The EU-IPFF Consultation Guide:
A guide to living with Idiopathic Pulmonary Fibrosis
Contents

1. Welcome ................................................................................................................ 4
2. About the EU-IPFF ................................................................................................. 7
3. How we developed this guide ................................................................................ 9
4. What is Idiopathic Pulmonary Fibrosis (IPF)? ...................................................... 11
5. What are the symptoms? ..................................................................................... 15
6. What are the risk factors? .................................................................................... 17
7. How is IPF diagnosed? ........................................................................................ 19
8. What is a multidisciplinary team (MDT)? .............................................................. 24
9. Centres of expertise for interstitial lung disease and IPF ..................................... 28
10. What treatments are available to me? ................................................................ 35
11. How will the disease progress? .......................................................................... 45
12. How I can take care of myself? .......................................................................... 48
13. Where can I find support? .................................................................................. 58
15. Annex 1: Questions for your doctor ................................................................... 67
15. Acknowledgements ............................................................................................ 72
16. References ........................................................................................................ 74

List of figures

Figure 1: Idiopathic Pulmonary Fibrosis definition
Figure 2: Understanding what Idiopathic Pulmonary Fibrosis is
Figure 3: Location of lungs and airways in the body
Figure 4: How Idiopathic Pulmonary Fibrosis affects the lungs tissue
Figure 5: Difference between the tissue of healthy lungs and affected by IPF lungs; and examples of X-rays taken of healthy lungs and affected by IPF lungs
Figure 6: Common signs and symptoms of IPF
Figure 7: IPF risk factors
Figure 8: IPF diagnosis pathway
Figure 9: IPF Multidisciplinary team
Figure 10: IPF Management
Figure 11: How IPF progresses over time
This document is for educational purposes only

Disclaimer: the European Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF) developed this document to educate people. Please consult your doctor if you need to discuss any medical information.

The diagrams and pictures only represent the various stages of the disease. We do not intend them to fully capture how the disease progresses.

F. Hoffmann-La Roche LTD and Boeringher Ingelheim financially supported the Consultation Guide.
Section 1

Welcome
1. Welcome

If you have recently been diagnosed with Idiopathic Pulmonary Fibrosis (IPF), you probably have a lot of concerns and questions that you wish to discuss with your doctor and healthcare professional team.

These might be about:

• the disease;
• your diagnosis and treatment options; or
• how IPF will affect your day-to-day life.

You may also be scared, worried, angry, or confused. We hope this guide can help you.

This guide has been compiled by the European Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF) to give you some important information about IPF.

However, this document has been developed for educational purposes only; please consult your doctor to discuss any medical information. We do not provide medical advice, and the content of this guide does not replace the advice and medical opinion of your doctor.

We have included sections about:

• what symptoms you may experience;
• tests you may have for diagnosis;
• monitoring your lung disease; and
• possible treatments (with or without medicines) available to you.

It can be emotionally challenging for you to discuss the disease, but some things can make managing your condition easier. This includes having access to a multidisciplinary team of doctors, nurses and specialists; and making lifestyle changes. This guide includes information about this. It also includes information about how the disease is likely to progress and what other related conditions could develop.
Since IPF is a rare disease, you may feel isolated or alone after your diagnosis. IPF patient associations and groups can have a huge impact on you and provide a platform for other patients and carers to share advice and experience with you, your family and friends. We have included contact details for relevant associations in section 13. In addition, we have also included patient and carer stories to provide a first-hand experience on the different subjects mentioned in this guide. These stories are clearly differentiated in the text.

We hope this guide is useful to you, your family and friends, or anyone you know who has recently received a diagnosis of IPF.

We welcome any suggestions for information that should be included in this guide – please get in touch (secretariat@eu-ipff.org).

Best wishes from,

Liam Galvin
Secretary
Section 2

About the EU-IPFF
2. About the EU-IPFF

The European Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF) is made up of 17 patient organisations from 13 European countries and is the first such pan-European organisation. We are committed to changing the IPF landscape in Europe.

The EU-IPFF aims to defend the rights of IPF patients in Europe. We do this by promoting improvement in the quality of life and prognosis of IPF patients, and of patients suffering from other interstitial lung diseases.

Interstitial lung disease describes a large group of disorders characterised by progressive scarring of the lung tissue. The tissue affected is between the air sacs (alveoli) and also supports them.

For more information please visit our website: www.eu-ipff.org
Section 3

How we developed this guide
3. How we developed this guide

We developed this guide based on current information provided to IPF patients by:

• hospitals;
• patient organisations; and
• industry.

This included:

• nineteen guides issued by patients’ associations;
• seventeen guides issued by hospitals; and
• four guides issued by industries.

Medical experts and patient groups reviewed, assessed and edited the information to compile a high-quality, unbiased and comprehensive patient guide.

Two meetings among patient representatives and members of the EU-IPFF Scientific Advisory Board took place (in Brussels, in November 2016 and in Milan, in February 2017) to review and apprise the information collected.

For more information or to share any concerns or comments you may have on this guide’s contents, please contact: secretariat@eu-ipff.org

References

We have used numbered references in square brackets throughout this document. You can find the complete references in section 16.
What is Idiopathic Pulmonary Fibrosis (IPF)?
4. What is Idiopathic Pulmonary Fibrosis (IPF)?

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<tr>
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<td>P</td>
<td>Pulmonary</td>
<td>Refers to the lungs</td>
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<tr>
<td>F</td>
<td>Fibrosis</td>
<td>Formation of scar tissue</td>
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Figure 1: Idiopathic Pulmonary Fibrosis definition (adapted from: [1])

Idiopathic Pulmonary Fibrosis (IPF) is a rare, long-term, progressive disease that affects the fragile tissue in the lungs. It leads to a gradual, persistent decline in lung function.[2] IPF typically occurs in people who are more than 45 years of age, and the average patient age is 65.[3] The disease is more common in men than women.[4] About 110,000 people in Europe have IPF, and 35,000 new patients are diagnosed each year.[5] The disease is irreversible, and without treatment, half of IPF patients will die within 2-5 years of diagnosis.[6]

Figure 2: Understanding what Idiopathic Pulmonary Fibrosis is (adapted from: [7])

<table>
<thead>
<tr>
<th>What IPF is:</th>
<th>What IPF is not:</th>
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<tr>
<td>Scarring on the lungs</td>
<td>It is not cancer</td>
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<tr>
<td>Difficulty in breathing</td>
<td>It is not cystic fibrosis</td>
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<tr>
<td>Unknown cause</td>
<td>It is not contagious</td>
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Figure 2: Understanding what Idiopathic Pulmonary Fibrosis is (adapted from: [7])
How does IPF happen?

In IPF, damage to the cells that line the air sacs, or ‘alveoli’, of the lungs leads to the formation of scar tissue. This is why IPF is one of the interstitial lung diseases (ILDs). Interstitial lung disease describes a large group of disorders characterised by progressive scarring of the lung tissue between and supporting the air sacs.

In IPF, scar tissue builds up around the alveoli and this makes it harder for oxygen to pass into the bloodstream.\(^{[8]}\) As a result, the brain, heart and other vital organs may not get the amount of oxygen they need to work properly.\(^{[9]}\) The amount of scarring usually increases over time, although how quickly it progresses varies. The scarring is generally irreversible.

