MEDICAL EDUCATION SYSTEMS, Inc.

Scleroderma: An Overview

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Scleroderma: an Overview

Learning Objectives

- Define what is meant by the term scleroderma
- Indicate how scleroderma is diagnosed
- Indicate how scleroderma is treated
- Indicate what causes scleroderma

Introduction

This course is an overview and has drawn from several sources. As a result there may some repletion or duplication of information.

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.

What scleroderma is not

Scleroderma is not contagious, it is not infectious, it is not cancerous or malignant.

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.

How is scleroderma diagnosed?

The diagnostic process may require consultation with rheumatologists (arthritis specialists), and/or dermatologists (skin specialists) and require blood studies and numerous other specialized tests depending upon which organs are affected.

Who develops scleroderma, and when?

There are an estimated 300,000 people in the United States who have scleroderma, about one third of whom have the systemic form of scleroderma. Since scleroderma presents with symptoms similar to other autoimmune diseases, diagnosis is difficult and there may be many misdiagnosed or undiagnosed cases as well.

Localized scleroderma is more common in children, whereas systemic scleroderma is more common in adults. Overall female patients outnumber male patients about four to one, and the average age at diagnosis is in the forties.

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.

However, 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. When doctors say "usually" or "for the most part," the reader should understand that variations frequently occur. Many patients get alarmed when they read medical information that seems to contradict their own experiences, and conclude that what has happened to them is not supposed to happen. There are many exceptions to the rules in scleroderma, perhaps more so than in other diseases. Each case is different, and information should be discussed with your own doctor.

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. It is known that scleroderma involves overproduction of collagen.

Is scleroderma genetic?

Most patients do not have any relatives with scleroderma and their children do not get scleroderma. Research indicates that there is a susceptibility gene which raises the likelihood of getting scleroderma, but by itself does not cause the disease.

What is the treatment for scleroderma?

At the present time, there is no cure for scleroderma, but there are many treatments available. Some are directed at particular symptoms like heartburn, which can be controlled by medications called proton pump inhibitors or medicine to improve the motion of the bowel. Some treatments are directed at decreasing the activity of the immune system. Some people with mild disease may not need medication at all and occasionally people can go off treatment when their scleroderma is no longer active. Because there is so much variation from one person to another there is great variation in the treatments prescribed.

What is scleroderma?

Scleroderma is an autoimmune disease of the connective tissue. Autoimmune diseases are illnesses which occur when the body's tissues are attacked by its own immune system. Scleroderma is characterized by the formation of scar tissue (fibrosis) in the skin and organs of the body. This leads to thickness and firmness of involved areas. Scleroderma, when it's diffuse or widespread over the body, is also referred to as systemic sclerosis.

The cause of scleroderma is not known. Researchers have found some evidence that genes are important factors, but the environment seems to also play a role. The result is activation of the immune system, causing injury to tissues that result in injury similar to scar tissue formation. The fact that genes seem to cause a predisposition to developing scleroderma means that inheritance at least plays a partial role. It is not unusual to find other autoimmune diseases in families of scleroderma patients. Some evidence for the role genes may play in leading to the development of scleroderma comes from the study of Choctaw Native Americans who are the group with the highest reported prevalence of the disease. The disease is more frequent in females than in males.

How is scleroderma classified?

Scleroderma can be classified in terms of the degree and location of the skin involvement. Accordingly, scleroderma has been categorized into two major groups, diffuse and limited.

The diffuse form of scleroderma (systemic sclerosis) involves symmetric thickening of skin of the extremities, face, and trunk (chest, back, abdomen, or flanks) which can rapidly progress to hardening after an early inflammatory phase. Organ disease can occur early on and be serious. Organs affected include the esophagus, bowels, lungs with scarring (fibrosis), heart, and kidneys. High blood pressure can be a troublesome side effect.

The limited form of scleroderma tends to be confined to the skin of the fingers and face. The skin changes and other features of disease tend to occur more slowly than in the diffuse form. Because a characteristic clinical pattern can occur in patients with the limited form of scleroderma, this form has taken another name which is composed of the first initials of the common components. Thus, this form is also called the CREST variant of scleroderma. This name represents the following features:

C...Calcinosis refers to the formation of tiny deposits of calcium in the skin. This is seen as hard whitish areas in the superficial skin, commonly overlying the elbows, knees, or fingers. These firm deposits can be tender, can become infected, and can fall off spontaneously or require surgical removal. This is the least common of the CREST scleroderma variant features.

R...Raynaud's phenomenon refers to the spasm of the tiny artery vessels supplying blood to the fingers, toes, nose, tongue, or ears.

These areas turns blue, white, then red after exposure to extremes of cold, or even sometimes with extremes of heat or emotional upset.

E...Esophagus disease in scleroderma is characterized by poorly functioning muscle of the lower two-thirds of the esophagus. This can lead to an abnormally wide esophagus which allows stomach acid to backflow into the esophagus to cause heartburn, inflammation, and potentially scarring. This can eventually lead to difficulty in passing food from the mouth through the esophagus into the stomach. Symptoms of heartburn are treated aggressively in patients with scleroderma in order to prevent injury to the esophagus.

S...Sclerodactyly refers to the localized thickening and tightness of the skin of the fingers or toes. This can give them a "shiny" and slightly puffy appearance. The tightness can cause severe limitation of motion of the fingers and toes. These skin changes generally progress much slower that those of patients with the diffuse form of scleroderma.

T...Telangiectasias are tiny red areas, frequently on the face, hands and in the mouth behind the lips. These areas blanch when they are pressed upon and represent dilated capillaries.

Patients can have variations of CREST, for example, CRST, REST, ST, etc. Patients can also have "overlap" illness with features of both CREST and the diffuse form of scleroderma. Some patients have overlaps of scleroderma and other connective tissue diseases, such as rheumatoid arthritis, systemic lupus erythematosus, and polymyositis. When features of scleroderma are present along with features of polymyositis and systemic lupus erythematosus, the condition is referred to as mixed connective tissue disease (MCTD).

Finally, scleroderma skin changes can be very localized. Morphea is scleroderma skin that is localized to a patchy area of the skin that becomes hardened and slightly pigmented. Sometimes morphea can cause multiple lesions in the skin. Morphea is not associated with disease elsewhere in the body. Linear scleroderma is scleroderma that is localized usually to a lower extremity, frequently presenting as a strip of hardening skin down the leg of a child. Linear scleroderma in children can stunt bone growth of the affected limb. Sometimes linear scleroderma is associated with a "satellite" area of a patch of localized scleroderma skin, such as on the abdomen.

What are symptoms of scleroderma?

The symptoms of scleroderma depend on the type of scleroderma present and the extent of external and internal involvement in the individual affected. Because scleroderma can affect the skin, esophagus, blood vessels, kidneys, lungs, blood pressure and bowels, the symptoms it causes can involve many areas of the body.

Scleroderma affects the skin to cause local or widespread signs of inflammation (redness, swelling, tenderness, itching, and pain) that can lead to skin tightness or hardening. These skin changes can be widespread, but it's most common for them to affect the fingers, feet, face, and neck. This can lead to decreased range of motion of the fingers, toes, and jaw. Tiny areas of calcification (calcinosis), while not common, can sometimes be noticed as hard nodules at the tips of the elbows or in the fingers.

Scleroderma affecting the esophagus leads to heartburn. This is directly a result of stomach acid flowing back up into the esophagus. Sometimes this can lead to scarring of the esophagus with difficulty swallowing and/or localized pain in the central chest.

Blood vessels that can be affected include the tiny arterioles of the finger tips, toes, and elsewhere. These vessels can have a tendency to spasm when the areas are exposed to cold, leading to blueness, whiteness, and redness of involved fingers, toes, and sometimes nose or ears. These color changes are referred to as Raynaud's phenomenon. Raynaud's phenomenon can cause inadequate supply of oxygen to the involve tips of fingers or toes, causing tiny ulcers or blackened (dead) skin. Sometimes Raynaud's phenomenon is also associated with tingling. Other blood vessels that can be involved in scleroderma are the tiny capillaries of the face, lips, mouth, or fingers. These capillaries widen (dilate) forming tiny, red blanching spots, called telangiectasias.

Elevated blood pressure is potentially serious and can lead to kidney damage. Symptoms include headache, fatigue, and in severe cases, stroke.

Inflammation of the lungs in scleroderma can cause scarring, resulting in shortness of breath, especially with physical exertion. Elevated pressure in the arteries to the lungs (pulmonary hypertension) can also cause shortness of breath and difficulty getting an adequate breath with activity.

Scleroderma affecting the large bowel (colon) most often causes constipation but can also lead to cramping and diarrhea. When this is severe, it complete stool blockage (fecal impaction) can result.

How is scleroderma diagnosed?

The diagnosis of the scleroderma syndrome is based on the finding of the clinical features of the illnesses. Nearly all patients with scleroderma have blood tests which suggest autoimmunity, antinuclear antibodies (ANAs). A particular antibody, the anticentromere antibody, is found almost exclusively in the limited, or CREST, form of scleroderma. Anti-Scl 70 antibody (antitopoisomerase I antibody) is most often seen in patients with the diffuse form of scleroderma.

Other tests are used to evaluate the presence or extent of any internal disease. These may include upper and lower gastrointestinal tests to evaluate the bowels, chest x-rays, lung function testing, and CAT scanning to examine the lungs, EKG and echocardiograms, and sometimes heart catheterization to evaluate the pressure in the arteries of the heart and lungs.

How is scleroderma treated?

Treatment of scleroderma is directed toward the individual feature(s) affecting different areas of the body.

