



INTERSTITIAL LUNG DISEASE

The purpose of this leaflet is to help you understand the unusual condition of interstitial lung disease. The name covers a number of different lung conditions, and is caused by inflammation and scarring which develops in the lungs. In most cases the cause of this is unknown.

What are the Symptoms?

The main symptom of interstitial lung disease is breathlessness. Initially, this may only occur after unusual exertion, such as running for a bus. As the condition progresses and the damage to the lung is more severe, you may become breathless after even minor physical effort such as showering. Usually there is no problem breathing while you sit quietly. The other main symptom is cough. This can be quite distressing and is often worse after physical exertion. The cough is usually dry, however some people produce a small amount of clear sputum or phlegm.

What causes this Disease?

This disease is the result of damage to the lungs caused by inflammation and scarring. In a few cases it is clear that this is caused by inhaling some foreign material to which the body reacts strongly. For example interstitial lung disease occurs in some pigeon breeders as a reaction to inhaling the fine dust from the bird aviary over a long period of time. Another group of patients do not appear to have inhaled any damaging material but have developed an internal body reaction which affects the lungs. For example, a number of patients with rheumatoid arthritis develop progressive lung scarring in addition to their damaged joints. Sarcoidosis (an immunological reaction in the lung) and several drugs may also result in interstitial lung disease. There are many people with interstitial lung disease where an inhaled substance or an underlying illness known to cause lung damage cannot be identified. The cause of lung scarring in these people is unknown and is the subject of much research around the world. The medical term for a condition where the cause is not known is called "idiopathic" and doctors refer to scarring in any tissue as "fibrosis" - hence the name "idiopathic pulmonary fibrosis". This condition is also known medically as "cryptogenic fibrosing alveolitis".

Special Tests

Your doctor will usually recommend a number of special tests to try and work out the cause of your lung condition. Blood, urine and lung function tests are usually performed. In most cases the doctor will recommend an internal examination of the lungs. This is called a bronchoscopy. During this procedure the doctor can obtain samples of lung fluid and lung tissue for examination in the laboratory. These samples are very helpful in making a diagnosis.

Treatment of Interstitial Lung Disease

In cases where the disease is due to inhaling foreign material, avoidance of the substance can often stop the progression of the illness or reverse it. For example pigeon breeders may have to give up all contact with birds or aviaries. In cases where the cause remains unknown after detailed testing, a trial or treatment with corticosteroid drugs is often used. In some cases, these are combined with other anti-inflammatory drugs, for example cyclophosphamide. This drug has very strong anti-inflammatory effects and can sometimes slow or stop the progressive scarring of the lungs. The progress of the disease is monitored by regular breathing tests and sometimes by measurement of the amount of oxygen in the blood. Regular x-rays of the chest can also be helpful. The disease may sometimes stop progressing of its own accord leaving an amount of permanent but stable lung scarring, or alternatively may progress gradually over months or years and require continual treatment. In some patients where the condition is severe, oxygen supplied at home can be helpful in the relief of breathlessness.

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