

Breathe Bulletin



A Quarterly Newsletter of the
Pulmonary Fibrosis Foundation
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President's Message

After years of frustration with research studies that failed to find an effective treatment for pulmonary fibrosis, there finally seems to be a light at the end of the tunnel with many of the latest research projects showing positive results. Although these studies may be in their preliminary stages, they are indeed promising. The other important development is the fact that there has been a significant increase in the quantity of pulmonary fibrosis research.

The research that is being carried out by the National Institutes of Health and universities throughout the United States and Europe cover two basic areas: the causes of the disease and effective treatments. Among the more prominent institutions are: UCLA, which is using Sildenafil (Viagra) with positive initial outcomes, and Johns Hopkins University, which is working with Thalidomide. For additional information about UCLA's findings in the Sildenafil study, turn to page 5.

Other drugs that are being studied are Pirfenidone and Gc1008. Because they are in the early stages of investigation, their value is still undetermined. In Europe, the Institut National de la Sante et de la Recherche Medicale of France is studying a drug called Octretide.

The University of Pittsburgh is attempting to identify genetic and biologic markers in patients with IPF in a program called "Genomic and Proteomic Analysis of Disease Progression in IPF." Many of these studies are being replicated at other centers, which is important to verify results.

In addition to these findings, there is promising work being done at the newly formed Center of Excellence at the University of Chicago, in which the Foundation has invested \$1,000,000. Unfortunately, research of this scale and magnitude requires enormous funding. It is my hope that, with your continued support, we can look forward to finding a cure in the not too distant future.

Michael Rosenzweig, Ph.D., President and CEO

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Foundation Mourns Loss of Congressman Norwood

United States Representative Charles Norwood of Georgia passed away on February 13, 2007 after battling idiopathic pulmonary fibrosis (IPF) and non-small cell lung cancer.

Diagnosed with IPF in 1998, Rep. Norwood battled the disease for eight years before undergoing a single-lung transplant in 2004. The following year, he was treated for non-small cell lung cancer, a possible side effect of the immuno-suppressant medications, which all lung transplant patients must take. In November of 2006, shortly after being re-elected to his seventh term, he and his family learned that the cancer had metastasized to his liver.

A native of Valdosta, Ga., Rep. Norwood graduated from Georgia Southern University and received a doctorate in dental surgery from Georgetown University in 1967, serving as president of the dental student body. He served in the Army Dental Corps, with a combat tour in Vietnam, where he was one of the first participants in an Army outreach program that delivered dentists to forward firebases, his office said. Rep. Norwood also provided some of the first field-based dental treatment of military guard dogs and assisted in non-dental trauma care in Mobile Army Surgical Hospitals. He received two Bronze Star

Medals during his service.

After his discharge, he began a private dental practice in Augusta, Georgia and founded a wholesale nursery and a laboratory that manufactured dental devices.



Rep. Norwood was first elected to the House of Representatives in 1994 when the relative unknown defeated Democratic incumbent Don

Johnson. With this victory, he became the first Republican to represent his northeastern Georgia district since Reconstruction. He continued to win re-election every year since 1998 by landslide margins.

In a move that garnered national recognition, Rep. Norwood, a longtime advocate for patients' rights, introduced the first comprehensive managed health care reform legislation in 1995, which passed through the House twice, but has yet to be enacted. He was also instrumental in health care reform for military retirees and veterans as co-author of the Keep Our Promises to Military Retirees Act in 1999.

The 65-year-old Republican is survived by his wife, Gloria, sons Charles and Carlton Norwood, and four grandchildren, all of Augusta, Georgia.

Our thoughts and prayers go out to his family and friends. As with all lives cut short by this dreadful disease, the Foundation mourns his death alongside the pulmonary fibrosis community. As tragic as this event is, we are optimistic that the attention surrounding his passing will refocus on a renewed effort to find a cure for this disease ensuring that his struggle was not in vain.

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Lung Transplants Explained

For the nearly 200,000 individuals in the United States currently suffering from pulmonary fibrosis, there are few options for treatment. However, for those in otherwise good health and who meet the criteria, a lung transplant may provide hope.

While lung transplants have the potential to both lengthen life expectancy and improve the quality of life for pulmonary fibrosis patients, there are many risks and complications that can accompany such an invasive procedure. If you are considering a transplant, it is important to educate yourself as much as possible on the process, risks, and results of undergoing a transplant operation. Below are the answers to a few frequently asked questions regarding lung transplants.

