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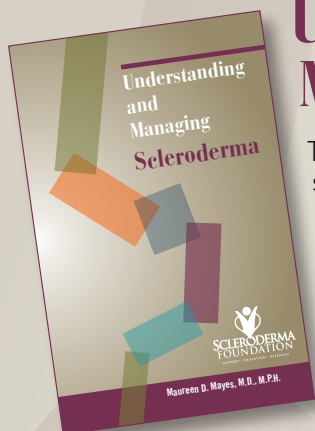
# Understanding and Managing Scleroderma



**SCLERODERMA  
FOUNDATION**

SUPPORT • EDUCATION • RESEARCH

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# Understanding and Managing Scleroderma

This booklet is intended to help people with scleroderma, their families and others interested in learning more about the disease to better understand what scleroderma is, what effects it may have, and what those with scleroderma can do to help themselves and their physicians manage the disease. It answers some of the most frequently asked questions about scleroderma.

## Disclaimer

The Scleroderma Foundation does not provide medical advice nor does it endorse any drug or treatment mentioned herein.

The material contained in this booklet is presented for general information only. It is not intended to provide medical advice, to answer questions specific to the condition or problems of particular individuals, nor in any way to substitute for the professional advice and care of qualified physicians. Mention of particular drugs and/or treatments is for information purposes only and does not constitute an endorsement of said drugs and/or treatments.

## Thanks!

The Scleroderma Foundation expresses its deep appreciation to the many physicians whose efforts have led to this booklet.

Special thanks are owed to Maureen D. Mayes, M.D., M.P.H., of the University of Texas McGovern Medical School, Houston.



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*Funding for this booklet was provided by an educational grant from Actelion Pharmaceuticals U.S., Inc.*

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## What is scleroderma?

Scleroderma is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases. The systemic form (SSc for systemic sclerosis) is different from the localized form (morphea) and this distinction will be discussed further in this brochure. To avoid confusion systemic scleroderma will be referred to as “SSc” and localized scleroderma will be called “morphea.” It is believed that these are two separate conditions that have little in common.

*The symptoms of scleroderma vary greatly from individual to individual.*

The word “scleroderma” comes from the Greek word “sclero” meaning hard, and the Latin word “derma” meaning skin. Hardening of the skin is one of the earliest and most visible manifestations of the disease. Previously SSc had been called “progressive systemic sclerosis,” but the use of that term has been discouraged since it has been found that SSc is not necessarily progressive. The disease may take several forms which will be explained later. There is also much variability among patients.

Scleroderma is a disease whose symptoms may be visible, as is the case when the skin is affected, or the symptoms may be invisible, as when internal organs are affected.

### What scleroderma is not

Scleroderma is not contagious, it is not infectious, it is not cancerous or malignant, and it is usually not hereditary.

### How serious is scleroderma?

Any chronic disease can be serious. The symptoms of scleroderma vary greatly from individual to individual. Morphea is never life-threatening, whereas SSc can range from very mild to very serious. The seriousness will depend on what parts of the body are affected and the extent to which they are affected. An early case of SSc can become more serious if not properly treated. Prompt and proper diagnosis and treatment by qualified physicians may minimize the symptoms of scleroderma and lessen the chance for irreversible damage.

## Who develops scleroderma, and when?

It is estimated that there are approximately 300,000 persons with scleroderma in the United States, including 80,000 to 100,000 with the systemic form (SSc) and the rest with the localized form (morphea). International prevalence varies from country to country and it has been reported worldwide.

Statistically, approximately three to four times more women than men develop the disease. Scleroderma can develop and is found in every age group from infants to the elderly but its onset is most frequent between 25 and 55.

Factors other than gender, such as race and ethnic background, may influence the risk of getting scleroderma, the age of onset and the pattern or severity of internal organ involvement. The reasons for this are not clear. Although scleroderma is not directly inherited, some scientists feel there is a slight predisposition to it in families with a history of rheumatic diseases. This suggests that there are some genes that can predispose toward getting scleroderma. In addition, some genes may influence the type and severity of this disease.

## What causes scleroderma?

The exact cause or causes of scleroderma are still unknown, but scientists and medical investigators in a wide variety of fields are working hard to make those determinations.

What is known about the disease process in scleroderma is that it involves three features:

- ◆ an overproduction of collagen.
- ◆ an autoimmune process.
- ◆ blood vessel damage.

Collagen is the major protein portion of the connective tissue of the body, which includes the skin, joints, tendons and parts of internal organs. Collagen is made up of tiny fibers, which weave together much like the threads forming a piece of cloth. When there is an overproduction of collagen, thickening and hardening of the affected areas takes place, often interfering with the normal functioning of those parts.

There are several theories about how collagen is overproduced. The “autoimmune theory” suggests that the body’s own immune system plays a part. Normally, the body’s immune system produces chemical signals in the blood called cytokines, which coordinate the body’s defense against bacteria, viruses and other foreign invaders. In addition, some cytokines help to repair wounds by stimulating collagen production that forms a scar. There are a number of theories in which the immune system is activated inappropriately, causing abnormal levels of cytokines to be produced. These, in turn, mount an attack not against a foreign invader but against the body’s own healthy tissues, stimulating an overproduction of collagen.

The “vascular theory” concerns blood vessels. Damage to the blood vessels, especially the small ones, is typical in scleroderma. Injury to blood vessels causes them to constrict and stiffen, and leads them to overreact to cold or stress. These reactions can cause further damage to the vessels themselves and to the organs, which they supply. There may also be a connection between the buildup of excess collagen and blood vessels, the processes which take place, and their significance for prevention and treatment.

Research is being done to study these and other theories. It is hoped that a better understanding of what causes scleroderma will lead to better treatment methods and, ultimately, to a cure.

## Are there different forms of scleroderma?

There are two major classifications of scleroderma: **localized scleroderma** (morphea) and **systemic sclerosis** (SSc). The systemic form is usually sub-divided into limited and diffuse SSc.

### Localized scleroderma

The changes, which occur in localized scleroderma, are usually found in only a few places on the skin or muscles, and rarely spread elsewhere. Generally, localized scleroderma is relatively mild. The internal organs are usually not affected, and persons with localized scleroderma rarely develop systemic scleroderma. Some laboratory abnormalities commonly seen in systemic scleroderma are frequently absent in the localized form.

