welcome.

It gives me great pleasure to welcome you to "Scleroderma Connections", an innovation of the Australian Scleroderma Interest Group (ASIG).

A diagnosis of scleroderma (a.k.a. systemic sclerosis) can present a difficult challenge for patients and health-care providers alike. The multitude of troublesome and at times, life-threatening symptoms can be overwhelming. But in recent years, there have been exciting developments in scientific research, resulting in better understanding of the factors underlying this disease and leading to therapeutic advances. Pulmonary arterial hypertension (PAH) and interstitial lung disease (ILD) are good examples of complications in which recent studies have demonstrated improved outcomes with new therapies or in the case of ILD, appropriate use of older treatments. A key message arising from these studies is that even greater improvements can be achieved by early detection and treatment.

Regular screening for PAH and ILD is now recommended, a point stressed at the 1st World Congress, held in Florence earlier this year.

Since its inception in 2006, ASIG has grown to include 12 centres around Australia, each co-ordinated by a group of doctors, nurses and research assistants dedicated to the care of patients with scleroderma and the related disease, mixed connective tissue disease. Each centre participates in the Australian Scleroderma Screening Programme (ASSP): a comprehensive clinical programme designed to facilitate annual screening of these patients whilst contributing to an ever-growing web-based database and decision-support tool. Since November 2007, 914 people have been enrolled. This valuable resource provides a wonderful opportunity for a plethora of research studies. There is no doubt that the clinical assessments and data entry can be time-consuming but it is gratifying to see the high quality data analyses emerging. This is due in no small part to the efforts of the centres but also to the IT Department at St Vincent's Hospital in Melbourne which hosts the database and especially to the patients who attend for tests, fill in forms and wait in clinics for consultations. ASIG thanks you all.

Designed in a concise, easy-to-read format, "Scleroderma Connections" will bring you the latest news about ASSP, ranging from handy hints for using the on-line database to research findings. Feature articles will provide pithy updates on what's new in scleroderma and in a special version for patients, experts will give practical advice on how to manage common problems arising in scleroderma. Future editions will include a profile of one of the screening centres and proposed future studies. If you would like to find out more about the centre nearest you or even setting up your own centre, contact details are included on the back page. Finally, I would like to thank our wonderful project co-ordinator, Dr Jill Byron, who makes all our ideas reality, including this newsletter.

Whether you have scleroderma, work in a screening centre or are just interested in what's happening in scleroderma, I hope you enjoy the first issue.

Susanna

Susanna Proudman
ASIG Chair

ASIG
AUSTRALIAN SCLERODERMA INTEREST GROUP

contact.

Australian Scleroderma Screening Centres

WESTERN AUSTRALIA

ROYAL PERTH
Janet Roddy & Madelynn Chan
Ph: 08 9224 1310

SOUTH AUSTRALIA

ROYAL ADELAIDE
Susanna Proudman
Ph: 08 8222 5190

QUEEN ELIZABETH
Catherine Hill
Ph: 08 8222 6688

QUEENSLAND

SUNSHINE COAST RHEUMATOLOGY, MAROOCHYDORE
Peter Nash & Louisa Voight
Ph: 07 5443 1033

NEW SOUTH WALES

JOHN HUNTER, NEWCASTLE
Gabor Major & Glenn Reeves
Ph: 024921 3000

ROYAL NORTH SHORE
Les Schrieber
Ph: 02 9926 7351

ST GEORGE SYDNEY
Allan Sturgess
Ph: 02 9113 2670

ROYAL PRINCE ALFRED
Peter Youssef
Ph: 02 9515 9337

AUSTRALIAN CAPITAL TERRITORY

CANBERRA RHEUMATOLOGY
Kathie Tymms
Ph: 0437 595 334

VICTORIA

ST VINCENT'S
Wendy Stevens
Ph: 03 9288 3983

MONASH MEDICAL CENTRE
Joanne Sahhar
Ph: 03 9594 3566

TASMANIA

MENZIES RESEARCH INSTITUTE
Jane Zochling
Ph: 03 6226 7776

This ASIG publication is supported through an unrestricted educational grant from Actelion

Australian Rheumatology Association
www.rheumatology.org.au

ASIG
AUSTRALIAN SCLERODERMA INTEREST GROUP

SCLERODERMA connections

PATIENT NEWS

EDITION 1 : MAY 2010

tips.

