CONDITION

# Interstitial lung disease

Interstitial lung disease is a group of conditions that inflame or scar the tissue around the lungs' air sacs.

**WHAT IT IS**
Inflammation or scarring of tissue within the lungs

**BODY SYSTEM**
Lungs and breathing

**MAIN SYMPTOMS**
Breathlessness, dry cough, tiredness, reduced exercise tolerance

**WHAT CAUSES IT**
Autoimmune disease, inhaled exposures, medicines, or unknown

**HOW IT IS DIAGNOSED**
Chest CT, breathing tests, and sometimes lung biopsy

**MAIN TREATMENT**
Cause-directed medicines, rehabilitation, oxygen, sometimes transplant

**CAN IT BE CURED?**
Sometimes - inflammation may resolve; established scarring usually remains

**IS IT AN EMERGENCY?**
Sometimes - sudden severe breathlessness or blue lips

**LONG-TERM OUTLOOK**
Stable for some; progressive scarring can shorten life

## Overview

Interstitial lung disease, usually shortened to ILD, is the name for a large group of conditions affecting the tissue in and around the lungs' tiny air sacs. This supporting tissue is called the interstitium.

ILD can cause inflammation, scarring, or both. Lung scarring is also called fibrosis. Scarred lungs become stiffer, making it harder to take a deep breath and move oxygen into the blood.

Some forms of ILD are linked to an immune condition, workplace exposure, medicine, or infection. In other cases, no cause is found. Idiopathic pulmonary fibrosis is one specific type in which progressive lung scarring has no known cause.

The different types do not all behave in the same way. Some improve when their cause is

removed or inflammation is treated. Some remain stable for years. Others gradually worsen. Finding the specific type matters because it guides treatment and helps explain what may happen over time.

ILD is not itself a type of cancer, and most forms are not contagious.

## Symptoms

Symptoms may appear slowly over months or years. Less often, they develop over days or weeks.

Common symptoms include:

You may begin avoiding hills or taking more breaks without realizing that your breathing has changed.

Some people develop clubbing. This means the ends of the fingers become wider and the nails curve more than usual. It does not happen in everyone and can have causes other than ILD.

Symptoms outside the lungs can provide clues to the cause. Joint pain, muscle weakness, skin changes, dry eyes, or cold-sensitive fingers may suggest an autoimmune condition. Autoimmune conditions happen when the immune system mistakenly attacks the body's own tissues.

Symptoms alone cannot confirm ILD. Asthma, heart conditions, infections, anemia, and other lung conditions can cause similar problems.

- Shortness of breath, first noticed during activity
- A dry cough that does not settle
- Tiredness
- Reduced ability to walk, climb stairs, or exercise
- Chest discomfort
- Unplanned weight loss in some forms of ILD

## Causes and risk factors

ILD has many possible causes. Sometimes more than one factor may be involved.

Autoimmune conditions associated with ILD include rheumatoid arthritis, systemic sclerosis, inflammatory muscle diseases, and Sjögren disease. Lung symptoms can occasionally appear before the other features of these conditions.

Repeated exposure to inhaled material can cause hypersensitivity pneumonitis, an immune reaction within the lungs. Possible triggers include mould, birds, feathers, farming materials, and contaminated water systems. The trigger is not always obvious.

Long-term workplace exposure to substances such as asbestos, silica, coal dust, or certain metal and wood dusts can also scar the lungs. The risk depends on the substance, amount, duration of exposure, and use of effective protection.

Some medicines can injure the lungs. These include certain cancer treatments, heart medicines, antibiotics like amoxicillin or doxycycline , and immune treatments. Radiation therapy involving the chest can also cause lung inflammation or scarring. Do not stop a prescribed medicine without medical advice, because stopping it suddenly may be harmful.

A small number of ILDs run in families. Inherited changes affecting protective chromosome structures called telomeres, or proteins in the air sacs, can increase risk. Having an affected relative does not mean that you will develop ILD.

