A quick guide to understanding pulmonary fibrosis

www.actionpf.org

Interstitial Lung Disease Interdisciplinary Network

Action for Pulmonary Fibrosis

Contents

Here to help...

Email our support team:
support@actionpf.org

Support line:
01223 785725

What is pulmonary fibrosis?

You might be told by your medical team you have 'lung scarring' or 'pulmonary fibrosis'. In this booklet, we talk about pulmonary fibrosis.

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

Pulmonary → **refers to the lungs**

Fibrosis → **refers to scarring**

Scar tissue can make it difficult for the lungs to work normally.

Some people with interstitial lung disease (ILD) may develop pulmonary fibrosis.

Interstitial lung disease: a group of diseases that affect the interstitium. The interstitium is the space in the lungs between the air sacs (alveoli) and the blood vessels. (See diagrams 2 and 3).

How do the lungs work?

The lungs have two main functions:

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

Healthy lungs are spongy and stretchy so air can move in and out of them. There's a network of tubes that carries air through the lungs. At the end of the tubes are small sacs called alveoli, surrounded by blood vessels.

Oxygen and carbon dioxide move between the alveoli and the blood. The space between the alveoli and blood vessels is called the interstitium. This space contains different cells that help the lungs work properly.

Diagram one:

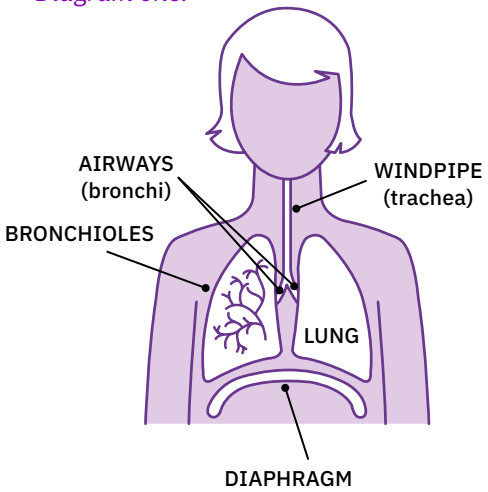
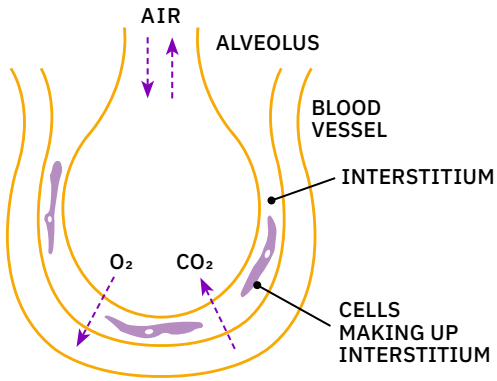


Diagram two:



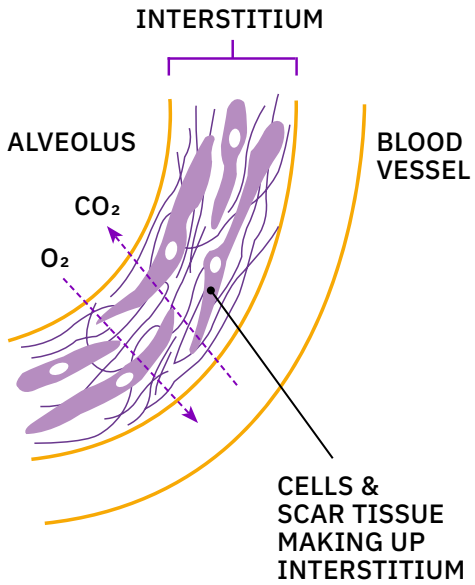
What happens in the lungs?

Scar tissue forms in the interstitium and this area becomes thicker. This makes the lungs stiff and less stretchy, which can make you feel short of breath (breathless).

Several different mechanisms can cause a cough. Change to the structure of the lungs is one potential cause.

Scar tissue makes it more difficult for oxygen to move between the alveoli and the blood. Because of this, there is less oxygen available for the body to use.

Diagram three:



Why does this happen?

We don't know exactly what happens in the lungs when someone has pulmonary fibrosis. There's more research looking into this. From what we have learned so far, two processes can lead to scar tissue in the lungs:

1 Inflammation

Inflammation is when something causes irritation or injury to the lungs. Some types of ILDs can cause inflammation in the lungs. These ILDs include, but are not limited to, sarcoidosis, autoimmune disease-associated ILDs, and hypersensitivity pneumonitis. Inflammation can lead to scar tissue in the lungs (pulmonary fibrosis).

2 Lung cells age too quickly

Pulmonary fibrosis can happen when cells in the lungs age too quickly. One of the normal functions of these cells is to repair damage. Ageing lung cells are unable to repair normally. Instead, they cause scar tissue to form. So, factors that cause lung cells to age too quickly increase the risk of pulmonary fibrosis. This includes genes (inherited from your parents), your environment (e.g. dust or smoke that your lungs have breathed in previously), and other risk factors below. Read about risk factors in the next section.

Damage to lung cells is normal. We breathe in over 10,000 litres of air each day. Air may contain dust, pollutants, viruses, bacteria or small amounts of stomach content. These can damage the delicate lining of the lungs. Cells that are lost to damage must be replaced. When a person has pulmonary fibrosis, their body is not able to replace cells as it normally would.

