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RESEARCH LETTER

Real-life use of validated quality of life questionnaires in patients with pulmonary arterial hypertension in Spain: a cross-sectional study

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This study aimed to determine how often health-related quality of life (HRQoL) is assessed in patients with pulmonary arterial hypertension (PAH) and to elucidate which tools are most frequently used in a real-life setting. We conducted a multicentric retrospective cross-sectional analysis of 3628 PAH patients enrolled in the Spanish Registry of Pulmonary Arterial Hypertension (REHAP) from 2007 to 2023. HRQoL questionnaires were used in only 67 patients (1.8%). Among them, generic tools were the most used (SF-36 in 16.9% and EQ5D in 10.2%). These findings highlight a marked underutilization of HRQoL assessment tools in PAH patients included in REHAP.

Keywords PROMS • Pulmonary arterial hypertension • Quality of life

Novelty

- Notably, there is a significant underutilization of validated pulmonary arterial hypertension (PAH)-specific questionnaires for measuring health-related quality of life (HRQoL) in Spain.
- Highlighting the inadequate assessment HRQoL in patients with PAH in everyday settings may serve as an initial step towards the broader adoption of these evaluative tools.
- An increased application of validated HRQoL questionnaires could foster interest in implementing non-pharmacological interventions and address the personal needs of this highly vulnerable population.

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Introduction

Pulmonary arterial hypertension (PAH) is a rare and disabling disease, which is characterized by progressive pulmonary vascular remodelling and increased pulmonary vascular resistance. It is included in Group 1 of the WHO Classification of Pulmonary Hypertension (PH) and is defined by the combination of mean pulmonary arterial pressure (mPAP) > 20 mmHg, a pulmonary vascular resistance > 2 Woods Units in the presence of a normal pulmonary capillary wedge pressure. Symptoms are usually non-specific such as dyspnoea, fatigue, decreased exercise tolerance, lack of energy, and syncope. Progression of the vasculopathy can trigger to right heart failure, the leading cause of death related to PAH.[1]

Despite the recent therapeutic advances, PAH currently has no cure and the burden of the disease on patients' lives remains high. Disabling symptoms, oxygen therapy, frequent hospitalizations, and both drug adverse effects, can negatively impact on patients' quality of life (QoL).[2] Health-related quality of life (HRQoL) instruments aim to measure impairment and assess its impact on individual's perceived social, psychological and physical well-being. Most HRQoL instruments comprise multiple domains, which vary in scope and content.[2] Recently, recommendations for obtaining Patient Reported Outcomes (PROs) in PAH have increased. The Nice World Symposium of Pulmonary Hypertension (WSPH) in 2018 and 2022 European guidelines strongly recommend this factor to be assessed.[1] At the 2024 WSPH, there was a strong emphasis on the critical importance of assessing it.[3]

Compared to generic or 'non-PAH-specific' questionnaires such as SF-36 or EQ-5D,[4,5] condition-specific measures such as CAMPHOR, emPHasis-10, and PAH-SYMPACT[6–8] may be more sensitive to treatment-related changes, as they address patients' most important concerns. However, the use of these instruments in clinical practice has only recently been evaluated.[9] Despite strong recommendations highlighting the importance of assessing HRQoL in PH patients, its implementation in routine clinical care remains limited. Between 2011 and 2022, the use of HRQoL assessments increased from 20% to just 65% across 20 high-volume PH centres in Europe.[10] Similarly, a 2021–2022 survey of 30 PH centres revealed that only half of the centres routinely implemented patient-reported outcome measures (PROMs) in clinical care (personal communication, ERN-LUNG). To our knowledge, there is limited evidence on the real-world use of HRQoL assessment in these patients. Although such assessments may provide valuable prognostic information and are widely recommended by experts, they remain underutilized.[1]

Table 1 Demographic and clinical characteristics of patients with pulmonary arterial hypertension included in Registry of Pulmonary Arterial Hypertension

	PAH patients Total	PAH patients until 2018	PAH patients 2019–2023	*P*-value
N (% from total)	3628	2729	899	
Age, mean ± SD	50.17 ± 18.13	48.17 ± 18.26	56.23 ± 16.22	<0.001
Female, *n* (%)	2458 (67.8)	1866 (68.4)	592 (65.9)	0.162
PAH aetiology				
Idiopathic, *n* (%)	1133 (31.2)	819 (30)	314 (34.9)	0.006
Hereditary, *n* (%)	56 (1.5)	35 (1.3)	21(2.3)	0.041
Connective tissue disease, *n* (%)	834 (23)	570 (20.9)	264 (29.4)	<0.001
CHD, *n* (%)	809 (22.3)	700 (25.7)	109 (12.1)	<0.001
PVOD, *n* (%)	184 (5.1)	128 (4.7)	56 (6.2)	<0.001
Toxics, *n* (%)	91 (2.5)	75 (2.7)	16 (1.8)	
Others, *n* (%)	521 (14.4)	402 (14.7)	119 (13.2)	0.068
Functional class				
NYHA. I, *n* (%)	191 (5)	134 (4.9)	57 (6.3)	
NYHA II, *n* (%)	1341 (37)	980 (35.9)	361(40.2)	0.013
NYHA III, *n* (%)	1805 (49.8)	1380 (50.6)	425 (47.3)	
NYHA IV, *n* (%)	291 (8)	235 (8.6)	56 (6.2)	
PVR (WU), mean ± SD	10.12 ± 6.12	10.61 ± 6.33	8.71 ± 5.24	<0.001
TAPSE, mean ± SD	17.56 ± 5.11	17.55 ± 5.45	17.55 ± 5.45	0.989
QOL Questionnaires	**67 (1.8)**	**58**	**9**	<0.001
SF-36, *n* (%)	10 (16.9)	8 (15.7)	2 (25)	<0.001
EQ-5D, *n* (%)	6 (10.2)	4 (7.8)	2 (25)	
EMPHASIS-10, *n* (%)	2 (3.4)	0	2 (25)	
Adapted CHD, *n* (%)	3 (5.1)	4 (5.9)	0	
KCCQ, *n* (%)	2 (3.4)	0	2 (25)	
Non specificized, *n* (%)	44 (65.67)	42 (72.41)	1 (12.5)	

