

Rheumatoid Arthritis-associated Interstitial Lung Disease (RA-ILD)

What is rheumatoid arthritis-associated interstitial lung disease?

Rheumatoid arthritis-associated interstitial lung disease, or “RA-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 rheumatoid arthritis. Rheumatoid arthritis, or “RA”, 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 RA, including systemic lupus erythematosus, scleroderma, dermatomyositis, and others. RA primarily causes arthritis, but people living with RA can also develop lung disease, problems with the nerves or eyes, and other types of inflammation in the body.

Many people with RA do not have lung disease. When RA does affect the lung, it can lead to many different lung conditions. ILD is the most common problem in the lungs of people living with RA. Other conditions that RA can cause include nodules in the lungs (known as “pulmonary nodules”), and problems with the airways in the lung, problems with the outside lining of the lung (known as “pleural disease”). People taking certain medications to treat RA can have side effects that involve the lung, including lung infections (due to a weakened immune system) and even ILD due to medications. When doctors use the term RA-ILD, they are usually referring only to ILD involving the lungs in someone living with RA.

What are the symptoms of RA-ILD?

People with RA-ILD can have symptoms related to problems in their lungs and joints. In the lungs, most people with early RA-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 RA-ILD?

No one is certain what causes either RA or RA-ILD. Some risk factors for RA include smoking and certain genetic mutations in a family of genes called “human leukocyte antigen”, or “HLA” genes. There is a theory that inflammation of the lungs might trigger RA itself, meaning that the condition might start in the lungs in some people and then spread to the joints. In people with RA, the major risk factor for developing ILD is smoking. Other risk factors for ILD include being older, being a man, and having more severe RA.

How is RA-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.

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

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.

Pain, stiffness, and swelling in the joints would be an important clue to the presence of RA. Blood tests, including “rheumatoid factor” (also known as “RF”) and “anti-cyclic citrullinated peptide antibody” (also known as “anti-CCP”) tests, can help identify the presence of RA. In most cases, a specialist in autoimmune diseases (a rheumatologist) would help establish a diagnosis of RA.

How is RA-ILD treated?

There is no FDA-approved therapy for RA-ILD, and there are no medical guidelines on how to treat RA-ILD. In some cases, medications that weaken the immune system are used to treat inflammation in the lung. In some patients, doctors will monitor the disease without medications. Treatments for joint disease with RA do not necessarily treat the lung disease.

Lung transplantation is an appropriate treatment for some people living with RA-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 RA-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 RA-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 RA-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.

Other Resources

[American College of Rheumatology](#)

[Arthritis Foundation](#)

[American Autoimmune Related Diseases Association](#)

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