IDIOPATHIC PULMONARY FIBROSIS (IPF)
MEDIA BACKGROUNDER

IPF overview

IPF is a debilitating and fatal lung disease.[1] It causes permanent scarring of the lungs, difficulty breathing and decreases the amount of oxygen the lungs can supply to major organs of the body.[2]

IPF progression is variable and unpredictable and over time the lung function of an IPF patient gradually and irreversibly declines. [1] The prognosis for those diagnosed with IPF varies, but median survival after diagnosis is 2–3 years.[3]

Sudden worsening in lung function, known as an acute exacerbation, can impact the course of the disease, often leading to death within a few months.[4] Approximately 5-10% of all IPF patients may experience an exacerbation over a year.[3] Acute IPF exacerbations are a risk to all IPF patients because they may occur at any time during the course of the disease without warning or known cause. [5]

Prevalence and possible causes

IPF affects approximately 3 million people worldwide.[6,7] The disease primarily affects patients over the age of 50 and affects more men than women.[3]

The cause of IPF is unknown, but risk factors may include smoking, lung injury, family history of the disease, abnormal acid reflux, environmental exposures and chronic viral infections.[1,3]

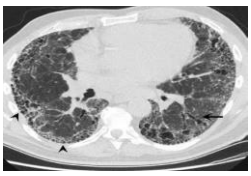
Symptoms of IPF

Individuals with IPF may experience symptoms such as breathlessness during activity, a dry and persistent cough, chest discomfort and finger clubbing.[3]

Diagnosing IPF

Diagnosing IPF can be difficult because it requires specific diagnostic testing, such as lung imaging using a high-resolution CT scan.[3,8] The median time from first symptoms to diagnosis is between one and two years.[1]

HRCT scan of an IPF patient



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Initial misdiagnosis occurs in around half of patients because symptoms are similar to other respiratory diseases like COPD, asthma and congestive heart failure.[9]

- Over 80% of patients with IPF have a distinctive, Velcro®-like crackling sound that can be detected through a stethoscope.[10]

Management options

Early and accurate diagnosis of IPF is important, as management options such as pharmacological treatment, supplemental oxygen treatment, cough management and pulmonary rehabilitation (which can include special exercises or breathing techniques) can help patients manage their condition.[3,11]

Treatments for IPF

Until recently, treatment options for IPF have been limited. There is a high unmet need for safe and effective treatments that can alter the course of IPF by slowing disease progression. Two treatments have been approved in the United States, European Union, Japan and other countries for the treatment of IPF.

The 2015 international evidence-based IPF guideline,[12] jointly developed by an international committee including the American Thoracic Society (ATS), the European Respiratory Society (ERS), the Japanese Respiratory Society (JRS) and the Latin American Thoracic Association (ALAT), provides physicians with updated treatment recommendations for patients with IPF. The guideline recommends the use of the following agents (conditional recommendation*):[12]

- Nintedanib
- Pirfenidone
- Antiacid therapy

*This is a conditional recommendation. This means that the majority of individuals in this situation would want the suggested course of action, but many would not. Clinicians should recognise that different choices will be appropriate for individual patients and are encouraged to discuss preferences with their patients when making treatment decisions.

ERS abstract information: Crestani B, et al. Long term treatement with nintedanib in patients with IPF: an updated from INPULSIS ®-ON. Abstract presented at the ERS International Congress 2016, London, September 3 - 7, 2016.

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