and recovered with CYC and anti-epileptic drugs. The second patient with blurred vision and headache had cerebral venous sinus thrombosis (CVST) and recovered with anticoagulant therapy. The third patient with muscle weakness died due to spondylodiscitis complicated with aortic pseudoaneurysm.

In 5 patients (26%), neurologic work-up did not lead to an underlying condition. The presenting symptoms of these patients were transient acute vision loss in 2, numbness of extremities in 1, syncope in 1 and vertigo in 1 patient. Neurologic symptoms resolved after high dose GC and RTX in the patient with vertigo. At the onset of neurologic symptoms, 3 patients were using IS therapy including azathioprine, MMF and CYC in 1 patient each. The fourth patient was off treatment. Neurologic symptoms were transient in these patients, and did not recur during our follow-up of 36, 52, 57, and 120 months.

Conclusion: CNS involvement appears to be rare in AAV and non-CNS entities including ocular, orbital and sinonasal involvement and complications such as PRES, CVST and infections may mimic CNS involvement in patients with AAV.

Disclosure of Interests: Yeliz Yagiz Ozogul: None declared, Sinem Nihal Esatoglu Speakers bureau: Sinem Nihal Esatoglu has received honorariums for presentations from UCB Pharma, Roche, Pfizer, and Merck Sharp Dohme, Murat Ozogul: None declared, Osman Kizilkilic: None declared, Yesim Ozguler Speakers bureau: Yesim Ozguler has received honorariums for presentations from UCB Pharma, Novartis, and Pfizer., Vedat Hamuryudan Speakers bureau: Vedat Hamuryudan has served as a speaker for AbbVie, Celgene, Novartis, and UCB Pharma., Grant/research support from: Vedat Hamuryudan has received grant/research support from Celgene., Gulen Hatemi Speakers bureau: Gulen Hatemi has served as a speaker for AbbVie, Celgene, Novartis, and UCB Pharma, Grant/research support from: Gulen Hatemi has received grant/research support from Celgene.

DOI: 10.1136/annrheumdis-2022-eular.4011

AB0636

RELATIONSHIP BETWEEN ORGAN DAMAGE AND IMPAIRMENT OF HEALTH-RELATED QUALITY OF LIFE IN PATIENTS WITH BEHÇET'S SYNDROME: RESULTS FROM A LONGITUDINAL EXTENSION OF THE BODI PROJECT.

A. Floris[1,2], R. Laconi[1,2], G. Espinosa[3], G. Lopalco[4], L. Serpa Pinto[5], N. Kougkas[6,7], J. Sota[8], A. Lo Monaco[9], M. Govoni[9], L. Cantarini[8], G. Bertsias[6], J. Correia[5], F. Iannone[4], R. Cervera[3], C. Vasconcelos[10], A. Mathieu[1], A. Cauli[1,2], M. Piga[1,2]on behalf of BODI project group. *[1]University of Cagliari , Department of Medical Sciences and Public Health, Cagliari, Italy; [2]AOU University Clinic and University of Cagliari, Rheumatology Unit, Cagliari, Italy; [3]University of Barcelona, Department of Autoimmune Diseases, Hospital Clinic, Institut d'Investigacions Biomediques August Pi i Sunyer (IDIBAPS), Barcelona, Spain; [4]University of Bari , Rheumatology Unit, Bari, Italy; [5]Hospital Santo Antonio Centro Hospitalar do Porto, Unidade de Imunologia Clinica, Porto, Portugal; [6]University of Crete, Rheumatology, Clinical Immunology and Allergy Unit, Heraklion, Greece; [7]Aristotle University of Thessaloniki, Fourth Department of Internal Medicine, Hippokration University Hospital, Medical School,Thessaloniki, Greece; [8]University of Siena , Rheumatology Unit, Siena, Italy; [9]University of Ferrara, Rheumatology Unit - AOU S. Anna, Ferrara, Italy; [10]University of Porto, UMIB Abel Salazar Biomedical Sciences Institute, Porto, Portugal*

Background: Preventing accrual of organ damage represents a primary goal in the treatment of Behçet's Syndrome (BS), as it may result in impairment of other outcomes, including the health-related quality of life (HR-QoL).

