symptoms (see table 1). Four patients failed to improve at all. Persistent remission of the arthritis/arthralgia was found in 41 patients (85%). One patient did not improve. Complete resolution of the urinary abnormalities were registered in 14 patients (40%), improvement – in 17 cases (48.5%), further deterioration in 4 patients (11.5%).

There were side effects in 13 patients (16%). In 4 (5.1%) cases there were headache and nausea, respectively. In 2 (2.5%) cases treatment was accompanied with skin itching and urticarial rash. Most severe complication presented with reversible cytotoxicity, characterised with increase of AST and ALT up to the 2–5 normal limits in 3 cases (2.5%).

Abstract FRI0483 – Table 1. Clinical response to SASP treatment in primary IgA-vasculitis (n=78)

Clinical features	Complete remission		Partial remission		No response		Worsening	
	n	%	n	%	n	%	n	%
Skin purpura (n=78)	46	58.9	27	35	4	5.1	-	-
arthritis or arthralgia (n=48)	41	85	6	13	1	1.2	-	-
hematuria and/or proteinuria (n=35)	14	40	17	48.5	-	-	4	11.5

Conclusions: To our knowledge, this is the first experience of SASP in treatment of primary IgA-vasculitis not reported before. Preliminary results look promising and worthy of further evaluation.

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FRI0484 OVERALL SURVIVAL AND MORTALITY RISK FACTORS IN TAKAYASU'S ARTERITIS: A MULTICENTER STUDY OF 318 PATIENTS

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Background: Takayasu arteritis (TA) is a rare large vessel vasculitis of unknown origin. Main epidemiological data come from Asia. TA significantly increases morbidity and mortality and there is an unmet need to identify patients with higher complications and mortality risks.

Objectives: To report the long term mortality in Takayasu arteritis (TA) and to identify risk factors associated with mortality.

Methods: We analysed epidemiological data according to gender and ethnic origins, the causes of death, and the factors associated with mortality in a cohort of 318 patients [median age at diagnosis was 36^{25–47} years and 276 (86%) patients were women] fulfilling American College of Rheumatology and Ishikawa criteria of TA. Overall survival and factors associated with death and vascular complications were identified. A prognostic score for death and vascular complications was elaborated based on a multivariate model.

Results: In a cohort of 318 patients, men tended to be more frequently smokers (42% vs. 26%, p=0.07), had more frequently cardiac insufficiency (14% vs. 9%, p=0.034), but less frequently aneurysmal lesion (51% vs. 76%, p=0.005). Ethnic origins were Caucasian/White (n=87, 37%), North-Africa (n=73, 31%), Black (n=56, 18%), and other (n=17, 5%). White patients were more frequently smokers and diagnosis was more frequently made after 35 years. Black patients had less frequently a supra-aortic vessels involvement and more frequently a vessel wall thickening at diagnosis (78% vs. 62%, p=0.033).

Among 318 TA patients, 16 (5%) died after a median follow-up of 6.1 years. The mean (±SD) age at death was 38^{25–47} years with 88% of women. Main causes of death included mesenteric ischemia (n=4, 25%), aortic aneurysm rupture (n=4, 25%), sudden death (n=3, 19%), septic shock (n=3, 19%), pulmonary embolism (n=1, 6%), and cardiogenic shock (n=1, 6%). One, five, and ten years' overall survival were 98.9% [97.8–100], 98.1% [96.4–99.8], and 96.1% [93.4–98.9%], respectively. Factors associated with mortality were ethnic origin (White 9.5% vs. North Africa 7.1% vs. Black 0%, p=0.049), and tobacco smokers (10.8% vs. 4.2%, p=0.002).

We define high risk patients for death and vascular complications according to the presence of two of the following factors (i.e a progressive clinical course, thoracic aorta involvement and/or retinopathy) elaborated based on the multivariate model. The probability of death and complication free survival at five years was 78.4% (69.4–88.6) and 51.5% (38.3–69.2) (p=0.001) in the low risk and high risk group, respectively.

