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The full report is titled “The Clinical Course of Patients with Idiopathic Pulmonary Fibrosis.” It is in the 21 June 2005 issue of *Annals of Internal Medicine* (volume 142, pages 963-967).

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What Is the Natural Course of Idiopathic Pulmonary Fibrosis?

What is the problem and what is known about it so far?

Idiopathic pulmonary fibrosis (IPF) is an uncommon disease in which the lungs become scarred and stiff for no apparent reason. Patients note shortness of breath and cough, and many eventually die of lung failure. Treatment may include medications to decrease inflammation or further scarring or lung transplantation. How IPF naturally worsens over time is uncertain because the disease is uncommon and many patients receive a diagnosis in more advanced stages of disease. The researchers of this study previously performed a study in which patients with mild to moderate IPF were randomly assigned to take an experimental treatment or a placebo. The study focused on whether patients improved with treatment. However, the researchers realized that observing the patients who took placebo could provide valuable information about what happens over time to patients with untreated IPF.

Why did the researchers do this particular study?

To see how IPF naturally worsens over time in a large number of patients with mild to moderate disease.

Who was studied?

168 patients randomly assigned to receive placebo in a study of treatment for IPF. Most were white men.

How was the study done?

The researchers collected information on patients' lung function and symptoms every 12 weeks from the time the patients enrolled in the study until the study's conclusion (about 1.5 years). They also collected information on the number of hospitalizations and deaths.

What did the researchers find?

Patients' lung function did not generally worsen over the observation period. About one third of patients were hospitalized, many more than once; many hospitalizations were for presumed infection. About 1 in 5 patients died, almost half within weeks of the first worsening of symptoms.

What are the limitations of the study?

The researchers could not provide detailed information about the reasons patients were hospitalized and why they died. The study included only patients who had not previously improved with steroids. The findings may not apply to patients who did not previously take steroids or to those who took steroids and improved.

What are the implications of the study?

IPF does not always slowly worsen over time. Instead, many patients with IPF remain stable for long periods, then suddenly get worse and die. This pattern suggests that patients with IPF who do not improve with steroids should be monitored closely and might possibly undergo lung transplantation while their symptoms are still relatively mild.