

SCLERODERMA ASSOCIATION OF B.C.

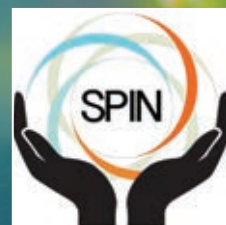
# The Bulletin

Spring-Summer 2020 | Volume 2 Number 1

**Research  
Across Canada**

**Protecting You  
and Your Estate**

**June Virtual  
Walks**



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## Board of Directors



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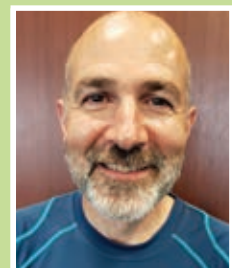
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## SABC ANNUAL GENERAL MEETING AND CONFERENCE

**SATURDAY  
OCTOBER 3<sup>RD</sup>, 2020**

This Fall, SABC invites and encourages you to join us **'virtually'** for the 36<sup>th</sup> annual opportunity to learn and share information and ideas at the 2020 AGM and Conference, held on **Saturday October 3.**

Please periodically check the website [sclerodermabc.ca](http://sclerodermabc.ca) for conference program details and updates regarding online registration timelines.



**Our mission statement hasn't changed since 1984 but you may have noticed that our logo has. We think it clearly portrays the beauty of BC:**

**The mountains, ocean and rivers!**



### **Living with Scleroderma**

Approximately 2,300 people live with this disease in BC, that's about 1 in 2,000. Sharing stories about others who have scleroderma is important to let you know that you are not alone. Others may be experiencing what you experience and the stories portray how strong individuals are facing the challenges of this disease.

I want to thank Beth (story on page 4) for sharing her story. I would appreciate receiving more personal stories to share with our community.

### **June Awareness**

Spreading awareness for this orphan disease is not only important to educate the public but also to highlight our sense of community. Awareness campaigns give our family and friends an annual opportunity to show us they care. The campaigns also show we are strong and that scleroderma does not define who we are!

New this June, due to the COVID-19 pandemic, we are introducing a virtual bike ride and virtual walks. We celebrate the communities holding these virtual events all around the province.

So now, everyone can join to help raise awareness generated by these events in June, knowing that 100% of the money raised will go to scleroderma-related research in BC and across Canada.

Check out page 9 (and the SABC website) for information on how to participate. Send the links to your family and friends. You will be surprised how many people will support you because as we've witnessed time and time again,

they do care. Don't forget to send us your 'selfies' so we can all share in each other's success! In this time of stress due to the pandemic, we need to stay connected and laugh!

### **Research**

There are many great research projects happening in BC and across Canada. None of the research happens without continued financial support through patient-driven fundraising. We can accomplish so much with the trust and support of our members, volunteers and very generous donors.

Canadian research projects cover a wide array of scleroderma-related topics to try and better understand and determine innovative ways to attack this disease. A visual display of current researchers is provided on pages 6 to 8 showing who, where and what, we, as a national scleroderma community, are studying to improve the future for all patients.

### **Something Extra**

In this issue of the Bulletin, we are talking about the often unspoken topic of wills and representation agreements. With our busy schedules, it's hard to find the time to make sure our financial and medical affairs are in order, but I urge you to do it sooner than later. There is a section on how you can maximize your charitable dollars when leaving legacies and save on taxes (page 5). Also find out how to more easily get your financial and medical affairs in order with the Tips on pages 12 to 14.

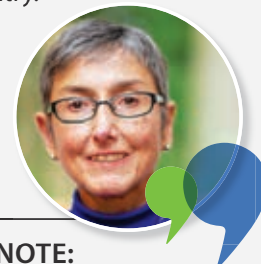
**October 3, 2020 -**

### **Save the Date! SABC's AGM and Annual Conference - Virtual**

Once again, our annual conference will be held in October to allow us to focus on our awareness efforts in June. In the months to come, updated information will be posted on our website.

### **Synergy of Three**

Since the fall of 2018, SABC has been working closely with Sclerodermie Quebec and Scleroderma Manitoba. This alliance has allowed us to strengthen our representation in the scleroderma community. Together our websites have improved along with our ability to share common information and resources. The other communication tool that we are extremely happy about is The Bulletin; the semi-annual magazine with its collaboratively developed content and organization-specific articles. In addition to The Bulletin being sent to our members, it is sent to hospitals, clinics, doctors and specialists across the province to increase awareness and understanding. The Synergy of Three is a solid collaboration that reaches out across the country.



