Scleroderma Education Program

Chapter 2

Raynaud's, Finger and Skin Care

Chapter Highlights

- 1. Cold Fingers: Raynaud's Phenomenon
 - Tips to help Raynaud's Phenomenon
- 2. Skin Problems: Scleroderma and the Skin
 - Help for Dry Skin
 - Itching
 - Color Changes
 - Skin Ulcers

This second chapter usually takes about 20 minutes.

Cold Fingers: Raynaud's Phenomenon

In the last chapter we introduced Raynaud's (pronounced RAY-KNOWDS) Phenomenon (see figure on next page). When someone has Raynaud's phenomenon their fingertips change color when exposed to cold temperatures. In general, an attack starts with the fingertips getting very pale, becoming numb and then turning purple or blue. These symptoms can happen within minutes of being exposed to cold temperatures and will last until the fingers get warm again. Sometimes the fingers will turn red as they warm up and there may be a painful throbbing or burning sensation as this happens.

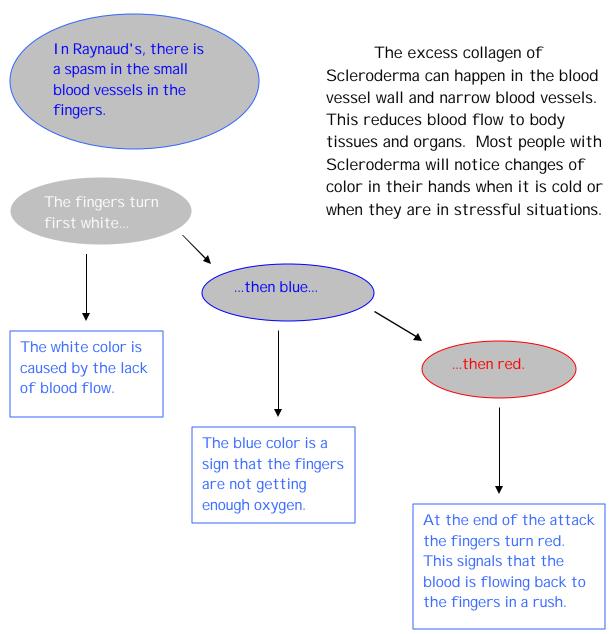
Who Gets Raynaud's?

Raynaud's phenomenon is a common condition. It affects about 1 out of 20 adults in the United States. Most of these people just have Raynaud's and will never develop Scleroderma or other connective tissue diseases. However, a small percentage of people will go on to develop Scleroderma. In these people the cold sensitivity is called *secondary* Raynaud's Phenomenon because it is due to the Scleroderma. Most people with Scleroderma do develop this kind of cold sensitivity as the first sign or symptom that something is wrong.

What Causes the Symptoms of Raynaud's?

A spasm in the small blood vessels in the fingers is what causes a Raynaud's attack. This spasm also causes the color changes. Many people also have Raynaud's attacks in their toes. The color changes of a Raynaud's attack are an exaggeration of what happens normally when hands are exposed to the cold. Everyone will develop cold hands when they are exposed to cold temperatures for a long enough time. However, the normal cold response is a blotchy red and white pattern. The color changes of a Raynaud's attack---white then blue or purple---are very different.

Raynaud's Phenomenon



Tips to Help Raynaud's

It is easier to prevent a Raynaud's attack than to stop it once it has started. So...

- Protect your fingers and toes from cold temperatures.
 This is the best way to prevent Raynaud's.
- Consider it cold outside if it's 65 degrees or lower.
 When the temperature or wind chill outside fall below 65 degrees wear protection on your hands.
- Wear mittens instead of gloves.

 Remember that mittens keep your hands warmer than gloves.

 Chemical hand warmers are helpful.
- Try to avoid sudden changes in temperature.

 Shifting temperatures may make things worse such as going from a warm outside temperature to cool air conditioning.
- Be careful, even in warm weather.
 The winter months aren't the only time to be careful of cold temperatures. Other cold things can trigger a Raynaud's attack.
- ✓ Seek peace.
 Stress or nervousness can make the response to cold worse.

THINGS THAT CAN TRIGGER A RAYNAUD'S ATTACK

- 1. Cold air, I NCLUDI NG air conditioning
- 2. Holding a cold drink
- 3. Washing something in cold water
- 4. Reaching into the refrigerator or freezer

Keep Gloves Everywhere!

