Figure 1. ORs and 95% CIs of individual studies and pooled data; comparison of endovascular intervention and surgical revascularization for stenosis.

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FRI0312 THE FREQUENCY AND SEVERITY OF PATIENT-REPORTED SYMPTOMS IN GIANT CELL ARTERITIS

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Background: A better understanding of the patients' perspectives is pivotal in the development of patient-reported outcomes (PROs) in vasculitis.

Objectives: To assessed patients' perspective of disease amongst cases with Giant Cell Arteritis (GCA) compared to comparator illnesses mimicking large vessel vasculitis (LVV) included in the Diagnostic and Classification Criteria in Vasculitis Study (DCVAS) database.

Methods: Patient Description of Illness (PDI) forms were circulated amongst Centres participating in the DCVAS study. The PDI form records up to 10 free-text severity ranked symptoms in descending order of severity, a body-map to localise the sites of pain and a free-text short summary of illness description. Free text was reorganized through content and thematic analysis.

Results: PDI forms from 89 patients with GCA and 28 comparators (COM) were analysed. There was no difference in age and sex distribution between groups (mean age 70±8 for GCA and 69±12 for COM). The symptoms description and frequency of the first most severe aspect of disease, including the patient's own words, is presented in Table 1. The symptom regarded as the most severe by

Abstract FRI0312 – Table 1. Top 10 most recurrent patient-reported symptoms and correspondent severity rank in giant cell arteritis (GCA) and Comparators (COM)

Item	Frequency in GCA	Severity in GCA	Frequency in COM	Severity in COM	Examples of patient's own words
Headache	1	1, 2, 8	1	1, 2, 5	Headaches; Sore head; Thumping headache
Jaw claudication	2	3, 6	6	0	Jaw ache; Pain in jaw and teeth
Shoulder/neck pain	3	3, 4, 5,	0	0	Shoulder upper arm pain
Fatigue	4	5, 6, 10	3	3, 6, 7	Severe tiredness; Fatigue; No energy and exhausted
Myalgia or muscle weakness	5	5, 7, 10	4	3	Aching muscles; Achey limbs; Loss of strength in arms and legs
Blurred vision	6	10	5	4, 8	Blurred vision
Scalp tenderness	7	8	0	4	Irritation to the scalp; Tender scalp
Loss of appetite	8		0	0	Lack of appetite
Flu-like symptoms	9	9	0	0	General ill feeling; Flu-like symptoms; Unwell
Arthralgia or arthritis	10	9	2	3	Hip, knee more on right side; Pain in back of neck, ankles, wrists, and chest
Other ENT	0	0	7	4	Severe sinusitis; sore inside gums
Sudden visual loss	0	0	8		Loss of eyesight to both eyes; Vision loss
Night sweats	0	0	9	4, 8	Night sweats; Night fever sweats
Painful eyes	0	9	10	0	Shooting pain left eye; Pain right eye

both groups was headache. While there were no differences in the frequency of sudden visual loss, visual symptoms were reported more commonly as the most severe feature by COM vs GCA (21% vs 8%, $p=0.05$). Arthralgia was more frequently reported by COM vs GCA (11% vs 1%, $p=0.01$). Headache was the most frequently reported symptom in both groups. Patients with GCA reported jaw claudication (37%) as the second most frequently reported symptom, while COM reported arthralgia/arthritis (32%). Shoulder/neck pain was the third most important symptom in GCA (33%), while fatigue was the third most common complaint among COM (21%). Fatigue was reported as the fourth most common feature by 30% of GCA patients.

Conclusions: Headache was the most frequent and most severe symptom reported by patients with GCA and comparators. However, the reported frequencies and severities of other symptoms were significantly different between the two groups.

Disclosure of Interest: None declared

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FRI0313 THE EFFICACY AND SAFETY OF TOCILIZUMAB THERAPY IN PATIENTS WITH POLYMYALGIA RHEUMATICA WHO WERE RESISTANT OR INTOLERANT TO GLUCOCORTICOIDS AND ADDITIONAL METHOTREXATE

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Background: A recent trial of tocilizumab (TCZ) in patients with newly diagnosed polymyalgia rheumatica (PMR), conducted in Europe and the United States, has shown its efficacy and safety.

Objectives: We examined the efficacy and safety of TCZ for patients with PMR who had been primarily resistant or intolerant to glucocorticoids (GC) and additional methotrexate (MTX).

Methods: Sixty patients had been diagnosed with having PMR since 2011. The patients are all compatible with the 2012 EULAR/ACR provisional classification criteria for PMR, and had been treated first with GC and then, if they were resistant or intolerant to GC, were added MTX, similarly to the 2015 EULAR/ACR recommendations for the management of PMR. The disease activity were measured by PMR-AS.

