SCLERODERMA & RAYNAUD'S UK

SRUK NEWS

#RareTogether

Read the first in our new series of readers stories

30 Years of Research

We explore our impact on the research landscape over the last 30 years

Doc Spot

Your questions answered by our medical professional

Check out our new shop range

2017-2018 Highlights

April

10 runners in the London Marathon - £20,000 raised

Attended FESCA meeting – bringing together Scleroderma organisations from across Europe

May

SRUK Information Stand, Ninewells Hospital, Dundee

June

Ran first ever mobile clinic, working with Dr Francesco Del Galdo in Leeds, to test for Primary & Secondary Raynaud's. Over 600 people were tested in 7 days

Professor Chris Denton gave the inaugural Anne Mawdsley Memorial Lecture, hosted by Fiona Bruce MP and Professor Dame Carol Black

Second round of SRUK Research Grants awarded £144k new grants and £153k towards on-going grants

July

Award winning garden at Hampton Court supported SRUK

Sept

The second Annual conference received very positive feedback with 100% saying they would attend another conference

Aug

SRUK Information Stand, Great Ormond Street Hospital, London

Oct

Secured 9 min slot on BBC2 'Trust Me I'm a Doctor' and a feature on BBC's R4 Gardener's World

Nov

Attended Rare Disease Forum meeting in Birmingham to highlight challenges faced by rare auto-immune conditions

Dec

Our materials were awarded the health and social care quality mark for reliability and trustworthiness.

Jan

SRUK Information Stand, Queen Alexandra Hospital, Portsmouth

Feb

Since Launch, over 6,000 people have taken part in our online Raynaud's test. 96% of participants were advised that they might have Raynaud's

Attended World Scleroderma Congress

The RAIRDA report, Reduce, Improve, Empower, published setting out recommendations to tackle the challenges faced by people living with rare auto-immune conditions

We highlighted that nearly 50% waited more than three years to be correctly diagnosed

Mar

Joint meeting with the UK Scleroderma Study Group

Event Calendar 2018-19

NOVEMBER

Big Give

27th - 4th December

theBigGive.org.uk CHRISTMAS CHALLENGE 2018

FEBRUARY

Raynaud's Awareness Month

Cosy Up with a Coffee for Raynaud's Awareness Month

Wednesday 22nd Canary Wharf Tube collection

Thursday 28th February Rare Disease Day

MARCH

Launch of Research Strategy

Sunday 24th London Landmarks Half Marathon

LONDON LANDMARKS HALF MARATHON

Dear Supporters

Over the last three decades SRUK, and it's two founding charities, the Raynaud's and Scleroderma Association (RSA) and the Scleroderma Society, have funded over 100 research grants, spending more than £10 million on cutting edge science.

The cumulative effect of this support over the last 30 years means that today, survival outcomes are much improved, quality of life is significantly more positive and there are more treatments available.

As a special sneak preview for our community members, we've included some highlights from the report on pages 11-14.

While we recognise that we've come so far, we know it's not far enough.

Last week we held a thought leaders roundtable, bringing together our community of people with Raynaud's and Scleroderma, clinicians, researchers and scientists to map out what we need to do next. It was a lively and very productive day with excellent presentations from members of the Community Research Panel and key Clinicians. The output from this event will inform our research strategy and what needs to be funded.

We continue to invest in research and this year we awarded funding for 4 new grants and you can read more on pages 6-7 about these projects.

The progress we're making is thanks to your support and we couldn't achieve what we're doing without you. This Christmas you could double the value of your donation. We're taking part in The Big Give Christmas Challenge. So, please consider donating to the SRUK project on the Big Give website between noon on 27th November and noon on 4th December. The value of your gift could be doubled thanks to a matched funded provided by major donors. See page 5 for more details.

We live in an age of unparalleled advances in technology. We have the connectivity to bring together some of the greatest minds from across the globe and at SRUK we believe it's only by working together that we can hope to eradicate disease and live longer, healthier lives.

Best wishes,
Sue

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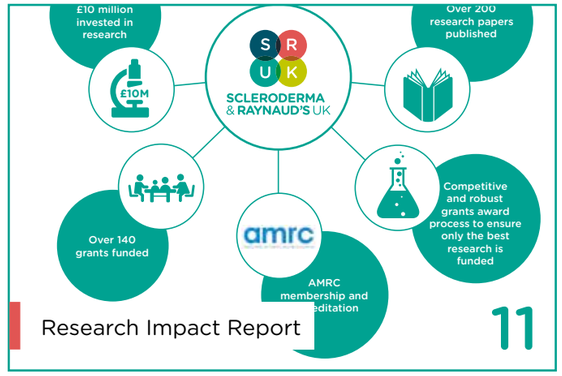
@farrsue01



Doc Spot

Your questions answered by our medical professional, Prof. Denton

What is pre-scleroderma?

(Pre-scleroderma) is a concept that can be confusing to doctors and patients but does have some value. One limitation is that there is no formal definition at the moment although relevant research is ongoing.

Like any complex long term disease, scleroderma in its various forms can take time to develop and so there will be individuals where some features that may reflect scleroderma have developed (such as Raynaud's phenomenon) but not enough to make a definite diagnosis (of scleroderma).

However, not all such cases will necessarily progress and so it is important to look at each patient individually to determine what features they have. The term is most often applied to patients with Raynaud's phenomenon, positive antinuclear antibodies (ANA) and abnormal nail fold capillaries.

Studies suggest that a substantial number of these cases (but not all) will later develop scleroderma or a related connective tissue disease over the next 5 years.

From a practical viewpoint when a patient is given this label or diagnosis they should ask what features they have so that they understand it better.

Can the contraceptive pill, contraceptive implant or injection have impact on Raynaud's? It has been suggested that my Raynaud's may have been caused by taking the pill and wonder about the association with hormonal contraception.

Female hormones can affect blood vessels and it is often observed that Raynaud's severity improves in pregnancy when hormone levels are more stable and generally oestrogen levels are higher.

Although contraceptives that alter oestrogen or progestogen levels may impact on Raynaud's, this is not always the case. If RP develops or clearly worsens within a few weeks of starting or changing a hormonal contraceptive this is a good clue that stopping or changing this may be worthwhile.

I suffer from what used to be called CREST. I have several difficult symptoms but recently I found I had a longish lump on my lower leg (just as if I had been wearing tight socks, but I haven't). It is not painful and I only discovered it by accident. I have shown my doctor and she wondered if it was to do with thickening skin due to my condition, but she is basically a bit baffled.

In scleroderma of all types there is thickening of the skin and this occurs in distinct patterns and distribution depending upon the subset or type of systemic sclerosis or localised scleroderma.

