Results: The mean age was 60.4 years and 91.5% were women (table 1). Sarcopenia was identified in 15 (18.3%) SSc patients. Malnutrition was diagnosed in 12 (14.6%) and was more common in patients with sarcopenia ($P=0.038$). Also, there were significant differences between patients with and without sarcopenia regarding Body Mass Index ($P=0.001$), Fat Free Mass Index ($P<0.001$), knee extension strength ($P=0.049$), and Timed Up and Go test ($P=0.041$). There were no differences regarding age, disease duration, Rodnan skin score (mRSS), FVC and DLCO.

Table 1. Clinical characteristics of SSc patients with and without sarcopenia. Data presented as number (%) of patients, except when indicated otherwise.

Patients features	Whole (n=82)	SSc patients without sarcopenia (n=67)	SSc patients with sarcopenia (n=15)	P
Females	75 (91.5)	62 (92.5)	13 (86.6)	0.606
Caucasian	68 (82.9)	53 (79.1)	15 (100)	0.258
Age (years) ^b	60.4 (10.6)	60.2 (10.3)	61.5 (12.3)	0.678
Diffuse skin involvement	16 (19.5)	16 (23.8)	0 (0)	0.082
Disease duration (years) ^a	12.8 (7.5, 19.2)	12.7 (7.1, 19.2)	13.4 (8.9, 19.8)	0.324
Rodnan skin score ^a	4.0 (2.0, 10.0)	5.0 (2.0, 10.0)	2.0 (2.0, 10.0)	0.076
Interstitial lung disease on HRCT	32 (39.5)	25 (37.3)	7 (10.4)	0.737
FVC (% predicted) ^b	88.4 (16.6)	88.4 (16.9)	88.3 (15.3)	0.991
DLCO (% predicted) ^b	63.7 (11.9)	63.6 (12.2)	64.2 (11.2)	0.855
BMI (kg/m ²) ^b	25.6 (4.6)	26.5 (4.4)	21.8 (4.1)	0.001
FFMI (kg/m ²) ^b	15.6 (1.7)	16.1 (1.6)	13.8 (1.2)	<0.001
FMI (kg/m ²) ^b	9.5 (3.7)	9.9 (3.6)	7.5 (3.5)	0.023
Malnutrition	12 (14.6)	7 (10.4)	5 (33.3)	0.038
MUST - Low risk	58 (70.7)	51 (76.1)	7 (46.7)	0.046
Moderate and high risk	24 (29.3)	16 (23.9)	8 (53.3)	
HAQ ^b	0.778 (0.591)	0.797 (0.575)	0.833 (0.576)	0.825
Timed Up and Go test (s) ^a	8.41 (7.41, 10.59)	8.39 (7.5, 9.4)	10.23 (7, 14)	0.041
Knee extension strength (kgf) ^a	21.40 (15.65, 27.52)	21.70 (18.05, 28.35)	14.82 (9.5, 24)	0.049

^a Median (25, 75th percentiles)

^b Mean (standard deviation)

Abbreviations: BMI: body mass index; DLCO: diffusion capacity of carbon monoxide; FFMI: fat free mass index; FMI: fat mass index; FVC: forced vital capacity; HAQ: Health Assessment Questionnaire; HRCT: high-resolution computed tomography; MUST: Malnutrition Universal Screening Tool; SSc: systemic sclerosis.

Conclusion: Sarcopenia is relatively common and is associated with malnutrition in patients with SSc. In our population, sarcopenia was not associated with other features related to a more severe disease. The role of sarcopenia in the prognosis of SSc needs to be better understood in longitudinal studies.

