SCLERODERMA ASSOCIATION OF B.C.

The Bulletin

SPRING-SUMMER 2024 | VOLUME 6 NUMBER 1

Scleroderma Association B.C.

SABC's 40th Anniversary

Calcinosis in Systemic Sclerosis

Sjögren's Syndrome associated with Systemic Scleroderma

Summary

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A Word from Our President

Spring has sprung and that means our June awareness campaign has begun!

Our **"Moving to Cure Scleroderma"** events were so successful last year that I'm looking forward to once again raising awareness and money for research.

Kenny Reid and his band will host their 1st dance and David and I will host our 13th **"Scleroderma Ride/Walk for Research".** Thank you to all for organizing and to the volunteers that will make these events a success.

We do have a large province so for those who can't attend these two events, I encourage you to join by hosting your own "virtual" walk, run, picnic or any other activity that you love to do. It really is easy to do. Get together with family and friends and have fun! Take photos, even make up a sign "Moving to Cure Scleroderma" and send us your photos to share. If you want to make a donation and or fundraise, go to our website and either support an existing team or start your own "virtual" team.

We are all in this together and together we can make a difference!

SABC's Board and volunteers have been steadily working to stay connected with our members and to expand our existing ways to share information. I encourage you to check out our virtual support group meetings. It's a great opportunity to share ideas, learn tips and trick from others on how to live with scleroderma. We know it's a complicated disease, but these zoom meetings are a reminder we are not alone in this journey.

We continue to also host "virtual" educational seminars so please continue to check our website for upcoming presentations under the "events" tab.

Finding moments of personal relaxation amidst our hectic schedules can be quite challenging. Therefore, I want to thank **Teressa Colosimo** *for offering both Easy Flow and Chair Yoga to our members*.

I would also like to thank **Sylvia Reimer** for sharing her incredible story. When I read these stories, I am never surprised how resilient "scleroderma warriors" continue to be.

In this edition you'll find educational resources on both Calcinosis and Sjögren's. As I cope with the symptoms of Sjörgens daily, I appreciate the tips and am reassured the information is from a reliable resource.

As we celebrate our 40th Anniversary this year, I hope you will join us at our in-person AGM & Conference on Saturday, October 26th at the Alan Emmott Centre in Burnaby. I am proud to be part of this volunteer organization that has not only grown but, has also remained steadfast in supporting its members throughout the years. Your presence will mean a great deal to us as we commemorate this milestone together.

Moving to Cure

SCLERODERMA

WALK - RIDE - GROOVE

2 EVENTS - JUNE 2024

June 1st, across Canada, starts Scleroderma Awareness Month which involves raising awareness of scleroderma and much needed funds for research.
The yearly campaign ends on June 29th, World Scleroderma Day.

With the participation of you: B.C.'s patients, families, and friends, we are once again excited to come together in a sea-of-blue and support each other as we live with this rare disease. Our ride/walk and new this year, a music concert in Ladner/Delta provide us the opportunity to connect over a shared cause; showcasing our enduring warrior spirit while raising funds to support research.

It's time again for Moving to Cure Scleroderma… please join us this June for all the fun!

For more details on the schedule of the events and to register, please visit

sclerodermabc.ca

*The Scleroderma Association of B.C. will accept donations before, during or after the month of June. All donors receive a receipt for the full amount of donations. This fundraising campaign is part of the Moving to Cure Scleroderma occurring each June.

A big Thank-You to the event coordinators who welcome you to join them and support us all this June. We are excited to see everyone in Vancouver, Ladner/Delta and Anywhere in B.C.!

With the support of you, our families and community, we can all make this June's events truly special. If you're interested in fundraising in your community this June, please contact the SABC.

Not able to attend any of the June events? 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.**

JUNE IS SCLERODERMA AWARENESS MONTH!

LADNER/DELTA

SATURDAY, JUNE 8TH / GROOVE FOR SCLERODERMA

LADNER COMMUNITY CENTER
– Ladner/Delta, V4K 3R8

80's cover band 17 West will be playing an evening of songs for you to Sing, Groove and Boogie the night away. Concert includes a cash bar with Beer (alcoholic or non), Wine and soft drinks for sale. Silent auction prizes and 50/50 draw, so please bring as much as you can spare to spend for a great cause. All profits go directly to scleroderma research and our Moving to Cure Scleroderma awareness campaign.

Doors open at 7 PM Come on by, as it is guaranteed to be a great night!

KENNY REID, LADNER/DELTA

VANCOUVER

SUNDAY JUNE 16TH / BIKE RIDE OR WALK

STANLEY PARK
– Vancouver, V6G 1Z4

As we approach our 13th "Scleroderma Ride/Walk for Research" this year, it's truly touching to see the support we've garnered. Whether you're biking up to UBC, walking or biking around the seawall or simply joining us for our famous potluck lunch, your presence means everything. We're driven by a shared passion to raise awareness and funds for the SABC Research Project, and your support deeply touches us all. We can't wait to see you there, knowing that together, we're making a meaningful impact.

Registration starts 9:00 AM, the fun starts 9:45 AM rain or shine!

ROSANNE AND DAVID QUEEN, NORTH VANCOUVER

ANYWHERE IN B.C.

