Objectives: To describe cross-sectional data of anti-TNF trough levels and ADA, combined with decision effects, in children with JIA.

Methods: Patients' records in children with JIA using etanercept, adalimumab or infliximab were retrospectively checked for measurements of anti-TNF trough levels and ADA. Anti-TNF trough concentrations and ADA were measured using an enzyme-linked immunosorbent assay (ELISA) and antigen-binding test. Data on age, sex, JIA subtype, reason for testing and the decision effect of trough level or presence of ADA on the current therapy were collected.

Results: Eighty-one anti-TNF trough levels were measured in 45 children with JIA. A wide variety of anti-TNF trough levels was found. Therapeutic drug concentrations, according to adult ranges in RA and IBD,^{4,5,6} were found in 11 (58%) patients on etanercept (n=19), 2 (14%) on adalimumab (n=14) and 8 (17%) on infliximab (n=48). Four patients on adalimumab and one patient on infliximab showed ADA. All of these five patients had non-detectable drug trough levels. Reasons for testing trough level and/or presence of ADA were loss of response (20%), partial or no response (40%), measurement after dosage increase (2%), remission (17%), uveitis flare (9%) and allergic reaction (11%). Treatment decisions were influenced by trough levels in 70/81 (86.4%) of measurements and in 5/5 (100%) of patients with ADA.

Abstract THU0557 – Figure 1. Dose vs Concentration (etanercept, adalimumab, infliximab)

Conclusions: Measuring anti-TNF trough levels and ADA was a valuable tool in making personalised treatment decisions in JIA. Treatment changes included dose/frequency increase, or stopping and switching treatment in the presence of ADA combined with undetectable drug levels. More data are needed to access optimal therapeutic drug levels in anti-TNF treatment in JIA and to implement this strategy more widely.

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THU0558

INCREASED FREQUENCY OF FEBRILE SEIZURES IN TWO PERIODIC FEVER SYNDROMES: FAMILIAL MEDITERRANEAN FEVER AND PFAPA SYNDROME

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Background: Fever and the fear of febrile seizure risk in parents are a common reason that lead parents to seek care in childhood emergency departments. The most frequently seen periodic fever syndromes in Turkey are periodic fever, aphthous stomatitis, pharyngitis, cervical adenitis (PFAPA) syndrome and familial Mediterranean fever (FMF). There is lack of data regarding the frequency of febrile seizures in periodic fever syndromes.

Objectives: To document the frequency of febrile seizure in patients with Familial Mediterranean Fever and PFAPA syndrome.

Methods: Patients with FMF and PFAPA, who were diagnosed according to Turkish paediatric FMF diagnostic criteria and Marshall criteria, were enrolled to the study. Past medical history of all subjects from postnatal six months to 6 years of age were assessed in terms of febrile seizure. Clinical, demographic and laboratory data of both patient groups were obtained from patient files. The frequency of febrile seizure in both disease groups was compared with the prevalence in healthy children of a previous Turkish study.

Results: A total of 417 FMF and 157 PFAPA subjects were recruited to the study with a female frequency of 49.9% and 42.8%, respectively. The mean age of the FMF and PFAPA patients at study time was 12.4±4.5 years and 5.3±2.1 years, respectively. The mean age at disease onset was 5.1±3.8 (IQR: 2–7) years in FMF group, whereas it was 21±16.5 months (IQR: 10–30) in PFAPA group. The frequency of febrile seizure in FMF and PFAPA syndrome was similar (8.4% versus 8.6%, p>0.05). Among the subjects with febrile seizure, 42.9% (n=15) in FMF group and 30.8% (n=4) in PFAPA group underwent electroencephalography. The EEG result of all subjects were reported to be normal. While 8 subjects with febrile seizure (22.9%) in FMF group required anticonvulsant therapy, 2 patients (15.4%) in PFAPA group used anticonvulsant treatment. Among the PFAPA subjects with febrile seizure (n=13), 53.9% (n=7) underwent tonsillectomy. Thereafter, 85.7% (n=6) of the patients with tonsillectomy never experienced a febrile seizure.

