

Connective tissue and interstitial lung disease (CTD-ILD)

Respiratory

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Information for Patients

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Introduction

This leaflet will inform you about what connective tissue disease associated with interstitial lung disease (CTD-ILD) is, including:

- signs and symptoms
- tests you may need
- treatments
- where to get more information

CTD-ILD is a lung condition that affects a small number of patients with a connective tissue disease. CTD-ILD causes inflammation or scarring (fibrosis) of the lungs, or both. The exact cause of the lung damage is not known. This damage is in the tissue of the lung rather than the airways.

Examples of connective tissue diseases which are 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

Patients are often diagnosed with the connective tissue disease first. Later they start having CTD-ILD, and in some cases the opposite happens.

**Health information and support is available at www.nhs.uk
or call 111 for non-emergency medical advice**

Visit www.leicestershospitals.nhs.uk for maps and information about visiting Leicester's Hospitals
To give feedback about this information sheet, contact InformationForPatients@uhl-tr.nhs.uk

Signs and symptoms

Some patients with CTD-ILD do not have any symptoms. For others, common symptoms include:

- shortness of breath when moving around
- cough
- tiredness (fatigue)
- "crackle" sounds which are heard when listening to the chest with a stethoscope
- symptoms of a connective tissue disease, such as joint pain and swelling, rash, dry eyes, dry mouth and acid reflux

The diagnosis of CTD-ILD is made by a team experienced in interstitial lung disease.

In making a diagnosis, your doctor will start by

- taking a thorough medical history and
- carrying out a physical examination, including your skin and joints.

This will include discussing your health history, reviewing medications and talking about your job and things in your environment that could be having an effect on your lungs.

Tests you may need

- **Pulmonary function testing:** This test involves a series of breathing exercises that measures how well air flows through your lungs, the volume of air in your lungs, and the ability of your lungs to get oxygen from the air into the body.
- **High resolution computed tomography (HRCT):** This is a special type of CT scan that gives very good images of your lungs.

Images may be taken in several different positions

- with you lying on your back,
- lying on your chest, and
- when breathing air out of your chest.

These various images are extremely valuable in finding out whether or not you have CTD-ILD. The test is done on an open-air table and takes only a few minutes.

- **Blood tests:** Blood tests look for various antibodies common in connective tissue disease problems. Some patients with connective tissue diseases start having lung problems before the more common symptoms arise. In these cases blood test results may be the only proof of CTD-ILD.

For some patients, interstitial lung disease is the first sign of a connective tissue disease. If this is the case, we may refer you to a rheumatologist for further tests or to clinic where you will see a rheumatologist and a respiratory consultant.

Treatments

CT-ILD is treated with anti-inflammatory or immunosuppressive medications. You may notice some or all of these medications if they were prescribed to you for your connective tissue disease.

Medication

The most common medications used to treat CT-ILD are:

- corticosteroid (prednisolone)
- cyclophosphamide (Cytoxan)
- mycophenolate mofetil (Cellcept)
- azathioprine (Imuran)
- rituximab (Rituxan)

These medications may need regular blood tests and you should ask for an information leaflet to understand these medications.

Exercise

Exercise is important to keep you as fit as possible, 1 of the easiest exercises is regular walking.

Pulmonary rehabilitation is another important and effective treatment for patients with CTD-ILD. Pulmonary rehabilitation is an exercise and education program designed for patients with long term lung disease. If you have not already, ask for a referral.

Diet

It is also important to have a good balanced diet and to keep an eye on your weight.

It is also important that any other associated medical problems, such as gastroesophageal reflux disease (GERD) and pulmonary hypertension are treated.

Lung transplantation may be an option for some patients whose symptoms are getting worse and are not getting better with medication.

Contact details

The ILD Team can be contacted on 0116 258 3949.

Further information

If you need further information, talk to the ILD team member or visit these websites:

- National Rheumatoid Arthritis Society: <https://www.nras.org.uk/groups>
- Scleroderma Foundation: <http://www.scleroderma.org>
- British Sjogren's Syndrome Association (BSSA): <https://www.bssa.uk.net>
- Sjogrens Syndrome Foundation (SSF): <https://www.sjogrens.org/home/get-connected/support-groups>
- LUPUS UK: <https://www.lupusuk.org.uk>
- Myositis Support Group – Myositis UK: <https://www.myositis.org.uk/>
- MCTD Foundation - Mixed Connective Tissue Disease: <https://mctdfoundation.org/>

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