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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

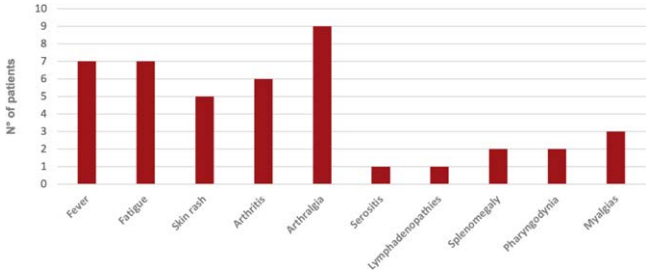

Figure 1. Graphic 1 Main clinical features at canakinumab introduction

mg at month 3 and 5 (0-7.5) mg at month 6; PDN was stopped in 3 patients (1 at month 3, 1 at month 6, 1 at month 12) due to optimal disease control. CNK was temporarily held-off in 3 patients (zoster reactivation, 1; prostatitis, 1; mild leukopenia, 1). We observed no case of primary inefficacy

Conclusion: Our real-life data confirm that CNK is highly effective and safe in AOSD treatment and has significant steroid-sparing effects. CNK showed its efficacy both as first-line therapy and after other bDMARDs failure, also in patients who have previously failed IL-1 inhibition through ANK

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FRI0507 COLCHICINE INTOLERANCE IN FMF PATIENTS AND PRIMARY OBSTACLES FOR OPTIMAL DOSING

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Background: Colchicine is the mainstay of treatment in FMF. However, in daily practice it is not easy to maintain effective colchicine doses in substantial number of patients, due to its side effects.

Objectives: It was aimed to investigate prevalence and risk factors for colchicine side effects that limit optimal drug dosing and permanent discontinuation.

Methods: All patients were recruited from "FMF in Central Anatolia" (FiCA) cohort, 915 adult subjects with minimum follow up time of 6 months and had compliance of treatment were included. Demographic and anthropometric data, FMF disease characteristics, disease severity, complications and treatment features were recorded on a web based registry. Prevalence of colchicine intolerance and characteristics of intolerant patients were analyzed.

Results: Effective colchicine doses cannot be maintained in 172 (18.7%) subjects. Main side effects that limit optimal dosing were as follows; diarrhea in 99 (10.8%), elevation in transaminases in 54 (5.9%), leukopenia in 10 (%1.1), renal impairment in 14 (1.3%), myopathy in 5 (0.5%) and allergic skin reaction in two. Colchicine had to be permanently ceased in 18 (2%) patients because of serious toxicity. Male gender and obesity were found to be associated with liver toxicity and having normal body weight was associated with diarrhea. Chronic inflammation and proteinuria were more common in colchicine intolerant patients and they had reported more frequent attacks compared to those tolerating optimal doses.

Table 1. Prevalence of all side effects of colchicine and reasons for drug discontinuation

Side effect	All side effects N=172*	Permanent cessation N=18*
Diarrhea	99	11
Liver toxicity	54	4
Leukopenia	10	1
Muscle toxicity	5	2
Skin reaction	2	-
Nausea	4	-
Infertility	2	-

* some patients had more than one clinically significant side effect

Table 2. Disease course in colchicine tolerant and intolerant patients

	Colchicine Tolerant N=743	Colchicine Intolerant N=172	p value
Chronic inflammation	115 (15.4%)	45 (26.1%)	**<0.001**
Number of attacks in the last year	4.05±6.08	7.60±9.6	**<0.001**
Proteinuria	44 (5.9 %)	20 (11.6%)	**0.025**
Amyloidosis	33 (% 4.4)	23 (13.3%)	**<0.001**
ADDI (median)	1 (1)	1 (1)	**<0.001**

ADDI: auto-inflammatory disease damage index, FMF: familial Mediterranean fever

Conclusion: Colchicine intolerance is an important problem in daily clinical practice, mainly due to diarrhea and liver toxicity. Suboptimal colchicine dosing associated with complications.

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FRI0508 MALIGNANCY AND IGG4-RELATED DISEASE: THE INCIDENCE, RELATED FACTORS AND PROGNOSIS FROM A PROSPECTIVE COHORT STUDY IN CHINA.

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Background: The association between IgG4-related disease (IgG4-RD) and malignancies is unclear. No epidemiological data for malignancies in Chinese IgG4-RD patients is available. It is also important to know the risk factors and prognosis for IgG4-RD patients harboring malignancies.

Objectives: To investigate the incidence, related factors and prognosis of IgG4-related disease (IgG4-RD) with malignancies in the Chinese cohort.

Table 1. Baseline characteristic of IgG4-RD patients with malignancy

Patient	Sex	Age	Age at diagnosis of IgG4-RD	Age at diagnosis of malignancy	Serum IgG4 (g/L)	Organs involvements of IgG4-RD (*Organ with biopsy)	Sites of malignancy
P1	F	59	58	54	1499	Parotid gland*, salivary gland	Breast cancer
P2	M	74	66	68	10402	Pancreas, bile duct, retroperitoneal fibrosis, kidney, prostate, lymph nodes	Rectal cancer
P3	M	46	42	40	2630	Lacrimal gland, parotid gland	Lipoblastoma
P4	M	70	68	64	5780	Pancreas, bile duct, lung, prostate, lymph nodes	Thyroid carcinoma
P5	F	62	61	61	11600	Pancreas, bile duct, salivary gland*, periaortitis, lymph nodes, pituitary	Thyroid carcinoma
P6	M	72	68	68	3490	Pancreas, bile duct, lymph nodes	Rectal cancer
P7	M	60	58	58	2410	Pancreas, bile duct, lymph nodes	Renal cancer
P8	M	68	63	68	3520	Pancreas, bile duct, retroperitoneal fibrosis, lung, kidney, artery, lymph nodes	Rectal cancer
P9	M	36	30	35	12400	Pancreas, bile duct	Skin cancer
P10	M	52	49	52	10000	Pancreas, parotid gland*, lacrimal gland, lung, prostate, lymph nodes	Thyroid carcinoma
P11	F	70	68	69	17300	Parotid gland, lacrimal gland, salivary gland, sinus	Lung cancer
P12	M	82	79	79	58000	Pancreas, lacrimal gland*	Colon cancer
P13	F	50	49	45	14300	Uterus*, ovary	Ovarian carcinoma
P14	F	52	46	50	10000	Parotid gland*, lacrimal gland	Breast cancer
P15	F	60	55	57	12500	Pancreas, parotid gland, lacrimal gland, lymph nodes, sinus	Lymphoma
P16	M	42	37	40	7490	Lung*, lymph nodes	Renal cancer
P17	M	71	68	69	415	Pancreas*, bile duct	Prostate cancer