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 others may live several years longer. Scientists have yet to determine which factors lead one IPF patient to survive longer than another. Research has shown that there may be different “subtypes” of IPF. It’s possible that patients with a certain IPF subtype may actually live longer than other patients with a different IPF subtype, although this is still being investigated.

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 or a decline in lung function. These complications may also lead to an appearance of new patterns 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 more difficult than usual, and this feeling may not get better over time (it may even last up to 30 days)
- You may experience a worsening cough
- You may develop fever or flu-like symptoms
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, although FDA-approved treatment can reduce the chances of them occurring.

Who Gets Exacerbations?

It’s important to note that not all people with IPF will experience acute exacerbations. One study found that only about 10% of people with IPF had exacerbations over a 2-year period. Also, the cause of exacerbations is still unknown. With this in mind, the best approach is to work with your doctor. He or she may help you find the right balance between taking part in those events that matter to you and your family and being appropriately cautious about situations that may pose a risk to someone with a serious lung disease like IPF.