

Chapter 1

Introduction

Chapter Highlights

1. This Manual is for You
2. What is Scleroderma?
3. Who gets Scleroderma?
4. What are the Early Symptoms of Scleroderma?
5. Is All Scleroderma the Same?
 - Limited Scleroderma - CREST
 - Diffuse Scleroderma
6. Working with Your Doctor
7. Some Problems You May Have Working with Your Doctor
8. What are Important Questions to ask About Medicines

This first chapter usually takes about 15 minutes.

This Manual is for You

Knowledge Can Help You Cope With Changes

Living with a chronic illness like Scleroderma is a constant source of stress.

Scleroderma is a rare illness so people don't know much about it when they are first diagnosed. Stress and worry can be made worse by not understanding your illness and not knowing what to expect.

One of the best ways to cope with an illness is to learn more about it. **Knowledge can give you a sense of control.** You can't change the illness but you won't have to cope with the unknown.

This manual was designed to help inform you about Scleroderma and to help you understand what is happening to your body. You will be better able to talk with your doctor, family and friends about Scleroderma by learning as much as you can about the disease.

You Can Learn About Main Areas of Change, One Chapter at a Time

Each chapter in this manual reviews an area of your body or your life that may be changed by Scleroderma. This first chapter is an introduction to Scleroderma. It will also teach you ways to better talk with your doctors. Each chapter has an audiotape. This tape reviews the material in the chapter with people with Scleroderma talking about their illness. After you read a chapter listen to the audiotape. Together you will learn more about your disease.

CAUTION

Try not to be afraid. Some changes may happen. Some may not. As you read this manual you may find some of the information frightening. Be careful not to think that everything we cover will happen to you. It is important to remember that everyone's experience with the illness is different.

- Your illness will also be different at different times.
- Because there are so many differences, we will discuss **all** of the possible ways in which Scleroderma can affect people.
- Many of the things described may NOT happen to you.
- Very few people will experience all of the symptoms we describe.

This manual can help. But rely on your doctor.

This manual is designed as a place for information. Only your doctor can tell you the type of Scleroderma you have. Even your doctor may not be able to tell you what symptoms you can expect to develop.

What is Scleroderma?

Scleroderma is an illness that causes hardening of the skin and other tissues of the body. It is an autoimmune, rheumatic, chronic disease that affects tissues by changing the **connective** tissue. This includes **collagen** and other extracellular component cells and structures. The word Sclero-derma means thick-skin.

What is connective tissue? Your skin, tendons, joints, ligaments, and the "capsules" around your organs can all be referred to as "connective tissue". Connective tissue is what holds you together.

What is collagen? Collagen is what connective tissue is made of. It is a kind of protein that your cells make and deposit outside of the cells in your body. Collagen is what makes your skin firm. It is also what your body makes to help heal wounds.

An Illness of the Immune System

Scleroderma is classified as an **autoimmune** disease. That means that your own immune system ends up working **AGAINST** you instead of just working **FOR** you. Your immune system is an internal "army" that fights off foreign invaders such as viruses and infections. Sometimes, something goes wrong with the immune system and it attacks your own body instead of something from the outside. This is what happens in Scleroderma. The problem in Scleroderma is that cells start making collagen as if there were an injury that needs to be fixed---but the immune attack actually **CAUSES** injury. Then, the cells don't turn off like they should and end up making too much collagen. The extra collagen can get in the way of the normal functioning of your organs.

An Illness that Varies from Person to Person

The severity of Scleroderma varies a great deal from person to person. For some people it is a nuisance - causing itching and cold fingers. For others, it can become life threatening. For most people, the illness may become more or less severe over time. Most people experience times when the illness improves and even goes into remission.

An Illness That Does Not Go Away

Scleroderma is **chronic**. It doesn't go away and lasts for your lifetime. **BUT**, it can be managed. Lots of illnesses aren't curable like diabetes, high blood pressure, heart disease and psoriasis. They differ in how severe they are but all—including Scleroderma—are **MANAGEABLE**. Unfortunately, we do not know what causes Scleroderma. We also don't have a cure. But we do have treatments that can limit the damage caused by this chronic disease.

