



EU-IPFF

EUROPEAN PULMONARY FIBROSIS FEDERATION



THE EU-IPFF CONSULTATION GUIDE

A guide to **living** with Pulmonary Fibrosis

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WHAT IS PULMONARY FIBROSIS?



Pulmonary Fibrosis (PF) is a progressive and incurable disease that affects the fragile tissue in the lungs. It results in lung scarring and leads to a decline in lung function and increasing breathlessness. PF can be of known cause or of unknown origin, like Idiopathic Pulmonary Fibrosis (IPF). PF typically occurs in people who are more than 45 years of age and the average patient age is 65 years old.

About 400,000 people in Europe live with pulmonary fibrosis and some 100,000 patients die each year from the disease.

Pulmonary fibrosis is irreversible and treatment can only slow the progression of the disease. On average, PF patients die within 3–5 years of diagnosis.

Common types of Pulmonary Fibrosis

- Idiopathic Pulmonary Fibrosis
- Non-Specific Interstitial Pneumonia
- Chronic Hypersensitivity Pneumonitis
- Rheumatoid Arthritis – ILD*
- Scleroderma – ILD
- Fibrotic sarcoidosis
- Unclassifiable ILD

HOW DOES PF HAPPEN?

In PF, damage to the cells that line the air sacs, or ‘alveoli’, of the lungs leads to the formation of scar tissue, which makes it harder for oxygen to pass into the bloodstream. As a result, the brain, heart and other vital organs may not get the amount of oxygen they need to work properly. The amount of scarring usually increases over time, although how quickly it progresses varies.

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Living with Pulmonary Fibrosis

* ILD: *Interstitial Lung Disease, which causes scarring of the lungs*

WHAT ARE THE SYMPTOMS?

The symptoms of PF develop over time and can vary from person to person. At diagnosis, the most common symptoms of PF are **shortness of breath, persistent dry cough and fatigue**. Over time, the cough often becomes productive, and in the later stages of the disease, other factors such as **loss of appetite, aching joints and muscles and gradual, unexplained weight loss** develop.

About half of people with PF may also experience **finger clubbing** (widening and rounding of the tips of the fingers or toes).

When someone uses a stethoscope to listen to the lungs of a person with PF, they may hear '**velcro-like crackles in your lungs**'. These are 'opening' sounds which are made by the small airways when inhaling and can be heard in both lungs.



HOW IS PF DIAGNOSED?

If you are suspected of having PF, several investigations are needed, starting with a CT scan of the lungs, but also lung function testing, bronchoalveolar lavage, blood tests or a biopsy.

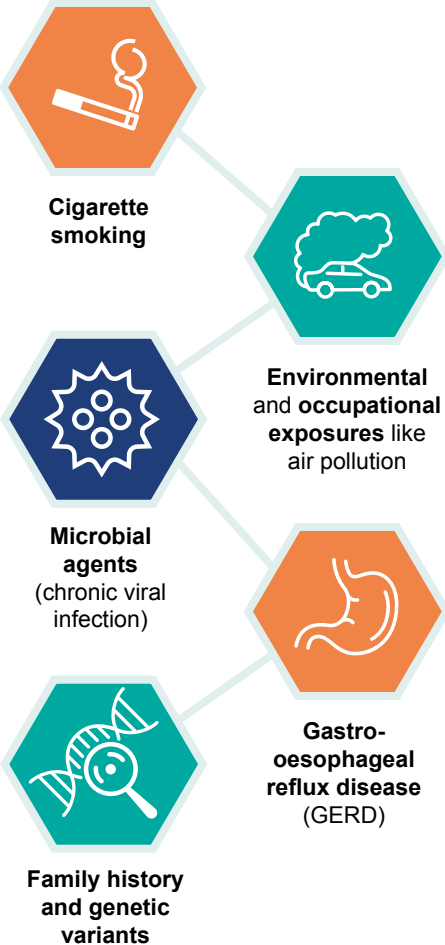
Delayed diagnosis of PF is common and can occur at all stages. On average, it takes 7-8 months for a patient to receive a diagnosis, and in 40% of patients it takes over a year. This may be due to symptoms of PF being like those of other, more common lung or heart diseases. And patients may be slow to consult their general practitioner (GP)/ primary care physician (PCP) once they show 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.

In addition, 37% of patients are misdiagnosed at least once. If you don't feel comfortable with your doctor or treated in the best possible way you might want to get a second opinion.