Objectives: The objective of this study was to investigate whether the recent accrual of organ damage, rather than its extent at a single time point, correlate with an impairment of the HR-QoL.

Methods: A sub-analysis of data from patients recruited in the longitudinal phase of the BODI Project validation cohort was performed. The HR-QoL and damage were measured by the Short-form 36 questionnaire (SF-36) and the BS Overall Damage Index (BODI), respectively, at the baseline visit and at a follow-up (FU) 24 ±3 months later. Then the possible increase of damage over FU was assessed by calculating the difference between the BODI score (Δ-BODI) in the two visits. Then, the relationship between the Δ-BODI and the individual and summary domains of the SF-36 was analysed by building multivariate regression models, including age, gender, concomitant fibromyalgia and/or depression, current disease activity as assessed by the BDCAF, as confounding variables.

Results: From the BODI validation cohort, 147 patients were recruitable for this sub-analysis;73 (49.8%) were males. The mean (SD) age and disease duration at enrolment were, respectively, 46.2 (12.4) and 13.4 (10.1) years. BODI score did not influence the SF-36 domains assessed at the baseline visit. In contrast, a significant correlation was recorded between the Δ-BODI and the following SF-36 domains: physical function (PF) (β -0.158 for 1 unit increase in BODI score, p 0.025), role physical (RP) (β -0.150, p 0.044), general health (GH) (β -0.199, p 0.004), role emotional (RE) (β -0.180, p 0.001), mental health (MH) (β -0.244, p 0.001), and the mental components summary (MCS) (-0.203, p 0.008) (Figure 1). Gender, age, fibromyalgia and disease activity were also confirmed to significantly influence HR-QoL (Table 1).

Table 1. Multiple regression for the assessment of the relationship between Δ-BODI and SF-36 domains

	Δ-BODI	Male	Age	FBM	DPR	BDCAF
Physical function (PF)	-0.158 **(p 0.025)**	0.180 (p 0.010)	-0.299 (p<0.001)	-0.358 (p<0.001)	-- (p 0.552)	-0.141 (p 0.044)
Role-physical (RP)	-0.150 **(p 0.044)**	0.154 (p 0.039)	-0.212 (p 0.001)	-0.278 (p<0.001)	-- (0.086)	-0.251 (p<0.001)
Body-pain (BP)	-- 0.868	0.266 (p<0.001)	-0.286 (p<0.001)	-0.276 (p<0.001)	-- (p 0.799)	-262 (p<0.001)
General health (GH)	-0.199 **(p 0.004)**	0.187 (p 0.010)	-- (0.136)	-0.296 (p<0.001)	-- (0.861)	-0.352 (p<0.001)
Vitality (VT)	-- (p 0.868)	0.238 (p 0.001)	-0.178 (p 0.008)	-0.213 (0.002)	-- (p 0.855)	-0.371 (p<0.001)
Social function (SF)	-- (p 0.239)	0.299 (p 0.004)	-0.166 (p 0.024)	-0.242 (p 0.001)	-- (0.831)	-0.202 (p 0.010)
Role emotional (RE)	-0.180 **0.003)**	0.158 (p 0.047)	-0.157 (p 0.048)	-0.233 (p 0.003)	-- (0.531)	-0.191 (p 0.016)
Mental health (MH)	-0.244 **(p 0.001)**	-- (p 0.142)	-- (p 0.142)	-0.292 (p<0.001)	-- (p 0.073)	-0.254 (p 0.001)
Physical Component Summary (PCS)	-- 0.105	0.229 (p 0.001)	-0.298 (p<0.001)	-0.296 (p<0.001)		-0.254 (p<0.001)
Mental Component Summary (MCS)	-0.203 **(p 0.008)**	-- (p 0.068)	-- (0.246)	-0.255 (p 0.001)	-- (0.122)	-0.302 (p<0.001)

FBM: fibromyalgia; DPR: depression

Conclusion: The recent accrual of organ damage, rather than its extent assessed in a single visit, is associated with impairment of different aspects of heath related quality of life, especially those mental related. Such phenomenon is similar to that observed in other systemic rheumatic disease, may be due to coping mechanisms.