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Disclosure of Interests: Vanessa Hax: None declared, Rafaela Cavaleiro do Espírito Santo: None declared, Emerson Pena: None declared, Luisa Rodrigues: None declared, Renata Ternus Pedo: None declared, Jordana Miranda de Souza Silva: None declared, Nicole Pamplona Bueno de Andrade: None declared, Andrese Aline Gasparin: None declared, Ricardo Xavier Consultant of: AbbVie, Pfizer, Novartis, Janssen, Eli Lilly, Roche, Rafael Mendonça da Silva Chakr: None declared

DOI: 10.1136/annrheumdis-2020-eular.3393

FRI0244

EOSINOPHILIC FASCIITIS-LIKE SECONDARY TO CHRONIC GRAFT-VERSUS-HOST DISEASE: CLINICAL DESCRIPTION OF 28 PATIENTS

C. Hidalgo¹, C. Román², M. E. Acosta¹, L. Gómez-Lechón¹, M. D. Sánchez³, L. Pantoja⁴, O. Compán¹, S. Pastor¹, C. A. Montilla-Morales¹, L. López Corral⁵.

¹Hospital Universitario de Salamanca (Spain), Rheumatology Unit, Salamanca, Spain; ²Hospital Universitario de Salamanca, Dermatology, Salamanca, Spain;

³Hospital Clínico Universitario de Valladolid, Rheumatology, Valladolid, Spain;

⁴Complejo Asistencial Hospital General de Segovia, Rheumatology, Segovia, Spain;

⁵Hospital Universitario de Salamanca, Haematology, Salamanca, Spain

Background: Eosinophilic fasciitis (EF) is an uncommon chronic inflammatory disease characterized by myalgia, soft tissue hardening, peripheral eosinophilia and increased acute phase reactants, often triggered after strenuous physical exercise. Its appearance has been described as a rare complication after allogeneic transplantation of hematopoietic progenitors in the context of chronic

graft-versus-host disease (cGVHD). Its etiopathogenesis is not well known and usually the treatment with systemic corticosteroids is effective

Objectives: Describe the clinical and transplant-related characteristics of a cohort of patients with eosinophilic fasciitis-like in the context of cGVHD

Methods: Observational, retrospective and descriptive study of the clinical characteristics of 28 patients affected by EF-like followed in a multidisciplinary consultation of cGVHD, started in March, 2014. Regular demographic variables, clinical characteristics related to the transplant and with the cGVHD, laboratory parameters, rescue therapies and their response were collected. The statistical analysis was done with Microsoft Excel 2007.

Results: Seventeen (60.7%) patients were male and 11 (39.35%) were women with a mean age of 48.75 years (range from 10 to 74). Acute myeloid leukemia was the most frequent cause of the transplant in 11 patients (39.3%). Transplant related characteristics are reflected in Table 1 and the clinical manifestations, therapies received and their response in Table 2. Four (14.2%) patients died during their follow-up, being the cause of death in 2 cases due to sepsis, and in 1 case attributable to GVHD.

Table 1. Baseline and transplant related characteristics (N = 28).

Variables	N (%)
Donor Type (related / not related)	13(46.4%)/15(53.6%)
Type of conditioning (reduced intensity / myeloablative)	18(64.25%)/10(35.7%)
Source of cells (Peripheral blood / bone marrow)	27(96.4%)/1(3.6%)
cGVHD type quiescent / de novo / progressive	11(39.3%)/13(46.4%)/4(14.3%)
Other affected organs (cGVHD score)	
-Mouth	6(21.4%)
-Eyes	10(35.7%)
-Lung	2(7.1%)
-Liver	3(10.7%)
-Gastrointestinal tract	0(0%)
-Genital	2(7.1%)
-Cutaneous	16(57.14%)
Global Score	4(14.2%)/14(50%)/10(35.7%)
NIH ¹ (mild / moderate / severe)	

¹ NIH: National Institute of Health.

Table 2. Clinical manifestations and therapies (N = 28).

Variables	N (%) / Median (range)
Prodromic symptoms: yes / no	20(71%)/8(29%)
- Stiffness	2(7.1%)
- Artromyalgia	17(60.7%)
- Edema	3(10.7%)
Time until first visit	31.3 months (range 9-73)
Contracture Yes / No	18(64.3%)/10(35.7%)
Mobility limitation (mild / moderate)	13(46.4%)/9(32.1%)
ECOG ¹ affected	11(39.2%)
Eosinophilia	17(60.7%)
Positive autoantibodies	8(28.5%)
First line therapies (corticosteroids)	28(100%)
Extracorporeal photopheresis	19(67.9%)
Therapies of 2nd line/ 3rd or more	6(21.4%)/12 (42.8%)
Physiotherapy	14 (50%)
Response: complete / sequels	10(35.7%)/18(64.2%)

