Idiopathic Pulmonary Fibrosis

What is Idiopathic Pulmonary Fibrosis?

Idiopathic pulmonary fibrosis (IPF) is a condition that causes persistent and progressive scarring of the tiny air sacs (alveoli) in the lungs. The alveoli perform the vital functions of transferring oxygen to your blood stream from the air you breathe in, and transferring the waste product, carbon dioxide from your blood, to the air you breathe out. The amount of scar tissue irreversibly increases over time. This is a serious condition, however the rate at which the disease progresses is variable, with some patients remaining stable for years while others may deteriorate rapidly.

What causes patients to develop IPF?

At present, it is not known what causes the disease to develop and progress. The term ‘idiopathic’ literally means ‘of no known cause’ although it is known that the disease is more common in smokers (70% of those diagnosed have a history of significant nicotine consumption). As a general rule, IPF is not passed on to siblings or children, but on occasions, several members of a family may be affected. This suggests that one’s genetic profile may be a factor in the cause of the illness.

How common is IPF?

We don’t really know how many people in Australia have IPF, but based on international rates, we estimate that approximately 2,300 new cases of IPF are diagnosed every year. The risk of IPF increases with age; the disease is uncommon below the age of 50 years. Men are slightly more commonly affected than women and the disease affects people of all races.

Many cases of IPF remain undiagnosed until later in the disease course. Patterns of scarring similar to IPF may be seen in a number of other medical conditions, including in people who have worked with animals or birds, silica or asbestos. Scarring can also be secondary to diseases such as rheumatoid arthritis, lupus and scleroderma. Careful attention to a patient’s past medical and occupational history along with complex testing is therefore required to avoid confusion with these conditions.

What are the symptoms?

The most common symptom of IPF is shortness of breath on exertion, which is particularly noticeable when walking up hills or stairs. Other symptoms may include cough which is generally dry, although in the latter stages of the disease, patients may produce clear phlegm. Some patients may also notice their fingernails and toenails change to a beak shape, known as clubbing. It is vital that any symptoms consistent with IPF are investigated by a specialist.
How is IPF diagnosed?

The diagnosis of IPF is challenging and requires careful evaluation of clinical, laboratory, x-ray data, high resolution computed tomography (HRCT) and sometimes lung biopsy material to make a confident diagnosis. This is usually made by a respiratory specialist in conjunction with other specialists with an interest in IPF. Investigations are likely to include:

Physical examination – your GP or specialist may hear “velcro-like” fine crackles in your chest which may be a valuable sign in early diagnosis. Fine crackles are present in more than 80 percent of patients in the early stages of the disease.

A chest x-ray - this may indicate some areas of scarring in your lungs however a CT scan is necessary to confirm the presence of scarring or fibrosis. Early in the disease, scarring is usually located in the lower zones of each lung. Occasionally, patients may be diagnosed with IPF on the basis of an abnormal x-ray before they develop symptoms.

Lung function tests - these are breathing tests to show how well your lungs are working and are an important monitor of how your disease is progressing over time.

Blood tests - these are generally performed to rule out other causes of pulmonary fibrosis.

High Resolution Computed Tomography (CT) scan - this is a detailed picture of your lungs that gives multiple images compared with a simple chest x-ray.

Surgical lung biopsy – In most cases, after assessment of medical history, physical examination findings, lung function tests and CT images of the chest, IPF experts can diagnose the condition with confidence. Nonetheless, in some instances you may be referred to a surgeon who will remove a small piece (called a biopsy) of your lung under general anaesthesia. This will require you to stay in hospital over a two to three day period. In patients with IPF who have undergone a biopsy, the pattern that is referred to by pathologists is usual interstitial pneumonia (UIP).

How is IPF treated?

