

ORIGINAL RESEARCH: EMPIRICAL RESEARCH –
QUALITATIVEPerceptions, experiences and needs of patients with idiopathic
pulmonary fibrosis

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Abstract

Aims. To understand the perceptions, needs and experiences of patients with Idiopathic Pulmonary Fibrosis.

Background. Idiopathic pulmonary fibrosis is a progressive interstitial lung disease, with a mean life expectancy similar to some forms of cancer of 2–4 years from diagnosis. Unlike the cancer literature, which is rich with studies exploring the needs of their disease group, few publications exist on patient needs with this severe fibrotic lung disease.

Design. A Qualitative study which took place between 2007–2012.

Methods. Seventeen patients with a multidisciplinary team confirmed diagnosis of Idiopathic Pulmonary Fibrosis, with moderate to advanced disease severity and six of their informal carers were interviewed. An interview topic guide was developed by the researchers and service user group. The interviews were audio-recorded, semi-structured and took place at a regional respiratory and lung transplant centre in North West England. Interviews were transcribed verbatim and data analysed using Framework Analysis.

Findings. Three main themes were identified: ‘Struggling to get a diagnosis’; ‘Loss of the life I previously had’; and ‘Living with Idiopathic Pulmonary Fibrosis’. Patients reported struggling to get a diagnosis and coping with a life-limiting, rapidly progressive illness with no good treatment and few support structures.

Conclusions. There is an urgent need for a better understanding of the difficulties faced by people with Idiopathic Pulmonary Fibrosis and their carers. This can be used to develop better supportive care in the United Kingdom and ultimately improve the quality of life of these patients.

Keywords: diagnosis, experiences, idiopathic pulmonary fibrosis, IPF, nursing, oxygen, palliative care, quality of life, respiratory, symptoms

Why is this research or review needed?

- To highlight the significant burden this disease has on quality of life in patients with Idiopathic Pulmonary Fibrosis.
- To generate discussion on how we can improve support structures for patients dying often unpredictably from Idiopathic Pulmonary Fibrosis.
- To remind us that Idiopathic Pulmonary Fibrosis is an uncommon but devastating cause of shortness of breath.

What are the key findings?

- Patients struggled to get a prompt diagnosis, often being misdiagnosed and treated for other more common respiratory diseases first such as asthma, COPD or recurrent chest infections.
- Patients experienced a sudden loss of life, as they knew it, with this rapidly deteriorating disease giving them little time to adjust and cope. Support structures were lacking in contrast to what is available for e.g. cancer care.

How should the findings be used to influence policy/practice/research/education?

- There is a clear need to establish properly funded regional Idiopathic Pulmonary Fibrosis networks with experienced staff conducting multidisciplinary teams similar to those used in cancer pathways to secure a timely diagnosis and treatment opportunities for patients.
- Patients should have easy early access to specialists who can provide information about the disease, discuss prognosis realistically and support them till death.
- There is a need for more healthcare education in recognising the signs and symptoms of this disease, the impact on quality of life and on how symptoms can be mitigated.

Introduction

Idiopathic pulmonary fibrosis (IPF) is a statistically significant progressive, incurable, interstitial lung disease (ILD). Navarantam *et al.* (2011) estimates 5000 new cases are diagnosed annually in the UK with 15,000 prevalent cases.

Current mean IPF life expectancy is 2–4 years from diagnosis (Wells & DuBois 1994) with a 5 year survival rate worse than for many cancers (Vancheri *et al.* 2010). Apart from its significant mortality, an IPF diagnosis leads to a high burden of morbidity with poor quality of life (Schoenheit *et al.* 2011).

In many regions of the UK, as yet, care networks to deal with this significant morbidity are underdeveloped or at least lack focus in this area. In addition a sizable propor-

tion of UK patients with IPF are still cared for solely by local chest physicians and do not make contact with the developing ILD networks.

