



Xmas Newsletter - Scleroderma Unit - Royal Free London



From all of us

www.royalfree.nhs.uk/services/services-a-z/scleroderma/

Christmas 2017

Dear Friends

Firstly let me wish you all a Happy Christmas and joyous New Year. This year when I attended the biannual Scleroderma Research workshop in Pittsburgh USA I was so pleased to see basic researchers wanting to get their findings translated into patient benefit as soon as possible. I know how much you wish for that too. I also know that Professors Denton and Abrahams and their teams are at the forefront of both clinical and basic research and I want to pay tribute to all they do for Scleroderma every week of the year. I am very proud of the Centre.

Keep warm and enjoy the festive season.

Warm regards

Professor Dame Carol Black



Update from the Scleroderma Clinical Clinic

The past year has continued to provide expected challenges to the scleroderma team that are inevitable in tackling a diverse disease with many potential complications and high unmet medical need. We are fortunate to have an excellent clinical team to provide expert assessment and treatment for the condition. This has benefited from growing links with other services including plastic surgery, dermatology and gastroenterology.

Our clinical work is closely linked to ongoing research projects and this benefits from the excellent facilities and infrastructure within the UCL Institute of Immunity. A new building is planned that will give much more laboratory space for immunological research. This is timely as our research projects are re-establishing a central role for the immune system in development of scleroderma. In addition the new building plans include hotel facilities that will permit even better use of the PITU unit, possible including patients who find it difficult to travel outside the hospital as required at present for courses of treatment such as iloprost for digital ulcers.

As usual we have seen excellent clinical fellows move on to other hospitals, Dr Stern is now back in renal medicine and Dr Garthwaite in respiratory medicine. We really benefit from having relevant specialists embedded in the clinic and will be working towards this more in the future. At present we have been able to increase the number of clinics where we have a dermatologist available for advice and this is a model that we plan to expand for other specialties. Dr Corrado Campochiaro has returned to Italy and will be missed but the research work that he started is now being undertaken by Dr Kristina Clark who previously worked as a clinical fellow and has returned to the unit to undertake PhD degree. We are also delighted that Ms Tani Ngcozana has returned to the unit and is now working as a senior member of the clinical trials team.

This year has been very exciting for clinical trials in scleroderma. There are several studies that will report results over the next few months including trials of nintedanib for lung fibrosis that recruited over 500 patients worldwide. The Royal Free was the second highest recruiter and this is exciting as this agent is already an approved treatment for other forms of lung fibrosis. Other trials are testing promising new therapies for skin fibrosis in diffuse scleroderma. Together with the promising results that have emerged from trials of stem cell transplantation in scleroderma these new avenues for therapy are very encouraging.

However, many challenges remain and some of these will be topics for discussion at the 2018 family Day that you are all warmly invited to attend.

Professor Chris Denton



News from the Scleroderma Research Laboratory

Research news

The last 12 months have been a very exciting time in the research laboratory. Our biomedical research has been progressing very well and there have been some major achievements which we would like to share.

- Our studies looking at disease biomarkers have identified different molecular sub-sets within scleroderma patients
- We have identified two new pathways by which fibroblast might be activated in scleroderma – the purine pathway and a metabolic pathway. These represent potential avenues for therapy of fibrosis.
- We have been studying the movement of fibroblasts when they are grown on different type of substrates and have identified that these cell behave in different ways depending on the features of the substrate

In addition, a number of our collaborative studies with colleagues have been very successful.

- We have been studying the role of B cells in scleroderma in collaboration with Prof Rizgar Mageed and Drs Taher and Jonas Byston at the William Harvey Institute/QMUL and have identified an abnormal B cell sub-set in scleroderma.
- Our work on monocytes with Dr Jacques Behmoaras at Imperial College has revealed abnormal gene signatures in scleroderma and potentially new targets for immunotherapy.
- Studies in collaboration with Prof Butler in Surgery at the Royal Free have noted the potential therapeutic benefit of adipose stem cell in tissue fibrosis.

