

U
nderstanding
&M
anaging

SCLERODERMA



Maureen D. Mayes, M.D., M.P.H and Khanh T. Ho, M.D.

Understanding & Managing Scleroderma

This booklet is intended to help persons with scleroderma, their families, and others interested in scleroderma 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 questions most frequently asked 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., and to Khanh T. Ho, M.D., both of the University of Texas Medical School, Health Science Center at Houston.



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



Khanh T. Ho, M.D.

Funding for this booklet was provided by an educational grant from Actelion Pharmaceutical Corporation.

Contents

	Page
■ What is scleroderma?	3
What scleroderma is not	3
How serious is scleroderma?	3
Who develops scleroderma, and when?	3
What causes scleroderma?	4
Are there different forms of scleroderma?	5
Localized scleroderma	5
Systemic scleroderma (systemic sclerosis)	6
How is scleroderma diagnosed?	7
■ What are the symptoms of scleroderma, and how are they treated?	8
Raynaud's phenomenon	8
Swelling or puffiness of the hands	10
Pain and stiffness of the joints	10
Skin disorders	10
Skin thickening	10
Skin ulcerations	10
Calcinosis	11
Telangiectasia	11
Dry skin	11
Itchy skin	11
Other skin symptoms	11
Sclerodactyly and joint contractures	11
Digestive system and gastrointestinal tract problems ...	12
Esophageal dysfunction	12
Swallowing difficulties	13
Diarrhea	13
Constipation	13

Contents

	Page
Sjögren's Syndrome	14
Oral, facial, and dental problems	14
Kidney involvement	15
Lung involvement.....	15
Heart involvement.....	16
Non-specific symptoms.....	17
■ Managing scleroderma	17
The course of scleroderma	17
Being alert to symptoms	18
Developing an individual treatment program.....	18
Physical therapy and exercise	19
Protecting the joints.....	19
Taking medications	20
Common-sense measures.....	20
The emotional aspects of scleroderma	21
Building a health and support network.....	21
■ Progress through research.....	22
■ The Scleroderma Foundation.....	23
■ Glossary	24
■ Additional resources.....	29

What is scleroderma?

Scleroderma, or systemic sclerosis, is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases.

The word “scleroderma” comes from two Greek words: “sclero” meaning hard, and “derma” meaning skin. Hardening of the skin is one of the most visible manifestations of the disease. The disease has been called “progressive systemic sclerosis,” but the use of that term has been discouraged since it has been found that scleroderma 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.

The symptoms of scleroderma vary greatly from individual to individual.

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, and the effects of scleroderma can range from very mild to life-threatening. The seriousness will depend on what parts of the body are affected and the extent to which they are affected. A mild case 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 200,000 persons with scleroderma in the United States, including 80,000 to 100,000 with the systemic form and the rest with the localized form. International incidence is unknown, but 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 the ages of 25 to 55.

Factors other than sex, 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.

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.

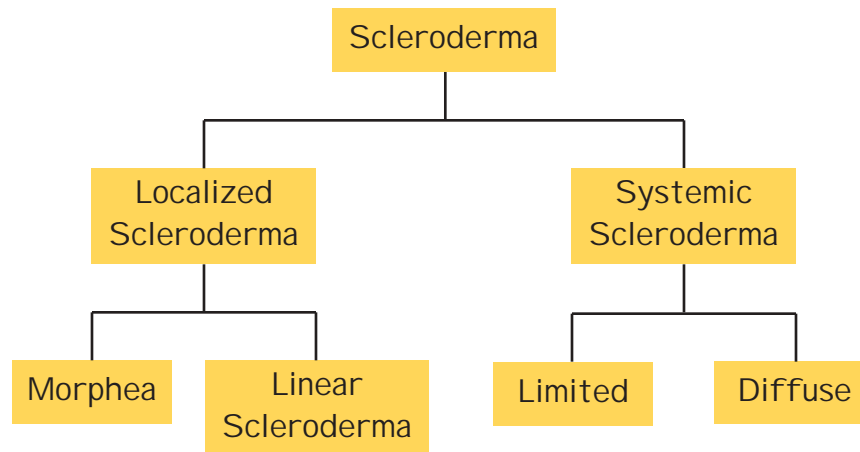
<hr/> <hr/> <p>The disease process in scleroderma involves overproduction of collagen, an autoimmune process, and blood-vessel damage.</p>	<p>What is known about the disease process in scleroderma is that it involves three features: 1) an overproduction of collagen; 2) an autoimmune process; and 3) blood-vessel damage.</p> <p>Collagen is the major protein portion of the connective tissue of the body, which includes the skin, joints, and tendons, and parts of internal organs. The structure of collagen is made up of tiny fibers, which are woven 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.</p> <p>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 on the way 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.</p> <p>Another theory, the “vascular theory,” concerns blood vessels. Damage to the blood vessels, especially the small ones, is typical in</p>
--	--

