

Living With

INTERSTITIAL LUNG DISEASE

ILD

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LEARN ABOUT ILD

ILD is short for “interstitial lung disease.” It refers to a group of diseases that cause lung inflammation and/or permanent scars (“fibrosis”). When you have ILD, your lung tissue becomes inflamed. The cause might be known or unknown. The onset of ILD is usually gradual, but it can occur quite fast in some people. Over time, lung inflammation can cause fibrosis. This scarring causes changes in the small air pockets in your lungs (called “alveoli”). They thicken, lose their normal shape, and become stiff

Eventually, it becomes hard for your lungs to work normally and get oxygen into your blood. Supplemental oxygen can help you stay active and strong.

Low blood oxygen levels may:

- *Cause shortness of breath and tiredness*
- *Reduce the function of your organs*
- *Make it hard to maintain your quality of life*

You deserve a happy and fulfilling life.

Effective treatments can help you manage your health.

That’s why it’s important to see a lung doctor early.

Determining the cause of your ILD (if there is one) is very important to your management and treatment.



APPROXIMATELY
400,000 PEOPLE IN THE UNITED
STATES **LIVE WITH ILD TODAY**

ILD SYMPTOMS ARE SIMILAR TO
THOSE OF OTHER LUNG DISEASES,
SO **HALF OF ALL CASES**
ARE INITIALLY MISDIAGNOSED

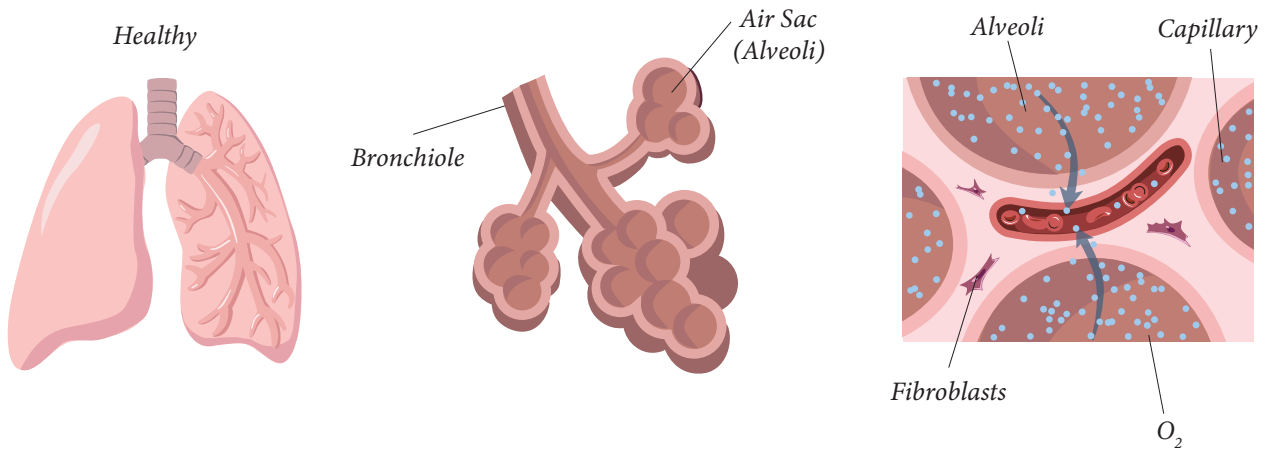
MANY ILD PATIENTS REPORT
WAITING MORE THAN ONE
YEAR FOR THE RIGHT DIAGNOSIS

IN MANY CASES **PATIENTS DO**
NOT NEED A LUNG BIOPSY
TO MAKE A DIAGNOSIS OF ILD

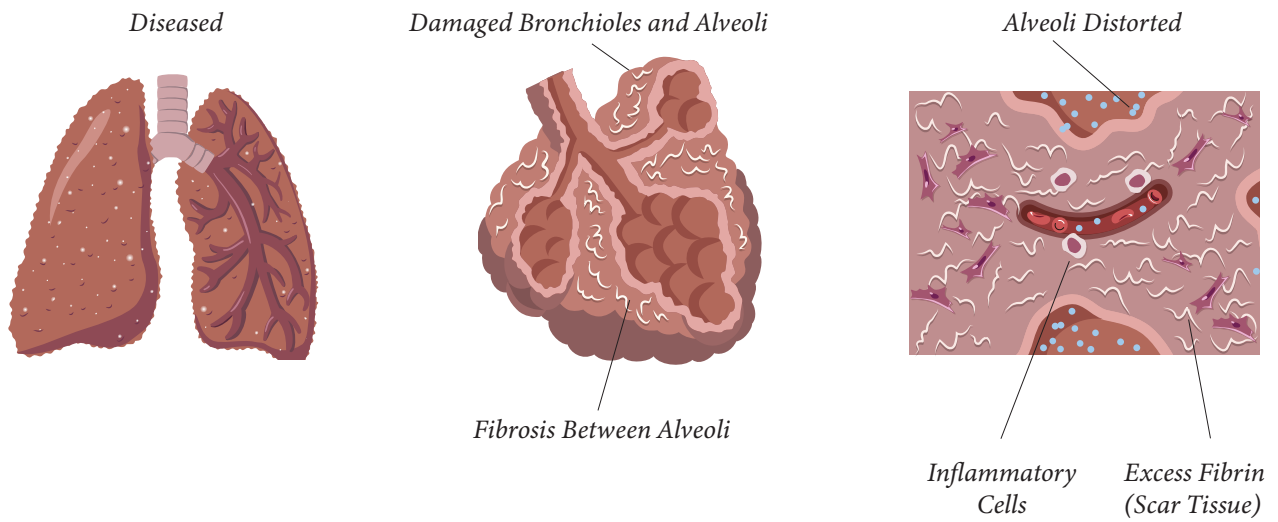
MANY ILD PATIENTS ALSO HAVE
GASTROESOPHAGEAL
REFLUX DISEASE (GERD)—SOME
DON'T HAVE REFLUX SYMPTOMS BUT
STILL HAVE ACID REFLUX

MONITORED EXERCISE TRAINING
(PULMONARY REHABILITATION)
CAN HELP MANAGE ILD AND
SIGNIFICANTLY IMPROVE
QUALITY OF LIFE

Healthy Lung



ILD Lung





Living with Different Types of ILD

David has an ILD called “idiopathic pulmonary fibrosis” (IPF for short), and Cheryl has “hypersensitivity pneumonitis.” Steroids and medications that suppress the immune system will help Cheryl get better, but these medications won’t help David. David has other medications that he can take to keep his IPF from worsening over time.



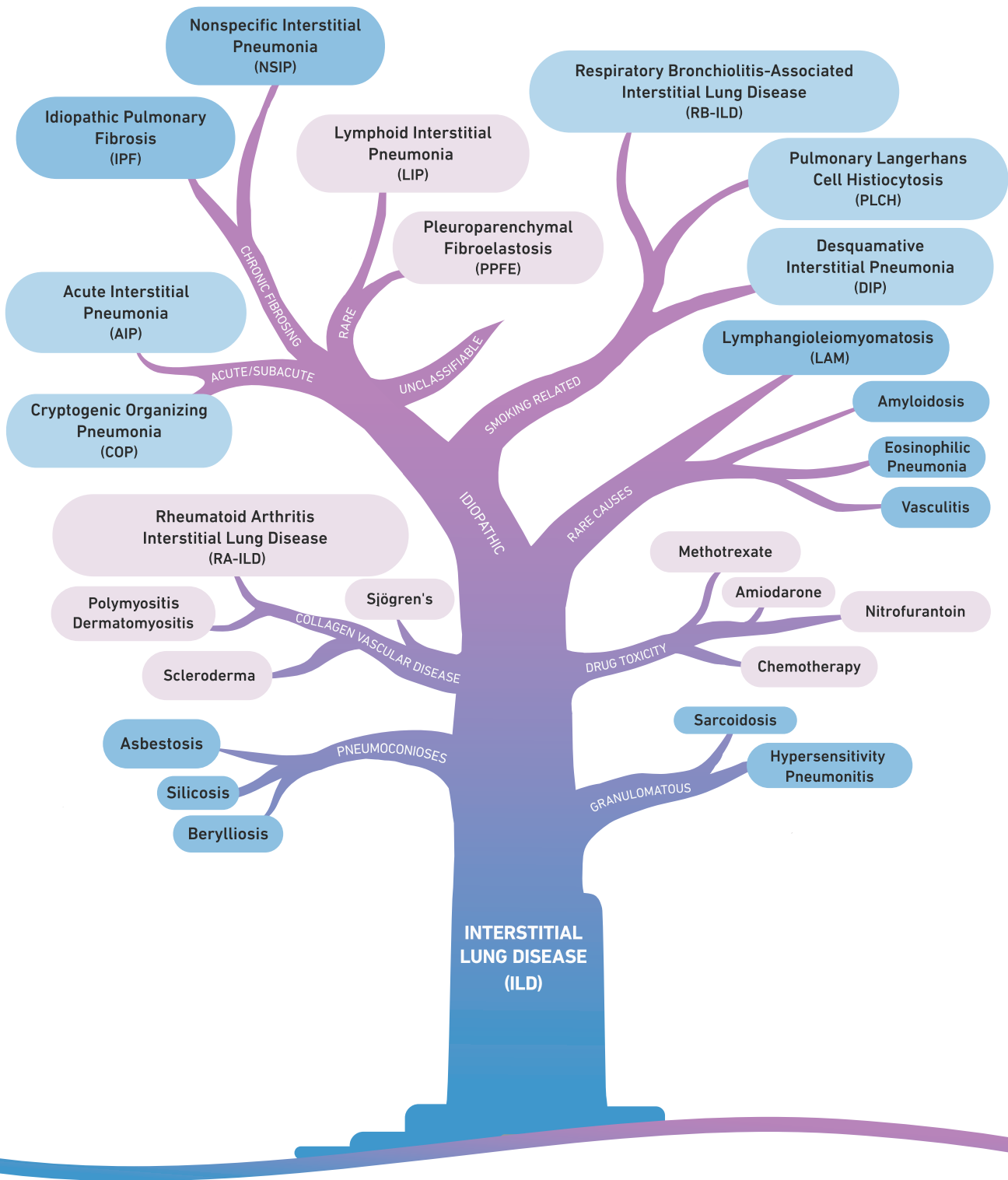
TYPES & CAUSES OF ILD

The causes of ILD can be hard to find. Sometimes ILD is caused by another systemic disease, like rheumatoid arthritis. At other times, it is caused by something in the environment in your home or at your job.

