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
Diazoxide	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.

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SAT0274

DESCRIPTIVE ANALYSIS OF FLARES DURING THE LONG-TERM FOLLOW-UP OF PATIENTS WITH BEHÇET'S DISEASE INCLUDED IN REGEBOH 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 REGEBOH cohort over the last decade.

Methods: The Spanish Registry of BD or REGEBOH (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, REGEBOH 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 REGEBOH 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 REGEBOH 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

Objectives: To analyze whether benefit in SF-36 MCS was maintained in patients originally assigned to TCZ compared with those originally assigned to placebo (PBO) plus a 26- or 52-week prednisone taper among patients who achieved clinical remission at week 52 and maintained treatment-free clinical remission in the 2-year, long-term extension of GiACTA.

Methods: At the end of part 1, patients entered open-label part 2, in which GCA therapy (including initiation/termination of open-label TCZ and/or GCs) was given at the investigator's discretion according to disease status. Change from baseline in SF-36 MCS score was compared for combined original TCZ (n = 33) and PBO (n = 17) patients who achieved clinical remission at week 52 and maintained treatment-free (no TCZ or GCs) clinical remission in part 2 using a repeated-measures model. The minimal clinically important difference (MCID) for SF-36 MCS is >2.5.²

Results: During treatment, SF-36 MCS scores in all 50 patients who maintained treatment-free clinical remission in part 2 had diverged between the TCZ and PBO groups as early as 36 weeks after baseline, with greater improvements evident in the TCZ group (Figure). The difference in least square means (LSM) change between TCZ and PBO was statistically significant at week 52 (p = 0.016) and maintained at weeks 100 (p = 0.023) and 156 (p = 0.002). The LSM difference (95% CI) between TCZ and PBO at weeks 52, 100, and 156 was 5.6 (1.1-10.2), 6.5 (0.9-12.1), and 7.4 (2.9-11.9), respectively, exceeding the MCID.

Conclusion: Among patients who maintained treatment-free clinical remission during part 2 of GiACTA, those originally assigned to receive TCZ plus a prednisone taper during part 1 maintained statistically significant and clinically meaningful improvements in SF-36 MCS up to week 156 compared with those originally assigned to receive PBO plus a prednisone taper in part 1. This was true even though neither of the patient groups received TCZ or GC treatment after they achieved clinical remission at week 52.

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SAT0276 CARDIAC MAGNETIC RESONANCE IMAGING IN PATIENTS WITH BEHCET'S DISEASE: A PILOT STUDY

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Background: Behçet's disease is a multisystemic vasculitis characterized by mucocutaneous, ocular, arthritic, and vascular manifestations.[1] However, the incidence and nature of cardiac involvement in Behçet's disease have not been clearly documented yet.

Objectives: This study aimed to assess the cardiac involvement in patients with Behçet's disease using cardiac magnetic resonance imaging (MRI).

Methods: This cross-sectional descriptive study was carried out on thirty consecutive patients with Behçet's disease (21 males, 9 females) with mean age 32.3±8.9 years and with no evidence of cardiac disease. They underwent cardiac MRI to determine morphological and functional changes of the heart and to detect areas of hyperenhancement after IV administration of gadolinium.

Results: At least one abnormality on cardiac MRI was observed in 20/30 patients (66.67%). Myocardial oedema was observed in 3 patients (10.0%) and late gadolinium enhancement in only 1 patient (3.3%). Pericardial effusion was found in 3 patients (10.0%), global hypokinesia in 6 patients (20.0%) and intra-cardiac thrombosis in only 1 patient (3.3%). Pulmonary artery was dilated in 4 patients (13.3%). Left ventricular and right ventricular end diastolic volume were altered in 4 patients (13.3%) and 7 patients (23.3%) respectively. Also, left ventricular and right ventricular end systolic volume were abnormal in 7 patients (23.3%) and 5 patients (16.7%) respectively. Moreover, there was aortic valve regurgitation in 2 patients (6.7%), tricuspid valve regurgitation in 9 patients (30%), and mitral valve regurgitation in 9 patients (30%). Dilated left main coronary artery in 2 patients (6.7%) and arrhythmogenic right ventricular dysplasia in only one patient (3.3%).

Conclusion: Behçet's disease may cause cardiac abnormalities without clinical manifestations and cardiac MRI may represent a tool for early detection of these subtle abnormalities.

References:

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Figure 1. Cardiac MRI in 32 year-old female patient with Behçet's disease for 3 years.

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SAT0277 HEAD AND NECK INVOLVEMENT OF IGA VASCULITIS: A CASE-CONTROL STUDY

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Background: IgA vasculitis (IgAV) is an immune-complex mediated, small-vessel vasculitis which predominantly involves the skin on the lower extremities. Head and neck involvement is rarely reported.

Objectives: To describe the presentation and outcome of a series of patients with head and/or neck involvement in comparison to patients with cutaneous findings isolated to the lower extremities.