AUTUMN

scleroderma victoria

NEWSLETTER AUTUMN 2019

EDITION 150

40 years Young!

Welcome to 2019!

We are very proud to announce that 2019 is Scleroderma Victoria's 40th anniversary of operation.

Along the way, our Scleroderma community have contributed to our success, and we are very grateful for your contributions and support. We couldn't have done it without you.

In 2019 we will have some special social media posts on our 40th anniversary, and hopefully obtain some great stories of our history from some of our founders and long-term members.

You can join in and use our hashtag #40Scleroderma. We hope to commemorate this achievement of supporting our community in many different ways – ideas and thoughts are welcome on how we can do this. Please send your ideas to newsletter@sclerodermavictoria.com.au

The success of our previous year's events has generated engaging dialogue for us to continue with both our World Scleroderma Day Lunch at the Grand Hotel in Richmond and our inaugural More than Skin Deep Fashion Parade.

You can find out more on these events in this newsletter, our website as well as our social media pages.

Our aim for 2019 is to raise the bar on our awareness campaigns, and grow as an organisation to provide substantial benefits and long-term sustainability for our Scleroderma community. I am looking forward to sharing another great year ahead filled with excitement and success.

Thank you to everyone, and I am pleased to say I am excited about the year ahead!

Amanda Lawrie-Jones
President, Scleroderma Victoria.

Scooter finds a home

We have found a very grateful recipient for our mobility scooter.

He is Robin Henry, a member of Scleroderma Victoria who attends our Gippsland Support Group.

Robin is pictured above with Jane Rhyder (committee member and North Eastern Support Group Leader).

Jane and her husband Andrew drove to Gippsland and delivered the scooter to Robin at his home recently.

Thank you again to Janice Burke who donated the scooter.

Kathleen's advice – Don't ever give in

Kathleen Flanagan talks to Bridget Naughton about Scleroderma and her days as secretary of Scleroderma Victoria.

Kathleen Flanagan is an energetic vibrant 84-year-old woman who lives in Moonee Ponds and spends her spare time reading to the children in the child care centre nearby.

She is one of two children her Irish mother and father had and grew up in Brunswick. Kathleen never married and does not have any children, however, does have second cousins who visit her on a regular basis.

Kathleen has a love of golf, table tennis, cooking and travelling.

Whilst she has travelled through many different countries, her favourite place is Margaret River in WA.

Kathleen's love of travel found its way into her work life when she landed herself a job as a radio broadcaster on 3RPH in early adulthood and had a segment about world travel.

Kathleen's first signs of any Scleroderma-related issues started when she was only 16=years-old when she was a volunteer at her local CFA. She played regular games of table tennis and started to experience what she termed "dead fingers". Due to her unfamiliarity of her issues, Kathleen found herself writing to Anne Mawdsley who was a campaigner and fundraiser for Raynaud's in England.

Kathleen said it was around this time she also found herself putting on weight for no reason and struggling with fatigue.

Kathleen's diagnosis of Scleroderma wasn't the first time she had heard of this condition, with a family friend having it from when she was young. Now that Kathleen thinks about it, she also had an aunt always complaining of "dead fingers" and a second cousin who suffered from Lupus.

It wasn't until Kathleen was in her 30's that she saw a vascular physician who diagnosed her initially with CREST and later Scleroderma. She was then referred to a Rheumatologist Dr Kim Boyden who counselled her on the condition and different medications. Kathleen is now a patient of Dr Wendy Stevens who has kept her as healthy and vibrant as she is today.

Later in life, Kathleen became a teacher however retired at 56 to take care of her father who was blind from Macular degeneration, the same thing her late sister lived with.

She said she didn't have time to think about her condition, as the primary carer for her father kept her busy. She dare not thought about dying, because who would look after the family dog?

Kathleen chats with Bridget Naughton.

Kathleen said there was one positive to getting older with Scleroderma and that was the onset of menopause, as circulation significantly improved and "dead fingers" disappeared. Kathleen was greatly disappointed it didn't take the sausage like fingers with it.

During this time she found herself looking up information about support groups in the phone book and found Scleroderma Victoria.

It wasn't long between Kathleen attending her first Scleroderma Victoria meeting, to filling in as secretary whilst her friend Giselle J'dour was unwell, to taking over this position from her in later years.

The committee at the time of Kathleen joining was made up of patients, parent's or spouses of patients, a little like Scleroderma Victoria today.

She said aside from the common duties of a secretary, she would often go and visit Scleroderma patients in hospital with this commonly being the first time the two of them had met.

Kathleen's remembers her time on the committee as a great opportunity to connect with like-minded people where she could gain greater understanding of others dealing with similar experiences with Scleroderma.

Kathleen advised that the biggest change she has witnessed over the years with Scleroderma is the way they treat it.

In her eyes medications have come a long way, especially when she was initially diagnosed they still wanted to perform sympathectomies. She is fortunate that she doesn't take a lot of medication these days as her Scleroderma is only limited with only minor heart issues, sausage fingers and the need for a walking frame, however it can be complicated by her overlapping Hashimoto's disease.

Kathleen now lives by her own advice to us all; "You've just got to keep going, get your hair done and do your makeup, otherwise you've given in."

Thank you to Kathleen for spending the afternoon with me and sharing your insights.

*- **Bridget Naughton***

HAIR RAISING FUNDRAISER

Making his mark!

It was three years ago when my wife Melissa was diagnosed with Scleroderma. It was a confusing time and we found it hard to find out information on what to expect.

We went to the Scleroderma Fashion Parade at the Collingwood Town Hall and this was the first time Melissa really got to speak with other sufferers of Scleroderma.

I could tell it helped her to know that there is a support network out there.

I have always wanted to do something to raise awareness of Scleroderma but didn't know how.

Funnily, it was Melissa who posted on social media that I would grow my hair to raise funds for Scleroderma research. She didn't tell me until all her friends had committed to a donation.

I think they were all amused because I am bald for a reason – something to do with genetics!

However, I thought I could give it a go and have people laugh at my efforts whilst raising money for Scleroderma.

