Results: The levels of BAFF were significantly higher in group 3 (0.88±0.19 ng/ml) compared with group 1 (0.68±0.13 ng/ml, $p<0.05$), but did not distinguish from the group 2 (0.90±0.35). There were not significant differences in ESR between groups. ROC analysis indicated that the AUC for ESR is 0.63±0.10 ($p=0.17$) and for BAFF – 0.72±0.12 ($p<0.05$), which indicates fair capacity for BAFF differentiate groups of pts with low activity or remission of SNV and pts with severe SNV (sensitivity - 61.5%, specificity - 88.9%), while it was poor for ESR (sensitivity - 85.7%, specificity - 46.4%).
Conclusions: There are significant differences in levels of BAFF between pts with BVAS≥24 and pts with BVAS≤11. According to ROC analysis evaluation of serum BAFF level distinguishes pts with active SNV from pts with low activity or remission better than determination of ESR.
Disclosure of Interest: None declared
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FRI0341 CLINICO-PATHOLOGICAL DISCREPANCIES AND CAUSES OF DEATH IN TAKAYASU ARTERITIS: A RETROSPECTIVE ANALYSIS OF 60 FATAL CASES

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Background: Takayasu arteritis (TA) may present with a wide spectrum of symptoms common to other diseases leading to delayed or missed diagnosis.
Objectives: To investigate the rate of undiagnosed clinically cases of TA, and provide a detailed analysis of the wrong diagnoses and the underlying causes of death.
Methods: A retrospective clinicopathological analysis of sixty autopsy cases (52 males and 8 females aged 18 to 45 years) of TA over period of 11 years have been performed. The median age at disease onset was 25, 7 years.
Results: In 33 cases (55%), TA was not diagnosed during the clinical stages but only in autopsy. The most common incorrect clinical diagnosis was atherosclerosis of the aorta and its branches (celiac trunk, renal, mesenteric and iliac arteries) that has misdiagnosed in 14 cases (42.4%) of TA. TA was misdiagnosed as myocardial infarction/ischaemic heart disease in 5 (15.1%), perforated peptic ulcer in 3 (9.1%), polyarteritis nodosa in 3 (9.1%), and infective endocarditis in 2 cases (6.1%). There were other discrepancies in 6 cases (18.2%); in these cases, cerebral haemorrhage, rheumatic heart disease, pulmonary embolism, pheochromocytoma, chronic glomerulonephritis and lung cancer were the wrong clinical diagnoses. The leading position in the mortality structure due to TA belongs to septic shock that observed in 19 cases (31.7%) due to peritonitis/acute abdomen caused by mesenteric artery occlusion with subsequent intestinal necrosis in 12 (63.1%), lower limb gangrene in 6 (31.6%), and prosthetic aortic graft infection in 1 (5.3%). The second-leading cause of death was acute heart failure due to myocardial infarction, and renovascular arterial hypertension (25%). The third common cause of death was haemorrhagic shock in 14 cases (23.3%). The acute bleeding was caused by ruptured aortic aneurysm in abdominal part in 6 cases (42.8%) and ascending aorta with cardiac tamponade in 4 (28.6%). In 2 cases (14.3%), the source of hemorrhage was ulcers of gastrointestinal tract. The surgery has complicated by lethal bleeding in 2 cases (14.3%). In other patients, causes of death were cerebral haemorrhage in 6 (10%), renal failure in 3 (5%), respiratory failure in 1 (1.7%), and revascularization syndrome in 1 (1.7%). In one case, TA coexisted with scleroma of larynx, and asphyxia was direct cause of death.
Conclusions: Our date show a high number of cases of TA (55%) that were identified at autopsy but were not diagnosed clinically. It can be assumed that aortic atherosclerosis, myocardial infarction/ischaemic heart disease were the most common wrong diagnoses in TA. The leading causes of death in TA are septic shock, acute heart failure, and haemorrhagic shock. In addition, it should be noted that in our series of autopsy cases of TA, males (86, 7%) were most frequently affected compared with results other investigations. Interestingly, that the correct diagnosis of TA was established before death in all females' cases. It seems to be a trend to miss the clinical diagnosis of TA in male patients.
Disclosure of Interest: None declared
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FRI0342 DISTRIBUTION OF VASCULITIDES IN EASTERN MEDITERRANEAN: RESULTS OF A PROSPECTIVE COHORT

