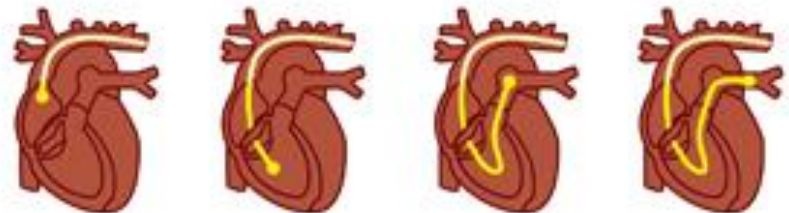
Characteristic intracardiac pressure waveforms during passage through the heart



# RIGHT HEART CATHETERIZATION



Characteristic intracardiac pressure waveforms during passage through the heart



# RIGHT HERAT CATHETERIZATION

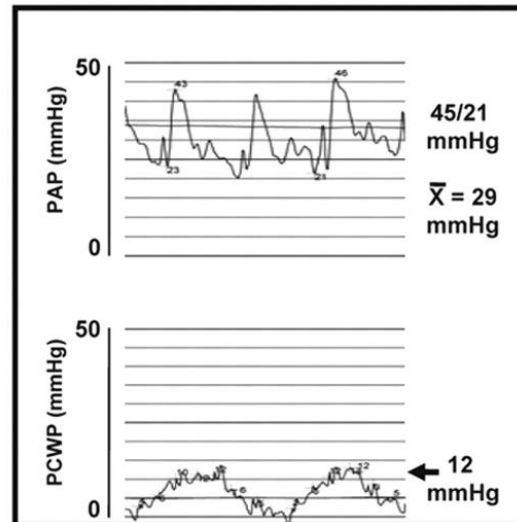
## Fluid Challenge

Obtain Baseline hemodynamic Profile

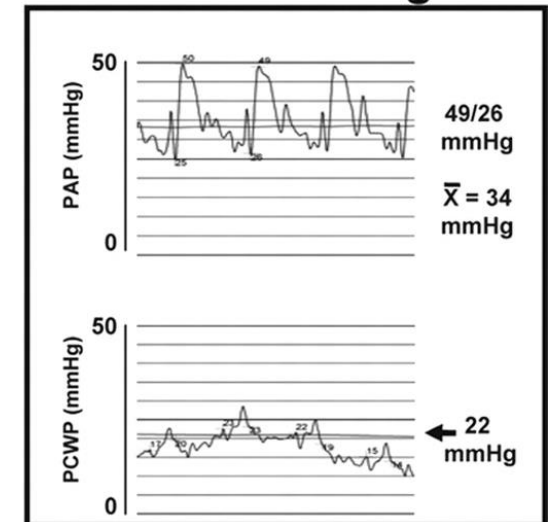
Administer 1000 cc 0.9% NaCl iv until completion over 20 minute period or until PCWP exceeds 15 mmHg with symptoms