WHAT ARE THE RISK FACTORS?

We don't know exactly what causes people to develop PF. However, there are several things that increase a person's risk of developing PF.

They include:



WHAT IS A MULTIDISCIPLINARY TEAM?

Multidisciplinary teams (MDTs) are made up of people who specialise in different medical skills. The diagnosis of PF is multidisciplinary, meaning it draws on experienced clinicians, radiologists and pathologists to diagnose PF. Therefore, they need to work together to confirm your diagnosis and provide you with the best possible treatment.

Internationally, this approach is proven to improve the accuracy of diagnosis.

CENTRES OF EXPERTISE FOR INTERSTITIAL LUNG DISEASES AND PF

Centres of Expertise (CEs) are expert centres for the management and care of rare disease (RD) patients. ILD/PF centres of expertise specialise in ILDs. They aim to provide patients with ILD/PF (among others) with the highest standards of care and to deliver timely diagnosis, appropriate treatments and follow up. They also contribute to research and collaborate with different stakeholders, including patient organisations.

What is more, 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. They unite several CEs and provide expert opinions, advice and referral for cross-border care when appropriate.

An ERN on respiratory diseases including fILDs was launched in March 2017: the ERN-LUNG. The ERN-LUNG (find more information [here](#)) is made up of many European Expert Centres ([see here](#)), but not all. We strongly recommend that you ask your local patient group or GP about your nearest nationally recognised centre.

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HOW WILL THE DISEASE PROGRESS?

PF progression varies from person to person and it is impossible to predict exactly how rapidly PF will progress for you.

Most people with PF experience a slow, but steady worsening of their disease. If you have PF 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.

Some patients may experience unpredictable acute (sudden and short-term) worsening of their disease, called acute exacerbations, which can happen at any time. An event like this may be fatal or may leave a person with PF with substantially worsened disease. Often, a person is not diagnosed with PF until their first acute exacerbation. The main risk factor for acute exacerbation of PF is advanced disease.

The danger of an acute exacerbation makes regular monitoring by your doctor essential so they can track how the disease is progressing and see how you are responding to treatment.

OTHER CONDITIONS PATIENTS WITH PF MIGHT HAVE (COMORBIDITIES)

PF can be a debilitating condition as, on top of the adverse effects of PF, many PF patients have other associated conditions, so-called **comorbidities**. These can have a negative effect on the quality of your life and your prognosis. This is why doctors also need to identify and treat any comorbidities you may develop. These conditions can affect the lungs (pulmonary comorbidities) or other parts of your body (non-pulmonary comorbidities).

You can find the definitions of co-morbidities [here](#).



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WHAT TREATMENTS ARE AVAILABLE?

To manage your PF, both pharmacological (drug-based) treatments and non-pharmacological treatments are recommended. You should also have regular check-ups and medical examinations. As soon as you are diagnosed with PF, you should be offered psychological support.

You might want to get in touch with patient groups, which can support you at all stages of your disease. If you experience side effects, e.g. diarrhoea or weight loss, talk to your doctor or specialist nurse.

A PHARMACOLOGICAL TREATMENTS

Decisions about pharmacological treatments depend on the diagnosis and the disease course. Antifibrotic drugs (Pirfenidone or Nintedanib) are recommended for pulmonary fibrosis. They help to prevent tissue scarring and have been shown to slow progression of the disease over time and may increase life expectancy.

For some PF patients, immunomodulation (substances that affect the functioning of the immune system) is generally recommended as initial treatment. If the disease still progresses, antifibrotic therapy such as Nintedanib can be considered.



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 PF – 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](https://www.clinicaltrials.gov) (worldwide), the [EU-IPFF clinical trial finder](https://www.eu-ipff.org/clinical-trial-finder) or the [EU Clinical Trials Register](https://www.eu-clinicaltrialsregister.eu).

B**NON-PHARMACOLOGICAL TREATMENTS**

While a lung transplant is the only effective cure for someone with PF, there are several non-pharmacological treatments which can help improve your quality of life.

• Pulmonary rehabilitation

Includes different activities to relieve the symptoms of PF and improve your overall quality of life such as exercising, nutritional counselling, breathing strategies. Pulmonary education and rehabilitation can be particularly effective in alleviating symptoms, increasing how much you can exercise and reducing how long you may need to stay in hospital.

