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 (%)

Disclosure of Interests: None declared

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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 annulariform) 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