After setting up the fundraiser in January we were so amazed and humbled that

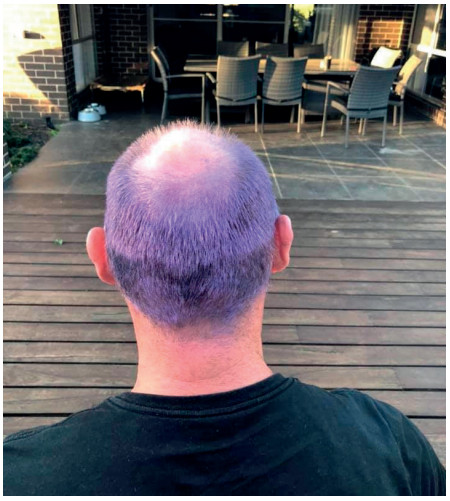
within three weeks we had raised over $4000 for Scleroderma Victoria.

I raised the goal after two weeks and committed to dying my hair a colour if we could raise enough; needless to say, my little amount of hair was dyed purple.

Melissa and I are so thankful to everyone who was so generous; we had friends, family, colleagues and people we had never met donate money.

It made us appreciate the support for Melissa and other sufferers of Scleroderma and we hope that it raised awareness of Scleroderma more broadly.

I am sure that Scleroderma Victoria will put the funds to good use and that one day there is a cure for this terrible disease.



Mark and his wife Melissa and below our purple-haired fundraiser

Dr Barnett's legacy continues to grow

- by Robyn Sims
President, Scleroderma Victoria 2011-2015

"From little things, big things grow".

We are all familiar with these famous lyrics written by Australian artist Paul Kelly.

These words also apply to our wonderful organisation Scleroderma Victoria.

Now celebrating its 40th Anniversary, our organisation was the inception of the late Dr Alf Barnett, who introduced three of his patients to each other. They then went on to form Scleroderma Victoria.

When I was involved with the committee, I was the person in charge of organising a roster for volunteers to man the office and monitor phone calls. I also spent some time organising old files.

I came across many photos and documents which highlighted the work of those in past years who, without the help of electronics, gradually built up Scleroderma Victoria into what it is today.

Two particular documents stand out. They were the handwritten minutes and treasurer's report, in pencil.

I recall the balance was somewhere in the vicinity of $300. The newsletters were typed, copied and then mailed to members.

There were very few people on the committee, and they worked tirelessly.

At a couple of stages during the past 40 years it seemed that the organisation would close. After many years Heather Holloway and her late husband John, were left with very little support and were unable to continue, suffering from what we called burn-out.

At a function at Emu Bottom in Sunbury, Heather spoke with Jan Elliott the owner of the venue, whose daughter had passed away due to scleroderma.

After some hasty negotiations Jan took up the President's role and so on rolled our charity.

With the introduction of the internet Scleroderma Victoria flourished. We were able to reach out to many more people.

A succession of new faces joined the committee with enthusiasm and new ideas.

These volunteers are the lifeblood of our organisation.

I feel privileged to have become a part of this team and have gained many wonderful friendships, travelled overseas to Conferences and have a greater understanding of systemic sclerosis and the work into research which is now coming to fruition.

SSc patients show improvements after CD34-selected stem cell transplant

by Santiago Gisler
Scleroderma News

Systemic sclerosis (SSc) patients showed reduced skin thickness and improved lung function after a hematopoietic stem cell transplant (HSCT) using blood progenitor cells positive for the CD34 marker, a study reports.

This finding suggests that CD34-selected autologous HSCT may have additional clinical benefits over conventional autologous HSCT. The study, "CD34-selected versus unmanipulated autologous haematopoietic stem cell transplantation in the treatment of severe systemic sclerosis: a post hoc analysis of a phase I/II clinical trial conducted in Japan," were published in the journal Arthritis Research & Therapy. Autologous HSCT is a process in which a patient's blood stem cells are collected and reintroduced back into the patient after a chemotherapy treatment course to eliminate faulty immune cells.

The technique is often used in patients with cancer or autoimmune diseases, such as SSc, because it is thought to reset the immune system by removing autoreactive immune T-cells and B-cells.

Studies have shown that autologous HSCT can relieve skin symptoms and improve lung function in SSc patients, and lead to significant long-term and event-free survival.

In cancer patients, physicians often choose to select stem cells for HSCT based on the CD34 marker, which allows them to obtain highly purified tumour-free cell collections from patients with malignancies.

In SSc patients, though, it is not clear if the selection of CD34-positive cells prior to transplant affects HSCT outcomes.

To test if CD34-selected HSCT could offer additional clinical benefits to patients with SSc, researchers in Japan evaluated data from a non-randomized Phase 1/2 clinical trial assessing 19 SSc patients (mean age of 53.7 years), 11 of whom received a CD34-selected autologous HSCT, and eight who received a non-manipulated autologous HSCT.

All patients were treated with high-dose cyclophosphamide (200mg/kg) monotherapy (chemotherapy regimen) before the stem cell transplant. Changes in skin sclerosis and pulmonary function were assessed over an eight-year follow-up period.

Most of the patients had moderate to severe skin sclerosis, established by a modified Rodnan skin score (mRSS; evaluating changes in skin thickness, a hallmark of SSc) of 15 or more (mean mRSS of 26.7). Higher mRSS values correlate with worse skin sclerosis.

Results showed a progressive improvement in mRSS after HSCT in both groups, with the mean mRSS decreasing from an initial 26.7 to 9.1 at the five-year mark. The CD34-selected group demonstrated significantly more improvement over the non-manipulated group.

To assess lung function, researchers measured forced vital capacity (FVC), which measures the amount of air forcibly exhaled from the lungs, and diffusing capacity of carbon monoxide (DCLO), which measures oxygen transfer from the lungs to bloodstream.

While DCLO levels were stable in both groups over the eight-year follow-up period, the researchers found a gradual improvement in FVC after the transplant in the CD34-selected group — from 71.5% at the start to 82.1% at five years and 84.8% at eight years.

In contrast, the non-manipulated group showed temporary FVC improvements at six months, which then returned to initial values three years after transplant.

