Ugh. The Echo Report says PH! Now What?

The Diagnostic Evaluation of Pulmonary Hypertension

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Conflicts of Interest

None

Primary Reference

ESC European Society of Cardiology

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ESC/ERS GUIDELINES

2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS).

Outline

- A clinical vignette
- What is pulmonary hypertension? Are there different types?
- Who gets it? How common is it?
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48 yo woman presents with slowly worsening shortness of breath on exertion over the last 2 years and swollen ankles for the last 3 months. Has gained 20 lbs. Belly feels bigger.

No cough. No fever. No wheezing.

Previously healthy. No chronic illness. Nonsmoker. No alcohol in past 6 months. Previously light drinker.

Mother died at age 58. Not sure why. Legs were swollen and also had chronic breathing problems. Died in her sleep. Father and siblings alive and well.

Has seen several physicians. CXR and echo and PFTs have been normal. Was told she should lose weight and exercise more.

HR 105 (regular). BP 98/72. Sat 91% on RA. JVP elevated. Lungs clear. HS regular. Normally split S2 with loud P2. Liver faintly pulsatile. 2+ LE edema. Cognition good. Affect worried.

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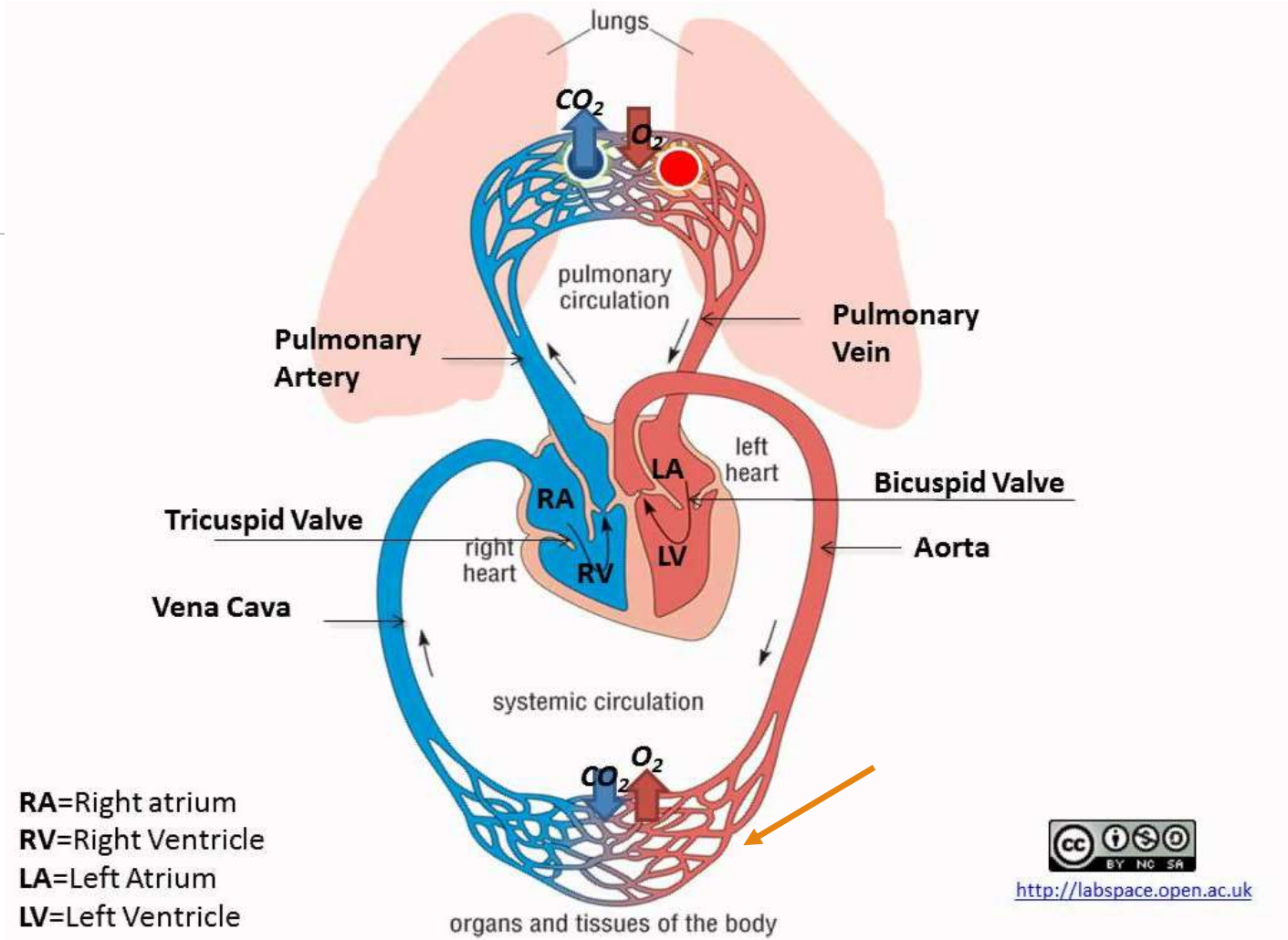
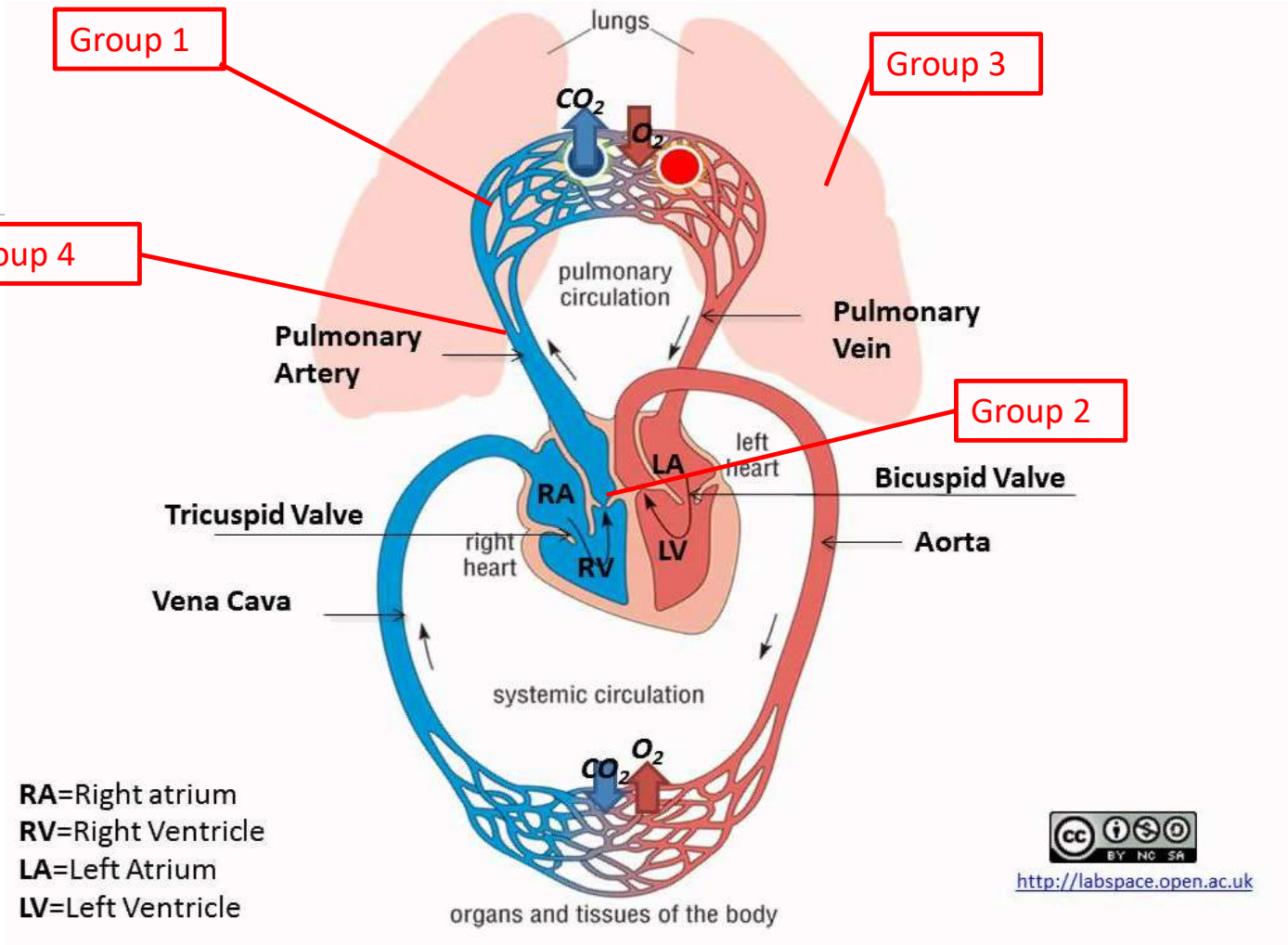


