SAT0525 AN UPDATE ON PULMONARY ARTERY INVOLVEMENT IN BEHCET'S SYNDROME: MORE PULMONARY ARTERY THROMBOTIC DISEASE AND A BETTER OUTCOME

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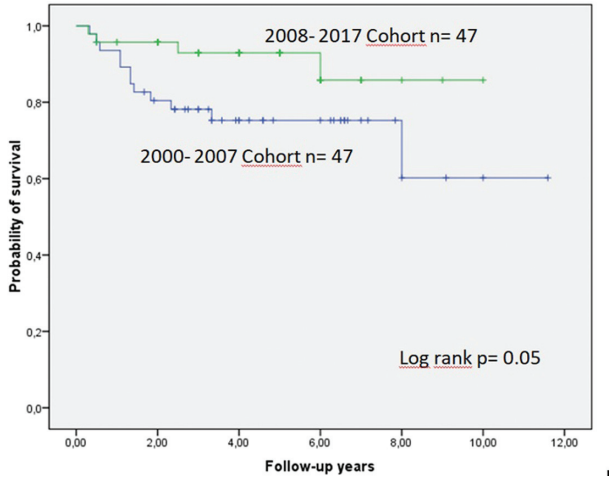
Background: Pulmonary artery involvement (PAI) is the most common form of arterial involvement in Behçet's syndrome (BS) and is a well-known cause of mortality and morbidity. A previous survey[1] by our group had analysed the clinical characteristics and outcome of 47 pts with PAI registered between 2000–2007 and shown that: 1.the overwhelming male predominance was decreasing; 2. 1/4th of the pts had isolated pulmonary artery thrombosis (PAT); and 3. the mortality rate was 26% after a mean follow-up of 7 years. Recently we had the impression that female/male ratio was perhaps increasing, we are becoming to see more pts with isolated PAT and that we started to use more biologics.
Objectives: This survey was done to look at these assumptions formally in a recent group of BS pts with PAI.
Methods: We reviewed the records of about 3390 pts with BD who were registered at our multidisciplinary BS clinic between $^{Jan\ 2008}$ and $^{Jan\ 2018}$ From this group we identified 47 (42 M/5 F) pts who were diagnosed with PAI and recorded all information regarding clinical characteristics, outcome, radiological studies and medical or surgical treatment.
Results: The prevalence of pts with PAI decreased from 1.9% to 1.4% in the recent cohort. The M/F ratio, the mean age at the onset of PAI were similar across 2 cohorts. The frequencies of other vascular involvement were almost similar to that observed in the previous cohort. However, there were more pts with neurological disease (parenchymal) in the recent cohort. As usual, PAT or PAA were mostly bilateral and involved descending lobar arteries. On the other hand, types of PAI involvement at presentation had changed substantially: those with isolated PAT reached a share of 45%. Forty-five (96%) pts received cyclophosphamide (Cy) pulses for a mean of 6±4 courses, which was significantly shorter compared to that observed in the previous cohort. A total of 23 (49%) pts received infliximab because of relapsing course, side effects or unresponsiveness to Cy for a mean follow-up of 8±4 mo while only 2 pts received anti-TNF's in the older cohort. 4 pts had lung surgery. These were lobectomies in 3 pts due to giant rapidly progressing aneurysms and cavitectomy in 1. Bronchial artery embolization was done in 3 pts because of refractory hemoptysis. ,$^{By\ Jan\ 2018}$ the outcome of information was available on 45/47 pts: 4 pts (all male) (8%) had died, 2 were lost to follow-up after 12 and 16 mo of follow-up and the remaining were alive after a median follow-up of 5 [IQR:3–9] years. The causes of deaths were massive hemoptysis in 3, severe pulmonary hypertension in 1. As shown in the figure, the survival has improved significantly in the recent yrs.



Conclusions: The surveys of 2 consecutive cohorts showed that the prevalence of PAI perhaps mildly decreased, isolated PAT type of involvement was with considerably higher frequency and the outcome was getting better. Cy is still the first agent in these pts however its duration of use became much shorter and anti-TNF's mainly infliximab was used in about half of the cohort. The survival seems to have improved significantly. This could have been due to a decreased severity of the type of PAI, with isolated PAT becoming the most frequent type, or a better management.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.6675

SAT0526 IS RELAPSE RATE OF GIANT CELL ARTERITIS IN REAL-LIFE EXPERIENCE LOWER THAN IN THE CONTROLLED TRIALS? RESULTS OF A RETROSPECTIVE, MULTI-CENTRE COHORT STUDY

