Results: We determined a higher level of homocysteine in patients with AN (12,15 (7,55-14,60) vs. 8,18 (6,57-10,70) $\mu\text{mol/l}$ in the control group, $p=0,04$). Also we observed almost equal levels of vitamin B12 in patients with AN in the control group (531,95 (340,80-793,55) vs. 532,75 (417,60-669,7) pg/ml , $p=0,99$). The level of folate was lower in the study group (5,66 (3,83-6,83) vs. 6,99 (5,02-10,38) ng/ml , $p=0,01$).

Conclusion: The data obtained show that homocysteine level in patients with AN is higher. This can be a consequence of folate deficiency, which takes part in homocysteine metabolism. Therefore, homocysteine metabolism disturbances can lead to endothelial dysfunction, which results in cardiovascular events in the heart, brain, kidneys [1] as well as in the bone, according to the study results. Folate supplement is the possible way of AN treatment and prevention.

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POS1378

COMPARISON OF DEMOGRAPHIC AND CLINICAL FEATURES OF FAMILIAL MEDITERRANEAN FEVER PATIENTS AND PATIENTS WITH AXIAL SPONDYLOARTHRITIS ACCOMPANYING FAMILIAL MEDITERRANEAN FEVER

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Background: The rate of co-occurrence of Familial Mediterranean Fever (FMF) and axial spondyloarthritis (axSpA) in adults is reported ranging from 0.5% to 7.5%. The clinical implications of this co-occurrence in the course of FMF is still a research question.

Objectives: To compare of demographic and clinical features of patients with FMF and FMF+axSpA.

Methods: A total of 9630 FMF patients was detected according to the ICD-10 code (E85.0) of FMF in Hacettepe University Hospital database. 241 of these patients also had axSpA according to the ICD-10 code (M45). FMF diagnosis was confirmed by Tel-Hashomer criteria. AxSpA was diagnosis was confirmed by either presence of sacroiliitis on sacroiliac radiography according to the Modified New York Criteria (mNY) or presence of active sacroiliitis according to ASAS criteria on magnetic resonance imaging. 136 patients were confirmed according to these criterias as having FMF+axSpA. As a control group, 231 consequent FMF patients without axSpA recorded on the "FMF in Central Anatolia (FICA) database" and followed up at our center were included in the analysis. Demographic and clinical features of those patients in both groups were compared. $p<0.05$ was considered as statistically significant, correction for multiple comparisons was not performed.

Results: 136 patients were included in FMF+axSpA group and 231 patients were included in FMF group. 114 (83.8%) patients in FMF+axSpA group had radiographic sacroiliitis according to mNY criteria; median cervical mSASSS was 0 (available for 49 patients, min-max, 0-36), median lumbar mSASSS was 4 (available for 121 patients, min-max, 0-36), 33 (27%) patients had cervical or lumbar syndesmophyte. Twenty-six (19.1%) of these patients had radiologically documented inflammatory hip disease 12 (8.8%) of these patients underwent total hip replacement. Female gender was more prevalent in FMF+axSpA group (53.7% vs 32.5%, $p<0.001$). Age at FMF symptom onset and diagnosis were earlier in FMF patients; however, symptom and disease durations were longer in FMF+axSpA group in our study cohort (Table 1). Frequency of FMF signs and symptoms were comparable except the rate of pleuritis was higher in FMF patients compared to FMF+axSpA group ($p=0.003$). Amyloidosis was more prevalent in FMF+axSpA group (6.6% vs. 1.7%, $p=0.014$). Results of MEFV gene analysis were available for 273 patients. Although presence of M694V mutation (either in 1 allele or 2 alleles) was comparable in 2 groups, homozygous M694V mutation was more prevalent in FMF+axSpA group (39.8% vs. 28.9%, $p=0.02$).

Conclusion: The coexistence of spondyloarthritis in FMF patients appears to be associated with the increased prevalence of amyloidosis. The inflammatory

burden of a second disease and the increased prevalence of the homozygous M694V mutation may explain this risk.

Table 1. Comparison of demographic and clinical features of two groups.

	FMF+AxSpA (n=136, 37.1%)	FMF (n=231, 62.9%)	p
Female, n(%)	73 (53.7)	75 (32.5)	<0.001
Age at FMF symptom onset, years med (IQR)	12 (5-20)	10 (6-18)	0.046
Symptom duration, years, med (IQR)	24 (18-32)	20 (14-29)	0.007
Age at FMF diagnosis, years, med (IQR)	24 (13-33)	20 (11-30)	0.10
Duration after diagnosis, years, med (IQR)	16 (10-22)	13 (7-17)	<0.001
FMF signs and symptoms, n(%)			
-Fever	128 (94.1)	204 (88.3)	0.067
-Abdominal pain	123 (90.4)	217 (93.9)	0.21
-Pleuritis	31 (22.8)	87 (37.7)	0.003
-Pericarditis	3 (2.2)	2 (1.0)	0.34
-Arthritis	64 (47.1)	92 (39.8)	0.17
-Erysipelas	24 (17.6)	38 (16.5)	0.77
-Febrile myalgia	9 (6.6)	13 (5.6)	0.70
Inflammatory back pain, n(%)	92 (67.6)	26 (11.3)	<0.001
Inflammatory bowel disease, n(%)	6 (4.4)	4 (1.7)	0.12
FMF family history, n(%)			
-Any degree	66 (48.5)	137 (59.8)	0.04
-First degree	48 (35.8)	97 (42.0)	0.24
-Second degree	25 (18.7)	86 (37.2)	<0.001
Number of attacks at recent year, med (min-max)	1 (0-12)	1 (0-10)	0.13
Amyloidosis	9 (6.6)	4 (1.7)	0.014
M694V status (N=273)			
-Present (one or two alleles)	91 (80.5)	120 (75.0)	0.28
-Two alleles	45 (39.8)	43 (28.9)	0.02

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POS1379

LONG-TERM EFFICACY AND SAFETY OF CANAKINUMAB IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER (FMF) - INTERIM ANALYSIS OF THE RELIANCE REGISTRY

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Background: Familial Mediterranean Fever (FMF) is characterized by recurrent attacks of fever and serositis as well as elevated inflammatory markers. FMF treatment goals according to EULAR are to control acute attacks and subclinical inflammation and to improve patients' quality of life¹. In a phase 3 pivotal study (CLUSTER study), FMF patients treated with the interleukin-1 β inhibitor canakinumab met all these goals².

