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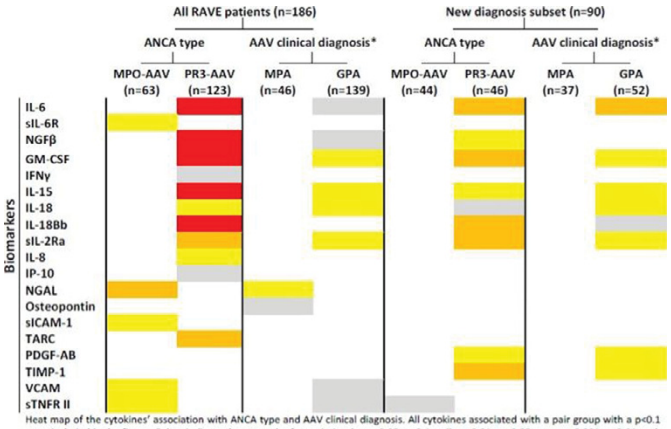
Background: Evidence supporting the classification of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV) based on ANCA type is accumulating.[1]
Objectives: To evaluate serum cytokine profiles in patients with AAV classified by ANCA specificity (proteinase 3 (PR3)-ANCA *versus* 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, NGFb, 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).



Heat map of the cytokines' association with ANCA type and AAV clinical diagnosis. All cytokines associated with a pair group with a $p<0.1$ were included in the figure. Colors indicate the strength of association (grey, $0.05<p\leq0.1$; yellow, $0.01<p\leq0.05$; orange, $0.001<p\leq0.01$; red, $p\leq0.001$). The tested panel of 30 biomarkers also included BAFF, BCA-1, bFGF, G-CSF, PAI-1, KIM-1, MMP-3, RANTES, IL-12(p70), ACE, Clusterin, which showed no association ($p\geq0.1$) and therefore were not included in the figure.

*One patient had an indeterminate type of AAV.

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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FRI0315 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 policlinic 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 acneiform 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)
Safenous 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.
Disclosure of Interest: None declared
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FRI0316 FACTORS ASSOCIATED WITH SURGICAL OUTCOMES OF SEVERE AORTIC REGURGITATION IN PATIENTS WITH BEHCET'S DISEASE

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Background: In rare cases, Behcet'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