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Background: The 2019 ACR/EULAR classification criteria for IgG4-RD have recently been published¹. In the criteria, patients with an inclusion criteria score of >20 without exclusion criteria are classified as having IgG4-RD.

Objectives: To validate the 2019 ACR/EULAR classification criteria for IgG4-RD in a Japanese kidney disease cohort.

Methods: The study involved Japanese patients diagnosed as having kidney disease between April 2012 and May 2019, for whom sufficient clinical information and data on serum IgG4 values and/or immunohistological staining for IgG4 in renal biopsy samples were known. These patients were classified as having IgG4-RKD or non-IgG4-RKD (mimickers) based on the 2019 ACR/EULAR classification criteria for IgG4-RD, and the results were evaluated by expert opinion.

Results: Among 105 included patients, the expert panel diagnosed 55 as having true IgG4-RKD and 50 as mimickers. The final diagnoses among the mimickers were vasculitis (n=11), idiopathic tubulointerstitial nephritis (TIN) (n=5), drug-induced TIN (n=5), Sjögren's syndrome (n=4) and others. Among the 55 true IgG4-RKD patients, 4 had exclusion criteria, and 50 of the remaining 51 had an inclusion criteria score of ≥20 points (sensitivity 90.9%). On the other hand, 49 of the 50 mimickers were classified as having non-IgG4-RKD (specificity 98.0%) (Table 1).

Table 1. General characteristics and prevalence of individual items of true IgG4-RKD and non-IgG4-RKD (mimicker)

	IgG4-RKD (true IgG4-RKD) (n=55)	Non-IgG4-RKD (mimicker) (n=50)	P
Age at diagnosis, mean ±SD (years)	69.9 ± 9.4	56.7 ± 17.4	<0.001
Male (%)	76.4	44	0.001
Elevated serum IgG4	54/55 (98.2%)	18/50 (36.0%)	<0.001
Serum IgG4 (mg/dl), mean±SD	1028 ± 796	226 ± 261	<0.001
Dense IgG4+Plasma cells (>10/hpf) in the kidney biopsy	48/51 (94.1%)	13/40 (32.5%)	<0.001
Storiform fibrosis in the kidney biopsy	28/51 (54.9%)	3/50 (6%)	<0.001
Hypocomplementemia	39/55 (70.1%)	7/42 (16.7%)	<0.001
Renal pelvis thickening/ soft tissue	5/55 (9%)	1/50 (2%)	0.20
Bilateral renal cortex low-density areas	29/55 (52.7%)	7/50 (14.0%)	<0.001
Exclusion criteria present	4/55 (7.3%)	22/50 (44%)	<0.001
Total inclusion criteria points >20 without exclusion criteria	50/55 (90.9%)	1/50 (2%)	<0.001

Conclusion: The 2019 ACR/EULAR classification criteria for IgG4-RD showed good agreement with expert classification in this Japanese kidney disease cohort.

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FRI0504

TOCILIZUMAB IN GRAVES' ORBITOPATHY. MULTICENTER STUDY OF 48 PATIENTS.

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Background: Tocilizumab (TCZ) has shown promising results in the treatment of inflammatory ocular disease, especially in uveitis (1-3). Graves orbitopathy (GO) is the most common complication of Graves' disease (GD). Conventional immunosuppressive drugs are not always effective or well tolerated. TCZ may be useful in the treatment of GO.

Objectives: To assess the efficacy of TCZ in corticoid-resistant GO.

Methods: Multicenter open study of corticoid-resistant GO. We measured clinical activity using the clinical activity score (CAS). CAS evaluates 10 different ocular items, ranging from 0 (no symptoms) to 10. We defined remission as the presence of CAS ≤ 3.

