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Action Alert

The Quarterly Publication of the Coalition for Pulmonary Fibrosis

National IPF Awareness Week 2006: September 25 – October 1

Plans are underway to make the fourth annual National IPF Awareness Week a success, including three full days of meetings for CPF staff, volunteers, medical experts and patient advocates on Capitol Hill in Washington, D.C.

Serving as a national voice for the IPF community and its more than 10,000 members, the CPF will send 17 IPF patients, family members, non-profit partners and CPF staff to the Hill. They will meet with Members of Congress regarding important legislation and issues that affect CPF members to help raise the awareness of IPF and the needs of the IPF community on Capitol Hill. In addition, they will be meeting with a representative from the National Institutes of Health (NIH) who will speak to them about the latest federal efforts underway in IPF research and what we can expect from the NIH in the fight to find viable treatments and ultimately a cure for IPF.

The CPF delegation will meet with Members of Congress to improve awareness of IPF, and advance legislative priorities important to the patients and physicians we serve, including:

- Ending the Medicare 24-month waiting period for IPF patients (H.R. 2869 and S. 1217)
- Supporting the Home Oxygen Patient Protection Act (H.R. 5513) that would protect patients' interests with respect to oxygen and oxygen equipment
- Supporting the Access to Medicare Imaging Act (S. 3795) that would provide a two-year delay on the enactment of a provision slipped into a budget savings bill (PL 109-171)
- Supporting increased government funding of rare disease research, particularly IPF
- Supporting the Stem Cell Research Enhancement Act of 2005 (H.R. 810)
- Supporting Patients Traveling with Oxygen

GET INVOLVED WITH NATIONAL IPF AWARENESS WEEK!

You can join the CPF's National IPF Awareness Week efforts in your local area. Contact Members of Congress from your state and ask to meet with them to discuss their support of IPF awareness and research. You can also join the CPF's Campaign ACT advocacy program, an efficient online tool to improve communication between our patients and their Members of Congress.

To join, simply visit <http://www.coalitionforpf.org/campaignact>.



The CPF's B.I.G. (Breathing Is Glorious!) Ball Fundraising Gala is Saturday, October 21 in Chicago!

Can't attend, but want to support the event? You can still be involved in this important effort and support the CPF. Just send in your contribution to the CPF earmarked "BIG Ball" and the funds you give will benefit the CPF and the University of Chicago's efforts to help patients fight IPF while advancing research to find a cure. Donating is simple – just visit the CPF website at www.coalitionforpf.org to contribute online through PayPal, or call us at 888-222-8541. You may also mail your contribution to:

COALITION FOR PULMONARY FIBROSIS

Attn: BIG Ball
Suite F, #227
1659 Branham Lane
San Jose, CA 95118



The CPF is Looking For Volunteers!

We need your help in getting the word out to doctors in your area about the CPF and the work being done to help the patients and families in the IPF community. If interested, you'll have the opportunity to hand deliver important CPF education materials to medical professionals in your area. Please call Teresa Geiger to get involved today at (888) 222-8541.

Share Your Story!

The CPF newsletter regularly profiles patients, family members and healthcare professionals like you! If you would like to share your story with our readers, please contact us and tell us something interesting about you and your experience with IPF. Please email Teresa Geiger at tgeiger@coalitionforpf.org or call (888) 222-8541.

Chicago Man Works to Find Treatments for Deadly Disease That Has Claimed Seven Family Members

The motivation behind a Chicago man's fundraising efforts for a deadly lung disease is simple. The disease, idiopathic pulmonary fibrosis (IPF), that claimed his mother's life 15 years ago, now threatens his life, the lives of his siblings and possibly the lives of his three young daughters.

Roc Roney decided a few years ago to dedicate time out of his busy life as a real estate developer in Chicago to raise money for the Coalition for Pulmonary Fibrosis (CPF). Roney's goal was to find new treatments for IPF, which claimed his mother's life just after he graduated from college and while most of his siblings were still teenagers. Seven other members of the Roney family have passed away from IPF. The disease, characterized by progressive scarring in the lungs, has no known cause, no FDA-approved treatment, and no cure. The average life expectancy upon diagnosis is just three years.

"I want to live to see my children grow up," Roney, 42, said. "I also want to prevent them from experiencing what I went through with my mother, and from becoming victims of IPF, themselves."

Roney and his wife, Debbie, co-chair the CPF's annual B.I.G. (Breathing Is Glorious!) Ball. Last year's event helped raise nearly \$110,000 in the Chicago area. This year, the couple hopes to have similar success with the 2nd annual B.I.G. Ball on Oct. 21, 2006 at the Chicago Renaissance Hotel.

Though IPF is believed to have a genetic origin in less than 10 percent of the 128,000 known cases in the U.S., it has a devastating impact on the affected families. IPF has claimed the lives of multiple members of the same family, like the Roney's, making their commitment to helping others truly inspirational.

"If I get this disease,
I can say to my
kids, we've done
something about it."

– Roc Roney,
IPF Family Member

Researchers believe the familial, or genetic form of IPF may hold the key to understanding why IPF affects all patients who suffer from the disease. If they can figure out what causes the disease, with the help of affected families, they may be able to find out what can be done to stop it. Led by Duke University, a multi-year, NIH-funded

research initiative is underway to help understand the role of genetics in IPF and try to obtain these answers.

In addition to his mother's death, Roney's grandmother and four of his uncles also had IPF. The first member of his generation, a 36-year-old cousin, is the latest to be diagnosed with the disease.

Roney wants not only to find treatments for IPF, he and his wife want to set an example for their young daughters, Isabel, 6, Lola, 4, and Stella, 3. "If I get this disease, I can say to my kids, we've done something about it."

"As our girls grow, they will know why we're doing this," said Debbie Roney. "We're a little bit more in control because we're not waiting for it to come and get us. We're going at it first."



Investments in IPF Research Paying Off Hope on the Horizon

Editor's Note: *Since the CPF was founded in 2001, much has changed in the world of idiopathic pulmonary fibrosis (IPF). As clinical understanding of the disease improves, new approaches to treatment are emerging at a rapid pace. Whereas in 2001, there were just a few therapies being investigated to treat IPF, today there are approximately 10 in various stages of clinical development, including three in Phase III clinical trials.*

Researchers studying IPF have begun to change their way of thinking about the disease and the way it is treated. Patients who suffer from it are starting to see signs of hope that treatments may be on the horizon. The CPF's Vice President for Patient Outreach and Advocacy, Teresa Geiger, spoke with doctors and patients about how the recent progress in IPF research and treatment has affected them professionally and personally. Their feedback gives us a glimpse into the changes that are taking place in the IPF community.