RA=Right atrium
RV=Right Ventricle
LA=Left Atrium
LV=Left Ventricle

PAH= World Symposium Goup 1



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

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CLINICAL CLASSIFICATION

Pulmonary arterial hypertension (PAH)	PH associated with left heart disease	PH associated with lung disease	PH associated with pulmonary artery obstructions	PH with unclear and/or multifactorial mechanisms
				
• Idiopathic/heritable • Associated conditions	• IpcPH • CpcPH	• Non-severe PH • Severe PH	• CTEPH • Other pulmonary obstructions	• Haematological disorders • Systemic disorders

PREVALENCE

Rare	Very common	Common	Rare	Rare
				

PULMONARY HYPERTENSION

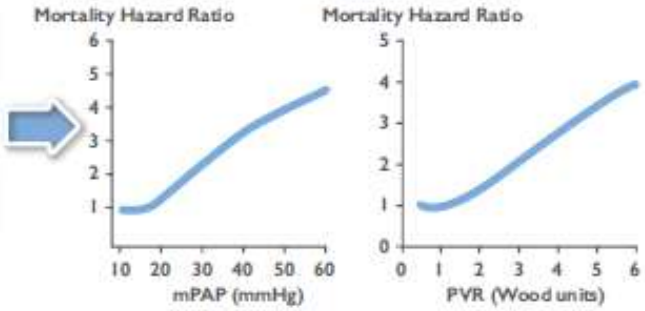
Prevalence

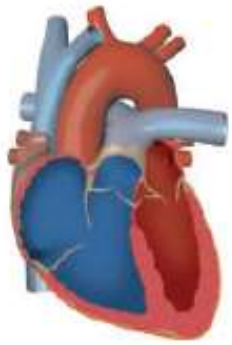
1%

Global population

Pulmonary congestion in post-capillary PH

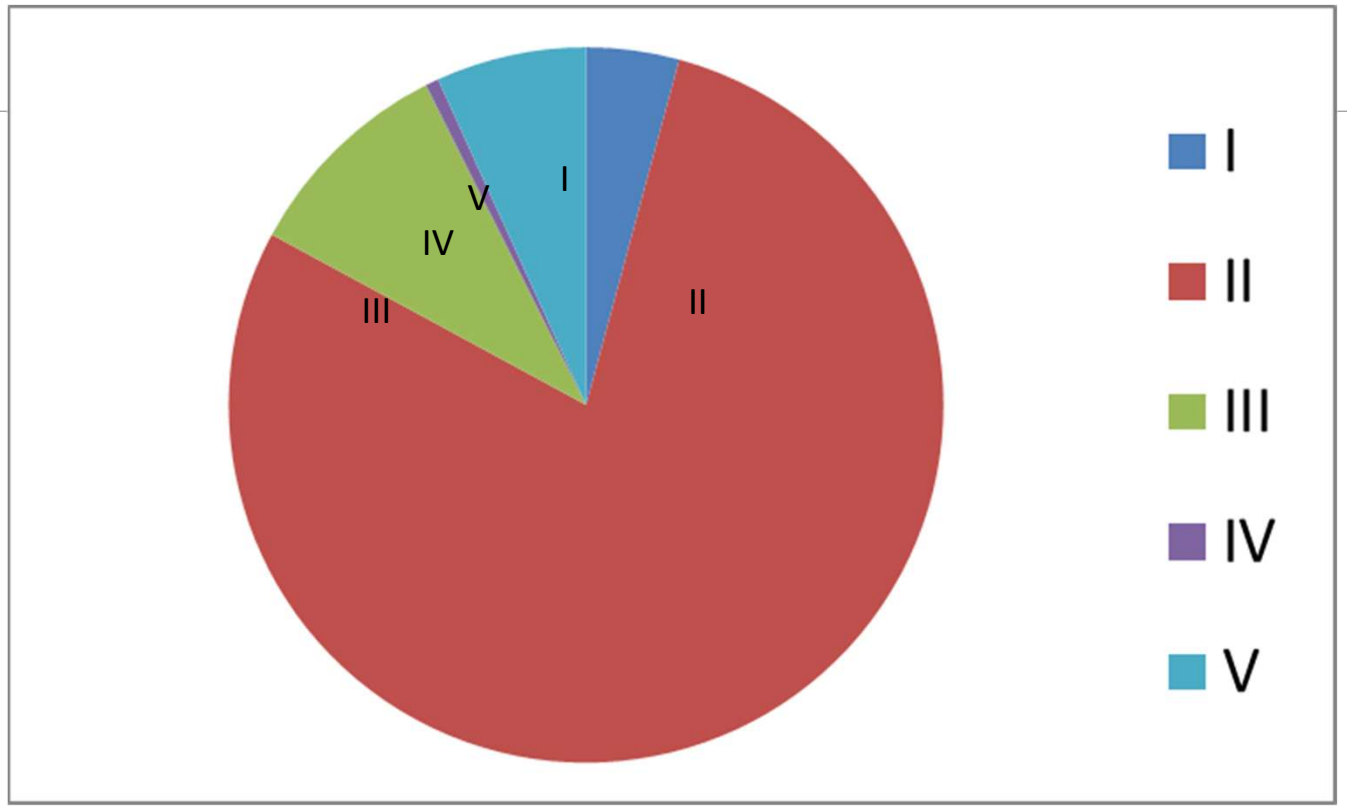
Pulmonary vascular disease / obstruction in pre-capillary PH





Right heart failure

Relative Frequency of PH Groups



N=483 patients with echo PASP >40 mm Hg.
Gabby E. *Am J Respir Crit Care Med*. 2007;175:A713.

