patients (5 infliximab and certolizumab each, 2 adalimumab and 2 tocilizumab). Remission was observed in 84% of the patients. At least one relapse was occurred in 43% and >1 relapse in 14 patients. At the last visit 26% were determined to have an active disease. A >4 mg of methylprednisolone dose was required in only 8.4%. Mortality rate was 3.7% (4 patients).

Table 1 Clinical characteristics and outcomes of patients

Gender (F)	96 (89.8%)
Takayasu Arteritis Type	
1	42 (38.8%)
2	8 (7.4%)
3	0 (0%)
4	2 (1.8%)
5	55 (51.8%)
Age at symptom onset	30 (14-67)
Age at diagnosis	33 (14-68)
Time for diagnosis delay (months)	12 (0-180)
Follow-up time (months)	72 (6-264)
Cardiovascular risk factors (Baseline)	
Hypertension	41 (38.3%)
Diabetes	6 (5.6%)
Dyslipidemia	25 (23.4%)
Coronary artery disease	13 (12.1%)
Congestive heart failure	4 (3.7%)
peripheral artery disease	13 (12.1%)
Stroke	10 (9.3%)
Smoking	
Current	30 (28%)
Former	6 (5.6%)
Family history of atherosclerosis	28 (26.2%)
Laboratory findings (baseline)	
ESR (mm/h)	43.8±27.9
CRP (mg/L)	18.2±22.6
Treatment at last visit n (%)	
Immunosuppressives	100 (93.4%)
Methotrexate	16 (15%)
Azathioprine	49 (45.8%)
Leflunomide	20 (18.7%)
Infliximab	2 (1.9%)
Adalimumab	1 (0.9%)
Certolizumab	3 (2.8%)
Tocilizumab	2 (1.8%)
Certolizumab+Azathioprine	1 (0.9%)
Certolizumab+methotrexate	1 (0.9%)
Infliximab+azathioprine	3 (2.8%)
Adalimumab+Leflunomide	1 (0.9%)
Glucocorticoids*	51 (47.7%)
≤ 4 mg	42 (39.2%)
> 4 mg	9 (8.4%)
Cumulative glucocorticoid dose (g)	7.5 (0-31.9)
Damage Scores	
VDI**	5.6±2.3
TADS***	8.2±3.1

*methylprednisolone **Vasculitis Damage Index ***Takayasu arteritis damage score

Conclusions: We have defined the long-term follow-up results of our Takayasu's arteritis cohort. Comparing with European and Asia series published recently, requirement for a surgical intervention was lower under immunosuppressive treatments in our series. However, disease activity and relapse rate were still high under conventional ISSs, suggesting a need for better therapeutic options.

Disclosure of Interest: None declared

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THU0463 COMPLEMENT FACTORS OF THE ALTERNATIVE PATHWAY IN GPA AND MPA

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Background: In antineutrophil cytoplasm autoantibody (ANCA)-associated vasculitis (AAV), involvement of complements, especially alternative pathway of complement, has been reported in researches using mouse models. In human, while some studies have identified levels of C3 as a renal prognostic factor, entire complement factors in alternative pathway have not been evaluated.

Objectives: To evaluate complement profiles of AAV patients at diagnosis and at 6 months after treatments (Month 6).

Methods: In total, 91 incident cases of granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) based on the European Medicines Agency algorithm were enrolled. They are a part of participants of the Japanese nationwide, prospective, inception cohort study from 22 tertiary Japanese institutions (RemiT-JAV-RPGN). Multiplex complement bead assays to measure levels of C1q, C2, C3, C3b/iC3b, C4, C4b, C5, C5a, C9, Factor D, Factor I, MBL, factor B, factor H, and properdin were performed using preserved sera at diagnosis and at Month 6. We compared complement levels at diagnosis of AAV patients with those of age- and sex-matched healthy donors, and compared those of the AAV patients at diagnosis and Month 6.

Results: Seventy-two of 91 patients had serum both at baseline and Month 6. Compared with healthy donors, GPA patients had significantly higher levels of C2, C5a, and Factor B, and significantly lower levels of C3b/iC3b, C4b, and properdin. MPA patients had significantly higher levels of C2 and Factor D, and significantly

lower levels of C3b/iC3b, C4, C5, Factor H, and properdin (table 1). At baseline, GPA had significantly higher levels of C4, Factor B and Factor H, and had significantly lower levels of C4b and Factor D compared to MPA. There are no significant differences in levels of C3, C5, Factor D, MBL, and properdin using Wilcoxon signed-rank test between at diagnosis and Month 6 both in GPA and MPA. Factor I significantly decreased at Month 6 only in GPA. Other complement factors significantly decreased at Month 6 both in GPA and MPA.