Keep gloves in your kitchen when you have to reach into the freezer.

Take gloves to the grocery store where the frozen food aisles may trigger attacks.

Keep gloves in your desk at your office or at home.

Keep Your Whole Body Warm

Bring extra layers of clothing when you are going somewhere that might be cold.

Use a space heater in the bathroom or at your office at work if it gets too cold.

Get your bed warmed up. Stay warm at night by preparing your bed ahead of time. Warm it up with an electric blanket or hot water bottle. Flannel sheets are warmer than regular ones.

Have your car warmed up and ready to go before you leave the house. If you don't have a loved one to do this for you, think about a remote car starter. A lambskin steering wheel cover will help protect your hands from cold temperatures when you drive.

Reduce Stress in Your Life

Stress can trigger an attack.

Stress can also be a trigger for Raynaud's attacks. Stress causes the blood vessels in the fingers to get smaller so that more blood goes to large muscles in the arms and legs. This helps the body prepare for the stressful event but may cause a Raynaud's attack.

Learn to control stress.

Learning relaxation and stress management skills may help decrease stress-related Raynaud's attacks. Pay attention to the things that cause stress and Raynaud's attacks. Try to prepare for them.

Medications Can Help

Your doctor may prescribe medication that helps with Raynaud's (see Table 1). The most common medications are called vasodilators. The calcium channel blockers are the most effective vasodilators for Raynaud's Phenomenon. Calcium channel blockers are commonly used for blood pressure control or angina (heart vessel spasms). They also stop the spasms of arteries in Raynaud's Phenomenon. Other vasodilators can be used including topical nitroglycerin cream.

For severe Raynaud's Phenomenon with digital ulcers (discussed later in this chapter) intravenous prostaglandins are given.

Biofeedback May Help

Biofeedback has been shown to be helpful in treating primary Raynaud's. Biofeedback uses a machine to give you information on the temperature of your hands. A biofeedback therapist can teach you skills for increasing blood flow and warming the temperature of your hands. Biofeedback machines can help you become more aware of small changes in the temperature of your fingers and what works to prevent attacks. Biofeedback will be discussed more in Chapter 8.

Table 1: Medications to Prevent or Reduce Raynaud's Symptoms

| MEDICATION | BASIC MECHANISM | PRINCIPAL SIDE EFFECTS |
|---|--|--|
| CALCIUM CHANNEL BLOCKERS Nifedipine (Procardia, Adalat) Diltazem (Cardizem, DilacorXR) Veropamil (Calan, I soptin) Nicardipine (Cardene) Bepridil (Vascor) I sradipine (DynaCirc) Amlodipine (Norvasc) Nisoldipine (Sular) | Relaxes blood vessels | Low blood pressure Constipation Nausea Increased trouble swallowing Lightheadedness Headache Fluid retention |
| OTHERS Prazosin (Minipress) Terazosin (Hytrin) Doxazosin (Cardura) | Relaxes blood vessels | - "first dose" low blood pressure |
| Pentoxifylline (Trental) | Enhances blood flow to skin | - Nausea - Dizziness |
| Carbaprostacyclin (Lloprost) Investigational for Scleroderma | Relaxes blood vessels, inhibit platelets | NauseaLoose stoolsFlushingJaw discomfort |
| Nitroglycerin ointment | Relaxes blood vessels | - Headache - Flushing |

Skin Problems: Scleroderma and the Skin



Skin Becomes Thick.

The thickened skin of Scleroderma is the most noticeable sign of the disease. Some of the changes to the skin may be limited and subtle and can be missed by doctors not used to treating Scleroderma.

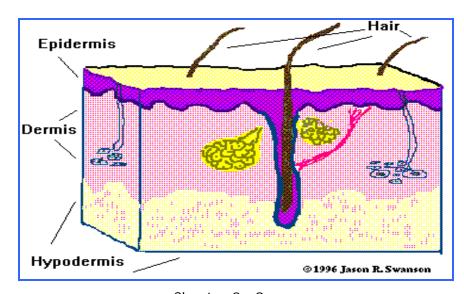
The skin thickening can be just on the fingers (this is called sclerodactyly) or it can be on the back of the hand, the forearm, the upper arm, the face, the

neck, the trunk, the legs or the feet. It usually starts on the hands or feet and may or may not spread to other areas. In general, the thickening will reach its peak within 1-2 years after it starts.