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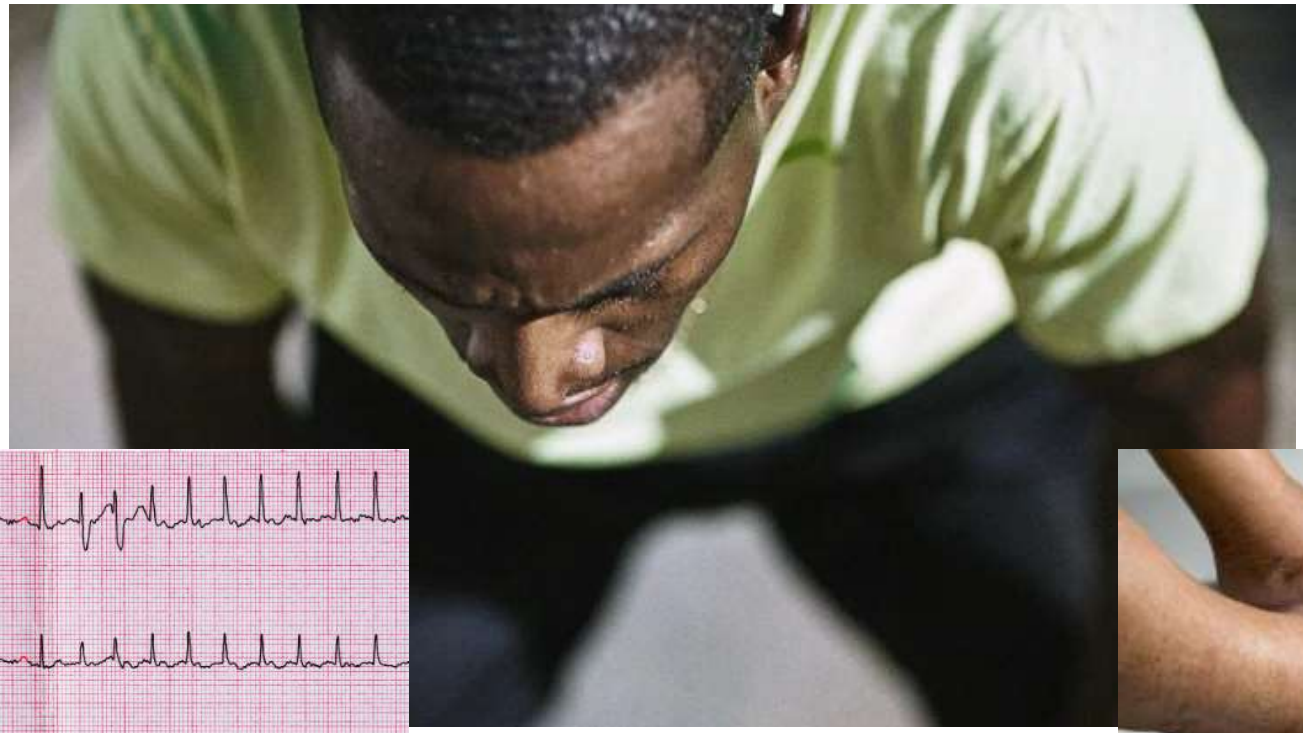
