

RAYNAUD'S PHENOMENON AND SCLERODERMA-RELATED DIGITAL ULCERS



Information booklet for Scleroderma patients
and their family members/informal caregivers

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Pages 5 and 11 - 6.2 Appearance and description of wounds

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1

INTRODUCTION



Systemic Sclerosis (SSc) is an autoimmune disease that can affect various organs, in addition to the skin. In almost all cases, the thickening and loss of flexibility of the skin affect the fingers. Raynaud's phenomenon is very common in SSc patients. It can sometimes lead to painful fingertip ulcers.

In Quebec, it is estimated that one in 2,500 might develop Scleroderma. To this day, there is no effective treatment against the disease. Efficient management of its manifestations can prevent or delay the onset of complications that may occur as disease progresses.

This document provides you with practical information about prevention and management of Raynaud's phenomenon and SSc-related digital ulcers. It should be seen as complementary to treatments and medication prescribed by your doctor.

2

REMINDERS ABOUT THE DISEASE



What "Systemic Sclerosis (SSc)" stands for?

The term "**Scleroderma**" means

hardening (sclero) of the skin (derma). This disease is characterized by the thickening and reduced flexibility of the skin of the fingers and other body parts.



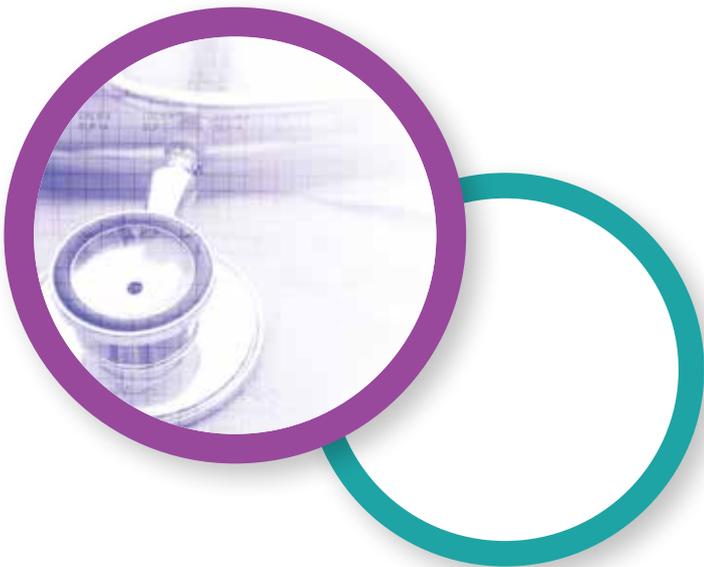
The term "**Systemic**" means

that the disease can affect, in addition to the skin, different organs (or systems):

- the heart
- the lungs
- the kidneys
- the digestive system
- muscles
- joints

What is the cause of Scleroderma?

The cause of Scleroderma is unknown. It is an autoimmune disease in which the body produces antibodies against itself. The overproduction of collagen in the skin and in some organs of the body causes their hardening, a process called "fibrosis".



Common manifestations of vascular involvement in SSc patients

1 **Raynaud's phenomenon:**

It is a disorder resulting in the reduction of blood circulation in the fingers and toes, which is typically characterized by color changes. The body's extremities can vary in colour from white through blue, and sometimes red. This is usually triggered by cold, sudden temperature changes or stress.



About 10% of the population is affected by Raynaud's phenomenon, whereas 95% of SSc patients have Raynaud's.

2 **Digital ulceration:**

It is a wound which develop mostly on the fingers. Ulcers are painful and difficult to heal. In the most severe cases, necrosis (death of tissue) and amputation may occur.

3

TYPES OF SSC-RELATED DIGITAL ULCERS

Digital ulcers must be taken seriously, because they are painful, affect everyday activities and tend to recur.

URGENT

It is essential to conduct regular self-examinations of the hands to detect the presence of ulcers and to notify your specialist physician as soon as one is suspected. Early treatment may be initiated to help reduce pain and prevent infection.