Results: We studied 48 (95 eyes) patients (TABLE). Besides oral corticosteroids, they had received iv Methylprednisolone (n=43), methimazole (n=20), selenium (n=11) or radioactive iodine (n=5). According to the classification of severity (EUGOGO group), before of TCZ onset our patients had severe (n=19) or moderate (n=29) disease. TCZ was used in monotherapy (n=45) or combined with methotrexate (n=2) or azathioprine (n=1) at a dose of 8 mg/kg/iv/4 weeks (n=43) or 162 mg/sc/week (n=5). TCZ yielded rapid and maintained improvement and most patients achieved remission (FIGURE).

After a mean follow-up of 16.1±2.1 months, most patients experienced ocular improvement, with TCZ withdrawal in 29 cases due to remission (n=25) or inefficacy (n=4). Only 5 relevant adverse events were observed (neutropenia, external otitis, otitis media, costal osteitis and gingival hyperplasia). None of these events resulted in discontinuation of the treatment.

Conclusion: TCZ appears to be an effective and safe treatment in corticoid-resistant GO.

TABLE:

Number of patients/eyes affected, n/n	48/96
Age, mean (SD), years	50.96 (11.78)
Sex, men/women, n/n (%)	10/38 (20.8 / 79.2)
Regimen of TCZ therapy	
Monotherapy/combined treatment, n (%)	45/3 (93.8/6.2)
• AZA	1 (2.1)
• MTX	2 (4.2)
TCZ dosage, n (%)	
• 8 mg/kg/iv/4 weeks	43 (89.6)
• 162 mg/sc/week	5 (10.4)
Follow-up on TCZ therapy, mean (SD), months	16.05±2.06
• Remission, n (%)	72 (75.8)
• Discontinuation treatment, n (%)	29 (60.4)
• Remission	25 (86.2)
• Inefficacy	4 (13.8)
• Side effects	0

FIGURE.

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FRI0505 TOCILIZUMAB DISCONTINUATION AFTER REMISSION ACHIEVEMENT IN PATIENTS WITH ADULT STILL'S DISEASE

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Background: The efficacy of tocilizumab, an interleukin (IL)-6 receptor inhibitor, has been proved in patients with adult Still's disease on suppressing systemic inflammation and decreasing glucocorticoid dose. However, whether tocilizumab can be discontinued after remission achievement is unclear.

Objectives: To clarify the possibility of tocilizumab discontinuation in patients with adult Still's disease who achieved remission with tocilizumab.

Methods: Consecutive patients with adult Still's disease diagnosed according to the Yamaguchi's criteria in our hospital from April 2012 until September 2019 were retrospectively reviewed. Patients who were in good control with tocilizumab were included in the analysis, and their clinical courses were collected from their medical charts. Patients were divided according to the presence of recurrence after tocilizumab discontinuation and compared.

Results: Among 42 patients with adult Still's disease who had a history of intravenous tocilizumab of 8mg/kg use, 13 patients discontinued tocilizumab following a good disease control. During the mean observation period of 26.4 months, six patients (46%) remained in remission while seven patients (54%) developed recurrence after tocilizumab discontinuation. The sex and the mean observation period were not different between the patients with recurrence and those without (71% vs 50%, $p=0.43$; 27.3 months vs 25.4 months, $p=0.93$, respectively), but the age at tocilizumab discontinuation tended to be higher in the recurrence group than the non-recurrence group (64.0 years vs 46.5 years, $p=0.08$). The disease activity including swollen joint counts and laboratory data at tocilizumab discontinuation were comparable between the two groups (serum ferritin levels, 88ng/mL vs 122ng/mL, $p=0.67$). While the duration of tocilizumab use was not different between the two groups (29.4 months vs 39.5 months, $p=0.40$), the mean interval of tocilizumab infusion at tocilizumab discontinuation in the recurrence group was 3.6 weeks, shorter than the 6.7 weeks in the non-recurrence group ($p=0.03$). The median dose of prednisolone at tocilizumab discontinuation was 5.0mg/day in the recurrence group and 0.0mg/day in the non-recurrence group ($p=0.06$). In the recurrence group, the duration from the last tocilizumab administration to recurrence was 7.8 months, and the median dose of prednisolone at recurrence was 5.0mg/day.

