
IDIOPATHIC PULMONARY FIBROSIS

THE COURSE OF A DISEASE

KATIE HARRIS

INTRODUCTION

What is Idiopathic Pulmonary Fibrosis (IPF)? The answer can be found by examining the name.

- Idiopathic: Arising from an unknown cause
- Pulmonary: Dealing with the lungs
- Fibrosis: A condition whose hallmark is increased fibrous (scar-like) tissue

So, Idiopathic Pulmonary Fibrosis (IPF) might best be described as a disease of unknown origin that is evidenced by scarring of lung tissue.

Those diagnosed by pulmonary fibrosis are generally between the ages of 50 and 70. While there seems to be no ethnic or racial component to risk, males are at higher risk than females. Also, those diagnosed often have one of several risk factors, such as use of cigarettes or antidepressants, viral infection, or gastroesophageal reflux. ¹ This mysterious disease follows a deadly course which, as of 2008, the medical community could do little or nothing to impede. This paper follows that course, acquainting you with the stages of idiopathic pulmonary fibrosis.

ONSET

A person who is developing IPF will first begin to notice a shortness of breath, a general fatigue, and often a dry, hacking cough which cannot be controlled. ² Most people would simply attribute these symptoms to a lingering cold or bronchial infection, perhaps made worse by an environmental stressor. And most of those people would be correct. Obviously, not everyone who develops a cough or is fatigued is suffering from IPF. After all, there are only 128,000 cases in the United States. ¹ But for those who are these symptoms are not an infection. Shortness of breath for patients with IPF is an indication that the oxygen which they are breathing into their lungs is not able to be easily passed into the blood stream. The cause of this problem is that the interstitial tissue in the lung is becoming scarred. Normal lung tissue is permeable to allow the exchange of gases. There are two main theories as to why this scarring occurs. The first is that the lungs are becoming needlessly inflamed and that this is resulting in scarring. The second is that IPF itself causes some problems with the body's wound repair system. This abnormal wound repair causes the excessive scarring. ³ In these early stages of the disease, pulmonary fibrosis often goes undiagnosed, because people confuse the symptoms with a more common respiratory disease and fail to go to the doctor. This leads to the undiagnosed progression of the disease.

PROGRESSION

As a patient with undiagnosed IPF continues their daily activities, the lack of oxygen circulation begins to become more and more obvious, and the patient will find themselves out of breath during even the most routine activities. Other symptoms often begin to surface as well, such as aching muscles or joints, unexpected weight loss, and a general fatigue. ⁴ Once a patient goes to the doctor, the difficult work of obtaining a diagnosis begins. The symptoms of IPF, even to a trained medical eye, often mimic those of other more common and more treatable respiratory illnesses. To have a conclusive diagnosis, both a high resolution special CT scan and a lung biopsy are needed. ⁵ Once the diagnosis has been made, the only treatments available are to try to prevent more lung scarring and to alleviate the symptoms which are nagging the patient. There is currently no way to reverse lung scarring which had already occurred. For this reason, it is important to obtain a diagnosis as soon as possible in order to prevent lung scarring before it occurs.⁶ The way in which the scarring is slowed is related to the inflammation theory of IPF. The doctor will often prescribe prednisone, an anti-inflammatory. Other accepted medications include immune system repressors such as cyclophosphamide and azathioprine. ⁶ Giving elderly people immune system repressors comes with obvious problems such as an increased risk of serious complications from influenza, even development into life-threatening pneumonia.

PROGNOSIS

From the time of diagnosis, there is nothing that doctors can do to halt this mysterious scarring that is blocking the air flow in the lungs. IPF invariably leads to scarring in the lungs to the point of non function. While the time which this process takes is dependent on many factors, including age at onset, health at onset, and previous risk factors, such as cigarette usage, average survival from the onset of symptoms is 3-6 years.³ As the disease progresses, it can cause problems beyond shortness of breath. These can include high blood pressure in the lungs (pulmonary hypertension) caused by compression of blood vessels by scar tissue, or right sided heart failure, due to the heart having to pump harder to circulate blood through the lungs.⁷ Any of these things can lead to the cause of death. Patients with IPF can die from heart failure, respiratory failure, or collapsed lungs.