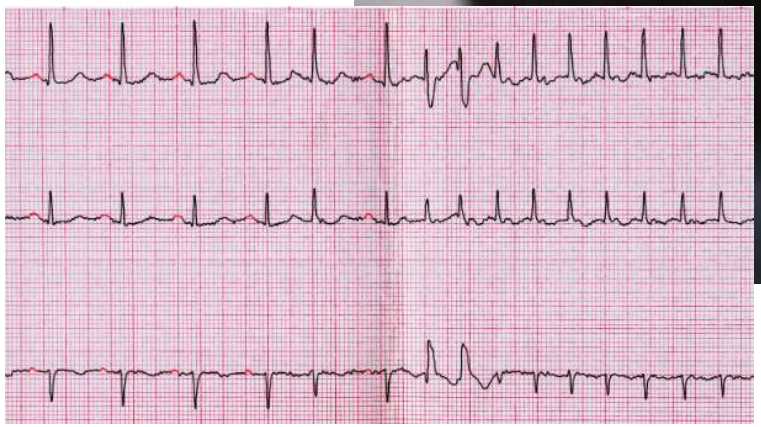
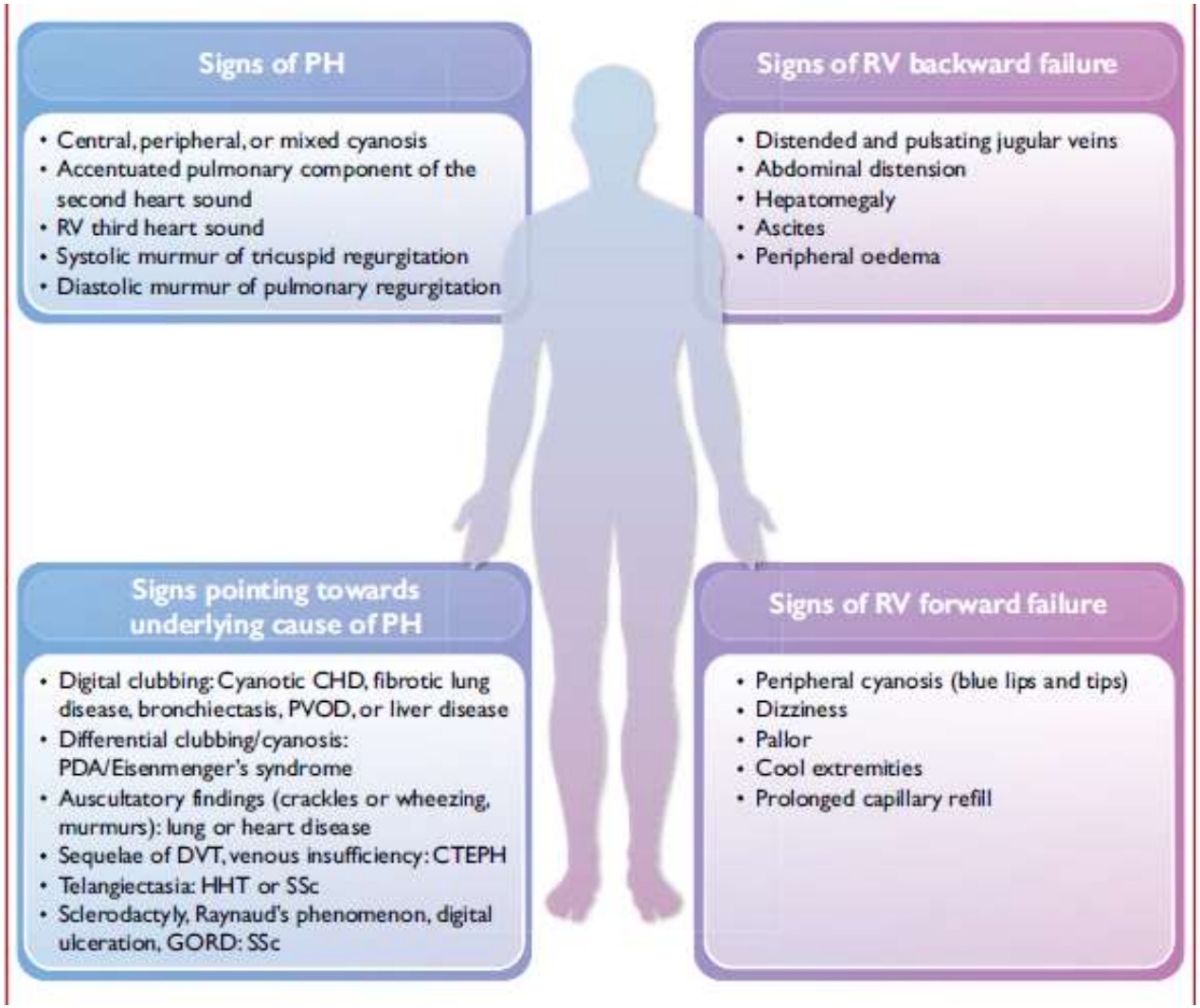
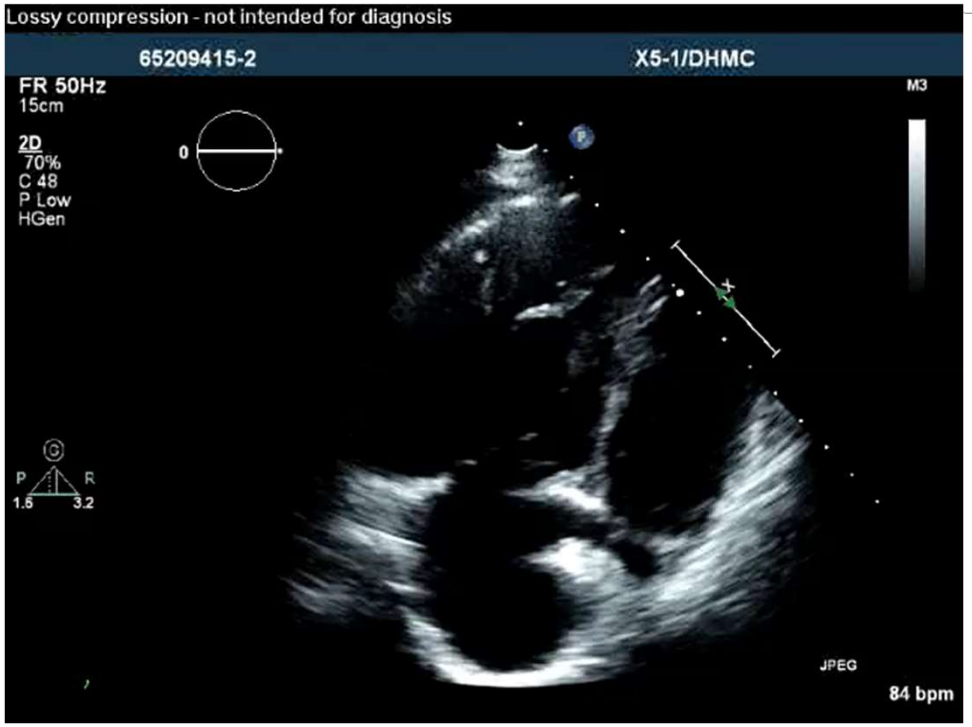





