Hipertensión Arterial Pulmonar
De los síntomas al tratamiento

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21-11- 2023

La hipertensión pulmonar (HP) es una enfermedad crónica, multifactorial, de carácter clínico progresivo que se asocia con una con elevada morbimortalidad.

Es una entidad heterogénea, que involucra desde la hipertensión arterial pulmonar (HAP), a entidades más frecuentes como son las asociadas a cardiopatía izquierda y enfermedades pulmonares crónicas

Galie N, Humbert M. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension.
Simmoneau G. Eur Respir J 2019; 53: 1801913

Unidad Multidisciplinaria de Hipertensión Pulmonar

Enfoque Diagnóstico y Terapéutico

Unidad Multidisciplinaria

Reumatología

Medicina Interna

Neumonología

Cardiología

Hemodinamia

C. Congénitas

Imagenes

Unidad Coronaria

Infectología

PPM > 20 mm Hg

Pruebas Funcionales

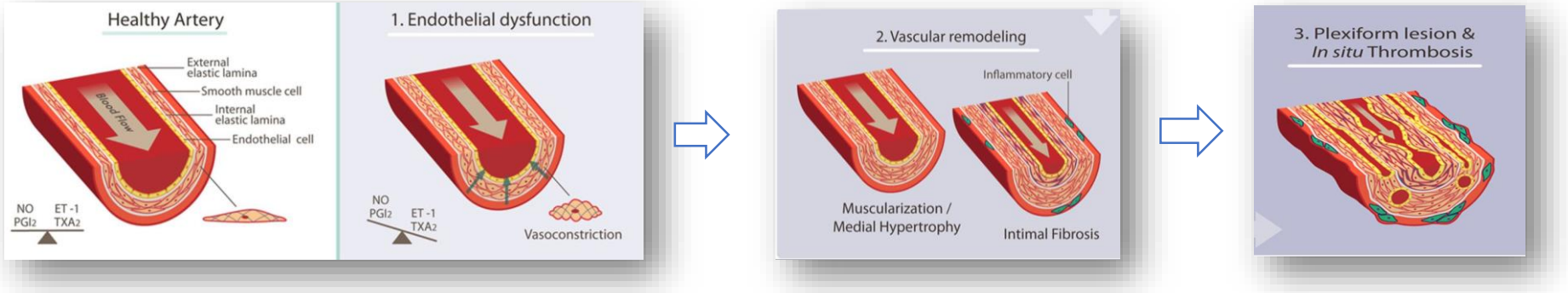
Anestesia

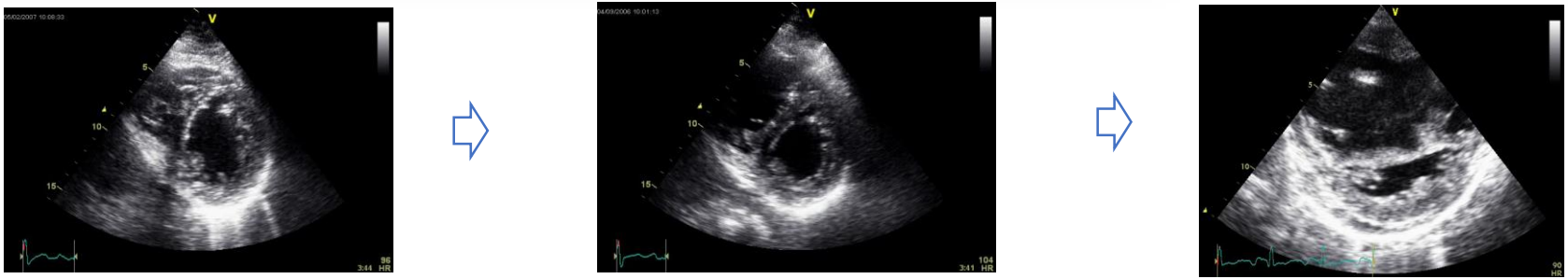
Hematología

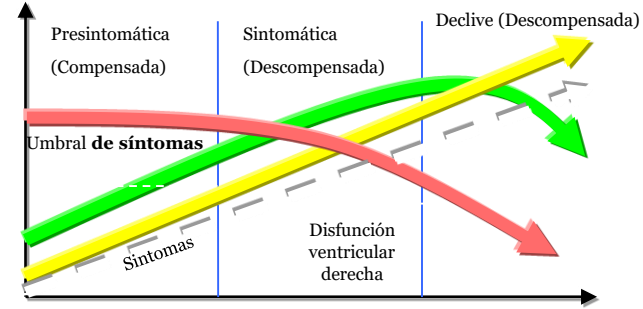
Obstetricia

Psicología

Rehabilitación

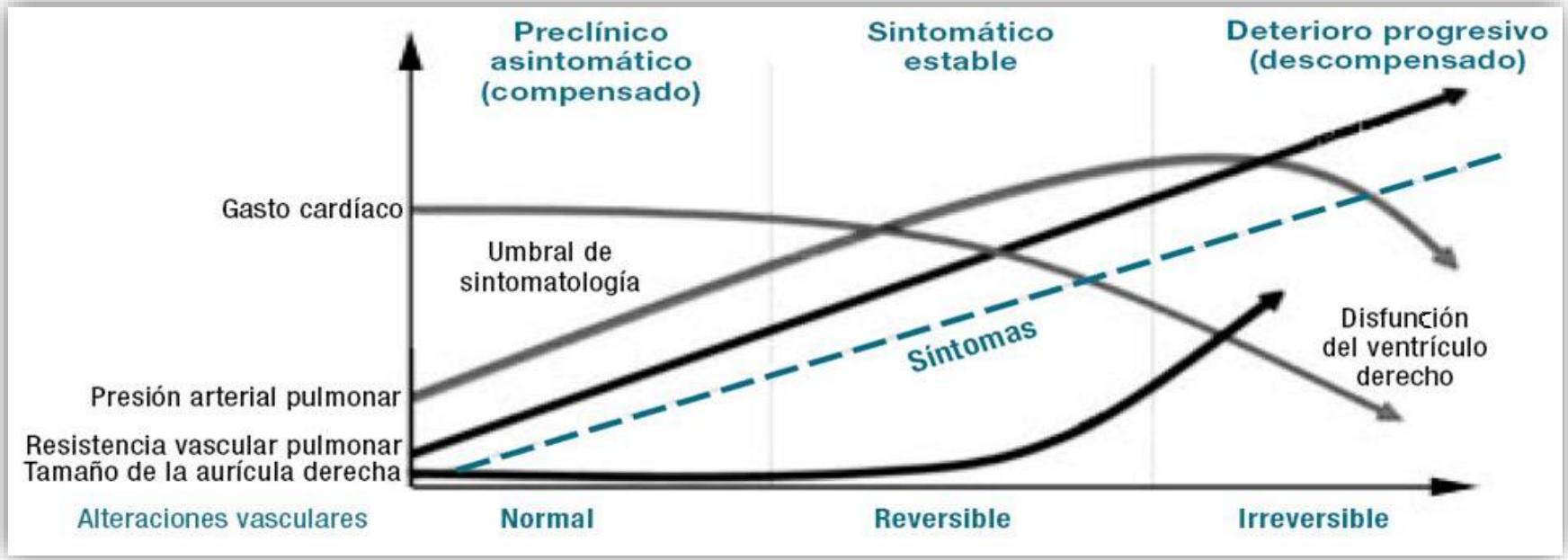

Healthy Artery
External elastic lamina
Smooth muscle cell
Internal elastic lamina
Endothelial cell
Blood Flow
NO
PGI2
ET -1
TXA2
1. Endothelial dysfunction
NO
PGI2
ET -1
TXA2
Vasoconstriction
2. Vascular remodeling
Inflammatory cell
Muscularization / Medial Hypertrophy
Intimal Fibrosis
3. Plexiform lesion & In situ Thrombosis




Presintomática
(Compensada)
Sintomática
(Descompensada)
Declive (Descompensada)
Umbral de síntomas
Síntomas
Disfunción ventricular derecha

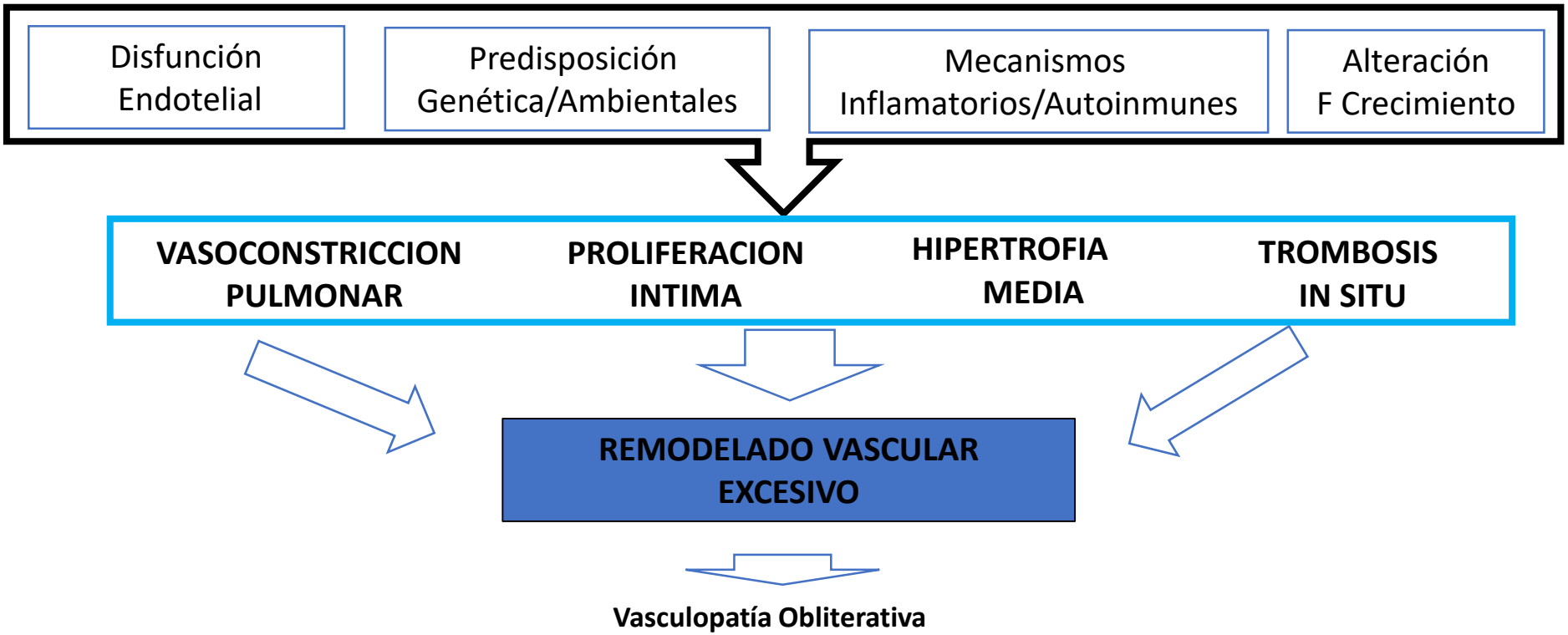
VD: Principal predictor de mortalidad

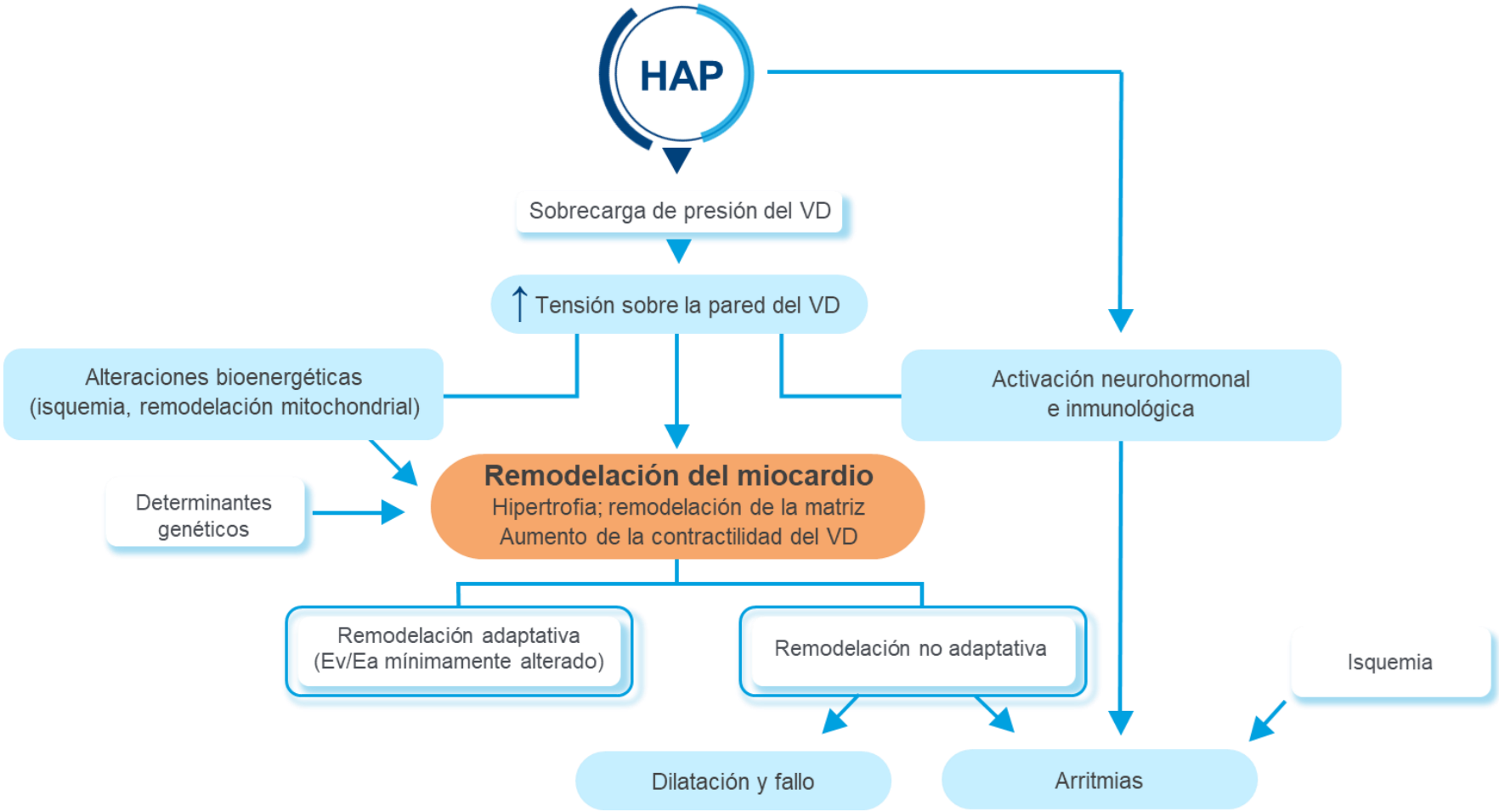
El estado clínico y el pronóstico depende principalmente de la capacidad del VD para adaptarse al aumento de la poscarga