Obtain hemodynamic measurements with every 250 cc volume

### Baseline

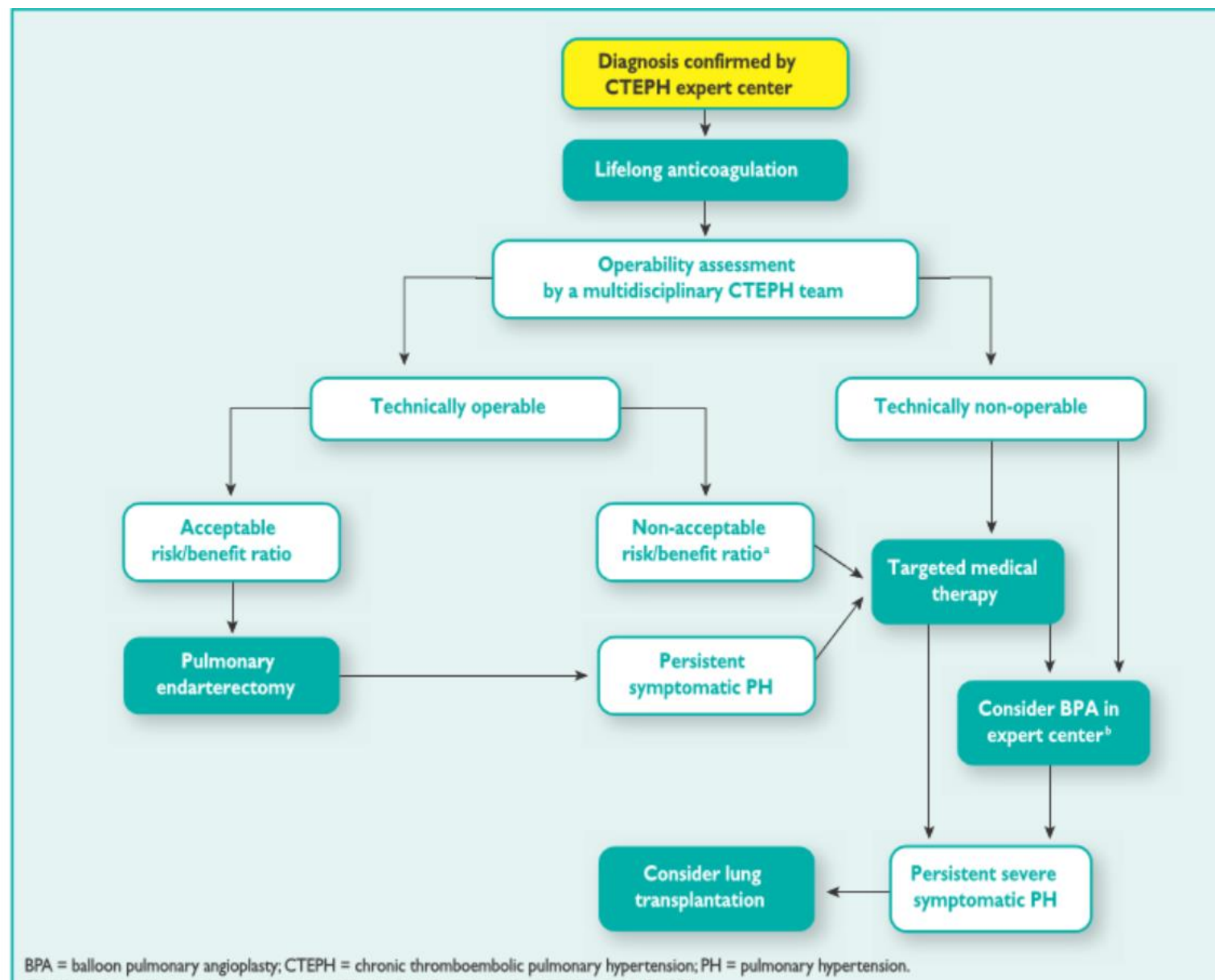


### Fluid Challenge



? 500cc 0.9% NaCl iv in 5 min ?

# CONTEMPORARY DIAGNOSTIC AND THERAPEUTIC ALGORITHM



BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic pulmonary hypertension; PH = pulmonary hypertension.

<sup>a</sup>Technically operable patients with non-acceptable risk/benefit ratio can be considered also for BPA.

<sup>b</sup>In some centers medical therapy and BPA are initiated concurrently.

