XXXIV Congreso Sociedad Andaluza de Medicina Interna (SADEMI)

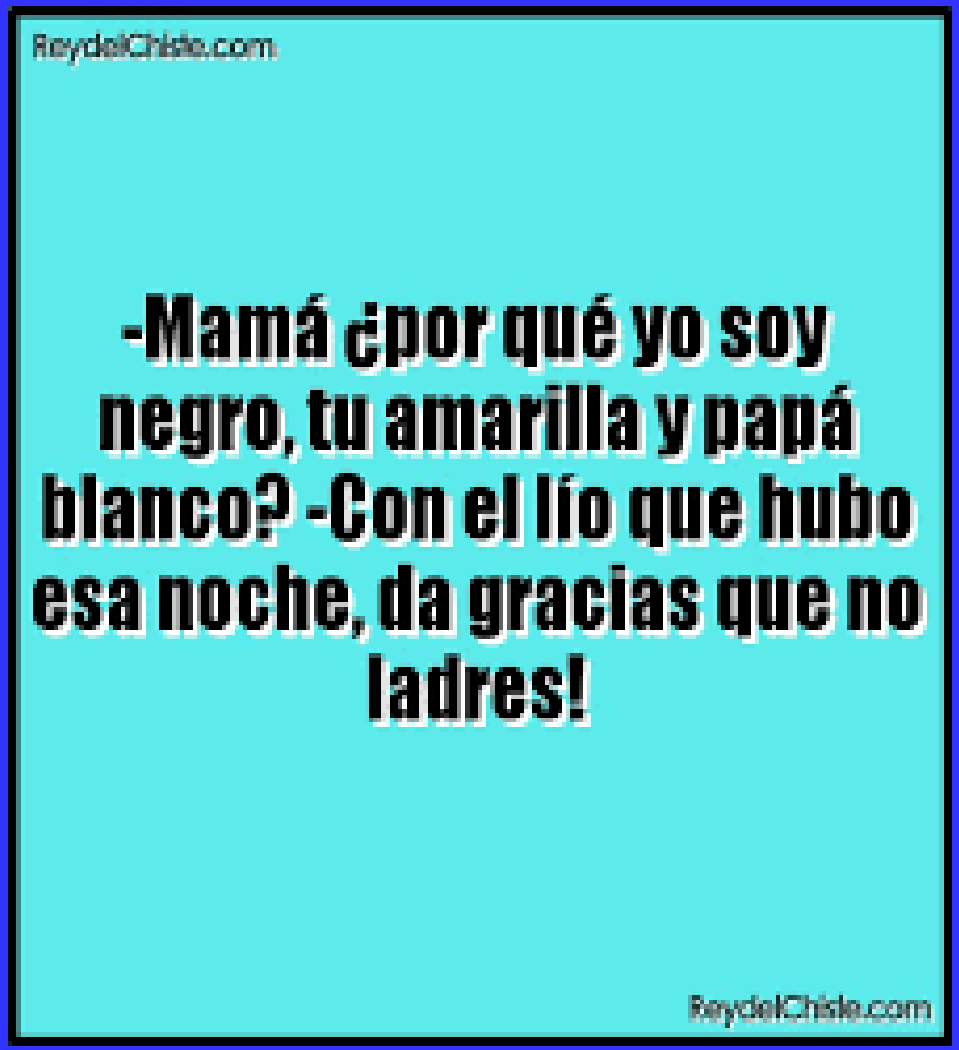
HIPERTENSIÓN PULMONAR PARA INTERNISTAS

UEAS Unidad de Enfermedades Autoinmunes Sistémicas

Jose Luis Callejas
Unidad de Enfermedades Sistémicas
Hopital. Univesitario S.Cecilio

HIPERTENSIÓN PULMONAR

El doctor Santos



HIPERTENSIÓN PULMONAR (HP)

HIPERTENSIÓN ARTERIAL PULMONAR (HAP)

PRESIÓN ARTERIAL PULMONAR SISTÓLICA (PAPs)

PRESION ARTERIAL PUMONAR MEDIA (PAPm)