Bortman G. Revista de Insuficiencia Cardiaca , vol 4, N° 1, 2009

Mecanismos Fisiopatológicos HAP: Grupo1





1. Vonk-Noordegraaf A, *et al*. Right heart adaptation to pulmonary arterial hypertension: physiology and pathobiology. J Am Coll Cardiol. 2013 Dec 24;62(25 Suppl):D22-33. doi: 10.1016/j.jacc.2013

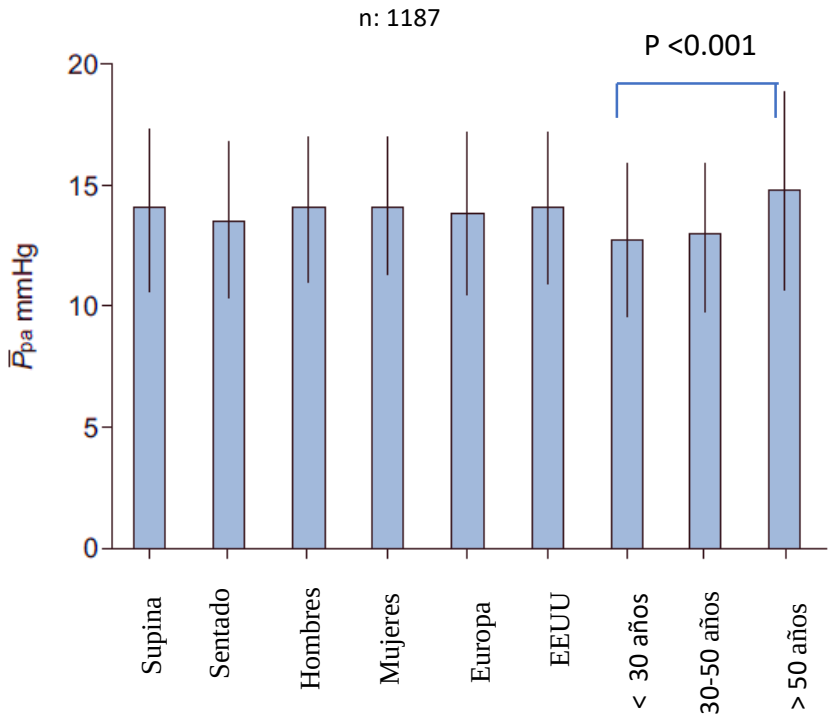
Definición NIZA 2018

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 ERJ Express. Published on December 13, 2018

PAPm en sujetos sanos en reposo

PmAP mmhg	14 ± 3.3
PSAP/PDAP mmhg	20.8 ± 4.4/ 8.8 ± 3
PCP mmhg	8 ± 2.9
FC min¹	76 ± 14
GC l.min¹	7.3 ± 2.3
IC l.min¹. m²	4.1 ± 1.3
RVP din.s.cm	74 ± 30



G. Kovacs et al. Eur Respir J 2009; 34: 888–894

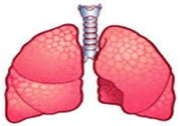
Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
IpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
CpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise >3 mmHg/L/min

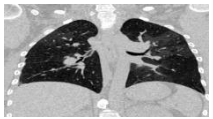
Humbert M et al. European Heart Journal (2022) 00, 1–114

Clasificación Clínica de la HP

HP

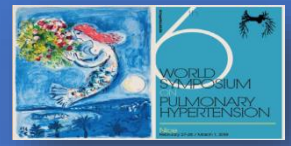
Grupo 1	Grupo 2	Grupo 3	Grupo 4	Grupo 5
HAP	Cardiopatía Izquierda	Enf. Pulmonares y/o hipoxemia	Obstrucción A pulmonar	Mec multifactoriales no aclarados





Simmoneau G. Eur Respir J 2019; 53: 1801913

Prevalencia según Grupo Clínico



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
- 1.7 Persistent PH of the newborn syndrome

< 5%

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

80%

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

10%

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

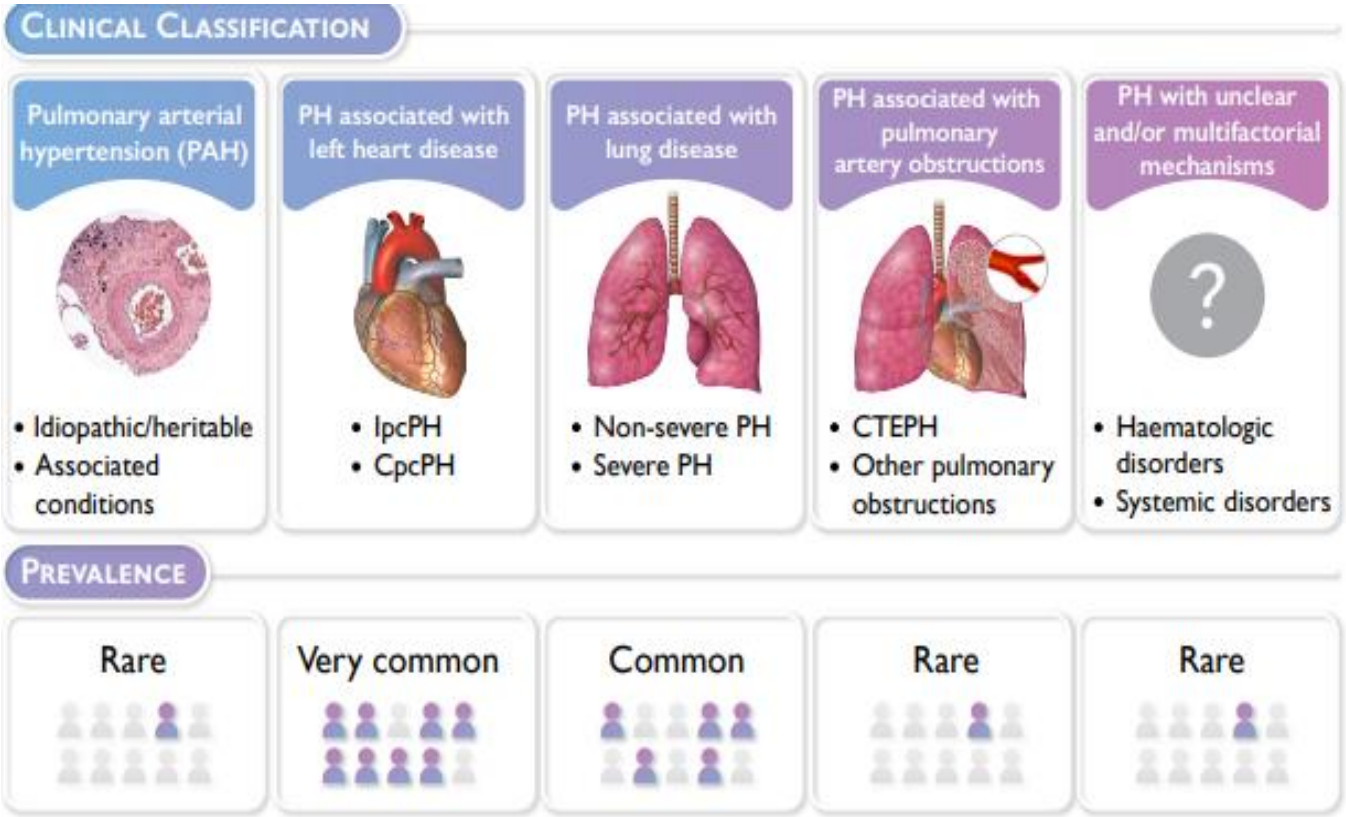
< 5%

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

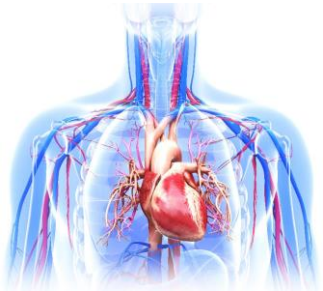
5%

Simmoneau G. Eur Respir J 2019; 53: 1801913

Clasificación Clínica de la HP



Humbert M et al. European Heart Journal (2022) 00, 1–114



Clasificación Clínica

ESC European Society of Cardiology

GROUP 1 Pulmonary arterial hypertension (PAH)
- 1.1 Idiopathic
  - 1.1.1 Non-responders at vasoreactivity testing
  - 1.1.2 Acute responders at vasoreactivity testing
- 1.2 Heritable[a]
- 1.3 Associated with drugs and toxins[a]
- 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 disease
  - 1.4.5 Schistosomiasis
- 1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
- 1.6 Persistent PH of the newborn

GROUP 2 PH associated with left heart disease
- 2.1 Heart failure:
  - 2.1.1 with preserved ejection fraction
  - 2.1.2 with reduced or mildly reduced ejection fraction[b]
- 2.2 Valvular heart disease
- 2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH

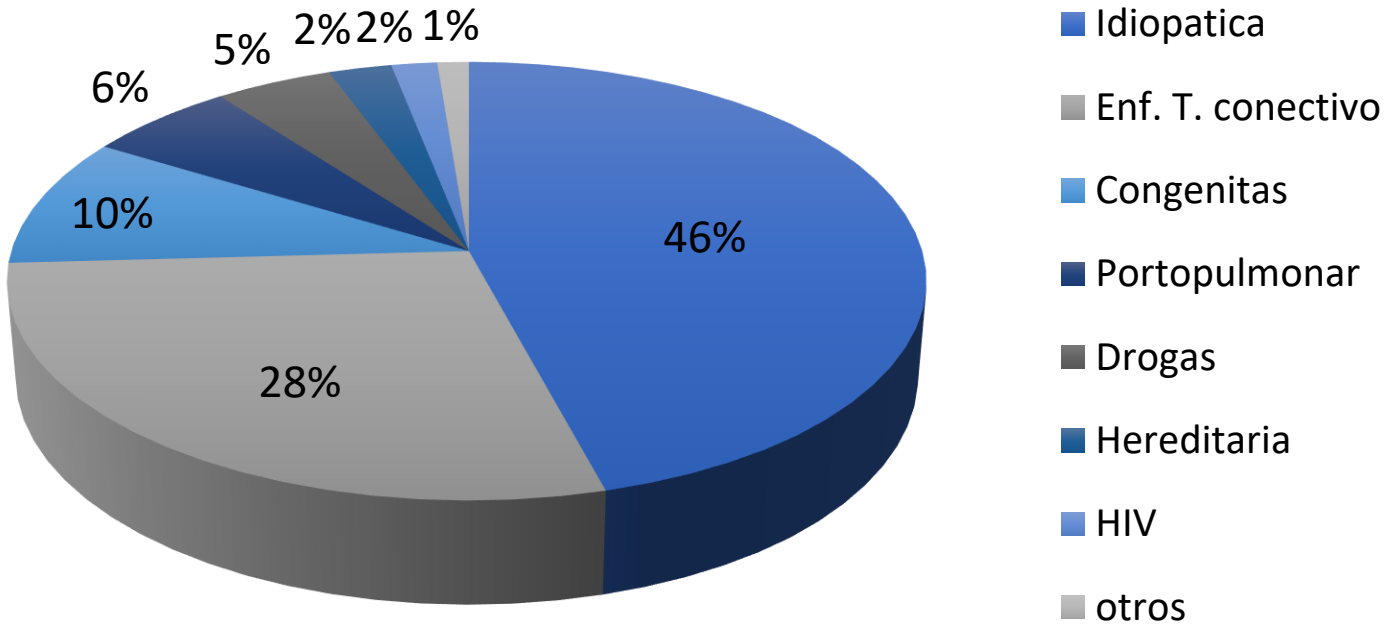
GROUP 3 PH associated with lung diseases and/or hypoxia
- 3.1 Obstructive lung disease or emphysema
- 3.2 Restrictive lung disease
- 3.3 Lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoventilation syndromes
- 3.5 Hypoxia without lung disease (e.g. high altitude)
- 3.6 Developmental lung disorders

