



**MERRY
CHRISTMAS**

From us all

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www.scleroderma-royalfree.org.uk

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Christmas 2009

Dear Friends

May I wish you all a very Happy Christmas and joyous New Year. I also bring you good wishes from The TriState patient organisation, whose headquarters are in New York. I was quest speaker at their annual research forum in November. They are an energetic well-informed group, who share the same hopes, fears and challenges as all of you who live with scleroderma. I never cease to be humbled by the courage and determination of the patients I have known and served over many years. In the lecture I gave in New York I reviewed the progress that has been made over the past twenty years. It is considerable and it is important to note that recent clinical research by Professor Denton and colleagues has shown a considerable improvement in mortality and early identification of lung fibrosis. We should enter 2010 with hope and optimism

A Happy Christmas

Professor Dame Carol Black

Update on the Clinical Scleroderma Service

Recent research confirms better outcomes for patients – Professor Chris Denton

There have been some very exciting and positive developments since the last newsletter. One of the most significant is the recent appointment of Dr Voon Ong as a new senior lecturer and consultant in the department. Many of you will know Dr Ong from the clinic, where he has been a valuable member of the team over several years. He completed his PhD researching scleroderma and then his specialist training in rheumatology in Oxford. We are delighted that he has come back as a permanent member of the scleroderma team. PhD success has been a common theme of the past few months with success also for Dr Rachel Hoyles, who worked in the clinic and lab in joint research with the Brompton, and Dr Richard Stratton. Dr Anna Nuttall also recently completed her research MD degree.

This is a very exciting time for scleroderma research and there is growing optimism about better treatments. There is more interest than ever and this is exemplified by studies from our unit recently presented by Dr Svetlana Nihtyanova in USA at the American College of Rheumatology international meeting. We show how better assessment and treatment mean that patients are now living much longer even with the most severe forms of the disease. This work will soon be published in a major medical journal.

There have been major successes in clinical research, for example the Royal Free will lead a major new European initiative to examine new blood tests for scleroderma funded by a grant from EULAR, the European umbrella organisation for rheumatology that has selected scleroderma as a target "orphan disease" for funding.

There are many challenges in the NHS at present but a real commitment to highest quality care for rare diseases. With this in mind the Royal Free Trust and University College London are establishing close links through UCL Partners, one of five Academic Health Science Centres in UK designated by the Department of Health. There are ongoing discussions to upgrade and expand the integrated clinical space for rheumatology, scleroderma and pulmonary hypertension and this is expected to lead to long needed improvements in the environment in which we can see patients and their families.

Website News

This year we have some IT support and we are currently in the process of updating our website. Our new website, which we hope will go live in January, will have a new format, links to the Royal Free NHS Trust site and updated information.





Laboratory and Research News – Professor David Abraham

As we approach the end of 2009, it would seem a good opportunity to reflect back on the last 12 months of scientific research in the Royal Free Raynaud's and Scleroderma Unit. It has been an exciting time in the laboratory with a number of important findings. We have continued our studies into the genetics of Scleroderma and have identified new genes that appear to be associated with the disease process. These are undergoing further evaluation. We have also been studying in more detail and refining our use of new model systems with which to study the development of scleroderma and the potential use of novel treatments. Research has also been progressing at a rapid pace into the study of the growth factors that we feel are important in driving tissue fibrosis in Scleroderma and we are building up a more complete picture of the precise role in Scleroderma and whether they can be used to modify the disease process.

Major achievements

- Identification of new Scleroderma related genes
- The study of new biomarkers to help with disease assessment and management
- Development and use of new model systems to understand scleroderma and to test new treatments
- Detailed investigation into some of the key growth factors that promote scarring and fibrosis

Who is supporting our research?

Our research continues to receive exceptional and long-term research support from the Raynaud's and Scleroderma Association. We receive this in the form of a joint programme grant held with Athol Wells at the Brompton Hospital which is aimed at understanding the development and treatment of pulmonary fibrosis and also as grant support for specific research projects. These include studies into the role of the epidermis in scleroderma and the influence of growth factors that drive the excessive scarring process. We also receive major grant support from the Arthritis Research Campaign (ARC) and Medical Research Council to study the links between blood vessel damage and fibrosis, and to develop and study new and novel model systems for vessel disease and fibrosis in Scleroderma.

