normal ($1.2x10^3/\mu l$; normal range 1.1 to $3.2x10^3/\mu$ l) but peripheral B cell numbers were markedly diminished ($37/\mu l$, normal range: 100 – $500/\mu l$). While the percentage of naïve B cells (53%) and marginal zone like B cells (10.5%) were within normal range, the percentage of class-switched memory B cells was high (28.2%). To assess the in vitro class switch capacity to IgE the number of IgE class-switched cells after 9 days of culture was determined by counting the events in the IgG/A/D-IgE+ gate. The percentage of IgE-switched cells was 6.8% (range: 0.3%>34%) and not statistically different from healthy controls.

Conclusions: In the EGPA-patients we report we observed markedly diminished B-cells despite of normal lymphocyte counts. Within the B cell compartment, there was a shift towards later B cell maturation stages. The in vitro B-cell development to IgE class-switched cells was not increased in EGPA-patients pointing towards a non-B cell intrinsic mechanism.

Disclosure of Interest: None declared

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THU0322 DESCRIPTIVE STUDY OF ASIAN INDIAN PATIENTS WITH RHEUMATOID VASCULITIS IN RETROSPECT: A SINGLE, TERTIARY CARE CENTRE EXPERIENCE

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Background: Rheumatoid vasculitis (RV) is a severe extra-articular manifestation of rheumatoid arthritis (RA), with high morbidity and mortality reported in literature

Objectives: To describe the Asian Indian perspective on RV patients, their clinico-laboratory features and their outcome along with the factors affecting them

Methods: A retrospective review of electronic medical records of 8984 RA patients from January 2007 to August 2016, was done for those satisfying Scott & Bacon criteria for RV[1]. Probable RV was defined as patients not satisfying Scott & Bacon Criteria, but were managed like RV after exclusion of alternate diagnosis. Birmingham Vasculitis activity score (BVAS) version 3[2] was used for monitoring activity of RV

Results: 63 patients of RV were identified, with a study period prevalence of 0.7%, in our RA cohort. 33 (52.4%) patients were female. Mean age of patients was 50.7±11.5 years with median duration of RA being 6 years. Involvement of Peripheral Nervous System (PNS) was the commonest manifestation of RV in 52/63 (82.5%) patients followed by skin in 34/63 (53.9%) patients. Rheumatoid Nodule was seen in 14/ 63 (22.2%) patients. Percentage of current and ex-smokers combined,was same as rheumatoid nodule prevalence. 52 (82.5%) patients had biopsy evidence of vasculitis.26/51 (50.9%)patients were started on mycophenolate mofetil, 13/51 (25.5%)patients on cyclophosphamide, 8/51 (15.7%) patients on azathioprine, 4/51 (7.8%) patients on Methotrexate as immunosuppressive (IS) agent along with mean dose of 46.6±13.7 (0.86±0.23mg/kg/day) prednisolone. Additionally, Rituximab & IVIg were used in 2 patients each respectively. 3 months after initiation of immunosuppression 26/50 (52%) patients on follow-up were in remission and 39/47 (82.9%) patients attained remission at 6 months. Mean time to achieve remission was 151.1±86.3 days. All IS agents were equally effective in inducing remission at 3 and 6 months and showed statistically similar BVAS reduction at 3 and 6 months from baseline (t test & chi-square test). 7 (11.2%) deaths noted in the cohort at their respective last visit during 195.3 patient years cumulative follow up. Multiple regression analysis showed that at baseline, presence of PNS involvement, eosinophilia, thrombocytosis, higher BVAS score and higher steroid requirement were predictors of persistently active vasculitis and absence of eye involvement and higher hemoglobin % at baseline were predictors for remission, at 3 months ($p<0.05$). 4/50 (8%) patients had relapse of vasculitic symptoms. 2 and 5 year survival rates were 96.2% and 83.9% respectively

Conclusions: Our cohort of Asian Indian RV was compartively younger with lesser RA duration, less percentage of ever-smokers, lesser rheumatoid nodule prevalence, higher PNS involvement with better survival/mortality rates compared to published literature. All IS agents showed equal rates of BVAS remission & BVAS reduction at 3 and 6 months of treatment

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THU0323 THE INCIDENCE RATES OF ANCA-ASSOCIATED VASCULITIDES IN NORTHERN GERMANY (SCHLESWIG-HOLSTEIN) REMAIN STABLE BETWEEN 1998 TO 2014

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Objectives: To assess the incidence of ANCA-associated vasculitides (AAV) in a stable population by registering all newly diagnosed patients in a large region in Northern Germany. To investigate the epidemiology and pattern of AAV over seventeen years in a prospective study.

