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Facts About Idiopathic Pulmonary Fibrosis

DEFINITION AND STATISTICS

- ❑ Idiopathic pulmonary fibrosis (IPF) is a progressive and generally fatal lung disease marked by inflammation and scarring of the interstitium—the tissue layer between the alveoli and the blood vessels.
- ❑ IPF is one of about 200 disorders called interstitial lung diseases (ILDs). IPF is the most common form of ILD.
- ❑ Scarring, or fibrosis, caused by IPF thickens and stiffens the interstitium, causing an irreversible loss of the tissues' ability to transport oxygen.
- ❑ Although other ILDs may be attributed to exposure to asbestos or certain medications, IPF has no known cause.
- ❑ Potential risk factors for IPF include cigarette smoking and exposure to wood or metal dust. In addition, researchers are exploring a potential genetic predisposition for the disease.
- ❑ IPF affects about 50,000 people in the United States, with about 15,000 new cases expected to occur annually.
- ❑ About two-thirds of IPF patients die within five years.
- ❑ IPF occurs more often in men than women. Men tend to present at a later stage in the disease.
- ❑ A recent study found that IPF may be 5 to 10 times more prevalent than previously thought. It is unknown whether this may be due to an increased prevalence of the disease or to a previous lack of definitive guidelines for diagnosing IPF.
- ❑ Although IPF's cause is not known, the disease is believed to result from a normal tissue repair process gone awry. Inflammation, which is the body's normal response to injury, is usually followed by restoration of healthy tissue. With IPF, inflammation is chronic, and tissue repair is uncontrolled and exaggerated.
- ❑ There is currently no FDA-approved treatment for IPF.

DIAGNOSIS

- ❑ Until recently there were no uniform diagnostic standards for IPF. Other diseases with similar symptoms have often been diagnosed as IPF, despite widely varying prognoses. In addition, IPF is known by a number of other names (e.g. cryptogenic fibrosing alveolitis), which has caused confusion for patients and physicians.
- ❑ Patients with IPF typically experience shortness of breath and a dry cough, which become progressively worse and debilitating.
- ❑ In 2000, the American Thoracic Society and the European Respiratory Society, in collaboration with the European Respiratory Society, issued an International Consensus Statement defining the diagnosis, evaluation and management of patients with IPF.
- ❑ Diagnosis of IPF requires a multidisciplinary approach, usually involving a pulmonologist, pathologist and radiologist, to exclude other known causes of ILD and other diseases that mimic IPF.
- ❑ Physicians must take a complete patient history, perform a thorough physical exam, assess pulmonary function, and examine chest X-rays and high-resolution computed tomographic images. Lung biopsy, with or without bronchoalveolar lavage (a “lung washing” technique used for the examination of cells and proteins from inside the lung), is frequently required to rule out alternative diagnoses.
- ❑ According to the International Consensus Statement, a critical indicator of IPF is the presence of usual interstitial pneumonia (UIP), which is determined by doing a lung biopsy and is characterized by alternating areas of normal tissue, interstitial inflammation, fibrosis and “honeycombing”—abnormal holes in the lung.

TREATMENTS AND THERAPIES

- ❑ IPF patients typically are treated with anti-inflammatory drugs, including corticosteroids and cytotoxic agents, despite the fact that there is no evidence that they have any effect on long-term patient survival.
- ❑ Patients may require supplemental oxygen to help reduce breathlessness and allow the patient to be more active.
- ❑ Lung transplantation is recommended for consideration in patients with severe functional impairment, dependency on oxygen and continued, rapid deterioration despite optimal medical management if they meet established transplantation criteria.
- ❑ Recent studies indicate that an imbalance of “cytokines”—biologically active proteins in the body—appears to contribute to pulmonary fibrosis. Researchers are seeking to better understand the cellular and molecular biology underlying IPF to enable the development of more targeted interventions.
- ❑ Researchers are exploring a number of drug treatments for IPF. The most advanced study (in Phase III trial) is looking at the effect of interferon gamma-1b—a cytokine—on reducing collagen deposition in the lungs.