ALL OF JUNE / VIRTUAL MOVING TO CURE SCLERODERMA

What is a virtual event? Engage in an activity of your choice, in a location of your choice! You can walk, bike, run or run a marathon, do yoga, kayak or any activity you enjoy. It's your choice so be as energetic, adventurous or creative as you can be!!

Let's get our family, friends, co-workers and even your community involved in spreading awareness and most of all having fun. No matter where you live, pick a day in June, and show us your favorite activity. I encourage you to go to Scleroderma Association of B.C.'s website and read "Our Stories". I am so much stronger, having these inspirational Scleroderma Warrior's in my life. Thank you for your continued support, raising awareness and donating to find a cure for this rare disease.

And **please DON'T FORGET to email your "MOVING TO CURE SCLERODERMA" blue t-shirt photos to info@sclerodermabc.ca.**

TERESSA COLOSIMO, VALEMOUNT

The Scleroderma Association of B.C. marks its 40-year anniversary in 2024!

Scleroderma Association B.C.

EST. 1984

This year SABC celebrates 40 years of supporting patients, promoting greater awareness of Scleroderma and supporting research toward finding a cure.

In 1984, five women diagnosed with Scleroderma came together with the help of a shared doctor to launch a support program for Scleroderma patients in B.C. With just $17 from their purses, they undertook the formation of an all-volunteer registered charity which became the Scleroderma Association of B.C. (SABC), with a three-fold mission:

- to promote patient outreach, support, and education.
- to raise Scleroderma awareness and understanding.
- to encourage and support Scleroderma research in B.C. and Canada.

Forty years later, we are delighted to celebrate the impact of our original founders on our community and the proceeding volunteers and supporters who continue to work in achieving the above objectives. We are honoured to carry forth the legacy of visionary leadership in service to those impacted by Scleroderma with the shared goal that others will have an easier journey.

REFERENCES HISTORY TABLE - PAGE 7

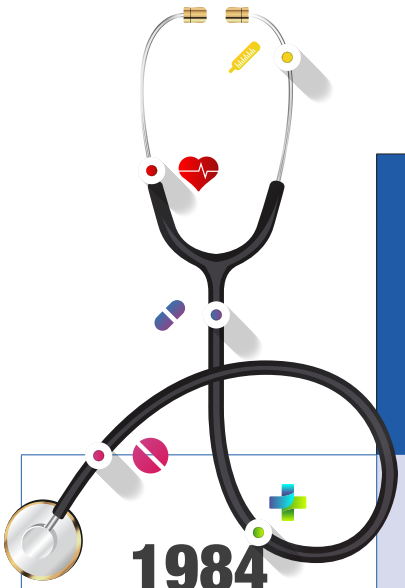
[1]The BC Scleroderma Clinic, a collaboration between SABC and Research Centre at St. Paul's Hospital, was the sole clinic of its kind in Western Canada, offering specialized care for patients across the province. This initiative created the potential to greatly improve the quality of life for scleroderma patients in the region, addressing a significant healthcare need. It is now a combined clinic with rheumatologists and respirologists for patients diagnosed with scleroderma and interstitial lung disease.

[2]SABC member Nobby Breen, a painter, and his employer convinced the Worker's Compensation Board that scleroderma was a work-related, occupational hazard, resulting in the WCB accepting his claim and providing full compensation benefits.

[3]The Health Sciences Fair evolved into a society of volunteers called PIE, Patients in Education. The purpose of PIE was to provide opportunities for patients and community organizations to have a voice in the education of health professionals. SABC made a small annual donation to PIE and had representatives on the organizing committee.

[4]Over 25 Information Sheets have been produced and placed on the website providing explanations, beneficial insights and current, credible information on the various aspects and symptoms of systemic sclerosis or scleroderma.

From SABC member Kelly Mauro, *"It's impossible for me to put it into a few sentences. I can only simplify it with, if it hadn't been for Nobby Breen coming forward to Joan Kelly with his story, and then Joan having the amazing ability and foresight to communicate and understand how much sharing his story would impact not only my husband Rino and our family, but many more over the years, encouraging us to come together to help others having their claims accepted. I can't express how much the Sclerodata meant to so many patients and families. Patient outreach is at the core of what makes support groups and networking a success! I will be forever grateful and appreciative to the SABC".*



