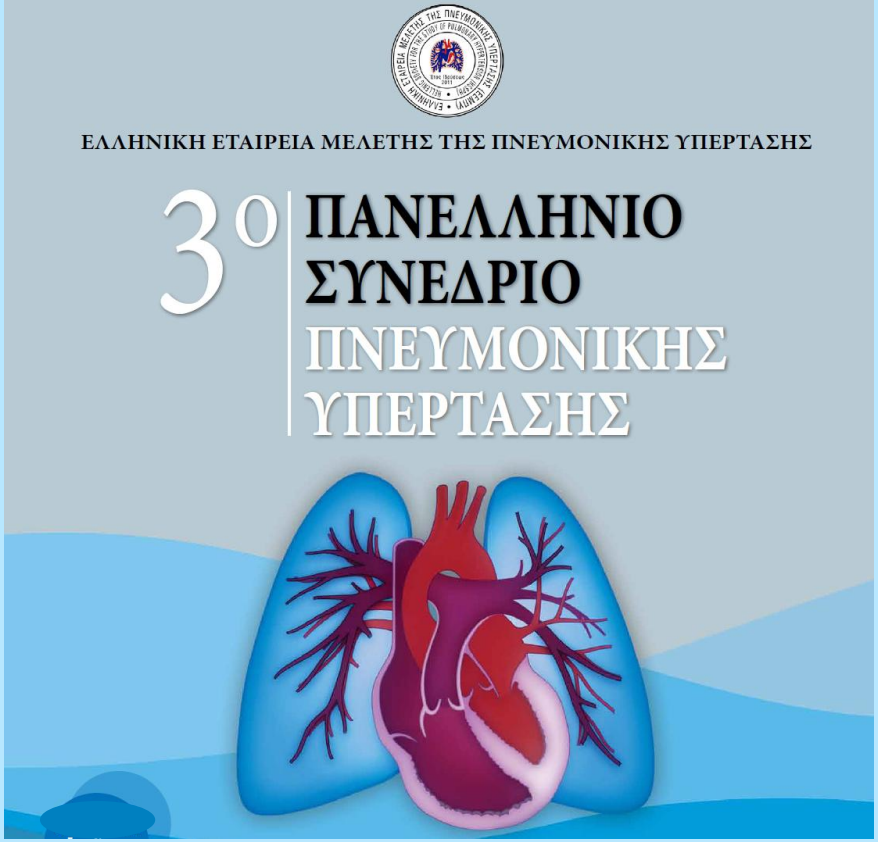

Τι άλλαξε στο 6ο Παγκόσμιο Συνέδριο στη διάγνωση της Πνευμονικής Υπέρτασης

Αναστασία Ανθη

Β΄ Κλινική Εντατικής Θεραπείας & Διακλινικό Ιατρείο Πνευμονικής Υπέρτασης
Π.Γ.Ν. «ΑΤΤΙΚΟΝ»

Classification of Pulmonary Hypertension

- **Geneva**, 1973: primary/ secondary pulmonary hypertension
  1st world symposium on PH

- **Evian**, 1998:
  2nd world symposium on PH *5 groups*

- **Venice**, 2003:
  3rd world symposium on PH »

- **Dana Point**, 2008:
  4th world symposium on PH »

- **Nice**, 2013:
  5th world symposium on PH »

- **Nice**, 2018:
  6th world symposium on PH »

Clinical classification of Pulmonary Hypertension (ESC/ERS guidelines 2015)

1. Pulmonary arterial hypertension

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2 mutation
  - 1.2.2 Other mutations
- 1.3 Drugs and toxins induced
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 Human immunodeficiency virus (HIV) infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease (Table 6)
  - 1.4.5 Schistosomiasis

1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

- 1'.1 Idiopathic
- 1'.2 Heritable
  - 1'.2.1 EIF2AK4 mutation
  - 1'.2.2 Other mutations
- 1'.3 Drugs, toxins and radiation induced
- 1'.4 Associated with:
  - 1'.4.1 Connective tissue disease
  - 1'.4.2 HIV infection

1''. Persistent pulmonary hypertension of the newborn

2. Pulmonary hypertension due to left heart disease

- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital / acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
- 2.5 Congenital /acquired pulmonary veins stenosis

3. Pulmonary hypertension due to lung diseases and/or hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental lung diseases (Web Table III)

4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions

- 4.1 Chronic thromboembolic pulmonary hypertension
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Angiosarcoma
  - 4.2.2 Other intravascular tumors
  - 4.2.3 Arteritis
  - 4.2.4 Congenital pulmonary arteries stenoses
  - 4.2.5 Parasites (hydatidosis)

5. Pulmonary hypertension with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders, sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: pulmonary tumoral thrombothic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension

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Categorization of multiple clinical conditions into 5 groups according to their similar:

- **clinical presentation**
- **pathological findings**
- **haemodynamic characteristics &**
- **treatment strategy**

Updated clinical classification of pulmonary hypertension

1 PAH
- 1.1 Idiopathic PAH
- 1.2 Heritable PAH
- 1.3 Drug- and toxin-induced PAH (table 3)
- 1.4 PAH associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
- 1.5 PAH long-term responders to calcium channel blockers (table 4)
- 1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement (table 5)
- 1.7 Persistent PH of the newborn syndrome

2 PH due to left heart disease
- 2.1 PH due to heart failure with preserved LVEF
- 2.2 PH due to heart failure with reduced LVEF
- 2.3 Valvular heart disease
- 2.4 Congenital/acquired cardiovascular conditions leading to post-capillary PH

3 PH due to lung diseases and/or hypoxia
- 3.1 Obstructive lung disease
- 3.2 Restrictive lung disease
- 3.3 Other lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoxia without lung disease
- 3.5 Developmental lung disorders

4 PH due to pulmonary artery obstructions (table 6)
- 4.1 Chronic thromboembolic PH
- 4.2 Other pulmonary artery obstructions

5 PH with unclear and/or multifactorial mechanisms (table 7)
- 5.1 Haematological disorders
- 5.2 Systemic and metabolic disorders
- 5.3 Others
- 5.4 Complex congenital heart disease

"Proceedings of the 6th World Symposium on PH" ***Eur Respir J* 2018**

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"Proceedings of the 6th World Symposium on PH" ***Eur Respir J*** **2018**

Table 3 Haemodynamic definitions of pulmonary hypertension[a]

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

[a]All values measured at rest.
[b]According to *Table 4*.
[c]High CO can be present in cases of hyperkinetic conditions such as systemic-to-pulmonary shunts (only in the pulmonary circulation), anaemia, hyperthyroidism, etc.
CO = cardiac output; PAP = pulmonary arterial pressure; PH = pulmonary hypertension; PWP = pulmonary wedge pressure; TPG = transpulmonary pressure gradient (mean PAP – mean PWP).

ESC/ ERS guidelines 2009

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Isolated post-capillary PH (Ipc-PH)	DPG <7 mmHg and/or PVR ≤3 WU[c]	
Combined post-capillary and pre-capillary PH (Cpc-PH)	DPG ≥7 mmHg and/or PVR >3 WU[c]	

PH guidelines 2015

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Haemodynamic definition of Pulmonary Hypertension

Definition	Characteristics[a]	Clinical group(s)[b]
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Pre-capillary PH	PAPm ≥ 25 mm Hg PAWP ≤ 15 mm Hg **PVR > 3 Wood Units**	**Pulmonary Arterial Hypertension**
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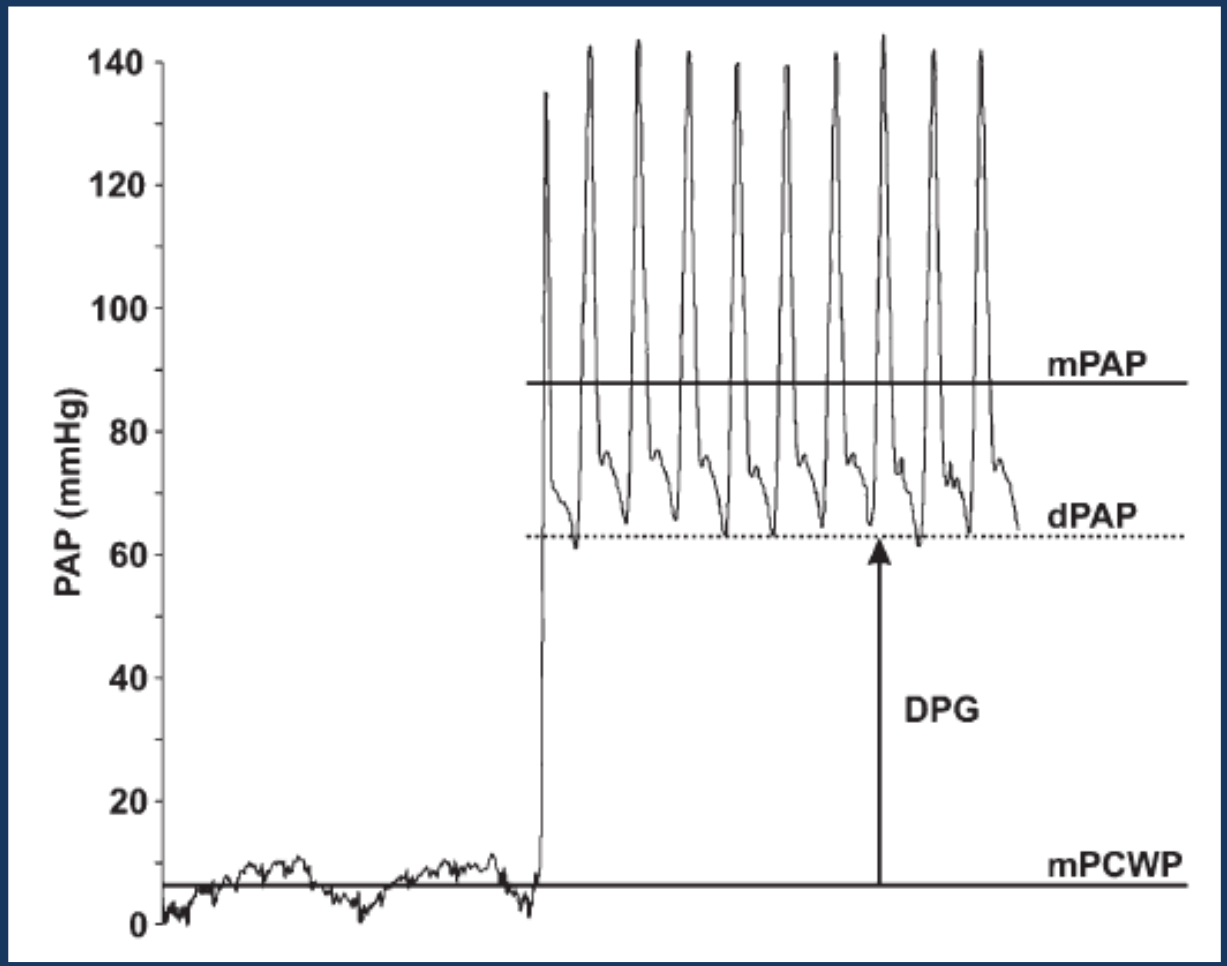
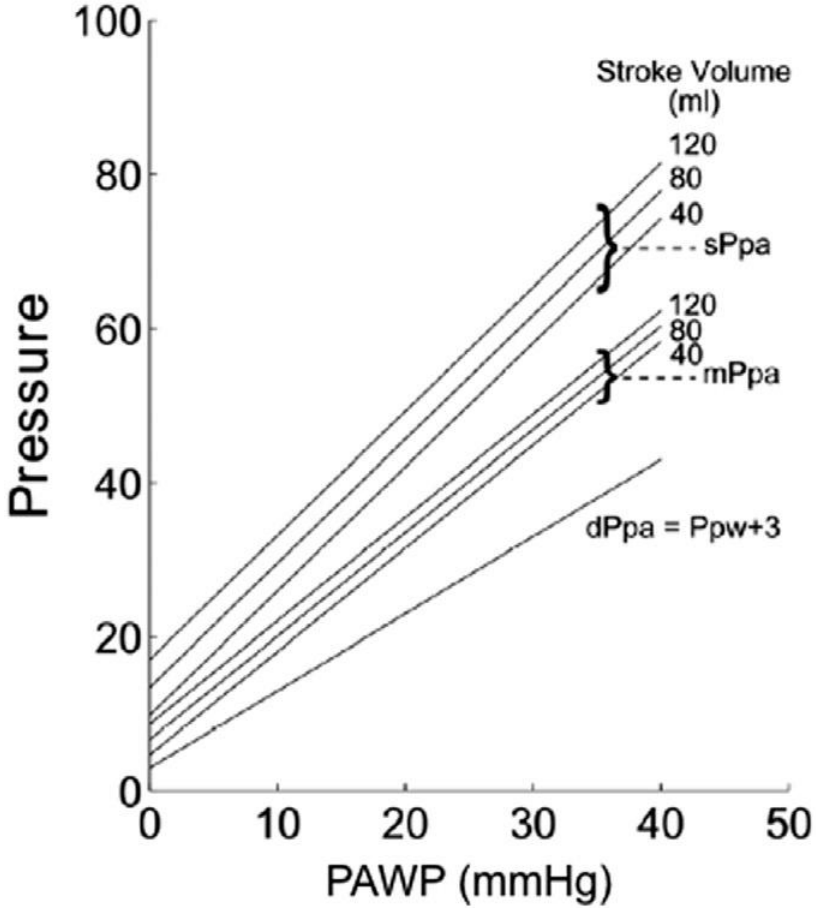
DPG

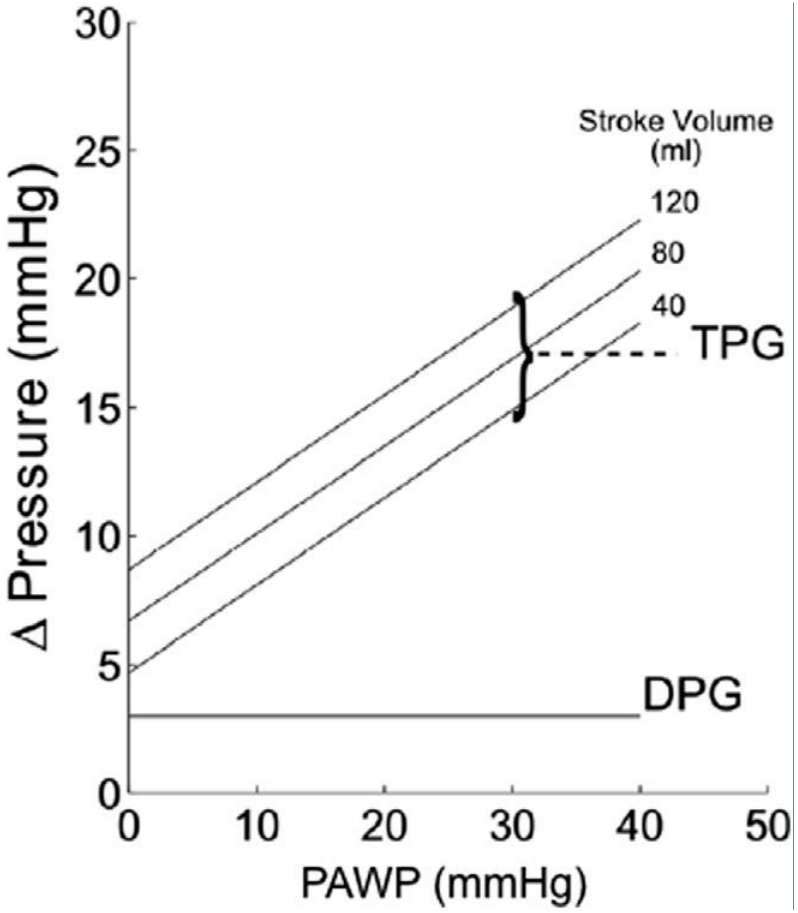
PH guidelines 2015

TPG (transpulmonary pressure gradient): mPAP - PAWP

DPD (diastolic pressure difference): diastolic PAP- PAWP




Pressure
100
80
60
40
20
0
0
10
20
30
40
50
PAWP (mmHg)
Stroke Volume
(ml)
120
80
40
sPpa
120
80
40
mPpa
dPpa = Ppw+3


Δ Pressure (mmHg)
30
25
20
15
10
5
0
0
10
20
30
40
50
PAWP (mmHg)
Stroke Volume
(ml)
120
80
40
TPG
DPG

Haemodynamic definitions of pulmonary hypertension

Definitions	Characteristics	Clinical groups[#]
Pre-capillary PH	mPAP >20 mmHg PAWP ⩽15 mmHg PVR ⩾3 WU	1, 3, 4 and 5
Isolated post-capillary PH (IpcPH)	mPAP >20 mmHg PAWP >15 mmHg PVR <3 WU	2 and 5
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"Proceedings of the 6th World Symposium on PH" ***Eur Respir J*** **2018**

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Pulmonary arterial pressure during rest & exercise in healthy subjects: a systematic review

TABLE 1 Haemodynamics at rest (supine)

$\bar{P}_{pa}$ mmHg	14.0±3.3
Systolic P_{pa} mmHg	20.8±4.4
Diastolic P_{pa} mmHg	8.8±3.0
P_{paw} mmHg	8.0±2.9
Heart rate min^{-1}	76±14
Cardiac output $L \cdot min^{-1}$	7.3±2.3
Cardiac index $L \cdot min^{-1} \cdot m^{-2}$	4.1±1.3
PVR $dyn \cdot s \cdot cm^{-5}$	74±30

Data are presented as weighted mean±weighted SD. $\bar{P}_{pa}$: mean pulmonary arterial pressure; P_{pa}: pulmonary arterial pressure; P_{paw}: pulmonary arterial wedge pressure; PVR: pulmonary vascular resistance. n=882 healthy volunteers.

