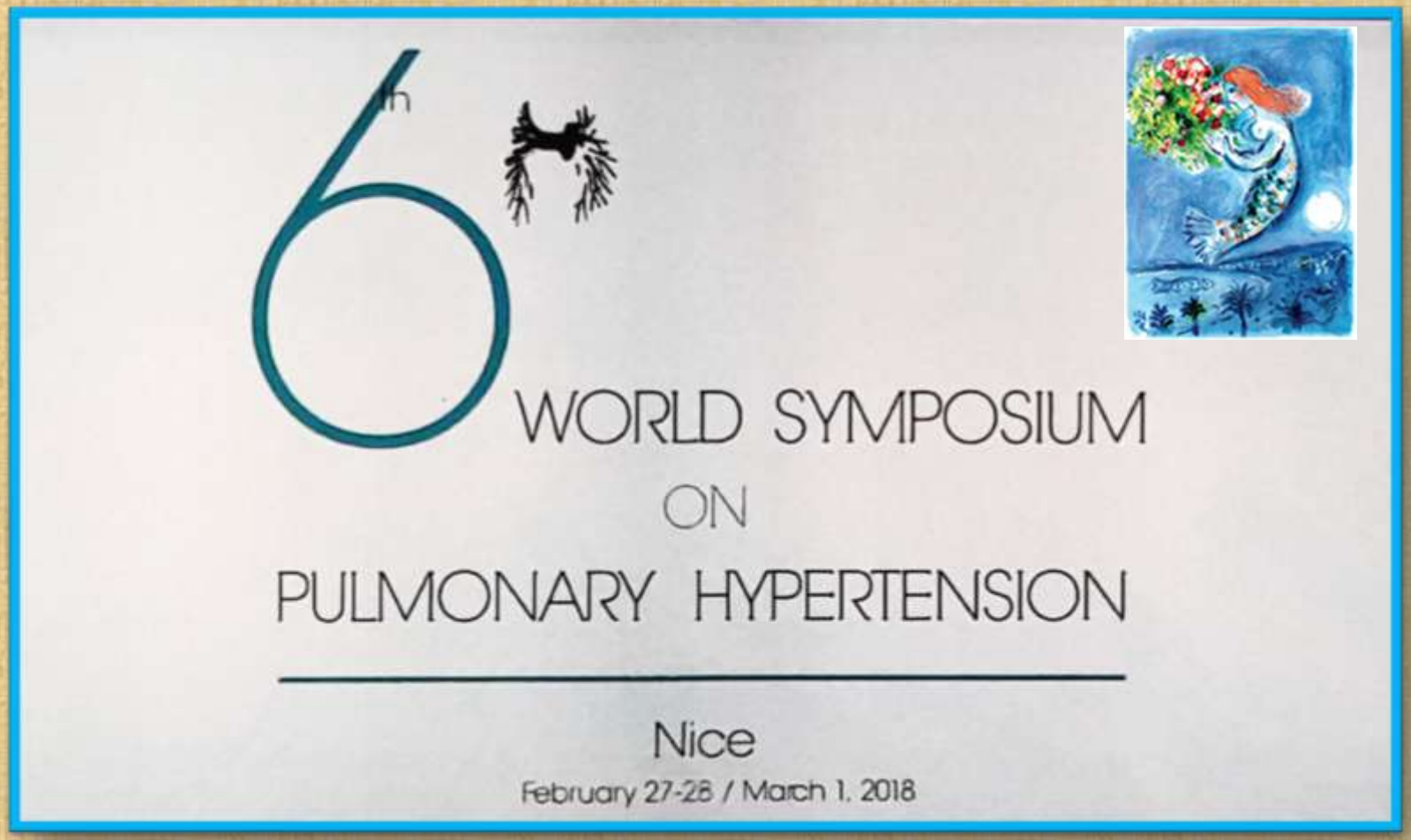
Πότε υποπτευόμαστε πνευμονική υπέρταση-Ο ρόλος του πνευμονολόγου

Φραντζέσκα Φραντζεσκάκη
Πνευμονολόγος- Εντατικολόγος
Β'Παν/κή Κλινική Εντατικής Θεραπείας
ΠΓΝ «ΑΤΤΙΚΟΝ»



"Proceedings of the 6th World Symposium on Pulmonary Hypertension"
Edited by N. Galiè, V.V. McLaughlin, L.J. Rubin and G. Simonneau

Eur Respir J, December 2018

TABLE 1 Haemodynamic definitions of pulmonary hypertension (PH)

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

Simonneau G et al. *Eur Resp J* 2018

TABLE 2 Updated clinical classification of pulmonary hypertension (PH)

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

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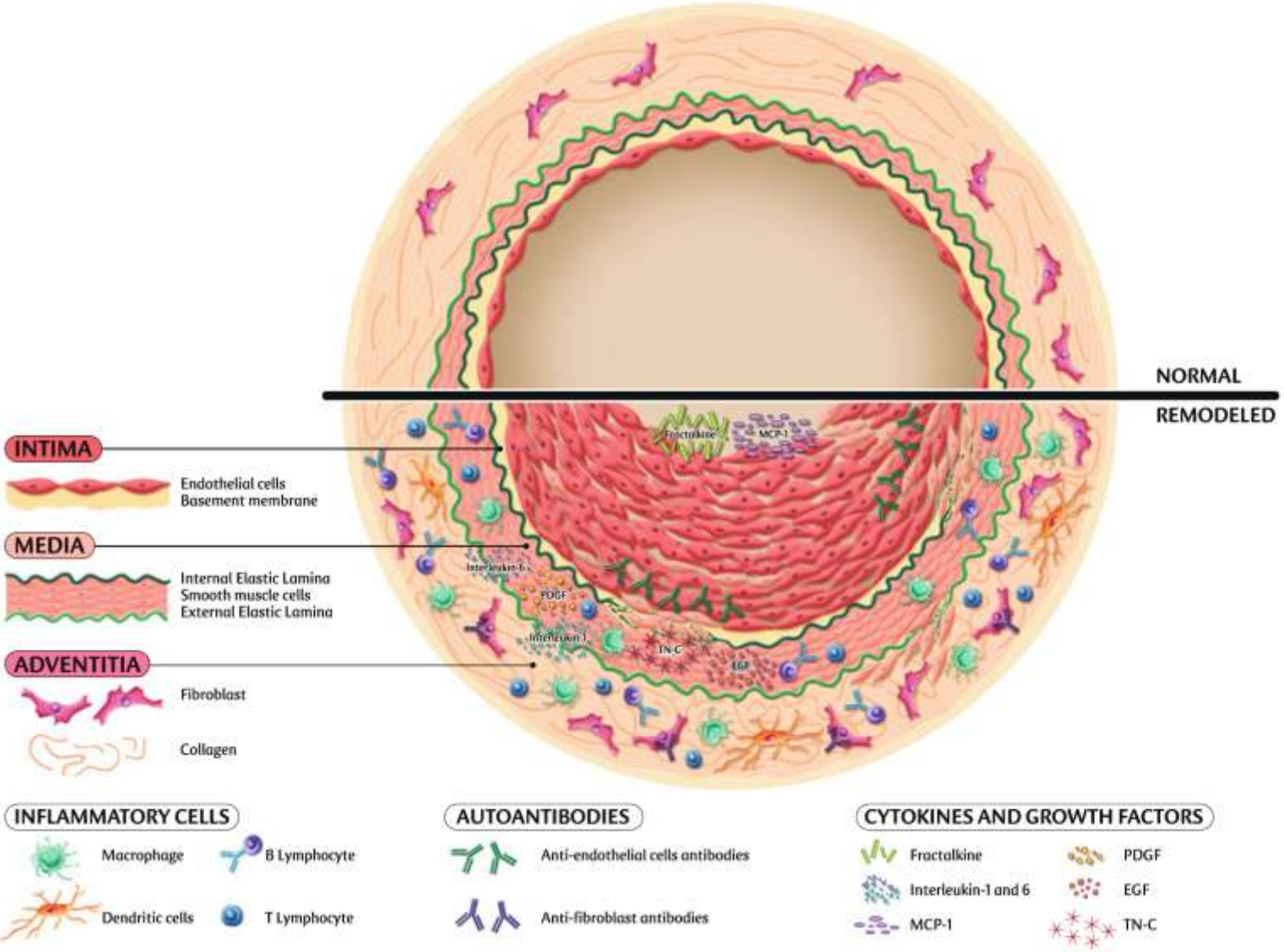
- 5.2 Systemic and metabolic disorders
- 5.3 Others
- 5.4 Complex congenital heart disease