Figure 3: Location of lungs and airways in the body (adapted from: [10])
Figure 4: How Idiopathic Pulmonary Fibrosis affects the lungs tissue (adapted from: [11])

Figure 5: Difference between the tissue of healthy lungs and affected by IPF lungs; and examples of X-rays taken of healthy lungs and affected by IPF lungs (adapted from: [12] [13])
Section 5

What are the symptoms?
5. What are the symptoms?

The symptoms of IPF develop over time, and can vary from person to person. The most common symptoms of IPF are shortness of breath (known as dyspnoea) along with the following:

- dry chronic cough;
- loss of appetite;
- gradual, unexplained weight loss;
- fatigue, tiredness and generally feeling unwell; and
- aching joints and muscles.[4]

About half of people with IPF may also experience finger clubbing (widening and rounding of the tips of the fingers or toes).[4]

When someone uses a stethoscope to listen to the lungs of a person with IPF, they may hear ‘velcro-like’ crackles in your lungs. These are ‘opening’ sounds made by the small airways when inhaling.[10]

![Figure 6: Common signs and symptoms of IPF (adapted from: [1])](image-url)
Section 6

What are the risk factors?
6. What are the risk factors?

It is sometimes unclear what the relationship between IPF symptoms and risk factors are, but research is ongoing. We don’t know exactly what causes people to develop IPF, however, there are several things that increase a person’s risk of developing IPF. They include:

- cigarette smoking;
- environmental and occupational exposures like pollution;
- microbial agents (chronic viral infection);
- gastro-oesophageal reflux disease (GERD) and
- someone else in your family who has pulmonary fibrosis.\(^{[14]}\)

Figure 7: IPF risk factors (adapted from: [1])
How is IPF diagnosed?
7. How is IPF diagnosed?

The symptoms of IPF are like those of other, more common lung diseases. Unfortunately, this means the diagnosis is often delayed and misdiagnosis occurs frequently.\[10\] This can then delay access to specialised care.\[15\]

The delay in diagnosing IPF ranges from six to 18 months.\[15\]\[16\] To avoid this delay, people should be referred to ILD specialised centres.

If you are suspected of having IPF, a multidisciplinary team to confirm diagnosis should include:

- respiratory specialist (i.e. pulmonologists);
- radiologists; and
- pathologists.

As a first step, your family doctor should refer you to a respiratory specialist. The pulmonologist may conduct the following diagnostic tests to confirm the diagnosis, or refer you to a centre with expertise in IPF.

**Diagnostic Tests**

- **Lung auscultation (listening to the lungs)**
  
  Lung auscultation is one of the most important investigations as it can diagnose IPF early.\[17\] If ‘velcro-like’ crackles can be heard when listening to the patient’s lungs it is a key clinical feature that we can use to recognise IPF.

- **Pulmonary function testing (PFT)**
  
  Pulmonary function tests are also called lung function tests. Spirometry is the most common type of pulmonary function test performed in people with IPF.

  During the PFT, you will be asked to breathe in and out in various ways. These tests can determine how IPF affects the way you breathe and get oxygen. They allow the doctor to measure the total amount of air in your lungs and assess the flow of air in and out of the lungs. This shows how well the lungs are working.\[18\]\[19\]

  The test is painless, but requires a lot of effort from you to complete.
Six-minute walk test

This test measures how far you can walk in six minutes, and how much oxygen is present in your body while walking.\textsuperscript{[20]} It finds out how fast you run out of oxygen while walking and if you need supplementary oxygen therapy.

Chest X-ray

Chest X-rays provide a two-dimensional image of your chest using a small amount of radiation. Chest X-rays can often reveal when something is wrong in the lungs. However, they are not good at finding out if you have IPF.

If your medical team thinks you may have IPF, they may do an X-ray to look for lung abnormalities that suggest scarring of the lung tissue.\textsuperscript{[1]} However, 5-15\% of patients with significant scarring will still have a normal chest X-ray. Therefore, it’s important to remember that IPF cannot be diagnosed from a chest X-ray alone.\textsuperscript{[10]}

Blood tests

Blood tests are done to investigate other known causes of interstitial lung diseases (ILD), such as autoimmune diseases.\textsuperscript{[21]}

High-resolution computed tomography (HRCT)

A High-Resolution Computed Tomography scan\textsuperscript{[4]}, or HRCT scan, is the most important imaging study we use to diagnose IPF.

With a high-resolution CT scan, doctors can create detailed images of the inside of the chest.

The scan is quick, painless, and uses relatively small amounts of X-ray radiation. In half of all cases, a HRCT is enough to confirm a diagnosis of IPF, and no lung biopsy is necessary.\textsuperscript{[12]}

Bronchoscopy for trans-bronchial biopsy

During a bronchoscopy, your doctor will insert a bronchoscope (small flexible tube) through your nose or mouth under local or general anaesthesia. The tube is lowered into the lungs through the trachea, or windpipe, and into the lungs.

The doctor then takes a small piece of lung tissue, known as a ‘trans-bronchial biopsy’, for testing. The limited amount of tissue they can take in this test may
make it difficult for your doctor to find out for certain what is causing your lung problem.[10]

* Autoimmune diseases are diseases where the body produces antibodies that attack its own tissues. This can lead to the deterioration and in some cases to the destruction of such tissue.

**Bronchoalveolar lavage (BAL)**

A BAL involves taking fluid from your lower respiratory tract using a small, flexible tube called a bronchoscope. A sample of cells is then taken from this fluid for testing. This is a minimally invasive procedure that can be very helpful to exclude or investigate other interstitial lung disease. However, there are risks associated with a bronchoscopy, which your doctor will discuss with you.

**Lung biopsy**

A lung biopsy is a surgical procedure in which a small piece of lung tissue is removed and analysed. You will only need a lung biopsy if doctors cannot diagnose IPF from:

- your medical history;
- a physical examination;
- blood tests; and
- a high-resolution CT scan.

A lung biopsy is used together with a high-resolution computed tomography (HRCT), to assess how the disease is progressing.[11] Depending on your individual risk factor as well as on the preference of the surgeon, lung biopsies can be performed as:

- so-called ‘open surgery’; or
- video-assisted thoracoscopic surgery (VATS).

VATS is less invasive than open surgery, but not all patients are suitable for lung biopsy.

**Genetic counselling**

Genetic counselling could be useful for patients with a family history of lung fibrosis.
How doctors diagnose idiopathic pulmonary fibrosis (IPF)

When doctors suspect you have pulmonary fibrosis, they must try different things to confirm the diagnosis and find the cause. If your doctor suspects you have idiopathic pulmonary fibrosis (IPF), you should be carefully evaluated to exclude any other interstitial lung disease (ILD).

If the medical team cannot find the cause of your ILD, an HRCT scan that shows ‘usual interstitial pneumonia’ (UIP) means you have IPF. If the HRCT scan shows you do not have UIP, doctors can diagnose IPF using specific HRCT scans and by identifying changes in lung tissue.

Diagnosis of IPF is most accurate when it is made through multidisciplinary discussion among ILD experts. Multidisciplinary discussion is when different medical experts work together. See explanation of multidisciplinary teams in the next section.

![IPF diagnosis pathway diagram](adapted from: [21])
Section 8

What is a multidisciplinary team (MDT)?
8. What is a multidisciplinary team (MDT)?