Smoking can contribute to certain types and may worsen overall lung health. Increasing age is associated with some fibrotic forms, including idiopathic pulmonary fibrosis.

In many people, no single cause is found even after a detailed assessment.

## Diagnosis

Diagnosis usually starts with questions about your symptoms, work, hobbies, home environment, medicines, smoking history, and family history. A physical examination may detect fine crackling sounds in the lungs or finger clubbing.

A high-resolution CT scan is central to diagnosis. This detailed chest scan can show the pattern and location of inflammation or scarring. Some CT patterns strongly suggest a particular type of ILD, while others need further investigation.

Breathing tests measure how much air your lungs hold, how quickly you can breathe out, and how well gases pass from the lungs into the blood. A test called DLCO measures gas transfer. These tests help show severity and provide a baseline for future comparisons.

You may also have:

During bronchoscopy, fluid may be used to wash a small area of the lung and collect cells. This is called bronchoalveolar lavage. It can help identify inflammation, infection, or exposure-related disease.

A biopsy is not needed for everyone. When it is considered, the team weighs whether the result is likely to change care against risks such as bleeding, infection, or a collapsed lung.

Because ILD can be complex, specialists often review the history, scans, laboratory results, and any tissue samples together. This multidisciplinary discussion may involve lung doctors, radiologists, pathologists, rheumatologists, and occupational health specialists.

- Blood tests for autoimmune conditions or other causes
- Oxygen measurements at rest and while walking
- An echocardiogram, which uses ultrasound to assess the heart
- A bronchoscopy, in which a thin camera passes into the airways
- A lung tissue sample when other findings do not provide an answer

## Treatment and management

Treatment is based on the specific ILD, its cause, whether inflammation or fibrosis is dominant, and whether it is changing.

If an inhaled trigger is identified, avoiding it may reduce further injury. This can require changes at home or work. Occupational health or environmental specialists may help locate and control the exposure.

Corticosteroids and other medicines that reduce immune activity are used for some inflammatory or autoimmune forms. They are not suitable for every ILD and may cause significant side effects, including infection risk.

Antifibrotic medicines can slow loss of lung function in idiopathic pulmonary fibrosis and certain other progressive fibrosing ILDs. Nintedanib and pirfenidone are examples. They do not remove existing scars, and they require monitoring for side effects.

Supplemental oxygen may be advised if oxygen levels are low at rest, during activity, or while sleeping. Oxygen can support activity and protect organs from low oxygen, but it does not directly treat scarring.

Pulmonary rehabilitation combines supervised exercise, breathing education, and support for managing breathlessness. It can improve fitness, confidence, and daily function even though it does not reverse ILD.

Other parts of care may include stopping smoking, staying current with recommended vaccinations, treating related conditions, receiving nutrition support, and managing distress or poor sleep. Palliative care can help control breathlessness and support quality of life at any stage; it is not limited to end-of-life care.

Lung transplantation may be considered for some people with severe or progressive disease.

Eligibility depends on overall health, the type and stage of ILD, and transplant-centre assessment.

## Complications

As fibrosis increases, the lungs may become less able to move oxygen into the blood. This can lead to long-term low oxygen levels and, in advanced disease, respiratory failure.

High pressure can develop in the blood vessels between the heart and lungs. This is called pulmonary hypertension. It places extra strain on the right side of the heart and may cause worsening breathlessness, faintness, chest discomfort, or swelling in the legs.

Some people have an acute exacerbation, meaning a sudden worsening of lung injury over days or weeks without a simple explanation. Infection, a blood clot, heart problems, and other treatable causes can produce similar symptoms and must be checked promptly.

Reduced activity can lead to loss of muscle strength and independence. Anxiety, low mood, poor sleep, and social isolation can also occur, especially when breathlessness limits everyday plans.

Treatment can create complications as well. Medicines that suppress the immune system may increase infection risk, while antifibrotic medicines can cause digestive or liver-related side effects. Monitoring is intended to identify these problems early.

