Πνευμονική Υπέρταση σχετιζόμενη με Νοσήματα του συνδετικού ιστού

πρώιμη διάγνωση
και
έγκαιρη παρέμβαση

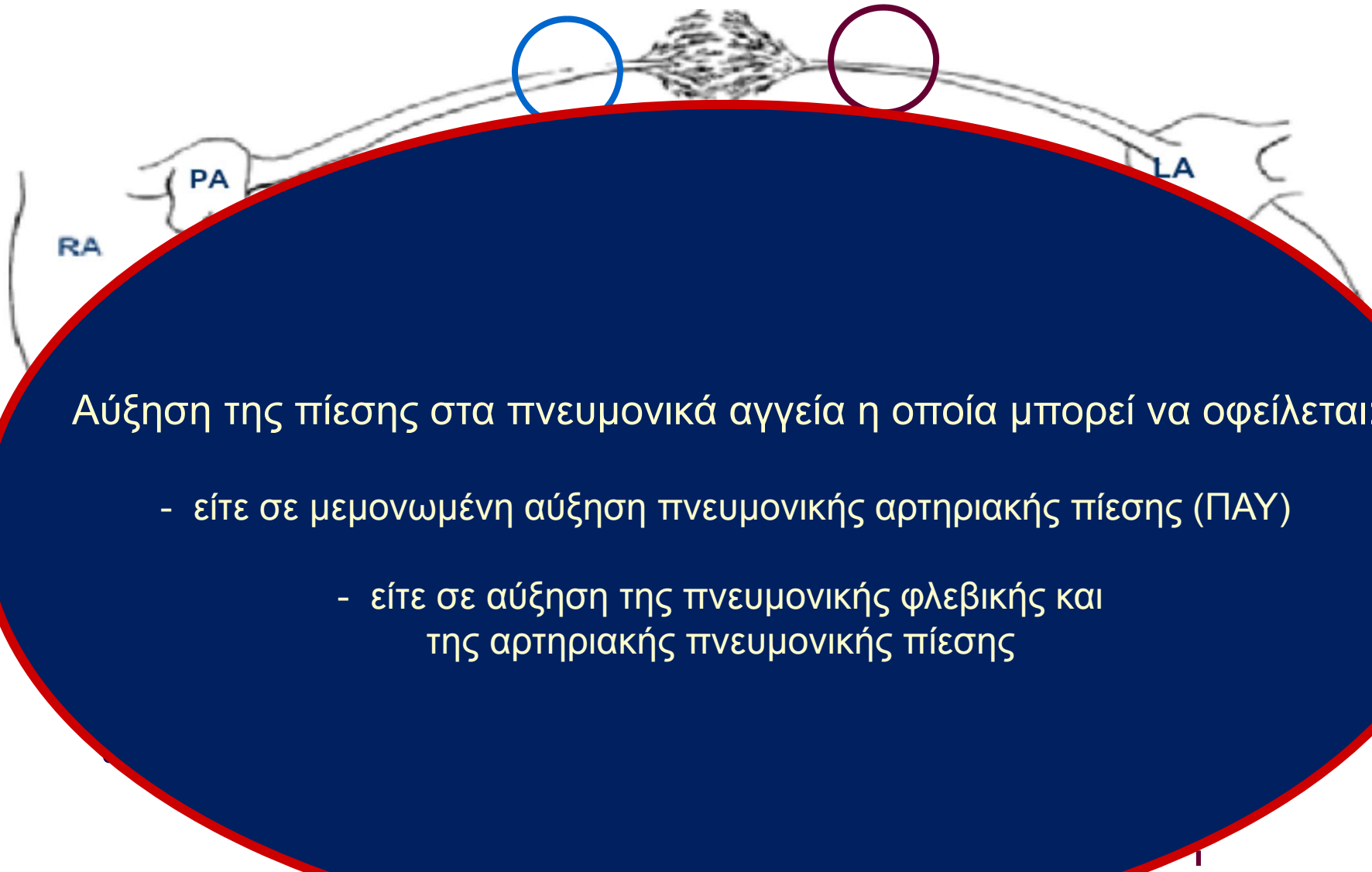
Ι Μητρούσκα
Πνευμονολογική Κλινική
Πανεπιστημιακό Νοσοκομείο Κρήτης

Πνευμονική Υπέρταση

Ορισμός

Αύξηση της πίεσης στα Πνευμονικά αγγεία

Current hemodynamic classification of PH *Define the lesion*



Αύξηση της πίεσης στα πνευμονικά αγγεία η οποία μπορεί να οφείλεται:

- είτε σε μεμονωμένη αύξηση πνευμονικής αρτηριακής πίεσης (ΠΑΥ)

- είτε σε αύξηση της πνευμονικής φλεβικής και της αρτηριακής πνευμονικής πίεσης

Reactive

Ppa-Pcwp ≤12 mmHg

Ppa-Pcwp >12 mmHg

Adapted from Galiè N, et al. Eur Heart J 2009;30:2493–537; Galiè N, et al. Eur Respir J 2009;34:1219–63.

Πνευμονική Υπέρταση
Ορισμός

- Πνευμονική Υπέρταση ορίζεται:
- **η αύξηση της μέσης Πνευμονικής Αρτηριακής Πίεσης (mPAP) σε τιμές ≥ 25 mmHg,**
  - όταν αυτή μετράται **με Καθετηριασμό Δεξιών Καρδιακών Κοιλοτήτων**

Αιμοδυναμικός
και όχι
Κλινικός ή υπερηχογραφικός ορισμός

Current hemodynamic classification of PH *Define the lesion*

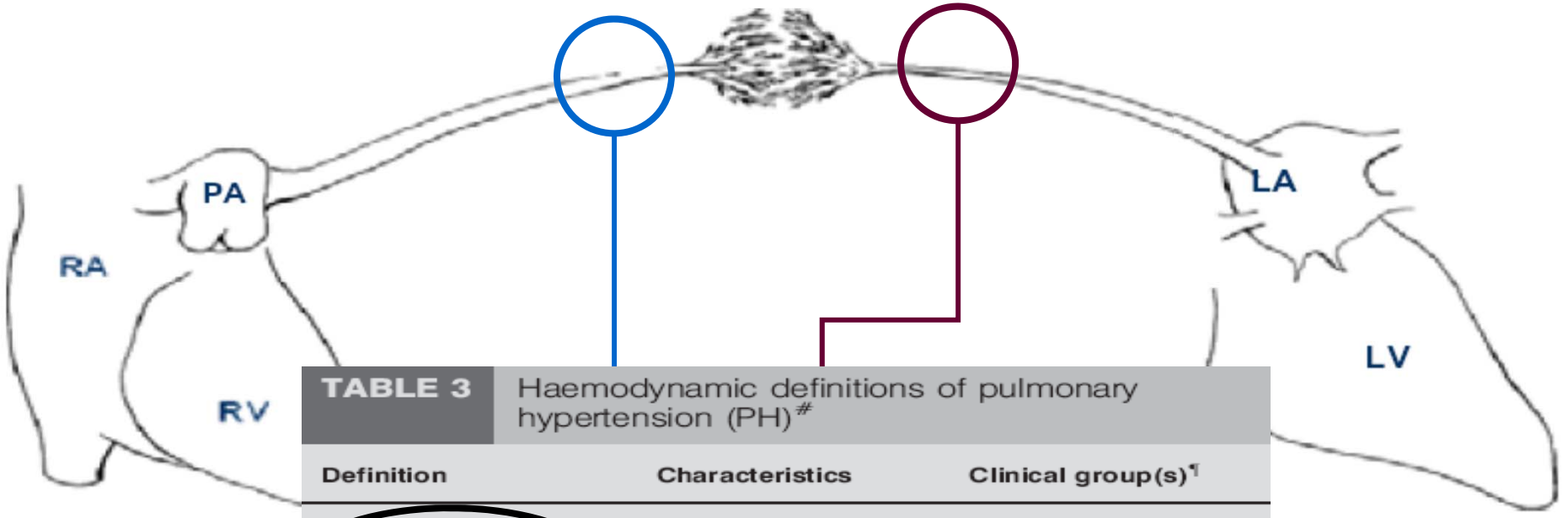


TABLE 3 Haemodynamic definitions of pulmonary hypertension (PH)#

Definition	Characteristics	Clinical group(s)¶
PH	$\bar{P}_{pa}$ ⩾25 mmHg	All
Pre-capillary PH	$\bar{P}_{pa}$ ⩾25 mmHg	1. Pulmonary arterial hypertension
	P_{pcw} ⩽15 mmHg	3. PH due to lung diseases
	CO normal or reduced+	4. Chronic thromboembolic PH
		5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH	$\bar{P}_{pa}$ ⩾25 mmHg	2. PH due to left heart disease
	P_{pcw} >15 mmHg	
	CO normal or reduced+	
Passive	TPG ⩽12 mmHg	
Reactive (out of proportion)	TPG >12 mmHg	

Pre-cap **PH**

- Ppa ≥2
- Pcwp <
- N or rec
- All but

active

o >12 mmHg

Adapt 63.

Table 1 Updated Classification of Pulmonary Hypertension*

1. Pulmonary arterial hypertension
   - 1.1 Idiopathic PAH
   - 1.2 Heritable PAH
   - 1.2.1 BMPR2
   - 1.2.2 ALK-1, ENG, SMAD9, CAV1, KCNK3
   - 1.2.3 Unknown
   - 1.3 Drug and toxin induced
   - 1.4 Associated with:
   - 1.4.1 Connective tissue disease
   - 1.4.2 HIV infection
   - 1.4.3 Portal hypertension
   - 1.4.4 Congenital heart diseases
   - 1.4.5 Schistosomiasis

1′ Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis

1″. Persistent pulmonary hypertension of the newborn (PPHN)

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

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

4. Chronic thromboembolic pulmonary hypertension (CTEPH)

5. Pulmonary hypertension with unclear multifactorial mechanisms
   - 5.1 Hematologic disorders: chronic hemolytic anemia, 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: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH

*5th WSPH Nice 2013. Main modifications to the previous Dana Point classification are in **bold**.

BMPR = bone morphogenic protein receptor type II; CAV1 = caveolin-1; ENG = endoglin; HIV = human immunodeficiency virus; PAH = pulmonary arterial hypertension.

Ταξινόμηση Πνευμονικής Υπέρτασης

The Three-Year Incidence of Pulmonary Arterial Hypertension Associated With Systemic Sclerosis in a Multicenter Nationwide Longitudinal Study in France

Eric Hachulla,[1] Pascal de Groote,[2] Virginie Gressin,[3] Jean Sibilia,[4] Elisabeth Diot,[5] Patrick Carpentier,[6] Luc Mouthon,[7] Pierre-Yves Hatron,[1] Patrick Jego,[8] Yannick Allanore,[7] Kiet Phong Tiev,[9] Christian Agard,[10] Anne Cosnes,[11] Daniela Cirstea,[12] Joël Constans,[13] Dominique Farge,[14] Jean-François Viallard,[15] Jean-Robert Harle,[16] Frédéric Patat,[17] Bernard Imbert,[6] André Kahan,[7] Jean Cabane,[9] Pierre Clerson,[18] Loïc Guillevin,[7] Marc Humbert,[19] and the ItinérAIR-Sclérodermie Study Group

Table 2. Estimated incidence of pulmonary hypertension during the 3-year followup period*

	Estimated incidence (no. of cases per 100 patient-years)	95% CI
All forms of pulmonary hypertension	1.37	0.74–2.00
Pulmonary arterial hypertension	0.61	0.26–1.20
Among patients with lcSSc	0.40	0.11–1.03
Among patients with dcSSc	1.25	0.34–3.20
Postcapillary pulmonary hypertension	0.61	0.26–1.20
Pulmonary hypertension secondary to pulmonary fibrosis	0.15	0.02–0.55

* 95% CI = 95% confidence interval; lcSSc = limited cutaneous systemic sclerosis; dcSSc = diffuse cutaneous systemic sclerosis.

Hachulla et al. Arthritis Rheum 2009

Table 1 Updated Classification of Pulmonary Hypertension*

1. Pulmonary arterial hypertension
   - 1.1 Idiopathic PAH
   - 1.2 Heritable PAH
   - 1.2.1 BMPR2
   - 1.2.2 ALK-1, ENG, SMAD9, CAV1, KCNK3
   - 1.2.3 Unknown
   - 1.3 Drug and toxin induced
   - 1.4 Associated with:
   - 1.4.1 Connective tissue disease
   - 1.4.2 HIV infection
   - 1.4.3 Portal hypertension
   - 1.4.4 Congenital heart diseases
   - 1.4.5 Schistosomiasis

1′ Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis

1″. Persistent pulmonary hypertension of the newborn (PPHN)

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

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

4. Chronic thromboembolic pulmonary hypertension (CTEPH)

5. Pulmonary hypertension with unclear multifactorial mechanisms
   - 5.1 Hematologic disorders: chronic hemolytic anemia, 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: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH

*5th WSPH Nice 2013. Main modifications to the previous Dana Point classification are in **bold**.

BMPR = bone morphogenic protein receptor type II; CAV1 = caveolin-1; ENG = endoglin; HIV = human immunodeficiency virus; PAH = pulmonary arterial hypertension.

BMPR-2: bone morphogenetic receptor protein2 gene,

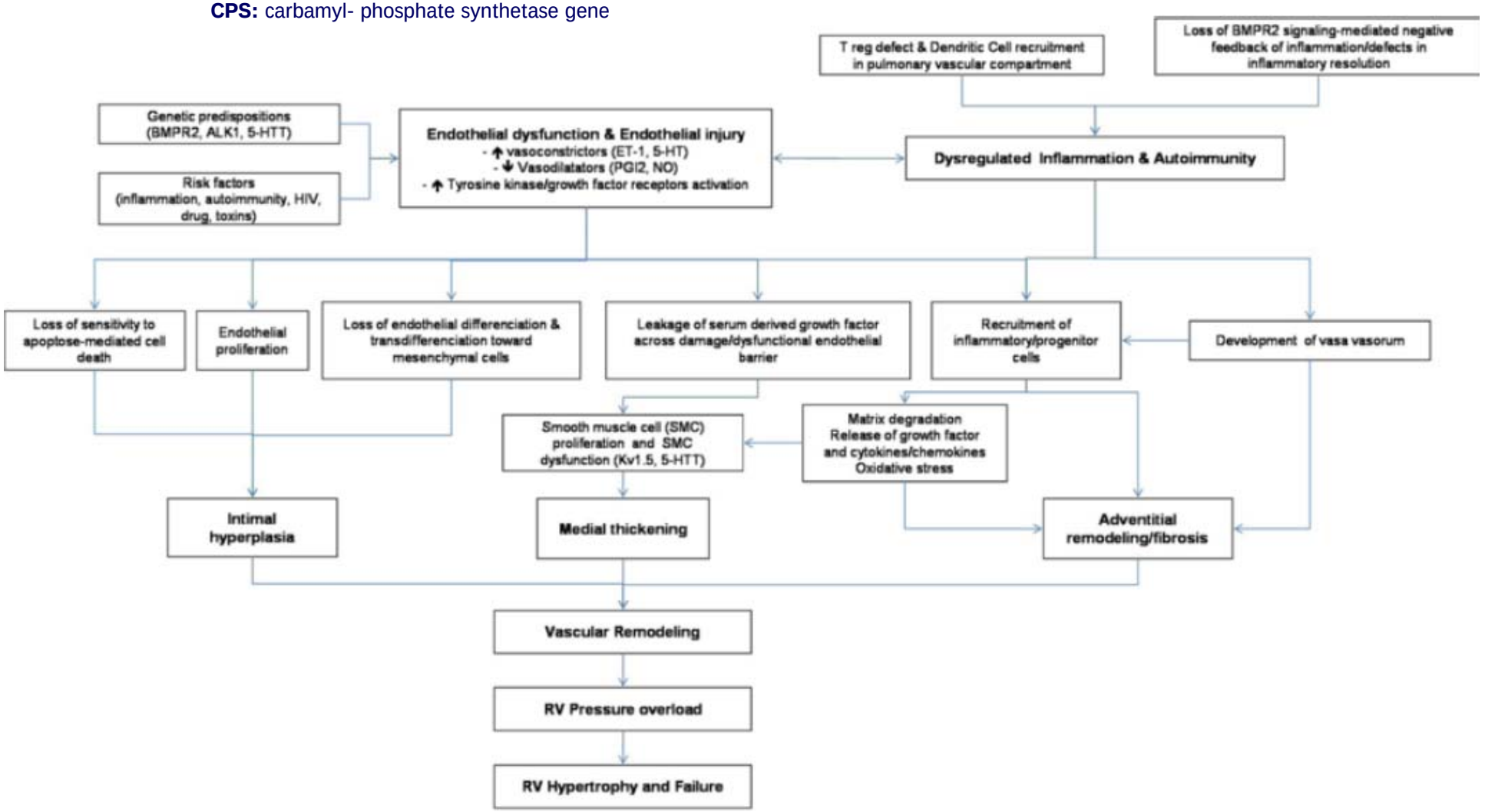
ALK 1: activin- receptor- like kinase 1 gene,

5-HTT: serotonin transporter gene,

ec-NOS: nitric oxide synthase gene;

