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Pulmonary Fibrosis

Also called: Idiopathic pulmonary fibrosis, IPF

Pulmonary fibrosis is a condition in which the tissue deep in your lungs becomes scarred over time. This tissue gets thick and stiff. That makes it hard for you to catch your breath, and your blood may not get enough oxygen.

Causes of pulmonary fibrosis include environmental pollutants, some medicines, some connective tissue diseases, and interstitial lung disease. Interstitial lung disease [<https://medlineplus.gov/interstitiallungdiseases.html>] is the name for a large group of diseases that inflame or scar the lungs. In most cases, the cause cannot be found. This is called idiopathic pulmonary fibrosis.

Symptoms include:

- Shortness of breath
- A dry, hacking cough that doesn't get better
- Fatigue
- Weight loss for no known reason
- Aching muscles and joints
- Clubbing, which is the widening and rounding of the tips of the fingers or toes

Your doctor may use your medical history, imaging tests, a biopsy, and lung function tests to diagnose pulmonary fibrosis. There is no cure. Treatments can help with symptoms and improve your quality of life. They include medicines, oxygen therapy, pulmonary rehabilitation, or a lung transplant.

NIH: National Heart, Lung, and Blood Institute

Start Here

- Introduction to Pulmonary Fibrosis [<https://www.lung.org/lung-health-diseases/lung-disease-lookup/pulmonary-fibrosis/introduction>] (American Lung Association)
- What Is Idiopathic Pulmonary Fibrosis? [<https://www.nhlbi.nih.gov/health/idiopathic-pulmonary-fibrosis>]  (National Heart, Lung, and Blood Institute)
- What Is Pulmonary Fibrosis? [<https://www.pulmonaryfibrosis.org/understanding-pff/about-pulmonary-fibrosis/what-is-pulmonary-fibrosis>] (Pulmonary Fibrosis Foundation)

Diagnosis and Tests

- Arterial Blood Gas (ABG) Test [<https://medlineplus.gov/lab-tests/blood-oxygen-level/>]  (National Library of Medicine)
Also in Spanish [<https://medlineplus.gov/spanish/pruebas-de-laboratorio/prueba-de-gasometria-arterial/>]
- Bronchoscopy and Bronchoalveolar Lavage (BAL) [<https://medlineplus.gov/lab-tests/bronchoscopy-and-bronchoalveolar-lavage-bal/>]  (National Library of Medicine)
Also in Spanish [<https://medlineplus.gov/spanish/pruebas-de-laboratorio/broncoscopia-con-lavado-broncoalveolar/>]
- Lung Function Tests [<https://medlineplus.gov/lab-tests/lung-function-tests/>]  (National Library of Medicine)

Also in Spanish [<https://medlineplus.gov/spanish/pruebas-de-laboratorio/pruebas-de-funcion-pulmonar/>]

- Spirometry [<https://www.lung.org/lung-health-diseases/lung-procedures-and-tests/spirometry>]  (American Lung Association)
- Spirometry [<https://www.mayoclinic.org/tests-procedures/spirometry/about/pac-20385201?p=1>] (Mayo Foundation for Medical Education and Research)
Also in Spanish [<https://www.mayoclinic.org/es/tests-procedures/spirometry/about/pac-20385201?p=1>]
- Tests for Lung Disease [<https://www.nhlbi.nih.gov/health/lung-tests>]  (National Heart, Lung, and Blood Institute)
Also in Spanish [<https://www.nhlbi.nih.gov/es/salud/pruebas-pulmonares>]

Living With

- Maintain Your Health [<https://www.pulmonaryfibrosis.org/patients-caregivers/education-resources/maintain-your-health>] (Pulmonary Fibrosis Foundation)
- Pulmonary Rehabilitation: MedlinePlus Health Topic [<https://medlineplus.gov/pulmonaryrehabilitation.html>]  (National Library of Medicine)
Also in Spanish [<https://medlineplus.gov/spanish/pulmonaryrehabilitation.html>]

Specifics

- Familial Pulmonary Fibrosis [<https://www.nationaljewish.org/conditions/familial-pf?modal=1>] (National Jewish Health)

