



Pulmonary fibrosis

What it is and how it affects your breathing

What is pulmonary fibrosis?

Pulmonary fibrosis is a term that covers many different conditions that cause scar tissue to build up in your lungs. This build-up of scar tissue, which makes your lungs stiff, is called fibrosis.

Pulmonary fibrosis is a type of interstitial lung disease (ILD). 'Interstitial' means the disease affects the interstitium, the lace-like network of tissue that supports the air sacs in your lungs. There are more than 200 different ILDs.

Some types of pulmonary fibrosis have an identifiable cause. But for many types, a definite cause cannot be found.

In ILDs, there can be scarring in your lungs or inflammation in your lungs. Some ILDs mostly cause scarring, some mostly cause inflammation. But often there is a combination of these processes going on. Which of these processes is dominant can determine what kind of treatment you may have.

The treatment and outlook for different types of pulmonary fibrosis vary considerably, so if you're not sure about your diagnosis, check with your doctor or nurse for the exact name of your lung condition.

All types of pulmonary fibrosis are rare. We have specific information about those seen most often in other PDFs:

- **idiopathic pulmonary fibrosis or IPF**
- **hypersensitivity pneumonitis formerly called extrinsic allergic alveolitis**
- **pneumoconiosis, also known as an occupational interstitial lung disease**
- **pulmonary fibrosis associated with connective tissue and autoimmune diseases**
- **drug-induced pulmonary fibrosis**

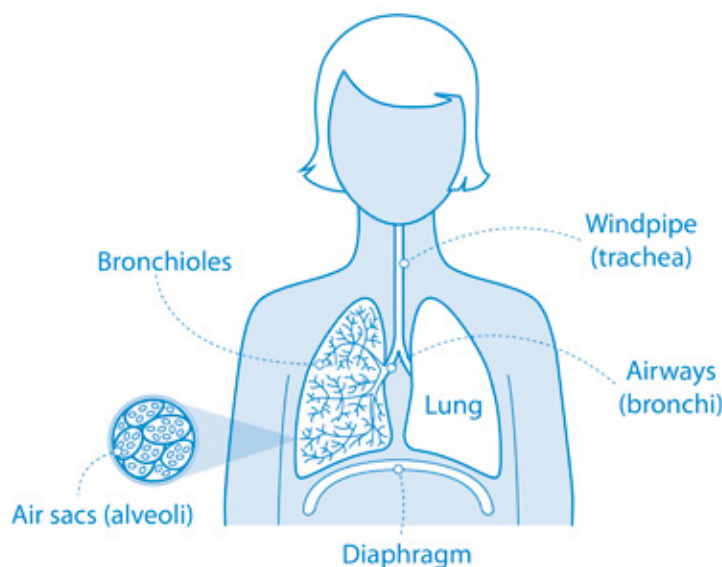
Although we do not always know what causes pulmonary fibrosis, we do know it is *not* a form of cancer or cystic fibrosis, and it is *not* contagious. Cystic fibrosis is not a type of ILD or pulmonary fibrosis.

Sarcoidosis is a relatively common type of disease that usually affects the lungs. It can sometimes cause pulmonary fibrosis but we don't know why. To find out more about this condition, go to blf.org.uk/sarcoidosis or call our helpline.

How does pulmonary fibrosis affect your breathing?

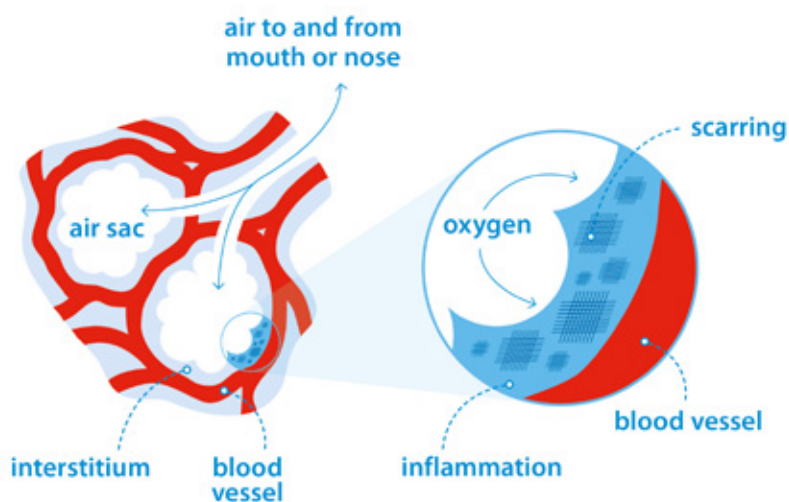
Pulmonary fibrosis scars your lungs and so reduces the efficiency of your breathing. Scarring causes your lungs to become stiffer and less elastic so they are less able to move and take oxygen from the air you breathe.

