The journey to diagnosis in patients with pulmonary fibrosis

An EU-IPFF report on patients' experiences throughout the journey to diagnosis in Europe

EU-IPFF
EUROPEAN IDIOPATHIC PULMONARY FIBROSIS & RELATED DISORDERS FEDERATION

Action for Pulmonary Fibrosis

Galápagos
Pioneering for patients

About the European Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF)

The EU-IPFF brings together 21 European national patient associations from 15 European countries. Its mission is to serve as the trusted resource for the IPF community by raising awareness, providing disease education, advancing care and funding research. The EU-IPFF collaborates with physicians, medical organisations, people with pulmonary fibrosis, caregivers and policy-makers throughout Europe.

For further information, please visit **www.eu-ipff.org**.

About Galapagos

Galapagos is a clinical-stage biotechnology company, specialised in the discovery and development of small-molecule medicines with novel modes of action.

Our ambition is to become a leading global biopharmaceutical company, focused on the discovery, development and commercialisation of innovative medicines that will improve people's lives.

Foreword from the EU-IPFF President

Steve Jones
EU-IPFF President
November 2020

Imagine that every breath you take is an effort. This is what it feels like for over 250,000[1] people in Europe living with the devastating lung disease, pulmonary fibrosis.

Pulmonary fibrosis causes scarring or stiffening of the lungs, making it hard for oxygen to pass from the lungs into the body.[2] Early symptoms of the disease include a persistent cough and breathlessness.[3] Over time, people find it increasingly difficult to breathe,[3] even becoming breathless when eating or talking. Everyday tasks become increasingly hard. In time, patients become dependent on oxygen and ultimately die from respiratory failure.[3]

I was one of these patients. I lived with idiopathic pulmonary fibrosis (IPF), a type of pulmonary fibrosis, for 8 years and would have died had I not received a lung transplant in 2016.

Since the median survival of patients with IPF is only 2–5 years from diagnosis, without treatment,[4] it is important that we are diagnosed swiftly and start treatment promptly. Sadly, we know from talking to patients across Europe that diagnosis with pulmonary fibrosis is often slow and that many patients are initially misdiagnosed. Given that IPF and potentially other types of progressive pulmonary fibrosis have a poorer prognosis than cancer,[5] any delay in diagnosis is unacceptable.

The EU-IPFF undertook this study to improve our understanding of the path to diagnosis for patients with pulmonary fibrosis.

We hope to identify the challenges patients and their families face and to generate evidence to use in advocating for early diagnosis.

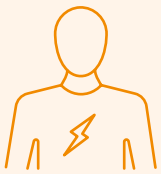
The EU-IPFF's mission is to improve the treatment and care of patients with pulmonary fibrosis in Europe. We hope that this study will also contribute to this goal.

We are extremely grateful to Galapagos NV for its active support in undertaking this study.

Rationale for this survey

There is an urgent need to understand the journey to diagnosis in patients with pulmonary fibrosis and identify ways in which it can be improved.

Given this, the aims of the survey were to:



Understand the experience of patients with pulmonary fibrosis, from first symptoms at home to receiving a diagnosis at a hospital

Identify any gaps in pulmonary fibrosis care prior to and following diagnosis, including timings, management approaches and taking account of patients' feelings during their journey

Understand how the process of diagnosis could be improved across Europe

The survey was undertaken by the EU-IPFF, supported by Galapagos.*

Key findings

Many patients across Europe experience a long and difficult journey to diagnosis with pulmonary fibrosis. Over 40% of patients wait longer than 12 months to be diagnosed

Delays occur at all stages along the pathway. Patients may be slow to consult their general practitioner (GP)/primary care physician (PCP) once they have symptoms. GPs/PCPs may not refer patients to pulmonary specialists quickly enough, and there can be delays in waiting for a hospital appointment and conducting the necessary tests to achieve a correct diagnosis

A quicker diagnosis would mean that patients can start treatment earlier and would reduce the worry and anxiety patients feel when waiting for a diagnosis

