CONDITION

Idiopathic pulmonary fibrosis

Idiopathic pulmonary fibrosis is progressive scarring of the lungs without a known cause.

WHAT IT IS
Progressive lung scarring without a known cause

BODY SYSTEM
Lungs and breathing

MAIN SYMPTOMS
Breathlessness, dry cough, tiredness, reduced exercise ability

WHAT CAUSES IT
Unknown, with age and genetic risk

HOW IT IS DIAGNOSED
High-resolution CT and sometimes lung biopsy

MAIN TREATMENT
Antifibrotic medicines, oxygen, rehabilitation, transplant assessment

CAN IT BE CURED?
No, current treatments cannot remove scarring

IS IT AN EMERGENCY?
Sometimes, with severe or suddenly worsening breathlessness

LONG-TERM OUTLOOK
Progressive, with an unpredictable rate of decline

Overview

Idiopathic pulmonary fibrosis, often shortened to IPF, is a long-term condition that causes scarring in the lungs. "Idiopathic" means that no specific cause has been found. "Fibrosis" means thickening and scarring.

The scarring affects the tiny air sacs where oxygen passes from the lungs into the blood. As the tissue becomes thicker and stiffer, the lungs cannot expand as easily or transfer oxygen as efficiently. This can make physical activity increasingly difficult.

IPF is one type of interstitial lung condition. The interstitium is the supporting tissue around the lung's air sacs. IPF has a characteristic pattern of damage called usual interstitial pneumonia, or UIP. Despite its name, this is a scarring pattern rather than an ordinary infection.

The condition usually progresses over time, but the speed is not predictable. Some people

decline gradually, while others have periods of stability followed by faster changes. Treatment cannot remove existing scar tissue, but it may slow further loss of lung function and help manage symptoms.

Symptoms

Shortness of breath is often the main symptom. It may first appear during exercise, climbing stairs, or walking uphill. As the condition progresses, less demanding activities may cause breathlessness.

A dry cough that does not bring up mucus is also common. It can be persistent and may interfere with talking, sleep, or daily activities.

Other possible symptoms include:

Symptoms usually develop gradually. Because breathlessness and cough can have many causes, these symptoms do not by themselves mean that you have IPF.

A sudden increase in breathlessness is not the usual day-to-day pattern. It may be caused by an infection, a blood clot, a collapsed lung, heart trouble, or an acute exacerbation of IPF. An acute exacerbation is a rapid worsening of lung function without another clear explanation.

- Tiredness and reduced stamina
- Unplanned weight loss or reduced appetite
- Chest discomfort
- Aching muscles or joints
- Rounded, widened fingertips called finger clubbing

Causes and risk factors

The direct cause of IPF is unknown. Researchers think repeated injury to the lining of the air sacs may trigger abnormal healing. Instead of repairing normally, the lung lays down increasing amounts of scar tissue.

IPF is not usually caused by a single exposure or behaviour. Factors linked with a higher chance of developing it include:

Most people with IPF do not have a clear family history. When pulmonary fibrosis occurs in several relatives, it may be called familial pulmonary fibrosis. Inheritance is complex, so a genetic change does not always mean that someone will develop the condition.

Other conditions can also cause lung scarring, including autoimmune conditions, medication reactions, and exposure to mould, birds, asbestos, or industrial dust. These must be considered before scarring is described as idiopathic.

IPF is not contagious. You cannot catch it from or pass it to another person.

- Older age
- A history of smoking
- Long-term exposure to certain wood, metal, stone, or agricultural dusts
- Having a close relative with pulmonary fibrosis
- Certain genetic changes that affect lung repair or ageing

Diagnosis

There is no single blood test that confirms IPF. Diagnosis usually involves a respiratory specialist and may be discussed by a team that includes lung doctors, radiologists, and pathologists.

Your assessment may include a detailed history of your symptoms, work, hobbies, medicines, smoking, home exposures, and family history. Blood tests may look for autoimmune conditions that can cause a similar pattern of lung damage.

A high-resolution CT scan produces detailed images of the lungs. Doctors look for a UIP pattern, which may include scarring around the outer and lower parts of the lungs, widened airways, and small clustered air spaces known as honeycombing. A typical pattern, combined with the right clinical history, may be enough to diagnose IPF without surgery.

Lung function tests measure how much air your lungs hold, how quickly you can breathe out, and how well oxygen crosses into your blood. These tests help measure severity and monitor change, but they do not confirm IPF by themselves.

You may also have an oxygen measurement at rest and while walking. An echocardiogram, which is an ultrasound scan of the heart, may be used if there are concerns about pressure in the lung's blood vessels.

If scans and other findings are unclear, doctors may consider bronchoscopy or a lung biopsy. A bronchoscopy passes a thin flexible tube into the airways. A surgical lung biopsy removes small tissue samples. Biopsy is not needed or suitable for everyone because its risks must be balanced against the likely benefit.

Treatment and management

Treatment aims to slow further scarring, maintain activity, relieve symptoms, and support quality of life. It cannot reverse scar tissue that has already formed.

The antifibrotic medicines nintedanib and pirfenidone can slow the average rate of lung function decline. They do not stop progression in everyone, and they may not make breathlessness or cough feel better.

Nintedanib commonly causes digestive problems, especially diarrhoea and nausea. Pirfenidone can cause nausea, reduced appetite, indigestion, tiredness, and increased sensitivity to sunlight. Both can affect the liver, so people taking them need routine blood tests. Your care team can discuss expected benefits, side effects, interactions, and whether a medicine is suitable for you.

