0.71, inter-observer mean of 8.64 and within-patient standard deviation (SD) of 4.25. For the second group of assessors who returned 2 days after training (n=14), compared to the experts' scores, the inter-observer and intra-observer variability was 0.73 and 0.85 respectively. The inter-observer mean was 7.39 with a within-patient SD of 3.65. The intra-observer mean was 6.92 and within-patient SD was 2.73.

Conclusions: There was substantial inter-observer reliability and excellent intra-observer reliability. This is the first study examining the training of assessors using the SCTC training guidelines and our results support the importance of standardised teaching for mRSS.

Disclosure of Interest: None declared

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SAT0481

SETTING THE STANDARD FOR LONGITUDINAL FOLLOW-UP OF SYSTEMIC SCLEROSIS: A EUSTAR DELPHI-BASED EXPERT CONSENSUS

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Background: Systemic sclerosis (SSc) is a severe multi-organ disease associated with substantial morbidity and mortality. Lung and heart involvement are currently the major causes of disease-related deaths. Skin, gastrointestinal and musculoskeletal involvement, digital ulcers and Raynaud's phenomenon have shown to be associated with high morbidity, reduced quality of life and lower social functioning. SSc is progressive and many of the disease features aggravate over time, while other features may commence during the disease course. However, to date, there are no established standardised international guidelines for follow-up of SSc patients.

Objectives: The aim was to establish an expert consensus regarding the longitudinal systemic assessment of organ involvement in SSc to improve the standard-of-care for SSc patients.

Methods: All experts in SSc from the European Scleroderma Trials and Research Group (EUSTAR) network and the scleroderma clinical trial consortium (SCTC) were invited to participate. The final expert panel consisted of a multidisciplinary team including rheumatologists, dermatologists, pulmonologists, cardiologists and nephrologists. The Delphi method was Internet based and completed from December 2016 until October 2017. The method entailed the entire group of experts who anonymously replied to in total 5 online questionnaires. The experts were asked to score each item in the survey to answer the following question: "Which domains and tools do you strongly suggest for the minimum annual systemic investigation of SSc patients". Every item in every questionnaire was asked to be rated between 0% and 100%, with 100% as 'very important/appropriate' and 0% as 'not important/appropriate at all'. Parameters rated >80% by more than 80% of the experts were rated as acceptable in all steps.

Results: Of the 269 invited centres, physicians from 132 (49.1%) centres participated in the DELPHI survey of 5 steps. Of the included participants, 71.3% were seeing >50 SSc patients annually and 48.3% of the centres seeing >100 patients on an annual basis. Of all, 98 of the centres were located in Europe (74.2%), 18 in North America (13.6%), 7 in Asia (5.3%), 5 in South America (3.8%) and 4 in Oceania (3.0%). In the first round, 23 domains were suggested by the expert panel. After the second Delphi step, 10 domains were included (figure 1). In the third round, tools for each domain were received. The tools were included in the fourth step and rated by all participating experts. The tools for each of the 10 domains that were rated appropriately by all experts were included in the last step of the DELPHI survey and were re-rated. The final tools for each domain are shown in figure 1 and can be seen as the collective opinions of the convened expert panel.

Abstract SAT0481 – Table 1. Overview of the tools for each domain

1. Domain: Raynaud's phenomenon
Investigation: Frequency and severity of the attacks
2. Domain: Skin and nails
Investigation: Skin (increasing or regressing) (qualitative reported)
Clinical assessment: Nail fold capillary (NFC) count, extent of skin thickening (distal, proximal, trunk, face), telangiectasia (yes), calcinosis (yes), modified Rodan skin test
3. Domain: Musculoskeletal
Investigation: Muscle weakness and stiffness (yes)
Clinical assessment: Joint and finger extensor (grip) strength (yes), joint contractures (yes), tender/swollen paraspinal joints (yes), calcification (yes), tendinopathy (yes), osteoarthritis (yes)
4. Domain: Respiratory
Investigation: Dyspnoea (yes), cough (yes), development of new rales during the last year (yes), no existing condition (no), asthma (yes), smoking status (yes)
5. Domain: Lung
Investigation: Dyspnoea (yes)
Functional assessment: Functional class (NYHA I-IV)
Clinical assessment: Spirometry (yes)
Investigation: Lung function test
6. Domain: Heart
Investigation: Dyspnoea (yes), increasing heart disease (yes)
Functional assessment: Functional class (NYHA I-IV), heart rate, blood pressure
Clinical assessment: ECG (yes)
Investigation: ECG (yes)
7. Domain: Gastrointestinal
Investigation: Upper and/or lower gastrointestinal (yes), dysphagia (yes), diarrhoea (yes), weight loss (yes)
Clinical assessment: Weight
8. Domain: Kidney
Functional assessment: Blood pressure
Investigation: Serum creatinine, eGFR, renal analysis, proteinuria
9. Domain: Laboratory
Investigation: Serum albumin, creatinine, CK, hematology, renal function test, liver function test
10. Domain: Treatment
Investigation: Type of treatment, date of initiation, date of discontinuation of drug usage