Signs of PH
• Central, peripheral, or mixed cyanosis
• Accentuated pulmonary component of the second heart sound
• RV third heart sound
• Systolic murmur of tricuspid regurgitation
• Diastolic murmur of pulmonary regurgitation
Signs of RV backward failure
• Distended and pulsating jugular veins
• Abdominal distension
• Hepatomegaly
• Ascites
• Peripheral oedema
Signs pointing towards underlying cause of PH
• Digital clubbing: Cyanotic CHD, fibrotic lung disease, bronchiectasis, PVOD, or liver disease
• Differential clubbing/cyanosis: PDA/Eisenmenger's syndrome
• Auscultatory findings (crackles or wheezing, murmurs): lung or heart disease
• Sequelae of DVT, venous insufficiency: CTEPH
• Telangiectasia: HHT or SSc
• Sclerodactyly, Raynaud's phenomenon, digital ulceration, GORD: SSc
Signs of RV forward failure
• Peripheral cyanosis (blue lips and tips)
• Dizziness
• Pallor
• Cool extremities
• Prolonged capillary refill


Lossy compression - not intended for diagnosis
65209415-2
X5-1/DHMC
FR 50Hz
15cm
2D
70%
C 48
P Low
HGen
M3
0
P
R
1.5
3.2
JPEG
84 bpm

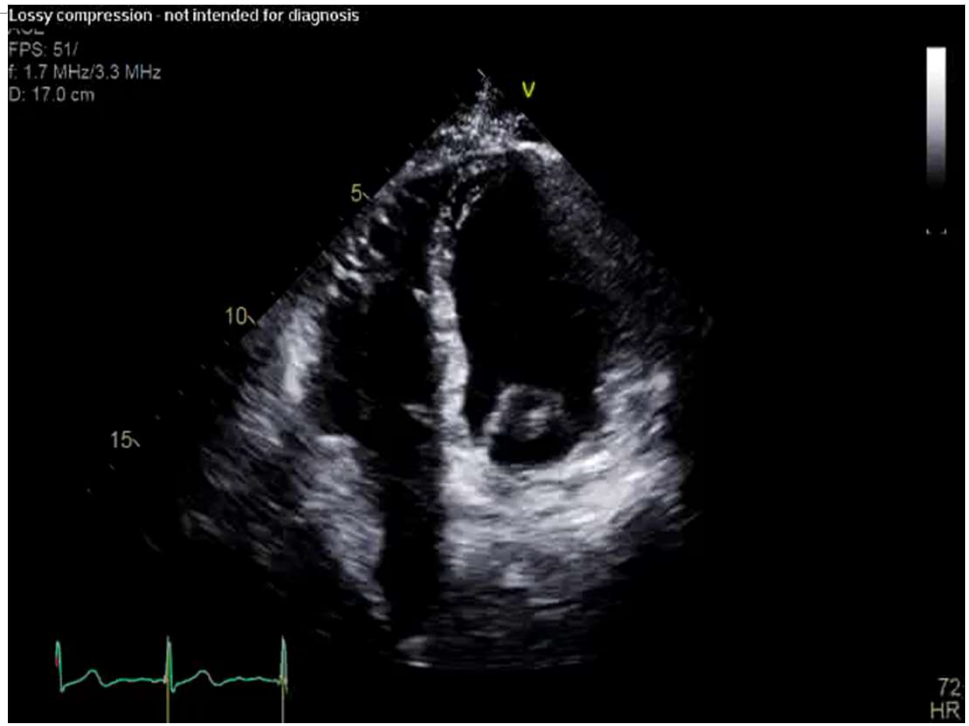

Lossy compression - not intended for diagnosis
FPS: 51/
f: 1.7 MHz/3.3 MHz
D: 17.0 cm
V
5
10
15
72
HR

