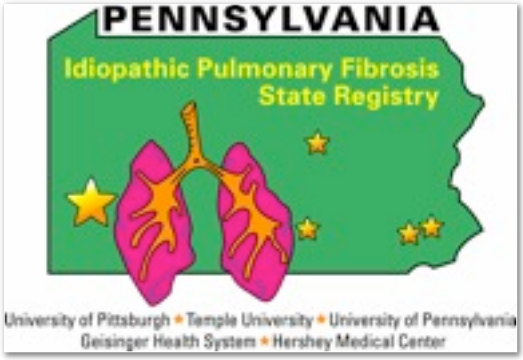
ISSUE 6

AUGUST 4, 2010

the PA-IPF Newsletter

A PROJECT SPONSORED BY THE COMMONWEALTH OF PENNSYLVANIA AND A GENEROUS DONATION FROM THE SIMMONS FAMILY



WELCOME

The PA-IPF newsletter is a part of the Pennsylvania State Idiopathic Pulmonary Fibrosis (PA-IPF) registry. The purpose of this newsletter is to provide you with up-to-date information about IPF research as well as current events to increase IPF awareness in the Commonwealth of Pennsylvania.

Share Your Story

If you would like to tell an inspirational story from your own experience with IPF, or as a caretaker for a patient diagnosed with IPF , please email your story to registry coordinator ,Trisha Black at blacktr@upmc.edu. It may appear in the PA-IPF newsletter and someone also with IPF may benefit from reading your story.

To Our Readers

Scientists across the nation and around the world are conducting research geared towards finding the cause of IPF as well as looking for a cure. One of the missions of the PA-IPF registry is to provide patients, family members, and physicians with a resourceful newsletter for IPF patient care. We hope that you enjoy the content, and we will keep you aware of our progress as the registry continues to grow. We invite you to respond to our newsletter with feedback, comments, or general questions. Please send your requests to Trisha Black, registry coordinator by email blacktr@upmc.edu or mail to the Dorothy P. & Richard P. Simmons Center for Interstitial Lung Diseases NW 628 UPMC Montefiore 3459 Fifth Ave. Pittsburgh PA 15213.

In This Issue

- **A Grandson's Journey to Finding a Cure for IPF**- Page 2
- **Pulmonary Hypertension-** Page 3
- **Golf Outing and Boat Cruise-** Page 3
- **PA-IPF Institutions**- Page 5-6

Editor:
Trisha Black, BA, research coordinator at the *Dorothy P. & Richard P. Simmons Center for ILD in Pgh, PA*

"The past is a source of knowledge, and the future is a source of hope. Love of the past implies faith in the future."

A Grandson's Journey to Finding a Cure for IPF

written by Marcus Magister

Three summers ago, in 2008, I learned about the awesome research that was being done at Dorothy P. & Richard P. Simmons Center for Interstitial Lung Disease at UPMC not from a help-wanted ad, but from a patient of the Simmons Center – my grandfather. At the time, my family and I had been living with a family member diagnosed with IPF for a number of years. Being the curious type of person I am, I never could get enough information about the disease and always wanted to know more. When I learned about the research lab, I was extremely excited to try to get involved in some way. Little did I know what was in store for me over the next three years! I've worked on various projects, many of which have dealt with gene regulatory molecules called microRNA's. My findings have sparked ideas on how a growth factor, TGFβ, affects these microRNA's. I hope one day, I can be a part of a useable treatment for IPF – one that extends the life expectancy after diagnosis or maybe even cures the disease all together.

Eventually, I would like to become a doctor. Though I have not determined a specialty yet, working in Dr. Kaminski's research lab has opened my eyes a great deal to pulmonology as well as the possibility of becoming a researching physician. In addition to being a great beginning step on my way to becoming a doctor, I am extremely happy to say that those who I work with everyday are among some of the best people I know. Ever since my first day, I was amazed at the immense compassion everyone in the Simmons Center has for those who suffer from the disease. This was something I thought only people like myself who were touched by the disease personally could comprehend, but I was wrong. I've forged relationships and friendships with many of my co-workers that will last for many years to come!

This year, Marcus attended the American Thoracic Society Conference in New Orleans as a co-author on a poster by Jadranka Milosevic PhD titled: The role and regulation of miR-154 family in Lung Fibrosis.

As I've mentioned, I originally learned about the Simmons Center through my grandfather. Since I began working in the research lab, my family and I have unfortunately suffered a tragic loss. My grandfather passed away in September of 2008, shortly after my first summer working in the lab came to a close. Though this was hard, it too became a learning experience for me. I've pushed through, and ever since, my determination and resolve to further the information we have of IPF has grown and led to better treatment of IPF.

