University of Pittsburgh & UPMC Scleroderma Center

COLLAGEN CONNECTION

Spring/Summer 2012

Interstitial Lung Disease in Systemic Sclerosis (Scleroderma)

By Kristen L. Veraldi, MD, PhD

Pulmonary (lung) involvement is common among individuals with Systemic Sclerosis (SSc; scleroderma) and negatively impacts both quality of life and life expectancy. SSc-associated pulmonary complications include interstitial lung disease, pulmonary hypertension, or a combination of both. This article will review the most common pulmonary complication of SSc and the current leading cause of death: Interstitial Lung Disease.

What is Interstitial Lung Disease?

Interstitial Lung Disease (ILD), often referred to as pulmonary fibrosis, is a group of disorders that includes over 100 separate lung diseases that share the common characteristics of scarring (also called fibrosis) and/or inflammation of the lungs. Fibrosis affects the lung's interstitium, which is the part of the lung that surrounds and supports the small blood vessels (capillaries) and air sacs (alveoli). This is where oxygen from the air we breathe enters the blood stream to supply our vital organs and tissues, and where carbon dioxide leaves the blood stream and is eliminated in our exhaled breath. Fibrosis of the interstitium makes it more difficult for oxygen to enter the blood stream, which is why people with ILD often need to use supplemental oxygen.

Dr. Kristen Veraldi is a graduate of the Medical Scientist Training Program at the University of Pittsburgh and Carnegie Mellon University, where she earned her MD and her PhD in Biochemistry and Molecular Genetics. She completed her internship and residency in Internal Medicine at the University of Michigan Health System in Ann Arbor, MI and her fellowship in Pulmonary Sciences and Critical Care Medicine at the University of Colorado Health Sciences Center and National Jewish Health in Denver, CO. After completing an additional post-doctoral research fellowship at the University of Pittsburgh, she joined the faculty and is currently an Assistant Professor of Medicine in the Division of Pulmonary, Allergy and Critical Care Medicine. Dr. Veraldi is board-certified in Internal Medicine, Pulmonary Disease, and Critical Care Medicine and is a Fellow of the American College of Chest Physicians. Dr. Veraldi provides care for individuals with interstitial lung diseases (ILD) at the UPMC Comprehensive Lung Center. In addition to her clinical activities, she serves as a member of the University of Pittsburgh Institutional Review Board and is actively involved in research projects focused on the molecular mechanisms of ILD, including characterizing how changes in the normal cellular stress response contribute to systemic sclerosis-associated ILD.

What are the Symptoms of Interstitial Lung Disease and how is it Diagnosed?

ILD can be present without any lung symptoms. The most common symptoms are slowly progressive shortness of breath with exercise and a dry cough. Unfortunately, these symptoms can occur for many other reasons and are not specific for ILD. Your rheumatologist may ask you to complete tests to screen for ILD and may refer you to a pulmonologist (lung doctor). The most common tests you will be asked to complete are pulmonary function tests (PFTs) and a high resolution chest computed tomography (CT) scan.

You may also be asked to complete an exercise test to see if your oxygen levels decrease during activity. This is usually done by having you walk on a treadmill while your oxygen

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level is monitored with a pulse oximeter placed on your earlobe or fingertip. An echocardiogram (ultrasound of the heart) may also be recommended to screen for other possible causes of shortness of breath, including pulmonary hypertension (high blood pressure in the lung). Depending upon the results of these tests, your pulmonologist may recommend additional tests such as a right heart catheterization or a surgical lung biopsy. The lung biopsy is often performed using a scope procedure that allows the surgeon to biopsy several areas of one lung through just a few small incisions in the chest.

What is the Natural History of Interstitial Lung Disease and How is it Treated?

The severity and progression of SSc-associated ILD is variable and challenging to predict for a given individual. However, the prognosis for the majority of patients is good, with published survival estimates of up to 85% at 5 years. Early studies performed at the University of Pittsburgh by Drs. Thomas Medsger and Virginia Steen demonstrated that those who experience severe progressive ILD tend to do so early in the course of disease, with the greatest decline in lung function usually occurring within the first 2-3 years. It should go without saying that it is vitally important to avoid smoking, which causes injury to your lungs regardless of whether or not you have ILD.

