**SAT0239**

**PREDICTION OF LONG-TERM EVOLUTIONARY PROFILES IN EOSINOPHILIC GRANULOMATOSIS WITH POLYANIGIITIS (CHURG–STRAUSS) BASED ON BASELINE AND FOLLOW-UP CHARACTERISTICS**

Matthias Pago1, Renato A. Sinico2, Vitor Teixeira3, Maria-Letizia Urban4, Juliane Mahfield5, Sara Monti6, Giulia Cassone7, Franco Schiavon8, Benjamin Seeliger9, Thomas Neumann10, Claas Kroegel11, Matthieu Groh12, Chiara Marvisi13, Maxime Samson14, Thomas Barba15, David Jayne16, Bernhard Heilmich17, Carlomaurizio Montecucco18, Carlo Salvarani19, Jean-Emmanuel Kahn20, Bernard Bonnotte21, Claus Kroegel22, Matthieu Groh23, Chiara Marvisi24, Vítor Teixeira25, Maria-Letizia Urban26, Benjamin Terrier27, Matthieu Groh28, Chiara Marvisi29, Vítor Teixeira30, Maria-Letizia Urban31

**Background:** Eosinophilic granulomatosis with polyangiitis (EGPA) (Churg–Strauss) is a small-vessel necrotizing vasculitis characterized by blood and tissue eosinophilia and asthma. Glucocorticoids (GCs) control the disease, but GC-dependence is frequent and is an important distinction to make. EGPA seems to evolve toward distinct phenotypic profiles, especially in terms of eosinophilia and asthma. The Long-Term EGPA Outcomes (LTEO) Study aims to describe long-term EGPA outcomes, predicting for the first time the predictive factors using a large international database.

**Objectives:** The primary endpoints of the LTEO study are remission and GC-free remission. Remission is defined as absence of clinical activity (i.e., no glucocorticoids at last follow-up visit). GC-free remission is defined as remission with no GC treatment at last follow-up visit. Secondary endpoints include classification of long-term EGPA outcomes based on eosinophilic and asthmic activity.

**Methods:** Baseline and follow-up characteristics were included. EGPA patients were classified into distinct subgroups based on criteria defined by the unmet need for GCs and phenotyping of asthma, eosinophilia, and disease activity.

**Results:** A total of 1,188 EGPA patients were included. The median follow-up was 5.8 years (IQR 4.3–10). Baseline and follow-up characteristics predicting long-term EGPA outcomes were identified.

**Conclusion:** The LTEO study provides new insights into the long-term evolution of EGPA, predicting the risk of GC-dependence and the likelihood of asthma remission.


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**SAT0240**

**DEVELOPMENT AND PRELIMINARY VALIDATION OF THE BEHÇET’S SYNDROME OVERALL DAMAGE INDEX (BODI)**

Matteo Piga1, Alberto Floris1, Gerard Espinosa2, Nikolaos Koukgas3, Andrea Lo Monaco4, Giuseppe Lopalco5, Ida Orlandi6, Vittorio Pirani7, Emetestina Santos8, Luisa Serpa Pinto9, Diego Bertsiats, Luca Cantarini10, Alberto Cau11, Ricard Cervera12, João Correia13, Marcello Goffoni14, Florenzo Iannone15, Ana Martins Da Silva16, Piergiorgio Neri17, Carlos Vascconcelos18, Monica Muntoni19, Alessandro Mattiace20, University of Cagliari, Rheumatology, Monserrato, Italy; 2University of Barcelona, Autoimmune Diseases, Barcelona, Catalonia, Spain; 3University of Cret, Rheumatology, Clinical Immunology and Allergy, Heraklion, Greece; 4University of Ferrara, Rheumatology, Ferrara, Italy; 5University of Bari, Rheumatology, Bari, Italy; 6University of Siena, Rheumatology, Siena, Italy; 7Università Politecnica delle Marche, Ophthalmology, Ancona, Italy; 8Centro Hospitalar do Porto/Hospital do Santos Antonio, Neurology, Porto, Portugal; 9University of Porto, UMB, Abel Salazar Biomedical Sciences Institute, Porto, Portugal; 10Hospital Santo Antonio Centro Hospitalar do Porto, Unidade de Imunologia Clinica, Porto, Portugal; 11Cleveland Clinic Abu Dhabi, Eye Institute, Abu Dhabi, United Arab Emirates; 12Patient delegate, Cagliari, Italy

**Background:** Irreversible organ damage is considered a core outcome by the OMERACT working group. However, no specific tools are currently available to detect and measure damage accrual in Behçet’s syndrome (BS).

**Objectives:** To develop and preliminarily validate the Behçet’s syndrome Overall Damage Index (BODI).

**Methods:** A preliminary version of the instrument (p-BODI) was developed by reviewing pre-existing tools [e.g. Vasculitis damage index (VDI)] and through an extensive literature review. p-BODI was then reviewed and implemented by a multi-rounds Delphi process, involving an international and multidisciplinary (5 rheumatologists, 4 internist, 1 ophthalmologist, 1 neurologist) panel of experts in BS management and a patients’ delegate. A group of clinicians (CG), not involved in the BODI development, was asked to independently score a set of clinical vignettes, in order to test the instrument reliability. A training process consisting of an user manual and a video-tutorial. Then, Cohen’s K and intra-class correlation coefficient (ICC) between assessors and gold standard were calculated. Afterwards, BODI validation was conducted according to the OMERACT Filter 2.0 in a multicenter BS cohort.

**Results:** Starting from a list of 120 candidate items, the final version of BODI consisted of 4 overarching principles, 30 items and 12 sub-items (each of them scores one point) grouped in 8 domains (figure). In terms of reliability, the mean K coefficient was 0.84 (95%CI 0.78 to 0.90) and the ICC was 0.88 (95%CI 0.80-0.95). Validation cohort consisted of 228 BS patients (49.1% males), with a median (IQR) age and disease duration of 46.9 (35.5-55.0) and 11.7 (5.9-20.7) years, respectively. Overall, prevalence of any BODI damage (BODI ≥1) was 56.1% with a median score of 1.0 (0.0-2.0). In regard to construct validity, BODI score significantly correlated with VDI (Spearmann’s rho 0.693, p <0.001). Besides, BODI score did not correlate with BDCAF (rho=0.016, p=0.807), contrary to VDI (rho 0.141, p=0.034). Such results support the validity of BODI, unlike VDI, in discriminating damage from non-disease activity in BS. Preliminary data encourage further validation of BODI in more extended and multi-ethnic BS cohorts before being applied in clinical practice and as a therapeutic outcome.

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