by Montgomery–Asberg Depression Rating Scale (MADRS) and Hamilton Anxiety Rating Scale (HAM-A).
Brain MRI including T1- weighted images, T2- weighted images and fluid-attenuated inversion-recovery images (FLAIR) was done in 44 (41,5%) BD patients.
Results: CD of mild to moderate severity were diagnosed in 82 (77,4%) of BD patients. The mechanical memory (50%) and attention deficit (80,5%) were the most frequent manifestations of CD, impairment of associative memory (31,7%) and logical thinking (36,6%) were less frequent. The presence of CD didn't depend on BD activity, severity and duration, as on patient's ethnicity, use of prednisone and immunosupressive agents. The frequency of neurological manifestations (headache, seizures, myelopathy, ataxia) did not differ significantly in patients with and without CD (28% vs 32%, p=0,44). The patients with CD were older (34,3±1,07 vs 29,0±2,14, p=0,006), more often had chronic/recurrent depressive disorders (84,1% vs 50,0%, p=0,001) of moderate severity (MADRS 16,1±0,74 vs 12,2±1,06, p=0,005), chronic stressful life events (91,5% vs 62,5%, p=0,001) and multifocal subcortical parenchymal MRI changes (57,6% vs 9%, p=0,005).
Conclusions: the results have shown high rates of different CD in BD patients. CD were not associated with BD activity and presence of neurological symptoms. CD were related to the diagnoses of stress-related mild to moderate chronic depressive disorders and minor brain multifocal subcortical parenchymal MRI lesions.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.3741

FRI0344 NOVEL BIOMARKERS OF SUBCLINICAL ENDOTHELIAL DYSFUNCTION IN BEHÇET'S SYNDROME: EVALUATION OF CROSS-SECTIONAL DISTENSIBILITY AND INTIMA-MEDIA-THICKNESS IN A HOSPITAL-BASED POPULATION

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Background: Growing interest exists on the role of markers of subclinical cardiovascular disease as independent predictors of cardiovascular events. Poor data are available on the role of these markers as prognostic factors for Behçet's syndrome (BS).
Objectives: The primary aim was to explore Intima-Media-Thickness (IMT), mean arterial diameter and distensibility (DC) in a group of patients with BS, comparing these data with a healthy control group and a disease control group; the secondary aim was to correlate the vascular parameters with demographic/clinical profile.
Methods: Thirty BS patients (females:12;mean age±SD:43±10.5; mean disease duration±SD:13±5.8) fulfilling the ISG criteria were prospective enrolled. Demographic data, level of disease activity, frequency of smokers, hypertension, family history of cardiovascular risk factors, body mass index (BMI) and current therapies were analysed. For each subject, ultrasound B-mode image sequences of right common carotid arteries were acquired and analysed by an automatic system (Carotid Studio,Quipu) for the measurement of IMT and mean arterial diameter. In addition, carotid pulse pressure (PP) was estimated by tonometry and DC coefficient was obtained. The systolic and diastolic carotid diameters were automatically measured on the distal wall of the common carotid artery, 1–2 cm beneath the bifurcation. Carotid diameter was calculated as the distance between media-adventitia interfaces. Cross-sectional DC was estimated through the variations in arterial cross-sectional area and blood pressure during systole. DC was computed as DC=ΔA/(PP*A) where A is the diastolic lumen area, ΔA is the stroke change in lumen area, and PP is the local pulse pressure.
Results: At time of evaluation, 4/17 patients presented active disease (50% ocular involvement, 25% joint involvement, 25% gastro-enteric involvement; mean BS activity score 5). Mean IMT±SD value resulted of 0.57±0.81, mean arterial diameter±SD value was 6.879±0.81 and mean DC± SD 27.3±14.34. All the vascular parameters considered were significant correlated with BMI, while only IMT and DC were also significant correlated with arterial hypertension. Using a correction analysis for age and sex, we found significant correlations between mean arterial diameter and disease activity and between DC and disease duration. These data resulted significantly different compared to healthy control and a disease control groups, in terms of smaller arterial diameter and higher DC.
Conclusions: Our data have shown that there is a consistent influence of disease activity and duration of disease on mean arterial diameter and DC, respectively. Thus, it is desirable that future pharmacological researches on BS target on this issue.
Acknowledgements:
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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.5907

