



# Patient education: Idiopathic pulmonary fibrosis (The Basics)

Written by the doctors and editors at UpToDate

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## What is idiopathic pulmonary fibrosis?

Idiopathic pulmonary fibrosis (also called "IPF") is a lung disease that makes it hard to breathe. It damages the air sacs in your lungs that send oxygen to the blood. This damage causes the lungs to be stiff. It also makes it hard for oxygen to reach the blood. This makes people with IPF cough and get short of breath.

People who get IPF are usually older than 50. It is a very serious illness that cannot be cured and gets worse over time. In some people, IPF can stay stable for several years before getting worse, or get worse gradually. But in others, it gets worse more quickly.

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## Why did I get IPF?

Doctors do not know exactly how IPF starts. The risk of IPF is greater for people who:

- Smoke or used to smoke
- Have breathed in a lot of toxic chemicals or pollution
- Have breathed in certain types of dust at work, over a long time
- Have family members with pulmonary fibrosis

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## What are the symptoms of IPF?

IPF can start very slowly, so it might not cause any symptoms at first. When it does, symptoms can include:

- Shortness of breath during exercise or other physical activity
- Dry cough

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## Are there tests for IPF?

Yes. Doctors can do:

- Blood tests to make sure that you don't have a different type of lung disease – There is no blood test for IPF.
- Breathing tests to see how well your lungs are working – Breathing tests can show if your shortness of breath is caused by IPF or another disease such as emphysema.
- A CT scan – This imaging test uses a special X-ray to create pictures of the inside of the body. It can show lung damage caused by IPF.
- If your doctor is not sure that you have IPF after the CT scan, they might do a lung biopsy. In this test, the doctor does surgery to take a small sample of tissue from your lung. Another doctor looks at the sample under a microscope for signs of IPF.

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## How is IPF treated?

There is no treatment to cure IPF. It usually gets worse slowly. But doctors can treat some of the symptoms. These treatments can include:

- Quitting smoking – If you smoke, the most important thing that you can do is to **stop smoking**.
- Flu and pneumonia vaccines – Get the flu shot every fall and the pneumonia vaccine at least once. Infections like the flu and pneumonia can hurt your lungs. It's important to try to prevent them.
- Oxygen – As IPF gets worse, some people need to breathe oxygen from a tank that they carry with them.
- Pulmonary rehab – In pulmonary rehab, people learn exercises and ways to breathe that can help with IPF symptoms.
- Medicine – Two medicines, [nintedanib](#) (brand name: Ofev) and [pirfenidone](#) (brand name: Esbriet), have been shown to slow lung damage. But they do not cure IPF.
- Clinical trials – If you are interested, you can join a research study of new medicines that might help people with IPF.
- Treatment for acid reflux – Acid reflux is when the acid that is normally in your stomach backs up into your esophagus. The esophagus is the tube that carries food from your mouth to your stomach. People who have acid reflux might need medicine to stop the acid reflux from making IPF worse.

- Lung transplant – This is surgery to replace 1 or both diseased lungs with healthy lungs. It is done only if a person with IPF meets certain conditions.

IPF sometimes gets worse very quickly, over a few days to weeks. If this happens, tell your doctor. Doctors can try to treat it with antibiotics and steroid medicines. (These are **not** the same as the steroids that some athletes take illegally.)

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