HIPERTENSION PULMONAR

Epidemiología, definición y clasificación

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CÁTEDRA DE FISIOLOGïA

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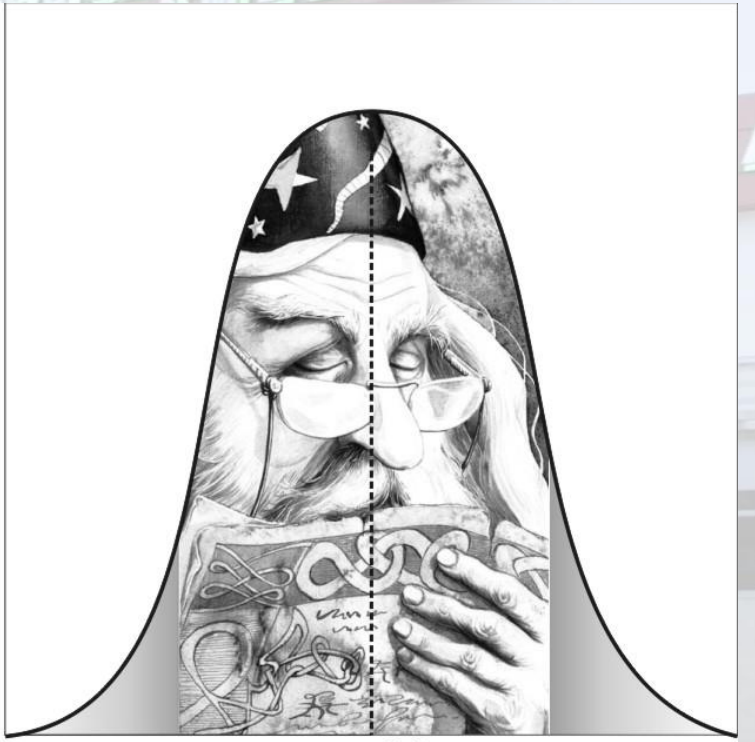
Cual es la presión pulmonar normal?



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Presión pulmonar normal

La reevaluación de los datos disponibles sugieren como normal una presión arterial **media** en reposo de 14+/-3 mmHg
Con un limite normal máximo de 20 mmHg.

El significado de las presiones entre 21 y 24 mmHg son inciertos

Badesch BD, et all. Diagnosis and assessment of pulmonary arterial hypertension. J Am Coll Cardiol 2009;54:S55–S56

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Definamos hipertensión pulmonar

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Incremento de la presión arterias pulmonar media por encima de 25 mmHg en reposo medido por cateterismo derecho

†la definición de hipertensión pulmonar de 30 mm hg durante el ejercicio no esta respaldada por bibliografía publicada.

† La hipertensión *arterial* pulmonar se caracteriza por aumento de la presión pulmonar pre capilar en ausencia de causas de hp pre capilar como enfermedad pulmonar , TEP crónico recurrente u otras causas

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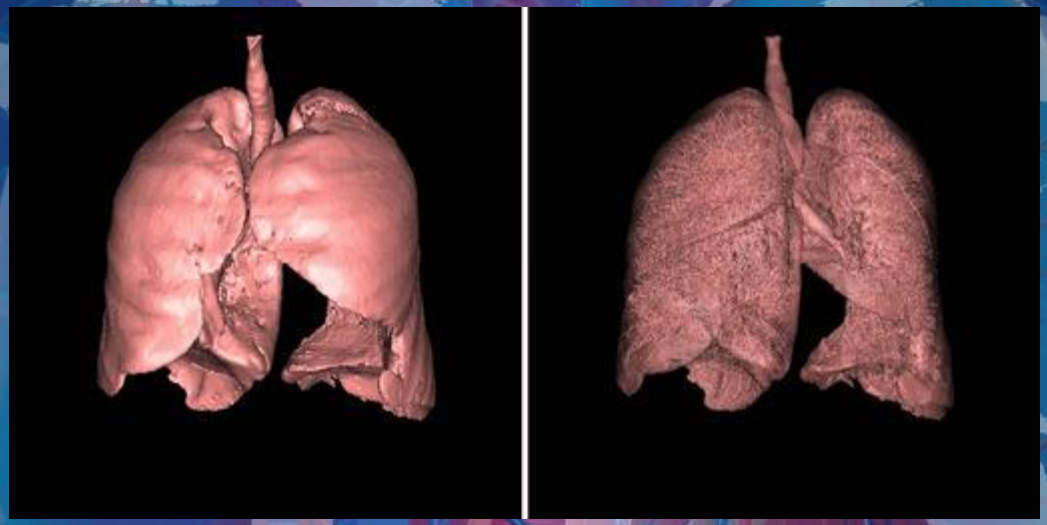
What is Pulmonary Hypertension (PH)?

Pulmonary hypertension (PH) is a rapidly progressive, deadly disease which affects the lungs and heart.[1,2]

It is characterized by high blood pressure in the arteries of the lungs.

There are 5 main types of PH which affect patients in different ways, all of which can lead to heart failure and death.[3]

5 types



What are the symptoms?

PH is a personal condition and symptoms vary in each individual, common symptoms may include:[4]

Breathlessness
Blue lips
Fatigue

Educating people to recognize the symptoms of PH could save lives.

Treatment

A range of pharmaceutical treatments are available but they only treat 1 of the 5 types of PH, called PAH.

There are currently

0 cures for 4 of the 5 forms of PH

- The only potentially curative treatment available is surgery for 1 form of PH called chronic thromboembolic pulmonary hypertension (CTEPH).

Some patients are eligible for lung or heart-lung transplant, although this is not always possible due to lack of available organs, or patients not being suitable for surgery.

Accurate and early diagnosis and treatment followed by continuous treatment monitoring can mean the difference between life and death.

More research is needed to improve understanding of how all 5 types can be treated effectively.[16]

The impact of PH

50% of patients die within two years if not treated.[5]

50%

The death rate for pulmonary arterial hypertension (PAH), the most studied form of PH, **is higher** than both breast or colorectal cancers.[6,7]

PH can have a profound impact on many aspects of daily life such as having difficulty climbing stairs, walking short distances or simply getting dressed.[8,9]

Time matters for people with PH

Diagnosis of PH takes approximately 2 years due to delay.[10]

- Symptoms are often non-specific, meaning PH is frequently mistaken for asthma or other conditions.

2 years

PH is a rapidly progressive disease and time lost in its progression cannot be entirely regained.[11,12,13]

With earlier diagnosis

and treatment, **survival and quality of life could be significantly improved.**[6]

Rapidly progressive disease

Who is affected?

It is thought that there are more than 25 million patients globally.[14]

25 million

One of the rare types of PH, called PAH, affects approximately 52 people per million.[15]

People of all ages, including children, can develop PH although **it is most likely to be diagnosed in people between 40-50 years of age.**[5]

40-50 years

References

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Mayor frecuencia en el sexo femenino
Entre 20-50 años
Poblaciones de riesgo con antecedentes familiares (3,8%),
consumo de anorexígenos,
colagenopatías, cardiopatías congénitas,
portadores de HIV
hipertensión portal

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2100 casos en Argentina

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Disnea, astenia , ocasionalmente sincope

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SUB DIAGNOSTICO

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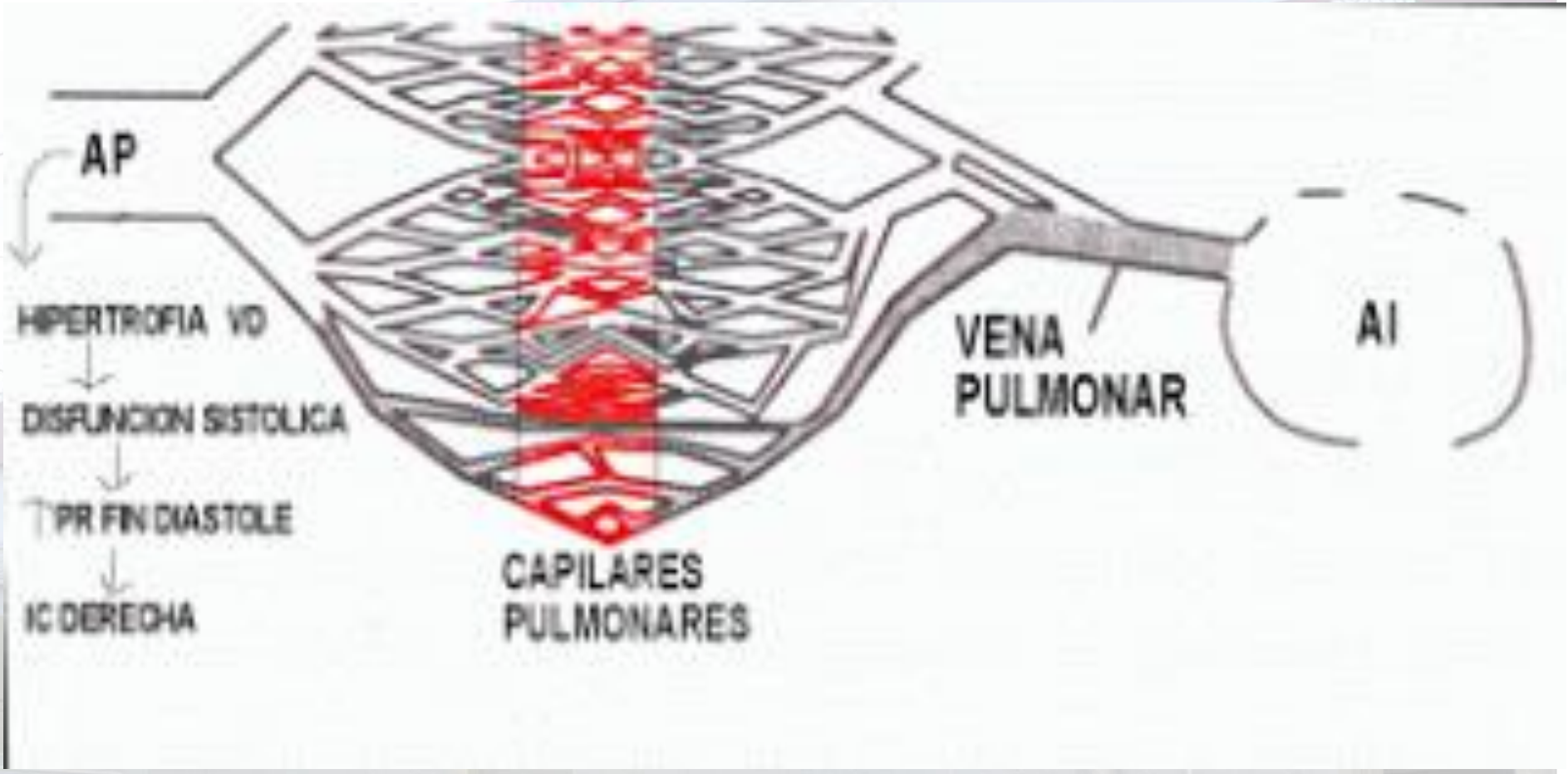

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European Heart Journal
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ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

Authors/Task Force Members: Nazzareno Galiè[*] (ESC Chairperson) (Italy), Marc Humbert[*a] (ERS Chairperson) (France), Jean-Luc Vachiery[c] (Belgium), Simon Gibbs (UK), Irene Lang (Austria), Adam Torbicki (Poland), Gérald Simonneau[a] (France), Andrew Peacock[a] (UK), Anton Vonk Noordegraaf[a] (The Netherlands), Maurice Beghetti[b] (Switzerland), Ardeschir Ghofrani[a] (Germany), Miguel Angel Gomez Sanchez (Spain), Georg Hansmann[b] (Germany), Walter Klepetko[c] (Austria), Patrizio Lancellotti (Belgium), Marco Matucci[d] (Italy), Theresa McDonagh (UK), Luc A. Pierard (Belgium), Pedro T. Trindade (Switzerland), Maurizio Zompatori[e] (Italy) and Marius Hoeper[a] (Germany)

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ESC Committee for Practice Guidelines (CPG) and National Cardiac Societies document reviewers: listed in Appendix

[a]Representing the European Respiratory Society; [b]Representing the Association for European Paediatric and Congenital Cardiology; [c]Representing the International Society for Heart and Lung Transplantation; [d]Representing the European League Against Rheumatism; and [e]Representing the European Society of Radiology.

ESC entities having participated in the development of this document:

ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

ESC Councils: Council for Cardiology Practice (CCP), Council on Cardiovascular Nursing and Allied Professions (CCNAP), Council on Cardiovascular Primary Care (CCPC).

ESC Working Groups: Cardiovascular Pharmacotherapy, Cardiovascular Surgery, Grown-up Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Valvular Heart Disease.

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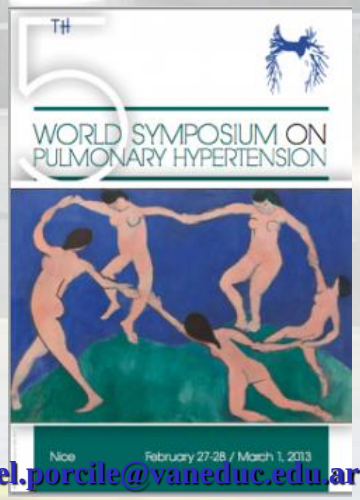
Actualización en clasificación de la hipertensión pulmonar

* Hipertensión arterial pulmonar (PAH)
* Secundaria a falla ventricular izquierda
* Secundaria a enfermedad pulmonar con o sin hipoxemia
* Hipertensión pulmonar secundaria a tromboembolismo crónico
* Mecanismos poco claros o multifactorial



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WHO Classification of Pulmonary Hypertension

Group 1: Pulmonary arterial hypertension

- Idiopathic PAH (iPAH)
- Heritable PAH (HPAH)
- Drug- and toxin-induced
- Associated with PAH (APAH)
  - Connective tissue diseases
  - HIV infection
  - Portal hypertension
  - Congenital heart disease (CHD)
  - Schistosomiasis
- Persistent pulmonary hypertension of the newborn (PPHN)

Group 2: Pulmonary hypertension due to left heart disease

- Systolic dysfunction
- Diastolic dysfunction
- Valvular disease

Group 3: Pulmonary hypertension due to lung diseases or hypoxemia

- COPD
- Interstitial lung disease (ILD)
- Restrictive or obstructive disorders

Group 4: Chronic thromboembolic pulmonary hypertension *(CTEPH)*

Group 5: Pulmonary hypertension with clear multifactorial mechanisms

- Myeloproliferative disorders
- Sarcoidosis, lymphatic vasculitis
- Glycogen storage diseases

Clasificación de hipertensión ARTERIAL pulmonar

afección de arterias de menos de 500 micrones

- **1 Idiopática**
- **1.2 Heredables**
  - **1.2.1 BMPR2b receptor tipo2 proteína morfogenetica**
  - **1.2.2 ALK1, gen kinasa 1 like**
  - **1.2.3 hereditaria desconocida**
- **1.3 Inducida por drogas y toxinas**
- **1.4 Asociadas**
  - **1.4.1 Enfermedades del tejido conectivo**
  - **1.4.2 HIV i**
  - **1.4.3 hipertensión portal**
  - **1.4.4 cardiopatías congénitas**
  - **1.4.5 Schistosomiasis**
  - **1.4.6 anemia hemolítica crónica**
- **1.5 HAP persistente del recién nacido**

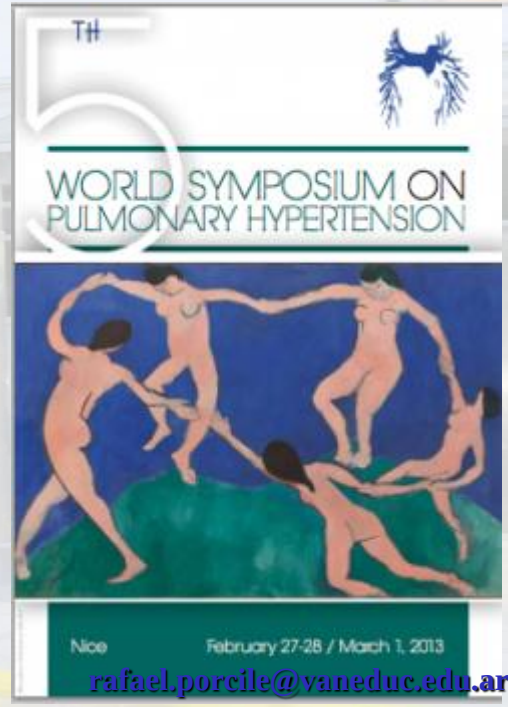

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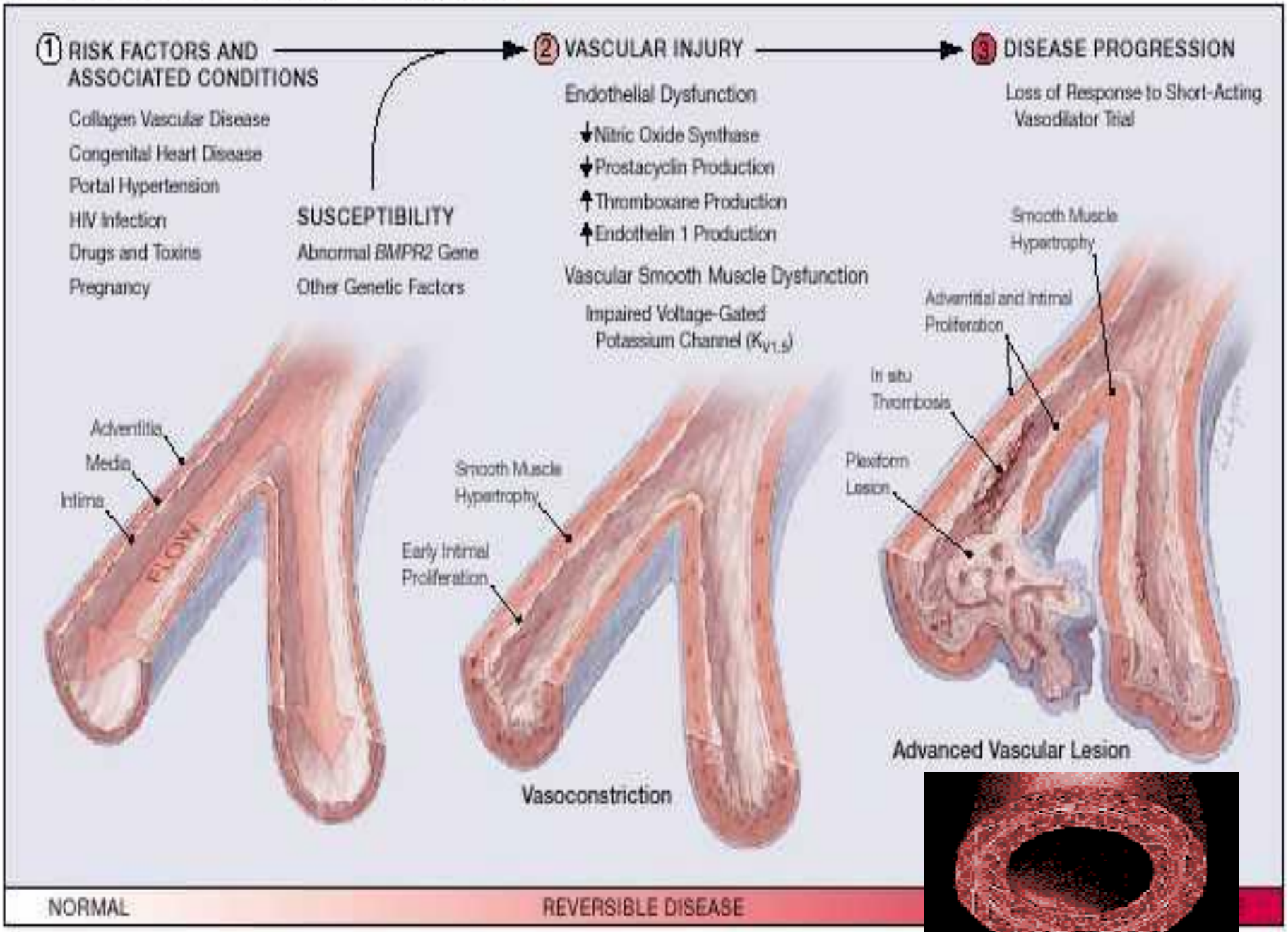
Figure 3. Pathogenesis of Pulmonary Arterial Hypertension



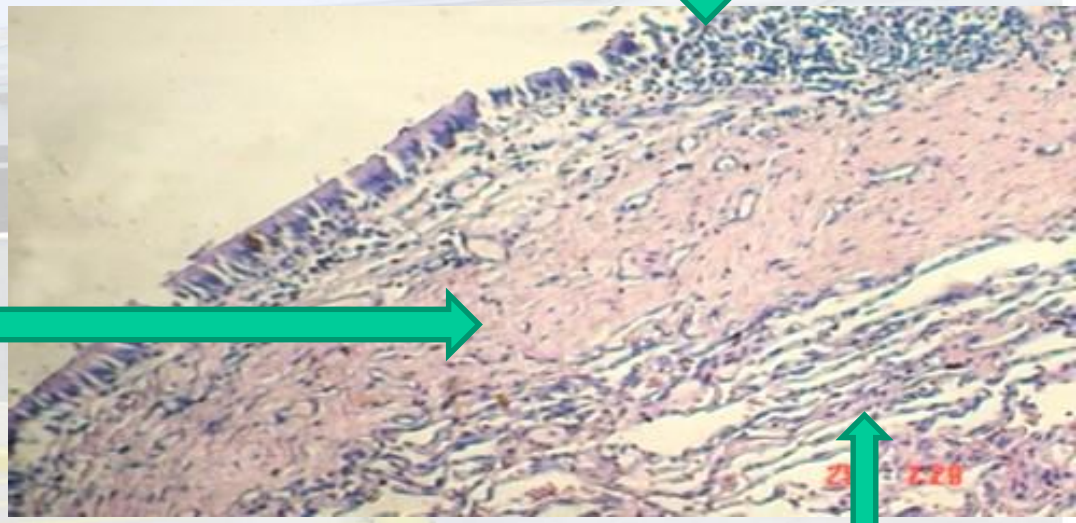
Pulmonary arterial hypertension occurs in susceptible patients as a result of an insult to the pulmonary vascular bed resul… the characteristic pathological features. HIV indicates human immunodeficiency virus; *BMPR2*, bone morphogenetic protein receptor II gene.

Hipertensión ARTERIAL Pulmonar

PAH: menores desde 500 micrones d y

Inflamación peri vascular plexiforme .

hipertrofia de la media



la intima con proliferación fibroblastica

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EVALUACIÓN PRONOSTICA

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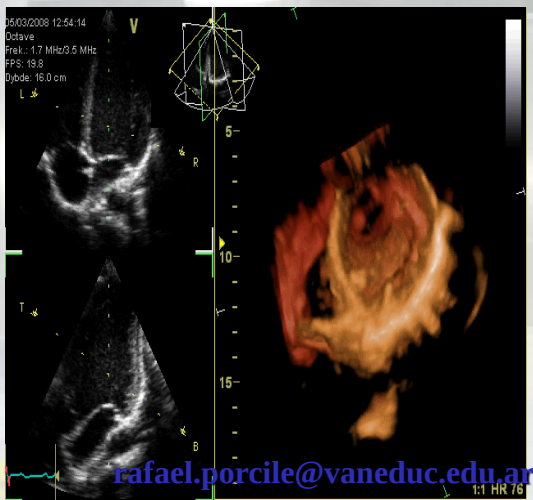
Clasificación funcional según New York Heart Association de acuerdo a O.M.S: 199876



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Class I .Hipertensión pulmonar **sin** síntomas de esfuerzo ni de reposo

Class II Sin síntomas de reposo , leve limitación al esfuerzo

Class III Sin síntomas de reposo Marcada limitación de la actividad física.

Class IV sintomáticos en reposo

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SOBREVIDA

WHO-FC IV, 2.5 AÑOS
WHO-FC III, 6 AÑOS
WHO-FC I and II. 8 AÑOS

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Seis pacientes mas fueron tratados con ambrisentan por hipertensión arterial pulmonar del 2009 a la fecha todo perteneciente al grupo UAI salud o circuitos de seguridad social atendidos por nuestro sistema

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6,7 años de seguimiento promedio

Los resultados globales de los 11 pacientes son muy similares.

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SOBREVIDA

O.M.S: 199876

Expectativa de vida según clase funcional

WHO-FC IV, 2.5 AÑOS

WHO-FC III, 6 AÑOS

WHO-FC I and II. 8 AÑOS

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PRONOSTICO

Cardiopatías congénitas

Idiopática

Secundaria a colagenopatías

Enfermedad veno oclusiva

MORTALIDAD

SOBREVIDA

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Biomarcadores de mal pronostico

- *NT-proBNP* mayor de 1400 pg/ml
- BNP mayor a 150 pg/ml
- La **proteína ligadora de ácidos grasos** específica del corazón: falla derecha
- Dímero D +factor de Von Willebrand marcadores de disfunción endotelial

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Better prognosis	Determinants of prognosis	Worse prognosis
No	Clinical evidence of RV failure	Yes
Slow	Rate of progression of symptoms	Rapid
No	Syncope	Yes
I, II	WHO-FC	IV
Longer (>500 m)[a]	6MWT	Shorter (<300 m)
Peak O_2 consumption >15 mL/min/kg	Cardio-pulmonary exercise testing	Peak O_2 consumption <12 mL/min/kg
Normal or near-normal	BNP/NT-proBNP plasma levels	Very elevated and rising
No pericardial effusion TAPSE[b] >2.0 cm	Echocardiographic findings[b]	Pericardial effusion TAPSE[b] <1.5 cm
RAP <8 mmHg and CI $\geq$2.5 L/min/m^2	Haemodynamics	RAP >15 mmHg or CI $\leq$2.0 L/min/m^2

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The REVEAL Registry risk score calculator in patients newly diagnosed with pulmonary arterial hypertension.

Chest. 2012 Feb;141(2):354-62. doi: 10.1378/chest.11-0676. Epub 2011 Jun 16.

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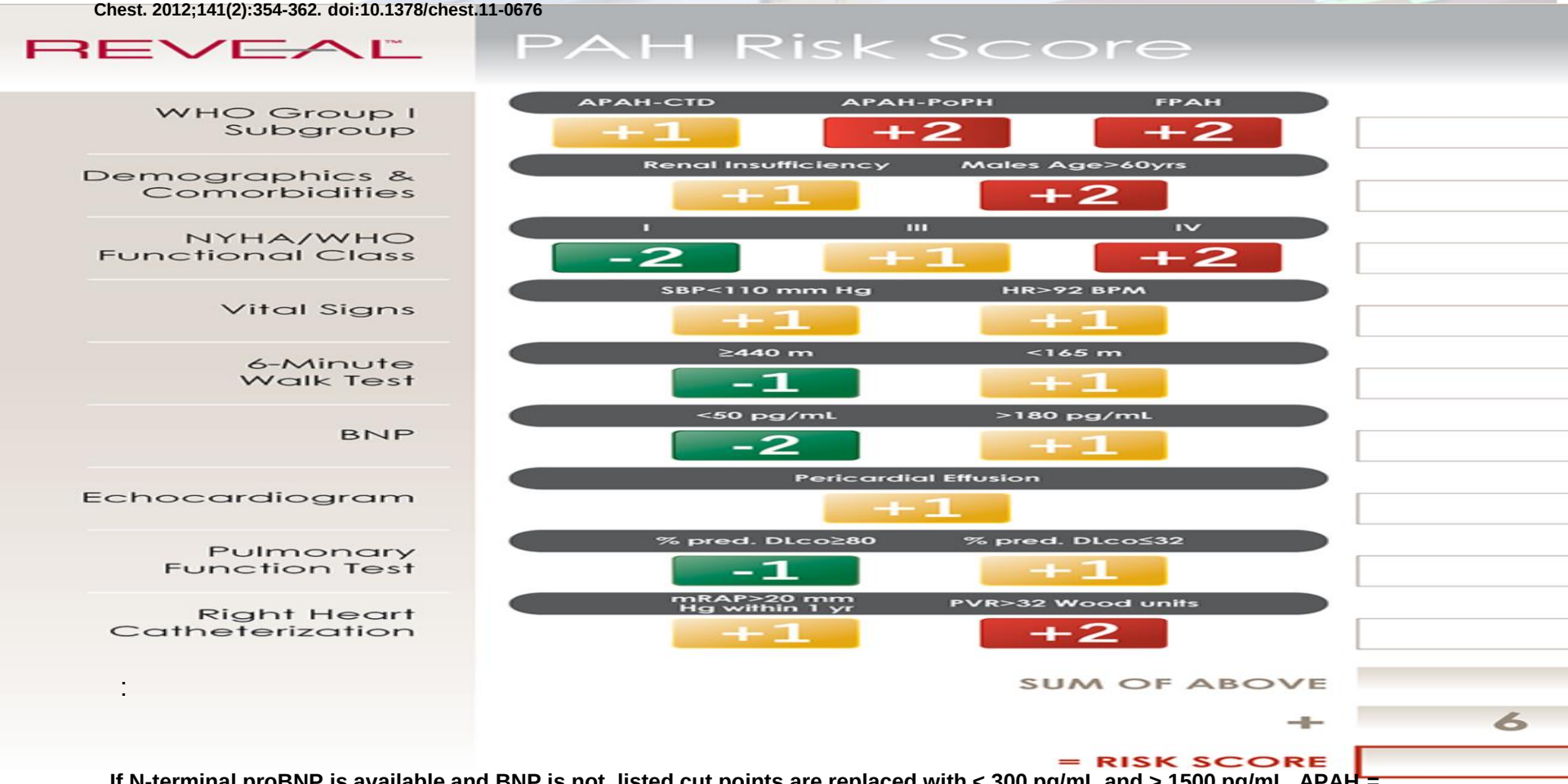
From: The REVEAL Registry Risk Score Calculator in Patients Newly Diagnosed With Pulmonary Arterial HypertensionValidation of the REVEAL Registry Risk Calculator

Chest. 2012;141(2):354-362. doi:10.1378/chest.11-0676



If N-terminal proBNP is available and BNP is not, listed cut points are replaced with < 300 pg/mL and > 1500 pg/mL. APAH = associated pulmonary arterial hypertension; BNP = brain natriuretic peptide; BPM = beats per minute; CTD = connective tissue disease; DLco = diffusing capacity of lung for carbon monoxide; FPAH = familial pulmonary arterial hypertension; HR = heart rate; mRAP = mean right atrial pressure; NYHA = New York Heart Association; PAH = pulmonary arterial hypertension; PoPH = portopulmonary hypertension; PVR = pulmonary vascular resistance; REVEAL Registry = Registry to Evaluate Early and Long-term Pulmonary Arterial Hypertension Disease Management; SBP = systolic BP; WHO = World Health Organization.

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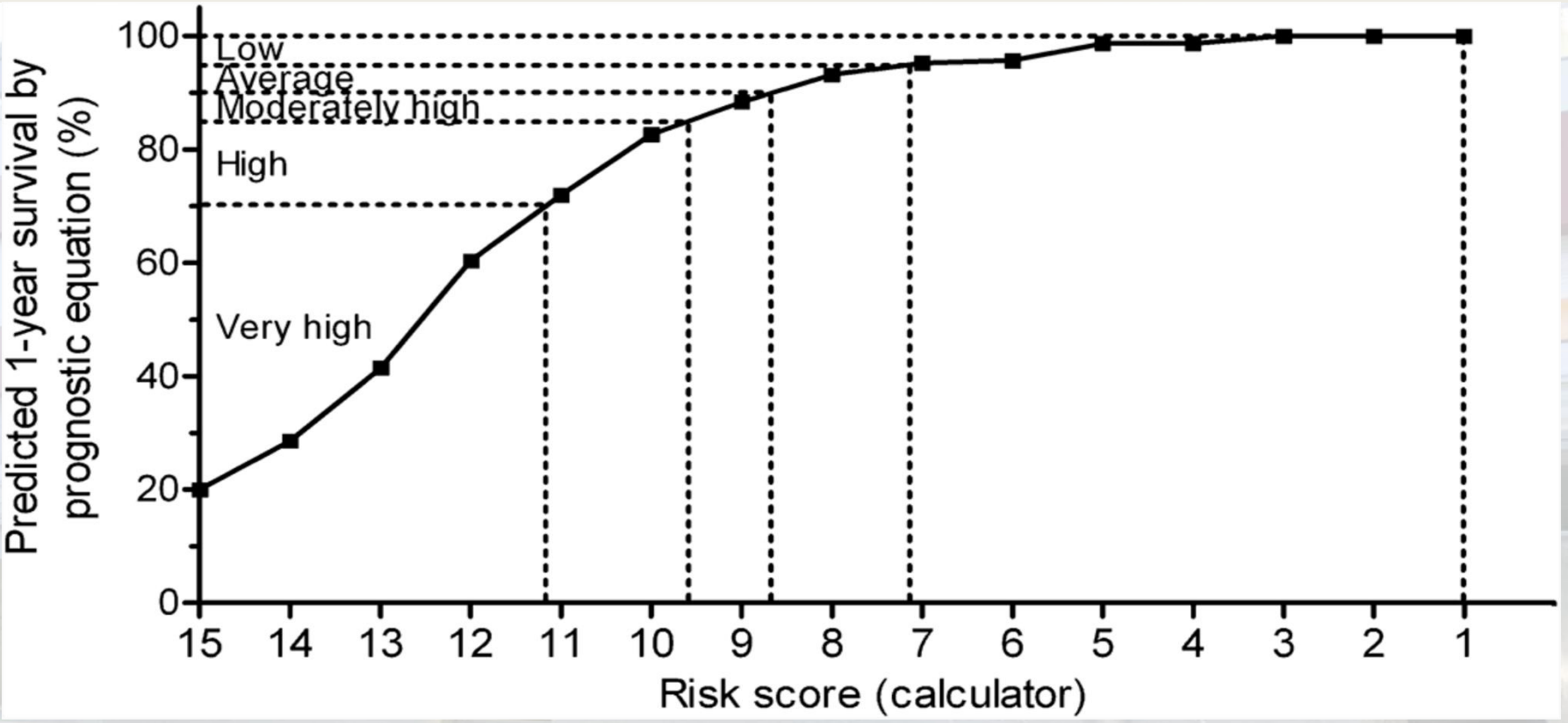

Twelve-month Kaplan-Meier survival estimate for the REVEAL Registry development cohort with predicted risk score. Risk strata are indicated by the lines: predicted 1-year survival is 95% to 100% in the low-risk group, 90% to < 95% in the average-risk group, 85% to < 90% in the moderately high-risk group, 70% to < 85% in the high-risk group, and < 70% in the very high-risk group. See Figure 1 legend for expansion of abbreviation.

