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FRI0332 IL-2 RESTORED THE REDUCED ABSOLUTE NUMBER OF TREG CELLS IN THE PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHY

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Background: Idiopathic inflammatory myositis (IIM) is a group of skeletal muscle non-suppurative inflammatory disease, including polymyositis (PM), dermatomyositis (DM), inclusion body myositis (IBM), DM/PM associated with malignancy and so on. The latest studies suggest that there is an increase of Th17 cells levels and a decrease of regulatory T cells (Tregs) levels in IIM, which lead to the Th17/Treg imbalance. IL-2 promotes the proliferation and differentiation of several subsets of CD4⁺T cells and inhibits Th17 cells. It is provided to Tregs from IL-2-producing cells such as activated CD4⁺CD25^{low} T cells, NK and NKT cells. Tregs express IL-2R $\alpha\beta\gamma$, a high affinity IL-2 receptor complex. Therefore, Tregs may be selectively stimulated by low-dose IL-2 and amplified.

Objectives: To explore the quantitative changes of peripheral Th17 cells and Tregs in IIM before and after receiving treatment with low-dose interleukin-2 (IL-2) and analyze the relationship between peripheral Th17 cells and Tregs and clinical indicators disease activity.

Methods: Total 151 IIM patients were enrolled, and 196 healthy adults were used as normal controls. Of them, 76 cases were treated with low dose IL-2 (5.0*10⁵ international units (IU) for 5 days). The absolute number of peripheral T, CD4⁺ T, CD8⁺ T, Th1, Th2, Th17 and Treg subsets were analyzed by flow cytometry. Laboratory examinations were analyzed retrospectively. Since the data was disregarded from the normal distribution, the median four quantile method was used for statistical description. Multiple samples were compared with Kruskal-Wallis H test, and the correlation between variables was Spearman rank correlation analysis.

Results: (1) In the patients, the absolute number of Treg cells was significantly decreased as compared with that in the control group (P<0.05) and increased by treated with a low dose IL-2. After IL-2 treatment, the absolute numbers of T, CD4⁺ T, CD8⁺ T, Th1, Th2 and Th17 cells and the ratio of Th17/Treg were still significantly lower than that in the control group (P<0.05); the ratio of Th17/Treg in the patients was not significantly different from that in the control group (P>0.05). (2) The peripheral Th17 cells levels were negatively correlated with ESR, CRP, LDH and HBDH (r=-0.42, r=-0.28, r=-0.21, r=-0.20, P<0.05); the peripheral Treg cells levels were negatively correlated with ESR, CRP, LDH and HBDH (r=-0.18, r=-0.20, r=-0.18, r=-0.20, P<0.05); the ratio of Th17/Treg was negatively correlated with ESR (r=-0.23, P<0.05).

Conclusion: The absolute number of peripheral Th17 cells and Tregs declined significantly in IIM, and had correlation with ESR, CRP, LDH and HBDH. However, after receiving treatment with low-dose IL-2, the Tregs levels elevated, which means that IL-2 may plays an essential role in the differentiation and development of Tregs. It also provides new thought and strategy for the therapy of IIM.

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FRI0333 ASSESSING SYSTEMIC SCLEROSIS CARDIAC INVOLVEMENT USING MAGNETIC RESONANCE T1-MOLLI MAPPING

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Background: Patients with systemic sclerosis (SSc) and symptomatic cardiac involvement have a disease-associated mortality of 20% in 10 years. Prevalence of subclinical cardiac involvement is 15%-35%. Early diagnosis and monitoring is therefore critical in the management of SSc-associated cardiac involvement. Conventional cardiac magnetic resonance imaging (cMRI) T1-weighted sequence with late gadolinium contrast has good accuracy for detecting localized fibrosis but not diffuse fibrosis that occurs in SSc. Modified look-locker inversion recovery (T1-MOLLI) mapping, a novel cMRI sequence, has been used to detect and quantify diffuse fibrosis in aortic stenosis. T1-MOLLI values have also been correlated with the degree of biopsy-quantified fibrosis.

Objectives: To investigate whether cMRI T1-MOLLI is able to detect fibrosis associated with SSc cardiac involvement.

Methods: We recruited 16 patients fulfilling the 2013 ACR/EULAR criteria for SSc and 17 healthy age-matched (within 5 years) controls. Patients underwent cMRI T1-MOLLI mapping on the Siemens Biograph mMR 3.0T (Erlanger, Germany).

T1-MOLLI values were compared between SSc patients and healthy controls using student t-test and statistical significance was taken to be p<0.05.

