SCLERODERMA ASSOCIATION OF B.C.

The Bulletin

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Scleroderma Association B.C.

Nutrition *and sclérodermie*

Nancy's story

The Scleroderma Patient-centered Intervention Network (SPIN)

Summary

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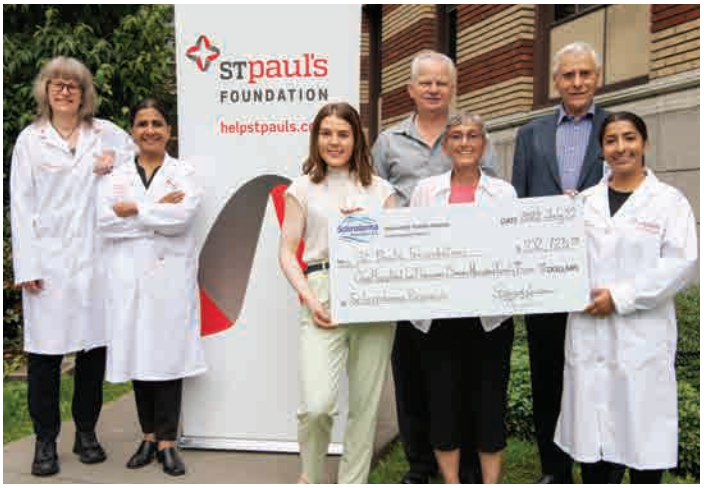
A Word from Our President

Three years ago, the world took a dramatic turn with the eruption of the coronavirus, adversely impacting people across this planet. Luckily, I have not yet directly crossed paths with COVID-19 but know of many people who've had to battle through its effects. For us that are immune compromised, practicing caution and diligence remains in our best interest. Staying safe and healthy.

With the past several years having to be 'virtual', this year's 'in-person' **Moving to Cure Scleroderma** campaign was a huge success. We ran live events across the province. I was especially pleased to be able to attend the inaugural walks in Kamloops and Surrey; meeting scleroderma patients and their family members who were not previously aware that SABC even existed. Some attendees were newly diagnosed, some had lost loved ones and family members and wanted to honour them by their presence at the walks and even their friends of friends joined us. The stories I heard just confirmed how far-reaching our efforts are in spreading awareness and educating the public. For me, this confirms the positive impact individuals and the SABC have on the community, making a difference in patients' lives. Similar heart-warming experiences were felt at both the Scleroderma Ride for Research in Stanley Park and the walk held in Victoria

Thank you to all the organizers, participants, volunteers, and donors who made this the best year to date. We collectively expanded the awareness of this disease and raised over $100,000 across the province for the SABC Research Project.



While remaining aware of the risks of COVID-19 to our members, and the population in general, SABC has expanded outreach activities using social media, virtual presentations, and online support group meetings. Although we still have members who meet face-to-face over a coffee, our zoom meetings allow people who may live in rural areas or not able to join in person, to attend what is offered. Our professional presentations and Valerie's "Scleroderma Education Podcast" series; two great examples of information available to everyone.

We had a very successful AGM & Conference in October. Dr. Leslie Laing's presentation on the Management & Treatment of Oral Complications Associated with Dry Mouth was very informative. She offered us many tips on how to better manage our Sjogren's symptoms. Dr. Marie Hudson's presentation on the Progress in Cellular Therapies for Scleroderma provided an important update on stem cell transplants. With the results of her ongoing research, we hope that in the future, cellular therapies will be less invasive for the patient. Both presentation videos are on our website, and I encourage you to take the time to watch them.

I hope that with winter upon us, you will continue to still stay connected with your friends and family. Covid is here to stay apparently and is now our new normal. We will have to be diligent in staying safe while continuing to still have a life!

June is Scleroderma Awareness Month!

All across BC, over 5 teams within the Scleroderma community moved together to raise awareness and fund research initiatives towards finding a cure. We raised $102,723.09 for scleroderma research. Congratulations to all for a successful June Awareness Campaign.

SURREY

We had an awesome first Surrey Walk on June 26th! Thank you to everyone who participated this year. Almost $8,500 was raised as our community united with a goal to find a cure. Over 50 people joined us for the 5 km walk in the sunshine. We are humbled and grateful for the support we received. Thank you to the dedicated team of volunteers who helped with the organization and set-up; the enthusiastic participants; and all those who generously donated to help patient driven research for scleroderma. It was an incredible day and left our hearts full!

- CHELSEA FITZPATRICK-LINDSAY & KELLY GRANT

KAMLOOPS

Kamloops, we did it! Our wonderful city hosted the first ever walk for scleroderma in history! Not only did we create amazing awareness in our community for scleroderma **but we crushed our fundraising goals and raised $7,760.49!** With the support of our 5 local sponsors, walk participants (local and out of town), family members, SABC, City of Kamloops, local media, and small businesses who kindly shared our posters in their stores, a lot of "amazingness" happened June 5th at Riverside Park! I would like to extend a special thank you to all our volunteers who worked really hard the past few months to help make this event successful and fun! My heart is full of gratitude as I watched our community show up for scleroderma! Thank you Kamloops. I am proud to call this city my home.

- JEN BECKETT

VANCOUVER RIDE FOR RESEARCH

Having the 11th Scleroderma Ride for Research "in person" was just wonderful. Over 50 people showed their support by riding the 30 km route, walking the 10 km around the seawall or just sitting and chilling out at the picnic site. Conversation was enjoyed over our potluck lunch, with people catching up with each other. It was great to see everyone after 2 years of Covid. We are humbled by the support we receive. **With the help of our team, we raised $65,723.50.** I always find that fundraising is a lot easier than people actually think, because people do care and they know how important it is to support research.

- ROSANNE & DAVID

VALEMOUNT & ACROSS B.C.

