Patient

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Pulmonary fibrosis

Pulmonary fibrosis is a serious lung disease where the tiny air sacs of the lungs (the alveoli) and the lung tissue next to the alveoli become damaged and scarred, resulting in lung fibrosis. The main symptom is shortness of breath that gradually gets worse. The cause is often not known but pulmonary fibrosis can be caused by a variety of underlying conditions. Treatments include treating any underlying cause in addition to steroids and other medicines. Increasingly lung transplantation may be considered.

What is pulmonary fibrosis?

Pulmonary fibrosis means a build-up of scar tissue (fibrosis) in your lungs. The scar tissue restricts the transfer of oxygen from the tiny air sacs in your lungs into your bloodstream. See the leaflet called The respiratory system for more information about the lungs.

Pulmonary fibrosis causes

The main causes of pulmonary fibrosis are:

Unknown (called idiopathic pulmonary fibrosis, or IPF) - see below for more information.

Hypersensitivity pneumonitis:

- This happens when something you breathe into your lungs causes a reaction, which causes inflammation. Over a period of time this gradually causes scarring (fibrosis).
- Examples include farmer's lung (caused by breathing in mould that grows on hay, straw and grain) and bird fancier's lung (caused by breathing in particles from feathers or bird droppings).

- There are many other substances which can cause pulmonary fibrosis. It can sometimes be difficult to know exactly which substance is responsible.

Occupational interstitial lung disease (pneumoconiosis):

- These lung diseases are caused by breathing in certain dusts when at work.
- Over a period of time the dust particles lodge in your lungs and cause scarring.
- The most common type is coal worker's pneumoconiosis, caused by breathing in coal dust. Other forms include silicosis (caused by silica dust) and asbestosis.
- There is often a long delay of many years between breathing in the dust and developing symptoms of pulmonary fibrosis.

Pulmonary fibrosis associated with autoimmune diseases:

- An autoimmune disease means that your immune system, which normally attacks germs (bacteria, viruses, etc), attacks part of your body.
- When the disease is active, parts of your immune system may attack your lungs, causing inflammation. Over a period of time, the inflammation slowly leads to scarring (pulmonary fibrosis).
- Examples include rheumatoid arthritis, Sjögrens syndrome and scleroderma.

Sarcoidosis:

Sarcoidosis often affects the lungs. It can sometimes cause pulmonary fibrosis.

Medicine-induced pulmonary fibrosis:

Certain medicines may occasionally cause pulmonary fibrosis as a side-effect. Examples include:

- Chemotherapy medicines.
- Amiodarone (a medicine used to treat certain heart conditions).

- Methotrexate (an immunosuppressant medicine).
- Nitrofurantoin (an antibiotic).

If pulmonary fibrosis is caused by an underlying cause then the symptoms, treatment and outcome will depend on the underlying cause. However, the symptoms and treatment of the lung symptoms will be similar to idiopathic pulmonary fibrosis (see below).

If a medicine has caused the fibrosis, people often get better quickly if the medication is stopped at an early stage. However if the medicine isn't stopped soon enough then the lung scarring will be permanent.

The rest of this leaflet is about idiopathic pulmonary fibrosis.

What is idiopathic pulmonary fibrosis?

Idiopathic pulmonary fibrosis (IPF) means lung scarring (fibrosis) of unknown cause. It is useful to understand what the following words mean:

- Idiopathic means 'of unknown cause'.
- Pulmonary means 'affecting the lungs'.
- Fibrosis means 'thickening' or 'scarring'.

For more information about the lungs and alveoli, see the leaflet called The respiratory system.

Idiopathic pulmonary fibrosis symptoms

The early symptom of pulmonary fibrosis is usually shortness of breath with activity (exertion), with or without a dry cough. Symptoms tend to develop gradually:

- Shortness of breath that gradually becomes worse over time is the main symptom. This is due to the reduced amount of oxygen that gets into the bloodstream. Shortness of breath on exertion may be noticed at first. This may be passed off as 'just getting older' for a while until the shortness of breath gets worse.
- A dry cough often develops (a cough with little or no sputum).

- Finger or toenail clubbing occurs in about half of cases. Clubbing is a painless swelling at the base of the nails. The cause of clubbing is not clear. It occurs in people with various chest and heart diseases.
- Tiredness.
- An increased risk of developing chest infections.
- Weight loss may occur, due to the extra energy needed as breathing becomes more difficult. Losing over 5% of body weight in the first year of diagnosis is associated with worse outcomes for those who do not have a lung transplant.

As the condition progresses, symptoms can sometimes quickly become worse (called an acute exacerbation). Periods of worse symptoms may be followed by periods where there seems to be some improvement.

What are the symptoms of severe and end-stage IPF?

Shortness of breath gradually becomes severe and present at rest as well as when you're active.

Heart failure may develop when IPF becomes severe. This is due to the reduced level of oxygen in the blood and to changes in the lung tissue, which can cause an increase in pressure in the blood vessels in the lungs. This increase in pressure can put a strain on the heart muscle, leading to heart failure. Heart failure can cause various symptoms, including worsening breathlessness and fluid retention. Pulmonary hypertension is an increased pressure in the right side of the heart - it is a complication of heart failure.