### **PERSONAL NOTE:**

**I would like to thank all the people that have supported SABC over the last 36 years. I would also like to thank the board members for their time and passion. We can't do all that we do, without you.**



## Beth's story

**My journey with Scleroderma began in the early 2000's with random, seemingly unconnected symptoms. I was a teacher and very active – skiing, hiking, dancing, which fit well with my lifestyle in Squamish. The initial symptoms included wrist and shoulder pain and stiffness, Raynaud's Syndrome, and some minor digestion issues. I was referred to the right doctors, any changes were dealt with as necessary, especially as my lungs gradually worsened.**

In 2015 I was still teaching but had reduced my workload by half. My lung capacity was about 50% and I had given up on skiing, hiking, even yoga (bending over? Nope!) I still travelled but moved much slower and needed oxygen to fly comfortably. I did resume exercising, thanks to St. Paul's Healthy Heart program, which provided the oxygen I needed. But with extensive pulmonary fibrosis and worsening pulmonary arterial hypertension it was time to seriously consider a transplant.

Pre-transplant assessment was a long, drawn-out process. I was seen by several specialists; I met with a psychologist, a social worker and a dietician, all to assess my readiness for the challenge ahead. I updated my immunizations and waited for the final word.

By 2016, after a restful, healthy summer, I returned to the classroom. The previous year, I had begun to experience a weakening of my ability to fight off viruses. That fall the cycle began again. One cold followed another, and finally, around November, I was diagnosed with pneumonia. Breathing was so hard that I now needed oxygen full-time. My doctor was away, and the other doctors didn't seem to take me seriously. The hospital emergency doctor patted my head and sent me home. For the first time I felt helpless, so I reached out, one of the hardest things I've ever done. I had spent years being independent and coping well, but this was something I couldn't rationalize away. It was a very humbling experience.

My first call was to my parents in Ontario, who came to spend Christmas caring for me. Luckily, I was connected to my specialists, who monitored me, set up tests, and then admitted me to St. Paul's for the antibiotic treatments that beat back the pneumonia. My doctor pointed me to an RT who contacted the Home Oxygen program and, the next day, I had the machinery

I needed. Finally, I was put on the lung transplant list. It was a difficult recovery, but I got over the pneumonia.

The main hurdle was admitting my dependence on oxygen. Looking back, I realize I had convinced myself that being out of breath was normal. Now I use oxygen all the time, at high levels for exercise, at lower levels for running errands, and doing chores. Supplemental oxygen has greatly improved my energy and my quality of life.

Though I loved teaching, I decided not to return to work. It was clear that constant exposure to bugs was affecting my health. I was approved for transplant, so I needed to stay as healthy as possible. I am grateful that my union has excellent benefits. Not everyone has the luxury to make such a decision without serious financial difficulties.

I have been on the transplant list, in limbo, for three years and am still waiting. My health has been stable, probably because I'm not working. I don't have to worry about episodes of fatigue or a gut freak-out. I manage my appointments and health-related obligations without stress and continue my exercise routine. I took up tai chi and bridge to keep my mind busy and developed a new social network in addition to my close friends. Turns out my schedule and speed are a perfect match for many retired folks!

I hope sharing my story helps others, whether you find comfort in someone else's experience, confidence to speak up for yourself with doctors and those who may not fully understand your daily challenges, or courage to consider a riskier treatment path. I regret nothing in my journey, I am grateful every day that I have so many people to support me and bring joy to my life. Scleroderma doesn't define me!

# Maximizing Your Charitable Dollars Meaningful Impacts and Saving Taxes

**AUTHORED BY: ROSEMARY WESTIE AND TROY IWANIK,  
INVESTMENT ADVISORS, HOLLISWEALTH®,  
A DIVISION OF INDUSTRIAL ALLIANCE SECURITIES INC.**

Every once in a while, we have an opportunity to do something really good that makes a big impact. Helping people with Scleroderma is near and dear to our hearts, as we know some fantastic people living with this often-painful condition. We will explore the benefits and tax savings of donating securities (mutual funds, stocks, or ETFs) to your preferred charitable cause.