Conclusion: Patients with adult Still's disease remaining in remission with a longer interval of tocilizumab administration and a lower dose of prednisolone was likely to succeed in withdrawal of tocilizumab.

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FRI0506 EFFICACY AND SAFETY OF CANAKINUMAB IN ADULT-ONSET STILL'S DISEASE: A SINGLE-CENTER REAL-LIFE EXPERIENCE

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Background: The pro-inflammatory cytokine interleukin (IL)-1 has a central role in the pathogenesis of adult-onset Still's disease (AOSD), a rare auto-inflammatory condition. Anakinra, has been for years the cornerstone of IL-1-blocking therapy in AOSD. More recently, the monoclonal antibody canakinumab, a new agent blocking IL-1, has become available

Objectives: To describe our real-life experience with CNK in a cohort of AOSD patients from a single Italian Center

Methods: AOSD patients diagnosed according to Yamaguchi's criteria followed-up at our Autoinflammatory Unit and treated with CNK for at least 3 months were included. Demographic features, disease characteristics, reasons for CNK introduction, concomitant therapies, variation in systemic steroids dose, adverse events, and response to treatment were retrospectively evaluated. Non-parametric tests were used for statistical comparison

Results: 13 patients (5 women; median age 49 years, range 21-74), treated with subcutaneous CNK 4mg/kg 4-weekly, were identified. Median disease duration before CNK introduction was 12 (6-240) months. After CNK introduction, 2 patients were followed-up for 18 months, 3 for 12 months, 6 for 6 months, 2 for 3 months. CNK was introduced as first-line biologic DMARD in 6 patients. The other 7 patients had been already treated with at least one other bDMARD, for a total of 15 treatment courses (7, anakinra, ANK; 4, tocilizumab; 4, TNF-inhibitors), with a median bDMARD therapy duration of 8 (4-178) months. Previous bDMARDs had been interrupted because of inefficacy (8 cases) or adverse events (AE, 7 cases); of the 7 ANK-treated patients, therapy interruption was due to inefficacy in 3 cases. At CNK introduction, 11 patients were on systemic steroid therapy, prednisone (PDN) equivalent dose 15 (5-80) mg, and 10 were concomitantly receiving a conventional DMARD (7, methotrexate; 2, colchicine; 1, cyclosporine-A). **Graphic 1** summarizes main clinical features at CNK introduction. After CNK start, a striking and rapid clinical response was observed, as demonstrated by a substantial decrease of modified Pouchot score and a normalization of acute phase reactants after only 3 months (see Table 1 for details). CNK showed also a significant steroid-sparing effect: median PDN dose was reduced to 7.5 (2.5-12.5)

Table 1. Disease activity and blood tests at canakinumab introduction and during follow-up

Daily prednisone dose mg	Baseline (n=13)	3 months (n=13)	6 months (n=11)	12 months (n=5)	18 months (n=2)
Pouchot score	15 (5-80)	7.5 (2.5-12.5)	5 (0-7.5)	5 (0-7.5)	2.5
VAS pain	3 (2-5)	1 (0-2)	0 (0-1)	0	0
Erythrocyte sedimentation rate mm/h	7 (2-10)	3 (1-8)	2 (1-4)	1 (1-2)	1
C-reactive protein mg/L	42 (8-120)	21 (2-69)	13 (2-55)	14 (2-41)	11
Ferritin ng/mL	20.8 (3-180)	3.1 (0.5-22.5)	1.6 (0.5-8.4)	1 (0.3-6.3)	0.5
Hemoglobin g/dL	379.5 (161-914)	282 (82-552)	215 (34-464)	177 (77-401)	199
	13.1 (9.4-15.7)	13.2 (10.7-15.3)	13.8 (11.5-15.5)	13.9 (11.3-14.3)	13.5

Figure 1. Graphic 1 Main clinical features at canakinumab introduction