

Breathe Bulletin



A Quarterly Newsletter of the
Pulmonary Fibrosis Foundation
Volume 7 Issue 4 - December 2007



President's Message

As 2007 draws to a close we would like to thank all those who have made financial contributions to enable us to conduct research towards finding a cure for Pulmonary Fibrosis. We also thank those who have written to their representatives and senators asking for additional funds for research and to the many others who in their small ways have promoted and advocated the cause of Pulmonary Fibrosis.

We realize that many of you have lost loved ones to this terrible disease during the past year and the Foundation wishes to express its sincerest condolences at the passing of your loved ones. I have lost a brother and sister to Pulmonary Fibrosis so I share your sorrow on a very personal level. Consequently, the Foundation is totally dedicated to finding a cure so that we do not continue to lose more than 40,000 individuals per year to this disease. In 2006, 88% of our expenditures went to our mission.

On the positive side, the research at our University of Chicago Center of Excellence is making significant progress. The University of Michigan has identified the factors which cause the lung scarring. Duke University has identified the genes and areas of the chromosomes which are responsible for the development of Pulmonary Fibrosis. Finally, researchers have begun to work with stem cells in the hope of creating new healthy lung tissue. 2008 may be the year that we see a breakthrough in a successful treatment program for Pulmonary Fibrosis.

I would like to take this opportunity to personally wish all of you the happiest of holidays and a new year in which all of our dreams and aspirations come true.

Michael Rosenzweig, Ph.D.,

President and CEO

1 **President's Message**

2 **Julie W. O'Connor
Board Members**

3 **Advocacy**

4 **Pulmonary Fibrosis**

5 **Current Research**

6 **Ways to Donate
Donation Form**

7 **Calendar of Events**

8 **To lung biopsy or not**

Disclaimer

The material contained in this newsletter is for educational purposes only and should not be considered as medical advice. Always consult your health care provider for treatment options.

Welcome to our newest Board Member Julie Willis O'Connor



I am a 50 year old married mother of 3 children. (ages 18, 16 and 13). I have been married to Dan O'Connor for 21 years. We have lived in Concord, MA for the past 16 years. Since the birth of my oldest child, I have been a stay at home mom. Currently I am on the Parent Executive Board at Lawrence Academy where my son is a senior. I am a active person who has run in several 6 and 10K races. I ran the Boston Marathon in 1996. I have played competitive team tennis in a Div I league in the greater Boston area for the past 8 years. I have been an avid hiker for the past 10 years.

I have climbed all 46 mountains in

the Adirondacks Park in honor of my father, Bo Willis who has Pulmonary Fibrosis. I was born and raised in Baltimore MD. I graduated from Garrison Forest School and went on to attend college at Tulane University and ultimately graduated from St. Lawrence University in Canton, NY. I also received an Associate of Applied Science form Parson's School of Design and worked in the interior design industry before having my children.

Professional Experience

Full time mom 1989 to date...
 Managing Partner, RHIVLB, LLC (residential real estate development and management)
 Partner, Willis Investments (commercial real estate management)
 Peabody Office Furniture Interior designer

Education

Tulane University
 St. Lawrence University BS Parson School of Design AAS

Activities

ADK 46r's (August 2007)... has climbed top 46 peaks over 4000 feet in Adirondack park Team Tennis Division I in DBH Tennis league past 6 years Boston Marathon 1997

Community Service

Advisory Board, Lawrence Academy, Groton, MA
 Carroll School Parents Assoc. Lincoln, MA

Julie has raised over \$61,000 in her 46r's event

We enthusiastically welcome her to our Board of Directors

If you are interested in joining the Foundation as a Board Member, Event host, Advocate or Volunteer, please call us: 312-587-9272

Foundation Staff

Michael Rosenzweig, Ph.D.
President and CEO

Leanne Storch
Executive Assistant and Patient Advocate

Vakarie Roberts
Director of Development

Myrrick Liontonia
Database Administrator

Monica Storch
Administrative Assistant

Medical Advisory Board

David Kamp, MD – Medical Director
 Kevin Brown, MD – Chairman
 Marvin I. Schwarz, MD – Past-Chairman
 Rany Condos, MD
 Roland M. du Bois, MD
 Gary W. Hunninghake, MD
 Naftali Kaminski, MD
 Joseph Lasky, MD
 Geoffrey Laurent, Ph.D.
 Joseph Lynch, MD
 Albert Niden, MD
 Imre Noth, MD
 Ralph J. Panos, MD
 Ganesh Raghu, MD
 Jesse Roman, MD
 Moises Selman, MD
 Charlie Strange, MD
 Robert Strieter, MD
 Jacob Iasha Sznajder, MD
 Galen Toews, MD