The province-wide, virtual, Moving to Cure Scleroderma had another successful year. **We raised over $9,300.00 for research!!** So proud of our Teams (Beth Miller: Squamish, Mimi's Minions: Duncan and Teressa Colosimo: Valemount). In June, it doesn't matter where you in live in our Province. We all wear our blue shirts, gather donations and move together, raising awareness for Scleroderma.

- TERESSA COLOSIMO

VICTORIA

Being back in person for the 2022 Victoria Scleroderma Walk, after a two year pandemic hiatus, made the 5th Walk a happy day for reconnection and meeting new friends. On a lovely sunny day, approximately 100 people took part, including those living with Scleroderma, their families and friends, volunteers and support personnel. We were delighted to be joined by Dr. James Dunne who spoke to the group about the progress and hopes for the research project at St. Paul's that our donations would continue to help fund. Participants enjoyed a lovely breakfast donated by Tim Hortons, and then were led in a 2 or 5 km walk by the lovely ladies of "Soul Sisters" running group. Returning to the picnic shelter at the Juan de Fuca field, the Silent Auction was enjoyed by many happy recipients of prizes and goodies. The newly designed shirts were a hit with all the participants and the sea of blue around the walk site attracted lots of questions and attention to spread the word about Scleroderma. **The icing on the cake for the day was the final donation tally: over $12,000 for Scleroderma Research.** Thank you to everyone who helped make this a very successful day. We hope to see you all back next year, and bring a friend!

- LINDA BARNES & JACKIE ALEXANDER

It seems like scleroderma has been a part of me my whole life. There are times when I can't remember what life was like before scleroderma.

I had to give up my lunch hour walks. I had just started a new job in 1991 at a major law firm in Toronto. It was a dream job for me. I commuted by train and had a busy workday, but the best part of the day was my lunch hour walks in the city. I would walk rain or shine, hot or cold. Then one day my fingers turned black after coming back from my walk and my hip joints started to ache. I bought warmer gloves and new running shoes, but that didn't help. These symptoms started a long series of tests and doctor visits. At first, I was scheduled for a carpal tunnel surgery, then finally referred to a doctor who said, 'No, it is Raynaud's.' I had never heard of Raynaud's Phenomenon, looked it up and thought, *'I can deal with this, I'll just wear warm mitts.'* ***Wrong!*** My hands puffed up like puff pastry making it hard to bend my fingers.

One cool Fall evening I was out walking my dog, wearing mitts, and caught frostbite on my index finger. It was the most severe pain I have ever experienced, and I was lucky I didn't lose the finger. Now I have no feeling in that finger. A few years later I applied for a Nexus pass. When I went for the interview and fingerprint scan it was discovered I had no fingerprints on my right hand. Try explaining that to border security!

But I made the decision to get on with life and live it the best I could. I was still able to continue to work, travel and enjoy life. I bundled up, even on warm days, as I am always cold, I always had gloves handy in my purse for when I go into air-conditioned establishments. I always drove with the windows open instead of using air conditioning.

I have to stop and think now about what I can and cannot eat. Having severe acid reflux has really changed my life. As a child I had so many upset stomachs and days of just not feeling well. I sometimes sit back and reflect, *'Did I have scleroderma even as a child?'*. I love spicy food, tomato-based food, and a good vodka tonic. However, all these trigger a severe reaction. And no, I haven't stopped eating those foods, I am my own worst enemy! But I did give up drinking as I would prefer to eat than drink!

I developed a cough in 2010. I went to my family doctor who performed a quick breathing test and I failed. She had no idea why as my lungs sounded clear. She referred me to a respirologist who walked into the waiting room, took one look at me and said, 'You have scleroderma, you have no lips.' I was astounded. First, that she would say that in a room full of people and secondly, what is scleroderma?

Then the fun began. I was referred to a scleroderma specialist in Hamilton. He was very blunt, told me I had lung scarring, should be on oxygen and had 5 years to live. I was in shock and denial. At that point I was breathing fine; it was just a cough! Like many of us, I went home and looked up scleroderma on the internet and thought, 'Yup this is it, I'm dying.' I asked to be referred to a doctor in Toronto. I was so thankful I did. I had two great doctors there who explained scleroderma to me. I was followed-up with them on a regular basis, had numerous tests and medications and things seemed to stabilize a little. However, my breathing did slowly get worse. Sadly in 2017 I decided I just couldn't do the commute any longer, and the fatigue was bad, so I retired and moved to Vancouver Island.

I was referred to the Scleroderma Clinic at St. Paul's Hospital. The last 2-3 years I have gone downhill quickly. I have severe pulmonary hypertension, interstitial lung disease, a non-working esophagus and the general aches, pains and fatigue of scleroderma. I came to the island with no doctor and now have 20! I am on home oxygen pretty much 24/7 now and take more pills than I care to mention. I've lost over 50 pounds.

In June I completed the testing, (wow, there are a LOT of tests), for a double lung transplant. I am a status 2 on the wait list. I'm told I have the best blood type for transplant so it could happen quickly. Fingers crossed that by the end of 2022, I will be breathing again. I know I will still have scleroderma after the transplant, and it will always be a part of me, but once I get my new lungs I am determined to not let scleroderma define me.

I still try my best to get out with friends, even though it is exhausting, because I need to get out and not let this disease own my life. I look forward to being able to walk my dogs, go shopping, travel and yes even vacuum! I have many goals for when I can breathe but the most exciting one is to be able to do the Scleroderma Walk in 2023.

See you all there!

SPINNING TOGETHER INTO THE FUTURE

Patients and Researchers Partnering to Make SPIN Research Even Better

The Scleroderma Patient-centered Intervention Network (SPIN) is a patient-oriented research organization that provides evidence and creates tools to better inform scleroderma patients and clinicians. SPIN comprises researchers, health care providers and people living with scleroderma worldwide. Currently, SPIN is working on a Patient Engagement Project that involves researching more effective ways to engage patients in scleroderma research.

