Syndrome International Study Group Criteria) and 30 healthy controls were included. Cases who were not able to tolerate posturography, with a history of orthopaedic surgery and with known balance problems were excluded. The age, gender, disease duration, anamnesis of falls (last 12 months), fear of falling (yes/no) and drugs used were recorded. Also disease activity (with Behçet's Disease Current Activity Form; BDCAF) and fall efficacy (with Tinetti's Falls Efficacy Scale) were evaluated. Fall risk analysis was performed by Tetrax Interactive Balance System which is a computerised posturography device. By this method, fall risk is obtained as a numeric value (0–100) and as ranges indicating low, moderate or high risk of fall. We investigated age, gender, disease duration, fall anamnesis, fear of falling, drug usage, fall efficacy, disease activity as possible related factors to fall risk. Mann-Whitney U, chi square and Spearman correlation tests were used for statistical analysis.

Results: The mean ages of the cases and controls were 35,17±9,48 and 33,03±11,813 years, respectively. Symptom duration of the cases was 7,70±6,613 years. 7 cases (23,3%) had anamnesis of falls during the last 12 months, whereas only 8 control (26,7%) had this anamnesis ($p>0.05$). Fear of falling was reported by 43,3% of the cases and 40% of the controls ($p>0.05$). There was no significant difference between case and control groups in terms of FES-I scores (15,97±9,257 and 12,53±30,48 respectively; $p=0.599$). With the computerised system used, significantly higher fall risk results were recorded in patients with BD than the controls (50,40±24 710 and 23,13±11,811, respectively; $p<0.001$). Low, moderate and high fall risks were recorded as 30%, 33,3% and 36,7% of the cases and 70%, 30% and 0% of the controls and this distribution was also significantly worse in cases than controls ($p<0.05$). No significant correlation was found between fall risk and other factors including age, gender, disease duration, fall anamnesis, fear of falling, drug usage, disease activity except arthralgia which was significantly correlated with fall risk assessment in cases ($p<0.05$).

Conclusions: With an objective computerised technique, fall risk was found to be higher in cases with BD than controls in our study. The higher fall risk in these patients seems to be affected by joint involvement. An increased awareness of the potential fall risk and future studies investigating the possible coexisting balance problems in BD may contribute to the management.

Disclosure of Interest: None declared

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SAT0538 AORTIC INVOLVEMENT IN RELAPSING POLYCHONDRIITIS: A SYSTEMATIC REVIEW

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Background: Aortic involvement (AI) is an important complication of relapsing polychondritis (RP). Current literature is based on case reports and small case series.

Objectives: To delineate the clinical characteristics and outcome of AI in RP through a systematic literature review (SLR).

Methods: The SLR covered all English articles retrieved with relevant keyword combinations listed in PubMed until October 2017. Initially the titles and abstracts were screened by two investigators and articles considered to be relevant (involvement of the aorta and the aortic valve) were identified. Data extraction was done by the same investigators.

Results: The SLR revealed 352 papers of which 162 were discarded at the first step and a further 114 after full reading. After excluding 5 articles reporting on the same patient, we finally had 71 papers reporting 97 patients. The sex distribution was identifiable in 79 patients of whom 39 were men and 40 were women. The median age at the first symptom of RP was 31.5 years [IQR:24–43 years], the median age at RP diagnosis was 36 years [IQR: 28–43 years]; median age at the diagnosis of AI was 37 years [IQR:29–49 years]. Median duration from first RP diagnosis to AI diagnosis was less than 1 year [Range:0–21]. AI was the presenting symptom in 3 patients.

Seventy patients (73%) had involvement of thoracic aorta and 16 (23%) had involvement of the abdominal aorta. Other involved arteries were: coronary (8 patients), subclavian (5 patients), renal (3 patients), iliac (3 patients) and others (8 patients). Two patients had neurologic and renal involvement, respectively.

The most common symptoms were dyspnea (40%), followed by chest pain (13%), abdominal pain (13%) and fever (9%).

The diagnosis of AI was made during surgery in 6 patients and with different radiologic methods ranging from chest x-ray to PET-CT.

All patients excluding 1 received corticosteroids either alone or in combination with classical immunosuppressives (cyclophosphamide, azathioprine, methotrexate, mycophenolate-mofetil) or biologics (infliximab, tocilizumab, adalimumab).

The majority of the patients (53/87 patients (61%) underwent surgery including aortic graft replacement, coronary by-pass, aortic valve or mitral valve replacement operations.

16 patients died after a median follow up of 24 [IQR:11–43] months. The reasons for deaths were mainly vascular (acute myocardial infarction, heart failure,

cardiovascular operation, abdominal aorta dissection, aortic rupture and acute aortic valvular dysfunction).