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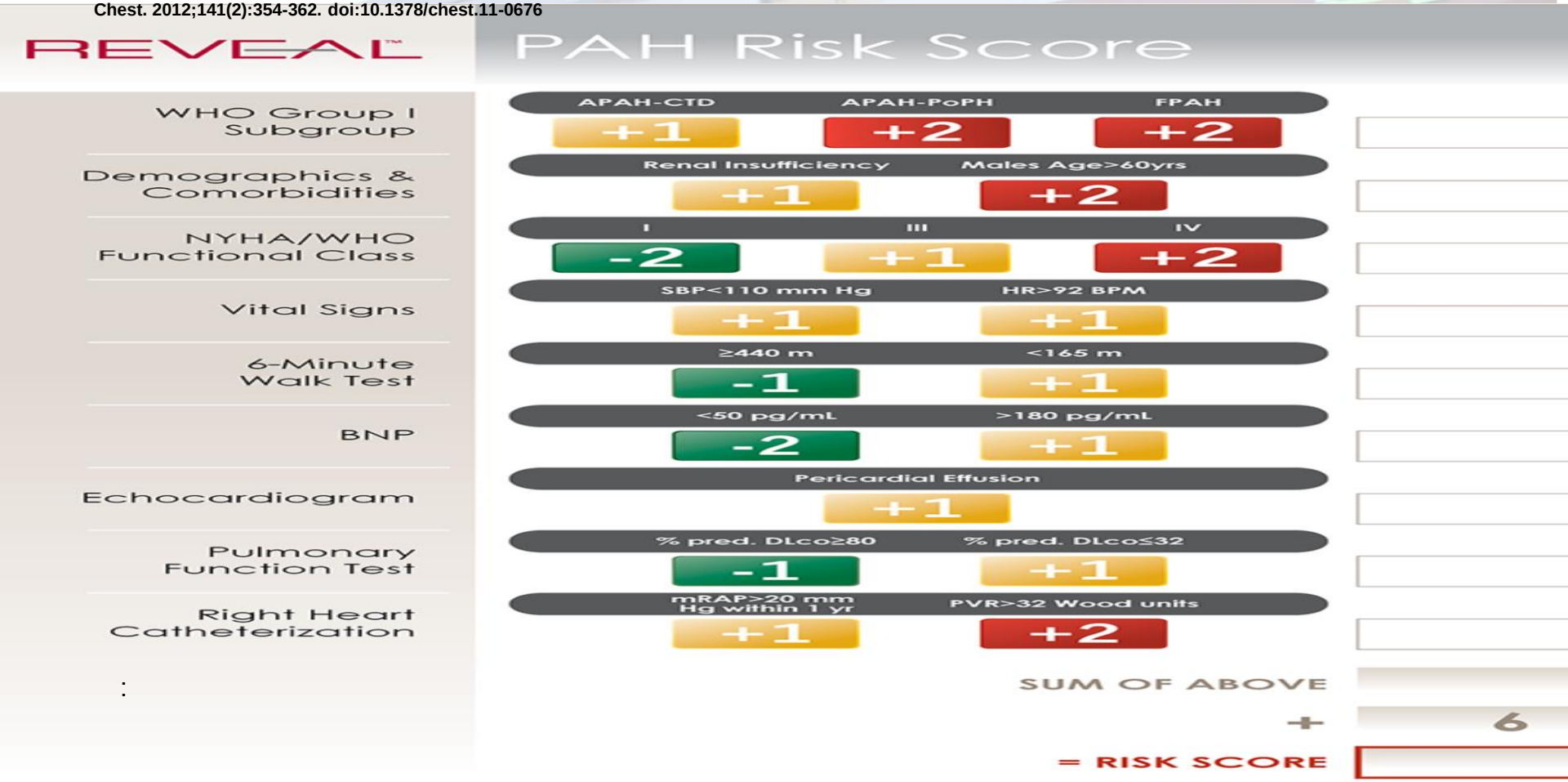

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Nuestra experiecia

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6,7 años de seguimiento promedio

Los resultados globales de los 11 pacientes son muy similares.

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¿Que herramientas utilizar
Para una evaluación ESTIMATIVA
De la evolución de esta pequeña población ?

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5 herramientas de estimación

- Calculador de riesgo del REVEAL
- Evaluación de corte de la evolución clínica a los 36 meses
- Curva Kaplan Meier del REVEAL
- Mortalidad prevista según test de caminata inicial
- Mortalidad prevista según clase funcional OMS.

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Reflexiones…

A los **12 meses** de

seguiminto

AÑOS
2005/2006

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2004-2005

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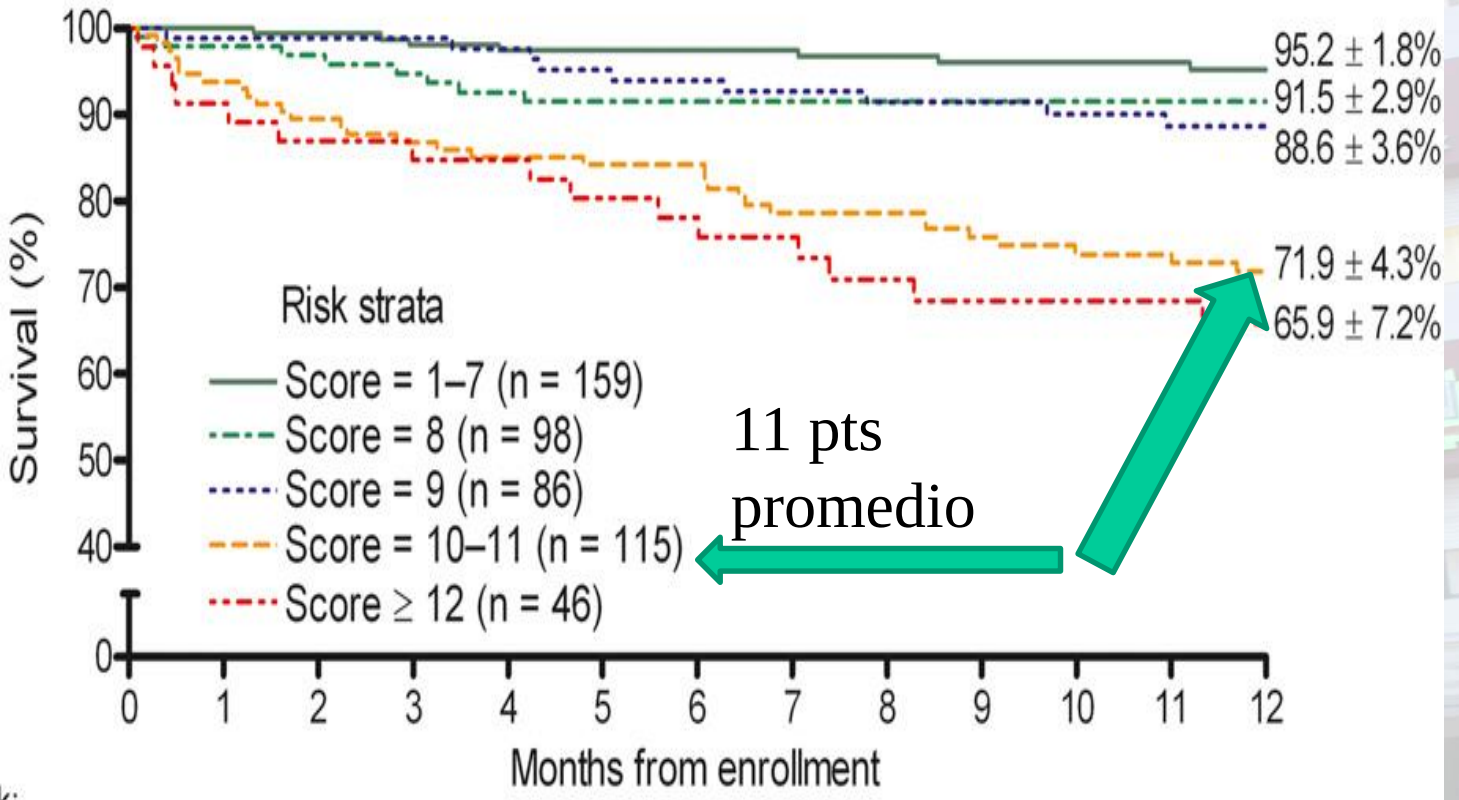
B

Risk calculator



11 pts promedio

No. at risk:													
Score = 1–7	159	156	155	151	150	150	150	141	140	139	120	120	119
Score = 8	98	93	91	89	87	86	86	84	81	81	71	71	71
Score = 9	86	84	84	81	80	78	77	73	72	72	65	64	64
Score = 10–11	115	107	102	99	96	95	95	85	85	82	74	74	72
Score ≥ 12	46	42	40	38	38	36	35	31	29	28	26	26	25

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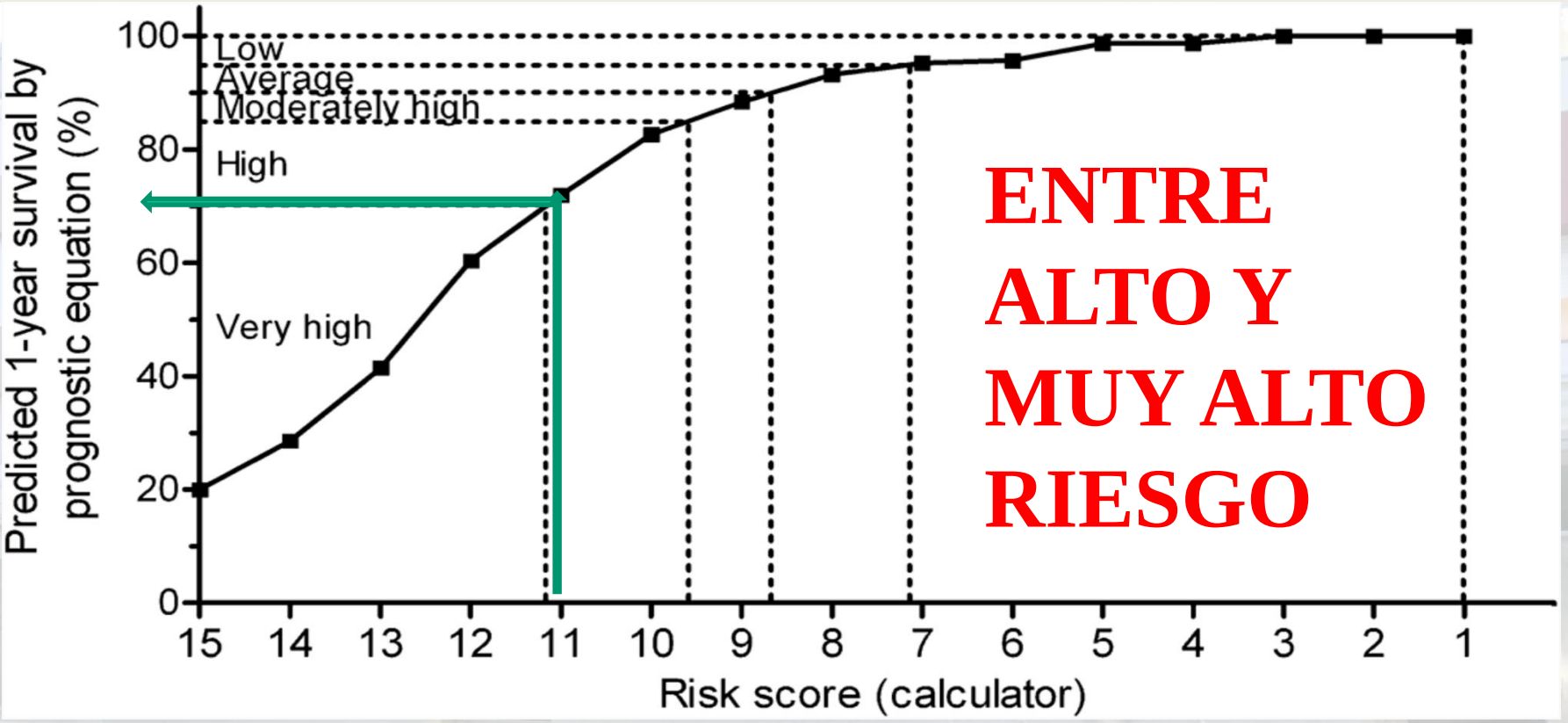

Twelve-month Kaplan-Meier survival estimate for the REVEAL Registry development cohort with predicted risk score. Risk strata are indicated by the lines: predicted 1-year survival is 95% to 100% in the low-risk group, 90% to < 95% in the average-risk group, 85% to < 90% in the moderately high-risk group, 70% to < 85% in the high-risk group, and < 70% in the very high-risk group. See Figure 1 legend for expansion of abbreviation.

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Reflexiones… No Conclusiones

- Sobrevida esperada promedio de los 5 pacientes al año subestimada por no ponderar BNP según REVEAL **71.9.%**

- **Sobrevida UAI observada al**
  - **1 año 100 %**

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Reflexiones…

A los

Tres años de

seguimiento

AÑOS
2007/2008

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EVOLUCIÓN CLINICA

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- **Buena tolerancia al fármaco sin hepatotoxicidad**
- **Caída del test de caminata respecto del mejor histórico en todos los casos mayor al 6-11 % que mejora al agregarle sildenafil a partir del ingreso a Aries Ext.**
- **Reducción de la presión arterial pulmonar a expensas predominantemente de la presión sistólicas**

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Reflexiones…

A los **4 años** de seguimiento

AÑOS
2008/2009

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Survival estimates of patients in REVEAL using Kaplan–Meier estimates stratified by newly versus previously diagnosed patients and survival estimated by a delayed entry model accounting for truncation

M.D. McGoon, and D.P. Miller
Eur Respir Rev 2012;21:8-18

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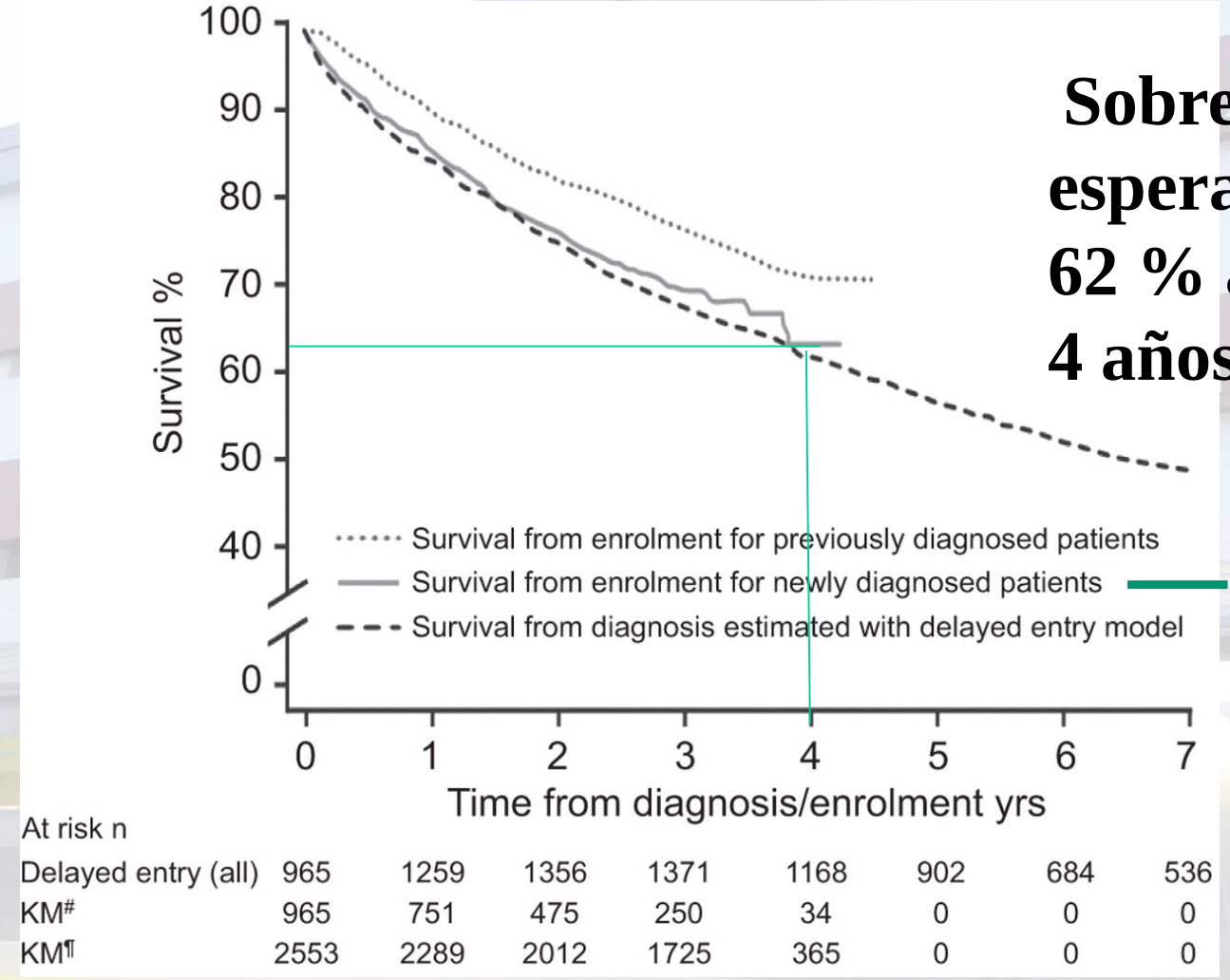
Survival estimates of patients in REVEAL using Kaplan–Meier estimates stratified by newly versus previously diagnosed patients and survival estimated by a delayed entry model accounting for truncation.



At risk n								
Delayed entry (all)	965	1259	1356	1371	1168	902	684	536
KM#	965	751	475	250	34	0	0	0
KM¶	2553	2289	2012	1725	365	0	0	0

Sobrevida esperada 62 % a los 4 años

M.D. McGoon, and D.P. Miller Eur Respir Rev 2012;21:8-18

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Reflexiones… No Conclusiones

- Sobrevida esperada promedio a los 4 años

62%

- Sobrevida UAI observada al
  - 1 año 100 %
  - 2 años 80%
  - 3 años 80 %
  - **4 años 80%**
  - 5 años 60 %
  - 11 años 60%

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Reflexiones…

A los **6 años** de seguimiento

AÑOS
2010/2011

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UAI
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UNIVERSITARIO

SOBREVIDA

O.M.S: 199876

Expectativa de vida según clase funcional

WHO-FC IV, 2.5 AÑOS

WHO-FC III, 6 AÑOS

WHO-FC I and II. 8 AÑOS

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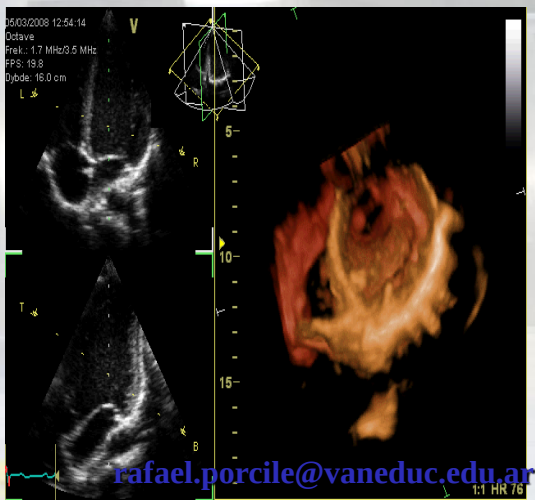
Clasificación funcional según New York Heart Association de acuerdo a O.M.S: 199876

WHO-FC IV, 2.5 AÑOS

WHO-FC III, 6 AÑOS

WHO-FC I and II. 8 AÑOS

Se esperaría que todos los pacientes hubiesen fallecido a los seis años



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Reflexiones… No Conclusiones

- Mortalidad esperada promedio a los 11 años

100%

- Sobrevida observada al
  - 1 año 100 %
  - 2 años 80%
  - 3 años 80 %
  - 4 años 80%
  - 5 años 60 %
  - **11 años 60%**

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FISIOPATOLOGIA DE LA HIPERTENSION PULMONAR

Rafael Porcile

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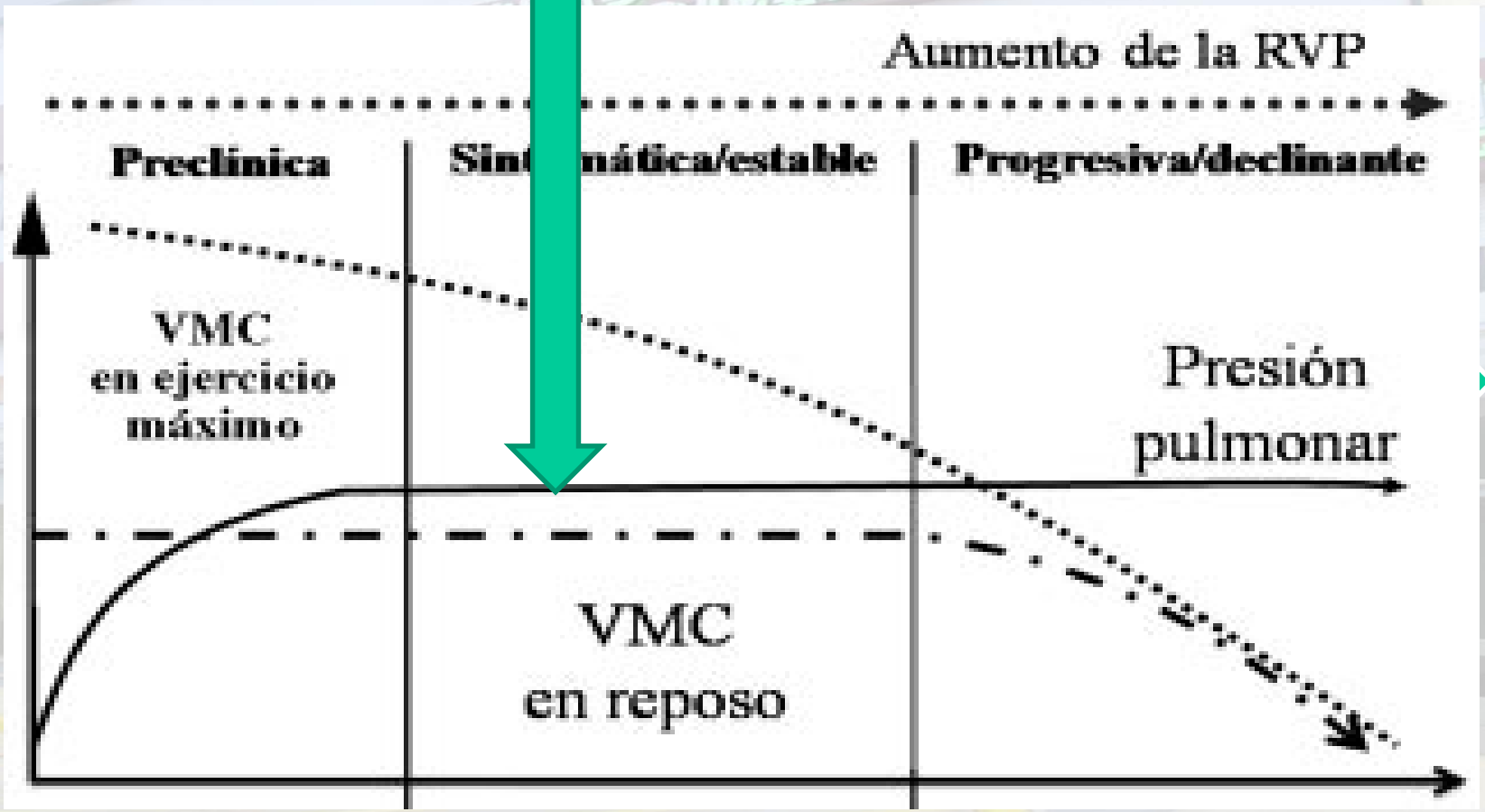
DEPARTAMENTO DE CARDIOLOGIA

CÁTEDRA DE FISIOLOGïA

Universidad Abierta Interamericana

AUMENTO DE LAS RESISTENCIAS CON PRESION CONSTANTE



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HIPERTENSION ARTERIAL PULMONAR

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¿HIPERTENSION PULMONAR ES EL NOMBRE ADECUADO?

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RESISTENCIAS VASCULARES PULMONARES

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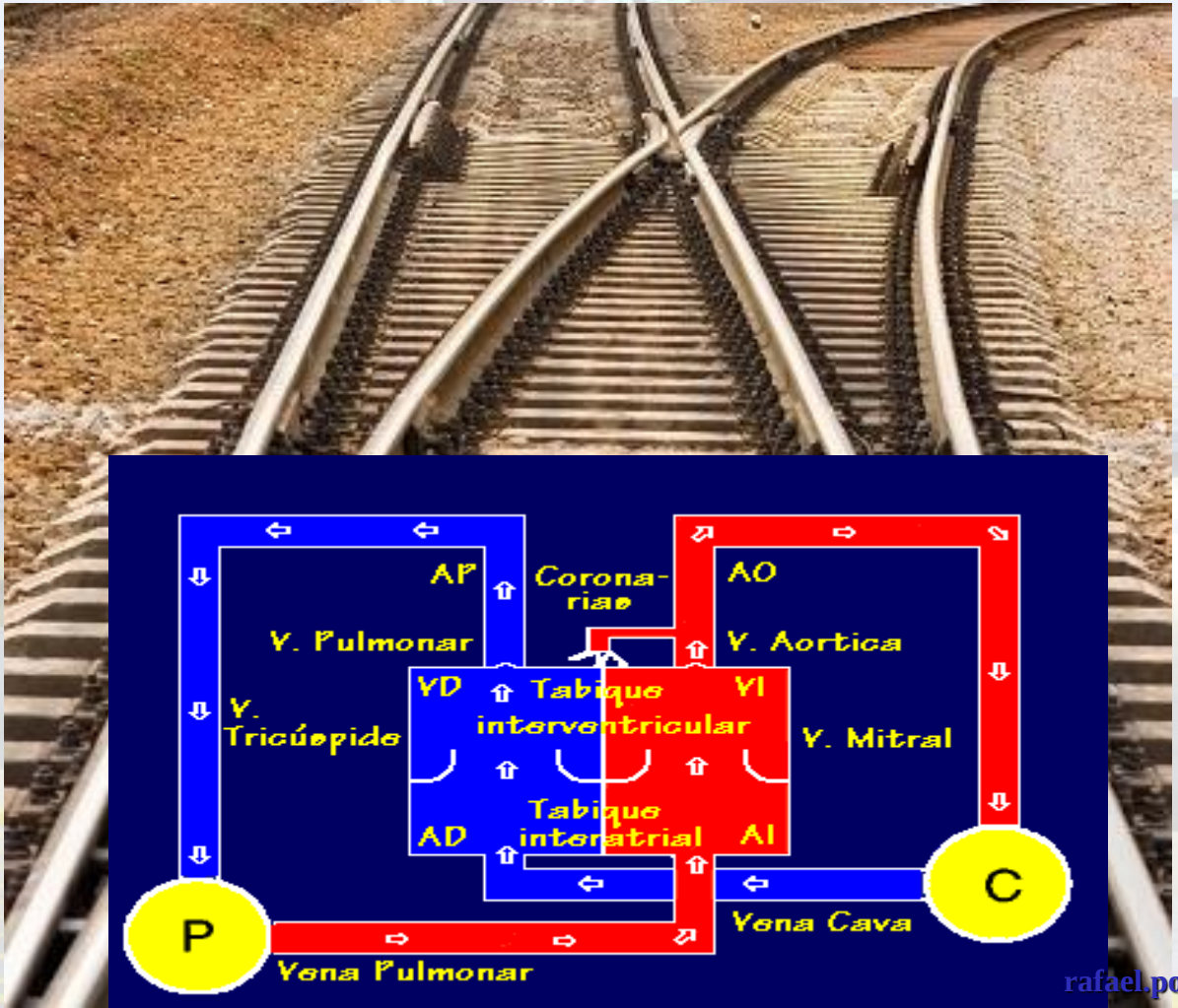
CONFLICTO IZQUIERDA DECECHA



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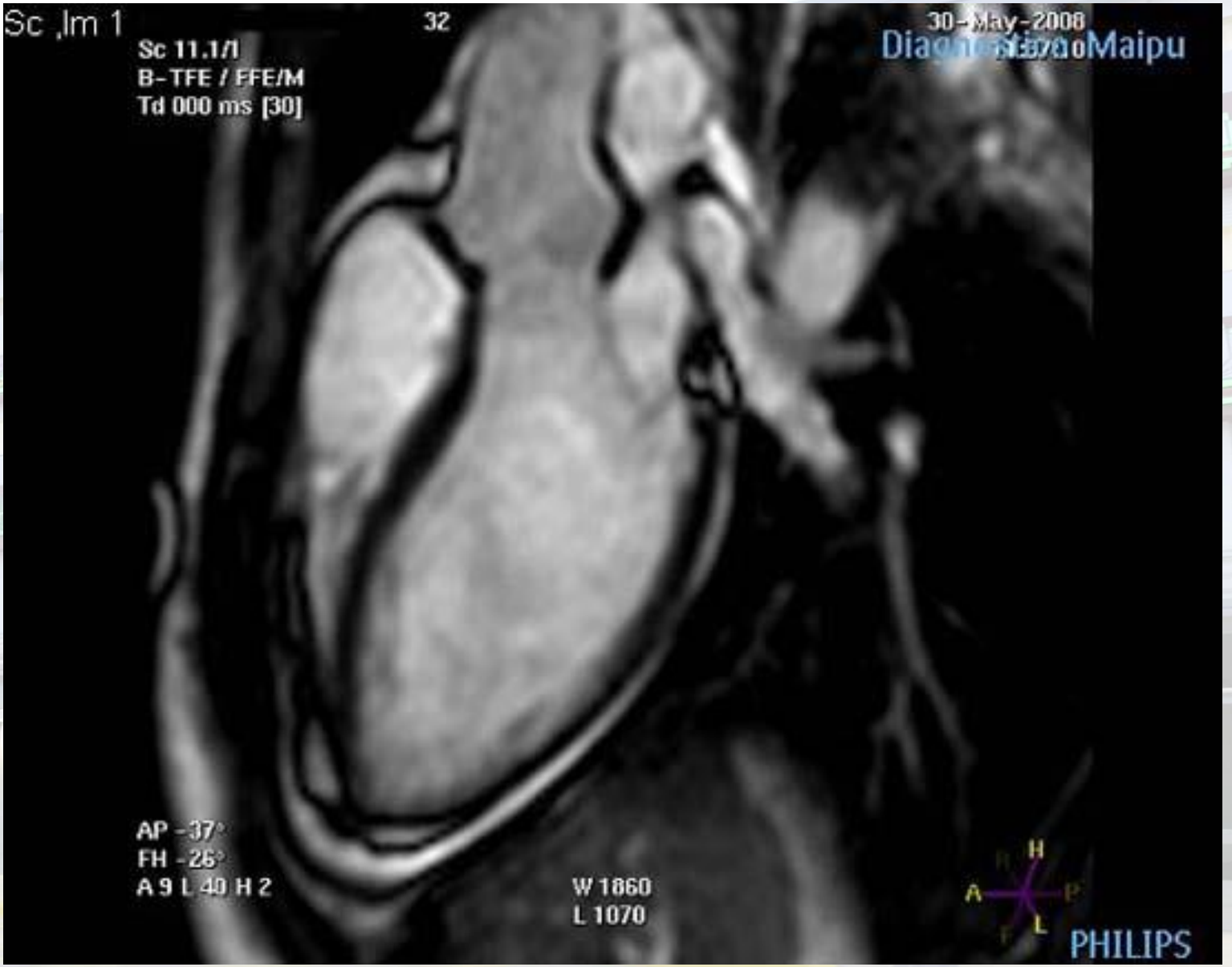

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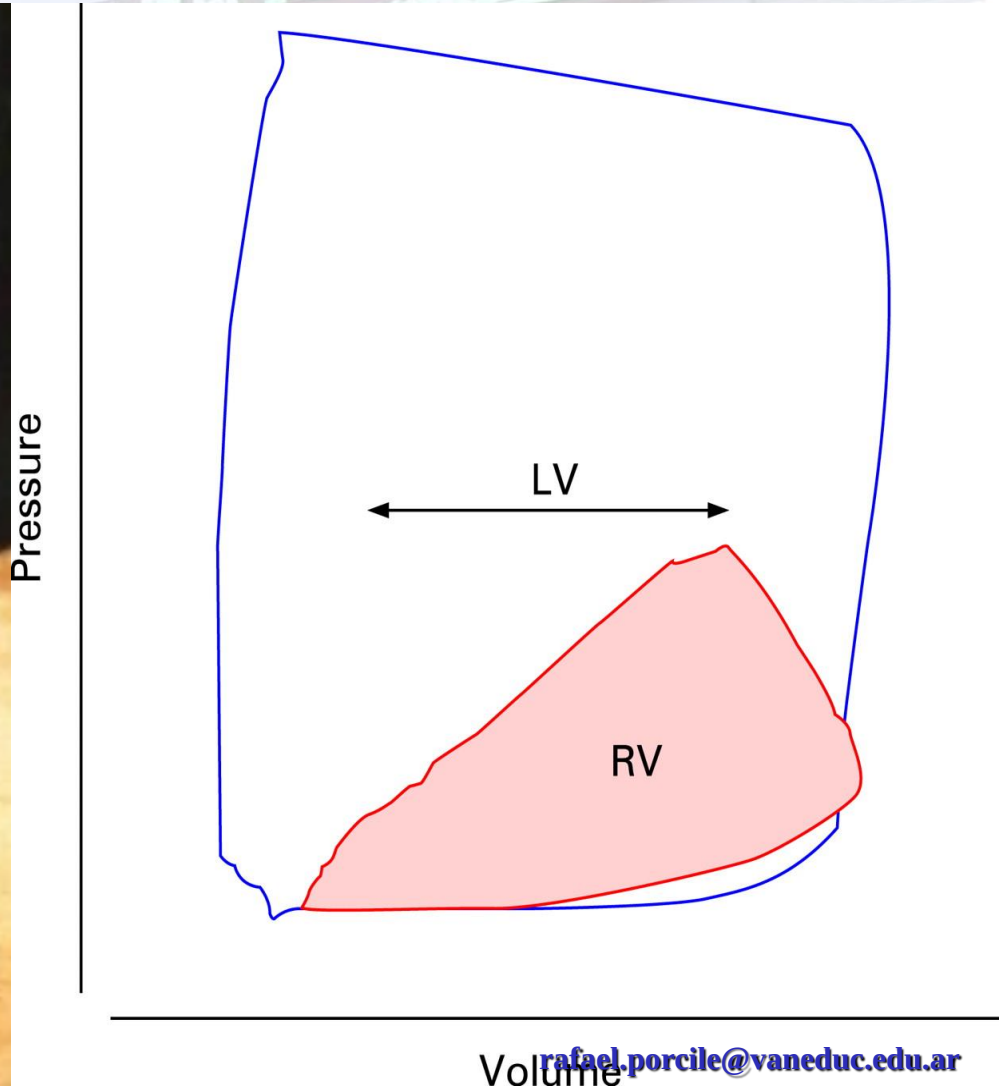
V.D. VS HIPERRESISTENCIA



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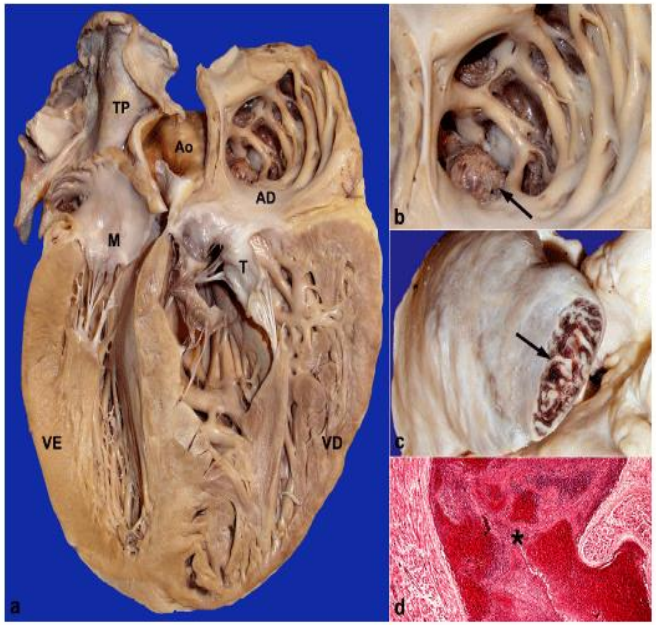
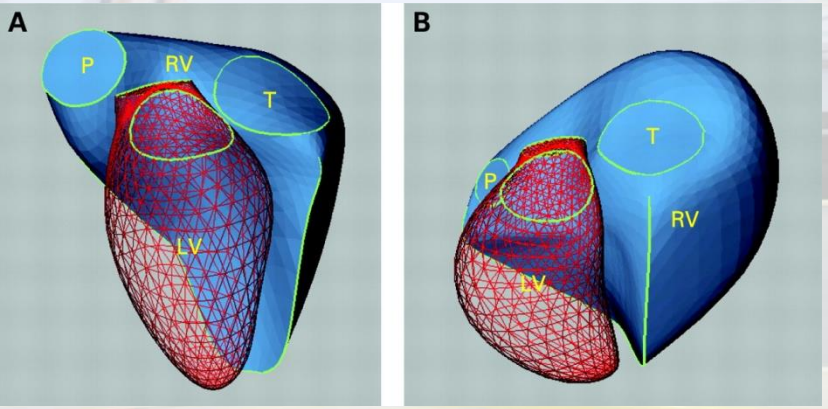
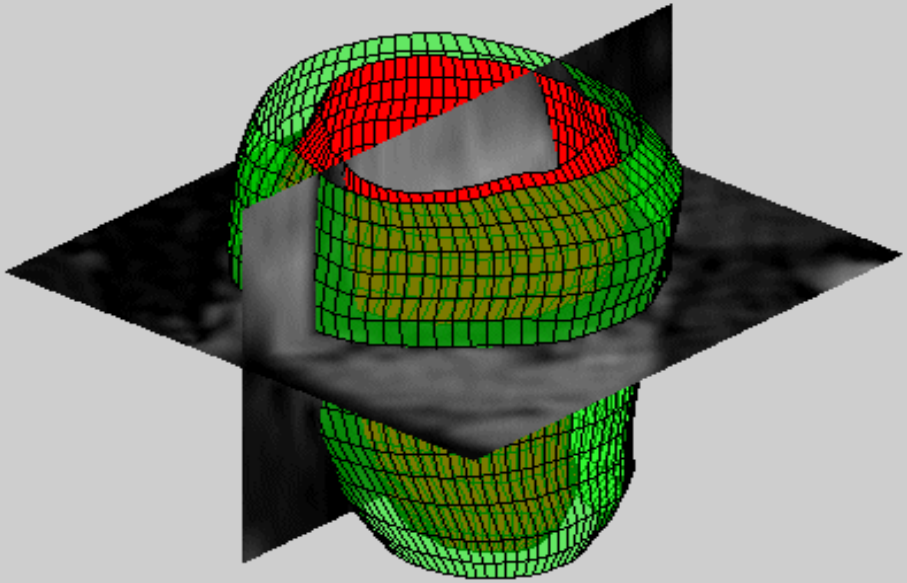


Figura 2 - Imágenes del corazón. En (a) observamos un gran volumen de las cámaras derechas, principalmente del ventrículo derecho (VD) con acentuada hipertrofia de la pared y dilatación cavitaria en relación con el ventrículo izquierdo (VI). El atrio derecho (AD) es mejor observado en la figura (b) que destaca la presencia de trombosis (flecha) traspasando la musculatura pectínea y llenando la aurícula derecha (flecha) cuya punta se cortó en (c). Corte histológico de la aurícula (d) evidencia trombo fibrinohemático (*) llenando la cavidad auricular (Hematoxilina & eosina; objetiva de 5X). TP - tronco pulmonar, Ao - aorta, M - mitral, T - tricúspide.





