Abstract M264 Table 1 Characteristics of the patients at baseline

	Pirfenidone N=96
Age – years (range)	67.1+/-8.1 (47–83)
Male sex - no (%)	71 (73)
BMI (range)	28+/-7.9 (14.4–43.4)
Former Smokers - no (%)	64 (66%)
Duration of Treatment – months (range)	9.3+/-8.3 (0–32)
FVC% Predicted (range)	72.9+/-23.1 (46–146)
FVC 51–80% Predicted - no (%)	69(72%)
FVC >80% Predicted – no (%)	20 (20%)
DLCO% Predicted (range)	43.6+/-19.9 (14–87)
DLCO <25% Predicted or unable – no (%)	12 (12)
DLCO 26–35% Predicted – no (%)	24 (25)
DLCO 36–65% Predicted – no (%)	45 (47)
DLCO >66% Predicted – no (%)	7 (7)
Use of supplementary oxygen - no (%)	22 (23)
Use of prednisolone – no (%)	26 (27)
Use of N-acetylcysteine – no (%)	22 (22)

Based on an annual unit cost of £22, 245.96 for pirfenidone (without undisclosed discount). To date 96 patients have been treated for a total of 876 months at a total cost of £1,623,955 in two and a half years.

Conclusion This study highlights both the health and economic impacts of pirfenidone over a two and a half year period of prescribing.

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M265 DAILY ACTIVITY MONITORING IN IDIOPATHIC PULMONARY FIBROSIS

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Introduction Idiopathic pulmonary fibrosis (IPF) is an incurable chronic progressive lung disease with a poor prognosis. Decline in forced vital capacity (FVC) is the primary outcome measure in most clinical trials. However, slowing lung function decline does not translate into patients feeling better. We investigated the acceptability of activity monitoring as a patient centred outcome measure in IPF and correlated results with lung function and quality of life (QoL) measures.

Methods IPF Subjects underwent activity monitoring 23 h a day for a minimum of 8 days using the SenseWear armband (Bodymedia, Philadelphia). Monitoring data from the first and last monitored days were discarded to prevent clinic visits impacting the results. Participants completed the St George's Respiratory Questionnaire (SGRQ) as a QoL measure. Lung function measurements performed within 3 months were collected and correlations assessed using Pearsons correlation coefficient. Data are presented as mean±SD.

Results 17 IPF subjects (Age 76 ± 6.3, 82% males, FVC%predicted 82.3 ± 16.1%, TLCO% predicted 48.3 ± 13.3%) were monitored. There was excellent compliance – armbands were worn for an average of 23 h and 9 min per day (range: 22 h and 10 min to 24 h) for 6.2 ± 0.6 complete days. Activity levels measured in METs were 1.25 ± 0.2 with a daily step count of 3364 ± 2504. IPF subjects were physically active (METs >3) for 83.8 ± 57.4 min per day. Mean daily METs inversely correlated with SGRQ score (r=-0.64, p = <0.01). Mean daily METs correlated with FVC (% predicted) (r = 0.50, p = 0.04) but there was no correlation with TLCO (% predicted) (r = 0.39, p = 0.13). Conversely TLCO inversely correlated with SGRQ score (r=-0.55, p = 0.03) but FVC did not (r=-0.29, p = 0.26).

Conclusion Activity monitoring is an acceptable, well tolerated means of measuring functional status in IPF patients. Mean daily activity level correlates well with QoL measures and FVC. Neither individual lung function measurement performed as well in terms of correlation with QoL and activity level. A larger longitudinal study is required to further evaluate the role of activity monitoring in IPF and identify its utility in prognostication.

