

# Pulmonary Fibrosis Foundation News

Spring 2002

## Lung Disease Moves to Third Place Killer

Lung Disease is now the third leading cause of death in the United States behind Heart Disease and Cancer. Approximately 361,000 people die each year from lung disease and there are 30 million people today, living, painfully, with chronic lung disease (American Lung Association).

## Pulmonary Fibrosis Still Has No Cure

There is no known cure for Pulmonary Fibrosis. More than five million people worldwide that are affected by this disease. In the U. S. alone 200,000 suffer from this malady. Of these more than 40,000 die annually. Pulmonary Fibrosis is an illness in which the air sacs or "alveoli" of the lungs are gradually replaced by scar tissue. As the disease progresses the increase in scar tissue interferes with the ability to transfer oxygen from the lungs to the bloodstream. The act of breathing becomes more difficult with the passage of time leading to a condition of general breathlessness or Dyspnea. Simple activities like walking or talking on the phone become difficult. Oxygen therapy is utilized at this point. The patient eventually succumbs to either heart or respiratory failure.

## Grant Award Program Funds Research

One of the ways the Pulmonary Fibrosis Foundation implements its mission of commitment to finding a cure for Pulmonary Fibrosis (IPF) is through its grant awards. The Young Investigator Fellowship Awards Program supports postdoctoral research for studies in the basic science or clinical investigation of Pulmonary Fibrosis (IPF). These yearly awards, intended primarily for salary support, are renewable annually for up to an additional two years based upon the availability of funding and approval of documentation of progress. To apply for a grant contact Michael Rosenzweig, Ph. D. at (720) 932-7850.

## Research Registry Helps Find a Cure

The Pulmonary Fibrosis Foundation Registry, was developed in accordance with the standards of the World Health Organization. It is a confidential database of individuals with Pulmonary Fibrosis. The largest hurdle to advances in Pulmonary Fibrosis clinical research is finding a sufficient number of volunteers to participate in clinical studies. By establishing a database of thousands of people with Pulmonary Fibrosis and making it available to the research community, the Registry will aid in the development of new therapies, and eventually a cure for Pulmonary Fibrosis. The Questionnaire, in itself, is a valuable piece of research material providing important data on Pulmonary Fibrosis.

Individuals enrolled in the Registry have the ongoing opportunity to participate directly in clinical trials of new therapeutic approaches. There is strict adherence to established confidentiality procedures that are intended to protect the identity of those who participate. The Registry is required to conceal a member's personal information from researchers and organizations unless consent is granted by the Registry participant.

The Registry represents a great research asset for Pulmonary Fibrosis patients. This is due to the concerted efforts and cooperation of physicians and the lay community. As a patient diagnosed with Pulmonary Fibrosis, your participation in a clinical research study offers all patients renewed hope for effective treatments, a greater understanding of this disorder, and the possibility that a cure will soon be discovered. Imagine, you can do all this just by participating in the registry. To participate in the Registry contact the registry committee at (720) 932-7850.

## New Research Partnerships

The Pulmonary Fibrosis Foundation and the National Jewish Medical and Research Center of Denver, Colorado began a new research program to find a cure for Idiopathic Pulmonary Fibrosis. The National Jewish Medical and Research Center has appointed Dr. Stephen K. Frankel, a noted investigator in the field of Pulmonary Fibrosis as head of the research program. The program is funded by an initial grant of \$50,000 from the Pulmonary Fibrosis Foundation. Our goal for 2002 is to raise \$500,000 to help set up additional Young Investigator Research Fellowship Programs throughout the United States. Corporations and individuals are invited to participate in this fund-raising effort. New research is the only way that we will find a cure.

## Support Groups

The Pulmonary Fibrosis Foundation was a sponsor at the first Interstitial Lung Disease (ILD) Support Group held at National Jewish Medical and Research Center on January 9, 2002. Dr. Kevin Brown and Dr. Greg Cosgrove were the speakers of the evening. They discussed the diseases listed under the ILD umbrella including Idiopathic Pulmonary Fibrosis (IPF). Dr. Brown explained the disease process and showed slides of healthy lung tissue as well as diseased tissue. Dr. Cosgrove gave an explanation of the research currently under way.

In Brooklyn, New York a group of IPF patients have started a support group called Hanging In There (HIT). If you would like to start a support group in your area write to Support Groups, Pulmonary Fibrosis Foundation, 1075 Santa Fe Drive, Denver, CO 80204 or call (720) 932-7850.