40% of ASIG scleroderma patients report ulcers on their fingers or toes...

Digital ulcers are a common problem for patients with scleroderma. Barbara Gemmell, the Scleroderma Nurse at St Vincent's Hospital has provided the following information and tips.

Prevention should be the aim – keep warm to maintain good circulation

- Dress warmly and keep your environment as warm as possible.
- Use disposable or reusable hand warmers that can be put in your pocket.
- Wear gloves – woollen out in the weather, cotton at night or under rubber gloves.
- Wear rubber gloves when putting hands in water.
- Keep skin moist by using a good moisturising cream regularly.
- If you have had problems with ulcers in the past talk to your doctor about medication to dilate the blood vessel in your fingers.

At least 13% of these patients were hospitalised due to digital ulcers...

Good management supervised by a health professional from the outset is crucial:

- Painful fingertips may be relieved by covering with an adhesive film dressing such as Tegaderm.
- Digital ulcers due to necrotic (dead) tissue eventually form a tough scab (or "eschar"). The scab should be softened with a gel such as Flaminal or Wound Aid (as they have added ingredients that help keep infection at bay) and use a dressing such as Tegaderm applied over the top until the scab is soft. Then cover with Tegaderm alone until the body's normal healing forms normal skin over the tip.
- For very painful or exposed areas, stronger non-stick dressing such as Melonin or an adhesive dressing such as Mefix or Hyperfix over the above may protect the finger tip/knuckle from further trauma/damage
- Watch out for signs of infection such as ulcer increasing in size, redness and pain in the skin around the ulcer or smelly discharge from the ulcer. If any of these signs occur please contact your health care professional for urgent advice.

1st World Scleroderma Congress

FLORENCE 2010

DR WENDY STEVENS

The first world Scleroderma Congress was held in Florence, Italy in February this year. The Congress was attended by over 1000 doctors interested in scleroderma and it was opened by Dame Carol Black who gave a review of the history of scleroderma research and highlighted the rapid growth in interest and research into this difficult illness that has occurred in the last 30 years.

There were over 500 abstracts at the meeting and for our relatively young group to be selected by the organising committee to present 2 oral presentations as well as our 2 posters was a great honour and an indication of the quality of our research project. The two oral presentations were on the Incidence of PAH in the Australian database, and RNA polymerase antibodies and their clinical association in our cohort. Both presentations were very well received and our national database received a lot of interest. We were commended on our database and research project and several groups expressed an interest in collaborating with us.

Research Highlights:

Dan Furst presented the five year follow-up from the scleroderma lung study which was a placebo controlled 12 month trial of cyclophosphamide in scleroderma lung disease. At 5 years:

- 30% of patients enrolled in the trial had died. Unfortunately they have not been able to track the cause of death in all patients and these data were not presented.
- 19% have been lost to follow-up.

Of the 108 who could be traced for subsequent analysis,

- Five have developed a malignancy
- 11% now require home oxygen therapy
- 2% have had lung transplants.

Poor prognostic factors were male sex, being Afro-American, having Scl70 or RNP antibodies and extent of disease being more than 20% on HRCT scan at enrolment.

Janet Pope presented her experience with imatinib, a tyrosine kinase inhibitor that has anti-fibrotic effects *in vitro*. Her trial, which was a placebo-controlled study, has now been ceased after enrolling only 10 patients. All patients had severe diffuse skin disease with a mean duration of 3.2 years and mean Rodnan skin score of 23. The main reason for ceasing the trial was that the therapy was very poorly tolerated with oedema being one of the most difficult side effects.

ROBYN SIMS

Robyn Sims, Vice President Scleroderma Australia, was asked by Scleroderma Australia to represent the association at the Scleroderma World Congress. She gave the following report.