Engaging patients in research is a fundamental aspect of the organization. SPIN aims to include patients in all steps of the research process. This includes deciding what topics or issues should be researched, what questions are asked to patients involved in SPIN, how patients are recruited for studies, understanding what the results mean for patients, and how findings are best communicated with patients and other stakeholders. SPIN actively practices patient-centred decision-making through its multiple committees and advisory teams. These patient advisory teams oversee and monitor each SPIN project, thus providing guidance on how the research process should operate. SPIN is a leader in including patients as decision-makers in research, but SPIN leaders still strive to improve their work in disseminating research findings to patients and including patients in the research process.

SPIN's focus with its new Patient Engagement Project is to work in partnership with people living with scleroderma to develop the most effective way of engaging patients in SPIN. This work will hopefully improve how SPIN works with patients and will guide other researchers and research organizations in making decisions regarding patient engagement.

SPIN aims to meet its patient engagement goals by first gathering existing information from other research groups on advantages and disadvantages of different strategies for engaging patients in research. Second, SPIN will develop tools (e.g., podcasts, infographics, lay summaries) designed, specifically, to communicate about research to patients in a way that is interesting, understandable, and provides the information desired by patients. SPIN will then test the tools with patients. Third, SPIN will use what it has learned to partner with patient advisors in formulating a SPIN engagement plan. SPIN expects this project to have many benefits, including that the research will provide evidence on how best to engage patients in health research, generally, and how best to communicate about complex research to patients. Finally, it will improve the quality of SPIN's research by more effectively integrating patient input in SPIN's research processes.

CARDIAC INVOLVEMENT IN SYSTEMIC SCLEROSIS

Dr. Sabrina Hoa, MD MSc FRCPC,
Rheumatologist, clinical researcher
at the CHUM Research Centre

Pulmonary arterial hypertension (PAH) means «high pressure in the arteries of the lungs». PAH is different from systemic arterial hypertension, which is usually referred to as «high blood pressure» and measured at the upper arm with a blood pressure monitor.

WHO IS AT RISK OF DEVELOPING CARDIAC INVOLVEMENT?

Cardiac involvement occurs in 10 to 30% of patients with systemic sclerosis. Cardiac involvement occurs in both the limited and diffuse forms of the disease, but is generally more frequent and severe in patients with diffuse disease with rapidly progressing skin involvement and in those with associated myositis (inflammation of the muscles).

WHAT ARE THE SYMPTOMS OF CARDIAC INVOLVEMENT?

Symptoms indicative of cardiac involvement include:

- the presence of respiratory symptoms attributable to pulmonary fibrosis;
- unusual shortness of breath or fatigue (tiredness) during physical exertion;
- shortness of breath at rest;
- shortness of breath when lying down;
- awakening during the night due to shortness of breath;
- painless swelling of the feet and legs (edema);
- chest pain that increases with physical exertion (angina);
- chest pain aggravated by inspiration (breathing in) and when lying down;
- palpitations or feeling that the heartbeat is irregular or abnormal;
- dizziness or fainting;
- generalized fatigue (tiredness).

It should be pointed out that a number of very different heart problems can cause identical symptoms. Therefore, the doctor will proceed to a more in-depth evaluation in order to determine the underlying cause of these symptoms and decide on the most appropriate course of treatment.

CARDIAC INVOLVEMENT IN SYSTEMIC SCLEROSIS

WHAT ARE THE CARDIAC MANIFESTATIONS OF SYSTEMIC SCLEROSIS?

Patients with systemic sclerosis can have cardiac problems that are directly caused by systemic sclerosis (small vessel abnormalities, inflammation, fibrosis), but can also have heart diseases commonly found in the general population around the age of 50, which is also the typical age at onset of systemic sclerosis (e.g., atherosclerotic coronary artery disease, valve problems, heart problems related to high blood pressure or "hypertension", etc.).

Cardiac involvement associated with systemic sclerosis can be classified according to the affected component of the heart (see Table for details):

Diseases of the pericardium (envelope of the heart):

- acute pericarditis: inflammation of the pericardium;
- pericardial effusion: accumulation of fluid around the heart; may be related to kidney damage or pulmonary arterial hypertension (high pressure in the arteries of the lungs;
- constrictive pericarditis: compression of the heart due to prolonged inflammation and excessive fibrosis of the pericardium (rare).

Diseases of the myocardium (heart muscle):

- myocardial fibrosis: hardening of the heart muscle caused by excessive collagen deposition;
- myocarditis (acute or chronic): inflammation of the heart muscle; may be associated with inflammatory muscle disease (myositis);
- heart failure: reduced ability of the heart to function; may be caused by myocardial fibrosis, acute or chronic myocarditis, high blood pressure (measured at the upper arm with a blood pressure monitor), pulmonary arterial hypertension, vascular disease or other causes.

Vascular diseases (vessels in the heart)

- microvascular ischemia: abnormalities of small blood vessels that constrict (narrow) spasmodically (as in Raynaud's phenomenon), leading to a decrease in oxygen supply to the tissues of the heart, which over time may lead to myocardial fibrosis.

Heart rhythm disorders (electrical system in the heart)

- arrhythmias and heart blocks: abnormal heart rhythm that is too fast or too slow, due to a disruption in the flow of electrical current through the different parts of the heart; can be caused by myocardial fibrosis, myocarditis, pulmonary arterial hypertension or other causes;
- dysfunction of the autonomic system: abnormal control of blood pressure and heart rate; may be an early sign of myocardial fibrosis.

HOW TO SCREEN FOR CARDIAC INVOLVEMENT IN SYSTEMIC SCLEROSIS?

