

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.



Non-drug strategies, also known as nonpharmacological therapies, play an important role in the treatment of IPF. These strategies include symptom management, oxygen therapy, and pulmonary rehabilitation. Peer coaching through support groups and palliative care experts are usually helpful. For some individuals lung transplantation is appropriate.

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 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 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 to find out more 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 few patients, because it is a big operation with major risks. Availability of donor lungs may also be limited. Lung transplant recipients frequently 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, who are relatively young, 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 should try to avoid situations where you can get an infection, even a simple cold or the flu can be very serious. You should get a flu vaccine every year. Check with your doctor about getting a pneumococcal pneumonia vaccine if you have not had it. Living with IPF can be particularly challenging during a pandemic; for more information on COVID-19 see www.thoracic.org/patients.

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 strategies that are possibly effective for 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)

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Rx 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.
- ✓ Avoid smoking or vaping exposure.
- ✓ If you have IPF, consider joining a support group.

Healthcare Provider's Contact Number:

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

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

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