M266 DEVELOPMENT OF AN IDIOPATHIC PULMONARY FIBROSIS (IPF) PATIENT REPORTED OUTCOME MEASURE (PROM): AN ITERATIVE APPROACH TO ITEM GENERATION

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Introduction Patients diagnosed with IPF experience debilitating symptoms which impact upon quality of life. To date there is no curative treatment and the tolerability andefficacy of existing andemergent therapies require further evaluation. We are developing a new concise IPF-PRoM (according to FDA criteria[1]) for use as a primary endpoint in studies exploring treatments of symptoms associated with IPF andas a secondary endpoint in clinical/therapeutic trials. Robust item generation is fundamental to the development of the IPF-PRoM reflecting what is important to patients and ensuring saturation is reached.

Methodology

- Domains and items were identified in existing symptom and quality of life measures used in IPF studies reported in the literature
- 5 focus groups were held at one of 3 UK centres. 28 patients (18 male) stratified for disease severity according to Composite Physiological Index (CPI) participated. Transcripts underwent inductive analysis and data was coded using NVIVO 10 ©QSR software
- Expert Opinion was sought from 10 ILD physicians utilising the Nominal Group technique. The importance of each descriptor identified in the literature was rated and then ranked according to overall importance. The top 5 were noted anddiscussed. Descriptors defined by focus group participants (n = 9) were added and the process repeated
- A multidisciplinary Research Support Group including patient and carer representatives contribute to the analysis at each stage and have the authority to mandate for the inclusion of 'grey' items.

Interim results A validation list applied to existing measures identified 208 items for inclusion. Systematic coding and recoding within NViVo reduced 28 categories initially identified to 10. Fatigue is identified as a dominant theme in patients with CPI ≥ 45 andmedication availability/impacts has emerged as a significant category in all groups. ILD experts place importance upon breathlessness and emotional and mental well-being (Table 1).

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Abstract M266 Table 1 Top '5' rankings from rounds 1 and 2 in the Nominal Group

Descriptor	R1	R2	Descriptor	R1	R2
Abandonment*			Planning analysing		
Chest Pain*			Relationships with others		
Control*			Sexual Relationships		
Cough			Shortness of breath	1	2
Energy Level	5		Sleep		
Fatigue*			Spirituality		
GORD			Therapies for IPF		
Level of independence		5	Uncertainty*		5
Mental & emotional wellbeing	2	3	Wheeze		
Mortality	2	1	Impact on Physical State		
Nausea*			Impact on Psychological health	4	3
Oxygen Therapy*			Impact on Social participation		
Perception by others*			Impact on Finances		
Phlegm - expectorating *			Impact of Chest Trouble on work		

*Key descriptors defined by focus group participants (n=9) added in Round 2

Discussion This methodological approach to item generation will enhance the content validity of the IPF-PROM instrument. Items generated to date will be modified further by 80 patients from 4 UK centres and 20 ILD physicians participating in 3 rounds of a Qualtrics Delphi survey. This study is ongoing.

1 http://www.fda.gov/downloads/Drugs/Guidances/UCM193282.pdf

M267 HEALTH STATUS AND QUALITY OF LIFE IN IDIOPATHIC PULMONARY FIBROSIS AND SARCOIDOSIS: EFFECT OF FATIGUE

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Introduction and Objective Sarcoidosis and Idiopathic Pulmonary Fibrosis (IPF) are two common forms of interstitial lung disease. Sarcoidosis frequently causes extra-pulmonary disease whereas IPF specifically affects the lungs. Fatigue is a common feature of sarcoidosis, but an association between fatigue and IPF has not been investigated. We investigated the frequency and severity of fatigue in sarcoidosis and IPF, how it correlates with quality of life (QOL) scores, and whether fatigue is affected by disease severity.

Methods This was a cross-sectional questionnaire study of patients with sarcoidosis and IPF. Questionnaire data was analysed to investigate health status, QOL, and symptom prevalence (fatigue, depression and sleepiness). Comparison of scores between groups, and an analysis of the effect of markers of disease severity on fatigue, was undertaken.