History of SABC

	Supporting Patients **Patient Outreach & Education: A person living with scleroderma no longer needs to feel alone**	Awareness **Raise scleroderma awareness and develop information tools for the general public and the medical community**	Research **Encourage and support, through fundraising, leading edge research in BC and Canada. To date, SABC members have raised over $1.5M in support of research!**
1984 **member count: 27**	Members connected over coffee or the phone, sharing and learning. The semi-annual ***Sclerodata*** newsletter provided updates and tips, while annual workshops, in collaboration with the Arthritis Society, served as conferences for the community.	Informational brochures and pamphlets detailing the impact of scleroderma were created for distribution at events.	Beginning as a grassroots effort, funds for scleroderma research were raised through tea parties, dances, casino days, raffle tickets, and entertainment book sales.
1990 **member count: 270**	Member gatherings expanded with the inclusion of Community Representatives, individuals with scleroderma volunteering to provide support and guidance in their local communities.	Participated in UBC's Health Sciences Fair[3] which introduced medical students to patient members, providing valuable hands-on experience with a rare disease, leading to earlier awareness.	SABC backed Dr. Connie Wong's tight-skin mouse scleroderma study at UBC with over $90,000. BC patients joined the Canadian Scleroderma Research Group (CSRG) registry.
2000 **member count: 400**	The Scleroderma Clinic[1] is established. ***Sclerodata*** celebrates a member's success in obtaining WCB recognition and acceptance of their claim[2]. Annual local AGMs and conferences are held in the Lower Mainland.	SABC and a Calgary-based support group joined forces to establish a national network of provincial support organizations, now recognized as Scleroderma Canada. The Gurmej Kaur Dhanda Memorial Scholarship Award, was presented to medical students showing dedication to scleroderma research.	Dhar Dhanda & family's annual gala supported the SABC Research Project and other scleroderma research in BC. The Scleroderma Skate events also raised funds for research. The SABC Research Program at St. Paul's Hospital intensified its study of scleroderma patients' blood and skin.
2010 **member count: 800**	SABC and Scleroderma Canada co-hosted a joint local/national conference in Burnaby, bringing together doctors, patients, and families nationwide. Members represented BC at various national and international scleroderma conferences.	Annual province-wide walks and bike rides in June raise awareness and research funds. ***The Bulletin***, funded by pharmaceutical grants and created with *Sclérodermie Québec*, contains updates on scleroderma symptoms, research, and SABC events. It is also mailed to doctor and rheumatologist offices.	With increased funds, SABC expanded support for national research initiatives at SPIN, the Scleroderma Research Chair at CHUM, and CSRG. Additionally, funds were allocated for acquiring an ultra-sound machine and Doppler ultra-sound for the Scleroderma clinic.
2020 **member count: 1000**	***Come Zoom With Us*** virtual support group meetings enable everyone in BC to attend. Educational Webinars are also hosted online.	SABC's website[4] is continually updated with the latest sclero-derma information, offering accurate resources for patients and boosting public awareness. Additionally, SABC establishes a social media presence on Meta, LinkedIn, and Instagram platforms. The **Lihong Yang Memorial Scholarship Award** supports two medical students dedicated to advancing their studies in fields related to scleroderma	The **Joan Kelly Memorial Research Fund**, established by generous donors, creates an endowment for sustained financial backing of scleroderma research. Capital is invested, with annual dividends dedicated to funding research in BC and Canada.

Community Contact Representatives
What the SABC has Meant to Me

Kelly Grant, Chilliwack - ***I was diagnosed in 2004; I've had scleroderma for 19 years.*** *Soon after I was diagnosed I called SABC and Joan answered. Joan reassured me and sent me brochures and information. Since then I have attended the AGMs and conferences where I met and connected with other people with Scleroderma. Three years ago I became a Community Rep and completed the SPIN/SSLED training. I have been facilitating Come Zoom With Us and Educational Webinars where I've met many scleroderma warriors. I'm grateful for SABC's resolution in finding a cure and improving the lives of people affected by scleroderma.*

Jackie Alexander, Campbell River - ***I was diagnosed in 2001 but had symptoms for a couple of years before my diagnosis****. Through SABC I have been able to access resources, information and make connections with some other wonderful people that also live with Scleroderma.*

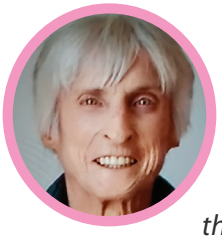
Susan Goss, Victoria - ***I have had Scleroderma for 24 years.*** The SABC connected me with other people that understood exactly what I was going through. Initially when you are scared and feel all alone this is a huge thing. They continue to offer support and useful resources all these year later and it is wonderful that I can repay all the support I received to others.

Angie Reglin, Kelowna - ***I was diagnosed in 2004; 19 years****. I reached out to SABC in about 2005 and spoke with Joan who put me in touch with the Kelowna rep. I was scared and did not want to know what was coming down the pipe, so I travelled this journey alone. In about 2016 I reached out to SABC again by attending a AGM conference, where I realized I could be helped and be helpful by sharing my journey. I became a rep for Kelowna shortly thereafter.*

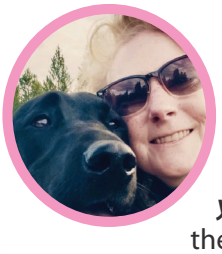
Linda Allen, Nanaimo - ***I have had scleroderma for 8 years****. I have been very appreciative of the SABC connection to others with this disease, the information provided through the Zoom webinars, The Bulletin magazine and the newsletter. The more information we can get about this strange and evolving disease, the better. The research being done is encouraging. Thank you SABC for all you do to support us.*

Suzanne Gavin, Vancouver - ***I was diagnosed with Scleroderma in 2012; 12 years****. I have found SABC to be a friendly supportive group. Over the last 2 years I have had the pleasure of supporting Kelly Grant with facilitation of Support Group Meetings. I have found the meetings to be a wonderful way to meet other people, listening to their journey and challenges with scleroderma.*

Community Contact Representatives
What the SABC has Meant to Me

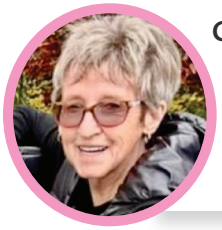


Betty Kuny, Creston - ***I was diagnosed in 1964 when I was 19 years old; I've had scleroderma for 60 years.*** *Through the educational, supportive and informative SABC, I have come to adapt my scleroderma life in coping and doing what I can to live my life as best I can.*



Teressa Colosimo, Valemont & Northern Rural Communities - ***I was diagnosed in 2011; 13 years ago.*** SABC has been there throughout my diagnosis and has given me the opportunity to help others through their journey with Scleroderma. Most of all SABC is an amazing and supportive community. I have met some of my closest friends through SABC.

