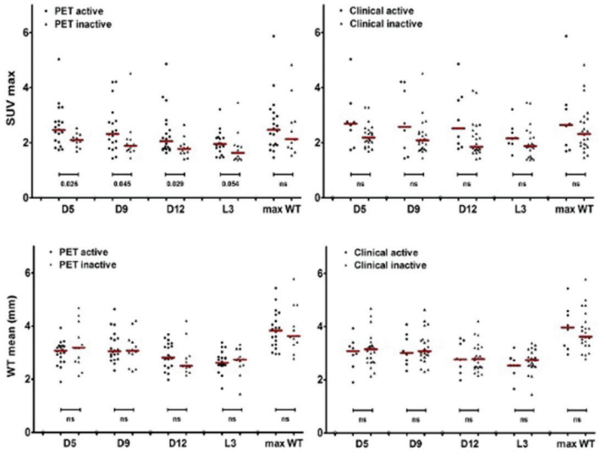
	All (n 23)	*GCA (n 13)*	*TAK (n 8)*	*p*
Female, n (%)	19 (82.6)	12 (92.3)	6 (75)	*ns*
Age at diagnosis	63 (51–68)	68 (63–73)	43.5 (30.5–57)	*0.003*
Diagnostic latency (months)	4.5 (2–12)	3 (2–10)	8 (3.5–12)	*ns*
ESR at disease onset (mm/h)	49 (38–68)	52.5 (45.5–59.7)	42 (40–61.5)	*ns*
CRP at disease onset (mg/L)	61.8 (13–132.5)	89 (32.5–106)	60.5 (9.3–132.5)	*ns*
Disease duration at PET/MR (months)	27 (18–36)	24 (13–29.5)	36.5 (14.75–129.3)	*ns*
ESR at examination (mm/h)	18 (9–35)	16 (7–31)	20.5 (11–44.5)	*ns*
CRP at examination (mg/L)	4.5 (2.55–8.9)	3.9 (3.48–4.72)	4.55 (2.05–10.4)	*ns*

Results: 23 LVV patients were included, 56.5% GCA, 34.8% TAK and 8.7% isolated aortitis, all Caucasian, mostly females (82%). We considered 55 PET scans, 32/55 in LVV group (from min. 1 to max. 3 scans/patient) mainly during follow-up (29/32 scans), and 23/55 in control group. Considering patients with abdominal aorta involvement, we found higher SUV max compared to controls, in all sites, regardless of disease activity. Mean WT resulted higher in patients than controls, but did not significantly differ between PET active or inactive patients (figure 1). Mean WT positively correlated with age in both cohorts, inversely correlated to disease duration in LVV patients, while no correlation with SUV max was observed. Despite clinical assessment was suggestive of remission in 24 (75%) cases before PET/MR acquisition, a normal uptake was present only in 12 (50%) of them. On the contrary, all patient with active disease at clinical examination (8, 25%) had also a positive PET/MR. Cohen's K coefficient between clinical assessment and imaging was poor (K Cohen=0, 33, 0.11–0.55). Finally, we found no significant correlation between SUV max and acute phase reactants.
Demographic and clinical data of LVV patients.



Conclusions: PET/MR is a safe imaging technique capable of detecting vasculitic inflammation, similar to PET/CT, but with a greater anatomical definition. The low radiological exposure represents a valid alternative to PET/CT for disease monitoring, especially in young women.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.4773

THU0461 COMPARISON BETWEEN CLINICAL PROFILE AND OUTCOME OF PATIENTS WITH JUVENILE ONSET AND ADULT ONSET TAKAYASU ARTERITIS

R.A Goel[1], D. Danda[1], G. Joseph[2], A. Nair[1], R. Ravindran[1], V. Jeyaseelan[3].
[1]Clinical Immunology and Rheumatology; [2]Cardiology; [3]Biostatistics, Christian Medical College, Vellore, Tamil Nadu, Vellore, India

Background: There is a paucity of data comparing juvenile onset Takayasu arteritis (jTA) and adult onset TA (aTA).
Objectives: We aimed to compare differences in clinical profile and outcome of patients with jTA and aTA attending our centre during 1998–2017.
Methods: Details of demography, clinical presentation, laboratory results, angiography and treatment response were collected prospectively for 252 and retrospectively for the rest of patients with TA. Disease activity was defined by Indian Takayasu Activity Score- A (ITAS-A)(CRP).[1] Complete remission (CR) was defined as ITAS-A=0 with no angiographic progression. Patients with onset of disease at ≤16 years of age were classified as jTA while the rest as aTA.