## Prevention

Not every form of ILD can be prevented. There is no proven way to prevent ILD caused by an inherited change or an unknown process.

Avoiding tobacco smoke and harmful dusts, fumes, moulds, or animal proteins can reduce some preventable lung injury. Workplace controls may include ventilation, approved protective equipment, exposure monitoring, and following safety procedures.

If you take a medicine known to affect the lungs, attend any recommended monitoring and report new breathing symptoms. Do not stop treatment on your own.

When a specific exposure has already caused ILD, preventing further contact may help limit additional injury. However, removing the trigger does not always reverse established fibrosis.

## Living with it and what to expect

The course of ILD differs widely. Some inflammatory forms improve with treatment or removal of a trigger. Some remain stable with regular monitoring. Fibrotic forms may progress slowly, while a smaller number worsen more quickly or have sudden setbacks.

Follow-up often includes symptom reviews, breathing tests, walking oxygen measurements, and occasional CT scans. Changes over several visits are usually more informative than one result. Report a clear decline rather than waiting for the next routine appointment.

Regular activity within your limits can help preserve strength. Pulmonary rehabilitation can teach you how to pace tasks, recover after exertion, and exercise safely. Using prescribed oxygen during activity may make movement easier.

Planning can reduce the impact on daily life. This may include allowing extra travel time, placing commonly used items within easy reach, sitting for demanding tasks, or discussing adjustments at work. Support groups and counselling can help with uncertainty and changes in independence.

Some people live active lives with mild, stable ILD for many years. Others need increasing oxygen, practical support, or assessment for transplantation. Your recent symptom pattern, lung function trend, CT findings, oxygen needs, and specific diagnosis provide more useful information than the general label "ILD."

## When to get medical help

Arrange a routine appointment if you develop a persistent dry cough, gradually worsening breathlessness, or a steady drop in how far you can walk. These symptoms have many possible causes, but they deserve assessment.

If you already have ILD, contact your healthcare team promptly if your symptoms are worsening faster than usual, your oxygen readings are repeatedly below the range in your care plan, or treatment side effects are difficult to manage.

Seek urgent same-day medical care for new or rapidly increasing breathlessness, fever with worsening respiratory symptoms, coughing up blood, new leg swelling, or chest discomfort.

Call emergency services for severe difficulty breathing, blue or grey lips, confusion, fainting, crushing chest pain, or an inability to speak in full sentences. Sudden major worsening should not wait for a routine ILD appointment.

## Common questions

Is interstitial lung disease the same as pulmonary fibrosis?

Not exactly. ILD is the broad group of conditions. Pulmonary fibrosis means lung scarring and is a feature of many, but not all, ILDs. Some forms mainly cause inflammation, particularly early in their course.

Can interstitial lung disease be cured?

Some exposure-related or inflammatory forms can improve greatly when the cause is removed or treated. Established fibrosis usually cannot be removed. Treatment may still slow further scarring, ease symptoms, and help you remain active.

Is interstitial lung disease cancer?

No. ILD is not cancer. A scan may sometimes show findings that need more investigation to distinguish scarring from infection, cancer, or another lung condition.

How quickly does ILD progress?

There is no single timeline. Some forms remain stable for years, while others worsen over months or have periods of sudden decline. Repeated breathing tests, oxygen measurements, symptoms, and scans help show your individual pattern.

Can I exercise with ILD?

Many people can and should remain active within safe limits. Exercise does not reverse scarring, but it can protect muscle strength and daily function. Pulmonary rehabilitation provides structured guidance, especially if activity causes breathlessness or low oxygen.

Can I fly if I have ILD?

Cabin oxygen levels are lower than at ground level, so some people need in-flight oxygen even if they do not use it every day. An assessment before travel may be needed. Airlines have different rules and require advance arrangements for oxygen equipment.

## Sources

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