## Dr. Rosenzweig Meets With Senators

On August 20, 2001, Michael Rosenzweig, Ph.D. President of the Pulmonary Fibrosis Foundation met with Senator John D. Rockefeller, IV (D - WV), at an event in Denver, Colorado. In a private conversation with the Senator, Dr. Rosenzweig had an opportunity to point out the similarities between Pulmonary Fibrosis and Black Lung Disease, which affects a great number of the Senator's constituents. The Senator was quite interested in the formation of a coalition to work to eradicate both of these terrible diseases. A subsequent meeting was held on February 6, 2002 in Washington in which Senator Rockefeller was asked help increase the funding for Pulmonary Fibrosis Research. He was asked to insert the following language into the 2003 appropriations bill.

### Departments of Labor, Health and Human Services, Education, and Related Agencies, Appropriations Bill, 2003

#### Requested Draft Language for Consideration National Institutes of Health - Office of the Director

**Pulmonary Fibrosis** — Pulmonary Fibrosis is a devastating, lethal disease of the lungs for which there is currently no cure. There are more than 5,000,000 sufferers worldwide and 200,000 victims in the United States alone. Of these, more than 40,000 die each year. The Committee encourages the NIH to enhance its research portfolio in regard to this disease through all available mechanisms, as appropriate, including clinical trials. The NIH is also encouraged to raise public awareness and to provide information about Pulmonary Fibrosis to the public and health professionals. The Committee believes that the amount of \$10,000,000 should be appropriated for these purposes.

The Senator agreed to help. On February 18, 2002 Dr. Rosenzweig attended another event with Senator Robert Torricelli (D - NJ) and asked him to support the inclusion of the above language. He agreed and said that he would contact Senator Rockefeller so that they could work jointly on this issue. It is extremely important that everyone write their Senators and Congressmen to get their support for the inclusion of this language in the 2003 appropriations bill. The addresses of your senator can be found at [www.senate.gov](http://www.senate.gov) and your congressman can be found at [www.congress.gov](http://www.congress.gov).

## Oxygen Delivery Systems

Do you understand the types of oxygen delivery systems? Many patients are not aware that there are different methods available to them. Gas tanks are the oldest system available. Large tanks for home usage and small tanks for portable usage are quite common. The advantage to the small tanks is that they can be stored in your car and they can be laid down. Concentrators manufacture oxygen from the room air. They work well for patients who require low flow oxygen. Liquid oxygen can be delivered from large tanks located in the home. They also have two different types of portable systems. One is a canister or

stroller type system. The standard canister can deliver up to 6 liters per minute. The companion canister can deliver up to 15 liters per minute. There is a newer system called Helios. Currently the Helios system is capable of delivering only up to about 4-6 liters of oxygen flow. The manufacturer is working on a system that will deliver a higher flow. The advantage to the Helios system is that the canisters are smaller, therefore, they weigh less and can be worn on a belt around the waist.

## Traveling With Oxygen

If you have ever tried to travel with oxygen you know how frustrating and exhausting the experience can be. Trying to coordinate with different companies in different cities to provide oxygen can be a logistical nightmare. Waiting at airline gates for your oxygen supply to arrive because security won't let your supplier through can add stress to the situation. Airlines are under no obligation to provide passengers with supplemental oxygen and those that do supply it charge an exorbitant fee for doing so. This is typical of the emails we receive:

*My Fran is on O2 and AA airlines charged us 75.00 per plane. We have to make a transfer so it costs us \$150.00 each way to travel back and forth to Boston. That \$300.00 could sure be used elsewhere.... Thanks for your concern. Please keep me informed I am very interested.*  
Jean

The foundation and other interested organizations have been lobbying congress, the FAA and the airlines to find a way to alleviate this travel stress for oxygen dependent passengers. After much pressure from the foundation and others the FAA has agreed to write new regulations governing this matter.

## Testing

Pulmonary Fibrosis patients are subjected to quite a number of tests over the course of their disease. Here is a list of some of the most common tests. The tests your doctor orders will depend on how far your disease has progressed and what course of treatment you are utilizing.

- Blood Tests
- Chest X-Ray
- CT Scan
- VQ Scan
- Bone Density Scan
- Lung Biopsy
- Pulmonary Function Tests
- Spirometry

Check with your physician to determine which of these tests are best for you.

Send in any articles that you would like to see included in future editions of the News.