THERE ARE 3 TYPES OF ULCERS :

ISCHEMIC



Reduction of the blood flow

MECHANICAL



- Traumas
- Dryness of the skin
- Hand deformations

On CALCINOSIS



Calcinosis refers to a calcium deposit under the skin

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PREVENTING RAYNAUD'S PHENOMENON AND ULCER CARE



- Stop smoking.
- Always tell your pharmacist when you start a new medication, either prescription or over-the-counter, because some may cause or worsen Raynaud's phenomenon.
- Protect your entire body against the cold and wind. It is easier to keep your body heat than to warm it once it's already cold:
 - Cover your head and neck;
 - Protect your hands with gloves (e.g. silk gloves in summer, mittens in winter);
 - Use gloves with hand-warmer packs or heated mittens in winter;
 - Wear footwear with thick soles and waterproof seams in cold weather;
 - Dress in layers, according to planned activities, for optimal moisture transfer and body heat retention.



- Avoid sudden temperature changes (e.g. air conditioning, cold water, food in the refrigerator or freezer, cold metal objects) by protecting your hands with gloves.
- Identify and avoid potential traumas for they may contribute to the development of ulcers:
 - Typing on the computer keyboard, sewing, gardening...;
 - Cooking, tinkering, cutting nails...
- Learn to manage stress because it can trigger Raynaud's phenomenon.

Despite these precautions, if Raynaud's occurs, quickly put your hands in a warm place (e.g. warm water or under your armpits) to improve blood circulation.

5

SKIN CARE



It is essential to provide special care for your skin. Ulcers on the fingers can be a source of pain, stress or frustration as they can affect all activities of daily living. Here are some ways to help you take care of your skin.

Products recommended by my healthcare team:

Skin Care

DOS

Use mild soaps or soap-free cleansers

Moisturize the skin every day during daily cleansing

Massage the skin to soften it with fragrance and allergen-free sensitive skin creams

Remember to moisturize the skin after each hand washing

Do regular exercises to fight against joint deformations, according to the recommendations of your specialist physician and occupational therapist

Wear an orthosis, if prescribed by your specialist physician

DON'TS

Use foaming scented soaps because they may dry out the skin and cause irritation

Take hot baths because by macerating the skin it could further compromise the skin integrity

Dry your hands by rubbing them together vigorously: blot them up gently instead

Use antiseptic gels because they can dry out the skin

6

PATIENT INVOLVEMENT IN ULCER CARE



6.1 Describing wound's appearance

Skin acts as a natural barrier against the external environment. When an ulcer occurs, a natural repair process begins: **wound healing (i.e. scarring)**. Your role in caring for digital ulcers is to:

NOTIFY

your specialist physician
as soon as an ulcer occurs
so that he/she can assess
it and start or adjust
treatment, if needed

Learn how

TO DESCRIBE THE APPEARANCE

of your wounds

6.2 Appearance and description of wounds

	CLASSIFICATION AND SIGNIFICATION	PURPOSE OF TREATMENT
	<p>Yellow Presence of yellow or grayish debris, sometimes moist.</p>	Cleaning the wound to eliminate the debris.
	<p>Red Healthy, bright red, shiny tissue. This indicates that wound healing is well under way.</p>	Stimulating the healing process.
	<p>Pink Pinkish or lavender, shiny tissue, pearlescent and fragile in appearance. The wound is now closed.</p>	Protecting the healing wound site.
 <p>The appearance of ulcers shown below is a warning sign indicating that you should be quickly assessed by a physician.</p>		
	<p>Black The wound is covered by a black or brownish crust with a leather-like texture. There is a risk of losing part of the finger.</p>	Cleaning the wound to eliminate this type of tissue.
	<p>Green The wound is infected. There is redness, a sensation of heat, discharge and increased pain. A bad smell can also emanate from the wound.</p>	Cleaning the wound and treating the infection.

6.3 Ulcer care



If the pain is too strong

take the prescribed pain medication 30 to 60 minutes before beginning care



Wash your hands

With a mild soap under tap water



Dry your hands

With a disposable towel (e.g. "paper towel") or a clean cloth towel

Blot them up gently without rubbing



Clean the ulcers to remove debris

With physiological saline solution and dry them with a clean gauze pad

Cover with the recommended dressing



The choice of dressing is very important to **promote wound healing**.

The idea is to keep ulcers moist (but not too much!) and protect them from traumas. The dressings prescribed by your doctor or nurse are chosen according to the appearance of your ulcers.