Many times, doctors cannot identify the cause of your ILD. In those cases, we say your ILD is “idiopathic.”

The progress and treatment of ILD will be different depending on the type of ILD you have. That is why your doctor will perform a thorough evaluation. Different tests will be ordered to search for any identifiable cause of your ILD.

Talk to your doctor about the medications that are best for your type of ILD. Knowing the cause of your condition will help you and your doctor decide on the best treatment. Treating your ILD will help you to live with a better quality of life.



ILD FAMILY TREE

Types of ILD

There are many types of ILDs. Sometimes a diagnosis can seem overwhelming. Here are some common types of ILDs. Those with similar causes or findings are grouped together.

PNEUMOCONIOSIS

Some people get ILD by being exposed to certain inorganic or organic dust particles. These harmful substances can be in places where you spend a lot of time, such as your work, but they can also be environmental.

Some harmful substances that may cause ILD are:

- Beryllium
- Birds or bird feathers (in down blankets, coats, and pillows)
- Dusts, such as aluminum, antimony, barium, graphite, iron, kaolin, mica, or talc
- Asbestos
- Silica
- Mold caused by water damage in your home or office or in hot tubs

WORKPLACE EXPOSURES BY JOB

Some common substances in the workplace can cause ILD, and you might be exposed to them in certain types of jobs.

HARMFUL SUBSTANCE	JOB / EXPOSURE
ASBESTOS	Construction worker (insulation); shipyard worker; mechanic (brake pads)
SILICA	Sand blaster
COAL DUST	Coal miner
BERYLLIUM	Denture manufacturer
GRAIN DUST	Peanut industry worker
HARD METAL DUST cobalt, tungsten, etc.	Steelworker

COLLAGEN VASCULAR DISEASE

When someone has an autoimmune disease or a connective tissue disease, their immune system attacks their body. Autoimmune diseases, like rheumatoid arthritis, can cause ILD.

If the autoimmune disease targets the lung, it can cause inflammation and scarring of the lung tissue. Sometimes people don't even know they have an autoimmune disorder until after they develop ILD.

Some autoimmune diseases that may cause ILD are:

- *Rheumatoid arthritis*
- *Systemic sclerosis*
- *Sjögren's syndrome*
- *Systemic lupus erythematosus*
- *Polymyositis/dermatomyositis*
- *Granulomatosis with polyangiitis*

GRANULOMATOUS DISEASE

Granulomas are collections of cells that form in some inflammatory conditions. They can occur all over the body. When they form in the lung tissue, different signs and symptoms of ILD can appear.

Some granulomatous conditions that can lead to ILD are:

- *Sarcoidosis*
- *Crohn's disease*
- *Hypersensitivity pneumonitis (common causes include dust, mold, and bird feathers)*

DRUG INDUCED (MEDICATION)

Some medications play a role in the development of ILD. The type of medicine, the amount of time someone takes it, and the dosage all play a part.

Some medications that can lead to ILD are:

- *Cancer treatments (bleomycin)*
- *Medications for rheumatoid arthritis*
- *Immunotherapy drugs (nivolumab, pembrolizumab)*
- *Heart medications (amiodarone)*
- *Antibiotics (nitrofurantoin)*

RARE ILD

Some ILDs are extremely rare but have specific features that distinguish them from other ILDs. These features can be noticed in a computed tomography (CT) scan or in laboratory tests (“pathology”).

Some rare ILDs are:

- *Amyloidosis*
- *Vasculitis*
- *LAM (short for “lymphangioleiomyomatosis”)*
- *Familial ILD*
- *Radiation pneumonitis*
- *Eosinophilic pneumonia*
- *Related to vaping or inhaling marijuana-like substances*

RADIATION TREATMENT | Radiation is often used to treat cancer in the lung, throat, or breast. Radiation might help treat cancer, but it sometimes causes ILD. A person may develop ILD weeks to years after having radiation treatments.



Familial ILD (Genetic)

Although not common, some ILDs tend to run in families. Idiopathic pulmonary fibrosis (IPF) runs in families 10% to 15% of the time. That means for every 100 people with IPF, 10 to 15 of them will have family members with the disease.

Genetic changes that cause IPF are passed down from a parent to a child. Genetic tests for IPF are not a part of a routine workup.

- If more than one person in your family has pulmonary fibrosis, ask your lung doctor about getting a genetic test.
- You may be referred to a special counselor who specializes in genetic diseases.

IDIOPATHIC (NO KNOWN CAUSE)

More than half of ILD cases are “idiopathic.” This simply means they have no known cause. IPF is the best example of an idiopathic ILD.

Before labeling your ILD as idiopathic, your doctor will do an extensive workup. Your doctor will find out if your ILD has an identifiable cause.

Idiopathic ILDs can be:

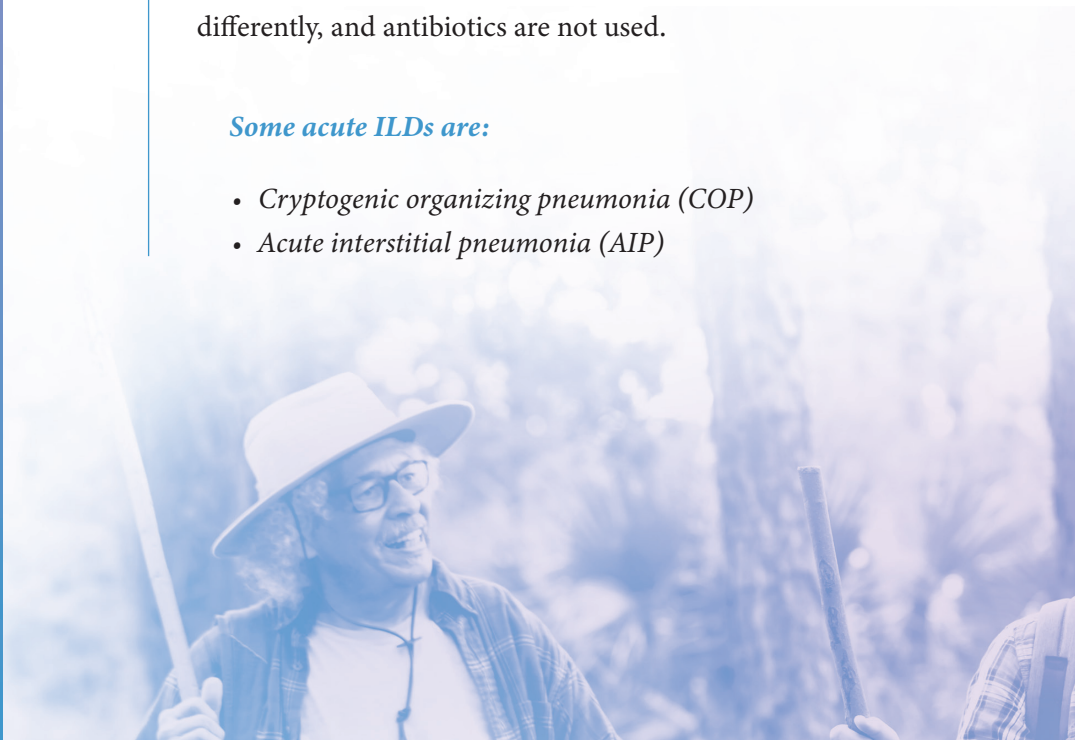
- *Acute: severe and sudden over days to weeks*
- *Subacute: uncomfortable over weeks to months*
- *Chronic: discomfort that is ongoing or recurring*

Idiopathic ILDs can be associated with smoking or other rare causes. Sometimes they are unclassifiable.

ACUTE | Acute Idiopathic ILDs usually present like pneumonia but are not infections. These ILDs are treated differently, and antibiotics are not used.

Some acute ILDs are:

- *Cryptogenic organizing pneumonia (COP)*
- *Acute interstitial pneumonia (AIP)*



CHRONIC FIBROSING | Chronic Fibrosing ILDs may take time to cause symptoms. By the time the patient has symptoms, the lung already has scarring that cannot be reversed. IPF is one type of a chronic fibrosing ILD with no underlying identifiable cause (some known causes can also trigger fibrosis). Another type is chronic nonspecific interstitial pneumonia (NSIP).

SMOKING RELATED ILDS | Smoking usually causes chronic obstructive pulmonary disease (COPD) or emphysema. Smoking rarely causes ILD. For smokers who do develop ILD, the first step to treating these ILDs is to stop smoking.

