Idiopathic Pulmonary fibrosis

Lung Transplant

Sometimes, with severe IPF, the only way to improve quality of life or increase survival may be a lung transplant. However, the decision to have a lung transplant must be made carefully. On average, only 50% of patients live for 5 years. Not all patients qualify for lung transplantation, depending on age or other medical problems.

Lung Infections

Due to lung damage, lung infections can cause serious problems for patients with IPF. If you notice the following symptoms:

- Worsening cough
- Change in the color or amount of sputum produced
- Fever

call your health care provider immediately.

Coping With Cough

A nagging, dry cough is one of the most common symptoms with IPF. Although cough is often caused by IPF, there are other things that can make it worse including:

- Postnasal drip
- Acid reflux disease
- Allergies and pollutants
- Cigarette smoke
- Some medications, such as beta-blockers or angiotensin-converting enzyme (ACE) inhibitors, for blood pressure control.

Nasal sprays, cough medicines, and cold remedies may help control cough.

Leg Swelling

IPF may cause swelling in the legs because lung scarring makes it difficult for the heart to pump blood through the lungs. If this happens, fluid can back up, causing the legs to swell. This is called right-sided heart failure and can get worse if blood oxygen levels are too low. If you have swelling in your legs, call your health care provider.

Travel

A loss of oxygen pressure during air travel often makes flying very difficult with IPF. Ask your health care provider if oxygen is needed during the flight. Airline rules for oxygen use often change. Contact the airline, and let them know that you will need oxygen during the flight (they may charge a fee). Also, make sure you can get oxygen, should you need it, when you get off the plane, particularly travelling to a high altitude area.

Life Support

If your lung disease becomes very severe, you may need life support (ventilation) to keep you alive. Newer forms of mask ventilation, called bi-level pressure ventilation (BiPAP), may help for a short time. However, most patients with IPF, who are placed on a ventilator, cannot be taken off. You may need this type of ventilation for the rest of your life. It is up to you to decide if you want this treatment.

End-of-Life Issues

Talking about end-of-life issues is not easy. However, it is important that you let your family and your doctor know how you feel. There are legal forms that you can fill out that state what treatments you want or do not want, should you become too ill to make your wishes known. You should also choose someone as a power of attorney. This person can make decisions about your treatment

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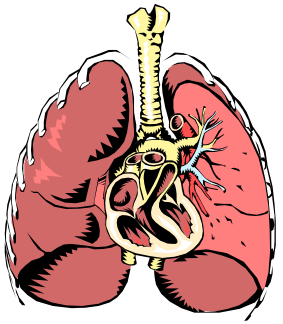
Idiopathic Pulmonary Fibrosis



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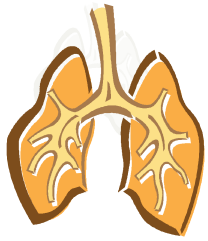
Idiopathic pulmonary fibrosis

Idiopathic pulmonary fibrosis (IPF) is a buildup of scar tissue in the lungs. The scar tissue damages the lungs, making it hard for oxygen to pass through the lungs to the rest of the body. Lack of oxygen can result in serious health problems and even death.



"Idiopathic" means the exact cause of scarring cannot be found. The scarring probably starts from something that injures the lung. Scar tissue increases as the lungs try to repair the injury. In time, so much scarring forms that patients have problems breathing. IPF usually worsens over time; however, while some patients get sick quickly, others may not feel sick for years. Even though IPF has no cure, certain treatments may slow down lung scarring.

Symptoms

Shortness of breath
Cough (nonproductive/dry)
Fatigue and weakness
Chest pain or tightness in the chest
Loss of appetite
Weight loss

Diagnostic Testing

Pulmonary Function Tests: measure how well the lungs are working and the severity of scarring.
CAT scan: Provides a detailed image of the lungs, and determines the degree of scarring
Blood tests: determine if infection is present and/or amount of oxygen is in the blood.
Bronchoscopy:A fiber optic scope inserted through the nose or mouth into the lung allows a small piece of lung tissue to be biopsied and tested
Thoracoscopic biopsy: A larger tissue sample is obtained via a surgical procedure in which small incisions are made in between
the ribs. Performed by a thoracic surgeon,

Treatment

Lung scars cannot be surgically removed and cannot be completely dissolved with medication. However some behavior modification and preventative measures may help:

Smoking Cessation

Cigarette smoke not only damages the lining of the lungs, it can also make you more likely to get a lung infection. Experts agree that you should stop smoking.

Supplemental Oxygen

As lung scarring gets worse, many patients need extra oxygen to go about their daily lives without getting short of breath. In later stages of IPF, oxygen may be needed even while sleeping or resting. Oxygen is not addictive, do not worry about using it too much. To monitor oxygen levels,
a small, easy-to-use device called a pulse oximeter is used initially to determine how much oxygen is needed and later to assure this amount remains adequate.

Vaccinations

Prevent severe infection
Influenza-annually each fall
Pneumonia-every 5 years until age 65, then once after age 65.
Tetanus-every 10 years with one booster given as a combination Tetanus/Pertussis before age 65

Exercise/Pulmonary Rehab

Regular exercise can help patients with IPF. Exercise keeps breathing muscles strong and increases energy because healthy muscles need less oxygen to perform work.
Ask your health care provider about a Pulmonary Rehabilitation (exercise) program.

Nutrition

Many patients with IPF lose weight because of the disease. Excessive weight loss may cause breathing muscles to become weak. . A well-balanced diet helps the immune system fight off infections

Medications

A variety of drugs can be used to treat IPF. Often given for long periods of time (3 to 6 months or longer), regular check-ups
are needed to see how well they are working.
Corticosteroids- a common treatment for many lung conditions. However, corticosteroids have many side effects and do not
usually work well for patients with IPF. In fact, they may do more harm than good. Most experts agree that these drugs should only be used for patients who have had improvement while taking them.
Cytotoxic and immune suppressing drugs:Bone marrow problems may occur so white blood cell counts must be watched closely to avoid levels becoming too low The efficacy of these drugs for IPF has never been well established.
Other medications;Colchicine may help stop scar tissue from forming; however, it has not been shown to be better than the more commonly used drugs. N-acetyl cysteine (an antioxidant), may have some benefit, but experience with the drug is limited, and more studies are needed. Many experts think that acid reflux is an important factor in IPF. Patients with this condition should take medicines to help control it.
Experimental treatments/clinical trials:many FDA-approved studies, called clinical trials, are being performed to find new medications for IPF. In a clinical trial, a new drug is compared to a "placebo" (an inactive medicine, such as a sugar pill). Participants do not know which treatment they are getting, the experimental