

FESCA news

summer 2013



New EULAR recommendations for SSc care needed



The current recommendations for the treatment of systemic sclerosis (SSc) were published in 2009, and included results of systematic literature search only up to the end of Feb 2007. Since then new drugs have become available, while others have been withdrawn from the market. Important new information has been published regarding treatment. Search of PubMed from Mar 2007 to Dec 2012 reveals 129 hits related to randomized clinical trials in SSc. Development of new EULAR recommendations is underway.

The DeSSciper project has begun



On April 1st 2013, the observational trials of the international EU-funded research project DeSSciper began. Currently the largest SSc project ever, DeSSciper will over

3 years cover different evolutionary phases of the disease from early manifestations such as digital ulcers and hand arthritis to more severe organ involvement such as interstitial lung disease, pulmonary hypertension, and heart disease. Patients can participate by asking their doctors to check out (www.dessciper.eu) or writing to dessciper@kerckhoff.med.uni-giessen.de

EUSHnet meets for kickoff at EULAR



To improve non-pharmacological care and reduce inequity through an international network of

SSc-specialising health professionals, EUSHnet complements the work of FESCA and the EUSTAR medical network. Activities will kick off at the EULAR congress, where national champions selected by EUSHnet will meet to discuss the diffusion of physical, psychosocial and educational therapies for SSc. Their aims are to improve expertise and knowledge of HPs, facilitate communication between them, and establish standards of care and rehabilitation.

FESCA holds successful AGM in Porto

FESCA held its AGM in Porto this year, where although the meeting ran all day on April 6, there was time also to enjoy the beauties of the city. Dinners were networking events, since nearly all the 23 associations were represented and new national representatives were introduced from Denmark, Finland, Norway, and both the UK associations.



Don't forget that we celebrate SSc Day worldwide on June 29th

Across the world, FESCA's European Scleroderma Day has now become World Scleroderma Day, using the same iconic Paul Klee painting as its symbol. Conferences, publicity campaigns and awareness-raising events of all sorts take place as the scleroderma community demonstrates its unity by celebrating together. This year, many are using the Wear Your Gloves campaign, for which the Scleroderma Society in the UK has created PR materials freely shared with anyone who also wants to join in. Next year's theme will be "Art and Scleroderma" so we will look for artists who have SSc in all countries.



Mark your Calendars for the World Congress in Rome

The Third World Systemic Sclerosis Congress will be held in Rome in 2014 **from Feb 6-8**. Actually two concurrent congresses, one will be for doctors, with hands-on sessions with experts and patients as well as lectures. The other will be a two-day programme for people who have scleroderma, based on the success of the last congress in Madrid. The Patient Programme for the Third World Systemic Sclerosis Congress will consist of interactive sessions of lecture and discussion, with consultants and patients speaking on topics including:

- Why did I get scleroderma and will I be cured?;**
- The lungs—why is breathing so difficult?;**
- What can I do to help digestion processes?;**
- What can be done about digital ulcers?;**
- Transplants of different kinds—are they for me?;**
- Face and hand exercises; Sexuality and Pregnancy; Dentistry in SSc.;**
- New treatments; Carers and family-members;**
- Fatigue and how to deal with it; Useful tips for daily life; and**
- How can I cope emotionally?**

Two exclusive sessions will address small-group needs:

Men with Scleroderma, and **Juvenile Scleroderma**. The patient congress will also include a round-table networking meeting for leaders of patient groups.

IMPRESS 2 Study of Pregnancy in Systemic Sclerosis

An interesting new project is underway for patients with systemic sclerosis who plan a pregnancy. Initiated by EUSTAR, this international observational study, IMPRESS 2 (International Multicentric prospective study on PREgnancy in Systemic Sclerosis) will follow women before, during and after pregnancy, and hopes to answer a number of important questions that remain open, such as whether pregnancy alters the course of SSc in the short or long term. Women whose doctors are not yet aware of this study can request that they join by contacting FESCA or the following doctors: Monika Østensen, National Center of Pregnancy and Rheumatic Disease, University of Trondheim, Norway monika.ostensen@gmail.com ; Angela Tincani, Rheumatology, Università degli Studi di Brescia, Italy tincani@bresciareumatologia.it ; Antonio Brucato, Internal Medicine, Ospedale Papa Giovanni XXIII, Italy albrucato@hpg23.it

New treatment being investigated for systemic sclerosis

Systemic sclerosis is a chronic autoimmune disease characterized by connective tissue fibrosis, involving organs such as the skin, heart, kidneys and lungs. Interstitial lung disease (ILD) is one of the main manifestations of SSc, and the varying degrees of pulmonary inflammation and fibrosis can lead to restrictive lung disease.

Progressive SSc with ILD (SSc-ILD) can result in a 10-year mortality rate of about 50%¹ with approximately 30% of SSc deaths attributed to pulmonary fibrosis.²

While the DeSSciper³ global research project includes observational trials⁴ with the goal to improve our understanding of the SSc condition and patients' quality of life, there is still unmet need for disease and/or organ-specific treatments for systemic sclerosis. The good news is that a Phase II trial has been initiated to investigate CC-4047 (pomalidomide) overall efficacy and safety in patients with SSc-ILD. CC-4047 has demonstrated potent antifibrotic effects in a preclinical skin fibrosis model and the investigators, led by Dr Jörg Distler of the University of Erlangen-Nuremberg, Germany, concluded that these data support clinical studies in SSc.⁴

The trial, entitled "*A Phase II, Proof-of-Concept, Multicenter, Randomized, Double-Blind, Placebo-Controlled Study to Evaluate the Safety, Tolerability, Pharmacokinetics, Pharmacodynamics and Efficacy of CC-4047 in Subjects with Systemic Sclerosis with Interstitial Lung Disease*" [NCT01559129]⁵ is designed to assess the safety and efficacy of CC-4047 treatment in SSc-ILD patients. Since initiation, there are now 47 active sites across 10 countries: Australia, France, Germany, Italy, Poland, Russia, Spain, Switzerland, the United Kingdom and the United States.

The target is to enroll 88 SSc-ILD patients over 1 year at approximately 55 sites. The progression of this potential therapy into Phase II is very encouraging, particularly in a disease which is so rare and for which there are no treatments available.

If you are interested in participating in the trial, or would simply like to find out more, please visit <http://www.clinicaltrials.gov/ct2/show/NCT01559129>. Alternatively, please contact FESCA or Eustar (<http://www.eustar.org>).

References:

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