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

Centre of Research Excellence in Pulmonary Fibrosis

Lung Foundation Australia

What is Connective Tissue Disease-associated Interstitial Lung Disease?

Connective Tissue Disease-associated Interstitial Lung Disease, commonly referred to as CTD-ILD, is a group of Interstitial Lung Diseases that are caused by a connective tissue disease such as systemic sclerosis, rheumatoid arthritis, inflammatory myopathies and many other autoimmune conditions. Connective tissue diseases are conditions affecting the parts of the body that connect body structures together such as joints and skin, but can involve other organs. Interstitial Lung Disease (ILD) is characterised by thickening or scarring of lung tissue. ILD can affect people with all types of connective tissue disease, but not everyone with connective tissue disease will get CTD-ILD.

If you have CTD-ILD, you are not alone:

About **18%** of people with an Interstitial Lung Disease have an associated diagnosis of CTD-ILD

What are the symptoms of CTD-ILD?

Symptoms of CTD-ILD vary. In addition to the symptoms you may experience from ILD, there may be a wide variety of other possible symptoms related to the underlying connective tissue disease. You may experience a few symptoms or none at all. Everyone is different.

General wellbeing symptoms
Fatigue, unusual weight loss or fever that is not due to an infection

Respiratory symptoms
Unusual breathlessness, for example, when going up stairs and/or a persistent dry cough

For some people, respiratory symptoms may occur before other typical symptoms of connective tissue diseases, such as joint and skin symptoms

Joint and skin symptoms
These will vary depending on your connective tissue disease, but may include:

- Muscle weakness or aches (such as difficulty lifting your arms or getting out of a chair)
- Joint pain, including stiffness in the morning
- Raynaud's phenomenon in which your fingers may turn different colours (such as white) in the cold and can be associated with discomfort or numbness
- Rashes or other changes of your skin
- Fingertip ulcers
- Dry eyes and mouth

Gastrointestinal symptoms
Gastro-oesophageal reflux that may be difficult to control, problems with swallowing or issues with your bowels

If you are experiencing any of these symptoms and they are new, changing or worsening, or if you have had the symptoms for a while and they just don't feel right, make sure you speak with your doctor.

How is CTD-ILD diagnosed?

Depending on your circumstances, the initial diagnosis may be made by your GP, respiratory physician, immunologist or rheumatologist. In most cases, a diagnosis of CTD-ILD is made at a multidisciplinary meeting, where members of your healthcare team meet to discuss a range of investigations including the following:

- **Blood tests** looking for autoantibodies and inflammatory markers
- **High-resolution Computed Tomography (HRTC) scan** looking at your lungs
- **Lung-function (breathing) tests** looking to see how well your lungs are working
- **Echocardiogram** which is an ultrasound of your heart
- **Other specialised tests** such as bronchoscopy or biopsy

"Diagnosis can be challenging…[make sure you] go back to the GP to let them know that you think something is going on and it may take multiple visits to figure it out".

Tania, NSW, living with CTD-ILD

Some people who receive a diagnosis of CTD-ILD may experience emotional distress. This is quite a normal reaction and it is important to seek support from family, friends, and your treating healthcare team. However, seeking more professional support from a psychologist is often a good idea too. A psychologist can help you with strategies to manage difficult feelings that may arise.

How is CTD-ILD treated?

Not everyone will need treatment for their CTD-ILD. There are a range of factors you and your treating healthcare team will consider when deciding what is right for you, including:

- **Goals of treatment:** which may include stabilising or reversing your connective tissue disease, or slowing progression of your lung disease.
- **The Type of medication:** which will depend on the type of CTD-ILD you have. Treatment for your lung condition may also improve your other symptoms of connective tissue disease. Potential treatment medications, as shown in the table below, include immunosuppressive medications that help treat inflammation, and anti-fibrotic medications that help slow down lung fibrosis (scarring).

Immunosuppressive medications	Anti-fibrotic medications
• **Prednisolone** • **Mycophenolate** • **Azathioprine** • **Methotrexate** • **Cyclophosphamide** • **Rituximab**	• **Nintedanib**

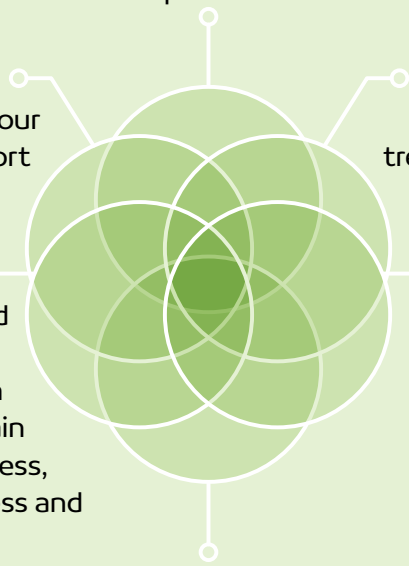
- **Treatment side effects:**
  - Some immunosuppressive and anti-fibrotic medications may have unwanted side effects. The types of side effects you may experience vary depending on what medication you are taking. It is important to tell your doctor as soon as possible if you notice any possible side effects or do not feel well while taking your medication. For more detailed information on the side effects of your medication, visit the Australian Register of Therapeutic Goods (ARTG) Consumer Medicines Information database.

- If you are considering having a baby and/or breastfeeding, make sure you discuss your plans with your treating healthcare team as there are some CTD-ILD medications that should not be taken.

- **Non-pharmacological treatments:** such as pulmonary rehabilitation, oxygen therapy, psychological support, nutrition and smoking cessation. For more information, see the Living with Pulmonary Fibrosis: Non-pharmacological treatments resource here.

Your treating healthcare team will work with you to develop a treatment plan that works for you, which may change over time.

Your treating healthcare team may include:



Medical team

Including your GP, respiratory specilaist/ ILD physician, rheumatologist, cardiologist and pharmacist

Community teams

Help you navigate your care and offer support when you need it

Specialist nurses

Help manage your treatment and disease

Physiotherapists

Help get you started with pulmonary rehabilitation, which can help you maintain or improve your fitness, reduce breathlessness and improve your mood

Palliative care
(if recommended by your healthcare team)

Help you manage your symptoms

Psychologists

Help you manage stressors associated with your condition

A team approach to the management of your condition is vital, and your GP or specialist will help co-ordinate with all of the different practitioners in your team.

What else can you do to look after yourself?

To help feel your best and get the most out of your CTD-ILD treatment, it is important to:

- Stay connected with your healthcare team to monitor your condition and discuss any questions or concerns you may have
- Keep your vaccinations up to date
  - Speak with your treating healthcare team about which vaccinations are suitable for you, as some medications can affect the effectiveness of vaccines
- Participate in regular exercise to stay fit and active
- Maintain a healthy diet
- Seek support when you need it
  - Stay connected with your family and friends
  - Join support groups to connect with people who are dealing with similar experiences

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

Therapeutic Goods Administration (TGA) - Consumer Medicines Information (CMI)	www.tga.gov.au
Pulmonary Fibrosis Foundation	www.pulmonaryfibrosis.org
Canadian Pulmonary Fibrosis Foundation	www.cpff.ca

This fact sheet was based on the 2022 'Introduction to Connective Tissue Disease-associated Interstitial Lung Disease' webinar by Dr Adelle Jee, Respiratory and Sleep Physician and CTD-ILD consumer advocate, Tania Hyde.

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