Sharp, C., McCabe, M., Hussain, M. J., Dodd, J. W., Lamb, H., Adamali, H., ... Smith, D. (2017). Duration of benefit following completion of pulmonary rehabilitation in interstitial lung disease: an observational study. *QJM*, *110*(1), 17-22. [hcw105]. https://doi.org/10.1093/qjmed/hcw105

QJM Advance Access published July 10, 2016

Title: Duration of benefit following completion of Pulmonary Rehabilitation in Interstitial Lung Disease – an observational study

Short title: Pulmonary Rehabilitation in ILD

Authors: C Sharp[1,2], M McCabe[3], M J Hussain[1], J W Dodd[2], H Lamb[1], H Adamali[1], AB Millar[2], D Smith[1]

1 – North Bristol Lung Centre, Southmead Hospital, Bristol BS10 5NB, UK

2 - Academic Respiratory Group, University of Bristol, Bristol BS10 5NB, UK

3 – Department of Geography, London School of Economics, Houghton Street, London, WC2A 2AE, UK

Corresponding author – ann.millar@bristol.ac.uk, 01179505050

Word count - 2496

Key words – Pulmonary rehabilitation, interstitial lung disease, idiopathic pulmonary fibrosis

Abstract

Background – It remains unclear for how long the benefits of pulmonary rehabilitation (PR) last in interstitial lung disease (ILD). An increasing number of ILD patients complete PR and it is vital they be offered the most beneficial approaches.

Methods – This is a retrospective, observational study of a cohort with ILD who had completed PR. Incremental shuttle walk (ISWT) and chronic respiratory disease questionnaire (CRDQ) were compared before PR, at course completion, and 6/12 months follow-up. Focus group discussions with ILD participants who had completed PR and their carers established qualitative views on existing and potential future PR provision.

Results – 79 participants with ILD were identified at course completion, with 39 followed to 12 months. 11 participants died during follow-up. Initial benefits from PR were not sustained at 6 months (ISWT change 0.0m (95% CI -23.2 to 23.2m), CRDQ change 2.5 (95% CI -2.4 to 7.4)) and 12 months (ISWT change -0.7m (95% CI -37.3 to 35.9m), CRDQ change 4.0 (95% CI -2.2 to 10.2)). Continued home exercise gave longer lasting benefit in exercise capacity. Focus group discussions highlighted the value attached to PR and suggested areas for improvement.

Conclusions – Standard PR gives initial benefits in participants with ILD who complete the course, however these are not sustained. Tailored approaches to this group would be appreciated by this group and should be explored.

Duration of benefit following completion of Pulmonary Rehabilitation in Interstitial Lung Disease – an observational study

Introduction

Disabling breathlessness frequently complicates Interstitial Lung Diseases (ILDs). Patients report significantly limited activities of daily living, along with low levels of physical functioning and high levels of fatigue and dyspnoea[1]. These limitations contribute to significantly reduced quality of life (QoL)[2]. While pharmacological treatments are now available for IPF[3, 4] and other ILDs[5, 6], their impact on QoL is unclear. International guidelines[7, 8] advocate referral for pulmonary rehabilitation (PR), while the UK NICE quality standard for IPF highlights its importance[9]. PR is well established in Chronic Obstructive Pulmonary Disease (COPD)[10] but evidence of benefit in ILD patient populations is of low quality[11, 12]. Limited data exists concerning longer term benefit from PR in this group. This is especially important given the greater likelihood of progressive deterioration in participants with ILD.

We report a retrospective, observational study of a well characterised population of consecutive patients with ILD who completed a PR course. To capture patient/carer experience and attitudes towards PR in ILD, we conducted focus group discussions with those who had completed PR.

The aims of this study were:

1) Determine if there is a sustained benefit from PR in ILD.
2) Identify patient characteristics associated with response to PR in ILD.
3) Undertake a qualitative assessment of PR experience in ILD.

We considered that this would help to inform future ILD-tailored PR programmes.

Methods

Study subjects

The Lung Education and Exercise Programme (LEEP) at North Bristol NHS Trust has over 20 years' experience providing a standard PR course to a heterogeneous population. Increasingly, referrals come from Bristol Interstitial Lung Disease (BILD) service; 10-15% of those completing PR have ILD.

We retrospectively identified consecutive participants with a diagnosis of ILD made by their referring physician, who completed PR from the LEEP database. Outcomes were recorded at course completion, and at follow-up visits after 6 and 12 months.

