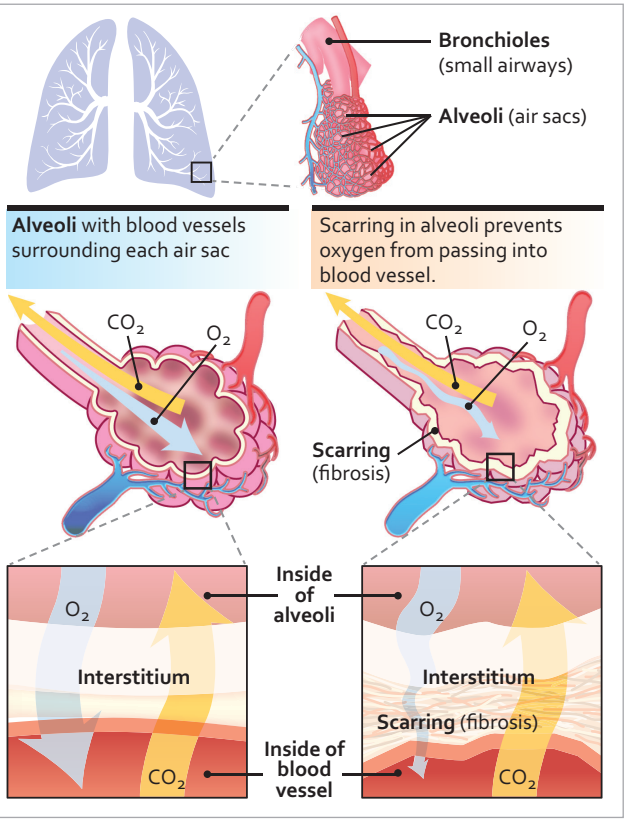
What Is Idiopathic Pulmonary Fibrosis?

IPF Part 1

Pulmonary fibrosis literally means scarring of the lungs. The scarring occurs in the interstitium, which is the lung tissue that provides the scaffolding (support structure) for the alveoli (air sacs). Fibrosis thickens the interstitium, making the air sacs stiff so that they are unable to fully expand and hold as much air as they normally should. This thickening also limits passage of oxygen through the wall of the alveoli into the bloodstream. Over time, fibrosis can worsen to the point that patients may need supplemental oxygen to raise low blood oxygen levels, relieve shortness of breath, and improve exercise ability. This fact sheet will describe IPF and how it is diagnosed—for more information on the Treatment of IPF see Parts 2 and 3 at www.thoracic.org/patients.



Pulmonary fibrosis can happen for many different reasons, including autoimmune disorders, environmental or occupational exposures, as a side effect of certain medications, and a variety of other causes. In many cases, despite extensive evaluation, the cause is unknown; we call such cases **idiopathic**. It is important that you have a full evaluation by an experienced lung specialist to rule out other causes of pulmonary fibrosis before being diagnosed with idiopathic pulmonary fibrosis (or IPF). While the cause of IPF is unknown, it is a form of pulmonary fibrosis, and specific criteria must be met before the diagnosis of IPF is made.

How does IPF develop and who is at risk?

IPF is usually diagnosed in people between the ages of 50 and 80 years. IPF is very uncommon in people under the age of 50 years. Men are at higher risk than women. A past or current history of smoking cigarettes or working in a dusty environment also increases the risk of developing IPF. IPF sometimes runs in families, and there are several genes associated with its development.

Is IPF a rare disease?

IPF is a rare disease, but it is still common enough that you may know someone with this diagnosis. Overall, about 1 out of every 5,000 people has IPF, which equates to about 50,000-100,000 people in the United States, though data from 2015 shows that as many as 200,000 people may be living with the disease. IPF is much more common in older age groups. In people over 65 years of age, up to 1 out of every 200 people have IPF.

What are the signs and symptoms of IPF?

The main symptom of IPF is shortness of breath with activity, which exists in almost everyone as the disease progresses. In fact, in IPF, shortness of breath often limits physical activity. People with IPF may have to slow down, rest and recover throughout an activity or even give up physical activities they once did with ease. Many people with IPF notice they have the most trouble walking quickly, climbing stairs, or going up inclines. About 85% of people with IPF will have a cough. The most frequently-occurring symptoms of IPF (shortness of breath, cough and

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fatigue) are also typical symptoms of other more common diseases; thus, other more common conditions may need to be ruled out before a diagnosis of IPF is made.

