SCLERODERMA ASSOCIATION OF B.C.

The Bulletin

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Scleroderma Association B.C.

Moving to Cure *Scleroderma*

B.C.'s Scleroderma Clinic

Summary

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A Word from Our President

Finally, after two years of 'virtually' spreading awareness while raising funds for research, our June is Scleroderma Month events will be in-person. Yes, we will all be incredibly careful at these outside events that are so well deserved and needed after being isolated from each other for so long. We have two new in-person walks this year joining the Victoria and Vancouver events. They will be in Kamloops and in Surrey. Valemount's Moving to Cure Scleroderma will once again be virtual. A big Thank You to all the organizers and their helpers.

I really hope SABC members, along with their family and friends, support this June's **Moving to Cure Scleroderma**. And not only by donating to research but by joining us in-person. I realize there may not actually be a walk right in your local community, so a nice drive to the nearest town might be in order! Ask your friends and families to join you safely outside this year. Nothing is more inspiring then seeing a walking wave of supporters **Moving to Cure Scleroderma**. I have already ordered clear weather for all our events!

As always, SABC is committed to every penny raised (not that we have those anymore) to support research here in B.C. and across the country. We are so lucky that for such a rare disease, there are so many unique and important research projects happening in Canada.

Raising awareness also involves the importance of educating medical students about scleroderma. Valerie Doyon, a SABC board member and third year medical student has been working long hours to put together 5 educational podcasts to share with medical students at UBC. These podcasts (available on the SABC website) consist of both a patient and a doctor sharing their experiences about a specific scleroderma topic. Titles include: Sclerodactyly, Raynaud's Phenomenon, The Role of Surgery, Esophageal Dysmotility & Interstitial Lung Disease. You can find the currently completed ones under the website tab ***Videos***.

The first ever article spotlighting **B.C.'s Scleroderma Clinic** is contained within! For those of us who attend the Clinic on a regular basis, we know only too well how lucky we are to have Fran & Co. running the show there.

SAVE THE DATE: In October the SABC hopes to reinstate our in-person **AGM & Conference**. Keep an eye out for our email notifications and website updates for the program and how to register. We will continue to have a on-the-day live-streaming of the Conference as well as Presenter sessions recorded for future viewing.

Once again, I would like to thank Sclérodermie Québec for creating and sharing their educational sheets. Having up-to-date and reliable information is so important; Dr. Google is not always a reliable source. Find even more educational sheets on our website, under ***Articles & Publications***.

SCLERODERMA

Awareness breeds **compassion**,

which leads to **funding**

which leads to a **cure**.

Moving to Cure

SCLERODERMA

JUNE IS SCLERODERMA AWARENESS MONTH!

With the participation of patients, their families and friends, each June we raise awareness of scleroderma and funds for research. We come together as a province and support the ground-breaking research in B.C. and elsewhere in Canada and continue to promote awareness about living with this rare disease.

This year our live events provide us with the long-awaited opportunity to participate in-person and in the same location, connect with others over a shared cause.

Please join us this June for all the fun!

With the support of you, our families and community, we can all make the June events truly special.

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

*** The Scleroderma Association of B.C. will accept donations before (on-line) or during these events. 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.**

KAMLOOPS

SUNDAY, JUNE 5TH / WALK

RIVERSIDE PARK - 100 Lorne St, Kamloops, BC V2C 1V9

KAMLOOPS is excited to host our first in-person walk on June 5th! Our goal this year is to create awareness for scleroderma and raise money for critical research in B.C. We look forward to collaborating and connecting with our wonderful community. Together, we are better.

JEN BECKETT - KAMLOOPS

VICTORIA

SUNDAY, JUNE 12TH / WALK

WEST SHORE PARK - 1767 Island Hwy, Victoria, BC V9B 1J1

We are looking forward to seeing everyone in-person at our Victoria "**Moving to Cure Scleroderma**" 2022 Walk on June 12th. It's been a difficult few years to fundraise for our cause so we are excited to join together again and set some new goals to raise awareness and funds for research for scleroderma. My family and friends are gathering around to make this the best event yet, and we warmly welcome you to join us and help spread the word about this debilitating disease. **Thanks for your support!**

LINDA BARNES - VICTORIA

I have been able to participate in these events for the last few years and each time I am surprised at how much love and support is received from friends and family. I look forward to joining this year's walk in Victoria for scleroderma awareness.

JACKIE ALEXANDER - CAMPBELL RIVER

For more details on the schedule of the walks and to register on-line, please visit

sclerodermabc.ca

A big thank-you to the walk coordinators who welcome you to join them and support us all this June.

Not able to make any of June's organized walks? No problem!

Wherever you are in B.C. please visit sclerodermabc.ca and help raise funds for research.

100% of funds raised supports scleroderma research here in B.C. and across Canada!

VANCOUVER

SUNDAY, JUNE 19TH / BIKE RIDE/WALK

STANLEY PARK - Vancouver, BC V6G 1Z4

David and I came up with the idea of starting a ride to raise awareness and funds for research on our way home from a two-month cycle tour of Ireland. We could not have imagined the impact that has come from the people that have joined us over the years and the generous donations that contribute directly to scleroderma research. We are all making a difference and are looking forward to really getting together this year to celebrate our 11th year.