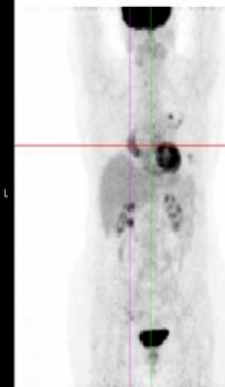
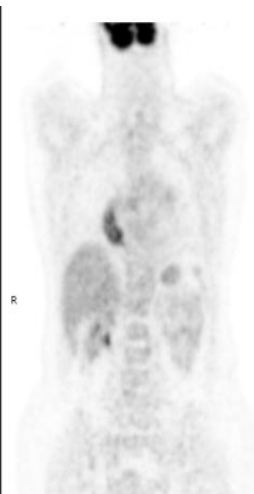
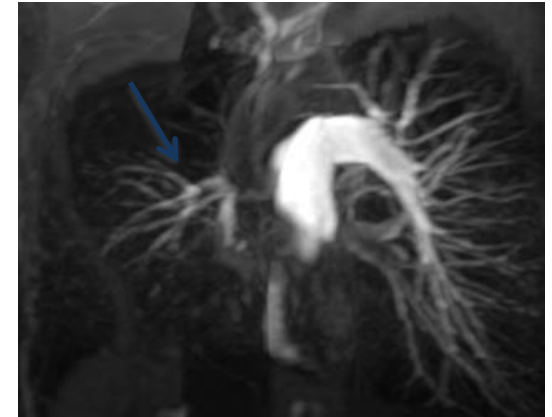
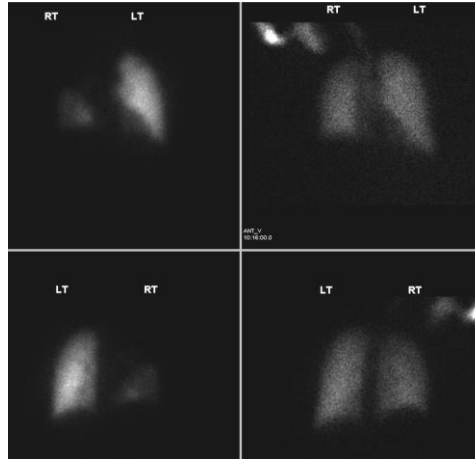
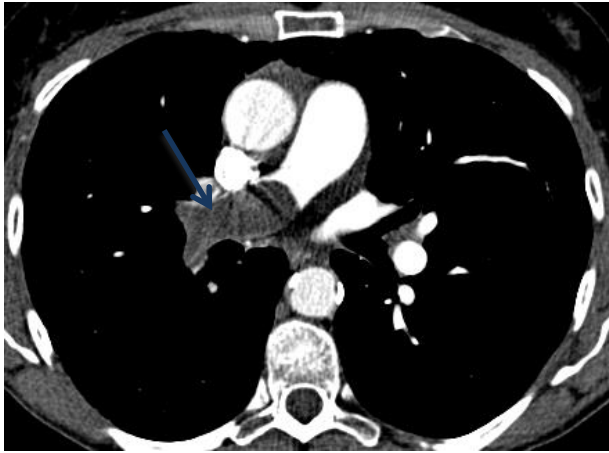
# **DIFFERENTIAL DIAGNOSIS**

- **Pulmonary Artery Sarcoma**
- **Tumor cell embolism**
- **Parasites (Hydatid cyst)**
- **Foreign body embolism**
- **Congenital or acquired PA stenosis**