On the 11th of February I attended the opening ceremony held at Auditorium of the Congress Centre, Florence. On first entering the Auditorium I was overwhelmed by the number of doctors and scientists who were attending this congress. Apparently there were over 1300 doctors from the four corners of the world, England, Ireland, Italy, Spain, Hungry, Netherlands, USA, Canada, Germany, China, France, Brazil and many from Australia. Professor Carol Black addressed the meeting giving an overall history of scleroderma and the prospects for the future. Professor Black mentioned that organisations such as ours were vitally important and that she had seen so much progress made around the world in the number of these patient associations and their willingness to work together, such as FESCA (the Federation of European Scleroderma Association).

Dr. T. Medsger another very experienced clinician and scleroderma researcher closed this opening Lecture with the very heartening words that he felt confident that in the next ten years there will be more progress made into the cause and cure of scleroderma than in the past fifty years.

There followed for these doctors and scientists three days of presentations. In all there were 322 presentations on subjects such as Musculoskeletal & Gastrointestinal system. Basic Immunology pathways, Pulmonary Artery Hypertension, Cardiovascular, Raynaud's Phenomenon and Ulcers, as well as scientific studies which as a layperson I was unable to grasp. Many different topics were covered during question and answer time, such as stem cell therapy, oral problems, incontinence etc.

We here in Australia have many dedicated specialists who speak at our seminars, and it was encouraging to hear that we are well up to date with all that is happening in other countries and our patient seminars compare favourably with those in Florence.

Maureen Worron-Sauve, President of the Scleroderma Society of Ontario gave an interesting presentation from an extensive survey of over 600 patients and the results of this survey give us a good indication of where our energies need to be put. It appears that fatigue was one of the main complaints from those surveyed and yet this aspect of the disease is often ignored. Of course, this is a very difficult symptom to treat. Diagnosis time was also interesting, as it seems to be rapidly improving from time of consulting a general practitioner to having a correct diagnosis.

There was an excellent opportunity to exchange ideas, listen to others, and to make contact with committee members from many other scleroderma associations. I deemed it a privilege to attend the conference and an exciting opportunity for our organisation to be recognised in the Northern Hemisphere. I wish to thank Scleroderma Australia for this wonderful opportunity and the sponsorship of Actelion for my airfares.

ASIG database: Incidence of PAH

Following is a summary of the oral presentation given by Dr Wendy Stevens on Pulmonary Arterial Hypertension in Australian Scleroderma Patients.

Of the 841 patients who had enrolled in the ASIG database project at 1st Jan 2010 a total of 108 were found to have PAH. 34 had already been diagnosed with PAH prior to their enrolment in the project and 74 have been diagnosed as a result of the screening project.

PAH was seen in equal frequencies in patients with diffuse and limited scleroderma, confirming the evidence that screening should be encouraged in both sets of patients. Patients with diffuse scleroderma had shorter disease duration at the time of diagnosis of PAH, with mean of 11 years when compared to those with limited scleroderma in whom the mean disease duration at diagnosis of PAH was 15 years. 33% had some associated interstitial lung disease.

Annual screening that includes Echo and lung function enables patients to be monitored according to their risk stratification.

RISK STRATIFICATION

High Risk PAH	sPAP on echo ≥ 50 mmHg	All should have RHC even if no symptoms
Moderate Risk PAH	sPAP 40–50 mmHg or DLCO ≤ 50% with FVC > 85%	6 min walk test and consider RHC if any symptoms of Dyspnea. Repeat echo in 6/12 if echo if no RHC
Lower Risk of PAH	sPAP 30-40 mmHG	RHC if symptoms of Dyspnea
Lowest Risk of PAH	sPAP ≤ 30	If unexplained Dyspnea consider RHC

RESULTS OF THE 127 PATIENTS IN THE DATABASE REFERRED FOR A RHC

PAH at rest	59 (46%)
PAH on exercise	15 (12%)
Raised mPA with raised wedge	6 (4.7%)
Normal	47 (37%)