Table 10 Additional echocardiographic signs suggestive of pulmonary hypertension

A: The ventricles	B: Pulmonary artery	C: Inferior vena cava and RA
RV/LV basal diameter/ area ratio >1.0	RVOT AT <105 ms and/or mid-systolic notching	IVC diameter >21 mm with decreased inspiratory collapse (<50% with a sniff or <20% with quiet inspiration)
Flattening of the interventricular septum (LVEI >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/s	RA area (end-systole) >18 cm^2
TAPSE/sPAP ratio <0.55 mm/mmHg	PA diameter >AR diameter PA diameter >25 mm	

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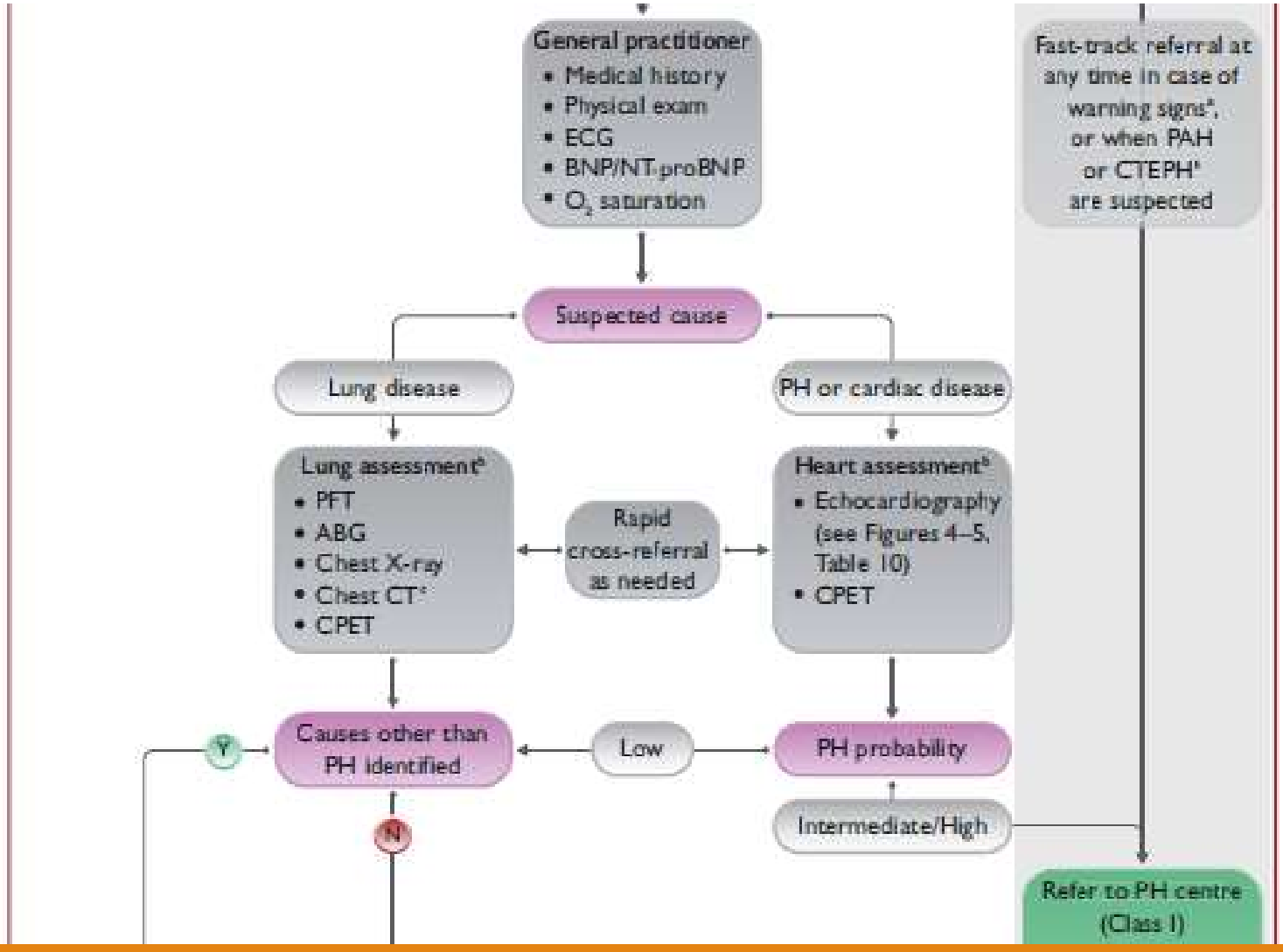
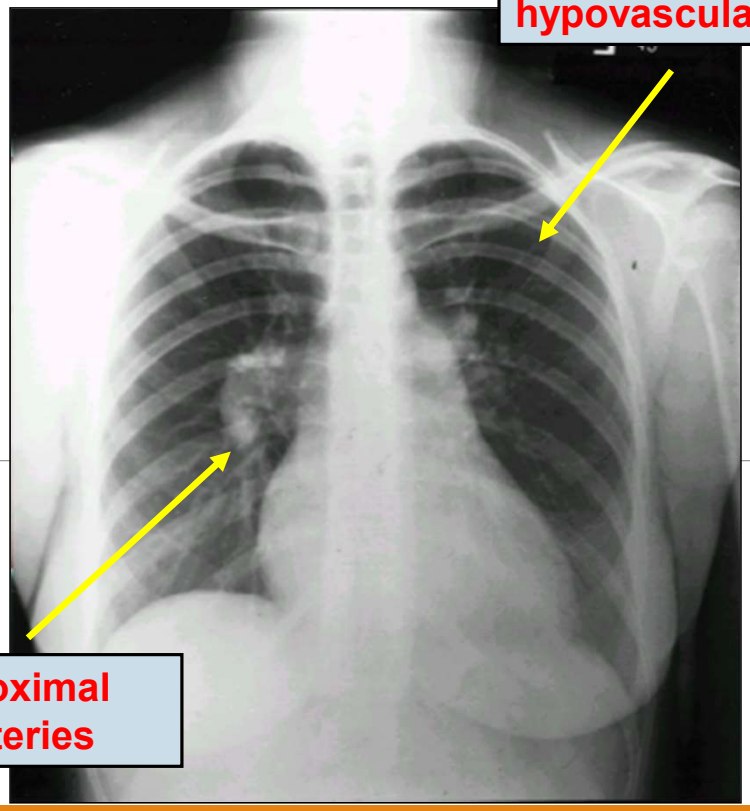
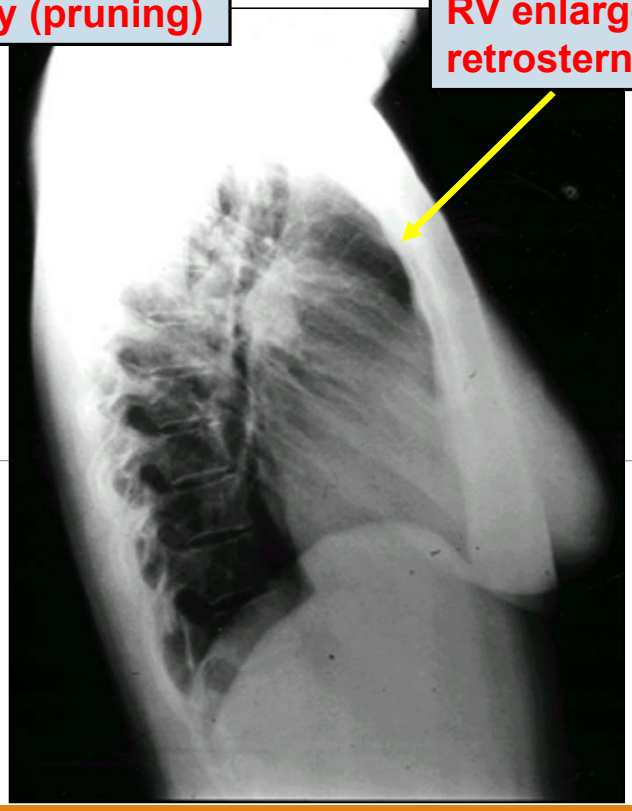