Results: Demographics and clinical features of our study cohort are as shown in Table 1. Two (12.5%) SSc patients were symptomatic with palpitations or had arrhythmia requiring treatment. Eleven (68.8%) SSc patients had elevated T1 MOLLI values, using a normal cut-off threshold of 1284 ms.

Mean cardiac T1 values were significantly higher in SSc patients (1329 ± 88.0 ms) than in controls (mean T1 = 1238 ± 93.7 ms, p=0.0075), indicating the presence of cardiac fibrosis.

Conclusion: cMRI T1-MOLLI mapping demonstrated that SSc patients who were predominantly asymptomatic showed evidence of cardiac fibrosis. cMRI T1-MOLLI is potentially a useful diagnostic and monitoring tool for SSc cardiac disease.

Table 1. Demographics and clinical features

	SSc patients (n=16)	Controls (n=17)
Limited/Diffuse SSc, n	11/5	Not applicable
Female, n (%)	14 (87.5%)	14 (82.3%)
Age, years	49.2 ± 13.9	51.5 ± 14.8
Mean disease duration from Raynaud's phenomenon onset, years	6.1 ± 7.3	Not applicable
Mean disease duration from non-Raynaud's phenomenon onset, years	6.6 ± 7.4	Not applicable
Hypertension, n (%)	4 (25%)	1 (5.9%)
Palpitations, n (%)	2 (12.5%)	0
Arrhythmia requiring treatment, n (%)	2 (12.5%)	0
Anti-centromere positivity, (n)	3	Not applicable
Anti-Topo-I positivity, (n)	4	Not applicable

Disclosure of Interests: None declared

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FRI0334 PERFORMANCE OF THE SCLERODERMA SKIN PATIENT-REPORTED OUTCOME (SSPRO) IN A PHASE 2 TRIAL WITH LENABASUM

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Background: Skin thickening is a distressing feature of systemic sclerosis (SSc). The severity and extensiveness of skin thickening, as traditionally

assessed by mRSS, may not directly correlate with its effect on patients' health related quality of life (HRQoL). The Scleroderma Skin Patient-reported Outcome (SSPRO) was specifically developed to assess the skin-related HRQoL in SSc. No skin-specific PRO in SSc has been prospectively validated in a clinical trial.

Objectives: Validate the SSPRO prospectively in a clinical trial.

Methods: The SSPRO, Patient Global Assessment (PtGA), Physician Global Assessment (MDGA), HAQ-DI, mRSS, FVC% predicted, PROMIS-29 questionnaire, and ACR CRISS were assessed prospectively in a Phase 2 study of lenabasum in dcSSc. SSPRO has 18 items that assess four SSc skin-related HRQoL domains. The Phase 2 study of lenabasum had a 4-month double-blinded portion (N = 41 completers) followed by an open-label extension (N = 38 entered). Spearman correlations of baseline values and change values were determined for SSPRO and other outcome measures. Mean change in SSPRO scores were determined in subjects with increasing levels of improvement in other efficacy outcomes.

Results: At baseline, SSPRO correlated moderately with all other outcome measures except for FVC%, as expected, with strongest correlations with PtGA, HAQ-DI, and PROMIS-29 pain interference and social role domains (Table 1). The mean change in SSPRO scores at 3 and 12 months correlated mostly moderately ($r = 0.25$ to 0.62) with the mean change in other outcome scores, but correlations were low or inconsistent with mRSS, FVC%, and PROMIS-29 anxiety domain (Table 1). Significance of correlations in some outcomes was hampered by small magnitudes of change.

Mean SSPRO generally increased in subjects with increasing levels of improvement in HAQ-DI, PtGA, mRSS, and ACR CRISS, although less consistently with mRSS and ACR CRISS at 3 months (Table 2).

Conclusion: SSPRO score correlates with both physician and patient-reported outcomes at baseline, though correlations with the latter were stronger. Change in SSPRO reflects changes in how the patient feels (PtGA, PROMIS-29 pain interference and depression domains) and functions (HAQ-DI, PROMIS-29 physical function and social role domains) and less consistently with physician-assessed outcomes. Subjects with improvement $\geq$ MID levels of -0.250 in HAQ-DI, -1 in PtGA, and -5 in mRSS had SSPRO improvements of -22 to -23 points at 12 months.

Disclosure of Interests: Ada Man Consultant for: Advisory Board Fee: Abbvie, Janssen, Boehringer Ingelheim, Nancy Dgetluck Employee of: Corbus Pharmaceuticals, Inc., Brian Conley Employee of: Corbus Pharmaceuticals, Inc., Barbara White Shareholder of: Corbus Pharmaceuticals, Inc., Employee of: Corbus Pharmaceuticals, Inc.

