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FRI0615

EVALUATION OF COMPLIANCE AND RELATED FACTORS IN COLCHICINE TREATMENT IN FAMILIAL MEDITERRANEAN FEVER PATIENTS

Emre Tekgoz¹, Seda Colak¹, Fatma İknur Cinar², Sedat Yılmaz¹, Muhammet Çınar¹. ¹University of Health Sciences, Gulhane Faculty of Medicine, Department of Internal Medicine, Division of Rheumatology, Ankara, Turkey; ²University of Health Sciences, Gulhane Faculty of Nursing, Department of Internal Medicine Nursing, Ankara, Turkey

Background: Familial Mediterranean Fever (FMF) is an autoinflammatory disease requiring long-term treatment. Increasing the compatibility with colchicine treatment in patients with FMF is the first step for preventing amyloidosis. Patients' beliefs about medicines and treatment may affect treatment adherence and treatment success.

Objectives: The aim of this study was to determine adherence to colchicine treatment and related factors in FMF patients. In addition, patients' beliefs about colchicine, which are one of the important factors affecting the treatment adherence of patients, were evaluated.

Methods: Total of 179 patients with FMF was included in this study. The demographic and clinical features and MEFV gene mutations were recorded. The treatment adherence of the patients was assessed using by Compliance Questionnaire on Rheumatology (CQR). The Beliefs About Medicines Questionnaire (BMQ-T) was used to assess patient's beliefs about colchicine. The relationship between compliance of treatment and clinical characteristics of patients were assessed.

Results: One hundred thirteen (63.1%) of the patients were male. The mean age of patients was 34.5 ± 12.7 years and mean delay in diagnosis was 6.7 ± 8.4 years. The mean dose of colchicine was 1.37 ± 0.43 mg/day and, the percentage of patients using colchicine regularly was 66.5%. Adherence to treatment was higher in patients with concomitant diseases than those without comorbidities (p = 0.028). In addition, treatment compliance was higher in married patients compared to single patients (p = 0.013). The colchicine dose used in compatible patients was higher than in non-compatible patients (p = 0.033) (Table 1). We also found that as the BMQ-T Specific Necessity scores increased, compliance with treatment increased. On the other hand, as the BMQ-T General Overuse and General Harm scores increased, non-compliance with treatment increased (Table 2).

Table 1. Relationships between socio-demographic and clinical characteristics of patients and CQR score (n = 179)

Characteristics	CQR score		
	≥80% n=29	< 80% n=150	p
Age (years) median (25th–75th percentile)	37.0 (28.5–44.5)	29.0 (24.0–44.0)	0.099 ^a
Gender (n,%)			0.164 ^b
Male	15 (13.3)	98 (86.7)	
Female	14 (21.2)	52 (78.8)	
Marital status, (n,%)			0.013 ^b
Married	21 (22.8)	71 (77.2)	
Single/widowed/divorced	8 (9.2)	79 (90.8)	
Comorbidities, (n,%)			0.028 ^b
Yes	12 (26.7)	33 (73.3)	
No	17 (12.7)	117 (87.3)	
Delay of diagnosis (years)			0.920 ^a
Median (25th–75th percentile)	3.0 (0.5–10.0)	3.0 (1.0–9.0)	

Table 1. Demographic features and treatment response during anakinra therapy

Patient ID #	Age	Sex	Diagnosis	Duration of pericarditis follow-up (mo)	Prior medications	Number of recurrences before anakinra	Anakinra treatment duration (mo)	Time to corticosteroid discontinuation (mo)	Number of recurrences after daily dose of anakinra
1*	23	M	IRP	129	Colchicine, NSAIDs, CS, HCQ	6	52	9	No
2	32	F	IRP	128	Colchicine, NSAIDs	7	4	NA	No
3*	40	F	IRP	21	Colchicine, CS	6	8	1	No
4	20	M	IRP	11	Colchicine, CS	3	8	2	No
5	25	M	FMF	30	Colchicine, CS	5	15	1	No

F: Female, M: Male, CS: Corticosteroid, HCQ: Hydroxychloroquine, NA: Not applicable

*Dose tapering was unsuccessful in these patients

Colchicine dose, (mg/day) 1.5 (1.25–1.75) 1.5 (1.0–1.5) **0.033^a**
 Median (25th–75th percentile)
 Data represented either as the median (25th–75th percentile) or as the frequency. a: Mann-Whitney test b: Pearson Chi-Square test, CQR, Compliance Questionnaire on Rheumatology

Table 2. Beliefs about Medicines Questionnaire Scale Scores in Adherent and Nonadherent Patients.

Variable	CQR score			
	Discontinuous		Z	p
BMQ-T-Specific Necessity	≥80% n=29	< 80% n=150	-5.260	< 0.001
BMQ-T-Specific Concerns	2.8 (2.2–3.8)	2.8 (2.4–3.6)	-0.100	0.920
BMQ-T-General Overuse	2.25 (1.75–2.63)	2.75 (2.25–3.25)	-2.936	0.003
BMQ-T-General Harm	1.75 (1.38–2.63)	2.38 (2.0–3.0)	-3.212	0.001

Z; Mann-Whitney test, Data represented as median (25th–75th percentile, CQR; Compliance Questionnaire on Rheumatology

Conclusion: In patients with FMF, it is important to evaluate the compliance with the treatment due to the importance of colchicine to prevent amyloidosis that may occur in patients without treatment. As this study shows it is also important to determine patients' beliefs about medicine in terms of their influence on patients' compliance with treatment.