Study ethical approval was granted by NRES Committee East of England - Cambridge South (REC reference 15/EE/0023).

Pulmonary Rehabilitation programme

LEEP is a PR programme, for all respiratory diagnoses, conforming to standard BTS recommendations[11]. The six week course includes twice-weekly sessions, with an educational component and supervised, monitored exercise and a third home exercise session, all tailored to participants according to their needs and disease severity. Exercise prescriptions for each participant are based on their medical history and exercise capacity, including upper and lower extremity training and incorporating both strength and endurance elements.

Measures

Demographics

Baseline variables included diagnosis, age, gender, co-morbidities, smoking history and pulmonary function tests.

Exercise capacity

An Endurance Shuttle Walk Test (ESWT) and an Incremental Shuttle Walk Distance (ISWT) were performed at pre-course assessment according to published American Thoracic Society/European Respiratory Society field walking test criteria[13]. At each follow-up visit, ISWT was repeated, as per standard practice for the LEEP course. ISWT has been used in previous studies and appears responsive to PR in ILD[14, 15]. Participants used oxygen or walking aids if required.

Health Status, Mood & Dyspnoea

Change was assessed in the Chronic Respiratory Disease Questionnaire (CRDQ), a 20-item questionnaire covering domains of dyspnoea, mastery, emotion and fatigue, which has been a standard measure in studies of PR in COPD[16] and ILD[15]. Higher scores represent more severe disease. The minimal clinically important difference (MCID) of 10 points was used as previously described[17]. The Hospital Anxiety and Depression Score (HADS) was used to assess anxiety and depression[18]; this is a 14-item questionnaire with higher scores representing more severe symptoms. Breathlessness was measured using Medical Research Council (MRC) dyspnoea scores.

Patient focus groups

A thematic content analysis approach was taken for this study, using semi-structured focus group discussions conducted with a convenience sample of subjects, identified and

approached at a bi-monthly patient support group. Group discussions lasted one hour and were moderated by a male physician (CS) and a female ILD specialist nurse (HL) and recorded by detailed field notes. Both moderators were known to the group participants. Topics focused on patient and carer experiences of PR and areas which may improve these experiences. Questions and discussion points used are shown in Supplemental table S1. The two moderators (CS and HL) independently agreed that data saturation had been achieved.

In qualitative analysis, field notes were reviewed for an overall impression. Candidate themes were identified and grouped. Descriptions were prepared based on these themes and presented as results. Results were discussed between the study authors.

Statistical analysis

Data are described using mean with standard deviation (SD) or mean difference with 95% confidence intervals (CI) unless otherwise indicated. Continuous variables were compared using t-tests. Categorical variables were compared using χ^2 tests. To assess the impact of PR on course completion and at follow-up visits (6 and 12 months), each visit was compared to the pre-course values. Sensitivity analyses following multiple imputation of missing cases were performed to assess the impact of loss to follow-up over the study period. Multiple imputation was not used in the remainder of the analyses.

Multiple linear regression models were constructed to ascertain predictive elements from amongst the baseline characteristics. Collinearity was assessed and ESWT, pulmonary function and baseline HADS were excluded from the model.

Data were analysed using SPSS software (v23.0.0; IBM Corp.; Armonk, NY, USA).

Results

Study patients

We identified 79 consecutive patients with ILD who completed PR from the LEEP database. Demographics are shown in Table 1, diagnoses in Table 2. The most common diagnosis was IPF (n=28, 35.4%). Patients had moderate physiological and functional impairment, with mean baseline Forced Vital Capacity (FVC) 75.6% predicted (SD 22.6%) and Diffusing Capacity for Carbon Monoxide (DL_{CO}) 47.2% predicted (SD 18.8%). 25.9% of participants had a decline in FVC of >10% in the year prior to course participation. There were no differences for FVC or FEV1/FVC ratio between ILD diagnostic groups (Supplemental table S3). ILD participants with potentially obstructive physiology did not significantly differ from others in this cohort. Baseline ISWT was 177.4m (SD 110.9m) and patients had poor QoL with mean CRDQ, 78.3 (SD 18.4). Other baseline measurements are shown in Table 1.