Lumps on the leg in a patient with limited cutaneous systemic sclerosis (the type of scleroderma also called CREST, especially in the USA) could be due to a localised area of scleroderma developing or to calcified material being deposited under the skin (calcinosis).

It is also important to consider that it might be an entirely unrelated process from scleroderma. I am sure these are things that your doctor has considered but sometimes further tests such as an X-ray, ultrasound scan or biopsy (small sample taken from the lump) are needed and you should discuss this with your doctor.

What is the cause of Raynaud's?

Raynaud's phenomenon results from excessive spasm of the blood vessels in the extremities (e.g. fingers, toes, ears etc.). This spasm is triggered by cold or emotional stress, which can be a normal response.

It causes symptoms in up to 10 million people in the UK, so is very common, but usually not associated with any other disease (primary Raynaud's phenomenon).

It probably is caused by complex alterations in the balance of chemicals that narrow blood vessels and those that widen or relax the muscle in the blood vessel wall. Other mechanisms may include an increased sensitivity of nerve endings in the blood vessel wall or damage to the small vessels.

I have Raynaud's and my hands swell up, am I developing scleroderma?

Most people with Raynaud's do not have scleroderma - and swelling of the hands can be normal in Raynaud's that occurs in otherwise healthy people (primary Raynaud's).

However you should discuss this with your doctor as you may require tests to look at the blood vessels around the finger (nailfold capillaroscopy) and blood tests for autoantibodies. If these tests are normal you are very unlikely to have scleroderma.

Can Raynaud's affect the tongue? My tongue swells up and goes blue at times and I find it difficult to talk. It feels as though my tongue is shrinking.

It is certainly possible for the tongue to be affected and this seems to occur in both primary Raynaud's or in association with connective tissue disease.

Blood vessel spasm in Raynaud's can involve any of the vessels that respond to or regulate temperature and this includes the tongue. Raynaud's therapies may improve these symptoms.

If you have a question you would like to ask Professor Denton, or have a comment on some of the answers given, please email info@sruk.co.uk

Or you can connect with us on Twitter, Instagram and Facebook.

/WeAreSRUK

Four New Research Grants

Fatima Sulaiman
Head of Research and Services

We are committed to investing even more into the most ground breaking and innovative research so that our community can feel the benefit of research sooner.

Your donations help us to make this difference and allow us to maintain the crucial investment into research.
As part of this commitment to research, we are very pleased to announce that we have awarded four exciting new grants in June 2018.

Here is an exclusive look at the four grants that we have awarded this year.

"Profiling of subpopulations of monocytes and monocyte-derived cells in scleroderma to identify targets for biomarker, patient stratification and therapy"

Dr Angela Tam (Royal Free Hospital/UCL)

Fibrosis (a key feature of scleroderma) is characterised as the thickening of connective tissue, which has very serious complications in systemic scleroderma for large organs such as skin, lungs, heart and the GI system.

This research team will be looking at monocyte cell populations in early scleroderma patients, as well as patients for whom the condition is more established. Monocytes are large white blood cells and comprise part of the immune system response. The research is focused on a particular type of monocyte, which belongs to the non-classical sub population, and has been implicated in fibrosis. Recent research has shown that these particular monocytes are found at higher levels in systemic sclerosis and has a correlation with the level of skin severity.

This work will be able to determine whether these sub populations are also present in patients

with early scleroderma, which could lead to the identification of therapeutic targets. The team will be profiling the monocyte sub population, e.g. determining the cytokine expression profiles, in order to determine the impact of targeting these cells as part of a therapeutic intervention.

"Elicitation of expert prior opinion for a future Bayesian randomised controlled trial for Juvenile Localised Scleroderma (JLS)"

Dr Clare Pain (Alder Hay Children's Hospital)

Mycophenolate mofetil is a potential alternative treatment that is also used to suppress the immune system and has been typically used in organ transplant procedures. It lowers the immune system just enough to ensure that the host body will not reject the donor organ therefore ensuring the success of the process. There is currently some evidence to suggest that mycophenolate mofetil can have the same efficacy as methotrexate in treating JLS, but with reduced toxic side effects.

As the condition is so rare, a traditional clinical trial to test the effects of these two drugs against each other will not be feasible as these require larger cohort numbers. In order to accelerate progress and bring a potentially beneficial treatment to children with JLS sooner, SRUK have funded the setup of a critical expert meeting and panel to design a novel clinical trial that does not rely on large numbers.
In order to carry this trial out, it's crucial that there is consensus amongst clinical experts and leaders in the design of this novel clinical trial. This meeting will create consensus and consolidate expert opinion, thus forming the basis of a trial that will maximise benefit to a small patient group within a shorter period of time.

"Preclinical pathologic signs of SSc in Raynaud's patients at risk of scleroderma"

Dr Francesco del Galdo (University of Leeds)

Raynaud's Phenomenon, in contrast to scleroderma, is fairly common in the UK with up to 10 million diagnosed. Although it is a common condition, a very small sub population of people with Raynaud's will exhibit more severe symptoms, which could be the precursor to an autoimmune condition such as scleroderma.

While the link between Raynaud's and scleroderma has been known for a number of years, we still don't know why this link exists or what the early signs for progression into scleroderma are.

Dr del Galdo is working with a group of Raynaud's patients who have signs of being at risk for developing scleroderma to determine what factors are at play.

Over 3 years, blood and skin samples will be collected from this group of people at 6-month intervals and analysed to determine the presence of biomarkers (such as antibodies and proteins) associated with scleroderma. While research has taken place into identifying biomarker association with scleroderma, this analysis has only taken place in patients who have been advanced in the progression of their condition. This study takes a novel approach by analysing biomarker association before the onset of scleroderma.

"Using microneedle patches to administer novel anti-fibrotic peptides in order to treat Scleroderma"

Dr Richard Stratton (Royal Free Hospital/UCL)

Skin fibrosis can be a severe and life limiting complication of scleroderma, with an impact on quality of life. It can be progressive and painful leading to permanent ulcers and restriction of movement.

Dr Richard Stratton, based at the Royal Free Hospital in London, will be carrying out a preclinical study investigating the efficacy of a novel treatment for the lesions caused by skin fibrosis.

Working closely with a biotechnology company, Dr Stratton is pioneering the usage of microneedle patches which will deliver a novel therapeutic directly into the skin lesions.

Macrophages, white blood cells that are a part of the immune system, have been implicated in playing a role in the progression of fibrosis through recruitment of other factors into the lesion sites. This may be perpetuating the formation of the lesion and leading to severe presentation. Dr Stratton has determined a peptide that can counteract the effects of macrophages and therefore prevent the progression of the skin lesions.

