bDMARDs. Systemic glucocorticoids and Methotrexate were used more frequently in scleritis (Table 1). The main indication for biologic therapy was related to underlying IMID in both groups, but 7 bDMARDs in scleritis were indicated for systemic and ocular compromise.
BVCA and IOP improved significantly after systemic treatment in scleritis (Figure 1).

Table 1. Underlying diseases and systemic treatment.

	Overall (n =175)	Episcleritis (n=135)	Scleritis (n=40)	p
Age (years), mean ± SD	48.9 ± 14.2	47.8 ± 14.3	52.6 ± 13.9	0.061
Sex (women), n (%)	106 (60.6)	81 (60)	25 (62.5)	0.920
UNDERLYING DISEASE				
-Idiopathic, n (%)	81 (46.3)	65 (48.1)	16 (40)	0.364
-Infectious, n (%)	11 (6.3)	7 (5.2)	4 (10)	0.276
-IMID, n (%)	76 (43.4)	57 (42.2)	19 (47.5)	0.563
◦Spondyloarthritis	21 (12)	17 (12.6)	4 (10)	0.787
◦Crohn's disease	16 (9.1)	14 (10.4)	2 (5)	0.469
◦Rheumatoid Arthritis	14 (8)	12 (8.9)	2 (5)	0.740
◦Granulomatosis with polyangiits	7 (4)	3 (2.2)	4 (10)	0.080
◦Relapsing polychondritis	6 (3.4)	4 (3)	2 (5)	0.621
◦Systemic lupus erythematosus	5 (2.9)	2 (1.5)	3 (7.5)	0.079
◦Ulcerative colitis	3 (1.7)	2 (1.5)	1 (2.5)	0.796
SYSTEMIC TREATMENT	72 (41.1)	37 (27.4)	35 (87.5)	**0.000***
-Systemic glucocorticoids, n (%)	72 (41.1)	37 (27.4)	35 (87.5)	**0.000***
-Methotrexate, n (%)	39 (22.3)	17 (12.6)	22 (55)	**0.000***
-Non-methotrexatecDMARD, n (%)	35 (20)	24 (17.8)	11 (27.5)	0.177
-TNFibDMARD, n (%)	27 (15.4)	19 (14.1)	8 (20)	0.362
-Non-TNFibDMARD, n (%)	8 (4.6)	5 (3.7)	3 (7.5)	0.386

*p<0,05*p<0,05

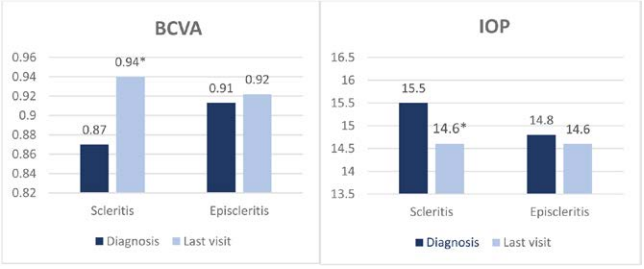

Figure 1. BVCA and IOP at diagnosis and last visit.

Conclusion: OSP is a relatively frequent entity. It is necessary to exclude an underlying systemic disease to establish correct systemic treatment.
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POS1358 THE EFFECTS AND SAFETY OF APREMILAST AND CYTOKINE EXPRESSION IN BEHCET'S DISEASE PATIENTS

