complications. To establish the relationship between its value and the AOSD-related mortality.

Methods: Retrospective study realized in two University Hospitals. Clinical, laboratory, AOSD-related complications data, administered biologic treatments and number of deaths (AOSD related or not) were recorded. Each patient was characterized for the presence of AOSD-related complications such as macrophage activation syndrome (MAS), myocarditis, lung involvement (pulmonary hypertension, interstitial infiltrate), renal involvement (tubulointerstitial nephritis, acute renal failure), secondary amyloidosis and AOSD-related death. SSS was applied at the onset of the disease development, assigning a point to each of the next 12 variables: fever, exanthema, pleuritis, pneumonitis, pericarditis, alteration of liver tests or hepatomegaly, splenomegaly, lymphadenopathy, odynophagia, leukocytosis $>15,000/\text{mm}^3$, myalgia and abdominal pain. A ≥ 7 score has been validated on other populations as the one which identifies the patients with high risk of complications. The relationship between SSS value and the next parameters was determined: clinical course, complications, biologic treatments administered and AOSD-related mortality.

Results: Data from 64 patients was analyzed (40,6% men, mean age 37 years). SSS values of <7 were obtained in 50 patients (78,3%) and of ≥ 7 in 14 patients (21,7%). MAS and renal involvement were significantly related with a score of ≥ 7 (Table 1). Lung involvement, myocarditis and secondary amyloidosis were not significantly associated with a high SSS value. Even so, analyzing each case, it was found that individually they had a score of ≥ 7 (except for lung involvement). Biologic treatment requirement along the disease course was related to SSS ≥ 7 . Moreover, SSS value was not related to the different clinical patterns or the not-related-AOSD deaths. It was not possible to determine the relationship between the score and the AOSD complications-related deaths due to having only two registered cases. Despite this, it was found that individually both had a SSS of ≥ 7 .

TABLE 1. AOSD RELATED COMPLICATIONS

COMPLICATIONS	SSS <7 N= 50	SSS ≥ 7 N= 14	P-Value
MAS	2	3	<0.05
Myocarditis	0	1	0,500
Lung involvement	1	1	0,920
Renal involvement	0	3	<0.05
Secondary amyloidosis	0	2	0,06
Biologic treatment	14	10	<0.05
All-cause deaths	3	2	0,970

Conclusion: The prognosis score described by *Pouchot et al* could be useful to identify those patients with high risk of developing clinical complications and those who will need biologic treatment along the course of their disease. It is necessary a higher number of patients to determine if the score could be useful to estimate the death risk related to AOSD complications.

REFERENCES:

- [1] Ruscitti, et al. Adult-onset Still's disease: evaluation of prognostic tools and validation of the systemic score by analysis of 100 cases from three centers. *BMC Medicine* (2016)14:194

Disclosure of Interests: Ivette Casafont-Solé: None declared, Susana Holgado: None declared, J. Narváez Consultant for: Bristol-Myers Squibb, Maribel Mora: None declared, Josep Roca: None declared, Anahy Brandy-Garcia: None declared, Lourdes Mateo: None declared, Melania Martínez-Morillo: None declared, Laia Gifre: None declared, Maria Aparicio Espinar: None declared, Águeda Prior-Español: None declared, Anne Ríveros: None declared, Clara Sanguesa: None declared, Jordi Camins-Fàbregas: None declared, Annika Nack: None declared, Joan Miquel Nolla: None declared, Alejandro Olive: None declared

DOI: 10.1136/annrheumdis-2019-eular.5798

THU0559

PSYCHOLOGICAL EFFECTS OF A BEHÇET'S DIAGNOSIS ON DISEASE ACTIVITY PERCEPTION- A PROSPECTIVE OBSERVATIONAL STUDY

Casandra Buzatu^{1,2}, Hannah Nicholson³, Roisin Cunningham^{2,3,4}, Robert J Moots^{2,3,4}. ¹Research Center of Rheumatic Diseases, Sf Maria Hospital, University of Medicine and Pharmacy Carol Davila, Bucharest, Romania; ²Liverpool Behçet's Centre of Excellence, Liverpool, United Kingdom; ³University of Liverpool, Liverpool, United Kingdom; ⁴Institute of Ageing and Chronic Disease, Liverpool, United Kingdom