Background

In a European study, Schoenheit *et al.* (2011) reported protracted times to diagnosis for IPF patients. They struggled to get through health services for 1.5 years (average) from a presentation, usually initially with shortness of breath. Three or more physicians were typically involved until finally they were clearly labelled as having IPF. During this journey patients reported experiencing a lack of empathy and support from their healthcare workers. They sensed a lack of competence; and some just clearly ‘lost faith’ in the system. Many were ‘misdiagnosed’ initially with more common respiratory conditions that also present with shortness of breath, for example, COPD, asthma, or recurrent chest infections.

After an often stressful path to a diagnosis problems seem to get compounded for patients with IPF. Bajwah *et al.* (2013), in a UK study, found that patients with ILD went onto receive inadequate information about their disease and its prognosis. This study took a broad stance assessing patients, carers and healthcare professionals. Unmet information needs were identified in all three groups that led to an inability to deliver key aspects of IPF care that can improve quality of life. This includes planning for end of life discussions and early referral to palliative care services for psychological support and symptom control.

Reporting unmet care needs in respiratory disease is not new. Diseases with comparable morbidity and mortality might include COPD and lung cancer. High symptom burden, poor quality of life and inadequate service provision especially at the end of life have been documented by many including (Gore *et al.* 2000, Elkington *et al.* 2005, Hasson *et al.* 2008, Pinnock *et al.* 2011). Raising awareness of these needs, however, has led to a stepwise improvement in care. Key quality of life management outputs are now expected in the day to day care of these patients and are included in their disease guidelines (National Institute of Clinical Excellence (NICE) 2011a,b). By raising the current plight of patients with IPF, we hope to evoke some positive changes in IPF care going forward.

Evidence has emerged that two main themes are deficient with IPF holistic care. The first relates to delays in diagnosis, the second around the support structures accessible to patients to deal with their serious diagnosis. If appropriate

UK ILD networks are established going forward both of these deficiencies could be tackled.

The study

Aims

The aim of this study was to understand the perceptions, experiences and needs of patients with IPF. Data collection occurred in 2007 and data analysis was completed in 2012. Despite this delay, the authors feel that patient experiences have not changed in a significant enough way yet in the UK and this data still reflects clinical events we are seeing in this patient group now in 2014. This data also supports other more current emerging evidence of poor patient experiences in IPF. We believe in further highlighting these issues so that clinicians will go on to incorporate solutions for these areas in their developing ILD services.

Design

The focus of this research was on 'what it is like to live with IPF', 'how patients cope with it' and what their support needs are. The design is qualitative after Silverman (2005), employing semi-structured interviews of approximately one hour duration.

Sample

Ethical and governance approvals were granted, respectively, from the Bolton Local Research Ethics committee and by the University Hospital of South Manchester Foundation Trust. As IPF is relatively uncommon it was more feasible to conduct the study at a regional specialist centre. Eligible patients with an MDT IPF diagnosis were approached by their clinicians to participate. They were given verbal information, an introductory letter and an information sheet and appropriate time (as per GCP guidance) to consider if they wanted to enter the study.

Seventeen patient interviews were conducted (seven males, 10 females). This represented approximately 90% of the patients with IPF at this centre at the time of data collection. Median age 67 years Ten patients were ex- or current smokers, seven never smokers. Mean %predicted FVC ($n = 17$) was 68% (range 44–104). Mean % predicted DLCO for 13 patients was 43% (range 23–67); four patients could not perform the manoeuvre. Most patients ($n = 11$) had lung function in 3 months of study. For other six, it was in 7–17 months from date of interview. For these six cases, their lung function data may

underestimate disease severity. Three patients had a family history of IPF.

Ten patients were receiving long-term oxygen therapy (LTOT) and ambulatory oxygen; two ambulatory oxygen only and five were not on any oxygen. Three patients were on a lung transplant waiting list. Three patients were in a clinical trial. In six of the 17 patient interviews, a family member contributed to the interviews in the form of – five spouses (three females, two males) and one daughter.

Data collection

Semi-structured interviews of approximately 1 hour duration were undertaken. They were with participants' permission, audio-recorded and transcribed verbatim, as were field notes made alongside the interviews. Data were collected by two researchers (AC and SB). Prior experience indicated that patients might want their main informal carer to be present during the interviews; this eventuality was therefore accommodated in study design and data collection. An interview topic guide (Appendix 1) was developed by drawing on the literature, expertise in the research team and guidance from an ILD patient/carers support group. Broad, open ended questions, supplemented by prompts from the researcher, gave participants the opportunity to tell their story.

