What is Idiopathic Fibrosis (IPF)?

Idiopathic Pulmonary Fibrosis, or IPF, is a serious condition that affects the delicate tissues of the lungs. Normally the lung tissue is soft and flexible which means it is easy to breathe, but in someone with IPF the lung tissue becomes stiffened and damaged from scarring. The process of the scarring of the lungs is called fibrosis, and once it has occurred the tissue does not recover.

- *Idiopathic (means a condition without any known cause)*
- *Pulmonary (relating to the lungs or respiratory system)*
- *Fibrosis (a rapid increase of fibrous connective tissue – this process occurs naturally in the formation of scar tissue to replace normal tissue lost through injury or infection, but in this instance causes irreversible damage of the lung tissue)*

IPF is just one of the many types of Interstitial Lung Disease (or ILDs). The *Interstitium* is the name given to the 'scaffolding' structure of the lung tissue, which normally gives the lungs their structure and shape. This means that ILDs are different to other lung diseases, such as asthma or COPD, which mostly affect the airways coursing through the lung tissue. It also means that inhalers are rarely of benefit in ILDs. You may hear of ILDs as a group of illnesses that allow medical teams to organise their care for illnesses with similar features – for instance in a specialist ILD clinic, or at an ILD Multidisciplinary Meeting (ILD-MDM). Because ILD and IPF sound quite similar, reading up about them online can be confusing. We hope this information helps explain the differences between ILD and IPF, as well as outlining the possible treatments of IPF.

IPF (formerly known as "cryptogenic fibrosing alveolitis", especially in the UK), is caused by repeated injury to the lungs, leading to scarring. Scars generally serve a good purpose on the skin – they heal injured areas – but in the lungs, scar tissue stops the lungs doing their job of taking oxygen from the air that we breathe in, and passing the oxygen that the air contains across into to the blood. The amount of lung scarring usually increases and is generally irreversible. The speed at which the condition progresses varies: some people with the condition stay stable for many years, and others worsen more quickly. Unfortunately IPF is a terminal disease with an average life expectancy of about 3–5 years after an early diagnosis; however within that average there are a small group of people who get worse very quickly and appear to have a disease very resistant to treatment, and another group who have a much more gentle time course with a slow reduction in physical condition over many years (about 20% of IPF patients or about 1 in 5 are alive 5 years after diagnosis). This makes estimating life expectancy for an individual quite difficult.

What are the causes of ILD? –The non-IPF diseases

There are many possible causes of interstitial lung disease, but IPF is the most common variety. The focus of this information is therefore on IPF, but it is worth mentioning other non-IPF fibrotic diseases, as it can be difficult sometimes to distinguish the different types, which can have quite similar appearances on medical tests like CT scans.

If ILD is due to a particular disease or drug, then that cause should be treated in its own right. For instance, rheumatoid arthritis and some other rheumatological diseases can cause pulmonary fibrosis that looks quite a lot like IPF on CT scan. Similarly some drugs such as amiodarone (used to keep heart rhythm regular), methotrexate (often used in rheumatological diseases, such as some types of arthritis), and nitrofurantoin (an antibiotic commonly used for urine infections) can cause ILD as well. If a drug treatment is likely to have caused fibrosis it should be stopped.

If an underlying disease can be treated, then it is reasonable to treat that disease aggressively – for instance, in a condition such as antisynthetase syndrome that has led to ILD, strong immunosuppressant treatment is likely to be tried. If control of the antisynthetase syndrome is achieved, there can be improvement in the lung function as well. If there is an underlying cause, the ILD cannot – by definition – be idiopathic, which means “of no known cause or spontaneously occurring”.

If there is no reason that can be found for the ILD, and certain criteria are met – usually based on CT scan appearances - it can be correctly labelled Idiopathic Pulmonary Fibrosis. Many medications have been tried for IPF, but currently (2018) there are only two medications that have reasonable evidence for benefit that are licensed for use in New Zealand. Neither reverse the disease, but the aim is to achieve stability if possible, or a slower rate of decline than might otherwise be seen.