Marcus, his Uncle Mike Magister, and Dr. Naftali Kaminski discuss IPF research at the Simmons Center, during the 5th Annual Fran Magister FORE IPF golf outing in August 2009.

In 2009, the Simmons Center renamed their annual golf outing in memory of my grandfather, Francis J. Magister. My family and I were extremely pleased to accept this honor especially because we knew how much the golf outing meant to him. Fran was instrumental in beginning the first golf outing and helping to get it to where it is today. This outing supports everything IPF, and I sincerely encourage everyone to get involved in some way with this great event.

If you're reading this newsletter, you have probably been affected by IPF in one way or another. Despite the long, and arduous road to a cure, I am hopeful that one day we will find one. And in doing so, give the many who suffer the answer they so rightfully deserve.

Pulmonary Hypertension

Written by: Hunter Champion, MD, Ph.D. Associate Professor of Medicine, Division of Pulmonary, Allergy, and Critical Care Medicine University of Pittsburgh

What it is: Simply put, pulmonary vascular disease (PVD) and pulmonary hypertension (PH) refer to an elevation of blood pressure in the lungs. PH is associated with a number of heart and lung conditions, including interstitial lung disease and idiopathic pulmonary fibrosis. It is generally believed that the increase in lung blood pressure is because of lack of oxygen in the blood, we now know that patients with any stage of ILD or IPF can have pulmonary hypertension. We believe that PH is a major cause of exercise-limiting shortness of breath in our patients with ILD. If undetected and untreated, the blood pressures in the lungs can increase to a point that the heart becomes weakened and patients begin to experience heart failure.

What are the symptoms?: Generally, the symptoms are slow in progression and include fatigue and shortness of breath (particularly with exertion). Patients may also experience chest pain, dizziness or even pass out with exertion. If the heart is affected by the lung pressures, patients may experience leg or abdominal swelling.

How is it diagnosed?: Fortunately the screening for pulmonary hypertension is non-invasive. Screening is generally performed by cardiac ultrasound (echocardiogram). The ultrasound technician is able (in most patients) to estimate the pressures in the lung. While the screening is non-invasive, the confirmation of the diagnosis must be made by a cardiac catheterization. The catheterization is performed on an outpatient basis and takes less than one hour to perform.

In addition to the echocardiogram and catheterization, it is critical that other tests be performed to make sure that the pulmonary hypertension is not caused by other conditions. These additional tests include blood work, chest X ray, CT scans, and other noninvasive tests.

Can it be treated?: We now have 9 drugs approved by the FDA for the treatment of certain types of pulmonary hypertension. These drugs have a number of different delivery routes (oral pills, inhaled therapy, subcutaneous, and intravenous) and side effect profiles. All of these medicines are generally well tolerated. Clinical studies have shown that patients taking these medications walk farther than patients not on therapy. For specific conditions, we can only treat pulmonary hypertension as a part of a clinical research protocol. At the University of Pittsburgh, we have a number of clinical protocols that we can offer to patients who may not be appropriate for FDA approved therapies.

How do you go about being screened?: Even more important than awareness of pulmonary hypertension is making sure that screening is performed. Many patients with interstitial lung disease (ILD, idiopathic pulmonary fibrosis (IPF), or sarcoidosis are already screened with cardiac echocardiograms on an annual or semi-annual basis. If there is a concern for pulmonary hypertension, patients can be seen in our integrated pulmonary hypertension program in which patients can be seen in the same clinic by a pulmonary hypertension specialist in cardiology and/or pulmonary medicine. The pulmonary hypertension program offers clinic times on Tuesday afternoons and Thursday mornings. Many of our patients can be seen on the same day that they see their Simmons Center caregivers.

How a clinic appointment can be scheduled: A call (1-877-PH4-UPMC) or email (PHprogram@upmc.edu) is all that it takes. We strive to offer appointments within 72 hours of your call.

More information can be found on our website: http://

Two FUN Upcoming Events!

August 30, 2010

Fran Magister "FORE IPF" Golfing Outing

Date: Monday August 30, 2010
Where: Pittsburgh National Golf Club (formerly Deer Run) 287 Monier Rd. Gibsonia
What: Golfing, Dinner, and Prizes!
If you're an IPF patient you golf for free!

To participate in or sponsor this event please contact Mary Williams 412-647-3156.

