



## Dear Friends

Lots of exciting things have happened this year, including the joining together of the two patient societies – this gives the opportunity to build an ever stronger voice to represent patients. I have been very busy this year looking after my College in Cambridge and also doing some very exciting work for the Prime Minister, who asked me to do a review on how to help those people who are addicted to drugs and alcohol or who are obese and who are in our benefits system return to work. I hope we will be able to make some meaningful recommendations.



Happy Christmas and a joyous New Year to you all.

**Dame Carol Black**

## Update from the Clinic –

As the colder weather approaches we are getting to maximum capacity in the clinic and hospital since many scleroderma complications become more troublesome in winter, especially those related to Raynaud's phenomenon.

Our nurse led digital ulcer clinic is thriving, led by Annalyn Nunag who many of you will know from clinic, working with Louise and Joseph within the specialist nurse team. We have had some excellent overseas visitors in the clinic this year including doctors from Portugal, Tenerife and Italy. These visitors come to learn about scleroderma and undertake research and also help to raise our international profile. This also benefited from the major research workshop in Cambridge in the Summer that was linked for the first time to a patient focused pre-meeting. This allowed world class international speakers an opportunity to meet UK scleroderma patients and share experiences and learning.

On a practical side our unit has taken a lead role working with other UK scleroderma centres to develop UK Guidelines for treatment of scleroderma under the auspices of the British Society for Rheumatology. This is work in progress but we hope that the guideline will be published early in 2016 and this will help to ensure that all patients in UK get appropriate treatments and also provide guidance about the organisation and delivery of scleroderma services. It is encouraging that more clinical trials are being planned or undertaken in scleroderma for skin and lung fibrosis than we have ever had before. Many of these are being supported by pharmaceutical companies. This means that if the results are positive there is a very real chance of developing approved therapies for the disease. This is an exciting time but also a challenging one and we are deeply grateful to all of our patients that agree to take part in scleroderma trials. Within the hospital and at UCL there is now an exciting initiative to develop and expand the Institute of Immunity. Planning permission is obtained and hopefully in the next few months building work on Phase 2 of the Institute will begin. Moving the clinical service and outpatient into the Institute in 2012 was a major step forward for patients and staff and we are all excited about the next phase of this important project.

Professor Chris Denton



# Xmas Newsletter - Scleroderma Unit - Royal Free London

## Research News – David Abraham

### **Biomedical science news from the laboratory**

2015 has been an exciting year in the laboratory with a number research project coming to fruition and several new studies have begun and are underway. This newsletter gives us the opportunity to up-date you on our research activities and several important events that have taken place over the latter part of the year. These include ongoing and new research, grants that have been awarded, new staff that have join the group as well as those that have move on to new positions and meetings and conferences where our work has been presented. We have made a number of discoveries in our studies of scleroderma. We have continued our studies into exploring the genetics of scleroderma in collaboration with colleagues in Grenada, Spain (Prof Javier Martin) and with colleagues in Cologne, Germany (Prof Thomas Krieg), Paris, France (Prof Luc Mouthon and Lund, Sweden (Prof Hesselstrand), we have identified a number of new markers which are risk factors for scleroderma and also performed gene profiling in peripheral blood studies to identify distinct scleroderma disease subsets. We have continued our studies on pulmonary hypertension and we have begun to look in more detail the complex processes involving cells arise from the blood vessel and molecules which cause these cells to change their behaviour and begin to occlude the vessels. We colleagues in Boston University, USA we have been studying an important process that appears to control and drive tissue scarring and fibrosis in the skin. In another study close to home we have embarked upon a collaboration with Dr Jacques Behmoaras at Imperial College, London to study in depth aspects of the immune system in the majors forms of scleroderma. These exciting discoveries will allow us to rapidly transfer our research finding into pre-clinical models and away from the laboratory in experimental medicine studies and onto clinical trials. As you can see from the research highlights above we work productively and successfully with a number of collaborators and colleagues around the world and in the UK to advance our research into and understanding of the complexities in the development of scleroderma. Our research studies have also been presented at prominent meetings and conferences around the globe and also this year we held our biennial research workshop in workshop at St John's College in Cambridge, UK.