**Morphea** is a form of localized scleroderma characterized by waxy, thickened patches of skin of varying sizes, shapes and pigment. There may be a single patch or several. The patches may enlarge or shrink, and often may disappear spontaneously. Morphea usually appears between the ages of 20 and 50, but it can also be seen in young children.

**Linear scleroderma** is a form of localized scleroderma which frequently starts as a streak or line of hardened, waxy skin on an arm or leg or on the forehead. Sometimes it forms a long crease on the forehead referred to as scleroderma **en coup de sabre** because it resembles a saber or sword wound. Linear scleroderma tends to involve deeper layers of the skin as well as the surface layers, and sometimes affects the motion of the joints which lie underneath. Linear scleroderma often develops in childhood although it can start in adults as well. In children, the growth of involved limbs may be affected.

### Systemic scleroderma (systemic sclerosis, SSc)

SSc can involve the skin, esophagus, gastrointestinal tract (stomach and bowels), lungs, kidneys, heart and other internal organs. It can also affect blood vessels, muscles and joints. The tissues of involved organs become hard and fibrous, causing them to function less efficiently. The term **systemic sclerosis** indicates that “sclerosis” (hardening) may occur in the internal systems of the body. There are two major recognized patterns that the illness can take - **diffuse** or **limited** disease. In diffuse scleroderma, skin thickening occurs more rapidly and involves more skin areas than in limited disease. In addition, people with diffuse scleroderma have a higher risk of developing “sclerosis” or fibrous hardening of the internal organs.

About 50 percent of patients have **limited SSc**. In limited scleroderma, skin thickening is less widespread, typically confined to the fingers, forearms, hands and face, and tends to develop more slowly. Although internal problems occur, they are less frequent and tend to be less severe than in diffuse scleroderma, and are usually delayed in onset for several years. However, persons with limited SSc, and occasionally those with diffuse disease, can develop pulmonary hypertension, a condition in which the blood vessels in the lungs become narrow, leading to decreased blood flow through the lungs and shortness of breath.

Limited SSc was formerly called the **CREST syndrome**. CREST stands for the initial letters of five common features:

- ◆ **C**alcinosis
- ◆ **R**aynaud Phenomenon
- ◆ **E**sophageal dysfunction
- ◆ **S**clerodactyly
- ◆ **T**elangiectasia

To further complicate the terminology, some people with diffuse disease will go on to develop calcinosis and telangiectasias so that they also have features of CREST.

Although most patients can be classified as having either diffuse or limited disease, different people may have different symptoms and different combination of symptoms of the illness.

### How is scleroderma diagnosed?

Diagnosis of morphea is usually made by a skin biopsy of one of the skin patches. Blood tests for SSc are usually negative.

Diagnosis of systemic sclerosis (SSc) may be very difficult, particularly in its early stages. Many of its symptoms are common to, and may overlap with, those of other diseases, especially other autoimmune connective-tissue diseases such as rheumatoid arthritis and lupus. Different symptoms may develop in stages over an extended period of time, and few people with SSc experience exactly the same set of symptoms and effects.

While SSc can often be suspected from its more visible symptoms, no single test can prove its presence. Doctors with extensive experience in the treatment of scleroderma usually make a diagnosis by a combination of the following: a patient’s medical history, including past and present symptoms; a thorough physical examination; and findings from a variety of laboratory tests and other studies. The American College of Rheumatology along with its European counterpart (European League of Associations for Rheumatology, EULAR) recently revised the classification criteria for SSc to help doctors make more accurate and early diagnosis. In managing SSc, it is important not only to confirm the diagnosis, but also to determine its extent and severity, particularly with regard to the involvement of internal organs.

Diffuse and limited scleroderma can sometimes be differentiated by the presence of different antibodies called **anti-nuclear antibodies (ANA)** in the blood. For example, **anti-Scl-70** is frequently associated with diffuse skin involvement whereas **anti-centromere** is usually indicative of limited scleroderma.

<u>Test</u>	<u>Diffuse patients</u>	<u>Limited patients</u>
Anti-nuclear antibody . . . . .	90 percent test positive	90 percent test positive
Anti-centromere antibody . . . . .	3 percent test positive	40 percent test positive
Anti-Scl-70 antibody . . . . .	30 percent test positive	20 percent test positive
Anti-RNA Polymerase III antibody . . . . .	25 percent test positive	10 percent test positive

# What are the symptoms of SSc, and how are they treated?

This section describes the most common symptoms of SSc and some of the treatments being used to control them. Scleroderma is a complex disease with many possible symptoms that can affect many parts of the body. Most people only develop a few of the symptoms mentioned. Each patient is different in the number of symptoms and their severity. Typically, the symptoms may also vary over time with periods of improvement and worsening. It is not possible in a booklet of this length to describe every symptom or all of the methods being used in scleroderma management. A great variety of treatments and medications have been tried over the years and new ones are constantly being tested. Physicians experienced in scleroderma should be consulted regarding any symptoms or treatments mentioned here, as well as for any other symptoms that may be experienced.

Even though scleroderma has no cure, many of the symptoms can be improved with medication or lifestyle changes. It is important to point out that not all complications develop in all patients even after many years.

*Even though scleroderma has no cure, many of the symptoms can be improved with medication or lifestyle changes.*

## Raynaud Phenomenon in SSc

Raynaud Phenomenon is the most common early symptom of SSc. It is present at one time or another in about 90 percent of patients. It is most obvious in the fingers and toes but can also involve the ears, nose and tip of the tongue. In Raynaud Phenomenon, the blood vessels constrict or narrow in response to cold or emotional upset and stress. The resulting disturbance in blood circulation causes a series of color changes in the skin: white, blanched

or pale, when circulation is reduced; blue as the affected part loses oxygen from decreased blood flow; and then red or flushed as blood flow returns and the part re-warms. Finally, as the episode subsides and the circulation returns to normal, usual skin color is restored. In the “white” or “blue” stages, sensations such as tingling, numbness and coldness

may be felt. In the “red” stage, a feeling of warmth, burning or throbbing may be noted. Some people find Raynaud attacks painful.