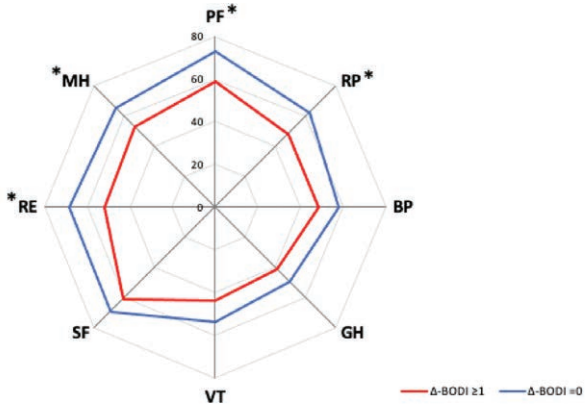

Figure. Impact of increase of damage accrual of 2 years of follow-up on different domains of the SF-36 questionnaire for measurement of the heath related quality of life. PF, physical function. RP, role-physical. BP, body-pain. GH, general health. VT, vitality. SP, social function. RE, role emotional (RE), MH, mental health. * p < 0.005 in univariate analysis using Mann Whitney test.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.4060

AB0637

THE ROLE OF ANTINEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA) IN ASSESSMENT OF DISEASE ACTIVITY IN SYSTEMIC VASCULITIS

L. Petelytska[1], O. Iaremenko[1], M. Nahirna[1]. *[1]Bogomolets National Medical University ,The Department of Internal Medicine #3, Kyiv, Ukraine*

Background: Despite the proven diagnostic significance, the prognostic role of ANCA, in particular for assessing disease activity, remains questionable. Numerous studies have attempted to estimate the role of ANCA monitoring with different results and a lack of consensus on reported outcomes [1].

Objectives: To analyze the relationship between ANCA level and clinical or laboratory parameters of disease activity.

Methods: This is a retrospective analysis of 38 patients with ANCA-associated vasculitis (granulomatosis with polyangiitis – 25 patients, microscopic polyangiitis – 6 patients and eosinophilic granulomatosis with polyangiitis – 7 patients) from a single center observed from 2015 till the end of 2020. The diagnosis of ANCA-associated vasculitis was performed according to the ACR 1990 criteria or the Chapel Hill Consensus Conference 2012 nomenclature. The study included 20 women (52.6%) and 18 men (47.4%). The average age of patients was 49 (27-62) years, the mean duration of the disease was 26 (6-120) months. The clinical data, initial Birmingham vasculitis activity score (BVAS), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), ANCA (ELISA test) against proteinase-3 (PR-3) and myeloperoxidase (MPO) were evaluated. Spearman's correlation analysis was used to investigate the relationship between ANCA levels and ESR, CRP levels, BVAS activity index. The diagnostic value of ANCA in determining the active disease was evaluated by ROC analysis with an estimation of the area under the ROC curve (AUC). The definition of active disease included new, persistent, or worsening clinical signs and/or symptoms attributed to GPA, MPA, or EGPA and not related to prior damage [2].
Results: Positivity for MPO-ANCA was observed in 23.7% of patients, and for PR3-ANCA - in 76.3% of patients. The BVAS activity index averaged 16 (IQR-13) points. The mean CRP level was 47.9 (IQR-90.0) mg/L and the ESR level was 30.1 (IQR-33.5) mm/h. There was a positive correlation between the level of both ANCA and the BVAS index (r = 0.43; 95% CI 0.11-0.66; p <0.01), as well as the level of ESR (r = 0.37; 95% CI 0.05-0.63; p <0.05). No relationship was found between CRP level and ANCA level (r = 0.22; 95% CI -0.15-0.54; p> 0.05), but a positive correlation was observed between CRP level and index BVAS activity (r = 0.41; 95% CI 0.05-0.67; p <0.05). When using ROC-analysis to determine the value of ANCA in the assessment of active disease, it was found that the AUC is 0.93 ± 0.04 (95% CI 0.84-1.01; p <0.01), which indicates excellent ability ANCA diagnose patients with active disease (sensitivity - 87.9%, specificity - 80.0%).
Conclusion: The level of ANCA in patients with ANCA-associated vasculitis correlates with the Birmingham vasculitis activity score, as well as with the level of ESR. Determination of ANCA level can be used not only to diagnose ANCA-associated vasculitis, but also to assess disease activity.