HP PRE Y POSTCAPILAR

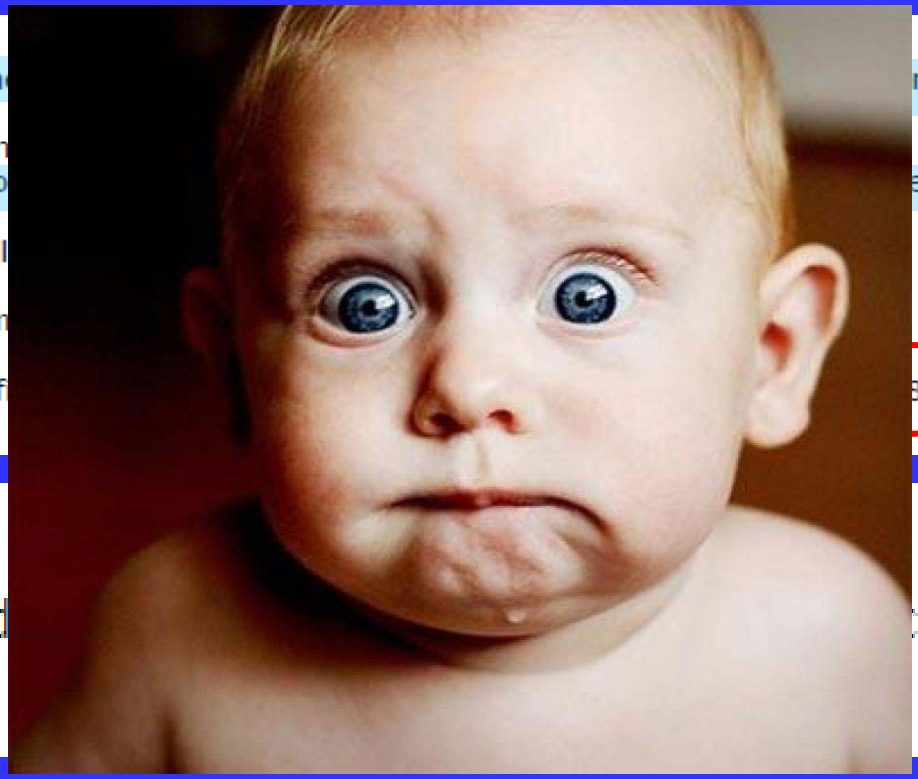
SERVICIO DE CARDIOLOGÍA
ECOCARDIOGRAFÍA
HOSPITAL U SAN CECILIO GRANADA

Fecha: 15/05/2018

Diagnóstico clínico: Ingresada por disnea con derrame pleural izqdo. I. respiratoria agudizada.

Comentario

Ventrículo derecho moderadam moderadamente deprimida (TAPSE 16 mm).
Aurícula derecha moderadamen
Ventrículo izquierdo no dilatado ervada. Aurícula izquierda no dilatada.
Válvulas mitral y aórtica normal
Válvulas derechas normales.
Arteria pulmonar y ramas pulm
Insuficiencia pulmonar ligera
Válvula tricúspide normal, insuf 3 mmHg.
Pericardio normal.



Conclusiones

HTP moderada, Ventrículo d ción sistólica global ligera-moderadamente deprimida


gif-finder.com

ESCENARIOS CLÍNICOS

PACIENTE CON HP
RUEGO ESTUDIO

PACIENTE CIRRÓTICO
PRE-TRASPLANTE HEPÁTICO
DISNEA

HIPERTENSIÓN PORTO-PULMONAR

Review article

Pulmonary arterial hypertension in the setting of scleroderma is different than in the setting of lupus: A review

Isabel S. Bazan[a,1], Kofi A. Mensah[b,1], Anastasiia A
Erica L. Herzog[a], Lenore Buckley[b], Wassim H. Far

HIV-associated pulmonary hy

Harish Jarrett and Christopher Barn

HIPERTENSIÓN PULMONAR
vs
HIPERTENSIÓN ARTERIAL PULMONAR

Clasificación de Hipertensión Pulmonar revisada en Niza 2013

1. Hipertensión Arterial Pulmonar
- 1.1. HAP Idiopática
- 1.2. HAP hereditaria
- 1.2.2 ALK-1,ENG, SMAD9, CAV1, KCNK3
- 1.2.3 Desconocida
- 1.3. Inducida por drogas y toxinas
- 1.4. Asociada con:
  - 1.4.1 *Enfermedad del tejido conectivo*
  - 1.4.2 Infección HIV
  - 1.4.3 *Hipertensión Portal*
  - *1.4.4 Enfermedad Cardiaca Congenita*
  - 1.4.5 Esquistosomiasis

4.2%

1' Enfermedad pulmonar veno-oclusiva y/o Hemangiomatosis pulmonar capilar

1'' Hipertensión Pulmonar persistente del recién nacido (PPHN)

2 Hipertensión Pulmonar debida a enfermedad izquierda
- 2.1. Disfunción sistólica ventricular izquierda
- 2.2. Disfunción diastólica ventricular izquierda
- 2.3. Enfermedad valvular
- 2.4. Obstrucción del tracto de entrada o de salida Congénita / Adquirida del corazón izquierdo