Some ILDs caused by smoking are:

- *Respiratory bronchiolitis-ILD (RBILD)*
- *Desquamative interstitial pneumonia (DIP)*
- *Langerhans cell histiocytosis (LCH)*

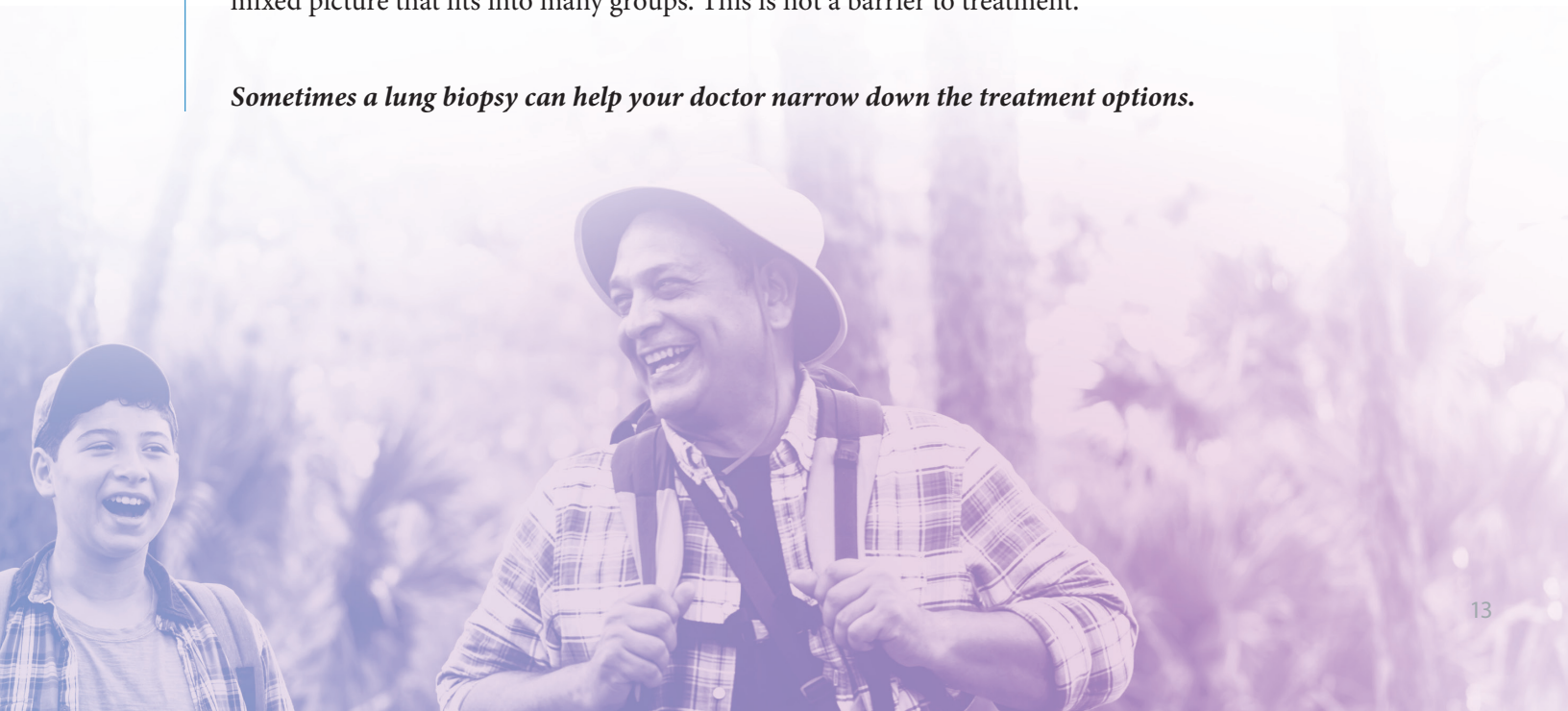
RARE IDIOPATHIC ILDS | Some ILDs are extremely rare but have specific features that distinguish them from other ILDs.

Some rare idiopathic ILDs are:

- *Lymphoid interstitial pneumonia (LIP)*
- *Pleuroparenchymal fibroelastosis (PPFE)*

UNCLASSIFIABLE ILDS | Some ILDs do not fall into any particular group. They may have a mixed picture that fits into many groups. This is not a barrier to treatment.

Sometimes a lung biopsy can help your doctor narrow down the treatment options.



SYMPTOMS OF ILD

Symptoms of ILD are often difficult to treat. They can be very debilitating for people living with ILD.

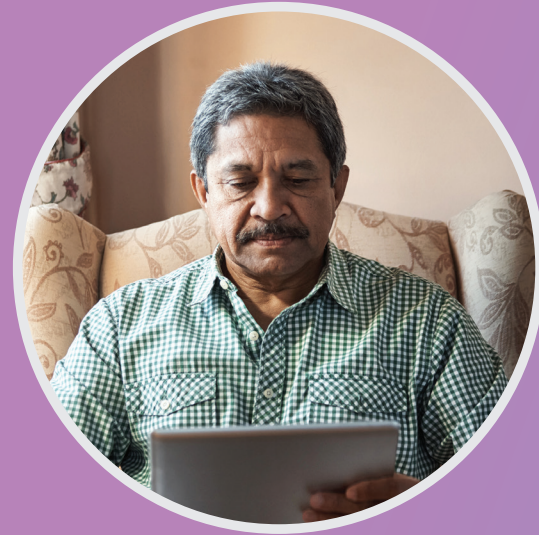
Shortness of breath is common for people with ILD. However, ILD symptoms could also point to other conditions:

- *Chronic obstructive pulmonary disease (COPD)*
- *Heart failure*
- *Anemia*
- *Blood clots*
- *Heart disease*

A dry cough is very common for people with ILD, but it could also point to:

- *Asthma*
- *Reflux*
- *Post-nasal drip (where mucus builds up and drips down behind the nose and throat)*
- *Allergies*

Symptoms of ILD can be hard to treat and can get in the way of your normal routine.



John is 57 years old and visited his doctor because he hasn't been feeling well.

He has a cough that seems to never go away, and he has been short of breath while doing his normal activities. He's even having a hard time exercising.

As his symptoms get worse, John finds it hard to keep his usual routine and do the things he once did without trouble.

John has been feeling worried and sad about his condition, and these feelings aren't going away. John decides to call his doctor for advice.

To find out for sure if John has ILD, his doctor will perform different tests. After a complete evaluation and finding out more about John's health and work history, John's doctor is sure that John has an ILD.

Together, John and his doctor will work to manage John's symptoms.



To feel less short of breath, John can:

- Treat the underlying cause of his ILD (if there is one)
- Stay active
- Participate in pulmonary rehabilitation

Low-dose opioids (morphine) can help reduce the feeling of shortness of breath in advanced ILD.

To find relief from a dry and persistent cough, John's doctor might prescribe gabapentin or opioids. Participating in a clinical trial can help find new treatments.

To exercise better and longer, John should start a pulmonary rehabilitation program to learn breathing techniques to use while doing physical activities.

To help with anxiety and depression, John's doctor might prescribe benzodiazepines and antidepressants. If you feel anxious or depressed, please seek advice from your physician. Your mental health is just as important as your physical health.

To keep his ILD from getting worse, John should:

Health

- Stop smoking
- Keep up with doctor visits, scheduled tests, and vaccinations
- Practice relaxation techniques
- Breathe with pursed lips
- Eat small meals
- Maintain an ideal body weight

Home

- Get rid of pets, birds, carpets, down pillows, and comforters
- Use fans to blow cold air
- Install bathroom aids
- Use a wheelchair or walker

DIAGNOSIS & WORKUP

At your first appointment, your pulmonologist (lung doctor) will ask a lot of questions. To make it easier for you, you may be given the questions ahead of time.

Expect to be asked about:

- *Your health history*
- *Any surgeries you've had*
- *Your close family's health history*
- *Any pets you have at home*
- *Your current and past jobs*
- *Your hobbies*
- *Your current and previous medications, including:*
 - * *herbal medication*
 - * *anything over the counter, like vitamins*

Your doctor will also run various tests.

Expect to have:

- *A complete physical exam*
- *Blood tests*
- *A breathing test, called a "pulmonary function test"*
- *Imaging tests*

Your doctor may also want to do a lung biopsy to better diagnose your type of ILD.

A biopsy looks at a small piece of tissue taken from your lungs. There are different ways your doctor can do a biopsy.



Types of Tests

BLOOD TESTS

Your doctor will order a comprehensive panel of blood tests. The blood tests will help search for any identifiable causes of ILD.

Basic blood tests

- *Complete blood count (CBC) to check the types of cells in your blood*
- *Complete metabolic panel to check how well your liver and kidneys work*
- *Tests to see how well your blood clots*

Other blood tests will be done to check for collagen vascular or autoimmune diseases such as:

- *Rheumatoid arthritis*
- *Lupus*
- *Scleroderma*
- *Sjögren's syndrome*
- *Sarcoidosis*
- *Vasculitis*
- *Dermatomyositis or polymyositis*

Blood tests may also be done to check for infections like tuberculosis and hepatitis.

To check your blood oxygen levels, your doctor may also take a small sample of blood from your wrist for an arterial blood gas (ABG) test. This will let your doctor know if you need supplemental oxygen.