Aggressive treatment of elevations in blood pressure have been extremely important in prevent kidney failure. Blood-pressure medications, such as captopril, are frequently used.

Recent data indicate that colchicine can be helpful in decreasing the inflammation and tenderness that periodically accompanies the calcinosis nodules in the skin. Skin itching can be relieved with lotions (emollients) such as Eucerin and Lubriderm.

Mild Raynaud's phenomenon may require only hand warming and protection. Low-dose aspirin is often added to prevent tiny blood clots in the fingers, especially in patients with a history of fingertip ulcerations. Moderate Raynaud's phenomenon can be helped by medications that open up the arteries, such as nifedipine (Procardia, Adalat) and nicardipine (Cardene), or with topical nitroglycerin applied to the most affected digit (most effective on the sides of the digit where the arteries are). Gently applied finger splinting can protect tender tissues. A class of medications that is typically used for depression, called serotonin reuptake inhibitors, such as fluoxetine (Prozac), can sometimes improve the circulation of the affected digit. Severe Raynaud's phenomenon can require surgical procedures, such as those to interrupt the nerves of the finger that stimulate constriction of the blood vessels (digital sympathectomy). Ulcerations of the fingers can require topical or oral antibiotics.

Esophagus irritation and heartburn can be relieved with omeprazole (Prilosec), esomeprazole (Nexium), or lansoprazole (Prevacid). Antacids can also be helpful. Elevating the head of the bed can reduce the back-flow of acid into the esophagus that causes inflammation and heartburn. Avoiding caffeine and cigarette smoking also helps.

Constipation, cramping, and diarrhea is sometimes caused by bacteria that can be treated with tetracycline or erythromycin. Recent studies have shown that erythromycin could also be used. Increased fluid intake and fiber intake are good general measures.

Irritated, itchy dry skin can be helped by emollients such as Lubriderm, Eucerin, or Bagbalm.

Telangiectasias, such as those on the face, can be treated with local laser therapy. Sun exposure should be minimized as it can worsen telangiectasias.

Approximately 10% of patients with the CREST variant develop elevated pressures in the blood vessels to the lungs (pulmonary hypertension). Abnormally elevated blood pressure of the arteries supplying the lungs is often treated with calcium antagonist medications, such as nifedipine, and blood-thinning drugs (anticoagulation). More severe pulmonary hypertension can be helped by continuous intravenous infusion of prostacyclin (Iloprost). A new drug taken by mouth, bosentan (Tracleer), is now available to treat severe pulmonary hypertension.

Additionally, medications are used to suppress the overly active immune system that seems to be spontaneously causing the disease in organs affected. Medications used for this purpose include penicillamine, azathioprine, and methotrexate. (Recent research has found that low-dose penicillamine [Depen, Cuprimine] [125mg every other day] is as effective as previously used high doses of penicillamine, with less toxicity.) Serious inflammation of the lungs (alveolitis) can require immune suppression with cyclophosphamide (Cytoxan) along with prednisone. The optimal treatment of scleroderma lung disease is an area of active research. Stem-cell transplantation is being explored as a possible option.

No medication has been found to be universally effective for all patients with scleroderma. In an individual patient, the illness may be mild and not require treatments. In some, the disease is ravaging and relentless.

What is the outlook (prognosis) for patients with scleroderma?

A patient's prognosis is optimized with close monitoring of overall health status and treatment of complications, especially elevated blood pressure. Recent data indicates that the critical period of organ risk is generally within the first three years of skin involvement. This means that patients can be reassured that their risk of organ-threatening complications is significantly less after three years of having skin symptoms.

Much more research is needed in all areas of scleroderma disease, from cause to treatment. Today scleroderma continues to baffle medical scientists. Researchers are evaluating the effectiveness of thalidomide for the treatment of scleroderma. More sensitive tests to detect early lung disease of scleroderma are also being evaluated. Psoralen and ultraviolet light therapy (PUVA) is being studied as a possible treatment for limited scleroderma.

Many researchers are investigating the roles of various cell messengers, called cytokines, in causing scleroderma. Researchers are also currently studying a hormone of pregnancy, called relaxin, for the treatment of scleroderma. Preliminary results suggest that it may improve scleroderma. Relaxin normally loosens the ligaments of the pelvis and ripens the womb for childbirth. How it might work in scleroderma is unclear.

Scleroderma At A Glance

Scleroderma is a skin disease that is associated with illness involving internal organs.
Scleroderma is classified into diffuse and limited forms.
CREST syndrome is a limited form of scleroderma.
Patients with scleroderma can have antibodies in their blood which suggest autoimmunity.
Treatment of scleroderma is directed toward the individual feature(s) that is(are) most troubling.

References:

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Kelley's Textbook of Rheumatology, W B Saunders Co, edited by Shaun Ruddy, et al., 2000.

Scleroderma - Describe Your Experience

The MedicineNet physician editors ask:
Comment from: kelticlady, 45-54 Female (Patient)

I am 49 years old right now. I was diagnosed with scleroderma in 1992. Two years later, I was in the hospital with congestive heart failure. I really have not progressed much since then. I do have trouble with my hands and arms. They do not straighten out very well. I take a series of medicines: Cuprimine, Captopril, and Diltiazem. I do suffer emotionally and from depression. That has been the worst for me. I do feel lucky to be here today, and I try to be thankful for the time I have been given. I always get my rest. Published: November 29 ::

Comment from: slick1, 55-64 Female (Patient)

In 1999, I was diagnosed with eosinophilia fasciitis. My doctor had me taking Remicade, which worked very well until I started having liver problems. I had a biopsy performed on the muscle, which showed that I had systemic scleroderma. I am now taking Cellcept. I have been taking it for only four weeks, but already my skin is looking better, and I am being told by friends and family that I sound and look as if I feel better. The Cellcept makes me very tired, but we will know more after it completely gets into my system. Published: November 18 ::

Comment from: 45-54 Female (Patient)

I too was diagnosed with systemic scleroderma. It started with Raynaud’s, since then have developed gastro intestinal problems due to the muscles in my esophagus not working anymore. I am undergoing testing for lung hypertension. I also have thyroid problems that have been taking med. for about 25 years. I also have been diagnosed with fibromyalgia. New research is suggesting that hormones play a big part in disease progression. I read where relaxin hormone slows the disease process. This is a hormone found in pregnant women and relaxes the muscles in turn keeps fibrosis at bay. I haven't been given anything that works yet. I can't take much medication due to lung involvement. I have found that sunshine and as much activity as I can muster keeps acute symptoms down. Good Luck and God Bless to all that suffer this crazy disease. I am 54 yrs old. Published: August 04 ::

Comment from: OBF1983, 25-34 Female (Patient)

I am 25 years old and was diagnosed with scleroderma in May of 2008, although my symptoms had started probably a year or so before that. When I first had trouble, I went to my GP, and she told me to cut back on salt and lose weight. Three times I went back to the doctor and got the same response, even though her advice wasn't helping. I was finally referred to a rheumatologist who ran numerous tests that all came back normal. When my skin began to thicken, and I developed ulcers on my knuckles, she sent me to a dermatologist. There, they did a skin biopsy and diagnosed me with scleroderma. It has been a really rough year. I miss a lot of work, I am in constant pain, and I just feel down a lot of the time. Published: November 29 ::

Comment from: Erin, 19-24 Female (Patient)

I was diagnosed with scleroderma when I was in elementary school. I started developing skin patches on my right calf, then my right thigh, my right buttock, and then the right side of my ribs. It had been itchy, and after a while, I lost fat in those areas with a large amount of discoloration. Being that I was only in elementary school, I grew up being very self-conscious and insecure about my scars. I, of course, had other problems, such as gastrointestinal ones, and I still do. But thankfully, the disease hasn't spread further or affected my internal organs. I am now 20 years old, and while I would like to say it has made me stronger, I still feel very insecure and unsure of what to tell those who see my scars and think they are bruises or burns. It could be a lot worse, but the external scars are so damaging to my ego. I am considering plastic surgery to see if they can't help fix the lesions. Published: November 18 ::

Comment from: JP, 45-54 Female (Patient)

I was diagnosed with CREST Syndrome in 1985. I have had many different complications. Leg ulcers have been my No. 1 problem. I have been doing well, and all my medications have been decreased until just this past week. I have been experiencing shortness of breath. I went to my regular physician and they checked my heart and pulmonary functions. I just went to my regular appointment for my rheumatoid arthritis (RA) and had taken along copies of all my tests. My doctor said they were looking at me like a regular person with shortness of breath and not a scleroderma patient. He increased my medication, and I am feeling better and hoping I didn't wait too long and have damaged my lungs. I just wanted others to be aware and not hesitate to call the doctor they see regularly for their scleroderma when you have serious symptoms. Published: November 18 ::

Comment from: Adelene, 25-34 Female (Patient)

Last year, I was infected with scleroderma. It mostly affected my arms and chest. The skin on the infected areas is red. I had an uneven skin tone. My finger and toe tips would turn purplish when cold. I even had difficulty in stretching my arms straight. It was very troublesome for me, as people had been questioning about my abnormal looking skin. So, I had been wearing long-sleeve clothes to cover up myself most of the time. I had gone for traditional, medical and nutritional healings. From that day forward, I changed to a healthier lifestyle. Besides, taking medicines by doctors, I began to take lots of nutritional food and supplements, such as fruits and vegetable enzymes, strath herbal yeast elixir, vitamin E, etc. I even took up tai chi lessons to keep myself fit.

