OP0091

TREATMENT OF OCULAR SARCOIDOSIS. STUDY OF 65 PATIENTS OF A SERIES OF 384 PATIENTS FROM A SINGLE UNIVERSITY HOSPITAL

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Background: Ocular involvement is a relatively frequent and potentially severe complication of sarcoidosis. Oral corticosteroids (OCS) are the first-line treatment. Conventional immunosuppressive agents (cIS) and biological therapy (BT) can be used in refractory cases (1-5).

Objectives: To evaluate the treatment and visual outcomes of a cohort of patients diagnosed with ocular sarcoidosis.

Methods: Study of a large cohort (n=384) of all consecutive patients diagnosed with sarcoidosis from January 1, 1999 to December 31, 2019 at a single University Hospital. Finally, 344 patients were included according the ATS/ERS/WASOG criteria (*Eur Respir J.* 1999; 14:735-7). Different ocular manifestations and the following systemic treatments were assessed: **a)** OCS, **b)** cIS, **c)** monoclonal TNF inhibitors, **d)** Etanercept (ETN), **e)** Tocilizumab (TCZ). Best Corrected Visual Acuity (BCVA) according to different systemic treatments was compared at diagnosis and after one year of follow-up (Kruskal Wallis test).

Results: 344 patients were reviewed. From these, 65 (18.9%) presented ocular manifestations as uveitis (83.1%), orbital lesions (7.7%), retinal vasculitis (6.2%), dry eye (6.2%) and scleritis (1.5%). All of them received systemic treatment. BT was particularly used in patients with retinal vasculitis (100%), panuveitis (75%) and orbital lesions (40%). Systemic treatment and BCVA outcome according to

Table 1. Median BCVA at onset and after one year according to ocular manifestations and type of systemic therapy.

Type of ocular affection	n (%)	Median BCVA at onset [IQR]	Median BCVA after 1 year [IQR]	OCS n (%)	cIS n (%)	monoclonal TNFi n (%)	ETN n (%)	TCZ n (%)
Uveitis	54 (83.1)	0.6 [0.3-0.8]	0.9 [0.6-1]	44 (81.5)	29 (53.7)	16 (29.6)	3 (5.5)	3 (5.5)
-Anterior	31 (47.7)	0.7 [0.3-0.8]	0.8 [0.5-1]	22 (70.9)	12 (38.7)	2 (6.5)	2 (6.5)	0
-Intermediate	2 (3.1)	0.5 [0.3-0.8]	0.7 [0.5-1]	2 (100)	1 (50)	1 (50)	1 (50)	1 (50)
-Posterior	5 (5.2)	0.5 [0.3-0.8]	0.9 [0.5-1]	4 (80)	4 (80)	3 (60)	0	0
-Panuveitis	16 (24.6)	0.4 [0.2-0.7]	0.9 [0.5-1]	16 (100)	12 (75)	10 (62.5)	0	2 (12.5)
Orbital lesions	5 (7.7)	0.5 [0.3-0.8]	1 [0.6-1]	4 (80)	2 (40)	2 (40)	0	1 (20)
Retinal vasculitis	4 (6.2)	0.6 [0.5-0.8]	1 [0.6-1]	4 (100)	4 (100)	1 (25)	0	1 (25)
Dry eye	4 (6.2)	1 [0.5-0.8]	0.9 [0.6-1]	2 (50)	1 (25)	0	0	0
Scleritis	1 (1.5)	1 [0.5-0.8]	1 [0.6-1]	1 (100)	0	0	0	0

Abbreviations: BCVA: Best Corrected Visual Acuity; IQR: Interquartile Range; OCS: Oral Corticosteroid; cIS: Conventional Immunosuppressants; Monoclonal TNFi: monoclonal tumour necrosis factor inhibitors; ETN: Etanercept; TCZ: Tocilizumab.

Figure 1. Median [25,75 IQR] BVCA after one year follow up according to type of systemic treatment in sarcoid uveitis. Median BCVA after one year Systemic treatment. **Abbreviations:** BCVA: Best Corrected Visual Acuity; OCS: Oral Corticosteroids; Conventional IS: Conventional immunosuppressants; Monoclonal TNFi: monoclonal tumour necrosis factor inhibitors; ETN: Etanercept; TCZ: Tocilizumab.

ocular manifestations are shown in table. Median BCVA at onset and after one year was 0.6 [interquartile range (IQR) 0.3-0.8] and 0.9 [0.6-1], respectively. No statistically significant differences were observed between systemic treatments in BCVA of patients with uveitis after 1 year of follow-up (Figure).

Conclusion: Panuveitis, intermediate uveitis and orbital lesions, require a more aggressive treatment than other manifestations of ocular sarcoidosis. In uveitis, an important improvement in BCVA after 1 year of follow-up was observed regardless of the type of treatment used.

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OP0092

LONG-TERM SAFETY AND EFFECTIVENESS OF CANAKINUMAB IN CRYOPYRIN-ASSOCIATED PERIODIC SYNDROMES (CAPS) – 30-MONTH DATA FROM THE RELIANCE REGISTRY

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Background: In clinical trials as well as in real-life, the IL-1β inhibitor canakinumab leads to rapid remission of symptoms in the treatment of CAPS, a monogenic autoinflammatory disease with severe systemic and organ inflammation.

Objectives: The RELIANCE registry is designed to explore long-term safety and effectiveness of canakinumab under routine clinical practice conditions in pediatric (≥2 years) and adult patients with CAPS, including Muckle-Wells syndrome (MWS), familial cold autoinflammatory syndrome (FCAS), and neonatal onset multisystem inflammatory disease (NOMID)/chronic infantile neurological cutaneous and articular syndrome (CINCA).

Methods: This prospective, non-interventional, observational study with a 3-year follow-up enrolls patients with clinically confirmed diagnoses of CAPS routinely receiving canakinumab. In 6-monthly visits, clinical data, physician assessments and patient-reported outcomes are evaluated starting at baseline with last update at 30 months of follow-up in the total cohort including the cohort with severe CAPS subtypes (NOMID/CINCA).

Results: 91 CAPS patients (50% female; 14 [15%] NOMID/CINCA subtypes) were enrolled by December 2020 (Table 1). At baseline, median age was 20.5 years and median duration of prior canakinumab treatment was 6 years. 11 drug related severe adverse events per 100 patient years were reported. 68% of patients