

# Christmas Newsletter

[www.scleroderma-royalfree.org.uk](http://www.scleroderma-royalfree.org.uk)

*Scleroderma Unit  
The Royal Free Hospital -  
December 2004*

## Seasons Greetings from all at The Scleroderma Research Unit

*Dear Friends*

*May I wish you all a very Happy Christmas. It has been a very busy one for me as President of the Royal College of Physicians and although I have kept in excellent research contact with the Department I have had very little time to be in the clinic. I have always enjoyed my out-patient clinics and I miss them. I hope this coming year to have the opportunity to be there more frequently. It is however a great pride to me that Drs Denton, Brough and Coghlan have developed the service so well.*

*In 2005 we celebrate 10 years of Family Open Days. We want this to be a great success so put it in your diary now! The date is Saturday 21 May. May 2005 bring you happiness and peace*

*With best wishes*

**Carol Black**



## Clinical Scleroderma at the Royal Free Hospital : Dr Chris Denton

*The past few months have been as busy as ever for the clinical staff. Several members of the medical team travelled to San Antonio in Texas for the American College of Rheumatology meeting. Drs Ong, Carulli and Shand and I presented our various research projects and there was considerable interest from other international groups. It is always a good chance to catch up with other scleroderma experts and to learn about new therapies being tested in pilot trials. We were able to share our own recent experience of anti-TGFbeta antibodies (the CAT192 trial) and anti-TNF for early diffuse scleroderma. The latter trial is still ongoing but the results should be available towards the end of 2005.*

*In the clinic there have been some changes. Dr Lynne Shand has been successful in securing a highly competitive training post as a specialist registrar in rheumatology on the North Thames rotation. She left the Royal Free in November and is sorely missed - especially in the scleroderma clinics. However she will maintain her research links and plans to complete her MD degree in scleroderma research. Dr Svetlana Nhitanyova has recently joined us as a clinical research fellow and is working in the clinic. She is from Bulgaria and quite new to rheumatology but is proving very accomplished and is learning very quickly.*

*We are fortunate to have received clinical research support for some of our database projects and are especially grateful for a 5 year grant from the Arthritis Research Campaign to support development of the National Scleroderma Central Register. This work has been progressing for the past 5 years but will now be able to maximise its potential.*

*We are hosting a second Pulmonary Hypertension Workshop for doctors from other European countries just before Christmas. The last one was very successful in 2003. This year we are expecting rheumatologists from the Benelux countries. We have the largest experience of scleroderma related pulmonary hypertension in the UK and are delighted to share our knowledge in this important area that represents a real area of progress for treatment in scleroderma.*



# Laboratory and Research News : Dr David Abraham

As we approach the end of 2004, I feel that it is timely to look back on what we have achieved in scientific research over the past 12 months. I believe that the laboratory work has gone very well with a number of studies yielding new and important information about the disease processes in Raynaud's and Scleroderma. Three particularly important findings have been the identification of a fibroblast receptor for connective tissue growth factor, one of the most potent factors involved in driving fibrosis in Scleroderma; the examination of genes that are activated by endothelin-1, another important factor involved in the vascular changes seen in Raynaud's and Scleroderma and the finding of a molecule called a chemokine acts as a mediator of both the inflammatory response and also as a fibroblast growth factor. These achievements were made possible with the research support from The Raynaud's and Scleroderma Association, The Arthritis Research Campaign and The Welton Foundation, and the scientists involved in these studies are Dr Y Chen, Dr A Leask, Dr X Shi-wen, Dr M Carulli and Dr V Ong. These studies are important as they show us the way in our quest to develop more effective therapies for Raynaud's and Scleroderma.

In addition to these studies, work carried out by Markella Ponticos has shown that a type of cell found in the blood stream (a T-cell) is able to produce growth factors that influence fibroblast growth and production of collagen. These cells are found in the initial stages of the disease and may be important as early drivers of the disease process. Also, Vineeth Rajkumar, one of our research assistants has shown that in early scleroderma some of the cells in the blood vessel wall can move into the surrounding tissues and become fibroblasts. This provides, for the first time, a potential link between the early blood vessel changes that are almost always present in scleroderma and the ensuing scarring process.

## Research Grants

Other important items of news are the research and equipment Grants that have been awarded this year: The Arthritis Research Campaign- have awarded us two project grants- both to study important aspects of collagen regulation in scleroderma. One of these grants is a collaborative venture with Dr George Bou-Gharios at Imperial College London and the other is to work in collaboration with Dr Gisela Lindahl and Dr Sankar Maity in Houston, Texas. The Rosetrees Trust- have continued their long-term support of our basic research in Scleroderma with an emphasis on translational research studies. The Raynaud's and Scleroderma Association - have been instrumental in supporting our research

endeavours over many years, and have continued to significantly support the laboratory research. The Scleroderma Research and Development Action Committee and The Scleroderma Society have both continued to support vital research projects in the department. The Welton Foundation – have awarded us additional funding to support our ongoing research.