ROSANNE & DAVID QUEEN - VANCOUVER

SURREY

SUNDAY, JUNE 26TH /WALK

TYNEHEAD PARK - Surrey, BC V4N 2E2

We are excited for our first "Moving to Cure Scleroderma" in the Lower Mainland. We invite everyone to help us raise money for research for scleroderma.

CHELSEY FITZPATRICK-LINDSAY - COQUITLAM
KELLY GRANT - CHILLIWACK

VALEMOUNT

ALL OF JUNE / VIRTUALLY 'MOVING'

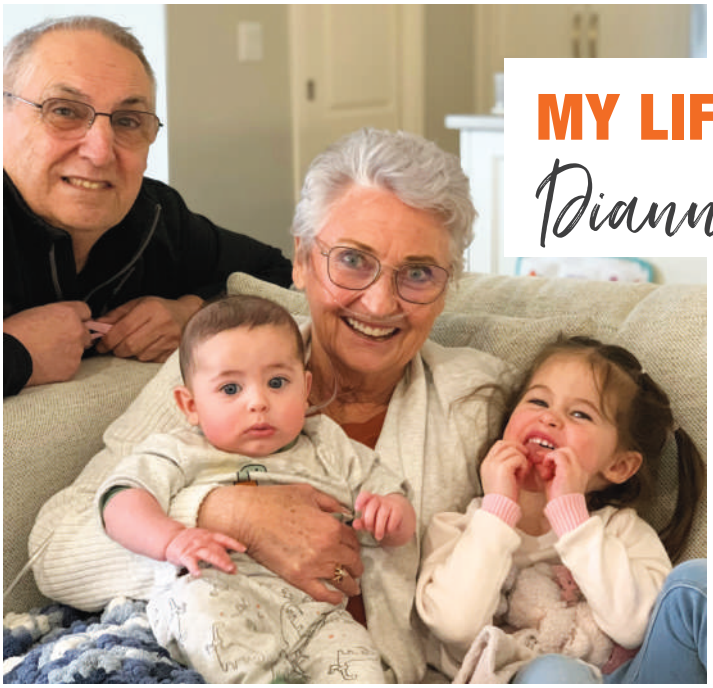
This will be the 5th year spreading awareness and raising money for research. We will be hosting another Virtual event "Moving to Cure Scleroderma". Let's see how many family members, friends and other communities we can get to participate.

TERESSA COLOSIMO – VALEMOUNT & NORTHERN RURAL COMMUNITIES

ANYWHERE IN B.C.

ALL OF JUNE / VIRTUALLY 'MOVING'

Can't get to any of the in-person walks this year? We'll miss seeing you, but it is still easy to continue your much needed support this month!



MY LIFE WITH SCLERODERMA

Dianne's Story

I was diagnosed with scleroderma in 1992 but had symptoms for about two years before that. My skin had been feeling tighter than normal and I was incredibly itchy. I was referred to a dermatologist who diagnosed me after a simple blood test. Finally, my symptoms made sense. The tight skin, itchiness, fatigue, sore joints, Raynaud's and depression, all of course part of scleroderma, but on their own I felt maybe I was just feeling sorry for myself! I just had never put the symptoms together. The biggest mistake I made was to go to the library and research scleroderma, an illness I had never heard of. The encyclopedia diagnosis suggested I had about 5 years to live. I was so angry that I got this disease when I was only 40 years old and at a point in my life where my son was in high school, I had a great job and we were enjoying doing things as a family. I managed to work for another year and then had to take a disability pension. I spent the next two years very depressed and spent a lot of time in bed.

I was referred to a rheumatologist who introduced me to one of the founders of the SABC who was very helpful making me feel that I was not alone with this illness. Becoming a SABC Board Member in 1993 helped me look at things with a different perspective, be glad to be alive and get the opportunity to give back and encourage others so that they could also move forward. I was given the opportunity to be involved with UBC medical students by participating in their new Health Sciences Fair. The Fair's objective was to introduce medical students to patients with different illnesses. Our scleroderma booth was larger than life with 8 scleroderma patients sharing their stories. We had pictures and pull up banners but most importantly we had scleroderma patients allowing students to touch their hands and ask questions. No question was unimportant or off limits. The Health Fair experience led to inviting medical students to SABC's annual AGM & Conference where they sit with patients and really get to understand this disease. Based on the student's uplifting comments about their experience, I know we have made a difference in how these future doctors will care for scleroderma patients.

After about 5 years my hands started to soften and I was once again able to go back to bowling and golfing, two sports I really enjoy. I also went back to coaching 5 pin bowling. Although my health was now stable, I told my rheumatologist I could feel a crackle in my chest. An x-ray confirmed I was in the beginning of lung involvement with a diagnosis of pulmonary fibrosis. My progression was very slow and things were stable for a long time. Then I got a phone call in October 2020 from my lung specialist to inform me that they had seen blood clots on my lung echo. I was put on blood thinners right away and went through another series of tests. Within a week, I was put on oxygen and referred to a cardiologist at the Pulmonary Clinic at VGH with a diagnosis of PAH (pulmonary arterial hypertension).

I think most scleroderma patients have some issues with indigestion but over the years mine has become worse. I have had my esophagus dilated 4 times. I am on some very strong drugs for GERD (Gastric Esophageal Reflux Disease) and now I am becoming more concerned as I have lost 40 lbs in the last 1 ½ years. Some specialized tests are going to be done; I am confident we will find a solution soon. So now my heart, lungs and digestive track are involved. If this was 30 years ago, I would have simply crawled back into bed and felt sorry for myself once again.

