0.607–0.816), being slightly lower. A proximal diameter ≥ 7.5 mm provides a specificity of 87.2% (95% CI: 76.7–96.7) and a sensitivity of 65.3% (95% CI: 51.9–78.6) for esophageal dysmotility, enabling correct classification of 75% of the patients. A distal diameter ≥ 12.9 mm provides a specificity of 76.6% (95% CI: 64.5–88.7) and a sensitivity of 71.2% (95% CI: 58.8–83.5), correctly classifying 73% of the patients.

Abstract THU0391 – Figure 1. ROC curves of the diameters for esophageal dysmotility detected by manometry.

Conclusions: CT, a less invasive technique than manometry, can be an acceptable diagnostic tool for esophageal dysmotility in SSc, when the maximum proximal or distal esophageal diameter are ≥ 7.5 mm and 12.9 mm, respectively. CT done in the daily clinical practice could be exploited when manometry is not preferred in selected patients with SSc. More studies need to be carried out to confirm this results.

Disclosure of Interest: None declared

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THU0392 **SYSTEMIC SCLEROSIS AND PRIMARY BILIARY CHOLANGITIS: AN OVERLAP SYNDROME? PRELIMINARY DATA FROM A MULTICENTRE EUSTAR STUDY**

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Background: The association between systemic sclerosis (SSc) and primary biliary cholangitis (PBC) is well known. However, classifications criteria of the 2 diseases have been recently revised and may change the relationships. Furthermore, the specific outcomes with regards to organ involvement and also of liver aspects have been only scarcely investigated.

Objectives: To describe clinical characteristics of SSc-PBC patients compared to SSc using a large series

Methods: A multicentre EUSTAR study collection data of SSc patients with known PBC or with PBC-specific antibodies (abs) and of SSc controls matched for disease duration and cutaneous subset. Data were recorded at baseline and at the last available visit.

Results: 229 patients were enrolled (85 SSc-PBC and 144 SSc). The mean age of the population at the SSc diagnosis was of 53 ± 12.8 . **Baseline characteristics.** The limited cutaneous subset was the most common. Anticentromere abs (ACA) were present in 82.9% of SSc-PBC patients (vs 68.5% in SSc, p -value=0.01) while anti-topoisomerase I abs were less frequently (1.2% vs 9.7%), p -value=0.02. Out of 85 SSc-PBC patients, antimitochondrial abs (AMA) were present in 80%, only 9.4% presented anti-gp210 abs and 7.1% anti-sp100 abs. The two populations did not differ for fibrosis at HRCT, lung function tests, the value of creatinine and for pulmonary arterial hypertension. A trend towards statistical significance was found in the prevalence of digital ulcers (DUs) as patients that have never suffered from past or current DUs were greater in the SSc-PBC group (78.6% vs 66%, p -value=0.051). Regarding other autoimmune associated diseases, a greater prevalence of Hashimoto thyroiditis was found in SSc-PBC (p -value=0.03). At baseline, transaminase, alkaline phosphatase and γ GT levels were all higher in PBC-SSc (p -value<0.001) (see Fig 1). **Outcomes of organ**

involvements. The mean follow-up was of 10.5 ± 6.8 years. During the follow-up the percentage of patients presenting ILD (defined as new appearance of fibrosis at control HRCT scan) was greater in SSc vs SSc-PBC (12.9% vs 2.8%, p -value=0.02) and PAH percentage of new cases was of 9.9% in SSc while it was not present in SSc-PBC (p -value=0.003).

