CONDITION

Fibrous interstitial pneumonia

Fibrous interstitial pneumonia is inflammation and scarring in the supporting tissue of the lungs.

WHAT IT IS
Scarring and inflammation in tissue around the lung air sacs

BODY SYSTEM
Lungs and breathing

MAIN SYMPTOMS
Breathlessness, dry cough, fatigue, reduced exercise tolerance

WHAT CAUSES IT
Often unknown; sometimes autoimmune disease, exposures, or medicines

HOW IT IS DIAGNOSED
High-resolution chest CT, sometimes bronchoscopy or lung biopsy

MAIN TREATMENT
Cause-specific care, antifibrotics, oxygen, and pulmonary rehabilitation

CAN IT BE CURED?
No - established lung scarring usually remains

IS IT AN EMERGENCY?
Sometimes - sudden severe breathlessness needs emergency care

LONG-TERM OUTLOOK
May stay stable or progress despite treatment

Overview

Fibrous interstitial pneumonia describes inflammation and scarring in the tissue that supports the lungs. This tissue, called the interstitium, surrounds the tiny air sacs where oxygen enters the blood.

When the interstitium becomes thick and stiff, the lungs cannot expand as easily, and oxygen may have difficulty passing into the bloodstream. This can cause breathlessness, a dry cough, and reduced stamina.

The term describes a lung condition rather than one specific disease. Several forms of interstitial lung disease can produce this pattern, including idiopathic pulmonary fibrosis, fibrotic nonspecific interstitial pneumonia, autoimmune-related lung disease, and damage linked to certain exposures or medicines. "Idiopathic" means no cause has been found.

Identifying the type matters because treatment varies. Inflammatory forms may respond to medicines affecting the immune system. Conditions involving mainly established fibrosis may be treated with medicines intended to slow further scarring.

Symptoms

Symptoms often develop gradually. You may first notice difficulty with activities that previously felt manageable, such as climbing stairs or walking uphill.

Common symptoms include:

Some people lose weight or develop clubbing, in which the fingertips become broader and the nails curve more than usual.

An underlying autoimmune condition may cause painful or swollen joints, muscle weakness, dry eyes or mouth, skin changes, or fingers that change colour in the cold.

Breathlessness does not always match the amount of scarring on a scan. Heart conditions, anaemia, infection, anxiety, muscle weakness, and loss of fitness can also affect breathing.

- Shortness of breath, especially during activity
- A persistent dry cough
- Tiredness or reduced energy
- Lower exercise tolerance

Causes and risk factors

In many cases, the cause is unknown. Doctors may use the word idiopathic when assessment has not found an exposure, medicine, or other condition that explains the scarring.

Possible causes and contributing factors include:

Hypersensitivity pneumonitis can develop when the immune system repeatedly reacts to something breathed into the lungs. Continued exposure may lead to fibrosis, although the cause is not always obvious.

When more than one person in a family has pulmonary fibrosis, it is called familial pulmonary fibrosis. Having an affected relative does not mean you will develop it, but genetic counselling or testing may be considered.

Several fibrosing lung conditions are more common later in life, although younger adults can also be affected.

- Autoimmune conditions, such as rheumatoid arthritis, systemic sclerosis, or inflammatory muscle diseases
- Repeated exposure to mould, birds, farming dust, metal dust, wood dust, or other particles
- Certain medicines, including some cancer treatments and heart-rhythm medicines
- Radiation treatment involving the chest
- Previous or continuing cigarette smoking
- Inherited susceptibility to pulmonary fibrosis

Diagnosis

There is no single test that identifies every type of fibrous interstitial pneumonia. Diagnosis usually combines symptoms, medical history, examination, scans, and lung function tests.

A clinician will ask about your work, hobbies, home, medicines, smoking history, and contact with birds, mould, dust, or chemicals. They may also ask about autoimmune symptoms and relatives with lung scarring.

A high-resolution CT scan provides detailed images of the lungs. The location and shape of scarring may suggest a particular pattern. One important pattern is usual interstitial pneumonia, or UIP. It can occur in idiopathic pulmonary fibrosis but may have other causes.

Pulmonary function tests measure how much air the lungs can hold and how effectively gases enter the blood. A walking test may show whether oxygen levels fall during activity. Blood tests can look for autoimmune conditions and other causes of breathlessness.

If the diagnosis remains uncertain, bronchoscopy may be used to collect fluid or small tissue samples through a flexible tube passed into the lungs. A larger lung biopsy is needed less often because it has meaningful risks and may not change treatment.

Specialists in lung medicine, radiology, pathology, and rheumatology may review the findings together to determine the likely type and whether fibrosis is progressing.

Treatment and management

Treatment depends on the lung pattern, its cause, your symptoms, and evidence of progression. Established scar tissue usually cannot be removed, but treatment may reduce inflammation, slow further fibrosis, improve activity, or manage low oxygen.

If an exposure is contributing, avoiding it may be important and could require changes at home

or work. Do not stop a prescribed medicine on your own if it is suspected as a cause; the prescriber can assess alternatives.

