SCLERODERMA ASSOCIATION OF B.C.

The Bulletin

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SABC

WALK
for Scleroderma

SPIN-SHARE
Support Program

Summary

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A Word from Our President

The Scleroderma Association of B.C. (SABC) was founded in 1984 by five patients diagnosed with scleroderma. SABC is an all-volunteer, registered charity created to support patients and their families. Our membership currently includes about 450 patients and their supporters.

Our Mission

SABC has a three-fold mission: to promote patient outreach, support and education; to raise public awareness and understanding of scleroderma; and to encourage and support scleroderma research in BC and Canada.

In addition to our Board Members, we are very grateful to our members who also volunteer as Community Contact representatives around the province. To find your community representative, see page 15.

This year we are very excited to partner with Scleroderma Quebec and Scleroderma Manitoba to expand the working relationship we have developed over the past years. I am honoured to represent the SABC in this new partnership as it is a great opportunity to share and exchange information which will benefit research and the scleroderma community.

Patient outreach, support and education

SABC has a website with a variety of information for patients and their supporters at **www.sclerodermabc.ca**

Patient Advocacy

SABC advocates for the scleroderma community across the province and is committed to promoting disease awareness and improving the quality of life for all patients. Through advocacy, we aim to raise public awareness and rally support around the issues affecting the scleroderma community (eg. access to medications) to ensure our voice is heard and needs are met by government decision-makers. SABC not only serves as a support for scleroderma patients and their families, but also advocates with other associations, such as the Pulmonary Hypertension Association of Canada, for those who are also affected by other related diseases.

Scleroderma Clinic at St. Paul's Hospital in Vancouver

Scleroderma patients in British Columbia are privileged to have access, by medical referral, to a Scleroderma Clinic, the only one of its kind in western Canada. The Clinic evaluates and treats patients from all areas of the province. The clinic is engaged in several areas of research, including research into the mechanisms of scleroderma, the risks of progression, and evaluation of new potential therapies which the SABC is proud to support. For doctors only: referrals may be directed to Dr. James Dunne - Scleroderma Clinic Tel: (604) 732-4993.

Raise public awareness and understanding of scleroderma

The month of June is Scleroderma Awareness Month. SABC helps organize walks/rides across the Province to raise awareness and funds for research. This year we are supporting fundraising events in Valemount, Vancouver and Victoria (see page 8).

On October 5, 2019 we will be having our Annual General Meeting & Conference at the Poco Inn & Conference Center at 1545 Lougheed Highway in Port Coquitlam. This year we are planning to incorporate the option of real-time, online streaming, ensuring those unable to travel to the lower mainland have access to and can partake in, the conference and its valuable information presented.

Encourage and support scleroderma research in BC and Canada

SABC funds and co-leads a Genome Research Study in BC that began recruiting scleroderma patients in 2017. This research program is creating a firm foundation for intensive research to control lung and skin damage in patients with scleroderma. SABC has contributed over $240,000 in donations to date.

In addition to research being undertaken in BC, there are several research consortiums in Canada involved in studies focused on scleroderma that SABC supports:

- Canadian Scleroderma Research Group, under the directorship of Dr. Murray Baron. The CSRG maintains a key database of scleroderma patient information used by scleroderma researchers of many disciplines.
- Scleroderma Research Group of the Centre Hospitalier de l'Universite de Montreal (SRG-CHUM) by Dr. Jean-Luc Senécal and his team.
- Hamilton Scleroderma Group (HSG), St Joseph's Healthcare McMaster University
- Scleroderma Patient Intervention Network (SPIN)

ROSANNE QUEEN
President

WHEN SCLERODERMA ENTERED MY LIFE

I was 19 when scleroderma hijacked my future goals. I spent a few years being angry with my body, grieving the future I had dreamed for myself, and feeling isolated, even when I was surrounded by people who loved me but could never truly grasp the degree to which my life was changing. It happened so quickly. I went from developing a pre-professional dance career to having limited capacity for sustained movement within a few weeks.

With a history of joint issues, I was easily mis-diagnosed and treated for rheumatoid arthritis. The prescription cocktails I had tried, however, were not preventing the skin on my hands, arms, chest, and face from tightening. The chronic fatigue and pain were beyond any physical discomfort I had ever experienced before. After a few years of struggling to develop an effective treatment plan, I was accurately diagnosed with scleroderma and learned that the symptoms I had been experiencing in my extremities were typical of Raynaud's Syndrome. However, it would still be a few years before finding health care professionals willing to work with me and for me.

