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FRI0215 **RESPONSE TO TOCILIZUMAB IN LARGE VESSEL VASCULITIS ACCORDING TO THE EXTENT OF BASELINE 18F-FDG VASCULAR UPTAKE**

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Background: ¹⁸F-FDG PET/CT is useful to establish the presence and extent of large vessel vasculitis (LVV)(1-2). Early therapy is needed to prevent severe compli-
 cations. Tocilizumab (TCZ) has shown efficacy in LVV (3-5). However, it is unknown
 if the extent of FDG vascular uptake may influence on clinical response to TCZ

Objectives: To assess the correlation of the extent of baseline FDG vascular
 uptake in PET/CT scan with clinical response to TCZ in patients with LVV.

Methods: Single center study of patients with LVV treated with TCZ who were
 divided into 2 groups depending on the extent of vascular uptake in baseline
 PET/CT scan: **a)** 1-2 affected areas **b)** ≥3 affected areas. Vascular uptake was
 qualitatively assessed by two experienced nuclear medicine physicians in five
 areas (supraaortic trunks, thoracic aorta, abdominal aorta, iliac and femorotibial
 arteries). We assessed clinical improvement (no improvement/partial/complete),
 normalization of CRP (≤0.5mg/dL) and/or ESR (≤ 20 mm/1st h) and reduction of
 prednisone dose (mg/day).

Results: 30 patients (24 w/6 m); mean age 65.3±10.6 yrs. In baseline PET/CT,
 vascular uptake was observed in 1 or 2 areas (n=13) and in ≥3 areas (n=17).
 There was a trend to higher ESR/CRP and shorter evolution of clinical symp-
 toms before TCZ onset in patients with ≥3 affected areas (TABLE 1). Clinical/
 serological evolution and reduction of prednisone dose is shown in TABLE 2.
 No statistical differences were found. However, patients with ≥ 3 affected areas
 tended to experience a slower clinical response.

TABLE 1.

	1-2 vascular affected areas (n=13)	≥3 vascular affected areas (n=17)	P
Demographic data			
Age, mean ± SD	66.0 ± 10.8	64.8 ± 10.7	0.77
Sex (women), n (%)	11 (84.6)	13 (76.5)	0.67
Evolution time before TCZ (months), median [IQR]	26.0 [3.5-34.0]	5.0 [1.5-10.0]	0.02
Laboratory markers			
ESR (mm/1st h), mean ± SD	30.0 ± 27.3	34.8 ± 27.6	0.64
CRP (mg/dL), mean ± SD	1.3 ± 1.2	1.8 ± 1.7	0.28
Previous treatment			
Prednisone dose (mg/day), mean ± SD	9.4 ± 6.2	7.9 ± 6.9	0.53
TCZ therapy			
Intravenous, n (%)	10 (76.9)	11 (64.7)	0.47
Combined with MTX, n (%)	6 (46.2)	8 (47.1)	0.96

TABLE 2.

	1-2 vascular affected areas (n=13)	≥ 3 vascular affected areas (n=17)	P
Complete clinical improvement, n/N (%)			
6 m	11/13 (84.6)	12/17 (70.6)	0.43
12 m	12/13 (92.3)	13/17 (76.5)	0.36
18 m	10/11 (90.9)	11/12 (91.7)	0.99
24 m	8/8 (100)	9/10 (90.0)	0.99
Normalization of ESR and/or CRP, n/N (%)			
6 m	13/13 (100)	16/17 (94.1)	0.99
12 m	13/13 (100)	16/17 (94.1)	0.99
18 m	11/11 (100)	11/12 (91.7)	0.99
24 m	8/8 (100)	10/10 (100)	0.99
Dose of Prednisone (mg/day), median [IQR]			
6 m	5.0 [1.3-5.0]	5.0 [0.0-5.0]	0.98
12 m	2.5 [0.0-3.8]	0.0 [0.0-5.0]	0.97
18 m	0.0 [0.0-2.5]	0.0 [0.0-1.9]	0.72
24 m	0.0 [0.0-2.2]	0.0 [0.0-2.5]	0.77

Conclusion: TCZ therapy was effective in patients with LVV regardless the
 extent of FDG vascular uptake in baseline PET/CT scan. However, a trend to a
 slower clinical response was observed in patients with ≥3 affected areas.

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FRI0216 **STERIOD SPARING EFFECT, LOWER INCIDENCE OF DISEASE RELAPSE AND DIABETES IN GIANT CELL ARTERITIS TREATED WITH IMMUNOSUPPRESSORS AB INITIO OR VERY EARLY: A MULTICENTER RETROSPECTIVE CASE-CONTROL STUDY**

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Background: Glucocorticoids (GC) are associated with serious side effects in
 giant cell arteritis (GCA). Immunosuppressive therapies (IT) gave conflicting
 results in GCA, regarding GC sparing effect. Recently, tocilizumab by blocking
 IL-6, has been licensed as first biologic treatment for GCA, being clinically effec-
 tive and saving GC (1).

Objectives: To evaluate the usefulness of IT for GCA in: 1) minimizing the rate of
 GC-induced adverse events (AEs) and 2) reducing the risk of relapse.

