



# The Scleroderma News

Winter Issue  
January 2007

## The Newsletter of The Scleroderma Society (UK)

### A Word from Kim

### Happy New Year

The last few months have been busy for the Society Steve Holloway and I went to the London Marathon Bondholders meeting in October. After years of being on the waiting list for a Gold Bond we were thrilled to become (because of our position on the waiting list) the recipients of the London Marathon's first wave of Silver Bonds. **More about this inside** but we must thank the existing Gold Bond Holders who have given up part of their allocation to allow this to happen.

The Musculoskeletal Service Framework was finally launched in London on the 31<sup>st</sup> October. Anyone wishing to view the framework on line can visit: <http://www.dh.gov.uk> and search for Musculoskeletal Service Framework.

We are very excited about our 25<sup>th</sup> AGM/Conference which will start earlier than usual to accommodate all our prestigious speakers.

**DAME PROFESSOR CAROL BLACK, President**, who needs no introduction as a founding member of the Society and World Scleroderma Expert.

We are very honoured that **PROFESSOR MARCO MATUCCI CERINIC** will join us from Italy. He is the President of EUSTAR, the EULAR scleroderma trials and research group.

**DR HENRY PENN** from the Royal Free will be talking about kidneys and scleroderma.

**SISTER NAOMI REAY** from Leeds will be giving us an update on the Quality of Life Study.

Last but certainly not least, back by popular demand, **SONIA TELLO** who inspired us all in 2002. Sonia has Scleroderma, had a Scleroderma renal crisis, works full time and will share her views and motivating coping strategies.

I urge you to make a note of the date 28<sup>th</sup> July 2007. Booking forms and details will follow in the next issue of "Scleroderma News" for this special Conference.

Lastly I would like to thank everyone who has sent Christmas Cards. The numbers are overwhelming. I have to apologise that I haven't sent Cards this year. I will be making a donation to the Society instead. I do hope that you all had a wonderful Christmas and best wishes for a Healthy and Peaceful New Year!

Take care Lol Kim

### THE QUARTERLY NEWSLETTER OF THE SCLERODERMA SOCIETY

CHARITY REGISTRATION NUMBER:286736

The Scleroderma Society 020 8961 4912

Advice Line Hours 9.00 till 9.00 7 days per week.

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*The Scleroderma Society is not responsible for any information, news or views that appear in this newsletter.*

**Please send YOUR articles, etc.** for the next issue to Carol Sanders,

The Laurels, Westport, Langport, Somerset. TA10 0BN  
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**Closing date for contributions 16<sup>th</sup> March 2007**

### SCLERODERMA FAMILY WEEKEND

The Raynaud's & Scleroderma Association will be holding its 6th Family Weekend (9th, 10th & 11th February) at the Crowne Plaza Hotel in Chester. Families who have a child under the age of 18 with scleroderma are welcome to attend.

The event will be a good opportunity for families to be educated by members of the medical profession about the condition and the best coping strategies as well as meeting other children in the same position. The Family Weekend is subsidised through sponsorship which means that there is only a minimal cost to the families. For more information and a booking form please contact the Raynaud's & Scleroderma Association on: 01270 872776 or email [fiona@raynauds.org.uk](mailto:fiona@raynauds.org.uk)

## HAMPSHIRE

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The Hampshire and West Sussex  
local group broke with tradition  
this year. Instead of the usual  
afternoon tea meeting in  
November, we had a most  
enjoyable Skittles and Fish and  
Chip supper, organised by David  
and Celia Graham. It was held at  
the Southern Electric Sports and  
Social Club (David's local!) and  
was attended by 12 members.  
After a competitive skittles  
match, the final knockout was  
won by Dan, son of Tracey and  
Colin James. Second place went  
to their very young daughter Flo.  
David presented a small trophy  
to the winner. As well as great  
fun and good fish and chips, we  
raised £75 from the raffle and  
sales of Celia's birthday and  
Christmas cards. Thanks to  
everyone for supporting this  
event and especially to David  
and Celia for organising it. A  
return match is already being  
planned for next year!

On her father's birthday Tracey  
organised a meal in The Balti  
House in Southsea, in his  
memory. Everything was  
extremely kindly provided by  
Tracey and Colin and anyone  
attending was asked to give a  
donation to the Society. It was  
mostly Tracey's family there and  
very friendly and generous they  
are. £185.50 was raised at the  
end of the evening as well as a  
good time being had by all. Very  
many thanks.

The next meeting will be on  
Wednesday May 2<sup>nd</sup> at 7.00pm in  
the Seminar room in North  
Building at QA. This is now the  
Rheumatology Outpatients Dept.