I applied virgin coconut oil and massage my arms every morning and night. I pray hard to God for healing day and night. Gradually, my skin condition began to improve. I realized that I have became a more optimistic and strong person. Although I have now fully recovered, I still continue to live a healthy lifestyle. I feel more confident with my looks now. I had faced lots of difficulties when having scleroderma, but it has made me a tough person now. I would also like to encourage people who are having such sickness not to give up easily. There is always hope.

Comment from: csr, 55-64 Female (Patient)

I have scleroderma on my left lower leg. It started as a small patch after knee replacement and has gradually increased in size. It has been a most difficult situation but one I face head on. I refuse to let it keep me from enjoying my life. I have a small area on my right leg that is also beginning to form. I truly have not been advised as to anything I can do. I do believe that my esophagus is also involved, but was not aware until visiting your sight. Every time I have a flare, these areas become very inflamed. Published: October 24 ::

Comment from: Wilhelmina, 35-44 Female (Patient)

Eight years ago I was diagnosed with scleroderma, I was never ill never even had a real flu, then suddenly my fingers started to get really sensitive, I couldn't tie my shoe laces. I went to a specialist and he found out I had scleroderma. I thanked him for the medication and left, not knowing what to do next. I went looking for a licensed doctor who was also a naturopath, he did a very simple hair analysis and we found out I had too much, mercury, lead, silver arsenicum and tin in my body. After a detox of three months with DMSA pills and removing my amalgam my scleroderma stabilized. My fingers and my toes are a bit stiff but for the rest I can do everything. I hope I can help other people with my story. Published: October 23 ::

Comment from: Brenna, 35-44 Male (Caregiver)

My dad has scleroderma, and it has attacked his kidneys. My auntie just died from scleroderma a year ago. The doctor who diagnosed my dad was shocked to hear that his sister also had the disease because it is rare to be in the same family, such as brother and sister. Seeing this disease attack my dad so drastically puts me in shock because my dad was always so healthy and hardly ever got sick. The physical effects it has had on my dad are: loss of muscle and weight, hardening and tightening of the skin, joint pain, struggling to breathe, blotches on his skin from kidney problems, purple hands and feet, and a huge change in diet to survive better and almost daily blood tests. Published: September 30 ::

Comment from: Mary, 55-64 Female (Patient)

I have had scleroderma since I was 12 years old. I am now a 59-year-old female. It started with a small, white patch underneath my right eye. Over the 47 years, it has progressed down the right side of my face, my arm, leg, back, and stomach. I always seem to be able to tell when it spreads a little more. It's as though it has become live and itches. Published: September 30 ::

Comment from: Annie, 35-44 Female (Caregiver)

Hi, I'm from the Philippines. My 44 year old female cousin (from my mother's side) just died last week of scleroderma. She was diagnosed in November last year. The hardening of her skin and its spread throughout her body was fast and I guess the doctors here have no idea as to mode of treatment. Published: September 25 ::

Comment from: ripeberry4, 65-74 Female (Caregiver)

My mother has scleroderma and she has had two fingers cut off is this usually what happens in the later stages Published: September 25 ::

Comment from: Lyndsey, 25-34 Female (Patient)

I am 26-year-old white female who was diagnosed with UMCTD with a leaning toward scleroderma because my first symptoms were finger swelling along with Raynaud's disease. I spent a year in pain before I saw a doctor, and initially the doctors said my disease was progressing rapidly. I had a finger ulcer and had two pulmonary effusions, besides the debilitation of joint pain. I wanted to write however, to let people know that it is possible to deal with this disease-and to find the right doctor. My doctor has made it possible for me to stay off as many medications as possible and to give me back my life. It has been almost a year since I have had any serious complications with my disease, and I am working again and working on a graduate degree. I know that there is always a possibility of it getting worse, but all I prayed for was for it to be manageable, and I believe that if you get the right doctor and take an active part in your healing that it is ... manageable. Published: August 29 ::

Comment from: Joan, 65-74 Female (Patient)

I got Raynaud’s disease in 2001, and now I have the lung disease scleroderma. I’m getting the chemotherapy drug Cytoxan. I will get this until December, and then I will be tested again. If it doesn’t work I will be going to Boston for an experimental drug. I have been on oxygen for a year and also have been on different drugs. Published: August 29 ::

Expert Answers About Scleroderma

By THE NEW YORK TIMES

Bryce Vickmark for The New York Times Marie Coyle, one of the founders of the Scleroderma Foundation, was 12 when she first noticed a hardened patch on her scalp, which turned out to be scleroderma; she is among those featured in Patient Voices: Scleroderma.

Does scleroderma, the mysterious disorder that causes thickening of the skin and other problems, run in families? Is it related to other diseases? How long can you live with scleroderma? Those are among the questions posed by readers of the Consults blog. This week, two experts joined Consults to answer readers' questions: Dr. John Varga, of Northwestern University Feinberg School of Medicine, and Dr. Virginia Steen, of Georgetown University. Both doctors also serve on the advisory board of the Scleroderma Foundation.

Is Scleroderma Inherited?

Q.

My daughter's paternal grandmother and her father's niece died of scleroderma. She is 48 years old — should she be tested? What are the odds of her inheriting this disease? (And where would she go to be tested?)
Edwina, Copiague, N.Y.

Q.

Does scleroderma run in families?
Jake, Charlotte, N.C.

A.

Dr. John Varga responds:

Scleroderma rarely runs in families. For every 100 families with a member who has scleroderma, only two families will have another family member with the disease. So the relative risk of a family member developing scleroderma is very small, although it is higher than in the general population.

But the odds of inheriting scleroderma remain extremely small. Furthermore, there is currently no predictive test that can identify someone with scleroderma. Only if a child develops symptoms, such as the blue fingers of Raynaud's phenomenon or puffy fingers and tight skin, should he or she be evaluated for possible scleroderma.

Q.

How does someone get scleroderma? Is it hereditary?
Ali, New York, N.Y.

A.

Dr. John Varga responds:

The cause of scleroderma is unknown. Viruses and exposure to certain chemicals and occupational toxins have been suspected, but nothing has been firmly proven to cause the disease.

To elaborate on what I said above, scleroderma is, strictly speaking, not a hereditary disease. However, inheriting certain groups of genes appears to make someone more susceptible to developing the disease. The identity of these susceptibility genes is not yet known, but ongoing research is narrowing down the list of possibilities.

Is Scleroderma Related to Other Autoimmune Disorders?

Q.

How is scleroderma related to other autoimmune diseases?
AC, Princeton, N.J.

A.

Dr. John Varga responds:

Scleroderma is an autoimmune disease. Virtually all patients with scleroderma have proteins in their blood called autoantibodies. Healthy individuals also have antibodies, but they are directed at foreign microbes, thereby helping fight off infections. But in patients with scleroderma, as in other autoimmune diseases, the antibodies are directed toward the individual's own tissue, and they can cause tissue damage.

The risk of developing another autoimmune disease is increased if you have scleroderma. However, this is still an uncommon occurrence. Nevertheless, your rheumatologist should be — and presumably is — monitoring you and watching out for signs of another autoimmune disease developing.

Ovarian Cancer and Scleroderma?

Q.

My sister had scleroderma from the age of 4 until she died at age 57 of ovarian cancer; no one else in our family has had it. She also had Raynaud's disease. Is there any relationship between cancer and scleroderma?

A.

Dr. Virginia Steen responds:

There is no direct causal relationship of cancer and scleroderma, although there may be an increased association with lung cancer in patients who have chronic lung fibrosis (scarring) and breast cancer around the time of the diagnosis of scleroderma. Patients with longstanding esophageal problems can be at higher risk for esophageal cancer and Barrett's esophagus, a precursor to cancer, but studies have not shown a significant increase in this type of cancer.

Raynaud's and Scleroderma?

Q.

What about Raynaud's and scleroderma?
Raven, Greenville, N.C.

A.

Dr. Virginia Steen responds:

Raynaud's disease is an integral part of scleroderma. Almost all patients have the cold fingers of Raynaud's, often for years before other problems or the diagnosis of scleroderma.

To learn more about Raynaud's and scleroderma, see The Times Health Guide: Raynaud's Disease.

Dermatomyositis and Scleroderma?

Q.

What are the latest research and treatment methods for adults with dermatomyositis?
W., Rock Creek, Ala.

A.

Dr. Virginia Steen responds:

Dermatomyositis is another autoimmune disease, so it's in the same family as scleroderma. Some patients with scleroderma have some kind of muscle problems, though the problem usually is not dermatomyositis. An exciting clinical trial has just been completed that looked at the drug Rituxan in the treatment of dermatomyositis, so hopefully we will have the results later this year.

Can Foods Combat Scleroderma?

Q.

The father of a good friend of my daughter has this terrible disease. He was diagnosed over 10 years ago and given a life expectancy of about five years.

Against all odds and doctors' expectations, he has continued to live. At this stage of his disease, his lungs are becoming incapable of breathing without the assistance of oxygen at night. The drugs to assist this aspect of his disease cause incredible spikes in his blood sugar (he is also diabetic), so that his treatment options are becoming increasingly limited, as the balancing act for treatment is going off kilter — that is, the treatment is beginning to kill him.

One thing that confounds me is my friend's diet. He eats ONLY fast food items, and his diet is to me absolutely atrocious. He's like the author of "Super Size Me," except he didn't just eat McDonald's for a month, he's been eating a chain food diet of what is, in essence, fat and salt for years. Isn't there a resource that could help him develop a diet that might arrest his malady? Thanks!
Craig, Newport Beach, Calif.

A.