Changing Perspective

No longer able to pursue dancing as a career, my attention shifted towards wanting to help others like myself find ways of moving without pain. The human body has always fascinated me, so I chose to study kinesiology. While beginning my undergraduate degree, I had begun to establish a community of health professionals that influenced my drive for a deeper understanding of what enables health and well-being. I experimented with holistic practices and found adaptations for my life style that helped me heal while still trying to find medications that would fix more problems than they created. When I located a rheumatologist, who included a holistic view of health into her practice, I finally established a complete "tool kit" for managing scleroderma that included medications that were preventative.

My frustrations navigating the health care system, and the impact social inequalities have on accessing treatments, fueled my desire to break down barriers to well-being for others. At nearly the end of my Kinesiology degree I chose instead to complete a bachelor's degree in Health Sciences at Simon Fraser University with a minor in Political Sciences. I realized that, had I not found health professionals who had affordable services and who were willing to hear my perspective as a patient, the struggle to complete my post-secondary education would have been defeating. Regardless of your physical abilities, these milestone achievements should be accessible to everyone and advocacy seems like the best way to make that a reality.

Directing My Desire to Do Something About It

Since discovering the Scleroderma Association of B.C. my quality of life has improved. I have connected with a community of people that are resilient and have witnessed their power to accomplish amazing things together. Supporting Canadian research initiatives that aim to develop new diagnostic and treatment options, raising awareness within our communities, and sharing information with patients and their families that assist in improving well-being are goals we continue to achieve. Most importantly, there is a constantly growing network of support for those of us who are affected by this intimidating autoimmune disease.

It has taken a decade to come to terms with my physical limitations and the uncertainty of my future, but I've chosen to not let scleroderma control my life. I have recently been married, begun a new career, and started dancing again – all milestones I did not imagine would happen for me ten years ago. Scleroderma does not define who I am, but it does contribute to the perspective from which I see the world and function within it. Through my role within the Scleroderma Association of B.C., I will continue harnessing my experiences and knowledge to bring social awareness, justice, and support to our community.

February 2019 Update

SCLERODERMA ASSOCIATION OF B.C. RESEARCH PROJECT

SABC funds and co-leads a research study that began recruiting scleroderma patients with and without interstitial lung disease (ILD) in July 2017. Blood samples have also been taken from patients with only idiopathic pulmonary fibrosis (IPF) and both blood and skin samples have been taken from control participants. This research program is creating a firm foundation for intensive research to control lung and skin damage in patients with scleroderma and lung damage in patients with IPF, with the expectation of receiving future support from donations and, hopefully, research funding agencies.

Research Study Progress

Nov 2017	Collection of blood and skin tissue samples completed and stored in The Scleroderma Biobank at St. Paul's Hospital in Vancouver
Dec 2017	Skin tissue growth completed
Apr 2018	Extraction of micro-RNA (miRNA) from tissue types
May 2018	Frozen tissue samples from three tissue types transferred to the BC Genome Sciences Centre for sequencing
Jul 2018	miRNA sequence data downloaded for one-third of the samples and quality control (QC) checks started by the scientific team members
Nov 2018	Completion of QC assessment for first two-thirds of the samples
Feb 2019	Completion of QC assessment for last third of the samples and QC examination for pilot study of fourth tissue

This proof-of-concept study is to discover miRNA sequences and their differential expression amongst six different types of cells. This study is important for the approximately 15,224 Canadians with scleroderma and the approximately 7,045 Canadians with IPF (as confirmed by CT, biopsy, or bronchoscopy). Discovering which miRNA sequences are too low or too high and correcting these imbalances could lead to effective treatment of skin damage in patients with scleroderma and treatment of lung damage in patients with IPF only and in patients with both scleroderma and ILD.

SABC has been behind this project for the last five years, contributing over $240,000 in donations to date. Your donations do make a difference in contributing to research that otherwise would not even be considered for funding for these two orphan diseases.

The research team brings together experts in respirology, rheumatology, bioinformatics, and genetical statistics to uniquely tackle this challenge. The team is led by **SABC President Rosanne Queen**, **SABC past President Bob Buzza**, **Drs. James Dunne** and **Kevin Keen**. **Drs. Raewyn Broady**, **Robert Holt**, **Chris Ryerson** and **Pearce Wilcox** round out the scientific research team. Both Rosanne and Bob participate on the leadership team to keep us informed on the progress of this SABC-funded research program and to ensure the interests of patients and their families are at the forefront.

