ANCA status (MPO-ANCA+ versus PR3-ANCA+), did not change these results. Despite similar immunosuppressive therapy approaches and relapse rates, elderly patients had a higher rate of severe infections compared to younger patients (OR 2.1, 95% CIs: 1.1-4.4, $p=0.043$; Figure below).

Abstract THU0290 – Figure 1

Conclusion: Our findings indicate that elderly patients had higher susceptibility to morbidity related to vasculitis or its treatment than younger patients, particularly to worst renal function recovery and higher infection rate.

REFERENCES:

- [1] Harper L, et al. ANCA-associated renal vasculitis at the end of the twentieth century—a disease of older patients. *Rheumatology (Oxford, England)*. 2005;44(4):495-501.
- [2] Pagnoux C, et al. Treatment of systemic necrotizing vasculitides in patients aged sixty-five years or older: results of a multicenter, open-label, randomized controlled trial of corticosteroid and cyclophosphamide-based induction therapy. *Arthritis & rheumatology (Hoboken, NJ)*. 2015;67(4):1117-27.
- [3] Yates M, et al. EULAR/ERA-EDTA recommendations for the management of ANCA-associated vasculitis. *Annals of the rheumatic diseases*. 2016;75(9):1583-94.

Disclosure of Interests: Alvise Berti: None declared, Mara Felicetti: None declared, Sara Monti: None declared, Augusta Ortolan: None declared, Roberto Padoan: None declared, Giuliano Brunori: None declared, Roberto Bortolotti: None declared, Roberto Caporali Speakers bureau: AbbVie, Bristol-Myers Squibb, Celgene, Roche, Genzyme, Lilly, MSD, Pfizer, UCB, Carlomaurizio Montecucco Speakers bureau: AbbVie, Bristol-Myers Squibb, Celgene, Sanofi, Genzyme, Lilly, MSD, Pfizer, UCB, Franco Schiavon: None declared, Giuseppe Paolazzi: None declared

DOI: 10.1136/annrheumdis-2019-eular.6078

THU0291 THE CHARACTERISTICS OF PEDIATRIC BEHÇET'S DISEASE IN TURKEY VERSUS ISRAEL

Yonatan Butbul¹, Ezgi Deniz Batu², Hafize Emine Sonmez², Betül Sözeri³, Nuray Aktay Ayaz⁴, Limor Baba¹, Gil Amarilio⁵, Seval Simsek³, Liora Harel⁵, Şerife Gül Karadağ⁴, Yelda Bilginer², Seza Özen². ¹Ruth Rappaport Children's Hospital, Rambam Medical Center, Department of Pediatrics, Division of Rheumatology, Haifa, Israel; ²Hacettepe University Faculty of Medicine, Department of Pediatrics, Division of Rheumatology, Ankara, Turkey; ³Umraniye Training and Research Center, University of Health Sciences, Department of Pediatrics, Division of Rheumatology, Istanbul, Turkey; ⁴Kanuni Sultan Suleyman Research and Training Hospital, University of Health Sciences, Department of Pediatrics, Division of Rheumatology, Istanbul, Turkey; ⁵Schneider Children's Medical Center of Israel, Department of Pediatrics, Division of Rheumatology, Petach Tikva, Israel

Background: Behçet's disease (BD) is a variable vessel vasculitis affecting all sizes of vasculature in both the arterial and venous systems. Children with BD comprises 3-24% of all BD patients.

Objectives: To compare the demographics, presentation, clinical manifestations, disease activity, and treatment in pediatric Behçet's disease in Turkey versus Israel.

Methods: Three centers from Turkey and two centers from Israel participated in this study. The diagnosis of BD was before 16 years of age and based on expert opinion at each center. BD current activity form (BDCAF) was used for assessing disease activity.