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 and systemic sclerosis (SSc). Other forms or sub-classifications, each with its own characteristics and prognosis, may be identified through future research.



Localized scleroderma

The changes, which occur in localized scleroderma, are usually found in only a few places on the skin or in the muscles, and only 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 patches on the skin of varying sizes, shapes, and coloration. The skin

underlying the patches may thicken. The patches may enlarge or shrink, and may often disappear spontaneously. Morphea usually appears between the ages of 20 and 50, but is often 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 head or neck, referred to as *en coup de sabre* because of its resemblance to 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 usually develops in childhood. In children the growth of involved limbs may be affected.

Systemic scleroderma (systemic sclerosis)

The changes occurring in systemic scleroderma may affect the connective tissue in many parts of the body. Systemic scleroderma can involve the skin, esophagus, gastrointestinal tract (stomach and

The changes occurring in systemic scleroderma may affect the connective tissue in many parts of the body.

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 one-half of patients have a slower and more benign illness called limited scleroderma. In limited scleroderma, skin thickening is less widespread, typically confined to the fingers, hands, and face, and develops slowly over years. 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 many years. However, persons with limited scleroderma can develop pulmonary hypertension, a condition in which the blood vessels of the lungs become narrow, leading to impaired blood flow through the lungs.

Limited scleroderma is sometimes called the CREST syndrome. CREST stands for the initial letters of five common features: Calcinosis; Raynaud's phenomenon; Esophageal dysfunction; Sclerodactyly; and Telangiectasia (these features are discussed later in this booklet). To further complicate the terminology, some people with diffuse disease will go on to develop calcinosis and telangiectasias so that they also have the 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 scleroderma may be very difficult, particularly in its early stages. Many of its symptoms are common to, or 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 a very long period of time, and few persons with scleroderma experience exactly the same set of symptoms and effects.

While scleroderma can often be suspected from its more visible symptoms, no single test can prove its presence. The diagnosis is usually made by physicians with extensive experience in the treatment of scleroderma by a combination of the following: the medical history, including past and present symptoms; a thorough physical examination; and findings from a variety of laboratory tests and other studies. In making the diagnosis, it is important not only to confirm the presence of scleroderma, but also to determine its extent and severity, particularly with regard to the involvement of internal organs.

While scleroderma can often be suspected from its more visible symptoms, no single test can prove its presence.

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 scleroderma whereas anti-centromere is usually indicative of limited scleroderma.

	<u>Diffuse patients</u>	<u>Limited patients</u>
Anti-nuclear antibody98% test positive	98% test positive
Anti-centromere antibody3% test positive	41% test positive
Anti-Scl-70 antibody29% test positive	16% test positive

What are the symptoms of systemic scleroderma, and how are they treated?

This section describes the most common symptoms of scleroderma 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 number of symptoms and 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 all of the symptoms or all of the methods being used in the management of scleroderma. 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 is not curable, many of the symptoms can be improved with medication or lifestyle changes.

Even though scleroderma is not curable, many of the symptoms can be improved with medication or lifestyle changes.