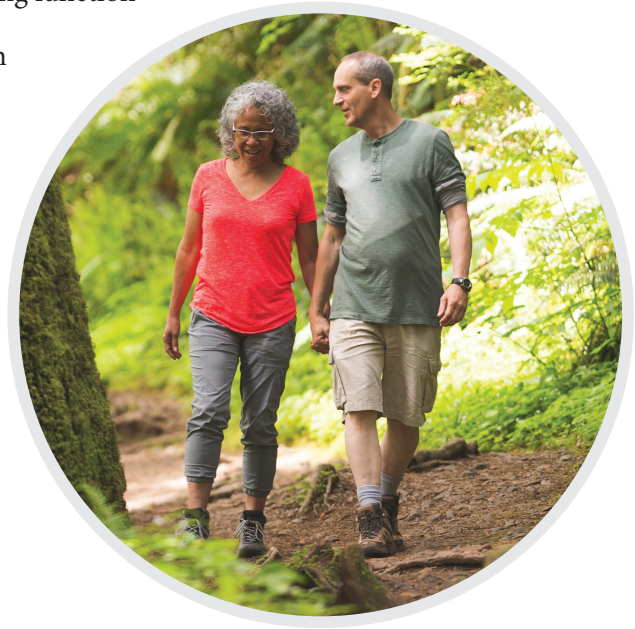
LUNG FUNCTION TESTS

PULMONARY FUNCTION TEST | The ***Pulmonary Function Test (PFT)*** is a series of breathing exercises to check how much air your lungs can hold and how quickly and forcefully you can breathe in and out. The PFT will also check how well oxygen can move in and out of your lungs.

The PFT takes about 45 minutes. It is done multiple times a year to check for any progression of your ILD.

SIX-MINUTE WALK TEST | The **Six-Minute Walk Test** checks how far you can walk in 6 minutes. It will be repeated during the year to see if your lung function has changed. You will wear a heart rate and oxygen saturation monitor during this test. It will also determine if you need to use oxygen.

OVERNIGHT OXIMETRY | **Overnight Oximetry** records your oxygen levels while you sleep. This test checks to see if you need oxygen therapy while sleeping. This test is different than an evaluation for sleep apnea, which requires an overnight sleep study.



IMAGING TESTS

X-RAY | A chest X-ray gives your doctor a quick view of your chest, lungs, and heart. This test will probably be one of the first performed if no other imaging tests have been done.

COMPUTED TOMOGRAPHY (CT) SCAN | A CT scan uses a computer to combine many X-rays taken from different angles. It gives a very precise view of your organs. A high-resolution CT (HRCT) scan will allow your doctor to see your lungs with more detail than a traditional CT scan. These scans take only a few minutes but give your doctor important information about your ILD.

A detailed picture of your lungs helps your doctor diagnose your ILD. It will also let your doctor see how much scar tissue is in your lungs. If the image is good enough, you may not need a lung tissue biopsy. CT scans will be repeated yearly or more often to closely follow your ILD.

ECHOCARDIOGRAPHY | An echocardiogram is an ultrasound of your heart. It uses sound waves to create pictures of your heart. These pictures help evaluate the function and strength of your heart and calculate pressures.

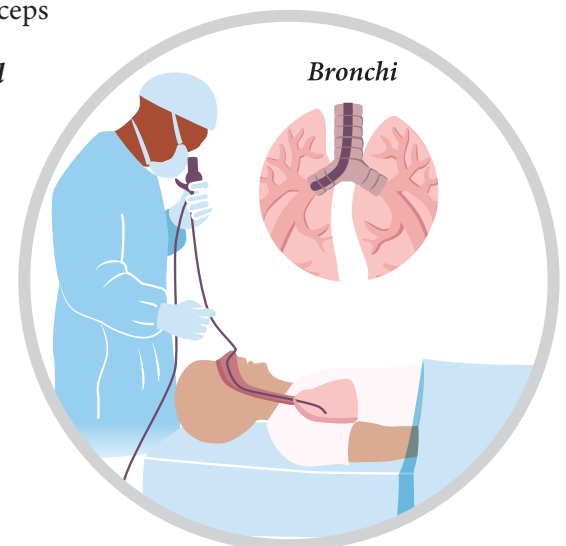
This test helps your doctor know if your ILD is affecting your heart. ***It will also tell your doctor if you have a type of high blood pressure that affects the blood vessels of your lungs (“pulmonary hypertension”).***

TISSUE SAMPLING TESTS

Sometimes a good history, imaging, and blood tests are good enough to make a diagnosis. However, your doctor might need to take a small amount of tissue from your lungs to make a correct diagnosis. There are different ways to obtain lung tissue, and each has its pros and cons.

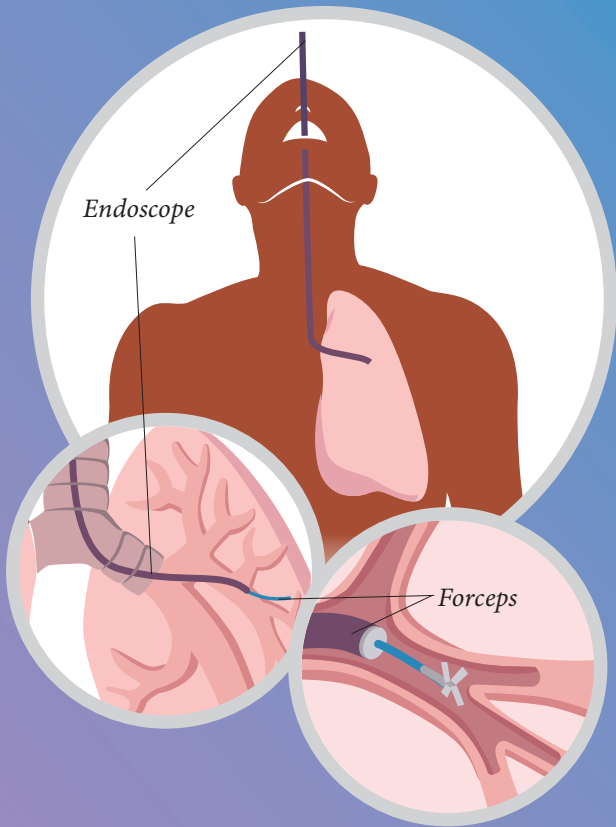
BRONCHOSCOPY | During a bronchoscopy, a long, bendable tube with a camera is placed in your nose or mouth and inserted into your windpipe (trachea). The camera allows your doctor to see inside your lungs. Tools can then be used to collect fluid. Forceps can also be used to obtain samples of lung tissue (***transbronchial biopsy***). The lung tissue samples are small—about the size of breadcrumbs.

You will need anesthesia for this test, but in most cases, you won't have to spend the night in the hospital. Anesthesia is medication to keep you from feeling pain. Some types of anesthesia can put you to sleep. Local anesthesia numbs the pain in an area of your body but does not make you tired. General anesthesia puts you to sleep, so you are not awake and do not feel any pain during your procedure.

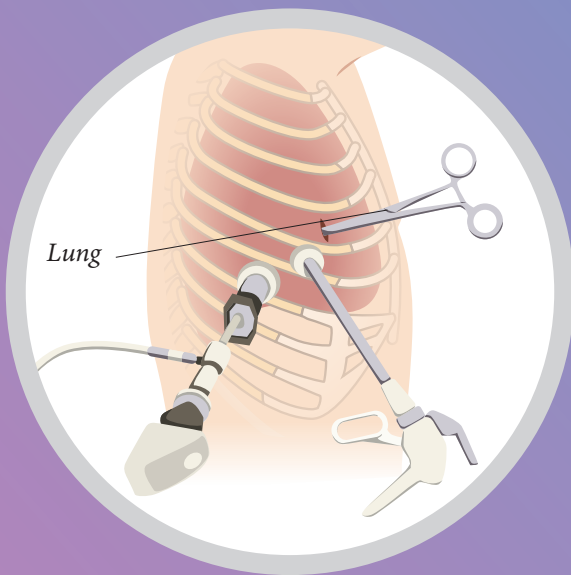


Bronchoscopy

Transbronchial Biopsy



VATS Biopsy



CRYOBIOPSY | Some medical centers do a newer tissue sample test called a *cryobiopsy*. A bronchoscopy is performed. But, for this test, a small region of your lung is frozen with a special tool. This allows your doctor to take a bigger sample of lung tissue than the sample obtained by traditional bronchoscopy forceps. To do this test, you will need anesthesia, and you may need to stay in the hospital for observation.

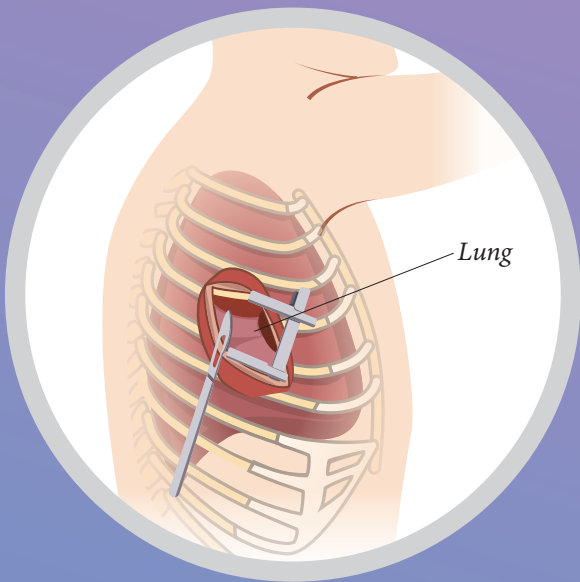
Cryobiopsies are not routinely done in all hospitals. They are usually performed at large academic hospitals. Cryobiopsies carry a higher risk of bleeding than a routine bronchoscopy.

