

# COLLAGEN CONNECTION

Summer 2013

## DR. CAROL FEGHALI-BOSTWICK ACCEPTS NEW POSITION



Dr. Carol Feghali-Bostwick has accepted the Kitty Trask Holt SmartState Endowed Chair

Professorship at the Medical University of South Carolina (MUSC), in Charleston, SC. She and her laboratory team will be relocating to MUSC in October 2013. MUSC became a Scleroderma Center in 1975 under the leadership of Professor E. Carwile LeRoy, who was the first to demonstrate the overproduction of collagen in skin cells (fibroblasts) from systemic sclerosis (SSc) patients grown in tissue culture and the first to propose a "vascular hypothesis" for the pathogenesis of SSc. Dr. Feghali-Bostwick will be joining Dr. Richard Silver, the Chief of the Division of Rheumatology and Immunology at MUSC. His group was among the first to examine cells from the lungs obtained by bronchoalveolar lavage for the study of lung disease in SSc, and the first to use cyclophosphamide as a treatment for SSc lung disease. The Division

is known for its outstanding patient care and for training clinicians and investigators, many of whom have gone on to establish research and teaching programs in scleroderma at other institutions.

Dr. Carol Feghali-Bostwick will also serve as Co-Director of the MUSC Scleroderma Center. She will continue her ongoing research into the cause of SSc and will pursue the development of E4, a peptide her group identified as a potential treatment for fibrotic disorders including SSc.

Dr. Feghali-Bostwick spent 20 years at the University of Pittsburgh doing SSc research. Faculty members and staff at the Pittsburgh Scleroderma Center wish her the best and hope to continue collaborating with her on several research projects currently in progress.

## ARTHRITIS FOUNDATION TRANSFERS ENDOWED RESEARCH FUND TO UNIVERSITY OF PITTSBURGH SCHOOL OF MEDICINE

The Arthritis Foundation, Great Lakes Region has completed transfer of nearly \$360,000 in endowed funds to the University of Pittsburgh's Division of Rheumatology and Clinical Immunology. The interest accrued by this endowment is restricted to support scleroderma research.

The endowment was created by the late G. Albert Shoemaker in 1983 in memory of his first wife, Jean O. Shoemaker. It has been a welcome source of funding for many scleroderma research projects over the past 30 years. Mr. Shoemaker's widow, Mercedes Shoemaker, has generously supplemented the endowment in past years. She is a member of the Scleroderma Center's Patient Advisory Group.



June is Scleroderma Awareness Month

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**HELLO**  
my name is

*Jennifer Peat*



The Scleroderma Center is pleased to introduce you to its new Clinical Care Coordinator, Jennifer Peat.

Jennifer coordinates new patient visits to our center and also to the Division of Rheumatology and Clinical Immunology's other centers devoted to Myositis, Lupus, Rheumatoid Arthritis and Vasculitis. New patients often need to see specialists with whom our faculty work in renal disease, pulmonary, gastroenterology, cardiology, etc. She arranges these additional appointments and helps patients to decide on travel, accommodations and restaurants. She is also responsible for coordinating the Rheumatology Telemedicine Program.

Jennifer is a graduate of Lenape School of Practical Nursing and has 10 years of experience in geriatric nursing. She is furthering her education in the nursing program at the Community College of Allegheny County. In her spare time she loves to travel and create wheel-thrown pottery. Welcome Jennifer!



# SCLERODERMA FOUNDATION

2013 NATIONAL PATIENT EDUCATION CONFERENCE

JULY 26 - 28, 2013  
HYATT REGENCY ATLANTA  
ATLANTA, GEORGIA



Join us for the 2013 National Patient Education Conference and celebrate the Scleroderma Foundation's 15th Anniversary!



*"KNOCK OUT SCLERODERMA"*  
FIFTEEN YEARS OF FIGHTING

## CURRENTLY RECRUITING PATIENTS FOR CLINICAL TRIALS

The Scleroderma Center is participating in two active multicenter clinical drug trials. Pittsburgh is a logical site to consider for participation if you reside in this area. If you live in another part of the country, you may wish to contact the participating site closest to you for additional details. For more information on these studies please visit 1) <http://clinicaltrials.gov>, or 2) contact our Research Coordinator, Dana Ivanco, at 412-648-7040 or [des2@pitt.edu](mailto:des2@pitt.edu).

