Gaps in care of patients living with pulmonary fibrosis: a joint patient and expert statement on the results of a Europe-wide survey

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ABSTRACT

Background

Pulmonary fibrosis (PF) and its most common form, idiopathic pulmonary fibrosis (IPF), are chronic, progressive diseases resulting in increasing loss of lung function, impaired quality of life and survival. The aim of this joint Expert and Patient Statement was to highlight the most pressing common unmet needs of patients with PF and IPF, putting forward recommendations to improve the quality of life and health outcomes throughout the patient journey.

Methods

Two online surveys for patients and healthcare providers were conducted by the European Idiopathic Pulmonary Fibrosis and Related Disorders Federation (EU-IPFF) in 14 European countries.

Results

The surveys were answered by 286 patients and 69 healthcare professionals, including physicians and nurses. Delays in diagnosis and timely access to ILD specialists and pharmacological treatment have been identified as important gaps in care. Additionally, patients and healthcare professionals reported that a greater focus on symptom-centered management, adequate information, trial information, and increasing awareness of PF/IPF was required.

Conclusion

The surveys offer important insights into the current unmet needs of PF/IPF patients. Interventions at different points of the care pathway are needed to improve patient experience.

Keywords

IPF; Interstitial Lung Diseases/ Pulmonary Fibrosis; Doctor-Patient Relationship; Early Diagnosis; Treatment access; Health outcomes.

INTRODUCTION

Interstitial lung diseases (ILDs) comprise a diverse collection of more than 200 lung disorders, affecting the interstitium of the lung (1). A large subgroup of patients with ILD have pulmonary fibrosis (PF); most forms of PF are characterized by a progressive phenotype, are associated with a high burden of disease and have devastating consequences for patients and their families (2-4). Idiopathic pulmonary fibrosis (IPF) is the most frequent form and accounts for 17-37% of all ILDs (5). A cure for IPF does not currently exist, although there are two drugs approved that slow disease progression (6, 7). Non-pharmacological treatment options include lung transplantation to prolong life and measures such as pulmonary rehabilitation and supplemental oxygen to ameliorate exercise tolerance and quality of life (8-10).

In 2016, a collaborative effort of patient associations and healthcare providers was undertaken to gain insights in the needs of patients with IPF, which led to a European IPF Charter (11). This charter was presented at the European Parliament to improve awareness and equal access to care around Europe for patients with IPF. We hoped that this would lead to improvement in the care and treatment of patients with fibrotic lung diseases. One of the aims of the current study was to see whether this happened or not. To do so, we aimed to identify the most pressing common unmet needs of patients with PF and IPF throughout Europe and to put forward recommendations in an Expert Statement to improve quality of life and health outcomes throughout the patient journey.

PARTICIPANTS AND METHODS

The study was conducted by the European Idiopathic Pulmonary Fibrosis and Related Disorders Federation (EU-IPFF) in association with the European Reference Network on Rare Lung Diseases (ERN-Lung). This Expert Statement is a result of the collaboration between patient representatives and medical experts. Two online surveys were developed: one for PF/IPF patients and one for practicing pulmonologists and nurses with ILD expertise. The questions for the surveys were developed by the EU-IPPF working group, consisting of four patient representatives and 14 ILD experts. The group met in person to discuss the topics of the surveys and to reach consensus on the questions. Both surveys contained 62 questions and were circulated between 29 June 2018 and 8 September 2018 in 14 countries. The survey has been created in SurveyMonkey (www.surveymonkey.com). An information sheet was developed to inform respondents about the purpose of the project. All respondents were asked to read and understand the terms of the questionnaire and provide their consent. The survey for healthcare profes-

sionals (HCPs) was distributed through the ERN-LUNG network and the patient survey through the EU-IPFF's 17 member organizations *via* an email that contained a link to the survey. Caregivers were allowed to respond to the survey on behalf of the patient. The surveys are available in the supplementary material. This study is exempt from ethics review because it solely consists of an online survey that was disseminated to patients *via* patient groups.

Results have been divided into four geographical sub-regions: Northern Europe (Denmark, Ireland and UK), Eastern Europe (Bulgaria, Czech Republic and Poland), Southern Europe (Greece, Italy and Spain), and Western Europe (Austria, Belgium, France, Germany, and the Netherlands) (12). Results were collected, tabulated in Excel (Microsoft, Redmond, WA, USA) and bar graphs were generated. Pearson chi-squared test has been used to compare between geographical sub-regions. Data were analyzed with R version 3.5.2 (www.r-project.org).

In addition, a literature search was conducted for articles about the care pathway and unmet needs of patients with PF/IPF. PubMed and Embase have been searched for articles published between January 2010 and March 2018, using the (MesH) terms "idiopathic pulmonary fibrosis", "pulmonary fibrosis", "interstitial lung disease" or "diffuse parenchymal lung disease" in combination with "care pathway", "unmet needs", and/or "barriers". The search was limited to adults and articles published in English. The reference lists of articles were manually screened for additional publications. Relevant articles were included in order to create an overview on the state of knowledge on the care pathway and unmet needs of patients with PF/IPF. Results of the literature search will be used to compare gaps in care from previous research with results of the current study.

RESULTS

Literature search

The literature search retrieved 1111 articles, of which 966 articles were excluded based on title and abstract. After full-text screening of the remaining articles and exclusion of studies without relevant information on unmet needs and the care pathway in PF/IPF, 22 studies were included (see the supplementary material for details). Unmet needs reported by patients and caregivers were extracted from these studies and displayed in **Table 1**.

