R-scale for pulmonary fibrosis: a simple, visual tool for the assessment of health-related quality of life

Ciaran Scallan [1,2], Lauren Strand[3], Jennifer Hayes[1], Suha Kadura [1], Bridget Collins[1], Lawrence Ho[1], Carolyn Spada[1], Will Canestaro[3,4], Martin Kolb [2] and Ganesh Raghu[1]

[1]Center for Interstitial Lung Disease, University of Washington, Seattle, WA, USA. [2]Firestone Institute for Respiratory Health, McMaster University, Hamilton, ON, Canada. [3]School of Pharmacy, University of Washington, Seattle, WA, USA. [4]Washington Research Foundation, Seattle, WA, USA.

Corresponding author: Ganesh Raghu (graghu@uw.edu)

Shareable abstract (@ERSpublications)
Health-related quality of life (HRQoL) can be significantly impacted in patients with idiopathic pulmonary fibrosis. This article describes a novel, rapid tool to assess HRQoL in the context of routine clinical encounters with good concurrent validity. https://bit.ly/3yUSex2

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Abstract

Rationale Patients with idiopathic pulmonary fibrosis (IPF) experience impaired health-related quality of life (HRQoL). Several tools have been developed to objectively assess HRQoL in this patient population, but none are in use in routine clinical practice.

Objectives To develop a rapid, specific tool that can be used for patients with IPF during routine clinic visits.

Methods A novel and simple five-item numerical rating scale was developed and compared with two other previously validated tools. 100 consecutive patients with IPF managed at a centre for interstitial lung disease were recruited to complete the Raghu scale for pulmonary fibrosis (R-Scale-PF), King's Brief Interstitial Lung Disease questionnaire (K-BILD), and the EuroQol Five-Dimensional Five-Level questionnaire (EQ-5D-5L) in addition to pulmonary function and 6-min walk tests.

Measurements and main results All 100 patients successfully completed the three HRQoL tools with 53 completing them again at follow-up visits. Internal consistency was high (Cronbach's α 0.825) with minimal floor/ceiling effect. Concurrent validity of the R-Scale-PF was moderate to high compared with the K-BILD (r=−0.713) and the EQ-5D-5L (r=−0.665). Concurrent validity was moderate with physiologic measures (forced vital capacity, r=−0.307, 6-min walking distance, r=−0.383). The R-Scale-PF demonstrated good known-groups validity when comparing scores across stages of disease severity.

Conclusions The R-Scale-PF correlates well with the K-BILD and EQ-5D-5L. It is hoped that this novel simple numerical rating scale tool, subject to validation in patients from other centres, will provide an opportunity to objectively measure HRQoL in routine clinical practice for patients with IPF.

Introduction

Idiopathic pulmonary fibrosis (IPF) is a chronic and progressive fibrotic lung disease causing impaired lung function and symptoms including dyspnoea and chronic cough. As disease severity increases, patients with IPF experience an increase in symptom burden that has a negative impact on health-related quality of life (HRQoL) [1]. In general, quality of life (QoL) refers to an individual's perception of their needs and position in life in the context of their culture and values [2]. Several tools have been used for the assessment of patient reported HRQoL in IPF which have been widely used in a research context but were not designed for and have not been used in routine clinical practice to date. Additionally, none of these tools used a simple numerical rating scale (NRS) to facilitate rapid assessment of symptoms and HRQoL in a routine clinical encounter. The aim of this study was to develop and evaluate the validity and reliability of a novel, simple NRS allowing for rapid assessment of symptoms and HRQoL among patients with IPF during routine clinic visits.