Information on follow-up was known in 30 patients for a median of 18.5 months [IQR 10–48 months].

Conclusions: AI is less frequently recognised but prognostically important complication of RP. Thoracic aorta is the most frequently involved site. Surgery is needed in the majority of patients. Medical treatment is empirical and is based on glucocorticoids and immunosuppressives including biologic agents. Despite treatment, mortality is high (18% during a median of 24 months).

Disclosure of Interest: None declared

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SAT0539 THE FREQUENCY OF RECURRENT ORAL ULCERS IN FAMILY MEMBERS OF PATIENTS WITH BEHÇET'S DISEASE

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Background: It is unknown whether the observed geographic disparities in Behçet's Disease (BD) occurrence reflect primary genetic susceptibility or environmental influences within specific populations. Family aggregation studies may help to discriminate between environmental and genetic components but there are no large family surveys.

Objectives: The aim of this study was to contribute to a better understanding of the genetic aspect of familial aggregation studies in Turkey.

Methods: The study group consisted of siblings and children of 133 unrelated consecutive patients followed up at the BD outpatient clinic in FSM Hospital, Istanbul. Surveyed individuals were interviewed via telephone regarding if they had ever suffered from recurrent oral ulcerations (ROU). Children less than ten years old were surveyed via discussion with their parents. Symptoms and signs of Behçet's disease in family members were also interviewed via telephone. Subjects experience ROU, at least three episodes in one year, were invited to attend further examination, and also were asked whether anyone in their family had BD.

Results: Total 133 patients with BD (86 F, 47 M, respective mean ages: 39.9±11.7, 43.9±10.4 years) had 271 children (137 F, 134 M) and 642 siblings (319 sisters, 323 brothers, respective mean ages: 42.1±13.6, 40.7±14.4 years). All 642 siblings were contacted by telephone; 62 siblings (33 F, 29 M, respective mean ages: 17.9±9.8, 18.2±9.9 years) and 84 children (63 F, 21 M) had positive ROU history. All probands were invited, only 36% family members attended. Apart from patients with BD, 146 family members had ROU (14%). The estimated ROU rate among siblings and children was 9.6%, and 31% respectively. Among members there were 13 patients with BD (2 fathers, 1 mother, 2 children, 3 brothers, 5 sisters). Eleven of these were diagnosed at another centre. We identified 2 additional patients who met ISG criteria during the survey. Only five spouses had ROU (0.5% of members).

Conclusions: In a study from Istanbul, ROU was reported as 9.5% in the general population. In our previous study, that was conducted in an area that was reported as having the lowest prevalence rate of BD, this was estimated as 2.1% in the general population. The ROU rate in the current study for total family members was 16%, and 31% for children, which was higher than in the general population. The rate of patients with BD in the studied cohort, apart from previously diagnosed patients, were 140 per 10, 000 while the reported prevalence rates changed regionally between 20 per 10, 000 and 42 per 10, 000 in Turkey. These findings suggest that BD risk is primarily influenced by genetics rather than environmental conditions. Familial aggregation supports a genetic background for BD

Disclosure of Interest: None declared

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SAT0540 TREATMENT WITH INTRAVENOUS IMMUNOGLOBULIN IN THE VASCULITIS ANCA POSITIVE. 27 CASES STUDIED IN A SINGLE REFERENCE CENTRE

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Objectives: Intravenous immunoglobulins (IGIV) is a therapeutic alternative in vasculitis ANCA+ specially in cases of refractory or superinfection. We study the efficiency and safety in the short and long term of the IGIV in the vasculitis ANCA.

Methods: Descriptive and observational study of 27 patients with vasculitis ANCA from a reference tertiary hospital. We analysed the treatments received, the clinical and analytical variables and the indexes of activity and evolution (TABLE). Birmingham Vasculitis Activity (BVAS) was the activity index used, and for prognosis Five Factory Score (FFS). Results are indicated as mean ±SD when

there is a normal distribution, or as median [25–75 IQR] when there is an usual one.

Results: 27 patients were analysed (14 W/13 M). At the beginning of the IGIV the average age was 57.8±15.98 and the vasculitis average development was 1.29 ±0.68. The vasculitis ANCA subtypes were: **a)**granulomatosis with polyangiitis (n=14; 51.8%), **b)**microscopic polyangiitis (n=9; 33.3%), **c)**eosinophilic granulomatosis with polyangiitis (n=2; 7.4%), **d)**pulmonary-renal syndrome with ANCA positive (n=1; 3.7%) and **e)** indeterminate vasculitis ANCA positive (n=1; 3.7%). Previously to the treatment with IGIV, apart from steroids, they also received: cyclophosphamide (n=12, 44.4%), metotrexate (n=6, 22.2%), infliximab (n=5, 18.5%), rituximab (n=4, 14.8%), azathioprine (n=3, 11.1%), mycophenolate (n=3, 11.1%) and plasmapheresis (n=1; 3.7%).

