SCLERODERMA LUNG DISEASE: WHAT THE PATIENT SHOULD KNOW

Lung disease can be a serious complication of scleroderma. The two most common types of lung disease in patients with scleroderma are interstitial lung disease, also called pulmonary fibrosis, and pulmonary hypertension. These pulmonary complications can occur in patients with limited or diffuse scleroderma. Although these are the most common manifestations, other less common forms of pulmonary disease can also occur in scleroderma (see the table below).

Types of Scleroderma Lung Disease

- Interstitial Lung Disease
- Pulmonary Hypertension
- Airways obstruction
- Aspiration
- Extrinsic restriction
- Lung cancer
- Muscle weakness
- Pleural Effusions
- Pneumonia
- Pneumothorax
- Pulmonary Hemorrhage
- Sarcoidosis

ASSESSMENT OF SCLERODERMA LUNG DISEASE

It is important to note that patients may have significant pulmonary involvement from their scleroderma before signs and symptoms appear. **Therefore, it is important to have routine screening for possible pulmonary involvement, in particular interstitial lung disease and pulmonary hypertension.**

Symptoms

- **Chest pain**, particularly on exertion, is a potentially serious symptom and should be evaluated promptly by your physician. Chest pain may be a warning sign that you are at risk of having a heart attack. Chest pain can also occur in patients that have pulmonary hypertension, or it may be a symptom of gastroesophageal reflux (GERD).

- **Cough** can be seen in patients with interstitial lung disease, pulmonary hypertension or with airways disease. However, the most common cause of chronic cough is gastroesophageal reflux disease, which is very common in patients with scleroderma. It is important to note that many patients with GERD-related cough do not have typical symptoms of GERD such as indigestion or heartburn. Typically, in scleroderma patients the cough is non-productive.
• **Light headedness or fainting** (*syncope*) with exertion is also a very serious symptom that should prompt immediate evaluation by your physician. It may be a result of pulmonary hypertension or a consequence of too little oxygen secondary to severe interstitial lung disease.

• **Muscle weakness** can be seen in patients with scleroderma, particularly in those patients who have an overlap with polymyositis or dermatomyositis. Patients with muscle weakness of their arms and legs can also have muscle weakness of the respiratory muscles.

• **Palpitations** (heart racing or fluttering) may be a symptom of pulmonary hypertension or heart disease and should be evaluated promptly by your physician.

• **Shortness of breath**, also called **dyspnea**, is the most common symptom in patients with scleroderma lung disease and should be promptly evaluated by your physician. However, patients may have significant pulmonary involvement without this symptom. Often patients gradually decrease their activity to avoid the unpleasant sensation of shortness of breath without even realizing it. *It is important to screen for scleroderma lung disease even if you do not have shortness of breath.*

• **Swelling** is also referred to as **edema**. Lower extremity swelling is a potentially serious symptom and should be evaluated promptly by your physician. Swelling can occur in patients with pulmonary hypertension, very advanced interstitial lung disease and in patients with left-sided heart disease (congestive heart failure). Some medications may also cause leg edema.

• **Wheezing** is not common in scleroderma, but can be seen in patients with scleroderma-related airways disease, or in patients with a history of asthma or smoking. Airways disease is more common in scleroderma patients who also have features of rheumatoid arthritis or Sjogren’s syndrome. Wheezing can also be a symptom of gastroesophageal reflux disease.

**Tests Your Doctor Might Order**

**Pulmonary Function Tests**
Pulmonary function tests (PFTs) are performed by blowing in a tube that is connected to a computerized machine.

It is important that you form a tight seal with the mouthpiece. Often patients with scleroderma need to use a mouthpiece made for children because they cannot open their mouth very wide.

Numerous measurements can be made during pulmonary function testing and can be suggestive of a specific diagnosis.
6 minute walk testing (6MWT) is simple and reproducible and correlates with findings on more formal exercise testing. It has been also shown to correlate with hemodynamics measured during heart catheterization, functional classification, prognosis and even survival. The 6MWT is routinely used in studies of pulmonary hypertension. During this test, you are asked to walk as far as you can during six minutes. You are allowed to rest if you need to. While doing this test, your heart rate, blood pressure, oxygen level, degree of shortness of breath and other symptoms are monitored. Ask your doctor to use a forehead probe to monitor your oxygen level as it is frequently difficult to detect oxygen levels with a finger probe in patients with Raynaud phenomenon.

Table: Recommendations for Pulmonary Function and Exercise Testing

*There are no official recommendations for pulmonary function and exercise testing in patients with scleroderma. However, many experts recommend the following:

- **Baseline** complete set of pulmonary function tests (spirometry, lung volumes, diffusion)
- **≤ 5 years of Scleroderma**
  - PFTs and 6MWT every 3-6 months
- **> 5 years of Scleroderma**
  - PFTs and 6MWT every 6-12 months

Chest radiography (Chest x-ray) is not a sensitive test for scleroderma lung disease. This means that you could have significant lung disease and still have a normal chest x-ray. However, there are features on the chest x-ray that should prompt your physician to do further testing.