REFERENCES:

[1] Adam D. Morris, Anthony W. Rowbottom, Francis L. Martin, Alexander Woywodt and Ajay P. Dhaygude Kidney360 March 2021, 2 (3) 586-597; DOI: https://doi.org/10.34067/KID.0006432020
[2] Chung, S. A., Langford, C. A., Maz, M., et al. (2021). 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. Arthritis & rheumatology (Hoboken, N.J.), 73(8), 1366–1383. https://doi.org/10.1002/art.41773

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.4223

AB0638 INTRAVENOUS IMMUNOGLOBULIN IN ANTINEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS. STUDY OF 28 CASES FROM A SINGLE UNIVERISTARY HOSPITAL AND LITERATURE REVIEW

F. Benavides-Villanueva[1], C. Corrales-Selaya[1], J. Loricera[1], V. Calvo-Río[1], S. Castañeda[2], N. Aviles Mendez[3], J. M. Blanco Madrigal[4], M. A. González-Gay[1], R. Blanco[1]. *[1]Marqués de Valdecilla University Hospital, Rheumatology , Santander, Spain; [2]Hospital de La Princesa, Rheumatology, Madrid, Spain; [3]Hospital Universitario Nuestra Señora del Perpetuo Socorro de Albacete, Rheumatology, Albacete, Spain; [4]Basurto University Hospital, Rheumatology, Bilbo, Spain*

Background: Anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitis (AAV) includes granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA) or microscopic polyarteritis (MPA). Standard treatment is often accompained by significant adverse events. Intravenous immunoglobulins (IVIG) may constitute a therapeutic alternative, however, the data are scarce.
Objectives: To assess the utility and safety of IVIG in AAV.
Methods: Observational study of patients with AAV from Spanish referral center treated with IVIG. AAV diagnosis was based on a compatible clinical presentation and/or positive ANCA serology and/or histology. Disease activity was assessed with the Birmingham Vasculitis Activity Score (BVAS).
Results: We included a total of 28 patients; GPA (n=15), MPA (10), and EGPA (3). The main features are summarized in Table 1. The reasons for using IVIG were: **a)** relapse/refractory disease (n=20), or presence/suspicion of infection (8). We observed a rapid and maintained Clinical improvement, since first month of IVIG onset, yielding a BVAS score of zero in 56.5% of patients at 24 months (Figure 1). Serious Adverse event was only observed in 1 patient who developed congestive cardiac failure and had to stop the IVIG therapy.

Table 1. Main general features of 28 patients with antineutrophil cytoplasmic antibody-associated vasculitis treated with intravenous immunoglobulins.