Eur Respir J 2009; 34: 888–894

Kovacs and Olschewski *Arthritis Research & Therapy* (2015) 17:123
DOI 10.1186/s13075-015-0649-7

arthritis research&therapy

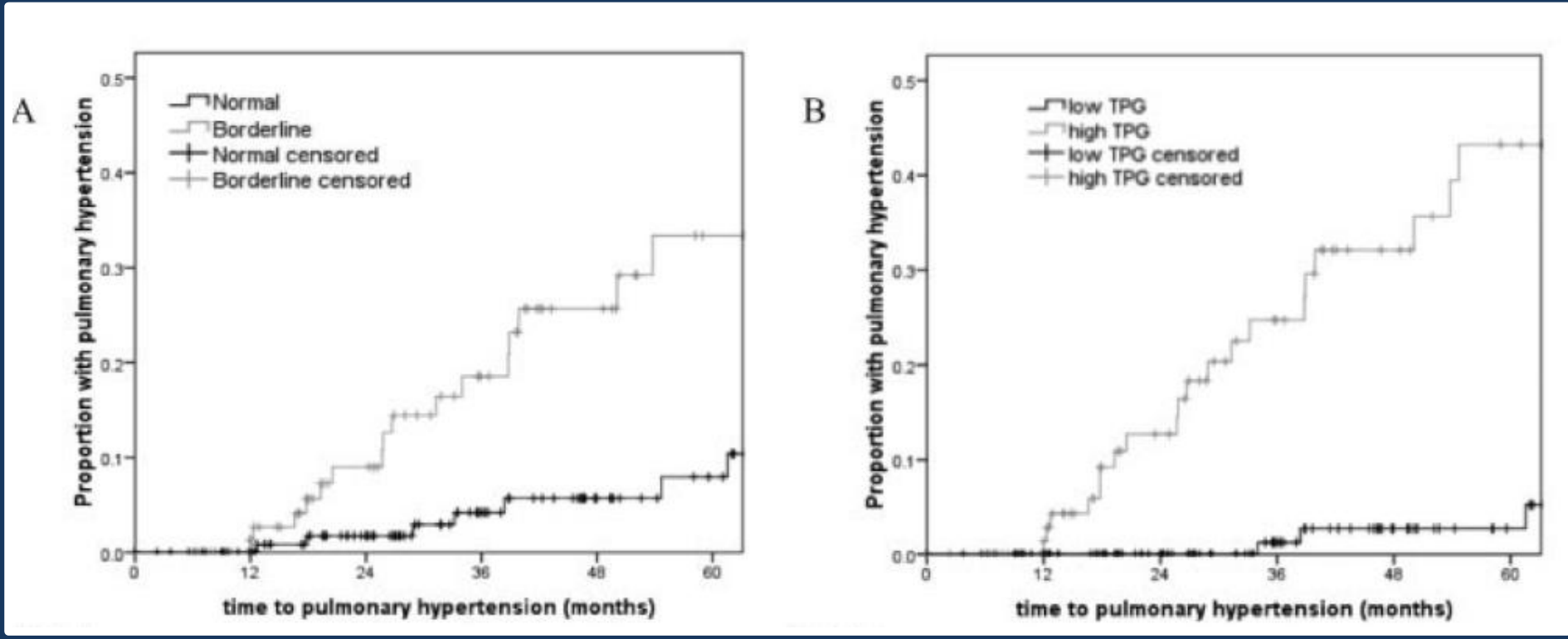
EDITORIAL Open Access

Borderline pulmonary pressures in scleroderma - a 'pre-pulmonary arterial hypertension' condition?

Gabor Kovacs[1,2*] and Horst Olschewski[1,2]

Borderline Mean Pulmonary Artery Pressure in Patients With Systemic Sclerosis

Transpulmonary Gradient Predicts Risk of Developing Pulmonary Hypertension



Valerio CJ et al. ARTHRITIS & RHEUMATISM 2013; 65(4): 1074-84

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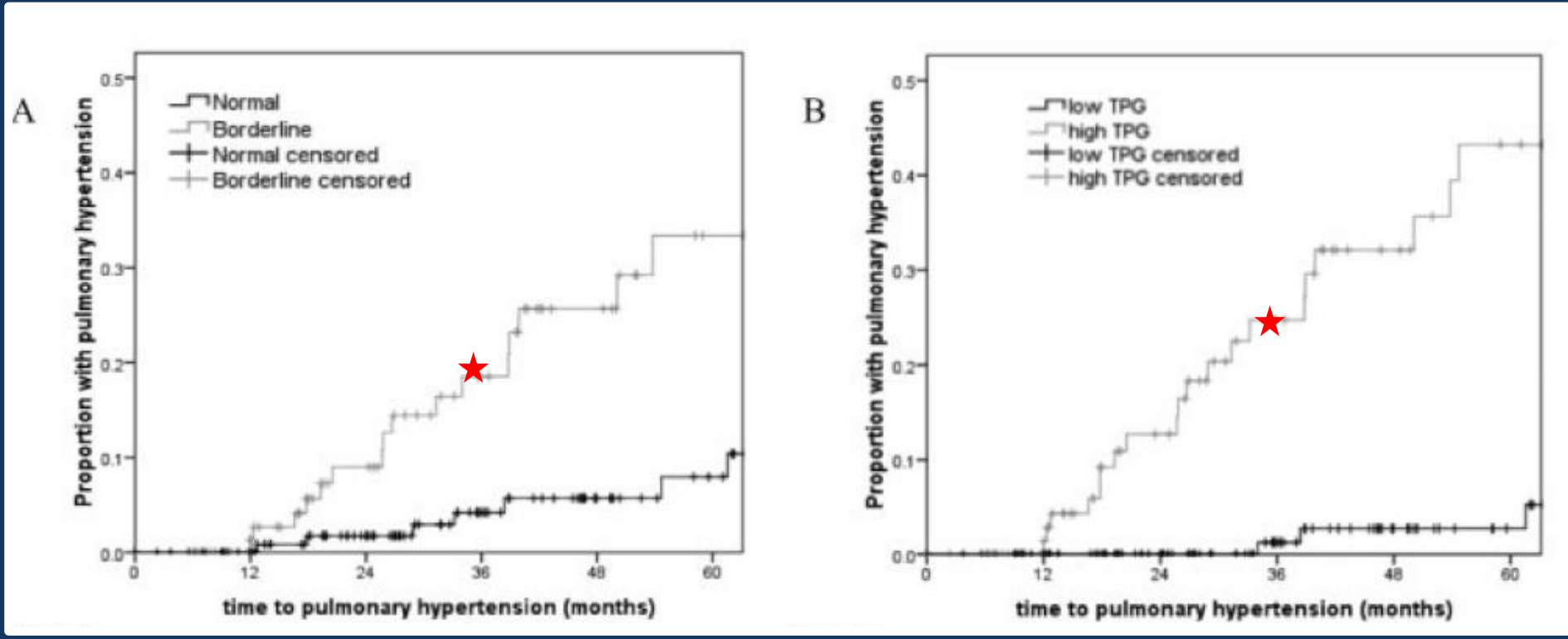

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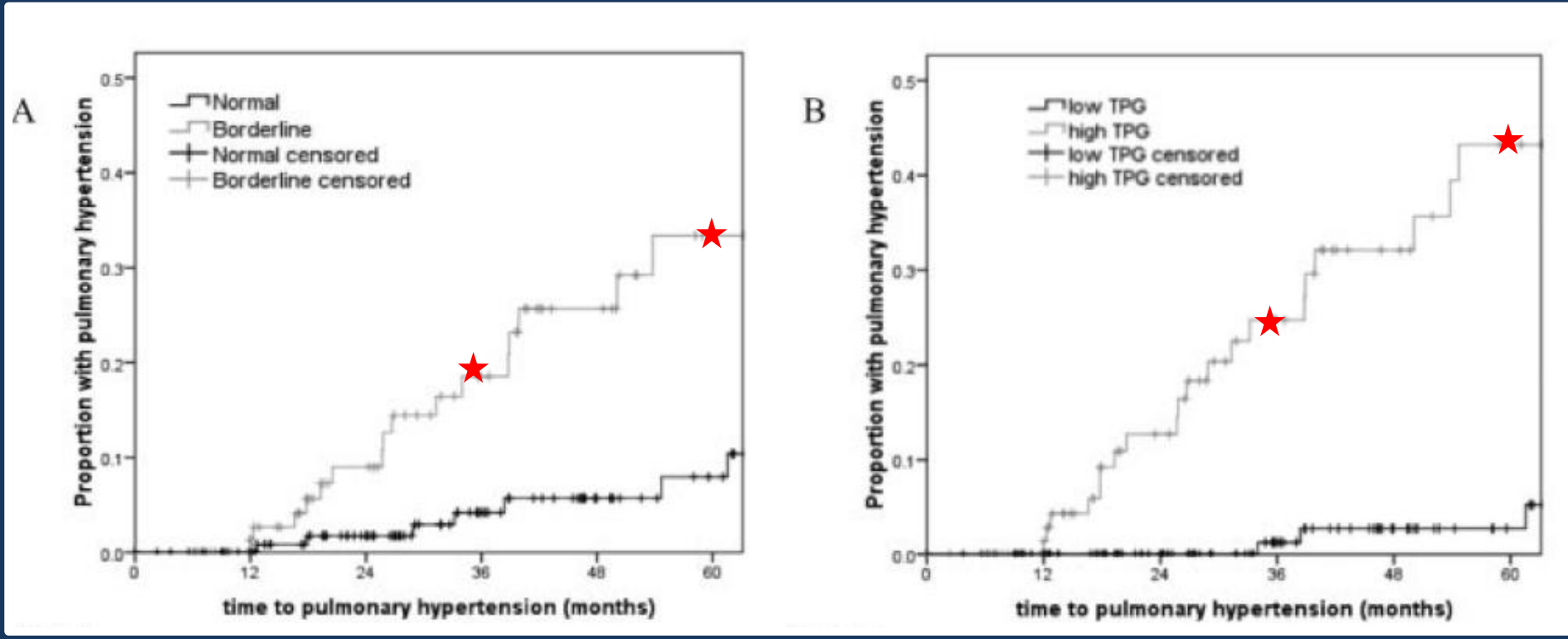
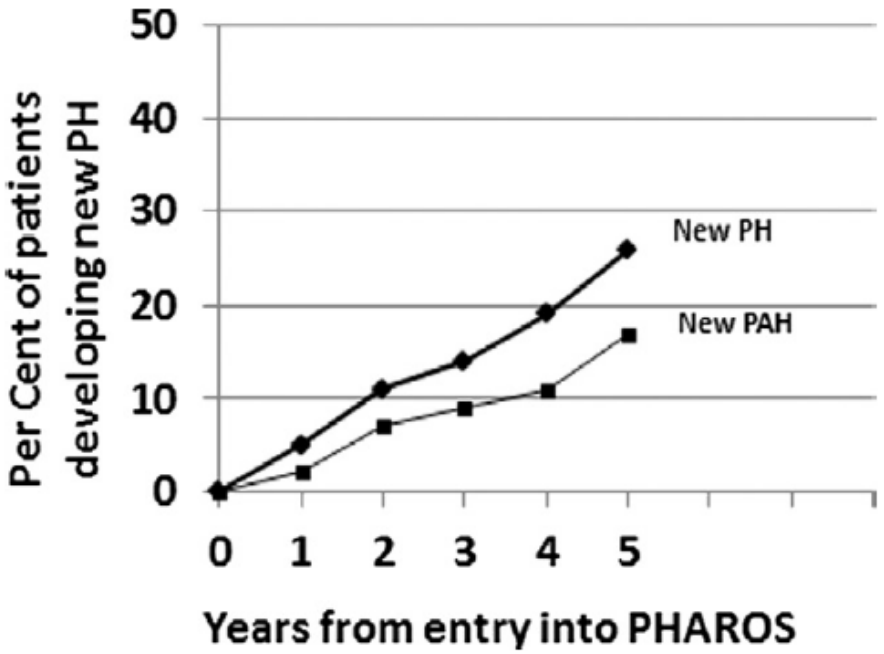


Valerio CJ et al. ARTHRITIS & RHEUMATISM 2013; 65(4): 1074-84

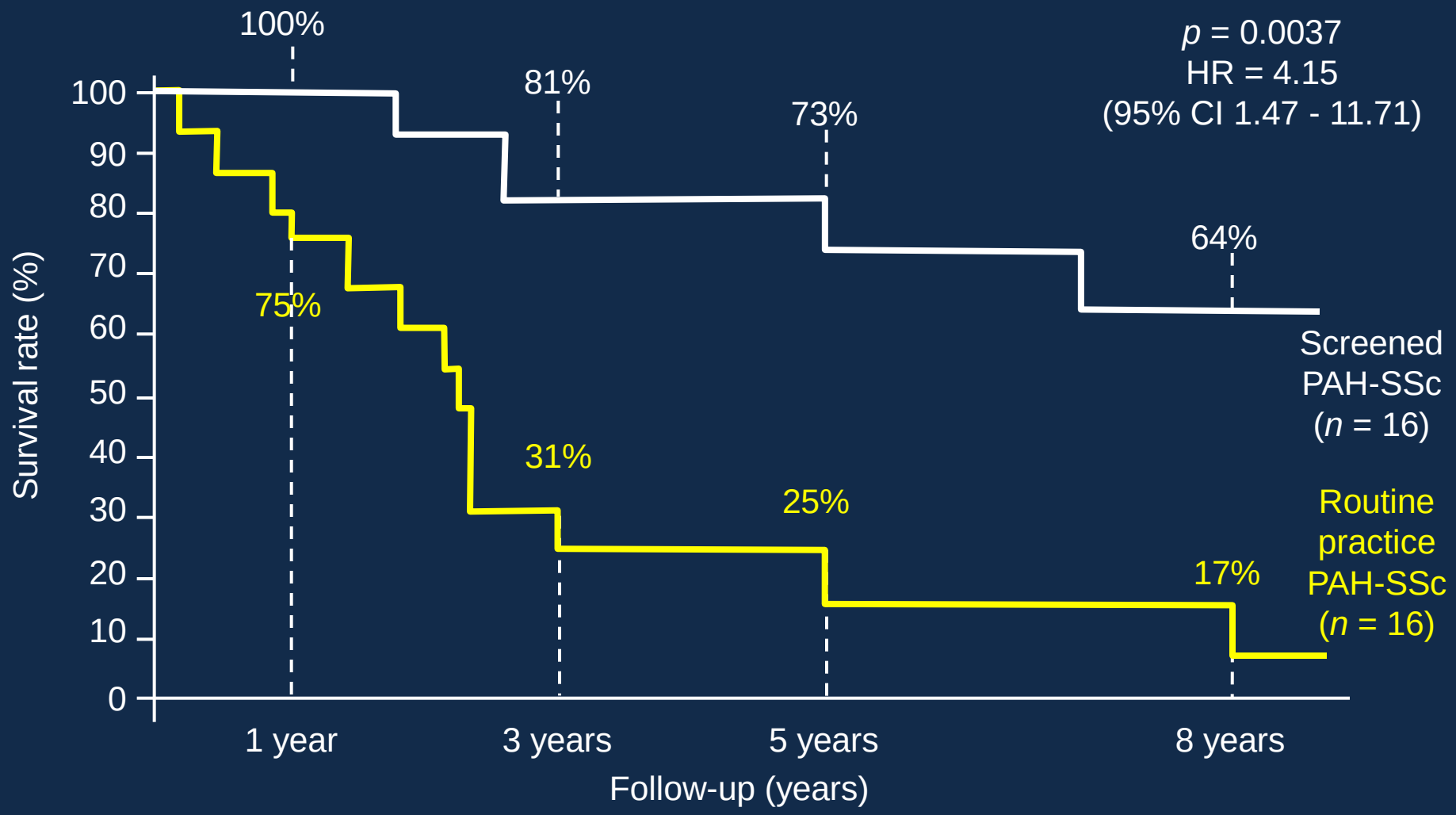
Development of pulmonary hypertension in a high-risk population with systemic sclerosis in the Pulmonary Hypertension Assessment and Recognition of Outcomes in Scleroderma (PHAROS) cohort study



Pts who are at high risk for PH, actually do develop definite PH during the 6-year follow-up time period and this risk increased with time

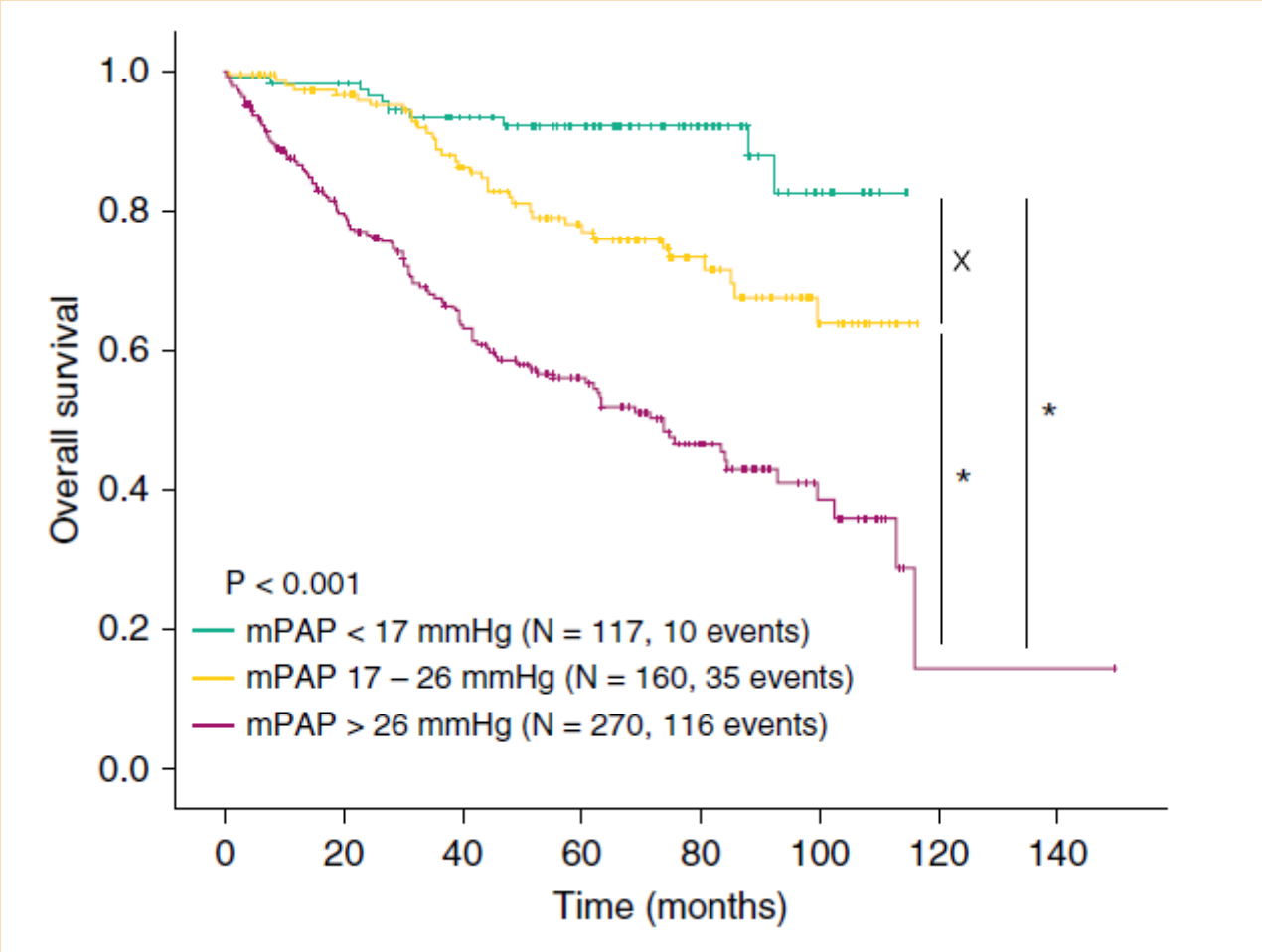
Seminars in Arthritis and Rheumatism 44 (2014) 55–62

