ASIG

AUSTRALIAN SCLERODERMA INTEREST GROUP

SCLERODERMA connections

EDITION 4: NOVEMBER 2011

SSC SCREENING CENTRE UPDATE

greetings,

The original motivation for establishing the Australian Scleroderma Screening Program was to provide a service to people with scleroderma and their doctors, for screening for the serious complication of pulmonary arterial hypertension (PAH).

But why all the fuss about PAH?

Timely identification of PAH can be complex, particularly in a busy rheumatology practice. The "one stop shop" visit to a screening centre for tests and clinical assessment aims to take the worry out of this process. While an elevated pulmonary arterial pressure on an echocardiogram is a strong predictor of PAH, this often occurs late in the disease, so detection relies on a combination of other factors including reduced diffusing capacity on lung function tests and reduced exercise tolerance. Unfortunately, people with scleroderma can have a poor exercise capacity for so many reasons, making it difficult to unravel the cause of their symptoms. The primary goal of the screening program is to increase rates of detection of PAH in its early stages. In addition, the spin-off Australian Scleroderma Cohort study is driving multiple research studies, some of which aim to find better markers of PAH in its early stages and which are summarised elsewhere in this newsletter.

These are worthy objectives indeed, but in all the complexities of screening tests and data collection, we must not lose sight of the individual caught in the vortex of all this activity, who is being poked and prodded, while all the while hoping that a right heart catheter (required for diagnosis of PAH) is never recommended and if it is, that PAH is never detected. Undoubtedly, this can be a serious and debilitating complication and a source of much anxiety but it is not all bad news. De-mystifying some of the issues around PAH is one of the important components of managing this disease.

Firstly, 90% of patients with scleroderma will never develop PAH. It is only because better non-invasive methods for early detection are yet to be developed, that international guidelines currently recommend annual screening. Newer therapies have measurable benefits in this condition. Furthermore, it appears likely that earlier detection and treatment leads to improved survival so there is much to be optimistic about. While there has been a growing and rewarding collegiality between the various specialists who treat this disease – cardiologists, respiratory physicians and rheumatologists – the "glue" which holds the PAH clinic together is the PAH nurse. In the patient version of this issue, Leah McWilliams, PAH nurse at the Royal Adelaide Hospital, the centre featured in this issue, provides helpful advice for the new patient with PAH. The clinical support and pastoral care provided to people with PAH and scleroderma by these dedicated PAH nurses is without doubt, second to none.

Susanna

Susanna Proudman
ASIG Chair

World Scleroderma Day

This day occurs in June. It is a good opportunity to educate health professionals and the wider community about scleroderma.

This year, at St Vincent's Hospital the scleroderma team held an information session for staff across the hospital. Presentations were given by Dr Wendy Stevens and Barbara Gemmell focusing on the clinical aspects of patient care, and the fellows Dr Vivek Thakkar and Dr Owen Moore gave an overview of their research projects. It was well attended and included nurses from the wards and from in-home care, technicians from the respiratory clinic and others interested in learning about the disease.

The lunch sponsored by Actelion was much appreciated.

profile.

A/Prof Susanna Proudman and Leah McWilliams

Royal Adelaide Hospital

Leah McWilliams began training as a nurse at the Royal Adelaide Hospital in 1987. She writes, "On completing my training, I secured a permanent position as a registered nurse in an orthopaedic/ rheumatology ward and soon became a clinical nurse. Travel beckoned, resulting in twelve months of backpacking in America, Europe and South Africa. On returning to Adelaide, I completed a Bachelor of Nursing degree at Flinders University and found myself back at the RAH in the Orthopaedic/ Rheumatology Outpatient department. It was here I was approached by A/ Prof Susanna Proudman to take up the position as clinical studies manager for the Rheumatology Unit.

I embarked upon a very steep learning curve, managing a clinical trial in patients with early rheumatoid arthritis and another in pulmonary arterial hypertension. I was also enlisted by the cardiology and respiratory physicians to assist with Medicare applications for highly specialised drugs for PAH. The staff weren't quite sure what a rheumatology nurse was doing in their OPD, but were welcoming, interested and accommodating.

The scleroderma screening program grew fast and we now have over155 patients enrolled, so my feet didn't really hit the ground till a new cardiac nurse came on board early this year.

My main role is to be a first point of contact for patients, answer questions, provide educational support, liaise with pharmacists, GP's, specialist physicians, community nurses and research staff. I prepare and collate patient files to help streamline patient visits, perform joint examinations and complete lots of paperwork! I provide support to the clerical staff by making sure appointments are as convenient as possible for patients. Often I catch up with patients admitted to the wards, but the aim is to provide enough 'accessible' support to keep admissions to a minimum.

It is A/Prof Susanna Proudman's everyday example that is a privilege to be near and the quiet bravery of our patients that makes working at the RAH so extraordinary.

A career highlight was receiving a 'Commendation award' on Susanna's nomination for services to 'Safety & Quality' within the South Australian Health Service; oh and having a patient name his race horse 'Leah' after me!"

Office: 8222 2746 Mon-Thurs
8222 4000 pager 1429

Often in clinics in the mornings, best time to catch me is afternoons in the office

ASIG research
Blood Samples

Now that the ASIG group has the data collection well established, greater effort is being put into the blood sample collection. It will be the linking of disease data with blood markers and genetic information that will provide the most valuable insight into the disease. We ask that patients provide blood for DNA (usually a couple of samples are taken to ensure that the quality is acceptable for research) and then annual sera samples. By collecting the sera annually we are able to investigate links between changes in sera and the onset of disease complications.