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Objectives: Corticosteroids (CS) are accepted as the standard first-line treatment for giant cell arteritis (GCA). However, controlled trials of tocilizumab and abatacept demonstrated relapse rates of up to 70%–80% in patients on CS-only protocols in 12–24 months. Though level of evidence is low and not suggested by guidelines (except for methotrexate), conventional immunosuppressives (ISs) are also commonly used. We aimed to assess the relapse rates in patients with GCA in routine practice, retrospectively.
Methods: We assembled a retrospective cohort of patients with GCA from Turkey. All data was abstracted from records. Relapse was defined as any new manifestation or increased acute-phase response leading to the change of the CS dose or use of a new therapeutic agent by the treating physician.
Results: The study included 156 (F/M: 95/61) patients with GCA (table 1). The mean age at disease onset was 67.8±9.1 years. Polimyalgia Rheumatica was also present in 48 (30,8%) patients. Diagnosis was proven histopathologically in 99 patients.All patients received 1 mg/kg/day CS for remission induction, additional CS pulses were given to 36 (23.1%) patients. Conventional ISs including methotrexate and azathioprine were used in 89 (56.1%) and 26 (16.6%) patients respectively, while 10 (6.4%) patients received biologic treatments (8 tocilizumab,2 etanercept). Fourty-four (28.2%) patients used only CS during follow-up. Follow-up of at least 6 months was available for 132 patients, and median follow-up duration was 35 (6–268) months. Relapses occurred in 27 (20.5%) patients during follow-up. Mortality rate was 7.5% (n=10) during follow-up. VDI score was 2.4±1.7. Main causes of damage were related to CS treatments such as cataract, osteoporosis and diabetes mellitus.

	Giant Cell Arteritis (n=136)
Manifestations of systemic inflammation, n/156 (%)	
Anemia (<12 mg/dl for female,<13 mg/dl for male)	94 (60.2 %)
Erythrocyte Sedimentation Rate (mm/hour) *	84.6 ± 30.1
C-reactive protein (mg/l)*	84.8 ± 65.3
Malaise	132 (84.6%)
Weight loss	72 (46.1%)
Fever	35 (22.4%)
Stiffness and/or pain muscles and joints	48 (30.8 %)
Manifestations of Vascular Ischemia , n /156 (%)	
Headache	144 (92.3%)
Scalp Tenderness	78 (50 %)
Jaw Claudication	57 (36.5 %)
Ocular Symptoms	72 (46.2 %)
Extremity Claudication	14 (8.9 %)
Neurological Manifestations	14 (9 %)

Conclusions: In this first multi-centre series of GCA from Turkey, we observed that only one fifth of patients had relapses during a mean follow-up of 35 months. This lower relapse frequency suggests a different clinical spectrum in routine

practice compared to patients included in controlled trials. Our results also suggest that there is a clear need for a steroid sparing agent in patients with GCA, that is a older aged population prone to CS side effects.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.3983

SAT0527 RITUXIMAB PRESCRIPTION PATTERNS AND EFFICACY IN THE INDUCTION TREATMENT OF ANCA-ASSOCIATED VASCULITIS IN A BELGIAN MULTICENTRIC COHORT

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Background: The RAVE trial has revolutionised the induction treatment of ANCA-Associated Vasculitis (AAV, including GPA, granulomatosis with polyangiitis and MPA, microscopic polyangiitis), by demonstrating non-inferiority of rituximab compared with cyclophosphamide[1].
Objectives: To study AAV patients' characteristics, rituximab prescription practices and efficacy in AAV induction treatment in 4 Belgian university hospitals. The patient population, selected according to the Belgian reimbursement criteria, is relatively homogeneous and comparable to the one of RAVE trial.
Methods: 57 patients, receiving rituximab as AAV induction therapy since May 2014, were enrolled in an observational retrospective multicenter trial involving 4 Belgian university hospitals. We have focused on the type of AAV (GPA/MPA), ANCA specificity (anti-PR3/MPO), prescriber's speciality, used reimbursement criteria, organ involvements, severity of the flares (according to BVAS-WG definition) and finally rituximab efficacy in AAV induction treatment by considering the RAVE primary (complete remission without prednisone) and secondary (complete remission with prednisone <10 mg, rates of relapses) outcomes at 6, 12, 18 and 24 months.
Results: The most frequent subtype of AAV was GPA (84%). The main indication was relapsing disease (54.4%), followed by contra-indication to cyclophosphamide (38.6%). 66.7% of the patients reached complete remission with prednisone <10 mg at 6 months, and 55.3% at 12 months, 40% at 18 months, 25% at 24 months respectively. In the "severe disease" subgroup, 73% reached complete remission with prednisone <10 mg at 6 months, 58.8% at 12 months, 50% at 18 months and 32% at 24 months. The rates of complete remission without steroids were very low at 6, 12, 18 and 24 months (between 0%–6%) but our patients were not asked to follow a glucocorticoid tapering scheduled for complete withdrawal of prednisone after 6 months and were usually maintained under low-dose prednisone. Relapse rates were high between 18 and 24 months both in the total group and in the severe disease subgroup (due to the fact that rituximab is not reimbursed for maintenance treatment in Belgium). The subtype of ANCA was not predictive of the risk of relapse.
Conclusions: Our results confirm – in a "real-life" cohort of patients selected according data of RAVE trial – those of RAVE regarding complete remission rates at 6 months with prednisone 10 mg/j. The high prevalence of relapses – in particular after 18 months – underlines the need to optimise maintenance treatment after an induction treatment with rituximab.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.1881