Raynaud's phenomenon

Raynaud's phenomenon is the most common early symptom of systemic scleroderma. It is present at one time or another in about 90% 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's phenomenon, the blood vessels constrict or narrow in response to cold or to emotional upset and stress. The resulting disturbance in circulation of the blood 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 attack 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 persons experience Raynaud's attacks as painful.

Many common-sense preventive measures can be taken by those susceptible to Raynaud's phenomenon. Most obvious is minimizing exposure to cold, such as outdoor weather, air conditioning, or reaching into a refrigerator or freezer. Keeping warm is of equal importance, primarily the extremities, but also the entire body. Gloves or mittens should be worn, and a number of warming devices are available to protect the hands. Hat, ear muffs, heavy socks, and warm layered clothing of fibers such as silk, cotton, wool, and down are effective in maintaining body temperature. Protecting the hands with gloves when touching refrigerated or frozen items is important. Electric heaters, electric blankets, and comforters can supplement the heat in the home or apartment. Keeping the entire body warm helps prevent Raynaud's episodes.

A warm bath or shower, or a heating pad or hot water bottle on the back, may relieve an attack better than just warming the hands. Avoidance of emotional upset and stress is helpful but not always possible. Relaxation techniques of various kinds, whether self-taught or learned through training courses, have proven effective for some people in managing stress. One particular technique, biofeedback, has been used to increase finger temperature.

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

When Raynaud's 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's 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 simple aspirin, or drugs that decrease the stickiness of platelets, may improve circulation. There are now over a dozen medications to improve circulation, and it is not possible to list them here.

Raynaud's phenomenon is not confined to persons with scleroderma. It is also seen in lupus, rheumatoid arthritis, and other connective tissue diseases. In addition, many healthy people have Raynaud's phenomenon without any other illness. In this situation, it is called "primary Raynaud's disease."

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

Swelling or puffiness of the hands

Swelling is another typical early symptom of scleroderma, and this may be especially noticeable upon awakening because of muscle inactivity overnight. The skin of the fingers may look full and sausage-like, making it difficult to close the hand into a fist. Exercising the fingers and toes is helpful. The physician may recommend medications to reduce inflammation.

Pain and stiffness of the joints

Symptoms of pain, stiffness, swelling, warmth or tenderness may accompany the arthritis-like joint inflammation which frequently occurs in scleroderma. Muscle pain and weakness are other important symptoms. Aspirin or aspirin-like medications (non-steroidal, anti-inflammatory prescription drugs) can be helpful.

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 disorders

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

Skin thickening. Hardening and thickening of the skin give scleroderma its name (“hard skin”). There are no proven treatments as yet to prevent or alter the course of the skin changes in scleroderma. Many medications and treatments are being tested. The skin sometimes softens spontaneously over time.

Skin ulcerations. Sores, especially on the fingertips, are a common symptom of systemic scleroderma. They may be very slow or difficult 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 scrupulously clean to avoid infection. If infection should develop, it may be helpful to

soak the affected area in warm water, apply an antiseptic such as betadine, or use an antibiotic ointment, such as Neosporin™ or Bacitracin™. Should these remedies prove unsuccessful in relieving

the pain or infection of ulcerated skin, the physician may prescribe oral antibiotics or take other measures.

Calcinosis. This condition is characterized by deposits of calcium in the skin, which may be painful. The calcium deposits may occur just below the skin surface in the form of hard lumps or nodules. They may break through the skin, becoming visible as 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 cosmetics 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 such as Neutrogena® and Dove™ are preferable to harsh soaps which dry out the skin. Frequent use of moisturizing skin creams containing lanolin is advisable. A room humidifier may be helpful during the winter months.

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

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

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 perspire. In addition, there may be an increase in pigment (which looks like a skin tan) or a spotty loss of pigment.

Sclerodactyly and joint contractures

Sclerodactyly means simply “hard skin of the digits”; that is, of the fingers and toes. It generally occurs after initial swelling has subsided. It is characterized by shiny, tight 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 a “flexion contracture.” Tightening and hardening of the skin and of the tissues surrounding the joints can cause decreased motion of the wrists, elbows, and other joints.