Conclusions: The overall mortality in our Takayasu cohort was 5% after a median follow-up of 6.1 years. Caucasian and tobacco smokers were associated with mortality in TA. We developed a simple and useful prognosis score to identify patients at risk for vascular complication and death.

Disclosure of Interest: None declared

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FRI0485 PHENOTYPE OF ANCA ASSOCIATED VASCULITIS ASSOCIATES WITH MAJOR CARDIOVASCULAR EVENTS: A RETROSPECTIVE OBSERVATIONAL STUDY FROM THE LEEDS VASCULITIS COHORT

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Background: Patients with ANCA-associated vasculitides (AAV) are at increased risk of cardiovascular (CV) disease than the general population. The factors associated with CV involvement in AAV are poorly understood.

Objectives: To explore whether patient and disease-related characteristics associated with CV disease from a single-centre AAV cohort.

Methods: Medical records from 145 patients with AAV were reviewed and patients' diagnoses reclassified according to a validated algorithm.¹ CV events (CVE) were defined as myocardial infarction, cerebrovascular accident, thromboembolic event, heart failure, or death/hospitalisation due to CV cause. Time to first CVE was calculated; patients without CVE were censored at most recent clinical follow-up. Data on CV risk factors (hypertension, hyperlipidaemia, diabetes mellitus, chronic kidney disease stage III or higher) were collected. Putative predictors included age, sex, diagnosis (GPA, MPA, EGPA, unclassifiable AAV – UAAV), ANCA (absent, PR3, MPO), number of BVAS items, prior CV disease and number of CV risk factors. Cox proportional hazards regression was used to assess the relationship between CVE and predictors. In this exploratory analysis we used p<0.10 to indicate potential associations.

Results: 122 patients who had been followed up for between 6 months and 23 years were included in the analysis. 11/122 patients (10%) had prior history of CV disease. 17 patients (14%) experienced a CVE within median (IQR) 5.8 (1.4, 7.6) years of diagnosis. Univariable analyses indicated older age at diagnosis, MPA and prior CVD were associated with increased risk of CVE, and that those with UAAV were at decreased risk compared to GPA. With the exception of previous CVD an adjusted model confirmed the same associations (table 1).

Variable	No CVE n=105	CVE n=17	Unadjusted HR (90% CI)	Adjusted HR (90% CI)	P value
Age: mean (SD)	49 (17)	57 (13)	1.04 (1.01–1.07)	1.03 (1.00–1.06)	0.049
Female:% (n)	55% (58)	41% ⁷	0.76 (0.34–1.67)	0.69 (0.28–1.64)	0.488
Diagnosis:% (n)					
GPA	54% (57)	65% ⁷	Ref.	Ref.	0.370
MPA	13% ¹⁴	6% ¹	0.71 (0.12–2.44)	0.38 (0.05–2.21)	0.031
EGPA	10% ¹⁰	29% ⁵	3.37 (1.34–7.76)	4.81 (1.46–15.36)	0.071
UAAV	23% ²⁴	- (0)	0.15 (0.00–0.91)	0.09 (0.00–0.83)	
ANCA:% (n)					
Absent	25% ²⁶	12% ²	Ref.	Ref.	0.259
PR3	45% ⁴⁷	65% ¹¹	1.65 (0.58–6.25)	0.33 (0.08–1.79)	0.207
MPO	30% ³²	24% ⁴	1.62 (0.46–6.77)	0.27 (0.05–1.52)	
BVAS items: median (IQR)	4 (3–5)	4 (4–5)	1.26 (0.96–1.65)	1.35 (0.95–1.92)	0.156
Prior CVD:% (n)	10% ¹¹	18% ³	1.28 (1.17–9.23)	2.58 (0.72–8.03)	0.211
CV risk factors: median (IQR)	1 (0–2)	1 (0–2)	0.92 (0.59–1.39)	0.64 (0.36–1.04)	0.135

Conclusions: This exploratory analysis suggests that, among AAV, MPA may have a higher CV risk, whereas UAAV may be associated with a decreased risk. Along with ANCA status, AAV phenotype may inform future CV risk reduction interventions in AAV.