Perhaps you would like to create a legacy and make a positive social impact at the same time. If you have investments in non-registered accounts (this would exclude RRSPs, RRIAs, or TFSAs) that have built up capital gains over the years, then please read on. The Canada Revenue Agency will request a larger share if you sell the investment, and then donate the proceeds to your selected charity, than if you simply donate it as is (in kind). From the chart below, we see an example of how this would look in practice. This strategy is mutually beneficial to you, and your chosen charity—you receive no tax liability, and your charity receives more money.

Reviewing the example in the table below, you may notice that by donating the securities directly to your charity will result in \$9960 more going to help the people that will benefit most. This is a real tangible benefit to the organization that is most important to you.

In Canada, if you donate to a charity you receive charitable tax credits. They will vary, depending on your province and marginal tax rate. In BC for 2019, if you donated \$50,000 of securities in kind, you could have received a charitable tax credit for \$22,848.52, versus \$18,634.92 by selling the securities first, then donating the cash. You could therefore increase your charitable tax credit by \$4213.60 donating the securities in kind and not selling them first.<sup>2</sup>

Furthermore, you do not have to claim the donation tax credit in the year you make the donation. You may carry forward the unclaimed donations for five tax years.

There are similar strategies that employ donating stock options to reduce concentration risk and lower taxes. You can also donate registered assets (RRSPs, RRIAs, and TFSAs) in kind upon one's passing to your preferred charity.

There are even more advanced strategies available as well related to the donation of securities. Contact your investment advisor for further details on any of the examples mentioned.

*This article may contain multiple strategies, not all of which will apply to your particular financial circumstances. The information in this article is not intended to provide legal or tax advice. To ensure that your own circumstances have been properly considered and that action is taken based on the latest information available, you should obtain professional advice from a qualified tax and/or legal advisor before acting on any of the information in this article.*

*This information has been prepared by Rosemary Westie and Troy Iwanik who are Investment Advisors for HollisWealth®. Opinions expressed in this article are those of the Investment Advisors only and do not necessarily reflect those of HollisWealth. HollisWealth® is a division of Industrial Alliance Securities Inc., a member of the Canadian Investor Protection Fund and the Investment Industry Regulatory Organization of Canada.*



	SELL SECURITIES FOR CASH. DONATE AFTER-TAX PROCEEDS	DONATE SECURITIES DIRECTLY TO CHARITY
Original Cost of Securities	\$10,000.00	\$10,000.00
Current Market Value	\$50,000.00	\$50,000.00
Capital Gain	\$40,000.00	\$40,000.00
Tax on Capital Gains @ 49.80%	\$9,960.00 <sup>1</sup>	\$0.00
Donation Amount After-Tax	\$40,040.00	\$50,000.00



<sup>1</sup> This assumes a 49.8% marginal tax rate for BC, highest tax bracket. For calculating capital gains tax, this rate is applied to 50% of the capital gain.

<sup>2</sup> This is an estimate based on CRA rates, 2019; always consult a tax professional before proceeding.

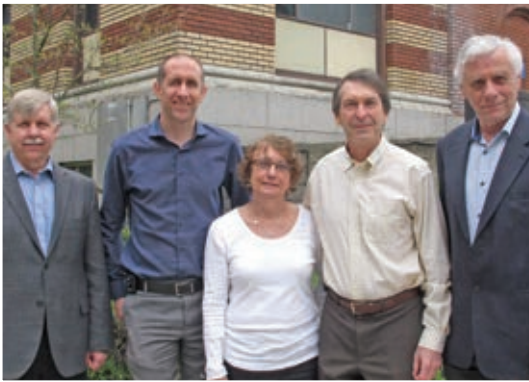


## Research Across Canada

**“Thank you”** to the inspiring and dedicated Canadian researchers and health care providers who are contributing to the development of ground-breaking interventions, improved diagnostic techniques and understanding of scleroderma. Though the disease may be rare, the community supporting quality of life improvement for people impacted by scleroderma provides a broad source of hope. Here is a sample of some of the outstanding research efforts taking place in our own country.

To read more about research across Canada, please visit the *Research section* of our website [sclerodermabc.ca](http://sclerodermabc.ca).

### VANCOUVER, BRITISH COLOMBIA



**DR. JAMES DUNNE, DR. KEVIN KEEN**

**Scleroderma Clinic, Mary Pack Arthritis Program, Vancouver Coastal Health Authority (Vancouver General Hospital)**  
**Scleroderma Lung Clinic & Combined Scleroderma Pulmonary Arterial Hypertension Clinic, Pacific Lung Health Centre, Providence Health Care (St. Paul's Hospital)**  
**Providence Health Care Research Institute (St. Paul's Hospital)**

*Scleroderma Association of B.C. Research Project* is a community-based patient-oriented study to discover the impact of certain RNA molecules on biochemical processes at the cellular level with the goal of targeted therapeutic interventions to correct the disturbed cycle of regeneration in heart, lung, and skin tissues in scleroderma and lung tissue in idiopathic pulmonary fibrosis.

