medical masterclass

Pulmonary Hypertension for Advanced Practice Providers: Risk Stratification and Diagnosis to Enhance Quality of Care and Outcomes

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Learning objectives

- Increase knowledge of pulmonary hypertension (PH) diagnostic and treatment approaches
- Recognize early signs and symptoms of pulmonary hypertension
- Utilize tools to appropriately complete risk stratification in patients with PH
- Demonstrate competency in communication with patients and other referral providers to reach underserved patients for confirmatory and earlier diagnosis

CASE 1

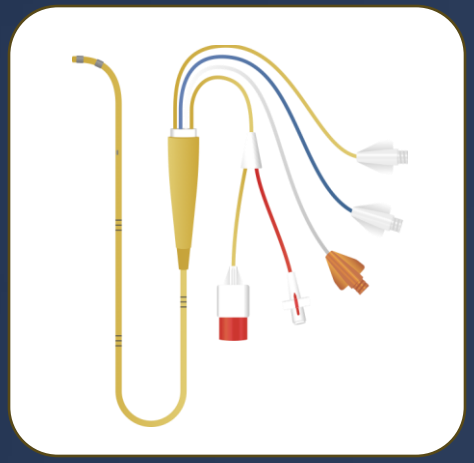
Case Study: 58-Year-Old Woman

Medical History

Physical Exam

Testing

Echo



Right Heart Catheterization

Treatment

Medical History

Medical History

- 58 F with scleroderma (dx 4 years ago), IBS, stage II CKD, no ILD
- 12 months of slowly progressing dyspnea with stairs and housework; not better with weight loss or exercise program
- No chest pain, palpitations, or syncope; some mild leg swelling
- Rheumatologist screened with DETECT algorithm and referred to PH clinic
- No allergies
- Meds: NTG ointment for fingers

Family History

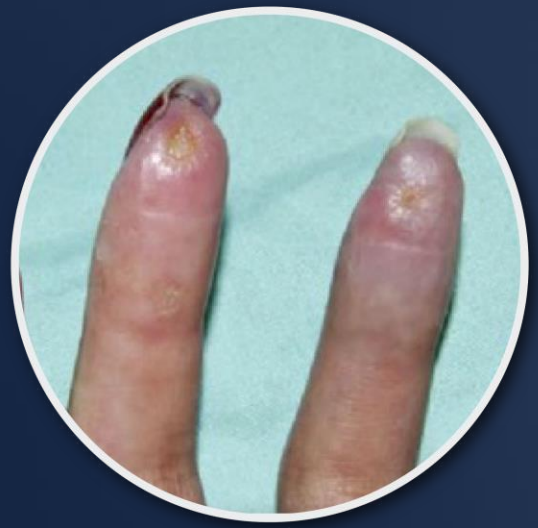
- No family history of PH or other rheumatologic disorders; no history of PE or stimulant use

Social History

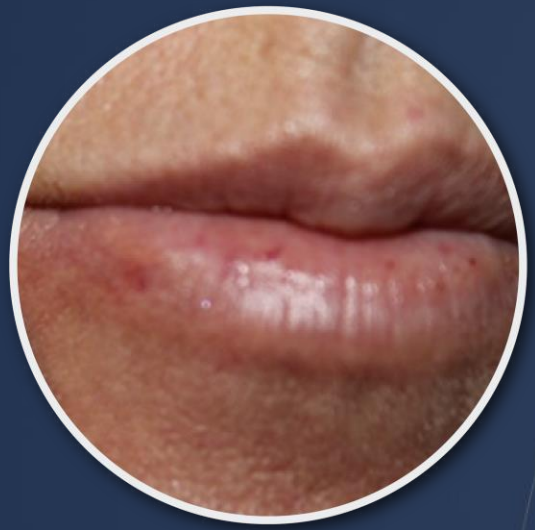
- Works full-time as a fundraiser in a rural community with unreliable internet access

Physical Exam

- Lungs: Clear
- Heart: II/VI HSM at LSB
- Abdomen: benign
- Ext: 1+ LEE bilaterally, digital ulcers, sclerodactyly, and telangiectasia



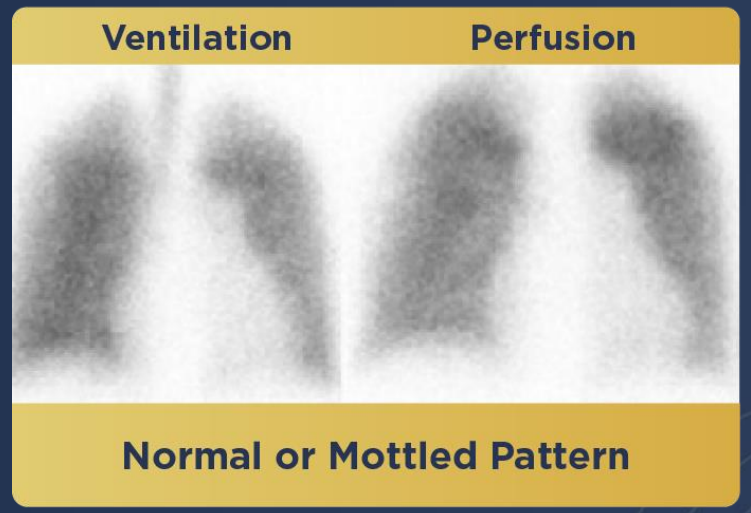
Digital Ulcers



Telangiectasia

Test Results

- 6MWT: 385 m, lowest sat 93%
- FC: III
- Labs:
  - Hgb 9.1, + scl-70, + anti-centromere AB, +ANA, creatinine 0.8
  - NT pro BNP 650
- PFTs
    - FVC: 92%
    - FEV_1/FVC: normal ratio
    - TLC: mildly reduced
    - DLCO: 50% (low)
- VQ scan: no perfusion defects
- HRCT: no ILD
- Overnight oximetry: no desaturations



PAH

2 Steps of the DETECT Algorithm

Step 1
Non-echocardiographic data
(6 variables)

Step 1

Non-ECHO variables

- FVC% predicted/Dlco% predicted
- Current/past telangiectasias
- Serum ACA
- Serum NT-proBNP
- Serum urate
- ECG/right axis deviation

Step 2
Echocardiographic data

Step 2

Total risk points from Step 1
plus ECHO variables
Right atrium area
TR velocity
N = 267

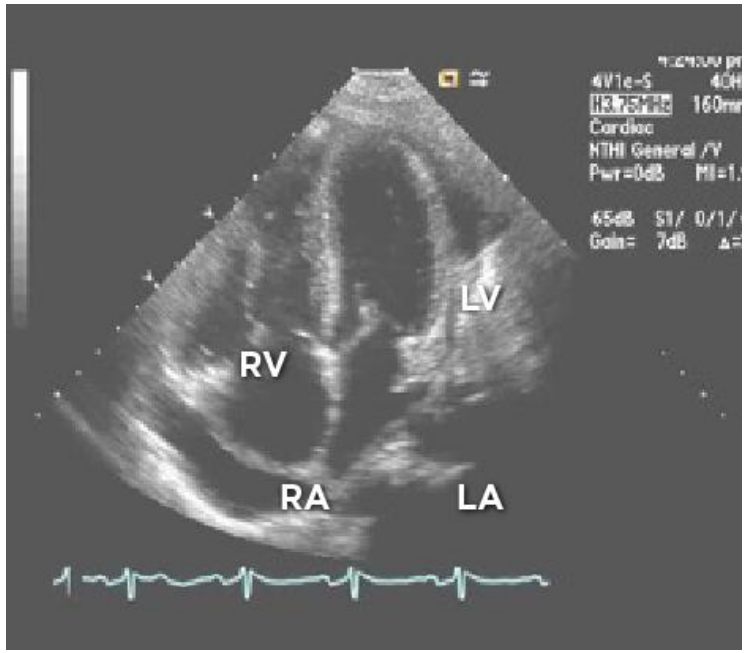
If echo evidence of PH, patients are referred for RHC

This patient's score was 42.

Coghlan JG, et al. *Ann Rheum Dis.* 2014;73(7):1340-1349.

Echocardiogram

- Mild RA and RV enlargement
- Normal LV and LA
- Estimated RVSP = 40 mmHg
- Mildly reduced RV systolic function
- No pericardial effusion