Results: There were 17 patients with GC/MTX resistant or intolerant PMR (28%). Of them, 9 patients with PMR agreed to the proposal of TCZ addition, and their therapeutic responses to TCZ and its safety were determined. They were at the age of 68.2±10.6, including three males and six females. Before TCZ addition, the patients were treated with prednisolone (PSL) at 7.6±3.0 mg/day plus MTX at 7.1±5.1 mg/week, and serum CRP were at 1.0±1.0 mg/dL. After 8.4±5.7 months of TCZ treatment, PSL and MTX had been reduced to 1.1±1.3 mg/day and 3.3±4.5 mg/week, respectively, with CRP at 0.02±0 mg/dL. GC were able to be withdrawn in 5 patients, and MTX were further withdrawn in 4 patients. Two patients reached drug-free remission (PMR-AS=0.02). During TCZ therapy, each one patient showed the worsening of depression and occlusion of the central retinal vein.

Conclusions: These results indicate that TCZ may provide a therapeutic option for patients with severe PMR who were resistant or intolerant to GC and additional MTX.

Disclosure of Interest: None declared

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FRI0314 DIFFERENT SERUM CYTOKINE PROFILES REFLECT ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODIES (ANCA) SPECIFICITY IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

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Background: Evidence supporting the classification of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV) based on ANCA type is accumulating.¹
Objectives: To evaluate serum cytokine profiles in patients with AAV classified by ANCA specificity (proteinase 3 (PR3)-ANCA versus myeloperoxidase (MPO)-ANCA) or by clinical diagnosis (granulomatosis with polyangiitis (GPA) versus microscopic polyangiitis (MPA)) and clinical phenotypes.
Methods: An antibody array testing 30 soluble mediators, already shown to be implied in the pathogenesis of AAV, was performed in each patient with active AAV at inclusion in the Rituximab in ANCA-Associated Vasculitis (RAVE) trial, as previously described.^{2,3} By means of Wilcoxon signed rank test for univariate analyses, we analyzed the association of levels of these cytokines with ANCA specificity, clinical diagnosis, and distinct clinicopathologic phenotype categories derived from the BVAS/WG items recorded at the time of enrollment (capillaritis, granulomatous manifestations, renal involvement, and alveolar hemorrhage; new diagnosis and relapsing disease), as described.⁴
Results: All cytokines tested (see Figure 1 and legend for the complete list), except for RANTES, ACE, bFGF and VCAM-1, were significantly increased in the RAVE cohort when compared to healthy controls ($p<0.05$). Median Birmingham vasculitis activity score and steroid use at screening did not significantly differ between PR3-AAV and MPO-AAV, and between GPA and MPA. Within both ANCA specificities, levels of 9 mediators were significantly higher in PR3-AAV (IL-6, NGF, GM-CSF, IL-15, IL-18, IL-18Bb, sIL-2Ra, IL-8, TARC; $p<0.05$), compared to 5 different cytokines that were higher in MPO-AAV (sIL6R, NGAL, sICAM-1, VCAM-1, sTNFR II; $p<0.05$). In contrast, only 4 cytokines (GM-CSF, IL-15, IL-18, sIL-2Ra) were higher in GPA than MPA, and 1 (NGAL) was higher in MPA than GPA ($p<0.05$). The association of the majority of cytokines was stronger with ANCA specificity than with the clinical diagnosis (Figure 1). Similarly, the defined clinical phenotypes were also not separated by cytokine signatures as clearly as the ANCA specificity was (*data not shown*). Restricting the analysis to newly-diagnosed patients ($n=90$) showed the most significant cytokine profile differences associated with PR3-AAV (Figure 1).

Conclusions: Cytokine profiles separate patients more clearly by ANCA specificity than by clinical diagnosis, suggesting important differences in underlying pathophysiology and validating stratification of patients by ANCA specificity for treatment trials.
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FR10315 CLINICAL CHARACTERISTICS OF VASCULAR INVOLVEMENT IN BEHCET'S DISEASE

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Background: Behçet's disease (BD) is a multisystemic inflammatory disorder classified as vasculitis, which can affect all types of vessels. The prevalence of vascular involvement has been reported at rates ranging from 15 to 50%.
Objectives: In this study, we aimed to determine the characteristics of vascular involvement in patients with BD.