### CALGARY, ALBERTA



**DR. MARVIN FRITZLER**

**University of Calgary**

*Autoantibodies to a novel Rpp38 (Th/To) derived B-cell epitope are specific for systemic sclerosis and associate with a distinct clinical phenotype:* This study screened certain proteins that target autoantibodies in scleroderma patients, and evaluated

their clinical relevance. Detection of antinuclear antibodies and specific autoantibodies is important in the diagnosis and classification of SSc. The new RNA P protein holds promise to increase the sensitivity in the detection of autoantibodies, enhancing the diagnosis of SSc.



**DR. JAN STOREK**

**University of Calgary**

*Myeloablative Autologous Hematopoietic Stem Cell Transplantation for Severe Scleroderma: Long-Term Outcomes 6-11 Years after Entry on a Randomized Study Comparing Transplantation and Cyclophosphamide:* This study reports subject

survivor status, late effects and outcomes 6-11 years after the Scleroderma: Cyclophosphamide or Transplantation (SCOT) trial. This follow-up analysis demonstrates the clinical benefits of stem cell transplantation (HSCT) remain 6-11 years after the study. Survival and functional status were significantly better with HSCT, and continuing control of scleroderma was demonstrated by 92% of transplant survivors remaining free of disease.

## WINNIPEG, MANITOBA



**DR. ADA MAN**  
University of Manitoba

*Development and validation of a patient-reported outcome instrument for skin involvement in patients with systemic sclerosis:* This study developed a patient questionnaire to assess the skin-related quality of life in patients with systemic sclerosis. It shows reliability and validity and is complementary to existing measures of SSc skin involvement with emphasis on the patient's experience.

## MONTREAL, QUEBEC



**DR. JEAN-LUC SENEAL**  
CHUM Research Center  
Scleroderma Research Chair  
University of Montreal

*The pathogenic roles of autoantibodies in scleroderma:* At the request of the Journal of Scleroderma and Related Disorders (JSRD), which is the only international medical journal

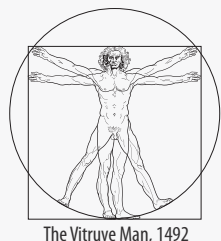
dedicated solely to scleroderma, this study analysed how autoantibodies in the blood of scleroderma patients contribute to disease mechanisms. It was concluded that anti-topoisomerase I is the single autoantibody with the most evidence in favor of a pathogenic role in scleroderma, followed by anticentromere autoantibody. A better understanding of how these autoantibodies contribute to scleroderma manifestations may lead to improved therapies.



**DR. SABRINA HOA**  
CHUM Research Center  
University of Montreal

*Association between immunosuppressive therapy and course of mild interstitial lung disease in systemic sclerosis (SSc-ILD):* The objective of this study was to determine whether use of immunosuppressive drugs, namely

cyclophosphamide or mycophenolate mofetil, was associated with an improved course of lung disease in patients with normal or mildly reduced lung function. In this setting, exposure to these immunosuppressive drugs at the beginning of the study was associated with higher forced vital capacity values and a lower risk of progression among patients with mild interstitial lung disease at two years of follow-up. These data suggest a possible window of opportunity to preserve lung function in mild SSc-ILD.



The Vitruve Man, 1492

**DR. HEENA MEHTA,  
DR. MARIKA SARFATI**  
CHUM Research Centre  
Immunoregulation Laboratory  
University of Montreal

*From bench to bedside to bench...*

Recent acquisition of cutting-edge instrumentation enables the comprehensive study of the immune profile in blood and at barrier tissues. 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.



**DR. MARIE HUDSON**  
Lady Davis Institute for Medical  
Research, Montreal Jewish  
General Hospital

*Generation of a Core Set of Items to Develop Classification Criteria for Scleroderma Renal Crisis Using Consensus Methodology:* This study helped to generate a method of

classification for scleroderma renal crisis (SRC), which will be used in future phases of this project to develop classification criteria for scleroderma renal crisis.

