

# What You Should Know About Scleroderma

**S**clerodema is the term for a chronic disease affecting the skin and internal organs. Literally, the term means “hard skin,” and a main feature of this condition is the build-up of hard, scar-like tissue in and underneath the skin, in blood vessels, and sometimes in body organs. Because the disease affects more than the skin, it was recently renamed systemic sclerosis.

Approximately one of every 4,000 adults—about 300,000 people—in the United States has systemic sclerosis. Women are about three to four times more likely than men to develop the condition, which most often starts between the ages of 35 and 55 years.

## Diagnosing Systemic Sclerosis

Systemic sclerosis is one of many diseases for which no specific cause has yet been found. Scientists do know that it is not contagious—you cannot get it from, or give it to, another person. It is not cancer, and it is not directly inherited from one or both parents.

Systemic sclerosis is one of a group of diseases, called autoimmune diseases, in which the immune system designed to protect your body damages it instead. Normally the immune system signals cells called fibroblasts to produce collagen, a thick, fibrous material, to repair damaged tissue. In systemic sclerosis, the collagen builds up around normal tissue—skin, blood vessels, and internal

organs—and interferes with their function.

Since there are no specific diagnostic tests, detecting the disease depends on history and physical examination. Women with suspected systemic sclerosis are questioned about symptoms and examined carefully for signs of the disease. Tests may include blood work to check for certain types of antibodies and a skin biopsy (removing a tiny piece of skin to be examined under the microscope).

Almost all women with systemic sclerosis have changes in both the skin and the blood vessels. The first noticeable symptom may be Raynaud’s phenomenon, in which the small blood vessels in the hands and feet become narrow and tight when exposed to cold. This causes severe pain, numbness and tingling, and fingers may turn purple and then white. In addition to the blood vessel changes, the skin begins to look abnormally tight, shiny, and nonstretchy, particularly over the hands, feet, face, mouth, or in patchy areas over the body. The fingers and other joints may become swollen and painful.

In many women, systemic sclerosis also affects the esophagus—the muscular tube through which food passes to the stomach from the mouth. If the esophagus is involved, common

symptoms are heartburn, difficulty swallowing, or a feeling that food is coming back up from your stomach.

## Living With Systemic Sclerosis

While there is no cure for systemic sclerosis, there are many effective treatments available. Like many other chronic diseases, living as comfortably as possible involves both the medical treatment your health care providers can supply and self-care measures. An important step is educating yourself (see Resources box) and enlisting the support of your family, friends, and/or a support group. Medical treatment is important, but having those close to you offer understanding, assistance, and support on a daily basis is also crucial.

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Stress management techniques, including relaxation exercises, meditation, yoga, or biofeedback can be helpful. These approaches may increase the blood flow in hands and feet affected by Raynaud’s phenomenon, keep joints flexible, and improve ability to think clearly, function calmly, and cope with physical and emotional challenges.

Until ways are found to halt or reverse the progress of the disease, medical

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treatments concentrate on controlling the symptoms. You will probably be referred to a specialist for medical care, and it is most helpful to work with a team of care providers. Physical therapists offer exercises to keep joints and muscles working, and occupational therapists can help you adapt if tightness and swelling make it difficult to care for yourself. Nurses or social workers can connect you with local support groups and provide educational resources for you and your family. Dietitians can advise you about nutrition, and therapists can provide counseling and medication if depression is an issue.

Raynaud's phenomenon can be treated with oral medication to keep the blood vessels open. If you live in a cold climate, dress warmly when you go out. Limit your exposure to cold, including air conditioning. It is also extremely important to stop smoking; in addition to its other harmful health effects, smoking causes blood vessels to narrow. Ask your health care provider for help in quitting smoking; contact local chapters of the American Lung Association, American Heart Association, American Cancer Association; or get online help at <http://www.4woman.gov/QuitSmoking/index.cfm>.

For skin tightness and thickening, you should avoid using very hot water, which dries skin out. Use oil-based skin creams and lubricants often; petroleum jelly is inexpensive and works well. Also, use sunscreen on exposed areas daily to avoid sun damage. If you must handle detergents, cleaning products, or other harsh, drying chemicals, wear protective gloves.

If you have problems swallowing or digesting food, try eating small meals every few hours rather than three larger meals per day. Wait at least 2 hours after eating before lying down, and if possible, prop up the head of your bed with wooden blocks. Prescription

medications are helpful for heartburn, diarrhea, or constipation.

Dental problems are common in women with systemic sclerosis since skin tightening makes it difficult to open your mouth, and affected salivary glands may produce less saliva. Brush and floss regularly and keep your mouth moist by chewing sugar-free gum, drinking water, or sucking on ice chips. Also, you should see your dentist regularly. For severe dry mouth, prescription medication is available.

Although not common, some women with systemic sclerosis develop kidney problems. You can help prevent kidney damage by monitoring your own blood pressure with a home blood pressure monitor, which is available without a prescription. Blood pressure must be checked on a daily basis. Your health care provider will ask you to notify him or her right away if your blood pressure rises.

Almost half of women with systemic sclerosis develop some lung damage. There are two types: scar tissue in the lungs (pulmonary fibrosis) and high blood pressure in the artery carrying blood from the lungs to the heart (pulmonary hypertension). Medical treatment is available for both conditions. Shortness of breath, unusual tiredness, or swollen feet may be signs of lung problems; report these to your health care provider right away. It is important for women with lung problems to get a pneumonia vaccine and a yearly flu shot.

Depression is a common problem for women coping with chronic diseases, including systemic sclerosis. If you are feeling down or sad most of the time, having problems with sleep (trouble falling asleep, waking up early, or sleeping too much), losing interest in your usual activities, and experiencing appetite changes (overeating or no interest in eating), don't wait. Ask your health care provider for a referral to a

mental health therapist, or discuss tak-

### **National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse/National Institutes of Health**

1 AMS Circle  
Bethesda, MD 20892-3675  
(301) 495-4484  
or (877) 22-NIAMS (226-4267)  
TTY: (301) 565-2966  
Fax: (301) 718-6366  
[www.niams.nih.gov/](http://www.niams.nih.gov/)

### **Scleroderma Foundation**

12 Kent Way, #101  
Byfield, MA 01922  
(800) 722-HOPE  
or (978) 463-5843  
Fax: (978) 463-5809  
[www.scleroderma.org](http://www.scleroderma.org)

### **Scleroderma Research Foundation**

2320 Bath Street, Suite 315  
Santa Barbara, CA 93105  
(800) 441-CURE (2873)  
or (805) 563-9133  
[www.srfcure.org](http://www.srfcure.org)

ing medication for depression.

### **Research Offers Hope**

Ongoing research is investigating many factors to determine why systemic sclerosis occurs—a key step in discovering measures for prevention, treatment, or a cure. Some researchers are looking at possible treatments, including medications to target the immune system or to prevent fibroblasts from producing too much collagen. If you are interested in learning about research in systemic sclerosis or participating in a research program, ask your health care provider about local research projects or check out programs through the resources listed.