Abstract THU0319 –Table 1. Prevalence and characteristics of inflammatory/autoimmune extravascular comorbidities in a cohort of 129 TAK patients, temporal correlation with TAK diagnosis, and need for biological therapy.

	DISEASE	N° OF CASES	ANTECEDENT TO TAK DIAGNOSIS	SUBSEQUENT TO TAK DIAGNOSIS	SYNCHRONOUS WITH TAK DIAGNOSIS	NEED FOR BIOLOGICAL THERAPY
AUTO-INFLAMMATORY	**Pericarditis**	4	3	0	1	3
	Relapsing Polychondritis	4	4	0	0	3
	Sarcoidosis	3	0	1	2	1
	TOT.	**11**	**7**	**1**	**3**	**7 (63.7%)**
GUT	**Ulcerative Rectocolitis**	3	2	1	0	2
	Crohn Disease	1	1	0	0	1
	Undifferentiated Colitis	1	1	0	0	1
	Celiac Disease	1	0	1	0	0
	TOT.	**6**	**4**	**2**	**0**	**4 (66.7%)**
ARTICULAR	**Psoriatic Arthritis**	4	0	4	0	3
	Enteropathic Arthritis	2	0	2	0	2
	Seronegative Rheumatoid Arthritis	1	1	0	0	1
	TOT.	**7**	**1**	**6**	**0**	**6 (85.7%)**
EYE	**Scleritis**	6	3	3	0	4
	Anterior Uveitis	3	2	1	0	1
	Posterior Uveitis	1	0	1	0	0
	Optic Neuritis	2	2	0	0	1
	Serpiginous Choroiditis	1	1	0	0	0
	TOT.	**13**	**8**	**5**	**0**	**6 (46.2%)**
SKIN/MUCOSA	**Erythema Nodosum**	18	8	8	2	10
	Chronic Urticaria	4	3	1	0	3
	Chronic Recurrent Oral Aphthous Ulcers	2	1	1	0	2
	Skin Psoriasis	1	1	0	0	0
	TOT.	**25**	**13**	**10**	**2**	**15 (60%)**
MISCELLANEOUS	**Retroperitoneal fibrosis**	1	0	0	1	0
	Autoimmune Hepatitis	1	0	1	0	0
		64 (46 patients)	**33 (51.6%)**	**25 (39%)**	**6 (9.4%)**	**38 (25 patients)**

history for inflammatory/autoimmune diseases (8 psoriasis, 5 rheumatoid arthritis, 3 inflammatory bowel disease, 1 sarcoidosis).

Conclusion: In TAK patients, extravascular involvement is a common finding and usually precedes the vascular involvement (the only significant exception is represented by inflammatory arthritis). Inflammatory and autoimmune extravascular comorbidities seem to be associated with a significant higher burden of vascular inflammation, as inferred by the higher rate of biological therapies use in these TAK patients.

REFERENCES:

[1] Numano, et al. Takayasu arteritis. Lancet (2000).
[2] Kwon, et al. Extravascular manifestations of Takayasu arteritis: focusing on the features shared with spondyloarthritis. Arthritis Res Ther (2018)

Disclosure of Interests: Alessandro Tomelleri: None declared, Corrado Campochiaro Consultant for: Dr Corrado Campochiaro received consultation honoraria from Pfizer., Silvia Sartorelli: None declared, Veronica Bandini: None declared, Elena Baldissera Consultant for: Consultation honoraria from Novartis and Rottapharm, Speakers bureau: Pfeizer, 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.953

THU0320 RISK FACTORS OF INTRAVENOUS IMMUNOGLOBULIN RESISTANCE AND CORONARY ARTERIAL LESIONS IN TURKISH CHILDREN WITH KAWASAKI DISEASE

Serkan Turkucar[1,1], Kaan Yildiz[2], Ceyhun Acari[1], Hatice Adiguzel Dundar[1], Mustafa Kir[2], Erbil Unsal[1]. *[1]Dokuz Eylul University, Department of Pediatric Rheumatology, Izmir, Turkey; [2]Dokuz Eylul University, Department of Pediatric Cardiology, Izmir, Turkey*

Background: Kawasaki Disease (KD) is an acute, systemic, febrile vasculitis that occurs during infancy and is the most common cause of childhood coronary artery disease. The incidence of coronary artery lesions (CALs) has declined with the routine use of intravenous immunoglobulin (IVIG) treatment, but there is still considerable risk for resistance to IVIG treatment and development of CALs (1).

