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LONG-TERM EFFICACY AND SAFETY OF CANAKINUMAB IN PATIENTS WITH AUTOINFLAMMATORY PERIODIC FEVER SYNDROMES – FIRST INTERIM ANALYSIS OF THE FMF/TRAPS/HIDS SUBGROUPS FROM THE RELIANCE REGISTRY

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Background: Autoinflammatory periodic fever syndromes characterized by excessive interleukin(IL)-1b release and severe systemic and organ inflammation have been successfully treated with the anti-IL-1 inhibitor canakinumab (CAN). In clinical trial situations and real life, rapid remission of symptoms and normalization of laboratory parameters were observed in most patients.¹⁻³

Objectives: The present study explores long-term effectiveness and safety of CAN under routine clinical practice conditions in pediatric and adult patients with CAPS (cryopyrin-associated periodic syndromes), FMF (familial Mediterranean fever), TRAPS (tumor necrosis factor receptor-associated periodic syndrome) and HIDS/MKD (hyperimmunoglobulinemia D syndrome/mevalonate kinase deficiency).

Methods: RELIANCE is a prospective, non-interventional, multi-center, observational study based in Germany with a 3-year follow-up period. Pediatric (age ≥2 years) and adult patients with clinically confirmed diagnoses of CAPS, FMF, TRAPS and HIDS/MKD that routinely receive CAN are enrolled in order to evaluate effectiveness and safety of CAN in clinical routine.

Results: This first interim analysis of patient subgroups diagnosed with FMF, HIDS and TRAPS includes baseline data of 41 patients (10 TRAPS, 2 HIDS, 29 FMF), including preliminary 6-month data of a FMF-subset (N=16). Evaluation of disease activity and fatigue by patients' assessment, days absent from school/work, inflammatory markers and physician global assessment (PGA) was performed at baseline and will be further assessed at 6-monthly intervals within the 3-year observation period. Preliminary results of a first subset of 16 patients diagnosed with FMF are available and indicate stable remission and disease control upon long-term CAN treatment: within the first study interval, no major changes were observed regarding the analyzed parameters (table 1).

Conclusion: Baseline characteristics of all subgroups and first interim data of FMF patients are available from the RELIANCE study, the longest running real-life CAN registry. Further interval data will be analyzed to assess efficacy and safety of long-term CAN-treatment in patients with autoinflammatory periodic fever syndromes.

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APREMILAST IN MONOTHERAPY OR COMBINED IN NON-ULCER MANIFESTATIONS OF BEHÇET'S DISEASE. NATIONAL MULTICENTER STUDY OF 34 REFRACTORY CASES OF CLINICAL PRACTICE

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Background: Apremilast (APR) has demonstrated efficacy in orogenital ulcers of Behçet's disease (BD). Response of other clinical manifestations remains unknown.

Objectives: To assess the efficacy and safety of APR in monotherapy or combined with disease-modifying anti-rheumatic drugs (DMARDs) in non-aphthous ulcers of BD.

Methods: National multicenter open-label study on 34 BD patients treated with APR at maintained standard dose of 30 mg twice daily.

Results: From a cohort of 51 patients with APR by refractory orogenital ulcers of BD, we selected 34 (24 women/10 men, mean age 43.8±14.3 years), cases with another clinical manifestation/s.

Excluding CTs, colchicine or NSAIDs, APR was given in monotherapy (n=21) or combined with conventional and/or biologic DMARDs in 13 cases (5 methotrexate, 3 azathioprine, 3 hydroxychloroquine, 1 sulfasalazine, 1 dapsone, 2 tocilizumab, 1 IFX). Other active manifestations present at APR onset were: arthralgia/arthritis (16, true arthritis in 5), folliculitis/pseudofolliculitis (14), erythema nodosum (3), furunculosis (2), paradoxical psoriasis by TNFi (2), intestinal ileitis (2), deep venous thrombosis (2), leg ulcers (1),

Table 1. Baseline characteristics of the TRAPS/HIDS/FMF-subgroups and preliminary 6-month data of FMF subset.

	TRAPS			HIDS			FMF					
	Baseline			Baseline			Baseline all FMF patients			Baseline FMF subset	6 months FMF subset	
Number of patients, N	10			2			29			16	16	
Mean age, years (SD)	22 (4; 43)			11 (5; 18)			26 (5; 56)			16 (5; 47)	16 (5; 47)	
Female (%), N=9 (1 missing)	5 (56)			1 (50)			15 (52)			7 (44)	7 (44)	
Mean duration of prior canakinumab treatment, years (min; max)	1 (0; 2)			3 (2; 4)			n. a.			2.2 (0; 6)	2.2 (0; 6)	
Patient's assessment of disease activity 0-10, mean (min; max)	2.1 (0; 5)			0 (0; 0)			3 (0; 10)			2.8 (0; 8)	2.2 (0; 7)	
Patient's assessment of fatigue 0-10	3.4 (0; 8)			0 (0; 0)			4.4 (0; 9)			4.6 (0; 9)	3.9 (0; 8)	
Number (%) of patients with days absent from school/work due to study indication during last 6 months	4 (44)			2 (100)			5 (17)			2 (13)	5 (31)	
Inflammatory markers, CRP/SAA, mean (mg/dL)	2.0	7.9		0.1	0.6		0.9	5.3	0.5	2.4	0.6	2.4
PGA of disease activity: no/mild to moderate/severe; N (%)*	1 (11)	6 (67)	0 (0)	2 (100)	0 (0)	0 (0)	10 (35)	8 (28)	5 (31)	7 (44)	6 (38)	7 (44)
SAE, N (%)	0 (0)			0 (0)			2 (6.9)	2 (7)	n. a.	1 (6)	2 (12.5)	0 (0)