Table 2. Comparison between F-SS-2 and ISSF Score

		ISSF Score (n=160)				p ^a
		Mild	Intermediate	Severe	Total	
F-SS-2 Score (n=160)	Mild	37 (62.7)	20 (33.9)	2 (3.4)	59 (100.0)	<0.001
	Intermediate	22 (53.7)	17 (41.5)	2 (4.9)	41 (100.0)	
	Severe	8 (13.3)	43 (71.7)	9 (15.0)	60 (100.0)	
	Total	67 (100.0)	80 (100.0)	13 (100.0)	160 (100.0)	

F-SS-2: The second set of Familial Mediterranean Fever severity score, ISSF: International Severity Scoring System for Familial Mediterranean Fever, ^aPearson Chi-Square test

Conclusion: Disease severity scores should be used for treatment choice, evaluation of disease prognosis and classify patients according to disease severity in clinical trials. In the current study, we found a difference between the classifications of disease severity, which could be a result of each scoring system uses different parameters of the disease.

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THU0588 CLINICAL PHENOTYPES OF IGG4-RELATED DISEASE IN SPAIN

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Background: Recently, several clinical phenotypes in IgG4-related disease have been described in a multinational and ethnically variate cohort¹.

Objectives: To assess the clinical presentation of IgG4-related disease (IgG4-RD) in Spanish patients and assess the distribution among different clinical phenotypes.

Methods: Clinical data were obtained from the Spanish IgG4-RD registry (REERIGG4) from October 2013 to December 2018, including 9 centers. We reviewed demographic data and organ involvement. The assignment of clinical phenotypes was done by 2 experts, based on organ involvement and clinical manifestations, following Wallace et al.¹ subsets. The phenotypes were: pancreato-hepato-biliary (HBP), retroperitoneum and aorta (RA), head and neck limited (HNL) and Mikulicz and systemic (MS). A fifth group designated not defined (ND), included the patients that did not fit in the previous phenotypes.

Results: One-hundred patients were included. Thirty-four (34%) were females, median age at diagnosis was 54.8 years (IQR 20.7). The ethnicity of the participants was: Caucasian 83%, Hispanic 12% and North-African/Middle-East 5%. Ninety-two percent were diagnosed with a biopsy. Regarding the diagnostic criteria, 85% met consensus pathology criteria and 94% comprehensive criteria.

Fifty-one patients (51%) had systemic IgG4-RD involving >1 tissue. The most commonly involved tissues were: retroperitoneum (35%), lymph nodes (19%), orbit pseudotumor (18%), salivary glands (16%) and pancreas (14%). Forty-two patients (42%) had elevated serum IgG4.

The representation of each clinical phenotype was: HBP 14%, RA 25%, HNL 26%, MS 20%, ND 15%. Patients were equally distributed from the perspective of ethnicity. Men were predominant in all groups (71, 84, 70, 73%) except in HNL (61% for women). Systemic disease was present in all the MS patients, but in 1/3 of the other groups. Serum IgG4 was elevated in 86% of the cases in HBP, 28% in RA, 19% in HNL, 60% in MS and 40% in ND.

Conclusion: The Spanish IgG4-RD population was mainly ethnically Caucasian. Few patients had serum IgG4 elevation. The most frequent phenotype was HNL, followed by RA. The HBP phenotype was less frequent than in previous reports. The influence of race could modify the clinical expression of IgG4-RD. Knowing the regional phenotypes of IgG4-RD may help clinicians improve disease management.

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THU0589 FREQUENCY, CHARACTERISTICS AND CLINICAL DETERMINANTS OF "PRODROME" IN FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Prodrome was defined by the presence of manifestations that precede an FMF attack and predict its emergence. This period might be turned to a window of the opportunity for the prompt treatment of impending attacks with fast-acting IL-1 inhibitors.

Objectives: We aimed to determine the frequency, characteristics and clinical determinants of prodrome in patients with FMF

Methods: 401 FMF patients were enrolled in this cross-sectional study. We applied a questionnaire to the FMF patients about attack types, pre-attack manifestations and latent time. Four hours was accepted as the cutoff point for prodrome as based on the literature, which is compatible with the pharmacokinetics of anakinra.

Results: The mean age was 37.7 (±11.0) years, and the disease duration was 20 years (3-58). 248 of the patients (61.8%) were female. MEFV gene mutations were achieved in 364 (90.8%) patients, of these 121 (30.2%) patients harbored homozygous exon 10 mutations. Patients with a latent time over four hours considered as prodrome positive (PP) patients (n=141). Male gender, homozygous MEFV mutations, peritonitis, pleuritis, arthritis and ELE were more pronounced in PP group. 25.9% of patients with peritonitis, 14.7% of patients with pleuritis, 13.2% with arthritis and 5.5% with ELE had prodrome with variable latent time durations (median, 6.7(4-72), 6 (4-48), 6 (4-72), and 7.7 (4-30) hours, respectively). Prodrome was found to be more common in those with peritonitis. Male gender, having peritonitis or arthritis were found to be the independent clinical determinants of having prodrome (RR 1.72 (1.07-2.76), P=0.02, 4.27 (1.80-10.1), P=0.001, 1.77 (1.04-3.04), P=0.04, respectively). Age, MEFV mutations, pleuritis and ELE were not found as clinical determinants of prodrome.

Conclusion: Especially male patients with peritonitis or arthritis tend to have prodrome more than other patients, and should be questioned. Prodrome positive patients are candidates for prevention of the impending attacks with on-demand treatments.

Table 1. Patients demographics and disease characteristics

	Prodrome (+) (n=141)	Prodrome (-) (n=260)	P
Age, years	37.7 (10.5)	37.8 (12.8)	0.92
Female	78 (55.3%)	170 (65%)	0.05
Age at FMF diagnosis, years	24(3-62)	25(2-59)	0.52
Disease duration, years	21 (4-58)	19 (3-58)	0.08
Fever	0	9 (3.5%)	0.03
Peritonitis	133 (94.3%)	197 (75.8%)	<0.001
Pleuritis	92 (65.2%)	123 (47.3%)	0.01
Arthritis	103 (72.9%)	129 (49.8%)	<0.001
Myalgia	0	6 (2.3%)	0.09
Erysipelas like erythema	46 (32.6%)	58 (22.3%)	0.02
Homozygous*	57 (43.2%)	64 (27.6%)	0.02
M694V/M694V	50(35.5%)	51(19.6%)	0.01
M694V/-	19 (13.5%)	46(17.7%)	0.19
M694V/M680I	13 (9.2%)	23 (8.8%)	0.98
M694V/V726A	12 (8.5%)	20 (7.7%)	0.88
M694V/E148Q	6 (4.3%)	15 (5.8%)	0.45
M680I/M680I	6 (4.3%)	9(3.5%)	0.76

* 364 patients MEFV mutations were achieved. 9.2% of patients MEFV mutations is not known, 7.7% of patients is gen negative FMF.

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