A significant downside to current available treatment is the toxic side effects that patients often experience. By administering the therapeutics directly to the skin lesions, the toxic side effects can be avoided as the treatment will only affect the lesions and not circulate throughout the body. This treatment has the potential to revolutionise treatment for both localised and systemic scleroderma, and will ultimately result in a significantly better quality of life.

Cosy Up this Christmas?

We've enclosed a letter and guide to help you get started

This Christmas we want to get people to talking about Scleroderma & Raynaud's, so that anyone who needs our services, knows that we are here to help and how they can get in touch with us, so that they are not alone. We know that Christmas is a time of giving and we hope that by telling people about us and raising awareness it will inspire people to donate to SRUK.

To help you suggest holding a Cosy Up at work, or your place of worship, we have enclosed a letter for you to introduce SRUK. We've also enclosed a guide to show how easy it is to raise awareness and funds with our new Cosy Up pack (details of how to order your pack are on the right of this page). By hosting a Cosy Up event you can raise awareness about the conditions, helping to remove public stigma and provide a space to discuss the conditions.

This letter is a way of introducing Scleroderma & Raynaud's UK to your local community and asking them to think of SRUK this Christmas.

Angela hosted a Cosy Up in collaboration with her Church, in memory of her friend Mary who had lived with Scleroderma and together they raised an amazing £689.

We know churches have many activities during the festive season such as concerts, collections, nativities, fetes and bake sales, we are hoping that this letter will help you share the causes closest to your heart with your place of work, faith or sport.

If you have questions about Cosy Up, would like more information about the conditions. To share or a Cosy Up pack, please get in touch and we will help in any way we can.

Our new Cosy Up pack is now ready - just in time for Winter. Why not have some fun raising awareness and funds at the same time. Please find your Cosy Up guide enclosed. To order a Cosy Up pack with a collection box, balloons, banners or bunting email fundraising@sruk.co.uk or call 020 3893 5993. Remember you can Cosy Up anywhere; at home, work or your place of worship! **#CosyUp**

London Landmarks Half Marathon

We are so excited to announce our new partnership with the London Landmarks Half Marathon event.

The event, only in its second year, is the most popular Half Marathon the UK has to offer. With ballot places being oversubscribed, the only way to take part now is with a Charity spot.

The route takes you along much of the prestigious London Marathon route but with half the distance to run. If you were caught up in the excitement on the London Marathon and wanted to get involved but fear taking on the 26.2 miles, this Half Marathon is the perfect taster, and an opportunity to take your running to the next level. Join #TeamSRUK and help us raise awareness and vital donations for SRUK. Secure your place for only £10 today (Minimum sponsorship £350) by calling 02038935993 or emailing shauna.creamer@sruk.co.uk

New Cosy Up for a Coffee pack now ready!

Grab your free Cosy Up pack now! By calling 02038935993 or emailing fundraising@sruk.co.uk. Remember you can Cosy Up anywhere; at home, work or church this winter! #CosyUp

Jane Potter

When did you first notice you had Raynaud's?

I regularly play football and during 2011, I noticed that my hands, nose, chin and feet were turning white and blue and were very painful. I saw the doctor in May 2011, had a number of blood tests and the results came back with a positive result for Raynaud's and Systemic Scleroderma.

How did the condition affect your life?

My hands, feet and face become so cold quite easily and it feels that they don't belong to me, it is very painful. I should stay out of the sun, wrap up and avoid any cold, wet environments.

Winning a football cup

How was your sport affected by your condition?

I still continue to participate in football and running, however I need to be mindful and plan for my condition, e.g. having a snood, gloves and handwarmers, extra socks and feet warmers and gels in my boots or running shoes. I play through the pain.

When diagnosed, what changes did you make to your lifestyle?

My specialist advised me to avoid outside sports and getting cold and wet – but I was not going

Jane doing Go Ape

to give up the sport I love and have adjusted by wearing more layers and using warming aids and painkillers. I get changed very quickly out of my sports gear into dry warm clothing.

What exercise do you partake in now and how does it help your condition?

I play football, go to the gym and run – the more I do the better I feel. It stretches me and exercise is good for mental stimulation – it is my way of not letting this be detrimental to what I love to do.

What top tips would you give to others with Raynaud's who love sport?

Stick at it and find ways to keep warm, prepare for the conditions and have a change of clothes and get a hot drink and get warm as soon as possible afterwards.

Playing in the cup

Would you recommend others who experience Raynaud's to participate in sport to help manage their condition?

Absolutely, it helps with circulation which reduces the pain, don't give in.

What is your next biggest sporting challenge?

In 2019 my aim is to run a half marathon and walk the Inca trail.

Carrie Steen's Story

It all started when I was two and my localised scleroderma presented as my first lesion appeared on my right leg with more quickly appearing on my back.

My parents quickly took me to the doctors who were told it was a skin fungal infection, so I was given medication for it. After the first bottle with no improvement I was then given a second, still with no improvement my parents looked up the medication and realised it could be harmful so stopped giving it to me and I was eventually referred to a local dermatologist. It was then that I was diagnosed with localised scleroderma.

With new lesions appearing, my parents were concerned for me. The local dermatologist just wanted to take the approach of doing nothing in order to watch it and see how far it spread. This was the case till I was 11 when my dad came across SRUK. By this time my localised scleroderma was getting worse with new lesions appearing affecting the fat and muscle leaving indentations. SRUK told us that there was help out there and a specialist unit at The Royal Free Hospital in London where a team deal with scleroderma and there was treatment for it.

Armed with this new information we went back to my dermatologist to ask to be referred to the specialist team. The dermatologist was reluctant to do so but after a long conversation he did. I was 12 when my first appointment came through. By this time I had started puberty which accelerated my condition.

The team at the Royal Free were amazing, they took the time to listen and investigate. At this first appointment it was suggested that I start a course of methotrexate and steroid infusions to try to slow down and even stop the localised scleroderma from getting worse.

I have to admit being 12 this prospect was really scary, hearing all the possible side-affects and being immuno-compromised was a daunting prospect. The medical team were fantastic with me and my parents, and so treatment began.

Treatment was tough and I was tired all the time and seemed to catch any bug and virus that was going around. Consequently, I had time off school, I was also being bullied which made school life harder. With regular visits to the Royal Free hospital, the methotreaxate was upped till I reached a therapeutic dose which worked for me and after time the localised scleroderma seemed to halt.

I was kept at this dose for quite some years as every now and then a lesion would get a bit bigger or a new one would appear, but I'd hate to think how bad it could have gotten without this medication.

By the time I was 20 I had come off the medication since the localised scleroderma was in remission.