Table 1. Unmet needs of patients with PF/IPF reported by patients and caregivers

	Reference(s)
Timely and accurate diagnosis	(11, 13-26)
More awareness of PF/IPF	(11, 13-15, 18-21, 23, 26, 27)
Adequate information and education	(11, 13-19, 22-24, 26-32)
Access to pharmacological treatment	(11, 14, 15, 19-21, 26)
ILD specialists	(11, 13-16, 18, 19, 22, 25-27)
Symptom relief	(17, 18, 20, 21, 23, 28, 30)
Psychological support	(11, 13-15, 17, 23, 26-28)
More involvement and support of partners	(14, 16, 17, 24, 28, 32, 33)
Non-pharmacological management (i.e. supplemental oxygen, pulmonary rehabilitation)	(11, 13, 15-17, 19, 21-23, 26, 30)
Access to a multidisciplinary team	(11, 16, 17)
End-of-life care	(11, 16, 19, 20, 22, 23, 26-30)

Survey results

Respondent characteristics

The patient survey was completed by 286 individuals from 14 different countries, of whom 79% were patients and 21% were caregivers (**Figure 1)**. The majority of patients had IPF (86%) and 14% of respondents had another type of PF. Patients reported diagnosis between 1987 and 2018. The mean age of patients was 66 years, and 70% were male. A fifth of respondents (21%) reported a history of PF/IPF in their families. The questionnaire for HCPs was completed by 69 respondents: 56 physicians (81%) and 13 specialist nurses (19%). Most HCPs (87%) were specialized in ILD and worked at recognized centers of expertise. There was a large variation in the reported number of patients with PF/IPF treated per center (range 5-3000). The estimated total number of patients managed per year among all participating centers collectively was 10,000-11,000 for IPF and 27,000-28,000 for other forms of PF.

Referral pathways and access to ILD specialist care

In order to assess the delay in access to a pulmonary physician, patients were asked to indicate how much time passed before their general practitioner referred them to a respiratory specialist. Almost half of patients (45%) reported that referral took place within one month. In contrast, time to referral was >1 year for 16% of patients. No evident differences in referral time were found across Europe (p=0.84) (**Figure 2**).

Furthermore, 33% of patients reported that their referral to a specialist center took <1 month, with 20% reporting a wait of >1 year. Fewer than half of patients (47%) reported that a referral to a specialist center was (very) easy to obtain, whereas 20% considered it a (very) difficult process. More than a third of PF/IPF patients (37%) reported at least one

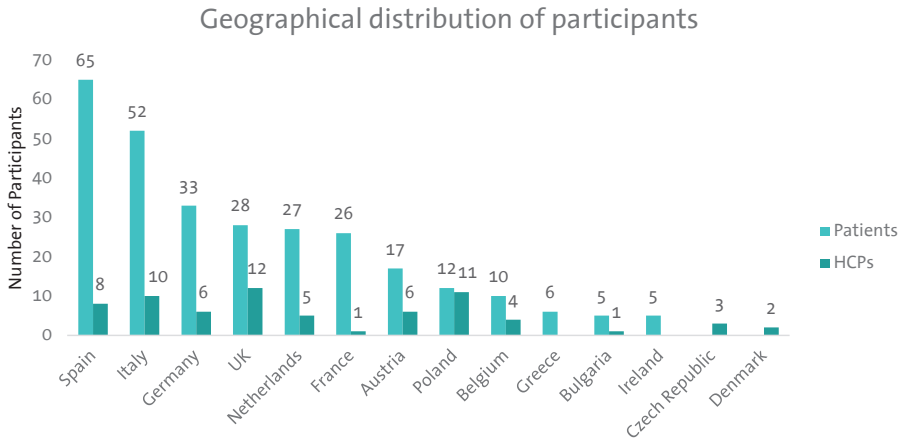

Figure 1. Number of participants (patients and HCPs) per country. HCP = healthcare professional

other diagnosis prior to being diagnosed with PF/IPF. Half of these patients indicated that >1 year passed before they were correctly diagnosed (**Figure 3**).

The vast majority of HCPs (94%) reported that there was access to a multidisciplinary team (MDT) for all IPF/PF patients in their center, but composition of the MDT varied greatly. In the patient survey, 58% of respondents stated that diagnosis has been confirmed in an MDT meeting. However, it is unknown if all patients were aware that their case was evaluated in an MDT. Around two-third of HCPs (65%) answered that ILD specialist nurses were available in their center, while 52% of PF/IPF patients responded that they had access to specialist nurses.

Reported access to genetic screening varied. Half of the participating HCPs (49%) stated that genetic screening was offered, either in their own center or *via* referral to another center. In total, 16% of surveyed patients underwent genetic testing; of these 45 patients, 42% stated that they did not receive enough information about their results of the genetic tests.

Access to pharmacological treatment for IPF patients

Both approved treatments for IPF, *i.e.* nintedanib and pirfenidone, were available in all participating countries. Almost all HCPs (93%) confirmed that antifibrotic drugs could be prescribed in their centers. The majority of respondents with IPF (82%) were treated with either nintedanib or pirfenidone at the time of the survey.

The time from diagnosis to initiation of treatment varied greatly, and this was reflected throughout Europe **(Figure 4)**. No statistical differences were found between sub-regions (p=0.16). Although antifibrotic treatment was initiated <1 month after diagnosis in 31% of patients, more than a quarter of patients (26%) reported that they

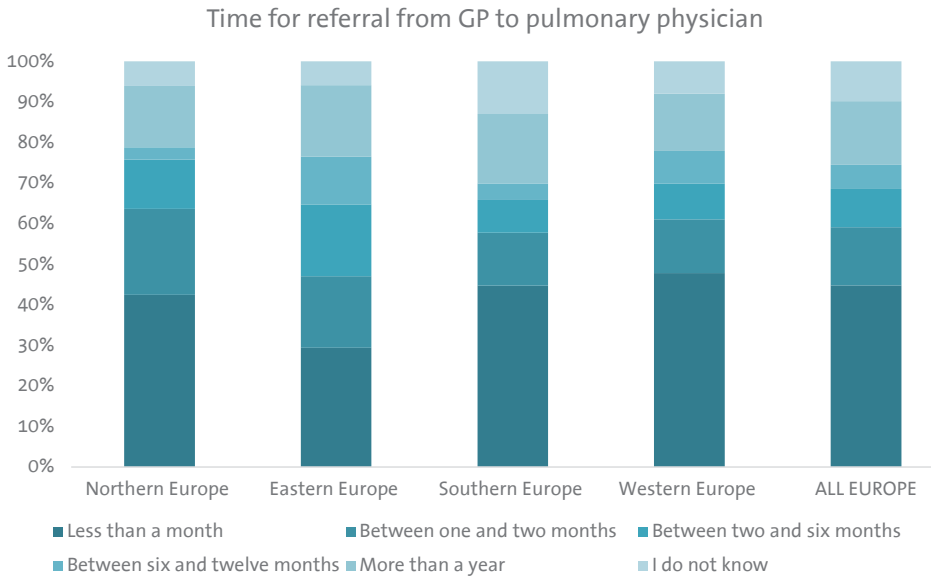

Figure 2. Time for referral from general practitioner (GP) to a pulmonary physician (patient survey)

