



Systemic Sclerosis: Diffuse and Limited

INTRODUCTION

Scleroderma literally means “hard skin.” But this disease is also called systemic sclerosis (SSc), because it can affect many other organs too. It’s a “systemic” disease that may affect your vascular system (blood vessels) and immune system.

Most people with systemic sclerosis have skin issues, such as skin scarring (fibrosis), and swollen, tight, hardened skin on the fingers, face or arms.

SSc often affects blood vessels, causing Raynaud phenomenon in about 90 percent of people. The skin on your fingers change color, turning red, white or blue when exposed to cold or if you’re stressed.

SSc also affects your immune system. About 95 percent of people with SSc have abnormal levels of autoimmune antibodies to the nuclei of their own cells (called anti-nuclear antibodies or ANA).

TYPES OF SYSTEMIC SCLEROSIS

For unknown reasons, SSc causes you higher than normal amounts of the protein collagen deposited in your skin, making skin thicker and tougher. You may have excess collagen in their fingers, or possibly on your face or hands. Some people have collagen skin deposits all over their body. Based on the extent of skin involvement, SSc is grouped into diffuse cutaneous scleroderma and limited cutaneous scleroderma.

Diffuse Cutaneous Scleroderma

People with diffuse cutaneous scleroderma often have thick or tight skin on their arms above and below the elbows. The skin on their legs above and below the knees, the face, and the chest and abdomen may also become tight, thick or hard.

Skin thickening often progresses rapidly, in just weeks or months. It’s very bothersome.

Your doctor measures skin involvement by palpating (touching with the hands) 17 body areas, then scoring the level of thickness using the modified Rodnan Skin Score (mRSS). Skin thickening in diffuse cutaneous scleroderma may continue for 1-3 years, then slow down and level off. After 1-2 years of stability, skin thickening usually eases, and skin begins to thin or soften.

Limited Cutaneous Scleroderma

People with limited cutaneous scleroderma have thick, tight, or hard skin below, but not above, their elbows and knees. It may or may not affect skin on their face.

Skin thickening frequently develops gradually, and it may be milder. Over time, the mRSS skin score is usually low and stays stable.

People with either diffuse or limited cutaneous scleroderma may have:

- Raynaud phenomenon
- Heartburn and other esophagus problems, like trouble swallowing food
- Skin sores, usually on the fingers, but possibly in the wrists, elbows or ankles
- Tummy grumbles, feeling “full” after eating only a small amount of food, bloated or swollen belly after eating, constipation, diarrhea or changes in bowel habits
- Lung fibrosis (scarring) in about 40 percent of people, causing shortness of breath
- Pulmonary hypertension (PAH), or high blood pressure in arteries that deliver oxygen to your lungs, in about 10–20 percent

Common problems for people with diffuse cutaneous SSc:

- Kidney failure in about 15–20 percent of people. Treatments are available to preserve kidney function and, if treated early, prolong life for many people.
- Heart involvement, including fluid around the heart, heart rhythm disturbances, and possibly heart failure in around 25 percent of people
- Joint aches and pains, reduced motion in joints like fingers, wrists, elbows, shoulders or knees, and, decline in hand function in many people

HOW OTHER ORGANS MAY BE AFFECTED:

Skin

Skin is all over your body. To measure skin involvement in SSc, your doctor touches and gently pinches your skin in 17 body areas. They assign a number to “score” skin thickness on a scale of 0–3. Zero is normal, 3 is very thickened. All 17 scores are added for a total score from 0–51.

Lungs

Most people with SSc develop lung tissue scarring that may be seen on a high-resolution chest computed tomography, or CT scan. Serious lung problems aren’t common, but about 40 percent of people do have a decline in lung function. Vital capacity, or how much air lungs can move in and out in one deep breath, may decline, because scarring in tissue makes your lungs stiffer. Immunosuppressive treatments like cyclophosphamide and mycophenolate can be prescribed to slow lung damage.

Kidneys

Kidney failure is a severe complication usually seen in people with diffuse cutaneous scleroderma who have had systemic sclerosis for less than five years. Blood flow to their kidneys declines, and this triggers a release of hormones that can increase blood pressure and decrease blood flow to the kidneys even more. This is called “renal crisis.” Your doctor will measure

creatinine levels in your blood to test your kidney function.

Right now, we can’t predict who will develop kidney complications. We do know that most people who develop kidney failure also have new-onset high blood pressure at the same time, so your doctor will check your blood pressure frequently. Get a home blood pressure monitor device to check your blood pressures three times a week. This may detect early signs of renal crisis. If your blood pressure is higher than 160/90 on two occasions, 12 hours apart, contact your doctor. They can prescribe an angiotensin-converting enzyme (ACE inhibitor) medicine to lower your blood pressure.

Unfortunately, some people will need to start on dialysis. Up to one half of people who start on dialysis are able to stop it within 18-24 months of kidney failure onset, as long as they continue taking ACE inhibitors or other blood pressure medications like angiotensin-receptor blockers (losartan or valsartan).

Heart

If your kidneys fail, you may also develop heart failure, at least temporarily. With successful treatment of kidney failure, your heart function may return to normal. A small percentage of people develop scarring of heart muscle that causes abnormal heart function.

Gastrointestinal

SSc often causes a “lazy” muscle to develop in the esophagus. This causes heartburn or a sensation of food “sticking” in your chest. Lazy muscles in your stomach or intestines can cause you to feel full after eating even a small amount. You may feel bloated after eating, or develop chronic diarrhea, chronic constipation, or disrupted bowel habits. Work with your doctor to find the area of your gut that is causing problems, so they can suggest treatments.

Musculoskeletal Pain

Systemic sclerosis often causes musculoskeletal pain in the first few years, including arthritis (joint pain or swelling), tendonitis, bursitis, muscle aches or fibromyalgia (sensitivity to pain, fatigue, very tender muscles). With diffuse

cutaneous scleroderma, musculoskeletal pain often eases once your skin starts to soften.

WHAT'S DOWN THE ROAD?

Significant heart, lung or kidney problems tend to appear in the first five years of SSc. If these complications haven't appeared, they are unlikely to occur later on. By the fifth year, SSc disease typically slows down. Skin begins to soften. Fatigue, joint and muscle aches, and joint mobility usually improves.

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Please note that this brochure is provided for educational purposes only. It is not intended to substitute for informed medical advice.

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