
Action for
Pulmonary Fibrosis
Navigating
pulmonary fibrosis

Contents

Navigating **PF**

Need this information in an alternative format?
Visit our website to explore our other Navigating PF resources, or contact our Support Line:

support@actionpf.org Support line: 01223 785725

Introduction

Being told you have pulmonary fibrosis can feel overwhelming. You may be facing uncertainty, questions and emotions that are difficult to put into words. This booklet is here to help you make sense of what lies ahead and to support you in the days and months to come.

You are not alone. Action for Pulmonary Fibrosis is here for you, alongside your healthcare team, family and wider support network. We are working to make sure that everyone affected by pulmonary fibrosis can access the information, care and support they need, when they need it.

We know there is still more to do. Too many people experience delays in diagnosis, unequal access to care and limited treatment options. That is why we are focused on driving change, improving access to treatments, investing in research and strengthening support for people living with pulmonary fibrosis across the UK.

This booklet is part of that commitment. It has been shaped by the experiences of people living with pulmonary fibrosis and those close to them. It aims to give you clear, reliable information and to help you feel more informed and more in control.

Above all, we want you to feel supported. We are here to help you find your way forward, with the right information, the right support and a community that understands.

Best wishes,

Daniel Saxton
Chief Executive Officer

About Action for Pulmonary Fibrosis

Action for Pulmonary Fibrosis (APF) is the UK's leading charity dedicated to supporting people affected by pulmonary fibrosis (PF), driving research and shaping policy.

We are rooted in the lived experiences of patients, families, carers and clinicians and united by a shared belief that people with pulmonary fibrosis deserve better.

We are committed to making a meaningful difference for everyone affected by pulmonary fibrosis by bringing hope and lasting support. Our work spans four areas:

Support

Providing trusted, compassionate support for anyone affected by pulmonary fibrosis.

Awareness

Raising the profile of pulmonary fibrosis and tackling stigma and misinformation.

Advocacy

Campaigning for faster diagnosis, better care and health equality.

Research

Funding and influencing science that improves lives and brings hope.

If you or someone you know is affected by pulmonary fibrosis, we're here to support you. We have a range of services that are free to access. Please find out more at **www.actionpf.org**

- **Support line**: our support line is available Monday to Friday, 9am to 5pm on 01223 785 725. Alternatively, you can email **supportline@actionpf.org**.

- **Support groups**: there are over 100 support groups across the UK offering emotional support, helpful information, practical guidance and signposting for anyone affected by pulmonary fibrosis.

- **Information resources**: we provide a variety of information from telling people you have pulmonary fibrosis, to the benefits you might be able to access if you have pulmonary fibrosis. Our resources are available on our website or in print to order.

- **Webinar series**: our Talking PF webinars include in depth talks, patient stories and insight from healthcare professionals and include topics such as managing fatigue and emotional wellbeing.

Thanks to the generosity of our donors, funders, volunteers and partners, APF is able to reach more people impacted by pulmonary fibrosis every year, offering hope and support so that no one has to face pulmonary fibrosis alone.

What is pulmonary fibrosis?

The information in this booklet is intended as general information only. It is not a substitute for medical advice. Please follow the advice of your medical team.

1. What is pulmonary fibrosis?

The term **pulmonary fibrosis** (PF) describes scar tissue in the lungs.

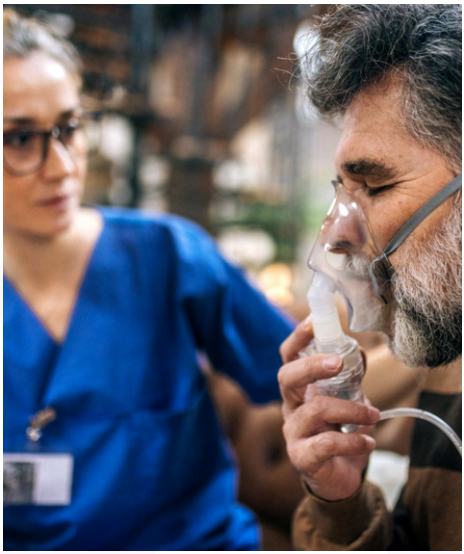
Pulmonary → refers to the lungs

Fibrosis → refers to scarring

Pulmonary fibrosis is a condition where the lungs become scarred over time, making it difficult for the lungs to function properly. Symptoms often include shortness of breath, a dry cough, fatigue and weight loss. For many people, pulmonary fibrosis may get worse over time.