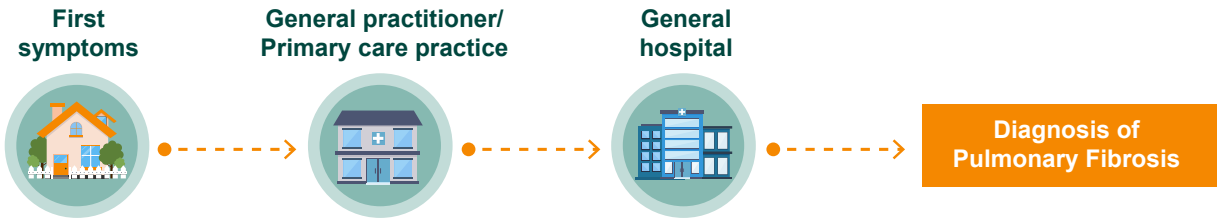
Methodology

A survey was designed to collect quantitative and qualitative data from patients diagnosed with pulmonary fibrosis across Europe. Patients completed a structured questionnaire online, which included both closed and open-ended questions. The questionnaire was translated into eights languages by a certified translation agency and disseminated by the EU-IPFF through its member organisations in Europe. Responses were collected from countries in which there was an EU-IPFF member organisation.

The questionnaire focused on the following stages of the patient journey:

- Time at home before seeking medical attention
- Time being cared for by a general practitioner (GP)/primary care physician (PCP)
- Time at a hospital and/or specialist centre
- Overall time to diagnosis and feelings throughout the patient's journey

Fig. 1: Journey to diagnosis for patients with pulmonary fibrosis.



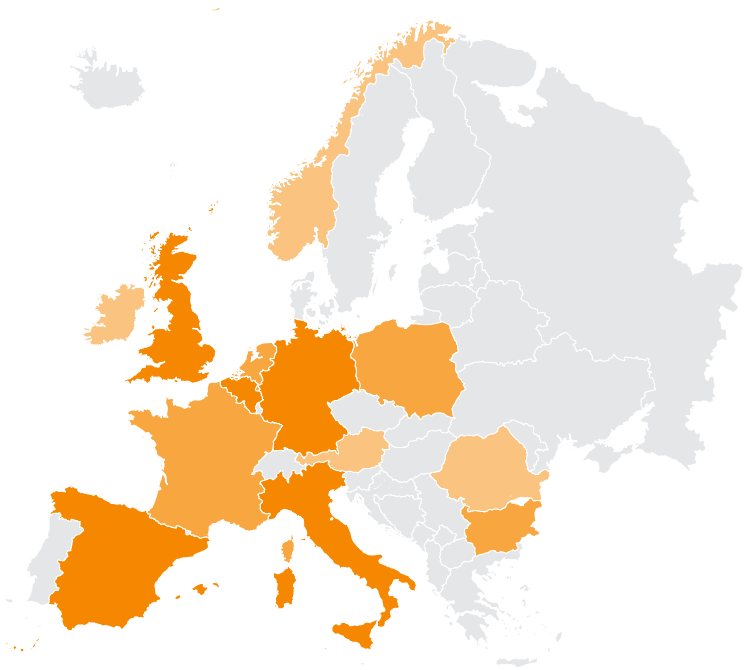
The results of the survey were compiled and analysed to generate insights into the patient journey (Fig. 1). Each of these stages will be examined in turn.

The survey was managed by the communications consultancy, emotive.

Europe at a glance

The survey was completed by a total of 273 patients from 13 European countries (Fig. 2). Almost 90% of the patients came from six countries: Spain, Belgium, the United Kingdom, Italy and Germany. As shown in Fig. 3, the majority of patients (78%) reported being diagnosed with IPF, followed by sarcoidosis (10%), autoimmune-related pulmonary fibrosis (4%) and chronic hypersensitivity pneumonitis (just under 1%).

Fig. 2: Countries with participants in the survey.



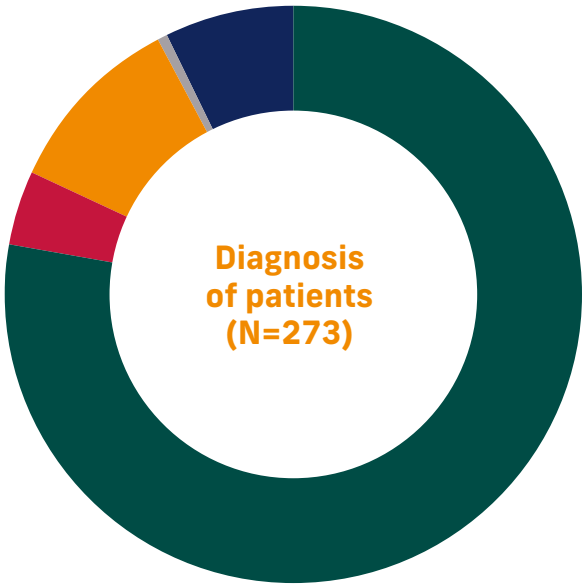
- Spain **59** (21.6%)
- Belgium **55** (20.1%)
- UK **50** (18.3%)
- Italy **47** (17.2%)
- Germany **29** (10.6%)