GROUP 4 PH associated with pulmonary artery obstructions
- 4.1 Chronic thrombo-embolic PH
- 4.2 Other pulmonary artery obstructions[c]

GROUP 5 PH with unclear and/or multifactorial mechanisms
- 5.1 Haematological disorders[d]
- 5.2 Systemic disorders[e]
- 5.3 Metabolic disorders[f]
- 5.4 Chronic renal failure with or without haemodialysis
- 5.5 Pulmonary tumour thrombotic microangiopathy
- 5.6 Fibrosing mediastinitis

Humbert M et al. European Heart Journal (2022) 00, 1–114

Etiología de HAP



50% HAPI, HAPH y HAP asociada a fármacos.
La HP en Esclerodermia ocupa el 60 a 70% de todas las ETC
LES (2.8-4.2%)

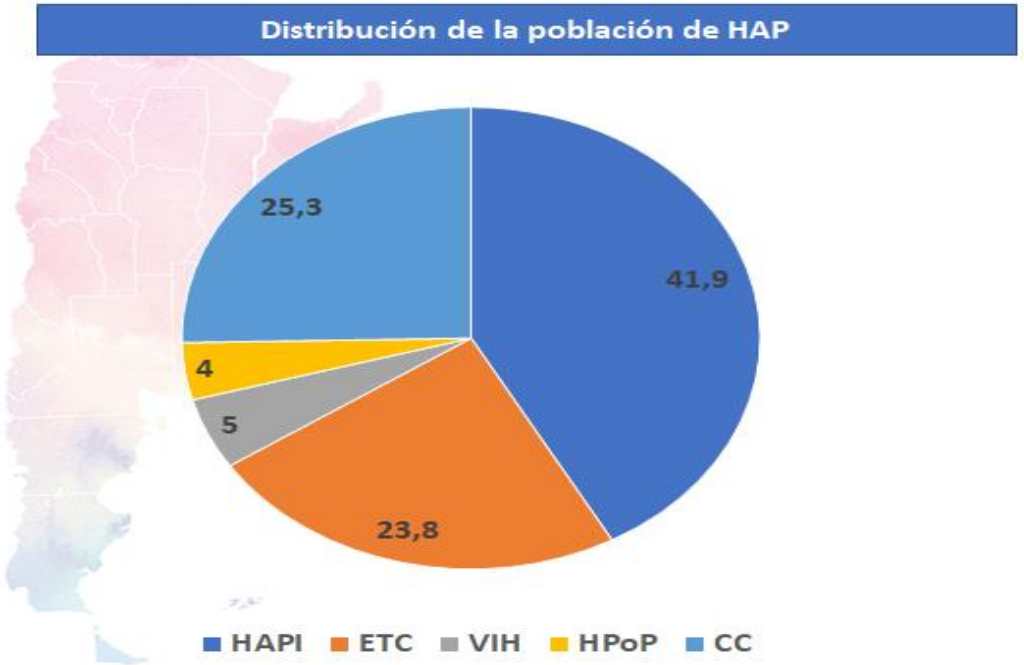
Benza et al CHEST 2012; 142(2):448–456

RECOPILAR

Resultados

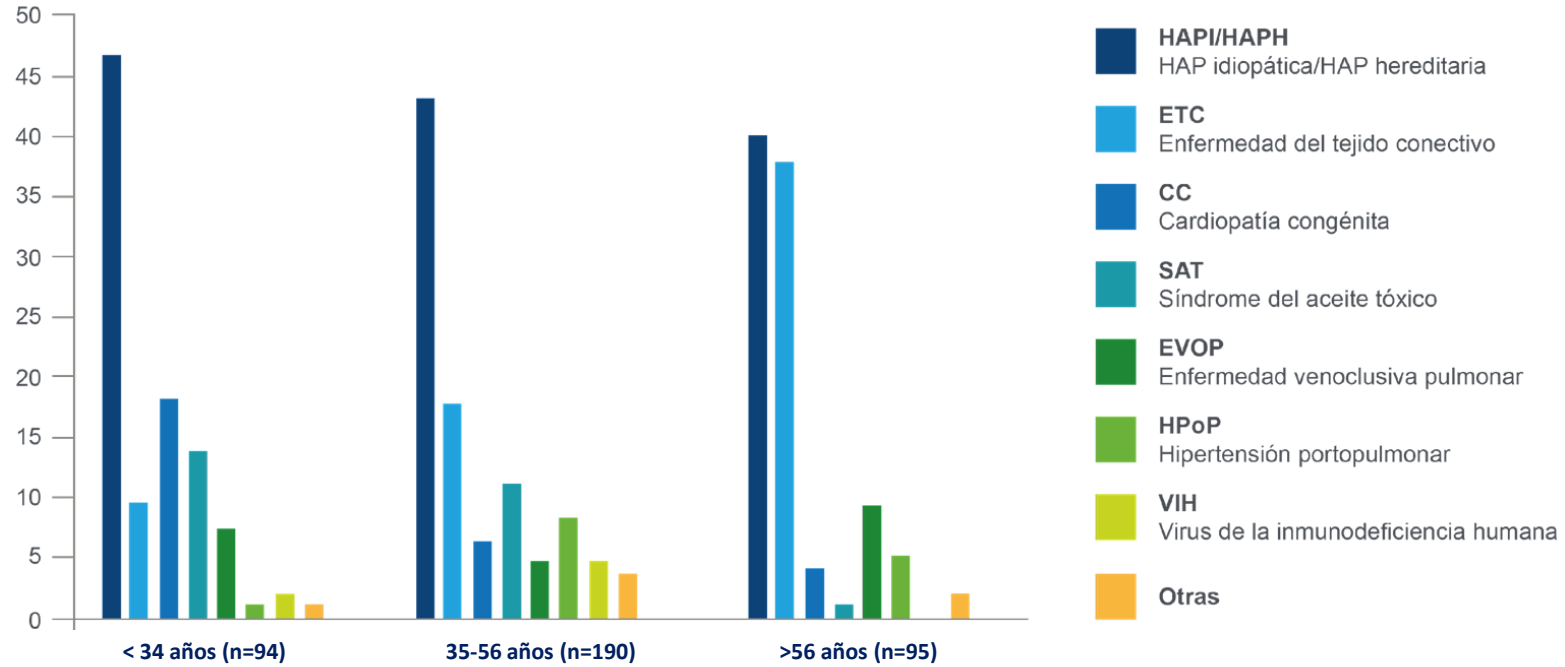
627 patients con PH.
Edad media 50.8±18.7 años,
434 (69.2%) mujeres.

Grupo 1: 399 p
Grupo 2: 100
Group 3: 52
Group 4: 61
Group 5: 15



GRUPO 1: HIPERTENSIÓN ARTERIAL PULMONAR (HAP)

Distribución por etiologías[1]



Adaptado de Figura 2 de Quezada-Loaiza C, *et al*. 2017. Dsitribución de los pacientes por percentil de edad., disponible en el anexo

1. Quezada Loaiza C, *et al*. Trends in Pulmonary Hypertension Over a Period of 30 Years: Experience From a Single Referral Centre. Rev Esp Cardiol. 2017 Nov;70(11):915-923. doi: 10.1016/j.rec.2016.12.044

Epidemiología de la HAP

Enfermedad Rara

Incidencia : 5,8 casos cada millón habitantes por año
(1,5 y 32 (ppm)/año)

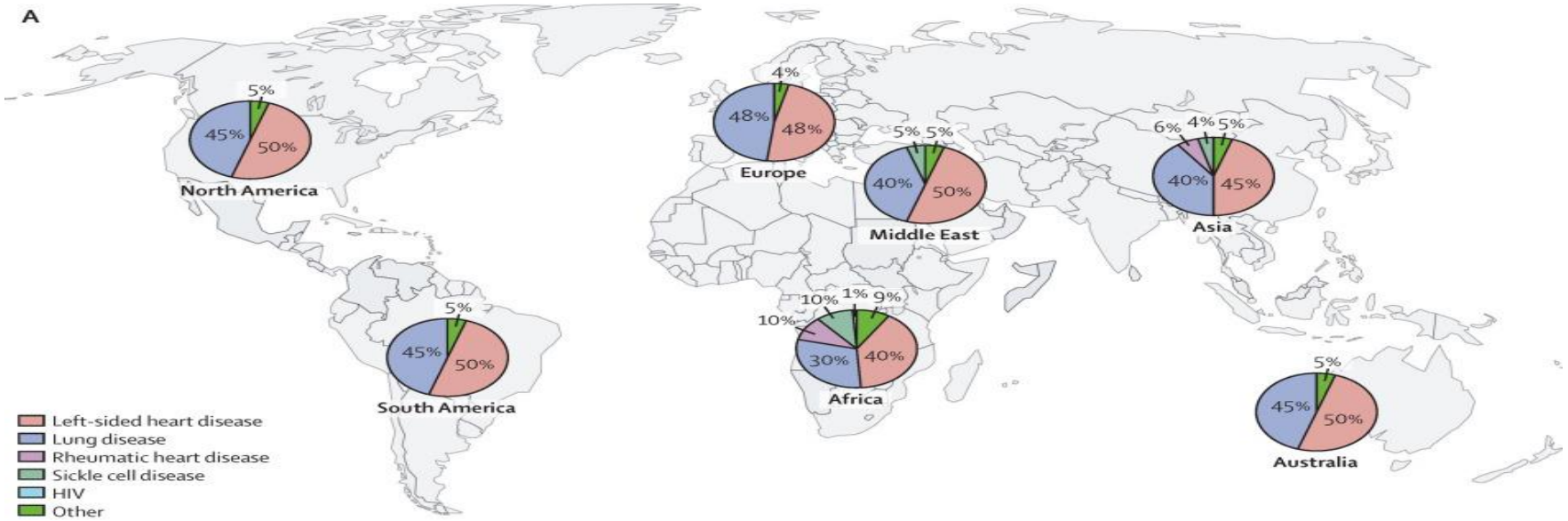
Prevalencia : 15 -26 casos por millón habitantes (47,6- 54,7)

Predomina en el sexo femenino : 2:1 3:1 4:1 (55-81%)

Edad: 43 - 67 años

Valverde et al. BMC Pulmonary Medicine (2018) 18:106
Leber. Pulmonary Circulation (2021):;11(1):2045894020977300

Prevalencia de HP: Etiología según región



Hoeper.Lancet Respir Medic. 2016;4:306-322

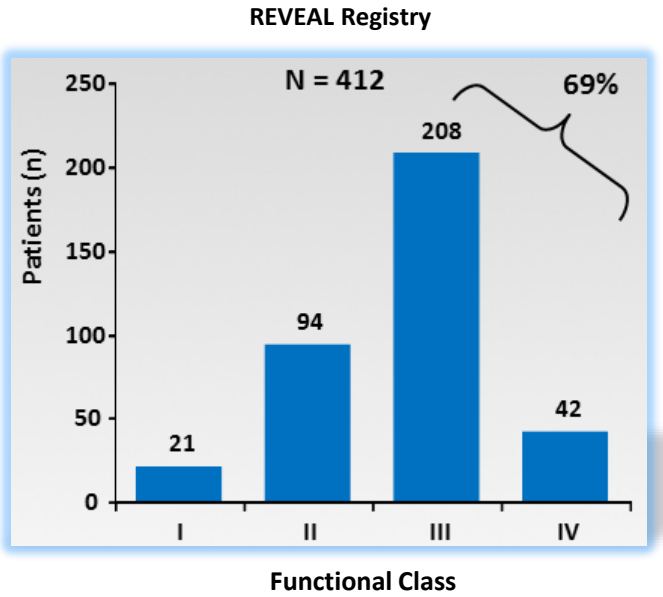
Diagnóstico Desafiante

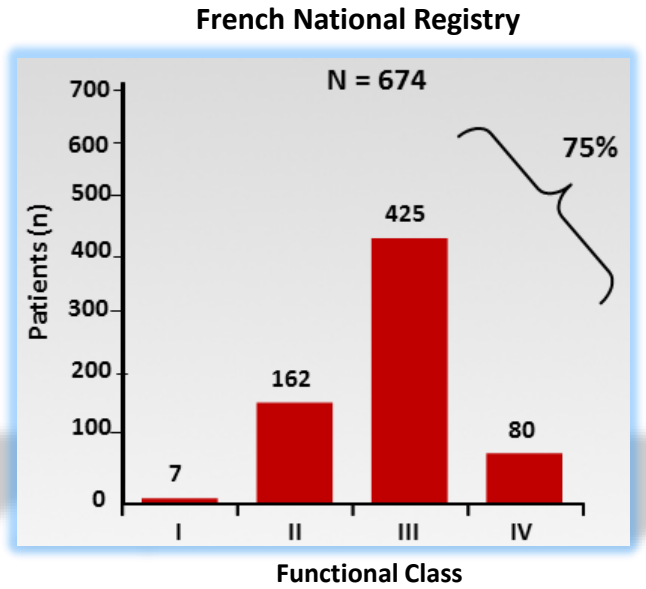
- Los síntomas inespecíficos de la HP demoran el diagnóstico de la patología
- Se suelen pensar diagnósticos alternativos más comunes (asma, enfermedad cardiovascular)
- Mayores probabilidades de demora:
  -Jóvenes (confusión con enf. psicológicas)
  -Comorbilidades (EPOC, SAHOS, enfermedad renal)
- No se piensa en esta entidad