Given the sometimes silent progression of the various cardiac abnormalities, it is important to perform a targeted cardiac assessment approximately once a year. This involves a questionnaire and physical examination, as well as certain screening tests such as an electro-cardiogram (or EKG, measures the passage of electrical current through the heart) and an echocardiogram. Further tests are sometimes necessary depending on the symptoms reported (stress tests, cardiac magnetic resonance, Holter, cardiac catheterization). Once detected, cardiac problems associated with systemic sclerosis usually require a consultation and concomitant follow-up with a cardiologist.

CARDIAC MANIFESTATIONS OF SYSTEMIC SCLEROSIS

TABLE

CARDIAC MANIFESTATION	DESCRIPTION	SYMPTOMS/SIGNS	INVESTIGATIONS	TREATMENTS
ACUTE PERICARDITIS	Inflammation of the envelope of the heart	Constant chest pain, aggravated by deep breathing and when lying on the back	• Electrocardiography • Echocardiography +/- Cardiac MRI	• Anti-inflammatory drugs • Colchicine • Prednisone
PERICARDIAL EFFUSION	Accumulation of fluid around the heart	Accumulation of fluid around the heart		• Observation (if asymptomatic) +/- Drainage
CONSTRICTIVE PERICARDITIS	Compression of the heart due to inflammation/fibrosis of the pericardium	Shortness of breath, fatigue, swelling of the stomach and legs		• Pericardiectomy (surgery to decompress the heart)
MYOCARDIAL FIBROSIS	Hardening of the heart muscle caused by excessive deposition of collagen	Often asymptomatic; symptoms of heart failure and/or heart rhythm disorders (see below)	• Echocardiography • Cardiac MRI	• Monitoring +/- treatments for heart failure and rhythm disorders
MYOCARDITIS	Inflammation of the heart muscle	Symptoms of heart failure and/or heart rhythm disorders (see below)	• CK, troponin (blood) • Echocardiography • Cardiac MRI • Catheterization, biopsy	• Prednisone • Immunosuppressants • Treatments for heart failure
HEART FAILURE	Reduced ability of the heart to contract and/or relax	Shortness of breath on exertion or at rest, aggravated when lying on the back; swelling of feet, fatigue, weight gain	• Echocardiography • Investigations for underlying causes	• Treatment according to underlying cause • Diuretics
MICROVASCULAR ISCHEMIA	Abnormalities in the small blood vessels of the heart	Chest pain aggravated on exertion or by cold weather (angina), shortness of breath	• Scintigraphy or echocardiography with exercise or stress test • Coronarography	• Aspirin, statin, nitrates, calcium channel blockers
ARRHYTHMIAS AND HEART BLOCKS	Abnormal heart rhythm that is too fast or too slow	Palpitations (abnormal heartbeat), shortness of breath, chest pain, dizziness, fainting	• Electrocardiography • «Holter» monitor for 24-48 hours	• +/- Anti-arrhythmic drugs • +/- Catheter ablation • +/- Pacemaker
DYSFUNCTION OF THE AUTONOMIC SYSTEM	Abnormalities in the control of blood pressure and heart rate	Symptoms of low blood pressure (dizziness) when making the transition from lying down to standing up, exercise intolerance, abnormal sweating	• Blood pressure and heart rate in lying and upright positions • Tilt-table test	• Staying hydrated • Salt intake • Compression stockings • Standing up slowly

NUTRITION AND SCLERODERMA

Audrey Potvin, TECH.DT.,T.N.
DIETARY TECHNICIAN AND TECHNOLOGIST IN NUTRITION

Knowing that scleroderma can manifest itself in several parts of the affected person's body (visible symptoms when the skin is affected and/or invisible symptoms when internal organs are affected), it is important to have the best nutritional intake to help in better coping with the symptoms of the disease.

Whether or not we are perfectly healthy, the food we eat is an integral part of a global approach to our health and can greatly influence our general well-being. It is in our best interest, especially when ill, to consume quality fuel that is adapted to the needs of our body.

Here are some of the principal symptoms of scleroderma and a few nutritional recommendations and useful advice to mitigate them.

ACID REFLUX AND SCLERODERMA

For several people affected by scleroderma, acid reflux (or gastroesophageal) is a particularly bothersome, sometimes painful symptom of the disease.

FIRST, WHAT IS GASTROESOPHAGEAL REFLUX?

It is acid reflux from the stomach to the esophagus, due to a malfunctioning lower esophagus sphincter (a valve that serves as a protective barrier) which, having lost its tone, can no longer close. This dysfunction of the esophagus sphincter can cause complications, such as inflammation, ulcers in the esophagus, dental cavities due to loss of enamel caused by acid regurgitations.

NUTRITION AND SCLERODERMA

GASTRIC REFLUX

Here are some tips that can help if you suffer from this symptom:

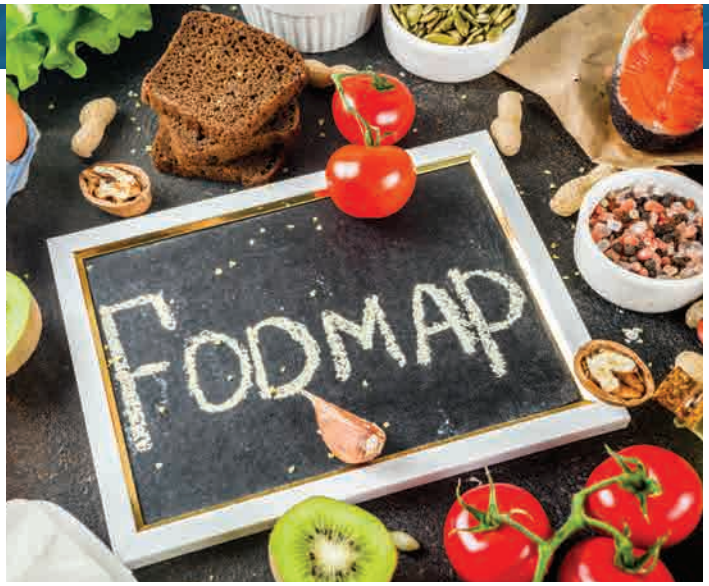
- maintain a healthy weight;
- considerably diminish, or even eliminate, certain foods such as: chocolate, coffee (caffeinated or decaf), alcohol, soft drinks, tomatoes, citrus, sugar, fried fatty foods, strong spices, white vinegar, mustard and mint;
- eat lots of vegetables, preferably raw or lightly steamed. At least half of our plate should consist of vegetables, one quarter of starchy foods, and one quarter of lean proteins;
- avoid eating two to three hours before laying down;
- have several small meals and snacks;
- eat slowly and chew properly;
- drink liquids at least a half-hour before or after the meal;
- avoid swallowing air (for example talking while eating, eating with your mouth open, drinking with a straw, drinking carbonated drinks, etc.);
- avoid chewing gum;
- if, on a special occasion, you are offered alcohol, don't drink on an empty stomach, as alcohol increases stomach acid and irritates the digestive system;
- avoid constipation by drinking a lot of water and by eating enough fibers (consult a nutritionist if needed).

