What Is Interstitial Lung Disease?

Interstitial lung disease is a group of conditions that cause inflammation and scarring in the lungs.

Patients with interstitial lung disease typically have shortness of breath with exertion that may progress to difficulty breathing while at rest. About 30% of people with interstitial lung disease have cough, and some have flare-ups (exacerbations) that cause episodes of rapidly worsening shortness of breath over several days to weeks.[1]

Types and Causes of Interstitial Lung Disease

The most common type of interstitial lung disease is idiopathic pulmonary fibrosis (IPF), which has no known cause. Conditions associated with interstitial lung disease include connective tissue diseases, such as rheumatoid arthritis or scleroderma; hypersensitivity pneumonitis, caused by environmental exposures such as mold, fungus, or birds; medications, such as amiodarone, nitrofurantoin, bleomycin, and certain cancer immunotherapies; and infections, such as COVID-19.

Who Is Affected by Interstitial Lung Disease?

The highest rate of interstitial lung disease occurs in people aged 80 to 84 years, although the average age at diagnosis is 67 to 72 years. In the US, more than 650 000 people have interstitial lung disease. Overall, interstitial lung disease is more common in females, but IPF affects males about 3 times more often.

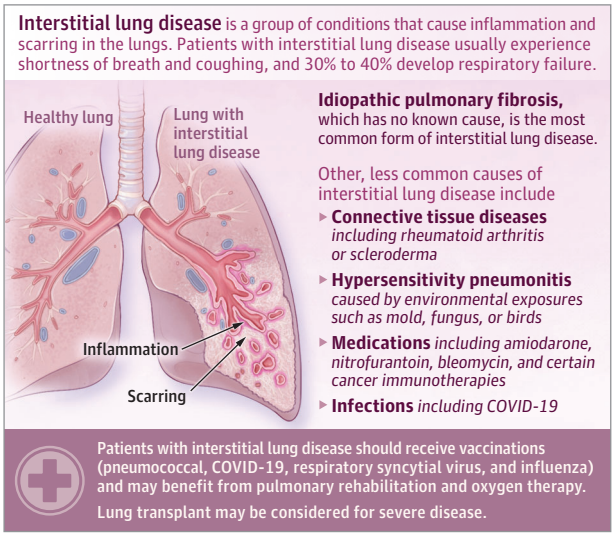
How Is Interstitial Lung Disease Diagnosed and Treated?

Diagnosis of interstitial lung disease is typically made based on symptoms, physical examination, and additional testing that typically includes chest computed tomography (CT), pulmonary function tests, and blood tests to evaluate for autoimmune diseases. Lung biopsy may be performed if the diagnosis is unclear.

Treatment depends on the cause and severity of interstitial lung disease. Medications known to cause interstitial lung disease should be discontinued, certain exposures (mold, birds) should be avoided, and medical conditions associated with interstitial lung disease (connective tissue diseases, infections) should be treated. First-line treatment for IPF is medication that may slow lung scarring (nintedanib or pirfenidone). Lung transplant may be considered for severe interstitial lung disease, although organ availability is limited, and many patients are excluded due to other medical conditions (diabetes, heart disease) and advanced age.

What Is the Prognosis of Interstitial Lung Disease?

Approximately 30% to 40% of patients with interstitial lung disease develop progressive lung scarring, which causes respiratory failure and is associated with an average survival of 2.5 to 3.5 years after diagnosis without lung transplant. Patients with interstitial lung disease who undergo lung transplant often have improved symptoms and an average survival of 5.2 to 6.7 years after transplant.



Supportive Care for Patients With Interstitial Lung Disease

Pulmonary rehabilitation, an 8- to 12-week program of endurance training and education, improves symptoms, quality of life, and walking distance in patients with shortness of breath due to interstitial lung disease. Pneumococcal, COVID-19, respiratory syncytial virus, and influenza vaccines should be offered to all patients with interstitial lung disease, and oxygen therapy is recommended for those with low blood oxygen levels. End-of-life planning and palliative care services are important for patients with respiratory failure. For patients with severe interstitial lung disease who are not candidates for lung transplant, mechanical ventilation should be avoided because it is associated with poor outcomes.

FOR MORE INFORMATION

National Heart, Lung, and Blood Institute

Authors: Amy E. Varghese, MD; Christopher D. Jackson, MD; Julio Lanfranco, MD, MPH

Published Online: July 3, 2025. doi:10.1001/jama.2025.9804

Author Affiliations: Division of General Internal Medicine, Department of Medicine, University of Tennessee Health Science Center, Memphis (Varghese); Division of General Internal Medicine, Department of Medicine and Medical Education, University of South Florida, Tampa (Jackson); Division of Pulmonary, Critical Care and Sleep Medicine, Department of Medicine, University of Tennessee Health Science Center, Memphis (Lanfranco).

Conflict of Interest Disclosures: None reported.

1. Maher TM. Interstitial lung disease: a review. *JAMA*. 2024;331(19):1655-1665. doi:10.1001/jama.2024.3669