PATIENT INFORMATION FOR IDIOPATHIC PULMONARY FIBROSIS (IPF)

What is IPF?

IPF is a disease that leads to persistent and often progressive scarring of the lungs. The condition is also known as cryptogenic fibrosing alveolitis and also as usual interstitial pneumonia. Unfortunately this can be confusing for both patients and doctors, so many doctors now try to use the term IPF to describe this disease. The scar tissue prevents the lungs from working normally and it becomes more difficult for oxygen to pass from the air sacs in the lungs into the blood, where it is needed. The amount of scarring usually increases with time and is generally irreversible. The speed of progression is very variable; some people may remain stable for several years whilst others deteriorate more rapidly.

There are about 4000 new patients diagnosed with IPF each year in the UK, and the condition occurs throughout the world. Furthermore the disease does seem to be becoming more common.

Who develops IPF?

Most people with IPF develop their symptoms between the ages of 60 to 85 years and the disease is uncommon below the age of 50 years. Men are affected more commonly than women. Despite the efforts of researchers in this area the cause of IPF is not known. IPF is not 'passed on' to children, but occasionally more than one family member may have IPF, suggesting that genes may be involved in causing the disease.

What are the symptoms of IPF?

The main symptom is usually shortness of breath on exertion, for example walking up hills or stairs. A cough, which is often dry, is also common and finger 'clubbing', which is a change in shape of the fingernails, may occur.

How is IPF diagnosed?

The diagnosis is usually made by a specialist, and so for most patients this means being referred by their general practitioner to the hospital. Most people will need to have the following investigations.

- A chest x-ray. This may show signs of the scarring even early on, so some people will be diagnosed by an abnormal chest x-ray before they develop symptoms.
- Lung function tests. These are breathing tests to show how well your lungs are working. They are also used later on to monitor the severity of the disease and how it is progressing.
- Blood tests. These are usually done to exclude other causes of lung disease.
- A CT scan. This will usually be done and is a special x-ray that gives a more detailed picture of your lungs. There are characteristic features on the CT scan that can allow your doctor to make the diagnosis of IPF.

In addition, some people may have a bronchoscopy which involves passing a telescope down into the lungs to collect samples. Finally some patients may also need to have an operation with a general anaesthetic to remove one or more pieces of lung for more detailed tests. This procedure is called surgical lung biopsy and sometimes is the only way to make a firm diagnosis.

How is IPF treated?

The drug treatments that are currently available for IPF do not work very well and may have side effects. For this reason we often do not treat people automatically, and may monitor breathing tests and symptoms first.

The treatments that are generally tried for IPF are steroids and other drugs which suppress the immune system. Unfortunately these drugs do not work for most patients and the side effects of the drugs may outweigh the benefits. For example high doses of steroids may cause weight gain, thinning of bones and diabetes, whilst other drugs may dampen down your immune response making you more likely to pick up infections. Patients on such drugs need careful supervision with regular blood tests.

New treatments for IPF are however becoming available, but we do not know yet if they will be effective. Your doctor may discuss taking part in a clinical trial of new drugs if this is suitable. If you are under the age of 65 years and otherwise well your doctor may discuss lung transplantation with you.

What will happen if my condition starts to deteriorate?

Unfortunately most patients with IPF will become more breathless with time and some will die of the disease. Your doctor will be able to provide medications to relieve the symptoms and may recommend oxygen therapy if you are very breathless. You may need to have assistance with mobility or simple changes to the house to make activities such as showering easier.

Further information

This sheet contains general information about IPF. As the disease affects different people in different ways, it is best to discuss your illness with your own doctor.