Some of the same tests used to diagnose ILD will also be repeated at intervals determined by your physicians to monitor the progression of lung disease. Currently available medication therapy is not thought to reverse scarring, but in some cases it may be possible to slow or prevent the development of further lung scarring by treating inflammation. Response to medication therapy varies widely and your physicians will weigh the potential benefits of treatment against the possible side effects of the various medication options. Even with treatment, some people will still experience progressive decline in their lung function. For some advanced cases of ILD that do not respond to medication therapy, referral for lung transplant evaluation at a center experienced with SSc may be an option. The University of Pittsburgh Lung Transplant Program has performed lung transplants on over 60 SSc patients with ILD, the largest experience world-wide.

Oxygen therapy is required for many people living with lung disease. There are many systems used to deliver oxygen that allow for an active lifestyle and travel while using oxygen. Some people may need oxygen all of the time, others may need to use it only with exercise or during sleep. All patients with ILD should have vaccinations to prevent pneumonia due to pneumococcus bacteria (Pneumovax) and an annual flu vaccine. If you have ILD and esophageal reflux symptoms, you should be on an acid-blocking drug to minimize the possibility of stomach acid “refluxing” into the lungs.

High resolution CT findings consistent with ILD are found in up to 90% of persons with SSc, but only a minority of them will experience severe, progressive lung disease.

Who is at Risk of Developing Interstitial Lung Disease?

ILD is common among patients with SSc. High resolution CT findings consistent with ILD are found in up to 90% of persons with SSc, but only a minority of them will experience severe, progressive lung disease. One challenge faced by physicians is trying to determine who will develop debilitating lung disease and who will experience only mild, slowly progressive, and often asymptomatic lung disease.

A number of factors have been described as being associated with an increased risk of developing pulmonary involvement, including antibodies such as anti-topoisomerase I (Scl-70) antibody and more extensive skin disease. Unfortunately, these characteristics are not reliable predictors. There is a critical need for continued research to discover better tests to predict the development, progression, and response to medication for SSc-associated lung disease.

An association between reduced esophageal motility and/or gastroesophageal reflux and ILD has been explored by a number of researchers, but the exact role for esophageal reflux in the development of SSc-ILD remains uncertain.

Your pulmonologist may recommend participation in a formal pulmonary rehabilitation program to help you to achieve your highest possible level of physical functioning. This is a comprehensive program that includes more than just supervised exercise conditioning and respiratory therapy evaluation; pulmonary rehabilitation also provides important education, nutritional counseling, breathing and energy saving techniques.

ILD and Lung Cancer

Many forms of ILD, including SSc-associated ILD, are associated with an increased risk of lung cancer, even in people who have never smoked cigarettes. Even if your ILD is mild, long-term followup is recommended with periodic CT scans done at intervals determined by your physicians.

Current Research on SSc-Associated ILD

For additional information on research for SSc-associated ILD, please visit our Center’s website and click on the “Research” tab.

http://www.dom.pitt.edu/rheum/sclero/scleroderma.html

JUNE IS SCLERODERMA AWARENESS MONTH

About the Authors

Linda McNamara, RN, MBA is a registered nurse, healthcare consultant, and certified health coach with over forty years experience in health and wellness. She has been living with systemic lupus since 1996.

Karen A. Kemper, PhD, MSPH is a health educator and university professor in health promotion and public health. She has certifications in health fitness and life coaching and has worked in health and wellness for twenty-five years. She has been living with scleroderma since 1992.

Book Review

by Mary L. Fike

A book given to my daughter by staff at The Scleroderma Center to help her understand those affected by lupus, scleroderma and related autoimmune diseases turned out to be a gift to me. The whimsical floral dress on the front cover was a colorful sign of hope. Having dealt with lupus for 38 years since a bleak diagnosis at the age of 16, the title and layout of the book fashioned a new approach toward self help for me. Was there something new to be learned about the "ugly dress"—my diagnosis—that I too had felt so uncomfortable in for years?

This is a well-written book about the authors' personal journeys with scleroderma and lupus. Being a nurse, I felt the book covered the clinical issues of autoimmune diseases in an understandable way. Thought-provoking and interspersed with reflective poetry, the book leaves the reader with a sense of peace, strength and direction.

