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

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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. Evolving concepts distinguish vasculitis-related symptoms from asthma and/or ENT manifestations. That distinction has become even more important since the development of B-cell and eosinophil-targeted therapies.

Objectives: This study aimed to describe and identify characteristics predicting long-term EGPA outcomes.

Methods: We set up a multicenter European cohort that included 636 EGPA patients. Based on recent consensus, we distinguished 4 EGPA-evolutionary profiles: GC-dependent asthma and/or ENT manifestations (requiring prednisone >7.5 mg/d), ≥1 vasculitis relapse(s) (excluding asthma and/or ENT flares), both phenotypes, and complete remission (no GC-dependent asthma/ENT signs, no vasculitis relapse and prednisone <5 mg/d at last follow-up). Baseline and follow-up characteristics predicting those outcomes were analyzed.

Results: After median follow-up of 63 (IQR 30-110) months, 35.8% had GC-dependent asthma and/or ENT manifestations, 12.9% had ≥1 vasculitis relapse(s), 14.3% had both phenotypes, 14.6% were in complete remission, 14.4% were in partial remission and 7.8% had not reach remission.

Patients with GC-dependent asthma/ENT manifestations were younger at diagnosis (p<0.0001), had more frequent GC-treated asthma before overt EGPA (p=0.002), had more ENT manifestations (p=0.01) and less frequent MPO-ANCA (p<0.0001). Their daily GC dose was higher at every time point (p<0.0001), and they had more frequent active asthma at last follow-up (p<0.0001).

Patients with vasculitis relapse(s) had more frequently neurological manifestations at diagnosis (p=0.002) and MPO-ANCA positivity (p<0.0001), and less frequently pulmonary infiltrates (p=0.031). Median time from diagnosis-to-1st vasculitis relapse was 25 (11–60) months. During follow-up, their daily GC dose was lower than those with GC-dependent asthma/ENT manifestations, but similar to those in complete remission. At last follow-up, neurological sequelae tended to be more frequent (p=0.06).

Finally, patients in complete remission were older (p<0.0001), had more fever (p=0.03), less GC-treated asthma (p=0.002) and ENT manifestations at diagnosis (p=0.01), lower daily GC dose during follow-up (p<0.0001), lower eosinophils count at 6 months (p=0.002) and less frequent sequelae (p=0.003).

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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SAT0240

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

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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 **B**ehçet's syndrome **O**verall **D**amage **I**ndex (BODI). NCT03803462.

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, after a training process consisting of a 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 (5.8-20.7) 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 (β0.221; 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.

Conclusion: BODI is the first tool specifically developed to assess damage 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.