Ethical considerations

Patients with advanced IPF can get very breathless even on speaking. We decided therefore to exclude patients with very advanced disease due to concerns about asking extremely breathless patients to be interviewed for one hour. We were also highly aware that we would be encouraging patients to talk about things that they might find distressing. However as the patient was leading the interview, we anticipated that they would only bring up things they wanted to discuss. Patients were reminded at the beginning of the interview that they could stop the interview at any time if they wished.

Data analysis

Data were analysed using Framework Analysis (Ritchie & Spencer 1994), a well-recognised method for analysis of qualitative data in health services research. This involves five steps: familiarization, identifying a thematic framework, indexing, charting and mapping and interpretation. One team member (AD) led on analysis, with a second (AC) supporting this. Pseudonyms have been used where extracts from data are presented.

Rigour

Principles of rigour in qualitative work, as described by Denzin and Lincoln (2005) and Silverman (2005) were adhered to. These included: having at least two project team members analyse each transcript, maintaining a clear audit trail in the analysis and inviting comments on the analysis/emerging themes from the ILD patient/carer support group. The authors said that this was more appropriate than sending transcripts/summaries back to participants due to the high mortality rate in the patient group and the length of time from data collection.

Findings

Most participants described their patient journey from noticing that something was wrong, to finally getting a diagnosis. The interviews typically shifted to reporting on symptoms, comparing current to past self and discussing the impact of the disease on daily living.

The main themes and sub-themes which emerged from the data were:

- 1 Struggling to get a diagnosis
- 2 Loss of the life I previously had
 - Loss of independence and roles within relationships
 - Loss of spontaneity
- 3 Living with IPF
 - Struggling with increasing breathlessness
 - Struggling with cough
 - Struggling with tiredness and lethargy
 - You cannot do this alone-support structures, including support from specialists,
 - Living with oxygen
 - Hoping for a successful treatment

Struggling to get a diagnosis

Most participants could identify a beginning to their problem, with phrases like:

‘...it started with a cough’ ‘... ‘I was getting a bit breathless going upstairs’ being typical. From there on, the journey to diagnosis was variable, usually causing anxiety and frustration. Participants described a lengthy delay from being treated in primary care initially for asthma, COPD or recurrent chest infections before being referred to a local chest physician

Even in secondary care the struggle to obtain a diagnosis often continued, as Margaret was told by one physician:

...its nothing more than stress and anxiety

One patient reported it taking 4 years to obtain a correct diagnosis which had a very negative impact on their well-being. Several others reported being ‘misdiagnosed’ and, as a result, being unresponsive to prescribed treatments usually inhaler therapy. Most participants found their diagnostic journey distressing and upsetting, feeling symptoms and problems were trivialized. Participants indicated that, despite being recommended in international guidelines (British Thoracic Society 2008, American Thoracic Society 2011), no information was given about prognosis or treatment options, including the option to enter a clinical trial:

...some kind of fibrosis.....so I just kept going to the clinic and they did lung function....which....at the time....were quite low....then he decided to refer me to Dr. X’s clinic and then we found out that I was too bad for the trial [Bridget]

Loss of the life I previously had

Participants described two main dimensions of loss:

Loss of independence and roles in relationships

Loss of independence was significant as disease progressed, with participants becoming very dependent on partners and others. They found it difficult to continue many of their usual roles, causing guilt and frustration, as Jim explained:

I’ve always been active, garden, DIY, you know I’d do anything around the house. I mean there was a little job there yesterday and I had to get Christine to (do it)...I sort of direct operations now, but physically do it, No’. [Jim]

Loss of spontaneity

Breathlessness severely impacted on patient’s lives, making it difficult for them to be spontaneous, necessitating meticulous planning of all activities inside and outside the house:

So it’s getting around I think is the big change...actually doing jobsyou’ve got to think before you do anything, if you want to..... just nip into the shops, you’ve gotta think ‘have I enough liquid oxygen? Have I filled the bottle? Have I done this? Have I done that? [Terry]