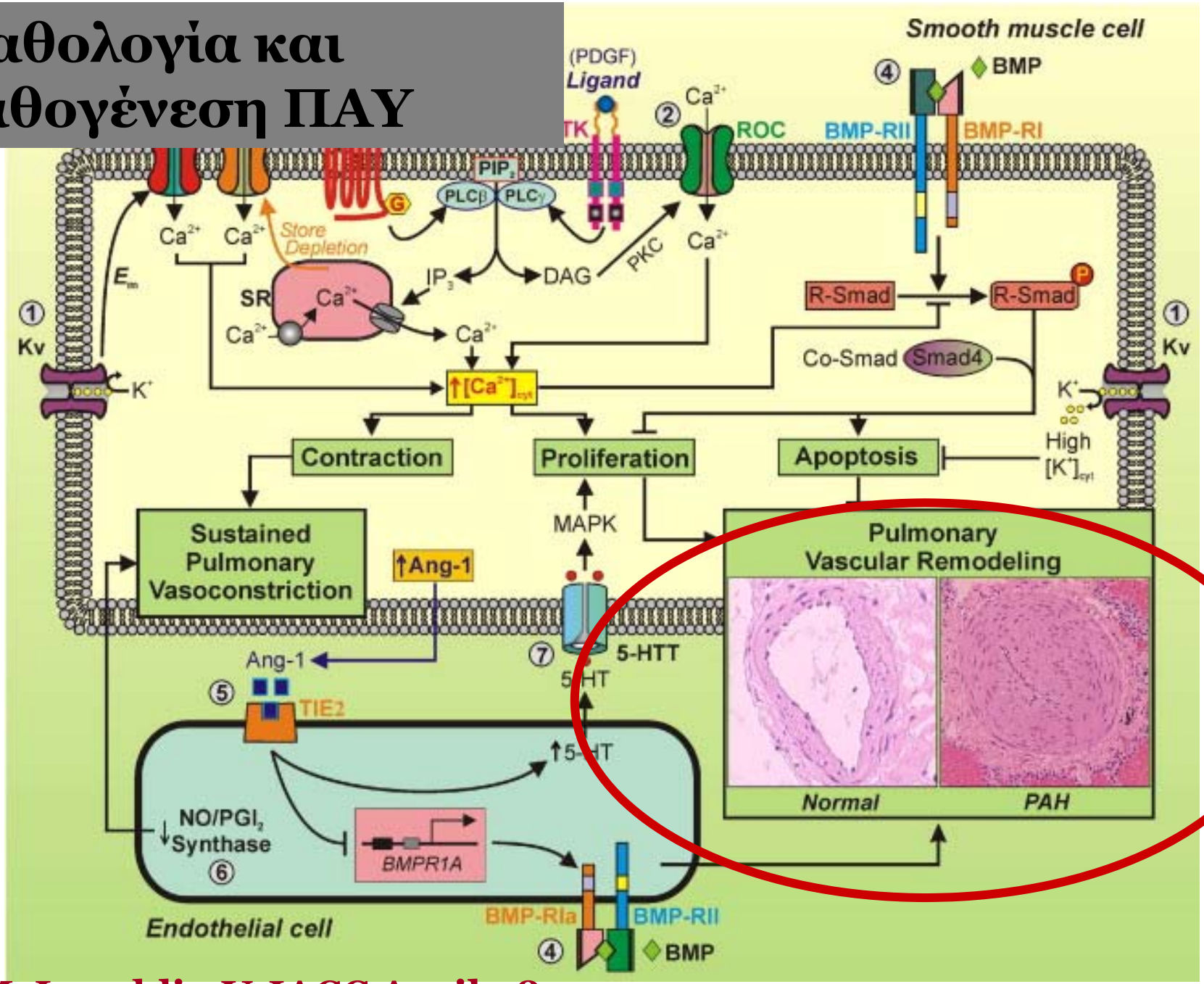
CPS: carbamyl- phosphate synthetase gene

Pathophysiology of PAH 2013



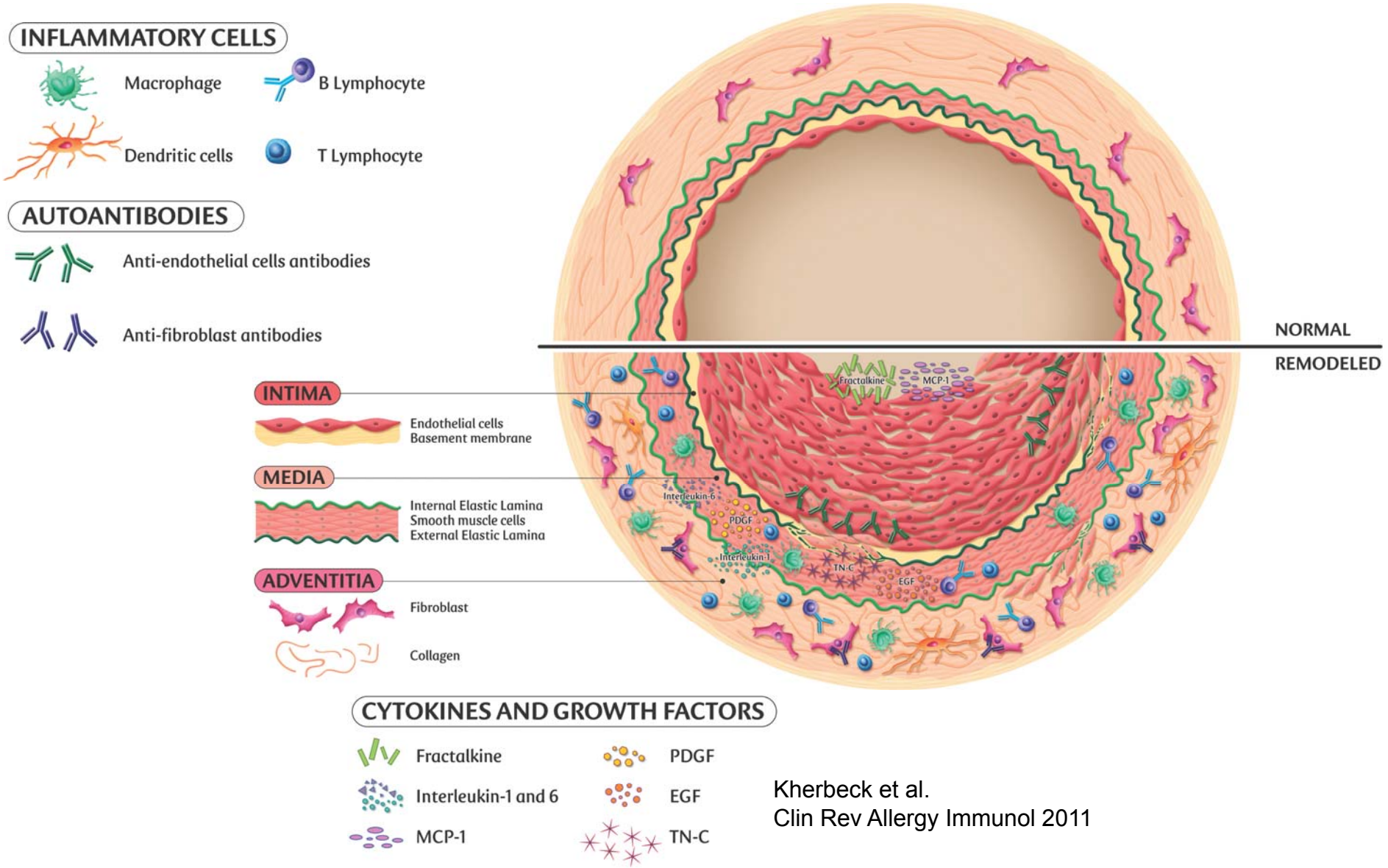
Montani D JOD 2013

Παθολογία και παθογένεση ΠΑΥ



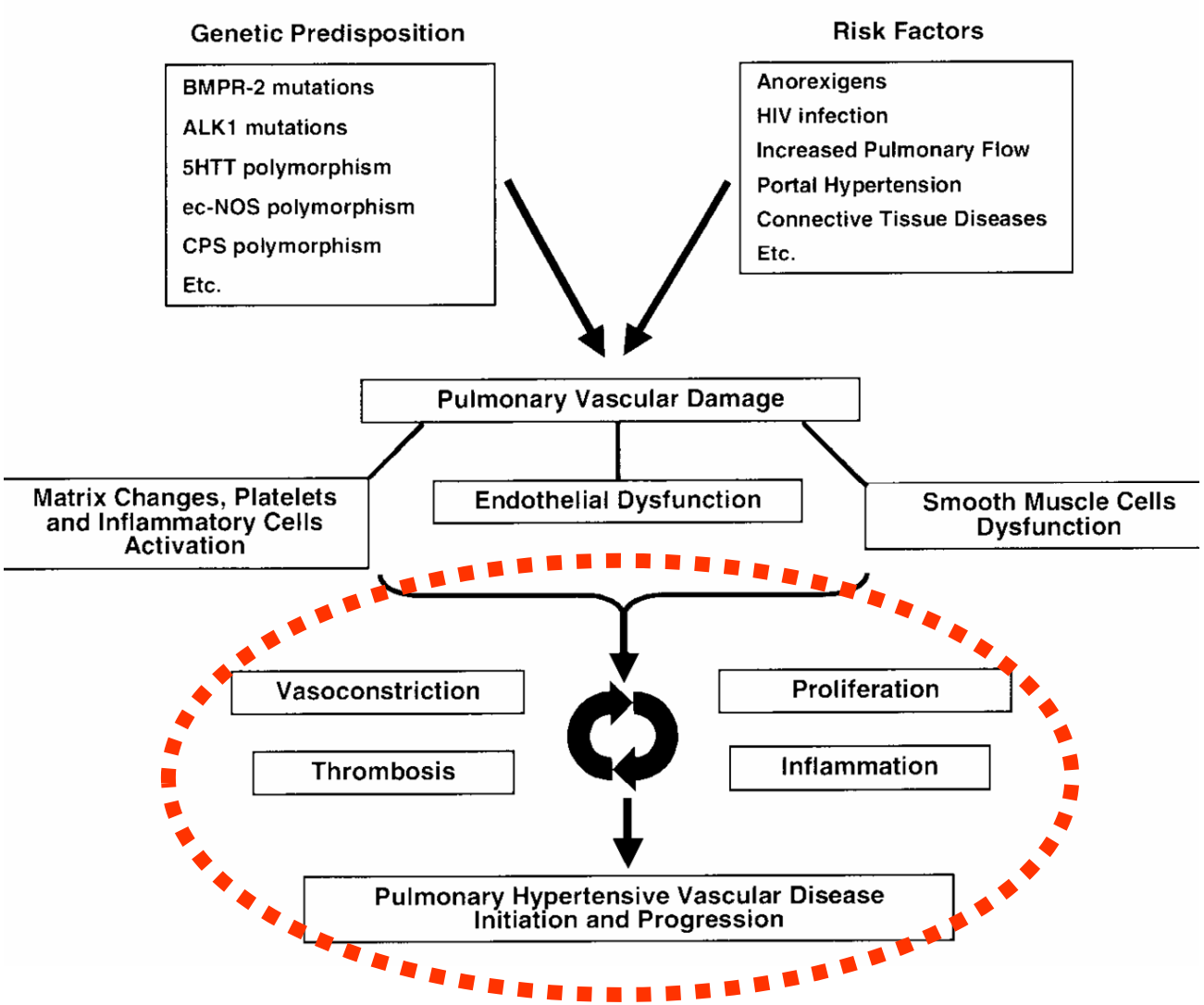
McLaughlin V JACC April 28 2009

FOCUS ON PAH PATHOPHYSIOLOGY



Kherbeck et al.
Clin Rev Allergy Immunol 2011

Αγγειακή Αναδιαμόρφωση



BMPR-2: bone morphogenetic receptor protein2 gene,

ALK 1: activin- receptor- like kinase 1 gene,

5-HTT: serotonin transporter gene,

ec-NOS: nitric oxide synthase gene;

CPS: carbamyl- phosphate synthetase gene

PAH-CTD in Registries

Ref	Methodology	Diagnosis	PAH prevalence
Mukerjee 2003 UK	n=722, monocenter Prospective 1998-2002	RHC	12%
Hachulla 2005 France	599, multicenter Prospective, transsectionnal	RHC	8%
Phung 2009 Australia	184, monocenter Prospective, transsectionnal	RHC	13%

PAH-CTD in Registries

French Registry (674 PAH patients)[1]

Idiopathic	39.2%
CTD	15.3%
CHD	11.3%
Portal hypertension	10.4%
Anorexigens	9.5%
HIV	6.2%
2 co-existing risk factors	4.3%
Familial PAH	3.9%

SSc	76%
SLE	15%
MCTD, Sjögren's, RA, polymyositis	9%

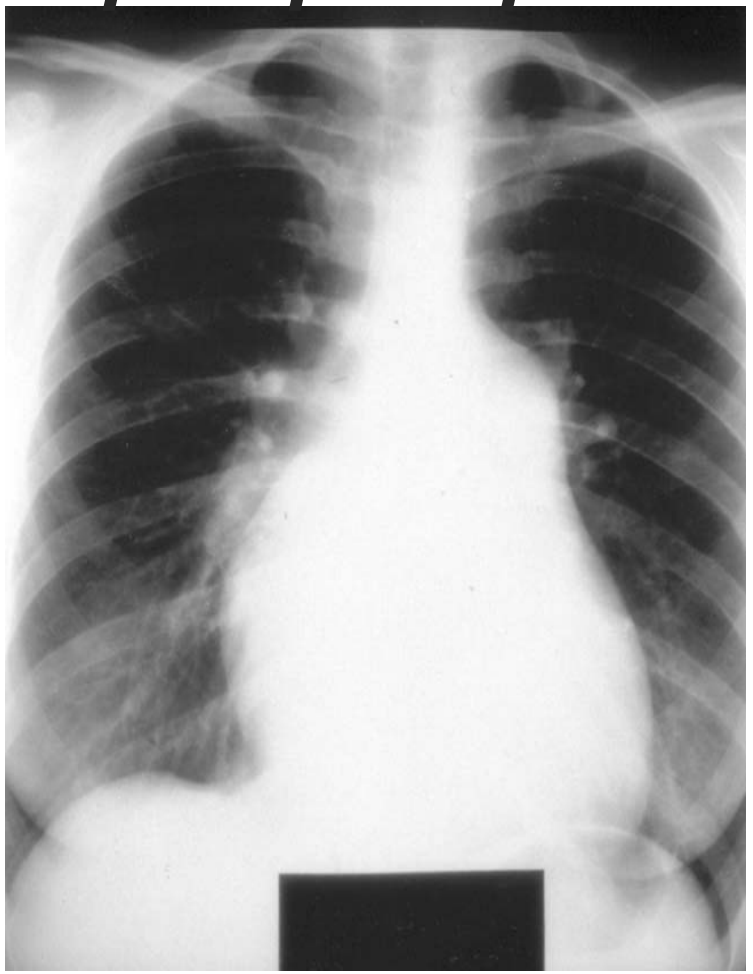
UK Registry (429 PAH-CTD patients)[2]

SSc	74%
MCTD	8%
SLE	8%
DM/PM	4%
RA	3%
UCTD	2%
Sjogren's	1%

1. Humbert M, et al. Am J Respir Crit Care Med 2006; 173:1023
2. Condliffe R, et al. Am J Respir Crit Care Med 2009; 179:151

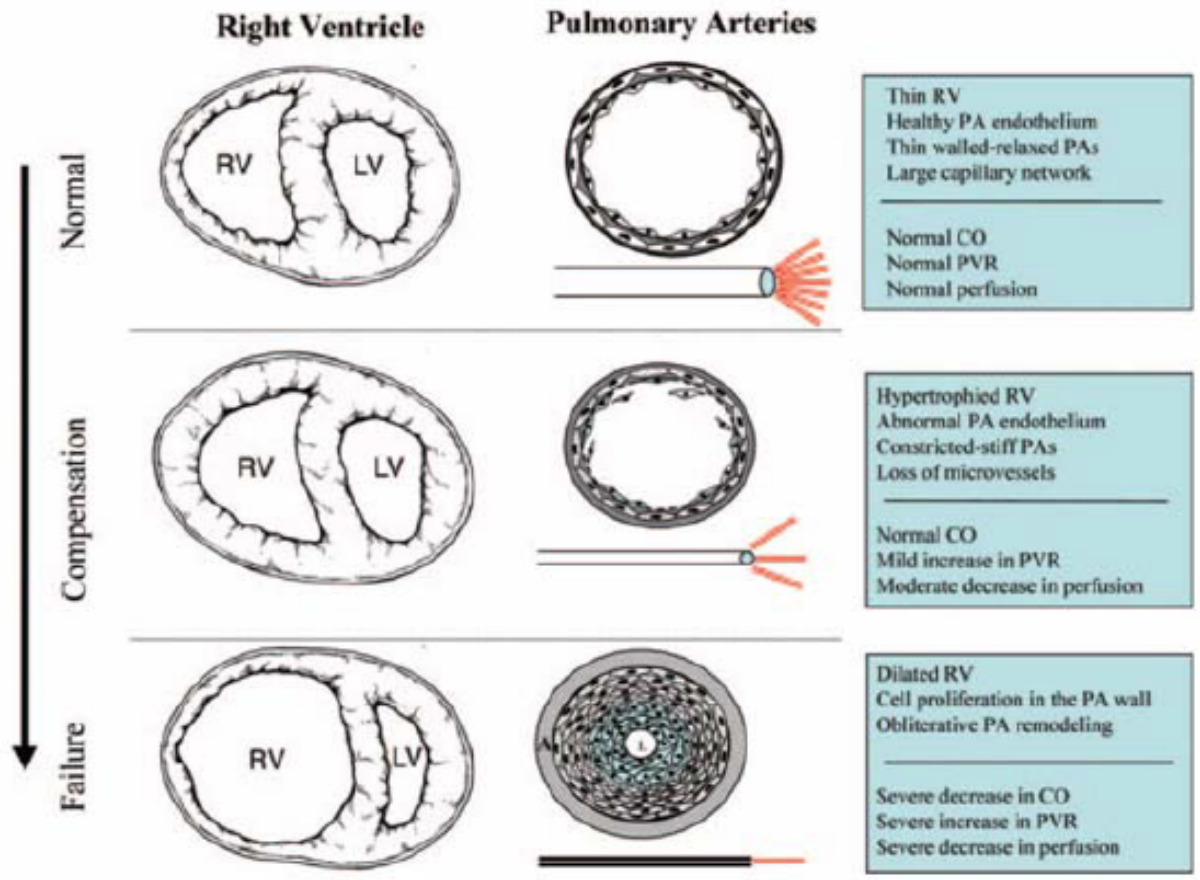
Εστιάζουμε στην Πνευμονική Αρτηριακή Υπέρταση
Προτριχοειδική Πνευμονική Υπέρταση