79%

3. Hipertensión Pulmonar debida a Enfermedad Pulmonar y/o Hipoxia
- 3.1. Enfermedad Pulmonar Obstructiva Crónica
- 3.2. Enfermedad del Intersticio Pulmonar
- 3.3. Otras enfermedades pulmonares con patron mixto restrictivo y obstructivo
- 3.4. Trastornos respiratorios del sueño
- 3.5. Trastornos de hipoventilación alveolar
- 3.6. Exposición crónica a grandes alturas
- 3.7. Enfermedades del desarrollo pulmonar

10 %

4. Hipertensión Pulmonar tromboembólica crónica

0.6%

5. Hipertensión pulmonar con mecanismos multifactoriales inciertos
- 5.1. Trastornos hematológicos: anemias hemolíticas crónicas trastornos mieloproliferativos esplenectomía,
- 5.2. Trastornos sistémicos
- 5.3. Trastornos metabólicos, enfermedades de depósito de Glucogeno, enfermedad de Gaucher, enfermedades tiroideas
- 5.4. Otros: obstrucción tumoral, mediastinitis fibrosante, insuficiencia renal crónica, HP segmentaria

6.2%

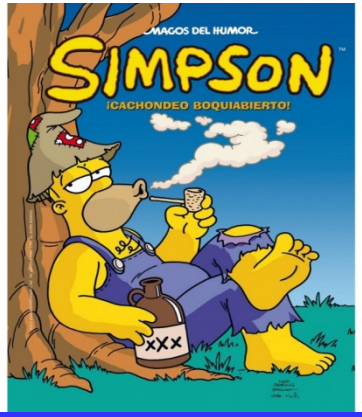
Escribano Subías P, et al. Rev. Esp. Card 2010;63(5):583-96

Pulmonary hypertension: the importance of correctly diagnosing the cause

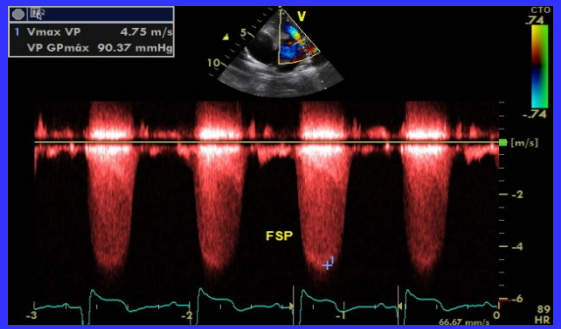
Sanjay Mehta[1,2] and Jean-Luc Vachiéry[3]

Eur Respir Rev 2016; 25: 372–380

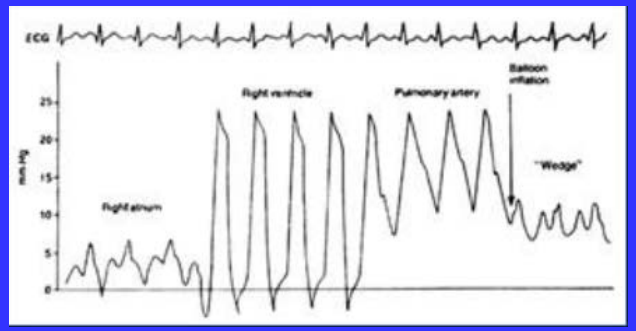
Contemporary PH patients are older and frequently have a multitude of comorbidities that may contribute to or simply coincide with their PH. Identifying the cause of PH in these complicated patients can be challenging but is essential, given that the aetiology of the disease has a significant impact on the management options available.



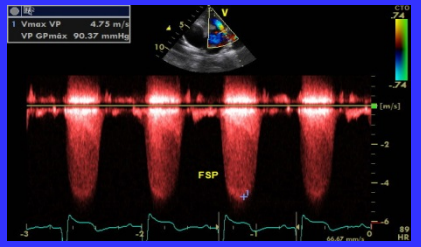
PRESIÓN ARTERIAL PULMONAR SISTÓLICA
VS
PRESIÓN ARTERIAL PULMONAR MEDIA



VS



PAPsistólica (PAPs)



Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'[a]	Echocardiographic probability of pulmonary hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

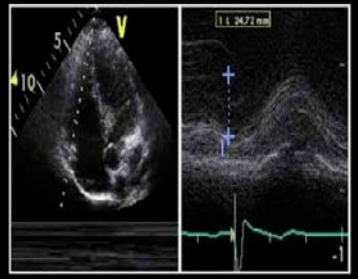
Fórmula de Bernouilli

PAPs=4 v^2 VRT + Pr AD (5-15mmHg)

PAH was defined as a pulmonary arterial systolic pressure (PASP) of >30 mmHg at rest as measured by TTE. PAH was classified as mild (PASP 30-<45 mmHg), moderate (PASP 45-59 mmHg) or severe (PASP >60 mmHg)

