

Scleroderma-Associated Interstitial Lung Disease *Managing Symptoms, Living Well*

By Sally McLaughlin, M.S.

The majority of scleroderma patients have some lung involvement in the form of either pulmonary hypertension or interstitial lung disease (ILD).

Lung involvement in scleroderma ranks fourth behind manifestations of the skin, peripheral blood vessels and esophagus. With fewer problems due to kidney involvement, thanks to the use of ACE inhibitors, lung involvement now is the most important predictor in a scleroderma patient's survival.

In general, the more skin involvement, the more lung involvement. Lung involvement can appear before other signs of scleroderma.

A different lung problem – pulmonary hypertension – may be more prevalent in patients with limited scleroderma, but this disease will not be discussed here.

What is interstitial lung disease?

Interstitial lung disease is a broad group of lung disorders, characterized by inflammation and/or scarring of the interstitium, the tissue through which oxygen circulates (diffusion) from the air sac (alveolus) into the bloodstream. When this tissue is inflamed or scarred, such as happens with scleroderma, it becomes difficult for oxygen to diffuse into the bloodstream. Consequently, the body's tissues and cells don't get enough oxygen. Chronic inflammation can lead to irreversible scarring.

How is interstitial lung disease diagnosed?

- A history of a dry cough, shortness of breath at rest or with activity.
- Fine "Velcro"-sounding crackles in the lungs heard through a stethoscope.
- A full pulmonary function test (PFT) shows decreased diffusion capacity, such as how well oxygen diffuses across the interstitium into the bloodstream.
- A high resolution CT (HRCT) of the chest showing evidence of inflammation or fibrosis.
- A biopsy using video-assisted thoracic surgery, called a VATS biopsy, of the lung may be necessary.

Interstitial lung diseases are not always easy to diagnose. Studies have shown that the accuracy of an interstitial lung disease diagnosis increases when a multidisciplinary team of pulmonologists, radiologists, pathologists and rheumatologists who have expertise in interstitial lung disease work together to review a case.

How do I manage scleroderma-associated interstitial lung disease?

Inflammation of the interstitium makes it difficult for oxygen to diffuse into the bloodstream. Because chronic inflammation can lead to scarring (fibrosis), which is irreversible, it is vital that inflammation of the interstitium be treated.

Immunosuppressants such as prednisone, mycophenolate (CellCept®) or cyclophosphamide (Cytoxan®) work to calm the body's immune system down and reduce lung inflammation. By reducing inflammation, it is hoped that these drugs will prevent the formation of lung fibrosis, and allow the inflamed lung to return to normal.

Definition: Comorbidity

Two or more coexisting medical conditions or disease processes that are additional to an initial diagnosis.

-Mosby's Medical Dictionary, eighth edition, 2009, Elsevier.

To keep the lungs as healthy as possible, it is important that any problems associated with scleroderma-associated interstitial lung disease (that is, any *comorbidities*) be treated as well. These might include gastroesophageal reflux disease (GERD), pulmonary hypertension secondary to interstitial lung disease or sleep apnea.

Will I need a lung transplant if I have scleroderma-associated interstitial lung disease?

The key to managing scleroderma-associated interstitial lung disease is to get a baseline pulmonary function test and high resolution CT (HRCT) of the chest early in the course of the disease, and then to follow up regularly. By doing this, any changes can be addressed early, rather than letting the disease worsen to the point that lung transplantation becomes the only option.

Some lung transplant centers are unwilling to perform transplants in scleroderma patients due to potential risks such as aspiration, when a person's stomach contents are accidentally breathed into the lungs. However, preliminary data from a recent study at the University of California at San Francisco indicates that esophageal problems are not significantly associated with worse outcomes in lung transplantation. The study also

found that there is no significant difference, when comparing transplantation in people with scleroderma-related lung disease versus that in people with other interstitial lung diseases, in survival outcomes or rate of rejection.

Can there be complications and symptoms? How do I manage them?

In summary, research shows that the more strategies one uses to manage shortness of breath, the more control one will have over these symptoms, and the less shortness of breath one will experience. In order to keep the lungs as healthy as possible in scleroderma-associated interstitial lung disease, it is important to do the following:

- **Prevent the flu.** Get a flu shot every year. Scleroderma patients should receive the shot made from the killed virus, not the nasal spray, which contains a live virus.
- **Prevent bacterial pneumonia.** Get a pneumococcal pneumonia shot. Follow the most recent advice from the Centers for Disease Control (www.cdc.gov/vaccines) regarding how often to get this vaccination.
- **Manage dyspnea (shortness of breath).** Dyspnea can be the most distressing symptom of interstitial lung disease, and is associated with a poor quality of life. Research shows that the key to managing dyspnea is to develop a list of strategies, many of which can be learned in a pulmonary rehabilitation program.