- ***Hemodynamic definition (right-heart cath)***
  - **Mean PAP** **≥ 25 mmHg at rest**
  - **Wedge PAP** **≤ 15 mmHg**
- ***Consequences***
  - **Right ventricular hypertrophy**
  - **Right heart failure**
  - **Dyspnea, chest pain, (pre-)syncope**
  - **(Sudden) death**



Galiè N et coll. Eur Heart J. 2009 & Eur Resp J 2009

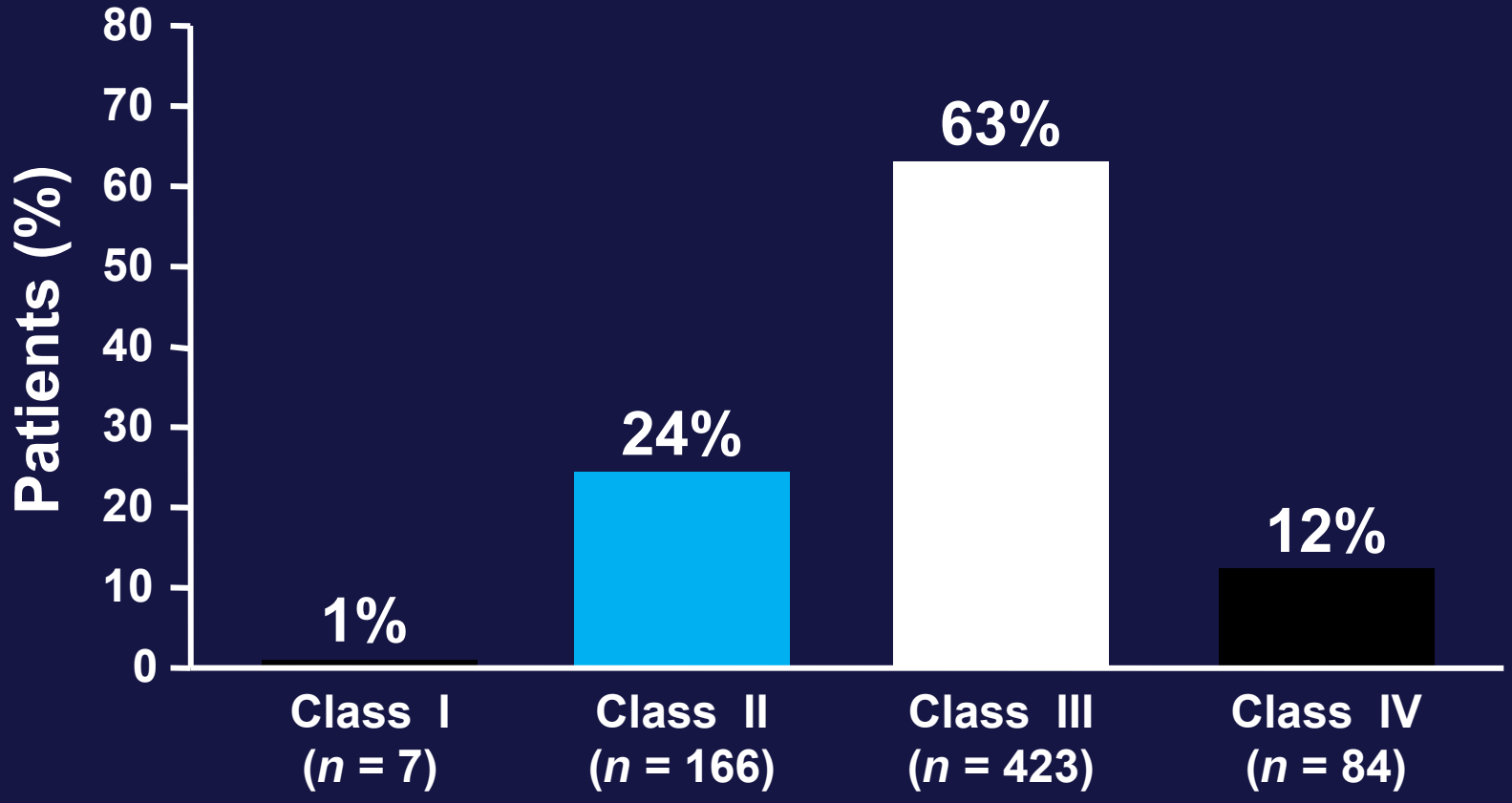
Progression of vascular disease



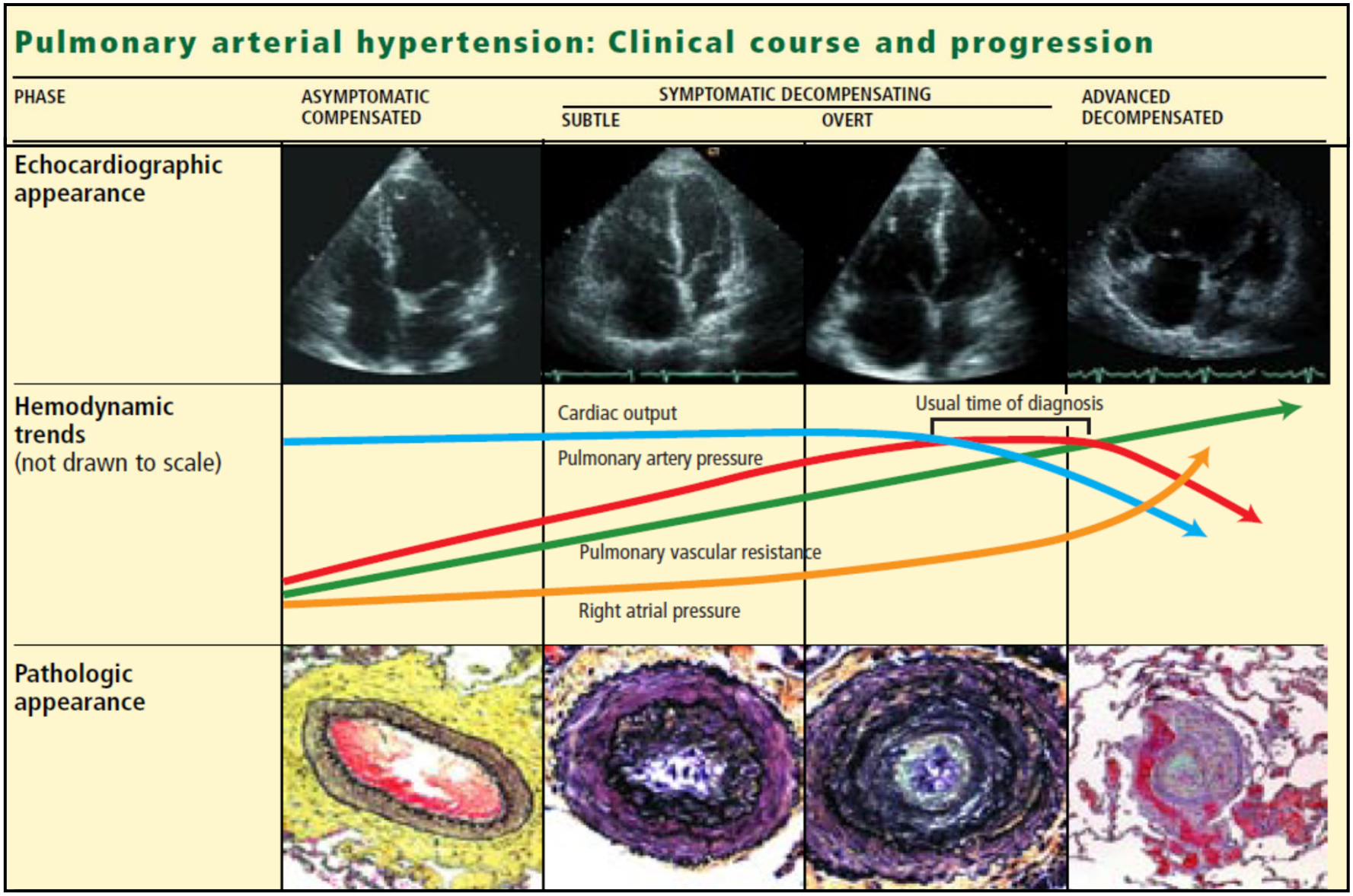
Hunter C. Champion, Evangelos D. Michelakis and Paul M. Hassoun

Circulation 2009;120;992-1007

Λειτουργική κατάσταση NYHA/WHO στην διάγνωση



Humbert M, *et al. Am J Respir Crit Care Med* 2006; 173:1023-30.



Cl C J M 2007;74:737-47

Επιδημιολογία

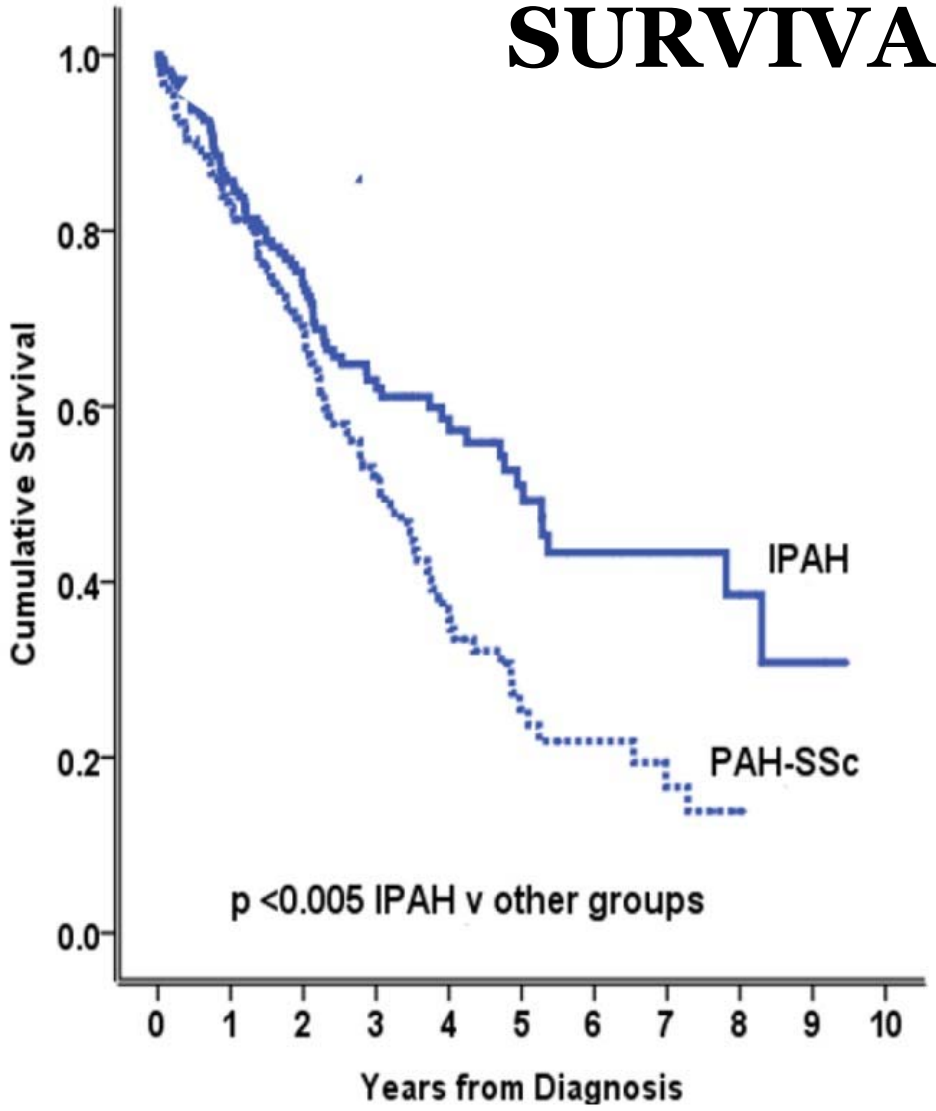
- Η Πνευμονική Αρτηριακή Υπέρταση (ΠΑΥ) προσβάλει 0.5-15% των ασθενών με νοσήματα του συνδετικού ιστού [connective tissue diseases (CTD) and mixed connective tissue diseases (MCTD)]

- Οι περισσότεροι ασθενείς με ΠΑΥ που σχετίζεται με νόσημα του συνδετικού ιστού πάσχουν από σκληρόδερμα (Systemic sclerosis-SS

7-12% of pts with SSc have PAH

- Η καρδιοπνευμονική συμμετοχή ευθύνεται για > 50% των θανάτων σε ασθενείς με SSC

SURVIVAL in PAH



108	98	85	67	46	32	PAH-Eisenmenger's
175	143	102	67	44	30	IPAH
156	123	83	51	31	14	PAH-SSc
439	364	270	185	121	76	Total

Condliffe R 2015

Potential reasons

Age

Pulmonary vasculopathy

(arter, venules)

Right ventricle

(reduced contractility)

Left ventricle

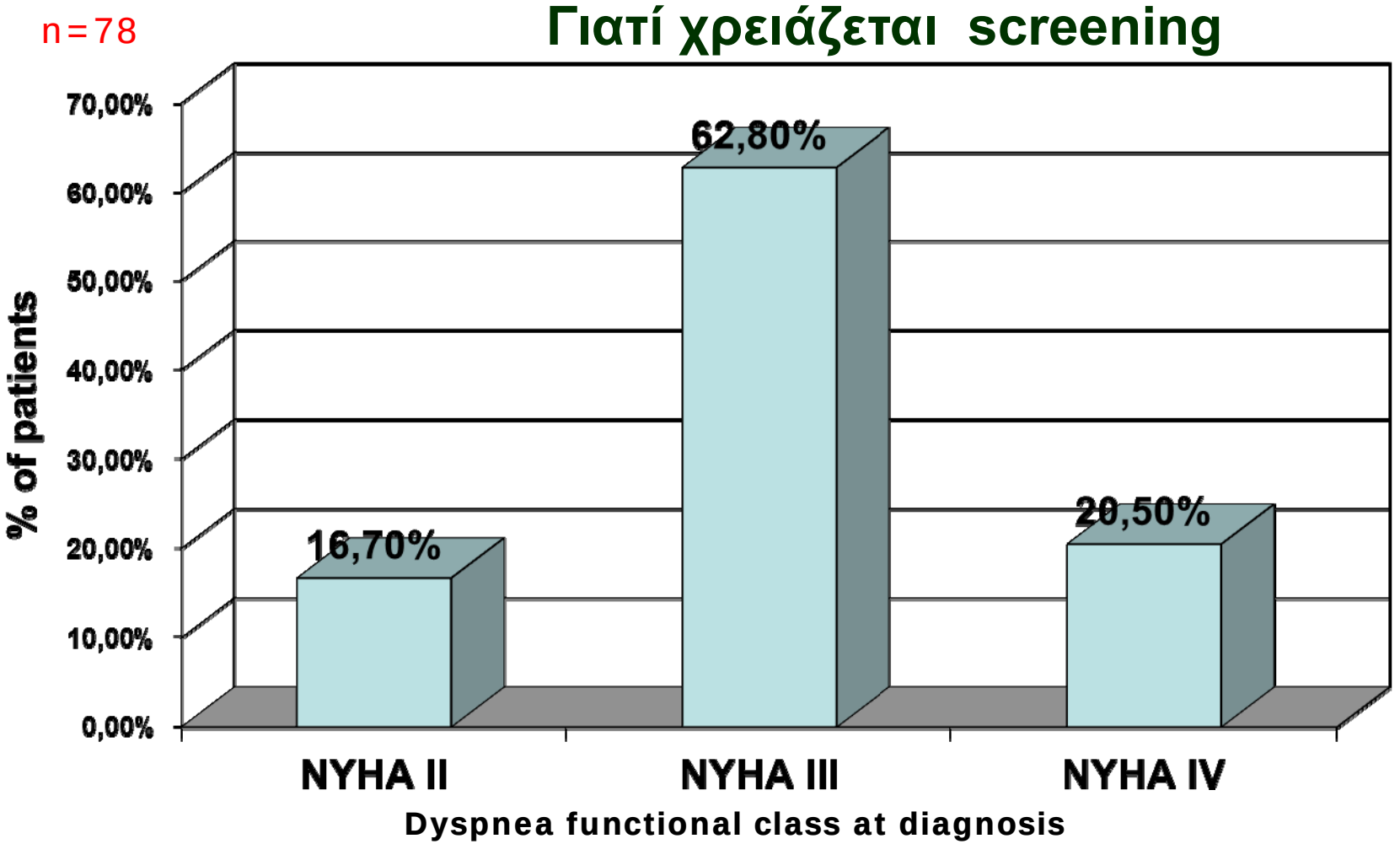
(S/D dysfunction)