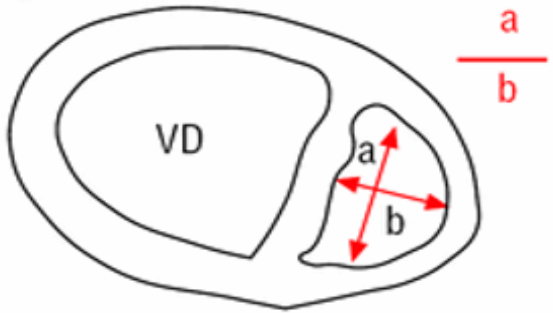
OTROS DATOS ECOCARDIOGRÁFICOS

TAPSE



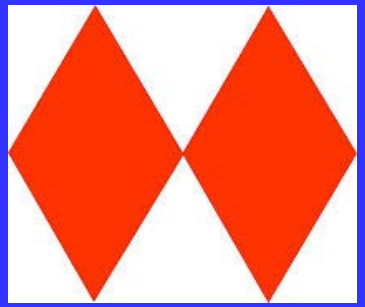
	TAPSE
Normal RV Function	> 16mm
RV Dysfunction	< 16mm

IE×D



PAPmedia: CATETERISMO DERECHO

Definition	Characteristics[a]	Clinical group(s)[b]
PH	PAPm ≥25 mmHg	All
Pre-capillary PH	PAPm ≥25 mmHg PAWP ≤15 mmHg	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	PAPm ≥25 mmHg PAWP >15 mmHg	2. PH due to left heart disease 5. PH with unclear and/or multifactorial mechanisms



NO SE RECOMIENDA

NI DIAGNOSTICAR

NI TRATAR

SIN LA REALIZACIÓN DEL CATETERISMO CARDÍACO DERECHO

HAP ≠ HP
PAPs ≠ PAPm

- Todos los pacientes con HAP tienen HP
- No todos las HP son HAP
- PAPs es un concepto ecocardiográfico
- PAPm es un concepto hemodinámico

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

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**

Hemodynamic Phenotyping of Pulmonary Hypertension in Left Heart Failure

Circ Heart Fail. 2017;10:e004082.

YES

NO

Consider most common causes of PH
(i.e, left heart disease, lung disease)