Methods

Instrument development

The initial concept of the Raghu scale for pulmonary fibrosis (R-Scale-PF) was developed by Ganesh Raghu at the Center for Interstitial Lung Disease, University of Washington (Seattle, WA, USA). To better understand the determinants of HRQoL and the impact of symptoms associated with IPF, participants were recruited from the local pulmonary fibrosis support group and the interstitial lung disease (ILD) clinic at the University of Washington. Several focus group sessions were conducted with the goal of identifying the factors and symptoms having a direct impact on HRQoL from a patient perspective. We recorded general themes and specific phrases generated by focus group members to facilitate concept elicitation and the generation of a list of the most important factors contributing to HRQoL from a patient perspective. Through discussion with the focus group, we identified an initial list of 11 domains/items followed by semi-structured interviews with patient groups conducted to rate these items using a Likert response scale (supplementary material). The top five items were selected for inclusion, which included: cough, shortness of breath, fatigue, mood and overall well-being. Five items were chosen based on expert opinion to balance an inclusive assessment with the ability to quickly and concisely assess HRQoL in a similar way to other validated and widely used tools [3]. When completing the tool, the patient was asked to select one point for each item along a 13 cm NRS with a range from 0 to 10 with 0.5-point increments. The item scores range from 0 to 10 and total scores from 0 to 50 with lower numbers indicating better HRQoL. The patients had utilised a 13 cm NRS that ranged from 0 to 10 with 0.5-point increments, and we had originally named the tool as RQ-LIFE (Raghu Quality-LIFE) at the time. On further thought, we named the same tool as R-Scale-PF for appropriateness. The example of the R-Scale-PF tool along a 10 cm NRS and the original RQ-LIFE NRS tool answered by patients are available in the supplementary material. The recall period of 2 weeks was established based on expert opinion and is consistent with several other tools previously developed in this field [4, 5]. After completing the study, participants were asked to rate the understandability, accuracy and ease of use of the tool for assessing HRQoL. The tool development process is outlined in figure 1.

Subjects

Patients with an ascertained diagnosis of IPF by multidisciplinary discussion, consistent with the 2018 American Thoracic Society/European Respiratory Society/Japanese Respiratory Society/Latin American Thoracic Association guidelines [6], were consecutively recruited from the ILD clinic at the University of Washington (24 September 2019 to 27 November 2019). Only patients who were clinically stable (as determined by the treating clinician) were included. Participants independently completed three tools in

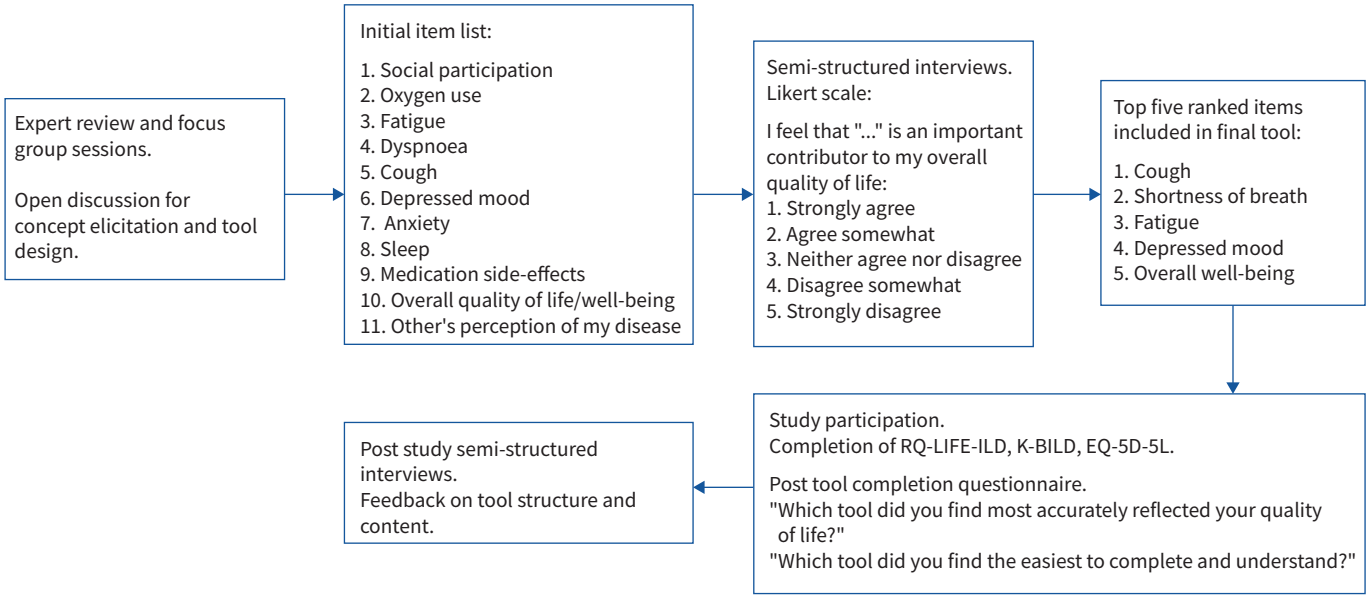

FIGURE 1 Flowchart of the tool development process including concept elicitation, item generation and ranking, tool testing, and post-study feedback. RQ-LIFE-ILD: Raghu Quality of Life in Interstitial Lung Disease; K-BILD: King's Brief Interstitial Lung Disease; EQ-5D-5L: EuroQol Five-Dimensional Five-Level questionnaires.