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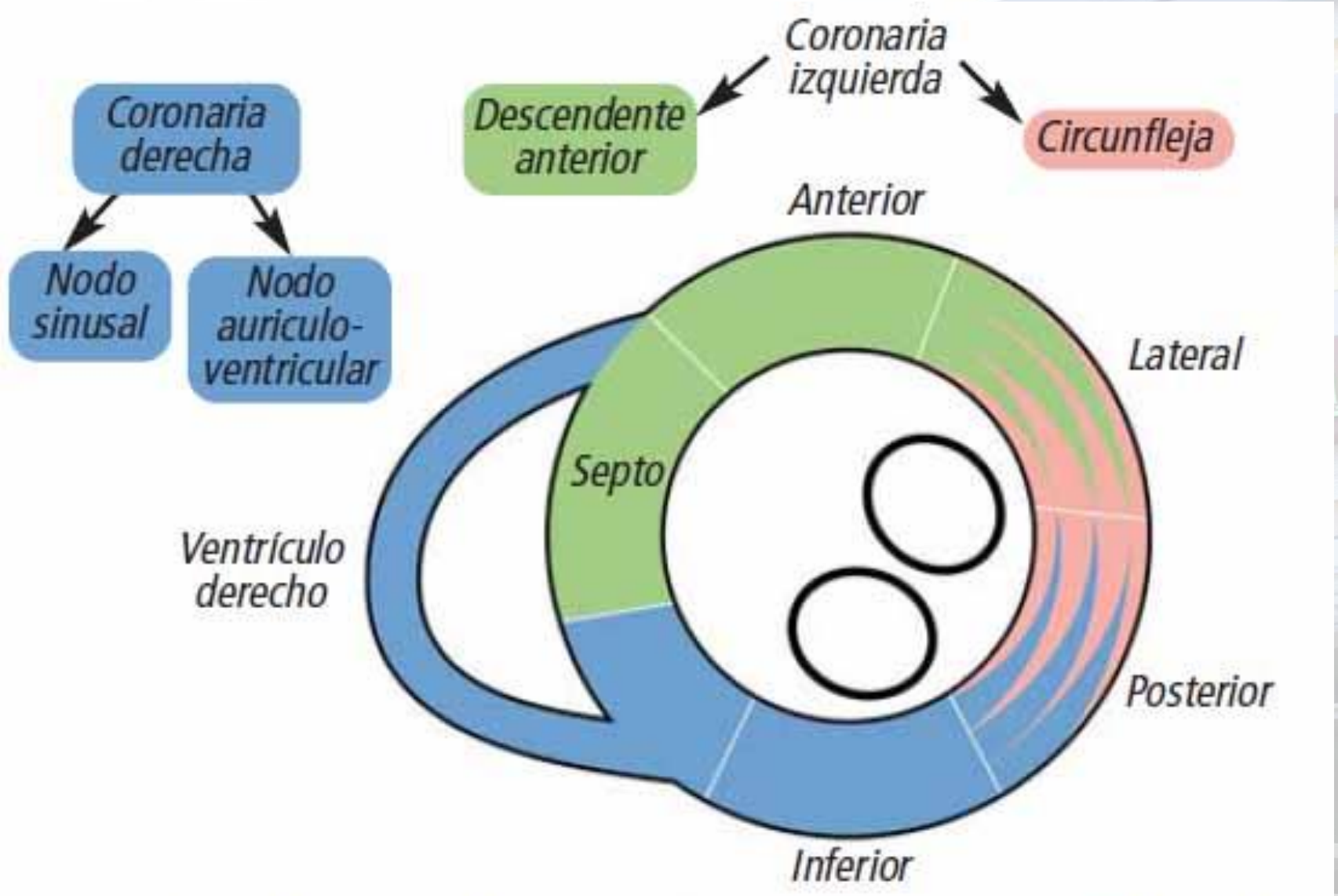

Figura 3. Corte transversal de los ventrículos con las áreas irrigadas por cada arteria coronaria.

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PRECAPILAR

POST CAPILAR

RVP=TAMP - PCP
VM

TAD+DIF/3

DIAMETRO ARTETIOLAR

AUMENTO DEL GRADIENTE TAMP-PCP
REDUCCION PRECARGA IZQUIERDA

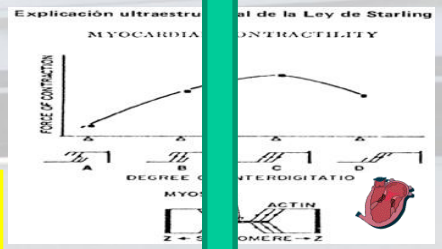
TROMBOSIS LOCAL

REMODELACION VASCULAR PULMONAR

EMBOLIA PULMONAR
DISNEA SUBITA

DISNEA PROGRESIVA

CAIDA DE LA DESCARGA SISTOLICA DEL VENTRICULO IZQUIERDO



FALLA PER FUSIÓN ANTERO GRADA

SINCOPE

AUMENTO DE POSTCARGA DERECHA
CONGESTION VENOSA
FALLA VENTRICULAR DERECHA CLINICA

HIPOTERFUSION MIOCARDICA Y HEPATICA

ASTENIA

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Right Ventricular Function Predicts Clinical Response to Specific Vasodilator Therapy in Patients with Pulmonary Hypertension.

Echocardiography. 2012 Sep 18.
doi: 10.1111

Over an average period of 1 year, almost half of patients showed signs of clinical deterioration despite specific vasodilator therapy. Parameters of right ventricular morphology and function had prognostic value in these patients.

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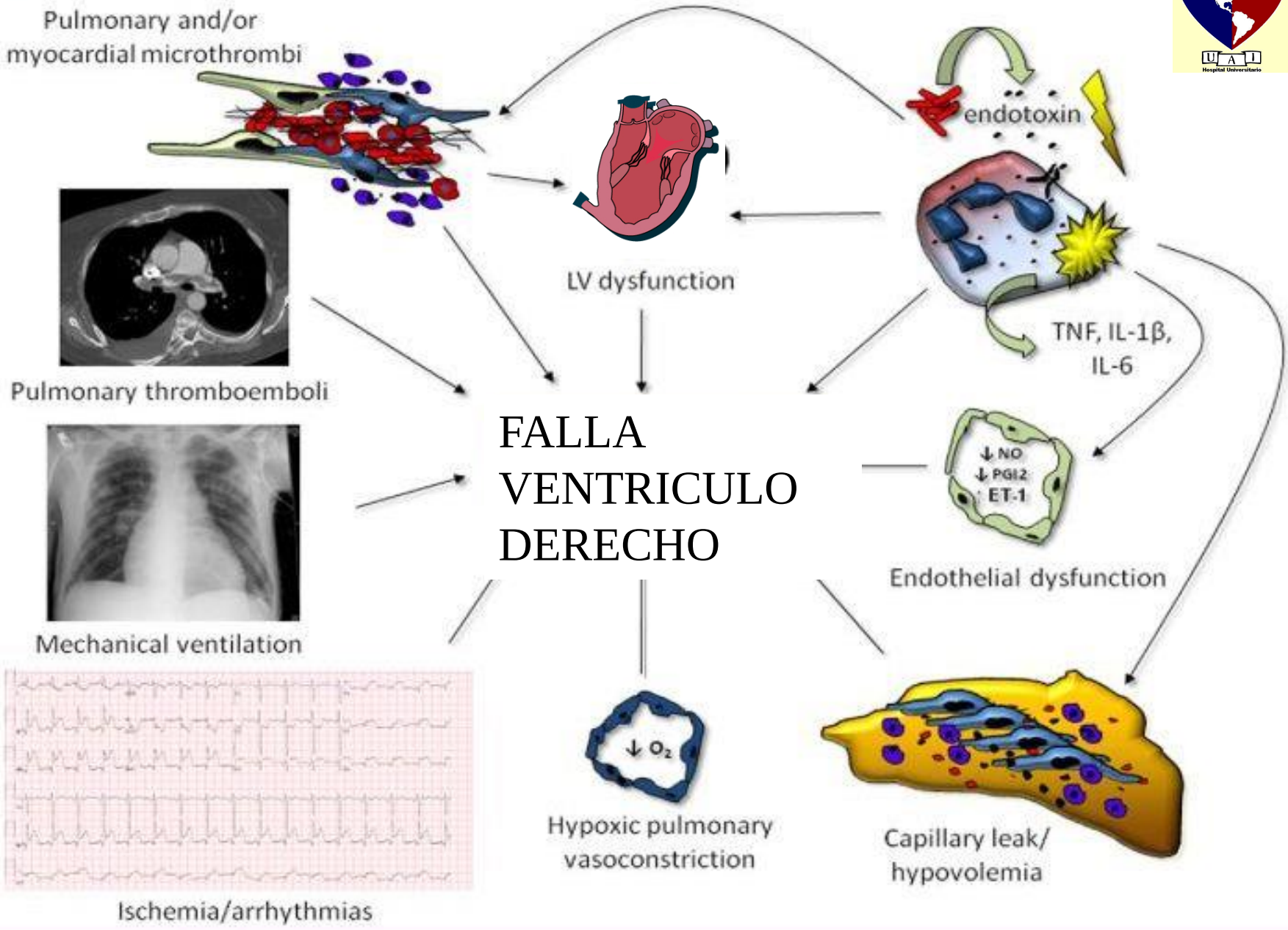

UAI
HOSPITAL UNIVERSITARIO

HIPERTENSION PULMONAR

DIAGNOSTICO

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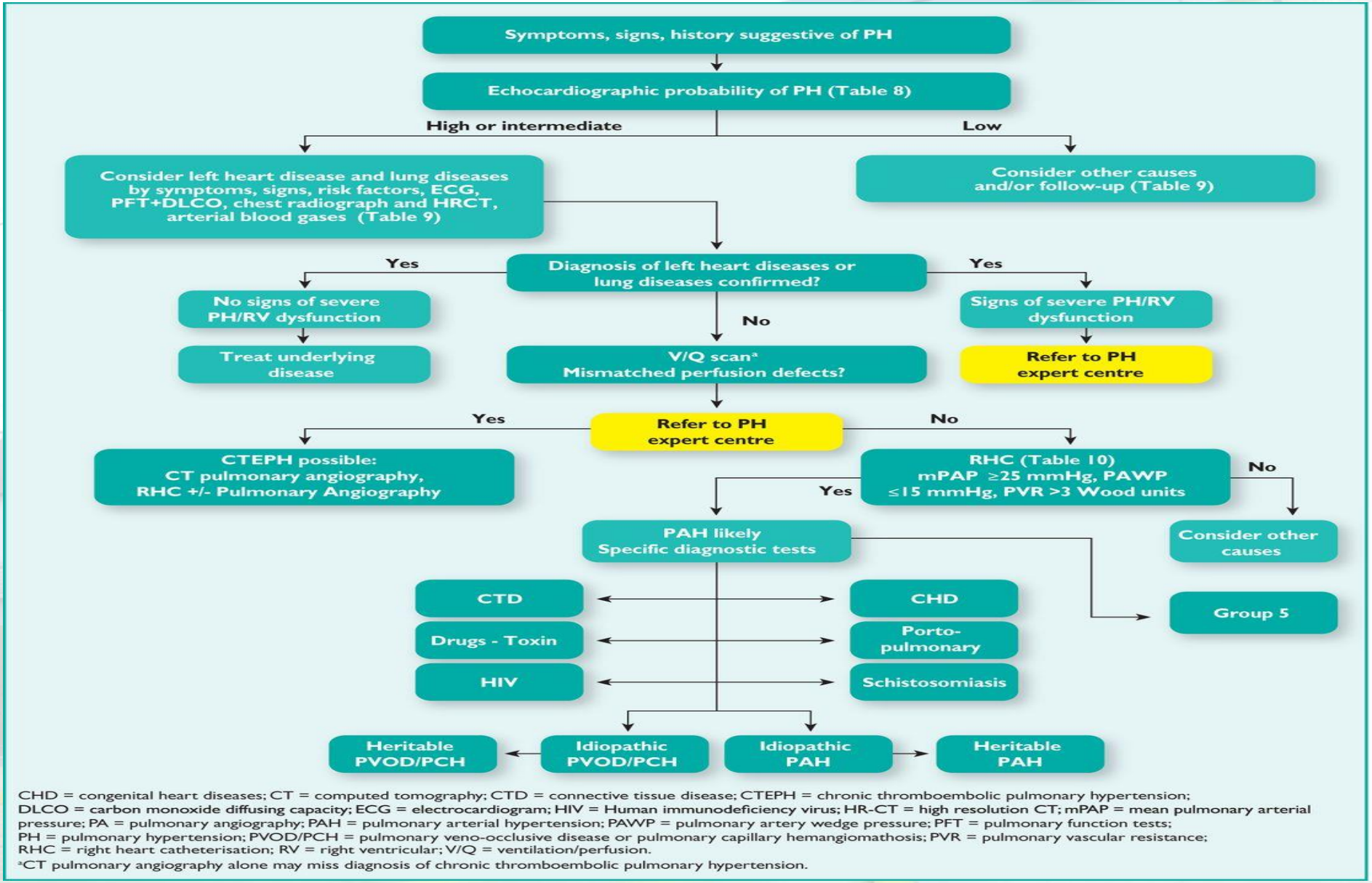
CÁTEDRA DE FISIOLOGïA

Universidad Abierta Interamericana

Diagnostic algorithm.



CHD = congenital heart diseases; CT = computed tomography; CTD = connective tissue disease; CTEPH = chronic thromboembolic pulmonary hypertension; DLCO = carbon monoxide diffusing capacity; ECG = electrocardiogram; HIV = Human immunodeficiency virus; HR-CT = high resolution CT; mPAP = mean pulmonary arterial pressure; PA = pulmonary angiography; PAH = pulmonary arterial hypertension; PAWP = pulmonary artery wedge pressure; PFT = pulmonary function tests; PH = pulmonary hypertension; PVOD/PCH = pulmonary veno-occlusive disease or pulmonary capillary hemangiomathosis; PVR = pulmonary vascular resistance; RHC = right heart catheterisation; RV = right ventricular; V/Q = ventilation/perfusion.
[a]CT pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.

Authors/Task Force Members: et al. Eur Heart J 2015; eurheartj.ehv317

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Symptoms/signs/history suggestive of PH

Search for
ther causes and/or
re-check

Group 2: Left

3: Lung diseases
l/or hypoxia?

Y
PH "proportion

Yes
proportion" PH

Treat under
and check fo

Search for
her causes

Consider
CTE

NO

Cons
PVOD

P ≥ 25 mm Hg
≤ 15 mm Hg

PVOD
PCH

Schistosomiasis
Other group 5

ronic
nolysis

Idiopathic or Heritable PAH

BMPR2, ALK1,
Endoglin (HHT)
Family history

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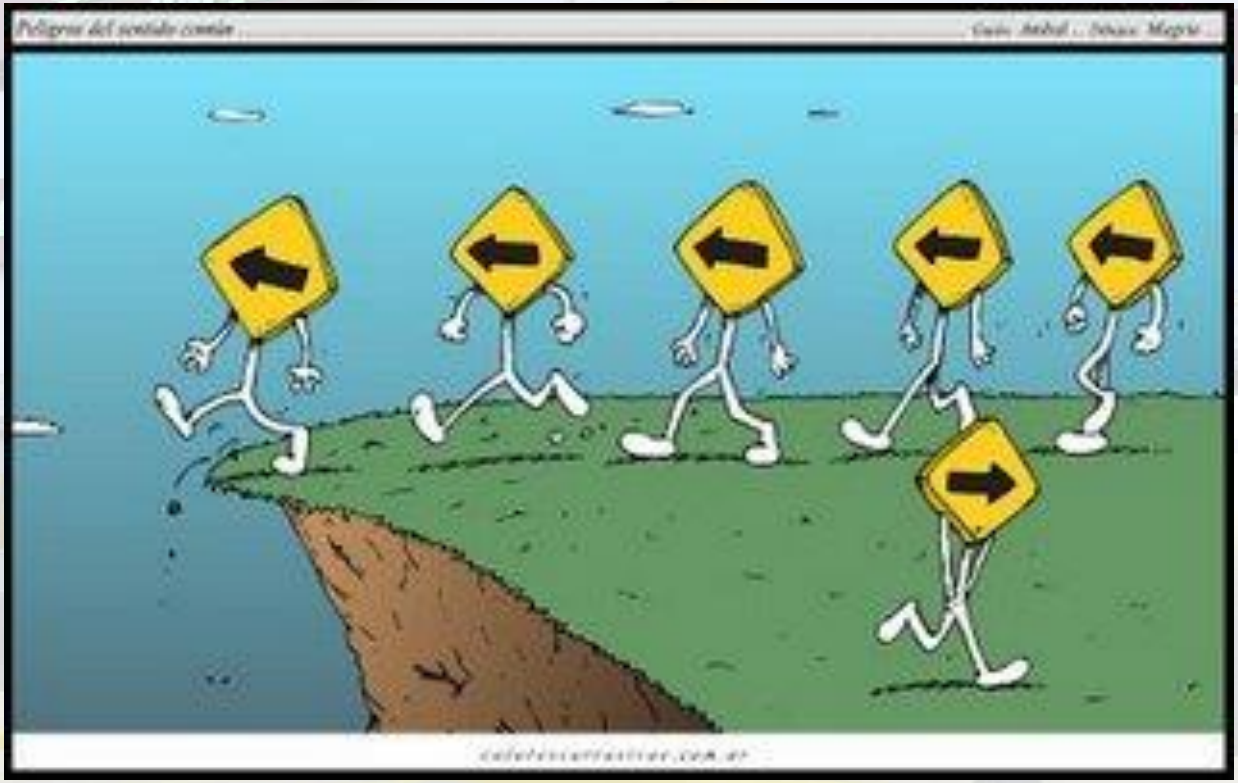
No… solo sentido común



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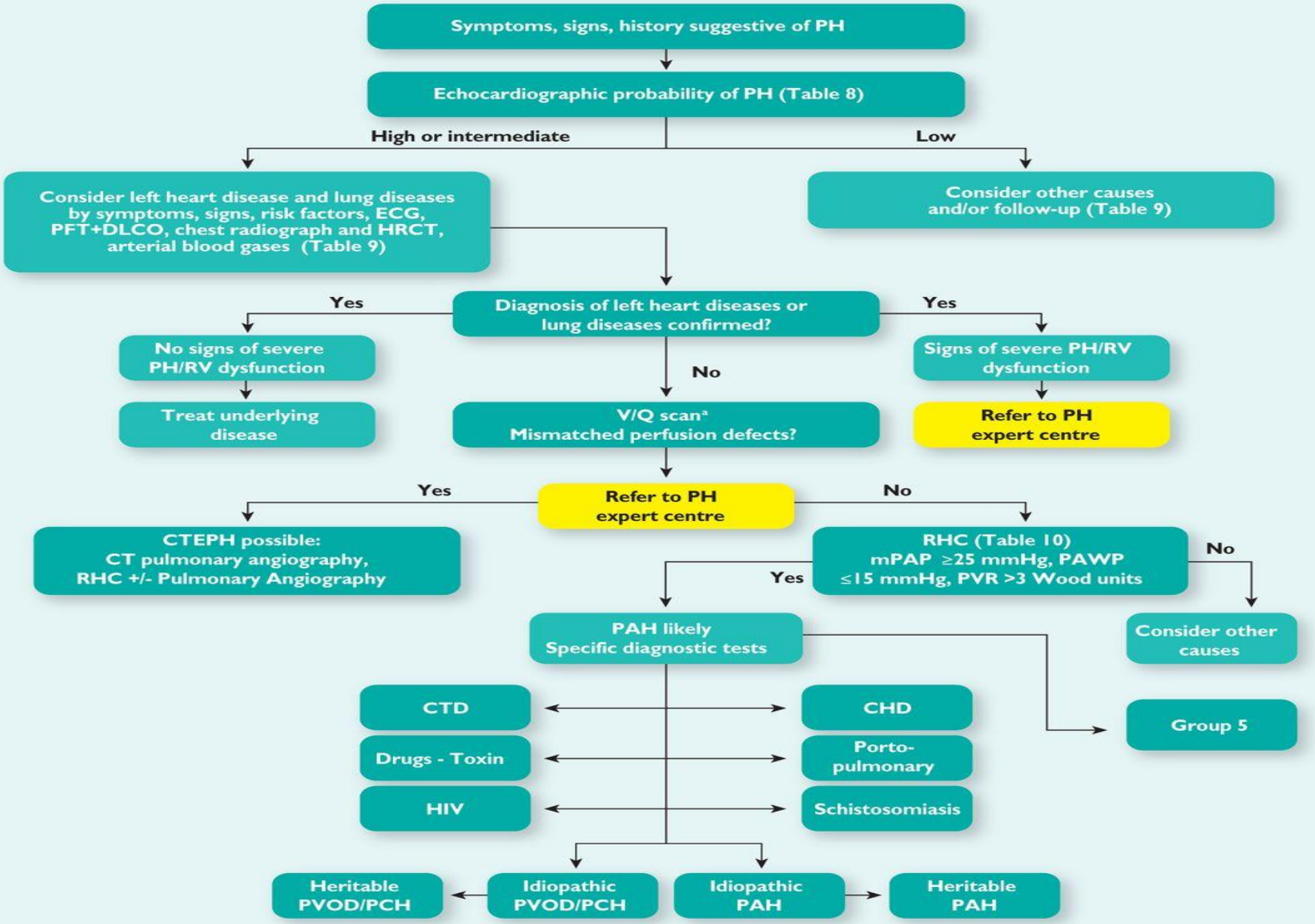

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[a] CT pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.

Authors/Task Force Members: et al. Eur Heart J 2015; eurheartj.ehv317

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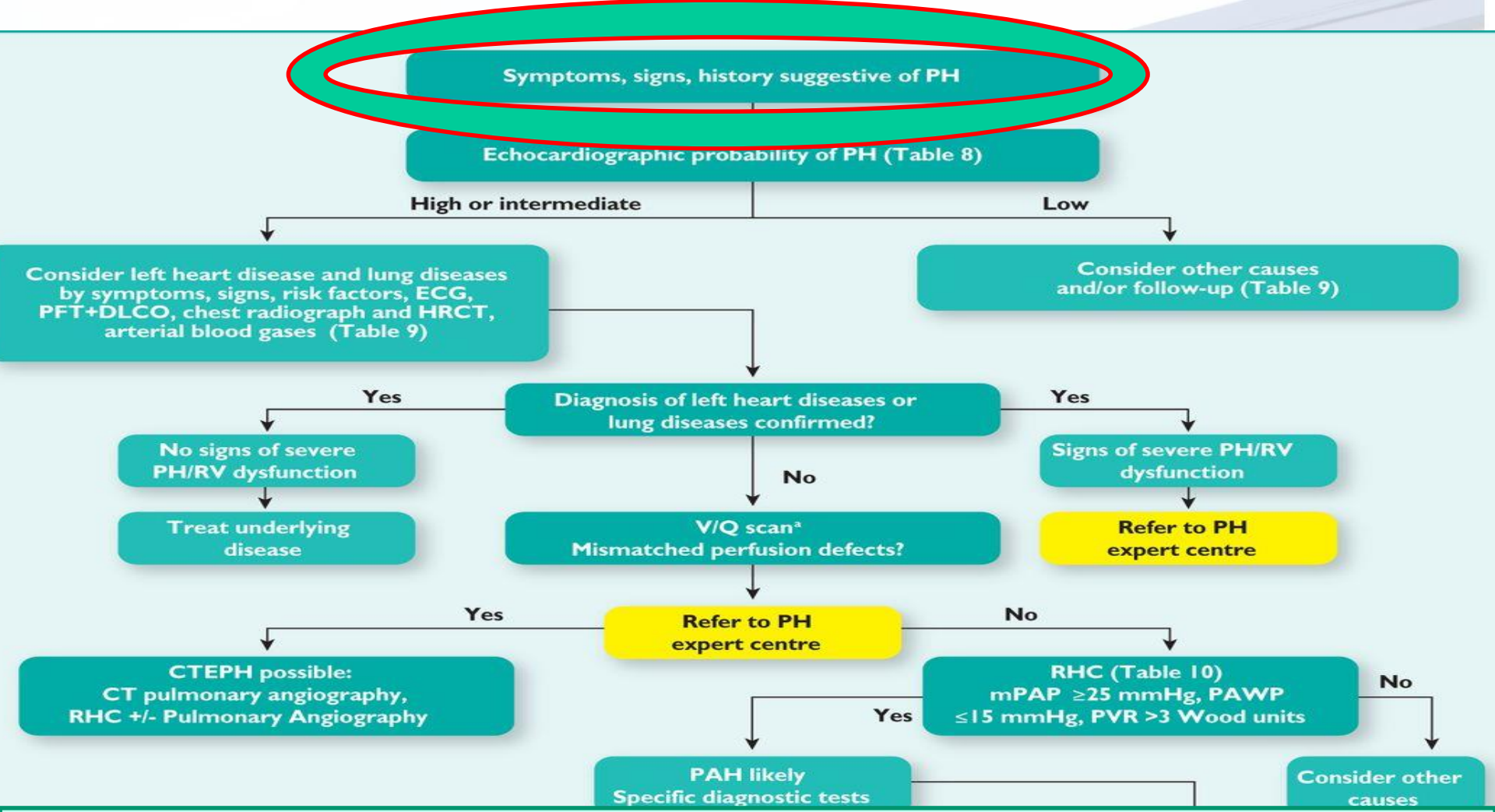

El estudio de la Disnea

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Clasificación
Calificación
Cuantificación de la disnea

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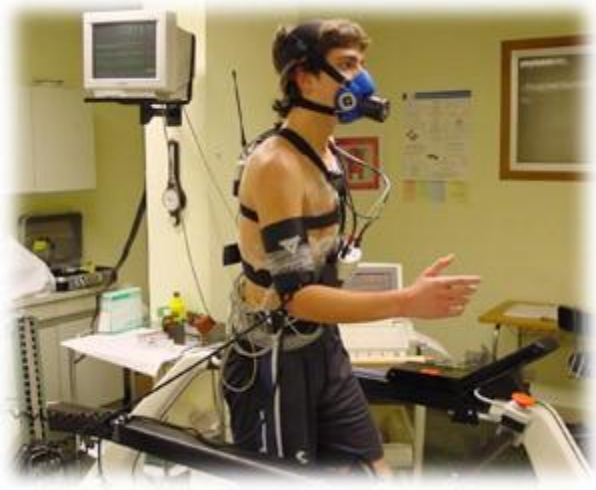
Pruebas de esfuerzo



Test de ejercicio cardiopulmonar

- Disminución del consumo máximo de oxígeno
- Disminución umbral anaeróbico
- Disminución de la reserva respiratoria
- Consumo máximo de O2 menor de 14 ml/kg/min mal pronostico
- Contraindicado en pacientes con sincope

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Pruebas de esfuerzo

Prueba de la caminata de los 6 minutos (6MWT)

- Correlaciona con la clase funcional de los pacientes
- Correlaciona con el estado hemodinámico
- Distancia recorrida :objetivo para evaluar el tratamiento

Menos de 332 metros: mal pronostico

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Once mas fueron tratados con ambrisentan por hipertensión arterial pulmonar del 2004 a la fecha todo perteneciente al grupo UAI salud o circuitos de seguridad social atendidos por nuestro sistema

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Reflexiones…

A los **5 años de**

seguimiento

AÑOS
2009/2010

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El promedio de test de caminata de seis minutos al ingreso al programa en nuestros paciente fue de 322 m

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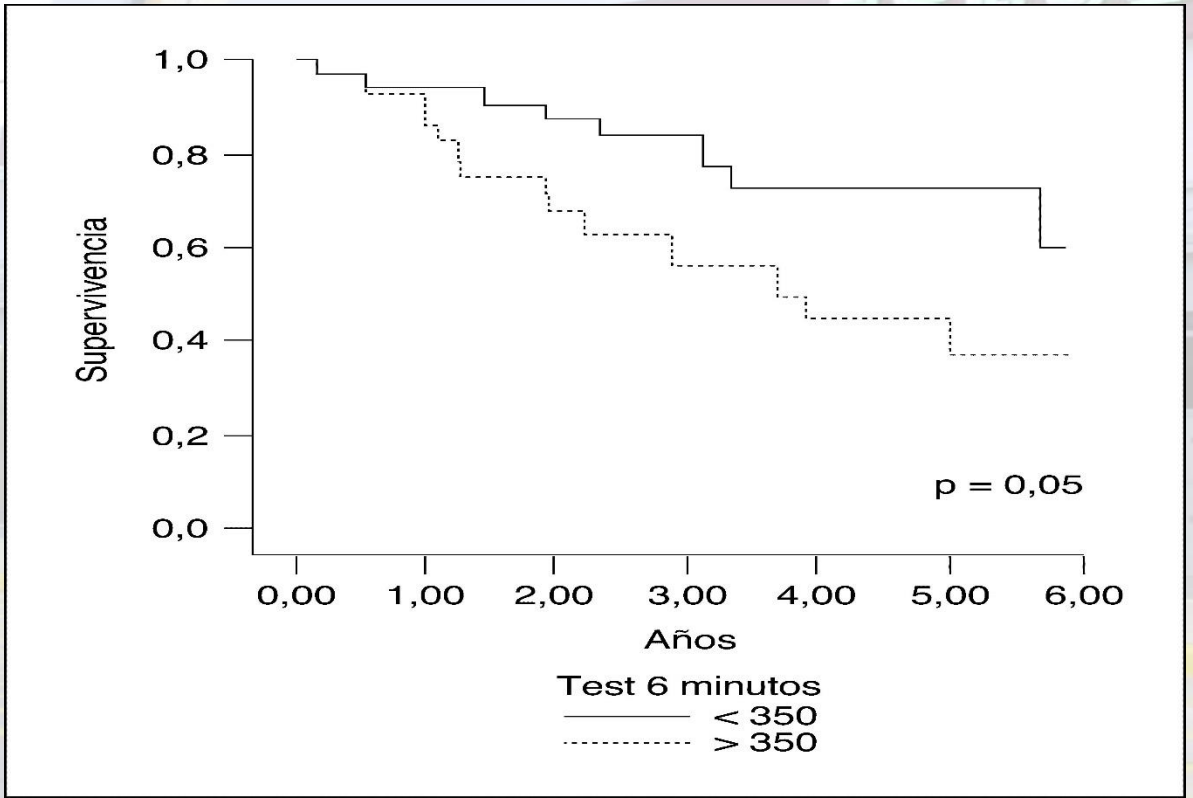
Supervivencia de los pacientes con hipertensión arterial pulmonar de acuerdo con el recorrido en el test de la marcha de 6 minutos en el momento del diagnóstico (Unidad de Colagenosis e Hipertensión Pulmonar del Hospital Universitario Virgen del Rocío, Sevilla).

