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-dependent asthma is frequent, and GC-related intolerance concepts have been described. EGPA patients may be classified into distinct GC-resistant phenotypes. The aim of this study was to establish new phenotypes and evaluate their clinical relevance.

METHODS: We set up a multicenter European cohort that included 636 EGPA patients. Based on recent consensus, we distinguished 4 EGPA-proliferative profiles: GC-dependent asthma and/or ENT manifestations, eczema, systemic symptoms, and pulmonary infiltrates. Patients with vasculitis relapse(s) had more frequently neurological manifestations (p = 0.01), past major organ involvement (p = 0.001), past hospitalization (p = 0.001), and they had more frequent active asthma at last follow-up (p < 0.0001), and they had more frequent active asthma at last follow-up (p < 0.0001).

RESULTS: After median follow-up of 63 (IQR 30-110) months, 35.8% had GC-dependent asthma and/or ENT manifestations, 12.9% had vasculitis relapse(s), 14.3% had both phenotypes, 14.6% were in complete remission, 14.4% were in partial remission, 7.8% had not reach remission.

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

Matthias Pago1, Renato A. Sinico2, Vitor Teixeira3, Maria-Letizia Urban4, Juliane Mahrho5, Sara Monte5, Giulia Cassone4, Franco Schiavon4, Benjamin Seeliger5, Thomas Neumann5, Klaus Kroege1, Matthieu Groh6, Chiara Marvin6, Maxime Samson6, Thomas Barba6, David Jayne6, Bernhard Hellmich7, Carloanmauro Monteuccio8, Carlo Salvareni8, Jean-Emmanuel Kahn9, Bernard Bonnotte10, Bernard Durel10, Luc Mouthon11, Xavier Puéchal11, Loïc Guillevin11, Giacomo Emmi11, None declared, Maxime Samson: None declared, Thomas Barba: None declared, David Jayne: None declared, David Kroege: None declared, Chiara Marvini: None declared, Maxime Samson: None declared, Thomas Barba: None declared, David Jayne: None declared, David Yame: Grant/research support from: David Jayne has received research grants from Chemocentryx, GSK, Roche/Genentech and Sanofi-Genzyme. He has received consultancy fees from Astra-Zeneca, Boehringer-Ingelheim, Chemocentryx, Chugui, GSK, Infra-RX, Insmed and Takeda, Bernhard Hellmich Consultant for: Roche, Speakers bureau: Abbvie, MSD, Roche, Novartis, Pfizer, Carloanmauro Monteuccio Speakers

Conclusion: EGPA seems to evolve toward distinct phenotypic profiles, which could be identified using baseline and follow-up characteristics. Early identification of those profiles could allow guided choices of the best therapeutic option.

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

Matteo Piagi1, Alberto Floris1, Gerard Espinosa2, Nikolaos Koukakis2, Andrea Lo Monaco3, Giuseppe Lopalo3, Ida Orlando3, Vittorio Pirani3, Emedesta Santos4,5,6, Luisa Serpa Pinto4,7, George Bertzias8, Luca Cantarini8, Alberto Caui8, Ricard Cervera9, João Correia9, Marcello Govoni10, Florenzo Iannone11, Ana Martins Da Silva12,13, Piergiovanni Neri11,13, Carlos Vasconcelos14, Monica Muntoni15, Alessandro Mathieu15, University of Cagliari, Rheumatology, Monasterrà, Italy, University of Barcelona, Autoimmune Diseases, Barcelona, Catalonia, Spain, University of Ciret, Rheumatology, Clinical Immunology and Allergy, Heraklion, Greece, University of Ferrara, Rheumatology, Ferrara, Italy, University of Bari, Rheumatology, Bari, Italy, University of Siena, Rheumatology, Siena, Italy, Università Politecnica delle Marche, Ophthalmology, Ancona, Italy, Centro Hospitalar do Porto/Hospital do Santo António, Neurology, Porto, Portugal, University of Porto, UMB, Abel Salazar Biomedical Sciences Institute, Porto, Portugal, Hospital Santo Antonio Centro Hospitalar do Porto, Unidade de Imunologia Clínica, Porto, Portugal, Cleveland Clinic Abu Dhabi, Eye Institute, Abu Dhabi, United Arab Emirates, Patient 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 internists, 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. The instrument validity was assessed using 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 (0-2.0). In regard of construct validity, BODI score significantly correlated with VDI (Spearman’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 current disease activity in BS. On multiple regression analysis, factors independently associated to higher BODI damage score were male gender (β coefficient 0.143; p = 0.014), longer disease duration (0.221; p < 0.001), past major organ involvement (β 0.377; p < 0.001) and required use of anti-TNFα inhibitors (β 0.222; p < 0.001).

