retrospectively included. The presence of a halo or occlusion was considered a positive CDU finding for GCA. In patients with PET-proven extra-cranial GCA, an axillary FDG uptake higher than liver uptake was considered positive. The reference was the clinical diagnosis (GCA or not) after 6 months.

Results: Of the 113 included patients, GCA was diagnosed in 41 and excluded in 72 patients at 6 months reference. PET-proven extra-cranial involvement was present in 20/41 GCA patients, of which 13 showed axillary involvement. An axillary halo was found in 8/13 of these patients, consistent with an increased FDG uptake. In 5/13, axillary involvement on PET was missed with axillary CDU (1/5 on steroids). However, in 4/5 of these an halo was found in the temporal arteries. Additionally, 6/20 patients with extra-cranial involvement did not show axillary involvement on both CDU and PET. All 6 had isolated involvement of a single artery on PET (2 aorta; 4 vertebral). CDU found a temporal halo in 3 of them. Overall, CDU of only the temporal arteries resulted in a sensitivity of 52% (95CI 35-67) and a specificity of 93% (95CI 84-97) for the reference diagnosis at 6 months. Adding CDU results of axillary arteries improved sensitivity to 71% (95CI 55-84), while specificity did not change. Although not systematically assessed, adding CDU assessment of the subclavian arteries improved sensitivity to 76% (95CI 59-87).

Conclusion: In case of an axillary halo no further FDG PET/CT scanning is required to diagnose extra-cranial GCA. In this study, investigation of both temporal and axillary arteries substantially increased the diagnostic performance of CDU. Furthermore, CDU and FDG PET arterial involvement is not always congruent, and occasionally arterial involvement is restricted to vessels not systematically assessed with CDU in our center. In patients in whom the axillary CDU is negative, a targeted CDU of selected vessels (e.g. subclavian, carotid or vertebral), or an FDG PET/CT is mandatory.

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Difficult to manage Sjögren's syndrome and Myositis

OP0212

DIGESTIVE INVOLVEMENT IN PATIENTS WITH PRIMARY SJÖGREN SYNDROME FROM THE SJÖGREN SPANISH REGISTRY

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Background: Primary Sjögren's syndrome (pSS) is a systemic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands. Among the extraglandular manifestations, digestive involvement (DI) is frequent, and may condition the quality of life of these patients. There are few studies that systematically analyze gastrointestinal involvement in pSS.

Objectives: To describe the prevalence of DI in patients with pSS included in SJÖGREN Spanish registry and the phenotype of these patients, and to analyze their association with other clinical manifestations, serological markers, activity index and treatments used.

Methods: Transversal multicenter study that includes all patients of SJÖGREN registry (patients who meet 2002 pSS classification criteria), a study conducted in 33 units of rheumatology in Spain between 2014-2016, which collected demographic and clinical and serological data (1). Patients were classified according to the presence of DI (that included esophagus, stomach, intestine, liver and pancreas). A descriptive analysis was done, with means and standard deviations, frequencies and proportions. Student t for quantitative variables and Chi2 for qualitative ones were performed to evaluate differences between groups. The association of DI with other variables was analyzed by bivariate and multivariate binomial logistic regression analysis. In the multivariate model all variables with p < 0.2 in the bivariate were included, and the effect of age and sex was controlled.

Results: From 437 patients included (95% women, median age of 58 years), 59 (13.5%) had some DI (21 (36%) chronic atrophic gastritis, 12 (20%) esophageal dysmotility, 3 (5%) lymphocytic colitis, 23 (39%) other). 54% of patients developed DI at the time of pSS diagnosis or later, and 45% before the diagnosis, with a mean age of 49 years at the onset of pSS symptoms. Patients with DI were older, both at diagnosis, onset of pSS symptoms and inclusion in the cohort, than those without it. There were no differences in the ESSDAI index between both groups. Comparison regarding clinical and serological characteristics is shown in Table 1. Patients with DI had more thyroid involvement and C3 hypocomplementemia. In addition, patients with DI had been treated more frequently with glucocorticoids and immunosuppressants.

In the multivariate analysis, DI was significantly associated with older age at diagnosis (OR 1.03 (1.00-1.05); p=.005), more C3 hypocomplementemia (OR 2.40 (1.10-5.26) p=.027) and absence of anti-Ro antibodies (OR 0.33 (0.12-0.87) p=.025).

Conclusion: In patients with pSS from the SJÖGREN cohort, the prevalence of digestive involvement is 13.5%. These patients are older at diagnosis, have more C3 hypocomplementemia and negative anti-Ro antibodies. These patients need more frequently glucocorticoids and immunosuppressants, suggesting a more severe disease phenotype.