# Research Across Canada

## MONTREAL, QUEBEC



**DR. CELIA GREENWOOD**  
Lady Davis Institute for Medical Research, McGill University

*Whole-genome bisulfite sequencing in systemic sclerosis provides novel targets to understand disease pathogenesis:*

This study investigated the role of DNA methylation in SSc and will be used for future studies on the same subject.



**DR. BRETT THOMBS**  
McGill University

*Scleroderma Patient-centered Interventional Network (SPIN),* maintains an international patient database used to study psychosocial and rehabilitative online programs and toolkits to improve the health-related quality of life of patients living with scleroderma.

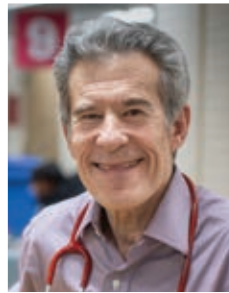
## TORONTO, ONTARIO



**DR. SINDHU R. JOHNSON**  
Toronto Scleroderma Program - University Health Network & Sinai Health Systems

*Nintedanib for Systemic Sclerosis Associated Interstitial Lung Disease:* An international, multicenter study that found treatment with nintedanib

(OFEV®) slows progression of scleroderma-associated interstitial lung disease. Thanks to this study, OFEV® was approved by Health Canada and is now available in Canada.



**DR. RONALD LAXER,  
DR. ELENA POPE**  
Toronto Hospital for Sick Children

*Developing comparative effectiveness studies for a rare, under-studied pediatric disease:* This study investigated the feasibility of conducting a comparative effectiveness study for juvenile localized

scleroderma, a rare pediatric disease, for which there is limited evidence on best therapy.

## LONDON, ONTARIO



**DR. JANET POPE**  
St. Joseph's Health Care, London ON  
Western University, London ON (London Health Sciences Centre)

*Changes in skin score in early diffuse cutaneous systemic sclerosis are associated with changes in global disease severity:* This study

determined that at 1 and 2 years, skin scores showed overall improvement in early diffuse cutaneous scleroderma patients, improving prognosis and quality of life.

## HALIFAX, NOVA SCOTIA



**DR. EVELYN SUTTON**  
Dalhousie University

*Clinical correlates of faecal incontinence (FI) in systemic sclerosis: identifying therapeutic avenues:* This study established the prevalence and severity of FI in SSc, its association with other intestinal manifestations and potential predictors of FI, and its

impact on quality of life. FI was found to be common and often severe in SSc. Loose stools, SIBO, constipation and urinary incontinence were strongly associated with FI. Other than targeting anorectal dysfunction, concomitant treatment of clinical correlates could lead to improvement in FI and quality of life in SSc.



# Join our **virtual** events across British Columbia

Due to COVID-19 we will not be able to be together physically this June – Scleroderma Awareness Month, but we can come together as a province to support the ground-breaking scleroderma research taking place here and continue to promote awareness about living with this rare disease.

100% of funds raised supports scleroderma research in BC and Canada! We welcome you to support us this June, wherever you are in BC at [sclerodermabc.ca](http://sclerodermabc.ca).



## WAYS TO PARTICIPATE

### 1 GENERAL SUPPORT

Use the **MAKE A DONATION** button at the top of the page at [sclerodermabc.ca](http://sclerodermabc.ca)

### 2 OVERALL WALK/RIDE SUPPORT

Follow the



Click here

Use the

**DONATE NOW** button

### 3 SPECIFIC TEAM SUPPORT

Scroll down and click on a Fundraising Team or Individual Fundraiser

Use the **DONATE TO TEAM** button

### 4 OR JOIN THIS TEAM

button to help with fundraising efforts, then forward the link to the people that care about you.

You will receive a receipt for the full amount of your donation. This fundraising campaign is part of the walks organized in June for Scleroderma.

## Questions regarding participation?

Contact SABC's Yvonne Alexander at

[yvonnea@shaw.ca](mailto:yvonnea@shaw.ca)

Capture your energetic, adventurous or simply creative "moving" in pictures or videos and send them to SABC's Alex MacDonald at [alexandria.cmacdonald@gmail.com](mailto:alexandria.cmacdonald@gmail.com). Include your location and any other details you want to provide. SABC will create and post a slideshow of everyone moving to demonstrate how we all came together in the month of June to raise awareness and funds to support scleroderma research.