Jennifer Beckett, Kamloops - ***I was diagnosed in the winter of 2015; over 8 years ago.*** As an area representative with SABC, I have had the opportunity to make some lasting friendships and amazing connections with other area representatives across the province. Along with existing members of SABC it's been a pleasure and privilege to meet some of the original founders of this organization. I am grateful for all the wonderful resources SABC shares.



Cecelia Jaeger, Williams Lake - ***I was diagnosed in 1994.*** By always being present for reassurance that I was not alone ... plus valuable information.

Beth Miller, Squamish - ***Since 2005, so 19 years.*** Providing information, both online through the website, and in person through conferences. Also providing a sense of connection and community, through "Come Zoom With Us" support groups and educational webinars. Also providing opportunities to participate in SPIN programs and other research.

Sylvia Reimer, Nelson - ***I was diagnosed with scleroderma in 2000; 24 years ago.*** Early on I became a member of the Scleroderma Association of BC and was appreciative of the newsletters and the information. Soon after becoming a member I became SABC area rep for the Nelson area. In 2023, after Rosanne reached out to me, I completed the SPIN/SSLD course. I'm now helping out with co-facilitating some of the SABC monthly virtual support group meetings, getting to know the other dedicated people involved and connecting to the broader Scleroderma community.

Helen White, Yellowknife, NWT - ***24 years.*** SABC has supported me through giving information, socialization, and providing contacts.

Testimony
Sylvia's Story

> I had no idea how Raynaud's would become a crystal ball into my future.

Raynaud's phenomenon was not new terminology for me as I had been having intermittent attacks since I was a teenager, but during my mid thirties the severity of attacks steadily increased involving both my hands and feet.

I had always been very fit, having taught fitness classes after work since my early twenties. So the random joint swelling, pain, the incredible fatigue, skin tightening, rashes on one side of my body for a few days at a time and itchy fingers raised significant red flags for me. My feet became so painful I had difficulty walking let alone teaching step/weight classes. Along with verbal cues, I always snapped my fingers to let my fitness participants know a change was upcoming. All of a sudden I was having difficulty with the 'snap'. It didn't take much longer for the day to arrive when I could no longer snap my fingers. The skin on my hands had tightened too much and this was the beginning of my fingers being forced to curl and disfigure. The year was 1999.

By year 2000, one short year later, my symptoms were like a runaway train. I had a background in science and was not a stranger to research. I turned to the (then) fairly new technology of the internet. When I read about Scleroderma, specifically the horrible itchiness and skin tightening, I knew I had found the answer. I was originally diagnosed with CREST syndrome, although that changed. All aspects of life as I had known it came to a screeching halt. I could no longer adequately perform my job in the finance department of Kootenay Lake Hospital and went on long term disability. I had to quit my fitness activities. The fatigue had me spending most days bed ridden, the outrageous itching spread over my entire body (lasting many years) along with severe skin sensitivity. My hair hurt, even the ambient air on my skin was painful.

Testimony
Sylvia's Story

At age 40 other odd symptoms began to appear!

During my early research I found the SABC.

The following 8 years were a blur. My kidneys failed within 3 years of diagnosis for which I spent 6 weeks in hospital. My weight plummeted over 20lb to 100lb. I developed watermelon stomach causing constant anemia and blood loss of approximately 1 unit of blood/week. Without blood transfusions at least biweekly for about 10 years I wouldn't be alive to tell you, my story. I had 20+ gastroscopies with laser trying to stem the bleeding.

Eventually, I started receiving EPO injections which allowed my body to slowly begin manufacturing more red blood cells. The ratio of blood manufacturing to blood loss became almost equal and I haven't needed transfusions for about five years. This was revolutionary for my day to day to life and something I am so thankful for.

During the course of managing my disease I have experienced multiple finger surgeries, ulcers, calcinosis, numerous serious infections, months of IV antibiotics, hospitalizations, reflux, major pain management, ongoing gastrointestinal challenges, food issues, weight loss, skin tightening and thickening. I saw so many specialists, including Dr. Furst (then in Seattle) and those at the Scleroderma Clinic at St. Paul's in Vancouver and Dr. Erin Browne, plastic surgeon specializing in hands.

Some of my early symptoms eventually backed off. After several difficult years the skin itching eventually became intermittent, the pain greatly reduced, and my blood loss and blood pressure were under control. After having spent so much time in bed I was weak and unstable, sometimes accessing a wheelchair; I needed to teach myself how to walk again. I used a walker (short term), progressed to a cane, then walked holding the cane (my security blanket!) but not using it and eventually left the cane at home. I now walk, even mildly hike on a regular basis.