Results Questionnaires were administered to 235 participants; 82 healthy volunteers, 76 sarcoidosis patients and 77 IPF patients. IPF patients had statistically higher St George's Respiratory Questionnaire ($p = 0.034$) and Epworth Sleepiness Scale scores ($p = 0.003$) than sarcoidosis patients, but there was no difference in mean fatigue scores. When stratified by questionnaire scores (Table 1), including pathological fatigue levels, no statistical difference was seen between IPF and sarcoidosis, although there was a trend towards a higher frequency of 'severe fatigue' in sarcoidosis. Fatigue scores correlated strongly with quality of life scores (King's Brief Interstitial Lung Disease score and St George's Respiratory Questionnaire) in both IPF (r=-0.615 and 0.659 respectively) and sarcoidosis (r=-0.529 and 0.502). In sarcoidosis, no measures of dyspnoea or disease severity (spirometry abnormality, immunosuppression use or extrapulmonary disease) were associated with fatigue scores. In IPF increasing dyspnoea scores were associated with increased fatigue scores ($p < 0.001$).

Conclusions Both sarcoidosis and IPF patients suffer with high levels of fatigue, although the sarcoidosis cohort showed a trend towards greater frequency of severe fatigue compared with IPF. In IPF patients increasing fatigue was associated with worsening dyspnoea, suggesting an association with disease progression, but no similar relationship was seen in sarcoidosis. This suggests that fatigue in sarcoidosis occurs independently of common markers of disease activity, whereas it occurs as a sequelae of progressive disease in IPF.

Abstract M267 Table 1 Prevalence of Anxiety, depression, sleepiness and fatigue symptoms

		Healthy Volunteers	Sarcoidosis	IPF
HADS-A – Anxiety	Normal (%)	73 (89.0)	42 (60.9)	54 (71.4)
	Mild (%)	5 (6.1)	18 (26.1)	14 (18.2)
	Moderate (%)	3 (36.6)	8 (11.6)	6 (7.8)
	Severe (%)	1 (1.2)	1 (1.4)	2 (2.6)
HADS-D - Depression	Normal (%)	78 (95.1)	51 (73.9)	63 (82.9)
	Mild (%)	3 (36.6)	10 (14.5)	3 (3.9)
	Moderate (%)	1 (1.2)	5 (7.2)	8 (10.6)
	Severe (%)	0 (0)	3 (4.3)	2 (2.6)
Epworth Sleepiness Scale	No sleepiness (%)	66 (85.7)	47 (67.1)	52 (85.2)
	Borderline (%)	7 (9.1)	19(27.1)	7 (11.5)
	Severe Sleepiness (%)	4 (5.2)	4 (5.7)	2 (3.3)
Fatigue Assessment Scale	Normal (%)	74 (91.4)	34 (49.3)	40 (54.8)
	Fatigued (%)	5 (6.2)	27 (39.1)	30 (41.1)
	Severe Fatigue (%)	2 (2.4)	8 (11.6)	3 (4.1)

†Due to incomplete responses for some questionnaires there are fewer patients stratified by depression, anxiety, sleepiness and fatigue than were enrolled.

M268 THE IPF DIAGNOSIS – COMMUNICATING A LIFE SENTENCE

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Background The aim was to explore the patients' emotional experience of receiving a diagnosis of IPF.

Methods Market research was conducted with an independent agency. Patients with IPF were asked to record a personal account of their experience on a hand-held camera. Face to face interviews with patients were conducted in their home. Carers were also interviewed to add an alternative perspective.

Results The sample included 13 male and 3 female patients with IPF. Patients with lung function impairment of all severities were included, five patients were treated with oxygen therapy and another had received a lung transplant. There was a national spread geographically throughout England.

There is a fine balance to providing information and patients can benefit from an individually tailored approach. Too much information at the start can be overwhelming. Too little information can leave the individual uncertain about how to deal with their future.

Gaps that were identified focused on the practicalities of living with IPF, including social care. There was a high expectation for their physician to explain the trigger for developing IPF. Patients felt a blame culture exists, whereby others felt that IPF is self-inflicted, like COPD, particularly when a patient was

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