SOSPECHARLA

Frost A et al. Eur Respir J. 2019;53(1); 2. Brown et al. Chest. 2011;140(1):19-26

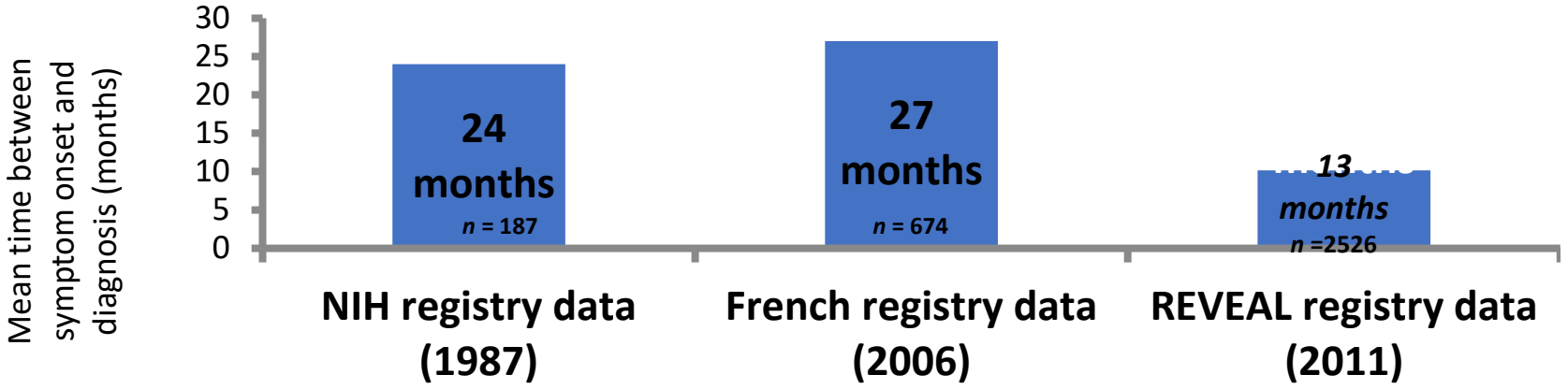
Diagnóstico Clínico HAP





Benza RL and col. Circulation. 2010;122:164-172. Humbert M and col. Am J Respir Crit care 2006;173:1023-30.

El retraso entre el inicio de los síntomas y el diagnóstico es ≥ 2 años



Rich S, *et al. Ann Int Med* 1987; 107:216-23.
Humbert M, *et al. Am J Respir Crit Care Med* 2006; 173:1023-30.
Brown LM, *et al. Chest* 2011; 140:19-26

Clínica

- **Síntomas→** inespecíficos y están presentes en otras patologías cardíacas y pulmonares más frecuentes

✓ **disnea**
✓ **fatiga**
✓ **debilidad**
✓ **angina**
✓ **síncope**
✓ **palpitaciones**

Inicialmente en esfuerzo → **<u>Casos avanzados</u>**
Síntomas en reposo

Sospechar la enfermedad en sujetos con disnea sin causa aparente

Hemoptisis (ruptura de AB hipertrofiadas)
Voz ronca (Compresión laríngeo recurrente)
Sibilancias (Compresión vía aérea)
Angina/Sincope (Compresión TCI)

Galie N, Humbert M. 2015 ESC/ERS .

Exámen Físico

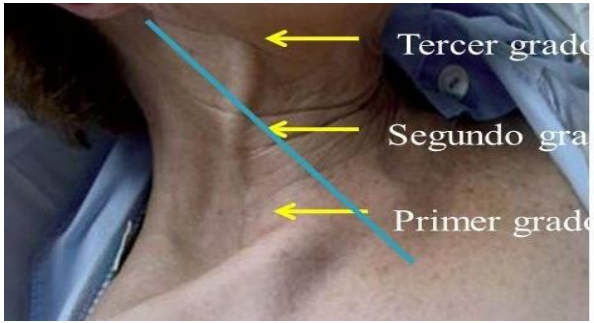
- Latido sagital: latido paraesternal izquierdo positivo con retracción de la zona de la punta(dilatación del VD) La inspiración acentúa intensidad y amplitud
- R2(P2) pulmonar acentuado
- R3 del VD (disminución de distensibilidad del VD)
- Soplo Holo sistólico de IT por dilatación anillo
- Soplo protomeso diastólico de IP por dilatación del anillo pulmonar (Soplo de Graham Steel)
- Pulso hepático, por transmisión flujo desde VD : IT severa

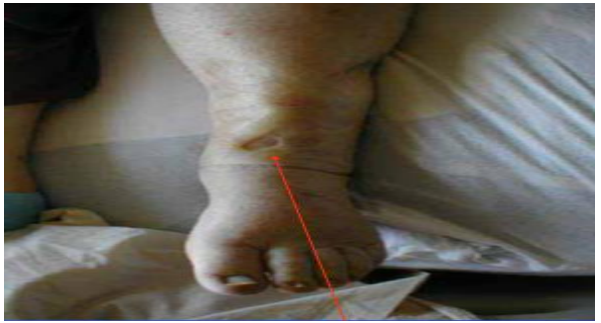
Galie N, Humbert M. 2015 .

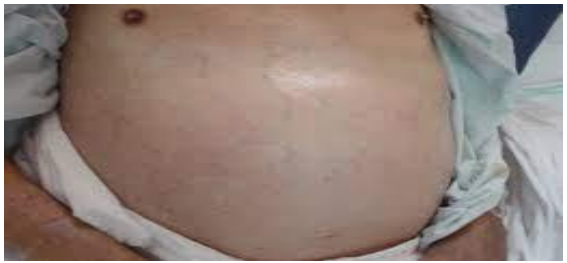
Exámen Físico

Disfunción del VD: **Signos de Congestión venosa:**

-Ingurgitación yugular
-RHY positivo
-Hepatomegalia
-Edemas MII
-Derrame Pleural
-Ascitis
-Cianosis



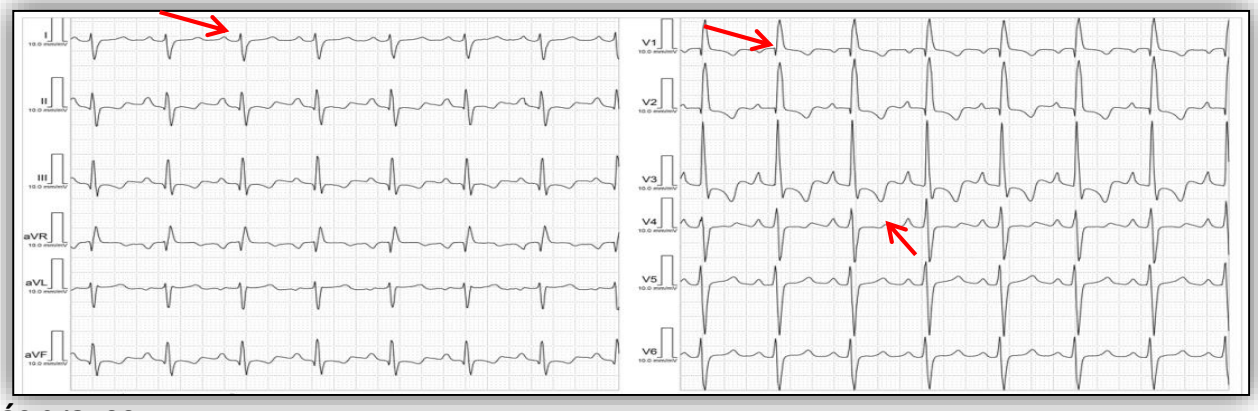




Strange G. Pulm Circ 2013 Jan-Mar; 3(1): 89–94

Electrocardiograma

ECG normal no excluye el diagnóstico



<u>ECG anormal es probable en casos más graves</u>:

- **Eje a la derecha** (desviación del eje QRS >90°-complejo negativo DI avl positivo DIII y avf)
- **Agrandamiento AD : onda P pulmonar** (amplitud onda P >2,5 mm DII)
- **HVD** (R altas en precordiales derechas,S profundas en izquierdas)
- **Trastorno repolarización en precordiales derechas (ondas T negativas asimétricas con depresión ST-T)**
- **BCRD**
- **Taquiarritmias : Taquicardia Sinusal**

Galie N, Humbert M. 2015 .

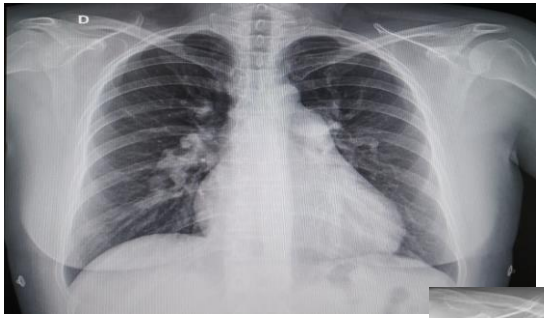
Rx torax

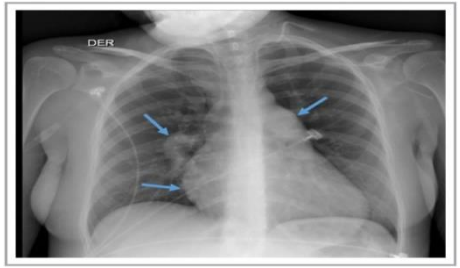
Normal en el 90% de los casos

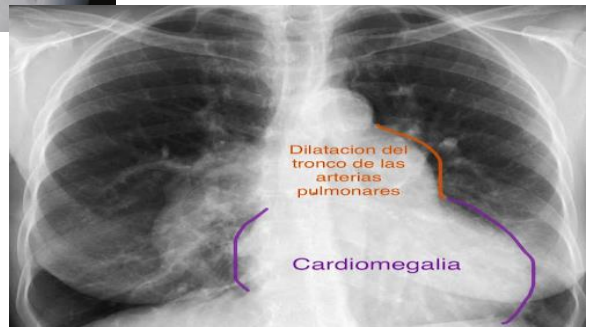
- Dilatación de AP central
(con perdida vascular periférica)

- Agrandamiento AD y VD

- Sin relación con la magnitud de HTP

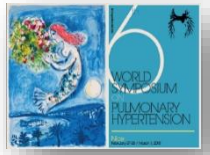


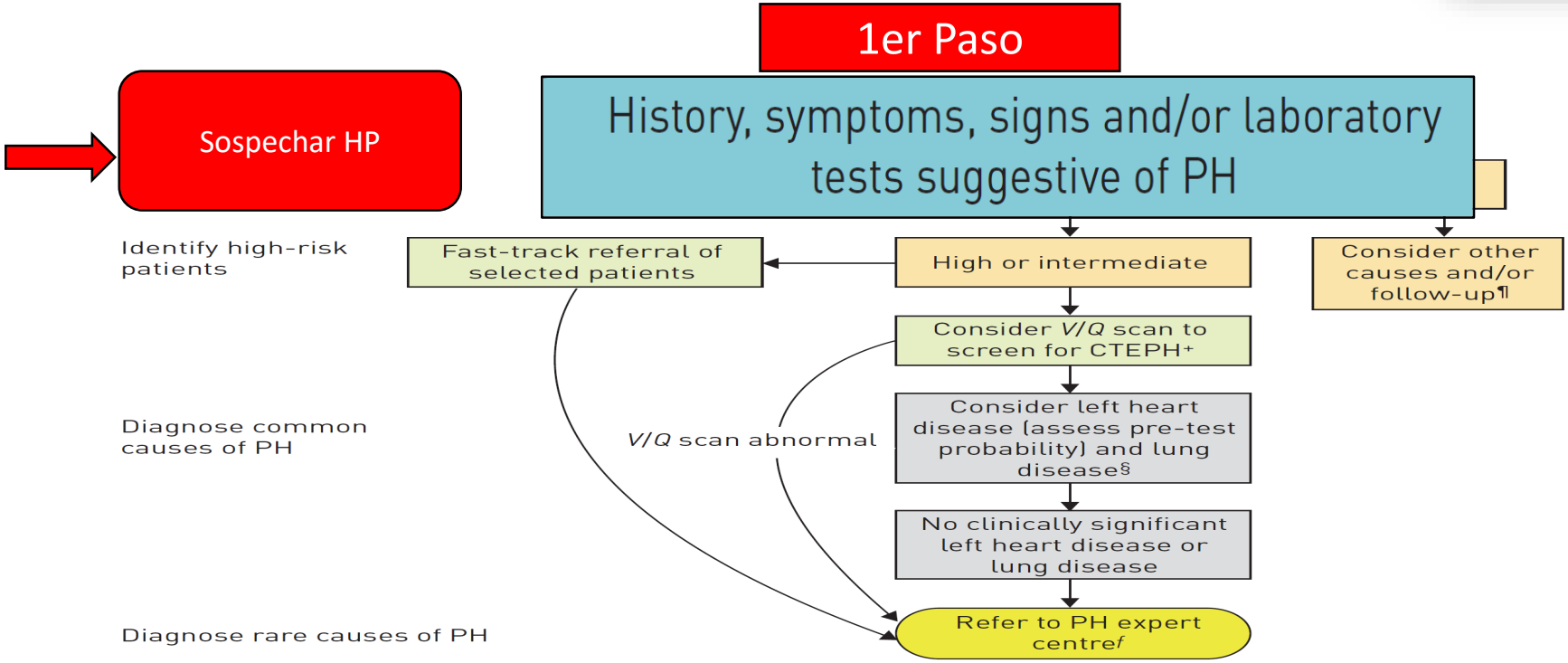




La radiografía de tórax no es suficiente para el diagnóstico de HP pero, al igual que el electrocardiograma, forma parte de los estudios iniciales.