# DIFFERENTIAL DIAGNOSIS

## Conditions That Mimic CTEPH *Pulmonary Artery Sarcoma*



# CTEPH PEA

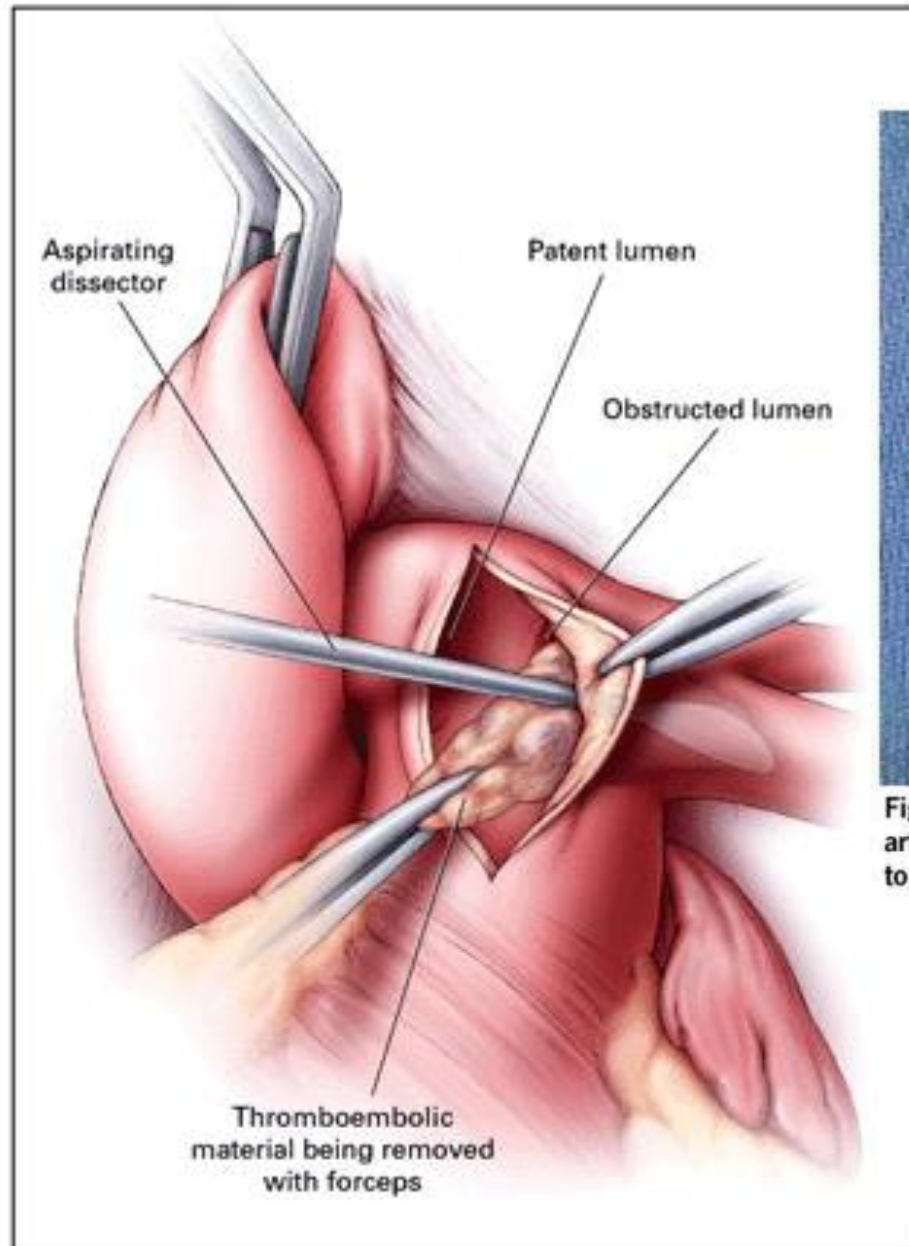
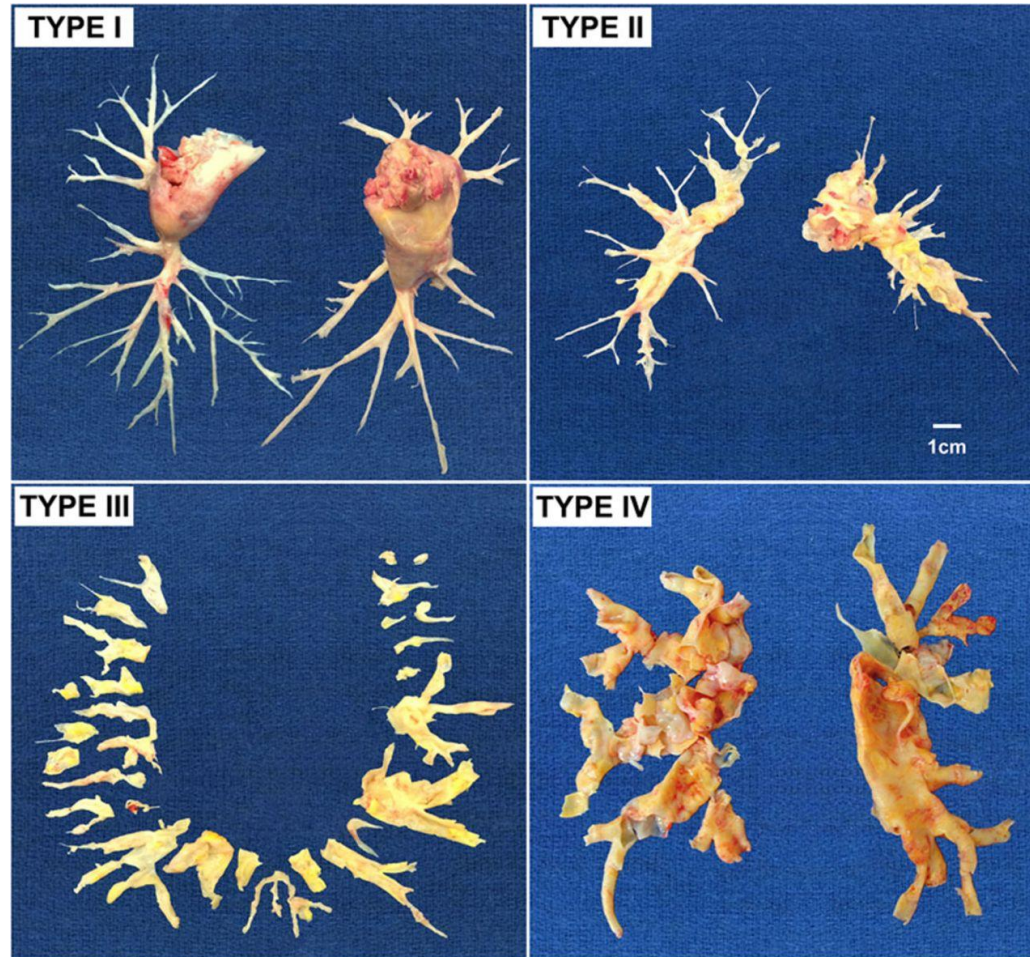