Screening improves long-term outcomes in SSc patients



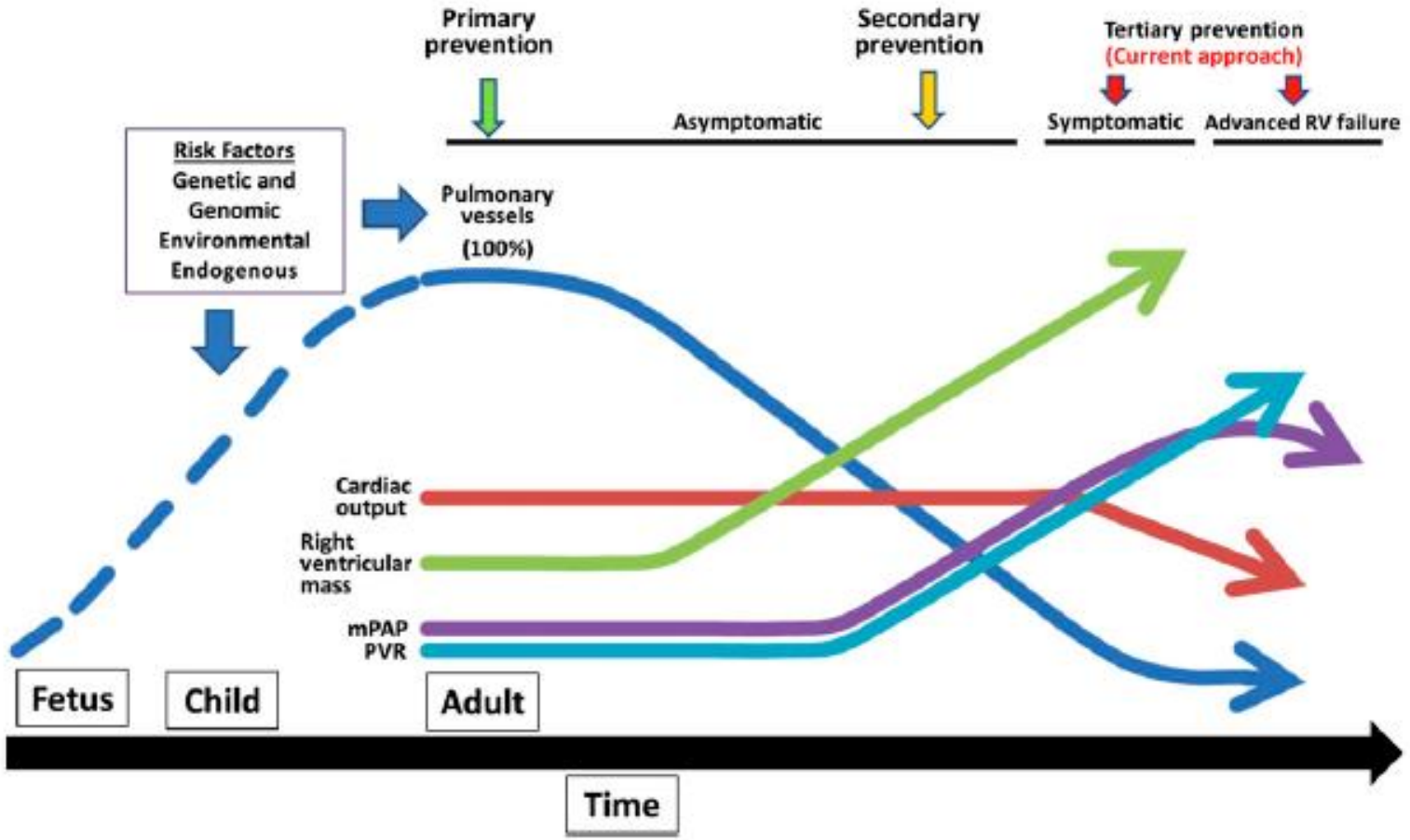
Humbert M, *et al. Arthritis Rheum* 2011; 63:3522-30.

Mild Elevation of Pulmonary Arterial Pressure as a Predictor of Mortality



Am J Respir Crit Care Med Vol 197(4): 509–516, 2018

State of the Science in PAH



Ann Am Thorac Soc Vol 11, Supplement 3, pp S178–S185, Apr 2014

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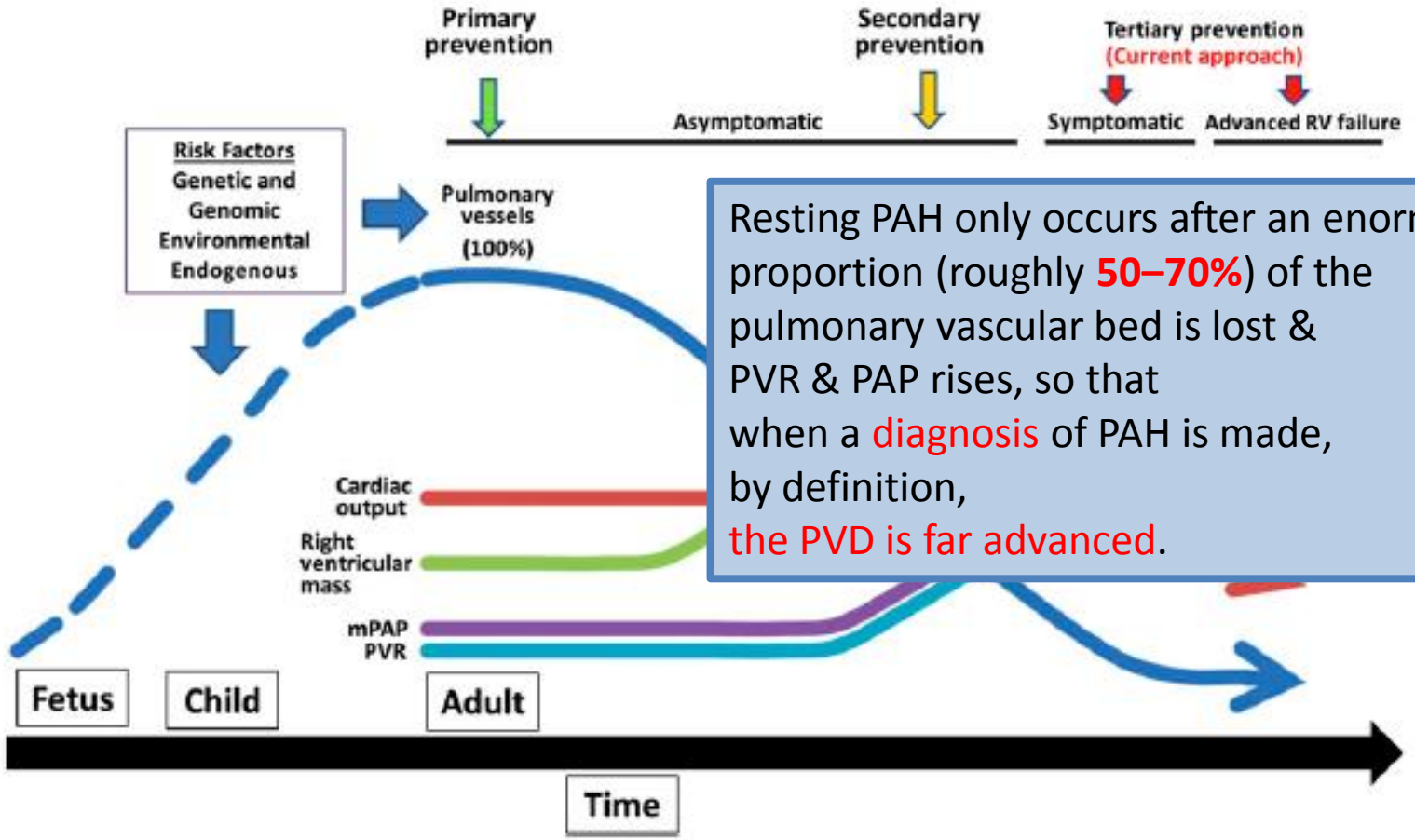
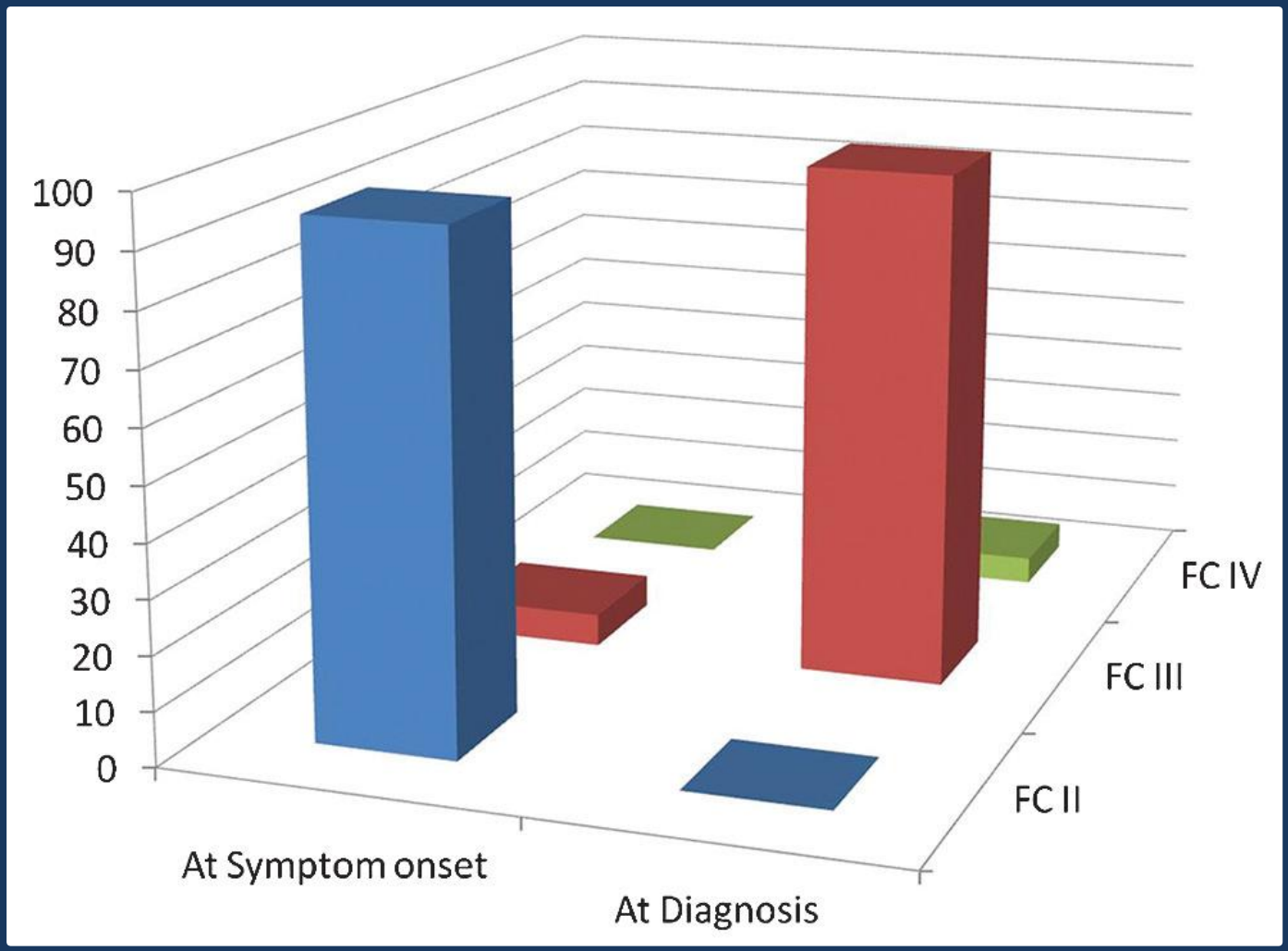


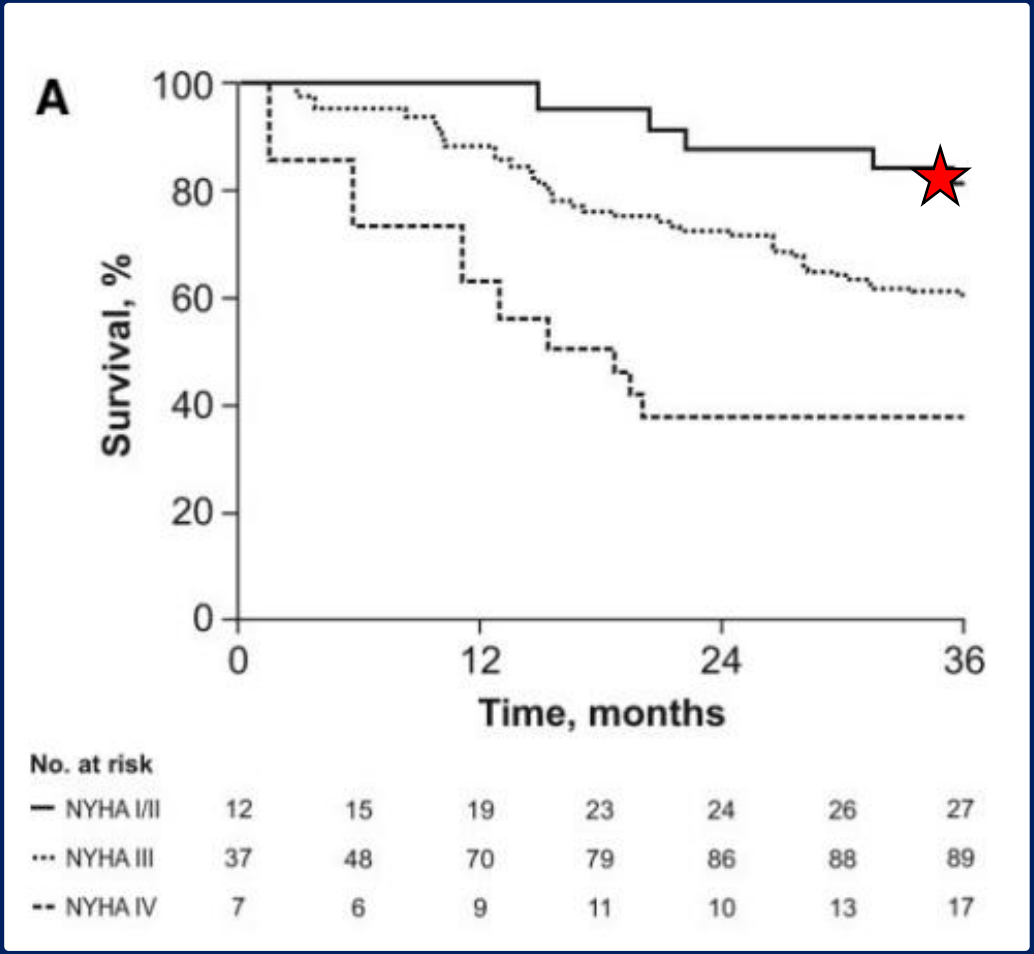
Resting PAH only occurs after an enormous proportion (roughly **50–70%**) of the pulmonary vascular bed is lost & PVR & PAP rises, so that when a diagnosis of PAH is made, by definition, the PVD is far advanced.