GENERAL FEATURES	RESULTS	GENERAL FEATURES (Continuation)	RESULTS (Continuation)
DEMOGRAPHIC FEATURES		ANALITICAL FINDINGS	
Age of Diagnosis of AAV, mean±SD	57.1±18	CRP (mg/dL), median [IQR]	13.02
Men/ Women; n, (% men)	15/13 (53.6%)	ESR, mm/1st hour, median [IQR]	70.4
AAV Subtype, n (%)		PR3-ANCA, n (%)	11 (39.3)
GPA	15(53.6%)	MPO-ANCA, n (%)	12 (42.8)
EGPA	3(10.7%)	ANCA negative, n(%)	5 (17.8)
MPA	10(35.7%)	FFS at AAV diagnosis, n (%)	
CLINICAL MANIFESTATIONS, n (%)		0	10 (35.7)
Fever	15 (53.6%)	1	11 (39.3)
Constitutional symptoms	26 (92.85%)	2	7 (25)
ORL involvement	7 (25%)	PREVIOUS TREATMENT, n (%)	
Pulmonary involvement	19 (67.9%)	Cyclophospamide	13 (46.4%)
Renal involvement	25 (89.3%)	Methotrexate	6 (21.4%)
Cutaneous involvement)	6 (21.5%)	Azathioprine	3 (10.7%)
Ocular involvement	4 (14.3%)	Cyclophosphamide	13 (46.4%)
Joint involvement	4 (14.3%)	Mycophenolate mofetil	4 (14.3%)
Neurologic involvement	8 (28.57%)	Rituximab	5 (17.9%)

Abbreviations: ANCA:antineutrophil cytoplasmic antibody; EGPA: eosinophilic granulomatosis with polyangiitis; FFS: Five-Factors Score; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis

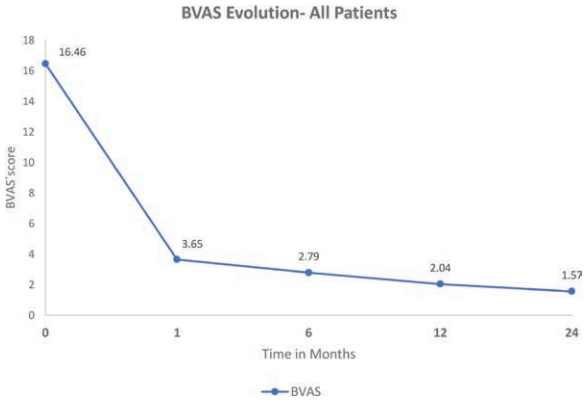

Figure 1. BVAS Evolution with IVIG treatment of all our patients.

Conclusion: IVIG seems to be an effectiveness and relative safe therapeutic option in relapse/refractory AAV or in presence of a concomitant infection.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.4369

AB0639 COMMON FEMORAL VEIN THICKNESS MEASUREMENT AS A DIAGNOSTIC TEST IN INCOMPLETE BEHÇET'S DISEASE

F. Alibaz-Oner[1], R. Ergelen[2], F. Temiz[3], S. Kutluğ Ağaçkıran[1], E. Soydemir[2], T. Ergun[4], H. Direskeneli[1]. *[1]Marmara University School of Medicine , Department of İnternal Medicine, Division of Rheumatology, İstanbul, Turkey; [2]Marmara University School of Medicine , Department of Radiology, İstanbul, Turkey; [3]Marmara University School of Medicine , Department of İnternal Medicine, İstanbul, Turkey; [4]Marmara University School of Medicine , Department of Dermatology, İstanbul, Turkey*

Background: Behçet's disease (BD) is characterized by recurrent oral and/or genital aphthous ulcers accompanied by cutaneous, ocular, articular, gastrointestinal and central nervous system lesions. Diagnosing BD can be a clinical challenge in patients presenting with a limited number of organ manifestations, especially with single major organ involvement. We reported the first controlled doppler ultrasound study showing increased common femoral vein (CFV) thickness in BD (1). We recently also showed that increased CFV thickness is a distinctive feature of BD, rarely present in other inflammatory or vascular diseases such as ankylosing spondylitis, systemic vasculitides, venous insufficiency, and non-inflammatory DVT with a specificity higher than 80% for the cut-off value of ≥ 0.5 mm. We suggest that CFV thickness measurement is an easy, non-invasive diagnostic test for BD (2).
Objectives: In this study, we aimed to assess the diagnostic performance of CFV thickness measurement in patients with 'Incomplete' Behçet's Disease diagnosed by expert opinion.