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

References:

- [1] Cavalli G, et al. Treating rheumatological diseases and co-morbidities with interleukin-1 blocking therapies. *Rheumatology* (2015)
- [2] Cavalli G, et al. Efficacy of Canakinumab as First-Line Biologic Agent in Adult-Onset Still's Disease. *Arthritis Res Ther* (2019)

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FRI0507

COLCHICINE INTOLERANCE IN FMF PATIENTS AND PRIMARY OBSTACLES FOR OPTIMAL DOSING

H. Satış¹, B. Armagan², E. Bodakci³, N. Atas¹, A. Sarı², D. Yapar¹, N. S. Yasar Bilge³, R. Bilici Salman¹, G. K. Yardımcı², H. Babaoglu¹, L. Kılıç², M. A. Öztürk¹, B. Goker¹, S. Haznedaroglu¹, U. Kalyoncu², T. Kaşifoğlu³, A. Tufan¹. ¹Gazi University Faculty of Medicine Hospital, Ankara, Turkey; ²Hacettepe University Faculty of Medicine, Ankara, Turkey; ³Eskişehir University Faculty of Medicine Hospital, Eskişehir, Turkey

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

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

inflammation and proteinuria were more common in colchicine intolerant patients and they had reported more frequent attacks compared to those tolerating optimal doses.

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

References:

- [1] Sönmez, H.E., E.D. Batu, and S. Özen, *Familial Mediterranean fever: current perspectives*. Journal of inflammation research, 2016. **9**: p. 13.
- [2] Sarı, İ., M. Birlık, and T. Kasifoğlu, *Familial Mediterranean fever: an updated review*. European journal of rheumatology, 2014. **1**(1): p. 21.
- [3] Özen, S., et al., *EULAR recommendations for the management of familial Mediterranean fever*. Annals of the rheumatic diseases, 2016. **75**(4): p. 644-651.

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FRI0508

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

H. Yang¹, H. Tang¹, P. Zhang¹, Y. Fei¹, H. Chen¹, X. Zhang¹, Y. Zhao¹, F. Zhang¹, W. Zhang¹. ¹Peking Union Medical College Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, Beijing, China

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 malign- ancy	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, retro- peritoneal 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, pros- tate, 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

Table 2. Related factors for malignancies in patients with IgG4-RD

Variables	Univariate analysis		Multivariate analysis	
	Univariate OR (95%CI)	P-value	Multivariate OR (95%CI)	P-value
Head and Neck involvement	0.304 (0.097- 0.952)	0.041	0.604 (0.152-2.401)	0.474
Autoimmune pancreatitis	5.359 (1.651-17.393)	0.005	6.230 (1.559-24.907)	0.010
Eosinophilia	0.117 (0.014-0.966)	0.046	0.094 (0.010-0.883)	0.039

Figure 1. Standardized incidence ratios of malignancies in patients with IgG4-RD from different studies.

Methods: We prospectively analyzed the IgG4-RD patients recruited in Peking Union Medical College Hospital from January 2011 to August 2018 and identified patients diagnosed with IgG4-RD complicating malignancies. Data regarding demographics, clinical features, treatment and prognosis of IgG4-RD patients complicating malignancies were collected and compared to those of age- and sex-matched controls.

Results: Among the 587 Chinese patients with IgG4-RD, 17 malignancies were identified. Ten of them developed malignancy after the diagnosis of IgG4-RD, given a standard incidence ratio (SIR) of 2.78 (95%CI 1.33-5.12). Multivariate logistic analysis indicated that autoimmune pancreatitis (OR= 6.230, 95%CI 1.559-24.907, $p=0.010$) was positively associated with malignancy, whereas eosinophilia (OR= 0.094, 95%CI 0.010-0.883, $p=0.039$) was negatively related with malignancies. During a median follow-up period of 61.4±26.4 months, all patients with IgG4-RD and malignancies survived.

Conclusion: An increased incidence of malignancy was found in Chinese IgG4-RD cohort. Autoimmune pancreatitis is a potential risk factor, whereas eosinophilia is a possible protective factor for complicating malignancies.

References:

- Wallace, Z. S., et al. Association of IgG4-Related Disease With History of Malignancy. *Arthritis Rheumatol* 68, 2283–2289 (2016).
- Hirano, K. et al. Incidence of malignancies in patients with IgG4-related disease. *Intern. Med.* 53, 171–176 (2014).

