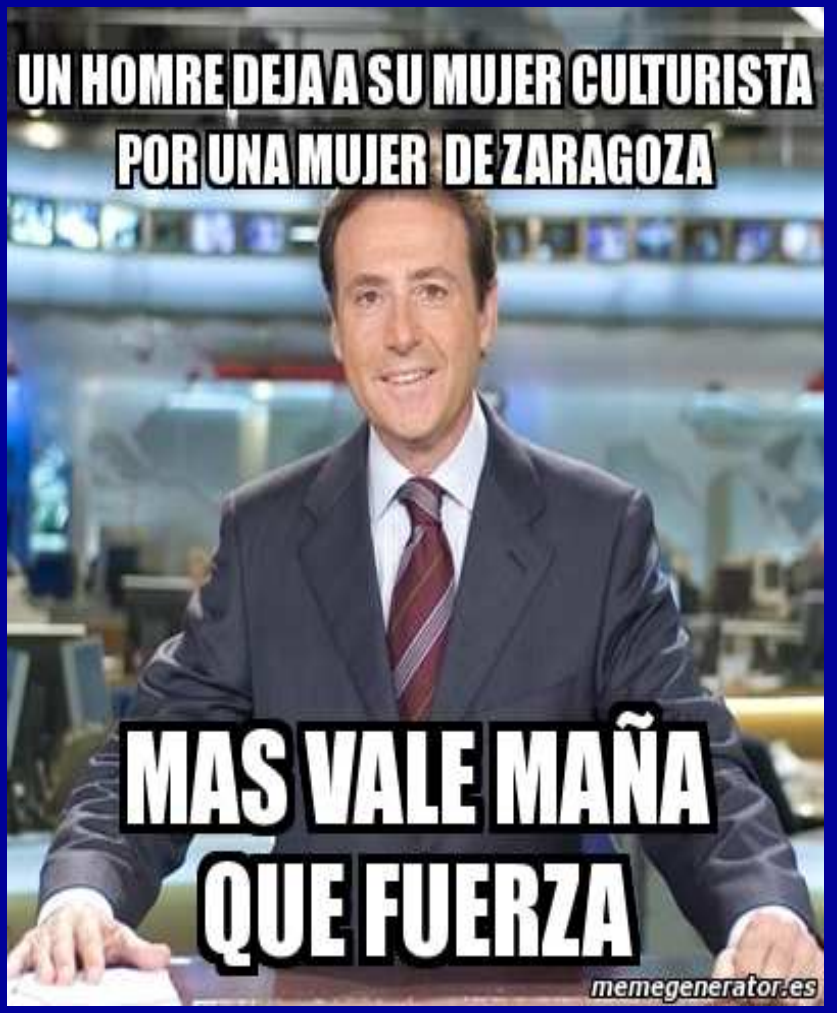
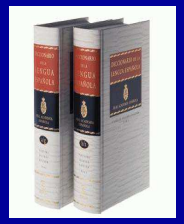
Hipertensión pulmonar en la esclerodermia: una encrucijada diagnóstica

UEAS Unidad de Enfermedades Autoinmunes Sistémicas

José Luis Callejas
Unidad de Enfermedades Sistémicas
H. Clínico San Cecilio de Granada


UN HOMRE DEJA A SU MUJER CULTURISTA
POR UNA MUJER DE ZARAGOZA
MAS VALE MAÑA
QUE FUERZA
memegenerator.es



Encrucijada:

1. Lugar en donde se cruzan dos o más calles o caminos

2. Ocasión que se aprovecha para hacer daño a alguien, emboscada, asechanza

3. Situación difícil en que no se sabe qué conducta seguir

HAP ≠ HP
PAPs ≠ PAPm

- Todos los pacientes con HAP tienen HP
- No todos las HP son HAP

Lugar en donde se cruzan dos o más calles o caminos…
Un paciente con ES e HP puede tener….

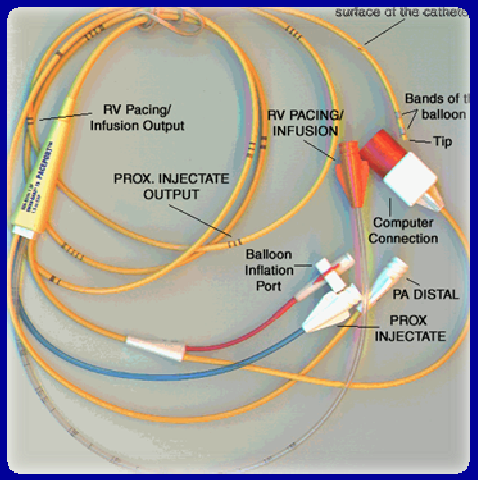
Properly diagnosing pulmonary arterial hypertension
Preston IR.

Am J Cardiol. 2013 Apr 16;111(8 Suppl):2C-9C

Clasificación Dana Point 2008/Niza 2013

Grupo 1: **HAP, incluye las asociadas a ETC**

- **2:** **HP causada por cardiopatía izquierda**
- **3:** **HP por enfermedades pulmonares**
- **4:** **HP tromboembólica crónica**
- **5:** **HP multifactorial**



CATETERISMO CARDIACO DERECHO

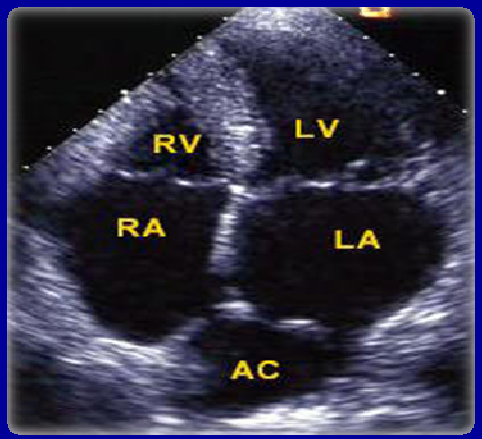
HP: Presión Arterial Pulmonar Media

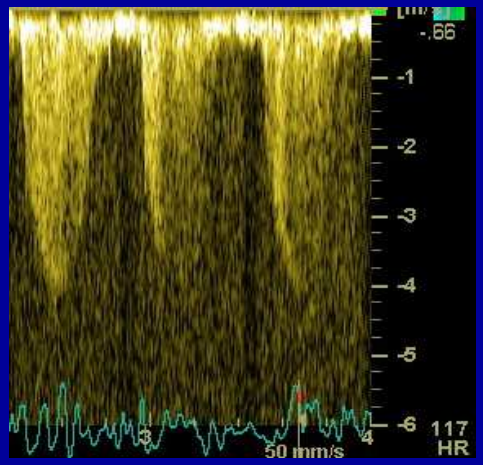
>25mmHg……HP

20-25mmHg…..HP limítrofe

HAP: PCP <15mmHg

HP asociada a cardiopatía: PCP>15





ECOCARDIOGRAMA-DOPPLER

Presión Arterial Pulmonar Sistólica

PSAP= 4 velocidad^2 + Pr Adcha

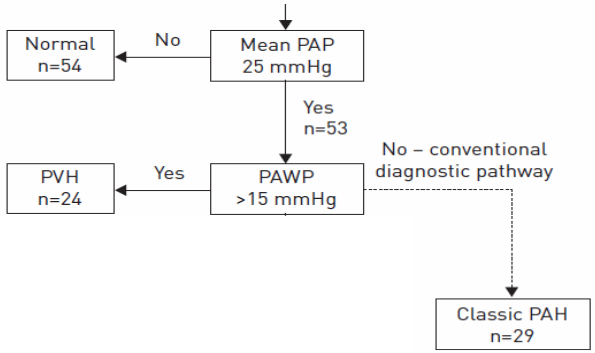
PSAP> 50mmHg

37-50mmHg

High prevalence of occult left heart disease in scleroderma-pulmonary hypertension

Eur Respir J 2013; 42: 1083–1091

PVH had high prevalence in our scleroderma-pulmonary hypertension population. Distinguishing PAH from PVH with only PAWP may result in some PVH patients being misclassified as having PAH.



Un paciente con ES puede tener….