VIDEO-ASSISTED THORACOSCOPIC SURGERY (VATS) |

This tissue sample test is a type of lung surgery. A surgeon will use a few small incisions (cuts) in your chest in between your ribs to insert tools into the chest cavity. Your doctor will use a camera to help guide the procedure. After that, other tools are used to take a larger tissue sample from your lung. This is a less invasive surgery than a traditional thoracotomy.

You will need to spend the night in the hospital for this surgery, and you will need general anesthesia to be put to sleep for the test.

Thoracotomy



CT-Guided Needle Biopsy



THORACOTOMY | This surgery is one of the most invasive ways of getting a tissue sample. A surgeon will do a traditional incision (cut) to open your chest and take a few samples of your lung tissue. You will have a tube in your chest (chest tube) for several days after the surgery as you recover in the hospital.

For this type of surgery, you will need to spend the night in the hospital. You will also need general anesthesia to be put to sleep for the test.

CT-GUIDED NEEDLE BIOPSY |

Sometimes your doctor will be able to take a sample of lung tissue with a needle while also performing a CT scan. However, this is not the best way to get a tissue sample and diagnose a person with ILD.

The skin where the needle is inserted is numbed with local anesthesia, so you won't need to be asleep. There is a moderate risk that your lung may collapse when the needle is inserted. If this happens, you will be observed closely, or a small chest tube will need to be inserted to re-expand your lung.

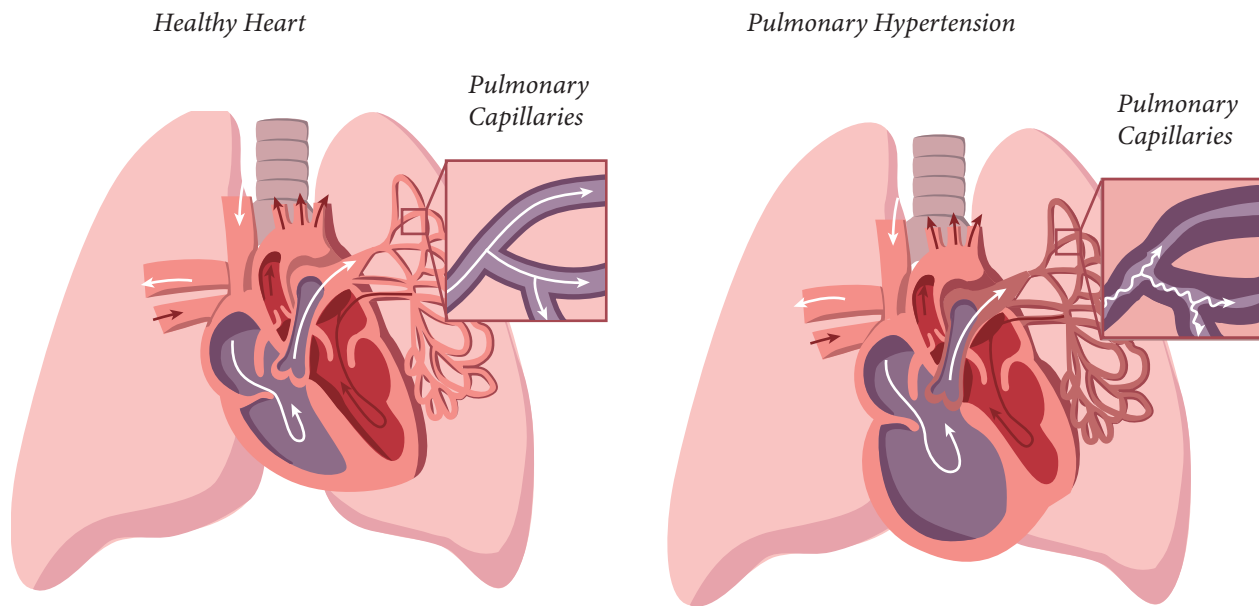
COMORBIDITIES

ILD mainly affects your lungs, but it can also cause problems in other parts of your body. Your doctor will check to see if other organs are affected and will monitor you regularly. Treating other existing conditions will help you and your doctor better manage your ILD.

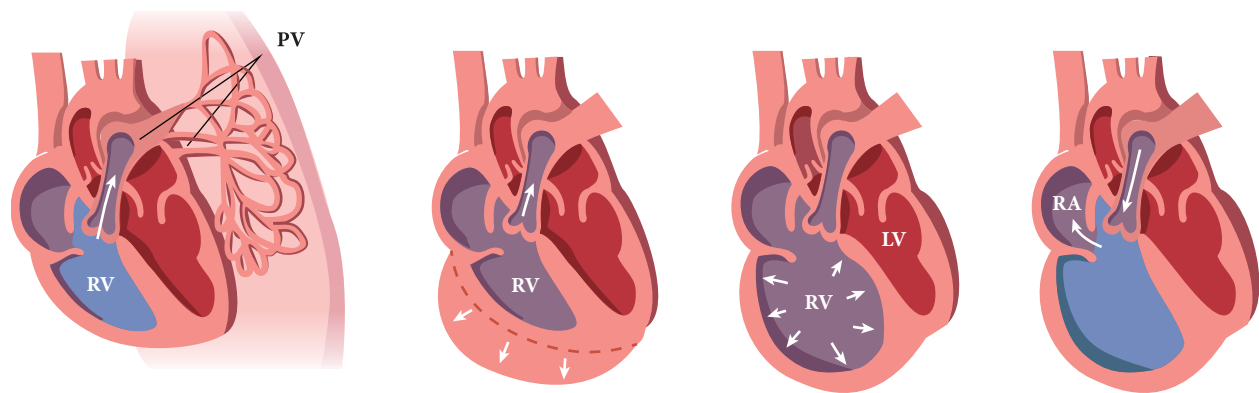
You can have one or more conditions along with your ILD. If you have symptoms or are worried about having other conditions, talk with your doctor.



Pulmonary Hypertension



Right Heart Failure Due to Pulmonary Hypertension



Narrowing of the pulmonary vessels (PV) leads to backward pressure on the right ventricle (RV).

The RV begins to enlarge to adapt to the increased pressure load.

The RV becomes so dilated it starts to push on the left ventricle (LV).

Blood starts to go backward into the right atrium (RA) and leads to signs of right-sided congestive heart failure.

PULMONARY HYPERTENSION

Over time, your lung scarring might get worse. This can affect the vessels that carry blood throughout your lungs. The pulmonary vessels may develop fibrosis, become narrower and limit normal blood flow. This is called ***pulmonary hypertension***. It is a very serious complication of ILD.

Some diseases that cause ILD also cause pulmonary hypertension. Pulmonary hypertension makes it very hard to breathe when you exercise. Work with your doctor to manage your pulmonary hypertension.

Your doctor might prescribe supplemental oxygen therapy to help with your symptoms and protect your heart.

CONGESTIVE HEART FAILURE

If you have ILD, it is important to monitor for heart failure. In congestive heart failure, the heart doesn't pump blood as well as it should. For people with ILD, this is usually caused by too much pressure in the blood vessels of the lungs (*pulmonary hypertension*).

Having too much pressure in your lungs' blood vessels, aka, pulmonary hypertension, causes a lot of stress on the right side of the heart. The right side of the heart pumps blood into the lungs. Congestive heart failure can cause shortness of breath.

As your ILD gets worse, your heart may not work as well as it used to. Your doctor will run tests to make sure your heart is working well:

- *An echocardiogram will check how well your heart is functioning. This test is an ultrasound of the heart.*
- *Cardiac catheterization will properly evaluate the pressure in your lungs. Your doctor will put a catheter into the blood vessel that goes into your heart and your lungs. This test is used to diagnose and treat pulmonary hypertension.*

LUNG CANCER

People with ILD have a higher chance of having lung cancer. Researchers are still studying the connection between lung scarring and lung cancer.

The things that may cause ILD (like cigarette smoke or being around chemicals) might also cause cancer. Your body might not be able to repair your lung tissue as well as it should if your lungs are inflamed or damaged.

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

GERD is also called acid reflux. Stomach acid goes backward into the tube that connects your throat and stomach (“esophagus”). This causes a burning feeling in the chest.

Doctors think that GERD may cause ILD or lead to the progression of lung disease, but this is not proven. What doctors do know is that GERD is more common in people with IPF.

Common symptoms of GERD are:

- *Heartburn*
- *Acidic or metallic taste in the throat*
- *Aspiration (fluid goes into your lungs)*
- *Regurgitation of food or drinks (undigested food goes back up the throat and into the mouth)*
- *Chest pain*
- *Cough*

People with ILD may have GERD without showing any symptoms. More research is needed to understand GERD in people with ILD.

If you have heartburn or other GERD-related symptoms, let your doctor know. Your doctor may be able to give you medications or suggest simple lifestyle changes.

Coexisting Conditions

ILD affects your lungs, but it can cause problems in other parts of your body, too. Your doctor will check and monitor your other organ functions regularly. Treating other conditions can help you and your doctor manage your ILD.

RESPIRATORY SYSTEM

OBSTRUCTIVE SLEEP APNEA

Excess soft tissue around the neck can block your breathing when you sleep. This can lower your oxygen level and keep you from having restful sleep.

LUNG CANCER

Scar tissue in ILD can turn into cancer in a small number of people with ILD. Usually, they have been exposed to some harmful substance that causes ILD (like cigarette smoke or asbestos).