Both the FMF and PFAPA syndrome group were found to have a higher frequency of febrile seizure compared with the healthy subjects of the previous Turkish study (FMF versus healthy children: 8.4% versus 3.2%) (PFAPA syndrome versus healthy children: 8.6% versus 3.2%); and this was statistically significant (p<0.001). However, the frequency of febrile seizure in FMF did not differ from the patients with PFAPA syndrome (8.4% versus 8.6%, p>0.05).

Conclusions: Compared to the healthy children, the frequency of febrile seizure in FMF and PFAPA syndrome was significantly increased. The recurrent fever, which is the main shared manifestation of these two diseases, is possibly a trigger for febrile seizure. This increased frequency of febrile seizure in both periodic syndrome seems to be a result of recurrent fever, other than a neurologic involvement of the FMF itself.

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THU0559

THE PERFORMANCE OF THE NEWLY PROPOSED EULAR/ACR CLASSIFICATION CRITERIA IN JUVENILE-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS-A PRELIMINARY STUDY

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Background: The sensitivity and specificity of the ACR-1997 and SLICC-2012 classification criteria in juvenile-onset systemic lupus erythematosus (SLE) are already studied. In previous reports, the main limitations of the ACR-1997 and

SLICC-2012 were low sensitivity and low specificity, respectively. To avoid misclassifications, a new set of classification criteria have been developed by the collaboration of the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) and the draft was presented at the 2017 ACR/ARHP Annual Meeting in San Diego, California. After application on 500 SLE patients and 500 controls, the sensitivity and specificity were found as 98% and 97%, respectively.

Objectives: To compare the sensitivity of the new EULAR/ACR criteria with those of the 1997 American College of Rheumatology (ACR) criteria and 2012 Systemic Lupus International Collaborating Clinics criteria in juvenile-onset SLE patients.

Methods: Patients initially were evaluated by ACR-1997, SLICC-2012 and EULAR/ACR classification criteria at baseline, when the diagnosis for the first time had been established by an expert paediatric rheumatologist (OK). All data were obtained from patient records. The diagnostic sensitivity of the three sets of classification criteria were further tested within 1 year of diagnosis and at last patient visit, longitudinally.

Results: A total of 104 juvenile-onset SLE patients were enrolled for the sensitivity performance of classification criteria at diagnosis. Since the follow-up period was less than 1 year, 12 subjects excluded after baseline evaluation. Finally, 92 subjects were eligible for sensitivity evaluation within 1 year of diagnosis and at last visit. The median age at diagnosis of clinician was 13.0 years (range 3.1–17.9 years, interquartile range (IQR) 11.1–16.5 years) with a median disease duration of 5.0 years (IQR 3.0–8.0 years). The female-to-male ratio was 4.7:1. The newly developed EULAR/ACR classification criteria were more sensitive than SLICC-2012 and ACR-1997 at diagnosis (93.3% versus 91.3% and 85.6%, respectively), and at first year (95.7% versus 94.6% and 90.2%, respectively ($p>0.05$)). At last visit the sensitivity of the new set of criteria and SLICC-2012 were same (97.8%), but higher compared to ACR-1997 criteria (95.7%).

Conclusions: Juvenile-onset systemic lupus erythematosus was classified by the newly proposed EULAR/ACR criteria with higher sensitivity compared with SLICC-2012 and ACR-1997 at disease onset and within one year of diagnosis. However, last visit assessment demonstrated equal sensitivity between new EULAR/ACR criteria and SLICC-2012. Although the difference was not significant, the new set of criteria seem to be capable of recruiting more children with juvenile SLE to clinical trials.

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THU0560 CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS: FOUR TERTIARY SPANISH HOSPITALS EXPERIENCE. A MULTICENTRIC STUDY

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Background: Chronic recurrent multifocal osteomyelitis (CRMO) is a rare autoimmune inflammatory polygenic bone disease characterised by aseptic bone inflammation in paediatric population. Its management, clinical, radiological findings and treatment have not yet been standardised.

Objectives: Retrospective, descriptive multicentric study of patients diagnosed of CRMO in four tertiary level hospitals' paediatric rheumatology section. There were 16 patients included. The clinical, radiological characteristics were analysed as well as response to treatment options.