"IPF research has really exploded," said world-renowned IPF researcher Marvin Schwarz, M.D., chairman of the CPF, the James C. Campbell Endowed Chair and professor of Pulmonary Medicine at the University of Colorado Health Sciences Center. "The interest in this disease has generated momentum that is affecting researchers and pharmaceutical companies around the world."

INCREASE IN CENTERS OF EXCELLENCE THAT TREAT AND STUDY IPF

Dr. Schwarz says an industry which had fewer than a handful of centers that specialized in this area of research just 10 years ago now has many centers across the United States that specialize in the treatment and study of interstitial lung diseases, including IPF. Dr. Schwarz would know. Over the last 30 years, he has trained a large number of physicians who now specialize in interstitial lung disease across the country. Dr. Schwarz himself has also been the recipient of numerous research grants in IPF.

Joe Lasky, M.D., chief of the section of pulmonary diseases at Tulane University, who has studied IPF and interstitial lung disease for more than 15 years, says the tides are beginning to turn in IPF research and investigators across the globe are closer than ever in uncovering viable treatments and possibly the predecessors to a future cure for the deadly lung disease. "I have a very biased opinion, but organizations such as the CPF and the National Institutes of Health (NIH), have been investing money in fibrosis research and it is beginning to pay off," said Dr. Lasky. "Some studies are suggesting some benefit [to patients and survival]."

Dr. Lasky recently received funding from the NIH to investigate the role of viruses such as Epstein Barr in the development of pulmonary fibrosis. In addition, he received funding for a pulmonary hypertension study that focuses on AIDS. He expects to learn things in the study that may apply to IPF as well.

FINDING THE ANSWERS

Much of the interest is in finding the possible causes of disease and mechanisms behind the scarring of lung tissue – believed to be the main culprit in IPF. There are various hypotheses in IPF research, and one of the prevalent theories is that this is a process of disordered healing; that is, the healing process gets 'turned on' in the body and never stops.

"There is tremendous interest in stopping the fibrotic process," said Dr. Schwarz. He says an important part of the research is to understand what happens when a patient's lungs first get injured and then understanding the process that leads to lung scarring. "When we know the pathogenesis of IPF – or how the disease develops - for sure, then we can target various therapies, like they do in cancer, for example, to treat this disease and really stop the process and possibly reverse it."

RESEARCHERS NO LONGER IN THE DARK

Gregory Cosgrove, M.D., is the assistant director of the Interstitial Lung Disease Program at National Jewish Medical and Research Center in Denver, and relatively new to IPF, having been involved in its study for just six years. But that timeframe has likely been the most important period of research in IPF history. He has now decided to focus his career on finding ways to treat and possibly cure the disease.

"We are no longer in the dark working in small groups," said Dr. Cosgrove. "We are now internationally linked so that patients as well as physicians

caring for those patients are interacting in a way so that the field is moving by leaps and bounds. It is going to lead to a fundamental breakthrough that could be just around the corner. I'm hoping at least to contribute in some small way to advance the field. I think the future for understanding as well as treating IPF is very bright."

Dr. Cosgrove also mentioned that for the first time, researchers are refocusing how they think about the disease due to the results of some sentinel studies that led to a better understanding of IPF.

PATIENTS HOPEFUL ABOUT THE FUTURE

Jan Lening is an IPF patient who lives in Littleton, Colo. She is a retired physical education teacher who has two grown children. Jan was diagnosed more than five years ago at age 50 and has already lived longer than two-thirds of all IPF patients. "I definitely think there is a reason to be hopeful," said Lening. "It may be soon enough for me...I plan to see my grandkids."

Another IPF patient, 42-year-old Beth Middlestadt of Denver, was pregnant with her son Blake, now 6, when she was diagnosed with pulmonary fibrosis in 1999. Middlestadt also has an 11-year-old daughter named Brianna. "They are the reason for fighting so hard," she said. "If it was just me, I don't think I would have put up nearly as big a fight."

Lening and Middlestadt share the same doctor at National Jewish Medical and Research Center in Denver. Kevin Brown, M.D., chief of the Interstitial Lung Disease center is a nationally recognized researcher

in IPF. "There are a million problems in the world – and you can only fix a handful," said Dr. Brown. "And, this is the one that certainly I and our group are focused on trying to fix. What we are going to see, I think probably within the next five years, are therapies that are going to significantly prolong people's lives, but more importantly provide that extension of their lives with a great quality of life."

CLINICAL TRIALS SHOW PROMISE

For patients who die on average three years after diagnosis, treatments are badly needed. And prevalence is on the rise: According to new research 128,000 Americans currently suffer from the disease and 48,000 new cases are diagnosed each year.

Though there are currently no FDA-approved treatments, no known cause and no cure for the disease, Dr. Lasky and others believe the answers are closer than ever. "Studies, so far, haven't shown a therapy to be a home run, but many studies show improvement over what we've seen in the past," said Dr. Lasky. "Maybe the treatment of IPF will require a combination treatment – maybe not a home run but a scorer run."

Imre Noth, M.D., assistant professor of medicine at the University of Chicago, says success in IPF is only a matter of time. "I think efforts are growing at an exponential rate and that is certainly reason for optimism," he said. "The number of scientific efforts and clinical trials are five to ten times greater now than they were just five years ago. Between increased bench research work and a ten-fold increase in the number of clinical trials now being conducted, it is no longer a question of 'if' we will make

a meaningful impact in this disease but 'when'. This should drive us all to do more." For the first time, in preliminary studies some drugs in clinical trials in pulmonary fibrosis appear to be having an impact on length of survival and mortality in some patients. Though more research is needed, the trend is incredibly promising.

"Until now, there haven't been any treatments that have provided any measurable benefits," said Dr. Lasky. "Though not perfect, drugs being investigated today hold much more promise than past treatments. And, the more we learn through continued research, the better our opportunities are for finding life-saving treatments sooner rather than later." Dr. Lasky and researchers also say viable treatments are likely not far away. "There are a number of ongoing studies that we should see information from in the next two to three years – promising studies," he said.

