

Diagnosis of IPF

A limited awareness of the cause and progress of the disease has often made diagnosis difficult and it is common for the condition to be incorrectly diagnosed initially. A diagnosis is usually made by a specialist, after a referral by a general practitioner (GP). How commonly IPF occurs is not clearly known, but a reasonable estimate would be around 5 cases per 10,000 individuals – which means most GPs may encounter a new case about once per decade of full time work.

Most people will have the following investigations:

History and physical examination: Talking to the patient and often their family to find any factors in the home or work environment, any medical illnesses that run through the family, any medications, treatments (such as radiotherapy) or any other medical conditions that might make a person at risk of getting ILD. When listening to the lungs with a stethoscope, a doctor may hear crackles (often referred to as “velcro crackles”). However as mentioned above, other conditions can also cause crackles, so they cannot be relied upon for a definite diagnosis.

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 used later to monitor both how bad the disease is, and assess rate of worsening. There are several different sorts of lung function tests, each of which give different bits of information. A simple peak flow rate (as measured in asthma with a peak flow meter) is of no particular relevance in IPF, as it relates more to flow through the airways.

Pulse oximeter: This test indicates the amount of oxygen carried in the blood. A device called an oximeter – usually a simple clip on device - is placed on an earlobe or a finger, and measures the oxygen concentration in the blood (usually referred to as “Sats”, as a much easier shortening of oxyhaemoglobin saturation), as it pulses past. Pulse oximetry is painless and simple. In room air the normal range is 95–100%. It is worth noting oximetry does not measure carbon dioxide levels (the ‘waste gas’ of breathing): to do so an Arterial Blood Gas (ABG) must be taken.

Arterial blood gases: A direct measurement is made of both oxygen *and* carbon dioxide levels in blood, by taking a sample with a needle and syringe from an artery, usually in the wrist. Venous blood has much lower oxygen concentration than arterial blood, and therefore cannot be used to make decisions about the need for oxygen treatment alone. Arterial blood gas measurement is often somewhat more uncomfortable than a standard blood test, as the artery is often small and has plenty of nerves around it.

Blood tests: These are usually done to exclude other causes of ILD (such as rheumatological diseases).

CT scan (HRCT): This is a special type of x-ray scan which produces very high resolution pictures of the inside of the lungs. There are typical features in the CT scan of lungs that can allow a doctor to make a diagnosis of IPF. An HRCT allows an image to be created of virtual “slices” of lung tissue, maybe only 1mm thick, that often show the changes needed to diagnose IPF confidently. Sometimes the diagnosis can be made with a very high degree of certainty (“Definite IPF”) and sometimes there can be less confidence (“Probable IPF”). It can be difficult to tell IPF from some other types of ILD – the international standard for diagnosing IPF is to have a formal team of experts called an ILD-MDM who review cases and decide on the cause of an ILD. Because ILDs are fairly rare, the ILD-MDMs often cover quite large areas including a number of different hospitals as they have to see enough cases to keep their skills ‘sharp’.

Bronchoscopy: Some people may also require a bronchoscopy, which involves passing a small flexible telescope down into the lungs to collect samples. This procedure is often with sedative drugs rather than a full general anaesthetic. This is a safe procedure that is usually very well tolerated. Occasionally lung biopsies may be taken during bronchoscopy, although as they are so small (1-2mm) they are rarely that helpful outside of a few special circumstances – for instance diagnosing pulmonary sarcoidosis, a disease that can also cause ILD. It is more likely that washings are taken from the inside of the lungs to make sure there are no unusual infections present.

Surgical lung biopsy: Some patients may also need to have a procedure called a surgical lung biopsy under general anaesthetic, to make a firm diagnosis of IPF. These biopsies are much bigger than those taken during bronchoscopy, although still only a tiny proportion of the whole lung tissue. Because it is often possible to diagnose IPF confidently on HRCT appearances and the clinical history, surgical lung biopsies are generally more useful for diagnosing other types of ILD than IPF.