*"In the past 2 years Islanders have participated in walks, raising much needed research funds.*

*We hope to continue to grow, albeit virtually, this year!"*

*– Linda and Jackie, Victoria and Campbell River*

*"Over the last 8 years, we have asked family, friends, and anyone and everyone we know, to support the research.*

*We need to find a cure for this horrible disease.*

*We couldn't do it without your support"*

*– Rosanne, Vancouver*

*"Please join me on Team Valemount Mountain*

*– Teresa, Valemount*

*"Don't see a team for your specific region?*

*Choose the Walking for a Cure – Anywhere in BC Team!"*

*– Yvonne*



**With the support of you, our families, friends and community, we can all make this event truly special.**

**FOR MORE DETAILS PLEASE VISIT**

**[sclerodermabc.ca](http://sclerodermabc.ca)**

# SYSTEMIC SCLEROSIS- ASSOCIATED PULMONARY FIBROSIS

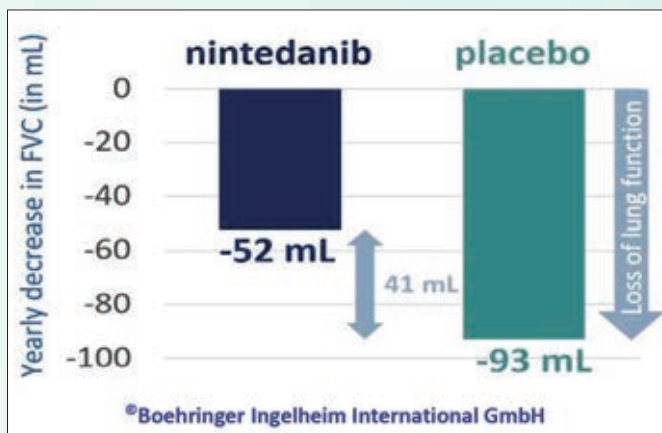
MARIELLE PELLETIER,  
R.N.



Scleroderma or systemic sclerosis (SSc) is 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 and fibrosis reach the lungs, it is called interstitial lung disease (ILD) or pulmonary fibrosis (PF). This is a common complication of systemic sclerosis, affecting more than 50% of patients. However, in only about 15% of patients is PF more severe. SSc-pulmonary involvement is often silent in the early stages of the disease. In more severe cases, patients may complain of unexplained fatigue, decreased exercise capacity, shortness of breath and/or persistent dry cough.

There are now medications available for the treatment of SSc-PF. The first are immunosuppressive drugs that work by decreasing the activity of the immune cells responsible for inflammation. Recently, a randomized trial showed that an anti-fibrotic drug, nintedanib (OFEV®), is effective in slowing the progression of scleroderma-associated PF compared to placebo. Slowing the progression of PF in SSc patients seems to work best when immunosuppressant and nintedanib are combined as treatment.

A study to compare nintedanib with placebo for patients with scleroderma-related lung fibrosis (SENSCIS® study, 1199.214)



Nintedanib has been approved by Health Canada since the fall of 2019 for patients with scleroderma-associated PF. Nintedanib is the only molecule approved by Health Canada to treat PF in SSc patients. Although this medication is new in the treatment of scleroderma, it has already been approved since 2015 for the treatment of idiopathic pulmonary fibrosis or IPF (a disease which involves progressive scarring of the lungs of unknown cause-hence the term idiopathic).

In patients with scleroderma-associated PF, fibrosis affects the alveoli in the lungs that allow oxygen to be transported from the lungs to the blood, and carbon dioxide to be removed from the blood. Because of scarring and stiffness, the lungs can no longer perform these tasks effectively. Nintedanib acts on the healing process by blocking certain kinases, enzymes that are partly responsible for this process.

Currently, certain private insurance companies can reimburse the medication, and we hope that patients will eventually be able to obtain reimbursement from their Provincial Health Insurance Plan.

**For more information on SSc-associated PF, please consult the educational fact sheet on Scleroderma Quebec's website at [sclerodermie.ca/en](http://sclerodermie.ca/en) in the section About scleroderma under Articles and publications.**



PRESENTS



# SPIN-HAND

The first publicly accessible SPIN toolkit.

## SPIN-HAND for hand function

The SPIN Hand Function (SPIN-HAND) toolkit provides exercises to improve hand function with sections to help you develop a personalized program, set goals, and track your progress. Instructional videos demonstrate how to perform each exercise properly with pictures to illustrate common mistakes.