MDTs are made up of people who specialise in different medical skills including:

- doctors in different specialties;
- nurses in different specialties; and
- other healthcare providers.

The diagnosis of idiopathic pulmonary fibrosis (IPF) is multidisciplinary, meaning it draws on experienced clinicians, radiologists, and pathologists to diagnose IPF. Internationally, this approach is proven to improve the accuracy of diagnoses.

Therefore, in idiopathic pulmonary fibrosis they need to work together to confirm your diagnosis and provide you with the best possible treatment. This approach can improve the accuracy of your diagnosis and avoid unnecessary testing, as well as resulting in a better experience for you.

Every leading ILD specialised centre has in place an MDT to determine treatment decisions. The type of people on an MDT differs depending on the country, but can include:

- consultant respiratory physician;
- consultant thoracic radiologist;
- consultant pathologist;
- interstitial lung disease specialist nurse;
- multidisciplinary team coordinator;
- physiotherapist or a rheumatologist or an immunologist*

Unfortunately, this multidisciplinary approach is not always possible. But, it is very important to try to exchange information between different specialties when evaluating a case.

* Within ERN-Lung (European Network on rare pulmonary diseases), the minimum composition of an MDT teams consists of: two pneumologists, one experienced lung radiologist (could be an external radiologist), one experienced lung pathologist (could be an external pathologist), one nurse, one physiotherapist, one GCP certified study nurse, one social worker, one secretary and access to an immunologist or rheumatologist, or both.
From Ireland

Patient story:
My experience with an IPF specialised nurse
D.C.

After being diagnosed with IPF, my wife and I were devastated. We didn’t know where to turn or what to do. We were numb. After the consultant left the room, we were handed over to our respiratory nurse. She explained my diagnosis, treatment and management plan repeatedly until I fully understood what I was dealing with.

We had a nurse dedicated to our case for any questions, worries or queries at any time. This was invaluable support to my wife and I, as we had a lot of concerns regarding my health, medication and future. The nurse was always very approachable, informative and reassuring.
From U.K.:

ILD specialised nurse story:
Annette Duck

The ILD [interstitial lung disease] specialist nurse is an important part of the regional multidisciplinary team.

Trained ILD nurses will understand the difference between IPF and other ILDs and why it is important to get a correct diagnosis for the management and treatment options of IPF.

They will be able to support patients and talk to them about their investigations and test results. They can also help them with their various treatment options, prognosis and management plans.

ILD nurses are knowledgeable about the disease trajectory and can answer concerns that patients may have about advancing disease and an uncertain future.

ILD nurses will remain the support anchor from diagnosis onwards providing information throughout. They will also know how and when to involve other healthcare specialists such as oxygen therapists, physiotherapists and palliative care teams for help with the management of troublesome symptoms.

ILD nurses can usually refer directly to pulmonary rehabilitation services and will liaise with general practitioners [family doctors] and the local support services including district nurses.

Having an ILD nurse who knows and understands about IPF and who can also navigate the healthcare system should be a ‘must’ for every patient diagnosed with IPF.
Section 9

Centres of expertise for interstitial lung disease and IPF
9. Centres of expertise for interstitial lung disease and IPF

Centres of Expertise (CEs) are expert centres for the management and care of rare disease (RD) patients. CEs are designated at a national level by each EU Member State.

ILD/IPF centres of expertise specialise in ILDs (interstitial lung disease). They aim to provide IPF patients (among others) with the highest standards of care and to deliver:

- timely diagnosis;
- appropriate treatments; and
- follow up. [24]

The European Union Committee of Experts on Rare Diseases (EUCERD) recommends that centres of expertise offer a wide range of specialised services. These should include:

- consultations;
- medical examinations;
- specialised equipment;
- genetic testing and counselling; and
- social care.

CEs also contribute to research and collaborate with different stakeholders, including rare diseases’ patient organisations. [25]

The European Commission has organised European Reference Networks (ERNs), aimed at tackling complex or rare diseases and conditions that require highly specialised treatment and resources. CEs are involved in European Reference Networks and organise travel of expertise or cross-border care when appropriate. More information about ERNs is available here. (https://ec.europa.eu/health/ern/policy_en)

A European Reference Network on respiratory diseases (ERN-LUNG) was launched in March 2017 and IPF is part of this programme. More information about ERN-LUNG is available here. (http://ec.europa.eu/health/sites/health/files/erm/docs/ernlung_factsheet_en.pdf)

The list of expert centres in Europe can be found here. (http://www.orpha.net/consor/cgi-bin/Clinics_Search_Simple.php?lng=EN&LnkId=7029&Typ=Pat&CnsGen=n&fdp=y&from=right-Menu)
From Germany

Patient story:
My diagnosis, and my life with IPF

K.G.

Even a few years before I was diagnosed with IPF, I had noticed myself breathing more heavily when exercising or hiking. In early 2012, I started coughing and suffering from shortness of breath as well. My doctor diagnosed bronchitis and prescribed the use of a cortisone spray, but my cough didn’t improve.

I then had an HRCT, a bronchoscopy and repeated monitoring of my lungs, but still didn’t have a clear diagnosis of what was wrong. The head physician at the lung clinic I was attending wanted to perform a biopsy, but I was reluctant to take the risk. Instead, I visited a university clinic specialising in lung disease. After reviewing my CT-scan and listening to my lungs, the doctor there immediately diagnosed IPF.

The doctor explained what this meant and started me on a course of pirfenidone medication, with check-ups every three months.

At first the shock from the diagnosis was huge. The more I read about IPF, learning how it is both fatal and incurable, with an average post-diagnosis prognosis of 2-3 years, the more I realised how serious the situation was.

I decided to learn everything I possibly could about the disease, its progression, possible treatments, and the latest research into it.

I joined the patient support organisation ‘Lungenfibrose e.V.’. I would highly recommend any fellow IPF sufferer to join a patient support organisation.

I also took part in two clinical studies (Panorama study and Riff study) at the Thorax-Clinic in Heidelberg, Germany. Taking part in such studies is highly beneficial – not only do you contribute to research into new treatments and drug trials, but you also undergo monthly examinations and medical monitoring throughout the study. This can help improve your prognosis.

So far, I am doing quite well. My lung function has remained relatively stable, I hardly cough at all, I can still travel long-distances by plane and I can still exercise (with some restrictions). I remain optimistic about potential progress in IPF-related research.
Maybe one day – and just in time – there will be a medical breakthrough. This is what I am hoping for, even if the approval process for new medications takes time.

I would advise every IPF patient to swap stories and experiences with other IPF patients, via a support group, and to fight the progression of the disease with an active mind, physical activity and a positive attitude towards life.
From Greece

Patient story:
Recognising IPF and facing its reality

S.P.

It was the autumn of 2008 and I worked with a friend for the harvest of olives at one of my properties. My friend, listening to my breath, asked me if I had a problem with my lungs. I replied that I had no problems, but that sometimes I had difficulties in breathing and I had to stop working to take a deep breath. In addition, I was a bit overweight and I knew I should have been on a diet. Therefore, I thought about going to a pulmonologist to have my lungs checked.

After this conversation, I did not have enough strength to walk or to do some manual work as I was experiencing even more difficulties in breathing, so I decided to go to the doctor.

