



*"When you can't breathe...
nothing else matters"*

Interstitial Lung Diseases – Adult

Interstitial Lung Diseases – Adult

There is no pretending that interstitial lung diseases (ILD) are easy to understand, even for senior doctors. To understand these conditions more easily, it helps to know about the "interstitium"; the part of the lung which is affected. The interstitium is the tissue through which oxygen from the lung's tiny air sacs (the alveoli) passes to enter the lung's small blood vessels (capillaries). It also provides a framework to maintain the lung's shape. A large group of different diseases can damage the interstitium and together these diseases are termed "interstitial lung disease".

When the interstitium is damaged, the major effect is to reduce the lung's normal ability to deliver oxygen to the bloodstream. ILD can distort the lungs' shape, generally making them smaller and stiffer, and change the shape of the airways that pass through damaged areas of lung. They can occur at any age, in both men and women. They are not contagious and are only rarely handed down from one generation to the next. There are several well-recognised types that have known triggers, but other types have no known cause, and are the subject of intense research to understand more about them. Many ILDs are rare, which has hampered research efforts.

What are the symptoms?

Although all ILDs can affect the interstitium, individual types have different patterns of where they cause most damage in the lung, and this can alter the symptoms that people notice. Most commonly, people with ILD notice breathlessness. Initially, breathlessness often occurs only during exertion, but with more advanced disease, it occurs even during rest. People with ILD often also suffer cough, particularly when the ILD pattern is based around the lungs' airways. Although these symptoms vary amongst individuals, they can be severe and disabling. This can be particularly so when people also suffer from anxiety or depression, each of which are common in ILD and should not be ignored if present. Sometimes, ILD is part of a condition that affects more than just the lung, and people experience additional symptoms related to other parts of their body, such as joint problems or rash. People with ILD also often have other diseases that can be caused by the ILD or make the symptoms of the ILD worse. These diseases, called co-morbidities, may also require treatment to improve symptoms.

What causes these diseases?

ILD is a group of diseases with many causes. It is very important to check whether it has been caused by something that could be avoided; perhaps at work, in the home, via hobbies, or by a medicine.

Examples of such externally triggered diseases include asbestosis and bird fancier's lung (hypersensitivity pneumonitis). Others occur as part of a condition, which can affect multiple body parts, such as rheumatoid arthritis, scleroderma or sarcoidosis. Usually, only the lung is affected, with the most common type of ILD seen in adults being idiopathic pulmonary fibrosis (IPF).

There are many other rare types, and recognising these can require considerable expertise.

Common types of interstitial lung disease

- **ILD limited to the lung**
 - Idiopathic pulmonary fibrosis
 - Other idiopathic interstitial pneumonias
- **ILD caused by diseases that can affect other parts of the body**
 - ILD associated with systemic diseases /connective tissue diseases – e.g., rheumatoid arthritis, scleroderma, sarcoidosis
- **ILD associated with known triggers**
 - Hypersensitivity pneumonitis – e.g. bird fancier's lung, farmer's lung
 - Asbestosis
 - Silicosis
 - Medication-induced lung disease

How is interstitial lung disease diagnosed?

When someone develops ILD, it is very important that they are referred to a specialist physician, who will often need to spend time taking a careful history and performing a thorough examination. This is because the types and causes of ILD can be difficult to diagnose, and it is vital that any environmental triggers are recognised so that they can be avoided. Guidelines recommend that the diagnosis of ILD is made by a team of specialists and increasingly, such teams are used in Australia to establish the most likely diagnosis. Multiple tests are often required, including:

- **Chest x-ray.** This is commonly a good first step as a screening investigation for ILD, although it can be normal in mild ILD. It is useful if previous chest x-rays are available for comparison to see how quickly ILD has developed, as this can guide diagnosis and allow some prediction of future outlook.
- **Lung function testing.** Also known as a breathing test, this test helps to confirm the presence of ILD and to allow its progress to be monitored. Although they can be quite tiring – requiring repeated breaths in and out at maximum force and depth - they are the most important test in many cases for monitoring the progression of ILD and response to treatment.
- **Blood tests.** These tests are used to look for systemic diseases (conditions involving several parts of the body) such as rheumatoid arthritis and for evidence of exposure to environmental triggers, such as birds. It is probable that in future, blood tests will also be used to monitor the progress of ILD by measuring components of the blood called biomarkers, although no such markers are available yet.
- **Computed Tomography (CT scan).** This is a radiology test that takes a very detailed picture of the lungs. The type of CT done for ILD is called a high resolution CT (HRCT), often the person must have pictures taken whilst lying on their front and back, and with their lungs full (breathing in)

and empty (breathing out). Many types of ILD have a unique pattern on HRCT that allows doctors to make a diagnosis without the need for more invasive tests, such as a surgical biopsy (which entails more risk). If a biopsy is needed, the CT scan can help identify the best target site for the biopsy. A repeat CT is sometimes needed in monitoring the progression of ILD, its response to therapy, or to assess for the development of new features.