Rev Clin Esp.2008;208:142-55 - Vol. 208 Núm.3
DOI: 10.1157/1311582



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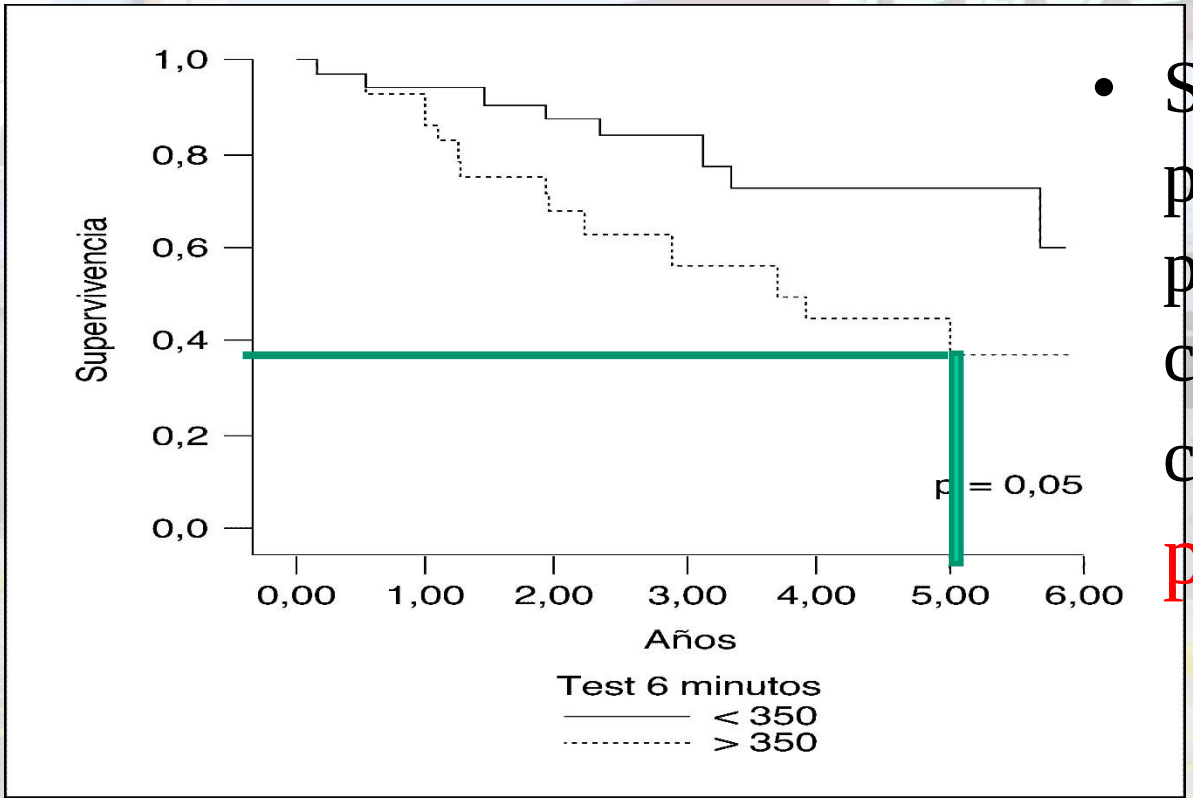
Supervivencia de los pacientes con hipertensión arterial pulmonar de acuerdo con el recorrido en el test de la marcha de 6 minutos en el momento del diagnóstico (Unidad de Colagenosis e Hipertensión Pulmonar del Hospital Universitario Virgen del Rocío, Sevilla).

Rev Clin Esp.2008;208:142-55 - Vol. 208 Núm.3
DOI: 10.1157/1311582



- Sobrevida esperada promedio de los pacientes a los 5 años clase según test de caminata 322m promedio

38 % aprox

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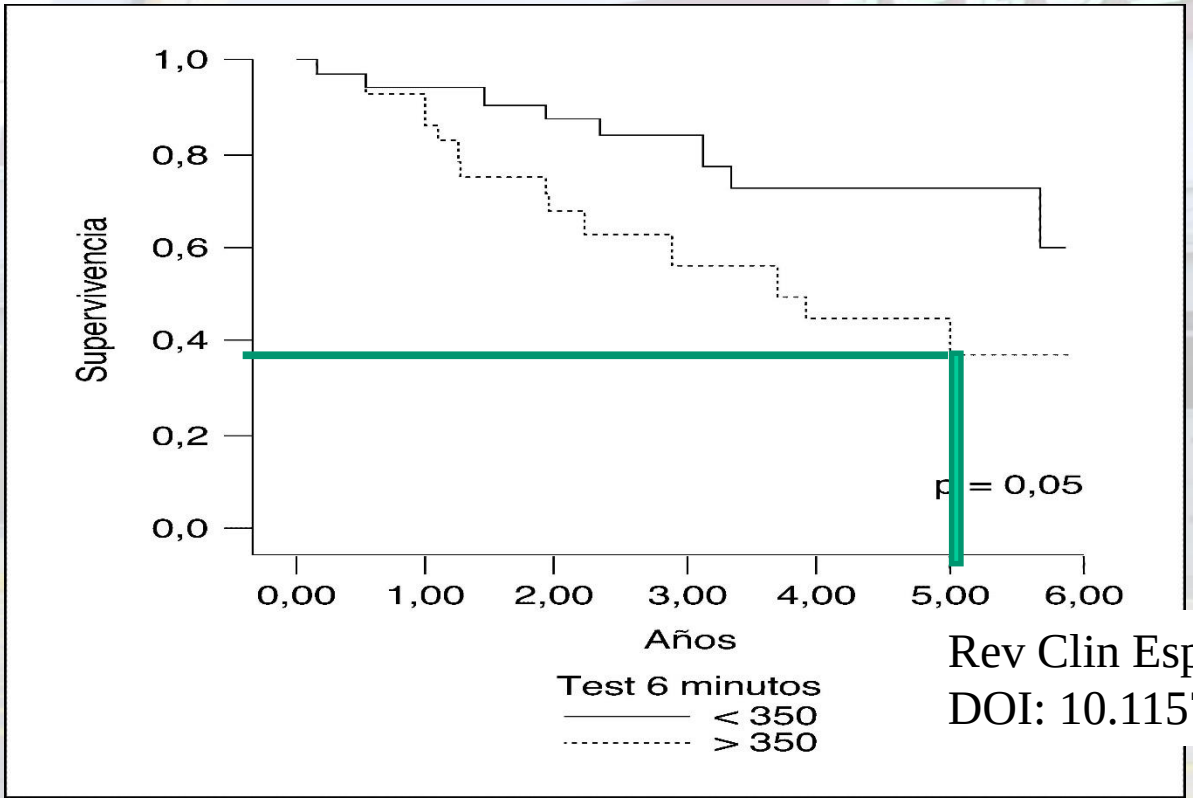
Reflexiones… No Conclusiones

Caminata 322m promedio



Sobrevida a 5 años 60%

Rev Clin Esp.2008;208:142-55 - Vol. 208 Núm.3
DOI: 10.1157/1311582

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Reflexiones… No Conclusiones

- Sobrevida esperada promedio a los 5 años

  38%

- Sobrevida observada al
  - 1 año 100 %
  - 2 años 80%
  - 3 años 80 %
  - 4 años 80%
  - **5 años 60 %**
  - 11 años 60%

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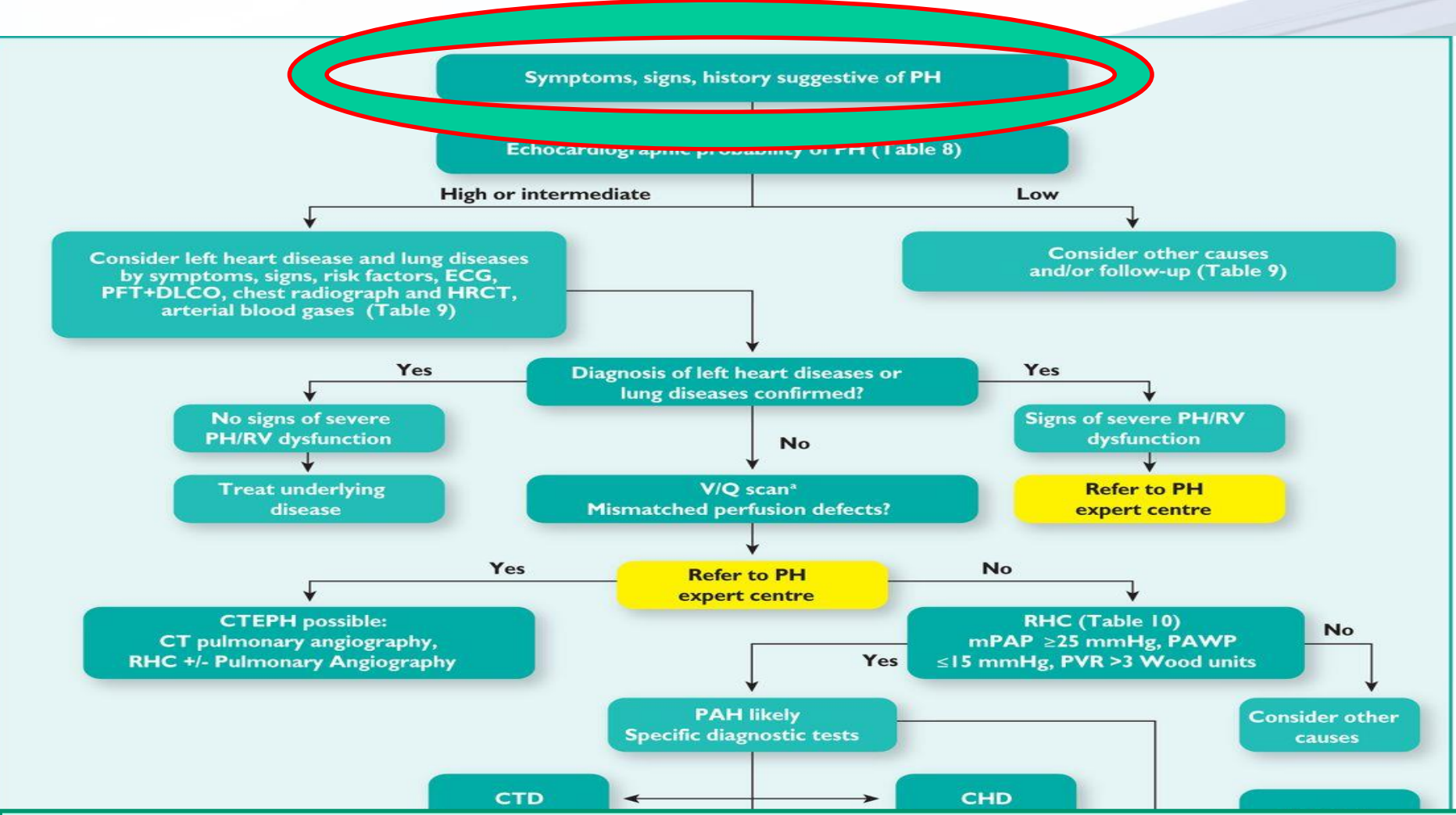

LA CLINICA

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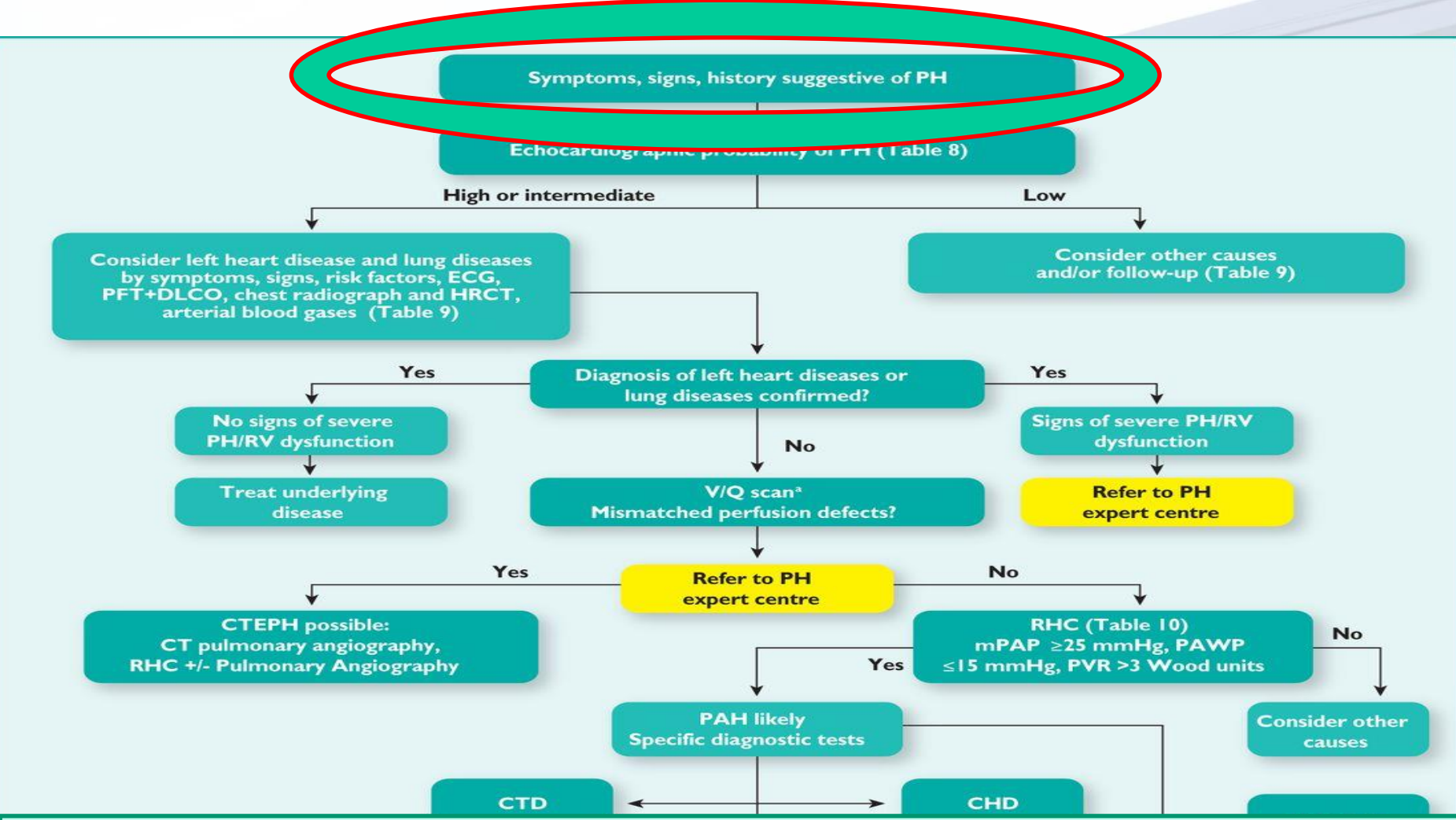

Signo de Dressler- Segundo ruido cardíaco aumentado,
– Insuficiencia tricuspídea -Insuficiencia pulmonar,
– Tercer ruido derecho Distensión yugular

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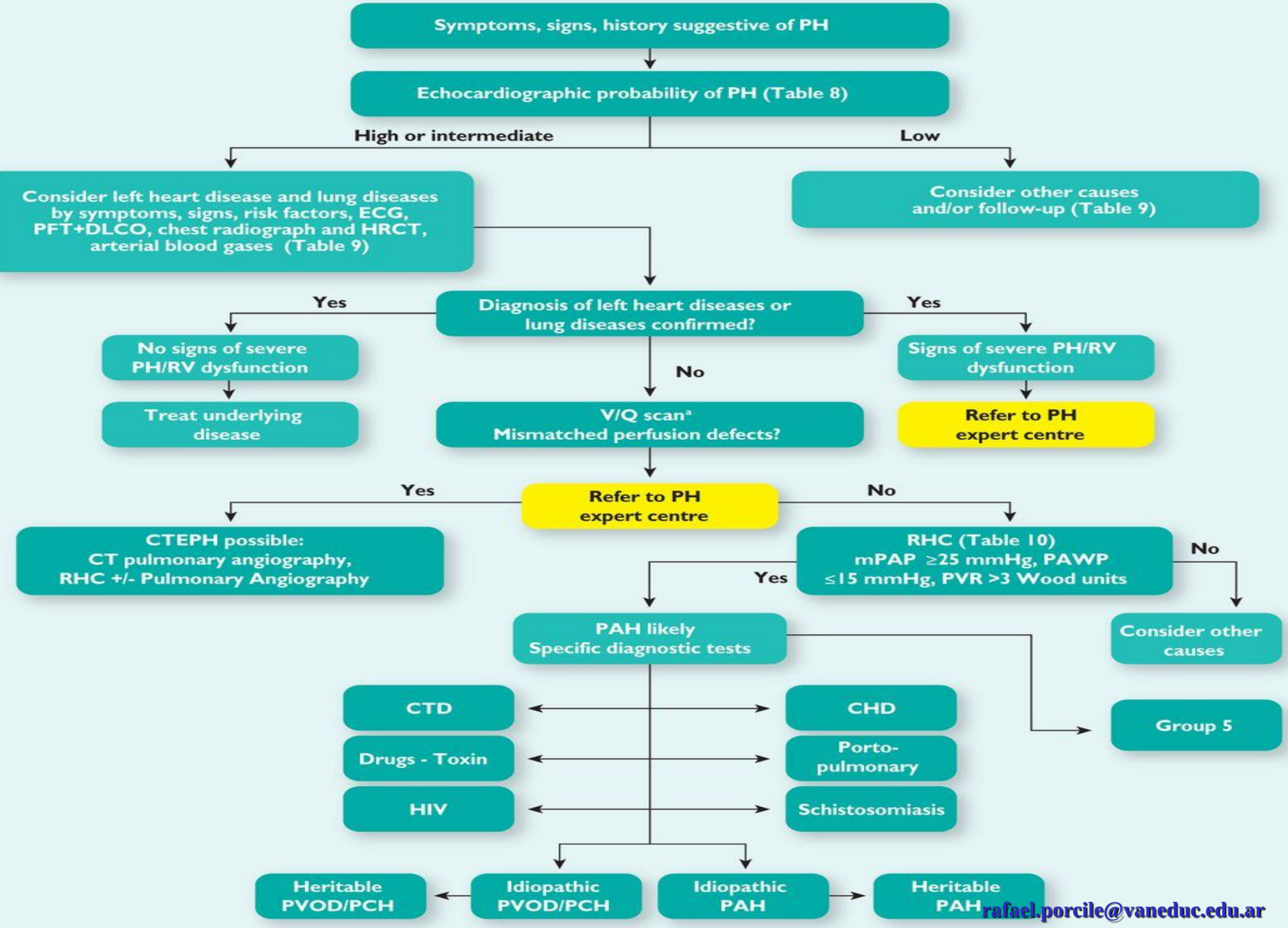

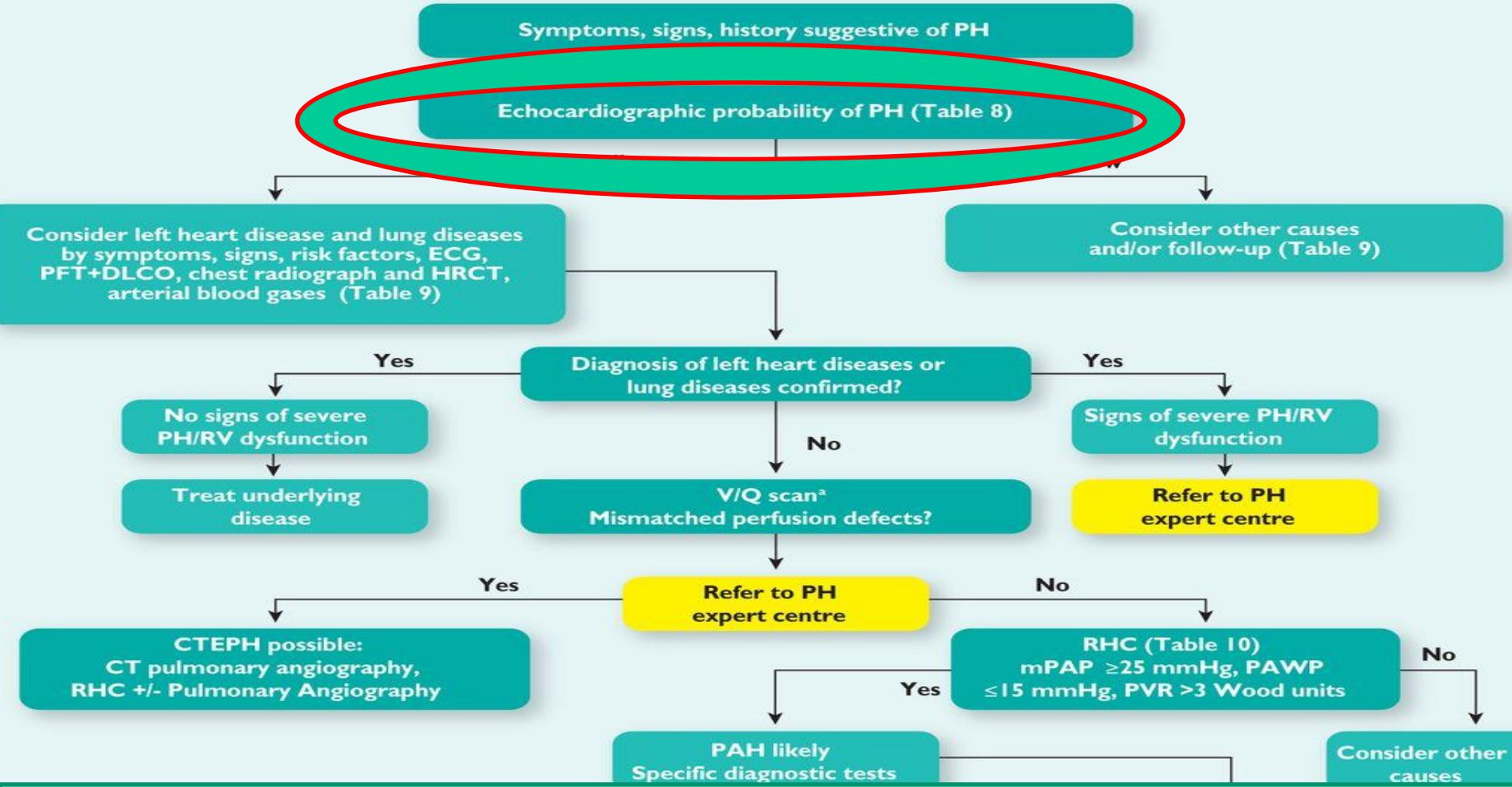

Ecocardiograma

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El ecocardiograma

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European Heart Journal Advance Access published September 15, 2015

European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

Authors/Task Force Members: Nazzareno Galiè[*] (ESC Chairperson) (Italy), Marc Humbert[*a] (ERS Chairperson) (France), Jean-Luc Vachiery[c] (Belgium), Simon Gibbs (UK), Irene Lang (Austria), Adam Torbicki (Poland), Gérald Simonneau[a] (France), Andrew Peacock[a] (UK), Anton Vonk Noordegraaf[a] (The Netherlands), Maurice Beghetti[b] (Switzerland), Ardeschir Ghofrani[a] (Germany), Miguel Angel Gomez Sanchez (Spain), Georg Hansmann[b] (Germany), Walter Klepetko[c] (Austria), Patrizio Lancellotti (Belgium), Marco Matucci[d] (Italy), Theresa McDonagh (UK), Luc A. Pierard (Belgium), Pedro T. Trindade (Switzerland), Maurizio Zompatori[e] (Italy) and Marius Hoeper[a] (Germany)

[*] Corresponding authors: Nazzareno Galiè, Department of Experimental, Diagnostic and Specialty Medicine – DIMES, University of Bologna, Via Massarenti 9, 40138 Bologna, Italy. Tel: +39 051 349 858, Fax: +39 051 344 859, Email: nazzareno.galie@unibo.it

Marc Humbert, Service de Pneumologie, Hôpital Bicêtre, Université Paris-Sud, Assistance Publique Hôpitaux de Paris, 78 rue du Général Leclerc, 94270 Le Kremlin-Bicêtre, France. Tel: +33 145212972, Fax: +33 145217971, Email: marc.humbert@aphp.fr

ESC Committee for Practice Guidelines (CPG) and National Cardiac Societies document reviewers: listed in Appendix

[a]Representing the European Respiratory Society; [b]Representing the Association for European Paediatric and Congenital Cardiology; [c]Representing the International Society for Heart and Lung Transplantation; [d]Representing the European League Against Rheumatism; and [e]Representing the European Society of Radiology.

ESC entities having participated in the development of this document:

ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

ESC Councils: Council for Cardiology Practice (CCP), Council on Cardiovascular Nursing and Allied Professions (CCNAP), Council on Cardiovascular Primary Care (CCPC).

ESC Working Groups: Cardiovascular Pharmacotherapy, Cardiovascular Surgery, Grown-up Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Valvular Heart Disease.

This article is being published concurrently in the *European Heart Journal* (10.1093/eurheartj/ehv317) and the *European Respiratory Journal* (10.1183/13993003.01032-2015). The articles are identical except for minor stylistic and spelling differences in keeping with each journal's style. Either citation can be used when citing this article.

A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50% with a sniff or <20% with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm.	

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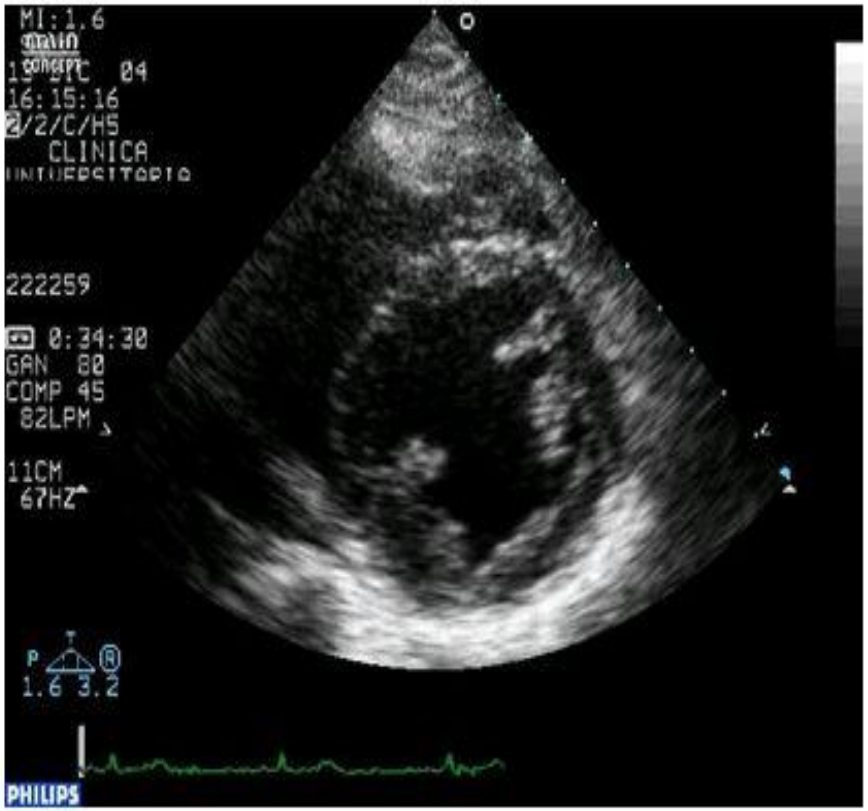
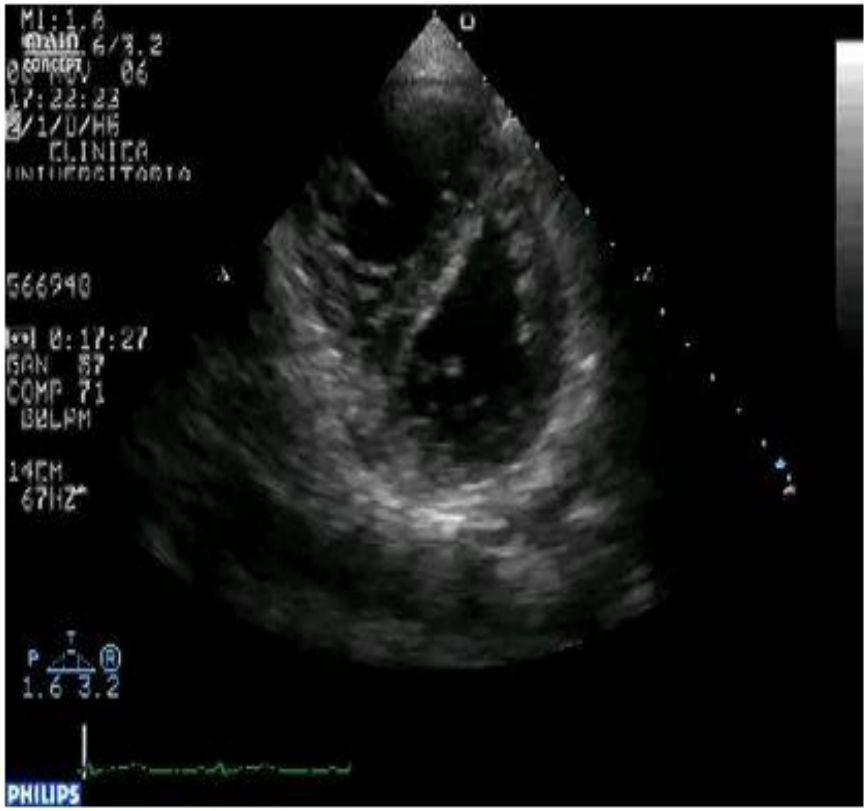
TABLA 4. Criterios ecocardiográficos para evaluar la posibilidad del diagnóstico de hipertensión pulmonar

	Clase[a]	Nivel[b]
Diagnóstico ecocardiográfico: HP improbable		
Velocidad de regurgitación tricuspídea ≤ 2,8 m/s, presión sistólica en AP ≤ 36 mmHg, y sin variables ecocardiográficas adicionales que parecen indicar una HP	I	B
Diagnóstico ecocardiográfico: HP posible		
Velocidad de regurgitación tricuspídea ≤ 2,8 m/s, presión sistólica en AP ≤ 36 mmHg, pero con variables ecocardiográficas adicionales que parecen indicar una HP	IIa	C
Velocidad de regurgitación tricuspídea de 2,9-3,4 m/s, presión sistólica AP de 37-50 mmHg con/sin variables ecocardiográficas adicionales que parecen indicar una HP	IIa	C
Diagnóstico ecocardiográfico: HP probable		
Velocidad de regurgitación tricuspídea > 3,4 m/s, presión sistólica en AP > 50 mmHg, con/sin variables ecocardiográficas adicionales que parecen indicar un HP	I	B
La ecocardiografía Doppler durante el ejercicio no es recomendable para explorar la HP	III	C

Ecocardiografia



Normal



Sobrecarga de presión VD

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Pronóstico mediante Ecocardiograma.

- Derrame pericárdico: signo indirecto de disfunción ventricular y PAP muy elevadas, posiblemente relación con alteración del drenaje linfático.
- Doppler tisular pulsado anillo tricuspideo: Velocidad pico de la onda S < 11.5 cm/s indica disfunción VD y <8 cm/s severamente deprimido pobre pronostico.
- TAPSE: cuando es < 15 se relaciona mortalidad alta.

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LIMITACIONES DEL ECOCARDIOGRAMA:

1) está ausente hasta el 16% de los pacientes
2) en algunos estudios se han documentado diferencias de hasta 20 mmHg al compararlo con cateterimo ventricular derecho.

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Por que usar el ecocardiograma solo como Screening?

- PSAP se tiende a subestimar a través de la medición de la velocidad de regurgitación tricuspidea y puede presentar diferencias en las medidas de mas de 10 mmHg por lo que no se debe utilizar para decidir cuando iniciar el tratamiento o monitorizar la respuesta al tratamiento.
- Los demás parámetros no se relación de una forma lineal con la presión pulmonar

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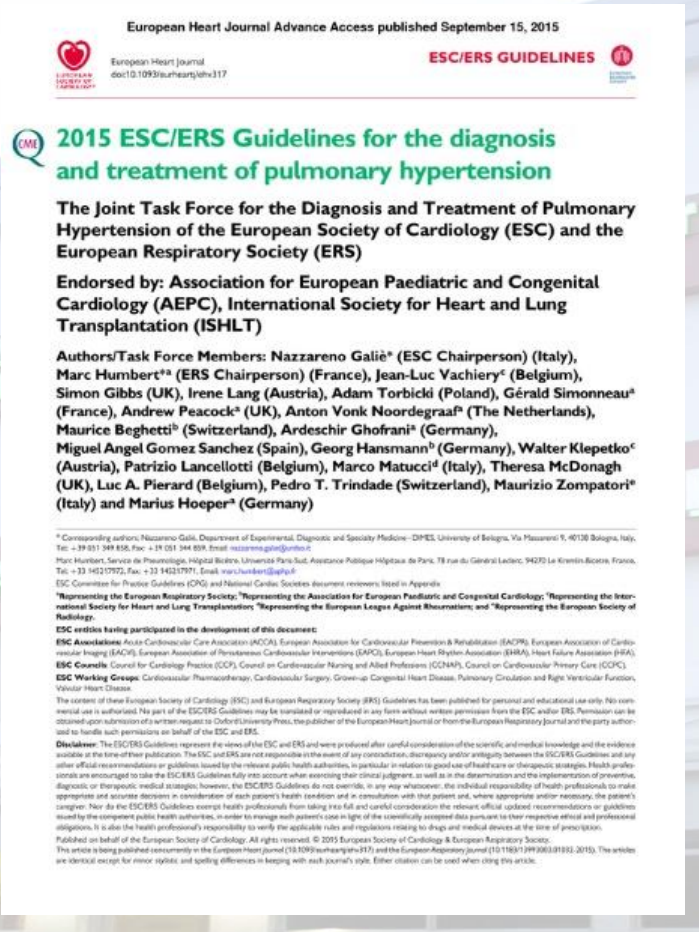

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European Heart Journal
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ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

Authors/Task Force Members: Nazzareno Galiè* (ESC Chairperson) (Italy), Marc Humbert*[a] (ERS Chairperson) (France), Jean-Luc Vachiery[c] (Belgium), Simon Gibbs (UK), Irene Lang (Austria), Adam Torbicki (Poland), Gérald Simonneau[a] (France), Andrew Peacock[a] (UK), Anton Vonk Noordegraaf[b] (The Netherlands), Maurice Beghetti[b] (Switzerland), Ardeschir Ghofrani[a] (Germany), Miguel Angel Gomez Sanchez (Spain), Georg Hansmann[b] (Germany), Walter Klepetko[c] (Austria), Patrizio Lancellotti (Belgium), Marco Matucci[d] (Italy), Theresa McDonagh (UK), Luc A. Pierard (Belgium), Pedro T. Trindade (Switzerland), Maurizio Zompatori[e] (Italy) and Marius Hoeper[a] (Germany)

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ESC Committee for Practice Guidelines (CPG) and National Cardiac Societies document reviewers: listed in Appendix

[a]Representing the European Respiratory Society; [b]Representing the Association for European Paediatric and Congenital Cardiology; [c]Representing the International Society for Heart and Lung Transplantation; [d]Representing the European League Against Rheumatism; and [e]Representing the European Society of Radiology.

ESC entities having participated in the development of this document:

ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

ESC Councils: Council for Cardiology Practice (CCP), Council on Cardiovascular Nursing and Allied Professions (CCNAP), Council on Cardiovascular Primary Care (CCPC).

ESC Working Groups: Cardiovascular Pharmacotherapy, Cardiovascular Surgery, Grown-up Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Valvular Heart Disease.

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Clinical classification of pulmonary arterial hypertension associated with congenital heart disease (updated from Simonneau et al.[5])

1. Eisenmenger's syndrome

Includes all large intra- and extra-cardiac defects which begin as systemic-to-pulmonary shunts and progress with time to severe elevation of PVR and to reversal (pulmonary-to-systemic) or bidirectional shunting; cyanosis, secondary erythrocytosis, and multiple organ involvement are usually present.

2. PAH associated with prevalent systemic-to-pulmonary shunts

- Correctable[a]
- Non-correctable

Includes moderate to large defects; PVR is mildly to moderately increased, systemic-to-pulmonary shunting is still prevalent, whereas cyanosis at rest is not a feature.

3. PAH with small/coincidental defects [b]

Marked elevation in PVR in the presence of small cardiac defects (usually ventricular septal defects <1 cm and atrial septal defects <2 cm of effective diameter assessed by echo), which themselves do not account for the development of elevated PVR; the clinical picture is very similar to idiopathic PAH. Closing the defects is contra-indicated.

4. PAH after defect correction

Congenital heart disease is repaired, but PAH either persists immediately after correction or recurs/develops months or years haemodynamic lesions.

