[Scleroderma](http://www.mayoclinic.org/diseases-conditions/scleroderma/home/ovc-20206014)

Overview

[By Mayo Clinic Staff](http://www.mayoclinic.org/about-this-site/welcome)

Scleroderma (skleer-oh-DUR-muh) is a group of rare diseases that involve the hardening and tightening of the skin and connective tissues — the fibers that provide the framework and support for your body.

In some people, scleroderma affects only the skin. But in many people, scleroderma also harms structures beyond the skin — such as blood vessels, internal organs and the digestive tract. Signs and symptoms vary, depending on which structures are affected.

Scleroderma affects women more often than men and most commonly occurs between the ages of 30 and 50. While there is no cure for scleroderma, a variety of treatments can ease symptoms and improve quality of life.

**Symptoms**

Scleroderma's signs and symptoms vary, depending on which parts of your body are involved:

* **Skin.** Nearly everyone who has scleroderma experiences a hardening and tightening of patches of skin. These patches may be shaped like ovals or straight lines, or cover wide areas of the trunk and limbs. The number, location and size of the patches vary by type of scleroderma. Skin can appear shiny because it's so tight, and movement of the affected area may be restricted.
* **Fingers or toes.** One of the earliest signs of scleroderma is an exaggerated response to cold temperatures or emotional distress, which can cause numbness, pain or color changes in the fingers or toes. Called Raynaud's disease, this condition also occurs in people who don't have scleroderma.
* **Digestive system.** In addition to acid reflux, which can damage the section of esophagus nearest the stomach, some people with scleroderma may also have problems absorbing nutrients if their intestinal muscles aren't moving food properly through the intestines.
* **Heart, lungs or kidneys.** Scleroderma can affect the function of the heart, lungs or kidneys to varying degrees. These problems, if left untreated, can become life-threatening.

**Causes**

Scleroderma results from an overproduction and accumulation of collagen in body tissues. Collagen is a fibrous type of protein that makes up your body's connective tissues, including your skin.

Doctors aren't certain what prompts this abnormal collagen production, but the body's immune system appears to play a role. In some genetically susceptible people, symptoms may be triggered by exposure to certain types of pesticides, epoxy resins or solvents.

**Risk factors**

Scleroderma occurs much more often in women than it does in men. Choctaw Native Americans and African-Americans are more likely than Americans of European descent to develop the type of scleroderma that affects internal organs.

**Complications**

Scleroderma complications range from mild to severe and can affect your:

* **Fingertips.** The variety of Raynaud's disease that occurs with scleroderma can be so severe that the restricted blood flow permanently damages the tissue at the fingertips, causing pits or skin sores (ulcers). In some cases, gangrene and amputation may follow.
* **Lungs.** Scarring of lung tissue (pulmonary fibrosis) can result in reduced lung function, reduced ability to breathe and reduced tolerance for exercise. You may also develop high blood pressure in the arteries to your lungs (pulmonary hypertension).
* **Kidneys.** When scleroderma affects your kidneys, you can develop elevated blood pressure and an increased level of protein in your urine. More-serious effects of kidney complications may include renal crisis, which involves a sudden increase in blood pressure and rapid kidney failure.
* **Heart.** Scarring of heart tissue increases your risk of abnormal heartbeats (arrhythmias) and congestive heart failure, and can cause inflammation of the membranous sac surrounding your heart (pericarditis). Scleroderma can also raise the pressure on the right side of your heart and cause it to wear out.
* **Teeth.** Severe tightening of facial skin can cause your mouth to become smaller and narrower, which may make it hard to brush your teeth or to even have them professionally cleaned. People who have scleroderma often don't produce normal amounts of saliva, so the risk of dental decay increases even more.
* **Digestive system.** Digestive problems associated with scleroderma can lead to acid reflux and difficulty swallowing — some describe feeling as if food gets stuck midway down the esophagus — as well as bouts of constipation alternating with episodes of diarrhea.
* **Sexual function.** Men who have scleroderma often experience erectile dysfunction. Scleroderma may also affect the sexual function of women, by decreasing sexual lubrication and constricting the vaginal opening.
* Diagnosis
* Because scleroderma can take so many forms and affect so many different areas of the body, it can be difficult to diagnose.
* After a thorough physical exam, your doctor may suggest blood tests to check for elevated blood levels of certain antibodies produced by the immune system. He or she may remove a small tissue sample (biopsy) of your affected skin so that it can be examined in the laboratory for abnormalities.
* Your doctor may also suggest breathing tests (pulmonary function tests), a CT scan of your lungs and an echocardiogram of your heart.
* In addition, nearly all patients with scleroderma have blood tests that suggest autoimmunity and **antinuclear antibodies (ANAs) are usually detectable**. Another particular antibody, the **anticentromere antibody**, is found almost exclusively in the limited, or CREST, form of systemic sclerosis. **Anti-Scl 70 antibody (antitopoisomerase I antibody)** is most often seen in patients with the diffuse form of systemic sclerosis.
* Other tests are used to evaluate the presence or extent of any internal disease. These may include upper and lower gastrointestinal tests to evaluate the stomach and bowels, chest X-rays, lung function testing (pulmonary function test), and CAT scanning to examine the lungs, [EKG](http://www.medicinenet.com/electrocardiogram_ecg_or_ekg/article.htm) and echocardiograms, and sometimes heart catheterization to evaluate the pressure in the arteries of the heart and lungs for pulmonary [hypertension](http://www.medicinenet.com/high_blood_pressure_hypertension/article.htm).

Treatment

In some cases, the skin problems associated with scleroderma fade away on their own in three to five years. The type of scleroderma that affects internal organs usually worsens with time.

**Medications**

No drug has been developed that can stop the underlying process of scleroderma — the overproduction of collagen. But a variety of medications can help control scleroderma symptoms or help prevent complications. To accomplish this, these drugs may:

* **Dilate blood vessels.** Blood pressure medications that dilate blood vessels may help prevent lung and kidney problems and may help treat Raynaud's disease.
* **Suppress the immune system.** Drugs that suppress the immune system, such as those taken after organ transplants, may help reduce scleroderma symptoms.
* **Reduce stomach acid.** Medications such as omeprazole (Prilosec) can relieve symptoms of acid reflux.
* **Prevent infections.** Antibiotic ointment, cleaning and protection from the cold may help prevent infection of fingertip ulcers caused by Raynaud's disease. Regular influenza and pneumonia vaccinations can help protect lungs that have been damaged by scleroderma.
* **Relieve pain.** If over-the-counter pain relievers don't help enough, you can ask your doctor to prescribe stronger medications.

**Therapy**

Physical or occupational therapists can help you to:

* Manage pain
* Improve your strength and mobility
* Maintain independence with daily tasks

**Surgery**

Used as a last resort, surgical options for scleroderma complications may include:

* **Amputation.** If finger ulcers caused by severe Raynaud's disease have developed gangrene, amputation may be necessary.
* **Lung transplants.** People who have developed high blood pressure in the arteries to their lungs (pulmonary hypertension) may be candidates for lung transplants.