FRI0345 CLINICAL IMPACT OF ALPHA-1-ANTITRYPSIN DEFICIENCY IN GRANULOMATOSIS WITH POLYANGIITIS

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Background: Deficiency in α-1-antitrypsin (AAT), which is the main proteinase 3 (PR3) inhibitor, is now recognized as a pathogenic factor in some cases of anti-PR3 anti-neutrophil cytoplasmic antibodies (ANCA) related granulomatosis with polyangiitis (GPA). However, the clinical impact of AAT deficiency remains poorly established in this setting.
Objectives: The purpose of our study was to describe the clinical phenotypes and outcomes of anti-PR3 GPA patients according to their AAT status.
Methods: A retrospective monocentric study carried out in Caen University Hospital led to identify anti-PR3 GPA patients, from 09/21/2011 to 06/10/2016. AAT dosage and phenotype (isoelectric focusing in agarose gel) were performed for all patients. Categorical variables were reported as percentages and compared using Chi^2 or Fisher's tests according to expected frequencies. Continuous variables were expressed as means and analysed using Student's t-test. Associations between survival, renal survival or relapse-free survival, and AAT phenotype were evaluated by the log-rank test. A p-value <0.05 was considered to be statistically significant.
Results: Among the 72 identified anti-PR3 GPA patients, 40 (56%) were male. Median age at diagnosis was 60.5 years old. Patients mainly had constitutional symptoms (51, 71%), pulmonary (52, 72%), ear, nose or throat (ENT) (49, 68%), rheumatologic (45, 63%), and renal (44, 61%) involvements. Median initial BVAS score was 38 (maximum score: 63). Twelve (17%) deaths and 33 (46%) relapsing patients were noted (median follow-up: 55 months). Forty-eight (67%) patients had MM phenotype, 10 (14%) MZ phenotype, 8 (11%) MS phenotype, 3 (4%) M-variant phenotype, 2 (3%) ZZ phenotype and 1 (1%) ZS phenotype. Allele frequencies of M, Z and S allele were 81, 10 and 6%, respectively. The whole patient cohort had the same immunosuppressant drug regimen.

Tab;e 1

	Z carriers (n=13)	Non-Z carriers (n=59)	p-value	Z or S carriers (n=21)	Non Z or S carriers (n=51)	p-value
Initial BVAS (median)	18	18	0.91	18	18	0.71
Death (%)	15	17	1	14	18	1
Age at diagnosis (years, median)	61	60	0.91	61	59	0.62
Relapse (%)	38	47	0.56	43	47	0.75
Intra-alveolar hemorrhage (%)	47	14	**0.004**	43	12	**0.009**
ENT (%)	92	63	**0.05**	67	69	0.88
AAT level (g/L, median)	1.07	1.65	**<0.001**	1.11	1.65	**<0.001**

Only intra-alveolar hemorrhage (IAH), that was more frequent in Z or S deficient allele patients, and ENT involvement, that was more frequent in those with Z allele, were significantly associated with AAT deficiency. Among the 13 patients carrying Z allele, 9 had normal AAT level, including 6 without any biologic inflammatory parameter at the time of AAT dosage. Global survival, renal survival or relapse-free survival were identical between Z carriers compared to non-Z carriers (respectively 0.77, 0.75 and 1), and between Z or S carriers compared to those without (respectively 0.91, 0.44 and 0.32).
Conclusions: This study confirms the epidemiological association of anti-PR3 GPA with AAT deficiency, and find an interesting higher risk of IAH in this setting. The identical prognosis of both subgroups should be related to other therapeutic added in IAH, including plasma exchanges. Prospective studies are required to specify these data and to assess the need for replacement therapy in AAT deficient patients.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.4500

FRI0346 RENAL INVOLVEMENT IN GRANULOMATOSIS WITH POLYANGIITIS

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Objectives: We assessed the frequency, clinical features and prognosis of renal involvement in granulomatosis with polyangiitis (GPA) and current treatment approaches.
Methods: We performed a retrospective analysis of 234 patients with GPA, diagnosed according to Chapel-Hill Consensus Conference 2012 classification, 81 male and 153 female, aged 53 (41; 62) years. 54 patients (23.1%) had localized GPA. 174 (74.4%) were ANCA positive. Median follow up was 61.5 (32; 105) months. 103 patients (44%) had a history of renal involvement. Frequencies of proteinuria, hematuria, hypertension, rapidly progressive glomerulonephritis (RPGN), acute kidney injury (AKI) and chronic kidney disease (CKD) grades were analyzed.
Results: In 25 patients (24.3% of 103) renal involvement developed at disease