emphasize the importance of an accurate joint examination in these patients. The OSd group showed more joint manifestations which might be explained by the coexistence of SLE and MCTD patients in this group. Currently, no association between the clinical subtypes of IIM, overall, these results are encouraging and suggest that joint assessment in follow up may be helpful in differentiating subtypes of IIM.

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FRI0386 THE VALIDATION OF THE SCLERODERMA HEALTH ASSESSMENT QUESTIONNAIRE IN TURKISH SPEAKING PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: The Health Assessment Questionnaire (HAQ) is one of the main instruments for assessing disability in rheumatic diseases.The Scleroderma HAQ (S-HAQ) combines the HAQ with five systemic sclerosis (SSc) related visual analogue scales for Raynaud's phenomenon (RP), digital ulcers, digestive symptoms, pulmonary symptoms, and overall disease severity.
Objectives: To perform cross-cultural adaptation and validation of the S-HAQ in patients with SSc.
Methods: Sixty patients who fulfilled the 2013 ACR/EULAR Classification Criteria for Systemic Sclerosis ACR criteria for SSc were were recruited.Fifty three percent of the patients fulfilled the criteria for limited systemic sclerosis (lcSSc).We evaluated test–retest reliability using the intraclass correlation coefficient (ICC); known-groups construct validity by stratifying patients according to severe organ involvement; and convergent validity using Spearman's correlation with mental and physical components of Short Form 36 version 2 (SF36v2).
Results: Eighty percent of the patients were female and the median age was 56 years old.Finger tip ulcers were observed in 16 patients (26.7%) and severe organ involvements were detected as intersititial lung disease in 32 (53%) and pulmonary hypertension in 5 (8%) of the patients.HAQ Disability Index (HAQ-DI), digestive VAS, pulmonary VAS, digital ulcer VAS, Raynaud's phenomenon VAS and overall disease severity VAS demonstrated high reliability (ICCs=0.819–0.908).The HAQ-DI showed higher correlation coefficients with physical function and role physical components of physical-related scores (r=-0.537 and – 0.453, respectively); social function and role emotional components of mental-related dimensions (r= -0.470 and -0.328, respectively) in the SF-36.Two among five SSc related VAS (VAS digital ulcer and VAS overall disease severity) were highly correlated with HAQ-DI (r=0.527 and 0.481 respectively).The instruments could not discriminate between severe organ involvements and both subsets of SSc.

Table 1. Correlation of HAQ-DI with physical and mental components of SF-36 and, SSc related VAS scores

N=60	Mean±SD and/or %	Correlation Coefficient	*p-value*
Physical function	55.4±23.83	-0.537	0.000
Role physical	33.82±42.67	-0.453	0.001
Body pain	53.82±28.84	-0.290	0.041
General health	39.9±18.43	-0.295	0.037
Vitality	41.08±18.39	-0.286	0.044
Social function	62.25±24.49	-0.470	0.001
Role emotional	49.02±32.9	-0.328	0.02
Mental health	61.12±18.39	-0.295	0.04
VAS digestive	0.39±0.65	0.304	0.028
VAS pulmonary	0.64±0.79	0.380	0.005
VAS RP	0.67±0.74	0.354	0.01
VAS digital ulcer	0.62±0.94	0.527	0.000
VAS overall severity	1.14±0.97	0.481	0.000

Conclusions: The Turkish version of the S-HAQ meet the requirements of reproducibility and validity.More studies are needed to examine the capacity of these instruments to discriminate between severe involvements and disease subsets.This study forms the basis for future studies to evaluate the properties of S-HAQ in Turkish speaking SSc patients more extensively.

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FRI0387 ANTI-RNA POLYMERASE III SUBSET OF SCLERODERMA PATIENTS: A MONOCENTRIC STUDY