Grupo 1: HAP, incluye las asociadas a ETC

2: HP causada por cardiopatía izquierda

3: HP por enfermedades pulmonares

4: HP tromboembólica crónica

5: HP multifactorial

Situación difícil en que no se sabe qué conducta seguir

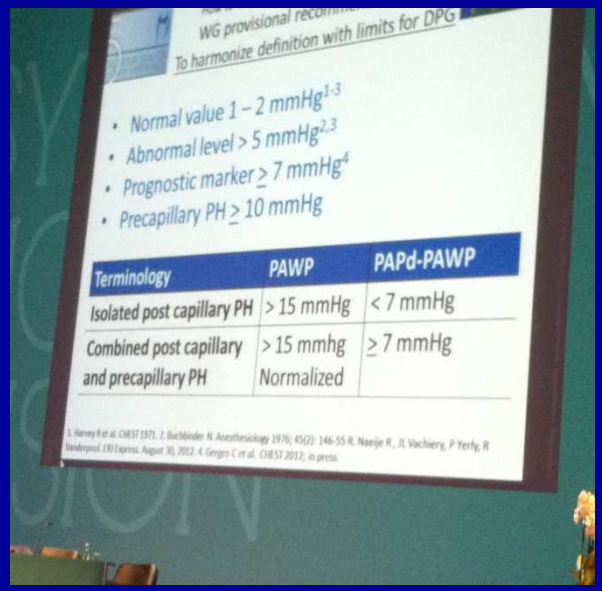
HP ~~reactiva~~

HP desproporcionada

HP PRE Y POSTCAPILAR

Gradiente transpulmonar > 15mmHg

Gradiente diastólico >7mmHg



ARTHRITIS & RHEUMATISM
Vol. 65, No. 4, April 2013, pp 1074–1084

Borderline Mean Pulmonary Artery Pressure in Patients With Systemic Sclerosis

Transpulmonary Gradient Predicts Risk of Developing Pulmonary Hypertension

Christopher J. Valerio, Benjamin E. Schreiber, Clive E. Handler, Christopher P. Denton, and John G. Coghlan

Registry of the Spanish Network for Systemic Sclerosis: Clinical Pattern According to Cutaneous Subsets and Immunological Status

Carmen Pilar Simeón-Aznar, MD, PhD,*
Vicent Fonollosa-Plá, MD, PhD,* Carles Tolosa-Vilella, MD, PhD,†
Gerard Espinosa-Garriga, MD, PhD,‡ Manel Ramos-Casals, MD, PhD,‡
Mercedes Campillo-Grau, PhD,§
Francisco José García-Hernández, MD, PhD,¶
María Jesús Castillo-Palma, MD, PhD,¶
Julio Sánchez-Román, MD, PhD,¶ José Luis Callejas-Rubio, MD, PhD,‖
Norberto Ortego-Centeno, MD, PhD,‖
María Victoria Egurbide Arberas, MD, PhD,**
Luis
Lu
Enrique de Ramón-Garrido, MD, PhD,
Eva María Esteban Marcos, MD,¶¶ Lucio Pallarés-Ferreres, MD, PhD,¶¶
Carmen Hidalgo-Tenorio, MD, PhD,‖‖
José Mario Sabio-Sánchez, MD, PhD,‖‖
Ricardo Gómez-de la Torre, MD, PhD,***
Gonzalo Salvador-Cervello, MD,††† Juan José Rios-Blanco, MD, PhD,‡‡‡
Antonio Gil-Aguado, MD,‡‡‡ and Miquel Vilardell-Tarrés, MD, PhD*

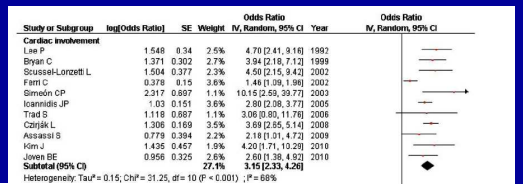
Table 2 Cumulative Clinical Manifestations Among Patients with SSc Patients According to Their Cutaneous Subsets

	a lcSSc (%)	b dcSSc (%)	c ssSSc (%)	*P* Value a vs b	*P* Value a vs c	*P* Value b vs c
PAH	91 (16.2)	53 (21.8)	17 (24.6)	ns	ns	ns

Semin Arthritis Rheum 41:789-800 2012

The impact of cardiopulmonary manifestations on the mortality of SSc: a systematic review and meta-analysis of observational studies

Rheumatology 2012;51:1027–1036



Study or Subgroup	log[Odds Ratio]	SE	Weight	Odds Ratio IV, Random, 95% CI	Year
Cardiac involvement					
Lee P	1.548	0.34	2.5%	4.70 [2.41, 9.16]	1992
Bryan C	1.371	0.302	2.7%	3.94 [2.18, 7.12]	1999
Scussel-Lorzetti L	1.504	0.377	2.3%	4.50 [2.15, 9.42]	2002
Ferri C	0.378	0.15	3.6%	1.46 [1.09, 1.96]	2002
Simeón CP	2.317	0.897	1.1%	10.15 [2.59, 39.77]	2003
Ioannidis JP	1.03	0.151	3.6%	2.80 [2.08, 3.77]	2005
Trad S	1.118	0.887	1.1%	3.06 [0.80, 11.76]	2006
Czirják L	1.306	0.169	3.5%	3.69 [2.65, 5.14]	2008
Assassi S	0.779	0.394	2.2%	2.18 [1.01, 4.72]	2009
Kim J	1.435	0.457	1.9%	4.20 [1.71, 10.28]	2010
Joven BE	0.956	0.325	2.6%	2.60 [1.38, 4.92]	2010
Subtotal (95% CI)			**27.1%**	**3.15 [2.33, 4.26]**	

Heterogeneity: $Tau^2 = 0.15$; $Chi^2 = 31.25$, df = 10 ($P < 0.001$); $I^2 = 68\%$

Study or Subgroup	log[Odds Ratio]	SE	Weight	Odds Ratio IV, Random, 95% CI	Year
Pulmonary arterial hypertension					
Simeón CP	2.382	0.979	0.6%	10.83 [1.59, 73.76]	2003
Trad S	1.409	0.525	1.6%	4.09 [1.46, 11.45]	2006
Hachulla E	2.343	0.296	2.7%	10.41 [5.83, 18.60]	2009
Joven BE	0.788	0.307	2.7%	2.20 [1.20, 4.01]	2010
Tyndall A	0.702	0.172	3.5%	2.02 [1.44, 2.83]	2010
Hissaria P	0.775	0.223	3.2%	2.17 [1.40, 3.36]	2010
Subtotal (95% CI)			**14.4%**	**3.50 [1.94, 6.30]**	

Heterogeneity: $Tau^2 = 0.27$; $Chi^2 = 43.40$, df = 11 ($P < 0.001$); $I^2 = 75\%$
Test for overall effect: $Z = 5.38$ ($P < 0.001$)

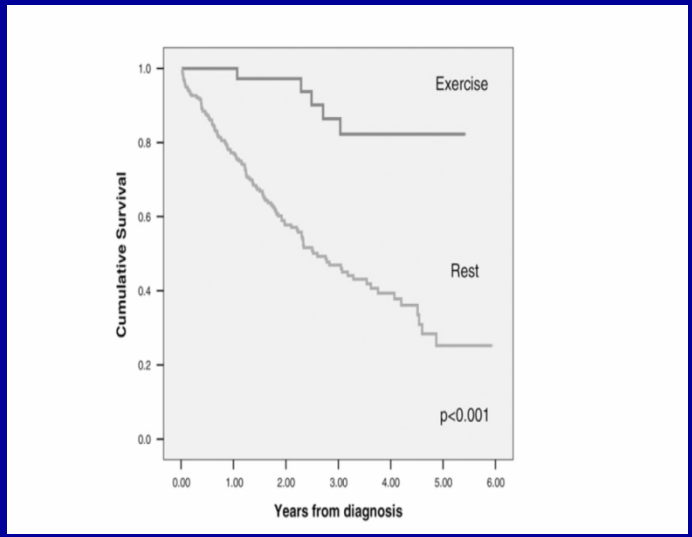
0.1 0.2 0.5 1 2 5 10

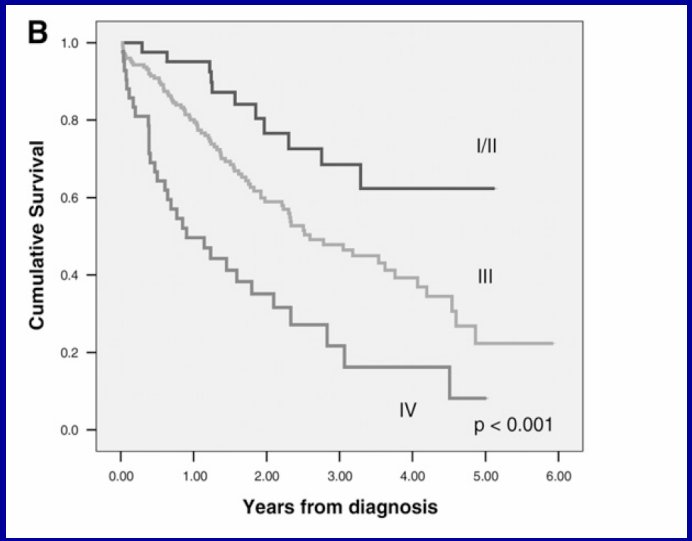
Odds Ratio
IV, Random, 95% CI

Connective Tissue Disease–associated Pulmonary Arterial Hypertension in the Modern Treatment Era

Robin Condliffe[1,2], David G. Kiely[1], Andrew J. Peacock[3], Paul A. Corris[4,5], J. Simon R. Gibbs[6], Florenc Vrapi[7], Clare Das[7], Charlie A. Elliot[1], Martin Johnson[3], Julia DeSoyza[4], Chantal Torpy[6], Kim Goldsmith[2], Denise Hodgkins[2], Rodney J. Hughes[2], Joanna Pepke-Zaba[2], and J. Gerry Coghlan[7]