# Area News

and is part of the main patient car  
park.

Thanks, Liz and Steve

## LONDON AND SOUTH EAST

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[info@sclerodermasociety.co.uk](mailto:info@sclerodermasociety.co.uk)  
The next meeting is on March 22<sup>nd</sup>  
at 7.30 in the Rheumatology Library  
on the Lower Ground Floor  
(Opposite the main lifts) at the  
Royal Free Hospital, Pond Street,  
Hampstead, NW3 2QG. Nearest  
tube: Belsize Park on the Northern  
Line and Silver Link at Hampstead  
Heath. Kim

## OTHER CONTACTS

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### GLASGOW & WEST SCOTLAND

For the time being please contact  
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### SOUTH WALES

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*If anyone has any news to go into  
this section of the newsletter  
please send details preferably via  
email to Kim (details earlier) or  
to Carol and Roy  
[roy4sanders@aol.com](mailto:roy4sanders@aol.com)*

## CONGRATULATIONS!

Dr Markella Ponticos and  
Professor David Abraham  
have a gorgeous baby girl  
called Eirini who was born at  
the Royal Free Hospital.

## GRANT AWARD

The Society has awarded an  
equipment grant of £17, 010 to  
Dr Mark Eastwood and Dr  
Catherine Sarraf for a multi-  
station tensioning-Culture  
Force Monitor at the School of  
Biosciences, Department of  
Biomedical Sciences,  
University of Westminster.  
The multi-station tensioning-  
Culture Force Monitor will be  
used to study the potential of  
anti-fibrotic agents and the  
role of mechanotransduction  
on fibroblasts from  
Scleroderma origin.

## TRUSTEES

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Liz Holloway, Belinda Thomson,  
Frances Bain.

## Treatment for Pulmonary Hypertension related to scleroderma in 2006

*Clive Handler, Consultant in Pulmonary Hypertension, Clare Das, Senior Specialist Nurse, Professor Christopher Denton, Consultant Rheumatologist, Geraldine Brough, Specialist in Rheumatology, Gerry Coghlan, Consultant Cardiologist  
The National Pulmonary Hypertension Unit and Centre for Rheumatology, Royal Free Hospital,  
London, NW3 2QG, UK*

As well as causing scarring or fibrosis of the skin, scleroderma (systemic sclerosis) can lead to similar changes in the blood vessels and other internal organs. A small proportion (8%) of patients, more commonly those with limited compared to diffuse SSc, develop another serious condition called pulmonary arterial hypertension (PAH) - a high pressure in the lung arteries. "Ordinary" hypertension - is a high pressure in the arteries that carry fresh blood around the body. Whereas ordinary hypertension is common, can be effectively treated with several different types of tablets and most patients don't know they've got it, PAH is much rarer, affecting around 40 people in every million. Patients get breathless and tired and it is more serious. Until recently, we had little to offer patients with PAH. Now we have several different drugs and new ones are coming along. Although none of these "cure" PAH, they can, in some patients, improve symptoms, reduce the pressure in the lungs, and help patients live a longer and more active life by stabilising the condition. This short article will tell you about PAH and how we treat it. Scleroderma patients, their families, and their doctors and nurses, are justified in feeling much happier and more optimistic about the future.

What is PAH? In people with PAH the walls of the pulmonary (lung) arteries, which are the blood vessels carrying "stale" blood from the right side of the heart to the lungs, get thick and narrowed. This makes it more difficult for blood to get through the lungs and puts an increasing amount of pressure on the right heart pumping chamber (right ventricle), which gets weak (heart failure). Patients feel progressively breathless and unable to walk more than short distances. Patients with scleroderma often do not get swollen ankles because of their tight skin. When the heart gets very weak, patients may lose their appetite and lose weight. Patients with PAH are also more vulnerable to getting blood clots in the lungs because the flow of blood in their lung arteries is slow.

### Are there other reasons why the pressure may be high in the lung arteries?

There are several other much more common causes of breathlessness (bronchitis, asthma, heart disease, and being unfit) than PAH. These have to be excluded before PAH is diagnosed.

Any problem with the left heart pump (the left ventricle) – for example, a weakness of the muscle due to a heart attack or high blood pressure - can cause a high pressure in the lungs. People who get blood clots in their lung arteries, and individuals who have holes in their heart, may also get high pressure in the lung arteries.

How serious is PAH? It depends on how high the pressure is. Whereas some patients with only a slightly high pressure in the lungs can lead a very active and productive life, as long as they don't over-do things, others with severe PAH get more and more breathless quite quickly. Without treatment the outlook is poor in more severe cases – such that in very severely affected patients their future is similar to some forms of aggressive cancer.