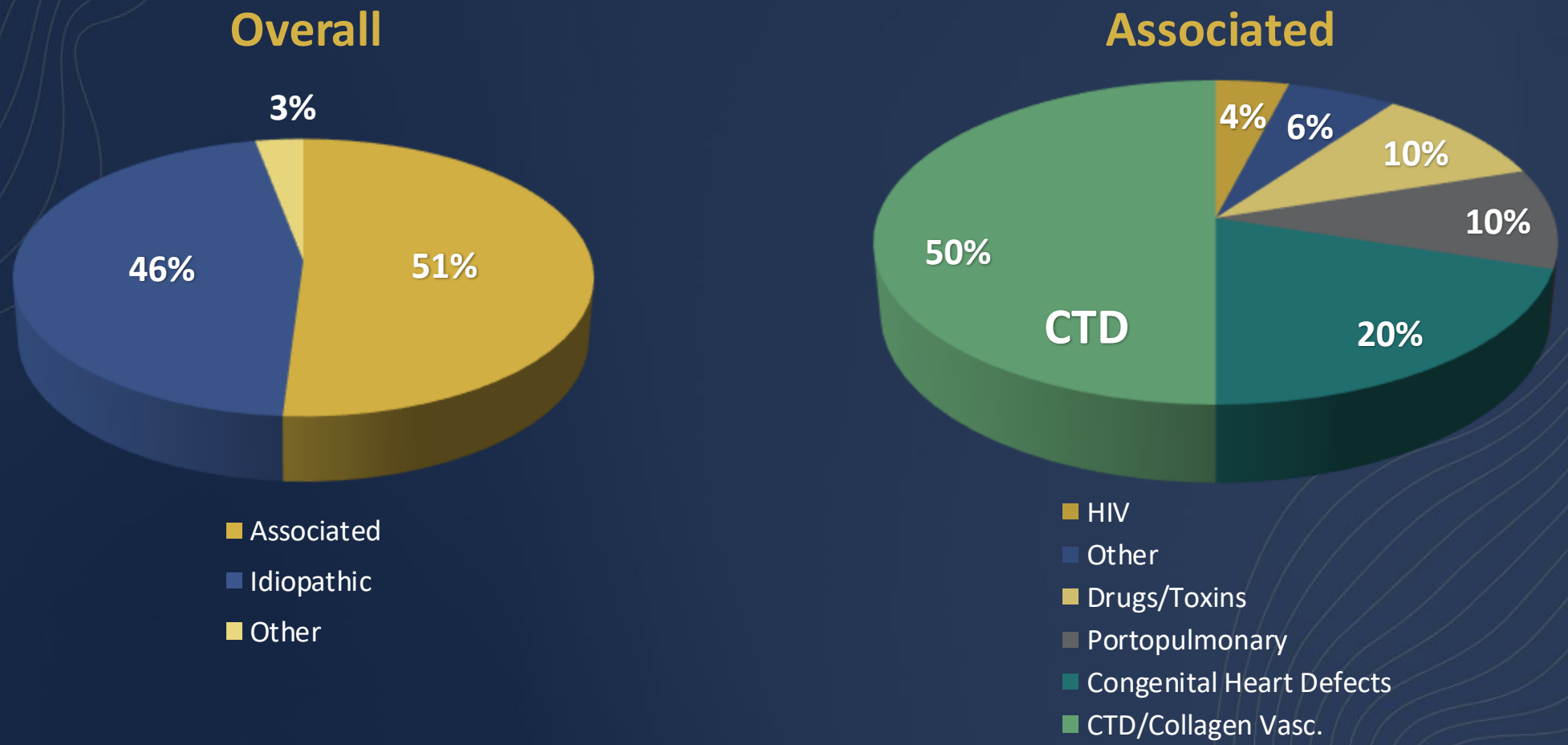


Right Heart Cath

- Hemodynamics:
  - RA = 7 mmHg
  - mPAP = 30 mmHg
  - mPCWP = 8 mmHg
  - CO 4.4/CI 2.7 (TD)
  - SVO_2 64%
  - PVR = 5 Wood units

- Diagnosis: Group 1 PAH associated with scleroderma; CTD-PH

Nearly 25% of PAH Is CTD Related

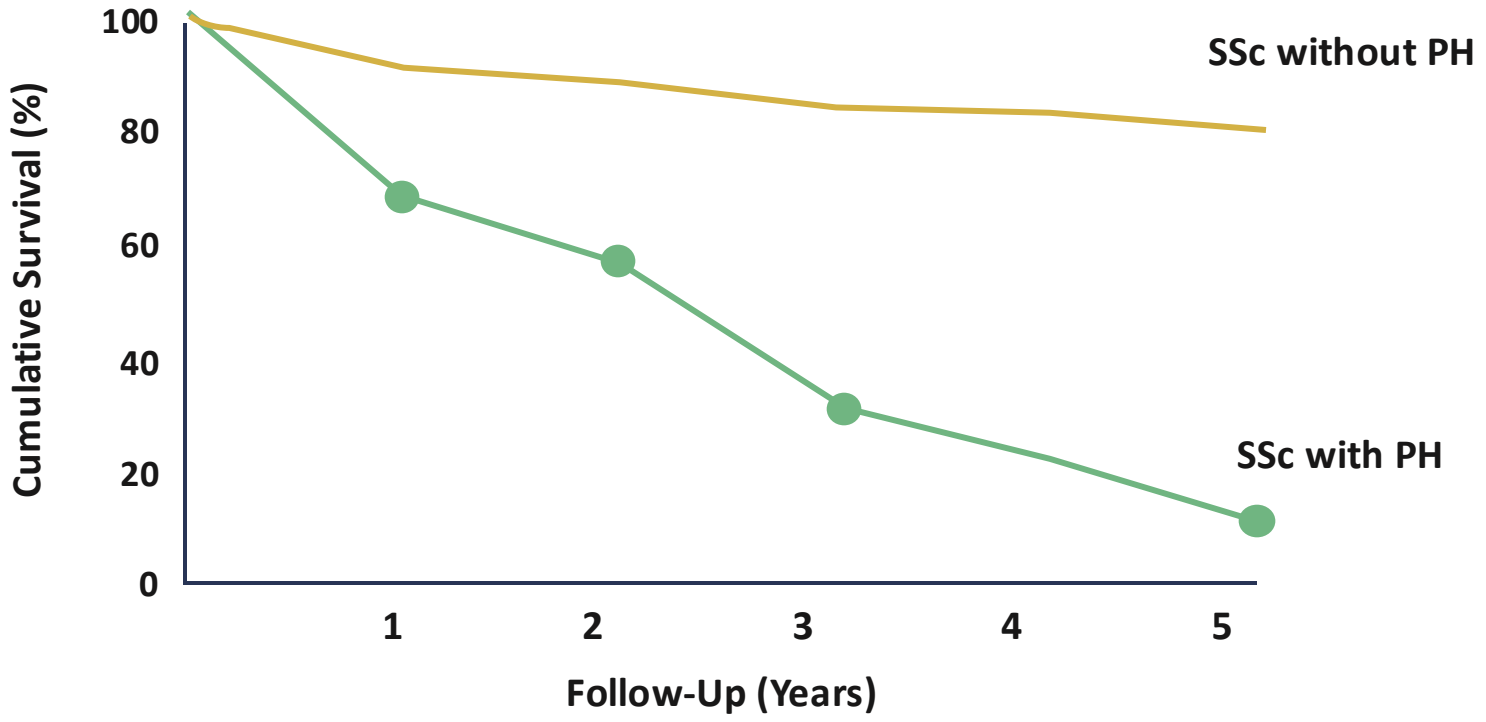


Based on Venice Clinical Classification (2003); 2,967 patients.

Adapted from Badesch DB, et al. *Chest*. 2010;137(2):376-387.

Survival in SSc With and Without PH

Median survival of 1-2 years in pretreatment era



Adapted from Steen V, Medsger TA Jr. *Arthritis Rheum*. 2003;48(2):516-522.

Risk Assessment at Baseline: 3-Strata Model

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

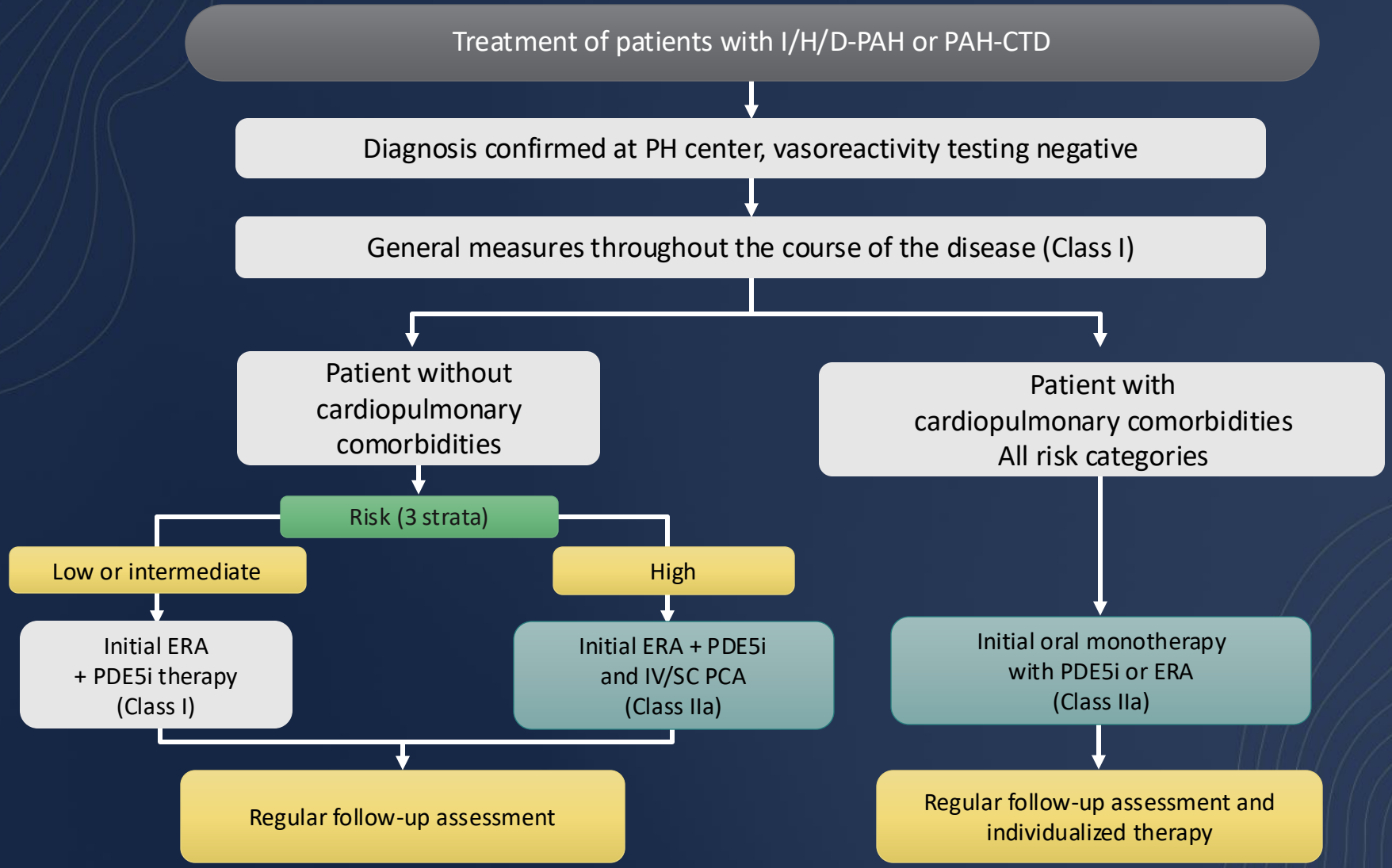
Humbert M, et al. *Eur Heart J*. 2022;43(38):3618-3731.