### **Support for the Research**

We have been awarded several research grants over the last months. These include support from the Rosetrees Trust and Royal Free Charity to Dr Richard Stratton to study scarring in scleroderma, a PhD studentship jointly funded by the Scleroderma Society and Royal Free Charity to allow research directed by Dr Voon Ong and Prof Justin Hsuan to investigate and characterise the composition and structure of the abnormal scar tissue in scleroderma. We have also attracted the interest of several industrial partners who have agreed to fund collaborative research into understanding the scleroderma disease process. We are also extremely grateful to the Raynaud's and Scleroderma Association and Arthritis Research UK for their continued support and for the research and the grants that we have been awarded this year. This funding has been essential in allowing us to pursue our biomedical research studies into the causes of Raynaud's and Scleroderma and the identification of pathways which may lead to new treatments.

### **National and International events**

A great amount of our work has been presented and prestigious national and international symposium including the British Society for Rheumatology, the American College of Rheumatology, the American Thoracic Society. In addition our International research workshop (the 14<sup>th</sup>) held in Cambridge in the summer was a resounding success, attracting over 250 participants from around the world and hosting all of the world leaders in scleroderma research. We also organised the annual translational medicine workshop, now in its 10<sup>th</sup> years at the Royal college of Physicians. This meeting has also become a regular feature in the academic calendar with nearly 150 attendees. These events are an important way by which we raise our profile and that of scleroderma throughout the biomedical research community and allow us to receive critical positive-feedback on our research endeavours.

### **Research staff and visitors to the Laboratory**

We have a number of new staff who have joined the group. Three new PhD students Rebecca Bolton (BHF), Elizabeth Evans (UK Research Council) and Daniela Gentile (Italian government) are undertaking their training rotation in the laboratory. And we have been joined by Dr Amanda Saracino who is undertaking her PhD studies with a fellowship form her home country Australia. Amanda is working on a collaborative study into localised scleroderma with Dr Cate Orteu (Royal Free Dermatology). We also have two new research staff who have joined us from Brunel University Mathew Delaney and Ms Geerthana Jeythana. Finally one of our scientist Ms Athina Dritsoula is currently visiting Yale University in the USA as part of a collaboration to study the abnormalities in blood vessels in scleroderma. Athina will bring back expertise acquired at Yale to the research group at the Royal Free.



# Xmas Newsletter - Scleroderma Unit - Royal Free London

## Scleroderma Specialist Nurses – Louise, Annalyn & Joseph

I am delighted to report that there are three of us in the team here at The Royal Free. Annalyn has now been with us 18 months and Joseph joined us at the end of last year. You may have met them on ward round, clinic or spoken to them on the advice line.

We continue to provide nurse led initiatives including the Raynaud's clinic and the digital ulcer clinic which is facilitated by Annalyn. Louise also provides support to the Dermatology service once a week for the joint clinic there which some of our patients attend. There is also a monthly joint Dermatology clinic held in The Institute which runs parallel to the scleroderma clinic to provide seamless care and to avoid repeat visits.

The Institute continues to be a great success and we have had good feedback on the improved environment and that blood tests can be carried out there too. The expansion of PITU is also working well and we have rapidly filled our capacity there for patients requiring Iloprost and other infusions. Again, the improved environment and familiar staff add to enhanced patient experience and continuity of care.

Annalyn and Joseph have started to support the outreach clinics outreach service where the team here come to other hospitals around the country to support scleroderma clinics closer to you.

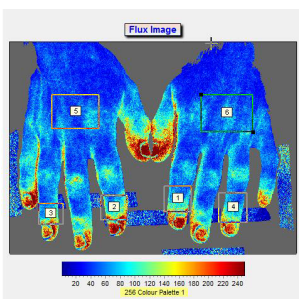
We wish you all an enjoyable and relaxing Christmas and look forward to seeing you soon.

## Microvascular Diagnostics – Dr. Kevin Howell

2015 has been one of our most interesting and productive since we began measuring the microcirculation in scleroderma back in the early 1990s...

Nailfold capillaroscopy is now established as key investigation in the diagnosis of early scleroderma, and we are one of just a very few centres that can offer this in the UK. As well as providing this test at the Royal Free, my role has expanded to deliver training and education to rheumatologists so that the technique is more widely recognised. To this end, I have lectured in Cambridgeshire, Kent and at several London hospitals on the topic during 2015.

Back in September I attended the XIII Annual Congress of the European Association of Thermology in Madrid, where I was honoured to be elected President of the Association. This is excellent news for the Royal Free, and will consolidate our position as a major stakeholder in infrared imaging and thermal biomedical research.