Board of Directors

Daniel Rose, M. D.
Board Chairman
 Michael Rosenzweig, Ph. D.
President and CEO
 Joseph Borus, Esq.
Secretary
 Daniel Beren
 Leslie Bull
 Nicol Corbin, M. D.
 Gregory Davis, Esq.
 Jennifer A. Galvin, M. D.
 Thomas Hales
 Evel Knievel
 Heather Leverone
 Joseph Maltese
 Julie Willis O'Connor
 Susan Rattner, M. D.
 Nancy Rodriguez
 Carl Salzano
 Thomas Terrill

Advocacy

Individual States support Stem Cell Research

States have taken on an extremely important role in advancing the field of stem cell research in the United States. For years, many of our country's leading scientists have had their progress greatly limited by the lack of resources and funding made available by the federal government in support of some of the most promising areas of this research. State efforts from coast to coast are helping to refuel that progress, and the hope of millions of Americans.

In 2005, more than 170 bills on stem cell research (both for and against) were considered by states across the country. With some of that legislation carrying over into this year, and many new bills planned, the National Conference of State Legislatures expects stem cell research to be one of the top ten state legislative issues in 2007. Already, significant progress has

been made on the state level. In 2004, California voters passed a landmark proposition that will provide nearly \$3 billion for stem cell research funding in California over the next ten years. In December 2005, New Jersey announced \$5 million in stem cell research grants, making it the first state to use public money to fund stem cell research. A growing coalition in Missouri is gathering support for an initiative to amend the state constitution to ensure that stem cell research, therapies and cures permitted by federal law will be allowed in the state.

Despite important steps forward, ensuring the advancement of this vital area of research remains a substantial challenge in many states. For example, Arkansas, Indiana, Iowa, Michigan, North Dakota and South Dakota have banned both therapeutic and reproductive cloning for any purpose, including research.

This means that scientists in these states are unable to pursue areas of stem cell research that are believed to have the greatest potential for developing treatments and cures. South Dakota has banned all forms of stem cell research.

Still, recent state and national polls indicate that a growing majority of voters believe in the promise of stem cell research and favor public support for it. This makes the tremendous work that states are doing to help secure the future of this research even more important.

**Write to your state
Senators and
Representatives
asking them to
support stem cell
Research**

Sample Stem Cell Letter

For much too long Congress and the medical community have turned a blind eye on the devastation wrought by idiopathic pulmonary fibrosis (IPF). It has been a disease that has received little attention. It is a progressive, incurable scarring and irreversible loss of lung tissue resulting in its consequential inability to transport oxygen. Neither its cause nor a cure is known. IPF is not as rare an occurrence as many may be led to believe.

Currently, there are approximately 200,000 Americans subjected to this disease with 48,000 new cases diagnosed each year. Two-thirds of IPF patients die within five years of diagnosis, and approximately 40,000 of them die each year. In my

case alone, I had two good friends who died with this disease and now I, too, have finally fallen victim.

According to researchers at the University of Colorado, between 1992 and 2003, the age-adjusted mortality rate of IPF rose nearly 28.4% in men and 41.3% in women. And they predict the rate will continue to rise.

However, Rice University is now planning to conduct human trials of a blood protein that has had some success at preventing fibrotic disease from developing in the hearts and lungs of lab animals. Gene therapy may eventually hold some promise as an effective treatment whenever a safe and efficient delivery system is found, but this is still

somewhere in the indefinite future.

Another, perhaps more proximate, treatment would be the use of stem cells to grow new lung tissue. However, this avenue has been strictly limited by the proscription of federal funding for research when using new embryonic stem cells. This regretful situation should be rectified as soon as possible. Senator Arlen Specter has offered an amendment to the 2008 Appropriations Bill to increase funding for lung research, particularly with respect to IPF, by the NHLBI. More specifically, the NHLBI is urged to convene a conference of experts in lung disease and other stakeholders to lay the groundwork for a Pulmonary Fibrosis Disease Action Plan for prevention and control of this lethal disease.

Pulmonary Fibrosis

What are the causes of Pulmonary Fibrosis

The process is one of gradual replacement of the lung tissue with fibrosis or scarring, resulting from some type of injury to the lung. In the past, the predominant theory was that this process began with inflammation which resulted in scar formation. However, it has recently been proposed that fibrosis itself, representing abnormal wound repair, is the primary process rather than inflammation, particularly in IPF.