Risk Assessment at Baseline: 3-Strata Model

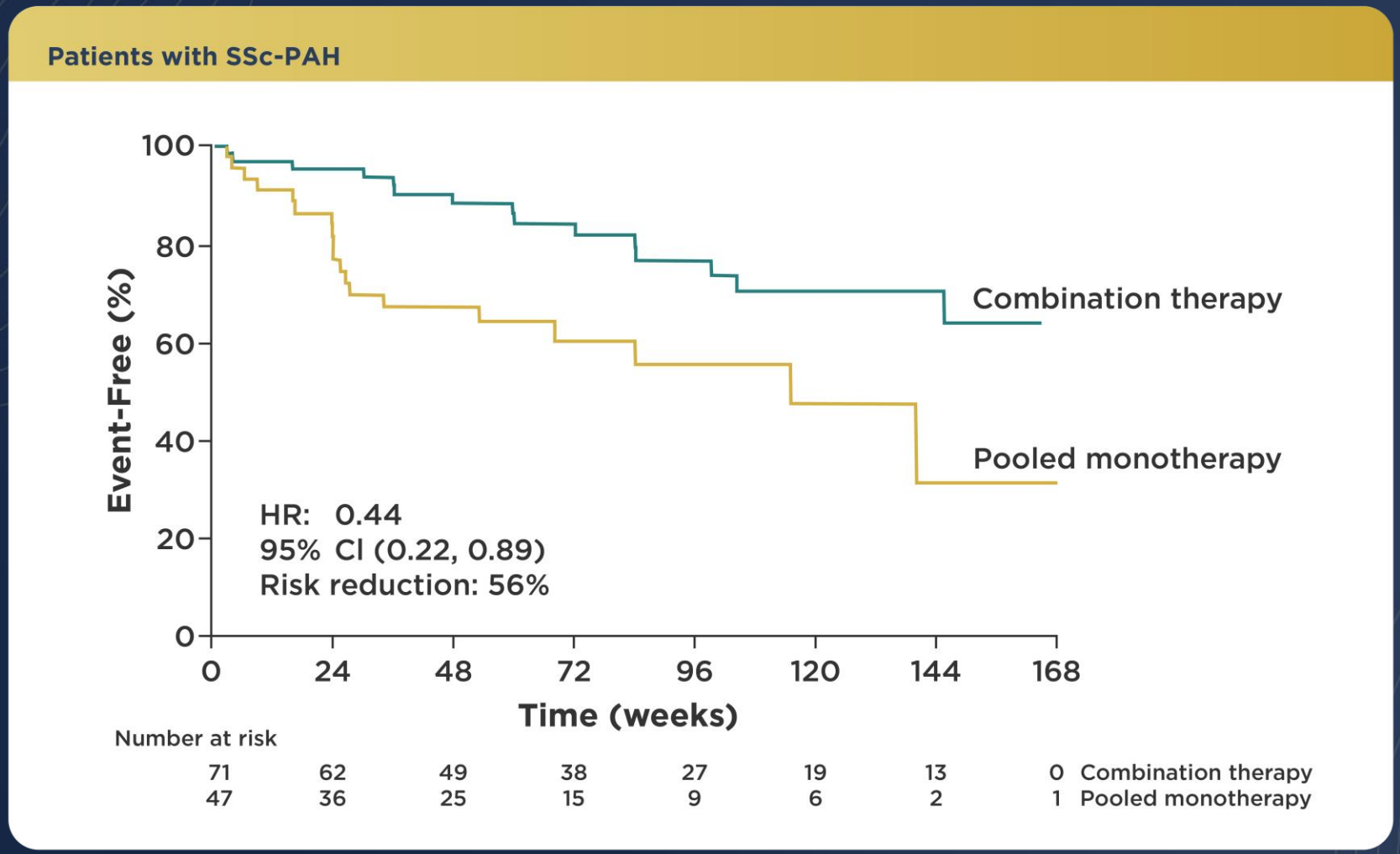
Determinants of prognosis (estimated 1-year mortality)	Low risk (<5%)	Intermediate risk (5–20%)	High risk (>20%)
Clinical observations and modifiable variables			
Echocardiography	RA area <18 cm^2 TAPSE/sPAP >0.32 mm/mmHg No pericardial effusion	RA area 18–26 cm^2 TAPSE/sPAP 0.19–0.32 mm/mmHg Minimal 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%

Humbert M, et al. *Eur Heart J*. 2022;43(38):3618-3731.

Initial Treatment



Ambrisentan/Tadalafil Initial Combo Therapy Reduces Morbidity/Mortality Relative to Monotherapy (SSc-PAH)



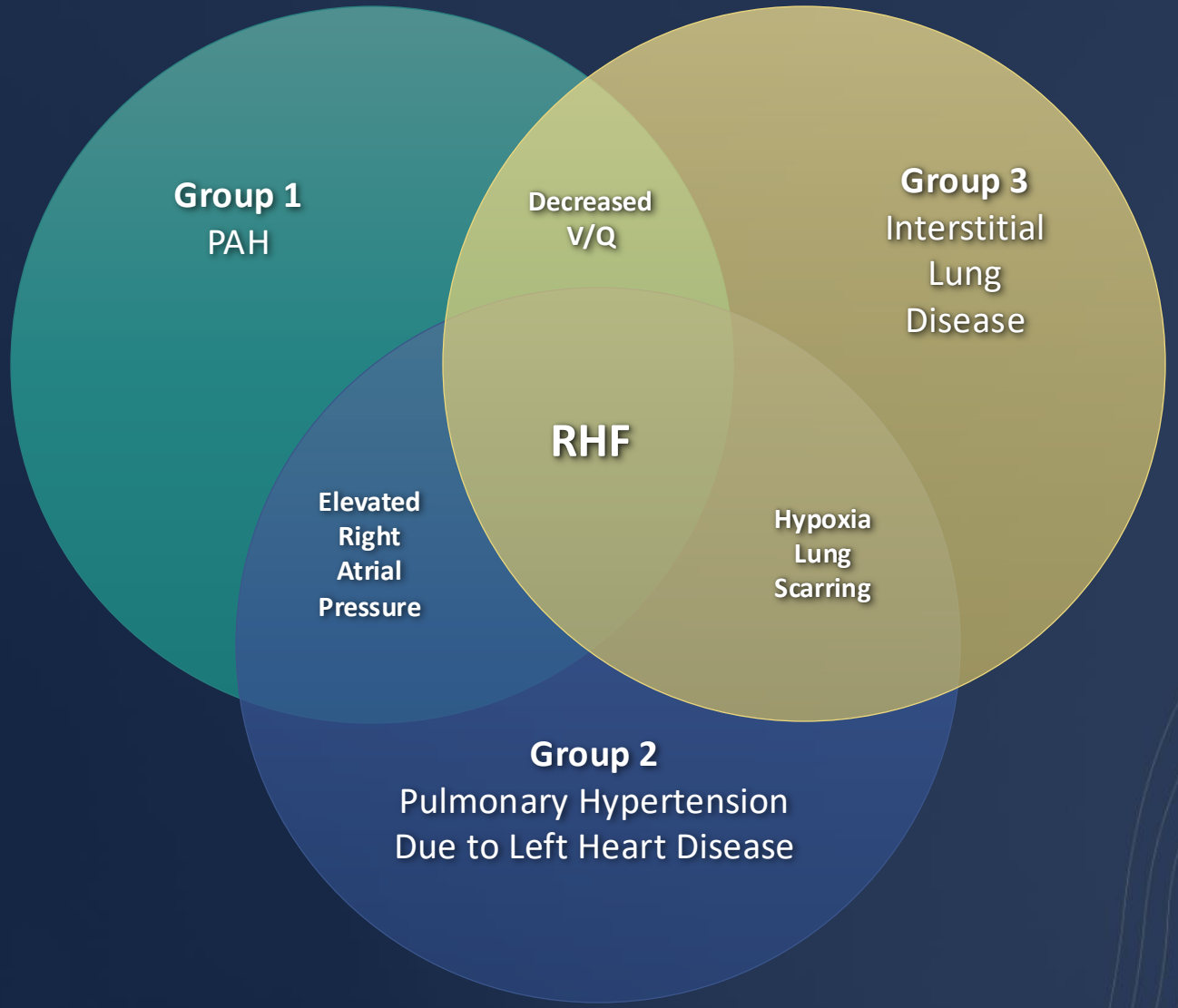
56% RR

Coghlan JG, et al. *Ann Rheum Dis.* 2017;76(7):1219-1227.