Background: There is a pressing need to understand more about the psychological experience in patients with Behçet's Disease (BD). The presence of poor wellbeing, low mood and anxiety may affect the ability to both manage and cope with this complex disorder and also impact upon the perception and experience of disease activity^{1,2,3}.

Objectives: Our aim was to investigate the relationship between patient rating of disease activity and their psychological experience (anxiety, low mood, illness perception and coping style) in a UK Centre of Excellence for BD.

Methods: This study had full UK research ethics approval.

A 12-month prospective observational study of a cohort of patients diagnosed with BD according to the International Criteria for BD (ICBD) under surveillance in the BD Centre of Excellence Liverpool, UK. All patients that agreed to a psychological evaluation were included. Participants were described for demographics and clinical presentation. BD Current Activity Form 2006 and a clinician's and patient's visual analogue scale (VAS) were recorded. Participants completed validated self-reported measures: Brief COPE, IPQ-R, PHQ-9, GAD-7 and WEMWBS. Spearman Rank correlations, multivariate regression analysis and mediation analysis were used to investigate the relationship between patient perception of disease activity, coping mechanisms and mood. For data analysis Excel and SPSS were used.

Results: 86 patients were selected (24 males, 68 females; mean age at time of evaluation 43.58 years ± 11.8 SD; mean disease duration 9.76years ± 10.01). The correlation between Clinician's VAS and Patient's VAS was strong 0.754 ($p<0.005$). Strong correlations were also observed between Patient's VAS and low mood (PHQ-9) $r=0.533$ ($p<0.005$), anxiety (GAD-7) $r=0.433$ ($p<0.05$), illness perception- emotional representation (IPQ-R) $r=0.377$ ($p<0.05$) and emotion-focused coping (COPE) $r=0.320$ ($p<0.05$).

The results of the mediation analysis indicated that patient's perception of disease activity was a significant predictor for Dysfunctional Coping Strategies ($b=1.5, se=0.57$, $p<0.05$). Dysfunctional Coping Strategies had a mediating impact on the relationship between patient perception of disease activity (VAS) and mood 0.645 (CE: 0.68-1.389).

Conclusion: Psychological experience plays an important part in the patient evaluation and experience of disease activity. Patient coping strategies and psychological representation of illness experience influence their perception of disease activity and their mood. This highlights the need of patients for psychological support and for improving coping mechanisms in order to obtain a better disease control and a higher quality of life.

REFERENCES:

- [1] RJ Moots, S Campbell, Is Psychological Status Related to Symptom Experience in Behçet's Syndrome? 2014 ACR/ARHP Annual Meeting
- [2] IM Atay, I Erturan, A Demirdas, GB Yaman, VA Yürekli The impact of personality on quality of life and disease activity in patients with Behçet's disease: a pilot study *Compr Psychiatry*. 2014 Apr;55(3):511-7
- [3] MA Melikoglu, M Melikoglu The relationship between disease activity and depression in patients with Behçet disease and rheumatoid arthritis. *Rheumatol Int*. 2010 May;30(7):941-6

Disclosure of Interests: Casandra Buzatu: None declared, Hannah Nicholson: None declared, Roisin Cunningham: None declared, Robert J Moots Grant/research support from: Biogen, Bristol-Myers Squibb, Chugai, Novartis, Pfizer Inc, Roche, Sandoz, and UCB, Consultant for: Biogen, Bristol-Myers Squibb, Chugai, Novartis, Pfizer Inc, Roche, Sandoz, and UCB, Speakers bureau: Biogen, Bristol-Myers Squibb, Chugai, Novartis, Pfizer Inc, Roche, Sandoz, and UCB