How is PAH diagnosed? In common with other internal complications of scleroderma it is important to diagnose PAH as early as possible. Therefore all scleroderma patients should have regular screening tests. In order to pick up those patients who may be developing PAH, we recommend that they also have

- an ultrasound examination of their heart (echocardiogram) to measure the pressure in the lung arteries. This is not very accurate but is a useful guide.
- breathing tests (lung function)
- a walking test to see how far they can walk in six minutes
- a blood test called BNP which can be done in nearly all hospitals. A very high level suggests that the heart may be under strain, though the test does not tell us why this is so – for that further investigations are required
- chest X ray
- ECG (electrical recording of the heart).

We analyse all these bits of information to help us decide whether further tests are necessary.

What is the most accurate test to diagnose PAH and how is it done? A right heart catheter is the most important test. It allows measurement of the pressure in the lung arteries. PAH cannot be reliably diagnosed without this test. Patients take all their drugs on the day of the procedure (except warfarin which should be stopped three days before, and diabetic tablets on day of the procedure). Patients lie on an X ray table and a thin tube is inserted under local anaesthetic, into the vein in the groin (femoral vein) and then positioned, using X ray guidance, into the right side of the heart and then into the lung arteries. Most patients do not feel anything more than a pushing sensation. A pressure of 25 mm Hg or more at rest or 30 mm Hg or more during arm exercise means that the patient has PAH. The catheter tube is also used to take blood samples and to measure the amount of blood pumped around the body, and the resistance to the flow of blood in the lungs. This is called the cardiac output and will be low in patients with a weak heart.

How does the result affect treatment? Only patients with PAH respond to the available drugs, patients with high pressure in the lung arteries for other reasons (such as left heart failure) need different treatments.

What if the tests are normal? If we think that a patient has *not* got PAH, we would see the patient in a year. If the tests are borderline, we would repeat the tests in six months. If we are certain that a patient does not have PAH, we refer them back to their usual physician for continued monitoring.

What treatments are given to patients with PAH? We divide the types of treatment into two categories – basic and advanced.

#### **BASIC TREATMENTS**

These do not affect the disease process but relieve symptoms of PAH and heart failure.

Oxygen We encourage patients who are very breathless to use oxygen for most of the day, provided the oxygen level in the blood is low. This can be done with a condenser obtained via the GP or arranged by their PAH centre. Small portable oxygen machines are also available and allow patients to leave home.

Warfarin This anticoagulant thins the blood and reduces the likelihood of blood clots in the lungs. It might also reduce the thickening in the wall of the lung arteries. The GP or local hospital monitor the blood International Ratio (INR) every two months, and advise patients of the correct dose to keep the INR between 2-3. Warfarin may have to be stopped if patients get recurrent nose bleeds or bleeding from the stomach.

Water tablets (diuretics) This reduces fluid retention in the legs and lungs and makes it easier for patients to breathe. Spironolactone and, in addition, when necessary, furosemide and bumetanide, are used. The dose of these diuretics may need to be increased if patients develop a lot of water retention. The dose is decreased if the kidney function deteriorates.

Digoxin (Foxglove) This is used to help strengthen the heart beat.

#### **ADVANCED TREATMENTS**

These treatments reduce thickening in the lung arteries. There are three different type of drugs. They have all been shown to improve the six minute walking distance and lower the pressure in the lungs.

##### Endothelin antagonists

*Bosentan* is usually the first to be used and we have most experience with this. It is given as a tablet twice a day. Blood tests are done monthly because a small proportion of patients get liver damage which disappears when the drug is stopped. It slows down the rate of progression of the thickening in the lung arteries and this improves breathing by reducing the resistance to blood flow through the lung arteries. It may lower the blood pressure and some patients feel light headed if they stand up quickly. We have shown that *Bosentan* has improved survival in patients with SSc-PAH compared to previous treatment approaches.

*Sitaxentan* is a relatively new drug but we do not yet know how it compares to *Bosentan*, although in clinical trials it appears to be similarly effective for PAH.

##### Sildenafil

Also known as *Viagra®*, sildenafil appears to be a useful drug in PAH. It is often added on to *bosentan* in patients who do not respond adequately to *bosentan* alone. We call this combination therapy. We sometimes

use sildenafil instead of bosentan as first line treatment, for example in patients who would not tolerate bosentan because of liver problems, or other medications. Patients are given sildenafil 20 mg twice a day and then, this dose is increased to 50 mg twice a day. Side effects include headache and lowering of the blood pressure.