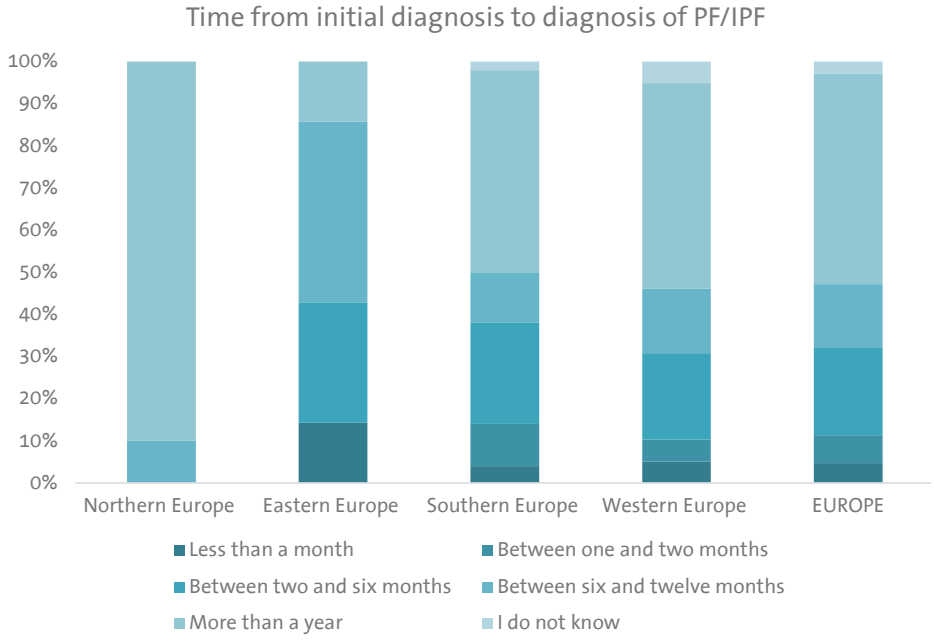

Figure 3. Time from initial diagnosis to diagnosis of PF/IPF (patient survey)

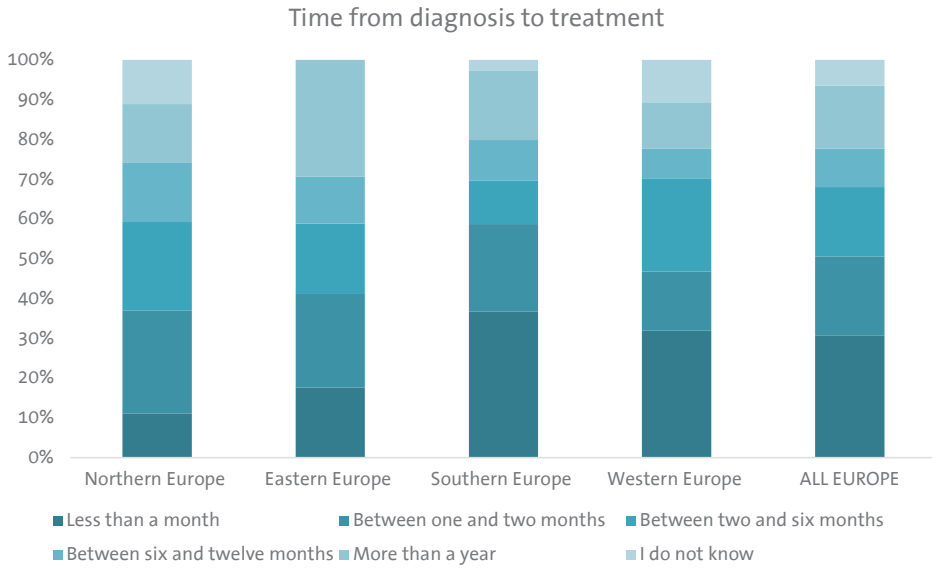

Figure 4. Time between diagnosis to start of antifibrotic treatment (patient survey)

had to wait >6 months before antifibrotic treatment was started. HCPs reported reimbursement restrictions as the main reason for this delay; 78% of respondents confirmed that reimbursement restrictions for prescription of antifibrotic treatment exist in their country. In some countries, antifibrotic drugs are only reimbursed when patients are diagnosed in an ILD specialist center, and in others lung function and/or age criteria exist. Specific lung function criteria were identified as the main barrier for prescription of antifibrotic medication by 70% of HCPs.

Access to non-pharmacological treatment

Almost all HCPs (97%) were able to prescribe oxygen therapy for PF/IPF patients. More than three quarters of patients (78%) reported full coverage for the costs of ambulatory oxygen therapy, and two-thirds of patients (64%) for the costs of oxygen at home.

The vast majority of HCPs (88%) could refer patients for pulmonary rehabilitation. A third of HCPs answered that pulmonary rehabilitation was not fully reimbursed in their country. Fewer than half of patients (42%) stated that they had access to outpatient pulmonary rehabilitation; 11% of patients also had access to inpatient pulmonary rehabilitation. Just over half of HCPs (58%) reported that their patients had access to psychological support at their center, with full reimbursement for 70% of patients. Patients were not specifically questioned about access to psychological support; however, 10% of patients spontaneously reported the need for (better) psychological support throughout their disease course.

The most reported eligibility criteria for lung transplantation concerned age and general health condition. Most HCPs (96%) reported that all eligible patients were referred for lung transplant. In one of the surveyed countries, lung transplantation was not possible at the moment of the survey.

