



finding a cure
Pulmonary Fibrosis Foundation
Through Research, Education and Advocacy

Pulmonary Fibrosis Foundation Newsletter

Volume 5 Issue 1 April 2004

OUR MISSION

The Pulmonary Fibrosis Foundation is dedicated to finding a cure for and raising awareness of Pulmonary Fibrosis, an often terminal disease. The Foundation is devoted to improving the quality of life for those afflicted with Pulmonary Fibrosis. The Foundation provides hope for patients through funding and supporting research, education and advocacy.

PRESIDENT'S MESSAGE



Our progress during the years 2003-2004 represents an expansion of our mission. We are proud of our achievements. We look forward to increasing our efforts until we can say that a cure for Pulmonary Fibrosis has been found.

Young Investigator Awards Program

- Awarded \$35,000 to Dr. Yingze Zhang of the University of Pittsburgh for research on "The Genetic Predispositions to Pulmonary Fibrosis"
- Awarded \$ 50,000 to Dr. Pyong Woo Park of the Baylor College of Medicine for research on the "Proteoglycans in Lung Inflammation and Fibrosis. (Co-funded with the American Lung Association).
- Awarded \$45,000 to University of Vermont, Burlington, Vermont. Awarded to Navdeep Singh, MBBS and Baylor College of Medicine, awarded to Dr. Joseph N. Walter. Co-funded with the American College of Chest Physicians for research in Pulmonary Fibrosis.
- Awarded \$50,000 to Brigham and Women's Hospital, Boston, Massachusetts, Dr. Caroline A. Owen. Co-funded with the American Thoracic Society for research in Pulmonary Fibrosis.

Conferences Funded

Awarded \$ 15,000 to the American College of Chest Physicians to fund a Conference on the Early and Accurate Diagnosis of Pulmonary Fibrosis held on March 22, 2003. The Conference was attended by 35 of the leading researchers in the United States. A permanent Network for the Early and Accurate Diagnosis of Pulmonary Fibrosis was established at the conference. The goal of the network is to provide physicians with the necessary tools to accurately diagnose this illness. Our plan is to establish a National Screening program with an early warning system so that doctors may initiate treatments long before it progresses to the advanced stage where patients are beyond medical help.

Other Achievements

- The American Thoracic Society awarded the Pulmonary Fibrosis Foundation its Public Service Award in recognition of service in public health in the area of lung-related health issues.
- The Pulmonary Fibrosis Foundation is represented on the Health Care Policy Committee of the American Thoracic Society.
- The Pulmonary Fibrosis Foundation is a founding member of the Rare Lung Disease Consortium.
- The Foundation is a founding member of the Public Advisory Roundtable of the American Thoracic Society.
- The Foundation is a founding member of the Early and Accurate Detection of Pulmonary Fibrosis Network.
- We have convinced the National Heart, Lung and Blood Institute of the NIH to initiate two studies in the area of Pulmonary Fibrosis.
- As a result of the Foundation's influence, National Heart, Lung and Blood Institute is establishing a National Network for Pulmonary Fibrosis Clinical Research. This is a multi-centered program which will design and perform multiple therapeutic trials for treatment of Pulmonary Fibrosis Patients.
- In addition, the NHLBI has established the Lung Tissue Resource Consortium to increase understanding of the pathogenic mechanisms of Pulmonary Fibrosis.

Achievements for the first quarter of 2004

- Awarded \$20,000 to Duke University to Dr. David Schwartz to study the genetics of Pulmonary Fibrosis. Co-funded with the William E. Simon Foundation.
- Participated in a Conference which examined the incidence, causality and genetics of Pediatric Interstitial Lung Disease. One of the targets of this conference is to develop a network and a partnership between the Foundation and the PILD research community.
- The same conference discussed the parameters and research efforts of the Rare Lung Disease Network.

Michael Rosenzweig, Ph. D., *President and CEO*

Research Issues

Patients suffering from Pulmonary Fibrosis desperately desire effective treatments for this disease. Unfortunately, at this time a successful treatment is still unavailable.

Why does research take so long?

In order to obtain FDA approval for a new drug, it must successfully complete the phases described below. Each Phase takes about one year to complete. Clinical trials are conducted in a series of steps, called phases - each phase is designed to answer a separate research question.

- Phase I: Researchers test a new drug or treatment in a small group of people for the first time to evaluate its safety, determine a safe dosage range, and identify side effects.
- Phase II: The drug or treatment is given to a larger group of people to see if it is effective and to further evaluate its safety.
- Phase III: The drug or treatment is given to large groups of people to confirm its effectiveness, monitor side effects, compare it to commonly used treatments, and collect information that will allow the drug or treatment to be used safely.
- Phase IV: Studies are done after the drug or treatment has been marketed to gather information on the drug's effect in various populations and any side effects associated with long-term use. (<http://clinicaltrials.gov/info/resources>)

New Studies by Drug Companies

After extensive lobbying on our part, five drug companies have initiated studies involving the development of new drugs with which to treat Pulmonary Fibrosis. These are: Wyeth - Enbrel, an Anti-TNF antibody.

