Abstract SAT0608 – Table 1. Results of univariate and multivariate analysis

Variables	Univariate Analysis			Multivariate Analysis		
	Odds Ratio	Confidence Interval	p value	Odds Ratio	Confidence Interval	p value
Favours Still's						
Fever at night	7,66	3,53–16,5	<0001			
Rash	10,08	4,80–21,2	<0001	31,3	3,6–271,9	0002
Arthritis	6,58	3,09–14,01	<0001			
Arthralgia	36	10,46–123,8	<0001	158,1	4,3–5755,8	0006
Sore throat	27,72	11,58–66,33	<0001	20,8	2,8–154,7	0003
Hemophagocytosis	4,79	0,96–23,89	0079			
Neutrophilia	10,87	4,90–24,13	<0001	18,4	2,6–132,3	0004
Ferritin $\geq$ 5 x UNL	4,88	2,34–10,16	<0001	132,8	7,1–2502,9	0001
LDH	7,12	2,35–21,59	<0001	6,2	0,76–50,9	0087
C3	3,20	1,47–7,00	0003			
Female	2,90	1,46–5,73	0002			
Favours FUO						
Pleuritis	2,04	0,68–6,12	0,19			
Fever peak number $\geq$ 3	3,66	1,16–11,52	0019	69	2,2–2114,4	0015
Lymphadenopathy	3,39	1,72–6,79	<0001			

UNL: Upper normal limit (for ferritin: 336 ng/ml)

Conclusions: Presence of arthralgia, hyperferritinemia, sore throat and neutrophilia strongly favour AOSD in patients presenting as FUO. This study demonstrates a clinician-friendly algorithm for the first time in current literature.

Disclosure of Interest: None declared

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SAT0609

VENOUS VESSEL WALL THICKNESS IN LOWER EXTREMITY IS INCREASED IN MALE BEHCET'S DISEASE PATIENTS WITH AND WITHOUT VASCULAR INVOLVEMENT

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Background: Vascular involvement is seen in up to 40% of the patients with Behçet's Disease (BD), especially in young males and is one of the major causes of mortality and morbidity. Lower extremity vein thrombosis due to vascular inflammation is the most frequent form of vascular involvement in BD. Recently, assessment of vessel wall thickness (VWT) and venous dilatation by US is suggested to be valuable in patients with vascular inflammation.

Objectives: In this study, we investigated whether vessel wall thickness or dilatation is present in young male BD patients prone to venous vascular disease.

Methods: Thirty male patients with BD without major organ involvement and 29 male patients with Vascular BD (VBD) followed in Marmara University Behçet's Clinics, 24 healthy male controls and 27 male patients with Ankylosing Spondylitis (AS) were included the study. Bilateral lower extremity venous doppler ultrasonography (US) was performed by an experienced radiologist blinded to cases. No patients except VBD were under immunosuppressive treatment. Bilateral common femoral vein (CFV) wall thickness and great/small saphenous vein dilatations were examined. Behçet Syndrome Activity Score (BSAS) was used for the general assessment of disease activity. In 10 patients, CFV wall thickness was measured by 2 different radiologist (RE, RA) in the same day to calculate "inter-observer reliability". Correlation between radiologists was good. ($r=0.765$, $p<0.001$).

Results: The mean disease duration was 9.1 ± 6 years in patients with BD. BSAS score was 24 ± 17 . All venous measurements were significantly higher in BD compared to AS and healthy controls ($p<0.001$ for all, table 1). When we compared mucocutaneous BD and VBD, all measurements of patients with VBD were higher than mucocutaneous BD. But only left CFV thickness and width of right great saphenous vein reached the statistical significance ($p<0.001$, $p=0.028$, respectively, figure 1). There were no correlations between BSAS, acute phase reactants and venous wall measurements.

Abstract SAT0609 – Table 1. Venous wall measurements of lower extremity in study groups.

	Behçet's Disease (n=59)	Ankylosing Spondylitis (n=27)	Healthy Controls (n=24)	P Value
Age, years	32.5 (23–42)	32 (20–37)	27.5 (25–42)	0.023
Body Mass Index (kg/m ²)	25.1 (18–33)	25 (18–32)	23.8 (20–29)	0.213
Right Common femoral VWT (mm)	0.8 (0.04–1.8)	0.3 (0.1–0.6)	0.25 (0.06–0.4)	<0.001
Left Common femoral VWT (mm)	0.8 (0.3–1.6)	0.3 (0.1–0.5)	0.2 (0.04–0.6)	<0.001
Right Great saphenous width (mm)	3.1 (0–6.4)	2.5 (1.1–3.5)	2.1 (1.3–3.5)	<0.001
Left Great saphenous width (mm)	3.1 (0–7.4)	2.6 (0.3–4.8)	2.4 (1.6–3.6)	<0.001
Right Small saphenous width (mm)	2.8 (0–5.3)	1.7 (1–3.1)	1.4 (0.9–3.7)	<0.001
Left Small saphenous width (mm)	2.7 (0–5.2)	1.8 (1.1–3.4)	1.6 (0.8–3.6)	<0.001

VWT: Venous wall thickness

Abstract SAT0609 – Figure 1. Venous Vessel Assessments of patients with Mucocutaneous Behçet and Vascular Behçet

Conclusions: In our study, an increased venous vessel wall thickness in lower extremity was shown in male BD patients with or without vascular involvement. As a similar change was not observed in controls, we think, increased VWT might be an early sign of venous inflammation in patients with BD rather than a result of non-specific systemic inflammation.