Antifibrotic medicines, including nintedanib and pirfenidone, can slow lung function loss in certain fibrosing conditions. They do not remove existing scars. Eligibility depends on the diagnosis and local approval criteria.

Corticosteroids or other medicines that reduce immune activity may be used when inflammation or an autoimmune condition is driving damage. They are not suitable for every form. In particular, routine immune suppression is not used for idiopathic pulmonary fibrosis in the same way as for inflammatory interstitial lung disease.

Pulmonary rehabilitation combines supervised exercise, breathing strategies, and education. It can improve stamina and help you use your available lung function effectively. Supplemental oxygen may be offered when blood oxygen is low at rest, during activity, or while sleeping.

Care may also include cough treatment, smoking cessation support, vaccination against respiratory infections, and help with nutrition or emotional wellbeing.

Lung transplantation may be considered for some people with advanced or worsening fibrosis. Eligibility depends on age, overall health, other conditions, and transplant-centre assessment.

Complications

Fibrosis can reduce the oxygen reaching the blood. Some people eventually develop chronic respiratory failure and need oxygen support.

Pulmonary hypertension, or high pressure in the lung blood vessels, can strain the right side of the heart. It may worsen breathlessness, fatigue, swelling, or faintness.

An acute exacerbation is a sudden, unexplained worsening of lung injury. It can cause rapidly increasing breathlessness over days or weeks and may require hospital care. Infection, a lung blood clot, and heart problems can cause similar symptoms, so prompt assessment is important.

Reduced activity can lead to muscle weakness and loss of fitness. Persistent cough and uncertainty may also affect sleep, mood, relationships, and daily life.

Prevention

Many cases cannot be prevented, particularly when the cause is unknown or inherited susceptibility is involved.

Avoiding harmful dust, fumes, mould, or bird proteins may lower risk when these contribute. Workplace controls and correctly fitted respiratory protection may be needed. Removing an exposure after fibrosis develops may help prevent further injury, although existing scars usually remain.

Not smoking reduces additional lung damage. If you take a medicine known to affect the lungs, attend recommended monitoring. Do not stop prescribed treatment without medical advice.

Vaccination and sensible infection precautions do not prevent fibrosis, but they can reduce infections that place extra strain on scarred lungs.

Living with it and what to expect

The course differs between people. Some conditions remain stable for long periods, particularly when a cause is removed or inflammation responds to treatment. Others progress over months or years. A period of stability does not guarantee the condition will remain unchanged.

Follow-up commonly includes symptom reviews and repeat pulmonary function tests. Walking tests may check exercise ability and oxygen levels. CT scans are generally repeated when they are likely to clarify a change rather than at every appointment.

Notice changes in what you can do, such as needing more pauses on a familiar walk. A steady increase in cough or breathlessness should be discussed at routine or earlier follow-up rather than waiting until symptoms become severe.

Regular activity is usually encouraged within safe limits. Pulmonary rehabilitation can help you find a manageable level and pace demanding tasks. If oxygen is prescribed, your care team can explain how to use it during activity, sleep, or travel.

Many people continue working, travelling, exercising, and participating in family life, although adaptations may be needed. If the condition progresses, discussions about future treatment preferences and supportive care can help you remain involved in decisions.

When to get medical help

Arrange a routine or earlier medical review for gradually worsening breathlessness, a persistent increase in cough, lower exercise tolerance, unexplained weight loss, or new medicine side effects. Mention new joint, muscle, skin, or circulation symptoms because they may suggest an autoimmune cause.

Seek urgent same-day assessment for a clear and rapid change in breathing, fever with worsening respiratory symptoms, new chest discomfort, coughing up blood, or oxygen readings that remain below your advised target.

Call emergency services for severe breathlessness at rest, blue or grey lips, confusion, fainting, crushing chest pain, or an inability to speak normally because of breathing difficulty.

Common questions

Is fibrous interstitial pneumonia the same as idiopathic pulmonary fibrosis?

No. Fibrous interstitial pneumonia is a broad description of inflammation and scarring. Idiopathic pulmonary fibrosis is a specific diagnosis with a recognised scan or tissue pattern and no identified cause.

Can lung scarring heal?

Established fibrosis usually does not return to normal lung tissue. Inflammation around the scarring may improve, and treatment may slow or prevent further damage.

Is it lung cancer or can it turn into cancer?

It is not lung cancer or a precancerous condition. Some fibrosing lung diseases are associated with a higher risk of lung cancer, particularly when smoking or older age adds to the risk.

Is fibrous interstitial pneumonia contagious?

No. Lung fibrosis cannot pass between people. Respiratory infections can still occur and may have a greater effect on someone with lung damage.

How quickly does it progress?

There is no fixed timetable. The course depends on the underlying type, amount of fibrosis, response to treatment, other health conditions, and whether lung function continues to decline.

Will I need oxygen?

Not everyone does. Oxygen is used when testing shows that blood oxygen is too low, which may initially happen only during activity or sleep. Needing oxygen is treatment for low oxygen levels, not a sign that you have done something wrong.

Sources

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