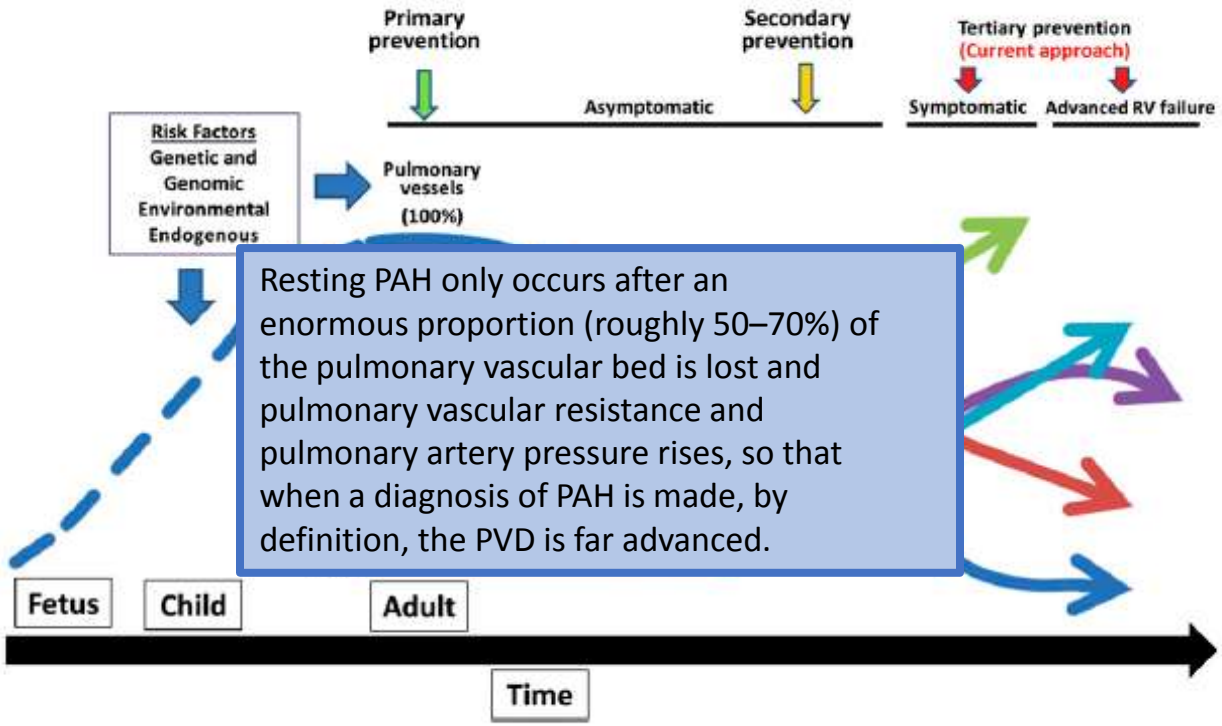
PAH: pulmonary arterial hypertension; PVOD: pulmonary veno-occlusive disease; PCH: pulmonary capillary haemangiomatosis; LVEF: left ventricular ejection fraction.

Simonneau G et al. *Eur Resp J* 2018



Kherbeck et al *Clin Rev Allergy Immunol* 2011

Nature of the PAH disease



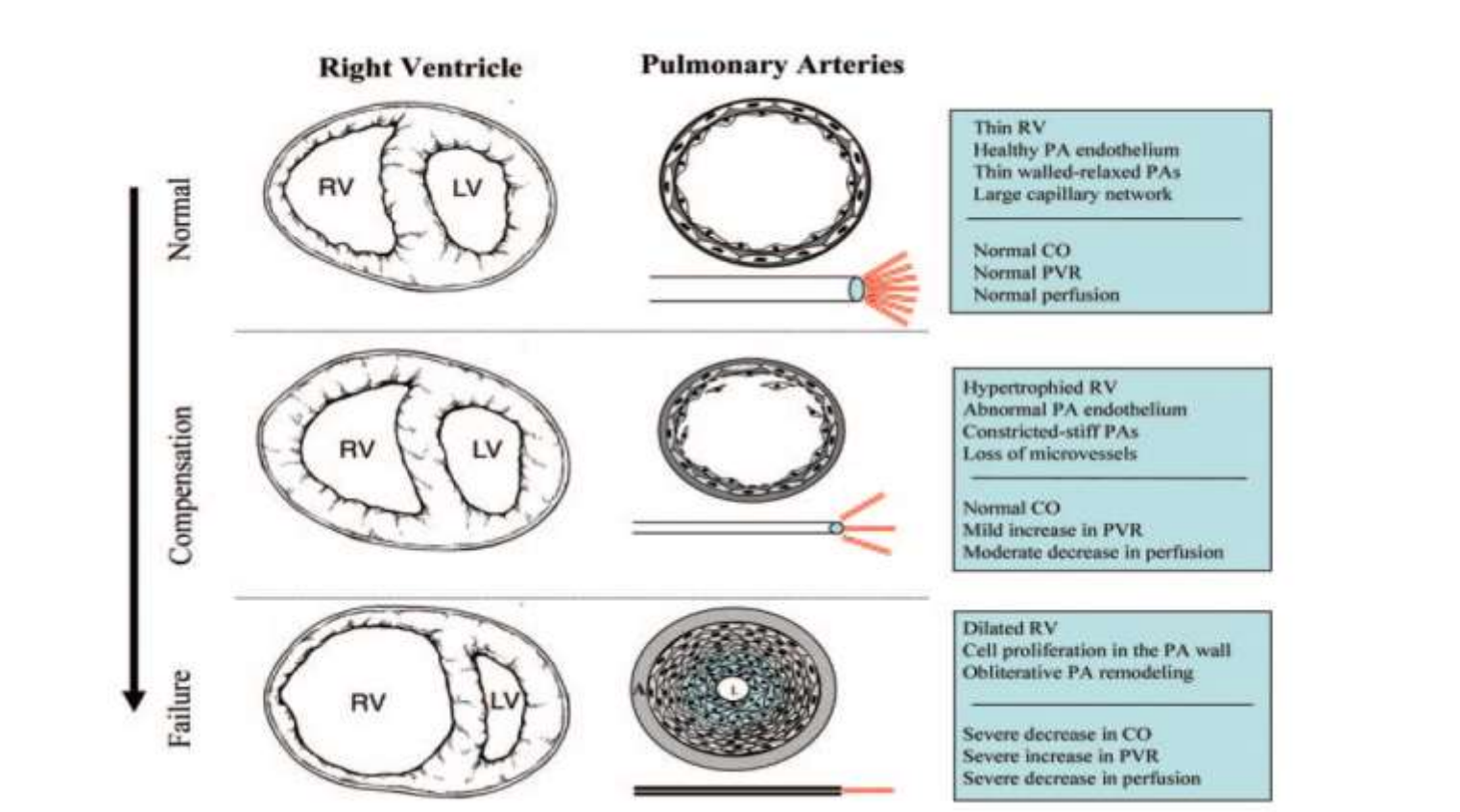
Resting PAH only occurs after an enormous proportion (roughly 50–70%) of the pulmonary vascular bed is lost and pulmonary vascular resistance and pulmonary artery pressure rises, so that when a diagnosis of PAH is made, by definition, the PVD is far advanced.

Figure 1. Due to the tremendous reserve of the pulmonary vasculature, resting pulmonary arterial hypertension (PAH) and symptoms occur long after the initial inciting events trigger pulmonary vascular dysfunction (PVD). Multiple known risk factors exist that can prompt the loss of normal pulmonary vessel function *in utero* and beyond. Risk factors are variable, including: (*1*) genetic (e.g., bone morphogenetic protein receptor type 2 [*BMPR2*] gene mutations and sickle cell lung disease); (*2*) environmental (e.g., dietary stimulants); and (*3*) endogenous (e.g., premature lung disease and portal hypertension). Resting pulmonary hypertension (PH) only occurs after an enormous proportion of the pulmonary vascular bed is lost, causing a rise in mean pulmonary artery pressure (mPAP) and pulmonary vascular resistance (PVR). By the time a diagnosis of PH is made, PVD is far advanced. Ultimately, advanced right ventricular (RV) failure will occur. Primary prevention efforts must focus upon the detection and prevention of PVD, before the onset of PH.

