Risk factors for death. Univariate and multivariate analysis

	Univariate analysis HR (95% CI)	p-value	Multivariate analysis HR (95% CI)	p-value
Age at first symptom	1.08 (1.07–1.09)	<0.001	1.07 (1.06–1.09)	<0.001
Gender (female)	0.40 (0.29–0.54)	<0.001	0.55 (0.38–0.78)	0.001
Disease subset				
lcSSc	0.57 (0.45–0.72)	<0.001	–	–
dcSSc	2.74 (2.14–3.50)	<0.001	2.33 (1.74–3.12)	<0.001
preSSc	0.05 (0.00–0.62)	0.020	–	–
Mode of onset				<0.001
RP	0.44 (0.33–0.58)	<0.001	1 (ref.)	–
Puffy Hands	2.18 (1.19–4.00)	0.011	2.11 (1.07–4.16)	0.031
Arthralgia	0.92 (0.53–1.61)	0.779	0.76 (0.40–1.45)	0.407
Skin sclerosis	2.23 (1.45–3.42)	<0.001	1.43 (0.88–2.33)	0.150
ILD	11.34 (4.97–25.85)	<0.001	3.97 (1.58–9.97)	0.003
PAH	31.00 (11.21–85.75)	<0.001	13.92 (4.79–40.5)	<0.001
Others	2.94 (1.51–5.75)	0.002	1.83 (0.85–3.96)	0.123
ILD	2.60 (2.01–3.35)	<0.001	1.90 (1.37–2.65)	<0.001
PAH	2.60 (2.01–3.35)	<0.001	1.64 (1.25–2.15)	<0.001
SRC	4.08 (2.64–6.30)	<0.001	1.94 (1.14–3.31)	0.014
Heart involvement	2.40 (1.89–3.05)	<0.001	1.38 (1.05–1.80)	0.020

Conclusions: The mode of onset can be considered as independent prognostic factor, and specifically non-RP onset can be considered as a risk factor related to poorer outcome and RP onset and arthralgias can be considered as protector factors.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.1675

FRI0403 INCREASED CRP LEVEL IS INTERRELATED WITH CLINICAL AND SEROLOGICAL PHENOTYPES OF SYSTEMIC SCLERODERMA AND ASSOCIATED WITH CRP RS1205 GENE POLYMORPHISM

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Background: Recent studies report on increased serum levels of C-reactive protein (CRP) in systemic scleroderma (SSc) pts.

Objectives: The current study was aimed at identifying potential correlations between CRP levels, immunologic SSc phenotypes and CRP rs1205 gene polymorphism in the Russian cohort.

Methods: 92 SSc pts were enrolled in the study. Mean age ($\pm$ SD) was 49.4 $\pm$ 12.6 years, mean ($\pm$ SD) SSc duration was 11.1 $\pm$ 9.0 years, increased CRP levels >5 mg/l were documented in 43% pts. CRP levels were analyzed in the following subgroups: with limited (lcSSc) and diffuse (dcSSc) types, with SSc duration <3 years >3 , with or without interstitial lung disease (ILD+) and (ILD-), as well as in subgroups with positive antibody titers to DNA topoisomerase I (ATA+) and antibody to centromeres (ACA+). CRP concentrations were measured with highly sensitive immunoturbidimetry method. Allele-specific real time PCR was used to study CRP rs1205 gene polymorphism.

Results: Mean CRP ($\pm$ SD) level was significantly higher in dcSSc vs lcSSc (11.9 $\pm$ 15.5 mg/l; vs 4.7 $\pm$ 7.4 mg/l, respectively, $p=0.006$). Pts with disease duration <3 years had higher mean CRP levels as compared to pts with SSc duration >3 years, although the difference was not significant. In pts with disease duration >3 years mean CRP levels were significantly higher among subjects with dcSSc than lcSSc (11.0 $\pm$ 15.5 mg/l vs 4.8 $\pm$ 7.7 mg/l, respectively, $p=0.027$). Mean CRP level was higher in (ATA+) pts than in (ATA-) pts (11.0 $\pm$ 15.4 mg/l vs 5.5 $\pm$ 13.7 mg/l, respectively, $p=0.032$). No statistical difference in CRP levels was found between (ILD+) and (ILD-) pts, as well as between (ACA+) and (ACA-) pts. Carriers of rs1205 CC genotype had higher CRP levels than carriers of rs1205 T allele, $p=0.087$. In pts with SSc duration >3 years and carriers of rs1205 CC genotype mean CRP level was significantly higher than in T-allele carriers (12.0 $\pm$ 17.4 mg/l vs 5.4 $\pm$ 7.4 mg/l, respectively, $p=0.021$). Similar trends for CRP levels depending on genotypes were established among (ATA+) pts (16.9 $\pm$ 20.1 mg/l and 7.3 $\pm$ 10.5 mg/l, respectively, $p=0.031$).