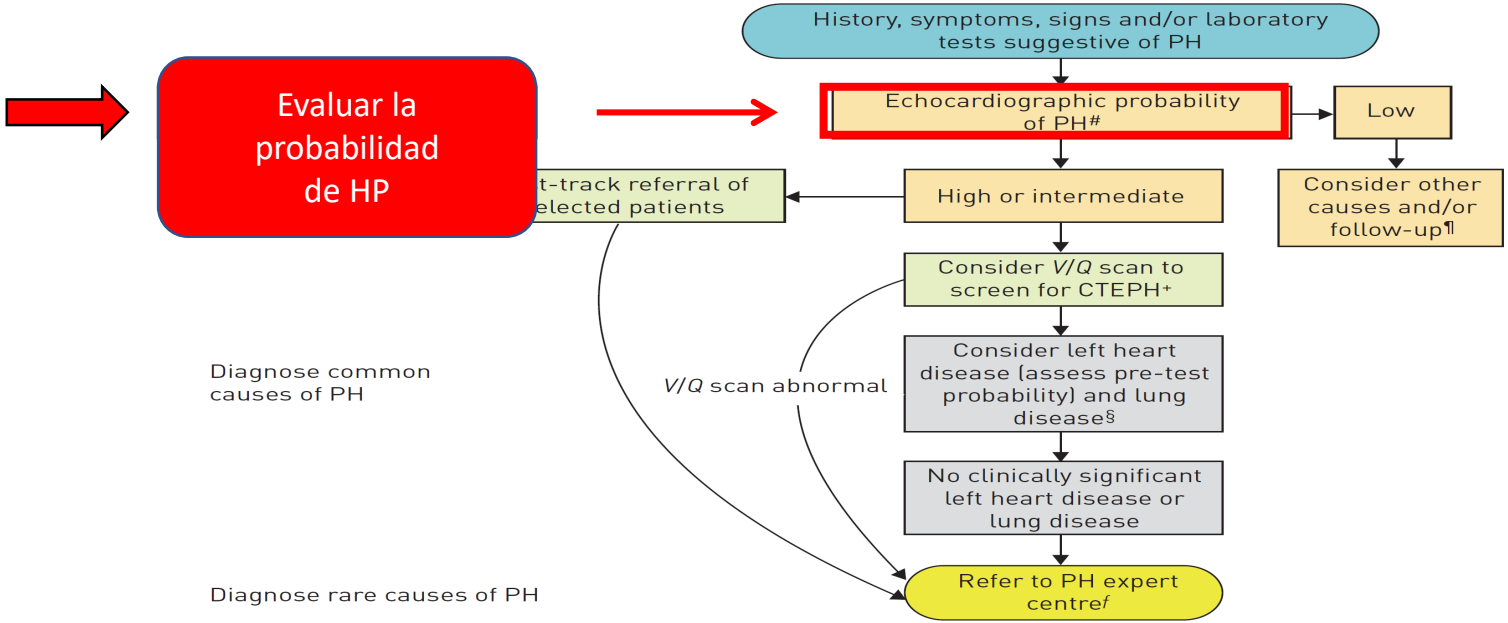
Algoritmo diagnóstico





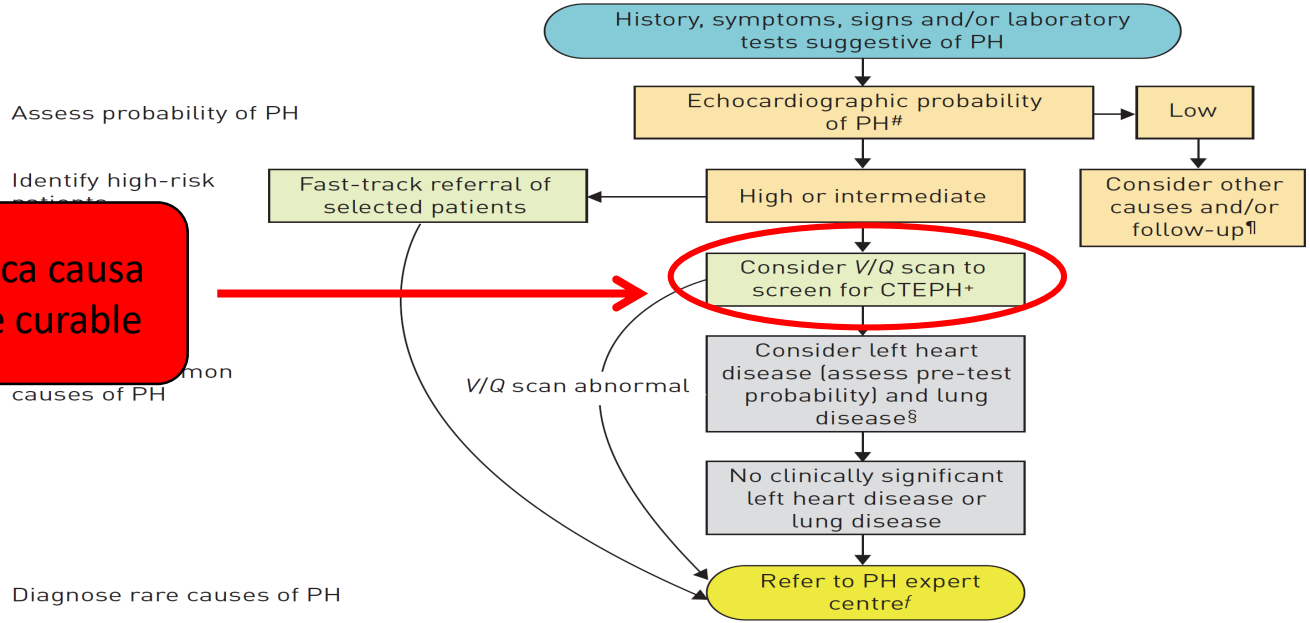
Adaani Frost et al. Eur Respir J 2019;53:1801904

2do Paso: Evaluar Probabilidad



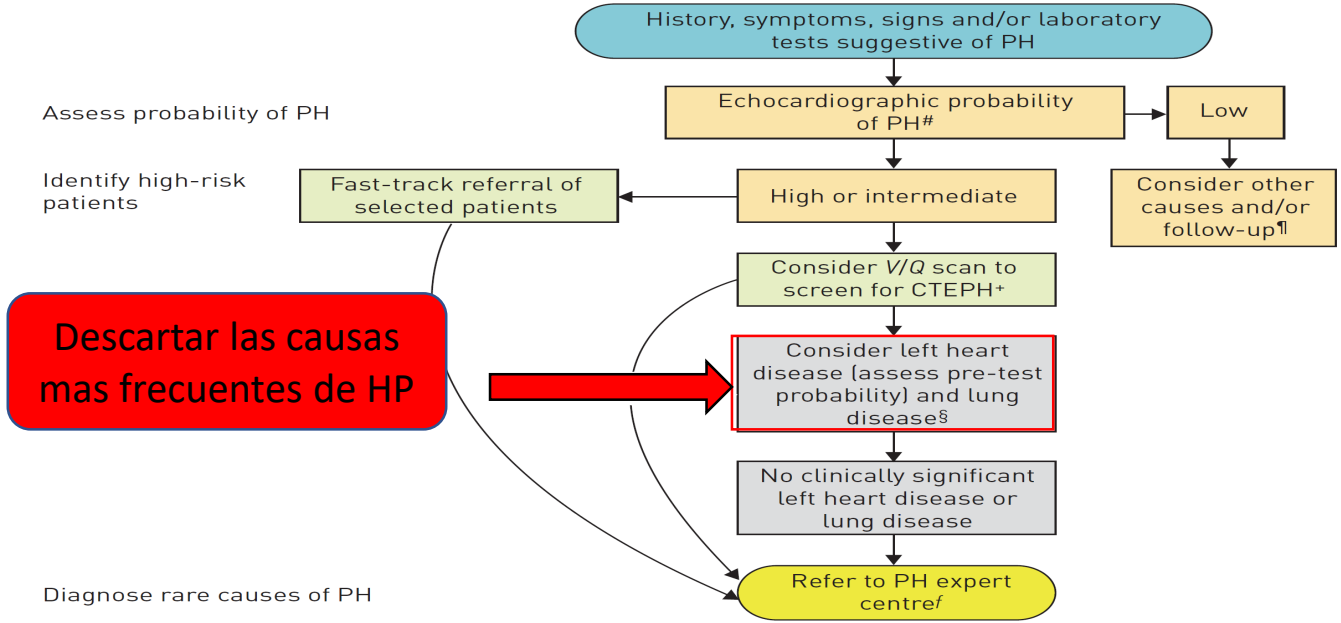
3er Paso: Algoritmo Diagnóstico

Descartar la única causa potencialmente curable



4to paso: Algoritmo diagnóstico

Descartar las causas mas frecuentes de HP

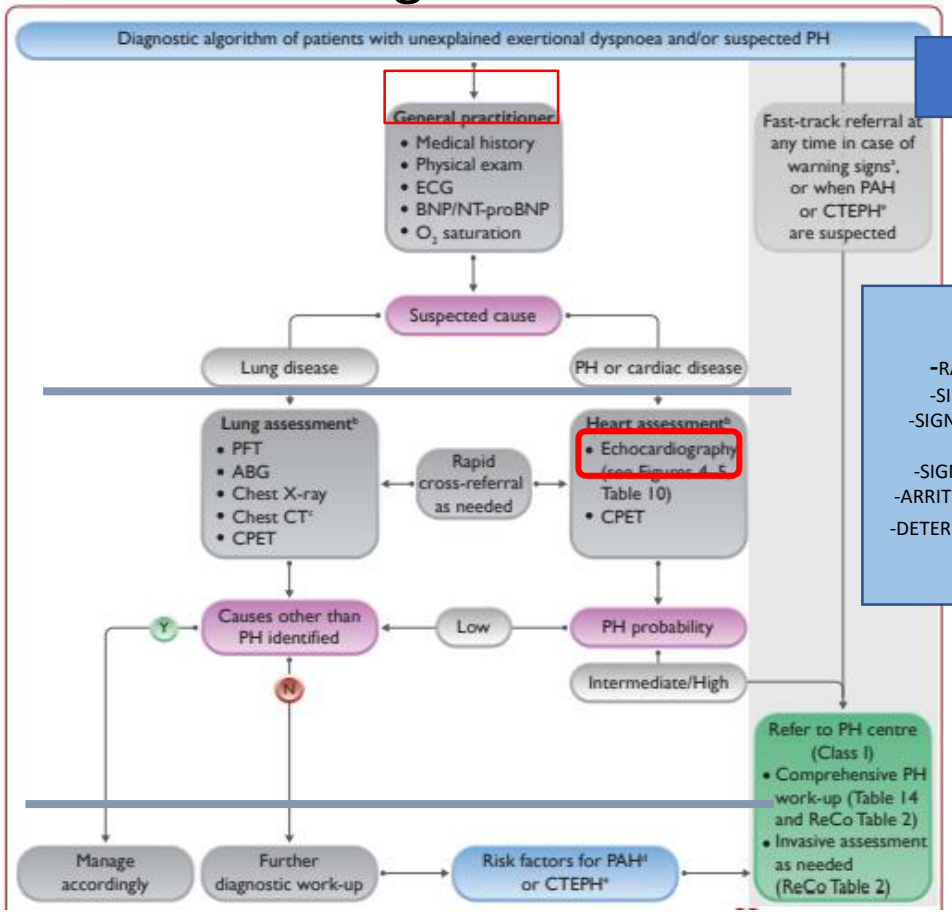


Diagnóstico

SOSPECHA

DETECCIÓN

CONFIRMACIÓN



DERIVACION

-RAPIDA EVOLUCIÓN.
-SINTOMAS SEVEROS.
-SIGNOS DE FALLA DEL VD.
-SINCOPE
-SIGNOS DE BAJO GASTO.
-ARRITMIAS MAL TOLERADAS.
-DETERIORO HEMODINÁMICO.