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

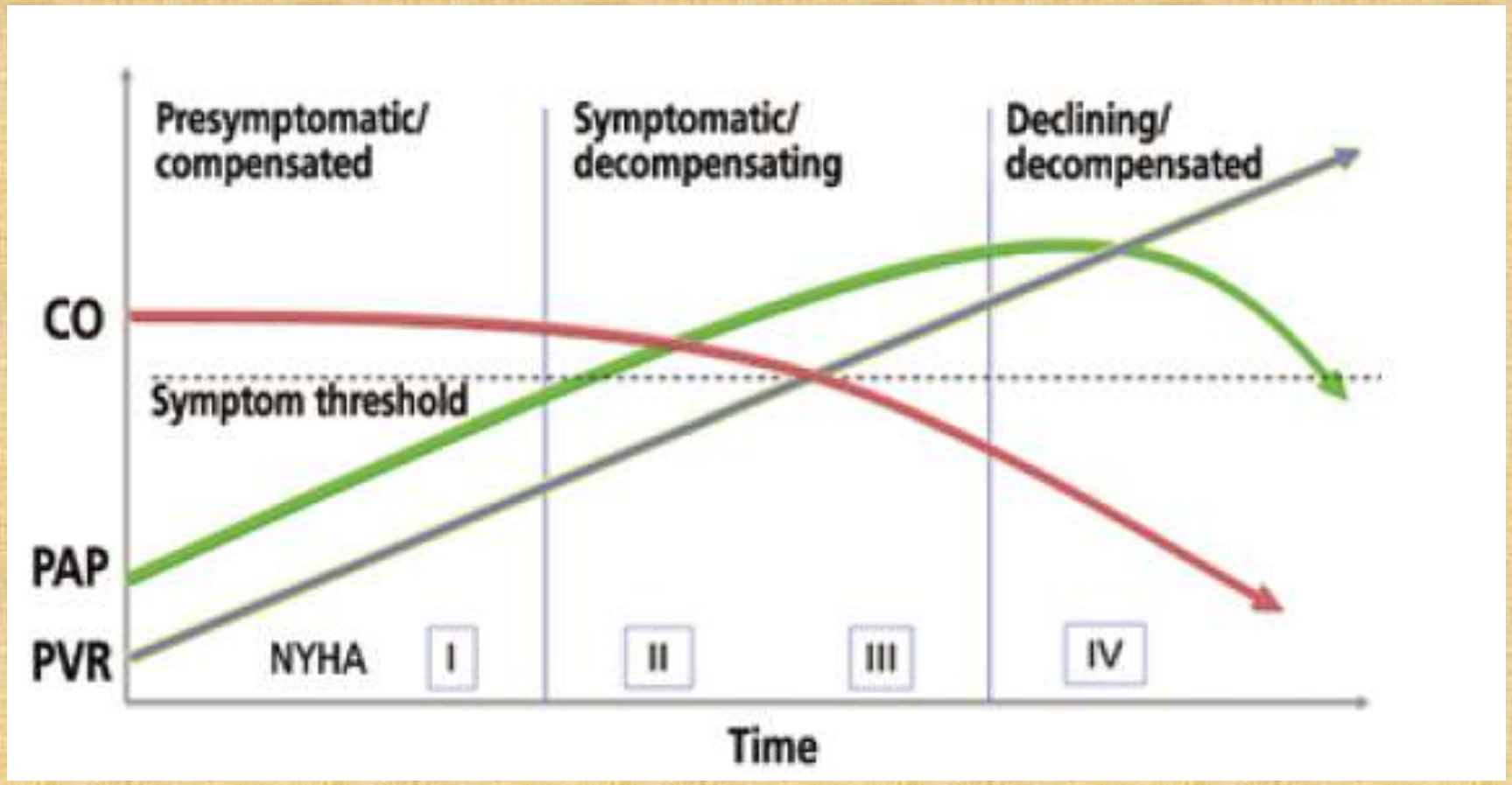
DOI: 10.1513/AnnalsATS.201312-443LD

Progression of vascular disease

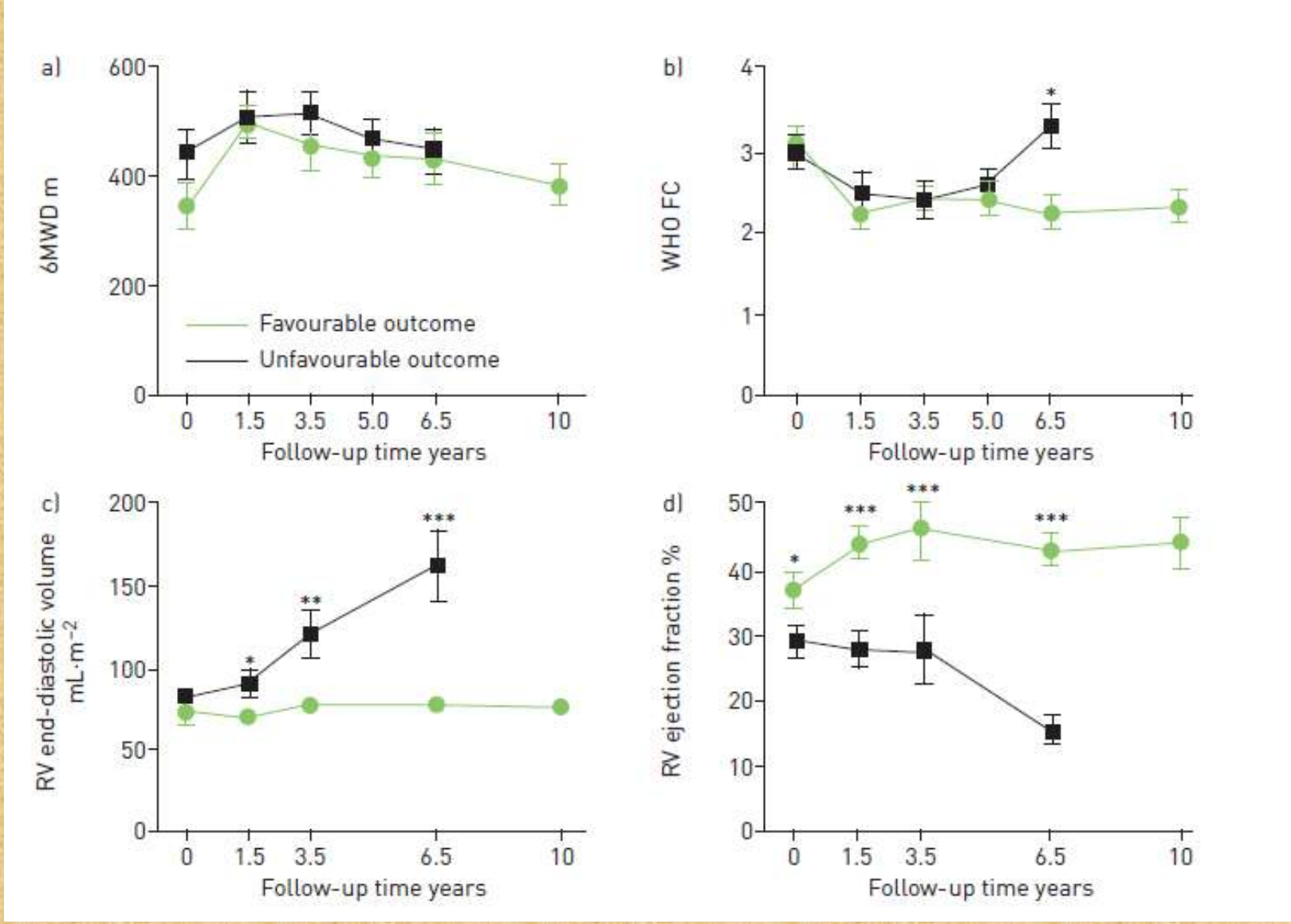


Hunter C. Champion, Evangelos D. Michelakis and Paul M. Hassoun

Circulation 2009;120;992-1007



Bhatnagar A et al. *Clev Cl J Med* 2018; 85(6)



Van der Veerdonk et al. *Chest* 2015;147:1063-71

CHEST

Original Research

PULMONARY VASCULAR DISEASE

Delay in Recognition of Pulmonary Arterial Hypertension

Factors Identified From the REVEAL Registry

CHEST 2011; 140(1):19–26

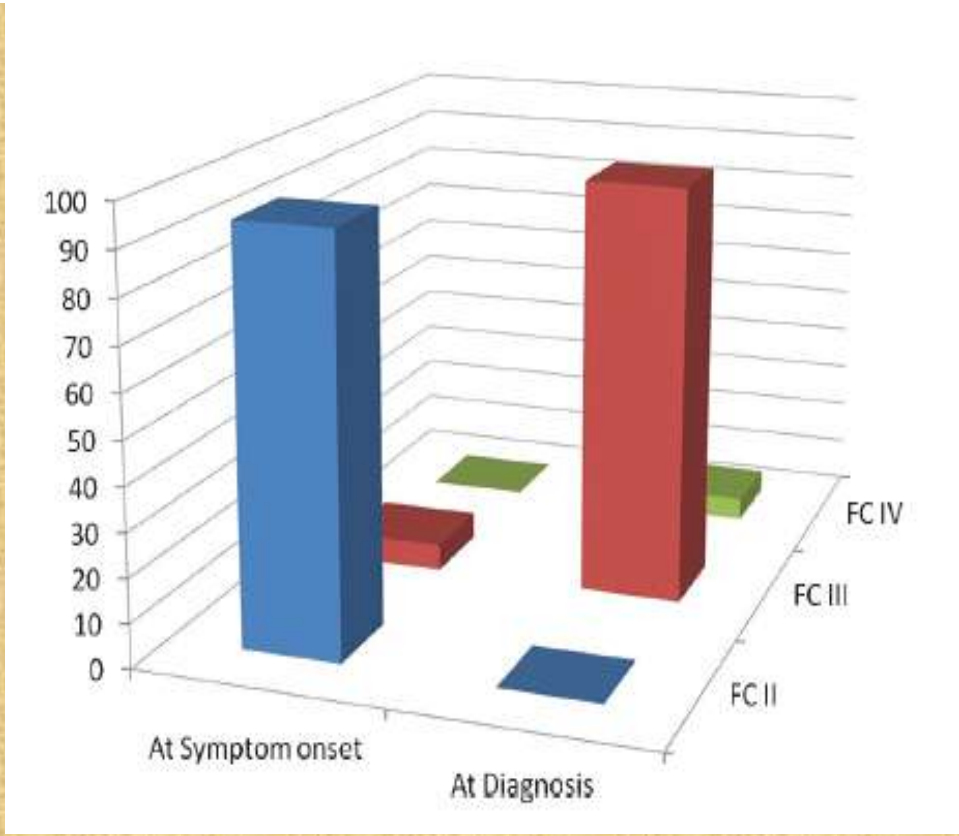
- 2493 pts from REVEAL registry
- Mean time from PAH symptoms onset to RHC:1.1 y
- Factors associated with >2 y interval (21%)
- Younger age, other resp disease
- Less severe RV dysfunction
- 6MWD<250m

Research Article

Time from symptoms to definitive diagnosis of idiopathic pulmonary arterial hypertension: The delay study

Geoff Strange[1], Eli Gabbay[2], Fiona Kermeen[3], Trevor Williams[4], Melinda Carrington[5], Simon Stewart[5], and Anne Keogh[6]

Pulmonary Circulation | January-March 2013 | Vol 3 | No 1



- Median: 44mo from symptom onset to diagnosis
- Alternative diagnosis:asthma

Risk assessment, prognosis and guideline implementation in pulmonary arterial hypertension