Fibrosis or scarring sometimes can be linked to particular causes

such as prolonged exposure to occupational or environmental contaminants or dusts. This can be due to inorganic dusts such as asbestos, silica, beryllium and hard metal dusts or organic dusts such as bacteria and animal proteins, which is also called Hypersensitivity Pneumonitis. Hypersensitivity Pneumonitis (or allergic alveolitis) is an allergic disorder caused by the inhalation of organic dusts. In some instances, an acute toxic reaction may occur at the time of exposure to a large dose of spores from a microbe, or within days, weeks or a few months. The acute reaction is generally in the form of bronchitis or asthma.

However, in most cases, a large

cumulative exposure of moderate to high levels of the contaminant is necessary over several years (10-20 years) for Pulmonary Fibrosis to develop. Genetic Predisposition is becoming recognized as playing an important role.

Fibrosis is also associated with auto immune diseases such as Rheumatoid Arthritis, Scleroderma or Lupus and can be caused by drugs or certain treatments, such as antibiotics (nitrofurantoin, sulfasalazine), antiarrhythmics (amiodarone, propranolol), anticonvulsants (phenytoin), chemotherapeutic agents (methotrexate, bleomycin) and therapeutic radiation.

[Patient Handbook](#)

Seasonal Health Concerns for Patients

As the cold and flu season approaches, the Foundation would like to remind patients of some small measures they can take to stay healthy.

- Get a flu shot
- Make sure your pneumonia shot is up-to-date
- Avoid overly crowded environments where there may be an abundance of germs
- Ask friends and relatives to call instead of visit if they suspect they may be ill
- Wash hands frequently

See a doctor immediately if you begin to experience any unusual symptoms.

NEWS ABOUT FAMILIAL PULMONARY FIBROSIS RESEARCH SIGNIFICANT SCIENTIFIC PROGRESS

We are very excited about the results we have obtained. We have identified multiple chromosome regions that point to the location of pulmonary fibrosis genes. The highest risk gene is located on the tip of chromosome 11. There are several lung-related genes in this region, and we are intensely studying these genes for abnormalities in our patients. We have also identified regions on chromosome 5, 10, and 12. We anticipate there will be several different genes that cause pulmonary fibrosis. We wish to thank the individuals and their families who have participated in our research study. It is only because of families

like yours that this research can continue to progress. During the next year, we will extensively test chromosome regions 10 and 11 to

- Identify individuals who are at risk of developing pulmonary fibrosis;
- Treat people in the early, potentially reversible stages of their disease: and

Discover new treatment for this form of life-threatening lung disease.

[Duke University](#)

Please remove me from your mailing list

Name _____

Address _____

City _____

State _____ Zip _____

Current Research

Update on Pulmonary Fibrosis research at the University of Chicago

As many of you know the University of Chicago was awarded a large grant by the Pulmonary Fibrosis Foundation to accelerate their work in translating their bench to bedside research in the area of pulmonary fibrosis research. As you can see from the last letter, the area of genetics and genomics in the study of IPF is an exciting and very productive route of investigation which the U of C is world renown. It is our hope that exploring the etiology of this devastating disease with novel techniques and strategies will lead to new thinking in applying potential treatments.

We have divided our attack into three parts. The first involves the development of a clinical core to help “flush” out the type of data which characterizes our IPF patients. This “phenotypic data” helps us understand how the disease is expressed differently from person to person with IPF and how it may differ from other interstitial lung diseases. The second approach involves developing an animal model for IPF which will allow us to test new therapies as they are developed. Last but certainly not least, the third project involves looking at the genetics and genetic expression in people with IPF. This involves novel technologies using chips to explore thousands of genes all at once. This is a very different approach; the classical hypothesis driven approach to most science in that we

know there will be differences, the issue is in discovering which ones. This “scattergun” approach allows for much more rapid progress than the classical approach of exploring only a few “target” genes.

We are happy to report that we have accumulated enough samples to begin some meaningful analyses this fall and hope to have preliminary results to share with you in the coming months. This is actually well ahead of the target we outlined in our proposal and is very exciting. It means that once we get results that we will have time and resources to continue on this path and hopefully continue to hone in on a targeted approach to the disease. As discussed in the last news letter, there has been ongoing work involving identifying people with IPF of an accelerated nature. It is our hope that our study will further identify which groups might be at risk for the expression that leads to progression and death.

We hope you share our excitement in these projects.

Joe G.N. Garcia, M.D.

Imre Noth, M.D.

Uncovering the Mechanisms Underlying Lung Scarring

ScienceDaily (Nov. 17, 2007) — Pulmonary fibrosis is an incurable disease where the lung becomes scarred due to pathologic accumulation of fibrous scar tissue. Telomerase is a protein most notable for its connections to

aging and cancer, but it has also been shown to have increased activity in mice with lung fibrosis.