Our major research project this winter is an ARUK-funded study called "VALIDS", which is co-ordinated by our colleagues at Salford Royal Hospital and is recruiting scleroderma patients at a total of six centres across England. The aim is to investigate the reproducibility of the "cold challenge" of hands in water as an assessment of Raynaud's phenomenon. For this we ask each patient to undergo the test twice (on consecutive days). Warming of the hands is monitored by thermography, and also by a new device (see image) which is able to measure blood flow simply by illuminating the hand with light. If we are able to establish that these devices give consistent results, it will open up important new ways for treatments of Raynaud's phenomenon to be evaluated.

If you are a scleroderma patient who could spare us a couple of hours on consecutive days at the Royal Free sometime over the next few weeks, please contact my colleague Dipa Ghedia on **0207 317 7544** and she will be happy to provide you with more information and check if you are eligible to participate.

From the Microvascular lab at the Institute of Immunity and Transplantation, I wish you all a Merry Christmas and a Happy New Year!

Kevin



# Xmas Newsletter - Scleroderma Unit - Royal Free London

## Pulmonary Hypertension Team

As we approach the end of 2015, it is a good opportunity to reflect on the exciting developments in the PH service over the last 12 months. The Service remains as busy as ever and the New Year promises continuing expansion. At the beginning of the year, we established an outreach clinic in Margate. This is currently taking place every three months at the Queen Elizabeth Queen Mother Hospital and is set to become more frequent as the service becomes more established. We now have 10 outreach clinics in total, ensuring that a greater number of our patients can receive their care closer to their homes. Furthermore, the Trust has recently purchased a new MRI scanner and we look forward to providing a new cardiac MRI service for our patients in the new year. This test is an important tool in both the diagnosis of Pulmonary Hypertension and in the evaluation of treatments.

In May, we welcomed our new nurse, Sihle Malapela. Sihle started her career as a coronary care nurse before moving into cardiovascular risk management as a Clinical Nurse Specialist. Sihle joins us with a wealth of experience and knowledge and has proven to be a fantastic addition to the nursing team. The team continues to be supported by Jo, Sarah and Mariam– our excellent administration team who continue to keep the service running efficiently.

**Wishing you a Merry Christmas and a Happy New Year!**

## Clinical Trials News

Another year has flown by and we are already saying goodbye to 2015. We continue to see patients in the Institute of Immunity and Transplantation and this has gone smoothly especially with the positive feedback from the patients themselves. We have had a few staff changes with Dr. Anna Gill, Catherine Oduka, Karl Salazar, Joanna Lansell and Loveth Paddy joining the team. Paul Fonseca will still be working with us during the summer.

We have a number of on-going and new trials at present which include the following:

### **Scleroderma Trials:**

**Duo Registry:** The initial report confirms the early occurrence and high frequency of digital ulcers in scleroderma patients with different groups of antibodies. This study is on-going.

**Tocilizumab:** We completed the phase 2 study a year ago and we are just about to start the phase 3 study. We will be approaching suitable patients for this study

**Selexipag:** This was a new study for patients with secondary Raynaud's which started in November 2014 and ran over the winter. The drug was well tolerated, and although some patients reported benefits, overall unfortunately no net benefit was seen. It is now completed.

**Spin:** The purpose of this study is to provide better patient-centred care to individuals living with chronic diseases. We are halfway through our recruitment and have positive feedbacks from the patients that this study is needed as a platform for them to express the daily functioning issues faced living with scleroderma.

**Rise:** This is a study of medication that has already been approved for use in CTEPH (thromboembolic PAH) PAH patients. We are actively recruiting for early diffused SSC patients, this maybe a new potential drug, in Scleroderma

**Valid:** This is an exciting study for patients with Raynaud's secondary to scleroderma. We are halfway through recruitment and planning to close by the end of January 2016.

Thank you to all those who continue to volunteer to take part in our trials. These are just a few of our on-going studies. If you are interested in participating or for more information please contact us on 0207 317 7544 or alternatively you can email us on [rf.clinicaltrials@nhs.net](mailto:rf.clinicaltrials@nhs.net).



# Xmas Newsletter - Scleroderma Unit - Royal Free London

## Kim Fligelstone

Hello, my name is Kim, I have had systemic scleroderma for 28 years and am generally available on Tuesday and Wednesdays if anyone would like to phone or pop in for a chat, it's best to call or email beforehand (see details below) as these days can change. I must emphasise I am not a counsellor, I just have lots of experience of living with scleroderma, although as you are probably aware we are all affected by scleroderma in different ways, I would love to hear from you if you want to talk.