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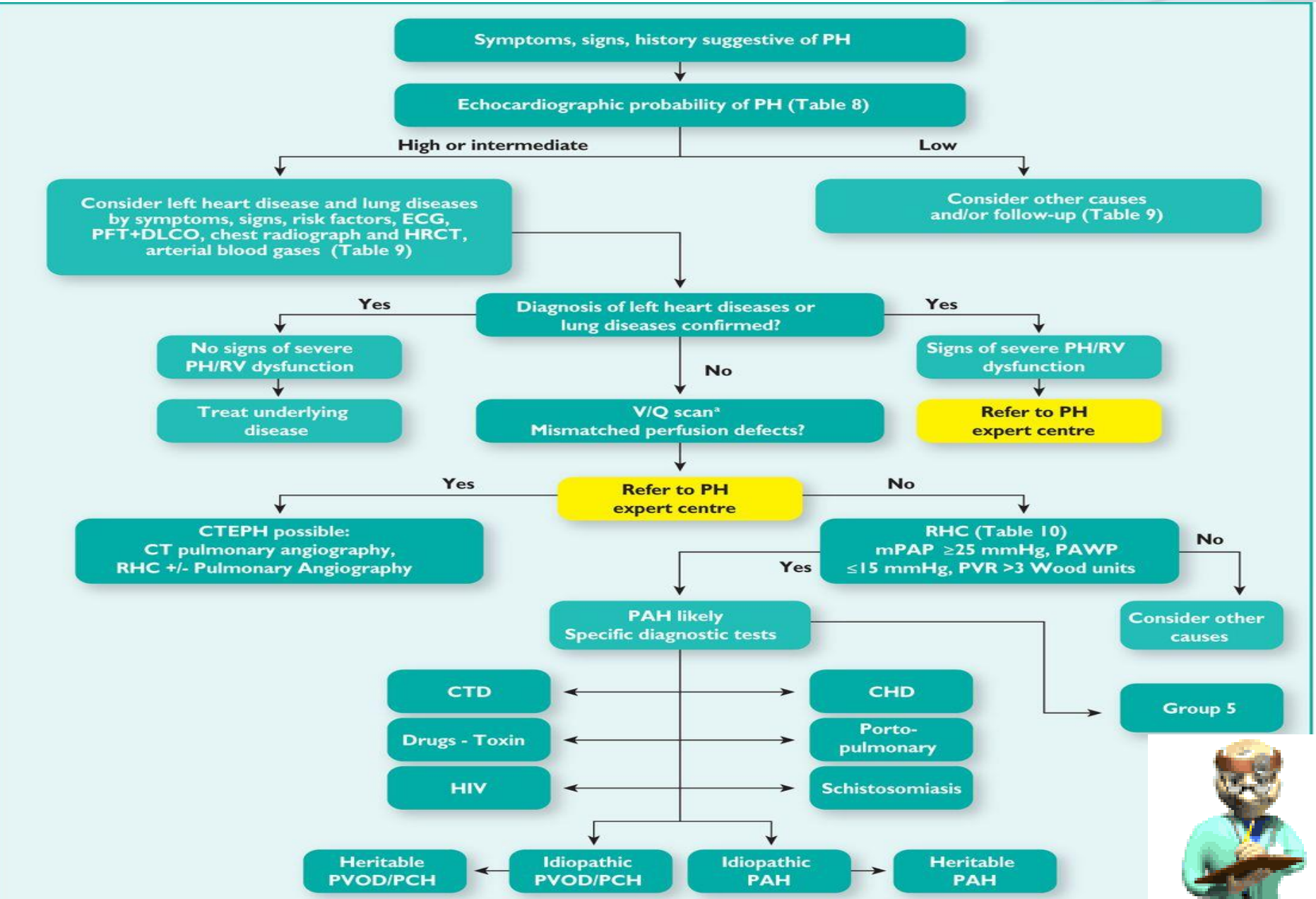
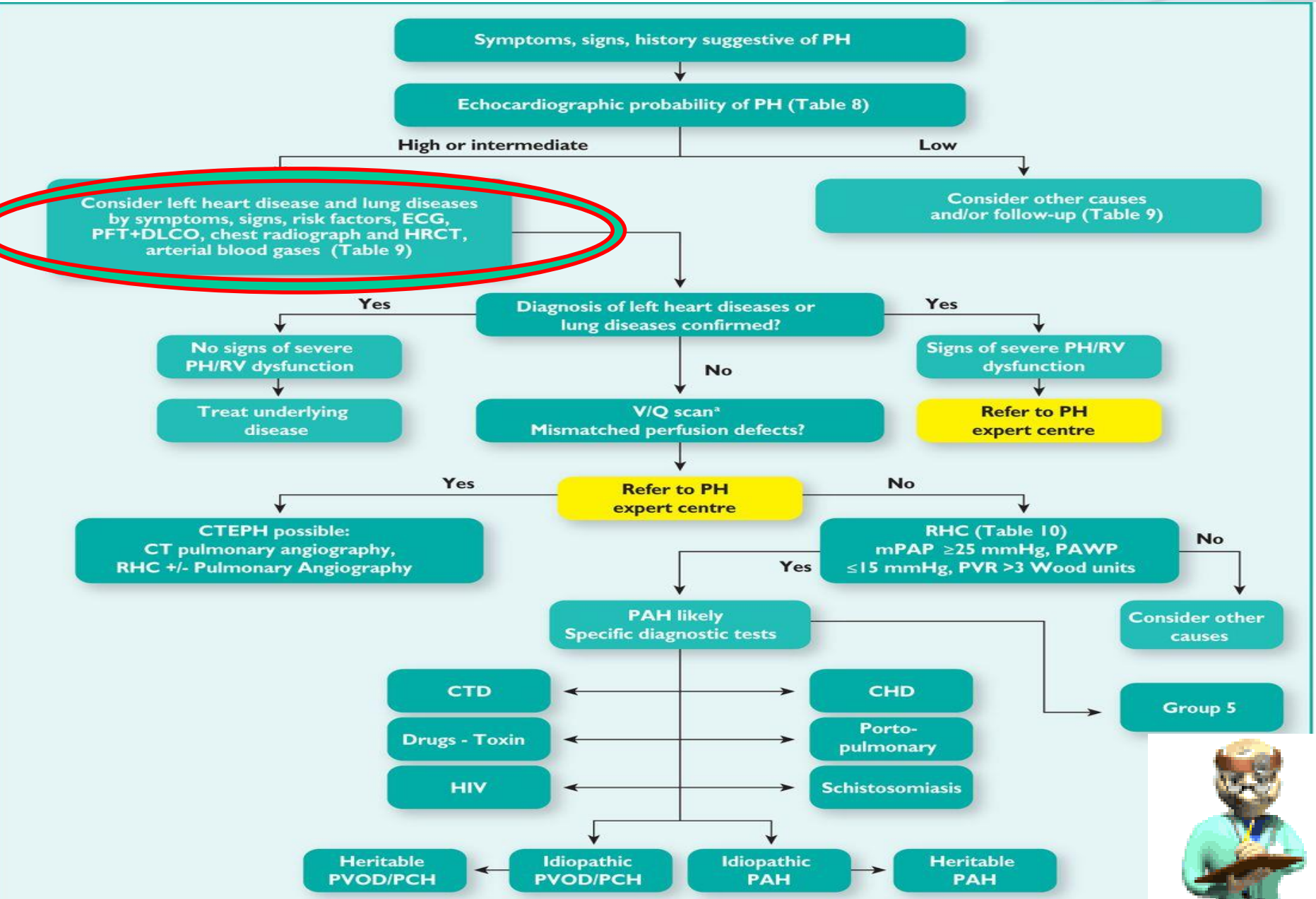


CHD = congenital heart diseases; CT = computed tomography; CTD = connective tissue disease; CTEPH = chronic thromboembolic pulmonary hypertension; DLCO = carbon monoxide diffusing capacity; ECG = electrocardiogram; HIV = Human immunodeficiency virus; HR-CT = high resolution CT; mPAP = mean pul pressure; PA = pulmonary angiography; PAH = pulmonary arterial hypertension; PAWP = pulmonary artery wedge pressure; PFT = pulmonary function tests; PH = pulmonary hypertension; PVOD/PCH = pulmonary veno-occlusive disease or pulmonary capillary hemangiomathosis; PVR = pulmonary vascular resistance RHC = right heart catheterisation; RV = right ventricular; V/Q = ventilation/perfusion.



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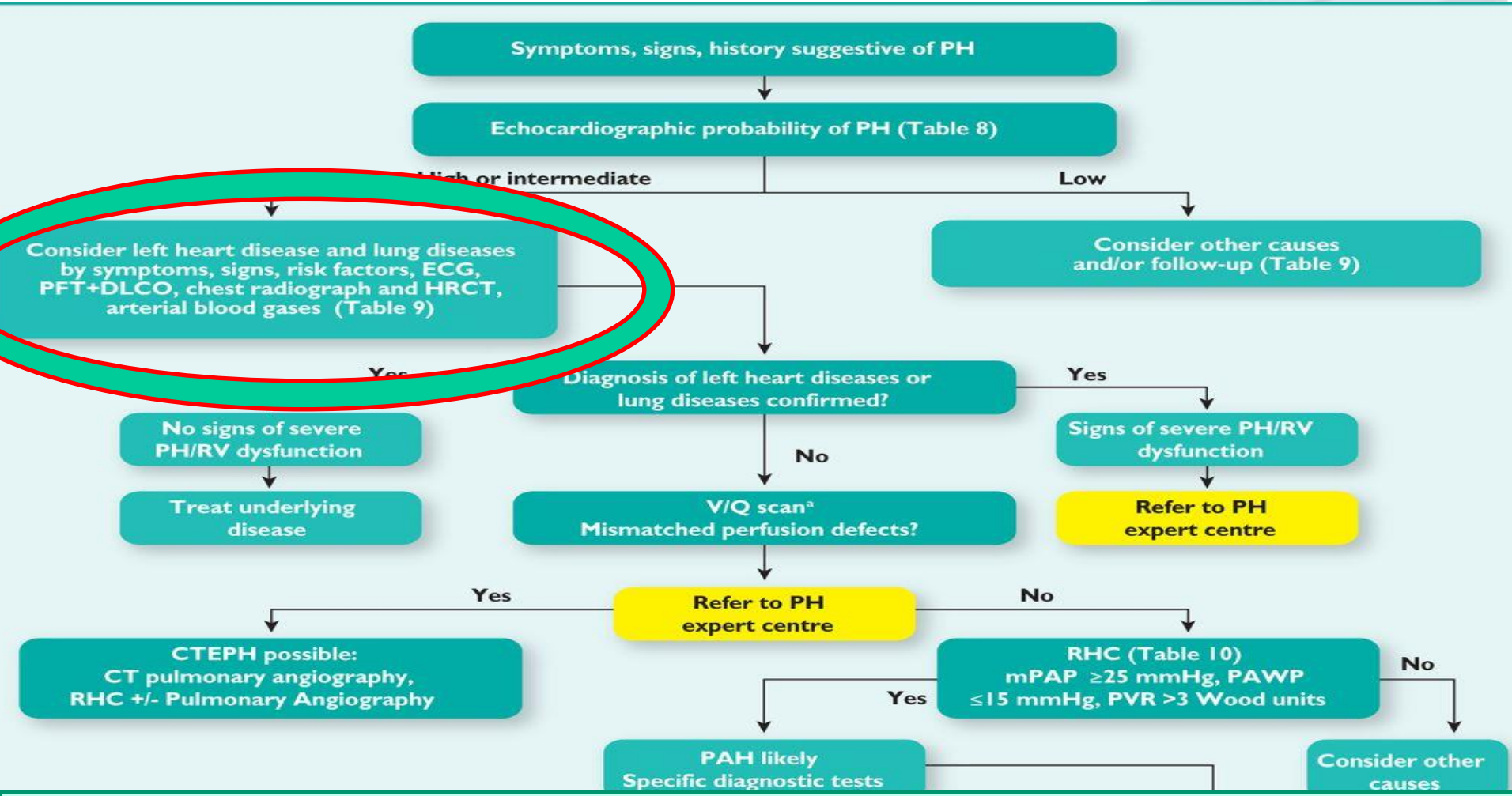

ECG+RX+GASES ARTERIALES

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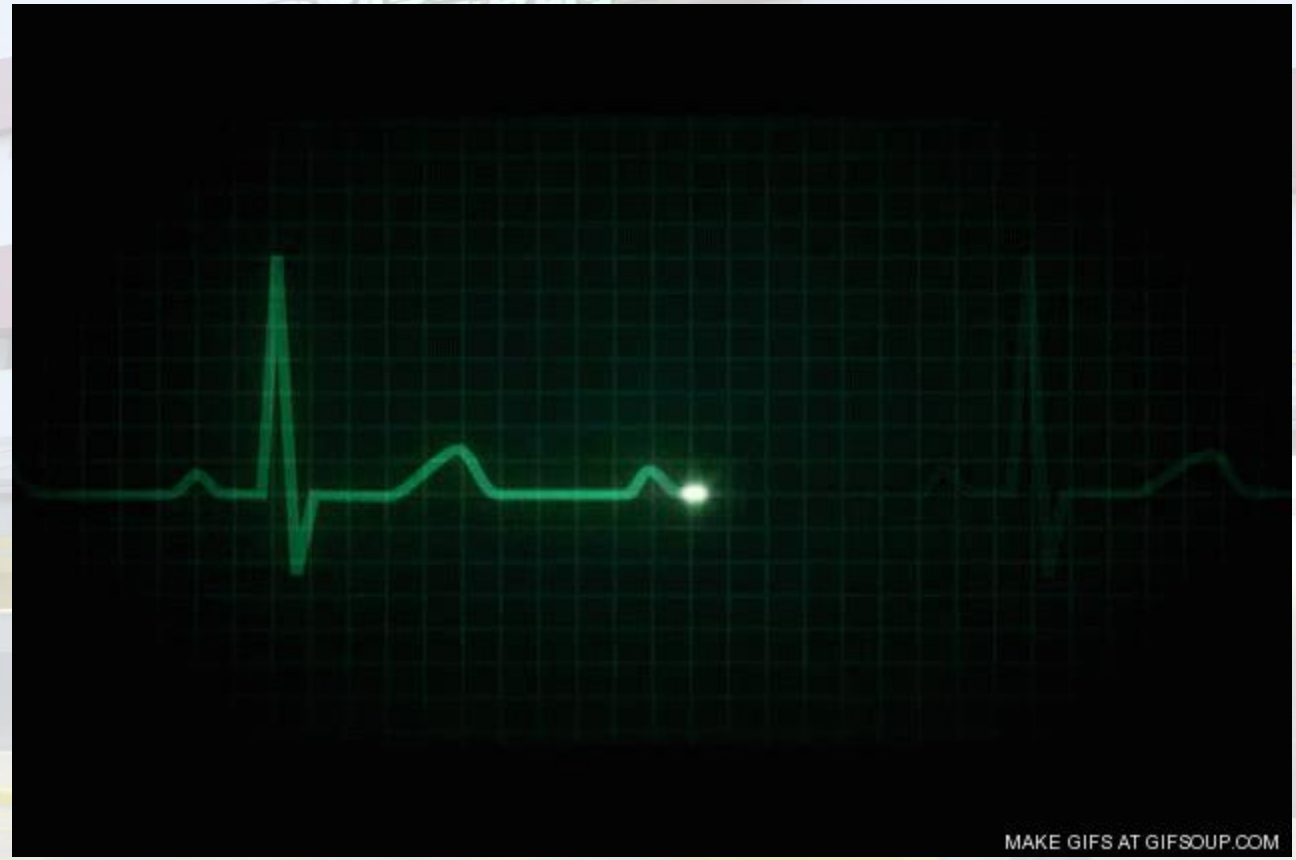
El electrocardiograma



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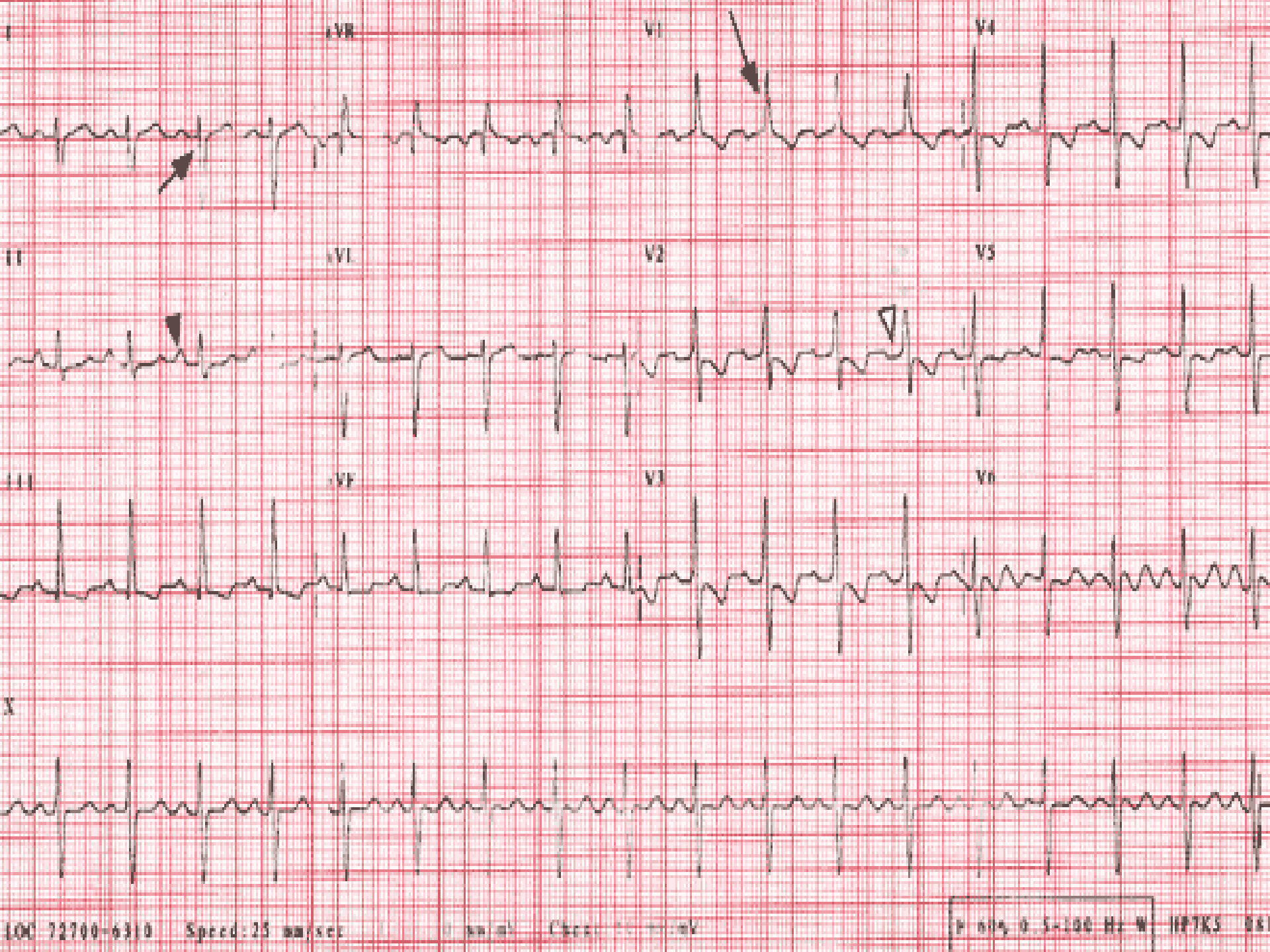
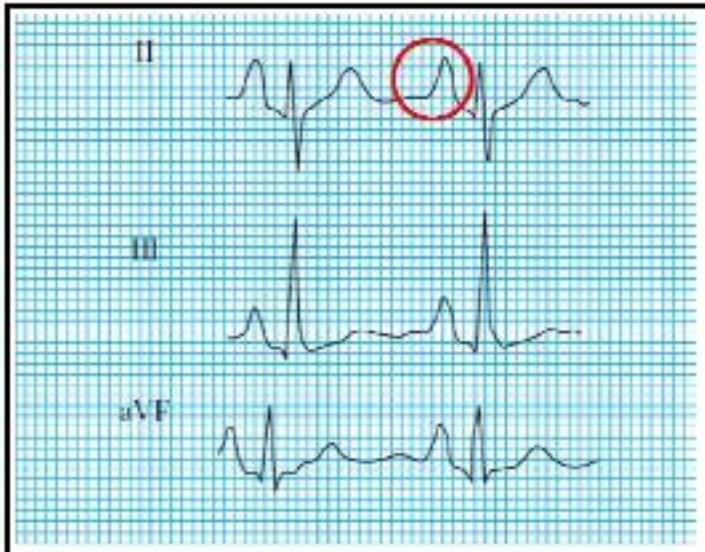

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
X
LOC 72700-6310 Speed:25 mm/sec
P 0.5-100 Hz W

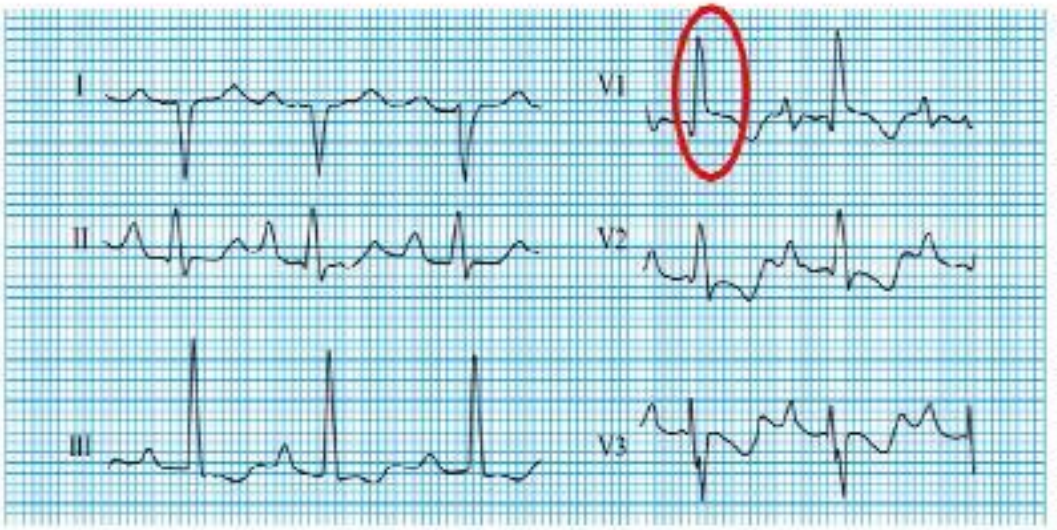


Large P waves in leads II, III, and aVF (P pulmonale)

CRECIMIENTO AURICULAR DERECHO

DERECHO

Aumento de la onda P: P PULMONAR



CRECIMIENTO VENTRICULAR

Aumento de la onda R

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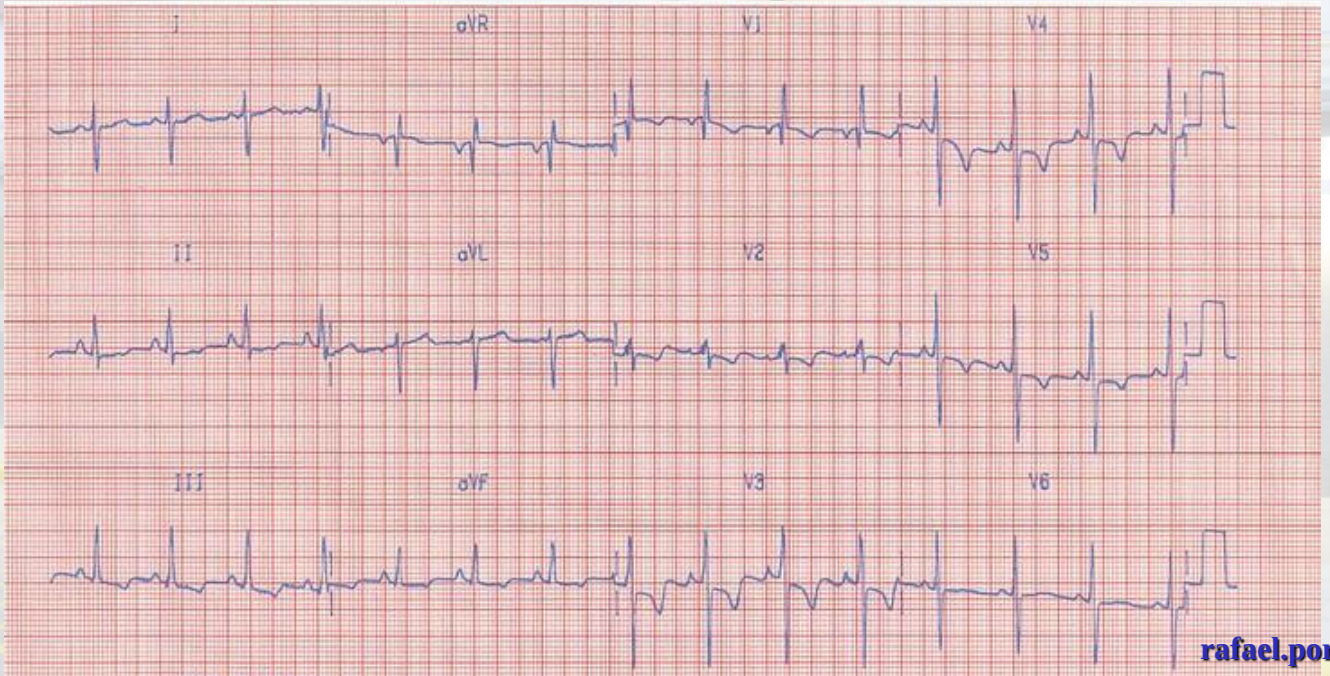
Electrocardiograma

- Sensibilidad del 55% especificidad del 70%, por lo que **no es útil como herramienta de detección**

  Hipertrofia ventricular derecha (87%)
- Desviación del eje a la derecha (79%)



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SE DESCARTA O CONFIRMA

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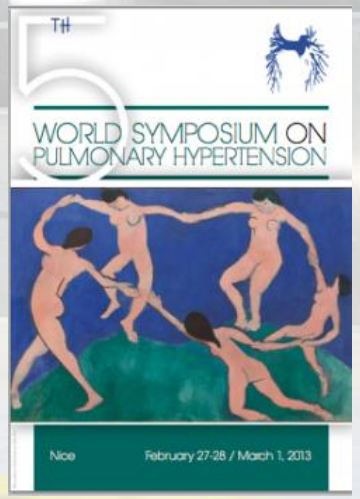
Actualización en clasificación de la hipertensión pulmonar

* Hipertensión arterial pulmonar (PAH)
* **Secundaria a falla ventricular izquierda**
* Secundaria a enfermedad pulmonar con o sin hipoxemia
* Hipertensión pulmonar secundaria a tromboembolismo crónico
* Mecanismos poco claros o multifactorial



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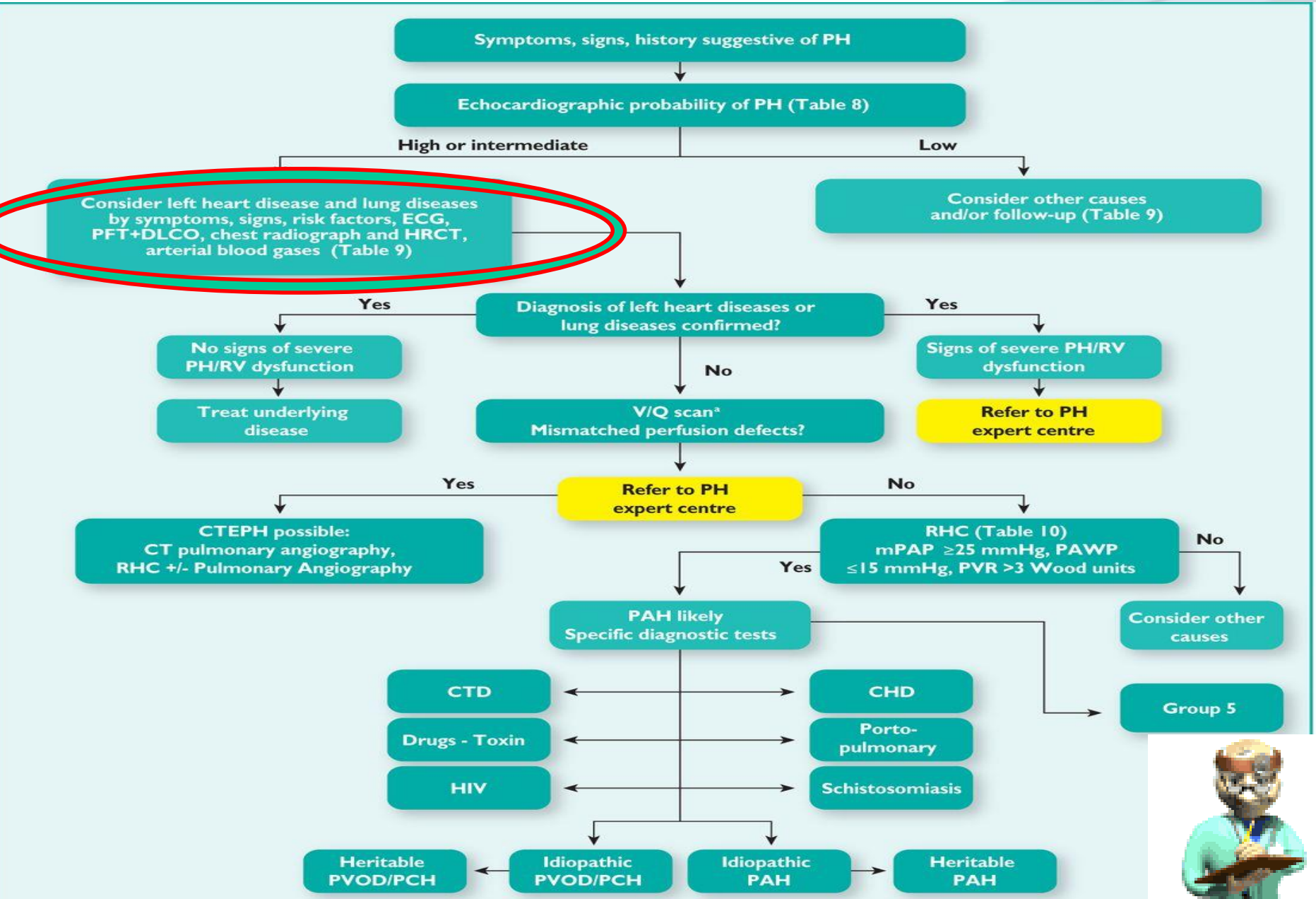

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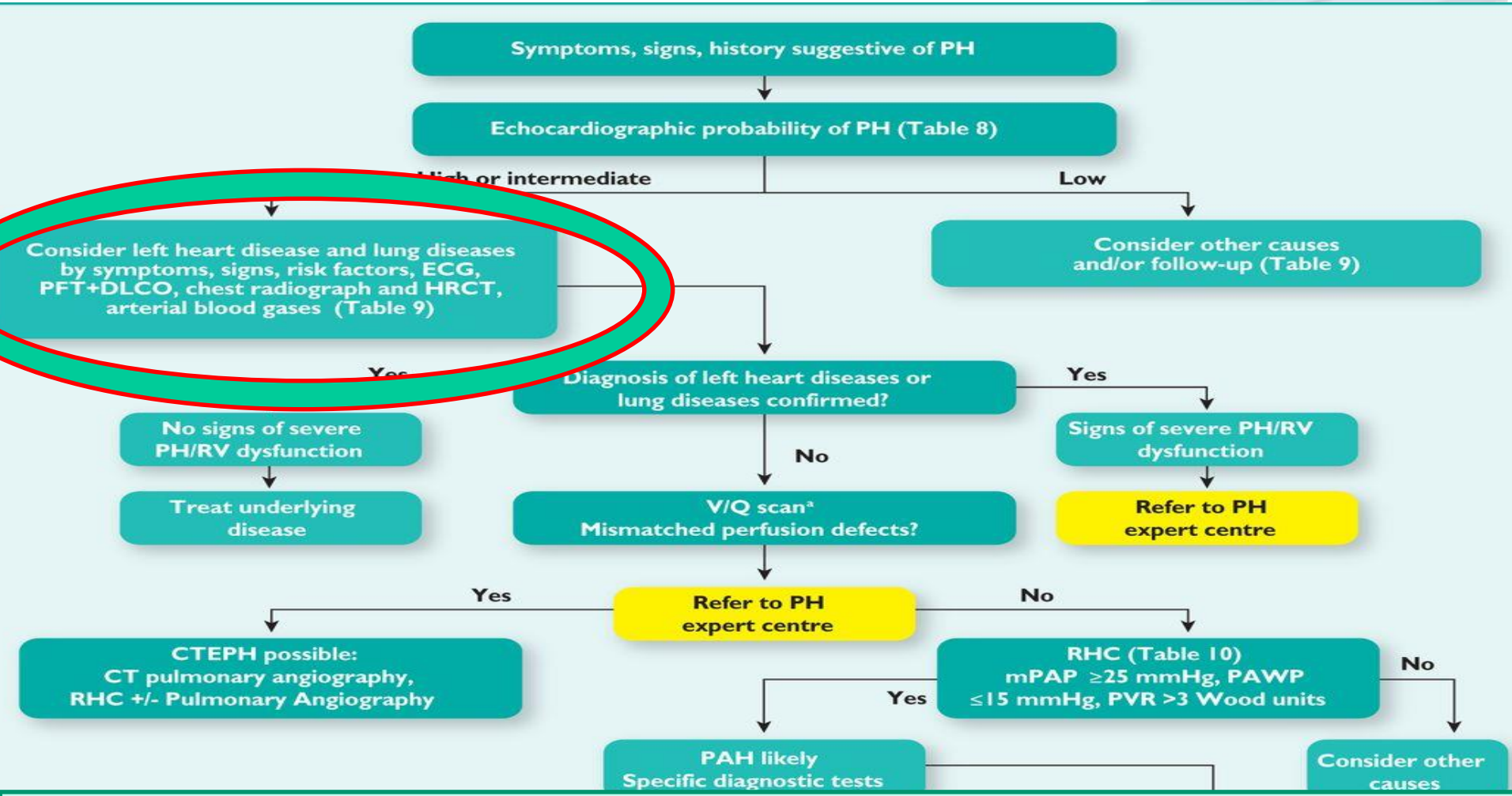

EVALUACIÓN NEUMONOLOGICA

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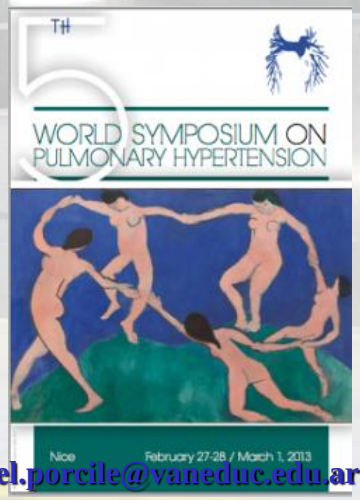