Initial Treatment

- General measures:
  - Furosemide 20 mg BID with 20 mEq of potassium chloride
  - BMP in 1-2 weeks
  - Initial PH therapy with shared decision-making:
  - Macitentan and tadalafil
  - Close follow-up in 3 months: consider switch to combination tab of macitentan/tadalafil if tolerating both meds
  - NOTE: Patient unable to do telehealth due to unreliable internet service

Patients With CTD Can Also Have Group 2, 3 PH



Case 1: Summary

- Pulmonary arterial hypertension (PAH) is a common complication of connective tissue diseases (CTD), particularly scleroderma
- When CTD complicates PAH, it significantly worsens survival and is a leading cause of death in these patients
- Early screening of at-risk patients with SSc and MCTD using the DETECT algorithm improves outcomes
- Aggressive treatment, including combination therapy and close follow-up, is recommended

CASE 2

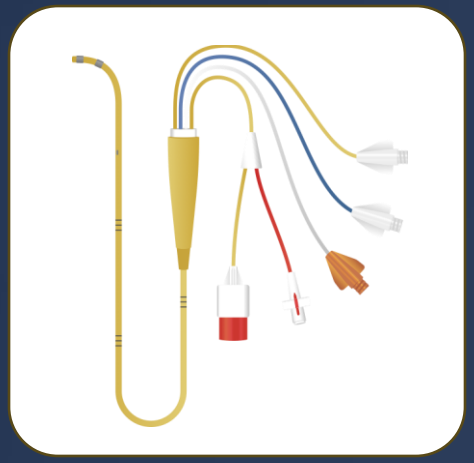
Case Study: 42-Year-Old Woman

Medical History

Physical Exam

Testing

Echo



Right Heart Catheterization

Treatment

Medical History

- History of DVTs (provoked while on OC), not on chronic anticoagulation
- Bipolar disorder
- History of polysubstance abuse
  - Cocaine
  - Methamphetamine
  - Alcohol
  - Tobacco
- History of alcohol-related pancreatitis
- Renal artery aneurysm
- Previous R great saphenous vein ablation

Social & Family History

Social History

- Education: high school
- Works as a waitress
- Low health literacy
- Rural living situation with poor access to Wi-Fi at home
- Single mother of a 2-year-old with a disability
- Smokes 1ppd since age 16
- Remote cocaine use history
- Drinks 4 alcoholic drinks per day, previously heavier consumption
- Uses methamphetamine 2x/day mostly by inhalation, occassionally snorting, no IVDU since age 16 (has been able to quit for up to 5 years at a time). Per her report, uses meth two times daily due to significant fatigue trying to work and care for 2 year old.
- Fragmented primary care

Family History

- Father: DVT
- No family history of pulmonary hypertension or rheumatologic conditions

Patient Presentation/Symptoms WHO Functional Class III

- Dyspnea with walking room to room in her home, climbing stairs, carrying items such as groceries and her 2-year-old son
- Fatigue, daily
- Palpitations with exertion and when lying down at night
- Occasional lightheadedness
- Denies syncope
- Denies PND, orthopnea
- Lower extremity edema for past 3 months

Diagnostic Tests

- ECG:
  - Normal sinus rhythm, right axis deviation, right bundle branch block, right ventricular hypertrophy, and nonspecific ST wave abnormality
- V/Q scan:
  - No evidence of thromboembolic disease
- ANA negative, HIV non-reactive
- 1+ bilateral lower extremity edema
- Transthoracic ECHO:
  - Left ventricular ejection fraction of 60%
  - Moderate tricuspid regurgitation
  - Estimated RVSP of 77 mmHg based on an RA pressure of 3
  - Severe right atrial enlargement
  - Severe right ventricular enlargement
  - Moderately reduced RV systolic function
  - Evidence of right ventricular pressure overload
  - Small pericardial effusion

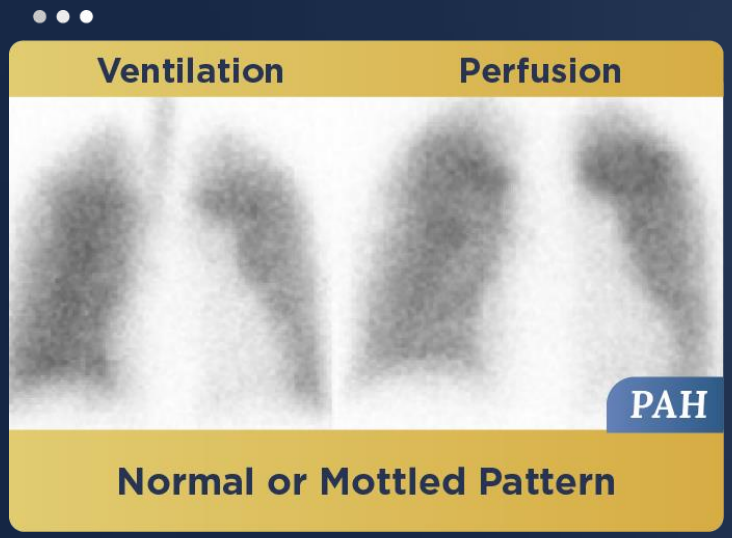
Physical Exam

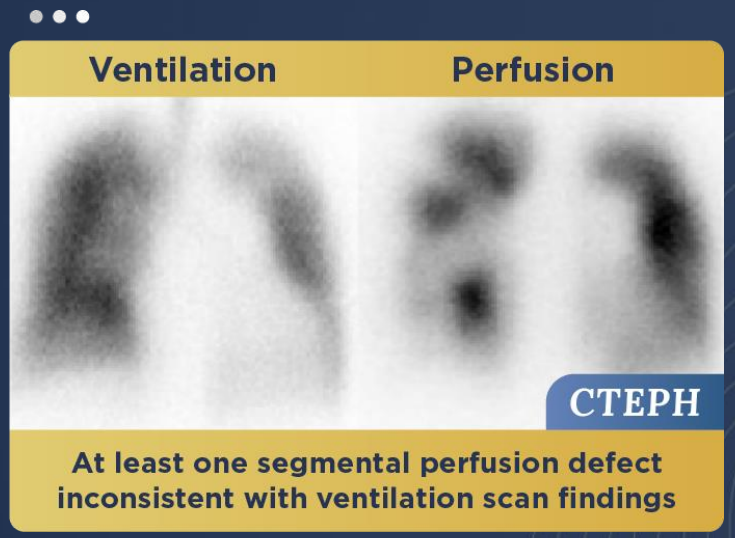
BP 109/62 (BP location: left arm; BP cuff information: adult 23-33 cm)
Pulse 89
Temp 36.6°C (97.8°F)
Resp 16
Ht 1.7 m (5' 6.93")
Wt 79.9 kg (176 lb 2.4 oz)
SpO_2 96%
BMI 27.65 kg/m²

General: Awake, alert, and oriented x 3
HEENT: No carotid bruit, JVP 12 cm
Chest: No crackles, no wheezing
Heart: Palpable RV heave, regular rate and rhythm, normal S1, loud pulmonic component to the second heart sound, + II/VI TR
Extremities: 1+ lower extremity edema bilaterally
Skin: No telangiectasias, no ulcerations, no sclerodactyly

Test Results

Date	FVC (%)	FEV-1 (%)	FEV1/FVC	TLC	DL_{CO}	Comments
03/08/2024	75	69	75		86	SPIROMETRY: No obstructive ventilatory defect; FVC is reduced, which can indicate a restrictive ventilatory defect; full PFTs, including TLC, are required for confirmation if clinically indicated DIFFUSION: Mild gas exchange impairment when corrected for hemoglobin OXIMETRY: Low but adequate oximetry on room air at rest COMPARISON: No prior studies for comparison OTHER: Blunting of inspiratory and expiratory limbs, which can indicate a fixed airway obstruction; clinical correlation recommended; recommend consideration of pulmonary consultation





Test Results

Study/Location	Date	Findings
CXR	3/1/24	No pneumothorax No focal airspace consolidation, frank pulmonary edema, or sizable pleural effusion Enlarged cardiopericardial silhouette with prominent central pulmonary arteries
HRCT		
CT Angiogram	2/28/24 OSH	Enlargement of the central pulmonary arteries and right cardiac chambers; findings consistent with pulmonary arterial hypertension There is eccentric, mural-based low-attenuation material involving the left pulmonary artery and left lower lobar pulmonary arterial branch; suspected to represent thrombus in situ

Test Results

Study/Location	Date	Findings
DECT Angiogram	03/06/2024	Chronic pulmonary embolism; there are peripheral filling defects within the left main pulmonary artery extending into the proximal left upper and left lower lobar pulmonary arteries Small pericardial effusion
V/Q scan	03/04/2024	There are no significant segmental mismatched perfusion defects Overall, the ventilatory study demonstrates greater parenchymal defects with prominent central airways and hilar radioaerosol deposition
DVU LE	03/02/2024	• The RIGHT lower extremity was imaged, assessed by Doppler, and appears patent with no evidence of DVT within the imaged veins • The right great saphenous vein appears ablated • The following LEFT-sided veins revealed evidence of ACUTE DEEP VENOUS THROMBOSIS (DVT): proximal femoral vein (minimal); mid femoral vein (totally occluding); distal femoral vein (totally occluding) • The left small saphenous vein was seen proximal and appears to show evidence of ablation

Test Results (Pulmonary Angiogram)

- Main pulmonary artery is widely patent
- Proximal lobar branches of right and left main pulmonary artery branches are widely patent
- Right A1: Distal arteriopathy with abnormal venous return
- Right A2: Distal arteriopathy with abnormal venous return
- Right A3: Distal arteriopathy with abnormal venous return
- Right A4: Distal arteriopathy with abnormal venous return
- Right A5: Distal arteriopathy with abnormal venous return
- Right A6: Distal arteriopathy with abnormal venous return
- Right A7: Distal arteriopathy with abnormal venous return
- Right A8: Distal arteriopathy with abnormal venous return
- Right A9: Distal arteriopathy with abnormal venous return
- Right A10: Distal arteriopathy with abnormal venous return
- Left A1: Distal arteriopathy with abnormal venous return
- Left A2: Distal arteriopathy with abnormal venous return
- Left A3: Distal arteriopathy with abnormal venous return
- Left A4: Distal arteriopathy with abnormal venous return
- Left A5: Distal arteriopathy with abnormal venous return
- Left A6: Distal arteriopathy with abnormal venous return
- Left A8: Distal arteriopathy with abnormal venous return
- Left A9: Distal arteriopathy with abnormal venous return
- Left A10: Distal arteriopathy with abnormal venous return

ECHO

Date	TR	RVSP	TAPSE - FAC - S'	RV	Atria	LV	Shunt	Comments
3/3/24	Mod	74+3	T 21 S' 11.4	Sev RVE, Mod Dysfx	RAE, Normal LA	EF 60%, RVPO		Small PEF E/A 0.5

