SCLERODERMA ASSOCIATION OF B.C.

# The Bulletin

Fall-Winter 2020-2021 | Volume 2 Number 2

Scleroderma Association B.C.

## Educational Sheets

# Summary




**Contact :**
**Scleroderma Association of B.C.**
PO Box 16155 Lynn Valley
North Vancouver BC V7J 3H2
Phone: 604-371-1005
Email: info@sclerodermabc.ca
**www.sclerodermabc.ca**




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**ISSN 2562-5780**


## Board of Directors

**Rosanne Queen**
**President**
604-984-9425
rq.sabc@telus.net

**Michele Gervais**
**Vice President**
604-761-7782
gordmich17@gmail.com

**Pat Thomasson**
**Member Liaison**
604-591-3033
pthomasson2001@shaw.ca

**Jeff Gammon**
**Adm. Director/Secretary**
604-809-6939
info@sclerodermabc.ca

**Patrick Livolski**
**Treasurer**
778-791-7834
treasurer@sclerodermabc.ca

**Tiasha Burch**
**Board Member**
778-984-3745
burchtiasha@gmail.com

**Valerie Doyon**
**Board Member**
250-202-9449
valerie.doyon@alumni.ubc.ca

**Jessica Jun**
**Board Member**
778-887-0523
jessjun@student.ubc.ca

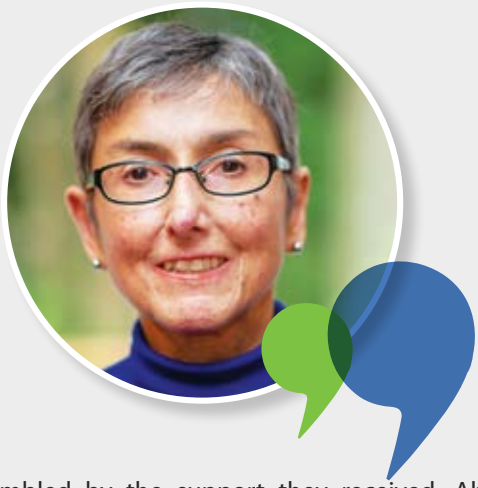
**Grace Kim**
**Board Member**
778-926-0118
ggkim@student.ubc.ca

**Beth Miller**
**Board Member**
604-815-8740
bethmiller@telus.net

**David Queen**
**Board Member**
604-984-9425
dq.sabc@telus.net

# COVID!!!

# It has been an interesting and challenging year to say the least!



**The global pandemic has turned our worlds upside down, mine included. The increased anxiety due to the isolation and concern of not ever having a normal life again has been a big challenge. Don't get me wrong, I love being home with my husband, but enough already! I'd like a bigger social bubble! And since this may well be the case for others, I decided for this report to talk about how we get through this together, COVID-19 plus the challenges of living with scleroderma.**

**Keeping informed.** With more time on our hands, there is ample opportunity to keep ourselves educated regarding our health. This issue of The Bulletin includes information sheets on topics related to scleroderma: how is systemic sclerosis diagnosed, how does it evolve over time, and what body systems are potentially affected. As we know, scleroderma is very individualistic and not every patient experiences the same challenges. Many more of these sheets are available for your reading pleasure on SABC's website.

**Keeping connected.** Despite COVID-19, SABC has continued to pursue our mandates of raising awareness, supporting research and providing education. As you can read in the articles in this issue, June was successful in raising awareness across the province as well as raising much needed research funds, and SABC's AGM and Conference went virtual this year.

June Awareness spread across the province with individuals "Moving for Scleroderma" and for some, stepping out of their comfort zone. When I asked both Donna Gervais and Pat Thomasson about their experience with participating in the first ever virtual fundraising event, I was not surprised at all by their responses. They, however, were surprised, pleased, and humbled by the support they received. Although the "Scleroderma Ride for Research" had its 9th year go virtual, it still managed to inspire generous donors despite this uncertain time. Each year I also continue to be humbled by the outpouring of support and caring both financially and emotionally. As Donna mentions, people do donate to causes that are close to their hearts. When they donate to research, they show that they do care about finding a cure. There are a lot of people around us that care.

I am also so incredibly grateful that "Zoom" is available to us and is not terribly technically challenging. It has allowed me to stay connected to family and friends while maintaining both our bubbles. It also comes in handy when I am reaching out to people I may not know just yet. Even a virtual face-to-face interaction helps those that may need that important personal link…whatever gets us through COVID-19.

With the limit to the size of gatherings and our members specific health concerns, the AGM and Conference organizers pivoted to a virtual event using "Zoom". Not only did it allow us to conduct our AGM & conference but allowed us to join in on conferences across Canada and the world.

In Dr. May Kazem's presentation, she stated studies have shown the survival rate of scleroderma patients has consistently increased over the years: in 1995, 61% of scleroderma patients survived 10 years past first diagnosis, 79% in 2002 and 82% in 2010. For a disease that presently has no cure, these numbers are extremely encouraging. There is much more hope, now based on science, than when I was first diagnosed.