Drugs similar to sildenafil are being tested and we look forward to the results of these trials.

### Prostanoids

These were the first drugs used as advanced therapy. The drug, prostacyclin, can be given by three routes: inhalation, into the skin (subcutaneous), usually in the abdomen, and most effectively, through a vein directly into the bloodstream. Inhalations should be given six times per day but this is time consuming. The subcutaneous form is painful and the indwelling needle has to be moved to different sites on the skin. The intravenous form uses a tube placed in a large vein under the collarbone. This procedure carries a small risk of a punctured lung and infection and patients need to be taught how to look after the tube to avoid infection. The doses of prostanoids are increased periodically. These drugs are very effective in helping patients feel better and being more active.

What treatments are offered when the condition deteriorates despite medication? Lung transplantation is considered for patients with severe PAH. It has become much more difficult to get donor lungs. The operation, which carries a significant risk is done at only a few hospitals in the UK.

We occasionally offer a treatment called atrial septostomy. This creates a hole between the collecting chambers but only a very small number of patients are suitable for this treatment.

Living with PAH Although a serious condition, a lot can be done by the team looking after you to provide support for you and your family, and practical advice to help you cope with the condition. You will be told to do as much as you can do comfortably, to enjoy the good days and do not overdo things when you are not in the mood. You will be advised what to do about work. It is useful to have a few good friends to talk to. Patient organisations provide wonderful support and advice on all aspects of your condition, including travelling abroad. They provide the opportunity for you to meet other people with SSc and PAH and many patients find this very helpful and comforting.

It is very important to take the medication prescribed in the correct dose and at the right time of day. Your PAH centre will have a **telephone advice line**. Keep in touch with your PAH centre and let them know how you are, particularly if you feel you are getting worse. Clinic appointments are important and essential for the medical team to be able to see you and examine you. Blood tests and other investigations may be necessary to follow your condition.

You may be asked to participate in a drug trial but this would usually involve extra visits to hospital for assessments. Many patients enjoy being part of trials because they are reviewed frequently and given prompt care. The choice to enter a trial or not, is completely up to the patient.

### Conclusion

As with all medical conditions, effective treatments and cures rely on a detailed understanding of the cause of the problem. We are making considerable progress and are working very hard to improve the health and well being of our scleroderma patients in several ways. The last five years has seen major advances in the outcome for patients with SSc-PAH and we anticipate further advances in the near future.

## WALK FOR SKIN



Due to the outstanding success of the first ever **Walk for Skin** earlier this year, the event will take place again from May 13<sup>th</sup> to 20<sup>th</sup> 2007. The event, proudly supported by Sanex, promises to be even more impressive in 2007, with 17 charities uniting in a bid to raise awareness and money for vital research and support for skin disease sufferers. The Walk for Skin is unique in that a number of charities are working together raising funds for skin disease, rather than each charity holding separate, smaller events. The charities represent a vast range of disorders, including eczema, skin cancer, **Scleroderma**, acne, birthmarks, psoriasis, vitiligo and many more. These diseases affect young and old, men and women and all ethnic groups alike, which is just one reason why we feel the Walk for Skin has been so popular. Three thousand people, including comedian Ricky Tomlinson, TV legend Jimmy Saville and other famous faces, took part in the last Walk for Skin, which raised a staggering £160,000. This money will fund three major grants for skin disease research projects, as well as helping to finance support groups, help lines, patient information services and much more. The walks themselves are approximately three miles long, suitable for all ages and held at scenic, accessible locations all across the UK, which also definitely adds to the appeal.

**If anyone is interested in supporting the Scleroderma Society please contact Caroline Walker at [carolinew7@btinternet.com](mailto:carolinew7@btinternet.com) or telephone: 020 8876 1917. If you register on line please remember to add the Scleroderma Society as your charity. This will entitle us to 50% of all sponsorship and gift aid raised by our supporters and please let Caroline know so that we can keep track of all our fundraisers. Last year the Society received £1,868.22 from the British Skin Foundation. Many thanks to everyone who took part.**

**This year there is a new venue:** Windsor Great Park (Thames Valley). The walk will take place around Virginia Water lake within Windsor Great Park. The lake is said to be named after Queen Elizabeth I, the Virgin Queen, and was created by the Duke of Cumberland, son of George II. Here you will find a one hundred-foot tall totem pole weighing twelve tons and an obelisk raised by George II to mark his son's success at Culloden. The walk is popular for its beautiful scenery and ideal location.