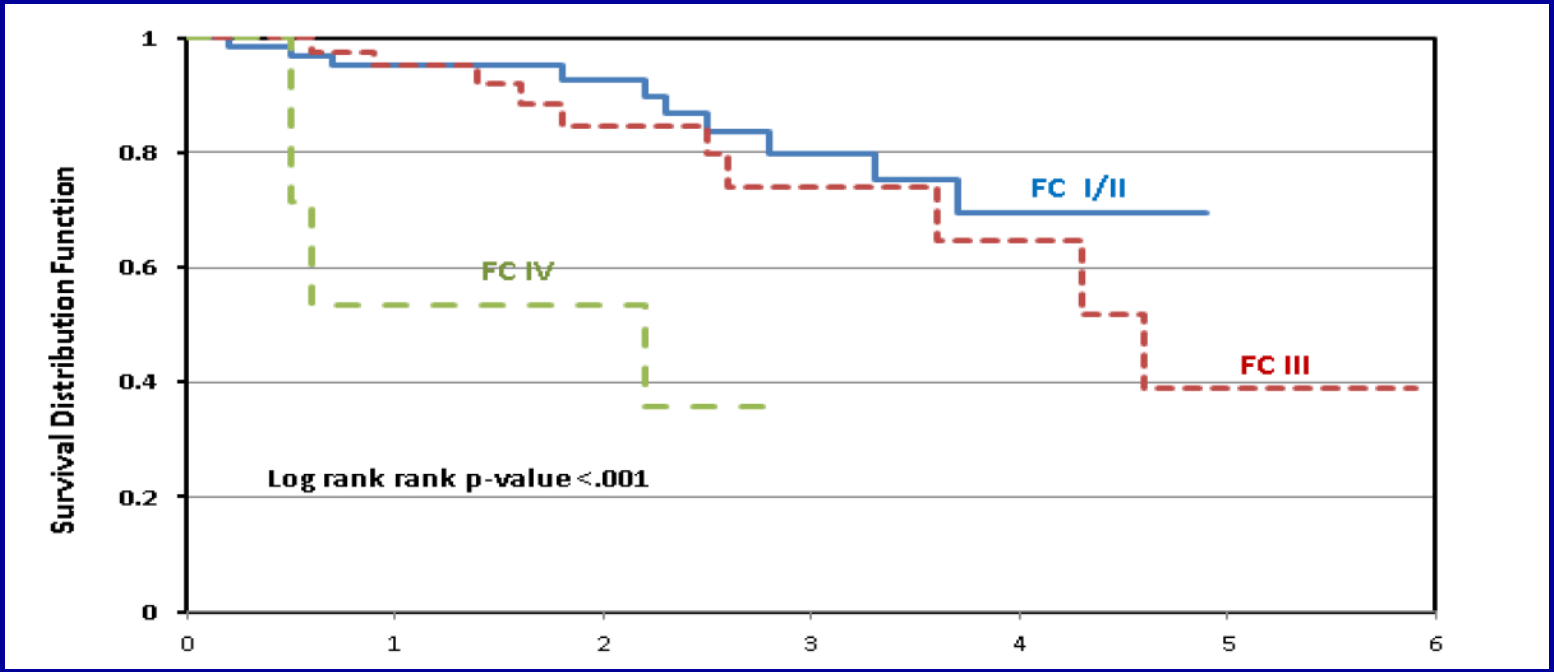
Am J Respir Crit Care Med Vol 179. pp 151–157, 2009





Survival and Predictors of Mortality in Systemic Sclerosis Associated Pulmonary Arterial Hypertension: Outcomes from the PHAROS Registry

Arthritis Care & Research



¿Podemos predecir la gravedad de la hipertensión arterial pulmonar en pacientes con esclerodermia?

María Isabel Acosta Colmán[a,*], Gabriela Avila Pedretti[a], María Eugenia Acosta, Carmen Pilar Simeón Aznar[b], Vicent Fonollosa Plá[b] y Miquel Villardel Torrés[b]

Reumatol Clin. 2012;8(5):259–262

Conclusiones: La disminución de la DLCO y el aumento de la VRT son factores predictores de HTAP, que al mismo tiempo condiciona un peor pronóstico en los pacientes con ES.

ARTHRITIS & RHEUMATISM
Vol. 65, No. 9, September 2013, pp 2412–2423

Survival and Prognostic Factors in Systemic Sclerosis–Associated Pulmonary Hypertension

A Systematic Review and Meta-Analysis

Guillaume Lefèvre,[1] Luc Dauchet,[2] Eric Hachulla,[1] David Montani,[3] Vincent Sobanski,[1] Marc Lambert,[4] Pierre-Yves Hatron,[4] Marc Humbert,[3] and David Launay[1]

ARTHRITIS & RHEUMATISM
Vol. 65, No. 9, September 2013, pp 2412–2423

Survival and Prognostic Factors in Systemic Sclerosis–Associated Pulmonary Hypertension

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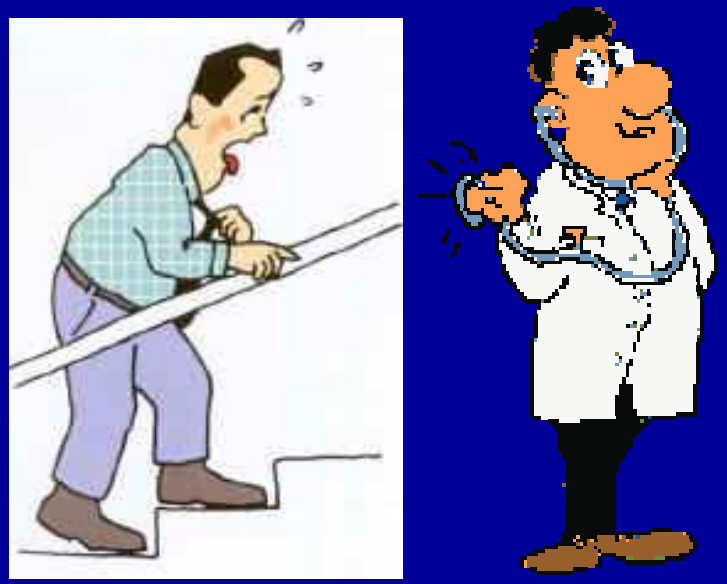
Guillaume Lefèvre,[1] Luc Dauchet,[2] Eric Hachulla,[1] David Montani,[3] Vincent Sobanski,[1] Marc Lambert,[4] Pierre-Yves Hatron,[4] Marc Humbert,[3] and David Launay[1]

In patients with SSc complicated by pulmonary arterial hypertension (PAH), age, male sex, diffusing capacity for carbon monoxide (DLco), pericardial effusion, and the parameters classically associated with the severity of idiopathic PAH, including the 6-minute walk distance, mean pulmonary artery pressure, cardiac index, and right atrial pressure, were significant prognostic factors.

HAP



Alto riesgo: ES

Survival in pulmonary hypertension in Spain: insights from the Spanish registry

Pilar Escribano-Subias*, Isabel Blanco#, Manuel López-Meseguer¶, Carmen Jimenez Lopez-Guarch*, Antonio Roman¶, Pilar Morales+, María Jesús Castillo-Palma§, Javier Segoviaƒ, Miguel A. Gómez-Sanchez* and Joan Albert Barberà# on behalf of the REHAP investigators**

Eur Respir J 2012; 40: 596–603

	PAH				
	IPAH	CTD	CHD	HIV	PoPH
Subjects n (%)	314 (30)	157 (15)	167 (16)	54 (5)	61 (6)
Age yrs	46±18	54±15 *	36±17*	39±5*	52±10*
Males %	27	10*	27	42*	49
6MWD m	382±117	309±115*	365±101	416±126	396±109
WHO FC %					
I–II	29	21	38	40	53
III	59	64	54	40	46
IV	11	15	8	19	2


¿POR QUÉ FROILAN SE DISPARÓ
UN TIRO EN UN PIE?
TONTERIAS.COM

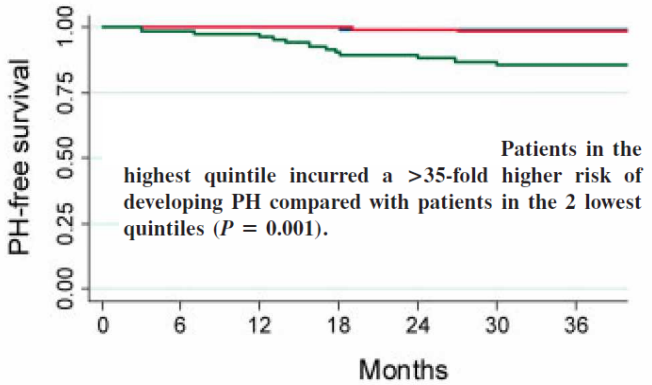
ARTHRITIS & RHEUMATISM
Vol. 63, No. 9, September 2011, pp 2790–2796

Prediction of Pulmonary Hypertension Related to Systemic Sclerosis by an Index Based on Simple Clinical Observations

Christophe Meune,[1] Jérôme Avouac,[1] Paolo Airò,[2] Lorenzo Beretta,[3] Philippe Dieudé,[4] Karim Wahbi,[1] Paola Caramaschi,[5] Kiet Tiev,[6] Susanna Cappelli,[7] Elisabeth Diot,[8] Alessandra Vacca,[9] Jean-Luc Cracowski,[10] Jean Sibilia,[11] André Kahan,[1] Marco Matucci-Cerinic,[7] and Yannick Allanore[1]

Calculation of the Cochin risk prediction score (RPS)

. We calculated the Cochin RPS score as follows: RPS = 0.0001107(age) + 0.0207818(100 − FVC) + 0.04905(150 − DLco/alveolar volume).