Full agreement among the CG was reached in judging BODI as a credible, comprehensive, easy to use, timesaving and acceptable instrument. BODI was extended and multi-ethnic BS cohorts before being applied in clinical practice and as a therapeutic outcome.
HEALED TEMPORAL ARTERITIS. ANALYSIS OF PATIENTS REFERRED TO RHEUMATOLOGY WITH A DIAGNOSIS OF TEMPORAL ARTERITIS

Lily Romero Karam1, Shrutti Agashe2, Ileana De Anda-Duran2, Ramona Mihu4, Jovan Popovich5, Houston Methodist, Internal Medicine, Houston, United States of America; 2Houston Methodist, Neurology, Houston, United States of America; 3Harvard T.H. Chan School of Public Health, MPH-Epidemiology, Boston, United States of America; 4Houston Methodist, Rheumatology, Houston, United States of America

Background: The diagnosis of giant cell arteritis (GCA), or temporal arteritis (TA), is a clinical one. Temporal artery biopsy (TAB) is usually obtained prior to treatment. Unfortunately, a biopsy does not always confirm the diagnosis; results can be reported as active, healed, or negative even in patients with all other features of giant cell arteritis. Histopathological features suggestive of resolving inflammation are evident in healed TA, but these are non-specific and can also be seen in other conditions that are unlikely to benefit from prolonged steroid treatment. It is not clear if a pathological diagnosis of healed TA helps to support treatment which may be associated with significant toxicity.

Objectives: We analyzed clinical features of patients with “healed” TA and compared them to patients with active and negative biopsies with the goal of identifying any features that may aid treatment decisions.

Methods: Retrospective analysis of 49 patients with established TA diagnosis, seen at a rheumatology practice in Houston, Texas from 8/2007 to 12/2018. Fifty seven percent were referred by Ophthalmology, the rest by Neurology. Patients were divided into 3 groups based on biopsy: Active, Healed, and Negative. Clinical features analyzed at the time of presentation were gender, age of onset, fulfillment of the American College of Rheumatology (ACR) 1990 criteria for the classification of Giant Cell Arteritis and the 2010 revised ACR criteria for early diagnosis of Giant Cell Arteritis (rACR), and the presence of vision abnormalities, headache, temporal artery tenderness/induration, jaw claudication, fever, anemia, sedimentation rate at onset, and flares during follow up. Descriptive data was reported as means ± standard deviation for continuous variables and as proportions for categorical variables; analyzed with ANOVA and chi squared, respectively. Post hoc analysis with Bonferroni corrections and Fisher’s exact tests were used. All analyses were performed on Stata version 15 (StataCorp LP, College Station, TX, USA). A p-value of ≤ 0.05 was considered statistically significant.

Results: Sixty-two (65%) of the 94 patients were females and 17 (35%) were male. Biopsy reports showed active disease in 14 (28.5%), healed in 31 (63.3%) and negative in 4 (8.2%) patients. Average age was 72.7 years ±6.3, 73.0 ±9.1, 61.25 ±1.3 years in these 3 groups, respectively (p = 0.008). The Active versus Healed groups were statistically different with respect to age (p = 0.028), those meeting ACR and rACR criteria (p = 0.029 and p = 0.002), and the incidence of jaw claudication: 8 active (57.1%) versus 0 healed (16.1%) patients (p = 0.03). There was no statistically significant difference in ophthalmological manifestations, headache, temporal artery palpation, fever, anemia, and flares. In addition, 90% of patients reported side effects from steroids.

Conclusion: The percent of healed TA was 63.3%, higher than in other studies. Patients with healed TA were less likely to fulfill ACR and rACR criteria for giant cell arteritis. Further studies are needed to identify the subset of patients with healed TA who would have a clear benefit from high dose steroids, that would outweigh the treatment side effects. At the end of the day, the diagnosis of TA remains a clinical one and early involvement of Rheumatology may help avoid unnecessary biopsies and treatment. Ultrasound, MRA, and PET-CT scan may be able to fill the gap when biopsy fails to confirm the diagnosis.