immunosuppressive treatments may modify gut microbiota in SSc. Overall these findings support the involvement of altered immune recognition of specific gut bacteria in early SSc.

Disclosure of Interests: Anna-Maria Hoffmann-Vold Grant/research support from: Boehringer Ingelheim, Consultant of: Boehringer Ingelheim, Actelion, Bayer, GlaxoSmithKline, Speakers bureau: Boehringer Ingelheim, Actelion, Roche, Kristofer Andréasson: None declared, Simen Hyll Hansen: None declared, Simon Midtvedt: None declared, Håvard Frøtheim: None declared, Henriette Didriksen Consultant of: Actelion, Torhild Garen: None declared, Espen Bækkevold: None declared, Øyvind Midtvedt: None declared, Roger Hesselstrand: None declared, Brian K Chung: None declared, Øyvind Molberg: None declared

DOI: 10.1136/annrheumdis-2020-eular.5481

THU0349

THE RELATIONSHIP BETWEEN DISEASE ACTIVITY AND SEVERITY IN SYSTEMIC SCLEROSIS: A PROSPECTIVE ANALYSIS OF 278 PATIENTS

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Background: Evaluating disease activity and severity in systemic sclerosis (SSc) is crucial to define the patients who are candidate for treatment options.

Objectives: We aimed to investigate the relationship between disease activity and severity in SSc in a large cohort.

Methods: This is a cross-sectional prospective analysis of 278 (253 females) patients fulfilling ACR/EULAR (2013) classification criteria for SSc. Disease activity and severity were calculated separately for cutaneous subsets (EscSG and Medsger). The patients were grouped as inactive if EscSG score=0, mildly active if EscSG score>0<3, active if EscSG score≥3.

Results: The mean age, duration of Raynaud's and non-Raynaud features were 48.5±13.1, 12.1±9.8 and 8.3±7.5 years respectively. Characteristics of the SSc patients were summarized in table-1.

Table-1: Characteristics of SSc patients

	All patients n=278(%)
ANA	223(80.2)
anti-centromer	128(46.0)
anti-Scl70	39(14.0)
Anti-RNAPol3	29/165(17.6)
limited cutaneous pattern	143 (51.4)
diffuse cutaneous pattern	135(48.6)
digital ulcers	101(36.3)
Synovitis	56(20.1)
flexion contractures	81(29.1)
Myositis	40(14.4)
lung involvement	140(50.4)
PAH	9(3.2)
GIS involvement	166(59.2)
renal crisis	8(2.9)
immunosuppressives	173(62.2)
Steroids	156(56.1)
specific vasodilatory drugs	47(16.9)

Ninety-three (34%) and 151(54%) patients were evaluated as having active and mildly active disease. Only 34(12%) patients had inactive disease. The patients with diffuse cutaneous involvement (dcSSc) who were active had higher modified Rodnan Skin score(mRSS) and severity scores of general, skin and joint-tendon involvements; the patients who had mildly active disease also had higher scores of mRSS and severity scores of skin compared to those with inactive disease (table-2). The patients with limited cutaneous involvement (lcSSc) who were assessed as having active disease had higher mRSS and higher severity scores of general, skin, peripheral vascular, lung, joint-tendon and gastrointestinal involvements; the patients who had mildly active disease

also had higher scores of mRSS and severity scores of skin, lung, joint-tendon and gastrointestinal involvements compared to those with inactive disease (table-2).