The Scleroderma Research Chair of the University of Montreal

Dr. Jean-Luc Senécal, M.D., and Dr. Martial Koenig, M.D.

THE CHAIR - WHAT EXACTLY IS IT?

First and foremost, it is a public recognition of the importance of scleroderma as a serious disease in society, and of the need to conduct medical research at the highest level to better understand the mechanisms of the disease and develop new treatments.

At the initiative of Scleroderma Quebec, the Chair was established in the early 2000s. The signatories to the Chair agreement are the University of Montreal and its Faculty of Medicine, the University of Montreal Health Centre (CHUM), the CHUM Research Centre (CRCHUM) and Scleroderma Quebec.

Financially, the Chair is a capitalized fund worth $1.5M at the University of Montreal, with interest paid annually to the Chairholder to finance research. Initially, the Chair was set up thanks to donations from the CHUM, the Faculty of Medicine of the University of Montreal, and thanks in particular to individual donations from patients and the public collected by Scleroderma Quebec.

What are the objectives of the Chair?

The main objective is to improve the quality of life and life expectancy of scleroderma patients.

To achieve this objective, the Chair uses the following means:

- expand and accelerate clinical research (with patients) and high-level fundamental research (in the laboratory),
- attract the best doctors and researchers,
- ensure the sustainability and continuity of research.

Who is the Chairholder?

The incumbent is Jean-Luc Senécal, MD, FRCPC, Full Professor in the Department of Medicine at the Faculty of Medicine of the University of Montreal, rheumatologist at the CHUM and researcher at the CRCHUM. The Chair's research activities are located in Montreal at the CRCHUM and the CHUM.

Are there any concrete examples of research discoveries and progress made as a result of the Chair?

Here are some examples, published in the best international medical journals.

First description of scleroderma in French Canada. This study, which involved more than 300 patients, described scleroderma in Quebec and made it possible to identify certain manifestations at the time of diagnosis that are strongly suggestive of a high mortality risk. The attending medical team can, therefore, intensify treatment and follow-up for those at risk, to improve their life expectancy[1].

Identification of predictive factors of scleroderma in patients with isolated Raynaud's disease. This major groundbreaking study uses scleroderma-typical circulating autoantibodies and nailfold capillary abnormalities detected by capillaroscopy to identify several years in advance people who are developing scleroderma. This study therefore makes it possible to diagnose the disease at an early stage – "prescleroderma" – and opens the door to new treatments which will stop scleroderma in its tracks from the onset[2].

Anti-centromere autoantibodies promote vascular complications of scleroderma. The four major pathophysiological mechanisms that cause scleroderma are autoimmunity (attack of the immune system on the person's body, including in the blood the presence of autoantibodies highly specific of the disease, such as antitopoisomerase I or anti-topo), microvascular anomalies, inflammation, and fibrosis. For some unknown reason, patients with anti-centromere autoantibodies are at particular risk of severe vascular complications, such as finger ulcers, and mortality from pulmonary arterial hypertension. The research team has discovered that the immune complexes formed by the anti-centromere autoantibodies and the centromeric B protein against which they are directed, promote a slow but progressive process of vascular obliteration, thus explaining how these autoantibodies contribute to the severe vascular manifestations of scleroderma[3].

Anti-topoisomerase I autoantibodies have pro-inflammation and pro-fibrosis properties. Patients with anti-topo autoantibodies have been found to be at particular risk of death from pulmonary fibrosis, but the etiological mechanism is still unknown. The research team discovered that the immune complexes formed by anti-topo autoantibodies with the topo itself have properties that promote inflammation and fibrosis, thus explaining how circulating anti-topo contribute to the manifestations of scleroderma[4].

Copy human scleroderma in mice. One of the major scientific challenges of scleroderma is the absence of an animal model that accurately reproduces its four major mechanisms. A good animal model allows experiments that ethics would not allow in humans (such as triggering scleroderma), gives access to a higher level of biological complexity than is possible in test tubes, and allows therapeutic testing of new curative and even preventive drugs, the success of which then provides a strong scientific justification for testing in humans.

