Abstract AB0986 – Table 1

Patient	Sex	Diagnostic age	Onset age	Arthritis	Location	Other musculoskeletal manifestations	Extra-articular manifestations	MEFV Mutation	Homo/ Heterocytosis
1	M	47	40	Yes	Knee	No	No	p Glu148Gln	Heterocytosis
2	M	14	13	Yes	Knee	No	No	R202Q	Homocytosis
3	M	22	22	No		No	Lymphocytic myocarditis	p Glu148Gln	Heterocytosis
4	M	52	47	No		Arthralgias	Abdominal pain. Pleuritis	R202Q	Heterocytosis
5	M	39	20	Yes	PIPs	Arthralgias	Abdominal pain	R202Q	Heterocytosis
6	M	61	54	No		Tendinopathy	Recurrent pericarditis	R202Q	Homocytosis
7	M	17	16	No		No	Abdominal pain	R202Q	Heterocytosis

Results: Seven patients with MEFV gene mutations were reviewed, all of them were women, ranging in age from 14 to 61 years old. Two of them had *recurrent knee monoarthritis*, one had a history of *arthritis* in the hands and erratic *arthralgias*, one had erratic arthralgias and two had no musculoskeletal manifestations. The 2 patients with intermittent hydrartrosis responded satisfactorily to *colchicine*, but not the other patients with other musculoskeletal manifestations.
Conclusions: *Genetic testing* of the common *mutations of the MEFV gene* should be considered in patients with *recurrent episodes of monoarthritis* without justifying cause (palindromic rheumatism, intermittent hydrartrosis, etc.)
Disclosure of Interest: None declared
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AB0987 THE RELATIONSHIP BETWEEN ANTI-INTERLEUKIN-1 THERAPIES AND MEFV GENE MUTATIONS IN FAMILIAL MEDITERRANEAN FEVER PATIENTS: A SINGLE CENTER EXPERIENCE

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Background: Familial Mediterranean fever (FMF) is an autoinflammatory disease characterizing recurrent self-limiting attacks of inflammation mainly placed in serosal surfaces of the body with fever (1). Colchicine is an important weapon of the humanity in FMF treatment yet. However, the puzzle of inflammation was solved, and so, inflammasome complex and interleukin-1 were discovered. Eventually, new era started with anti-interleukin-1 agents in colchicine resistant and/or intolerant FMF patients (2).
Objectives: The aim of this study is to evaluate the effectiveness of anti-interleukin-1 (anti-IL-1) agents on the characteristics of attacks, the adverse effects associated with anti-IL-1 treatment, and relation between therapy and MEFV gene mutations in 23 FMF patients with resistant and/or intolerant to colchicine.
Methods: Between January 2015 and December 2016, twenty-three-FMF patients that following-up at Cumhuriyet University Medical Faculty Rheumatology-Internal Medicine Department were included in to the study. Anakinra (69,6%), and canacinumab (30,4%)] were used in 23 FMF patients. 20 cases were resistant to colchicine, 3 were intolerant to colchicine.
Results: The median age of the patients was 28 years,(18–54) and the median age at diagnosis was 20 years (3–50). Of the FMF patients, nine (39.1%) were female and fourteen (60.9%) were male. The distribution of MEFV gene mutation frequencies in the FMF patients was no mutation in one (4,3.%) patient, M694V heterozygous in nine (39,1%), M694V homozygous in 7 (30,4%), E148Q heterozygous in one (4,3%), and compound heterozygous mutation in 3. (13%) patients. 16 patients used anakinra (100mg/day) for 6 mounths (median) and 7 used canacinumab (150mg/2 months) for 3 mounths (median). Four of 7 patiens were resistant to anakinra. After a median follow up 6 months overall clinical response [no attack (18 patients) or decreased frequency of attacks (5 patients)] was %100. In an FMF patient with no mutation, his brother and mother had FMF with no mutations on MEFV gene. We achieved good clinical and laboratory responses in the patient by canacinumab.
Conclusions: Anti-IL-1 agents can be got involved in a new place safely and effectively in FMF patients along with colchicine. In addition, whole gene analysis should be done in refractory FMF patients with no mutations in order to could investigate new mutations, epigenetic mechanisms or other unexplained reasons.
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AB0988 BEHÇET'S DISEASE IN A DEFINED AREA OF NORTHWESTERN SPAIN

