Background: Juvenile Idiopathic Arthritis (JIA) is a heterogeneous group of pediatric diseases. Different response to biological treatment (BT) has been reported according to disease subtype.

Objectives: To analyze the prescription and withdrawal of BT in JIA patients with focus on JIA category.

Methods: A retrospective observational study was conducted on JIA patients followed in a referral hospital and who had received at least one BT between 1999 and 2019.

Results: 130 JIA patients were analyzed: 29 (22,4%) were Oligoarticular Persistent (OligP), 22 (16,9%) Enthesitis related Arthritis (ERA), 20 (15,4%) Systemic (sJIA), 19 (14,6%) Polyarticular RF- (PolyRF-), 14 (10,8%) Polyarticular RF+(PolyRF+), 13 (10%) Oligoarticular-Extended (OligE), 11 (8,4%) Psoriatic Arthritis (APso) and 2 (1,5%) Undifferentiated (Und).

The main characteristics are summarized in table 1.

The first line BT most frequently indicated was Etanercept up to 40% in all the categories except for ERA, where the most frequent BT was Adalimumab and sJIA, where the most frequent BT was Anakinra. The time between diagnosis and start of BT was different among the categories ($p=0,007$). In the Und category, the time until BT was the shortest (median: 1 month), since both patients had coxitis, followed by APso [median: 9 months IQR(1-57)] and sJIA [median: 17,5 months IQR(0,3-146,8)]. The survival of the first BT was different among the categories ($p=0,006$): 94,7% of the ERA continue receiving the first BT, followed by 76,2% of OligP and 50% of PolyRF+ and APso. Only 42% of sJIA continue on the first BT prescribed [up to 53,3% were TNF inhibitors (TNFi)]. The categories with less retention of the first BT were: OligE (25%); PolyRF- (27,3%) and Und (0%). The most frequent cause of discontinuation, among these categories, was secondary failure.

In the survival analysis between categories, there were differences on OligP ($p=0,004$), OligE ($p=0,042$) and PolyRF- ($p=0,017$). Tocilizumab and Adalimumab were the BT with highest survival with regards to Infliximab, Etanercept, Rituximab (OligE, PolyRF-), Abatacept (OligE, PolyRF-) and Certolizumab (OligP). The survival rate of IL1 inhibitors and IL6 inhibitors was higher regarding to TNFi in sJIA patients ($p=0,013$).

Conclusion: Taking into account JIA category is mandatory to choose BT and to understand the response and discontinuation of BT. OligE and PolyRF- showed a high rate of change of the first BT related to secondary failure of Etanercept and Infliximab when compared to Adalimumab and Tocilizumab, as described in the survival analysis. The category with the highest retention of the first BT was ERA. UND patients started sooner BT due to the presence of coxitis. In sJIA, IL1 inhibitors and IL6 inhibitors were superior to TNFi in the survival analysis, as reported in existing literature.

Table:

Table 1	OligP	ERA	sJIA	PolyRF-	PolyRF+	OligE	Apso	UND
Sex, n%								
M	4(13,8)	17(77,3)	11(55)	2(10,5)	2(14,3)	1(7,7)	4(36,4)	1(50)
F	25(86,2)	5(22,7)	9(45)	17(89,5)	12(85,7)	12(92,3)	7(63,6)	1(50)
Age at diagnosis me, IQR	4 (2,6-5)	12 (9,8-15)	7 (3-13)	8 (2-13)	12 (8,5-15)	3,5 (2-8,3)	12 (3-15)	12,5
Uveitis, n%	12(41,4)	7(31,8)	0(0)	3 (15,8)	0 (0)	3(25)	2 (18,2)	1(50)
ANA, n (%)	22(75,9)			8 (42,1)	12 (85,7)	9(75)	5 (45,5)	
ACPA, n (%)		18(81,8)			9(64,3)		3(27,3)	
B27, n (%)								
BT lines: n	1 st : 29 2 nd : 11 3 rd : 1	1 st : 22 2 nd : 2	1 st : 20 2 nd : 10 3 rd : 5 4 th : 1 5 th : 1	1 st : 11 2 nd : 9 3 rd : 6 4 th : 3	1 st : 14 2 nd : 7 3 rd : 2 4 th : 2	1 st : 13 2 nd : 9 3 rd : 2 4 th : 1 5 th : 1	1 st : 11 2 nd : 5 3 rd : 4 4 th : 3 5 th : 1	1 st : 2 2 nd : 2