DOI: 10.1136/annrheumdis-2019-eular.6274

THU0560

DISTINCTIVE IMAGING FEATURES BETWEEN IGG4-RELATED OPHTHALMIC DISEASE AND GRAVES' ORBITOPATHY: A COMPARATIVE STUDY

Eduardo Martín-Nares¹, Oziel Amaya-Piña², Rosa Delia Delgado-Hernández², Gabriela Hernandez-Molina¹. ¹Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán, Immunology and Rheumatology, Mexico City, Mexico; ²Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán, Department of Radiology and Molecular Medicine, Mexico City, Mexico

Background: IgG4-related ophthalmic disease (IgG4-ROD) may present as a cause of orbital myositis leading to proptosis and diplopia. This clinical scenario can be mistakenly diagnosed as Graves' orbitopathy (GO), preventing a timely and adequately treatment.

Objectives: To elucidate if there are specific radiological features that might differentiate between IgG4-ROD and GO by imaging.

Methods: We included 19 patients with diagnosis of IgG4-related disease (IgG4-RD) according to the Comprehensive Diagnostic Criteria for IgG4-RD, who regularly attended a tertiary referral center in Mexico City. All the patients had ophthalmic involvement and available computed tomography (CT) and/or magnetic resonance imaging (MRI) of the orbits. We also included 32 patients with GO with available CT and/or MRI of the orbits. Imaging studies were evaluated by a blinded head and neck radiologist for the following features: exophthalmos, extraocular muscles (EOM) size and morphology, lacrimal gland enlargement, orbital fat involvement, stretching of the optic nerve (ON), ON sheath thickening and orbital bone changes.

Results: Both groups were similar in age (49.1±15.8 vs. 51.6±14.7, p=0.58) and gender (men 58.9% vs. 40.6%, p=0.23). In addition to ophthalmic involvement, 18 (94.7%) IgG4-RD patients had extra-ophthalmic involvement with a median number of organs involved of 7 (1-12), mainly submandibular glands (73.7%), lymph nodes (68.4%), parotid glands (63.2%) and pancreas (47.4%). Three patients were misdiagnosed as GO before IgG4-RD diagnosis. Graves' disease was the underlying thyroid disorder in 28 (87.5%) GO patients, Hashimoto's thyroiditis in 2 (6.3%), papillary thyroid carcinoma in one (3.1%) and one patient was euthyroid with positive thyroid stimulating immunoglobulin. The prevalence of exophthalmos (78.9% vs. 93.8%), bilateral involvement (78.9% vs. 87.8%) and overall EOM involvement (47.4% vs. 68.8%) was similar between IgG4-ROD and GO groups. However, IgG4-ROD patients had a higher frequency of lacrimal gland involvement (73.7% vs. 10.7%, p=0.001) and a tendency for the lateral rectus to be the most frequently involved EOM (22.2% vs. 0%, p=0.07); conversely they had a lower prevalence for the inferior rectus to be the most frequently involved EOM (33.3% vs. 72.7%, p=0.04), orbital fat involvement (47.4% vs. 81.3%, p=0.01), ON stretching (57.9% vs. 87.5%, p=0.02) and orbital bone changes (0% vs. 25%, p=0.02). EOM bellies were involved in all the IgG4-ROD and GO cases, whereas EOM tendon involvement was present in 9% of GO and in none of the IgG4-ROD group. Patients with IgG4-ROD had more frequently the combination of lacrimal gland and lateral rectus (31.6% vs. 3.1%, p=0.008) and less frequently the combination EOM and orbital fat involvement (21.2% vs. 59.4%, p=0.008).

