Poster #CO114

Humanistic Burden in Patients With Pulmonary Hypertension Associated With Interstitial Lung Disease Using Patient-Reported Outcome Measures

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WHAT WAS KNOWN

- PH-ILD is associated with substantial burden, including severe symptoms and worsening functional status, which can impact many aspects of patients' lives[1-3]
- In patients with PH-ILD, physical, social, and emotional functioning can be impaired and may lead to personal/social isolation, anxiety and depression, stress, fear, sadness, sleep disorders, and financial burdens[4-6]

OBJECTIVES

- This study assessed the humanistic burden in patients with PH-ILD using generic and disease-specific PROMs
  - PROMs scores were differentiated across WHO-FC groups to assess burden among patients with different disease severity

METHODS

Data Source and Methodology

- Data were analyzed from the Adelphi Real World PH-ILD DSP™, a retrospective cross-sectional survey of physicians and their patients diagnosed with PH-ILD in the US, UK, France, Germany, Italy, and Spain (EU4+UK), conducted between November 2024 and June 2025[7-10]
- Physicians were recruited in a geographically representative manner by local fieldwork agencies
- Physicians completed patient-record forms for up to 3 patients with a confirmed PH-ILD diagnosis, using clinical records, their own judgement, and diagnostic/interpretation skills. Patients for whom a form was completed were invited to complete a survey
- Generic PROMs included the SF-12 PCS for the US and the EQ-5D-5L and EQ-5D-VAS scores for EU4+UK, disease-specific instruments included the L-PF Impacts Module total score and emPHasis-10 total score

Physician and Patient Inclusion Criteria

- Physicians were eligible if they were pulmonologists, cardiologists, or internal medicine specialists (France only) managing patients with PH-ILD and seeing at least 3 patients in a typical month
- Included patients had a physician-confirmed diagnosis of PH-ILD, were ≥18 years old, and were not involved in a clinical trial at survey date

Statistical Analysis

- Comparisons between WHO-FC groups (FC **I** vs FC **II** vs FC **III**/IV) were made using ANOVA, followed by Bonferroni-adjusted pairwise comparisons
  - Due to the small number of patients with FC IV, all analyses were conducted with data for FC **III** and IV combined
- EQ-5D utility scores were derived using the UK crosswalk value set[11]
- Correlations between generic and disease-specific measures were assessed using Pearson's correlation coefficient and interpreted according to Schober and Boer[12]

RESULTS

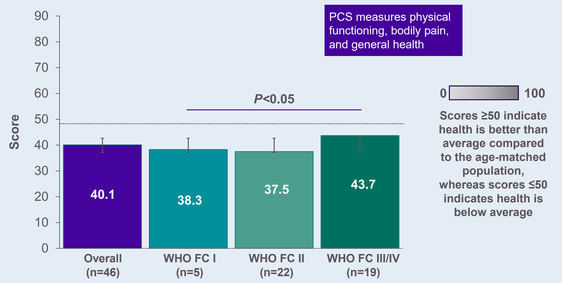
Data Collection and Patient Demographics

- Physician and patient-reported data were available for 234 patients
- For patients completing a questionnaire, mean (SD) age was 64.7 (10.5) years, 44.0% were female, and 94.6% were White
- At survey date, most patients were WHO FC **II** (52% [122/234]), followed by WHO FC **III** (32% [75/234]); WHO FC groups **I** and IV comprised 10% (24/234) and 6% (13/234), respectively
- Idiopathic pulmonary fibrosis was the most predominant ILD subtype (45.3%)
- The most common (≥15%) comorbidities at the survey date were hypertension (55.1%), dyslipidemia (26.5%), elevated cholesterol/hyperlipidemia (20.1%), and cardiac arrhythmias (15.0%)
- At the most recent test, 52.7% of patients (n=93) had a 6-minute walk distance of <250 m
- Physicians in the EU4+UK (n=141) reported consulting their patients in an academic setting (59.0%), whereas in the US, physicians (n=35) reported seeing their patients in an office setting (74.9%). Overall, physicians reported being affiliated with a PH/ILD or transplant center (64.8% [144/176])
- Overall, most physicians reported low current involvement in clinical trials across PH-ILD (8.0% [14/176])

Patient-Reported Data

- The mean (SD) SF-12 PCS was 40.1 (6.8), indicating below-average health for patients in the US compared to the age-matched population[13] (**Figure 1**)
- PCS scores were generally similar when stratified by WHO FC groups (**Figure 1**)