Who Gets Scleroderma?

A few people get it, but no one really knows why.

Scleroderma is a **rare** disease. Fewer than half a million people in the United States have Scleroderma. Scleroderma has few rules about who gets it and why. It is upsetting for patients and their loved ones to learn that we don't know why someone gets



Scleroderma. Why do you have Scleroderma? Unfortunately, we can't answer that. We do know that more women than men develop Scleroderma. Some experts say that for every 7 people with Scleroderma, 6 of them are women. The most common age to develop Scleroderma is between 35 and 50 years of age. But young children and older adults can also develop Scleroderma.

Some families may be more affected by autoimmune disorders than others, but no one is sure how this happens.

Scleroderma, itself, does not seem to run in families. But family members may have other autoimmune diseases like Multiple Sclerosis, Rheumatoid Arthritis or Lupus. These diseases are also autoimmune diseases like Scleroderma. This makes scientists think there may be inherited autoimmune genes. African Americans and Native Americans generally have more severe Scleroderma. This fact also leads experts to suspect genes may play a role.



It is clear that more women get it than men.

Scleroderma happens to many more women than men. It is thought that the female hormones may play a role in the development of Scleroderma, but the evidence is not clear.

People who are exposed to certain substances may be at risk.

Things in the environment may cause Scleroderma. For example, some men exposed to silica have developed Scleroderma but for most people, silica is not a factor. Also, certain drugs seem to cause a Scleroderma-like reaction in some people.

What are the Early Symptoms of Scleroderma?

Changes in your Fingers: Raynaud's Phenomenon

The earliest symptoms of Scleroderma may be:

- fingers that become very sensitive to cold
- fingers that sometimes change color

These symptoms do not happen for everyone. But for many, these are the first hint of Scleroderma.

Most people with Scleroderma will notice changes of color in their hands when it is cold or when they are in stressful situations.

Finger changes are caused by narrowing blood vessels.

The excess collagen of Scleroderma can narrow blood vessels. This reduces blood flow to body tissues and organs.

The cold sensitivity and color changes are called Raynaud's (pronounced RAY-KNOWDS) Phenomenon.

There are two types of Raynaud's Phenomenon

Raynaud's Phenomenon is a common condition. Most people with Raynaud's Phenomenon will **NOT** develop Scleroderma. But there are 2 types:

1. **Primary Raynaud's**

People who have Raynaud's, but not Scleroderma, have primary Raynaud's

2. **Secondary Raynaud's**

A small percentage of people with Raynaud's Phenomenon develop Scleroderma. But, almost everyone with Scleroderma has Raynaud's symptoms. This is called secondary Raynaud's Phenomenon, because it is secondary to the Scleroderma disease.