October 7th, 2010

7th Annual Gateway Clipper Boat Cruise & Luncheon

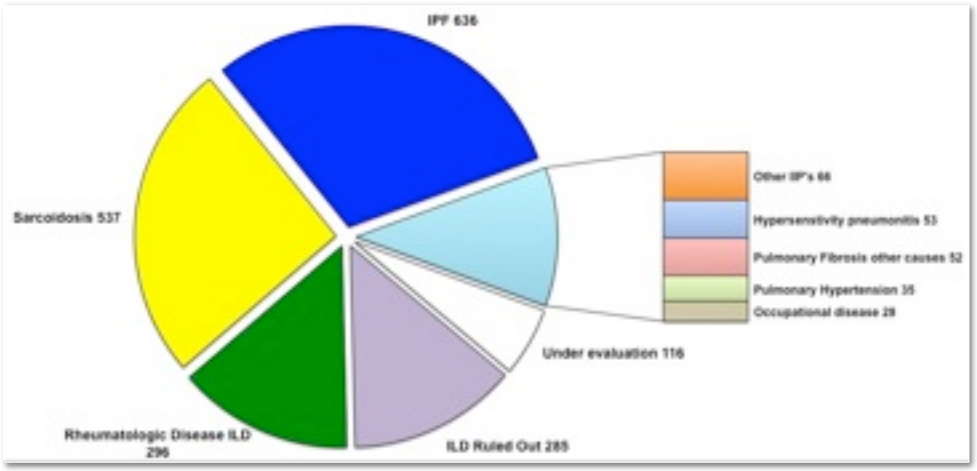
Date: October 11, 2010
What: A free lunch with fellow patients, pulmonologists, researchers, and staff of the Dorothy P. & Richard P. Simmons Center
Boarding Time--12:00 pm
Sailing Time--12:30
Docking Time--2:30

Please contact Mindy for more information 412-802-6860.

PA-IPF Institutions

The five institutions described in this section create an extensive network across the state of Pennsylvania. Each center aims to provide advanced comprehensive care and access to cutting-edge research to patients with interstitial lung disease.

The Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease at the University of Pittsburgh Medical Center Increases IPF Awareness



Distribution of diagnoses of patients with Interstitial Lung Disease at the Simmons Center- August 5, 2010

The pulmonary doctors at the Simmons Center treat patients with Interstitial Lung Disease (ILD). A majority of the patients have IPF/UIP, sarcoidosis, or pulmonary fibrosis associated with a rheumatoid condition, as shown on the diagram to the above. The center follows more than 1,900 patients with Interstitial Lung Disease. The ILD experts at the Simmons Center work closely with a multi-disciplinary team of experts in pathology, radiology, rheumatology, quality of life and lung transplantation to provide the best care. Physicians and scientists at the Simmons Center are recognized as international leaders in research on mechanisms of lung inflammation and fibrosis, and promoting the translation of scientific discoveries into new treatments for patients with these lung disorders. The Simmons Center was established through a generous donation from the Simmons family. Research funding is provided by the National Institute of Health (NIH), industry sponsors and private donations. Because there is no approved therapy for IPF, the scientists and physicians at the Simmons Center dedicate their research to finding a cure for IPF. In the last 5 years scientists at the Center have been involved in multiple studies and published more than 70 research papers in the top medical journals that are expected to have a strong impact on patient management and care. You can follow these journal updates on the Simmons Center's twitter page at www.twitter.com and look for simmonscenter

Temple Lung Center a Patient Centered Facility

Growing recognition of Temple Lung Center's quality care has prompted more patients to come to Temple over the past decade. To meet growing demands, Temple has moved into a spacious new outpatient facility. The 18,000 square foot facility provides patients with access to the most advanced capabilities for disease management and the opportunity to participate in cutting-edge therapies offered by clinical research trials. Located in the Ambulatory Care Center at the Temple University Hospital, this new facility allows patients to receive all their care in one convenient spot. Services include, office visits with Temple pulmonologists in one of 22 exam rooms, routine pulmonary function and exercise testing, blood draws, electrocardiograms, sleep and non-invasive ventilator equipment instruction and titration, and an on-site rehabilitation center.

Patients requiring evaluation or treatment for lung transplantation or other medical-surgical conditions, consultations with Temple's multidisciplinary team, including cardiologists, surgeons and other specialists, take place in one location.

Temple's clinicians helped ensure that the new space was designed around the special needs of pulmonary patients. This patient-centric design is reflected in the layout of the unique spaces and thoughtful details. Patients are often concerned about depleting their portable oxygen tanks when they leave home for an extended period, so multiple oxygen hook-ups throughout the clinic, waiting rooms, and exam rooms make visits more convenient. The lack of carpeting throughout the space also makes appointments easier for those using wheel chairs. Other touches to make patients more comfortable include flat-screened televisions located throughout the open waiting area as well as a complimentary coffee service.