Living with IPF

Participants indicated that IPF affected every aspect of their lives and described ways that they sought to live with the condition and its associated limitations:

Struggling with increasing breathlessness

Breathlessness on even minimal exertion was a significant symptom for all participants, affecting most activities of daily living:

...when I shave, I put me (my) shaving cream on, I've got to take the oxygen off and I know it doesn't take long...but I can't wait to get the oxygen back on....(I am) thinking about getting an electric shaver' cause I can just lie down and use it'. [Frank]

Struggling with cough. Cough was often described as a presenting symptom, with or without breathlessness, some describing it as a '...silly little cough' and others describing it as an '...unpredictable hacking cough that leaves you exhausted', often associated with exertion '...I definitely cough when I'm doing something physical'

Many described bringing up thick white phlegm that looks like '...wallpaper paste'. Cough was cited by several participants, of both sexes, as a cause of urinary incontinence and, in one female, of faecal incontinence:

...when I go upstairs and sometimes I walk up, no trouble, ...cough very, very little...other times I walk up, start coughing and then I've really got to go to the toilet and if I don't go to the toilet, well...sometimes I wet myself [Terry]

Struggling with tiredness and lethargy

Participants described feeling tired all the time. Even simple tasks such as reading and watching television was an effort and respondents reported lacking concentration to complete activities that they would had once enjoyed:

Over 30 years we've been playing cards, alternate weeks....playing poker and that's stopped 'cause she can't concentrate when we're playing. [Carer of Bridget]

A few patients did admit to feeling depressed which seemed to be associated with feelings of lack of control, losing a sense of self and having to give up roles they once enjoyed. Other participants described having good and bad days, with periods of tiredness and weariness.

You cannot do this alone

Participants described becoming more dependent on their partners, to help with personal cleanliness and adjusting oxygen. This disease also impacted on carers as Jim described:

.....you are in this together, its not me,..its not Christine...we are in this together...like going up and down the stairs....and I say to Christine when you come down will you bring me so and so.

Many participants felt this was an 'unseen' disease as Bridget comments:

...sometimes I think....do people think I am lazy?...because they will say, you look so well. [Bridget]

Respondents report having increased confidence when they were being cared for by clinicians who understood their condition, as Terry indicated when describing his first visit to the specialist centre:

I came in and saw the nurse and she knew more and understood more about my complaint than anyone I'd seen prior...it gives you confidence...[Terry]

Participants measured disease progression by loss of functional ability over time. They described things that they could do in the past but now no longer could. They constantly referred back to the person they had once been, illustrating how quickly the disease had got a grip and completely turned their worlds upside down.

...cause I've been so active you see I used to go out and do 40 miles (on the bike) before I went to work then I used to go out and do the same at night... and I've rode across the country in 3 days and 216 miles in 12 hours....[Arthur]

Telling the interviewer about his past life as a racing cyclist seemed to animate Arthur. As he told his stories, he seemed to fantasise about the person he was and it seemed important to him to impress on the interviewer that he had not always been like this.

The study site had a support group for patients with IPF. Participants valued this support group and found it useful talking to others with the same condition. Gauging themselves against others at the support group helped some to put their disease in perspective, as Stella commented:

.....there were people there that had it 7 years who were on oxygen and looked really really poorlyI started picking up on it...it came out in conversations and I was twigging away there thinking gosh I'm at the beginning.and I thought I was getting near the end. [Stella]

Living with oxygen

Most participants were using long-term oxygen therapy (LTOT) and/or ambulatory oxygen. Most participants said that oxygen helped their breathlessness and some felt it helped their cough. Despite being symbolic of deteriorating disease, most participants described how oxygen improved their confidence and enabled them to do the things they wanted as Terry described:

...with the oxygen on at least I can get around, not as fast as normally, but I can get around...and I don't seem to cough as much. [Terry]

Learning to use oxygen, to help manage breathlessness helped participants to feel more in control of their lives. By contrast, however, being so dependent on oxygen meant that there was the constant worry that it would run out, meaning that participants needed to do careful calculations to know how long they could be out of the house. Some participants found it difficult to go without oxygen even briefly; removing oxygen to dress, wash or brush their teeth caused increased breathlessness and anxiety:

....I couldn't sit here this long talking to you without oxygen. [Frank]

Hoping for successful treatment

Participants who were waiting for a lung transplant or in a clinical trial were more hopeful than other participants, anticipating that receipt of a transplant or success of the trial drug would yield a 'cure' and a return to a 'normal' life, as Paul's carer commented:

...it's his only chance really, unless there is something else....