**Rituxan:** This is an immune suppressing drug currently used by hematologists for certain malignancies such as lymphoma. It is also approved for use in rheumatoid arthritis. Rituxan eliminates B cells from the blood stream. These cells participate in immune responses and may be responsible for some types of immune injury to tissues in patients with rheumatoid arthritis, lupus, and other related diseases, including Scleroderma. It is given by vein twice, two weeks apart. This

study is directed at Scleroderma patients who have confirmed pulmonary arterial hypertension (PAH or high blood pressure in the lungs), regardless of how much skin thickening they have. Half of the patients will receive Rituxan and half placebo. Other PAH medications can be continued throughout the study. Patients will be followed for 1 year or more.

**Pomalidomide:** This drug has the potential to reduce fibrosis (formation of excessive scar tissue) in the skin and lungs. Pomalidomide is currently being studied in a number of diseases, including different types of cancers, and is FDA approved for the treatment of multiple myeloma (a malignancy of the bone marrow). The purpose of this study is to examine the safety and efficacy of a daily oral form of Pomalidomide in patients with early scleroderma (< 5 years of symptoms) who also have interstitial lung disease (pulmonary fibrosis). Half of the patients will receive Pomalidomide and half placebo. Patients will take the drug or placebo once daily for 52 weeks in this study.



# American College of Rheumatology Annual Meeting

The annual meeting of the American College of Rheumatology (ACR) was held in Washington, D.C. in November 2012. Investigators from around the world presented results of recent basic and clinical research on systemic sclerosis and localized scleroderma, as well as other rheumatic diseases. There were 4 presenters from the University of Pittsburgh Scleroderma Center.

Dr. Thomas A. Medsger Jr. gave the Paul Klemperer, MD Memorial Lecture on autoantibodies in scleroderma and their usefulness in patient subsetting and predicting outcomes.

Dr. Robyn Domsic discussed autoantibodies in clinical practice during 2 Meet the Professor sessions. She was the primary author on 2 abstracts regarding

early diffuse systemic sclerosis and co-author on 2 abstracts for the PHAROS (Pulmonary Assessment and Recognition of Outcomes in Scleroderma).

Dr. Kathryn Torok was co-author on 5 abstracts regarding pediatric localized scleroderma, 2 of which focused on localized scleroderma affecting the face and scalp, termed 'en coup de sabre' or Parry-Romberg syndrome.

Dr. Christine Peoples presented her work on "Gender Differences in Systemic Sclerosis" and participated in the ACR Knowledge Bowl with Dr. Ximena Ruiz, a rheumatology fellow at the University of Pittsburgh who is working on a scleroderma kidney involvement project in the Center.



Dr. Christine Peoples discusses Gender Differences in Systemic Sclerosis



Drs. Ruiz and Peoples compete in the ACR Knowledge Bowl



Dr. Kathryn Torok at her poster on Pediatric Localized Scleroderma.



## Walk with Tori

The annual walk will be held at Doubs Woods Park in Hagerstown, Maryland on September 8th. All monies raised from the event will be used for scleroderma research. Last year, Tori and her team raised nearly \$40,000! For more information please email:

[walkwithtori@yahoo.com](mailto:walkwithtori@yahoo.com)





Martha and Dave Merry enjoying Maui

## A PATIENT GIVES BACK

When Martha Merry first began feeling the effects of scleroderma on her body, she had no idea what was happening. An avid equestrian, Martha was in the midst of a lengthy major horse competition when she felt her energy level quickly and suddenly being drained. “When it was over, I literally got off the horse, handed him over and collapsed,” Martha remembers.

just one month. “Carol really was the key to getting treatment started so quickly. I don’t know what we would have done without her.” Martha’s husband Dave remembers

During her first visit with Dr. Medsger in April 2003, Martha found a renewed faith that her disease could be managed. “Between Dr. Fritz and Dr. Medsger, I knew I was in the best hands. This truly has been a successful team approach to treatment,” she says. Dr. Medsger was able to increase the medication she was already taking and soon she was starting to feel better, thanks to the way Dr. Medsger considered both her disease and the way that it impacted her life.

Over the past ten years, Martha and Dave have spent a lot of time learning about a disease that was once so unfamiliar. They’ve educated

Medsger and the physicians like him who are advocating for getting the knowledge out there to referring physicians about the importance of early referrals for treatment, along with the breakthroughs that were made in Pittsburgh.”

