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relevant information in the management of patients with scleroderma-spectrum

Recently, an international survey on non-invasive techniques to assess the microcirculation performed under the aegis of members of the European League Against Rheumatism (EULAR) Study Group on Microcirculation in Rheumatic diseases (SG\_MC/RD) showed that nailfold capillaroscopy was the one most used technique in both clinical and research settings by adult physicians and paediatric rheumatologists to assess patients with Raynaud's phenomenon.

A number of different instruments are available to perform the exam. They have different characteristics in terms of their cost, quality of images, magnifications, training period, portability, software for image analysis and storage.

Some of these instruments can be used both in clinical and research settings such as the stereomicroscope and the videocapillaroscope. The stereomicroscope allows the widefield visualization of the nailfold with low magnifications, the training is relatively short, but the examination is difficult to perform in patients with digital flexion contractures.

There appears to be consensus regarding the use of videocapillaroscopy that allows a detailed visualisation of capillary morphology using higher magnifications (100-300x). Contact probe with polarized light microscopy permits easier observation of the skin surface, and the training period is briefer. Specific softwares are available for images analysis, storage, and complete medical reports (text + images) can be produced.

By contrast, in a clinical setting, nailfold capillaries can generally be visualised using more simple but also efficient tools such as a dermatoscope. USB microscope, ophtamloscope or smartphone device. The quality of images can be quite good, although the lower magnification means that some details are unlikely to be seen, and they often lack the possibility of image storage and measurement. In particular, the dermatoscope with magnification of the order of x10 is a small, inexpensive and easily portable piece of equipment that has been suggested to be comparable to videocapillaroscopy in routine clinical practice.

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#### SATURDAY, 17 JUNE 2017

# Workshop: strengthening your organisation - how to manage volunteers \_

# SP0172 MANAGING VOLUNTEERS- A UK PERSPECTIVE

C.B. Jacklin. External Affairs, National Rheumatoid Arthritis Society, Maidenhead, United Kingdom

Volunteers are an integral part of any charity and it would be impossible to run a charitable organisation without the support of volunteers. Like paid staff they need to be trained, nurtured and rewarded but as volunteers they need to be handled in a very different way to employees.

People who volunteer do so for many differernt reasons and not always perhaps for the right reasons so managing volunteers takes great skill and diplomacy. My talk will cover how to value volunteers, lessons we have learned from managing volunteers over many years as well as how to manage issues with volunteers and the lessons I've learned from my mistakes!

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# SP0173 THE CHALLENGES OF A SMALL ORGANIZATION

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When a group of citizens establishes a non-profit and a non-government organisation in our country, those volunteers are carried by great enthusiasm. At the beginning when founding an NGO the main problems are lack of experience and financial resources. Those deficiencies can be overcome by some other qualities such as the personal competencies of volunteers.

As NGOs are seen by the public rather critically in our country, our organization had to face several additional challenges. In my presentation I will illustrate the following aspects: the non-attractiveness of NGOs for volunteers, the lack of awareness how volunteering is important for a society, the lack of knowledge how to attract volunteers and how to manage them, the lack of knowledge how to define the volunteers' positions and how to monitor their work, the lack of their systemic, continuing education and the lack of rewards, recognition and appreciation to acknowledge the most dedicated volunteers.

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## SP0174 WAYS OF SUPPORTING VOLUNTEERS

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The promotion of volunteer workers is an indispensable part of the human resources policy of self-help organizations and requires specific concepts. Using the example of the rheumatism league Baden-Württemberg -an organization with 65,000 members, 3,000 volunteers and 10 fulltime employees- there will be shown best- practice examples.

A successful concept covers the areas of recruitment, training, support and integration.

The support of volunteers should include four key areas:

- 1. Transfer of knowledge and professional competences
- 2. Individual support for personal development
- 3. Promotion of teamwork and social skills
- 4. Framework conditions (insurance cover, reimbursement of expenses)

The implementation of these requirements should be carried out by specifically trained volunteer managers on the basis of a strong and motivating personal relationship.

This strategy can be seen as a precondition for establishing a long-term relationship of the volunteer with the association, a successful local work, satisfied members and volunteers who perceive their work as satisfying and fulfilling.

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#### SATURDAY, 17 JUNE 2017

### WIN & HOT session —

## SP0175 WHAT IS NEW IN JUVENILE IDIOPATHIC ARTHRITIS

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Juvenile Idiopathic Arthritis comprises 7 subcategories. As the insights in pathogenesis progress so does the need for reclassification that is based more on biology than on clinical phenotypes. After a series of clinical trials for new biologicals, now trials are started that test specific treatment strategies such as treat to target an d step down studies. Especially rapid induction of remission is currently a major aim, followed by biomarked guided tapering of medication.