Access to palliative care

Of the surveyed patients, 29% confirmed access to palliative care and 36% answered that they were involved in palliative care decisions. The majority of HCPs (88%) stated that they discuss possibilities for end-of-life care with patients, and almost all HCPs (93%) could prescribe (palliative) medication for symptom relief.

HCPs were asked to explain at which point in the disease course they initiate palliative care for their patients. Around one third answered that palliative care was started at an early stage of the disease if desired by patients. Most HCPs reported that palliative care is initiated in more advanced stages of PF/IPF. One fifth stated that palliative care was only initiated at the end-of-life.

Communication and education

The majority of patients (60%) had a positive experience while discussing their diagnosis with the pulmonary physician. However, a fifth of patients answered that they did not receive any information about their disease at the time of diagnosis. Three out of four patients (73%) and 60% of HCPs felt that there was enough time to discuss diagnosis and treatment options. Only 39% of HCPs reported that they received training on how to effectively communicate information on diagnosis and treatment of PF/IPF with their patients. Three quarters of patients received a treatment plan following their diagnosis, which was clearly explained in 73% of cases. Less than a third of patients (31%) were involved in development of their treatment plan; this involvement was mostly related to the selection and dosage of antifibrotic medication, initiation of non-pharmacological management and participation in clinical trials.

Patients were asked to give recommendations on how healthcare staff could work more effectively with them and their caregivers. Many patients answered that they would like to have more time allocated for their questions and concerns, and receive more information about PF/IPF including practical issues such as reimbursement. Furthermore, patients mentioned the need for timely referral to a specialist center and more awareness of PF/IPF amongst general practitioners, nurses and physicians in community hospitals. Around two-thirds of participating centers (65%) offered educational activities specifically for PF/IPF patients, such as nurse led education sessions, information meetings, eHealth programs and patient support groups. Among the surveyed patients, 39% attended educational sessions in their treating center.

Involvement in research

The majority of HCPs (95%) reported that their center participated in clinical trials and that they inform their PF/IPF patients on ongoing clinical trials. Half of patients (53%) were aware of ongoing clinical trials, 31% had been asked to participate and 25% had actually participated in a clinical trial. Patient registries for IPF and PF existed in 75% and 48% of centers respectively. A quarter of patients (27%) declared that they contributed to the collection of registry data.

General recommendations

In general, 61% of patients described that their experience with the healthcare system as either good or excellent. Both patients and HCPs were asked about suggestions to improve the patient experiences at different stages of the disease. Based on the answers on this question five recommendations by the expert panel were proposed **(Figure 5)**.

DISCUSSION

This is the first study investigating unmet needs of patients with pulmonary fibrosis in a Europe-wide survey. Despite recent advances in PF/IPF care and research, the unmet needs and gaps in care revealed in this study are in line with previous research (**Table 1**).

Referral pathways and access to specialist care

One of the major unmet needs in PF/IPF care is a timely and accurate diagnosis (13, 15, 19, 24, 25). In the current study, a significant number of patients received another diagnosis prior to being diagnosed with of PF/IPF; time from initial diagnosis to diagnosis of PF/IPF was often >1 year. This is in agreement with previous studies, which showed that many patients receive at least one misdiagnosis, consult more than three physicians before receiving a final diagnosis, and have a delay in diagnosis of >1 year (15, 25, 26, 34). Although the current study shows less delay than some previous reports, one out of five patients in this study still had to wait >1 year for referral to an ILD specialist center. It is of utmost importance to reduce delays in diagnosis and referral, since previous research indicated that a lengthy diagnostic trajectory can have an adverse effect on quality of life, and that delayed access to tertiary referral centers is associated with a higher risk of death in IPF (13, 25, 35). In fact, access to ILD specialist centers may increase the perceived quality of care (15, 26). Access to MDTs appears to have increased in recent years. In contrast to the European IPF patient charter in 2016 (11), almost all HCPs in the current study reported access to an MDT, while the composition of the MDT still widely varies.

Figure 5. General recommendations to improve the IPF/PF patient journey

One of the reasons for delayed diagnosis is the lack of knowledge regarding PF/IPF among the general public, GPs and physicians in community hospitals (5, 11, 15, 26). Improving knowledge about IPF, through education and awareness campaigns, could facilitate earlier diagnosis and referral (11, 13, 34). A prior study suggested to develop symptom-based algorithms for GPs, to help identify which patients should be referred for further analysis (19).

Pharmacological treatment

Although antifibrotic medication can be prescribed in all participating countries in this study, timely access to treatment has been highlighted as an issue by both patients and healthcare professionals. A recent study found that up to 40% of patients with a confirmed IPF diagnosis do not receive treatment with antifibrotic medication (36). Barriers

to pharmacological treatment include delayed access to specialist care and reimbursement restrictions (36). Moreover, a watch-and-wait approach is sometimes preferred in patients with mild or relatively stable disease, despite the fact that the importance of early treatment initiation has been emphasized in recent years (5, 36-39). Our results show that reimbursement restrictions continue to be an important cause of delayed access to antifibrotic treatment. Treatment delays vary due to different prescription criteria. To ensure equal access to antifibrotic medication across Europe, fewer reimbursement restrictions and uniform criteria acknowledging the patient needs reported in this statement are imperative.

Non-pharmacological treatment

Non-pharmacological treatment options, such as pulmonary rehabilitation, oxygen therapy, psychological support, and lung transplantation are a vital part of holistic care for patients with PF/IPF (2, 40, 41). Previous studies demonstrated that non-pharmacological treatment options are not equally available for patients in different European countries (11, 19). In the current survey, the vast majority of HCPs indicated that they could refer patients for lung transplantation and pulmonary rehabilitation, as well as being able to prescribe oxygen therapy. In contrast, fewer than half of the patients reported that they had access to pulmonary rehabilitation. This discrepancy could be due to the fact that pulmonary rehabilitation is often not fully reimbursed, that many patients are unaware that pulmonary rehabilitation programs exist for PF/IPF, and that patients often have to travel long distances for pulmonary rehabilitation (11). The need for better emotional and psychological support for patients and caregivers has been frequently reported and is underlined by the findings from our study (11, 13, 14, 17-19, 21, 23, 28, 33). Nevertheless, reimbursement and access to psychological support for PF/IPF patients remains restricted. If referral to a psychologist is not possible, other options for emotional support should be explored. Previous work shows that many patients also benefit from psychological and emotional support through peer support groups, pulmonary rehabilitation, and ILD specialist nurses (11, 13, 14, 23, 30, 42, 43). Strikingly, only half of the surveyed patients in this study had access to ILD specialist nurses, demonstrating that more specialist nurses should be trained.

