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.

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

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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

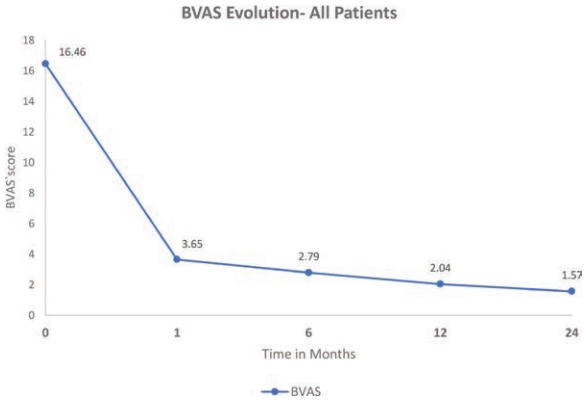

Figure 1. BVAS Evolution with IVIG treatment of all our patients.

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

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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.

Methods: We included 28 patients with incomplete BD (15 male, 12 female) diagnosed with expert opinion and followed in the Marmara University Behcet's Clinic. Demographic, clinical characteristics and treatment data were recorded during routine visits. Common femoral vein wall thickness was measured by an experienced radiologist at the same day.
Results: Median age was 34.3 years and median disease duration 2 years (0-16). Four patients were newly diagnosed. At follow-up onset, oral ulcers were present in 22 (78.6%), genital ulcers in 6 (21.6%), papulopustular lesions in 4 (14.3%) and pathergy positivity in 5 (17.9%) patients. Ten (35.7%) patients had familial BD. While 24 (85.7%) patients had major organ involvement, 4 (14.3%) patients had mucocutaneous disease. Distrubution of major organ involvements were given in Figure 1.

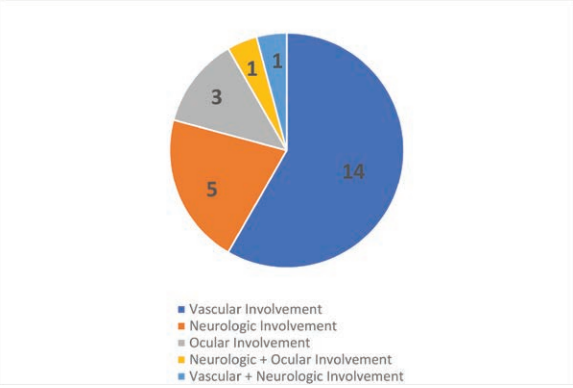

Figure 1. Distribution of major organ involvements (n) in study group

All patients except 1, had CFV thickness value above the cut-off value of ≥0.5 mm. Right CFV thickness was 0.71 (0.3-1.3) mm and left CFV thickness 0.72 (0.4-1.2) mm. Bilateral femoral vein thicknesses were similar in patients with and without an history of familial BD.
Conclusion: Diagnosing BD can be challenging in patients presenting with one major organ involvement such as oral ulcers and posterior uveitis, brain-stem disease or arterial aneurysms, especially in countries with a low prevalence. These patients are generally diagnosed as 'incomplete' BD by 'expert opinion'. Early diagnosis is of utmost importance in some of these cases, especially with venous thrombosis as their management differs from non-inflammatory venous thrombosis, necessitating immunosuppressive use rather than anticoagulant therapy. Our results show that CFV thickness measurement with Doppler US, a non-invasive, fast and cost-effective radiological modality, is a valuable diagnostic test in incomplete BD, especially with major organ involvement.
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AB0640 PREVALENCE AND DETERMINANT FACTORS OF ENDOTHELIAL DYSFUNCTION IN ANCA ASSOCIATED VASCULITIS