Many common-sense preventive measures can be taken by those with Raynaud Phenomenon. Most obvious is minimizing exposure to cold, such as outdoor weather, air conditioning, or reaching into a refrigerator or freezer. Keeping your extremities and body warm is very important. Gloves or mittens should be worn, and a number of warming devices are available to protect the hands. Hats, ear muffs, heavy socks and warm, layered clothing made from silk, cotton, wool and down feathers can help maintain central body temperature. It is important to protect the hands with gloves when touching refrigerated or frozen items. Electric heaters, electric blankets and comforters can supplement the heat in the home or apartment. Keeping the entire body warm helps prevent Raynaud episodes. A warm bath or shower, or heating pad or hot water bottle on the back of the neck may relieve an attack better than just warming the hands. Avoidance of emotional upset and stress can help but isn't always possible.

*Many common-sense preventive measures can be taken by those susceptible to Raynaud phenomenon.*

Various relaxation techniques, whether self-taught or learned through training courses, prove effective for some people to manage stress. One particular technique, biofeedback, has been used to increase finger temperature.

Smoking definitely worsens Raynaud Phenomenon. For this and other reasons, persons with scleroderma should not smoke.

When Raynaud Phenomenon does occur, carefully waving the arms in an underhand, circular motion (like a softball pitcher) can help to restore blood circulation. Rubbing or massaging the hands and feet may also help.

Physicians use a number of different medications to prevent, reduce the frequency or minimize the effects of Raynaud Phenomenon. Most of these drugs dilate or open up the blood vessels. Drugs commonly used include vasodilators such as prazosin, or calcium channel blockers such as nifedipine. Mild blood thinners such as aspirin, or drugs that decrease the stickiness of platelets, may improve circulation. There are now more than a dozen medications to improve circulation, and it is not possible to list them all here.

Decreased circulation to the fingers can result in skin breakdown at or near the tip of the finger resulting in painful ulcers. About 40% of SSc patients will develop at least one of these painful ulcers at some time in the course of this disease. These sores initially develop “spontaneously” due to lack of blood supply and are usually not due to an infection. However, if the area remains open for any length of time, it can become secondarily infected. Treatment of these ulcers include optimizing the circulation with medicine to dilate or open up the small blood vessels (vasodilators), being careful to keep the area clean and protected from injury. Typically, these ulcers can last for weeks and sometimes months.

Although almost everyone with SSc will also have Raynaud Phenomenon, the opposite is not true, that is, not everyone with Raynaud Phenomenon has SSc. Primary Raynaud’s disease (in the absence of an autoimmune disease) is actually quite common, affecting about 10 % of adult American women and about 5% of men. It can also be seen in people with lupus, rheumatoid arthritis and other connective tissue diseases.

### Swelling or puffiness of the hands

Swelling is another typical early symptom of SSc, and this may be especially noticeable upon waking up in the morning with some improvement as the day wears on. The skin of the fingers may look full and sausage-like, making it difficult to close the hand into a fist. Some patients may have to get their rings resized. Exercising the fingers and toes can help. Your physician may recommend medications to reduce inflammation.

### Pain and stiffness of the joints

Symptoms of pain, stiffness and swelling, may accompany the arthritis-like joint inflammation which can occur in SSc. Muscle pain and weakness are other important symptoms. Anti-inflammatory medications may help. Be sure to discuss these medications with your doctor if you have digestive problems because they may irritate the stomach. Other treatments and suggestions are included in the sections on “Physical Therapy and Exercise” and “Protecting the Joints.” A physical therapist can develop an exercise plan after consulting with your physician.

### Skin Complications

**Skin thickening.** Hardening and thickening of the skin give scleroderma its name (“hard skin.”) Some medications have been shown to improve the skin changes in scleroderma although the improvement has been relatively modest.. Many other medications and treatments are being tested. The skin will sometimes soften spontaneously over time.

**Skin ulcerations.** Sores, especially on the fingertips, are a common symptom of SSc. They typically are painful and slow to heal because of poor circulation. These sores or ulcerations may also occur on the knuckles, elbows, toes or other sites of the body where the skin is especially tight or stretched. The affected area should be kept warm to increase blood flow and carefully cleaned to avoid infection. If an infection develops, it may help to soak the affected area in warm water, apply a topical antiseptic or use an antibiotic ointment. Should these remedies prove unsuccessful in relieving the pain or infection of ulcerated skin, your doctor may prescribe oral antibiotics or take other measures.

**Calcinosis.** This condition is characterized by deposits of calcium in and just under the skin, which may be painful. The calcium deposits form hard lumps or nodules. They may break through the skin and drain chalky white material, and may become infected. Care should be taken not to bump or injure affected areas. Warm-water soaks may be helpful. Antibiotics may be prescribed to prevent or control infection. In severe cases, surgery to remove calcium deposits may be required.

**Telangiectasia.** This abnormality consists of the dilation of small blood vessels near the surface of the skin, which become visible as small red spots, usually on the fingers, palms, face and lips. The spots usually fade with pressure, but turn red again when the pressure is released. These spots are generally not harmful. Special make-up may be used to mask the spots or to reduce their visibility.

**Dry skin.** Excessive dryness of the skin may lead to skin breakdown and ulcerations. Excessive bathing and handwashing should be avoided, and rubber gloves worn to avoid direct contact with household detergents. Keeping the skin moist and well-lubricated is important to avoid complications from dry skin. Bath oils and moisturizing soaps are preferable to harsh soaps which dry out the skin. Frequent use of moisturizing skin creams containing lanolin is advised. During the winter months, a humidifier may help.

*Hardening and thickening of the skin give scleroderma its name (“hard skin.”) The skin sometimes softens spontaneously over time.*



**Itchy skin.** If moisturizing creams do not work, your doctor may prescribe a topical cortisone cream to rub on the skin to relieve itching. Antihistamines have been effective for some people.

*Keeping the skin moist and well-lubricated is important to avoid complications from dry skin.*

**Other skin symptoms.** There may be a decrease in hair over affected areas of the skin, as well as a decrease in the ability to sweat. In addition, there may be an increase in pigment (which looks like a skin tan) or a spotty loss of pigment (salt and pepper appearance). The pigment changes usually occur early in SSc and then can fade back to a more normal skin tone after 5 years or so.

### Sclerodactyly and joint contractures

Sclerodactyly means “hard skin of the digits,” particularly the fingers and toes. It generally occurs after initial swelling has subsided. It is characterized by shiny, tight and thickened skin of the fingers.