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FRI0486

POTENTIAL PREDICTORS OF VISCERAL INVOLVEMENT IN ADULT IGA VASCULITIS

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Background: Predictors of severity of visceral involvement in acute adult IgA vasculitis (IgAV) are poorly recognised.

Objectives: The aim of our study was to evaluate the role of smoking and extension of skin lesions on the visceral manifestations of acute adult IgA vasculitis.

Methods: We analysed medical records of adult, histologically proven IgAV cases, diagnosed at our secondary/tertiary rheumatology centre between 1 January 2010 and 31 December 2017. Purpura was defined as generalised when skin lesions extended above the waistline. Gastrointestinal (GI) disease was considered severe in case of bloody diarrhoea, ileus or bowel perforation. Renal disease was defined as severe when nephritic syndrome with acute renal failure or nephrotic syndrome developed.

Results: During the study period we identified 230 incident IgAV cases (57.8% males, median (IQR) age 64.8 (45.6–77.3) years). Ninety-eight (42.6%) patients were smokers (56 past and 42 current). Skin, joint, GI, renal and involvement were present in 230 (generalised purpura in 114 (49.6%), necrotizing in 108 (47.0%), 93 (40.4%), 70 (30.4%; severe in 17) and 102 (44.3%; severe in 27) patients, respectively. Smoking was associated with renal disease (RR 1.3 (95% CI 1.0–1.8)) and its severity (RR 3.2 (95% CI 1.5–7.0)), but not with GI involvement or its severity. Generalised purpura was associated with GI involvement (RR 2.9 (95% CI 1.8–4.7) and its severity (RR 3.3 (95% CI 1.1–9.8)), as well as with renal involvement (RR 1.4 (95% CI 1.0–1.9)). Data of combined influence of smoking and purpura extension on visceral involvement are presented in table 1. The risk of severe renal involvement in IgAV was the highest in ever-smoker with generalised purpura (RR 8.1 (95% CI 1.9–34.7) in comparison to IgAV non-smoker with localised purpura).

Table 1 The influence of smoking and purpura extension on visceral involvement in IgAV

Visceral involvement	Nonsmokers with localized purpura	Ever-smokers with localized purpura	Nonsmokers with generalized purpura	Ever-smokers with generalized purpura
Number of cases	69	47	63	51
GI (%)	13.0	19.1	47.6	43.1
Severe GI (%)	2.9	4.3	11.1	11.8
Renal (%)	30.4	46.8	47.6	56.9
Severe renal (%)	2.9	14.9	9.5	23.5

Legend: GI gastrointestinal; severe GI involvement: bloody diarrhea or ileus or bowel perforation; severe renal involvement: nephritic syndrome with acute renal failure or nephrotic syndrome;

Conclusions: Smoking and generalised purpura were associated with visceral involvement in adult IgAV.

Disclosure of Interest: None declared

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FRI0487

SERUM INTERLEUKIN-6 LEVELS IN ANTINEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS

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Background: The deregulated overproduction of interleukin (IL)–6 has been implicated in several inflammatory and antibody-mediated autoimmune diseases.

Objectives: To investigate serum IL-6 levels (sIL-6) during active disease, remission, and relapse in antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV), and to explore the association of changes in sIL-6 with repopulation of blood B cells and disease relapse.

Methods: sIL-6 levels (explanatory variable) were measured longitudinally over 18 months in 78 patients with AAV enrolled in a prospective, double-blinded, randomised, control trial comparing treatment with rituximab (RTX) (n=45) or cyclophosphamide (CYC)/azathioprine (AZA) (n=33). Outcome variables included baseline clinical features, ANCA type and titers, disease activity (status of active disease versus complete remission (CR)), time to B cell repopulation, relapse and severe relapse.