Variables shown in bold correspond to quality of life questionnaire data, which represent the core focus of the study.
CHD, congenital heart disease; EQ-5D, EuroQol 5 dimensions questionnaire; KCCQ, Kansas City Cardiomyopathy Questionnaire; NYHA, New York Heart Association; PAH, pulmonary arterial hypertension; PVOD, pulmonary venooclusive disease; PVR, pulmonary vascular resistance; QOL, quality of life; SD, standard deviation.

Thus, the aim of this study was to find out how often HRQoL was assessed in patients with PAH at the time of diagnosis, as well as to elucidate which tools were the most frequently used.

Methods

We conducted a multicentric retrospective cross-sectional analysis of 3628 PAH patients enrolled in the Spanish Registry of Pulmonary Arterial Hypertension (REHAP). REHAP is a voluntary national registry that gathers clinical data of patients aged ≥14 years diagnosed with PAH from 58 Spanish hospitals from 2007 and ongoing. The promoting centre's ethical committee authorized the study. The study was appraised and accepted by the institutional review board or local committee of each participating hospital. All patients signed informed consent.

Diagnosis was haemodynamically defined by mPAP ≥ 25 mmHg measured by right heart catheterization until 2022. After the publication of the 2022 European Guidelines, definition of haemodynamic PH was redefined to mPAP ≥ 20 mmHg. Clinical and demographic variables were collected at baseline, as 'QOL Questionnaire'. Patients were stratified into 2 groups according to the year of diagnosis, 2007–2018 and 2019–2023. This stratification reflects the publication of the first formal recommendation for QoL assessment in PAH, issued in 2018.

An exploratory analysis was performed through the calculation of the central tendency and dispersion or percentages. Bivariate analyses were carried out using Student's *t*-tests for independent samples and chi-square to independent samples and to the characteristics of the variables analyzed. All the analyses were performed with the SPSS v.25 package (IBM Corp., Armonk, NY, USA).

Results

Patients with pulmonary arterial hypertension were mostly female (67.8%), with a mean age of 50.17 years SD 18.1. The most frequent aetiology was Idiopathic Pulmonary Arterial Hypertension (31.2%), followed by Connective Tissue Disease PAH (CTD-PAH) (23%) and Congenital Heart Disease (CHD-PAH) (22.3%). Most of them were in NYHA functional class II and III (37% and 49.8%, respectively). Detailed demographic and clinical characteristics of the population are described on *Table 1*.

The HRQoL of only 58 (2.1%) and 9 patients (1%) was analyzed, until 2018 and 2019–2023, respectively ($P < 0.01$). Generic tools were more often used [SF-36 10 (16.9%) and EQ5D in 6 (10.2%)] than PAH-specific tools (EMPHASIS-10 in only 2 patients). However, in 44 (65.7%), the tool used was not reported (*Table 1*).

Discussion

In our study, we found an extremely low rate of assessment of HRQoL among patients with PAH in a real-life setting. Although this under-use of PROs instruments in clinical practice has been frequently assumed in literature,[1] this is, to the best of our knowledge, the first study to quantify the actual proportion of PAH patients in whom QoL was assessed as part of routine care.

Despite the increasing importance of addressing PROs in contemporary health, especially in such a disabling disease as PAH, the lack of accessibility to validated and cross-culturally adapted questionnaires to Spanish, as well as the high fees required for using some of these instruments, may be some of the main reasons for their under-use. Furthermore, there is also a lack of awareness about the importance of measuring some important constructs such as QoL by health-care professionals. A more extensive use of these instruments could promote the implementation of non-pharmacological interventions and address the personal needs of such a highly vulnerable population.

Limitations of this study include (i) potential missing QoL data in the registry; (ii) as the data were drawn from a national cohort in Spain, the generalizability of our findings to other healthcare systems or cultural contexts may be limited.

However, we believe that sharing these results with the PAH health carers is the first step towards a paradigm shift. Encouragingly, the REHAP registry is currently working in the implementation of better and easier tools for the assessment of QoL in PAH patients.

Author contributions

Ada del Mar Carmona-Segovia (MsC RN), Víctor M. Becerra-Munoz (PhD (Supervision: Lead; Writing—review & editing: Lead)), Josefa Jiménez Arjona (Data curation: Equal), Virginia Naranjo (Data curation: Equal), Nuria Ochoa Parra (Data curation: Lead; Investigation: Lead), Joan Albert Barberà PhD MD (Project administration: Equal)), Manuel López Meseguer (Writing—review and editing: Lead), José Miguel Morales Asencio (PhD (Supervision: Lead; Validation: Lead)), Isabel Blanco (Project administration: Equal), and Pilar Escribano Subías (Conceptualization: Lead; Project administration: Lead; Writing—review & editing: Lead)

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Conflict of interest: The authors declare no conflicts of interest related to this report.

Data availability

The data used in this study come from the REHAP (Registro Español de Hipertensión Arterial Pulmonar) cohort. Due to confidentiality agreements and data protection policies, the dataset is not publicly available. Data are available on request from the corresponding author.

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