Pulmonary fibrosis is a type of interstitial lung disease (ILD). There are many different types of pulmonary fibrosis and ILDs. Not all pulmonary fibrosis will get worse and the rate of progression can vary significantly between different types and individuals. Your medical team may use different terminology to describe your pulmonary fibrosis. Please reach out to your healthcare team for clarification if you are unsure.

- **Non-progressive pulmonary fibrosis** – the result of a one-off event. It does not get worse over time.
- **Progressive pulmonary fibrosis (PPF)** – pulmonary fibrosis that gets worse over time.
- **Idiopathic pulmonary Fibrosis (IPF)** – the cause of the pulmonary fibrosis is unknown. This is the most common type of pulmonary fibrosis.
- **Interstitial lung disease (ILD)** – a group of over 200 diseases that affect the interstitium (including autoimmune ILDs, hypersensitivity pneumonitis and sarcoidosis). The interstitium is the space in the lungs between the air sacs (alveoli) and the blood vessels.



The following sections will talk you through the causes, signs, symptoms, diagnosis, treatments and life expectancy of pulmonary fibrosis.

2. How does pulmonary fibrosis affect the lungs?

The **lungs** have two main functions:

1. Bring oxygen into the blood.
2. Remove carbon dioxide from the body.

Healthy lungs are spongy and stretchy so air can easily move in and out of them. When you breathe in, air enters your airways and travels down into the air sacs (alveoli) in your lungs. Oxygen and carbon dioxide move between the alveoli and the blood. The space between the alveoli and blood vessels is called the interstitium.

With pulmonary fibrosis, scar tissue forms in the interstitium. This scar tissue is thicker and makes the lungs stiff and less elastic. This can lead to shortness of breath, reduced lung capacity and if severe, impaired movement of oxygen between the air and the blood.

Diagram one:

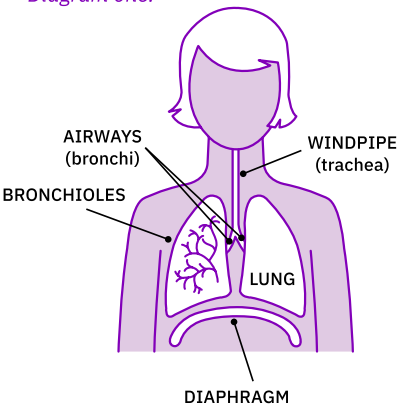
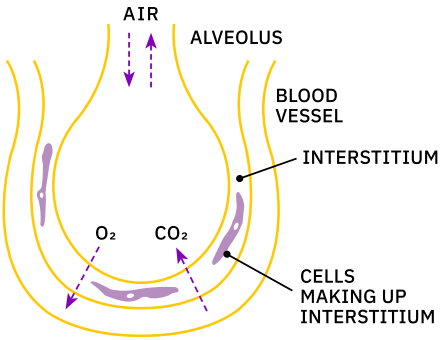


Diagram two:

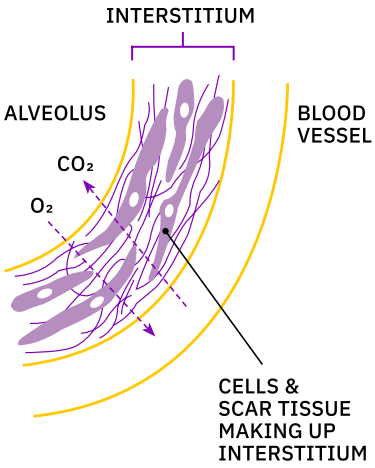


3. What causes pulmonary fibrosis?

Research into pulmonary fibrosis is still developing. From what research into pulmonary fibrosis has learnt so far, there are two processes that can lead to scar tissue forming in the lungs:

1. Post inflammatory scarring
Inflammation in the lungs happens when something causes irritation or injury to the lungs. This injury can leave a scar. For example, radiotherapy.