Affected digits may be difficult to move, and they may become fixed in a bent or flexed position called a “contracture” or “flexion contracture.” Tightening and hardening of the skin and tissues surrounding the joints can cause decreased motion of the wrists, elbows and other joints.

“Range of motion” exercises performed daily are important to prevent or slow down the development of such contractures and to maintain limber joints. They may also help to increase blood supply to the tissues. These exercises are simple to perform and can be done at home. A typical exercise consists of laying the hand as flat as possible on a table, placing the heel of the other hand across the fingers, and gently pressing down to straighten the fingers. An occupational therapist can develop an exercise plan after consulting with your physician. He/she also may provide devices to help perform common personal care and household tasks more easily.

### Digestive system and gastrointestinal tract problems

People with systemic scleroderma may develop abnormalities of the digestive system and gastrointestinal tract from the mouth to the anal canal. The overproduction of collagen typical of scleroderma can cause thickening and fibrosis (or scarring) of the tissues. This can result in weakened muscles, and lead to the abnormally slow movement of food (dysmotility) in the digestive process.

**Esophageal dysfunction.** Food travels from the mouth and throat into the stomach through a tube called the esophagus. Normally, the lower esophageal sphincter, or valve, acts as a gate which opens to allow food to enter the stomach and then closes promptly to prevent food from coming back up. In SSc, this valve does not close properly and the result is a backwash of acid and a burning sensation (heartburn, gastroesophageal reflux disease or GERD) as food and acid return into the esophagus. The acid may also injure the lining of the lower portion of the esophagus, causing ulcers or a narrowing (stricture) of the tube.

Acid production can be reduced, and the problems of acid reflux and heartburn helped, by avoiding alcohol, greasy or fatty foods, spicy foods, carbonated beverages, chocolate, tobacco and caffeine. Antacids (particularly in liquid form) can help neutralize acids and reduce heartburn. Some antacids cause constipation while others cause diarrhea. Consult your physician or pharmacist when choosing over-the-counter products. Your doctor may prescribe antacid medications such as proton pump inhibitors or H-2 blockers to decrease acid production in the stomach. The physician also may prescribe a drug such as metoclopramide which promotes muscular activity and causes the muscles of the GI tract to work better.

The force of gravity helps to keep food and acid in the stomach; therefore, an upright position during and after meals is helpful. Other common-sense measures to prevent acid from coming up into the esophagus include eating smaller and more frequent meals, not eating for several hours before bedtime, and elevating the head of the bed six-to-eight inches with wooden blocks. Being overweight is harmful, and you should avoid wearing girdles or other tight-fitting garments.

**Swallowing difficulties.** Abnormally slow movement of food and narrowing of the esophagus may cause swallowing difficulties. Eating slowly and chewing thoroughly are important. Swallowing and digesting are made easier by eating softer foods (many foods can be prepared in a blender) and avoiding foods which tend to stick in the throat. If the esophagus has narrowed significantly, the physician may need to dilate the esophagus periodically to permit easier swallowing.

*“Range of motion” exercises performed daily are important to prevent or slow down the development of such contractures and to maintain limber joints.*

**Diarrhea.** In SSc, there can be damage to the muscles of the small bowel (small intestine). The weakened muscles do not work effectively to push food through the bowel. Simply put, things sit rather than move well.

One consequence can be an overgrowth of bacteria, leading to diarrhea. There also may be a bloated, distended feeling and some pain if the bowel is stretched. Another effect is that the nutrients of food remain in the bowel instead of being absorbed into the body. This condition is called malabsorption, and it may lead to weight loss and stool abnormalities.

For diarrhea or malabsorption, the physician may prescribe an antibiotic, or supplementary fat-soluble vitamins, and/or iron. Your doctor also may suggest that you reduce the amount of fatty foods in your diet.

**Constipation.** Weak or scarred muscles in the colon (large bowel) make it difficult for the bowel to work well, resulting in constipation. Fresh fruits and vegetables are natural laxatives. Exercise also helps to keep bowel movements regular. Your doctor also may recommend stool softeners. Although the routine use of laxatives is usually to be avoided, this may be necessary if there is persistent constipation unresponsive to the measures suggested above.

## Sjögren Syndrome

Sjögren Syndrome (dry eyes and dry mouth) is characterized by a decrease in secretions of the tear glands and the salivary glands, which provide lubrication for the eyes and mouth. The unusual dryness of the eyes resulting from this condition can lead to serious irritation and inflammation. Excessive dryness of the mouth may lead to difficulties in swallowing and speaking, an increase in tooth decay and cavities, and a reduced sense of taste. The lack of secretions in Sjögren Syndrome also may involve the vagina. Dry eyes may be lubricated by the frequent use of artificial tears and ophthalmic ointments. Regular visits to an ophthalmologist are important. The mouth should be kept as well-lubricated as possible by sipping fluids during the day (a plastic squirt bottle filled with water may help), and by chewing sugar-free gum or sucking sugar-free sour candy to stimulate salivary activity. Artificial saliva is also available. (See next section for preventive dental care.)

Your doctor may prescribe a propionic acid gel preparation or vaginal cream to lubricate the vagina and facilitate sexual relations. Avoiding pantyhose and other tight-fitting clothing may help to reduce irritation and prevent infection. Choose cotton rather than nylon underwear.

## Oral, facial and dental problems

People with SSc may experience a general tightening of skin over the face. The opening of the mouth may be decreased in size, making lip and mouth movements as well as oral hygiene difficult.

The best approach to treatment is by means of facial grimacing and mouth stretching exercises, including the use of oral augmentation props inserted between the upper and lower teeth. When doing these exercises, be careful to avoid further damage, especially to the jaw bone and teeth. Particular care is advisable in using augmentation devices. Consult with your dentist before starting any exercises or using any augmentation device.

Preventive dental care including regular flossing and brushing of the teeth and gums is very important, as are regular dental visits for oral health and for the early detection and prompt correction of any abnormalities. The dentist also can recommend a good oral hygiene program. Floss holders, pump toothpaste tubes and built-up handles on toothbrushes can help people with hand impairment. (These measures are equally important for those with Sjögren Syndrome.)