Several studies are expected to be completed in the coming 24 months: A recent clinical trial of Gleevec (imatinib mesylate) is expected to be completed in the fall of 2007, a Phase III study of Actimmune (interferon gamma 1-b) expected to be completed in early 2008, a Phase II study of Ventavis (inhaled iloprost) is expected to be completed in 2007, and a Phase III trial of Pirfenidone is expected to be completed in late 2008. There is also a large, multi-center research consortium led by the National Institutes of Health called the IPF Network which expects to begin human trials of experimental therapies within twelve months.

New Research Shows IPF is More Common Than Previously Thought: Prevalence Now Estimated at 128,000

Understanding that idiopathic pulmonary fibrosis (IPF) is a chronic interstitial lung disease whose epidemiology in the United States has not been well characterized, Ganesh Raghu, M.D. - from the University of Washington Chest Clinic - and colleagues recently initiated a retrospective study to better understand the incidence and prevalence of the disorder. The study concluded that IPF is probably more common in the United States than previously reported. The results were scheduled for publication in the American Journal of Respiratory & Critical Care Medicine as this issue of the CPF's Action Alert went to print.

The objective of the study was to estimate the annual incidence and prevalence of IPF in the United States. The researchers employed a retrospective cohort design utilizing a large healthcare claims database spanning the period of January 1996

through December 2000. Persons with IPF were identified based on diagnosis and procedure codes.

Using broad case-finding criteria, prevalence was estimated to range from 4.0 per 100,000 persons aged 18-34 years to 227.2 per 100,000 among those 75 years of age and older; annual incidence was estimated to range from 1.2 to 76.4 per 100,000.

Using narrow case-finding criteria, prevalence ranged from 0.8 to 64.7 per 100,000 persons; comparable figures for incidence were 0.4 to 27.1 per 100,000 persons. Extrapolating these rates to the overall U.S. population, prevalence was estimated to be 42.7 per 100,000 (incidence, 16.3 per 100,000) using broad criteria; with narrow criteria, prevalence was estimated to be 14.0 per 100,000 (incidence, 6.8 per 100,000).

Source: Abstract from the American Journal of Respiratory & Critical Care Medicine (Am J Respir Crit Care Med. 2006 Jun. 29 published online ahead of print): Investigators: Ganesh Raghu, Derek Weycker, John Edelsberg, Williamson Z Bradford, and Gerry Oster.

Editor's Note: Using the broad criteria noted above, and combining the data with the most recent U.S. Census report showing the population of the United States as 300 million, the new estimated number of cases in the United States is 128,100, and the new estimate of incidence is 48,900 per year. CPF has confirmed the accuracy of the new estimates with the study authors. The abstract and complete manuscript can be viewed online at <http://ajrccm.atsjournals.org/>: the abstract is free of charge; the full manuscript requires a subscription to the journal.

Candidate Gene for Familial Idiopathic Pulmonary Fibrosis Possibly Identified

Researchers at the University of Helsinki and the University Hospital of Helsinki, Finland have identified a candidate gene for Familial Idiopathic Pulmonary Fibrosis (FIPF), according to a recent study published in the American Journal of Human Genetics.

The pathogenesis and etiology of IPF remain unknown, but the reports of

multiple affected family members in the same family support the influence of genetic factors. This study, published in the July 2006 edition of the AJHG, suggests the ELMOD2-gene is a prime candidate gene for FIPF.

In their previous studies researchers at the University of Helsinki and the University Hospital of Helsinki

observed that the prevalence of idiopathic pulmonary fibrosis was distributed unevenly in Finland. The prevalence was two times higher in eastern and southern Savo (45 cases per 100,000 inhabitants) compared to the prevalence in Finland (16-18 cases per 100,000).

The researchers identified families with IPF and noticed that familial

Viruses Play Role in Lung Disease

By Fran Simon
Tulane University Health Sciences Center

The Tulane University Section of Pulmonary Diseases and Critical Care Medicine recently received a major grant from the National Institutes of Health to study the role of viruses in chronic lung diseases. Joseph Lasky, M.D., chief of the section of pulmonary diseases, received a four-year grant of nearly \$1.5 million to study the role of Epstein-Barr virus in idiopathic pulmonary fibrosis.

Dr. Lasky's co-investigators on the EBV/IPF study are Eric Flemington, Gilbert Morris and Debbie Sullivan in the pathology department, and Gary Hoyle and Bin Shan in the pulmonary diseases section.

"Latent viruses may alter cell behavior and cause the lungs to undergo abnormal repair, such as exuberant scarring, in response to insults or injuries," Lasky says.

Lasky is now the principal



Joseph Lasky, M.D., chief of the section of pulmonary diseases at Tulane, is studying lung diseases along with one of his co-investigators, Bin Shan.

investigator on three NIH grants; in 2005 he received a five-year grant to lead a Gulf South regional network for clinical research with patients who have idiopathic pulmonary fibrosis.

IPF causes shortness of breath with exertion and usually leads to death, predominantly in people over the age of 50. There are as many as 48,000 newly diagnosed cases of IPF in the United States annually.

The Tulane Gulf South Clinical Research Network is reaching out to IPF patients in Louisiana, Mississippi, Alabama, Texas and Florida, and is combining forces with 10 other national centers to

determine which pharmacological agents show the most promise to treat lung scarring and prolong life for IPF patients.

Prior NIH support of Lasky has led to an ongoing investigator-initiated national trial using imatinib mesylate (Gleevec) for the treatment of IPF, in collaboration with investigators at the Mayo Clinic.

"About half the patients with IPF will die within three years of their diagnosis, but a minority of patients may survive for 10 years," Dr. Lasky says. "Why do some patients rapidly lose lung function and die, whereas others get worse only slowly? Patients with IPF who carry Epstein-Barr, or other herpes virus, have a latent viral infection or may have undergone a viral genetic rearrangement that causes a more precipitous decline in lung function. One could look at the underlying disease of IPF as a smoldering fire, and the virus is a hot, dry wind that fans the flames."

Continued from previous page

IPF patients were clustered within Savo and Carelia, the same areas with the high prevalence, suggesting that they most likely share a common disease-causing gene or genes introduced by a common ancestor. The researchers performed a genome-wide scan with six multiplex families.

Since ELMOD2 is potentially involved

in apoptosis, phagocytosis, cell engulfment, and cell migration, and its expression is significantly decreased in IPF lungs when compared healthy lungs, researchers determined ELMOD2 to be a prime candidate gene for familial IPF. The complete manuscript can be viewed at www.journals.uchicago.edu/AJHG/ home by subscription only.

