

Understanding IDIOPATHIC PULMONARY FIBROSIS (IPF)



An educational health series from





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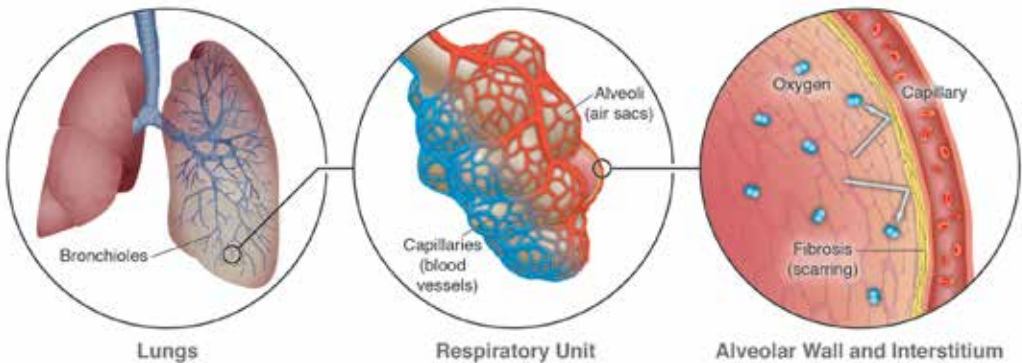
What is Idiopathic Pulmonary Fibrosis or IPF?

Interstitial lung disease (ILD) is a broad category of lung diseases that includes more than 200 disorders that can be characterized by fibrosis (scarring) and/or inflammation of the lungs.

Despite an exhaustive evaluation, in many people the cause of ILD remains unknown. In these cases, the ILD is considered “idiopathic” (meaning of unknown cause).

Idiopathic pulmonary fibrosis is a term used to refer to one particular form of pulmonary fibrosis of unknown cause. Idiopathic pulmonary fibrosis is commonly called IPF.

Let’s take a look inside the lungs to see what is happening with IPF.



In IPF, scarring or fibrosis occurs in the interstitium. The interstitium of the lung refers to the microscopic area within the walls of the alveoli (air sacs). Like the leaves on a tree, the alveoli arise from the tiniest bronchioles (airways). There are hundreds of millions of alveoli in a lung: 150 alveoli would fit in a cubic millimeter. Each alveolus (individual air sac) is surrounded by a network of tiny blood vessels (capillaries) — like mesh encircling the alveolus. The air sac and blood vessels together are called a respiratory unit. This is where the exchange of oxygen and carbon dioxide take place. In IPF, scarring or fibrosis disrupts the interstitium; the air sac wall thickens; oxygen cannot pass into the bloodstream as it normally should; and the lungs get stiff and difficult to expand.

What are Symptoms of IPF?

The most common symptom of IPF is shortness of breath, also known as dyspnea. Many people describe this as a feeling of breathlessness. The earliest symptom of IPF is shortness of breath with exertion or strenuous activity. Many people ignore this symptom or ascribe it to being old or out of shape and simply reduce the amount of physical activity they do. For example, earlier in the course of IPF, a person may notice shortness of breath only when running or hurrying to catch a bus. Later in the course of IPF, he or she may notice shortness of breath when casually walking across the street.

IPF is isolated to the lung and does not cause eye or ear problems, chest pain, swelling or pain in the joints or skin rash.

Other common symptoms include a dry, hacking cough and fatigue. Symptoms of IPF may develop and worsen gradually or, less commonly, they may also worsen rapidly. Instances of acute IPF worsening occur in about 10 percent of people.



The most common symptom of IPF is shortness of breath.

How is IPF Diagnosed?

The first step in diagnosing IPF is a thorough evaluation. Idiopathic pulmonary fibrosis can be diagnosed only after ruling out other lung diseases. When the diagnosis of IPF is suspected, your doctor may have you do a number of tests.

These may include:

- A complete medical history
- A complete physical examination
- Chest imaging, including a chest X-ray and high resolution computed tomography (HRCT) scan
- Pulmonary function tests
- Arterial blood gas
- Exercise test and/or an assessment of blood oxygen levels during exertion
- Blood tests looking for hints at the cause of the lung disease
- Bronchoscopy with bronchoalveolar lavage
- Surgical lung biopsy

Your doctor may order other tests based on your history and physical exam.

You and your health care provider may choose to have you seen by a specialist, such as a pulmonologist (lung specialist) to confirm a diagnosis and treat your IPF once it's diagnosed.