Objectives: Previously defined risk scoring systems have limited predictive capacity for IVIG resistance of KD in Turkish children. The present study was aimed to determine the risk factors in Turkish children with IVIG resistant KD and coronary artery involvement.

Methods: Clinical, laboratory and echocardiographic data were retrospectively analyzed in 94 Kawasaki patients. IVIG resistant and responsive groups were compared.

Results: Of the 94 patients included in the study, 55 (58.5%) were male and 39 (41.5%) were female and the ratio was 1.41. The median (25-75 percentage) age at the time of diagnosis was 35 (19-52) months. CALs were observed at echocardiographic evaluation in 31 patients (33%) and 17 patients (18.1%) were IVIG resistant. IVIG resistant group had a higher rate of CALs compared to the IVIG responsive group ($p<0.05$). When two groups were compared prior to IVIG, neutrophil/lymphocyte ratio (NLR) and C-reactive protein (CRP) parameters were statistically higher in IVIG resistant group. NLR values still remained as statistically higher in the IVIG-resistant group after initial IVIG.

Regarding IVIG resistance, duration of fever $\geq$ 9.5 days, CRP $\geq$ 88 mg/L and NLR $\geq$1.69 were the best cutoff values. The criteria for at least two of these three predictors were considered to be statistically significant risk factors for detecting IVIG resistance in KD before treatment (76.47% sensitivity, 71.05% specificity and 95% confidence intervals were 50.1-93.19% and 59.51-80.89%, respectively).

Regarding risk for CALs, duration of fever $\geq$ 9.5 days before IVIG, (OR: 3.4) and Plt count after IVIG $\geq$ 670x10^3/uL, (OR: 5.5), were the best cutoff values.

Abstract THU0320 –Table 1. ROC analyses and Odds ratios for best cut-off values of variables for predicting IVIG resistance before IVIG treatment.

Variable	*Value*	*Sensitivity*	*Specificity*	*Discriminative ability*	*Odds ratio*
Duration of fever before IVIG	9.5 days	58.8%	72.7%	0.667±0.080 (95% CI: 0.510-0.824, p=0.032)	3.809 (95% CI: 1.28-11.31, p=0.016)
CRP before IVIG	88.0 mg/L	70.0%	64.7%	0.669±0.076 (95% CI: 0.520–0.817, p=0.032)	4.40 (95% CI: 1.38-13.97 p=0.012)
NLR before IVIG	1.69	93.3%	43.4%	0.679±0.070 (95% CI: 0.541–0.817, p=0.031)	4.40 (95% CI: 1.38-13.97 p=0.012)

p<0.05

Ann Rheum Dis: first published as 10.1136/annrheumdis-2019-eular.1341 on 27 June 2019. Downloaded from http://ard.bmj.com/ on September 2, 2021 by 193.140.151.30. Protected by copyright.

Conclusion: This study defined three criteria for IVIG resistance in KD prior to treatment: Duration of fever before IVIG ≥ 9.5 days, CRP ≥ 88 mg/L and NLR value ≥ 1.69. Presence of two of these three criteria were found as a significant risk factor for IVIG resistance. Following initial therapy with IVIG, if NLR value is ≥ 1.25, it also predicted ongoing inflammation and IVIG resistance, possibly a need for steroid therapy instead of second IVIG.
Based on the clinical and laboratory features, we established a new risk-scoring system for predicting IVIG resistance in Turkish children with KD. This may be useful for choosing optimal treatment for KD before coronary artery involvement.

REFERENCE:

[1] Newburger JW, Takahashi M, Burns JC, Beiser AS, Chung KJ, Duffy CE, Glode MP, Mason WH, Reddy V, Sanders SP, et al. The treatment of Kawasaki syndrome with intravenous gamma globulin. N Engl J Med. 1986 Aug 7 ve 315(6):341-7.