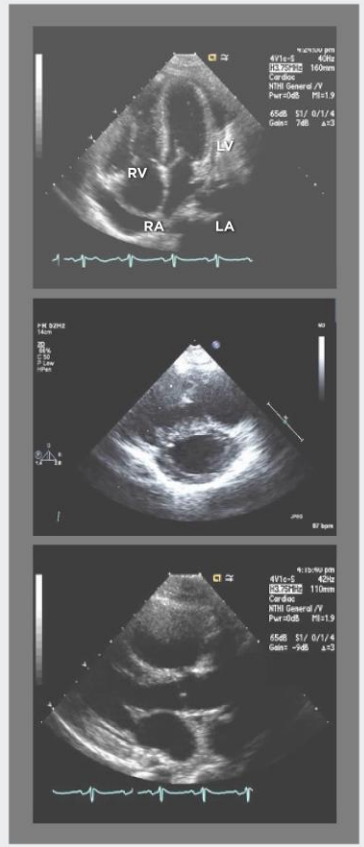
Right Heart Cath

Date	RA	PA (s/d/m)	PCWP/ LVEDP	PVR (Wood units)	Cardiac Output (TD/F)	Cardiac Index (TD/F)	TPG/ DPG	Comments
3/7/24	8	79/40/53	11	13	3.2/3.1	1.7/1.6	42/13	No response to iNO

REVEAL 2.0
Risk Calculator

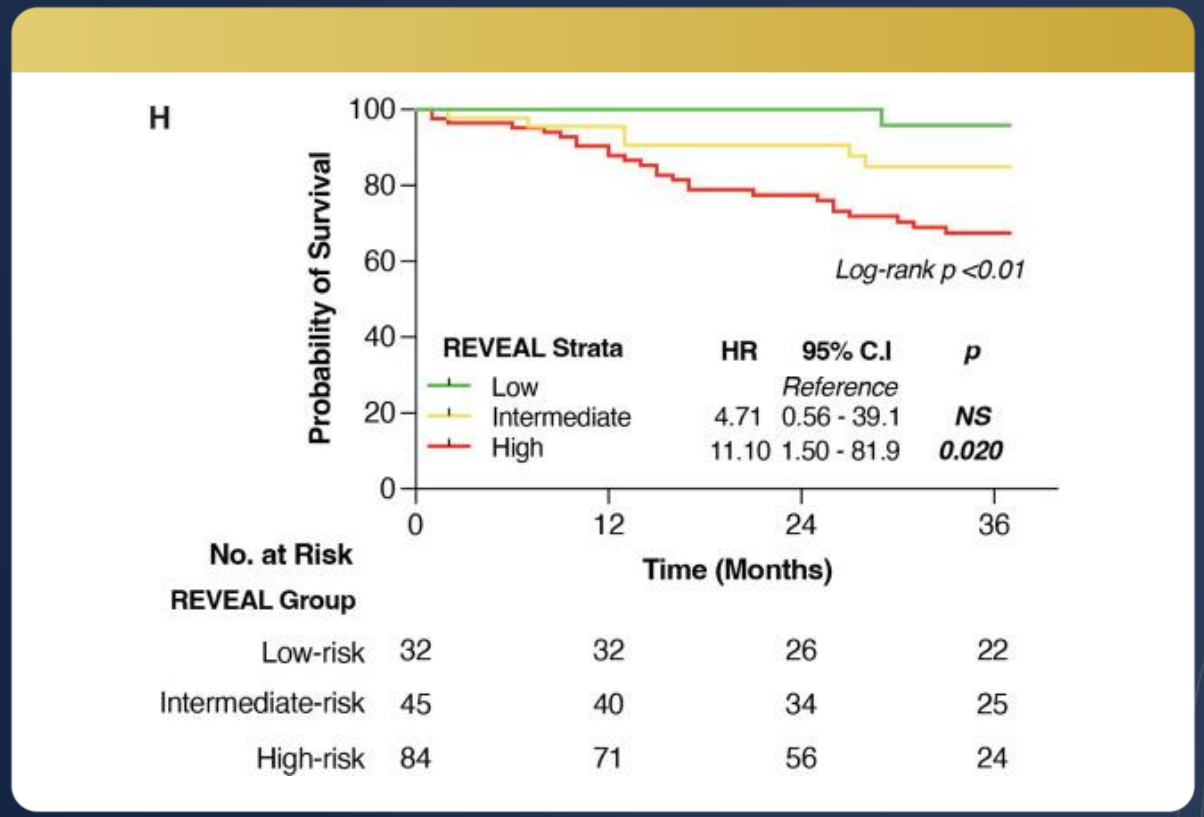
Select all variables that apply. A minimum of 7 variables are required to generate a score. Calculation accuracy increases with more selections.

					Score
WHO Group 1 Subgroup	CTD-PAH 1	Heritable 2	PoPH 3	Other 0	0
Demographics - Male age > 60 years		No 0	Yes 2		0
eGFR<60mL/min/1.73m² or renal insuffiency		No 0	Yes 1		0
NYHA/WHO Functional Class	I -1	II 0	III 1	IV 2	1
Systolic BP (mm Hg)		SBP≥110 0	SBP<110 1		1
Heart Rate (BPM)		HR≤96 0	HR>96 1		0
All-Cause Hospitalizations ≤ 6 mo		No 0	Yes 1		1
6-Minute Walk Test (m)	≥440 -2	320 to 440 -1	<320 to 165 0	<165 1	--
BNP (pg/mL)**	50 -2	50 to <200 0	200 to <800 1	≥800 2	--
or					
NT-proBNP (pg/mL)	<300 -2	300 to <1100 0	≥1100 2		0
Pericardial Effusion on Echocardiogram		No 0	Yes 1		1
% predicted DL_{co} ≤40		No 0	Yes 1		0
mRAP >20 mm Hg Within 1 Year		No 0	Yes 1		0
PVR < 5 Wood units on right heart catheterization		No 0	Yes -1		0
					+6
				Risk Score	10



Risk Scores: REVEAL 2.0

	Low risk	Intermediate risk	High risk
Risk score	0-6	7-8	≥9



Sharp-Dimitri D, et al. *Res Sq*. Preprint published online June 25, 2024. doi:10.21203/rs.3.rs-4474171/v1

Meth-APAH Is a Severe and Progressive Form of PAH With Poor Outcomes

- Urine drug screening is recommended for all patients with idiopathic PAH or a history of substance use disorder
- Methamphetamine use may be a contributor when screening in cases formerly classified as idiopathic PAH
- Routes of use: (Meth)amphetamine can be inhaled (vaporized), smoked, snorted (intranasal), orally ingested, or injected
- Cardiovascular toxicity of (meth)amphetamine:
  - Myocardial ischemia
  - Infarction
  - Cardiomyopathy
- Respiratory:
  - Pulmonary hemorrhage
  - Pulmonary edema
  - Acute lung injury
  - Pneumothorax
  - Pulmonary hypertension

Drugs and toxins associated with pulmonary arterial hypertension (PAH)

Definite association	Possible association
Aminorex	Alkylating agents
Benfluorex	Amphetamines
Carfilzomib	Bevacizumab
Dasatinib	Bortezomib
Dexfenfluramine	Bosutinib
Fenfluramine	Cocaine
Methamphetamines	Diazoxide
Mitomycin C[#]	Direct-acting antiviral agents against hepatitis C virus (sofosbuvir)
Toxic rapeseed oil	Indigo naturalis (Chinese herb Qing-Dai)
	Interferon-α and -β
	Leflunomide
	L-tryptophan
	Phenylpropanolamine
	Ponatinib
	Solvents (trichloroethylene)[#]
	St John's wort

[#]:PA with features of venous (pulmonary veno-occlusive disease)/capillary (pulmonary capillary haemangiomatosis) involvement.

Initial Treatment

- PAH (group 1 PH) associated with drug/toxin use
- WHO Class III
- Social work support for community resources for mental health wellness, application for social security disability, durable power of attorney, etc.
- Support for cessation of substance use (methamphetamine, alcohol, tobacco) and durable contraception
- Pulmonary hypertension patient mentor and support group connection
- Shared decision-making regarding PH medical therapy
  - Start tadalafil 20 mg daily with plan for up-titration to 40 mg daily as outpatient
  - Macitentan 10 mg daily to be started as outpatient
  - Inhaled treprostinil to be started
    - Patient is not a candidate for parenteral prostacyclin therapy due to social factors and patient preferences
- Plan
  - Close patient follow-up with frequent telehealth visits, every 2 weeks after hospital discharge
  - Repeat echocardiogram at 3 months
  - Consider timing of repeat RHC based on clinical response, echo findings, and follow-up risk stratification