National Jewish Health is a Specialized Center of Research for ILD as designated by the National Institutes of Health. Members of our team work together in an attempt to better understand the causes of ILD (including IPF) and to develop and test new therapies. Doing so ensures that you have access to the latest, groundbreaking research, to participation in ongoing clinical trials and to the most effective therapies available for IPF.



What are Your Goals?

Put a check (✓) in the boxes with your goals

- Decreasing shortness of breath
 - Read more about medication, breathing techniques and exercise for ideas on decreasing and controlling shortness of breath.
- Reducing exacerbations
 - Read more about avoiding infections and symptoms for ideas on how to try to reduce exacerbations.
- Becoming more active
 - Read more about breathing techniques, healthy lifestyle and exercise for ideas on becoming more active.
- Decreasing anxiety or stress
 - Read more about breathing techniques and living with IPF for ideas on decreasing anxiety and stress.
- Improving mood
 - Read more about living with IPF and exercise for ideas on improving mood.
- Learning more about IPF

Think of other goals you may have.

Write any other goals in the space below:

Talk with your health care provider about your goals.



Talk with your healthcare provider about your goals.



Living A Full Life with IPF

How is IPF Managed?

People with IPF can become actively involved in the management of their disease. Participation in the treatment plan will help people with IPF, their caregivers and family members achieve the best possible results.

IPF Management Includes:

- Partnering with your health care provider to formulate a comprehensive treatment plan
- Learning more about IPF
- Living a full life that includes adopting a healthy lifestyle involving
 - Regular exercise
 - Healthy eating
 - Rest
 - Support from others
- Giving up smoking (cigarettes, cigars, pipes, marijuana)
- Avoiding infections
- Taking medication as prescribed
- Using oxygen therapy when prescribed
- Using breathing techniques
- Determining whether lung transplantation is an option for you
- Following an Action Plan
 - Everyday Action Plan
 - Action Plan for exacerbations
- Learning about palliative care
 - Needs of care, physical, emotional, social, intellectual, spiritual
 - Community resources
 - Advanced care needs

Now that you have an overview of IPF management, let's talk about each topic in more detail.

Remember your goals.



People with IPF can become actively involved in the management of their disease.



Healthy Lifestyle

A healthy lifestyle is important for everyone, including people with IPF and their caregivers.

A healthy lifestyle includes:

- Regular exercise
- Giving up smoking
- Healthy eating
- Support from others

Exercise

An exercise program is perhaps the most important aspect in the treatment of IPF. It is common for people with lung disease to limit physical activities, because they are afraid of becoming short of breath. But this is the wrong approach. Regular exercise can improve your heart, lungs and muscles, and it can help you breathe easier and feel better. Many people with IPF enjoy walking, water aerobics and riding a stationary bike. Some people with IPF need oxygen therapy while exercising. Portable oxygen units are available. Talk with your health care provider about recommending an exercise program for you.

Getting Started

Before starting an exercise program, always talk to your doctor about your plan to be sure that it is safe. Then, start with slow, small steps rather than attempting a quick “lifestyle overhaul.” Changing lifelong patterns is a gradual process. Succeeding in many little steps can be encouraging, while failing at a quick major change is definitely discouraging. Pulmonary rehabilitation provides an excellent way to jump start an exercise program. Talk to your provider about whether pulmonary rehabilitation is right for you and if there is a program nearby.

People with IPF can benefit from a formal rehabilitation program. Most pulmonary rehabilitation programs will include medical and nursing management, exercise, breathing retraining, education, emotional support and nutrition counseling.

A successful pulmonary rehabilitation program (such as the program offered by National Jewish Health) addresses the needs of each person and tailors the treatment to meet those needs.



Regular exercise helps you improve your overall fitness by improving your heart and muscle function.

Exercise Tips

- While exercising, remember to inhale (breathe in) before starting the exercise.
- Exhale (breathe out) through the most difficult part of the exercise.
- Purse your lips while breathing.
- Don't hold your breath while exercising.
- Count out loud as you do the exercises.

Giving Up Smoking

Giving up smoking is a critical step in controlling your disease and preventing further lung damage.



Giving up smoking is important to help control your disease and prevent further damage to your lungs.

Steps to help you quit smoking

- Make a firm commitment to quit. Start thinking of yourself as someone who doesn't smoke.
- Talk to your health care provider about quitting and a plan to help you quit. Your provider can discuss the use of medicines to help you give up smoking.
- Consider joining in a program that provides guidance and support for learning to live without cigarettes. Every state offers free telephone counseling through the Quitline. **The Quitline number is 1-800-QUIT-NOW, or 1-800-784-8669.** In addition, there are many online and face to face group quit smoking programs.

