Scleroderma Canada | Sclérodermie Canada

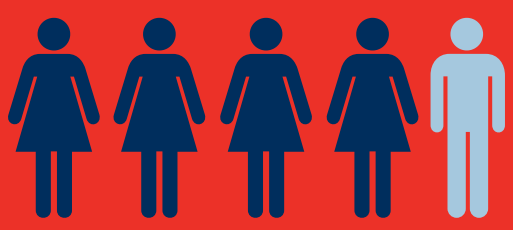
WHAT IS SCLERODERMA?

Scleroderma (also known as systemic sclerosis) is a rare, chronic, multi-symptom autoimmune disease that affects the body's connective tissue. Connective tissue (fibrous cells and proteins such as collagen) provides strength and support to structures including skin, blood vessels and internal organs. In scleroderma, a person's immune system works against itself resulting in excess tissue collagen throughout the body. This overproduction of collagen can prevent the body's organs from functioning properly. In some forms of scleroderma, patches of hard, tight skin are the extent of this irregular collagen over-production process. In the systemic forms, the problem goes much deeper, and can severely affect blood vessels and internal organs such as the heart, lungs, kidneys, and digestive tract.

WHO GETS SCLERODERMA?

Scleroderma is not contagious, not infectious, and not cancerous. Although scleroderma is more common in women (average age of onset is 25 to 55 years), scleroderma also occurs in men and children, from infants to the elderly. It affects people of all races and ethnic groups, but Indigenous persons and persons of African ancestry have a higher incident rate of scleroderma than Caucasians. Men who develop scleroderma get sicker quicker, and they have a poorer long-term prognosis compared to women.

Women are **4 times** more likely to get scleroderma compared to men.



Worldwide, **2 million** people live with scleroderma.

In Canada, **1 in every 2,500** persons live with scleroderma.

LOCALIZED

Scleroderma primarily affecting the skin without major organ involvement, is referred to as localized scleroderma. Although this type of scleroderma typically develops in children, it has a much better long-term prognosis.

2

TYPES

SYSTEMIC

Scleroderma that affects the skin, as well as the underlying tissues, is referred to as systemic sclerosis. It is characterized by vascular lesions in the tiny blood vessels and major organs, which often leads to organ damage. Symptoms of systemic sclerosis tend to be more severe with less desirable long-term outcomes.

WHAT ARE THE SYMPTOMS OF SCLERODERMA?

Symptoms of scleroderma vary greatly from individual to individual, and the effects of scleroderma may range from mild to severe. The seriousness of the illness will depend on what parts of the body are affected and the extent to which they are affected. Some people experience tight skin, swollen fingers, sensitivity to cold in fingers and toes, and extreme fatigue. Others may have no outer skin affected but instead, the disease may cause serious damage to internal organs. With time, internal organs such as the kidneys, lungs, digestive tract, and the heart can sometimes become involved. To look out for these complications, your rheumatologist will invite you to have regular screening assessments done for bloodwork, lung (pulmonary) function testing, and heart testing with electrocardiography (ECG) and Echocardiography (ECHO).

SYMPTOMS MAY INCLUDE:

*Symptom statistics are taken from results of a 2019 Canadian Patient Scleroderma Needs Assessment Survey.

Skin: **9/10** Canadians experience gradual thickening and tightening of the skin on face, hands, arms, and sometimes the whole body.

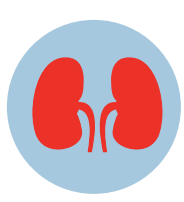
Raynaud's Phenomenon (RP): **9/10** Canadians experience extreme sensitivity to cold, which causes the blood vessels in fingers and toes to constrict which leads to reversible digital discolouration; almost 98% of persons with scleroderma also have RP.

Digestive (Gastrointestinal) Issues: **6-8/10** Canadians experience common symptoms which include swallowing difficulties, poor absorption of nutrients, constipation, diarrhea, incontinence, and gastroesophageal reflux disease, a condition that causes a "backwash of stomach acid".

Joints, Muscles, Pain & Fatigue: **8-9/10** Canadians experience stiff, sore, swollen joints, severe fatigue, muscle weakening, and pain.

Sjogren's Syndrome: **8-9/10** Canadians experience dry eyes and mouth due to decrease in secretions from the tear ducts and salivary glands; dry mouth can cause difficulties in swallowing and tooth decay.

Lungs: **8/10** Canadians experience fibrosis and scarring in the lungs and weakened respiratory muscles which leads to shortness of breath and persistent coughing; fibrosis affects oxygen absorption and may lead to Pulmonary Arterial Hypertension. Another serious complication with poor prognosis is Interstitial Lung Disease.



Kidney: 2/10 Canadians experience high blood pressure and excess protein in the urine can be signs of renal crisis which may quickly lead to kidney failure.

Heart: The heart muscle can become thickened and scarred, and may result in fluid retention, palpitations (irregular heart beats), shortness of breath, and in some cases, heart failure. This equates to **2/10** Canadians experiencing common heart symptoms.