In close collaboration with Marika Sarfati, MD, PhD, of the CRCHUM's Immunoregulation Laboratory, and thanks to a major research grant from the Canadian Institutes of Health Research, the team was able to copy human scleroderma in mice by developing a new model that reproduces its four cardinal anomalies. Using dendritic immune cells loaded with topo and injected into mice, this innovative model reproduces the production of anti-topo autoantibodies, as well as the inflammation and fibrosis of the skin and lungs typical of patients with anti-topo autoantibodies. Four weeks of scleroderma with this model is equivalent to approximately four years of human scleroderma, resulting in a major decrease in the time required with the model for future trials of new therapeutic molecules[5].

This research was also supported by Scleroderma Quebec, Scleroderma Society of Ontario, Scleroderma Society of Canada and Scleroderma Association of Saskatchewan.

Are there any other concrete examples of the Chair's usefulness in the fight against scleroderma?

Excellence of care. The Chair is helping to create a centre of excellence in the rheumatology department of the CHUM for the diagnosis and care of scleroderma. Patients receive the best care from their rheumatologists thanks to a highly qualified and dedicated multidisciplinary medical team at the forefront of knowledge.

Advanced teaching. All residents in the rheumatology subspecialization program learn to expertly diagnose and treat scleroderma and, at the end of their training, apply their knowledge to their rheumatological practice for the benefit of patients. No less than 35 new young rheumatologists now working in Quebec have been trained since the establishment of the Chair.

Training the next generation. The Chair awards scholarships to the best master's and doctoral students, thereby contributing to the advancement of knowledge and ensuring that the future of scleroderma care and research is in good hands.

Acquisition of exceptional research equipment. The achievements of the Scleroderma research Chair, the excellence of its research team and its international influence have led to an exceptional private donation of $400,000 from Scleroderma Quebec to Dr. Marika Sarfati and the Chairholder. This donation, combined with a major grant from the Canada Foundation for Innovation, enabled the CRCHUM to acquire a Becton-Dickinson FACSymphony device in the fall of 2018. It is a high-end, state-of-the-art multi-parameter cellular analyzer that can determine up to 50 characteristics of a single cell. Worth $1.5M, this device has a very limited distribution in Canada. In particular, it will enable the research team to better understand, both in the experimental model and in patients, the complex molecular and cellular mechanisms of scleroderma in order to identify potential new therapeutic targets.

Scientific references for this article are available on our website at: *http://sclerodermabc.ca/research-chair-in-scleroderma/*

The Vancouver Scleroderma Ride for Research

DETAILS ON THE SCHEDULE OF THE WALKS AND TO REGISTER

ONLINE AT:
www.sclerodermabc.ca

VANCOUVER

Sunday, June 16, 2019

(BIKE RIDE + WALK)

STANLEY PARK
Vancouver, BC V6G 1Z4

REGISTER BY PHONE
Rosanne Queen - 1.604.984.9425

BY EMAIL
rq.sabc@telus.net

No registration fee in Vancouver

VICTORIA

Sunday, June 23, 2019

WEST SHORE PARKS
1767 Island Hwy,
Victoria, BC V9B 1J1

WEST SHORE PARKS & RECREATION

REGISTER BY PHONE
Linda Barnes 1.250.380.9794

BY EMAIL
linda.barnes@shaw.ca

Registration fee Adult $35
(includes t-shirt),
Children under 10 free

VALEMOUNT

Saturday, June 29, 2019

CENTENNIAL PARK
5th avenue,
Valemount, BC

VILLAGE OF VALEMOUNT

REGISTER BY PHONE
Teressa Colosimo 1.250.566.4172

BY EMAIL
pattess72@hotmail.ca

No registration fee in Valemount

IF YOU'RE INTERESTED IN ORGANIZING A WALK OR FUNDRAISER IN YOUR COMMUNITY, CONTACT US!

* Scleroderma Association of B.C. accepts donations for these three fundraising activities.
This fundraising campaign is part of the walks organized in June for scleroderma.

JUNE IS SCLERODERMA AWARENESS MONTH!

BECAUSE OUR FAMILY & FRIENDS CARE, IN 2018 WE NOT ONLY RAISED AWARENESS BUT $52,556.53 FOR SCLERODERMA RESEARCH!

Please join us this June for even more fun!