Patients in the highest quintile incurred a >35-fold higher risk of developing PH compared with patients in the 2 lowest quintiles (P = 0.001).

Arthritis Care & Research
Vol. 64, No. 3, March 2012, pp 303–310
DOI 10.1002/acr.20693

REVIEW ARTICLE

Practical Approach to Screening for Scleroderma-Associated Pulmonary Arterial Hypertension

ARYEH FISCHER,[1] TODD M. BULL,[2] AND VIRGINIA D. STEEN[3]

Table 3. Decision algorithm for screening and performing RHC in scleroderma*

	Low risk	Mild risk	Moderate risk	High risk
Dyspnea, or Raynaud's phenomenon duration >8 years, or positive for anticentromere, or positive for isolated nucleolar-ANAs	No	Yes	Yes	Yes
DLco% (without extensive emphysema or ILD)	>70	>70	<70	<60
FVC%/DLco%	<1.6	<1.6	>1.6	>1.6
RVSP, mm Hg	<35	<35	>35	>40
Next step	Repeat PFTs annually Repeat echocardiogram in 2–3 years	Repeat PFTs and echocardiogram annually	Consider repeat echocardiogram in 3–6 months or proceed to RHC	Proceed to RHC

* The presence of echocardiographic features of right ventricular hypokinesis or dilatation or an increased B-type natriuretic peptide (BNP) and N-terminal proBNP in a dyspneic scleroderma patient should lead to right-sided heart catheterization (RHC) irrespective of the estimated right ventricular systolic pressure (RSVP). ANAs = antinuclear antibodies; DLco = diffusing capacity for carbon monoxide; ILD = interstitial lung disease; FVC = forced vital capacity; PFT = pulmonary function testing.

Internal Medicine Journal **43** (2013)

ORIGINAL ARTICLES

Performance of screening algorithms in systemic sclerosis-related pulmonary arterial hypertension: a systematic review

V. Thakkar,[1,3] W. M. Stevens,[1] O. A. Moore[1,4] and M. Nikpour[1,2]

Table 1 Screening algorithms in prevalent studies

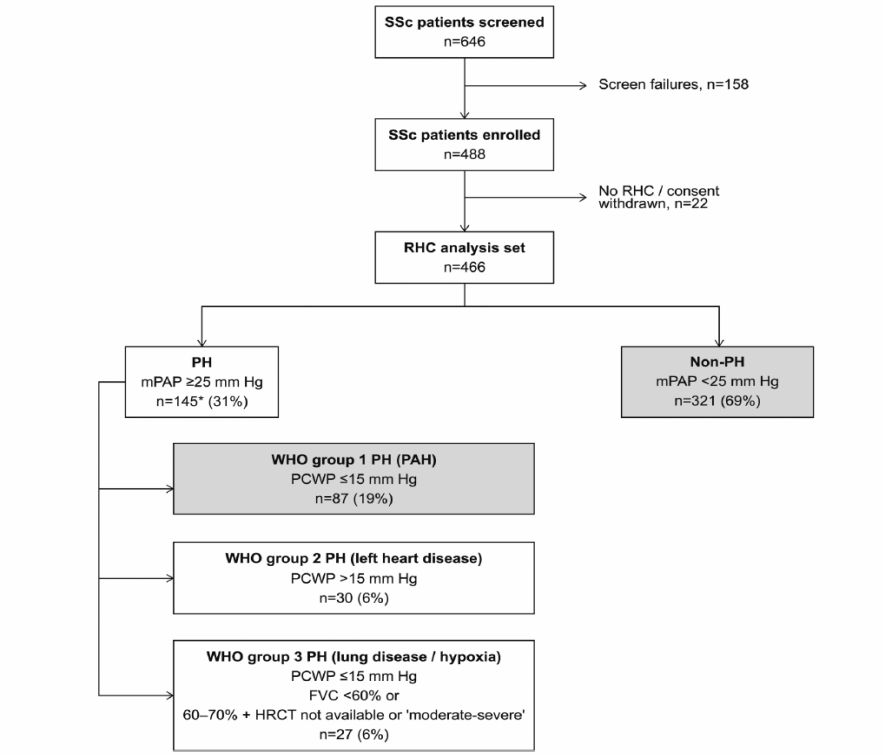
	Avouac *et al.*[1]	Mukerjee *et al.*[24]	Hachulla *et al.*[21]	Jansa *et al.*[23]	Cavagna *et al.*[18]	Ciurzynski *et al.*[19]
Intent to Screen, *n*	1165	722	570	203	135	71
Screening method	$sPAP_{TTE}$ >40 mmHg or DLCO <50% or unexplained dyspnoea	TG >35 mmHg or DLCO <50% or DLCO ↓20% or unexplained dyspnoea	VTR >3 m/s; or VTR 2.5–3m/s & unexplained dyspnoea	TG >30 mmHg	$sPAP_{TTE}$ >36 mmHg or DLCO <50% or DLCO↓20% or unexplained dyspnoea	TG >31 mmHg or ↑TG by 20 mmHg on exercise
Definition of precapillary PH	Rest	Rest or exercise	Rest or exercise	Rest	Rest	Rest or exercise

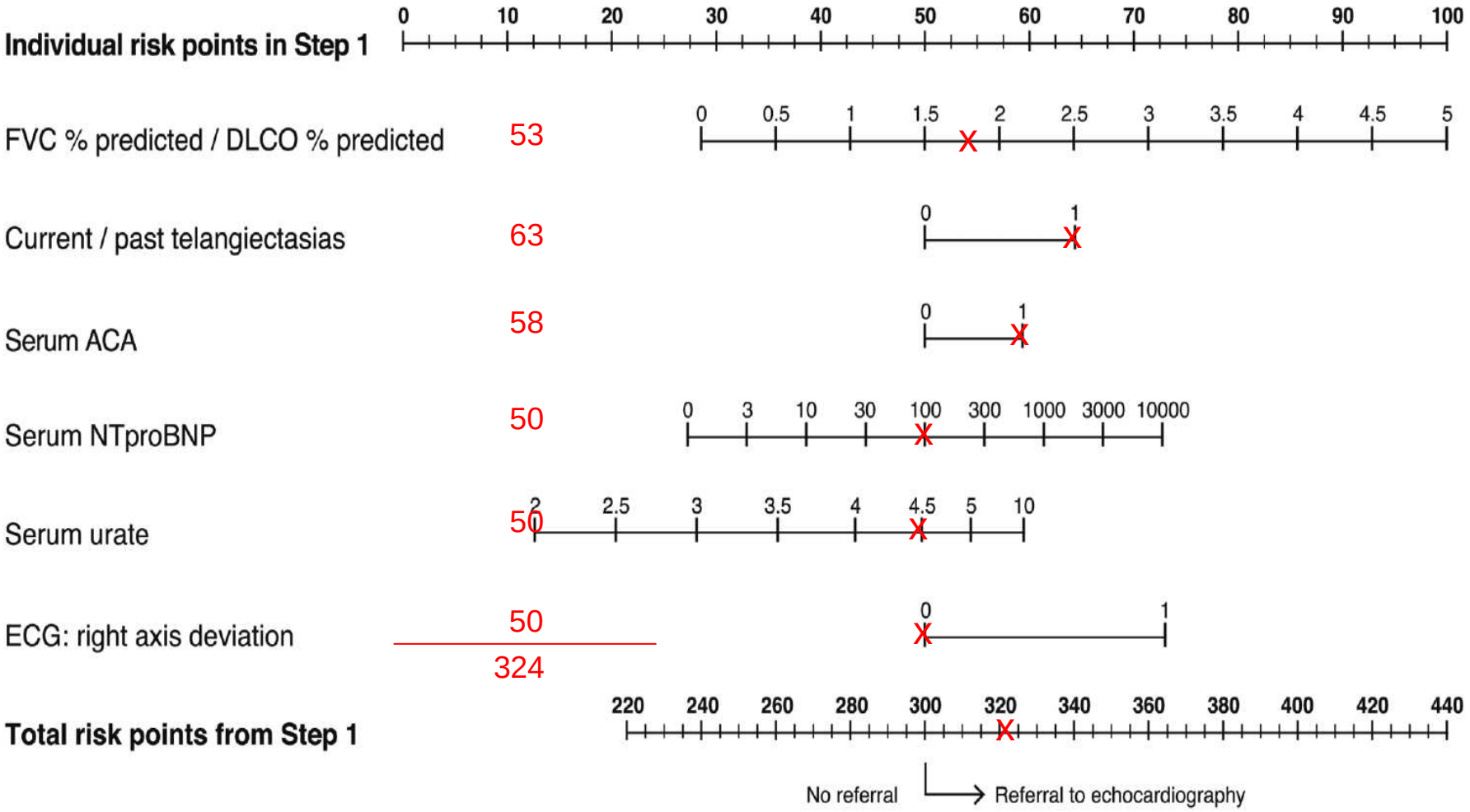
Conclusion: The low to moderate PPV of current screening algorithms, coupled with the inability to determine accurately the negative predictive value, sensitivity and specificity, suggests that there is a need to validate further these algorithms before making recommendations regarding screening for SSc-PAH.