ILD

Multisystem disease

Antibodies

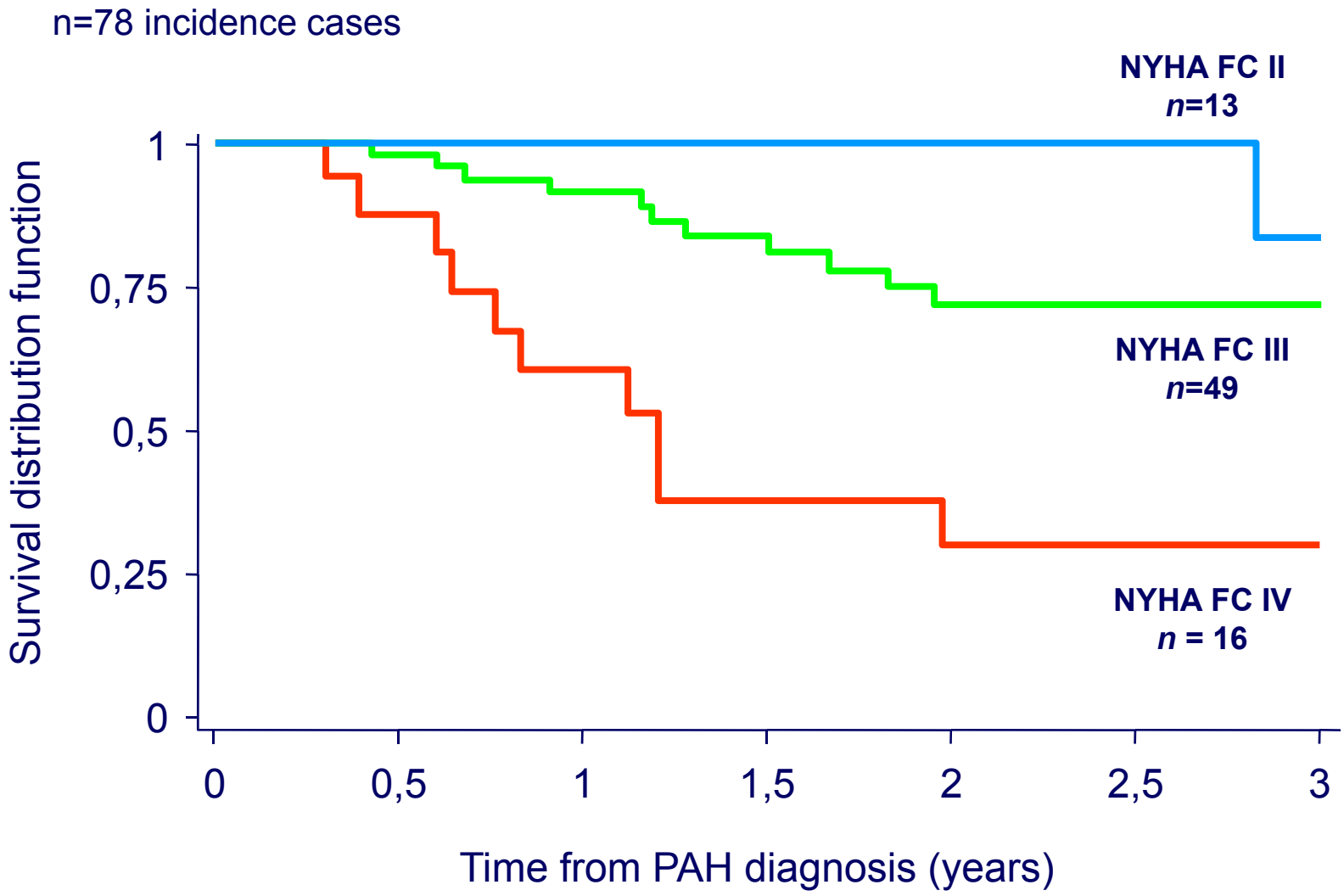
Most of SSc-PAH patients have severe symptoms



Hachulla E. Chest 2009; 136: 1211

Γιατί χρειάζεται screening

Survival depends on NYHA FC at diagnosis

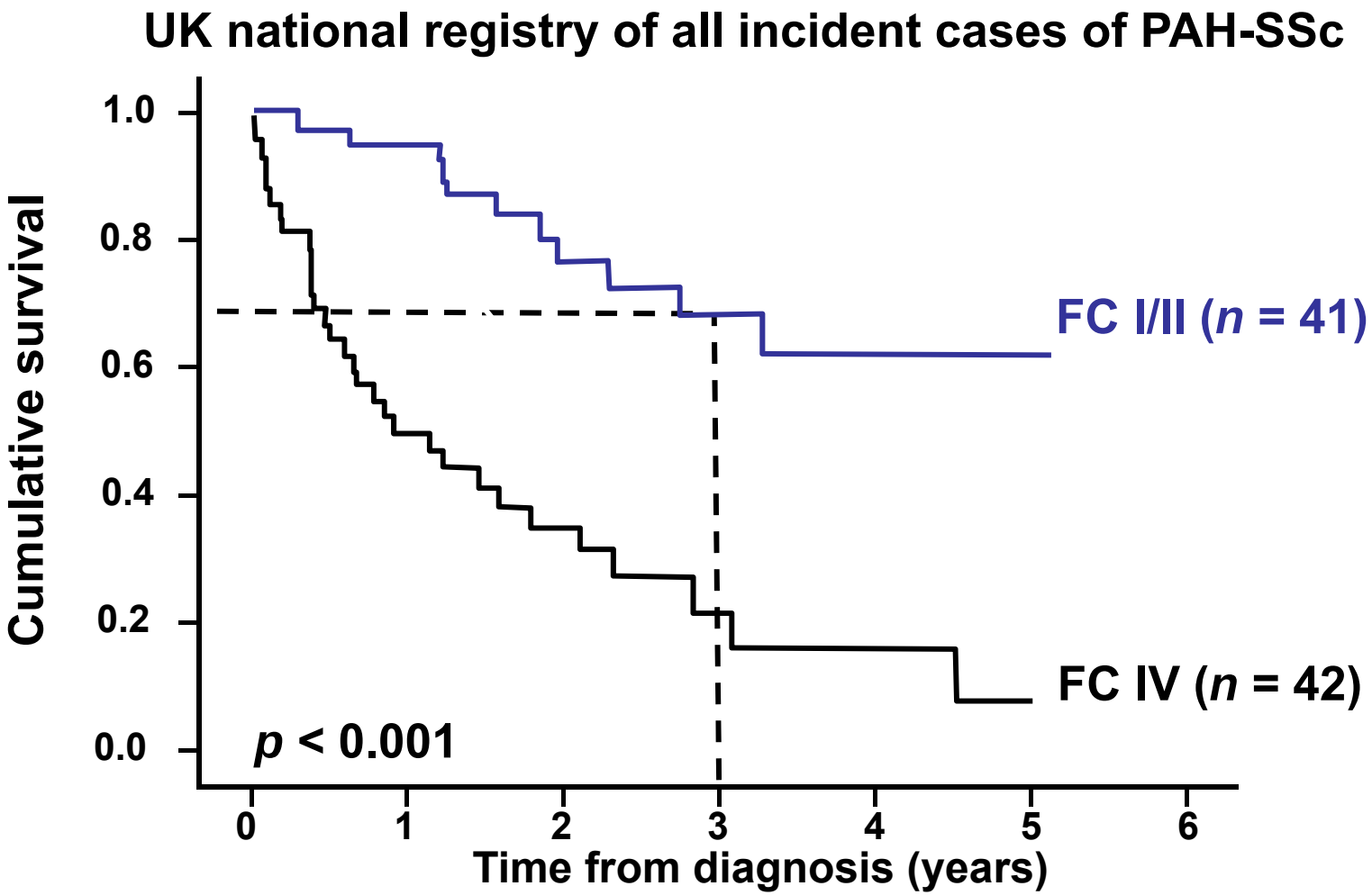


Log rank test p=0.002

Hachulla E. Chest 2009; 136: 1211

Γιατί χρειάζεται screening

Survival depends on NYHA FC at diagnosis



Condliffe R. Am J Respir Crit Care Med 2009; 179:151

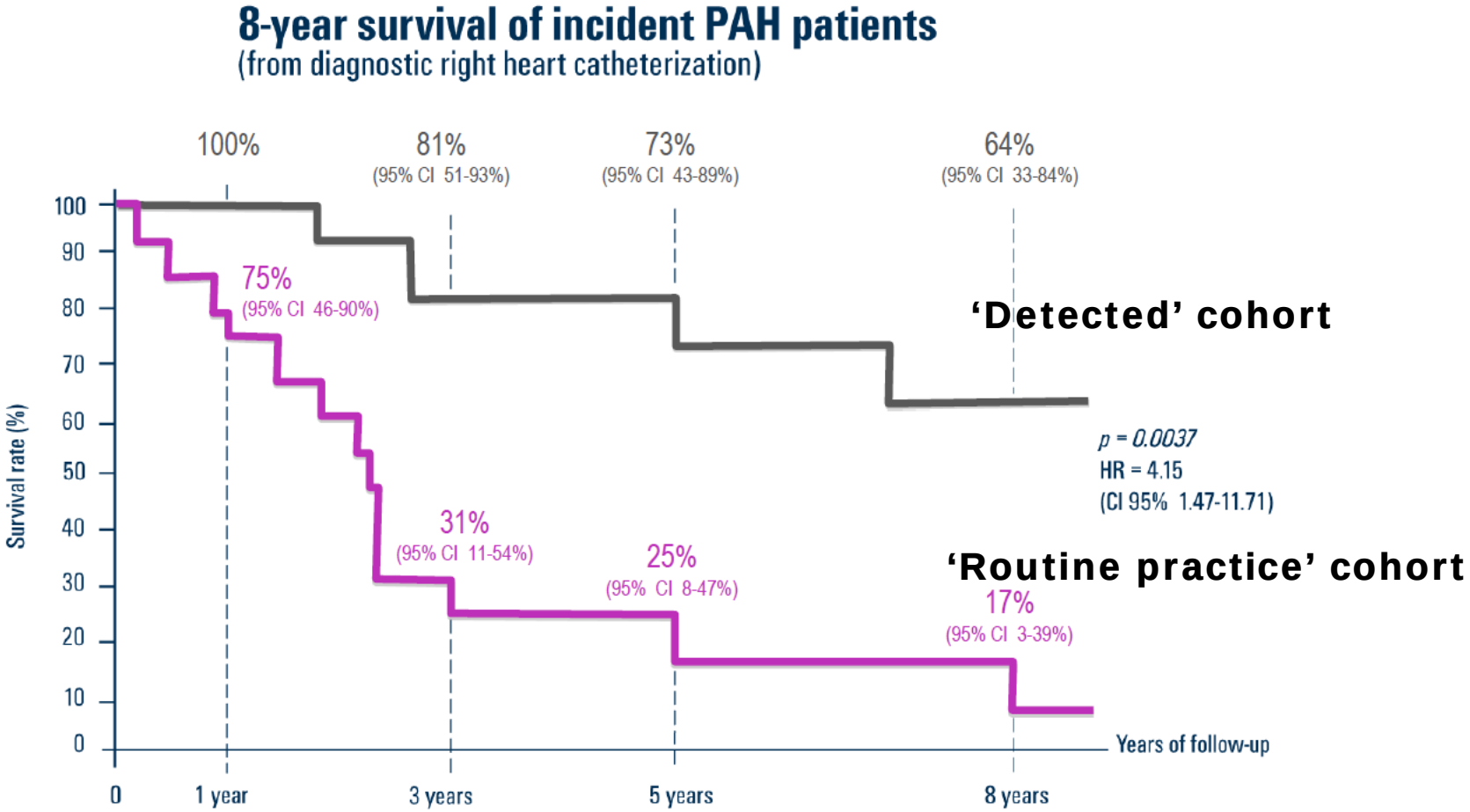
Condliffe R ERJ 2012

Clinical Features
screening

- ***Risk factors for the development of PAH in SSc patients:***
  - Late-onset disease
  - An isolated reduction in DLCo
  - An FVC%/DLCO% ratio greater than 1.6 or
  - A combined decreased DLCO/alveolar volume with elevation of serum N-terminal pro-natriuretic peptide levels

Boueiz A Ann Thor MED 2014 Sep

A screening programme may improve the prognosis



Humbert M. Arthritis Rheum 2011: 63: 3522

Αρχική εκτίμηση ασθενών με SSc και νοσήματα στο φάσμα του σκληροδέρματος

- Πνευμονικές δοκιμασίες (PFTs) High QE
  - Σπιρομέτρηση με στατικούς όγκους
  - Μέτρηση διαχυτικής ικανότητας (DLCO)
- Διαθωρακικός υπέρηχος καρδιάς High QE
- N-terminal pro-B-type natriuretic peptide (NT-ProBNP) Moderate QE
- DETECT αλγόριθμος αν DLCO <60% pred και διάρκεια νόσου > 3 έτη Moderate QE

Dinesh Khanna Dec 2013 Arthritis & Rheumatism

Πώς πρέπει να ελέγχονται οι ασθενείς;;

Echocardiography: the best screening tool

Table 9 **Arbitrary criteria for estimating the presence of PH based on tricuspid regurgitation peak velocity and Doppler-calculated PA systolic pressure at rest (assuming a normal right atrial pressure of 5 mmHg) and on additional echocardiographic variables suggestive of PH**

	Class[a]	Level[b]
Echocardiographic diagnosis: PH unlikely		
Tricuspid regurgitation velocity ≤2.8 m/s, PA systolic pressure ≤36 mmHg, and no additional echocardiographic variables suggestive of PH	I	B

ESC-ERS guidelines ERJ 2009

PAH-CTD screening: ESC/ERS recommendations

Table 26 Recommendations for PAH associated with connective tissue disease

Statement	Class[a]	Level[b]
Echocardiographic screening for the detection of PH is recommended in symptomatic patients with scleroderma spectrum of diseases	I	B
Echocardiographic screening for the detection of PH is recommended in symptomatic patients with all other CTDs	I	C
Echocardiographic screening for the detection of PH may be considered in asymptomatic patients with the scleroderma spectrum of disease	IIb	C

[a]Class of recommendation.
[b]Level of evidence.

Galiè N. Eur Heart J 2009; 30:2493

Πώς πρέπει να ελέγχονται οι ασθενείς;;

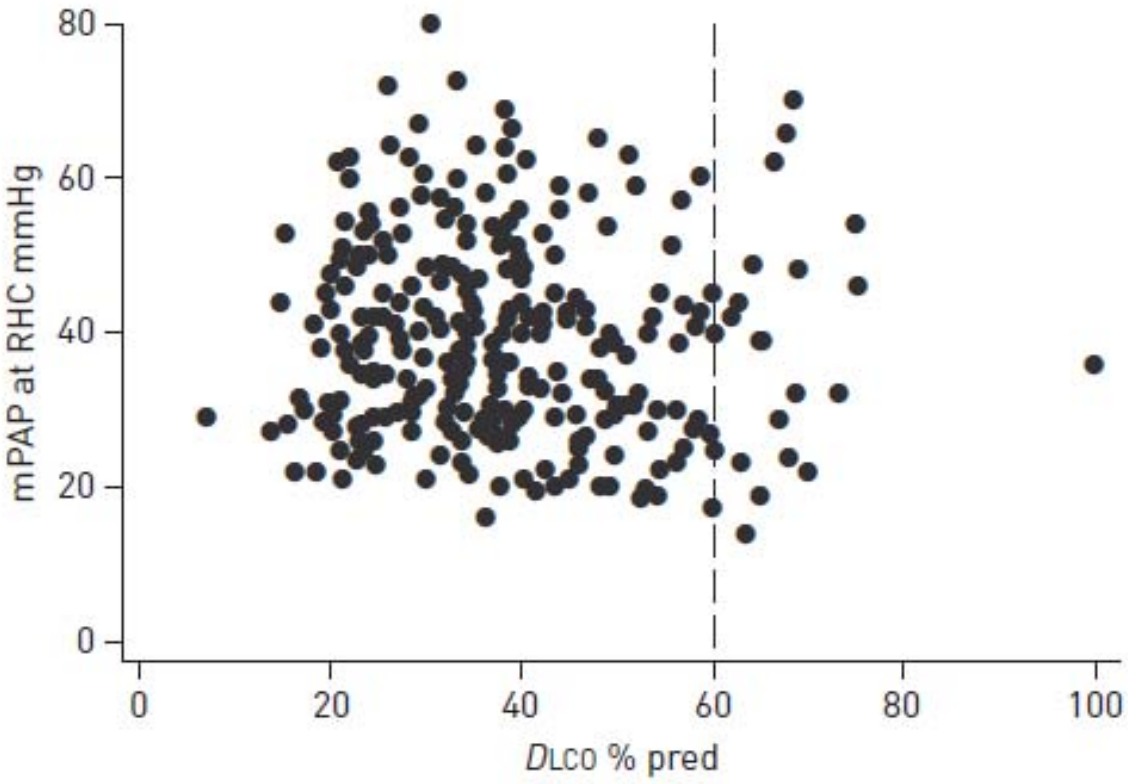
DLCO/VA; NT-proBNP: a PAH predictive factor ?