Luckily the pulmonologist I went to knew about IPF. Through a high-resolution computed tomography [HRCT], he could already see the scarring on my lungs. After that I needed to go to the hospital for some additional diagnostic tests. I went to a public hospital in Athens as I live in the countryside and there is no pulmonologist there. I did all the tests, including a bronchoscopy and a biopsy thanks to which I got my IPF diagnosis.

I went to the doctor in the hospital, together with my wife, who has always supported me.

He asked me, in a severe tone: “Who suggested you have your lungs tested?”

And I replied: “A friend of mine, who heard the difficulties I had in breathing.”

But he said: “Forget about it, just ignore it.”

So, I was happy about it and I left the room but I heard him saying, behind my shoulders: “You could only live for another 3-5 years.”

So, I turned around and I asked him: “What did you say, doctor? So, there is no cure?”

He did not answer me. He said only that he could give me cortisone at a later stage, when the stage of the disease would be advanced and I would need oxygen.
Given his behaviour, my understanding was that the doctor was not willing to discuss anything further about it. I turned to my wife who heard all the discussion and she was really shocked.

She asked the doctor: “Should you not monitor the progression of the disease?”

He replied: “It is okay to do a CT scan in six months to see how fast the disease is progressing.”

The conversation finished there and we went back to our house.

I followed the doctor’s advice and I managed to forget about my disease. However, my wife never stopped reminding me that I needed to do a CT scan after six months. Nevertheless, I was trying to follow my doctor’s advice and to forget about the disease. So, on the one hand there was my wife reminding me about the CT scan, and on the other hand, I could hear the doctor’s voice saying: “Forget about it. Forget about it.”

After some time, I noticed that my breathing was getting worse and two years later I finally decided to do the test. It was spring 2012.

So, I went back to the same doctor in Athens and I brought him the new CT scan that I did, together with the old one.

He looked at them and told me: “Unfortunately, your disease is progressing”.

He described how the disease would progress even more, until I died, since there was no cure for IPF.

I left and I was really disappointed. I lost the earth beneath my feet; I could not even walk and everything around me began to seem black to me. So many thoughts in my head, I did not know what to do and I was really desperate.

I came back home and I went straight to the bathroom as I wanted to take a shower. I was lost and I moved in a mechanical way.

I went to the room where I have my computer and I desperately started looking for information about IPF, until I found the website of a pulmonologist. So, I read the description of the disease and it said that there was no cure. However, it said a new medication had been found and it showed good results – allowing the patient to survive for longer.
So, I called the pulmonologist and we had a nice conversation. He was completely different from the other doctor, way more polite and kind. He heard all my story, he encouraged me and he supported me psychologically, and he has been doing this for all these years.

I slowly began to realise that I even if I had a rare disease, I was not alone and my life could not stop. I had to go on and cope with my disease. I started to look for other IPF patients as I realised that being introvert was the worst thing. I needed to talk about IPF. Sharing experiences can help because you realise that other people are experiencing the same and having the same concerns, but they are still struggling, not giving up. Some of them have defeated the disease, avoided depression and looked at the right opportunity to fight IPF.

Finally, being the president of the National Association of Pulmonary Fibrosis, I have met many patients with IPF and I have listened to their stories. Therefore, I am carrying the weight of my own disease, plus that of all the other members of the association.

I hope my testimonial can help other IPF patients outside Greece as well.
Section 10

What treatments are available to me?
10. What treatments are available to me?

To manage your IPF, both drug treatments and other treatments are recommended. You should also have regular check-ups and medical examinations.

As soon as you are diagnosed with IPF, you should be offered psychological support. You should also get in touch with patient groups, which can support you at all stages of your disease.

![Figure 10: IPF management (adapted from: [1])](image)
Pharmacological treatments

There are currently no curative treatments for people living with IPF. However, two drugs which have been approved by the European Medicines Agency are able to slow down the disease progression. [26] [27] One of these drugs contains the active substance (ingredient) nintedanib and the other contains the active substance pirfenidone.

You can find more information about these two drugs at the following links:


Both of the approved drugs are antifibrotic drugs. They help to prevent tissue scaring and have been shown to reduce the loss of lung function by about 50% in 1 year [26] [27] [28]. (This was measured by spirometry – a method of measuring breathing capacity.)

Recent IPF guidelines do not favour one drug over another; they provide a conditional recommendation for the use of either nintedanib or pirfenidone in the treatment of IPF [28]. This means that your doctor will evaluate which drug to prescribe on a patient-by-patient basis.

You should always consult your doctor to discuss treatment options, as every patient is different and no two treatment plans are alike. Furthermore, each European country has their own national official guidelines on IPF and its treatment. Payment and reimbursement criteria for these drugs may also differ between countries. A final decision on your treatment should be taken together with your doctor as many factors need to be taken into consideration.

If you have any further questions about possible pharmaceutical treatments and/or if you notice and want to report any side effects, talk to your doctor or pharmacist.
Clinical trials

A clinical trial is a study which tests the effectiveness and safety of a new therapy on humans. There have been several clinical trials into treatments designed to treat IPF – you should check with your doctor if there are any ongoing trials you may be able to take part in. You can also find wider information on on-going clinical trials using ClinicalTrials.gov (worldwide) or the EU Clinical Trials Register.
From Germany

Patient story:
What made me take part in two clinical studies

K.G.

Clinical studies are an important part of research into new active substances and therapies for the effective treatment of diseases, including IPF, which still does not have a cure. Pharmaceutical researchers and specialised doctors, at university clinics with study centres, work together to try out or study new active substances in patients.

Taking part needs a high level of commitment and willingness to take risks. The participant does not find out the results of the study until long after the study is complete. Bearing all this in mind, what made me – as a patient – take part in two studies?

First and foremost, without these studies there will be no longed-for medical progress in the treatment of my disease. As participant in a study I am serving the development of new therapies. I hope that I will not waste valuable time taking a placebo (tablet with no active ingredient), but that I will be receiving the new active substance. I try to guess this by following closely how my disease progresses throughout the course of the study (usually 12 months).

This is the advantage of taking part in a study. During the study, I was carefully examined by the very experienced study nurses and the study physician every month. This included my blood values, my lung function values, my physical state and performance and other details. These tests are particularly important and interesting for a patient with a progressive disease like IPF.

As participant in a study you feel under control, which is quite calming. I felt I was well informed about my health developments. I very much welcomed the monthly opportunity to discuss with the study personnel all the questions I had. These monthly assessments are extremely important, interesting and reassuring for a patient with a severe disease and an unforeseeable progression.

I can only recommend taking part in a study – even though the patient is not included in the design and strategic goal-setting. Unfortunately, you are expected to submit to the study with very little information.
Non-pharmacological treatments

While a lung transplant is the only effective cure for someone with IPF, there are several non-pharmacological treatments which can help improve your quality of life. They and other treatments can help lessen the burden on you, your family and your carers.

Pulmonary rehabilitation

Pulmonary rehabilitation includes different activities designed to relieve the symptoms of IPF and improve your overall quality of life. This can include:

- exercise training;
- nutritional counselling; or
- learning breathing strategies.

Pulmonary rehabilitation can be particularly effective in:

- alleviating symptoms;
- increasing how much you can exercise (which in turn can reduce your tendency to psychosocial issues such as anxiety or depression); and
- reducing how long you may need to spend in hospital.