In a time where I feel more confined and with winter coming, I'll have a continued need to connect with people and other scleroderma patients. Please reach out to those that provide you support and 'share your caring'. Together, we can get through this.

# Teressa's Story

**I would like to tell you all a little bit about the story of my Scleroderma journey and diagnosis. I was diagnosed 9 years ago after a few years of questioning and trying to figure out what was happening. In 2009 I was living my typical active life doing all the outdoor activities, sports and coaching that I loved to do. Then out of nowhere my symptoms started with just some unexplained muscle and joint pain, fingers and toes going white and being out of breath.**

After several years of going to specialist's appointments and trying to self-diagnose and heal myself with physiotherapy, chiropractors, acupuncture and much more, I started to think as if everything I was experiencing was all in my head. No one could physically see the pain I was in because it was all inside my body which was very difficult to explain. Finally, in 2011 after only a simple blood test I was diagnosed with scleroderma. This blood test proved that I had an overlap of syndromes which was characterized as a mixed connective tissue disorder.

Once diagnosed, I was referred to see a specialist in Vancouver, who specialized in scleroderma. My first visit with Dr. Dunne was a blur, I just couldn't wrap my head around the diagnosis. How did this happen? What did I do?

> **I remember travelling back to Valemount with my husband and we looked at one another and said, "What the heck is scleroderma??"**

I was so scared, wondering how long was my life span? What am I going to tell my daughter… my family?

This is my life with scleroderma. I live with GERD, Raynaud's syndrome, dermatomysositis, SLE, Sjogrens syndrome and rheumatoid arthritis. My daily challenges living with this disease vary from day to day. It is very frustrating, since I am a "control freak", not knowing what the day will look like. With each symptom there are medications to take. I used to hate taking medications and now I need to take them to manage symptoms and pain. But no matter how I feel, I put a smile on my face, go for a walk with my puppy, breathe in the fresh air and be thankful.

So, after several years of family, friends and co-workers asking the same what is scleroderma question, I had a friend, who inspired me 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, so we can find a cure for all of us fighting this rare disease.

Last fall I had the opportunity to participate in the SPIN program. What an amazing program!! This taught me how to be a support group leader, to create a positive environment, to be a great listener and to help and encourage people with all types of scleroderma. But most of all, to let you know that you are not alone and there will always be someone there for you. As of right now I am looking into setting up virtual group meetings in Northern BC. If you have any interest in joining, please email.

**Thank you for reading my story.**

# AGM and Conference Highlights

**On Saturday October 3rd, SABC conducted the very first virtual AGM & Conference via Zoom. Fifty-eight patients and their supporters registered for the event. The morning began with the Annual General Meeting, followed by presentations from two conference speakers: Dr. Alyson Wong and Dr. May Kazem. The conference concluded with an opportunity for attendees to 'socially meet' using Zoom's break-out rooms and talk about various topics of interest.**

## AGM HIGHLIGHTS

Included SABC President Rosanne Queen's Director's Report for 2019-2020 and the election of SABC's 2020-2021 Board of Directors.

## RAISING FUNDS FOR RESEARCH

Work on the Scleroderma Association of B.C. sponsored Research Project continues. SABC funds and co-leads a research study that began recruiting scleroderma patients with and without interstitial lung disease (ILD) in July 2017. 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 ILD. In 2019 a $71,009.10 cheque was presented to the St. Paul's Foundation for the SABC Research Project. A $43,000 cheque was virtually presented for 2020.

For a full update, visit SABC's website at sclerodermabc.ca/sabc-research-project/

## RAISING AWARENESS

For World Scleroderma Day, on June 29th, Canada Place, B.C. Place Stadium and Telus World of Science (all in Vancouver) were lit up in scleroderma "blue" to show our support for all those living with scleroderma around the world.


## CONFERENCE HIGHLIGHTS

Included **Dr. Alyson Wong's** presentation of interstitial lung disease in systemic sclerosis and **Dr. May Kazem's** presentation of what to expect with scleroderma over time.

**Dr. Alyson Wong** completed her respirology training at Dalhousie University, followed by an interstitial lung disease (ILD) fellowship and Master of Health Science at the University of British Columbia. She is a Clinical Instructor at UBC and physician in the St. Paul's Hospital ILD clinical and research programs.

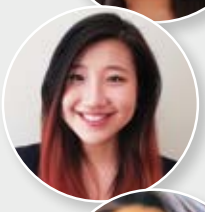
**Dr. May Kazem** completed her rheumatology training at Schulich School of Medicine at University of Western Ontario, after completing her Internal Medicine Specialty, MD, and MHA at UBC. Dr. Kazem's professional interests include connective tissue disorders such as Systemic Sclerosis and how they affect different body systems.