	SSc-PBC population	SSc controls	p-value
Cutaneous subset			
Limited cutaneous subset	77 (90.6%)	132 (91.7%)	0.75
Diffuse cutaneous subset	2 (2.4%)	5 (3.5%)	
Unclassified cutaneous subset	6 (7.1%)	7 (4.6%)	
CT fibrosis	15 (17.6%)	28 (19.4%)	0.69
PVC (forced vital capacity) [mean $\pm$ SD]	104.3 $\pm$ 21.5	103.2 $\pm$ 17.1	0.72
DICO (diffusing capacity for carbon monoxide) [mean $\pm$ SD]	78.3 $\pm$ 18.3	76.5 $\pm$ 17.5	0.50
Portal hypertension	4 (4.7%)	-	
Arrhythmia	4 (4.7%)	4 (2.8%)	0.55
Conduction block	7 (8.2%)	7 (4.9%)	0.39
PAH	5 (5.9%)	2 (1.4%)	0.10
Never DUs	66 (77.6%)	95 (66%)	0.051
GI symptoms			
GERD	44 (51.8%)	90 (62.5%)	0.24
Malabsorption	2 (2.4%)	5 (3.5%)	>0.99
Anorectal incontinence	1 (1.2%)	4 (2.8%)	0.66
Thyroid diseases			
Basedow	1 (1.2%)	1 (0.7%)	>0.99
Hashimoto	10 (11.8%)	8 (5.6%)	0.034
SSc specific abs			
Anti-topoisomerase I	1 (1.2%)	14 (9.7%)	0.023
ACA	68 (80%)	98 (68.1%)	0.012
PBC specific abs			
AMA	68 (80%)	-	
gp210	8 (9.4%)	-	
sp100	6 (7.1%)	-	
Therapy			
Immunosuppressant therapy	18 (21.2%)	43 (29.9%)	NS
Steroid therapy	19 (22.4%)	27 (18.8%)	NS
Deoxicholic acid	58 (68.2%)	-	
Creatinine [median(IQ)]	70 (61-81.3)	70 (61-79.5)	0.99
Alanine transaminase [median(IQ)]	34.5 (26-59.25)	20 (15-26)	<0.001
Aspartate transaminase [median(IQ)]	36 (28-54.25)	21 (19-27)	<0.001
Alkaline phosphatase [AP] [median(IQ)]	147 (94-296)	71 (56-90)	<0.001
γ GT [median(IQ)]	143 (59.5-253.5)	18 (11-31.25)	<0.001

Abstract THU0392 – Figure 1. Clinical features at baseline

Conclusions: PBC is more present in ACA positive IcSSc; SSc-PBC patients have a higher risk of polyautoimmunity. Of the most interest, SSc-PBC patients seem to have a milder SSc phenotype with less severe organ involvement and progression. Regarding PBC phenotype, only 4 patients presented portal hypertension and nobody was subjected to liver transplantation. At baseline, the SSc-PBC group had higher cholestatic liver enzymes and more than 60% were treated with deoxycholic acid. In the future, it will be useful to evaluate the PBC phenotype during the follow-up enrolling a greater number of patients.

Disclosure of Interest: None declared

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THU0393 **THE PREDICTOR OF MALNUTRITION IN SYSTEMIC SCLEROSIS (PREMASS) SCORE: A VALIDATED COMBINED INDEX PREDICTIVE OF FUTURE WEIGHT LOSS IN SYSTEMIC SCLEROSIS**

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Background: Malnutrition and severe gastrointestinal dysfunction are the cause of mortality in 4%–15% of systemic sclerosis (SSc) patients whereas overall gastrointestinal involvement is observed in 75%–90% of cases.¹ Hence, a reliable tool for stratification of risk for malnutrition would be of great value in the clinical management of SSc.

Objectives: Here we set out to identify a combined index predictive of significant weight loss at 12 months employing Malnutrition Universal Screening Tool (MUST) and serum adiponectin to leptin ratio (A/L) already used in other conditions.

Methods: This was an international, multicentre, longitudinal study employing 180 SSc patients in two independent cohorts: a study cohort (110 consecutive SSc patients) enrolled from University of Messina (60) and University of Padova,²⁰ and a validation cohort (70) at the University of Leeds. Serum A/L ratio was measured by ELISA. MUST score, which includes BMI and weight loss reported by the patient in the last 3–6 months, was calculated as described: 0=no, 1=mild, >2= moderate/severe risk of malnutrition. End point of the study was weight loss >10% of baseline weight at 12 months.

