onset, the other 78 patients (75.7%) developed glomerulonephritis after a median of 3 (1; 12) months. Hematuria was present in 94 (91.3%), proteinuria in 85 patients (82.5%), nephrotic syndrome in 11 patients (10.7%), hypertension in 31 patients (30.1%). Rapidly progressive glomerulonephritis (RPGN) defined by the doubling of serum creatinine within ≤3 months developed in 29 patients (28.2% of 103). Myeloperoxidase-ANCA-positive patients (n=12) developed RPGN significantly more often than proteinase-3-ANCA-positive patients (n=78): 7 (58.3%) vs 22 (28.2%) (p=0.0499). 11 (10.7%) patients developed AKI, stage 3 being the most common (in 8 patients). 40 (38.8%) patients exhibited indolent renal disease course, i.e. without worsening of renal function. By the end of the follow-up, 33 (32.0%) patients were diagnosed with CKD grade 3b-5, among them 11 patients (10.7%) developed end-stage renal disease (ESRD). 6 patients (5.8%) died. As the first line induction therapy all patients received corticosteroids in combination with cyclophosphamide in 93 patients (90.3%), rituximab (RTX) in 3 patients (2.9%), mycophenolate mofetil in 7 patients (6.8%). RTX was used for remission induction in 27 (26.2%) patients due to refractory or recurrent disease. During follow-up, 74 patients (71.8%) developed one or more severe relapses with a relapse rate of 0.27 per patient-year. 50 (48.5%) patients had renal relapses (0.096 per patient-year).

**Conclusions:** Prevalence of renal involvement in the studied group was lower than expected, most likely because of a large proportion of patients with localized GPA. Our study showed that RPGN was not the most common feature of renal involvement. Almost 40% of patients developed indolent course of kidney disease. Nevertheless one third of patients developed CKD G3B or worse by the end of the follow up. Despite effective induction therapy with high-dose corticosteroids, cytotoxic agents and/or RTX relapses, both renal and extrarenal, are frequent.

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## FRI0347 RHEUMATOID FACTOR IS CORRELATED WITH DISEASE ACTIVITY AND INFLAMMATOTY MARKERS IN ANTINEUTROPHIL CYTOPLASMIC ANTIBOTY-ASSOCIATED VASCULITIS

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**Background:** Raised levels of serum rheumatoid factors (RFs) of different immunoglobulin classes had been reported in a high proportion of patients with rheumatoid vasculitis. RF used to be studied and was shown elevated in some forms of vasculitides. Patients with antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) are often positive for RF. However, the clinical significance of RF has been seldom examined in AAV.

**Objectives:** The aim of this study was to investigate association between the presence of RF and clinical features and outcomes in AAV.

**Methods:** Eighty-one patients were diagnosed with AAV from 2006 to 2015 in our hospital. Among 81 patients, forty-seven patients (17 males, median age 67 years) who were not complicated with rheumatoid arthritis and in whom RF was measured before the treatment, were studied, retrospectively. Patients were classified using the European Medicines Agency vasculitis classification algorithm. AAV included eosinophilic granulomatosis with polyangitis (n=10), granulomatosis with polyangitis (n=14), microscopic polyangitis (n=16) and unclassifiable vasculitis (UV) (n=7). Patients with UV with MPO-ANCA or PR3-ANCA were included in this study. IgM-RF was measured using a latex agglutination assay. Disease activity was assessed with Birmingham vasculitis activity score (BVAS). IgM-RF, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), serum ferritin, IgG, IgM, IgA, MPO-ANCA and PR3-ANCA were obtained from hospital records. Clinical manifestations between RF-positive subset (n=29) and RF-negative subset (n=18) were analyzed using Fisher's exact and Wilcoxon rank sum tests. Correlation coefficients were established with Spearman's correlation coefficient. Data were shown as medians (interquartile range).

