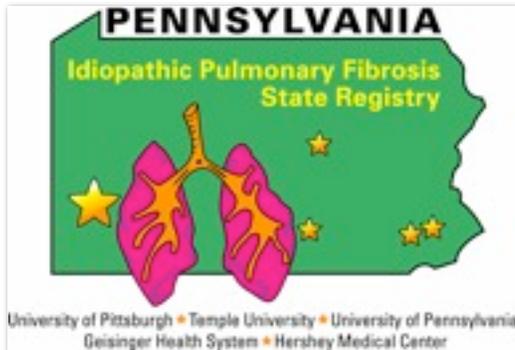


I S S U E 5

JANUARY 27, 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.*

## TO OUR READERS

*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](mailto:blacktr@upmc.edu). It may appear in the PA-IPF newsletter and someone with IPF may also benefit from reading your story.*

*leahleah*

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. 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](mailto: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.

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### EDITOR:

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



*They may forget what you said, but they will never forget how you made them feel.*

-Carl W. Buechner

# LIVING WITH IPF

Written by: PF Simonelli, MD, Ph.D.  
Director, Thoracic Medicine, Geisinger Health System



Dr. Simonelli is an expert in pulmonary fibrosis. He treats IPF patients at Geisinger Health System in Danville, PA.

Myrna was the first person with IPF whom I ever met. It was the beginning of my pulmonary fellowship, and she was a newly assigned patient in our chest clinic. She was an older, retired cleaning woman who lived locally in upper Manhattan. Myrna described becoming more short of breath over a long period of time, often in fits and starts, but progressively worse over time. By the time that I met her, she was already short of breath just walking from the waiting room. She had been to an emergency room many times over the course of the prior few years, and each time she was treated as if she had asthma, and sent home with instructions to use a variety of inhalers. At the end of our first meeting, I remember telling her that I didn't yet know what she had, but it wasn't asthma.

She missed her next appointment because she was hospitalized due to a collapsed lung that ultimately required surgery for repair. When I learned about this during our next meeting, I explained that I wished I knew about this event, because a lung biopsy would have helped explain what was wrong, and they could have done the biopsy at the time of the repair. As fate would have it, the other lung collapsed before her next appointment. As they were wheeling her into the operating room, she told the surgeon "my doctor is Paul Simonelli, and he said that if this ever happened again, I should have a lung biopsy". She got her biopsy. Unfortunately, it confirmed the impression of IPF.

I cared for Myrna for the next many months. I prescribed prednisone as it was the standard therapy of the time, and supplemental oxygen. She continued to worsen, but she never missed another appointment, each time lugging her oxygen and as usual, a big shopping bag. I finally learned what was in the bag during one visit after I explained that I would be away in New Orleans for a meeting when I would otherwise be scheduled to see her again. She reached into the bag and pulled out a brochure for K-Paul's, telling me that I had to go there. It is still one of my favorite restaurants in that city. It turns out that Myrna was a gourmet cook – who would have guessed – and she collected and carried around all sorts of cooking-related brochures and recipes.

I learned so much about patient care, and IPF, from Myrna. Firstly, IPF is usually misdiagnosed, often for a period of years. Most of the patients whom I have seen with IPF were first told that they had asthma, COPD, heart disease, or any other number of problems. Physicians are getting better in this regard, but getting the word out about this disease remains one of our challenges. I learned that conventional therapies like prednisone don't much help in the long run (there can still be a role for short periods in the treatment of IPF). She taught me how important it is for patients to be involved in their care. After all, she was the one who managed to "arrange" to get her lung biopsy. Most importantly, she taught me that one can – and should – lead their life, despite IPF. I never heard her complain once, although she could have. What she did express was her joy

in living each day as long as she could.

Our knowledge and skill in treating patients who have IPF is light-years ahead of where it was when I met Myrna.

Unfortunately, we're still far from truly understanding and treating this disease effectively. So what are the positives? To start, there is a remarkable effort in Pennsylvania that has the goal of identifying just who does get this disease, and possibly why.

This is the PA-IPF registry, an effort funded by the state to address what has for too long been an ignored problem. Imagine, five competing academic medical systems coming together with the state's help to share knowledge and resources to help understand and combat a disease like IPF. This type of effort is almost unique. We're still at a point of gathering information about those who have IPF, but our hope is to soon offer trials of new and promising therapies to the group at large, as such opportunities arise.

