

Idiopathic Pulmonary Fibrosis (IPF)

WHAT PATIENTS AND CAREGIVERS NEED TO KNOW

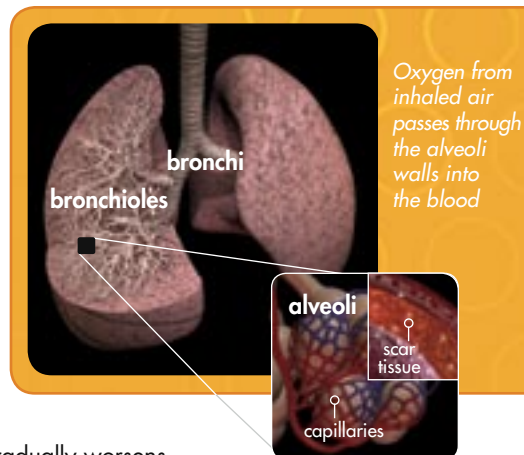
This fact sheet provides basic information about IPF and its treatment. It will answer some of your questions and offer resources for additional information and support.

WHAT IS IPF?

More than 80,000 people in the United States have been diagnosed with IPF, and each year about 30,000 new cases are identified.

You can understand more about IPF by learning what each part of its name means:

- **Idiopathic:** Of unknown cause. IPF is not thought to be related to any other disease or condition, such as cancer or asthma.
- **Pulmonary:** Occurring in the lungs. IPF affects the small air sacs (*alveoli*) in the lungs where oxygen enters the blood.
- **Fibrosis:** The development of scar (*fibrotic*) tissue. This scarring starts in the tissue between the alveoli and gradually worsens.



Over time, fibrosis continues to develop in the lungs, causing alveolar tissue to become thick and stiff. This makes it more difficult for the lungs to expand and harder for oxygen to get into the bloodstream.

POSSIBLE CAUSES AND RISK FACTORS

Although no one knows the exact cause of IPF, it is thought that a change takes place in the healing process of the lungs. Normal lungs would recover from inflammation caused by an illness, but lungs with this healing “malfunction” would allow scar tissue to develop without stopping. What causes this change is unknown.

It is known that people who smoke, or who have relatives who have IPF, are at higher risk for developing IPF. When more is known about IPF, it may be possible to identify specific causes so that people can take steps to reduce their risks.

SIGNS AND SYMPTOMS OF IPF

Early in IPF, a person might develop a dry cough (one that does not produce phlegm or mucus) and may also notice that normal physical activities like climbing stairs cause shortness of breath.

As fibrosis continues to develop, symptoms include shortness of breath and/or cough that occur without physical activity—such as while sitting, eating, or talking. Some patients also experience “clubbing” (enlargement of the fingertips) or fluid retention in their legs and arms which causes swelling and pain.

HELP FOR PATIENTS WHO HAVE IPF

There is no cure for IPF. However, there are therapies your doctor may recommend to help ease symptoms and enable you to breathe more easily. You will work closely with your doctor, nurse, and other team members to develop a therapy plan. It may include these important elements:

- **Pulmonary rehabilitation:** A personal program of exercise training, breathing techniques, medical education, nutrition advice, and weight management provided by a team of specially trained healthcare professionals.
- **Oxygen therapy:** Supplemental oxygen delivered from a tank at your home or from a unit you can take with you; this can help reduce breathlessness and enable you to be more active.
- **Lung transplant:** Your doctor will discuss this option with you if he or she thinks you may be a candidate.