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