Access to palliative care

As of yet, there are no (international) guidelines on palliative care in PF/IPF. This leads to underuse of and varying access to palliative care across Europe, which is also influenced by differences in local resources, cultural and religious beliefs, and misconceptions about the meaning of palliative care (2, 11). It is important to acknowledge that palliative care comprises more than just end-of-life care alone, and aims to improve quality of life during the whole disease course (2, 44, 45). Still, our results indicate that many HCPs

in Europe start palliative care in more severe stages of PF/IPF. The majority of HCPs in this study stated that they discuss end-of-life care with all patients. However, the optimal timing of end-of-life discussions and referral to palliative care services remains difficult in PF/IPF (16, 19, 29) and depends on various factors including culture, religion etc. Prior reports suggest that early palliative care can potentially reduce symptom burden for patients with IPF, but needs to be tailored to the preferences of individual patients (2, 28). Hence, palliative care should be an integral part of comprehensive care for patients with PF/IPF (2).

Communication and education

Education plays an important role in the management of PF/IPF. To enable shared-decision making and enhance communication, patients must be well informed about their disease and its prospects (40, 46). While our results show that three quarters of patients receive a treatment plan after their diagnosis, only a third of patients are actually involved in developing this plan. Possible reasons are the lack of time to discuss treatment plans with patients and the fact that patients need to be better educated to become more involved (46). Adequate information about PF/IPF, more education, and continuous counselling were among the frequently reported suggestions for improvement of the care pathway in the patient survey. The need for more information is in agreement with findings from previous surveys and interviews (11, 15-18, 23, 24, 27-29). Whereas two-thirds of centers in the current study offer education for patients, only a minority of patients attended any educational activity. This suggests that greater awareness of the educational activities amongst patients may be required, or that some patients might prefer to receive written information and/or use online resources (24, 31, 46). To improve experiences for patients and caregivers, educational material about PF/IPF needs to be easily accessible, understandable, updated frequently and adapted to individual patient's needs (14, 23, 24, 46).

Involvement in research

Results of this study highlight that patients should be better informed about clinical trials and patient registries. Only half of patients were aware of ongoing clinical trials and only a quarter actually participated in a trial. Previous research suggested that many patients wish to be informed about possibilities to participate in clinical trials and that patients treated in specialist centers were more likely to be participating in a clinical trial (13, 15, 27, 37). Moreover, one study reported that patients who participated in a clinical trial were more hopeful regarding treatment than other patients (13). Efforts should therefore be made to inform all PF/IPF patients about clinical trials, and to refer patients to specialist centers for participation in trials. Many countries have local or national registries for PF/IPF; however, only a quarter of patients indicated that they contribute data

to a registry. Improved collaboration with patients and between countries is needed to collect data and establish a multinational registry. Such a registry will not only enhance understanding of disease behavior, but may also provide insights to improve care and outcomes for patients with PF/IPF(47).

Limitations

This study has several limitations**.** First, the results are only representative of the situation in 14 EU-IPPF member countries; newer EU Member States, in particular, have been under-represented. Moreover, the HCP survey was distributed through the ERN-LUNG network. This resulted in a high number of responses from physicians in ILD specialist centers, representing an important bias. Similarly, the patients who participated in the survey may have better access to information and specialist care, because they were recruited *via* support groups. There may also have been a bias towards less impaired patients amongst the respondents, which makes it difficult to compare answers of HCPs and patients. Further limitations of this online survey distributed *via* patient member organizations were a self-reported diagnosis and an unknown response rate.

CONCLUSIONS

This survey and literature search offers important insights into the current unmet needs of PF/IPF patients in Europe and should be considered for healthcare decisions. Recommendations set out in this statement could provide a useful tool to healthcare providers and policy makers to improve the patient journey and overall care of these rare diseases. Better international collaboration between clinicians, researchers, patients, caregivers, industry partners, and governments should be established to solve unmet needs, improve outcomes, and develop evidence-based multidisciplinary care for PF/IPF patients.

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SUPPLEMENT

Supplementary file S1

Your experience of living with Idiopathic Pulmonary Fibrosis (IPF) or another form of Pulmonary Fibrosis (PF)

Thank you for agreeing to take part in this survey about living with IPF or PF. You can fill in the questionnaire yourself of your carer can do it for you. It will take about half an hour. The information you share will give EU-IPFF, the patients' European federation, a better understanding of the needs of IPF and PF patients and may help to improve services in the future. Your name will not be requested to complete this questionnaire.

English plain review
Translated in DE/FR/EL/ES/BG/IT/NL/PT/PL
Time to complete: 25 minutes
Online distribution
Consent form requested before questionnaire is completed

General information

1) I am a patient / carer. (If you are a carer or family member, please answer on behalf of the person you care for).
2) I have IPF or another type of PF
3) I am male / female
4) My age:
5) Country I live in

Information about your PF/IPF

6) Is there a history of IPF or PF in your family?
7) In which year were you diagnosed with IPF?
8) Some countries have a national registry which collects health information from patients and uses it to improve knowledge and treatment of the disease.
9) Do you know if there is an IPF (not PF) registry in your country? Yes / No / I don't know
10) Do you know if there is a registry for PF (not IPF) in your country? Yes / No / I don't know
11) Do you contribute to the collection of registry data? Yes / No

Your experience of interacting with doctors

12) Did your doctor give you information on your disease when you were diagnosed?