Video recording of the speaker presentations are available on SABC's website at sclerodermabc.ca

## WELCOME TO NEW BOARD MEMBERS

**VALERIE DOYON** is a second-year UBC medical student at the Island Medical Program, in Victoria.

**JESSICA JUN** is a first-year medical student at the University of British Columbia.



**GRACE KIM** is starting her path towards a career in healthcare as a first-year medical student at the University of British Columbia.

**BETH MILLER** is a teacher and has been living with Scleroderma since 2005.

# "2020 MOVE FOR SCLERODERMA" *Virtual*

## THE TEAMS ACROSS THE PROVINCE RAISED:

| | |
|---|---|
| **PEDALING FOR A CURE** - VANCOUVER | **$36,060** |
| **WALKING FOR A CURE** - ANYWHERE B.C., | **$4,296** |
| **VALEMOUNT MOUNTAIN WARRIORS** | **$2,660** |
| **VICTORIA "VIRTUAL" SCLERODERMA WALK** | **$2,470** |
| **WALKING FOR A CURE** - CAMPBELL RIVER | **$1,860** |

The campaign has raised **$47,546**

*Thank You*

**All of this money will be going to research in Canada. SABC continues to be the only financial supporter of the SABC Research Project being run locally through St. Paul's Foundation with a commitment this year of $43,000. The remaining funds will be available to research projects across Canada.**

## SUPPORT BECAUSE IT MEANS SOMETHING TO YOU.

That is my philosophy when donating to charitable organizations. When family and friends send me a link to support them in their fundraising campaigns for cancer, diabetes, heart and stroke, war amps, etc., it is my connection to their personal stories of courage and resilience in living with their challenges every day, that drives me to 'click' on their link to show them my admiration and that I care. The same goes for my annual 'walk for scleroderma' ask for donations for scleroderma research. I thank my family and friends (and this June, even those unknown to me) who clicked to help me virtually raise an astonishing **$1,445**! It is a great feeling when we all support and show we care for one another. **— Donna Gervais**

## I'VE NEVER FELT COMFORTABLE FUNDRAISING.

I have no problem donating but having to actually ask for money was extremely difficult for me. When my son was involved in sports and asked to sell chocolates, I would buy the case and gave them away to people I worked with. When I joined the SABC Board, I always declined to be involved with anything to do with fundraising. When it became time for fundraising this year, we knew it would be difficult due to COVID-19 so I decided to move out of my comfort zone and get involved. I posted a picture on Facebook of me "Moving for Scleroderma" with my young granddaughter with a note asking for donations for scleroderma research. I expected to get maybe $100 but was surprised when I raised close to a **$1,000**. I realize now that friends and family donate because they care for you. From now on I'll be more involved in raising funds every year. It's surprising how easy it is. **— Pat Thomasson**

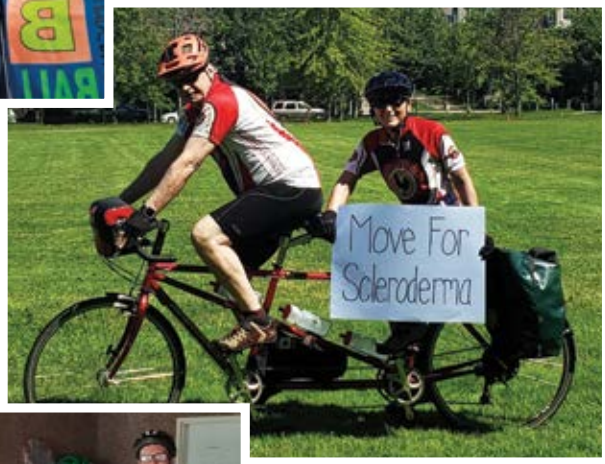
# THE SABC VIRTUAL WALK/RIDE

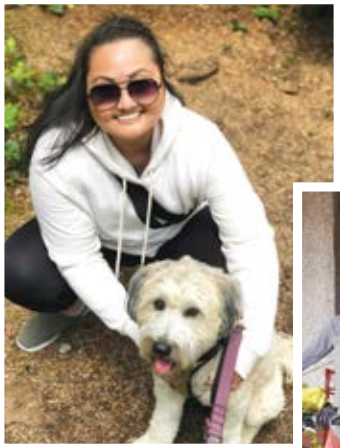
**has been a tremendous success, with participants from across the province.**

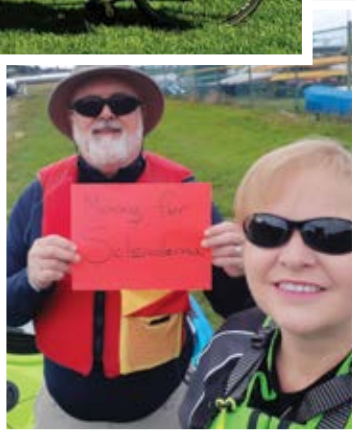

**Thank You**

**to everyone who participated in the first virtual "Move for Scleroderma".**

Thank you to all that donated, fund raised and got active. Check out the video on SABC's website, **sclerodermabc.ca**, capturing everyone "moving", showing how we all joined together in spreading awareness and raising funds to support scleroderma research!