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Background: Apremilast, the small-molecule phosphodiesterase (PDE) -4 inhibitor, was approved for the treatment of recurrent oral ulcers associated with Behcet's disease (BD) in Japan from September 2019, following the success of the phase 3 RELIEF study (1). However, the efficacy of apremilast on domains other than oral ulcers in BD patients is unclear. On the other hand, it has been reported that apremilast may decrease the production of proinflammatory cytokine and increase the production of anti-inflammatory mediators in psoriasis (PS) and psoriatic arthritis (PsA) (2).
Objectives: To evaluate the effects and safty of apremilast on clinical symptoms and the changing of serum cytokine expression.
Methods: BD patients who had treated with apremilast for active oral ulcers were included in the study. We investigated the improvement rate of oral and genital ulcers, skin lesions, arthritis. In addition, serum cytokines (IFN-γ, IL-10, IL-8, and TNF-α) before and after apremilast treatment were measured using a multiplex immunoassay (Luminex Assay, R&D Systems).
Results: Fourteen patients (3 males and 11 females) were enrolled in this study. The mean age was 46.6 ± 13.0 years and the mean duration of disease was 10.2 ± 8.8 years. All patients had active oral ulcers, five had genital ulcers, six had skin lesions, and four had arthritis. Three months after the treatment with apremilast, oral ulcers improved in 13 patients (92.9%). The improvement rates of genital ulcers, skin lesions and arthritis were 60%, 25% and 25%, respectively. Changes in serum cytokines were different from those previously reported in PS. Adverse events were gastrointestinal symptoms such as nausea and diarrhea in 6 patients and sensorineural deafness in 1 patient. Medication was reduced in 2 patients, and discontinued in 1 patient due to nausea and diarrhea.
Conclusion: Apremilast is useful not only for oral ulcers, but also for other lesions in BD patients. The effect of apremilast for other domain such as genital ulcers, skin lesions, arthritis was not comparable to that of active oral ulcers. Additionally, BD may have different cytokine profile from PS and PsA.
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POS1359 REGIONAL DIFFERENCES IN DISEASE CHARACTERISTICS OF FAMILIAL MEDITERRANEAN FEVER IN TURKEY: PRELIMINARY REPORT

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Background: Familial Mediterranean fever (FMF) is a hereditary autoinflammatory disease accompanied by recurrent attacks of fever and serositis. It is prevalent among Mediterranean populations, mainly Turks, Armenians; Jews and Arabs. As genetic factors are variable in the population, environmental factors can also affect phenotypic characteristics.
Objectives: This study aimed to determine the geographic differences in disease characteristics and burden in patients with FMF in Turkey.
Methods: Patients diagnosed with FMF according to the Tel-Hashomer criteria were included in this multi-center study. Patients were included from the different regions of Turkey. Demographic features and clinic characteristics of the patients including disease duration, medications, comorbid conditions, attack characteristics, amyloidosis, acute phase reactants, FMF gene mutations, arthritis, sacroiliitis, and febrile myalgia were recorded. PRASS disease activity score, FMF-QoL, HAD, and HAQ were assessed. Patients from different parts of Turkey were divided into 3 groups the Central Anatolia, Western, and Eastern. Disease activity, characteristics and burden were also investigated among 3 distinct geographic regions.
Results: A total of 281 patients with FMF (195 women, 86 men) were enrolled in this study. The mean age of the patients was 34.9 (SD:12.3) years. While the patients in the eastern areas of Turkey were diagnosed earlier age (*p<0.001*), the patients in the western area had a longer diagnostic delay time (*p<0.001*). Patients enrolled from western regions tended to have higher ESR and PRASS scores than those from eastern and central Anatolian regions, but attack numbers per 6 months were similar among the regions. The highest proportion of patients who were M694V/M694V homozygous patients were in western, and then eastern and central Anatolia (19.5%, 18%, and 5.4%). While fever and arthritis were more common in the eastern, pleuritis and sacroiliitis were more common in the central anatolia. Peritonitis and erysipelas like erythema rates were similar among the regions. The majority of patients were receiving colchicine treatment in all three regions. FMF-QoL scores were highest in the eastern and lowest in the western (*p=0.006*). Patients enrolled in the central Anatolia region experienced more functional disability than those from the western and eastern regions (p=0.009). Anxiety and depression scores were similar between groups (p=0.385 vs p=0.549).
Conclusion: These findings suggest that patients with FMF have diversity concerning the age at diagnosis, diagnostic delay time, disease activity, quality of

life, and functional disability among the 3 regions. In the genotypic analysis, the M694V mutation is the most common pathogenic mutation in all regions. Regional genetic and environmental varieties may explain the areal differences. These relationships can occur more clearly in larger patient populations.

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Table 1. Clinical and demographic characteristics of patients by geographic region

