

Other possible causes for your symptoms may need to be ruled out. This includes things like a chest infection, blood clot or a heart problem. You will have blood tests and an ECG. The doctor may suggest more investigations depending on your case.

If you are more breathless, and your oxygen levels are low, you will probably be admitted to hospital. It generally takes some time for an acute exacerbation to be diagnosed. This is because the symptoms and findings on a chest X-ray look like a patient with a chest infection, pneumonia or pulmonary oedema (fluid on the lungs).

Treatment for an acute exacerbation normally includes:

- oxygen and
- medications to help with symptoms.

Steroids and antibiotics are normally prescribed and sometimes other medications, like drugs that suppress the immune system.

How can I reduce the risk of having an acute exacerbation?

There is no clear evidence on the best way to reduce the risk of acute exacerbations. However, you can reduce the risks if you do the following.

Get vaccinated

This reduces your risk of infection. Relevant vaccines include:

- the 'flu vaccine (every autumn)
- pneumococcal vaccine (check with your doctor if you need a booster)
- the COVID-19 vaccine.

Hygiene

Wash your hands regularly, particularly after being in a public place.

Good air

Exercise outdoors where air quality is good. Avoid outdoor exercise in places where air quality is poor.

Avoid infected people

Avoid contact with people who are unwell and may have an infection.

Symptom diary

Keep a symptom diary that can help you to keep track of your disease and recognise changes that may be signs of an acute exacerbation.

Use a spirometer at home

Use this to check your lung function regularly. It can help you recognise a decline in your lung function. Remember, the results can be affected by the amount of effort you put in, so always make a maximum effort.

Live well

The following will all help you to maintain your overall health:

- exercise
- eat a balanced diet
- get enough sleep
- find ways to manage stress
- keep hydrated by drinking water
- keep in touch with your healthcare team
- join a support group.

Do you suffer from acid reflux?

If you suffer from acid reflux, you should know that there may be a relationship between acid reflux disease and the risk of acute exacerbations. Talk to your doctor about controlling your acid reflux.

Planning for the future

Although not everyone will experience an acute exacerbation of their pulmonary fibrosis, they can happen to anyone at any point during their disease. Living with this uncertainty is difficult, both for pulmonary fibrosis patients and their loved ones.

Talk with your loved ones and your doctor about your wishes should your health suddenly deteriorate.

The Irish Hospice Foundation has excellent resources. They help support people with life-limiting conditions (and their families) to consider plans for the future. See www.irishhospicefoundation.ie for more information.

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Irish Lung Fibrosis Association

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Acute exacerbations of pulmonary fibrosis

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This leaflet gives you information on acute exacerbations (sometimes called 'flare-ups') of pulmonary fibrosis. It tells you:

- what acute exacerbations are
- how they affect your condition
- how to recognise if you might be having an acute exacerbation
- how they are diagnosed and treated.

It also tells you what you can do to reduce the risk of having a flare-up.

What is pulmonary fibrosis?

There are many different types of pulmonary fibrosis diseases. You will sometimes hear pulmonary fibrosis being called 'Interstitial Lung Disease' (shortened to ILD). With ILD, the tissue damage happens in the interstitial space in the lungs, which is the area around the tiny air sacs in the lungs.

Sometimes the underlying cause of the fibrosis is known. For example, your lungs may have been exposed to dusts or certain chemicals. Most of the time the cause is unknown. When the cause for the disease is unknown, it is called idiopathic. In the case of pulmonary fibrosis with an unknown cause, it is called idiopathic pulmonary fibrosis or IPF.

How does pulmonary fibrosis progress?

Pulmonary fibrosis is a progressive disease. This means it gets worse with time. How quickly the disease gets worse varies from person to person. For some people, it progresses slowly, while others get worse much more quickly.

Tests to assess your pulmonary fibrosis

Healthcare staff can monitor how quickly your pulmonary fibrosis is progressing by doing some tests.

Lung function tests (which include tests called spirometry and gas transfer tests): The forced vital capacity or FVC for short is obtained when you perform spirometry. FVC is the maximum amount of air you can forcefully breathe out in one single breath. Another test, named the DLco, is measured by performing gas transfer tests. DLco measures the extent to which oxygen passes from the air sacs of the lungs into the blood. Both your FVC and DLco reduce as your pulmonary fibrosis gets worse.

Oximetry – to measure your oxygen saturation: This measures the amount of oxygen in your blood; it is a sign of how well your lungs are working.

Chest X-rays or CT scans: These show the amount of scarring inside your lungs.

Exercise tests: These show how the disease is affecting your ability to do everyday activities like walking.

For some people with pulmonary fibrosis, their disease gets worse at a constant or steady rate. Other people have a steady rate of decline for months or years, but then experience a large and sudden worsening of their condition. These sudden deteriorations are called acute exacerbations.

What is an acute exacerbation?

An acute exacerbation of pulmonary fibrosis is a sudden worsening or 'flare-up' of your disease. It causes:

- a decline in your lung function (your FVC and DLco)
- a rapid worsening of your breathlessness and other symptoms.

When an acute exacerbation occurs, your doctor will be able to see new changes or scarring on your chest scan.

Doctors aren't sure what causes acute exacerbations. However, some event (like an infection or breathing in something) may trigger a change which speeds up the rate of 'fibrosis'. Fibrosis is the scarring on the inside of your lungs.

Acute exacerbations can occur at any time. About 5 to 20% of people with pulmonary fibrosis may get an exacerbation over a year. Having one acute exacerbation may mean you are more likely to have another in the future.

Acute exacerbations are serious

Acute exacerbations are a serious concern as they often mean people need to be admitted to hospital to receive treatment and can result in death. Even after recovery from the event itself, people don't regain the lung function they had before the acute exacerbation. Acute exacerbations, therefore, speed up the progression of pulmonary fibrosis. This means people get worse more quickly than they would if they hadn't had an acute exacerbation.

How can I recognise an acute exacerbation and what should I do?

You suddenly feel worse: An acute exacerbation normally causes a sudden worsening of your symptoms. Your breathlessness may get much worse over a short time (normally over a few days, but certainly less than 30 days).

Other symptoms also get worse: This includes symptoms like a cough.

You need more oxygen: If you use oxygen, you may feel you suddenly need to use more than usual.

Flu-like symptoms: You may develop flu-like symptoms, including a high temperature, chills, and muscle/joint aches.

Your health results worsen: If you monitor your lung function or oxygen saturation at home, you might notice that your results are lower than normal.

See a healthcare professional

Any rapid change in your symptoms needs to be assessed by a healthcare professional. If you think you might be having an acute exacerbation, call your specialist hospital team straight away.

If they aren't available, call your GP or their out-of-hours service.

The important thing is not to wait – if you are having an exacerbation, you might get worse very quickly. If you are having severe difficulties in breathing, go straight to your hospital emergency department or call 999 or 112 for emergency help.

How are acute exacerbations diagnosed and treated?

If you have already been diagnosed with pulmonary fibrosis, a healthcare professional will ask you about the changes in your symptoms and how quickly they have occurred. They will measure your oxygen saturation and you will have a chest X-ray and often a CT scan. ▶▶