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 endpoints 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 nonnephrotic 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 heretozygos 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 ($p \leq 0.01$), duration ($p \leq 0.01$), frequency ($p \leq 0.01$), C-reactive protein (CRP) ($p \leq 0.01$) and erythrocyte sedimentation rates ($p \leq 0.01$) 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

	Colchisine	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 aminotranspherase, ESR: erythrocyte sedimentation rate

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

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