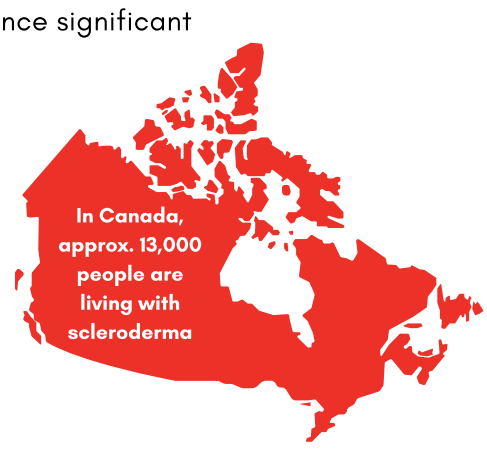
WHAT TYPE OF TREATMENT IS AVAILABLE?

Although at present, there is no cure for scleroderma, there are effective treatment options to help alleviate symptoms and slow down disease progression. Current medications are designed to treat scleroderma by targeting four key factors: inflammation, autoimmunity, vascular disease, and tissue fibrosis. Due to the highly variable nature of the disease, treatment is often simultaneously managed by several specialists. Typically, primary disease management is provided by rheumatologists that specialize in the treatment of autoimmune diseases and musculoskeletal disorders. Depending on disease progression, other specialists may become involved such as nephrologists (for kidneys), cardiologists (for the heart), gastroenterologists (for the digestive tract) or pulmonologists (for the lungs).

WHAT DO WE KNOW ABOUT SCLERODERMA?

Scientists are certain that scleroderma is not contagious, not infectious, and not cancerous. Studies of twins also suggest that scleroderma is not inherited. Although the cause of scleroderma is unknown, and currently, there is no cure for scleroderma, there are important treatments that can help slow the process down and improve the quality and quantity of life for persons affected by the disease. More painful, debilitating, disfiguring, and life-threatening than other autoimmune diseases such as rheumatoid arthritis or lupus, scleroderma can affect every part of a person's body. Often, this leads people with scleroderma to experience significant disability, reduced quality of life, depression, and premature death.

Currently, the life expectancy for people with systemic scleroderma is a little over 10 years. The leading cause of death for people with scleroderma is lung failure due to interstitial lung fibrosis (scarring) and pulmonary hypertension. In Canada, approximately 13,000 people are living with scleroderma, many of whom do not have equitable access to appropriate medical treatment and community supports.



WHAT MEDICAL CHALLENGES DO PEOPLE LIVING WITH SCLERODERMA FACE?

Because scleroderma is a rare disease, often people suffer for years with its devastating symptoms before getting a proper medical diagnosis. This is problematic since scleroderma treatments work best to slow down the disease progress if administered within the first 6-months of disease onset. Although there are new medications that are coming to market and have proven to slow down the progress of serious scleroderma symptom complications that affect the lungs, heart, and kidneys, the costs of these drugs are expensive and can range up to $30,000/month. Because these medications are not covered under Provincial and Federal Government Health Benefit Programs, most people with scleroderma cannot afford them. Although some people may have access to these medications through clinical trials, once the trials end, continued treatment access is challenging, if not impossible.

WHAT SOCIAL CHALLENGES DO PEOPLE LIVING WITH SCLERODERMA FACE?

Because scleroderma is a rare disease, it comes with loneliness and isolation. People feel overwhelmed and distressed in the absence of information, support, and an understanding peer group. Social stigma and misunderstanding about scleroderma often lead people with the disease to experience marginalization and significant barriers to societal participation. Identifying meaningful ways to participate in and contribute to community life is challenging for many people with scleroderma. For Canadians with scleroderma living in rural/remote and northern populations this issue is even more pronounced due to the lack of resources available in these areas.

VOICES

I felt lost, hopeless, like a failure in life, to my family and as a man.

JOE C.

Everyday I have to push myself.

TAMANYA J.

My Respirologist suggested a lung transplant.

PAUL O.

Sometimes I feel "invisible" since many people don't realize how sick I am.

LIZ V.

NEXT STEPS IN CANADA

Everyone deserves the right to be part of a vibrant community where they may contribute, feel valued, recognized, and in turn, lead others to live more fulfilling and engaged lives. For persons living with the rare and debilitating condition of scleroderma, performing everyday tasks (e.g., dressing, cooking, grocery shopping, childcare), not to mention work-related responsibilities, poses significant physical and emotional challenges—resulting in feelings of helpless, hopeless, and being a burden to others.

Scleroderma Canada needs help from the Provincial and Federal Government of Canada to remove barriers and increase equitable access for persons with scleroderma to 1) affordable life-saving medications and treatments and 2) meaningful opportunities to participate in and contribute to community life.

This means:

- Quicker market authorization/approval of scleroderma drugs.
- Assurance that drugs for scleroderma, as a rare disease, are included as part of a national pharmacare program.
- Open-label drug extensions to enable people participating in clinical trials to receive continued treatment access after trial completion.
- Improved patient access to specialized treatment centers in northern Canadian communities.
- Specialized community funding through the Provincial and Federal Government Grant Programs for the development and expansion of community outreach, support group programing, and educational activities for persons living with scleroderma. Community support programs provide an important connection that will help people diagnosed with scleroderma learn to manage the variety of issues that affect their daily lives.

To learn more about scleroderma, including ways you can help improve the lives of people living with scleroderma, please contact:
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