The authors cite writer Flannery O'Connor, herself a lupus sufferer, who said that lupus is more instructive than a trip to Europe, but it is a trip you have to take entirely on your own. This observation is so true in spirit. We feel trapped inside a body unwilling to cooperate and with a condition that is unpredictable. But in reality, as the book outlines, there is so much we as patients and caregivers can do to "accessorize" the ugly dress that we must wear.

The book's first section was difficult to read amidst empathetic tears for so many of the emotions and struggles that the authors shared. The second section resurrects the spirit; it is an excellent resource of helpful management strategies and "accessories." It teaches us that we must take care of ourselves and exercise "healthy selfishness" in order to wear our "dresses" with style.

This is truly a "girls' club" book: experienced women share support, humor, tears and practical advice with their friends, their readers. For those of us who struggle each day with wearing the "ugly dress" or our illnesses, this book is indeed a gift.

PLEASE JOIN US

The Second Annual

Walk With Tori

Date: Sunday, September 9, 2012

Location: Doubs Woods Park, Hagerstown, Maryland

Time: Registration starts at 1 p.m. Walk begins at 3 p.m.

We are in need of donations for the silent auction.

For additional information, please email Tori at walkwithtori@gmail.com or call Joe Dill at 301-797-3566

Informational Reception

Sheldon Marstine, friend of the University of Pittsburgh and UPMC Scleroderma Center, graciously hosted an event at the Pittsburgh Golf Club on May 19, 2012. At this intimate gathering, approximately 50 people came together to learn more about the latest activities of the Scleroderma Center.

Those in attendance had a unique opportunity to learn about current research along with the Center's vision for the future through updates by Thomas Medsger, MD and Carol Feghali-Bostwick, PhD (Center Co-Directors); Robyn Domsic, MD, MPH; Kristen Veraldi, MD, PhD; Kathryn Torok, MD; and Christine Peoples, MD.

By showcasing the outstanding work that is currently being done "from bench to bedside" here in Pittsburgh, the speakers provided clear examples of why supporting scleroderma research, education and patient care is vital to strengthening the Center's overall goal of finding a cure for scleroderma.

For more information on ways in which you can support the Scleroderma Center, please contact Gary Dubin at (412) 647-9113 or at dgary@pmhsf.org.

Dr. Carol Feghali-Bostwick, Dr. Thomas Medsger, Sheldon Marstine and Annette Blum

Dr. Thomas Medsger

Dr. Carol Feghali-Bostwick and Advisory Group Member, Merdie Shoemaker

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Judi Feinberg and Dr. Basil Zitelli

Mary Lucas, Jessica Fike and Maureen Laffoon (Center Staff)

Annette Blum, Sheldon Marstine, Dr. Robyn Domsic and Anne Medsger

Elise Yoder, Yvonne Keairns, Dr. Carol Baker, Dr. Christine Peoples

Gregg and Cynthia Kimmy and Beenie and George Smith

The Bindernagel Family

Ask the Expert

Tonya V. from Cleveland, OH asks the following:

Question: I found Dr. Domsic's article about Raynauds very helpful. Can she offer advice on sun exposure and scleroderma?

Summer Skin "Scare"

by Robyn T. Domsic, MD, MPH

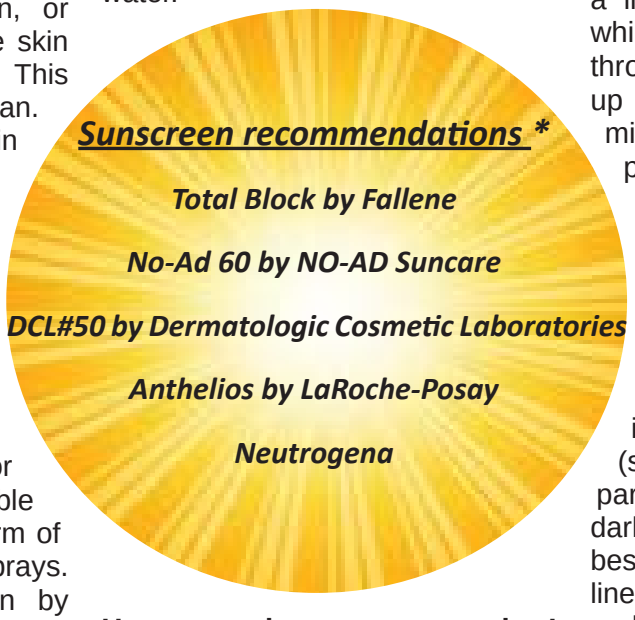
Scleroderma patients can experience hyperpigmentation, or skin darkening, as part of the skin changes in their disease. This can often be mistaken for a tan. However, scleroderma skin needs to be protected from sun exposure during the summer. Here are some tips for understanding and managing sun exposure in the upcoming summer months.