New Research grants awarded include:

- Two new project grants from the ARC to further study the interactions between different cell types in the skin and a novel protein that appears to be involved in blood vessel disease
- A project grant from the Scleroderma Society to study the role of contractile proteins in Scleroderma

New Staff in Rheumatology Research

We have a number of new staff who have joined the research team. Two new research scientists, Johanna Donovan and Helen Jones joined at the end of the summer, and new student training for her research Doctorate Angela Tam has also recently joined the department. We have a new laboratory technician, Ronald Johnson who helps the senior members of staff with the smooth running of the laboratory and also joins Dr Carmen Fonseca to carry out a research project into the genetics of scleroderma. We are also hoping to appoint a new young investigator to the research team early in 2010.

National and International Meetings

This year our research work has been presented at both National and International scientific symposia. Our work was well received at the British Society for Rheumatology meeting in Glasgow at the end of April with both oral and poster presentation given. We also presented our new research finding at the European societies meeting in Limoges in France at the end of August. Highlights include two of our studies being given awards at the American College of Rheumatology meeting in Philadelphia in October. One presented by Korska Khan on fibrocyte recruitment in lung injury, and the other by Dr Emma Derrett-Smith on studies into the vascular disease in Scleroderma.

The Future – A busy 2010 lies ahead!

The calendar for 2010 is already looking very busy for us, with two of our most important meetings taking place. Firstly the first World Congress in Scleroderma takes place in Florence in February and our 11th International Scleroderma Research Workshop, scheduled for Boston at the beginning of August. We are key active participants in both meetings where we will present some of our new research work to the wider Scleroderma research community.





Xmas Newsletter - Scleroderma Unit - Royal Free Hospital



Kim Fligelstone

I can't believe that it's Christmas so soon, it's great to be back at work and this is just a little reminder that if anyone would like to talk to someone with scleroderma your very welcome to give me a call, I usually work on Tuesday and Wednesday's although that can change so do call(number below) and I will always get a message when I come to work alternatively if your on any of the wards and would like me to pop up and say hello call 0207 794 0500 ext 35131. I'm not a counsellor/therapist but have had the systemic type of scleroderma for past 22 years.

Keith Hunt and his team of therapists have been offering their massage service at the Royal Free since 1993. If you would like a massage just ask one of the ward staff to get in touch with Complimentary Therapy Services to book one. The hospital is in the process of employing a patient pathway co-ordinator who should be in post by the end of 2009 who will book tests to coincide with appointments. For now the general appointment number to ring is 0207 794 0500 Ext 36000.

I wish you all a lovely Christmas and as healthy and peaceful New Year as possible.

Thermography and Capillaroscopy – Kevin Howell

Autumn 2009 has been a particularly busy period for our work in thermography and capillaroscopy. The service continues to assess large numbers of patients with Raynaud's phenomenon and early scleroderma to look at the important changes which occur to the small skin blood vessels. Localised scleroderma is an area where thermography can certainly assist in assessment, and our relationship with the paediatric rheumatologists and dermatologists at Great Ormond Street Hospital remains close for this purpose.

Kevin has been in Germany twice in recent weeks for meetings: firstly in Mannheim for the European Association of Thermology conference, and secondly in Hamburg at the invitation of paediatric rheumatologist Dr. Ivan Foeldvari for a workshop on improving outcome measures in juvenile scleroderma.

Kevin's PhD studies are at an advanced stage: his thesis on the use of thermography in scleroderma is now complete. More news about that in the spring!

Wishing you all a very Merry Christmas and a Happy New Year.



Specialist Nurses

It has been a busy and changeable year for the Scleroderma team. Rachel left the team this summer due to family commitments and we wish her well. Helen is currently on maternity leave and is due back early 2010.

Louise and Tanaka have been busy with all the aspects of running the service. Louise continues with the Raynaud's clinic, which gets busier every year! She is also committed to completing her Masters degree having taken a brief break from her studies to get married in August. Louise's role has also increased this year as she is currently managing the Scleroderma and Pulmonary Hypertension teams on a temporary basis.

Tanaka joined the team in February and some of you may have met her in clinic or spoken to her on the helpline. Tanaka has settled in well to the role and is keen to develop other aspects of the service. She is currently undertaking a specialist course in order to assess various wounds and in particular digital ulcers. We will give you more information in the next newsletter to how she will use her new skills!

