Scleroderma with Interstitial Lung Disease

Boehringer Ingelheim

Centre of Research Excellence in Pulmonary Fibrosis

Lung Foundation Australia

What is Scleroderma associated Interstitial Lung Disease?

Scleroderma (also known as systemic sclerosis) is an autoimmune disease that involves the thickening and tightening of skin as well as the narrowing of blood vessels. If you have scleroderma with Interstitial Lung Disease (ILD), the condition is known as Systemic Sclerosis-related ILD or SSc-ILD for short.

Scleroderma can damage internal organs including the lungs, oesophagus, kidneys, heart and blood vessels. It is not known exactly what causes scleroderma, but it is thought to be a combination of many factors including a genetic predisposition, as well as some environmental triggers. While there is no cause found in most patients with scleroderma, the best known environmental trigger is crystalline silica, encountered in industries such as stone masonry, mining and construction.

ILD is frequently diagnosed in people living with scleroderma. In 10-20% of newly diagnosed scleroderma cases, the lungs are identified as the first site of involvement.

50–65% of people with scleroderma show some evidence of SSc-ILD on a lung scan, however, not everyone who has SSc-ILD develops symptoms or needs treatment.

What are the signs and symptoms of SSc-ILD?

The signs and symptoms of scleroderma can differ depending on which organs are involved. Some of the common signs and symptoms of scleroderma are listed below, but you may experience additional or different signs and symptoms to these:

Scleroderma signs and symptoms

Skin changes including thickening, tightening or hardening of the skin, colour changes or red spots on the skin (telangiectasias), hair loss and reduced sweating

Raynaud's phenomenon or chill blains, in which fingers or toes turn white and blue and then appear red as blood flow returns to normal – this occurs in response to extreme cold or emotional stress. Puffy fingers, sores and pitted scars on the fingers

You may experience breathlessness, cough and/or fatigue if your lungs are involved

Reflux, heartburn or problems swallowing

Heart problems such as high blood pressure or pulmonary hypertension

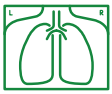
Stiff and painful joints

How is SSc-ILD diagnosed?

Scleroderma may affect many different parts of the body, which can make diagnosis difficult. Your doctor will talk to you about your symptoms, perform a physical examination and perform a blood test to check for antibodies made by your immune system. They will also order a range of other tests to see if any other parts of your body are involved. A team of healthcare professionals will assess your individual case. For example, sometimes a specialist opinion from a rheumatologist is required, who may also perform a detailed examination of your nailbeds to help confirm the diagnosis.

If it is suspected that scleroderma has affected your lungs, your doctor will perform additional tests to determine the extent of the ILD. It is rare to need a biopsy to confirm a diagnosis of SSc-ILD.

Tests which may be used to diagnose SSc-ILD include:



High-resolution Computed Tomography (HRCT) scan of your chest to help determine if you have SSc-ILD and the effect on your lungs

Lung-function tests help to determine how well your lungs are working and to help determine if you need treatment.

I have been diagnosed with SSc-ILD – what next?

Many cases of SSc-ILD remain stable and do not require any specific treatment. In cases where SSc-ILD does progress, the rate varies and is difficult to predict the course of the disease. In some cases, SSc-ILD may progress slowly, in others it may deteriorate more quickly. Patients with more lung damage are at higher risk of developing complications.

Advanced SSc-ILD can cause severe symptoms and is life-threatening. It is therefore important that your lungs are monitored regularly by a respiratory specialist, so that treatment can be started if your lung health deteriorates.

How is SSc-ILD treated?

Where treatments for SSc-ILD are required, the aim of treatment is to preserve and stabilise lung function. While it is unlikely your lungs will improve significantly on therapy, in most cases the goal is to prevent further deterioration.

Your treatment will be tailored to suit you. You may not need any medication at all to start with, or you may receive a combination of medications. Your medication requirements might also change over time. If you have SSc-ILD, there are a range of treatments available to help your lungs such as:

- **Immunosuppressive medications** such as prednisolone, mycophenolate, azathioprine or cyclophosphamide
- **Monoclonal antibodies** such as rituximab may be considered if your condition doesn't respond to immunosuppressive medications. These medications work by lowering the activity of some of the immune system pathways that trigger inflammation.
- **Anti-fibrotic therapies** such as nintedanib may be used if you have progressive scarring (fibrosis) on your lungs despite immunosuppressive treatment
- **Stem-cell and lung transplantation** may be possible in rare severe cases

When visiting your respiratory specialist clinic, you will also be assessed for Pulmonary Hypertension (PH), a condition that causes high blood pressure in the lungs and symptoms such as breathlessness.