88%

- The Netherlands **9** (3.3%)
- Bulgaria **7** (2.6%)
- France **5** (1.8%)
- Poland **5** (1.8%)
- Austria **4** (1.5%)
- Romania **1** (0.37%)
- Ireland **1** (0.37%)
- Norway **1** (0.37%)

12%

Fig. 3: Diagnoses of patients participating in the survey.



78.0%
IPF

4.0%
Autoimmune-related pulmonary fibrosis

10.3%
Sarcoidosis

0.7%
Chronic hypersensitivity pneumonitis

7.0%
Other/no response

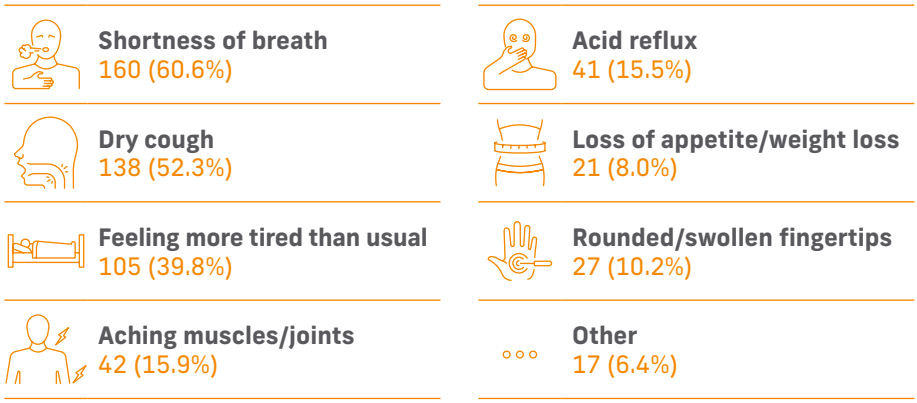
Patient journey: at home

Call to action:

Raise awareness of pulmonary fibrosis with the general public and encourage people to visit their GP/PCP if they feel breathless, unusually fatigued or have a persistent cough.

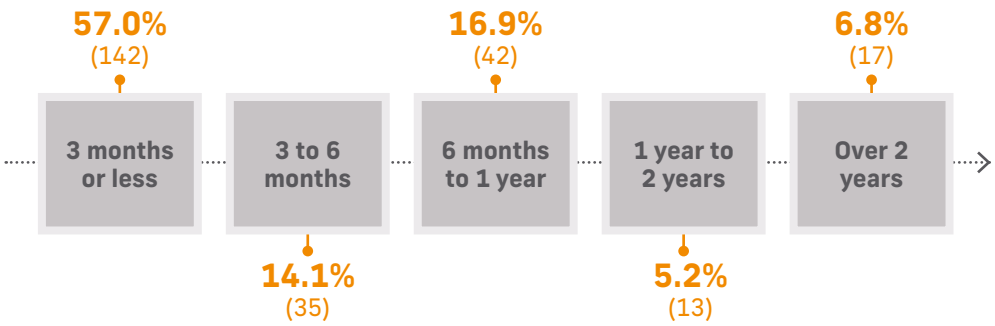
The patient journey starts when somebody first experiences a symptom of the disease. Most respondents to the survey reported first going to the GP/PCP because they experienced shortness of breath, a dry cough or fatigue (Fig. 4). These responses are consistent with those commonly reported in previous studies.[6,7]

Fig. 4: Symptoms of patients attending a GP/PCP for the first time.



Almost 60% of patients saw their GP/PCP within 3 months of symptoms starting; however, 29% waited more than 6 months to make an appointment with their doctor (Fig. 5). In the majority of cases, patients delayed either because they were not concerned about their symptoms or thought they were related to age. The majority of patients finally decided to visit their doctor when their symptoms got worse and/or because they were spurred on by family members or friends.