Here is more advice that may help neutralize gastric acid:

- at the onset of the reflux, drink a tall glass of water to dilute the stomach acid;
- try raw potato juice if you have a juicer. Thoroughly wash the unpeeled potato before putting it in the juicer and mix with equal parts of water. Drink three times a day.
- in case of acid reflux between meals, eating half a ripe banana as a snack can bring some relief;
- drinking fennel or ginger tea stimulates digestion and helps neutralize acid. Aloe juice or gel is effective in alleviating the burning sensation of the digestive tract and stomach acidity. Avoid as much as possible baking soda and milk. Even though they are effective at temporarily reducing the burning sensation and digestive discomfort caused by gastric reflux, in the long run they will cause the stomach to produce even more acid and make the problem worse!

NUTRITION AND SCLERODERMA

GASTROINTESTINAL DISCOMFORT



If you have scleroderma, you may have intestinal transit problems, causing symptoms such as constipation and/or diarrhea, bloating, pain, abdominal distention, etc.

In 2005, a new approach to nutrition was developed by Sue Shepherd, an Australian nutritionist. She discovered that the range of foods that cause gastrointestinal disorders extends much further than consumption of wheat and dairy products. What is this new approach? It is called the **FODMAP** diet, which consists in reducing consumption of certain foods that contain carbohydrates. These, when fermenting in the colon, produce bloating, gas, and abdominal pain. They are called "fermentable".

BUT WHAT DOES THE ACRONYM FODMAP MEAN?

F	=	Fermentable (rapidly fermented by bacteria in the colon)
O	=	Oligosaccharides
D	=	Disaccharides
M	=	Monosaccharides
A	=	And
P	=	Polyols (sorbitol, mannitol, xylitol et maltitol)

NUTRITION AND SCLERODERMA

Here are some examples of foods rich in FODMAP to watch for and, eventually, to limit or eliminate in order to improve intestinal comfort:

Fructose:
Apple, watermelon, mango, corn syrup

Lactose:
Milk, yogurt, cottage cheese, ricotta

Frutane:
Asparagus, cabbage, onion, wheat and rye (in large quantity), apple

Galacto-oligosaccharides:
Legumes (chickpeas, lentils, soybeans)

Polyols:
Apple, pear, watermelon, cauliflower, mushrooms, peppers, sugar "alcohol" (sorbitol, xylitol, maltiltol), gum and unsweetened candy, etc.

Here are a few of my personal tricks to prevent or soothe discomfort following consumption of fermentable foods:

- add a piece of kombu seaweed when you cook legumes, to improve digestion. Some canned legumes already contain kombu;
- germinate legumes before cooking;
- add ginger to your meals;
- eat more gluten-free cereal (quinoa, buckwheat, etc.)
- replace milk with lactose-free milk or plant-based milks (almond, coconut, etc.)
- season with tamari sauce, miso, or baking powder. These condiments contain probiotics and/or enzymes that aid digestion (on top of adding flavor to dishes!);
- avoid eating dessert after a meal containing fermentable components;
- mint, fennel, ginger and cinnamon teas are excellent to promote digestion and eliminate gas after a heavy meal;
- puree vegetables to break the fibers that could cause irritation. Some people suffering from scleroderma sometimes have difficulty chewing raw vegetables, which can prevent adequate digestion. For example, cabbage-type vegetables, grated in a salad, will cause much less symptoms than if they are cut in large pieces;
- avoiding chewing gum (especially on an empty stomach, between meals), soft drinks or talking less while eating, can reduce bloating;
- eat slowly, chew your food well, and watch for repletion signals.

NUTRITION AND SCLERODERMA

ARTERIAL HYPERTENSION AND SCLERODERMA

Arterial hypertension (AHT) is a frequent health problem for a lot of people. It happens when arterial blood pressure is abnormally high.

