Pulmonary Fibrosis, Idiopathic, IPF
Fibrosing Alveolitis, Hamman Rich Syndrome

Idiopathic pulmonary fibrosis is an *inflammation* (soreness and irritation) in the lungs which eventually causes scarring. This usually shows in middle age over a several year time period. The cause is unknown (*idiopathic*). Usually death occurs after several years but there are no time tables which will predict perfectly what the course of the illness will be. It affects males and females equally. In this condition there is a formation of *fibrous* (tough leathery) tissue in the small ducts which carry air to and from your lungs. Because of this, the lungs do not work as well as they should for taking in oxygen from the air you breathe and getting rid of wastes (carbon dioxide). Because the lungs are damaged, there may be more problems with infections or the heart.

Other problems may include *pulmonary hypertension* (high blood pressure in the lungs), and formation of blood clots. Some symptoms of this illness are:

- Coughing and breathing difficulties; cough is usually dry and hacking.
- Bluish (cyanotic) skin and lips due to lack of circulating oxygen.
- Loss of appetite.
- Loss of strength which comes from just the increased work of breathing.
- Rapid, shallow breathing occur with moderate exercise and later even while resting.
- Increasing shortness of breath (dyspnea) which progresses as the disease gets worse.
- Weight loss and fatigue due partly to the increased work of breathing.
- Clubbing of the fingers (the ends of the fingers become rounded and enlarged).

DIAGNOSIS

A diagnosis of idiopathic pulmonary fibrosis may be suspected based on exam and a patient's history.

- Specialized x-rays, pulmonary function tests, pulse oximetry, and laboratory tests including blood gasses help confirm the problem.
- Sometimes a biopsy is done and a small piece of lung tissue is removed. This may be done through a bronchoscope or during an operation in which the chest is opened. This is looked at under a microscope by a specialist who can tell what the lung problem is.

KNOWN CAUSES OF PULMONARY FIBROSIS

If the cause of pulmonary fibrosis is known, it is no longer known as idiopathic. Several causes of pulmonary fibrosis include:

- Occupational and environmental exposures to asbestos, silica, and or metal dusts.
- Illegal or street drug use.
- Agricultural workers may inhale substances, such as moldy hay, which can cause an allergic reaction in the lung. This reaction is called Farmer's Lung and can cause pulmonary fibrosis. Some other fumes found on farms are directly toxic to the lungs.
- Exposure of the lungs to radiation.
- Collagen diseases.
- Sarcoidosis is a disease which forms *granulomas* (areas of inflammatory cells), which can attack any area of the body but most frequently affects the lungs.
- Drugs. Certain medicines may have the undesirable side effect of causing pulmonary fibrosis. Check with your doctor about the medicines you are taking and ask about any possible side effects.
- Some cases of pulmonary fibrosis seem to be genetic.

TREATMENT

- There are no drugs currently approved for the treatment of pulmonary fibrosis. Steroids (a potent medication which cuts down on inflammation) are sometimes given to prevent lung changes before they become permanent. High doses may be recommended at first, followed by lower maintenance dosages. Other

medications may be tried if steroids do not work.

- Lung disease may be monitored with x-rays and laboratory work.
- Oxygen may be helpful if oxygen in the blood is diminished. This improves the quality of life. Your caregiver will give you a prescription for this if it is helpful.
- Antibiotics are used for treatment of infections.
- Exercise may be beneficial.
- Lung transplants are being investigated and a single lung transplant may be considered for some patients.
- Influenza vaccine and pneumococcal pneumonia vaccine are both recommended for people with IPF or any lung disease. These two shots may help keep you healthy.

Document Released: 03/09/2005 Document Re-Released: 06/15/2009
ExitCare® Patient Information ©2011 ExitCare, LLC.