My husband, dog, family and most of all, my fabulous home care nurses and doctors all gave me undying support. I'm so thankful. Do I still have numerous oandréngoing symptoms? You bet I do, but from the get-go I decided Scleroderma wasn't going to get every itty-bitty part of me. I was going to fight. To quote a line from the movie 'Shawshank Redemption', "you either get busy livin' or you get busy dyin." Well, I certainly wasn't going to settle for option number 2.

I cautiously rejoined life in ways that were 'Scleroderma Friendly'. I became a SABC member, signed up for the Sclerodata newsletter and eventually became the Community Representative for the Nelson area. I read articles and found links to other resources. These were people who were living in my world and spoke my disease language. It was and still is a way to stay connected and informed, especially since there is no support group in my area.

Fast forward to 2020, the onset of the pandemic. In the Sclerodata I had read about the SPIN/SSLED course being offered. Rosanne Queen reached out to me, encouraging me to sign up; not only because they wanted all community reps to have the training, but also for the educational/leadership benefits for anyone living with scleroderma. So at the beginning of 2023 I decided I would like to become more involved with SABC and a good starting point was enrolling for the online SPIN/SSLED program. I'm so glad I did! For 13 weeks, one day per week, my group met virtually. The course was really well done and is a free resource anyone with Systemic Sclerosis can sign up for. I'm now helping out with co-facilitating some of the SABC monthly virtual support group meetings, getting to know the other dedicated people involved and connecting to the broader Scleroderma community.

The more smiles and laughter the better. Scleroderma continues to throw me curve balls and will never be something that comes to a close. It remains poorly understood and only other people experiencing the disease can fully grasp the broad impact it has on our lives, making connection with those individuals even more special. Yet I don't want Scleroderma to be the definition of my life. I count my many blessings every day, strive to live as "normal" a life as possible, and care about the quality of my days rather than the quantity! We're all in this together and hopefully, one day, there will be broad medical advancements to address this difficult disease.

CALCINOSIS IN SYSTEMIC SCLEROSIS

Dr. Thaisa Cotton, MD
Rheumatology fellow,
Centre hospitalier de l'Université de Montréal

Dr. Sabrina Hoa, MD MSc FRCPC
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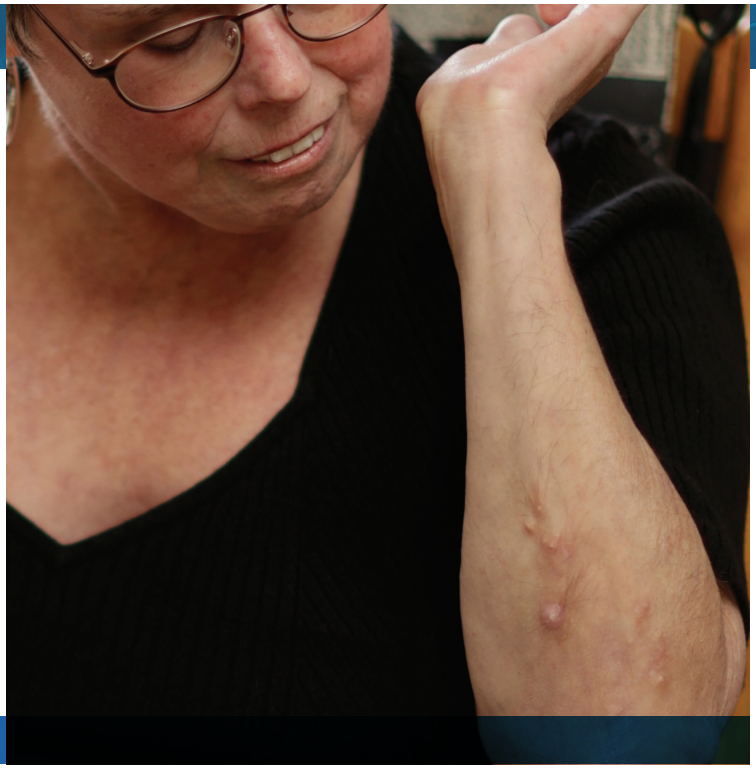

We warmly thank Josée Fontaine for allowing us to publish her photos on pages 12 and 13.

Systemic sclerosis (or scleroderma) is characterized by many skin changes, including calcinosis. In this article, we will discuss what is calcinosis, how it is diagnosed and available treatments.

WHAT IS CALCINOSIS?

Calcinosis is an accumulation of calcium in the skin and surrounding tissues. It can affect up to 20-40% of people with scleroderma, with a similar rate between limited and diffuse scleroderma. It often occurs on the hands, forearms, elbows, and knees, despite normal calcium levels in the blood. Small lesions may go unnoticed, but may also cause pain, interfere with joint function, or be complicated by ulcers, infections, or nerve compression. Unfortunately, few cases of calcinosis improve spontaneously with time, with most lesions remaining stable or worsening after 1 year.

CALCINOSIS IN SYSTEMIC SCLEROSIS

WHY DOES CALCINOSIS DEVELOP, AND WHICH SCLERODERMA PATIENTS ARE MOST AT RISK?

We still do not completely understand why calcinosis develops. Studies have found that people with scleroderma who are men, who have ulcers on their fingers, who have osteoporosis, or internal organ involvement of scleroderma (especially lung fibrosis) are at higher risk for calcinosis. Some autoantibodies have also been associated with calcinosis, including anti-centromere, anti-PM-Scl and anti-RNA-polymerase-3 autoantibodies. Other potential factors include longer disease duration, poor blood circulation, and trauma.