Abstract THU0463 – Table 1. Complement profiles of patients with AAV at baseline and healthy donors

	GPA (median)	MPA (median)	HD (median)	GPA vs. MPA	GPA vs. HD	MPA vs. HD
C1q, ng/mL	1 04 220	96 890	1 08 242	N.S.	N.S.	N.S.
C2, ng/mL	50 181	54 422	18 610	N.S.	p<0.05	p<0.05
C3, ng/mL	1265500	1371550	1416900	N.S.	N.S.	N.S.
C3b/iC3b, ng/ mL	10433000	11066000	17364500	N.S.	p<0.05	p<0.05
C4, pg/mL	3 10 020	2 66 554	3 08 085	p<0.05	N.S.	p<0.05
C4b, ng/mL	18 064	27 740	31 287	p<0.05	p<0.05	N.S.
C5, ng/mL	30 014	27 805	32 015	N.S.	N.S.	p<0.05
C5a, pg/mL	7783	6592	4836	N.S.	p<0.05	N.S.
C9, ng/mL	6934	5905	6742	N.S.	N.S.	N.S.
Factor D, ng/ mL	5335	7706	5658	p<0.05	N.S.	p<0.05
Factor I, ng/ mL	29 917	25 633	25 653	N.S.	N.S.	N.S.
MBL, ng/mL	3583	3638	3023	N.S.	N.S.	N.S.
Factor B, ng/ mL	2 54 961	1 80 045	2 12 153	p<0.05	p<0.05	N.S.
Factor H, ng/ mL	2 58 187	2 28 238	2 95 480	p<0.05	N.S.	p<0.05
Properdin, ng/mL	18 794	19 665	32 521	N.S.	p<0.05	p<0.05

GPA, granulomatosis with polyangiitis; MPA, microscopic polyangiitis; HD, healthy donor.

Conclusions: We found some differences in complement factors among GPA, MPA, and healthy donors. There were no differences of levels of C3, C5, Factor D, and properdin, which suggested involvements of alternative pathway, both in GPA and MPA between at diagnosis and Month 6.

Disclosure of Interest: None declared

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THU0464 INCREASED FREQUENCY OF OBSTRUCTIVE SLEEP APNEA SYNDROME IN BEHÇET'S SYNDROME PATIENTS WITH VENA CAVA SUPERIOR THROMBOSIS

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Background: Superior vena cava syndrome (SVCS), is a medical emergency and can also be seen in Behçet's syndrome (BS). Contrary to the severe outcome seen in malign conditions, SVCS in BS usually has a benign course, complicated rarely by hemoptysis, pleural effusion, and a chylothorax. We had noted that BS patients with SVCS frequently complained of sleep disturbances, snoring and sleep apnea, suggesting an obstructive sleep apnea (OSA) disorder.

Objectives: We formally surveyed the degree of risk for OSA among BS patients with SVCS and suitable controls using the Berlin questionnaire, a screening questionnaire for OSA with a high sensitivity and modest specificity.¹

Methods: Because of the lower frequency of female patients with VCSS (n=2), only males were included. We studied 28 BS patients with SVCS (Group 1), 80 BS patients with vascular involvement without a SVCS (Group 2), and 59 BS patients with no vascular involvement (Group 3). Also, 80 apparently healthy individuals (Group 4) of similar age and gender to BS patients were studied. Polysomnography was performed in patients at high risk for OSA according to the Berlin questionnaire.

Results: There were no differences regarding demographic characteristics, disease duration, and variables associated with OSA among the groups (table 1). The Berlin questionnaire categorised 57.1% (16/28) of the BS patients with SVCS (Group 1) as having a high risk for OSA and this was significantly higher compared to that found in the control groups. The frequency of those at high risk for OSA was 15%, 8.5%, 11.3% in Group 2, 3 and 4, respectively (p>0.05). Until now, polysomnography was performed in 12 subjects (5 patients with SVCS, 1 patient with vascular involvement without a SVCS and 6 healthy controls). OSA was detected

in 3/5 patients with SVCS and 1/1 patient with vascular involvement without a SVCS and 4/6 healthy controls.

Abstract THU0464 – Table 1. Demographic characteristics and variables associated with obstructive sleep apnea

	Group 1 (BS patients with SVCS) (n=28)	Group 2 (BS patients with vascular involvement without SVCS) (n=80)	Group 3 (BS patients with no vascular involvement) (n=59)	Group 4 (Healthy controls) (n=80)	p value
Age, mean ±SD, years	44.3±9.7	42.1±7.8	41.9±5.9	42.7±9.7	0.051
Disease duration, mean±SD, years	18.7±9.4	14.6±7.7	12.5±6.5		0.001
Hypertension, n (%)	4 (14.3)	6 (7.5)	2 (3.4)	4 (5)	NS
BMI, mean ±SD	26.8±4.7	26.4±3.9	26.2±3.3	27.0±3.5	NS
High risk for OSA, n (%)	16 (57.1)	12 (15)	5 (8.5)	9 (11.3)	<0001

Conclusions: This study shows that BS patients with a history of VCSS are at high risk of OSA. This is probably due to the external pressure of the significant venous collaterals on the upper airways.

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THU0465 A LONGITUDINAL STUDY OF NEUTROPHIL PHENOTYPE CHANGES IN GIANT CELL ARTERITIS

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Background: Neutrophils with differential surface protein expression were recently implicated in pathogenesis of Giant Cell Arteritis (GCA)¹. However, data are lacking with regard to treatment-naïve GCA and their long-term follow-up.