PULMONARY HYPERTENSION

This type of high blood pressure occurs in the blood vessels of the lungs. It is caused by ILD or collagen vascular disease (long-term low oxygen levels in the blood).

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

Changes in the airways or tissues of the lungs make it hard to breathe. Common causes are emphysema, chronic bronchitis, or asthma. These conditions can coexist with ILD.

PULMONARY EMBOLISM (PE)

Blood clots in the lungs can happen suddenly. They often start in the legs or the lung vessels. Multiple clots can cause shortness of breath and pulmonary hypertension.

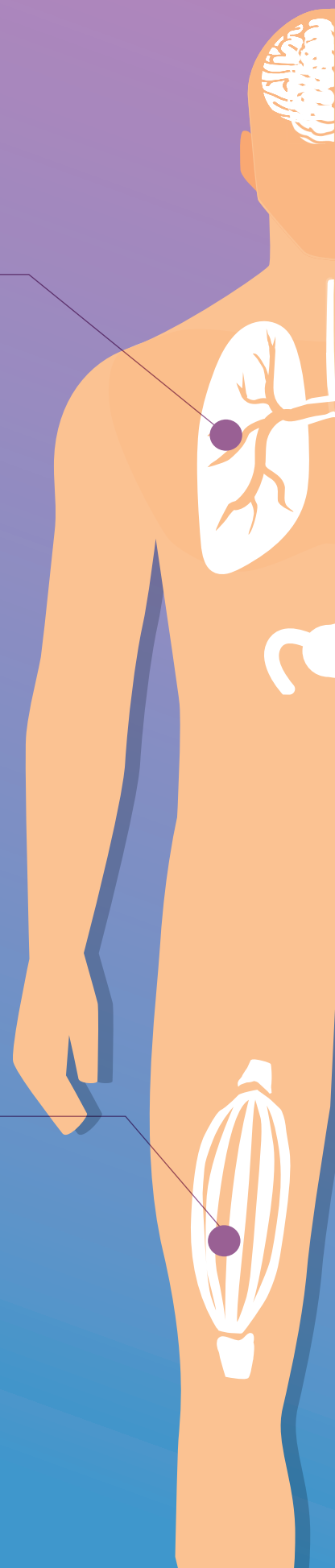
MUSCULO-SKELETAL SYSTEM

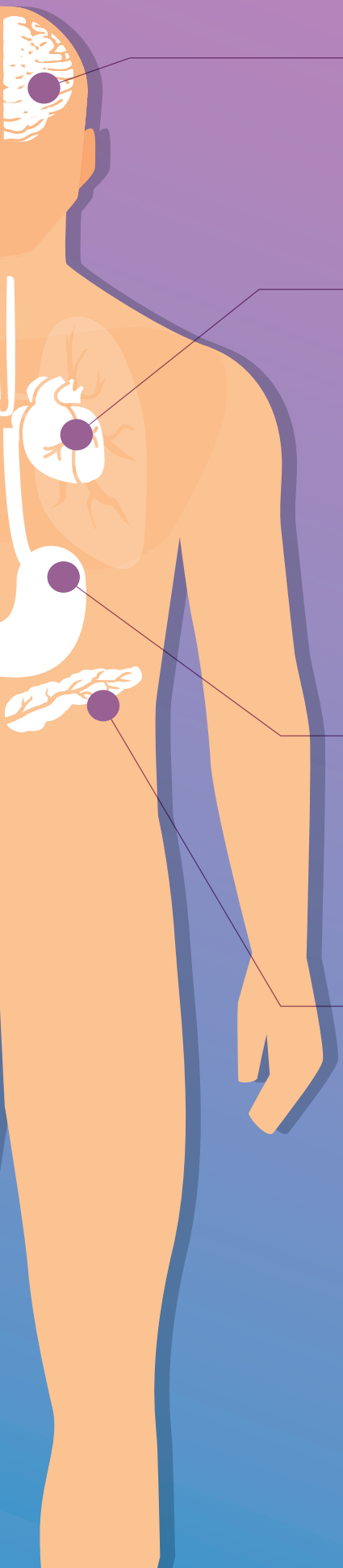
SARCOPENIA

This is mainly loss of muscle over time. In patients with ILD, it is caused by low blood oxygen, lack of exercise, long-time steroid use, inflammation, or malnutrition. Pulmonary rehabilitation and exercise can reduce loss of muscle.

DECONDITIONING

This loss of muscle strength and endurance can be caused by a lack of physical activity.





NEUROLOGIC SYSTEM

DEPRESSION / ANXIETY

A constant feeling of sadness that interferes with your usual routine, sometimes with anxiety and fear. Mental health disorders can have various symptoms.

CARDIOVASCULAR SYSTEM

CONGESTIVE HEART FAILURE

When the heart is under too much stress, it does not pump blood well to the body or the lungs. Low oxygen levels in the blood in people with ILD can stress the heart by making it work overtime. This can cause congestive heart failure.

CORONARY ARTERY DISEASE

Inflammation in patients with ILD can cause fat and plaque to build up in the blood vessels that supply the heart. This makes you more likely to have a heart attack or stroke.

DIGESTIVE SYSTEM

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

Stomach acid goes backward into the esophagus, causing a burning sensation in the chest. This acid can flow into the lungs and cause inflammation, which worsens ILD. It can also cause a constant cough due to irritation of the throat.

ENDOCRINE SYSTEM

DIABETES MELLITUS (DIABETES)

The body becomes less sensitive to insulin (the hormone that regulates blood sugar control), and there is too much sugar in the blood.

You can have one or more of these conditions along with your ILD.
If you have symptoms or are worried about having any of these conditions, talk with your doctor.

DEPRESSION AND ANXIETY

People with ILD live with their condition for a long time. This can be hard to handle. Some people with ILD are always short of breath and cough most of the time. This is stressful and can affect mental health.

One in every 4 people with ILD has depression or anxiety. Depression is a constant feeling of sadness that keeps you from doing your usual routine. People with anxiety feel worried or afraid almost all the time.

If you feel sad for a long time or worry a lot about your ILD, let your doctor know. Your doctor may be able to give you medication to help you feel better. Your doctor can also help you find other ways to cope with your condition.



DISEASE MANAGEMENT

Living with ILD can be difficult at first. There are things you can do to help manage your disease. Living a healthy lifestyle and following your doctor's recommendations will help improve your quality of life.

Start Pulmonary Rehabilitation

Your doctor may recommend pulmonary rehabilitation. This supervised exercise program is for people with lung disease. It can help you learn to breathe better so you can still do your normal activities and hobbies.

Rehabilitation helps your breathing muscles gain strength. It also teaches you ways to control your breathing. You will probably need pulmonary rehabilitation a few times a week for up to three months. If you practice the exercises at home, you can maintain your progress.

If you have been in the hospital recently, your doctor may suggest a pulmonary rehabilitation program when you leave the hospital.

Manage GERD

Your doctor may tell you to change what you're eating to control acid reflux, or GERD. Other lifestyle changes might be recommended. Your doctor may also give you medication to help control acid reflux.

Get Tested for Sleep Apnea

ILD is associated with sleep apnea, a sleep disorder. If you have any of these symptoms of sleep apnea, let your doctor know. You'll need an appointment to be tested if you:

- *Stop breathing or choke during sleep*
- *Snore*
- *Have daytime headaches*
- *Feel sleepy during the day (daytime sleepiness)*

Remember, you can live a fantastic life with ILD. Talk to your doctor about ways that you can continue to do the things you love, like traveling, playing sports, and cooking.

Things you can do to keep healthy with ILD:



If you smoke, quit!

Avoid secondhand smoke, vaping, and inhaled substances that could hurt your lungs

Exercise regularly

Maintain a healthy diet and weight



Take your medication as prescribed

Go to your follow-up visits with your doctor

Keep up with all lab, radiology, and lung function tests



Use oxygen therapy the way your doctor has prescribed



Get your flu vaccine every year

Get vaccinated for pneumonia

Control acid reflux/GERD

Treat sleep apnea for good sleep



Follow CDC guidelines regarding quarantine, handwashing, and social distancing

Use face masks during pandemics

MEDICATION OPTIONS

The medication used to treat your ILD depends on the cause of your disease, if there is one. Your doctor will assess all potential risks and benefits and recommend the medication that is best for you.

No medication will cure your ILD, but taking it will help keep your ILD from getting worse. Medication also helps prevent more scar tissue from forming in the lungs. This improves your lung function and your quality of life.

Medications for idiopathic pulmonary fibrosis (IPF)

Two medications are FDA approved to treat IPF:

- *Nintedanib (Ofev®)*
- *Pirfenidone (Esbriet®)*

Both medications help to slow lung tissue scarring, which preserves your lung function. Nintedanib (Ofev) may also lower your chances of having a serious and quick worsening of your IPF (IPF exacerbation).

Your doctor will need to perform blood tests to ensure that your liver is okay while you're taking these medications. The most common side effect of nintedanib (Ofev) is diarrhea. More serious but rare side effects can include liver injury, blood clots, or gastrointestinal perforation. You should not take either of these medications if you are pregnant or thinking of becoming pregnant. The most common side effects of pirfenidone (Esbriet) are nausea and GERD, or acid reflux.

Medications for other types of ILD

If you have ILD but not IPF, your medication options may be different. The type of medication depends on the type of ILD. Risk factors and other health conditions will also help your doctor decide what medication to prescribe.

Remember, these drugs will not cure your ILD. They will help improve your symptoms and maintain your lung function and overall health.