What is a lung transplant and how is it performed?

There are two types of lung transplants: a single-lung or a double-lung. The transplant team will determine if one or both lungs should be transplanted.

A transplant operation is a lengthy procedure ranging anywhere from 4 to 8 hours for a single-lung transplant, and 6 to 12 hours for a double-lung transplant. During the operation, your lung is removed and the donor lung is set in its place. The surgeon connects the blood vessels to and from the lung and the main airway of the donor lung to your airway. The same procedure is followed for the other lung if you are having a double-lung transplant.

What are the risks of a lung transplant?

There are two major complications that can arise following a lung transplant: rejection and infection.

Rejection can occur because your body sees the newly-transplanted lung as “foreign,” which causes your body’s immune system to attack and try to destroy it. To prevent rejection, immunosuppressive medications are prescribed by your doctor to control the rejection process. However, rejection can still occur despite the medication. The first three months following the surgery are the most vulnerable time for rejection. Your transplant team will instruct you of any symptoms to be aware of as well as perform regular check-ups to detect rejection as early as possible.

While the medications work to prevent rejection of the new lung, they also render your body less able to fight off infection from bacteria and viruses. It is important to avoid exposure to infection and seek medical attention at the first sign of symptoms.

There are also additional risks associated with lung transplants including side effects from the medications, long-term problems with the function of the transplanted lungs, and certain types of cancer. It is important to thoroughly discuss all potential complications with your doctor prior to undergoing the operation.

What is the survival rate following a lung transplant?

According to the Organ Procurement and Transplantation Network, the survival rate for single-lung transplants is more than 82 percent at one year, nearly 60 percent at three years, and more than 43 percent at five years. The survival rate for double-lung transplants is similar — nearly 82 percent at one year, nearly 64 percent at three years, and more than 48 percent at five years.

Who is a candidate for a lung transplant?

Each transplant center has its own criteria for evaluating viable candidates for lung transplantation. Most will consider your overall health (Are you healthy enough to survive the surgery?), age, any medical conditions that could hinder the transplant’s success, and your available support network including family who can help you through this stressful time. For more exact criteria, contact the transplant center you are considering and discuss its candidacy evaluation process.

How do I get my new lung?

If the transplant team decides that you are a good candidate for a lung transplant, you will be placed a nationwide waiting list. Once a donor organ becomes available, the donor-recipient matching system administered by the United Network for Organ Sharing (UNOS) finds an appropriate match based on specific criteria, including: Blood type, Geographic distance between donor organ and transplant recipient, and Lung allocation score, which is determined by the type and severity of lung disease and the likelihood of a transplant being successful.

Where do I go for more information?

For more information regarding lung transplants, contact the United Network for Organ Sharing (UNOS) at (888) 864-6361 or visit their website at www.unos.org.

**Is there something you would like to see in our newsletter?
Let us know! To make a suggestion, please call our office at
(312) 587-9272 and speak with Leanne.**

Pulmonary Fibrosis

Surprising Results in Groundbreaking Stem Cell Trial

Researchers with Osiris Therapeutics, Inc. received surprising results in a recent groundbreaking clinical trial evaluating the use of PROVACEL, an adult stem cell therapy for the treatment of heart disease.

In the 53-patient, double-blind, placebo-controlled study evaluating the safety and preliminary efficacy of the intravenous administration of PROVACEL, heart attack patients receiving the therapy not only had significantly lower rates of adverse events, but also experienced improvements in lung condition.

“The results consistently show patient improvement with regard to heart and lung function, and indicate global improvements in well-being. These findings strongly support the ongoing development of PROVACEL for acute myocardial infarction and possibly other forms of heart and lung disease,” says Dr. Joshua Hare, the trial’s lead investigator.

As part of the study, lung function tests were performed to monitor subjects for potential adverse changes related to the treatment. Surprisingly, patients who received PROVACEL had significantly improved pulmonary function following treatment compared to placebo.

Preclinical studies performed at Osiris Therapeutics had previously demonstrated that the anti-inflammatory and anti-fibrotic effects of the stem cells may be of benefit to patients with conditions such as Chronic Obstructive Pulmonary Disease (COPD) and Idiopathic Pulmonary Fibrosis (IPF). However, this finding is the first placebo-controlled evidence in humans that indicates stem cells may play a beneficial role in the treatment of certain lung disorders.