Vancouver

The 8th annual **'Ride for Research'** (bike ride/walk in Stanley Park and beyond), again organized by Rosanne and David Queen, will be held on Sunday, June 16, 2019.
Contact Rosanne 604-984-9425 or rq.sabc@telus.net

" Many ask How and Why we do it. Simple answers, really. The "How" is I ask every family member, friend & acquaintance to help us find a cure for this horrible disease, (and I ask more than once!). The "Why" is I've lived with this disease for more than 20 years and I feel I'm one of the lucky ones. Bike Rides & Walks enable patients and those that care about them to raise awareness. Because most scleroderma research in Canada is patient-driven, monies must be raised to continue the research and the hope for a cure. Research studies in BC, Canada and around the world give us that hope. Last year, the 7th Annual Scleroderma Ride for Research raised $41,600. In the last 7 years we have raised more than $220,000. It's amazing what a fun-filled morning with family and friends can do! " ***- Rosanne***

Victoria

The 2nd annual **Walk for Scleroderma**, on June 23rd. Please join Linda Barnes and all Vancouver Island scleroderma supporters, for this event.
Organizer: Linda Barnes 250.380.9794 or linda.barnes@shaw.ca

" As CEO of West Shore Parks and Recreation, and after a long career in Recreation, I understand the healthy benefits of regular activity. Six years ago, when my diagnosis of Scleroderma started to change my ability to lead an active lifestyle, I was devastated, in pain and depressed. In the spring of 2017, I could no longer work full time, but I decided that I wasn't letting Scleroderma steal my joy in living the best life I could. The doctors and staff at St Paul's Scleroderma Clinic have helped me to continue to be active and to manage my symptoms in the best way possible. After attending the Ride for Research in Vancouver in 2017, I decided to organize the 1st Walk for Scleroderma in Victoria in order to raise money for continued research at St Paul's, and to gather a community of Scleroderma patients and supporters on Vancouver Island. As "Oma" to 13 wonderful grandkids, I intend to stick around and fight for a cure as long as possible! Please join us in raising awareness and funds to fight back. " ***- Linda***

Valemount

The 3rd annual **Walk for Scleroderma** in Valemount, again organized by Teressa Colosimo, will take place on Saturday, June 29. **Organizer: Teressa Colosimo 250.566.4172 or pattess72@hotmail.ca**

" I was diagnosed 7 years ago after years of trying to figure out what was happening to my body. During those uncertain years, I was starting to think these symptoms were all in my head. While a diagnosis of Scleroderma was a name to call it, it then became 'What the heck is Scleroderma?'. After several years of family, friends and co-workers asking me that same question, I was inspired by a friend to start a walk here in Valemount. The goal of my walk is to raise awareness about Scleroderma by spreading the word and raising funds for research. We need to find a cure for all of us fighting this rare disease. With continued support from my family, friends and the community of Valemount, we will be hosting our 3rd Annual Walk this June. Our 2nd Annual Walk raised $2153.38 " ***- Teressa***

Immunoregulation lab at CRCHUM

HEENA MEHTA, PHD AND MARIKA SARFATI, MD PHD

The "initial trigger" for scleroderma is still unknown, but it is now thought to be an "insult" to the immune system leading to dysregulation of immune pathways, and the cascade of events that lead to clinical manifestations. Key interests of the research team at CRCHUM (Research Center of the University of Montreal Health Center) include study of immune cell populations such as T cells, and dendritic cells, which are the sentinels of the body's defence system, as well as their interactions at barrier sites such as skin, lungs, and gut. In recent years, the rapid advancement of technology available to immunologists to study cells in affected tissues at a single-cell level, and concomitant development of tools for analysis of high-dimensional data make it an exciting time to be studying the immunopathology of scleroderma. Here, we highlight some milestones we have reached with generous support from various scleroderma associations in Canada including Sclérodermie Québec, Scleroderma Society of Ontario, and Scleroderma Society of Canada.

Milestones

January 2011: Pilot grant from Sclérodermie Québec.

November 2011: Experiments initiated to develop a small animal model (mouse) that mimics diffuse cutaneous scleroderma using autologous (self) dendritic cells and peptides from topoisomerase protein, a known scleroderma autoantigen **(TOPO I).**

2012-2016: Student fellowships from the Scleroderma Research Chair (Dr. Jean-Luc Senécal).

August 2015: Received a federal research grant from the Canadian Institutes of Health Research.