History, signs, risk factors, ECG, X-ray, PFT incl.
DLCO, consider BGA, HR-CT

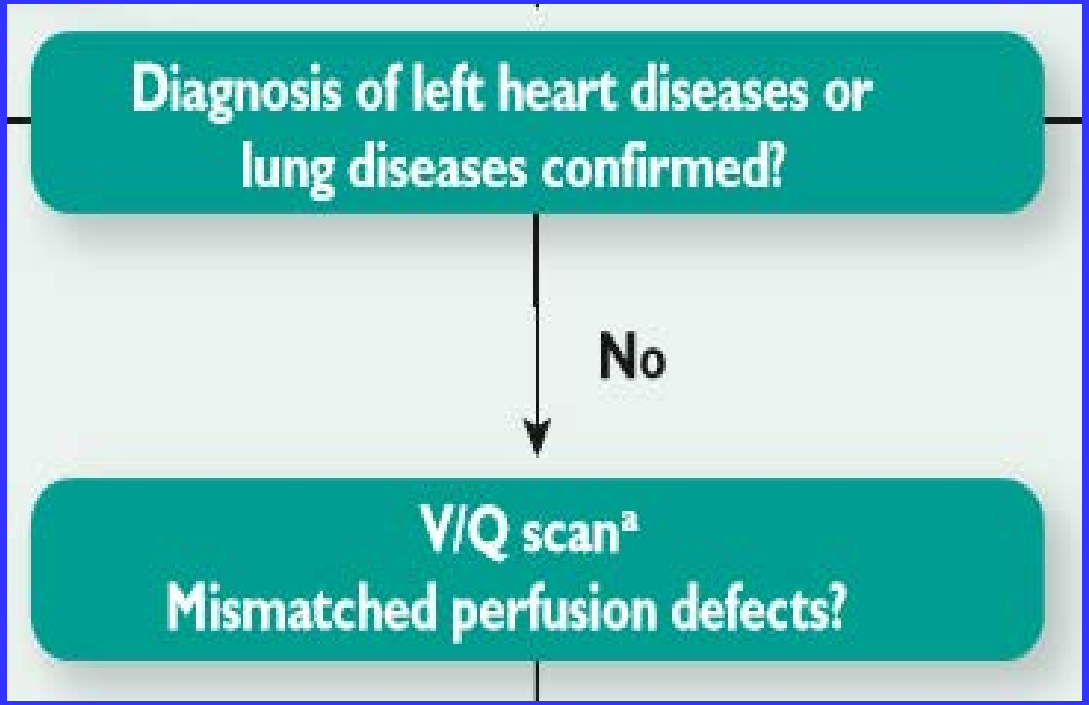
PCH

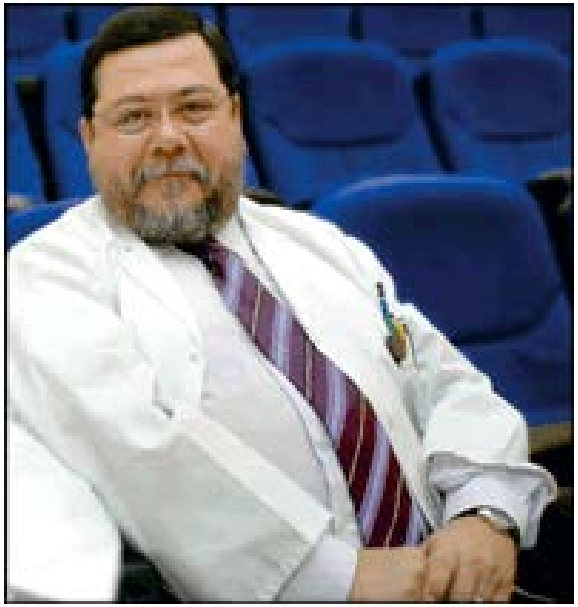
Other (group 5?)

Idiopathic or Heritable PAH

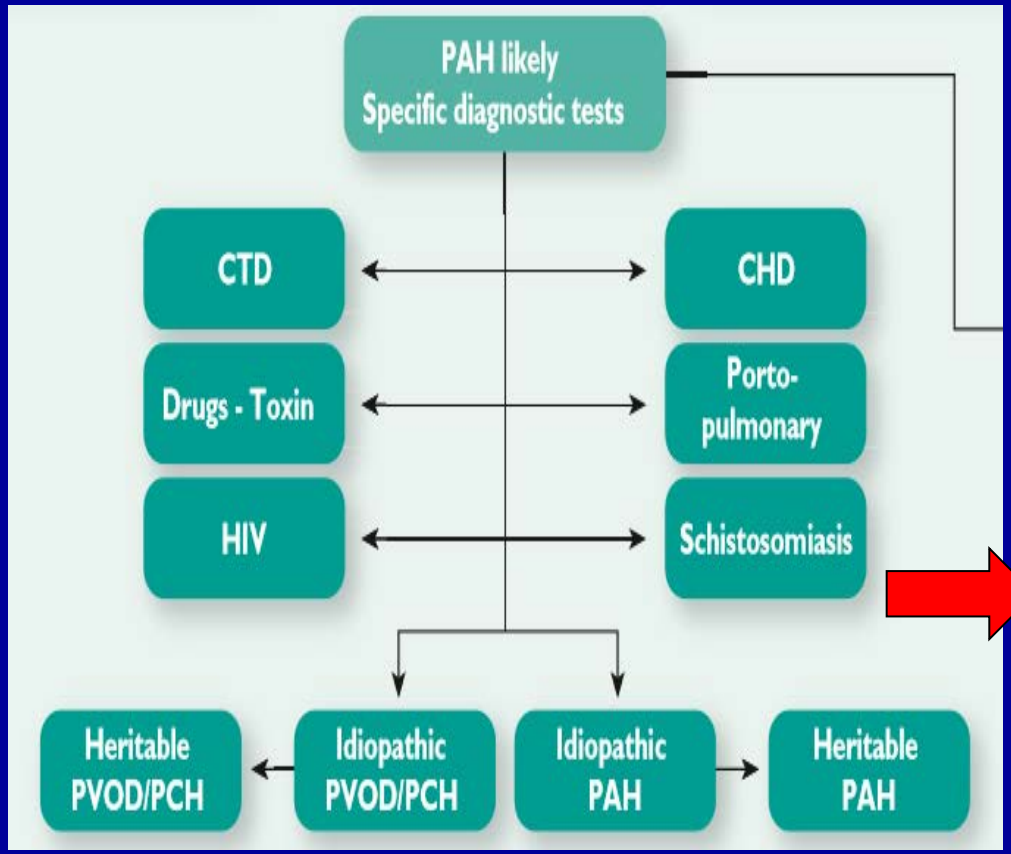
Family history; consider genetic studies
(expert centers only)

DESCARTAR UNA HIPERTENSIÓN PULMONAR TROMBOEMBÓLICA CRÓNICA





El doctor Santos



Recommendations	Class[a]	Level[b]
Echocardiography is recommended as a first-line non-invasive diagnostic investigation in case of suspicion of PH	I	C
Ventilation/perfusion or perfusion lung scan is recommended in patients with unexplained PH to exclude CTEPH	I	C
Contrast CT angiography of the PA is recommended in the workup of patients with CTEPH	I	C
Routine biochemistry, haematology, immunology, HIV testing and thyroid function tests are recommended in all patients with PAH to identify the specific associated condition	I	C
Abdominal ultrasound is recommended for the screening of portal hypertension	I	C
Lung function test with DLCO is recommended in the initial evaluation of patients with PH	I	C

Clinic Rev Allerg Immunol January 2010
DOI 10.