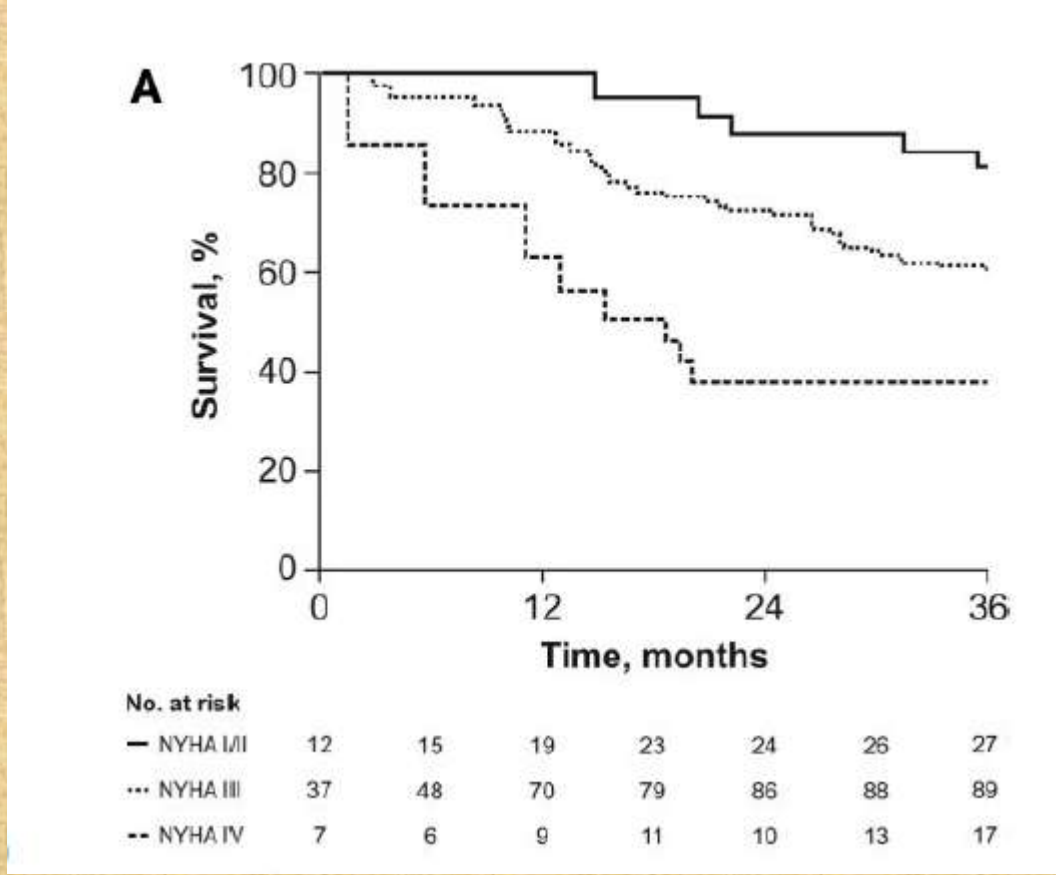
Athénaïs Boucly[1,2,3], Jason Weatherald[2,3,4], Laurent Savale[1,2,3], Xavier Jaïs[1,2,3], Vincent Cottin[5], Grégoire Prevot[6], François Picard[7], Pascal de Groote[8], Mitja Jevnikar[1,2,3], Emmanuel Bergot[9], Ari Chaouat[10,11], Céline Chabanne[12], Arnaud Bourdin[13], Florence Parent[1,2,3], David Montani[1,2,3], Gérald Simonneau[1,2,3], Marc Humbert[1,2,3] and Olivier Sitbon[1,2,3]

Eur Respir J 2017; 50: 1700889

TABLE 1 Demographics and baseline characteristics of overall and study populations of patients with newly diagnosed idiopathic, heritable and drug-induced pulmonary arterial hypertension (PAH)

	Overall population	Study population
Subjects	1591	1017
Female/male	924 (58)/667 (42)	598 (59)/419 (41)
Age years	60±17	57±17
BMI kg·m^{-2}	27.8±7.0	27.7±6.4
BMI >30 kg·m^{-2}	474 (30)	291 (29)
PAH diagnosis		
Idiopathic	1228 (77)	762 (75)
Heritable	109 (7)	94 (9)
Drug-induced	254 (16)	161 (16)
Acute vasodilator responder	139 (8.7)	97 (9.5)
WHO/NYHA functional class		
I–II	441 (28)	261 (26)
III	925 (58)	624 (61)
IV	225 (14)	132 (13)

WHO-FC and mortality



FC-II: 20% mortality in 3 ys

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

Table 2. Findings Suggestive of Pulmonary Hypertension

History
Angina
Dyspnea on exertion
Exercise intolerance
Fatigue
History of medical illness associated with pulmonary hypertension
Syncope or presyncope
Physical examination
Abnormal pulse oximetry, elevated jugular venous pressure, ascites, lower extremity edema, right ventricular heave, tricuspid regurgitation murmur (increased pulmonic component of S2)
Signs of right heart failure
Office-based diagnostic tests
Chest radiography: right ventricular enlargement, engorged pulmonary arteries
Electrocardiography: right ventricular enlargement, right bundle branch block, right ventricular strain pattern, S1Q3T3 pattern
Laboratory studies: elevated brain natriuretic peptide level

Dunlap B et al. *Am Fam Phys*, 94 (6);2016

Συμπτώματα και σημεία

- Δυσλειτουργία της δεξιάς κοιλίας
- Δύσπνοια στην κόπωση
- Αδυναμία, κ
- Επεισόδια εμ
- Οιδήματα κά
- Περικαρδιακ
- Συγκοπτικά
- Μηχανικές επιπλοκές

- Ασθενείς με συμπτώματα δυσανάλογα με την υποκείμενη νόσο
- Απουσία ανταπόκρισης στη θεραπεία

ERS/ESC Guidelines for the diagnosis and treatment of pulmonary hypertension 2015
Bhatnagar A et al. *Clev Cl J Med* 2018; 85(6)

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Dunlap B et al. *Am Fam Phys*, 94 (6);2016

RIGHT SIDED ♥ FAILURE

(Cor Pulmonale)

- Fatigue
- ↑ Peripheral Venous Pressure
- Ascites
- Enlarged Liver & Spleen

- May be secondary to chronic pulmonary problems
- Distended Jugular Veins
- Anorexia & Complaints of GI Distress
- Weight Gain
- Dependent Edema

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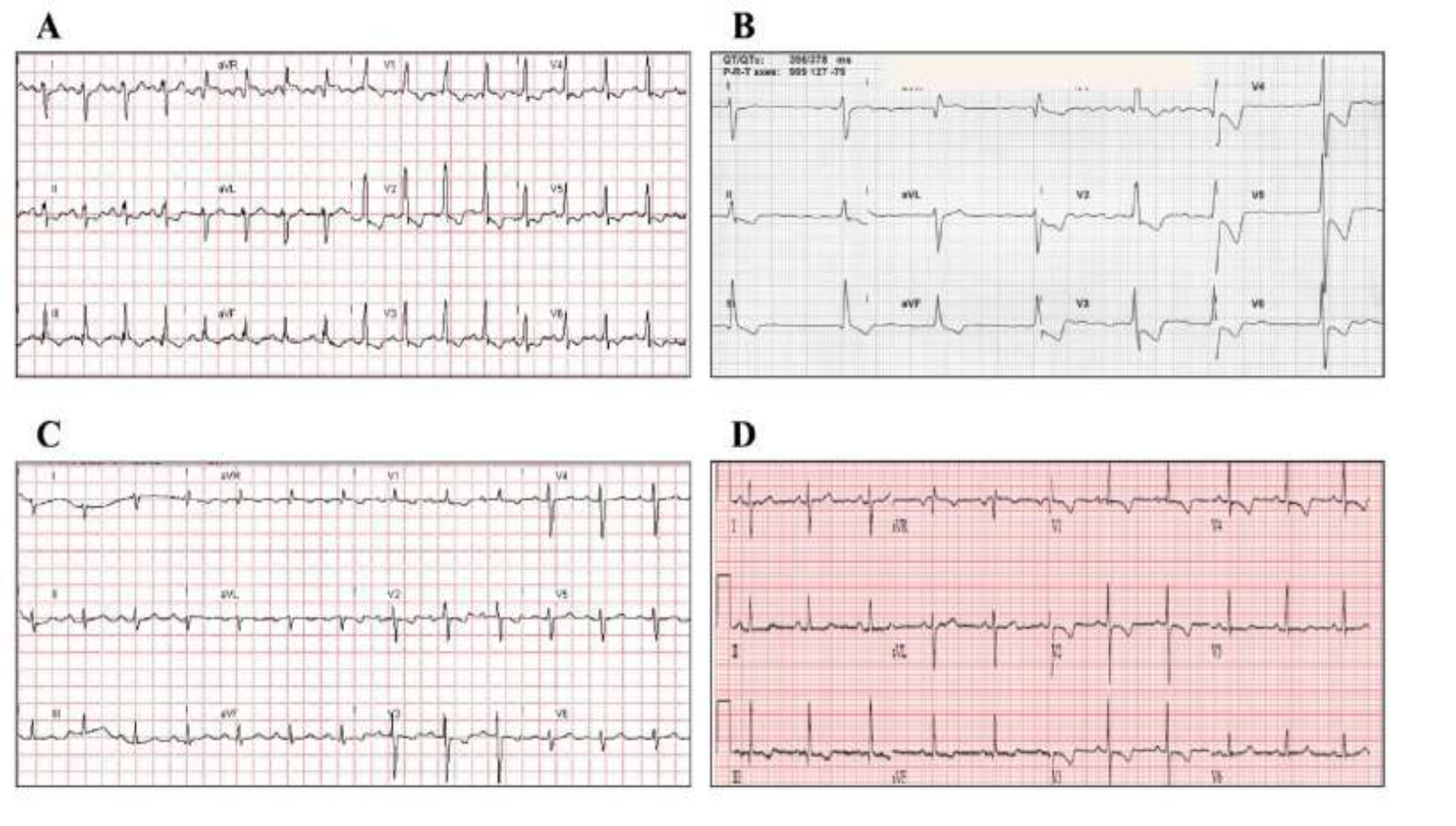
Dunlap B et al. *Am Fam Phys*, 94 (6);2016