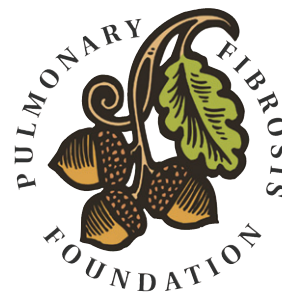
New evidence provided by Sem Phan and colleagues from the University of Michigan Medical School, Ann Arbor, has identified a role for telomerase in the progression of this disease in mice.

Telomerase activity is dependent on the presence of a related protein named TERT. The authors studied the effect of this enzyme in mice with induced lung injury and subsequent fibrosis. They found that mice with reduced TERT levels also had reduced telomerase activity in the lungs, and less severe lung fibrosis. This effect was reversed if the TERT-deficient mice were transplanted with TERT-sufficient bone marrow cells prior to lung injury.

Conversely, when normal mice received bone marrow cells from TERT-deficient donors, subsequent telomerase activity and fibrosis of the lung was reduced.

From these results, the authors concluded that bone marrow cells expressing TERT are important in the development of pulmonary fibrosis.

Adapted from materials provided by Journal of Clinical Investigation (2007, November 17).



Various ways to donate

The Foundation frequently receives calls about the various forms of donations. Below is a clarification how we organize donations.

General Donations

A general donation is any gift made to the Foundation in the form of cash, check or credit card payment. These can be general or in honor or memory of a loved one. If the donation is made in someone's honor or memory, please include a short note with the person's name and the names and addresses of any family members who you would like to be notified of the donation.

Greatest Need vs Research

Any donation made to the Pulmonary Fibrosis Foundation can be restricted for research or used where the need is the greatest. A Research Restricted

donation means that it cannot be used for any administrative costs. A Greatest Need donation means that it can be used for overhead costs such as postage, office equipment, and publications. In order for a donation to be restricted, it must be acknowledged that it should be. To do so, attach a small note to the donation or check the appropriate box if using a Foundation form. The exception is those donations made to an established memorial fund, which are always restricted for research.

Memorial Funds

While donations made in a loved one's honor or memory are welcomed, they may not necessarily be considered memorial funds. A memorial fund is established when the amount donated in a person's memory reaches \$10,000. This can be done either by an initial donation of that amount, or it can be

accumulated over time. Families may request that the Foundation regularly update them with donation totals in order to monitor progress toward a memorial fund. Memorial funds are 100% restricted for research.

Matching Gifts

The Pulmonary Fibrosis Foundation accepts matching gifts from any company willing to join its employees in the fight against Pulmonary Fibrosis. When you make a donation to the Foundation, be sure to check with your employer to see whether a matching gifts program is offered. Donations can also be made in the form of securities, wills and bequests, annuities, and remainder trusts. For more information on these or any forms of donations, contact the Foundation at 312.587.9272. As always, the Foundation greatly appreciates all gifts regardless of size.

Yes! I want to support the work of the Pulmonary Fibrosis Foundation by making a contribution.

Donation Amount: \$5,000 \$1,000 \$500 \$100 \$50 Other _____

Restrict my donation for Research Use my gift where the need is greatest

Name _____

Address _____

City _____ State _____ Zip _____

Phone _____ E-mail _____

Please send your check to:
Pulmonary Fibrosis Foundation,
1332 N. Halsted Street, Suite 201,
Chicago, IL 60622

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) 587-9272 or Fax (312) 587-9273

50/50 Calendar

The 50 Events in 50 States Fundraising Campaign creates awareness of pulmonary fibrosis and raises funds for research. These fundraising events are vital to our success. More information can be found at: www.pulmonaryfibrosis.org/events.htm. To host an event call (312) 587-9272

By press time 27 events have been completed

California

- 6th Annual Bernice F. Dunlop Golf Tournament hosted by her daughter Jasmin Powell
- "An evening in Sonoma and Napa Valley" Winetasting event hosted by Kathryn Smith in memory of Wilbur Smith on September 5th

Connecticut

- Jon Allard ran a half marathon on June 24th in memory of Ann Urciuoli while raising funds for research

Georgia

- John Wade Event - Concert performed by the Dread Clamptitts - Hosted by Jim Vianneau

Illinois

- Susan Heizer conducted a letter writing campaign to raise funds for the Foundation in honor of her sister Diana Larson
- Erin Parizek is hosting an on-going Hearts and Crafts Workshop for kids in honor of her dad Dan Fitzgerald