“Range of motion” exercises performed daily are important in keeping the joints limber.

“Range of motion” exercises performed daily are important in preventing or slowing down the development of such contractures and in keeping the joints limber. 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 may also provide devices to help perform common personal care and household tasks more easily.

Digestive system and gastrointestinal tract problems

Persons 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 permit food to enter the stomach and then closes promptly to prevent food from coming back up. In systemic scleroderma, the gate does not close properly and the result is a backwash of acid and a burning sensation (“heartburn”) as food and acid return into the esophagus. The acid may also injure the lining of the lower portion of the esophagus, causing scarring and a narrowing (“stricture”) of the tube.

Acid production can be reduced, and the problems of acid reflux and heartburn alleviated, by avoiding (to the extent possible) alcohol, greasy or fatty foods, spicy foods, chocolate, tobacco, and caffeine. Antacids (particularly in liquid form) are helpful in neutralizing acids and reducing heartburn. Some antacids cause constipation

while others cause diarrhea. Consult your physician or pharmacist when choosing even over-the-counter products. The physician may prescribe antacid medications such as proton pump inhibitors or H-2 blockers to decrease acid production in the stomach. The physician may also prescribe a drug such as metoclopramide which promotes muscular activity and causes the esophagus to work better.

The force of gravity helps to keep food and acid in the stomach; therefore, an upright position 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 detrimental, and wearing girdles or other tight-fitting garments should be avoided.

To prevent acid reflux: eat smaller meals, don't eat before bedtime, and raise the head of the bed.

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.

Diarrhea. In systemic scleroderma, 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 may also 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. The physician may also suggest that the amount of fatty foods in the diet be reduced and the amount of carbohydrates increased.

Constipation. Weak or scarred muscles in the colon wall make it difficult for the bowel to work well, resulting in constipation or other

abnormalities of the colon. Maintaining a diet high in fiber, and drinking at least six to eight glasses of fluids daily, especially water, will help prevent constipation. Fresh fruits and vegetables are natural laxatives. Exercise also helps to keep bowel movements regular. The physician may also recommend stool softeners and bulking agents like Metamucil or Fiberall.

Sjögren's Syndrome

Sjögren's Syndrome (dry eyes, 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

Sjögren's
Syndrome is
characterized by
dry eyes and
mouth.

lead to difficulties in swallowing and in speaking, a pronounced increase in tooth decay and cavities, and a reduced sense of taste. The lack of secretions in Sjögren's Syndrome may also involve the vagina and other areas of the body.

Dry eyes may be lubricated by the frequent use of artificial tears and ophthalmic ointments. Regular visits to the ophthalmologist are important. The mouth should be kept as well-lubricated as possible by sipping fluids throughout the day (a plastic squirt bottle filled with water may be useful), 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.)

The physician 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

Persons with scleroderma may experience a general tightening of skin over the face. The opening of the mouth may be decreased in size ("microstomia" or "small mouth"), 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. In performing

these maneuvers, care should be taken to avoid further damage, especially to the jaw bone and teeth. Particular care is advisable in using augmentation devices. An evaluation by a dentist experienced in dealing with these problems is suggested prior to undertaking such exercises.

Preventive dental care through 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 can also recommend a program of good oral hygiene. 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's Syndrome.)

Kidney involvement

Kidney or renal involvement in systemic scleroderma 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 of systemic scleroderma, may occur quite suddenly. Its most important warning signal is an abrupt 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 in controlling blood pressure and in stabilizing and even improving kidney function. In cases of severe kidney failure, dialysis may be required. Persons with scleroderma are advised to have their blood pressure and kidney function monitored at regular intervals. People may recover successfully from renal crisis, but only if the problem is recognized and treated quickly.

Persons with scleroderma should have their blood pressure and kidney function monitored regularly.

Lung involvement

Lung involvement in systemic scleroderma may be caused by multiple factors. Build-up of collagen thickens lung tissue and causes fibrosis or scarring, making the transport of oxygen into the bloodstream more difficult. 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), or special breathing tests (pulmonary function tests) to detect or confirm lung involvement.