Case 2: Summary

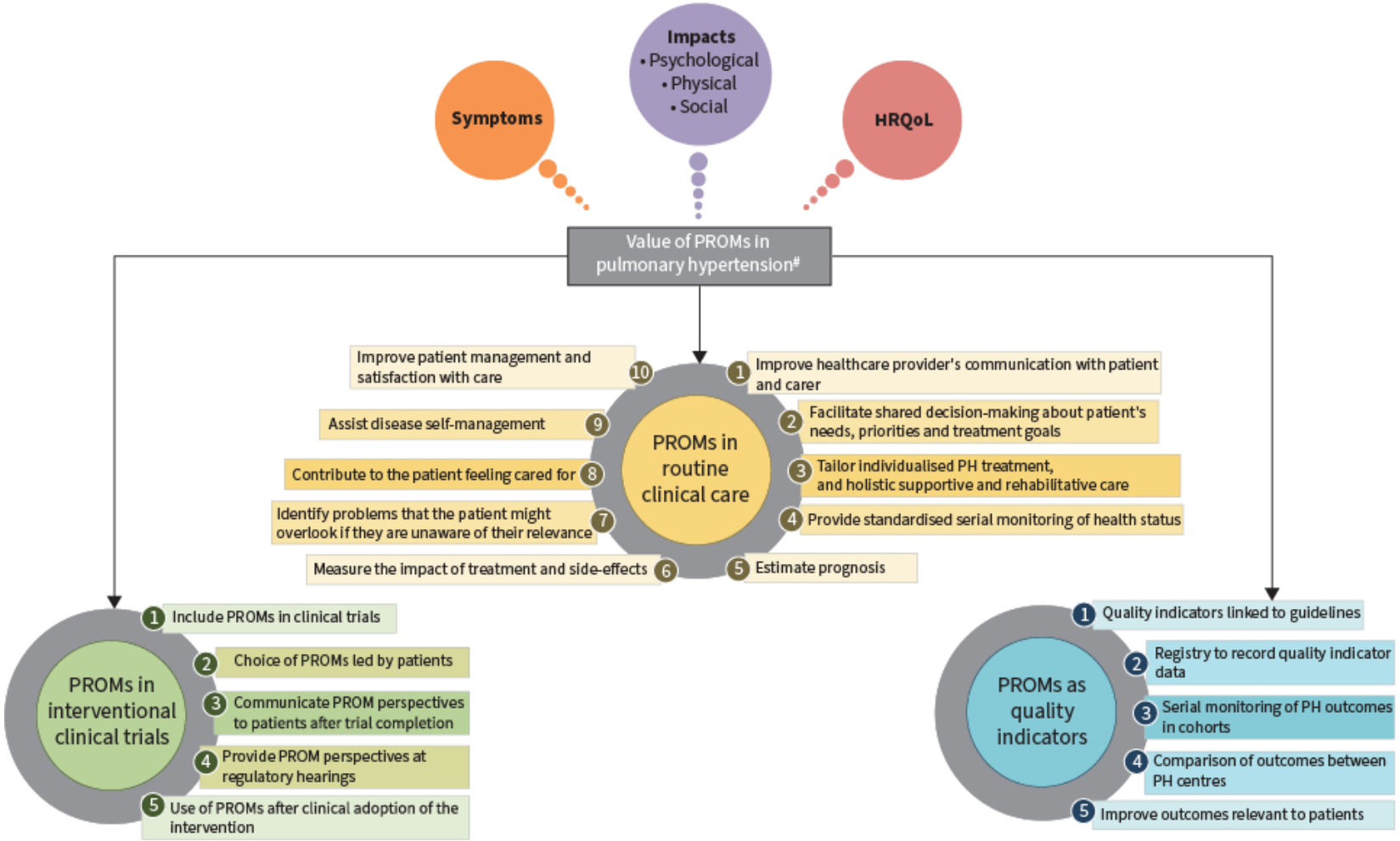
- Meth-APAH is a severe and progressive form of PAH with poor outcomes
- Close follow-up and frequent visits can help build trust and promote adherence to PH medical recommendations
- Recommend comprehensive team approach to care in setting of high-risk social determinants of health
- Aggressive treatment, including combination therapy and repeat risk stratification, is recommended

medical masterclass

A Sampling of Updates From the 7th World Symposium

Patient Perspective

Patient-Related Outcome Measures



Ford HJ, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01129-2024

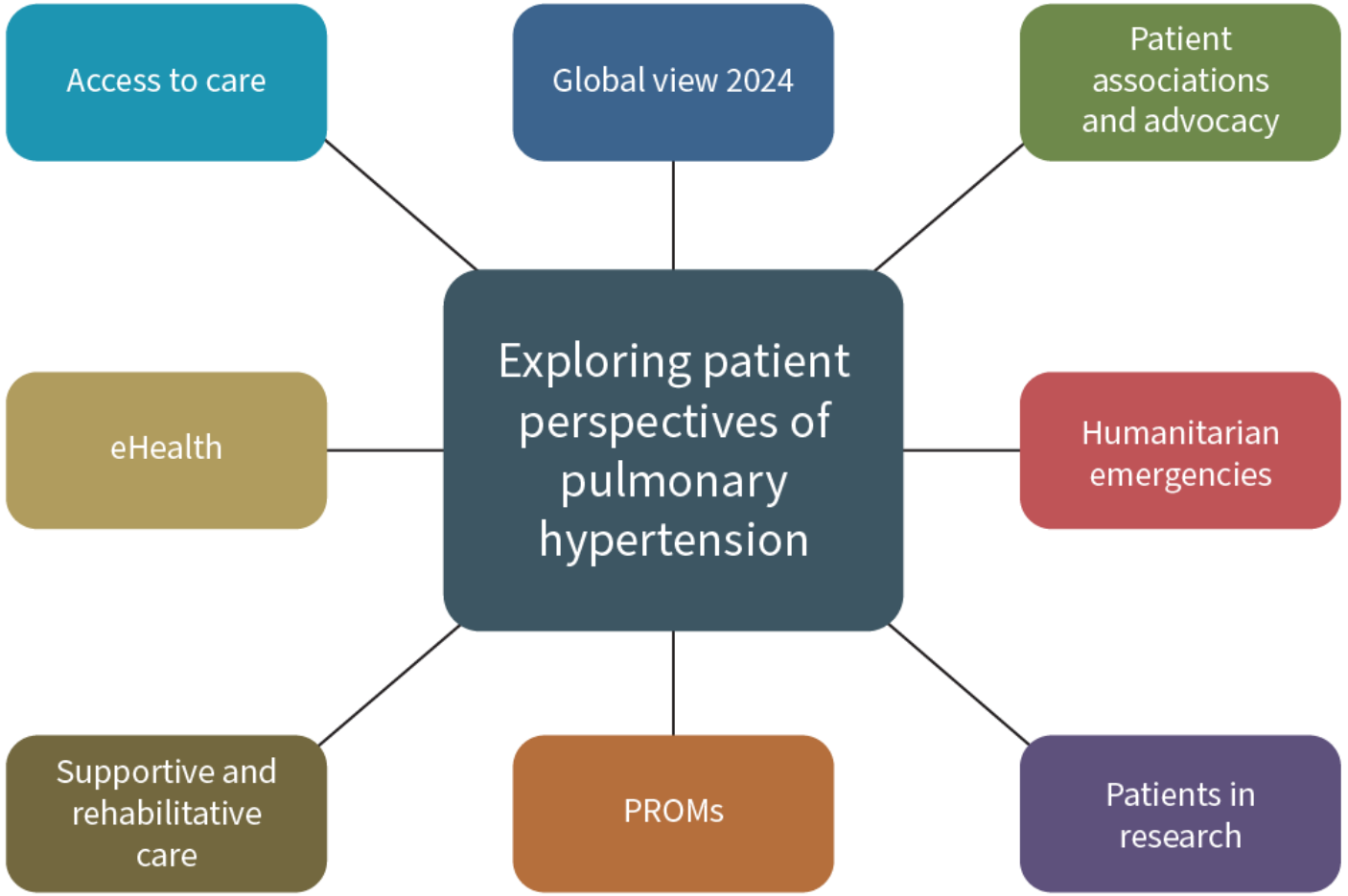


FIGURE 3 Overview of various factors and perspectives affecting and informing the pulmonary hypertension patient's experience of living with the disease. PROMs: patient-reported outcome measures.

Ford HJ, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01129-2024

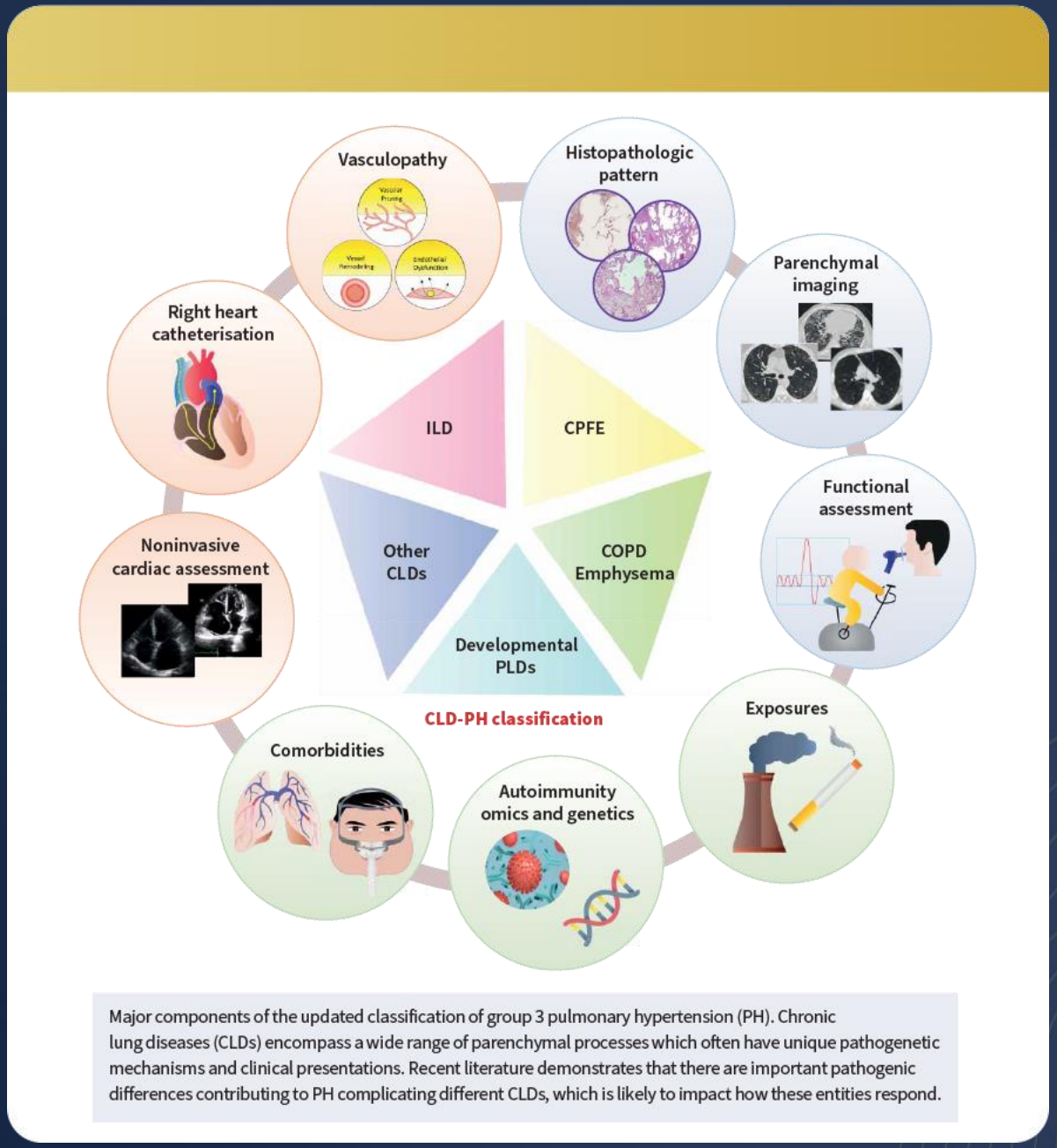
Classification

Updated clinical classification of pulmonary hypertension (PH)
Group 1: PAH
1.1 Idiopathic
1.1.1 Long-term responders to calcium channel blockers
1.2 Heritable[#]
1.3 Associated with drugs and toxins[#]
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
2.1.3 cardiomyopathies with specific etiologies[¶]
2.2 Valvular heart disease:
2.2.1 aortic valve disease
2.2.2 mitral valve disease
2.2.3 mixed valvular disease

Kovacs G, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01324-2024

Group 3 Classification Changes

Instead of “restrictive/ obstructive lung disease,” now using COPD, ILD, CPFE. Also introduced the term “nonparenchymal restrictive disorder.”