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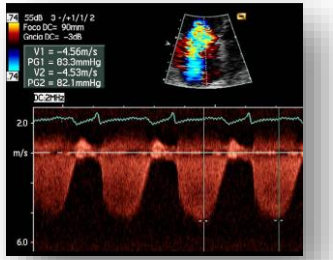
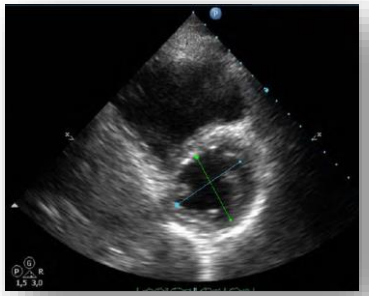
Ecocardiograma Doppler

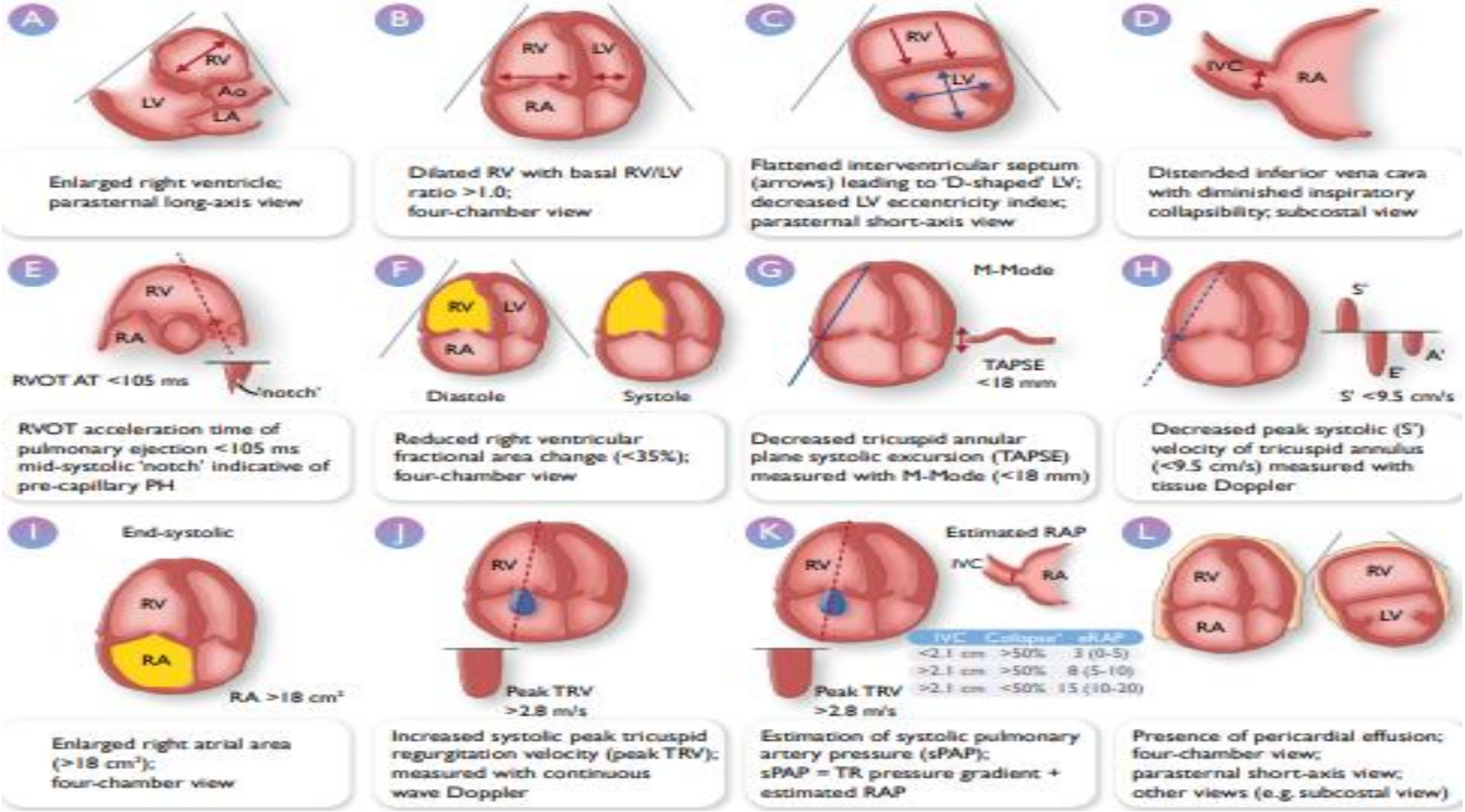
Herramienta más utilizada para screening para guiar la indicación de CCD

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	





Galiè N, et al. Eur Heart J 2009; 30:2493-537.
Rudski LG, et al. J Am Soc Echocardiogr 2010; 23:685-713.



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DIAGNÓSTICO – Cateterismo Derecho

Measured variables	Normal value
Right atrial pressure, mean (RAP)	2–6 mmHg
Pulmonary artery pressure, systolic (sPAP)	15–30 mmHg
Pulmonary artery pressure, diastolic (dPAP)	4–12 mmHg
Pulmonary artery pressure, mean (mPAP)	8–20 mmHg
Pulmonary arterial wedge pressure, mean (PAWP)	≤15 mmHg
Cardiac output (CO)	4–8 L/min
Mixed venous oxygen saturation (SvO_2)[a]	65–80%
Arterial oxygen saturation (SaO_2)	95–100%
Systemic blood pressure	120/80 mmHg
Calculated parameters	
Pulmonary vascular resistance (PVR)[b]	0.3–2.0 WU
Pulmonary vascular resistance index (PVRI)	3–3.5 WU·m^2
Total pulmonary resistance (TPR)[c]	<3 WU
Cardiac index (CI)	2.5–4.0 L/min·m^2
Stroke volume (SV)	60–100 mL
Stroke volume index (SVI)	33–47 mL/m^2
Pulmonary arterial compliance (PAC)[d]	>2.3 mL/mmHg

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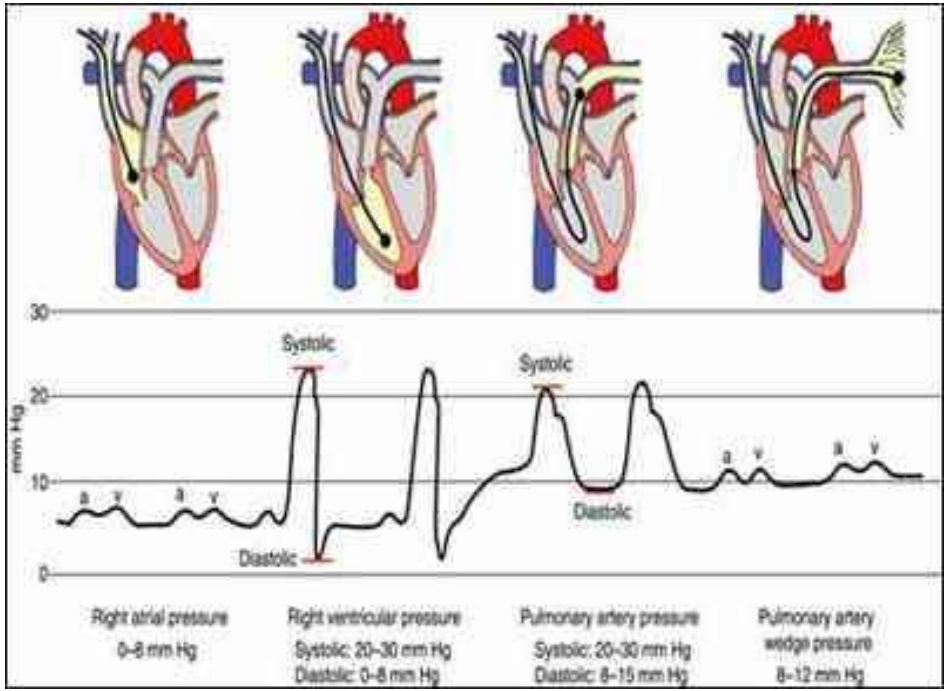
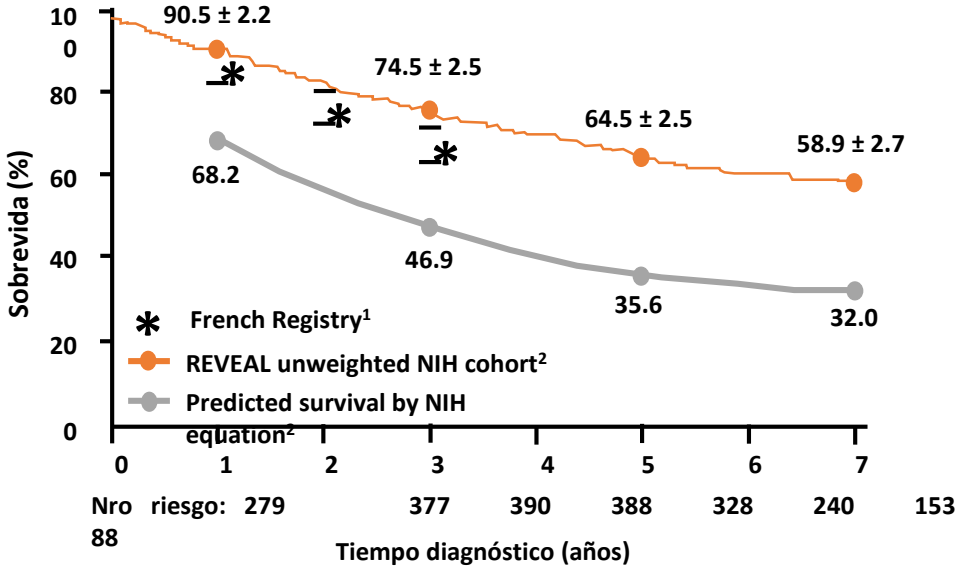
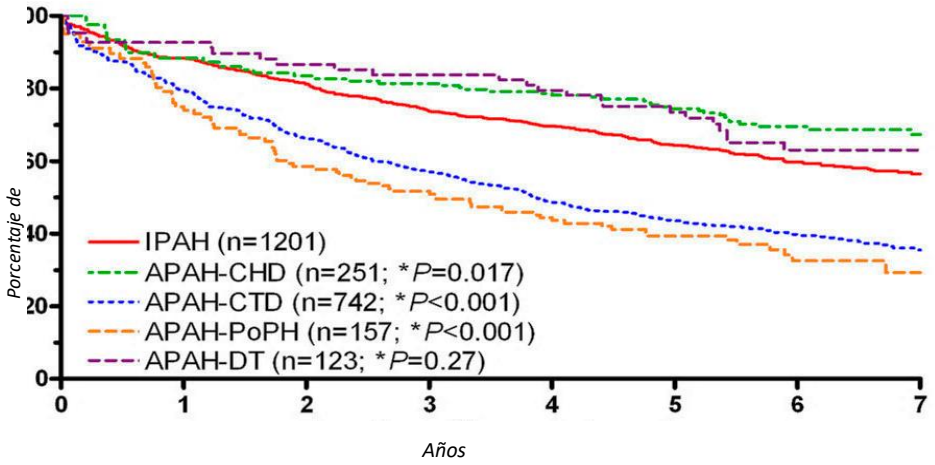

30
20
10
0
mm Hg
Systolic
Systolic
Diastolic
Diastolic
a
v
a
v
a
v
a
v
Right atrial pressure
0–8 mm Hg
Right ventricular pressure
Systolic: 20–30 mm Hg
Diastolic: 0–8 mm Hg
Pulmonary artery pressure
Systolic: 20–30 mm Hg
Diastolic: 8–15 mm Hg
Pulmonary artery wedge pressure
8–12 mm Hg

Pronóstico





1. Humbert M, et al. Eur Respir J 2010; 36:549–555. 2. Benza RL, et al. Chest 2012

Danzker R and col. Ann Intern Med 1987:107;216-223

Determinants of prognosis (estimated 1-year mortality)	Low risk (<5%)	Intermediate risk (5–20%)	High risk (>20%)
Clinical observations and modifiable variables			
Signs of right HF	Absent	Absent	Present
Progression of symptoms and clinical manifestations	No	Slow	Rapid
Syncope	No	Occasional syncope[a]	Repeated syncope[b]
WHO-FC	I, II	III	IV
6MWD[c]	>440 m	165–440 m	<165 m
CPET	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	Peak VO_2 <11 mL/min/kg (<35% pred.) VE/VCO_2 slope >44
Biomarkers: BNP or NT-proBNP[d]	BNP <50 ng/L NT-proBNP <300 ng/L	BNP 50–800 ng/L NT-proBNP 300–1100 ng/L	BNP >800 ng/L NT-proBNP >1100 ng/L
Echocardiography	RA area <18 cm^2 TAPSE/sPAP >0.32 mm/mmHg No pericardial effusion	RA area 18–26 cm^2 TAPSE/sPAP 0.19–0.32 mm/mmHg Minimal pericardial effusion	RA area >26 cm^2 TAPSE/sPAP <0.19 mm/mmHg Moderate or large pericardial effusion
cMRI[e]	RVEF >54% SVI >40 mL/m^2 RVESVI <42 mL/m^2	RVEF 37–54% SVI 26–40 mL/m^2 RVESVI 42–54 mL/m^2	RVEF <37% SVI <26 mL/m^2 RVESVI >54 mL/m^2
Haemodynamics	RAP <8 mmHg CI ≥2.5 L/min/m^2 SVI >38 mL/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 L/min/m^2 SVI 31–38 mL/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 L/min/m^2 SVI <31 mL/m^2 SvO_2 <60%

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Re estratificación de Riesgo en el Seguimiento

Determinants of prognosis	Low risk	Intermediate–low risk	Intermediate–high risk	High risk
Points assigned	1	2	3	4
WHO-FC	I or II[a]	-	III	IV
6MWD, m	>440	320–440	165–319	<165
BNP or NT-proBNP,[a] ng/L	<50 <300	50–199 300–649	200–800 650–1100	>800 >1100

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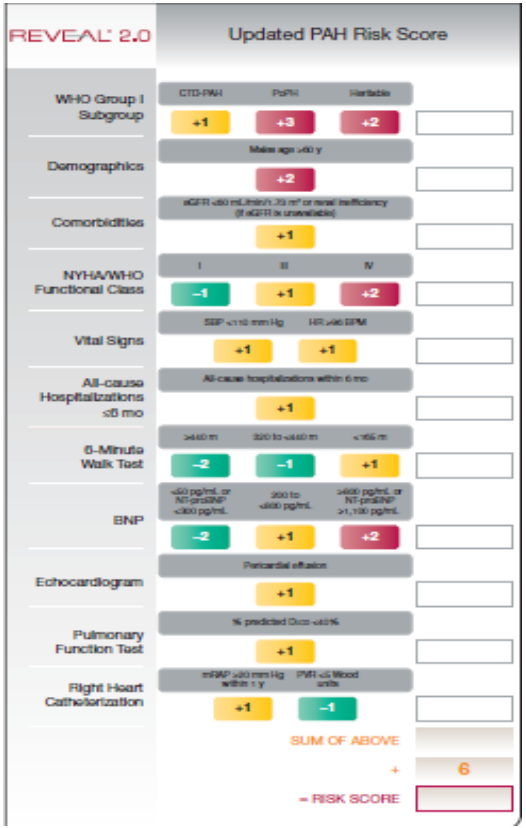
Score REVEAL 2.0