Methods: Between 1/1/98 and 31/12/14 all newly diagnosed cases of AAV (Granulomatosis with polyangiitis (GPA), microscopic polyangitis (MPA), eosinophilic granulomatosis with polyangitis (EGPA)) as defined by the Chapel Hill Consensus Conference 1992 were identified in a large mixed rural/urban northern German region consisting of 2,83 million habitants in a population-based prospective study. The following sources were explored on a regular basis: (a) departments of all hospitals, including their outpatient clinics; (b) all departments of pathology, and (c) the reference immunological laboratories serving the catchment area. All cases were re-evaluated by the authors.

Results: Within the observation period of seventeen years 551 patients were identified as having a new diagnosis of AAV in the catchment area, the incidence rate was 12 (range 8 -16)/million/year. GPA represents 70% of the AAV with an incidence rate of 7.9 (range 6–12)/million/year (95% CI 1;11 - 5*;19*), MPA with an incidence rate of 2.4 (range 1–5)/million/year (CI 0;3 – 1;9) and EGPA 1.1 (range 0–2)/million/year (CI 0;4).

Incidence rates of GPA were on average three times higher than those of MPA and seven times higher than CSS. The incidence rates were stable between 1998 and 2014, our observations did not reveal significant differences. Gender differences were not observed. People diagnosed with MPA were on average 66.4 years old.

Conclusions: Results of our unique population based vasculitis register over seventeen years assessing the incidence of AAV in a region of 2.83 million habitants in northern Germany demonstrate stable incidence rates for AAV. Variances between years have been observed, but no pattern or periodicity. Compared to other studies investigating small regions or referral centers the incidence rates for AAV were comparable to the incidence rates observed in Norway, Sweden and the United States, lower than those in United Kingdom, but higher than in Spain and Vilnius. The incidence rates of MPA are significantly higher in Japan, whereas GPA and CSS have rarely been diagnosed.

Disclosure of Interest: None declared

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THU0324 NEUTROPHILS IN GIANT CELL ARTERITIS: MONITORING DISEASE PROGRESSION DURING THERAPY TAPERING

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Background: Giant Cell Arteritis (GCA) represents a medical emergency due to risk of permanent vision loss and/or cerebrovascular insults. Polymyalgia rheumatic (PMR) frequently coexists with GCA[1]. No diagnostic or prognostic markers are yet known for GCA and predicting relapses during steroid therapy tapering is difficult. Biomarkers, such as serum amyloid A (SAA) and interleukin-6 (IL-6) provide added value for monitoring inflammation and a recent investigational study indicated the potential use of neutrophils and their surface markers in GCA pathogenesis[2].

Objectives: To determine the percentage of neutrophils, T and B cells, and the mean fluorescence intensity (MFI) of L-selectin (CD62L) and integrin αM (CD11b) on $CD16^+$ neutrophils in peripheral blood of newly diagnosed, untreated GCA and PMR patients vs. healthy controls (time 0) and at GCA follow-up (7, 30 and 90 days after therapy).

Methods: Flow cytometry of stained, lysed and fixed whole blood[2] was performed in 10 GCA patients (6 followed longitudinally), 7 PMR patients and 5 healthy controls (7-colour immunophenotyping kit, Miltenyi). Levels of SAA and IL-6 were measured in sera of GCA patients (n=6) using nephelometry and ELISA, respectively.

Results: Percentage of neutrophils was significantly higher at time 0 in GCA and PMR patients compared to healthy controls. Expression of both CD62L and CD11b on $CD16^+$ neutrophils was also higher in GCA and PMR patients, as compared to healthy controls. Longitudinally, GCA patients showed an initial decrease in percentage of neutrophils at day 7 in comparison with time 0, increasing on days 30 and 90, while both T ($CD4^+$) and B cells exhibited a significant elevation in % at day 7, with a decline at days 30 and 90. The MFI of neutrophil CD62L steadily decreased from day 0 (85.80±45.45) to day 7 (71.99±29.93) and day 30 (57.61±42.17), while showing a substantial increase on day 90 (88.54±60.35). CD11b expression diminished initially and remained reduced on day 90. Levels of SAA and IL-6 declined sharply from day 0 to 7 (>10-fold drop) and day 30, with gradual elevation of both on day 90.