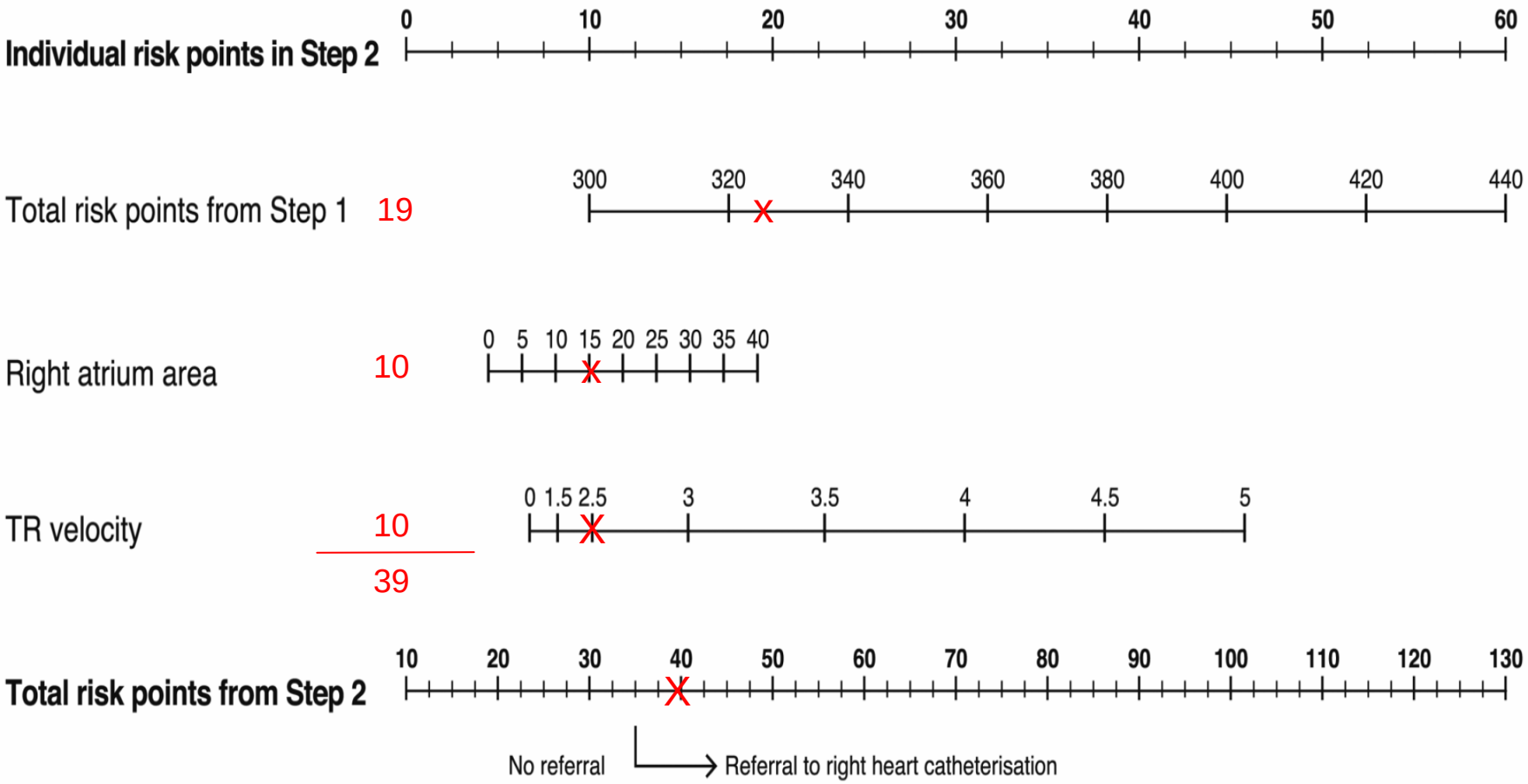
Evidence-based detection of pulmonary arterial hypertension in systemic sclerosis: the DETECT study

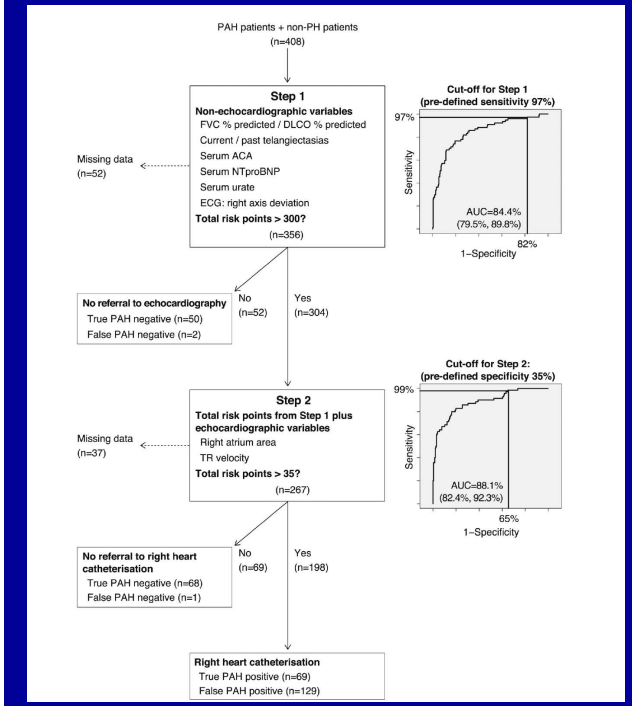
J Gerry Coghlan,[1] Christopher P Denton,[2] Ekkehard Grünig,[3] Diana Bonderman,[4] Oliver Distler,[5] Dinesh Khanna,[6] Ulf Müller-Ladner,[7] Janet E Pope,[8] Madelon C Vonk,[9] Martin Doelberg,[10] Harbajan Chadha-Boreham,[11] Harald Heinzl,[12] Daniel M Rosenberg,[11] Vallerie V McLaughlin,[6] James R Seibold,[13] on behalf of the DETECT study group

Ann Rheum Dis 2013;**00**:1–10.




Individual risk points in Step 1
0 10 20 30 40 50 60 70 80 90 100
FVC % predicted / DLCO % predicted
53
0 0.5 1 1.5 2 2.5 3 3.5 4 4.5 5
Current / past telangiectasias
63
0 1
Serum ACA
58
0 1
Serum NTproBNP
50
0 3 10 30 100 300 1000 3000 10000
Serum urate
50
2 2.5 3 3.5 4 4.5 5 10
ECG: right axis deviation
50
0 1
324
Total risk points from Step 1
220 240 260 280 300 320 340 360 380 400 420 440
No referral
Referral to echocardiography


Individual risk points in Step 2
0 10 20 30 40 50 60
Total risk points from Step 1
19
300 320 340 360 380 400 420 440
Right atrium area
10
0 5 10 15 20 25 30 35 40
TR velocity
10
0 1.5 2.5 3 3.5 4 4.5 5
39
Total risk points from Step 2
10 20 30 40 50 60 70 80 90 100 110 120 130
No referral
Referral to right heart catheterisation


PAH patients + non-PH patients
(n=408)
Step 1
Non-echocardiographic variables
FVC % predicted / DLCO % predicted
Current / past telangiectasias
Serum ACA
Serum NTproBNP
Serum urate
ECG: right axis deviation
Total risk points > 300?
(n=356)
Missing data
(n=52)
No
(n=52)
Yes
(n=304)
No referral to echocardiography
True PAH negative (n=50)
False PAH negative (n=2)
Cut-off for Step 1
(pre-defined sensitivity 97%)
97%
Sensitivity
AUC=84.4%
(79.5%, 89.8%)
82%
1−Specificity
Step 2
Total risk points from Step 1 plus
echocardiographic variables
Right atrium area
TR velocity
Total risk points > 35?
(n=267)
Missing data
(n=37)
No
(n=69)
Yes
(n=198)
No referral to right heart
catheterisation
True PAH negative (n=68)
False PAH negative (n=1)
Cut-off for Step 2:
(pre-defined specificity 35%)
99%
Sensitivity
AUC=88.1%
(82.4%, 92.3%)
65%
1−Specificity
Right heart catheterisation
True PAH positive (n=69)
False PAH positive (n=129)