- *Subgrupos HAP G 1 (HAP-ETC, HTPPP, Hereditable)*
- *Sexo masculino > 60 a*
- *IR (TFG < 60 ml/kg/min)*
- *Clase Funcional WHO/NYHA*
- *TAS < 110, FC > 96*
- *Hospitalizaciones dentro de 6 meses*
- *TC6M (mts)*
- *BNP/ NTProBNP*
- *Derrame pericárdico en Ecocardiograma*
- *DLCO < 40*
- *Cat. Derecho PmAP > 20 durante 1 a, RVP*

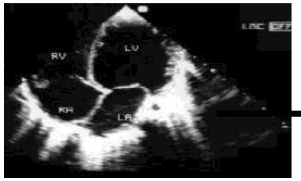
13: Alto riesgo



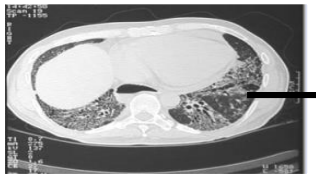
CHEST 2019; 156(2):323-337

Tratamiento de la HP: no son todas iguales

HP G II-III-IV (90%)



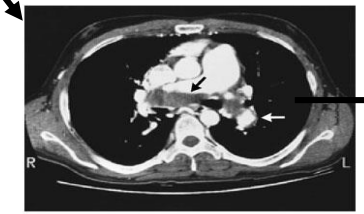
ARNI/ IECA/ARA2- iSGLT2 BBloqueantes ; Antagonistas Aldosterona; diuréticos etc.



Oxígeno, Broncodilatadores,

Drogas específicas FPI

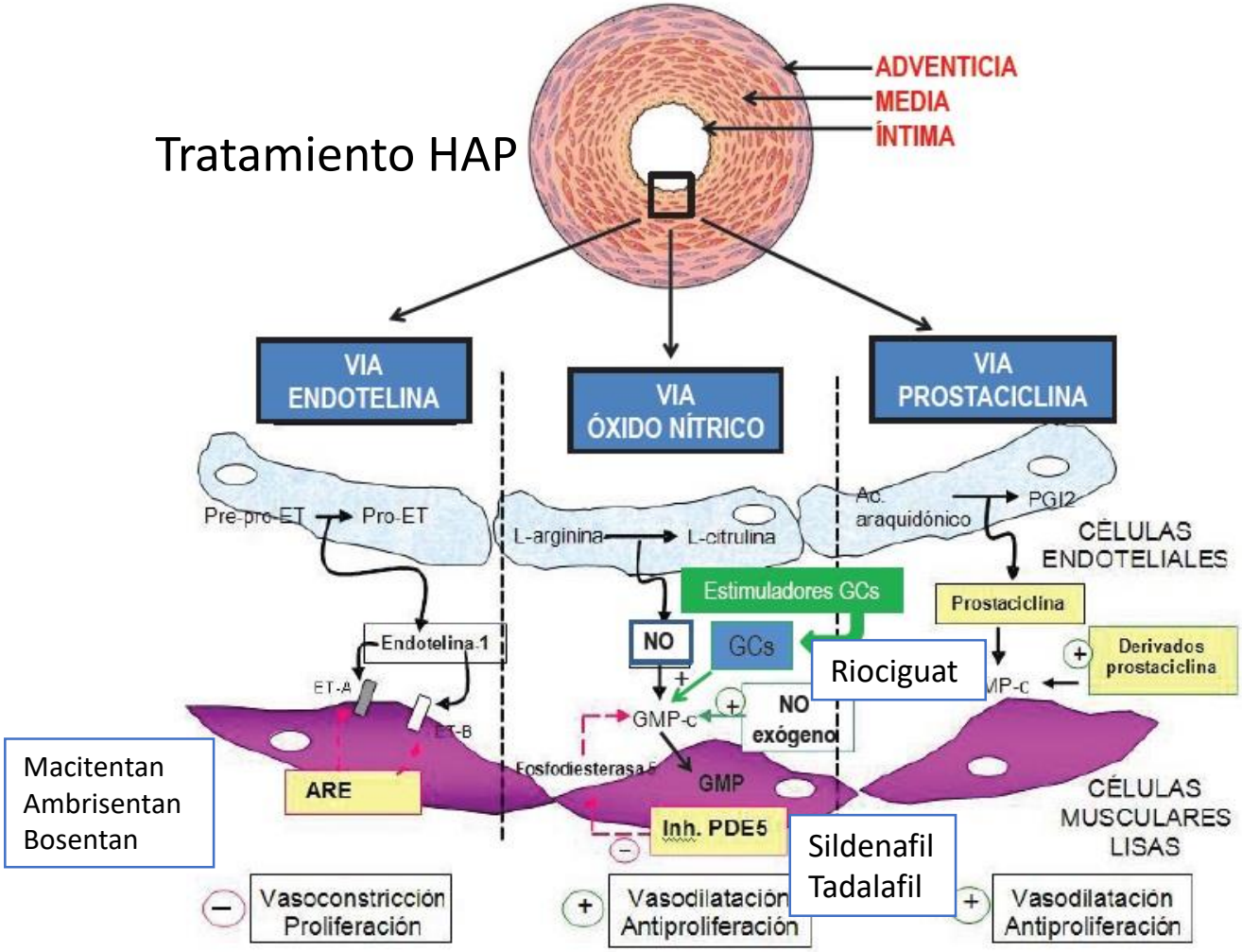
Treprostinil inhalado



Antigoagulantes orales
Endarterectomía, APB,

Riociguat

Tratamiento HAP



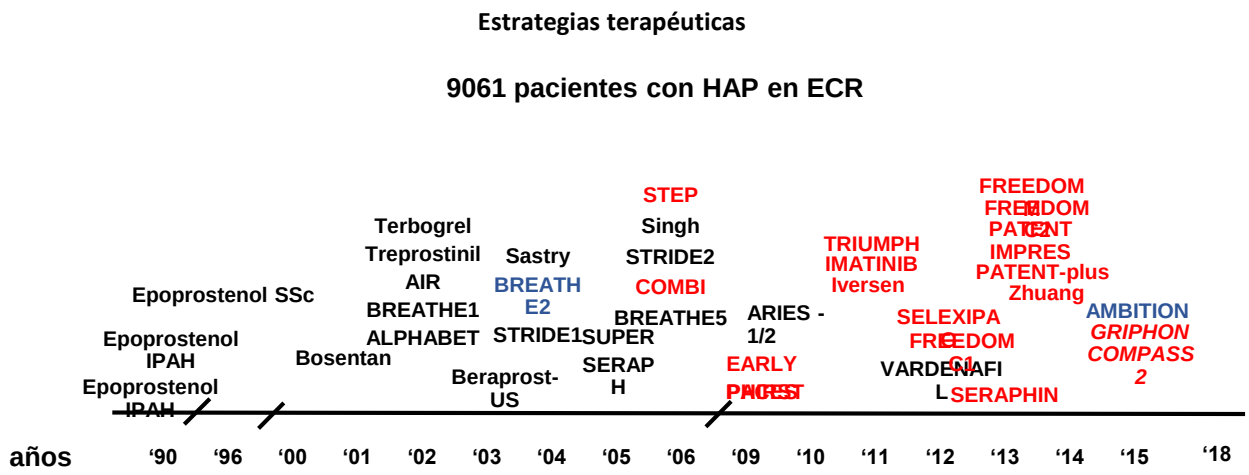
Riociguat

Macitentan
Ambrisentan
Bosentan

Sildenafil
Tadalafil

Iloprost inh.
Treprostinil sc/inh.
Epoprostenol ev.
Selexipag

Historia de los Ensayos Clínicos Randomizados en HAP (41)



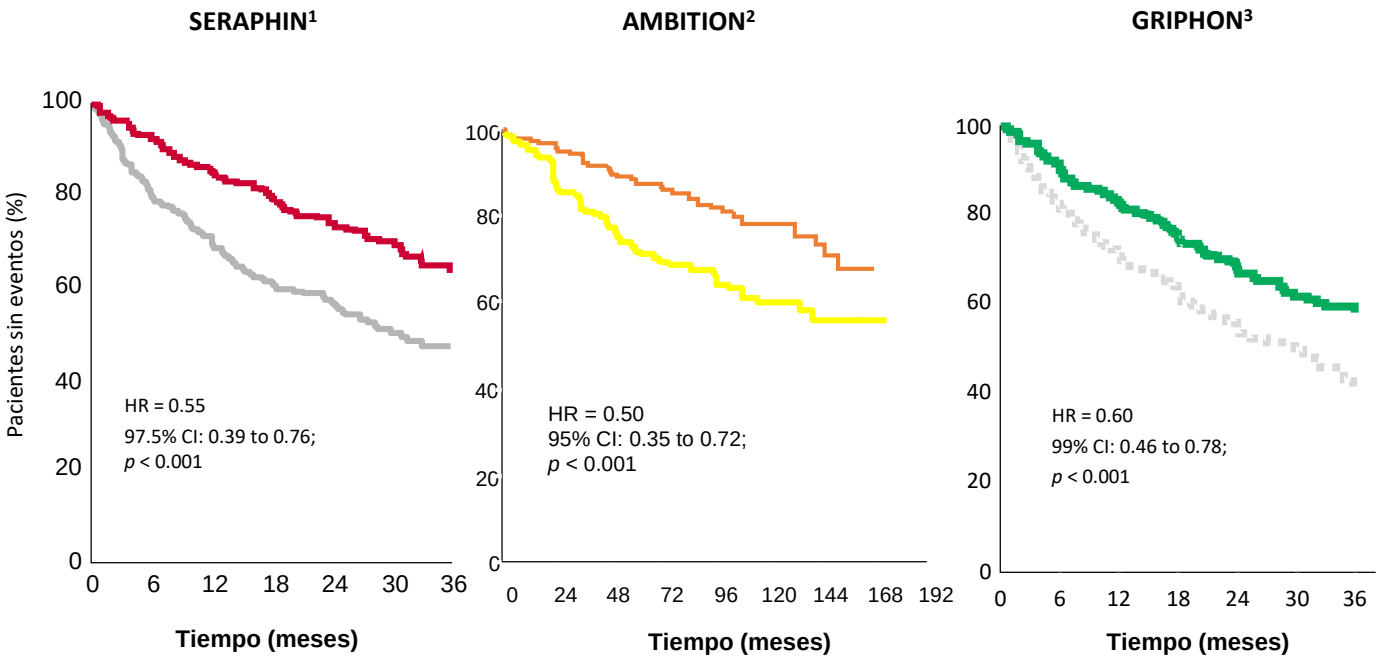
RCTs on monotherapy vs placebo or vs monotherapy (21)

RCTs on monotherapy and/or sequential combination vs placebo (18)

RCTs on initial combination vs monotherapy (2)

Modified from Galiè N, et al. Eur Heart J 2010; 31:2080-6.

EVIDENCIA EN TRATAMIENTO COMBINADO



1. Pulido T, *et al. N Engl J Med* 2013; 369:809-18;
2. Galiè N, *et al. N Engl J Med* 2015; 373:834-44;
3. Sitbon O, *et al. N Engl J Med* 2015; 373:2522-33.

