gastrointestinal involvement (GI), nutritional status and medications may lead to Vit B12 deficiency.
Objectives: We aimed to investigate the frequency of Vit B12 deficiency and its determinants in SSc patients.
Methods: Sixty-two (90.3% female) SSc patients were enrolled in to the study. The nutritional status of patients was assessed with Malnutrition Universal Screening Tool (MUST). Serum Vitamin B12, homocysteine and Helicobacter Pylori Immunoglobulin G (H. Pylori IgG) levels were measured in all patients. Serum Vit B12 levels of patients were classified as; Low (<200 pg/ml), Borderline (200 - 300 pg/ml) and Normal (>300 pg/ml). Serum homocysteine levels of patients were classified as; Elevated (>9 μmol/L) and hyperhomocysteinemia (>15 μmol/L). H. Pylori IgG antibody level >5 U/ml considered as positive. Serum Vit B12 level <200 pg/ml or being on Vit B12 replacement therapy was considered as B12 deficiency.
Results: The mean age of the patients was 50.2 (12.5) years and mean disease duration was 12.0 (7.5) years. Forty-four (71.0%) patients were limited and 18 (29.0%) patients had diffuse SSc. The mean serum Vit B12 level of the patients was 323.6±291.5 pg/ml. Seventeen (27.4%) patients had normal, 23 (37.1%) patients had borderline and 22 (35.5%) patients had low serum Vit B12 level. Forty-four (71.0%) patients were considered as Vit B12 deficient; 22 had serum Vit B12 level <200 pg/ml (4 of these patients were on vitamin B12 replacement therapy), 22 were already on Vit B12 replacement therapy and Vit B12 level ≥200 pg/ml. The mean homocysteine levels were higher in the group with Vit B12 <200 pg/ml as compared to other groups (p=0.005). In the group with Vit B12 level <200 pg/ml, 33.3% (7/21) of the patients had hyperhomocysteinemia and 76.2% (16/21) had 'elevated homocysteine levels (Table). Fifty-one (82.3%) patients had GIS involvement and 16 (25.8%) patients had medium-high risk MUST score. H. Pylori IgG antibody was positive in 40 (64.5%) patients. There were no statistically significant differences between the patients with and without Vit B12 deficiency regarding to age, mean disease duration, hemoglobin level, GI involvement, medium-high risk MUST score, H. Pylori IgG antibody positivity and other clinical features (p>0.05 for all).

	Vit B12 level			
	>300 pg/ml n=16	200–300 pg/ml n=22	<200 pg/ml n=21	p
Homocysteine μmol/L, mean (SD)	9.1 (3.4)	10.4 (3.0)	14.1 (6.5)	0.005
Homocysteine >9 μmol/L, n (%)	7 (43.8)	16 (72.7)	16 (76.2)	0.084
Homocysteine >15 μmol/L, n (%)	1 (6.3)	0	7 (33.3)	0,004

Homocysteine level was not measured in 3 patients.

Conclusions: SSc patients are at risk for Vit B12 deficiency. Using homocysteine level seems to be unpractical for confirmation of Vit B12 deficiency in a complex disease such as SSc because its level is influenced by many factors. Patients with SSc should be closely monitored for Vit B12 deficiency and replacement therapy should be planned if necessary.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.6385

FRI0398 INFLAMMATORY MYOPATHIES WITH CAMPTOCORMIA OR DROPPED HEAD SYNDROME ARE ASSOCIATED SCLEROMYOSITIS WITH LATE ONSET AND DELAYED DIAGNOSIS