By definition, IPF affects only the lungs. If a person has joint stiffness or inflammation, rash, or symptoms coming from outside the lungs, then another condition might be present that requires special testing. In late stages of the disease, IPF can put a strain on the right side of the heart. This is called pulmonary hypertension, which can be associated with swelling in the ankles and worse lung symptoms.

How is IPF diagnosed?

The diagnostic evaluation begins with a thin slice chest CT scan (computerized x-ray imaging). A pulmonologist (lung specialist) with specific expertise in pulmonary fibrosis will review your symptoms, medical history, past medication use, and occupational history. Potential exposures that cause lung scarring in the home or workplace will be discussed. A history of pulmonary fibrosis in other family members is an important clue to the diagnosis. Often your doctor will hear crackles in your lungs when listening with a stethoscope.

Your doctor should also send you for blood tests that can be used to identify blood markers of autoimmune diseases. Currently, there is no single blood test or genetic test to diagnose IPF. Pulmonary function tests (breathing tests) and tests of oxygen levels (e.g., walking tests) are often used to rate the severity of IPF and to look for worsening over time, but these are not helpful in actually diagnosing IPF, as they can be abnormal in other conditions as well. An echocardiogram (ultrasound of the heart) can be used in more advanced IPF to evaluate whether there are any heart complications, such as pulmonary hypertension.

Some people will need additional tests to make a diagnosis, including flexible bronchoscopy or a surgical lung biopsy. Bronchoscopy is a procedure in which a thin flexible tube is passed into the lungs through the mouth or nose usually done when a person is sedated (asleep). Samples can be taken through this tube that can be sent for various tests. (For more information, see ATS Patient Information Series on Flexible bronchoscopy at www.thoracic.org/patients.) A surgical lung biopsy is a more invasive procedure in which two or three small incisions are made in the side of the chest under general anesthesia. During this procedure, a surgeon takes two or three samples of lung tissue that can be looked at under a microscope.

Making a final diagnosis of IPF typically requires a lung specialist who has experience in evaluating and treating patients with pulmonary fibrosis. Patients are also usually discussed in a multidisciplinary conference, in which various clinical features, imaging findings, and biopsy results are reviewed in a group discussion. This discussion most often includes a pulmonologist, radiologist, and pathologist.

Is IPF a serious disease?

IPF is a serious disease. Most people with IPF will have shortness of breath, exercise limitation, and cough as the disease progresses. Many people will require oxygen at some point in their life. On average, IPF shortens a person's life. In previous studies, half of people with IPF live less than 3-5 years, and half live longer than 3-5 years. Some people may have lung transplantation for advanced disease. Over the last decade, advances in the management of IPF have improved the outlook for patients. There are FDA medications approved for IPF and many clinical trials attempting to find new therapies.

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Rx *Action Steps*

- ✔ If you have shortness of breath or cough that does not go away, talk with your doctor.
- ✔ Do not smoke or vape and ask your doctor for support if you need help stopping.
- ✔ If you have been told you may have IPF, ask to see a lung specialist who has experience with its diagnosis and treatment.

Healthcare Provider's Contact Number:

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For More Information

American Thoracic Society
- www.thoracic.org/patients/
  - Flexible bronchoscopy
  - Oxygen therapy
  - Part 2: Medications for Idiopathic Pulmonary Fibrosis
  - Part 3: Nondrug Treatments for Idiopathic Pulmonary Fibrosis

Pulmonary Fibrosis Foundation
- https://www.pulmonaryfibrosis.org/

National Heart Lung and Blood Institute (NHLBI)
- https://www.nhlbi.nih.gov/health-topics/idiopathic-pulmonary-fibrosis

Medications for Idiopathic Pulmonary Fibrosis

IPF Part 2

Idiopathic pulmonary fibrosis (IPF) is a chronic lung disease that gets worse over time. Scarring of the walls of the alveolar sacs (interstitium) causes the lungs to stiffen and reduces the ability of the lungs to absorb oxygen. This leads to shortness of breath that occurs with less and less exercise. While there is no cure for IPF, there are medications that can help with symptoms and slow progression of the disease. Treatment of IPF is important because lung function and symptoms of people with IPF typically worsen over time. There are different kinds of treatments available for people suffering from IPF. This fact sheet discusses medications that are used to help treat IPF. For more information about IPF and other ways to manage and live with it in addition to medication, see parts 1 and 3 at www.thoracic.org/patients.