The team also found that the CD34-selected group showed better progression-free survival (81.8%) at five years than the non-manipulated group (50%).

Results showed that the frequency of severe adverse events, such as bacterial infections or organ toxicity, was similar between the two groups. Viral infections were more common in the CD34-selected group (affecting nine patients), than in the non-manipulated group (two patients).

Nonetheless, all patients were successfully treated with or without anti-viral medications. No treatment-related deaths occurred in either group.

The team concluded that "the results of this study show that improvements in skin sclerosis and pulmonary function due to CD34-selected [autologous] HSCT are maintained for at least 8 years, which is superior to results achieved with unmanipulated [autologous] HSCT."

They suggest that given the fact that immune T-cells are "extremely deleted by the positive selection of CD34+ cells," the use of CD34-selected autologous HSCT "has a benefit in that it prevents reinfusion of autoreactive [T-cells] that may be associated with SSc."

The history of research into stem cells is not as recent and one might imagine. More than 100 years ago scientists presented evidence that stem cells existed, were germinal in nature (being in the earliest stages of development) and were undifferentiated.

The term regenerative medic9ine is often used to describe medical treatments and research that restore the function of organs or tissues.

Fast forward to the present day, Stem Cell Therapy is touted as the news frontier of regenerative therapy.

As with all new innovative methods the way forward will be cautious, steady, while developing expertise and methods.

Scleroderma spells *fatigue*

A summary of St Vincent's Scleroderma Nurse Barbara Gemmell's address to the Gippsland Support Group on fatigue.

Fatigue is a well-documented symptom in Scleroderma.

When the disease is active, symptoms of fatigue and low energy are common, often coupled with an inability to sleep normally.

The process of inflammation is considered a major cause of fatigue – white blood cells and cytokines (chemicals that cause pain and swelling) are the main suspects – so working to reduce inflammation, whether it be medication, exercise, diet or relaxation techniques may actually, assist in relieving this disabling problem

Barbara said there are many ways of coping with fatigue. These include:

- Pace your activities throughout the day
- Ensure sufficient sleep (low caffeine, bath, warm milk, lavender pillows, exercise during day, +/- medications)
- Delegate some chores
- Rest when you need
- Exercise regularly
- Reduce stress
- Maintain a well-balanced diet
- Learn to accept (but not surrender to) fatigue

Barbara also outlined some of the contributors to fatigue:

- Anaemia - consider timing of supplementation,
- Diet
- Depression
- Stress
- Pain

She said there could also be other medical conditions such as an underactive thyroid.

Side effects of medications can also contribute to fatigue, such as some pain killers. Changes in fatigue that coincide with medication adjustments should be reported to your specialist.

With winter fast approaching, Barbara offered some tips on Raynaud's. They were:

- Avoid allowing the blood vessels to spasm
- Avoid cold exposure
- Use gloves whenever outside
- Think about time of day
- Avoiding water immersion
- "Holiday at home in the Tropics"

Help – when and what is available?

Informal Assistance:

Neighbours, friends and family are often happy to help short term or in crisis.

Community Aid:

District nurses, home help, meals on wheels, drivers for appointments, - generally accessible through your GP or council

Care packages:

For people who struggle to manage either the cost or the day-to-day problems of organising home and/or medical care, there are government subsidised options: Either aged care, or non-age restricted

Low- and high-level care

Managed by a variety of agencies, some with religious affiliation, others not so.

Low cost to the patient

Flexible to the patient's needs at the time

Indefinitely on-going usually

Summary

Help is available from a variety of sources – use it!

Independent Health Solutions have some great gizmos to help with fatigue – try to visit their website

www.independenceaustralia.com/ and see what they have on offer.

Picture shows Dee, Trevor, Barbara Gemmell, Corrie (Gippsland Support Group Leader), Judy and Elaine.

Meet your committee

Jane Rhyder

Jane joined the committee in 2015 as a representative with special interest in support groups having started the North East support group in 2011.

Jane received a swift diagnosis of scleroderma whilst living in the UK in 2001. Raynaud's symptoms prompted a visit to the GP and then to a hospital connective tissue clinic.

Scleroderma was something she knew nothing about.

What made you support Scleroderma Victoria and what were the reasons you became a paid member?

Once back in Melbourne it was at a seminar at St Vincent's Hospital that I first heard about Scleroderma Victoria and decided to become a member. By joining I have benefited by being part of a larger scleroderma community.

Also, it's good to know my contribution is helping to raise awareness of this disease and fund future research.

In 2011 I was encouraged to start a support group where I live and as a result have met many others in my area living with scleroderma in all its many forms. It has been a pleasure to organise get togethers over the years that have enabled us to share our journey with scleroderma with each other.

It's great meeting with others who understand what you are going through and where no explaining is required.

Why did you become a Committee Member?

After attending many seminars and fund raising events over the years and getting to know some of the committee members, I decided to join the committee.

I'm keen to help promote support groups and would love groups to be set up across Victoria so more people have the opportunity to meet with others living with scleroderma and receive much needed support.

What is the best thing about being involved with Scleroderma Victoria?

I have got to know and am working alongside enthusiastic and interesting people from a variety of different backgrounds, most of whom are navigating their way living with scleroderma.

I enjoy being a part of the different events the committee have organised, helping where I can.

Our first 'More than Skin Deep' fashion parade in 2017 was a highlight for me especially meeting our ambassador Dyson Heppell.

What would you like to see Scleroderma Victoria achieve in 2019?

Last year was exceptionally good for raising awareness and much needed funds.

It would be good if we can build on this success.

Tell us about your other interests and what drives you?

I am a member of a local Christian community church assisting with the running of two playgroups.

I love arts and crafts especially embroidery and crochet. Healthy eating is important to me as is taking my dog out for regular walks which is good for both of us.

What is Iloprost?

What is it used for? How does it work? How does it help?

Iloprost is a drug that comes under the prostaglandins group of treatments. It is used to treat a range of conditions, such as scleroderma, Raynaud's phenomenon, pulmonary hypertension and other diseases where blood vessels are constricted, and so blood cannot flow to the tissues in affected areas.