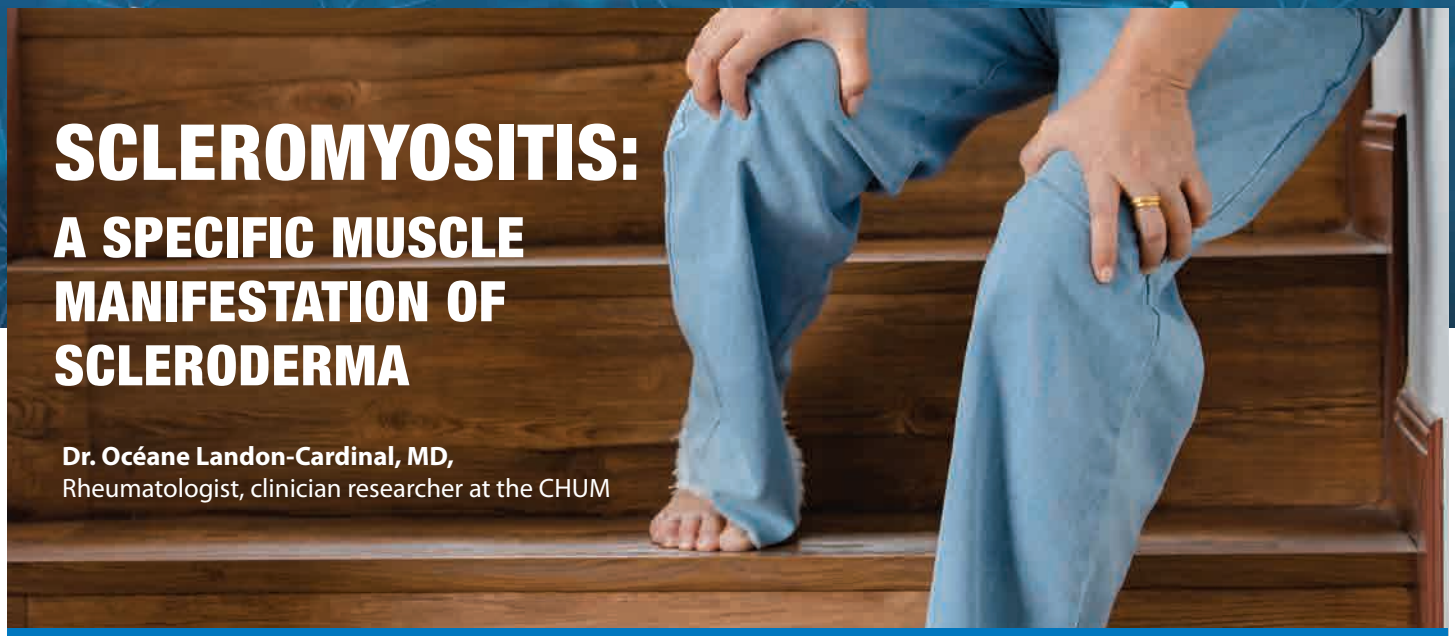
When arterial tension is caused by a disease, for example in case of renal involvement in a person suffering from scleroderma, or by frequent use of certain medications, it is called secondary arterial hypertension.

Even though we can't eliminate every risk factor, here are some recommendations that, mainly for primary AHT, but also for secondary AHT, can clearly contribute to a better control of arterial tension:

- reduce salted foods as much as you can (especially those that are highly processed such as chips, crackers, canned soups and sauces, etc.) as well as added salt. Sodium is one of the main causes of AHT;
- eat fresh fruits and vegetables that are rich in potassium, such as green vegetables (asparagus, spinach and peas), cruciferous vegetables (cabbage and broccoli), squash, sweet potato, apples, bananas, plums, grapes, cantaloupe, eggplant and melon;
- eat whole grain products;
- eat foods rich in omega-3, such as linseed, chia or hemp (vegetal sources) or oily fish such as salmon and trout (animal sources);
- avoid cold-cuts and cheese, as well as commercially processed dishes, often too high in sodium, or choose, in moderation, reduced-salt options;
- avoid caffeine, alcohol and tobacco;
- maintain a healthy weight;
- the following foods can, by promoting better blood circulation, help to lower arterial tension: Cayenne pepper, garlic, saffron, ginger, non-pasteurized apple-cider vinegar, olive oil and dark chocolate.

The above recommendations are for information only and do not replace medication prescribed by your doctor. Scleroderma is a serious condition but I firmly believe in a global health approach that aims at adopting healthy life habits, starting with the food we eat.

References: Nutrition and Recipe Book for people with Scleroderma and articles from Sclérodermie Québec The Bulletin.



SCLEROMYOSITIS:
A SPECIFIC MUSCLE MANIFESTATION OF SCLERODERMA

Dr. Océane Landon-Cardinal, MD,
Rheumatologist, clinician researcher at the CHUM

Scleroderma is a disease characterized by abnormalities in the functioning of small blood vessels and the immune system, ultimately leading to inflammation and excessive fibrosis (hardening) of the skin and various organs. When the inflammation reaches the muscles of scleroderma patients, it is called "scleromyositis". Scleroderma patients frequently report weakness, which may be due to a variety of causes (e.g., skin thickening, joint contractures, heart or lung involvement, and deconditioning). Weakness due to muscle inflammation (myositis) can therefore easily be missed.

WHAT ARE THE SYMPTOMS OF SCLEROMYOSITIS?

MUSCLE WEAKNESS:

The main symptom of myositis is usually muscle weakness, mainly in the shoulders and hips. People with myositis may have difficulty lifting their arms above their shoulders, lifting heavy objects, climbing stairs or getting up from a seat. Neck and back muscles may also be involved with difficulty lifting the head from a pillow or holding the head upright (dropped head). In some cases, the swallowing muscles are involved resulting in difficulty swallowing food.

RAYNAUD'S PHENOMENON:

Scleromyositis is often associated with a discoloration (successively white, blue and/or red) of the fingertips caused, in particular, by cold exposure. Raynaud's phenomenon is often the first clinical manifestation of scleroderma and may precede the onset of myositis by several years.

OTHER SCLERODERMA MANIFESTATIONS:

All the usual organ involvement of scleroderma can be found in scleromyositis. It should be noted, however, that the classic skin thickening of scleroderma is not always present at the time of onset of myositis, which may result in a delay in diagnosis.

SCLEROMYOSITIS: A SPECIFIC MUSCLE MANIFESTATION OF SCLERODERMA

CARDIOPULMONARY SYMPTOMS:

Myositis can weaken the muscles needed to breathe and cause shortness of breath. Some people may also develop inflammation and/or **fibrosis of the lungs**, which can contribute to shortness of breath and coughing. More rarely, myositis can cause inflammation of the **heart muscle** ("myocarditis") and eventually lead to heart rhythm problems ("arrhythmia") or heart weakness ("heart failure") which can cause shortness of breath or swelling of the legs.

