Objectives: To assess the frequency of depression and anxiety in patients with Familial Mediterranean Fever (FMF)

Methods: In this study, 77 FMF patients aged 18 and over who were followed up in Sakarya University Education and Research Hospital, Department of Rheumatology, and 78 healthy volunteers aged 18 and over as the control group. Beck depression scale and Beck anxiety scale were used to depression and anxiety, respectively. Beck's depression scale was evaluated as 9 and below normal, 10-16 mild depression, 17-29 moderate depression, 30-63 severe depression. Beck anxiety scale was evaluated as 0-8 normal, 8-15 mild anxiety, 16-25 moderate anxiety, 26 and above severe anxiety. FMF disease severity was determined by Prasad scoring.

Results: The study group, comprised 77 diagnosed with FMF with a mean age of 37.18 and a control group comprised of 78 healthy controls (C) with a mean age of 35.32 ($p=0.058$). In study group (P) %63.6, control group (C) %53.8 as female. %36.4 of the study group (C), %46.2 of the control group are male. ($p=0.216$). The prevalence of depression was significantly higher in FMF patients compared to the control group (in order P:C: normal %24.7; %47.4, mild depression: %40.3; %26.9, moderate depression %26; %19.2, severe depression %11.7; %6.4 $p<0.015$). Similarly in depression results; the prevalence of anxiety was significantly higher in FMF patients compared to the control group (in order P:C normal %23.4; %57.7, mild anxiety %26; %20.5, moderate anxiety %26; %15.4, severe anxiety %24.4; %6.4 $p<0.001$). Depression status was not correlated with FMF disease severity ($p=0.645$). A correlation was found between FMF severity and anxiety which it is which was found statistically significant ($p=0.005$). There was no relationship between erythrocyte sedimentation rate and C-reactive protein with depression and anxiety.

Conclusion: Both anxiety and depression frequency are increased in FMF patients compared to healthy controls.

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AB1035

INTESTINAL MICROBIOTA COMPOSITION OF ADULT PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER AND HEALTHY CONTROLS (THE RHEUMA-BIOTA STUDY)

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Background: Although Familial Mediterranean Fever (FMF) is a monogenic disease, microbiota composition may play role in the pathogenesis or phenotypic expression.

Objectives: We aim to evaluate the intestinal microbiota composition in patients with FMF and to compare with healthy controls.

Methods: In this prospective cohort study, a group of 10 adult patients with FMF and 10 age-appropriate healthy controls, for which there was strict inclusion/exclusion, were enrolled. Fecal samples were stored at -80°C until DNA extraction. A region of the 16S rRNA gene (V3-V4) was selected and sequencing was performed on the Illumina MiSeq platform at the Sequencing and Bioinformatics Service of FISABIO foundation.

Results: Alpha and beta diversity tests were similar between FMF and control groups except that Chao1 index. Chao1 index was modestly decreased in FMF group comparing the healthy controls ($p<0.05$). Our results showed differences in the intestinal microbiota composition of patients with FMF, with a higher

abundance of *Eggerthella*, at genus level. At species level, *Eggerthella sinensis* and *Eggerthella lenta* were more abundant in patients with FMF.

Conclusion: *Eggerthella lenta* was previously shown to be higher in type II diabetes, multiple sclerosis, rheumatoid arthritis and some disseminated infections. In this study we firstly showed abundance of *Eggerthella* in patients with FMF, especially in *E. sinensis* and *E. lenta*; in addition to. Whether any of observed associations are causal, or the direction of causality is unclear yet and further studies with patients with FMF at the first diagnosis might clarify this issue.

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AB1036

CLINICAL MANIFESTATIONS, CLINICAL COURSE, AND OUTCOMES OF IMMUNOGLOBULIN G4 RELATED DISEASE

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Background: Immunoglobulin G4 related disease (IgG4-RD) is an uncommon chronic systemic autoimmune disease, pathologically characterized by lymphoplasma cell, IgG4 plasma cell or storiform fibrosis infiltration with elevated serum IgG4 level. IgG4-RD is a new disease and not widely recognized.

Objectives: The aim of this study was to describe clinical manifestations and outcomes of IgG4-RD in Thai patients

Methods: This multicenter retrospective cohort study included patients who aged ≥ 18 years and were diagnosed with IgG4-RD according to 2011 comprehensive or consensus diagnostic criteria, between 2000 and 2019 in four academic centers in Thailand. Baseline characteristic, laboratory and pathologic findings, treatments, and outcomes were systematically reviewed.

Results: Of the 110 patients included, 71% were male with mean age (SD) of 59.6 (13.3) years and median disease duration (IQR) of 28.8 (14.6-53.5) months. Single organ involvement was observed in 60 patients (54.5%). The most common presenting organ involvement was the orbit (29%), followed by the salivary glands (19%), lacrimal glands (18%), bile duct (16%), and pancreas (11%). The most frequently affected organs were the orbits (34%), followed by the salivary glands (26%), lacrimal glands (20%), bile duct (19%), and lymph nodes (19%). Ninety-six percent (96%) had IgG4 level of more than 135 mg/dl at presentation. Most patients (92%) were treated with corticosteroid (CS) alone or in combination with immunosuppressive agents. Azathioprine (47%) and methotrexate (11%) were the most commonly used immunosuppressive agents. Additionally, 20% required surgery, and 6.4% underwent stent insertion. One-fourth (26%) were in remission with successfully CS tapering, while 37%, and 29% had complete, and partial response. Nevertheless, 22% relapse with median time to relapse (IQR) of 22.2 (12.8-41.1) months. Relapse was common in patients with orbital ($p = 0.001$) and lung ($p = 0.007$) involvement, and patients with longer disease duration (median 44.1 and 23.1 months, $P=0.001$), while serum IgG4 level was insignificantly higher in relapse group (median 1,085 vs. 850 mg/dL, $p=0.28$).

