Value of an Interim Patient Reported Outcome Measure by a Specialty Clinician

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INTRODUCTION:

Introduction: The recent evidence-based treatment algorithm in the management of pulmonary arterial hypertension (PAH) was proposed at the 7th World Symposium on Pulmonary Hypertension and published in the European Respiratory Journal in 2024 (figures 1, 2 and 3). Monotherapy is no longer recommended. Frequent reassessment and risk stratification is suggested to determine appropriateness for escalation of therapy. Risk is based on laboratory values (BNP, NT-proBNP), functional class (FC), and 6-minute walk test distance (6-MWD). FC and 6-MWD correlate to the Living with PH Questionnaire (LPHQ) (figure 4): administration can be facilitated by a specialty pharmacy clinician.

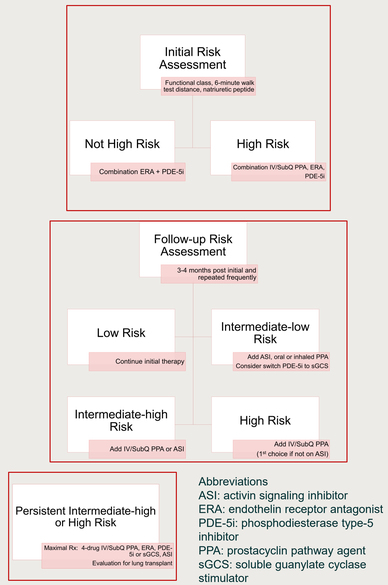
Figure 4

LIVING WITH PAH SURVEY (LPHQ)

Responses: on a scale of 0-5, with 0 being no interference and 5 being extreme interference did your PAH prevent you from living as you wanted during the past 7 days by:

- ❑Causing swelling in your ankles, legs?
- ❑Making you sit or lie down to rest during the day?
- ❑Making it difficult to walk about or climb stairs?
- ❑Making it difficult to work around the house or in the garden?
- ❑Making it difficult to go anywhere away from home?
- ❑Making it difficult to sleep well at night?
- ❑Making it difficult to have relationships or do things with your friends or family?
- ❑Making it difficult to work to earn a living?
- ❑Making your recreational pastimes, sports or hobbies difficult?
- ❑Making your sexual activities difficult?
- ❑Making you eat less of the foods you like?
- ❑Making you short of breath?
- ❑Making you tired, fatigued or lacking in energy?
- ❑Making you stay in the hospital?
- ❑Costing you money for medical care?
- ❑Giving you side effects from treatments?
- ❑Making you feel you are a burden to your family or friends?
- ❑Making you feel a loss of self-control in your life?
- ❑Making you worry?
- ❑Making it difficult for you to concentrate or remember things?
- ❑Making you feel depressed?

Figures 1, 2 and 3



Abbreviations
ASI: activin signaling inhibitor
ERA: endothelin receptor antagonist
PDE-5i: phosphodiesterase type-5 inhibitor
PPA: prostacyclin pathway agent
sGCS: soluble guanylate cyclase stimulator

METHODS:

A report identifying patients with at least two LPHQ scores between 12-20-2023 and 12-20-2024 was generated. Medical records were retrospectively reviewed for precise PAH diagnosis, FC at the time of the most recent medication addition, and therapy regimen by classification. Additional LPHQ scores were added when identified. On manual chart review. Patient data were included in the final deidentified dataset when the following criteria were met – aged 18-89, diagnosis WHO group I PAH, first/last scores a minimum of four weeks apart, at least one score greater than zero and drug covered by a payer without data use restrictions.

RESULTS:

Eighty-seven records met full inclusion criteria: 29% [25/87] monotherapy, 32% [28/87] dual therapy, 26% [23/87] triple therapy, and 13% [11/87] quadruple therapy (figure 5). Monotherapy was most often achieved with an sGC stimulator, dual therapy added an ERA, triple therapy a PPA and quadruple therapy an ASI (figure 6).