At the logistic regression analysis we found an association of lacrimal gland involvement (OR 64.4, 95% CI 6.8-609.5, p=0.001) with IgG4-ROD. In a second model including combined variables, the combination of lacrimal gland and lateral rectus involvement was associated with IgG4-ROD (OR 62.5, 95% CI 3.31-1000, P=0.006), whereas the presence of EOM and orbital fat involvement was protective (OR 0.05, 95% CI 0.006-0.48, p=0.009).

Conclusion: Imaging features may reliably differentiate between IgG4-ROD and GO. The presence of both lacrimal gland and lateral rectus enlargement must alert clinicians to consider IgG4-ROD diagnosis.

REFERENCES:

- [1] Tiegs-Heiden CA, Eckel LJ, Hunt CH, et al. Immunoglobulin G4-related disease of the orbit: imaging features in 27 patients. *AJNR Am J Neuroradiol.* 2014 Jul;35(7):1393-7.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5144

THU0561

CANAKINUMAB FOR THE TREATMENT OF ADULT ONSET STILL'S DISEASE TO ACHIEVE REDUCTION OF ARTHRITIC MANIFESTATION AT WEEK 12: AN INVESTIGATOR-INITIATED MULTI-CENTRE, PLACEBO-CONTROLLED STUDY (CONSIDER)

Claudia Kedor¹, Joachim Listing², Jan Zernicke¹, Anja Weiß², Frank Behrens³, Norbert Blank⁴, Jörg Henes⁵, Jörn Kekow⁶, Andrea Rubbert-Roth⁷, Hendrik Schulze-Koops⁸, Eva Seipelt⁹, Christof Specker¹⁰, Eugen Feist¹, The Study was supported by Novartis Germany. ¹Charité – Universitätsmedizin Berlin, Rheumatology and clinical Immunology, Berlin, Germany; ²Deutsches Rheuma-Forschungszentrum (DRFZ), Epidemiology, Berlin, Germany; ³Goethe University Frankfurt, Fraunhofer Institut für Molekularbiologie und Angewandte Onkologie IME, Frankfurt a.M., Germany; ⁴University Hospital Heidelberg, Internal Medicine 5, Rheumatology, Heidelberg, Germany; ⁵University Hospital Tuebingen, Centre for Interdisciplinary Clinical Immunology, Rheumatology and Auto-inflammatory Diseases (INDIRA) and Internal Medicine II, Tübingen, Germany; ⁶Helios Fachklinik Vogelsang-Gommern, Klinik für Rheumatologie, 39245 Vogelsang-Gommern, Germany; ⁷Kantonsspital St Gallen, Klinik für Rheumatologie, St Gallen, Switzerland; ⁸University of Munich, Department of Internal Medicine IV, Division of Rheumatology, München, Germany; ⁹Immanuel Krankenhaus Berlin, Rheumatologie und Klinische Immunologie, Berlin, Germany; ¹⁰Evangelisches Krankenhaus • Kliniken Essen-Mitte, Klinik für Rheumatologie und Klinische Immunologie, Essen, Germany

Background: Inhibition of interleukin-1 (IL-1) represents a promising treatment option in adult-onset Still's disease (AOSD). Canakinumab (CAN) is approved for treatment of systemic juvenile idiopathic arthritis (sJIA) since 2013 and recently for AOSD in Europe. In sJIA, CAN has a proven marked impact on systemic and articular disease activity, but for AOSD RCT data is limited.

Objectives: Investigation of efficacy and safety of CAN in AOSD patients with joint involvement by means of a multi-centre, double-blinded, randomized, placebo controlled trial.

Methods: AOSD patients with joint involvement (tender and swollen joint count ≥ 4 each) were stratified by pre-treatment status with biologics and randomized in a ratio of 1:1 to CAN (4 mg/kg up to a maximum of 300 mg) or placebo (PBO) s.c. every 4wks. The primary endpoint was defined as the proportion of patients with a clinical-relevant reduction (≥ 1.2) in disease activity as measured by change in disease activity score (DAS28) at week 12. There were two major protocol violations: two PBO patients received blinded CAN at week 4. Therefore, a per-protocol (PP) analysis, which excluded data of these two patients, was performed.