Conclusions: Increased CRP levels were found almost in 50% of Russian SSc pts. Higher CRP levels were associated with specific clinical and immunological SSc parameters and CRP rs1205 CC genotype, signifying unfavorable prognosis.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2676

FRI0404 THE EFFECT OF A HOME-BASED OROFACIAL EXERCISE PROGRAM ON ORAL APERTURE OF SYSTEMIC SCLEROSIS PATIENTS

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Background: Microstomia is one of the major problems that causes multiple dysfunctions and affects quality of life in systemic sclerosis (SSc).¹

Objectives: The purpose of this study is to evaluate the effect of a home-based orofacial exercise program on the oral aperture (OA) of SSc patients.

Methods: SSc patients who fulfilled the American College of Rheumatology (ACR) 2013 criteria with an OA of <40 mm were included in the study. Patients were randomly divided into two groups. Group 1 was given oral augmentation and mouth stretching exercises with combination of strengthening exercise to masseter muscle twice a day for one month in addition to oral hygiene care advices; followed by no activity but oral hygiene care for the next four weeks. Group 2 received oral hygiene care advices for the first one month followed by the same orofacial exercise program twice a day for the next four weeks. Patients' OA was measured by a dentist at baseline, 1-month, and 2-months.

Results: A total of 52 patients were included in the study. Twenty-seven patients were assigned to group 1 (mean age 53.70 $\pm$ 9.79 years; 96.3% female) and twenty-five patients to group 2 (mean age 50.68 $\pm$ 11.93 years; 96.0% female). A total of 28 patients (group 1, n=18; group 2, n=10) completed the study. After four weeks period, OA increased in group 1 ($P<0.001$), whereas no change was observed in group 2 ($P=0.906$). At the end of 2 months the intragroup analysis showed no additional increase in group 1 ($P=0.283$), but a statistically significant increase in group 2, regarding the measurement of OA ($P=0.010$). We didn't find any statistically significant difference between OA measurements of the groups 1 and 2 at the end of the trial ($P=0.830$).

Conclusions: The results of this pilot study suggested that an intervention of combined home-based orofacial exercise program might improve oral aperture of SSc patients. Future studies with longer follow-up period and higher number of patients are needed to observe the long-term benefits of the presented home-based exercise program.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.6274

FRI0405 PREVALENCE, CLINICAL CORRELATES AND POSSIBLE CAUSES OF NEUROPATHIC PAIN IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Objectives: To assess prevalence, possible causes and clinical correlates of neuropathic pain (NP) in patients with systemic sclerosis (SSc).

Methods: In 42 patients with SSc, presence of NP was assessed using the PainDetect questionnaire and subsequently confirmed by neurological evaluation. Patients with previously diagnosed neurological disorders and diabetes were excluded. Relationship between NP and disease status, symptoms of depression and quality of life was estimated. Index of disease status (IDS) was assessed using the Scleroderma Assessment Questionnaire (SAQ), occurrence of depressive symptoms by the Beck's Depression Inventory (BDI), whilst quality of life was evaluated using the EQ-5D index. In order to evaluate possible causes of NP in SSc, all patients with NP, except one (who did not agree with further assessments) underwent detailed neurological and electroneurography (ENG) examination, as well as HgbA_{1c} and vitamin B12 level testing.

Results: 12/42 (28.6%) of patients were found to have NP. Patients with and without NP were similar in age and disease duration. NP was found more frequently in patients with lcSSc (32.3%), than in dcSSc (18.2%), as well as in patients with ACA (46.7%) compared to patients with ATA (17.6%) and both ACA and ATA negative patients (20%), but the differences were not statistically significant. Patients with NP had significantly higher mean values of IDS (1.28 vs 0.66) and BDI (18.3 vs 9.9) than patients without NP ($p<0.01$). Mean value of the EQ-5D index was significantly lower in patients with NP (0.39 vs 0.79, $p<0.01$). Patients with NP had significantly higher mean value of Index of vascular status, than patients without NP (2.26 vs 1.36, $p=0.02$). HgbA_{1c} and vitamin B12 levels were normal in all SSc patients with NP. Clinical signs of polyneuropathy were noticed in 10 patients by neurological evaluation. Typical symmetric glove-and-stocking hyperesthesia was found in 10/11 (90.9%) of SSc patients with NP, along with symmetric hyporeflexia in 6/11 (54.5%) patients on lower, and 7/11 (63.6%)