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Background: Severe weakness of axial muscle leads to dropped head syndrome or camptocormia. The signification of these symptoms has not been studied in inflammatory myopathies (IM).
Objectives: To assess the signification of dropped head syndrome and/or camptocormia in patients with IM.
Methods: All practitioners of the French Myositis Network and the Club Rhumatisme et Inflammation (>1000 physicians) were invited to report their patients suffering from IM (myopathy with myositis specific autoantibody and/or typical muscle biopsy according to the ENMC criteria) with camptocormia and/or dropped head syndrome. These axial IM cases were included only if no other explanation of axial weakness was found. IM patients without axial involvement (non-axial IM group), were randomly selected from the participative centers, and included as control patients (ratio 1:2). Clinical, serological, muscle pathological features, management and outcomes were studied using a standardized form.
Results: Twenty patients (sex ratio 2.6) with axial involvement (camptocormia: 60%, dropped head syndrome: 40%) were included. Compared with the control group, these axial IM-patients were older (64.05±11.64 y. vs. 48.22±18.28. p<0.005) and diagnosis of IM was delayed (16.4±4.5 months vs. 8.5±2.7, p<0.05).
All patient except one had also proximal weakness of the limbs. CK blood level was 2794±870 UI/L, which was similar to the controls (2864±559 UI/L). According to the ENMC classification, non-specific myositis was the most frequent finding on muscle biopsy (n=5/15, 30% vs. n=1/24, 4% in the non-axial group, p<0.05) and dermatomyositis (DM) pattern tended to be less frequent in axial-IM patients (3/12 20% vs. 11/24, 46%, p=0.10).
Most of the patients (75%) had also extra muscular involvement including acrosyndrome (45%), interstitial lung disease (35%), sclerodactyly (30%), telangiectasia (25%), digital tip ulcer (10%), sclerodermy (5%). By contrast, no patient had polyarthrithis (vs. 20% in the controls, p<0.05). DM rash was hardly threefold less frequent in axial IM patients (15% vs. 42%, p<0.05).
Auto-antibodies associated with scleromyositis were the most frequent in axial-IM patients (30% vs. 10.3%, p=007, manly anti-PM/Scl). One patient (5%) had cancer within the 3 years before or after IM diagnosis (NS vs. controls).
Thus, most frequent diagnosis in axial IM-patients were scleromyositis (35% vs. 5% in the controls, p<0.05) and inclusion body myositis (20% vs. 2.6% in the controls, p<0.05). DM was twofold less frequent than in control (10% vs 41%, p<0.05). Other IM subtypes were not statistically different from the control groups.
Except patients with diagnosis of sIBM, all axial IM-patients received corticosteroids with another immunomodulatory drugs (median number 2, range 1–5). Half of the axial-IM patients received intravenous immunoglobulin. After a mean follow up of 68.4±3.76 months all patients had improvement, including in axial weakness, except in patients with sIBM. One patient died from ischemic cardiomyopathy.
Conclusions: In IM, camptocormia and dropped head syndrome are associated with late onset scleromyositis and sIBM with delayed diagnosis.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.6564

FRI0399 EFFICACY OF AN INTENSIVE 24-WEEK PHYSIOTHERAPY PROGRAMME IN PATIENTS WITH SYSTEMIC SCLEROSIS - PRELIMINARY DATA FROM A SINGLE-CENTER CONTROLLED STUDY

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Background: Involvement of skin and musculoskeletal system in systemic sclerosis (SSc) leads to loss of function, disability and reduced quality of life. Data on efficacy of non-pharmacologic care in SSc is very limited due to variety in studied interventions/outcomes.
Objectives: To address the limitations of existing studies, and evaluate the effect of a controlled, long-term (24-week intervention, 24-week follow-up), intensive (1h physiotherapy + 0.5h occupational therapy twice weekly, and home-exercise for 0.5h 5x weekly), tailored physiotherapy programme on function/impairment of hands/face, and quality of life/disability in cohorts with a substantial number of SSc patients.
Methods: All patients fulfilled ACR/EULAR 2013 criteria, had skin involvement of hands/mouth, and were consecutively recruited from 2014 to 2016 at the Institute of Rheumatology in Prague. Both groups received educational materials and instructions for home exercise at baseline, however, only intervention group underwent the intensive physiotherapy programme. At months 0,3,6,12 all patients were assessed by a physician (physical examination, mRSS-Modified Rodnan's skin score, EUSTAR SSc activity score, Medsger SSc severity score), and a physiotherapist blinded to intervention [validated measurements (dFTP-delta finger to palm, inter-incisor/inter-lip distance, grip strength using Baseline dynamometer); tests (HAMIS-Hand Mobility In Scleroderma)], patients filled out patient reported outcomes/questionnaires (CHFS-Cochin Hand Function Scale, MHISS-Mouth Handicap In SSc Scale, HAQ, SHAQ, SF-36) and provided blood for routine laboratory analysis and biobanking. Normality of data was tested, inter-group analysis was performed with 2-way ANOVA, and intra-group analysis by Friedmann's test with Dunn's post hoc test.
Results: 25 SSc patients (22 female/3 male, 14 limited cutaneous (lc)SSc/11 diffuse cutaneous (dc)SSc, median of age 54.0 and disease duration 7.0 years, mRSS 12) were recruited into the intervention group (IG) and 29 patients into the control group (CG) (25 female/4 male, 16 lcSSc/13 dcSSc, median of age 49.0 and disease duration 5.0 years, mRSS 11). Compared to observed statistically significant deterioration in CG over the period of m0-m6, we found statistically significant improvement in dFTP, grip strength, HAMIS, inter-incisor and inter-lip