Research funding

This year we have been awarded a number of new research grants to study Raynaud's and scleroderma. We have several research grants supported by Scleroderma & Raynaud's UK (SRUK) to explore important aspects of blood vessel/endothelial cells, immune cell (monocytes and B cells) and fibroblast function in scleroderma. These studies are led by Prof Chris Denton and Drs Alan Holmes and Richard Stratton. The Rosetrees Trust and Royal Free Charity have continued supporting our studies into the molecules, pathways and cells that contribute to scarring in scleroderma. These studies are led by Dr Ioannis Papaioannou. We also have four new PhD students who have started. The first is jointly held with Prof John Dart and Dr Jill Norman at UCL to investigate the role of cell metabolism in tissue fibrosis. The second is with Dr John Atkinson at UCB to study the way in which cells respond to the altered environment in scar tissue, and the third with Prof Julie Daniels which is a study aimed at understanding how cells respond to tissue injury. And the four with Prof Lucie Clapp investigation blood vessel damage in inflammatory diseases. Our work had also attracted interest of several industrial partners who have agreed to fund collaborative research into understanding the scleroderma disease process and to begin on the pathway to develop novel therapies.

National and International scientific meetings

Our work has been presented widely at prestigious national and international meeting including the International Scleroderma Research workshop held at the University of Pittsburgh, USA in the summer. In addition posters, oral presentations and lectures of our research studies have appeared at the British Society for Rheumatology, the American Thoracic Society, the American college of Rheumatology symposium in San Diego USA. Our annual translational medicine workshop at the Royal college of Physicians, which has now in its 12th year was a great success attracting over 150 attendees. We are now looking forward to the next Scleroderma world congress in France in early 2018. These national and international conferences are an important part of our calendar and a god way by which we raise our profile world-wide and that of scleroderma throughout the international biomedical research community.

New developments, staff and visitors.

The Unit has been leading on several new initiatives including the UCL Partners programme in inflammation and tissue repair and the London inflammation Network consortium. We have also been developing a London-wide PAH consortium- with the web-site going live in early 2018. Several new staff have joined the group. New PhD students include Jennyfer Parnasse, Ashkon Seyed-Safi, Dafni Gyftaki Venieri and Hamda Aboujassoum. Two new staff have also joined from Brunel University to undertake training in laboratory based and clinical research Sian Morris and Yaseen Siddiqui. And we have a visitor from Brazil – Dr Jones Albuquerque who is a bio-information and will help in the analysis of large data-sets.

Professor David Abraham





Scleroderma Specialist Nurses update

The scleroderma nurses remain busy supporting the outpatient clinics, PITU and the inpatient wards. During this time of year PITU is at capacity with Iloprost infusions to help you get through the winter months!

We have had some very interesting speakers at our support group this year. This has included Prof Stephen Porter who talked about dental and oral care and Richard Leigh our Lead Podiatrist who gave a talk about foot care. This is a quarterly event held at The Royal Free; if you would like to attend do get in touch for more details.

Sadly, due to funding difficulties the specialist nursing service has now fallen from three nurses to two; however, Annalyn has taken up a role with the clinical trials team so you will still get to see her on the wards and in clinic.

Thank you to those of you who continue to help with our research and audit commitments by spending time filling out questionnaires and feedback forms, it is very much appreciated.

Just a reminder – if you have queries regarding appointments, Echo and Lung functions tests please contact our coordinator Norlan in the first instance on: 020 7794 0500 Ext 32909. And for clinical or medication queries call us directly on 020 7830 2326.

We hope you have a relaxing Christmas and a peaceful new year.

Louise and Joseph

Clinical Trials News

It has been another busy year, thank you to our patients for the positive feedback on their experiences at the institute. This year we have had several new members joining the clinical trials team whom some of you may have already met including Molly, Anjinee, Stephanie, Tani and Annalyn. They have all settled well into the team. We have also sent out questionnaires to some of our patients who have participated in clinical trials in the past. We are currently still collecting these and plan to utilise the feedback to improve our service.