Joining the Walk for Skin couldn't be easier – simply choose a venue and register free of charge, either online at [www.walkforskin.org.uk](http://www.walkforskin.org.uk) or by phone on 0207 391 6341, from February 8<sup>th</sup> 2007. Once registered, entrants will receive a sponsor form, walkers' pack and information about their chosen venue.

### **Sunday May 13<sup>th</sup> Beamish, the North of England Open Air Museum, near Newcastle**

Wollaton Deer Park, Nottingham  
Bramham Park, near Leeds  
University of East Anglia, Norwich  
Belfast Castle, Belfast

### **Sunday May 20<sup>th</sup> Beaulieu Motor Museum, near Southampton**

Cosmeston Lakes County Park, near Cardiff  
Hollyrood Park, Edinburgh  
Cheddar Gorge, Somerset  
Tatton Park, near Manchester  
Windsor Great Park, Thames Valley

### **Dates / venue to be confirmed:**

West Midlands  
London

Join the Walk for Skin and together we can ease the suffering of the eight million people whose lives are blighted by skin disease.

## SCLERODERMA SOCIETY ENTRY IN THE 2007 FLORA LONDON MARATHON

2007 is the 25<sup>th</sup> Anniversary of the Scleroderma Society. To celebrate we are making a special effort to increase our fundraising. As part of this initiative we have been able to purchase a Flora London Marathon Silver Bond. This gives the society a guaranteed entry in the marathon in April 2007 and thereafter once every 5 years. The competition for entry places is extremely fierce. 100,000 people apply for 20,000 places in the public allocation, and there are 14,000 charity places already permanently allocated. The final 1,000 places are reserved for elite runners and high profile celebrities. The Flora London Marathon is the world's largest charity fundraising event, and receives a great deal of publicity nationally.



We are delighted to announce that our entry in 2007 has been taken up very enthusiastically by **Richard Bennett** (shown here training hard) who approached the society after reading about our Silver Bond in an earlier Scleroderma News. Richard will be using the 2007 Flora London Marathon to raise money exclusively for the Scleroderma Society. Richard is 52 years old and is the partner of society member **Gail Tytherleigh** who has systemic sclerosis with lung involvement. They live in Kingston on Thames. Unusually, Gail's Mum was also diagnosed with scleroderma 35 years ago, which was why Gail originally went to the doctor when she started with the same symptoms that her Mum had had all that time ago.

Richard told us: **“Like most people I've watched the Marathon over the years and always promised myself that the following year I would apply. I have applied unsuccessfully over the past 3 years and was about to apply again when Gail read your Society Newsletter - perfect, a cause obviously close to my heart and a place.”**

**“Why am I doing it? Because I appreciate being able to run as I am all too aware that it can quickly be taken from you and more importantly it's for a very good cause. I have played sports from the age of 10, Football to 30, then Rugby to 40, and now it's Tennis, all to a stunningly very average level. I have got myself up to running 12 miles (this is not a total to date figure! ) and feel surprisingly comfortable in doing so. I am aiming to be able to run 15 miles by the end of December, and then keep between 15-18 during January, getting up to 20 miles, one in February and one in March. I am running twice a week, playing tennis 2 to 3 times a week and going to the Gym 3 lunch times a week. In the New Year the emphasis will be towards more running.”**

Richard and Gail will be organising all sorts of fundraising and sponsorship events in the run up to the big day in April. If you would like to support Richard, you can sponsor him by visiting his special sponsorship web page at: [www.justgiving.com/richbenn](http://www.justgiving.com/richbenn). Here you will be able to make your sponsorship pledge securely on line with a credit card. Better still, tell all your friends and send them Richard's web page address by email, so they can sponsor him too.

Alternatively you could support Richard by sending a cheque made out to The Scleroderma Society to Steve Holloway, Secretary, 80 St Agnes Place, Chichester, PO19 7TU. Please write **London Marathon** on the back of your cheque. If you are a UK tax payer we can increase the value of your donation by claiming Gift Aid from the taxman. If you wish to do this, please put your **full name and address** on the back of your cheque and write **Gift Aid**.

**Good Luck Richard!**

## LIVING WITH SCLEROMYXEDEMA

### *THE IMPACT IT HAS HAD ON MY LIFE*

Scleromyxedema is a rare progressive disorder that mimics scleroderma and is characterized by widespread thickened skin and multi-system disease secondary to mucin deposition in the skin and internal organs. I was diagnosed 15 years ago by biopsy of the papules that I had on my hand, Scleromyxedema was confirmed. I was told of its rarity and of an unknown future. I was prescribed various treatments and felt like a guinea pig. Treatments ranged from Melphalen, high dose steroids, plasma phoresis and both Alpha and Beta interferon. Topical treatment was also prescribed by way of a tube of cream which I discovered was given to men with genital warts.