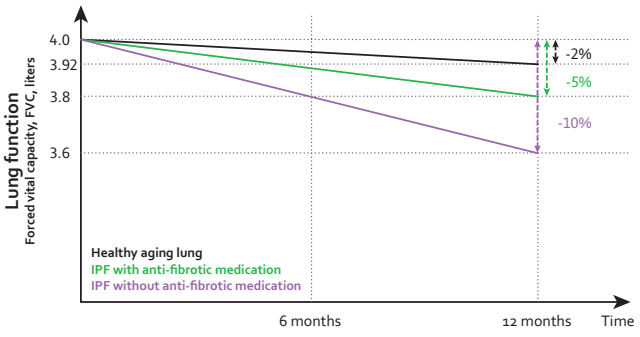
What anti-fibrotic drugs are available for the long-term treatment of IPF?

Pirfenidone and nintedanib are two anti-fibrotic drugs that are used for the long-term treatment of IPF. Anti-fibrotic means that the drugs block scarring. Scar formation in the lungs is complex and involves many different types of cells. By decreasing the formation of scar tissue in IPF, pirfenidone and nintedanib slow the decline in lung function. These are both approved for use in many countries, including the US.

What benefit can I expect from anti-fibrotic drugs?

Both anti-fibrotic drugs slow down the worsening of IPF, but neither are able to cure the disease or reverse existing fibrosis. A healthy person loses around 35 milliliters (ml) of lung function volume per year with increasing age. Those affected by IPF who are not treated with an anti-fibrotic drug lose, on average, around 200-400 milliliters (ml) of their vital capacity (lung function) per year. This is a loss of up to 10% of the total lung function volume each year. People who are treated with an anti-fibrotic drug lose, on average, around 100-200 ml of their vital capacity per year, or about half of the loss that is seen in people not taking an antifibrotic drug. This slowing of lung function decline in people treated with pirfenidone or nintedanib has been confirmed in several large clinical research studies that include more than 2800 people with IPF. However, it is important to note that not everyone with IPF experiences benefit from these medications and that both medications can have associated side effects. (Figure 1)

Figure 1. *Decline in lung function in healthy adults (black, nonsmokers), in patients with IPF treated with antifibrotic medication (green), and in patients with IPF not treated with anti-fibrotic medication (purple).*



Anti-fibrotic therapy might reduce shortness of breath over time, and some people may have a decrease in cough severity, but these benefits have not been seen consistently in all people taking part in the research studies. Some, but not all, studies have suggested that people treated with anti-fibrotic medications may need to be admitted to the hospital less often. Further, people on these medications may live longer than without these medications. Taking both drugs together is not yet well studied; it is unknown if there would be additional benefit or harm to this approach.

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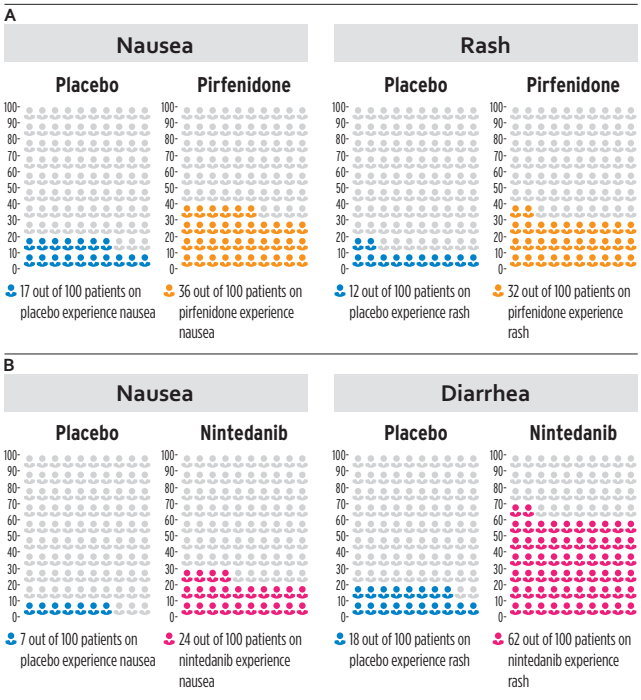
How do I take pirfenidone (brand name Esbriet®) and what side effects can occur?