It is now March 2022 and my prognosis is fine. The specialists are working on my GERD issues, my cardiologist has me on Tadafil, which is keeping my arteries open, and I have such an amazing team of doctors at the Scleroderma Clinic at St. Paul's Hospital. My oxygen needs have increased but are manageable. I have an oxygen concentrator in my home, I have liquid oxygen when I am out and about, and I purchased an Inogen battery operated oxygen system so that I can continue bowling with my special backpack and hopefully get back to golfing this spring.

I truly believe that wellness is a state of mind. I can either give in to this disease or I can fight it on my terms. I am not giving up and will let you in on my next chapter whatever that may be.

DIANNE MCPHEE

WHAT IS B.C.'S SCLERODERMA CLINIC?

The Scleroderma Clinic is located within the Pacific Lung Health Centre, on the 8th Floor, Providence Building at St. Paul's Hospital in Vancouver. It should be clarified that the Scleroderma Clinic is a combined clinic, specifically for patients diagnosed with scleroderma and Interstitial Lung Disease (ILD) secondary to scleroderma.

Scleroderma patients, their families and their health care professionals might first learn of the good work being done at the Scleroderma Clinic at the time of diagnosis. Perhaps due to little or no familiarity with this rare condition, a patient's general practitioner, rheumatologist or dermatologist may request a referral appointment at the Scleroderma Clinic.

Fran Schooley, the Patient Educator/Clinic Coordinator/ Research Assistant for both the ILD Clinic and the Scleroderma Clinic was interviewed for this spotlight article.

Linda Barnes, a patient of the Scleroderma Clinic and living on Vancouver Island provided the ever-important-patient's perspective of attending the Clinic.

BACK IN TIME. In 2004 the Pacific Lung Clinic was formed at St. Paul's Hospital. At that time, respirologist Dr. Pearce Wilcox was seeing the majority of scleroderma patients. It was quickly noted that scleroderma patients attending the clinic needed their own clinic due to the time needed to tend to the possible array of issues they presented with; not the least of which was the lung component that may have gone unrecognized. A group of physicians started the monthly Scleroderma Clinic on Monday afternoons in collaboration with Vancouver rheumatologist, Dr. Jim Dunne. The Department of Rheumatology used the clinic to teach their medical residents and fellows, the "anatomy of a scleroderma patient".

For two years, there was a steady routine of moving 3-4 patients from the Lung Clinic to the monthly Scleroderma Clinic. The oversight of 100-120 scleroderma patients was somehow manageable. Within 4 more years however, there was much more need. In May 2012, the now hospital-run Scleroderma Clinic was upped to 2 clinics per month.

Spotlight on B.C.'s Scleroderma Clinic

It's terrifying to be diagnosed with a rare disease and to find your life changing in so many ways as you struggle to understand what's happening to your body, your life and your future. In 2012, after a long year of tests and anxiety, with a suspected diagnosis from my family doctor of scleroderma, I found myself on the ferry from my home in Victoria, heading to the office of Dr. Dunne. I had a million questions and not many answers.

Almost 20 years later, over 200 returning patients attend the clinic from not just all over B.C., but from across Canada as well. While we celebrate the increased awareness of the clinic by our health care professionals, their patients and families, we recognize the challenge of being able to meet the need. The referral list contains well over 400 names, with patients currently waiting an average of 5 months for a non-urgent appointment. The clinic's goal is to include at least one new patient at each clinic. There are presently 3 clinics each month.

Generally, I attend the clinic every 6 months but sometimes more often depending on any particular issues that may come up. As an out-of-town patient, there are some logistics to work through. First, I have Fran or my family doctor provide me with a "pink sheet", the provincial form that allows me to access the ferry free of charge. Sometimes my husband comes with me, so his free fare is also included on the form. The same form can also be used for a reduced fare on Harbour Air flights.

I always ask Fran to schedule my Pulmonary Function Test, or any other tests a little later in the morning of my clinic day, as I ferry over the night before so I can visit with my grandkids. It is nice not to have to rush in the morning! My mindset about my Clinic day is that I'm getting a little holiday, travelling free of charge and just happen to have a few appointments as well. I even try to arrange something fun to do in Vancouver while I'm there!

BACK TO THE CLINIC APPOINTMENT. The clinic schedules scleroderma patients who have been diagnosed with ILD in one of the three Monday mornings each month, depending on their scleroderma-related issues. Patient referrals are reviewed by the clinic's Coordinator, Fran.

A team of physician specialists rotate through the clinic each Monday,

- Dr. Pearce Wilcox – Respirologist
- Dr. Chris Ryerson – Respirologist
- Dr. Alyson Wong - Respirologist
- Dr. James Dunne - Rheumatologist
- Dr. Iman Hemmati - Rheumatologist
- Dr. Hyein Kim - Rheumatologist

Patients complete a questionnaire each time they attend the clinic, after which they receive at least an hour of a specialist's time, (respirologist and rheumatologist). The data captured on the questionnaire is used to:

1. Help the doctors identify issues and discuss health concerns related to scleroderma
2. Introduce patients to and invite them to join any recruiting clinical research trials at the clinic. The answers to the questionnaire become part of a database used for research purposes by associate doctors and scientists
3. Track each patient's individual progress

After the tests are complete, I head to the Clinic with my results in hand. Having the Clinic at St. Paul's means there is rapid access to testing and results. If issues with any of my results present themselves, I have been sent for further tests on the same clinic day, without having to wait weeks for the same test in Victoria. However, if needed, Fran can also arrange for other tests to be completed in Victoria upon my return. Fran then follows up with those results and gets the information to the Clinic doctors. I have even called Fran from home to check in with one of the doctors about a particular problem and it's a relief to get a quick response to concerns specific to scleroderma.