Completion of follow-up is shown in Figure 1; data were available for 52 participants at 6 months following course completion and 39 participants at 12 months. 11 participants died during 12 months follow-up. Sensitivity analyses suggested no significant impact on overall findings with loss to follow-up (Supplemental table S2). Amongst those lost, there was no significant difference in baseline ISWT compared to those completing 12 months' follow-up (difference 42.8m (95% CI -95.29m to +9.72m), p=0.11).

Short term impact of PR

PR was effective in ILD for those who completed the course, improving both ISWT and CRDQ (Figure 2). ISWT improved by a mean of 29.5m (95% CI 13.7 to 45.2m). CRDQ improved by 11.6 (95% CI 8.5 to 14.7). 38 participants (48.7%) achieved the MCID of a 10 point improvement. Pre-course HADS values indicated no clinical depression or anxiety and did not change after PR. Effects were similar between the subgroups of patients with IPF and non-IPF ILDs (Supplemental table S3). There was no difference in response to PR between participants with and without progressive FVC decline (>10%) before the course (Difference in ISWT change 38.2m, p=0.101, difference in CRDQ change 1.95, p=0.647).

Long term impact of PR

Benefits of PR were not sustained at 6 months or 12 months for participants with ILD following course completion (Figure 2). There was no difference between subgroups of participants with IPF and non-IPF diagnoses in mean change in ISWT from baseline at 6 or 12 months (0.0m (95% CI -23.2 to 23.2m) and -0.7m (95% CI -37.3 to 35.9m) respectively). Analyses were repeated excluding those lost to follow-up; the values did not significantly change. There was no difference in ISWT change at 6 months between participants with and without progressive FVC decline before the course (Difference in ISWT change 4.5m, p=0.882).

Initial improvements in CRDQ were not sustained, with mean change from baseline at 6 months of 2.5 (95% CI -2.4 to 7.4) and at 12 months of 4.0 (95% CI -2.2 to 10.2). At 6 and 12 months, 31.2% and 29.6% of participants achieved the MCID. There was no difference in CRDQ change at 6 months between participants with and without progressive FVC decline before the course (Difference in CRDQ change 9.36, p=0.193).

Of those completing 6 months' follow-up, 58.3% reported continuing with their home exercise prescription, while 29.2% continued to attend a weekly supervised exercise session. At 12 months, these numbers reduced to 39.3% and 10.7% respectively. Those reporting having discontinued home exercise were more likely to experience a fall in their ISWT at 6 months (-27.4m vs 18.6m, 95% CI for the difference 0.1m to 91.8m, p=0.049). There were no other differences between those reporting continued home exercise, or those attending regular supervised exercise sessions (Figure 3).

Prediction of response to PR

A multiple linear regression model predicted 17.4% of the variance in ISWT change 6 months following course completion, $F(3,46)=3.026$, $p=0.04$. Co-efficients of variables included in the model are reported in Table 3. Shorter baseline ISWT ($\beta=-0.368$, $p=0.026$), and younger age ($\beta=-0.386$, $p=0.016$) were predictive of more sustained improvement in ISWT at 6 months.

A similar model constructed for improvement in CRDQ at 6-month follow-up, predicted 16.6% of the variance, $F(3,47)=2.929$, $p=0.044$. Co-efficients of variables included in the model are shown in Table 4. A lower pre-course CRDQ was predictive of greater improvement in QoL at 6 months ($\beta=-0.432$, $p=0.008$).

Focus group discussions

Twenty-three patients and 20 carers participated in focus group discussions, with a range of ILD diagnoses and aged between 62 and 83 years (15 male, 8 female). No patients or carers who were approached declined to participate. Analysis of discussions identified several themes.

Experiences of PR

All felt that PR was valuable, with particular importance being attached to social elements of the course and its role in reducing feelings of isolation. Some negative views were expressed; many felt there was insufficient time for exercise, however this was debated within the focus group. Overall the course was felt to be too short. There was also concern over the predominance of COPD, with patients reporting feeling distinct and isolated from the group, along with feelings of stigma due to oxygen use. Many patients expressed anxiety about unsupervised exercise after PR.

Suggestions for development of PR in ILD

Patient and carer suggestions to improve PR for those with ILD included:

- Longer sessions to give more time for exercise
- Longer course duration
- To enable carers to be present for at least some of the course, to benefit from the educational component. Carers were especially keen on this idea
- Ongoing supervised exercise sessions after course completion
- Specific ILD focused lectures and content more tailored to ILD concerns including oxygen use, diet, control of breathing and entitlements to social support and benefits advice.