Massachusetts

- 3rd Annual Leverone/O'Leary "drive to a cure" golf tournament planned and organized by the O'Leary and Leverone families on October 1st
- Doug Bernard and family hosted a fundraiser in memory of his mother, Judith A. Bernard and his Aunt Cheryl Williamson on April 21st
- CARS4ACURE - Car Show hosted by Doug Reinbold for Tom Mournighan on June 9th

Michigan

- 3rd Annual "Paddle out Pulmonary Fibrosis" canoe outing in memory of the Dery and Willacker families on August 4th organized by Matt & Fred Dery

Missouri

- "Washers for Wellnes" - In memory of Hank Hopfinger hosted by Jen Etling. www.washersforwellnes.com

Montana

- Evel Knievel days in Butte, July 26, 27 and 28th

New Hampshire

- Cathleen Brown completed a half marathon in honor of her mother Patricia Vaudreuil while raising funds for research on April 1st

New Jersey

- "A Breath of Fresh Air" cocktail party hosted by Sandra Lewis in honor of her mother Judy Bean on May 11th in Princeton
- Marilyn Fread participated in the NJ Marathon in memory of her good friend Bob Schmuldt

New York

- 3rd annual wine tasting hosted by Arizona State University Alumni in memory of Jim Stephens held on January 18th

Completed events continued

- Texas Hold 'Em Tournament on June 23rd hosted by Mary Jane Borst in memory of her mother, Georgia Jean Williams Gorton
- "Love to find a cure" Tennis tournament hosted by Nancy Feldman on June 23rd in memory of her mother Ruth Lang
- 4th Annual "Drive to a Cure" in memory of Joe Maltese Sr. on August 19th
- Team Toby - Ongoing fundraiser in memory of Toby Wilgoren - www.teamtoby.net
- Julie Willis O'Connor has climbed all 46 of the Adirondack Park's highest peaks in honor of her father Bo Wills who suffers from Pulmonary Fibrosis. (see article on page 2)

Ohio

- "Comedy Rocks" Benefit held on January 19th and 20th in memory of Fred Leeds
- Lawrence Bray memorial barn sale hosted by Vicki Mountain
- Charity Golf Scramble in memory of Patricia J. Haueter
- Team Poleon - Race for Hope. Christine Poleon is running her 1st Marathon in honor of her father Robert Poleon

Pennsylvania

- 2nd annual benefit bike run and picnic hosted by Rob Fiorillo and family in memory of Barbara A. Fiorillo on June 30th
- 3rd Annual Wescoe Walk planned by Jennifer Wescoe-Schaninger - October 27th

Texas

- Fathers's Day Concert held in memory of Greg Miller hosted by Janan Miller and family and friends on June 17th
- Fundraising calendar sent out to family and friends in loving memory of Lisa Herschelman by her husband, Randy and family and friends

Total raised to date from 50 in 50 events - \$294,095

**If you would like to host an event,
please call the Pulmonary Fibrosis
Foundation at:
312-587-9272**

Ask for the Fundraising Handbook.

To Lung Biopsy or NOT to Lung Biopsy,

-that is the question.

The Pulmonary Fibrosis Foundation receives many calls about whether or not getting a lung biopsy is a good idea. Unfortunately, we hear of too many families who have lost a loved one die right after biopsy. But, we also hear that a patient didn't get an accurate diagnosis until a biopsy was performed. It

is a personal decision, one that should be made after educating yourself about biopsies.

OLB – Open Lung Biopsy – After a general anesthetic is given, the surgeon makes an incision in the skin on the chest and surgically removes a piece of lung tissue. This procedure requires a hospital stay of 3-7 days.

VATS – Video Assisted Thoracic Surgery – After a general anesthetic is given, an endoscope is inserted through the chest wall into the chest cavity. Various types of biopsy tools can be inserted through the endoscope to obtain lung tissue for examination.

BRONCHOSCOPY – is a

diagnostic procedure in which a pulmonologist inserts a bronchoscope into the trachea, main stem bronchi and some of the small bronchi. This procedure is typically used to take samples. It is the least invasive of the three biopsies.

WE WOULD LIKE YOUR STORY.

If you've had any of the above procedures and had a good experience or a bad experience, we would like to hear your story.

Please email to: pulmonaryfibrosisinfo@yahoo.com or send via mail to Leanne Storch, PFF, 1332 N. Halsted, Suite 201, Chicago, IL 60622.



Pulmonary Fibrosis Foundation

1332 N. Halsted Street, Suite 201
Chicago, IL 60622-2691
phone: 312.587.9272 fax: 312.587.9273
e-mail: pulmonaryfibrosisinfo@yahoo.com
web: www.pulmonaryfibrosis.org

Nonprofit Org.
U.S. POSTAGE PAID
Oak Brook, IL
PERMIT No. 100