13) If yes, what type of information did you receive?
14) Did you understand the information you were given?
15) Did you have enough time to discuss your diagnosis with your doctor? Yes/ No
16) How would you describe your experience of discussing your diagnosis with your doctor? (scale 1 to 5: 1= strong positive; 2=positive; 3=neutral; 4=negative; 5=strong negative)
17) Besides your family doctor, did you have access to any other healthcare professionals to discuss your diagnosis? Yes / no
18) If yes, please say which of the following you spoke with :
    - respiratory (lung) doctor (also known as a pulmonologist)
    - radiologist
    - specialist respiratory nurse
    - physiotherapist
    - rheumatologist
    - psychologist
    - thoracic (chest) surgeon
    - other (please specify)
19) Did you get training or educational activities from your treating centre? Yes / No
20) How could health care staff work more effectively with you and your carers? Please give us your suggestions and recommendations

Genetic screening

21) Did you have a genetic test? This is a test to examine your genes to find out how likely you are to get IPF or PF. Yes / No
22) If yes, did you get enough information about this test?

Your referral

23) How much time passed before your family doctor referred you to a respiratory doctor?
    - Less than a month
    - Between one and two months
    - Between two and six months
    - Between six and twelve months
    - More than a year
24) How much time passed between the first consultation with your family doctor and your referral to a specialist lung centre?
    - Less than a month
    - Between one and two months
    - Between two and six months

- Between six and twelve months
- More than a year

25) Did you get a wrong diagnosis before you were diagnosed with IPF or PF? Yes / No
26) If yes, how much time passed between your first (wrong) diagnosis and your diagnosis with IPF or PF?
    - Less than a month
    - Between one and two months
    - Between two and six months
    - Between six and twelve months
    - More than a year
27) In your country, how it easy is it to get a referral to a specialist lung centre? Please choose from the following scale where 1=very easy; 2= easy; 3=neither easy nor difficult; 4=difficult; 5=very difficult.
28) How far is your centre from your home?
    - Less than 10 km (about 6 miles)
    - Between 10 km and 30 km (between about 6 miles and 18 miles)
    - Between 30 km and 50 km (between about 18 miles and 31 miles)
    - Between 50 km 100 km (between about 31 miles and 62 miles)
    - More than 100 km (more than 62 miles)
29) Do you have access to a multidisciplinary team? Yes / No
30) Do you have access to a specialist lung nurse? Yes / No
31) Which healthcare professional has been monitoring your IPF or PF since your diagnosis?
    - Respiratory doctor (pulmonologist) from the specialist lung centre
    - Family doctor
    - Specialist nurse
    - Physiotherapist
    - Palliative care doctor or nurse
    - Other, please specify:
32) On average, how often do you meet your multidisciplinary team to discuss your condition?
    - Once a month
    - Every 3 months
    - Every 6 months
    - Every 12 months
    - Other, please specify:
33) Following your diagnosis, did you get a treatment plan for your disease? Yes / No
34) Was your treatment plan explained clearly to you? Yes / No If no, please tell us more.

35) Were you involved at any stage in the development of your treatment plan? Yes / No

If yes, how were you involved (please describe)?

36) Were you given different options for your treatment? Yes / No

If yes, please say which options.

37) Do you feel you got enough support from your doctors and healthcare team? Yes / No

If no, please tell us what support you would have liked to get.

Access to medicines (only for IPF patients)

38) Are you being treated with medicines for your IPF? Yes / No

If no, were you treated with any medicines in the past? Yes / No

39) In what year did you start taking medicines for your IPF?

40) Did you have to change your medicines at any point? Yes / No

If yes, please say why you had to change.

41) Overall, how long did you have to wait from diagnosis to receiving treatment?
    - Less than a month
    - Between one and two months
    - Between two and six months
    - Between six and twelve months
    - More than a year

42) In case you started late taking medicines, what are the main reasons?
    - Late diagnosis
    - Not referred in time to a specialist lung centre
    - Treatment not available for sale in your country
    - Treatment is not paid for
    - Treatment is only partially paid for
    - Treatment is not reimbursed for your type of IPF
    - Treatment is not prescribed by your centre
    - Other, please explain

43) Are you fully (100%) covered for the costs of medicines by health insurance or the health service in your country? Yes / No

44) If no, how much do you have to pay for medicines out of your own pocket each year?
    - Less than €200
    - Between €200 and €500
    - Between €500 and €1,000
    - Between €1,000 and €2,000
    - Between €2,000 and €5,000

- More than €5,000

45) In your opinion, what would help increase the use of medicines for treating IPF? Please choose all that apply.
    - Making the prescription of medicines easier for doctors
    - Encouraging family doctors to refer more patients to specialist lung centres
    - Making the procedures for reimbursement easier
    - Cheaper medicines
    - Other, please explain

Access to other treatments

46) Has your doctor prescribed portable oxygen therapy for you? Yes / No
47) Is the cost of portable oxygen paid for by your health insurance or the health service in your country?
    - Yes, fully (100%)
    - Yes, in part
    - No
48) What do you have to do to be fully or partially reimbursed? Please describe.
49) Has your doctor prescribed oxygen therapy for you to use at home? Yes / No
50) Is the cost of oxygen at home paid for by your health insurance or the health service in your country?
    - Yes, fully (100%)
    - Yes, in part
    - No
51) What do you have to do to be fully or partially reimbursed for home oxygen therapy? Please describe.
52) What are the main difficulties you face in accessing portable and home oxygen therapy? Please choose all that apply.
    - High cost
    - No availability of liquid oxygen portables
    - No availability of portable oxygen concentrators
    - Oxygen therapy was not prescribed
    - Other, please specify
53) Do you have access to a pulmonary (lung) rehabilitation programme?
    - Yes, out-patient
    - Yes, in-patient
    - No, none at all
54) Is the cost of the pulmonary rehabilitation programme paid for by your health insurance or the health service in your country?
    - Yes, fully (100%)

- Yes, in part
- No

55) What do you have to do to be fully or partially reimbursed for pulmonary rehabilitation? Please describe.
56) Do you have access to palliative care? Yes / No
57) Is palliative care paid for by your health insurance or the health service in your country?
    - Yes, fully (100%)
    - Yes, in part
    - No
58) What do you have to do to be fully or partially reimbursed for palliative care? Please describe.
59) Are you and your family involved in decisions about palliative care?
60) Who supported you during your treatment (whatever type of treatment you had)? Please choose all that apply.
    - respiratory doctor (pulmonologist)
    - radiologist
    - pathologist
    - specialist nurse
    - multidisciplinary team coordinator
    - physiotherapist
    - rheumatologist
    - immunologist
    - thoracic surgeon
    - interventional pulmonologist (who uses minimally invasive endoscopic techniques)
    - psychologist
    - patient organisation or peer group
    - other, please indicate
61) Do you have access to a patient organisation to support you during your treatment? Yes / No
62) How would you describe your overall experience with the healthcare system in relation to your IPF or PF? Please choose from the following scale where 1= excellent; 2=good; 3=neither good nor bad; 4=bad; 5=very bad.

