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 $13,5 \pm 3,8$ years, age of disease onset $7,8 \pm 4,2$ years; mean disease duration before biologic therapy initiation $3,3 \pm 3,4$ 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 sulfasalazine (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 ($75,7\% \pm 3,5$) with non-systemic JIA, which was significantly more frequently than other biologics together – 50 ($24,3\% \pm 6,2$) ($t=7,1$; $P>99,7\%$). 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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Disclosure of Interests: Anna Ignatova: None declared, Nina Seylanova: None declared, Elena Zholobova Grant/research support from: Roche, Pfizer, Novartis, Consultant for: Pfizer, AbbVie, Roche, BMS, Novartis, MSD, Speakers bureau: Pfizer, AbbVie, Roche, BMS, Novartis, MSD
DOI: 10.1136/annrheumdis-2019-eular.3462

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-1 β . Overview of pathogenesis, clinical features and management in Japanese patients with FMF had been reported¹. 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 ($17,5 \pm 12,2$), 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: $22,3 \pm 4,5$, $26,8 \pm 14,6$ years old (yo) and $29,5 \pm 8,7$ yo, O: $11,6 \pm 2,4$ yo, $18,5 \pm 13,8$ yo and $18,5 \pm 12,0$ D: $3,0 \pm 1,4$ hrs, $6,3 \pm 3,2$ hrs, and $8,1 \pm 8,5$ hrs, FF: $1,2 \pm 0,2$ /month (M), $1,2 \pm 1,1$ /M, and $1,1 \pm 0,2$ /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 patients¹, indicating that E148Q is the polymorphism or accelerating factor.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.1327

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 synthesized in items.

Results: All of the 46 JIA patients seen at the IAGI outpatient service and their caregivers completed the questionnaire. Patients were mostly females (78%) with early disease onset (median 4.8 years) and positive ANA status (61%). The majority (52%) had persistent oligo-JIA, followed by extended oligo-JIA (26%), RF-negative polyarthritis (17%), ERA and systemic JIA (2%, respectively). The median age at IAGI was 14.7 years (IQ 11.3-19.5). Forty patients (87%) had previous hospitalization for IAGI, mostly under general sedation (64%). All patients, except a 12 year old girl (0.02%) with uncontrolled needle phobia and previous IAGI in general sedation, (99.98%) and all caregivers (100%) were satisfied with the IAGI outpatient service. In Table 1 reasons of satisfaction are detailed in items. Procedural pain was median rated 3 (IQ 2-6). Most of the families came from the city or province of the study center (76%); 17% from other provinces of the same geographical region; 7% from other regions.

Abstract AB0989 Table 1. Frequency of specific reasons of satisfaction on the IAGI outpatient service, synthesized in items, during the 56 procedures of the study.

Items	Patients, n (%)	Parents, n (%)
Time saving	45 (80)	30 (54)
Stress saving	5 (9)	-
Reduction of absence from school/work	27 (48)	23 (41)
Less negative impact on personal commitments	-	26 (46)
Improvement of family burden organization	-	54 (96)
High quality of dedicated Staff	55 (98)	19 (34)

Figure 1. Proportion of specific sites injected in the 46 JIA patients in the 56 procedures performed at the outpatient dedicated service during the study

Figure 1

Figure 2. Proportion of procedures with one or multiple injected sites at the IAGI outpatient service during the study

Figure 1

Conclusion: The results outline the afternoon IAGI outpatient service foster the management of flares or persistently active arthritis in JIA patients, particularly at older ages, with high rate of satisfaction and lower impact on patients' and family burden compared to hospital admission and, of note, despite moderate pain rating and multiple site injections. This supports the development of Diagnostic Therapeutic Care

Pathways for JIA patients, in the view of improving patients' quality of life and also towards resource optimization.

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Disclosure of Interests: Hanan Jadoun: None declared, Aurora Pucacco: None declared, Angela Aquilani: None declared, Andrea Uva: None declared, Fabio Basta: None declared, Rebecca Nicolai: None declared, Fabrizio De Benedetti Grant/research support from: Abbvie, SOBI, Novimmune, Roche, Novartis, Sanofi, Pfizer, Silvia Magni-Manzoni Consultant for: Abbvie, Speakers bureau: Abbvie

DOI: 10.1136/annrheumdis-2019-eular.1720

AB0990 FINAL DIAGNOSIS OF THE PATIENTS WITH MUSCULOSKELETAL COMPLAINTS: PRELIMINARY RESULTS OF ONE-YEAR STUDY

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Background: Musculoskeletal (MS) complaints are one of the most common reason for administration to outpatient clinics.

Objectives: The study aimed to summarize the final diagnosis of the patients who suffered from musculoskeletal (MS) findings.

Methods: We prospectively evaluated the patients who were referred with the complaints of MS systems in a year period.

Results: A total of 940 patients with the complaint of musculoskeletal systems were examined. Among them, 577 patients suffered from arthralgia, 234 had arthritis, 39 had low-back pain, 26 had limping, and 64 had other symptoms such as myalgia, heel pain, hip and neck pain. A diagnosis of rheumatic disease was made in 430 of patients, while 510 had non-rheumatic conditions. Final rheumatological disease diagnoses were as follows: juvenile idiopathic arthritis (n=195), familial Mediterranean fever (n=101), reactive arthritis (n=46), acute rheumatic fever (n=39), toxic synovitis (n=17), psoriasis (n=8), Raynaud's phenomenon (n=6), chronic recurrent multifocal osteomyelitis (n=5), vasculitis (n=5), systemic lupus erythematosus (n=4), juvenile dermatomyositis (n=2) and juvenile scleroderma (n=1). Among patients with non-rheumatic conditions, most of them had vitamin D deficiency, infections, mechanic-orthopedic conditions, and growing pains.

Conclusion: Evaluation of a child presenting with MS findings requires a comprehensive, multidisciplinary, and systematic approach. As a busy pediatric rheumatology center, we demonstrated the final diagnosis of referred patients with MS symptoms and only less than half of the patients were diagnosed as a rheumatological condition. Whilst pediatric rheumatology centers are limited in number, our results put forth the need of formulating recommendations for clinicians in order to prevent unnecessary referrals

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.1991

AB0991 PRELIMINARY RESULTS OF REFERRALS TO A TERTIARY PEDIATRIC RHEUMATOLOGY OUTPATIENT CLINIC: A YEAR IN REVIEW

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Background: Previously, the profile of pediatric rheumatology practice was demonstrated from several countries(1,2). However, there is not a