Conclusion: IgG4-RD is a chronic systemic autoimmune disease with diverse manifestations, response to treatment, and outcomes. Most patients responded well to CS and immunosuppressive agents with notable relapse rate, while minority required surgery or mechanical intervention.

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AB1037

CANAKINUMAB FOR TREATMENT OF ADULT ONSET STILL'S DISEASE-RESULTS OF THE 24 WEEKS TREATMENT AND BEYOND: A MULTI-CENTRE, PLACEBO-CONTROLLED STUDY (CONSIDER)

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Background: Inhibition of interleukin-1 (IL-1) represents a promising treatment option in adult-onset Still's disease (AOSD). Canakinumab is approved for treatment of systemic juvenile idiopathic arthritis and has a marked impact on systemic as well as articular activity of the disease.

Objectives: To investigate the efficacy and safety of canakinumab in patients with AOSD and active joint involvement by means of a multi-centre, double-blinded, randomized, placebo controlled trial over a period of 24 weeks with the option of a long-term extension.

Methods: Patients with AOSD and active joint involvement (tender and swollen joint count ≥ 4 each) were stratified by pre-treatment status with biologic DMARDs to canakinumab (4 mg/kg, maximum 300 mg s.c. q4w) or placebo. After approval of canakinumab for AOSD by the European Medicines Agency, recruitment was stopped prematurely with enrollment of 35 out of 68 planned patients. The primary endpoint was the proportion of patients with a clinically-relevant reduction in disease activity at week 12 as determined by the change in disease activity score (Δ DAS28 > 1.2).

Results: At enrollment, patients had high active disease with a mean DAS28(ESR) of 5.4 in the canakinumab group (n=18, [CI 43.1; 85.2]) and 5.3 in the placebo group (n=17, [CI 20.1; 65.0]). In the intention-to-treat analysis, 12 (67%) canakinumab and 7 (41%) placebo patients fulfilled the primary outcome criterion (p=0.18). Figure 2 shows the DAS28-ESR disease activity by treatment groups and visits with imputation. In the per-protocol analysis, significantly higher ACR30 (61% vs. 33% p=0.033), ACR50 (50% vs. 6.7% p= 0.009) and ACR70 (28% vs. 0% p=0.049) response rates were observed in the canakinumab group compared with placebo (Figure 1). Two patients in the canakinumab group experienced an SAE.

Figure 1. Response rates (PP)

Figure 2. DAS28-ESR disease activity by treatment groups and visits with imputation

Conclusion: Although the study was terminated prematurely and the primary endpoint was not achieved, treatment with canakinumab led to an improvement of several outcome measures in AOSD. The overall safety findings were consistent with the known profile of canakinumab. Thus, our data support indication for IL-1 inhibition with canakinumab in AOSD.

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P-values are shown above each pair of bars; P-values in red are significant ACR, American College of Rheumatology; CRP, C-reactive protein; DAS, disease activity score; EULAR, European League Against Rheumatism; PP, per-protocol

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AB1038 INFLAMMATORY ORBITAL DISEASES: THE EXPERIENCE OF A TERTIARY RHEUMATOLOGY CENTRE

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Background: Inflammatory lesions of orbital disease encompass a wide spectrum of clinical entities including rheumatic disorders.

Objectives: To describe our experiences in adult patients who applied to a tertiary rheumatology center due to orbital disease.

Methods: This is a retrospective descriptive study and data were extracted from patient charts. We described the clinical, laboratory, radiologic, histopathological presentations and final diagnoses of patients with inflammatory orbital disease who applied to our rheumatology clinic between January 2014 and December 2019.

Results: Thirty-eight patients (Female: 63.2%) were identified; median age at onset of orbital symptoms was 44.5 (min.-max 5-72) years. Swelling (57.9%) and orbital pain (47.4%) were the most common symptoms, followed by erythema (13.2%), vision loss (13.2%), proptosis (7.9%) and diplopia (7.9%). Table summarizes the demographic and clinical characteristics of the patients. Imaging (MRG) was performed in all patients and 63.2% had an orbital biopsy. Orbital imaging revealed extraocular muscles (71.1%), lacrimal glands (50.0%) and optic nerve (42.1%) involvement. Of patients 34.2% had bilateral and 18.4% had retroorbital involvement. The final diagnoses of patients were: IgG4-related disease (34.2%, n=13), idiopathic orbital inflammatory pseudotumor (36.8%, n=14), granulomatosis with polyangiitis (18.4%, n=7), Sjogren's syndrome (n=1), relapsing polychondritis (n=1), thyroid-associated orbitopathy (n=1) and fungal granulomatous angitis (n=1).

Conclusion: Inflammatory lesions of the orbit are rare and the diagnosis may be challenging. Differential diagnosis is based on clinical, laboratory, radiologic and histopathologic findings. Although IgG4-related disease is a relatively new diagnostic cause for orbital inflammation, it should be considered more in differential diagnosis.