Results: The study was prematurely stopped with 35 out of 68 planned patients enrolled due to approval of CAN for AOSD by EMA. At enrollment, mean DAS28 represented high disease activity for both: 5.4 in CAN (n=18) and 5.3 in PBO (n=17). In the intention-to-treat analysis, 12 (67%) CAN and 7 (41%) PBO patients met the primary endpoint. The 25.5% [95%CI: -10.3%; 55.9%] difference in the response rate did not achieve statistical significance (p=0.18). Using the PP analysis, the treatment difference for primary endpoint was 33.4% [95%CI: -10.3%; 55.9%, p=0.08]. We observed higher ACR50 (50% CAN vs. 6.7% PBO, p=0.009), ACR70 (28% CAN vs. 0% PBO, p=0.049) response rates, (PP analysis). Higher mean improvements in the CAN vs PBO group for DAS28(ESR and CRP), ESR, CRP, 66 swollen and 68 tender joint counts at week 12 were observed (PP analysis). There were 47 and 21 non-serious adverse events (AE) reported in CAN and PBO groups, respectively. Two CAN patients experienced an SAE (hepatotoxicity and patellofemoral pain syndrome).

Conclusion: Although the study was terminated prematurely and the primary endpoint did not achieve statistical significance, treatment of patients with active AOSD with CAN led to improvement of several outcome measures. In addition, safety profile was similar to that reported in sJIA and no unexpected safety issues were observed. This data support the treatment of AOSD patients with CAN.

REFERENCES:

- [1] Ruperto. *NEJM* (2012): 2396-2406

Disclosure of Interests: Claudia Kedor Grant/research support from: Sobi, Consultant for: Novartis, Speakers bureau: Novartis, Roche, Joachim Listing: None declared, Jan Zernicke: None declared, Anja Weiß: None declared, Frank Behrens Grant/research support from: AbbVie, Pfizer, Roche, Chugai, Prophylax, Bioline, Novartis, Consultant for: AbbVie, Pfizer, Roche, Chugai, UCB, Bristol-Myers Squibb, Celgene, MSD, Novartis, Biotech, Janssen, Genzyme, Eli Lilly, Speakers bureau: Ad board: AbbVie, Pfizer, Roche, Chugai, UCB, Bristol-Myers Squibb, Celgene, Novartis, Biotech, Janssen, Genzyme, Eli Lilly, Norbert Blank Grant/research support

Table 1. Characteristics of 9 patients from 1 center

Patient	Age/ Sex	Tumor type	Checkpoint Inhibitor	Fulfilled EULAR/ACR Classification Criteria*	Unfulfilled Criteria	Atypical Features	Treatment
1	63 M	RCC	Nivolumab	No; 5 points	Acute phase reactants		Prednisone 40 mg; tocilizumab 162 mg q2 weeks
2	69 M	Melanoma	Ipilimumab/ nivolumab	Yes; 6 points		Sicca symptoms; ANA 1:1280, anti-SSA >8 IU	Prednisone 30 mg
3	79 M	Melanoma	Ipilimumab/ nivolumab	Yes; 7 points**		Sicca symptoms	Prednisone 20 mg
4	57 M	Melanoma	Pembrolizumab	No; 3 points	Acute phase reactants; RF 35 IU/ml; knee involvement		Prednisone 60 mg
5	60 M	Melanoma	Pembrolizumab	No; 3 points	RF 45 IU/ml; knee involvement		Prednisone 60 mg
6	66 M	Melanoma	Nivolumab	Yes; 5 points	Hand involvement		Prednisone 20 mg; MTX
7	69 F	RCC	Nivolumab	Yes; 5 points	Hand involvement		Prednisone 10 mg; tocilizumab 162 mg q2 weeks
8	66 M	RCC	Durvalumab; tremilimumab	Yes; 5 points	Hand involvement		Prednisone 20 mg
9	72 F	RCC	Avelumab	Yes; 4 points	Acute phase reactants; hand and knee involvement		Prednisone 20 mg