Objectives: To determine the expression of I-selectin (CD62L) and integrin αM (CD11b) on CD16⁺ neutrophils in peripheral blood of newly diagnosed, treatment-naïve GCA cases, at the time of diagnosis (time 0) and during follow-up - at week 1, 4, 12, 24 and 48. In parallel, we aimed to measure also sera levels of serum amyloid A (SAA) and interleukin-6 (IL-6).

Methods: Peripheral blood from 33 treatment-naïve GCA patients and 16 healthy blood donors (HBD) was stained, lysed, fixed and analysed by flow cytometry (Miltenyi). SAA and IL-6 were measured by nephelometry and ELISA, respectively. 3/33 GCA patients experienced relapse and were analysed separately during follow-up, at the time of relapse and 12 weeks after relapse. At the time of diagnosis, all patients received steroid treatment and therapy tapering started after 4 weeks. At week 12, some of the patients (14/22) received leflunomide, in addition to steroid therapy.

Results: Expression of CD62L, but not CD11b, was significantly higher on CD16⁺ neutrophils in treatment-naïve GCA patients (median; IQR: 72.5; 56.2–100.7), as compared to HBD (55.9; 44.5–70.6, p=0.017). Longitudinally, the expression of CD62L significantly decreased in GCA patients from day 0 to week 4 (p=0.005). At week 12 there was an elevation in CD62L which escalated also at week 24. Patients receiving steroids only showed a marked increase at week 48, while patients receiving also leflunomide exhibited a decrease (Fig 1). Expression of CD11b declined from day 0 to week 4, but substantially increased throughout weeks 12, 24 and 48, regardless of therapy used. SAA and IL-6 declined sharply from day 0 to week 1 and 4, with gradual elevation of both at week 12. There was a decline in SAA levels observed in all patients at week 24, while IL-6 increased in patients receiving only steroids. These patients exhibited further elevation of both markers at week 48, while patients receiving steroids, in combination with leflunomide, showed a decrease. In 2/3 patients who experienced a relapse, we could observe an increase in the expression of CD62L at the time of relapse, which was found to be decreased again 12 weeks later. A similar trend was observed for IL-6.

Conclusions: Neutrophil CD62L could represent a good surface marker for detection of relapse in GCA. A distinct dichotomy was found for CD62L, as well as SAA and IL-6 in GCA during long term follow-up, with the combination of steroids with leflunomide showing more optimal results.

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THU0466 SEXUAL DYSFUNCTION AND DEPRESSION IN BEHCET'S DISEASE – ARE THERE DIFFERENCES REGARDING PATIENT'S ORIGIN

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Background: Behcet's disease (BD) is a systemic vasculitis of veins and arteries characterised by oral and genital ulcers (aphthae), skin lesions and uveitis. BD is more common in Middle Eastern countries and Asia but also occurs in Caucasian people.

Objectives: Aim of this study was to evaluate the prevalence of sexual dysfunction (SD) in patients with BD as well as analysing differences between patients from different origins. Additionally we investigated the prevalence of depression in both ethnic groups.

Methods: This prospective, monocentric study included 106 patients with BD. The International Index of Erectile Function (IIEF) and the Female Sexual Function Index (FSFI) were used for assessing sexual dysfunction and the Beck Depression Inventory (BDI) was used for depression assessment.

Results: The mean age of our group was 40.5 years. Half of the patients had Middle Eastern and half Caucasian origin. SD was found in 24.5% of all subjects. Only 6.9% of the male patient's group showed signs of SD, while half of the women's group was suffering from SD (p<0.001). The prevalence for SD was significantly higher in women with Middle Eastern origin compared to women with Caucasian origin (75% vs. 33.3%, p=0.024). Genital ulcers affected 73.6% of all patients. Depression was found in 36.7% of all subjects. Both SD and depression correlated positively in males (p=0.017) and females (p=0.013).

Conclusions: SD and depression are very common problems in BD and should be addressed by the treating physician. Both manifestations are intensifying each other.

Disclosure of Interest: None declared

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THU0467 EFFICACY AND PATENCY OF REVASCULARISATION IN PATIENTS WITH THROMBOANGIITIS OBLITERANS (BUERGER'S DISEASE)

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Background: The cornerstone of therapy in thromboangiitis obliterans (TAO) is complete abstinence from tobacco. In addition to discontinuation of cigarette smoking, very few pharmacological and surgical options of controversial efficacy are available to date. TAO is associated with a high amputation rate because of tobacco continuation, medical therapy failure, and desert foot with no revascularisation option. Endovascular or bypass revascularisation patency and efficacy are poorly described in the literature.

Objectives: To describe the results of the French National Reference Centre on TAO on revascularisation modalities, their patency and efficacy.

Methods: Among the 198 patients followed in our centre, we retrieved data from 19 patients for who a revascularisation procedure has been attempted. Patency was assessed on angioCT scan and Duplex ultrasounds performed during follow-up. Efficacy was assessed according to clinicians in charge of the patient, and if