INVESTIGATORS: Hodgson U, Pulkkinen V, Dixon M, Peyrard-Janvid M, Rehn M, Lahermo P, Ollikainen V, Salmenkivi K, Kinnula V, Kere J, Tukiainen P, Laitinen T:

REFERENCE: ELMOD2 Is a Candidate Gene for Familial Idiopathic Pulmonary Fibrosis. Am J Hum Genet. 2006 Jul;79(1):149-54. Epub 2006 May 9

Source: Medical News Today and Wire Services

Amarillo Biosciences Gets FDA Approval for Phase II IPF Clinical Trial

Amarillo Biosciences' investigational new drug (IND) application to evaluate interferon in idiopathic pulmonary fibrosis has successfully passed the FDA's 30-day review process, clearing the way for a phase II trial to begin. With the FDA's backing, Amarillo now aims to begin a 60-patient phase II study to test low-dose oral interferon as a treatment of chronic cough in idiopathic pulmonary fibrosis (IPF) patients in the fourth quarter of 2006. According to the company, approximately 85-90% of idiopathic pulmonary fibrosis (IPF) patients complain of a bothersome persistent cough that negatively impacts their quality of life.

The trial is a follow-up to a positive pilot study conducted by investigators at the Texas Tech University Health Sciences Center, in which five out of six IPF patients given orally administered IFN-alpha reported a significant reduction in chronic cough with corresponding improvement in their quality of life.

Source: Amarillo Biosciences



FDA Announces New Initiative to Protect Clinical Trial Participants, Resulting Data

The Food and Drug Administration (FDA) recently unveiled a new initiative aiming to strengthen oversight of clinical trials and to protect participants through policy making and regulatory changes. The program, called the Human Subject Protection and Bioresearch Monitoring (HSP/BIMO) initiative, is part of the Department of Health and Human Services' effort to improve development and review of therapies.

"As clinical trials continue to evolve, in particular becoming increasingly large, decentralized and global, the FDA's approach to bioresearch monitoring and human subject protection must also evolve and modernize," said Janet

Woodcock, FDA deputy commissioner for operations at a recent Drug Information Association conference where the initiative was originally announced.

FDA regulations concerning clinical trials date back to the early 1980s. Since then, clinical trials have changed "dramatically," Woodcock said. Some changes already made under HSP/BIMO include draft guidances for handling referrals to the agency, safeguards for children in trials, and establishing a centralized institutional review board — a group of professionals who review protocols and trial material to establish safety for participants for multicenter trials.

According to an FDA release, the agency spent the past year and a half going over its programs and identifying issues for HSP/BIMO. The FDA said it will continue to examine potential issues as well as seek related information from internal and external stakeholders and the public.

Source: Congressional Quarterly HealthBeat News

University of Pittsburgh Warm Autopsy Program Could Lead to Better Understanding of IPF

Researchers at the University of Pittsburgh have begun to analyze the progression of idiopathic pulmonary fibrosis (IPF) through the examination of lung tissue recovered shortly after a patient passes away. The process, known as 'warm autopsy', is described by University of Pittsburgh School of Medicine (UPSM) researchers in the June 2006 issue of PLoS Medicine, an online journal of the Public Library of Science.

"Up until now, what we have been able to do has been limited by the lack of availability of lung tissue for study," said Naftali Kaminski, M.D., director of the Dorothy P. & Richard P. Simmons Center for Interstitial Lung Disease in the division of pulmonary, allergy and critical care medicine at UPSM and senior author of the PLoS essay, "Lessons from Our Patients: Development of a Warm Autopsy Program."

Warm autopsy - a practice involving retrieval of organs within six hours of death - has been in use for more than 25 years, chiefly in the study of Alzheimer's disease and multiple sclerosis. However, the University of Pittsburgh's program, which began at the suggestion of an IPF patient, is among the first to use the technique for lung disease research. "Obtaining such prompt access to the organs will, we hope, allow us to approximate disease conditions in a living patient," said Kathleen Oare Lindell, R.N., clinical nurse specialist at the Simmons Center and the essay's first author. "The goal is to learn as much as we can about how IPF works."

Since 2003, Dr. Kaminski and his colleagues have examined donated lungs from 12 patients, beginning with tissue from a retired firefighter who suggested the idea after participating in support group sessions. "We had never had patients wanting to donate their lungs before and had not even discussed the possibility," said Ms. Lindell, adding that research into the proposal revealed there were no such programs in the country for lung disease.

"Obtaining such prompt access to the organs will, we hope, allow us to approximate disease conditions in a living patient."
— Kathleen Oare Lindell, R.N

"The tissue that is available from lungs is rare and primarily from cancer patients," explained Dr. Kaminski. "Most IPF patients will have one biopsy for diagnostic purposes, and because of surgical risks and their fragile health, some may not get a biopsy at all."

But thanks to warm autopsy donors, tissue studied soon after death can be compared to tissue collected at biopsy - usually years earlier. Preliminary analysis has revealed some promising paths for further research.

"Using these donated lungs, we can retrieve tissue from multiple locations within the organ and see how they compare," said Dr. Kaminski.

The warm autopsy program also has a bonus of strengthening connections between patients, their family members and the health professionals who care for them, said Ms. Lindell. "This program is one example of how a patient can make a difference and initiate something that will help others. It conveys a message that the team respects patients' wishes and allows them to contribute, even in their last days."

Source: University of Pittsburgh School of Medicine

CoTherix Completes Enrollment of Phase II Study of Potential Treatment for Pulmonary Hypertension with Associated IPF

CoTherix, Inc. of South San Francisco, Calif. announced on Aug. 2, 2006 that it has completed enrollment of a Phase II clinical trial investigating the safety and potential effectiveness of inhaled Iloprost (Ventavis) in the treatment of patients with pulmonary hypertension associated with idiopathic pulmonary fibrosis (IPF). Results from this study will be announced in 2007.

According to CoTherix, approximately 20 percent of IPF patients also have pulmonary hypertension (PH) associated with their IPF. This association is due to progressive scarring of the lung tissue that causes increased pulmonary artery pressure. Inhaled Iloprost allows for direct alveolar deposition so the drug can act locally and produce potent pulmonary vasodilation when compared to systemic vasodilators.

For more information about this clinical trial, please contact CoTherix at 877-483-6828, or visit www.cotherix.com.

Source: CoTherix, Inc.

