

PATIENT INFORMATION FOR IDIOPATHIC PULMONARY FIBROSIS (IPF) at Heart of England NHS Trust

What is IPF?

IPF is a disease that leads to persistent and often progressive scarring of the lungs. The condition is also known as usual interstitial pneumonia and used to be called cryptogenic fibrosing alveolitis.

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. Idiopathic means of no known cause, however the disease is more common in smokers, people who have worked with metals, wood, stonecutting, textiles and working with animals or birds. A similar disease may complicate rheumatic diseases such as rheumatoid arthritis and scleroderma, and may be due to treatments for these diseases and some other drugs. IPF is not 'passed on' to children, but sometimes occurs in several members of a family suggesting that genes may be involved in causing the disease. A search for underlying causes is an important part of the initial investigation.

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 done to find other causes of lung disease.

- CT scan.
This 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 about half of those with IPF, others requiring a biopsy if the changes on the CT are not classical.
- A bronchoscopy
This involves passing a telescope down into the lungs to collect samples.
- A biopsy
If the diagnosis is not clear after the above tests some patients may 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 slow disease progression but are not cures; they may have side effects. Pirfenidone and Nintedanib are currently available on the NHS for some patients with IPF, They delay progression by about one third. The NHS will only pay for these (about £20,000 per year) when the forced vital capacity (FVC, a breathing test which measures the maximum amount of air that you can breathe out) is between 50 and 80% of your predicted value. There are lots of patients with IPF who have values above this, some of whom have advanced disease. Your doctor may discuss taking part in a clinical trial of new drugs if this is suitable.

High doses of steroids were used to treat IPF, but are no longer recommended apart from short-term use during exacerbations. They often cause weight gain, thinning of bones and diabetes.

If you are under the age of 65 years and otherwise well you may be suitable for lung transplantation if your disease becomes severe.

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. Sometimes the disease does not progress, more usually there is continuing scarring with increased breathlessness.

Some patients have acute exacerbations when breathlessness increases suddenly. This is often confused with pneumonia and needs urgent hospital care with a repeat CT scan and different treatment.

You will be followed up by a specialist nurse who will help you through your disease. We will be able to provide medications to relieve the symptoms and provide oxygen therapy when needed. You may need to have assistance with mobility or simple changes to the house to make activities such as showering easier. State benefits are available to help with this.

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 specialist nurse and doctor. This information sheet is based on the Department of Health DOAS document, and the British Thoracic Society ILD guidelines, and has

been updated to include the practice of the Interstitial Lung Disease Service at the Heart of England NHS trust, September 2008.