HOW IS CALCINOSIS DIAGNOSED?

It is often felt by the physician on physical exam and can be confirmed by plain radiographs. Other ways to identify calcinosis include ultrasound, computed tomography and magnetic resonance imaging.

HOW DO WE TREAT CALCINOSIS?

Calcinosis remains a challenging aspect of scleroderma to manage, as there is no cure or highly effective treatment. We generally do not treat calcinosis that does not bother a patient. At present, no medical therapies have proven to be efficacious in large randomized controlled trials.

General measures

Generally, it is recommended to avoid injuries and ensure good blood flow to prevent new calcinosis, as these are thought to play a role in calcinosis development. This includes smoking cessation, avoiding exposure to cold and stress, and managing Raynaud and ulcers on the fingers. If a wound develops, proper wound care is important, and antibiotics should be given for nonhealing and infected ulcers. If there is an open wound, warm soaks can help squeeze out calcinosis and may help prevent infections.

Medical therapies

Sodium thiosulfate is thought to improve calcinosis because it binds to calcium. A systematic review that looked at 40 studies (including 1 small randomized controlled trial) found that the topical form of this medication improved calcinosis in over 80% of patients over an average of 5 months. Topical sodium thiosulfate needs to be prepared by a pharmacist in a mixture with either Vaseline or zinc oxide. Possible side effects include skin irritation, allergy to zinc and pain with application. Sodium thiosulfate can also be injected directly into calcinosis lesions, usually by a dermatologist. A case series of 5 scleroderma patients showed improvement or complete resolution of calcinosis with weekly injections for 4 weeks. A common side effect is a burning sensation in the area of the injection site.

Several medications have also been shown to improve calcinosis in small studies. Two pill form medications, colchicine and minocycline, may decrease the inflammatory aspect of calcinosis, when there is associated pain, redness, warmth and swelling, as shown in small studies involving less than 10 patients each. One of the side effects of colchicine is diarrhea, while the side effects of minocycline include nausea, dizziness, and a blue-black discoloration of calcinotic lesions. Other medications, such as diltiazem, bisphosphonates, rituximab, and intravenous immunoglobulins, have been reported to help calcinosis in small studies, but with no larger or higher quality studies to support these observations.

CALCINOSIS IN SYSTEMIC SCLEROSIS

Finally, neem oil with Hypericum plant extract is an herbal therapy that is often used in wound care. A pilot study of 21 scleroderma patients with open calcinotic wounds followed over 40 days found that neem oil helped complete healing in 45% of patients, perhaps by softening and facilitating excision of calcium deposits. This would need to be further supported in larger studies.

Interventions

Surgery to remove calcinosis can be an effective treatment and is considered for lesions that are affecting hand or joint function, compressing a nerve, or causing severe discomfort. Complications include recurrence in 15%, delayed wound healing in 13%, and wound infection in 10% of cases.

Extracorporeal shockwave therapy uses acoustic shock waves to break apart and destroy calcinosis. It is performed by physiotherapists who have received the appropriate training. It can help relieve pain associated with calcinotic lesions that are not amenable to surgery. Several prospective studies have found improvements in pain and size of calcinotic lesions. It often involves 3 to 5 sessions done every 7 to 10 days.

IN SUMMARY

Calcinosis is a feature of scleroderma that is common but unfortunately difficult to treat. Important preventative measures include avoiding trauma and ensuring good blood circulation such as through smoking cessation, cold avoidance, proper management of Raynaud, and treating ulcers. For calcinotic lesions that are symptomatic, topical or injected sodium thiosulfate may be helpful. Surgery may be considered for lesions that significantly affect function and quality of life. More research is needed to understand the cause and optimal treatment of calcinosis.