It stayed this way till I was 23 when I became pregnant, I had already been warned that pregnancy could cause a flare up of my condition which it did, but - as I was pregnant - not a lot could be done at the time. I went on to have a healthy baby boy.

Knowing we wanted to have more children I decided not to start treatment again until we had another baby, and to my surprise we went on to have twins.

However, once again during my pregnancy my localised scleroderma was active. By this time I had quite a few lesions on my right ankle, leg, tummy, side, back and arm.

I have recently re-started treatment of methotrexate injections to hopefully put my condition back in remission.

The impact of research: 30 years of support

SRUK has a 30 year legacy of supporting vital research into Scleroderma and Raynaud's phenomena. Existing originally as the two founding charities, the Raynaud's and Scleroderma Association (RSA) and the Scleroderma Society, and eventually coming together to achieve more. We have made it our mission to transform outcomes for patients, injecting much needed funding into highly promising research areas.

We've come a long way since 1985, when a diagnosis of scleroderma meant that you had a 60% chance of surviving more than 5 years. Now, in 2018, a diagnosis of scleroderma does not have to be so scary. The survival rate has gone up to 85%, and even more amazingly survival post renal failure has dramatically increased from 25% to 80%.

We also now have 6 specialist centres in England, some of which are providing support to other clinics to ensure that more people are receiving the appropriate treatment and care.

As part of our commitment to ensuring that our community is at the forefront of understanding the research that is carried out by our clinician researchers, we are highlighting how research has enabled the amazing progress that has been achieved over the last 30 years.

We've focused on four key areas:

- Understanding the cause of Scleroderma and Raynaud's
- Enabling better diagnosis for Scleroderma
- Earlier detection of Scleroderma and Raynaud's
- Developing the best treatments available

As a special sneak peek for our community members, here are some highlights from the report.

Understanding the cause of Scleroderma and Raynaud's

A genome wide association study to get to the root of the problem

Professor Chris Denton, Royal Free Hospital/University College London

A team of world class scleroderma scientists at the Royal Free hospital made a breakthrough, uncovering a potential mechanism behind pulmonary hypertension, one of the biggest risk factors in scleroderma. They have spotted how certain genes may be behind blood vessel changes, potentially spotting those patients most at risk of this deadly complication.

Every single thing that a cell can do, from colouring your eyes through to conducting thoughts across your brain, depends on a detailed and complex instruction manual called the genome. There are thousands of different instructions (or genes) that make us who we are. Yet cells don't follow all the genes all the time. A nerve cell and a skin cell behave very differently, and this is because they subscribe to different parts of this instruction manual.

Over £10 million invested in research

The symptoms of a disease occur because cells start to follow instructions that they normally ignore. Sometimes this is because the cells themselves are faulty, other times it is because something, perhaps a bacteria or virus, is stressing the cell, changing its behaviour. In scleroderma, something causes the immune system to begin attacking the body, and these attacks cause damage to the cells, which defend themselves by changing the genes they use. In areas of extreme damage where repair is impossible, the cells form scar tissue.

If we can understand which genes are in play for Scleroderma or Raynaud's, we can begin to anticipate how each person's disease will behave, and begin to explore ways to intervene or even correct the problem.

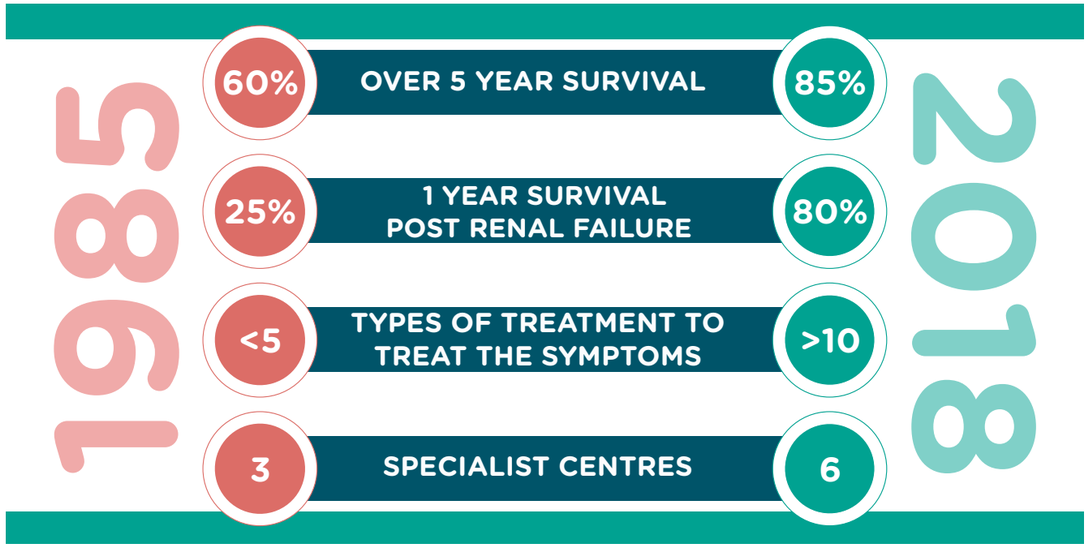
"The diagnosis, management and treatment of Raynaud's and scleroderma has improved considerably in the time Scleroderma & Raynaud's UK (SRUK), the Raynaud's and Scleroderma Association and Scleroderma Society have been awarding research grants."
Dame Carol Black, President

This is not a simple challenge, yet it is one to which Professor Chris Denton has devoted almost 25 years of research, becoming a world authority in the process. His work is improving the ability of doctors to predict how scleroderma may behave, identifying which patients will respond best to certain treatments and even helping to develop new treatments.

Using cutting-edge techniques to study the genes involved in different forms of scleroderma, Chris' team uncovered a cluster of active genes that normally help control blood vessels and may be linked tomany symptoms of scleroderma, including pulmonary hypertension, a major risk-factor in diffuse systemic scleroderma.

They discovered that the highly specialised cells that normally line our blood vessels start to follow incorrect genetic instructions, transforming them into a different type of cell without some of their specialised properties. The consequence is that the barriers to the blood vessels become very leaky, contributing to pulmonary hypertension, possibly by allowing a type of white blood cell that helps form scar tissue to interact with the vital tissue.

These major advances in understanding the mechanisms and genes behind some of the most serious scleroderma symptoms has opened up many opportunities for trialing new treatments, opportunities that Chris' team are set up to build upon.



Enabling better diagnosis for Scleroderma

Staging Interstitial Lung Disease: A world class system for clinical diagnosis

Dame Carol Black, Professor Athol Wells, Professor Chris Denton, Royal Free/Royal Brompton

A team of world-leading clinicians and scientists developed a life-saving algorithm that allows clinicians to rapidly diagnose lung fibrosis as well as an expert. Led by Dame Carol Black and Professor Chris Denton from the Royal Free Hospital and Professor Athol Wells from the Royal Brompton, this dream-team's solution is one of the most important advances in scleroderma diagnosis in recent years.