Source: <http://investor.osiris.com/releasedetail.cfm?ReleaseID=235227>

InterMune Discontinues Actimmune Trial

InterMune, Inc. announced March 5, 2007 that it has discontinued the Phase 3 INSPIRE clinical trial evaluating Actimmune(R) in patients with idiopathic pulmonary fibrosis.

Actimmune(R) is a synthesized version of interferon gamma, a naturally occurring protein believed to stimulate the immune system.

In a planned interim analysis that included a total of 115 deaths, the overall survival result showed a lack of benefit of Actimmune(R) relative to placebo. Among the 826 randomized patients, there was not a statistically significant difference between treatment groups in overall mortality. Based on a preliminary review of the interim safety data, the adverse events associated with Actimmune(R) therapy appear generally consistent with prior clinical experience including constitutional symptoms, neutropenia, and possibly pneumonia.

Gene Mutations Linked to Hereditary PF

Scientists at Johns Hopkins may have identified the genetic culprits that trigger a hereditary form of pulmonary fibrosis. The findings, published in the March 29, 2007 issue of the *New England Journal of Medicine*, may provide new directions in diagnosis and treatment for families that inherit genes for the disease, as well as for those that develop non-inherited forms of the illness.

As many as 20 percent of IPF sufferers are thought to have inherited genetic mistakes that predispose them to the disease; and until now, these gene flaws remained unknown. To locate the genetic problem, Hopkins investigators screened DNA from blood samples of 73 people with inherited IPF and discovered that six of them (eight percent) had mutations in two genes that produce an enzyme which

helps lengthen the fragile ends of chromosomes. Chromosome ends, or telomeres, contain repetitive bits of DNA code that wear down each time a cell divides. The mutations were spotted in two genes that regulate the enzyme telomerase, which keeps telomere length extended just beyond the borders of needed genes. With mutations in telomerase, however, chromosome ends fray and wear down far more quickly, which can trigger cell death.

In the current study, mutation carriers had telomeres about one-third the length of those in family members with no gene mistakes. Short telomeres also were found in seven younger relatives who had gene mutations but not IPF. Gene tests are currently not available for IPF, but scientists are evaluating ways to assess risk of disease by

screening telomere length. “If we follow the genetic threads of families that inherit IPF, it may lead us to understand the genetic properties causing more common forms of the disease,” says Armanios. Patients with non-inherited IPF also may have short telomeres, so, says Armanios, “there may be other causes for short telomeres, such as older age and smoking, which also happen to be the main risk factors for IPF.”

To determine the link between short telomeres and non-inherited IPF, investigators will need to study a larger group of these patients. If studies reveal a solid link between the two, Armanios says that it may change the way IPF is treated.

Source: <http://www.hopkinsmedicine.org/press/2001/FEBRUARY/010222.HTM>

Research Edition

Viagra May Help Improve Exercise Capacity in PF Patients

Researchers at UCLA led by Foundation Medical Advisory Board member Dr. David Zisman have found that Viagra, also known medically as Sildenafil, may help patients with idiopathic pulmonary fibrosis. The study was published in the March 2007 issue of the journal *Chest* and shows that more than half of the patients treated with Viagra saw improved exercise capacity.

Many patients with PF also have pulmonary hypertension, which constricts arteries and lessens blood flow to the lungs, resulting in diminished lung capacity and breathing difficulties. According to Zisman, Viagra may help breathing by opening or dilating blood vessels to allow more blood flow to the lungs.

“Over five million worldwide suffer from this devastating disease, so we are hopeful that this drug may prove an effective therapy for pulmonary fibrosis,” said Zisman.

The study examined 14 IPF patients who initially took a standard six-minute walking test. All patients were then given oral Sildenafil for three months, followed by a second walking test to gauge performance changes. Researchers noted that 57 percent of the patients improved their walking distance by 20 percent or more.

“In this small pilot study, the drug was well-tolerated,” said Zisman. “The next step is to confirm this finding in a large, randomized clinical trial.”

Source: <http://www.newsroom.ucla.edu/page.asp?relnum=7741>

Frequent Asked Questions About Clinical Trials

What is a clinical trial?

A clinical trial is a research study using human volunteers to answer specific health questions. Carefully conducted clinical trials are the fastest and safest way to find treatments that work in people. Interventional trials determine whether experimental treatments or new ways of using known therapies are safe and effective under controlled environments.

