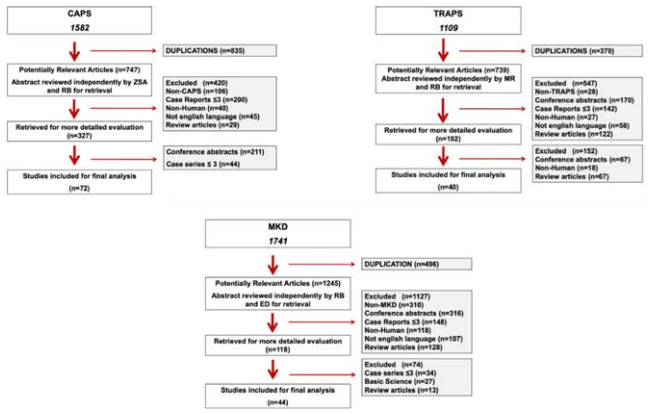

Figure 1. Flow-charts of systematic review for CAPS, TRAPS and MKD.

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FRI0483 SUBCLINICAL INFLAMMATION AND RELATED PARAMETERS IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF), which is more common in groups in the Mediterranean basin, is a monogenic autoinflammatory disease characterized by recurrent attacks of febrile peritonitis, pleuritis and arthritis.

Objectives: In this study, we aimed to investigate the clinical, demographic and genotypic features that may be associated with subclinical inflammation in FMF and to determine the related parameters with subclinical inflammation.

Methods: FMF patients according to the Tel-Hashomer criteria were included into the study. The demographic characteristics of the patients, duration of the disease, concomitant diseases, MEFV genotype mutation, colchicine use and resistance were collected. Acute-phase reactants such as white blood cell count, erythrocyte sedimentation rate, and C-reactive protein levels during the attacks and attack-free periods were noted. Subclinical inflammation was defined as the continuation of the acute phase response (CRP) between episodes. We divided study population into two groups as; patients with or without subclinical inflammation (Group 1 and Group 2, respectively) and these group were compared with the parameters described above. Patients with infectious disease (viral or bacterial) in the past two months were excluded from the study

Results: Eigthy patients (72.5% female) with mean age 37.1 SD 11.2 years were recruited into the study. Twenty-three (28.7%) patients were determined with subclinical inflammation. Group 1 had significantly higher rate of concomitant rheumatic disease(i.e spondyloarthropathy), erythrocyte sedimentation rate and MEFV homozygous mutation compared with Group 2 ($p<0.05$, for each). Disease duration, months PRASS score, FMF quality of life, age at onset of symptoms, family history of FMF, response to colchicine, attack time, attack in the last 6, delay in diagnosis parameters were not significantly different between groups ($p> 0.05$).

Conclusion: FMF patients whose elevated erythrocyte sedimentation rate and MEFV homozygous mutation should be closely monitored for subclinical inflammation even during attack-free periods. Concomitant disease should be detected in FMF patients with subclinical inflammation.

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Table 1: Demographic and clinical features of the patients with familial Mediterranean fever

	Without Subclinical inflamation n= (57)	With Subclinical inflamation n= (23)	P value
Age (years; mean SD)	37,78 SD 13,22	36,82 SD 10,49	0.987
Female, gender, n (%)	45(%78)	13(%56)	0.055
Disease duration (month; mean SD)	255,3 SD 195,1	180,2 SD 121,1	0.191
PRASS score (mean SD)	6,08 SD 2,15	5,36 SD 1,59	0.147
BMI, kg/m^2	26,12 SD 4,8	32,13 SD 28,48	0.629
Current smoking status (%)	17(%29)	3(%13)	0.067
Age at onset of symptoms (month; mean SD)	15,69 SD 9,41	17,28 SD 10,34	0.54
Family history of FMF(%)	37(%64)	18(%78)	0.295
Response to colchicine(%)	6(%10)	4(%17)	0.462
Attack time (day; mean SD)	1,9 SD 1,1	2,26 SD 1,4	0,523
Attack in the last 6 months (mean SD)	2,79 SD 3,1	4,56 SD 5,5	0,184
FMF quality of life (mean SD)	31,5 SD 13,6	25.7 SD 16,4	0,130
Delay in diagnosis(month; mean SD)	12,29 SD 10,9	14,3 SD 14,9	0,840

Disclosure of Interests: None declared

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FRI0484 SAFETY PROFILE, CLINICAL AND RADIOLOGICAL EFFICACY OF ANAKINRA, TARGETED AND COMBINED TREATMENT IN ERDHEIM-CHESTER DISEASE

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Background: Erdheim-Chester disease (ECD) is a rare non-Langerhans histiocytosis. Combined treatment with anakinra (ANK) and targeted MAPK-inhibiting therapies (vemurafenib – VMF - or cobimetinib - CBM) has been recently used to treat severe cases of ECD.

Objectives: To evaluate the safety and the clinical and radiological efficacy of ANK, targeted and combined treatments in ECD patients in a real-world setting.

Table 1. Disease characteristics and therapy-related adverse reactions of Erdheim-Chester patients treated with vemurafenib, cobimetinib and/or anakinra.

	Vemurafenib (n=19)	Cobimetinib (n=10)	Anakinra (n=12)
Clinical Manifestations			
Cardiovascular	79%	70%	75%
Retroperitoneal	84%	60%	42%
Pleuropulmonary	63%	80%	42%
Neurological and/or orbital	90%	90%	92%
Adverse reactions	74%	60%	25%
Renal	26%	10%	0%
Cutaneous	26%	30%	17%
Systemic Inflammation	11%	10%	0%
Cardiovascular	11%	10%	0%
Gastrointestinal	0%	30%	0%
Haematological	0%	10%	0%
Herpes Zoster	0%	0%	8%