APso, Psoriatic Arthritis; BT, biological treatment; ERA, Enthesitis related Arthritis; F, female; M, male; OligE, Oligoarticular-Extended; OligP, Oligoarticular-Persistent; PolyRF-, Polyarticular RF-; PolyRF+, Polyarticular RF+; sJIA, Systemic JIA; Und, Undifferentiated

Disclosure of Interests: None declared

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AB0985 THE EFFECT OF GENDER ON CHILDREN AND ADOLESCENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Background: In the literature, it was reported that children with with Familial Mediterranean fever (FMF) have lower functional capacity and muscle strength than healthy children. With reduced functional capacity, daily activities are negatively affected. The individual starts to adopt an inactive lifestyle and decreases in muscle strength are observed. A vicious circle occurs and results in exacerbation of symptoms and worse quality of life.

Objectives: The aim of this study is to investigate the effect of gender on dynamic muscular endurance, physical activity and quality of life with FMF.

Methods: Forty-eight children and adolescents (26 girls, 22 boys, mean age=12.43±3.04 years, age range=7-18 years) were included. Exclusion criteria: The presence of another disease. Intraarticular steroid injection or surgery in any joint in the last 3 months. Evaluations were made by the same pediatric rheumatologist and physiotherapist by face to face interview method. Dynamic muscle endurance was evaluated by use of curl up test (30 sec), push up test (30 sec) and one-legged stationary hop test (15 sec); physical activity level by Physical Activity Questionnaire and quality of life by Pediatric Quality of Life Inventory (PedsQL) 3.0 Arthritis Module. Physical Activity Questionnaire contains nine items and evaluates physical activities in last seven days and frequency of these activities. As score increases, level of physical activity increases. In this study, child and parent forms of PedsQL were used to evaluate the quality of life. High scores mean high quality of life. Independent Samples Test was used in the analysis because the data fit the normal distribution.

Results: Difference was significant in Physical Activity Questionnaire ($p=0.028$) in favor of girl gender whereas in child form ($p=0.017$) and parent form ($p=0.040$) of PedsQL ($p=0.003$) in favor of boy gender.

Conclusion: We see that the physical activity are lower in children and adolescents with FMF who have a gender of girls and, accordingly, lower quality of life. Therefore, we believe that these individuals should increase their physical activities.

References:

- Gualano B, S'a Pinto AL, Perondi B, Leite Prado DM, Omori C, Almeida RT, Sallum AM, Silva CA. Evidence for prescribing exercise as treatment in pediatric rheumatic diseases. *Autoimmun Rev* 2010;9(8):569-573.
- Alayli G, Durmus D, Ozkaya O, Sen HE, Nalcacioglu H, Bilgici A, Kuru O. Functional Capacity, Strength, and Quality of Life in Children and Youth With Familial Mediterranean Fever. *Pediatric Physical Therapy* 2014;26(3):347-352.