# HOW TO DIAGNOSE SYSTEMIC SCLEROSIS

**Dr. Sabrina Hoa, MD MSC,**
Rheumatologist, clinical researcher
at the CHUM Research Centre

**The diagnosis of systemic sclerosis (SSc, systemic scleroderma) is usually based on the presence of a combination of symptoms and signs typical of systemic sclerosis:**

- Raynaud's phenomenon;
- skin thickening or puffy «sausage-like» swelling of the fingers;
- autoantibodies associated with systemic sclerosis in a blood sample;
- abnormalities in small blood vessels at the base of the nails;
- other skin and internal organ involvement associated with systemic sclerosis.

## RAYNAUD'S PHENOMENON

The most common and earliest problem observed in systemic sclerosis is Raynaud's phenomenon. This phenomenon is characterized by a change in color of the fingers (typically from a white discoloration to a blue then red color) and is most commonly provoked by exposure to cold. Raynaud's phenomenon can be "primary", meaning that it is isolated and not associated with a systemic autoimmune disease such as systemic sclerosis. However, when Raynaud's phenomenon occurs after the age of 40-50 years or is associated with ulcerations of the fingers (open lesions of the skin that heal very slowly) or other symptoms and signs of systemic sclerosis, a diagnosis of systemic sclerosis should be suspected and sought.

## THICKENING OF THE SKIN OF THE FINGERS

Thickening of the skin of the fingers, especially when it extends to the back of the hand, or puffy swelling of the fingers with a sausage-like appearance in the earlier stages, are characteristic signs of systemic sclerosis.

## AUTOANTIBODIES

Systemic sclerosis is an autoimmune disease in which the immune system becomes dysfunctional and turns against its own cells. Evidence of this autoimmunity can be found by the presence of autoantibodies in the blood, i.e., antibodies directed against oneself. Several autoantibodies specific to systemic sclerosis have been identified, including anti-centromere, anti-topoisomerase I (Scl 70) and anti-RNA polymerase III. Their presence, detected by a blood test, supports the diagnosis of systemic sclerosis when it is associated with Raynaud's phenomenon and other symptoms and signs of systemic sclerosis.

## NAILFOLD CAPILLARY ABNORMALITIES

Systemic sclerosis is also a disease of the small blood vessels. Abnormalities of these small vessels, or "capillaries", can be seen at the base of the nails. Specialized examination by high magnification microscopy of the capillaries in the nail bed, or "capillaroscopy", is often useful to support a diagnosis of systemic sclerosis.

## OTHER SKIN AND INTERNAL ORGAN INVOLVEMENT

Systemic sclerosis can also present with skin ulcerations on the fingertips, telangiectasias (dilations of small blood vessels that form red or sometimes purple spots on the surface of the skin) and calcinosis (small, white bumps of calcium deposits under the skin). Systemic sclerosis can also affect the digestive, pulmonary, cardiac and renal systems. In the presence of other symptoms and signs suggestive of systemic sclerosis, these additional manifestations support the diagnosis of systemic sclerosis.

## CLASSIFICATION CRITERIA

In scientific research, classification criteria are used to standardize the definition of systemic sclerosis (see Table below for the classification criteria issued jointly in 2013 by the American College of Rheumatology and the European League Against Rheumatism). Patients with a score of at least 9 points are classified as having systemic sclerosis. However, a diagnosis of systemic sclerosis (often at an early stage) can be made in a patient who does not meet the classification criteria.

## SUMMARY

In summary, the diagnosis of systemic sclerosis is based on a constellation of symptoms and signs typical of systemic sclerosis, particularly Raynaud's phenomenon and thickening of the skin of the fingers, as well as the presence of specific autoantibodies in blood samples and characteristic abnormalities on examination of the small blood vessels (capillaries) at the base of the nail.

**TABLE:**

**Classification criteria for systemic sclerosis, issued in 2013 by the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR)**

| Critères | Points |
|---|---|
| Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints | 9 |
| Puffy fingers ***or*** | 2 |
| Sclerodactyly (skin thickening of the fingers) | 4 |
| Digital tip ulcers ***or*** | 2 |
| Fingertip pitting scars | 3 |
| Telangiectasia | 2 |
| Abnormal nailfold capillaries | 2 |
| Pulmonary arterial hypertension ***or*** | 2 |
| Interstitial lung disease | 2 |
| Raynaud's phenomenon | 3 |
| Systemic sclerosis-related autoantibodies (anti-centromere, anti-topoisomerase I, anti-RNA polymerase III) | 3 |