Results: Among 602 patients with TA during this period, 119 (19.8%) were jTA, while 483 were aTA. Female predominance was less striking in jTA (71.4%) than aTA (79%), p=0.047. Patients with jTA had presented more commonly with fever (29% vs 17.4%, p=0.002), headache (31% vs 18%, p=0.002), pain abdomen (11% vs 5.6%, p=0.031), systolic hypertension (66.4% vs 48.4%, p<0.001), cardiomyopathy (15.1% vs 5.4%, p<0.001) and raised creatinine (16% vs 4.7%, p<0.001) while claudication as presenting symptom was less common in jTA (39%) as compared to aTA (55%), p=0.003. Pulse abnormality tended to be commoner in aTA. Angiographically, type-I disease (5.1% vs 22.6%, p<0.001) and coronary involvement (8.3% vs 20.6%, p=0.016) was less common while type-IV disease occurred more frequently (25% vs 14.3%, p=0.004) in jTA than in aTA. Logistic regression showed similar results after adjustment for gender. Median ITAS ^{2010} score was higher in jTA [7 (2–14)] than aTA [5 (2–11)].
Follow up was available for 77 and 287 patients with jTA and aTA respectively. Median follow up duration was $32^{10–61}$ months for jTA and $27^{10–59}$ months for aTA. **CR** was attained more frequently in jTA (n=67; 87%) than aTA (n=190; 66.2%), p=0.001. Another, 7 (9%) and 55 (19.2%) of patients with jTA and aTA respectively achieved partial response with immunosuppression. Among patients with initial CR, relapse of active disease during further follow up was observed more frequently in jTA [n=20, (29.9%)] as compared to aTA [n=50,(26.6%)], p=0.029. Altogether, persistently stable disease course was more common in jTA (62.3%) than aTA (47.5%), p=0.029.
Conclusions: In our large cohort of TA treated with uniform immunosuppression protocol, systemic features, hypertension, cardiomyopathy, renal dysfunction and type IV disease are more commonly observed in jTA while claudication, pulse abnormality, coronary involvement and type I disease are more frequent in aTA. Patients with jTA respond better to immunosuppression but relapse more frequently than aTA. Persistent stable disease course is commoner in jTA patients.

REFERENCE:

[1] Ruchika Goel, Debashish Danda, et al. Long term outcome of 251 patients with Takayasu Arteritis on combination immunosuppressant therapy: single centre experience from a large tertiary care teaching hospital in southern India. Seminars in Arthritis and Rheumatism, http://dx.doi.org/10.1016/j.semarthrit.2017.09.014

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.5158

THU0462 LONG TERM FOLLOW-UP RESULTS OF TAKAYASU ARTERITIS COHORT: A TERTIARY-SINGLE CENTRE STUDY

S. Kaymaz Tahra, F. Alibaz-Oner, H. Direskeneli. *Department of Rheumatology, Marmara University, ıstanbul, Turkey*

Objectives: To assess the clinical characteristics and long term follow-up outcomes of patients with Takayasu's arteritis (TAK) in a tertiary referral centre.
Methods: In this retrospective study, 107 (F/M: 96/11) patients fullfilling ACR 1990 criteria for Takayasu Arteritis and referred to our centre between 2004 and 2017 were investigated. All clinical and demographic data during first diagnosis and longitudinal follow-up were abstracted from medical records. Relapse was defined according to the physician's global assessment (PGA).
Results: The median age was $30^{14–67}$ years at symptom onset and $33^{14–68}$ years at diagnosis. Median follow-up duration was 72 (6–264) months. According to Hata Angiographic Classification, Type 5 (51.8%) and Type 1 (38.8%) were the most common patterns with the most frequently affected vessel subclavian artery (82.2%). At diagnosis 0.5–1 mg/kg/day corticosteroid treatment was started in 94.6% patients and a steroid-sparing immunsuppressive (IS) agent in 96.3% of the patients. An initial pulse steroid (1 g/day) therapy was chosen for 8 patients. Before diagnosis 24% patients had a history of a revascularisation procedure. After IS treatments, 24% of the patients were undergone a new revascularisation procedure. During follow-up, biologic agents were chosen for 13.8% of the

Ann Rheum Dis: first published as 10.1136/annrheumdis-2018-eular.4790 on 12 June 2018. Downloaded from http://ard.bmj.com/ on February 26, 2022 by guest. Protected by copyright.

patients (5 infliximab and certolizumab each, 2 adalimumab and 2 tociluzumab). Remission was observed in 84% of the patients. At least one relapse was occured 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)
Dislipidemia	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 (%)	
Immunsupressives	100 (93.4%)
Methotrexate	16 (15%)
Azathiopurin	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+ Leflunamide	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

*metilprednisolone ** 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 ISs, suggesting a need for better therapeutic options.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4790

THU0463 COMPLEMENT FACTORS OF THE ALTERNATIVE PATHWAY IN GPA AND MPA

S. Fukui[1], K. Ichinose[1], K.-E. Sada[2], M. Harigai[3], A. Kawakami[1], on behalf of Japan Research Committee of the Ministry of Health, Labour, and Welfare for Intractable Vasculitis (JPVAS). *[1]Immunology and Rheumatology, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki*; *[2]Department of Nephrology, Rheumatology, Endocrinology and Metabolism, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama*; *[3]Department of Epidemiology and Pharmacoepidemiology, Tokyo Women's Medical University, Tokyo, Japan*

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, health 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

DOI: 10.1136/annrheumdis-2018-eular.3384

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

A. Gokturk[1], S.N. Esatoglu[1], Y. Ozguler[1], E. Atahan[2], B. Musellim[2], V. Hamuryudan[1], H. Yazici[1], E. Seyahi[1]. *[1]Istanbul University, Cerrahpasa Medical School, Department of Internal Medicine, Division of Rheumatology*; *[2]Istanbul University, Cerrahpasa Medical School, Department of Chest Diseases, Istanbul, Turkey*

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

Ann Rheum Dis: first published as 10.1136/annrheumdis-2018-eular.4790 on 12 June 2018. Downloaded from http://ard.bmj.com/ on February 26, 2022 by guest. Protected by copyright.