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FRI0335

PROGNOSTIC IMPACT AND CLINICAL CHARACTERISTICS OF INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES IN A MULTIDISCIPLINARY SETTING

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Background: Interstitial Pneumonia with Autoimmune feature (IPAF) is a recently defined disease that includes Interstitial Lung Disease (ILD) patients with features of autoimmunity, not satisfying any of the established classification criteria for connective tissue diseases (CTD). Even if IPAF patients share clinical and serological findings with CTD, we are unaware about the clinical evolution of IPAF patients.

Objectives: To define the characteristics and evolution of IPAF patients in a multidisciplinary setting.

Methods: We selected patients with a diagnosis of IPAF referring to a multidisciplinary Rheumatology/Pneumology/Radiology team at our hospital. We excluded from the analysis patients positive for antisynthetase antibodies, as diagnosed with antisynthetase syndrome. Data were retrospectively collected from our hospital medical records.

Results: We analyzed 25 patients (19 females, 76%, 6 males, 24%), with a median onset age of 67 years (interquartile range, IQR, 59-74) and a follow-up of 32 months (IQR 22-69). ANA test was positive in 23 (92%) cases (fig 1), whereas cytoplasmic positivity was observed in 17 (68%). Anti-ENA screen was positive in 9 patients (36%) (7 [28%] anti-Ro52 and 2 [8%] anti-RNP). One patient (4%) was positive for anti-PM-Scl, 1 (4%) for anti-Mi2 and 1 (4%) for anti-Ku antibodies. These 2 latter antibodies are not included in the serological domain of IPAF. Patients had mainly a Non Specific Interstitial Pneumonia (NSIP) pattern (19 [76%]; fibrosis in 7 [28%], and with concomitant organizing pneumonia (OP) in 3 [12%]). Three (12%) patients had Usual Interstitial Pneumonia-like and 3 (12%) an OP pattern. The majority of patients (15 [60%]) satisfied only the morphological and serological domains. Clinical domains satisfied were: arthritis (4 [16%]), Raynaud's phenomenon (5 [20%]), palmar telangiectasias (2 [8%]), mechanic's hands (1 [4%]) and hiker's feet (1 [4%]). We observed other findings not included in IPAF criteria but suggestive for CTDs, both clinical (inflammatory myopathy 3 [12%]; dry eye 1 [4%]; sclerodactyly 1 [4%]), and instrumental (scleroderma pattern at nailfold capillaroscopy 1 [4%]; dilated esophagus at barium X-rays 2 [8%]). Clinical spectrum time course was variable in 5 (20%) cases: 3 (12%) patients developed arthritis after ILD, and 2 (8%) developed ILD respec-

Table 1. Spearman Correlations for SSPRO and Other Efficacy Outcomes at Baseline and for Change in SSPRO and Other Efficacy Outcomes at 3 and 12 months (r, P-value)

Time	PtGA	MDGA	HAQ-DI	mRSS	FVC,%	ACR CRISS	Pain Interference	Social Role	PROMIS-29 Domain Physical Function	Depression	Anxiety
Baseline	0.61 <0.0001	0.51 0.0007	0.52 0.0005	0.44 0.004	-0.07 0.670		0.62 <0.0001	-0.57 0.0001	-0.33 0.034	0.51 0.0007	0.41 0.008
Change 3 months	0.26 0.124	0.30 0.067	0.29 0.083	0.02 0.932	-0.12 0.483	-0.20 0.235	0.46 0.005	-0.40 0.015	-0.46 0.004	0.33 0.044	0.14 0.395
Change 12 months	0.28 0.103	0.15 0.394	0.25 0.145	0.20 0.251	-0.53 0.001	-0.48 0.004	0.41 0.014	-0.62 < 0.0001	-0.25 0.152	0.27 0.119	0.21 0.234

Table 2. Mean Change in SSPRO Associated with Increasing Levels of Improvement in Other Outcomes

Efficacy Outcome	HAQ-DI improvement				PtGA improvement			mRSS improvement				ACR CRISS ¹			
Improvement at least	None	-0.125	-0.250	-0.500	None	-1	-2	None	-3	-5	-8	< 0.30	≥ 0.30	≥ 0.60	≥ 0.90
3 months (N=37)	-9.9	-17.6	-20.2	-22.5	-11.9	-15.3	-16.5	-11.0	-14.7	-16.3	-10.3	-10.2	-17.8	-17.1	-14.5
12 months (N=35)	-17.9	-20.2	-22.7	-27.2	-16.5	-21.9	-21.5	-11.6	-19.6	-23.2	-23.1	-13.0	-23.5	-26.6	-27.3

¹ACR CRISS score is absolute, not change, score.