## New Lung Procedure Found Effective

WASHINGTON March 13, 2002 — In June 2001 Navy Capt. John Pasko was dying of Pulmonary Fibrosis. This disabling disease was robbing his lungs of their ability to breathe. He didn't know a little-known surgery and a couple of selfless naval officers were to save his life. Pasko was diagnosed in January 2001 and in desperate need of a transplant by June. Doctors couldn't explain his rapid descent. He was put on the waiting list for a donor lung in the end of May. By June 26, his condition had deteriorated to the point where his doctors at Bethesda (MD.) Naval Medical Center hospitalized him and wanted to put him on a ventilator. Pasko knew once he went on the vent, he wasn't likely to ever get off unless he got a donor lung.

Early the next morning, his wife, Sherry, called 911 because Pasko could hardly breathe, even with supplemental oxygen. He was placed on a ventilator, and his family began the painful vigil of waiting for a donor lung to be available in time. The call came in early July. A donor had been found, and Pasko was moved to the local transplant center of Inova Fairfax in Virginia. False start. The family soon learned that the lungs weren't acceptable for transplant. It was then that the Inova Fairfax lung-transplant coordinator suggested an experimental procedure available only in California.

The University of Southern California runs a hospital that is one of only three in the world that performs living-donor, double-lobe lung transplants. While Pasko was in a drug-induced coma with a machine breathing for him, his wife set a chain of events in motion that would end on the West Coast with Pasko receiving pieces of two other officers' lungs. A call went out through the entire close-knit submarine force: John Pasko was dying and needed volunteer donors. The two volunteer doctors eventually picked were Capt. Don Boland, Pasko's brother-in-law, and Lt. Cmdr. Scott Fever.

During the operation, doctors would take one lobe from one lung of each donor and transplant the two into Pasko. Lungs are elastic, so the men's remaining lobe would expand to fill the space left after the surgery. Pasko explained that doctors removed both his lungs and transplanted one lobe in each side. X-rays today show him with complete lungs, he said. The Navy had few concerns about allowing the men to be living donors, because their lung capacity was expected to return to normal with no lasting effects from the surgery.

The five-hour surgery took place at the University of Southern California Hospital July 9. The doctors said it was in the nick of time. Pasko was kept unconscious for a week after the operation to allow his body more time to recover. Recovery has been a long road for Pasko. His body began rejecting his new lungs while he was still in the hospital in California, and drugs did little to stem the effects. The answer came in the form of monthly photophoresis treatments. The experimental procedure bathes his white blood cells in ultraviolet light and helps keep the white cells from attacking and damaging his new lungs.

For a man who ran marathons before he got sick, this has been hard to take. But, Pasko said, he's coming back bit-by-bit. "As I get stronger, things get easier," he said. "Now I can take the stairs three at a time." He's still not running as much as he'd like. "I thought I'd be running by the end of December," he said. "It's been hard to accept that it's taking longer than I thought I would. But still, I see progress week to week."

Reprinted from (edited to save space) **With a Little Help From Friends — Little-Known Surgery Saves Officer's Life** By Sgt. 1st Class Kathleen T. Rhem, USA American Forces Press Service

## Albert Rose, Founder of the Pulmonary Fibrosis Foundation Died of Pulmonary Fibrosis

After a prolonged illness, Albert Rose, a Denver Businessman and founder of the Pulmonary Fibrosis Foundation succumbed to Pulmonary Fibrosis on February 6th, 2002. Mr. Rose began the Foundation when he was informed that he was suffering from the disease. He was a fighter all his life and stated "that he wasn't going to sit and wait for the undertaker". He contributed time, energy and finances to the development of the Foundation.

His enthusiasm was contagious and within a short time a Board of Directors and Medical Advisory Board were created. Despite his weakened condition, he was an active participant in Foundation work until the very end.

He was loved by all who knew him and will be sorely missed. There is now emptiness where once stood a giant. An Albert Rose Memorial Fellowship is being planned to commemorate him.

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Comments, suggestions and criticisms are always welcome. Send your letters to the address above or email: [breathe@pulmonaryfibrosis.org](mailto:breathe@pulmonaryfibrosis.org). Additional information is available at: [www.pulmonaryfibrosis.org](http://www.pulmonaryfibrosis.org)

## Sign Me Up!

I believe in the work of the Pulmonary Fibrosis Foundation and would like to contribute to it.

Here's my check for \_\_\_\$5,000 \_\_\_\$1,000\_\_\_\$500\_\_\_\$100\_\_\_\$50\_\_\_Other

Name\_\_\_\_\_

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Phone\_\_\_\_\_ Email\_\_\_\_\_

*Please send your contributions to:*

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