Once you stop smoking it is important to avoid being around smoke. This will help decrease irritation to your lungs. Also, a smoke free environment is healthier for everyone.

- Talk with family members about avoiding tobacco smoke.
- Encourage family members and caregivers who smoke to quit. If they are not ready to quit, ask them not to smoke in your home or car.

Healthy Eating

It is unknown whether a specific diet is beneficial for people with IPF, but eating a balanced diet with plenty of vegetables, fruits and whole grains and maintaining a healthy weight are important parts of managing IPF. While it can be difficult to focus on this aspect of your life, eating well plays a big role in feeling good and staying healthy.

If you are underweight, focus on gaining weight. The average person requires approximately 50 calories per day for breathing. People with IPF often expend more calories per day on breathing alone. This increases daily calorie requirements making it difficult to maintain weight.

If you are overweight, focus on losing weight. There are many benefits of weight loss, including reducing shortness of breath by decreasing pressure on the diaphragm and respiratory muscles. A good weight loss program should include a variety of foods and physical activity.

Rest

Rest is important. Make sure you get enough sleep each night. Some people with IPF need a nap or dedicated rest time during the day. If you are having trouble sleeping, talk with your health care provider.

Support From Others

Living with IPF is a unique and special challenge that you and your family must deal with on a daily basis. But the more you know about IPF, the better suited you are to manage the various aspects of your disease. As you take control, your quality of life will improve.

Support groups, both in-person and online, can provide an important social outlet for people with IPF and their caregivers. The meetings often provide education, discussions about a variety of health topics and social outings.





Treatment of IPF

A healthy lifestyle is important for people with IPF. In addition, IPF management includes:

- Avoiding infections
- Medications
- Oxygen therapy
- Breathing retraining
- Lung transplant

Avoiding Infections

People with IPF are at increased risk of lung infections. There are a number of measures that can help avoid infections.

- Vaccines are recommended. The influenza or flu vaccine is recommended yearly, often in the fall. There are two pneumonia vaccines; both are recommended. These vaccines help prevent severe infection from bacteria that is a common cause of pneumonia.
- Good hand washing with soap and water is also very important to help prevent the spread of germs and infections. Alcohol-based gels are also effective. Use a quarter-sized dollop in your palm, and rub your hands together until they are dry.
- Avoid touching your eyes, nose and mouth. Germs are often spread this way.
- Avoid contact with others when they are sick.





Develop a routine to remember your medications.

Medication Therapy

There are two medications that have been approved by the Food and Drug Administration (FDA) for treatment of IPF: nintedanib and pirfenidone. Both are considered anti-fibrotic drugs, meaning that they can help reduce new lung scarring. These drugs are not a cure, and they do not stop progression of IPF. However, they have been shown to slow its progression.

Pirfenidone (Esbriet®)

Pirfenidone is an oral medication taken three times daily. Pirfenidone slows the progression of disease for some people with IPF.

Some side effects include:

Nausea, loss of appetite, stomach upset and photosensitivity (a tendency to easily develop sunburn or rash when exposed to the sun)

Nintedanib (OFEV®)

Nintedanib is an oral medication taken twice daily. It is a triple kinase inhibitor that blocks several pathways that lead to the development of fibrosis. Like pirfenidone, nintedanib slows the progression of disease for some people with IPF.

Some side effects include:

Diarrhea, and less often, nausea and vomiting.

Should I take an anti-fibrotic for IPF? And how am I monitored if I do?

The decision to treat people with IPF involves a careful weighing of the potential risks and benefits of therapy. The potential benefits often outweigh the risk from the medication side effects. Neither medication is recommended over the other, but one may work best for you. Your doctor will work with you to prescribe treatment tailored to your individual needs.

Because of the potential side effects of anti-fibrotic medications, your doctor will carefully monitor you while on therapy. This will include routine blood work to assess your liver.

Oxygen Therapy

People with IPF need oxygen therapy as the disease progresses. Oxygen therapy is used to normalize the oxygen level in blood during sleep, rest, activity and during acute illnesses in the hospital.

It is sometimes difficult for you to know when oxygen therapy is required. If the oxygen level is low in your blood, you may experience symptoms like shortness of breath, irritability, morning headaches or ankle swelling. If you notice any of these symptoms, see your health care provider. He or she can review your overall medical condition and decide what treatment you need. Your oxygen can also be low without any symptoms. Just because you feel well doesn't necessarily mean you don't need oxygen.

Testing Your Oxygen Level

There are two methods to test the oxygen level in the blood: oximetry and arterial blood gases. One or both methods may be used to determine your need for oxygen therapy.