Scle

Jérôme
Paul M

Prevalence and factors associated with left

a

lbir,[13]

);69:218–221

graphy

):410-415

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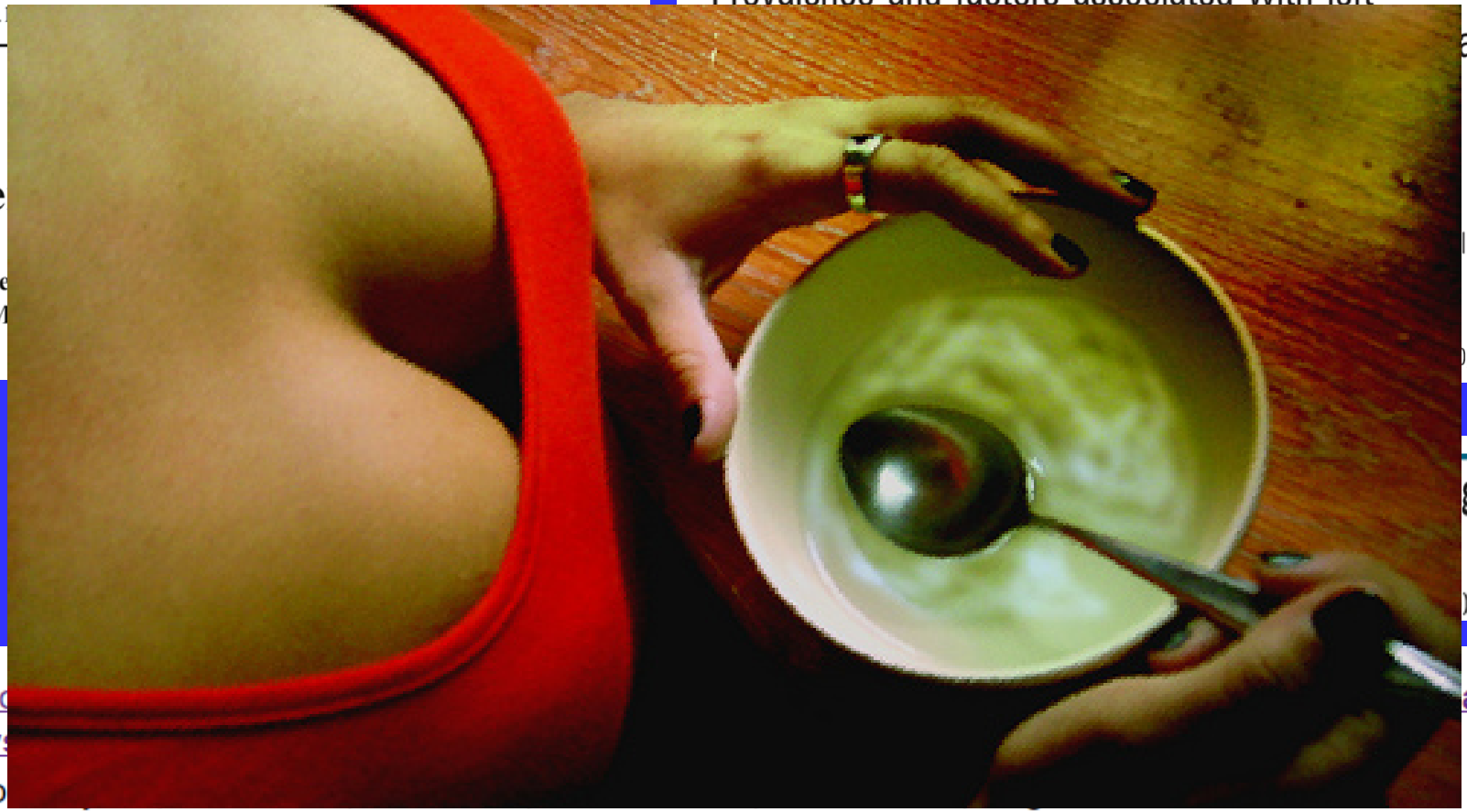

Rheumatology (Oxford). 2017 Dec 1;56(12):2197-2203. doi: 10.1093/rheumatology/kex351.