General practitioner
• Medical history
• Physical exam
• ECG
• BNP/NT-proBNP
• O_2 saturation
Fast-track referral at any time in case of warning signs[a], or when PAH or CTEPH[a] are suspected
Suspected cause
Lung disease
PH or cardiac disease
Lung assessment[b]
• PFT
• ABG
• Chest X-ray
• Chest CT[c]
• CPET
Rapid cross-referral as needed
Heart assessment[b]
• Echocardiography (see Figures 4–5, Table 10)
• CPET
Y
Causes other than PH identified
Low
PH probability
N
Intermediate/High
Refer to PH centre (Class I)

CXR



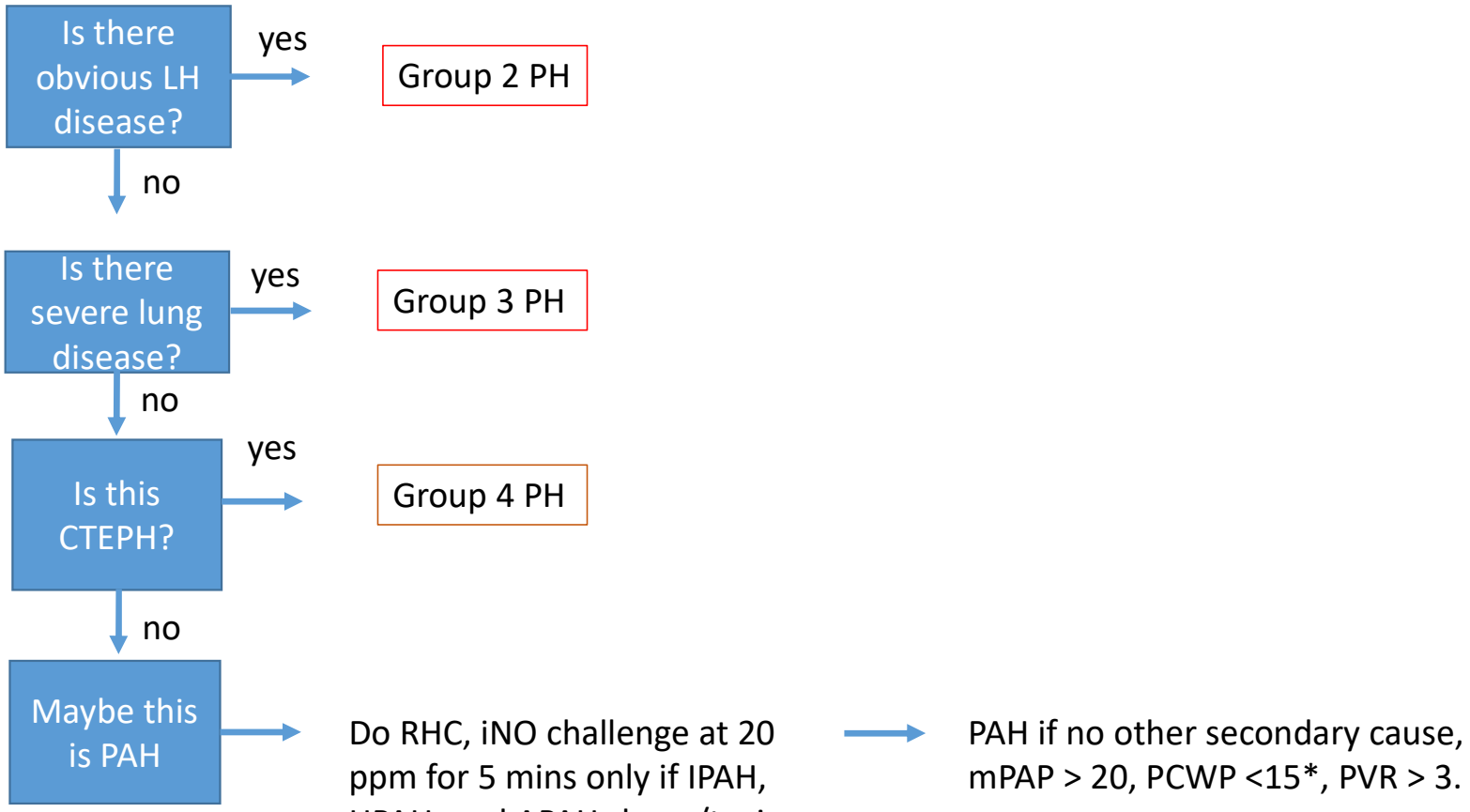


Peripheral hypovascularity (pruning)

RV enlargement into retrosternal clear space

Prominent proximal pulmonary arteries

Diagnostic Approach



Imaging		
Ventilation/perfusion or perfusion lung scan is recommended in patients with unexplained PH to assess for CTEPH[105]	I	C
CT pulmonary angiography is recommended in the work-up of patients with suspected CTEPH[104]	I	C
Routine biochemistry, haematology, immunology, HIV testing, and thyroid function tests are recommended in all patients with PAH, to identify associated conditions	I	C
Abdominal ultrasound is recommended for the screening of portal hypertension[164]	I	C
Chest CT should be considered in all patients with PH	IIa	C
Digital subtraction angiography should be considered in the work-up of patients with CTEPH	IIa	C
Other diagnostic tests		
Pulmonary function tests with DLCO are recommended in the initial evaluation of patients with PH[78]	I	C
Open or thoracoscopic lung biopsy is not recommended in patients with PAH	III	C

PH Risk Factor Review

Risk Factor	Yes	No	Comments
Family hx of PH like illness			
Anorexic drug use			
Cocaine use			
Prior thromboembolism			
Chronic Lung disease			
Symptoms of sleep apnea			
Connective tissue disease			
Raynaud's phenomenon			
Chronic liver lisease			
Congenital heart disease			
Malignancy			
Splenectomy			
Hemoglobinopathy			
Myeloproliferative disorder			
Hereditary Hemorrhaghic Telectangasia			
Thyroid disease			
Parasitic infection			
Chronic high altitude			

PH Relevant Test	Date	Comments
LFTs, ANA, HIV		
CT of chest		
PFTs		
V/Q scan		
Left heart function by echo		
Evaluation for OSA		
Right heart cath		

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Referral to PH Center

- Severe PH or RV dysfunction, regardless of underlying cause
- No lung disease or left heart problems
- FH of pulmonary hypertension
- Uncertainty about diagnosis
- Abnormal V/Q scan
- Not responding as expected

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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%

Used for initial assessment

3 Item Risk Stratifier

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

Used for subsequent visits

Follow Up of PAH

	At baseline	3–6 months after changes in therapy[a]	Every 3–6 months in stable patients[a]	In case of clinical worsening
Medical assessment (including WHO-FC)				
6MWT				
Blood test (including NT-proBNP)[b,c]				
ECG				
Echocardiography or cMRI				
ABG or pulse oximetry[d]				
Disease-specific HR-QoL				
CPET				
RHC				

Rest Stop

Quick Review

What is the hemodynamic definition of PH?

A. mean PA pressure > 20

B. Mean PA pressure > 100

Quick Review

How common is PH?

It affects what % of the general population?

A. 0.001%

B. 1%

C. 10%

Quick Review

What are the 5 different groups of PH?

I	Pulmonary arterial hypertension (pulmonary vascular disease)
II	PH due to left heart failure (back up of pressure from the left heart)
III	PH due to chronic lung disease
IV	PH due to obstruction of pulmonary arteries (mostly chronic thromboembolism)
V	PH due to miscellaneous causes

Quick Review

After history and exam and basic labs, what test is generally done first?

1. Colonoscopy
2. CT of the head
3. Echocardiography
4. Liver biopsy

Quick Review

What complications of PH are we trying to avoid?

- progressive right heart failure (RV dilating and becoming weak)
- worsening disability
- chronic liver disease due to congestion of the liver
- premature death

A Clinical Vignette

48 yo woman presents with slowly worsening shortness of breath on exertion over the last 2 years and swollen ankles for the last 3 months. Has gained 20 lbs. Belly feels bigger.

No cough. No fever. No wheezing.

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