Chest Computed Tomography Scan (CT) is a special type of x-ray that allows one to see more details of the lungs. During a CT, you lie on your back and slide through a machine that looks like a giant doughnut. You may also be asked to also lie on your stomach (prone). Images are typically taken while holding your breath in inspiration. However, images may also be taken during expiration to see if air is trapped in your lungs. Finally, you may be given contrast dye in order to get a better look at the blood vessels in your lungs and to look for evidence of blood clots. A high resolution chest CT (HRCT) (or thin-slice CT) is considered the gold standard for evaluation for interstitial lung disease. Findings that are suggestive of interstitial lung disease include ground glass opacities (also called “alveolitis”), which looks like a haziness over the lung; septal thickening, which are extra lines in the lung.
because of thickening of the interstitium; and honeycombing, which represents holes in the lung. Ground glass opacities are suggestive of inflammation in the lungs that may be reversible, whereas honeycombing usually represents irreversible fibrosis.

**Bronchoalveolar lavage (BAL)** by bronchoscopy is sometimes done to look for inflammation or evidence of infection in the lung. A fiberoptic scope is passed through your nose or mouth down into your airways. Small amounts of sterile saline are then injected in an area of your lung and suctioned back out and sent for laboratory analysis. Although bronchoscopy and BAL to look for infection is a routine procedure, the analysis of the fluid for the presence of inflammatory cells (alveolitis) needs to be done by experienced personnel, usually at a scleroderma or pulmonary referral center.

**Echocardiogram** is actually an ultrasound of your heart. By rubbing a probe across your chest, the cardiologist and can look to see how well your heart is pumping, see if your heart valves are working, evaluate the size of the various chambers of the heart, see if there is fluid around the heart (pericardial effusion) and estimate your pulmonary artery pressure.

*There are no official recommendations for echocardiogram testing in patients with scleroderma. However, many experts recommend the following:

- Baseline and yearly screening echocardiogram
- Patients with pulmonary hypertension
  - Echocardiogram every 3-6 months
  - Echocardiogram if deterioration

**Heart catheterization**

- **Right heart catheterization** is essential for the evaluation of pulmonary hypertension. The cardiologist will thread a wire through either your femoral vein in your groin or internal jugular vein in your neck to the right side of your heart and into your pulmonary artery in order to measure the pressure on the right side of your heart and in your lungs. The right heart catheter can also be used to measure your cardiac output (how well your heart is pumping) and can estimate the pressures on the left side of your heart. *A right heart catheterization needs to be done by an experienced cardiologist or pulmonologist before starting medicine for pulmonary hypertension.*

- **Left heart catheterization** is usually performed by threading a wire through your femoral artery in your groin. The cardiologist will do a left heart catheterization if he/she is looking for blockages of your coronary arteries, or needs to measure pressures on the left side of the heart in conjunction with pressures on the right
side of the heart. A left sided heart catheterization is also sometimes needed in patients with heart valve problems.

**Ventilation/Perfusion Scan (V/Q scan)** is a special nuclear x-ray to see if areas of your lung that get air also are getting blood. A mismatch or defect suggests a blood clot. This test should be considered in patients with pulmonary hypertension because if blood clots in the lungs are identified, they can sometimes be surgically removed and the pulmonary hypertension cured.

**Lung Biopsy** is rarely indicated in patients with scleroderma and is especially risky if you have pulmonary hypertension.

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**Due to the complexity of the diagnosis and treatment of scleroderma lung disease strong consideration should be given to referral of patients to physicians with expertise in scleroderma, interstitial lung disease and PAH. This requires close collaboration between you, your rheumatologist, pulmonologist and cardiologist.**

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**Treatment of Scleroderma Lung Disease** – This is very much dependent on the type of lung disease as determined by the outcome of the studies described above. For example, interstitial lung disease might be treated with immunosuppressive drugs, e.g. cyclophosphamide, azathioprine, or mycophenolate mofetil, depending upon the stage and degree of activity of the pulmonary fibrosis. Pulmonary hypertension requires different treatments, and there are a number of oral, inhaled and intravenous medications now available to treat this complication of scleroderma. Other pulmonary complications might require different treatment approaches. Adjunctive therapy is appropriate for scleroderma patients regardless of the particular type of lung disease. Adjunctive treatment might include one or more of the following: (1) avoid tobacco exposure; (2) take steps to minimize acid reflux; (3) exercise appropriately; and (4) use supplemental oxygen when prescribed by your physician.

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