Actelion - Bosentan, a Anti-Endothelin inhibitor.

Genzyme - An Anti-TGF beta, a broad spectrum, monoclonal antibody.

Novartis - Gleevec, an Anti-TNF antibody.

Pfizer - Viagra, a blood vessel dilator.

Studies needing patients

Safety and Tolerability Study of FG-3019 in Patients with Idiopathic Pulmonary Fibrosis Sponsored by FibroGen, Inc.

The purpose of this study is to evaluate the safety and tolerability of FG-3019, a therapeutic antibody designed to block the pro-fibrotic activity of connective tissue growth factor (CTGF). CTGF triggers the production of collagen and fibronectin, which cause scarring and thickening of the lungs. Approximately 18 to 27 males and females, 21 to 80 years of age with a diagnosis of idiopathic pulmonary fibrosis (IPF) will be enrolled in this study. The duration of the study is approximately one month, during which patients will receive a single infusion of FG-3019. In addition, there will be two follow-up visits 6 and 12 months after receiving the study drug.

Condition Treatment or Intervention Phase: Idiopathic Pulmonary Fibrosis Drug: FG-3019 Phase I (<http://clinicaltrials.gov/>)

Efficacy and safety of oral bosentan in patients with Idiopathic Pulmonary Fibrosis. Sponsored by Actelion

Endothelin-1 (ET-1) is expressed in a variety of pulmonary pathological conditions including pulmonary vascular disease and pulmonary fibrosis. Bosentan (an oral dual ET-1 receptor antagonist) could delay the progression of idiopathic pulmonary fibrosis (IPF), a condition for which no established treatment is available. The present trial investigates a possible use of bosentan, which is currently approved for the treatment of symptoms of pulmonary arterial hypertension (PAH) WHO class III and IV, to a new category of patients suffering from IPF.

Condition Treatment or Intervention Phase: Idiopathic Pulmonary Fibrosis Drug: bosentan Phase II Phase III (<http://clinicaltrials.gov/>)

The safety and efficacy of etanercept in comparison with placebo. Sponsored by Wyeth-Ayerst Research

The objective of the study is to evaluate the safety and efficacy of etanercept in comparison with placebo in a double-blind, parallel, randomized fashion in subjects with idiopathic pulmonary fibrosis (IPF) who failed previous therapy. The treatment period will be up to 1 year. The primary objective is evaluation of safety and efficacy. Secondary: The secondary objective is to evaluate quality of life (QoL) and pharmacokinetics (PK).

Condition Treatment or Intervention Phase: Pulmonary Fibrosis Drug: Etanercept Phase II (<http://clinicaltrials.gov/>)

Research on Familial Pulmonary Fibrosis

Jennifer A. Galvin, M.D.

One of the critical aspects to understanding the pathology of pulmonary fibrosis (PF) is to understand the genetic components of this disease. Last year, I had a rewarding experience using my molecular biology background to study the genetic basis of PF with Dr. David Schwartz at Duke.

The Schwartz lab is one of three NIH-funded labs studying familial PF. This study includes over 400 families. We were looking for regions within the genetic material (chromosomes) that were mutated in the PF families. During my three months in the lab, we designed a way to look at and compare gene expression in the genetic material of PF families. We observed that certain regions within the genetic material (chromosomes 10 and 13) showed potential gene mutations in many, but not all, of the PF families.

These findings were presented at a conference in December and a paper is currently being written.¹ From my experience at Duke, I learned the importance of the applicability of clinical medicine at the laboratory bench. Moreover, being a member of the PF research team was inspiring. The dedication and commitment of the physician/scientists, post-doctoral researchers, and pulmonary fellows was evident in every aspect of the project's many demands.

¹ Ivana V. Yang, Lauranell H. Burch, Jennifer A. Galvin, Mark P. Steele, Jordan D. Savov, David A. Schwartz. *Gene Expression Analysis of the Idiopathic Interstitial Pneumonias*. National Institutes of Environmental Health Sciences (NIEHS) Toxicogenomics Research Consortium (TRC), Seattle, WA, December 7-8,th 2003.

Living With Pulmonary Fibrosis

By Suszette McKay, R.N. B.S.

Living with Pulmonary Fibrosis. What a concept. In April, 2001 I didn't think that was a possibility. You know, we've all heard the words: Lung scar, no treatment, no cure, terminal. Now, I've been a nurse for what seems like an eternity. I've been on both sides of the bed. I've been the one bearing bad news. When it was "my turn" I was just like everyone else, in shock. We all have heard the words. Lung scar. No treatment. No cure. Terminal. "They" were confused. I was confused.

It is now 3 years later. Life goes on and here I am. How'd I do that?

