

distance (Table 1). Only numerical improvement in IG compared to numerical deterioration in CG, which have not reached statistical significance, were observed in patient reported outcomes (CHFS, MHIS, HAQ, SHAQ, SF-36).

Conclusions: Our physiotherapy program not only prevented the natural course of progressive deterioration of function of hands/mouth (observed in the control group), but led to a significant improvement in monitored parameters, which was clinically meaningful in a substantial proportion of patients.

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FRI0400 EFFICACY OF INTRAVENOUS IMMUNOGLOBULIN THERAPY IN REFRACTORY DYSPHAGIA IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Polymyositis (PM) and Dermatomyositis (DM) are systemic inflammatory diseases affecting skeletal muscles and other organs, particularly hypopharynx and the upper tract of the esophagus, causing motility dysfunction. The current treatment for PM and DM relies on steroids and immunosuppressants, but esophageal involvement is often refractory to standard therapies.

Objectives: To assess the efficacy and safety of intravenous immunoglobulins (Ig-iv) as therapy of refractory dysphagia (1) in patients affected with PM/DM who have received conventional treatments for at least three months. These are the extended data of a previous abstract entitled "Improvement of refractory dysphagia in patients with idiopathic inflammatory myopathies receiving immunoglobulin intravenous therapy" accepted to EULAR 2015 Congress.

Methods: We enrolled 17 patients (15 F), 8 with DM, 9 with PM according to Bohan and Peter criteria, treated with prednisone (mean dose 20 mg/day) ± Methotrexate (15 mg/wk) or Azathioprine (100 mg/day) for at least 3 months, without improvement of dysphagia. All patients received one infusion of Ig at a standard dose (2 g/kg in a day) every two months for 3 cycles. In all patients, at entry and before each infusion, creatine kinase (CK) levels and Manual Muscle strength Test on 12 muscle groups (MMT12) were evaluated. Dysphagia assessment included: Eating Assessment Tool (EAT) impaired for score ≥3; Three-oz Swallow Test (3oST) and fibrolaryngoscopy (FEES) for pharyngeal muscle propulsion and stasis of liquid/solid bolus detection. Student T-test and Chi-square test were used to compare baseline and follow-up data.

Results: At entry, all patients had an EAT score ≥3 with a mean score of 16.2±9, 3 patients presented a 3oST for solid (STPNS) impaired, and only 1 patient also for liquid (STPNL). CK levels were slightly increased (474±720 UI/L), and MMT12 score was 4.45±0.35. Following Ig-iv therapy we observed a significant improvement of EAT score after the first dose (9±8, p=0.006) with a further reduction after the second dose (7.4±8, p=0.007) and stability after the third one (7.6±8, p=0.008). An improvement of pharyngeal muscle propulsion, with a significant resolution of solid stasis and a partial resolution of liquid stasis were observed. In addition, MMT12 score progressively improved after each dose, increasing to 4.86±0.16 after the third dose (p=0.001). Noteworthy, after the third Ig-iv dose, three patients stopped the steroid and the remaining 12 were treated with a significant lower dose of 5 mg/day of prednisone (p=0.04).

Conclusions: According to literature, our findings provided evidence that Ig-iv induced a meaningful improvement of dysphagia, also having a steroid-sparing effect. An improvement of muscle strength was also detected. Although further studies of larger cohort of patients and a longer follow-up time are needed, our findings corroborate the utility of Ig-iv in the treatment of more resistant idiopathic inflammatory myositis related dysphagia.

References:

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FRI0401 INITIAL MANIFESTATION DETERMINES CLINICAL ENTITY AND COURSE IN PATIENTS WITH ANTI-CENTROMERE ANTIBODY: A SINGLE CENTER LONGITUDINAL RETROSPECTIVE COHORT STUDY

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Background: Anti-centromere antibody (ACA) is often detected in sera from autoimmune diseases, such as limited cutaneous systemic sclerosis (SSc) and primary biliary cirrhosis (PBC). ACA-positive Sjögren's syndrome (SjS) is recently

considered as a distinct clinical subgroup in SjS [1-2]. The other autoimmune diseases associated with ACA-positive have not fully elucidated and the role of ACA in their pathogenesis still remains unclear. In addition, comprehensive and enough clinical information from the point of view of ACA positive cases has not been accumulated.

Objectives: To clarify clinical features of patients with ACA and their association with autoantibodies.

Methods: Patients with discrete-speckled pattern in anti-nuclear antibody (ANA) test and/or positive ACA, who visited to our department during over 20 years between May 1995 and December 2016, were enrolled. Clinical information and immunological tests including immunoglobulin (Ig) and serum autoantibodies were collected and statistically analyzed.