Previously, there have been very few treatment options available for IPF patients. Many medicines, such as powerful immune suppressant agents, have been trialed in patients with IPF. Unfortunately, no medicine of this nature has ever been shown in a well-designed clinical trial to alter the outlook for patients with this condition in a positive way. A clinical trial of the combination of prednisolone/azathioprine/N-Acetylcysteine (NAC) was associated with significantly increased mortality and hospitalisation compared to placebo.

a) Anti-fibrotic therapy

In May 2014, two clinical trials were published in the medical literature. These trials, called ASCEND and INPULSIS 1 and 2, evaluated the role of new medicines called pirfenidone and nintedanib, respectively, in patients with mild to moderate IPF disease. Prescription of each drug compared to placebo was associated with a significant reduction in decline in lung function over a 12 month period. Submissions
are in the process of being formulated with the Pharmaceutical Benefit Advisory Committee (PBAC) to have such treatment available in Australia on the Pharmaceutical Benefits Scheme. The results of this process may not be known until early 2016. Your specialist may discuss with you options to potentially access these therapies via a compassionate access program. Whilst these therapies are revolutionary in terms of IPF management, the agents slow disease progression rather than halting altogether or even reversing scarring.

b) Clinical trials

The future of IPF therapy still rests with well-designed clinical trials to evaluate the ability of new therapies to stop or reverse existing lung scarring. It is recommended to discuss the possibility of taking part in a clinical trial with your specialist. A list of the clinical trials which are being conducted in Australia can be accessed at http://lungfoundation.com.au/patient-support/other-lung-conditions/idiopathic-pulmonary-fibrosis-ipf/.

c) Enrol in the Australian IPF Registry

In addition, you may also want to discuss joining the Australian IPF Registry with your specialist. The purpose of the Registry is to collect anonymous information from patients with IPF that can be used by approved Australian researchers to better understand this complex condition. Your data may also be used to identify participants for clinical trials, helping to develop better treatments. Participating involves completing a questionnaire and allowing the Registry to review and store your medical information and test results.

d) Regular follow-up

Regardless of treatment, you are encouraged to visit your specialist regularly to have your lung function assessed and to have other clinical assessments performed. This enables your condition to be closely and regularly monitored and any treatment adjusted accordingly. There is also the possibility that you may be referred to a hospital specialising in lung transplantation where your suitability for transplantation will be assessed.

e) Other

Other aspects of your clinical management will include discussing your vaccination schedule for protection against influenza and pneumonia and you may be referred to a physiotherapist for a structured exercise program, also known as a pulmonary rehabilitation program. Lung Foundation Australia can help you find out if there is a program near you. It is important to keep as fit as possible.

f) Treatment of acute exacerbation

Some patients with IPF may experience a sudden increase in breathlessness (also known as an acute exacerbation). In such situations, urgent assessment by your specialist or presentation to your nearest hospital will be required for you to begin treatment. Sometimes, pneumonia, a clot in the lungs or heart problems cause similar symptoms and will need to be ruled out.

g) Oxygen and symptomatic care

As scarring increases, you may be eligible for oxygen therapy to improve your exercise capacity. Palliative care is a central part of the management of IPF patients with advanced disease, with an aim to relieve shortness of breath, cough and assist with providing emotional support. You may need assistance with
mobility or be required to make simple changes to your home to ensure activities such as showering are made easier. Community services coordinated by respective State Governments are available to help with this and can usually be accessed with the help of your general practitioner or specialist. Finally, psychological and spiritual support to assist patients in end of life planning is available with the help of community organisations.

Other support for patients with IPF

Having a chronic lung condition such as IPF is frequently stressful. In addition to the physical symptoms such as breathlessness, you may feel stressed and angry at your diagnosis, depressed, anxious, worried, isolated or confused. As the disease is ongoing, it may take time for you to accept your diagnosis. Psychological support is very important. Patient Support Groups can help you to manage and control your symptoms, overcome negative feelings, improve confidence and build new friendships.

Patient support can also help your family and carers who are often affected by your diagnosis and may feel stress and concern about what is happening. Lung Foundation Australia provides information and support services, including overseeing a network of Patient Support Groups for people affected by chronic lung disease across Australia. For more information, or to find out if a Patient Support Group exists near you, please call 1800 654 301.

Further Information

Lung Foundation Australia
www.lungfoundation.com.au

Australian IPF Registry

Pulmonary Fibrosis Foundation (US)
www.pulmonaryfibrosis.org

Coalition for Pulmonary Fibrosis (US)
www.coalitionforpf.org

Clinical Trials

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