It is important that issues like these are treated, otherwise the tissues can become damaged and lead to high blood pressure.

Iloprost is prescribed if a person is suffering from ulcers of the fingers, if there is gangrene, or if a person has severe Raynaud's Phenomenon, and if other drugs such as nifedipine have not been successful in relieving symptoms.

What is it used for?

Iloprost is a synthetic (man-made) version of prostacyclin. This is a molecule that is produced in the body and is a natural vasodilator. This means that it relaxes the walls of blood vessels, so that it is easier for blood to flow through.

Iloprost is helpful as:

1. It widens/dilates the blood vessels, helping them to transport more blood to all areas of the body. This increases warmth in affected areas such as the hands and feet.
2. It reduces the tendency of the blood to clot.
3. It helps to prevent and repair damage to the blood vessels.

It usually starts to come into force and show benefits immediately after administration, but it can sometimes take up to six weeks. Cold hands and feet may warm up straight away, and ulcers may begin to improve and heal within a few days.

The positive effects of iloprost may carry on for weeks and sometimes even months after treatment.

How is it administered?

Infusion

Iloprost may be given through an infusion (drip) into your arm. This is usually continuous for ~6 hours a day for 3-5 days in a row in a hospital or in a clinic.

It can sometimes be given continuously over 24 hours. In some hospitals, you will stay on the ward for three to five days, whereas in others you will attend the day-case unit during the day, and then return home in the evenings.

The frequency of iloprost infusions is normally every six months, but this can be adjusted depending on the person's needs.

Inhalation

Iloprost can also be administered through inhalers rather than infusion.

Initially, the process of iloprost inhalation will be started for the patient in hospital, where they will be shown how to inhale the solution, using a nebuliser. A nebuliser is a device that turns the iloprost solution into a fine mist so that it can be inhaled into the lungs.

An individual may have to stay in hospital for up to three days to allow for training and for monitoring their response. After this, the person can return and continue taking the medicine independently.

What are the side-effects?

When receiving treatment in hospital or in a clinic, medical professionals will observe you for potential side effects and how you respond to the treatment, through monitoring of blood pressure and electrocardiograms (test done to check heart's rhythm and electrical activity). If not under observation, such as if inhaling iloprost at home, it is important for the person to be alert to any sudden side-effects.

All side-effects disappear very quickly once iloprost treatment is stopped.

Common adverse reactions include facial flushing, cough and a fall in blood pressure. Others are headache, flu syndrome, nausea and insomnia.

It is crucial to get emergency medical help if any signs of allergic reactions are noticed, such as hives, difficulty breathing and swelling of the face, lips, tongue or throat. Paracetamol and anti-sickness drugs can be taken if side-effects are noticed, but it is important to check with your doctor first.

If any these serious side effects are noticed, call your doctor at once:

- feeling faint;
- pounding heartbeats or fluttering in your chest;
- coughing up blood;
- unusual bleeding (nosebleeds, bleeding gums);
- fever, chills, cough with yellow or green mucus;
- chest tightness, stabbing chest pain, wheezing, feeling short of breath;
- anxiety, sweating, pale skin, severe shortness of breath, wheezing, gasping for breath, cough with foamy mucus, chest pain, fast or uneven heart rate.

Restrictions

There are some circumstances where iloprost cannot be taken. These include if you suffer from:

- unstable angina;
- a heart condition such as heart valve defects;
- any problems with liver function;
- breathing problems such as asthma or chronic obstructive pulmonary disease;
- a peptic ulcer (open sores in the lining of your stomach or the upper part of the small intestine);
- a recent stroke or heart attack;

Your doctor will be able to inform you if you are unable to have iloprost treatment and the full reasoning behind this.

Additional medications

Pending your doctor's advice, usual medications can still be taken before and after a course of iloprost treatment. Your doctor will advise the patient on whether or not they should continue taking medications during a course of treatment, as certain drugs can also act to widen blood vessels or lower blood pressure. To ensure that this is done, SRUK recommends individuals to take a list of their medications with them when going to their first day of treatment.

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Low vitamin D levels linked to Scleroderma

Vitamin D deficiency has been linked to several autoimmune diseases, such as rheumatoid arthritis, lupus and multiple sclerosis, and is seen almost universally in patients with systemic sclerosis (SSc).

The frequent reports of vitamin D deficiency, especially in people from developed countries, may explain the high incidence of autoimmune disorders in these regions.

This has garnered significant attention, alongside recent data suggesting that vitamin D plays a role in the functioning of the immune system.

Vitamin D synthesis is regulated in the kidneys by three molecules: parathyroid hormone (PTH), fibroblast growth factor 23 (FGF23) and -Klotho.

A team led by Dr P Kotyla at the Medical University of Silesia aimed to assess the levels of Vitamin D, -Klotho, and FGF23 in individuals living with SSc, and to determine if there is a relationship between these factors and disease activity.

The role of vitamin D in the pathogenesis of autoimmune conditions is heavily debated and has been addressed in a previous study by An et al. The results from this study showed a reduction in the level of vitamin D in people with SSc who lived in Israel, Italy, China and Brazil; areas of high sun exposure.

The results of the study by Kotyla et al. indicated that a group living with the diffuse variant of SSc exhibited a lower level of vitamin D, significantly reduced levels of FGF23, and similar -Klotho concentrations compared to the control group.

None of these factors correlated with the extent of skin involvement and disease activity. This study only examined Caucasians with low sun exposure, so vitamin D deficiency is not solely related to sun exposure.

This study is the first demonstration of a possible relationship between vitamin D and SSc, through the levels and activity of -Klotho and FGF23. Based upon the results, the authors have suggested that the FGF23: -Klotho ratio is a potential marker of disease activity that may reflect the progression of SSc in individuals, and that this ratio may be contributing to a potential correlation between vitamin D and SSc.

A question that now remains to be answered is if vitamin D deficiency contributes to progression of the condition. It is necessary for there to be further testing before it is distinguished that there is a role that Vitamin D plays in SSc.