The expending number of potential biomarkers forms the basis for the creation of personalized medicin, a strategy aimed at providing individualized medication choices. Since most pediatric rheumatic conditions are rare, international collaboration is vital. The recently created European Reference Networks (ERN) will prove instrumental here

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## SATURDAY, 17 JUNE 2017

## Systemic sclerosis \_

## SP0176 TARGETING VASCULOPATHY FROM THE BEGINNING

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In systemic sclerosis (SSc), the natural history of microvascular damage progresses from capillary dilation to capillary loss and reactive angiogenesis, as detectable by nailfold videocapillaroscopy (NVC) [1]. The process is systemic and determines multiple clinical manifestations, from the early appearance of Raynaud's phenomenon, trough formation of digital ulcers (DUs), until severe organ involvement, impairing patient's quality of life or leading to main death causes, including interstitial lung disease and pulmonary arterial hypertension (PAH), hearth involvement, scleroderma renal crisis [2,3]. Although microvascular and macrovascular abnormalities frequently coexist in disease such as diabetes mellitus and other vascular diseases, the possible association between microvascular failure and macrovasculopathy in SSc patients has not been deeply investigated. However, significant correlations seem to exist between increased Intima-Media Thickness (IMT) of periferic small-caliber arteries (macrocirculation) and altered peripheral BP (LASCA) at the level of hand microvessels (microcirculation) in SSc patients.

Siln addition, sgnificant capillary loss, observed at NVC, is peculiar of the "Late" scleroderma pattern of microangiopathy and is mainly preceded by progressive capillary enlargement, microhemorrhages and their collapse, leeding to presence of large avascular areas [4]. The importance of capillary loss was already demonstrated by a simple and reliable prognostic index, capable to predict digital trophic lesion development in SSc-related microvascular disease, when evaluated as part of the semi-quantitative NVC scoring [5]. Moreover, microvascular function and its alterations in SSc, can be reliably assessed by laser-doppler flowmetry (LDF) and laser speckled contrast analysis (LASCA), evaluating blood perfusion at fingertips or at larger body areas [6-9].

The most frequently used drugs for treatment of complications in SSc patients, approved with evidence grade la, are vasoactive drugs. In particular, for severe Speakers Abstracts Saturday, 17 June 2017 43

RP and for prevention of DUs onset and treatment of PAH, both intravenous prostanoid iloprost (ILO), and the dual endothelin-1 receptor antagonist (ERA) BOSE are used, respectively [10]. Bosentan seems to block pathogenic activities of Endothelin-1, the endothelial derived mediator determining both vasoconstriction and also the fibrosis genes induction [11-14].

Previous long-term follow up studies, showed that treatment with BOSE in combination with ILO interferes with progression of nailfold microvascular damage, evaluated through both capillary number semi-quantitative scoring at NVC and fingertip blood perfusion (FBP) at LDF [15,16].

In particular, an open label, prospective study of 4 follow up years showed that long-term treatment with BOSE added to ILO infusions administered quarterly in SSc patients, exerts a remodelling effect on structural and functional microvascular alterations and on stabilization of lung function, compared to ILO mono-therapy [17].

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## SP0177 NEW APPROACHES BY TARGETING SOLUBLE MEDIATORS

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Although the aetiopathogenesis of systemic sclerosis (SSc) remains incompletely understood there is now sufficient knowledge about the pathobiology of the disease and mechanisms underlying development of fibrosis, autoimmunity and vasculopathy to permit informed selection of candidate soluble mediators that may be important drivers of the disease. This has permitted testing of potential targeting approaches in preclinical models and in vitro tissue culture systems. The data from these studies has informed understanding of the disease and has started to be translated into clinical trials that test hypotheses in vivo in patients. This approach has in turn fueled the concepts of reverse translation that are being applied to further understand SSc mechanisms in model systems and observational cohort studies. Soluble mediators that have emerged as strong targets for therapeutic intervention include TGFbeta superfamily members, connective tissue growth factor, IL13, IL4 and IL6. Studies of agents that target these proteins have been proposed or undertaken. Targeting TGF-beta appears to have benefit for skin thickening and attenuate some of the characteristic TGF-beta regulated genes and proteins in skin. However, the most encouraging data have emerged from targeting the IL6 axis using anti-IL6R neutralizing antibodies. This approach was promising in a Phase II study and further trials are ongoing. Interestingly this approach appears to attenuate macrophage gene expression signatures in the skin, may prevent worsening of lung fibrosis and showed significant benefit in the skin by biomarker analysis and a strong trend of benefit over 48 weeks for skin sclerosis score. Other small clinical trials testing TNF-alpha blockade have been more disappointing. Finally, it is possible that soluble mediators may be identified that directly attenuate the fibroproliferative mechanism of SSc. Experimental studies have suggested that increasing alpha-MSH may have benefit for skin fibrosis in a small controlled clinical trial. Clinical trial design is critical to the success and interpretation of studies targeting soluble mediators to define target engagement, mechanism of action, and possible clinical benefit. Advances in trial design are facilitating progress and it seems likely that, as in other rheumatic diseases, targeting key soluble mediators will become a therapeutic reality of the next few years. Defining the place of such approaches compared to other emerging treatment strategies will remain challenging.