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ANAKINRA TREATMENT IN RECURRENT PERICARDITIS: SINGLE CENTER EXPERIENCE

Zeynep Toker Dincer, Osman Corbali, Serdar Ugurlu, Huri Ozdogan. University of Istanbul-Cerrahpasa, Division of Rheumatology, Department of Internal Medicine, Istanbul, Turkey

Background: Recurrent pericarditis (RP), however the etiology is unknown in the majority, may be observed in autoinflammatory diseases such as familial Mediterranean fever (FMF) and tumor necrosis factor receptor-1 associated periodic syndrome (TRAPS). Colchicine has long been used to treat pericarditis related to FMF as well as patients with idiopathic recurrent pericarditis (IRP) (1). Alternative treatments have been reported for cases with colchicine resistant RP.Objectives:

Objectives: The aim is to present our data regarding anakinra treatment in recurrent pericarditis either related to FMF or idiopathic, who are resistant to colchicine.

Methods: Patients who had received anakinra with a diagnosis of recurrent pericarditis either idiopathic or secondary to FMF followed in our autoinflammatory disease center between 2014–2018 are evaluated retrospectively. From patients' files, demographic and clinical features, response to other treatment approaches such as NSAID, corticosteroid, colchicine, were evaluated. All patients have been genetically screened for monogenic autoinflammatory diseases (MEFV, TRAPS, MVK, NLRP3, NOD2). Patients who had at least 3 attacks were administered anakinra 100 mg/day. Therapeutic efficacy, as well as side effect profile of anakinra is also assessed.

Results: There were 5 patients (3 male and 2 female) with the diagnosis of RP, 1 was related to FMF and 4 were idiopathic. The mean age of

the group was 28±8 (range 20-40). All patients diagnosed with IRP were negative for autoinflammatory genetic screening, while a MEFV variant (K695R het.) was detected in the FMF patient. Median duration of follow-up was 30 months (range 11-129). In table 1, demographic and clinical features are given. The median number of recurrence was 6 before anakinra treatment. No episode of pericarditis was observed in any of the patients after the initiation of anakinra. The response to anakinra persisted even after the dose was reduced to 100 mg/alternate day in 3 patients, however in 2, recurrence of pericarditis was observed and anakinra was escalated to initial dose. It was possible to discontinue corticosteroid treatment in all patients. Currently all patients continue anakinra treatment. No side effect including injection site reaction, has been observed by now.

Conclusion: Anakinra seems to be a safe and effective treatment approach for colchicine resistant recurrent pericarditis. However recurrence may occur during dose tapering.

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FRI0617 APPLICATION OF AUTO-INFLAMMATORY DISEASE DAMAGE INDEX (ADDI) TO PATIENTS WITH FMF AND RELATED FACTORS WITH DAMAGE

Hakan Babaoglu¹, Berkan Armagan², Erdal Bodakci³, Timuçin Kaşifoğlu³, Hasan Satış¹, Nuh Atas¹, Alper Sari², Nazife Sule Yasar Bilge³, Gözde Kübra Yardımcı², Reyhan Bilici Salman¹, Levent Kılıç², Mehmet Akif Öztürk¹, Berna Goker¹, Semir Haznedaroğlu¹, Umut Kalyoncu², **abdurrahman tufan¹**.

¹Gazi University Faculty of Medicine, Department of Internal Medicine-Rheumatology, Ankara, Turkey; ²Hacettepe University Faculty of Medicine, Department of Internal Medicine-Rheumatology, Ankara, Turkey; ³Eskisehir Osmangazi University Faculty of Medicine, Department of Internal Medicine-Rheumatology, Eskisehir, Turkey

Background: Familial Mediterranean Fever (FMF) is the most frequent auto-inflammatory disease caused by MEFV gene mutations. Available reports investigated only specific components of damage such as amyloidosis. All possible organ targets of damage have not been entirely evaluated before. Such as Disease severity index which is emerged especially for FMF do not cover entire damage domains related to FMF. Recently, a new scoring system (Auto-inflammatory disease damage index) was developed and validated for autoinflammatory diseases.

Objectives: We aimed to investigate damage accrual caused by FMF and associated features with damage.

Methods: All patients recruited from FMF in Central Anatolia (FiCA) cohort, which is a duplication disabled, internal and externally controlled, cross-sectional, multicenter accessible web-based cohort. This study is comprising 970 adult patients (mean age 35.3 ±12.1 years, 61.5% female). Demographic data, FMF disease characteristics, co-morbid conditions, disease complications were meticulously questioned and laboratory features and genotype data (if available) were recruited from patient files. FMF caused damage was assessed by auto-inflammatory disease damage index (ADDI) which is recently validated. Association between damage and demographic, disease and treatment characteristics were analyzed.