2016-2017: Publication of peer-reviewed articles; report of animal model displaying diffuse skin, and lung fibrosis as well as anti-TopoI autoantibodies[1], proof-of-concept study with dabigatran (a direct thrombin inhibitor) demonstrating the need for caution when using this drug in scleroderma patients[2], and establishing a link between early-life gut dysbiosis, and skin and lung fibrosis[3,4].

October 2018: Acquisition of BD FACSymphony at the CRCHUM with support from Sclérodermie Québec and Canadian Foundation for Innovation.

Future directions

The continued commitment to research for this orphan disease with unmet medical needs brings us closer to fulfilling the promise of personalized medicine for patients. Since inflammation precedes fibrosis, the immune system is central to the disease process. With the insights we gained from our animal model studies, we are now very excited to explore immune pathways in affected tissues of patients with different forms of scleroderma. Recent acquisition of cutting-edge instrumentation, the BD FACSymphony, will enable us to comprehensively study the immune profile in blood and at barrier tissues. This single-cell level analysis that was not possible before, would require only limited amount of blood and small skin biopsies. Establishing a link between different clinical presentations of scleroderma and the immune landscape at a given timepoint of the disease will open the doors for identifying new immune therapeutic targets and novel clinical management strategies. Partnership between patients, basic science researchers, clinicians, and generous support from organizations and individuals will be crucial to improve scleroderma patients, and ultimately find a cure for this devastating disease.

Scientific references for this article are available on our website at: *http://sclerodermabc.ca/immunoregulation-lab-at-crchum/*

Interstitial Lung Disease in Systemic Sclerosis:

Should Mild Disease Be Treated?

DR. SABRINA HOA, MD MSC

From left to right:
Dr. Sasha Bernatsky, Mr. Gaétan Baril, President of Scleroderma Quebec,
Dr. Sabrina Hoa and Dr. Marie Hudson

Systemic sclerosis (SSc) is a rare autoimmune disease characterized by various degrees of skin fibrosis and internal organ involvement. Interstitial lung disease (ILD), or pulmonary fibrosis, is a common complication of SSc, affecting up to 50% of patients. Immunosuppressive drugs are currently used to treat SSc-ILD, particularly in patients with pulmonary symptoms (e.g. shortness of breath or cough), severe lung function impairment, or progressive worsening disease. However, the usefulness of these drugs in patients with normal or mildly impaired lung function is currently unclear, given that this patient population was mostly excluded from randomized controlled trials. Yet, SSc-ILD with normal or mildly impaired lung function is frequent, representing approximately half of all SSc-ILD patients.

In the course of her Masters of Epidemiology thesis project at McGill University, Dr. Hoa was interested in exploring the role of immunosuppressive drugs, namely cyclophosphamide and mycophenolate mofetil, in the treatment of patients with mild SSc-ILD. Using data collected in the Canadian Scleroderma Research Group (CSRG) registry from 2004 to 2017, she studied the characteristics and outcomes of 116 patients with mild SSc-ILD. About 10% of these patients were exposed to cyclophosphamide or mycophenolate mofetil mostly for joint and skin disease. After one year, the forced vital capacity (a measure of lung function) of patients who were exposed to these drugs was compared with that of patients who were not exposed to these drugs.

What were the results? Patients with mild SSc-ILD who were exposed to immunosuppressive drugs for reasons other than lung disease had better lung function test results at one year compared to patients who were not exposed, even after adjusting for baseline lung function values, disease duration, extent of skin disease and shortness of breath scores. Moreover, none of the patients who were exposed to immunosuppressive drugs progressed over the next two years, whereas up to 25% of patients who were not exposed to these drugs progressed over this timeframe.

These preliminary results are very interesting in that they suggest that immunosuppressive drugs may have a disease-modifying role in the treatment of early SSc-ILD. Future randomized controlled studies looking at efficacy of therapies in SSc-ILD should include patients with mild SSc-ILD, as these patients may also benefit from disease-altering therapies. Further research also needs to be done to better characterize the predictors of disease progression among patients with mild SSc-ILD.