paper format by random order at the end of the routine, scheduled clinic visit which included the R-Scale-PF, the King's Brief Interstitial Lung Disease questionnaire (K-BILD) and the EuroQol Five-Dimensional Five-Level questionnaire (EQ-5D-5L). Pre-specified clinical data were abstracted from the participant's medical records including clinical and demographic characteristics, antifibrotic use, comorbid conditions, pulmonary function and simple pulmonary exercise testing, *i.e.* – the 6-min walk test (6MWT). Participants also completed the tools at their next routine in-person follow-up visit subject to their clinical needs amid changes in clinical practice as a consequence of the coronavirus disease 2019 (COVID-19) pandemic.

Other HRQoL tools

K-BILD is a 15-item health status questionnaire completed by patients with a seven-point Likert scale. Three major domains included in the questionnaire are: psychological impact, breathlessness and activity, and chest symptoms. Total and domain scores are calculated using a pre-defined algorithm that has been previously validated [5]. A recent update with logit-transformed raw scores provides increased sensitivity for the mild and severe ends of HRQoL scores [7]. The domain and total scores range from 0 to 100 with higher numbers indicating better HRQoL. K-BILD was developed and validated in a population of patients with a range of ILD diagnoses and takes several minutes to complete.

The EQ-5D-5L is a general, non-disease-specific questionnaire that assesses five dimensions including mobility, self-care, usual activities, pain/discomfort and anxiety/depression. Each domain has five possible responses (levels) that are combined to form a sum utility score which is then converted into a specific index value using a country-specific set (in our case USA in English) [8]. The index-based score is interpreted along a continuum where 1 represents best possible health and 0 represents dead, with some health states being worse than dead (<0). Those completing the questionnaire also rate their overall health on a 20 cm visual analogue scale (VAS) with a score of 0 representing "the worse health you can imagine" and 100 representing "the best health you can imagine".

Validity

All patients recruited completed the questionnaires in full. We assessed the floor and ceiling effects of the R-Scale-PF, which has been previously established as >15% of participants rating the maximum or minimum possible scores [9]. Internal consistency was evaluated with Cronbach's coefficient alpha (α) where an α coefficient $\geqslant$0.7 is considered acceptable [10]. Concurrent validity was assessed by using Pearson's correlation between the R-Scale-PF to K-BILD, the EQ-5D-5L, pulmonary function testing and the 6MWT. We also evaluated the discriminatory ability of our tool or the known-groups validity by stratifying participants into groups based on lung function, oxygen use and the gender, age, physiology (GAP) index [11]. The GAP index and staging system was developed as a means to predict mortality using a combination of clinical and physiologic variables to identify three stages with associated increasing 1-year mortality.

Mean and standard deviation were used to describe parametric data. The independent two-sample t-test was used for analysis of normally distributed data and the Wilcoxon–Mann–Whitney test for non-normally distributed data. Effect size is reported as Cohen's *d*. The correlation between the tool scores and continuous variables was measured using the Pearson correlation coefficient (*r*). We considered correlations <0.3 to be weak, those $\geqslant$0.3 and <0.7 to be moderate, and those $\geqslant$0.7 to be strong [12]. GraphPad Prism 9 for macOS and SPSS 27 software were used for statistical analysis.

Ethical approval

This study was reviewed and approved by the Institutional Review Board at the University of Washington (Study ID 8283). Written informed consent was obtained from all participants.

Results

Between September and November 2019 (preceding the COVID-19 pandemic), the novel tool was completed by 100 consecutive patients with an ascertained diagnosis of IPF who were seen in the clinic for routine clinical care. 92 patients were established patients followed for continuity care at the centre for ILD; eight out of 100 recruited for this study were newly referred for further management of IPF. The demographics of the study participants are summarised in table 1. The mean±SD age was 71.2±8.2 years with the majority being male (67%) and Caucasian (91%). Mean values for lung function were forced vital capacity (FVC) % predicted (70.5±18.7%) and diffusing capacity of the lung for carbon monoxide (D_{LCO}) % predicted (51.7±18.9%). Patients with mild to moderate impairment in FVC and D_{LCO} were more represented based on GAP index stages I (38%), II (44%) and III (18%). Due to the unforeseen and abrupt onset of the COVID-19 pandemic in March 2020, the intended regular in-person follow-up visits were all