Recipe to prepare physiological saline solution at home:

1/2 teaspoon of salt in 250 ml of water boiled for at least one minute. This solution can be stored for 24 hours at room temperature.

Care and dressings recommended by my healthcare team:



TIP:

Avoid applying antiseptics, disinfectants and creams or antibiotic ointments directly on the wounds unless advised by your doctor.



REGULAR ULCER SELF-MONITORING



To closely monitor the evolution and promote optimal treatment for your ulcers, it is strongly recommended that you keep a Self-Monitoring Weekly Log (see page 18).

- 1 Number each ulcer
- 2 Enter assessment date for each ulcer
- 3 Describe the appearance of each ulcer
- 4 Note the type of dressing being used
- 5 Keep the recommended weekly log provided in Annex until your ulcers have fully healed



TIP:

If you can, take pictures of your ulcers once a week in order to facilitate follow-up assessments by your healthcare team.

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CHECKLIST

Remember that it is essential to:

Protect	your hands from the cold and traumas
Moisturize	your skin with fragrance-free, sensitive skin products
Consult	your specialist physician promptly when an ulcer occurs
Take	time to record the appearance of each of your ulcers and initiate proper treatment

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TO LEARN MORE

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REFERENCES

- Berezne, A., et Mouthon, I. (2010). Prise en charge des ulcères digitaux dans la sclérodémie systémique. *Réalités en rhumatologie*, 28(1): 37-41.
- Brown, S. (2010). Management of digital ulcers related to systemic sclerosis. *Nursing Standard*, 24(32): 53-62.
- Bryant, R. A. & Nix, D. P. (2012). *Acute and chronic wounds: current management concepts*. 4th Éd. St-Louis, Missouri: Elsevier Mosby.
- Francès, C., Allanore, Y., Cabane, J., Carpentier, P., Dumontier, C., Hachulla, E., ..., Sibilia, J. (2008). Prise en charge des ulcères digitaux de la sclérodémie systémique. *Presse Médicale*, 37(2): 271-285.
- Hughes, M., Ong, V. H., Anderson, M. E., Hall, F., Moinzadeh, P., ..., Herrick, A. (2015). Consensus best practice pathway of the UK systemic sclerosis study group: digital vasculopathy in systemic sclerosis. *Rheumatology*, 54(11): 2015-24. doi: 10.1093/rheumatology/kev201. Epub 2015 Jun 26.
- Joyal, F. (2015). Enfin l'hiver. Longueuil, Québec: Sclérodémie Québec. Récupéré de <http://sclerodermie.ca/wp-content/uploads/2015/01/Fascicule-Enfin-lhiver.pdf>
- Lok, C., Mouthon, L., Ségard, M., Richard, M. A. et Guillevin, L. (2011). Les ulcères digitaux de la sclérodémie systémique. *Annales de dermatologie et de vénéréologie*, 138: 762-768.
- Ordre des Infirmières et Infirmiers du Québec [OIIQ]. (2007). *Les soins de plaies: au coeur du savoir infirmier*. Westmount, Québec : OIIQ.
- Sibbald, R. G., Orsted, L., Coutts, P. M., & Keast, D. H. (2006). Recommandations de pratiques exemplaires pour la préparation du lit de la plaie: mise à jour 2006. *Wound Care Canada*, 4(1): 73-86.

ULCER SELF-MONITORING WEEKLY LOG



		year/month/day	year/month/day	year/month/day	year/month/day	year/month/day
Ulcer/Site N° _____ Site: _____	Date:					
	Appearance					
	Dressing					
Ulcer/Site N° _____ Site: _____	Date:					
	Appearance					
	Dressing					
Ulcer/Site N° _____ Site: _____	Date:					
	Appearance					
	Dressing					

ULCER SELF-MONITORING WEEKLY LOG



year/month/day	year/month/day	year/month/day	year/month/day	year/month/day
Date : _____				
Appearance _____				
Dressing _____				
Date : _____				
Appearance _____				
Dressing _____				
Date : _____				
Appearance _____				
Dressing _____				

Ulcer/Site
N° _____
Site: _____

Ulcer/Site
N° _____
Site: _____

Ulcer/Site
N° _____
Site: _____

SCLERODERMA QUEBEC

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Please don't hesitate to contact us.

You wish to advance promising
Scleroderma research
and help provide support to SSc patients?

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