Results: The median age at diagnosis was 10.5 years, female: male ratio 62.5:37.5%. The delay in the diagnosis had a median of 4.5 months, being less than one year in 11 patients. Bone pain was the first symptom in 100% of the patients accompanied by fever in 25% of them. A single patient presented perilesional arthritis. A slight-moderate increase on acute phase reactants was observed at the debut of the disease: median ESR 47.5 mm/h.

The median number of locations at debut was 2.5 (range 1–14), with multifocal involvement in 75%. The most frequent location was tibia (56%), followed by pelvis (44%) and vertebrae (31.25%). Other locations less frequent were: carpus (12.5%), femur (12.5%) mandible (6%) and sternum (6%).

Biopsy was performed in 14/16 patients and bone scintigraphy with Tc99 in 12/16 patients, with pathological uptake observed in 91.6% of cases. MRI was the radiological test of suspected diagnosis in 15/16 patients.

NSAIDs were the initial treatment. 5 patients received different antibiotic therapy regimens, without clinical or radiological improvement. 56.25% of patients required other treatments. Systemic corticosteroids were used in 12.5% of patients and bisphosphonates in 43.75% (100% of patients with axial involvement). After 6 months of treatment with bisphosphonates, 57.14% had complete remission, 28.57% partial remission and 14.28% worsening. 12.5% of the patients had a torpid evolution, receiving sequential therapies with multiple synthetic or biological DMARDs (Anakinra, Canakinumab, Etanercept), and another 12.5% required surgery.

Conclusions: The diagnosis of CRMO is a challenge in the absence of pathognomonic features which leads to delay in diagnosis and the initiation of treatment. In our centres the bisphosphonates were the treatment strategy used in patients with spinal involvement with 85.67% response at 6 months.

Disclosure of Interest: None declared

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THU0561 ANAKINRA FIRST- OR SECOND-LINE IN SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS PATIENTS

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Background: Treatment of systemic juvenile idiopathic arthritis (sJIA) still remains a challenge.

Objectives: Patients newly starting treatment with Anakinra (ANA) as first-line biologic were compared to ANA second-line users regarding efficacy and safety.

Methods: ANA treatment courses are documented by the German Biologics registry (BiKeR). Demographics, clinical characteristics, disease activity parameters as well as efficacy and safety parameters were compared in pts using ANA as first biologic with those previously treated with other biologics. For efficacy analyses ACR inactive disease, JADAS minimal disease activity (MDA), JADAS remission (REM) and active joint count as well as functional ability assessment (CHAQ) were used. "Intention to treat" (ITT) and "as observed" (AO) analyses were performed. Safety assessments were based on adverse events (AE) reports.

Results: 47 pts started treatment with ANA, 33 pts were pretreated with other biologics (Etanercept n=29, Tocilizumab n=4, Infliximab and Canakinumab n=1). ANA first line pts had significantly shorter disease duration and had received DMARDs and i.a. steroids significantly less frequent than second line users. At baseline, ANA first-line users had higher disease systemic activity indicators (fever, rash, ESR), functional disability (CHAQ) and impaired parent/patient global assessment of overall well-being significantly more often, while there were no significant differences in Physician global assessment of disease activity, active joint count and JADAS 10. At last follow-up 43%>66% of pts in both groups reached ACR inactive disease, 43%>63% JADAS MDA, 29%–50% JADAS REM and 43%>69% reached status of no functional disability. Rates did not differ between the 2 cohorts (table 1). 13 pts experienced adverse events (AE) with a rate of 53/100PY, 5 pts experienced serious AE (9/100PY) and in 11pts infections were reported (33/100PY). Macrophage activation syndrome was reported once in each group. One case of AML was reported in a patient one year after last exposure to ANA. No deaths were reported.

Conclusions: Anakinra is a feasible first-line biologic for treatment of sJIA. In the BiKeR cohort ANA treated pts showed a marked treatment response, as first or consecutive biologic. Rates of AE, SAE and infectious AE seem acceptable.

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THU0562 EVOLUTION OF SERUM CALPROTECTIN IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS IN CLINICAL PRACTICE

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Background: Serum Calprotectin (MPR8/MPR14) is a promising biomarker in the management of juvenile idiopathic arthritis (JIA), mainly as a predictor of flare, especially in treatment de-escalation

Objectives: To describe the evolution of serum Calprotectin in patients with JIA, their clinical evolution and its impact on therapeutic decisions