At the Clinic, I chat with Fran while filling out a form to assess any changes in my health. Then I am installed in an examining room where the doctors who see me cycle through, instead of me having to move from room to room.

Can scleroderma patients who have no ILD be seen by the team at the Scleroderma Clinic? Not at this time, no. However, the rheumatologists at the clinic, with their scleroderma-specific experience, act as a referral resource, pointing patients and their families in the right direction for care. St. Paul's hospital has a Rapid Access Clinic on the 3rd floor. Here, patients are seen and then subsequently referred to a rheumatologist, and depending on their symptoms, a dermatologist, cardiologist, gastroenterologist, nephrologist, etc.

Patients who present with Pulmonary Arterial Hypertension (PAH), and are referred to the medical team at the PAH Clinic at Vancouver General Hospital usually continue to remain active members of the Scleroderma Clinic.

Spotlight on B.C.'s Scleroderma Clinic

This is a picture of the girls walking me to my appointments at St. Paul's!

THEY'VE GOT YOUR BACK. As mentioned above, often patients arrive for their referral appointment at the clinic from a situation in which their current scleroderma treatment was prescribed from a physician who may not have had much exposure to this rare disease. The Scleroderma Clinic provides patients with referral access to experienced staff who are not only willing to treat all the conditions associated with scleroderma but whose knowledge of scleroderma is constantly increasing. It is important to a patient's care to confirm the correct diagnosis in order to guide them on the path of effective treatment.

Generally, I'm followed by the same doctors but over the years I've been seen by all of them at various times. All are knowledgeable of scleroderma and work collaboratively to come up with any changes in treatment plans. They are respectful of my concerns and understand my particular situation and my wishes. They send all the information to my family doctor, always keeping him in the loop.

The clinic is well-known among the medical community and patients are benefiting from its excellent reputation. The success of the clinic, with its 6 physicians, draws the attention of and is attractive for other doctors to work and learn there. Senior fellows and medical students studying dermatology, rheumatology and/or respirology can complete a rotation at the clinic as part of their education. These future specialists can then become, after graduation, experienced physicians to whom the clinic can refer scleroderma patients.

The ongoing clinical trials at the clinic also interest the research community, attracting visiting doctors from across Canada. Research collaboration is a vital component of the doctors' practice, with different studies being conducted at St Paul's Hospital.

Also contributing to the clinic's success is the fact that it is a 'patient-driven' clinic, meaning patients become more knowledgeable of their health by the explanations/information provided by the physicians and the educational materials provided by Fran. Knowledge is power, as they say, but in the case of scleroderma patients, knowledge specifically facilitates independence.

This is not to say that patients are left on their own. Far from it. The clinic has the important patient support liaison role who is an actual person simply a phone call or email away, acting as a safety net for patients when they are back at home. With the help of the Coordinator in this role, patients are able to report any issues they are experiencing between clinic visits.

With the mandate of 'lets work this out together', the patient is able to constantly have access to the medical expertise of the Clinic staff. The physicians can efficiently respond to patients' questions, posed on their behalf by the Coordinator, as all specialists are working full-time in their own clinics. In a typical day this patient support liaison can receive 17-20 phone calls. Patients can be assured that when they need it, something is always happening in the background on their behalf.

Fast forward to 2022, and many, many ferry rides later and I am a knowledgeable patient, managing my disease well with the professional care of the Scleroderma Clinic at St Paul's. It feels like a lifetime ago when my consultation with Dr. Dunne resulted in a confirmed diagnosis of scleroderma with ILD. I'm so grateful I was then referred to the Scleroderma Clinic where I met "the Team" with their kindness, caring and expert advice, grounded by the administration skills of Fran! I feel incredibly fortunate B.C. has this Clinic model and such caring doctors to staff it.

The ultimate measurement of success is when a scleroderma pulmonary patient may become clinically stable and 'graduate' from the need to go to the clinic. An extreme example of this are patients who've received a lung transplant. When patients leave the clinic, they 'celebrate the stability' but are still reminded to ensure they are followed and monitored by their rheumatologist.

Thank You so much Fran, patient support liaison/clinic Coordinator extraordinaire, and to Linda for sharing her experience and for both being SABC's first Spotlighted Clinic contributors!

MICHELE GERVAIS
Vice president
Scleroderma Association of B.C.

HOW TO PREPARE FOR AN APPOINTMENT WITH YOUR HEALTH CARE TEAM

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

Here are a few tips to prepare for your appointment with your medical team.

MEDICATIONS

Have your medication list up to date and note if there are any side effects to the medications. If you are not taking some of the medications as prescribed, it is important to report this to your doctor so that a fair assessment can be made of the effectiveness of the treatments. Also, tell your doctor if you take over-the-counter medications or natural products.