Methods: Six hundred and ninety-two patients with BD, who applied to the multidisciplinary BD polyclinic between 2006–2016, were retrospectively analyzed. The diagnosis of vascular involvement was made on clinical signs, by Doppler ultrasonography and/or angiography using computed tomographic or magnetic resonance techniques where appropriate.
Results: One hundred and seventy-six patients (25.4%) had vascular involvement. The mean age of patients with vascular involvement ($n=176$) and non-vascular involvement ($n=516$) was similar to 42.5 ± 12.3 and 42.2 ± 12.2 years, respectively, while male sex frequency was significantly higher in patients with vascular involvement (72% to 42%, $p<0.001$). Uveitis (45%) and erythema nodosum (EN) (47.7%) were significantly higher in patients with vascular involvement than those without vascular involvement (for Uveitis OR: 1.7, $p=0.003$ /for EN: OR: 2.5, $p<0.001$) (Table 1). Deep vein thrombosis was detected in 142 (80.7%) of the patients with vascular involvement and this was the most common type of vascular involvement (Table 2). Arterial involvement was detected in 52 (29.5%) of 176 patients with vascular involvement. Deep venous thrombosis alone was present in 125 (71%) patients, arterial involvement alone in 12 (6.8%) patients, arterial and venous involvement together detected in 40 (22.7%) patients. Arterial involvement was most commonly seen as pulmonary artery involvement in 45 (25.6%) patients [pulmonary artery thrombosis 35 (20%), aneurysm 10 (5.7%) and thrombosis + aneurysm 6 (3.4%) patients]. Twenty (11.3%) patients had other arterial involvement except pulmonary, 8 of them had thrombosis and 12 had aneurysm.

Table 1. Clinical features in patients with and without vascular involvement

	With vascular involvement	Without vascular involvement	OR	95% CI	P value
n (%)	176 (25.4)	516 (74.6)			
Age (year)	42.5±12.3	42.2±12.2			0.762
Male n (%)	127 (72.2)	217 (42.1)	3.57	2.46–5.19	<0.001
Genital ulceration n (%)	123 (69.9)	392 (76.1)	0.728	0.498–1.065	0.101
Uveitis n (%)	79 (44.9)	166 (32.4)	1.698	1.196–2.409	0.003
Papulopustular erythema or acniform rash n (%)	101 (51.4)	186 (57.4)	1.274	0.886–1.832	0.190
EN n (%)	84 (47.7)	98 (27.1)	2.460	1.689–3.582	<0.001

Table 2. Venous involvement (%)

Deep vein thrombosis	142 (80.7)
Budd-Chiari syndrome	7 (4.0)
Inferior and superior vena cava syndrome	26 (14.7)
Iliac vein thrombosis	18 (10.2)
Safenus vein thrombosis	23 (13.1)
Femoral vein thrombosis	36 (20.5)
Popliteal vein thrombosis	43 (24.4)
Crural vein thrombosis	12 (6.8)
Cerebral sinus thrombosis	20 (12.2)

Conclusions: Vascular involvement in Behçet's disease is commonly seen in males. Deep vein thrombosis is the most common form of vascular involvement. Arterial involvement is important because of its higher risk of mortality (especially pulmonary artery aneurysm). The incidence of EN and uveitis is higher in patients with vascular involvement.
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FR10316 FACTORS ASSOCIATED WITH SURGICAL OUTCOMES OF SEVERE AORTIC REGURGITATION IN PATIENTS WITH BEHCET'S DISEASE

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Background: In rare cases, Behçet's disease (BD) can cause severe aortic regurgitation (AR) or aortic root destruction that might have lethal outcomes. Conventionally, simple aortic valve replacement (AVR) is performed for the management of severe AR in BD patients; however, the reoperation rate is as high as 78 to 100%, while mortality rate range from 20 to 47%. Recently, several series of case reports showed that compared to AVR, aortic root replacement (ARR) improved the surgical outcomes of AR in BD patients.
Objectives: To identify the factors associated with the long-term surgical outcomes of AR in BD patients.
Methods: We identified 23 patients who had been surgically treated for AR caused by aortic root involvement of BD from January 1996 through December 2013. We evaluated the occurrence of post-surgical adverse events, which were defined as follows: death, aortic valve/graft problem, infective endocarditis, cerebral infarction, and/or re-operation of aortic valve or root. Types of surgery were classified as simple aortic valve replacement (AVR), bioprosthesis aortic root replacement (bARR), and mechanical valved composite graft aortic root replacement (cARR). Clinical parameters including baseline characteristics, C-reactive protein (CRP), erythrocyte sedimentation rate, and medications were extracted from electronic medical records.
Results: Appropriate aortic valve or root surgery cases were 35 in total, with a mean follow up duration of 11 ± 5 years in 23 patients. Out of the 11 cases