Idiopathic pulmonary fibrosis is a devastating lung scarring disease that kills more people than many cancers

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Idiopathic pulmonary fibrosis develops insidiously and is often misdiagnosed as more benign conditions. There are anti-scarring drugs, but they do not cure or stop the disease from getting worse. There is hope on the horizon with new drugs and patient advocacy playing a leading role.

Idiopathic pulmonary fibrosis (IPF) is a form of progressive scarring disease of the lung. Progressive pulmonary fibrosis (lung scarring that gets worse over time) is an uncommon condition and can be associated with inhaled dusts such as asbestos, exposure to parrots or pigeons and diseases such as rheumatoid arthritis. However, the most common form of lung scarring has no obvious cause and is therefore called idiopathic pulmonary fibrosis.

IPF is a devastating lung disease that leads to progressive disability and ultimately death on average three years after diagnosis. Only 25% of people survive with the condition for five years. The prognosis is worse than most cancers. Although less common than most other lung diseases, it affects approximately 30,000 people in the UK and it is the fourth biggest killer after lung cancer, COPD and pneumonia.

Strong genetic trends with IPF

While the precise cause of IPF is not known, risk factors associated with its development include exposure to cigarette smoke, dust and pollution. Furthermore, there is a strong genetic component to the illness and 6% of people with IPF have a parent or sibling with the condition.

Screening studies have shown that approximately 7.5% of the population have evidence of lung scarring on a CT scan. Fortunately, only a minority of patients will go on to develop progressive, symptomatic IPF.

Unfortunately, at the current time, it is not possible to predict who will get progressive IPF. The excess scar tissue in the lung starts gradually and most patients don't realise that their symptoms of breathlessness on exertion are anything serious. As the disease progresses, patients are often treated for other conditions such as chest infections, asthma or heart failure before being diagnosed with IPF, on average, three years after the onset of symptoms.

> *"Only 25% of people survive with the condition for five years"*

Treatment cannot stop or cure IPF

Anti-scarring drugs are available that have been shown to slow down loss of lung function as IPF progresses. Anti-scarring drugs are expensive and have a number of side effects. The National Institute of Health and Care Excellence (NICE) has approved anti-scarring drugs for patients with advanced IPF based on lung function criteria (forced vital capacity between 50 and 80% of the predicted value), but the best time to start therapy is not known.

Unfortunately, these drugs do not stop or cure the fibrosis and patients will still get worse on treatment but more slowly than without the medication. However, the speed at which the disease progresses for individual patients is difficult to predict.

As well as anti-scarring drugs, people with IPF should be offered supportive care such as pulmonary rehabilitation, cough suppressants and oxygen therapy as this can help alleviate some of the severe symptoms such as fatigue, cough and breathlessness associated with IPF.

What does the future hold for people with Idiopathic Pulmonary Fibrosis?

Despite the grave prognosis, there is considerable hope for patients with IPF. There are a large number of new, anti-scarring drugs that are being developed to treat this condition. Furthermore, the genetic insight into this condition has raised the prospect of precision medicine using targeted treatments tailored to patients with specific genetic or molecular abnormalities.

Artificial intelligence as well as wearable, and other smart technologies may help identify patients earlier, who may then gain more benefit from starting these new or conventional therapies sooner. Patient charities, such as Action for Pulmonary Fibrosis, have galvanised the community by raising money, funding research, helping to set up over 65 local support groups and providing educational resources for patients, carers and healthcare professionals.

This is a time of real optimism to be involved in treating people with progressive pulmonary fibrosis, not only because of the scientific advances, but due to the real collaboration between doctors and patients with a desire to end the suffering associated with this devastating illness.

"IPF is a deadly disease killing more and more people each year, but the increased collaboration between doctors, scientists and patient advocacy groups is leading to real improvements in patient outcomes that I am sure will get even better in the future." ■