

## Pulmonary fibrosis

Pulmonary fibrosis is a chronic disease that causes scarring and inflammation in the lungs. This scar tissue replaces healthy tissue, slowly reducing a person's ability to absorb oxygen. It also causes lungs to become stiff, making breathing difficult.

It can be difficult to predict how pulmonary fibrosis will progress over time in an individual. It can be a mild disease causing few symptoms or it can be severe and possibly fatal.

### What causes pulmonary fibrosis?

Pulmonary fibrosis is a complicated illness with many possible causes:

- Some occupations, mainly those that involve working with organic substances, asbestos, silica dust, ground stone or metal dust
- Certain chronic conditions (e.g. lupus, rheumatoid arthritis, scleroderma)
- Infections (e.g. tuberculosis, pneumonia)
- Certain strong medications (e.g. antibiotics, chemotherapy)
- People who smoke or former smokers are more likely to develop pulmonary fibrosis than people who have never smoked

The causes of Idiopathic Pulmonary Fibrosis (IPF) are unknown. Idiopathic means "of unknown origin". Some researchers believe that IPF may result from an autoimmune disorder, a condition in which the body's immune system attacks its own tissues.

### What are the symptoms of pulmonary fibrosis?

Breathlessness is the most common symptom of pulmonary fibrosis. A dry hacking unexplained cough, usually lasting longer than eight weeks, is another common symptom. Other symptoms may include chest pain, loss of appetite and energy, and joint and muscle pain.

### How is pulmonary fibrosis diagnosed?

Recognizing pulmonary fibrosis can be a challenge since the symptoms of breathlessness and cough are similar to common lung diseases, such as asthma.

A chest X-ray and CT-scan can help with the diagnosis. Pulmonary function testing (e.g. spirometry) determines how well your lungs function. Samples of cells or tissue from inside the lungs can be sent to a laboratory for analysis.

### What is the treatment for pulmonary fibrosis?

Although research for effective drugs is on-going, unfortunately, currently there is no cure for pulmonary fibrosis. When there is a known environmental cause of the pulmonary fibrosis, removal of such cause is the first priority.

For IPF, when the cause is unknown, treatment is targeted at treating the inflammation that occurs in the lungs. This may help prevent the scar formation and the progression of the disease, as well as lessen the symptoms of cough and shortness of breath.

Pulmonary rehabilitation and education programs can teach patients how to breathe more efficiently and to perform their activities of daily living with less breathlessness. Sometimes supplemental oxygen therapy is required in order to treat breathlessness. Early treatment of chest infections is important. Lung transplantation is also a possibility. Smoking must be discontinued since it will aggravate the shortness of breath.

To learn more about pulmonary fibrosis and available resources,  
Call the **Ontario Lung Association's Helpline at 1-888-344-LUNG (5864)** or visit  
[http://www.lung.ca/diseases-maladies/a-z/pfibrosis-fibrosep/index\\_e.php](http://www.lung.ca/diseases-maladies/a-z/pfibrosis-fibrosep/index_e.php)