People with IPF who are prescribed pirfenidone will typically take tablets or capsules three times daily with food. You will usually be started on one 267 mg tablet or capsule taken three times per day, and will be instructed to gradually increase the dose to 801 mg three times per day. During this period, your lung specialist will ask you about side effects that might occur and check for abnormal liver blood tests. Blood tests are usually performed less often after the first few months in those who are tolerating a stable dose without significant side effects. Side effects of pirfenidone that occur in more than 10% of people include nausea, acid reflux (heartburn), vomiting, decreased appetite, weight loss, headache, dizziness, fatigue, and skin problems such as a "photosensitivity" rash that develops in reaction to sun exposure (Figure 2). Overall, side effects from pirfenidone occur frequently, but usually these can be controlled sufficiently so that most patients can continue treatment (Figure 3). Talk to your healthcare provider about possible side effects and concerns you may have about your medication.

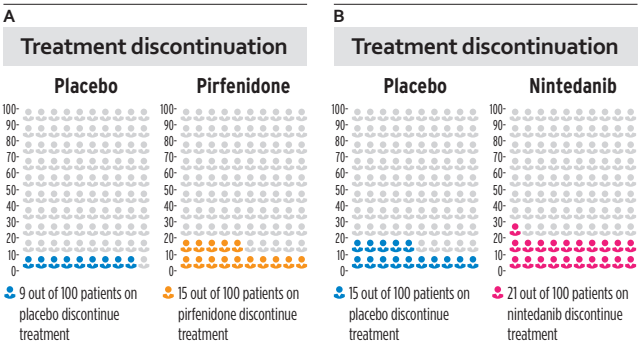
Figure 2. *Most common side effects from pirfenidone (A) and nintedanib (B).*



How do I take nintedanib (brand name Ofev®) and what side effects can occur?

People with IPF who are prescribed nintedanib will typically take one 150 mg tablet twice daily with food. The dose can be reduced to 100 mg tablets if side effects occur. The most common side effect of nintedanib is diarrhea, which occurs in about two thirds of patients. Additional side effects can include decreased appetite, weight loss, nausea, vomiting, abdominal pain, and abnormal liver blood tests (Figure 2). Your doctor will monitor your liver using simple blood tests that need to be repeated more frequently in the first few months of treatment and typically monthly thereafter. Overall, side effects from nintedanib occur often, but usually these can be controlled well enough that most can continue treatment (Figure 3). Talk to your IPF healthcare team about possible side effects and concerns you may have about your medication.

Figure 3. *Number of patients that discontinue treatment due to side effects from pirfenidone (A) or nintedanib (B) in clinical trials.*



How can I manage common side effects of pirfenidone and nintedanib?

Nausea, diarrhea, or loss of appetite occur in up to 50% of people with IPF taking anti-fibrotic drugs. Therefore, management of these side effects with medications or a dietary change is often needed. Your IPF healthcare team or a dietician can provide advice about how to manage these symptoms. This is particularly important for any weight loss, either as a consequence of your disease or from side effects of the anti-fibrotic medication. For patients taking pirfenidone, it is essential to wear sun-protective clothing (e.g., long sleeves, wide-brimmed hat) and sunblock to prevent a bad sunburn.

These medicines can interact with other medications or supplements. It is also important to check with your physician or pharmacist before starting any new medications or supplements to make sure these are safe to take along with your anti-fibrotic drug.

How do I know if the anti-fibrotic drug is working?

Despite using these medications, your lung function is still likely to get worse over time, and your shortness of breath will likely increase. The goal of anti-fibrotic treatment is to slow this progression, and preserve your lung function as much as possible. As a result, your

physical performance and quality of life will likely be better in the future.

How long do I have to take the anti-fibrotic drug?

Anti-fibrotic medications can be continued for as long as they are considered effective. The treatment can also be changed from pirfenidone to nintedanib, or vice versa, if severe side effects occur, or if one drug is considered to be ineffective. Decisions about whether to change or discontinue anti-fibrotic medications should be made along with your lung specialist and healthcare team.

What happens if I don't want to be treated with an anti-fibrotic drug?

You might decide not to take any medications for IPF. This may be because you suffer from other severe diseases, or you have concerns about possible side effects or have had significant side effects that appear to outweigh potential benefit from the drugs. Sometimes your lung specialist will advise you not to use anti-fibrotic treatment. Regardless of whether you take an anti-fibrotic medication or not, it is important to remember that there are other strategies that can help manage your disease. Other ways to help manage IPF are covered in Part 3 found at www.thoracic.org/patients.

Which drugs should I not take for the treatment of IPF?

Currently, only pirfenidone and nintedanib are approved for the long-term treatment of IPF. Although you can find suggestions for a variety of other treatments on the Internet, these lack proven benefit and are not recommended at this time. We advise that you discuss your questions related to such alternative treatments with your lung specialist.