Dr. Hoa has now completed her Masters of Epidemiology and is very grateful to her supervisors, Dr. Marie Hudson and Dr. Sasha Bernatsky, for their exceptional mentorship. She is also thankful to Dr. Murray Baron, director of the CSRG, and all the patients and researchers who have contributed to the richness of the CSRG registry. She is deeply grateful to Sclérodermie Québec, the Scleroderma Research Chair of the University of Montreal, The Arthritis Society, the Canadian Institutes for Health Research and the Centre Hospitalier de l'Université de Montréal (CHUM) Foundation for their financial support during her postdoctoral fellowship training. Dr. Hoa has now been appointed Assistant Professor in the Department of Medicine, Faculty of Medicine, at University of Montreal and in the Division of Rheumatology at the CHUM. As a clinician-researcher, she hopes to further knowledge on the optimal use of therapies to improve outcomes of all people affected by scleroderma.

The *SPIN-SHARE* at the National Scleroderma Conference:

MOVING FORWARD WITH PATIENT-CENTERED SUPPORT PROGRAMS

The Scleroderma Patient-centered Intervention Network (SPIN) was created to bring together people living with scleroderma, healthcare professionals who care for them, and scleroderma researchers from around the world to develop and test accessible support tools for scleroderma patients.

At last September's National Scleroderma Conference in Calgary, Alberta, SPIN was proud to announce the launch of our online platform for sharing our scleroderma support programs: SPIN-SHARE. Once ready, these programs will be available to the public free-of-charge through patient organization websites, including Sclérodermie Québec Scleroderma Association of B.C., and Scleroderma Manitoba.

You can keep up to date with SPIN's projects on our website (**www.spinsclero.com**),

Facebook (**www.facebook.com/spinsclero**) or Twitter (**www.twitter.com/spinsclero**).

Dr. Brett Thombs, the Founder and Director of SPIN, gave a preview of SPIN's first support program, which addresses hand function limitations in scleroderma, and will be released to the public in Spring 2019. The hand program was jointly developed by patients and rehabilitation specialists, and includes instructional videos for hand exercises, illustrations showing common mistakes, advice on how to develop a personalized exercise routine, goal-setting and tracking features, and patient stories of their experiences with hand disability and exercises.

Dr. Thombs's presentation described other SPIN patient programs that are currently in development, including a disease self-management program, a program to support positive coping with emotions, and a program to help patients deal with body image distress. SPIN also recently pilot tested a videoconference-based program to provide training and support for scleroderma peer support group leaders.

SPIN's first online toolkit to support hand function in scleroderma includes instructional videos designed by medical experts in scleroderma.

Health-related quality of life (HRQoL)

Dr. Marie Hudson, MD MPH
CSRG – GRCS

Health-related quality of life (HRQoL) is defined as an individual's perception of his or her well-being, and takes into account both physical and psychological states. It is, therefore, a highly subjective concept, as only the individual can determine his or her own HRQoL. Furthermore, it is a multidimensional concept, including physical, psychological, functional and social health domains, as well as satisfaction with health care.

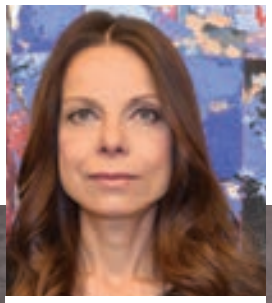
The study of HRQoL is useful for several reasons:

1. to provide information about aspects of the illness other than the usual biometrics, such as morbidity and mortality;
2. to inform public health authorities of the impact of various health conditions, and allow comparisons with other disease in order to distribute resources equitably;
3. to aid in the identification of new targets of intervention that are of importance to the patients; and
4. to evaluate new treatments to establish whether the benefits outweigh risks and side effects.

Until now, very little research has been done to document HRQoL in systemic scleroderma (SSc). Dr. Hudson and her colleagues in the Canadian Scleroderma Research Group (CSRG) have undertaken several research projects in this area. To start, a systematic review of studies on HRQoL in SSc was undertaken and identified 12 articles including more than 1000 SSc patients. A meta-analysis of these studies showed that HRQoL in SSc is one and a half standard deviation lower than the general population. In other words, on average, 85% of the general population has a better HRQoL than those with SSc, or that is to say people with SSc are among the lowest 15% of the population in terms of HRQoL. Also, there is very little difference in the HRQoL among people with diffuse and limited scleroderma, and those differences are almost imperceptible.