Disclosure of Interest: None declared

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SAT0610

A DECLINING TREND IN FREQUENCY OF SECONDARY AMYLOIDOSIS IN BEHCET'S SYNDROME

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Background: A decline in the frequency of AA amyloidosis secondary to rheumatoid arthritis and infectious diseases has been reported. This is probably due to more effective treatment strategies. We had previously reported that although amyloidosis occurs in less than 0.5% of BS patients, it is one of the leading causes of death.^{1–3} We had an impression that the frequency of amyloidosis is decreasing among our patients with BS.

Objectives: We aimed to determine the change in the frequency of AA amyloidosis over years in BS pts in addition to elaborating on clinical characteristics and outcomes.

Methods: We performed a chart review to identify all pts with amyloidosis in our BS centre since 1976. We noted demographic characteristics, BS manifestations, age at BS and amyloidosis diagnosis, treatment modalities of these pts. Our end-points were death and end stage renal disease (ESRD) requiring renal replacement therapy. The prevalence of amyloidosis was calculated separately for two periods (pts registered between 1976–2000 and 2000–2017)

Results: Among our 9410 BS pts, 27 (0.29%) had amyloidosis. We identified 24 pts with amyloidosis among the 3820 pts in the earlier cohort and 3 additional amyloidosis among the 5590 pts in the recent cohort. The frequency of AA amyloidosis had declined from 0.62% to 0.054% in the recent cohort. M/F ratio was 22/5 and mean age at BS diagnosis was 29.5±7.4 years. Twenty-two (82%) of the pts with amyloidosis had major organ involvement (vascular involvement in 15, eye involvement in 13 and neurologic involvement in 2). Five (18%) of 27 pts had only mucocutaneous involvement. AA amyloidosis was diagnosed after a mean duration of 9.8±6.7 years (mean age at amyloidosis: 39.3±9.3 years) and was confirmed with renal biopsy in 14 pts and rectal biopsy in 13. Eight pts had non-nephrotic range proteinuria at amyloidosis diagnosis. After amyloidosis diagnosis, 24 pts continued their previous immunosuppressives and colchicine. Two of these 24 were on anti-TNFs at AA diagnosis. Biologics were initiated in 3 pts who were most recently diagnosed to have amyloidosis, anti-TNFs in 2 and tocilizumab in 1. Fourteen (52%) pts had died after a median follow-up of 3 (IQR:1–8.75) years, 3 were lost to follow-up just after amyloidosis diagnosis and 10 (37%) are still alive after a median follow up of 16 (IQR:10–23) years. The reasons for death were infections in 5, related to ESRD in 5, subarachnoid haemorrhage, gastric adenocarcinoma, liver cirrhosis and iatrogenic bowel perforation in 1 patient each. 10 (71%) of these 14 pts had developed ESRD before their deaths. Overall, 15/27 pts developed ESRD after a median follow-up of 3.5 (IQR:1.25–6.5) years after amyloidosis diagnosis. 5 of them had renal transplantation, all but 1 are still alive after 3, 4, 6, and 12 years.

Conclusions: AA amyloidosis appears to be a rare, but fatal complication of BS. Around 50% of patients died after a median follow-up of 3 years after amyloidosis. This study showed a decreasing trend of AA amyloidosis due to BS similar to that observed in other inflammatory and infectious causes. The shorter follow-up duration may be contributing for the lower prevalence of AA amyloidosis in the recent cohort.

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SAT0611

CANAKINUMAB USE IN ADULT FAMILIAL MEDITERRANEAN FEVER PATIENTS: A LARGE SINGLE CENTRE EXPERIENCE

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Background: IL-1 blocking agents have been shown to be effective in the prevention of attacks in colchicine resistant FMF (crFMF) patients. Canakinumab is FDA approved long acting recombinant IL-1 receptor antagonist for use in crFMF patients which is available for off label use in Turkey. Herein, we aimed to share our real life single centre experience for use of canakinumab in adult crFMF patients.

Methods: Data was derived from Gazi FMF cohort which was established in year 2010. From that date patients with FMF who were diagnosed according to the Tel Hashomer criteria were registered. Co-morbidities, detailed attack characteristics, type, duration, severity, treatments, laboratory parameters and impact of FMF on their life in terms of quality of life and work productivity were recorded either by FMF diary or a mobile phone application (FMF AIDD free to download from App-Store and android market). A retrospective cohort analysis was made from records of patients who were treated with canakinumab.

