How IPF Affects the Body

If you think you could be suffering from idiopathic pulmonary fibrosis (IPF)—or even if you’ve already been diagnosed—you may feel confused and overwhelmed. We’re here to help you better understand what IPF is and how it impacts the body. Once you understand the condition, you’ll have the information you need to take an active role in managing it.

While there is no cure for IPF, there is a treatment available that may help to slow the progression of the disease.

Read About a Treatment for IPF

How Does IPF Impact the Lungs?
Healthy vs. IPF Lung

- **Healthy Lung**
  - Bronchioles (tiny airways)
  - Alveoli (or air sacs)

- **Lung with IPF**
  - Areas of fibrosis alternate with areas of normal lung
  - ‘Honeycombing’ - clustered cystic air spaces
  - Irreversibly enlarged, damaged bronchioles (airways) and distorted alveoli (air sacs)
  - Fibrosis between alveoli greatly decreases gas exchange, reducing oxygen transferred to the bloodstream
Let’s explore what actually happens in the lungs of a person with IPF. Normally, oxygen in the air is inhaled through the mouth or nose and travels down the throat and into the lungs.

Once inside the lungs, oxygen passes through tiny air sacs (alveoli) before being released into the bloodstream. At the same time, carbon dioxide is passed from the bloodstream to the lungs to be exhaled. After oxygen is released into the bloodstream, it’s carried to other organs inside the body. These processes are necessary for survival.

In a person with IPF, however, air sac walls thicken with scar tissue, which leads to progressive lung destruction. As the air sac walls thicken, it becomes more difficult to inhale enough air and for oxygen to pass into the bloodstream. Eventually, more air sacs may no longer work. When this happens, organs may not receive the oxygen they need to function properly. This lack of oxygen can lead to symptoms like shortness of breath, a dry, hacking cough, tiredness, and clubbing of the fingertips or toes, meaning they are rounded and widened.

What Happens as IPF Progresses?

Over time, the scarring of the lungs causes them to stiffen, making it difficult for the lungs to expand fully. Breathing becomes difficult when this happens. While IPF will eventually progress for everyone, it’s important to know that it may progress at different speeds for different people. For some, IPF worsens fairly quickly. These people are said to have “rapid progression.” For others, symptoms may stay the same for long periods of time, known as “stable course.” The majority of people with IPF experience “slow progression,” a slow but steady worsening of their disease.

Some people may also experience a serious and sudden worsening of symptoms associated with accelerated disease progression known as acute exacerbations. The cause of these exacerbations in IPF is still unknown and is being investigated.
Acute exacerbations cause a sudden and often permanent worsening of the disease. More than just “having a bad day,” an exacerbation may lead to hospitalization. Almost half of them can be fatal. During an acute exacerbation, you may experience a worsening cough and a fever.

Learn how IPF progresses, and why slowing this progression is so important

Learn About IPF Progression

What Can Be Done to Help Manage IPF?
Although there is no cure for IPF, there may be things that you and your doctor can do to help manage the disease.

- Your doctor may prescribe certain treatments that may help to slow the progression of the disease.
- Your doctor may also recommend oxygen therapy if breathing on your own becomes too difficult.
- Pulmonary rehabilitation may help to improve your quality of life.
- A lung transplant may be an option for some patients. Talk with your doctor about whether or not you could be a candidate.

Although IPF can be an overwhelming disease, we are here to help you understand the condition and possible treatment options. Using the
information here, you can have an informed and meaningful conversation with your doctor about how IPF impacts the body and possible treatment options.