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Background: The ethnic and geographical differences have been commented on, mainly from European data. We lack large data sets from the eastern part of Europe.
Objectives: This study is aimed to present the distribution of vasculitides in the Pediatric and Adult Vasculitis patient groups.
Methods: Hacettepe University is one of the main referral centers in the capital of Turkey, serving approximately 470,000 adult and pediatric patients/a year. Hacettepe University Vasculitis Centre (HUVAC) was established to organize a prospective cohort in 2014. All patients starting from October 2014 have been recorded to the database. Additionally electronic patient recording database between October 2014-December 2016 was searched for the patients having any of the 10th revision of the International Statistical Classification of Diseases (ICD)-10 code for the particular vasculitis. The study group of Hacettepe University Vasculitis Centre (HUVAC) re-evaluated the ascertained patients' hospital records to review their diagnosis according to the 2012 revised Chapel Hill nomenclature criteria.
Results: A total of 1196 patients had been newly followed in this period. 271 (22.7%) of them were pediatric patients and 31.8% of them were newly diagnosed. The leading vasculitis among adult patients was Behcet's Disease whereas in pediatric patients it was HSP/IgA Vasculitis (Table). Granulomatous polyangiitis was the most common small vessel vasculitis in adults. Takayasu arteritis was more frequent than giant cell arteritis among the adult patients. There was a female predominance in patients with large vessel vasculitis.
During prospective follow up, 22 (1.9%) patients deceased; 7 due to the primary disease, 6 due to infections, 7 due to cardio and cerebrovascular diseases.

Table. Distribution and gender of Adult and Paediatric Vasculitides Patients

Adult Patients (n=925)		
Type	Frequency,%	Female, %
Behcet's Disease	50.5	53.7
ANCA Associated Vasculitis	11.9	46.4
• GPA	• 8.9	47.9
• EGPA	• 2.2	52.3
• MPA	• 0.8	57.1
Takayasu's Arteritis	10.7	90
IgA Vasculitis	4.3	47.5
Giant Cell Arteritis	3.8	71.4
Cutaneous Vasculitis	3.2	70
Polyarteritis Nodosa	3.1	44.8
IgG4 Related Disease	2.8	48.2
CTD-Secondary Vasculitis	2.3	71.4
Polimyalgia Rheumatica	2.2	95
Primary CNS Vasculitis	0.8	42.8
Isolated Aortitis	0.8	50
Cryoglobulinemic Vasculitis	0.4	25
Others	3.2	57.3
Paediatric patients (n=271)		
Type	Frequency,%	Female, %
IgA Vasculitis	46.5	45.6
Behcet's Disease	22.5	45.9
Kawasaki Disease	17.3	45.6
Polyarteritis Nodosa	3.7	50.0
Takayasu's Arteritis	2.6	100
ANCA Associated Vasculitis	1.5	75
Vasculopathies (SAVI, DADA2)	1.5	37.5
Others	4.4	52.3

Conclusions: Behcet's Disease is the most frequent vasculitis in Turkey and more than half of the patients had organ/system involvements. GPA is the most common small vessel vasculitis whereas Microscopic polyangiitis is very rare. Among children Kawasaki was less and Behçet's disease markedly more when compared to Western European figures. Even though this study is a single-center one, the number of annual admitted patients of our hospital along with more than 2 years-prospective recruitment, reflects the true burden of vasculitides in Turkey, at the cross-roads between Europe and Middle East.
Disclosure of Interest: None declared
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FRI0343 COGNITIVE DISORDERS IN BEHÇET'S DISEASE: SOME CLINICAL AND PATHOGENESIS RELATIONS

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Background: the cognitive disorders (CD) is a special psychopathological problem for Behçet's Disease (BD) patients. The causes of CD aren't enough investigated. Anxiety/depressive disorders and primary neural parenchymal lesions due to BD usually related to CD in these patients.
Objectives: to determine the prevalence of CD in BD patients and its associations with some clinical and magnetic resonance imaging (MRI) scan features.
Methods: the investigation has been realized in accordance with the inter-disciplinary program "Stress factors and mental disorders in immune-mediated inflammatory rheumatic diseases".
106 BD patients were enrolled in the study. The majority of patients were men (72,6%), natives of the North Caucasus (51,9%), with mean age (M±m) 33,3±0,98 years. All the patients met the criteria of the International Study Group for BD (1990) classification. The disease activity was assessed by scoring system BDCAF.
CD were diagnosed with psychology and neuropsychology methods. Mental disorders (MD) were diagnosed by psychiatrist in accordance with the ICD-10 in semi-structured interview. The severity of depression and anxiety was evaluated

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
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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.
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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.
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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