The SPIN-HAND program was first tested by the SPIN cohort.

Explore the following features  
by logging in or registering at:

**tools.spinsclero.com**

01

4 modules with different exercises to address common hand limitations.



Thumb



Fist



Extension



Wrist

Detailed video instructions for hand exercises, with 2 difficulty levels.

02



Under each video, you will find a written transcript if ever you would like to refer back to what was said within the video, and tips to avoid common exercise mistakes with visual references.

03

Video testimonies of other people's experiences with hand limitations in scleroderma.



**More toolkits to come in 2021!**





# *Tips for Protecting You and Your Estate*

## *(and maintaining family harmony)*

**Waiting to create your estate plan until you are old and dependant can open your estate up to a challenge by beneficiaries alleging your Will is invalid on the basis of diminished capacity or the undue influence of one of your beneficiaries.**

It is recognized that someone who is old, frail, and dependant is more susceptible to the influence of others and to those looking to receive a benefit you otherwise would not have left them when you were younger with full command of your faculties.

### **ENSURE YOU HAVE AN ESTATE PLAN IN PLACE WHICH INCLUDES A:**

- 1. WILL**
- 2. GENERAL ENDURING POWER OF ATTORNEY**
- 3. REPRESENTATION AGREEMENT FOR HEALTH CARE WITH AN ADVANCE DIRECTIVE**



**Author: Mary Anne Richter of Mary Richter Law Office (Vancouver, Kerrisdale Area)**

Mary is a solicitor with a general practice of law (wills and estates, real estate conveyancing, and corporate). She has practiced law on the West side of Vancouver for over 30 years.

**If possible, discuss with your family how you intend to distribute your estate and have these documents prepared by a legal professional so that there are no errors. Review and update your documents regularly to ensure they reflect your current wishes.**

## 1. WILL

- Choose Executors who reside in Canada. Choosing non-resident Executors can pull your estate out of Canada for income tax purposes. The residency of your estate is determined by the residency of a majority of your acting Executors.
- Specify the amount to be paid as an Executor's fee. Beneficiaries often do not value the work carried out by the Executor in administering the estate and can refuse to sign a consent, requiring costly court approval. If you specify in your Will the amount or percentage to be paid to your Executor, the consent of the beneficiaries is not required. The maximum fee payable to an Executor is 5% of the gross value of your estate.
- Executors can be beneficiaries. If you appoint more than two Executors, include a statement if you want them to make decisions by majority, otherwise unanimous decisions are required.
- Your Will distributes assets in your sole name. These are the assets on which the 1.4% probate fee is charged. Registering your assets in joint names can expose them to creditor and matrimonial claims being made against the joint owners and can trigger capital gains tax if not properly done. However jointly owned assets with named beneficiaries (such as TFSA, RRIF, RRSP and Life Insurance) automatically pass to those beneficiaries.
- Make it clear if you want loans forgiven – if you want loans repaid to your estate make sure you have loan documentation and complete records which confirm the balance outstanding.
- If you want gifts you made to one of your beneficiaries during your lifetime equalized with the other beneficiaries, make it clear leaving a cash gift off the top to the other beneficiaries who did not receive such gifts during your lifetime.
- Include a distribution list of your personal effects among your beneficiaries. This helps to eliminate any conflict that may arise if your beneficiaries themselves are tasked with dividing and agreeing on the value of your items.
- Your current Will is not revoked if you legally marry. Therefore, you should review it if you legally marry to make sure it still reflects your wishes.
- If you separate from a common law spouse, your Will will be interpreted as if your separated common law spouse predeceased you and will be excluded from receiving anything. The rules are slightly different if you separate from a legal spouse. If you want any separated spouse to receive something, you need to make your intentions clear.
- When a Will is probated it becomes public. Be sensitive to what you write as you can cause pain and humiliation if you use your Will to criticize someone you perceive has wronged you. Your grievances and reasons for distributing your estate the way you do can be described in a private Statutory Declaration rather than in your Will. It is important to document your reasons if you are giving a smaller share to your spouse or children, (the people who can apply to vary your Will - see below). Your reasons just do not need to be detailed in your Will.
- Your Will (on your death) is widely circulated. If it is probated, it is sent to all beneficiaries as well as your next of kin, being the people who would have received your estate if there was no Will. Whether it is probated or not, it is sent to the third parties with whom you have dealings such as the Canada Revenue Agency, financial institutions where you hold assets, utility suppliers, etc.