Your oxygen level can be measured as a percentage of your blood's oxygen carrying capacity. This is called oxygen saturation. Oxygen therapy may be necessary if your saturation is below 89 percent. To measure oxygen saturation in your blood, a small clip is placed on the finger or earlobe. This is a simple, convenient, painless way to determine your need for oxygen. This test may be done at rest, during sleep and while you are walking.

The second method of testing is the arterial blood gas. While this blood test is more complex, the results can give your health care provider more information about how your lungs are working. For this test, blood is drawn from an artery in your wrist, and both oxygen and carbon dioxide levels are measured.

When the test determines that oxygen is needed, your health care provider will write a prescription for oxygen. The prescription will tell you how much oxygen to use and when to use it. You will feel better using oxygen.

Once oxygen is prescribed, many people use a portable pulse oximeter to measure oxygen saturations when exercising, when resting or with changes in altitude.



Oxygen therapy is used to ensure there is enough oxygen in the blood to provide for the body's needs.



Talk with your health care provider if you have safety concerns with oxygen.

There are three systems which can supply oxygen:

- Concentrators
- Compressed gas
- Liquid

Each system has portable systems available when outside the home. Each system also has advantages and disadvantages. Your health care provider can help you select the system that is best for you. The durable medical equipment (DME) company will provide the oxygen and provide an explanation and demonstration of whichever system you choose, working with your health care provider. It is important to choose the system which best fits your lifestyle.

Concerns With Oxygen

You may be concerned about how oxygen therapy may change your lifestyle, how oxygen affects your body and whether oxygen therapy is safe. You may worry that oxygen treatment will prevent you from leaving your home. However, many convenient portable systems are available. In fact, oxygen therapy allows you to be more active by providing the oxygen that your body needs. Oxygen therapy does not cause any harm to your lungs or your body, if used as prescribed. You will not develop an addiction to oxygen. Oxygen therapy is very safe. The only thing you need to remember about safety is that oxygen is flammable; keep your face and your oxygen away from flames. Talk with your health care provider if you have specific safety concerns.

Breathing Techniques

Learning new breathing techniques will help you move air in and out of your lungs. It is helpful to use effective breathing techniques with exercise to minimize shortness of breath and assure adequate oxygen to your working muscles. Breathing retraining has the added benefit of helping you relax when you are anxious or stressed. Two types of breathing techniques are pursed lip breathing and coordinated breathing with exercise.

Pursed Lip Breathing

The purpose of pursed lip breathing is to help keep your airways open. This helps your airways to remain open. Pursed lip breathing also slows down your breathing rate and calms you down.

Here are the steps for pursed lip breathing:

- Inhale slowly through your nose with your mouth closed; try to take in a normal amount of air.
- Exhale slowly through your mouth with your lips in the whistling or kissing position.
- Breathe out for twice as long as you breathe in.

Do not take in a large deep breath. Never try to force out the air.

Coordinated Breathing

The purpose of coordinated breathing is to help assure adequate oxygen to your working muscles and to prevent you from holding your breath.

Here are the steps for coordinated breathing.

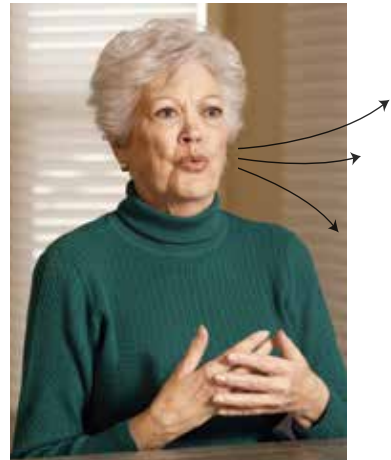
- Inhale through your nose before starting the exercise or activity.
- Exhale through pursed lips, during the most exerting part of the exercise or activity.

If coordinating your breathing with exercise is difficult, as you perform the movement, count out loud. This helps prevent you from holding your breath. If you become very short of breath, stop the exercise, use pursed lip breathing to help control your breathing, then start exercising again.

Practice these breathing techniques daily. When you feel short of breath, anxious or just wound up, use these breathing techniques.



Pursed lip breathing inhale



Pursed lip breathing exhale



Think of whether lung transplant is an option for you personally, and share this information with your pulmonologist.

Lung Transplant: Is It Right For You?

IPF may progress despite therapy. If this happens, lung transplantation may be an option for you. Lung transplant surgery replaces one or two diseased lungs with healthy lungs from a non-living donor. With improved surgical techniques and post-transplant care, this may offer you improved quality of life and longer survival. IPF doesn't recur in transplanted lungs.