Ann Am Thorac Soc Vol 11, Supplement 3, pp S178–S185, Apr 2014



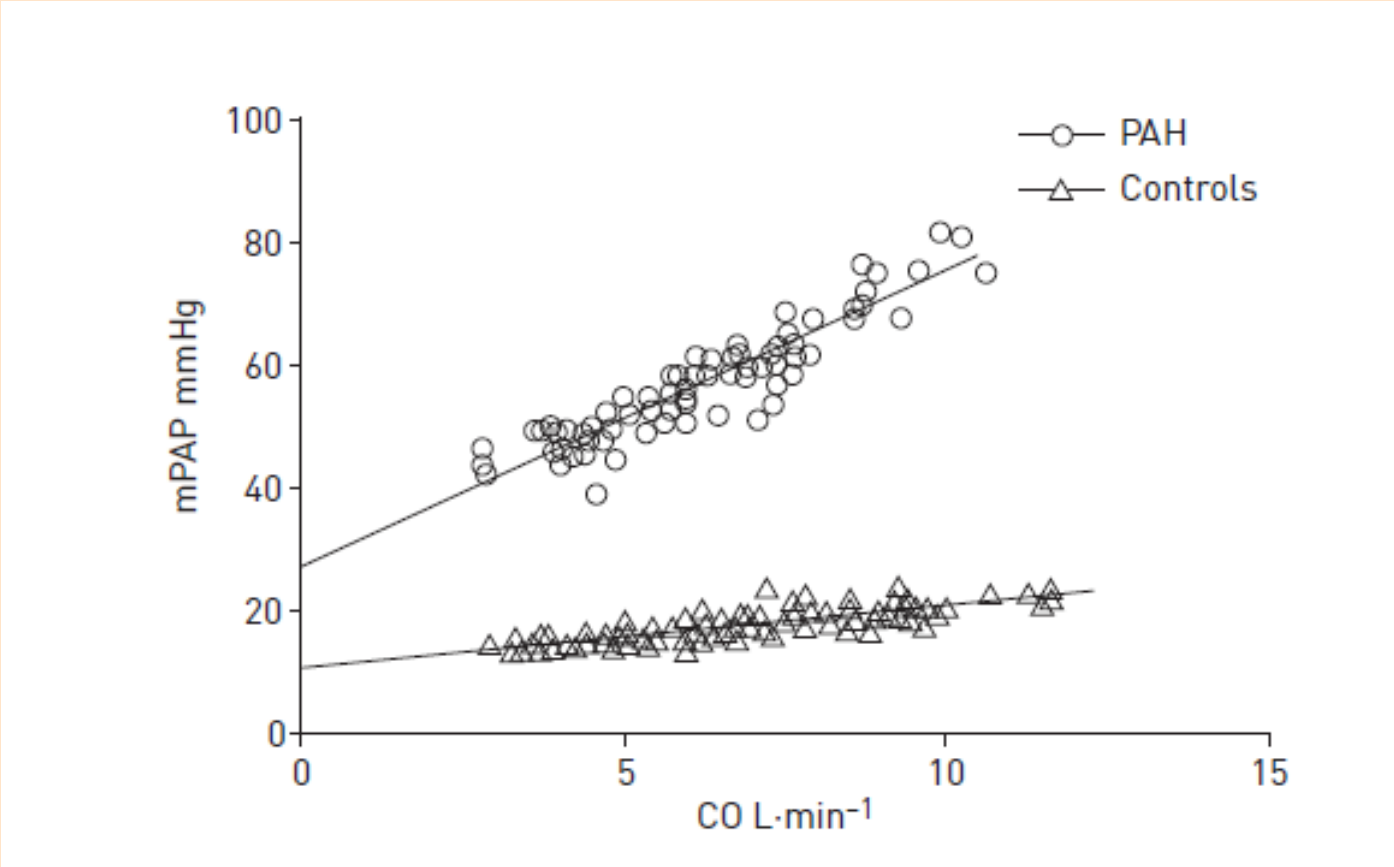
Strange et al.: Pulmonary Circulation, January-March 2013

In the modern management era, pulmonary arterial hypertension remains a progressive, fatal disease



Humbert M et al. Circulation 2010;122:156-163

American Heart Association
Learn and Live

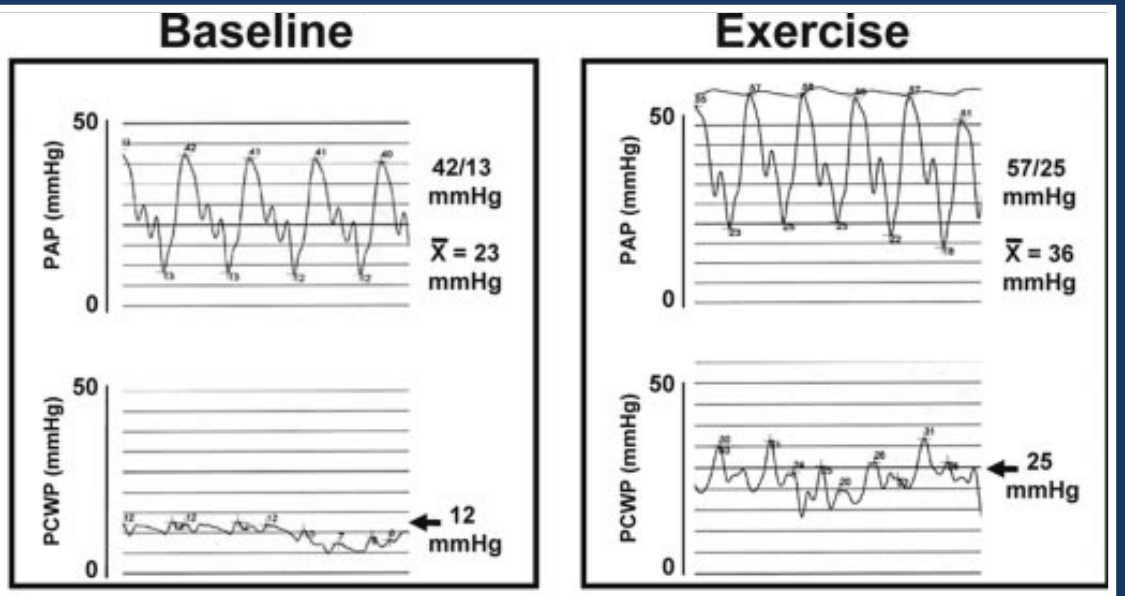


"Proceedings of the 6th World Symposium on PH" ***Eur Respir J*** **2019**

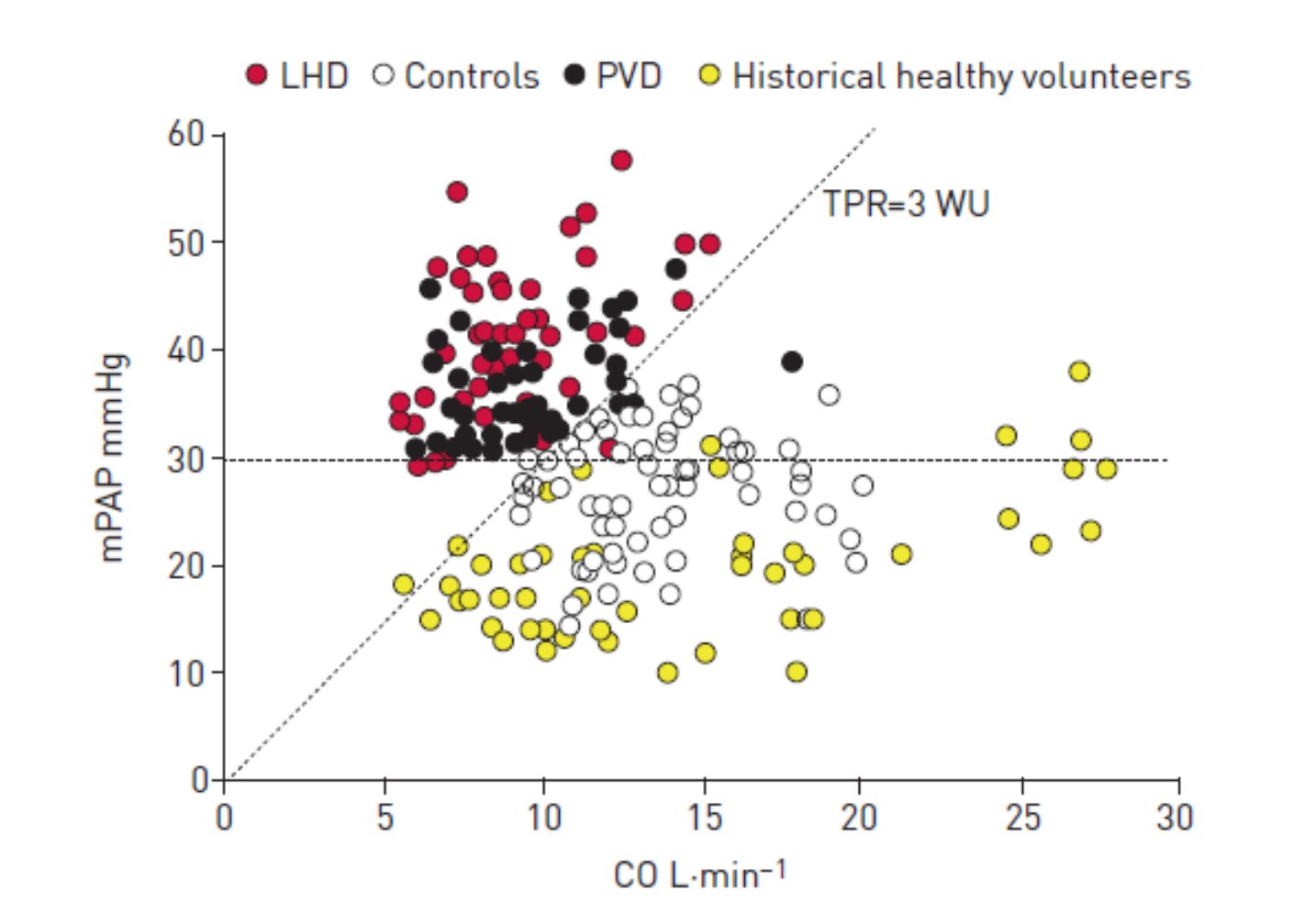
Exercise during right heart catheterization

Exercise Right Heart Catheterization

- Obtain Baseline hemodynamic Profile
- Perform arm or leg exercise
- Goal is 85% age predicted maximal heart rate or elevated PCWP with symptoms
- Retake measurements during exercise including CO and PA oxygen saturation



Hunter C. Champion, Evangelos D. Michelakis and Paul M. Hassoun. Circulation 2009; 120:992-1007



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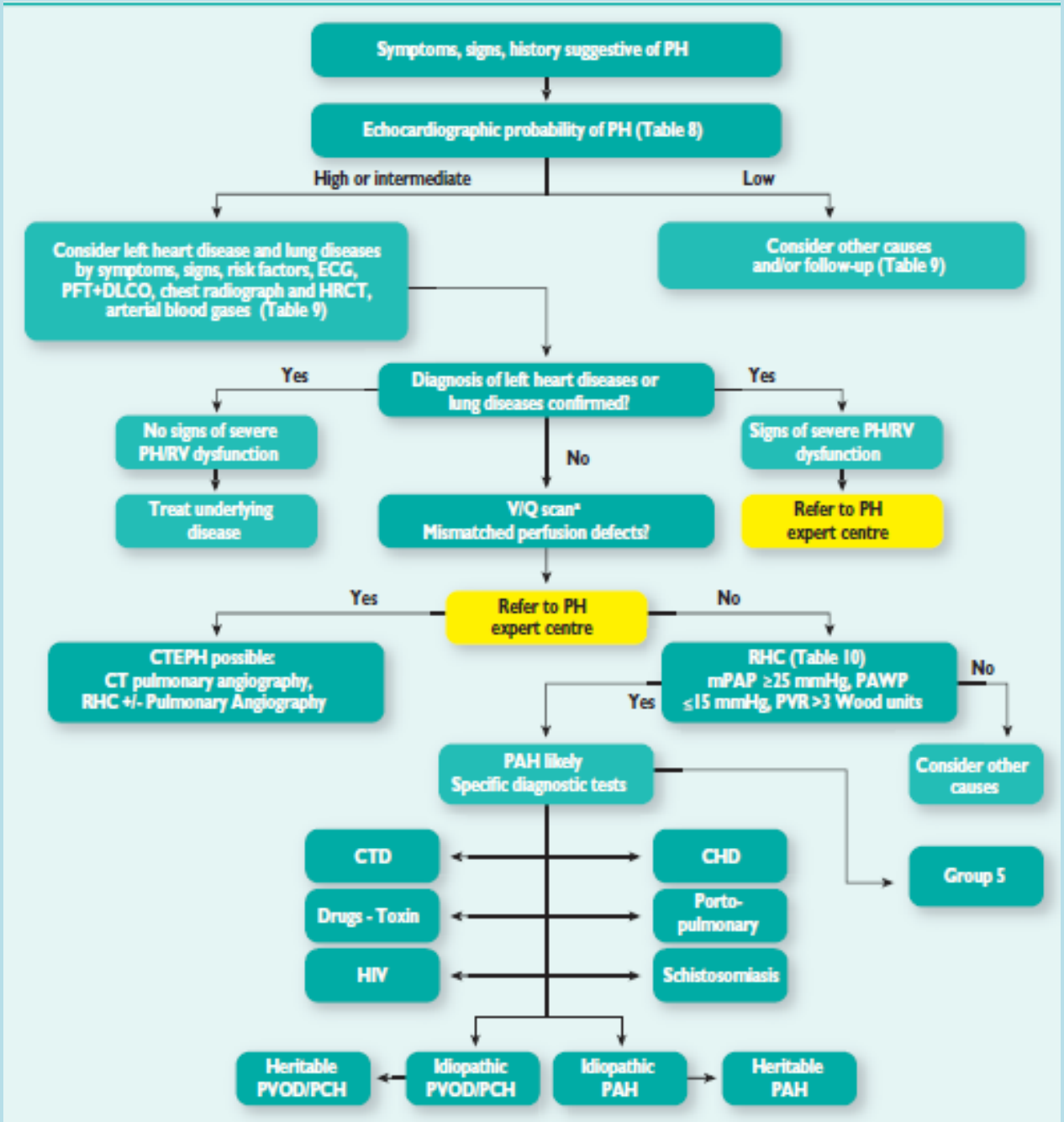
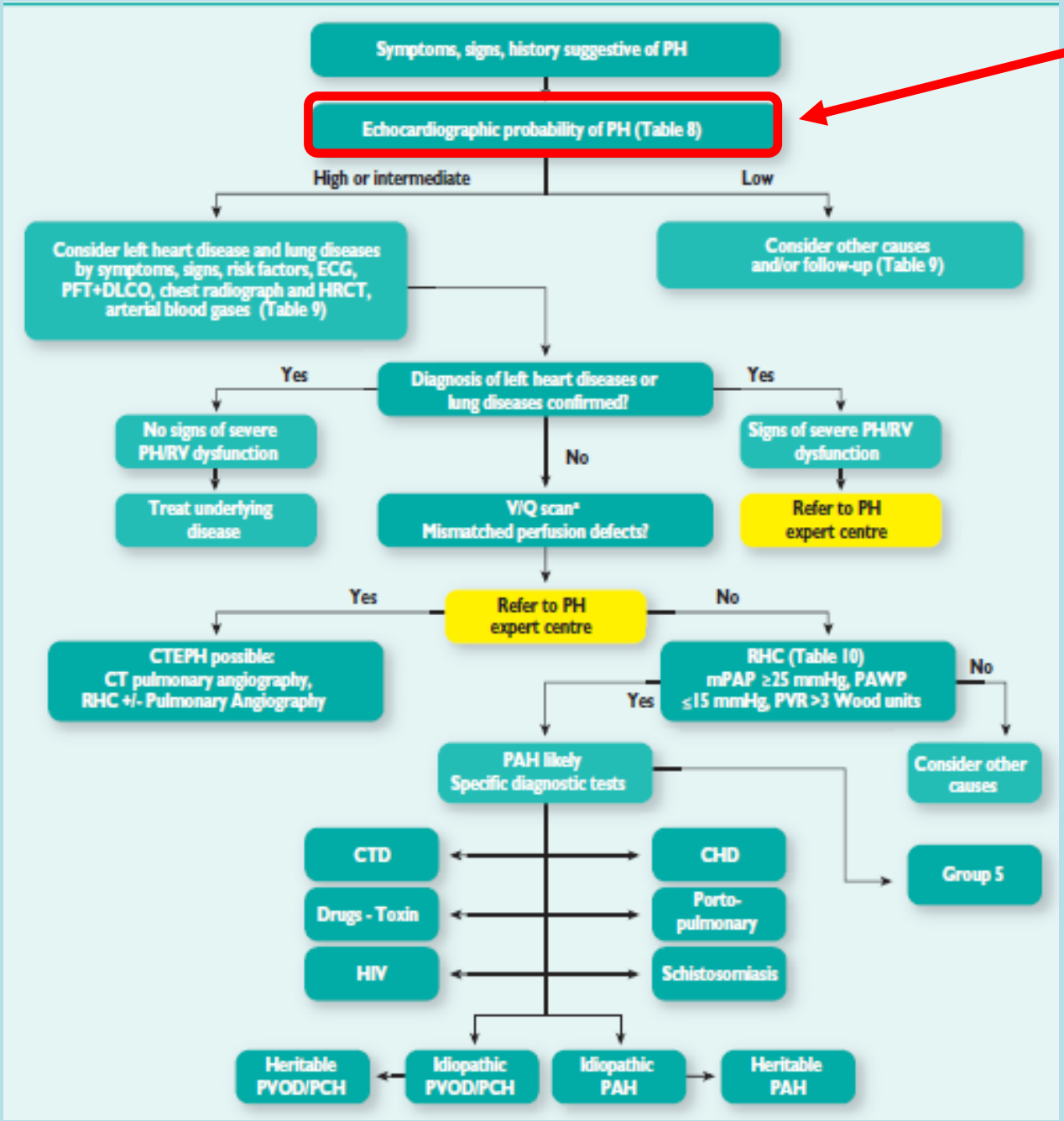
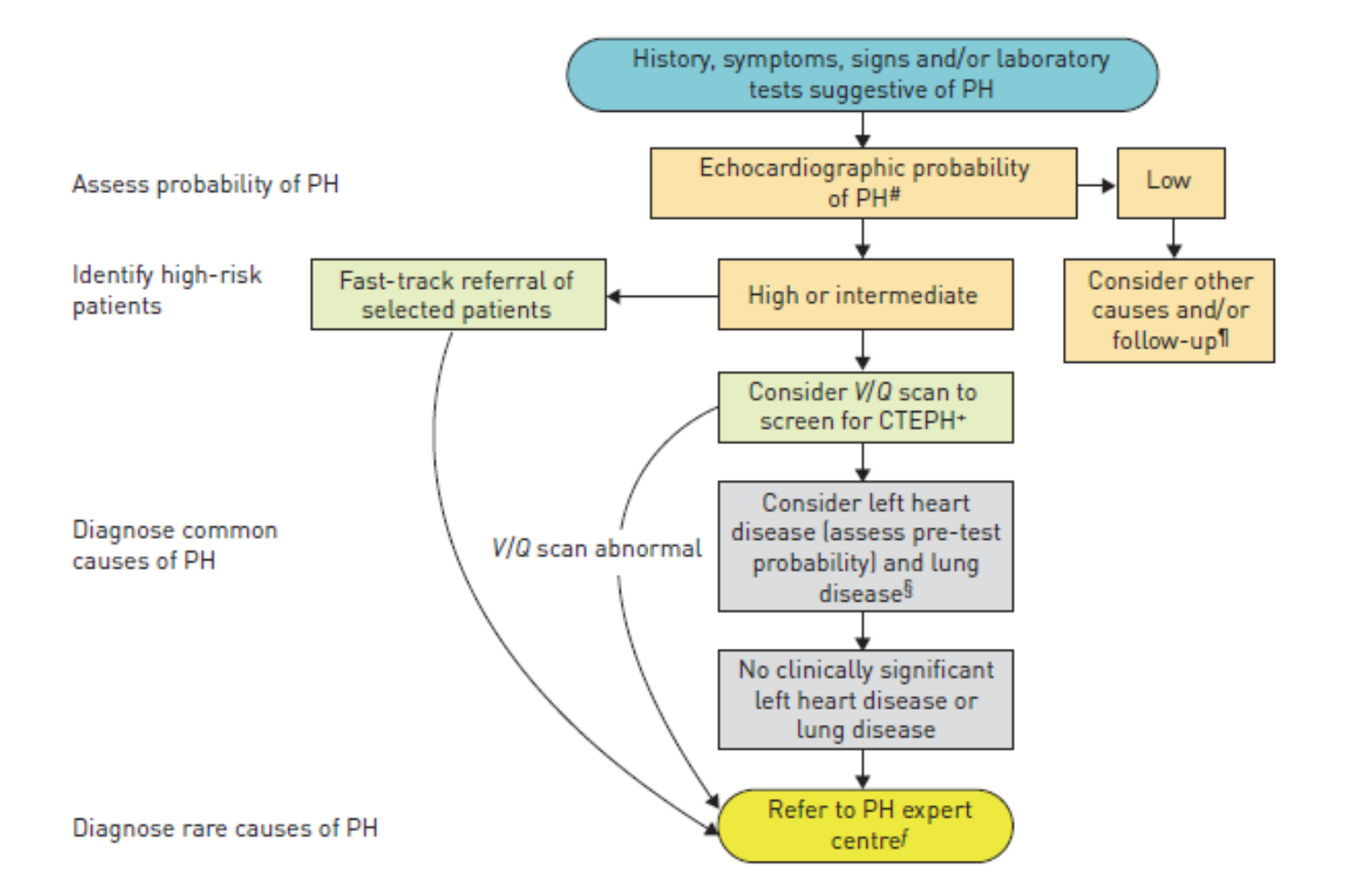
Diagnostic algorithm (guidelines 2015)