Methods: A multicenter retrospective case-control study included 165 GCA
 was performed. The first group of patients (GCA-IT) included 114 patients
 who were treated with at least one IT given ab initio or within 3 months from
 the start of GC. The control group included 51 GCA who received only GC
 or an IT later than 3 months (GCA-steroid). The primary endpoints were the
 rate of GC-related side effects: infections, hospitalized infections, new
 onset systemic arterial hypertension, GC-induced diabetes and osteoporotic
 fractures.

Results: Methotrexate up to 20 mg/week (138 patients), followed by cyclophos-
 phamide (48 patients) and tocilizumab (27 patients) were the most frequently
 used IT. No difference was observed as concerns the follow-up time between the
 two groups [48.5 (IQR 26-72) vs 40 (IQR 24-69), p=0.3, rank-sum test]. The two
 groups were similar as concerns sex (p=0.13), while the first group (69±8 yrs)
 was slightly younger than the second one (72±7 yrs) (p=0.005). Comorbidity
 was similar between groups. Patients in the GCA-IT group showed a significant
 lower incidence of GC-induced diabetes (8/114, 7% vs 12/51, 23.5%; p=0.003,
 chi-square test), while no differences were documented for rate of infections
 (p=0.64), including hospitalized infections (p=0.44), new onset systemic arterial
 hypertension (p=0.68), or osteoporotic fractures (p=0.32). Forty-four patients in
 the GCA-IT group (38.6%), while 34 patients in the GCA-steroid group (66.7%)
 experienced at least one relapse (p=0.001, chi square test). There was no dif-
 ference in terms of time to first relapse between the two groups (p=0.53, log-
 rank test). GCA-IT group was exposed to lower dose of GC at first (p<0.0001,

rank-sum test) and third ($p < 0.0001$, rank-sum test) month, while no differences were recorded at the other time points. Clinical outcomes were similar between the two groups.

Conclusion: Very early introduction of IT in GCA provided a greater steroid sparing in the first 3 months of treatment, leading to a lower incidence of diabetes. Relapse rate was even lower. IT was usually well tolerated without an increase incidence of infections. A randomized prospective trial is required to support this strategy in the management of GCA.

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FRI0217 SENSITIVITY AND SPECIFICITY OF 2019 DCVAS DRAFT CLASSIFICATION CRITERIA FOR GIANT CELLS ARTERITIS AND TAKAYASU ARTERITIS IN A MONOCENTRIC COHORT OF PATIENTS WITH CLINICAL DIAGNOSIS OF LARGE VESSEL VASCULITIS

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Background: Recently, a new set of classification criteria for Giant Cells Arteritis (GCA) and Takayasu Arteritis (TA) has been developed by the DCVAS project and presented as draft criteria at the 19th International Vasculitis and ANCA Workshop held in Philadelphia in 2019.

Objectives: The purpose of the present study is to analyze the performance of the 2019 DCVAS Draft Classification Criteria in differentiating GCA and TA in a cohort of patients with Large Vessel Vasculitis (LVV), comparing their sensitivity and specificity to 1990 ACR Classification Criteria.

Methods: 2019 DCVAS Draft Criteria and 1990 ACR Criteria were retrospectively applied to a cohort of 130 consecutive patients with Large Vessel Vasculitis. In all patients the diagnosis of vasculitis was histologically and/or radiologically confirmed.

Results: One-hundred patients had a clinical diagnosis of GCA, 25 patients of TA and 5 patients of other form of LVV, different from GCA and TA (idiopathic isolated aortitis n:2, aortitis with retroperitoneal fibrosis n:2, isolated pulmonary arteritis n:1).

Among the 100 patients clinically diagnosed as GCA (F/M: 68/32, age: 74 (60-83)) only 82 fulfilled the 1990 ACR Criteria for GCA, while all of them fulfilled the 2019 DCVAS Draft Criteria for GCA.

Instead, among the 25 patients with a clinical diagnosis of TA (F/M: 21/4, age: 34 (16-48)), 22 (88%) could be classified as TA according to the 1990 ACR Criteria, 25 (100%) according to 2019 DCVAS Draft Criteria.

In the group of patients diagnosed with other form of LVV (F/M: 2/3, age: 56 (38-71)) 4 patients (80%) fulfilled the 2019 DCVAS Draft Criteria for GCA, while none of them fulfilled the 2019 DCVAS Draft Criteria for TA or the 1990 ACR Criteria for GCA or TA. One of these patients did not fulfilled any classification criteria.

On the contrary, one GCA patient could be classified both as GCA or TA according to the 2019 DCVAS Draft Criteria but didn't fulfilled the 1990 ACR Criteria for GCA or TA.

For GCA, 2019 DCVAS Draft Criteria shown a sensitivity of 100% and a specificity of 80%, compared to 82% and 100% of 1990 ACR Criteria. For TA, 2019 DCVAS Draft Criteria shown a sensitivity of 100% and a specificity of 99%, compared to 88% and 100% of 1990 ACR Criteria.