Our ability to diagnose IPF, and to distinguish it from several other similar diseases has improved remarkably. This is important, as many diseases that resemble IPF might have already-existing effective therapies. Using a therapy where it has a good chance of success is worthwhile, even though many of the therapies (such as prednisone) have important undesirable side-effects. On the other hand, avoiding therapies that are not likely to work is also important; side effects without much benefit are all too often the result of using conventional therapies to treat IPF. Even the need for biopsies has decreased, as we are often able to confidently make a diagnosis of IPF without needing a biopsy, something that was not possible at the time when I met Myrna.

So what does one do if they have IPF. First, get an accurate and secure diagnosis. Very often, this means seeing someone who has considerable experience in managing this disease. Secondly, take very good care of yourself and don't ignore the rest of your health. At least as many people with IPF who succumb die because of some other problem. Thirdly, don't use unproven therapies unless they are being tested as part of a clinical trial. Unproven therapies - there are many – are not likely to help, and may make things worse. In contrast, medications that reach clinical trial testing have been examined quite rigorously, and those who are enrolled in such trials are watched extremely closely for complications or worsening of their disease. Fourthly, stay as active as you can: maintaining muscle fitness is one of the best ways to combat the effects of IPF on one's ability to get things done. For some, using supplemental oxygen can help with this goal. And lastly, live your life to the fullest – like Myrna, never give up.

# Traveling with Oxygen written by Patricia Jennette

The word “travel” for those on oxygen often evokes stress and confusion. There is the question, “Can I travel with oxygen?” followed by the next question, “Can I take my own oxygen supply with me?”

For those traveling by vehicle, the answer is generally ‘yes’ to both questions. However, for those who want to travel by other means, particularly by airplane, there are some challenges.

The challenges are not insurmountable, and the opportunity to take a vacation can be accomplished with some planning and a bit of research.

Such has been the case for my husband John Jennette, who has been part of the IPF Support Group at the Simmons Center since the group’s inception.

John and I are avid travelers in between running a community magazine publishing business.

Most recently, we planned, implemented, and completed a two-week journey to the islands of Hawaii to celebrate our silver wedding anniversary.

John opted to rent an Inogen portable concentrator from Blackburn’s Physician’s Pharmacy. Batteries and 120 AC current power the machine. These batteries allow the machine to make its own oxygen. It is a widely acceptable and approved oxygen equipment option by the major airlines.

Given that Hawaii is made up of many islands, the concept of hopping islands by airplane wasn’t the easiest choice.

John and I decided on a trip that would allow us to visit the islands by cruise ship. NCL’s Pride of America was built specifically for cruising the Hawaiian Islands, and was our choice of travel for the first week of the journey. Pre-notification to the cruise line that John was bringing Inogen made for a smooth registration at the dock during pre-boarding.

The cruise line only allows liquid or cylinder oxygen bottles on board, supplied by their vendor to insure safety. They also allow portable concentrators such as the Inogen.

Making airplane reservations to fly from Pittsburgh to Honolulu was kept simple. I decided that the fewer

legs in the journey there were, the less hassle it would be to get from one gate in the airport to another. We booked a flight from Pittsburgh to Phoenix, non-stop, which was about four hours. A two-hour layover in Phoenix allowed John time to plug in his Inogen for recharging and to take a quick break for a mid-morning meal before getting onto the 6-1/2-hour flight to Honolulu. Again pre-notification to the airline that John was carrying an Inogen machine was helpful. Also, the airline required that John bring a written prescription, called an “In-flight Authorization Form,” from his physician confirming that he was required to use the oxygen and could carry the Inogen on board the flights. It is helpful to bring along extra copies of this form. At the gate for each flight, John simply showed the In-flight Authorization Form to the airline personnel. They also allowed him to board the flight in advance in order to get the concentrator machine under the seat and settled properly before other passengers got on board.

Be aware that it is important to have plenty of batteries for a trip this long, as airlines don’t provide a means to recharge batteries.

Upon arrival in Honolulu, we arranged for shuttle transportation via a shuttle to their hotel in downtown Waikiki. We made sure the hotel had elevator accessibility and was close enough to restaurants and other amenities, since walking would be factored in once we arrived at the hotel.