Fig. 5: Patient waiting time before consulting a GP/PCP.



29% of patients waited 6 months or more before making an initial doctor's appointment

n=249 (the number of patients who were able to recall time to making an appointment).

"I was becoming less and less able to undertake physical activities such as gardening and rambling, so I went to see my GP."

–A patient from the UK

Patient journey: at general practice/primary care practice

Call to action:

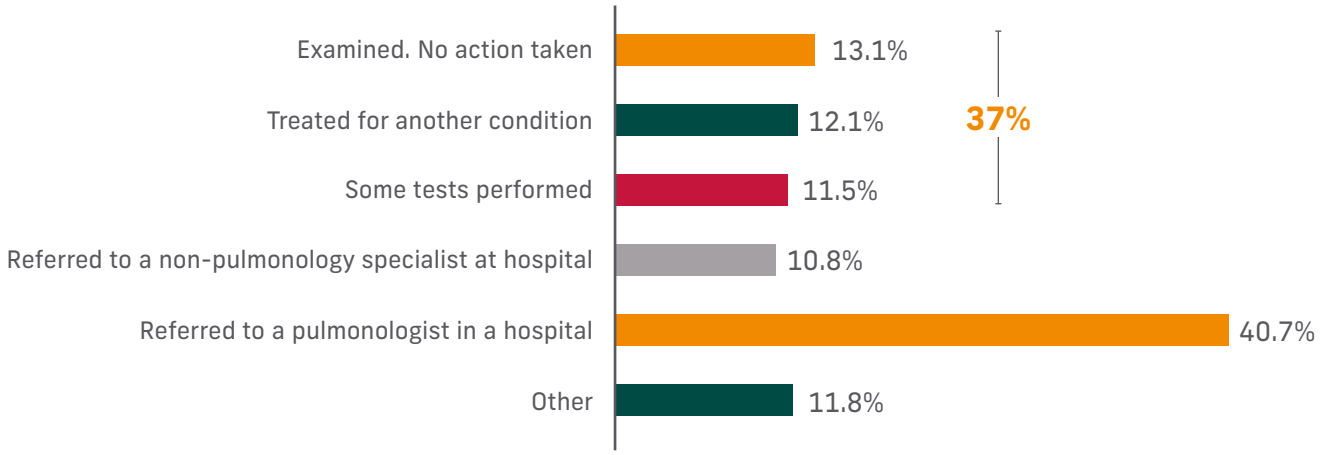
Make GPs/PCPs more aware of the signs and symptoms of pulmonary fibrosis and cut hospital waiting times for patients with suspected pulmonary fibrosis.

Although pulmonary fibrosis accounts for an increasing number of deaths in Europe, the disease is not widely recognised by GPs/PCPs.[8] The key symptoms of breathlessness and cough are commonplace in general practice. As a result, early diagnosis can often be challenging and symptoms may be attributed to other respiratory conditions, such as asthma or chronic obstructive pulmonary disease.[9]

Patients reported that the approaches adopted by doctors during the initial consultation varied widely (Fig. 6). Almost half of patients (41%) were referred immediately by their GP/PCP to a pulmonologist, whereas 37% were treated or given further tests at the GP/PCP practice. As reported in other studies,[10] a minority of patients (12%) were diagnosed and treated for another condition, illustrating the challenges GPs/PCPs face in diagnosing pulmonary fibrosis.

The majority of patients saw a specialist within 3 months of being referred by their GP/PCP; however, it took over 6 months for 10% of patients (Fig. 7).

Fig. 6: Actions taken by GP/PCP at a patient's first visit.



These percentages were calculated as a percentage of the total responses (305) rather than as a percentage of patients in the survey. Patients were able to select more than one answer.

"The GP thought it might be severe pneumonia and put me on antibiotics for a month."

–A patient from Belgium

"I went to see a doctor who diagnosed me with bronchitis."

–A patient from Germany

Fig. 7: Time taken to see a specialist after referral by GP/PCP.

n=264 (the number of patients who were able to recall time taken to see a specialist).

After the onset of breathlessness, delayed referral to a hospital is associated with an increase in mortality, irrespective of disease severity.[11] This underlines the importance of being referred to a specialist without delay.