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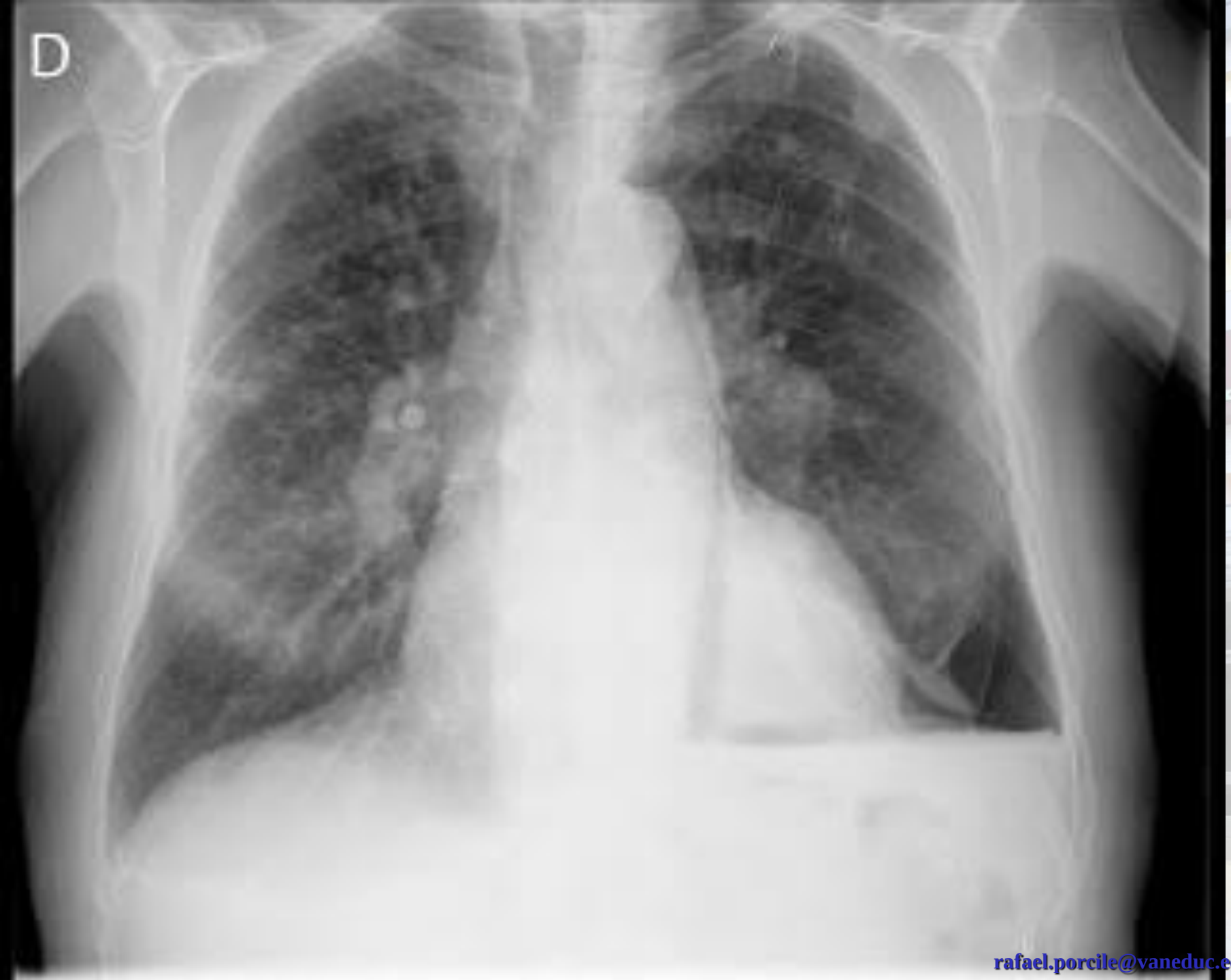

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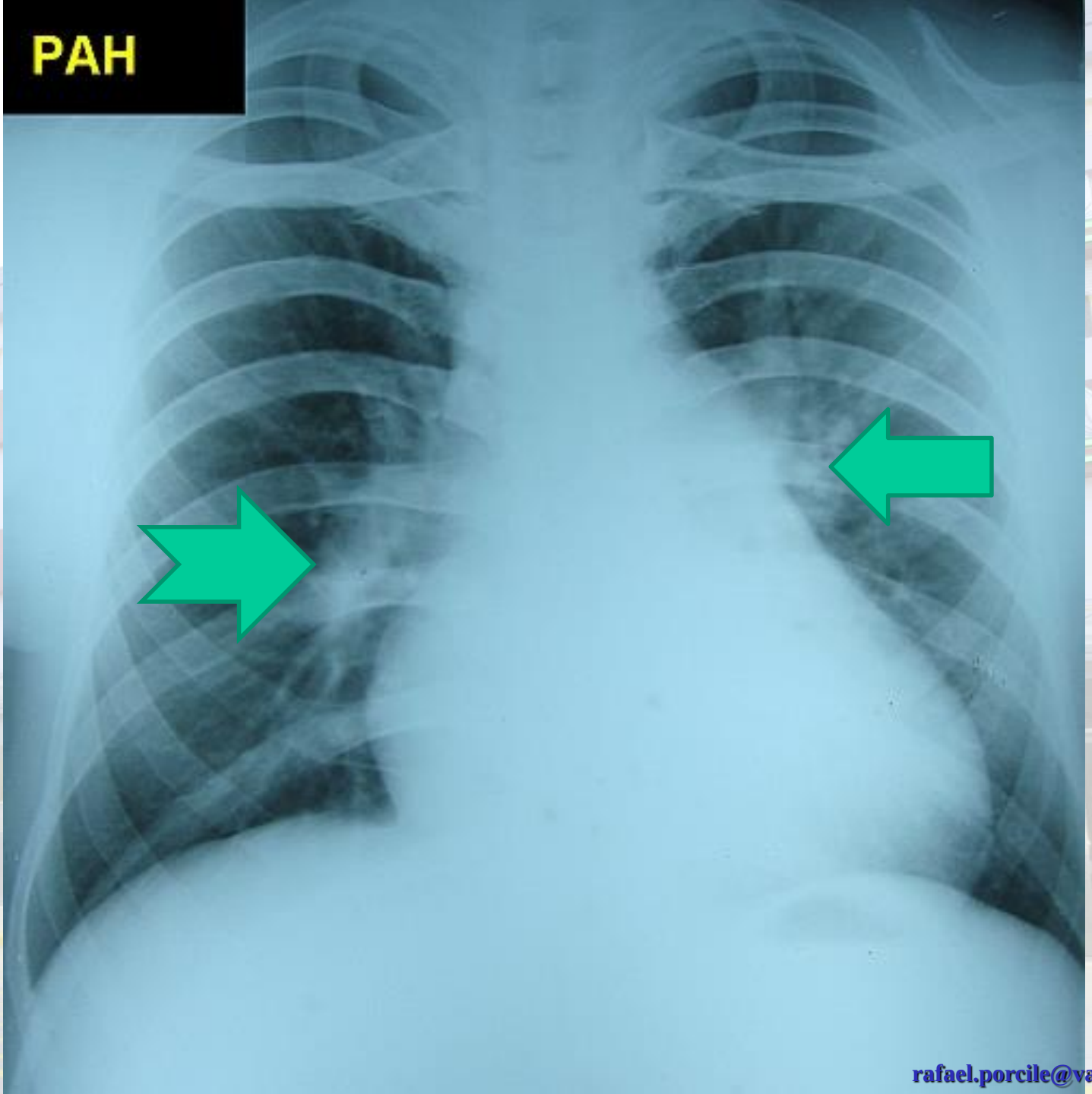

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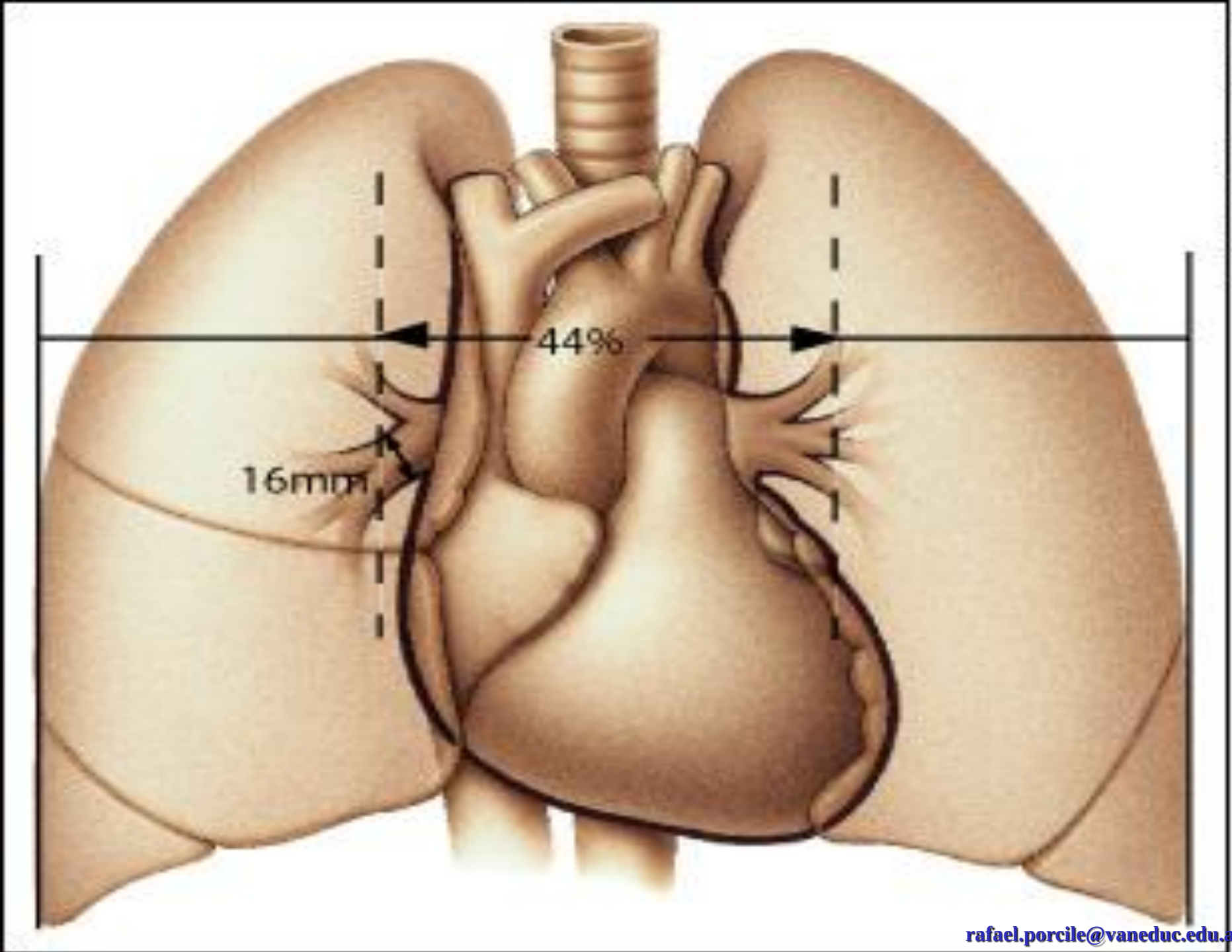

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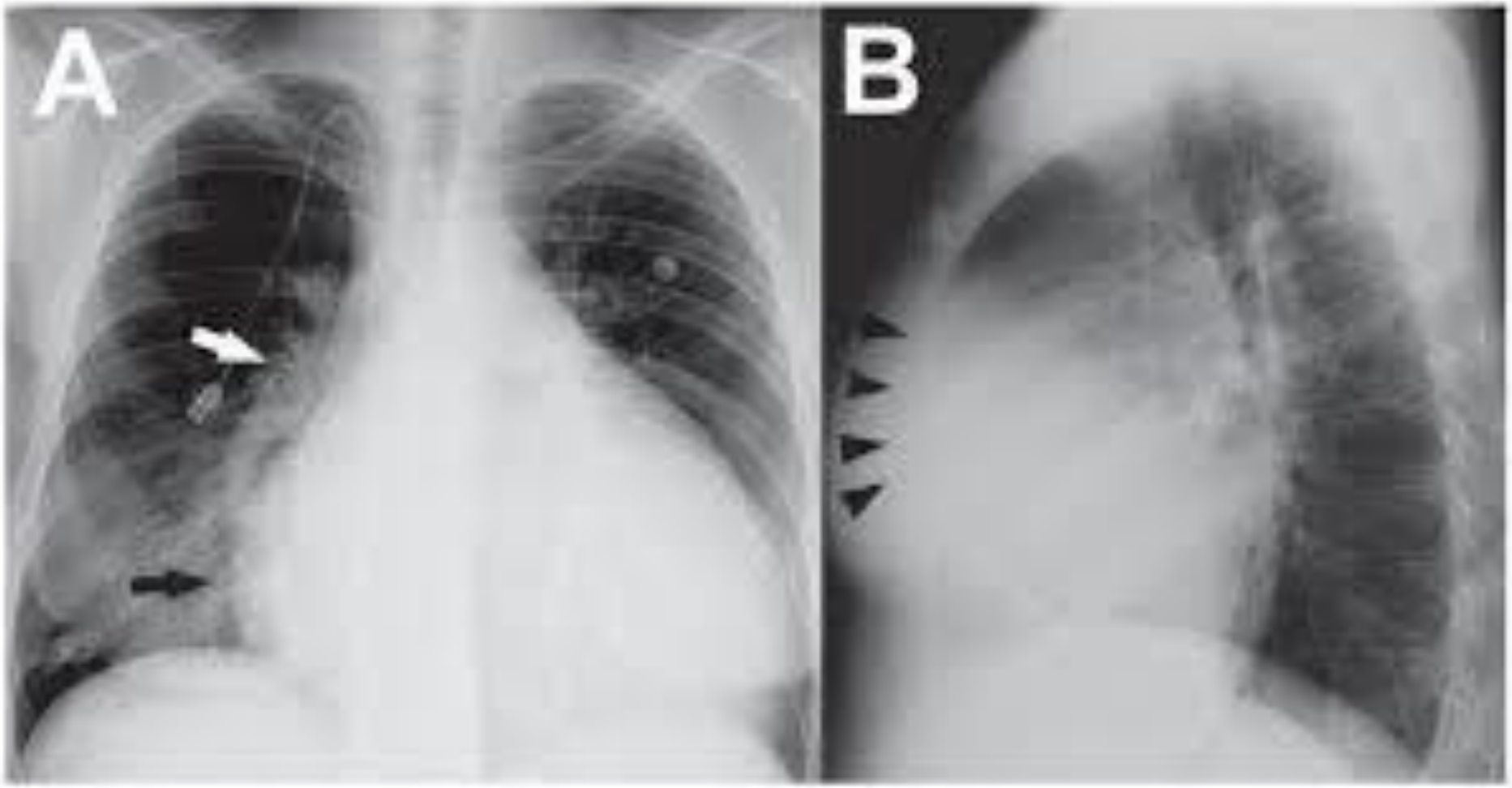

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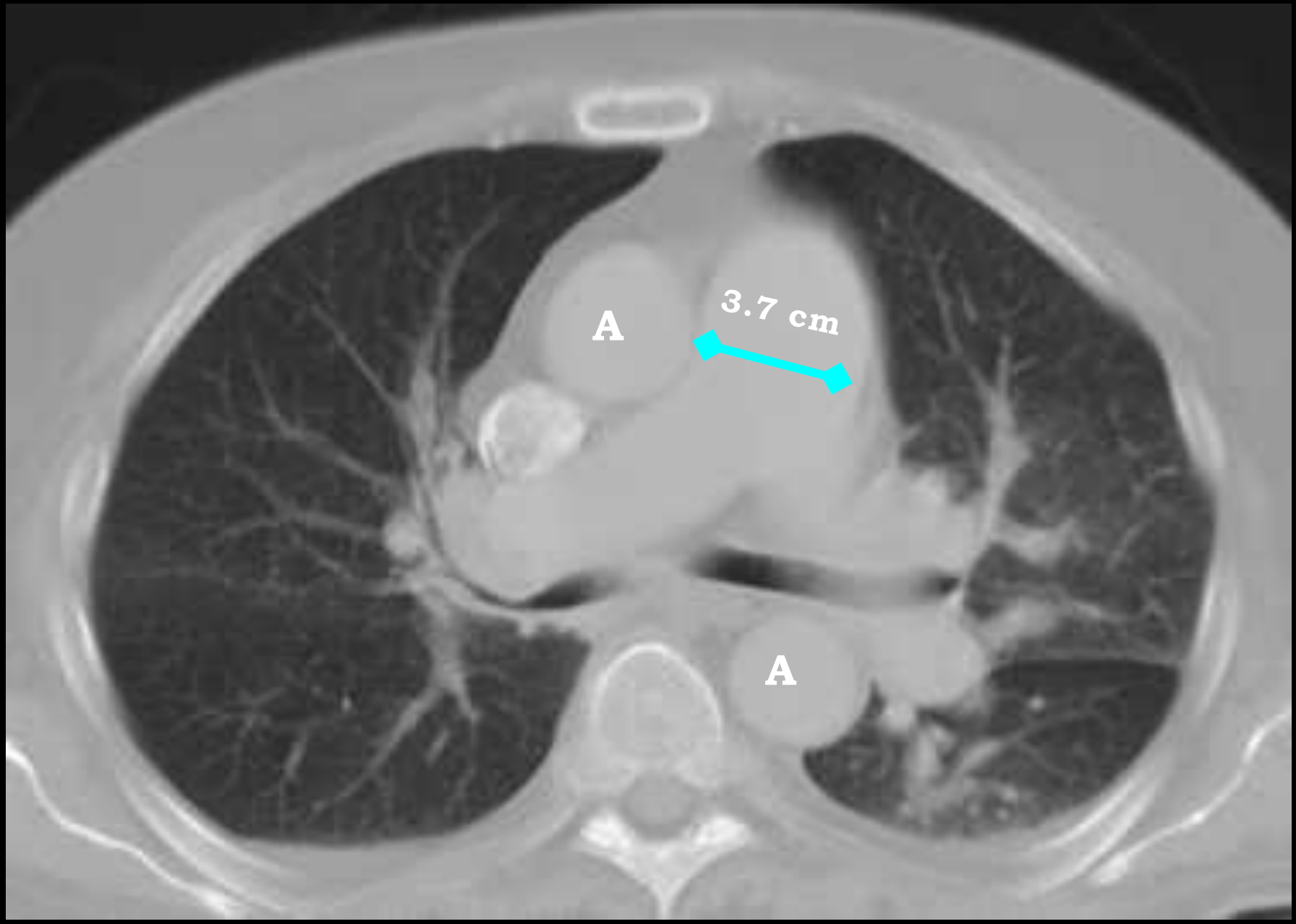

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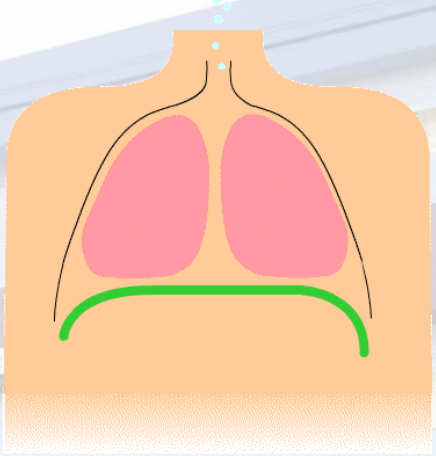
Estudio funcional respiratorio



Test prácticamente normales

Reducción leve de los volúmenes pulmonares y de la capacidad de difusión de monóxido de carbono

Test de función pulmonar anormal

si reducidos: enfermedad pulmonar

Los gases en sangre arterial en reposo, evidencian PaO2 normal o levemente disminuida si hipoxia: enfermedad cardiaca

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Test de respiración única para la capacidad de difusión de monóxido de carbono (DLCO) y su interpretación en enfermedades autoinmunes. Aplicación en la práctica clínica

Tabla 2. Grados de severidad y porcentaje predicho de DLCO.

Grado de severidad	% predicho DLCO
Leve	> 60% y < LIN
Moderado	40-60%
Severo	< 40%

LIN: límite inferior de lo normal

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Actualización en clasificación de la hipertensión pulmonar

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* Enfermedad veno oclusiva pulmonar con o sin haemangiomatosis capilar
* Secundaria a falla ventricular izquierda
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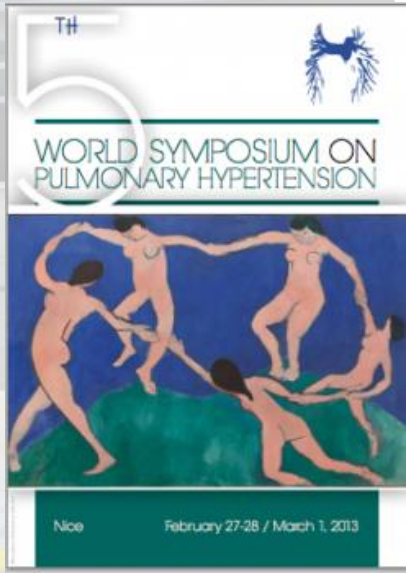

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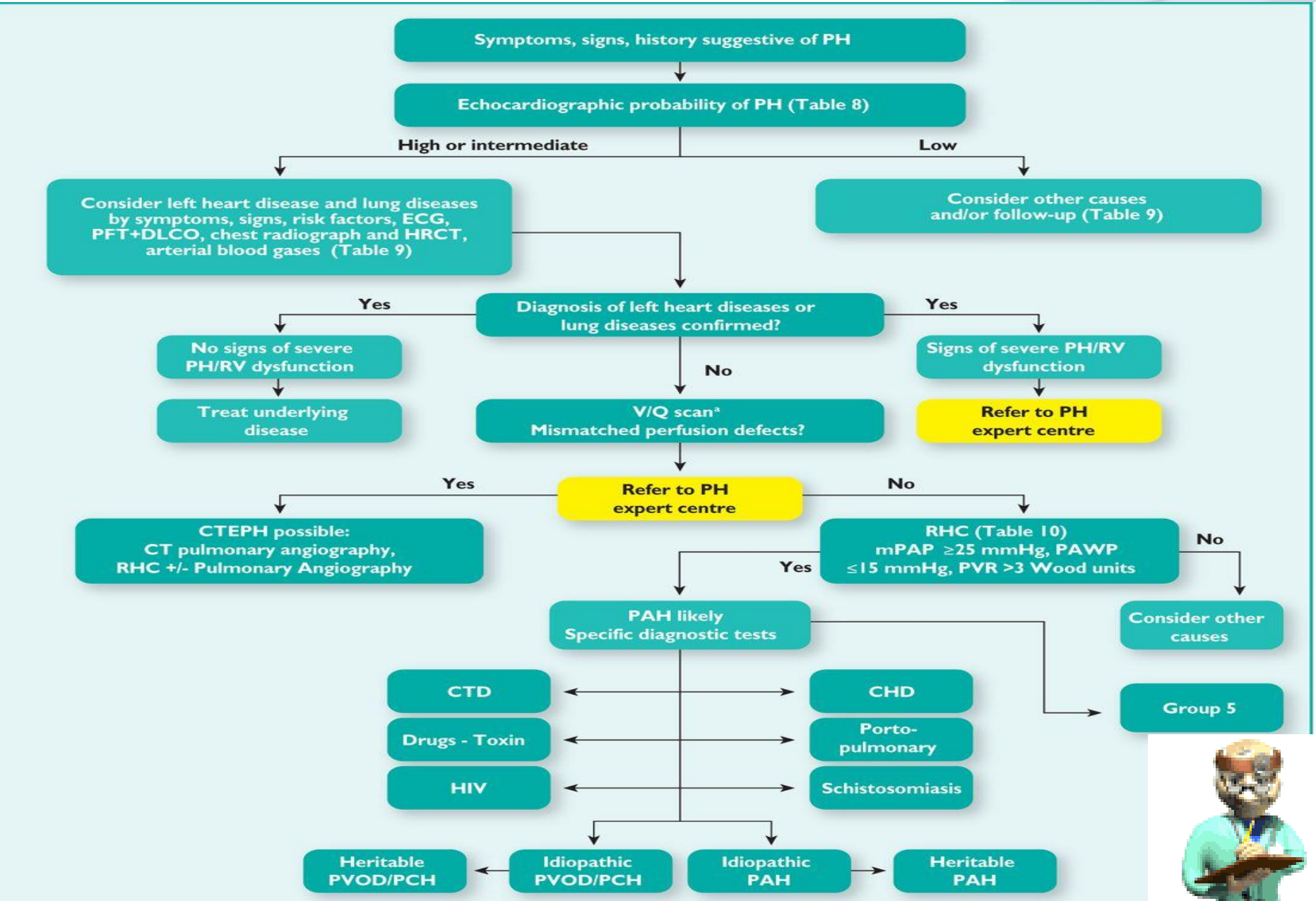

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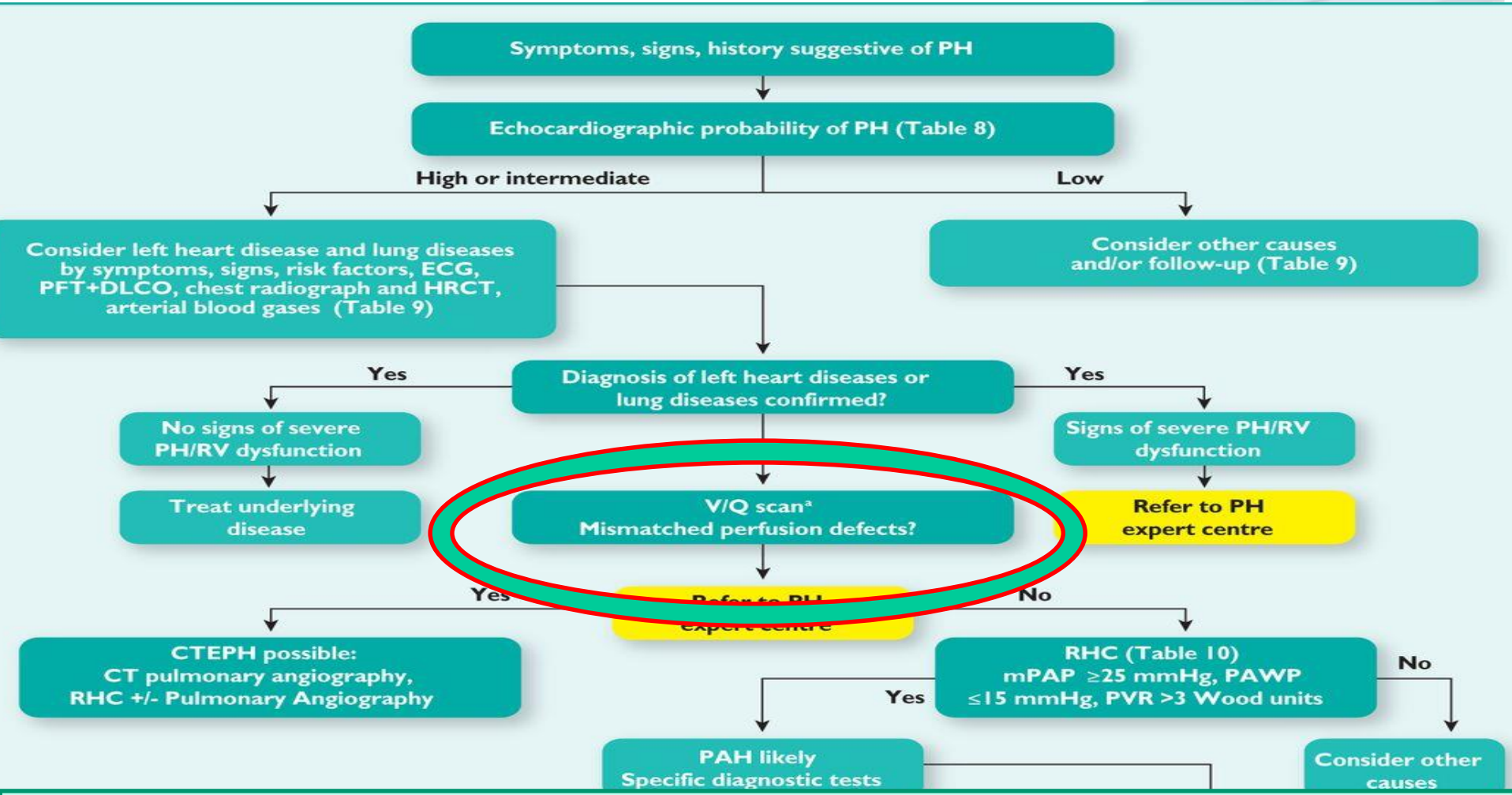

PULMONES Y CORAZON SANO…… CENTELLO V/Q

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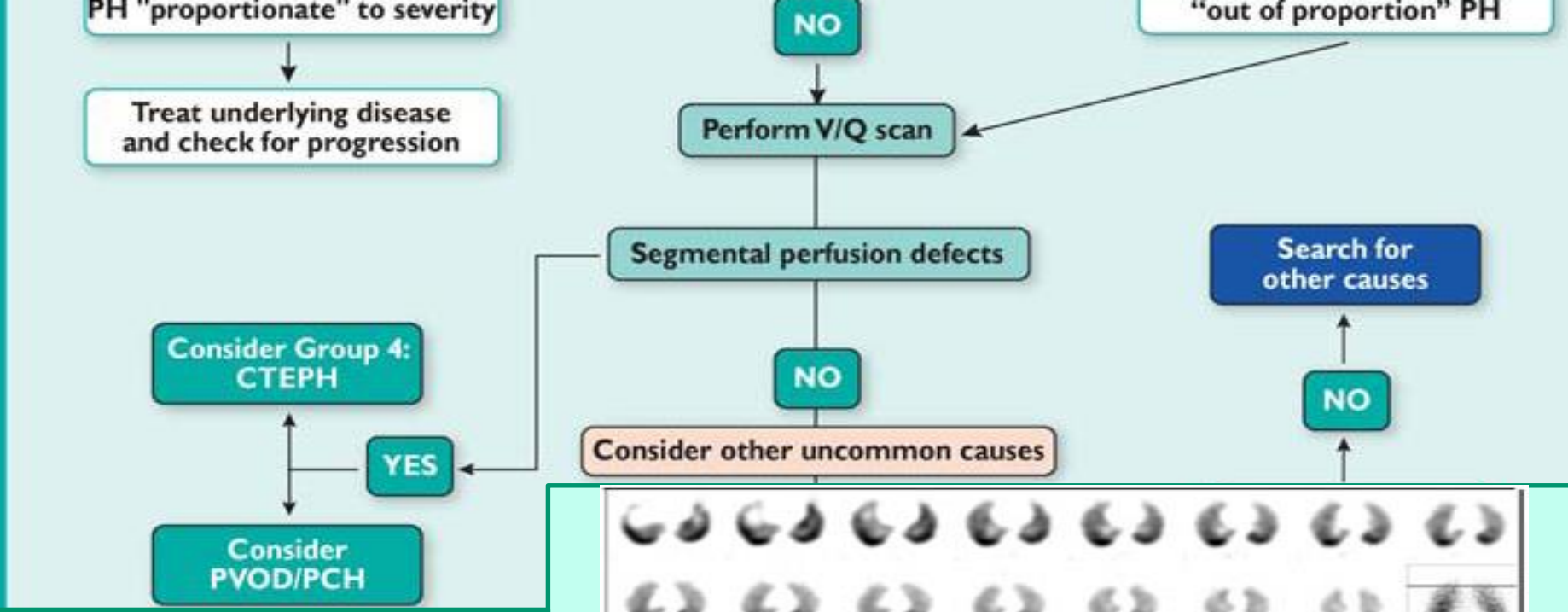
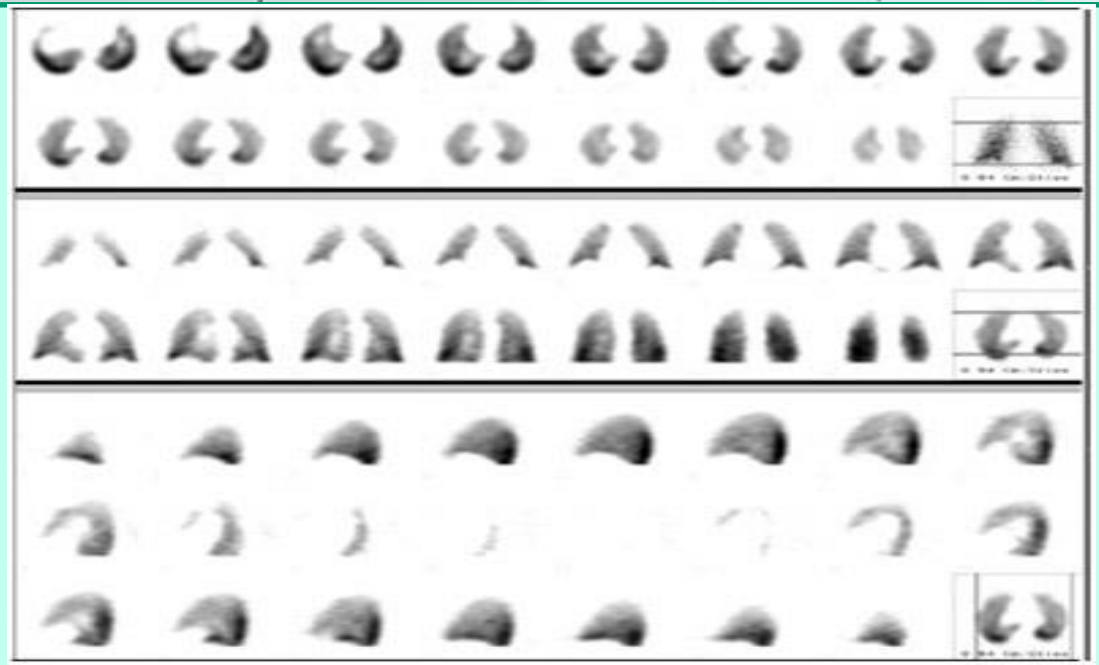


sensibilidad entre el 90% y el 100%, y una especificidad entre el 95% y el 100% para diferenciar una HAP secundaria a patología tromboembólica crónica de la HAP primaria o idiopática.



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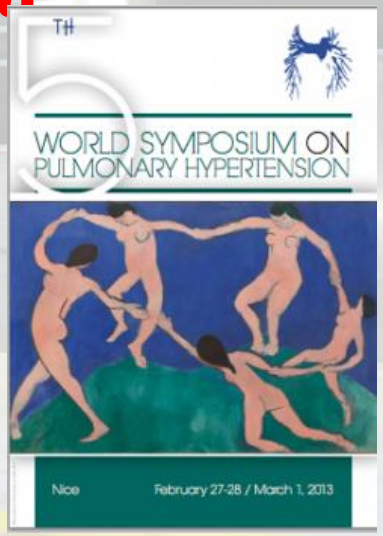

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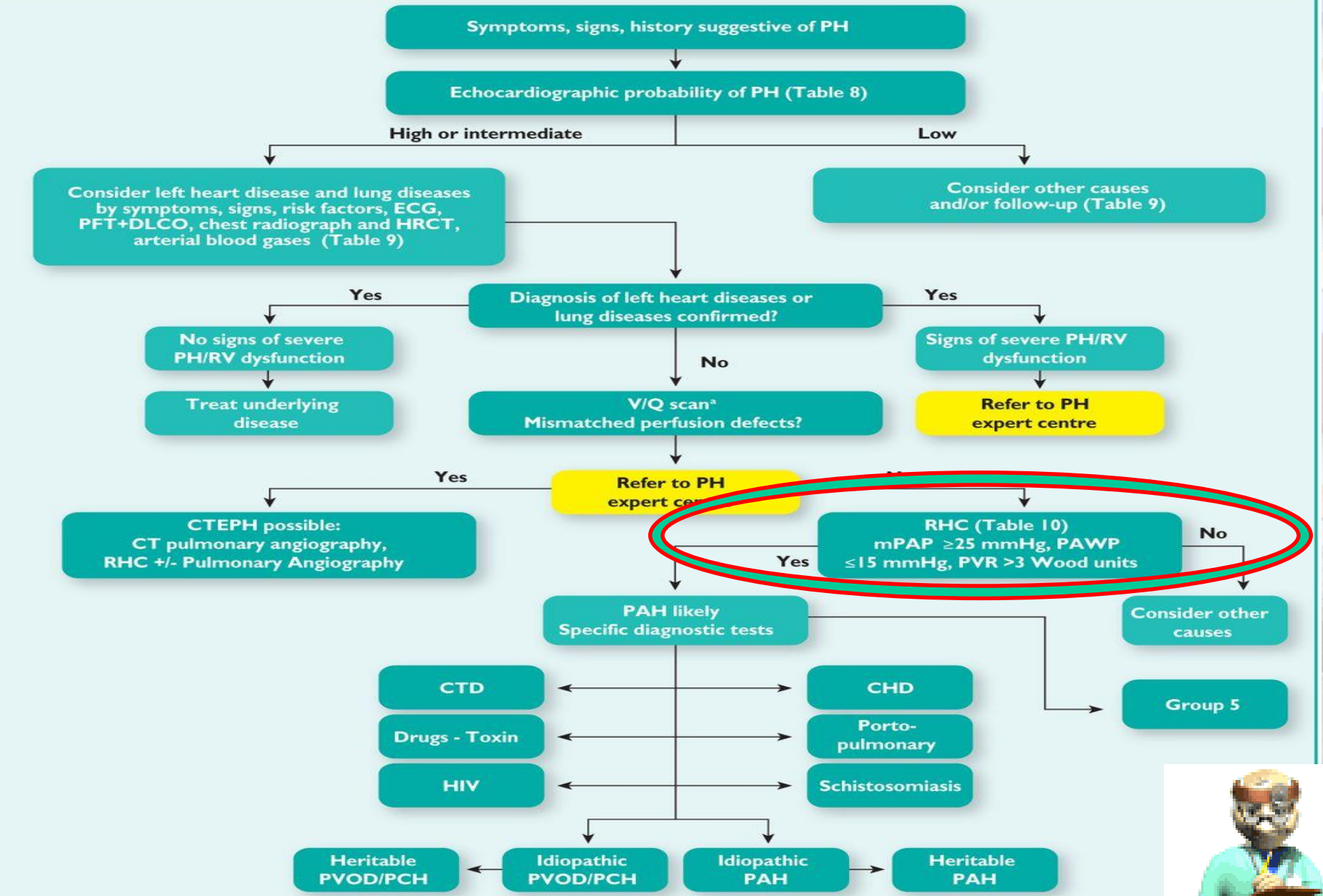

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T pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.