Conclusions: Neutrophil CD62L may represent a good surface marker for monitoring disease progression following glucocorticoid tapering. SAA and IL-6 exhibit a sharp decrease at early time points, increasing at day 90. In the future, a larger, longer study of neutrophils and their CD62L expression could support clinicians in their decision when and how to re-evaluate therapy in GCA patients.

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THU0325 RELATIONSHIP OF THE INITIAL SYMPTOMS TO THE DIAGNOSIS DELAY AND POOR PROGNOSIS IN PATIENTS WITH TAKAYASU ARTERITIS

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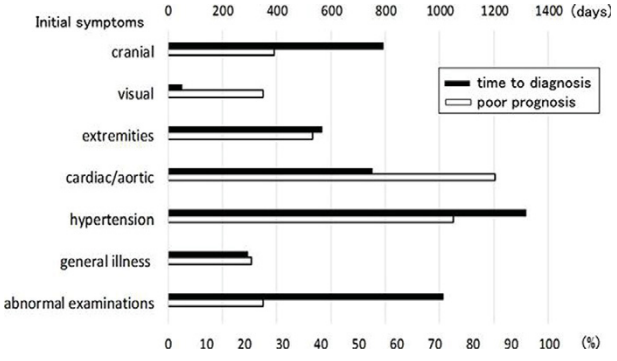
Background: Clinical courses of Takayasu arteritis are of great variety. Its non-specific initial symptoms can cause the delay in diagnosis and lead to poor prognosis. However, the symptoms of very early phase of Takayasu arteritis and their effects on diagnosis delay and prognosis are unclear.
Objectives: To clarify the characteristics of initial symptoms of Takayasu arteritis, the delay in diagnosis, and its relationship with prognosis.
Methods: The consecutive patients with Takayasu arteritis with analysable information in our hospital were enrolled. Initial symptoms, laboratory findings before diagnosis, the duration from symptom onset to diagnosis, and prognosis were investigated. Initial symptoms were divided into 7 groups; cranial symptoms (dizziness, syncope, headache, neck pain, hemi-paralysis, and jaw claudication), visual symptoms (vision loss and visual field loss), extremities symptoms (claudication of extremities, coldness of limbs, bilateral difference in blood pressure, and limb numbness), cardiac/aortic symptoms (dyspnea on exertion, palpitation, and chest compression), hypertension, general illness (fever, fatigue, body weight loss, and arthralgia), and abnormal medical examinations (heart murmur, bruit on any extremities, and abnormal chest X-ray). Poor outcome was defined as a cardiovascular surgery or death.
Results: A total of 98 patients were enrolled with the median observation period of 12.1 years (range, 1 month to 59 years). Eighty-seven (88.7%) were female and the mean age at diagnosis was 37 years. The mean duration from the initial symptoms to diagnosis was 600 days. Thirty-four (34.7%) patients had poor outcomes. The initial symptoms before diagnosis were cranial symptoms in 25%, visual symptoms in 5%, extremities symptoms in 20%, cardiac/aortic symptoms in 7%, hypertension in 9%, general illness in 26%, and abnormal medical examinations in 8%. The duration from symptom onset to diagnosis was 792, 52, 567, 752, 1318, 293 and 1014 days (p=0.10), respectively; the rate of poor outcome was 28, 25, 38, 86, 75, 22, and 25% (p=0.010), respectively. The duration from symptom onset to diagnosis was longer in the patients with poor prognosis than those without (837 vs 493 days, p=0.06). The patients with extremities symptoms were younger (29.6 vs 38.9 years, p=0.026) than those with the other symptoms, and patients with cardiac/aortic symptoms were older (49.7 vs 36.3 years, p=0.044). The patients without general illness showed lower levels of C-reactive protein (3.26 vs 5.86 mg/dl, p=0.048), erythrocyte sedimentation rate (38.3 vs 66.6 mm/h, p=0.01), and platelet counts ($29.4*10^4$ vs $34.0*10^4/\mu l$, p=0.09), and resulted in poor outcome more frequently than those with (42.6 vs 22.6%, p=0.058).