## Kidney involvement

Kidney or renal involvement in SSc may be mild or very serious in nature. Early signs of kidney involvement may include mild hypertension (high blood pressure), protein in the urine and blood test abnormalities. Renal crisis, a highly dangerous complication may occur quite quickly. The most important warning sign is a sudden rise in blood pressure. Other symptoms are headache, visual disturbances, shortness of breath, chest pain or discomfort, or mental confusion. Unless treated promptly, renal crisis leads to kidney failure, a condition in which the kidneys lose their ability to eliminate waste products from the body. The treatment of choice involves anti-hypertensive drugs that belong to the category of ACE inhibitors. These medications are quite effective to control blood pressure and stabilize or improve kidney function. In cases

*The most important warning sign is a sudden rise in blood pressure.*

of severe kidney failure, dialysis may be required. People with early SSc are advised to check their blood pressure at home and to have their kidney function monitored at regular intervals. People may recover successfully from renal crisis, but only if the problem is recognized and treated quickly.

### Lung involvement

Multiple factors can cause lung involvement in systemic scleroderma. Build-up of collagen thickens lung tissue and causes fibrosis or scarring, making the transport of oxygen into the bloodstream less efficient.

Pulmonary arterial hypertension (PAH), a state of increased resistance to blood flow through the lungs, can result from damage to blood vessels, and may lead to additional strain on the heart resulting in heart failure. Respiratory muscle weakness may decrease lung function.

Symptoms of lung involvement include shortness of breath, a decreased tolerance for exercise and a persistent cough. The physician may order a chest x-ray, an echocardiogram (ultrasound

of the heart), special breathing tests (pulmonary function tests) or a CAT scan of the lungs to detect or confirm lung involvement.

In the early stages of lung fibrosis, medications may be given to decrease the inflammation which is thought to lead to lung scarring.

Recently two different medicines have been shown to improve or slow the progression of lung fibrosis in SSc. These include cyclophosphamide (a form of chemotherapy) and mycophenolate mofetil (a medicine that causes immunosuppression). In addition, several other drugs are being studied and hold promise for this complication. It is also important for SSc to take whatever measures are within his or her control to avoid further damage to the lungs. It is essential to avoid smoking, a major cause of lung disease. Exposure to air pollutants may worsen breathing problems and should be avoided to the extent possible. Your doctor may recommend medications to make breathing easier and may also suggest deep breathing exercises and a graduated aerobic exercise program.

People with pulmonary arterial hypertension may be treated with special medications targeted at dilating or opening up the blood vessels of the lungs, and possibly changing the underlying nature of the disease. This is another complication of SSc for which new medications have proven successful; there are now multiple medications which can be used.

*Symptoms of lung involvement include shortness of breath, a decreased tolerance for exercise and a persistent cough.*

### Heart involvement

If the heart muscle becomes thickened and fibrous scar tissue accumulates, the force of heart contractions may be decreased, which limits the efficiency of the heart. If the scarring involves the electrical conduction system of the heart, palpitations or changes in heart rhythm can result. There are medication which can improve this and sometimes a pacemaker is needed. Inflammation of the outer heart lining (pericarditis) may cause pain and accumulation of fluid around the heart. An irregular heartbeat may also occur. These conditions require careful evaluation and treatment by the physician.

### Non-specific symptoms

The person with SSc may experience a variety of non-specific symptoms, including fatigue (ranging from mild to severe), lack of energy, generalized weakness, weight loss, and a vague aching of muscles, joints or bones. Treatments or medications recommended by the physician will depend on his/her evaluation of the causes of these symptoms.

## Managing scleroderma

You may know other forms of treatment that have been used or are proposed for use to manage scleroderma in addition to those discussed in this booklet. Scleroderma is a difficult disease to study because of its variable nature, its prolonged course and the relatively small number of persons affected by it. Currently, there are several large clinical trials being conducted for the different manifestations of SSc. Your physician must often make treatment decisions based on incomplete information. He/she must weigh the possible benefits against the potential risks or side effects.

### The course of scleroderma

Scleroderma has many forms and a number of different symptoms that may present themselves alone or in combinations at various times throughout the course of the disease. Some symptoms develop with relative suddenness; others may take years to develop. The exact course in any individual patient is somewhat unpredictable, and the prognosis (long-term outcome) will vary for each person. SSc is a chronic, life-long disease. Currently, there is no known cure, but as with other chronic diseases there are many ways to control or manage its symptoms. It

is helpful to keep scleroderma in perspective. Many persons with the disease have few or minimal symptoms and are able to lead a normal or nearly-normal life.

There may be periods of time when the person with scleroderma will be free of troubling symptoms and feel well. At other times, he or she may feel quite ill. Spontaneous improvements may occur. The skin, in particular, sometimes softens and becomes more pliable after a number of years.

### Being alert to symptoms

This booklet describes many symptoms although each person with scleroderma usually develops only a few of them. Its purpose is not to overwhelm people but to provide them with useful information on what to look for, what may occur during the course of the disease, and some of the things that can be done if symptoms do develop.

Learning to recognize early symptoms of disease activity can lead to earlier detection and diagnosis of scleroderma and to prompt start of treatment. Some of the more promising medications in current use are slow-acting and the sooner treatment begins, the better the results may be. If one has already been diagnosed with scleroderma, it is especially important to watch for and report to the physician new or changed symptoms. Early treatment may prevent symptoms from worsening and may decrease the chance of permanent tissue or organ damage.

In being alert to symptoms, it would be a mistake to assume that every symptom or condition that develops is necessarily related to, or the result of, scleroderma. People with or without scleroderma do suffer accidents, contract infectious diseases and develop other illnesses.

The physician can help to distinguish what is related to scleroderma and what is not, and recommend appropriate treatment.

### Developing an individual treatment program

While there is no proven cure for scleroderma, much can be done to prevent, minimize or alleviate its effects and symptoms. Just as SSc symptoms vary greatly from individual to individual; the manner in which each person responds to treatment may also vary; and there are many treatment options. It is important that a physician experienced in scleroderma management works out an individually-tailored program to meet the specific needs of a person with this disease. Close cooperation with the physician will help him or her develop such a program.

Many forms of treatment have been discussed already in the chapter titled "What are the symptoms of scleroderma, and how are they treated?" The next six subheads will discuss other important elements of a program for managing scleroderma.

