Interstitial Pneumonia
a guide for patients

What is ‘Interstitial Pneumonia’?

In everyday speak pneumonia refers to a bad chest infection. Medically it is distinguished from other types of respiratory infections such as bronchitis by the presence of shadowing on a chest X-ray or scan. Bacteria or viruses cause inflammatory material to accumulate in the airspaces of the lung (alveoli) which absorb X-rays. Usually infection responds to antibiotic treatment, and the chest X-ray returns to normal after six to eight weeks.

Sometimes, symptoms occur associated with X-ray shadowing which is not caused by infection. This shadowing can persist for months or longer. Many of the diseases affect the lining of the alveoli (known as the lung interstitium) rather than the airspaces themselves. These conditions are known as ‘Interstitial pneumonia’, although they are not caused by infection as standard pneumonia is.

In some of these diseases the X-ray changes are largely due to scarring or ‘fibrosis’ of the lungs which can be hard to reverse, but fibrosis is not seen in all cases of interstitial pneumonia. It is best to think of interstitial pneumonia as a group of related diseases rather than a single diagnosis, as we know that the conditions differ in their treatments and outcomes. It is possible to separate those diseases where a cause is known or suspected (secondary interstitial pneumonia) from those where no cause is known.

Causes and associations of secondary interstitial pneumonia

Interstitial pneumonia can be caused by exposure to inorganic dusts and particles. These diseases are known as pneumoconiosis. The common dusts, and their associated diseases, are: asbestos (asbestosis), silica — sand and rock dust — (silicosis), and coal dust (coal-worker’s pneumoconiosis). People with these conditions may be entitled to compensation.

Exposure to organic particles can also trigger an interstitial pneumonia in susceptible individuals. The preferred term for this is hypersensitivity pneumonitis (HP) which was previously known as extrinsic allergic alveolitis (EAA). The condition can be caused by exposure to proteins from bird feathers (bird-fancier’s lung) or mould growing on hay and straw (farmer’s lung), but a long list of other organic particles can also lead to the condition.

In certain individuals, interstitial pneumonia can develop as a side effect of medications. A variety of drugs can cause it, including the cardiac drug amiodarone as well as drugs used in rheumatological disease (especially methotrexate) and in chemotherapy.

Finally, interstitial pneumonia can occur together with joint, muscle or skin inflammation as part of rheumatological diseases. Associated diseases include rheumatoid arthritis as well as the less common conditions systemic sclerosis (scleroderma), Sjögren’s disease, polymyositis and dermatomyositis.
**Idiopathic Interstitial Pneumonia**

When none of the above associations are found, the disease is said to be **idiopathic** (meaning of unknown cause). What we now call idiopathic interstitial pneumonia (IIP) was traditionally known as **cryptogenic fibrosing alveolitis (CFA)** in the UK, and as **idiopathic pulmonary fibrosis (IPF)** elsewhere. It is important to highlight that the term IPF is nowadays reserved for a more specific subset of IIP (see below).

**Lung biopsy in Idiopathic Interstitial Pneumonia**

It has been found that a variety of different patterns of interstitial pneumonia can be seen when lung biopsies taken at operation are examined under the microscope. The distinction is important as it determines the response to various treatments, and the overall prognosis.

The most common, or usual, pattern is known as **Usual Interstitial Pneumonia (UIP)**. When this pattern is seen at biopsy the disease is called **Idiopathic Pulmonary Fibrosis (IPF)**. This is distinct from its old use as a term for all idiopathic interstitial pneumonia.

Other disease patterns are the so-called **’Non-specific interstitial pneumonia’ (NSIP)** and **cryptogenic organising pneumonia (COP)**, where the inflammation is in the airspaces as well as the interstitium. An old term for COP which is now no longer used was **bronchiolitis-obliterans organising pneumonia (BOOP)**. Together with **lymphocytic interstitial pneumonia (LIP)** — typically seen in Sjögren’s disease and in HIV — NSIP and COP can occur in patients with rheumatological disease as well as in idiopathic disease.

Although traditionally counted as idiopathic interstitial pneumonia two patterns of disease are now known largely to relate to **tobacco smoking**. A common response of the lung to inhaled smoke is the accumulation of immune cells called macrophages which attempt to clear the particles. When these accumulate largely around the very smallest airways the pattern is called respiratory bronchiolitis and the disease known as **respiratory bronchiolitis-associated interstitial lung disease (RBILD)**. If the macrophages accumulate throughout the airspaces the condition is called **desquamative interstitial pneumonia (DIP)**.

**Diagnosis of interstitial pneumonia**

Diagnosis requires careful questioning and examination, especially looking for secondary causes. Blood tests are taken to look for associated rheumatological conditions. So-called **high resolution CT (HRCT)** scans show the pattern of lung involvement much better than chest X-rays. Some diseases have very typical HRCT appearances. In particular, if a patient with IIP has a particular HRCT appearance the doctor can confidently diagnose IPF. In other cases a biopsy taken by a surgeon under general anaesthetic is needed to make a diagnosis. We aim to discuss all cases in a multidisciplinary team (MDT) of doctors, surgeons, pathologists and X-ray specialists.