What drugs are used to treat acute exacerbations of IPF?

Different medications are sometimes used for a short period of time if patients experience a sudden worsening of their symptoms. This situation can occur with a variety of predisposing factors, with a resulting increase in the amount of inflammation in the lungs. These short-term medications most often include antibiotics against lung infections and corticosteroids for the treatment of inflammation.

Are there medications that relieve the symptoms of IPF?

Medications that can relieve symptoms include opiate medications such as morphine to treat severe shortness of breath, cough suppressants, and medications against anxiety. Although these can help reduce symptoms, these do not affect the underlying fibrosis itself.

Will there be new drugs available soon?

For many decades, studies of potential medications for IPF showed no benefit, and some of these treatments were even found to be harmful. Pirfenidone and nintedanib are the first and only drugs to date that have been shown to reliably slow the progression of lung fibrosis. There is no medication to cure IPF, but research is ongoing to find more effective and better tolerated treatments. It is therefore still important that we continue to research new ways for the treatment of IPF. Taking part in and supporting clinical trials might help you or other people affected by IPF in the future. You can find ongoing clinical trials listed on the Pulmonary Fibrosis Foundation and the clinicaltrials.gov webpages below. Please talk to your lung specialist to get more information on new clinical trials that are testing potential treatments of IPF.

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Rx *Action Steps*

- ✔ If you have been diagnosed with IPF ask your lung specialist about antifibrotic drugs.
- ✔ Do not take any medicines for IPF without discussing them with your lung specialist.
- ✔ Take medicines regularly as prescribed and talk to your IPF healthcare team about any concerns you have about the medicines including side effects.
- ✔ Talk about other non-drug therapies you can do for IPF.

Healthcare Provider's Contact Number:

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For More Information

American Thoracic Society

- www.thoracic.org/patients/
  - Flexible bronchoscopy
  - Oxygen therapy
  - Part 1: What is Idiopathic Pulmonary Fibrosis?
  - Part 3: Nondrug Treatments for Idiopathic Pulmonary Fibrosis
- https://trials.pulmonaryfibrosis.org/
- http://clinicaltrials.gov/

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Nondrug Treatments for Idiopathic Pulmonary Fibrosis

IPF Part 3

IPF is a rare lung disease that causes shortness of breath and low oxygen levels because of lung scarring. There is no cure, but there are several medications used to treat IPF that help slow the worsening of the disease or reduce symptoms. There are also non-drug strategies that play an important role in IPF. This fact sheet discusses the non-drug strategies you can do to help manage IPF in addition to medications. For more information about IPF and its treatment, see parts 1 and 2 at www.thoracic.org/patients.

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Do I need oxygen therapy?

Scar tissue that builds up in the lungs leads to thickening of the walls of the air sacs. Oxygen from the air we breathe must move across this scar into the blood vessels of the body. When you are active, your body needs more oxygen to fuel your muscles. If the lungs are not able to meet those needs, the oxygen levels in your blood might drop. Because low oxygen levels can cause shortness of breath, oxygen therapy can relieve shortness of breath by increasing blood oxygen levels.

People with pulmonary fibrosis often require oxygen with activity before they require oxygen at rest. Your oxygen needs may change over time, so your oxygen levels should be evaluated regularly with an oximeter to find out how much oxygen you need at rest, with activity, and during sleep. Please talk to your healthcare provider about what oxygen saturation goals are best for you. There are multiple kinds of oxygen delivery systems such as oxygen concentrators and liquid oxygen. Some devices are designed for home use while others are designed for outdoor activities and travel. (See also the ATS Patient Information Series fact sheet Oxygen Therapy.)

Should I exercise and what is pulmonary rehabilitation?

Yes, you should exercise! Regular exercise helps you improve your stamina and keep in shape so that you are able to stay active and do things for yourself. People with IPF can have trouble with physical activity because of shortness of breath, muscle weakness, low oxygen levels, and lack of fitness.

Finding exercise you enjoy may be hard at first, but you can get help to build up your exercise level and tolerance over time.

Pulmonary rehabilitation is a safe way to exercise under supervision. The staff at these programs will prepare an exercise regimen that fits your abilities and needs. They will monitor your exercise tolerance and oxygen requirements, and gradually increase the intensity of exercise as you get fitter and your muscles get stronger. Joining a pulmonary rehabilitation program can help you improve your energy level, reduce your shortness of breath, give you a better understanding of your IPF and oxygen use, and teach you self-management skills.

