TREATING IPF

Treatment varies among patients, and thus could be tailored to a person’s need. Factors that determine the treatment plan include a patient’s age, the stage of the disease, the presence of other medical conditions and/or a disability, and a patient’s preferences and wishes. Medical efforts are focused primarily on early diagnosis as well as ways to decrease inflammation, remove the source of the problem if known, and reduce complications of the disease, which would prevent further lung damage. Early diagnosis and treatment of the inflammation will improve a patient’s quality of life.

There are two new drugs, Pirfenidone (Esbriet) and Ofev (Nintedanib) to treat IPF and possibly slow it down. There are also a number of non-pharmacological treatments:

Exercise. Daily walks or regular use of a treadmill or a stationary bicycle can vastly improve overall strength (especially in the muscles) and breathing ability.

Oxygen therapy. Some patients may need oxygen only during sleep and exercise, others (especially those with severe symptoms) may require it all the time.

Pulmonary rehabilitation program. While a rehab program combing exercise training, education and psychosocial support will not normalize lung functions that have already been disrupted, it will help improve a patient’s exercise endurance, breathing process, as well as quality of life.

Lung transplantation. This should only be considered in patients who meet the criteria for lung transplantation and who, despite optimal treatment, have a progressive lung condition.

Pneumococcal and influenza vaccine. Both are recommended to people with IPF and other chronic disease.
WHAT IS IPF?

Idiopathic Pulmonary Fibrosis (IPF) is one type of pulmonary fibrosis, a broad category of interstitial lung disease characterized by the scarring (fibrosis) of lung tissues. Idiopathic Pulmonary fibrosis is a chronic disease causing inflammation and scarring of the alveoli (air sacs) and interstitial tissues of the lungs.

IPF affects more males than females in their fifth or sixth decade of life. The disease takes a very progressive course with most patients dying of respiratory failure within two to five years of diagnosis. Five million people are currently affected worldwide, and there are 40,000 deaths annually.

WHAT ARE THE SYMPTOMS?

Shortness of breath and dry cough that progressively worsens are among the symptoms of IPF.

Most people tend to ignore the early signs, and postpone seeing their health professional until they are already short of breath even while doing simple routine activities.

As the disease progresses, patients experience breathing difficulties with greater frequency. Because the symptoms can be disabling, most patients will need oxygen by this time.

Other symptoms of IPF may include fatigue, weakness, chest discomfort, and loss of weight and appetite.

IPF can affect the heart as a complication.

WHAT CAUSES IPF?

Presently, IPF has no known single cause. However, several factors have been associated with the disease:

- Environmental and occupational pollutants, such as organic dust (bacteria, animal proteins), inorganic dust (metal and wood dust, silica), and gases and fumes
- Drugs and poisons, such as chemotherapy medicines, antibiotics (very rare) and radiation therapy
- Infection
- Connective tissue diseases such as rheumatoid arthritis, sarcoidosis and progressive systemic sclerosis

HOW IS IPF DIAGNOSED?

Diagnosis is usually based on two things: first, a careful and thorough study of a person’s medical history, particularly his/her exposures to environmental and occupational pollutants; second, a complete physical examination.

Physicians confirm the diagnosis by:

Chest x-ray. The chest x-ray of someone with IPF would reveal shadows, mostly on the lower portion of the lungs. As well, the lungs would appear smaller than normal. In 10% of patients, however, the chest x-ray may still appear normal despite IPF.

CT-scan. This type of scan provides more detailed information on what is happening inside the lungs.

Pulmonary function test. This test indicates how well the air sacs are functioning and expanding.

Lung biopsy. A biopsy determines how far the disease has progressed (its stage) and give a patient’s medical outlook.

Bronchoalveolar lavage (BAL). This involves the removal and examination of cells from the lower respiratory tract, which will provide information on the kind of inflammatory cells that are present. The process helps exclude other causes of the patient’s symptoms, but is not diagnostic of IPF.

Arterial blood gas. This is a measure of the amount of oxygen and carbon dioxide in one’s blood. Results may be normal or show a reduced blood oxygen level.