Mycophenolate Mofetil a Promising Treatment for Connective Tissue-Related Interstitial Lung Disease

By David Douglas, Reuters Health

Mycophenolate mofetil, an inhibitor of proliferating lymphocytes, appears to improve lung function in patients with interstitial lung disease as a result of connective tissue disease, Colorado-based researchers report in the July 2006 issue of *Chest*.

Lead investigator Dr. Jeffrey L. Swigris told Reuters Health that the study findings provide "the impetus for further study of mycophenolate mofetil in patients with connective tissue disease-related interstitial lung disease." It appears "that mycophenolate mofetil is safe and well-tolerated and may be an effective alternative to other immunosuppressant drugs in patients with this spectrum of diseases," he said.

Dr. Swigris of National Jewish Medical and Research Center in Denver and colleagues conducted a retrospective study of 28 patients who were treated with mycophenolate mofetil over a total of 35.9 patient-years.

Scleroderma, in nine patients, was the most common diagnosis. Other condition included undifferentiated connective tissue disease and dermatomyositis.

Most patients started taking mycophenolate mofetil because of adverse effects of another immunomodulatory agent. Six patients had significant mycophenolate mofetil-related side effects, but these resolved after dose reduction. After starting

mycophenolate mofetil, patients on prednisone were able to significantly decrease their mean daily dosage from 15 mg/d to 10 mg/d and five patients were able to discontinue prednisone use altogether.

Patients also had a 2.3 percent increase in forced vital capacity and an increase in average percentage of total lung capacity of 4.0 percent. The patients' lung diffusing capacity for carbon monoxide also increased by 2.6 percent.

Despite these encouraging results, Dr. Swigris said "prospective studies will be needed to confirm the inferences suggested by our study."

Reference: *Chest* 2006;130:30-36.
Source: Reuters Health Information 2006; Medscape.com





Education. Support. Hope.



CPF Announces 2006 Fall/Winter IPF Seminars

University of California, San Francisco & Stanford University

The Coalition for Pulmonary Fibrosis (CPF) in partnership with the University of California San Francisco and Stanford University in San Francisco, Calif. is hosting a free seminar on Saturday, Nov. 4, 2006 for patients and families living with idiopathic pulmonary fibrosis (IPF). This is a unique opportunity for patients, family members, caregivers — anyone affected by IPF to learn about the latest in IPF diagnosis, research and treatment from leading IPF researchers and pulmonary experts.

What: “Living with IPF” Free Educational Seminar

When: **Saturday, Nov. 4, 2006**

8:45 a.m. – 2:00 p.m.

Complimentary continental breakfast and lunch

Where: Mission Bay Conference Center at UCSF

1675 Owens Street

San Francisco, CA 94143-3000

University of California, Los Angeles

The Coalition for Pulmonary Fibrosis (CPF) in partnership with the University of California, Los Angeles (UCLA) is hosting a free seminar on Friday, Dec. 1, 2006 for patients and families living with idiopathic pulmonary fibrosis (IPF).

What: “Living with IPF” Free Educational Seminar

When: **Friday, Dec. 1, 2006**

8:30 a.m. – 3:15 p.m.

Complimentary lunch included

Where: California Room at Faculty Center on the campus of UCLA

For more information, or to register for either of these events, please contact the CPF at (888) 222-8541 or visit www.coalitionforpf.org.

Georgia Congressman Norwood, Other House Panel Members, Vocal About Medicare Imaging Cuts; New Bills Introduced to Delay Medicare Cuts

Editor's Note: According to the Consensus Statement of the ATS, medical imaging is a critical procedure to accurately diagnose and treat IPF. HRCT scans are considered one of the most effective tools for making an IPF diagnosis. The CPF supports legislation that would continue to allow physicians to appropriately use medical imaging in diagnosing lung disorders like IPF.



Rep. Charlie Norwood (R-GA) was loud and clear with his questions and concerns at a recent House hearing on cuts in Medicare payments for medical imaging that are scheduled to begin January 1, 2007.

The cuts were mandated in a provision slipped into a fiscal 2006 budget savings law (PL 109-171) that will trim \$2.8 billion from those payments over five years. But a number of lawmakers and witnesses at the House Energy and Commerce Health Subcommittee hearing said the cuts should be delayed until 2009, pending a Government Accountability Office study of their impact.

Rep. Norwood, who appeared to be grimacing through testimony by an administration official explaining the cuts' rationale, led the critical questioning.

Noting his own recent brush with death from lung disease (IPF) and opining that medical

imaging probably helped save his life, Norwood fired off a series of questions aimed at showing that Medicare can't be confident the cuts won't slice into medically necessary treatment because the agency doesn't know enough about the reasons for rapid increases in imaging costs.

Rep. Anna G. Eshoo (D-CA) warned against a meat-axe approach to imaging cuts, noting Norwood's suggestion that imaging may have helped save his life and reiterating the doubt that CMS [Centers for Medicare & Medicaid Services] thoroughly understands the impact that imaging cuts would have on savings generated by imaging technology on hospital care.

Imaging use should not be discouraged without a thorough understanding of its impact, said Rep. Edolphus Towns (D-NY). "I know we're trying to cut costs . . . but we really have to be careful," he said.

Kuhn noted that imaging spending has been rising at a fast clip. "Between 2000 and 2005, spending for imaging services paid under the physician fee schedule more than doubled from \$6.6 billion to \$13.7 billion, an average annual growth rate of 15.7 percent. This compares to an annual growth rate of 9.6 percent for all physician fee schedule services."

Spending for "advanced" imaging, a category largely consisting of CAT scans and MRI procedures, grew by 25 percent during 2005 and 82 percent from 2003-05, he said.

Those rapid increases raise questions about over-utilization of imaging services, Hackbarth testified. While individual patients' imaging clearly improves medical outcomes, research shows that more imaging spending in the aggregate is not necessarily tied to better outcomes, he said.

At least three bills have been introduced in Congress - including bills from Rep. Joe Pitts (R-PA), and Sens. Gordon H. Smith (R-OR), and John D. Rockefeller IV, (D-WV) - to remedy the concerns of Congressional leaders and the House Energy and Commerce Health Subcommittee.

Source: CQ HealthBeat News, content edited and summarized for space

Federal Funding of Stem Cell Research Still on Hold

Less than a year after Senate Majority Leader Bill Frist rocked his party by throwing his support behind a bill to expand embryonic stem cell research, the Tennessee Republican was able to generate more votes than expected but not quite enough to achieve a two-thirds majority to override a presidential veto of new legislation.