Thick Skin is Caused by Too Much Collagen.

As we learned in Chapter 1, Scleroderma means "thick skin", which is caused by too much collagen. Collagen is a normal part of skin, bones, cartilage, lungs, and just about every organ in the body. Collagen is important in helping the body repair itself after injury.

In Scleroderma, for some unknown reason, too much collagen gets made in the dermis layer of the skin. This overproduction of collagen can happen in large or small areas or may occur in a line (also called linear Scleroderma). It can start in the hands and progress to other parts of the body. It can even occur in the internal organs (also called systemic Scleroderma).



Some Cases are Mild, and Some Spread Over the Body

Unfortunately, we don't know why some people get a mild case (such as tightening in one hand) and why other people get skin thickness and tightness over their whole body.

Types of Skin Problems in Scleroderma

There are several problems that can happen with the skin of Scleroderma patients. The most common are:

- ? Dry skin
- ? Itching
- ? Ulcers
- ? Calcium deposits
- ? Discolored skin
- ? Enlarged blood vessels known as telangiectasias (pronounced TEL-an-jick-ta-zhas)

The rest of this chapter will talk about these changes and give you tips for protecting your skin.

Dry Skin

The dry skin seen in Scleroderma is caused by several things. First, the extra collagen in the inner layer of skin destroys normal sweat and oil glands. Second, the outer layer of the skin usually has fatty substances that moisten the skin. In Scleroderma this outer layer often gets thinner so there is less oil.



12 ways to put a **STOP** to your dry skin problems

1. Turn the thermostat down

Keep the thermostat set to the lowest comfortable temperature in the winter (since heat can dry out your skin).

2. Use a mild soap

Wash with mild unscented soaps or moisturizing soaps only.

3. Limit soap to certain areas

Use soap only in areas that are likely to have body odor like under the arms and groin. Be sure to rinse well.

4. Use special soaps and shampoos

Use hypoallergenic, fragrance-free cosmetics, soaps and shampoos.

5. Use warm, not hot, water.

6. Use baby oil

When you bathe try adding 1/2 cup of baby oil to the water. Be careful when you get out since the tub will be slippery.

7. Use moisturizers

Put moisturizers over your entire body immediately after all baths and showers. Water evaporating dries out the skin.

8. Moisturize often

Apply moisturizers to your hands after each hand washing.

9. Use gloves

Use gloves when using any substances that are rough on your hands. This would include detergents, soaps, salts or household cleaners.

10. Avoid excess cold or heat exposure

Both excess cold and heat can dry your skin. Use sunscreens or don't stay in the sun too long (see Table 5).

11. Avoid certain ointments and medicines

Do not use topical numbing agents (e.g. Dermoplast) or antihistamines since they tend to be very drying.

12. Avoid electric blankets

(Except to warm your bed before you get in) since their constant heat will cause you to lose body moisture.

Treat dry skin using special products:

Unscented moisturizers (see Table 2)
Unscented moisturizing soaps and shampoos (see Table 3 and 4).

PRODUCTS THAT CAN HELP TREAT DRY SKIN

TABLE 2. MOISTURIZERS

| Eucerin Cream or Lotion | |
|---|--|
| Neutroderm | |
| Lubriderm | |
| Complex - 15 | |
| Alpha - Keri Lotion | |
| Eutra Lotion (1-800-63-EUTRA) | |
| Neutrogena Hand Cream - Norwegian Formula | |
| Candermyl Cream | |
| Moisturel | |
| Aquaphor Ointment | |
| Lacticare Lotion | |
| U-Lactin Lotion | |
| Lachydrin Lotion (prescription only) | |
| Theraplex Lotion and Emollient | |

TABLE 3. BLAND TOILET SOAPS

| DRUGSTORE | GROCERY STORE |
|------------------------------------|-----------------------------------|
| Alpha Keri | Dove |
| Aveeno Bar (Dry Skin Formula) | Lever 2000 |
| Lowilla Cake | |
| Neutrogena (Dry Skin Formula) | LIQUID SOAPS |
| Purpose | Cetaphil |
| Oilatum | Cam Lotion |
| Basis Unscented Bar | Moisturel Sensitive Skin Cleanser |
| Eucerin Dryskin Care Cleansing Bar | |

TABLE 4. MILD NONDRYING SHAMPOOS

| Neutrogena Regular Shampoo | |
|----------------------------|--|
| DHS Clear Shampoo | |
| Duplex | |
| Progaine | |

TABLE 5. SUNSCREENS

| Sundown |
|--------------|
| Solbar |
| Photoplex |
| Sea and Ski |
| Pre Sun |
| Eclipse |
| Estee Lauder |
| Neutrogena |

Only use sunscreens containing a Sun Protection Factor (SPF) greater than or equal to 15.

