

Scleroderma (Systemic Sclerosis)

Scleroderma (also called systemic sclerosis) is a rare disease characterized most often by skin thickening. Scleroderma also involves internal organs, and often leads to intestinal, kidney, lung, and heart problems. It is a chronic disease, which means it lasts a long time.

When scleroderma only involves the skin, it is called “localized” or “linear” scleroderma or “morphea”. When there is internal organ involvement, it is called “generalized” scleroderma or “systemic sclerosis”. Early diagnosis and treatment are essential. This may help prevent many of the complications of scleroderma.



What are Some of the Symptoms of Scleroderma?

Scleroderma may have many symptoms. Common skin symptoms include:

- Thickening, hardening, and tightness,
- Swelling of hands and toes,
- Hair loss in involved skin,
- Sores over fingertips,
- Generalized itching and
- Color changes of hands and feet with cold exposure (known as Raynaud's).
- Other common symptoms include:
 - Fatigue,
 - Arthritis,
 - Muscle pain and weakness,
 - Dryness of the eyes and mouth,
 - Heartburn, bloating, and other digestive symptoms,
 - Shortness of breath,
 - Cough and
 - High blood pressure (known as hypertension).

Who Gets Scleroderma?

Scleroderma is a rare disease. Approximately 10-20 new cases are diagnosed per million people each year. People of all races and ethnic backgrounds get scleroderma. Approximately 75% of people with the disease are women. It can occur at any age. However, scleroderma most commonly begins when people are between the ages of 30- and 60-years-old.

What Causes Scleroderma?

The cause of scleroderma is not known. We do know that scleroderma is an autoimmune disease. This means that the body's natural immune system does not behave normally. Instead of serving to fight off infections from bacteria, viruses and the like, the immune system of a person with scleroderma attacks its own body. This damages blood vessels.

The body's response to damage to its blood vessels is to make specific proteins (called collagen). The collagen repairs the damage by forming diffuse scars (called fibrosis) throughout the blood vessels. The excess collagen can deposit in the skin. This causes the many skin changes seen with scleroderma. In addition, the fibrosis of the blood vessels leads to derangements of many of the internal organs.

How is Scleroderma Diagnosed?

Often times it is difficult to diagnose scleroderma. Most often, a specialist in autoimmune diseases such as scleroderma, (known as a rheumatologist), is required to establish the diagnosis. The diagnosis of scleroderma is made based on the careful analysis of many factors.

A thorough history and physical examination are essential for the diagnosis. There are certain laboratory studies that can be helpful when considering the diagnosis. In addition to specific blood and urine tests, specialized lung and heart evaluations are needed to evaluate for any internal organ involvement from scleroderma.

It is important to know that the diagnosis cannot be made based on any specific blood test alone.

How is Scleroderma Managed?

Early recognition of scleroderma is essential. This will allow for early treatment for people with scleroderma. It is important to recognize that there is no cure for scleroderma. In addition, because it is a chronic disease, people often require medical therapy for many years to keep scleroderma under good control.

Goals of therapy vary for each person. The various organs involved in a given person with scleroderma guide treatment. There are a number of effective organ-specific treatments available for people with scleroderma. Combinations of anti-inflammatory and immunosuppressing medicines are often needed. These manage the underlying problems with the immune system. Various other medicines are often needed to control the skin, and internal organ problems seen in scleroderma. In particular, chemotherapy may be required to control the underlying lung problems seen with scleroderma.

In addition to medication therapy for scleroderma, many people benefit from physical therapy and

rehabilitation. Under the guidance of rehabilitation therapists, people with scleroderma often learn how to appropriately rest, exercise, strengthen and maintain joint and muscle function.

What is the Role of National Jewish Health?

National Jewish Health is one of the world's leaders in the study and management of immune diseases, such as scleroderma. National Jewish Health also specializes in Interstitial Lung Disease (ILD), a common lung abnormality of people with scleroderma. The National Institutes of Health has designated and funded National Jewish Health as a Specialized Center of Research for ILD.

Our health care providers have vast experience in treating people with scleroderma. We provide the expertise needed for the comprehensive evaluation and management of people with scleroderma. We aim to design an individualized treatment plan best suited for each person with scleroderma. In addition, in order to provide for comprehensive care of our patients with scleroderma National Jewish Health also provides physical, occupational, and recreational rehabilitation services in our rehabilitation department.

Visit our website for more information about support groups, clinical trials and lifestyle information.

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