Other factors will also need to be considered, such as kidney function in those with SSc compared to a control group. The results could be important however, as they represent a line of questioning that has the potential to lead to a new target for therapy.

Iloprost? Its uses and side effects

...continued from page 6

Iloprost should not be taken with the following medicines except upon the advice of your doctor:

- Antihypertensives (blood pressure medicine)
- Anticoagulants and antiplatelets (blood thinning medicine)
- Non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen

Iloprost does not affect vaccinations, so these can be had before and after a course of treatment. In the small chance that a vaccination is being administered during treatment, a medical professional will be able to provide guidance.

Pregnancy/breastfeeding

There have been no conclusive results on whether or not iloprost affects fertility. The guidelines currently are that iloprost should only be prescribed to a pregnant woman in extenuating circumstances where the disease is severe.

If pregnant or if a person is planning on starting a family, the patient should tell their doctor prior to treatment. Likewise, there has been no research on the drug's activity and influence in breastfeeding, therefore breastfeeding is best avoided whilst taking iloprost.

CONCLUSION

If you have any questions regarding iloprost treatment or any other medications, Scleroderma Victoria strongly encourages you to bring these up with a medical professional.

References:

1. https://www.versusarthritis.org/about-arthritis/treatments/drugs/iloprost/ 2. https://bnf.nice.org.uk/drug/iloprost.html 3. https://www.gosh.nhs.uk/medical-information/medicines-information/iloprost 4. https://www.drugs.com/cdi/iloprost.html 5. https://patient.info/medicine/iloprost-for-pulmonary-arterial-hypertension-ventavis

Impact of Raynaud's on mental health

With winter fast approaching and despite the drastic impact that Raynaud's phenomenon can have on an individual's quality of life, the detrimental effects that the condition may have on mental health are often overlooked.

Raynaud's phenomenon is characterized by the recurrent narrowing of blood vessels in fingers and toes (can also occur at other body sites) due to exposure to cold or stress.

A person can experience colour changes to white, blue and red, as well as sensations of pain, numbness and swelling.

Primary Raynaud's phenomenon (PRP) is when the symptoms occur in the absence of other conditions, whereas secondary Raynaud's phenomenon (SRP) is when the symptoms occur alongside other connective tissue diseases and rheumatologic disorders. SRP is much more progressive and is normally complicated by irreversible structural changes such as ulcers and tissue loss.

The most common approaches to manage Raynaud's tend to be a combination of pharmacological therapies and preventions, but these treatments may not always address the full impact of Raynaud's.

As a psychological reaction to diagnosis is also likely, it is necessary to consider the likelihood of individuals living with Raynaud's also developing depression and anxiety, and suffering from a worse quality of life, and hence to develop appropriate techniques to combat this.

A team led by Dr B Fábián at the University of Debrecen in Hungary carried out an investigation ('Comparison of mental and physical health between patients with primary and secondary Raynaud's phenomenon') that aimed to compare anxiety, depression, physical health and quality of life in people living with primary Raynaud's phenomenon (PRP) versus those with secondary Raynaud's phenomenon (SRP).

Both groups were recruited from the Raynaud's Outpatient Clinic of the Department of Internal Medicine at the university, where they received regular follow-up care from September to December 2017.

This was the first study that aimed to identify the prevalence of symptoms of depression and anxiety in patients with PRP and SRP, to investigate whether SRP patients have more depressive and anxiety symptoms, and to assess if patients with SRP have a more deteriorated physical condition and a worse quality of life than those with PRP.

By assessing the aspects of quality of life that are most important in Raynaud's, it becomes possible to further understanding of the disease and to establish a stronger therapeutic strategy.

Different measures and tests were used to assess the severity of anxiety and depressive symptoms, physical health and quality of life; these included the General Anxiety Disorder Scale (GAD-7), the Beck Depression Inventory (BDI), a Physical Component Scale (PCS-12) and the Raynaud Specific Quality of Life Questionnaire (RQLQ).

In total, 60 individuals with PRP and 41 with SRP were studied – gender distribution, family status, employment status and smoking were similar in the two groups.

Link between Raynaud's and Scleroderma

Most people living with Raynaud's have the primary form of the condition with symptoms of cold hands/ feet, blue and white or purple fingers, where there is no link to any other disease, including scleroderma.

Secondary Raynaud's is the link to scleroderma and is where you have Raynaud's, and another underlying condition.

Secondary Raynaud's can be a sign of scleroderma which can be fatal, of which the three main symptoms are having Raynaud's, swollen fingers and reflux/ heartburn.

There is a very small chance that you will develop scleroderma if you have been diagnosed with Raynaud's, only 0.1% of people diagnosed with Raynaud's will go onto develop scleroderma.

As Raynaud's is often the first symptom of scleroderma to be noticed (for 95% of people with scleroderma, Raynaud's was their first symptom), it is important to get tested and establish whether you have the primary or secondary form of Raynaud's. Your doctor will be able to organise these tests.

Although we talk about people with scleroderma having 'Raynaud's symptoms', there are important differences in the actual disease processes of the two conditions.

The results highlighted major differences between the two groups.

The findings reported the presence of higher levels of anxiety in people living with SRP compared to PRP, indicating greater emotional distress in this group, and anxiety and depression were also reported as being more common in those with SRP compared to PRP. Furthermore, people living with SRP also had lower overall PCS-12 and RQLQ scores than people with PRP.

These results resemble those from previous studies focussing on other rheumatological and autoimmune diseases.

The results of this study contribute to the understanding that a diagnosis of SRP can indicate more severe impact on overall wellbeing than PRP. This may be because in the course of the condition, after the initial symptoms, greater physical disability and more extensive symptoms can occur as a result of SRP.

Despite the implications that anxiety and depression are more common in people living with SRP, it is essential that quality of life impairments are considered when managing all individuals with Raynaud's. Studies such as these represent a shift in the quality of care that clinicians are becoming increasingly interested in providing alongside pharmacological treatment.

Scleroderma Victoria strongly encourages anyone who is living with either primary or secondary Raynaud's and struggling with their mental health to not suffer in silence and tell a medical professional.