Διαγνωστική προσέγγιση

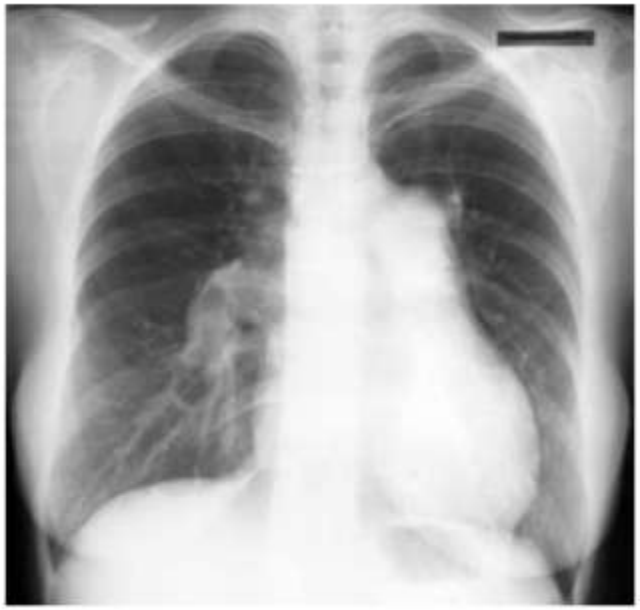
- **ΗΚΓ:** Το φυσιολογικό δεν αποκλείει τη διάγνωση



Διαγνωστική προσέγγιση

- Ακτινογραφία θώρακος. Αποκλεισμός αναπνευστικής νόσου (Group III) ή πνευμονικού οιδήματος (Group II)

Pulmonary Hypertension

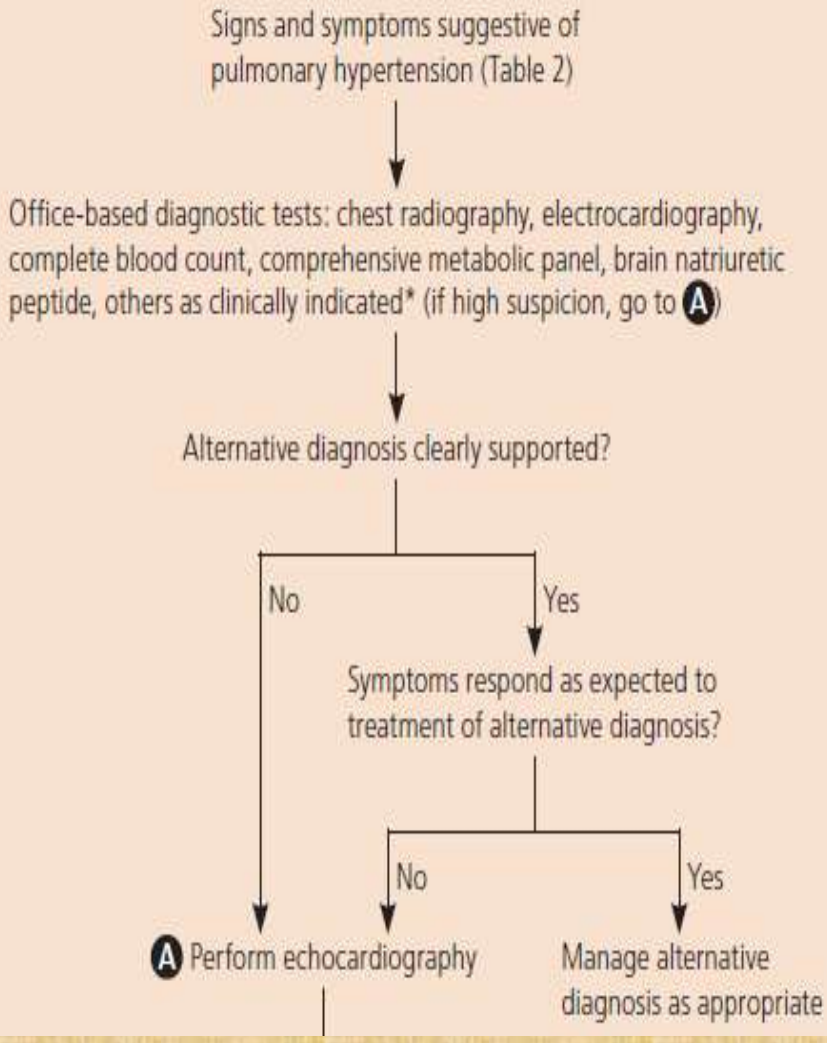


- Pulmonary hypertension. Chest radiograph in a patient with secondary pulmonary hypertension reveals enlarged pulmonary arteries. This patient was found to have an atrial septal defect.

Λοιπές εργαστηριακές εξετάσεις

- PFTs: Υποκείμενη νόσος παρεγχύματος ή αεραγωγών
- Ήπια – μέτρια μείωση των πνευμονικών όγκων
- Μείωση DLCO: PVOD, SSc, παρεγχυματική νόσος
- DLCO<45%: πτωχή πρόγνωση
- Νυχτερινή οξυμετρία
- Πολυσωματογραφική μελέτη ύπνου
- Brain natriuretic peptide

Evaluation of Suspected Pulmonary Hypertension in Primary Care



Dunlap B et al. *Am Fam Phys*, 94 (6);2016

Σύνθετα κλινικά σενάρια

- Ασυμπτωματικοί ασθενείς που ανήκουν σε ομάδες υψηλού κινδύνου (screening)
- Ασθενείς με νόσημα αναπνευστικού και δυσανάλογη αγγειακή πνευμονική νόσο που θα μπορούσαν να ωφεληθούν από ειδική αγωγή ΠΥ

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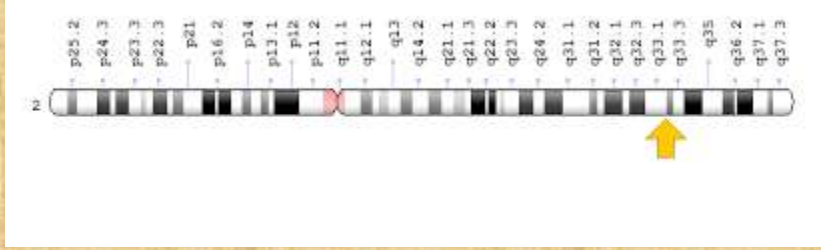
PAH: pulmonary arterial hypertension; PVOD: pulmonary veno-occlusive disease; PCH: pulmonary capillary haemangiomatosis; LVEF: left ventricular ejection fraction.

Simonneau G et al. *Eur Resp J* 2018

Κληρονομική ΠΑΥ



- Ετερόζυγος BMPR2 (*bone morphogenetic protein receptor-II gene)* μετάλλαξη
- 75% οικογενών περιπτώσεων ΠΑΥ
- 25% σποραδικών
- Κίνδυνος ανάπτυξης ΠΑΥ σε ασυμπτωματικούς φορείς: 14% σε άνδρες-42% σε γυναίκες
- Δυνατότητα γενετικού ελέγχου σε συγγενείς ασθενών
- Ετησίως echo σε ασυμπτωματικούς φορείς

Girerd B et al. *Current opinion in pulmonary medicine*. 2017;23(5):386-91.

TABLE 3 Updated classification of drugs and toxins associated with PAH

Definite	Possible
Aminorex	Cocaine
Fenfluramine	Phenylpropanolamine
Dexfenfluramine	L-tryptophan
Benfluorex	St John's wort
Methamphetamines	Amphetamines
Dasatinib	Interferon-α and -β
Toxic rapeseed oil	Alkylating agents
	Bosutinib
	Direct-acting antiviral agents against hepatitis C virus
	Leflunomide
	Indirubin (Chinese herb Qing-Dai)