Supplemental oxygen may be offered if the oxygen level in your blood becomes too low at rest, during activity, or while sleeping. Oxygen does not treat the scarring itself, but it may improve low oxygen levels and make activity safer or more comfortable.

Pulmonary rehabilitation combines supervised exercise, breathing strategies, education, and practical support. It can improve fitness and help you manage breathlessness, although it does not change the lung scarring.

Management may also include:

Palliative care is not limited to the final stage of illness. It can be provided alongside antifibrotic treatment and rehabilitation.

Lung transplantation may be considered for some people whose condition is progressing. It is the only treatment that replaces the scarred lungs, but it involves major surgery and lifelong anti-rejection treatment. Age, general health, other medical conditions, and transplant availability affect whether it is an option. Early assessment may be useful because the evaluation process takes time.

- Support to stop smoking
- Vaccination against respiratory infections
- Nutrition advice if appetite or weight is affected
- Treatment for anxiety, low mood, troublesome cough, or severe breathlessness
- Management of other lung, sleep, or heart conditions
- Palliative care for symptom relief and planning at any stage

Complications

As IPF progresses, the lungs may become less able to supply the body with oxygen. This can lead to chronic respiratory failure, meaning oxygen remains too low or breathing becomes inadequate.

Possible complications include:

People with IPF also have a higher risk of lung cancer than the general population, although most do not develop it.

An acute exacerbation can occur without warning and may be serious. It causes new or sharply worsening breathlessness over days or weeks, along with fresh changes on lung scans. Similar symptoms can result from treatable problems, so urgent assessment is important.

- Pulmonary hypertension, or high pressure in the lung's blood vessels
- Strain on the right side of the heart
- Acute exacerbations with rapid worsening
- Respiratory infections
- A collapsed lung
- Reduced mobility and muscle strength
- Anxiety, depression, or social isolation

Prevention

There is no proven way to prevent IPF because its cause is unknown. Having a risk factor does not mean that the condition could have been avoided.

You can still protect your remaining lung health by not smoking and by reducing exposure to harmful dusts, fumes, and vapours. Use appropriate workplace respiratory protection where required.

Vaccination and sensible infection precautions may reduce the chance of illnesses that place extra strain on the lungs. These steps do not prevent or cure IPF itself.

If several close relatives have pulmonary fibrosis, a specialist may discuss genetic counselling. Genetic testing is not automatically useful for everyone and cannot predict the future with certainty.

Living with it and what to expect

IPF is usually progressive, meaning lung function tends to decline over months and years. The rate differs greatly between people and can also change over time. A stable period does not guarantee that the condition has stopped, while one difficult week does not always mean permanent decline.

Regular follow-up usually includes discussion of symptoms, lung function testing, oxygen measurements, medicine side effects, and changes in daily activity. Tell your care team if you are walking less, avoiding stairs, losing weight, or taking longer to recover after activity. These changes can be useful even when test results appear similar.

Staying active within your limits can help preserve strength. Pulmonary rehabilitation can teach you how to pace tasks, plan rest breaks, and recover from breathlessness. If you use oxygen, equipment should be chosen around your needs at home and while moving about.

Travel may remain possible, but air travel and high altitude can lower oxygen levels further. An oxygen assessment may be needed before flying, even if you do not normally use oxygen.

IPF can shorten life, but population averages cannot predict one person's course. Age, lung function, oxygen needs, other health conditions, acute exacerbations, and response to treatment all affect the outlook.

Planning ahead can give you more control. This may include discussing future oxygen needs, transplant assessment, help at home, symptom relief, and the care you would want if your breathing became much worse.

When to get medical help

Arrange routine medical advice if you develop an unexplained cough or breathlessness that persists, especially if it limits activity. These symptoms have many possible causes and need an appropriate assessment.

If you have diagnosed IPF, contact your care team promptly about:

Seek urgent medical help for a marked or rapid change in breathing over hours or days.

Call emergency services for severe breathlessness, blue or grey lips, new confusion, fainting, severe chest pain, or an inability to speak in full sentences. Do not drive yourself if you are severely unwell.

- Gradually worsening breathlessness or cough
- Lower oxygen readings than your agreed usual range

- New fever, mucus, or other signs of infection
- Unplanned weight loss or difficulty eating
- Medicine side effects
- Increasing difficulty with normal daily activities

Common questions

Is idiopathic pulmonary fibrosis cancer?

No. IPF is lung scarring, not cancer. However, people with IPF have a higher risk of developing lung cancer than people without it.

Is IPF hereditary?

Most cases do not follow a simple inherited pattern. Some people have genetic changes or several affected family members. If pulmonary fibrosis runs in your family, genetic counselling may help explain what testing can and cannot show.

Can lung scarring from IPF heal?

Current treatments cannot remove established scar tissue. Antifibrotic medicines may slow the development of further scarring, while oxygen, rehabilitation, and symptom treatment can help you function more comfortably.

How quickly does IPF progress?

There is no reliable timetable. Some people change slowly over several years, while others decline more quickly or experience sudden exacerbations. Regular monitoring helps identify changes and adjust support.

How long can I live with IPF?

IPF is a serious, life-shortening condition, but no single estimate applies to everyone. Your outlook is influenced by lung function, oxygen needs, other health conditions, acute exacerbations, and whether treatments or transplantation are suitable.

Can I exercise or fly with IPF?

Many people can remain active and travel with planning. Pulmonary rehabilitation can help you exercise safely. Before flying, ask about an oxygen assessment because aircraft cabins contain less oxygen than ground-level air.

Sources

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