Research

63) Are you aware of any ongoing clinical trials? Yes / No
64) Were you ever asked to be involved in a clinical trial? Yes / No
65) Have you ever taken part in a clinical trial? Yes / No

General suggestions

66) Please tell us how you feel the experience of patients and carers could be improved at different stages of the disease.

Thank you for completing this survey

SUPPLEMENTARY FILE S2

Questionnaire for healthcare professionals on the patient's journey through Idiopathic Pulmonary Fibrosis (IPF) and other forms of Pulmonary Fibrosis (PF)

Time to complete: 30 minutes

English only

Online distribution

1) General information
2) Indicators
3) I am a doctor / nurse
4) Your organisation's name and location
5) Your country of residence
6) Is your organisation a recognised Centre of Expertise specialising in interstitial lung diseases (ILDs)? Yes / No
7) Is your organisation a member of ERN-LUNG? Yes / No

Epidemiological data

8) How many patients with Idiopathic Pulmonary Fibrosis (IPF) are managed in your centre each year?
9) How many patients with Pulmonary Fibrosis (PF), but excluding IPF, are managed in your centre each year?
10) How many new patients were diagnosed with IPF within the past 12 months at your centre?
11) How many new patients have been diagnosed with other forms of PF within the past 12 months at your centre?
12) Is there a national registry for IPF in your country? Yes / No
13) Is there a registry for IPF in your centre? Yes / No
14) Is there a national registry for other forms of PF in your country? Yes / No
15) Is there a registry for PF in your centre? Yes / No
16) Do you contribute to the collection of registry data? Yes / No

Interactions between patients and doctors

17) Have you received training to effectively communicate information on diagnosis and treatment to IPF and PF patients and carers? Yes / No
18) If yes, please specify the training.
19) Do you feel equipped to give clear and easy-to-understand information to IPF and PF patients and carers? (Please choose from the following scale where 1=strongly agree; 2=agree; 3=neither agree nor disagree; 4=disagree; 5=strongly disagree)

20) Do you feel you have enough time to discuss their diagnosis with patients? (Please choose from the following scale where 1=strongly agree; 2=agree; 3=neither agree nor disagree; 4=disagree; 5=strongly disagree)
21) Do you feel you have enough time to discuss their treatment with patients? (Please choose from the following scale where 1=strongly agree; 2=agree; 3=neither agree nor disagree; 4=disagree; 5=strongly disagree)
22) Does your centre offer training or educational activities to IPF and PF patients? Yes / No. If yes, please specify.
23) How do you think interactions with patients could be improved?

Screening

24) Does your centre have access to genetic testing to diagnose IPF and PF? Yes / No
25) If yes, is the test provided to IPF and PF patients in your centre or in another centre?
    - In my centre
    - In another centre
26) Does your centre have access to genetic counselling? Yes / No. Please specify for both answers. If yes, is it provided to IPF and PF patients?

Referral

27) In your country, how easy is it to identify a centre of expertise specialising in ILDs for IPF and PF patients? (Please choose from the following scale where 1=very easy; 2=easy; 3=neither easy nor difficult; 4=difficult; 5=very difficult).
28) Do IPF and PF patients have access to a multidisciplinary? Yes / No
29) Do IPF and PF patients have access to a specialist ILD nurse? Yes / No
30) Which of the following professionals is part of the multidisciplinary team or supportive interdisciplinary team? (Please tick all that apply)
    - consultant respiratory physician or pulmonologist
    - consultant thoracic radiologist
    - consultant pathologist
    - interstitial lung disease specialist nurse
    - multidisciplinary team coordinator
    - physiotherapist
    - rheumatologist
    - immunologist
    - thoracic surgeon
    - pharmacist
    - other (please describe)
31) On average, how often do you or the multidisciplinary team review an IPF or PF patient's case?

- Once a week
- Twice a month
- Once a month
- Every 3 months
- Every 6 months
- Every 12 months
- Other (please specify)

32) On average, how often do you or the multidisciplinary team invite the IPF or PF patient to the clinic to discuss their case?
    - Once a month
    - Every 3 months
    - Every 6 months
    - Every 12 months
    - Other (please specify)
33) Are IPF and PF patients involved in developing their treatment plan? Yes / No If yes, how?
34) Do you provide different treatment and support options to your IPF and PF patients? Yes / No
35) How are IPF and PF patients referred to you? Please indicate the top-two options that apply:
    - General practitioner
    - Pulmonologist
    - Other pulmonologist
    - Self-referral
    - Other (please specify)

Access to pharmacological treatment (specific to IPF)

36) The following treatments for IPF are approved by the European Medicines Agency (EMA). Please indicate if they are prescribed in your country
    - Active substance Available Not available
    - Pirfenidone
    - Nintedanib
37) Please indicate if the following EMA-approved treatments for IPF are available for prescription in your centre
    - Active substance Available Not available
    - Pirfenidone
    - Nintedanib
38) Please indicate if the following EMA-approved treatments for IPF are fully (100%) reimbursed or not in your country

- Pirfenidone Yes / No
- Nintedanib Yes / No

39) If no, please elaborate.

40) Are there any inclusion or exclusion criteria when administering the following treatments:
    - Pirfenidone Yes / No
    - Nintedanib Yes / No

41) If yes, please specify.