Fig. 2—Surgical specimen removed from right and left pulmonary arteries indicating type II disease. Note the extent of dissection down to the tail end of each branch. The ruler measures 6 inches.



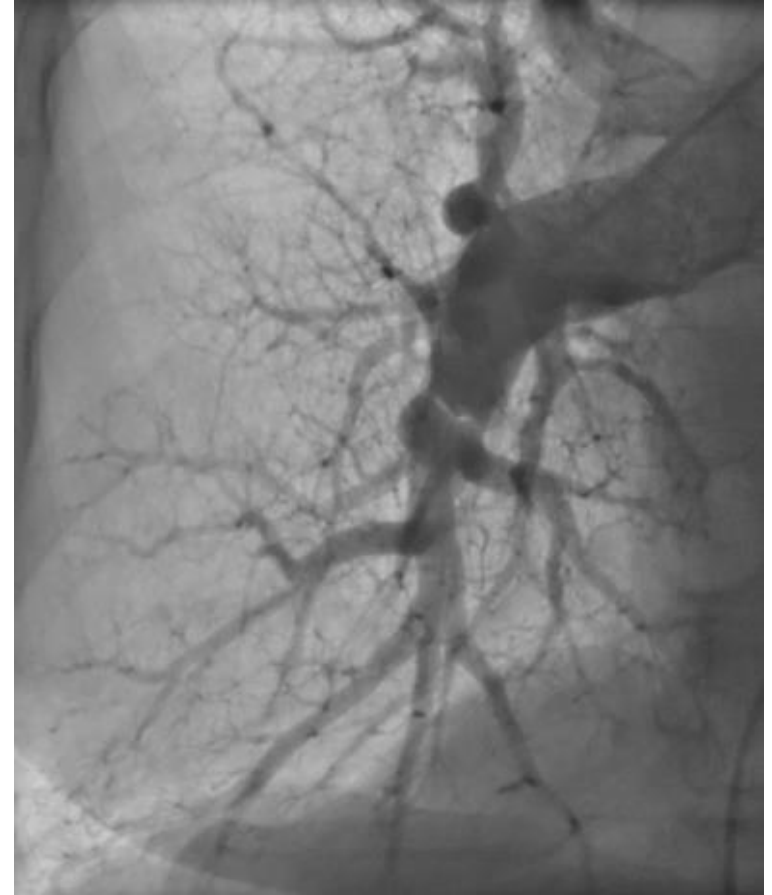
# CTEPH PEA

University of California–San Diego surgical classification (Jamieson) of pulmonary endarterectomy specimens.<sup>29</sup> Typical surgical specimens classified by the most proximal level of obstruction for types I through III are shown.