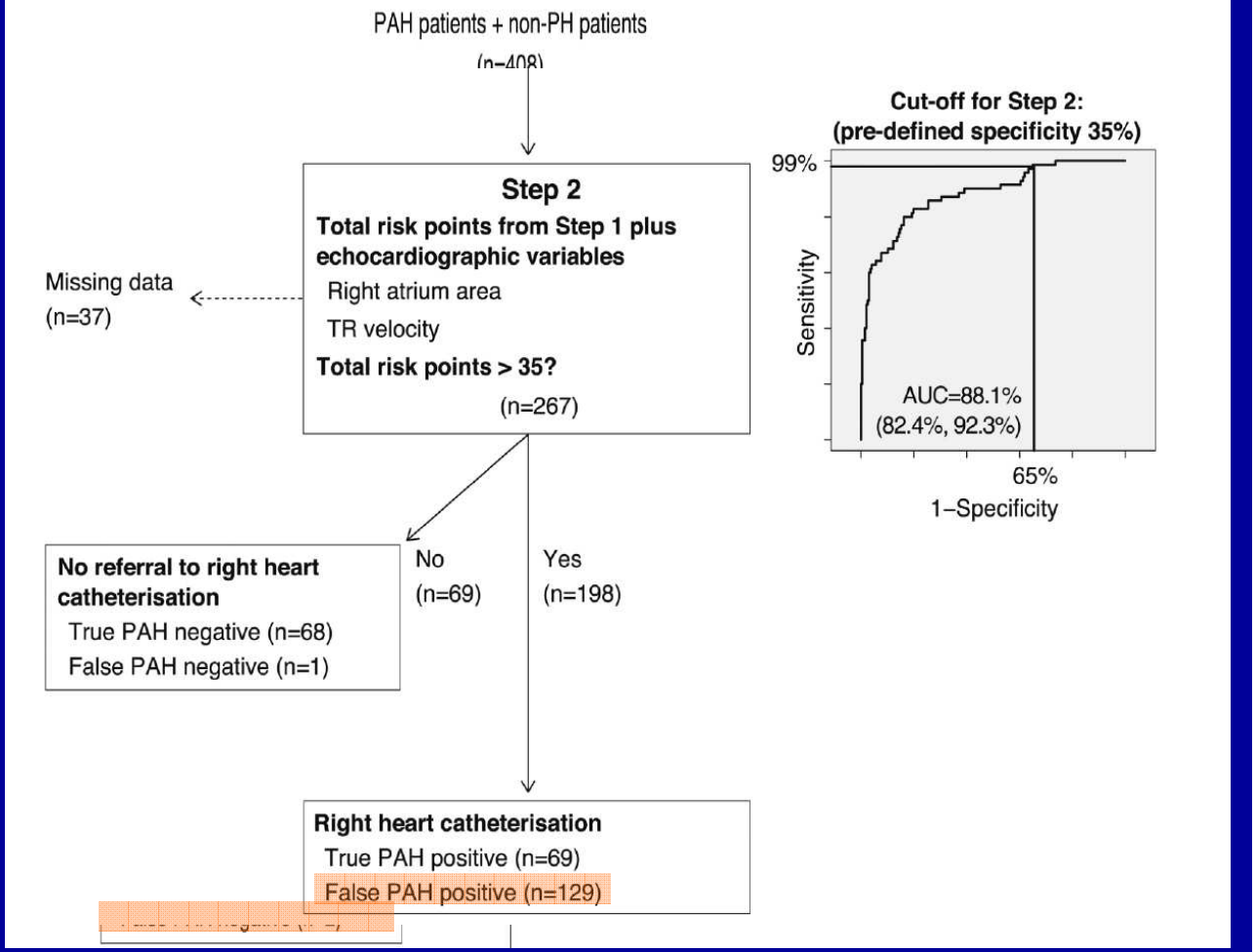

PAH patients + non-PH patients
(n=408)
Step 2
Total risk points from Step 1 plus
echocardiographic variables
Right atrium area
TR velocity
Total risk points > 35?
(n=267)
Missing data
(n=37)
No
(n=69)
Yes
(n=198)
No referral to right heart
catheterisation
True PAH negative (n=68)
False PAH negative (n=1)
Right heart catheterisation
True PAH positive (n=69)
False PAH positive (n=129)
Cut-off for Step 2:
(pre-defined specificity 35%)
99%
Sensitivity
AUC=88.1%
(82.4%, 92.3%)
65%
1−Specificity

Table 1 Patient characteristics

	Non-PH group (N=321)	PAH group (N=87)
Functional capacity		
6-min walk distance (m), n	243	66
Mean (SD)	412.5 (107.2)	389.7 (106.6)
Borg dyspnoea index, n	240	66
Mean (SD)	2.6 (1.8)	3.1 (2.1)
WHO functional class, n	306	87
I/II/III/IV, n (%)	133 (43.5)/123 (40.2)/50 (16.3)/0	16 (18.4)/40 (46.0)/30 (34.5)/1 (1.1)
TR velocity (m/s), n/N (%)		
'No TR' ticked	48/303 (15.8)	6/84 (7.1)
≤2.8	214/303 (70.6)	30/84 (35.7)
>2.8 to ≤3.4	37/303 (12.2)	23/84 (27.4)
>3.4	4/303 (1.3)	25/84 (29.8)

Approach	Overall missed PAH diagnoses, % (false negatives)
Primary analysis	
DETECT algorithm	4

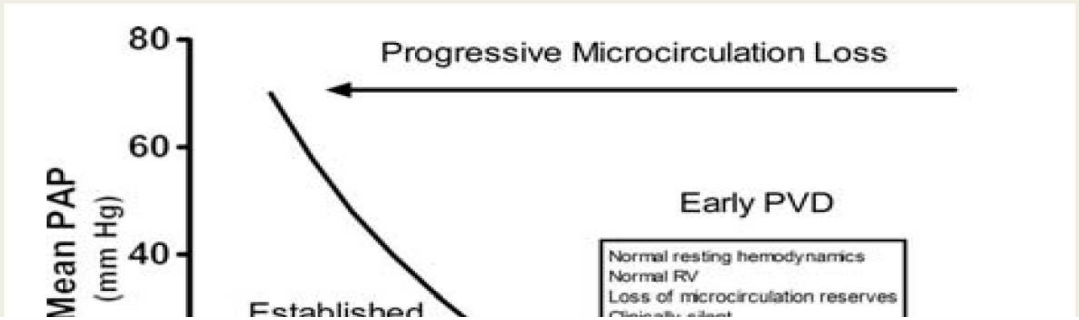
TABLA 7. **Clasificación del estado funcional de la NYHA/WHO de los pacientes con hipertensión pulmonar**

Clase	Descripción
I	Pacientes con hipertensión pulmonar que no presentan limitación de la actividad física normal; la actividad física normal no causa un aumento de la disnea, fatiga, dolor torácico o presíncope
II	Pacientes con hipertensión pulmonar que presentan una limitación leve de la actividad física. No sienten malestar en reposo, pero la actividad física normal provoca el aumento de la disnea, fatiga, dolor torácico o presíncope
III	Pacientes con hipertensión pulmonar que presentan una marcada limitación de la actividad física. No sienten malestar en reposo, pero la mínima actividad física provoca un aumento de la disnea, cansancio, dolor torácico o presíncope
IV	Pacientes con hipertensión pulmonar incapaces de desarrollar cualquier actividad física y que pueden presentar signos de insuficiencia ventricular derecha en reposo. También la disnea y el cansancio pueden estar presentes en reposo y los síntomas aumentan con la mínima actividad física

Early detection of pulmonary vascular disease in pulmonary arterial hypertension: time to move forward

Edmund M.T. Lau[1], Alessandra Manes[2], David S. Celermajer[1]*, and Nazzareno Galiè[2]

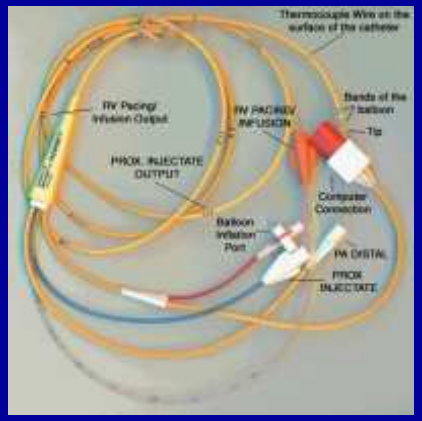
European Heart Journal (2011) **32**, 2489–2498