However, for those awaiting a transplant, anxiety increased with progressive functional disability and the longer they waited. Not knowing when they might get a call for a lung transplant meant that these participants lived with the daily anxiety of being called, but also of not being called for a transplant in time. Those participants for whom a transplant or clinical trial was not an option held a much bleaker outlook:

I just couldn't come to terms with it...I was getting panic attacks...two or three times a week...and used to break down...cry like a baby...there were times I just wanted to smash my head against a wall [Geoffrey]

Discussion

Participants in this study struggled to get an accurate prompt diagnosis. They experienced functional limitation, rapid disease progression with limited support and few positive treatment options. Participant stories centred round loss of sense of self, the life they had and the person they were. Learning to live with IPF was a struggle, with increasing dependency on partners and specialists. Oxygen became a lifeline to many and helped them to feel 'in control'.

Patients in this study could identify a beginning to their disease in contrast to studies in COPD (Pinnock *et al.* 2011), where their diagnosis seemed to creep up on them more. Being very aware of how long they had symptoms and how long it took them to get to a diagnosis appears to have contributed significantly to their anxieties. Earlier diagnosis could have relieved some of this stress. In line with Schoenheit *et al.* (2011) most participants in our study were initially misdiagnosed with asthma, COPD or recurrent chest infections. This compounded their feelings of frustration and helplessness because the treatments being given to patients for these diseases were not helping them and their symptoms continued to trouble them. Education in primary care is warranted to try and avoid diagnostic delays and to raise awareness of IPF. Patients who report breathlessness with or without cough unresponsive to inhaled therapy, with crackles heard in their lungs on auscultation should be considered for a chest x-ray even with normal spirometry. These fairly simple tests available in general practice should permit earlier identification of an 'at risk group' that would benefit from a hospital chest specialist referral. If local chest specialists then suspect IPF as a potential diagnosis a referral onwards to a specialized ILD MDT centre can be considered.

In our study and similar to lung cancer, increased breathlessness was symbolic of deteriorating disease (NICE 2011a,b) and negatively impacted on quality of life. Despite symbolizing deteriorating disease oxygen, as described by our patients was a lifeline that relieved breathlessness. Oxygen was liberating and helped patients maintain social interaction, leave the home and do the things that were important to them. An ambulatory prescription of oxygen in the home meant that patients could continue to complete activities of daily living and were less dependent on their spouse or carer. Ensuring that patients have regular oxygen review as disease progresses is important in this patient group and their specific ambulatory needs inside and outside the home should be considered by oxygen assessment services.

Pulmonary Rehabilitation is recommended in the National Institute of Clinical Excellence (NICE) (2013) IPF guidelines as a possible supportive therapy. Pulmonary rehabilitation is already an established supportive therapy for patients with COPD (Singh *et al.* 1998) and can improve distance walked shortness of breath and help develop coping strategies. Despite differing disease pathophysiology, Holland *et al.* (2013) suggest that exertional dyspnoea, fatigue and depression seen in patients with IPF is similar to that seen in COPD patients. Worsening functional impairment clearly has an impact on quality of life

(Holland *et al.* 2013) and therefore small, but modest benefits experienced from pulmonary rehabilitation in a disease where there is little else to offer may well be worthwhile to patients with IPF. There is still unequal access to pulmonary rehabilitation throughout the UK, which needs to be addressed and more research needed on the value of this therapy in this patient group.