Dr. Virginia Steen responds:

There is no diet or foods that can successfully arrest or treat scleroderma. But yes, sometimes the treatment is difficult because of toxicity. Treating the diabetes is also very important. The drugs that aggravate the diabetes are usually corticosteroids. In general, we try to avoid high doses of these in patients with the lung disease of Scleroderma.

How Long Can You Live With Scleroderma?

Q.

My mother died of scleroderma many years ago. At the time, we were told that this is a disease that tends to take a chronic course, say, over 20 to 25 years. Her disease expressed itself in a very rapid form, and she died within a little more than two years. Is there a reason that this happened? Thank you.
Phoenix, California

Q.

My brother died at age 57 of heart failure which was probably connected to scleroderma which he was diagnosed with within the last five years, probably systemic (diffuse form). Based on the studies I examined, systemic scleroderma appears to be fatal with high mortality, especially within 5 or 10 years of diagnosis. Those afflicted with systemic scleroderma need to be told the truth and not given false hope. Treatments barely contain the symptoms and the disease continues to progress. Why are some doctors unwilling to be direct about telling the patient that they will probably die sooner than later?
Anonymous

A.

Dr. Virginia Steen responds:

Scleroderma patients have an extremely wide spectrum of manifestations of the disease. Over half of patients — about 53 percent — have very mild disease and die of causes not related to the scleroderma. In many of the other patients, it is often hard to determine how long someone will live.

There are different subsets of the disease, and survival depends on what type of disease you have and how much damage occurs to the internal organs. Yes, scleroderma can be a deadly disease, and doctors should try to be honest with their patients. But many times we can not predict events for an individual patient.

Testing for Scleroderma?

Q.

I know there are a variety of autoantibodies which can be found in scleroderma. Should patients with scleroderma be tested for all of these antibodies? Why, or why not?

I've been tested for anti-Scl-70 and anticentromere, and both are positive, which I know is unusual. However, would it be of interest to know if I or other patients also carry the other autoantibodies (anti-Th/To, PM/Scl, U1-RNP, U3-RNP, RNA polymerase III)?

Clinical info: I face the complications of interstitial lung disease, absent esophageal peristalsis, diffuse skin disease, arthritis and recently rising pulmonary artery pressures (and Raynaud's, skin pigment changes, etc.) after just three years of symptoms and despite very aggressive treatment.
Holly, San Diego

A.

Dr. Virginia Steen responds:

You are right that there are a variety of antibodies in scleroderma, and these are often helpful in predicting the course of the disease. Not all the antibody tests are commercially available, but yes, all scleroderma patients should have the ones that are available (Scl-70, anticentromere, U1-RNP, RNA polymerase III and PM/Scl).

There are also other antibodies, which appear as a specific type of pattern (known as a nucleolar pattern) when the standard immunofluorescent anti-nuclear antibody, or ANA, test is performed. The ANA is the screening test for many autoimmune diseases, and it is almost always positive in scleroderma, though some of the newer ANA methods result in negative test results. Unfortunately, the commercial laboratories have changed the way they do the antibody tests, and we get many false negative ANAs — that is, many tests come back negative even though the patient has the antibody. There are also quite a few false positive tests for the specific antibodies — patients test positive, though the antibody isn't actually present.

It is very unusual to have more than one antibody. The fact that you tested positive for two could be a lab error or misinterpretation of the tests. Rapidly progressive disease and interstitial lung involvement classically is associated with Scl-70 as opposed to anticentromere antibodies.

Stem Cell Transplants and Scleroderma

Q.

I have read about using stem cell transplants as a way of "replacing" the immune system that is infected by scleroderma. It seems this is only being tried in cases of advanced systemic scleroderma. Is anyone looking at whether a stem cell transplant could stop the disease in its early stages? And what are your thoughts about what is commonly known as AP (antibiotic protocol) therapy for scleroderma?
Marcy, Ohio

A.

Dr. Virginia Steen responds:

Stem cell transplant is being carefully studied as a treatment for severe *early* disease. The goal is to prevent the progression of the disease. Although there are very promising effects on the severe skin manifestations, the variability in the natural course of the disease makes it necessary to do a

careful comparison with other, more standard treatments. Information about the United States study, the SCOT trial, can be found on ClinicalTrials.gov.

The antibiotic protocol has not been adequately studied in scleroderma. Although there are many "testimonials" to its effectiveness, there has been only one study of 50 patients who were followed closely on it for a year, and there was no evidence of any effectiveness.

Naltrexone for Scleroderma?

Q.

People are having tremendous success with low-dose naltrexone. This information needs to get to people A.S.A.P. Google it and see for yourself. No time should be wasted — people's lives are too precious to keep them on wild goose chases with toxic drugs and other snake oil.
redplanet, California

A.

Dr. Virginia Steen responds:

Although low-dose naltrexone has recently been proposed as an effective treatment for a range of autoimmune diseases, experience with the drug has been limited. There is one published study, and one ongoing trial, of naltrexone for multiple sclerosis. Like the antibiotic protocol, its purported "effectiveness" is based on patient testimonials.

Hopefully, there will be more research with this drug to see its effect on autoimmune diseases. We clearly would love to have a drug that helps 100 percent of patients, as their Web site claims.

Handout on Health: Scleroderma

This booklet is for people who have scleroderma, as well as for their family members, friends, and others who want to find out more about the disease. This booklet describes the different forms of scleroderma and provides information on their symptoms, diagnosis, and treatment, including what patients can do to help manage their disease and the problems associated with it. This booklet also highlights research efforts into the understanding and treatment of scleroderma, many of which are supported by the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) and other components of the U.S. Department of Health and Human Services' National Institutes of Health (NIH). If you have further questions after reading this booklet, you may wish to discuss them with your doctor.

What Is Scleroderma?

Derived from the Greek words "sklerosis," meaning hardness, and "derma," meaning skin, scleroderma literally means hard skin.

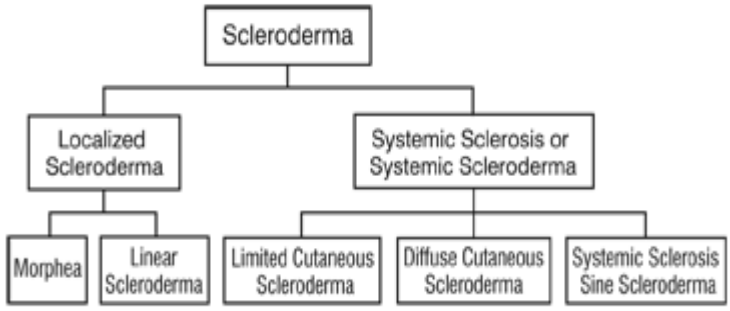
Although it is often referred to as if it were a single disease, scleroderma is really a symptom of a group of diseases that involve the abnormal growth of connective tissue, which supports the skin and internal organs. It is sometimes used, therefore, as an umbrella term for these disorders. In some forms of scleroderma, hard, tight skin is the extent of this abnormal process. In other forms, however, the problem goes much deeper, affecting blood vessels and internal organs, such as the heart, lungs, and kidneys.

Scleroderma is called both a rheumatic (roo-MA-tik) disease and a connective tissue disease. The term rheumatic disease refers to a group of conditions characterized by inflammation or pain in the muscles, joints, or fibrous tissue. A connective tissue disease is one that affects tissues such as skin, tendons, and cartilage.

In this booklet we'll discuss the forms of scleroderma and the problems associated with each of them, as well as diagnosis and disease management. We'll also take a look at what research is telling us about their possible causes and most effective treatments. And we will describe ways for people with scleroderma to live longer, healthier, and more productive lives.

What Are the Different Types of Scleroderma?

The group of diseases we call scleroderma falls into two main classes: localized scleroderma and systemic sclerosis. (Localized diseases affect only certain parts of the body; systemic diseases can affect the whole body.) Both groups include subgroups (see chart). Although there are different ways these groups and subgroups may be broken down or referred to (and your doctor may use different terms from what you see here), the following is a common way of classifying these diseases:



Localized Scleroderma

Localized types of scleroderma are those limited to the skin and related tissues and, in some cases, the muscle below. Internal organs are not affected by localized scleroderma, and localized scleroderma can never progress to the systemic form of the disease. Often, localized conditions improve or go away on their own over time, but the skin changes and damage that occur when the disease is active can be permanent. For some people, localized scleroderma is serious and disabling.

There are two generally recognized types of localized scleroderma:

Morphea: Morphea (mor-FEE-ah) comes from a Greek word that means "form" or "structure." The word refers to local patches of scleroderma. The first signs of the disease are reddish patches of skin that thicken into firm, oval-shaped areas. The center of each patch becomes ivory colored with violet borders. These patches sweat very little and have little hair growth. Patches appear most often on the chest, stomach, and back. Sometimes they appear on the face, arms, and legs.

Morphea can be either localized or generalized. *Localized morphea* limits itself to one or several patches, ranging in size from a half-inch to 12 inches in diameter. The condition sometimes appears on areas treated by radiation therapy. Some people have both morphea and linear scleroderma (described below). The disease is referred to as *generalized morphea* when the skin patches become very hard and dark and spread over larger areas of the body. Regardless of the type, morphea generally fades out in 3 to 5 years; however, people are often left with darkened skin patches and, in rare cases, muscle weakness.

Linear scleroderma: As suggested by its name, the disease is characterized by a single line or band of thickened or abnormally colored skin. Usually, the line runs down an arm or leg, but in some people it runs down the forehead. People sometimes use the French term *en coup de sabre*, or "sword stroke," to describe this highly visible line.

