



# VASCULITIS

## Overview

Scleroderma is a disease which is known for causing thickening and hardening of the skin. It is much more than that, as it can affect many other parts of the body, including the lungs, gut, and kidneys. As of today, there is still no cure, but management options are improving.

## Frequently Asked Questions

### What is scleroderma?

- Scleroderma, also known as Systemic Sclerosis, is a condition with certain characteristic findings, most common increasing thickness and hardening of the skin. However, it can affect the skin in other ways, as well as other organs, including the kidneys, lungs and stomach.

### How common is scleroderma?

- Scleroderma is not a common disease, with a new diagnosis in 1 in 100,000 people per year. It more commonly affects women, particularly between the ages of 30 and 50.

### Why does scleroderma happen?

- The exact cause of scleroderma remains unknown. It is possible an unidentified trigger in a patient with a certain but unknown genetic makeup sets off an immune mediated reaction which causes the different manifestations to appear.

### What are the common manifestations of Scleroderma?

- The most common manifestations of scleroderma include:
  - Skin changes
  - The skin can thicken and harden, most often in the hands but can occur anywhere
  - Calcium deposits can occur under the skin, called calcinosis
  - Small blood vessel in the skin can become dilated, called telangiectasias

- Raynaud's phenomenon can be a benign process when the hands or feet changes colours – classically white, blue then red – associated with pain in the cold. It is also one of the first signs of scleroderma or other connective tissue diseases. When associated with an underlying disease like scleroderma, Raynaud's can be more severe, causing pits or ulcers in the fingers which do not always heal easily.
- Heartburn, Difficulty Swallowing, and/or Slower GI Motility
- Kidney Problems – Called Scleroderma Renal Crisis, it is an acute kidney failure
- Lung problems – Either Lung Fibrosis (scarring of the lungs) or high blood pressure in the lung vessels, which can cause shortness of breath in some patients
- Other organs that can be involved include bones, joints, muscles, and rarely the heart.

### **When do these typically occur? Do they all happen at once?**

- For some patients, Raynaud's phenomenon is one of the first things noticed, and can precede other symptoms by years. The remaining manifestations can occur at any time. However, some research suggests that most manifestations will appear approximately within the first 5 years of diagnosis.

### **How do you diagnose Scleroderma?**

- An expert in scleroderma, often a rheumatologist, should diagnose scleroderma. Your doctor will listen to your medical history for symptoms suggested from the above list, will complete and physical exam and may review specific laboratory results to come to a diagnosis. There is no blood test that definitely diagnoses scleroderma.

### **I have Raynaud's. Does that mean I am going to get Scleroderma?**

- Raynaud's phenomenon is common affecting about 4% of the general population, meaning very few patients with Raynaud's also have scleroderma.
- Primary Raynaud's (meaning Raynaud's not associated with another disease like scleroderma) often occurs earlier in life, as a teenager or young adult. It is much more common in females than males, and it usually does not cause skin ulcerations or pits.

## How do you treat Scleroderma?

- As of today, there is no cure of scleroderma. Neither is there one strategy to treat the overall disease. Instead, the individual manifestations are each treated on their own as necessary.
- Speak to your physician about options which are best for you. Below are a few practical suggestions:
  - Raynaud's phenomenon:
    - The key first step is to keep the hands warm. Ensure you have a warm pair of mitts and plan your day to limit unnecessary cold exposure.
    - It is also important to keep your core body warm. Layered clothing can be helpful. In addition to mitts, a warm toque and scarf for cold weather can be important.
    - STOP Smoking!
    - Discuss with your physician to ensure you are not taking any other prescription medications that may exacerbate Raynaud's phenomenon.
    - If you are still having difficulties, discuss with your physician medication options which may be helpful.
  - Skin tightening:
    - While difficult to notice early on, if caught very early, there may be medications available to slow the skin progression.
    - More important to maintain flexibility through exercise and stretching of the affected areas.
  - Heartburn:
    - Discuss with your physician medication options which are available
  - Lung problems:
    - Many physicians will arrange regular screening tests to look for any early signs of lung problems prior to developing symptoms
  - Kidney problems:
    - Any problems with passing urine or a sudden rise in blood pressure could suggest a serious kidney problem. You should contact your doctor immediately.

## What is the prognosis for Scleroderma?

- The prognosis for Scleroderma can be different in every patient, based on how it presented. Patients with less skin involvement often do better than those with more

skin involvement. Those with lung, heart or kidney involvement often do worse. When possible, early treatment can be helpful.

- 5 year survival rates have been estimated between 70-90%, depending on the specific presentation of scleroderma.

*For more information on scleroderma, please discuss it further with your rheumatologist.*