from: SOBI and Novartis, Speakers bureau: Novartis and SOBI, Jörg Henes: None declared, Jörn Kekow: None declared, Andrea Rubbert-Roth Consultant for: Chugai, Eli Lilly, Roche, and Sanofi

, Speakers bureau: AbbVie, Bristol-Myers Squibb, Chugai, Hexal/Novartis, Janssen, Eli Lilly, Merck Sharp & Dohme, Pfizer, Roche, and Sanofi, Hendrik Schulze-Koops Grant/research support from: Novartis, Pfizer, Roche, Consultant for: AbbVie, Actelion, Amgen, AstraZeneca, Biogen International, BMS, Celgene, Chugai, GSK, Hospira, Janssen-Cilag, Leo Pharmaceuticals, Lilly, MSD, Medac, Merck, Novartis, Pfizer, Hexal Sandoz, Sanofi, Roche, UCB, Paid instructor for: AbbVie, Actelion, Amgen, AstraZeneca, Biogen International, BMS, Celgene, Chugai, GSK, Hospira, Janssen-Cilag, Leo Pharmaceuticals, Lilly, MSD, Medac, Merck, Novartis, Pfizer, Hexal Sandoz, Sanofi, Roche, UCB, Eva Seipelt: None declared, Christof Specker: None declared, Eugen Feist Grant/research support from: Pfizer, Roche, Novartis, BMS, Sanofi, Lilly, Consultant for: Pfizer, Roche, Novartis, BMS, AbbVie, Celgene, Sanofi, MSD, Lilly, Speakers bureau: Pfizer, Roche, Novartis, BMS, AbbVie, Celgene, Sanofi, Lilly

DOI: 10.1136/annrheumdis-2019-eular.1179

THU0562

PREVALENCE, UNDERLYING FACTORS AND DAMAGE ASSOCIATED WITH CHRONIC PERSISTENT INFLAMMATION IN PATIENTS WITH FMF

Hakan Babaoglu¹, Berkan Armagan², Erdal Bodakci³, Timuçin Kaşifoğlu³, Hasan Satış¹, Nuh Atas¹, Alper Sarı², Nazife Sule Yasar Bilge³, Gözde Kübra Yardımcı², Reyhan Bilici Salman¹, Levent Kılıç², Mehmet Akif Öztürk¹, Berna Goker¹, Semir Haznedaroglu¹, Umut Kalyoncu², **abdurrahman tufan¹**, FICA. 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 characterized by recurrent, self-limiting attacks of fever, peritonitis, pleuritis, synovitis, myalgia, and erysipelas-like erythema (ELE). Attacks has a robust inflammatory response which completely normalize when attacks subside. However, substantial number of FMF patients possess chronic persistent inflammation even in between attacks. Clinical significance of persistent inflammation and its contribution to damage accrual are yet to be determined.

Objectives: We aimed to determine the prevalence and underlying factors of chronic persistent inflammation and its association with the domains of Auto-inflammatory disease damage index (ADDI) in a large cross-sectional multicenter cohort.

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. Demographic data, disease characteristic, attack features (ever presented) were meticulously questioned. Genotype data (if available) and laboratory features including inflammatory markers were recorded. FMF related damage scores was assessed by auto-inflammatory disease damage index (ADDI) which is recently validated. Patients were stratified according to their antecedent acute phase responses as having persistent inflammation (PI

group) or not, both groups compared in terms of disease characteristics, attack features, inflammatory comorbidities and damage domains.