Table-2: Severity scores of dcSSc and lcSSc patient groups according to activity of disease.

	dcSSc				lcSSc			
	inactive n=7	mildly active n=72	active n=64	*p	inactive n=27	mildly active n=79	active n=29	*p
mRSS	7.0±3.6*	16.3±7.6**	20.4±8.4*	<0.001	4.0±4.1*	7.9±5.1**	9.9±7.5*	<0.001
Severity Score								
General	1.0±1.6	0.4±0.7*	0.8±0.8*	0.004	0.4±0.6*	0.4±0.6	0.7±0.7*	0.004
PVI	1.5±0.6	1.8±0.8	2.1±0.9	NS	1.3±0.6*	1.6±0.7**	2.1±0.9*	<0.001
Skin	0.8±0.4*	1.7±0.7**	1.8±0.7*	0.003	0.7±0.5*	1.1±0.4**	1.2±0.6*	<0.001
Joint/tendon	0.5±0.8*	1.5±1.0	1.9±1.3*	0.023	0.3±0.7*	1.0±1.0	1.2±1.1*	0.003
Myositis	0.3±0.8	0.2±0.4	0.2±0.5	NS	0.1±0.6	0.1±0.4	0.2±0.5	NS
GIS	0.3±0.5	0.7±0.6	0.7±0.6	NS	0.3±0.5*	0.6±0.5**	0.7±0.5*	0.003
Lung	1.0±0.63	1.3±1.1	1.6±1.4	NS	0.3±0.7*	1.2±1.0**	1.5±1.1*	<0.001
Heart	0.2±0.4	0.1±0.6	0.1±0.5	NS	0.0±0.0	0.1±0.5	0.0±0.0	NS
Renal	0.0±0.0	0.0±0.0	0.9±0.46	NS	0.0±0.0	0.1±0.6	0.0±0.0	NS

*when inactive and mildly active group compared; p=0.005 for mRSS, p=0.036 for skin severity in dcSSc; p=0.005 for mRSS, p=0.012 for skin, p=0.004 for joint-tendon, p=0.021 for GIS and p<0.001 for lung severity.

**when active and mildly active group compared p=0.004 for PVI severity.

Conclusion: One third of our cohort was found to have active disease despite treatment and only 12% had inactive disease. Skin involvement and severity of different organs were shown to be higher in patients with active disease in both cutaneous subsets, together with severity of lung, peripheral vascular and gastrointestinal involvements in active lcSSc. LcSSc and dcSSc patients who had mildly active disease also had severe disease similar to those with active patients. Disease activity and severity should be assessed as separate measurements to highlight the course of the disease and may guide to the management of patients with SSc.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6129

THU0350

LOGOPEDIC TESTING IN SSC PATIENTS REVEALS HIGH FREQUENCY OF OROPHARYNGEAL DYSFUNCTION: A MONOCENTRIC EXPERIENCE

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Background: Up to 98% of patients with systemic sclerosis (SSc) show involvement of the gastrointestinal system (GI) [1]. While meteorism, heartburn and GI dysmotility are very common and accessible to pharmacologic treatment on an evidence based level [1–3], checking for oropharyngeal dysfunction is usually not part of the standard diagnostic algorithm. However, in a survey of the German Network for Systemic Sclerosis (DNSS) patients reported coughing and/or a sore voice in up to 78% [1]. As impairment in speaking or swallowing for example does not only substantially reduce quality of life, it can also be very stigmatizing. In addition, the usual prokinetic therapy of GI-involvement, e.g. metoclopramide, does not appear to improve these symptoms. As the first step to approach this problem is the qualitative and quantitative description, we evaluated the oropharyngeal function in our cohort of SSc patients by detailed logopedic assessment.

Objectives: To evaluate the frequency and type of oropharyngeal dysfunction, e.g. swallowing or speaking, in patients with SSc and to elucidate the correlating and associated factors, e.g. disease duration or modified Rodnan Skin Score.

Methods: After obtaining written consent, oropharyngeal function using a standardized assessment protocol was evaluated in patients with SSc fulfilling the ACR/EULAR criteria by a speech therapist. Furthermore, we investigated whether oropharyngeal dysfunction is associated with patients' characteristics. In addition, all patients received instruction for a training program to treat their individual oropharyngeal dysfunction.

Results: 37 patients with d/SSc were assessed for eligibility. 34 patients met the inclusion criteria (3 patients did not speak German) and written consent was obtained.