Oxygen therapy

Since IPF makes the oxygen in your lungs less able to travel into the bloodstream and around your body, you may suffer from abnormally-low levels of oxygen in the blood in your arteries. This is called hypoxemia. You may have hypoxemia while you are:

- resting;
- sleeping (nocturnal hypoxemia); or
- exercising.

To deal with the effects of hypoxemia, you may be given extra oxygen to improve some symptoms of IPF and your overall quality of life. Supplemental oxygen can help to reduce shortness of breath and improve your ability to perform everyday tasks.

At first, you may only need extra oxygen during exercise and while sleeping, but as the disease progresses you may need oxygen all the time to keep the oxygen levels in your blood at a healthy level. There are several different ways you can take in extra oxygen (oxygen therapy), through:

- a concentrator;
- face mask;
- oxygen cylinders (compressed gas); and
- liquid oxygen.
Oxygen concentrator

The oxygen concentrator is a small electric machine that takes in air and separates the oxygen from the other gases. It allows only oxygen to flow through a tube and be delivered to your lungs through a nasal cannula.

Oxygen cylinders

‘Ambulatory oxygen’ is provided in cylinders that you can use when walking or doing any exercise inside or outside the home. Many patients use a portable oxygen concentrator for ambulatory oxygen. Oxygen cylinders may be fitted with an oxygen conserver that will deliver a ‘pulse-dose’ of oxygen – only when you breathe in.

Liquid oxygen

If you are using a lot of ambulatory oxygen at higher flow rates, then your doctor may recommend that you use liquid oxygen. It is in a large reservoir unit that you use to fill a smaller device.

Administrating oxygen

Oxygen can be administered or delivered through a face mask or nasal cannula:

• the face mask fits over your nose and mouth and straps onto your head;
• the nasal cannula is two small plastic tubes, or prongs, that fit within both of your nostrils.

Oxygen and travel

You can travel with oxygen, however, different flight companies may have different rules so please check with them. Your doctor and your local patient organization can help you with this.

Your doctor will tell you which type of oxygen delivery device you should use, as well as how much oxygen you need and how often. Always check with your doctor if you are unsure about how you should use your oxygen.
Psychological support

Psychological support is a very important part of living with IPF. You can discuss this with your doctor and they may be able to offer it to you as soon as you are diagnosed with IPF. This will help you accept the disease, relieve physical and emotional suffering and improve your quality of life.

Psychological support can help you accept the treatment and the drug’s side effects as well. In an advanced stage, psychological support can help you in end-of-life planning and choosing hospice care or the comfort of your home.

It is important to involve your friends and family members in the psychological support programme as they can help you manage your IPF.

Palliative and end-of-life care

Learning about IPF and its treatment can improve your quality of life. Since IPF is a progressive disease with no known cure, you can often feel better if you can discuss end-of-life issues with knowledgeable professionals. When possible, your close family members should be included in these discussions.

Palliative care can be used for symptom control and is part of a holistic approach to managing IPF.[32] It aims to provide you with relief from both the physical pain, and the broader stresses and problems, associated with your condition. Palliative care includes things like:

• advance care planning; and
• patient and carer education.[33]

Palliative care may also include physical, mental, social, and spiritual activities – depending on your needs and preferences. The goal is to improve your quality of life, and that of your family or carers.

No matter what stage of disease progression you are at, palliative care should be an integral part of your overall care. It is essential for IPF patients nearing the end of their life.[26] You should speak with your doctors to discuss how palliative care may help you and your family.

Learning to manage breathlessness

Palliative care can also help you manage episodes of breathlessness. It is very important that you learn how to minimise, cope and manage breathlessness. Learning to manage and live with breathlessness can make a big difference to your
quality of life. Your emotions can affect your breathing and being anxious about your breathlessness can make your breathing even worse. Relaxation can help to reduce breathlessness and it is useful for relieving anxiety, so try to find what relaxes you.

Managing your cough

Palliative care can involve treatments to help reduce the symptoms of cough as well. Being able to manage your cough plays an important role in managing IPF because the cough can make you feel isolated and embarrassed unless you can control it. This can make it difficult to interact and communicate with other people. In fact, managing your cough is a significant factor in maintaining your overall quality of life.

Lung transplantation

Your doctor may refer you to a lung transplant centre, where you will be evaluated as a candidate for transplant of one or both of your damaged lungs from a donor. This is the only surgical intervention procedure which can:

• reverse the progression of IPF;
• improve your quality of life; and
• improve your life expectancy.\(^{[34]}\)

Some 30% of lung transplants worldwide are performed on IPF patients.\(^{[35]}\)

Unfortunately, only 5% of all IPF patients qualify for a lung transplant, since the criteria for selection as a viable transplant candidate are quite strict. These consider a range of factors such as a patient’s;

• age;
• general health;
• likely outcome following the procedure;
• severity of IPF; and
• progression of IPF.

The criteria are established by local or national health authorities and you should speak to your doctor about whether or not you may qualify.\(^{[26]}\)

If you do qualify, it is important to remember that there are risks associated with the procedure, including serious complications, infection, or rejection of the new lung or lungs.
Tips: how to make the most of your discussion with your doctor

Having a good conversation with your doctor is very important when living with IPF.

Ask questions
Don’t be afraid to ask your doctor questions or to schedule another appointment if needed.

Bring someone with you
Don’t go to your doctor’s appointment alone; having someone else’s support can help you a lot.

Share information with your doctor
Share with your doctor what you think it is important for them to know.

Record the meeting
Ask your doctor if you can record the conversation so that you can listen to it again later.

Take notes
Take notes while your doctor is giving you some suggestions.
Section 11

How will the disease progress?
11. How will the disease progress?

IPF progression varies from person to person

It’s important to remember that the progression of IPF can vary significantly from patient to patient. It is impossible to predict exactly how rapidly IPF will progress for you.

Slow progression
Most people with IPF experience a slow, but steady, worsening of their disease. In you have IPF that is progressing slowly, you could have the symptoms for a long time before being diagnosed and then your disease could still progress relatively slowly.

Stable progression
Some people with IPF remain stable.

Rapid progression
Some people with IPF suffer a fast decline to death. This is called rapid progression.

Acute exacerbations
A minority of patients may experience unpredictable acute (sudden and short-term) worsening of their disease, called acute exacerbations. An event like this may be fatal or may leave a person with IPF with substantially worsened disease.

If it is not possible to identify the cause of this acute respiratory decline, the term ‘idiopathic acute exacerbation of IPF’ is sometimes used, where idiopathic refers to the unknown.

An acute exacerbation can happen at any time while someone has IPF. Sometimes a person is not diagnosed with IPF until their first acute exacerbation.

The main risk factor for acute exacerbation of IPF is advanced disease.

Figure 11: How IPF progresses over time (adapted from: [21])
The danger of an acute exacerbation, makes regular monitoring by your doctor essential so they can:

• track how the disease is progressing;
• see how well you are responding to treatment; and
• decide the next steps that should be taken.

As part of your regular monitoring, you will have lung function tests to monitor the progression of your lung disease. [32]

**Other conditions IPF patients might have (co-morbidities)**

IPF can be a debilitating condition as on top of the adverse effects of pulmonary fibrosis, most IPF patients have other, associated conditions. When you have more than one condition at the same time, it is called a co-morbidity. If you have co-morbidities alongside your IPF, they can have a negative effect on the quality of your life and your prognosis. This is why when doctors are managing your IPF they also need to identify and treat any co-morbidities you may develop. [39]

These co-morbid conditions can affect the lungs (pulmonary comorbidities) or other parts of your body (non-pulmonary comorbidities).