Refractoriness (n=18) and suspicious of infection (n=9) were the reasons for the application of IGIV. The IGIV guideline was 0.4 g/kg/day for 5 consecutive days. 66.6% received methylprednisolone IV concomitant (0.5–1 g/day for 3 days). After a follow-up of 80±68.4 months we observed clinical and analytical improvement, as well as, in the activity indexes (**TABLE**). The majority of the side effects were lower and IGIV was suspended in just one patient due to severe effects of congestive heart disease.

	Initial (n=27)	1-2 weeks (n=25)	1 month (n=25)	6 months (n=25)	1 year (n=25)	2 years (n=25)	Last visit (n=25)
Glaxi, n (%)							
Constitutional symptoms	24 (88.9%)	4 (16.0%)	4 (16.0%)	1 (3.7%)	0 (0%)	1 (3.7%)	0 (0%)
Ocular	3 (11.1%)	0 (0%)	0 (0%)	2 (7.7%)	1 (3.7%)	0 (0%)	0 (0%)
ORL	7 (25.9%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	2 (7.4%)	3 (11.1%)
Pulmonary	10 (36.4%)	5 (20.0%)	4 (16.0%)	7 (28.0%)	6 (22.2%)	4 (16.0%)	3 (11.1%)
Articular	4 (14.8%)	0 (0%)	0 (0%)	1 (3.7%)	1 (3.7%)	0 (0%)	1 (3.7%)
Renal	25 (92.2%)	18 (69.6%)	19 (76.0%)	12 (48.0%)	13 (52.0%)	10 (37.7%)	12 (48.0%)
Cutaneous	4 (14.8%)	1 (3.7%)	1 (3.7%)	2 (7.7%)	2 (7.7%)	1 (3.7%)	0 (0%)
SNP	5 (18.5%)	3 (11.1%)	2 (7.4%)	1 (3.7%)	0 (0%)	0 (0%)	0 (0%)
SNC	2 (7.4%)	1 (3.7%)	4 (16.0%)	2 (7.7%)	2 (7.7%)	2 (7.7%)	0 (0%)
Abdominal	4 (14.8%)	1 (3.7%)	1 (3.7%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Laboratory, mean±SD							
VSG (mm/h)	69.14 ^a 36.55	63.22 ^a 30.26	39.40±35.97	33.19 ^a 27.63	25.72 ^a 21.16	23.66±29.09	19 ^a 15.56
PCR (mg/dl)	12.34 ^a 10.44	2.83±5.15	1.59 ^a 3.66	6.02 ^a 22.39	10.56±22.39	0.76 ^a 1.30	5.32 ^a 20.94
Creatinine (mg/dl)	1.50 ^a 1.08	1.59 ^a 1.26	1.38 ^a 0.82	1.36±0.63	1.24±0.63	1.33 ^a 0.83	1.19±0.582
Urea (mg/dl)	74 ^a 44.21	81.25 ^a 55.19	73.64±41.91	55.3 ^a 23.43	57.85 ^a 30.64	56.95 ^a 40.61	51.23±29.31
Proteinuria 24h (mg)	843.77 ^a 1082.81	664.27±1126.35	867.28±1211.74	474.13±844.30	335.47±374.66	258.42±475.76	312±545.69
Titre ANCA	52.88 ^a 52.88	21.38 ^a 47.30	14.52±26.5	4.18 ^a 10.34	3.85 ^a 13.93	4.71 ^a 13.93	3.14 ^a 17.09
Index of activity/Prognosis							
BUS	15.92	N/D	2.82	2.56	1.45	1.13	N/D
FTS	0.88	0.58	0.72	0.45	0.40	0.36	N/D

Conclusions: IGIV seems to be an effective and secure therapy in the treatment of vasculitis ANCA.

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SAT0541 INCIDENCE OF LARGE VESSEL GCA IN NORTHERN ITALY DURING A 12-YEAR PERIOD (2005–2016)

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Background: There are no studies regarding the incidence of large vessel GCA (LV GCA).

Objectives: To investigate the incidence of LVGCA in the Reggio Emilia Area from 2005 through to 2016.

Methods: All patients with incident large vessel GCA diagnosed between 1 January 2005 and 31 December 2016 and living in the Reggio Emilia area, were identified by capture and re-capture checking of computerised discharge diagnosis codes (ICD10) and using outpatients databases from rheumatology, internal medicine, surgery, pathology, imaging departments of Reggio Emilia Hospital as well as by examining the Reggio Emilia district database for rare diseases. To be included in the study, patients must satisfy the following 2 criteria: Age at disease onset ≥50 years; evidence of large-vessel vasculitis by angiography, MRA, CTA, PET/CT and/or ultrasonography. We included in the study also patients associating biopsy proven GCA with evidence of LVV. We also evaluated the incidence of biopsy proven GCA without LV in same time period.