If your ever on the ward and would like a gentle massage please let your nurse know and they will contact complimentary therapy, this is a voluntary service and donations are very welcome.

The 4<sup>th</sup> World Systemic Sclerosis Congress is being held between the 18<sup>th</sup> and 20<sup>th</sup> February 2016 in Lisbon, Portugal. The patient congress program consists of interactive sessions, lectures and discussions. The main lectures will be given in English and translated into major audience languages. Also included are interactive exercises and breathing sessions, and a session for men with SSc and another for juvenile scleroderma. It only costs 35 euros for patient registration. The deadline for registration is the 31<sup>st</sup> January. [www.sscworldcongress.org](http://www.sscworldcongress.org) for booking and accommodation information. It's a great opportunity to meet other's living with SSc and to forge lifelong friendships.

The next Royal Free scleroderma local group meeting will be on Friday the 4<sup>th</sup> March 2pm to 4pm, if you have any thoughts about topics please let the specialist nurses know.

Contact details: 020 7794 0500 ext. 35131. You can email me at [kim.fligelstone@nhs.net](mailto:kim.fligelstone@nhs.net) or [kim@mediasolutions.co.uk](mailto:kim@mediasolutions.co.uk). Leave a message with Millie if I'm not around and I'll get back to you asap. Have a lovely Christmas and a Healthy and Peaceful New Year.

## Scleroderma Family Day – Saturday 14 May 2016

The 2015 Family Day was our 20<sup>th</sup> anniversary and a great success and some of the talks are on the Royal Free website - [www.royalfree.nhs.uk/services/services-a-z/scleroderma/scleroderma-family-day/](http://www.royalfree.nhs.uk/services/services-a-z/scleroderma/scleroderma-family-day/).

I look forward to seeing you on 14<sup>th</sup> May - the provisional programme is on page 6 of this Newsletter. If you are unable to attend we try to put as many of the talks as possible up on he Royal Free website.

## Newsletter

We hope you enjoy reading the Newsletter but if you prefer not to receive them please let me know and I will take you off the list.

## Fundraising

Thank you to everyone who raised money for the research unit this year – your support is very much appreciated. A big thank you goes out to the amazing marathon runners.

If you have any questions about Family Day, the newsletter, fundraising raffle or anything else, please do contact me at [p.yeomans@nhs.net](mailto:p.yeomans@nhs.net) or by phone on 01273 672686 or 020 7794 0500 x 33471 on Mondays.

**Pamela**

## Contact Numbers

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Nurses Scleroderma Nurses Help-line	020 7830 2326



# Xmas Newsletter - Scleroderma Unit - Royal Free London

Scleroderma Family Day – Saturday 14th May 2016

The Atrium, Royal Free Hospital, Pond Street, London NW3 2QG

Chair: Professor Chris Denton

## PROVISIONAL PROGRAMME

09:30 – 10:00	Registration and Coffee	
10:00 - 10:30	Welcome & Review of Progress	Dame Carol Black Prof Chris Denton Prof David Abraham
10:30 – 10:50	Emerging techniques in vascular imaging	Dr Kevin Howell
10:50 – 11:10	Heart of the matter – tackling a hard problem	Dr Gerry Coghlan
11:10 – 11:30	Vascular disease in the extremities	Dr Arriane Herrick
11:30 – 11:50	Revisiting an old idea – Factor XIII in scleroderma	Anna Gill
11:50 – 12:10	Update on the gut in scleroderma	Dr Charles Murray
12:10 – 14:00	Lunch break— discussion groups – see below	
14:00 - 14:30	The BSR Scleroderma guideline	Chris Denton
14:30 - 15:30	International Speaker	To be confirmed
15:30	Raffle	

### Lunchtime Discussions Groups /Demonstrations include

Clinical Trials	Rachel Ochiel and team
Drug information/monitoring	Gillian Murray/Jay Pang
Skin Camouflage	Changing Faces formerly British Red Cross
Massage	Mr Keith Hunt, Complementary Therapies
Pulmonary Hypertension Education/nursing	Sally Reddecliffe/Adele Gallimore
Rheumatology Laboratory	Research Laboratory Staff
Scleroderma Education/Nursing	Louise Parker/Annalyn Nunag/Joseph Cainap
Thermography	Dr Kevin Howell