In recent times fibrosis of the lungs has become the number one cause of death in people with systemic scleroderma. For people with the most severe lung disease it is important to treat aggressively, squashing the immune response. But these treatments are harsh and can have severe side-effects. It is therefore vital for doctors to know which patients have the most advanced lung fibrosis so they can treat appropriately.

Unfortunately the standard way of assessing lung function just wasn't good enough. There is too much variability, and the complexity of achieving an accurate understanding is beyond all but the most experienced doctors.

Dame Carol Black and her colleagues' approach was to combine High Resolution Computed Tomography, a technique that takes accurate cross-sectional x-rays of the body and reconstructs an internal view of the lungs, with standard tests of the lung's performance. The revolutionary element was to create an algorithm that can draw on this information and classify a patient's degree of lung disease severity. They developed this test by working with over 200 people with scleroderma, and then evaluated their final model. Incredibly, in the trials, doctors with less than 2 years' experience were able to classify the severity of disease in a way comparable to highly experienced radiologists after only 10 minutes training. This technique has put the ability to assess a potentially fatal aspect of scleroderma into the hands of doctors everywhere and has gone on to become the international standard for assessment, quite literally revolutionizing a vital part of scleroderma medicine.

Earlier detection of Scleroderma and Raynaud's

Nailbed micro capillaroscopy: A tool for earlier diagnosis

Dr Arianne Herrick, University of Manchester
Scientists at the University of Manchester

Scientists at the University of Manchester developed an essential diagnostic tool that can pick up the earliest signs of scleroderma in patients with Raynaud's Phenomenon. Led by Dr Graham Dinsdale, they overcame significant technical difficulties to develop a long sought-after method of spotting microscopic changes to the nailbed capillaries. This work has increased the chances of diagnosing early, helping to minimise the effects of the disease for many people.

The smallest blood vessels we have in our bodies are called capillaries. In normal Raynaud's these capilaries are unaffected, but if the disease is progressing into scleroderma, it leaves its first marks on the tiny capillaries, scarring and distorting them. Spotting this damage gives doctors the best chance to get on top of the disease, minimizing the window where lasting systemic damage can occur.

Unfortunately, spotting this capillary damage reliably is hard. All people are built slightly differently, with no typical 'sign' of scleroderma. The solution is to monitor changes to the capillaries in the hand over time, but the hundreds of meters of capillaries in just one hand makes it hard to compare the same area. It is all too easy for doctors to miss the warning signs, and finding a way to help is a huge technical challenge.

This has changed. For 12 years a team of scientists at the University of Manchester, led by Dr Graham Dinsdale and Professor Ariane Herrick, have been tackling this challenge. Their breakthrough insight was to develop computer software that can weave individual images of capillaries into a detailed and highly reproduceable image.

At a glance doctors can now see the entire nailbed, and compare it with the entire nailbed from a previous date to look for the signs of the disease. Our research has unlocked a solution that is equipping doctors with the tools they need to monitor people with Raynaud's, something which could be the difference between lifelong disability and a relatively normal life.

Developing the best treatments available

A topical treatment for calcinosis

Professor Richard Winpenny, University of Manchester

Calcinosis is a debilitating problem, not just in scleroderma but in other diseases like arthritis too. This team led a revolutionary project to develop a treatment that can dissolve them.

Calcinosis is the formation of painful, hard lumps over the skin, particularly at pressure points such as joints. Unsurprisingly this can be debilitating. Professor Richard Winpenny and Professor Ariane Herrick were interested in learning what these lumps are actually made of, and crucially, whether treatments can be given to dissolve them safely, and SRUK made this a reality, continuing their long tradition of identifying promising projects at an early stage and giving them a chance to flourish.

Using advanced chemistry, the team assessed various reagents that might have the necessary properties to dissolve calcinoids, combining them with a 'nanotechnology' delivery system that can transport this chemical across the skin and into the calcinoid. The goal is now to make a topical treatment that can be easily applied and will rid a person of a painful condition.

Eating well: Dietary recommendations for scleroderma

We should all be aiming towards a healthy diet, but people living with scleroderma should take some special care to look after themselves.

Scleroderma can often lead to gastrointestinal involvement (GI) including, incontinence, constipation, gastroesophageal problems, diarrhoea, weight loss/ nutritional issues and abdominal pain. All of these can be helped or hindered by your diet but finding a diet that works for you can be a long journey of trial and error.

The recommended diet for those with scleroderma, naturally, is not the same for everyone, but there are some general guiding principles. We've outlined a few below, as recommended by the NHS 'Eating Well' guidelines.

1 **Consider eating less meat:**
A way to aid the production of healthy gut bacteria is to cut down on meat consumption. Studies have shown that those who subscribe to a plant-based diet have less inflammation and lower cholesterol than their meat eating peers.

However, followers of plant based diets aren't always healthier. Even vegetarians and vegans can eat their share of processed foods. Avoiding overly processed food can be extremely beneficial to the growth of healthy bacteria in the stomach and aid digestion.

2 **Add some spice to your life:**
Adding some spices, such as cinnamon, ginger, turmeric liberally to foods can provide some benefit to a balanced diet. Research has shown that spices such as turmeric and cinnamon have anti-inflammatory effects. Cinnamon in particular has been shown to help maintain ideal blood glucose levels. Cutting down on added sugars is beneficial in many ways, so be sure to check the ingredient labels for added sugars such as sucrose, corn syrup and glucose.

3 **Stay hydrated:**
Making sure that you are well hydrated can aid digestion and helps to flush out any lingering toxins. The NHS guide recommends drinking between 6-8 glasses of water per day, and if you don't like the taste of plain water you can try adding lemon slices.

4 **Take some extra vitamins:**
You can also consider taking an over-the counter multivitamin for supplements such as zinc, magnesium, vitamin B12 and D. This can help you get some added nutrients into your diet.

Having a well balanced diet can be the key to helping you live better each day with your condition. We always recommend speaking with your GP if you have any concerns about foods you might be eating or if you're developing digestive issues.

Exercising with Scleroderma: A few handy tips

It's well-known among people living with scleroderma that regular exercise can help keep the joints flexible. However, how the condition effects every person can change case to case.

The effects of scleroderma may affect your posture and range of movement; both of these problems will have a knock-on effect on your muscle strength. You may have problems with your diet, and so not be getting enough nutrition to keep your muscles strong. Or you may well be less active than previously.