Objectives: The present study explores the long-term efficacy and safety of canakinumab (CAN) in routine clinical practice in pediatric (age ≥ 2 years) and adult FMF patients.

Methods: RELIANCE is a prospective, non-interventional, multi-center, observational study based in Germany with a three-year follow-up period. Patients with clinically confirmed diagnosis of FMF who routinely receive CAN were enrolled in order to evaluate effectiveness and safety of CAN under standard clinical practice conditions. Disease activity and remission by physicians' assessment,

Table 1. Baseline characteristics and third interim analysis data of patients with FMF

	Baseline		6 months		12 months		18 months	
All patients patients without prior CAN therapy								
Number of patients, N	54	11	35	7	27	5	16	3
Number (%) of patients in disease remission (physician assessment)	18 (48.6)	1 (20.0)	19 (73.1)	3 (75.0)	13 (65.0)	1 (50.0)	8 (61.5)	1 (100.0)
Physician Global Assessment, percentage* of absent/mild-moderate/severe rating	43/38/11	0/40/60	65/27/0	50/50/0	55/35/0	0/50/0	46/46/0	0/100/0
Patient assessment of current disease activity; 0–10, median (min; max)	3.0 (0; 10)	7.0 (0; 10)	2.5 (0; 7)	2.0 (0; 5)	2.0 (0; 7)	2.0 (0; 2)	2.0 (0; 6)	0.5 (0; 1)
Patient assessment of current fatigue; 0–10, median (min; max)	5.0 (0; 10)	5.0 (0; 9)	3.5 (0; 10)	3.0 (1; 6)	3.0 (0; 10)	0.0 (0; 4)	3.0 (0; 7)	0.5 (0; 1)
Number (%) of patients without impairment of social life by the disease	19 (46.3)	3 (37.5)	18 (66.7)	3 (75.0)	14 (66.7)	4 (80.0)	5 (55.6)	2 (66.7)
CRP, median (mg/dl)	0.2	1.1	0.2	0.1	0.2	0.0	0.1	0.5
SAA, median (mg/dl)	0.7	6.8	0.8	0.4	0.8	0.6	0.6	0.7
ESR, median (mm/h)	9.0	18.5	6.0	5.0	5.5	4.0	8.0	5.0
SAE	Number of events		Incidence rate per 100 patient years					
Total	11		16.23					
Arthritis	5		7.38					
SARS-CoV-2 infection	2		2.95					
Familial Mediterranean Fever	1		1.48					
Intestinal Hemorrhage	1		1.48					
Pyrexia	1		1.48					
Tonsillectomy (SADR)	1		1.48					

*not reported for all patients

CRP, c-reactive protein; ESR, erythrocyte sedimentation rate; n. a., not annotated; SAA, serum amyloid A; SADR, serious adverse drug reaction; SAE, serious adverse event

disease activity, fatigue and impact on social life by patients' assessment, inflammatory markers and AIDAI (Auto-Inflammatory Diseases Activity Index) score were recorded at baseline and were assessed at 6-monthly intervals within the three-year observation period of the study.

Results: This interim analysis of FMF patients (N=54) enrolled by December 2020 includes baseline as well as 6-, 12- and 18-month data. Mean age in this cohort was 25 years (4–56 years) and the proportion of female patients was 46 % (N=25). At baseline, median duration of prior CAN treatment was 2.0 years (0–6 years).

While physician ratings report around 62% of patients in disease remission, 52% with absent and 34% with mild-moderate disease activity, patient-reported disease activity decreased from moderate (PPA 3.0) to low (PPA 2.0) during the observation period. A decrease was observed regarding disease activity parameters, in particular in patients without prior CAN therapy (Table 1, Figure 1). A total of 11 serious adverse events was reported, of which one case of tonsillectomy was classified as drug-related.

Conclusion: Interim data of FMF patients from the RELIANCE study, the long-term running real-life CAN registry, confirm efficacy and safety of long-term CAN treatment.

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Figure 1: Median AIDAI score (0–390) of the FMF main group (N = 54, at baseline) and the FMF subgroup without prior CAN therapy (N=11) within 18 months

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POS1380

LONG-TERM EFFICACY AND SAFETY OF CANAKINUMAB IN PATIENTS WITH TRAPS (TUMOR NECROSIS FACTOR RECEPTOR-ASSOCIATED PERIODIC SYNDROME) - INTERIM ANALYSIS OF THE RELIANCE REGISTRY

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Background: Tumor necrosis factor receptor-associated periodic syndrome (TRAPS) is a rare autoinflammatory condition characterized by severe systemic and organ inflammation. In a phase 3 pivotal trial (CLUSTER study), TRAPS patients have been successfully treated with the interleukin-1 β inhibitor canakinumab. 45% of patients reached clinical remission after 16 weeks (primary endpoint)¹. Canakinumab has been approved and applied for the treatment of TRAPS patients since 2017².

Objectives: The present study explores the long-term efficacy and safety of canakinumab under routine clinical practice conditions in pediatric (age ≥ 2 years) and adult TRAPS patients.