Objectives: The aim of our retrospective observation was to describe trends in the biologics prescription in first and subsequent lines for the treatment of non-systemic JIA.
Methods: We recruited 252 patients with non-systemic JIA, who received medical treatment with biologics at the Department of Paediatric Rheumatology of Sechenov University from January 2015 to December 2017. 18% of them (n=46) had chronic anterior uveitis at the moment of biologic therapy initiation. Eye involvement influenced treatment decisions in these patients, and therefore they were excluded from the analysis. 206 children with non-systemic JIA and without uveitis were included in a study group.
Patients' characteristics: 128 girls and 78 boys (1,6/1). Mean age 13,5 ± 3,8 years, age of disease onset 7,8 ± 4,2 years; mean disease duration before biologic therapy initiation 3,3 ± 3,4 years. JIA categories: 135 (65,5%) had polyarticular RF-, 29 (14,1%) – oligoarticular, 26 (12,6%) – enthesitis-related, 9 (4,4%) – polyarticular RF+, and 7 (3,4%) – psoriatic subtype. At baseline, 204 patients (99,0%) received concomitant therapy with conventional DMARDs: 175 children (85,0%) received Methotrexate (MTX), 3 (1,5%) – combination of MTX and sulfasalazin (SSZ), 9 (4,4%) – SSZ, 9 (4,4%) – leflunomid, 4 (1,9%) – combination of MTX and leflunomid, 4 (1,9%) – cyclosporine A. Two children (1,0%) received biologics as monotherapy.
Results: As a first biologic abatacept was used in 26 patients, adalimumab in 14, tocilizumab in 6 and infliximab in 4. Etanercept was prescribed to 156 patients (75,7%± 3,5) with non-systemic JIA, which was significantly more frequently than other biologics together – 50 (24,3%± 6,2) (t=7,1; P>99,7%). Later 24 children (11,7%) were switched from a first to a second biologic agent. The main reason for switching (n=14) was inefficacy of the first-line drug. When etanercept was ineffective, abatacept (n=3) and adalimumab (n=5) were prescribed as second-line biologics. Etanercept was used as a second-line biologic in 6 children, who were initially treated with other biologic agents. The other reason for switching was the appearance of chronic anterior uveitis (n=7): 6 of these children were initially treated with etanercept, 1 with abatacept; they were switched to adalimumab (n=6) and abatacept (n=1). In 2 cases adverse events were observed – one episode of intrathoracic lymphatic nodes tuberculosis in a patient receiving etanercept, and one allergic reaction (rush, asphyxia) after the infliximab injection. At the end of our observation etanercept was chosen as treatment for 148 (71,8%) patients with non-systemic JIA.
Conclusion: Thus, etanercept was preferred biologic agent in the treatment of non-systemic JIA. The presence of uveitis requires a different treatment approach.

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AB0988 THE ASSOCIATION BETWEEN CLINICAL FEATURES AND ANALYSIS OF MEFV GENE IN 20 JAPANESE PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER (FMF)

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Background: FMF is recessive systemic auto inflammatory disorder characterized by recurrent fever, peritonitis, pleuritis, pericarditis and arthritis accompanied with skin rash. Mutation of MEFV gene encoding pyrin resulted in inflammasome activation and the uncontrolled production of IL-1β. Overview of pathogenesis, clinical features and management in Japanese patients with FMF had been reported[1]. However, the differences of clinical features between mutated and non-mutated of MEFV still remain unclear.
Objectives: We have analyzed 20 Japanese patients with FMF to clarify the association between various clinical features and mutation of *MEFV*.
Methods: Genomic DNA were purified from white blood cells in 20 FMF patients, and mutated MEFV has been explored. We have analyzed *MEFV, TNFRSF1A, MVK and NLRP3 genes* in 20 patients with FMF. Therefore, we excluded another autoinflammatory diseases such as TNF receptor-associated syndrome (TRAPS), mevalonate kinase deficiency and cryopyrin-associated periodic syndrome. Clinical symptoms and laboratory data were analyzed around onset time. Each patient had been treated with colchicine (0.5-2 mg).
Results: Characteristics of Patients with FMF (13 female/7 male) were as follows; Onset time were 0-53 years old (17.5 ±12.2), and teen aged patients were most. Frequencies of clinical symptoms such as periodic fever, headache, arthralgia, abdominal pain, chest pain, myalgia, and cervical lymph nodes swelling were 20/20, 7/20, 6/20, 5/20,4/20,2/20 and 1/20, respectively (double symptoms were observed). Patients with FMF were divided to 3 groups as follows; Patients with typical compound heterozygous mutations of MEFV (E148Q/M694I) which indicated exon 10 mutation, were 3 cases (group 1). Patients with atypical mutations, except for mutations in exon 10, such as exon 1 (E84K, L110P), 2 (E148Q), 3 (P369S, R408Q), 5(S503C) and 9(I591M) were 8 cases (group 2). Patients with no mutations in MEFV gene were 9 cases (group 3). There were no significant differences of age at first visiting hospital (FV), onset age of fever attack (O), duration of fever attack (D) and frequency of fever attack (FF) between group 1, group2 and group 3 (FV: 22.3 ± 4.5, 26.8 ± 14.6 years old (yo) and 29.5 ± 8.7 yo, O: 11.6 ± 2.4 yo, 18.5 ± 13.8 yo and 18.5 ± 12.0 D: 3.0 ± 1.4 hrs, 6.3 ± 3.2 hrs, and 8.1 ± 8.5 hrs, FF: 1.2 ± 0.2/month (M), 1.2 ± 1.1/M, and 1.1 ± 0.2/M), respectively. Laboratory examinations such as WBC, CRP and serum amyloid A (SAA) were not significantly different between 3 groups. All of those patients were effective for colchicine treatment except for 2 patients in group 1 because of severe diarrhea and alopecia. Finally, 2 patients in group 1 received canakinumab treatment. Mutations of E148Q were found in 9 patients (45%).
Conclusion: We have examined association between clinical features and mutations of MEFV in 20 Japanese patients, suggesting no positive findings in Japanese patients with FMF. Mutations of E148Q in exon 2 were observed in 16-23 % of normal Japanese patients[1], indicating that E148Q is the polymorphism or accelerating factor.

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AB0989 PATIENTS' AND CAREGIVERS' ASSESSMENT OF A DEDICATED OUTPATIENT SERVICE FOR INTRAARTICULAR GLUCOCORTICOID INJECTIONS IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: Patients with juvenile idiopathic arthritis (JIA) may require several hospital admissions over the disease course, due to flares or persistently active arthritis, with a negative impact on the patients' and family's daily life. To provide timely intervention and support the patients' and families' quality of life, in 2018 an afternoon outpatient service for intraarticular glucocorticoid injections (IAGI) in JIA has been created at the study center.
Objectives: To evaluate the patients' and caregivers' assessment of the outpatient service for IAGI in JIA; to investigate demographic and clinical features of patients entering the service.
Methods: All consecutive JIA patients and their caregivers seen at the IAGI outpatient service from February 2018 to January 2019 completed a satisfactory questionnaire just after the IAGI procedure. The patient's part included: satisfaction on the overall service and on dedicated personnel (yes/no, why), procedure pain assessment (VAS 0-10, 0=none; 10 worst); whereas the caregiver's part: satisfaction on the overall service (yes/no, why), facilitation of family burden (yes/no, why). Demographic and clinical data of patients, including previous hospitalization for IAGI under general sedation or local anesthesia and geographical provenance, were registered during the questionnaire completion. Descriptive analysis was performed on data. Open answers were synthetized in items.