Arrhythmia linked to fibrosis in Asymptomatic patients with Scleroderma

- by Jose Marques Lopes (PHD) in Scleroderma News

The expanding nature of inquiring research for scleroderma continues to facilitate the understanding of its impact.

Not long ago the heart was considered unaffected, as we now are aware this is not the case.

Improved investigative methods, regularity of follow up and worldwide collaboration is providing earlier detection and therapy.

A device designed to continuously record heart rhythm showed that asymptomatic (no symptoms of disease) patients with scleroderma have significant arrhythmia, according to a pilot study.

Findings also suggested that arrhythmia — irregular heart rhythm — is associated with fibrosis, or scarring, and the levels of cardiovascular biomarkers.
The study, "Incidental significant arrhythmia in scleroderma associates with cardiac magnetic resonance measure of fibrosis and hs-TnI and NT-proBNP," was published in the journal Rheumatology.

Heart disease in scleroderma is characterised by fibrosis in the myocardium, the cardiac muscle, with or without inflammation.

Although arrhythmia is common, it is not clinically relevant in all patients with fibrosis.

Most research on heart disease in scleroderma has not differentiated between incidental (associated with another disorder) and primary disease.

Researchers have also not evaluated the value of an implantable loop recorder (ILR) — a device that records heart rhythm for up to three years — to screen for significant arrhythmia in asymptomatic but high-risk patients.

Furthermore, researchers have yet to determine the correlation between cardiac magnetic resonance (CMR) findings — such as functional measures and fibrosis — and abnormalities in the cardiac system. To shed more light on these issues, a team led by researchers at Leeds Institute of Rheumatic and Musculoskeletal Medicine, University of Leeds, in the U.K., analysed 20

scleroderma patients with no known cardiovascular disease based on readings from an ILR inserted under the skin and cardiac imaging.

Patients were evaluated every three months over three years.

Assessed parameters included fasting lipid (fat) profile and glucose, creatine kinase — a biomarker of muscle disease activity — high-sensitivity cardiac troponin I (hs-TnI, a biomarker of acute myocardial infarction), and N-terminal pro-brain natriuretic peptide (NT-proBNP), which is produced in response to changes in pressure inside the heart and is a biomarker of pulmonary arterial hypertension.

ILR findings were available for 19 patients — mean age of 53 years, mean disease duration of 7.5 years.

Results showed that eight patients experienced significant arrhythmias over the three-year period: one with complete heart block, two with non-sustained ventricular tachycardia (abnormally fast heartbeat), and five with atrial arrhythmias — originating in the heart's upper chambers.

The median time from ILR insertion to significant arrhythmia was 12 months.

All three patients with serious arrhythmias (complete heart block or non-sustained ventricular tachycardia) had diffuse cutaneous scleroderma with interstitial lung disease. Their mean disease duration was two years.

Patients with significant arrhythmia showed higher initial serum levels of hs-TnI and NT-proBNP, but an unchanged amount of creatine kinase. They also had higher CMR-extracellular volume, which is indicative of diffuse fibrosis.

Five of the 14 patients with CMR data available exhibited focal fibrosis, though only one developed significant arrhythmia.

Greater initial levels of hs-TnI and NT-proBNP were also observed in patients with fibrosis. The higher these values, the lower the myocardial perfusion reserve — the maximum possible increase in myocardial blood flow above initial conditions. Six participants needed medical treatment, including one permanent pacemaker implant, one anticoagulation treatment, and four cases of anti-arrhythmic pharmacotherapy.

The researchers concluded that almost half of the scleroderma patients analysed "had a clinically significant arrhythmia," and that "ILR arrhythmias appeared to be associated with hs-TnI, NT-proBNP and CMR measure of diffuse ... fibrosis."

They suggested that the "disease phenotype [namely diffuse scleroderma], CMR-extracellular volume (indicating diffuse fibrosis), and cardiac biomarkers may identify at-risk patients that would benefit from ILR screening."

Telangiectasia and Autoimmune Disease

by Horatio Wildman, MD
Associate Professor of Clinical Dermatology, Weill Cornell Medical College. Featured in the Scleroderma, Vasculitis & Myositis eNewsletter

What is Telangiectasia?

Telangiectasias are dilated blood vessels located near the surface of the skin or mucous membranes.

They often appear as fine pink or red lines, which temporarily whiten under pressure. Matted telangiectasias are clusters of these small dilated blood vessels that form a pink or red patch on the skin. Although otherwise healthy individuals may develop this condition, telangiectasias are a cardinal feature of systemic and limited scleroderma, as well as dermatomyositis.

Acquired Causes of Telangiectasias

There are many causes of telangiectasias. Perhaps most commonly, telangiectasias are found in areas of chronic sun damage in fair-skinned individuals. They can also be seen on the sides of the nose in healthy adults.

Conditions associated with telangiectasias include:

- Rosacea
- Pregnancy
- Liver Disease
- Chronic systemic or topical corticosteroid use

Connective tissue diseases associated with telangiectasias include:

- Scleroderma
- Dermatomyositis
- Systemic Lupus Erythematosus

Telangiectasias in Scleroderma, Dermatomyositis, and Lupus

Connective tissue diseases often cause telangiectasias to develop on the face and fingernail folds (where the skin meets the nail).

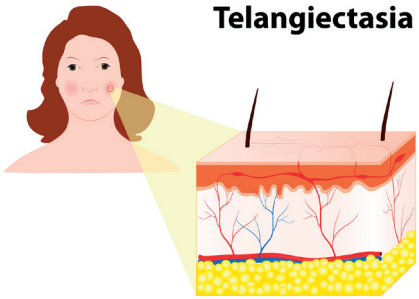
Scleroderma

Patients with scleroderma may develop telangiectasias on the face, mucous membranes, and hands. The condition occurs with both types of scleroderma:

- ***Limited scleroderma,*** (CREST syndrome – Calcinosis, Raynaud's, Esophageal Dysmotility, Sclerosis, and Telangiectasia) that affects primarily the skin of the face, hands and feet (with possible involvement of other organs).
- ***Diffuse scleroderma,*** which has a more rapid onset and affects internal organs as well as the skin.