JOINT PAIN/SWELLING:

Occasionally, inflammation of the small joints of the hands may precede or accompany muscle weakness.

HOW IS SCLEROMYOSITIS DIAGNOSED?

DETAILED QUESTIONNAIRE AND PHYSICAL EXAMINATION:

- Assessment of muscle strength
- Evaluation for cutaneous signs of scleroderma (e.g., skin thickening, vessel abnormalities visible at the nailfold)
- Cardiopulmonary examination
- Evaluation for joint pain and/or swelling

LABORATORY TESTS:

- ***Muscle enzymes:*** Measurement of creatine kinase (CK) or other muscle enzymes (AST, ALT, LD, aldolase) that may be increased following muscle injury. These markers are however not specific for myositis and may be increased in the blood for other reasons.
- ***Autoantibody assay:*** The presence of autoimmune markers in the blood may be useful in supporting a diagnosis of myositis, predicting associated organ involvement, and predicting a patient's response to certain treatments. However, it should be noted that autoantibodies in scleroderma are not found in all patients.

ELECTROMYOGRAM (EMG):

EMG measures electrical activity in the muscles using electrodes applied to the skin and may be abnormal in scleromyositis.

MAGNETIC RESONANCE IMAGING (MRI):

MRI is an imaging technique that uses magnetic fields rather than radiation to produce an image of the muscles. It can detect inflammation and muscle damage that may result from myositis.

MUSCLE BIOPSY:

The muscle biopsy consists of taking a small piece of muscle tissue (usually from the shoulder or thigh) under local anesthesia, which is then examined under a microscope.

Recent research has identified abnormalities of small blood vessels (capillaries) in muscle biopsies from patients with scleromyositis. The presence of multiple layers ("reduplication") in the wall ("basement membrane") of the majority of capillaries assessed in muscle biopsies is specifically found in scleromyositis. Identification of these vascular abnormalities, in addition to other autoimmune markers, on muscle biopsy is useful to support a diagnosis of scleromyositis, even when the patient does not have other scleroderma manifestations.

CAPILLAROSCOPY:

Capillaroscopy is a simple and painless examination performed on the hands to look for abnormalities in the small blood vessels called capillaries, located at the nailfold.

CARDIAC INVESTIGATIONS:

Additional tests such as an electrocardiogram (ECG), cardiac ultrasound or cardiac MRI will help to assess the presence and severity of **cardiac involvement**.

PULMONARY INVESTIGATIONS:

Additional tests such as a pulmonary function tests or a chest CT scan will help to assess the presence and severity of **pulmonary involvement**.

SCLEROMYOSITIS: A SPECIFIC MUSCLE MANIFESTATION OF SCLERODERMA

GASTROINTESTINAL INVESTIGATIONS:

Additional tests to evaluate the esophagus, stomach, small intestine and large intestine will help to assess the presence of various digestive system disorders associated with scleroderma.

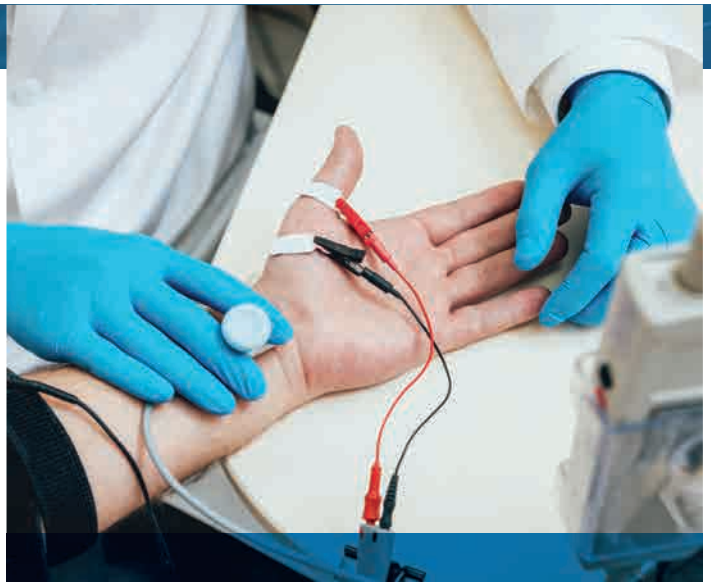
All or some of these different diagnostic tools can be implemented by your treating physician depending on your situation.

WHAT ARE THE TREATMENTS FOR SCLEROMYOSITIS?

Myositis can be treated with immunosuppressive drugs and muscle rehabilitation.

Immunosuppressive drugs will help regulate the immune system and block muscle inflammation. Current data suggest that these treatments are most effective in inflammatory muscle disease and less effective in fibrosing forms. Muscle biopsy is therefore very important for diagnosis but also to help guide treatment and anticipate the clinical response of patients. Immunosuppressive drugs are usually administered in combination with corticosteroids ("cortisone"), initially at a high dose (> 60 mg per day) and then gradually reduced.

High doses of corticosteroids are associated with an increased risk of developing a scleroderma renal crisis, a rare but urgent complication of scleroderma, which is caused by an acute decrease in blood flow to the kidneys and rapidly leading to their loss of function. Clinical factors such as the disease duration, the form of scleroderma (diffuse vs. limited) and the autoantibody profile will allow the physician to assess which patients are most at risk of developing this complication. Monitoring of certain symptoms and blood pressure at home will be recommended while taking corticosteroids. If the risk of a scleroderma renal crisis is considered too high, the doctor may suggest a blood product called intravenous immunoglobulin for a few months to allow the dose of corticosteroids to be lowered.



Muscle rehabilitation with physical therapy is an important aspect of scleromyositis patients treatment and is aimed at reducing inflammation and rebuilding muscle strength.

