Myositis-Associated Interstitial Lung Disease

Pulmonary Fibrosis Foundation™

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What is myositis-associated interstitial lung disease?

Myositis-associated interstitial lung disease (ILD) is a chronic lung disease in which inflammation and/or scar tissue ("fibrosis") builds up in the walls of the air sacs of the lungs. Myositis-associated ILD is one of the clinical manifestations in patients with idiopathic inflammatory myopathies, also known as myositis. The term myositis is used to refer to a disease involving chronic inflammation of the muscles, often occurring together with other symptoms. It is characterized by skeletal muscle weakness and inflammation, abnormal muscle enzymes, and myositis-related autoantibodies. Autoantibodies are proteins in the blood directed against self (Table 1). Common types of myositis include polymyositis, dermatomyositis, and anti-synthetase syndrome. Myositis-associated ILD falls into a large category of diseases called "interstitial lung diseases" or "ILD." There are over 200 types of ILD. ILDs include all of the diseases that have inflammation and/or scarring in the walls of the air sacs. "Pulmonary fibrosis" (PF) is used to refer to any type of ILD in which fibrosis (also known as scarring) is present.

What are the symptoms of myositis-associated interstitial lung disease?

People with myositis-associated interstitial lung disease can experience symptoms including breathlessness during activities or when coughing. People can also experience muscle weakness involving arms, legs, and neck, as well as fevers, joint pain, discoloration of fingers or toes, fatigue, or rash. These symptoms can also overlap with other disease conditions and are not specific to a myositis-associated interstitial lung disease diagnosis.

What causes myositis-associated interstitial lung disease?

A specific cause for myositis-associated interstitial lung disease has not been identified but there are genetic associations between myositis and interstitial lung disease.

How is myositis-associated interstitial lung disease diagnosed?

The diagnosis of myositis-associated ILD is usually made by a lung doctor (a pulmonologist) in conjunction with a specialist in autoimmune diseases (a rheumatologist). When a doctor or other healthcare provider suspects that a patient with myositis has ILD, they will perform tests that include pulmonary function tests (PFTs), blood work, and a high-resolution CT scan (also known as CAT scan). Pulmonary function tests provide an estimate of lung function.

Brief overview of findings on HRCT

Normal lung tissue looks dark gray/black on a CT scan. Inflammation and/or scar tissue appears light gray or white. Your doctor can review the results of your CT scan to show you the areas that are abnormal.

How is myositis-associated interstitial lung disease managed?

Your doctor will likely recommend monitoring via regularly scheduled office visits, pulmonary function tests, and blood work. Depending on your symptoms, test results, and disease activity, your doctor may monitor the disease without medications. In other situations, your doctor may use medications that affect the immune system to help stabilize your lung function or other medications to slow decline of the lungs. Steroid therapy is the main management strategy for myositis-associated interstitial lung disease. To avoid unwanted side effects from long-term steroid use, most patients will eventually transition from steroids to medications known as steroid-sparing agents. These medications include: mycophenolate mofetil, azathioprine, rituximab, cyclophosphamide, and/or intravenous immunoglobulin (IVIG).

Pulmonary rehabilitation, supplemental oxygen, smoking cessation, vaccinations (including COVID-19, influenza, and pneumonia vaccines) are vital to living with myositis-associated interstitial lung disease.

You can learn more about supplemental oxygen at **pulmonaryfibrosis.org/oxygentherapy.**
You can learn more about pulmonary rehabilitation at **pulmonaryfibrosis.org/pulmonaryrehab**.

Lung transplantation may be an option for some people living with myositis-associated interstitial lung disease. Early evaluation for lung transplant is important because the process involves a series of appointments and tests to determine if patients are appropriate candidates. You can learn more about lung transplantation at **pulmonaryfibrosis.org/lungtransplant.**

What is my prognosis?

The prognosis in myositis-associated interstitial lung disease is variable. Some cases remain stable while others can experience lung function decline over time. The rate of progression can be slow or rapidly progressive. Progression of the lung condition can lead to respiratory failure which is a life-limiting condition. No one can predict exactly how long you will live with myositis-associated interstitial lung disease, and every individual is different. Your doctor may be able to give you more detailed information about your prognosis.

Are there experimental therapies available?

Myositis-associated interstitial lung disease is an emerging area of research. Research studies can be found at PFF Clinical Trial Finder: **trials.pulmonaryfibrosis.org** or **clinicaltrials.gov**.

Other References

https://www.atsjournals.org/doi/10.1164/rccm.202202-0399ST (ATS guidelines 2022)

Myositis Autoantibodies*	
Anti-Jo1	Anti-PM/Scl
Anti-PL7	Anti-Ro-52
Anti-PL12	Anti-Ku
Anti-EJ	Anti-U1RNP
Anti-OJ	Anti-U2 SNRNP
Anti-MDA5	Anti-Fibrillarin (U3 RNP)
Anti-NXP2	Anti-SRP
Anti-Mi2	

* this is an abbreviated list.

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