The state-of-the-art waiting room is outfitted to be a "smart classroom" for patient and family support groups, disease management lectures, and educational programs for community physicians. The Temple Lung Center staff takes pride in its new home because it is a physical manifestation of the patient-centered philosophy that drives the entire Temple staff.

The Penn State Milton S. Hershey Medical Center-Occupational Research Interest

Penn State Hershey Medical Center was established in 1967 and is located in Hershey Pennsylvania, 10 miles east of Harrisburg. Penn State Hershey, has a specific focus on rural medicine. The Division of Pulmonary, Allergy and Critical Care Medicine has ten faculty members and nine fellows who care for patients with interstitial lung disease. New emphasis is being developed in environmental and occupational respiratory research, pulmonary hypertension associated with interstitial lung disease, and collaborating with basic scientists to identify determinants of susceptibility to respiratory diseases.

The Penn Lung Center Taking on ILD Initiatives

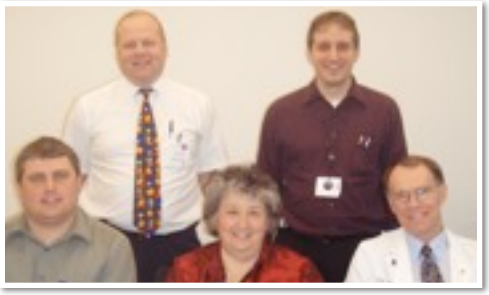
The Penn Lung Center is now housed in the Ruth and Raymond Perelman Center. The Perelman Center is a state-of-the-art, 500,000 square foot outpatient facility adjacent to the Hospital of the University of Pennsylvania. The Perelman Center links Penn's expert physicians and clinical researchers in new ways, by putting them just an idea's reach away from one another, always prepared to collaborate and create groundbreaking, individualized treatment plans.

The Penn Lung Center brings physicians together in new ways that will lead to closer collaboration between specialists. Patients, in turn, can expect quicker diagnosis and treatment. In the Lung Center, for example, a thoracic surgeon can upload a patient's high-resolution CT scans to an imaging center 10 feet away from the exam room. There, the doctor can confer about the images with an expert chest radiologist, and together the two will help craft a personalized treatment plan for the patient. The care team will relay the plans to the patient right away, paving the way for quick access to a team of other specialists – nutritionists and social workers, for instance – without leaving the department. If the patient requires further testing, they're just a short escalator ride away from the appropriate testing areas.

Currently, the Penn Lung Center participates and/or coordinates research studies for IPF, cancer, bronchiectasis, and lung transplant among others. According to Dr. Gregory Tino, Chief, Pulmonary Clinical Service, "the Lung Center is the culmination of our vision and dedicated effort to build a comprehensive, multi-disciplinary practice to provide the highest quality of care to patients with diseases of the chest. The Lung Center will also enhance our clinical research endeavors."

To make an Appointment at the Perelman Center call 1-800-789-PENN.

Geisinger Center for Health Research a "Green" Facility

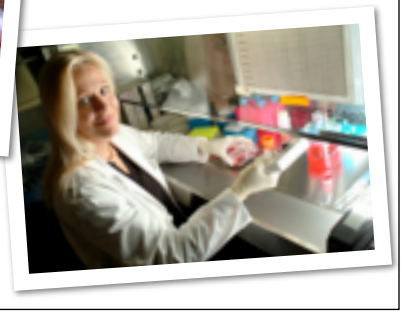


This institution is one of the nation's leading fully integrated healthcare providers. Founded in 1915, Geisinger serves more than two million residents throughout central and northeastern Pennsylvania. The Physician-led organization is at the forefront of the country's rapidly emerging electronic health records movement. Geisinger is a leader in the region's green building initiatives. Geisinger's Pulmonary Medicine Department includes board-certified pulmonologists supported by a staff of registered respiratory therapists and registered nurses. With extensive experience in the management of chronic obstructive pulmonary disease (COPD) and lung tumors, this team is recognized both nationally and locally as leaders in pulmonary care.

ISSUE 6
AUGUST 4, 2010

the PA-IPF Newsletter



PA-IPF *Newsletter*
Dorothy P. & Richard P. Simmons
Center for Interstitial Lung Disease
NW 628, UPMC Montefiore
3459 Fifth Ave.
Pittsburgh, PA 15213