Chapter 1

Idiopathic Pulmonary Fibrosis

The word “idiopathic” means unknown cause, "pulmonary" means “lung” and "fibrosis" means scar tissue. So, idiopathic pulmonary fibrosis (IPF) means scarring in the lungs due to an unknown cause. Over time, scar tissue makes air sacs thicken which makes it harder to pass oxygen into (and gases out of) the blood stream. This results in a decreased level of oxygen in the blood to circulate throughout the body, ultimately affecting all organs.

IPF is a serious disease that most often strikes middle-aged and older adults, but it can also attack younger people. Symptoms vary from person to person. In some, fibrosis happens quickly. For others, the process is much slower. In still others, the disease may stay the same for many years. Many who suffer from IPF have a life expectancy of only three to five years after diagnosis. Genetics may play a role in IPF; if more than one family member has IPF, they are said to have ‘Familial IPF’.

Like most chronic lung diseases, the primary symptoms of IPF are shortness of breath, a dry cough and fatigue. Diagnosis of IPF can be difficult. Using a stethoscope, doctors will listen for a crackling sound in the lungs that sounds like Velcro being slowly pulled apart. Many times people will be wrongly diagnosed with bronchitis or pneumonia, as they are far more common than IPF. A definitive diagnosis is usually obtained via a high-resolution chest CT scan, bronchoscopy and/or a surgical lung biopsy.

At any given time, about 132,000 Americans are living with IPF. A famous example is the popular American daredevil, Evel Knievel, who survived an amazing number of crashes and traumas over his long life, but passed away just five years after being diagnosed with IPF.

Many treatment and therapy options exist for people with IPF. Some of the most widely used options are medications, pulmonary rehabilitation therapy, support groups, lung transplantation, exercise and diet.

As yet, there is no cure for IPF. However, research is making strides in slowing the progression of the disease. Many clinical trials are underway searching for a cure as well as ways to improve longevity and quality of life. Clinical research trials are always seeking both ill and healthy volunteers; if you are interested in participating in a clinical trial, please follow the link below.

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