# HOW DOES SYSTEMIC SCLEROSIS EVOLVE

**Dr. Sabrina Hoa, MD MSC,**
Rheumatologist, clinical researcher
at the CHUM Research Centre

**The evolution of systemic sclerosis is variable, depending on the extent of skin thickening (limited or diffuse systemic sclerosis), the presence of specific autoantibodies in the blood and the presence of internal organ involvement.**

## LIMITED OR DIFFUSE SKIN INVOLVEMENT

When skin thickening is limited to the hands, forearms, feet, legs below the knees, face and/or neck, this is referred to as **limited systemic sclerosis**. This form of systemic sclerosis is usually associated with a lower risk of developing severe internal organ involvement, except for pulmonary arterial hypertension, the risk of which increases after 5 to 10 years of disease.

When skin involvement extends above the elbows and knees, affecting the skin of the upper arms, thighs, trunk and/or abdomen, this is referred to as **diffuse systemic sclerosis**. In this form of systemic sclerosis, internal organ involvement is generally more common and extensive.

Limited or diffuse skin involvement does not usually change in the same patient. Thus, a patient with the limited form does not progress to the diffuse form of the disease. However, at disease onset, it may be difficult to be certain of the limited nature of the disease. A patient may initially have limited involvement of the hands (and be classified as having limited disease), but the involvement may rapidly progress to diffuse involvement over the next few months. The presence of swelling of the fingers and hands, and the presence of autoantibodies associated with the diffuse form of systemic sclerosis (e.g., anti-topoisomerase I/Scl-70 or anti-RNA polymerase III) usually point to a possible evolution toward the diffuse form of the disease.

In the diffuse form, skin thickening generally progresses in the first 2 to 5 years of the disease, then progression halts with a tendency towards spontaneous "softening" of the skin. The skin then becomes thinner and more fragile, but less "hard" than in the initial phase. In the limited form, skin involvement is restricted to the areas defining the limited form and does not progress further.

## SYSTEMIC SCLEROSIS-SPECIFIC AUTOANTIBODIES

Anti-centromere (or anti-CENP-B) autoantibodies are usually associated with the limited form of systemic sclerosis, a slower disease course at the onset of disease and less severe involvement of internal organs, but with more pulmonary arterial hypertension later in the course of the disease.

Anti-topoisomerase I (or anti-Scl-70) autoantibodies are usually associated with the diffuse form of systemic sclerosis, a more rapidly progressive disease course at the onset of disease, and an increased frequency of pulmonary fibrosis.

Anti-RNA polymerase III autoantibodies are usually associated with the diffuse form of systemic sclerosis, a more severe disease course and a higher risk of developing scleroderma renal crisis. In these patients, blood pressure should be closely monitored and corticosteroids should be avoided.

Anti-Th/To autoantibodies are associated with the limited form of systemic sclerosis, as well as an increased risk of pulmonary fibrosis and pulmonary hypertension. Anti-fibrillarin (or anti-U3-RNP) autoantibodies are associated with diffuse systemic sclerosis and an increased risk of pulmonary fibrosis. These autoantibodies are not available in all centres, but give a "nucleolar" pattern on the antinuclear antibody (ANA) test.

## INTERNAL ORGAN INVOLVEMENT

Unlike skin involvement which tends to improve over the years, internal organ involvement usually does not regress. In the case of pulmonary fibrosis, disease progression is highly variable: some patients will have a mild and relatively stable disease, some will have a disease that progresses slowly over the years, and some will progress rapidly. Patients whose thoracic CT scan shows involvement that extends beyond the lung bases are usually at higher risk of developing more progressive pulmonary fibrosis.

It is commonly said that organ involvement (skin, lungs, heart, kidneys, and others) occurs in the first 3 to 5 years of the disease, with the exception of pulmonary arterial hypertension, which occurs after 5 to 10 years of disease progression. However, more recent studies have questioned this notion. Pulmonary fibrosis frequently occurs early in the course of the disease, but can also appear later on. Inflammatory damage to the heart (myocarditis) usually occurs early in the disease, but fibrotic damage gradually progresses throughout the disease course. Digestive involvement also becomes progressively more important as the disease moves from the vascular and inflammatory phase to the more fibrotic phase of disease. The tissues of the digestive system then become weaker and unable to contract, leading to greater motility disorders and malabsorption over the course of the disease.

## IN SUMMARY

The course of systemic sclerosis is highly variable. An initial assessment of skin and internal organ involvement and the search for systemic sclerosis-specific autoantibodies in the blood can help predict the course of systemic sclerosis in an individual and inform the approach to screening for internal organ involvement.