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Background: Atherosclerosis and its complications are one the leading cause of death in patients with anti-neutrophil cytoplasmic antibodies (ANCA)- associated vasculitis (AAVs), despite the recent remarkable improvements in prognosis. The mechanism by which atherosclerosis is accelerated in these diseases is not explained by the classical cardiovascular risk factors (CVRFs) and is still under investigation.
Objectives: To evaluate the prevalence of endothelial dysfunction (ED) in AAV patients and to identify the determinant factors of endothelial responses among vasculitis characteristics, such as ANCA status and organ involvement.
Methods: Thirty patients (15 men and 15 women) with AAVs [13 with eosinophilic granulomatosis with polyangiitis (EGPA), 7 with granulomatosis with polyangiitis (GPA), 10 with microscopic polyangiitis (MPA)] were enrolled in the study. For all subjects CVRFs, blood pressure, lipid profile, renal function, acute phase reactants, ANCA status and titers were recorded at diagnosis and at the enrollement in the study. Vasculitis disease activity was measured using the Birmingham Vasculitis Activity Score (BVAS) and organ damage was assessed with the vasculitis damage index (VDI). The prognosis was evaluated through the Five Factor Score (FFS). Patients were excluded from the study if they had previous cardiovascular or cerebrovascular events, liver failure, end stage renal disease under hemodyalisis, or cancer. Microvascular peripheral dysfunction was assessed by pulse amplitude tonometry (PAT) of the small digital artery. In particular, Log-transformed reactive hyperemia index (LnRHI) was evaluated using the EndoPAT2000 device: values of Ln-RHI < 0.51 were considered indicative of peripheral ED.
Results: At diagnosis, 23 subjects (76.7%) were ANCA positive (8 c-ANCA+, 15 pANCA+), and at the enrollment in the study 10 patients (33.3%) were still seropositive. Nineteen patients (63.3%) had inactive disease (BVAS=0), 7 (23.3%) were in low disease activity (1≤BVAS≤2) and 4 (13.3%) had an active disease (BVAS≥3). The presence of ED was documented in 9 AAV patients (30%). AAV patients with ED had higher C-reactive protein (CRP) values at diagnosis compared to subjects without ED (*p=0.05*). Moreover, patients with altered endothelial response were in higher percentage smokers (55.6%) and ANCA positive at enrollment (44.4%), compared to subjects with normal Ln-RHI (28.6%- p=0.12; 19%, p=0.15 respectively). There were no differences regarding age and other traditional CVRFs, disease duration, BMI, BVAS, VDI, FFS. There was an inverse correlation between CRP values at diagnosis and LnRHI (r=-0.42, *p=0.04*). In multiple logistic regression analysis, ANCA positivity at enrollment [OR (95% CI) = 15.68 (0.98-250.28)] (*p=0.05*), and higher CRP concentrations [OR (95% CI) = 1.03 (1.00–1.06)] (*p=0.03*) were independently associated with the presence of peripheral ED.
Conclusion: As observed in other chronic inflammatory autoimmune diseases, ED occurs in AAVs and is mainly related to the chronic systemic inflammation and seems to be also influenced by ANCA positivity, which is probably involved in the accelerated endothelial cell damage.
Further studies are needed to clarify the role of vasculitis related characteristics in the atherosclerotic process.
Disclosure of Interests: None declared
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AB0641 AN ULTRASONOGRAPHIC STUDY OF THE SALIVARY GLANDS IN A COHORT OF PATIENTS AFFECTED BY CRYOGLOBULINEMIA VASCULITIS: PRELIMINARY RESULTS

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Background: Xerostomia and xerophthalmia can occur in many diseases, rheumatological or not and are often reported by patients affected by cryoglobulinemia vasculitis[1], particularly if related to the Hepatitis C virus infection (HCV)[2]. On the other hand, the Ultrasound (US) was extensively used for detecting pathological findings of the salivary glands[3], possibly associated to many rheumatological diseases.
Objectives: The aim of this study was to detect, in patients affected by cryoglobulinemia vasculitis, the presence and the grade of pathological findings at level of the major salivary glands.
Methods: From January to December 2021, we enrolled consecutive oupatients patients affected by cryoglobulinemia (age ≥ 18 yo and a diagnosis of cryoglobulinemia vasculitis according to 2002 or 2011 classification criteria). Exclusion criteria were a previous diagnosis of other vasculitis/connective tissue diseases/inflammatory arthropathies, concomitant not rheumatological diseases or the intake of drugs possibly related to sicca syndrome symptoms. For each patient, the demographic, anthropometric and clinical history data, particularly about the cryoglobulinemia symptoms (either previous and ongoing) and past or current therapies have been collected. Furthermore, the values of erythrocyte sedimentation rate [ESR], C-reactive protein [CRP], Rheumatoid Factor [RF] and C4 serum complement fraction have been recorded. The presence and the titre of the cryoglobulins have been recorded. For each patient, the Schirmer's Test and the evaluation of the unstimulated whole salivary (UWS) flow were performed. The Schirmer Test was considered positive if < 5 mm/min at least in one eye and the UWS flow was defined pathological if < 1,5 ml/15 min. Each patient was submitted to the US examination (Esaote MyLab Twice) of both parotid and submandibular glands. Each gland was evaluated using a semiquantitative score 0-3, according to the OMERACT definitions[3]. The US score was considered pathological if ≥2. Thus, at patient level, a sum dichotomous score (normal/pathological>) was provided, defining it as "pathological" if any parotid or submandibular glands scored ≥ 2.
Results: We enrolled 20 patients, all females, with a mean age of 68.8 years (SD± 13.32) and a mean disease duration of 11.76 (SD±8.25). For 18 patients was reported a previous HCV infection. 16/18 HCV + subjects reached the remission of the