Diagram three:



2. Lung cells aging too quickly
Pulmonary fibrosis can happen when cells in the lungs age too quickly. Accelerated ageing is caused by a combination of genes and environment. Genetics can affect how cells age, which is why pulmonary fibrosis sometimes runs in families. Prolonged inhalation of smoke or dust, for example from tobacco or occupational exposures earlier in life, can also cause accelerated ageing.

You can find out more about pulmonary fibrosis research and how you can get involved on our website, **actionpf.org**

Risk factors

From our current understanding, there is no single cause of pulmonary fibrosis. Instead, we talk about different risk factors. Risk factors can lead to inflammation or lung cells aging too quickly.

Risk factors can include:

- older age
- family history (the genes inherited from your parents)
- breathing in something that irritates the lungs, such as smoke or dust – this will usually happen repeatedly over a long time
- some medications, for example Nitrofurantoin and some anticancer drugs
- other conditions, such as autoimmune rheumatic diseases
- Gastro-oesophageal reflux disease (GORD).

4. What are the signs and symptoms of pulmonary fibrosis?

The signs and symptoms of pulmonary fibrosis vary between individuals. They are also likely to change if the disease progresses.

Symptoms: Your experience of pulmonary fibrosis

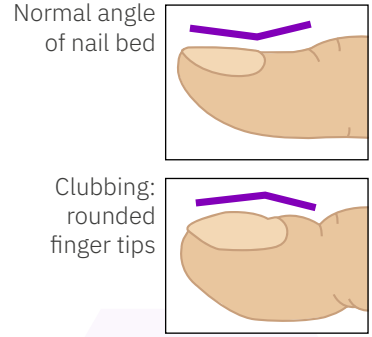
Common symptoms can include:

- shortness of breath (breathlessness)
- dry cough
- tiredness (fatigue)
- unexplained weight loss

Signs: Abnormalities found on physical examination

A common sign is hearing crackles when listening to your chest with a stethoscope. This sounds similar to Velcro being pulled apart.

Another sign can be clubbing of fingers and toes (swollen and rounded finger and toe tips – see diagram opposite).



5. How is pulmonary fibrosis diagnosed?

The healthcare professionals involved in your diagnosis will gather information in different ways. This can include:

- **Asking questions about your symptoms and your personal and family medical history**. This will be a very full and detailed history and will enable them to decide what tests might be needed.
- **Asking questions about situations** where you might have breathed in something that can irritate the lungs at work.
- **A physical examination**. This could include listening to your breathing with a stethoscope (both front and back of your chest) and a full body examination.
- **Further tests** normally include pulmonary (lung) function tests or a high-resolution computerised tomography (HRCT) scan.
- **Blood tests**, which may be required to be repeated periodically.

The results of these investigations, along with a discussion at a specialist ILD multi-disciplinary meeting (MDT), will help your healthcare team to make a diagnosis. Sometimes, additional procedures may be needed. These may include:

- a **bronchoscopy** (a camera into the lungs)
- or **lung biopsy** (taking a sample of lung tissue). Your healthcare team will discuss this with you if they think it is necessary.

There are several types of pulmonary fibrosis. Your medical team will use information from your diagnostic test results, alongside a detailed and full medical history, to decide which type of pulmonary fibrosis you have. Sometimes this only becomes clear by observing if and how the disease changes over time.

In most cases, your medical team will check if your disease changes. This is to see if your pulmonary fibrosis is progressing. It's important to know this to help decide on the right support and treatment for you.

6. How is pulmonary fibrosis treated?

Currently, we can't stop pulmonary fibrosis. However, some treatments can slow down the rate of further scarring. Other treatments can help to manage your symptoms. Not all treatments are suitable for everyone, so your medical team will discuss the best treatment options for you.

Some people have pulmonary fibrosis that is not progressive and might not need treatment.

Possible treatments to reduce further scarring include:

- Antifibrotic medication (nintedanib or pirfenidone).
- Immunosuppressants and steroid medications are sometimes used to treat inflammation.
- Lung transplantation.

Possible symptom management includes:

- Pulmonary rehabilitation.
- Techniques to manage breathlessness and cough (including medication and non-medication methods).
- You may have a secondary condition that has the potential to increase symptoms, such as acid reflux, which may require medication management.
- Oxygen therapy.
- Palliative care, which helps you to manage your difficult symptoms enabling you to live well with a life-limiting condition.
- Support to maintain a healthy lifestyle.