Lung transplantation is only performed at specialty medical centers. Your health care team may determine that lung transplant is the best option for you and that you are healthy enough for surgery. After an extensive evaluation, appropriate candidates are placed on a waiting list. A position on the waiting list is determined by disease severity. Wait times vary from transplant center to transplant center.

After a lung transplant, people need to take anti-rejection medicines for the rest of their lives to help prevent the body from rejecting the new lungs. These medicines can cause complications, including infection. Having emotional support is an important part of recovering and staying healthy after lung transplant surgery.



An Action Plan for IPF

Sometimes it is hard to know what to do when your symptoms change or worsen. Do you call your doctor? Go to the emergency room? Your health care provider can tell you what you can do when this happens. This is an Action Plan. Knowing when your symptoms are getting worse can help you and your health care provider decide what you should do in your home, at your health care provider's office or in the emergency room.

A change or increase in the symptoms you usually have may be the only early warning sign.

When your symptoms are getting worse, you may notice one or more of the following:

- An increase in shortness of breath or coughing
- A general feeling of ill health
- Feeling of lack of energy or fatigue
- Fever.

Symptoms do not go away when they are ignored. Therefore, call your health care provider if you have new or worsening symptoms lasting longer than a day.

Talk with your caregiver and family members about symptoms and your Action Plan.

Advance Directive

As part of the action plan, talk with your health care provider, caregiver and family about treatment if you become seriously ill and unable to express your wishes. A written, legal document expressing your wishes is called an Advance Directive. This may include what medical decisions are made and who will make them if you are unable to respond or make your own decisions.

Palliative Care

Palliative care is an approach to care that focuses on the physical, emotional, social, intellectual and spiritual needs of people who have a serious illness. Palliative care is not the same as end-of-life care. Palliative care focuses on improving or maintaining quality of life through symptom relief; assessing tolerance for curative treatment; and easing transitions when curative treatment is no longer an option. Palliative care incorporates the hopes and values of people with IPF and their family members.



For People with IPF and Caregivers: Living a Full Life at Any Stage of IPF

There is no formal staging system for IPF. However, thinking of IPF in four stages can help people with IPF and their caregivers prioritize their needs and keep them from feeling overwhelmed. Although education regarding a variety of topics can occur at any time, the stages may help people with IPF and their caregivers focus on what is needed and what to expect as time goes by.



The Four Stages of IPF

- **Stage 1:** Recently diagnosed
- **Stage 2:** Needing oxygen with activity, but not at rest
- **Stage 3:** Needing oxygen 24 hours a day, with activity, at rest and during sleep
- **Stage 4:** Advanced oxygen needs (needing high-flow oxygen or when a lightweight, portable delivery system is unable to meet a oxygen needs).

Stage 1: Recently Diagnosed

What should I focus on when I am recently diagnosed?

When you are diagnosed with IPF you may want to:

- Partner with your pulmonologist to develop and individualize your Action Plan.
- Learn about IPF, including the symptoms, treatment and prognosis.
- Live a full life with IPF through:
 - Regular exercise. Stay active. Physical exercise is the most important thing you can do for yourself.
 - Healthy eating
 - Rest
 - Support from others
- Consider joining a support group in your area.
- If you smoke, work with your doctor on quitting smoking.
- Learn what you can do to avoid infections.
- Begin thinking and talking about Advance Directives.
- Focus on today — on what you can do, not what you can't.
- Live life one day at a time.

Stage 1: How Can the Caregiver Help?

- Empower yourself by learning about IPF with your partner.
- Attend health care appointments to be a second set of ears.
- Discuss and write down questions to ask the health care provider.
- Help keep a record of care.
- Help keep track of medications.
- Encourage the person with IPF to live a full life including regular exercise, healthy eating, rest and support from others.
- Keep a good supply of waterless soap around the house and in a to-go bag.
- Consider attending a support group, either alone or with your loved one.
- Build your own support network consisting of family, friends and/or other caregivers.
- Remember to take care of yourself. Having a loved one with a serious illness can lead people to forget about themselves and their own health. You will not be a good caregiver if you are physically ill or emotionally unwell. Spend some time doing things you enjoy to avoid getting burned out.

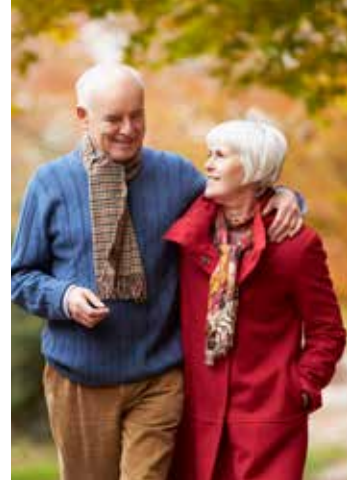


Empower yourself by learning about IPF with your partner.