Diagnostic algorithm (guidelines 2015)



Algorithm for the diagnosis of PH & its causes: triage of urgent cases & diagnosis of common conditions



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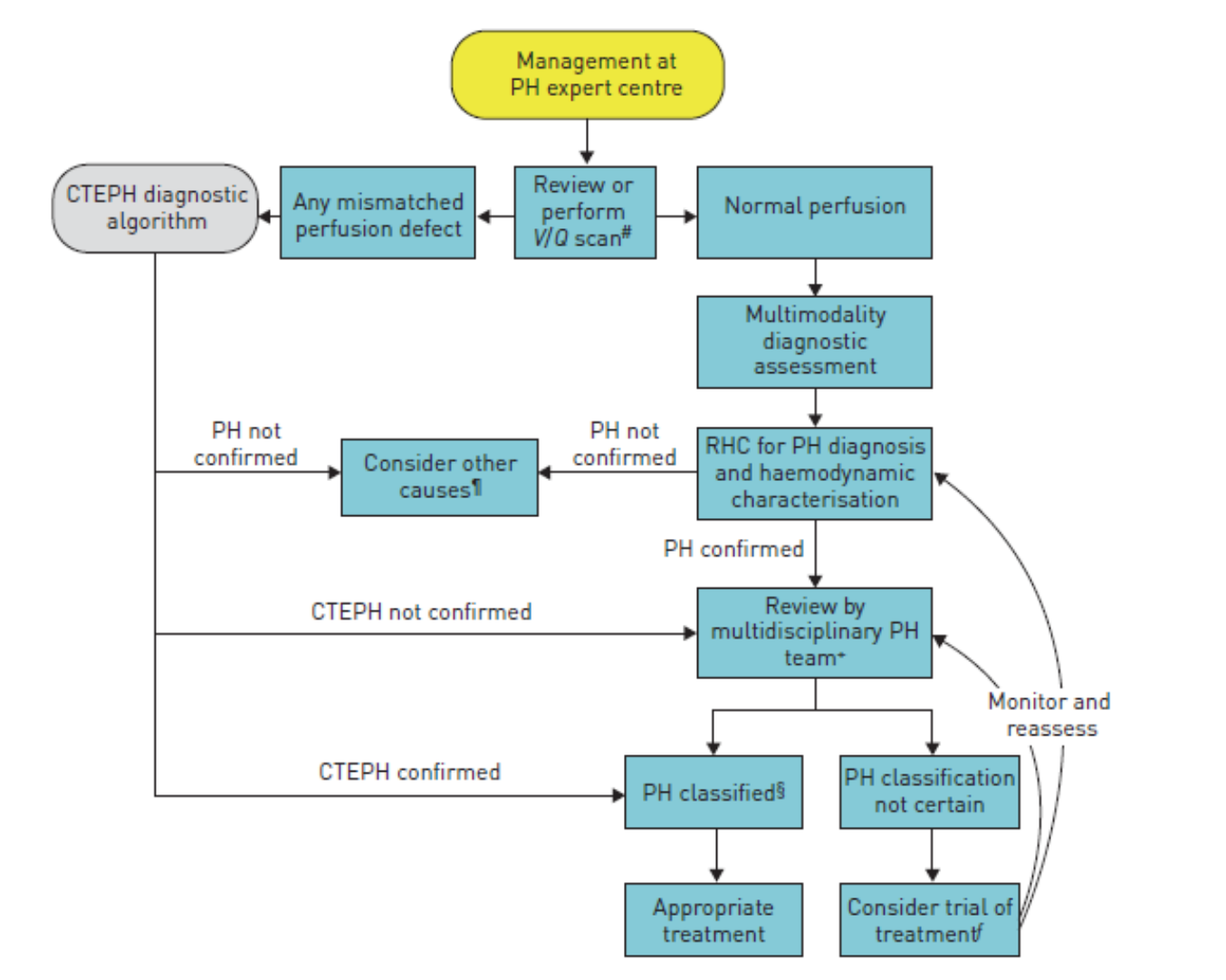
Algorithm for the diagnosis of PH & its causes: role of the PH expert centre



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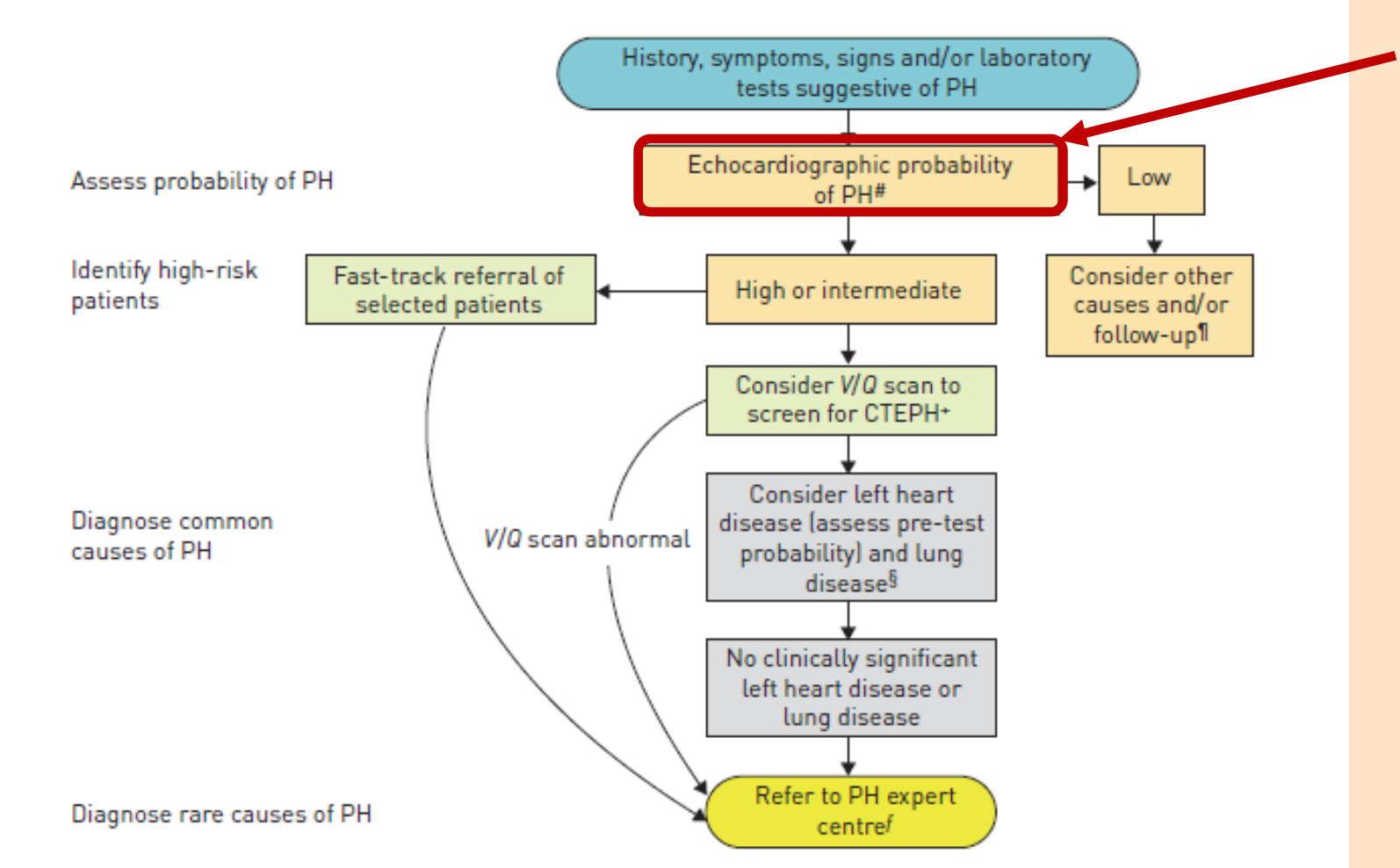

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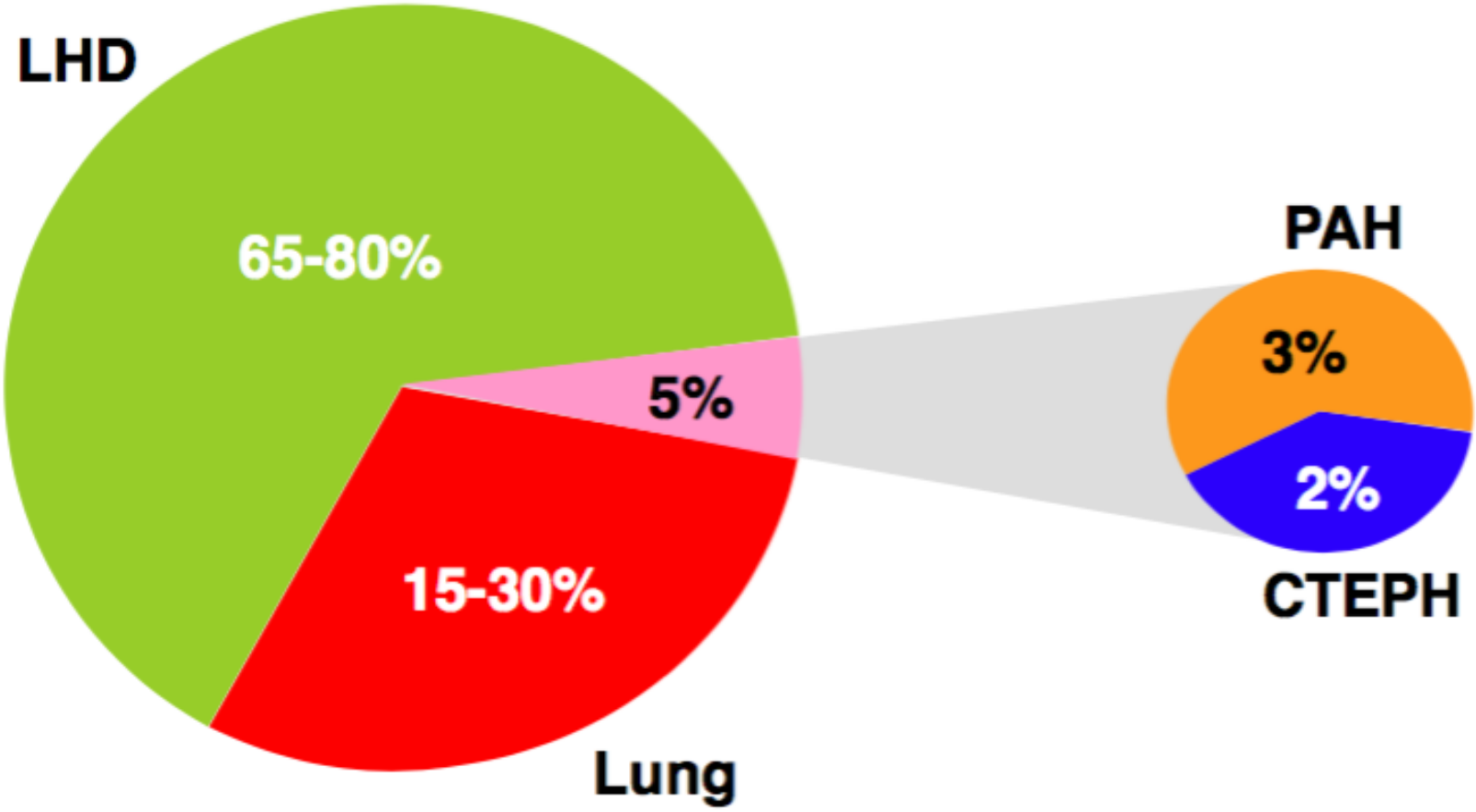
Most Common Forms of Pulmonary Hypertension



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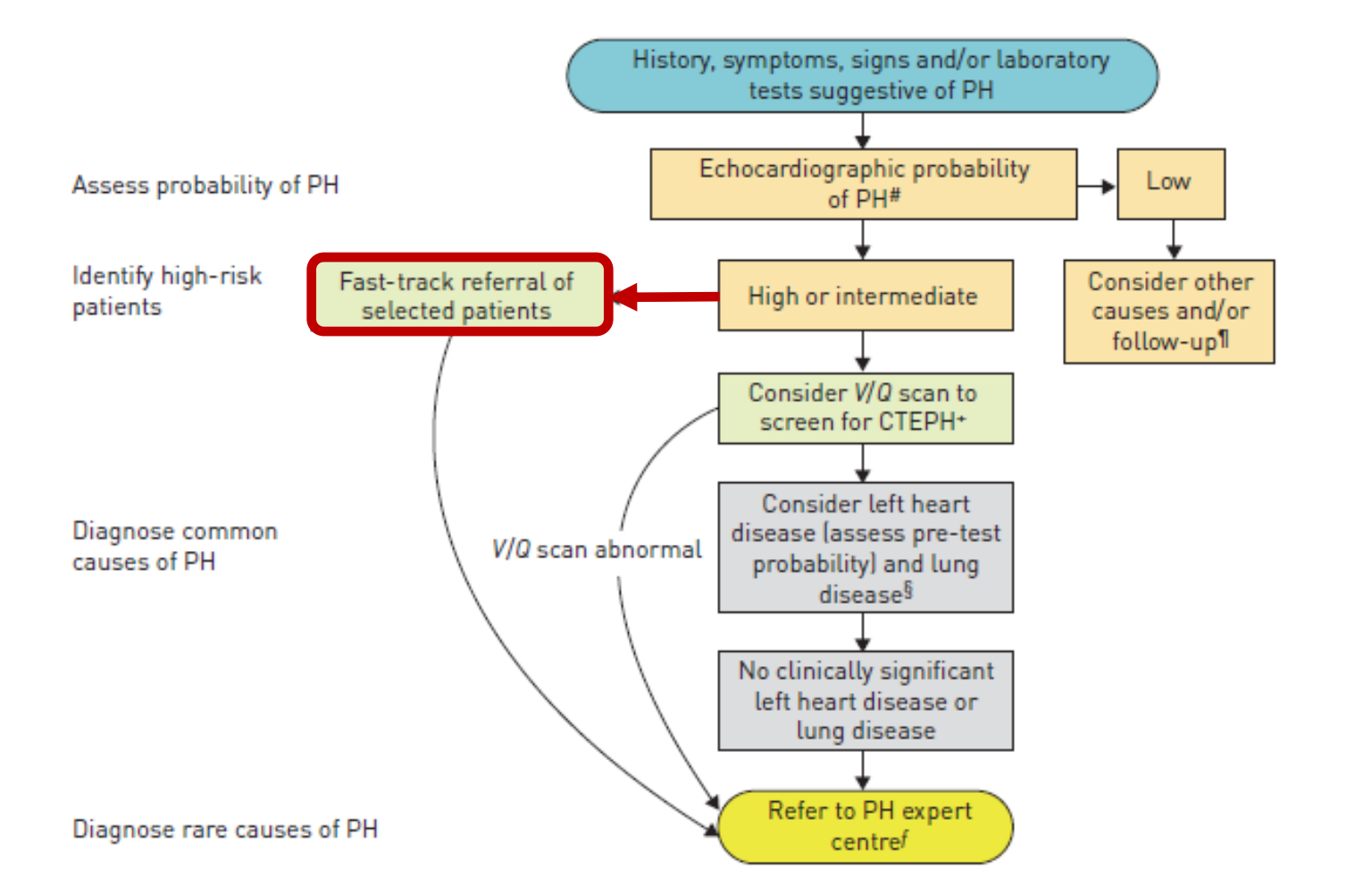