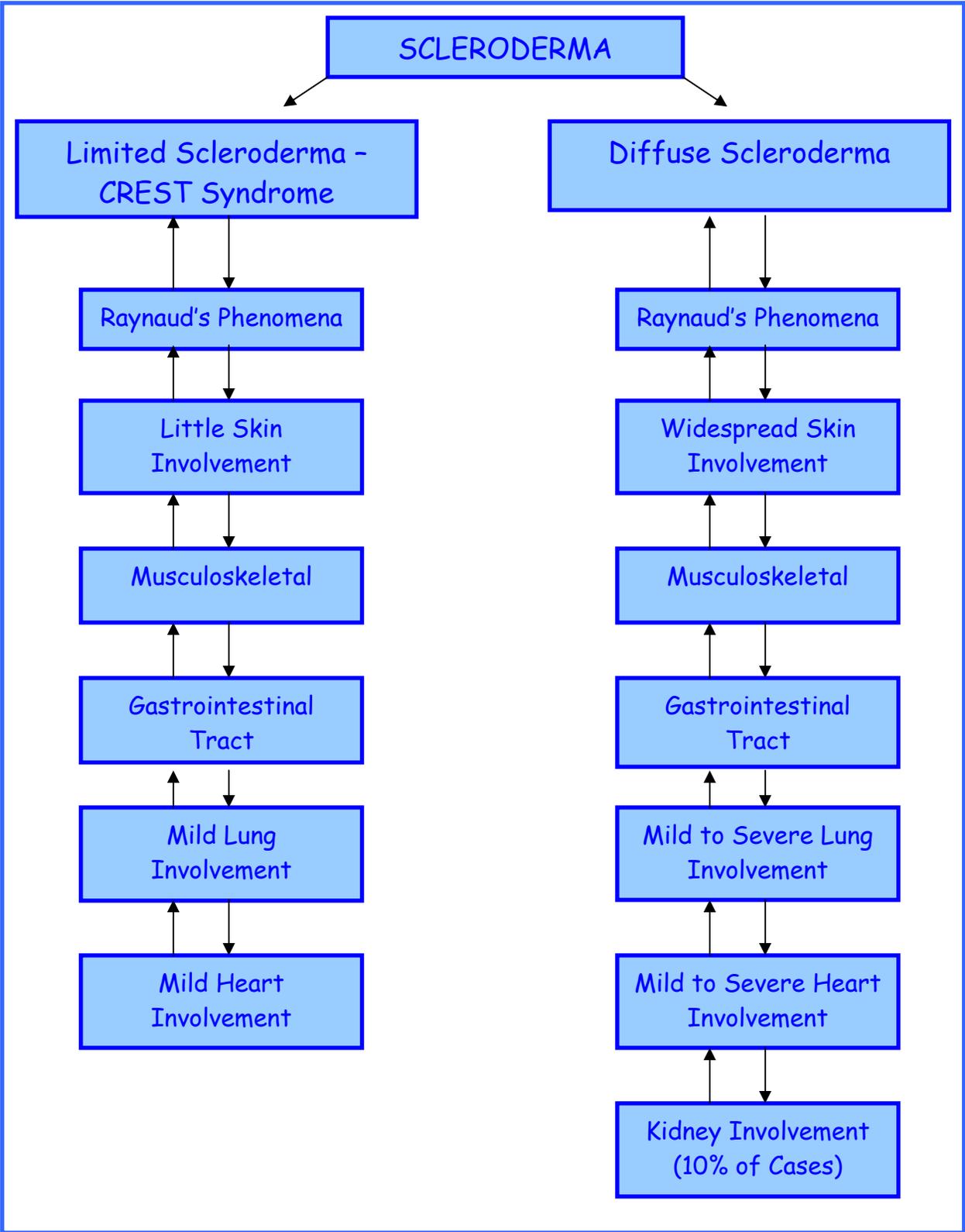
Is All Scleroderma the Same?

No. People do vary.

People vary greatly in how their illness progresses and changes over time. In the beginning of the illness, most people experience tight swollen fingers. They may also have Raynaud's phenomenon, described above. After the early symptoms, it may take months to years to see the full extent of the disease. It appears that the more skin involved in the process, the more severe the case of Scleroderma.

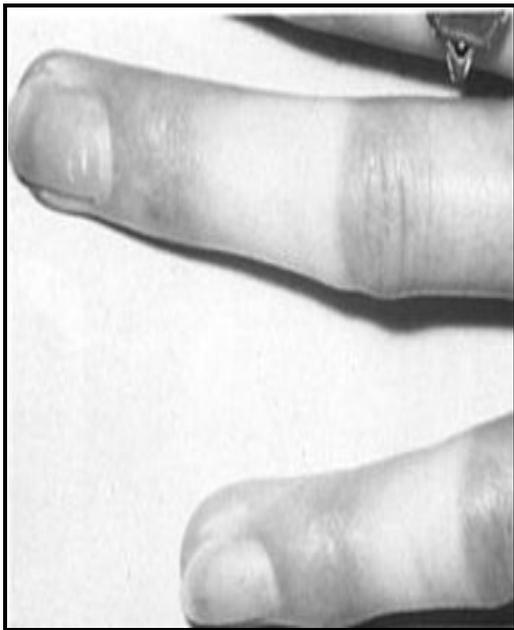
In fact, there are several types of Scleroderma

There are **two types** of Scleroderma we will discuss: Limited and Diffuse. The other types of Scleroderma - localized Scleroderma such as Morphea - are not discussed in this manual.