Stage 1: How Might a Caregiver Be Affected?

You may experience a range of emotions when your loved one is diagnosed; these could include denial, fear, stress, anger/frustration, grief or depression. It is important to know that your loved one may not experience the same emotions as you, or they may not experience emotions at the same time.

- **Denial:** It may be hard to believe or accept that your loved one has IPF. Experiencing denial for a short period of time is natural; however, longer-term denial is unhealthy and may affect your loved one and delay the initiation of appropriate therapy.
- **Fear:** Fear of the unknown is common. Not knowing how IPF will behave over time is unsettling. Try to focus on the here and now, rather than the what-ifs.
- **Stress:** Not knowing how to help your loved one with IPF can lead to stress. Try to find activities to relieve your stress. Planning for the future can help alleviate stress.
- **Anger/frustration:** It is normal to feel anger and/or frustration at IPF when a loved one is diagnosed. Try to work through it.
- **Grief/depression:** You have not lost your loved one. He or she and you have a lot of living to do. Focus on today and on what you can do, not what you can't.



You may experience a range of emotions when your loved one is diagnosed.

Stage 1: What to Expect From the Person With IPF in This Stage

- The person with IPF in Stage I may be entirely asymptomatic; however, it would not be uncommon for them to have shortness of breath when exerting, dry cough and/or fatigue.
- Shortness of breath may only be present with extreme exertion (e.g., climbing a hill). Remember, even people without IPF are at least a little short of breath going up two flights of stairs.
- Cough can be frequent and bothersome. It may only occur when the person with IPF talks for long periods of time or when they are exerting.
- Fatigue is common and challenging to treat. Eating well, exercising and getting plenty of high quality sleep can help.
- The person with IPF who is just diagnosed may experience an array of emotions. They may be afraid or angry, anxious or short-tempered.
- Many people with IPF in Stage I want to know more about the disease. We discourage online searching at random sites, because much of the information is outdated or inaccurate.

Stage 1: Questions to Ask Your Health Care Provider

- What treatments are available to me now?
- How will those treatments affect my quality of life?
- What should my caregiver and I expect as my disease progresses?
- Are there any support groups available for me to connect with fellow people with IPF?
- Are there good websites that provide helpful, reliable information?
- Do I need any vaccines?
- Do I have or need treatment for acid reflux?
- Do I have pulmonary hypertension? Should I be tested?
- Do I have obstructive sleep apnea? Should I be tested?

Stage 2: Needing Oxygen with Activity, But Not At Rest

What should I focus on when I am needing oxygen with activity, but not at rest?

- Work with your health care provider. The health care provider will write a prescription for oxygen, based on your oxygen saturations with rest and activity. A durable medical equipment (DME) company will provide the oxygen.
- Learn about the medical equipment. This includes the stationary/in-home and portable oxygen systems that are available.
- Use a pulse oximeter to check your oxygen saturations when you are active or exercising to make sure you are getting the right flow of oxygen to maintain your saturation at equal to or greater than 89 percent.
- Ask about the medications that may be used for IPF. Learn the dose and time the medicine is taken, how it may be helpful and what side effects to watch for.
- Breathing techniques may be helpful with the shortness of breath that can occur with exercise. Learn and practice the breathing techniques.
- Remember to continue living a full life with IPF activities.
 - Regular exercise is important, and oxygen will help you feel better while you exercise.
- Think about whether lung transplantation is an option for you personally, and share this information with your pulmonologist.



Stage 2: How Can the Caregiver Help?

- Continue to be a second set of ears with the health care provider, and now with the durable medical equipment (DME) company also. This is a new company you will both need to work with.
- Continue to discuss and write down questions.
- Learn the ins and outs of the new medical equipment. Examples include oxygen systems, portable oxygen systems and pulse oximetry
- Encourage the person with IPF to live a full life, including regular exercise, healthy eating, rest and support from others.
- Read the book *Adventures of an Oxy-Phile* by Dr. Thomas Petty.
- Remember to take care of yourself.



Stage 2: What to Expect From the Person With IPF in This Stage

- The person with IPF in Stage II is short of breath with exertion or activity.
- Cough can be frequent and bothersome. It may only occur when the person with IPF talks for long periods of time or when they are exerting.
- Fatigue is not uncommon at this stage and can be challenging to treat. Eating well, exercising and getting plenty of high quality sleep can help.
- Being prescribed oxygen for use during the day is perceived by many people with IPF as a major step in the wrong direction. But, oxygen is not a death sentence! Even so, some people with IPF feel self-conscious using their oxygen in public. This is natural and usually short-lived.