Results: The two cohorts showed no significant differences in demographic and clinical features. Overall, median BMI decreased over time in both study and validation cohorts (23.5 vs 22.35 and 23.44 vs 22.49, respectively; p <0.0001). A/L

ratio correlated significantly with BMI in both cohorts ($r^2=0.19$ for study cohort, $r^2=0.25$ for validation cohort; $p<0.0001$). MUST score had only moderate value in predicting weight loss in the study cohort (AUC 0.7; 95% CI: 0.58–0.82). Specifically, 46.5% of SSc patients lost $>10\%$ wt despite having “no” or “mild” MUST scores. Logistic regression analysis identified the combination of BMI and A/L as the best PREDictor of MAInutrition in Systemic Sclerosis (PREMASS). The formula $12.18 - (0.63 \times \text{BMI}) + (1.51 \times \text{A/L})$ predicted the end point with AUC=0.91 (95% CI: 0.77–0.84). A PREMASS score >0.23 showed 91.3% sensitivity (95% CI: 79.79–100) and 80.46% specificity (95% CI: 72.13–88.79) for $>10\%$ wt loss with an overall 55.26% positive predictive value (PPV) (95% CI: 39.45–71.07) and 97.22% negative predictive value (NPV) (95% CI: 93.43–100) and a relative risk (RR) of 19.90 (95% CI: 4.93–80.37). In the validation cohort, PREMASS showed 76.47% sensitivity (95% CI: 56.31–96.63) and 75.47% specificity (95% CI: 63.89–87.06) with an overall 50% PPV (95% CI: 30.78–69.22) and 90.91% NPV (95% CI: 82.41–99.4) and a RR of 5.5 (95% CI: 2–15.10).

Conclusions: PREMASS is the first validated index for weight loss risk stratification in the following 12 months in SSc. Prediction of future weight loss in SSc could aid both in clinical management and stratification/enrichment in clinical trials.

Disclosure of Interest: None declared

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THU0394 ENTHESTITIS IN SYSTEMIC SCLEROSIS (SSC): AN ULTRASOUND (US) PILOT STUDY

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Background: Articular involvement is frequently encountered in SSc and previous US studies suggest that synovitis is the commonest manifestation. Recently, it has been reported that SSc patients may show typical “hall-marks” of spondyloarthritis (SpA). Apart from tendon involvement which is a common event, sacroiliitis has been estimated to have a prevalence of 23%.

Objectives: To estimate the prevalence of enthesal and Synovio-Enthesal Complex (SEC) modifications in SSc

Methods: 30 SSc patients (2013 ACR/EULAR classification criteria) without a history of articular involvement (4 male, 26 female, mean age 53.3 ± 16.6 years) were included in this pilot cross sectional US study. 12 healthy subjects (2 male, 10 female, mean age 46.9 ± 5.8 years) were used as controls. The enthesal sites were the lateral epicondylar common extensor tendons (CET), and sites of the Glasgow Ultrasound Enthesis Scoring System. The GUESS score was also calculated as a soft tissue score (GUESS soft tissue) and as a bone score (GUESS bone). US assessment was performed with a MyLab 70XVG scanner equipped with a 6–18 MHz linear transducer (Esaote). Only the epicondylar region was evaluated with PowerDoppler US (PDUS), using semi-quantitative graded from 0 to 3. Involvement of SEC was evaluated at the epicondylar region in SSc patients who presented a PDUS signals ≥ 1 in the CET closer than 2 mm from the bony surface. SEC involvement was defined as the presence of a PDUS signal ≥ 2 in the elbow epicondylar synovial fold proximal to the annular ligament (AL), inferior to the bone insertion of CET and to the radial collateral ligament (RCL). Statistical analyses were carried out using Mann-Whitney U, Spearman correlation and Chi-square tests. Results were considered significant if $p<0.05$

Results: In SSc, the GUESS and GUESS soft tissue scores were significantly higher (5.67 ± 0.87 and 3.43 ± 0.44 , respectively) than in controls (1.25 ± 0.41 and 0.92 ± 0.29 , respectively) ($p<0.0001$) as was the GUESS bone score (SSc 2.23 ± 0.55 vs controls 0.33 ± 0.22 ; $p<0.05$). The CET entheses of SSc patients showed significantly more US B-mode alterations than controls (hypoechoogenicity $\chi^2=5.95$, $p=0.015$, cortical irregularity $\chi^2=7.90$, $p=0.005$, calcification/enthesophytes $\chi^2=3.78$, $p=0.05$). A PD signal in the CET entheses was found in 18/60 sites in SSc and in 1/24 in controls. The PD signal of the CET entheses was significantly higher in SSc patients than in healthy controls (0.47 ± 0.10 and 0.08 ± 0.08 respectively, $p=0.018$) as was the presence of SEC inflammation ($\chi^2=4.54$, $p=0.033$). In SSc, there was a strong correlation between the presence of PD signal at CET entheses and concomitant SEC inflammation ($\rho=0.655$, $p<0.0001$) but there were no correlations between GUESS score or CET enthesitis and disease subset, disease duration, antibodies, DLCO, FVC, DLCO/VA.

