Table 2. Characteristics of patients with and without poor outcome

	With Poor Outcome n= 16	Without Poor Outcome n= 78	p value
Gender (male, %)	9 (56.3)	40 (51.3)	0.72
Type of neurological involvement, p-NBS	15 (93.8)	37 (47.4)	0.001
Age at Behçet diagnosis (year)	30.7 ± 11.0	28.9 ± 9.2	0.50
Age at neurological involvement (year)	38.9 ± 12.2	35.5 ± 11.9	0.307
Initial mRS ≥ 3	14 (87.5)	19 (24.4)	<0.001
Genital ulcer	10 (62.5)	61 (78.2)	0.21
Skin involvement (PPE+EN)	12 (75)	60 (76.9)	1.00
Musculoskeletal involvement	10 (62.5)	36 (46.2)	0.23
Ocular involvement	10 (62.5)	37 (47.4)	0.27
Vascular involvement (except SVT)	1 (6.3)	28 (35.9)	0.019
Gastrointestinal involvement	1 (6.3)	4 (5.1)	1.00
HLA B51 positivity (n=32)	1 (50)	13 (43.3)	1.00
Clinical characteristics			
Headache	6 (37.5)	67 (85.9)	<0.001
Fever	0 (0)	6 (7.7)	0.585
Paresis	12 (75)	14 (17.9)	<0.001
Pyramidal signs	14 (87.5)	20 (25.6)	<0.001
Sensory symptoms	11 (68.8)	23 (29.5)	0.003
Cerebellar symptoms	7 (43.8)	12 (15.4)	0.017
Cranial nerve involvement	3 (18.8)	10 (12.8)	0.69
Convulsion	1 (6.3)	2 (2.6)	0.43
Papilledema	2 (12.5)	22 (28.2)	0.23
Nausea-vomiting	3 (18.8)	20 (25.6)	0.753
Treatment			
Colchicine	16 (100)	73 (93.6)	0.584
Anticoagulation	5 (31.3)	26 (33.3)	0.87
Azathioprine	15 (93.8)	70 (89.7)	1.00
Cyclophosphamide	9 (56.3)	13 (16.7)	0.002
Pulse steroid	13 (81.3)	27 (34.6)	0.001
Oral steroid	16 (100)	59 (75.6)	0.036
Interferone	1 (6.3)	8 (10.3)	1.00
Cyclosporine	2 (12.5)	12 (15.4)	1.00
Methotrexate	1 (6.3)	1 (1.3)	0.313
Acetylsalicylic acid	5 (31.3)	34 (43.6)	0.36
Diazemide	1 (6.3)	13 (16.7)	0.451
Total follow-up time (month)	66.84 (IQR: 67.3)	49.4 (IQR: 80.7)	0.36

mRS: Modified Rankin Score, PPE: Papulopustular eruption, EN: Erythema nodosum, SVT: Sinus Vein Thrombosis

sequela and a total of 5 (5.3%) deaths occurred. Characteristics of patients with and without poor outcome are presented in table 2. In multivariable logistic regression analysis, factors associated with poor outcome were initial mRS (OR 24.2 (95% CI 3.16 – 108.67)) and age ≥ 40 at NBS diagnosis (OR 4.59 (95% CI 1.02 – 20.69)), meanwhile, headache at presentation was associated with a lower risk for poor outcome. (OR 0.22 (95% CI 0.05 – 0.91)).

Conclusion: Neurologic involvement is a detrimental manifestation of BS and causes disability, even death. Patients who have an initial disabling presentation in advanced age are more likely to have poor prognosis. Treatment intensification in this subpopulation might be considered.

References: None

Disclosure of Interests: None declared

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SAT0274

DESCRIPTIVE ANALYSIS OF FLARES DURING THE LONG-TERM FOLLOW-UP OF PATIENTS WITH BEHÇET'S DISEASE INCLUDED IN REGEH COHORT

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Background:

Objectives: To describe the clinical manifestations and treatment of flares during follow-up in patients with Behçet's disease (BD) included in the REGEH cohort over the last decade.

Methods: The Spanish Registry of BD or REGEH (REGistro de la Enfermedad de Behçet as Spanish nomenclature) Project Group was created by the Spanish Internal Medicine Society in 2009 with the aim of compiling a large cohort of Spanish patients with this rare disorder. By July 2012, REGEH has collected 635 BD patients with a multicentre, longitudinal and consecutive design. Diagnosis of BD was performed on the basis of the International Study Group criteria for BD. Since inclusion in the registry, patients have been followed

prospectively and new flares have been recorded, defined as those clinical manifestations which have required initiation or modification of immunosuppressive treatment, or prednisone dose at or higher than 10mg/d of prednisone during more than 1 month.

Results: A prospective follow-up of 326 patients included in the registry has been analysed. There are 182 women (56%) and their average age (SD) at the time of diagnosis was 33 (13) years. The majority of them were Caucasian (91%). The median follow-up time from BD diagnosis was 180 months, and the prospective follow-up period from the inclusion in the registry was 80 months. The cumulative clinical manifestations until the initial registration were oral ulcers in 100% of patients, genital ulcers in 221 (68%), arthritis in 147 (45%), erythema nodosum in 96 (29%), fever in 82 (25%), thrombosis in 74 (23%), anterior uveitis in 76 (23%), retinal vasculitis in 46 (14%), posterior uveitis in 35 (11%), aseptic meningitis in 32 (10%), and other neurological manifestations in 15 (5%) patients. One hundred and fifty-six (48%) patients received immunosuppressants and 47 (14%) biological therapy.

During follow up (period from the inclusion in the REGEH to the last visit) 68 (23%) patients presented at least a severe outbreak. The median time from the BD diagnosis to the first flare were 170 months. The main clinical manifestations were oral-genital ulcers in 43 (63%) patients, uveitis in 31 (45%), arthritis in 13 (19%), neurological in 16 (24%), vascular in 10 (15%), and gastrointestinal in 3 (4%) patients. Immunosuppressants were used in 37 (54%) patients and biological therapy in 14 (21%). Biological therapy was mostly used due to refractory disease, the majority of cases because ocular manifestations. There were no differences in terms of age, sex, race and previous clinical manifestations between patients who suffered from flares and those who did not. Flares were more frequent in patients who have received previous immunosuppressive or biological treatment (35% vs 13% and 48% vs 19%, respectively) (p<0.001 in both cases), probably reflecting a more severe disease.

Conclusion: The long-term follow-up of BD patients from REGEH cohort showed that 10 years after diagnosis, a fifth of them may continue to present severe flares requiring systemic treatment. The use of biological therapy increased over time and their main indication was refractory disease.

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SAT0275

MAINTAINED BENEFIT IN HEALTH-RELATED QUALITY OF LIFE OF PATIENTS WITH GIANT CELL ARTERITIS TREATED WITH TOCILIZUMAB PLUS PREDNISONE TAPERING: RESULTS FROM THE OPEN-LABEL, LONG-TERM EXTENSION OF A PHASE 3 RANDOMIZED CONTROLLED TRIAL

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Background: In part 1 of the 52-week, double-blind GACTA trial, patients with giant cell arteritis (GCA) who received weekly tocilizumab (TCZ) plus prednisone tapering reported improvement in the 36-item Short-Form Health Survey (SF-36) Mental Component Summary (MCS) and Physical Component Summary scores and FACIT-Fatigue scores that were statistically significant and clinically meaningful compared with patients who received prednisone alone.¹