Each time you breathe in, you draw air into your nose or mouth, down through your throat and into your windpipe, also called your trachea. Your windpipe splits into two smaller air tubes called bronchi, which go to your lungs. The air passes down the bronchi, which divide again and again, into thousands of smaller airways called bronchioles.



The bronchioles have many small air sacs, called alveoli. Inside the air sacs, oxygen moves across paper-thin walls to the capillaries - tiny blood vessels - and into your blood. The air sacs also pick up the waste gas, carbon dioxide from your blood, ready for you to breathe it out.

If you have pulmonary fibrosis, scarring affects the air sacs in your lungs. The air sacs are supported by the interstitium, a network of tissue, a bit like lace. Scarring fills in the gaps between and around the air sacs and limits the amount of oxygen that gets into the blood.



As scarring increases, your lungs are less able to expand to allow you to take deep breaths and the level of oxygen in your blood can start to drop. Breathing may feel like harder work and you can feel breathless from everyday activities like walking.

What causes pulmonary fibrosis?

In a small number of pulmonary fibrosis cases, it's possible to identify a specific cause. Some causes are:

- being exposed to certain types of dust - including wood or metal dusts or asbestos
- being exposed to allergens - such as bird feathers or mould
- a side effect of a drug

Some types of pulmonary fibrosis occur when you have another condition such as rheumatoid arthritis or scleroderma.

In a very few cases, two or more members of a family may develop pulmonary fibrosis. But current research suggests the genetics of familial interstitial lung disease is complex and there is no clear inherited predisposition to develop pulmonary fibrosis.

In most types of pulmonary fibrosis a specific cause cannot be found. One of the most frequently occurring forms of fibrosis is called idiopathic pulmonary fibrosis (IPF). The word 'idiopathic' means there is no known cause.

Researchers recently set out some common types of interstitial lung disease, grouped by what causes them, in the table below. It is not agreed by all doctors and there's lots of research underway to improve our understanding of the causes.

Classification of common interstitial lung diseases

Idiopathic disorders	<ul style="list-style-type: none">• Idiopathic pulmonary fibrosis (IPF)• Acute interstitial pneumonia (AIP)• Idiopathic non-specific interstitial pneumonia (NSIP)• Sarcoidosis
Connective tissue and autoimmune disease	<ul style="list-style-type: none">• Scleroderma / progressive systemic sclerosis• Systemic Lupus erythematosus (Lupus)• Rheumatoid arthritis• Polymyositis / dermatomyositis
Occupational and environmental	<ul style="list-style-type: none">• Inorganic dust• Organic dust• Gases and fumes• Radiation
Drug-induced	<ul style="list-style-type: none">• Chemotherapeutic agent• Radiation therapy• Antiarrhythmics• Antibiotics• Anticonvulsants

Infections	<ul style="list-style-type: none"> • Viral infections • Bacterial infections
Genetic / inherited	<ul style="list-style-type: none"> • Familial pulmonary fibrosis • Hermansky-Pudlak syndrome

Adam Wallis and Katherine Spinks: The diagnosis and management of ILDs, British Medical Journal 2015

What are the symptoms?

The different types of pulmonary fibrosis have similar symptoms, which is one of the reasons why you need to take tests and speak to a specialist consultant to find out exactly which type you have.

The first symptom a lot of people notice is **getting out of breath** when they're exerting themselves, such as climbing a hill or stairs. But you might feel constantly short of breath, and not just when you're moving about.

Several forms of pulmonary fibrosis usually occur after the age of 60, so you might think you're getting breathless because you are not as young as you were. This breathlessness will get worse over time, if not treated. Your shortness of breath may also be affected by other long-term conditions, such as chronic obstructive pulmonary disease (COPD), heart disease and being overweight.

A **cough that doesn't go away** and **feeling very tired** all the time are two other symptoms of pulmonary fibrosis. Some people with pulmonary fibrosis can also have a fever, lose weight or experience muscle and joint pain.

You might not think that a lung condition could affect your fingers and toes, but this is often a distinctive sign of pulmonary fibrosis. It's called **clubbing**, and you might notice:

- your nails feel too soft or as if they are coming loose
- the shape of your nails changes
- the tips of your fingers or toes bulge out

Clubbing can also occur with various other lung conditions, and as a result of heart or liver disease.

Get in touch with us to find support near you.

Helpline: **03000 030 555**

Monday to Friday, 9am-5pm

Ringing our helpline never costs more than a local call and is usually free, even from a mobile.

helpline@blf.org.uk

blf.org.uk

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Registered charity in England and Wales (326730), Scotland (038415) and the Isle of Man (1177)



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We value feedback on our information. To let us know your views, and for the most up to date version of this information and references, call the helpline or visit **blf.org.uk**