Systemic Scleroderma (also known as Systemic Sclerosis)

This is the term for the form of the disease that not only includes the skin, but also involves the tissues beneath, the blood vessels, and the major organs. Systemic sclerosis is typically broken down into *limited cutaneous scleroderma* and *diffuse cutaneous scleroderma*. Some doctors break systemic sclerosis down into a third subset called *systemic sclerosis sine (SEEN-ay, Latin for "without") scleroderma*. This means that patients have other manifestations of scleroderma but they do not have any overt skin thickening.

Limited cutaneous scleroderma: Limited cutaneous scleroderma typically comes on gradually and affects the skin only in certain areas: the fingers, hands, face, lower arms, and legs. Most people with limited disease have Raynaud's phenomenon for years before skin thickening starts. Telangiectasia and calcinosis often follow (see definitions below). Gastrointestinal involvement occurs commonly, and some patients have severe lung problems, even though the skin thickening remains limited. People with limited disease often have all or some of the symptoms that some doctors call CREST, which stands for the following:

- **Calcinosis (KAL-sin-OH-sis):** The formation of calcium deposits in the connective tissues, which can be detected by x ray. These deposits are typically found on the fingers, hands, face, and trunk and on the skin above elbows and knees. When the deposits break through the skin, painful ulcers can result.
- **Raynaud's (ray-NOHZ) phenomenon:** A condition in which the small blood vessels of the hands or feet contract in response to cold or anxiety. As the vessels contract, the hands or feet turn white and cold, then blue. As blood flow returns, they become red. Fingertip tissues may suffer damage, leading to ulcers, scars, or gangrene.

- **Esophageal (eh-SOFF-uh-GEE-ul) dysfunction:** Impaired function of the esophagus (the tube connecting the throat and the stomach) that occurs when smooth muscles in the esophagus lose normal movement. In the upper and lower esophagus, the result can be swallowing difficulties. In the lower esophagus, the result can be chronic heartburn or inflammation.
- **Sclerodactyly (SKLER-oh-DAK-till-ee):** Thick and tight skin on the fingers, resulting from deposits of excess collagen within skin layers. The condition makes it harder to bend or straighten the fingers. The skin may also appear shiny and darkened, with hair loss.
- **Telangiectasia (tel-AN-jee-ek-TAY-zee-uh):** A condition caused by the swelling of tiny blood vessels, in which small red spots appear on the hands and face. Although not painful, these red spots can create cosmetic problems.

Diffuse cutaneous scleroderma: This condition typically comes on suddenly. Skin thickening begins in the hands and spreads quickly and over much of the body, affecting the hands, face, upper arms, upper legs, chest, and stomach in a symmetrical fashion (for example, if one arm or one side of the trunk is affected, the other is also affected). Some people may have more area of their skin affected than others. Internally, this condition can damage key organs such as the intestines, lungs, heart, and kidneys.

People with diffuse disease often are tired, lose appetite and weight, and have joint swelling or pain. Skin changes can cause the skin to swell, appear shiny, and feel tight and itchy.

The damage of diffuse scleroderma typically occurs over a few years. After the first 3 to 5 years, people with diffuse disease often enter a stable phase lasting for varying lengths of time. During this phase, symptoms subside: joint pain eases, fatigue lessens, and appetite returns. Progressive skin thickening and organ damage decrease.

Gradually, however, the skin may begin to soften, which tends to occur in reverse order of the thickening process: the last areas thickened are the first to begin softening. Some patients' skin returns to a somewhat normal state, while other patients are left with thin, fragile skin without hair or sweat glands. Serious new damage to the heart, lungs, or kidneys is unlikely to occur, although patients are left with whatever damage they have in specific organs.

People with diffuse scleroderma face the most serious long-term outlook if they develop severe kidney, lung, digestive, or heart problems. Fortunately, less than one-third of patients with diffuse disease develop these severe problems. Early diagnosis and continual and careful monitoring are important.

What Causes Scleroderma?

Although scientists don't know exactly what causes scleroderma, they are certain that people cannot catch it from or transmit it to others. Studies of twins suggest it is also not inherited. Scientists suspect that scleroderma comes from several factors that may include:

Abnormal immune or inflammatory activity: Like many other rheumatic disorders, scleroderma is believed to be an autoimmune disease. An autoimmune disease is one in which the immune system, for unknown reasons, turns against one's own body.

In scleroderma, the immune system is thought to stimulate cells called fibroblasts so they produce too much collagen. The collagen forms thick connective tissue that builds up within the skin and internal organs and can interfere with their functioning. Blood vessels and joints can also be affected.

Genetic makeup: Although genes seem to put certain people at risk for scleroderma and play a role in its course, the disease is not passed from parent to child like some genetic diseases.

Environmental triggers: Research suggests that exposure to some environmental factors may trigger scleroderma-like disease (which is not actually scleroderma) in people who are genetically predisposed to it. Suspected triggers include viral infections, certain adhesive and coating materials, and organic solvents such as vinyl chloride or trichloroethylene. But no environmental agent has been shown to cause scleroderma. In the past, some people believed that silicone breast implants might have been a factor in developing connective tissue diseases such as scleroderma. But several studies have not shown evidence of a connection.

Hormones: By the middle-to-late childbearing years (age 30 to 55), women develop scleroderma 7 to 12 times more often than men. Because of female predominance at these and all ages, scientists suspect that hormonal differences between women and men play a part in the disease. However, the role of estrogen or other female hormones has not been proven.

Who Gets Scleroderma?

Although scleroderma is more common in women, the disease also occurs in men and children. It affects people of all races and ethnic groups. However, there are some patterns by disease type. For example:

- Localized forms of scleroderma are more common in people of European descent than in African Americans. Morphea usually appears between the ages of 20 and 40, and linear scleroderma usually occurs in children or teenagers.
- Systemic scleroderma, whether limited or diffuse, typically occurs in people from 30 to 50 years old. It affects more women of African American than European descent.

Because scleroderma can be hard to diagnose and it overlaps with or resembles other diseases, scientists can only estimate how many cases there actually are. It is estimated that 49,000 adults in the United States have systemic sclerosis.

For some people, scleroderma (particularly the localized forms) is fairly mild and resolves with time. But for others, living with the disease and its effects day to day has a significant impact on their quality of life.

How Is Scleroderma Diagnosed?

Depending on your particular symptoms, a diagnosis of scleroderma may be made by a general internist, a dermatologist (a doctor who specializes in treating diseases of the skin, hair, and nails), an orthopedist (a doctor who treats bone and joint disorders), a pulmonologist (a lung specialist), or a rheumatologist (a doctor specializing in treatment of musculoskeletal disorders and rheumatic diseases). A diagnosis of scleroderma is based largely on the medical history and findings from the physical exam. To make a diagnosis, your doctor will ask you a lot of questions about what has happened to you over time and about any symptoms you may be experiencing. Are you having a problem with heartburn or swallowing? Are you often tired or achy? Do your hands turn white in response to anxiety or cold temperatures?

Once your doctor has taken a thorough medical history, he or she will perform a physical exam. Finding one or more of the following factors can help the doctor diagnose a certain form of scleroderma:

- changed skin appearance and texture, including swollen fingers and hands and tight skin around the hands, face, mouth, or elsewhere
- calcium deposits developing under the skin
- changes in the tiny blood vessels (capillaries) at the base of the fingernails
- thickened skin patches.

Finally, your doctor may order lab tests to help confirm a suspected diagnosis. At least two proteins, called antibodies, are commonly found in the blood of people with scleroderma:

- Antitopoisomerase-1 or Anti-Scl-70 antibodies appear in the blood of up to 30 percent of people with diffuse systemic sclerosis.
- Anticentromere antibodies are found in the blood of many people with limited systemic sclerosis.

A number of other scleroderma-specific antibodies can occur in people with scleroderma, although less frequently. When present, however, they are helpful in clinical diagnosis and may give additional information as to the risks for specific organ problems.

Because not all people with scleroderma have these antibodies and because not all people with the antibodies have scleroderma, lab test results alone cannot confirm the diagnosis.

In some cases, your doctor may order a skin biopsy (the surgical removal of a small sample of skin for microscopic examination) to aid in or help confirm a diagnosis. However, skin biopsies also have their limitations: biopsy results cannot distinguish between localized and systemic disease, for example.

Diagnosing scleroderma is easiest when a person has typical symptoms and rapid skin thickening. In other cases, a diagnosis may take months, or even years, as the disease unfolds and reveals itself and as the doctor is able to rule out some other potential causes of the symptoms.

In some cases, a diagnosis is never made, because the symptoms that prompted the visit to the doctor go away on their own.

Some patients have some symptoms related to scleroderma and may fit into one of the following groups:

- **Undifferentiated connective tissue disease (UCTD):** This is a term for patients who have some signs and symptoms of various related diseases, but not enough symptoms of any one disease to make a definitive diagnosis. In other words, their condition hasn't "differentiated" into a particular connective tissue disease. In time, UCTD can go in one of three directions: it can change into a systemic disease such as systemic sclerosis, systemic lupus erythematosus, or rheumatoid arthritis; it can remain undifferentiated; or it can improve spontaneously.
- **Overlap syndromes:** This is a disease combination in which patients have symptoms and lab findings characteristic of two or more conditions.

What Other Conditions Can Look Like Scleroderma?

A number of other diseases have symptoms similar to those seen in scleroderma. Here are some of the most common scleroderma "look-alikes."