Stage 2: Questions to Ask Your Health Care Provider

- Does staying active actually extend my life or does it just improve my quality of life?
- What resources are available to help me be active with oxygen? Can I travel with oxygen?
- Do I have the freedom to adjust flow rates based on how I am feeling, or am I bound to the prescribed flow rate?
- Do I need to be extra careful when exercising when using oxygen, or is it based on what I can tolerate?
- Do you recommend formal pulmonary rehab or self-initiated physical activity or both?
- Will you monitor my oxygen needs at clinic visits? If so, how?
- Are my vaccinations up to date?
- Is there a transtracheal oxygen program nearby? Am I a candidate for the procedure?



Stage 3: Needing Oxygen 24 Hours a Day, With Activity, At Rest and During Sleep

What do I focus on when needing oxygen at rest, activity and sleep?

- Work with your health care provider in the oxygen prescription change and the best oxygen system to meet your needs for using oxygen 24 hours a day.
- Learn about stationary oxygen systems and portable systems.
- Work with the DME company regarding the change in | oxygen needs.
- Plan for oxygen use with activities when away from home and traveling. Plan ahead for travel.
- Use your pulse oximeter to monitor your oxygen level. Maintain an oxygen saturation equal to or greater than 89 percent when you rest and are active.
- Your health care provider may discuss palliative care. Ask any questions you have.
- Remember to stay active, and continue living a full life with IPF.
 - Oxygen is now important 24 hours a day and with regular exercise. Regular exercise is still important, and oxygen will help you feel better while you exercise.
- If transplant is an option for you, make sure you are evaluated at a transplant center.

Stage 3: How Can the Caregiver Help?

- Learn the ins and outs of oxygen systems for use at home, when away from home and when traveling.
- Continue to be a second set of ears, with the durable medical equipment (DME) company as the oxygen needs change.
- Continue to discuss and write down questions.
- Encourage the person with IPF to live a full life, including social interactions, regular exercise, healthy eating, rest and support from others.
- If you haven't yet, get the book *Adventures of an Oxy-Phile* by Dr. Thomas Petty.
- Remember to take care of yourself.



Remember to stay active and continue living a full life with IPF.

Stage 3: What to Expect From the Person With IPF in This Stage

- The person with IPF in Stage III is short of breath with exertion or activity. Even though they need oxygen at rest, they will not be short of breath at rest.
- Cough can be frequent and bothersome. It may only occur when the person with IPF talks for long periods of time or when they are exerting.
- Fatigue is not uncommon at this stage and can be challenging to treat. Eating well, exercising and getting plenty of high quality sleep can help. Napping is okay and encouraged.
- Being prescribed oxygen for use 24 hours per day is perceived by many people with IPF as yet another major step in the wrong direction. But, even 24/7, oxygen is not a death sentence! Nonetheless, people with IPF may feel afraid, anxious or depressed and be short-tempered with this new stage.



Stage 3: Questions to Ask Your Health Care Provider

- Do I need to be careful about physical activity?
- Am I a candidate for a lung transplant? If so, what do I do?
- Are my vaccinations up to date?
- Should I enroll or re-enroll in pulmonary rehabilitation?
- Do I have pulmonary hypertension?

Stage 4: Advanced Oxygen Needs (needing High-Flow Oxygen or When a Lightweight, Portable Delivery System is Unable to Meet Oxygen Needs)

- Work with your health care provider regarding the oxygen prescription change and the best oxygen system to meet your needs for using oxygen as the liter flow increases 24 hours a day. High liter-flow is described as greater than 4 liters. Transtracheal oxygen may be an option.
- Learn about stationary oxygen systems and portable oxygen systems that will meet your oxygen needs at high liter-flows.
- Learn how long the portable oxygen system will last at higher liter-flows.
- Work with the DME company regarding the change in oxygen needs.
- Use your pulse oximeter to monitor your oxygen level. Maintain an oxygen saturation greater than or equal to 89 percent when you rest and are active.
- Talk with your health care provider about palliative care.
- Your health care provider may talk with you about the possibility of a lung transplant.
- Remember to stay active, and continue living a full life with IPF.

Stage 4: How Can the Caregiver Help?