• Oxygen therapy

Since PF 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 and breathlessness. You may experience this while exercising, resting or sleeping. Therefore, you might be given extra oxygen through a concentrator (a small electric machine), stationary & portable gas or liquid oxygen systems (connected via face masks or nose tubes).

• Lung transplantation

Lung transplantation is the only surgical intervention procedure which can reverse the progression of PF, improve your quality of life and your life expectancy. Some 30% of lung transplants worldwide are performed on PF patients. Unfortunately, less than 5% of all PF 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, severity and progression of PF. You can find them [here](#).

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fILD

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- Palliative and end-of-life care

Palliative care is a holistic approach to care, given to improve the quality of life of patients throughout the course of their disease. It aims to provide you with relief from both the physical pain, and the broader stresses and problems associated with your condition.

For PF patients, key aspects of palliative care include advance care planning, managing side effects of medications and symptoms like cough and breathlessness, psychological support, pulmonary rehabilitation and end-of-life care.

Palliative care can involve physical, mental, social or spiritual activities – depending on your needs and preferences.

- Learning to manage breathlessness and your cough

Being able to manage your cough plays an important role in managing PF because the cough can make you feel isolated and embarrassed unless you can control it.

It is very important that you learn how to reduce, cope with and manage breathlessness as it 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.

You can find more advice on managing breathlessness [here](#).



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QUESTIONS TO ASK MY DOCTOR

It can be difficult to know what to ask your GP, nurse or specialist. It's important to think about the questions you might like to ask as you see different health care professionals throughout your diagnosis and treatment.



Best to ask

The British Lung Foundation has put together [this list](#) of questions patients can ask at different times during their diagnosis and treatment.

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HOW CAN I TAKE CARE OF MYSELF?

It is essential that you manage some parts of this complex condition yourself. This allows you to control your care, set realistic goals and prepare for your future.



● Give up smoking

Or avoid exposure to second-hand smoke, so you can increase oxygen levels in your blood, lower your blood pressure and heart rate and reduce your risk of cancer and heart diseases.



● 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 can include guided exercise with a physiotherapist, but also exercising independently.



● Eat healthily

A balanced, nutritious diet can help you stay as healthy as possible. It can include fruit, vegetables, whole grains, lean meats or low-fat dairy products and should be low in saturated fats, sodium (salt) and added sugar.



● Adapt your home

Adapt your home to ensure safety and enable autonomous living, e.g. by installing a stairlift, handrails or an adapted bathroom. Your national health service, health insurance companies or local agencies may offer grants and assistance.



- **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**

You should also avoid exposure to infections as they can make PF worse. You can do that by washing your hands, avoiding large crowds and public transport and by staying up to date with flu, pneumonia and Covid-19 vaccines.



- **Avoid stress**

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



- **Relaxation and mindfulness**

There are different relaxation techniques that can help you to handle the emotional and psychological challenges that may come with a PF diagnosis. Mindfulness can help you manage the physical and lifestyle challenges of living with PF, and to calm you down when you are distressed, discouraged, scared, or in pain.



- **Yoga and similar activities**

Yoga is beneficial for PF 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.

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

You can find more information on how to take better care of yourself and live with PF [here](#).



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PSYCHOLOGICAL SUPPORT

Psychological support can help you come to terms with your condition and any side effects. In an advanced stage, it 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 from the start as they are usually supportive and can help you manage your PF. It might be difficult, but talking can help you to overcome your fears and difficulties.

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GET IN TOUCH WITH A PATIENT ORGANISATION AND JOIN A PATIENT SUPPORT GROUP

Joining a local support group can be helpful for you, as you can meet other PF patients, their partners and carers and discuss and share your experiences. Support from your family and friends is essential, but reaching out to fellow patients going through the same experience can be particularly beneficial.

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 through the organisations listed on our [website](#).



Finding out as much as possible about PF can help you feel more in control. Patient organisations are a good place to start because they provide information dedicated to people living with PF as well as mutual support.

GENERAL ADVICE FOR CARERS

Caring for someone with PF is a difficult and demanding task which can be both emotionally and physically draining. If you're caring for someone suffering from PF, let your family doctor know so they can advise you on your own health and refer you to specialised support if you need it. Oftentimes, there are national support groups for carers.

Remember to take care of yourself, as the day-to-day life of a caregiver can be challenging. Do not be afraid to ask the doctors and nurses supporting your relative or friend with PF for specific advice on how to best 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 will be able to tell you more.



Caregiving Tips

You can find some suggestions that may help you to be an effective caregiver [here](#).



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