What does a healthy lifestyle mean?

Everyday choices can help you live well with pulmonary fibrosis. This might include:

- stopping smoking
- staying active
- getting support for mental health
- eating well
- keeping up to date with flu, pneumococcal and COVID-19 vaccines if eligible. Please discuss this with your clinical team.

7. How does pulmonary fibrosis progress?

Pulmonary fibrosis can be a progressive condition. If you are unsure if your pulmonary fibrosis is progressive, please discuss this with your clinical team, as not all pulmonary fibrosis progresses.

Your symptoms may stay the same for a long time, or you may deteriorate quickly. Your healthcare team will monitor your pulmonary fibrosis to see if it progresses. This may be done by repeating a series of tests including:

- lung function tests
- CT scans.

They will then adjust your treatment options accordingly.

If you feel that your symptoms are worsening, it is important that you do not ignore this and that you contact your clinical team for an earlier review. Some potential signs that your pulmonary fibrosis is progressing include:

- increased breathlessness
- rapid weight loss
- increased fatigue
- any other significant changes to your ability to navigate your daily life.

8. Life expectancy

Reading about life expectancy can be difficult. You might want to read this information with someone you trust and who can support you. The APF support line is also here for you.

Contact our support team:
supportline@actionpf.org Support line: 01223 785725

Progressive pulmonary fibrosis is a terminal disease (sometimes also called 'life-limiting'). This means there is currently no cure. Many people who are diagnosed with the disease will eventually die from it.

Many factors affect how long a person may live with pulmonary fibrosis, including:

- age
- the severity of lung scarring
- how quickly your pulmonary fibrosis gets worse (if your disease is progressive)
- your other health conditions
- if you get infections
- your lifestyle (e.g. if you smoke).

It's important to note that life expectancy research needs updating. It may no longer be accurate.

Previous research looked at the average life expectancy of people with idiopathic pulmonary fibrosis (IPF). Life expectancy was suggested to be 3-5 years from diagnosis. However, researchers found this number before antifibrotic medications were widely used.

At this time, diagnosis also took longer and less was known about the disease.

There is also little information on the life expectancy of people with other types of pulmonary fibrosis. However, our current understanding is that people with progressive pulmonary fibrosis may follow a similar disease trajectory as those with IPF.

There have been improvements in pulmonary fibrosis treatment and care. However, it's important to acknowledge there is still work to do. This will ensure timely access to care and better treatments.

Researchers are working to understand why pulmonary fibrosis happens and to find more effective treatments.

It's understandable to have questions or concerns about your life expectancy.

Your medical team are there to support you and answer questions about your personal situation. They can also help you manage your symptoms and live a healthy lifestyle.

This can improve your health and chances of living well with pulmonary fibrosis.

Your healthcare team

The healthcare professionals in a team will vary. Your medical team might not have all the professionals listed below.

ILD Consultant: a senior doctor who will lead your medical treatment. They work alongside other doctors and health professionals to help you make decisions about your treatment.

ILD nurse: a specialist nurse who will support your ongoing care. They can advise on all aspects of pulmonary fibrosis including symptom management and information on your condition.

ILD pharmacist: a specialist pharmacist who will advise on medication.

Physiotherapist: they will support symptom management including breathlessness and getting around and advise on all aspects of physical well-being.

Occupational therapist: they will support you to manage everyday activities alongside your symptoms.

Community respiratory team: a team of different professionals who deliver care at your home or local facilities.

Palliative care team: a specialist team who will support you to have the best quality of life at any point after your diagnosis.

It may be helpful for you to write down the contact details for each member of your healthcare team.

Questions you may want to ask

Diagnosis and treatment:

- Do you know why I have developed pulmonary fibrosis? Do you know how advanced my pulmonary fibrosis is?
- Is my pulmonary fibrosis progressive?
- What treatments can I have?
- What's the aim of the treatment?
- If I choose not to have the treatment, what would happen?
- Can I be treated privately or by a different hospital?
- Are there any research studies I can take part in?
- What is my follow up plan? Who is responsible for my follow up plan?