We look forward to seeing all of you in clinic and on the ward round and please get in touch with us on the helpline if you need us anytime – we are here to help.

We hope you have an enjoyable festive season, a merry Christmas and best wishes for 2010.

Pulmonary Hypertension Team

It's nearly Christmas and the PH service remains as busy as ever! The team continues to expand. Katie joined the team in July, bringing the total number of specialist nurses to four. On the administration side, Alison is now joined by Innes, who was appointed permanently in September. Outreach clinics are growing in number and frequency. We are also in the process of setting up a one-stop new referrals clinic so that patients can have their clinic appointment and tests done on the same day. Clare is currently on maternity leave having had a baby girl called Maya in June. Mum and baby are doing well and Clare will hopefully be returning to work in the New Year.

Hello from Katie - Thank you to everyone for being so welcoming. I'm really enjoying my new role, and look forward to meeting more of you in 2010.

Wishing you a Merry Christmas and a Happy New Year!





Clinical Trials – Rachel Ochiel

We remain very busy in the clinical trials unit with several studies at different stages. The unit also continues to expand in order to cope with the number of exciting studies we are currently involved in. The team works alongside the specialist nurses and doctors. The unit now also has an additional research nurse, admin support and data entry/IT person. Despite the long delay we still plan to provide a summary of the results of completed studies. Finally we would like to thank all those who continue to volunteer in our trials. Below is a summary of our clinical trials activity

Ongoing Scleroderma Trials

1. DETECT - An observational study to find out early, simple and reliable methods that can be used in the detection of pulmonary Hypertension.
2. AIMSPRO - A study to find out if the drug AIMSPRO improves the quality of life in long standing Diffuse Scleroderma and to confirm that it is safe and well tolerated.
3. DISTOL - A study to find out the effect of a drug called Treprostinil diethanolamine (UT-15C) on digital ulcers in patients with scleroderma.
4. DUO Register - This is a multi-centre prospective, observational study non-interventional programme to follow-up patients with digital ulcers over the course of the disease. The outcome of this study will inform on how to improve patient care in the long run.
5. Gleevec (Imatinib) - A trial to evaluate the efficacy and safety of an oral dose of STI571 for diffuse patients with fibrosis
6. P144 - This is a trial to evaluate the efficacy and safety of the topical application of the drug "P144" for skin fibrosis in patients with Scleroderma

Completed Studies for which results will be provided

1. SStep - This was a six year trial that started in 2002 looking at the use of iloprost tablets in lowering the incidence of heart attacks and angina. We are still waiting for the results of this study including the un-blinded data.
2. GTN study - This was a Raynaud's trial involving a topical gel formation of Nitroglycerine MQX-503, and matching placebo, to prevent Raynaud's Phenomenon

Ongoing Pulmonary Hypertension Trials

1. SERAPHIN - This is a study to find out the effect of a drug called macitentan in pulmonary arterial hypertension.
2. FREEDOM STUDIES - These are studies to see the effect of UT-15C sustained release tablets in pulmonary arterial hypertension.
3. TRIUMPH - An extension study to find out the effect of Treprostinil Inhalation in the Management of severe Pulmonary Arterial Hypertension (PAH)
4. PATENT - A study to assess the effectiveness and safety of different doses of a new drug (BAY-63-251) given orally to patients with pulmonary hypertension.
5. Tadalafil - This is a study to evaluate the long term safety and efficacy of a drug called Tadalafil in the treatment of patients with pulmonary hypertension.
6. VOLT - This is a post marketing observational surveillance programme of a drug called Ambrisantan which is already licensed for treatment of PAH.

If you are interested in participating in any trial please contact the team on 02073177544.

Scleroderma Family Day at The Royal Free

Saturday 15 May 2010

Scleroderma Day is one of our favourite days. We will start to put the programme together in the New Year, if there are any areas that you would like us to include please let me know. We have included a book of raffle tickets with your Newsletter - the raffle raises much needed funds and we are very grateful to all of you who buy/sell tickets on our behalf. If you do not wish to use the tickets please discard them - you do not return them to us. If you would like more tickets please contact Pamela on 01273 672686 or pamela.yeomans@scleroderma-royalfree.org.uk.

Newsletter

We hope you enjoy reading our Newsletters. We send them out to all Raynaud's & Scleroderma patients. If you would prefer a paper-free copy these can be download from our website. Pamela

Contact Numbers

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