TABLE 1 Participant demographics

	All patients
Total subjects n	100
Male:female n	67:33
Age, years	71.2±8.2
Ethnicity, %	
Caucasian	91
Other	9
Ever smoker, %	53
Family history of ILD, %	21
Time since diagnosis, years	2.8±3.1
Surgical lung biopsy, %	43
Supplemental oxygen use, %	41
Pulmonary hypertension, %	14
Current antifibrotic use, %	59
FVC, % predicted	70.5±18.7
D_{LCO}, % predicted	51.7±18.9
6-min walk distance, feet	1194±420
GAP stage, %	
I	38
II	44
III	18
EQ-5D-5L index score	0.79±0.15
K-BILD total score	58.6±10.2
R-Scale-PF total score	15.4±9.7

Data are presented as mean±SD, unless otherwise stated. ILD: interstitial lung disease; FVC: forced vital capacity; D_{LCO}: diffusing capacity of the lung for carbon monoxide; GAP: gender, age, physiology; EQ-5D-5L: EuroQol Five-Dimensional Five-Level questionnaire; K-BILD: King's Brief Interstitial Lung Disease questionnaire; R-Scale-PF: Raghu scale for pulmonary fibrosis.

cancelled due to public health restrictions. Telehealth visits were increasingly utilised, which did not allow for assessment of lung function or the 6MWT. Follow-up HRQoL tool responses were, however, obtained from a subset of patients who at varying intervals presented for in-person assessment.

Internal consistency

Cronbach's α was high for the total score (0.825) indicating a good internal consistency. Floor and ceiling effects are summarised in table 2. Aside from the "depressed mood" item, which had a floor effect of 36%, the remaining items had ⩽15% of the participants selecting the highest or lowest scores. When testing the impact of item removal on internal consistency, exclusion of each item did not result in any significant increase in the α coefficient and all five items (including "depressed mood") were included in the final version of the tool (supplementary material).

Concurrent validity

The R-Scale-PF total score had a moderate to strong negative correlation with previously established HRQoL tools including the K-BILD total score (r=−0.71, p<0.01), the EQ-5D-5L index score (r=−0.67, p<0.01) and the VAS (r=−0.64, p<0.01). This suggests good agreement between the R-Scale-PF and other tools, as lower scores indicate better HRQoL (table 3). There was a moderate negative correlation between FVC % predicted (r=−0.31, p<0.01) and 6MWT (r=−0.38, p<0.01) but a weak correlation with D_{LCO} % predicted (r=−0.27, p<0.01).

Known-groups validity

The R-Scale-PF total score was evaluated across different measures of disease severity (table 4 and figure 2). Total scores were higher in those patients with more severe impairment in FVC % predicted and D_{LCO} % predicted. When examining exertional tolerance, total scores were higher in patients with a 6-min walking distance in the lower tertile compared to the upper tertile; R-Scale-PF totals scores were significantly higher in patients who were receiving supplemental oxygen and those with increasing disease severity based on the GAP index.

TABLE 2 Floor and ceiling effects

Item	Floor/ceiling %
Cough	9/1
Shortness of breath	12/1
Fatigue	10/0
Depressed mood	36/0
Well-being	15/0
Total score	1/0

Follow-up measurements

All study participants were eligible to repeat completion of the three HRQoL tools at their next scheduled follow-up visit. 53 out of 100 participants completed the tools at varying timepoints beyond 5–6 months after initial participation. The baseline measurements from patients were obtained from September 2019 to November 2020. Unfortunately, due to the significant limitations and restrictions related to the COVID-19 pandemic, the intended follow-up lung function and walk testing was only possible for a very small number of patients who only returned for in-person follow-up visits based on clinical needs and these were at varying intervals and different timepoints beyond the timeframe of 2–3 weeks of completing the HRQoL tools. Thus, the data was insufficient to allow for meaningful analysis and correlation with the HRQoL tools administered. Data collected from the 53 patients revealed that the mean±SD total scores had not significantly changed from the initial study visit: R-Scale-PF 17.3±9.1, K-BILD 57.9±12.5 and EQ-5D-5L index score 0.81±0.2. The R-Scale-PF total score demonstrated strong correlation with the K-BILD total score (r=−0.77, p<0.01), but weak to moderate correlation with the EQ-5D-5L index score (r=−0.32, p=0.049) and the VAS (r=−0.27, p=0.017). This suggests consistent concurrent validity across repeated measurements, particularly with K-BILD (table 5).