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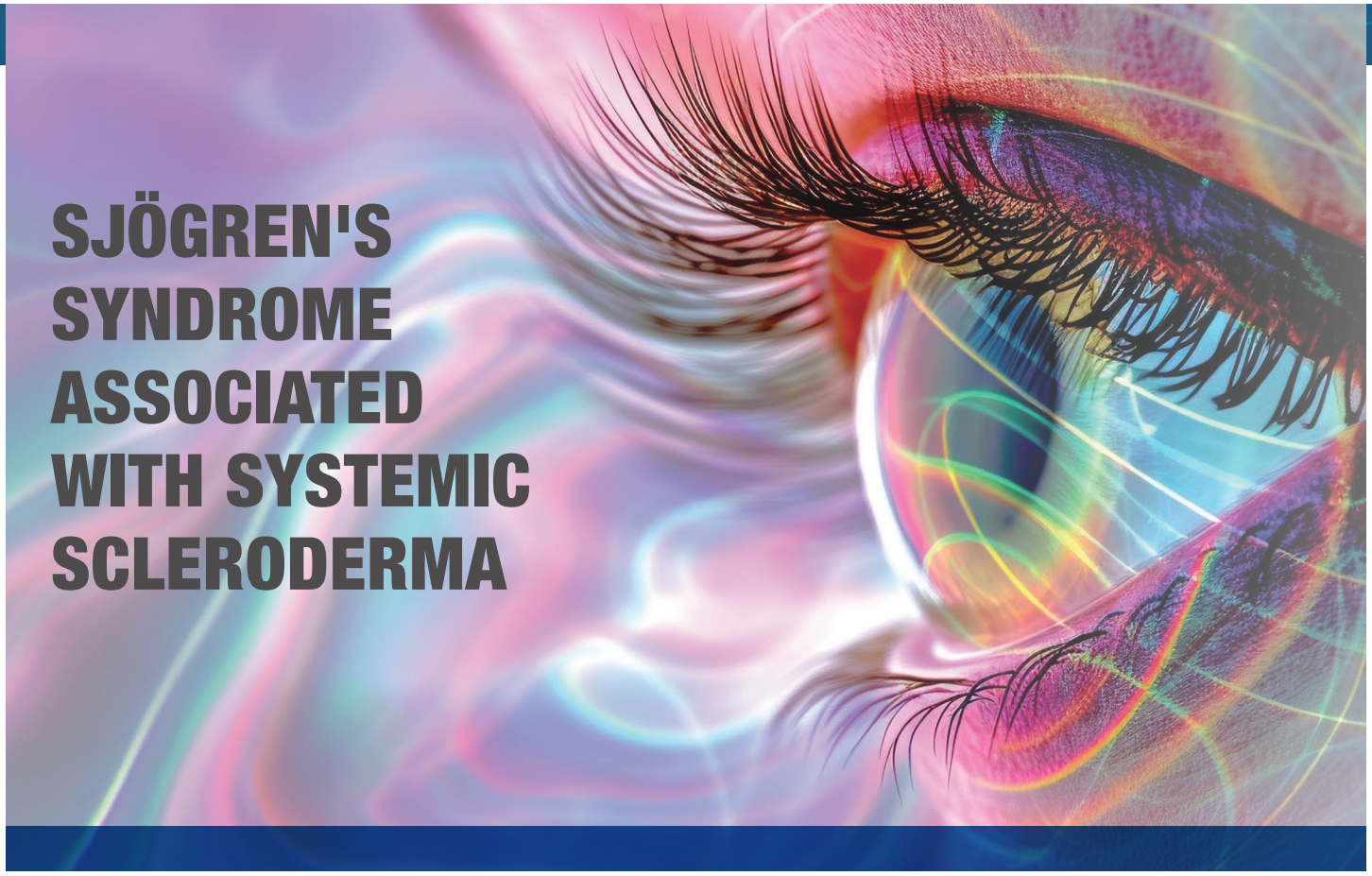

SJÖGREN'S SYNDROME ASSOCIATED WITH SYSTEMIC SCLERODERMA

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Systemic scleroderma is an autoimmune disease that affects the functioning of small blood vessels and leads to excessive scarring. Individuals with scleroderma are at a higher risk of developing other autoimmune diseases, such as Sjögren's syndrome. In this article, we will discuss what Sjögren's syndrome is, and how it is diagnosed, treated, and monitored.

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WHAT IS SJÖGREN'S SYNDROME?

Sjögren's syndrome is a chronic autoimmune disease that causes the immune system to attack the glands responsible for producing saliva (salivary glands) and tears (lacrimal glands). This results in reduced saliva production, leading to a dry mouth and dental problems such as tooth decay. Damage to the lacrimal glands causes excessive dryness in the eyes, with a sensation of having sand in the eyes, which requires the regular use of artificial tears. Although Sjögren's syndrome is commonly known as "dry eyes and dry mouth syndrome," it can also affect other organs such as the lungs, kidneys, lymphatic system, and neurological system.

I HAVE SCLERODERMA. HOW LIKELY IS IT THAT I ALSO HAVE SJÖGREN'S SYNDROME?

It is estimated that about 20% of patients with systemic scleroderma also have Sjögren's syndrome. This is the autoimmune disease that is the most commonly associated with scleroderma. Patients with limited scleroderma or with anti-centromere autoantibodies are more likely to develop Sjögren's syndrome. However, patients may experience dry eyes or mouth due to other causes like medication side effects.

HOW CAN WE DIAGNOSE SJÖGREN'S SYNDROME?

The diagnosis of Sjögren's syndrome is based on a combination of clinical symptoms, blood markers, and disease-specific tests. Blood markers such as anti-SSA, anti-SSB, and anti-Ro52 can be markers of Sjögren's syndrome. An oral medicine specialist can measure the amount of saliva produced to confirm a dysfunction of the salivary glands, and an ophthalmologist can assess dry eyes using the Schirmer test (which measures the amount of tears produced over a five-minute period) and tests to detect any damage to the surface of the eye caused by a lack of tears. A biopsy of the salivary glands in the lip may also be performed to confirm the diagnosis; however, this procedure is rarely required in the context of scleroderma, as the results usually do not change the treatment being administered for Sjögren's syndrome.

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HOW DO WE TREAT SJÖGREN'S SYNDROME?

Treatment for Sjögren's syndrome primarily aims to relieve the symptoms of dry eyes and dry mouth. Some environmental and lifestyle changes are recommended to prevent worsening of these symptoms (see Tables 1 and 2).