Eosinophilic fasciitis (EF) (EE-oh-SIN-oh-FIL-ik fa-shi-EYE-tis): This disease involves the fascia (FA-shuh), the thin connective tissue around the muscles, particularly those of the forearms, arms, legs, and trunk. EF causes the muscles to become encased in collagen, the fibrous protein that makes up tissue such as the skin and tendons. Permanent shortening of the muscles and tendons, called contractures, may develop, sometimes causing disfigurement and problems with joint motion and function. EF may begin after hard physical exertion. The disease usually fades away after several years, but people sometimes have relapses. Although the upper layers of the skin are not thickened in EF, the thickened fascia may cause the skin to look somewhat like the tight, hard skin of scleroderma. A skin biopsy easily distinguishes between the two diseases.

Skin thickening on the fingers and hands: This also appears with diabetes, mycosis fungoides, amyloidosis, and adult celiac disease. It can also result from hand trauma.

Generalized scleroderma-like skin thickening: This may occur with scleromyxedema, graft-versus-host disease, porphyria cutanea tarda, and human adjuvant disease.

Internal organ damage: Similar to that seen in systemic sclerosis, this may instead be related to primary pulmonary hypertension, idiopathic pulmonary fibrosis, or collagenous colitis.

Raynaud's phenomenon: This condition also appears with atherosclerosis or systemic lupus erythematosus or in the absence of underlying disease.

An explanation of most of these other diseases is beyond the scope of this booklet. What's important to understand, however, is that diagnosing scleroderma isn't always easy, and it may take time for you and your doctor to do this. Although having a definite diagnosis may be helpful, you do not need to know the precise form of your disease to receive proper treatment.

How Is Scleroderma Treated?

Because scleroderma can affect many different organs and organ systems, you may have several different doctors involved in your care. Typically, care will be managed by a rheumatologist (a doctor specializing in treatment of musculoskeletal disorders and rheumatic diseases). Your rheumatologist may refer you to other specialists, depending on the specific problems you are having. For example, you may see a dermatologist for the treatment of skin symptoms, a nephrologist for kidney complications, a cardiologist for heart complications, a gastroenterologist for problems of the digestive tract, and a pulmonary specialist for lung involvement.

In addition to doctors, professionals such as nurse practitioners, physician assistants, physical or occupational therapists, psychologists, and social workers may play a role in your care. Dentists, orthodontists, and even speech therapists can treat oral complications that arise from thickening of tissues in and around the mouth and on the face.

Currently, there is no treatment that controls or stops the underlying problem—the overproduction of collagen—in all forms of scleroderma. Thus, treatment and management focus on relieving symptoms and limiting damage. Your treatment will depend on the particular problems you are having. Some treatments will be prescribed or given by your doctor. Others are things you can do on your own.

Here is a listing of the potential problems that can occur in *systemic scleroderma* and the medical and nonmedical treatments for them. These problems do not occur as a result or complication of *localized scleroderma*. This listing is not complete because different people experience different problems with scleroderma and not all treatments work equally well for all people. Work with your doctor to find the best treatment for your specific symptoms.

Raynaud's phenomenon: More than 90 percent of people with scleroderma have this condition, in which the fingers and sometimes other extremities change color in response to cold temperature or anxiety. For many, Raynaud's phenomenon precedes other manifestations of the disease. In other people, however, Raynaud's phenomenon is unrelated to scleroderma, but may signal damage to the blood vessels supplying the hands arising from occupational injuries (from using jackhammers, for example), trauma, excessive smoking, circulatory problems, drug use, or exposure to toxic substances. For some people, cold fingers and toes are the extent of the problem and are little more than a nuisance. For others, the condition can worsen and lead to puffy fingers, finger ulcers, and other complications that require aggressive treatment.

If you have Raynaud's phenomenon, the following measures may make you more comfortable and help prevent problems:

- Don't smoke! Smoking narrows the blood vessels even more and makes Raynaud's phenomenon worse.
- Dress warmly, with special attention to hands and feet. Dress in layers and try to stay indoors during cold weather.
- Use biofeedback, which governs various body processes that are not normally thought of as being under conscious control, and relaxation exercises.
- For severe cases, speak to your doctor about prescribing drugs called calcium channel blockers, which can open up small blood vessels and improve circulation. Other drugs are in development and may become available.
- If Raynaud's phenomenon leads to skin sores or ulcers, increasing your dose of calcium channel blockers (ONLY under the direction of your doctor may help. You can also protect skin ulcers from further injury or infection by applying nitroglycerine paste or antibiotic cream. Severe ulcerations on the fingertips can be treated with bioengineered skin.

Stiff, painful joints: In diffuse systemic sclerosis, hand joints can stiffen because of hardened skin around the joints or inflammation within them. Other joints can also become stiff and swollen.

- Stretching exercises under the direction of a physical or occupational therapist are extremely important to prevent loss of joint motion. These should be started as soon as scleroderma is diagnosed.
- Exercise regularly. Ask your doctor or physical therapist about an exercise plan that will help you increase and maintain range of motion in affected joints. Swimming can help maintain muscle strength, flexibility, and joint mobility.
- Use acetaminophen or an over-the-counter or prescription nonsteroidal anti-inflammatory drug, as recommended by your doctor, to help relieve joint or muscle pain. If pain is severe, speak to a rheumatologist about the possibility of prescription-strength drugs to ease pain and inflammation.
- Learn to do things in a new way. A physical or occupational therapist can help you learn to perform daily tasks, such as lifting and carrying objects or opening doors, in ways that will put less stress on tender joints.

Skin problems: When too much collagen builds up in the skin, it crowds out sweat and oil glands, causing the skin to become dry and stiff. If your skin is affected, try the following:

- Apply oil-based creams and lotions frequently, and always right after bathing.
- Apply sunscreen before you venture outdoors to protect against further damage from the sun's rays.

- Use humidifiers to moisten the air in your home in colder winter climates. Clean humidifiers often to stop bacteria from growing in the water.
- Avoid very hot baths and showers, as hot water dries the skin.
- Avoid harsh soaps, household cleaners, and caustic chemicals, if at all possible. Otherwise, be sure to wear rubber gloves when you use such products.
- Exercise regularly. Exercise, especially swimming, stimulates blood circulation to affected areas.

Dry mouth and dental problems: Dental problems are common in people with scleroderma for a number of reasons. Tightening facial skin can make the mouth opening smaller and narrower, which makes it hard to care for teeth; dry mouth caused by salivary gland damage speeds up tooth decay; and damage to connective tissues in the mouth can lead to loose teeth. You can avoid tooth and gum problems in several ways:

- Brush and floss your teeth regularly. If hand pain and stiffness make this difficult, consult your doctor or an occupational therapist about specially made toothbrush handles and devices to make flossing easier.
- Have regular dental checkups. Contact your dentist immediately if you experience mouth sores, mouth pain, or loose teeth.
- If decay is a problem, ask your dentist about fluoride rinses or prescription toothpastes that remineralize and harden tooth enamel.
- Consult a physical therapist about facial exercises to help keep your mouth and face more flexible.
- Keep your mouth moist by drinking plenty of water, sucking ice chips, using sugarless gum and hard candy, and avoiding mouthwashes with alcohol. If dry mouth still bothers you, ask your doctor about a saliva substitute—or prescription medications such as pilocarpine hydrochloride or cevimeline hydrochloride—that can stimulate the flow of saliva.

Gastrointestinal (GI) problems: Systemic sclerosis can affect any part of the digestive system. As a result, you may experience problems such as heartburn, difficulty swallowing, early satiety (the feeling of being full after you've barely started eating), or intestinal complaints such as diarrhea, constipation, and gas. In cases where the intestines are damaged, your body may have difficulty absorbing nutrients from food. Although GI problems are diverse, here are some things that might help at least some of the problems you have:

- Eat small, frequent meals.
- To keep stomach contents from backing up into the esophagus, stand or sit for at least an hour (preferably 2 or 3 hours) after eating. When it is time to sleep, keep the head of your bed raised using blocks.
- Avoid late-night meals, spicy or fatty foods, alcohol, and caffeine, which can aggravate GI distress.

- Eat moist, soft foods, and chew them well. If you have difficulty swallowing or if your body doesn't absorb nutrients properly, your doctor may prescribe a special diet.
- Ask your doctor about prescription medications for problems such as diarrhea, constipation, and heartburn. Some drugs called proton pump inhibitors are highly effective against heartburn. Oral antibiotics may stop bacterial overgrowth in the bowel, which can be a cause of diarrhea in some people with systemic sclerosis.

Lung damage: Virtually all people with systemic sclerosis have some loss of lung function. Some develop severe lung disease, which comes in two forms: pulmonary fibrosis (hardening or scarring of lung tissue because of excess collagen) and pulmonary hypertension (high blood pressure in the artery that carries blood from the heart to the lungs). Treatment for the two conditions is different:

- Pulmonary fibrosis may be treated with drugs that suppress the immune system, such as cyclophosphamide or azathioprine, along with low doses of corticosteroids.
- Pulmonary hypertension may be treated with drugs that dilate the blood vessels, such as prostacyclin, or with newer medications that are prescribed specifically for treating pulmonary hypertension.

Regardless of your particular lung problem or its medical treatment, your role in the treatment process is essentially the same. To minimize lung complications, work closely with your medical team. Do the following:

- Watch for signs of lung disease, including fatigue, shortness of breath or difficulty breathing, and swollen feet. Report these symptoms to your doctor.
- Have your lungs closely checked, using standard lung-function tests, during the early stages of skin thickening. These tests, which can find problems at the earliest and most treatable stages, are needed because lung damage can occur even before you notice any symptoms.
- Get regular flu and pneumonia vaccines as recommended by your doctor. Contracting either illness could be dangerous for a person with lung disease.

