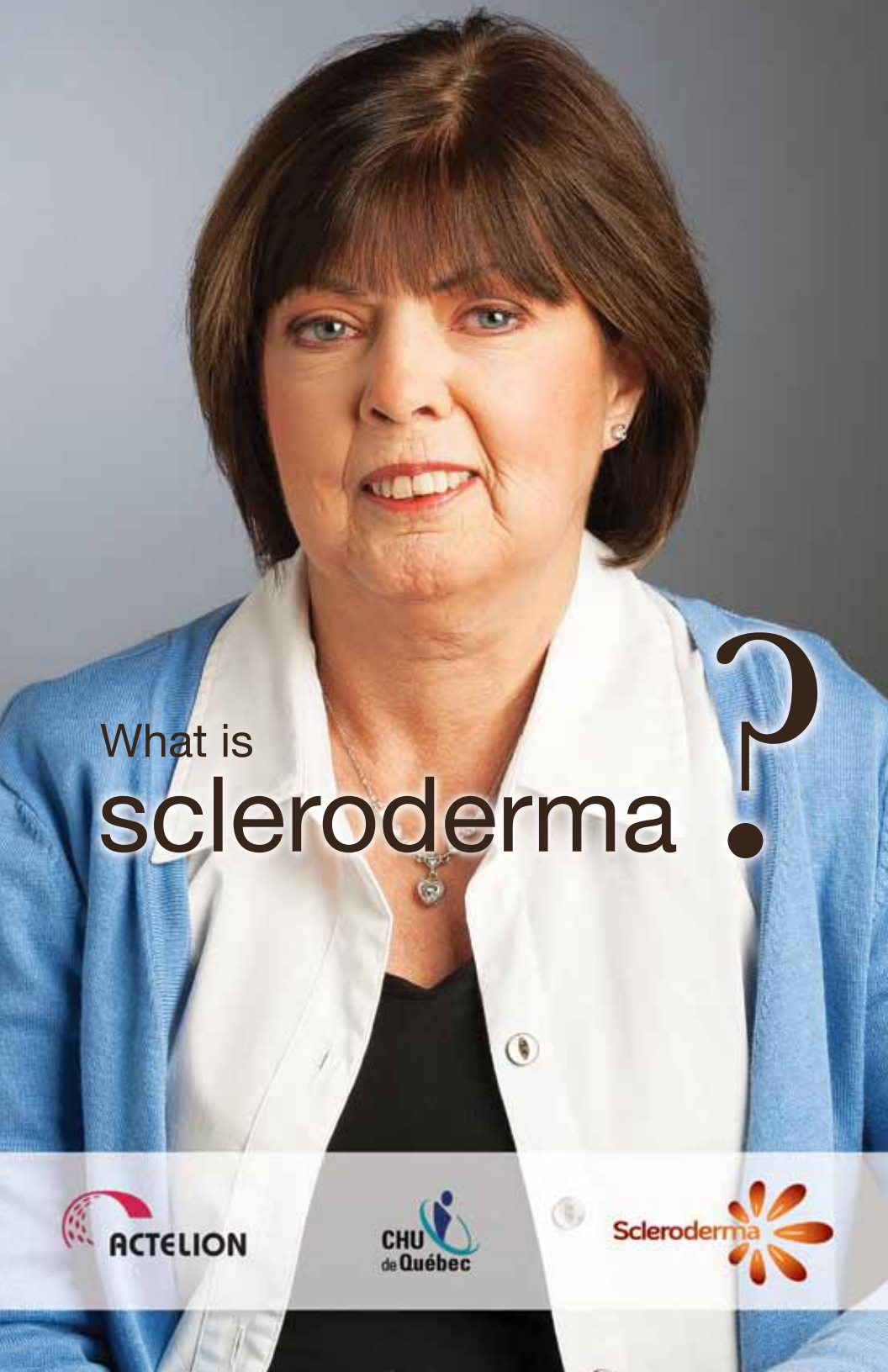

What is
scleroderma ?
ACTELION
CHU de Québec
Scleroderma

What is scleroderma?