**Results:** BVAS was higher (14 (12–22) vs 12 (6–16), P=0.026) in the RF-positive subset than the RF-negative subset. CRP and ESR were higher (P=0.020 and 0.007, respectively) in the RF-positive patients. IgM-RF titers significantly correlated with BVAS (r=0.50, P=0.0004). In addition, CRP, ESR, IgM and IgG also had a significant correlation with IgM-RF titers. The frequency of initiation of dialysis therapy (14% vs 6%), usage of mechanical ventilation (14% vs 0%) and mortality (10% vs 0%) were higher in the RF-positive subset than in the RF-negative subset although no significant differences were shown.

**Conclusions:** In AAV, IgM-RF titers are significantly correlated with disease activity and inflammatory markers. Presence of RF could be a poor prognostic factor in patients with AAV.

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## FRI0348 ASSOCIATED INFLAMMATORY SYNDROMES IN TAKAYASU'S ARTERITIS: MANY FACES OF A DISEASE

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**Background:** Case reports and series suggest that Takayasu's arteritis (TA) can co-exist with various inflammatory disorders.

**Objectives:** We conducted a formal study to look specifically at the frequency of inflammatory disorders and symptoms in a large cohort of TA followed by a single tertiary center.

**Methods:** There were 238 patients registered with a diagnosis of TA. Of these, 19 died, 18 were lost to follow-up and 3 did not wish to response our questionnaire. The remaining 198 patients were called back at the outpatient clinic. A standardized form sought whether the patient was also diagnosed as inflammatory bowel disease (IBD), ankylosing spondylitis (AS), Behçet's syndrome (BS), amyloidosis, uveitis, rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), systemic sclerosis (pSS), Sjögren syndrome, psoriatic arthritis, inflammatory myositis, small vessel vasculitis, autoimmune/demyelinating or any other inflammatory disorder.

**Results:** Overall, 198 (175 F/ 23 M) patients were studied. The mean age at the time of TA diagnosis was 34±12 years. Subclavian artery was the most common involved artery (84%), followed by common carotids (78%) and aorta (65%). Currently, while 25 (16%) patients were off treatment, 72 (47%) patients were using glucocorticoids, 47 (31%) azathioprine, 32 (21%) methotrexate and 44 (29%) biological agents.

We identified in total 37 (19%) patients with inflammatory diseases (IBD: n=12; AS: n=15; and BS: n=10). Table shows their demographic characteristics. Among the remaining 161 patients, the most frequent feature was inflammatory back pain (36%), followed by recurrent oral ulcers (15%), erythema nodosum (11%), arthritis (10%), papulopustular lesions (7%), uveitis (4%), and genital ulcer (1%). It was noted that inflammatory back was mostly located on the dorsal area. Regarding autoimmune diseases, we also observed RA (n=3), psoriasis (n=2), autoimmune hepatitis (n=2), SS (n=1) and SSc (n=1).

Table 1. Demographic features of 37 TA patients with inflammatory bowel disease (IBD), ankylosing spondylitis (AS) or Behçet's syndrome (BS)

| Concomitant disease | F/M | Mean age at TA diagnosis (SD) | Mean age at concomitant disease (SD) | Time of TA diagnosis in relation to concomitant disease |
|---|---|---|---|---|
| AS (n=15) | 13/2 | 31±8 | 27±11 | Simultaneous (n=3)<br>TA preceded (n=7)<br>AS preceded (n=5) |
| IBD (n=12) | 11/1 | 33±9 | 31±9 | Simultaneous (n=9)<br>TA preceded (n=1)<br>IBD preceded (n=2) |
| BS (n=10) | 7/3 | 35±13 | 32±13 | Simultaneous (n=5)<br>TA preceded (n=1)<br>BS preceded (n=4) |

**Conclusions:** TA does co-occur with IBD, AS or BS in about 1/5 of the patients, at least in a hospital setting and without a clear temporal pattern. This could be due to the close association of TA with MHC class-1 diseases. In addition, the high prevalence of inflammatory back pain in the dorsal spine in TA needs further scrutiny.

**Disclosure of Interest:** None declared

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## FRI0349 THE SNP RS6871626 LOCATED IN IL12B REGION MAY INFLUENCE ON VASCULAR LESIONS OF TAKAYASU ARTERITIS

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**Background:** Takayasu arteritis (TAK) is a type of large vessel vasculitis, which affects aorta and its main branches. We previously found a single nucleotide polymorphism (SNP), rs6871626 located in *IL12B* region, as a susceptible gene to TAK and reported that the risk allele at the SNP is associated with the risk of aortic regurgitation (AR)[1)]. However, there have been no studies on the association of the SNP with the organ involvements other than AR in TAK patients.