In the very early stages of lung fibrosis, medications may be given to decrease the inflammation which is thought to lead to lung scarring. This treatment is currently in the experimental phase.

Although many investigations are under way, there are no present proven medications to reverse lung changes once they have occurred. It is important, therefore, for the person with scleroderma 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. The physician may recommend medications to make breathing easier and may also suggest deep breathing exercises and a graduated aerobic exercise program.

Persons 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 one complication of scleroderma for which new medications have proven successful; there are now three different medications which can be used. Two prostacyclin drugs are available: epoprostenol (brand name Flolan™ is given through a continuous intravenous line; treprostinil (brand name Remodulin™) is given by a continuous infusion through a needle under the skin. Bosentan (brand name Tracleer™), an endothelin receptor antagonist, is given in pill form.

Heart involvement

If the heart muscle becomes thickened and fibrous scar tissue accumulates, the force of heart contractions may be decreased, which may ultimately result in heart failure. Spasm of the coronary arteries (the main blood vessels to the heart), may cause chest pain and, rarely, lead to a heart attack. The spasm appears similar to that involving the fingers in Raynaud's phenomenon. Inflammation

of the outer heart lining (pericarditis) may cause pain and accumulation of fluid around the heart. An irregular heartbeat may also be experienced. These conditions require careful evaluation and treatment by the physician.

Non-specific symptoms

The person with systemic scleroderma may experience a variety of non-specific symptoms, including fatigue (ranging from mild to severe), lack of energy, generalized weakness, weight loss, and 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

The reader may be aware of, or learn about, other forms of treatment that have been used or are proposed for use in managing 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. Under these circumstances, it is difficult to conduct scientifically sound studies proving the value of a particular drug or treatment. Therefore, the physician must often make decisions about treatment based on incomplete information. He/she must weigh the possible benefits against the potential risks or side effects. Further investigation will ultimately determine which treatments are beneficial and which treatments are not.

The course of scleroderma

Scleroderma has many forms and a number of different symptoms that may present themselves singly or in combinations at various times throughout the course of the disease. Some symptoms develop with relative suddenness; others take years to develop. The exact course the disease may take is unpredictable, and the prognosis will vary from individual to individual. Systemic scleroderma is a chronic, life-long disease. At present there is no known cure, but as with other chronic diseases there are many means available 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. Spontaneous remissions, times when symptoms may actually disappear, may also occur and may continue for long periods. The person with scleroderma should be cautious about attributing such improvements or remissions to a particular treatment, diet, or so-called "cure."

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 persons with scleroderma or those who suspect they may have the disease 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.

Early treatment
may prevent
symptoms from
worsening and
may decrease the
chance of
permanent tissue
or organ damage.

Learning to recognize early symptoms of disease activity can lead to earlier detection and diagnosis of scleroderma and to prompt initiation of treatment. Some of the more promising medications in current use are slow-acting and the sooner treatment is begun, the better the results may be. If one has already been diagnosed as having 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. Persons 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. The symptoms of scleroderma vary greatly from individual to individual; the manner in which each person responds to treatment also varies

greatly; and there are many treatment options. It is important, therefore, that a physician experienced in the management of scleroderma work 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 carried out at various locations, including a hospital physical therapy department and in the home.

The physician 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.

Physical therapists can help the person with scleroderma develop an appropriate program.

Protecting the joints

The goals of joint protection are to minimize further damage and to 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 their 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 to alleviate stress on the joints while carrying on the activities of daily living. Occupational therapists can demonstrate such devices and give further instruction on joint protection.

There are also a number of books, pamphlets, and cassettes that discuss how to apply the principles of joint protection, describe many

of the self-help aids and adaptive mechanical devices, and cover other topics of interest to the person with scleroderma. Two books, written especially for those with arthritis but very helpful to the person with scleroderma, are *The Arthritis Helpbook* by Kate Lorig and James F. Fries, and *Self-Help Manual for Patients with Arthritis* prepared by the Arthritis Health Professionals Section of the Arthritis Foundation.