Results: Study is comprised 970 adult patients (mean age 35.3±12.1, 61% female). 54 of them were excluded for their first inclusion. 15% of patients had persistent inflammation. PI group had significantly younger age of diagnosis, male dominance, homozygous M694V, more frequent attacks it the last year, more pleuritis, arthritis, myalgia and ELE and more severe disease according to ISSF than other patients (Table-1). Moreover, colchicine resistance and ADDI damage scores were remarkably higher in PI group.

In multi-variate analyzes colchicine resistance OR=2.71 [95%CI 1.38-5.35], ISSF OR=11.06 [95%CI 5.23-23.39], male gender OR= 2.38 [95%CI 1.51-3.76], homozygous M694V mutation OR= 2.31 [95%CI 1.41-3.78] and ELE OR= 1.72 [95%CI 1.04-2.857] found as independent predictors of persistent inflammation. PI group had more damage in multiple domains like joint restriction, musculoskeletal pain, proteinuria, amyloidosis, renal insufficiency and developmental delay. Same variables and persistent inflammation further analyzed in in terms of predicting the damage. Persistent inflammation found to be an independent predictor of proteinuria OR=2.02 [95%CI 1.05-3.90], amyloidosis OR=2.71 [95%CI, 1.34-5.48]), and renal insufficiency OR=4.36 [95%CI 1.84-10.32].

Conclusion: Persistent inflammation is relatively common in FMF patients particularly in those harboring M694V homozygous mutation and is associated with major complications of disease indicating poor prognosis.

Acknowledgement: None

Table 1 Patients demographics and disease characteristics |

	Persistent inflammation (n=142)	Others (-) (n=784)	p
Age, years	35(12.1)	35 (12.1)	0.09
Female	74 (52.1%)	492 (62.8%)	0.017
Disease onset <20 years	128 (90.1%)	585 (74.6%)	<0.001
Age at FMF diagnosis, years	22.6 (14.2)	25.1 (13.0)	0.041
M694V/M694V*	91 (71.1%)	240 (36.8)	<0.001
Fever	116 (81.7%)	655 (83.5%)	0.708
Peritonitis	125 (88%)	723 (92.2%)	0.158
Pleuritis	84 (59.2%)	351 (44.8%)	0.001
Arthritis	93 (65.5%)	301 (38.4%)	<0.001
Myalgia	57 (40.1%)	138 (17.6%)	<0.001
Erysipelas like erythema	68 (47.9%)	170 (21.7%)	<0.001
Attack frequency in the last year	8.37 (9.9)	3.78 (5.3)	<0.001
ISSF			
Mild Disease	9 (6.3%)	440 (56.1%)	
Intermediate Disease	91 (64.1%)	325 (41.5%)	<0.001
Severe disease	42 (29.6%)	19 (2.4)	
Colchicine resistance	38 (26.8%)	38 (4.8%)	<0.001
ADDI	2.2 (2.5)	0.9 (1.2)	<0.001
Joint restriction	13 (9.2%)	14 (1.8%)	<0.001
Musculoskeletal pain	84 (59.2%)	364 (46.4%)	0.006
Proteinuria	27 (19%)	37 (4.7%)	<0.001
Amyloidosis	28 (19.7%)	27 (3.4)	<0.001
Renal Insufficiency	20 (14.1)	15 (1.9%)	<0.001
Developmental delay	5 (3.5%)	9 (1.1%)	0.049
Solenomegaly	21 (14.8%)	34 (4.3%)	<0.001

ISSF: international severity scoring system for FMF. ADDI: autoinflammatory disease damage index
*n=14 missing in PI, n=132 missing in others

Disclosure of Interests: Hakan Babaoglu: None declared, Berkan Armagan: None declared, Erdal Bodakci: None declared, Timuçin Kaşifoğlu: None declared, Hasan Satış: None declared, Nuh Atas: None declared, Alper Sarı: None declared, Nazife Sule Yasar Bilge: None declared,