



www.arthritis.org
800-283-7800

Scleroderma

WHAT IS SCLERODERMA?

Scleroderma (skler-o-DER-muh) is a condition that causes the skin to get tight and thicken. It affects women more often than men and it usually begins between the ages of 30 and 50. There are two types of scleroderma: localized and systemic.

Localized Scleroderma

Localized scleroderma mainly affects the skin. It also can affect muscles and bone, but does not affect internal organs. There are two main types of localized scleroderma: morphea and linear.

In morphea scleroderma, hard, oval-shaped patches form on the skin. The patches usually are whitish and sometimes have a purplish ring around them. The patches can form anywhere on the body surface. Over time, this type of scleroderma often improves on its own.

In linear scleroderma, lines or streaks of thickened skin form on the body, such as on the arms, legs or head. These lines can form in more than one area and can cause deep scarring, affecting the bones and muscles beneath the involved skin. These changes in skin, muscle and bone can affect the motion of joints.

Systemic Scleroderma

Systemic scleroderma, also called sclerosis, affects many parts of the body, such as the skin, blood vessels, digestive system (esophagus, stomach and bowel), heart, lungs, kidneys, muscles and joints.

In rare cases, systemic scleroderma may involve only internal organs and not the skin.

Systemic scleroderma is classified into two main groups by the degree of skin involvement: limited and diffuse.

Limited scleroderma has less extensive skin thickening. Raynaud's phenomenon and puffy fingers appearing several years before other signs or symptoms, and signs of internal organ involvement tend to appear much later. Limited scleroderma usually only affects the skin on the face, fingers and hands. Trouble with swallowing can occur because of loss of movement of the esophagus. Limited scleroderma may also seriously affect internal organs, such as the lungs or intestines.

One form of limited scleroderma is called the CREST syndrome. The CREST syndrome is associated with calcinosis, Raynaud's phe-

nomenon, esophageal dysfunction, sclerodactyly and telangiectasia. Each of these conditions is explained on the following pages.

Diffuse scleroderma generally develops more suddenly, with more widespread skin thickening. It may affect the skin on the hands, arms, thighs, chest, abdomen and face. Other body parts, such as the blood vessels, joints, muscles, esophagus, intestines and lungs, also can be affected. Kidney problems may lead to high blood pressure, and if left untreated, kidney failure.

WHAT CAUSES SCLERODERMA?

The exact cause of scleroderma is unknown. Evidence supports the notion that scleroderma is an autoimmune disease because abnormalities of the immune system, particularly antinuclear antibodies (ANAs), are found in most people with scleroderma. Scientists also know that too much collagen is produced in people with scleroderma. Collagen is a protein found in skin, bone, cartilage and ligaments. Extra collagen is deposited in the skin and internal organs, causing the skin to thicken and harden and affecting the function of internal organs. Scleroderma is also associated with damaged blood vessels in the skin and other involved organs.

WHAT ARE THE SYMPTOMS?

Scleroderma affects everyone differently. You may have some of the following symptoms:

Raynaud's Phenomenon

Raynaud's (ray-NODES) phenomenon occurs when blood flow decreases in response to cold temperatures or emotional stress. Raynaud's phenomenon most commonly occurs in the fingers, but also may develop in the toes, ears or tip of the nose. In Raynaud's phenomenon the blood vessels become narrowed in response to cold, which

results in decreased blood flow to the skin. If you have Raynaud's phenomenon:

- Your fingers, toes and sometimes the tips of your ears, nose or tongue are very sensitive to cold and turn bluish or very pale.
- Your fingers feel tingly, cold or numb in cold weather or during temperature changes.
- You may develop ulcers (sores) on the tips of your fingers or around your fingernails caused by decreased blood supply.

When your hands warm up, the blood vessels open and the normal skin color returns and symptoms improve as the blood supply to your fingers becomes more normal. Most people with scleroderma have Raynaud's phenomenon, but most people with Raynaud's phenomenon do not have scleroderma.

Swelling

Swelling or puffiness of the hands and fingers may occur. This may make it much more difficult to close your fist tightly, or use your fingers for such things as dressing, cooking, sewing, or holding a golf club.

Skin Changes

Skin changes occur in most people with scleroderma. However, the disease is different for everyone. These skin changes can include:

- The skin on your fingers and toes or other areas may look and feel swollen.
- Your skin can appear shiny.
- Usual skin creases disappear.
- You have difficulty making a fist.
- Your skin hardens and thickens, especially on the hands, arms and face.
- You lose hair over the affected area.

- You experience change in skin color, which appears darker or has areas of decreased pigment.

Sclerodactyly

Sclerodactyly (skler-o-DAK-tuh-lee) means hardness of the digits (fingers). You may notice that the skin on your fingers becomes hard and shiny and you have difficulty bending or straightening your fingers.

Telangiectasia

Telangiectasia (tel-an-jek-TAY-zhuh) occurs when tiny blood vessels near the skin's surface become dilated and show through the skin. Small reddish spots may appear on your fingers, palms, face, lips, tongue or other areas. The spots are harmless and do not mean that the disease is getting worse. Cosmetics can help hide many of the spots, and ones on the face can be removed by laser therapy.

Calcinosis

Small white calcium lumps may appear under the skin, often affecting the fingers. This is called calcinosis and is common in limited scleroderma. Most of the time these are without symptoms. Occasionally, the lumps may break through the skin and leak a chalky white liquid. They may become infected, especially when the skin over the lumps breaks down or there is drainage from the lumps.