# RAYNAUD'S PHENOMENON AND DIGITAL ULCERS

**Dr. Sabrina Hoa, MD MSC,**
Rheumatologist, clinical researcher
at the CHUM Research Centre

## WHAT IS RAYNAUD'S PHENOMENON?

**The most common and earliest problem observed in systemic sclerosis is Raynaud's phenomenon (RP). This phenomenon is due to a narrowing of the blood vessels in the fingers caused by exposure to cold and strong emotions, and is manifested by a change in colour of the fingers, which turn white, then blue and finally red.**

The white ("syncopal") phase represents a partial stoppage of blood flow and is characterized by a well-defined pallor of the affected fingers. It may also be associated with numbness in the fingers. This phase is always present in RP, unlike the other phases which may or may not be present. The blue ("cyanotic") phase is due to a lack of oxygen in the tissues. Finally, the red ("hyperemic") phase is due to the return of blood flow and may be accompanied by pain with a burning sensation. RP mainly affects the fingers, but can also affect the toes, nose or ears.

RP can be "primary", i.e., isolated and not associated with an underlying autoimmune disease. However, when it occurs after the age of 40 or is associated with finger ulcerations (open lesions in the skin that heal very slowly) or other symptoms and signs of systemic sclerosis, a diagnosis of underlying systemic sclerosis should be suspected and sought.

## DIGITAL ULCERS

**When RP is secondary to systemic sclerosis, RP is initially caused by a reversible hyperreactivity of the vessels to cold. Later, excessive proliferation of several cells causes a fixed narrowing of the opening of the vessels, such that circulation is reduced due to permanent structural changes. In this context, the loss of blood circulation can become severe to the point of causing ulcerations (open skin lesions) at the fingertips. These lesions can be slow to heal and can be complicated by local infection of the skin (cellulitis) or of the underlying bone (osteitis). In extreme cases, RP can be severe to the point of necrosis (death of part of the finger) or, rarely, self-amputation (loss of the fingertip).**

## WHAT CAN BE DONE TO AVOID AGGRAVATING RAYNAUD'S PHENOMENON?

In order to improve RP symptoms and prevent complications associated with RP, certain environmental factors that can aggravate RP should be avoided, including cold, stress, tobacco smoking, as well as medications and products with a vasoconstrictive effect (vessel narrowing). Here are some general measures suggested for patients with RP. **See page 15 of this bulletin for some general measures suggested to patients with RP.**

These simple measures may be sufficient to treat RP in the early stage (before structural damage has occurred). However, when there is structural damage to the vessels (fixed narrowing), these non-pharmacological measures become insufficient and the treating physician will often use medications to help dilate the vessels.

## MEDICATIONS TO TREAT RAYNAUD'S PHENOMENON

Most medications for the treatment of RP work by relaxing the blood vessels, allowing better blood flow to the extremities.

The first line of drug treatment for uncomplicated RP (without ulceration, necrosis or risk of self-amputation) is a calcium channel blocker (e.g., Nifedipine XL or Amlodipine). The dose is gradually increased as tolerated until symptoms improve. If the medication is not tolerated (dizziness, headache, swelling of the feet) or is not effective at the highest tolerated dose, the medication is switched to another calcium channel blocker.

If there is still no efficacy or if there is intolerance, a medication of another class, alone or sometimes in combination, such as a phosphodiesterase type 5 (PDE5) inhibitor (e.g., Sildenafil or Tadalafil) may be used. The addition of an angiotensin II receptor antagonist (e.g., Losartan) or a selective serotonin reuptake inhibitor (e.g., Fluoxetine) may also be considered.

If RP does not respond to these treatments or is severe with impairment of function and quality of life, a prostacyclin (e.g., Flolan) or its analogues can be given intravenously on an outpatient basis.

## WHAT IF THERE ARE DIGITAL ULCERS?

**When digital ulcers are present, aggressive treatment is required to accelerate healing and prevent necrosis or infection of the finger. The dose of the calcium channel blocker already prescribed is increased to the maximum tolerated dose, with the addition of a PDE5 inhibitor. The addition of low-dose aspirin (81 mg daily) may also be useful to promote better local circulation if there is no contraindication (e.g., gastrointestinal bleeding). A statin (e.g., Atorvastatin) may also be added for its potential protective effects on the blood vessels. Ulcers can be extremely painful and the use of narcotic drugs may be necessary.**

In more severe cases, daily intravenous treatment for five consecutive days with a prostacyclin (Flolan), followed by maintenance treatment every three weeks thereafter, may be given on an outpatient basis to accelerate ulcer healing.

Injecting an anesthetic product (lidocaine or bupivacaine) into the ulcerated fingers ("local chemical sympathectomy") may be effective in improving the blood flow locally in some cases, but with benefits that are usually temporary.

In extremely severe cases where the physician feels there is a risk of necrosis and self-amputation of fingers (fingers that remain permanently white or become purplish despite treatment), emergency treatment is required: the patient should be hospitalized and placed in a warm, quiet place for increased monitoring during treatment and further investigation (blood samples and assessment of vessel permeability in radiology after local injection of a dye, or "arteriography"). Treatment of ulcerations is sometimes maximized by the addition of an intravenous or subcutaneous anticoagulant (heparin), and Flolan and local chemical sympathectomy should be initiated promptly if necessary.

