

# Scleroderma-associated Interstitial Lung Disease (SSc-ILD)

## What is scleroderma-associated interstitial lung disease?

Scleroderma-associated interstitial lung disease, or “SSc-ILD”, is a chronic lung disease in which scar tissue (“fibrosis”) and/or inflammation builds up in the walls of the air sacs of the lungs in a person with a diagnosis of scleroderma (also called “systemic sclerosis” or “SSc”).

Scleroderma is an “autoimmune” condition. The word “autoimmune” means that the body’s immune system is causing inflammation and other problems in the body’s own organs. There are many autoimmune diseases other than scleroderma, including systemic lupus erythematosus, rheumatoid arthritis, dermatomyositis, and others. People living with scleroderma often have thickening and tightening of the skin on the fingers and other parts of the body. Scleroderma can also affect the kidneys, GI tract (particularly the esophagus), and lungs.

Many people with scleroderma do not have lung disease. When scleroderma does affect the lungs, it most frequently leads to one of two conditions: ILD or pulmonary hypertension. Some people have both ILD and pulmonary hypertension. People taking medications that treat scleroderma by weakening the immune system (such as mycophenolate mofetil, cyclophosphamide, rituximab, and others) can also develop lung infections, such as pneumonia.

## What are the symptoms of SSc-ILD?

Most people with early SSc-ILD will have no respiratory symptoms or might have a bothersome cough. As the disease progresses, breathlessness during exercise and daily activities becomes common. Fatigue, depression, and anxiety are also common.

## What causes SSc-ILD?

No one is certain what causes either scleroderma or SSc-ILD.

## How is SSc-ILD diagnosed?

When a doctor or other healthcare provider suspects that a patient has ILD, they will perform tests that might include pulmonary function tests, a chest x-ray, blood work, and a high-resolution CT scan (also known as CAT scan). Pulmonary function tests measure how much air the lungs can hold. The more scar tissue the lungs have, the less air they will hold. A doctor will also use a special kind of X-ray of the chest, called a high-resolution computed tomography (HRCT) scan, which gives detailed images of the lung. Healthy lung tissue looks nearly black on a CT scan). Scar tissue and inflammation both appear grey or white.

Thickening or tightening of the skin on the fingers, arms, face or neck are an important clue that scleroderma might be present. Certain other changes in the fingertips can also help identify someone who has scleroderma. Blood tests, such as an anti-nuclear antibody (“ANA”) and an anti-Scl70 antibody, can also help identify scleroderma. In most cases, a specialist in autoimmune diseases (a rheumatologist) would help establish a diagnosis of scleroderma.

TYPE OF PULMONARY FIBROSIS	CLUES THAT DOCTORS USE
Drug-induced	Prior or current use of amiodarone, nitrofurantoin, chemotherapy, methotrexate, or other drugs known to affect the lungs
Radiation-induced	Prior or current radiation treatment to the chest
Environmental (called hypersensitivity pneumonitis)	Exposure to mold, animals, or other triggers
Autoimmune (called connective tissue disease-related)	Joint inflammation, skin changes (particularly on the fingers and face), dry eyes or mouth, abnormal blood tests
Occupational (called pneumoconiosis)	Prior or current exposure to dusts, fibers, fumes, or vapors that can cause PF (such as asbestos, coal, silica, and others)
Idiopathic	When no cause can be identified

## How is SSc-ILD treated?

There is no one best way to treat SSc-ILD. For some patients, monitoring without therapy is appropriate. For others, immunosuppressive medications (that suppress the immune system) such as mycophenolate mofetil, or cyclophosphamide, or azathioprine are used. **Nintedanib** is an antifibrotic medication that can slow the progression of SSc-ILD.

**Lung transplantation** is an appropriate treatment for some people living with SSc-ILD. Early evaluation for lung transplant is important because the process involves a series of appointments to provide the patient with information about transplantation and to determine if they are an appropriate candidate.

**Pulmonary rehabilitation, supplemental oxygen**, smoking cessation, routine vaccinations (such as influenza and pneumonia vaccination) are important parts of living with SSc-ILD.

You can learn more about supplemental oxygen at:

<https://www.pulmonaryfibrosis.org/life-with-pf/oxygen-therapy>.

You can learn more about pulmonary rehabilitation at:

<https://www.pulmonaryfibrosis.org/life-with-pf/pulmonary-fibrosis-treatment-options>

## What is my prognosis?

The prognosis in SSc-ILD is highly variable. Some cases remain stable while others get worse over time. The rate of progression can vary from being slow over years and at other times rapid. Progression of the ILD may lead to lung failure (medically called “respiratory failure”), which is a life-threatening condition. No one can predict exactly how long you will live with SSc-ILD. Everyone is different. Your doctor can give you more detailed information about your prognosis.

## Are there experimental therapies available?

You can search for research studies closest to you on our PFF Clinical Trial Finder: [trials.pulmonaryfibrosis.org](https://trials.pulmonaryfibrosis.org).

## Other Resources

[Scleroderma Foundation](#)

## Acknowledgements

The Pulmonary Fibrosis Foundation is thankful to the following for their assistance in writing and reviewing this fact sheet:

David J. Lederer, MD, MS  
Former PFF Senior Medical Advisor, Education and Awareness