Risk factors

From what we have learned so far, there is no single cause of pulmonary fibrosis. Instead, we talk about different risk factors. Risk factors can lead to inflammation or lung cells ageing too quickly. This can increase the risk of scarring.

Risk factors can include:

- Ageing
- 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
- Other conditions, such as rheumatoid arthritis
- Gastro-oesophageal reflux disease (GORD)

Many people with pulmonary fibrosis have a combination of risk factors. Your medical team will try to find your risk factors. This helps them to diagnose the correct type of pulmonary fibrosis.

Types of pulmonary fibrosis

There are several types of pulmonary fibrosis. Your medical team will use information from different sources to decide which type of pulmonary fibrosis you have. You can read more about this in the **Diagnosis** section.

Your medical team might say whether your pulmonary fibrosis is progressive or non-progressive. Progressive pulmonary fibrosis gets worse over time. Non-progressive pulmonary fibrosis is the result of a one-off event. It does that does not get worse 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 determine decide the right support and treatment for you.

Diagnosing pulmonary fibrosis

A doctor will diagnose pulmonary fibrosis. They are often part of a specialist ILD multi-disciplinary team (MDT).

Multi-disciplinary team:

A team of different healthcare professionals.

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.
- Asking questions about situations where you might have breathed in something that can irritate the lungs.
- A physical examination. This could include listening to your breathing with a stethoscope.
- Further tests which could include pulmonary (lung) function tests or a computerised tomography (CT) scan.

CT scan:

Combines lots of X-rays to create a more detailed image.

Sometimes, you might need other procedures to help determine the type of lung disease. These procedures might include a bronchoscopy (a camera into the lungs) or lung biopsy (taking a sample of lung tissue). The team will discuss this with you.

The information your medical team find will help them make the most likely diagnosis.

Signs and symptoms

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

Signs:

Information that healthcare professionals find

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

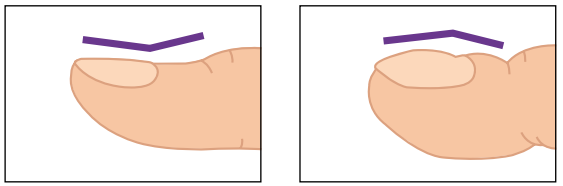
Symptoms:

Your experience of pulmonary fibrosis

Common symptoms can include:

- Shortness of breath (breathlessness)
- Dry cough
- Tiredness (fatigue)
- Unexplained weight loss
- Clubbing of fingers and toes (swollen and rounded finger and toe tips – see diagram below)

Diagram four:



Normal angle of nail bed

Clubbing: rounded finger tips

Treatments

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 treatments 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)
- Lung transplantation
- Immunosuppressants and steroid medications are sometimes used to treat inflammation

Possible symptom management includes:

- Pulmonary rehabilitation
- Techniques to manage breathlessness and cough
- Other medications to manage acid reflux, breathlessness or a cough
- Oxygen therapy (this involves breathing air that has more oxygen than normal air)
- Palliative care, which helps you to manage your symptoms
- 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

> Click here to find out more information.

Research

Research studies and clinical trials help us to:

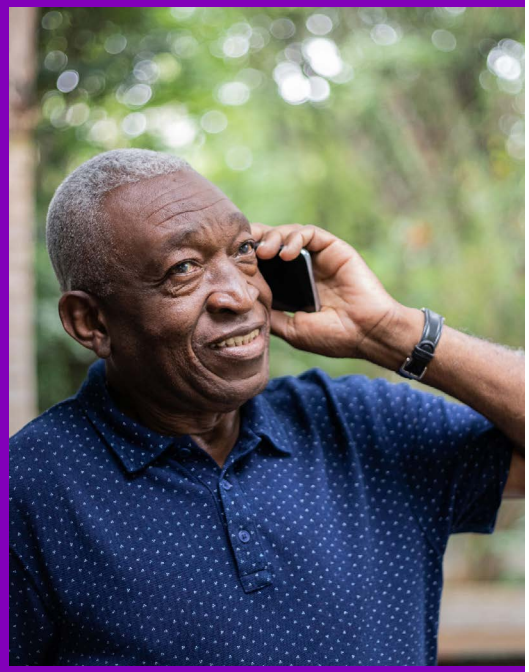
- Learn about pulmonary fibrosis
- Find new treatments
- Improve clinical services and care

People living with pulmonary fibrosis tell us that being involved in research can be a rewarding experience. By getting involved, you can:

- Gain a better understanding of pulmonary fibrosis
- Build links with medical teams
- Access new treatments
- Help others with the disease

Speak to your ILD team about opportunities to get involved in research. Visit **actionpf.org** for more information.

Stages of pulmonary fibrosis



Pulmonary fibrosis can be a progressive condition, meaning it can get worse over time. Not everyone has progressive pulmonary fibrosis.

Progression can look different for everyone. Your symptoms may stay the same for a long time or get worse quickly.

Life expectancy

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

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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:

- The severity of lung scarring when you are diagnosed
- 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.

There have been huge 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. Read more about living well in the Treatment section.

Action for Pulmonary Fibrosis is a patient-driven charity. Our vision is to stop pulmonary fibrosis so everyone affected has a better future.

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If you need this information in a different format such as large print, easy read or another language, please contact us.

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