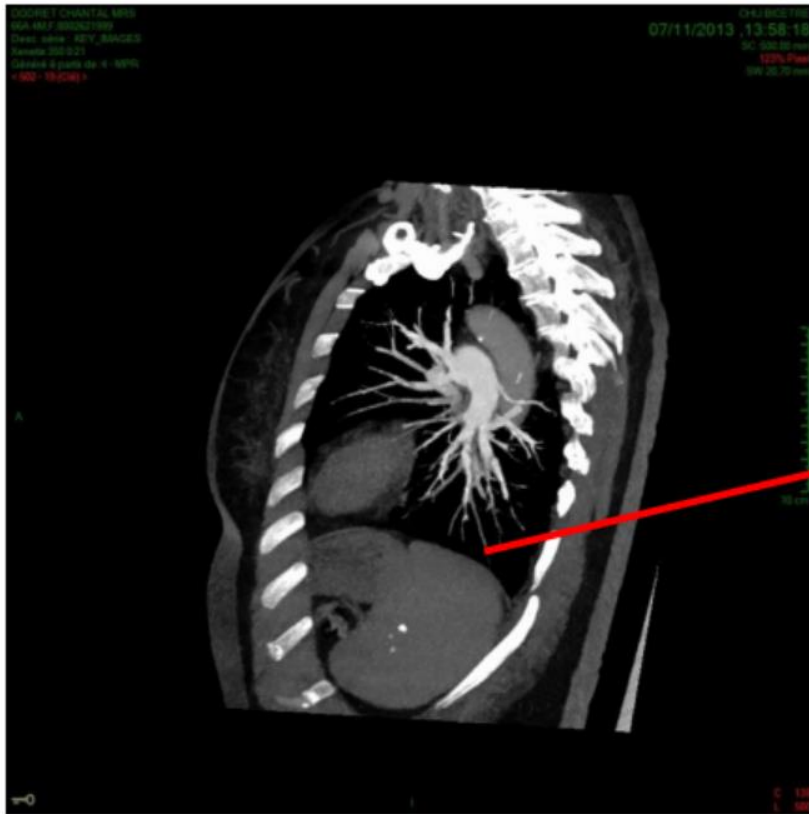


Lang I M , and Madani M Circulation. 2014;130:508-518

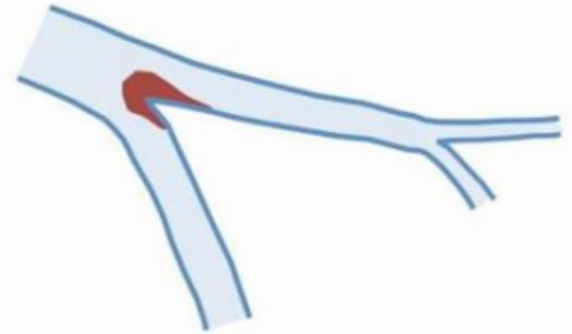
# CATHETER PULMONARY ANGIOGRAPHY



## CATHETER PULMONARY ANGIOGRAPHY

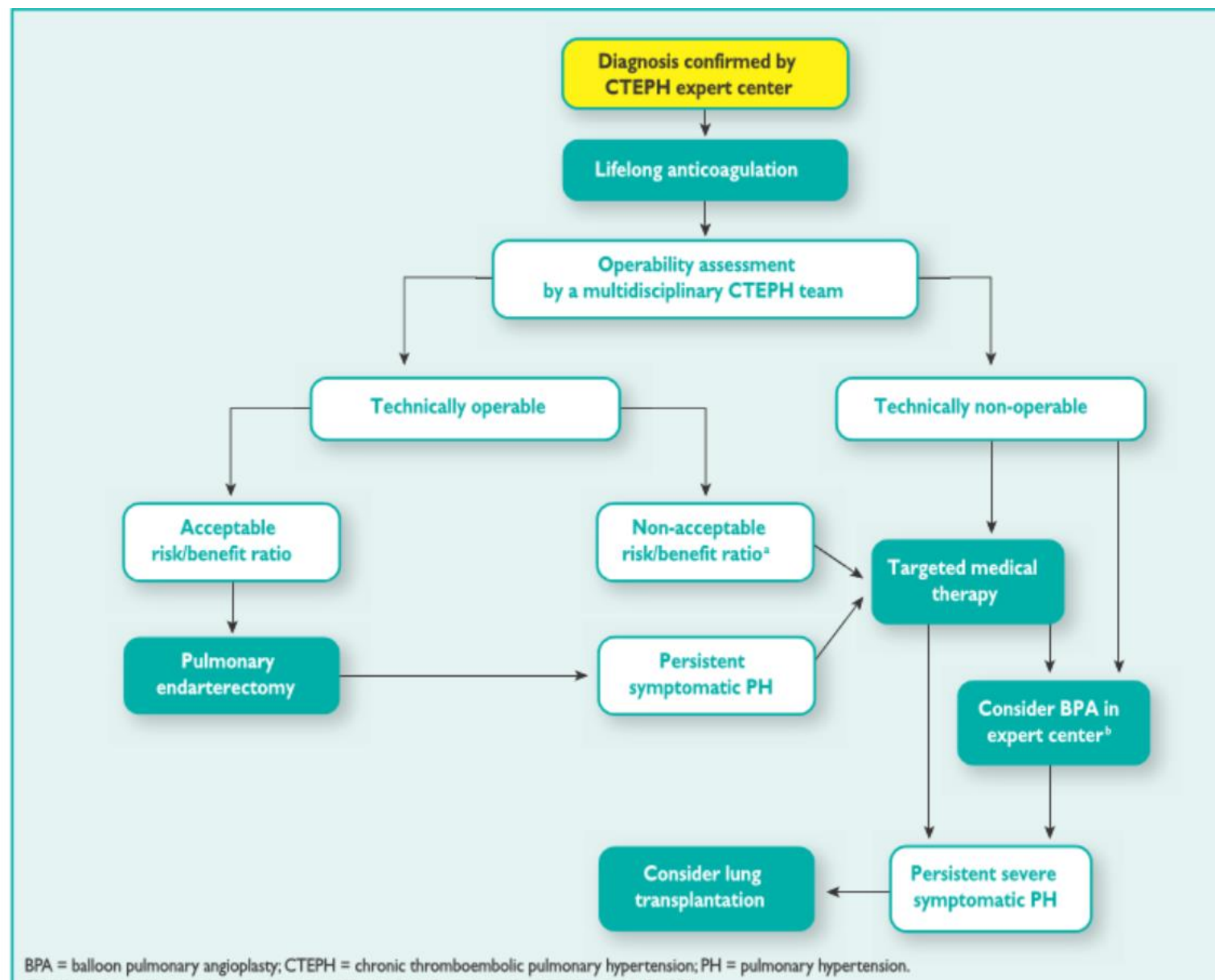


### Type 1: Webs



1st. Session in February 2014:  
Left pulmonary artery dilatation : A7 and A8.

# CONTEMPORARY DIAGNOSTIC AND THERAPEUTIC ALGORITHM

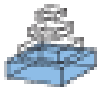


BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic pulmonary hypertension; PH = pulmonary hypertension.

<sup>a</sup>Technically operable patients with non-acceptable risk/benefit ratio can be considered also for BPA.

<sup>b</sup>In some centers medical therapy and BPA are initiated concurrently.

