frequencies of inflammatory diseases in families of patient with FMF haven't been investigated.

Objectives: In this study, we have aimed to evaluate the comorbid disorders in a large cohort of families of patients with FMF.

Methods: Four hundred and ninety-eight children with FMF, one hundred and forty patients with JIA and ninety-two healthy children were interviewed between December 2019 and January 2020. In JIA group and healthy control group, patients who have family history for FMF were excluded from the study. Patients were asked about characteristics of their disease attacks and if there is a relative with any inflammatory diseases who does not have FMF in patient's 1st and 2nd degree relatives.

Results: Demographic features of study group have shown in Table 1. The most common MEFV mutations in patients with FMF were: M694V homozygotes (13.2 %), M694V heterozygotes (12 %), M694V homozygotes and R202Q homozygotes (6,8 %). Type II diabetes, asthma and hypothyroidism were the most commonly detected diseases in all cohorts. Frequency of Behçet's disease, allergic rhinitis and type II diabetes were significantly higher in families of patients with FMF than other groups (p<0.05) (Table 2).

Table 1. Demographic features of study population.

	FMF†	JIA††	Healthy Control
	n: 498 (%) mean +/- SD	n: 140 (%) mean +/- SD	n: 92 (%) mean +/- SD
Female	284 (57)	91 (65)	55 (59.8)
Age (years)	12.9 ± 8.2	11.7 ± 5.1	7.4 ± 4.6
Age at Onset (years)	4.3 ± 3.3	5.4 ± 4.1	-
Age at Diagnosis (years)	6.3 ± 3.6	6.3 ± 4.5	-
Delay in Diagnosis (months)	23.8 ± 29.2	11.3 ± 28.2	-
Follow-up Duration (years)	6.9 ± 8.3	5.3 ± 4.0	-
Consanguinity	100 (20)	25 (17.8)	8 (8.6)
Family History of FMF	282 (56.6)	0 (0)	0 (0)
JIA subgroup	-	-	-
Oligoarticular	-	72 (51.4)	-
Polyarticular (RF negative)	-	16 (11.4)	-
Polyarticular (RF positive)	-	3 (2.1)	-
Enthesitis Related Arthritis	-	14 (10)	-
Psoriatic Arthritis	-	7 (5)	-
Systemic	-	23 (16.4)	-
Other	-	5 (3.5)	-
Clinical Findings	-	-	-
Fever	392 (78.1)	-	-
Abdominal Pain	429 (86.1)	-	-
Chest Pain	102 (20.5)	-	-
Arthralgia	334 (67.1)	-	-
Arthritis	157 (31.5)	-	-
Extremity Pain	64 (12.8)	-	-
Heel Pain	44 (8.8)	-	-
Myalgia	43 (8.6)	-	-
*ELE	13(2.6)	-	-
Serositis	10 (2)	-	-

†Familial Mediterranean Fever ††Juvenile Idiopathic Arthritis *Erysipelas like erythema

Table 2. Comparison of frequencies of diseases detected among families of patient groups (shortened).

Diseases	FMF	JIA	Healthy Control	p ¹
Type II Diabetes	284 (57)	64(45.7)	44 (47.8)	0.02
Asthma	139 (27.9)	30 (21.4)	20 (21.7)	0.19
Hypothyroidism	122 (24.4)	27 (19.2)	14 (15.2)	0.09
Eczema	68 (13.6)	14 (10)	5 (5.4)	0.06
Psoriasis	49 (9.8)	6 (4.2)	7 (7.6)	0.10
Allergic Rhinitis	49 (9.8)	3 (2.1)	1 (1)	0.001
Hyperthyroidism	40 (8)	9 (6.4)	3 (3.2)	0.24
Behçet's Disease	31 (6.2)	1 (1)	2 (2.1)	0.01
Rheumatic Fever	30 (6)	10 (7.1)	2 (2.1)	0.25

Conclusion: In this study, we have reported increased frequencies of Behçet's disease, allergic rhinitis and type II diabetes in families of patients with FMF. Our results suggest that possible increased mutation load among families of patients with FMF may cause increased inflammatory diseases.

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FRI0456

EARLY IMPLEMENTATION OF TREATMENT WITH ETANERCEPT INCREASES THE LIKELIHOOD TO ACHIEVE REMISSION

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic inflammatory rheumatic disease in children and adolescents. A consistent therapy is required to avoid consequential damage and permanent loss of function. Biologic disease modifying anti-rheumatic drugs (bDMARDs) provide a well-accepted option for treatment of patients with a severe course of JIA. Etanercept (ETA) is still the most commonly prescribed bDMARD for JIA in Germany.

Objectives: To analyze adherence to treatment with ETA with special attention on discontinuation after achieving an inactive disease and recurrence of active disease after ETA withdrawal.

Methods: Data from two ongoing prospective, multicenter, non-interventional registries BiKeR and JuMBO were used for the analysis. JuMBO is the follow-up study to BiKeR and follows patients who have reached the age of 18. Both registers provide treatment data, individual trajectories of clinical data and outcomes from childhood into adulthood in JIA patients treated with bDMARDs and csDMARDs. Clinical disease characteristics, such as disease activity, were reported by the rheumatologists in addition to patient-reported outcomes at each six-months follow-up. Remission was defined as inactive disease defined by the Wallace Criteria (1).

Results: Data from 2,500 patients who were included in BiKeR and had an age ≥18 at the time of analysis were considered. A subset of 1,535 were enrolled in JuMBO. The mean follow-up was 8.6 (SD 4.2) years for the JuMBO patients. The majority of them had polyarthritis (35%), followed by enthesitis-related arthritis (20%). A total of 1,779 (68.8% of 2,584) patients were ever treated with ETA, providing 2,178 ETA treatment courses. There were 1,724 (67%) patients with first, 338 patients with a second and 54 with a third course of ETA treatment course. 710 (41.2%) discontinued ETA by ineffectiveness in the first course with similar rates of discontinuation due to ineffectiveness in the first and second course. A total of 332 (+/-MTX, 19.3%) discontinued ETA after achieving remission in the first ETA course. Among those, 129 (38.9%) patients did not require treatment with any other bDMARD subsequently until last follow-up (3.9 years, SD 3.5), while 169 (50.9%) re-started treatment with ETA, 14 (4.2%) with adalimumab and 4 with other bDMARDs. The likelihood of discontinuing ETA due to an inactive disease was positively associated with a younger age (hazard ratio (HR) 1.08, p<0.001), persistent oligoarthritis (HR 1.89, p=0.004), a shorter duration between JIA onset and ETA start (HR 1.10, p<0.001) as well as a good response to therapy within the first six months of treatment (HR 1.11, p<0.001). 209 (of 332) had ETA monotherapy at withdrawal. Of those, 77% (n=161) experienced recurrence of disease with a mean time to flare of 12.1 (SD 13.7) months. 129 patients restarted bDMARD therapy (n=117 ETA). We could not identify any correlates for the risk of flare. 70% re-achieved remission and 20% again discontinued therapy thereafter.

Conclusion: The study confirms the good effectiveness of ETA, even in the re-treatment of patients with JIA. Our data highlight the association of an early bDMARD treatment with a higher likelihood to achieve an inactive disease indicating a window of opportunity.

References:

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FRI0457

LONG-TERM OUTCOMES AND TREATMENT EFFICACY IN PATIENTS WITH TNF RECEPTOR-ASSOCIATED AUTOINFLAMMATORY SYNDROME (TRAPS) FROM THE EUROFEVER INTERNATIONAL REGISTRY

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