Conclusions: The initial symptoms of Takayasu arteritis before diagnosis varied widely, and majority of them were non-specific. Lacking inflammatory signs were related with delayed diagnosis and poor prognosis.
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THU0326 SHORT AND LONG-TERM FOLLOW-UP WITH ADALIMUMAD IN REFRACTORY UVEITIS ASSOCIATED TO BEHÇET'S DISEASE. MULTICENTER STUDY OF 74 PATIENTS

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Objectives: To evaluate the efficacy of adalimumab (ADA) in short and long term follow-up in refractory uveitis of Behçet's disease (BD)
Methods: Multicenter study. Ocular inflammation was evaluated according to "SUN working Group" (Am J Ophthalmol 2005;140:509–516), and the macular thickening with OCT. A comparison was carried out between baseline, and follow-up visits. Results are expressed as mean±SD or median [IQR]. Continuous variables were compared with Wilcoxon test.
Results: We studied 74 patients/132 affected eyes (39M/35W); mean age 38.7±11.3. The ocular pattern was panuveitis (n=45), posterior uveitis (n=14), anterior uveitis (n=14) and intermediate uveitis (n=1). Before ADA, systemic treatment with corticosteroids, iv metilprednisolone (n=23), Cyclosporin A (58), azathioprine (33), metotrexate (31) and other drugs (28) was used. The dose of ADA was 40 mg/2 weeks/ sc in monotherapy (n=22) or combined (n=52). Most patients showed a rapid and progressive improvement (TABLE). The 24 patients (37 affected eyes) with CME showed a significant improvement.
ADA was optimized in 23 (31.1%) that were in remission for 15.3±9 months. Interval of administration was increased to 3 (n=6), 4 (13), 5 (1), 6 (1) and 8 weeks. After a mean follow-up of 13.0±9.7 months after optimization, 21 patients were stable and 2 had a severe flare. In 4 patients ADA was stopped after 35.2±9.3 months in remission. The main adverse effects observed were lymphoma (n=1), pneumonia (1), and 2° bacteriemia by E. Coli (1)

TABLE. EVOLUTION OF OCULAR PARAMETERS

	Basal	2nd week	1st month	6th month	1st year	2nd year	3rd year
VA (mean±SD)	0.5±03 n=146	0.6±0.3* n=144	0.6±0.2* n=143	0.7±0.2* n=120	0.8±0.2* n=112	0.8±0.2* n=89	0.8±0.2* n=41
Cells in the anterior chamber (median [IQR])	1 [0-2] n = 144	0 [0-2]* n= 142	0 [0-1]* n=142	0 [0-0]* n= 120	0 [0-0]* n = 116	0 [0-0]* n = 92	0 [0-0]* n = 42
Vitritis (median [IQR])	1 [0-2] n= 145	1 [0-1]* n= 143	0 [0-1]* n= 143	0 [0-0]* n= 124	0 [0-0]* n= 116	0 [0-0]* n= 92	0 [0-0]* n= 42
Retinal vasculitis (% affected eyes)	53.3% n= 144	43.2%** n= 141	36.4%* n= 142	11.4%* n= 123	4.1%* n= 116	0.6%* n= 92	0.6%* n= 42
OCT (μ) (mean±SD)	345.5±134.5 n= 82	341.1±108.6* n= 65	309.8±8* n= 64	268.1±38.4* n= 52	254.4±38.5* n= 55	259.5±41.2* n= 42	254.5±48.9* n= 20

*p<0.05
Abbrev: VA= visual acuity

Conclusions: ADA was effective in short and long-term follow-up in refractory uveitis associated to BD. Optimization or even suspension of ADA is possible.
Disclosure of Interest: None declared
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THU0327 ASSOCIATION BETWEEN THE TC/HDL RATIO AND DISEASE ACTIVITY IN PATIENTS WITH TAKAYASU ARTERITIS

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Background: Accelerated atherosclerosis has become the main cause of morbidity in patients with autoimmune diseases such as RA and SLE [1]. The Cholesterol/High-density Lipoprotein Cholesterol (TC/HDL-C) ratio is a high discriminatory power index for coronary heart disease. A high TC/HDL-C ratio has been intensively used as a predictor of CVDs [2]. EULAR Task Force recommended that the TC/HDL-C ratio should be regarded as an important prognostic indicator for future cardiovascular disease (CVD) in patients with rheumatoid arthritis (RA) [3]. However, the relationship between the TC/HDL-C ratio and disease activity of Takayasu arteritis (TAK) is unclear.
Objectives: To investigate changes in the TC/HDL-C ratio and to evaluate the relationship between the TC/HDL-C ratio and disease activity of TAK.
Methods: A retrospective study of 103 patients with TAK and 73 healthy controls was performed. We compared the triglyceride (TG), TC, low-density lipoprotein cholesterol (LDL-C), HDL-C and TC/HDL-C ratio between patients and healthy controls, and we analyzed correlations between the lipid parameters and indexes