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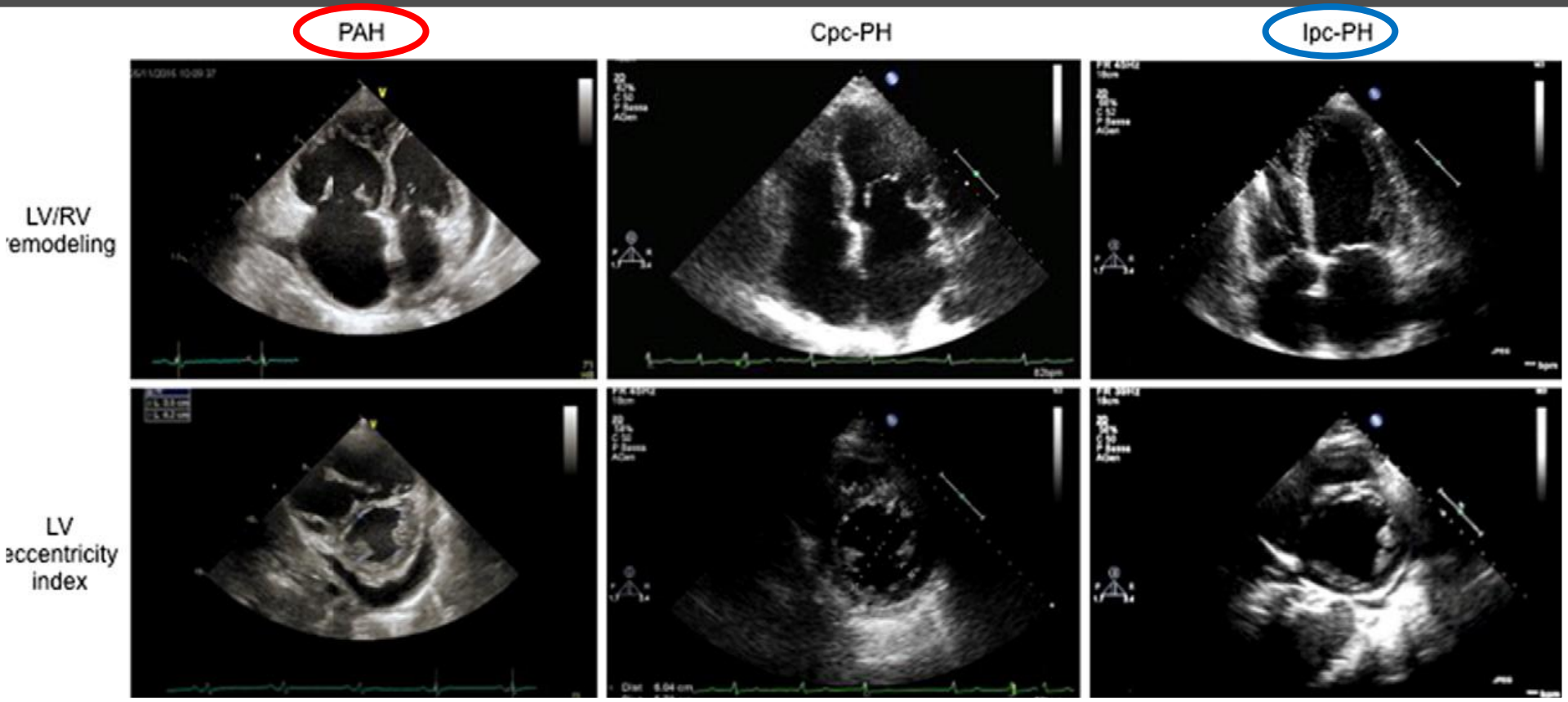
Typical examples of heart remodeling in different types of PH



PROGRESS IN CARDIOVASCULAR DISEASES 59(2016)22–29

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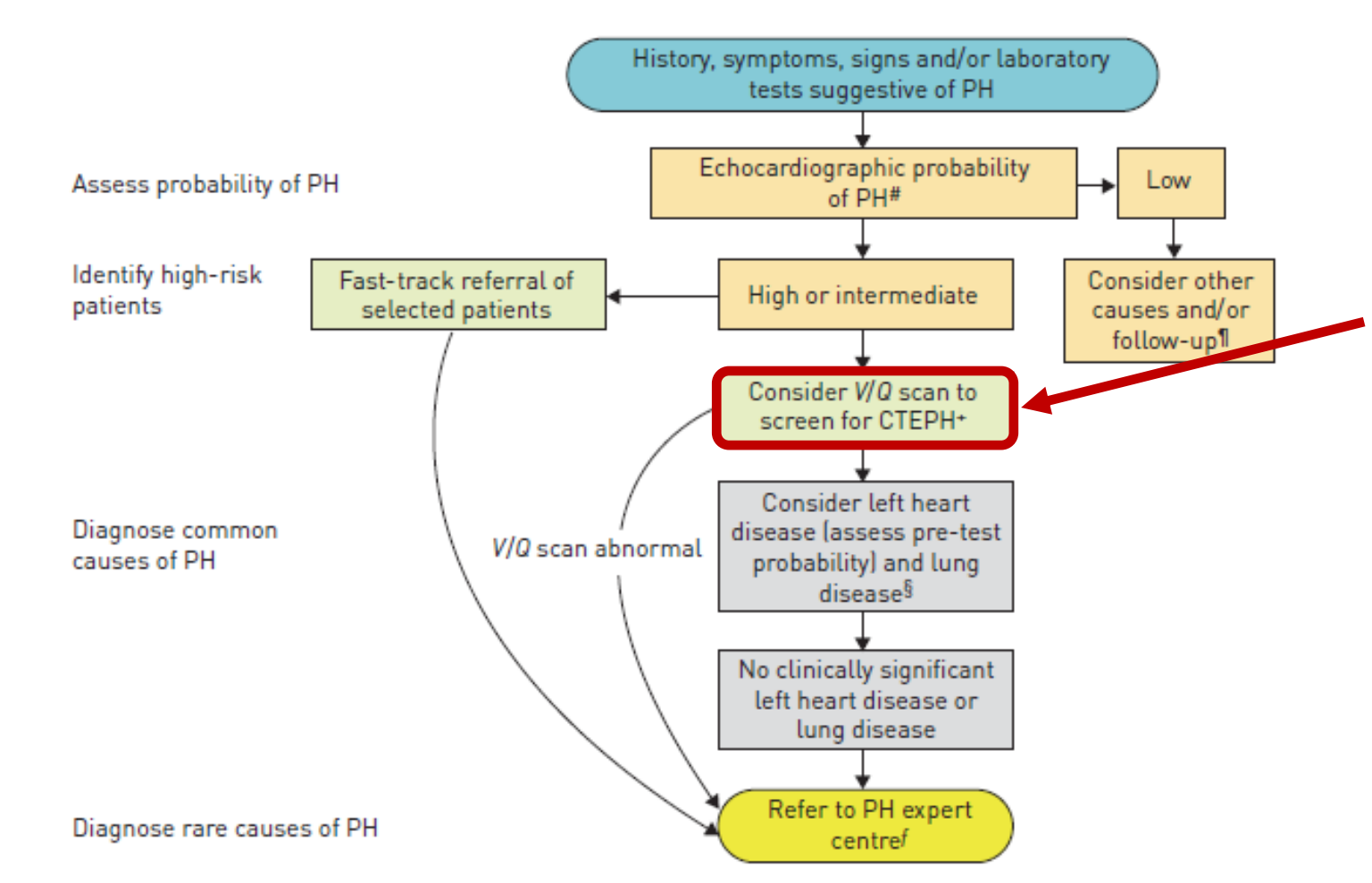

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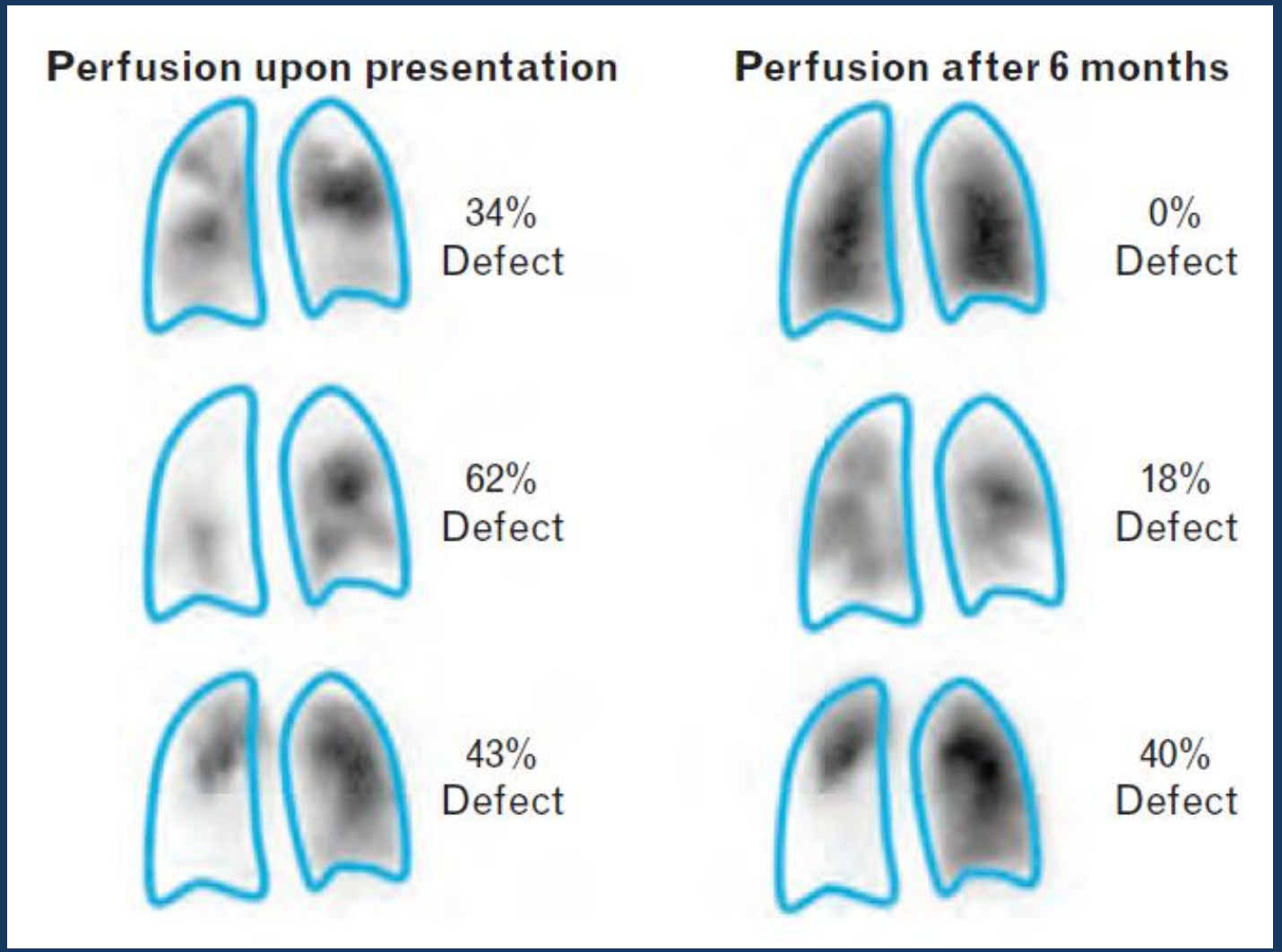

Curr Opin Pulm Med 2013, 19:422–429

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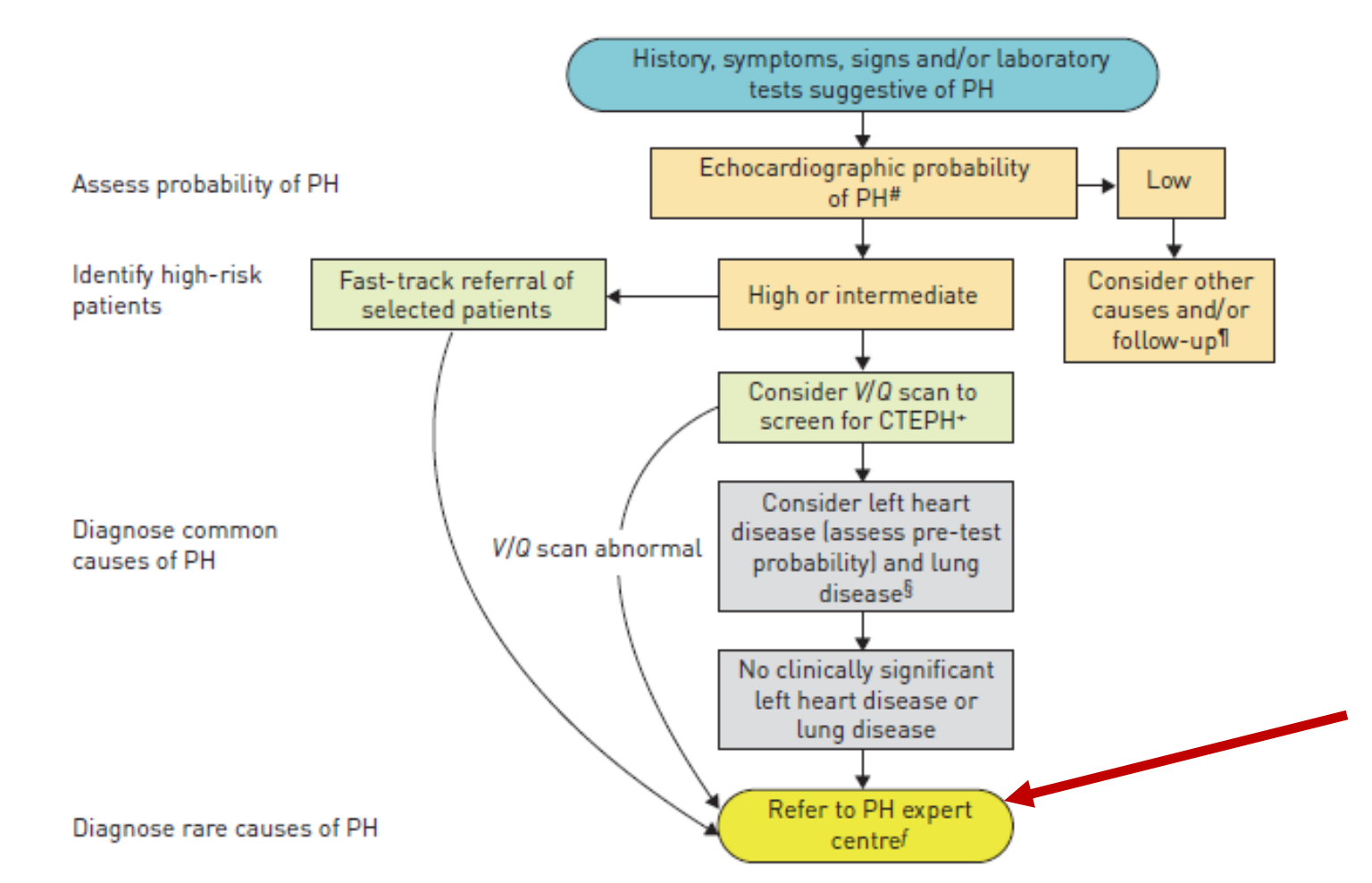

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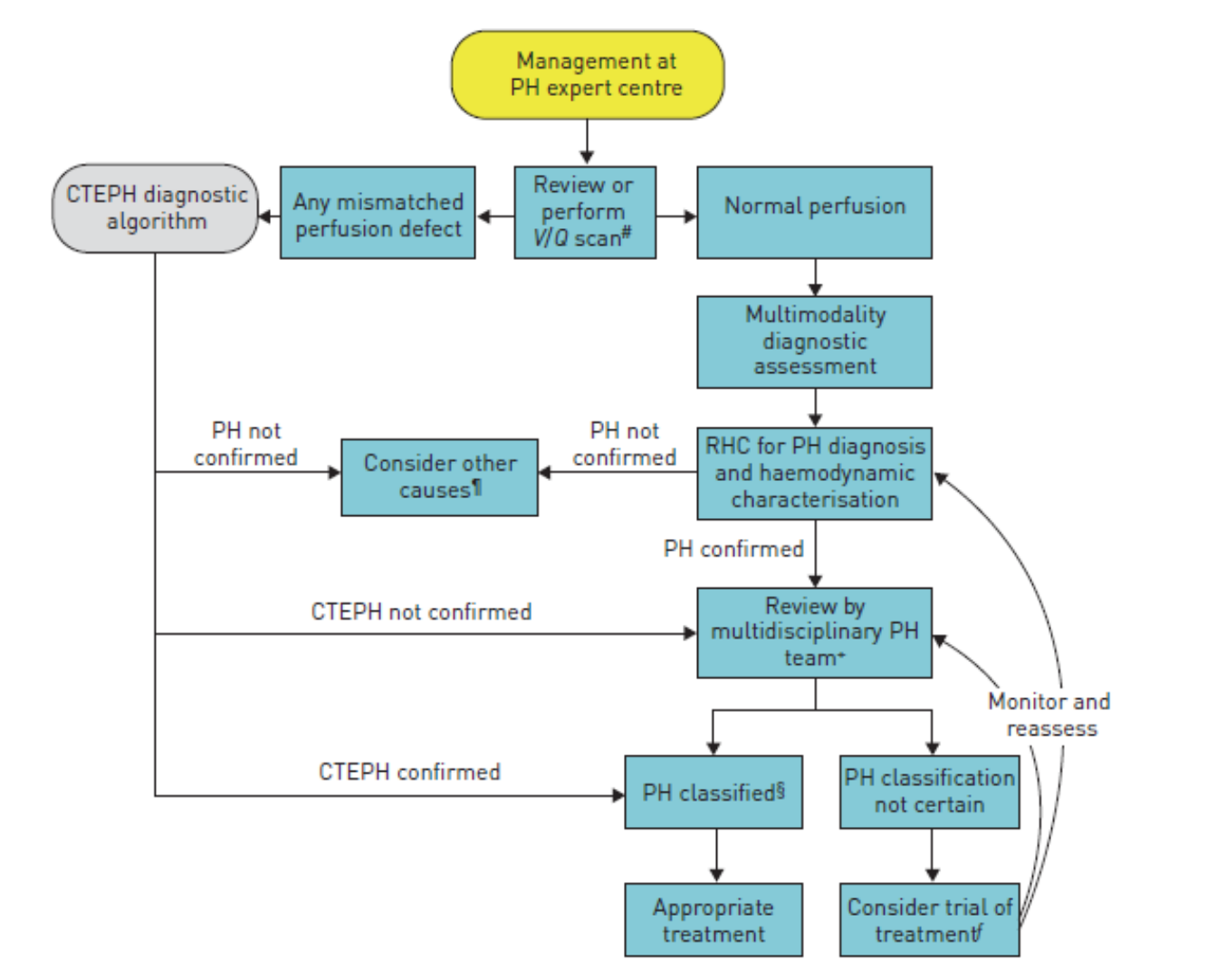
Algorithm for the diagnosis of PH & its causes: role of the PH expert centre