Discussion

In this study we have developed and evaluated a novel, rapid tool for the evaluation of HRQoL in patients with IPF and compared it to two validated tools used in the context of research studies. There have been multiple tools evaluated for the assessment of HRQoL in IPF, including disease-specific tools, general tools and tools developed for other respiratory diseases (table 6) [3, 5, 13–23]. Several tools have been developed for the assessment and analysis of HRQoL and have demonstrated utility in the context of clinical studies and trials for IPF but are not used in routine clinical practice to date. Over the last few years, there has been increasing awareness of the impact of HRQoL in patients with IPF and other fibrotic ILDs, particularly those of the progressive fibrotic form [24]. HRQoL is a generally accepted endpoint, but, to date, treatment interventions have regrettably shown no improvement in reported outcomes by patients with IPF. Most recent trials have used a combination of general instruments (*i.e.* not

TABLE 3 Correlation between impaired health-related quality of life tools and lung function

	R-Scale-PF total	K-BILD total	K-BILD breath	K-BILD psych	K-BILD chest	EQ-5D-5L VAS	EQ-5D-5L index score	6MWT distance	D_{LCO} % pred	FVC % pred
R-Scale-PF total	1									
K-BILD total	−0.713	1								
K-BILD breath	−0.674	0.888	1							
K-BILD psych	−0.553	0.877	0.619	1						
K-BILD chest	−0.674	0.733	0.683	0.545	1					
EQ-5D-5L VAS	−0.642	0.646	0.64	0.527	0.529	1				
EQ-5D-5L index score	−0.665	0.624	0.64	0.429	0.621	0.634	1			
6MWT distance	−0.383	0.528	0.599	0.358	0.457	0.459	0.56	1		
D_{LCO} % pred	−0.274	0.471	0.5	0.373	0.328	0.386	0.314	0.674	1	
FVC % pred	−0.307	0.421	0.421	0.342	0.347	0.277	0.213*	0.369	0.495	1

R-Scale-PF: Raghu scale for pulmonary fibrosis; K-BILD: King's Brief Interstitial Lung Disease questionnaire; EQ-5D-5L: EuroQol Five-Dimensional Five-Level questionnaire; VAS: visual analogue scale; 6MWT: 6-min walk test; D_{LCO}: diffusing capacity of the lung for carbon monoxide; FVC: forced vital capacity. Correlation shown as Pearson's coefficients (r); all p<0.01, unless indicated. *: p<0.05.

TABLE 4 Known groups validity analysis

Variable	Patients n	Mean±SD R-Scale-PF total score	Mean difference (95% CI)	p-value	ES
FVC					
>75% predicted	40	12.7±8.9	6.8 (0.02–13.5)	0.05	0.78
<45% predicted	8	19.4±7.5			
D_{LCO}					
>60% predicted	32	14.2±10.5	5.3 (0.1–10.5)	0.046	0.51
<40% predicted	31	19.4±10.1			
GAP index					
Stage I#	46	12.9±9.5	8.8# (3.7–13.8)	0.001	0.96
Stage II	36	15.3±9.4			
Stage III#	18	21.6±7.9			
Supplemental oxygen therapy					
No supplemental oxygen	57	13.1±9.2	5.2 (1.5–9.0)	0.007	0.56
On supplemental oxygen	43	9.6±9.6			
6MWT					
Lower tertile	32	19.4±9.6	7.9 (3.4–12.8)	0.001	0.87
Mid tertile	33	15.1±9.7			
Upper tertile	33	11.6±8.5			
Pulmonary hypertension					
Absent	86	15.1±9.6	2.26 (–7.8–3.3)	0.419	0.23
Present	14	17.3±10.0			

R-Scale-PF: Raghu scale for pulmonary fibrosis; ES: effect size (Cohen's *d*); FVC: forced vital capacity; D_{LCO}: diffusing capacity of the lung for carbon monoxide; GAP: gender, age, physiology; 6MWT: 6-min walk test. #: mean difference between GAP stage I and stage III.