↑ **CAPACITANCIA**

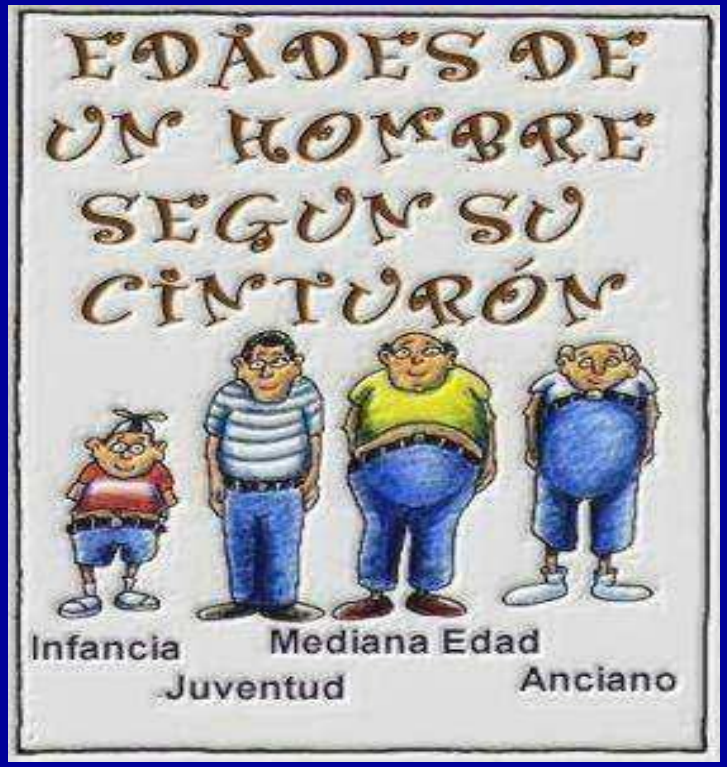
↓ **PRESION**

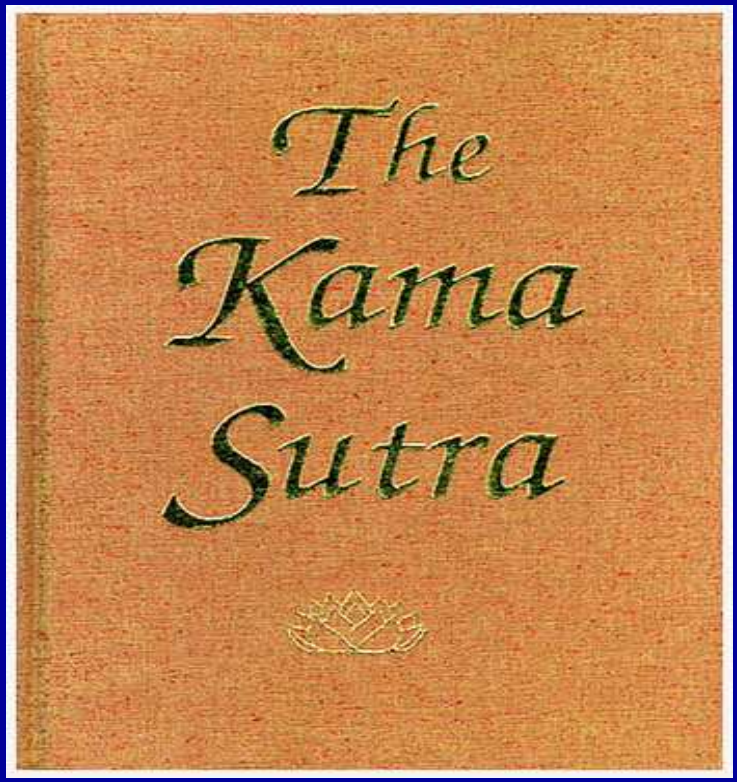
Therefore, it is imperative to reduce the time between the beginning of the pathobiological processes leading to PAH and the initiation of effective medical therapy.

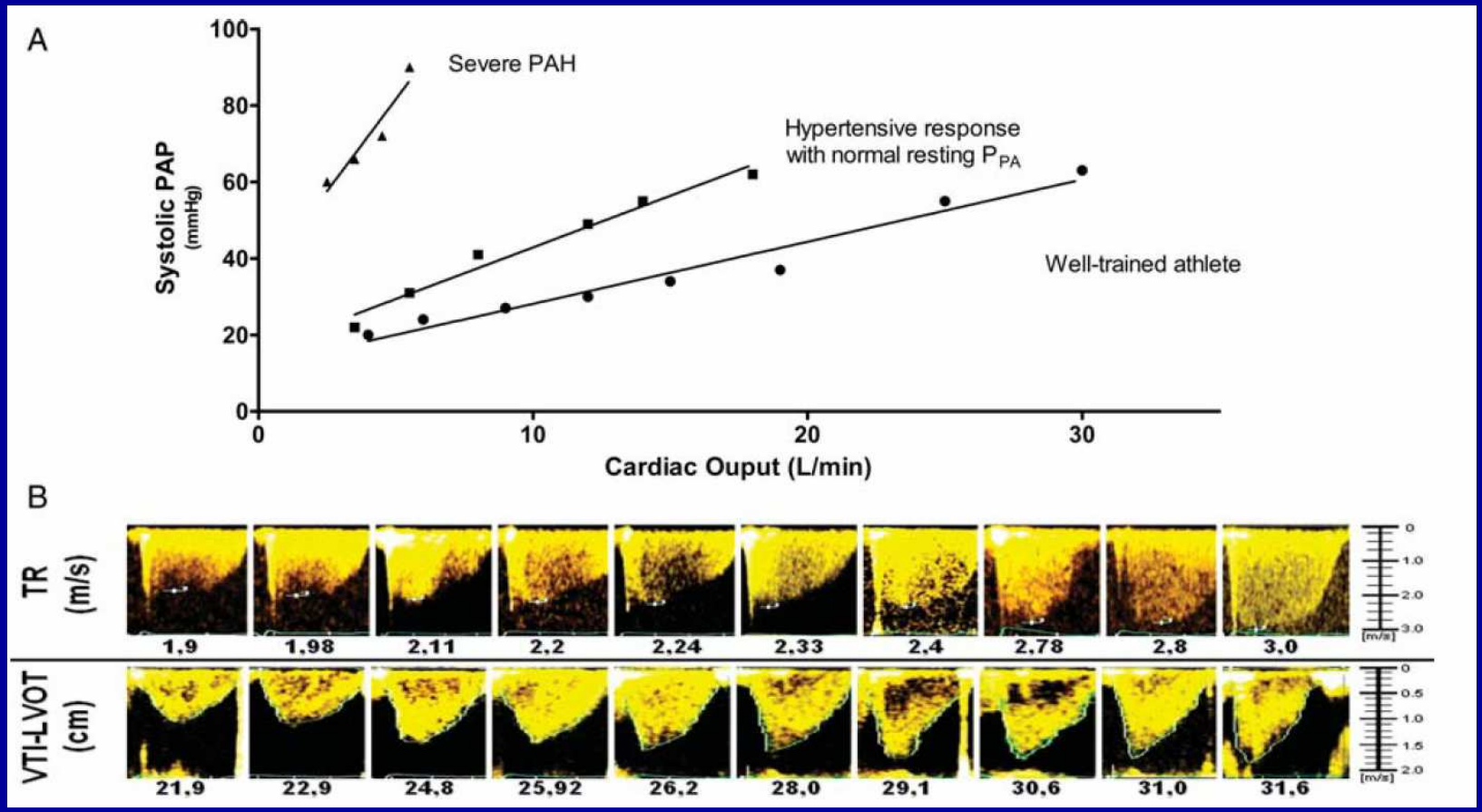


Therefore, using the current haemodynamic definition of PH (resting PAP $\geq$25 mmHg),[14] PVD is necessarily identified at a relatively late stage, potentially reducing the efficacy of any current or future anti-remodelling therapeutic strategies.

La hipertensión pulmonar (HP) se define como la presencia de una presión media en la arteria pulmonar (PAPm) > 25 mmHg en reposo o > 30 mmHg durante el ejercicio[17].


