Delayed access to tertiary care associated with higher death rate from type of pulmonary fibrosis

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Idiopathic pulmonary fibrosis (IPF)—scarring and thickening of the lungs from unknown causes—is the predominant condition leading to lung transplantation nationwide. Columbia University Medical Center researchers confirmed that delayed access to a tertiary care center for IPF is associated with a higher risk of death. The findings were published online in the *American Journal of Respiratory and Critical Care Medicine* on June 30, 2011.

A group led by Columbia researcher David J. Lederer followed 129 IPF patients at an academic medical center. They looked at the length of time from the onset of shortness of breath to the first visit to the center. A longer delay was associated with increased risk of death, independent of age, gender, socioeconomic status, lung capacity, disease severity, type of health insurance, or education. The researchers also found no association between the length of delay and the likelihood of the patient's receiving a lung transplant.

IPF leads to respiratory failure and death, usually within three years. It is a relatively rare disease, which afflicts 100,000 Americans, almost all over the age of 50. Characterized by shortness of breath upon exertion, it is often misdiagnosed, especially in people with other ailments.

A delay in making a correct diagnosis can lead to ineffective, or even harmful, treatments. For example, doctors sometimes still treat IPF with

steroids, because the disease was originally thought to have an inflammatory component. Now scientists know that steroids are counterproductive. A delay in diagnosis can also delay evaluation for a lung transplant. Although research is underway on potential drug therapies, currently lung transplantation is the only effective treatment.

"The initial symptoms of IPF are subtle, and accurate diagnosis may not be feasible for community-based pulmonologists," explains Lederer, Herbert Irving Assistant Professor of Clinical Medicine and co-director of the New York-Presbyterian Hospital Interstitial Lung Disease Program and Lung Transplant Program.

For that reason, earlier access would be aided by improved methods of early detection. But until then, the recognition, or even suspicion, of IPF should prompt referral to a tertiary care center.

Provided by Columbia University Medical Center