### Physical therapy and exercise

Physical therapists can help the person with scleroderma develop an appropriate program. Such a program may consist of "range of motion" exercises (as mentioned in the previous chapter under the subhead "Sclerodactyly and joint contractures"), paraffin wax baths, hydrotherapy or water therapy, strengthening exercises for muscle weakness and gentle massage. These treatments can be done at various locations, including a hospital physical therapy department or at home.

Your doctor may recommend an exercise program involving activities such as stretching, walking or swimming. Persons with scleroderma may find that their tolerance for activity and movement is below normal, so activities should be carried out in moderation, resting when tired. Individual exercises should be performed gently and with due care, and the exercise program should be built up gradually.

*Your doctor may recommend an exercise program involving activities such as stretching, walking or swimming.*

### Protecting the joints

Joint protection helps minimize further damage and reduce the possibility of skin ulcers and infection. Its basic principles include avoiding or minimizing pressure or stress on the joints by their proper use, and maintaining mobility and function by stretching and "range of motion" exercises. A variety of self-help aids and adaptive mechanical devices are available to help protect and alleviate stress on the joints while still completing daily activities. Occupational therapists can demonstrate such devices and give further instruction on joint protection.

### Taking medications

It is essential that a person with scleroderma take all medications wisely; take only those prescribed; read label warnings and follow instructions carefully; and take the medications when, for how long, and in the

dosages prescribed by the physician. The person with scleroderma should advise the physician of any drugs taken for other conditions including over-the-counter preparations, supplements or vitamins. Any side effects encountered should be promptly reported and discussed with your doctor.

One should not be concerned if the physician prescribes different medications for different people. Scleroderma symptoms vary from person to person, requiring different treatment. Some may benefit from certain drugs, while others may not. Furthermore, individual tolerance for the drugs used in scleroderma varies greatly. The physician may find it necessary to adjust the medication program accordingly.

### Common-sense measures

We have discussed treatment options for specific symptoms in other sections of this booklet. There also are a number of general common-sense measures that

a person with scleroderma can take to enhance his/her well-being. These measures include:

- ◆ Avoiding over-fatigue by taking it easy and getting sufficient rest. Knowing your own limits does not indicate you are lazy.
- ◆ Learning to control and minimize stress.
- ◆ Eating well-balanced meals and maintaining a sensible weight.
- ◆ Practicing good hygiene habits, especially of the skin, teeth, gums and feet (including the wearing of cushioned and well-fitted shoes).
- ◆ Avoiding smoking. The health risks of smoking are well known but frequently ignored. It is particularly dangerous to persons with scleroderma because it can have effects on blood circulation and lung function.

### The emotional aspects of scleroderma

A common reaction to being told that one has a disease such as scleroderma is "Why me?" It is not known why some people develop this disease and others do not. One does not bring scleroderma upon himself or herself; therefore, one shouldn't feel guilty or responsible for the illness.

A person newly diagnosed with scleroderma may feel alone and uncertain about where to turn for help. He or she may experience a number of other feelings and emotional reactions from time to time, including initial shock or disbelief, fear, anger, denial, self-blame, guilt, grief, sadness or depression. Family members may have similar feelings.

Feelings in themselves are neither good nor bad. One simply has them. Sharing them with family and friends or with others who have had similar experiences can help. Professional counseling also can help people with scleroderma and their family members who are having difficulty coping with their feelings.

The person with scleroderma may be a "patient" in the physician's office, hospital or clinic, but he or she is much more than that. Thinking of oneself as a total person with a full life to lead may help to keep scleroderma in perspective and enable one to maintain a positive but realistic attitude.

### Building a health and support network

Participating actively in one's own health care is of prime importance to the person with scleroderma. It is equally important to cooperate and communicate effectively with the physician who is managing the disease. While these two—the person with scleroderma and the physician—are the focal point of the management "team," many other people and resources also form a health and support network.

Family and friends can provide emotional support for the person with scleroderma, encourage him/her to follow the recommended treatment program, and assist in carrying out activities that he/she finds difficult.

The health team begins with the physician, but can include many other health professionals such as medical specialists, nurses, physical and occupational therapists, and psychologists or others trained in counseling.

Directories of community resources typically list a large number of voluntary and governmental agencies providing health, social and rehabilitation services that may benefit a person with scleroderma.

*One does not bring scleroderma upon himself or herself; therefore, one shouldn't feel guilty or responsible for the illness.*

*Participating actively in one's own health care is of prime importance to the person with scleroderma.*

Joining a scleroderma support group, such as one affiliated with the Scleroderma Foundation, enables the person with scleroderma to meet and exchange information with others who have similar problems, as well as to learn more about scleroderma. The Foundation also manages an online support group community at <http://www.inspire.com/groups/scleroderma-foundation>.

The extent of the health and support network is limited only by the imagination and resourcefulness of those helping to create it.

## Progress through research

Is there hope and help, for a person with scleroderma?

Emphatically, yes!

As this booklet has discussed, there are many treatments and medications available to help a person with scleroderma, and more and more physicians are becoming interested in the disease.

Investigators throughout the United States and other countries are intensifying their efforts to understand the nature and discover the cause of scleroderma, to find better means of prevention and treatment, and to find a cure. These efforts reflect the increased interest in all of the connective-tissue and rheumatic diseases.

Research has already resulted in better laboratory tools to detect the early stages of scleroderma and improved methods of measurement to evaluate disease progression and the results of treatment. Various animal models of scleroderma have been developed.

Investigators currently are studying the role of the immune system in scleroderma, exploring the relationship between blood vessel changes and fibrosis, and seeking markers to identify the various forms and subsets of scleroderma. These are just a few of the many studies in progress.

Scleroderma poses many questions. Answers may come from a variety of medical and scientific fields or from totally unexpected sources...but they will come!

*Participating actively in one's own health care is of prime importance to the person with scleroderma.*

## The Scleroderma Foundation

The Scleroderma Foundation is the national nonprofit organization in the U.S. representing and advocating for persons with scleroderma. The Scleroderma Foundation is the leading nonprofit supporter of scleroderma research, funding on average more than \$1 million of new grants each year to find the cause and cure of scleroderma. The Foundation's three-fold mission is support, education and research.