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	WITH DI (n=59; 13.5%)	WITHOUT DI (n=378; 86%)	P value
Age symptoms pSS onset	49 ± 13	45 ± 13	0.006*
Age diagnosis pSS	54 ± 12	49 ± 12	0.008*
Age cohort inclusion	62 ± 10	58 ± 13	0.036*
sex	57F (96%), 2M	359F (95%), 19M	0.585
Glandular involvement	23 (39%)	119 (31%)	0.253
Lung involvement	9 (15%)	34 (9%)	0.121
Thyroid involvement	13 (22%)	66 (17%)	0.017*
Raynaud	17 (28%)	75 (19%)	0.116
anti-Ro antibodies	52 (88%)	357 (94%)	0.066
Hypocomplementemia C3	14 (25%)	51 (14%)	0.034*
Hypocomplementemia C4	12 (21%)	50 (14%)	0.131
Glucocorticoid treatment	36 (63%)	177 (47%)	0.026*
DMARD treatment	36 (64%)	180 (49%)	0.035*

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OP0213

MYOFASCIA-DOMINANT: INFLAMMATION DETECTED ON WHOLE BODY MRI PREDICTS RAPIDLY PROGRESSIVE INTERSTITIAL LUNG DISEASE IN PATIENTS WITH DERMATOMYOSITIS

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Background: Rapidly progressive interstitial lung disease (RPILD) is a major cause of death in patients with dermatomyositis (DM). Early diagnosis and aggressive immunosuppressive therapy are required in RPILD to improve the prognosis. Clinically amyopathic dermatomyositis (CADM) and anti-melanoma differentiation-associated gene 5 antibody are known as the risk factors for RPILD. However, 70% of the patients with CADM do not develop RPILD¹. Whole-body magnetic resonance imaging (WB-MRI) can detect inflammation of whole muscle and myofascia. Although recent studies have indicated that MRI could be

useful for assessing disease activity², the relation between MRI findings and RPILD has not been investigated.

Objectives: In this study, we assessed whether WB-MRI findings are related with RPILD in patients with DM.

Methods: This retrospective study comprised 33 patients with DM who underwent WB-MRI in our hospital before the initiation of treatment for myositis. Muscular and myofascial signal abnormalities were scored on 42 muscular groups. The ratio of myofascial score to muscular score (myofascia/muscle ratio) was calculated and used to evaluate myofascia-dominant inflammation. RPILD was defined as the acute onset and rapid worsening of dyspnea, hypoxemia and radiographic ILD/fibrosis within 1 month.

Results: Of 33 patients, 16 were CADM, and 24 had ILD, including 8 patients with RPILD. All patients including CADM showed abnormal signal intensity in muscle and myofascia (scores median: 15 and 21, respectively). Muscle scores positively correlated with serum level of creatine kinase ($r=0.672$, $p<0.001$). The patients with RPILD showed a significantly higher myofascia/muscle ratio compared with that in the non-RPILD patients (2.167 vs 1.000; $p=0.018$). Logistic regression analysis identified myofascia/muscle ratio >1.53 (odds ratio: 141.90, $p=0.042$) and older age (odds ratio: 1.15, $p=0.041$) as independent risk factors for RPILD.

Conclusion: We newly defined myofascia-dominant inflammation using WB-MRI as a predictor to develop RPILD in patients with dermatomyositis.

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Anergy, exhaustion or post-activation in autoimmunity – facts and future consequences_____

OP0214

DISTURBED PD-L1 UPREGULATION IS CHARACTERISTIC OF ANERGIC B CELLS IN SLE

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Background: Prior studies on systemic lupus erythematosus (SLE) B cells reported an altered responsiveness to TLR9 stimulation, such as proliferation, cytokine production as well as indications for impaired T and B cell interaction [1]. The role of co-inhibitory and co-stimulatory (check-point) molecules in this setting has not been delineated in detail so far.

Objectives: Assess the expression of co-inhibitory (PD-1, PD-L1 and PD-L2) and co-stimulatory molecules (CD86 and CD40) by SLE B cells after *in vitro* stimulation and their potential contribution for B cell hyporesponsiveness in SLE.

Methods: PBMCs from 10 SLE patients and 10 healthy donors (HD) were stimulated with IL-2/IL-10, anti (α) - B cell receptor (BCR), CpG and CD40L alone or in combination. Expression of PD-1, PD-L1, PD-L2 as well as CD86, CD40 on CD19+CD20+ B cell subsets after 48h stimulation was analyzed by FACS. CD71 was employed to measure proliferation of CD19+CD20+ B cells after 48h stimulation.

Results: SLE B cells exhibited a substantially decreased upregulation of PD-L1 ($p=0.0006$) and CD86 ($p=0.0188$, Fig.1) associated with significantly reduced B cell proliferation ($p=0.0039$) after 48h stimulation with CpG alone and in combination compared with HD. While TLR9 engagement in SLE B cells appeared to be abnormal, activation of CD40 resulted into a consistent upregulation of both, inhibitory and stimulatory molecules. PD-L1 was positively correlated with B cell proliferation ($p=0.003$). Notably, the expression of PD-L1 and CD86 correlated inversely with Siglec-1, as surrogate marker for interferon signature ($p<0.0001$ and $p=0.0021$ respectively). PD-L1 expression correlated inversely with cSLE-DAI ($p=0.0087$).

Fig. 1 PD-L1 and CD86 expression by SLE B cells upon stimulation is lower than in healthy donors. A. PD-L1 and B. CD86 expression by CD19+CD20+ B cells after stimulation with IL2/IL10, CpG, anti-BCR and CD40L in healthy donors and SLE patients.

Conclusion: Hyporesponsive/anergic lupus B cells are characterized by a functionally diminished PD-L1 and CD86 upregulation associated with reduced proliferation and clinical activity. The data mandate evaluations of innovative therapeutic interventions in SLE.

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