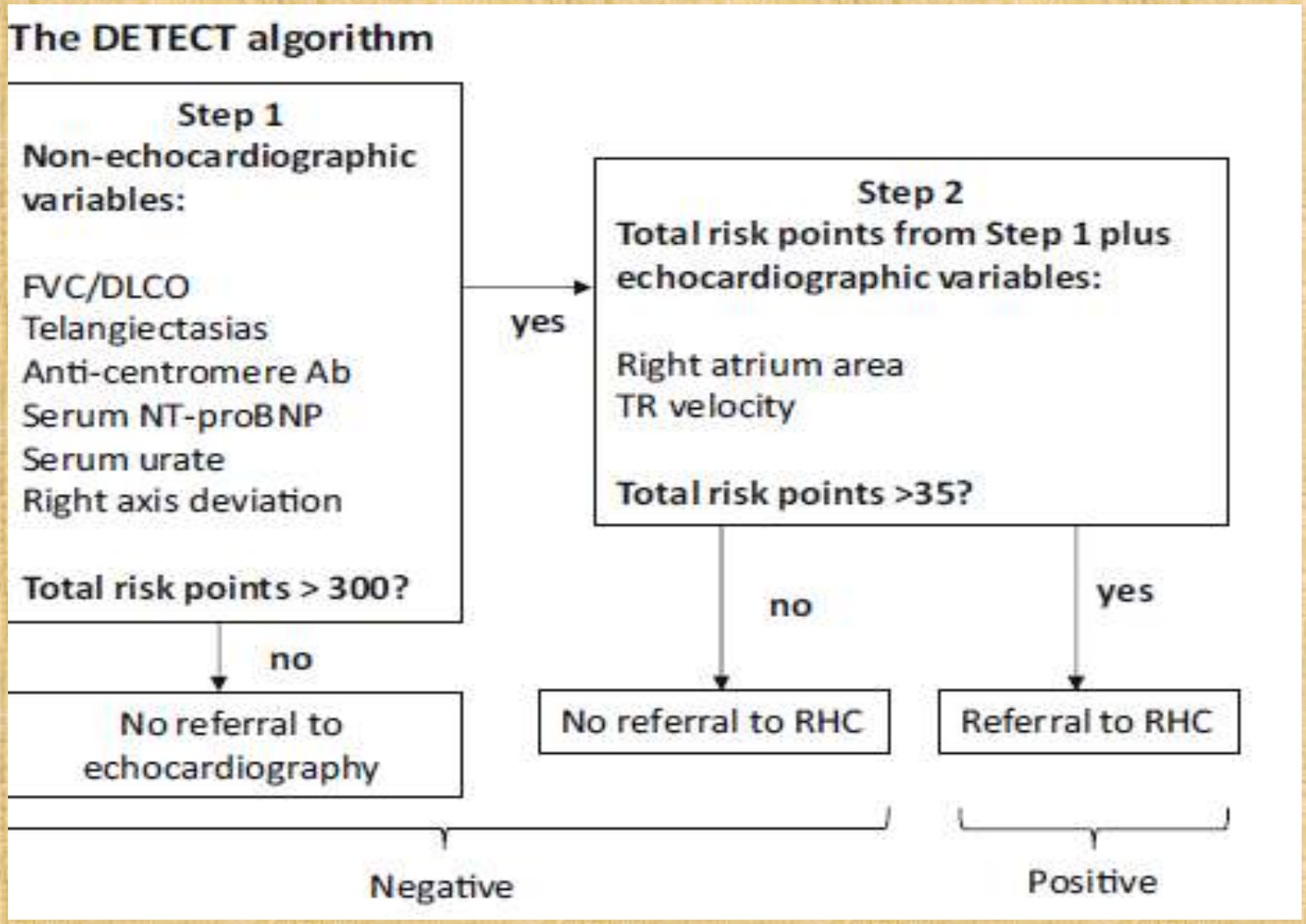
Simonneau G et al. *Eur Resp J* 2018

ΠΥ συνδεόμενη με νόσο του συνδετικού ιστού

- **Σκληρόδερμα** – ΠΥ 5-25% ανάλογα με τη μελέτη
  Σημαντική αιτία νοσηρότητας και θνητότητας
  Η συχνότερη αιτία θανάτου
- **ΣΕΛ** – ΠΥ 1-14% (Ασία, Κίνα)
  Τρίτη αιτία θανάτου
  Ανεξάρτητος παράγοντας θνητότητας
- **Μεικτή νόσος του συνδετικού ιστού**

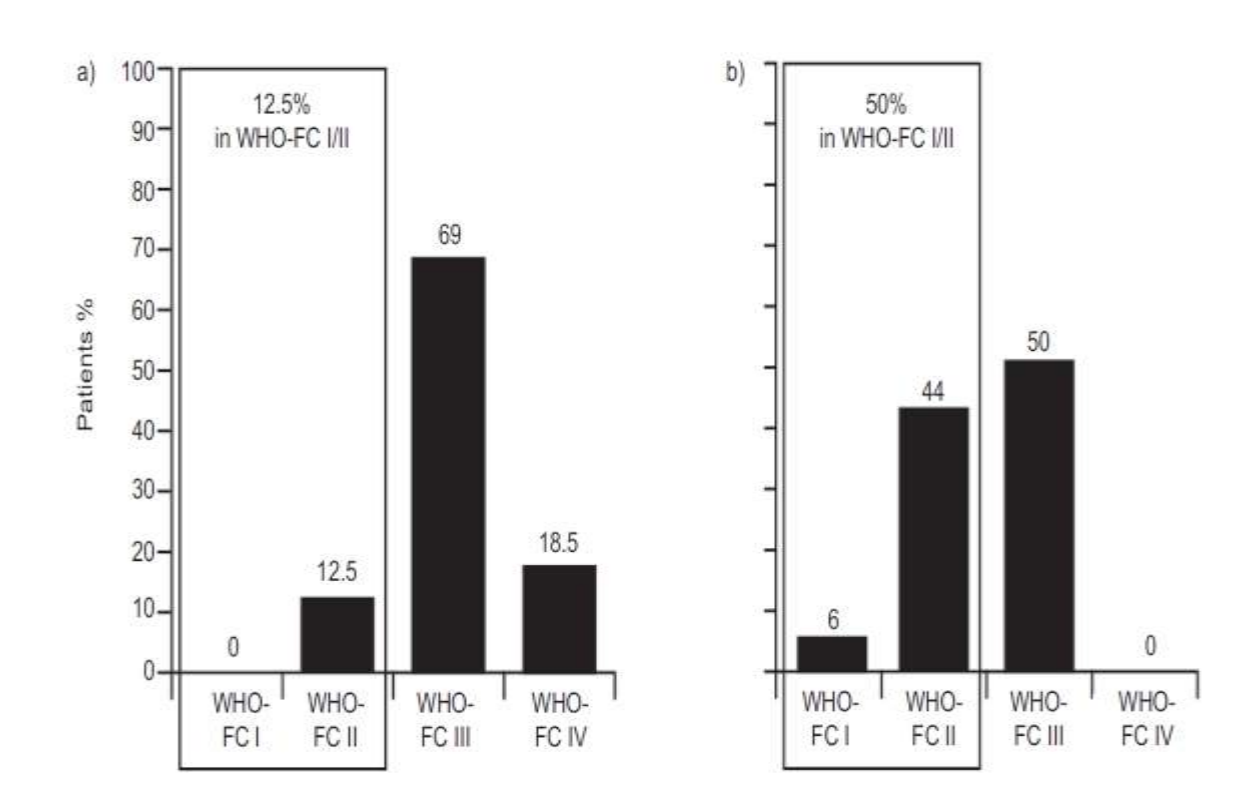
TTE, DETECT ετησίως

Denton CP et al. *Nat Rev Rheym*. 2018, 19 (2)
HK Min, JH Lee et al. *Korean J Int Med*, 2015;30,232-245



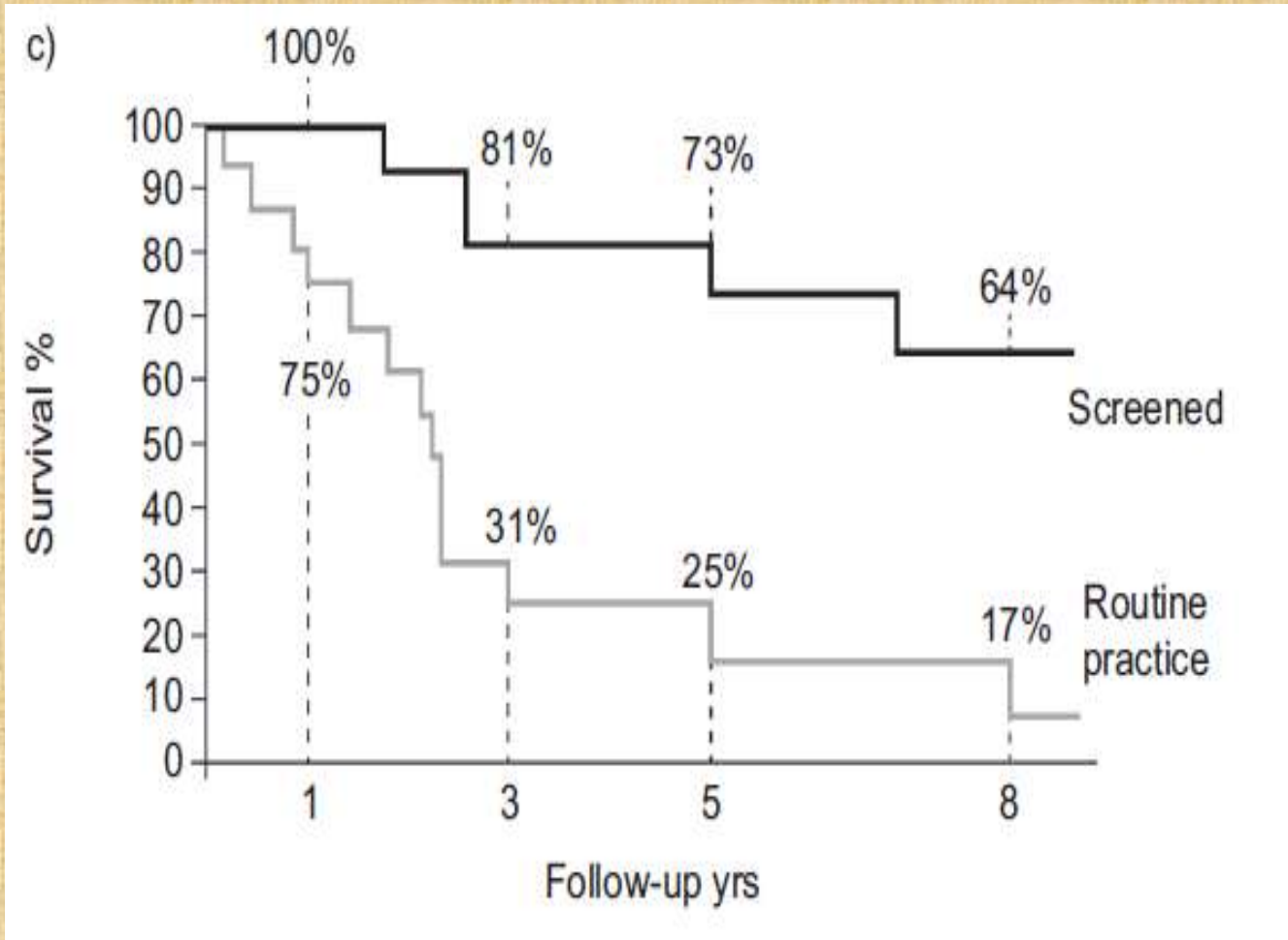
Coghlan JG et al. Annals of the rheumatic diseases. 2014;73(7):1340-9.