Discussion

In this analysis of real-world delivery of standard PR for participants with a variety of ILDs following course completion, we observed initial benefits in exercise capacity and health status. Improvements in exercise capacity and health status were not sustained to 6 months. Patients and carers with ILD felt that PR is a valuable component of their disease management, but could be improved.

These observations suggest this group benefits from PR. The regression analyses to identify predictors of benefit offer conflicting results, with greater benefits seen where PR is delivered in more severe disease as assessed by exercise capacity, but in those with less impairment to QoL and at a younger age.

We acknowledge limitations to this study. We followed patients from completion of PR. Therefore these results can only be generalised to ILD patients completing PR. There was also loss of participants to follow-up in this observational study, reflecting real-world conditions. Subject characteristics did not significantly differ between those lost and those completing follow-up. Additionally, sensitivity analyses demonstrated that this did not change our findings. Any bias is likely to be towards over-stating the degree of benefit in this study, as those not completing PR or subsequent follow-up may have more severe disease. As such this adds weight to our conclusion that benefits of PR are not sustained in ILD.

These limitations are balanced by the real-world world relevance and generalisability of a significant population with one of the longest periods of follow up reported. We were able to enrich our analysis using focus group discussions to capture patient experiences of PR. This adds to previous interview-based data highlighting the specific educational needs of those with ILD[19].

While PR is recognised to have a sustained response in participants with COPD[20], the current literature is inconclusive regarding duration of benefit of PR in ILD. A recent meta-analysis of randomised, controlled trials (RCTs) of standard PR programmes in ILD demonstrated short term effectiveness in both walking distance and QoL[12]. Reports conflict on how sustained this benefit is.

Ryerson et al reported benefit sustained above the MCID to 6 months in walking distance, but not for QoL or dyspnoea in a cohort study[21]. A small RCT by Vainshelboim showed no differences between groups at 11 months[22]. In a cohort study, Holland et al indicated that improvements in walking distance and QoL were not sustained[23]. Kozu et al compared patients with IPF with a cohort with COPD, demonstrating only modest short term gains, not sustained on 6-month follow-up[24]. Ochmann et al examined the effect of PR in occupational lung diseases and found that while exercise capacity benefits were sustained on 12-month follow-up for asthma and COPD, benefit was not sustained beyond 3 months in silicosis and asbestosis[25]. The long term efficacy of standard PR in ILD remains unproven.

In keeping with our observations of the benefits of continued exercise, Dalichau has demonstrated preserved benefit in a cohort of asbestosis patients to 18 months where they continued exercise, with loss of benefit where exercise was ceased[26, 27].

Data surrounding predictors of response to PR in ILD are conflicting. Previous work has suggested a reduced response to PR in a more severe, oxygen dependent group, as compared to those with no such requirement[14], while Holland et al observed maximum benefit in those with milder disease[23]. In contrast, Ferreira et al observed reduced response in those with greater baseline exercise capacity[28] and Ryerson et al reported that greatest benefit was seen in those with the most severe impairment[21]. Our data also conflict. It

appears appropriate to offer PR to ILD patients of all but the most severe levels of impairment, which would preclude course completion.

Our heterogeneous cohort of ILD patients did not demonstrate any differential response to PR when separated by diagnosis, however there is some evidence to suggest that patients with IPF are most likely to benefit[29]. An important question for future work will be to identify which groups benefit most.

Tailoring of PR courses to ILD patients' needs is an area which needs further exploration. Strength and endurance are impaired in fibrotic lung diseases[30, 31], however endurance responds well to PR[32]. The need to sustain activity and exercise beyond the end of the course is highlighted by more sustained improvements observed in those continuing home exercise to 6-month follow-up. It is possible, however, that a failure to sustain such activity is a marker of pulmonary decline. Guidelines have advocated the importance of the educational component to patients' self-efficacy[33], however at present course content is not tailored to patients' diseases. This was important to the focus group participants, and also in work conducted by the British Lung Foundation, who found that 28% of IPF patients surveyed found their PR to be of average quality or worse and 98% would appreciate a tailored approach[34].

Summary

Despite initial benefits from PR, overall improvements are not sustained in participants with ILD following course completion. Focus group discussions indicate that a tailored, ILD-specific programme would be valued. This is an area in which no quantitative data have been produced and there is a pressing need for a robust, large scale, randomised trial to test this question.