Results: There were 93 incident cases of LV GCA (66 women, 71%) during the 12 year study period; Mean ±SD age at diagnosis was 72±9 years. Incidence per 1 00 000 persons aged ≥50 years was 3.78 (95% confidence interval [95% CI 3.01, 4.55]). In particular incidence was 1.60 in LV GCA with biopsy proven GCA and 2.18 in LV GCA not biopsy proven GCA (pts biopsy negative and pts in which the biopsy had not been performed). Incidence was significantly higher in women (4.89 [95% CI 3.69, 6.09]) than in men (2.50 [95% CI 1.56, 3.45]) (p<0.0006). The highest incidence in women was observed in the 70–79 years age group (7.72 [95% CI 4.89, 11.58]) while in men the peak of incidence was in the 80–89 age group (4.23 [95% CI 2.50, 8.45]). A progressive increase in total incidence rates was observed during the 4 three years periods from 3.13 (2005–2007) to 4.85 (2014–2016). The incidence per 1 00 000 persons aged ≥50 years of GCA

biopsy proven without LV during the 12 year study period was 4.48 [95% CI 2.22, 9.45].

Conclusions: The incidence of LV GCA in the Reggio Emilia area 100.000/aged >50 was 3.78 and it was lower than that of patients with biopsy proven GCA without LV; the incidence of LV GCA was significantly higher in women and increased during the study period.

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Disclosure of Interest: None declared

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SAT0542 INTERLEUKIN-12 AND INTERLEUKIN-23 ARE KEY PATHOGENIC PLAYERS IN GIANT CELL ARTERITIS

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Background: Giant cell arteritis (GCA) is the most common form of systemic vasculitis; the pathogenesis is unclear. Current evidence suggests both the T_H1 and T_H17 pathways are important but the proximal initiators and effector cytokines are unknown. IL-12 and IL-23 secreted by dendritic cells are hypothesised as stimulators of these pathways. We have previously reported the efficacy of IL-12/23 blockade with ustekinumab in refractory GCA in a prospective clinical trial.

Objectives: To assess the role of IL-12 and IL-23 in GCA pathogenesis.

Methods: IL-12 and IL-23 were quantified by immunohistochemistry in temporal artery (TA) biopsies. TA explant, peripheral blood mononuclear cell (PBMC) and myofibroblast outgrowth culture models were established from patients with GCA and disease controls. PBMCs and TA explants were cultured for 24 hours in the presence or absence of IL-23 (10 ng/ml) or IL-12 (50 ng/ml). Gene expression was quantified by Real-time PCR and cytokine secretion by ELISA. Myofibroblast outgrowths were assessed following 28 days culture and quantified by counting the number of outgrowths/high-power field (hpf).

Results: Immunohistochemistry demonstrated IL-12p35 and IL-23p19 in inflammatory cells in TA biopsies (n=33). IL-12p35 and IL-23p19 were only detected in positive TA biopsies. IL-12p35 was increased in those with cranial ischaemic complications (p=0.026) and those with large vessel vasculitis (p=0.006). IL-23p19 was increased in those with two or more relapses (p=0.007). In cultured PBMCs, IL-12 stimulation increased IL-6 (n=17, p=0.009), IL-22 (n=16, p=0.003), and IFN-γ (n=14, p=0.0001) secretion and decreased IL-8 (n=15, p=0.0006) secretion, while IL-23 stimulation increased IL-6 (n=40, p=0.029), IL-22 (n=16, p=0.001), IL-17A (n=16, p=0.0003) and IL-17F (n=9, p=0.012) secretion. In the TA explant culture model, IL-23 stimulation increased gene expression of IL-8 (n=13, p=0.0001) and CCL-20 (n=9, 0.027) and protein expression of IL-6 (n=61, p=0.002) and IL-8 (n=60, p=0.004). IL-12 stimulation (n=14) had no effect; however, IFN-γ and IL-17A were not detectable in this model. IL-12 (n=20, p=0.0005) and IL-23 (n=33, p<0.0001) stimulation increased the quantity of myofibroblast outgrowths from TA biopsies. In all experiments there were no significant differences between biopsy positive GCA, biopsy negative GCA, and disease controls.

Conclusions: IL-12 and IL-23 play central and distinct roles in stimulating inflammatory and proliferative pathways in GCA. Our results were consistent in patients with biopsy positive and negative GCA, and in disease controls, suggesting that IL-12 and IL-23 play proximal roles in inducing these pathways.

Disclosure of Interest: None declared

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