Terapia Combinada versus Monoterapia: Meta-análisis

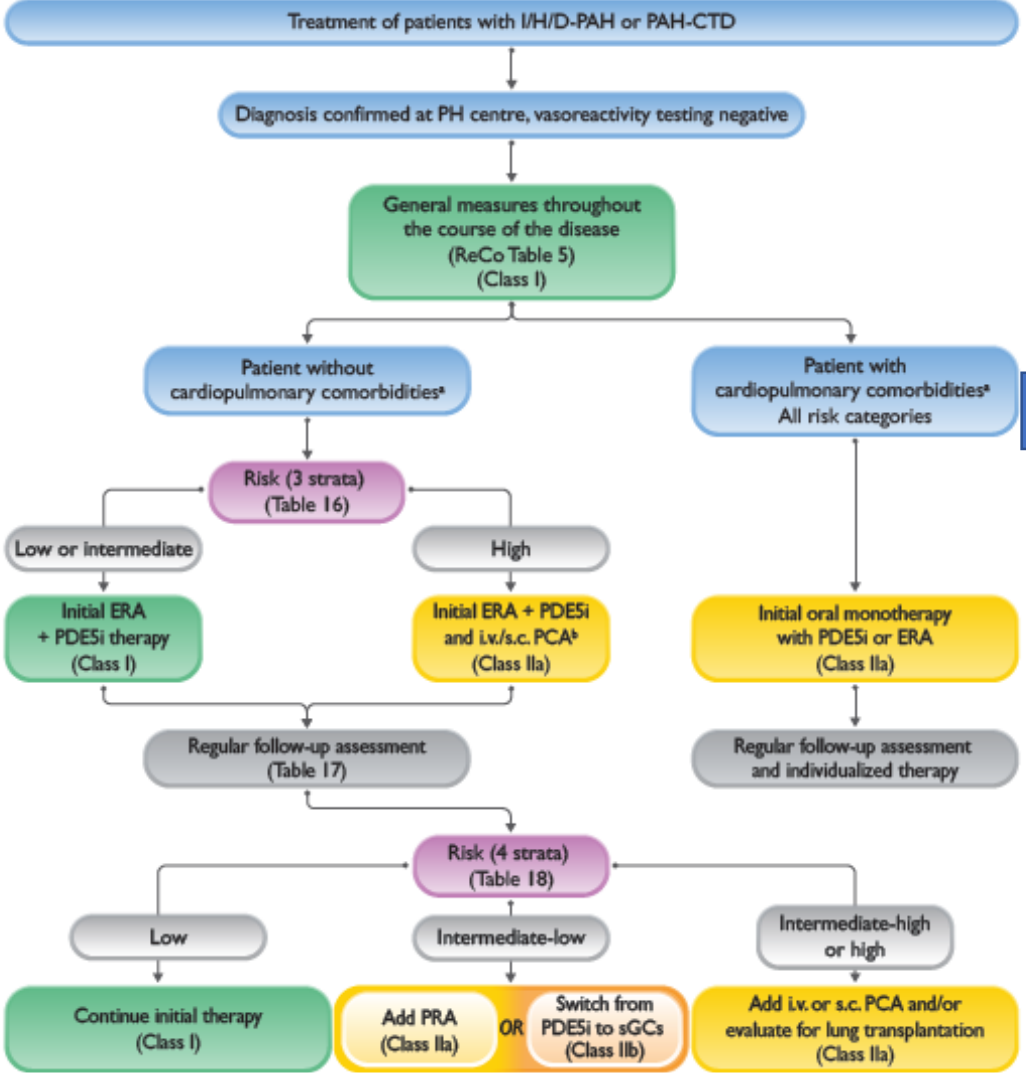
	Number of studies	Proportion of events (%)			Fixed-effect model		Random-effects model		Homogeneity	
		With combined therapy	With monotherapy	Total	Pooled RR	95 % CI (p value)	Pooled RR	95% CI (p value)	P value	I^2 (%)
Primary outcome										
Clinical worsening (all events)	15[11-14,29-32,34-40]	332/1940 (17%)	517/1862 (28%)	849/3802 (22%)	0·65	0·58–0·72 (p<0·00001)	0·65	0·56–0·76 (p<0·00001)	0·25	18%
Secondary outcomes as first event of clinical worsening										
All-cause mortality	12[11-14,30,32,34-40]	54/1711 (3%)	60/1712 (4%)	114/3423 (3%)	0·92	0·65–1·32 (p=0·65)	0·97	0·63–1·49 (p=0·88)	0·33	13%
Admission to hospital (PAH-related)†	8[11-13,32,34-37]	172/1658 (10%)	245/1680 (15%)	417/3338 (13%)	0·71	0·60–0·85 (p=0·0002)	0·71	0·53–0·96 (p=0·03)	0·12	37%

RR=rate ratio. PAH=pulmonary arterial hypertension. *Atrial septostomy were not reported in any study. †Data from the combination subgroup of the SERAPHIN trial are included in this analysis because they were available in a subsequent article.[45] However, these data were not included in the primary analysis because admission to hospital was not included in the definition of clinical worsening in SERAPHIN. ‡All deaths, including those as first event of clinical worsening and those after censoring for another event. Data from PHIRST,[31] EARLY,[40] PATENT-1,[29] and SERAPHIN[35] for the subgroup of patients already on background therapy were not available. Therefore, data from the entire study population were included for this secondary analysis. However, the exclusion of these studies did not modify the results for the all-cause mortality (RR 0·88 [95% CI 0·72–1·06], p=0·18). §Data from SERAPHIN[35] for the subgroup of patients already on background therapy were not available. The exclusion of this study yielded similar RRs for PAH-related mortality (RR 0·81 [95% CI 0·61–1·08]).

16 ECR 4538 pacientes. Seguimiento 37 semanas (%)

	RRR(%)	valor de p
Empeoramiento clínico	-35	0.00001
1ra Hospitalización IC	-29	0.0002
Mortalidad Global	-14	0.09

Lajoie AC and col. Lancet Respir Med 2016



-Disfunción diastólica.
-Obesidad.
-HTA
-Diabetes.
-Enfermedad coronaria.
-Enfermedad del parénquima pulmonar.

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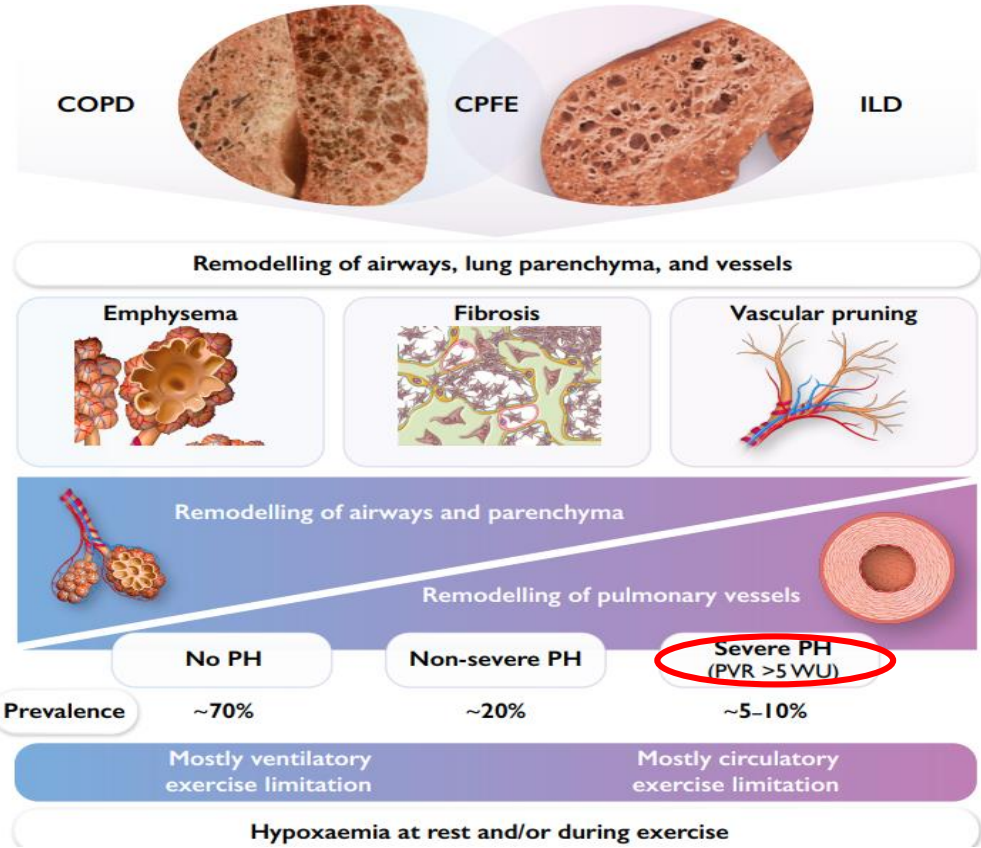
Hipertensión Pulmonar Grupo 3



HP severa previa:

PAPm>35 mmHg ó

PAPm≥ 25 mmHG con IC < 2,5L/min/m2

La RVP > ó < 5 U Wood predice mejor pronóstico

HP severa actual:

La RVP > 5 U Wood

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	COPD without PH	COPD with post-capillary PH	"Proportionate" mild-moderate PH	Pulmonary vascular phenotype
Risk of mortality due to PH				
FEV_1	↓ / ↓↓ / ↓↓↓	↓/ ↓↓ / ↓↓↓	↓↓/ ↓↓↓	↓
DLCO	N / ↓	↓	↓/ ↓↓	↓↓↓
6MWT CPET	↓	↓	↓ / ventilatory limitation	↓ / cardiovascular limitation
PaO_2	N / ↓	N / ↓	↓/↓↓/↓↓↓	↓/↓↓/↓↓↓
Pulmonary hemodynamics	mPAP ≤20 mmHg or mPAP 21-24 mmHg & PVR <3 WU	mPAP >20 mmHg PAWP >15 mmHg	mPAP 21-24 mmHg & PVR ≥3 WU or mPAP 25-34 mmHg	mPAP 25-34 mmHg & CI<2.5 L/min/m^2 or mPAP ≥35 mmHg

Criteria favouring group 1 (PAH)	Testing	Criteria favouring group 3 (PH due to lung disease)
Extent of lung disease		
Normal or mildly impaired: • FEV_1 >60% pred (COPD) • FVC >70% pred (IPF) • Low diffusion capacity in relation to obstructive/restrictive changes	Pulmonary function testing	Moderate to very severely impaired: • FEV_1 <60% pred (COPD) • FVC <70% pred (IPF) • Diffusion capacity "corresponds" to obstructive/restrictive changes
Absence of or only modest airway or parenchymal abnormalities	High-resolution CT scan¶	Characteristic airway and/or parenchymal abnormalities
Haemodynamic profile		
Moderate-to-severe PH	Right heart catheterisation ~~Echocardiogram~~	Mild-to-moderate PH
Ancillary testing		
Present	Further PAH risk factors (*e.g.* HIV, connective tissue disease, *BMPR2* mutations, *etc.*)	Absent
Features of exhausted circulatory reserve: • Preserved breathing reserve • Reduced oxygen pulse • Low $CO/\dot{V}O_2$ slope • Mixed venous oxygen saturation at lower limit • No change or decrease in P_{aCO_2} during exercise	Cardiopulmonary exercise test+ (P_{aCO_2} particularly relevant in COPD)	Features of exhausted ventilatory reserve: • Reduced breathing reserve • Normal oxygen pulse • Normal $CO/\dot{V}O_2$ slope • Mixed venous oxygen saturation above lower limit • Increase in P_{aCO_2} during exercise
Predominant haemodynamic profile		Predominant obstructive/restrictive profile

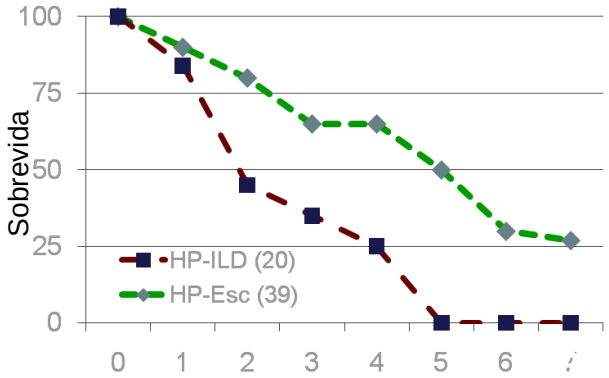
ERJ Express. Published on December 13, 2018

Esclerodermia

Hipertensión Pulmonar Aislada

Hipertensión Pulmonar con Enfermedad pulmonar intersticial

CPT < 60%
TAC : Fibrosis pulmonar



Aumenta la incidencia de neoplasias pulmonares
Disfunción ventricular izquierda
Derrame pericárdico (34%)

Arthritis & Rheumatism 2009;60:569-577

TABLE 1 Randomised controlled trials (RCTs) of pulmonary vasodilators in pulmonary hypertension (PH)-associated interstitial lung diseases (ILDs)

First author [reference]	Year	Name of RCT	Study population	Drug	Duration	Primary end-point	Primary end-point reached
King [76]	2008	BUILD-1	IPF	Bosentan	12 months	Change in 6MWD	N
Seibold [77]	2010	BUILD-2	SSc	Bosentan	12 months	Change in 6MWD	N
Zisman [78]	2010	STEP-IPF	IPF	Sildenafil	12 weeks	Change in 6MWD	N
King [79]	2011	BUILD-3	IPF	Bosentan	57 months	Combined[#]	N
Raghu [80]	2013	MUSIC	IPF	Macitentan	12 months	Change in FVC	N
Raghu [81]	2013	ARTEMIS-IPF	IPF	Ambrisentan	12 months	Combined[¶,+]	N
Corte [82]	2014		IPF/NSIP	Bosentan	16 weeks	Change in PVRI	N
Kolb [83]	2018	INSTAGE	IPF	Sildenafil	12 weeks	Change in SGRQ	N
Nathan [84]	2019	RISE-IIP	IIP	Riociguat	12 months	Change in 6MWD[+]	N
Nathan [85]	2020		PF-ILD	Inhaled NO	8 weeks	Change in MVPA[§]	Y
Behr [86]	2021	SP-IPF	IPF	Sildenafil	12 months	Combined[f]	N
Waxman [87]	2021	INCREASE	IIP	Inhaled treprostinil	16 weeks	Change in 6MWD	Y

Mensajes

- La HP grupo II y III son las mas frecuentes.
- La HAP es una entidad heterogénea con elevada morbimortalidad.
- El diagnostico precoz es fundamental. El ecocardiograma doppler es la herramienta inicial de sospecha de esta entidad.
- Los síntomas son inespecíficos por lo que el interrogatorio y los estudios complementarios pueden ayudar a sospecharla y derivar a centro de referencia de HP.
- La confirmación diagnostica se realiza por cateterismo cardiaco derecho en estado de euvolemia y reposo.
- Una vez realizado el diagnostico y definido el pronostico, es fundamental comenzar con el tratamiento especifico en HAP.

Muchas Gracias