Psychologically over fifteen years the disease has been difficult to cope with. When my body started “growing” very visible papules, these were hidden by my clothes. When the papules started covering my face and neck emotionally it was a very different matter.

As well as this, smiling, opening my mouth, moving my face, moving my tongue and swallowing solid food became impossible. Painful muscles and severe fatigue were, and to a certain extent still are, a problem. - Psychologically, just being rare is daunting enough!

Raynauds is also a problem but three-monthly infusions of Iloprost have helped to relieve this aspect.

In 2005 I carried out some research on the internet and came across the John Hopkins Scleroderma Center in Baltimore (USA), which has patients with Scleromyxedema and discovered how the use of Immunoglobulins (IVIG) by regular infusions has changed their lives.

In May 2006 my Consultants agreed to let me have a trial of IVIG. After just one treatment my face started to be more supple and after the second month's treatment I started eating solid food for the first time in five years! By the time I had received the third monthly infusion the papules had cleared from my face and arms and I understand with regular infusions the disease will be kept under control.

I now feel like a new person and would like to conclude by saying during the fifteen years of coping with a rare disease my Consultants and medical staff have always treated me with kindness and done all they could to alleviate my illness. IVIG has changed my life and I cannot thank my Consultants enough for allowing me to have the opportunity of a more normal life. Thank you. Ann Harris.

### **FEEDBACK FROM APOM 2006**

The 9<sup>th</sup> Annual meeting of APOM (Arthritis People on the Move) took place in Rome over the weekend of 25<sup>th</sup> November 2006. This is the autumn meeting of the EULAR user groups, or ‘Social Leagues’ and nearly everyone present was someone with personal experience of arthritis as well as a representative role in a national user group. The equivalent of Arthritis Care in Italy, called ANMAR, hosted the event and presented on their work.

ARMA, as the UK Social League, was represented by Kim Fligelstone and Steve Mc Bride; while ARMA Chair/Arthritis Care Chief Executive Neil Betteridge attended as Chair of PARE Manifesto, the EULAR Social Leagues’ campaigning project.

In recent years this two day conference has focussed on one or two major themes as a means of unifying the activities of the user group network. As such, the topic of ‘Health Economics’ was a major theme of this year’s event, in recognition of the fact that policy makers often need ‘hard’ incentives to do more for people with musculoskeletal conditions.

The event had several interactive sessions but also some ‘keynote’ presentations including:

**David McDaid**, UK Health Economist, who urged delegates to consider the broader impacts of a cost/cost savings, e.g. not only in health but also areas such as work, transport, leisure activities etc.

**Amye Leong** from the U.S.A., a representative of the global Bone and Joint Decade, who discussed collaboration in campaigning.

**Professor Tony Woolfe**, from the Royal Cornwall, who is involved in EULAR and the Bone and Joint Decade. This principal session focussed on getting the most from your consultation with a health professional.

Amye Leong and Professor Tony Woolfe gave some useful pointers on how to make best use of a consultation by role play, highlighting the best/worst use of limited time restraints. “Be prepared” was the overwhelming message, a useful example was to keep a strict daily record to monitor activity between

appointments and condense this information before the next appointment, which may show patterns of disease activity. Research any treatments that you wish to discuss before hand and finally always recap what action is to follow, i.e. treatments, tests, follow up appointments etc.

Neil presented on the work of Arthritis Care and ARMA, specifically on encouraging UK governments to develop national strategies and in ARMA's Standards of Care. There was a high level of interest from all parts of Europe in the England and Wales strategies and in the original and pending Standards of Care. Ideas for future themes to be used for World Arthritis Day were pooled with favourite suggestions including looking at relationships and sex when you have arthritis. The theme for World Arthritis Day on 12<sup>th</sup> October 2007 is: "Small Things Matter" which is the title for the Edgar Stene Prize. The deadline for submission of essays for this prestigious prize is January 31<sup>st</sup> and all entries are to be sent to ARMA by this date.

*Neil Betteridge, Arthritis Care / ARMA / PARE Manifesto, Kim Fligelstone, Scleroderma Society / ARMA*

**5 December 2006.**

This Essay is the winner of the  
2005 Edgar Sterne Prize.  
It is by Moonika King of Basel  
Switzerland.  
*See above*

## MY TIREDNESS – AN ENCOUNTER

At first ... there was the morning stiffness. The doctor said I had juvenile polyarthritis. I did not know much about it, I just knew that I was not the same as before ... And some time later ... the tiredness arrived. It was not easy to repeatedly tell my friends that (once again) I could not go dancing or to a party ... And now? They are both there. It's not that they are both always there, and

that my life comes to a stop. Not always. But they always come back. It could be described as an "advancing standstill", as Martin Walser writes in "Mesmer's Travels".