Table 2. Results of univariate and multivariate analyses of candidate predictors of PAH, by model analyzed*

Model, variable	Univariate analysis		Multivariate analysis	
	P	HR (95% CI)	*P*	HR (95% CI)
Main model				
DLco/Va <70%	0.0043	21.3 (2.5–181.3)	0.014	18.81 (1.7–206.8)
High NT-proBNP	0.0048	10.1 (1.96–51.72)	0.053	6.35 (0.94–82.8)
Systolic PAP >40 mm Hg	0.0078	1.08 (1.63–30.87)	0.54	0.40 (0.02–7.79)
ESR >28 mm/hour	0.015	5.6 (1.35–23.01)	0.15	6.19 (0.49–76.9)

Allanore Y. Arthritis Rheum 2010;58 : 284

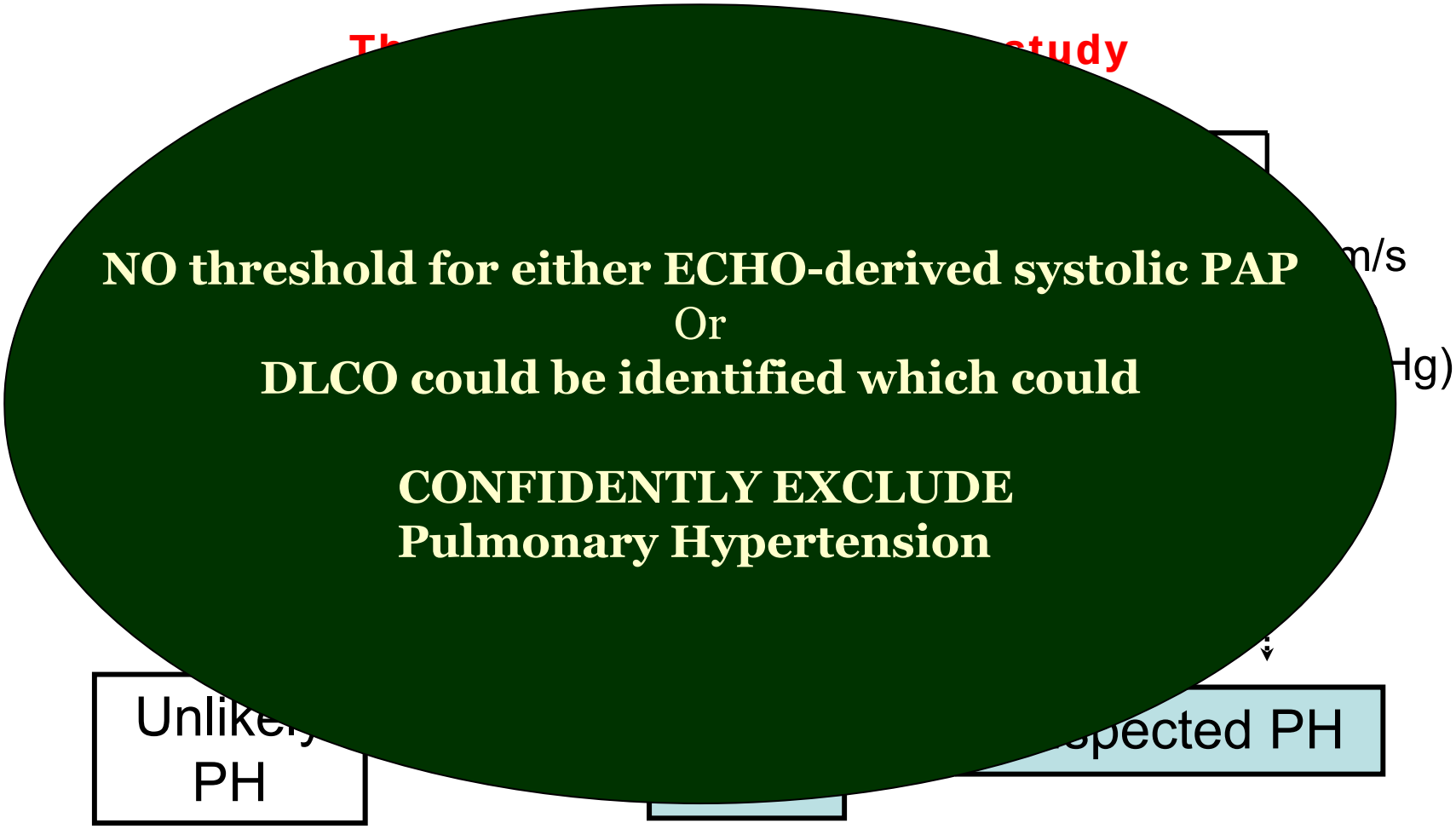
DLCO IN CTD-PAH



Schwaiger J ERV 2013;22:515

Πώς πρέπει να ελέγχονται οι ασθενείς;;

Doppler echocardiography



TJV= tricuspid jet velocity

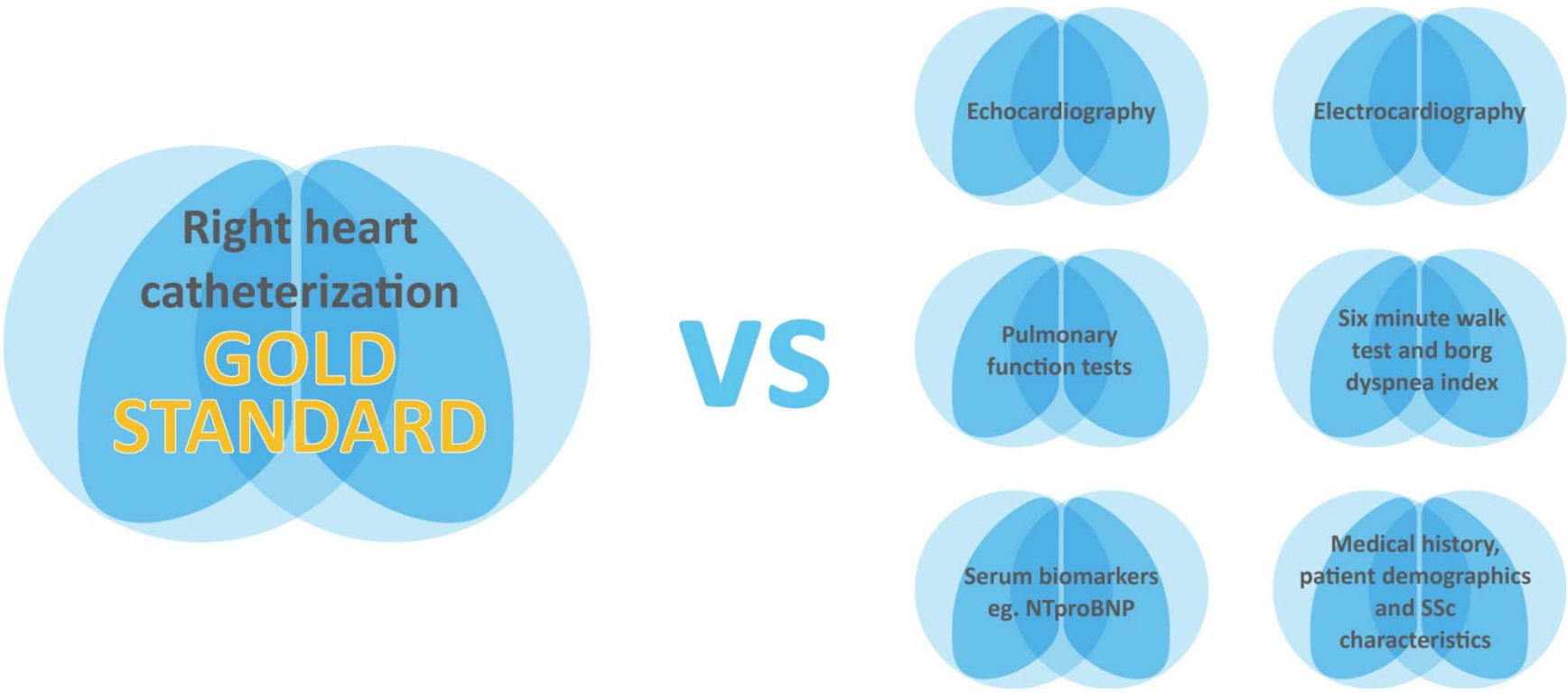
Hachulla E. Arthritis Rheum 2009; 60: 1831

Πώς πρέπει να ελέγχονται οι ασθενείς;;

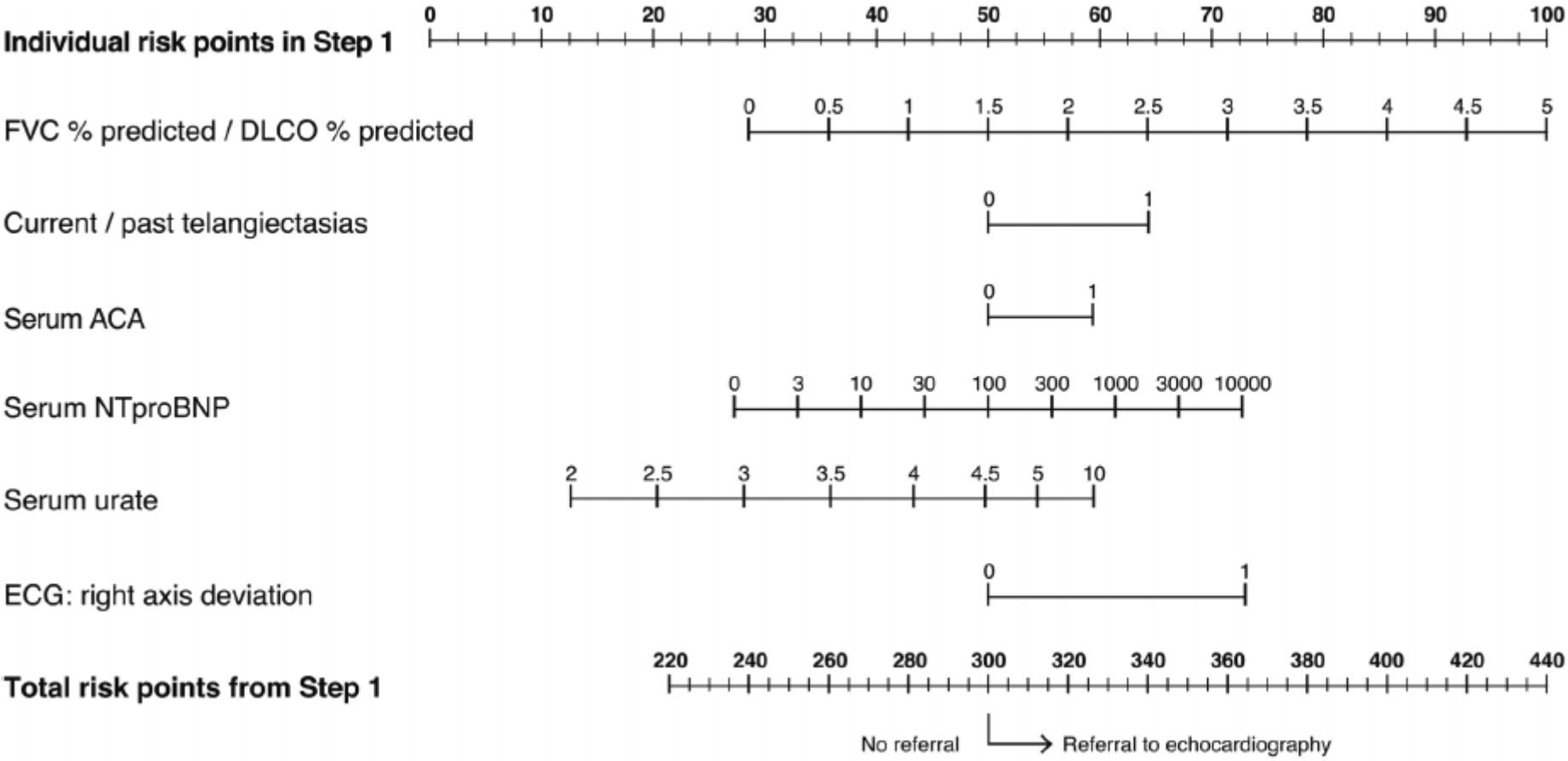
Combination predictive factors ?

The DETECT Study

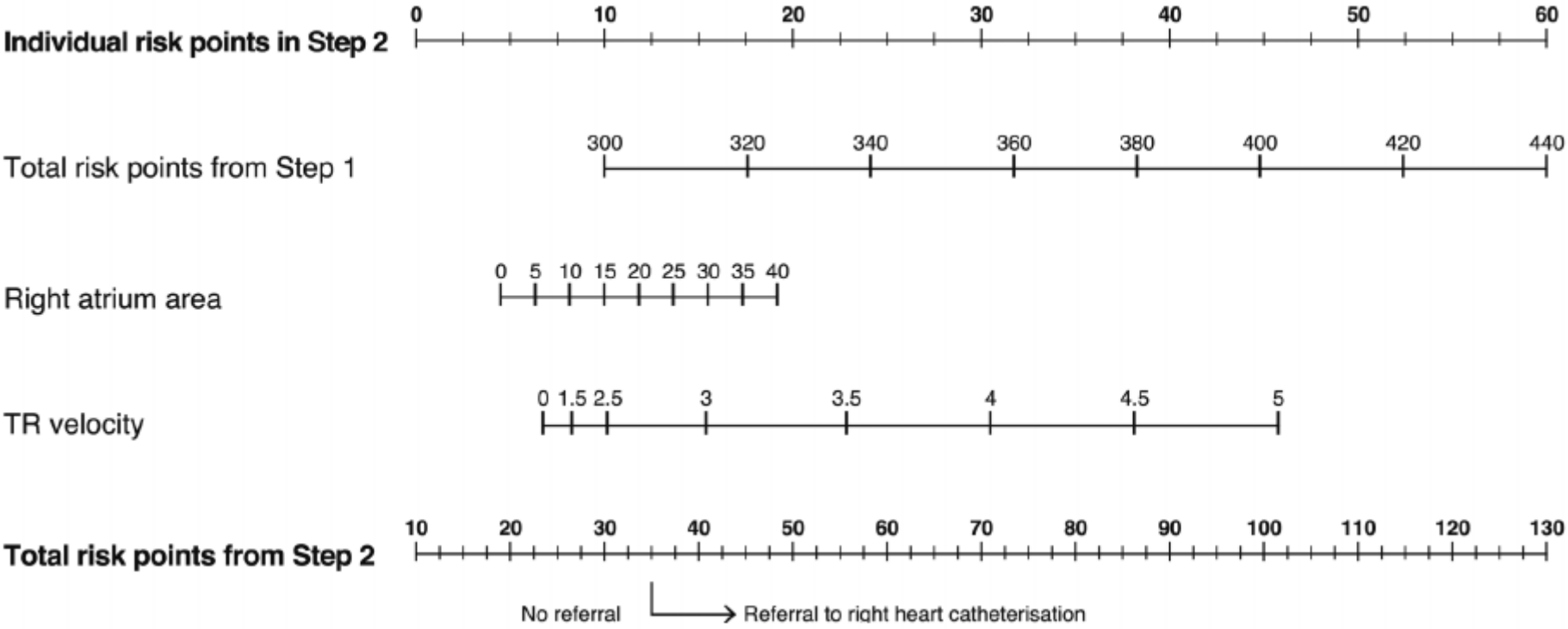
- **DETECT will evaluate more than 10 screening tools and their combinations against the confirmatory gold standard diagnostic test for PAH in SSc patients having a DLCO<60%**