BLOOD PRESSURE

Blood pressure should ideally be taken at home twice a week, or daily in patients at higher risk of developing scleroderma renal crisis. Bring your blood pressure log to help your doctor determine if further investigation is needed to detect a kidney complication.

BLOOD TESTS

If blood tests were prescribed at the last appointment, do them early enough (1 to 2 weeks) before the appointment so that the results can be available at the time of the appointment.

HOW TO PREPARE FOR AN APPOINTMENT WITH YOUR HEALTH CARE TEAM

SYMPTOMS OF SCLERODERMA

Take note of any change in symptoms associated with the disease. For example, have there been any new symptoms? Have the symptoms worsened, or have they improved following a change in treatment? More specifically for each symptom, the following details are particularly relevant:

- **Raynaud's phenomenon:** frequency and duration of attacks;
- **digital ulcers:** pain, discharge, redness, fever;
- **skin:** progression of thickening, itchy skin;
- **cardiopulmonary:** chest pain, palpitations, shortness of breath (after what level of effort), cough, loss of consciousness, leg swelling;
- **digestive:** reflux, heartburn, early satiety, nausea and vomiting, abdominal pain, bloating, diarrhea, constipation, fecal incontinence, weight loss;
- **renal:** high blood pressure at home, decreased urine output, headaches, blurred vision, confusion;
- **arthritis:** joint pain (where and when), morning stiffness (duration);
- **myositis:** difficulty climbing stairs or getting up from a chair without using your arms due to muscular fatigue.

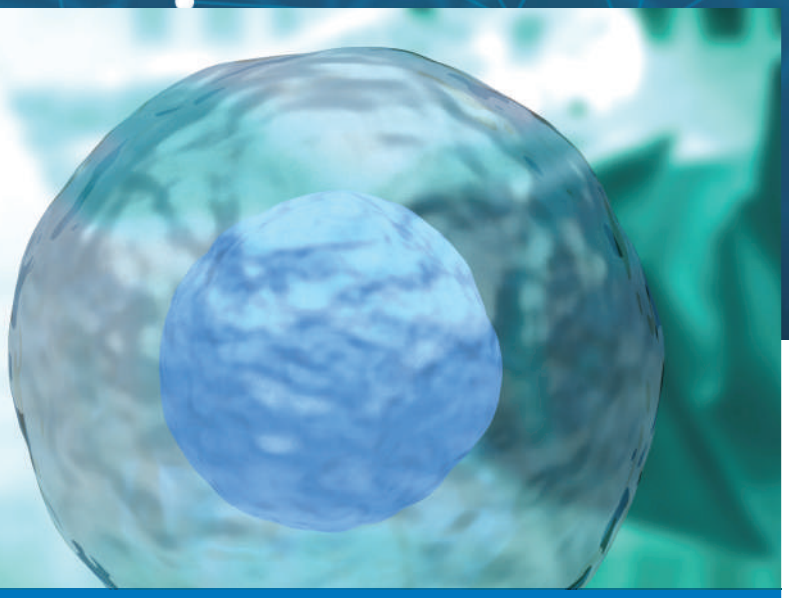
VACCINES

Vaccination against certain infections, such as influenza and pneumococcus, is recommended to prevent serious infection-related complications in the context of a chronic disease such as systemic sclerosis, particularly in patients with pulmonary involvement and in patients taking immunosuppressive medications. Make sure to bring your immunization record to your appointment so your doctor can determine if you need an update.

INVESTIGATIONS

Consultations and hospitalizations: if you have undergone additional examinations or consulted other doctors, or if you have been hospitalized since your last appointment, make a note of the details: dates, clinic or hospital center, reason for consultation, change in medication, etc. It is important that your doctor taking care of your systemic sclerosis is familiar with the general state of your health. If necessary, the doctor may ask for a copy of the investigation or hospitalization reports.

CELLULAR THERAPIES FOR SCLERODERMA



Dr. Marie Hudson, MD
Rheumatologist, epidemiologist, Associate Professor in the Department of Medicine at McGill University.

Introduction

Systemic sclerosis (SSc) is a chronic, systemic autoimmune disease characterized by a pathogenic triad of vasculopathy, immune dysregulation and fibrosis that results in multi-organ dysfunction affecting primarily the skin, gastrointestinal tract, lungs, heart and kidneys. Mortality rates for SSc compared to the general population have remained high (standardized mortality ratios above 3.5) in the past 50 years, in sharp contrast to the significant reductions in mortality in numerous other diseases, including cancer and cardiorespiratory diseases.

In addition to reduced survival, SSc is also notable for significant morbidity that results from Raynaud's phenomenon, finger ulcers, joint contractures, gastro-esophageal reflux disease, malabsorption, diarrhea, constipation, fecal incontinence, and exertional dyspnea, among others. These translate into limitations in physical mobility and function, disfigurement, pain, fatigue, sleep disturbance, and depression. SSc is associated with significant impairment in health-related quality of life on average 1½ standard deviations below the general population, comparable to or worse than that of patients with other chronic conditions, including heart disease, lung disease, hypertension, diabetes and depression.

SSc remains an orphan disease with high unmet needs in the field of therapeutics. The recommended treatments are mostly symptomatic, with drugs alleviating the symptoms mentioned above rather than targeting the disease as a whole. Also, immunosuppressive drugs such as cyclophosphamide and mycophenolate mofetil have at best modest effects aimed at stabilizing disease, without improving survival.