Genetics

- Hermansky-Pudlak syndrome: MedlinePlus Genetics [<https://medlineplus.gov/genetics/condition/hermansky-pudlak-syndrome>]  (National Library of Medicine)
- Idiopathic pulmonary fibrosis: MedlinePlus Genetics [<https://medlineplus.gov/genetics/condition/idiopathic-pulmonary-fibrosis>]  (National Library of Medicine)

Clinical Trials

- ClinicalTrials.gov: Pulmonary Fibrosis [<https://clinicaltrials.gov/search?cond=%22Pulmonary+Fibrosis%22&aggFilters=status:not%20rec>]  (National Institutes of Health)

Journal Articles

References and abstracts from MEDLINE/PubMed (National Library of Medicine)

- Article: The Role of Serum Prolidase Activity, MMP-1, MMP-7, and TGF- β Values... [<https://www.ncbi.nlm.nih.gov/pubmed/40733571>]
- Article: GSR Deficiency Exacerbates Oxidative Stress and Promotes Pulmonary Fibrosis. [<https://www.ncbi.nlm.nih.gov/pubmed/40723921>]
- Article: Current Understanding of Pulmonary Fibrosis: Pathogenesis, Diagnosis, and Therapeutic Approaches. [<https://www.ncbi.nlm.nih.gov/pubmed/40697404>]
- Pulmonary Fibrosis -- see more articles [<https://pubmed.ncbi.nlm.nih.gov/?term=%22Pulmonary+Fibrosis%22%5Bmajr%3Aexp%5D+AND+humans%5Bmh%5D+AND+english%5Bla%5D+AND+%22last+1+Year%22+%5Bdat%5D+NOT+%28letter%5Bpt%5D+OR+case+reports%5Bpt%5D+OR+editorial%5Bpt%5D+OR+comment%5Bpt%5D%29+AND+free+full+text%5Bsb%5D+>]

Reference Desk

- How the Lungs Work [<https://www.nhlbi.nih.gov/health/lungs>]  (National Heart, Lung, and Blood Institute)
Also in Spanish [<https://www.nhlbi.nih.gov/es/salud/pulmones>]

Find an Expert

- American Lung Association [<https://www.lung.org/>]
- National Heart, Lung, and Blood Institute [<https://www.nhlbi.nih.gov/>] 

- Pulmonary Fibrosis Foundation [<https://www.pulmonaryfibrosis.org/>]

Patient Handouts

- Blood gases [<https://medlineplus.gov/ency/article/003855.htm>] (Medical Encyclopedia)
Also in Spanish [<https://medlineplus.gov/spanish/ency/article/003855.htm>]
- Idiopathic pulmonary fibrosis [<https://medlineplus.gov/ency/article/000069.htm>] (Medical Encyclopedia)
Also in Spanish [<https://medlineplus.gov/spanish/ency/article/000069.htm>]
- Lung diffusion testing [<https://medlineplus.gov/ency/article/003854.htm>] (Medical Encyclopedia)
Also in Spanish [<https://medlineplus.gov/spanish/ency/article/003854.htm>]
- Pulmonary function tests [<https://medlineplus.gov/ency/article/003853.htm>] (Medical Encyclopedia)
Also in Spanish [<https://medlineplus.gov/spanish/ency/article/003853.htm>]



MEDICAL ENCYCLOPEDIA

- ACE blood test [<https://medlineplus.gov/ency/article/003567.htm>]
- Blood gases [<https://medlineplus.gov/ency/article/003855.htm>]
- Bronchoscopy [<https://medlineplus.gov/ency/article/003857.htm>]
- Chest x-ray [<https://medlineplus.gov/ency/article/003804.htm>]
- Idiopathic pulmonary fibrosis [<https://medlineplus.gov/ency/article/000069.htm>]
- Lung diffusion testing [<https://medlineplus.gov/ency/article/003854.htm>]
- Pulmonary function tests [<https://medlineplus.gov/ency/article/003853.htm>]

Related Health Topics

- Interstitial Lung Diseases [<https://medlineplus.gov/interstitiallungdiseases.html>]
- Pulmonary Rehabilitation [<https://medlineplus.gov/pulmonaryrehabilitation.html>]

National Institutes of Health

The primary NIH organization for research on *Pulmonary Fibrosis* is the National Heart, Lung, and Blood Institute [<http://www.nhlbi.nih.gov/>]

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