With so many samples now in storage and several studies underway that have required the retrieval of stored samples, we are looking at the development of a database to track these. You'll hear more about this soon.

ASIG Research Output

With longitudinal projects it can be several years before it is possible to publish the results. ASIG will be presenting several abstracts at the second World Scleroderma Congress to be held in Spain next February.

Our PhD fellow, Dr Vivek Thakkar, submitted three abstracts on behalf of the group. They were:

1. "N-Terminal pro-Brain Natriuretic Peptide (NT-proBNP) levels predict incident pulmonary arterial hypertension in systemic sclerosis (SSc)".

In the absence of LV dysfunction, NT-proBNP is a useful screening biomarker for PAH in SSc, with levels>189.2 pg/ml and <82.9 pg/ml defining patients with a high and low likelihood of PAH, respectively. Further prospective studies are required in unselected patients in order to confirm these findings.

2. "Serum ICAM-1 levels are related to the presence of interstitial lung disease in systemic sclerosis".

ICAM-1 (intercellular adhesion molecule-1) levels are associated with significant SSc-ILD. However, ICAM-1 level does not appear to be a specific marker for the presence of PAH. VCAM-1 (vascular cell adhesion molecule-1) levels are raised in SSc patients but are not characteristic of any particular phenotype. Further studies of ICAM-1 in SSc-ILD are warranted.

3. "Novel biomarkers of dysregulated angiogenesis are not specific to pulmonary arterial hypertension in systemic sclerosis".

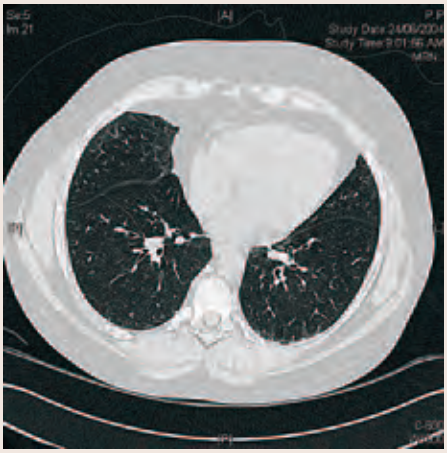
We did not find an association between serum IL-6, IL-13, FGF-2, VEGF, fractalkine and SSc-PAH. While these factors may play a role in the pathogenesis of SSc and SSc-PAH, their serum levels do not appear to correlate with clinical PAH.

St Vincent's Rheumatology Fellow, Dr Owen Moore, has been working on an ILD project with ASIG. He submitted two abstracts on behalf of the group. He has provided a summary of his research and initial findings:

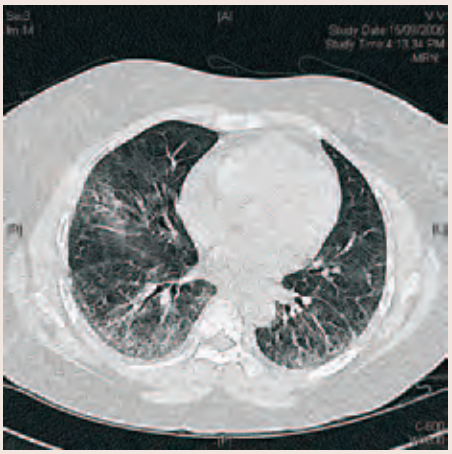
Interstitial lung disease (ILD) is a leading cause of mortality in SSc. It has been proposed that a simple grading system of total disease on high-resolution CT (HRCT) lung be used to determine prognosis in SSc-ILD. The scoring system examines the extent of disease and categorises; >20% - extensive, <20% - limited, unclear - indeterminate. Indeterminate results were converted to limited or extensive using an FVC threshold of 70%. We collected retrospective data from the ASIG database and patient notes. Scans were collected at St Vincent's Hospital, Melbourne where they were read by two respiratory physicians and one radiologist.

Our findings have confirmed that this semi-quantitative grading system for extent of lung disease on baseline HRCT has prognostic significance in SSc-ILD. Extensive changes (>20%) on HRCT are associated with a five-fold increased likelihood of deterioration or death compared with limited changes.

Further analysis using a time-varying covariate survival model sought to establish factors that predict deterioration and mortality in SSc-ILD over time. This method establishes that, though extent of lung disease on HRCT at baseline is predictive of deterioration and death in SSc-ILD, serial HRCTs performed over time add no prognostic information. In contrast, serial FVC and DLCO are significantly predictive of outcome and offer prognostic information in follow-up of patients with SSc-ILD.



"Limited" lung disease



"Extensive" lung disease

Dr Jane Zochling's abstract entitled "An immunochip based interrogation of scleroderma susceptibility variants" has also been accepted.

Additional Research

One manuscript has been submitted to a journal and one is in the final stages of writing. We'll keep you posted on their progress.

Further information about ASIG can be found at:
http://rheumatology.org.au/rheumatologists/asig-public.asp

ASIG
AUSTRALIAN SCLERODERMA INTEREST GROUP

contact.

RESEARCH QUERIES

Jill Byron, Project Manager
Ph: 03 9288 3986 Email: jill.byron@svhm.org.au

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