We have a number of on-going and new studies in Scleroderma, some like the Spin and Strike studies are simple and any scleroderma patient can take part.

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 feedback from the patients that this study is needed as a platform for them to express the daily functioning issues faced living with scleroderma.

Strike: This study focusses on the disease progression of patients diagnosed with Scleroderma. The aim is to collect data to enable us to better predict who will be likely to undergo disease progression in the future.

We also have a number of more complex studies testing potential new drugs in scleroderma for example:

1. GSK2330811 – This study looks at the mechanism of the new drug in diffuse cutaneous systemic sclerosis. We also want to check how much of the drug gets into the bloodstream and how long the body takes to get rid of it. GSK2330811 has been tested already in healthy people but this is the first time GSK2330811 has been tested in people with systemic sclerosis.

2. Tocilizumab - This research study is being carried out to compare the effects of the drug Tocilizumab (TCZ) with placebo, in patients with systemic sclerosis. It is hoped that TCZ will reduce the symptoms of systemic sclerosis.

3. Nintedanib – This study investigates how well treatment with the trial drug can demonstrate a reduction in worsening of lung fibrosis (scarring of the lungs) and how well the treatment has an effect of the skin fibrosis.

Thank you to all those who continue to volunteer, we appreciate you giving up your time to take part in our trials we know it is a huge commitment. If you are interested in participating or for more information feel free to contact us on **0207 317 7544** or alternatively you can email us on rf.clinicaltrials@nhs.net





Microvascular Diagnostics – Dr Kevin Howell

I am writing this from Vienna, where the Board of the European Association of Thermology (EAT) has gathered to discuss the plans for a thermography congress in London next July. I am currently EAT President, and will be Chair of the local organising committee of the London meeting. We are hopeful that 80-100 scientists from Europe and beyond will attend, to present work on thermography in Raynaud's phenomenon and many other medical disorders.

For this reason, 2018 is likely to be a particularly busy period, but 2017 has also been a year of achievement for our Microvascular service... Back in the summer I submitted a portfolio of the work the lab does in scleroderma to the Royal Photographic Society, and was duly elected a Fellow of the Society, along with their "Accredited Senior Imaging Scientist" qualification. At this time, I returned to the committee of the "Physiological Measurement Special Interest Group" of the Institute of Physics and Engineering in Medicine. Whilst contributing to these organisations does involve some time commitment, we feel it is very important that the Royal Free scleroderma microvascular service is at the forefront of policy-making in the physiological measurement sciences, and maintains a strong profile.

This year marks 25 years since I joined the scleroderma unit, then headed by Dame Professor Carol Black (although back then "Dr. Black"). I'm sure many of you will remember those early years of our unit, in rather cramped and uncomfortable conditions on the lower ground floor. We have come a long way since then, and are now so fortunate to have the excellent facilities of the Institute of Immunity and Transplantation at our disposal. This is in no small part due to the drive and enthusiasm of Carol and latterly Prof. Chris Denton, who have constantly strived to develop and improve every aspect of the scleroderma service.

Here's to the next 25 years...and I wish you all a Merry Christmas and Happy New Year! **Kevin**

Kim Fligelstone

We also have a number of more complex studies testing potential new drugs in scleroderma for example: Hello, It's 30 years since I was diagnosed with scleroderma and I'm generally available on Tuesday and Wednesdays for phone calls or if your local & want to pop in for a chat it would be lovely to see you, it's best to call or email beforehand (see details below) as these work days often change. I must emphasize I am not a counselor, just 30 years experience of living with scleroderma, although as you are probably well aware we are all affected 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 the complimentary therapy department, this is a voluntary service and donations are very welcome.

The next **Royal Free scleroderma local group meeting** will be on Friday the 23rd March 2pm to 4pm, the speaker will be Consultant Dermatologist Amanda Saracino.