Why participate in a clinical trial?

Participants in clinical trials can gain access to new research treatments before they are widely available, and help others by contributing to medical research.

Who can participate in a clinical trial?

All clinical trials have guidelines about who can participate. Using inclusion/exclusion criteria is an important principle of medical research that helps to produce reliable results. These criteria are based on such factors as age, gender, the type and stage of a disease, previous treatment history, and other medical conditions. Some research studies seek participants with illnesses or conditions to be studied in the clinical trial, while others need healthy participants.

What happens during a clinical trial?

The clinical trial process depends on the kind of trial being conducted. The clinical trial team includes doctors, nurses and other health care professionals. They check the health of the participant at the beginning of the trial, give specific instructions for participating in the trial, monitor the participant carefully during the trial, and stay in touch after the trial is completed. For all types of trials, the participant works with the research team. Clinical trial participation is most successful when the protocol is carefully followed and there is frequent contact with the research staff.

What is informed consent?

Informed consent is the process of learning the key facts about a clinical trial before deciding whether or not to participate. To help someone decide whether or not to participate, the doctors and nurses involved in the trial explain the details of the study. Then the research team provides an informed consent document that includes details about the study, such as its purpose, duration, required procedures, and key contacts. Risks and potential benefits are explained in the informed consent document. The participant then decides whether or not to sign the document. Informed consent is not a contract, and the participant may withdraw from the trial at any time.

What are the risks?

- There may be unpleasant, serious or even life-threatening side effects to experimental treatment
- The experimental treatment may not be effective for the participant
- The protocol may require more of their time and attention than would a non-protocol treatment, including trips to the study site, more treatments, hospital stays or complex dosage requirements
- There have been cases where participants have died while in the trial
- In most trials half of the participants receive a placebo which has no medical benefit
- Adverse reactions to the medication range from mild to life threatening

Clinical trials are generally conducted by Universities, research institutes and government agencies. They normally run for a period of 2 to 4 years. You should be prepared to devote the necessary time to these trials before making a commitment to them.

Clinical trials that are currently accepting participants change over time. To get a list of the latest available trials, visit www.clinicaltrials.gov and search for pulmonary fibrosis.

A Breath of Fresh Air

Pulmonary Fibrosis Foundation board member Sandra Lewis has found a way to breathe some added life into the Foundation's research endeavors. Lewis, the founding member of the Foundation's Princeton Area Committee, has organized "A Breath of Fresh Air," an elegant cocktail reception held at the American Boychoir School in Princeton, NJ on May 11, 2007, to benefit the newly-formed Pulmonary Fibrosis Foundation Center of Excellence.

Lewis's mother, Judith Bean, was diagnosed with pulmonary fibrosis almost five years ago leaving her family feeling devastated and powerless watching her suffer. "I was sitting outside Starbucks in my hometown with my sister and son when she told me. I found the Foundation's web site as soon as I returned home and was devastated and infuriated that nothing could be done to help her," says Lewis, recalling the day the family received the news.

For four years, Lewis struggled to come to terms with the diagnosis until lightning struck. "It finally dawned on

me that my sorrow was not helping her and my tears were not the cure," says Lewis. "I couldn't find the cause or cure but I could do something to help. I could raise money for research and I could raise awareness."

Armed with determination and a group of eager friends, Lewis decided on the idea of an open house cocktail party to benefit research. "It was a simple, yet festive way to bring a group together to accomplish both of my goals."

Lewis believes raising awareness and funding go hand in hand. "We will not discover the cause and cure for this disease until researchers are paid to do it, and we will only continue to raise money to continue the research if awareness is

heightened among the general public and people contribute funds."

"A Breath of Fresh Air" will certainly make a large step in the right direction to Lewis's goals. She and Princeton Area Committee have set a fund raising goal of \$20,000 after expenses through the help of event sponsors, ticket sales, and auction items.

"For the longest time, I kept my mother's illness a secret," says Lewis. Now I tell everyone about the disease and what I'm doing to help. So many people say they've never heard of it. . .now they know. I hope that some day we can make sure everyone knows what the warning signs are and join us in our fight to find help."

For more information about "A Breath of Fresh Air," visit <http://www.pulmonaryfibrosis.org/nj.htm> or call 609-730-0343.