For dry eyes, over-the-counter artificial tears in the form of eye drops can be used throughout the day. If eye drops are not effective enough, artificial tears in the form of gels can be tried. Ointments are best used at bedtime to avoid blurred vision. Preservative-free products are preferred and can be applied every 2 to 4 hours, whereas products with preservatives may increase inflammation if used more than 4 times a day. If symptoms persist, your ophthalmologist may recommend more intensive treatments. Omega-3 supplements (2000 to 3000 mg per day) may also help with symptoms.

For dry mouth, sugar-free products such as chewing gum and candy can help stimulate salivation. It's important to use sugar-free products to prevent tooth decay. Long-lasting lozenges can also be inserted inside the mouth. Saliva substitutes (artificial saliva) are available over the counter in mouthwash, spray, or gel form.

Oral medications to stimulate saliva and tear production (pilocarpine - Salagen®; anethole trithione - Sialor®) may be prescribed if the above treatments are not effective. These medications are effective in improving dryness in 60-70% of patients and may also improve dryness of the skin, nose and vagina. These drugs can cause side effects such as hot flashes, sweating, nausea, headaches and increased urination, but can be well tolerated when dosages are adapted and when taken with food. They are not recommended for people with angle-closure glaucoma, severe asthma, or liver dysfunction.

It is strongly recommended that people with Sjögren's syndrome have regular dental checkups every 3 to 6 months because of the increased risk of tooth decay. High fluoride toothpastes, mouthwashes, gels, and varnishes may be prescribed to prevent cavities.

ARE THERE ANY OTHER COMPLICATIONS TO WATCH FOR IN SJÖGREN'S SYNDROME?

People with Sjögren's syndrome should also be monitored for complications involving internal organs, such as the lungs, kidneys, lymphatic system, and neurological system. This includes a medical questionnaire, physical examination, and annual blood tests. It is important to report symptoms such as involuntary weight loss, persistent swelling of the salivary glands (in front of the ears or under the jaw), or swollen lymph nodes to your doctor, as these symptoms can be indicative of lymphoma, a complication that can affect around 5% of people with Sjögren's syndrome.

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TABLE 1
TIPS TO IMPROVE DRY EYES BY MAKING ENVIRONMENTAL AND LIFESTYLE CHANGES

- Avoid being in low-humidity environments.
- Use a humidifier in your bedroom to increase moisture in the air.
- Avoid being in areas with cold air currents, such as those created by air conditioners or fans.
- Stay away from smoke and dust.
- Avoid using eye makeup.
- Take short breaks with eyes closed when reading or using a computer to reduce strain.
- Apply warm compresses to the eyelids for 5 to 10 minutes at a time, 2 to 4 times a day to increase glandular secretion.
- Avoid wearing contact lenses. If you must wear contact lenses, use disposable lenses and replace them daily.
- Wear safety glasses with side shields or moisture chamber glasses that slow the evaporation of tears (such as Ziena® or 7eye® glasses).
- Consult your doctor about medications that may cause dry eyes and that can be avoided.

* Table adapted from the *Guide de traitement de la xérophtalmie et de la kératoconjonctive sèche chez les patients atteints du syndrome de Sjögren* (A Treatment Guide for Xerophthalmia and Keratoconjunctiva Sicca in Patients with Sjögren's Syndrome), with kind permission from Dr. Alexandra Albert and Dr. Marie-May Collin-Castonguay.

TABLE 2
TIPS TO IMPROVE DRY MOUTH BY MAKING ENVIRONMENTAL AND LIFESTYLE CHANGES

- Stop smoking, as it can dry and irritate the mouth and increase the risk of developing candidiasis (fungal infection) and periodontitis (gum disease).
- Stay hydrated by drinking small amounts of water frequently to keep your mouth moist.
- Gargle with olive oil or coconut oil to soothe your mouth.
- Avoid acidic or sweet drinks such as carbonated soft drinks, caffeinated drinks (coffee, tea, energy drinks) and alcohol.
- Avoid acidic foods such as citrus fruits, kiwi, pineapple, strawberries, etc.
- Avoid very hot drinks or foods.
- Avoid very dry or hard foods.
- Avoid very spicy foods.
- Accompany foods with sauces to make them easier to swallow.
- Prefer meats that have been simmered or cooked in foil.
- Prefer fatty meats.
- Avoid sweet, sticky foods to reduce the risk of tooth decay.
- Brush your teeth after each meal. If you cannot, rinse with water.
- Floss daily.
- Consult your doctor about medications that may cause dry mouth and that can be avoided.

* Table adapted from the *Guide de traitement de la xérostomie chez les patients atteints du syndrome de Sjögren* (A Guide to the Treatment of Xerostomia in Patients with Sjögren's Syndrome) with kind permission from Dr. Alexandra Albert and Dr. Marie-May Collin-Castonguay.

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CONCLUSION

Sjögren's syndrome is an autoimmune disease that can affect people with scleroderma and cause dry mouth and dry eyes. The main focus of treating this syndrome is to relieve the symptoms by optimizing the environment and lifestyle, using local treatments, and prescribing medications when needed. Regular check-ups with specialists in rheumatology, ophthalmology and dentistry are recommended to ensure proper treatment and to detect possible complications.

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Community Contact Representatives

CONNECT WITH THE SCLERODERMA COMMUNITY IN YOUR AREA!

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

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