In terms of physiotherapy and exercise, there is no set plan that will work for everyone with scleroderma. The best thing you can do is to work out what works for you, preferably with the support of a registered and trained physiotherapist.

However, there are some general guidelines that you may find helpful.

- **Maintaining your strength**
  You can maintain your muscle strength by keeping as active as possible, within the limitations of your scleroderma. You may need to perform some specific exercises to keep specific muscles strong or to re-strengthen muscles that have become weak. These exercises could well include things you do at home such as stair climbing, walking, cooking and other household jobs.

  If you are in any doubt how much exercise is appropriate for you, it is important to check with your doctor.
- **Keeping flexible**
  Some physiotherapists have noted that poor posture can add further discomfort and increase pain amongst people living with scleroderma. A good recommendation is to try outs balance and core strengthening based exercise such as yoga and pilates. If you also have Raynaud's Phenomenon, this can be a useful form of exercise to take up as it's easy to do in the comfort of your own home.

There are a number of easy exercises that you could try right now:

- Facial exercises such as massaging your face with a warm flannel and then squeezing your eyes closed. Next, wink tightly with either eye. You can also frown your forehead to wrinkle the bridge of your nose, while raising your upper lip to increase the stretch.
- Back exercises while sitting on a chair where you would then twist to one side to feel a stretch in the mid-back. You can also lie on your back on the bed with your knees bent up, and then gently roll your knees from side to side.
- Neck exercises while sitting on a chair. Tilt your head so that the ear gets nearer to shoulder while also being sure to not look backwards. You should feel a firm but comfortable stretch at the opposite side of the neck, running down towards the shoulder. You can also look over the shoulder, turning the head to the side without looking up or down. You should feel a firm but comfortable stretch on the opposite side of the neck to which you are looking.
- Wrist exercises to help extend your stretch. Place palms together and then lower them down the chest, keeping the base of the palms as close together as possible until you feel a firm but comfortable stretch at the inside of the wrists. You can also place backs of hands together and then lower them down the chest, keeping the base of the backs of the hands as close together as possible until you feel a firm but comfortable stretch at the outside of the wrists.
- Shoulder stretches while standing up. Hold a stick (e.g. rolling pin, walking stick, golf club, umbrella) in both hands with the palms facing upwards. Push one arm up to your side until you feel the stretch. Repeat for the other side. If it's easier, this stretch can be performed while sitting.

You can request physiotherapy appointments through the NHS or privately. This can be useful as physiotherapists will consider the body as a whole being instead of focusing on individual aspects.

You may need a referral from your GP to have physiotherapy on the NHS, although in some areas it's possible to refer yourself directly.

To find out whether self-referral is available in your area, ask the reception staff at your GP surgery or contact your local NHS Clinical Commissioning Group (CCG) or trust.

Short overview of presentations for SRUK Annual Conference 2018

BSR Guidelines and Annual Tests

Christopher Denton

To promote best practice care for systemic sclerosis (scleroderma) the British Society for Rheumatology (BSR) developed guidelines to help professionals give high quality care. One of the key points for effective management is that complications are found as soon as possible so that treatment can be given. Annual testing for heart, lung or other complications is recommended. The presentation will give an overview of the BSR Guidelines for Systemic Sclerosis and highlight the best approach for regular testing.

"It was great meeting up with other members. The break-out sessions were good. Learning about new treatment was very interesting." – Joanna

Top Line Results of Research: 12 treatments in clinical trials

Christopher Denton

There has been recent progress in clinical trials for scleroderma (systemic sclerosis). This has led to licensed drugs for pulmonary hypertension and digital ulcers. Treatments in routine use for skin and lung fibrosis have also been shown effective but are far short of a cure. Fortunately, there are more potential drugs in clinical trial than ever before and new approaches including stem cell therapy. This presentation summarised recent

Professor Chris Denton

"hits" and "misses" and reviewed some drugs in current trial that will soon report results. Although progress is inevitably slow, the prospect of more effective treatment for skin and lung disease is possibly within our grasp.

Biosimilars and biologics – What does personalisation mean for you?

Ian Bruce

This session focused on biologics and biosimilars and how these are being used increasingly across the CTD spectrum. In particular, Ian discussed the relative lack of success of clinical trials of these agents to date in CTDs. He also discussed how the way we think about these conditions is changing and how this may pave the way for newer trials to address the common molecular mechanisms across these conditions.

"[I loved] The opportunity to listen to experts. The commitment to live streaming is excellent as many people were unable to attend for a variety of reasons. Live streaming enables all interested parties to benefit from the presentations." – Edith

Our first Carers' Session

Face facts – A look at facial, oral and dental care including microstomia and Sjogren's

Elizabeth Price

Many patients with scleroderma will also have features of Sjogren's syndrome. Sjogren's is an autoimmune connective tissue disease that can occur alone or in combination with another rheumatic disease. It causes dryness of the eyes, mouth and skin. Simple, practical management can ease the symptoms and help prevent complications such as tooth decay.

"It was great to hear from Carol Black about research. The plastic surgeon given hope to patients. Biosimilars explained in a way for patients to understand." – Helen

Professor Ian Bruce

"Excellent show, some basic points missed but overall went from very little knowledge to good knowledge in 1 day so very pleased overall." – Brian

"Glad to have attended. In spite of possible bad progression of my systemic scleroderma, left feeling positive, and impressed by dedication of SRUK and specialist rheumatologist." – Joan

Louise Parker

My talk discussed the difficulty of living with DU and Calcinosis, the burden, treatment options, lifestyle factors and recognising signs so we can empower patients to intervene as early as possible.

Conference drinks reception

"I enjoyed listening to the research and meeting new people with the same rare disease. Thank you to all the SRUK team who organised it." – Monica

"Today's the first time I have met anybody else with scleroderma, so that's been a great thing. I live in the very far north of Scotland and feel pretty isolated with this diagnosis that I had last year. This has given me another sort of kick on that things may be not so bad, but knowing new things that I can do and people that I can go and see to help me". – Jane

"Great to be able to attend event like this. Not long been diagnosed so good to learn and go over things." – Karen

SRUK Shop

Christmas is on the way, so it's a good time to trade your old for new and buy some stocking fillers for others. You'll find some new products and some old favourites at **srukshop.co.uk** this winter.

Treat yourself or a loved one this Christmas Season

To get your order by Christmas please ensure we receive your form by the 14th December

Silver gloves

These gloves are seam free, flexible and will allow you to operate your mobile devise and visit a cash point without having to remove them. Made using 12% silver thread woven on the inside, they have light stretch for improved fit and have been designed to accommodate swollen fingers. Available in 8% or 12%. (NB 8% = silver, 12% = black).

xs, s/m, l/xl

12% = £20.99, 8% = £10.99

Hand warmers – individual pack/box

These handy warmers can be quickly activated for instant warmth whenever you are out and about. Just open the packet for 7 hours of constant heat. Please note these are disposable and cannot be reused once opened.