For GCA the agreement between the two different sets of criteria was 85.5% (Cohen's k coefficient: 0.64), for TA the agreement was 85.1% (k: 0.58).

Conclusion: The new draft classification criteria shown a lower specificity if compared to the older ones, but also a higher sensitivity: in particular 2019 DCVAS Draft Criteria can better identify GCA patients with extracranial involvement, historically excluded from the 1990 ACR criteria.

both GCA and TA 2019 DCVAS Draft Criteria, demonstrating that this classification well performs in differentiating GCA and TA.

Table:

	GCA (n:100)	TA (n:25)	Other LVV (n:5)
Morning stiffness in shoulders or neck	43 (43%)	3 (12%)	0 (0%)
Sudden visual loss	33 (33%)	2 (8%)	0 (0%)
Jaw or tongue claudication	35 (35%)	0 (0%)	0 (0%)
New headache/	77 (77%)	5 (20%)	1 (20%)
Scalp tenderness	30 (30%)	0 (0%)	0 (0%)
Temporal Artery Exam: pathological findings	34 (34%)	0 (0%)	0 (0%)
Elevated ESR or CRP	100 (100%)	16 (64%)	5 (100%)
Temporal Artery Biopsy: vasculitis	53 (53%)	0 (0%)	0 (0%)
Temporal Artery halo sign (US)/	11 (11%)	0 (0%)	0 (0%)
Bilateral axillary involvement	11 (11%)	4 (16%)	0 (0%)
FDG-PET activity throughout aorta	32 (32%)	13 (52%)	4 (80%)
Angina or ischemic cardiac pain	5 (5%)	4 (16%)	1 (20%)
Arm or leg claudication	5 (5%)	12 (48%)	0 (0%)
Arterial bruit	2 (2%)	19 (76%)	1 (20%)
Reduced pulse in upper extremity	1 (1%)	15 (60%)	0 (0%)
Carotid: reduced pulse or tenderness	0 (0%)	8 (32%)	0 (0%)
SBP difference in arms: >10/ >20 mmHg	0 (0%)	14 (56%)	0 (0%)
	1 (1%)	6 (24%)	0 (0%)
Pathological Angiography or AngioCT:	19 (19%)	22 (88%)	5 (100%)
number of affected arteries: 1/2/>3	7/3/9	0/1/19	3/0/2
Paired branch arteries involvement	7 (7%)	20 (80%)	2 (40%)
Abdominal aorta with renal or mesenteric involvement	0 (0%)	17 (68%)	0 (0%)

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FRI0218 PREVALENCE, BURDEN OF DISEASE AND HEALTHCARE UTILIZATION AMONG PATIENTS WITH EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA) IN JAPAN 2005-2017

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Background: EGPA is a rare vasculitis condition with very limited data available from real-world settings on burden and health care utilization (HCU), particularly in Japan.

Objectives: To estimate the prevalence (overall and age, gender stratified) and describe HCU and treatment patterns among Japanese EGPA patients.

Methods: This was a retrospective descriptive cohort study using a large administrative claims database covering up to more than 5 million corporate employees and their dependents (JMDC claim database) in Japan. Annual prevalence from 2005-2017 was estimated using two EGPA case definitions: a) patients with ≥ 1 ICD-10 code (2003 version) for EGPA (M30.1), b) patients with ≥ 2 ICD-10 codes for EGPA (M30.1) during the year in which prevalence was calculated. Among newly identified EGPA patients with no EGPA code in at least 12 months before, clinical burden, comorbidities, after hour visiting (AHV), all cause hospitalization, and treatment with drugs, including oral corticosteroid (OCS) use was described. OCS dose was expressed as prednisone equivalent.

Results: The total number of newly identified patients in 2006-2016 was 45 persons and the mean (SD) age was 42.3 years (SD 14.7 years). The prevalence (per 1,000,000 patients) of EGPA with case definition a) in Japan in 2017 was estimated to be 38.0. The stratified prevalence (per 1,000,000) by age was: 2.3 in the group aged <18 years, 34.0 in those aged 18-50 years, and 91.1 in those aged ≥ 50 years, respectively. The prevalence in females (50.0) was approximately 1.7-fold higher than that in male (28.7). The prevalence, including stratified results, with definition b) was similar to that with definition a). In the newly identified patients, 60% of patients had at least one hospitalization and 55.6% had AHV, in the year after the first observed EGPA code during the study period. Following index date, new patients were treated: 77.8% with OCS, 11.1% with Azathioprine, 8.9% with intravenous immunoglobulin, 6.7% with Cyclophosphamide, 4.4% with Methotrexate, and 2.2% with Rituximab (non mutually exclusive). The mean (SD) maximum recorded daily dose of OCS in the 12 months follow up period was 53.5 (39.9) mg in new patients. The average dose (SD) of OCS in first month and last month in new patients was 39.1 (29.0) and 9.8mg (4.8), respectively. Among those with at least a 14-day supply of OCS, 73.1% could be classified as adherent ($\geq 80\%$) based on their 1-year proportion of days covered. 6.7% of EGPA patients experienced a potentially worsening with an increase of ≥ 10 mg daily OCS dose prescription following a previous prescription of <10mg.