Pulmonary rehabilitation is offered at certified inpatient and outpatient centers. Please ask your healthcare team if pulmonary rehabilitation is right for you.

What are patient support groups?

Support groups offer patients and their caregivers the opportunity to receive additional education and support outside of the office visit. People who take part in support groups connect with other patients and caregivers who are going through the same changes in their life and facing similar challenges. You can learn how to cope and adapt to your life with pulmonary fibrosis, get valuable information about the disease and its impacts, and benefit from practical and emotional support. Pulmonary fibrosis support groups are available in many places and there are also some you are able to connect to on-line.

Could I benefit from palliative care?

Palliative care is a medical and nursing specialty focusing on maximizing quality of life during all stages of a serious illness. The goal is to provide relief from the symptoms and stresses of a serious illness like pulmonary fibrosis, and to improve quality of life for both you and your family. Palliative care may be appropriate at any stage of IPF and can be combined with other treatments that focus on slowing the disease

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progression, such as anti-fibrotic medications. Palliative care is based on patient and family needs, and not on prognosis.

End-of-life care (which is a part of palliative care) can help improve quality of life for people with pulmonary fibrosis by addressing physical, psychological, and spiritual distress at the end of life. Sometimes end-of-life care is delivered via a hospice service.

Please ask your healthcare team about whether palliative care approaches might be right for you. (See also the ATS Patient Information Series fact sheet on Palliative Care.)

Is lung transplantation an option for me?

Because IPF is an incurable disease that worsens over time, lung transplantation may be a treatment option for some patients. The diseased lungs are surgically removed and replaced by healthy lungs from a person who has died and donated his or her lungs. Lung transplantation is only an option for a few patients, and it is a big operation with major risks. Availability of donor lungs may also be limited. People who have had lung transplant often face short- and long-term problems. Medications to suppress the immune system are necessary to prevent the body from rejecting the new lung(s), and these medications also make patients more likely to get a severe infection. While lung transplant can be lifesaving, it is only an option for patients with very severe lung disease, otherwise healthy, and pass an extensive work-up. (For more information on lung transplantation, see www.thoracic.org/patients.)

What else can I do to manage my IPF?

Because you have a chronic lung disease, you may be more likely to become seriously ill from infections such as influenza (flu), pneumonia, COVID-19 or even a simple cold. You should try to avoid exposure to ill persons as possible and wash your hands often. You can help protect yourself from some infections by getting vaccinated. Ask your healthcare provider about what vaccines may be right for you and talk about any questions or concerns you may have about getting vaccinated.

A small percentage of people with IPF develop rapid worsening or breathlessness over a few weeks, which is called an acute exacerbation. If you experience such worsening, you should contact your healthcare provider or seek immediate medical attention.

Other things you can try to help with your shortness of breath include use of hand fans, mindfulness meditation, and yoga. Remedies that might soothe your cough include hot tea with honey and lemon, lozenges, or cough drops. (See the ATS Patient Information Series fact sheet on Mindfulness)

Action Steps

- ✔ Try to stay active and get regular exercise, even if you need to use oxygen.
- ✔ Consider going to pulmonary rehabilitation to get help with a safe exercise program.
- ✔ Get recommended vaccines such as flu and COVID, use good handwashing, and avoid ill contacts as possible.
- ✔ Avoid smoking or vaping exposure.
- ✔ If you have IPF, consider joining a support group.

Healthcare Provider's Contact Number:

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For More Information

American Thoracic Society

- www.thoracic.org/patients/
  - Oxygen therapy
  - Exercise and Lung Disease
  - Pulmonary Rehabilitation
  - Palliative Care
  - Mindfulness
  - Lung Transplantation (series)
  - Part 1: What is IPF
  - Part 2: Medications for Idiopathic Pulmonary Fibrosis
  - www.livebetter.org (pulmonary rehab resource)

US National Heart Lung and Blood Institute

- https://www.nhlbi.nih.gov/health-topics/idiopathic-pulmonary-fibrosis

United States United Network for Organ Sharing (Lung transplantation)

- https://unos.org/

Patient support groups:

- https://www.pulmonaryfibrosis.org/life-with-pf/support-groups
- https://www.blf.org.uk/support-for-you/pulmonary-fibrosis/support-groups
- https://www.actionpulmonaryfibrosis.org/find-a-support-group/
- https://www.eu-ipff.org/about-us#support
- https://bc.lung.ca/how-we-can-help/patient-support-groups/pulmonary-fibrosis-support-groups