THE FUTURE

While the outlook for this disease is currently very bleak, work is being done to ameliorate the lack of knowledge surrounding IPF. One of the most important movements in the medical research community is a shift from the traditional view of IPF as an inflammatory disease. Evidence is mounting that the lung scarring found in IPF patients is not a result of inflammation: the lung scarring characteristic of IPF can be induced in transgenic lab animals without the accompanying inflammation, degree of inflammation in biopsied lungs is not a reliable indicator of IPF progression, and there is no evidence that treatment with anti-inflammatory medication improves at all the prognosis for the afflicted. Given this information, there is now growing support for the theory that underlying IPF is an abnormal system of wound healing in the lungs.⁹ As knowing the underlying cause of a disease is often the first step towards developing a viable treatment, these advances should bring hope to the IPF community.

In addition to basic research on the etiology of IPF, there are also clinical drug trials being conducted, to varying degrees of success. One such study deals with an experimental drug called pirfenidone, and initial results are showing progress in the progression of the disease.¹⁰ Other trials do not fare as well, such as one testing the efficacy of Interferon Gamma-1b, which showed no difference between Interferon Gamma-1b and a placebo.¹¹ However, every trial performed, whether with a positive or negative result, provides more information to scientists about how the disease works and what can be done to treat it. It is imperative that advances continue to be made in the treatment of and knowledge about this disease. The current inability to halt or even slow this pulmonary disease is unacceptable and the need for more research in this area is undeniable. As the search for a cause of the fibrosis, the method of scarring, and a way to halt or even reverse this lung scarring continues, the need for increased funding for medical research will become even more apparent. Those afflicted with IPF and their families can do their part to encourage research by raising awareness in their communities, becoming politically involved in order to let their elected representatives know that there are people who care about IPF, or even become involved in the research field themselves. While encouraging steps are being made, the fight against IPF is far from over, and much work remains to be done.

REFERENCES

1. Coalition for Pulmonary Fibrosis. "Epidemiology and Risk Factors"
<http://www.coalitionforpf.org/Healthcare/riskfactors.asp>. 6 March 2008
2. United States Government. National Institutes of Health: Diseases and Conditions Index. "What are the Signs and Symptoms of Idiopathic Pulmonary Fibrosis?"
http://www.nhlbi.nih.gov/health/dci/Diseases/ipf/ipf_signsandsymptoms.html. 6 March 2008
3. Pulmonary Fibrosis Foundation. "Patient Information Handbook." 1332 N. Halsted Street, Suite 201, Chicago, IL 60622. 2004. <http://www.pulmonaryfibrosis.org/patient.pdf>
4. Mayo Clinic: Diseases and Conditions. "Pulmonary Fibrosis: Signs and Symptoms"
<http://www.mayoclinic.com/health/pulmonary-fibrosis/DS00927/DSECTION=2>. 6 March 2008
5. Mayo Clinic: Diseases and Conditions. "Pulmonary Fibrosis: Screening and Diagnosis"
<http://www.mayoclinic.com/health/pulmonary-fibrosis/DS00927/DSECTION=6>. 6 March 2008
6. United States Government. National Institutes of Health: Diseases and Conditions Index. "How is Idiopathic Pulmonary Fibrosis Treated?"
http://www.nhlbi.nih.gov/health/dci/Diseases/ipf/ipf_treatments.html. 6 March 2008
7. Mayo Clinic: Diseases and Conditions. "Pulmonary Fibrosis: Complications"
<http://www.mayoclinic.com/health/pulmonary-fibrosis/DS00927/DSECTION=7>. 6 March 2008.
8. United States Government. National Institutes of Health: Diseases and Conditions Index. "Key Points of Idiopathic Pulmonary Fibrosis"
http://www.nhlbi.nih.gov/health/dci/Diseases/ipf/ipf_summary.html. 6 March 2008
9. Molsés Selman, MD; Talmadge E. King Jr., MD; and Annie Pardo, PhD. (2001.) Idiopathic Pulmonary Fibrosis: Prevailing and Evolving Hypotheses about Its Pathogenesis and Implications for Therapy. *Annals of Internal Medicine*, Vol. 134, Issue 2, pp. 136-151.
10. Azuma, Arata et al. (2005.) Double-Blind, Placebo-Controlled Study of Pirfenidone in Patients with Idiopathic Pulmonary Fibrosis. *American Journal of Respiratory and Critical Care Medicine*, Vol. 171, pp. 1040-1047
11. Ganesh Raghu, MD, et al. (2004.) A Placebo-Controlled Trial of Interferon Gamma-1b in Patients with Idiopathic Pulmonary Fibrosis. *The New England Journal of Medicine*, Vol. 350, No. 2, pp.125-133.