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Background: Behçet's disease (BD) is a multisystemic inflammatory chronic disease. There is a wide variation in the clinical features of BD among geographical groups.
Objectives: To determine the demographic and clinical characteristics of BD in a defined area of northwestern Spain (Vigo).
Methods: Patients with BD (International Criteria BD) and seen in the University Hospital of Vigo in Spain, from 1994 to 2016, were retrospectively enrolled. Demographic, clinical, treatment and evolution data were recorded and analyzed using SPSS 22.0.
Results: Our patients were 26 male and 25 female. The mean age at the onset of the disease was 33±11.95 years (11–62). Oral and genital ulcers were seen in 100% and 84.3% respectively and skin lesions in 88.2%. Ocular involvement occurred in 35.3%, neurological disease in 39.2% and gastrointestinal involvement in 29.4% (the area worst affected was colon and small bowel). Vascular disease was present in 33.3%. See table 1. Pathergy test was performed in 18 patients (35.29%) and 10 (55.5%) were positives. HLA B51 was studied in only 13 patients (25.5%) and 8 (61.5%) were positives. 62.7% of patients had no cardiovascular risk factors (CVRF), 27.4% were smokers, 7.8% were hypertensive and 3.9% were hyperlipidemic and diabetic respectively. CVRF were not related to thrombotic events ($p>0.05$). In regard to gender influence, only pseudofolliculitis was significantly more frequent in men ($p<0.001$). There was a trend for increased prevalence of ocular disease and elevated erythrocyte sedimentation rate/serum C-reactive protein in men, and anemia in women, which, however, did not reach statistical significance. Treatment consisted of corticosteroids (92.2%), colchicine (68.6.%) and another immunosuppressive agent (35.3%). During the disease course 78.4% of the patients had an outbreak, 45.1% initiated or changed to immunosuppressive agent and 19.6% used biological drugs. Most of our patients (92.1%) were admitted to hospital and these constitute an evident bias. Two patients died during the followup period, but only one in relation of BD (upper gastrointestinal bleeding and seizures).
Conclusions: Our series has some particular aspects especially the high frequency of gastrointestinal lesions and neurologic involvement. CVRF do not seem to play a role in the development of thrombotic events. Our results confirm the ethnic and geographic variation of BD expression.
Disclosure of Interest: None declared
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Abstract AB0988 – Table 1. Clinical manifestation of our BD patients

Skin lesions 41/55 (88.2%)	Ocular disease 18/51 (35.3%)	Neurologic disease 20/51 (39.2%)	Vascular disease 17/51 (33.3%)	Gastrointestinal involvement 15/51 (29.4%)
Pseudofolliculitis 58.8%	Uveitis 23.5%	Aseptic meningitis 11.7%	Venous thrombosis 21.6%	Pain 11.7%
Erythema nodosum 39.2%	Retinal vasculitis 5.9%	Seizures 7.8%	Arterial thrombosis 7.8%	Bleeding 9.8%
Leukocytoclastic vasculitis 5.9%	Episcleritis 3.9%	Cerebral venous thrombosis 5.9%	Mixed thrombosis 2%	Diarrhea 5.9%
	Vascular occlusion 2%	White matter lesions 5.9%	Aneurysm 2%	Fever 2%
		Stroke 3.9%		
		Paresthesias 3.9%		
		Others 5.9%		

AB0989 RHEUMATOLOGIC COMPLICATIONS OF THALASSEMIA: SHOULD RHEUMATOLOGISTS JOIN THE MANAGEMENT TEAM?

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Background: Beta-thalassemia major is accompanied by progressive multiple organ systems involvement due to the disease pathophysiology as well as iron overload from blood transfusions on a regular basis. Rheumatologists are not frequently involved in the multidisciplinary management of the disease, in which rheumatologic complications are relatively common.