Results: Proportions of FMF manifestations were fever 83.1%, peritonitis 91.5%, pleuritis 47.9%, arthritis 43.3% and skin rash 26.2%. Dominant attack types were fever in 6.2%, serositis in 65.7%, musculoskeletal in 16.8% and all types of attacks were common in rest of patients. MEFV mutations were available in 814 subjects and 75.9% of these subjects were harboring M694V mutation (42.5% homozygous for M694V). Among all 63.1% patients were well responded to colchicine and 8.8% were non-responders. Median ADDI score was 1 (min 0-max 11). Most common FMF related damages were observed in musculoskeletal, reproductive and kidney domains. Chronic musculoskeletal pain was present in 49%, joint deformity in 2.9%, infertility in 6.6%, amenorrhea in 3.9%, proteinuria in 6.9%, amyloidosis in 5.9% and renal failure in 3.7% of the patients. 411 (%42.3) of patients had no damage accrual. M694V homozygous mutation, male gender and colchicine nonresponse were found to be the independent predictors of damage.

Conclusion: M694V homozygous mutation, colchicine non-response and male gender are predictors of damage and effective therapeutic interventions must be undertaken to prevent from damage in these patients.

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FRI0618 ANALYSIS OF NEW REFERRALS TO A SPECIALIST UK ADULT AUTOINFLAMMATORY DISEASE SERVICE

Serdal Ugurlu¹, Philip N. Hawkins², Charalampia Papadopoulou², Tamer Rezk², Dorota Rowczenio², Helen J. Lachmann². ¹Cerrahpaşa Faculty of Medicine, Division of Rheumatology, Istanbul, Turkey; ²UCL Division of Medicine and Royal Free Hospital London NHS Foundation Trust, National Amyloidosis Centre, London, United Kingdom

Background: Diagnosis of the systemic autoinflammatory diseases (SAIDs) requires a high index of suspicion and previous series has suggested that there are often long diagnostic delays, particularly in TRAPS and MKD.

Objectives: To look at the case mixed referred to a single adult clinic in London specialising in assessment of potential SAIDs over the course of the year of 2017.

Methods: All new referrals were accepted for clinical assessment. At the first visit patients had a full history and examination, genetic testing – varying from single gene to a 20 gene panel depending on clinical features, and laboratory testing including fortnightly blood draws for serial analysis of the hepatic acute phase response proteins, CRP and SAA over a 3 month period.

Patients with a non suggestive history, non contributory genetic testing and no evidence of inflammation accompanying symptoms were felt not to have SAIDs and referred back to their local hospitals for further management. Other cases were diagnosed based on full clinical assessment, other investigations – for example ferritin in AOSD, genetic testing results, serial monitoring of CRP and SAA and therapeutic trials, for example colchicine in presume FMF and anti IL-1 therapies in CAPS and Schnitzler's syndrome.

Results: 273 new patients were referred. Median age at referral was 37.4 years, the oldest patient was 84.3 years old and 59% were female. 174 (64%) were of northern European ancestry, 68 (25%) were eastern Mediterranean, west Asian or southern European ancestry, 19 (7%) were of south or east Asian ethnicity and 4% were of African or Afro-Caribbean ancestry. 76% of referrals were from hospital specialities. The referral source was: rheumatology 38%, general practitioner 24%, dermatology 8%, immunology 8%, gastroenterology 6%, infectious diseases 3%, clinical genetics 3%, nephrology 2%, haematology 2%, gynaecology 2%, emergency department 1%, respiratory 1%, other 2%.

After work up 135 (49.5%) were felt not to have a SAID as the cause of their symptoms. Of the remaining 138 patients who did have evidence of a SAID the diagnoses made were: FMF 33%, uncharacterised SAID 26%, CAPS 9%, AOSD 8%, recurrent idiopathic pericarditis 6%, Schnitzler's Syndrome 5%, TRAPS 4%, variant PFAPA 4%, DADA2 1%, MKD 1%, CRMO 1%, Behçets 1%, Cattleman's disease 1%.

The median interval between reported symptom onset and diagnosis were as follows: 16 yrs for FMF, 28.1 yrs for CAPS, 5.0 years for recurrent idiopathic pericarditis, 4.5 yrs for Schnitzler's Syndrome, 5.7 yrs for TRAPS, 20.5 yrs for variant PFAPA, 12.5 yrs for DADA2, 17 yrs for MKD and 2 years for CRMO.

Conclusion: This series suggests that recognition and diagnosis of the SAIDs remains a challenge. More than 1/3 of referrals were from rheumatology, referrals from primary care were almost exclusively from patients with a known family history of one the inherited syndromes. The wide variety of referring specialities reflects the diverse nature of SAIDs and the importance of almost all specialities considering the possibility of SAIDs. Only just over 50% referrals had evidence of diseases falling within the recognised SAID spectrum and 26% of these have currently uncharacterised disease with no diagnostic genetic testing. Of those in whom a diagnosis could be made there are significant diagnostic delays fortunately despite late initiation of treatment no patients had evidence of systemic AA amyloidosis.