Telangiectasias become more numerous over time in both types of the disease, however, they are thought to occur more frequently in patients with limited scleroderma.

Although the precise factors involved in the development of telangiectasias are unknown, some experts believe that they are a manifestation of the body's attempt to increase blood flow to organ tissue with poor circulation. Thus, in scleroderma, telangiectasias may be a marker of ongoing vascular injury and failed repair.



Researchers have also identified the following facts and findings about telangiectasias in individuals with scleroderma:

- The total number of telangiectasias has been shown to correlate with disease duration; whereas the longer you have had the disease, the more telangiectasias can develop.
- The number of telangiectasias also correlates to the risk of developing pulmonary arterial hypertension.
- Telangiectasias are associated with the presence of the *centromere antibody,* (an antibody to a portion of the chromosome that is active in cell division).
- One study found that body image dissatisfaction was higher in scleroderma patients with numerous telangiectasias.

Dermatomyositis

In individuals with dermatomyositis, telangiectasias are typically found in sun-exposed areas, such as the V-shaped area of the neck and chest or in a "shawl" distribution over the shoulders, arms, and upper back.

Lupus

Telangiectasias of the nailfolds also occur in individuals with lupus and correlate with systemic disease activity and Raynaud's phenomenon. Telangiectasias may also be found on the edges of lesions of discoid lupus.

What should I do if I have telangiectasias?

Dermatologists commonly evaluate and treat patients with telangiectasias. Because the skin can function as a window to internal health, the dermatologist will determine the cause of the telangiectasias and initiate the appropriate work-up.

What can you expect from a doctor's visit for telangiectasias?

The dermatologist will perform a history and physical examination. He or she will also review your medications and effects of associated health conditions.

Prevention and Treatment

Can telangiectasias be prevented?

Activities that trigger blushing or facial redness can worsen telangiectasias.

Although triggers are different for each individual, common culprits include:

- ultraviolet radiation
- heat
- cold
- strong wind
- alcohol
- smoking
- hot drinks and food
- spicy food

Products that result in irritation of the skin, such as abrasive cleansers, can also worsen telangiectasias.

The following tips are recommended:

- Protect skin from the sun with sunscreen, sunglasses, and hats
- Use mild cleansers
- Minimize exposure to extremes of temperature
- Avoid application of topical steroids

Are there treatments for telangiectasias?

Although telangiectasias themselves pose no adverse health risk, there are treatments available to improve their appearance.

Cosmetic Camouflage

Cosmetic camouflage is a technique using topical creams or powders to conceal conspicuous skin conditions.

Flesh-toned cover-up can immediately hide mild telangiectasias. For more prominent telangiectasias and facial redness, a slightly green-tinted foundation or moisturizer can neutralize the color.

These compounds are cost effective and readily available. Look for products with the terms "redness concealer," "redness relief," or "redness solutions."

Laser therapy

This form of therapy uses a specific wavelength of light to selectively heat haemoglobin (the protein responsible for the red color of blood) and seal dilated blood vessels. Superficial facial telangiectasias are amenable to laser treatment.

In scleroderma, due to thickened collagen fibers, telangiectasias are more resistant to laser therapy, but can be effectively cleared with multiple treatments.

However, this does not prevent new telangiectasias from forming and subsequent treatments may be required to maintain the desired effects. Individuals considering laser therapy for telangiectasias should be aware that insurance plans deem these treatments to be "cosmetic" and do not yet cover this expense.

Electrodessication

Electrodessication entails the insertion of fine needle into the blood vessel. An electrical current is then applied, which seals the vessel. This treatment may be helpful for simple facial telangiectasias; however, it has a higher risk of scarring compared to laser therapy.

Support Groups in full swing

Support Groups are undoubtedly the lifeblood of Scleroderma Victoria and once again they are back in full swing with get togethers across the State.

The photo right shows our North Eastern suburbs group at a recent luncheon. Pictured from left relaxing on the couch are: Vanessa, Jane (NE Support Group Leader), Ingrid, Monica, Sylvia and Sheryl.

Check the Scleroderma Victoria Facebook page for notices on where and when these meetings are held.

It's an excellent way to get to know others who suffer from Scleroderma and how they cope on a day to day basis.

Friends and family are more than welcome too!

Grand lunch for a *grand* day!

Scleroderma Victoria's World Scleroderma Day luncheon is on again, this time falling on the day itself – Saturday, June 29.
Once again, The Grand Hotel in Richmond has agreed to hold the luncheon striking a deal in which they return a substantial amount of the entry price to Scleroderma Victoria.
The lunch will be held at the hotel from 12 Noon on World Scleroderma Day. Mark it in your diaries now, or better still, phone Kirsty at The Grand on 9429 2530 and make a booking. It's strictly limited to 60 participants.
The cost is $125 a head which includes a five-course lunch with matching wines. The Grand is renowned for its Italian food.
Just to get your taste buds working here is the menu for the big day.

CANAPE
Suppli – porcini mushroom risotto balls with truffled aoli
Vegetable skewer - GF

ANTIPASTO
House made gnocchi with burnt butter & sage
Capsicum salad – GF

SECONDO
Roast free range cornfed chicken with roasted artichokes and mashed potato
Vegetarian Risotto - GF, V

DOLCI
Lemon Tart with Vanilla Cream
Fruits and sorbets – GF

TEA & COFFEE INCLUDED

There will be a silent auction and special guests for the day which will go from Noon until 4pm.

Don't miss out, book in early to avoid disappointment.

Ahoy there yon pirates in Tassy!

After last year's successful fundraiser 'Sunflowers for Scleroderma' in Tasmania, a Pirate-themed fundraiser is already underway to be hosted in August 2019 to again raise both awareness and funds for Scleroderma Australia.

This Pirate Treasure Hunt fundraiser is promising to be a fun-filled evening full of swash-buckling excitement and entertainment – including the main competition to complete a range of challenges to unlock a treasure chest full of prizes!