The Scleroderma Foundation has chapters and support groups nationwide to help people living with scleroderma and their families. The Foundation's services include:

- ◆ a toll-free telephone hotline, **800-722-HOPE** (4673);
- ◆ an email address for patient questions, **[sfinfo@scleroderma.org](mailto:sfinfo@scleroderma.org)**;
- ◆ a website with news and information, **[www.scleroderma.org](http://www.scleroderma.org)**;
- ◆ a quarterly magazine, *Scleroderma Voice*;
- ◆ a full line of informational literature;
- ◆ a national conference for persons with scleroderma and their families, featuring leading medical experts and other health professionals;
- ◆ educational meetings and workshops across the country;
- ◆ medical referrals; and many other support services. The Foundation and its chapters and support groups also put people with scleroderma in contact with each other.

At the national and international levels, the Scleroderma Foundation supports people with scleroderma and their families in many ways. The Foundation increases public and professional awareness of scleroderma through national publicity campaigns. It distributes information about scleroderma, and produces educational literature (such as this booklet). The Foundation advocates in Washington, D.C., for increased federal funding of scleroderma research, and for legislation benefiting persons with scleroderma.

The Scleroderma Foundation can put you in touch with the chapter or support group nearest to you. If there is no support group nearby, the Foundation can help you start one.

# Glossary

Here are some useful definitions of medical words and terms.

**Acid reflux, heartburn.** Stomach acid which abnormally travels up into and irritates the esophagus. (Acid production is a normal part of digestion in the stomach.) Heartburn refers to pain in the center of the chest caused by acid reflux. (See **Esophagitis**.)

**Analgesic.** A medication which reduces or eliminates pain. Example: aspirin and non-steroidal anti-inflammatory drugs.

**Antacid.** An agent which neutralizes excess stomach acid. This may be liquid/tablets which act immediately in the stomach, or long-acting medications taken regularly and absorbed into the blood in order to suppress acid production. (See **Acid reflux**.)

**Antibiotic.** Medication used to treat an infection. Each antibiotic kills or inhibits the growth of specific microorganisms, so antibiotics are prescribed based on the type of infection present.

**Arthralgia.** Pain in a joint.

**Autoimmune.** Disease or antibody which acts against the person's own tissues. (See **Immune system**.)

**Biofeedback.** A technique used to regulate a body function usually involuntarily controlled, such as a finger temperature or pulse rate. By observing a machine monitoring the function, a person can practice relaxation techniques and learn to control the function. Later, the machine becomes unnecessary. (See **Relaxation techniques**.)

**Biopsy.** The removal and examination of tissue, cells or fluid from the body.

**Blanched.** To become white or pale. In Raynaud Phenomenon, the fingers and toes blanch due to insufficient circulation of blood.

**Calcinosis.** Abnormal accumulation of calcium in the skin.

**Capillaries.** The smallest blood vessels of the body, connecting arteries and veins.

**Collagen.** A normal, fibrous protein found in the connective tissue of the body.

**Connective tissue.** Tissue which pervades, supports and binds together other tissues including mucous, fibrous, reticular, adipose, cartilage, skin and bone. Connective-tissue diseases are a group of diseases with similar cellular changes, but with the site where the changes occur determining the specific disease. Included are scleroderma, systemic lupus erythematosus, dermatomyositis and rheumatoid arthritis.

**Constrict (vessels), stricture (esophagus).** An abnormal narrowing.

**Contraction (of intestinal muscles).** The rhythmic squeezing action of the muscles of the wall of the intestine which moves food through the system. Also called peristalsis. (See **Motility**.)

Coronary arteries. Blood vessels which supply blood to the heart itself.

**CREST.** Form of scleroderma, whose initials stand for Calcinosis, Raynaud Phenomenon, Esophageal dysmotility, Sclerodactyly and Telangiectasia.

**Cutaneous.** Of the skin.

**Cyanosis.** Blue or purple color due to lack of blood oxygen. In Raynaud Phenomenon, cyanosis of the fingers and toes may follow blanching.

**Digits.** Fingers and toes.

**Dilate.** (esophagus, blood vessels). To widen or enlarge.

**Diuretic.** Medication to increase the flow of urine, thereby decreasing fluid retention in the tissues. Also called "water pills." (See **Edema**.)

**Dysfunction, disfunction.** Impaired or abnormal functioning.

**Dysphagia.** Difficulty in swallowing.

**Edema.** An abnormal excess accumulation of fluid in tissues or cavities of the body.

**En coup de sabre.** A form of localized scleroderma which forms a long crease of waxy skin, resembling a cut by a saber or sword wound usually on face or neck.

**Esophagus, esophagitis.** The muscular swallowing tube connecting the mouth and the stomach. When properly functioning it contracts in smooth waves to send food to the stomach. At its lower end a sphincter (ring-like muscle) opens to allow food to pass into the stomach, but closes again to prevent stomach acid or partially digested food from backing up into the esophagus. Esophagitis is an inflammation or irritation of the esophagus.

**Fatigue.** Weariness, a sense of being overwhelmingly tired, or exhaustion.

**Fibrous.** Consisting of, or resembling fibers.

**Fibrosis.** Abnormal formation of excess fibrous tissue.

**Gastrointestinal tract, bowel, diarrhea, constipation.** The gastrointestinal tract is the digestive system which breaks down food, allows absorption of nutrients, removal of cellular waste products, and elimination of solid waste from the body. It begins with the mouth and esophagus and leads to the stomach. The small intestine consists of the duodenum, jejunum and ileum. Lastly, the large intestine (also called colon) leads to the rectum. The term bowel refers to the intestine. The anal sphincter is the muscle which controls discharge of stool. Diarrhea is abnormally frequent or excessive passing of stool, usually watery. Constipation is the abnormally delayed or infrequent passage of stool, usually in a dry and hardened state. Normal bowel movements vary from person to person and with diet.

**Hypertension, anti-hypertensive.** Abnormally high blood pressure. An anti-hypertensive medication lowers blood pressure.

**Immune system.** The system of organs, cells and proteins which protect the body from foreign substances by producing immune responses. The immune system organs include the thymus, spleen, lymph nodes and bone marrow. The cells include white cells, lymphocytes, T cells and B cells. Immunoglobulins (antibodies) are proteins that can react with and/or neutralize corresponding proteins called antigens (usually damaged or foreign material). The immune system is essentially protective and helpful to the body, but can be the cause of disease and allergy when it attacks parts of the normal body in a process called autoimmunity.

