MEDIA BACKGROUNDER
Idiopathic pulmonary fibrosis (IPF)

Background information

What is idiopathic pulmonary fibrosis?

Idiopathic pulmonary fibrosis (IPF) is an irreversible lung disease, which is more deadly than most cancers.[1] Half of IPF patients die 2-5 years after diagnosis.[2]

IPF is a disease in which scarring and hardening of lung tissue stops the lungs from working properly. In people with IPF, the cells in the tissue surrounding the air sacs become damaged and begin to die. As a result, scarring and hardening of the lung tissue occurs and the air sacs are not able to expand as much as needed, which reduces the amount of oxygen entering the bloodstream, causing functional impairment and disability.[3]

The unmet need in IPF

IPF is a rare disease and awareness of it is low.[3] IPF is more deadly than most cancers, including breast cancer, prostate cancer, some forms of leukaemia and lymphoma.[4] A recent study comparing IPF to certain cancers shows only patients with lung and pancreatic cancer have a worse survival rate.[5]

The only cure at this time for IPF is a lung transplant, but less than 5% of IPF patients will receive one, and outcomes of transplantation for IPF are thought to be worse than for other indications. Survival estimates after transplantation for IPF have been shown to be 95% at 30 days, 73% at one year, 56% at three years, and 44% at five years.[6]

Causes and risk factors

The cause of IPF is not yet known; however, some evidence suggests that genetic factors may have a role in its development, and 5% of IPF patients have family members with the disease.[7] Other risk factors include:[7]

- Smoking (current or past)
- Occupational exposure to certain types of dust
- Viral infections
- Gastroesophageal reflux disease (GERD)
- Environmental factors

Epidemiology

- IPF is a rare disease. Approximately 100,000 people in the United States, and 110,000 people in Europe have IPF and 35,000 new patients are diagnosed in Europe each year.[8]
- According to Olmstead county data, it may be more prevalent than other rare diseases.[9]
- IPF is less well known than other rare diseases like lymphomas and pancreatic cancer.

- IPF is more common in men than women[10] and typically occurs in people over 45 years of age, with the average age being 65.[11]

Symptoms

Early signs and symptoms of IPF typically include:

- Cough that does not go away[3]
- Shortness of breath while doing everyday physical activities like going up stairs and, eventually, even while resting[3,12]
- Bilateral inspiratory crackles heard when listening to the chest, which have been described as sounding like Velcro® being slowly pulled apart.[13]

Late symptoms of IPF may include:

- Shortness of breath and/or cough while at rest, affecting routine daily activities such as eating, talking on the phone or showering[12]

IPF progresses differently in each patient. Some experience rapid progression and 'lung attacks' that lead to hospitalisations.[14] Others, if treated immediately after diagnosis, have been known to live much longer.[15]

Secondary conditions:
IPF affects the lungs but can eventually lead to heart failure or have serious effects on other vital organs.[8]

Diagnosis

The persistent cough and characteristic Velcro® crackles in breathing associated with the disease is often not recognised as IPF. Half of people with IPF are initially misdiagnosed, as the symptoms are often confused with other respiratory or cardiac illnesses, such as asthma, COPD, or heart failure.[16,17]

Diagnosis involves ruling out all other known causes of fibrotic lung disease, and may include one or more of the following:[18]

- Medical history and physical examination
- Chest X-ray
- Breathing tests
- CT scan
- Lung biopsy
- Bronchoscopy

For half of the people with IPF, it takes at least a year to receive a correct diagnosis.[19] This is a critical period as, without treatment, IPF only gets worse over time[20] and can progress rapidly.[13]

Treatment

60% of people diagnosed with IPF are not given treatment right away,[21] or do not get the best treatment available.[22] The patients most likely to receive treatment for IPF are those in the later stages of the disease whose lung function and quality of life is already severely impaired.[23]

Until 2011, there were no approved medicines for people with IPF. Nowadays, there is treatment available with the aim to slow the decline in lung function and slow the worsening of the disease. Treatments include medications such as anti-fibrotics, an anti-fibrotic kinase inhibitor, as well as oxygen support and pulmonary rehabilitation.[24]

References

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