Today it is back. I'm tired again. So tired that I cannot lift my arm. It's no good, I must lie down. Eyes closed ... Everything is heavy ... I am like a stone. My limbs can hardly move. I lie there, eyes shut ... And then suddenly I see it! Yes – a small figure, with something sad about it ... Two small, red eyes. Pale face. Very, very strange. I open my eyes – what was it? So alive, so real! Oh, I was probably half asleep and dreaming ... I close my eyes again. It's there again!!! It IS real, there's no doubt.

"Who are you then?" I ask curiously but carefully.

The figure directs her two small red eyes towards me and answers quietly: "I am your tiredness. You know me well enough." ... Yes, I can sense her disappointment, even a degree of reproach.

"Oh, YOU are my tiredness," I say rather unsurely. I am actually quite pleased about this encounter and hope to learn more about her. I therefore ask: "How did you come to me, how did you become MY tiredness?"

I could see an ever so slight sparkle in her eyes as she began to explain. She told me how lonely she was and still is (now and again). She said that she felt so misunderstood. That she wanted to share her being, her thoughts and feelings with someone. She explained that she has lots and lots of brothers and sisters who are all like her ... lonely and misunderstood. The great big tiredness family by its very nature needs a human being who is on the weak side. Because only these beings – in her opinion – can understand it ... that's why she came to me, the small, sad tiredness. She came to me, to share her life with ME. And that is what I am doing. Until now without knowing. And now that I have got to know her I become more and more aware that I have already learnt a great deal from her. She has made me aware of very important things. For instance, my relationship with time. My relationship with myself. My relationships with other people. Taking time for myself. Taking time for other people. Slowing down the pace of life ... She has taught me to stay for a while in the slow lane every now and then in order to speed up again later on the motorway of everyday life. Tiredness has also shown me that it is important to "celebrate" the quiet hours – yes, these are my very own sacred hours – I devote this time in my thoughts to dear people, to myself, and to God. And I also think that there are so many people who are accompanied by tiredness but who fail to understand that they are not alone ... Yes, these are the VERY important things which she has taught me. I think that on my part I can help her to bear her loneliness. Apparently it was the right time to get to know her better – especially now, at Christmas – that is a very special present. I have also told her this – "Thank you, my little tiredness!" And – I see in her little sad eyes a ray of happiness, yes even a few tears of joy ... Now I know I am here for my tiredness – she is here for me too. It is our own quite special relationship. Perhaps even a friendship? Yes, I think so – we have a lot that we can tell each other, we have a lot that we can share ...

Moonika King

## FUNDRAISING

**We have had so many donations for Christmas and sadly do not have enough room to list everyone, THANK YOU ALL SO MUCH! Your generosity is much appreciated and enables us to continue to help people with Scleroderma and their families in a variety of ways including: providing information, an advice line, raising awareness and funding research.**

Rosemary Terry has recently sent cheques to the value of £323 which she and her family have raised on behalf of the society.

Ann & Bob Hall have recently paid in £106 from their fund raising efforts.

Pat Aley sent us a cheque for £175.

Thomas Murphy has made a donation of £120.

Medical Student Julia Flint is still sending donations making her total £2,007

Mrs PM Smith and friends play golf with member Margaret Potts in Cumbria and sent us £30

Andrea and Toby Steele took part in the Beaulieu Duathlon. The Duathlon (a relay with a 10K Run 30K cycle and 5K run) went really well and the



money has finally come in and we have raised £237.00.

“The T-shirts fitted really well and I hope I am raising the profile as when I go out running I wear the T-shirt. The

picture is of Toby and me at the end of the race. We both agreed that if dad (Bill Storey who had scleroderma) were here he would have thought it very amusing, as I always said 'I don't do running'”.

### THE GREAT SOUTH RUN



On Sunday 22 October 2006, mother (me) and daughter (Dionne) team joined 17,998 other mad people to wheeze our way around Portsmouth for the Great South Run (10 miles....phew!). Having been diagnosed with

Systemic Sclerosis 2 years ago and being so grateful

for the wonderful support I get from my family (especially Dionne for doing this with me) and friends and the Society itself, this was a golden opportunity to raise funds for the Scleroderma Society and to raise awareness of the disease. It was a great challenge personally, as prior to the start of all the hard training, neither of us had completed a race of this length before.