Similar to lung or kidney transplant where a whole organ is harvested from a healthy donor and transferred to someone with lung or kidney failure, cell therapies involve the harvesting of healthy human cells (whether from a donor or even the patients themselves) which are then transfused into a patient to restore or repair a diseased cell or organ. The best known type of cell therapy is blood transfusions. Some cells used as cell therapies have long-lasting effects and are therefore valued for their regenerative properties.

The purpose of this information brochure is to provide an overview of the cellular therapies presently available or currently under investigation for the treatment of SSc.

HEMATOPOIETIC STEM CELL (HSC) TRANSPLANTATION

OVERVIEW

Hematopoietic stem cells (HSCs) are special undifferentiated cells that reside in our bone marrow and give rise to mature cells that circulate in the blood, including red blood cells (which carry oxygen), platelets (which prevent excessive bleeding) and white blood cells (which protect us from infections). In certain blood cancers (e.g., leukemias), treatment is given to eradicate diseased HSCs and 'new - healthy' HSCs are transplanted. In scleroderma, some abnormal white blood cells are involved in the development of the disease by causing excessive inflammation and fibrosis. So as in leukemia, those diseased cells are removed and replaced with healthy HSC that regenerate a 'new – healthy' immune system. Many good studies have shown that HSC transplant can improve outcomes in scleroderma, including survival. However, HSC transplant is associated with considerable risks (e.g., acute infections, heart toxicity) and transplant-related mortality (5-10%).

WHO IS AND IS NOT A CANDIDATE FOR HSC TRANSPLANT FOR SCLERODERMA?

Because of the toxicity associated with the transplant itself, **HSC transplant is not for everyone.** Patients are carefully selected. The main selection criteria are the following:

MAIN INDICATIONS FOR HSC TRANSPLANT

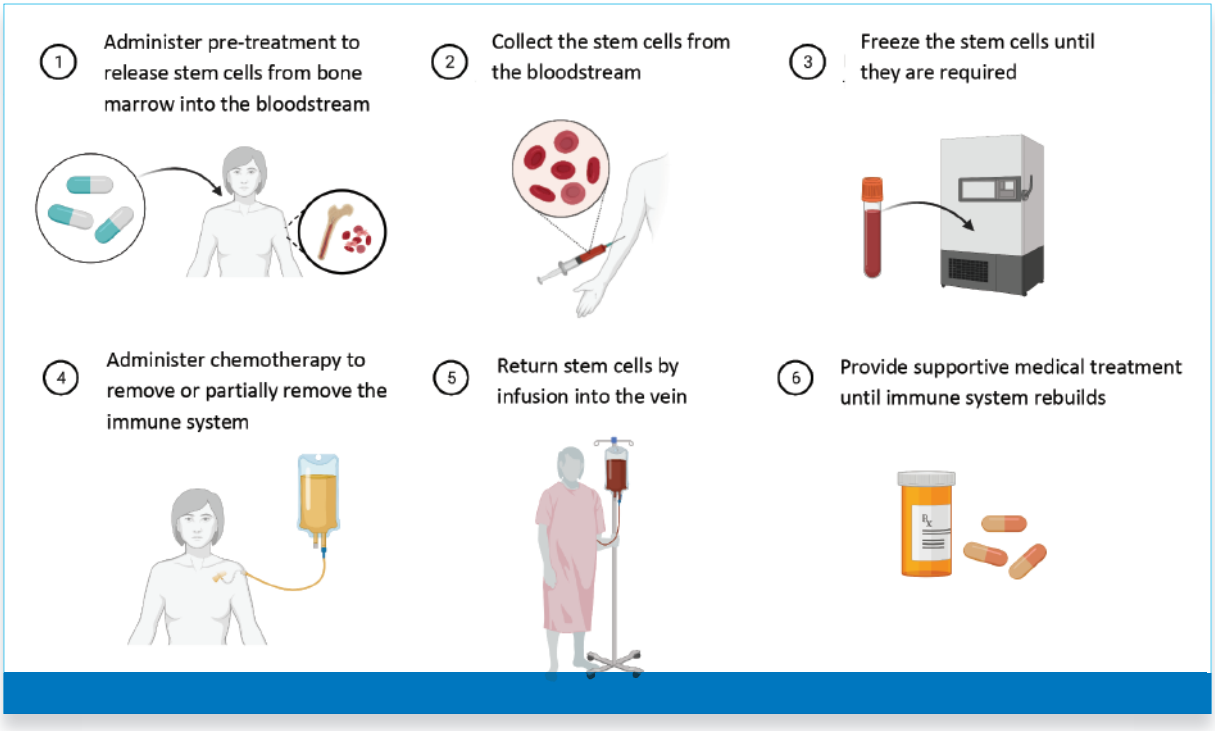
- Early disease (< 5 years)
- Rapidly progressive disease refractory to standard treatments
- Mild to moderate organ damage.

MAIN CONTRAINDICATIONS FOR HSC TRANSPLANT

- Longstanding disease
- Mild, slowly progressive disease
- Irreversible organ damage, including pulmonary arterial hypertension.

CELLULAR THERAPIES FOR SCLERODERMA

WHAT ARE THE STEPS FOR A HSC TRANSPLANT?



STEP 1: MOBILIZATION (A FEW DAYS)

First, you receive injections of a medication that allows your hematopoietic stem cells to be released from your bone marrow into your circulation.