Assessment for PH may include an echocardiogram (ultrasound of the heart) or a right heart catheter investigation with a cardiologist. If PH is diagnosed, your specialist will discuss the treatment options available to treat PH and lower the blood pressure in your lungs.

It is important to look after yourself when living with SSc-ILD to get the most out of your treatment and care. There are a range of ways to help manage your symptoms and maintain your health, including:

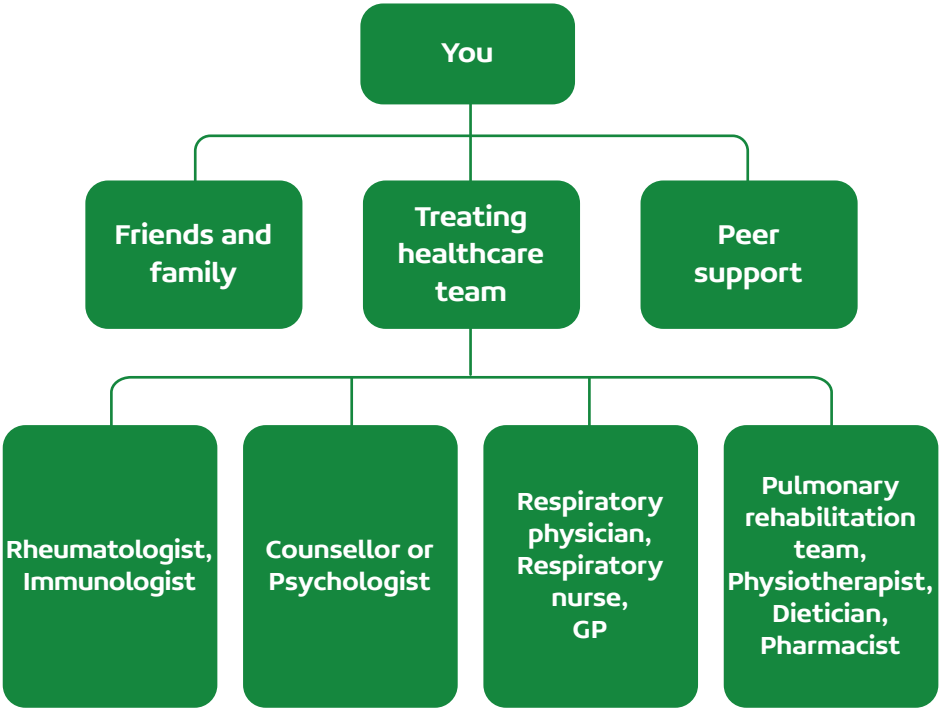
- Participating in a pulmonary rehabilitation program
- Maintaining a healthy diet and daily exercise – to learn more, visit the **Life with PF** booklet
- Keeping up to date with vaccinations
- Oxygen therapy if prescribed – to learn more, visit the **How can oxygen therapy help?** booklet
- Speaking with your healthcare team about whether a referral to palliative care would be beneficial as part of your ongoing care. To learn more about palliative care, see our **How can palliative care help you** resource.

For more information on looking after yourself, see our **Living with PF: Non-pharmacological Treatments** resource.

What support is available?

Being diagnosed with SSc-ILD can be difficult to process. It can be a serious condition that can cause, in some cases, severe symptoms, which is why it is important to stay connected with your healthcare team and seek support when you need it.

Your healthcare and support team may include:



To connect with support services and additional information, contact Lung Foundation Australia's Information and Support Centre on **1800 654 301** (option 3) or learn more **here.**

LUNG FOUNDATION AUSTRALIA SERVICES

- Information and Support Team
- Lung disease information resources
- Education webinars
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- E-newsletter

EXTERNAL LINKS

Scleroderma Australia	www.sclerodermaaustralia.com.au
American College of Rheumatology	www.rheumatology.org/patients/scleroderma

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