

Connective tissue disease-associated interstitial lung disease (CTD-ILD)



UHN

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

Connective tissue disease-associated interstitial lung disease (CTD-ILD) is a type of lung disease that may happen to some patients with connective tissue disease.

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

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

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

CTD-ILD causes inflammation, scarring (fibrosis) or both of the lungs. The exact mechanism that leads to lung damage is unknown.

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

Sometimes patients with CTD-ILD do not have any symptoms. However, some common signs and symptoms of CTD-ILD are:

- Shortness of breath with activity
- Cough
- Fatigue
- "Crackles" on examination of the chest with a stethoscope
- Symptoms and signs of a connective tissue disease (for example, joint pain or swelling, rash, dry eyes, dry mouth, acid reflux)

Please visit the UHN Patient Education website for more health information:

www.uhnpatienteducation.ca

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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 also 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.

In addition to a careful medical history and physical examination, you may need to have the following tests to help your doctor make an accurate diagnosis:

Pulmonary Function Testing (PFT) – This test involves a series of breathing tests that measure your lungs' airflows, the volume of air in your lungs, and the ability of your lungs to extract oxygen from the air.

High Resolution Computed Tomography (HRCT) – This is a special type of CT scan that provides your doctor with high- resolution images of your lungs. Images may be taken in several different ways: while you are lying on your back, lying on your chest, and possibly, while having you breathe air out of your lungs. The HRCT is an extremely valuable test to help your doctor find out whether or not you have CTD-ILD.

- Having a HRCT is no different than having a regular CT scan but requires a special protocol; they both are performed on an open-air table and take only a few minutes.

Blood Tests – Blood tests (serology) that will look for various antibodies may be needed to see if you have a connective tissue disease, such as Rheumatoid arthritis or Scleroderma.

- Autoantibodies are antibodies (immune factors normally produced to fight infection) that react to and damage your own body tissues, Some patients with these connective tissue diseases develop lung problems before other parts of the body are affected.

Referral to a Rheumatologist: For some patients, 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) for further evaluation.

What is the treatment for CTD-ILD?

The treatment for CTD-ILD usually involves the use of 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.

The following 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 the above medications, just 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 designed just 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.

Where can I find more information about CTD-ILD?

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

Online Resources

<http://www.scleroderma.org>