



Frequently Asked Questions About Scleroderma

Scleroderma, or systemic sclerosis, is a chronic, connective tissue disease generally classified as an autoimmune rheumatic disease. The disease stems from the over-production of collagen (a connective tissue) in the body that creates hardening (sclerosis) of the skin (derma) and internal organs such as the lungs, kidneys, heart and gastrointestinal tract.

The word “scleroderma” comes from two Greek words, “sclero” meaning hard and “derma” meaning skin. Skin hardening is one of the most visible manifestations of the disease. The disease can take several forms and varies from person-to-person.

Here are some of the most frequently asked questions about scleroderma to help you better understand this disease:

Q: How serious is scleroderma?

A: Any chronic disease can be serious. The symptoms of scleroderma vary greatly for each person, and the effects can range from mild to life threatening. The severity depends on which parts of the body are affected, and to what extent in which they are affected. A mild case can become more serious if not properly treated. Early and proper diagnosis and treatment by qualified doctors may minimize scleroderma symptoms and prevent irreversible damage.

Q: How is scleroderma diagnosed?

A: The diagnostic process may require a visit with a rheumatologist (someone who specializes in conditions such as arthritis) or a dermatologist (a doctor who specializes in the skin), and a blood study or other specialized tests depending on which organs are affected.

Q: What causes scleroderma?

A: The exact cause or causes of scleroderma are unknown. Scientists and researchers are working hard to find the cause. It is known that scleroderma involves overproduction of collagen.

Q: Who develops scleroderma, and when?

A: An estimated 80,000 to 100,000 in the U.S. have the systemic form of scleroderma with many more people diagnosed with the localized form of the disease. The international incident rate is unknown. However, cases have been reported around the world. Since scleroderma presents with symptoms similar to other autoimmune diseases, diagnosis is difficult, and there may be many misdiagnosed or undiagnosed cases.

Localized scleroderma is often seen more in children. Systemic scleroderma is more common in adults. Overall, female patients outnumber male patients about 4-to-1.

While scleroderma can develop in every age group, the onset is most often between the ages of 25 and 55. Symptoms, onset age and other factors vary for each patient. There are many exceptions to the rules in scleroderma, perhaps more so than any other disease. Each case is different, and information should be discussed with your own doctor.

Q: What is the treatment for scleroderma?

A: Currently, there is no cure for scleroderma, but there are treatments available. Some are directed at particular symptoms such as heartburn, which can be controlled by medications called proton pump inhibitors or medications that improve the motions of the bowel. Other treatments are directed at decreasing activity in the immune system. Some people with a mild form of the disease may not need any

medication, and occasionally, people can go off treatment when their scleroderma is not active. Because there is so much variation from one person to the next, treatments vary for each patient.

Q: Are people living with scleroderma at risk for any specific lung involvement?

A: Patients with scleroderma are at increased risk for developing pulmonary hypertension (PH), also known as high blood pressure in the blood vessels of the lungs. This reduces the blood oxygen level, which in turn, may cause a reflex increase in blood pressure in the pulmonary arteries.

If the high blood pressure in the lungs is due to narrowing of the pulmonary arteries, it is called pulmonary arterial hypertension (PAH). When the blood pressure inside the pulmonary vessels is high, the right side of the heart has to pump harder to move blood into the lungs to pick up oxygen. This can lead to failure of the right side of the heart.

Q: What are the symptoms of PH?

A: Patients with mild PH may have no symptoms. Patients with moderate or severe PH usually notice shortness of breath (dyspnea), especially with exercise. Patients may also notice unusual chest pains and symptoms of right-sided heart failure, such as worsening shortness of breath and swelling of the feet and legs. Other symptoms that patients may experience include cough, light-headedness or fainting, palpitations (heart racing or fluttering) and swelling.

Q: What tests are done to diagnose PH?

A: The test most commonly used to diagnose PH is the echocardiogram. It can estimate the pulmonary artery pressure fairly well in most patients in a noninvasive manner. The physician may order a cardiac catheterization to measure the actual pressure in the pulmonary arteries. This invasive test is done to more accurately measure the pressures in the lung blood vessels and to assess the blood flow generated by the heart, known as the cardiac output.

Q: Are there any other lung issues that I should worry about?

A: Pulmonary hypertension is not the only type of lung disease that can occur with scleroderma. Interstitial lung disease (ILD), also called pulmonary fibrosis, is another serious complication.

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

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 PH. This requires close collaboration between you and your health care team.

For more information about scleroderma, visit www.scleroderma.org or call (800) 722-HOPE (4673).