What Should I Expect as IPF Progresses?

The symptoms of idiopathic pulmonary fibrosis (IPF) are not always noticeable or bothersome until the disease has already progressed. As the disease continues to progress, most patients with IPF experience a gradual worsening of lung function. But the course of IPF varies a great deal from one patient to another.

Unlike many other chronic conditions, there are no "stages" of IPF. Everyone experiences it differently. Some patients may only live a few years, while some others may live several years longer. Scientists have yet to determine which factors lead one IPF patient to survive longer than another.

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

Learn About IPF Progression

About Breathlessness

Most people with IPF experience shortness of breath, which doctors call “dyspnea” ("DISP-knee-ah"). Some patients ignore occasional breathing troubles, believing it’s just a sign of old age or being out of shape.
However, as IPF progresses, the damage to the lungs grows more severe, and breathlessness becomes more common. It may occur with minor physical activity—such as getting dressed—or while at rest. With advanced disease, something as simple as brushing your teeth may even lead to breathlessness. Breathlessness has also been reported to lead to other problems, such as trouble with swallowing.

What Are Exacerbations?
People with IPF may experience complications that may cause an increase in symptoms and a decline in lung function. These complications may also lead to an appearance of new lung damage on an HRCT image. (Learn more about tests used to monitor progression of IPF.) When this happens, it is known as an “acute exacerbation” (“ex-zas-sir-BAY-shun”) of IPF. The cause of exacerbations in IPF is still unknown and is being investigated.
What Can Happen During an Exacerbation?

- You may feel your breathing become worse within a short period of time (within 30 days)
- You may experience a worsening cough
- You may develop fever or flu-like symptoms
- You may have trouble breathing on your own, increasing the need for supplemental oxygen

Acute exacerbations of IPF are a very serious concern because they can lead to a rapid decline in lung function. More than just “having a bad day;” an exacerbation can lead to hospitalization. It may ultimately lessen your independence and prevent you from continuing to take part in certain activities. As of now, there are no clear data to suggest that therapies used to treat acute exacerbations of IPF have any benefit while they are occurring.

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]
Who Gets Exacerbations?

It’s important to note that not all people with IPF will experience acute exacerbations. Only about 5-10% of people with IPF have exacerbations in a year. Also, the cause of exacerbations is still unknown.