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50/50 Calendar

The 50 Events in 50 States Fundraising Campaign aims to spread awareness of pulmonary fibrosis and raise funds for research across the nation. These fundraising events are vital to our success. The Foundation would like to thank the following individuals for offering their time to host an event. More information can be found at www.pulmonaryfibrosis.org/events.htm.

The following events scheduled for 2007:

California

- Winetasting event hosted by Kathryn Smith in memory of Wilbur Smith on September 5th

Georgia

- Bike run in memory of John Wade organized by Jim Venneau beginning on July 21st and continuing through July 23rd

Massachusetts

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

Michigan

- 3rd Annual "Paddle out Pulmonary Fibrosis" canoe marathon in memory of the Dery and Willacker families on August 4th

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, NJ
- Jessica Gilmore will be hosting a dinner/cocktail party on September 27th in memory of her grandfather, Robert Minugh

New York

- Texas Hold 'Em Tournament on June 23rd hosted by Mary Jane Borst in memory of her mother, Georgia Jean Williams Gorton
- Tennis tournament hosted by Nancy Feldman on June 23rd in memory of her mother Ruth Lang

Pennsylvania

- 2nd annual benefit bike run and picnic hosted by Rob Fiorillo and family in memory of Barbara A. Fiorillo on June 30th

Texas

- John Robertson has plans to run a triathlon in memory of William Mosley Robertson to raise funds for the Foundation in November

If interested in hosting a fundraising event of your own, please contact the Foundation's Events Coordinator, Krysten Knievel, at (312) 587-9272 to request a copy of the Event Planning Handbook.

By press time, five events have been completed:

Illinois

- Susan Heizer conducted a letter writing campaign to raise funds for the Foundation in honor of her sister Diana Larson

Massachusetts

- Doug Bernard and family hosted a fundraiser at the Quincy Knights of Columbus in memory of his mother, Judith A. Bernard on April 21st

New York

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

New Hampshire

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

Ohio

- "Comedy Rocks" Benefit held on January 19th and 20th in memory of Fred Leeds

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Exploring the Benefits of NAC

At the Foundation, we frequently receive calls from patients looking for ways to alleviate the persistent cough that often accompanies pulmonary fibrosis. While everyone deals with the cough differently, many have found relief in a natural supplement called N-Acetyl-Cysteine or NAC.

Produced by the body, NAC is a form of the amino acid cysteine. Because it enhances the production of the enzyme glutathione, one of the body's powerful antioxidants, NAC can help prevent disease and play an important role in boosting the immune system. Glutathione is made up of three amino acids and is a powerful antioxidant and the body's principal agent for safeguarding against lung damage due to oxidation.

While scientists have known for years that respiratory complications are widely due to oxidation, only recently have researchers attributed this oxidation to glutathione deficiency. This knowledge

has led to clinical trials showing that supplementing the diet with NAC boosts glutathione levels and inhibits free radical activity in the lungs.

For more than 30 years, NAC has been used as a mucolytic (mucus dissolving) agent to break up thick mucus often present in people suffering from chronic respiratory ailments.

A recent study conducted at University Hospital Gasthuisberg in Leuven, Belgium randomly assigned 182 patients to receive 600 milligrams of NAC three times a day or a placebo plus standard drug therapy.

The researchers found that, compared with placebo, NAC slowed the deterioration of vital lung capacity by nine percent and diffusing capacity by twenty percent after one year.

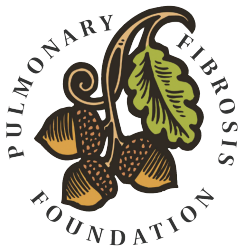
Still, some experts think that the benefits of NAC for IPF patients cannot be substantiated based on this study

alone. However, those doubting its benefits admit that taking NAC won't do any harm. "It will almost certainly do no harm, and it may be of value," says Foundation Medical Advisory Board member Dr. Gary W. Hunninghake, a professor of internal medicine and director of the Pulmonary Program in Internal Medicine at the University of Iowa.

NAC is available from most natural food stores or from online sites such as www.puritanspride.com. The recommended dosage of NAC is 1800 milligrams per day.

There are no known drug or nutrient interactions associated with NAC. However, some people complain of a sick stomach. As with any medication, natural or otherwise, you should consult your pulmonologist before taking NAC.

Source: Health Daily News & Reuters News Service



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