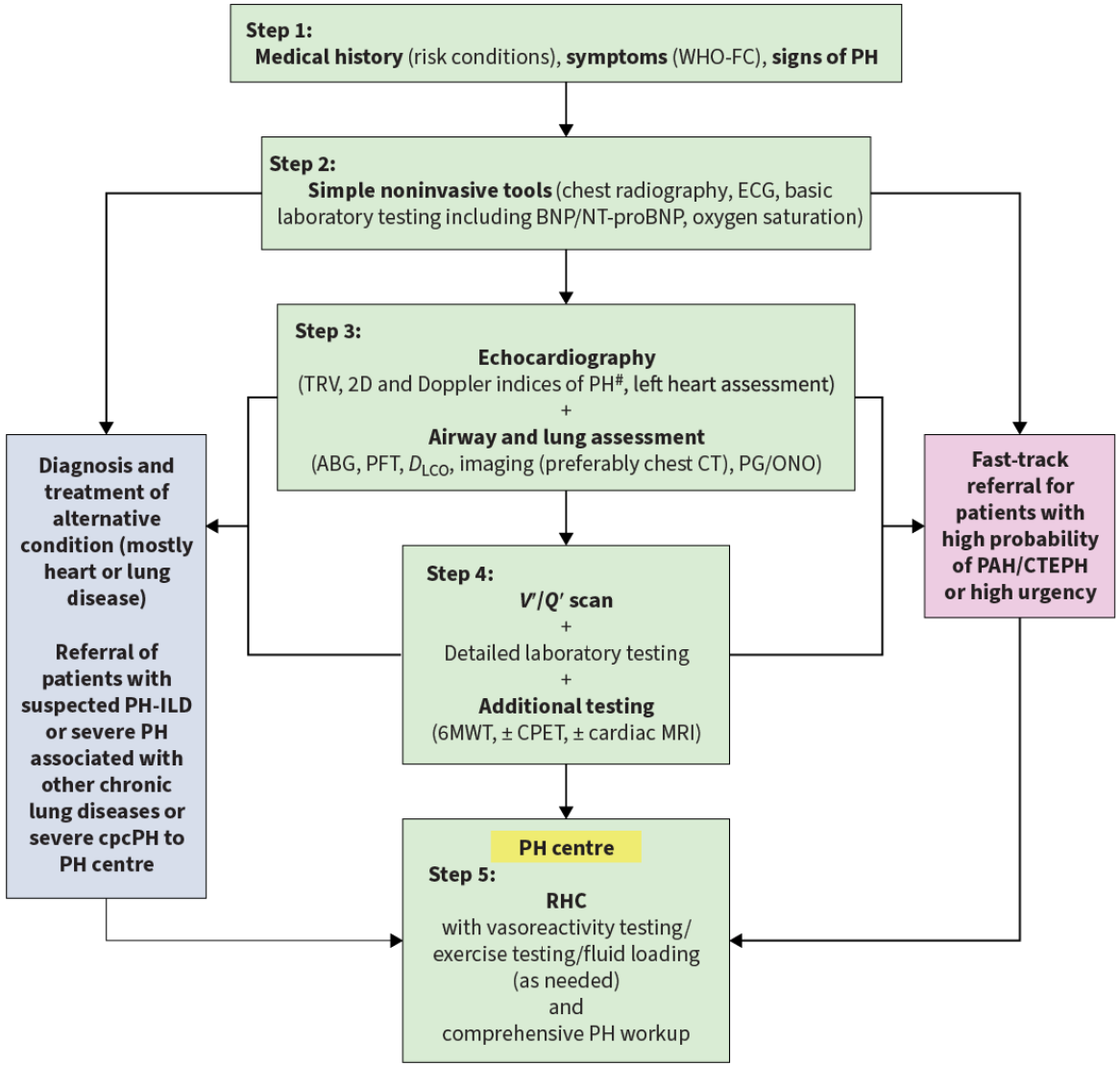


Major components of the updated classification of group 3 pulmonary hypertension (PH). Chronic lung diseases (CLDs) encompass a wide range of parenchymal processes which often have unique pathogenetic mechanisms and clinical presentations. Recent literature demonstrates that there are important pathogenic differences contributing to PH complicating different CLDs, which is likely to impact how these entities respond.

Shlobin OA, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01200-2024

Diagnosis

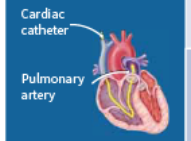
Diagnostic Algorithm



Kovacs G, et al. *Eur Respir J*. Published online August 29, 2024.
doi:10.1183/13993003.01324-2024

Therapy Update

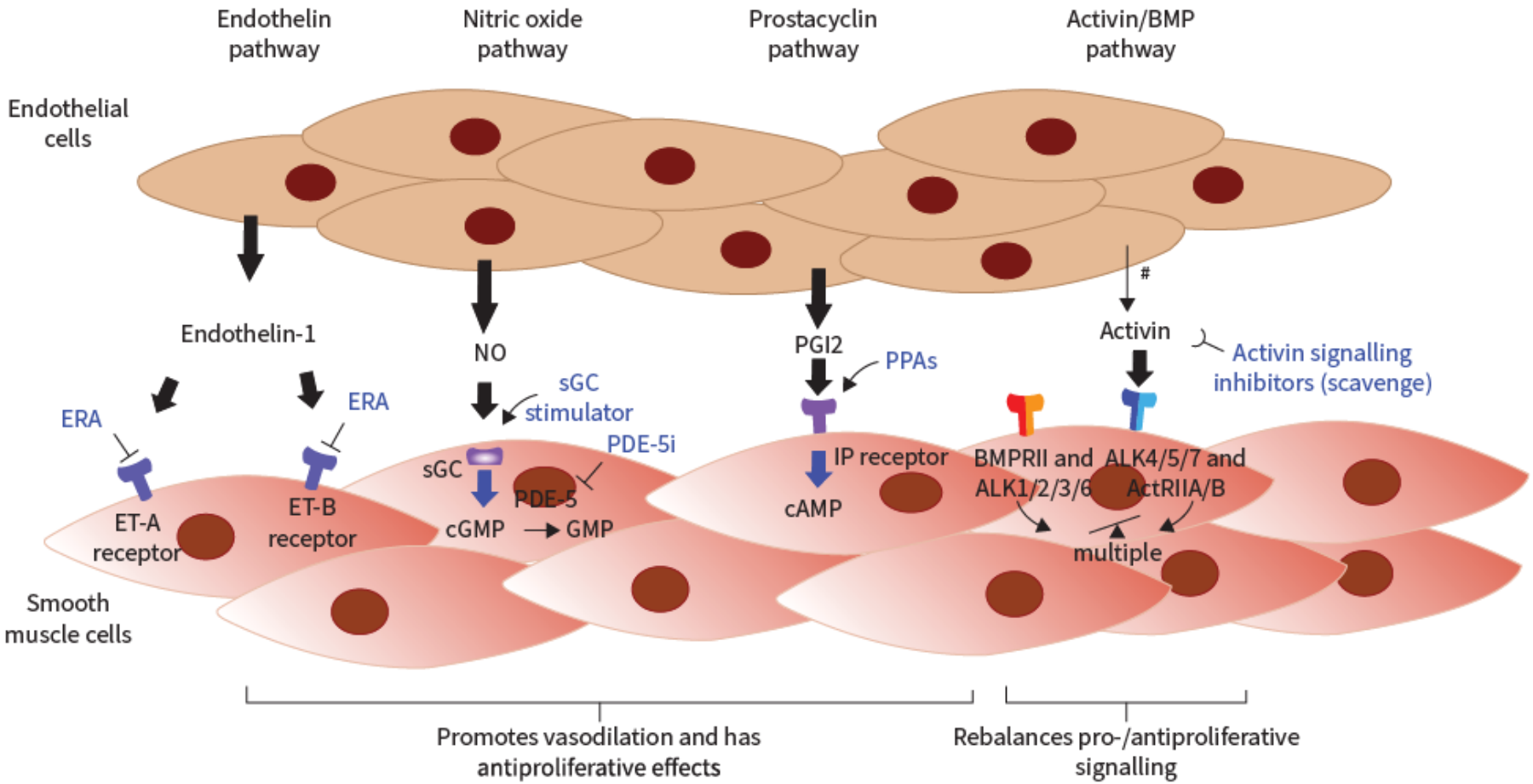
Goals of Therapy

Domain	Treatment goals	Comments	Limitations
Exercise tolerance 	**6MWD >440 m** **WHO-FC I or II**	Not disease-specific, potentially affected by conditions other than PAH	Goals may not be achievable in patients with other conditions limiting exercise capacity
RV function and strain 	**BNP <50 ng·L^{-1}** **NT-proBNP <300 ng·L^{-1}**	Not disease-specific, potentially affected by conditions other than PAH	Goals may not be achievable in patients with interfering conditions
	Need for research prioritisation: RA area <18 cm^2 TR, none or trace TAPSE/sPAP >0.32 mm·mmHg^{-1}	Other imaging parameters from echocardiography and MRI are emerging	TAPSE/sPAP threshold requires further validation
Haemodynamics 	**RAP <8 mmHg** **CI ≥2.5 L·min^{-1}·m^{-2}** **SVI >37 mL·m^{-2}** **S_{vO_2} >65%** **PVR <5 WU**	Uncertain added value in low-risk patients according to ESC/ERS 4 strata model PVR <5 WU treatment goal may not apply to patients with congenital heart disease	Established prognostic value; however, not necessarily independent of noninvasive parameters
	Need for research prioritisation: mPAP <30–35 mmHg PAC ≥2.5 mL·mmHg^{-1}	With emerging therapies and effective combination treatment strategies, comprehensive haemodynamic assessment of treatment response is expected to play a prominent role in the management of patients with PAH	The proposed thresholds may be associated with long-term survival; however, this is not evidence-based and requires further validation