If ulcer episodes recur or are very severe despite the treatments listed above, Bosentan, an endothelin antagonist, may be added. This medication works on the blood vessels and can prevent the recurrence of digital ulcers.

Well-established digital ulcers can take up to several months to heal. They must be closely monitored by the doctor for the onset of infection or progression to necrosis. Signs that an infection may be present include pus draining from the ulcer, significant redness and swelling around the ulcer and on the affected finger, or rapid progression of the ulcer. There may also be fever and chills. The doctor will then check for germs (bacterial culture) and, depending on the severity, prescribe antibiotics by mouth or intravenously.

## WHAT CAN BE DONE TO AVOID AGGRAVATING RAYNAUD'S PHENOMENON?

In order to improve RP symptoms and prevent complications associated with RP, certain environmental factors that can aggravate RP should be avoided, including cold, stress, tobacco smoking, as well as medications and products with a vasoconstrictive effect (vessel narrowing).
Here are some general measures suggested for patients with RP.

| AGGRAVATING FACTOR | RECOMMENDATIONS |
|---|---|
| **COLD** | ▶ Keep the whole body warm (e.g., wear thermal underwear, heat-retaining hat). |
| | ▶ Keep fingers warm (e.g., warm mittens or electric hand warmers). |
| | ▶ Avoid sudden exposure to cold. |
| | ▶ Avoid abrupt changes in temperature (e.g., from a heated room to an air-conditioned room), cold breezes and cold humid air. |
| | ▶ Quickly apply methods to end an RP attack: place hands in a warm place (e.g., warm water, under the armpits) or move your arms in rotation to promote circulation in the extremities. |
| **STRESS** | Reduce stress. |
| **TOBACCO** | Avoid smoking and/or being exposed to cigarette smoke (quitting smoking can significantly improve RP). |
| **MEDICATIONS AND PRODUCTS WITH VASOCONSTRICTIVE EFFECTS** | **Avoid or consult your pharmacist before using:** |
| | ▶ Nasal decongestants |
| | ▶ Amphetamines |
| | ▶ Some antihypertensive medications (e.g., clonidine, beta-blockers) |
| | ▶ Medications to treat attention deficit hyperactivity disorder, ADHD (e.g., methylphenidate or dextroamphetamine) |
| | ▶ Anti-migraine medications (e.g., ergotamine) |
| | ▶ Weight loss pills |
| | ▶ Natural products containing ephedra |
| **CAFFEINATED BEVERAGES** | Reduce consumption of caffeine-containing beverages (e.g., coffee, tea, colas). |
| **SYMPATHOMIMETIC RECREATIONAL DRUGS** | Avoid cocaine and amphetamines (e.g., speed). |

# PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS

**Dr. Sabrina Hoa, MD MSC,**
Rheumatologist, clinical researcher at the CHUM Research Centre

**Dr. Tamara Grodzicky, MD FRCPC**
Rheumatologist, clinical researcher at the CHUM Research Centre

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

## UNDERSTANDING PAH IN SYSTEMIC SCLEROSIS

PAH in systemic sclerosis is due to an exaggerated and progressive narrowing of the small blood vessels in the lungs. This is caused, on the one hand, by the increased presence of molecules (chemical signals) that promote the contraction and obliteration of the pulmonary arteries, and on the other hand, by a relatively insufficient quantity of molecules that promote their dilation. The factor that triggers this imbalance is unknown.

When the blood vessels in the lungs are narrowed, it is harder for the blood to circulate and get oxygenated. As a result, the level of oxygen in the blood becomes reduced, leading to suboptimal oxygenation of the body's organs and tissues.

The danger of having persistently very high pressure in the arteries of the lungs is also due to the fact that it makes it harder for the right side of the heart to pump blood through the lungs. Over time (several years), this eventually causes the right side of the heart to fail (right-side heart failure).

## WHO IS AT RISK OF DEVELOPING PAH?

**PAH occurs in 10 to 15% of patients with systemic sclerosis. PAH is most often associated with the limited form of systemic sclerosis and with more than 5 years of disease. The presence of certain autoantibodies, including anti-centromere, anti-U1-RNP, anti-Th-To and anti-U3-RNP (fibrillarin), are also risk factors for PAH.**

## WHAT ARE THE SYMPTOMS OF PAH?

PAH is often silent at first, but over time it can cause a variety of symptoms: shortness of breath and fatigue during physical exertion, chest pain, impending loss of consciousness or even unconsciousness (syncope) in more advanced cases.

Physical examination by the doctor is often not very revealing in the early stages, but it will show signs of heart dysfunction in more severely affected people. In these latter cases, the examination will show, for example, abnormal auscultation of the heart with the stethoscope and, in the event of heart failure, abnormal distension of the neck veins and swelling (edema) of the feet and legs.