¹ECOG: Eastern Cooperative Oncology Group scale to assess the quality of life

Conclusion: Nonspecific joint symptoms such as stiffness, edema or arthromyalgia in patients undergoing allogeneic transplantation of hematopoietic progenitors may be factors that predict the development of sclerotic GVHD type eosinophilic fasciitis-like and should be closely monitored in order to be able to perform early stage diagnoses of the disease. It is necessary to deepen the pathogenesis of this entity and the multidisciplinary approach to improve the prognosis of patients with GVHD

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3389

FRI0245

PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS IS NEARLY ALWAYS ACCOMPANIED BY A LOW DIFFUSING CAPACITY

A. Hu¹, T. Nevskaya², M. Baron², J. Pope¹ on behalf of Canadian Scleroderma Working Group (CSRG). ¹Western University, London, Canada; ²McGill University, Montréal, Canada

Background: Scleroderma (systemic sclerosis; SSc) has high morbidity and mortality. Pulmonary hypertension (PH) and pulmonary arterial hypertension

(PAH) is common with a high mortality (1). SSc patients are screened with pulmonary function tests (diffusing capacity of the lung for carbon monoxide; DLCO) (2).

Objectives: The DLCO%predicted was analyzed comparing patients with and without PAH to determine if it is always low at time of PAH diagnosis.

Methods: The Canadian Scleroderma Research Group (CSRG) database was used containing more than 1300 SSc patients with a mean disease duration of 8 years. All patients with at least one follow up visit and DLCO recorded at least twice were eligible for enrolment into this nested case control study. Diagnosis of PH was verified using several algorithms within the database including R heart catheterization, use of PH medications and physician response of 'yes' to question has this patient been diagnosed with pulmonary hypertension. Sensitivity, specificity and positive (PPV) and negative predictive values (NPV) were calculated for DLCO%predicted <50% and presence of PH/PAH.

Results: At time of PH diagnosis, the mean DLCO% predicted was 47% (N=30) vs no PH 73% (N=960) $P<0.0001$, and proven documented PAH also showed the differences (PAH, N=22 DLCO% predicted 51% vs. PAH negative (N=968) DLCO% pred 72%, $P<0.0001$) (Table 1). The OR of a DLCO%predicted less than 60 was 4.7 for PAH and 7.6 for PH (both $P<0.001$) and even higher if DLCO<50% (OR 11.5 for PH and 7.6 for PAH). Table 2 shows the PPV of DLCO at varying levels.

Table 1. DLCO comparison between PH+ and PH- SSc patients and between PAH+ and PAH- SSc patients, at the time of diagnosis.

	PH+ (n=30)	PH- (n=960)	P-value	PAH+ (n=22)	PAH- (n=968)	P-value
mean±SD	47.17±17.53	72.74±20.79	<0.0001	51.23±17.55	72.44±20.99	<0.0001
Range	18-81	13-147		25-81	13-147	

Table 2. Sensitivity, specificity and predictive values in SSc-PH and -PAH for DLCO at various cut points.

		OR	PPV	Specificity	NPV	Sensitivity
PH	DLCO<50%	11.5 (CI 95% 5.4-24.8), $p<0.0001$	13.2%	87.0%	98.7%	63.3%
	DLCO<60%	7.6 (CI 95% 3.3-17.2), $p<0.0001$	7.9%	73.3%	98.9%	73.3%
	DLCO>80%	0.06 (CI 95% 0.008-0.46), $p=0.007$	0.3%	64.5%	95.5%	3.3%
PAH	DLCO<50%	7.6 (CI 95% 3.2-17.9), $p<0.0001$	8.3%	86.4%	98.8%	54.5%
	DLCO<60%	4.7 (1.9-11.3), $p<0.001$	5.0%	72.7%	98.9%	63.6%
	DLCO>80%	0.1 (0.01-0.7), $p=0.018$	0.3%	64.8%	96.8%	4.5%

Conclusion: A low DLCO is associated with a high odds of PH/PAH in SSc and the NPV is very high at both DLCO<50% predicted and <60% predicted. This may aid in determining who should receive a right heart catheterization in SSc patients.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3394