Comprehensive treatment goals inpulmonary arterial hypertension (PAH). RV: right ventricle; 6MWD: 6-min walk distance; WHO-FC: World Health Organization functional class; BNP: brain natriuretic peptide; NT-pro BNP: N-terminal pro-BNP; RA: right atrium; TR: tricuspid regurgitation; TAPSE/sPAP: tricuspidannular plane systolic excursion/systolic pulmonary artery pressure ratio (estimated by echocardiography); RAP: right atrial pressure; CI: cardiac index; SVI: stroke volume index; S_{vO2}:mixed venous oxygensaturation; PVR: pulmonary vascular resistance; WU: Wood Units; mPAP: mean pulmonary artery pressure; PAC: pulmonary arterial compliance; ESC: European Society of Cardiology; ERS: European Respiratory Society.

Dardi F, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01323-2024

4 Pathways



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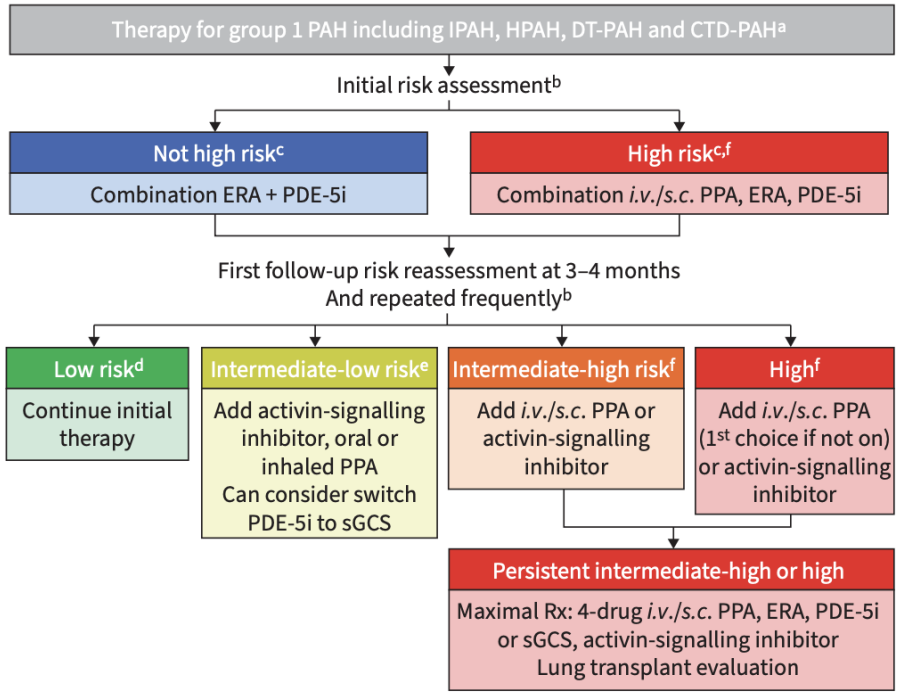
Treatment Algorithm



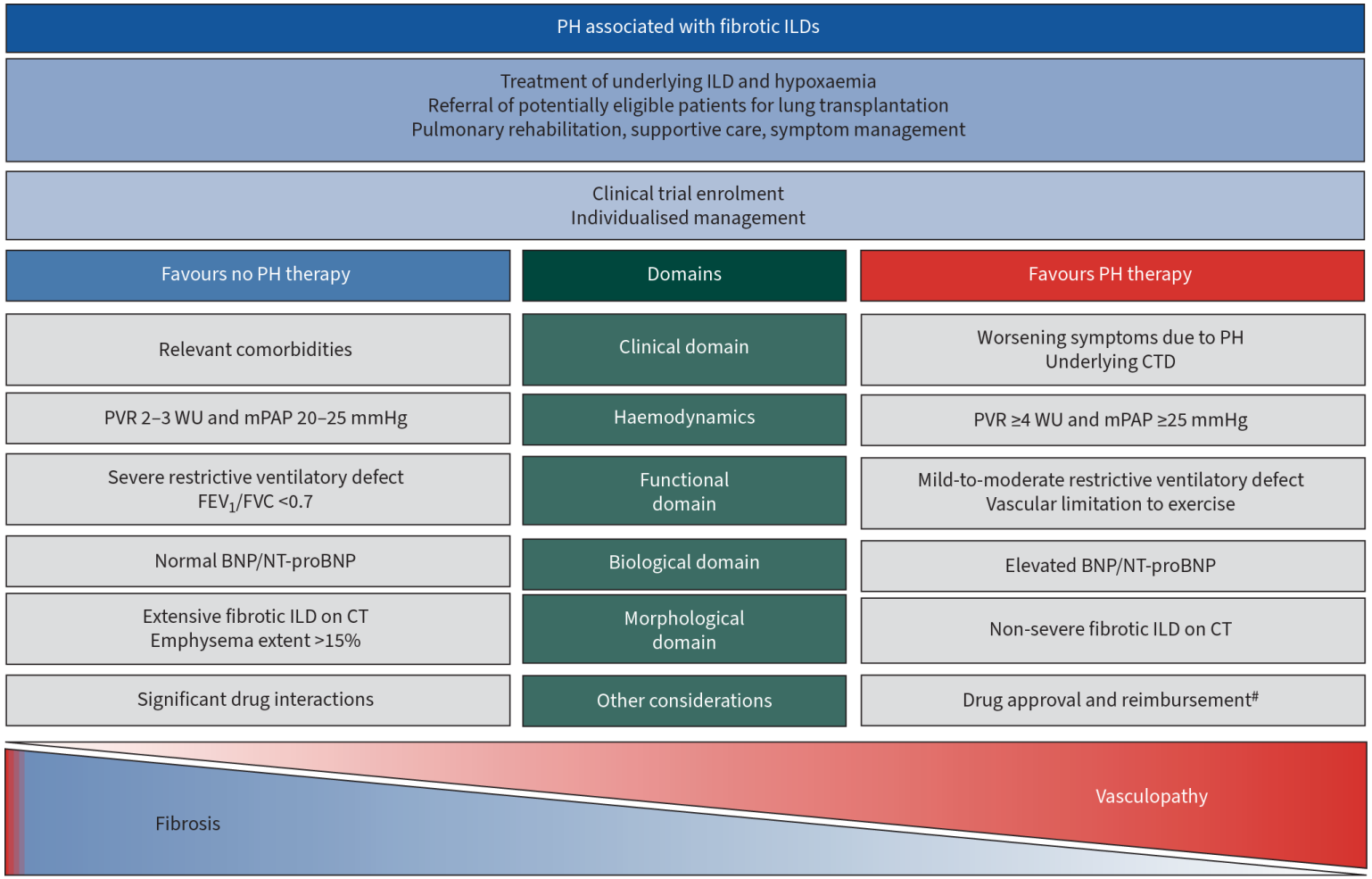
Treatment algorithm key points

a. The treatment algorithm is intended for patients with confirmed group 1 PAH (phenotypically clear-cut, including **mPAP ≥25 mmHg and PVR >3 Wood Units** and no significant response on acute vasoreactivity testing). See text for treatment in PAH with complex phenotypes.
b. ***Risk assessment*** should be performed at baseline, within 3–4 months and periodically thereafter, and using FC, 6MWD and natriuretic peptides as a part of a validated risk calculator. Haemodynamics, RV imaging and other measures should be used to supplement risk assessment.
c. ***Initial triple therapy*** with an *i.v./s.c.* PPA is recommended in high-risk patients and may be considered in non-high risk with severe haemodynamics and/or poor RV function.
d. Most ***low-risk patients*** at follow-up should continue initial therapy.
e. Clinical trials with oral and inhaled treprostinil included ***only patients on monotherapy***, while studies of selexipag and sotarcept included patients on combination therapy.
f. ***Transplant referral*** should be considered for select high-risk patients at diagnosis, and for intermediate-high and high-risk patients at first or subsequent follow-up.

FIGURE 1 Treatment algorithm. PAH: pulmonary arterial hypertension; IPAH: idiopathic PAH; HPAH: hereditary PAH; DT: drug and toxin; CTD: connective tissue disease; ERA: endothelin-1 receptor antagonist; PDE-5i: phosphodiesterase-5 inhibitor; *i.v.*: intravenous; *s.c.*: subcutaneous; PPA: prostacyclin pathway agent; sGCS: soluble guanylyl cyclase stimulator; Rx: prescription; mPAP: mean pulmonary artery pressure; PVR: pulmonary vascular resistance; FC: functional class; 6MWD: 6-min walk distance; RV: right ventricle.

Chin KM, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01325-2024

Fibrotic Lung Disease: To Treat or Not?



Shlobin OA, et al. *Eur Respir J*. Published online August 29, 2024. doi:10.1183/13993003.01200-2024

Don’t forget to complete the Evaluation!

The link will be emailed to you shortly.

Thank you!