1 box (includes 40 warmers) = £37.99,

1 pack (includes 4 warmers) = £4.99

Warmies – unicorn/penguin

These adorable soft toys are a great way to keep warm. Ideal for children and adults alike, just pop them into a microwave to warm them up, then enjoy some lavender scented hugs! Choose between our penguin or unicorn. Not suitable for children under 3.

£11.99

12% silver long socks

It's that time of year again! SRUK Shop is proud to present its new set of Christmas cards. Choose between our stunning Partridge, "Tis the Season", or Winter in the Forest cards.

s/m/l/xl

£14.99

Christmas cards

It's that time of year again! SRUK Shop is proud to present its new set of Christmas cards. Choose between our stunning Partridge, "Tis the Season", or Winter in the Forest cards.

£3.99 (box of 10)

Intelex Pillow Boots

Treat your hard working feet to these luxurious Pillow Boots. Super soft materials for the ultimate in comfort, they're perfect for cosy nights in and keeping your feet warm in winter

One size

£14.95

HotRox Double Sided Handwarmer

So simple and easy to use, this HotRox handwarmer heats up to around 45°C in just 15 seconds, has two heat settings to choose from, and lasts up to 6 hours. It's rechargeable via USB, therefore it's really easy to keep it topped up for your next adventure.

£28.99

Slipper socks kids frozen/star wars

These super snuggly character socks are perfect as a gift for the little ones this Christmas! Heat Holders incredible warmth together with their slip resistant grip makes them perfect for wearing around the home.

£10.99

Snuggle up blanket

Relax with a Heat Holders premium fleece blanket- perfect for wrapping up warmly on those cold winter nights. With a 1.4 tog rating, its generous size and incredible softness make it a must-have item for peaceful days at home.

£24.99

Gift Wrap

Wonderfully fun wrapping paper with animals sporting christmas hats and jumpers. The wrap will give that perfect finish to your presents and will bring joy to anyone opening a present from you this Christmas. 2 each of 3 different sheets and 12 matching tags.

£4.99

SRUK shop Product Details

Christmas Items	Cost	Qty	Size	Colour
Christmas Cards: Partridge	£3.99			
Christmas Cards: Tis the Season	£3.99			
Christmas Cards: Winter in the Forest	£3.99			
Heatholders Socks Starwars	£7.00		9-12, 12.5-3.5	
Heatholders Socks Frozen	£7.00		9-12, 12.5-3.5	
Warmies Penguin	£10.00			
Warmies Unicorn	£10.00			
HotRox Hand Warmers	£28.99			
Snuggle Up Blanket	£25.00			Cranberry
Gift Wrap	£4.99			
Stocking Fillers				
12% Silver Gloves (S/M or L/XL)	£20.00			Black
Mycoal Hand Warmers (4 packs)	£6.00			
Silver Socks & Gloves				
8% Gloves Per Pair (XS or S-M or L/XL)	£10.99			
8% Fingerless Gloves (S-M or L/XL)	£10.99			
12% Short Socks Pair (S or M or L or XL)	£12.99			
12% Long Socks Pair (S or M or L or XL)	£14.99			
Postage & Packing Costs				
Postage and packing for single items	£2.99			
Postage and packaging for multiple items	£1.00 extra per item			
Total Order				
Total cost of goods				
Postage & Packaging				
To include a donation please add here				
Total enclosed				

Name	
Address	
Postcode	
Telephone	
Email	

Please return to:
Bride House,
18 – 20 Bride Lane,
London EC4Y 8EE

Get your order form to us by the 14th December to receive it in time for Christmas

For more products and information go to www.srukshop.co.uk or call 020 3893 5998

Support & Useful Contacts

Our SRUK Helpline is available to anyone who is affected by the conditions to receive support.

0800 311 2756

The helpline operates 365 days a year from 9am-7pm. This service does get busy so if you receive a voicemail please leave your name and number and you will receive a call back within 24 hours.

We currently have eleven volunteers who man the helpline on a rota basis; Amelia, Brigid, Helena, Jean, Katherine, Kim, Liz, Paula, Penny, Rosemary & Susie.

Our volunteers update their skills regularly and having external accreditation, as a member of The Helpline Partnership, means that we conform to their standards of excellence.

If you call the helpline, the volunteer that you talk to may have Scleroderma and/or Raynaud's but as we know, everyone is different and the manifestations from person to person are varied and complex. Therefore, we refrain from swapping backgrounds and symptoms but listen positively to your issues and try to help you with your particular enquiry.

We are always keen to hear from people who have an understanding of Raynaud's and/or Scleroderma who may be interested in volunteering on our helpline.

Specialist Nurses		
Rheumatology Telephone Advice Line		01225 428823
Belfast	Audrey Hamilton	0289 056 1310
Leeds	Specialist Nurse Team	0113 392 4444
Liverpool	Jan Lamb & Jenny Fletcher	0151 529 3034
Manchester	Specialist Nurse Team	0161 206 0192
Newcastle Upon Tyne	Karen Walker	0191 223 1503
Portsmouth	Paula White & Julie Ingall	0239 228 6935
Royal Brompton	Lucy Pigram	020 7352 8121 (Main Switchboard)
Royal Free, London	Sally Reddecliffe & Adele Gallimore (For Pulmonary Hypertension Enquiries)	020 7472 6354
Royal Free, London	Specialist Nurse Team	020 7830 2326
Sheffield	Specialist Nurse Team	0114 271 3086

We are working towards providing contacts at key hospitals in Wales, Ireland and Scotland please check the website for up to date information or call the Rheumatology telephone advice line (emboldened in green, listed above) with your medical query.

Local Support Contacts

Our local support contacts provide support on a local level by organising support group meetings or by being available to local residents via the phone or email. If you are interested in joining one of our local groups or wish to receive some support then please contact us and we will be happy to put you in touch with your **local support contact: 020 3893 5998**

Local Support Contacts	
Bedfordshire	Rita Boulton
Exeter	Mike Corbett
Hampshire	Tracey James
Merseyside & Cheshire	Diane Unsworth
Newcastle & Northumberland	Lindsay Wilkinson
Norfolk	Lucy Reeve
South London	Celia Bhinda
South Wales	Belinda Thompson
Leeds	Lynne Lister

Delivering better outcomes for people with rare conditions

The numbers for each rare disease may be small, but collectively they represent up to 3.5 million people in the UK. About 20% of these conditions are non-genetic, which includes rare autoimmune rheumatic diseases like scleroderma.