Limited Scleroderma-CREST

This is the **milder** form of Scleroderma and more common among Caucasians. Patients with limited Scleroderma, also known as CREST usually only have thickness of skin on the fingers. The letters in the word CREST stand for the symptoms of the disease. Everyone is different and has a different pattern of symptoms.



C is for the **Calcium** deposits in the body tissues seen in CREST.

R stands for **Raynaud's phenomenon**.

E is for **Esophageal reflux**. This is heartburn that is often experienced by CREST patients.

S is for **Sclerodactyly**; that means thick skin on the fingers.

T is for **Telangiectasias**, which are enlarged blood vessels. These appear as red spots on the face and other areas.

These symptoms will be reviewed in depth in future chapters.

Raynaud's attacks can be uncomfortable. The reduced blood supply to the fingers can also cause finger ulcers. Reflux can be quite uncomfortable. Generally, CREST does not involve the body organs. But scar tissue in the lungs can build up. This manual will provide more information about each of these symptoms.

Another Type of Scleroderma

Diffuse Scleroderma

The second type of Scleroderma is called **Diffuse Scleroderma**. It is considered to be **the most involved form** of Scleroderma.

In diffuse Scleroderma thickness of skin on the arms, legs and trunk occur, and internal organs are more likely to be involved. Tightened skin makes movement of some joints difficult. People with diffuse Scleroderma can have trouble bending their fingers, hands and other joints. They will also have a smaller mouth opening. This is caused by tightness of the skin on the face. This makes it difficult to open the mouth wide and makes it hard to close the mouth completely over the teeth.

The skin will also have areas of lighter and darker color. Some people will lose hair, have dry skin and sweat less all due to damage in the skin. Since skin care is quite important, this topic is presented in the next chapter. Diffuse Scleroderma can cause changes to your organs, such as the gastrointestinal tract, heart, lungs or kidneys. These changes will be discussed in more detail in future chapters.

Working with Your Doctor



Perhaps, you used to be fine.

Most people have little knowledge of the medical profession before becoming ill with a chronic illness like Scleroderma. Before you got Scleroderma you may have gone to the doctor for a problem like a Strep Throat. You were examined, a simple test was done and a treatment—an antibiotic—was given. You got better and didn't go back.

Now, you have an illness.

Having a chronic illness is very different. You may have seen doctor after doctor and had test after test before you were correctly diagnosed with Scleroderma. This can be frustrating, time-consuming and expensive. As medicine has become more advanced, more and more specialists are needed to give you proper care. It can be overwhelming to see a different doctor for each test or each problem but it may be necessary. There is no cure or easy treatment for Scleroderma. This means you will try lots of things with lots more doctor visits.

You need to work with a doctor.

Treating a chronic illness like Scleroderma is complicated. This is why **you, the patient, have to take an active role in your care.** The most important part is learning to communicate with your doctor. How do you this?

To get the most from your doctor, practice **CAD:**

Come prepared

Ask questions

Discuss problems

Steps for working with your doctor

1. Ask questions!

Go to your appointment with a written list of questions and concerns you want answered. That way, you won't forget to mention something.

2. Be clear about what you really need or want.

Think about exactly what you want from the visit.

3. Don't wait for the doctor to ask for questions.

He or she might not. Ask questions when you first enter the office.

4. Can't follow certain advice? Let the doctor know.

If you know you won't or can't follow the doctor's advice, for some reason, be sure to let the doctor know. For example,

- I can't take that medicine. It makes my reflux worse.
- I can't afford that medication.
- I hate to exercise.
- Physical therapy isn't covered by my insurance.

If your doctor knows why you can't or won't follow advice a different plan can be made. If you don't share your problems with your doctor there is no hope for finding a solution.