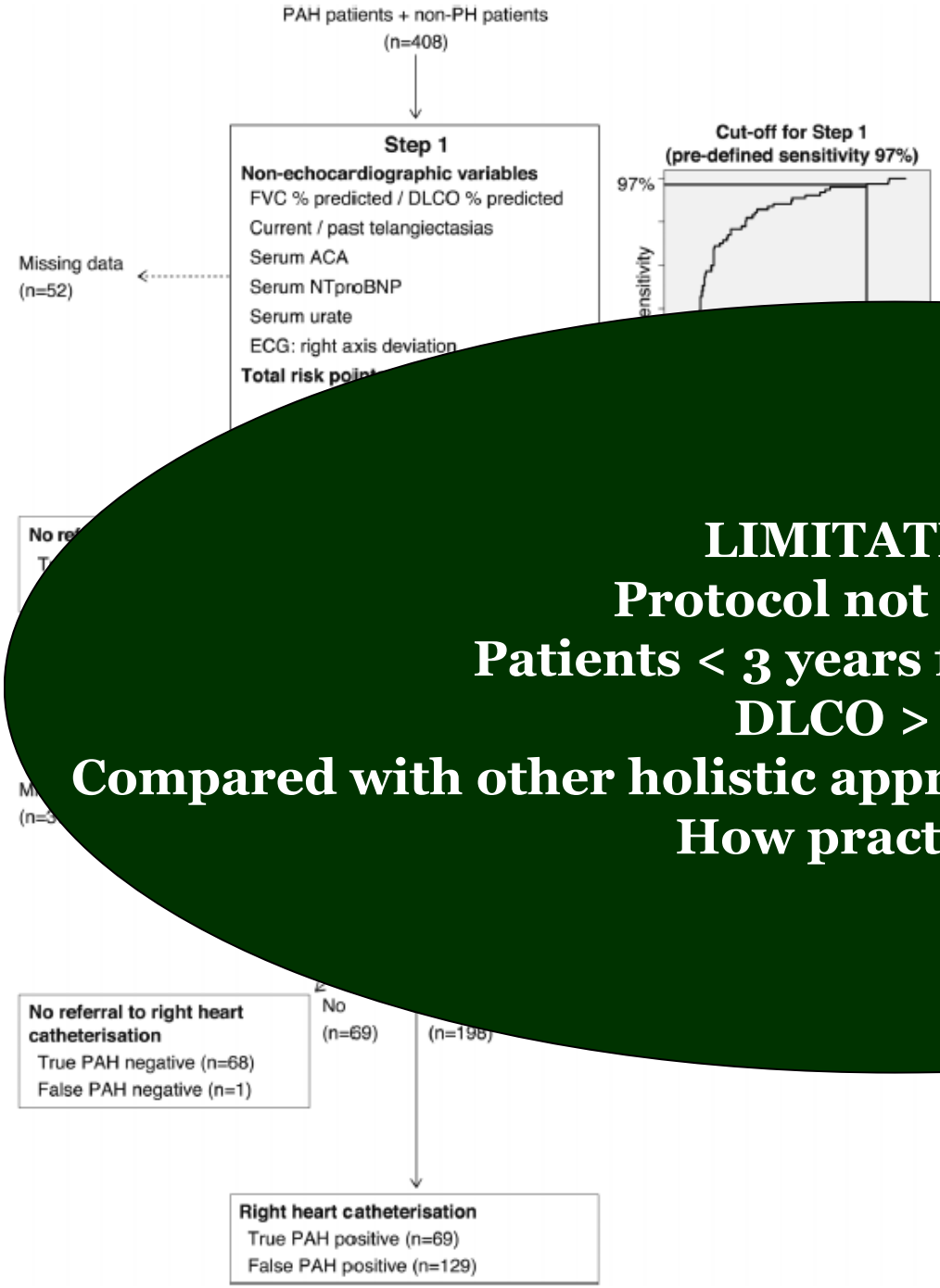
Seibold et al. American College of Rheumatology 2008; Distler et al. Swiss Society for Pulmonary Hypertension 2009; Vonk et al. Systemic Sclerosis World Congress 2010



DETECT study



DETECT study



DETECT study
Coghlan JG Am Rheum Dis 2013;00 1-10

detection of
rtension

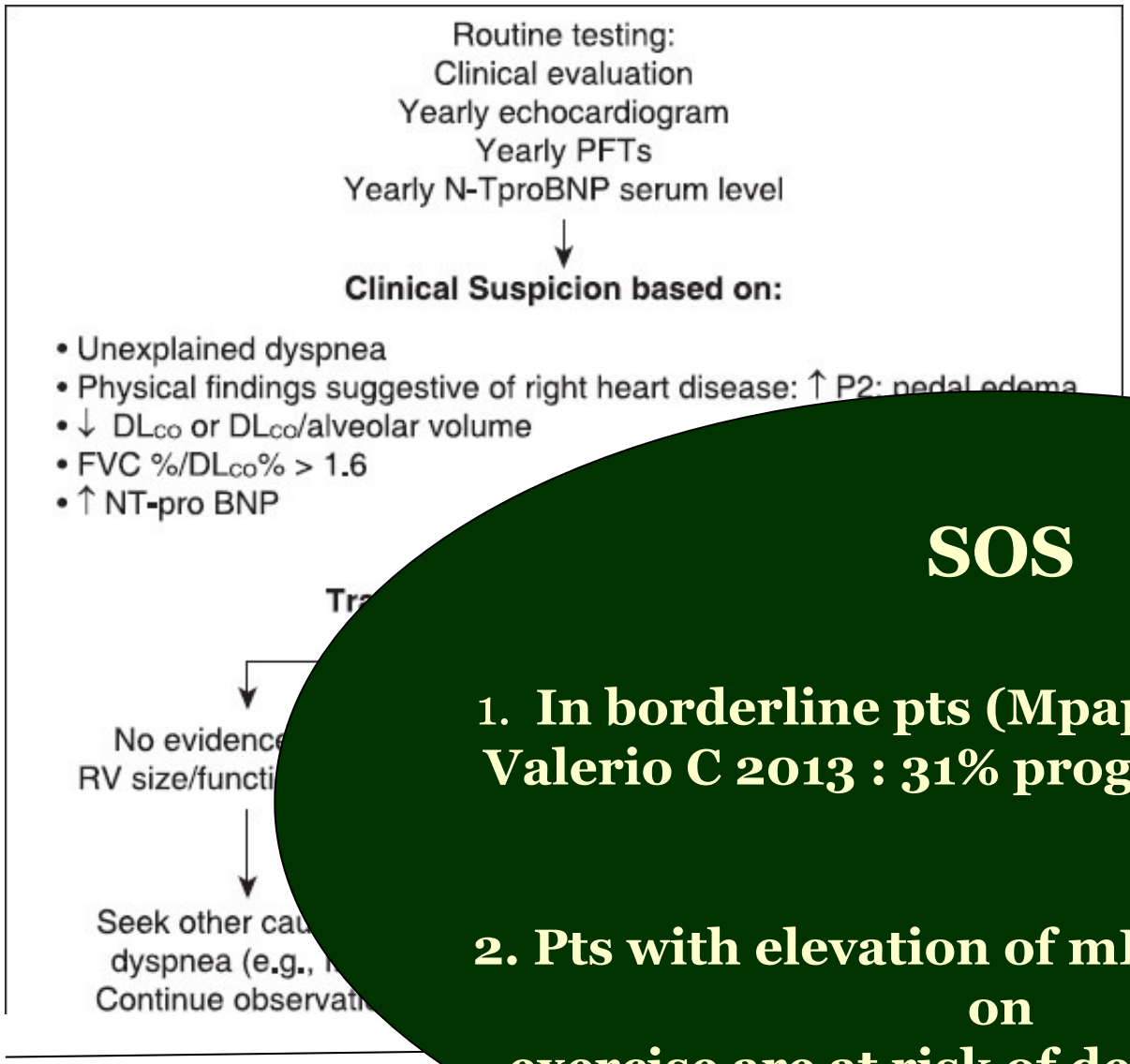
LIMITATIONS
Protocol not validated
Patients < 3 years from diagnosis
DLCO > 60%
Compared with other holistic approach (echo, symptoms, DLCO)
How practical is?

ied

Σύσταση για Καθετηριασμό Δεξιών Κοιλοτήτων

	Signs or symptoms required for RHC†	Quality of evidence
Transthoracic echocardiogram		
TR jet velocity		
2.5–2.8 meters/second	Yes	High
>2.8 meters/second	No	High
Right atrial or right ventricular enlargement (right atrium major dimension >53 mm and right ventricle midcavity dimension >35 mm), irrespective of TR jet velocity	No	High
PFTs		
FVC:DLCO ratio >1.6 and/or DLCO <60% predicted‡	Yes	High
FVC:DLCO ratio >1.6 and/or DLCO <60% predicted and NT-proBNP >2 times upper limit of normal‡	No	High
Composite measure		
Meets DETECT algorithm in patients with DLCO <60% predicted and disease duration >3 years‡	No	Moderate

Dinesh Khanna Dec 2013 Arthritis & Rheumatism



Guidelines

PAH in CTD

2014

SOS

1. **In borderline pts (Mpap= 21-24mmhg)**
Valerio C 2013 : 31% progression to PAH

2. Pts with elevation of mPAP >30 mmHg
on
exercise are at risk of developing PAH

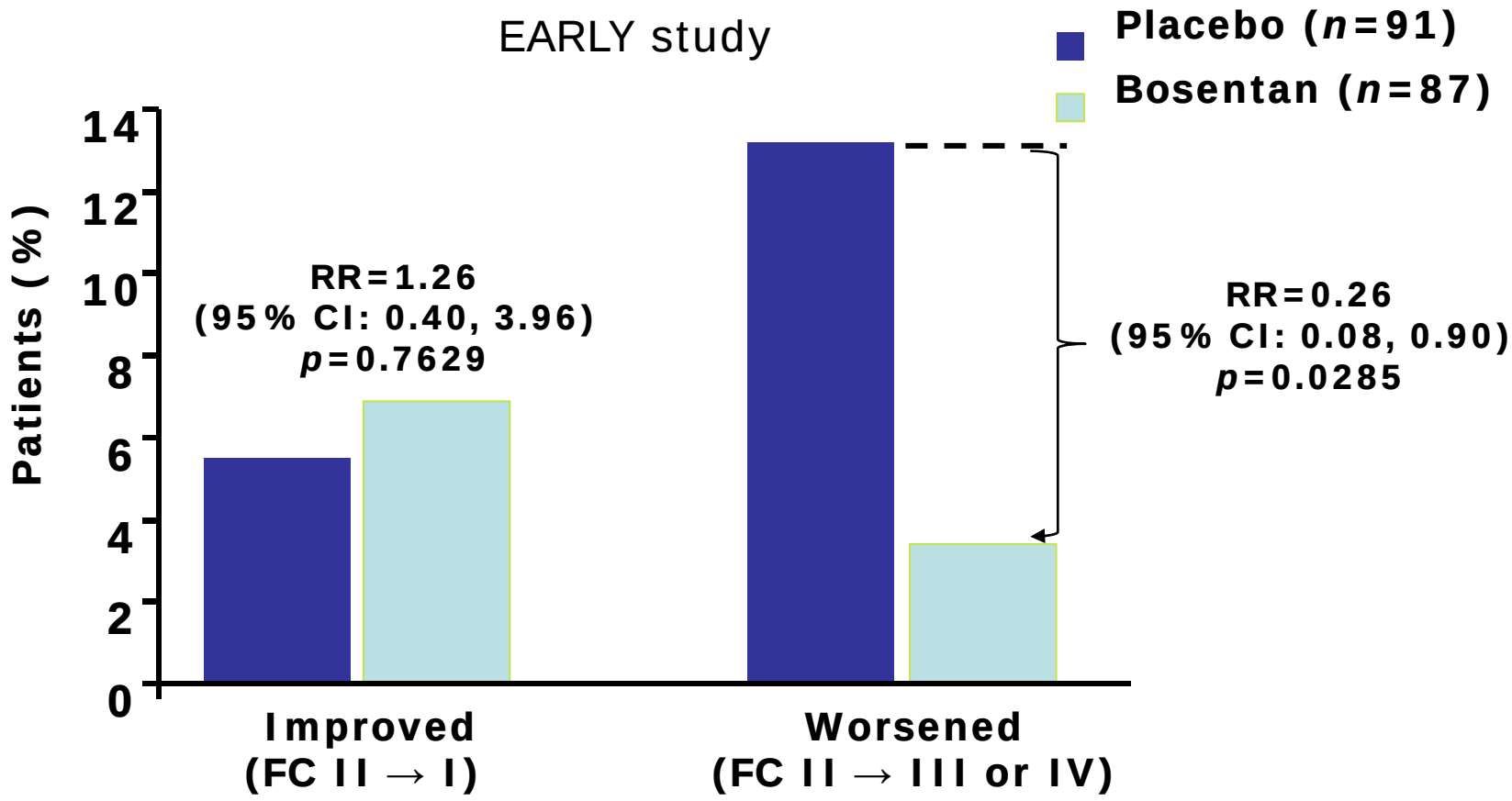
Boueiz A Ann Thor M

Prognosis of SSc-PAH in selected major registries

Registry	Year	n	Age (yrs)	Incident cases (%)	WHO FC I&II/III/IV (%)	mPAP (mmHg)	PVR (dyns.s.cm5)	1 yr survival (%)	3 yr survival(%)
UK [26]	2009	259	64	100	16/68/16	42	715	78	47
REVEAL [20]	2010	399	62	18	25/60/15	45	768	82	n/a
ASPIRE [25]	2012	156	66	100	19/67/14	43	678	82	52
French [27]	2013	85	65	100	21/67/12	41	680	90	56
PHAROS [28]	2014	131	60	100	56/38/6	36	448	93	75

