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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)	2 (7)	5 (31)	7 (44)	1 (6)	6 (38)	7 (44)	0 (0)
SAE, N (%)	0 (0)			0 (0)			2 (6.9)			n. a.	2 (12.5)				

TABLE.

Non-apthous manifestations at APR onset (n)	1-2 Weeks		6 Months		12 Months		24 Months	
	C	M	C	M	C	M	C	M
Folliculitis/pseudofolliculitis (14)	n=5 CR (2) PR (1) NC (2)	n=9 CR (4) PR (2) NC (3)	n=4 CR (4)	n=4 CR (2) NC (1) ND (1)	n=1 CR (1)	n=2 ND (2)	n=0	n=0
Arthralgias (11)	n=5 NC (5)	n=6 CR (1) NC (5)	n=4 CR (3) NC (1)	n=3 PR (2) ND (1)	n=4 CR (2) PR (1) ND (1)	n=3 CR (1) PR (1) ND (1)	n=0	n=0
Arthritis (5)	n=3 PR (1) NC (2)	n=2 NC (2)	n=3 CR (3)	n=1 PR (1)	n=0	n=1 PR (1)	n=0	n=0
Erythema nodosum (3)	n=0	n=3 CR (2) PR (1)	n=0	n=1 CR (1)	n=0	n=0	n=0	n=0
Psoriasis/ erythematous-scaly skin lesions (3)	n=0	n=3 NC (2) ND (1)	n=0	n=2 PR (2)	n=0	n=2 PR (2)	n=0	n=1 PR (1)
Ileitis (2)	n=0	n=2 CR (1) PR (1)	n=0	n=2 CR (2)	n=0	n=1 CR (1)	n=0	n=0
Deep venous thrombosis (2)	n=1 CR (1)	n=1 PR (1)	n=1 CR (1)	n=1 PR (1)	n=1 CR (1)	n=0	n=0	n=0
Furunculosis (2)	n=1 CR (1)	n=0	n=1 CR (1)	n=0	n=0	n=0	n=0	n=0
Leg ulcers (1)	n=1 PR (1)	n=0	n=1 CR (1)	n=0	n=0	n=0	n=0	n=0
Unilateral anterior uveitis (1)	n=1 CR (1)	n=0	n=0	n=0	n=0	n=0	n=0	n=0
Neurobehçet (1)	n=0	n=1 NC (1)	n=0	n=1 NC (1)	n=0	n=0	n=0	n=0
Fever (1)	n=0	n=1 NC (1)	n=0	n=0	n=0	n=0	n=0	n=0

CR= complete remission; ND= non-data; NC= no changes; PR= partial remission

erythematous and scaly skin lesions (1), fever (1), unilateral anterior uveitis (1) and neurobehçet (1).

After a median follow-up of 6 [3-12] months, folliculitis and ileitis improved, neurobehçet remained stable and musculoskeletal manifestations evolved in a variable way. (TABLE)

Conclusion: In addition of orogenital ulcers, APR in monotherapy or combined, seems to be useful in skin manifestations of BD

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CLINICAL AND LABORATORY FEATURES OF PATIENTS WITH REMITTING SERONEGATIVE SYMMETRICAL SYNOVITIS WITH PITTING EDEMA COMPARED TO PATIENTS WITH SERONEGATIVE RHEUMATOID ARTHRITIS

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Background: In the case of seronegative arthritis, it was difficult to make a differential diagnosis between remitting seronegative symmetrical synovitis with pitting edema syndrome (RS3PE) and seronegative rheumatoid arthritis (seronegative RA) because the distribution of affected joints was similar and the patients with RS3PE or seronegative RA may have edema.

Objectives: To compare the clinical characteristics of RS3PE and seronegative RA
Methods: We retrospectively examine consecutive patients diagnosed with RS3PE or seronegative RA in our hospital from 2007 to 2019. Patients in whom both ACPA and RF were negative were included. The patients with RS3PE met the criteria of McCarty et al.: (1) pitting edema of the dorsum of both hands and both feet, (2) sudden onset of polyarthritis, (3) seronegative for ACPA and RF, (4) no radiologically evident erosions developed. The patients with seronegative RA met the EULAR/ACR 2010 criteria. The patients who were diagnosed with RS3PE at first and then diagnosed with seronegative RA afterward were included in seronegative RA group. The first analysis was performed on the affected joints, CRP, ESR, Hb, LDH, edema, the history of malignancy 2 years before and after the diagnosis, treatment, and the history of infection requiring hospitalization after the start of treatment. The affected joints were shoulders, elbows, wrists, finger joints (the MCP, and PIP joints), hips, knees, ankles, and toe joints (the MTP and PIP joints). The secondary analysis was performed on the above evaluations with a propensity score (PS) matching for age.

Results: In the first analysis, 20 patients with RS3PE and 122 patients with seronegative RA were enrolled. The mean ages (RS3PE, seronegative RA) were 81.1, 67.4 years old. Females were 60.0%, 63.1%. The mean observation period was 25.4, 63.6 months. The proportion of affected joints were shoulders