Dealing with a chronic illness, of any kind, on a daily basis is a challenge. Our personality, the way we view the world and cope with problems will color the way we cope with chronic illness.

I've had to make some adjustments in the last three years, due to that "breathing problem" I have going on. I have to use oxygen now. I've dealt with being the "Poster Child for the Smiley Face" thanks to prednisone. I lost my hair and wore wigs (4 different lengths and colors just to keep people guessing) thanks to Cytosan. I left a very busy full time consulting business and had to ponder if I was defined by my career or if my career was defined by me.

I've adjusted, but in reality, I live my life the way I always did. I've always dealt with road blocks head on. I am an optimist. I believe in "I can." I believe that if "I wish I could," then I should. I believe in God. I believe that life is really funny, and I don't want to miss out on any of the fun.

The down side to carting oxygen around is "carting oxygen around." It's heavy, cumbersome, tubes get tangled, I'm forever getting unplugged, I step on my tube and nearly rip out my nose, it dries out my nose, pulls on my ears and is noisy in theaters. However, it provides me the opportunity to be active, see my friends and family and be independent.

You see, I don't have a choice regarding my disease, but I have a choice about my attitude. As soon as I open my eyes I can decide. Do I want to be mad, sad, or glad? Do I want to be negative or positive? I like positive.

Foundation Staff

Michael Rosenzweig, Ph. D., President & CEO
Traci Toutant, PHR, Executive Director
Jennifer Bulandr, Director of Media & Community Relations
Leanne Storch, Executive Assistant

Pulmonary Fibrosis Foundation Letter Writing Campaign

The Foundation attributes much of its fund-raising success to its patient and family community. Many of you have asked how you can help raise funds for the Pulmonary Fibrosis Foundation. Consider participating in a letter writing campaign. The time and cost commitment is minimal on your part, but the benefit to the Foundation is remarkable!

Write your own personal story about your connection with Pulmonary Fibrosis – friend, patient, family member, etc., include a donation form and send it out to your network of contacts. Once donations are received, you will receive an acknowledgment of the donation and we will thank the generous donor for their tax-deductible contribution. Your participation in raising funds for the Pulmonary Fibrosis Foundation will have a direct impact on the effectiveness of our mission to find a cure for Pulmonary Fibrosis. For more information, please visit our website at www.pulmonaryfibrosis.org and select the Fund Raising tab on our home page. You can also contact us at 312.377.6895.

Fund Raising Events!

We have planned a series of events that not only will raise funds but will also provide inspiration, hope and a sense of participation in an activity that advances a good cause.

Board Chairman to compete in Ironman Race

Daniel Rose M.D., Chairman of the Board of the Pulmonary Fibrosis Foundation, will be participating in the Coeur d'Alene Ironman competition to raise money for the Pulmonary Fibrosis Foundation as part of the Janus Charity Challenge. The event will take place on June 27, 2004. An Ironman competition involves a 2.4-mile swim, 112-mile bike-ride and a 26-mile run. Please support this incredible feat by making a contribution today! Donations, go to: www.active.com/donations/fundraise_public.cfm?key=D0pff

The New York State Pulmonary Fibrosis Foundation.

First annual "Drive to a Cure" Golf & Reception Benefit on Sunday, August 22nd at the Central New York Valley View Golf Club, Utica, NY. This event was established by the family and friends of Joseph F. Maltese, Sr., of Utica, who died from Pulmonary Fibrosis at the age of 66 on October 25, 2003.

A brand new Honda will be given to a person if he/she gets a hole-in-one on the 18th hole. Great prizes for best score, worst score, longest drive, closest to the hole. Putting Contest. 50/50 Raffle. Numerous randomly drawn prizes. Band and/or DJ. Possible special 'local celebrity' guests, former Major League baseball player(s). Hole Sponsorships (\$100). Golf and Reception \$67, includes 18 holes of golf, cart, lunch and dinner including beer, wine and soda and entertainment. Reception-only \$29, includes dinner, beer, wine, soda and entertainment and a chance to win the randomly-drawn prizes. Even if you'll be unable to participate, donations would be appreciated. Please let others know about it.

Headquartered at 450 Oakdale Avenue, Utica, NY 13502, 315-724-6512.

Yes, Sign Me Up! I want to support the work of the Pulmonary Fibrosis Foundation and would like to make a contribution.

Here's my check for: \$5,000 \$1,000 \$500 \$100 \$50 Other_____

Name_____

Address_____

City_____ State_____ Zip_____

Phone_____ Email_____

Please send your check to: Pulmonary Fibrosis Foundation, 1440 W. Washington Blvd., Chicago, IL 60607

If you would like to charge your Contribution: Visa MasterCard Discover American Express

Account Number_____ Expiration Date_____

Credit Card Contributions may also be made by Phone: (312) 377-6895 or Fax (312) 377-6896

Published quarterly by the **Pulmonary Fibrosis Foundation**, 1440 West Washington Blvd., Chicago, IL 60607
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