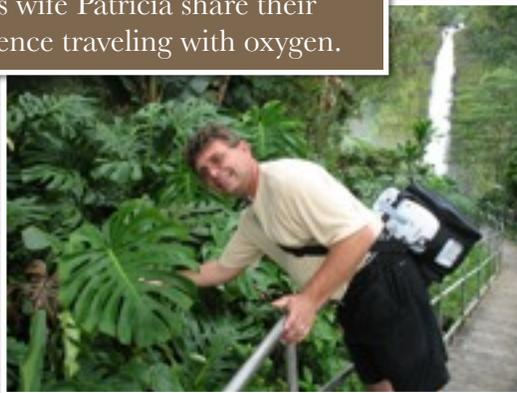
After a one-night stay, we took a pre-arranged shuttle to the cruise dock, which took us directly to the entrance of the pre-boarding area.

Suitcases with easy to use wheels and pullout handles made it less cumbersome to get from the shuttle through the pre-boarding area. John was able to pull one suitcase and carry his Inogen on his shoulder, and he attached a backpack onto the suitcase handle for his carry-on from his flight.

Throughout the cruise, the concentrator proved to be a valuable asset in order to enjoy the many opportunities to see the sites on the islands.

For example, we visited the Haleakala National Park on Maui. An ascent by rental car to the top of the 10,000-foot-high mountain to see the sunrise above this dormant volcanic crater was worth every minute. John

When John was diagnosed with IPF, he thought traveling was out of the question. In this newsletter, John and his wife Patricia share their experience traveling with oxygen.



was quick to admit that this height challenged him to use the maximum output of the concentrator machine. However, by pacing himself and moving modestly once we reached the visual points at the top of the mountain, he was able to enjoy everything that all the other visitors enjoyed.

Individuals who would like to go to sights such as this that have high elevations or long strenuous walks should always consult with their physician first.

The Road to Hana, also on Maui, is a very enjoyable ride by rental car that takes a full day. Slow going, with more than 50 one-lane bridges to cross, it is filled with tropical beauty, waterfalls, and overlooks of the Pacific Ocean.

On the Big Island of Hawaii, John and I were able to enjoy a tropical rainforest side trek near Hilo on the way to Akaka Falls. These falls are immersed within the rainforest, and a very easy to navigate walkway and stairs, with rails, makes it possible to enjoy with the use of the Inogen machine.

We next went on to Volcanoes National Park, the only live volcano on the Hawaiian Islands at this time. There, some caution is advised, as there are some areas where the steam vents emit sulfur and breathing the atmosphere should be done with caution. Depending on the downdrafts and the winds, one can enjoy the sights at various points throughout the park with no worries. The park's lookout points are on the level, and are quite easily reachable by car. A ranger station as well as Jagger Museum at one overlook provided excellent educational opportunities to learn about the park.

The island of Kauai is an excellent choice to enjoy the sites by vehicle, with very little challenges for oxygen use. Among the most popular sites is Waimea Canyon. Known as the Grand Canyon of the Pacific, the canyon is an enjoyable drive with numerous lookout points, all easily accessible and with parking areas that are relatively close to the lookouts. The height of the canyon does not pose any breathing challenges, and John found that the Inogen did an excellent job on this trip.

Also on Kauai are numerous beaches that can be enjoyed along the outer edge of the island on an easily navigable major highway.

While taking long drives, the concentrator can be run from the cigarette lighter to be charged, something that can't be done on bus tours and taxis. Renting a car was the most economical as well as convenient way to travel on oxygen. Plus, much of the scenery can be viewed from inside the vehicle with the windows up, if necessary.

The other major highlight on Kauai is the Na Pali coastline. While many adventurers will trek the trails to reach this gem, those traveling on the Pride of America will be able to enjoy a two-hour cruise past this coastline at the end of their two-day stay in Kauai.

Arriving back in Honolulu, John and I checked into a Sheraton Hotel one block from Waikiki Beach, again making sure it was accessible to restaurants and other amenities. They spent four days on Oahu, visiting Pearl Harbor, the Polynesian Cultural Center, the North Shore, Dole Pineapple Plantation, and Hanauma Bay. All of these sites are completely accessible by car and navigable by foot using the concentrator. We also hiked to the top of the Diamond Head Crater. The Inogen proved to be absolutely necessary for this journey, which John admitted, was among the most challenging activities for him throughout the entire vacation. It took some time to ascend, and several times to stop and rest, before reaching the top.