Acknowledgements

We thank the members of the LEEP team at North Bristol NHS Trust for data collection and entry.

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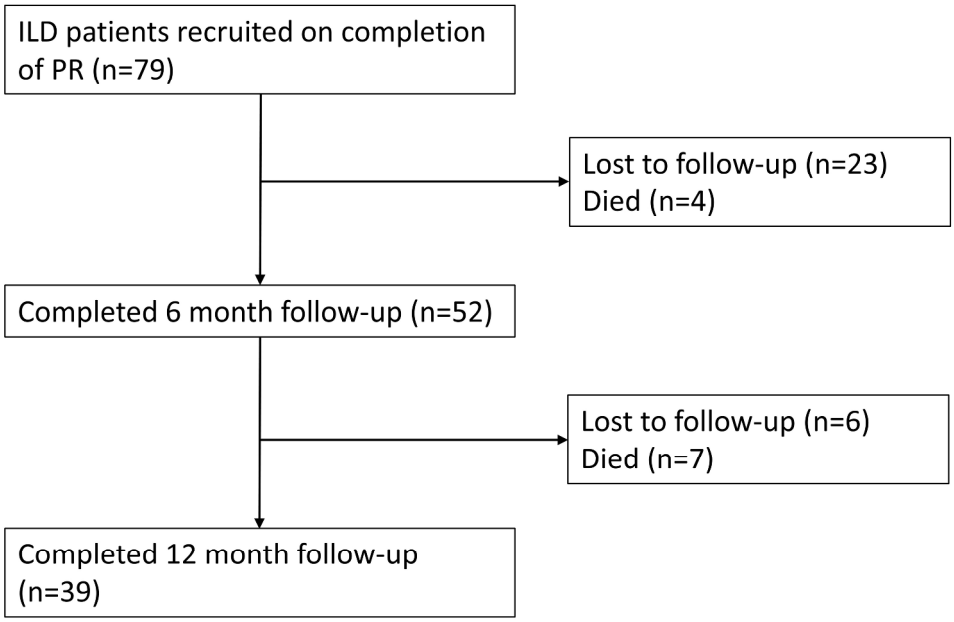
Table 1 – Baseline demographic, physiological and quality of life values, Interstitial Lung Disease participants			
		Mean/Value	SD
Age (years)		68.8	11.9
Male (%)		55.7	
Smoking history (%)	Ex-smoker	60.8	
	Never smoked	39.2	
	Current smoker	0	
Comorbidities (%)	None	38.0	
	1-2	45.1	
	3 or more	16.9	
Death <12 months (%)		13.9	
Follow-up (months)		32.3	25.5
FEV1 % predicted		74.9	21.5
FVC % predicted		75.6	22.6
DL_{CO} % predicted		47.2	18.8
TLC % predicted		69.6	18.8
Incremental Shuttle Walk Distance (m)		177.4	110.9
Endurance Shuttle Walk Distance (s)		329.2	286.4
MRC score		3.3	1.0
CRDQ overall		78.3	18.4
	Mastery	18.4	5.2
	Emotion	31.9	8.6
	Fatigue	13.8	4.5
	Dyspnoea	14.1	5.0
HADS overall		12.6	6.6
	Anxiety	6.4	4.0
	Depression	6.2	3.3
Abbreviations – FVC (Forced Vital Capacity), FEV1 (Forced Expiratory Volume in 1 second), TLC (Total Lung Capacity), DL_{CO} (Diffusing Capacity for Carbon Monoxide)			

Table 2 – Diagnoses of participants		
Diagnosis	n	%
Idiopathic Pulmonary Fibrosis	28	35.4
Asbestosis	10	12.7
Hypersensitivity Pneumonitis	8	10.1
Idiopathic Non-Specific Interstitial Pneumonia	8	10.1
Sarcoid	8	10.1
Combined Pulmonary Fibrosis and Emphysema	5	6.3
Connective Tissue Disease-related ILD	4	5.1
Respiratory Bronchiolitis-ILD	3	3.8
Drug induced ILD	2	2.5
Rheumatoid Arthritis with Usual Interstitial Pneumonia	2	2.5
Lymphangioleiomyomatosis	1	1.3
Total	79	100