FRI0246

GENOTYPING OF CYP3A5 IS USEFUL FOR TREATMENT WITH TACROLIMUS IN INTERSTITIAL LUNG DISEASE IN DERMATOMYOSITIS

M. Katayama¹, S. Horibata^{2,3}, S. Takahashi^{1,2}, M. Takahashi², S. Mukohara¹, N. Amano¹, K. Yoshida¹, S. Hatachi¹, K. Yorifuji^{2,3}, S. Kumagai^{1,2}. ¹Center for Rheumatic Disease, Shinko Hospital, Kobe, Japan; ²Shinko Institute for Medical Research, Kobe, Japan; ³Department of Pharmacy, Shinko Hospital, Kobe, Japan

Background: Tacrolimus (TAC), an immunosuppressant, can be used in second-line maintenance therapy of interstitial lung disease (ILD) in patients with dermatomyositis (DM) [1]. In Japan, TAC is approved for DM-ILD and often used as induction therapy for severe cases, especially in patients with anti-MDA5-Ab (melanoma differentiation-associated gene 5 antibody) positive, in combination with glucocorticoids (GC) and intravenous cyclophosphamide (IVCY). Some studies reported the clinical efficacy of initial high-trough level TAC for DM-ILD in combination with GC and IVCY [2]. Adjustment to target concentration of TAC in early stage of treatment is important for controlling disease activity. The

concentration of TAC depends on genetic polymorphisms in cytochrome P450 (CYP) 3A5 enzyme expression and several reports showed that the bioavailability and concentration of TAC in patients with a CYP3A5 *1 allele (*1) was lower than those with a CYP3A5 *3/*3 (*3/*3) [3].

Objectives: To examine the usefulness of CYP3A5 polymorphisms in decision of initial dose to achieve the target concentration of TAC and to evaluate the clinical efficacy in patients of DM-ILD who achieved the enough concentration of TAC in early stage of treatment.

Methods: We investigated CYP3A5 polymorphisms and TAC concentration in 9 patients of DM-ILD without renal dysfunction (eGFR>60). TAC was taken after both morning and evening meals and blood samples were taken 12h after TAC administration. The blood concentration of TAC normalized to the corresponding dose per body weight (C/D, ng/ml per mg/kg) was analyzed according to genetic variation in CYP3A5. Based on the retrospective analysis, we chose proper dose of TAC in initial treatment for an anti-MDA5-Ab positive DM-ILD patient, whose genotype was *3/*3.

Results: The C/D value in the *3/*3 group (n=6) was 154.6±25.6, which was significantly higher than that in the *1 group (n=3; 79.0±2.8; $P=0.028$). When the target concentration was set at 5-10 ng/ml, the required dose was (0.0316 to 0.0633) mg/kg in the *1 group and (0.0162 to 0.0324) mg/kg in the *3/*3 group. The *1 group needs more dose than that of the *3/*3 group to achieve the same target trough of TAC, suggesting that the examination of CYP3A5 genotype is useful for deciding initial dose of TAC (Fig.1). We started TAC 6mg/day with setting target concentration at 15-20 ng/ml to a *3/*3 patient of DM-ILD with anti-MDA5-Ab positive, resulting in good clinical course with making rapid attainment of target concentration in early stage of treatment (Fig.2).

Conclusion: To examine the CYP3A5 genotype is valuable for deciding the initial dose of TAC, especially in patients who need achievement to target concentration rapidly.

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Acknowledgments: none

Disclosure of Interests: motoko katayama: None declared, Shinji Horibata: None declared, Soshi Takahashi: None declared, Miho Takahashi: None declared, Saki Mukohara: None declared, Norihiko Amano: None declared, Katsuyuki Yoshida: None declared, Saori Hatachi: None declared, Kennosuke Yorifuji: None declared, Shunichi Kumagai Grant/research support from: Astellas, Chugai, Mitsubishi Tanabe Co.Ltds, Consultant of: Sysmex Co.Ltd, Speakers bureau: many companies

DOI: 10.1136/annrheumdis-2020-eular.4122