Conclusions: This proof-of-principal study demonstrates that DNN processing of stained dermal biopsy sections are sensitive to clinically relevant features of SSc skin. These results suggest that DNNs dramatically expand the quantifiable SSc phenome and that histological samples can now be incorporated into models of SSc. Moreover, our results indicate that the gene expression underlying SSc may be driving histological differences in SSc skin.

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THU0388

DIFFERENCES IN CLINICAL COURSES AND SERUM MARKERS OF INTERSTITIAL LUNG DISEASE ASSOCIATED WITH ANTI-AMINOACYL-TRANSFER RNA SYNTHETASE ANTIBODY AND ANTI-MELANOMA DIFFERENTIATION-ASSOCIATED GENE 5 ANTIBODY-POSITIVE POLYMYOSITIS/DERMATOMYOSITIS

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Background: Polymyositis/dermatomyositis (PM/DM) is a chronic autoimmune disease that is often complicated by interstitial lung disease (ILD). Anti-aminoacyl-transfer RNA synthetase antibody (ARS-Ab) and anti-melanoma differentiation-associated gene 5 antibody (MDA5-Ab) are highly detected in PM/DM with ILD. It was reported that ARS-Ab-positive-ILD (ARS-ILD) is often recurrent¹, and patients with MDA5-Ab-positive-ILD (MDA5-ILD) develop fatal rapidly progressive ILD².

Objectives: To evaluate the differences in clinical courses between ARS-ILD and MDA5-ILD, including the changes in serum ILD markers.

Methods: We retrospectively investigated 25 patients with ARS-ILD and 26 patients with MDA5-ILD who received induction therapy between April 2002 and September 2017 at Kobe University Hospital. The survival rate and relapse-free survival rate were analysed with Kaplan-Meier estimation and the log-rank test. The differences in serum ILD markers between patients with ARS-ILD and MDA5-ILD were evaluated with the Student's t-test.

Results: Disease subtypes at diagnosis with PM/DM-associated ILD were as follows: Eleven ARS-ILD and no MDA5-ILD patients had PM, 10 ARS-ILD and 5 MDA5-ILD patients had DM, and 4 ARS and 21 MDA5 patients had amyopathic DM. The survival rate for MDA5-ILD was significantly lower than that for ARS-ILD ($p < 0.01$, figure 1A). On the other hand, there was no significant difference in the relapse-free survival rate between ARS-ILD and MDA5-ILD ($p = 0.25$, figure 1B). The serum level of Krebs von den Lungen-6 was not significantly different between ARS-ILD and MDA5-ILD (1044.3 ± 768.1 U/ml in ARS-ILD vs $1044.863.1 \pm 566.5$ U/ml in MDA5-ILD, $p = 0.33$), but the serum level of ferritin was significantly higher in MDA5-ILD than in ARS-ILD (286.4 ± 422.3 ng/ml in ARS-ILD vs 696.2 ± 839.5 ng/ml in MDA5-ILD, $p = 0.04$). Although the serum level of surfactant protein D (SP-D) in ARS-ILD was high, the SP-D level in MDA5-ILD was normal (158.9 ± 82.1 ng/ml in ARS-ILD vs 46.3 ± 22.1 ng/ml, $p < 0.01$).

Conclusions: MDA5-ILD patients should be monitored for both rapidly progressive disease and relapsing disease. A normal SP-D level is a feature of MDA5-ILD.

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THU0389

BORDERLINE PULMONARY HYPERTENSION WAS ASSOCIATED WITH REDUCED CARDIAC OUTPUT DURING EXERCISE IN PATIENTS WITH CONNECTIVE TISSUE DISEASES

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Background: In patients with systemic sclerosis (SSc) borderline mean pulmonary arterial pressures (mPAP; 21–24 mmHg at rest) are a frequent finding and could represent an intermediate stage between normal pulmonary pressures and manifest pulmonary hypertension (PH).¹

Objectives: The objective of this prospective study was to compare right ventricular function and pulmonary arterial compliance (PAC) at rest and during exercise between systemic sclerosis (SSc)-patients with normal and borderline mean pulmonary artery pressures, respectively.

Methods: SSc-patients (n=112) underwent clinical assessment, including right heart catheterization at rest and during exercise and were divided in three groups according to their resting mPAP values: normal mPAP (≤ 20 mmHg), borderline mPAP (21–24 mmHg) and manifest pulmonary hypertension (PH, mPAP ≥ 25 mmHg). Results were compared between groups by ANOVA followed by post-hoc student's t-test.

Results: SSc Patients with borderline PAP showed significantly lower cardiac index (CI) increase during exercise and higher PVR values than SSc patients with normal PAP at rest. Six-Minute-walking distance (6MWD) and PAC (stroke volume/(systolicPAP-diastolicPAP)) were significantly lower in the borderline PAP group compared to patients with normal PAP.

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.

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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