Since receiving the scleroderma diagnosis a decade ago, Martha and Dave have tried to keep their lives as normal as possible. Martha’s biggest passion is still riding horses – which she has done since she was five years old, beginning competitive riding at just age 12. Together, they have a horse, two beagles – therapy dogs who they often take to hospitals to help lift patients’ spirits, one cat and one miniature donkey. Plus, they love spending time with their extended families – often having close to 30 people seated around their large Thanksgiving dinner table or at evening bonfires. And, each year

*“Every day that I have with Martha is a gift. And, that’s all thanks to Dr. Medsger and the physicians like him...” - Dave Merry*

After having physicians tell her that what she was experiencing was simply “in her head”, Martha saw rheumatologist Deborah Fritz, MD in Cincinnati, near her hometown of Lebanon, Ohio. It was then that she first heard the word ‘scleroderma’. “I was worried about my future and the progression of a disease that I knew very little about,” she says. Dr. Fritz started her on Penicillamine and helped her begin treating this disease. “When I asked Dr. Fritz what she would do if she were in my shoes, she remembered hearing someone who she considered an expert in the field at a continuing education conference – Dr. Thomas Medsger – and suggested I come to see him.”

Martha called Dr. Medsger’s office, only to find that there was a 3-4 month waiting period until the next available appointment. However, after she explained her accelerated disease progression to Dr. Medsger’s nurse Carol Blair, Carol helped Martha get an appointment within

themselves by reading about the latest research and therapeutic options. However, they’ve noticed that, compared to many other conditions, scleroderma receives very little when it comes to financial support. “Because it’s so unknown, there isn’t a lot of funding available for cutting-edge research or to help those who can’t afford care,” Martha says.

Throughout the years, Martha and Dave have decided to make philanthropic gifts in support of the work being done in Pittsburgh for scleroderma. “We just think it is so important to be a part of making a difference in the future of this disease,” Dave says. “Everyone can help. The size of the donation doesn’t matter – what does matter is that every little bit allows the doctors and researchers to gain momentum and keeps talented people in the pipeline doing important work. Every day that I have with Martha is a gift. And, that’s all thanks to Dr.

they take the time to visit St. Thomas – an annual tradition that began when Martha’s former boss at Mars Petcare suggested the vacation spot back in 2004 as a way to rejuvenate themselves after the initial upheaval of their lives caused by scleroderma. There, Martha and Dave still spend time snorkeling and enjoying time together.

“In the beginning, I believed that I had to fight my way through,” says Martha. “But, now I know the importance of **not fighting it**. I’ve learned to **live with it** and take it stride for stride.”



Martha riding rescue horse, Barney



**Pulmonary Fibrosis and Air Quality** by *Robyn Domsic, MD, MPH and Kristen Veraldi, MD, PhD*

Pulmonary fibrosis, or interstitial lung disease (ILD), is a significant complication of systemic sclerosis and the most common scleroderma-related cause of death. The risk of developing pulmonary fibrosis is variable and related to the subtype of scleroderma (diffuse vs. limited) and the scleroderma-related blood antibody a patient has. Symptoms include the gradual onset of shortness of breath accompanied by a dry cough. Because these symptoms are not specific for pulmonary fibrosis and are not present in mild or early disease, scleroderma patients considered by their rheumatologists to be at increased risk to develop ILD are screened on a regular basis with pulmonary function tests and high resolution computed tomography (CT) scans of the lungs.

Normal lungs contain millions of tiny air sacs called alveoli. Air that one breathes in containing oxygen reaches the alveoli and oxygen is transported into blood vessels

adjacent to the alveoli. At the same time, blood releases carbon dioxide into the alveoli, and the carbon dioxide is exhaled when one breathes out. The tissue between the alveoli and blood vessels is called the interstitium. It contains elastic fibers that allow the alveoli to stretch as they fill with air with each breath. In pulmonary fibrosis the interstitium becomes thickened and scarred due to excessive laydown of collagen, similar to thickening of skin. The alveoli are then not able to expand adequately and it is difficult for oxygen to cross into the bloodstream.

Air pollution results in reduced lung function in healthy adults and is recognized to trigger flareups of other lung diseases such as cystic fibrosis, asthma, and chronic obstructive pulmonary disease (COPD or emphysema). Although there is no direct evidence that air quality will worsen scleroderma-associated ILD, many patients report worse breathing symptoms when the air quality is poor. During the summer, air quality tends to be worse, and can vary from day to day.