Condliffe R 2015

Early PAH management may improve the prognosis

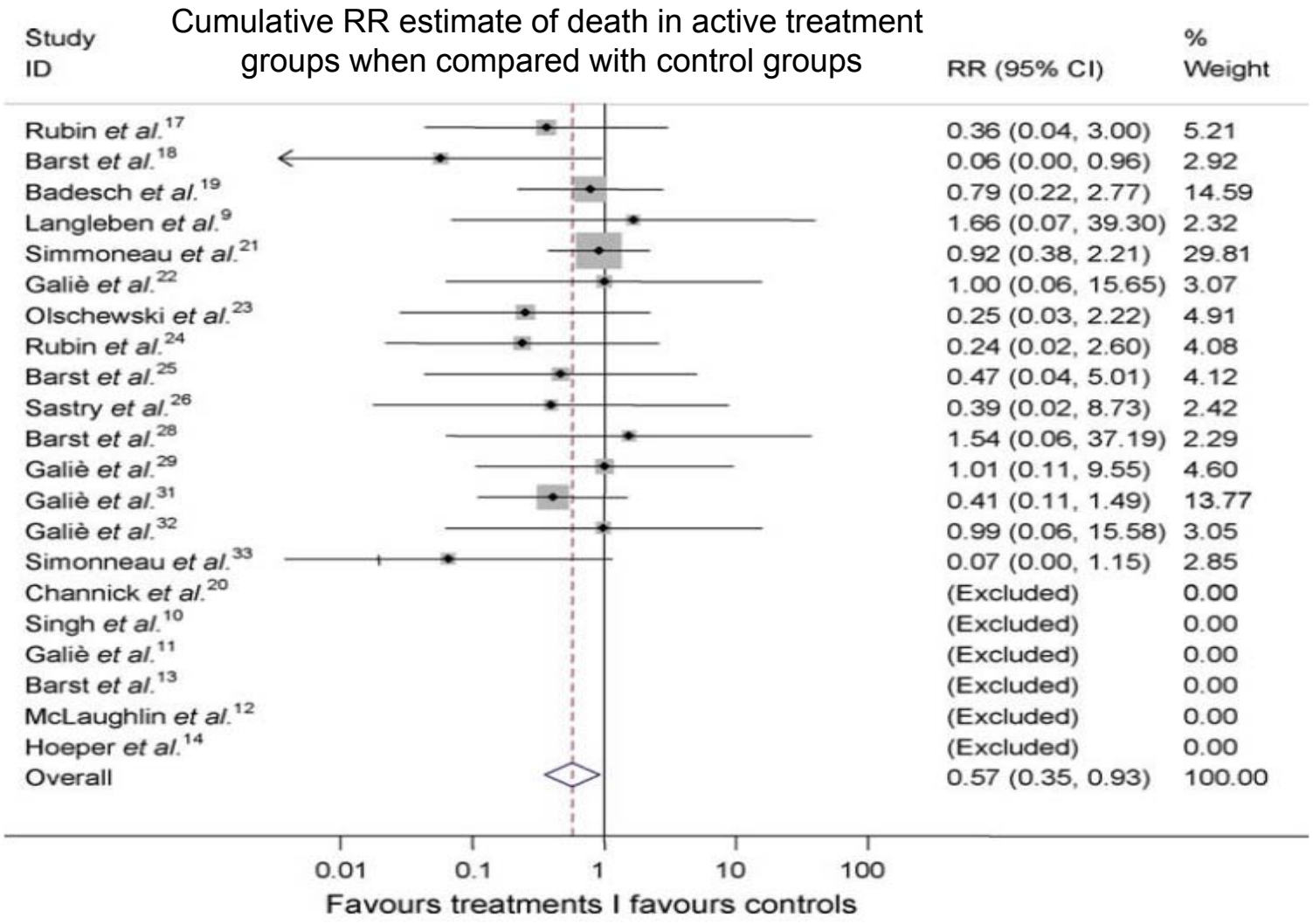


Effect on NYHA/WHO functional class

Galiè N et al. Presented at ESC 2007
Galiè N et al. Lancet 2008; 371:2093

Γιατί χρειάζεται screening

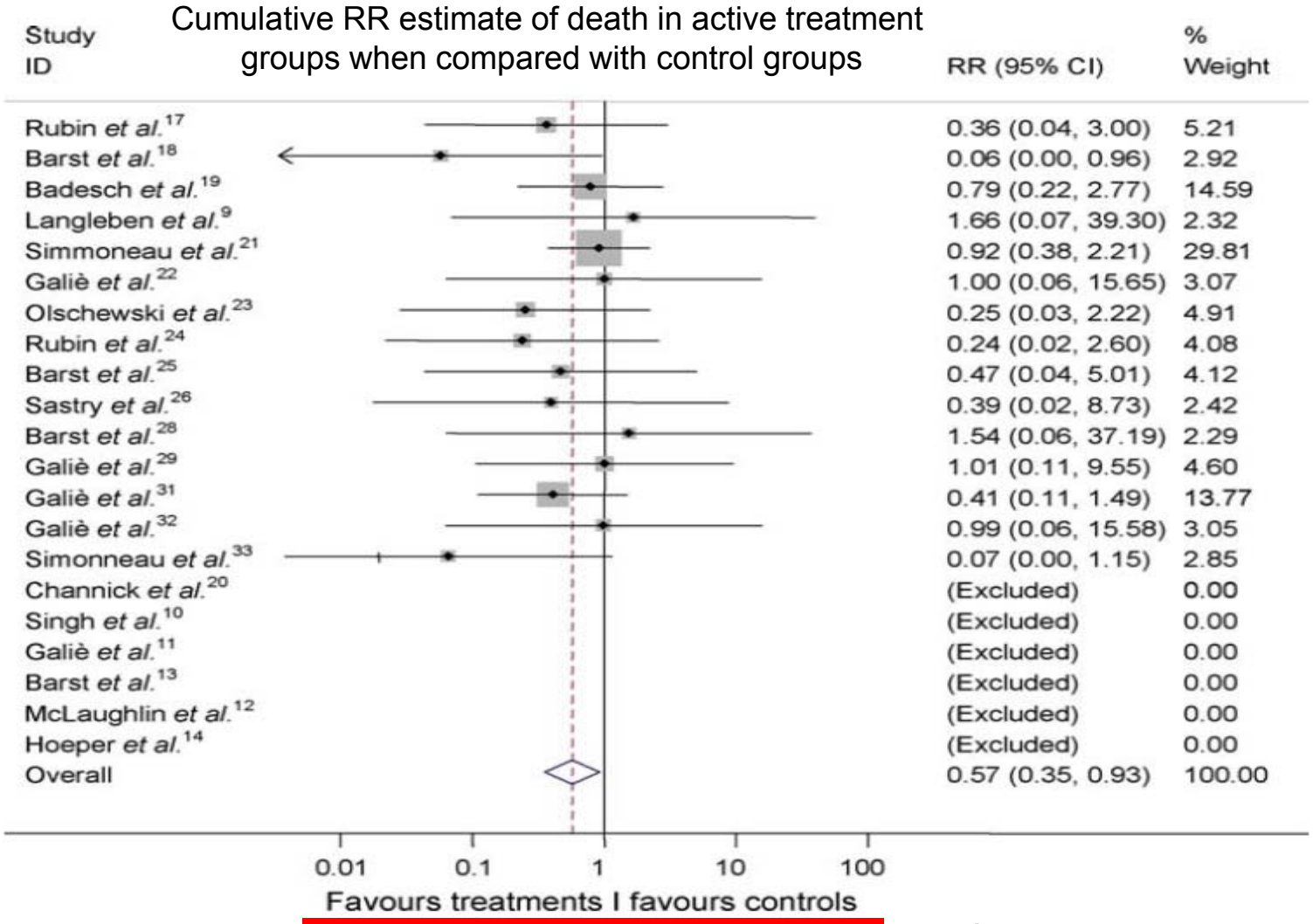
Effective PAH targeted therapies are available



Galiè N. Eur Heat J 2009; 30: 394

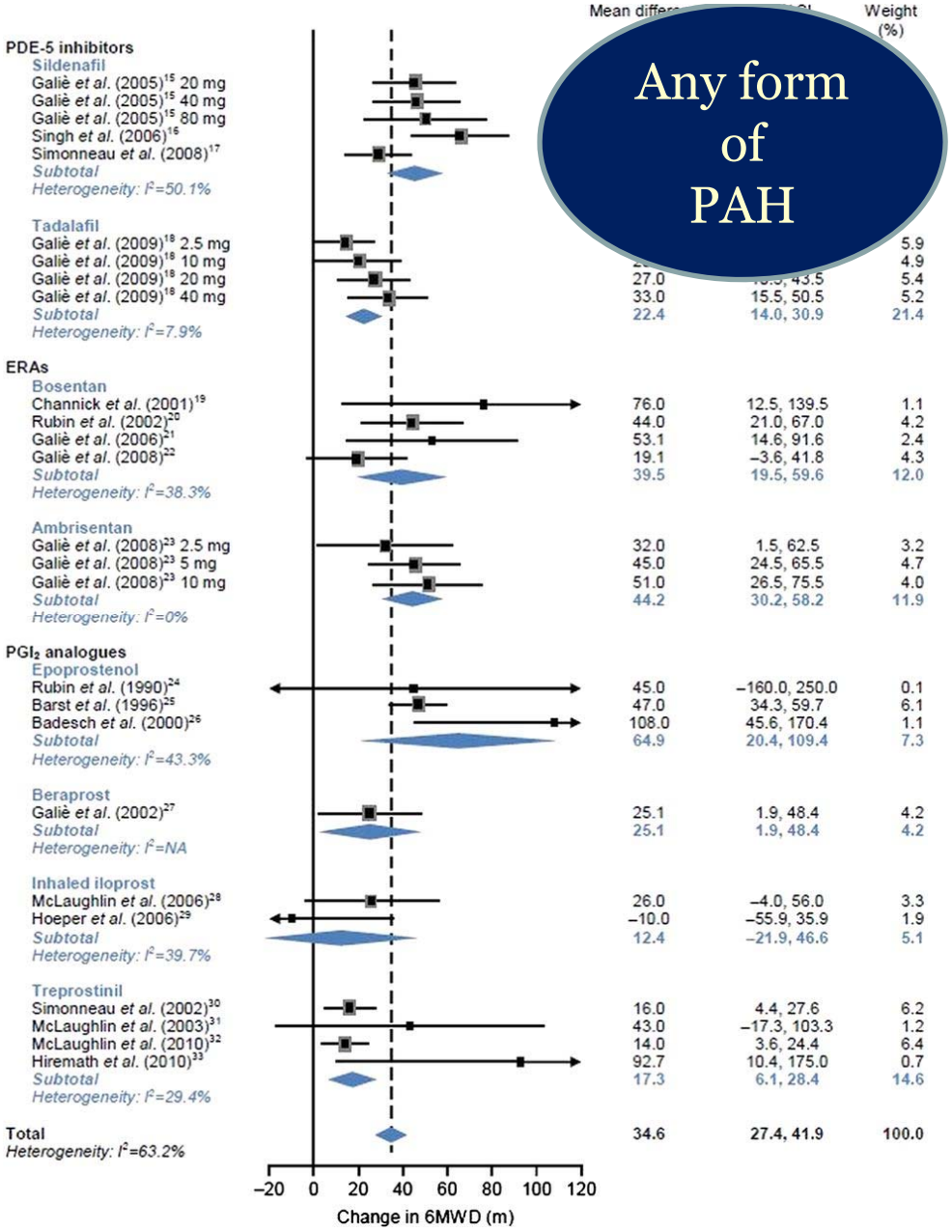
Γιατί χρειάζεται screening

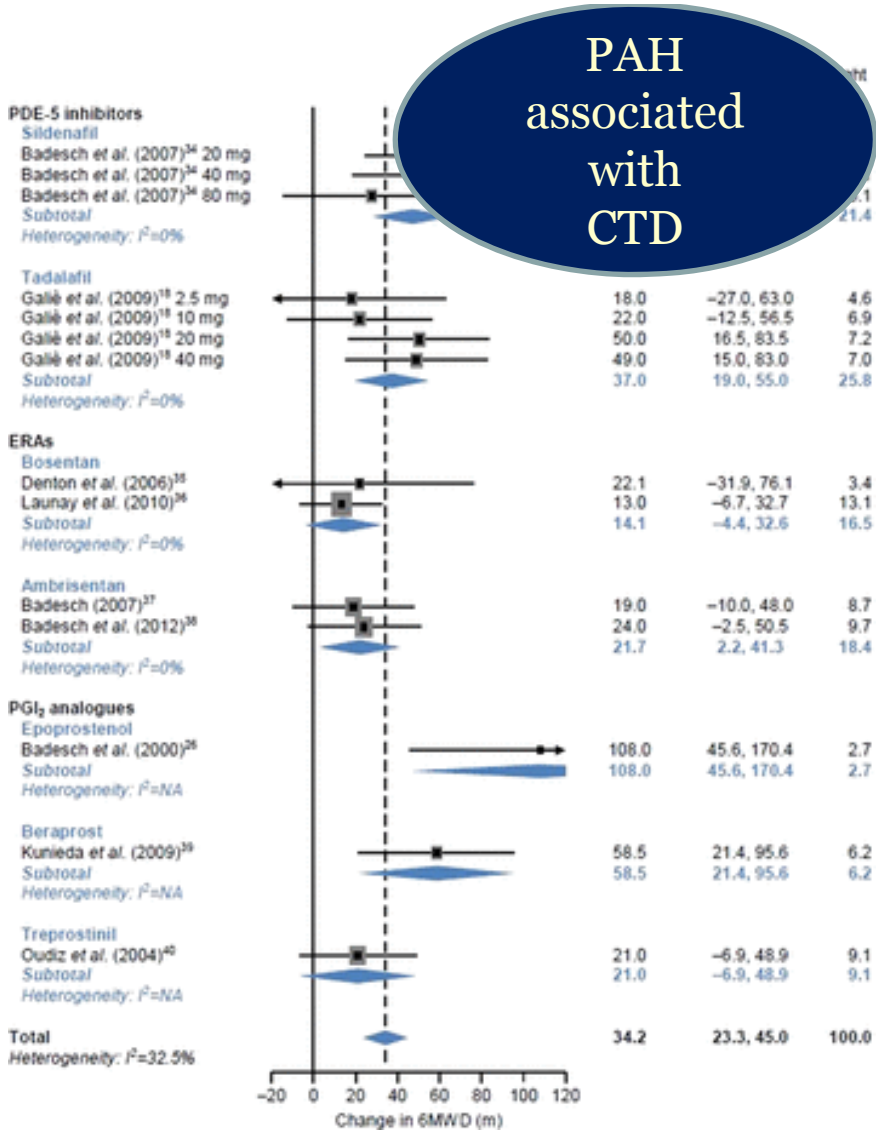
Effective PAH targeted therapies are available



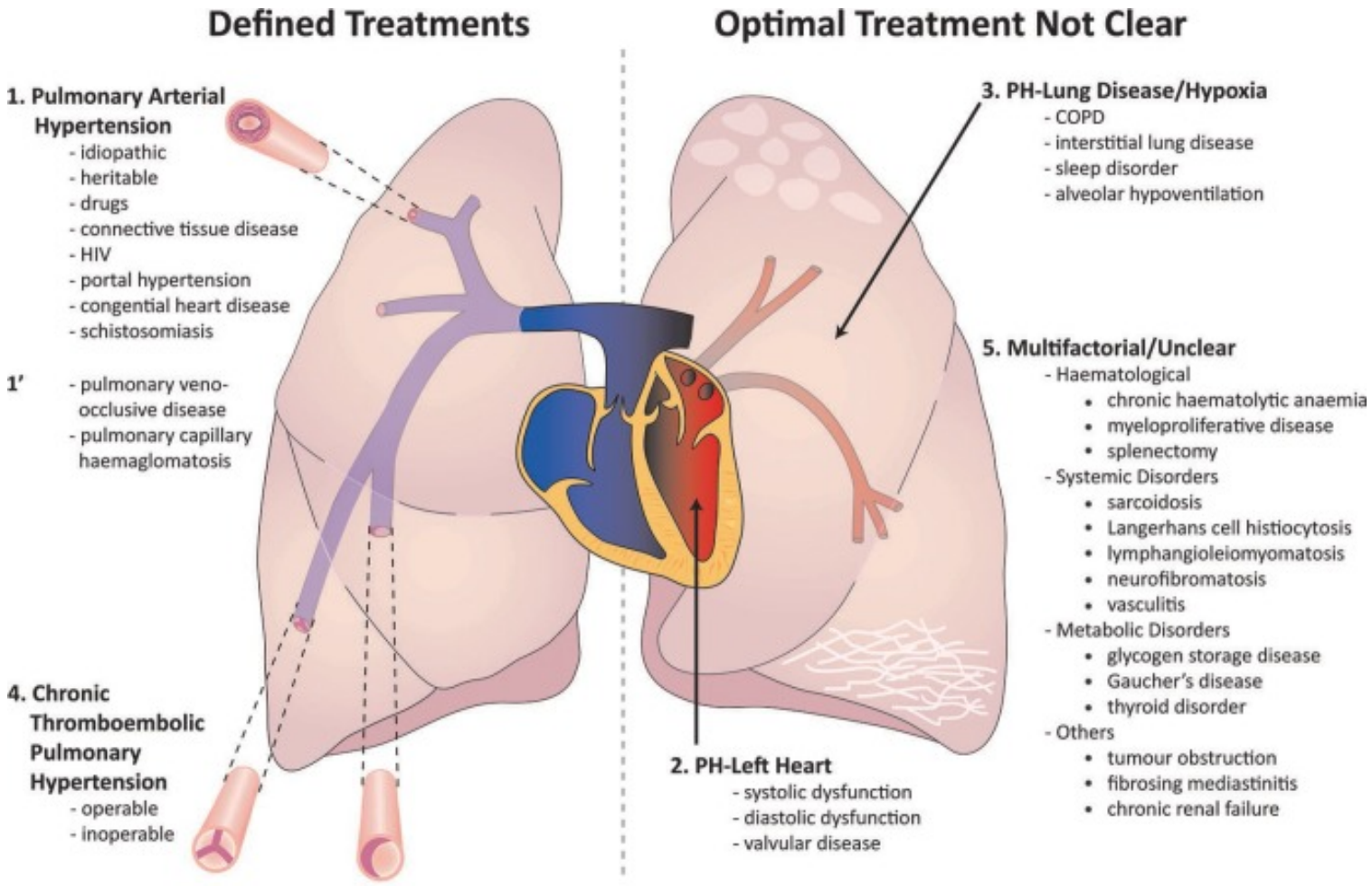
reduction in mortality of 43%

Galiè N. Eur Heat J 2009; 30: 394



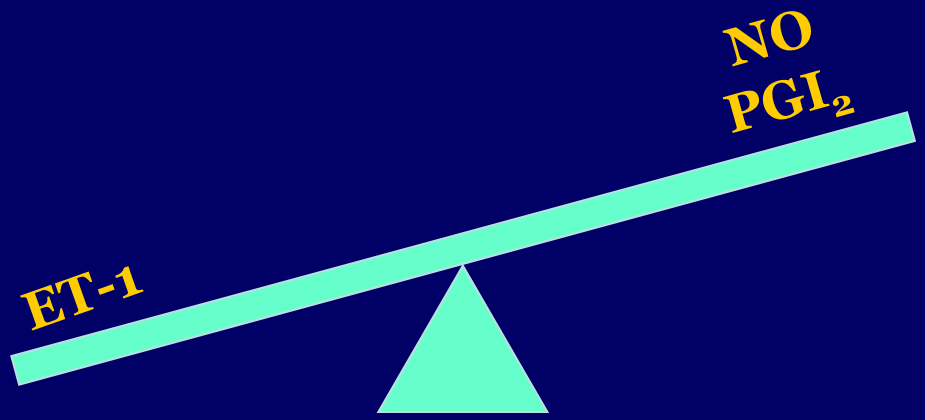


Kuwana Masataka BMJ 2013;3



Condliffe R 2015

Endothelial Cell Dysfunction in PAH



ET-1 is elevated (+)

- **Vasoconstriction**
- **Cell proliferation / Hypertrophy**

NO and PGI_2 are reduced (-)

- **Vasodilation**
- **Anti-proliferation**
- **Anti-inflammation**

Spieker LE et al. J Am Coll Cardiol. 2001;37:1493-1505.
Albrecht EW et al. J Pathol. 2003;199:8-17.
Luscher TF and Barton M. Circulation. 2000;102:2434-2440.
Hankins SR and Horn EM. Curr Cardiol Rep. 2000;2:244-251.