The CSRG researchers have since focused on the HRQoL of the 1000 SSc patients in their cohort. This enabled the researchers to make important observations. First, although the group of patients had mild to moderate disease, their HRQoL was as poor as those previously studied in the medical literature. In other words, their HRQoL was comparable to the 15% in the lowest end of the spectrum. Second, although the CSRG subjects reported that skin, respiratory, digestive and joint involvement had strong negative impacts on HRQoL, fatigue, pain and depression also figured as other major sources. Finally, the impact of SSc on HRQoL was at least as severe, if not more so, than other, better known chronic illnesses, including heart and lung disease, diabetes, hypertension and depression.

To conclude, this research emphasizes the negative impact of SSc on HRQoL and highlights issues which have, to date, been under-studied in SSc, such as fatigue, pain and depression. With this knowledge, the CSRG has several new research projects underway to address these problems, including fatigue and depression.



AUDREY POTVIN,
TECH.DT

Follow the New Food Guide!

Twelve years after the last 2007 edition, the new Canadian Food Guide (CFG) is finally here! The current version, inspired by Harvard's Healthy Eating Plates and by the Brazilian Food Guide, has replaced the rainbow with its four food groups and its portions concept. Canada's 2019 Food Guide now recommends that we fill half our plate with fruits and vegetables, one quarter with protein sources and the other quarter with whole grain foods. The "Milk and substitutes", and the "Meat and substitutes" groups have been excluded to give a larger place to plant-based proteins sources.

Furthermore, Health Canada is putting forward new recommendations such as:

"Be mindful of your eating habits";

"Cook more often";

"Enjoy your food";

"Eat meals with others".

So, in 2019, vegetal sources of food are preferred over animal sources of proteins. And as for beverage, it's simple: water! Fruit juices, which used to be part of the "vegetables and fruits" group, are now considered to be a sugary drink to be avoided.

Regarding the recommendations, the new edition of the CFG focuses on people's eating habits, by suggesting they enjoy healthy meals with family, friends, and neighbours; do more home-cooking with fresh ingredients rather than buying industrially transformed, pre-packaged meals; take the time to eat well, enjoying the taste of the food; read labels and limit the quantity of foods high in sodium, in sugar and in saturated fats, and be vigilant towards the food industry marketing strategies. The Guide promotes better cardiovascular health because it has less saturated fats, prevents obesity because it is more satisfying, preserves the health of the microbiota due to its high fiber content, and helps prevent certain types of cancer.

What else? Health Canada has also produced a 60-page document that provides guidelines to support improvements to the Canadian food environment by making better food choices.

And last, I leave you with one of my favourite home-made wafer recipes, made with healthy ingredients, for rushed mornings or healthy snacks!

References:
L'Épicerie (TV show) of February 6 2019
Text from Bernard Lavallée (Le Nutritionniste Urbain) titled: «Guide alimentaire 2019 : Santé Canada en route vers le futur»
Canada Food Guide (2007 and 2019 editions)
Recipe : https://petitevanille.weebly.com/recettes

OATMEAL, BANANA, DATES, WITH ALMOND BUTTER, COOKIE

12 Cookies

INGREDIENTS

1 tbsp **chia seeds**

2 tbsp water

2 very **ripe bananas**

1 tsp **vanilla extract**

½ cup **almond butter** (or other nut butter)

1 cup **oatmeal**

½ cup chopped **dates**

½ tsp baking soda

A pinch of salt

PREPARATION

1. Preheat oven to 375°F.
2. Mix the chia seeds and water in a small bowl and allow it to swell for a few minutes. Set aside.
3. In a blender, mix (until smooth) the expanded chia seeds, bananas, vanilla, almond butter, oatmeal, baking soda and salt.
4. Add the dates.
5. Set 12 dough balls on a greased cookie sheet and cook for 8-10 minutes.

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.

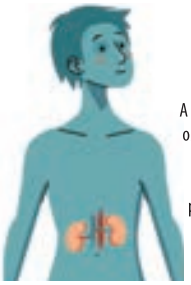
The symptoms of the disease may be visible, as is the case when the skin is affected, or the symptoms may be invisible, as when internal organs are affected.

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 FACIES
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.

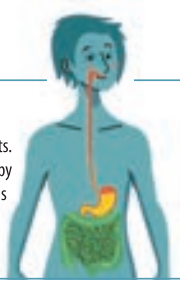
CALCINOSIS
Calcium deposits under the skin that may require antibiotics to cure occasional infections and sometimes surgery to drain calcium deposits and relieve pain.

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.

SCLERODERMA QUEBEC
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