EDADES DE
UN HOMBRE
SEGUN SU
CINTURÓN
Infancia
Juventud
Mediana Edad
Anciano


The
Kama
Sutra


A
Systolic PAP (mmHg)
100
80
60
40
20
0
0
10
20
30
Cardiac Ouput (L/min)
Severe PAH
Hypertensive response with normal resting P_{PA}
Well-trained athlete
B
TR (m/s)
1,9
1,98
2,11
2,2
2,24
2,33
2,4
2,78
2,8
3,0
0
1.0
2.0
3.0
[m/s]
VTI-LVOT (cm)
21,9
22,9
24,8
25,92
26,2
28,0
29,1
30,6
31,0
31,6
0
0.5
1.0
1.5
2.0
[m/s]

Stress-Induced Pulmonary Systolic Hypertension in Patients With Scleroderma

Jose Luis Callejas, MD
Eduardo Moreno, MD
Pilar Martin, MD
Lourdes Lopez-Perez, MD
Norberto Ortego, MD
Hospital Clinico San Cecilio
Granada, Spain

Chest 2007;131;1267

Prevalence of Exercise Pulmonary Arterial Hypertension in Scleroderma

JOSE LUIS CALLEJAS-RUBIO, EDUARDO MORENO-ESCOBAR, PILAR MARTÍN de la FUENTE, LOURDES LÓPEZ PÉREZ, RAQUEL RIOS FERNÁNDEZ, DANIEL SÁNCHEZ-CANO, JOSÉ POMARES MORA, and NORBERTO ORTEGO-CENTENO

J Rheumatol 2008;35:1812–6

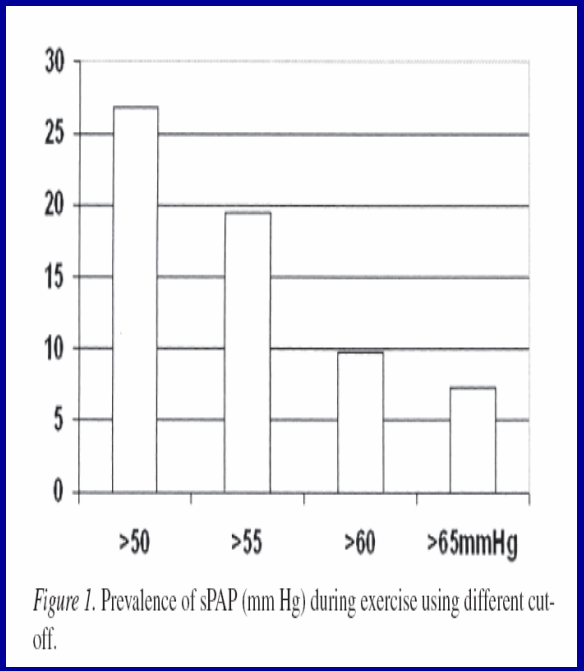


Figure 1. Prevalence of sPAP (mm Hg) during exercise using different cut-off.

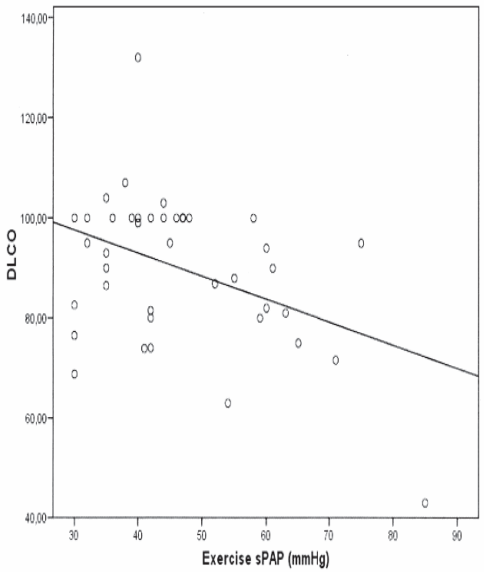


Figure 2. Correlation between sPAP (mm Hg) and DLCO (% predicted); r = -0.4, p = 0.008.

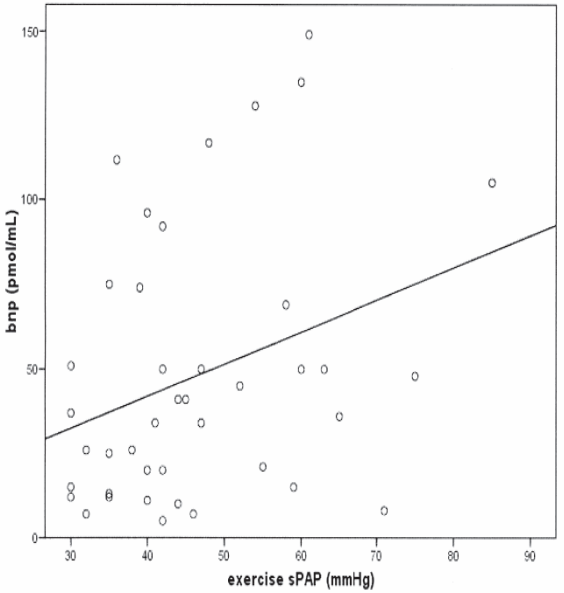


Figure 3. Correlation betwwen sPAP (mm Hg) and BNP (pmol/ml); r = 0.31, p = 0.04.

✓**Podría identificar a pacientes con alto riesgo de desarrollar HAP en reposo**

✓**Podría seleccionar a pacientes para realización de cateterismo cardíaco derecho de reposo y de esfuerzo.**

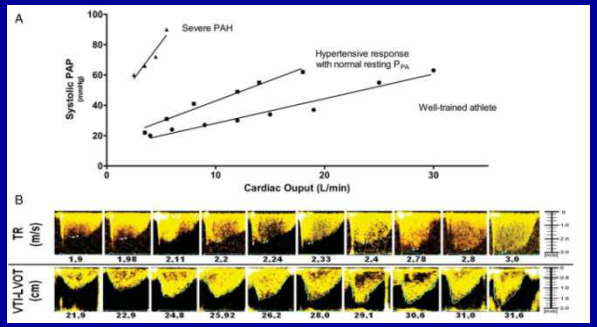
Stress Doppler Echocardiography in Systemic Sclerosis

Evidence for a Role in the Prediction of Pulmonary Hypertension

Veronica Codullo,[1] Roberto Caporali,[1] Giovanna Cuomo,[2] Stefano Ghio,[1] Michele D'Alto,[3] Chiara Fusetti,[1] Elena Borgogno,[1] Carlomaurizio Montecucco,[1] and Gabriele Valentini[2]

ARTHRITIS & RHEUMATISM
Vol. 65, No. 9, September 2013, pp 2403–2411

Conclusion. **An inappropriate response to exercise among patients with SSC is detectable by stress Doppler echocardiography. Independently of other clinical associations, increased Δpulmonary artery systolic pressure heralds PH. Stress Doppler echocardiography may represent an additional screening tool for this severe complication.**



Exercise-Induced Pulmonary Hypertension in Scleroderma Patients: A Common Finding but with Elusive Pathophysiology

Echocardiography 2013;30:378-384

Clinical and echocardiographic correlations of exercise-induced pulmonary hypertension in systemic sclerosis: A multicenter study

Am Heart J. 2013 Feb;165(2):200-7

The elevation of PASP during exercise in most patients of this cohort seems to be related to a reduced pulmonary vascular reserve, and not to an increase in PCWP. Further

Peak exercise PASP is affected by age, interstitial lung disease, and right and left ventricular diastolic dysfunction and, only in 5% of the patients, is associated with an increase in PVR during exercise, suggesting heterogeneity of the mechanisms underlying exercise-induced pulmonary hypertension in SSc.

Inappropriate exercise-induced increase in pulmonary artery pressure in patients with systemic sclerosis

Michele D'Alto,[1] Stefano Ghio,[2] Antonello D'Andrea,[1] Anna Sara Pazzano,[2] Paola Argiento,[1] Rita Camporotondo,[2] Francesca Allocca,[1] Laura Scelsi,[2] Giovanna Cuomo,[3] Roberto Caporali,[4] Lorenzo Cavagna,[4] Gabriele Valentini,[3] Raffaele Calabrò[1]

Heart 2011;**97**:112–117.

Clin Respir J 2013 Jul;7(3):227-36

Exercise during cardiac catheterization distinguishes between pulmonary and left ventricular causes of dyspnea in systemic sclerosis patients

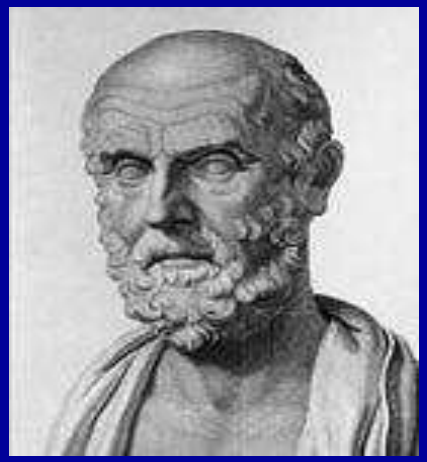
An abnormal ΔmPAP/ΔCO ratio,
an exercise transpulmonary gradient (TPG) ≥ 15,
a PAWP < 20,
a ΔTPG > ΔPAWP and
a pulmonary vascular resistance (PVR) which increased defined exercise induced pulmonary arterial hypertension (EIPAH).

Should patients with systemic sclerosis-related pulmonary arterial hypertension be anticoagulated?

M. Nikpour,[1,2] W. Stevens,[2] S. M. Proudman,[5] R. Buchbinder,[6] D. Prior,[1,3] J. Zochling,[7] T. Williams,[8] E. Gabbay[9] and H. Nandurkar[1,4]

Internal Medicine Journal **43** (2013)

En resumen….



vs

- Síntomas
- DLCo
- Ergo-espirometría
- TACAR
- NT-proBNP
- Ecocardiograma reposo y esfuerzo
- Cateterismo cardiaco dcho e izqdo, de reposo y esfuerzo