Heart problems: Common among people with scleroderma, heart problems include scarring and weakening of the heart (cardiomyopathy), inflamed heart muscle (myocarditis), and abnormal heartbeat (arrhythmia). All of these problems can be treated. Treatment ranges from drugs to surgery and varies depending on the nature of the condition.

Kidney problems: Renal crisis occurs in about 10 percent of all patients with scleroderma, primarily those with early diffuse scleroderma. Renal crisis results in severe uncontrolled high blood pressure, which can quickly lead to kidney failure. It's very important that you take measures to identify and treat the hypertension as soon as it occurs. These are things you can do:

- Check your blood pressure regularly. You should also check it if you have any new or different symptoms such as a headache or shortness of breath. If your blood pressure is higher than usual, call your doctor right away.

- If you have kidney problems, take your prescribed medications faithfully. In the past two decades, drugs known as ACE (angiotensin-converting enzyme) inhibitors, including captopril, enalapril, and lisinopril, have made scleroderma-related kidney failure a less threatening problem than it used to be. But for these drugs to work, you must take them as soon as the hypertension is present.

Cosmetic problems: Even if scleroderma doesn't cause any lasting physical disability, its effects on the skin's appearance—particularly on the face—can take their toll on your self-esteem. Fortunately, there are procedures to correct some of the cosmetic problems scleroderma causes:

- The appearance of telangiectasias—small red spots on the hands and face caused by swelling of tiny blood vessels beneath the skin—may be reduced or even eliminated with the use of guided lasers.
- Facial changes of localized scleroderma—such as the *en coup de sabre* that may run down the forehead in people with linear scleroderma—may be corrected through cosmetic surgery. (However, such surgery is not appropriate for areas of the skin where the disease is active.)

How Can Scleroderma Affect My Life?

Having a chronic disease can affect almost every aspect of your life, from family relationships to holding a job. For people with scleroderma, there may be other concerns about appearance or even the ability to dress, bathe, or handle the most basic daily tasks. Here are some areas in which scleroderma could intrude.

Appearance and self-esteem: Aside from the initial concerns about health and longevity, people with scleroderma quickly become concerned with how the disease will affect their appearance. Thick, hardened skin can be difficult to accept, particularly on the face. Systemic scleroderma may result in facial changes that eventually cause the opening to the mouth to become smaller and the upper lip to virtually disappear. Linear scleroderma may leave its mark on the forehead. Although these problems can't always be prevented, their effects may be minimized with proper treatment. Also, special cosmetics—and in some cases plastic surgery—can help conceal scleroderma's damage.

Caring for yourself: Tight, hard connective tissue in the hands can make it difficult to do what were once simple tasks, such as brushing your teeth and hair, pouring a cup of coffee, using a knife and fork, unlocking a door, or buttoning a jacket. If you have trouble using your hands, consult an occupational therapist, who can recommend new ways of doing things or devices to make tasks easier. Devices as simple as Velcro fasteners and built-up brush handles can help you be more independent.

Family relationships: Spouses, children, parents, and siblings may have trouble understanding why you don't have the energy to keep house, drive to soccer practice, prepare meals, or hold a job the way you used to. If your condition isn't that visible, they may even suggest you are just being lazy. On the other hand, they may be overly concerned and eager to help you, not allowing you to do the things you are able to do or giving up their own interests and activities to be with you.

It's important to learn as much about your form of the disease as you can and to share any information you have with your family. Involving them in counseling or a support group may also help them better understand the disease and how they can help you.

Sexual relations: Sexual relationships can be affected when systemic scleroderma enters the picture. For men, the disease's effects on the blood vessels can lead to problems achieving an erection. For women, damage to the moisture-producing glands can cause vaginal dryness that makes intercourse painful. People of either sex may find they have difficulty moving the way they once did. They may be self-conscious about their appearance or afraid that their sexual partner will no longer find them attractive. With communication between partners, good medical care, and perhaps counseling, many of these changes can be overcome or at least worked around.

Pregnancy and childbearing: In the past, women with systemic scleroderma were often advised not to have children. But thanks to better medical treatments and a better understanding of the disease itself, that advice is changing. (Pregnancy, for example, is not likely to be a problem for women with localized scleroderma.) Although blood vessel involvement in the placenta may cause babies of women with systemic scleroderma to be born early, many women with the disease can have safe pregnancies and healthy babies if they follow some precautions.

One of the most important pieces of advice is to wait a few years after the disease starts before attempting a pregnancy. During the first 3 years, you are at the highest risk of developing severe problems of the heart, lungs, or kidneys that could be harmful to you and your unborn baby.

If you haven't developed severe organ problems within 3 years of the disease's onset, your chances of such problems are less and pregnancy would be safer. But it is important to have both your disease and your pregnancy monitored regularly. You'll probably need to stay in close touch with both the doctor you typically see for your scleroderma and an obstetrician who is experienced in guiding high-risk pregnancies.

How Can I Play a Role in My Health Care?

Although your doctors direct your treatment, you are the one who must take your medicine regularly, follow your doctor's advice, and report any problems promptly. In other words, the relationship between you and your doctors is a partnership, and you are the most important partner. Here's what you can do to make the most of this important role.

- ***Get educated:*** Knowledge is your best defense against this disease. Learn as much as you can about scleroderma, both for your own benefit and to educate the people in your support network.
- ***Seek support:*** Recruit family members, friends, and coworkers to build a support network. This network will help you get through difficult times: when you are in pain; when you feel angry, sad, or afraid; or when you're depressed. Also, look for a scleroderma support group in your community by calling a national scleroderma organization. (See "For More Information.") If you can't find a support group, you might want to consider organizing one.

- ***Assemble a health care team:*** You and your doctors will lead the team. Other members may include physical and occupational therapists, a psychologist or social worker, a dentist, and a pharmacist.
- ***Be patient:*** Understand that a final diagnosis can be difficult and may take a long time. Find a doctor with experience treating people with systemic and localized scleroderma. Then, even if you don't yet have a specific diagnosis, you will get understanding and the right treatment for your symptoms.
- ***Speak up:*** When you have problems or notice changes in your condition, don't feel too self-conscious to speak up during your appointment or even call your doctor or another member of your health care team. No problem is too small to ask about, and early treatment for any problem can make the disease more manageable.
- ***Don't accept depression:*** Although it's understandable that a person with a chronic illness such as scleroderma would become depressed, don't accept depression as a normal consequence of your condition. If depression makes it hard for you to function well, don't hesitate to ask your health care team for help. You may benefit from speaking with a psychologist or social worker or from using one of the effective medications on the market.
- ***Learn coping skills:*** Meditation, calming exercises, and relaxation techniques may help you cope with emotional difficulties and relieve pain and fatigue. Ask a member of your health care team to teach you these skills or to refer you to someone who can.
- ***Ask the experts:*** If you have problems doing daily activities, from brushing your hair and teeth to driving your car, consult an occupational or physical therapist. They have more helpful hints and devices than you can probably imagine. Social workers can often help resolve financial and insurance matters.

Research Highlights

No one can say for sure when—or if—a cure will be found. But research is providing the next best thing: better ways to treat symptoms, prevent organ damage, and improve the quality of life for people with scleroderma. In the past two decades, multidisciplinary research has also provided new clues for understanding the disease, which is an important step toward prevention and cure.

Leading the way in funding for this research is the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a part of the U.S. Department of Health and Human Services' National Institutes of Health (NIH). Other sources of funding for scleroderma research include pharmaceutical companies and organizations such as the Scleroderma Foundation, the Scleroderma Research Foundation, and the Arthritis Foundation. Scientists at universities and medical centers throughout the United States conduct much of this research.

Studies of the immune system, genetics, cell biology, and molecular biology have helped reveal the causes of scleroderma, improve existing treatment, and create entirely new treatment approaches.

Some advances in the understanding or treatment of scleroderma include the following:

- Building on research that identified a gene associated with scleroderma in Oklahoma Choctaw Native Americans, scientists are using new technology to look for other genes associated with the disease's development and severity.
- The drug cyclophosphamide has been found effective in treating lung fibrosis. One recent study suggested that treating lung problems early with this immunosuppressive drug may help prevent further damage and increase chances of survival. In further research assessing the impact of cyclophosphamide on quality of life in people with lung involvement, 47 percent of people on the drug reported their health was somewhat or much better after 1 year, compared with 18 percent in the placebo group.
- ACE inhibitors are used increasingly for scleroderma-related kidney problems. For the past two decades, ACE inhibitors have greatly reduced the risk of kidney failure in people with scleroderma. Now there is evidence that use of ACE inhibitors can actually heal the kidneys of people on dialysis for scleroderma-related kidney failure. As many as half the people who continue ACE inhibitors while on dialysis may be able to go off dialysis in 12 to 18 months.
- Several drugs are now available to treat pulmonary hypertension. Previously, pulmonary hypertension was associated with a poor outcome, but medications such as prostacyclins, endothelin-receptor antagonists, and phosphodiesterase inhibitors—epoprostenol sodium, bosentan, and sildenafil—have increased the quality of life and life expectancy for people with this dangerous form of lung damage.

Other studies are examining the following:

- The identification of several early immune system targets in scleroderma. Building on this research, scientists are continuing to look for ways to treat scleroderma earlier, before it has a chance to cause irreparable damage.
- The theory that scleroderma is a more aggressive disease associated with more internal organ damage and a worse prognosis in non-Caucasians. Researchers believe that although factors related to both genetics and socioeconomic status may play a role, autoantibodies may be the primary reason that African Americans have such severe disease. A current study is examining that theory. Researchers hope that by better understanding the factors involved in scleroderma, they can design interventions that would improve the course and outcome of the disease.
- Changes in the tiny blood vessels of people with scleroderma. By studying these changes, scientists hope to find the cause of cold sensitivity in Raynaud's phenomenon and a way to control the problem.
- Studies have shown that certain chemicals called cytokines, made from cells in the body, enhance the development of increased collagen. New agents that counteract these cytokines may be helpful in preventing skin thickening.

