Table 2. Distribution of patients according to organ involvement.

Organ involvement	Number (%)
Primary sclerosing cholangitis	1 (1.4)
Musculoskeletal	1(1.4)
Submandibular gland	1(1.4)
Central nervous system	1 (1.4)
Multisystem involvement	9 (12.86%)

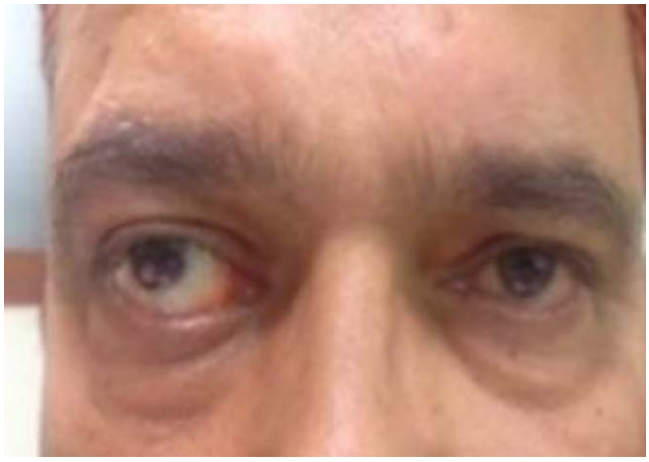

Figure 1. Right eye proptosis

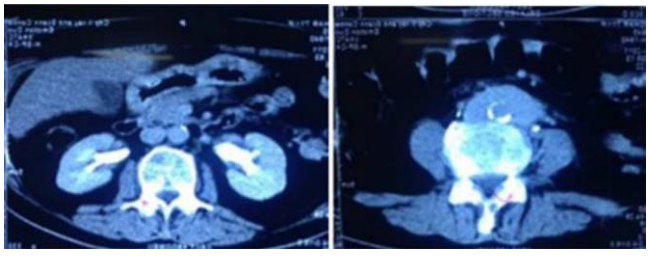

Figure 2. CT abdomen showing hydronephrosis due to retroperitoneal fibrosis

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AB1031 DYSKERATOTIC CELLS IN PERSISTENT PRURITIC SKIN LESIONS AS A PROGNOSTIC FACTOR IN ADULT-ONSET STILL'S DISEASE.

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Background: Adult-onset Still's disease (AOSD), a systemic inflammatory disorder, is characterized by high fever, evanescent rash, arthritis, and hyperferritinaemia. AOSD is also reported to be associated with other skin lesions, including persistent pruritic papules and plaques.
Objectives: This study aimed to assess the significance of dyskeratotic skin lesions in Japanese AOSD patients.
Methods: We retrospectively assessed the histology of persistent pruritic skin lesions and evanescent rashes and the relationship between dyskeratotic cells, serum markers, and outcomes in 20 Japanese AOSD patients, comparing AOSD histology with that of dermatomyositis (DM), drug eruptions, and graft-versus-host disease (GVHD).
Results: Persistent pruritic lesions were characterized by scattered single keratinocytes with an apoptotic appearance confined to the upper layer of the epidermis and horny layer without inflammatory infiltrate. In contrast to AOSD, the histology of DM, drug eruption, and GVHD demonstrated dyskeratotic cells in all layers of the epidermis with inflammatory infiltrate. AOSD with evanescent rash showed no dyskeratotic cells. The dyskeratotic cells in pruritic AOSD lesions stained positive for ssDNA and terminal deoxynucleotidyl transferase-mediated dUTP nick end-labeling, indicating apoptosis. Serum IL-18 was significantly higher in AOSD patients with dyskeratotic cells than those without, and generally required higher doses of glucocorticoids, immunosuppressants, and biologic agents. Two of ten AOSD patients with dyskeratotic cells died from haemophagocytic lymphohistiocytosis.
Conclusion: Persistent pruritic AOSD skin lesions are characterized by dyskeratotic cells with apoptotic features, involving the upper layers of the epidermis. There may be a link to elevated IL-18. This dyskeratosis may be a negative prognostic indicator.
Disclosure of Interests: None declared
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AB1032 CONTRIBUTION OF BONE BIOPSY DURING REVELATORY BONE METASTASES

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Background: Bone metastases (BM) are tumor cells that originate in a primary malignant tumor and are localized remotely in bone tissue. They more or less faithfully reproduce the morphological and biological characteristics of the primary tumor. Histological analysisis essential to confirm the diagnosis of BM and to identify the primary tumor if possible and sometimes to help in the selection of treatment.
Objectives: The aim of this work is to study the contribution of bone biopsy during revealing BM in diagnostic strategy and therapeutic decision.
Methods: We retrospectively studied the files of 105 patients hospitalized in a Rheumatology department of for BM revealing from January 2000 until December 2015. For each patient we collected epidemioclinical and anatomopathological data to arrive at the diagnosis of primary neoplasm and histological type.
Results: The patients were divided into 86 men (81.9%) and 19 women (18.1%) with a sex ratio (M / F) of 4.52. The average age of our patients was 64.91 ± 13.29 years. Pain was the most frequent reason for consultation found in 97.1%. This pain was either of bone site (61.9%) or of radicular topography (41.9%). Bone swelling or a pathological fracture revealed BM in 4.8% and 8.6% of the cases, respectively. The onset of neurological damage was noted in 13.3% of the cases.
Histologically, the bone biopsy performed in 64 patients made it possible to specify the histological type (carcinoma, adenocarcinoma) in 64% of the cases and to lead to primary cancer in 57.8%. A non-radio-guided percutaneous bone biopsy was performed in 44 patients (68.75%) including 41 osteo-medullary biopsy in iliac crest (BOM) and 3 in the sternum, a bone biopsy directed under scanner in 16 cases (25%) and a surgical bone biopsy in 4 cases.
The BOM was positive in 21 cases (51.2%) showing a poorly or moderately differentiated adenocarcinoma or carcinoma. It allowed referral to a primitive in 20 cases: a prostatic origin in 11 cases, a pulmonary origin in 5 cases, a digestive origin in 2 cases, a mammary origin in one case and a neuroblastoma in one case.
Conclusion: Thanks to improved sampling and immunohistochemistry techniques, the precise histological type and location of the primary tumor could be identified, there by improving the quality of care for patients with increased life expectancy.
Disclosure of Interests: None declared
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AB1034 DEPRESSION AND ANXIETY IN FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is a hereditary autoinflammatory disease characterized by recurrent attacks of fever, peritonitis, pleuritis, arthritis, and skin eruption (1). It is shown by studies that chronic diseases like diabetes mellitus, chronic heart disease, hypertension which other than inflammatory – rheumatologic disease increase depression and anxiety (2). There are a few studies evaluating depression and anxiety in FMF patients, and these results are conflicting (3,4).

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 thecontrol group. Beck depression scale and Beck anxiety scale were used to depression and anxiety, respectively. Beck'sdepression 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 Pras scoring.
Results: The study group, comprised 77 diagnosed with FMF with a meanage of 37.18 and a control group comprised of 78 healthy controls (C) with a meanage of 35.32 (p=0,058). İn studygroup (P) %63.6, control group (C) %53.8 as female. %36.4 of thestudy 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 -80ºC 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.
Disclosure of Interests: None declared
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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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Disclosure of Interests: None declared
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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)

C. Kedor[1], J. Listing[2], J. Zernicke[1], A. Weiß[2], F. Behrens[3], N. Blank[4], J. Henes[5], J. Kekow[6], A. Rubbert-Roth[7], H. Schulze-Koops[8], E. Seipelt[9], C. Specker[10],