Therefore, more severe disease may be link with subclinical inflammation, amyloidosis and frequent, prolonged and widespread attacks. All these components may augment the frequency of non-disease related co-morbidities.

Methods: We enrolled 158 FMF patients who fulfilled modified *Tel-Hashomer* Diagnosis Criteria2. The patients dichotomized based upon disease severity (mild disease or severe disease). Patients with ISSF scores lower or equal to 2 were accepted to have mild disease. Then, we compared frequency of non-disease related co-morbidities between the groups. These co-morbidities are hypertension, hypothyroidism, hyperthyroidism cardiovascular diseases, coronary artery diseases, cerebrovascular diseases, chronic renal disease (non-FMF related), chronic obstructive pulmonary diseases, and diabetes mellitus. This study was approved by the Local Research Ethics Committee and carried out in compliance with the Helsinki Declaration. All the patients gave written informed consent. P-value lower than 0.05 was considered as statistically significant.

Results: Demographic features, disease duration, smoking history and body mass index (BMI) were similar between the groups. Frequency of co-morbidity in severe disease group was statistically higher than mild disease group ($p=0.02$). Most frequent co-morbidity was hypertension in both groups.

Table. Features of mild and severe FMF groups

	Mild (n=135)	Severe (n=23)	p
Gender (M/F)	47/88	11/12	0.23
Age	36.4±11.3	36.5±14.3	0.68
Smoking (%)	38 (28.1)	5 (21.7)	0.52
BMI (kg/m ²)	24.3±9.2	24.0±8.9	0.34
Disease duration (year)	7.7±11.3	8.6±14.3	0.09
Amyloidosis (%)	2 (1.4)	3 (13.0)	0.02
Exon 10 homozygote (%)	35 (25.9)	9 (39.1)	0.19
Colchicine dosage (mg/day)	1.2±0.4	1.4±0.5	0.02
ISSF scores	0.7 ±0.7	3.4±0.5	<0.001
Co-morbidity (%)	25 (18.5)	9 (39.1)	0.02

Conclusion: In our FMF patient cohort, we found that severity of the disease may be associated with higher frequency of co-morbidities. Therefore, clinicians should be aware of the high possibility of co-morbidities in patients with more severe FMF and addressed these co-morbidities timely and properly.

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SAT0542 UTILITY OF QUANTITATIVE ANALYSIS OF 18FDG-PET/CT IN IGG4-RELATED DISEASE

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Background: In IgG4-related diseases (IgG4-RD), usefulness of ¹⁸F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) imaging for detecting the organ involvement of IgG4-RD have been shown and, ¹⁸FDG PET/CT was more accurate and appeared to be more sensitive as compared to other imaging technics^{1,2}. However, until now, the studies regarding about quantitative analysis of PET/CT imaging in IgG4-RD were few. To avoid unnecessary biopsy and select suitable lesion for biopsy on multi-organ involvement disease such as IgG4-RD, the information which lesion is suspected as disease-involvement lesion in a non-invasive test is important.

Objectives: The purpose of this work is to evaluate the usefulness of ¹⁸FDG-PET/CT imaging in management of IgG4-RD using quantitative analysis of PET/CT imaging.

Methods: 21 patients with IgG4-RD, in whom PET/CT was undertaken at the time of diagnosis between December 2009 and July 2018, were enrolled. We retrospectively investigated the association between histological findings in which biopsy was performed for diagnosis of IgG4-RD and findings of PET/CT. ¹⁸FDG uptake was assessed in site of major organ involvement of IgG4-RD which could be differentiated from the normal uptake of background tissue with ¹⁸FDG-PET/

CT. For quantitative analysis, we measured the highest standardized uptake value (SUV) of the pixels within the region of interest (ROI) (SUV_{max}) and the average SUV within ROI (SUV_{mean}). We also measured SUV_{mean} of liver as reference tissue. Then, we calculated ratio between SUV_{mean} of ROI and SUV_{mean}/liver.

Results: The age at diagnosis was 64.5 ± 11.9 years, serum IgG4 was 743.8 ± 584.1 mg/dl, and biopsy was performed at 24 sites (Submandibular gland 10, prostate gland 4, pancreas 2, thyroid gland 1, lung 1, retroperitoneum 1, kidney 1). Histological findings were consistent with IgG4RD (positive) at 19 sites. Although SUV_{max} at the biopsy site was not correlated with the biopsy results, SUV_{mean} at the biopsy site were significantly higher in the biopsy-positive group (figure 1). As similar, SUV_{mean}/liver SUV_{mean} were also higher in the biopsy-positive group (2.17 vs 1.52, respectively $P<0.05$). To establish cut-off value of SUV_{mean} to consider biopsy, A receiver operating characteristics (ROC) curve was constructed. ROC curve analysis indicated SUV_{mean}=4.074 as cut-off value which discriminate IgG4-RD related lesion.