**Pulmonary co-morbidities**

Pulmonary co-morbidities include:

• pulmonary hypertension;
• emphysema;
• venous thromboembolism;
• chronic obstructive pulmonary disease (COPD); and
• lung cancer. [40][41]

**Non-pulmonary co-morbidities**

Non-pulmonary co-morbidities include:

• coronary artery disease;
• congestive heart failure;
• sleep-disordered breathing;
• gastro-oesophageal reflux disease (GERD); and
• anxiety or depression.

To find the full definitions of co-morbidities, please visit [http://erj.ersjournals.com/content/46/4/1113](http://erj.ersjournals.com/content/46/4/1113)
Section 12

How can I take care of myself?
12. How I can take care of myself?

IPF is a highly complex condition. The disease needs to be managed along with its symptoms. It is essential that you manage some parts of the condition yourself. This allows you to:

• control your care;
• set realistic goals; and
• prepare for your future.

You need to work closely with your care team to take an active and positive role in your care pathway and outcomes.

Your doctor can best advise you on how to deal with the challenges associated with your condition, but here are a few suggestions for lifestyle changes you can make to improve your quality of life:

**Quit smoking**

If you give up smoking, or avoid exposure to second-hand smoke, you can:

• increase oxygen levels in your blood;
• lower your blood pressure and heart rate; and
• reduce your risk of cancer and heart disease.

Doing these things can help you prevent further damage to your lungs.

**Stay active**

If you take regular, moderate exercise, it not only helps you maintain a healthy weight, but also strengthens your muscles and keeps your body working efficiently. This will help you to manage shortness of breath.

**Eat healthily**

It can help you stay as healthy as possible if you eat a balanced, nutritious diet including:

• fruit;
• vegetables;
• whole grains;
• lean meats; and
• low-fat dairy products.

Your diet should be low in:

• saturated fats;
• sodium (salt); and
• added sugar.
Get plenty of rest

Sleep is essential for boosting your immune system and improving your overall sense of well-being.

Stay up-to-date with vaccinations

It is especially important that you stay up to date with flu and pneumonia vaccines. You should also avoid exposure to infections – these can make IPF worse.

Avoid stress

If you are physically and emotionally relaxed it may help you to avoid excessive oxygen consumption. Relaxation techniques can help and they can also teach you to manage the panic that can go with shortness of breath.

Join a patient group

It is important to become actively involved and to be proactive in managing the challenges of living with IPF. It can help you cope with your disease if you:

• get together with other people with IPF;
• create a sense of community;
• share experiences; and
• feel understood.

Involvethy family and caregivers

It might be difficult to talk about the disease with your family and friends, but talking can help you to overcome your fears and difficulties. Keep them close to you, involve them and keep them informed about your disease. This can help you to cope with your IPF diagnosis and feel less alone.

Relaxation and mindfulness

There are different relaxation techniques that can help you to handle the emotional and psychological challenges that may come with the IPF diagnosis. Mindfulness can help you to:

• focus on what is most important in your life;
• find motivation;
• be positive while coping with the physical and lifestyle challenges of living with IPF; and
• calm down when you are distressed, discouraged, scared or in pain.

Yoga

Yoga is beneficial for IPF patients as it can stimulate your lungs through focused breathing. It also helps workout your diaphragm (the muscle that operates the lung). Basic breathing exercises may improve lung function and ease breathing problems.
Tips and tricks that healthcare professionals say can help

Your doctor

Make sure you have a respiratory specialist who is an expert in lung fibrosis and whom you trust.

Discuss with your respiratory specialist what to do if the symptoms get worse.

Your doctor should give you all the information you need about managing side effects and prophylactic therapy (therapy to prevent symptoms or disease). In some cases, after talking to you, your doctor may change the dose of your medicines until symptoms resolve. Sometimes treatments need to be temporarily interrupted.

Tell your doctor if someone else in your family has IPF or another type of pulmonary fibrosis. In familial pulmonary fibrosis, smoking is especially dangerous.

Your patient history

Keep a detailed and full record of your patient history so your doctor has a full picture of your healthcare. This can include a copy of:

- your clinical tests; and
- medicines used.

Stay informed, read international guidelines and go to postgraduate courses and international congresses.
From the Netherlands

Patient story:
The importance of meditation during the IPF journey

When I was diagnosed with IPF, I realized that I was thinking about it all day long and could not get it out of my mind. When doing different activities with my wife, I always realized that it could be the last time for me. So, we decided to look for some psychological help, since it was not offered to us.

The second step was to find a psychologist with whom I could feel confident.

We went on holiday to Kos, in Greece, and I attended meditation training with my wife and this helped us a lot. We learnt how to clear our minds and how to choose what we want to think about. And thanks to this, I can now handle IPF better.

I think that not enough attention is given to the mental implications of being diagnosed with IPF.
From Ireland

Patient story:
My experiences of mindfulness

M.C.

Mindfulness has helped me to reduce stress and anxieties that I have experienced due to having IPF. Being mindful has helped me to see (at times we look but do not see,) and enjoy even the small things in life that we sometimes take for granted.

To be happy in the moment was a very valuable learning experience for me. Time can be precious with IPF, so mindfulness has meant that I cherish such moments.

Crucially, mindfulness has helped me to let go of things that I used to get worked up about (such as being stuck in traffic) and I am a calmer person as a result.

I found that the importance of breathing properly was a key feature of mindfulness. It helps get oxygen to the brain and has a calming effect, which can only benefit people with IPF.
From Italy

Teaching mindfulness to IPF patients

Roberto Ferrari

We have experimented with the techniques of mindfulness on patients with IPF. [41] IPF patients were enrolled in a 12-month study, aiming at showing that mindfulness represented an adequate and feasible approach to the different stages of the disease. It found that mindfulness could lead to significant improvements, especially in the mood of the patients and in their level of stress.

Practising mindfulness means bringing attention to what is happening in that precise moment in your body, in your mind and in the external environment. It is not only about being focused, but also about staying calm and being aware, with an attitude of curiosity, kindness and non-judgment.

It is a way of being, that you need to practise and includes:

• different meditation exercises;
• listening to your body; and
• being aware of what is happening around you in that precise moment.

It allows you to develop the capacity to face difficulties in your everyday life. Everyone can practise mindfulness, but it can be enhanced following a programme which lasts eight weeks, called MBSR (Mindfulness Based Stress Reduction).

During the last 30 years, its effectiveness has been tested with different health conditions that involve suffering. Today, mindfulness is considered to be a fundamental part of complementary medicine that can be integrated with traditional pharmaceutical therapy. Neuroscience has shown that mindfulness can influence the activities and the structure of the brain, facilitating the capacity to be in control of your own emotions.

I have been the teacher of a group of IPF patients practising mindfulness in the hospital since 2011, and this has allowed me to follow these patients and to see how the quality of their life has been increasing. Mindfulness is beneficial even if practised for only few minutes per day. It can be a useful tool to calm someone down when they have a respiratory crisis. If you practise mindfulness for a half an hour every day, it can help you to change your mood during a difficult day and to deal with your disease.