Over 18% of patients visited their doctor more than four times before being referred to a hospital

At this stage of their journey, patients became increasingly anxious about their health. Many patients were aware that they may have a serious disease and were worried that their symptoms would get worse, which caused them to feel helpless. Patients suggested that improving GP/PCP awareness, earlier testing and better information could help to reduce the time taken to achieve a diagnosis.

"I should have gone to see the doctor sooner and got a second opinion more quickly."

–A patient from Germany

"My doctor did not refer me to the pulmonologist because my chest X-ray and spirometry results were normal."

–A patient from Italy

Patient journey: at the hospital

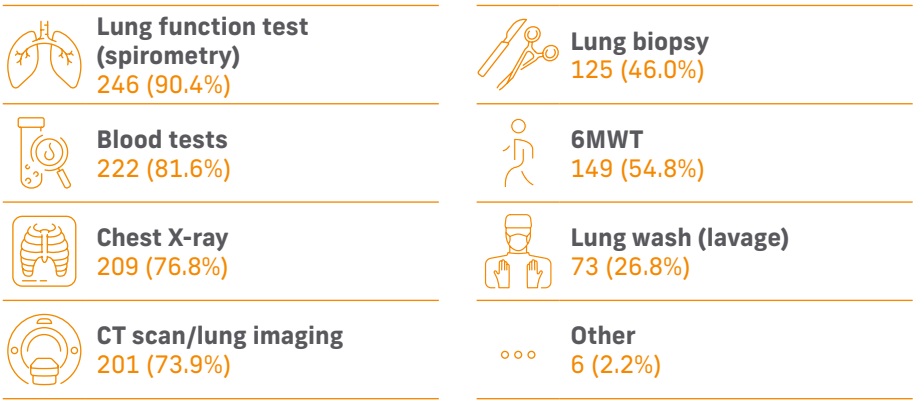
Call to action:
Reduce the time to diagnosis and improve support for patients diagnosed with pulmonary fibrosis.

In a primary care-based health system, a diagnosis of pulmonary fibrosis usually requires a GP/PCP to refer the patient to a respiratory physician in a secondary care hospital. Often, patients are also referred on to a specialist in an interstitial lung disease (ILD) centre.[12]

Achieving an accurate diagnosis of pulmonary fibrosis is complex and requires radiological, clinical and pathologic tests. In most cases, these tests were carried out at hospitals before diagnosis (Fig. 8). The vast majority of respondents had lung function and blood tests (90% and 82%, respectively) and chest X-ray and computed tomography (CT) scans (77% and 74%, respectively).

Given that CT scans are mandatory in diagnosing most ILDs,[13] we would have expected more patients to report having had CT scans. Similarly, almost half of the patients reported undergoing a lung biopsy procedure. This figure is higher than reported in the literature,[14] which may suggest the need for improved communication between healthcare professionals and patients in terms of the tests being undertaken.

Fig. 8: Tests undergone prior to diagnosis.



CT, computerised tomography; 6MWT, six-minute walk test.

Over 60% of patients received a diagnosis of pulmonary fibrosis within 3 months of their initial hospital visit, whereas a small but signifcant group of patients (13.5%) took over a year (Fig. 9).

Fig. 9: Time taken to receive a correct diagnosis after first visit to the hospital.

64.3% of patients received a diagnosis within 3 months of first visiting a hospital

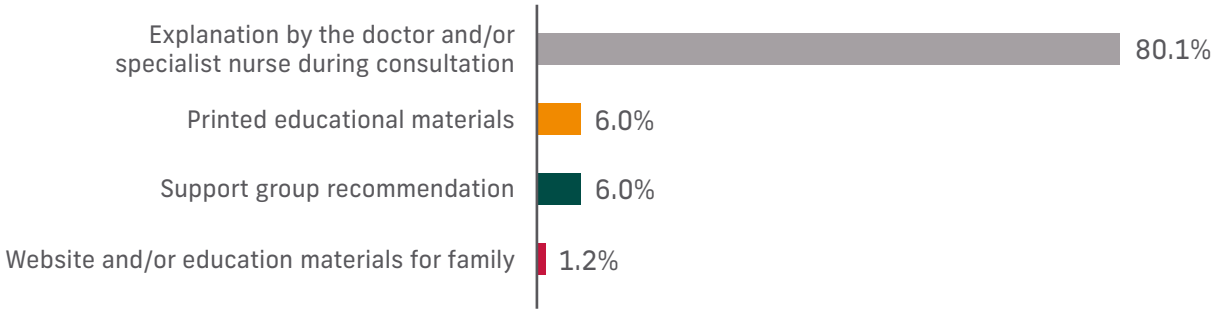
22.2% 3 months to 1 year

13.5% took over a year

n=266 (numbers refer to the number of patients who were able to recall the time it took for them to receive a correct diagnosis).

