PH Management in Unique Patients and Conditions

June 24, 2025

12:00 PM - 01:00 PM EDT

TOTAL CME

PATIENT CASE 1: DEBORAH

67-YEAR-OLD WOMAN

Case Description:

- Scleroderma and mild hypertension
- 6-month history of worsening dyspnea on exertion (DOE)
- Seen by her primary care provider (PCP) who referred her to a pulmonologist
- Pulmonologist ordered pulmonary function tests (PFTs) and an echocardiogram
  - PFTs: unremarkable
  - Echocardiogram:
    - Mild right ventricle (RV) enlargement and mild RV dysfunction
    - Right ventricular systolic pressure (RVSP) of 45
    - Mildly enlarged left atrium (LA)

Risk Score Assessment

REVEAL Lite 2 Risk Calculator

Directions: Select all variables that apply. **A minimum of 3 variables** are required to generate a score, where at least 2 are from most predictive values denoted **.

Step 1 Select at least 2 of the most predictive variables						Score
BNP (pg/mL)**	<50		50 to <200	200 to <800	≥800	
	–2		0	1	2	
OR						
NT-proBNP (pg/mL)**	<300		300 to <1100		≥1100	
	–2		0		2	
6MWT (m)**	≥440	320 to 440	<320 to 165	<165		-1
	–2	–1	0	1		
NYHA/WHO Functional Class**		I	II	III	IV	
		–1	0	1	2	
Step 2 Select additional variables.						
SBP (mm Hg)			SBP ≥110	SBP<110		
			0	1		
HR (BPM)			HR ≤96	HR >96		
			0	1		
eGFR < 60 mL/min/1.73m² or renal insufficiency			No	Yes		
			0	1		
				Step 3 Sum of above (min. 3 variables)		
						+6
				Step 4 Risk score		5

	Low Risk	Intermediate Risk	High Risk
Risk Score	≤5	6–7	≥8

BNP, B-type natriuretic peptide; BPM, beats per minute; DLCO, diffusing capacity of the lungs for carbon monoxide; eGFR, estimated glomerular filtration rate; mRAP, mean right atrial pressure; NYHA, New York Heart Association; 6MWT, 6-minute walk test; SBP, systolic blood pressure; WHO, World Health Organization.

Benza RL, et al. *Chest*. 2021;159(1):337-346.

Factors to Consider When Assessing Severity

For Deborah:

- Risk score
  - Score of 5 falls into low risk
- Echocardiogram results and cardiac function
  - Mild RV enlargement and mild RV dysfunction
  - RVSP of 45
  - Mildly enlarged LA
- Comorbidities
  - Scleroderma and mild hypertension

PATIENT CASE 1: DEBORAH

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Case Description:

- Additional testing needed:
  - Basic labs to evaluate for other possible contributing factors
  - Ventilation/perfusion (V/Q) scan to rule out chronic thromboembolic pulmonary hypertension (CTEPH)
  - Overnight oximetry to see if she needs oxygen at night
- PH center test results and treatment plan:
  - V/Q was negative
  - No oxygen needed on overnight oximetry
  - Repeat NT-proBNP 580
  - Started on a phosphodiesterase-5 inhibitor (PDE5i) and endothelin receptor antagonist (ERA)
    - Patient falls into the “not high risk” category
    - PDE5i – sildenafil – was started first, which she tolerated well
    - Developed some lower extremity edema (LEE) a few days after starting the ERA

Group 1 PAH: Managing LEE

- Counsel patients in advance
- Work to mitigate LEE, and any other side effects, as a first step
  - As opposed to withdrawing effective treatment
- Monitor labs, NT-proBNP, and weight over time
- Consider adding a diuretic, if appropriate

PATIENT CASE 1: DEBORAH

67-YEAR-OLD WOMAN

Case Description:

- PH center test results and treatment plan:
  - V/Q was negative
  - No oxygen needed on overnight oximetry
  - Repeat NT-proBNP was 580
  - Started on a PDE5i and an ERA
    - Patient falls into the “not high risk” category
    - PDE5i – sildenafil – was started first, which she tolerated well
    - Developed some LEE a few days after starting the ERA
- Returns after 3 months
  - Complains of worsening DOE and trouble climbing stairs in her home
  - Appears to be euvolemic after starting the diuretic

Is It PAH, or Is It Something Else?

Reassessment

- Is NT-proBNP rising?
- How does right heart appear upon repeat echocardiogram?
- Are there any new parameters, such as pericardial effusion?
- Has 6MWD changed?
- Is supplemental oxygen needed?
- What is her FC?
- Is RHC indicated?

Reassessment at 3 Months: Test Results

Parameter	Initial Status	Current Status	Trend
mPAP	32 mmHg	38 mmHg	↑ Worsening
PCWP	15 mmHg	12 mmHg	↓ Lower filling pressure
CO/CI	4.0 L/min, CI 2.0	3.8 L/min, CI 1.9	↓ Worsening
PVR	4.25 WU	6.8 WU	↑ Worsening
FC	II	III	↑ Worsening symptoms
BP	115/67	105/67	↓ Borderline low
HR	87 BPM	87 BPM	Stable
Echocardiography	Mild RV dysfunction	Moderate RV dysfunction, TAPSE 1.5, mild pericardial effusion	↓ Worsening
NT-proBNP	600	1,000	↑ Worsening
6MWD	370 m	315 m	↓ Worsening

TAPSE, tricuspid annular plane systolic excursion; WU, Wood units.