Results: A total of 205 patients were included (165 from Turkey; 40 from Israel). HLA-B51 positivity (68.3% vs 46.2%, $p=0.028$), male gender (52.1% vs 30%, $p=0.012$), and skin involvement (especially necrotic

folliculitis) (55.2% vs 22.5%, $p<0.001$) were more frequent among patients from Turkey compared to the ones from Israel. The first disease-related symptom was oral aphthosis in both groups followed by genital aphthosis and arthralgia/arthritis in patients from Turkey and Israel, respectively. The median BDCAF values were worse at the first visit for patients from Turkey than those from Israel (4 vs 2, $p<0.001$). The most commonly prescribed drug was colchicine in the whole group (96.6%). Significantly more patients were treated with corticosteroids (50% vs 28.5%, $p=0.006$), methotrexate (17.5% vs 3%, $p=0.002$), infliximab (10% vs 1.8%, $p=0.026$), and nonsteroidal anti-inflammatory drugs (12.5% vs 1.8%, $p=0.007$) in Israel than Turkey. For the classification of BD in the whole group, ICB criteria had the highest sensitivity (73.2%) followed by PED-BD (pediatric BD) (47.8%) and ISG (The International Study Group) (42%) criteria sets.

Abstract THU291 – Table 1. The characteristics of children with Behçet's disease

Characteristics, n (%)	All patients (n=205)	Patients from Turkey (n=165)	Patients from Israel (n=40)	P value
Male	98 (47.8)	86 (52.1)	12 (30)	0.012
Age at symptom onset, months, median (min-max)	133 (12-191)	132 (12-191)	134 (12-190)	1
Age at diagnosis, months, median (min-max)	156 (48-191)	156 (48-191)	153 (55-191)	0.96
Oral aphthosis	204 (99.5)	165 (100)	39 (97.5)	0.195
Genital aphthosis	134 (65.4)	107 (64.8)	27 (67.5)	0.75
Skin involvement	100 (48.8)	91 (55.2)	9 (22.5)	<0.001
Eye involvement	29 (14.1)	22 (13.3)	7 (17.5)	0.49
Neurologic involvement	30 (14.6)	26 (15.8)	4 (10)	0.35
Vascular involvement	22 (10.7)	19 (11.5)	3 (7.5)	0.77
Pathergy positivity	45/167 (26.9)	42/154 (27.3)	3/13 (23.1)	1
HLA-B51 positivity	122/187 (65.2)	110/161 (68.3)	12/26 (46.2)	0.028
BDCAF at first visit	3 (0-15)	4 (1-15)	2 (0-5)	<0.001
BDCAF at last visit	1 (0-13)	1 (0-13)	1 (0-4)	0.15

Conclusion: This is the largest cohort of pediatric BD reported to date. The disease manifestations and disease activity significantly differ among pediatric BD patients from Turkey and Israel which emphasizes the effect of the ethnicity on disease phenotype.

REFERENCE:

- [1] Kone-Paut I, et al. Consensus classification criteria for paediatric Behçet's disease from a prospective observational cohort: PEDBD. *Ann Rheum Dis* 2016;75:958-64.

Disclosure of Interests: Yonatan Butbul: None declared, Ezgi Deniz Batu: None declared, Hafize Emine Sonmez: None declared, Betül Sözeri: None declared, Nuray Aktay Ayaz: None declared, Limor Baba: None declared, Gil Amarilio: None declared, Seval Simsek: None declared, Liora Harel: None declared, Şerife Gül Karadağ: None declared, Yelda Bilginer: None declared, Seza Özen Consultant for: Seza Ozen is receiving consultancy fees from Novartis, Speakers bureau: Roche