42) Are there any barriers to prescribing EMA-approved IPF treatments in your centre? Please select all that apply
    - Severity of disease (mild, moderate, severe)
    - Disease progress
    - Patient clinical status
    - Age
    - Other, please explain

43) What is the average time from suspected diagnosis to starting treatment with Nintedanib or Pirfenidone?
    - Less than a month
    - Between 1 month and 6 months
    - Between 6 months and 1 year
    - More than a year

44) If IPF patients in your centre experience delays in accessing treatment, what are the main reasons for the delay? Please select all that apply
    - Wrong recognition of symptoms and signs
    - Time from suspected diagnosis to confirmed diagnosis
    - Delays in referral to a centre specialising in ILDs
    - Treatment not available for sale in your country
    - Treatment not reimbursed
    - Treatment only partially reimbursed
    - Treatment not reimbursed for all types of IPF or PF
    - Treatment not prescribed by your centre
    - Other, please explain

45) Do you have patients who might benefit from access to pharmacological treatment but who are not eligible?
    - No
    - Less than 10%
    - Less than 30%
    - More than 30%

46) In your opinion, what would improve access to treatment for IPF patients?

- No limitations for prescription
- Reimbursement
- Timely referral to specialist ILD centre
- Better pricing policies
- Other, please explain

Access to non-pharmacological treatment

47) Does your centre prescribe ambulatory oxygen for IPF and PF patients? Yes / No
48) Is ambulatory oxygen reimbursed? Yes / No / Partially
49) Can your centre prescribe oxygen for IPF and PF patients to use at home? Yes / No
50) Is oxygen for use at home reimbursed? Yes / No / Partially
51) Can your centre prescribe pulmonary rehabilitation for IPF and PF patients? Yes / No
52) Is pulmonary rehabilitation reimbursed? Yes / No / Partially
53) Can your centre prescribe medication for palliative care or symptom relief for IPF and PF patients? Yes / No
54) When do you start palliative care or symptom relief for IPF and PF patients? Please specify according to the stage of progression of the disease.
55) Is end-of-life care discussed with IPF and PF patients and their families? Yes / No
56) In your country, what are the eligibility criteria for a lung transplant? Please select all the criteria that apply.
    - Age, please specify
    - General health condition
    - Likelihood of developing risks associated with transplant (for example, infections, rejection of new lung)
    - Severity of IPF or PF
    - Progression of IPF or PF
    - Availability of organs
    - Surgical procedure not available
    - Other, please specify
57) In patients eligible for lung transplantation, do you refer these patients for a lung transplant? Yes / No
58) Do patients in your centre have access to psychological support such as counselling? Yes / No.
59) If yes, please specify if psychological support is reimbursed: Yes / No / Partially
60) Do patients in your centre have access to educational materials on IPF and PF?

Research

61) Are you aware of current IPF and PF clinical trials? Yes / No If yes, please state which trials.
62) Do you tell IPF and PF patients that they could take part in clinical trials? Yes / No
63) If yes, please specify if:
    - your centre participates in trials
    - your centre refers patients for trials to another centre
    - you refer to the patient association

General recommendations

64) Please share your suggestions for improving the patient's experience of the IPF or PF journey.

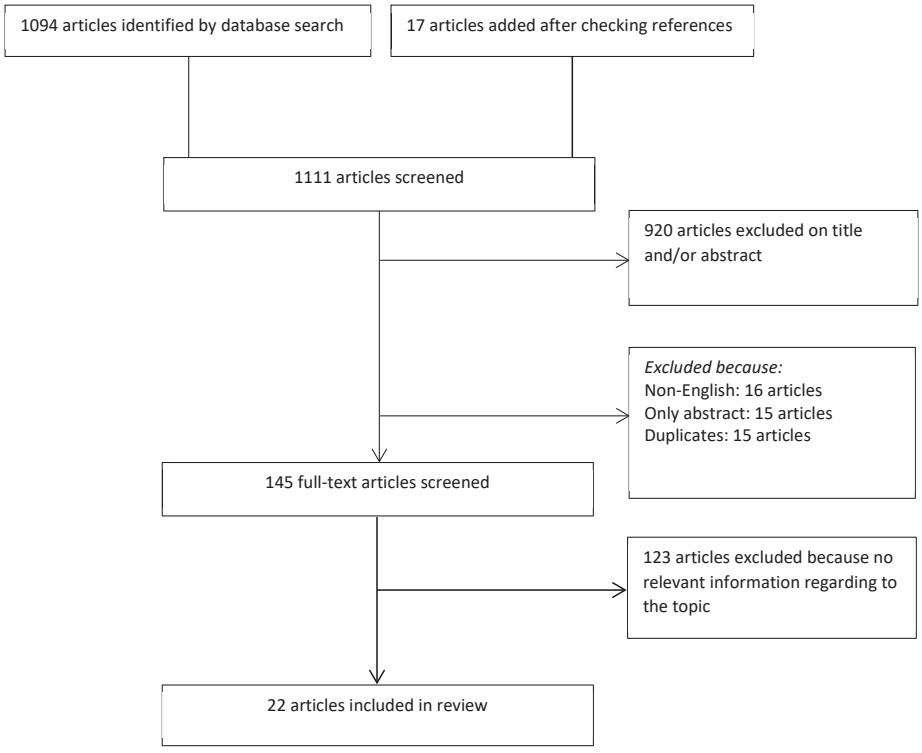
Thank you for completing this survey.

SUPPLEMENTARY FILE S3

Literature search

A systematic literature search was performed using PubMed and Embase for studies that evaluated the care pathway for pulmonary fibrosis patients, published between January 2010 and March 2018. For this search the following (MeSH) terms were used: ("idiopathic pulmonary fibrosis" OR "pulmonary fibrosis" OR "interstitial lung disease") AND ("care pathway" OR care OR barrier OR need OR unmet need). Additionally, references of included articles were checked to identify other potentially relevant articles.

This search retrieved 1094 articles and 17 articles were added after checking references. These 1111 articles were screened on title and abstract by three authors (DB, CM and LP). 145 potentially relevant articles have been fully assessed by the reviewers, and in case of disagreement the article has been discussed until consensus was reached. Non-English articles and abstracts were excluded. After exclusion of studies not related to the topic, 22 articles were included (see flowchart in figure 1).



Supplementary file S3: Literature search flow diagram