While 80% of respondents had their disease explained to them by the doctor or specialist nurse at the time of diagnosis, a quarter were diagnosed without being given adequate explanation. Also, only 6% were given printed educational materials at the time of diagnosis or signposted to a support group (Fig. 10). Helpful information given to the respondents included facts on pulmonary fibrosis, the nature of the disease, and their life expectancy. Respondents would have liked more information on living with pulmonary fibrosis day-to-day, access to a psychologist and information on support groups.

Fig. 10: Information received by patients at time of diagnosis.



n=251 (numbers refer to the number of patients who were able to recall the information they received).

"We were not given a lot of information. Most of what I learned was from attending a support group. I was told when the illness was diagnosed that it was terminal, which came as a shock to both me and my wife."

–A patient from the UK

"I felt desperate and alone. After such an awful diagnosis, my next appointment was in 6 months and nothing in between!"

–A patient from the UK

"I was not told how serious the illness was."

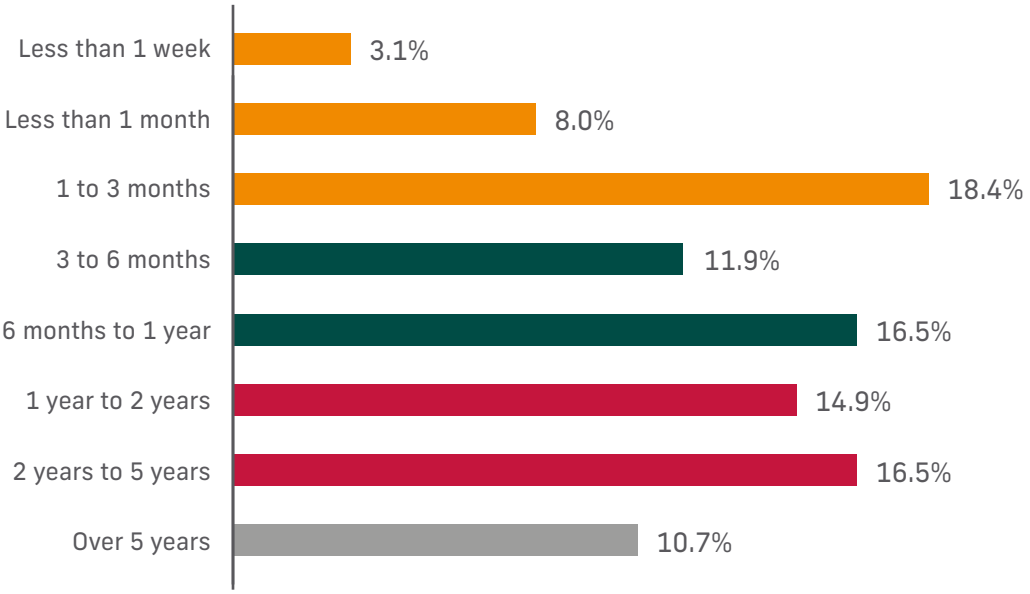
–A patient from Spain

Overall time to diagnosis and feelings throughout the patient journey

Call to action:
Strive for a diagnosis within 3 months of first symptoms to improve patient outcomes.

The total time from the first onset of symptoms to a final diagnosis varied greatly among patients. (Fig. 11). Nearly 30% had received a diagnosis within 3 months. However, more than 40% of patients had to wait over a year to be diagnosed, and treatment would have been delayed. This is likely to have affected both their quality of life and outcomes.[15]

Fig. 11: Overall time to diagnosis.