**There is no  
“Reading of the Will”  
as you see on TV  
where beneficiaries  
are assembled to be  
told what they are to  
receive under your  
Will. Sounds strange to  
mention this, but it is a  
question often asked!**

## Wills Variation Claims (Will Challenges)

**In British Columbia, only a spouse and natural or legally adopted children (not stepchildren, grandchildren, siblings, etc.) have the right to apply to vary the distribution in your Will and seek a larger share of your estate.**

A spouse can be a person you are legally married to or a person with whom you have been in a marriage-like relationship for at least 2 years, and includes same-gender relationships. If you are concerned that someone may claim after your death to be your common law spouse, you should consider swearing a Statutory Declaration setting out particulars of your relationship with that person and reconfirm those facts on a regular basis so that it can be used to defend your estate against a claim.

One of the most common reasons people have for challenging a Will is disappointment or surprise at its terms. For example, your children might expect that you will split your assets equally among them. When they later realize that you have allocated assets to third parties such as charities, friends, or other members of your family, they could attempt to challenge that.

Estate litigation by a spouse or children not only will pit your loved ones against each other, it also can tie up your estate for years and cost thousands of dollars. Try to avoid estate litigation by treating your spouse and children fairly. Resist using your estate distribution to exact revenge, or the temptation to leave more of your estate to a needy child thereby penalizing your more successful children.

## 2. GENERAL ENDURING POWER OF ATTORNEY – FINANCIAL DECISIONS

A Power of Attorney is the document by which you authorize an agent to act on your behalf on financial matters. It is an important document to protect you and to make sure your financial affairs are managed if you are unable to look after them yourself.

**“General”** as opposed to **“Limited”** means it covers all financial matters.

**“Enduring”** means it is in effect and can be used from the date you sign it until your death or revocation.

If you lose your capacity to manage your finances before executing a Power of Attorney, there is a lengthy and costly court process to have someone appointed to look after your financial affairs.

## 3. REPRESENTATION AGREEMENT FOR HEALTHCARE WITH AN ADVANCE DIRECTIVE – MEDICAL DECISIONS

A Representation Agreement for Healthcare is the document by which you appoint someone (your medical advocate) to make medical decisions for you if you are unable to make your own.

As we have all seen with the recent pandemic, a person of any age can need medical assistance. Accidents and other unforeseen events do happen. Appointing a medical advocate in a Representation Agreement for Healthcare will mean that you have a lasting person of your choosing, who knows you, in case you need someone to advocate for medical care for you.

Not only does your appointed representative advocate for your medical treatment and care but he/she can also discontinue life support if you have no reasonable chance of recovery. The Advance Directive is a legally binding document by which you can provide instructions with respect to giving or refusing consent to health care treatment or procedures. You can specify in your Advance Directive the kind of medical treatment you want, where you wish to live (i.e. your own home with care brought in or in a care home) and other personal wishes regarding your medical care.

**If you don't have** a Representation Agreement for Health Care with an Advance Directive, then the doctors and health care providers (rather than you) will select someone to be your Temporary Substitute Decision Maker (a “TSDM”) according to a list in the Health Care Consent Act.

The first on the list is a spouse, then an adult child, and if you have more than one adult child, the doctor can choose the child who will be making your health care decisions. If you have no spouse and no children, next in line are parents, followed by siblings, then grandparents, then grandchildren, and then other relatives.

- The TSDM is a ‘temporary’ decision maker. He/she cannot make decisions about your personal care. A TSDM cannot decide where you will live, or whether you will have additional personal care assistance such as with grooming, diet, etc. Proper personal care is very important. It affects our quality of life. It can be particularly important for someone who has a chronic illness or a condition such as Alzheimer's or Parkinson's disease
- A TSDM is only there to make a specific decision, typically in an emergency type situation, and is only able to make a medical decision for you if your doctor determines that a decision needs to be made and you are not capable of making the decision yourself
- There is no continuity of decision making and there is no ongoing medical advocacy with a TSDM



# 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.

### VANCOUVER

We are seeking a volunteer representative.  
Please contact Rosanne Queen at **604-984-9425**  
or by email at **[rq.sabc@telus.net](mailto:rq.sabc@telus.net)**

### Campbell River

Jackie Alexander  
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### Chilliwack

Kelly Grant  
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### Creston

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### Williams Lake

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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.

SCLERODERMA ASSOCIATION OF B.C.  
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