DOI: 10.1136/annrheumdis-2019-eular.2671

THU0292 EXTRACRANIAL VESSEL INVOLVEMENT IN PATIENTS WITH GIANT CELL ARTERITIS

Monica Calderón-Goercke¹, J. Loricera¹, D. Prieto-Peña¹, J. Narváez², Santos Castañeda³, Elena Aurrecoechea⁴, Ignacio Villa-Blanco⁴, Catalina Gomez-Arango⁵, Eva Perez-Pampin⁶, Vicente Aldasoro⁵, Noelia Alvarez-Rivas⁷, Nagore Fernández-Llanio⁸, María Álvarez del Buero⁹, Luisa Marena Rojas¹⁰, Francisca Sivera¹¹, Eva Galindez¹², Roser Solans-Laqué², Susana Romero-Yuste¹³, Norberto Ortego¹⁴, Marcelino Revenga³, Rafael Melero¹⁵, Eva Salgado-Pérez¹⁶, Sabela Fernández¹⁷, Vanesa Calvo-Río¹, Natalia Palmou-Fontana¹, Jose Ignacio Banzo¹, Isabel Martínez-Rodríguez¹, Carmen González-Vela¹, Raquel Dos-Santos⁵, J. Luis Hernández¹, Miguel A González-Gay¹, Ricardo Blanco¹. ¹Santander, Santander, Spain; ²Barcelona, Barcelona, Spain; ³Madrid, Madrid, Spain; ⁴Torrelavega, Torrelavega, Spain; ⁵Mondragón, Mondragón, Spain; ⁶Santiago, Santiago de Compostela, Spain; ⁷Lugo, Lugo, Spain; ⁸Lérida, Lérida, Spain; ⁹Palencia, Palencia, Spain; ¹⁰Alcázar San Juan, Alcázar de San Juan, Spain; ¹¹Alicante, Alicante, Spain; ¹²Bilbao, Bilbao, Spain; ¹³Pontevedra, Pontevedra, Spain; ¹⁴Granada, Granada, Spain; ¹⁵Vigo, Vigo, Spain; ¹⁶Ourense, Ourense, Spain; ¹⁷Avilés, Avilés, Spain

Background: Giant cell arteritis (GCA) is a large vessel vasculitis with a predisposition for the cranial branches of the external carotid artery. However, aorta and/or its main branches may also be involved.

Objectives: In a series of patients with GCA who presented extracranial vessel involvement, our aim was to assess **a)** the vascular territories most frequently affected and **b)** correlation of a major extension of extracranial vascular involvement with a more severe clinical and analytical features.

Methods: Multicenter study of 68 patients with GCA who presented a compromise of extracranial vessels confirmed by PET/CT. Visual analysis of vascular uptake was performed on supra-aortic trunks (SAT), aortic arch (AA), thoracic aorta (TA), abdominal aorta (AbA), iliac arteries (IA), lower limb arteries (LLA), and upper limb arteries (ULA).

Results: We evaluated 68 patients with GCA (51w/17m) with a mean age of 68.0±8.3 years. The vascular territories affected were: TA (n=58, 85.29%), SAT (n=38, 55.88%), AbA (n=28, 41.18%), AA (n=18, 26.47%), LLA (n=17, 25%), IA (n=13, 19.12%) and ULA (n=6, 8.82%). We considered 3 groups according to the number of vascular territories affected: a) 1 or 2 territories, b) 3 or 4 territories and c) 5 or more territories and made a comparative study between this groups. In patients with ≥5 vascular territories affected, we observed a higher baseline ESR, and the most frequent systemic manifestations were polymyalgia rheumatica and constitutional symptoms with statistical significance (**TABLE**). Distribution of categorical variables was compared by the Pearson Chi-squared test. Quantitative variables were analyzed using the ANOVA test.

Conclusion: In patients with GCA the involvement of TA is very frequent, followed by the SAT and the AbA. Regarding the laboratory findings, patients with higher levels of ESR presented a major extension of extracranial vascular involvement, as well as presenting PMR and/or constitutional symptoms was also related to more affection of extracranial territories.

Abstract THU0292 – Table 2.

ACUTE PHASE REACTANTS	NUMBER OF VASCULAR TERRITORIES AFFECTED			p
	1-2	3-4	≥5	
CRP, mg/dl, mean (SD)	3.1 ± 1.0	5.4 ± 1.8	7.3 ± 3.2	0.315
ESR, mm/h, mean (SD)	36.8 ± 5.3	44.4 ± 6.2	74.9 ± 13.0	0.031
Hemoglobin, g/dl, mean (SD)	12.4 ± 0.2	12.0 ± 0.4	11.6 ± 0.5	0.438
SYSTEMIC MANIFESTATIONS				
Constitutional symptoms, n (%)	6 (17.1)	6 (22.2)	5 (83.3)	0.002
PMR, n (%)	28 (52.9)	23 (85.2)	4 (66.7)	0.03
ISCHEMIC MANIFESTATIONS				
Headache, n (%)	11 (31.4)	10 (37)	2 (33.3)	0.888
Joint claudication, n (%)	1 (2.9)	3 (11.1)	1 (16.7)	0.307
Visual involvement, n (%)	2 (5.7)	2 (7.4)	-	0.783