**Inflammation, anti-inflammatory.** Tissue reaction to cell injury marked by redness, heat, pain, swelling and often loss of function. Capillary dilation and white blood cell infiltration help eliminate foreign substances and damaged tissue, so normally, inflammation is a natural part of the healing process. Excessive or inappropriate inflammation can, however, cause further damage. Anti-inflammatory drugs counteract inflammation.

**Joint contracture, flexion contracture.** Fixation of a joint in one position preventing full range of motion. In scleroderma, frequently affecting the fingers, due to tightening and hardening of the skin around the joint. In flexion contractures, the fingers become fixed in a bent or flexed position.

**Lacrimal glands.** Tear-producing glands, also spelled lachrymal.

**Laxative.** A medication which stimulates emptying of the bowels.

**Lubrication, secretion.** Substance which makes a surface slippery or oily, either artificially by applying lubricating fluids or naturally by secreting fluids made by cells for this purpose. Example: tears.

**Malabsorption.** The reduced ability to take nutrients from food into the cells of the body from the digestive tract.

**Microstomia.** Abnormally small mouth opening.

**Mixed Connective Tissue Disease.** Overlap or presence of symptoms of two or more diseases simultaneously. (See **Collagen** and **Connective tissue**.)

**Morphea.** Localized scleroderma.

**Motility, dysmotility.** Contractions of the digestive-tract muscles occurring in rhythmic waves, propelling food, allowing absorption of nutrients, and elimination of wastes (feces). Dysmotility indicates weakened or absent waves of contraction resulting in abnormally slow movement of food and feces. (See Malabsorption, Gastrointestinal tract, Contraction.)

**Occupational therapy.** Therapy using activity prescribed to promote recovery or rehabilitation. Often designed to increase ability to perform acts of daily living, such as grooming and eating, and concentrating on the hands and small muscle control. (Abbreviated "OT." See also Physical therapy.)

**Ophthalmic.** Related to, or situated near the eye.

**Pericarditis.** Tissue inflammation of the sac enclosing the heart.

**Peripheral blood circulation.** The flow of blood to the arms and legs.

**Phenomenon.** An unusual, significant, or unaccountable fact or occurrence which, when observed, is of scientific interest.

**Physical therapy.** Treatment of disease and injury by mechanical means such as massage, regulated exercise, water, light, heat and electricity. Often concerned primarily with joint motion, large muscle groups, and activities such as walking and aerobic and isometric exercise. (Abbreviated "PT." See also **Occupational therapy**.)



**Pleurisy.** Tissue inflammation of the sac enclosing the lungs.

**Prognosis.** Prediction of the progression and end result of a disease, or estimate of chance of recovery.

**Pulmonary fibrosis.** A process in which the lungs are scarred, decreasing the transfer of oxygen to the blood. Also called restrictive lung disease.

**Pulmonary hypertension.** Elevated pressure in the blood vessels of the lungs, decreasing blood oxygen and straining the right side of the heart.

**Raynaud Phenomenon.** Also called Raynaud Syndrome. A disorder with recurring spasms of the small blood vessels upon exposure to cold; characterized by fingers and toes turning white, blue, and red as circulation abnormally overreacts to normal conditions. Emotional stress may also trigger an attack. Named for the French physician (Dr. Maurice Raynaud, pronounced "Raynode") who first described it.

**Relaxation techniques.** Stress-reducing procedures, which can also be used to help regulate body functions such as finger temperature or pulse rate. These include tensing and relaxing muscles, imagery, breathing techniques, and medication. (See also **Biofeedback**.)

**Remission, spontaneous remission.** A period during which the symptoms of a disease decrease or go away. If the reason for remission is not related to treatment but seems to occur for no apparent reason, it is called spontaneous.

**Renal.** Relating to the kidneys.

**Respiratory.** Pertaining to breathing or the lungs.

**Salivary glands.** Glands which secrete fluid (saliva) into the mouth.

**Sclerodactyly.** Thick, tight skin of the fingers and/or toes. (See Joint contracture.)

**Sclerosis.** An abnormal hardening of tissue.

**Sjögren Syndrome.** A chronic inflammatory disease characterized by decreased secretions, especially dry eyes and dry mouth, named for the Swedish physician who first described it. It may occur alone, or as a part of scleroderma or other auto-immune diseases. (Pronounced "show-gren.")

**Skin ulceration.** A break in the skin with loss of surface tissue. It may also be associated with inflammation, calcium deposits and infection.

**Spasm.** Involuntary and abnormal contraction of muscle.

**Stasis.** A slowing or stoppage of body fluids as in venous stasis. Also, reduced motility of the intestines with retention of feces.

**Systemic.** Affecting the whole body rather than one of its parts. Opposite of localized.

**Telangiectasia.** An abnormal dilation of skin capillaries causing red spots on the skin.

**Vascular.** Pertaining to, or composed of blood vessels.

**Vasodilator.** A medication (or other substance) which causes widening of blood vessels.

# Additional resources

Here are some more sources of good and reliable information about scleroderma.

## Websites

*Scleroderma Foundation*

[www.scleroderma.org](http://www.scleroderma.org)

The national nonprofit organization in the U.S. representing and advocating for persons with scleroderma.

*National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)*

[www.nih.gov/niams](http://www.nih.gov/niams)

NIAMS, a division of the National Institutes of Health (NIH), oversees federal research funding and other federal programs pertaining to scleroderma and related diseases.

*American College of Rheumatology*

[www.rheumatology.org](http://www.rheumatology.org)

The American College of Rheumatology is an organization of physicians, health professionals, and scientists that advances rheumatology through programs of education, research and advocacy to foster excellence in the care of people with arthritis and rheumatic and musculoskeletal diseases.

*American Academy of Dermatology*

[www.aad.org](http://www.aad.org)

This national professional association for dermatologists can provide physician referrals in your local area and information about skin diseases.

## Recommended Reading

*The Scleroderma Book*, Second edition

By Maureen Mayes, M.D., M.P.H.

A comprehensive guide to the disease written especially for patients and their families.

# My notes

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# SCLERODERMA FOUNDATION

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