Post-capillary PH	PAPm ≥25 mmHg PAWP >15 mmHg
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]

TENGO CLARO ES GRUPO 1

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 ml/min/kg (35–65% pred.) VE/VCO_2 slope 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 slope ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal, pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60%

Bases fisiopatológicas

Opciones terapéuticas

FÁRMACOS APROBADOS PARA EL TRATAMIENTO DE LA HAP

ANTAGONISTAS DE LA ENDOTELINA

Bosentan
Ambrisentan
Macitentetan

PRODUCTORES DE NO

IPDE-5

Sildenafilo
Tadalafilo

GC

Riociguat

PROSTAGLANDINAS

Ilopros inh/iv
Treprostinil iv/sc
Epoprsotenol iv

Selexipag oral

Measure/treatment			Class[a]-Level[b]					
			WHO-FC II		WHO-FC III		WHO-FC IV	
Calcium channel blockers			I	C[d]	I	C[d]	-	-
Endothelin receptor antagonists	Ambrisentan		I	A	I	A	IIb	C
	Bosentan		I	A	I	A	IIb	C
	Macitentan[e]		I	B	I	B	IIb	C
Phosphodiesterase type 5 inhibitors	Sildenafil		I	A	I	A	IIb	C
	Tadalafil		I	B	I	B	IIb	C
	Vardenafil[g]		IIb	B	IIb	B	IIb	C
Guanylate cyclase stimulators	Riociguat		I	B	I	B	IIb	C
Prostacyclin analogues	Epoprostenol	Intravenous[e]	-	-	I	A	I	A
	Iloprost	Inhaled	-	-	I	B	IIb	C
		Intravenous[g]	-	-	IIa	C	IIb	C
	Treprostinil	Subcutaneous	-	-	I	B	IIb	C
		Inhaled[g]	-	-	I	B	IIb	C
		Intravenous[f]	-	-	IIa	C	IIb	C
		Oral[g]	-	-	IIb	B	-	-
	Beraprost[g]		-	-	IIb	B	-	-
IP receptor agonists	Selexipag (oral)[g]		I	B	I	B	-	-

Low or intermediate risk (WHO FC II–III)[a]

High risk (WHO FC IV)[a]

Initial monotherapy[b] (Table 19)

Initial oral combination[b] (Table 20)

Initial combination including i.v. PCA[c] (Table 20)

Patient already on treatment

Inadequate clinical response (Table 15)

Consider referral for lung transplantation

Double or triple sequential combination (Table 21)

Inadequate clinical response (Table 15)

Consider listing for lung transplantation[d] (Table 22)

Determinants of prognosis[a] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[b]	Repeated syncope[c]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
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NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
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WINNERS
LEAGUE

Unidad de HIPERTENSIÓN PULMONAR GRANADA

Dr Norberto Ortego Centeno
Dra Raquel Ríos Fernández
Dr Sánchez Cano

UEAS SAN CECILIO

Dr. Eduardo Moreno
Dra.Pilar Martin de la Fuente

CARDIOLOGÍA
SAN CECILIO

Dra Emilia Navascues
Dra Alicia Conde
Dr Ignacio Casado

NEUMOLOGÍA
SAN CECILIO Y
VIRGEN NIEVES

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 2007,14(8)67-74

[Stress echocardiogram in the early diagnosis of systemic sclerosis-associated **pulmonary** hypertension].

Callejas Rubio JL, Parra Rosado P, Ortego Centeno N.

Med Clin (Barc). 2016 Jul 15;147(2):91. doi: 10.1016/j

An MIF Promoter Polymorphism Is Associated with Susceptibility to **Pulmonary** Arterial Hypertension in Diffuse Cutaneous Systemic Sclerosis.

Bossini-Castillo L, Campillo-Davó D, López-Isac E, Carmona FD, Simeon CP, Carreira P, **Callejas-Rubio JL**, Castellví I, Fernández-Nebro A, Rodríguez-Rodríguez L, Rubio-Rivas M, García-Hernández FJ, Madroñero AB, Beretta L, Santaniello A, Lunardi C, Airó P, Hoffmann-Vold AM, Kreuter A, Riemekasten G, Witte T, Hunzelmann N, Vonk MC, Voskuyl AE, de Vries-Bouwstra J, Shiels P, Herrick A, Worthington J, Radstake TRDJ, Martin J; Spanish Scleroderma Group.

J Rheumatol. 2017 Oct;44(10):1453-1457. doi: 10.3899/jrheum.161369. Epub 2017 Jul 1.

[Echocardiography and scintigraphy in chronic thromboembolic **pulmonary** hypertension].

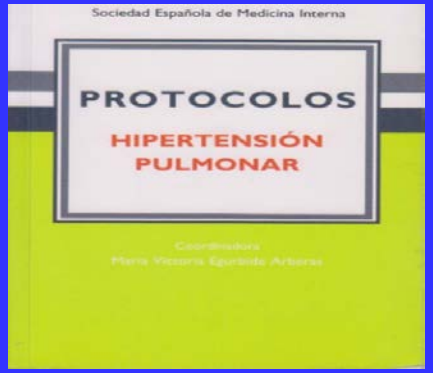
Callejas Rubio JL, Conde Valero A, Moreno Escobar E, Ortego Centeno N.