The close vote, just four votes shy of the necessary two-thirds majority required by Congress to override a presidential veto, demonstrates the growing support on and off Capitol Hill of stem cell research.

During a House floor debate in May, several GOP lawmakers noted their anti-abortion voting record but said the illnesses of friends and relatives motivated them to support the bill.

Scientists say embryonic stem cells have the potential to help understand and offer cures for life threatening illnesses such as diabetes, Parkinson's disease, some cancers and possibly Alzheimer's. Support for the research has also been bolstered by former first lady Nancy Reagan, whose backing has raised the profile of the issue.

The CPF supports Congress' efforts to end the ban on federal funding of research on new stem cell lines, as stem cell research may have potential to improve the lives of millions of Americans living with life-threatening illnesses such as idiopathic pulmonary fibrosis. A 2005 public opinion survey found that 74 percent of Americans agree with this position.

Source: Congressional Quarterly and Wire Services; KR Research

Lung Ailments No Longer Standing in the Way of Flight

Travelers with pulmonary disorders are thrilled that they have better access to the skies, making it easier to visit clients or family, even to take far away vacations. But some medical experts say in-flight respiratory emergencies are common, and some fliers with serious pulmonary problems are not healthy enough to fly.

There's also concern that the breathing devices used by passengers with lung ailments could pose some risk to other fliers. The government warns that devices should not be near an open flame and says airlines must ensure that they don't interfere

with navigational and other aircraft equipment.

Until recently, most airlines didn't provide or allow supplemental oxygen aboard planes. That made it difficult or impossible for most people with lung disease to fly commercial jets. But prodded by the government to give equal access to the medically impaired, airlines began changing their policies in the past year.

The Federal Aviation Administration issued a rule in July 2005, that said airlines can allow passengers to bring approved oxygen devices aboard. Two months later, the Department of Transportation issued a proposed rule requiring airlines to allow the devices and to supply oxygen for anyone with a medical problem.

Today, nine of 19 U.S. airlines surveyed by USA TODAY now provide bottled oxygen for a fee, and 11 airlines allow passengers to carry on their own portable oxygen concentrators — a relatively new product that converts cabin air into oxygen. Continental Airlines this month will become the 12th carrier to allow concentrators aboard, says spokesman Dave Messing.

BREATHING UNDER PRESSURE

Joan Garrett, CEO of MedAire, which provides in-flight medical assistance to about 90 airlines, supports the move to make it easier for pulmonary patients to fly. But she's worried that people with only one lung or serious breathing problems might think that an oxygen device guarantees their well-being in-flight. It doesn't, she says.

Continued on page 14

Continued from page 13

Many people with pulmonary disorders have multiple problems, such as heart disease, kidney problems and diabetes, and probably should not be on planes, Garrett says. The FAA requires that passengers have a doctor's permission before flying with an oxygen device, but Garrett says some doctors give that clearance without understanding the detrimental effects that altitude can have on an impaired passenger. Air inside an aircraft cabin is pressurized to an altitude of 8,000 feet.

Claude Thibeault, a doctor and aviation medicine expert, agrees that most doctors aren't well-versed with cabin altitude issues. It's a "legitimate concern" whether a person with a pulmonary disorder should fly, he says.

Frederick Tilton, the FAA's federal air surgeon, declined a request for an interview, but provided written responses to questions. He says individuals with medical conditions that require oxygen "should consult with their personal physician to determine fitness to fly before contemplating air travel." Individuals shouldn't fly "if they are medically unstable, and physicians should advise them against doing so," he says.

But Paul Billings, vice president at the American Lung Association, says he's not aware of any data that shows a traveler with lung disease is at any greater risk in-flight than someone with another disease. New technology "holds promise to open the skies" to those with respiratory problems and should be celebrated, he says.

For the latest airline specific guidelines on traveling with oxygen, as compiled by the CPF, visit our website at coalitionforpf.org.

Airlines aren't equipped to handle many types of in-flight medical emergencies. Flight crews can administer first aid and assist choking victims, Garrett says, but they aren't trained and don't have the equipment to deal with more complex illnesses.

AIR OF FREEDOM, CONCERN

Though not required to do so, some airlines began allowing oxygen concentrators on planes after the FAA approved two manufacturers' units last year. Among other conditions, the FAA requires those airlines to ensure that the concentrators do not interfere with a plane's electrical, navigation and communications equipment, and that no smoking or open flame is permitted near a passenger with a concentrator.

In comments submitted in January regarding the Department of Transportation's proposed medical oxygen rules, the Air Transport Association said safety experts have expressed concern that oxygen concentrators' batteries could

short-circuit and cause a fire. The group called on the agency to adopt the FAA's stowage and packaging rules for the batteries. New technology has incorporated safeguards that prevent a fire, says Daryl Risinger, vice president of Inogen, one of the approved manufacturers. Concentrators "use the same battery technology as laptop computers," he says.

Airlines oppose a Department of Transportation proposal that would require them to provide free bottles of oxygen for passengers with lung problems. In comments submitted to the DOT this year, the Air Transport Association said it would cost \$103 million annually to provide oxygen, and more to train the flight crew on its use. Providing free bottles of oxygen might also discourage passengers from bringing their own oxygen concentrators aboard, the airlines say. The concentrators store only enough oxygen for a person's next breath, says Risinger, and are safer than bottles of oxygen, which can accelerate a fire and is considered, according to DOT regulations, a hazardous material. Billings says he understands safety and cost concerns, but air travel is "very important" to people with pulmonary disorders. It gives them "freedom of mobility," he says.

Source: Usa Today, August 11, 2006 – content edited for space

Having Trouble with Insurance, Medication, or Disability? The Caring Voice Coalition Can Help!

Since its founding in 2003, the Caring Voice Coalition (CVC) has been instrumental in providing desperately needed support for patients suffering from serious and chronic disorders like IPF. The CPF has referred hundreds of IPF patients to the CVC since their inception. The CVC gives a helping hand to IPF patients who are having difficulties with affording the healthcare expenses they face when dealing with serious illness. They even help patients who don't have insurance to qualify for coverage.

"We get in the trenches to figure out where we need to land the punches in order to get the work done," said Pam Stockwell, the CVC's reimbursement team leader who has 25 years experience in insurance reimbursement.

The CVC assists more than 3,000 patients a year in three different pulmonary diseases: IPF, Alpha 1 Antitrypsin Deficiency (Alpha-1) and Pulmonary Hypertension. They have helped more than 100 IPF patients so far this year.