Figure 5

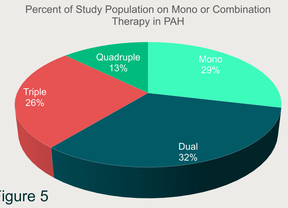
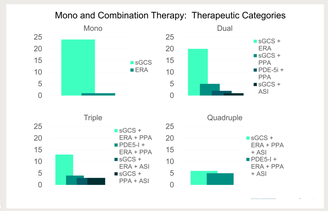


Figure 6



LPHQ scores ranged from 0-73 (median 27) for patients on monotherapy, 0-97 (median 23) for dual therapy, 4-85 (median 33) for triple therapy, and 6-79 (median 30) for quadruple therapy.

Patients receiving mono, dual and triple therapy were more likely to show improved LPHQ scores over time, 68% (17/25), 75% (21/28) and 70% (16/23) respectively, while only 27% (3/11) of patients on quadruple therapy met the same benchmark (figures 7-10).

Figure 7

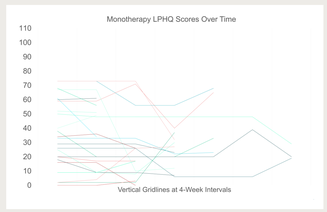


Figure 8

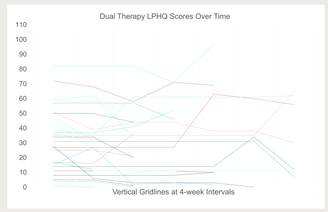


Figure 9

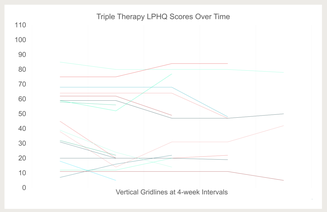
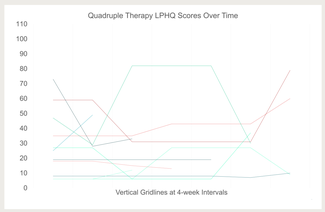


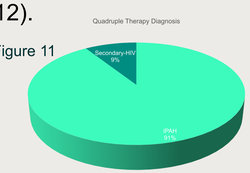
Figure 10



QUADRUPLE THERAPY:

The first ASI was FDA approved in March 2024. This agent has made quadruple therapy (with a PDE-5i or SGCs, ERA, and PPA) possible. A subanalysis of the eleven patients receiving quadruple therapy revealed that the majority (10/11; 91%) carried a diagnosis of idiopathic PAH (figure 11). PPA therapy was most often achieved with an oral medication (8/11; 73%). Of note, no patients on quadruple were receiving a PPA via the inhaled route of administration (figure 12).

Figure 11

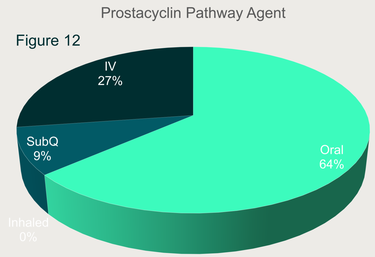


Breakdown of PPA dosing can be found in table 1.

PPA	Dose	Patients
treprostinil IV	80ng per kg per min	1
treprostinil subQ	54ng per kg per min	1
treprostinil oral	1.375 po tid	1
selexipag	800mcg po bid	1
selexipag	1200mcg po bid	3
selexipag	1600mcg po bid	1

Table 1

Figure 12



Three patients received the PPA from an alternate specialty pharmacy and exact dosing was unknown.

LIMITATIONS:

LPHQ is most often administered during an in-home skilled nursing visit, therefore sample is skewed toward those medications with this level of patient support (i.e., sGCS, PPA, ASI). Additionally, it was difficult to definitively determine the timing of the survey to the extract start of therapy for all drugs – particularly those provided by an alternate specialty provider.

CONCLUSION:

Administration of the LPHQ by a specialty pharmacy clinician provides a valuable interim outcome measure between medical appointments which may assist in informing appropriateness for escalation of therapy through an evidence-based treatment algorithm.

REFERENCES and DISCLOSURES

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*Salary, benefits, stock associated with employment at The Cigna Group