NINTEDANIB (OFEV)

Nintedanib (Ofev) has also been approved for patients with chronic fibrosing ILD with a progressive phenotype. This means that regardless of the underlying condition that has caused a person's fibrotic ILD, they may be eligible to benefit from this medication, just like patients with IPF.

CORTICOSTEROIDS (PREDNISONE)

Inflammation of the lungs is sometimes a problem with ILD. Corticosteroids can be very helpful for controlling it in certain types of ILD. Corticosteroids lower your body's immune response and reduce inflammation.

Steroids help decrease inflammation, but they will not get rid of scar tissue that has already formed in the lungs. It is important to treat inflammation before scar tissue forms.

Steroids have many side effects that you should review with your doctor. Because of these side effects, you might use them for only a short amount of time. If you need long-term treatment for ILD, your doctor will suggest a transition to a “steroid-sparing drug.”

MYCOPHENOLATE MOFETIL (CELLCEPT)

CellCept lowers your immune system by targeting a type of blood cell called a lymphocyte. CellCept is often used by people who have received an organ transplant. Some people with ILD may use it instead of corticosteroids because the side effects can be easier to manage.

While taking CellCept, your doctor will need to perform regular blood tests to check your blood cell counts. Your doctor will also tell you about other risks of taking this medicine. The most common side effect is diarrhea. Your doctor will want to watch you closely since you are at a higher risk for infections, lymphoma and skin cancer. You cannot take this medication if you are pregnant or thinking about becoming pregnant.

AZATHIOPRINE (IMURAN)

Imuran lowers the immune system by suppressing protein, DNA, and RNA production in cells. This medication is often used by people who have had an organ transplant or who have an autoimmune disease (such as rheumatoid arthritis, Crohn's disease, or lupus). Some people with ILD may also use it in place of corticosteroids.

While taking Imuran, your doctor will need to perform blood tests to check your blood cell counts. Your doctor will also tell you about the other risks of taking this medicine.

CYCLOPHOSPHAMIDE (CYTOXAN)

Cytoxan lowers your immune system faster than the medications listed above do. It is usually used to treat cancer. This medication may be given as an intravenous solution or as a pill.

Cytoxan is used to treat ILD that is rapidly getting worse or that does not get better with the usual medications. Sometimes it is given when a patient is hospitalized with a complication from their ILD.

Possible side effects of Cytoxan include infection, bone marrow suppression, bladder inflammation, and infertility.

OXYGEN THERAPY

Oxygen therapy is the use of supplemental oxygen prescribed by your doctor as a medical treatment. Your doctor may prescribe oxygen therapy if your blood oxygen levels are too low.

People with more severe lung scarring may need supplemental oxygen for the rest of their lives. People with reversible ILD may only need oxygen supplementation for a short amount of time.

Why do I need oxygen?

Every cell in your body needs oxygen. It is the job of your lungs to properly transmit the oxygen that you breathe in and transport it into your bloodstream. Having ILD makes it more difficult for your lungs to do this because of scarring and inflammation.

Oxygen levels can drop in patients with lung disease. This makes it hard for oxygen to effectively reach all your organs. Supplemental oxygen therapy helps ensure that your organs receive the oxygen they need.

Oxygen also helps protect your heart. If your lungs aren't working well, there is more stress on your heart. Sometimes ILD may cause *pulmonary hypertension*, a type of high blood pressure that affects your lungs. If this type of high blood pressure in your lungs goes untreated, it can eventually lead to heart failure.

When should I use oxygen?

Your doctor will test your oxygen levels at rest and while active. If your blood oxygen level is low, your doctor will prescribe oxygen.

Specific oxygen levels can qualify you to receive oxygen from your insurance company.

Use your oxygen exactly how your doctor tells you to use it. Your doctor will probably tell you to use oxygen 24 hours a day.

How will I get the oxygen I need?

Your insurance company will cover the costs of your oxygen therapy if you qualify for oxygen and your doctor gives you a prescription. You will need a face-to-face visit with your doctor to do an oxygen evaluation.

An oxygen evaluation checks your oxygen level at rest and with exertion (physical effort), both with and without supplemental oxygen. You will repeat this test at least once a year—sooner if your condition worsens. Your condition may get worse if you get a lung infection or your disease advances.

Your prescription will tell you how much oxygen you need. This is called the “oxygen flow rate.” The flow rate may be different for when you are resting, exercising, or sleeping.

Once you decide which oxygen company you want to use, your doctor will send them your prescription. The company will come to your home to deliver and set up your home oxygen machine. The oxygen company will give you the correct type and amount of oxygen that your doctor prescribes.

What are the dangers of oxygen?

Oxygen therapy is safe, and oxygen tanks are well built. Don't worry! Your oxygen tank won't explode unless it is near very high heat. But you should keep your oxygen tank at least 6 feet (about two-and-a-half arm lengths) away from open flames.

You should not smoke while on oxygen therapy. If you do, your hair and clothes may catch on fire. People who have normal blood oxygen levels should not use your oxygen therapy machine.

What is the best type of oxygen for me?

Many types of oxygen devices can be prescribed. The best choice will depend on what you need, how much it costs, and what your insurance will cover.

LUNG TRANSPLANT

A lung transplant for ILD can help you have a better quality of life, but it is a serious surgery. A diseased lung is replaced with a healthy lung from a donor. In the US, more than 2,700 lung transplants are performed every year.

Successful lung transplants can help people live longer. However, the prognosis for lung transplants is not as good as for other organ transplants. About half of those who receive a lung transplant are alive after 5 years, but many of them can live longer.

Having a lung transplant involves important risks and commitments:

- You may need to stay in the hospital for some time after the transplant.
- In some cases, life-threatening complications can occur from surgery.
- You will take medication for the rest of your life to prevent rejection of the transplanted lung.
- The medication may make you more likely to get infections and can have other side effects.
- Frequent visits with your doctor and transplant team will be necessary.
- If you don't live near a transplant center, you will have to move close to one for a year or two.

Deciding to have a lung transplant is a serious decision. You will need to talk it over with your family and caregivers. It takes a lot of time and effort to get evaluated for a lung transplant. You will have a lot of doctor appointments before and after the surgery.

Many factors determine whether a person is a good candidate for a lung transplant. Each lung transplant program in the US has its own criteria and transplant team. The transplant team has its own way of deciding which people are good candidates for a transplant.

Some programs will perform lung transplants on people over age 64 and on those who need the help of a machine to breathe (ventilator). For more information, visit the website of the transplant centers you are interested in.

Your doctor may consider referring you for a lung transplant if:

- You have shortness of breath at rest or with small amounts of activity
- You need more oxygen than you previously required
- Your disease is worsening and there are no other medications or treatments available
- Your other organs (liver, kidneys, heart) are working well
- You do not have any other major health problems
- You are mentally strong enough to have a lung transplant
- You can follow your doctor's orders exactly
- You have friends and family to help you before and after your transplant
- Your doctor says you have 2 years or less to live at your current health condition and on your current medication

You are not a good candidate for a lung transplant if:

- You have infections that are untreated or do not get better with treatment
- You have tuberculosis
- You have had cancer (except non-melanomatous skin cancer) within the past 2-5 years
- You have a mental health condition that is not being controlled
- You don't always take your medication or follow your doctor's orders
- You tend to miss your doctor appointments
- You don't have family or friends to help you before and after the transplant
- You have severe problems with your heart, kidneys, or liver
- You smoke or you quit smoking less than 6 months ago
- You currently use drugs or drink alcohol
- You have a bleeding disorder that cannot be controlled or cured
- You have a chest or spinal deformity
- You are extremely overweight or obese

In general, a lung transplant may be riskier for you if:

- You have osteoporosis, a condition that causes your bones to be weak and break easily
- You are of older age and frail
- You've had a part of your lung removed or you've had a procedure called a pleurodesis in the past
- You have an infection that does not get better with treatment
- You have HIV or hepatitis B or C
- You take high doses of steroids
- You have problems with your esophagus and severe acid reflux
- You don't eat enough or eat balanced meals, so your body doesn't have the vitamins and minerals it needs for proper wound healing

HELP YOUR DOCTOR EVALUATE YOU FOR ILD



TIP:

PRINT OR REMOVE
PAGES 41–43
AND FILL THEM
OUT TO TAKE TO
YOUR DOCTOR
APPOINTMENT.

Working closely with your doctor is very important when managing your disease and symptoms. Your doctor will need to ask you a lot of questions at your initial appointment. The answers will help your doctor know how best to treat your lung disease.

Sometimes this process can be overwhelming. You may want to answer these questions ahead of time to make it easier.

WORK HISTORY

List all the jobs you've had—especially if you worked in construction or farming, or with dust or metals. Mention if you've worked with birds or animals.

Have you been worried about anything you were exposed to at a job?

Have you worked at any jobs where powdered asbestos or any other particles were in the air?

Did you ever cut asbestos or work in jobs where asbestos was in the air (like construction, shipbuilding, or factory work)?

Did you ever do sandblasting or rock cutting?

Have you been around water damage or mold at work?

LIFESTYLE

What are your hobbies (for example, woodworking or metal cutting)? Do you wear a mask?

What pets do you have?

Do you raise any animals (for example, poultry)?

Have you been exposed to any of these animals or animal products?

- Cockatiels or exotic birds
- Bird feathers
- Down or feathers in pillows, comforters, cushions, or jackets

Where have you traveled in the past 5 years?

Does your house or basement have water damage?

Have you been exposed to mold at home?

Do you use hot tubs or humidifiers?