SAT0528 EFFECTIVENESS OF REMISSION-INDUCTION THERAPY WITH CONCOMITANT CYCLOPHOSPHAMIDE AND GLUCOCORTICOID FOR MICROSCOPIC POLYANGIITIS AND GRANULOMATOSIS WITH POLYANGIITIS IN JAPAN: A PROPENSITY SCORE MATCHED ANALYSIS OF TWO NATIONWIDE PROSPECTIVE COHORT STUDIES

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Background: Although concomitant use of cyclophosphamide (CYC) with glucocorticoids (GC) is considered to be one of the standard remission-induction therapies for antineutrophil cytoplasmic antibody (ANCA) associated vasculitis over 30 years, there are few reports about clinical efficacy or effectiveness of CYC.
Objectives: To evaluate effectiveness and safety of concomitant CYC as remission induction therapy in Japanese patients with microscopic polyangiitis (MPA) and granulomatosis with polyangiitis (GPA) using data sets from two nationwide prospective cohort studies.
Methods: Newly diagnosed MPA and GPA patients treated with GC with or without CYC for remission-induction therapy were enrolled. The patients treated with other immunosuppressants or plasma exchange were excluded. A propensity score for the use of CYC was estimated using age, types of AAV, serum creatinine level, Birmingham Vasculitis Activity Score (BVAS), and initial GC dosage at baseline. After propensity score matching at 1:1, remission, overall survival, and end-stage renal disease (ESRD)-free survival rates, Vasculitis Damage Index (VDI), and incidence of serious infection within 6 months were compared between patients treated with and without concomitant CYC.
Results: Of enrolled 327 patients, concomitant CYC was used in 119 (36%) patients during the initial 3 weeks of treatment. After propensity score matching, 95 patients with concomitant CYC (CYC group) and 95 controls (non-CYC group) were selected. Demographics, baseline characteristics and treatments were balanced between the two groups except for myeloperoxidase ANCA positivity (**Table**). The remission within 6 months was achieved in 85% in both groups. The survival and ESRD-free survival rates were also similar between the two groups (log-rank test; p=0.77 and 1.0, respectively). Median VDI at the time of last observation did not differ between the two groups (CYC, 3 [interquartile {IQR}: 2–4]; non-CYC, 2 [IQR:1–3], p=0.26). The accumulated GC dosage of the CYC-group from 3 to 24 months was lower than the non-CYC group, the GC-related damage did not differ (CYC, 1 [IQR: 0–2]; non-CYC, 0 [IQR:0–2], p=0.69).

Table. Comparison of patients treated with concomitant cyclophosphamide and with glucocorticoid alone

Variables at baseline and treatments	Cyclophosphamide users (n=95)	Non-users (n=95)	*p*-value
Male/female, n	51/44 (46%)	60/35	0.24
Age, years	70 (65–78)	71 (62–75)	0.92
GPA/MPA	26/69	27/68	1.0
Myeloperoxidase ANCA, n (%)	78 (82)	89 (94)	0.02*
Proteinase-3 ANCA, n (%)	17 (18)	8 (8)	0.15
Serum creatinine, mg/dL	1.2 (0.8–3.2)	1.1 (0.7–2.5)	0.39
C-reactive protein, mg/dL	7.5 (2.3–12.6)	7.2 (2.1–11.7)	0.35
BVAS	15 (12–20)	16 (12–21)	0.99
Interstitial lung disease, n (%)	38 (40)	37 (49)	0.55
Glucocorticoid (mg/kg/day) [d]	0.83 (0.72–0.97)	0.83 (0.67–0.99)	0.89

Values expressed as a number of patients (%) or median (interquartile). *p<0.05.

Conclusions: In Japanese patients with MPA and GPA, concomitant CYC could not show any benefits on clinical outcomes within 24 months. Dosage and treatment duration of CYC, as well as tapering methods of GC, could be confounding factors. Longer observation may be necessary to confirm the effectiveness of CYC as GC sparing agent.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.1689