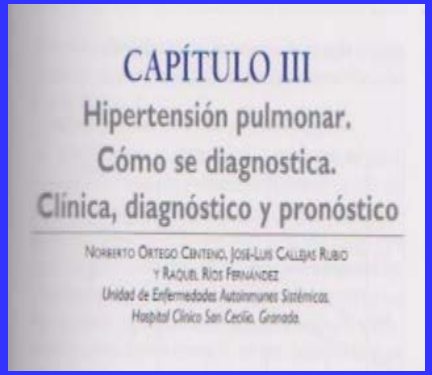
Med Clin (Barc). 2013 Apr 20;140(8):380. doi: 10.1016/j.medcli.2012.09.040. Epub 2013 Jan 17. Spanish. No abstract

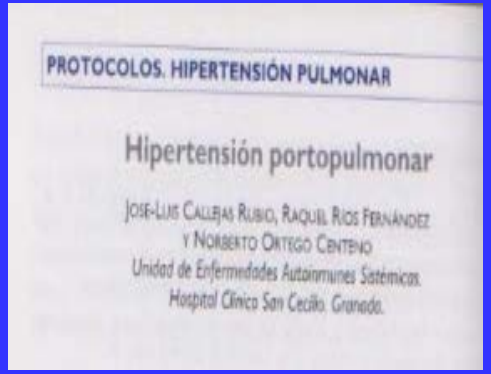
[**Pulmonary** arterial hypertension or **pulmonary** hypertension in patients with human immunodeficiency virus infection?].

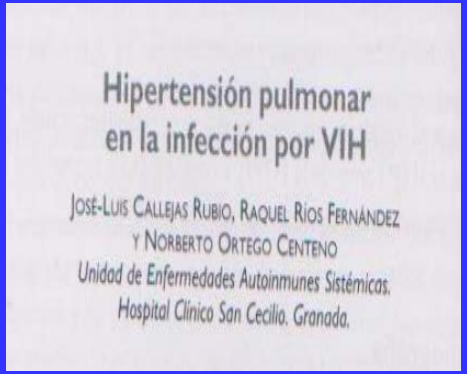
Callejas Rubio JL, Moreno Escobar E, Navascués E, Ortego Centeno N.

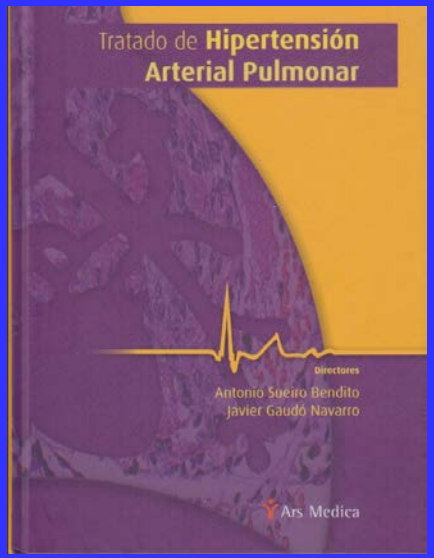
Med Clin (Barc). 2015 Feb 2;144(3):137. doi: 10.1016/j.medcli.2014.03.032. Epub 2014 Jul 26. Spanish. No abstract


Sociedad Española de Medicina Interna
PROTOCOLOS
HIPERTENSIÓN PULMONAR


CAPÍTULO III
Hipertensión pulmonar.
Cómo se diagnostica.
Clínica, diagnóstico y pronóstico
Norberto Ortego Centeno, José-Luis Callejas Rubio
y Raquel Ríos Fernández
Unidad de Enfermedades Autoinmunes Sistémicas.
Hospital Clínico San Cecilio. Granada.


PROTOCOLOS. HIPERTENSIÓN PULMONAR
Hipertensión portopulmonar
José-Luis Callejas Rubio, Raquel Ríos Fernández
y Norberto Ortego Centeno
Unidad de Enfermedades Autoinmunes Sistémicas.
Hospital Clínico San Cecilio. Granada.


Hipertensión pulmonar
en la infección por VIH
José-Luis Callejas Rubio, Raquel Ríos Fernández
y Norberto Ortego Centeno
Unidad de Enfermedades Autoinmunes Sistémicas.
Hospital Clínico San Cecilio. Granada.


Tratado de Hipertensión
Arterial Pulmonar
Directores
Antonio Sueiro Bendito
Javier Gaudó Navarro
Ars Medica


Capítulo 23
Nuevos métodos diagnósticos
de la hipertensión arterial pulmonar
José Luis Callejas Rubio, Eduardo Moreno Escobar y Norberto Ortego Centeno


Introcan Safety