Conclusion: Our present study suggested that quantitative analysis of ¹⁸FDG-PET/CT imaging is useful for selecting the biopsy site in IgG4-related disease.

Figure 1. XXXXX

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SAT0543 PREVALENCE OF FABRY'S DISEASE IN MILD AND SEVERE FMF PATIENTS

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Background: Fabry disease (FD) is a rare metabolic disorder caused by the mutations in the α-galactosidase A (GLA) gene. FD patients present with heterogeneous clinical manifestations, which may overlap with systemic diseases including familial Mediterranean fever (FMF). Recurrent episodes of fever,

abdominal pain, and arthralgias can be observed in both disorders and this may lead to misdiagnosis.

Objectives: To investigate FD prevalence in mild and severe FMF patients.

Methods: A total of 66 FMF patients, according to the Tel-Hashomer criteria, were included in the study. Patients were grouped into mild (Group 1) and severe (Group 2) subsets according to the severity score. α -GLA enzyme activity and mutations in the GLA gene were performed. Demographic features, clinical findings, MEFV mutations and treatments were recorded.

Results: The clinical and demographic characteristics of the patients were given in Table 1. In severe form, 27 patients were using biological drug and 40.7% had amyloidosis. Symptoms related to FD including hypohidrosis, acroparesthesias, and painful neuropathies, were not different between the groups. Only one patient in group 1 had a low GLA enzyme activity (0.1 nmol/h/ml; Normal >2.5) which also had mutations in the GLA gene but MEFV mutation test was negative. (Table 2). This patient was a 39-year-old female with recurrent abdominal pain, distal extremity pain and the presence of fever during the attacks. She was heterozygous for R301Q. In detailed history, she reported mild acroparesthesias, hypohidrosis, and tinnitus.

Table 1. Demographic and clinical findings

	All patients n: 66	Group 1 n: 32	Group 2 n: 34	p-value
Age, median (min./max.)	34 (17/64)	27 (17/59)	36 (18/64)	0.192
Male, n (%)	36 (54.5)	14 (43.8)	22 (64.7)	0.137
Disease duration, median (min./max.)	20.5 (1/57)	12.5 (2/50)	25 (1/57)	0.006
Family history of FMF, n (%)	41 (62.1)	22 (68.8)	19 (57.6)	0.443
Alpha-galactosidase A (nmol/h/ml), median (min./max.)	5.9 (0.1/16)	5.6 (0.1/9.6)	6 (3.1/16)	0.330
Abdominal pain, n (%)	58 (87.9)	31 (96.9)	27 (79.4)	0.030
Fever, n (%)	54 (81.8)	25 (78.1)	29 (85.3)	0.532
Arthritis, n (%)	34 (51.5)	10 (31.3)	24 (70.6)	0.003
Pleuritis, n (%)	31 (47)	19 (59.4)	12 (35.3)	0.083
Painful neuropathy, n (%)	23 (34.8)	13 (40.6)	10 (29.4)	0.440
Acroparesthesias, n (%)	9 (13.6)	6 (18.8)	3 (8.8)	0.240
Angiokeratomas, n (%)	0 (0)	0 (0)	0 (0)	-
Cardiac abnormalities	1 (1.5)	1 (3.1)	0 (0)	0.485
Tinnitus, n (%)	4 (6.1)	3 (9.4)	1 (2.9)	0.274
Hearing loss, n (%)	2 (3)	2 (6.2)	0	0.086
Hypohidrosis, n (%)	2 (3)	1 (3.1)	1 (2.9)	0.965
Cornea verticillata, n (%)	0 (0)	0 (0)	0 (0)	-
Proteinuria, n (%)	13 (19.7)	2 (6.3)	11 (32.4)	0.012
Cochine dosing (mg/day), median (min./max.)	2 (1/3)	1 (1/2)	2 (1/3)	<0.001

Table 2. MEFV mutant alleles and GLA mutations

	All patients n: 66	Group 1 n: 32	Group 2 n: 34
Alpha -galactosidase A (GLA) gene mutations, n (%)	1 (1.5)	1 (3.1)	0 (0)
M694V mutations, n (%)	47 (35.6)	17 (26.5)	30 (44.1)
Non-M694V mutations, n (%)	36(27.2)	20 (31.2)	16 (23.5)

Conclusion: In this study, we showed the following: 1) the FD rate in the total FMF group was 1.5% (3.1% in group 1), 2) none of the patients in the severe FMF subset had abnormal enzyme activity or mutations related with FD, 3) symptoms related with FD such as hearing loss, hypohidrosis, acroparesthesias, and painful neuropathies also noted in FMF patients particularly in the milder group. Based on our results, FD should be considered in the differential diagnosis of FMF particularly in patients with atypical symptoms.

