HOW IS THE DISEASE DIAGNOSED?

Certain clues can help diagnose the disease:
A chest x-ray reveals opaque abnormalities that are small and diffuse, somewhat like a frosted window. During pulmonary function tests, we notice a decline in lung capacity. A blood test taken from the radial artery at the wrist will demonstrate respiratory insufficiency that is characterized by a significant decrease in oxygen in the blood and an increase in waste gas, i.e. CO2. A lung biopsy by means of a bronchoscopy and bronchoalveolar lavage will allow cells inside and around the alveoli to be collected and examined, in order to establish a diagnosis.

TREATMENT

If the cause is known, we treat the underlying disease or problem inducing fibrosis. In the case of mineral dust inhalation, workers must avoid all contact with these substances.

Treatment begins with the use of cortisone, a potent anti-inflammatory drug. This medication is used in an attempt to decrease the inflammatory reaction that affects the lung, thereby possibly preventing the onset of scar tissue or fibrosis, and thus slowing the destruction of the lung.

If there is no improvement, we may offer other treatments, but there is currently little evidence indicating that they are superior to the use of cortisone.

If the drug therapy for pulmonary fibrosis rarely provides beneficial results, there are nevertheless many types of alternative treatments to relieve the shortness of breath that accompanies fibrosis:

- **First, patients must stop smoking. Pulmonary fibrosis and especially asbestosis increase the risk of cancer;**
- **Patients should enroll in educational programs for disease management and respiratory rehabilitation in order to increase their exercise tolerance, improve tissue oxygenation and maximize the effectiveness of the healthy tissue;**
- **In some cases, oxygen therapy is recommended;**
- **Treatment of respiratory infections should be initiated promptly. Vaccination against influenza and pneumococcal pneumonia is strongly recommended;**
- **A lung transplant may be proposed if the patient's condition warrants such surgery.**

Research is still required to discover effective ways to combat this disease and to improve the quality of life of those affected by it.

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Help us help you!

Donations made during our various fundraising campaigns, bequests, commemorative gifts and planned giving are also practical ways of supporting the Quebec Lung Association.

By calling our toll-free number, you can speak to a professional specializing in respiratory health. You can ask all your questions and get advice about your respiratory health.

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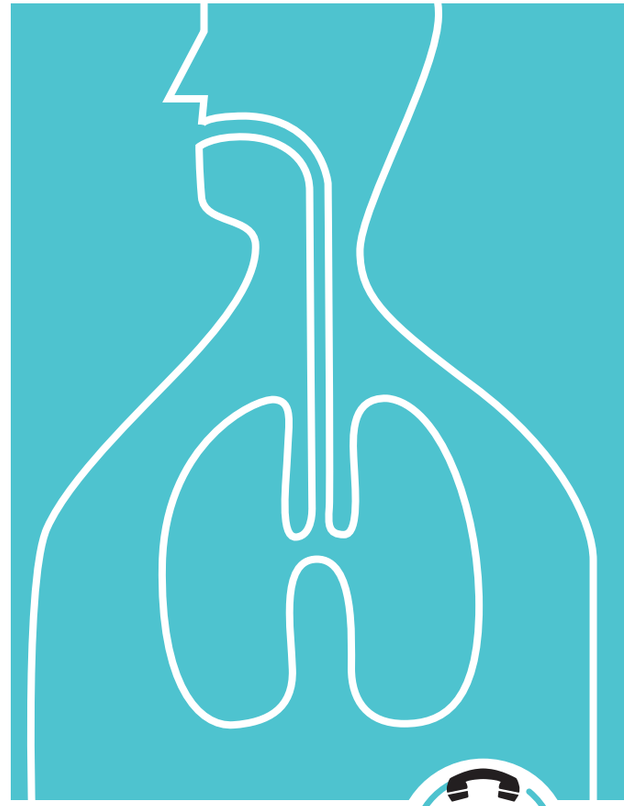
PULMONARY FIBROSIS



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PULMONARY FIBROSIS

Pulmonary fibrosis is a chronic disease with many potential causes (over 140 known causes). It is induced by inflammation of the alveoli, leading to thickening and hardening of the alveoli and interstitial tissues of the lungs, which eventually shrink, becoming smaller and more rigid.

Many diseases can trigger pulmonary fibrosis, but in half the cases the cause remains unknown.

If the disease is of unknown origin, it is termed idiopathic pulmonary fibrosis or diffuse pulmonary fibrosis.

If the exact cause can be identified, it is referred to as pulmonary fibrosis secondary to this cause: for instance, certain medications or certain rheumatic diseases.

Similarly, because of their profession, some workers may come into contact with substances that can cause fibrosis. The most common in Quebec are the fibers of asbestos and silica dust. When these are inhaled and damage the alveoli, they provoke pulmonary fibrosis. Less commonly, inhaled organic substances, such as moldy hay, can cause pulmonary fibrosis in farmers.

In addition, many powerful drugs like those used to treat certain heart conditions or cancers, radiation therapy at the thorax or the inhalation of industrial gases in high concentrations, can also cause the formation of pulmonary lesions like fibrosis.

Idiopathic pulmonary fibrosis affects both men and women and is usually diagnosed between the ages of 40 and 70. There is no connection between the disease and the ethnic or geographic origin of affected patients. Children can also develop idiopathic pulmonary fibrosis.

Fibrosis may be benign and only display a few symptoms, but it could gradually progress and lead to respiratory failure and death. If this happens, the impact on the autonomy of the affected individual may be significant. Even everyday tasks can become difficult to accomplish when the disease is advanced. It is essential for people with pulmonary fibrosis to stay physically active while still conserving their strength, slowing their pace, knowing their limitations and prioritizing their activities.

LE MÉCANISME DE LA MALADIE

“De nombreuses maladies peuvent être à l’origine de la fibrose pulmonaire...”

Inflammation of the lungs will usually occur for unknown reasons. This inflammation results in the invasion of white blood cells in the alveoli, which are small air sacs where oxygen is transferred into the blood.

If this inflammation persists in the alveoli, it will cause a thickening and hardening of the alveoli walls.

The blood vessels of the lungs are separated from the alveoli by solid walls called interstitial tissues. This interstitial tissue allows oxygen to enter the blood while carbon dioxide (CO2) is expelled in the opposite direction to the exterior of the lungs.

By thickening and hardening the interstitial tissue, fibrosis reduces the lung’s capacity to enrich the blood with oxygen and makes removal of the carbon dioxide (CO2) contained in the blood much more difficult. This leads to respiratory failure.

SYMPTOMS

Generally, the disease begins to manifest itself in the early fifties. Two symptoms make diagnosis difficult because they are present in several lung diseases and are therefore not very specific:

- Shortness of breath during physical exertion that steadily worsens
- Dry cough

These two symptoms may signal the onset of fibrosis, but other warning signs may be present:

- Loss of endurance
- Loss of appetite
- Fatigue
- Weight loss

Thereafter, these symptoms may appear:

- Breathlessness while at rest
- Cyanosis (blue lips, nails and sometimes skin caused by insufficient oxygenation of tissues)
- Deformation of finger tips resembling drumsticks, or finger clubbing seen especially with asbestosis.

Pulmonary fibrosis can lead to serious complications such as pulmonary hypertension. This is due to contraction of the pulmonary artery caused by lack of oxygen. The pressure in the pulmonary artery increases the work of the right side of the heart. Over time, the heart weakens, causing right heart failure.