AVOID: All sunscreens that contain Paba or Paba Ester since they may cause skin reactions.

| Choose four products that may work with your skin. | | |
|--|--|--|
| 1 | | |
| 2 | | |
| 3 | | |
| 4 | | |
| | | |

THINGS NOT TO USE FOR YOUR SKIN

DO NOT USE:

| I vory | Dial |
|--------------|-----------|
| Zest | Safeguard |
| Jergans | Lava |
| Lifeboy | Camay |
| Coast | Cuticura |
| Irish Spring | |

I tching

I tching is another major skin problem in Scleroderma. I tching is mostly a problem in patients with **Diffuse Scleroderma**. It can be caused by dry skin. It can also be from histamine.

Diffuse Scleroderma patients often have a greater number of the cells that produce histamine in their skin. This leads to itching. I tching generally occurs in the first 6 months to 2 years of the disease and then gets better. However, some people may have problems with itching for many years. There are many practical ways to prevent itching.

THINGS TO AVOID IF YOUR SKIN IS ITCHY

Avoid:

- 1. Scratchy fabrics such as wool.
- 2. Rubbing alcohol or other products containing alcohol (e.g., perfume).
- 3. Any product that contains ingredients which you are allergic to.
- 4. Harsh soaps, detergents and household cleaners (pg 2-16).
- 5. All fabric softeners.
- 6. Hot baths.
- 7. Excess cold exposure.
- 8. Vigorous exercise.
- 9. Active or passive cigarette smoke.
- Fatigue and anxiety. Get plenty of rest since these can increase itching.

TABLE 6. RECOMMENDED LAUNDRY DETERGENTS

| All Powder | |
|-------------|--|
| Dash Powder | |
| I vory Snow | |
| Safeskin | |

These products are recommended because they rinse out of clothes better. When residue is left in clothes this can cause irritation and itching.

MORE THINGS TO AVOID

DETERGENTS

Avoid:

- 1. Wisk
- 2. Lestoil
- 3. Arm and Hammer
- 4. Woolite
- 5. Cheer
- 6. Tide
- 7. Liquid Soaps
- 8. Detergents with extra power or brighteners
- 9. Strong perfumes and Store Brand soaps.

GET COMFORTABLE

There are several comfort measures that can be used to relieve itching. They include:

1. Rubbing skin rather than scratching.

Keep fingernails short to avoid damage from scratching. Damaged skin can lead to infections.

2. Taking soothing baths.

Bathe with Aveeno Oatmeal, Oilated Aveeno, Cornstarch (make a paste with 2 cups cornstarch and 4 cups water and add to bath), Mineral Oil and Cottonseed Oil.

3. Treat dry skin as recommended earlier in the chapter.

4. Use a topical anesthetic called Pramoxine.

It may be used alone or in combination with hydrocortisone 0.5%, 1%, or 2.5%. This is the only topical anesthetic that is safe to use on skin.

5. Using cooling agents.

Cooling agents such as menthol or camphor (Sarna lotion) may be used.

6. Using oral antihistamines.

This may be helpful but can also make dry skin worse.

CAUTION

Beware of drowsiness.

Some antihistamines can make you drowsy such as Benedryl, Atarax, Doxepin or Periactin. Newer antihistamines are not supposed to cause sleepiness. They are Allegra, Zyrtec, and Claritin.

Be aware of side effects.

Low dose steroids (such as prednisone) can be helpful. These medications have many side effects so they are only given as a last resort.

Color Changes

What may happen:

Skin color (pigment) changes often happen in Scleroderma. Some areas may have more pigment---get darker. This looks like a tan that doesn't fade even when you haven't been in the sun. Other areas of the skin may develop a patchy loss of skin color in a "salt and pepper" fashion. Large patches of pigment loss can also happen. This is called vitiligo.

No pain, but a change in appearance.

These changes aren't painful but are a cosmetic problem and may make you feel self-conscious or more focused on your appearance (Changes in your appearance will be reviewed in Chapter 6).

What can be done?