Results: Baseline sIL6 levels were detectable (>0.49 pg/ml) in 81% of patients. At baseline, sIL-6 positively correlated with proteinase (PR3)-ANCA levels ($r_s=0.36$, $p<0.01$), but not with myeloperoxidase (MPO)-ANCA levels ($r_s=-0.17$, $p=0.47$). Higher baseline sIL-6 levels were associated with the presence of fever, pulmonary nodules/cavities, PR3-ANCA, and absence of renal involvement ($p<0.05$ for all comparisons).

The median sIL-6 level was higher at baseline and promptly declined with induction therapy at following time-points (baseline, median [25%–75% IQR], 2.66 [0.76–20.98]; month 6th, 0.49 [0.49–1.16]; $p<0.01$) in a similar fashion in both treatment arms.

An increase in sIL-6 during clinical remission was a predictor for subsequent severe relapse in RTX-treated patients (Hazard Ratio (HR) 7.24, $p=0.01$), but not in CYC/AZA-treated patients (HR 0.62, $p=0.50$).

In RTX-treated B cell depleted patients (CD19 +B cell/microliter<10), the rise of sIL-6 levels did not precede B cell reappearance, regardless of whether a cut-off of ≥ 10 CD19+B cell/microliter (HR=0.97; $p=0.97$) or of ≥ 69 CD19+B cell/microliter (HR=1.50; $p=0.41$) was considered. Eighty percent of the patients who subsequently had a severe relapse in the RTX arm had B cell redetection before or at the time of the IL-6 increase, with a mean time between the redetection of B cells and sIL-6 increase of 55 days (25%–75% IQR: 0–184.25; range 0–210 days).

Conclusions: At baseline, sIL-6 correlates with PR3-ANCA titers and associates with the presence of fever and pulmonary nodules/cavities. A rise in sIL-6 levels after complete remission is associated with subsequent severe relapse only in RTX-treated patients. The observed discrepancies between treatments deserve confirmation and further study.

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FRI0488

INSUFFICIENT IMMUNOSUPPRESSIVE USE IS THE LEADING CAUSE OF VASCULAR RELAPSES IN BEHCET'S DISEASE

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Background: Vascular involvement is observed in up to 40% in Behcet Disease (BD) patients, as an important cause of mortality and morbidity, especially for males (Kural-Syahi E et al,1984).

Objectives: Purpose of this study is to describe clinical-demographic properties, treatments and prognosis of vascular BD patients in a tertiary rheumatology clinic.

Methods: BD patients fulfilling ISG 1990 criteria are recruited from the multi-disciplinary Behcet's Clinic in Marmara University, Istanbul for this retrospective study. All data is collected from patient files (ISG for BD, 1990).

Results: Mean age of BD patients (M/F:102/22) was 29.3±7.3 years at diagnosis and 32.4±9.5 years old during first vascular event. Median follow up was 47^{17–71} months. Mean age of female patients was significantly older during first vascular event (table 1). 73.2% of vascular involvement was venous, mostly deep vein thrombosis (table 2). 32% (n=40) of patients presented first with a vascular event and diagnosed as BD. Twenty (16%) patients were diagnosed with a median of 12^{1–120} months after the first vascular event. 15 (6.5%) patients were using immunosuppressive (IS- mainly azathioprine) drugs either for resistant mucocutaneous symptoms or major other organ involvement during the first vascular event.

Vascular relaps rate was 40.7% and it was similar between sexes (F: 33.3% vs M: 42.2%, $p=0.6$). After the first vascular event, 96 (85.7%) patients had been treated with ISs and 58.9% used anticoagulants. Median IS and anticoagulant usage duration was 25.5 (5–48) and 2 (0–12) months respectively. Relaps rate was higher in patients who had stopped ISs (87.5% vs 32.3%). IS treatment duration