Finally, it can help you to reconnect to life again and fully experience it.
Testimonials from other people in Italy with IPF who use mindfulness

“I was not feeling well for a really long time. Now, mindfulness helps me to breathe easily and lightly.

“Mindfulness made me understand that it is very important not to deny the disease. It is better to face your disease without anxiety and without reacting in an impulsive way.”

“I thought that nothing could surprise me anymore. But I had to change my mind and to open it to mindfulness.

“Yesterday my elevator broke and I was locked inside for 45 minutes. Generally, I am a very fearful lady and in such cases I usually panic and I have respiratory crises. But this time I told myself ‘Let’s try to put in practice what I have learnt with mindfulness. I started thinking about my breathing until the rescue team came. I was not stressed, nor anxious and the time passed in a quiet way. I think it is important to share this experience with all IPF patients.”

“The effect of mindfulness has been really positive. Today, while having a respiratory crisis I realized I needed to focus on my breathing and mindfulness helped me to breathe normally again and to calm down. This technique appears to be really important for managing my crisis.”

“This week I have had a long operation at the dentist. Generally, something like this causes me a lot of coughing. However, thanks to what I have learnt during mindfulness, I managed to breath consciously and I have not been coughing at all. Before experiencing mindfulness, I did not think something like that would have been feasible for me.”

“Thanks to mindfulness I have learnt how to listen to my body and my breathing. It takes me longer than expected, but it is an incredible exercise. It is not only a matter of establishing certain objectives, but it is about enjoying the moment itself, as it is so perfect.”

“I am not good at practising mindfulness on my own, but I try do to this exercise during my daily routine, trying to listen to my breathing and to my body. It turned out to be really important for me, a new dimension and one which I am not going to give up.”

“It does not happen that often, but when I am alone, coughing during the night, focusing on my breathing helps me to connect to my body and to focus on the present.”

“Thanks to mindfulness I have learnt that in every moment, even if it is difficult, I am alive and I am living my own present. Before practising it, I was very nervous; right now, I am not always calm and sometimes I still get anxious, but I am just trying to live my own life in the present. I am still very excited about it.”
The benefits of Nordic walking for people with IPF

Debora Arletti

Nordic walking is feasible all year long and is accessible to everyone. It makes it possible to exercise outside, training all the parts of your body while having fun. It is a good way to find your own balance, in a safe and dynamic way. This technique is based on recovering your natural way of walking, with the help of two sticks, which will support you. Nordic walking engages all your muscles and is beneficial for your cardiovascular system and your posture.

This activity allows you to practise your natural way of walking – the most appropriate for your physical shape. And you can use Nordic walking to also work on your posture, while being more aware of your own way of moving. Distributing your weight among your legs and the two sticks, and having a correct posture will allow you to exercise properly while feeling little fatigue.

Nordic Walking can be of great value for patients with different diseases, allowing them to exercise together, in a calm outside space.

This activity can be particularly beneficial for people with IPF, because the two sticks give physical support to help you walk. You can practise this kind of activity every day.

With the help of an experienced instructor, Nordic walking can significantly benefit your respiration.
From Italy

The benefits of yoga for people with IPF

Gaetano Zanni

I have been practising and teaching yoga for more than 25 years. I discovered it could be beneficial for people with IPF when I started collaborating with Rosalba Mele and the Italian patients’ association ‘AMA fuori dal buio’.

Practising yoga can help people with IPF because they suffer shortness of breath and breath is particularly important while practising yoga.

I took part as a guest at a meeting held in the hospital, where healthcare professionals and patients with IPF were sitting around the same table. There I realized that these patients were the ones who really needed to find techniques which could help them cope with their shortness of breath.

This lead to a successful cooperation with the AMA fuori dal buio and with the healthcare professionals dealing with IPF. We designed yoga events for people with IPF. Those who took part in these events managed to achieve great benefits to their breathing. Yoga has enabled them to breathe better both during and after exercise.

We are only at the beginning of this experience. We believe it is important to work in a multidisciplinary team that understands that yoga has positive effect on the wellbeing of people living with IPF.

Practising yoga has delivered many positive results for people with IPF and it would be great if all the patients’ associations promoted it.
Section 13

Where can I find support?
13. Where can I find support?

For people living with IPF

Living with IPF can take its toll, physically and emotionally. Learning to cope with your condition and coming to terms with your prognosis is particularly difficult if doing so alone. Finding out as much as possible about IPF can help you feel more in control.

Patient organisations are a good place to start because they provide information dedicated to people living with IPF. Joining a local support group can be helpful for you, as you can meet other IPF patients and discuss your experiences. Support from your family and friends is essential, but reaching out to fellow IPF patients going through the same experience can be particularly beneficial. Such groups can be a great source of shared wisdom, experience, strength and solidarity.

Many support groups are organised within hospitals or in the local community – your doctor or respiratory nurse should be able to tell you more. You can also get in touch with patients in your country using the organisations in the table on page 69. They are all EU-IPFF members.

General advice for carers

Caring for someone with IPF is a difficult and demanding task, which can be both emotionally and physically draining. If you’re caring for someone suffering from IPF, let your family doctor know so they can advise you on your own health and refer you for specialised support if you need it.

Remember to take care of yourself, as the day-to-day life of a caregiver can be challenging. Finally, do not be afraid to ask the doctors and nurses supporting your relative or friend with IPF for specific advice on how best to help them.

There are many support organisations for carers that can also help you through this experience. And in some EU countries, carers may be eligible for financial assistance or in-kind benefits – your doctor should be able to tell you more.

Here are some suggestions that may help you to be an effective caregiver.

Be involved
Be actively involved in your loved one’s medical treatment and take part in visits to doctors.
Attend patient support group or association’ meetings and take an active role
Know which medications your loved one takes and create a medication list.
Know how to operate any medical equipment used by your loved one.

Encourage independence and a healthy lifestyle
Let your loved one do everything they can for as long as possible.
Help your loved one maintain a healthy diet and exercise programme. Help them pay attention to their weight.

Learn about palliative care
Learn about palliative care options, even long before they seem to be needed.

Help prevent infections
Help prevent respiratory infections by, for example:
• washing your hands;
• getting vaccinations;
• avoiding public transport; and
• avoiding large mass gatherings of people.

Look after yourself
Take care of yourself. Find someone who can take over your tasks when you are tied up with other things.
Advice from one carer to other carers

Work together
Plan your daily routine together and reflect in the evening on how the day went.

Encourage mental and physical health
Be aware of what is physically and psychologically possible and provide support.
Encourage a feeling of security and comfort.
Encourage regular physical activity like hiking and biking. If possible, organise these activities together with friends, families or patient groups.
Prepare healthy and digestible food together with your partner.

Find things to do that you enjoy
Make use of experiences in your social and cultural environment. Take up tasks that make you and your partner happy and that connect you to others.
Caregiver testimonial: Caring for someone with IPF

H.W.

Since my husband was diagnosed with IPF, everything has changed. He needs me much more now – life is very different.

For him, everything started with a severe flu-like infection. He had a fever, was sweating heavily and was exhausted. Then came the weight loss and the previously non-existent shortness of breath when climbing stairs. We moved into an apartment, which allowed my husband to take fewer steps.

We then embarked on the search for a diagnosis. Our family doctor prescribed a chest X-ray but nothing unusual was noticed. A pulmonary specialist then prescribed inhalation sprays, but they didn’t help; his shortness of breath remained. Allergic alveolitis was then suspected and treated with a short course of cortisone medication – to no effect.