	Western, n=107	Central, n=74	Eastern, n=100	p
Age	37.7 (12.5)	36 (12.6)	31.2 (10.9)	**<0.001**
Sex, female	79 (73.8%)	58 (78.4%)	58 (%58)	**0.007**
Age at the symptom onset, year	16.8 (11.3)	20.4 (13.2)	18.5 (11)	0.229
Age at the diagnosis, year	29.6 (13.4)	27.7 (13.3)	22.1 (12.1)	**<0.001**
Diagnostic delay time, year	12.3 (12.9)	7.3 (10.5)	3.6 (5.3)	**<0.001**
ESR, mm/hr	26.8 (18.9)	15.5 (12)	13.1 (13.1)	**0.003**
PRASS		41 (55.4%)		0.577
Low	50 (46.7%)	28 (37.8%)	51 (51%)	
Moderate	48 (44.9%)		45 (45%)	
High	9 (8.4%)	5 (6.8%)	4 (4%)	
Fever	83 (77.6%)	60 (81.1%)	96 (96%)	**0.001**
Peritonitis	98 (91.6%)	64 (86.55)	92 (92%)	0.412
Pleuritis	35 (32.7%)	27 (36.5%)	18 (18%)	**0.013**
Arthritis	18 (16.8)	16 (21.6)	36 (36%)	**0.005**
Sacroiliitis	8 (7.5%)	14 (18.9%)	1 (1%)	**0.000**
Erysipelas like erythema	2 (1.9%)	7 (9.5%)	7 (7%)	0.075

Data are presented as mean (SD), median (min-max), and n (%)

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POS1360 MONOGENIC AUTO-INFLAMMATORY DISEASES IN ADULTS IN THE PRACTICE OF A RHEUMATOLOGIST

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Background: The group of the most common monogenic auto-inflammatory diseases (mAIDs) includes FMF, TRAPS, HIDS, CAPS. Descriptions of these diseases are widely presented in the literature, but they are not well covered among adults. MAIDs in the adult rheumatological practice are found: 1) in pts with the onset and diagnostics of the disease in their childhood; 2) in pts with juvenile onset, but diagnostics of the disease in their adulthood; 3) in pts with the onset of the disease in adulthood. Adult specialists, in contrast to pediatricians, are not very well aware of the clinical and laboratory picture, course, age characteristics, approaches to treatment, consequences of mAID, which leads to late diagnostics and untimely prescription of targeted therapy, development of complications (amyloidosis) and damage to organs, life quality impairment.

Objectives: To characterize adult patients with mAIDs in the practice of a rheumatologist.

Methods: Within the period from 2009 to 2020, 123 patients with mAIDs (FMF-53, CAPS-44, TRAPS-21, HIDS-5) were diagnosed, of which 50 were adults. All pts underwent a standard rheumatological examination, including ESR, CRP, ophthalmological and other instrumental examinations, audiogram, if required. Molecular genetic analysis of genes MEFV, TNFRSF1A, MVK, NLRP3 was performed in all pts. All but one patient with CAPS (CINCA / NOMID) showed feature genetic mutations.

Results: The study included 50 pts aged 18 to 66 years, 35 women, 15 men. The age of onset was from 0 to 53 years, in 88% of cases the onset of the disease was noted in childhood, 6 pts debuted at the age over 18 years (4 with FMF - 23-35 years; 2 CAPS - 51 and 53 years). As of the time of inclusion into the study, the duration of the disease ranged from 6 months to 59 years. All patients had fever, skin rashes were present in the majority (72%) of pts with CAPS (urticaria), TRAPS (erythematous and anulyariform) and HIDS (spotted), in half of pts with FMF (erysipeloid erythema). Articular manifestations were present in the majority of pts (80%), oligoarthritis in 38%, mainly in pts with CAPS (35%) and FMF (47.8%). Polyarthritis occurred in 18% of pts. Ophthalmic changes (32%) in the form of conjunctivitis and/or uveitis were more common in patients with CAPS (65%), sensorineural deafness only in pts with CAPS (40%), stomatitis in 16% in pts, more often in case of CAPS (35%). Family history is aggravated in almost half of pts. All pts showed an increase in ESR and CRP performance. Colchicine treatment was used mainly in patients with FMF. Glucocorticoids (GC) and biological agents (IL-1 inhibitors) were more often received by pts with CAPS (55% and 60%).

Table 1. Clinical and demographic characteristics of adult patients with mAIDs

AIDs	FMF (23)	CAPS (20)	TRAPS (5)	HIDS (2)	N (%)
Fever	23(100)	20 (100)	5 (100)	2 (100)	50 (100)
Rash	11(47,8)	18 (90)	5 (100)	2 (100)	36 (72)
Joints	17 (73,9)	16 (80)	5 (100)	2 (100)	40(80)
eye symptoms	1(4,3)	13 (65)	2 (40)	-	16(32)
Sensorineural hearing loss	-	8 (40)	-	-	
Family history	11(47,8)	8 (40)	3 (60)	-	22 (44)
Colchicine	23	1	2	-	26 (52)
Steroids	3	11	3	2	19 (38)
Biological therapy	5	12	3	1	21 (42)
Etanercept	2	-	1	-	3 (6)
Adalimumab	1	1	-	-	2 (4)
Tocilizumab	1	2	-	1	4 (8)
Canakinumab	1	7	2	-	10 (20)
Anakinra	1	5	-	-	6 (12)

Conclusion: The study of the peculiarities of the course of mAIDs in adults, increase in the awareness of adult rheumatologists about these diseases will improve and speed up diagnostics, develop approaches to treatment and management algorithms in order to improve the prognosis and quality of patients' life.