Oropharyngeal dysfunction (impairment of speaking, swallowing, breathing or oropharyngeal muscle function) was found in 29 of 34 (85%) of both l/dSSc patients. Neither the subtype of SSc, disease duration nor mRSS were significantly correlated with oropharyngeal dysfunction in general. Only GI involvement in general was associated with oropharyngeal dysfunction.

After logopedic therapy, 28 of the 34 (82%) patients with oropharyngeal dysfunction reported a benefit after 3 days of training and were motivated to continue logopedic training at home.

Oropharyngeal dysfunction	Present (n=29)	absent (n=5)	p-values (χ ² -test)
dcSSc	8	0	0,027
lcSSc	22	4	0,027
disease duration (mean)	12,7 y	12 y	0,462
mRSS<4	9	2	0,322
mRSS>4	20	3	0,322
Raynauds syndrome	29	5	0,673
digital ulcers	19	2	0,812
lung fibrosis	11	2	0,596
esophageal dilatation	15	2	0,566
esophageal dysmotility	19	0	0,015
GI-involvement	23	1	0,007

Conclusion: Logopedic assessment revealed a high incidence of oropharyngeal dysfunction in our cohort of SSc patients. Oropharyngeal dysfunction was not associated with disease duration, skin- or lung-involvement or dcSSc/lcSSc differentiation. A logopedic training program seems to be of benefit for this currently not pharmacologically treatable problem.

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Disclosure of Interests: Miriam Wirths: None declared, Ole Hudowenz: None declared, Ulrike Hoffmann: None declared, Ulf Müller-Ladner Speakers bureau: Biogen, Uwe Lange: None declared, Philipp Klemm Consultant of: Lilly, Medac
DOI: 10.1136/annrheumdis-2020-eular.3467

THU0351

MUSCLE INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS AND ANTI-PM/SCl+ ANTIBODIES IS ASSOCIATED WITH CARDIAC AND PULMONARY INVOLVEMENT. ANALYSIS OF THE MULTICENTRE EUSTAR COHORT.

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Background: Anti-PM/ScI antibodies positivity has been associated with frequent skeletal muscle involvement in patients with Systemic Sclerosis (SSc) in different studies, including the EUSTAR cohort (1). Moreover, although myositis has been previously associated with heart involvement in SSc patients (2), this issue has never been explored among anti-PM/ScI+ patients.

Objectives: To evaluate the cardiac involvement in anti-PM/ScI patients with SSc in the large multicentre EUSTAR database, with focus on the subgroup of patients with muscle involvement.

Methods: Patients from the EUSTAR database were included when the item anti-PM/ScI was fulfilled in at least one visit.

Results: Anti-PM/ScI status was available in 7,353 SSc patients from EUSTAR database: 295 were anti-PM/ScI+. After exclusion of 151 patients with multiple autoantibody positivity, 144 anti-PM/ScI+ patients were compared with 7,058 anti-PM/ScI- patients. Among them, 3,120 (44.2%) were positive for ACA, 2,361 (33.5%) for anti-Topo I and 274 (3.88%) for anti-RNAP3.

Regarding the specific cardiac outcomes, in the anti-PM/ScI+ as compared to the anti-PM/ScI- group, a decreased rate of elevated sPAP at ECHO was recorded (12.8% vs 25.0%, p:0.001), while no differences were observed in the frequency of conduction blocks (26.2% vs 23.7%, p:0.526), abnormal diastolic function (33.9% vs 36.4%, p:0.582), pericardial effusion (10.2% vs 10.9%, p:1.000) and LVEF ≤50% (4.76% vs 6.11%, p:0.818). In multivariate analysis, adjusted for age at disease onset, sex, and disease duration, the negative association of anti-PM/ScI with elevated sPAP was not confirmed (p:0.061).