Table 3 – Co-efficients of variables in multiple linear regression model for change in ISWT at 6 month follow-up				
Model	Unstandardized Coefficients	Standardized coefficients	t	Sig.
	Beta	Beta		
(Constant)	142.22		1.693	0.098
Age (years)	-2.849	-0.386	-2.518	0.016
Baseline ISWT	-0.266	-0.368	-2.307	0.026
Baseline CRDQ	1.35	0.316	2.011	0.051
Dependent Variable: Change in ISWT at 6 months				
Abbreviations – Incremental Shuttle Walk Test (ISWT), Chronic Respiratory Disease Questionnaire score (CRDQ)				

Table 4 - Co-efficients of variables in multiple linear regression model for change in CRDQ at 6 month follow-up				
Model	Unstandardized Coefficients	Standardized Coefficients	t	Sig.
	Beta	Beta		
(Constant)	29.612		1.646	0.107
Age (years)	-0.001	-0.001	-0.005	0.996
Baseline ISWT	0.023	0.150	0.956	0.344
Baseline CRDQ	-0.397	-0.432	-2.798	0.008
Dependent Variable: Change in CRDQ at 6 months				
Abbreviations – Incremental Shuttle Walk Test (ISWT), Chronic Respiratory Disease Questionnaire score (CRDQ)				



Study flow diagram
Figure 1
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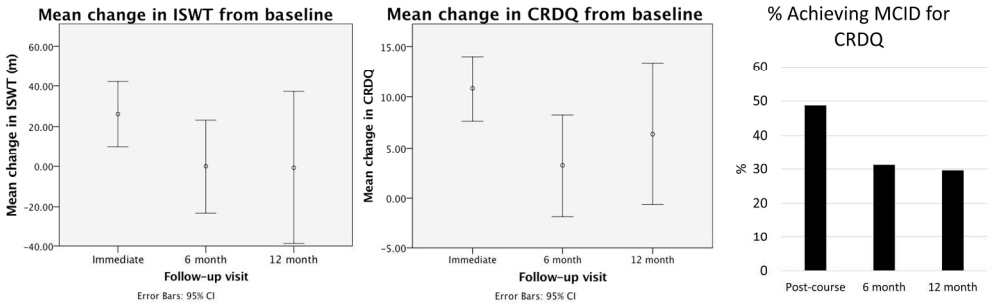
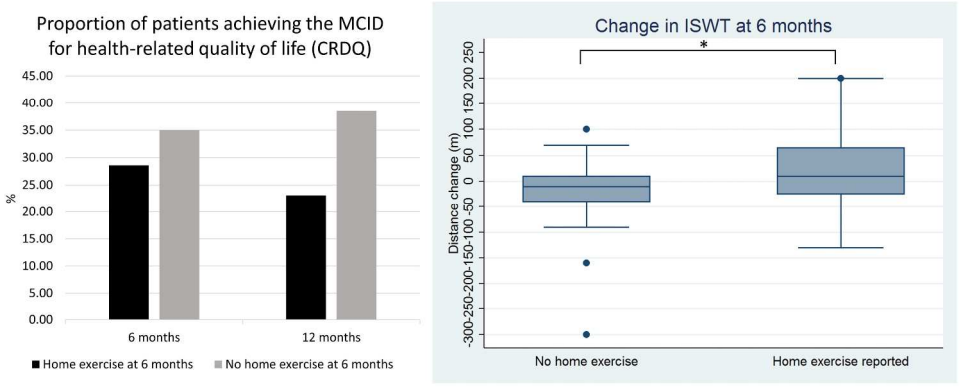


Change in exercise capacity and quality of life on follow-up.
ISWT - Incremental Shuttle Walk Test, CRDQ - Chronic Respiratory Disease Questionnaire, MCID - Minimal Clinically Important Difference
Figure 2
110x36mm (600 x 600 DPI)



Impact of continued home exercise on quality of life and exercise capacity.
ISWT - Incremental Shuttle Walk Test, CRDQ - Chronic Respiratory Disease Questionnaire, MCID - Minimal Clinically Important Difference
Figure 3
129x53mm (600 x 600 DPI)

Supplemental Data

Table S1 – **Focus group questions and discussion points**
How useful did you find the pulmonary rehabilitation course?
What limitations did you find as a patient with lung fibrosis?
Was the educational part of the course relevant to you?
What could be done to improve the course for patients with interstitial lung disease?
If you could design a course for patients with lung fibrosis, what would you include? How long would the sessions and the course in general last?

