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.

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Disclosure of Interests: Serkan Turkucar: None declared, Kaan Yildiz: None declared, Ceyhan Acari: None declared, Hatice Adiguzel Dunder: 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

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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.

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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, Cytro, 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

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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.

Methods: Patients with IgAV from January 1, 1997, through December 31, 2016 were retrospectively identified. Clinical characteristics, laboratory parameters and outcomes were abstracted from direct medical chart review. Proteinuria was classified as non-nephrotic (≥ 0.2 g/d, ≤ 3.5 g/d) or nephrotic (>3.5 g/d). Microscopic hematuria was defined as ≥ 5 RBCs/hpf or $\geq 2+$ on dipstick. Disease activity at each follow-up visit was categorized as complete response (normalization of all baseline abnormalities due to IgAV), partial response, non-response (lack of improvement of any abnormalities) or relapse (development of clinical signs of IgAV after a symptom-free period of at least one month). Prevalence of disease activity and competing risks were estimated using multi-state models.

Results: A total of 243 IgAV patients were identified (97% Caucasian, 58% male). 174 patients were adults (>21 years) and 69 were <21 years. Compared to patients <21 years, adults at baseline had more frequent ulcerative skin lesions (11% vs. 1%; $p=0.02$) and nephrotic-range proteinuria (22% vs. 3%; $p=0.007$) but less commonly had abdominal pain (34% vs. 61%; $p<0.001$), ischemic gastrointestinal involvement (10% vs. 20%; $p=0.04$) and arthralgias (38% vs. 61%; $p<0.001$). Frequency of microscopic hematuria was similar between groups (47%). Oral corticosteroids were the most common initial treatment used (80%).

Dialysis was required in 13 patients (8 adults) and renal transplant was performed in 4 cases (1 adult). Of 137 patients with hematuria during the study, 72% had complete resolution by 1 year after onset, compared to 50% of 179 patients with proteinuria. The prevalence of disease activity state at each follow-up time point is shown in figure 1. During 389 person-years of follow-up, 29 deaths were observed. The main causes of death were cancer, cardiovascular disease, infection and vasculitis. Five year survival rates (95% CI) for patients aged <21 , 21-50, and 51+ years were 100%, 94% (87, 100) and 40% (26, 63), respectively ($p<0.001$). Standardized mortality ratio for patients aged 21-50 years at diagnosis was 5.62 (0.68, 20.3) and 7.60 (5.0, 11.1) for those 51 or older.

Conclusion: IgAV in adults is associated with more severe skin/kidney involvement and poorer renal outcome. Among adults with IgAV, patients aged 51 years or older at diagnosis have significantly higher mortality.

Figure 1. Prevalence of IgA vasculitis disease state at each study time point during follow-up. At 1 year after diagnosis, 42% (95% CI: 30-51%) of patients were in complete response, 32% (95% CI: 24-39%) in partial response, 10% (95% CI: 6-13%) in non-response, 5% (95% CI: 0-10%) in relapse and 12% (95% CI: 5-19%) were deceased.

Abstract THU0322 – Figure 1

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7798

THU0323

THE MANAGEMENT OF BIOPSY NEGATIVE GIANT CELL ARTERITIS BY AUSTRALIAN RHEUMATOLOGISTS

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Background: The diagnosis of Giant Cell Arteritis (GCA) by Rheumatologists is based on a typical history of headache, jaw claudication and visual disturbance, examination findings such as scalp tenderness and abnormal temporal arteries, and confirmed by temporal artery biopsy (TAB) (1). Temporal artery biopsy is the gold standard for GCA diagnosis (2, 3). However clinicians may diagnose biopsy-negative GCA if there is a high enough pre-test probability for this condition, even if the biopsy is negative. There are few guidelines to assist clinicians with management of biopsy-negative GCA.

Objectives: The primary aim of this research project was to determine if there were differences in the management of patients with biopsy-positive versus biopsy-negative GCA with respect to duration of corticosteroid (CS) use, relapse rates, and use of steroid-sparing agents over a 12-month follow-up period in an Australian cohort. We also sought to determine if there were differences in patient characteristics, examination findings and investigation results between the two groups.

Methods: All patients who underwent a temporal artery biopsy during 2013-2016 for suspected GCA at three geographic sites (Prince of Wales Hospital, Royal Prince Alfred Hospital and Mid-North Coast Arthritis Clinic, Coffs Harbour) had their files reviewed for clinical presentation, examination findings, investigation results, and management over the 12-months following presentation. Comparison was made between biopsy-positive and biopsy-negative patients.

Results: One hundred and forty-two patient files were reviewed. One hundred and one patients were excluded. Of those remaining, 23 were biopsy-positive and 18 were biopsy-negative GCA. Biopsy-negative patients were younger at presentation compared to biopsy-positive GCA patients (69.3 years vs 76.1 years, $p = 0.01$). There was no difference in the frequency of headache, jaw claudication, polymyalgia rheumatica symptoms or visual disturbance between the two groups of patients. There was also no difference in ESR, CRP, platelet count, total WCC, ALP or Hb level. However, there was a trend towards higher ALP levels in biopsy-positive patients ($p = 0.058$). The length of temporal artery biopsy was similar between biopsy-positive and biopsy-negative patients (19.4mm vs 19.7mm, $p = 0.902$). Fewer biopsy-negative patients remained on corticosteroids at 12 months following presentation (65% vs 100%) and of those that did remain on corticosteroids, their dosage tended to be lower than for those with positive biopsies (3.4mg/d vs 7.5mg/d, $p = 0.002$). Fewer biopsy-negative patients were commenced on methotrexate (5.6% vs 34% $p = 0.02$). There was no difference in the frequency of relapse between biopsy-positive vs biopsy-negative patients ($p = 0.40$). Biopsy-positive patients were more likely to present in December or January compared to biopsy-negative patients ($p = 0.07$). There was no difference in corticosteroid duration prior to biopsy ($p = 0.42$).

Conclusion: Biopsy-positive patients were treated with corticosteroids for longer and at higher doses than those with a negative biopsy. They were also more likely to present in summer and be treated with methotrexate compared to biopsy-negative patients. The seasonal difference in presentation may suggest a different trigger compared to biopsy-negative patients.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5140