Strategies to manage dyspnea can be divided into four categories:

1. **Decrease the sense of effort of breathing** through exercise, using oxygen during activity, anti-anxiety medications, relaxation, distraction, yoga, blowing a fan in the face. Breathing retraining to make breathing more efficient; energy conservation and activity modification strategies such as avoiding bending over by using a reacher; sitting on a plastic chair in the shower; wearing oxygen during showers; using a terry cloth bathrobe to dry rather than a towel; or wearing oxygen during meals.
2. **Reduce airway resistance.** Avoiding infections and irritants such as smoke, dust and toxic vapors.
3. **Maximizing inspiratory muscle function** to maintain a good body weight, and getting plenty of protein in the diet.
4. **Altering the brain's experience or perception of shortness of breath** by learning or knowing that it's OK to be short of breath, knowing strategies for dealing with shortness of breath, increasing confidence that shortness of breath can be controlled, etc.

- **Manage hypoxemia (low oxygen levels in the blood).** Hypoxemia in interstitial lung disease is caused by poor diffusion of oxygen across the interstitium into the bloodstream. One can be hypoxemic at rest, with activity, during sleep, at different altitudes or all the time. A pulse oximeter measures hypoxemia in terms of oxygen saturation (O₂ sat). Normal oxygen saturation is 98 to 100 percent at all times; when it falls below 89 percent; supplemental oxygen becomes necessary to keep the tissues and cells of the body healthy.

Hypoxemia and shortness of breath do not necessarily go hand in hand. One can be short of breath, but not have hypoxemia, and one may not experience shortness of breath, yet be hypoxemic. It's important to be tested for hypoxemia at rest and with activity, and sometimes, during sleep and at varying altitudes, to know if and when one is hypoxemic.

Hypoxemia can be treated with supplemental oxygen. Supplemental oxygen should be used when the oxygen saturation falls below 89 percent. The amount of supplemental oxygen needed is how much it takes to keep a person's oxygen saturation above 89 percent, or preferably, higher.

An important part of managing hypoxemia is choosing oxygen equipment that *can* and *will* be used, and that will enable a person to stay active. Today, there are many choices of oxygen systems to accommodate an individual's lifestyle, needs, preferences and strength. Newer portable oxygen concentrators (POCs) make travel with oxygen easier than ever before, because it is the only oxygen equipment that can be taken on-board an airplane. A respiratory therapist at an oxygen supplier is an excellent resource to help you determine what equipment you will need. Three good websites to learn more about supplemental oxygen equipment are:

- www.portableoxygen.org
- www.homeoxygen.org
- www.emphysema.net

A word about air travel and hypoxemia

Cabin pressure altitude on airplanes is approximately 8000 feet. This would be like sitting on an 8000-foot mountain peak for the duration of a flight, where the air contains only 15 to 17 percent oxygen, as opposed to 21 percent oxygen at sea level.

In someone without lung problems, oxygen saturations can drop to 85 to 91 percent. Therefore, someone who needs, or is close to needing, supplemental oxygen at sea level, will surely need it during a flight. If there is a question about the safety of flying, an altitude simulation test at a pulmonary function laboratory should be performed before a flight .

If supplemental oxygen is required for the flight, you must notify the airline in advance. The airline can supply you with oxygen for your flight, or you can rent a portable oxygen concentrator from an oxygen supply company to take on-board.

Are there any other things that I should monitor?

- ***Manage weight and nutrition.*** Good nutrition is crucial to maintain a healthy respiratory system. It can facilitate better breathing and help prevent infections. Here are some tips for maintaining good nutrition in people with lung disease:
 - Achieve and maintain a healthy body mass index (BMI).
 - Eat a diet that is high in protein, low in carbohydrates and sodium.
 - Eat small, frequent meals.
 - Eat larger meals earlier in the day.
 - Avoid foods that cause gas, bloating and fluid retention.
 - Avoid foods that increase stomach acid.

People with lung disease can gain weight due to decreased physical activity and use of prednisone. For those who need to lose weight, it's important to watch portion size, seek support and exercise regularly.

People with chronic lung disease can lose weight. You can burn 10 times more calories due to the work of breathing. You may need an extra 250 to 300 calories per day to maintain a good body weight. It is especially important that a person eat meals that are high in protein, calories and nutrients.

For more information about lung disease nutrition, visit www.ucsfhealth.org/ild.

- ***Manage cough.*** A dry, persistent cough can be one of the most distressing symptoms accompanying interstitial lung disease. Before attributing cough to interstitial lung disease, however, it is important to assess for, and treat, other common reasons for cough, such as asthma, GERD or post-nasal drip (now called upper airway cough syndrome.)

The exact reason for chronic cough in interstitial lung disease is unknown. Medications such as prednisone, benzonatate and codeine-containing cough

syrups sometimes are used. Distraction and relaxation techniques are also used to treat patients with persistent coughs.

- ***Manage deconditioning, decreased mobility and fatigue.*** Respiratory impairment leads to a cycle of: shortness of breath with activity, avoiding activity, deconditioning and more shortness of breath. This sometimes is called the Dyspnea Spiral.

Participating in a pulmonary rehabilitation program, that includes supervised exercise and learning dyspnea management, optimizing oxygen equipment, avoiding infections and more. Through a rehab program, you can significantly improve function, strength, endurance, mood, overall wellbeing, stress, independence and quality of life. Pulmonary rehabilitation programs are usually held twice a week for six to nine weeks. Many programs are covered by insurance.

Sally McLaughlin, M.S., is the advanced lung disease nurse working with the Interstitial Lung Disease Program at the University of California San Francisco Medical Center since 2003.