- **Bronchoscopy.** This test is performed in some cases of ILD, using a narrow flexible fibre-optic tube to collect fluid and tissue from the lung. It is done under sedation and is usually a safe and well-tolerated procedure. A new technique done via bronchoscopy called cryo-biopsy is being used in some centres. It is hoped that this technique might provide specimens large enough to avoid the need for surgical lung biopsy, and with fewer complications.
- **Lung biopsy.** This procedure is done in select cases of ILD, where other investigational steps have not revealed a diagnosis. It requires a general anaesthetic and admission to hospital. It is done using a keyhole technique with small cuts on the side of the chest to insert a telescope and specialised instruments to take several pieces of lung, each about the size of a cherry. It is generally only done when the test result will make a difference in treatment choices or will provide important information about diagnosis and outlook.

Disease treatment

The treatment of ILD is complex and usually involves a variety of approaches. Lifestyle changes such as smoking cessation, optimising body weight and undertaking regular exercise are helpful. A dedicated program of exercise and education called pulmonary rehabilitation can maximise the benefits of these changes. Treating other medical conditions that can contribute to symptoms such as sleep apnoea, gastro-oesophageal reflux (heart burn) or heart conditions can reduce the severity of symptoms, particularly exertional breathlessness.

In consultation with a specialist, environmental, work-related or other triggers may be identified that can be avoided. Sometimes this can involve major changes to work or home life and therefore should not be undertaken without specialist advice. People with low oxygen levels may also benefit from oxygen therapy.

Specific medical approaches for ILD are available, depending on the type of ILD and its severity, and are best decided by a physician specialising in ILD. This is most likely to be a respiratory physician but may sometimes be an immunologist, rheumatologist or general physician, depending on local circumstances and availability. For some conditions, immunosuppressant drugs that reduce the immune system's activity are used. For example, in sarcoidosis, prednisolone is commonly prescribed, while combinations of immunosuppressants may be prescribed for ILD associated with connective tissue disease. International guidelines recommend anti-fibrotic agents for IPF and it is hoped that they will soon be available in Australia. In some cases, where ILD is severely disabling and the outlook guarded, lung transplantation may be appropriate.

Research initiatives

Clinical trials are frequently recommended to people with ILD, particularly those with IPF. This is because, while advances have been made in the last few years with the use of anti-fibrotics for IPF, the disease

remains progressive and disabling. Randomised, placebo-controlled, blinded trials are the best way of establishing the effectiveness of new treatments but have not been performed in other ILDs. Hence, the evidence for current clinical practice needs to be strengthened and treatments improved. When a clinical trial is recommended in Australia, strict guidelines are set by federal, state and local hospital authorities to ensure that the trial is conducted in an ethical manner. In all cases, researchers are obliged to ensure that participants in clinical trials have a clear understanding of what the trial involves, and those who receive the placebo (non-experimental therapy) must receive the standard of care currently available.

There are many ways that people with ILD can become involved in research, apart from receiving trial therapies. ILDs are rare and can only be properly researched when there is a big enough group to detect unique findings. A common way of collecting information about groups with rare diseases is via a registry. Lung Foundation Australia runs a nationwide registry for people with IPF and some large hospitals collect information about other ILDs. Inclusion within these registries can be arranged through local respiratory physicians. Medical research is expensive and government grant funding is increasingly difficult to obtain. Sadly, it is well recognised that rare diseases can be overlooked by funding bodies, such that the term “orphan disease” is sometimes used to denote the paucity of research and/or treatment options. By donating money or leaving a bequest, people with ILD can help ensure that their disease is studied so that one day better treatments will become available. Lung Foundation Australia is able to help administer such donations and bequests.

Patient support

Lung Foundation Australia provides information and support to patients with all forms of lung disease including ILD and advocates on their behalf. We encourage you to call our Information and Support Centre free call on 1800 654 301.

A number of internet sites exist that provide useful information relating to ILD:

- www.lungfoundation.com.au
- www.lungusa.org
- www.nhlbi.nih.gov
- www.mayoclinic.com/health/interstitial-lung-disease
- www.bpold.co.uk
- www.pulmonaryfibrosis.org

Content updated November 2015

This brochure is one in a series produced by Lung Foundation Australia to provide information on lung disease, its treatment and related issues. The information published by Lung Foundation Australia is designed to be used as a guide only, is not intended or implied to be a substitute for professional medical treatment and is presented for the sole purpose of disseminating information to reduce lung disease. Any information relating to medication brand names is correct at the time of printing. Lung Foundation Australia has no control or responsibility for the availability of medications, which may occasionally be discontinued or withdrawn. Please consult your family doctor or specialist respiratory physician if you have further questions relating to the information contained in this leaflet. For details of patient support groups in Australia please call 1800 654 301.