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Background: Systemic sclerosis (SSc) is an autoimmune disease characterized by heterogeneous clinical features and variable disease course. Specific antinuclear antibodies (ANA) identify different clinical subsets and are very useful in defining the prognosis of the patients. Recently, the anti-RNA polymerase III antibodies (anti-RNAP) were defined as the third specific ANA of SSc, together with anti-centromere (ACA) and anti-topoisomerase I (anti-Scl70).
Objectives: The aim of this study was to analyze the clinical picture of a group of SSc patients with anti-RNAP in a monocentric cohort of patients and to compare it with other subsets of patients with different specific ANA. In particular, the visceral involvement and the concomitant malignancies were investigated.
Methods: Among the SSc patients referring to the Rheumatology Unit of Padova University, 49 cases with anti-RNAP specific ANA were considered. Patient's demographics data and major clinical manifestations were compared with those of 50 ACA-positive and 52 anti-Scl70-positive patients. The skin score and the presence of digital ulcers, arthritis, interstitial lung disease (ILD), cardiac involvement including pulmonary hypertension, gastrointestinal disease (GI) and scleroderma renal crisis (SRC) were assessed. The presence and type of concomitant malignancies were also evaluated.
Results: 53% of anti-RNAP-positive patients had a diffuse cutaneous form of SSc. The skin score was significantly higher than in ACA-positive patients ($p<0.001$). Digital ulcers were active in 73.5% of cases, without significant difference with the other two subsets. Arthritis was found in 20.4% of cases in comparison to 4% in ACA-positive patients ($p<0.02$). Regarding visceral involvements, GI disease was the most common manifestation in all subsets; SRC was observed in 24.4% of anti-RNAP-positive patients and was significantly more frequent in comparison to ACA-positive ($p<0.005$); ILD showed a prevalence of 42.8% in anti-RNAP subset, lower than in anti-Scl70-positive ($p<0.05$) and higher than in ACA-positive patients ($p<0.01$); cardiac involvement was observed in 32.6% of anti-RNAP-positive patients, without significant difference with the other subsets. Malignancies were found in 33 (67.3%) anti-RNAP-, 18 (36%) anti-Scl70- and 7 (14%) ACA- positive patients. The most common cancer-sites were breast (36%), lung (14%) and colon (10%). The risk of developing cancer was higher in anti-RNAP-positive patients than in other subsets (*OR*:6.35).
Conclusions: Our data demonstrated that anti-RNAP specific antibodies can identify a subset of SSc patients characterized by a severe clinical picture, with a high prevalence of diffuse cutaneous form, SRC, cardiac involvement and ILD. These patients showed also an elevated risk of developing cancer. Our results were consistent with recent published papers on this topic (1,2).

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FRI0388 LONG-TERM TREATMENT WITH RITUXIMAB IN SYSTEMIC SCLEROSIS PATIENTS: UPDATE OF OUR CLINICAL EXPERIENCE

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Background: Systemic sclerosis (SSc) is an immune-mediated disorder characterized by abnormal fibrosis and diffuse microangiopathy with skin and internal organ involvement. The treatment of SSc represents a great clinical challenge because of the complex disease pathogenesis including vascular, fibrotic, and immune T- and B-lymphocyte-mediated alterations. Therefore, SSc should be treated by combined or sequential therapies according to prevalent clinico-pathogenetic phenotypes. Some preliminary data suggest that rituximab (RTX) may be usefully employed in SSc patients.
Objectives: The present study aimed to evaluate the efficacy of RTX in our SSc patients' series as well as the long-term effects of this treatment.
Methods: A series of 15 SSc patients (M/F 6/9, mean age 52.7±17.9SD years, mean disease duration 10.3±7.1SD years, L/D cutaneous subsets 5/10) were treated with one or more cycles of RTX (4 weekly infusions of 375 mg/m^2). In all patients RTX was repeated every 6 months for a total of 2–6 cycles. Patients' clinical-serological evaluation, including the self-evaluation of quality of life by means of HAQ and visual analogical scale (VAS) assessment, was performed every 6 months for a mean follow-up period of 42±24SD months.
Results: After the first 6 months following RTX treatment the extent of skin sclerosis measured with modified Rodnan skin score (mRSS) significantly improved (from 17.3±10.4 to 13.4±7.6; $p<.01$), and remained stable at the end of the follow-up (13.3±8.1; p=.009). The usefulness of RTX on skin sclerosis was more evident in patients with diffuse cutaneous SSc (n=10) showing a significant decrease of mRSS after the first 6 months (from 24.2±5.1 to 18.1±4.7; p=.006) and at the end of the follow-up period (18.0±6.0; p=.005). Similarly, a valuable improvement of other cutaneous manifestations, namely hypermelanosis (12/12 pts), pruritus (11/13 pts), and calcinosis (3/6 pts) was observed. Moreover, arthritis revealed particularly responsive to RTX treatment leading to a clear-cut reduction of swollen and tender joints in 12/13 patients; while lung fibrosis detected in