Reassessment at 3 Months: Risk Stratification

- ESC/ERS 4-strata model
  - Provides better discrimination within the intermediate-risk group, which helps guide therapeutic decision-making

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

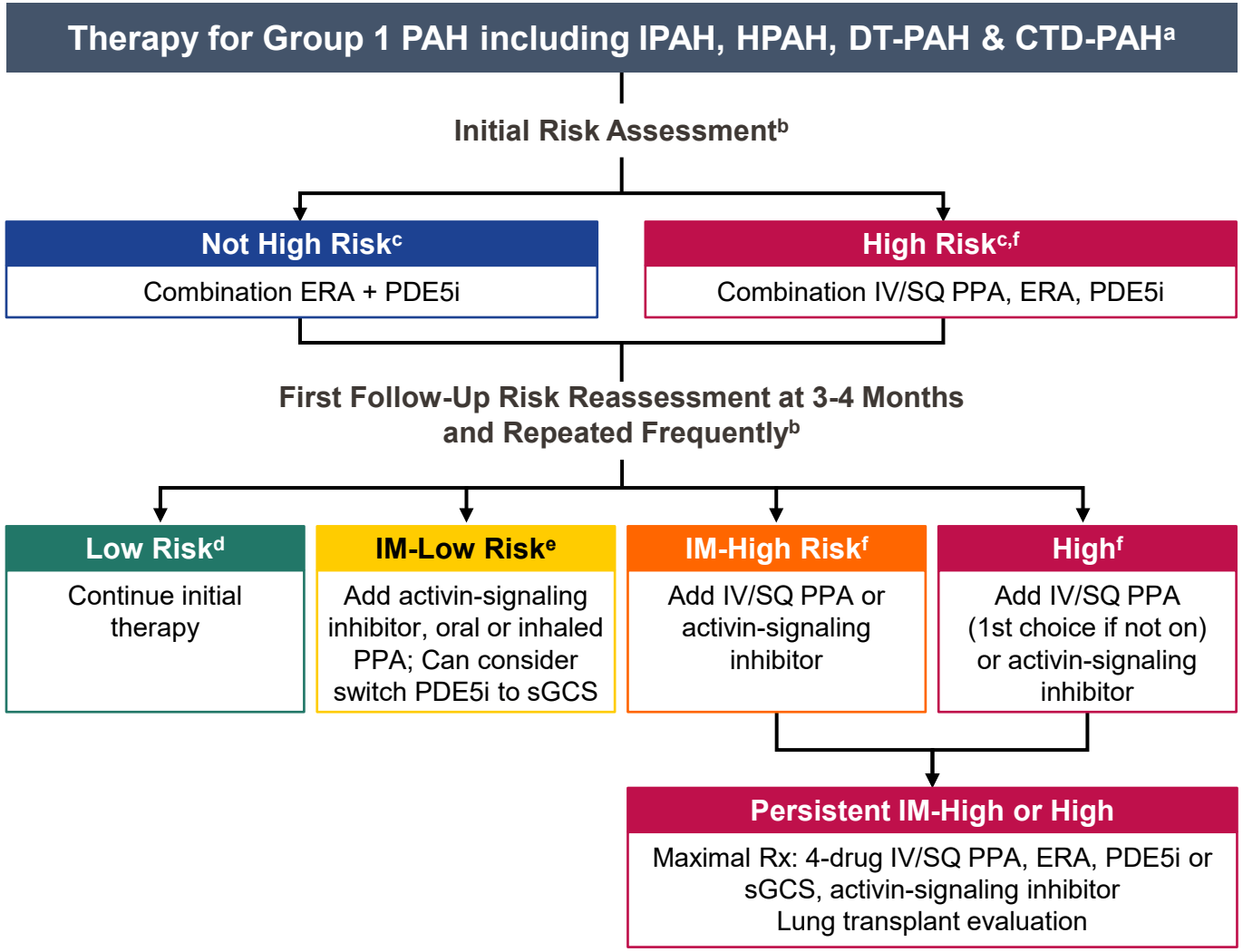
[a]WHO-FC I and II are assigned 1 point as both are associated with good long-term survival.
ERS, European Respiratory Society; ESC, European Society of Cardiology.
Humbert M, et al. *Eur Heart J*. 2022;43(38):3618-3731. Humbert M, et al. *Eur Respir J*. 2023;61(1):2200879.

PAH Does Not Always Remain Stable

Reassessment

- At 3-4 months, or earlier if patient is having problems
- Perform risk assessment and watch for changes
  - Consider continuing therapy for patients who remain at low risk
  - For other risk groups or a change in risk, reevaluate therapy

7th WSPH Treatment Algorithm



Treatment Algorithm Key Points

a. 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). Treatment nuances 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. Hemodynamics, RV imaging, and other measures should be used to supplement risk assessment.

c. Initial triple therapy with an IV/SQ PPA is recommended in high-risk patients and may be considered in non-high risk with severe hemodynamics and/or poor RV function.

d. Most ***low risk*** at follow-up patients should continue initial therapy.

e. Clinical trials with oral and inhaled treprostinil included ***only patients on monotherapy***, while studies of selexipag and sotatercept included patients on combination therapy.

f. Transplant referral should be considered for select high-risk patients **at diagnosis**, and for IM-high and high-risk patients at ***first*** or subsequent follow-up.

Chin KM, et al. *Eur Respir J*. 2024;64(4):2401325.

Key Takeaways

- A complete diagnostic workup, including risk assessment to guide therapy decisions, is important
- Follow patients closely and escalate therapy in cases of worsening PH
- A combination ERA/PDE5i tablet is available that may be appropriate for certain patients
- Proactive side effect management is important, especially when starting prostacyclin or advanced therapy
- Diagnosing condition as true PAH is key
- Monitor patients closely and avoid complacency, especially in cases of low risk
- Be on the lookout for any worsening in order to react quickly and manage appropriately