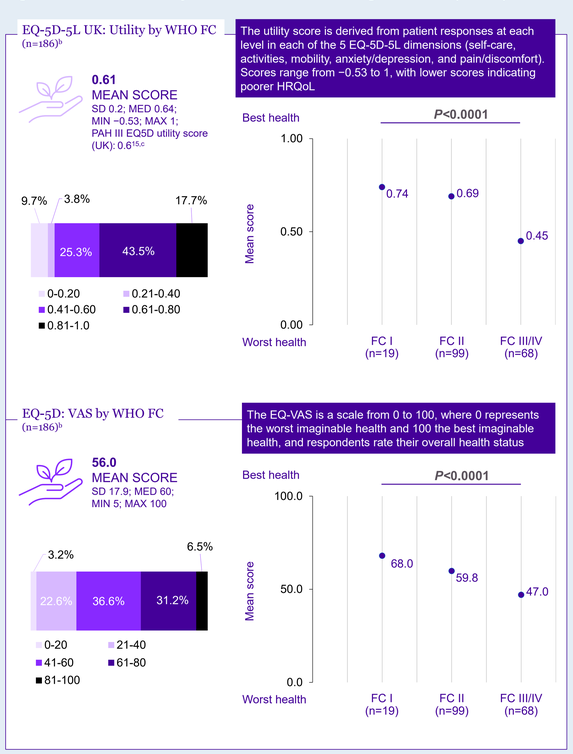
Figure 1. PCS SF-12 Score in US patients with PH-ILD (n=46)



Patients were classified into WHO-FC groups[14] as follows: FC I: No limitations. Ordinary physical activity does not cause undue dyspnea (shortness of breath), fatigue, chest pain, or near-syncope; FC II: Slight limitations. Comfortable at rest, but ordinary physical activity causes symptoms; FC III: Marked limitations. Comfortable at rest, but less than ordinary activity causes symptoms; FC IV: Inability to perform any physical activity without symptoms. Symptoms of right-heart failure (eg, edema) and dyspnea at rest may be present.

- Overall, mean (SD) scores for EQ-5D-5L utility and EQ-5D VAS were 0.61 (0.2) and 56.0 (17.9), respectively (**Figure 2**), indicating poor HRQoL
- Both generic measures worsened with deteriorating WHO-FC, with higher scores in FC **I** (utility, 0.74; VAS, 68.0) versus FC **III**/IV (utility, 0.45; VAS, 47.0; both *P*<0.001; **Figure 2**)

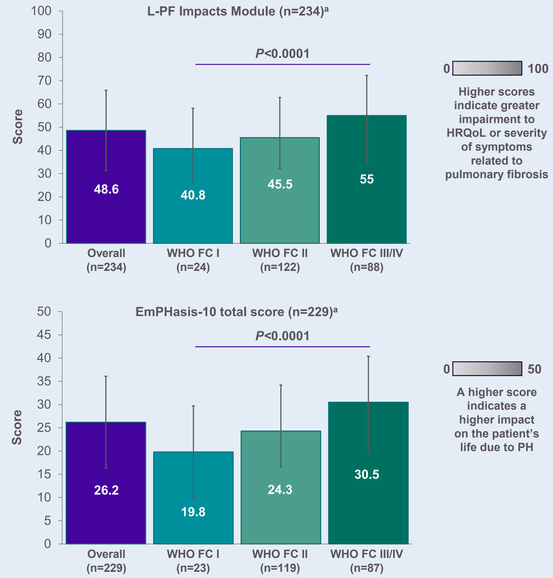
Figure 2. EQ-5D-5L Utility and EQ-5D Visual Analogue Scores (n=186)[a]



[a]EU4 (France, Germany, Italy, Spain) + UK patients only.
[b]P<0.05 indicates a significant difference between the WHO FC groups.
[c]EQ-5D-5L utility score in the PAH DSP population ranged from 0.6 in the UK to 0.9 in France.

- For disease-specific instruments, mean (SD) for L-PF Impacts domain and EmPHasis-10 scores were 48.6 (17.3) and 26.2 (9.9), respectively, with higher scores in FC **III**/IV compared with FC **II** and FC **I** (L-PF Impacts: 55.0 vs 45.5 and 40.8; EmPHasis-10: 30.5 vs 24.3 and 19.8; all *P*<0.001); **Figure 3**)