Table S2 - Summary of missing value analysis estimated means and standard deviations (SD)

		Age	Initial change in ISWT (m)	Initial change in ESWT (s)	Change in ISWT at 6 months (m)	Change in ISWT at 12 months (m)	Initial change in CRDQ	Change in CRDQ at 6 months
All Values	Mean	68.8	26.2	510	.0	-0.8	11.7	2.5
	SD	11.9	68.3	494	79.1	96.2	13.5	16.9
Expectation maximisation	Mean	68.78	26.2	517	2.7	-1.6	11.6	2.4
	SD	11.9	67.8	496	78.6	95.6	13.8	17.0
Regression	Mean	68.78	25.3	529	0.9	-11.5	11.7	1.0
	SD	11.9	66.5	493	75.9	104.6	13.8	16.5

ISWT (incremental shuttle walk test), ESWT (endurance shuttle walk test), CRDQ (chronic respiratory disease questionnaire)

Table S3 – Baseline characteristics of participants with IPF and Non-IPF ILD							
	Non-IPF ILD cohort n=51		IPF cohort n=28		95% Confidence Interval for difference		p value for comparison
	Mean	SD	Mean	SD			
Age (years)	66.33	13.08	73.25	7.63	2.27	11.56	<0.01
Male (n (%))	24 (47.1)		20 (71.4)				0.04
Smoking history (n (%))							
Ex-smoker	30 (58.8)		18 (64.3)				0.63
Never smoked	21 (41.2)		10 (35.7)				
Current smoker	0 (0)		0 (0)				
Death <12 months (n (%))	3 (5.9)		8 (28.6)				0.01
Followup (months)	36.20	27.59	25.25	19.86	-21.69	-0.21	0.05
FEV1 % predicted	75.20	23.16	74.38	18.10	-10.63	8.99	0.88
FVC % predicted	78.14	23.84	70.46	19.31	-17.99	2.63	0.14
DL_{CO} % predicted	51.11	19.57	39.24	14.37	-20.35	-3.39	0.01
TLC % predicted	75.37	18.93	59.76	14.04	-24.69	-6.53	<0.01
FVC trend prior to PR (% decline/yr)	1.05	17.03	-6.45	7.56	-13.97	-1.02	0.02
DL_{CO} trend prior to PR (% decline/yr)	1.88	17.09	-5.91	10.57	-15.28	-0.30	0.04
ISWT (m)	186.47	115.15	159.62	101.80	-78.26	24.55	0.30
MRC score	3.20	0.98	3.61	0.96	-0.43	0.87	0.08
CRDQ overall	77.08	17.24	80.39	20.44	-5.32	11.94	0.47
Mastery	18.16	4.84	18.96	5.79	-1.78	3.4	0.53
Emotion	30.92	8.24	33.75	8.95	-1.28	6.94	0.17
Fatigue	13.47	4.11	14.25	5.28	-1.54	3.1	0.50
Dyspnoea	14.53	5.41	13.43	4.23	-3.3	1.1	0.32
HADS overall	13.16	6.34	11.54	7.05	-4.83	1.59	0.316
Anxiety	6.98	4.01	5.43	3.96	-3.43	0.32	0.103
Depression	6.18	3.07	6.11	3.61	-1.69	1.55	0.932
Abbreviations – FVC (Forced Vital Capacity), FEV1 (Forced Expiratory Volume in 1 second), TLC (Total Lung Capacity), DL_{CO} (Diffusing Capacity for Carbon Monoxide), ISWT (Incremental Shuttle Walk Test), CRDQ (Chronic Respiratory Disease Questionnaire), HADS (Hospital Anxiety and Depression Score)							

List of abbreviations

QoL – Quality of Life

ILD – Interstitial lung disease

IPF – Idiopathic Pulmonary Fibrosis

PR – Pulmonary Rehabilitation

NICE – National Institute for Health and Care Excellence

COPD – Chronic Obstructive Pulmonary Disease

LEEP – Lung Education and Exercise Programme

BTS – British Thoracic Society

ESWT – Endurance Shuttle Walk Test

ISWT – Incremental Shuttle Walk Test

CRDQ – Chronic Respiratory Disease Questionnaire

MCID – Minimal clinically important difference

HADS – Hospital Anxiety and Depression Score

MRC – Medical Research Council

SD – Standard Deviation

CI – Confidence Interval

FVC – Forced Vital Capacity

DLCO – Diffusing Capacity for Carbon Monoxide

FEV1 – Forced Expiratory Volume in 1 second