My husband found information via a self-help organisation whose director suggested we get in touch with a specialist. We went to the University Clinic at Vienna General Hospital (AKH Wien) where, after two years without a diagnosis, the doctors determined it was idiopathic pulmonary fibrosis. It was a relief to finally have a diagnosis and to know that it was not cancer. However, our relief was short-lived when we learnt more about the disease prognosis and understood that no cure was available.

From then on everything changed. I was very worried about what would happen next and about what needed to be done to make life worth living. We now knew the prognosis and adapted our life accordingly.

My husband started working with a patient group, established contacts with IPF specialists and together with them, developed an information brochure on IPF for patients. He was physically weakened, but dedicated to the fight against IPF. This gave – and continues to give him today – the drive and the optimism to do something purposeful for the community.

Often, the situation is more burdensome for me than for my husband. I try to relieve him of as much work as possible while encouraging him to remain physically fit, doing some training and walking outside, but away from traffic.
His muscles become weaker if he doesn’t move enough. He also suffers from constant tiredness and needs a lot of support to make any effort. Associated conditions such as a neuropathy further limit his level of activity. Every day is different.

Any travel, short or long, must be planned down to the very last detail. Oxygen needs to be on hand for more strenuous activities. Some days our routes are shorter, sometimes they are longer. To limit exacerbation, we avoid events and public transport during flu season. Almost everything requires additional planning. Nevertheless, we continue to experience many fulfilling moments together.

My advice to IPF patients is to keep:
• up communication with family and friends; and
• doing activities that fulfil your life, your humour, and your courage.
This will help you to sustain your own fitness.
From the Netherlands

Carer’s story:
My wife has IPF and I am her carer

Henk is a man of few words. However, the words he says, come directly from his heart. Three years ago, his wife Carla was diagnosed with IPF and at that time they had been together for only few years. Nevertheless, this made their relationship stronger and as Henk says, they had to learn how to deal with it. Carla is now 61 and Henk is 64.

The biggest change is that Carla does not work anymore and she is at home all day, while Henk is taking care of the house. He was used to doing this before and he does not mind the fact that his wife can now do fewer things.

Henk says:

We can still do many things. We can still walk, although we need to use a pushchair and to carry the oxygen. Indoors it is not so bad, but we needed some time to get used to it.

I need to walk next to someone with a tube in the nose, and people are looking at me. At the beginning, I used to turn my head to the other side but now I look at these people, directly in their eyes, as if I want to tell them 'Come on, do you have something to say?'.

Maybe they think it is pathetic and of course it is not fun for me, but it is very far away from being pathetic. At the beginning, Carla could already see all the obstacles, but I did not want to see. Maybe it reflects a different approach between men and women because women are more likely to think ahead.

Carla tells me that she does not know what is going on in my head, and of course she doesn’t. Sometimes I sit down and cry, even when I am at the doctor and some other times I only get wet eyes. But we cannot see into the future and sometimes it is very difficult to deal with it.

I have deep respect for Carla because she has gone through so many things in her life.

It was difficult to deal with the fact that she was not eligible for lung transplantation, so we spent one entire week in the hospital for nothing. But Carla is a very strong woman and together we are strong enough to go through this.
So we are trying to do spend as much time as possible together, doing fun things. And now that I am retired, I am glad that I will not have to work anymore.

Carla has her own things to do and she goes to visit her grandchildren, as she should continue doing so. I can now stay at home and do the things in the house; so since January I have been at home and now we can go on holidays wherever we want, and we can enjoy it!
## European support for people with IPF

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<td>UK</td>
<td>British Lung Foundation (BLF)</td>
<td><a href="http://www.blf.org.uk/ipf">www.blf.org.uk/ipf</a></td>
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<tr>
<td>UK</td>
<td>Action for Pulmonary Fibrosis (APF)</td>
<td><a href="http://www.actionpulmonaryfibrosis.org/">http://www.actionpulmonaryfibrosis.org/</a></td>
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<tr>
<td>UK</td>
<td>Pulmonary Fibrosis Trust</td>
<td><a href="http://www.pulmonaryfibrosistrust.org/">http://www.pulmonaryfibrosistrust.org/</a></td>
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14. Annex 1:

Questions for your doctor
15. Annex 1: Questions for your doctor

When you are diagnosed

1. What can I still do now that I have IPF?

2. What can I no longer do now that I have IPF?

3. What problems should I expect?

4. Do the symptoms get worse over time?

5. How do external factors affect my lungs, for example:
   • strong smells,
   • pets’ fur, and
   • the weather?

6. How can I get a second opinion?

Treatment

7. Are there any other treatments available, apart from pharmacological (medicines) treatments?

8. Why have you prescribed this particular treatment?

9. What should I do to cope well with my disease?

10. What should I do if my symptoms suddenly get worse?

11. What should I do if suddenly I get very afraid?

12. Can I go to a nurse practitioner?

13. Can I get my treatment plan on paper?

14. Would the use of supplemental oxygen be helpful for me?
Medicines

15. I feel the medication I am taking is not helping me; how do I make sure I am taking the right medication?

16. How do I know if I am using my oxygen correctly?

17. I am switching from one drug to another; how do I know if the new drug is working better than the previous one?

Mobility and sport

18. How will exercising or working out affect my IPF?

19. What is the best form of exercise for me?

20. When I exercise, should I be supervised by a physiotherapist or can I do sport on my own?

21. Should I take more medicine before or after exercising?

22. Can I fly with my oxygen bottle?

Diet

23. Does my diet affect my IPF? If yes, how?

24. Should I follow a particular diet and should I avoid certain foods?

25. Should I gain or lose weight?

26. Would it make sense for me to go to a dietician?
Smoking

27. What happens if I do not stop smoking?

28. I have tried many times to quit smoking, but haven't been able to; what should I do?

29. Where can I get help to quit smoking?

30. Could an e-cigarette be a good alternative?

31. What can I do if others continue smoking around me and it affects my health?

Lung function test

32. Is the pulmonary function test painful? Would it make me tired?

33. Should I stop taking my medication before the pulmonary function test?

34. How does the lung function test work?

35. How does the lung function test affect my lungs and my overall health?

36. How long will it take to get the results of a lung function test?

37. What is a good result of a pulmonary function test?

38. How often is a pulmonary function test done?
Problems with feelings and thoughts

39. Sometimes I feel sad, anxious or angry about my lung disease. Is it normal to feel like this?

40. What can I do about my feelings about my IPF?

41. Who can help me to cope with my feelings about IPF?

42. What feelings do other people have about their lung disease?

43. Where and how can I get in touch with other people with a similar lung disease?

End of life

44. If treatment is not improving the health of my lungs, does it mean that I don't have long to live?

45. What should I do if my health gets worse very quickly?

46. If I get morphine, will I die faster?

47. Is there any possibility that I will die because I am very worried?
15. Acknowledgements

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EU-IPFF members

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- Action for Pulmonary Fibrosis (APF), United Kingdom.
- British Lung Foundation (BLF), United Kingdom.
- Pulmonary Fibrosis Trust, United Kingdom.
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• Anne-Marie Russell, Imperial College London, United Kingdom.
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A guide to living with Idiopathic Pulmonary Fibrosis


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