## HOW TO SCREEN FOR PAH?

Given the absence of specific symptoms at the onset of PAH, rheumatologists routinely screen all systemic sclerosis patients using pulmonary function tests (PFTs) and echocardiograms. A blood test for NT-proBNP, a specific marker for the heart, can also be used for screening. These tests are done annually in patients at higher risk of developing PAH, such as patients with the limited form of systemic sclerosis and long disease duration, or those with autoantibodies specified earlier.

When PAH is suspected, a more invasive type of investigation, i.e., catheterization of the right (and often also the left) side of the heart, is necessary to confirm the diagnosis. This is performed by a cardiologist. The pressure in the pulmonary arteries is then measured directly using a catheter inserted through a vein in the crease of the elbow or the groin.

Other tests may also be performed at this time to rule out other potential causes of pulmonary hypertension, such as heart disease, small clots in the lungs, lung fibrosis or emphysema, or sleep apnea.

## TREATMENT FOR PAH IN SYSTEMIC SCLEROSIS

Given the complexity of the diagnosis, initial assessment, administration of certain medications and follow-up, patients suspected of having PAH are referred to specialized centres for management, with concomitant follow-up by the treating rheumatologist.

Indications for starting treatment for PAH include confirmation of the diagnosis by cardiac catheterization and the presence of symptoms (shortness of breath on physical exertion) with moderate to severe functional impairment. Treatments for PAH act by dilating vessels that are too narrowed, thus reducing the high pressure in the pulmonary arteries. Medications for PAH work through different mechanisms:

- Endothelin-1 receptor antagonists: bosentan (Tracleer®), ambrisentan (Volibris®) and macitentan (Opsumit®);
- Phosphodiesterase-5 inhibitors: sildenafil (Revatio®) and tadalafil (Adcirca®); Soluble guanylate cyclase stimulator: riociguat (Adempas®);
- Prostacyclins: epoprostenol (Flolan®), treprostinil (Remodulin®) and selexipag (Uptravi®).

In patients with moderate functional impairment, medications from the first two categories above are used alone or in combination. These medications are given by mouth and require periodic blood tests to monitor for side effects. If the disease is progressive or severe with symptoms at the slightest exertion, then inhaled, subcutaneous or intravenous prostacyclins may be added.

Concomitant treatment with medications such as diuretics (e.g., furosemide/Lasix®) and inotropic agents (improve the contractility of the heart muscle) are also useful for treating heart failure. Home oxygen therapy is reserved for patients with very severe disease. As a last resort, there is the option of lung or heart-lung transplantation, after a detailed medical and multidisciplinary assessment.

The follow-up of patients with PAH is done through a medical questionnaire, physical examination, and periodic investigations: blood samples, echocardiogram, pulmonary function test and in some cases, a repeat cardiac catheterization. The 6-minute walking distance can also be used to evaluate the effectiveness of treatment, typically associated with decreased shortness of breath and improved tolerance to physical exertion, which translates to the ability to walk further in 6 minutes.

## IN SUMMARY

PAH is a serious complication of systemic sclerosis. However, over the past two decades, several new medications have been studied and approved for the treatment of PAH and can improve the quality of life and life expectancy of systemic sclerosis patients with PAH. Because PAH is often a silent disease in its early stages, it is essential to screen at-risk patients in order to make an early diagnosis and begin treatment if indicated.

# 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**

### Campbell River

**Jackie Alexander**
250-830-7287
rjsjalexander@telus.net

### Chilliwack

**Kelly Grant**
604-378-1806
thekellygrant@gmail.com

### Creston

**Betty Kuny**
250-428-8875
rkuny@telus.net

### Kamloops

**Jen Beckett**
250-574-3151
jenniferbecketts@hotmail.com

### Kelowna

**Angie Reglin**
250-860-5700
angiereglin@gmail.com

### Nanaimo

**Linda Allen**
llallen.52.14@gmail.com

### Penticton

**Barb Creighton**
250-770-7836
mischief2@shaw.ca

### Quesnel

**Leah McAnena**
250-925-0281
almcanena@hotmail.com

### Squamish

**Beth Miller**
604-815-8740
SABCBeth@gmail.com

### Surrey

**Pat Thomasson**
604-591-3033
pthomasson2001@shaw.ca

### Valemount area

**Teressa Colosimo**
250-566-3165
pattess72@hotmail.ca

### Vernon

**Lisa VanDyk**
250-542-5231
SanNicolaswest@shaw.ca

### Victoria

**Susan Goss**
250-479-8586
susangoss@shaw.ca

### Williams Lake

**Cecelia Jaeger**
250-392-3656
cecejaeger@gmail.com

### Yellowknife

**Helen White**
867-873-5785
hwhite@theedge.ca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