Living with pulmonary fibrosis:

- Where can I get more information about pulmonary fibrosis? Is there any support available for me and my family?
- What can I do to help myself?
- Is there a patient support group nearby that you could refer me to?
- Is there a healthcare professional who I can phone from home if I am worried about anything?
- Where can I learn more about managing my symptoms?
- What holistic supportive care is available to me?
- Can you tell me more about pulmonary rehab and if it is suitable for me?
- What vaccinations am I eligible for?

Coming to terms with your diagnosis

There is no right way or wrong way to respond to a life-changing diagnosis.

How do I deal with these emotions?

- Be nice to yourself. It's okay not to feel okay.
- Talk to friends and family.
- Pace yourself – you can't expect yourself to process your emotions instantly.
- Look after your body – your emotional and physical health are linked.
- Make new hopes for what you can achieve instead of focusing on what you can no longer do.
- Try to take it day by day. Some days you may even feel you need to manage hour by hour, and that is okay.
- Ask for professional help if you need it.

How can I support someone who has received a life changing diagnosis?

When someone reaches out emotionally, they are often not looking for facts or solutions. They want to feel heard and understood.

Try to:

1. Meet people where they're at, not where you think they should be.
2. Listen without interrupting.
3. Respond with empathy rather than solutions.
4. Invite them to share more.

You can find more information about caring for someone with pulmonary fibrosis on our website **actionpf.org**

Telling friends and family

There's no right or wrong way to tell your family and friends you have pulmonary fibrosis. You don't have to tell anyone if you don't want to, but talking about your diagnosis can help you feel more supported. If you find it difficult to tell people, these ideas might make it a little easier.

When do I tell people?

Tell people when it feels right for you and when you are emotionally ready to talk about your diagnosis. Some people want to learn more about pulmonary fibrosis and process their diagnosis on their own before telling others.

How do I tell people?

How you choose to tell people is very individual. You might want to try:

- Having one-to-one conversations with individual people.
- Telling one person who acts as a 'messenger' to tell others.
- Talking to a group of family and friends at the same time.
- Bringing family and friends to medical appointments. Healthcare professionals can help to explain your diagnosis.
- Giving your family and friends something to read so you don't have to explain everything. We have online and printed versions of this booklet that may help people understand what pulmonary fibrosis is.

> There is no right or wrong way of doing it. Initially we found it very difficult because of course we were in shock and we didn't really know that much about the illness itself anyway. We decided to tell our closest family and friends first. We decided that we were going to be open and honest. We told them how it affected us, because it does affect both of us, and how it made us feel.
>
> **George, living with pulmonary fibrosis**

Where do I have the conversation?

Choose a place where you feel comfortable and able to talk openly. This might be at home over a coffee or whilst out for a walk in a quiet area.

What should I say?

- If you're unsure where to start, you could use a simple phrase to describe pulmonary fibrosis overall. This could be something like: 'I have a condition called pulmonary fibrosis. This means my lungs have scar tissue, which stops them from working properly. The amount of scar tissue might increase over time.'
- Think about the most important things you want them to know. Is it how pulmonary fibrosis affects your daily life? Is it that you are worried about the impact of your diagnosis on your work or family?
- Consider your own needs. It can be tempting to downplay your diagnosis or put a positive spin on it, but tell people how you feel so they can support you.
- Let people know how they can help and support you. Whether that's practical help with things like shopping, or having someone to talk to when needed.
- You can invite them to support groups to learn more about pulmonary fibrosis and feel supported by a community. There is a specific support group for carers, family members and friends, but they can also attend the regional support groups that are open to everyone.

How should I expect the conversation to go?

- Telling people about your diagnosis might make you feel very emotional, and this is okay. Let yourself feel your emotions and let others know how you feel.
- Don't worry too much about silences. Some people need time to take in the information before responding, especially in an emotional situation.
- Consider their potential reactions and how you might deal with this. For example, some people might be very emotional and others may not say much.

- When you tell people you have pulmonary fibrosis, some people might do their own research to find treatments or cures. Whilst they are well-meaning, there is a lot of information out there that is not accurate. You should always ask your healthcare team before trying any suggestions.

If someone does want to read more about pulmonary fibrosis, you can direct them to recognised websites such as Action for Pulmonary Fibrosis, the NHS, or ones related to your specialist clinic.