**Objectives:** To investigate the association of the SNP with arterial and organ involvements except for AR.

**Methods:** We examined the medical records of 85 patients with TAK, stratified

**Abstract FRI0349** – Table 1. Organ involvements in patients

| Organ involvements | AA | AC | CC | p-value | p-values of multiple comparisons† | | |
|---|---|---|---|---|---|---|---|
| | | | | | AA vs. AC | AA vs. CC | AC vs. CC |
| Carotid arterial lesion | 78.9% (15/19) | 63.9% (23/36) | 80.0% (8/10) | 0.48* | 1.00 | 1.00 | 1.00 |
| Descending aortic lesion | 75.0% (18/24) | 44.2% (19/43) | 25.0% (4/16) | 0.0048* | 0.064 | 0.0096 | 0.71 |
| eGFR (ml/min/1.73m$^2$) | 61.3 ± 26.9 | 72.9 ± 22.8 | 81.5 ± 28.8 | 0.011** | 0.21 | 0.042 | 0.75 |

Mean ± SD, *Fisher's exact test, **one-way ANOVA, †p-value is corrected by Bonferroni's method.

them into three groups according to the allele at the SNP, AA (n=26), AC (n=43) and CC (n=16) (A is a risk allele), and investigated the association of the SNP and organ involvements.

**Results:** There were no differences in the complication rates of carotid arterial lesions among the groups (AA 78.9%, AC 63.9% and CC 80.0%). The proportion of patients with lesions in descending aorta (Numano classification[2] IIb∼V) was 75.0% in AA, 44.2% in AC 44.2% and 25.0% in CC and the proportion in AA was significantly higher than in CC (p=0.0096). Moreover, estimated glomerular filtration rate (eGFR) was significantly lower in AA than in CC (61.3±26.9ml/min/1.73m$^2$ vs. 81.5±28.8 ml/min/1.73m$^2$, p=0.042).

**Conclusions:** The SNP rs6871626 located in *IL12B* may influence on the occurrence of descending aortic lesions in TAK patients and this may lead to renal dysfunction.

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## FRI0350 INVESTIGATING THE LINK BETWEEN ISCHEMIC HEART DISEASE AND BEHCET'S DISEASE: A CROSS-SECTIONAL ANALYSIS

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**Background:** Behcet's disease (BD) is a multi-systemic chronic inflammatory disorder which involves the vasculature tree of all sizes. Subclinical endothelial injury as a result of chronic inflammation in BD has been associated with thrombosis and arterial stiffness, risk factors for ischemic heart disease (IHD)[1–4]. However, data regarding the association between BD and IHD and the impact of various risk factors on this association, is scarce[3–5].

**Objectives:** To examine the association between BD and the development of IHD and its potential impact as a cardiovascular risk factor.

**Methods:** This study was conducted as a case-control study utilizing the database of Clalit Health Services. The proportion of IHD was compared between patients diagnosed with BD and age- and gender-matched controls. Univariate analysis was performed using Chi-square and student *t-test* and a multivariate analysis was performed using a logistic regression model.

**Results:** 871 BD patients and 4,439 age- and gender-matched controls were included in this study. The proportion of IHD amongst BD patients was increased in comparison with controls (10.9% vs. 7.52% respectively, *p*-value =0.001). After controlling for cofounders, BD was associated with IHD on multivariate analysis (odds ratio 1.485, 95% confidence interval 1.106–1.982, Table 1). When the comparison between BD patients with IHD and controls with IHD was stratified according to risk factors, age younger than 70 and male gender were both found to be significantly elevated in the BD group.

