

# Scleroderma

### Overview

Scleroderma (sklair-oh-DUR-muh), also known as systemic sclerosis, is a group of rare diseases that involve the hardening and tightening of the skin. Scleroderma also may cause problems in the blood vessels, internal organs and digestive tract.

Scleroderma is often categorized as limited or diffuse, which refers only to the degree of skin involvement. Both types can involve any of the other vascular or organ symptoms that are part of the disease. Localized scleroderma, also known as morphea, affects only the skin.

While there is no cure for scleroderma, treatments can ease symptoms, slow progression and improve quality of life.

## **Symptoms**

Scleroderma symptoms vary from person to person, depending on which parts of the body are affected.

### Skin-related symptoms

Nearly everyone who has scleroderma experiences hardening and tightening of the skin.

The first parts of the body to be affected are usually the fingers, hands, feet and face. In some people, the skin thickening also can involve the forearms, upper arms, chest, abdomen, lower legs and thighs. Early symptoms may include swelling and itchiness. The color of affected skin can become lighter or darker, and skin may look shiny because of the tightness.

Some people also have small red spots, called telangiectasia, on their hands and face. Calcium deposits can form under the skin, particularly at the fingertips, causing bumps that can be seen on X-rays.

### Raynaud's phenomenon

Raynaud's phenomenon is common in scleroderma. It happens because of an exaggerated contraction of the small blood vessels in the fingers and toes in response to cold temperatures or emotional distress. When this happens, the digits may feel painful or numb and turn white, blue, gray or red. Raynaud's phenomenon also can occur in people who don't have scleroderma.

#### **Digestive symptoms**

Scleroderma can affect any part of the digestive system, from the esophagus to the rectum. Depending on which parts of the digestive system are affected, symptoms may include:

- · Heartburn.
- · Difficulty swallowing.
- Bloating.
- Diarrhea.
- · Constipation.
- · Fecal incontinence.

### Heart- and lung-related symptoms

When scleroderma affects the heart or lungs, it can cause shortness of breath, decreased exercise tolerance and dizziness. Scleroderma can cause scarring in the lung tissues that may result in increasing shortness of breath over time. There are medicines that may help slow the progression of this lung damage.

Scleroderma also can cause the blood pressure to increase in the circulation that goes between the heart and the lungs. This is called pulmonary hypertension. In addition to causing shortness of breath, pulmonary hypertension also can cause excess fluid to build up in the legs, feet and sometimes around the heart.

When scleroderma affects the heart, heartbeats can become irregular. Heart failure also may happen in some people.

### Causes

Scleroderma happens when the body produces too much collagen and it builds up in body tissues. Collagen is a fibrous type of protein that makes up the body's connective tissues, including the skin.

Experts don't know exactly what causes this process to begin, but the body's immune system appears to play a role. Most likely, scleroderma is caused by a combination of factors, including immune system problems, genetics and environmental triggers.

### Risk factors

Anyone can get scleroderma, but it is more common in people assigned female at birth. People typically get scleroderma between ages 30 and 50. Black people often have earlier onset and are more likely to have more skin involvement and lung disease.

Several other combined factors appear to influence the risk of having scleroderma:

- **Genetics.** People who have certain gene changes appear to be more likely to develop scleroderma. This may explain why scleroderma appears to run in families in a small number of people and why some types of scleroderma are more common for people in certain racial and ethnic groups.
- Environmental triggers. Research suggests that in some people, scleroderma symptoms may be triggered by exposure to certain viruses, medicines or drugs. Repeated exposure, such as at work, to certain harmful substances or chemicals also may increase the risk of scleroderma. An environmental trigger is not identified for most people.
- Immune system conditions. Scleroderma is believed to be an autoimmune disease. This means that it occurs in part because the body's immune system begins to attack the connective tissues. People who have scleroderma also may have symptoms of another autoimmune disease such as rheumatoid arthritis, lupus or Sjogren syndrome.

# **Complications**

Scleroderma complications range from mild to serious and can affect the:

- Fingertips. In systemic sclerosis, Raynaud's phenomenon can become so severe that the restricted blood flow permanently damages the tissue at the fingertips, causing pits or skin sores. In some people, the tissue on the fingertips may die.
- Lungs. Scarring of lung tissue can impact the ability to breathe and tolerance for exercise. High blood pressure in the arteries to the lungs also may happen.
- **Kidneys.** A serious kidney complication, called scleroderma renal crisis, involves a sudden increase in blood pressure and rapid kidney failure. Prompt treatment of this condition is important to preserve kidney function.
- Heart. Scarring of heart tissue increases the risk of irregular heartbeats and heart failure. Scleroderma also can cause inflammation of the sac surrounding the heart.
- Teeth. Serious tightening of facial skin can cause the mouth to become smaller and narrower. This may make it hard to brush the teeth or to have them professionally cleaned or restored. People who have scleroderma often don't make typical amounts of saliva, so the risk of dental decay increases even more
- **Digestive system.** Digestive complications of scleroderma can include heartburn and difficulty swallowing. Scleroderma also can cause bouts of cramps, bloating, constipation or diarrhea. Some people who have scleroderma also may have problems absorbing nutrients due to overgrowth of bacteria in the intestine.
- Joints. The skin over joints can become so tight that it restricts flexibility and movement, particularly in the hands.

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