5. Don't be afraid to question your treatment.

If you have problems with your treatment, let your doctor know. Don't just stop or change doctors. Sometimes treatment of Scleroderma involves trial and error.

6. Don't be afraid to ask financial questions.

It is your right to know how much a visit costs. If a treatment is too expensive, ask about alternatives.

Some Problems You May Have Working with Your Doctor

My doctor never has time.

It is true that doctors are busy. Here's what you can do:

- **Ask for more time** when you make your appointment if you know you want to talk to your doctor. Most doctors' offices are run with a certain time planned for a brief follow-up, longer for a short visit and even longer for a physical or new visit. You can ask for as much time as you need. However, you need to ask when you make the appointment---NOT when you arrive at the office. You should expect to pay extra because that time you are using can't be used by another patient.
- **Make the most of the time you have** by going to the office prepared. Bring your written list of questions. Hand it to the doctor when you first arrive and be sure they are answered before you leave. Don't count on remembering. Most people are a little nervous when they go to the doctor so it's easy to forget.

My doctor never tells me anything about my medicines.

Your doctor may have been pressed for time. You may have forgotten to ask.

- **If you want more information ask your doctor.** If this doesn't help
- **Ask the pharmacist who fills the prescription.**

What are important questions to ask about medicines?

- What is the name of the medication?
- Do I really need this medication?
- What is the medication supposed to do?
- How and when do I take the medication and for how long?
- What foods, drinks, medications or activities should I avoid while taking this medication?
- What are the possible side effects? What should I do if they occur?

- Are there any tests I'll need to monitor the medication?
- Can I use an alternative or generic medication that is less expensive?

My doctor just tries one pill after another.

Trial-and-Error is the Only Way

Unfortunately, there is no way your doctor can know for sure what medication will work for you. You may need to try a number of medications before you find the best one. This trial-and-error method can be costly.

How to Keep the Costs Down and Your Spirits Up

- When you start a new medicine ask how long it will be until you know whether the drug will work for you. If you will know in a short time, ask for a prescription for only a week or two with refills. That way you can try the medication and if it doesn't work you don't have to throw away a bunch of expensive pills.
- Ask the doctor if he has any sample packages.
- Don't be discouraged if you have to try several different things.
- Call your doctor if you have a problem with a medication and don't have an appointment in the near future.

I don't feel comfortable talking with my doctor.

We've already talked about several ways to make communication easier but here's another important one to remember.

- **Plan to be dressed**
Plan to have a serious conversation with your doctor when you are dressed. It is hard to feel comfortable talking in your underwear or examination gown. Sometimes the personalities of doctors and patients just don't fit.
- **Find another doctor**
If you've tried to open up communications and it hasn't worked it might be time to find a doctor you are more comfortable with. Good doctor-patient relationships are important.

I can't understand my doctor.

Many doctors are so used to talking in "medical-talk" that they forget that their patients don't speak the language. They don't do this on purpose. They don't even realize they are doing it.

- The solution is simple. If you don't understand something, ask. Never be afraid to speak up.

My doctor never listens to me.

If you feel your doctor is not listening to you we suggest you discuss it.

- You can start by saying something like, "Dr. Smith, sometimes I feel you don't hear what I'm saying". This takes some courage but it will open up communication.
- Another way to get your doctor to listen is to be brief and to the point. You can practice this before your appointment. Think out exactly what you want to say.

There isn't a cure so there's nothing my doctor can do.

It is true there isn't a cure for Scleroderma. But there is a lot your doctor can do to help manage Scleroderma.

- Treatment can make you more comfortable.
- Treatment can help control the disease and keep it from getting worse.

Summary

Scleroderma is a chronic disease and the severity varies a great deal. Most people will have periods when the disease is almost in remission. There is no cure for Scleroderma, but there is a lot you and your doctor can do to take care of the symptoms. If you don't already know, you will want to ask your doctor whether you have Limited or Diffuse Scleroderma.

This first tape usually takes about 20 minutes.