REFERENCES:

- [1] Loricera J, Blanco R, Hernández JL, et al. Use of positron emission tomography (PET) for the diagnosis of large-vessel vasculitis. *Rev Esp Med Nucl Imagen Mol*. 2015; 34: 372-377.
- [2] Loricera J, Blanco R, Hernández JL, et al. Non-infectious aortitis: a report of 32 cases from a single tertiary centre in a 4-year period and literature review. *Clin Exp Rheumatol*. 2015; 33: S19-31.

Disclosure of Interests: Monica Calderón-Goercke: None declared, J. Loricera: None declared, D. Prieto-Peña: None declared, J. Narváez Consultant for: Bristol-Myers Squibb, Santos Castañeda Consultant for: Amgen, BMS, Pfizer, Lilly, MSD, Roche, Sanofi, UCB, Elena Aurrecoechea: None declared, Ignacio Villa-Blanco: None declared, Catalina Gomez-Arango: None declared, Eva Perez-Pampín: None declared, Vicente Aldasoro: None declared, Noelia Alvarez-Rivas: None declared, Nagore Fernández-Llanio: None declared, María Álvarez del Buero: None declared, Luisa Marena Rojas: None declared, Francisca Sivera: None declared, Eva Galindez: None declared, Roser Solans-Laqué: None declared, Susana Romero-Yuste: None declared, Norberto Ortego: None declared, Marcelino Revenga: None declared, Rafael Melero: None declared, Eva Salgado-Pérez: None declared, Sabela Fernández: None declared, Vanesa Calvo-Río: None declared, Natalia Palmou-Fontana: None declared, Jose Ignacio Banzo: None declared, Isabel Martínez-Rodríguez: None declared, Carmen González-Vela: None declared, Raquel Dos-Santos: None declared, J. Luis Hernández: None declared, Miguel A González-Gay Grant/research support from: Prof. MA Gonzalez-Gay received grants/research supports from Abbvie, MSD, Jansen and Roche., Speakers bureau: Consultation fees/participation in company sponsored speaker's bureau from Pfizer, Lilly, Sobi, Celgene, Novartis, Roche and Sanofi., Ricardo Blanco Grant/research support from: Abbvie, MSD, and Roche, Consultant for: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen, Speakers bureau: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen

DOI: 10.1136/annrheumdis-2019-eular.2184

THU0293

DRUG RETENTION AND DISCONTINUATION REASONS BETWEEN SEVEN BIOLOGICS IN PATIENTS WITH TAKAYASU'S ARTERITIS: A SINGLE-CENTER EXPERIENCE

Corrado Campochiaro, Alessandro Tomelleri, Silvia Sartorelli, Giulio Cavalli, Giacomo De Luca, Elena Baldissera, Lorenzo Dagna. *San Raffaele Scientific Institute, Unit of Immunology, Rheumatology, Allergy and Rare Diseases (UnIRAR), Milan, Italy*

Background: Biologic disease-modifying anti-rheumatic drugs (bDMARDs) have been showed to be effective in refractory Takayasu arteritis (TA) [1]. Data about bDMARDs drug retention rate (DRR) in TA are scarce.