Table 1. Multivariate logistic regression for IHD (*n*=5,220)

| Variable | OR | 95% CI | *p*-value |
|---|---|---|---|
| Behcet | 1.485 | 1.106–1.982 | 0.008 |
| Age | 1.059 | 1.047–1.071 | <0.001 |
| Male gender | 2.929 | 2.248–3.841 | <0.001 |
| BMI | 1.010 | 0.987–1.034 | 0.388 |
| High SES *vs.* low | 0.709 | 0.503–0.994 | 0.048 |
| Intermediate SES *vs.* low | 0.793 | 0.600–1.048 | 0.104 |
| Arab ethnicity | 0.872 | 0.618–1.221 | 0.430 |
| Hypertension | 3.050 | 2.332–4.010 | <0.001 |
| Hyperlipidemia | 7.895 | 5.119–12.781 | <0.001 |
| Diabetes | 1.624 | 1.268–2.078 | <0.001 |
| Smoking | 1.849 | 1.445–2.371 | <0.001 |

IHD = ischemic heart disease, OR = odds ratio, CI = confidence interval, BMI = body mass index, SES = socioeconomic status.

**Conclusions:** BD patients have an increased proportion of IHD in comparison with matched controls. The population of BD patients with IHD has a higher proportion of younger males in comparison with controls with IHD. Preemptive screening for IHD may be warranted in this subgroup.

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## FRI0351 PAPULOPUSTULAR LESIONS ACCORDING TO AGE, SEX AND LOCALIZATION IN BEHCET'S SYNDROME PATIENTS COMPARED HEALTHY AND DISEASED CONTROLS

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**Background:** Papulopustular lesions (PPL) are the most common skin lesions in Behçet's syndrome (BS).

**Objectives:** To assess whether PPL are different in BS according to localization, age, sex and medications used when compared to rheumatoid arthritis patients (RA) and apparently healthy subjects (HS).

**Methods:** 209 consecutive BS patients who were routinely followed in our dedicated BS center were studied. Patients with RA (n=146) who were followed up in the rheumatology outpatient clinic of the same unit and HS (n=149) were used as controls. All subjects were clinically evaluated by the same dermatologist and all skin lesions (papules, pustules, comedones, folliculitis, cysts, nodules) on the face, trunk and legs were separately counted. Information regarding the demographic and clinical features of primary disease and medications used were obtained from patients' charts.

**Results:** Subjects without PPL were excluded before analyses. Demographic features and mean number of PPL according to site of body were summarized in Table-1. Mean number of total PPL were similar between BS and HS and significantly higher than in RA (p<0.001). Mean number of total PPL according to sex were similar in RA and HS but higher in male BS patients compared to female BS patients (p=0.04). When we analyzed the number of PPL according to different body sites, we observed that BS patients had significantly more lesions on the legs when compared to the RA patients and HS (p<0.0001). Number of PPL lesions tend to decrease as the patient ages in BS similar to RA and HS. When leg lesions were analyzed according to age, this difference remained in the age groups 31–50 and >50 but not in the age group ≤30. Corticosteroid use did not impact the results.

Table 1

| | BS | RA | HS |
|---|---|---|---|
| N | 155 (74%) | 57 (39%)* | 101 (68%) |
| F/M | 95/60 | 46/11 | 52/49 |
| Mean age (SD) | 38.6 (10.0) | 47.5 (13)† | 34.7 (12.0) |
| N of pts on corticosteroids | 24 (15 M/9 F) (15%) | 37 (9 M/28F) (65%) | – |
| Mean n of PPL (SD) | 7.8 (8.0) | 3.9 (3.8)‡ | 9.7 (9.5) |
| Trunk | 107 (58F/49M) (70%) | 36 (28F/8M) (63%) | 81 (37F/44M) (80%) |
| Face | 120 (80F/40M) (77%) | 37 (32F/5M) (65%) | 84 (50F/34M) (83%) |
| Legs | 48 (21F/27M)§ (31%) | 2 (1F/1M) (3.5%) | 13 (5F/8M) (13%) |

*p<0.001, †p<0.02, ‡p<0.05, §p<0.0001.

**Conclusions:** BS patients have significantly more PPL on the legs when compared to HS and RA. Number of PPL tend to decrease as the patient ages in BS similar to RA and HS however BS patients still have more PPL on the legs after the age of 50 suggesting that these lesions somehow differ from acne vulgaris, in pathogenesis.

**Disclosure of Interest:** None declared

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