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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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.

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

	Baseline	6 months	12 months	18 months
Number of patients, N	16	13	10	6
Median age, years (min; max)	23 (3; 43)	17 (3; 43)	16 (4; 38)	25 (4; 43)
Females (%)	11 (69)	9 (69)	7 (70)	3 (50)
Median duration of prior CAN therapy at baseline, years (min; max)	1.0 (0; 4)	1.0 (0; 4)	1.0 (0; 4)	1.5 (0; 2)
Number (%) of patients in disease remission (physician assessment)	9 (60.0)	9 (81.8)	7 (77.8)	4 (80.0)
Physician Global Assessment, percentage of absent/mild-moderate/severe rating	40 / 53 / 0	82 / 9 / 0	44 / 44 / 11	80 / 20 / 0
Patient assessment of current disease activity; 0–10, median (min; max)	1.5 (0; 5)	1.0 (0; 4)	1.0 (0; 6)	0.0 (0; 3)
Patient assessment of current fatigue; 0–10, median (min; max)	2.0 (0; 8)	1.0 (0; 7)	2.5 (0; 8)	4.0 (0; 7)
Number (%) of patients without impairment of social life by the disease	4 (50)	5 (63)	2 (33)	3 (60)
Number (%) of patients with days absent from work/school during last 6 months	8 (50)	5 (39)	5 (56)	3 (50)
CRP, median (mg/dl)	0.1	0.1	0.1	0.0
SAA, median (mg/dl)	0.5	0.4	0.4	0.3
ESR, median (mm/h)	7.0	5.0	5.0	5.0
SAE	Number of events		Incidence rate per 100 patient years	
Total	3		14.7	
Circulatory collapse (non fatal)	1		4.9	
Dizziness	1		4.9	
Headache	1		4.9	

CRP, c-reactive protein; **ESR**, erythrocyte sedimentation rate; SAA, serum amyloid A; SAE, serious adverse event

Methods: RELIANCE is a prospective, non-interventional, multi-center, observational study based in Germany with a 3-year follow-up period. Patients with clinically confirmed diagnoses of TRAPS, CAPS, FMF or HIDS/MKD who routinely receive canakinumab are enrolled in order to evaluate efficacy and safety of canakinumab under standard clinical practice conditions. Disease activity and remission by physician assessment, disease activity and fatigue by patient assessment, days absent from school/work due to study indication, inflammatory markers, and AIDAI (Auto-Inflammatory Diseases Activity Index) score were assessed at baseline and at 6-monthly intervals.

Results: The interim analysis of TRAPS patients enrolled by December 2020 includes baseline (N=16, including 1 patient with atypical TRAPS) and preliminary 18-month data. Mean age in this cohort was 23 years (3–43 years) and the median duration of prior CAN treatment was 1.0 year (0–4 years).

Physician assessment indicated 60–80% remission and laboratory parameters were within normal range. Disease control by patient assessment showed no major changes regarding the analyzed parameters (Table 1, Figure 1). Of the three serious adverse events reported none was classified as drug-related.

Conclusion: Preliminary analysis of 18 month interim data of TRAPS patients treated with CAN available from the RELIANCE study indicate stable efficacy and safety of CAN long-term treatment.

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Figure 1: Median AIDAI score (0-390) of the TRAPS patients (N = 16 at baseline) within 18 months

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