Conclusions: Our data show that SSc patients frequently present the usual US features of enthesitis. Moreover, CET enthesitis were correlated with SEC inflammation suggesting that enthesal inflammation in SSc may share same microanatomical targets as SpA.

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THU0395

INFLUENCE OF SETTING AN UPPER LIMIT OF THE MRSS AS AN INCLUSION CRITERION IN SSC CLINICAL TRIALS ON THE RATIO OF SKIN FIBROSIS PROGRESSION VS. IMPROVEMENT – AN ANALYSIS OF THE GENISOS COHORT

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Background: Skin involvement is a main domain in the assessment of patients with systemic sclerosis (SSc), and the modified Rodnan skin score (mRSS) is a primary outcome measurement in SSc clinical trials. Recent studies on large SSc cohorts have shown that lowering the upper threshold of mRSS as a study inclusion criterion leads to cohort enrichment with patients with progressive skin disease. Limitations of these studies were lack of racial diversity and low proportion of patients with anti-RNA-Polymerase III (Pol3) antibodies.

Objectives: As the Texas-based GENISOS is an ethnically diverse cohort and includes a large proportion of Pol3-positive patients, this study aimed to assess the effect of different mRSS cut-off values at baseline on progression of skin fibrosis after one year of follow-up.

Methods: We extracted data from GENISOS for patients fulfilling the 1980 ACR criteria for SSc and the Le Roy criteria for diffuse cutaneous SSc, who had a mRSS ≥ 7 at inclusion and a follow-up visit with documented mRSS at 12±2 months. Progressors were defined as having an increase in mRSS >5 points and $\geq 25\%$ from the baseline to 2nd visit, while regressors were defined as having a decrease in mRSS of >5 points and $\geq 25\%$. To identify the optimal cut-off of baseline mRSS that yields the highest sensitivity for progressive skin fibrosis, we developed ROC curves and logistic regression models with “progression” as outcome variable and a binary variable of baseline mRSS cut-off point as predictor.

Results: We identified 152 patients (age and disease duration [median, Q1–Q3, years] 49.5, 40.2–57.3 and 2.2, 1.1–3.3 respectively, 22.4% males) who matched the inclusion criteria. The proportion of patients of African American ethnicity was 31/152 and 50/152 were Pol3-positive patients, both substantially higher than in European cohorts.

After one year, 17 patients (11.2%) classified as progressors and 51 (33.6%) as regressors. Progressors were more frequently positive for anti-topoisomerase antibodies (37.5% vs. 15.3%, $p=0.028$), negative for anti-Pol3 antibodies (93.8% vs. 62.3%, $p<0.012$), had a shorter disease duration (median, Q1–Q3: 1.3, 0.5–2.2 vs. 2.4, 1.1–3.5 years, $p<0.005$) and lower mRSS (median, Q1–Q3: 21, 11–25 vs. 24, 16–31, $p<0.012$) than non-progressors.

Sixteen of 17 progressors, but only 33 of 51 regressors had a baseline mRSS ≤ 27 . The mRSS cut-off of ≤ 27 had the highest probability of progression (odds ratio 9.1, 95% confidence interval 1.2–70.9, $p<0.035$, area under the curve 0.652). Using this cut-off as an inclusion criterion (vs. no cut-off) would have included 94% of all progressors, but only 65% of all regressors and 67% of all patients. The figure 1 displays absolute numbers of progressors and regressors at 1 year for each mRSS cut-off.

Conclusions: This analysis reconfirmed, in a population rich in patients of African American origin and with high prevalence of Pol3 antibodies, that setting a lower upper threshold of mRSS at study inclusion increases the proportion of progressors and reduces the absolute number of regressors. This confirms that this recruitment strategy should be used for clinical trial design in early diffuse SSc.

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