WHO-FC at the time of diagnosis is different if a screening program is used



Marc Humbert*, et al. *Eur Respir Rev* 2012; 21: 126, 306–312

Survival in PAH/SSc patients is better if a a screening program is used



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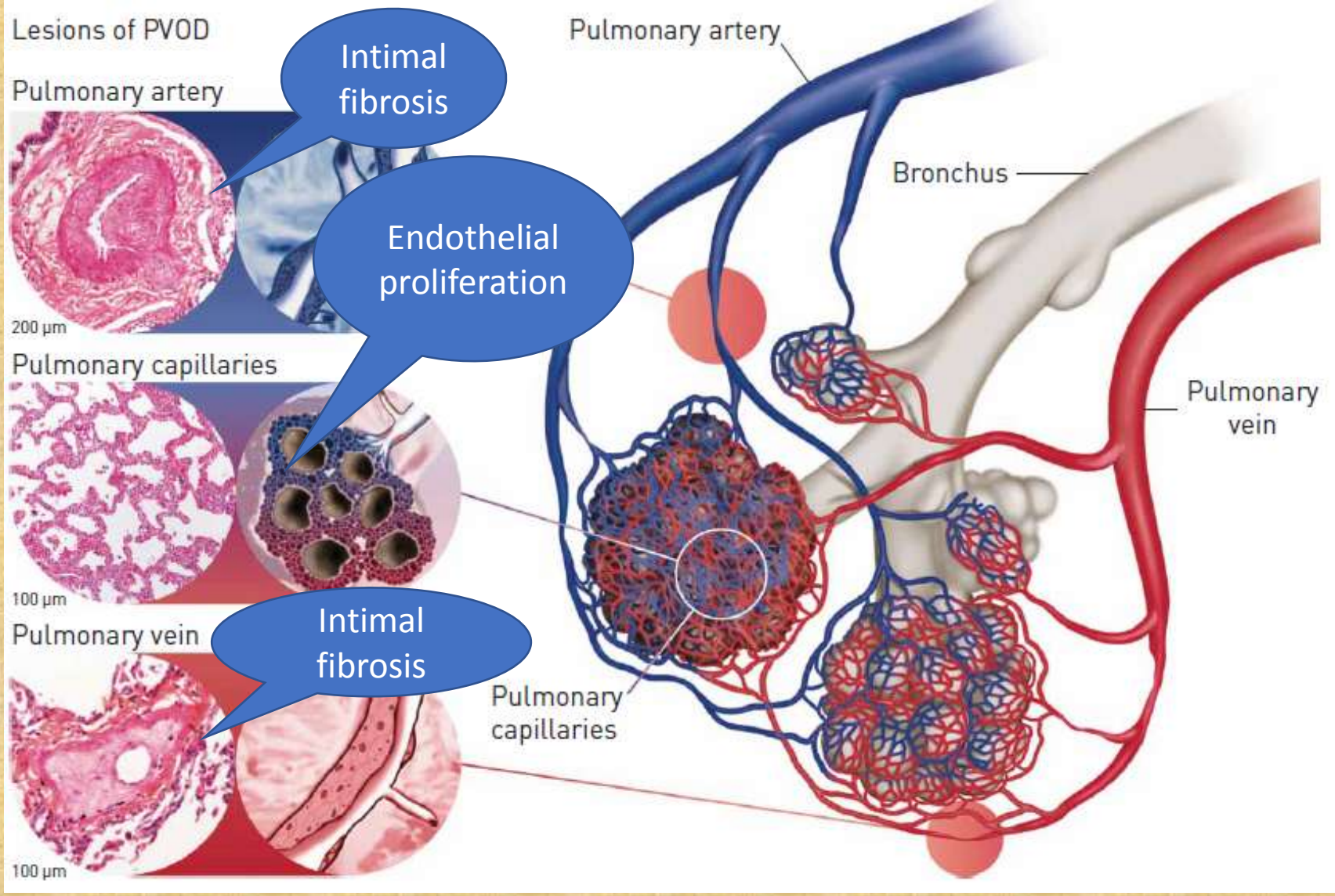
Συμπτώματα
Iv drug
HCV

TTE
ΠΥ 2-6%

Frost A et al. *Eur Resp J* 2018

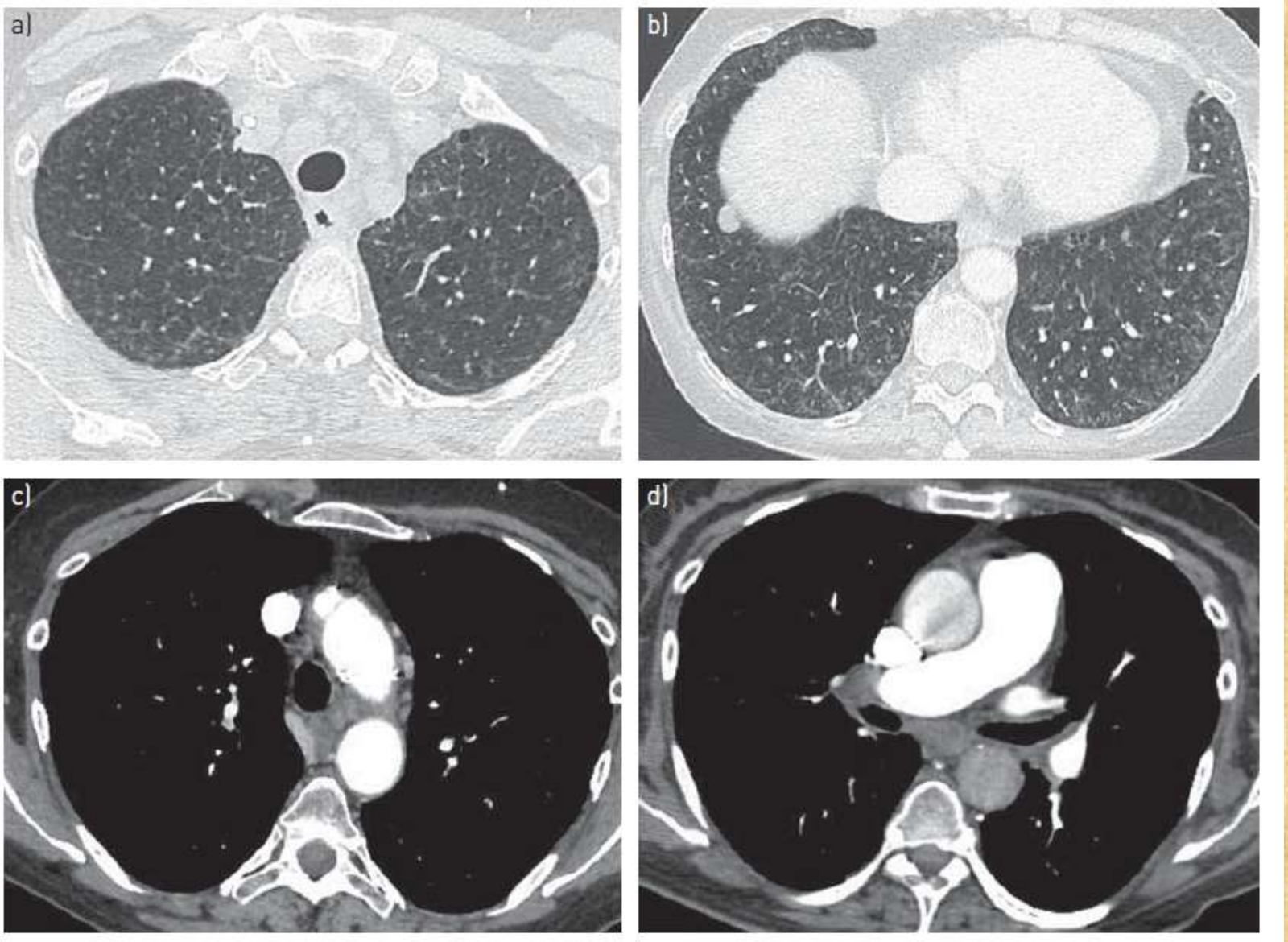


Montani D et al. *ERJ 2016*;47, 1518-1534

Πότε υποπτευόμασε PVOD

- Very low DLCO
- Hypoxemia
- Severe desaturation on 6-min walking test
- 2 of 3 characteristic radiological signs on HRCT (thickening of interlobular septa, ground glass opacities, lymph node enlargement)
- Occult alveolar haemorrhage (post capillary block)

