Conclusions: The results of this study suggest that impaired 6MWD in SSc patients with borderline PAP (and normal RV function at rest) might be caused by reduced RV contractile reserve (reduced RV output) and reduced PAC during exercise rather than by elevated pressures in the borderline range alone. These findings give further evidence for borderline PAP being an early stage of pulmonary vascular disease.

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THU0390 PATIENTS WITH SYSTEMIC SCLEROSIS DEVELOP FOCAL FIBROSIS OVER TIME, AND INCREASED ECV DIFFUSE FIBROSIS SEEN IN POOR PROGNOSTIC GROUP – A FIRST LONGITUDINAL CARDIAC MRI STUDY

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Background: Subclinical SSc-cardiomyopathy is described in up to 2/3 of the patients when sensitive methods such as cardiac MRI (CMR) are used. The prognostic implications and the natural history of these findings is unknown.

Objectives: To evaluate in SSc patients, free of cardiovascular (CV) risk factors and CV disease (CVD) the prevalence, clinical association and natural history of CMR abnormalities over 3 years.

Methods: 35 SSc patients, fulfilling the ACR/EULAR criteria, with no CVD, diabetes and ≤1 CV risk factor had 2 CMRs, 3 years apart. A 3T CMR with late gadolinium enhancement (LGE), T1 mapping for extracellular volume (ECV) quantification and stress perfusion (data available later) was undertaken. Initial CMR was compared with CMR results of 30 healthy controls (HC).

Results: 35 pts had an initial CMR: 26 (74%) female, mean (SD) age 55 (15) (43%) dcSSc, 11 (31%) ACA+, 12 (34%) Scl70+, 15 with (43%) interstitial lung disease (ILD) and 13 (37%) with history of digital ulcers (DU). 21 (60%) received any DMARD over the 3 year period and 10 (29%) prior treatment with cyclophosphamide. The first CMR (CMR1) of SSc pts vs HC showed higher ECV% values and comparable left ventricle (LV) volumes (table 1). LGE was present in 9/22 pts vs 1/30 HC. 22/35 pts had a second CMR (CMR2) at year 3 (Y3). A further 5 pts had evidence of LGE, total of 14 (40%): 7/14 had dcSSc, 6/14 males, 2 ACA positive, 6 Scl70 positive. LGE distributed in the basal and mid segments, mainly in a linear or patchy pattern. Of those with LGE at CMR1 (5/9 pts with LGE on CMR1 had CMR2) no change in the LGE pattern at CMR2 was observed. None of the initial CMR measures associated with LGE development at Y3 ($p > 0.05$). Whilst ECV had an overall decrease, ECV increased in patients with ILD [mean diff. (CI) 3 (-

1,6), $p = 0.14$] and in those with higher mRSS at baseline ($r = 0.455$, $p = 0.04$). A significant decrease over the 3 years was observed in LV end-diastolic volume (LVEDV/BSA), LV end-systolic volume (LVESV/BSA) and left ventricular stroke volume (LVSV/BSA) (table 1). A decrease in LVEDV/BSA was noticed for those with a history of DU [mean diff. (CI) -5 (-12,2), $p = 0.1$], ILD [mean diff. (CI) -6 (-12,0,5), $p = 0.07$] and shorter disease duration ($r = -0.504$, $p = 0.02$).

Abstract THU0390 – Table 1

CMR variable	HC Mean (SD)	SSc patients Mean (SD)	SSc patients Mean (SD)	Change (95% CI) in CMR between SSc patients	P value
LGE	3% (1/30)	21% (9/35)	22% (10/22)	23%	McNemar 0.063
ECV%	25(3)	30(4)	29(3)	-0.1 (-0.3, 0.1)	0.2
T1 native	1202 (35)	1199 (58)	1243 (67)	47 (-1, 94)	0.053
LVEDV/BSA (ml/m ²)	80 (11)	78(16)	71(15)	-7 (-10, -3)	<0.001
LVESV/BSA (ml/m ²)	31(6)	30(9)	26(9)	-4 (-6, 2)	0.001
LVSV/BSA (ml/m ²)	49(7)	48(9)	45(8)	-3 (-6, 0)	0.028
Lv mass/BSA (g/m ²)	49(8)	44(13)	50(21)	6 (-2, 13)	0.129
LVEF (%)	62(5)	62(5)	64(6)	2 (-0, 4)	0.08
Distensibility (10-3mmHg ⁻¹)	5 (2)	4 (2)	5 (3)	0 (-1, 2)	0.491
Torsion	15(4)	13(4)	13(5)	0 (-2, 3)	0.828

Conclusions: This longitudinal CMR study in SSc demonstrates that CMR is sensitive to change over time. More individuals developed LGE (of focal fibrosis) despite immunosuppressive treatment, and ECV% (diffuse fibrosis) appeared to worsen in a poor prognostic group. Functional changes were also observed. These data justify larger studies to inform stratification strategy for CMR in SSc, and also provide new insights for further investigation.

Disclosure of Interest: None declared

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THU0391 CLINICAL VALUE OF COMPUTED TOMOGRAPHY FOR THE DIAGNOSIS OF ESOPHAGEAL DYSMOTILITY IN SYSTEMIC SCLEROSIS

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Background: Esophageal dysmotility is common in Systemic Sclerosis (SSc), affecting 50%–80% of patients, usually associated with poor prognosis. SSc leads to atrophy and fibrosis of the smooth muscle of the oesophagus, modifying peristaltic contractions and motility. Manometry is considered the gold standard for the diagnosis of esophageal motility disorders, but dilatation can also be observed with computed tomography (CT), even if its diagnostic validity is still unknown.

Objectives: To compare esophageal dilatation observed with CT to manometry, in patients with SSc and to confirm whether CT can be used in the assessment of esophageal dysmotility.

Methods: Forty six patients meeting the 2013 ACR/EULAR Classification Criteria for SSc, and 53 healthy controls were included and retrospectively studied. Patients with overlapping syndromes, active infections or with longstanding diabetes were excluded. Epidemiological and clinical data were collected from medical records. All patients and controls had undergone at least one manometry and one CT, requested in daily clinical practice for another purpose. The most recent exams were selected for the study. Esophageal involvement was assessed using manometry (aperistalsis, inefficient peristalsis, nonspecific dysmotility and normal peristalsis) and compared with the largest coronal esophageal luminal diameters proximally, near the carina, and distally observed by CT. Data analysis was performed by STATA. All patients signed written informed consent, approved by the Research Ethics Committee.

Results: The sample included 76 women (10 dcSSc, 28 lcSSc, 3 MCTD, 35 controls) and 23 men (3 dcSSc, 2 lcSSc, 18 controls). Esophageal dysmotility was seen in 40/46 patients with SSc (87%) by manometry (defined as inefficient peristalsis or aperistalsis). Esophageal dilatation (≥ 10 mm) was present proximally in 23/44 patients (52.3%), distally in 35/46 patients (76.1%), and near the carina in 26/44 patients (59.1%). Esophageal dilatation at any level was statistically associated with esophageal dysmotility ($p < 0.05$). The areas under the ROC curves (figure 1) suggest that the esophageal proximal diameter in the coronal plane is good for detecting esophageal dysmotility (0.798, 95% CI 0.705–0.890), with the distal diameter (0.759, 95% CI 0.661–0.857) and the carinal diameter (0.712, 95% CI

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 (61.6-81.3)	70 (61.6-79.5)	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)	<0.002
γ GT [median(IQ)]	143 (59.5-253.5)	18 (11-31.25)	<0.002

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.

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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