PERSONAL AND FAMILY HEALTH HISTORY

Bring a list of the medications you're taking or have taken in the past.

Also bring a list of any over-the-counter medications, like vitamins, herbal medicine, or drugs you are taking.

Do you have any of these symptoms of autoimmune diseases?

- Bald patches in your hair
- Unexplained rashes
- Dry mouth or dry eyes
- Heartburn, frequent belching, reflux, or trouble swallowing
- A star-shaped spot on the skin
- Red, swollen, or tender joints in your fingers
- Scaly skin on your hands
- Shoulders or hips that are getting weaker and weaker
- Change in eyesight
- A history of strokes or blood clots
- Blood in urine or stool

Does anyone in your family have pulmonary fibrosis or an unexplained lung disease?

Did anyone in your family have their hair go gray early (in their teens or twenties)?

Does anyone in your family have scarring of the liver (cirrhosis), unexplained low red blood cells (anemia), or trouble making healthy blood cells (myelodysplastic syndrome)?

QUESTIONS TO ASK YOUR DOCTOR

TIP:

REMOVE OR PRINT THIS PAGE AND TAKE IT TO YOUR DOCTOR APPOINTMENT.

An ILD diagnosis can be scary at first. Having the right information will help you to feel in control of your condition. Here are some questions you may want to ask your doctor to better understand how to live with ILD.

● ASK ABOUT YOUR ILD DIAGNOSIS:

- What type of ILD do I have?
- Can I get a second opinion to confirm my diagnosis?
- Will I need to have a lung biopsy?
- What caused my ILD?
- How severe is my disease?
- How will ILD affect my everyday life?
- Will my disease get worse?
- How will I know if it's getting worse?
- Will I need oxygen?
- What kinds of tests will I need to monitor my ILD?
- How often will I have to go to the doctor?

● ASK ABOUT TREATMENT AND SUPPORT:

- What can I do about my persistent cough?
- Where can I find a support group?
- Will pulmonary rehabilitation help?
- Are there medications for my ILD?
- Do the medications for my condition have side effects?
- Can I have a lung transplant?
- Do I need palliative care now? Will I need it in the future?
- Are there any clinical trials that I could participate in?

● ASK ABOUT OXYGEN THERAPY:

- Should I use my supplemental oxygen all the time or just sometimes?
- What oxygen flow rate should I use?
- Can I travel with my supplemental oxygen?
- Should I be checking my oxygen saturation levels at home?

● ASK ABOUT HOW TO STAY HEALTHY:

- What should I do if I get a cold, the flu, or pneumonia?
- How will my doctor monitor my ILD?
- What health problems might arise with my condition?
- What should I do if my symptoms get worse suddenly?



**If you're
struggling, don't
be afraid to ask
for help.**

SUPPORT

Managing ILD may seem hard. Some of the best information to help you manage your condition can come from others living with ILD.

Support Groups

Local and online support groups can be a great help as you navigate life with ILD. Talking can help reduce stress and anxiety about your condition. Communication can ease the feeling of loneliness that often comes with a new diagnosis. Sometimes it's just good to be around people who know what you're going through every day.

Ask your health care team if a local support group exists. Join one, or set one up. Check with area medical centers and hospital systems for resources.

Support is also available online.

PF Warriors

www.pfwarriors.com

This online, international support group was formed by volunteers for people with pulmonary fibrosis (PF), ILD, and other lung diseases. Every month, there are in-person and online meetings.

Topics include:

- Diet
- Medication to treat lung disease
- Exercise
- Living a better life with lung disease

PF Warriors on Facebook

<http://www.facebook.com/pfwarrior>

The PF Warriors Facebook page shares websites and news articles relevant to PF, ILD, and other lung diseases. It also provides up-to-date information on PF Warriors and highlights current projects.

Pulmonary Fibrosis Foundation Support Groups

<https://www.pulmonaryfibrosis.org/life-with-pf/support-groups>

The Pulmonary Fibrosis Foundation (PFF) has local, virtual, and telephone-based support groups for people living with lung disease and their caregivers. The groups talk about worries or problems in living with lung disease. The goal is to help people live a better life.

PFF's support group resources include:

- **PFF Voices:** A support group for people who have PF and ILD
- **PFF Caring Conversations:** A support group for family members and caregivers
- **PFF Support Group Leader Network:** This network helps to train support group leaders and teach them how to start their own support group. Every three months, the support group network holds teleconferences to support group directors.

Two companies that make medicines to treat IPF—Boehringer Ingelheim and Genentech—sponsor the PFF Voices and PFF Caring Conversations groups.

Medication Manufacturers

Information is available from the companies that make FDA-approved medications used to treat IPF.

These websites may help you understand more about ILD and related medications.

Boehringer Ingelheim makes nintedanib (Ofev): www.boehringer-ingelheim.com or <https://www.ofev.com>

Genentech makes pirfenidone (Esbriet): www.gene.com or www.esbriet.com

Pulmonary Fibrosis Foundation

<https://www.pulmonaryfibrosis.org/>

PFF is one of the top organizations for information on pulmonary fibrosis, a type of ILD. The PFF website has information on education, clinical trials, events, and a medical provider phone line.

Other resources include:

- **PFF Care Center Network:** *This network helps people find doctors and medical centers that are skilled in the diagnosis and treatment of PF.*
- **PFF Ambassador Program:** *This program gives people with PF, their families, and their doctors the chance to share what they know about PF and their personal stories about PF. PFF Ambassadors spread the word about PF and reach out to government officials to get support for PF research and the PF community. Ambassadors also speak to local PF support groups and at PF educational events.*
- **PFF National Meetings:** *Every two years, PFF hosts a national meeting. At the meeting, top scientists explain their latest research on PF. This meeting educates people with PF, caregivers, and medical teams (doctors, nurses, and therapists). The best part is that everyone gets to connect and plan ways to spread the word about and gain support for PF.*

More About Lung Disease

To learn more about lung health, lung disease, and ILD, visit these national and international organizations:

- **American College of Chest Physicians (CHEST):** chestnet.org
- **American Lung Association:** lung.org/
- **American Thoracic Society:** thoracic.org
- **British Lung Foundation:** blf.org.uk

ILD MYTHS



Here are some common myths about ILD and some facts to help you better understand your condition.

MYTH

If I start using oxygen, I'll gradually have to use it more and more.

FACT

Your body is like a car, and oxygen is like gas. If your tank is almost empty, you fill it with gas. Putting in gas won't make your car need more gas each time. And, using oxygen won't make your body need more oxygen.

Your doctor will check the oxygen levels in your blood using a machine called a pulse oximeter. This device slides over your finger. The doctor will test your oxygen levels at rest and while you're walking. If your blood oxygen level is low, your doctor will prescribe oxygen therapy. Your prescription will tell you how much oxygen you will need when you are sitting or walking.

At follow-up appointments, your doctor will check your blood oxygen levels again to see if your oxygen requirements have changed. If you don't use your oxygen the way your doctor tells you to, the pressure could increase in the vessels of the lungs (pulmonary hypertension). If that happens, it can put excess strain on your heart and lead to heart failure.

Oxygen not only helps you breathe better and exercise more but also helps your lungs and heart work optimally. Don't be afraid to start using oxygen. It will help you keep your usual routine. Oxygen will also help prevent other health problems.

MYTH**FACT**

Pulse-flow portable oxygen concentrator devices deliver the same amount of oxygen as continuous-flow devices because the flow rate settings are the same (2 liters per minute, 3 liters per minute, etc.)

The number on a pulse-flow concentrator does not mean the same thing as the number on a continuous-flow oxygen canister.

Pulse-flow oxygen means that oxygen is flowing only when the patient takes a breath. Continuous-flow oxygen means that oxygen is flowing through the tubing constantly, causing some oxygen to be wasted when the patient is not breathing.

Although pulse flow helps to conserve oxygen in the device, it may not be suitable for all patients who need to use oxygen. Some people are very short of breath and have very poor lung function. They need a continuous-flow setting, whereas others may be comfortable on a pulse-flow setting.

Many portable devices only allow for pulse-flow oxygen. You should discuss the appropriate options for portable and stationary oxygen with your doctor and oxygen supply company.

The therapies for IPF—nintedanib (Ofev) and pirfenidone (Esbriet)—will help with my shortness of breath, and I'll feel better.

It may be disappointing, but the two medications used to treat IPF will not make your shortness of breath go away. Nintedanib (Ofev) and pirfenidone (Esbriet) slow the worsening of pulmonary fibrosis by half. They may also prevent serious sudden worsening or exacerbation of IPF. They cannot cure the disease.

Taking these medications means that you'll be able to keep your current lifestyle and your usual routine for longer than if you didn't take them.

Lung transplantation will cure my ILD.

Lung transplants can cure most ILDs. However, in rare cases, the disease can come back in the new, transplanted lung.

My oxygen saturations are normal as long as I'm not feeling severely short of breath.

If you feel short of breath because of low oxygen, your blood oxygen levels are already too low. Please don't use shortness of breath to figure out whether your blood oxygen levels are normal. They aren't necessarily related.

If you need oxygen and you don't feel short of breath, it may mean that your other organs (such as the heart) are working too hard to compensate. Your doctor will check your blood oxygen levels during your visits. If you want to check your levels at home, buy a finger pulse oximeter at your local pharmacy. Your doctor can teach you how to use it at home.

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This patient education guide was made possible by the Popovich Endowment and supported by a grant from Boehringer Ingelheim Pharmaceuticals.