As with all forms of lung disease, we recommend that ILD patients understand air quality warnings and be aware of their own personal activity limitations. ILD patients should minimize outdoor activities when the Air Quality Index (AQI) is at orange or above. Healthy persons should reduce outdoor activities when the AQI is at red or above. ILD patients should discuss any specific questions or concerns that they have about air pollution and their lung health with their physicians.

The US EPA has a very useful website to check daily air quality in your city. <http://airnow.gov>. The following information was taken from this website:

AIR QUALITY INDEX		
Levels of Health Concern	Numerical Value	Meaning
Good	0 to 50	Air Quality is considered satisfactory and air pollution poses little or no risk
Moderate	51 to 100	Air Quality is acceptable; however, for some pollutants there may be a moderate health concern for a very small number of people who are unusually sensitive to air pollution.
Unhealthy for Sensitive Groups	101 to 150	Members of sensitive groups, including lung diseases may experience health effects. The general public is not likely to be affected.
Unhealthy	151 to 200	Everyone may begin to experience health effects; members of sensitive groups may experience more serious health effects.
Very Unhealthy	201 to 300	Health alert; everyone may experience more serious health effects.
Hazardous	301 to 500	Health warnings of emergency conditions. The entire population is more likely to be affected.



## ASK THE EXPERT

By Kathryn S. Torok, MD

In pediatric scleroderma clinic I encounter the following questions:

### 1. Is scleroderma inherited?

Scleroderma, both systemic sclerosis (affecting the skin and internal organs) and localized scleroderma (affecting mostly the skin and fat/muscle underneath), are considered to be complex autoimmune diseases that occur in individuals who have a “genetic predisposition” and encounter one or more environmental exposures. When an exposure occurs, such as a virus causing the common cold, the immune system “over-reacts”. It responds appropriately to the environmental “trigger” but stays “on” rather than turning “off” or slowing down. The result is an immune attack against one’s own skin and organs. Though there is likely a genetic “predisposition” or genetic make-up that makes an individual more prone to these autoimmune diseases, scleroderma is not considered an inherited disease. To date, there have not been any genes identified as “causing” scleroderma. Rather multiple genes have been found to have slightly increased frequency in scleroderma compared with non-scleroderma patients. Thus it may be necessary for a patient to have a number of “predisposing” genes to be susceptible.

### 2. What is the chance of my other children developing scleroderma?

Very few patients with scleroderma have a relative with scleroderma, even if they are one of identical twins who have the exact same DNA. In a Pittsburgh Scleroderma Center study looking at identical twins, only 5% of the twin pairs both had scleroderma. Therefore, one’s genetic background is not sufficient to develop scleroderma. I have been asked by families if they should have their other children ‘screened’ for scleroderma. There is no screening tool available to do this and it is so uncommon for scleroderma to develop in other family members that this is not advised. Of course family members with Raynaud Phenomenon or other symptoms concerning for scleroderma should be evaluated by a rheumatologist.

### 3. What is the chance of my daughter’s children (my grandchildren) having scleroderma?

As in the case of siblings discussed above, it would be extremely uncommon to have a grandchild with scleroderma

if you have scleroderma.

### 4. Are there other autoimmune conditions that accompany scleroderma?

A few autoimmune diseases can occur with scleroderma in the same patient, such as autoimmune thyroid disease, rheumatoid arthritis and Sjogren syndrome. However, it is more common for there to be an autoimmune ‘clustering’ in the family. For example, occasionally an individual with scleroderma will have an uncle with psoriasis, an aunt with ulcerative colitis, and grandparent with rheumatoid arthritis.

In summary, although scleroderma is not directly inherited, some scientists feel there is a slight predisposition to it in families with a history of autoimmune rheumatic diseases. Very few scleroderma patients have a relative with scleroderma and their children extremely rarely develop scleroderma. Research indicates that a patient’s genetic background may make him or her more susceptible to developing scleroderma if and when exposed to certain environmental triggers. Ongoing research is focusing on what these triggers may be.

**Do you have a question regarding scleroderma that you would like answered by our Scleroderma Center team of experts?**



**Please send your question to  
Maureen Laffoon at  
[laffoonm@pitt.edu](mailto:laffoonm@pitt.edu).**

**If your question is selected, the question and answer will be published in a future issue of the newsletter.**

# Thank You

**Thank you to the the following recent donors for their support of scleroderma research**

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YOU  
SUMMERTIME  
FUN!



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