Objectives: To investigate DRR and reasons for discontinuation of 7 bDMARDs in a real-world setting

Methods: TA patients fulfilling the 1990 ACR criteria and treated with ≥1 bDMARD were selected. Data about disease duration, number of bDMARDs, reasons for bDMARDs discontinuation (classified as inefficacy, toxic reactions, non-toxic reasons), concomitant conventional synthetic (cs) DMARDs were collected. Survival curves were examined by the Kaplan-Meier method and compared using a stratified log-rank test. DRR was evaluated at 24 months. Hazard ratio (HR) for concomitant csDMARDs therapy and for previous bDMARDs was performed. A comparative sub-analysis between anti-TNFα drugs and tocilizumab (TCZ) was performed

Results: We identified 50 patients and 86 bDMARD-courses. At baseline, mean age was 39.1±12.1 years, mean disease duration was 119.5±110.1 months. No significant differences were observed in age and disease duration between the 7 groups. 31 patients were treated with 1 bDMARD, 11 with 2, 4 with 3, 2 with 5, 1 with 6, 1 with 7. Infliximab was the most frequent first-line bDMARD (78.6%). At bDMARDs initiation all patients were on prednisone (mean dose 13.5±10.3 mg), 85.2% on csDMARD (53 methotrexate, 13 azathioprine, 6 sirolimus, 3 mycophenolate-mofetil, 1 sulfasalazine). 43% of treatment courses were stopped by 24 months. Table 1 summarizes DRR and reasons for treatment discontinuation for each bDMARD. Golimumab had the highest DRR (71.4%), followed by infliximab (69%), adalimumab (56.3%), abatacept (50%), tocilizumab (41.1%), anakinra (0%) and rituximab (0%), p=0.016. Only concomitant csDMARDs therapy (HR=2.87, 95%CI=1.19-6.92, p=0.019) showed positive effects on DRR. Anti-TNFα drugs had significantly higher DRR compared to TCZ (67.2% vs 41.1%, p=0.028). Even in these subgroups csDMARDs (HR=3.79, 95% CI=1.49-9.6, p=0.005) showed positive effects on DRR

bDMARDs (n° of treatment courses)	N° of previous bDMARDs	csDMARD associated	24- month DRR	Reasons for bDMARDs discontinuation			24- month DRR
				Inefficacy	Toxicity	Non- toxic events	
IFX (42)	0 (0-2)	38 (90%)	69%	5 (12%)	6 (14%)	2 (5%)	67%
ADA (12)	0 (0-5)	11 (92%)	56%	4 (33%)	1 (8%)	0	
GOL (7)	2 (0-6)	5 (71%)	71.4%	1 (14%)	1 (14%)	0	
TCZ (17)	1 (0-4)	15 (88%)	41%	7 (41%)	2 (12%)	1 (6%)	41%
ANK (4)	2.5 (1-3)	2 (50%)	0%	2 (50%)	1 (25%)	1 (25%)	
ABT (2)	4.5 (4-5)	1 (50%)	50%	1 (50%)	0	0	
RTX (2)	2.5 (2-3)	1 (50%)	0%	2 (100%)	0	0	
Overall (86)	0.5 (0-6)	75 (86%)	60%	22 (26%)	11 (13%)	4 (5%)	

Conclusion: In our TA cohort, anti-TNFα agents had the highest DRR and, notably, they had a higher DRR in a head-to-head comparison with TCZ. Concomitant use of csDMARDs had a significant positive effect on bDMARDs DRR

REFERENCES:

- [1] Management of large-vessel vasculitis. Salvarani C, Hatemi G. *Curr Opin Rheumatol*. 2019 Jan;31

Disclosure of Interests: Corrado Campochiaro Consultant for: Dr Corrado Campochiaro received consultation honoraria from Pfizer., Alessandro Tomelleri: None declared, Silvia Sartorelli: None declared, Giulio Cavalli Consultant for: Dr Giulio Cavalli received consultation honoraria from Pfizer, Novartis and SOBI., Giacomo De Luca Speakers bureau: Pfizer, Sobi, Elena Baldissera Consultant for: Consultation honoraria from Novartis and Rottapharm, Speakers bureau: Pfizer, Sobi, Novartis, Lorenzo Dagna Consultant for: Prof Lorenzo Dagna received consultation honoraria from Abbvie, Amgen, Biogen, Bristol-Myers Squibb, Celltrion, Novartis, Pfizer, Sanofi-Genzyme, and SOBI.

DOI: 10.1136/annrheumdis-2019-eular.3537