STEP 2: STEM CELL COLLECTION (1 DAY)

The stem cells are then collected using an IV. The collection is sometimes manipulated to remove any remaining diseased cells and purify the stem cells.

STEP 3: STORAGE

The stem cells are stored in a cell therapy laboratory until they are needed.

STEP 4: PRE-TRANSPLANT TREATMENT (I.E., CONDITIONING, 5-10 DAYS)

High dose chemotherapy, and sometimes radiotherapy, is given to eliminate your unhealthy immune cells.

STEP 5: GETTING YOUR STEM CELLS BACK (I.E. HSC TRANSPLANT, AROUND 30 MINUTES)

Your own stem cells that were previously collected and stored are infused back into you through an IV.

STEP 6: RECOVERY

Close monitoring is required for weeks in the hospital and then months as an out-patient until your stem cells recover their normal function and regenerate a healthy immune system.

CELLULAR THERAPIES FOR SCLERODERMA

WHAT TO CONSIDER BEFORE A TRANSPLANT?

Studies have shown that the most common questions from scleroderma patients considering HSC transplant were the following:

1. **Will I be supported by a multidisciplinary team?**
2. **What are the financial risks?**
3. **Where can I get reliable information about HSC transplant?**
4. **Are there physical risks associated with HSC transplant?**
5. **What are the benefits of HSC transplant for someone in my condition?**

Here are some resources to help with answering some of these important questions:

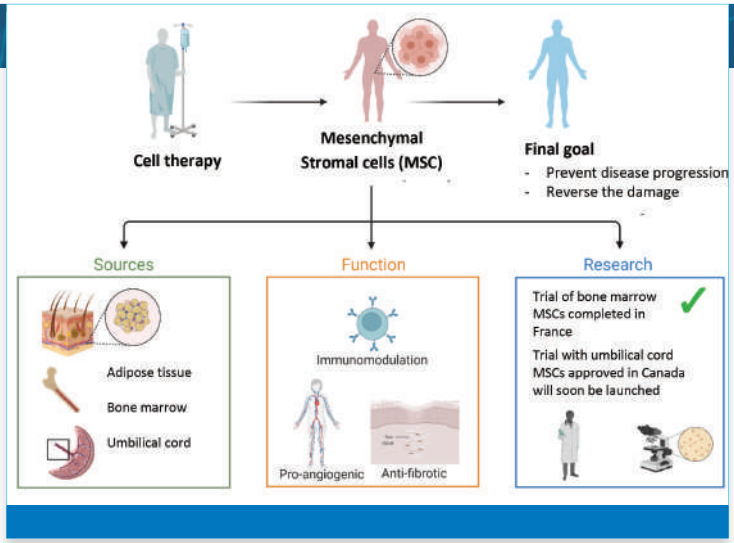
www.astemcelljourney.com

https://mathec.com

WHAT TO EXPECT AFTER TRANSPLANT?

HSC transplant is not a cure for scleroderma and the damage that has occurred prior to transplant is generally not reversible. However, HSC transplant is currently the best treatment to prevent disease progression and the only one shown to increase survival in scleroderma. Also, people with scleroderma have much better function and quality of life after HSC transplant compared to those treated with standard treatments.

MESENCHYMAL STROMAL CELLS (MSCS)



Because HSC transplant can be toxic and is not for everyone, different types of cellular therapies are being investigated. Mesenchymal stromal cells (MSCs) have immunomodulatory, pro-angiogenic, and anti-fibrotic properties and therefore have the potential to target all three axes of the SSc pathogenic triad. MSCs are present in nearly every tissue. They are responsible for keeping those tissues healthy. For therapeutic uses, they are usually harvested from bone marrow, adipose tissue, and umbilical cord. Injections of MSCs have been studied in a wide range of diseases, including other autoimmune diseases, and have been shown, first, to be safe (in large part because they do not need to be administered with chemotherapy) and second, to help in tissue repair and regeneration.

In scleroderma, MSCs are thought to work by re-setting the diseased immune system. This in turn can lead to less fibrosis and better circulation. To date, an early trial of bone marrow derived MSCs has been completed in France and results are promising. However, harvesting MSCs from the bone marrow is fairly invasive. Umbilical cords represent a promising alternative. A study of umbilical cord derived MSCs in scleroderma has been approved in Canada and should begin soon.

Many questions remain around the benefits of MSCs for scleroderma. What is the best source of MSCs? How can the function of MSCs from different donors be standardized? Will the benefits be sustained or will repeated infusions be required? If so, can MSCs from different sources and donors be used? In addition to infusions through the veins, can MSCs be injected locally, for example in the fingers, face or muscles? Can MSCs be produced in sufficient quantities to meet the demands? Instead of whole cells, are there specific components of MSCs or alternative cell products that could be more effective and more easily manufactured? And the list goes on!

CONCLUSION

Research is key to answering these and other questions. Identifying new, safe and accessible therapies that not only prevent disease progression but ideally reverse the damage that has already accumulated should remain the top priority for scleroderma research.

ADDITIONAL RESOURCES

Richard K. Burt, Dominique Farge, Milton A. Ruiz, Riccardo Saccardi, John A. Snowden (Editors). *Hematopoietic Stem Cell Transplantation and Cellular Therapies for Autoimmune Diseases*. CRC Press;
1st edition (November 15, 2021).
ISBN-13: 978-1138558557.
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PREGNANCY AND SYSTEMIC SCLERODERMA

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Internal Medicine resident, University of Montreal

WILL I BE ABLE TO HAVE CHILDREN?