Arthritis and Muscle Weakness

Arthritis and muscle weakness also may be symptoms of scleroderma. You may notice pain, stiffness, swelling, warmth and tenderness in your joints; general fatigue; or muscle weakness, often in your upper arms or thighs.

Digestive Problems

Scleroderma can damage your esophagus and your intestines. As a result of decreased motion in the esophagus, acid reflux may lead to inflammation that causes narrowing of your esophagus. Gastrointestinal problems may cause difficulty swallowing, heartburn, bloating, nausea or even vomiting, weight loss, diarrhea or constipation.

Dryness

Dryness of the mouth, eyes, skin and vagina (called Sjögren's syndrome) is common in people with scleroderma. The dryness is caused by decreased secretions from and scarring of the tear ducts, salivary glands and other areas of your body, such as the vagina. Without proper therapy, dental problems or damage to the surface of the eye may occur.

Heart and Lung Problems

Scleroderma may affect the heart, causing irregular heart rhythms or heart failure. The lungs may be involved by scarring (fibrosis) or thickening of the walls of blood vessels in the lungs. This can cause high blood pressure in the lungs. When the lungs or heart are affected, you may experience shortness of breath, a persistent cough, chest pain, palpitations, or swelling of your legs and feet.

Kidney Problems

People with diffuse scleroderma may have kidney problems that cause severe high blood pressure and sometimes kidney failure. Sometimes, there are no symptoms of high blood pressure, so periodic blood pressure checks are important.

HOW IS SCLERODERMA DIAGNOSED?

First, your doctor will ask you questions about your symptoms and your medical his-

tory. Then he or she will conduct a physical examination. Your doctor also will do blood tests that will help him or her make a diagnosis of scleroderma. Your doctor may do special testing, including chest X-rays, an echocardiogram and special tests to check your lungs to help find scleroderma in your internal organs.

WHAT IS THE TREATMENT?

Medication

Many drugs can help the symptoms of scleroderma. Medications used to treat scleroderma include:

- Medications that increase blood flow to your fingers and toes to treat Raynaud's phenomenon;
- Medications that treat heartburn and protect the esophagus and stomach by decreasing stomach acid;
- Nonsteroidal anti-inflammatory drugs (NSAIDs) to treat joint pain and swelling;
- Blood pressure medication to treat the high blood pressure caused by kidney involvement;
- Topical ointments and creams to help the skin;
- Disease-modifying drugs, such as methotrexate, which may decrease skin thickening and internal organ scarring; and
- Pain medications and antidepressants to improve quality of life.

Exercise

Exercise is an important part of staying healthy. Exercise helps you keep your skin and joints flexible, maintain better blood flow, and prevent contractures (shortening of the muscles and tightening of the joints). Talk with your doctor or physical therapist about which exercises are best for you.

Joint and Skin Protection

Occupational and physical therapists can teach you ways to relieve pain and increase function. They may recommend assistive devices, which are devices that can protect your joints and make doing daily tasks easier. Talk to your therapist about which assistive devices are right for you.

Skin protection can keep blood flowing to your skin and can protect your skin from injury. Keeping your body warm helps open the blood vessels in your arms, hands, legs and feet. Protect your skin by:

- avoiding cold temperatures;
- avoiding strong detergents or other substances that can dry and irritate your skin;
- using soaps, creams and bath oils designed to prevent dry skin;
- leaving calcium deposits alone; and
- keeping finger ulcers clean.

Stress Management

Emotional and social stresses can be part of living with a chronic disease. Be open about your feelings, and ask your doctor or other members of your health-care team for ways to reduce the effects of stress and fatigue.

THE ARTHRITIS FOUNDATION

The mission of the Arthritis Foundation is to improve lives through leadership in the prevention, control and cure of arthritis and related diseases.

The Arthritis Foundation supports research with the greatest potential for advances and has invested more than \$320 million in these efforts since its inception in 1948. Additionally, the Arthritis Foundation supports key public policy and advocacy efforts at a local and national

level in order to make a difference on behalf of 70 million people living with arthritis.

As your partner in taking greater control of arthritis, the Arthritis Foundation also offers a large number of programs and services nationwide to make life with arthritis easier and less painful and to help you become an active partner in your own health care.

Contact us at (800) 283-7800 or visit us on the Web at www.arthritis.org to become an

Arthritis Advocate or to find out how you can become involved.

The Arthritis Foundation acknowledges with appreciation Alverna Jenkins, Cincinnati Children's Hospital Medical Center, Cincinnati; Richard M. Silver, MD, Medical University of South Carolina, Charleston; and Frederick Wigley, MD, Johns Hopkins University, Baltimore, for their assistance in preparing this booklet.

For more information: The Arthritis Foundation offers a wide variety of books, brochures and videos about different forms of arthritis, treatment and self-management techniques to help you take control of your arthritis. To order any of these products, become an Arthritis Foundation member or to subscribe to the Arthritis Foundation's award-winning consumer health magazine, *Arthritis Today*, call (800) 283-7800. Call or visit our Web site (www.arthritis.org) to find out how you can take control of your arthritis and start living better today!

MISSION STATEMENT:

The mission of the Arthritis Foundation is to improve lives through leadership in the prevention, control and cure of arthritis and related diseases.

This brochure has been reviewed by the
AMERICAN COLLEGE OF RHEUMATOLOGY.



© 2001. Arthritis Foundation, Inc., Atlanta, GA. All rights reserved. For individual use only. Material in this publication may not be reprinted without permission. Send requests for reprint permissions in writing to 30 W. Peachtree St., Suite 100, Atlanta, GA 30309. For more information about arthritis, call 800/283-7800.

www.arthritis.org
800-283-7800