Results: Eighteen adult crFMF patients (%61 female) treated with canakinumab were enrolled in this study. The median age was 31^{24–58} years and the median disease duration was 28^{16–40} years. All patients harbour homozygous or compound heterozygous exon 10 *MEFV* mutations. Treatment reasons for canakinumab were colchicine resistance (n=14) and amyloidosis (n=4). In three patients canakinumab was initiated directly, while in 15 it was switched from anakinra (seven was allergic to anakinra, one patient had significant leukopenia, in six fail to control attacks). The median duration of canakinumab use was 8 (min 1- max 22) months. In two patients canakinumab was used as 300 mg/monthly, and in

remaining as 150 mg/monthly. Pre- and post-canakinumab periods of patients were compared (Table). Patient reported attack severity (p<0.01), duration (p<0.01), frequency (p<0.01), C-reactive protein (CRP) (p<0.01) and erythrocyte sedimentation rates (p<0.01) were significantly improved while serum creatinine and alanine aminotransferase (ALT) levels remained same (p=0.2, p=0.35, respectively). Canakinumab achieved complete disease remission in 5 patients. Side effects requiring discontinuation of canakinumab were observed in none of patients.

Table. Comparison of attack characteristics of 18 Adult crFMF patients before and after canakinumab

	Colchicine	Colchicine +canakinumab	P
Attack severity, VAS	8,5 (5–10)	3,5 (0–10)	<0,01
Attack duration, hours	108 (48–144)	24 (0–96)	<0,01
Attack frequency*	6 (1–10)	1 (0–3)	<0,01
CRP, mg/L	35,8 (3,8–85,0)	4,2 (2,45–55)	<0,01
ESR, mm/h	38 (11–67)	12 (3–44)	<0,01
Serum creatinine	0,66 (0,40–3,00)	0,70 (0,43–3,00)	0,20
ALT	20 (10–55)	19 (9–70)	0,35
Treatment duration, months	204 (48–456)	8 (1–22)	

*Attack frequencies adjusted for 3 months intervals. VAS: visual analogue scale, CRP: C-reactive protein, ALT: alanine aminotransferase, ESR: erythrocyte sedimentation rate

Conclusions: Canakinumab is effective in the prevention of attacks with a favourable safety profile.

Disclosure of Interest: None declared

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SAT0612

THE EFFICACY OF CALCINEURIN INHIBITORS IN PATIENTS WITH ADULT-ONSET STILL'S DISEASE: SINGLE-CENTRE HISTORICAL COHORT STUDY

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Background: Adult-onset Still's disease (AOSD) is a systemic inflammatory disorder generally responsive to corticosteroid therapy, whereas refractory AOSD is often encountered that not controlled by corticosteroid monotherapy. In such cases, methotrexate and biologics including TNF- α , IL-1, or IL-6 inhibitors are used.¹ However, more treatment options are required for refractory AOSD patients who are intolerant to methotrexate or biologics. Calcineurin inhibitors downregulate T cell activation through inhibiting IL-2 transcription and signal transduction. Therefore, calcineurin inhibitors are reasonable therapeutic medication for AOSD since T cells and subsequently activated macrophages play a pathophysiological role in AOSD.² Nevertheless, only a few case series have indicated effects of calcineurin inhibitors in clinical practice.

Objectives: To evaluate the efficacy of calcineurin inhibitors in patients with AOSD.

Methods: This is a single-centre historical cohort study comprised of the consecutive patients with AOSD according to Yamaguchi classification criteria, who were attending our Rheumatology Department between January 2000 and December 2016. The primary endpoint was set as the time from the initiation of treatment to event defined as death of any causes, relapse of AOSD requiring an increase of corticosteroid dose, or serious adverse effects. Secondary endpoints were set as the minimum dose of corticosteroid, persistency rate of calcineurin inhibitors, and safety. Based on the recurrent event data analysis, these endpoints were evaluated for each event. We divided the events into two groups according to the treatment that included calcineurin inhibitors (CI+) or conventional therapy without calcineurin inhibitors (CI-), and compared them after adjustment using inverse probability of treatment weighting (IPTW) methods.

Results: Forty-two patients (31 female and 11 male) were enrolled in this study. Mean age was 41 year-old, and median follow-up period 38 months. Thirty-one events in 21 patients were treated with therapeutic regimen including calcineurin inhibitors (CI+, cyclosporine: 7, tacrolimus: 24), and 34 events in 25 patients were treated with the conventional therapy excluding calcineurin inhibitors (CI-). After adjustment, the CI +group had significantly longer event-free survival than the CI-group (figure 1). The weighted hazard ratio (HR) was 0.49 (95% CI, 0.25–0.93, p=0.03). In addition, the CI +group had lower doses of corticosteroid (3.5 vs 6.1 mg/day, p=0.03) when the events occurred. The persistency rate of