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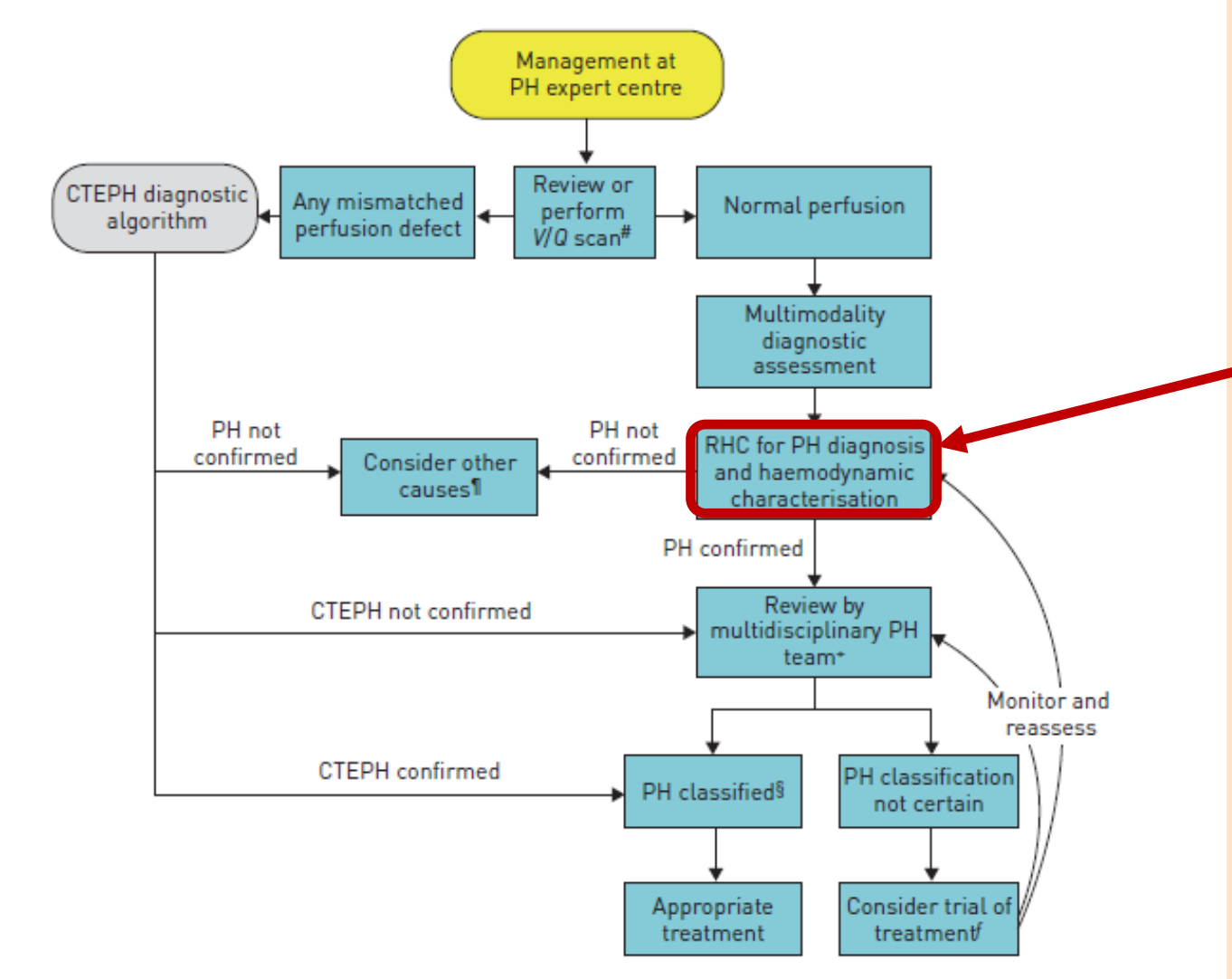

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Haemodynamic definitions of pulmonary hypertension

Definitions	Characteristics	Clinical groups[#]
Pre-capillary PH	mPAP >20 mmHg PAWP ⩽15 mmHg PVR ⩾3 WU	1, 3, 4 and 5
Isolated post-capillary PH (IpcPH)	mPAP >20 mmHg PAWP >15 mmHg PVR <3 WU	2 and 5
Combined pre- and post-capillary PH (CpcPH)	mPAP >20 mmHg PAWP >15 mmHg PVR ⩾3 WU	2 and 5

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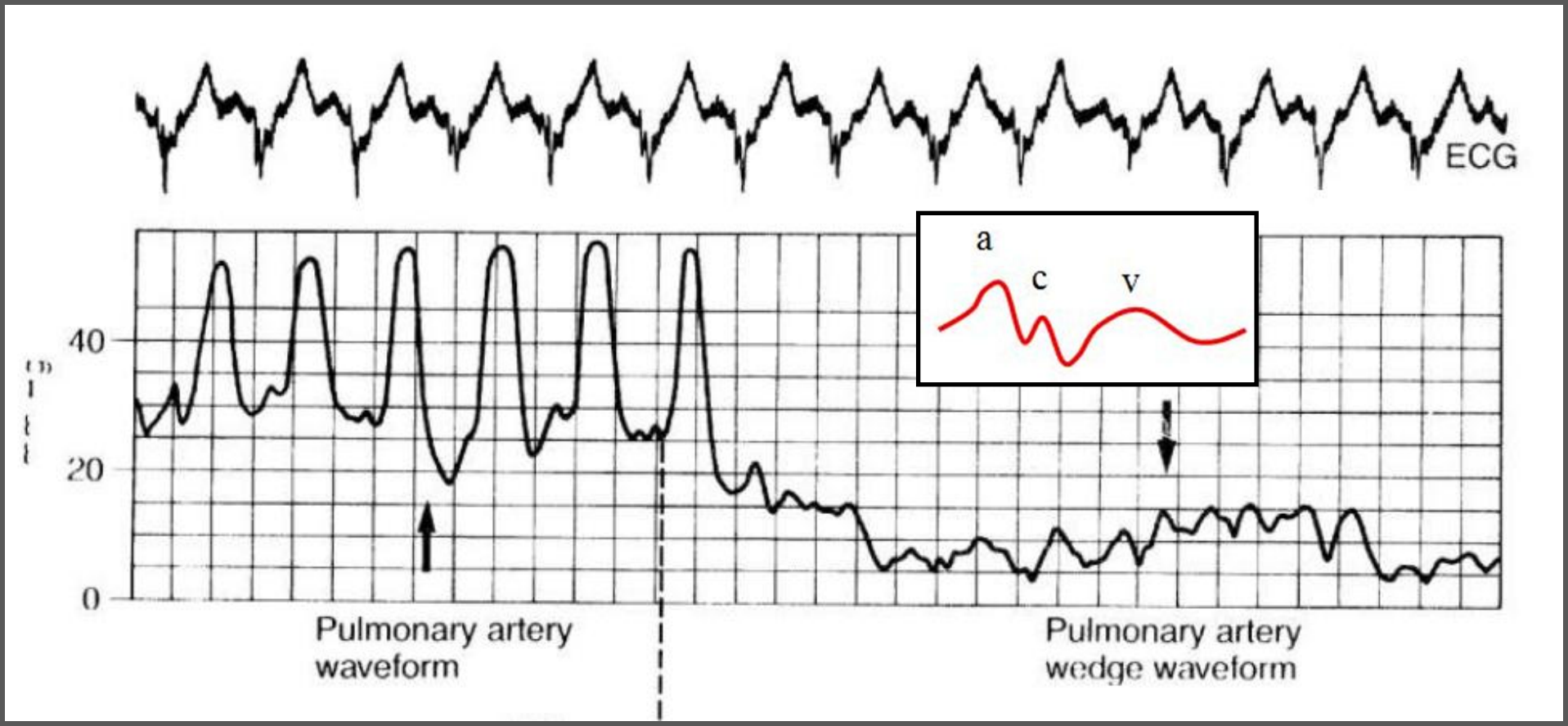
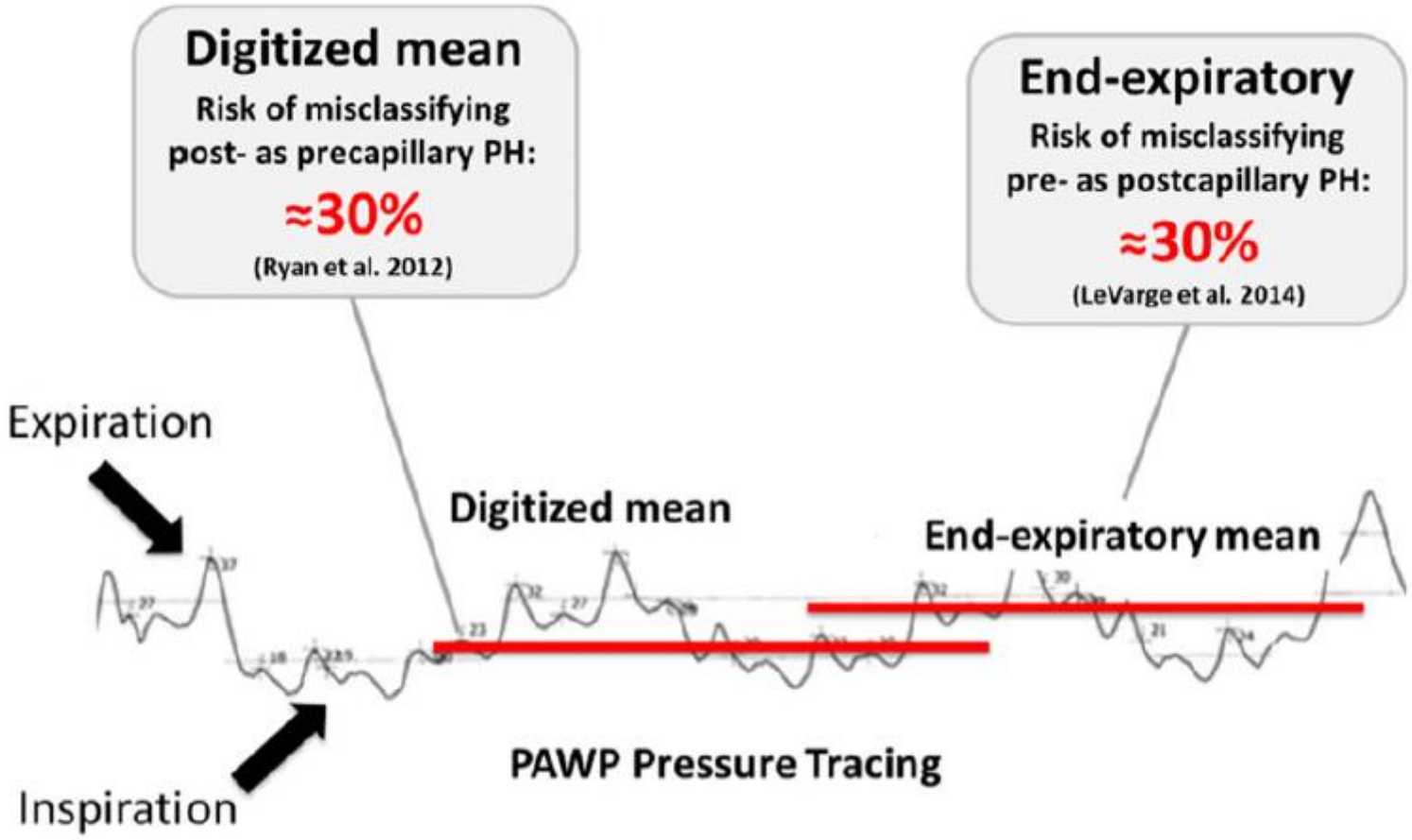
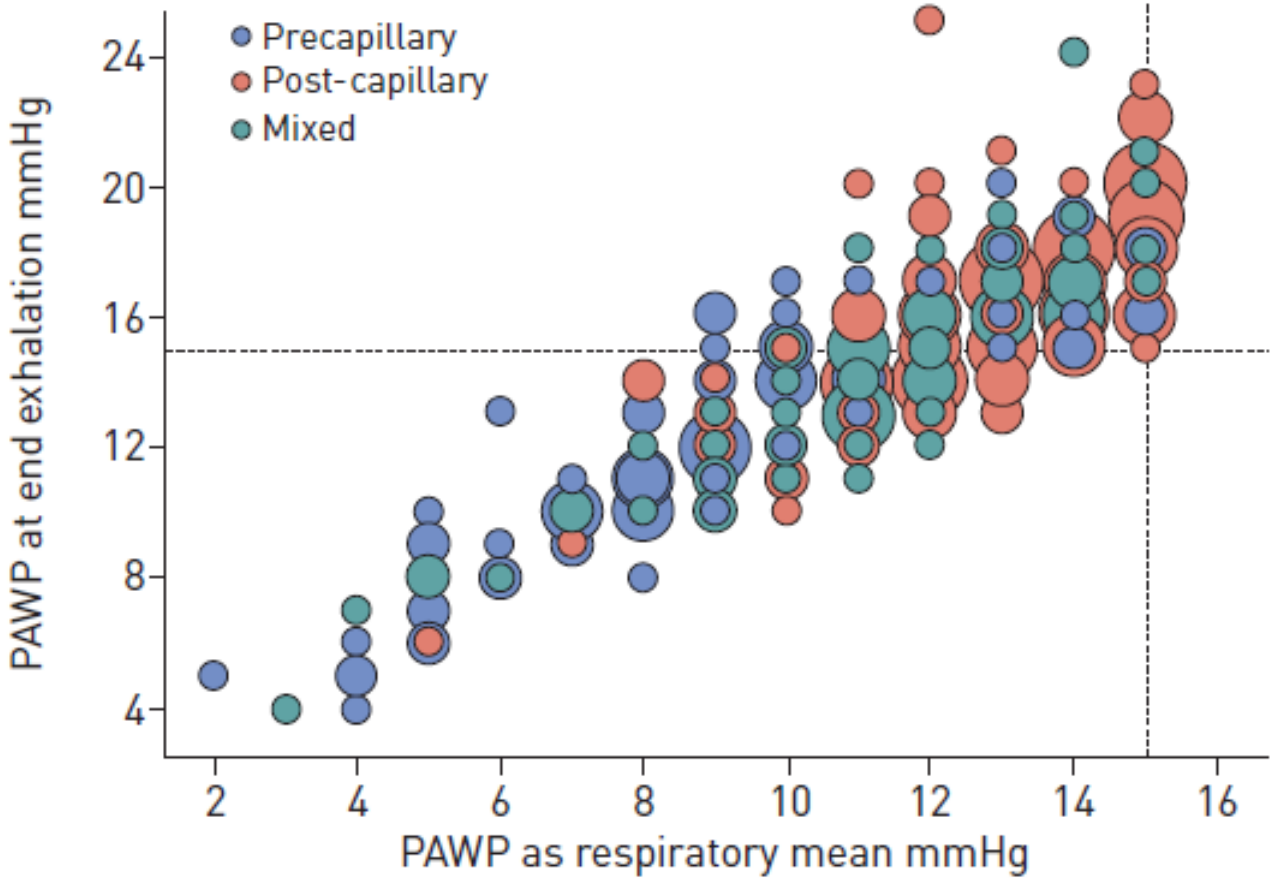