Sunscreen: There is a wide variety of sunscreens, or sun-blocking agents, available today. These come in the form of gels, creams, lotions and sprays. Sunscreen protects the skin by absorbing or reflecting ultraviolet (UV) rays. You should look for a broad-spectrum sunscreen, meaning that it protects against both UVA and UVB rays, and preferably water-resistant. New FDA rules require a sunscreen to state how long the water resistance lasts. Sunscreens that are not water resistant will not carry a warning.

In general, gels are more drying than lotions or cream. Patients with scleroderma tend to have dry skin, and thus lotions or creams may be a better choice. PABA-free may be more comfortable if sunscreen tends to irritate or sting the skin.

What is SPF? SPF stands for sun protection factor. This is a measure of the ability of the product (sunscreen or clothing) to prevent sunburn from UVB rays. However, both UVA and UVB light have been linked to skin cancer and aging of the skin.

What SPF is best? The American Academy of Dermatology recommends using a sunscreen with an SPF of 30 or above for everyone, however an SPF of 50 or above is preferred in patients with scleroderma or other autoimmune illness. You may need a higher SPF if you will be in the sun for a long time, or anticipate intense sun exposure, such as when on or near water.



How much sunscreen do I need? One ounce (approximately two tablespoons) of a lotion to cover an adult's arms, legs, necks and face. You may need more sunscreen to cover the chest and back. Applying less sunscreen will effectively reduce the sunscreen's SPF rating, or the amount of time it may take to become sunburned.

When do I apply sunscreen? Apply sunscreen 20-30 minutes before exposure. Exposed skin is any skin not protected from the sun. Sunscreen should be reapplied every 2-3 hours when you are dry. You should reapply sunscreen after sweating, drying off with a towel, or being in the water.

Does sunscreen expire? Many sunscreens list an expiration date, as chemical sunscreens may become less effective over time. The decrease in sun protection ability may be hastened by leaving them in high temperatures, such as in a car or sitting in a bag at the beach. If sunscreen does not have an expiration date, many dermatologists will recommend throwing it away after 3 years. Expired sunscreen may be less effective, essentially having a lower SPF rating.

What about other products for the face? Protect your lips with a lip balm of SPF 30 or higher, which also needs to be re-applied throughout the day. Some make-up products (liquid foundation or mineral powders) may contain sun-protective ingredients. However, many of these products provide little or no UVA protection.

What about clothing? Covering exposed skin, such as with a wide-brimmed hat and long-sleeved clothing or pants, is helpful. Tightly-woven material (such as canvas) is the best, particularly for hats. In clothing, darker, tightly-woven fabrics are the best. There are many manufactured lines of clothing with SPF available, and some can be found at national chains such as Target, Old Navy and LLBean. The more popular sun protection clothing manufacturers are 1) Sun Precautions 2) Coolibar and 3) Sunday Afternoons.

Minimize sun exposure: UVB sun exposure is the greatest in the summer months between 10:00 am and 4:00 pm, while UVA exposure is more constant throughout the day. Cloudy days still allow sufficient UVB light to pass through and cause sunburns. Thus, sunscreen and sun-protective measures should still be used, including hats and umbrellas.

Special Thanks to Dr. Lisa A. Pawelski, a Pittsburgh dermatologist who gave her recommendations on sunscreen, sun protection clothing and other useful tips.

Thank You

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Clinical Trials Update

The Scleroderma Center has recently started recruiting for two new investigational drug trials. The first trial is for scleroderma patients with active digital (fingertip) ulcers, and the second is for patients with diffuse skin disease and pulmonary fibrosis. For more information on either of these trials please contact our research coordinator, Dana Ivanco at des2@pitt.edu or phone number 412-648-7040.

The ongoing clinical trials can be found on our website:

http://www.dom.pitt.edu/rheum/sclero/scleroderma.html

HAVE A FUN SUMMER!

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