How common is idiopathic pulmonary fibrosis?

IPF is rare. About 2 in 10,000 people develop this disease. However, it seems to be becoming more common. It can affect anybody at any age but it most commonly affects older age groups. It seems to be more common in men than in women.

Idiopathic pulmonary fibrosis causes

The current thinking is that somehow the cells that line the alveoli are damaged in some way. The cells then try to heal themselves. But, this healing process becomes out of control, causing thickening and damage to the walls of the alveoli, and scarring of the alveoli and lung tissue. The thickening and scarring reduces the amount of oxygen that can pass into the blood vessels from affected alveoli. Therefore, as the disease progresses, less oxygen than normal is passed into the body when you breathe.

Various things have been suggested as potential causes, or triggers, of the initial damage to the cells lining the alveoli. These include:

- Cigarette smoking is the main risk factor. The disease is more common in people who smoke or have been smokers at some time, and continuing to smoke after diagnosis is associated with poorer chances of survival.
- Viral infections. These include viruses such as the Epstein-Barr virus that causes glandular fever, and the hepatitis C virus.
- Certain medicines.
- Pollutants in the environment.
- Gastro-oesophageal reflux disease. In this condition there is long-standing reflux of stomach contents into your gullet (oesophagus), which you may then inadvertently inhale into your lungs. See the separate leaflet called Acid Reflux and Oesophagitis (Heartburn) for more details.

A similar fibrotic lung condition sometimes occurs as a result of exposure at work to certain metals, wood dusts and other chemicals, and as a side-effect to certain medicines. These conditions are not idiopathic, as the cause can be identified. If you have suspected lung fibrosis, your doctor is likely to ask you about your work history to rule out these conditions.

Is idiopathic pulmonary fibrosis hereditary?

IPF seems to run in some families. However, in about 4 out of 5 cases, there is no family history of the disease.

How is idiopathic pulmonary fibrosis diagnosed?

The above symptoms, combined with crackles heard by a doctor listening to your lungs with a stethoscope, can suggest the diagnosis of IPF.

Chest X-ray

A chest X-ray may show a typical scarring pattern in the lungs, suggestive of the disease. Special breathing tests can also show changes that raise suspicion. Your doctor may advise referring you to a chest specialist if they suspect that you have IPF.

Lung function tests

These are also called spirometry - they are often requested by the GP when someone presents with shortness of breath. The pattern of breathing restriction can make a difference as to whether a disease such as pulmonary fibrosis is more or less likely, but these tests cannot diagnose pulmonary fibrosis.

Computerised tomography (CT) scan

To confirm the diagnosis and to assess the severity of the disease, a computerised tomography (CT) scan of the lungs and/or a lung biopsy are usually needed.

High-resolution CT (HRCT) scan

A special type of CT scanning, called high-resolution CT (HRCT) scanning, is commonly used.

Lung biopsy

A lung biopsy is a procedure where a small sample of lung tissue is taken by a small operation. A keyhole procedure is normally used. The sample is looked at under a microscope. IPF causes a typical appearance of the tiny air sacs of the lungs (the alveoli) and nearby lung tissue. This can be seen when the biopsy sample is examined.

Bronchoscopy

Sometimes the specialist may suggest that they take samples of your lung cells in another way to examine them. This is usually done through a procedure known as a bronchoscopy. A small, flexible telescope is passed down your breathing tubes so that the samples can be collected.

An ultrasound

An ultrasound examination of your heart (echocardiogram, or echo) may be done if it is suspected that you have developed heart failure.

Idiopathic pulmonary fibrosis treatment

At present there is no cure for IPF and the optimal treatment has not yet been found. The aim of treatment is to suppress the symptoms as much as possible.

There is no conclusive evidence for using any medicines to improve survival for people with IPF. Steroids are not recommended and can be harmful.

One or more of the following options may be advised as a treatment option:

- **Oxygen treatment** used in the home may be required if symptoms become severe.
- **Pulmonary rehabilitation courses** may help some people. The courses include education about IPF and physical exercise, as well as psychological and social support. Ask the chest clinic that you attend about your nearest course.
- Stop smoking if you are a smoker.
- **Get immunised** against influenza and pneumococcus. They protect against infections that can be particularly severe if you have a disease of the lung.
- **Pirfenidone** and **nintedanib** are medicines that may be used as treatment options for IPF if certain criteria are met. Decisions about whether to use/continue these drugs may be based on factors such as how well the lung is functioning (seen on lung function tests) and whether the disease continues to progress after the drug is started.
- **A lung transplant** may be an option. This is becoming more common, particularly in younger people who develop severe disease despite medication.

Idiopathic pulmonary fibrosis prognosis

The progression of the disease can vary greatly. Some people respond to medication which may slow the progression of the disease but, in others, it makes little difference. Some people remain stable for many years after being diagnosed but others deteriorate much more rapidly. It is difficult to predict at the outset how fast the disease will progress for each affected individual.

Lung transplantation has been shown to improve survival in those people for whom it is suitable and so is being increasingly used as a treatment. Newer treatments with medicines may also be shown to be of benefit in the future.

Further reading

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