Currently people's quality of life is at risk through not accessing timely and effective healthcare. This lack of accurate diagnosis and effective treatments represents an enormous unmet need and a major challenge for public health. This has to change.

Current Landscape

In 2013, the UK Strategy for Rare Diseases was published, which for the first time since the establishment of the NHS, provided direction for the healthcare needs of people living with rare diseases.

There is however a concern that the focus of the strategy and implementation to date has been on rare genetic conditions, with a worrying lack of focus on the 700,000 or so people living with non-genetic rare diseases.

Getting our voice heard

The lack of focus on rare autoimmune diseases means there is a risk that the needs of people living with conditions like scleroderma are being left behind. This led to the formation of the Rare Autoimmune Rheumatic Disease Alliance (RAIRDA), bringing together clinical and patient organisations, to seek change in the system and better outcomes for our community.

In June 2016, Lupus UK; Scleroderma & Raynaud's UK (SRUK); Vasculitis UK and the British Society for Rheumatology (BSR) came together to create an umbrella body for organisations with an interest in rare autoimmune rheumatic diseases. In 2017, the British Sjögren's Syndrome Association (BSSA) became an affiliate member.

RAIRDA aims to provide a single, strong voice that will raise the profile of this group of conditions, influence policy and guide future research.

Patient Experience

Last summer, RAIRDA conducted a survey of 2,300 people living with lupus, scleroderma and vasculitis, from across the 4 nations, to assess experiences of care highlighted by the UK Strategy.

The resulting report, Reduce, Improve, Empower, published in February 2018, shows that half of respondents waited more than three years between experiencing their first symptom and correct diagnosis – and that they continue to face ongoing struggles.

The impact of having a rare condition can be devastating – one in five people have missed more than three months of work in the past year, with a similar number having to give up work entirely. The research highlights the challenges people living with rare autoimmune rheumatic diseases face. These struggles – diagnosis, accessing treatment, coping with the impacts – should not exist.

The report sets out solutions to these problems with three aims to:

- reduce delays in diagnosis
- improve the coordination of care
- improve access to broader support and service

1 in 10 respondents asked for psychological support but did not receive any help.

Almost half of respondents

46%

indicated that they waited more than three years from first symptom to receiving their correct diagnosis.

Among people diagnosed within the past three years (approximately one third of all respondents), diagnostic delays were very similar to the overall findings, with

42%

of the total waiting more than three years for their diagnosis.

A third (**33%**) of respondents routinely visit two locations for their care, with the other third visiting 3 or more sites as a matter of course. One in twenty patients visited five or more hospitals in the past year for their routine treatment.

93% of respondents reported seeing clinicians from multiple medical specialists as part of their routine treatment. Yet, among those people, less than one in five

17%

were able to see multiple specialists at a joint clinic.

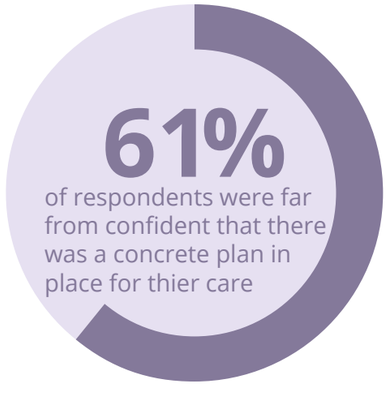
of respondents reported feeling either not very or not at all confident in their GPs knowledge of their condition

61%

of respondent's state that they are struggling to cope with their condition.

44%

of patients feel that their condition has had a negative effect on their family.



of respondents were far from confident that there was a concrete plan in place for thier care

Over a third

(40%)

say that they don't feel that they have enough information and support from the hospital in living with their condition.

of respondents reported that either they or their partner or carer had reduced their working hours as a result of their condition, and a further 20% reported that either they or a partner or carer had been forced to give up working due to their condition

theBigGive.org.uk

CHRISTMAS CHALLENGE 2018

27TH NOVEMBER 12PM — 4TH DECEMBER 12PM

This Christmas we have been selected to take part in The Big Give Christmas Challenge.

The Christmas Challenge means that your donation will be worth twice as much, as it will be match funded by private donors.

Double your donations

Your £10 donation

X2 will be doubled X2

£20 will be received by SRUK

On 27th November we're launching a one-week appeal with the Big Give. During the Big Give you can donate online to help our work to improve earlier diagnosis. Your gift could be doubled thanks to a matched fund already pledged by a couple of our major donors.

As you know, for many people earlier diagnosis can mean slowing the progress of Scleroderma, which will also save lives. Donations will be used to educate GP's and to develop digital tools for earlier diagnosis.

We would like to say a huge thank you to Jo and Tom for raising £6,165 for SRUK by taking part in Ride Across Briton which is a 980 mile cycle over 9 days with 52,000ft of climbing from Lands End to John O'Groats.

We would like to say a massive thank you to Megan and Richie for raising nearly £1,000 by taking part in the Robin Hood Half Marathon as a show of support for her dad who is living with Scleroderma.

Our Fantastic Fundraisers

A group of students from Trinity School, Croydon, successfully completed a 21-mile relay swim of the English Channel. Raising money for Scleroderma & Raynaud's UK (SRUK), the team swam for 14 hours and 8 minutes from Dover to France last week and raised an amazing £1,757

Do you want to take on a personal challenge for SRUK? Get in touch today and we can talk you through how on 0203 893 5993

Ways to support us

Firstly, we would like to say thank you. By receiving this newsletter you are helping us to continue our vital work to make a difference to the lives of people affected by Scleroderma and Raynaud's.

We could not achieve as much as we do without you and we are always striving to achieve more. If you have an idea as to how SRUK can further support the community then we would love to hear from you.

Your Magazine, Your Way

Thanks to everyone who provided feedback on the last issue of the magazine. We know that not all comments have been covered in this issue but we will be working hard to cover your feedback in future issues. Your feedback is really important to us. If you have a comment or suggestion on how we can improve future issues then call our team on 020 3893 5998 or email: info@sruk.co.uk

Donate to us through our website www.sruk.co.uk by clicking the donate button or by phoning our friendly team on 020 3893 5998 using your debit/credit card.

Your card details are not stored by the charity and the systems used to process your payment are secure.

Scleroderma & Raynaud's UK
Bride House, 18 - 20 Bride Lane,
London EC4Y 8EE

www.sruk.co.uk
Helpline: 0800 311 2756
Office: 020 3893 5998

@WeAreSRUK /WeAreSRUK

We hope you enjoyed your edition of the SRUK magazine. If you have finished with your copy then please do pass it on to a friend or your local GP surgery. Alternatively pop it into your recycling and help us look after our planet.