- Skin changes in laboratory mice in which a genetic defect prevents the breakdown of collagen, leading to thick skin and patchy hair loss. Scientists hope that by studying these mice they can answer many questions about skin changes in scleroderma. Scientists are also working to establish mouse models for other problems related to scleroderma. These models will make it easier to understand these problems and develop treatments for them.

Scleroderma research continues to advance as scientists and doctors learn more about how the disease develops and its underlying mechanisms. NIAMS funds a research center specializing in scleroderma at the University of Texas Health Science Center at Houston. Scientists there are conducting laboratory and clinical research on the disease with the goal of translating basic science findings quickly into improved treatment and patient care.

More Questions? Count on More Answers

Scleroderma poses a series of challenges for both patients and their health care teams. The good news is that scientists, doctors, and other health care professionals continue to find new ways to make earlier diagnoses and manage disease better. In addition, active patient support groups share with, care for, and educate each other. The impact of all of this activity is that people with scleroderma do much better and remain active far longer than they did 20 or 30 years ago. As for tomorrow, patients and the medical community will continue to push for longer, healthier, and more active lives for people with the diseases collectively known as scleroderma.

For More Information

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)
Information Clearinghouse
National Institutes of Health

1 AMS Circle
Bethesda, MD 20892-3675
Phone: 301-495-4484
Toll Free: 877-22-NIAMS (226-4267)
TTY: 301–565–2966
Fax: 301-718-6366
Email: NIAMSinfo@mail.nih.gov
Website: http://www.niams.nih.gov

Other Resources

American Academy of Dermatology (AAD)

Web site: http://www.aad.org

American College of Rheumatology (ACR)

Web site: http://www.rheumatology.org

Scleroderma Foundation

Web site: http://www.scleroderma.org

Scleroderma Research Foundation

Web site: http://www.srfcure.org

Arthritis Foundation

Web site: http://www.arthritis.org

Key Words

Antibodies. Special proteins produced by the body's immune system. They recognize and help fight infectious agents, such as bacteria and other foreign substances that invade the body. The presence of certain antibodies in the blood can help to diagnose some diseases, including some forms of scleroderma.

Atherosclerosis. Aabnormal fatty deposits in the inner layers of large or medium-sized arteries, which can lead to hardening and narrowing of the arteries and blockages of the blood supply, especially to the heart.

Autoimmune disease. A disease in which the body's immune system turns against and damages its own tissues.

Calcinosis. The formation of calcium deposits in the connective tissues, which can be detected by x ray. These deposits are typically found on the fingers, hands, face, and trunk and on the skin above elbows and knees. When the deposits break through the skin, painful ulcers can result.

Calcium channel blockers. Medicines that lower blood pressure, relieve chest pain, and stabilize normal heart rhythms by inhibiting calcium movement into the heart muscles and smooth muscle cells. They are used to treat a variety of conditions and to prevent circulatory and kidney problems in scleroderma.

Collagen. A fabric-like material of fibrous threads that is a key component of the body's connective tissues. In scleroderma, either too much collagen is produced or it is produced in the wrong places, causing stiff and inflamed skin, blood vessels, and internal organs.

Connective tissue. Tissues such as skin, tendons, and cartilage that support and hold body parts together. The chief component of connective tissue is collagen.

CREST syndrome. An acronym for a collection of symptoms that occur to some degree in all people with systemic sclerosis. The symptoms are calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasia. Because of the predominance of

CREST symptoms in people with limited systemic sclerosis, some people use the term CREST syndrome when referring to that form of the disease.

Eosinophilic fasciitis. A scleroderma-like disorder (often considered to be a localized form of scleroderma) featuring inflammation of the fascia (the thin, sheet-like connective tissues surrounding the muscles and other body structures) and an abnormally high number of a specific kind of white blood cells (eosinophils). The result of the inflammation may be fibrous buildup in the skin of arms and legs, contractures, and carpal tunnel syndrome.

Esophageal dysfunction. Impaired function of the esophagus (the tube connecting the throat and the stomach) that occurs when smooth muscles in the esophagus lose normal movement. In the upper and lower esophagus, the result can be swallowing difficulties. In the lower esophagus, the result can be chronic heartburn or inflammation.

Fibroblast. A type of cell in connective tissue that secretes proteins, including collagen.

Fibrosis. A condition marked by increased fibrous tissue that develops between the cells of various organs or tissues. It is a common feature of scleroderma and some other diseases. Fibrosis causes hardening or stiffening of tissues in the skin, joints, and internal organs.

Graft-versus-host disease. A major complication of bone marrow transplantations and sometimes blood transfusions in which white blood cells called lymphocytes, which are found in the marrow or blood, attack tissues in the body into which they were transplanted.

Pulmonary fibrosis. Hardening or scarring of lung tissue because of excess collagen. Pulmonary fibrosis occurs in a small percentage of people with systemic sclerosis.

Pulmonary hypertension. Abnormally high blood pressure in the arteries supplying the lungs that may be caused by a number of factors, including damage from fibrosis.

Raynaud's phenomenon. A condition in which the small blood vessels of the hands or feet contract in response to cold or anxiety. As the vessels contract, the hands or feet turn white and cold, then blue. As blood flow returns, they become red. Fingertip tissues may suffer damage, leading to ulcers, scars, or gangrene.

Rheumatic. An adjective used to describe a group of conditions characterized by inflammation or pain in the muscles, joints, and fibrous tissue. Rheumatic diseases or disorders can be related to autoimmunity or other causes.

Sclerodactyly. Thick and tight skin on the fingers, resulting from deposits of excess collagen within skin layers. The condition makes it harder to bend or straighten the fingers. The skin may also appear shiny and darkened, with hair loss.

Systemic condition. A condition involving the body as a whole, as opposed to limited conditions that affect particular parts of the body.

Systemic lupus erythematosus. A systemic rheumatic disease that occurs predominantly in women and is characterized by autoimmune activity, a facial rash across the bridge of the nose and cheeks, Raynaud's phenomenon, joint pain and swelling, fever, chest pain, hair loss, and other symptoms. Many of its symptoms overlap with those of scleroderma.

Telangiectasia. A condition caused by the swelling of tiny blood vessels, in which small red spots appear on the hands and face. Although not painful, these red spots can create cosmetic problems.

Scleroderma Examination

Select the ***best*** answer to each of the following items. Mark your responses on the Answer form.

1. There are an estimated _____ people in the United States who have scleroderma, about one third of whom have the systemic form of scleroderma.

a. 15,000
b. 150,000
c. 300,000
d. None of the above

2. Scleroderma, or systemic sclerosis, is a chronic _____ generally classified as one of the autoimmune rheumatic diseases.

a. congenital disease
b. connective tissue disease
c. skin disorder
d. None of the above

3. 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.

a. True
b. False

4. Scleroderma is not _____.

a. contagious
b. infectious
c. cancerous or malignant
d. All of the above

5. 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. True
b. False

6. The diagnostic process may require consultation with rheumatologists (arthritis specialists), and/or dermatologists (skin specialists) and require _____ and numerous other specialized tests depending upon which organs are affected.

a. blood studies
b. skin tests
c. CT scans
d. None of the above

7. Localized scleroderma is more common in children, whereas systemic scleroderma is more common in adults. Overall female patients outnumber male patients about _____ to one, and the average age at diagnosis is in the forties.

a. two
b. three
c. four
d. six

8. 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. It is known that scleroderma involves overproduction of collagen.

a. True
b. False

9. Most patients do not have any relatives with scleroderma and their children do not get scleroderma. Research indicates that there is a susceptibility gene which raises the likelihood of getting scleroderma, but by itself does not cause the disease.

a. True
b. False

10. At the present time, there is no cure for scleroderma, but there are many treatments available. Some are directed at particular symptoms like _____, which can be controlled by medications called proton pump inhibitors or medicine to improve the motion of the bowel.

a. swelling
b. heartburn
c. arthritis
d. None of the above

11. The diffuse form of scleroderma (systemic sclerosis) involves symmetric thickening of skin of the _____ which can rapidly progress to hardening after an early inflammatory phase.

a. extremities
b. face
c. trunk
d. All of the above

12. The symptoms of scleroderma depend on the type of scleroderma present and the extent of external and internal involvement in the individual affected. Because scleroderma can affect the skin, esophagus, blood vessels, _____ the symptoms it causes can involve many areas of the body.

a. kidneys
b. lungs
c. blood pressure and bowels
d. All of the above

13. Scleroderma affecting the esophagus leads to heartburn. This is directly a result of stomach acid flowing back up into the esophagus. Sometimes this can lead to scarring of the esophagus with difficulty swallowing and/or localized pain in the central chest.

a. True
b. False

14. Inflammation of the lungs in scleroderma can cause _____, resulting in shortness of breath, especially with physical exertion. Elevated pressure in the arteries to the lungs (pulmonary hypertension) can also cause shortness of breath and difficulty getting an adequate breath with activity.

a. scarring
b. swelling
c. soreness
d. All of the above

15. Aggressive treatment of elevations in blood pressure have been extremely important in prevent kidney failure. Blood-pressure medications, such as captopril, are frequently used.

a. True
b. False

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