Scleroderma, or systemic sclerosis, is a relatively misunderstood chronic disease affecting about four people per 10,000 of population. In Quebec, it is estimated that several thousands of people suffer from scleroderma.

Disease onset usually occurs in the fourth decade of life and is five time more prevalent in women than men. Scleroderma is one of the so-called “autoimmune” diseases in which the body’s antibodies attack its own cells. Scleroderma is not a contagious disease.

Scleroderma Quebec wishes to thank Dolorès Dupuis who has generously granted us the permission to publish her picture.

Dolorès is in charge of the Mauricie support and self-help group chapter of Scleroderma Quebec.

SCLERODERMA

The disease is characterized by the overproduction of collagen and damages to small blood vessels that causes excessive scaring in multiple internal organs. The overproduction of collagen causes an hardening (fibrosis) of the skin and, sometimes, internal organs, such as the lungs, heart, kidneys and the gastrointestinal tract.

There are two types of scleroderma:

- limited scleroderma (or **CREST** syndrome, which includes **C**alcinosis, **R**aynaud's phenomenon, Esophageal dysmotility, **S**clerodactyly and Telangiectasia);
- diffuse scleroderma.

The distinction between these two types of scleroderma is based on the pattern and severity of skin and internal organs involvement, which is more extensive and have a more rapid progression in the diffuse type. The skin, digestive system, heart, lungs and kidneys are the most commonly affected organs that can be negatively impacted by the potentially serious complications of scleroderma. At first, these complications may go unnoticed.

There is still no cure for scleroderma. However, medical treatments currently exist for most of the complications associated with scleroderma, hence the clinical importance of screening for patients, even in absence of symptoms.

To learn more about the most common complications of scleroderma, we encourage you to **familiarize yourself with the warning signs that you and your loved ones should regularly be watching for.**

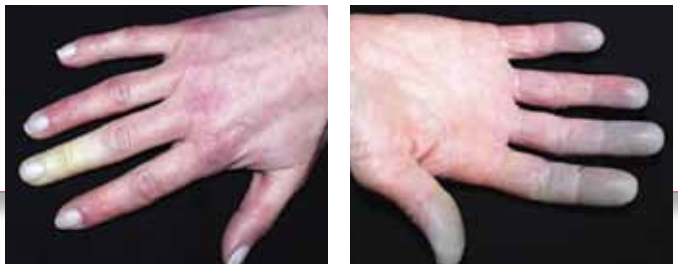
THE SKIN

An abnormal and temporary decrease in the size (calibre) of blood vessels In fingers and/or toes triggered by exposure to cold or emotional stress is referred to as Raynaud's phenomenon. The narrowing of blood vessels can cause a decrease in blood flow that manifests itself as a blanching of fingers and/or toes which may be associated with numbness. **Raynaud's phenomenon** is present in over 95% of patients with scleroderma, but can also be seen outside of this disease. Treatments and medications are currently available for Raynaud, when indicated.

It is to be noted that the prolonged narrowing (constriction) of blood vessels can cause painful ulcers that may be very slow or difficult to heal because of poor circulation, and in rare cases might even require amputation.

Thus, it is recommended to inform your doctor if you notice:

- **An increase in the severity or frequency of Raynaud's symptoms.**
- **The presence of a wound at the extremity (pulp) of a finger and/or toe which is slow to heal or is accompanied by a discharge.**



Raynaud's Phenomenon

To limit this complication, we suggest that you examine your hands and feet regularly (especially the pulp), in addition to:

- Avoid trauma to the fingers and toes;
- Wear two pairs of mittens, socks, and warm boots when it is cold;
- Always cover your head outside during the cold season;
- Use lukewarm water in your daily tasks;
- Stop smoking;
- Apply moisturising cream on dry skin.

Calcinosis is characterized by calcium deposits in the skin, which occur just below the skin surface in the form of hard lumps or nodules. It is found mainly around the pulp of the fingers or the joints.