Avoid pregnancy (I-C)
Influenza and pneumococcal immunization (I-C)
Supervised rehabilitation (IIa-B)
Psycho-social support (IIa-C)
Avoid excessive physical activity (III-C)

General measures and supportive therapy

Diuretics (I-C)
Oxygen* (I-C)
Oral anticoagulants:
IPAH, heritable PAH and PAH due to anorexigens (IIa-C)
APAH (IIb–C)
Digoxin (IIb-C)

Expert Referral (I-C)

Acute vasoreactivity test
(I-C for IPAH)
(IIb-C for APAH)

VASOREACTIVE

NON VASOREACTIVE

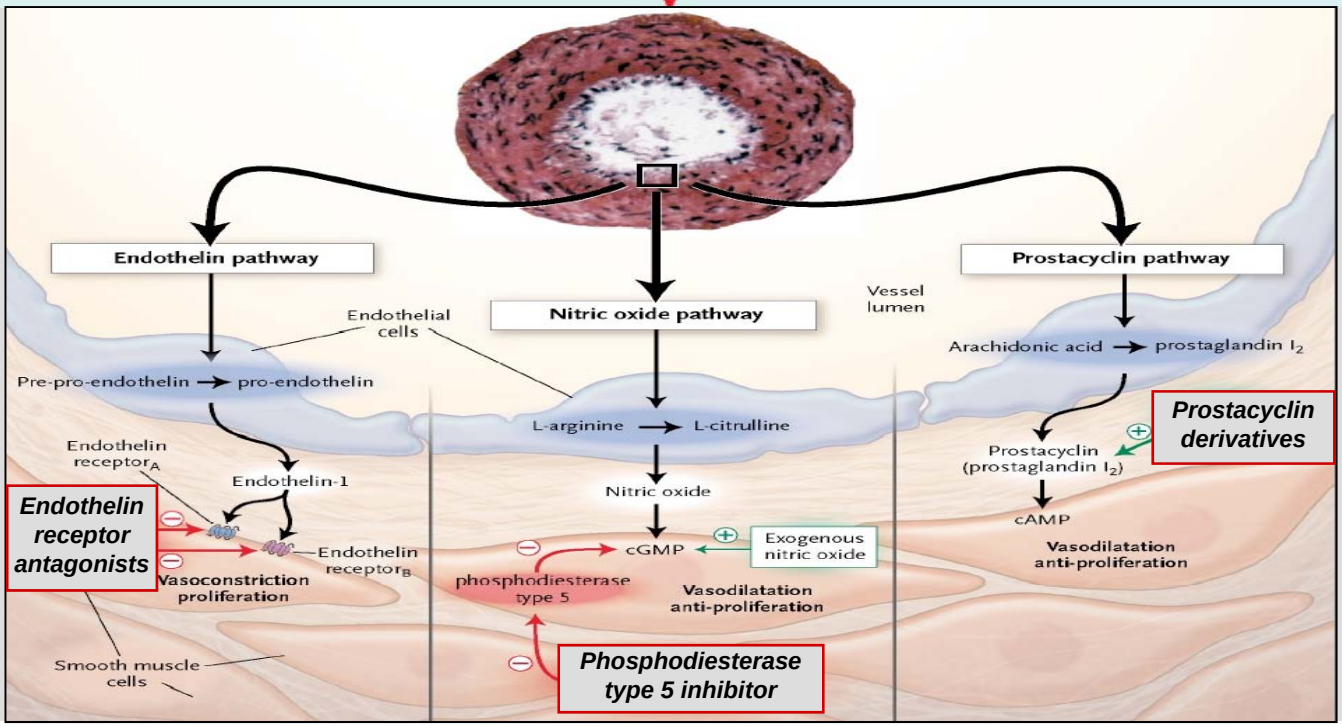
WHO-FC I-III
CCB (I-C)

Sustained response (WHO-FC I-II)

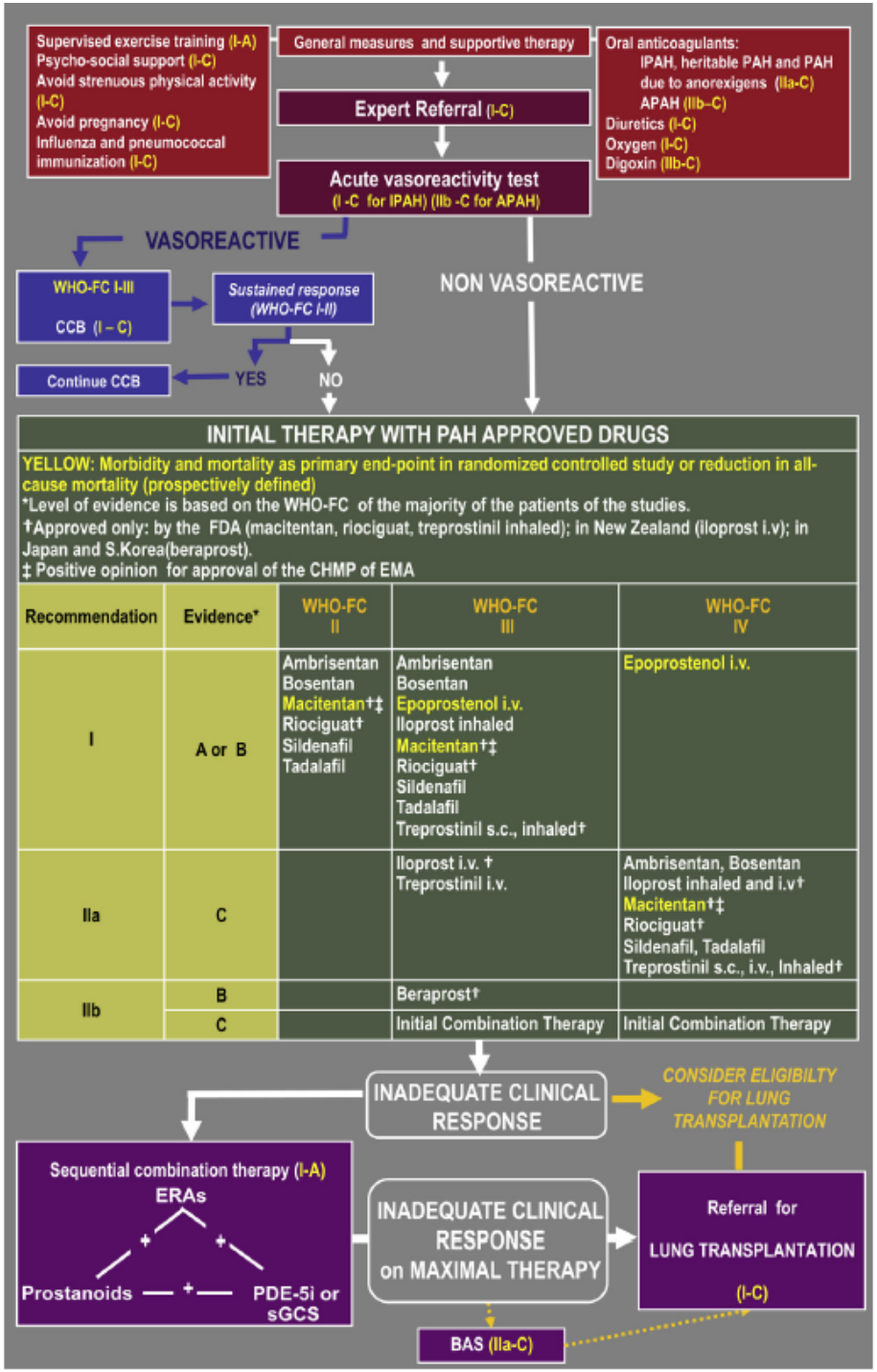
YES

NO

Continue CCB



Θεραπευτικός Αλγόριθμος ΠΑΥ



Γενικά μέτρα

Αρχική θεραπεία

Απάντηση στη θεραπεία

Galie N JACC; 2013 (25)

Derived treatment goals from risk stratification

Assessment parameter	Stable and satisfactory	Stable and not satisfactory	Unstable and deteriorating
Clinical evidence of RV failure	No	Only some of the "green" parameters are fulfilled (Grey zone)	Yes
Rate of progression	Slow		Rapid
Syncope	No		Yes
WHO-FC	I, II		IV
6-MWD	Longer (> 500 m)		Shorter (< 300 m)
CPET	Peak VO_2 > 15 ml/min/kg		Peak VO_2 < 12 ml/min/kg
BNP/NT-proBNP plasma levels	Normal or near-normal		Very elevated and rising
Echocardiographic findings	No pericardial effusion TAPSE > 2.0 cm		Pericardial effusion TAPSE < 1.5 cm
Haemodynamics	RAP < 8 mmHg and CI ≥ 2.5 l/min/m^2		RAP > 15 mmHg or CI ≤ 2.0 l/min/m^2

Galiè N, *et al. Eur Heart J* 2009; 30:2493-537.

Γενικές αρχές έγκαιρης διάγνωσης Πνευμονικής αρτηριακής υπέρτασης σε Νοσήματα του Συνδετικού Ιστού

All patients with SSc should be screened for PAH
Patients with mixed CTD or other CTD with scleroderma features (referred to as scleroderma spectrum disorders) should be screened in a similar way to patients with SSc
Screening of asymptomatic patients is not recommended for mixed CTD or other CTD patients without features of scleroderma (including systemic lupus erythematosus, rheumatoid arthritis, inflammatory myositis and Sjögrens syndrome)
All SSc and scleroderma spectrum patients with a positive, noninvasive screening (as presented in these recommendations) should be referred for RHC
RHC is mandatory for diagnosis of PAH
Acute vasodilator testing is not required as part of the evaluation of PAH in patients with SSc, scleroderma spectrum disorders or other CTD

Schwaiger J ERV 2013;22:515

Πνευμονική υπέρταση σε ασθενείς με ΣΕΛ:

1. Low prevalence of PH

2. Screening in asymptomatic lupus patients are not recommended

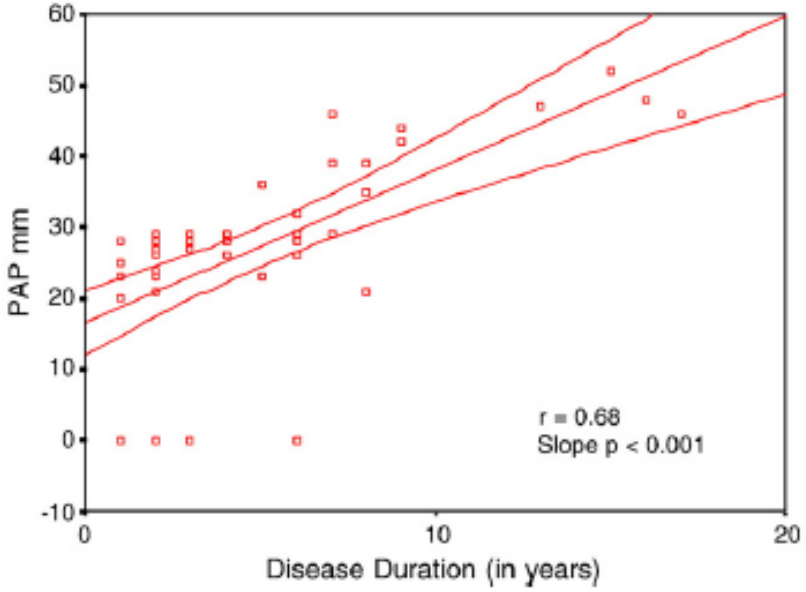
3. Two consecutive PAP values ≥40 mmHg by echocardiogram is the best screening cutoff for starting investigations in SLE patients with suspected PH

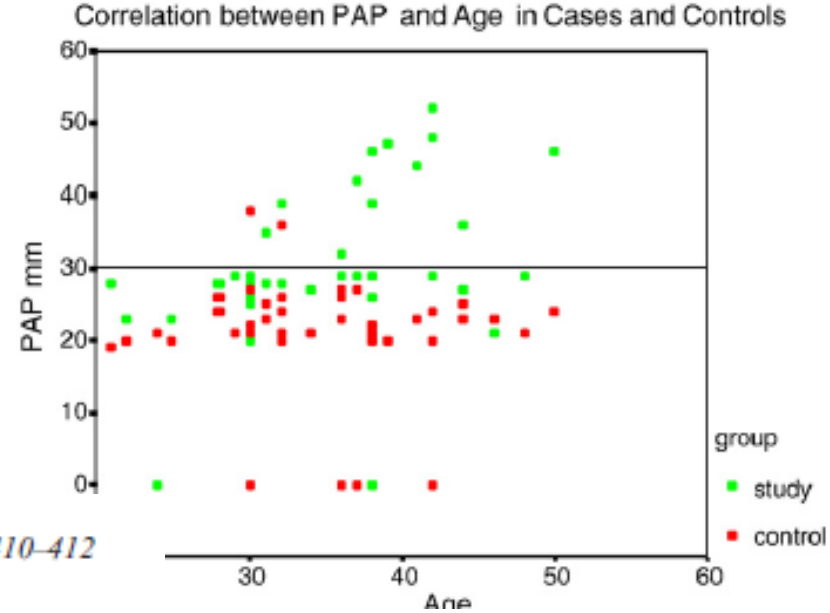
G. Ruiz-Irastorza et al. / Autoimmunity Reviews 12 (2013) 410–415

Πνευμονική υπέρταση σε ασθενείς με
MCTD: ~50% (περιστασιακά απαντούν στα ανοσοκατασταλτικά)

pSS: Σπάνια
Γυναίκες 50 ετών

RA: 1. Μεγαλύτερη ηλικία
2. Μεγαλύτερη διάρκεια νόσου



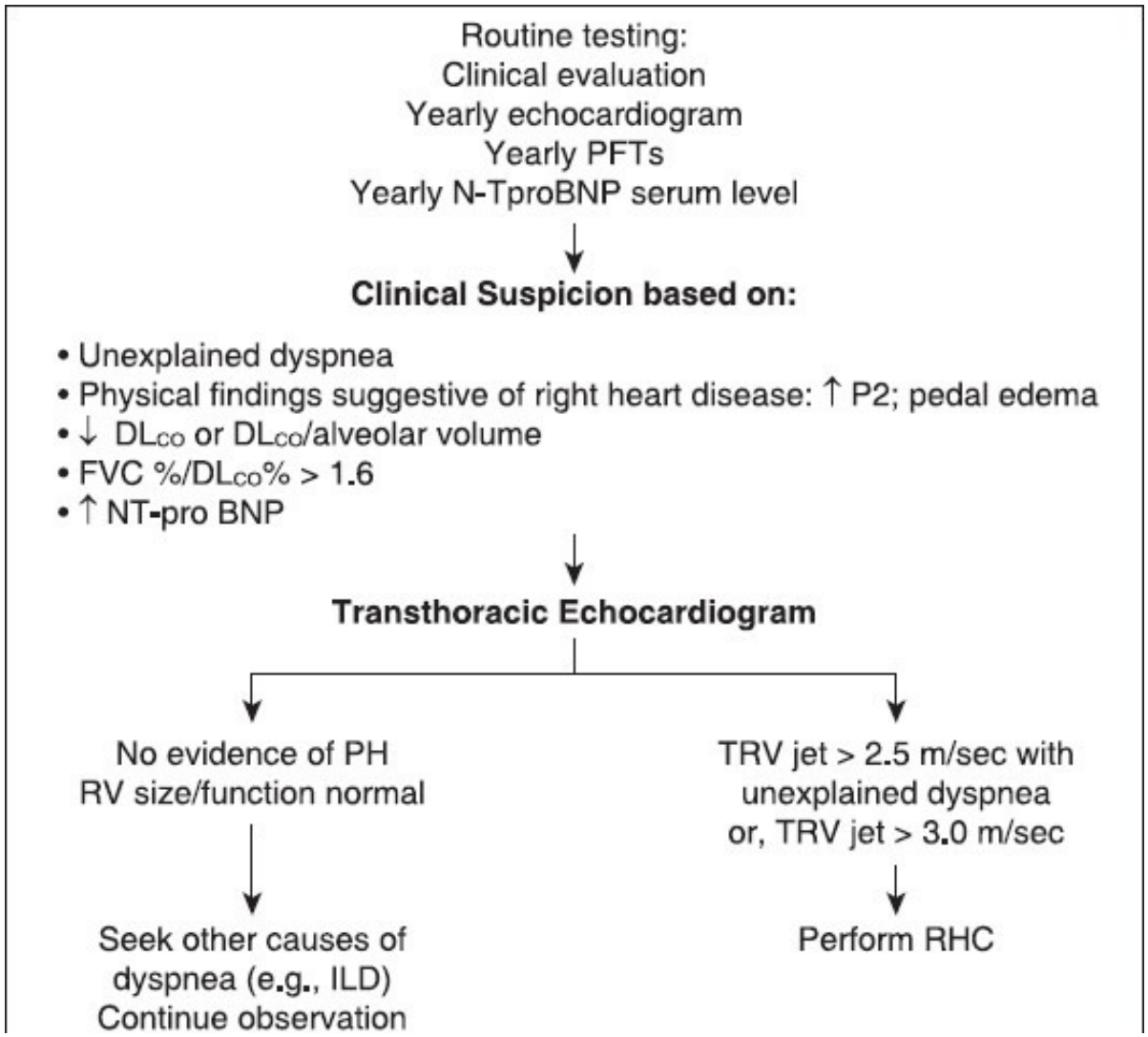


N. Udayakumar et al. / International Journal of Cardiology 127 (2008) 410–412

Γενικές αρχές έγκαιρης διάγνωσης Πνευμονικής αρτηριακής υπέρτασης σε Νοσήματα του Συνδετικού Ιστού

All patients with SSc should be screened for PAH
Patients with mixed CTD or other CTD with scleroderma features (referred to as scleroderma spectrum disorders) should be screened in a similar way to patients with SSc
Screening of asymptomatic patients is not recommended for mixed CTD or other CTD patients without features of scleroderma (including systemic lupus erythematosus, rheumatoid arthritis, inflammatory myositis and Sjögrens syndrome)
All SSc and scleroderma spectrum patients with a positive, noninvasive screening (as presented in these recommendations) should be referred for RHC
RHC is mandatory for diagnosis of PAH
Acute vasodilator testing is not required as part of the evaluation of PAH in patients with SSc, scleroderma spectrum disorders or other CTD

Schwaiger J ERV 2013;22:515



Guidelines PAH in CTD 2014

Boueiz A Ann Thor Med 2014

Take home messages

- **PAH prevalence in SSc is about 8-10%**
- **Current survival of SSc-PAH patients is not acceptable**
- **Diagnosis of PAH in FC I or II dyspnea is challenging**
- **Echocardiography is so far the most effective screening tool to suspect PAH in SSc**
- **Early diagnosis and intervention may translate into better long-term outcomes**