

Connective Tissue Disease-Associated Interstitial Lung Disease (CTD-ILD)

Read this handout to learn about:

- signs and symptoms of CTD-ILD
- how your doctor will know if you have CTD-ILD
- tests and treatments you may need
- where to get more information

What is Connective tissue disease-associated interstitial lung disease (CTD-ILD)?

CTD-ILD is a type of lung disease that may happen to some people with connective tissue disease.

Examples of connective tissue diseases (also known as rheumatologic, collagen vascular, or autoimmune diseases) are:

- Scleroderma
- Rheumatoid arthritis
- Sjogren's syndrome
- Mixed or undifferentiated connective tissue disease
- Systemic lupus erythematosus
- Polymyositis
- Dermatomyositis

In many cases, people are diagnosed with the connective tissue disease first and develop CTD-ILD later. In some cases the lung disease develops first.



CTD-ILD causes inflammation, scarring (fibrosis) or both of the lungs. The exact reason leading to lung damage is not known.

What are some of the signs and symptoms of CTD-ILD?

Sometimes people with CTD-ILD do not have any symptoms. But some common signs and symptoms of are:

- Shortness of breath with activity
- Cough
- Fatigue (feeling very tired)
- “Crackles” in the lungs that your doctor can hear with a stethoscope
- Symptoms and signs of a connective tissue disease (for example, joint pain or swelling, rash, dry eyes, dry mouth, acid reflux)

How will my doctor know if I have CTD-ILD?

To find out if you have CTD-ILD, your doctor will start by talking to you about your medical history and he or she will do a physical examination. This will include talking with you about your medications, any occupational and environmental exposures that might cause CTD-ILD, listening to your chest with a stethoscope, and examining your skin and joints.

What tests will I need?

Pulmonary Function Testing (PFT)

This breathing test will measure:

- the flow of air in your lungs
- the volume of air in your lungs
- how well your lungs take oxygen from the air

High Resolution Computed Tomography (HRCT)

This is a special type of CT scan that makes detailed pictures of your lungs. The scan can take pictures while you lie on your back and while you lie on your chest. It can also take pictures while you breathe air out of your lungs. The HRCT is a very valuable test to help your doctor find out whether or not you have CTD-ILD.

Having an HRCT is the same as having a regular CT scan. Both scans have you lie on an open-air table and only take a few minutes. Only the steps for doing a HRCT are different.

Blood Tests

Blood tests will show if you have other diseases. They will check for antibodies. These will show whether you have a connective tissue disease, such as rheumatoid arthritis or scleroderma. Some people with these diseases have lung problems before they get any other symptoms.

Referral to a Rheumatologist

For some people, interstitial lung disease is the first sign that you may also have a connective tissue disease. If this is the case, you may need to see a Rheumatologist (a specialist in connective tissue diseases and auto-immune diseases).

What is the treatment for CTD-ILD?

The treatment for CTD-ILD usually involves taking anti-inflammatory or immunosuppressive medications. You may recognize some of these medications as they may have been prescribed to you for your connective tissue disease in the past.

These are the most common anti-inflammatory and immunosuppressive medications used to treat CTD-ILD:

- Corticosteroid (Prednisone)
- Cyclophosphamide (Cytoxan)
- Mycophenolate mofetil (Cellcept)
- Mycophenolate sodium (Myfortic)
- Azathioprine (Imuran)

If you have any questions about these medications, ask one of our team members for a pamphlet, or talk to your doctor or pharmacist.

Pulmonary rehabilitation is an important and effective treatment for patients with CTD-ILD. Pulmonary rehabilitation is an exercise and educational program for patients with lung disease.

Lung transplant may be an effective treatment option for some patients. Your doctor will talk to you about this, if needed.

Careful attention to certain other medical problems you may have such as gastroesophageal reflux disease (GERD) or pulmonary arterial hypertension (PAH) may also be important.

Who can I talk to for more information about CTD-ILD?

If you have questions about CTD-ILD, please talk to your doctor.

Where to get more information online

Scleroderma Foundation

Website: <http://www.scleroderma.org>



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