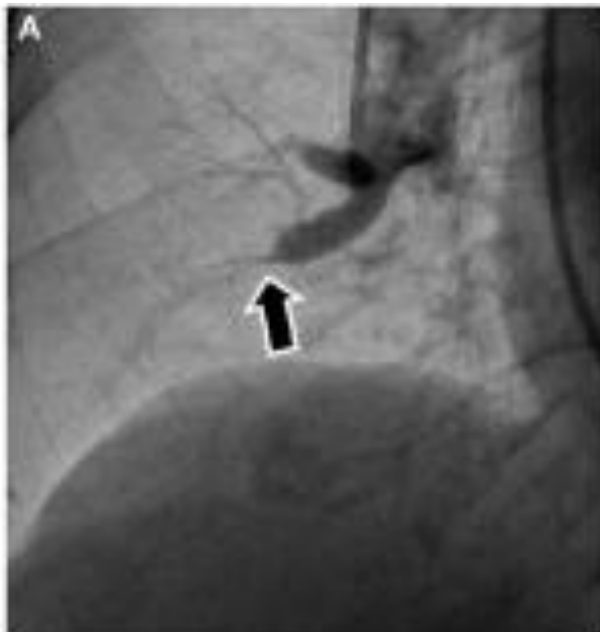




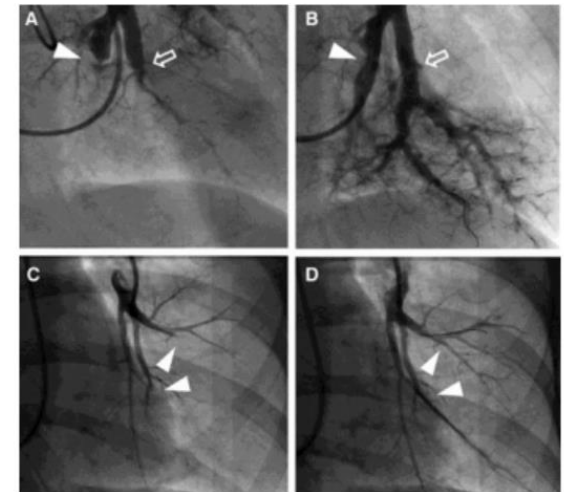
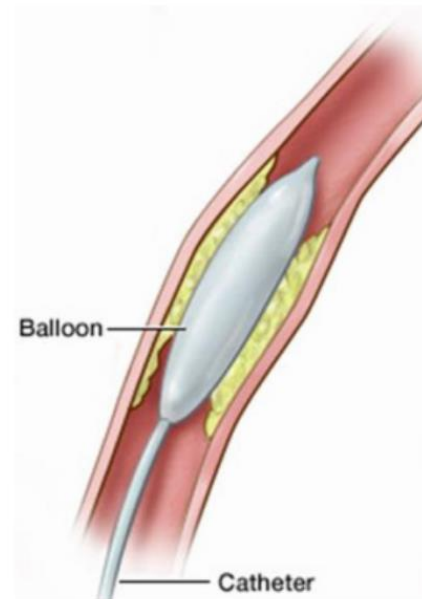
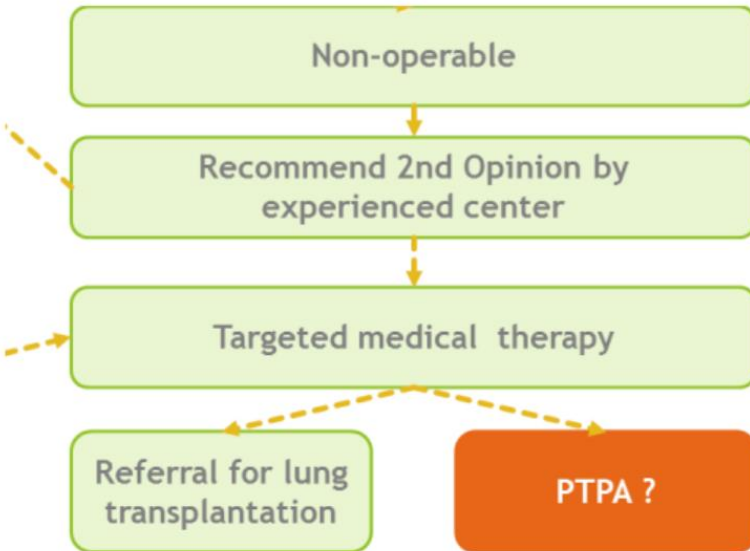
## Balloon pulmonary angioplasty: a treatment option for inoperable patients with chronic thromboembolic pulmonary hypertension

**Aiko Ogawa\* and Hiromi Matsubara**

*Department of Clinical Sciences, National Hospital Organization Okayama Medical Center, Okayama, Japan*



# BALLOON PULMONARY ANGIOPLASTY



Fukui et al. Right ventricular reverse remodelling after balloon pulmonary angioplasty.  
Eur Respir J 2014

[Circ J](#). 2016 Mar 25;80(4):980-8. doi: 10.1253/circj.CJ-15-1212. Epub 2016 Feb 24.

**Multiple Beneficial Effects of Balloon Pulmonary Angioplasty in Patients With Chronic Thromboembolic Pulmonary Hypertension.**

[Tatebe S](#)<sup>1</sup>, [Sugimura K](#), [Aoki T](#), [Miura M](#), [Nochioka K](#), [Yaoita N](#), [Suzuki H](#), [Sato H](#), [Yamamoto S](#), [Satoh K](#), [Fukumoto Y](#), [Shimokawa H](#).

### ***Underdiagnosed and Treatable***

- **CTEPH is more common than people realize.**
  - **It is hard for general physicians to make the diagnosis because many of the symptoms are nonspecific, such as fatigue and breathlessness on exertion.**
- **It is one of the most treatable forms of PH.**
  - **There is a curative treatment available in the form of PEA.**



**END**