Amy Atkisson, 63, was diagnosed with IPF two years ago. The Norcross, Georgia woman contacted the CVC for help with coverage for her medications and found much more. "They helped me get some medicine," she said. "They have helped me a lot. I had back surgery and [Caring Voice] called a company who donated a mattress. The Caring Voice will do everything in their power to help you. I love them with all of my heart."

The organization provides assistance to IPF patients in three basic departments within the Caring Voice: The Personal Support Program, which provides counseling and support and connects patients with resources to help them deal with the mental and emotional side of lung disease; The Reimbursement Team, which helps patients overcome reimbursement difficulties and insurance denials; and the Financial Assistance Program, which in certain circumstances can provide patients some relief from high drug costs.

Pam Harris, president of the Caring Voice Coalition, says it's the staff of the organization that makes things happen for their patients. "The thing that is making us so successful is all of the staff is passionate about the work they do," she said. Harris says their success also comes from their employees' vast experience in their respective fields, with most of their 26 employees having more than 10 years experience in their areas.

"Through the three programs, there is a great chance of getting some assistance from us (Caring Voice)," said Doyle Hull, director of patient services for Caring Voice.



Caring Voice Coalition

To learn more about the Caring Voice Coalition, please visit their Web page at www.caringvoice.org, or call (888) 267-1440.

Patricia Barnett, 63 from Ringgold, Georgia was diagnosed with IPF five years ago. Barnett says Caring Voice came through for her when she needed help paying for a \$5,000 a month experimental drug. "At the time, Medicare had stopped paying for my shots and I couldn't afford them." Though she didn't expect the Caring Voice to be able to help her, at the time, they were able to get the medication reinstated by Medicare. "I don't know what I would have done without them," she said. "It is incredible to know that someone out there cares for you...someone who I don't know who would take the time to call someone they don't know. They say 'hey, we love you. If you need anything, let us know.' I think it is what has kept me going. They are wonderful people."



The Caring Voice Coalition staff

Oxygen and Exercise Make a Difference in Managing IPF

Individualized rehab helps patients manage debilitating disease

When respiratory care professionals assure adequate support and instruction, patients with exertional dyspnea [shortness of breath upon exertion] requiring supplemental oxygen can stick with a tailored, independent exercise schedule. Individualized oxygen supplementation during exertional activities can make the difference for patients who might otherwise avoid these activities due to increased dyspnea [shortness of breath] and fatigue.

Patients with idiopathic pulmonary fibrosis (IPF) often desaturate [lose oxygen saturation] with exertion and thus provide a good example of the appropriate use of supplemental oxygen during exertional activities. IPF patients typically require higher levels of supplemental oxygen as their disease progresses than patients with other lung diseases such as chronic obstructive pulmonary disease.

The causes of IPF haven't been discovered, and an effective treatment regimen has yet to be determined. This incurable disease is fatal, and the median survival is three years with the currently available "treatment regimen" used by physicians without convincing evidence.

IPF is diagnosed by a thorough clinical assessment that includes a high resolution CT scan of the chest, histopathology and pulmonary function test.¹ Because lung volume

is decreased due to deposition of excess fibrotic tissue and decreased elasticity in the lungs, it's difficult for the IPF patient to take deep breaths, especially during exertional activities. As a result, the patient's anxiety and discomfort level increase with decreased exercise tolerance.

The object of oxygen therapy and rehabilitation for patients with IPF is to improve the patient's ability to be more physically active and cope with the frustration associated with management of their debilitating disease.

DETERMINING OXYGEN NEEDS

It's important that the patient maintain oxygen saturation of greater than 88 percent during exercise. Many patients with advanced/severe lung disease require supplemental oxygen to do this.

Before beginning any type of exercise or increased physical activity, a patient with lung disease should be evaluated by a pulmonologist and respiratory care professional. The respiratory care professional monitors the patient in a clinical situation to determine the oxygen requirements for the patient during exercise.

IPF is a progressive lung disease, therefore oxygen desaturation during exertional activities worsens over time, and the levels of oxygen supplementation should be reassessed during follow-up visits every three to four months.

Pulse oximetry during walking and/or climbing stairs helps determine appropriate levels of oxygen supplementation and provides guidance to the patient based on continuous SpO₂ [oxygen saturation] measurements. The patient is encouraged to walk at his own set pace on a pre-measured flat level surface for six minutes. Distance, heart rate, SpO₂ as measured indirectly by a pulse oximetry, and shortness of breath score (Borg scale) are recorded.

As oxygen saturation decreases, the patient's dyspnea usually will increase. If the patient's saturation is below 89 percent, supplemental oxygen is indicated. The rate of oxygen flow needed is determined by placing the patient on a nasal cannula [plastic tubing inserted in the nose] with supplemental oxygen, initially at 2 Lpm [liters per minute], and assessing the SpO₂ during the six-minute walk test.

Oxygen should be titrated to keep oxygen saturation above 89 percent. SpO₂ below 80 percent may precipitate cardiac events, especially in patients with occult cardiac disease, so it's appropriate to stop the six-minute walk test before the time is up if a patient demonstrates desaturation to 80 percent.

Based on the stage and clinical course of disease, determinants of the six-minute walk test may vary from visit to visit, and such longitudinal measurements may reflect progression of the disease. Determining significant

Most lung disease patients, including those with pulmonary fibrosis, should work to complete the rehab program and continue with exercising, either in a supervised program or on their own, to maintain their conditioning.

SpO₂ desaturation and other variables in a six-minute walk test and a timed walk test —a modified version of the six-minute walk test — has been associated with survival in IPF.^{2, 3}

OXYGEN DELIVERY

Supplemental oxygen can improve oxygenation, decrease dyspnea, and allow patients to increase their activity levels. It also will make the patient feel comfortable with exercising and during exercise. It's important to limit the stress on the heart due to increased hypoxia, so adequate oxygenation with appropriate levels of supplemental oxygen, when needed, is imperative.

Oxygen is a prescribed medication and must be ordered by a physician in accordance with the amount necessary to maintain saturation above 89 percent. Home care companies deliver supplemental oxygen delivery devices to the patient's home and instruct the patient on appropriate use and care of the devices.

Three oxygen delivery systems are currently available: a liquid oxygen system, compressed gas oxygen system and a concentrator system.

- Liquid oxygen - cools the oxygen gas, which changes it to a liquid

form. Larger amounts of oxygen can be stored in smaller, more convenient containers than compressed oxygen, making it more convenient during exercise and activity. However, it can't be kept for a long time due to evaporation.