- If someone has suggested you try something you don't want to try, you can tell them that you need to discuss this with your healthcare team first, as not everything is suitable for everyone.

Explaining pulmonary fibrosis to children

Talking about pulmonary fibrosis can be difficult but it's usually best to be honest about your diagnosis with the children in your life, when you feel ready to do so. They might be able to tell that something has changed and this uncertainty can lead to them making assumptions and worrying.

Give reassurance

- It can be helpful to reassure children that this is not usually something that happens in childhood and only a very small number of adults get it.
- You can also reassure them they will still be safe and looked after, whatever happens to you.

Where and when?

- Have a conversation when they can think about what you've said and ask questions. You might want to avoid telling them just before they go to school or before bedtime.
- Choose a place where they feel safe and comfortable.

> "When I found out I had pulmonary fibrosis, I told my children and we dealt with it using a lot of humour. They now tell me it took some of the anguish out of the situation. It's not what I'd recommend to everyone, but it got us through."

Andy, Birmingham

What to say

- What you say might depend on their age and understanding. If you're telling multiple children with significant age gaps, it might be helpful to tell them separately. This way you can adjust what you say to their level of understanding.
- For many children, you can explain pulmonary fibrosis by saying that your lungs are not working as well as most people's, making breathing harder for you.
- You might want to describe how this affects your daily life so they understand that you might need to do some things differently.
- Be honest and use straightforward language so they don't misinterpret what you're saying.
- You don't have to tell them everything in one conversation.
- It's okay to say 'I don't know' to something they ask you.

Get support

Involve another adult that the children are close to. If you are their grandparent, you could talk with the children and their parents. This can help to build a support network, so you and the children have someone else to go to with questions or worries.

If you need support telling your family and friends about your diagnosis, call our support line:

supportline@actionpf.org Support line: 01223 785725

Support available

At Action for Pulmonary Fibrosis, we have a range of support available for anyone affected by pulmonary fibrosis.

Call our Support Line

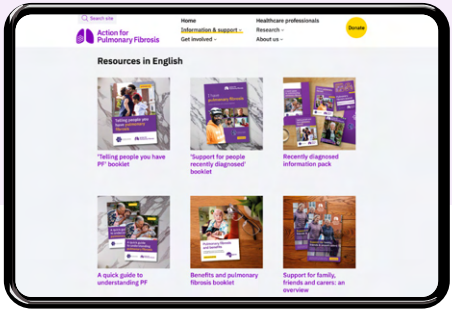
Our Support Line is available on **01223 785 725**, 9am to 5pm, Monday to Friday. If you ever feel like you need support, please call our support line. If something's on your mind, we're here to listen and help whenever you need us.

Not ready to call?
You can email us on **supportline@actionpf.org** instead.

Find a Support Group

There are over 100 local support groups across the country, as well as several online groups run by APF. Connecting with others who understand can make a real difference. You can find a support group that feels right for you by visiting our website, **actionpf.org**.

Learn more about pulmonary fibrosis



We have a range of printed and online information about living well with pulmonary fibrosis. To find out more, visit our website or call our Support Line.

Join our online webinars

Talking **PF**
Webinar series

In our Talking PF sessions, our panel of experts explore different PF-related topics in depth. Hear from healthcare professionals and people affected by pulmonary fibrosis and have the opportunity to ask the experts your questions. Find out more on our website or call our Support Line.

Join our Involvement Network

You can help shape our services and research into pulmonary fibrosis by taking part in our involvement activities and sharing your experiences. Email **involvement@actionpf.org** to find out more and hear about opportunities.

Stay in touch

We would love to stay in touch with you about our work. Please sign up to be sent regular information, including our e-newsletter and Insider magazine. Register online at **www.actionpf.org/information-and-support/signup** or call the Support Line.

For our latest news and updates, you can also follow us on our social channels:

@actionpulmonaryfibrosis
@actionpulmonaryfibrosis
@action-for-pulmonary-fibrosis

Scan this QR code to visit our website, or alternatively type **actionpf.org** into your browser.


Together, we will make the difference.
For Every Breath
For Every Journey
For Every Future

Action for Pulmonary Fibrosis

Here to help... Contact our support team:

supportline@actionpf.org

Support line: 01223 785725

actionpf.org **01733 839642** **info@actionpf.org**

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