A range of fantastic prizes have already been donated with many businesses and individuals generously contributing to make this event a success, including a three-hour wilderness cruise, a Nikon sports adventure camera, swim centre passes, assorted vouchers, and a Mimco clutch bag & rose gold necklace.

These are only some of the prizes available up for grabs in the raffle and within the Pirate Treasure Chest.

In addition to the treasure hunt challenge and raffle, a range of pirate-themed activities have also been organised including pirate bingo, best-dressed costume competition, and karaoke.

After the success and enjoyment of last year's fundraiser, this year's themed event is expected to be even more popular – people are already being encouraged to gather their crew together and prepare to set sail so as not to miss out!

Shire gives us $1000

The Mornington Peninsula Shire has donated $1000 to our Mornington Support Group.

The funds were made available through the Shire's Disability Community Capacity Buildings Projects.

The money will subsidise Support Group meetings and go towards the hire of buses to Scleroderma Victoria events for existing and new group members.

Donations to Scleroderma Victoria

Mornington Peninsular Shire
Pakenham CWA
William Angliss Charitable Trust
Marlene Anthony
Taina Archer
Damian Conroy
Melanie Edwards
Corrie Hemming
Heather Hollaway
Julian Inglese
Donald Irwin
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Mari-Alexia Phillipas
Dr Robert Sward
Marlene Swinstead
Marcus Walford
Nicholas Walford
Jill Yu

Increased production of Nestin Protein Involved in PAH development

- a new study suggests

by Alice Melao, Scleroderma News

Increased production of a protein called Nestin by cells lining the blood vessels in the lungs may contribute to the development and progression of pulmonary arterial hypertension (PAH), a study suggests.

Additional studies are still warranted to further understand whether targeting Nestin might represent a viable and effective approach to treating this life-threatening disease. The study, "Endothelial cells are a source of Nestin expression in Pulmonary Arterial Hypertension," was published in the journal PLOS One.

PAH is a serious respiratory disease, characterized by remodelling and thickening of blood vessels in the lung, potentially leading to blockage of blood flow and severe impairment of the heart.

The proliferation and modification of cells lining the blood vessels (known as endothelial cells) are critical contributing factors in the progression of the disease. However, the underlying biological mechanisms involved in these processes are still poorly understood.

A previous study showed that pulmonary arteries from PAH patients have increased levels of a protein called Nestin. This protein is known for its important function in promoting self-renewal and proliferation of stem cells. Still, its role in PAH remains unknown. Now, a team led by researchers from Virginia Commonwealth University evaluated the role of Nestin in experimental cells and lung tissue samples from PAH patients and healthy volunteers.

Results showed that Nestin was in general more abundant in PAH samples. In particular, Nestin was found to be mainly present in endothelial cells close to vascular lesions. This finding was further confirmed when the team analysed Nestin levels in the lungs and pulmonary arteries of mice with induced severe pulmonary hypertension.

Additional experiments with isolated lung endothelial cells from healthy rats revealed that Nestin levels were particularly higher when these cells were in proliferating conditions and undergoing cell expansion.

Supported by these results, the team reanalysed PAH patients' lung tissue samples to see whether Nestin was associated with the abnormal expansion of endothelial cells in vascular lesions. They found that Nestin was mainly accumulated close to the lesions within proliferating cells.

When researchers forced the production of Nestin in rat endothelial cells through genetic engineering methods, a significant increase in cells' proliferation rate was noted. Also, increased production of Nestin promoted the formation of new vessels (a process called angiogenesis) in an experimental setting.

"Our data suggest that in PAH, the abnormal endothelial cells undergoing aberrant proliferation and angiogenesis in the lung vascular lesions are a source of Nestin," researchers said. In turn, "elevated Nestin expression likely contributes to unchecked pulmonary vascular proliferation and angiogenesis."

Several questions remain regarding the underlying mechanism that contributes to the enhanced production of Nestin in the particular setting of PAH. Therefore, more studies are needed to provide insights on Nestin involvement in PAH development, and to determine whether Nestin could be used as a therapeutic target.

SCLERODERMA VICTORIA DIRECTORY

Incorporation No. A001798A
ABN 45 674 166 348
All correspondence to: Scleroderma Victoria, St. Vincent's Hospital, 41 Victoria Parade, Fitzroy, Victoria 3065 or phone 9288 3651
Website: www.sclerodermavictoria.com.au
Email: enq@sclerodermavictoria.com.au

Our office is attended Tuesday mornings from 10am to 12 Noon. Phone messages may be left on our message service.

MANAGEMENT COMMITTEE

COMMITTEE MEMBERS

NEWSLETTER EDITOR

VICTORIAN SUPPORT GROUPS

Co-ordinator - Judy Browning (Bendigo)
E: judybrowning2@bigpond.com (03 5437 3305)

Bendigo As Above.

Albury/Wodonga Kaye Dando
E: kcamry54@hotmail.com (0459 234 690)

Ballarat Paul Hadfield
E: paul.hadfield67@bigpond.com (0484 342 925)

Geelong Janelle Blight
E: dermageelong@gmail.com (0413 046 110)

Gippsland Corrie Hemming
E: corriehemming@bigpond.com (0400 331 829)

Inner South Eastern Suburbs Lynette Slade
E: l.slade@bigpond.com (9576 4066)

Mornington/Westernport
Annette Gaiardo PM Online Group
15 Nursery Lane, Tooradin 3989 (5998 3553)

Narre Warren Barbara Handasyde (0457 123 447)

North Eastern Suburbs Jane Rhyder
E: janerhyder@optusnet.com.au (9444 6900)

Seymour Heather Hollaway
E: hthr_hollaway@yahoo.com.au (57842322)

Western Suburbs Yvonne Hoffmann
E: hhoffmann@netspace.net.au (9743 1009).

TASMANIA

Hobart Sally Westbury E: sally@westbury.name (0414096881)

Contacts but no support group –

Margery Cairns E: Geelongmjcairns22@gmail.com
Cheryl Rosini - Maryborough: 03 5461 1356
Sharon Egberts - Mildura 5024 2337
E: shajoy54@gmail.com
Launceston - Margaret Murray 6391 8561