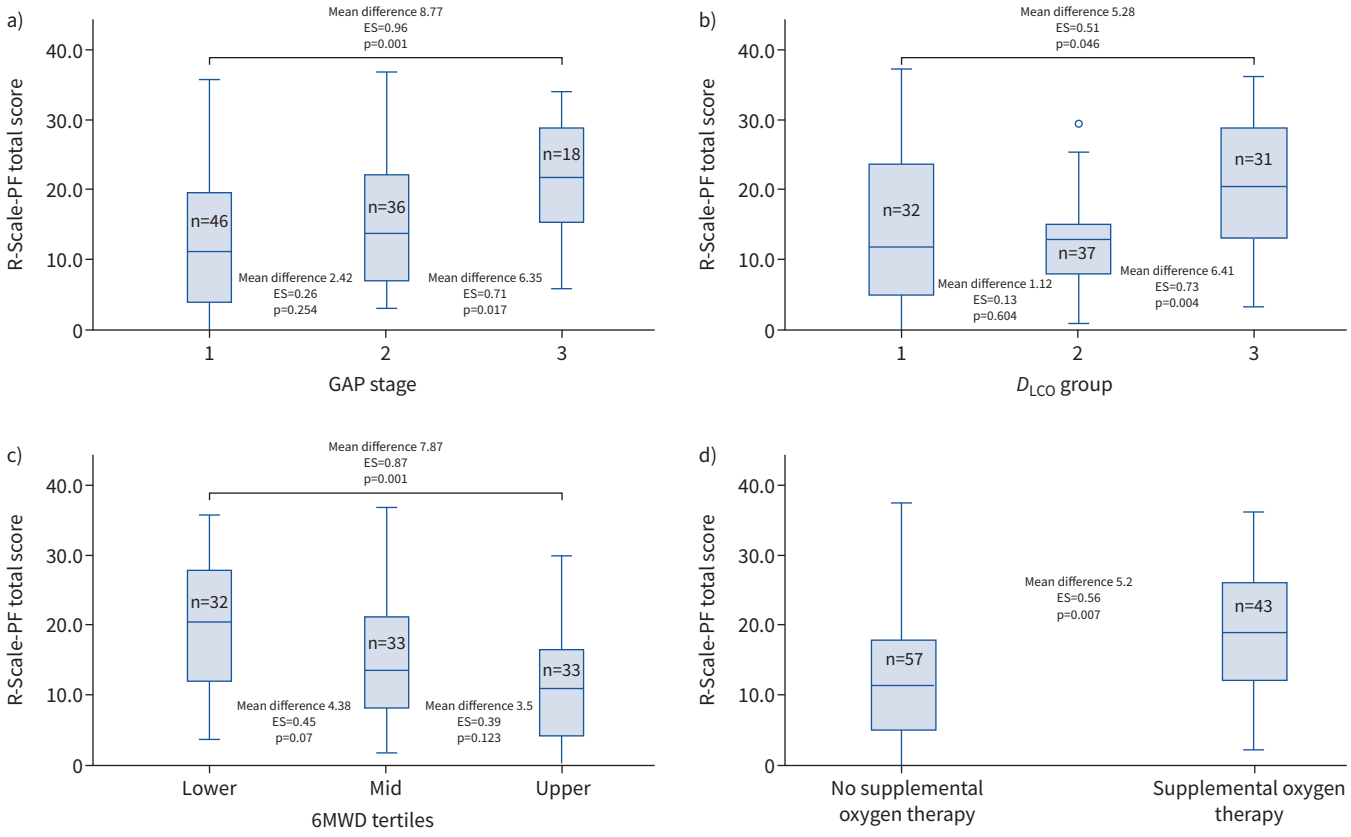

FIGURE 2 Raghu scale for pulmonary fibrosis (R-Scale-PF) total scores compared across a) gender, age, physiology (GAP) index, b) diffusing capacity of the lung for carbon monoxide (D_{LCO}) % predicted (group 1 >60%, group 2 40–60%, group 3 <40%), c) 6-min walk distance (6MWD) tertiles and d) supplemental oxygen therapy. ES: effect size (reported as Cohen's *d*).

TABLE 5 Correlation between health-related quality of life tools for follow-up completion

	R-Scale-PF total	K-BILD total	EQ-5D-5L VAS	EQ-5D-5L index score
R-Scale-RF total	1			
K-BILD total	−0.772 (p<0.01)	1		
EQ-5D-5L VAS	−0.272 (p=0.049)	0.131 (NS)	1	
EQ-5D-5L index score	−0.323 (p=0.017)	0.283 (p=0.040)	0.618 (p<0.01)	1

Follow-up health-related quality of life tool completion for 53 patients. Tool completion was at least 5 months after initial enrolment for all participants. R-Scale-PF: Raghu scale for pulmonary fibrosis; K-BILD: King's Brief Interstitial Lung Disease questionnaire; EQ-5D-5L: EuroQol Five-Dimensional Five-Level questionnaire; VAS: visual analogue scale; NS: nonsignificant. Correlation shown as Pearson's coefficients (*r*).

disease-specific) such as EQ-5D-5L, the Short-Form 36 Health Status questionnaire and St George's Respiratory Questionnaire (SGRQ), with more disease-specific instruments, such as K-BILD, A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis (ATAQ-IPF), and the Living with Idiopathic Pulmonary Fibrosis questionnaire, increasing in use [15].