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#### SATURDAY, 17 JUNE 2017

## How diet influences musculoskeletal diseases

#### SP0178 GUT DYSBIOSIS AND OTHER CHALLENGES PRECIPITATE **ARTHRITIS**

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Environmental factors contribute to development of autoimmune diseases. For instance, human autoimmune arthritis can associate with intestinal inflammation, cigarette smoking, periodontal disease, and various infections. The cellular and molecular pathways whereby such remote challenges might precipitate arthritis or flares remain unclear. We have defined many of the pathways in the gut that contribute to homeostasis, particularly GPCRs such as GPR43 and their ligands the short chain fatty acids. Such receptors and their ligands are anti-inflammatory. To probe peripheral inflammation connections to arthritis, we used a transfer model of self-reactive arthritis-inducing CD4 cells from KRNtg mice that, upon transfer, induce a very mild form of auto-inflammatory arthritis in recipient animals. This model enabled us to identify external factors that greatly aggravated disease, such as microbiota effects or disruption of gut homeostasis. We show that several distinct challenges precipitated full-blown arthritis, including intestinal inflammation through DSS-induced colitis, and bronchial stress through Influenza infection. Both triggers induced strong IL-17 expression primarily in self-reactive CD4 cells in lymph nodes draining the site of inflammation. Moreover, treatment of mice with IL-1b greatly exacerbated arthritis, while transfer of KRNtg CD4 cells lacking IL-1R significantly reduced disease and IL-17 expression. Thus, IL-1b enhances the autoaggressive potential of self-reactive CD4 cells, through increased Th17 differentiation, and this influences inflammatory events in the joints. We propose that diverse challenges that cause remote inflammation (lung infection, colitis, gut dysbiosis) result in IL-1b -driven Th17 differentiation, and this precipitates arthritis in genetically susceptible individuals. Thus the etiology of autoimmune inflammatory arthritis likely relates to diverse triggers that converge to a common pathway involving IL-1b production and Th17 cell distribution.

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## SATURDAY, 17 JUNE 2017

# Can targeting disease activity in hand osteoarthritis improve our treatment in the 21st century -

# SP0179 WHAT IS DISEASE ACTIVITY IN FINGER OA?

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Osteoarthritis of the interphalangeal finger joints constitutes one of the most prevalent musculoskeletal diseases with variable clinical impact ranging from nearly asymptomatic to severe inflammatory pain in and around affected joints, presence of soft tissue and bony swelling, stiffness and gradual loss of function. Current therapeutic options are limited to analgesic treatment but research on targeted therapies is increasing.

In order to improve the current treatments, a critical appraisal of the needs for improvement in finger OA is needed and how disease activity is best defined. Assessing disease activity or joint activity in finger OA is challenging: disease activity can comprise pain, inflammatory activity and structural damage.

In clinical practice, a combination of patient-reported (e.g. visual analogue scale or Likert scale pain), more objective and performance-based measures (e.g. grip strength) are used to assess and follow disease activity. In clinical research, pharmacological trials and epidemiological studies, a standardized approach to assess the disease activity is necessary to estimate the burden of disease and to evaluate the efficacy of potential new treatments. The instruments being used, depend mostly on the aim of the study or research question. In case structural disease progression is being studied, imaging-based outcome measures are mostly used. Structural changes can be assessed on joint level or on patient level, both by conventional radiographs, ultrasound and magnetic resonance imaging. Several imaging based outcome measures and scoring systems are being suggested but true consensus about the instruments of preference is still lacking. Therefore, further validation of these instruments is warranted.

This lecture will give an overview of the current instruments to measure activity in finger OA in several domains and discuss its strengths and limitations.

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#### SP0180 BIOLOGICS AND OTHER INFLAMMATORY THERAPIES IN FINGER OA. WHAT HAVE WE LEARNED?

X. Chevalier. Dept. of Rheumatology, University Paris XII, UPEC, Créteil, France

Hand osteoarthritis (HOA) is the most frequent form of osteoarthritis. A subset of digital hand OA is marked by an "inflammatory" like presentation with painful interphalangeal joints, refractory to NSAIDS and analgesics.