Parameter (unit)	Intervention group Mean ± SEM	Control group Mean ± SEM	Intra-group analysis (Friedmann+Dunn)		Inter-group analysis (2WA)
			Interevention gr.	Control group	
dFTP (cm)	m0: 5.7 ± 0.5	m0: 6.6 ± 0.5	m0-m3: p<0.001	m0-m3: p<0.01	p<0.0001
	m3: 6.2 ± 0.5	m3: 6.1 ± 0.4	m3-m6: p<0.05	m3-m6: p<0.05	
	m6: 6.8 ± 0.6	m6: 5.8 ± 0.4	m0-m6: p<0.001	m0-m6: p<0.001	
Hand grip strength (kg)	m0: 17.2 ± 1.8	m0: 16.6 ± 1.3	m0-m3: p<0.05	m0-m3: p=NS	p<0.0001
	m3: 19.2 ± 1.9	m3: 14.9 ± 1.4	m3-m6: p=NS	m3-m6: p=NS	
	m6: 19.7 ± 1.9	m6: 13.9 ± 1.9	m0-m6: p<0.001	m0-m6: p<0.01	
HAMIS	m0: 9.8 ± 1.3	m0: 3.9 ± 1.1	m0-m3: p<0.01	m0-m3: p<0.01	p<0.0001
	m3: 7.1 ± 1.2	m3: 6.4 ± 1.2	m3-m6: p<0.01	m3-m6: p<0.001	
	m6: 4.1 ± 0.9	m6: 9.3 ± 1.1	m0-m6: p<0.001	m0-m6: p<0.001	
Inter-incisor distance (mm)	m0: 30.6 ± 1.6	m0: 32.9 ± 1.3	m0-m3: p<0.01	m0-m3: p<0.001	p<0.0001
	m3: 33.3 ± 1.6	m3: 30.2 ± 1.4	m3-m6: p=NS	m3-m6: p=NS	
	m6: 36.2 ± 2.0	m6: 29.8 ± 1.4	m0-m6: p<0.001	m0-m6: p<0.001	
Inter-lip distance (mm)	m0: 39.2 ± 1.6	m0: 41.7 ± 1.1	m0-m3: p<0.01	m0-m3: p<0.05	p<0.0001
	m3: 42.4 ± 1.7	m3: 39.9 ± 1.2	m3-m6: p=NS	m3-m6: p=NS	
	m6: 44.6 ± 1.8	m6: 40.0 ± 1.3	m0-m6: p<0.001	m0-m6: p=NS	

Acronyms: SEM, standard error of the mean; Friedmann, Friedmann's test; Dunn, Dunn's post hoc test; 2WA, two way ANOVA; dFTP, delta finger to palm; HAMIS, Hand Mobility in Scleroderma; m0, month 0 (= at the baseline); m3, month (= in the middle of intervention period); m6, month 6 (= at the end of intervention); p, p-value; NS, not significant