- Learn the ins and outs of high-flow oxygen system at home, when away from home and when traveling. Calculate how long the portable oxygen system will last when away from home.
- Continue to be a second set of ears, with the durable medical equipment (DME) company as the oxygen needs change.
- Continue to discuss and write down questions.
- Encourage the person with IPF to live a full life, including social interactions, regular exercise, healthy eating, rest and support from others.
- Remember to take care of yourself. Consider respite care. Consider palliative care and/or hospice care as indicated.

Stage 4: What to Expect From the Person With IPF in This Stage

- The person with IPF in Stage IV is short of breath with exertion or activity. Even though they need oxygen at rest, they will not usually be short of breath at rest.
- Cough can be frequent and bothersome. It may only occur when the person with IPF talks for long periods of time or when they are exerting.
- Fatigue is not uncommon at this stage and can be challenging to treat. Eating well, exercising and getting plenty of high quality sleep can help. Napping is okay and encouraged.
- High-flow oxygen cannot be delivered by portable oxygen concentrators, so many people with IPF feel even more constrained. Remember, even high-flow oxygen is not a death sentence! Nonetheless, people with IPF may feel afraid, anxious or depressed and be short-tempered with this new stage. They are at risk for social isolation and mood disturbance.



Stage 4: Questions to Ask Your Health Care Provider

- What is the normal life expectancy at this point?
- What palliative care resources are available?
- What does the end look like?
- Can you guarantee that I will not suffer?




Remember Your Goals.

- Decreasing shortness of breath
- Reducing exacerbations
- Becoming more active
- Decreasing anxiety or stress
- Improving mood
- Learning more about IPF

Think of other goals you may have.

Write any other goals in the space below:

Talk with your health care provider about reaching your goals to live a full life with IPF. Ask any questions you have. Your health care provider is your partner in managing your IPF.

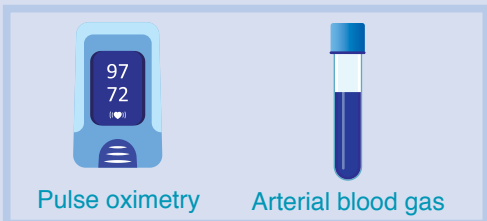


On The Go with Oxygen



If your oxygen level is 88 percent or below, oxygen therapy is often recommended to ensure your blood has enough for your body's needs.

HOW OXYGEN LEVEL IS MEASURED



Pulse oximetry

Arterial blood gas

Your oxygen level can be measured from a device on your finger or through a blood test.

OXYGEN PRESCRIPTION INCLUDES



- How much oxygen to use in liters
- When to use the oxygen, (rest, activity, sleep, altitude)
- Which oxygen system is recommended

Your doctor prescribes the oxygen.

A durable medical equipment company provides the oxygen.

TYPES OF OXYGEN SYSTEMS

- **Concentrators**
- **Portable concentrators**
- **Compressed gas**
- **Liquid**

ON DEMAND OR PULSE FLOW

VS

CONTINUOUS FLOW

Provides oxygen when you breathe in by triggering oxygen release (not recommended for sleep or everyone)

Provides a set liter flow of oxygen

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Busting Myths about Oxygen Therapy



If oxygen therapy is recommended for you, ignore the myths and get the facts.

Myth #1

Oxygen means I can't be active.



You will have less shortness of breath and can be more active on oxygen.



Myth #2

Oxygen is addictive.

Oxygen is not addictive.

Myth #3

Oxygen means this is the end.

No, oxygen therapy helps you to live longer, feel better and be more active.

Myth #4

Portable oxygen concentrators meet all oxygen needs.

Portable oxygen concentrators are not good for continuous flow or higher liter flow needs.



Myth #5

Petroleum jelly is best for nose dryness.

Water-based moisturizers are best for nose dryness.

Myth #6

I can't travel with oxygen.

You CAN travel with oxygen; it just takes some planning.



Myth #7

I can use a portable oxygen concentrator during sleeping.

Portable oxygen concentrators don't provide enough oxygen for sleeping.



Myth #8

Oxygen will cause a fire.

Oxygen is not flammable; it supports a flame, but does not cause fires.



Myth #9

Everyone will see the oxygen tubing on my face.

Maybe, but there are many options for oxygen delivery, even glasses.

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Four HO₂PES

Being prescribed oxygen therapy can be life-changing. However, maybe not all of the change is for the worse. Maybe there are some benefits to using oxygen therapy. Some people find that oxygen therapy allows them to do more, to be more active, to have greater stamina, to revisit hobbies they've left behind. What do you hope using oxygen therapy will do for you?

HO₂PE #1: _____

HO₂PE #2: _____

HO₂PE #3: _____

HO₂PE #4: _____

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