Taking medications

It is essential that the person with scleroderma take all medications wisely; take only those prescribed; read label warnings and follow instructions carefully; and take the medications exactly when, for how long, and in the dosages prescribed by the physician. The person with scleroderma should advise the physician of any drugs being taken for other conditions including over-the-counter

preparations, herbal supplements, or vitamins.

Any side effects encountered should be promptly reported and discussed.

Scleroderma

symptoms vary from person to person, requiring different treatment.

One should not be concerned if the physician prescribes different medications for different persons. 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

Treatment of specific symptoms has been discussed in other sections of this booklet. There are also a number of general common-sense measures that the 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 habits of good hygiene, especially of the skin, teeth, gums, and feet (including the wearing of cushioned and well-fitted shoes).

One key measure, mentioned previously, is 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 persons develop the disease and others do not. One does not bring scleroderma upon himself or herself; therefore, one need not 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, or 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 be helpful. Professional counseling can also help persons with scleroderma and their family members who are having difficulty coping with their feelings.

The term “person with scleroderma” has been used throughout this booklet instead of “scleroderma patient.” 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.

Participating actively in one’s own healthcare is of prime importance to the person with scleroderma.

Building a health and support network

Participating actively in one’s own healthcare 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 persons and resources can be enlisted to 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.

Joining a support group allows you to meet and exchange information with others who have similar problems.

Directories of community resources typically list a large number of voluntary and governmental agencies providing health, social, and rehabilitation services that may be of benefit 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 to exchange information with others who have similar problems, as well as to learn more about scleroderma.

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 the person with scleroderma? Emphatically, yes!

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

Investigators throughout the United States and in 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 in improved methods of measurement to evaluate disease progression and the results of treatment. Various animal models of scleroderma have been developed.

Investigators are currently 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!

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 over \$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 persons with scleroderma and their families. The Foundation’s services include

- a toll-free telephone Info Line, 800-722-HOPE (4673);
- an email address for patient questions, sfinfo@scleroderma.org;
- a website with news and information, 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 persons with scleroderma in contact with each other.

At the national and international levels, the Scleroderma Foundation supports persons with scleroderma and their families in

many ways. The Foundation increases public and professional awareness of scleroderma through national publicity campaigns. It acts as a clearinghouse for the exchange and dissemination of information about scleroderma. It produces educational literature (such as this booklet) which is widely distributed. 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.

Auto-immune. Disease or antibody which acts against the patient'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's 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's phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia.

Cutaneous. Of the skin.

Cyanosis. Blue or purple color due to lack of blood oxygen. In Raynaud's 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 auto-immunity.

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. A form of 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's phenomenon. Also called Raynaud's 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 "Ray-node") 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's 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's.")

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.

Books

All books on this list, and many informative brochures on specific topics related to scleroderma, may be purchased from the Scleroderma Foundation by calling toll-free 1-800-722-HOPE (4673).

The Scleroderma Book

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

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

The Best of the Beacon

Edited by Marie Coyle

A marvelous collection of practical and inspirational articles for those living with scleroderma.

Perspectives on Living with Scleroderma

By Mark Flapan, Ph.D.

Dr. Flapan's insightful articles on coping, including his personal story.

Scleroderma: A New Role for Patients and Families

By Michael Brown

Offers resources to help you take charge.

Scleroderma: Surviving a 17-Year Itch

By Dana Lovvorn

Self-help book for newly diagnosed patients, emphasizing exercise and lifestyle adaptations.

Successful Living with Scleroderma: Guidebook

By Robert Phillips, Ph.D.

Video

Living Well With Scleroderma

Created by patients for patients, this video is intended for newly diagnosed patients and their families. Inspirational and informative. Available for purchase from the Scleroderma Foundation; can be watched free of charge by visiting the Foundation's website (below).

Websites

Scleroderma Foundation

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

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

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

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



Scleroderma Foundation
12 Kent Way, Suite 101
Byfield, MA 01922
Info Line 800-722-HOPE (4673)
Email sfinfo@scleroderma.org
Website www.scleroderma.org