The 5th World Systemic Sclerosis Congress will be held in Bordeaux 15th-17th February <http://web.aimgroupinternational.com/2018/patientcongress/index.html> the link takes you to the menu where you can access the programme, register and find Hotel information.

Raynaud's Focus Group

Join a closed patient focus group for a forthcoming Raynaud's/Botox study

<https://www.facebook.com/groups/1469365719825976/>

Contact details: 020 7794 0432. You can email me at kim.fligelstone@nhs.or 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. **Kim**





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Scleroderma Family Day – Saturday 19 May 2018

The 2017 Family Day really well and received very positive feedback. We look forward to seeing you on 19th 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 on the Royal Free website.

Newsletter

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

Raffle Tickets

Please find enclosed with the Newsletter a book of raffle tickets. Money raised from the raffle is tremendously helpful for the unit. If you do not wish to buy raffle tickets please discard them, you do not need to return unused books.

If you have any questions about please do contact me at p.yeomans@nhs.net or by phone on 01273 672686 or 020 7794 0500 x 33471 on Mondays.

Pamela Yeomans

Fundraising

Lynn Steblecki's book 'Prizewinner' is now out with a donation from each sale going to scleroderma research.

News from Nikki Whitehill.

This year has been a very encouraging year for raising vital funds for the unit via Just Giving. A huge thanks to the Sarah, Sue and Adrian for their cross country run, Jen with her sponsored hair cut and to the family and friends of Kevin East, who raised an amazing £2000, at their first and now annual Golf Event. On 15th March Ben Milan (benmboxing@gmail.com) is organising a boxing match, in memory of his Dad Chris. 28 February is Rare Disease Day and the theme is 'Research', please follow Nikki's blog for updates raynaudsscleroderma.blogspot.co.uk – **Nikki**

If you would like to raise money for us please contact

Pamela

Contact Numbers

The Royal Free have a new direct line to the rheumatology department - 020 375 82042. There is a prompt to press 1, 2, 3 etc - I have added the prompt number where applicable below

NHS Secretariat	0203 758 2042 x 2
Norlan Dubissette [Scleroderma Co-ordinator]	0203 758 2042 x 4
Scleroderma Nurses Help-line	0203 758 2042 x 5
Clinical Trials	020 7317 7544
Kevin Howell	020 7794 0500 x 22516
Pulmonary Hypertension Nurses	020 7472 6354
Kim Fligelstone & Millie Williams	020 7794 0432
Pamela Yeomans	01273 672686 [Monday x 33471]



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Scleroderma Family Day – Saturday 19th May 2018

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 – 10:20	Welcome	Dame Carol Black Profs Chris Denton & David Abraham
10:20 - 10:45	Getting to the heart of scleroderma	Dr Dan Knight
10:45 – 11:10	The importance of classification in scleroderma	Dr Kristina Clark
11:10 – 11:35	Emerging biological therapies for scleroderma	Dr Voon Ong
11:35– 12:00	Update on fat stem cells to treat scleroderma	Prof Peter Butler
12:00 – 14:15	Lunch break – see below	
14:15 - 14:45	Psychological aspects of chronic immunological disease	Dr Mari Campbell Clinical Psychologist, RFH
14:45 - 15:30	International Speaker Systemic sclerosis in children – similarities and differences	Dr Ivan Foeldvari
15:30	Raffle	

Lunchtime Discussions Groups /Demonstrations include

Clinical Trials	Rachel Ochiel and team
Drug information/monitoring	Pharmacy
Massage	Mr Keith Hunt, Complementary Therapies
Pulmonary Hypertension Education/nursing	Sally Reddecliffe/Adele Dawson
Rheumatology Laboratory	Research Laboratory Staff
Scleroderma Education/Nursing	Louise Parker/Joseph Cainap
Thermography	Dr Kevin Howell
Sjogren's Syndrome	British Sjögren's Syndrome Assoc