When muscle disease is very active, a light exercise program is usually recommended. Once inflammation is under control, muscle training should be intensified to prevent loss of strength and endurance. With the guidance of a physician and a physiotherapist, patients can be directed to an exercise program that is appropriate for their cardiorespiratory capacities.

IN SUMMARY

Scleromyositis is a muscular manifestation of systemic scleroderma and may be the earliest feature of the disease. Weakness may be multifactorial and careful evaluation must be performed to make the diagnosis of scleromyositis and provide optimal patient management. The identification of specific vascular abnormalities on muscle biopsy is useful to support an early diagnosis of scleromyositis especially when the patient does not have other scleroderma manifestations or scleroderma autoantibodies.

Visit the **sclerodermabc.ca**
to see the reference for each sheet highlighted
in blue in this document.

Community Contact Representatives

CONNECT WITH THE SCLERODERMA COMMUNITY IN YOUR AREA!

Give us a call, send us an email, and meet other people living with scleroderma.

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HOW SCLERODERMA CAN AFFECT THE HUMAN BODY

The symptoms of scleroderma vary greatly from person to person, so that patients will not necessarily develop all the complications of the disease.

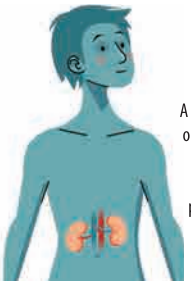
The symptoms of the disease may be visible, as is the case when the skin is affected, or the symptoms may be invisible, as when internal organs are affected.

SYMPTOMS AND MANIFESTATIONS OF SCLERODERMA

SKIN HARDENING
Thickening and loss of elasticity of the skin on different parts of the body. Hence the name «scleroderma», which means hard skin.

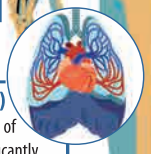
PULMONARY FIBROSIS
A potentially serious complication where normal lung tissue is gradually replaced by scarred fibrotic tissue, making it difficult to breathe and deliver needed oxygen to the body.

Pulmonary fibrosis causes shortness of breath and also sometimes a dry cough.



RENAL CRISIS
A renal crisis, which is due to an acute obstruction of arterioles and capillaries in the kidneys, leads to a sudden and sharp increase in arterial blood pressure. The symptoms are those of a hypertensive crisis: new and severe headaches, marked shortness of breath (left heart failure), and even epileptic seizures (convulsions). This is a very serious complication which requires urgent medical attention. Often during a scleroderma renal crisis, the kidneys stop functioning and dialysis (filtering the blood to avoid uremia) is then needed.

BLOOD VESSELS
The narrowing of the arteries, small blood vessels, and capillaries, can lead to many complications, including the development of pulmonary arterial hypertension (PAH), digital ulcers, and other conditions.



PULMONARY ARTERIAL HYPERTENSION (PAH)
Increased pressure in the pulmonary arteries due to the narrowing of small arteries in the lungs. Blood flow to the lungs is significantly restricted, making the heart work harder to pump blood through the lungs.

As arterial blood pressure rises in the pulmonary arteries, small pulmonary vessels slowly become clogged (a process which may take several years). This occurs through fibrosis of the small vessels, eventually leading to thrombosis, and the blood can no longer reach all parts of the lungs. Thus, it becomes more difficult for the lungs to supply enough oxygen to the body.

Sustained high blood pressure in the arteries of the lungs puts a strain on the heart, making it more difficult to circulate the blood through the lungs. Over time, this can eventually lead to congestive heart failure, particularly the right side, what is referred to as right heart failure (RHF). Right heart failure is indicative of significant PAH and is a serious complication of scleroderma.

PAH results in one or more of the following symptoms:

- Shortness of breath on exertion and at rest
- Palpitations (heart rhythm disorder)
- Fatigue
- Chest pain
- Dizziness
- Temporary loss of consciousness (syncope)
- Swelling of the ankles and legs

SCLERODERMA FACES
Hollow eyes, pinched nose, thin pursed lips, mask-like face, small puckered mouth (microstomia), and peri-oral folds.

Thinning lips and facial muscle atrophy can make the teeth appear more prominent.

EYES
Dry eyes caused by a decrease in tear production.

TELANGIECTASIA
Small dilated capillaries visible on the face and hands, sometimes referred to as «spider veins».

RAYNAUD'S PHENOMENON
Raynaud's is present in up to 95% of people with scleroderma. Whitening of fingers and/or toes triggered by cold or severe stress. The whiteness phase can be followed by a blue phase and then a red phase.

SCLERODACTYLY
The skin of the fingers, which have become infiltrated with collagen (fibrosis), may look full and sausage-like. Functional loss or decreased range of motion.

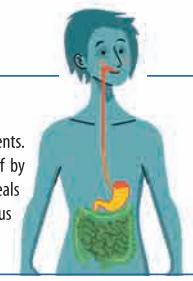
CALCINOSIS
Calcium deposits under the skin that may require antibiotics to cure occasional infections and sometimes surgery to drain calcium deposits and relieve pain.

DIGITAL ULCERS
Ulcers occur on the fingertips or on the top of the fingers. They are painful and difficult to heal.

In the most severe cases, it can lead to necrosis and amputation may be needed.

SKIN PIGMENTATION
Dark or pale spots occurring in one-third of patients.

DIGESTIVE SYSTEM
Gastrointestinal disorders affect the vast majority of patients. Gastric reflux is a common symptom that manifests itself by a burning sensation radiating up to the throat after meals and may cause inflammation of the lining of the esophagus (esophagitis reflux) if left untreated.



MUSCLE AND JOINT PAINS
Joint pain is common. It is caused by inflammation of the joints and tendons, which quite often leads to joint swelling and stiffness that can become quite debilitating.

Muscular pain (myalgia) can be intermittent or continuous. It can also be associated with muscle weakness (myositis). Symptoms include difficulty in climbing stairs, lifting objects and getting up, and also difficulty swallowing.

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Scleroderma Association B.C.