## National and International Meetings

Much of our research work has been presented at scientific meetings in the UK, Europe and the US as both abstract or poster presentation and oral communication. In September Drs Andrew Leask and Xu Shi-wen, and Alan Holmes attended the CCN meeting in St Malo Northern France to present their work on growth factors in Scleroderma. These were poster presentations and lectures. In October many of the research and clinical staff attended the American College of Rheumatology meeting in St Antonio, Texas. This, with the British Society of Rheumatology earlier in the year, is one of the major meetings in the calendar, and provides a very active forum and interactive environment to display our work.

Equally prestigious is the American Society for Matrix Biology Symposium, which was held in November in San Diego. Drs Gisela Lindahl and Markella Ponticos both attended this symposium and presented some of their most recent work. It also provided an opportunity to meet-up with some of our most loyal and long-term collaborators; Professor Benoit de Crombrughe and Professor Francesco Ramirez, whom have work with us for many years unravelling the complex processes in Scleroderma.

Nearer home the Department is still active in hosting the once-monthly London matrix group ([www.londonmatrix.org](http://www.londonmatrix.org)).

This meeting consists of a series of lectures throughout the year, and brings together all the groups working in London and whom have interests in connective tissues and associated diseases. This has been a very successful series of lectures, having attracted some very distinguished speakers in the past. It has also served to promote and widen the interest in Raynaud's and Scleroderma in London and throughout the UK.



# News From Our Nurse Specialists: Helen Clare Rachel Joanna Jane

Christmas is here again and the year, as always, has been very busy and has brought changes for the specialist nursing team for scleroderma and pulmonary hypertension: Helen (Clinical Nurse Specialist), Clare (Pulmonary Hypertension Specialist Nurse), Rachel (Education Nurse), Joanna (Pulmonary Hypertension Nurse) and Jane (Trials/Research Nurse), not forgetting Sharon, our clerical assistant, who is worth her weight in gold.

Leo our research and trials nurse left us in the summer and now works as an infection control nurse at Northwick Park hospital in London. She has been replaced by Jane Eagle whom some of you will have already met. Jane has started at a very busy time in terms of clinical trials as we continue to be able to offer trial treatments in the hope of finding useful therapies for scleroderma or pulmonary hypertension (PAH). As always if you may be interested in taking part in a trial you should contact Jane or Helen on 020 7830 2326.

Part of our role is to raise awareness of these sometimes devastating conditions and therefore we are delighted when asked to speak at meetings or patient groups. This year Helen and Rachel gave a talk about tests in scleroderma at the Raynaud's and Scleroderma Association annual conference in Chester, Joanna gave a talk about PAH in Connective Tissue Disease at the Pulmonary Hypertension Association (UK) patient conference in Cambridge, Clare spoke to delegates from the Department of Health about the services we offer at the Royal Free and some of you may have seen us when we all took our turn on Family Day back in May. Rachel also gives regular teaching sessions to the nursing staff on the wards at the Royal Free so that they understand the conditions and are able to look after patients well when they are admitted to the hospital. All the nurses have also attended various national and international meetings where they have learned about new developments in research into both the causes and treatments of scleroderma. We are then able to bring this information directly back to you the patients.

Joanna was lucky enough to be sponsored by the PHA (UK) to attend the patient conference of their North American group in Florida in the summer where she had a chance to meet both American sufferers of this awful condition and the dedicated medical teams and nurses looking after them. Clare was unable to take up the opportunity to attend as she had a prior engagement – our congratulations to Clare who became Mrs Das in July. The PAH nursing service continues to expand. This year has seen the introduction of a second weekly clinic which it is hoped will eventually become nurse-led, as well as out-reach clinics held in other hospitals which Clare often attends. Clare and Rachel have also written a

comprehensive booklet about PAH which it is hoped will give patients a better understanding of their rare condition.

Helen has been pleased to be able to develop her team this year thereby enabling us to offer more help to our patients. She has been busy this year managing the team, setting up the many new and ongoing clinical trials, and dealing with other departmental issues. However she still runs her Raynaud's clinic and enjoys having the chance to see and help patients.

2005 already promises change within the team: Rachel will be leaving us in February to go on maternity leave and although she is expected back later in the year we will be taking on another permanent member of the team when she leaves as the workload continues to expand. Amongst other plans we also hope to set up a nurse-led service for the administration of new therapies for scleroderma, rheumatoid arthritis and other rheumatic conditions.

We'd like to take this chance to wish you all a very happy Christmas and hope to be able to see many of you in the coming year.