Yes. Fertility in women with systemic scleroderma is generally comparable to that of women without the disease. Overall, the risk of miscarriage is also not increased compared to the general population.

On the other hand, pregnancy is not recommended in patients with the following conditions due to the high risk of complications:

- pulmonary arterial hypertension;
- progressive pulmonary fibrosis;
- kidney failure or severe high blood pressure;
- heart failure;
- a recent diagnosis of diffuse systemic scleroderma.

Pregnancy is also to be avoided in patients using medications that pose a risk to the babies (e.g., certain immunosuppressants). It is therefore important to use effective contraception and to discuss the desire for pregnancy with your doctor before becoming pregnant.

WHAT ARE THE RISKS ASSOCIATED WITH PREGNANCY IN THE CONTEXT OF SYSTEMIC SCLERODERMA?

If systemic scleroderma is well controlled and there is no cardiac, pulmonary, or renal involvement, there is a good chance of an uncomplicated pregnancy. Approximately 70-80% of women will have a successful pregnancy.

The risks associated with pregnancy in the context of scleroderma are

- premature birth (2-3 times more common);
- intrauterine growth retardation and/or low birth weight (3-4 times more common);
- renal failure, especially in patients with baseline renal failure;
- a difficulty in case of general anesthesia due to the limited opening of the mouth; local or epidural anesthesia is to be preferred.

A consultation with a gynecologist specializing in high-risk pregnancies is essential to ensure adequate follow-up in the context of scleroderma.

WHAT ARE THE POSSIBLE EFFECTS OF PREGNANCY ON SYSTEMIC SCLERODERMA?

In general, pregnancy does not appear to have any effect on the overall course of systemic scleroderma. However, pregnancy may worsen gastroesophageal reflux disease (GERD), with more heartburn and shortness of breath, especially if these problems were present before pregnancy. On the other hand, Raynaud's phenomenon may improve in about 30% of patients, due to increased body temperature and blood supply to the extremities in the context of pregnancy.

Rare cases of scleroderma renal crisis occurring in late pregnancy or after delivery have been reported in patients with a recent diagnosis of diffuse scleroderma (within the first 4 years). Pregnancy is therefore not recommended at this time in these patients but could be planned at a less progressive stage of the disease.

WILL SCLERODERMA AFFECT MY BABY?

The majority of babies are not affected by their mother's diagnosis of scleroderma. However, if the mother has anti-Ro or anti-La autoantibodies, these antibodies can cross the placenta and occasionally cause inflammation in the baby's heart, leading to heart block (heart rhythm disorder) in 1-2% of pregnancies. Serial fetal echocardiograms are then necessary during pregnancy to quickly detect a heart problem in the fetus. Also, the presence of anti-phospholipid autoantibodies in the mother is associated with an increased risk of miscarriage and preeclampsia. Ongoing monitoring of high-risk pregnancies is essential to ensure adequate assessment according to the level of risk.

WILL I BE ABLE TO BREASTFEED?

Yes, breastfeeding is possible and encouraged even in women with scleroderma. Some medications that can be passed into breast milk should be avoided. Scleroderma patients with active Raynaud's phenomenon in the post-pregnancy period may experience Raynaud's disease in the nipples, especially after breastfeeding or with any exposure to cold, which causes pain and discomfort. The use of heating pads to improve blood circulation before breastfeeding may be beneficial. A breast pump can also be used when Raynaud's is most active.

WHAT ABOUT MEN?

Few studies have looked at the fertility of men with Systemic Scleroderma. However, scleroderma can cause erectile dysfunction, possibly due to reduced blood flow to the penis. Some immunosuppressive drugs can also decrease fertility in men, most often reversibly.

IN SUMMARY

The majority of women with systemic scleroderma can become pregnant and have healthy children. Close collaboration between the patient, the rheumatologist and the gynecologist specializing in high-risk pregnancies is essential to minimize the risk of complications.

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**

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For this reason, we want to emphasize the generous gesture of our partner, Boehringer Ingelheim Canada Ltd which, thanks to an educational grant, made production, printing and distribution of this Spring Bulletin possible.

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.

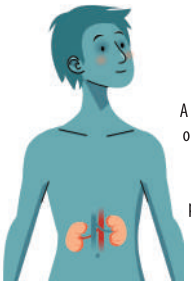
The symptoms of the disease may be visible, as is the case when the skin is affected, or the symptoms may be invisible, as when internal organs are affected.

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.

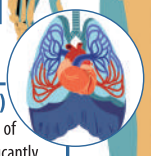
PULMONARY FIBROSIS
A potentially serious complication where normal lung tissue is gradually replaced by scarred fibrotic tissue, making it difficult to breathe and deliver needed oxygen to the body.

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.

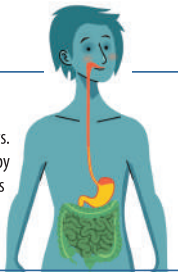
CALCINOSIS
Calcium deposits under the skin that may require antibiotics to cure occasional infections and sometimes surgery to drain calcium deposits and relieve pain.

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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Scleroderma Association B.C.