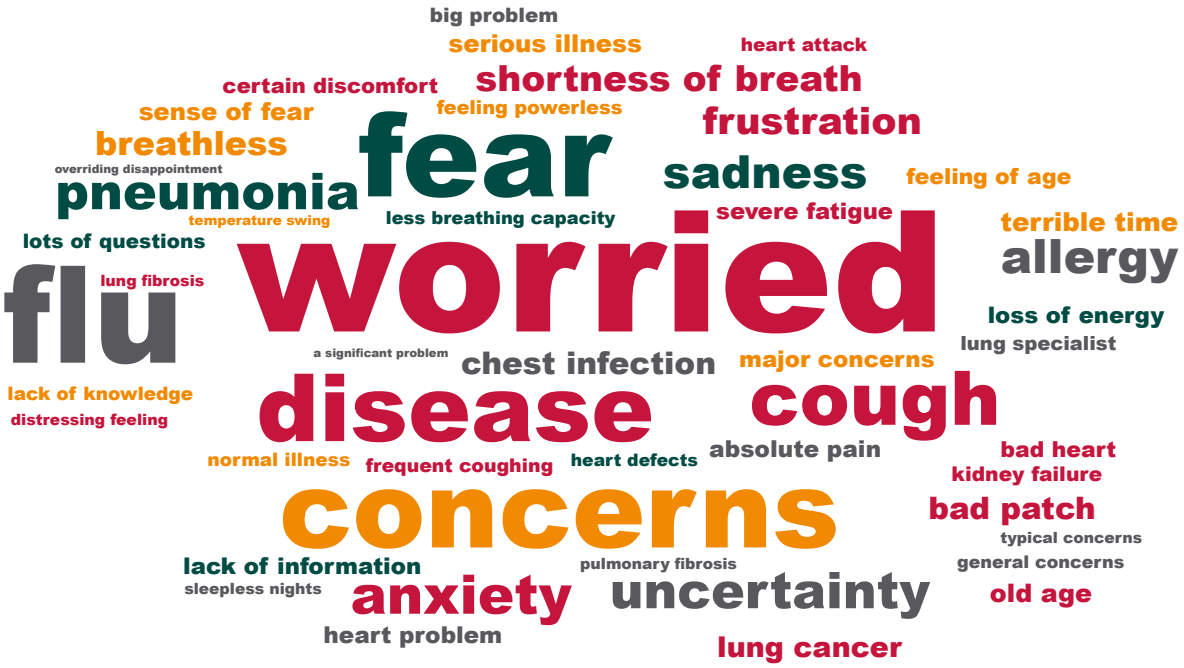


n=261 (numbers refer to the number of patients who were able to recall time to diagnosis).

- **29.5% received a diagnosis within 3 months**
- **42.1% took over a year for diagnosis**
- **>10% took over 5 years**

Patients experienced a multitude of feelings before being diagnosed with pulmonary fibrosis. Fear, worry and concern were the most common feelings experienced (Fig. 12), highlighting the need for a more active role in managing the psychosocial aspect of the condition.

Fig. 12: Feelings experienced by patients on the path to diagnosis.



Advice and top tips from patients

Respondents displayed a good understanding of the patient journey and its different stages. Their advice and the top tips they offered reflected this. They recommended:

Seeking help early (especially if breathless) and pushing for a speedier diagnosis

Seeking as much information as possible from healthcare professionals at all stages

Taking regular exercise and joining pulmonary rehabilitation classes to assist with breathlessness and to ultimately help improve quality of life

Joining support groups, remaining positive, pacing themselves and making the most of their time

Conclusions

Many patients experience a long and difficult journey to diagnosis, with over 40% of patients waiting more than a year to be diagnosed. This is not adequate given that IPF and potentially other types of progressive pulmonary fibrosis have a poorer prognosis than many cancers.[5] Those that have a reduced life expectancy due to their disease should not have to wait over a year to be diagnosed.

The slow and uncertain journey to diagnosis causes patients anxiety and delays access to treatments that could prolong their lives and slow down disease progression. There is, therefore, an urgent need to speed up the diagnostic pathway and provide patients with more information at all stages to improve daily management of the disease and ultimately their quality of life. This information should come from healthcare professionals working with patient organisations. Patient support groups have a key role to play in providing opportunities for mutual support among patents and exchanging information on living with the disease.

"Never give up, keep your morale up, do physical exercise, maintain social relationships and surround yourself with good people. Put certain negative issues into perspective and enjoy time with those close to you, and life in general, because it is worth living."

–A patient from France

*Galapagos financially supported the survey, and had direct involvement in developing the questionnaire and report in collaboration with the EU-IPFF. Logistical and medical writing support was provided by emotive.

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