The R-Scale-PF tool has good internal consistency, minimal floor or ceiling effects, strong concurrent validity and the ability to discriminate between degrees of disease severity across different measures. Our NRS tool has minimal text that requires focused reading to understand the five items and it is thus very easy for participants to see and circle the severity of their subjective perception. Our results are encouraging for consideration of this tool in future clinical trials and patients endorsed it as being very simple and well representative of their overall QoL. The "depressed mood" item of the R-Scale-PF tool demonstrated a significant floor effect with 36% of participants choosing 0 or "none" on the rating scale, suggesting no impact of mood on their overall HRQoL. Depression has been shown to be highly prevalent in patients with IPF, ranging from 19% to 49% across different studies and depression assessment tools [25–28]. Depression alone has been demonstrated to have a significant impact on overall HRQoL and

TABLE 6 Summary of health-related quality of life tools previously used in clinical studies in patients with idiopathic pulmonary fibrosis (IPF)

Name of tool	Domains assessed	Number of items
IPF-specific		
King's Brief Interstitial Lung Disease questionnaire (K-BILD) [5]	Breathlessness and activities, chest symptoms and psychological impact	15
IPF-specific version of St George's Respiratory Questionnaire (SGRQ-I) [13]	Symptoms, activity and psychosocial impact	34
A Tool to Assess Quality of Life in Idiopathic Pulmonary Fibrosis (ATAQ-IPF) [14]	Cough, shortness of breath, planning, sleep, mortality, energy, mental health, spirituality, social activities, finances, independence, sexuality, relationships and treatments	41–86[#]
Living with Idiopathic Pulmonary Fibrosis Questionnaire (L-IPF) [15]	Dyspnoea, cough and energy	44
Respiratory-specific		
St George's Respiratory Questionnaire (SGRQ) [16]	Symptoms, activity and psychosocial impact	50
University of California San Diego Shortness of Breath Questionnaire (UCSD-SOBQ) [17]	Shortness of breath	24
Cough and Sputum Assessment Questionnaire (CASA-Q) [18]	Cough	20
COPD Assessment Test (CAT) [19]	Cough, sputum, dyspnoea and chest tightness	8
Medical Research Council Scale (MRC) [20]	Dyspnoea	5
Modified Medical Research Council Scale (mMRC) [21]	Dyspnoea	5
General		
EuroQol Five-Dimensional Five-Level questionnaire (EQ-5D-5L) [3]	Mobility, self-care, usual activities, pain/discomfort and anxiety/depression	5
World Health Organization-Five Well-Being Index (WHO-5) [22]	Psychological well-being	5
Short-Form 36 Health Status Questionnaire (SF-36) [23]	Physical functioning, physical role, emotional role, bodily pain, general health, vitality, social functioning and mental health	36

[#]: varies for different versions of the tool.

previous work has suggested specific treatments for depression could improve other symptoms related to IPF in addition to overall HRQoL [29]. The significant floor effect observed in this study is consistent with the prevalence of depression reported in prior studies. This tool was not designed with the intent of being a comprehensive assessment of depression but instead to serve as a screening tool for those patients requiring further evaluation. Future validation studies of the R-Scale-PF should include concurrent administration of validated depression questionnaires to determine a threshold or significant change in the R-Scale-PF "depressed mood" score that would prompt additional investigation.

We chose to evaluate concurrent validity by comparing our novel tool against an established and validated questionnaire in patients with IPF and a general HRQoL tool that has been evaluated for a range of chronic diseases including IPF. K-BILD has now been applied across multiple studies and has been demonstrated to be a suitable measure of HRQoL in multiple countries [5, 30]. The EQ-5D-5L assesses generic HRQoL across disease states encompassing not only the burden of disease symptoms but also the effects of disease-specific treatments and comorbidities. The EQ-5D-5L has been used in many large clinical trials evaluating interventions in patients with IPF. Our study population had a slightly better mean EQ-5D-5L index score compared to these studies 0.79 *versus* 0.67–0.76 (INSIGHTS-IPF, BUILD-1, STEP-IPF and INPULSIS). We elicited direct feedback from study participants after completing the HRQoL tools to assess both the ease of use and the accuracy in assessing HRQoL (supplementary material). While both K-BILD and the R-Scale-PF were similarly ranked in ease of understanding and completion, followed by the EQ-5D-5L, K-BILD was rated by the most participants as being the most accurate reflection of their QoL followed by the R-Scale-PF and then the EQ-5D-5L.