ECG
a
c
v
40
20
0
Pulmonary artery waveform
Pulmonary artery wedge waveform

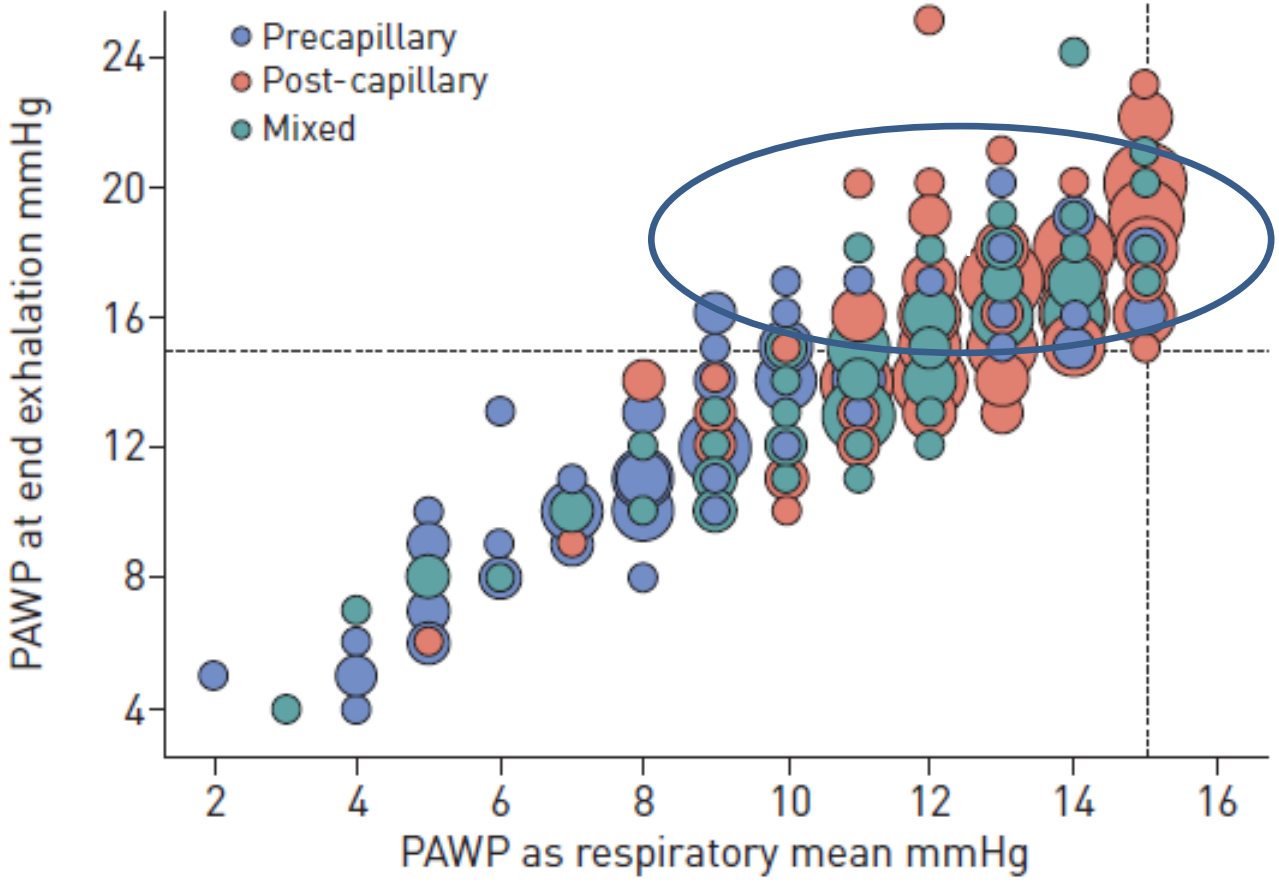
Potential misclassifications between pre- and post-capillary pulmonary hypertension depending on the method of PAWP reading



S. Rosenkranz et al. Eur Heart J. 2016;37(12):942-54



Eur Respir J 2014; 44: 425–434



Eur Respir J 2014; 44: 425–434

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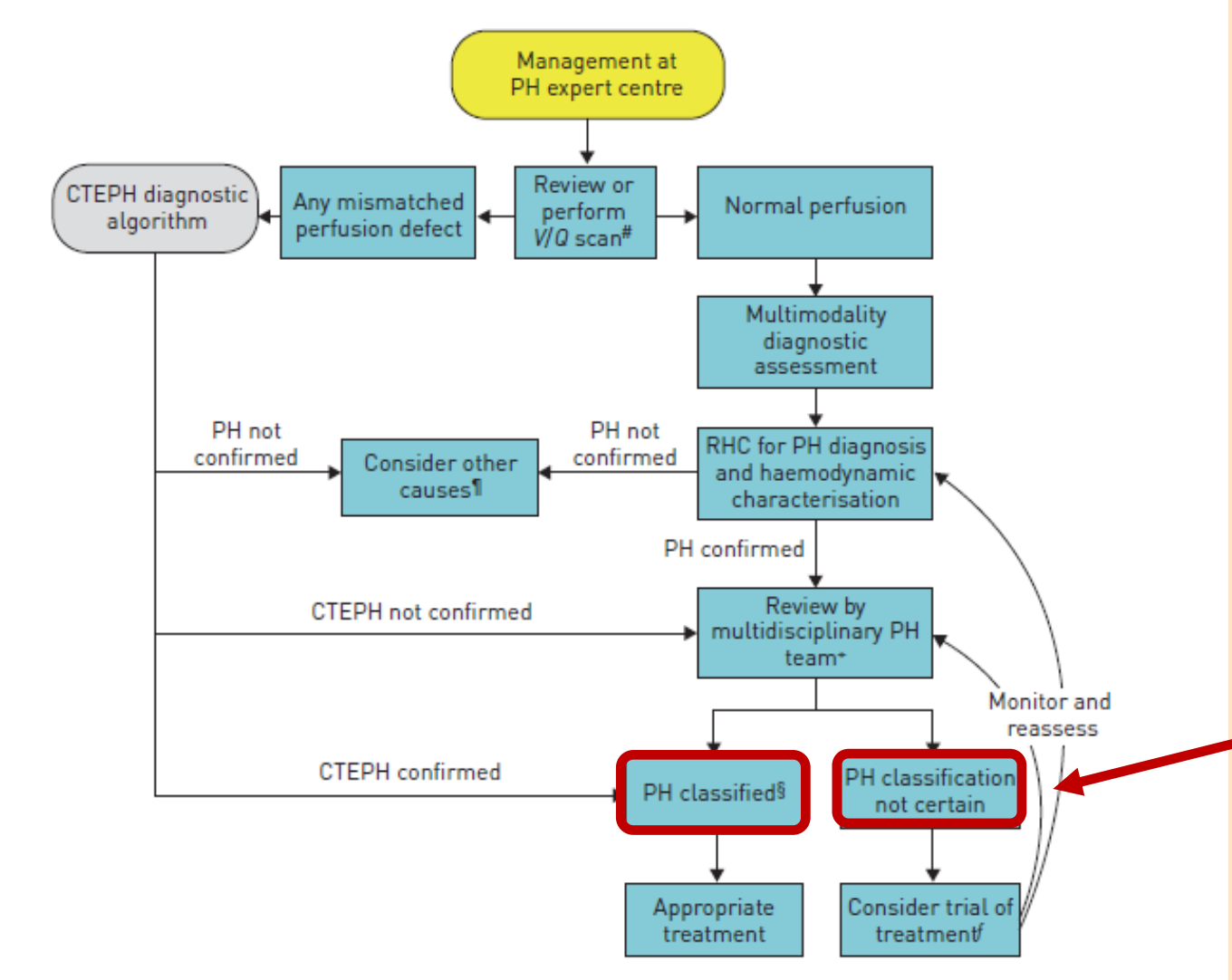

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Criteria favouring group 1 versus group 3 pulmonary hypertension (PH)

Criteria favouring group 1 (PAH)	Testing	Criteria favouring group 3 (PH due to lung disease)
Extent of lung disease		
Normal or mildly impaired: • FEV_1 >60% pred (COPD) • FVC >70% pred (IPF) • Low diffusion capacity in relation to obstructive/restrictive changes	Pulmonary function testing	Moderate to very severely impaired: • FEV_1 <60% pred (COPD) • FVC <70% pred (IPF) • Diffusion capacity "corresponds" to obstructive/restrictive changes
Absence of or only modest airway or parenchymal abnormalities	High-resolution CT scan¶	Characteristic airway and/or parenchymal abnormalities
Haemodynamic profile		
Moderate-to-severe PH	Right heart catheterisation Echocardiogram	Mild-to-moderate PH
Ancillary testing		
Present	Further PAH risk factors (*e.g.* HIV, connective tissue disease, *BMPR2* mutations, *etc.*)	Absent
Features of exhausted circulatory reserve: • Preserved breathing reserve • Reduced oxygen pulse • Low $CO/\dot{V}O_2$ slope • Mixed venous oxygen saturation at lower limit • No change or decrease in P_{ACO_2} during exercise	Cardiopulmonary exercise test+ (P_{ACO_2} particularly relevant in COPD)	Features of exhausted ventilatory reserve: • Reduced breathing reserve • Normal oxygen pulse • Normal $CO/\dot{V}O_2$ slope • Mixed venous oxygen saturation above lower limit • Increase in P_{ACO_2} during exercise
Predominant haemodynamic profile		Predominant obstructive/restrictive profile

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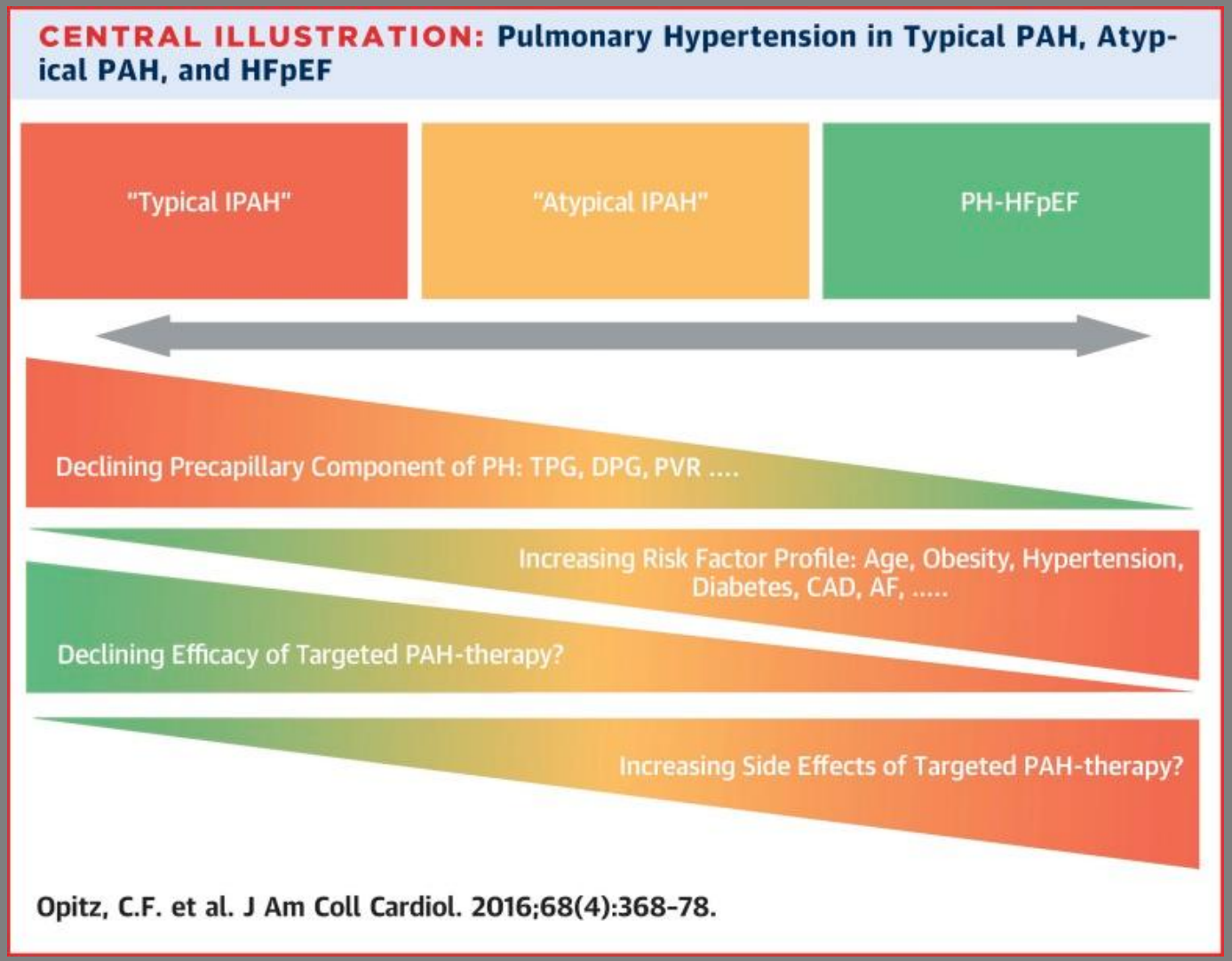
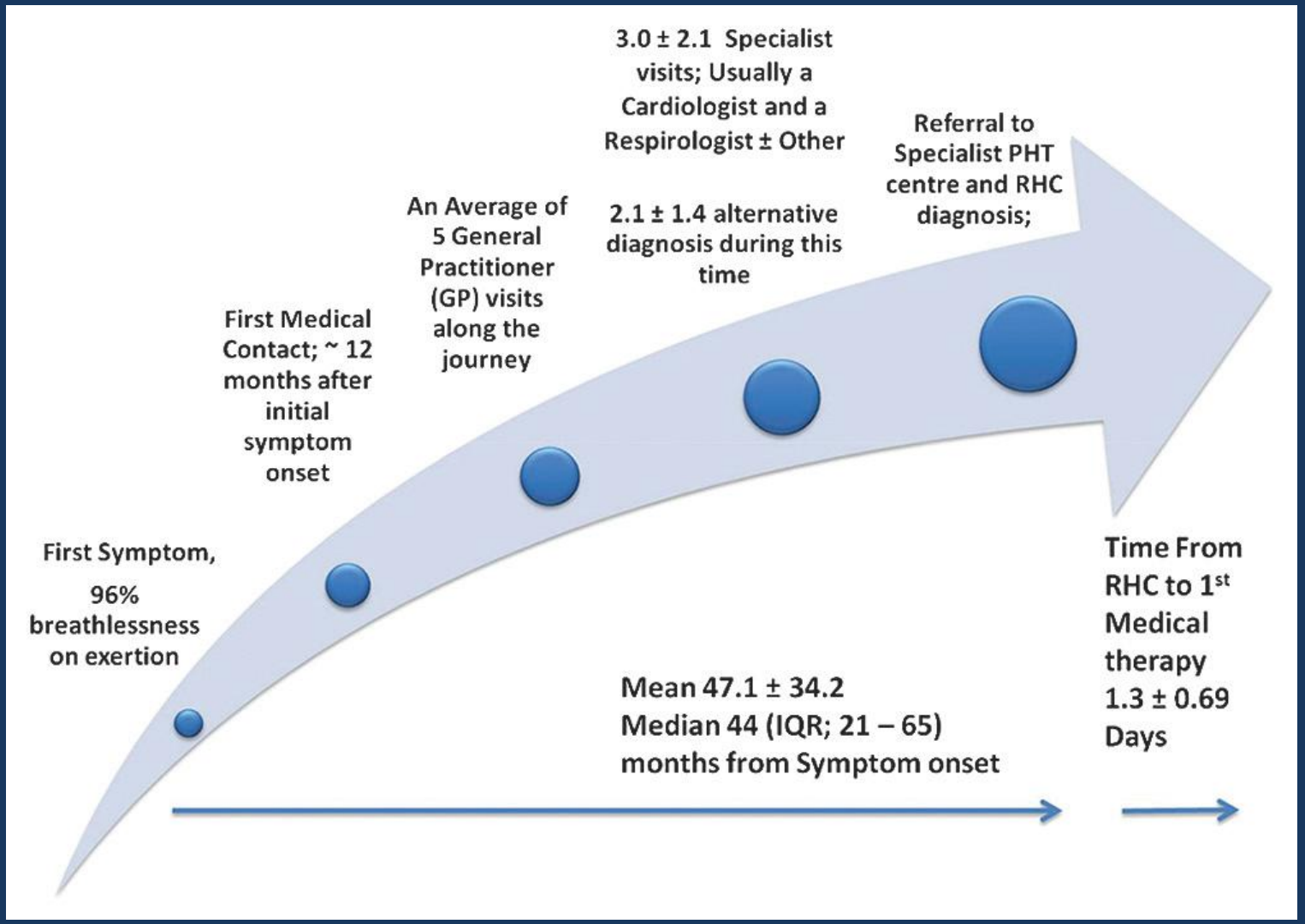
Pre-Capillary, Combined, and Post-Capillary Pulmonary Hypertension

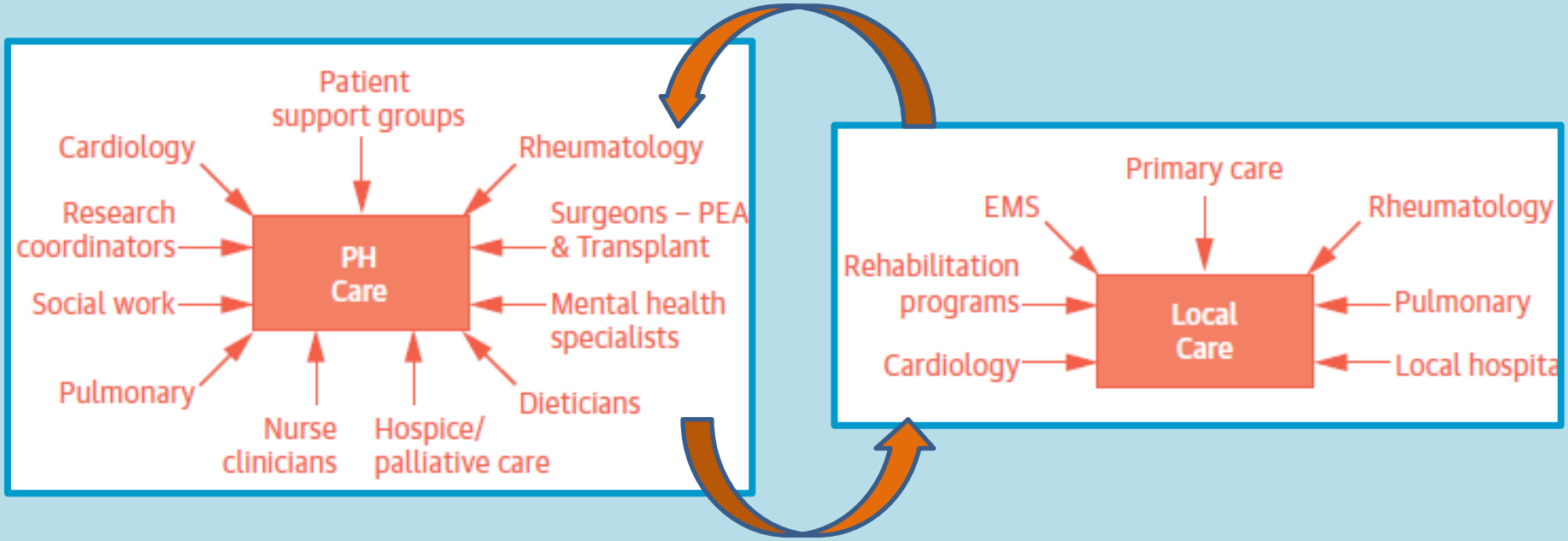
A Pathophysiological Continuum?





Strange et al.: Pulmonary Circulation, January-March 2013

Collaboration: PH Care- Local Care



JACC 2015;65:1976–97