Disclosure of Interests: Serkan Turkucar: None declared, Kaan Yildiz: None declared, Ceyhun Acari: None declared, Hatice Adiguzel Dundar: None declared, Mustafa Kir: None declared, Erbil Unsal Grant/research support from: Novartis, AbbVie, Roche, Koçak Pharma, Speakers bureau: Novartis, AbbVie, Roche, Koçak Pharma
DOI: 10.1136/annrheumdis-2019-eular.1341

THU0321 CLINICAL OUTCOMES OF PATIENTS WITH GIANT CELL ARTERITIS AND POLYMYALGIA RHEUMATICA SYMPTOMS TREATED WITH TOCILIZUMAB IN ROUTINE CLINICAL PRACTICE

Sebastian Unizony[1], Robert Spiera[2], Jinglan Pei[3], Paris Sidiropoulos[3], Jennie H. Best[3], John H. Stone[1]. [1]*Massachusetts General Hospital, Harvard Medical School, Boston, United States of America*; [2]*Hospital for Special Surgery, Department of Medicine, New York, United States of America*; [3]*Genentech, Inc, South San Francisco, United States of America*

Background: Approximately 50% of patients with giant cell arteritis (GCA) also have polymyalgia rheumatica (PMR) symptoms.[1]
Objectives: To determine whether the presence of PMR symptoms upon GCA diagnosis impacts GCA clinical outcomes and to evaluate the effectiveness of tocilizumab (TCZ) for controlling PMR symptoms in patients with GCA.
Methods: Retrospective analysis of patients with GCA treated with TCZ at a single center (2010-2018). Disease flares, defined as re-appearance of clinical manifestations (eg, cranial or PMR signs or symptoms), were assessed among patients with or without PMR at the time of GCA diagnosis. PMR symptoms during disease flares were assessed before and after TCZ initiation.
Results: A total of 60 patients with GCA (71.7% female; mean age, 69.3 years) were followed for a mean (SD) of 2.3 (2.17) years. At diagnosis, 32 had PMR symptoms (2 patients with PMR symptoms only). **Table 1** shows baseline patient characteristics. Among patients with PMR at diagnosis, 75% (n = 24) had a total of 60 flares before TCZ initiation and 34.3% (n = 11) had a total of 23 flares after TCZ initiation (**Table 2**). Among 28 patients without PMR at diagnosis, 67.9% (n = 19) had a total of 42 flares before TCZ initiation and 25% (n = 7) had a total of 14 flares after TCZ initiation (**Table 2**). TCZ was associated with a significant reduction in the annual flare rate both in patients with PMR symptoms (P = 0.003) and without PMR symptoms (P = 0.03) at diagnosis (**Table 2**). Median time to flare was significantly longer after initiation of TCZ in patients with PMR symptoms (hazard ratio [HR] [95% CI]: 0.18 [0.06 to 0.53]; P = 0.002) and without PMR symptoms (HR [95% CI]: 0.21 [0.05 to 0.87]; P = 0.032) at diagnosis. Before TCZ initiation, PMR symptoms were observed in 55.9% of flares (57/102) occurring in 43 patients (71.1%). After TCZ initiation, PMR symptoms were observed in 56.8% of flares (21/37) occurring in 18 patients (30%).
Conclusion: TCZ improved clinical outcomes in patients with GCA regardless of the presence or absence of PMR symptoms at diagnosis. These real-world findings suggest that TCZ is also effective in patients with GCA who have symptoms of PMR.

REFERENCE:

[1] Buttgereit F, et al. JAMA. 2016;315(22):2442-2458.

Abstract THU0321 –Table 1.

Table 1. Patient characteristics at GCA diagnosis and treatment received

	With PMR Symptoms (n = 32)	Without PMR Symptoms (n = 28)
Age, mean (SD)	67.0 (9.27)	71.8 (9.05)
Female, n (%)	23 (71.9)	20 (71.4)
White, n (%)	27 (84.4)	26 (92.9)
Clinical manifestations at disease onset, n (%)		
Localized headache	28 (87.5)	19 (67.9)
Scalp tenderness	15 (46.9)	11 (39.3)
Jaw claudication	16 (50.0)	15 (53.6)
Amaurosis fugax	5 (15.6)	6 (21.4)
Blurry vision	9 (28.1)	9 (32.1)
Permanent vision loss	2 (6.3)	6 (21.4)
Diplopia	1 (3.1)	1 (3.6)
Weight loss	6 (18.8)	14 (50.0)
PMR symptom(s) only, n (%)	2 (6.3)	0
ESR (mm/hr), mean (SD)	67.9 (36.52)	78.3 (29.41)
CRP (mg/L), mean (SD)	63.1 (59.69)	92.4 (84.85)
Positive TA biopsy, n (%)*	14/27 (43.8)	11/24 (39.3)
Disease duration before TCZ initiation, median (IQR), years	0.7 (0.2-1.9)	0.6 (0.2-1.4)
Duration of TCZ treatment (years), median (IQR)	0.6 (0.3-1.3)	0.5 (0.2-1.5)

CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; PMR, polymyalgia rheumatica; TA, temporal artery; TCZ, tocilizumab.
* Of patients with TA biopsies performed.

Abstract THU0221 –Table 2. Clinical outcomes before and after TCZ initiation

	With PMR at Diagnosis (n = 32)		Without PMR at Diagnosis (n = 28)	
	Before TCZ Initiation	After TCZ Initiation	Before TCZ Initiation	After TCZ Initiation
Patients with ≥ 1 flare, n (%)	24 (75.0)	11 (34.3)	19 (67.9)	7 (25.0)
With PMR symptom(s)*	21 (65.6)	8 (25.0)	4 (14.3)	2 (7.1)
Without PMR symptom (s)*	3 (9.4)	3 (9.4)	15 (53.6)	5 (17.9)
No. of flares	60	23	42	14
With PMR symptom(s), n (%)*	50 (83.3)	18 (78.3)	7 (16.7)	3 (21.4)
Without PMR symptom (s), n (%)*	10 (16.7)	5 (21.7)	35 (83.3)	11 (78.6)
Annual flare rate (95% CI)†	1.34 (0.84 to 2.13)	0.54 (0.28 to 1.05)	1.59 (0.78 to 3.23)	0.71 (0.29 to 1.74)
Time to flare (years), median (IQR)	0.3 (0.2-0.7)	2.1 (0.6-2.6)	0.5 (0.3-0.8)	2.6 (0.9-2.6)

GC, glucocorticoids; GCA, giant cell arteritis; IQR, interquartile range; PMR, polymyalgia rheumatica; TCZ, tocilizumab.
* Symptoms are after GCA diagnosis.
† Rates are estimated from a Poisson regression model with treatment, age, smoking history and new and relapsing GCA as covariates and random patient effect.

Disclosure of Interests: Sebastian Unizony Grant/research support from: F. Hoffmann-La Roche, Genentech, Consultant for: Kiniksa, Sanofi, GSK, Robert Spiera Grant/research support from: Roche-Genentech, GlaxoSmithKline, Bristol-Myers Squibb, Boehringer Ingelheim, Cytori, Chemocentryx, Corbux, Consultant for: Roche-Genentech, GlaxoSmithKline, CSL Behring, Sanofi Aventis, Jinglan Pei Employee of: Genentech, Paris Sidiropoulos Employee of: Genentech, Jennie H. Best Shareholder of: Genentech, Employee of: Genentech, John H. Stone Grant/research support from: F. Hoffmann-La Roche, Genentech, Xencor, Consultant for: Chugain, F. Hoffmann-La Roche, Genentech, Xencor
DOI: 10.1136/annrheumdis-2019-eular.2676

THU0322 CLINICAL CHARACTERISTICS OF BIOPSY-PROVEN IGA VASCULITIS IN CHILDREN AND ADULTS: A RETROSPECTIVE COHORT STUDY

Michel Villatoro Villar[1], Cynthia S. Crowson[1,2], Ashima Makol[1], Kenneth J Warrington[1], Steven R. Ytterberg[1], Matthew Koster[1]. [1]*Mayo Clinic, Rheumatology, Rochester MN, United States of America*; [2]*Mayo Clinic, Health Sciences Research, Rochester MN, United States of America*

Background: Differences in both presentation and outcome based on age of diagnosis have been described in patients with IgA vasculitis (IgAV) but data are limited due to cohort size and follow-up duration.
Objectives: To describe the differences in clinical characteristics and outcome between adult- and child-onset biopsy-proven IgA vasculitis (IgAV) in North America.

Ann Rheum Dis: first published as 10.1136/annrheumdis-2019-eular.1341 on 27 June 2019. Downloaded from http://ard.bmj.com/ on September 2, 2021 by 193.140.151.30. Protected by copyright.