## **How You Can Help Us Help You...**

The department is very committed to research into scleroderma and pulmonary hypertension but without patients willing to take part in clinical trials potential therapies may never be identified. If you are interested in taking part in any of these trials or would like to be considered for future trials then please call Jane or Helen on 020 7830 2326

**SSTEP** – recruitment for this five-year trial into the use of Iloprost tablets continues. It is hoped that scleroderma patients who take Iloprost tablets may report a lower incidence of heart attacks, angina and strokes. Once commenced on the trial you would need to visit the hospital only every six months and where possible we try to coincide these visits with your clinic appointments. Almost all patients with scleroderma are eligible to take part but recruitment will stop in January 2005.

**INFLIXIMAB** – this is a trial of a new treatment for early or worsening diffuse scleroderma and has already been used with much success in rheumatoid arthritis. Patients receive five infusions over the course of the trial which lasts 48 weeks. We already have some patients who have taken part but are still able to offer it to more.

**ARIES / STRIDE** – these two trials are both for patients with pulmonary hypertension and are looking at two new tablet medications, ambrisentan and sitaxsentan to see whether they are useful treatments for slowing down progression of PAH.

## Blood Flow Studies

### Kevin Howells

*Since you last heard from us in the spring, it's been non-stop action for me!*

*In early November I attended the UK Symposium on Medical Imaging at the National Physical Laboratory, and spoke about our work on thermal imaging of morphea patients. I've been assisting in arranging the meeting all year, so it was gratifying to see that the meeting was a success, with around 30 delegates attending from across Europe. The meeting was staged with the support of the UK Thermography Association (UKTA). Currently I am medical representative on the UKTA committee of management, and a recent task I've been given is to keep our website updated. Check out [www.ukta.org](http://www.ukta.org) to see what UKTA gets up to, and in particular the medical section!*

*September was also a busy month. I started up in York for the Annual Scientific Meeting of the Institute of Physics and Engineering in Medicine (IPEM). This year the meeting had a session on "clinical temperature measurement" and it was good to find that the venue was standing room only for this very important topic. Professor Black also addressed the conference dinner in her capacity as president of the Royal College of Physicians (IPEM and the RCP collaborate closely on many healthcare issues).*

*Unfortunately I missed the address because I was en-route to Genoa at the invitation of Professor Cutolo, for the first ever European course on capillaroscopy, where I gave short lectures to the students. The course was a great success, with more than 90 students attending from over 20 countries, some as distant as Dubai and South Africa. Capillaroscopy is recognised worldwide as an important tool in diagnosing connective tissue diseases like scleroderma, but up to now only a few centres have had the equipment and expertise to offer the kind of service to patients that we provide at the Royal Free. Courses like Prof. Cutolo's will ensure that capillaroscopy is more accessible in the future. Further courses are already planned: I shall be delivering the material again in June 2005 in Vienna!*

*Back at the Royal Free, our morphea imaging research continues apace. We are due to see more patients just before Christmas, fitting around a special clinic that some of these children attend, run by our colleagues at Great Ormond Street Hospital.*

*So all in all, another busy year in 2004. See you all in 2005!*

## Talking About the Disease

### Kim Fligelstone

*I am a long standing patient at the Royal Free called Kim Fligelstone. I have had scleroderma (Diffuse) since 1988. In 1991 I had a renal crisis and started dialysis which is kidney replacement therapy. Since then my condition appears to have stabilised and my mobility has vastly improved. I work in the Rheumatology unit part time and one of the more pleasurable aspects for me is visiting patients on the ward, so if you would like to talk to a patient with experience, I must stress that I am not a Counsellor or a therapist, then please ask the ward clerk to call Ext 5131 or you can call me at on 020 7794 0500 ext 5131 if you would like to chat on the phone, I usually work on Tuesdays and Wednesdays.*

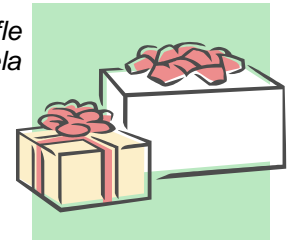
*Have a very Happy Christmas and as Healthy as possible New Year.*

## Family Day

### Saturday 21 May 2005

*A date for your diary in 2005. We have enclosed a book of raffle tickets — for the Family Day Draw. The raffle has, over the past few years, raised valuable funds and we are very grateful to all those who buy tickets.*

*If you would like more raffle tickets please contact Pamela Yeomans on 01273 672686*



## How to Contact Us

### Direct

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Helen/Jane/Rachel	020 7830 2326
Clare/Joanna	020 7472 6354
Scleroderma Appts	020 7830 2151
Millie	020 7794 0432
Kevin Howell	020 7472 6550
Pamela	01273 672686

### Via the switchboard on 020 7794 0500

Kim Fligelstone	Ext 5131
Angie Locke [Bed Manager]	Bleep 232