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SAT0544

USE OF BIOLOGICAL DMARDS IN PATIENTS WITH ADULT-ONSET STILL'S DISEASE: RESULTS FROM TURKBIO REGISTRY

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Background: Adult-onset Still's disease (AOSD) is a rare multisystemic inflammatory disorder, and is diagnosed by exclusion. AOSD is generally treated with nonsteroidal antiinflammatory drugs, corticosteroids, and conventional disease modifying antirheumatic drugs (cDMARDs). Biological disease modifying antirheumatic drug (bDMARD) therapy are recommended in AOSD patients who are

refractory to traditional therapy, and bDMARDs is becoming increasingly important in AOSD treatment.

Objectives: To evaluate the use of bDMARDs and drug survival in AOSD patients.

Methods: TURKBIO registry is the Turkish version of Danish DANBIO rheumatological database which has been established in 2011. All patients with AOSD who received biological agents registered in TURKBIO registry between dates of October 2011 and October 2019 were included in this study. The demographic data, response of therapy, frequency of using and switching biological agents were collected.

Results: As of October, 21 AOSD patients were recruited. Mean age of patients was 34.6±7.3 (min-max: 24-49) years, mean disease duration was 9.3±7.4 (min-max: 1-22) years, and 57.1% of patients was female. Mean duration from onset to start of bDMARDs was 7±6.1 (min-max: 0.5-21) years. It was observed that 13 patients (61.9%) received tocilizumab (TCZ), 6 patients (28.6%) received IL-1 inhibitors (5 anakinra and one canakinumab), 2 patients (9.5%) received certolizumab and one patient (4.8%) received etanercept as a first-line bDMARDs. The most frequently used biological agents in current treatment were as follows: 52.4% of patients received TCZ and 33.3% received IL-1 inhibitors (4 anakinra, 3 canakinumab), and the most frequently used concomitant drugs were methotrexate (47.6%) and hydroxychloroquine (14.3%). The switching rate was 33.3%, and in half of them the reason of switching was adverse events. The median drug survival for bDMARDs was 28.6 months (Table).

Table. Demographic, laboratory features and management of AOSD

(median;25-75)	n=21
Age (year)	34.7 (28.3-40.6)
Gender (Female) n(%)	12 (57.1)
Disease duration (year)	8 (2-17)
Duration from onset to start of bDMARDs (year)	6 (1.5-10)
ESR (on onset)	44 (21-66)
CRP (on onset)	65 (3.1-108)
Current bDMARDs n(%)	
Tocilizumab	11(52.4)
IL-1 inhibitors	7 (33.3)
Etanercept	1 (4.8)
Certolizumab	2 (9.5)
Concomitant cDMARD n(%)	
Methotrexate	10 (47.6)
Leflunomide	4 (19)
Sulfasalazine	1 (4.8)
Hydroxychloroquine	3 (14.3)
bDMARDs Survival (months)	28.6 (5.5-75)
Switching Rate n(%)	7 (33.3)
Adverse Event n(%)	3 (14.3)

Conclusion: This is the first evaluation of AOSD patients who used biological agents from TURKBIO registry. According our data, TCZ and anti-IL1 agents were the most frequent biological choices. The limitation of this study was the low number of the patients with AOSD who used biological agents.

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SATURDAY, 06 JUNE 2020

Diagnosics and imaging procedures

SAT0545

ULTRASONOGRAPHIC ENTHESEAL AND JOINT INVOLVEMENT IN PATIENTS WITH ACUTE ANTERIOR UVEITIS: A MONOCENTRIC CROSS-SECTIONAL STUDY.

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Background: Few studies have been investigating ultrasonographic (US) changes at peripheral entheses and joints in patients with anterior acute uveitis (AAU)¹ and none has been comparing non granulomatous uveitis (AAGU) with granulomatous uveitis (AAGU).

Objectives: To investigate prevalence of US entheses and joints abnormalities in a consecutive series of AAU patients and to compare US findings in AAGU HLA-B27 positive and HLA-B27 negative, with AAGU patients.