In her study looking at information and support needs, Bajwah *et al.* (2013) describes how patients with interstitial pneumonia (including IPF) were not given sufficient information about disease progression which caused anxiety and delayed planning for the future. When dealing with uncommon diseases like IPF inexperienced clinicians may not have a good feel for prognosis, which will limit their ability to open up discussions around end of life. There is much evidence that addressing issues of prognosis and end of life care can be beneficial for patients. Van Der Molen (1999) suggests that in order to develop effective coping strategies, individuals need to know the extent of future challenges. Living with uncertainty causes fear, anxiety and frustration particularly when it is associated with healthcare practitioners not understanding the significance of progressive, debilitating, unresponsive symptoms. Miller (2000) proposes that not knowing what the future may hold means that patients are unable to mobilize their internal resources and support structures to develop effective coping strategies.

Lindell *et al.* (2010) introduced a psycho-educational intervention programme to patients with IPF to explore whether having more knowledge about their disease could help patients develop coping strategies. There were some positive benefits from this study in that patients described feeling less isolated, were more able to put their disease into perspective and they had an improved mental picture of their status.

In our study a theme that came through as a positive source of support for patients was the peer support group. Patients valued meeting others in a similar situation to them and it reduced feelings of social isolation. This theme is in common with other studies (Milne *et al.* 2009, Berendes *et al.* 2010, Rustoen *et al.* 2010, Schoenheit *et al.* 2011). There is a need to further develop IPF patient support networks in the UK. This work is currently underway driven by charities such as Action for Pulmonary Fibrosis and the British Lung Foundation.

Patients with IPF experience the rapid onset of debilitating symptoms that will shorten their lives and diminish quality of life (Swigris *et al.* 2005). Gore *et al.* (2000) and Elkington *et al.* (2005) have already highlighted how patients with COPD have support needs comparable with those seen in cancer. Our research suggests that patients

with IPF have an even greater need of similar services. Patients with cancer are offered many specialist support services (Murray *et al.* 2002). Against a backdrop of increasing IPF prevalence, (Navaratnam *et al.* 2011), service provision however, for patients in the UK with IPF still remains poor.

This research suggests the need for improved diagnostic and support services for people with IPF; development of regional networks of specialist centres with MDT diagnosis and properly funded support networks. The UK National Health Service (NHS) is in the process of developing regional ILD networks. These networks will aim to deliver specified service functions that will improve the national delivery of ILD care. Services should be safe, accessible, effective and evidenced based where possible. Until regional ILD networks have been fully developed patients will continue to be managed in an ad hoc manner depending on local service provision.

As recommended in National Institute of Clinical Excellence (NICE) (2013) guidance, all specialist ILD centres should have patient access to an ILD specialist nurse from diagnosis through to end of life care. The ILD specialist nurse should attend the ILD MDT so that he/she can accurately convey information to the patient about their diagnosis, treatment options, management plan and prognosis.

The ILD specialist nurse who is familiar with the disease trajectory could act as the patient anchor and main support and can co-ordinate other healthcare clinician involvement at the appropriate time. The ILD specialist nurse, understanding his/her local, community healthcare provision has the ability to reach out to involve other health care services. This might include community respiratory, palliative care and social care teams, district and community nurses. For patients dealing with disease progression is one matter in itself but suffering additionally from unaddressed symptoms and having no-one to turn to is a burden that could be easily removed. ILD nurses can provide a vital support to patients and link to other specialists. All clinicians need to be able to recognise advancing disease and increasing symptoms and consider earlier use of drugs to treat breathlessness and know when to refer to palliative care services. Clinical specialists and palliative care teams working together can improve care where there is an uncertain disease trajectory (Murray *et al.* 2005) with both treatment management and symptom control running in parallel.

Limitations of study

It is important to note that this research was conducted in a single specialist respiratory and lung transplant centre,

where respondents had access to specialist ILD clinicians and a support group. The majority of patients do not have this in the UK, hence our participants' experiences may represent 'the tip of the iceberg'. However, the patients attending this centre were travelling from a fairly wide geographical area so this work at least reflects care across a region at the time. The authors are aware that data collection started in 2007 and times might have moved on. However, reflecting on current clinical interactions with newly referred patients and in addition to other more recent published work which reflects similar findings to our study the problems for patients with IPF continues (Schoenheit *et al.* 2011, Bajwah *et al.* 2013). Interest in developing support structures for this patient group is continuing to emerge. Byrne *et al.* (2014) describes a protocol study using mixed methods at different stages of the disease to look at support and information needs across three respiratory centres in the UK. It is hoped that this work will add more to the growing body of knowledge of the need to improve patient pathways and access to care for this as yet vulnerable patient group.