Finally the day dawned and we made our way to the event, suitably attired in our Scleroderma T-shirts, shivering with nerves and a lot of “oh dear what have we done” kind of thoughts... but soon settled down as the atmosphere was tremendous. The weather, however, was not kind to us and turned out to be the worst ever. From the first moment the race started, the rain fell (lightly at first) and then proceeded to pour down heavily after a very short time, along with a howling wind, for the rest of the way. We were running really well (despite the conditions) and enjoying the whole atmosphere, supporters along the way were fantastic (and very damp!), when unfortunately at the 5 mile mark, disaster struck with a dreaded knee injury. Being absolutely determined to finish, hobbling or no hobbling and being completely drenched and freezing cold, we managed to complete the whole 10 miles. We raised a total amount of £575 for the Scleroderma Society, which is absolutely fantastic. Dionne and I are extremely grateful to all of our sponsors/supporters for giving us the motivation and determination to achieve our goal. Thank you so very much.

Finally, I would just like to mention that the on-line fundraising method via Justgiving.com, is an excellent way of raising funds and has proved to be very successful in this instance. My thanks go to the Society for making this possible. Lynn Morton

Monica Watson held her annual fundraising event for Scleroderma in memory of her sister in law. This year the event took place at Monica's beautiful house in Horton. At least 30 to 40 of Monica's friends came to lunch and raised £1428. Thanks to June Gardner's friend, Mrs Joan Randall, who requested and received £200 from the Hornchurch Lodge 5790 in Essex. Our thanks to all involved.

Yvette Sagers sent another cheque for £200 making the total raised for her handmade cards £710. Yvette has asked us to thank all her customers. Please keep your orders coming. To contact Yvette Tel: 01768 894057 or 07984831221

## Ask The Expert

Q. My Father had a very aggressive form of systemic sclerosis and I recently had an ENA Blood test which proved positive for lupus antibodies. I have since had further blood and organ tests which are negative. Could you comment please?

A. It is common to have a connective tissue antibody in a close relative of a patient with scleroderma or any of the other connective tissue conditions. It reflects "background" susceptibility but is extremely unlikely to develop into disease as multiple other triggers are needed.

Q. What is the likely outcome and expected survival with systemic sclerosis?

A. This largely depends on which organs are involved. In the past patients died more often from renal complications but we are much better able to treat patients with this complication nowadays. Renal crisis occurs more often in patients with diffuse cutaneous systemic sclerosis in the first two years after presentation, so by educating patients to report the early warning signs we can save their kidneys by initiating early treatment. Pulmonary hypertension which occurs in isolation in some patients with limited cutaneous systemic sclerosis carries a poor prognosis, but again we have much better treatments for this and by monitoring our patients with yearly lung function and echocardiogram we are able to treat them early. For the vast majority of patients the outlook is good. So that we can usually be reassuring and treat them for their circulation problems and bowel symptoms with well tried treatments.

Q. Are plaques in Morphoea a sign of inflammation and increased blood flow? Is there a difference between new and old plaques?

A. New plaques of morphoea generally tend to show increased blood flow as part of the inflammatory response so they look red and can be itchy and uncomfortable. Thermography, which is used to detect temperature changes related to increase blood flow, can be useful to monitor these lesions as far as their activity is concerned. Older more mature lesions tend to look silver or brown and may fade altogether.

## Nurse Specialist Advice Lines



**Rona McIvor**

Woolmanhill Hospital, **Aberdeen**  
01224 555 403

**Sue Brown**

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**Karen Walker (Scleroderma)**

Freeman Hospital, **Newcastle Upon Tyne**  
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**Rachael Crackett & Julia De-Soyza (Pulmonary  
Hypertension)**

Freeman Hospital, **Newcastle Upon Tyne**  
0191 213 7418

**Liz Wragg**

Hope Hospital, **Manchester** 0161 206 0192

**Sally Smith**

General Infirmary, **Leeds** 0113 3922189

**Steve McSwiggan**

Ninewells Hospital, **Dundee** 01382 633957

**Royal Free Hospital, London**

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Lung Enquiries only - **Ross Ellis**

**Brompton Hospital, London** 020 7352 8121

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**Sally Marsh**

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The Nurse Specialist Advice Lines are part funded by the Raynaud's and Scleroderma Association. **Please Note:** These advice lines are for general inquiries. Unfortunately it is impossible for any health professional to comment on individual treatment without seeing the patient.

If there is no reply, please leave a message with your telephone number.