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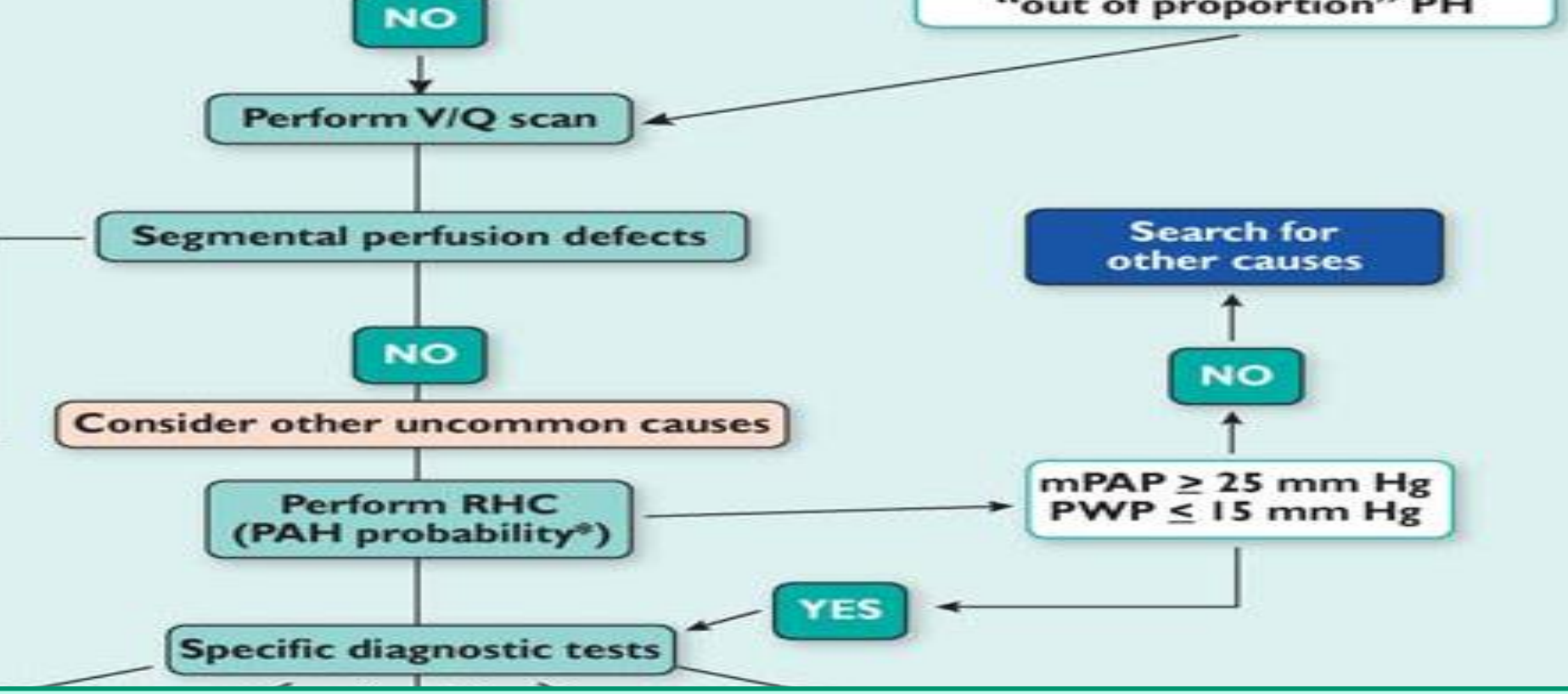

Angiografía pulmonar. Y cateterismo derecho Se halla indicada la algunos pacientes, sobre todo cuando se piense resolución de la HAP secundaria a TEP estudio que hagamos habitualmente, y presenta riesgos.

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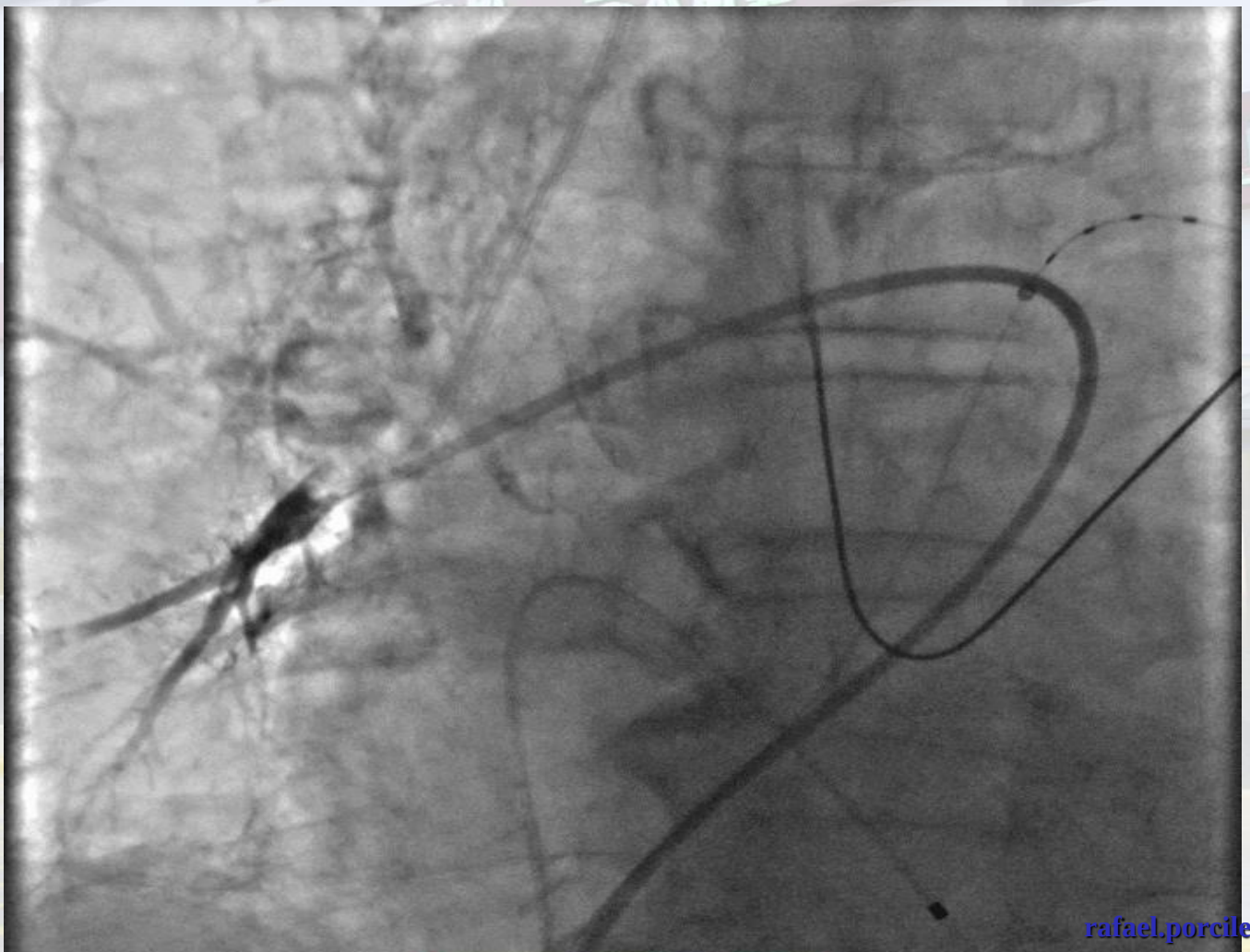
INTERRUPCION DE LA CIRCULACION PULMONAR



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ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

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Recommendations	Class[a]	Level[b]	Ref.[c]
RHC is recommended to confirm the diagnosis of pulmonary arterial hypertension [group 1] and to support treatment decisions	I	C	
In patients with PH, it is recommended to perform RHC in expert centres [see section 12] as it is technically demanding and may be associated with serious complications	I	B	[69]
RHC should be considered in pulmonary arterial hypertension [group 1] to assess the treatment effect of drugs [Table 16]	IIa	C	
RHC is recommended in patients with congenital cardiac shunts to support decisions on correction [Table 24]	I	C	
RHC is recommended in patients with PH due to left heart disease [group 2] or lung disease [group 3] if organ transplantation is considered	I	C	
When measurement of PAWP is unreliable, left heart catheterization should be considered to measure LVEDP	IIa	C	
RHC may be considered in patients with suspected PH and left heart disease or lung disease to assist in the differential diagnosis and support treatment decisions	IIb	C	
RHC is indicated in patients with CTEPH [group 4] to confirm the diagnosis and support treatment decisions	I	C	

Recomendaciones de cateterismo derecho

-Indicado en todos los pacientes con hipertensión pulmonar a los fines confirmatorios I C

-Confirmación de efectividad terapéutica IIa C

-Confirmación de progresión de la patología IIa C

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CATETER EN LA AD

REGISTRO DE PERSION AD

CATETER EN EL VD

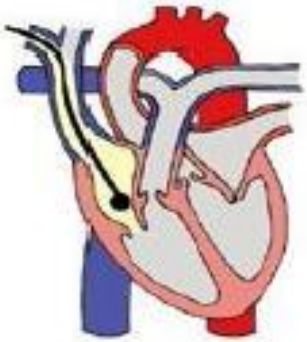
REGISTRO DE PRESSION DEL VD

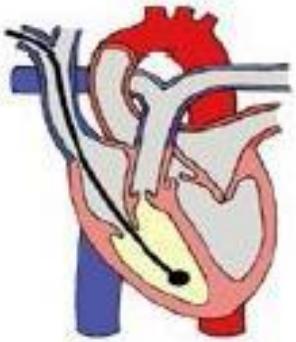
CATETER EN LA AP

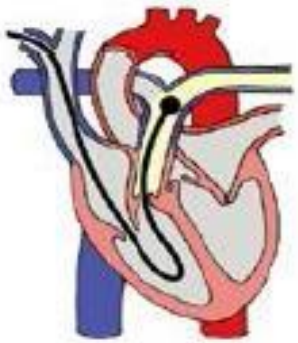
REGISTRO DE PRESSION DE LA AP

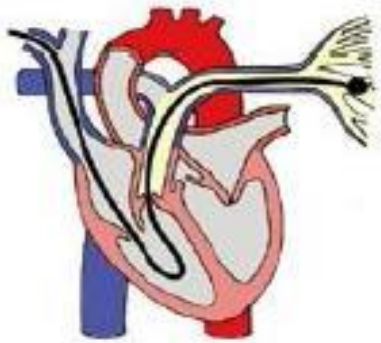
CATETER ENCLAVADO

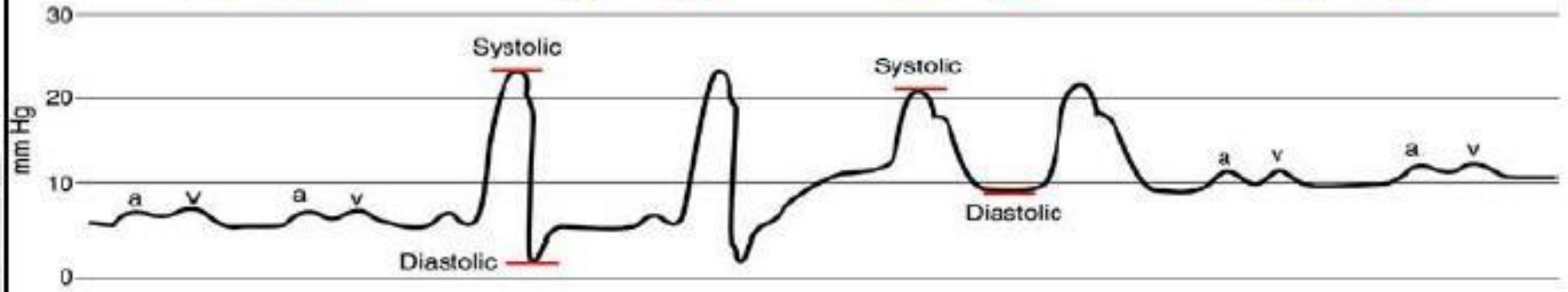
REGISTRO DE PRESION DE LA AI









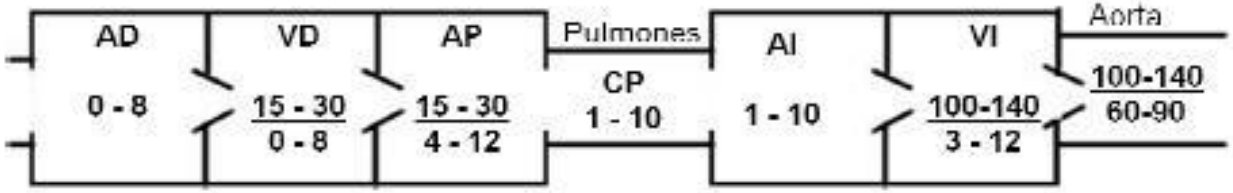


Right atrial pressure
0–8 mm Hg

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



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CATETER EN LA AD

REGISTRO DE PERSION AD

CATETER EN EL VD

REGISTRO DE PRESSION DEL VD

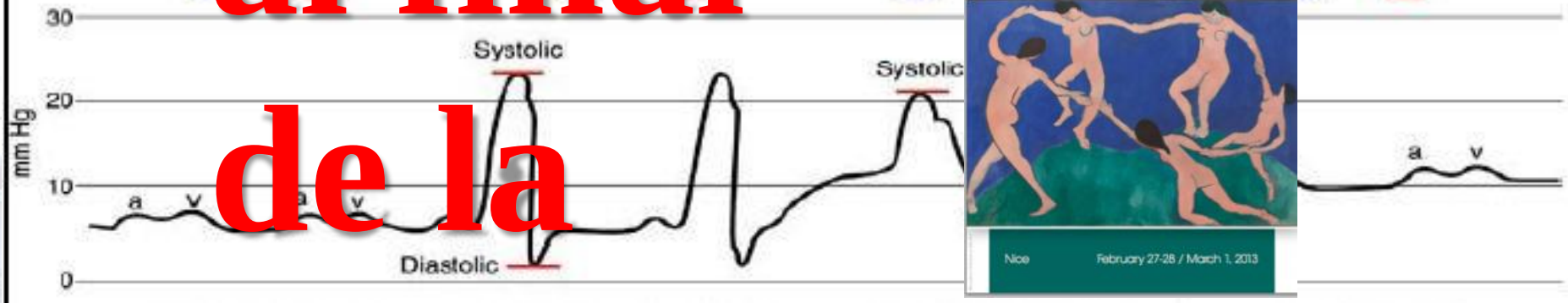
CATETER EN LA AP

REGISTRO DE PRESSION DE LA AP

CATETER ENCLAVADO

REGISTRO DE PRESION DE LA AI

La PCP se mide al final de la ESPIRACION

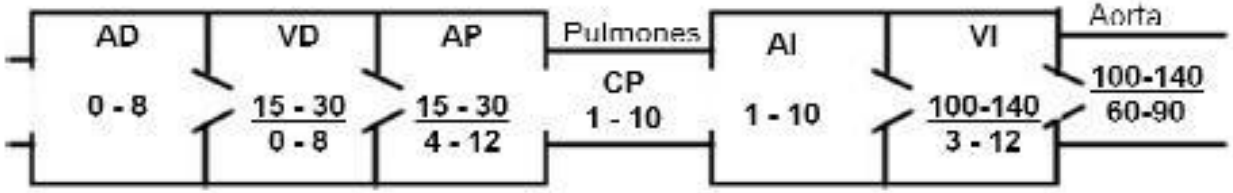


Right atrial pressure
0–[illegible] mm Hg

Right ventricular pressure
S[illegible] 20–[illegible] mm Hg
D[illegible]: 0–[illegible] mm Hg

Pulmonary artery pressure
Systolic: 2[illegible]30 mm Hg
Diastolic: 8[illegible]15 mm Hg

Pulmonary artery wedge pressure
[illegible]–12 mm Hg



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¿Es solo hipertensión pulmonar pre capilar?

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Eur Respir J. 2013 Jan;41(1):217-23.

The transpulmonary pressure gradient for the diagnosis of pulmonary vascular disease.

The transpulmonary pressure gradient (TPG), defined by the **difference** between **mean pulmonary arterial pressure** (P(pa)) and left atrial pressure (P(la); commonly estimated by pulmonary capillary **wedge pressure:** P(pcw)) has been recommended for the detection of intrinsic pulmonary vascular disease in left-heart conditions associated with increased pulmonary venous pressure

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Eur Respir J. 2013 Jan;41(1):217-23.

The transpulmonary pressure gradient for the diagnosis of pulmonary vascular disease.

a TPG of >12 mmHg would result in a diagnosis of "out of proportion" pulmonary hypertension

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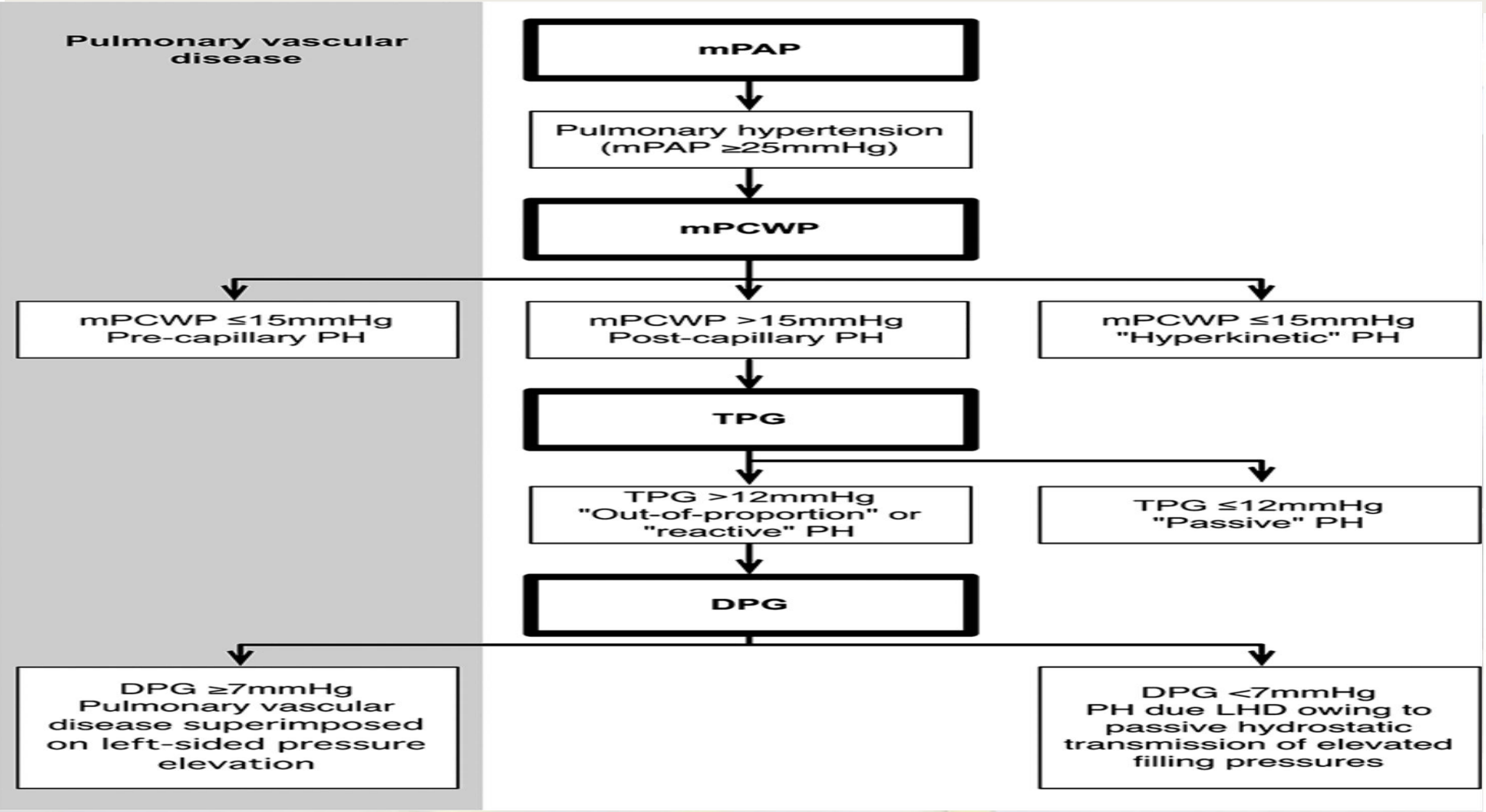
From: Diastolic Pulmonary Vascular Pressure GradientDiastolic Gradient and Prognosis: A Predictor of Prognosis in "Out-of-Proportion" Pulmonary Hypertension

Chest. 2013;143(3):758-766. doi:10.1378/chest.12-1653



Hemodynamic algorithm. Hemodynamic algorithm for the diagnosis of a high-risk subgroup of "out-of-proportion" PH. The gray shaded area indicates conditions where pulmonary vascular disease is expected. LHD = left-sided heart disease. See Figure 1 and 2 legends for expansion of other abbreviations.

From: Diastolic Pulmonary Vascular Pressure GradientDiastolic Gradient and Prognosis: A Predictor of Prognosis in "Out-of-Proportion" Pulmonary Hypertension

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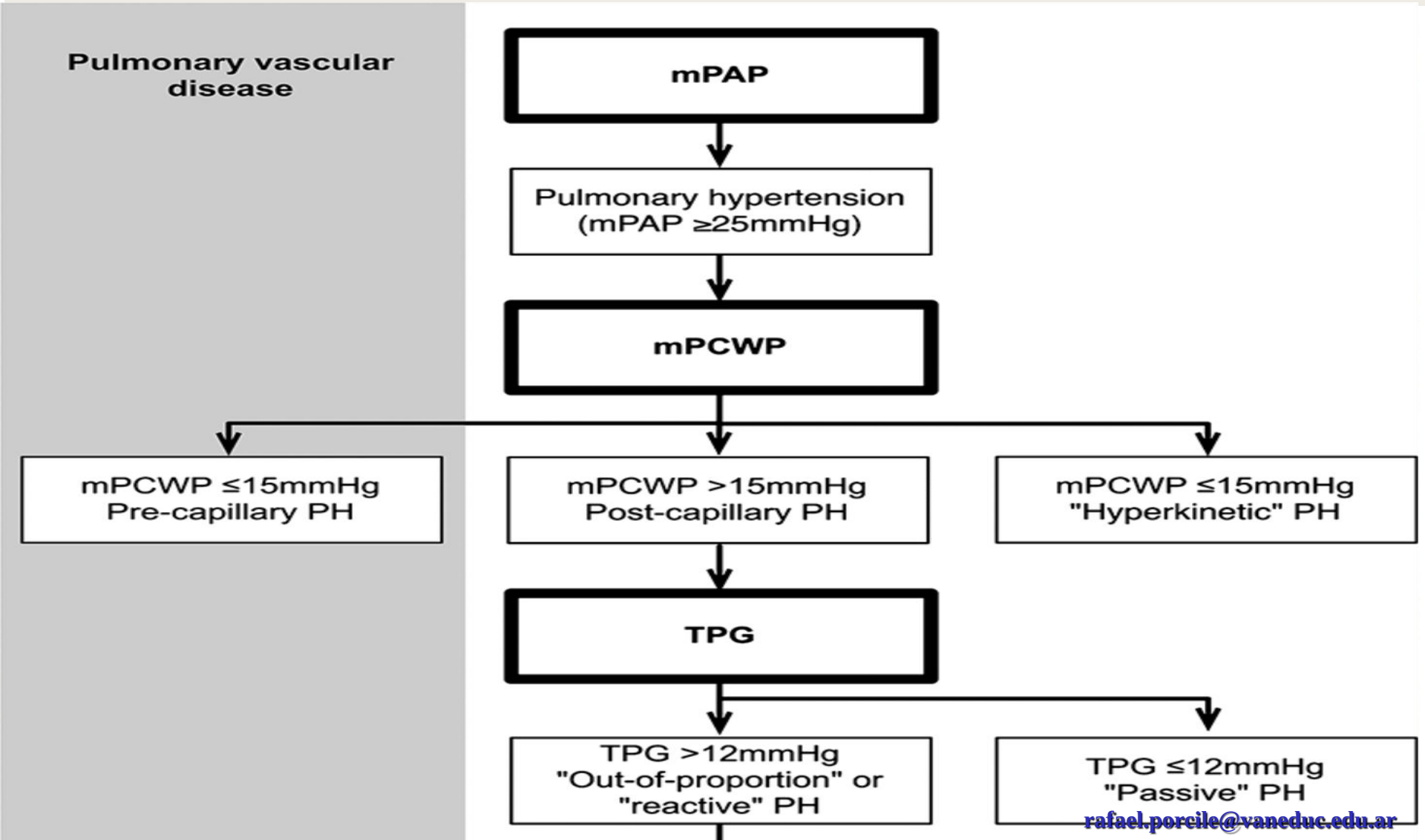
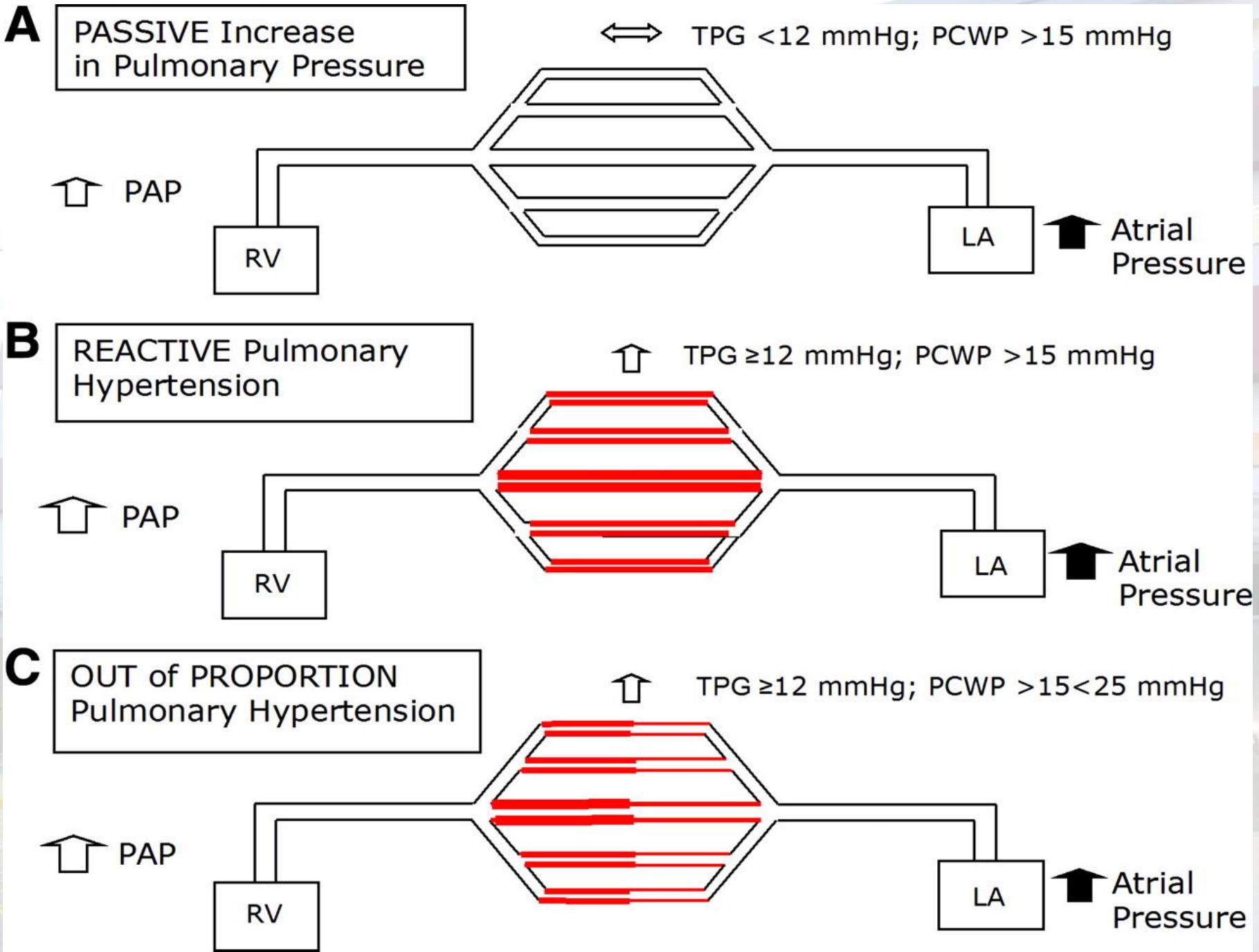


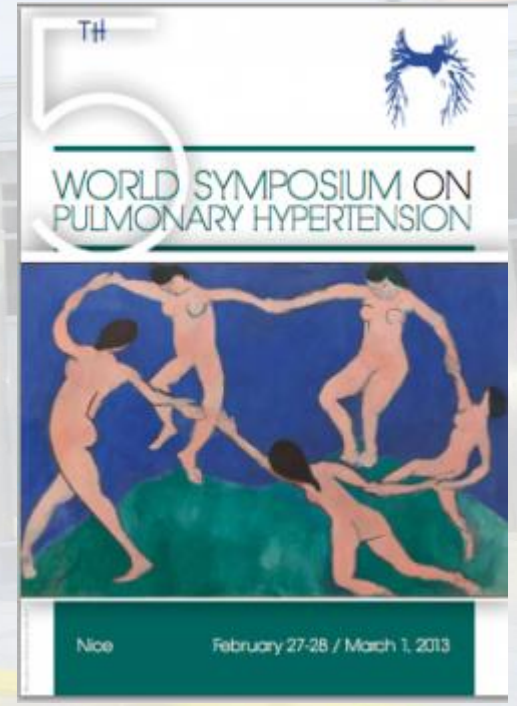

A
PASSIVE Increase in Pulmonary Pressure
TPG <12 mmHg; PCWP >15 mmHg
PAP
RV
LA
Atrial Pressure
B
REACTIVE Pulmonary Hypertension
TPG ≥12 mmHg; PCWP >15 mmHg
PAP
RV
LA
Atrial Pressure
C
OUT of PROPORTION Pulmonary Hypertension
TPG ≥12 mmHg; PCWP >15<25 mmHg
PAP
RV
LA
Atrial Pressure

Debe tomarse el **DPG**

Gradiente pulmonar diastólico

Es la diferencia entre diastólica pulmonar y W

Si es mayor a 7 hay Hipertensión pulmonar



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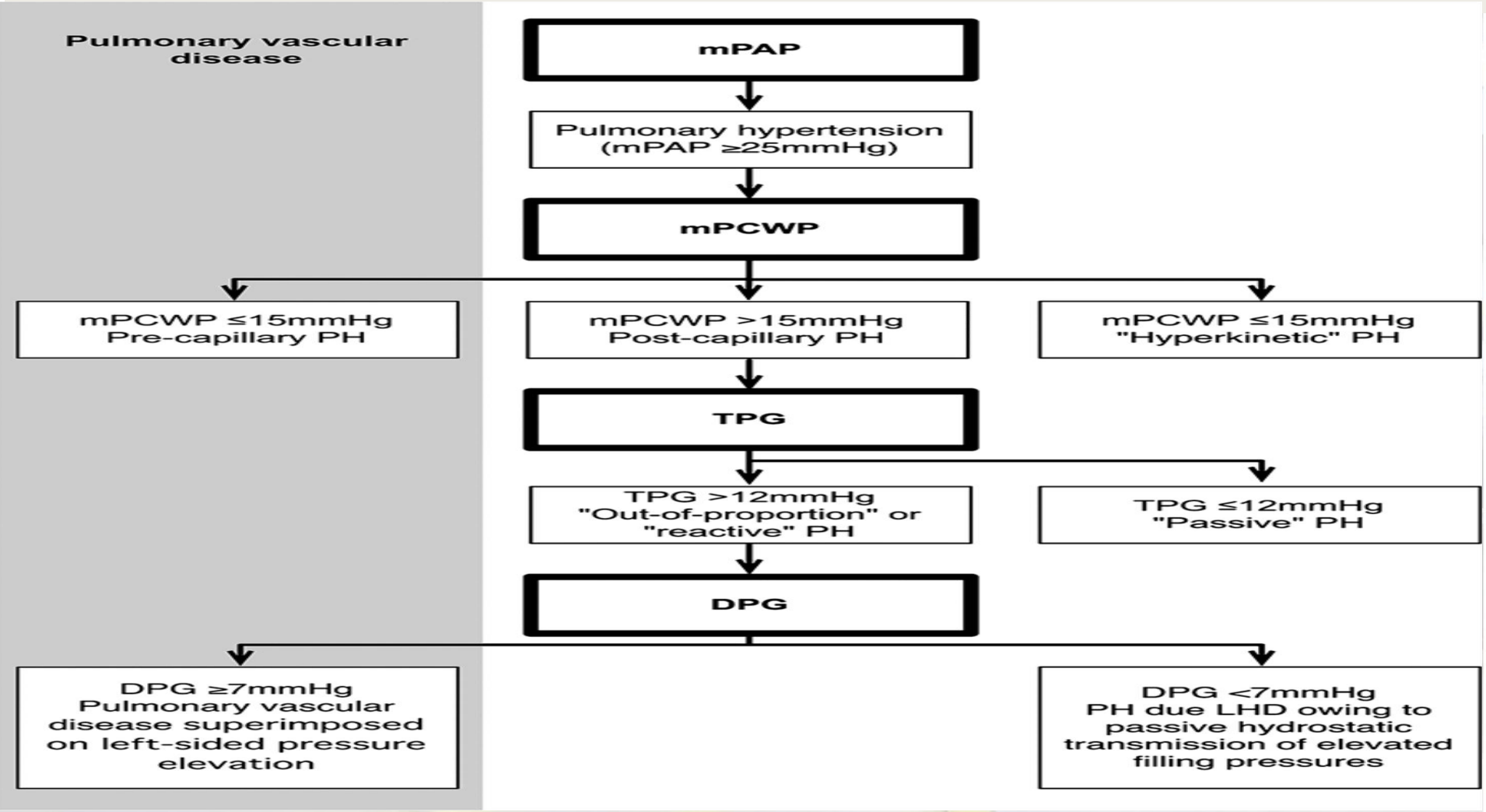