Prescription creams with vitamin A are sometimes given. These medications are usually used for acne and are very drying to the skin and take months to work. Skin bleaches can be used to even out areas with too much pigment. Makeup can be used to cover up the skin changes on the face and neck. You can get good advice about makeup at cosmetic counters in large department stores---ask for a makeover. Heavier cosmetics that cover better are most helpful.

It can get better.

The pigment changes get better over time and the skin tone usually returns to a more normal appearance after several years.

Sores on the Fingers or Toes (Digital Ulcers)

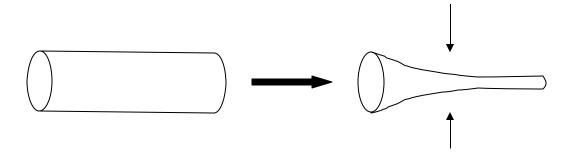
What can happen.

Digital ulcers are sores on the fingers or toes. These are seen in both CREST and Diffuse Scleroderma. They occur most often on the fingertips where they are caused by poor circulation. They can develop anywhere on the finger or toe and may be also caused by the skin being stretched too tight.

Sometimes a small cut or injury to the fingertip develops into a sore that will not heal because the blood supply is not good enough. Sores can also occur at pressure points—like over the elbows. Wherever sores occur they are very painful and can make it difficult to use your hands or other parts of your body.

How this is connected to poor circulation

In Scleroderma the excess collagen collects on the inside of blood vessels causing them to be more narrow (see figure below). This makes it harder for blood to flow.



Blood is needed to nourish the skin and other tissues of your body with nutrients and oxygen. When there isn't enough blood flowing to parts of your body – the tissues may become damaged. The areas farthest away from the heart are most often affected such as the toes and fingertips.

Another thing that can contribute to ulcers forming is Raynaud's Phenomenon. This is when the blood vessels constrict (get smaller) in response to cold or stress. This further reduces blood flow to the fingers and toes.

Preventing Digital Ulcers

There are many things that can be done to prevent digital ulcers:

1. Avoid stress and cold.

Avoid situations that bring on Raynaud's Phenomenon like stress and cold.

2. Keep as warm as possible.

Your overall body temperature impacts your finger temperature.

3. Wear layers of clothing.

4. Wear hats in cool weather.

Most of your body heat is lost through your head.

5. Wear the right shoes.

If you have trouble with your feet. Don't wear tight shoes or socks that make your circulation worse.

6. Use Band-Aids.

Using a Band-Aid can help prevent infections.

Treating Digital Ulcers

What to do if you do get an ulcer:

1. See a doctor.

Any ulceration should be seen by your doctor.

2. Keep it clean.

Keep the ulcer area clean with soap and water twice a day.

Apply a topical antibiotic (like Bacitracin) with a light fitting Band-Aid or other cover.

3. Use medicine, if it gets infected.

- ? If an infection does occur, medications are needed.
- ? There can also be a severe blockage of blood flow to the fingers. If they do not get the blood they need, the tissue may become damaged and die.
- ? Medications can help prevent ulcers by improving the blood supply.
- ? When severe infections develop, some people may lose parts of their fingers or toes.

Telangiectasias

Telangiectasias are red spots caused by the widening (dilation) of small blood vessels (capillaries) in the skin. In Scleroderma these spots tend to occur on the hands and face. In later disease they can happen on the trunk and chest. They don't cause any harm but people don't like them because they change their appearance. Laser treatments can be used to remove the spots but they tend to come back. Some people use make-up to cover the spots.

Calcinosis

Small calcium deposits can occur in the fingers, over the elbows, over the knees and in other places in Scleroderma. They start under the skin and feel hard. An X-ray can show that they are calcium deposits. They cause problems only if they are in certain areas of the hands and fingers and get in the way of hand function. Sometimes they break through the skin and drain a white material. Sometimes they can cause inflammation of the skin like that seen in gout. If open and draining they can become infected. These infections need immediate attention by your doctor. Doctors avoid operating on them to remove them because they tend to come back.

Summary

Raynaud's Phenomenon is a chronic struggle for patients with Scleroderma and many patients develop finger ulcers or problems with their skin. However, many of these symptoms can be managed by planning ahead and using the recommended products, making changes in your day to day life like keeping warm, using different products and treating sores early and very carefully. Your doctor can also prescribe medications to help prevent, improve or treat these problems.

This second tape usually takes about 15 minutes.