Disclosure of Interests: None declared

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POS1361 EVALUATION OF PATIENTS WITH (SUSPECTED) BEHÇET SYNDROME; DIAGNOSTIC VALUE OF PATHERGY TESTING AND OPHTHALMOLOGIC EXAMINATION

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Background: Behçet syndrome (BS) is a multisystem vasculitis of unknown etiology. A positive pathergy test (PT) is part of the ISG criteria for the classification of BS [1]. The added value of PT in the diagnostic workup is unknown. Also, it is unknown whether real time assessment of the pathergy reaction after 48 hours by a physician can be replaced by patient assessment.

The diagnostic value of routine eye examination in patients with suspected BS is unclear.

Objectives: Assess the diagnostic value of PT and ophthalmologic examination in (suspected) BS patients. Examine the correlation between physician and patient assessment of PT results.

Methods: This prospective cohort study in (suspected) BS patients was conducted from 2009 to 2020.

At baseline, patients were classified as "true" (≥3 criteria), "probable" (2 criteria) or "no" (0-1 criteria) BS according to ISG criteria and were referred to a dermatologist and/or ophthalmologist for PT and/or eye examination. The percentage of positive PT and eye examination in each group was assessed and the percentage of patients with a changed classification (probable to true BS) was determined.

PT results were scored by both patients and physicians.

Results: Baseline characteristics are reported in Table 1. Figure 1 displays the flowchart of diagnostic value of PT.

Table 1. Baseline characteristics of (suspected) BS patients.

	All patients n=153	ISG +[1] n=59	ISG+ after FU[2] n=11	ISG –[3] n=83	P-value (+ vs -)
Age (mean ± SD[4] years)	39.5 (12)	41.8 (11.6)	33.6 (7.5)	38.5 (12.6)	0.294
Male n (%)	47 (30.7)	16 (27.1)	4 (36.4)	27 (32.5)	0.360
HLA-B51+ n/n (%)	8/18 (44.4)	5/7 (71.4)	1/3 (33.3)	2/8 (25)	0.070
Ethnicity: endemic[5] n (%)	82 (53.6)	38 (64.4)	6 (54.5)	38 (45.8)	0.114
Oral aphthae n (%)	145 (94.8)	59 (100)	11 (100)	75 (90.4)	**0.008**
Genital ulcers n (%)	82 (53.6)	50 (84.7)	8 (72.7)	24 (28.9)	**0.000**
Skin n (%)	71 (46.4)	51 (86.4)	1 (9.1)	19 (22.9)	**0.000**
Pathergy + n/n (%)	23/58 (39.7)	17/33 (51.5)	2/3 (66.7)	4/22 (18.1)	**0.000**
Eye n (%)	28 (18.3)	17 (28.8)	1 (9.1)	10 (12)	**0.035**
Superficial thrombophlebitis n (%)	15 (9.8)	11 (18.6)	1 (9.1)	3 (3.6)	**0.018**
Vascular n (%)	11 (7.2)	5 (8.5)	1 (9.1)	5 (6)	0.360
Venous n (%)	10 (6.5)	5 (8.5)	1 (9.1)	4 (4.8)	
Arterial n (%)	2 (1.3)	1 (1.7)	0	1 (1.2)	
Neurological n (%)	5 (3.3)	3 (5.2)	0	2 (2.4)	0.433
Arthritis n (%)	40 (26.1)	20 (33.9)	2 (18.2)	18 (21.7)	0.197
Gastro- intestinal n (%)	6 (3.9)	3 (5.1)	0	3 (3.6)	1.00
Epididymitis n (% of males)	5 (10.6)	5 (31.3)	0	0	**0.002**

1.patients fulfilling the ISG criteria at enrollment2.patients who fulfilled the ISG criteria during FU3.patients did not fulfill the ISG criteria after FU4.standard deviation5.Turkey, Asia, Middle and Far Eastern, Arabic countries and Northern Africa