When examining the correlation of the R-Scale-PF total score with measures of lung function, only a weak to moderate correlation for D_{LCO}, FVC and 6-min walking distance was appreciated. A moderate correlation between tool scores and lung function has also been observed in other ILD questionnaires, including K-BILD, ATAQ-IPF and the SGRQ-I (IPF specific version) [5, 14, 31]. This effect has also been observed in COPD [32]. This highlights the complex nature of the perception of physical and psychological symptom burden and suggests that HRQoL tools assess information that is distinct from physiologic measurements of disease severity. The ability to discriminate between groups of disease severity is an important feature of any tool and the R-Scale-PF demonstrates this ability across several measures including 6-min walking distance, oxygen use and the GAP index (table 4 and figure 2). When comparing those patients with GAP stage I and III scores, the R-Scale-PF demonstrated significantly higher scores in patients with more severe disease corresponding to a perception of worse HRQoL. While the impairment in HRQoL observed with increasing disease severity (based on physiologic markers) may be driven by more severe physical or emotional impairment, it is imperative to evaluate the variation in individual patients' perception of symptoms and disease burden. As one participant in our focus group highlighted "cough is not a big issue for me, but I find that my fatigue really limits my ability to spend time with friends and family…".

While our study is prospective in a relatively large number of patients with IPF, in the context of assessing HRQoL tools in patients with IPF, several limitations warrant further investigation. First, we unfortunately could not evaluate test–retest reliability in all 100 patients because of the inevitable consequence of the COVID-19 pandemic that disallowed follow-up in-person clinic visits, lung function assessments and 6MWTs. Secondly, this study was completed in a single tertiary referral centre with 91% of participants of Caucasian ethnicity and, thus, the generalisability of this tool should be further validated in patients from other centres. Thirdly, the tool was developed by one long-standing, experienced clinical expert in ILD in combination with patients recruited for focus groups and semi-structured interviews. While we completed post-study interviews with non-study participants, additional cognitive interviews to elicit further feedback and refinement of the tool will need to be undertaken prior to validation studies. Acknowledging that the R-Scale-PF tool will need to be validated in a separate cohort of patients with IPF at another centre, which is not feasible until the COVID-19 pandemic slows down in a consistent manner, we have elected to share the tool development process and pilot testing results to provoke its immediate utilisation in other clinical studies from other centres and/or multi-centre studies, as has been done with other IPF-specific HRQoL tools [5, 14, 15]. Despite the changes to clinical practice and in-person contact during the ongoing pandemic, we are pleased to have the validated R-Scale-PF tool against the previously validated tools at baseline and to obtain follow-up data in a majority of the initial participants. Fourthly, repeatability is a critical component of any QoL tool and it was not assessed in this study. This analysis will be included in future validation studies. Finally, the utility of the R-Scale-PF in other ILD diagnoses needs to be evaluated as there may be disease-specific factors that contribute to HRQoL. In connective tissue disease-associated ILD, for example, rheumatologic disease manifestations contribute to symptoms and disease burden independent of any pulmonary manifestations.

Conclusion

In conclusion, this pilot, prospective study in a relatively large number of patients with IPF suggests that the R-Scale-PF is a valid measure of IPF-related symptoms and their impact on HRQoL. It demonstrates good concurrent and known-groups validity and has good correlation with both disease-specific and general tools. The R-Scale-PF provides an opportunity to assess HRQoL rapidly and objectively in patients with IPF. The limited number of essential items and simple numerical scale format as compared to existing tools allows for incorporation into both routine clinical practice and research settings. While not designed as a comprehensive assessment of HRQoL, the R-Scale-PF can highlight specific items affecting the individual patient which can prompt further exploration by the treating clinician. Due to limited in-person visits caused by the COVID-19 pandemic, telemedicine and remote monitoring have become increasingly adopted for both clinical and research purposes [33, 34]. The R-Scale-PF provides a rapid, easy to understand method of assessing HRQoL and could be easily applied to remote-monitoring programmes, which could include home spirometry measurements. Additional research is needed to assess the reproducibility, responsiveness, validity and test–retest reliability of the tool, thresholds prompting formal depression screening, and to explore its application in other ILD diagnoses.

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