It is recommended to inform your doctor if you notice:

- **An ulcer or discharge of pus at the site of the calcinosis.**

THE DIGESTIVE SYSTEM

Involvement of the digestive system, particularly the esophagus (swallowing tube), occur in nearly all SSc patients.

Gastroesophageal reflux is acid regurgitation in the esophagus that manifests itself by a burning sensation radiating up to the throat after meals. If left untreated, it may be further complicated by an inflammation of the esophagus and lead to a narrowing (stricture) which might then require esophageal dilatation. In rare cases, it may develop into cancer of the esophagus.

It is recommended to inform your doctor if:

- **You have a persistent sensation of blockage in the esophagus when you swallow. He/she will determine if you need a gastroscopy and if your digestive condition requires treatment.**

GASTROSCOPY RECORD

Date	Comments

THE LUNGS AND ITS BLOOD VESSELS

An abnormal elevation of blood pressure in the pulmonary arteries, also known as **pulmonary arterial hypertension (PAH)**, develops in 10% of SSc patients. This is a serious complication that can quickly lead to death if left untreated. Although patients with pulmonary arterial hypertension are usually short of breath on on exertion, **screening** for this condition is critical and should be done regularly **even in absence of symptoms**. This screening helps your doctor determine if your pulmonary condition requires treatment. Regular echocardiogram is recommended; — in most cases, yearly.

The lungs can also suffer from a condition called **pulmonary fibrosis (PF)** which can manifest itself as shortness of breath or dry cough. To check for early signs of PF, your physician might prescribe chest X-rays and pulmonary function tests.

Inform your doctor immediately if you notice any of the following symptoms:

- **An increased shortness of breath while carrying out activities of daily living.**
- **Episodes of dizziness (lightheadedness) and/or loss of consciousness.**
- **Chest pain on exertion.**
- **Swelling of the legs.**

ECHOCARDIOGRAM RECORD

Date	Comments

CHEST X-RAY RECORD

Date	Comments

PULMONARY FUNCTION TESTS (PFTs) RECORD

Date	Comments

THE KIDNEYS

The most severe complication affecting the kidneys, called **scleroderma renal crisis (SRC)**, which occurs in 5% of SSc patients, manifests itself as an increase in blood pressure. This may be accompanied by shortness of breath, headaches, visual changes or an altered state of consciousness. Patients experiencing this complication usually require temporary, or even sometimes, permanent dialysis. It should be noted that those who take cortisone (Prednisone) or have diffuse skin involvement are at greater risk of developing SRC.

We recommend that you **take and record your blood pressure** at least once a month, or more often according to your doctor recommendations. We also recommend that you show your blood pressure readings (numbers) to your doctor so that he/she can determine whether medical treatment is required in your case.

It is recommended to inform your doctor:

✶**If your blood pressure is consistently higher than usual**
(Normal blood pressure is less than 140/90 mmHg.)

✶**Be sure to inform any physician who is considering prescribing you cortisone that you suffer from scleroderma.**

BLOOD PRESSURE RECORD

Date	Time	Systolic (1st number)	Diastolic (2nd number)	Pulse

CHECKLIST

Medical treatments currently exist for most of the complications of scleroderma mentioned in this document. Therefore, it is important to screen **all patients** for these problems, even in the absence of clinical symptoms, so that appropriate treatment can start as soon as possible.

I immediately inform my doctor if:

- I notice an increase in the severity and/or frequency of Raynaud's symptoms.
- I notice a sore that does not heal or presents a discharge flow.
- I notice a recurring blockage in the eophagus when I swallow.
- I feel more short of breath or dizzy while carrying my usual everyday activities (e.g. being unable to do activities with friends, difficulty keeping up with a task, such as answering the phone, doing housework, etc.).
- My legs are swollen.
- My blood pressure is consistently higher than usual.

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Rheumatology Department, CHU of Québec, nov. 2011

You have scleroderma?
You know someone who has the disease?
You want more information?
You wish to receive our newsletter?

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