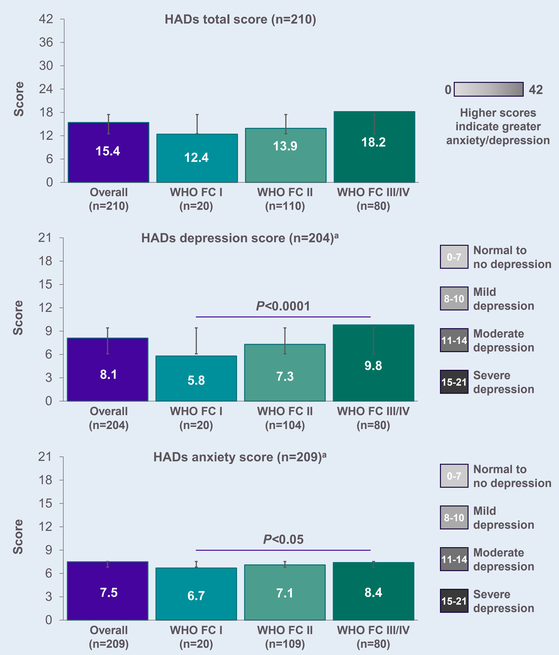
Figure 3. Disease-Specific Instruments: L-PF Impacts Module and EmPHasis-10 Total Scores



[a]P<0.05 indicates a significant difference between the WHO FC groups.

- Mean HADS total score indicated mild to moderate levels of anxiety and depression in patients with PH-ILD (**Figure 4**)
- The HADS depression and anxiety scores worsened with deteriorating WHO-FC (**Figure 4**)
  - Depression score: FC **III**/IV (9.8) compared with FC **I** (5.8) and FC **II** (7.3); both *P*<0.001
  - Anxiety score: FC **I** (6.7), FC **II** (7.1), and FC **III**/IV (8.4); overall *P*<0.05

Figure 4. Hospital Depression and Anxiety Scale Scores



[a]P<0.05 indicates a significant difference between the WHO FC groups.

- Correlation analyses suggested strong negative associations between L-PF and SF-12 PCS (r=−0.80, *P*<0.001); moderate negative associations were observed between L-PF and EQ-5D VAS and EQ-5D utility scores (r=−0.55, *P*<0.001; r=−0.57, *P*<0.001, respectively)
- Moderate negative associations were also reported between EmPHasis-10, PCS SF-12, EQ-5D VAS, and EQ-5D utility scores (r=−0.65, *P*<0.001; r=−0.62, *P*<0.001; r=−0.63, P<0.001, respectively)
  - For the disease-specific measures (EmPHasis-10 and L-PF Impacts), higher scores suggest worsening severity, whereas, for the generic measures (SF-12 PCS, 5D VAS, and EQ-5D), lower scores indicate worsening severity. Therefore, these negative associations suggest concordance between disease-specific and generic measures

WHAT THIS STUDY ADDS

- Most PROM scores differentiated across WHO-FC groups, suggesting responsiveness to worsening PH-ILD severity with disease-specific instruments (EmPHasis-10 and L-PF Impacts) being particularly sensitive to worsening WHO-FC (ie, symptoms and physical function)
  - Correlation analyses highlighted moderate to strong associations between generic and disease-specific measures
- Generic and disease-specific PROMs provided complementary insights into the substantial humanistic burden on patients with PH-ILD
  - Overall, generic measures indicated a greater disease burden in these patients compared to the age-matched general population. Patients reported lower physical functioning (SF-12 PCS) across all WHO FC severity groups and overall health scores (EQ-5D-5L Utility) that decreased with increasing severity groups, all indexes of poor HRQoL
  - Additionally, disease-specific instruments (L-PF, Emphasis-10, and HADS scores) demonstrated impairment in HRQoL, negative impact on patients' lives, and increased anxiety and depression that worsened with increasing FC severity
- Limitations of the study include the following:
  - Data across functional classes were collected in different patients, not across the disease trajectory in each patient
  - The disease-specific instruments assessed in this study have not been validated in patients with PH-ILD
  - The DSP™ is not based on a true random sample of physicians or patients. Though minimal inclusion criteria governed the selection of the participating physicians, participation was influenced by patient and physician willingness to complete the survey
  - Identification of the target patient group was based on the judgment of the respondent physician and not a formalized diagnostic checklist, but was representative of the physician's real-world classification of the patient
  - Physicians were asked to provide data for a consecutive series of patients to avoid selection bias, but no formal patient selection verification procedures were in place

ABBREVIATIONS:
ANOVA, analysis of variance; DSP, Disease Specific Programme; EmPHasis-10, 10-item Pulmonary Hypertension quality-of-life questionnaire; FC, functional class; HADS, Hospital Depression and Anxiety Scale; HRQoL, Health-Related Quality of Life; ILD, interstitial lung disease; L-PF, Living With Pulmonary Fibrosis; Med, median; PCS, Physical Component Summary; PH, pulmonary hypertension; PH-ILD, pulmonary hypertension associated with interstitial lung disease; PROMs, patient-reported outcome measures; SF-12, 12-Item Short Form Health Survey; US, United States; UK, United Kingdom; VAS, visual analogue scale; WHO, World Health Organization.

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DISCLOSURES:
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