Conclusions: Through five Delphi rounds with world leading experts in SSc, an expert consensus was established on strongly suggested tools for a minimum

longitudinal systemic assessment of organ involvement in SSc to improve the standard-of-care for patients with SSc.

Disclosure of Interest: None declared

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SAT0482

UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE AT RISK FOR SYSTEMIC SCLEROSIS: PREDICTIVE ROLE OF ANTI-TOPOISOMERASE AND AVASCULAR AREAS

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Background: Undifferentiated connective tissue disease at risk for systemic sclerosis (UCTD-risk-SSc) is a condition characterised by Raynaud's phenomenon and either SSc marker autoantibodies or typical capillaroscopic findings or both, unsatisfying classification criteria for SSc¹ and reported to evolve into definite SSc in about 50% of 60 cases over a 12–102 months follow-up time.² We found marker autoantibody positivity to predict the evolution into SSc satisfying 2013 ACR criteria for the disease.³

Objectives: To investigate in patients followed-up for a longer time if distinct marker autoantibody specificities have a different predictive value.

Methods: Sixty-five patients consecutively admitted to a tertiary Rheumatology Unit from November 1 st 2000 to December 31 st 2016 and diagnosed as UCTD-risk-SSc were enrolled in the study. Patients were monitored for a median of 27 months (range 6–144) and were evaluated twice yearly to assess disease progression. Kaplan-Meier curves and the log-rank test were used to analyse differences in fulfilling the criteria for SSc between subsets. Risk prediction was assessed by univariate Cox regression analysis.

Results: During follow-up 40/53 marker autoantibody-positive patients (75.5%) versus 3/12 (25%) marker autoantibody negative ones satisfied SSc criteria (p=0.006). Out of them, 11/12 (91.7%) anti-topoisomerase (Scl70) positive versus 29/40 (72.5%) anti-centromere (ACA) positive patients evolved into definite SSc (p=0.04). In univariate analysis, anti-Scl70 positivity increased by 2-fold the risk of a definite SSc outcome (HR 2.1 95% CI 0.9–4.4) with respect to ACA positivity (HR 0.5 95% CI 0.2–1.0) (p=0.05). In addition 3/3 (100%) patients with avascular areas at baseline versus 40/62 (64.5%) with megacapillaries only or no capillaroscopic abnormalities satisfied SSc criteria over a 12–38 months follow-up time (p=0.06).

Conclusions: We confirm that autoantibody positivity patients presents a faster evolution. Moreover we first detected an increased HR of Scl-70 versus ACA positivity and a potential role of baseline detected avascular areas.

REFERENCES:

- Valentini G. Undifferentiated Connective Tissue Disease at risk for systemic sclerosis (SSc) (so far referred to as very early/early SSc or pre-SSc). *Autoimmun Rev* 2015.
- Valentini G, et al. Early Systemic Sclerosis: Analysis of the Disease Course in Patients With Marker Autoantibody and/or Capillaroscopic Positivity. *Arthritis Care Res* 2014.
- van den Hoogen F, et al. 2013 classification criteria for systemic sclerosis: an American college of rheumatology/European league against rheumatism collaborative initiative. *Ann Rheum Dis* 2013.

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SAT0483

FEMALE SEXUAL DYSFUNCTION IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease leading to various physical and psychological impairments including sexual dysfunction.

Objectives: To assess sexual functions/quality of life and pelvic floor function in female SSc patients compared to age-/sex-matched healthy controls (HC), and to analyse the potential impact of disease activity, fatigue, physical activity and depression.

Methods: In total, 41 women with SSc (mean age: 50.9, disease duration: 5.8 years, lcSSc/dcSSc: 18/23, mRSS: 13.6, ESSG activity index: 2.5), who fulfilled the ACR/EULAR 2013 criteria, and 41 healthy controls (mean age: 50.9) without rheumatic diseases filled in 12 well-established and validated questionnaires assessing sexual function/quality of life, pelvic floor function, fatigue, physical