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Test de vaso reactividad pulmonar

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Recommendations	Class[a]	Level[b]	Ref.[c]
Vasoreactivity testing is indicated only in expert centres	I	C	69
Vasoreactivity testing is recommended in patients with IPAH, HPAH and PAH associated with drugs use to detect patients who can be treated with high doses of a CCB	I	C	84,85
A positive response to vasoreactivity testing is defined as a reduction of mean PAP $\geq$10 mmHg to reach an absolute value of mean PAP $\leq$40 mmHg with an increased or unchanged cardiac output	I	C	85,86
Nitric oxide is recommended for performing vasoreactivity testing	I	C	85,86
Intravenous epoprostenol is recommended for performing vasoreactivity testing as an alternative	I	C	85,86
Adenosine should be considered for performing vasoreactivity testing as an alternative	IIa	C	87,88
Inhaled iloprost may be considered for performing vasoreactivity testing as an alternative	IIb	C	89,90
The use of oral or intravenous CCBs in acute vasoreactivity testing is not recommended	III	C	
Vasoreactivity testing to detect patients who can be safely treated with high doses of a CCB is not recommended in patients with PAH other than IPAH, HPAH and PAH associated with drugs use and is not recommended in PH groups 2, 3, 4 and 5	III	C	

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1) Oxigeno al 100 % durante 30 minutos
2) Medición de swan completa
3) Suspender el Oxigeno
4) Nifedipina Sublingual 10 – 20 mg
5) Medición a los 15, 30 y 45 minutos después de administración hipotensión
6) Suspender la nifedipina esperar 60 minutos
7) Nitroglicerina Intravenosa 5-10 µg/min, con incremento gradual cada 10 min hasta noventa de sistólica
8) Medir swan
9) De no haber logrado l reducción de la tensión arterial pulmonar media de al menos 15 % Nitroprusiato Intravenosa 0.1 µg/kg/min con incrementos de 0.2 µg/kg/min
10) Medir swan
11) Esperar 60 minutos
12) De no haber logrado l reducción de la tensión arterial pulmonar media de al menos 15 %

Adenosina Intravenosa Inicio 50 µg /kg/min con incrementos de 50 µg Cefalea, náusea, dolor torácicocada 2-5 min hasta una dosis máxima de mareo, hormigueo500 µg/kg/min

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TEST DE VASORREACTIVIDAD

Positivo; Descenso de PAPm > 10 mm Hg. Consiguiendo una PAPm < 40 mm Hg. con GC normal o elevado

Tipos de Respuesta:

Pacientes que empeoran: Sospecha de EVOP o HC. Contraindicación para el uso de Vasodilatadores

Test negativo: Tto Vasodilatador según guías

Test positivo: Utilización de Bloqueantes de canales del Calcio y confirmar respuesta en 3 meses

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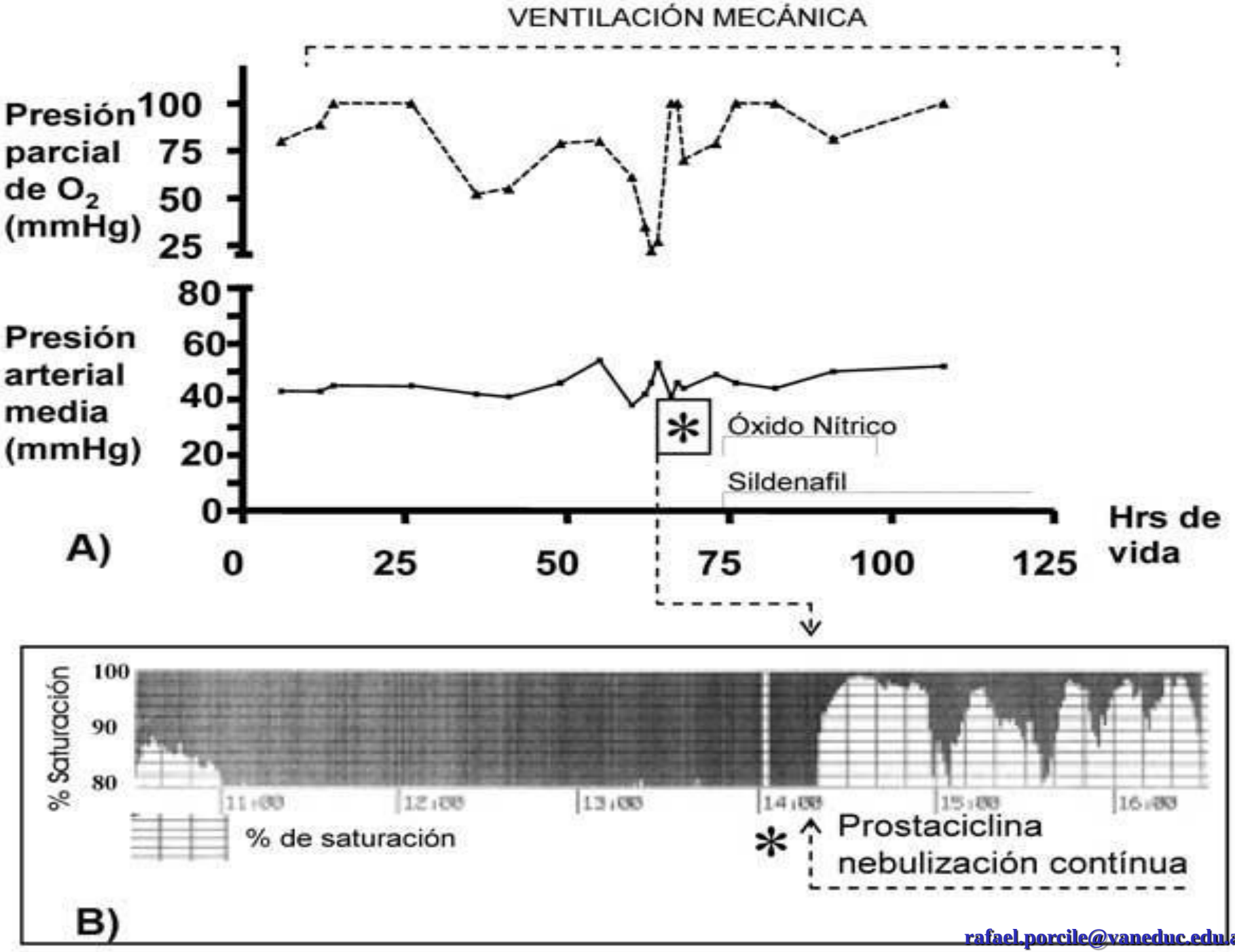

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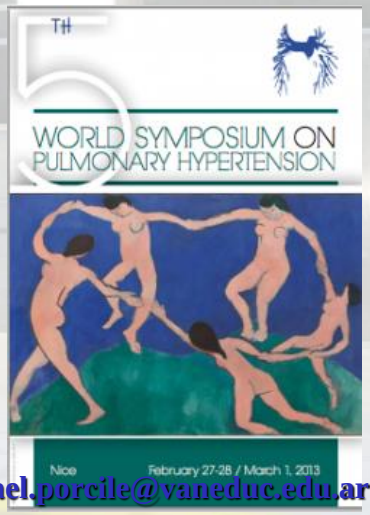
SE
DESCARTA
O
CONFIRMA

Actualización en clasificación de la hipertensión pulmonar

* **Hipertensión arterial pulmonar (PAH)**
* Enfermedad veno oclusiva pulmonar con o sin haemangiomatosis capilar
* Secundaria a falla ventricular izquierda
* Secundaria a enfermedad pulmonar con o sin hipoxemia
* Hipertensión pulmonar secundaria a tromboembolismo crónico
* Mecanismos poco claros o multifactorial



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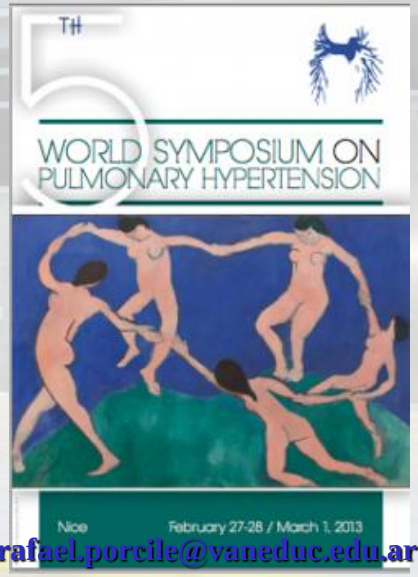
Clasificación de hipertensión ARTERIAL pulmonar

afección de arterias de menos de 500 micrones

- **1 Idiopática**
- **1.2 Heredables**
  - **1.2.1 BMPR2b receptor tipo2 proteína morphogenetica**
  - **1.2.2 ALK1, gen kinaa 1 like**
  - **1.2.3 hereditaria desconocida**
- **1.3 Inducida por drogas y toxinas**
- **1.4 Asociadas**
  - **1.4.1 Enfermedades del tejido conectivo**
  - **1.4.2 HIV i**
  - **1.4.3 hipertensión portal**
  - **1.4.4 cardiopatías congénitas**
  - **1.4.5 Schistosomiasis**
  - **1.4.6 anemia hemolítica crónica**
- **1.5 HAP persistente del recien nacido**



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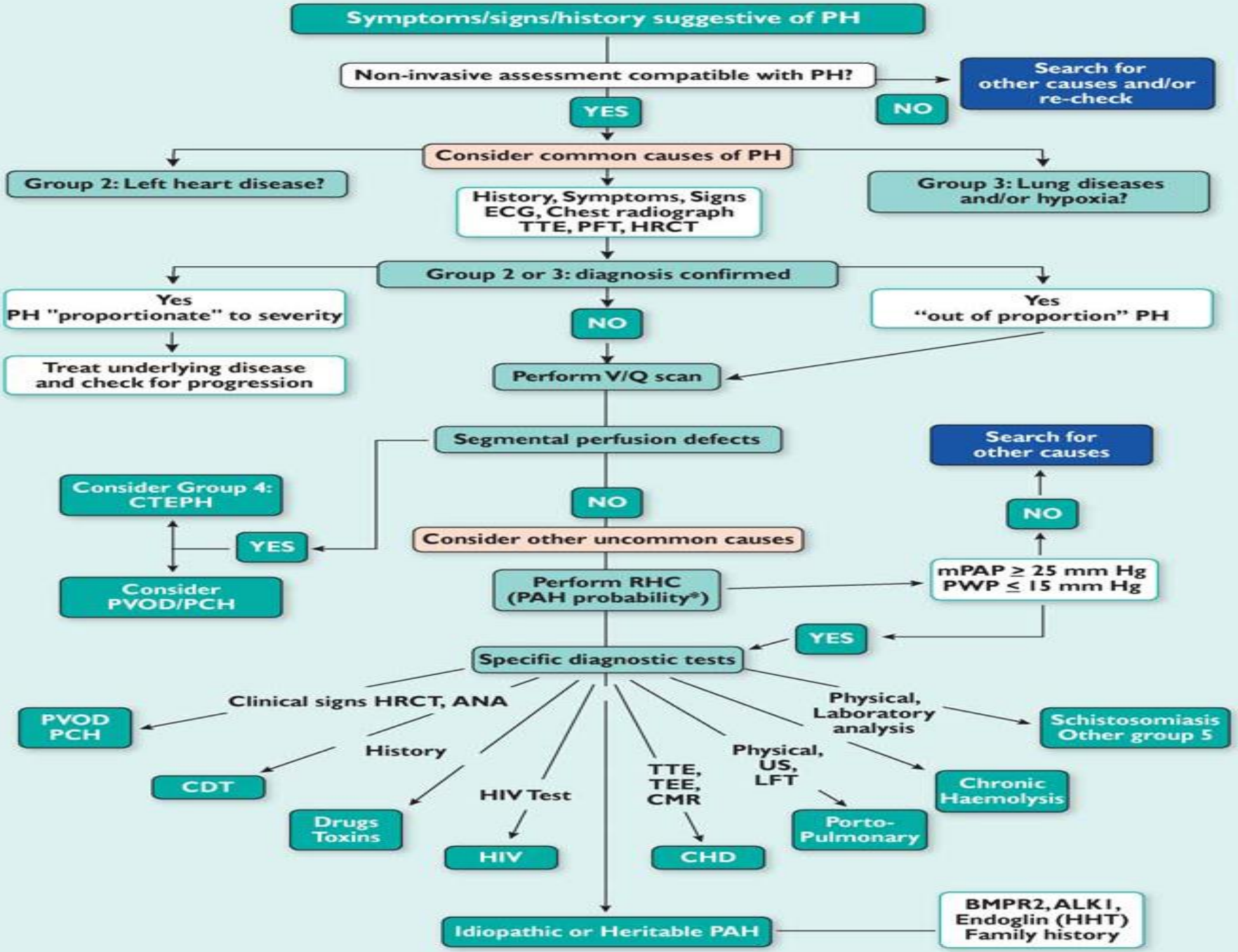
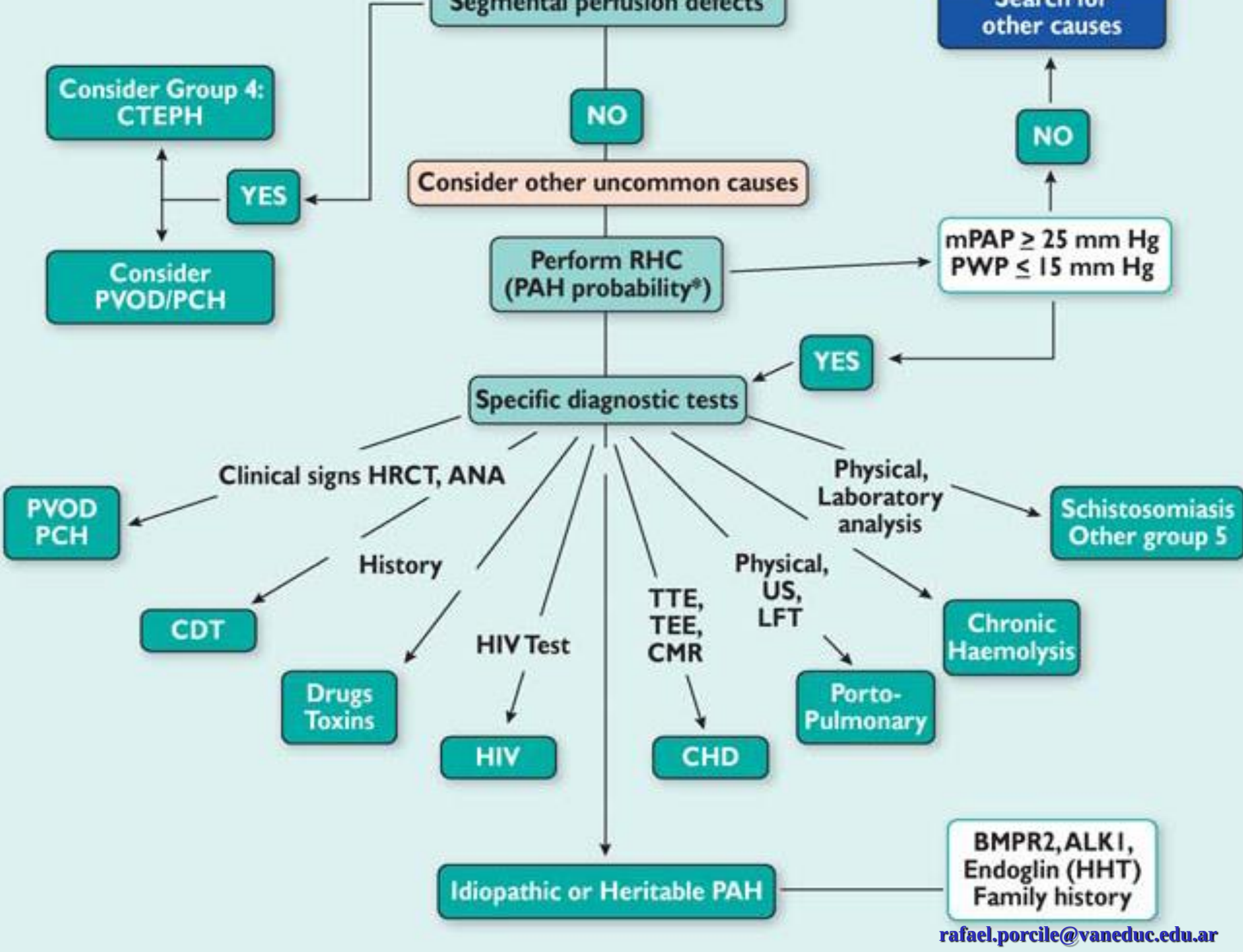

Symptoms/signs/history suggestive of PH
Non-invasive assessment compatible with PH?
NO
Search for other causes and/or re-check
YES
Consider common causes of PH
Group 2: Left heart disease?
History, Symptoms, Signs
ECG, Chest radiograph
TTE, PFT, HRCT
Group 3: Lung diseases and/or hypoxia?
Group 2 or 3: diagnosis confirmed
Yes
PH "proportionate" to severity
Treat underlying disease and check for progression
NO
Yes
"out of proportion" PH
Perform V/Q scan
Segmental perfusion defects
Search for other causes
Consider Group 4: CTEPH
YES
NO
NO
Consider other uncommon causes
Consider PVOD/PCH
Perform RHC (PAH probability*)
mPAP ≥ 25 mm Hg
PWP ≤ 15 mm Hg
YES
Specific diagnostic tests
Clinical signs HRCT, ANA
PVOD PCH
History
CDT
Drugs Toxins
HIV Test
HIV
TTE, TEE, CMR
CHD
Physical, US, LFT
Porto-Pulmonary
Physical, Laboratory analysis
Schistosomiasis Other group 5
Chronic Haemolysis
Idiopathic or Heritable PAH
BMPR2, ALK1, Endoglin (HHT) Family history



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HIV Y CORAZÓN

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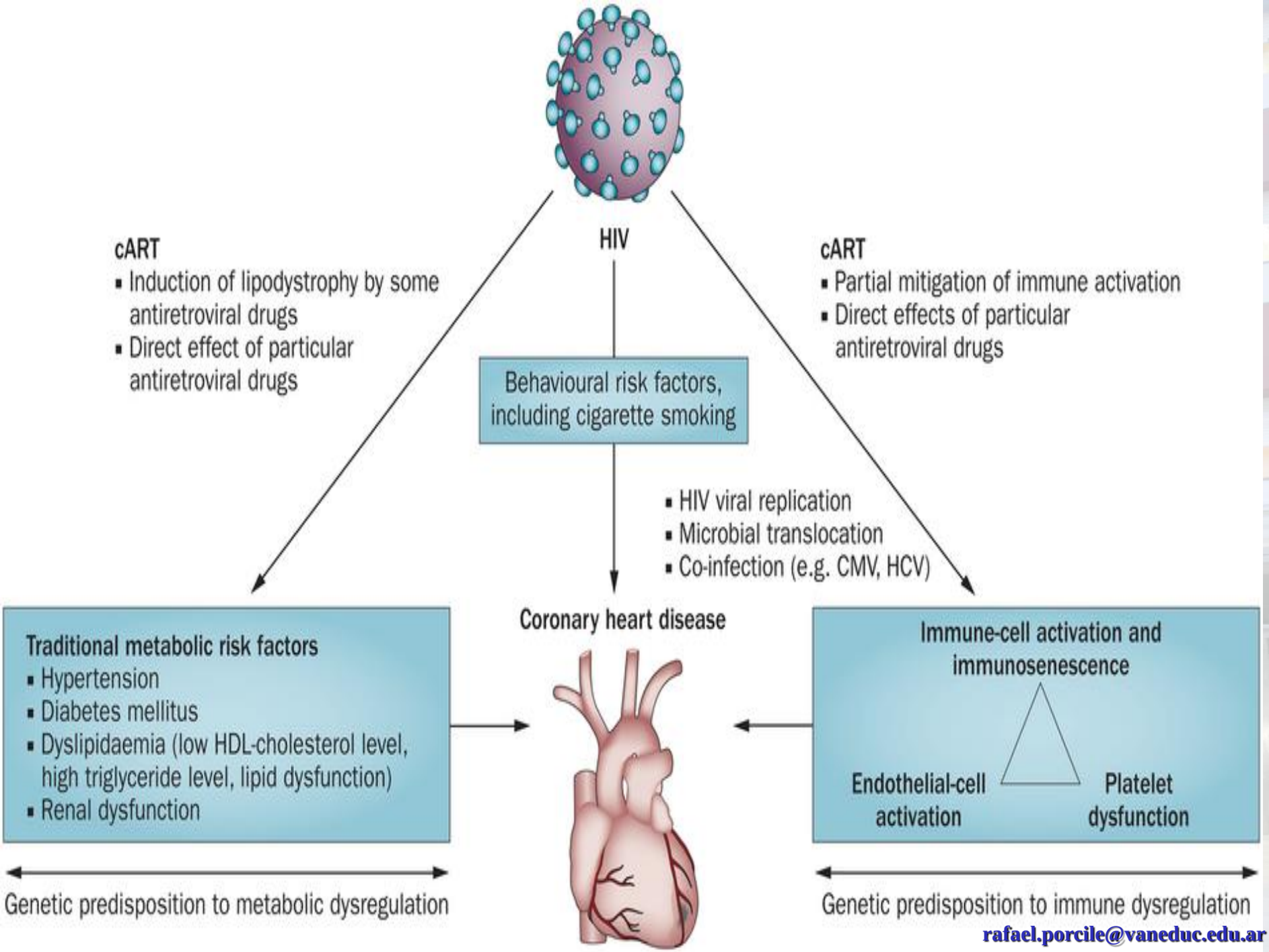

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Chapter 12

HIV-Associated Pulmonary Hypertension

G. Barbaro

About 14 years ago, Kim and Factor reported the first case of HIV-associated pulmonary hypertension [1]. Since then more than 131 cases have been described in the literature [2]. For this reason, HIV-associated pulmonary hypertension has been included as a definite cause of precapillary pulmonary hypertension according to the executive summary of the World Health Organization (WHO) [3]. The incidence of HIV-associated pulmonary hypertension is 1 in 200, much higher than the 1 in 200,000 found in the general population [3]. No differences have been found in the clinical, histologic, and hemodynamic features between patients with HIV-associated pulmonary hypertension and HIV-uninfected patients affected by primary pulmonary hypertension.

Pathogenesis of HIV-Associated Pulmonary Hypertension

The histopathology of HIV-associated pulmonary hypertension is similar to that of primary pulmonary hypertension. The most common alteration in HIV-associated pulmonary hypertension is plexogenic pulmonary arteriopathy (Fig. 1), while thrombotic pulmonary arteriopathy and pulmonary veno-occlusive disease are more rare histologic findings. This observation may suggest that similar etiopathogenetic mechanisms are at the basis of both HIV-associated pulmonary hypertension and primary pulmonary hypertension.

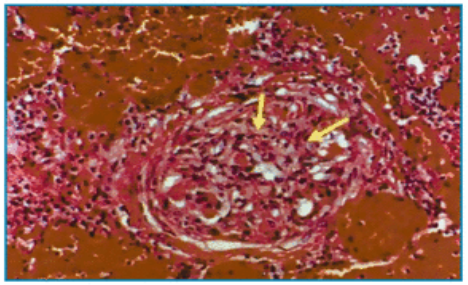

Fig. 1 Plexogenic pulmonary arteriopathy (*arrows*) in a patient with HIV-associated pulmonary hypertension (autopsy specimen). H&E, x20

The finding of an increased incidence of pulmonary hypertension in HIV-infected patients was at first related to viral infection. Although a direct role of HIV-1 in HIV-associated pulmonary hypertension has not been demonstrated [4, 5], several indirect mechanisms may link HIV infection to the pulmonary vascular changes. The principal pathogenetic hypotheses formulated for development of HIV-associated pulmonary hypertension with related clinical evidence are reported in Table 1.

Clinical Manifestations and Diagnosis of HIV-Associated Pulmonary Hypertension

In the largest clinical series of HIV-associated pulmonary hypertension, 47–54% of all

HIPERTENSION PULMONAR

- EL HIV NO INFECTA LAS CELULAS ENDOTELIALES PULMONARES
- EL HIV INDUCE INDIRECTAMENTE EL DESARROLLO DE H.P AL AUMENTAR CITOQUINAS INFLAMATORIAS EN LOS LINFOCITOS Y MACROFAGOS ALVEOLARES INFECTADOS
- ESTUDIOS RECIENTES HAN DEMOSTRADO QUE EL HIV Y LA PROTEINA gp120 AUMENTAN LA SECRECION DE ENDOTELINA-1

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HIV

ET-1 PDGF IL6 IL1β TNFα

Endothelial cell ⟵ **Endothelial cell** — **Smooth-muscle cells**

- Reduction of NO synthesis
- Proliferation
- Vasoconstriction
- Proliferation
- Migration of smooth muscle cells

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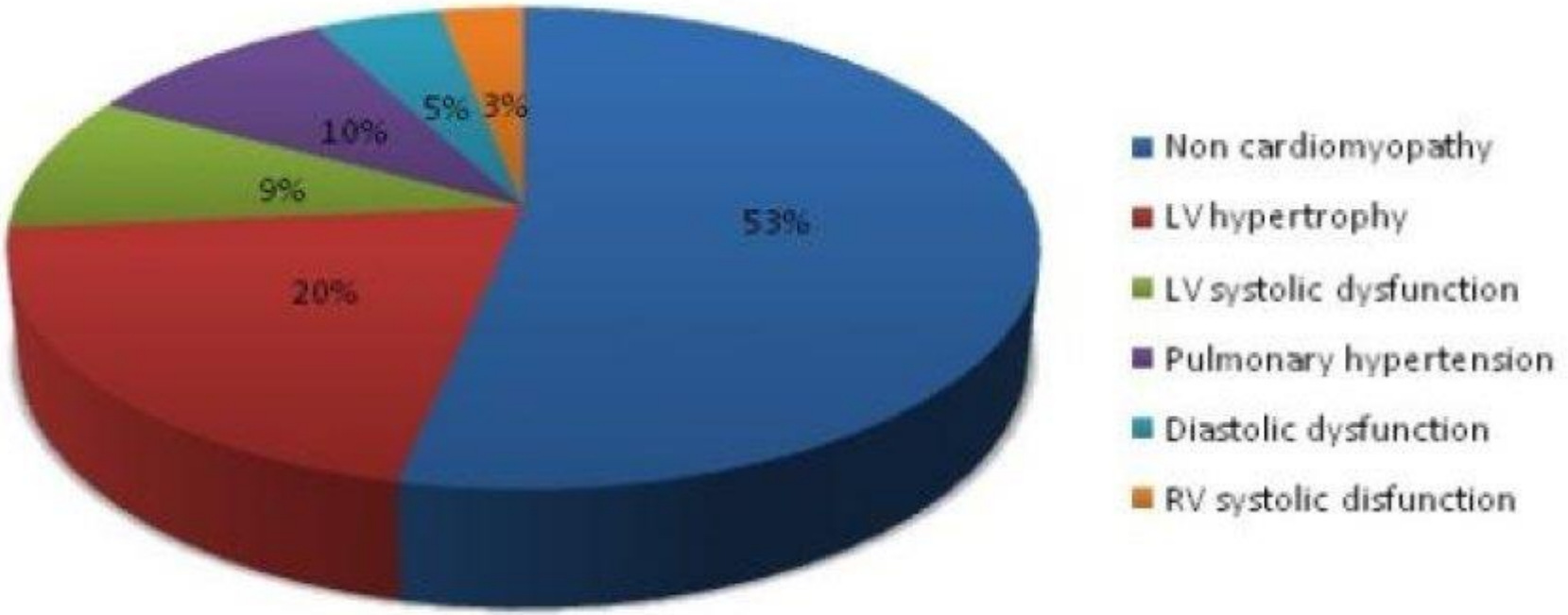
Prevalence of structural heart disease in HIV patients



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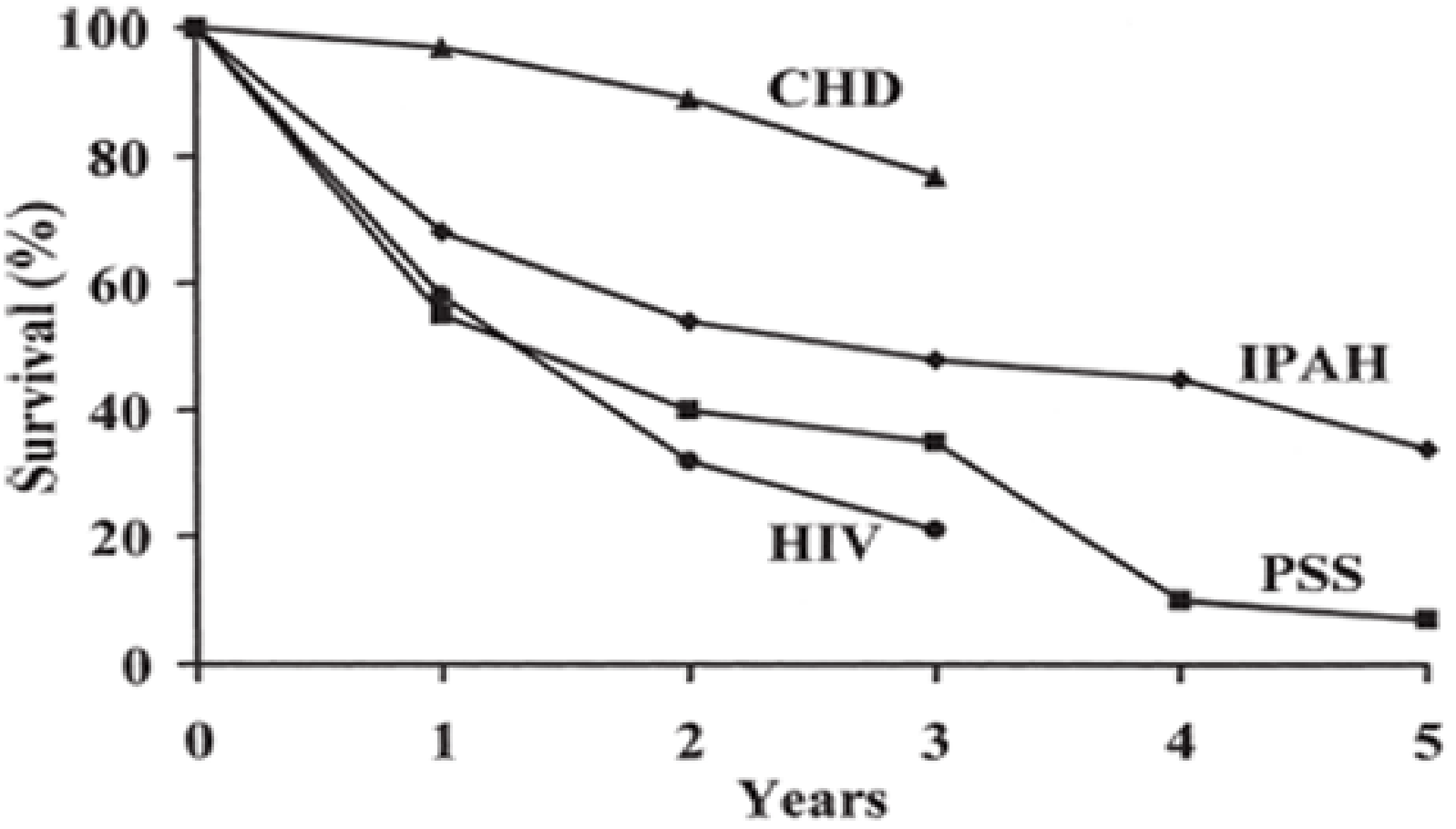
Sobrevida de la Hipertensión pulmonar según etiología

Survival by pulmonary arterial hypertension (PAH) condition. IPAH, idiopathic pulmonary arterial hypertension (D'Alonzo et al[3]); CHD, congenital heart disease (Hopkins et al[13]); PSS, progressive systemic sclerosis (Stupi et al[9]); HIV, human immunodeficiency virus (Opravil et al[10]).



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ESC/ERS GUIDELINES

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

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

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ESC entities having participated in the development of this document:

ESC Associations: Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

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Haematological disorders

a. Chronic haemolytic anaemia

The common feature of the haemolytic anaemias is that when there is intravascular haemolysis, there is release of cell-free haemoglobin into the plasma which scavenges the nitric oxide. A loss of nitric oxide, the physiological vasodilator of the pulmonary circulation, may cause vasoconstriction and vascular obstructive pathologic changes.

b. Sickle cell anaemia

Cells containing sickle cell haemoglobin (HbS) ma
additional factor leading to pulmonary hypertension (PH) is that these patients can suffer either functional or surgical asplenia, putting them at risk for
thromboembolism and chronic thromboembolic pulmonary hypertension (CTEPH). There are, however, a few small uncontrolled studies

c. Beta-thalassaemia

PH in patients with thalassaemia is also multifactorial, involving intravascular haemolysis (see above), changes in the coagulation system, and local

d. Hereditary spherocytosis/stomatocytosis

Hereditary stomatocytosis is a rare autosomal red cell membrane disorder and the red cells are subject to intravascular haemolysis. In addition, there is a
high risk of thrombotic complications but, once again, this is often in association with splenectomy which is done to prevent the haemolysis.

e. Myeloproliferative disease

Chronic myeloproliferative disease (CMPD) is associated with PH. There are thought to be 2 main aetiologies.
1. CMPD may have excess risk of venous thrombosis.
2. CMPD may have pre-capillary proliferative vasculopathy. It is of interest that dasatinib, a tyrosine kinase inhibitor, which is one of treatments for
chronic myeloid leukaemia, also appears to cause partially reversible PH.45, 46

f. Splenectomy

Splenectomy causes an increased risk of CTEPH and also even idiopathic pulmonary arterial hypertension.

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Systemic disorders associated with pulmonary hypertension

These disorders include sarcoidosis, histiocytosis, and lymphangiomyomatosis.

a. Sarcoidosis

PH occurs in 5–15%.47

mediastinitis, pulmonary vasculitis, portopulmonary hypertension, and pulmonary veno-occlusive disease.48

b. Langerhans cell histiocytosis (LCH)

PH associated with parenchymal lung disease itself related to smoking.

c. Lymphangioleiomyomatosis (LAM)

PH associated with parenchymal lung disease occurs in approximately 7% of unselected patients with LAM.

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Metabolic disorders

a. Thyroid disease

PH associated with hypo- or hyper-thyroidism.50

b. Glycogen storage diseases

Pathogenesis of PH unknown but may include pulmonary veno-occlusive disease. Enzyme replacement therapy seems to have little effect, unlike Gaucher's disease (see below).

c. Gaucher's disease

Approximately 30% of untreated patients with Gaucher's disease develop PH which is caused by a combination of factors including plugging of the vasculature by the abnormal macrophages, abnormal pulmonary vascular cell proliferation, and asplenia (see above). Treatment with enzyme replacement therapy (ERT), which is now the dominant therapy for Gaucher's disease, may improve the PH However, ERT initiation can also unmask underlying PH.

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Gracias por su atención

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