Results: Discrete-speckled pattern in ANA test and/or positive ACA were identified in 309 patients. There were 292 female and 17 male and the average age was 60 years old. Proportion of concomitant ANA patterns were speckled (16%), homogenous (7%), cytoplasmic (3%) and/or nucleolar (3%) pattern. 186 patients (60%) had Raynaud's phenomenon and 149 patients (48%) had sclerodactyly. 162 patients (52%) had oral and/or ocular dryness symptoms. 21 patients (6%) had interstitial pneumonia and 13 patients (4%) had pulmonary hypertension. 42 patients (13%) had arthritis. 19 patients (6%) had thyroid disease. 214 patients (69%) were classified into 17 autoimmune diseases (including overlap cases) from symptoms at initial visit, while other 95 patients (31%) who did not meet the criteria were not diagnosed. IgG, IgM and IgA in the patients who were classified into autoimmune diseases were higher than that in other patients who were not classified (p=0.0007, p=0.0057 and p=0.0158, respectively). In the 214 patients (69%), there were most of patients who were diagnosed as SSc (n=113, 36%) and/or SjS (n=78, 25%). 25 patients (8%) and 22 patients (7%) were diagnosed with rheumatoid arthritis (RA) and PBC, respectively. On the other hand, the patients who diagnosed as systemic lupus erythematosus were 3 (1%), and nobody was diagnosed as myositis. The mean observation period was 80 months. 2 patients in 95 patients who were unclassified at initial visit were diagnosed as RA or autoimmune hepatitis by developing new symptoms during observation. A patient was newly diagnosed as SSc in addition to SjS because sclerodactyly was newly developed. In other patients, the diagnosis was not changed and added during observation.

Conclusions: Our single center longitudinal retrospective cohort study confirmed that ACA-positive cases were classified into various types of autoimmune disease. Few patients had new symptoms during observation but the initial diagnosis was unchanged. Our data demonstrated that initial manifestation determined clinical entity and course in patients with ACA.

References:

[1] *J Rheumatol* 2001; 28: 2238.

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FRI0402 ONSET CLINICAL INVOLVEMENT AS PROGNOSTIC FACTOR IN SYSTEMIC SCLEROSIS (SSC)

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Background: In most patients with SSC, the onset is in the form of Raynaud's phenomenon (RP), but in a few is in the form of visceral or musculoskeletal involvement or skin sclerosis. Several risk factors are related to poor outcome in patients with SSC, but never before has the mode of onset been described as one of them.

Objectives: To assess the mode of onset as prognostic factor in patients with SSC.

Methods: A retrospective and multicenter cohort study including 1625 patients with SSC, based on the National Spanish Scleroderma Registry (RESCLE) which includes 28 referral centres.

RP, puffy hands, arthralgia/arthritis, skin sclerosis, interstitial lung disease (ILD), pulmonary arterial hypertension (PAH) and esophageal hypomotility where included as clinical modes of onset.

Results: The cohort was split up in two groups: 1342 patients (83%) presented as RP and 283 (17%) presented as non-RP (see graphic below)

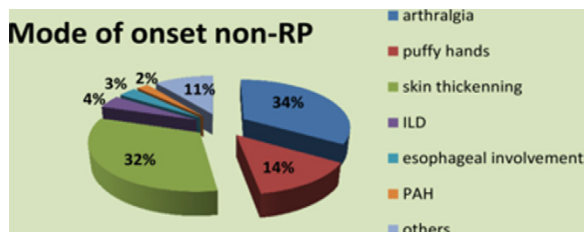
Non-RP patients were younger at onset of disease (44.7±16.5 vs 49±15.6 years, p<0.001), showed higher percentage of diffuse subtype (36% vs 19%, p<0.001), developed more visceral involvement during the course of the illness and where more frequently anti-RNAP III positive (29 vs 10%, p<0.001).

Overall mortality was 16% in patients presented as RP vs 23% in non-RP patients (p=0.003). Few differences were noted among causes of death related to SSC, but the higher mortality was observed in RP group due to PAH (19% vs 8.1%, p=0.05) and due to ILD in non-RP group (24% vs 8.3%, p=0.002).

Survival of patients with RP at onset was better at anytime than those with a non-RP onset: 0.97 vs. 0.89 at 5 years, 0.93 vs. 0.82 at 10 years, 0.83 vs. 0.63 at 20 years and 0.71 vs. 0.51 at 30 years from onset (p<0.001).

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				
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

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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 sclerosis (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

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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 <40mm 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.

References:

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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 24 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 electroencephalography (ENG) examination, as well as HgBA₁C 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, $p=0.02$). HgBA₁C 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%)