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FRI0509

COMPARISON OF MS SCORE AND HSCORE FOR THE DIAGNOSIS OF ADULT-ONSET STILL'S DISEASE ASSOCIATED MACROPHAGE ACTIVATION SYNDROME

L. Zhang¹, X. Yang¹, T. Li¹, S. Liu¹. ¹1st affiliated hospital of Zhengzhou University, Zhengzhou, China

Background: Adult-onset Still's disease (AOSD) is a rare multisystemic auto-inflammatory disease. Macrophage activation syndrome (MAS) is a life-threatening complication of rheumatic disorders, including sJIA, adult-onset Still's disease (AOSD) and lupus. Many tools have been developed for the diagnosis of MAS, including HScore and MS score. Although some authors have tried to apply these tools for the diagnosis of AOSD-associated MAS, there are no studies comparing the diagnostic ability of HScore and MS score.

Objectives: We aimed to compare the capacity of HS score and MS score for the diagnosis of AOSD-associated MAS.

Methods: Patients diagnosed as AOSD during January, 2012 and October, 2019 in our hospital were retrospectively analyzed. Clinical and laboratory data were compared between patients with pure AOSD and those with AOSD associated MAS. HScore and MS score were calculated respectively and the diagnostic capacity of MAS was compared by means of receiver operating characteristic (ROC) curve analysis.

Results: We included 174 patients with pure AOSD and 35 patients with AOSD-associated MAS. Patients with AOSD-associated MAS were younger than those with pure AOSD (32.4±11.4 yrs vs 36.9±13.5 yrs, $P=0.028$). More death were observed among patients with AOSD-associated MAS (17.1% vs 3.4%, $P=0.001$). Patients with AOSD-associated MAS had higher HScore (median [range] 196 [98-333] vs 68 [33-156], $P<0.001$) and higher MS score (median [range] 1.05 [-1.26-26.55] vs -1.17 [-1.26-2.52], $P<0.001$) than those with pure AOSD. The difference of different parameters of these two groups of patients

Figure 1. ROC curve of HScore and MS score. HScore=120, sensitivity=90.6%, specificity=89.6%. MS score=-0.45, sensitivity=75%, specificity=73%. AUC-HScore=0.973, AUC-MS score=0.865, $P<0.001$

were detailed in Table 1 and Table 2. ROC curve analysis (Figure 1) revealed that HScore has stronger ability to diagnose AOSD-associated MAS compared with MS score (AUC=0.973 and 0.865 for HScore and MS score respectively, $P<0.001$). HScore≥120 perform best (sensitivity 90.6% and specificity 89.6%). MS score≥-0.25 yielded a sensitivity of 75% and a specificity of 73%.

Conclusion: HScore seems to perform much better than MS score for the diagnosis of AOSD-associated MAS in our cohort.

References:

- Siddiqui M, Putman MS, Dua AB. Adult-Onset still's disease: current challenges and future prospects. *Open Access Rheumatol* 2016;8:17–22.
- Bracaglia C, Prencipe G, De Benedetti F. Macrophage activation syndrome: different mechanisms leading to a one clinical syndrome. *Pediatr Rheumatol Online J* 2017;15.
- Guilpain P, Le Quellec A. About the complexity of adult onset still's disease... and advances still required for its management. *BMC Med* 2017;15:5.
- Minoia F, Bovis F, Davi S, et al. Development and initial validation of the MS score for diagnosis of macrophage activation syndrome in systemic juvenile idiopathic arthritis. *Ann Rheum Dis* 2019;78:1357–1362.
- Fardet L, Galicier L, Lambotte O, et al. Development and validation of the HScore, a score for the diagnosis of reactive hemophagocytosis syndrome. *Arthritis Rheumatol.* 2014 Sep;66(9):2613-20.
- ED Batu, A Erden, E Seyhoğlu, et al. Assessment of the HScore for reactive haemophagocytic syndrome in patients with rheumatic diseases. *Scand J Rheumatol.* 2017 Jan;46(1):44-48.
- Wang R, Li T, Ye S, et al. Application of MS score in macrophage activation syndrome patients associated with adult onset Still's disease. *Ann Rheum Dis.* 2019 Oct 14. pii: annrheumdis-2019-216286.

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Public health, health services research, and health economics

FRI0510

THE FREQUENCY OF CONTRACEPTION DOCUMENTATION IN WOMEN ON AND OFF TERATOGENIC ANTI-RHEUMATIC MEDICATIONS IN THE RISE REGISTRY

M. Clowse¹, J. Li², M. Birru Talabi³, A. Eudy¹, G. Schmajuk². ¹Duke University, Durham, United States of America; ²UC San Francisco, San Francisco, United States of America; ³UPMC, Pittsburgh, United States of America

Background: Several of the most commonly prescribed medications for women with rheumatic disease are teratogens, posing a risk for pregnancy loss and birth defects if taken in pregnancy. To prevent these life-altering complications, it is important that all women taking teratogenic medications avoid pregnancy through abstinence or contraception.

Objectives: We sought to understand the accessibility to contraceptive data within the RISE Registry and to test whether, compared to other women,