- Concentrator - concentrates the fraction of oxygen in room air, runs it through filters to remove nitrogen from the air, and provides approximately 94 percent to 96 percent oxygen. The rate can be adjusted anywhere from one to 10 Lpm [liters per minute]. The concentrator units are big, so they're often equipped with 50-foot hoses that deliver the oxygen to the patient. This system depends on electricity and requires a backup power source. However, a new portable concentrator has been approved for air travel.
- Compressed gas - is 100 percent oxygen compressed to fit into portable tanks of varying sizes. These tanks are flow regulated up to 15 Lpm. They can be fitted with a pulse dose system regulator that extends the oxygen tank's life by giving a puff of oxygen when the patient initiates the flow; it doesn't provide a continuous flow like most tanks.

STICKING WITH IT

The patient needs to work out at an increased effort to reap any benefits of training. A good rehabilitation facility will have adequate support so that a professional is available to the patients throughout the session. Most lung disease patients, including those with pulmonary fibrosis, should work to complete the rehab program and continue with exercising, either in a supervised program or on their own, to maintain their conditioning.

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Source: By Ganesh Raghu, M.D., FCCP, FACP, and Jeffrey Moniz, CRTT, RRT as published in Advance for Managers of Respiratory Care. Content edited for space.

The CPF Celebrates Five Years!

It has been five years since the CPF was founded to become the definitive resource for accurate, reliable and helpful patient and physician information. Since our inception, the CPF has expanded its capabilities to fund promising research into treating and curing IPF!

Today, there are studies underway which may uncover the cause(s) of IPF and drugs under investigation which may lead to IPF treatments. Treatments are desperately needed. However, even if an effective treatment is found, it is still not a cure. Additional research funding is needed.

The CPF remains dedicated to finding a cure for this deadly disease that affects more than 128,000 people in the United States. There is a critical need to understand the history and progression of IPF and to understand better pathways that will ultimately lead to a cure. A cure can only come when the research community, the non-profit community and the government can work together.

The CPF serves as a conduit between the research community and sources of funding. To date, we have directed more than \$250,000 in research gifts to medical centers specializing in the treatment and study of IPF. When you give to the CPF, your gift goes towards this research, as well as supporting the patients and families currently fighting IPF.

The CPF relies on public contributions to further research into a cure, expand patient support and care, and increase public and professional awareness of IPF throughout the country.

We need your help!

Encourage your friends, family – even your doctor and respiratory therapist – to join the CPF – it's free!

Contribute to the CPF's efforts through a tax-deductible gift.

Raise awareness in your community about IPF - ask us how by emailing Teresa Geiger at tgeiger@coalitionforpf.org or call (888) 222-8541.

Access our website at www.coalitionforpf.org for patient information and resources, to learn about how you can be an advocate for all IPF patients, and contribute to this important work.



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Jeffrey A. Golden, M.D., is professor of Clinical Medicine at the University of California, San Francisco (UCSF). He received his BA degree from Yale University in New Haven, Connecticut, and his medical degree from Washington University in St. Louis, Missouri. Dr. Golden completed his internship at San Francisco General Hospital. He completed his residency in internal medicine and his research fellowship at the Cardiovascular Research Institute of the University of California, San Francisco, where he studied ozone inhalation in humans as a model of asthma. Dr. Golden has been on the faculty in UCSF's Department of Medicine since 1977 and is currently director of the Bronchoscopy Service, medical director of Lung Transplantation, and co-director of the Interstitial Lung Clinic.

Dr. Golden is involved primarily with clinical practice and clinical research in three main areas of interest: bronchoscopy, interstitial lung disease, and lung transplantation. He has been responsible for specific drug trials for idiopathic pulmonary fibrosis (IPF) that include a completed initial study of gamma interferon and ongoing studies with bosentan, gamma interferon, and inhaled iloprost.

The interstitial lung disease (ILD) clinic, which Dr. Golden has developed for over 25 years, includes six faculty members. The UCSF ILD program was awarded a Center of Excellence Grant from the Department of Medicine on which Dr. Golden is a co-investigator. In 2005, the program was selected to be one of 11 centers in the U.S. to receive NIH sponsorship for clinical research. Dr. Golden also initiated the UCSF lung transplant program in 1991. Since 2002 it has become one of the top 10 percent in the world in terms of yearly procedures.

Dr. Golden has been published in numerous scientific journals, including the American Review of Respiratory Disease, Chest, The Journal of Nuclear Medicine, Western Journal of Medicine, and the New England Journal of Medicine.

Supporting the CPF

The Coalition for Pulmonary Fibrosis relies on the contributions of individuals, corporations and associations who share our commitment to improving awareness and education of IPF, and improving the quality of life for patients fighting IPF nationwide. Through your generous support, the CPF will continue to provide information, resources and support to more than 128,000 IPF patients, caregivers and families, and to the healthcare professionals who treat them.

To contribute by phone using any major credit card, please call the CPF at (888) 222-8541.

Should you wish to make a tax-deductible contribution to the CPF, we encourage you to send your check or money order to:

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If you have any questions about your contribution to the CPF, or if you would like to make a restricted donation to advance specific CPF programs or research efforts, please contact CPF Executive Vice President of Development Mishka Michon at (888) 222-8541, or by email at mmichon@coalitionforpf.org.

About the Coalition for Pulmonary Fibrosis

The Coalition for Pulmonary Fibrosis (CPF) is a 501(c)(3) nonprofit organization, founded in 2001 to accelerate research efforts leading to a cure for pulmonary fibrosis, while educating, supporting, and advocating for the community of patients, families, and medical professionals fighting this disease. The CPF is governed by the nation's leading pulmonologists, individuals affected by pulmonary

fibrosis, medical research professionals and advocacy organizations. With more than 10,000 members nationwide, the CPF is the largest nonprofit organization in the United States dedicated to advocating for those with pulmonary fibrosis. The CPF's nonprofit partners include the American Thoracic Society, the Anne Harroun Landgraf Foundation, the Caring Voice Coalition, the Genetic Alliance,

the Mary D. Harris Memorial Foundation, the National Coalition of Autoimmune Patient Groups, the National Organization for Rare Disorders (NORD), The Pulmonary Paper, Second Wind Lung Transplant Association, and more than 35 leading medical and research centers nationwide. For more information please visit www.coalitionforpf.org or call (888) 222-8541.



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