When comparing anti-PM/ScI+ patients with (n=47) and without (n=87) CK elevation, the former group had a higher frequency of conduction blocks (43.2% vs 17.5%, p:0.005; OR 95% CI 3.47, 1.51-7.97) and left ventricular dysfunction, both diastolic (45.6% vs 27.2%, p:0.050; OR 95% CI 2.25, 1.05-4.81) and systolic (LVEF ≤50% 13.3% vs 0%, p:0.018; OR 95% CI 16.8, 0.87-324). Moreover, anti-PM/ScI+ patients with CK elevation had significantly increased rate of lung

fibrosis on HRCT (p:0.045), intestinal symptoms (p:0.017), joint contractures (p:0.045) and tendon friction rubs (p:0.034).

Conclusion: In the largest series of anti-PM/ScI positive SSc patients so far reported, muscle involvement in anti-PM/ScI+ patients (defined as increased serum CK) seems to represent a marker of a more severe disease phenotype, including a higher frequency of cardio-pulmonary involvement.

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Acknowledgments: Authors would like to thank the patients' association GILS (Gruppo Italiano Lotta Scleroderma) for the grant that supported the project.

Disclosure of Interests: Maria Grazia Lazzaroni: None declared, Stefania Zingarelli: None declared, Paolo Airò: None declared, Yannick Allanore Grant/research support from: BMS, Inventiva, Roche, Sanofi, Consultant of: Actelion, Bayer AG, BMS, BI, Oliver Distler Grant/research support from: Grants/Research support from Actelion, Bayer, Boehringer Ingelheim, Competitive Drug Development International Ltd. and Mitsubishi Tanabe; he also holds the issued Patent on mir-29 for the treatment of systemic sclerosis (US8247389, EP2331143), Consultant of: Consultancy fees from Actelion, Acceleron Pharma, AnaMar, Bayer, Baecon Discovery, Blade Therapeutics, Boehringer, CSL Behring, Catenion, ChemomAb, Curzion Pharmaceuticals, Ergonex, Galapagos NV, GSK, Glenmark Pharmaceuticals, Inventiva, Italfarmaco, iQvia, medac, Medscape, Mitsubishi Tanabe Pharma, MSD, Roche, Sanofi and UCB, Speakers bureau: Speaker fees from Actelion, Bayer, Boehringer Ingelheim, Medscape, Pfizer and Roche

DOI: 10.1136/annrheumdis-2020-eular.3525

THU0352

DIAGNOSTIC PERFORMANCES OF HAND ULTRASOUND PARAMETERS AND THEIR IMPACT ON THE 2013 ACR/EULAR CLASSIFICATION CRITERIA FOR SYSTEMIC SCLEROSIS.

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Background: Recent studies have highlighted that ultrasound (US) examination could offer a better assessment of hand manifestations of systemic sclerosis (SSc). Indeed, US allows a simultaneous evaluation of vascular, fibrotic and inflammatory hand features of the disease. Power Doppler US can especially explore macrovascular involvement characterized by an obliteration of digital arteries or ulnar arteries. Ulnar artery occlusion (UAO) is especially frequent in SSc patients and could be a relevant marker of the severity of SSc-associated vasculopathy. Among other hand manifestations of SSc, US evaluation can notably explore tenosynovial involvement such as fibrotic tenosynovitis (TS), which is considered to be SSc-specific.

Objectives: This study aims to assess the diagnostic performances of these hand US parameters for the diagnosis of SSc.

Methods: 244 patients with suspected SSc were consecutively included. They all had US evaluation assessing the presence of fibrotic TS and UAO. The final diagnosis of SSc was based on the evaluation of an expert, independently from US results and from any pre-established classification criteria.

Results: 166 patients were finally diagnosed as SSc. 62 SSc and 8 non-SSc patients had UAO (uni or bilateral) (p=0.001). 23 SSc patients and 1 non-SSc patient had US fibrotic TS (p=0.007). A US SSc-pattern (presence of UAO and/or fibrotic TS) was reported in 73 SSc patients and 9 non-SSc patients (p<0.001). UAO had an area under ROC curve (AUC) for the diagnosis of SSc of 0.618 (95%CI 0.539-0.697); with Se= 0.373 (0.304-0.449) and spe=0.862