Conclusion

This study reinforces the growing body of knowledge highlighting that patients with IPF currently have many unmet healthcare needs. It is novel in that it contributes to the literature with powerful accounts of patients' difficulties in their words and those of their carers. Support solutions for patients with IPF need to be implemented. Some of these are fairly obvious, from the literature particularly around end of life care and symptom management. Once implemented follow up work will be required to assess the effectiveness of these supportive interventions.

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Conflict of interest

No conflict of interest has been declared by the authors.

Author contributions

All authors have agreed on the final version and meet at least one of the following criteria [recommended by the ICMJE (http://www.icmje.org/ethical_1author.html)]:

- substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data;
- drafting the article or revising it critically for important intellectual content.

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Appendix: Interview Topic Guide

Symptom

- Which ones experienced? Respiratory/non-respiratory
- How often/when?
- How severely?
- How bothersome? Which are the most troublesome/distressing?
- Characteristics of the symptom

Impact

- On daily living activities
- Psychological impact
- Concerns/worries/fears
- Views re future outlook
- Effect on work/social/family roles (what impacted upon? how much?)

Treatment

- On any? Which?
- Benefits
- Side effects/difficulties
- Worries/concerns re treatment

Understanding

- Awareness of diagnosis
- Understanding of diagnosis
- Understanding of prognosis
- Worries/concerns re diagnosis/prognosis

Coping

- What helps them? What do they do to cope with/minimise their symptoms/problems?
- What have they found unhelpful?

Struggling to get a diagnosis

'I was diagnosed 4 years ago, but I was being treated for asthma and being overweight'. Carol
'..... there is (was) a query of pulmonary embolism' but then, '.....(he) carried on treating (him) for asthma'. Terry

Loss of the life I had

'I was a person who would decorate in one day.and I would have the flowers in the window.but I'm findingI don't know what I'm doing.....and I'm getting myself in a dither where I was never like that. I do a lot of sewing, but I'm finding I can't be mithered.I'm in such a mess'. Sarah

You cannot do this alone

'I should be looking out for him and I always thought I would have been because he was diabetic.and its turned out the other way round, he's turned out to be my carer. I couldn't cope without him,..' Marion
'...I don't really feel you have got any back up...like you know you can ring your doctor, which they don't really know a lot about it...I just feel that you're left with this condition and it's a horrible condition' Carol
'I could pick that phone up now and leave a message on her answer-phone and tomorrow morning she'd be on the phone to me. she never lets you down.its nice to know that you have somebody to talk too' Marion
'.....its much nicer to see doctors who seem to understand the condition, who specialise in it...the point is it gives you confidence'. Terry
'Those groups are invaluable. its surprising how you pick things up...it is a lot easier to talk to somebody who has it as well' Jim

Living with Idiopathic Pulmonary Fibrosis

'...everything I do is snail's pace and that way, well I still get breathless but I can control it' Felicity
'I just couldn't get my breath, I was fighting for air...and you put your oxygen on and it doesn't help straight away...you've got to try and relax' Geoffrey
'.....you will have to get me some sanitary towels cause every time I cough I wee, I dribble' Patricia
'...I could be working in the office and I'd be required to go and give a talk..... and I'd start coughing and choking and couldn't get me breath for 10 mins because I was changing atmospheres'. Terry
'...we've been to Tescos and I've run out (oxygen).... I was really gagging for breath. That's when the panic attack starts setting in and you know they're terrible. I mean I have to carry valium all the time now'. Geoffrey
'...if I am going upstairs, my wife will turn the oxygen up so that once I'm upstairs, getting undressed, going to the toilet, once I'm settled upstairs I shout down for her to turn the oxygen level down.and then morning when I get up (it's the) reverse'. Jim

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