Montani D et al. *ERJ 2016*;47, 1518-1534



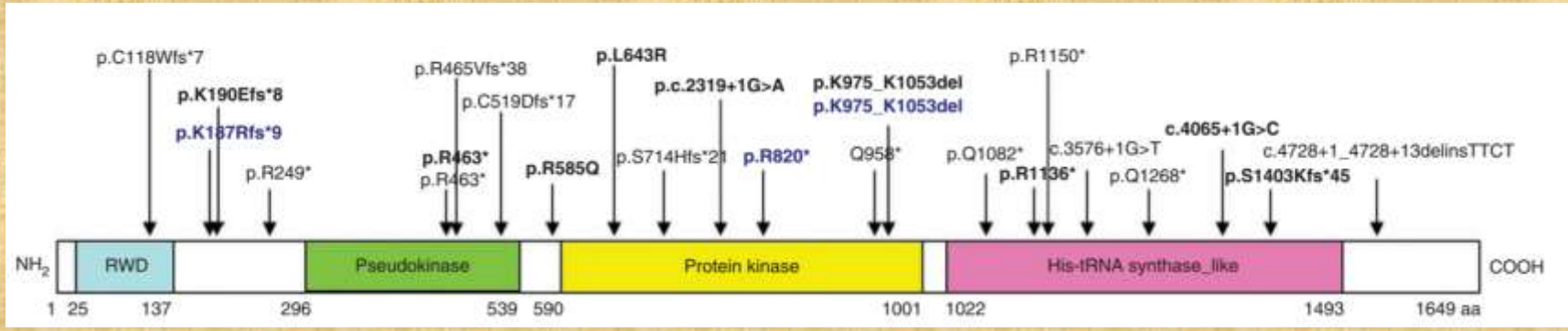
Montani D *et al. Eur Respir J* 2016

Heritable PVOD

EIF2AK4 mutations cause pulmonary veno-occlusive disease, a recessive form of pulmonary hypertension

Mélanie Eyries[1–3], David Montani[4–6], Barbara Girerd[4–6], Claire Perret[3,7], Anne Leroy[2], Christine Lonjou[8], Nadjim Chelghoum[8], Florence Coulet[2,3], Damien Bonnet[9,10], Peter Dorfmüller[6,11], Elie Fadel[6,12], Olivier Sitbon[4–6], Gérald Simonneau[4–6], David-Alexandre Tregouët[3,7], Marc Humbert[4–6] & Florent Soubrier[1–3]

nature genetics



Eyries et al. *Nat Genet* 2014

Κληρονομική μορφή PVOD

- Eukaryotic translation initiation factor 2 alpha kinase 4
- Activator of cellular stress response pathway
- Autosomal recessive transmission
- 100% of familial cases of PVOD
- 20-25% of sporadic cases
- Biallelic *EIF2AK4* in PH pt: PVOD

Hadinnapola C et al. *Circulation* 2017; 136:2022-2033

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- 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

PAH: pulmonary arterial hypertension; PVOD: pulmonary veno-occlusive disease; PCH: pulmonary capillary haemangiomatosis; LVEF: left ventricular ejection fraction.

Frost A et al. *Eur Resp J* 2018

CTEPH

- Group 4 of the present clinical classification
- Rare and late complication of PE
- Thrombi in proximal pulmonary arteries
- Small vessel disease
- “Honeymoon period”
- Incidence after symptomatic PE: 0.4-6.2% (3.4%)
- Systematic V/Q scanning after PE: Not recommended

Simonneau et al. *Eur Respir Rev* 2017;26

Σύνθετα κλινικά σενάρια

- Ασυμπτωματικοί ασθενείς που ανήκουν σε ομάδες υψηλού κινδύνου (screening)
- Ασθενείς με νόσημα αναπνευστικού και δυσανάλογη αγγειακή πνευμονική νόσο που θα μπορούσαν να ωφεληθούν από ειδική αγωγή ΠΥ

TABLE 2 Updated clinical classification of pulmonary hypertension (PH)

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
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Frost A et al. *Eur Resp J* 2018

ΠΥ σε ασθενείς με χρόνια αναπνευστικά νοσήματα

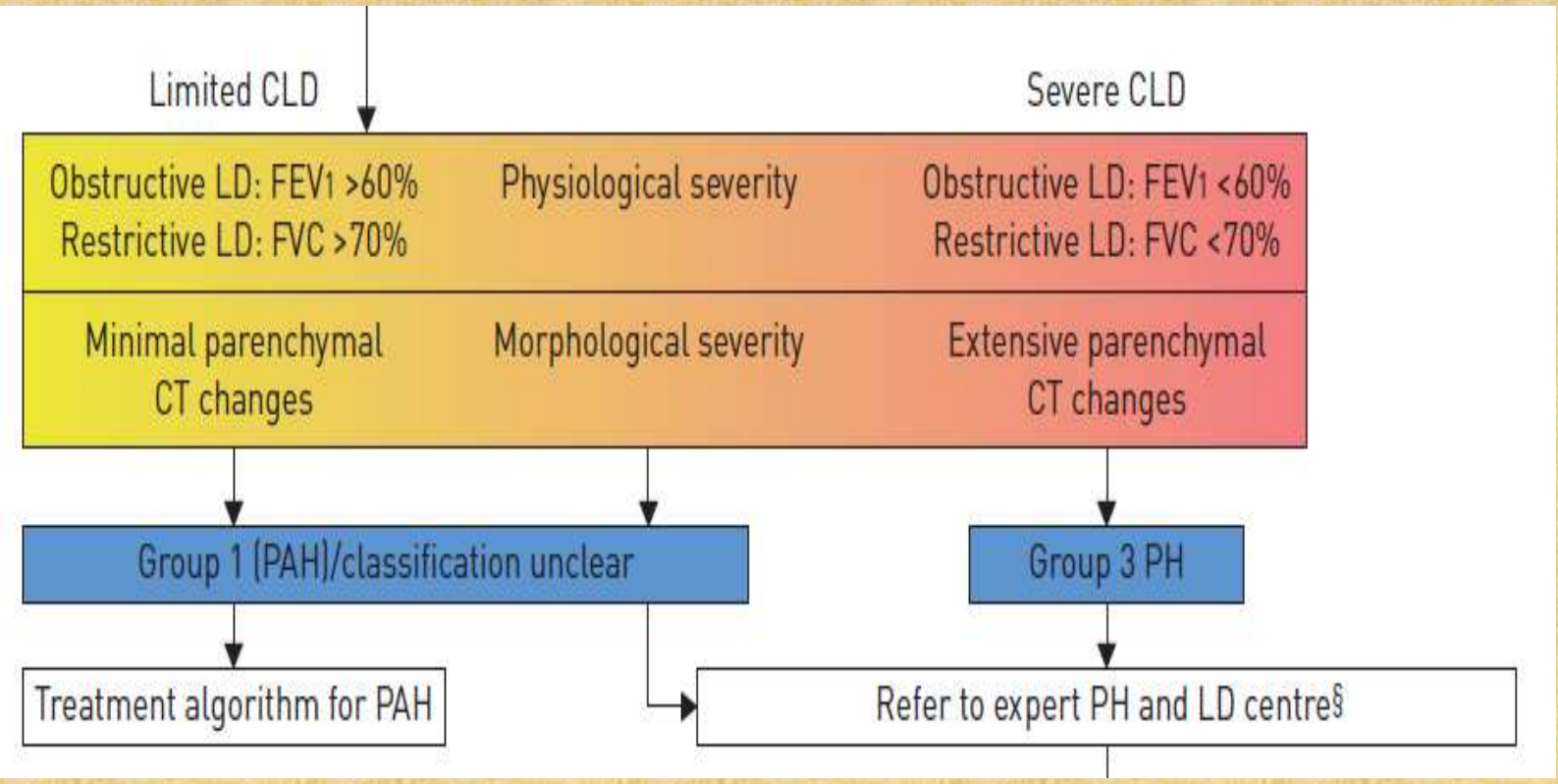
- IPF: mPAP>20 mmHg 8-15%
- COPD: ΠΥ 20-50%
- Στάδιο IV: mPAP>20 mmHg 90%
- Συσχέτιση με θνητότητα
- Pulmonary vascular COPD phenotype

  Ήπιος περιορισμός αεροροής/ ↓ DLCO

  Ήπια υποξαιμία/υπο ή νορμοκαπνία

  Επηρεασμένη καρδιοαναπνευστική άσκηση
- Παραπομπή σε ειδικά κέντρα-κλινικές μελέτες

Maron B, N.Galie; *JAMA Cardiol.2016*
Nathan S et al. *Eur Respir J*, December 2018



Nathan S et al. *Eur Respir J*, December 2018

Μηνύματα για το σπίτι…

- Η ΠΥ παραμένει μία νόσος με δυσμενή πρόγνωση
- Τα συμπτώματα είναι μη ειδικά – Δυσλειτουργία δεξιάς κοιλίας
- Η διάγνωση γίνεται σε προχωρημένο στάδιο της νόσου
- Οι συνοσηρότητες περιπλέκουν την εικόνα
- Παρακολούθηση ασθενών με παράγοντες κινδύνου
- Επιλογή των ασθενών με φαινότυπο σοβαρής πνευμονικής αγγειακής νόσου

Ευχαριστώ !!!