Individuals with lung and heart conditions also have to be aware of ascents to high elevations – by car, plane, or helicopter – can rise and descend quickly at times. It is important to do each very slowly to let one's body get acclimated and to monitor one's condition. And, it is always a good idea to carry a pulse oximeter since some of these changes are subtle and deceiving.

Also, when traveling on long flights or in a long car ride, one should stand and stretch their legs and walk around to avoid blood clots forming in their legs.

After two weeks, we returned home, first traveling the 6-1/2-hour

trip to Phoenix, and then the four-hour trip back to Pittsburgh.

With a little bit of planning, pacing the travel agenda,

and making sure that all arrangements were made in advance, they were able to enjoy all that Hawaii has to offer, all the while carrying the concentrator with us in the car, the airplane, or on foot.

As always, tell one's physician what travel plans are being made so they can advise.



# ARRA FUNDING FOR GROUND-BREAKING IPF RESEARCH

*The American Recovery and Reinvestment Act of 2009 (ARRA or “Recovery Act)* is allocating over \$12,405,765 towards IPF research. Listed below some of the currently funded research studies. Please visit [projectreporter.nih.gov](http://projectreporter.nih.gov), and enter idiopathic pulmonary fibrosis in the “term search” box for more information about these and other studies.

**Lung Genomics Research Consortium--** A consortium of National Jewish Health, University of Pittsburgh, Boston University, and Harvard are planning to create a genetic, molecular, and quantitative clinical phenotyping data warehouse using bioinformatic tools. This platform will empower investigators to make fundamental discoveries in disease pathogenesis, refine diagnostic criteria, and lead to advances in personalized medicine. They are extending the National Heart, Lung, and Blood Institute (NHLBI), Lung Tissue Research Consortium (LTRC) biorepository and creating the Lung Genomic Research Consortium (LGRC) to advance research discoveries. The goal is to: 1) identify individuals at risk of developing chronic lung diseases; 2) diagnose these conditions at an earlier stage; 3) identify novel mechanisms that cause chronic lung disease; and 4) eventually develop personalized therapeutic strategies for intervention.

**CTRIP: Molecular phenotypes of rapidly progressive idiopathic pulmonary fibrosis—**University of Michigan is collecting multi-specimens from a population of 135 IPF patients. The samples will be analyzed by the Nation’s leading IPF scientists and clinical research teams, to identify the early indicators of health stability. The results of this study will allow physicians to diagnosis patients sooner, and provide patients with optimal treatment.

**Genome-wide association and exon sequencing study in IPF—**The University of Chicago and the University of Pittsburgh are planning to recruit over 700 IPF patients to identify inheritable genetic factors that may affect an individual’s susceptibility to IPF. The identification of the genes and the specific genetic variants can be used as a guide to lead to new approaches for preventing and treating this deadly disease.

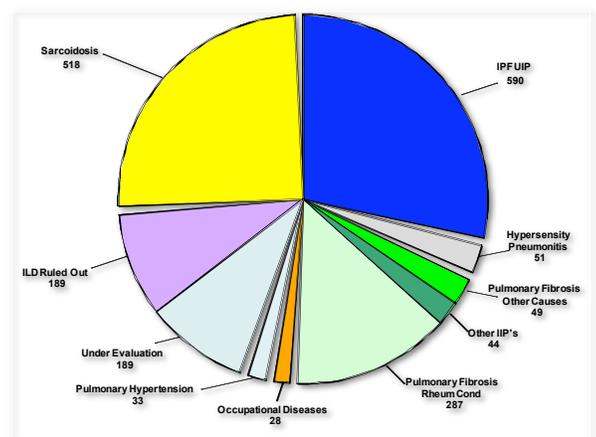
## PA-IPF Institutions

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

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 right. 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

**Distribution of diagnoses of patients with Interstitial Lung Disease at the Simmons Center- January 26, 2010**



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](http://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, the newest medical school in Pennsylvania, 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.



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**PA-IPF** *Newsletter*



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Center for Interstitial Lung Disease  
NW 628, UPMC Montefiore  
3459 Fifth Ave.  
Pittsburgh, PA 15213