Table 1. Demographic characteristics

Variables	Girls (n=26)	Boys (n=22)
	M±SD	M±SD
Age (years)	12.96±2.91	11.86±3.24
Body weight (kg)	42.76±12.55	40.17±11.89
Height (m)	1.50±0.13	1.48±0.15
BMI (kg/m ²)	18.25±2.90	17.85±3.39
Age of onset (years)	7.57±4.07	7.22±3.99
Dose of colchicine (per day)	1.18±0.60	1.63±0.68
Duration of treatment (years)	5.30±2.90	4.61±3.34
C-reactive protein (mg/dl)	0.88±2.28	1.25±2.91
Pras et al severity score	6.35±2.68	5.31±2.60
	n (%)	n (%)
FMF in the family (Yes/No)	4(15,4)/16(61,5)	12(54,5)/10(45,5)
Arthritis (Yes/No)	9(34,6)/17(65,4)	6(27,3)/16 (72,7)
Arthralgia (Yes/No)	20(76,9)/6(23,1)	14(63,6)/ 8(36,4)
Knee (Yes/No)	6(23,1)/20(76,9)	7(31,8)/15 (68,2)
Elbow (Yes/No)	1(3,8)/25(96,2)	0(0)/22(100)
Ankle (Yes/No)	6(23,1)/20(76,9)	7(31,8)/15(68,2)
Hand (Yes/No)	1(3,8)/25(96,2)	1(4,5)/21(95,5)

Table 2. Comparison of data in terms of gender

Variables	Girls (n=26)	Boys (n=22)	p*
	M±SD	M±SD	
Curl Up Test (repeat)	10.70±3.58	12.80±3.66	0.075
Push Up Test (repeat)	10.55±5.06	11.75±6.29	0.511
One-legged Stationary Hop Test (repeat)	37.60±12.74	35.35±6.02	0.480
Physical Activity Questionnaire	16.93±5.70	20.70±5.77	0.028
PedsQL- child form	19.15±13.48	10.68±9.38	0.017
PedsQL- parent form	22.30±17.85	13.04±11.06	0.040

Disclosure of Interests: None declared

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AB0986 NT-PROBNP LEVEL IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: The defeat of the cardiovascular system is considered a proven comorbid state in rheumatic diseases, including rheumatoid arthritis in adults. One of the markers of the cardiovascular failure formation is BNP, namely, its N-terminal inactive fragment (NT-proBNP76), which accumulates in specific granules of cardiomyocytes. Its diagnostic value increases with the appearance of minimally expressed symptoms. The long-term course of JIA is also characterized by changes in the state of the cardiovascular system, and there may be no visible clinical manifestations. For their diagnosis a 6-minute walk test is widely used, including in children.

Objectives: To study the content of NT-proBNP in patients with juvenile idiopathic arthritis and compare with the level of exercise tolerance.

Methods: 10 patients with JIA (9 girls, 1 boy), average age 12.78 ± 0.95 years, were examined. All children had a polyarticular RF negative subtype of JIA with a disease duration of more than three years (average disease duration 69.56 ± 17.07 months), received basic methotrexate therapy and did not have dysfunction of the lower extremities joints. The control group included 7 healthy children, comparable by sex, average age 14.25 ± 0.73 years. An ECG, an ultrasound scan of the heart, and a 6-minute walk test (6MTX) were carried out with determining the distance traveled (6MWD) and the increase in heart rate. The level of the N-terminal polypeptide of cerebral natriuretic hormone (B-type) (NT-proBNP) was determined in the morning, after waking up, and studied by competitive immunoassay on an IMMULITE 2000 analyzer ("Siemens").

Results: In children with JIA a decrease in myocardial contractility was not detected. Left ventricular ejection fraction ($62.17 \pm 0.83\%$ ($60.02 - 64.02$) versus $69.84 \pm 0.85\%$ ($62.3 - 80.3$), $p < 0.05$) in children with JIA were within normal limits, but significantly lower than in the control group. According to the results 6MTX indicator 6MWD in JIA-patients was 490.51 ± 11.40 m and in the control group 516.85 ± 8.84 m ($p < 0.05$) and heart rate growth was $27.75 \pm 2.30\%$ versus in the control group ($37.38 \pm 3.86\%$), $p < 0.05$. A negative correlation between the increase in heart rate and the duration of the disease was found ($r = -0.7$, $p = 0.05$). The level of NT-proBNP in patients with JIA was within physiological values and amounted to 47.5 ± 14.09 pg / ml ($20 - 128$ pg / ml), but this was higher than in children of the control group (20.29 ± 0.29 pg / ml ($20-22$ pg/ml), $p < 0.05$). A high correlation was found between 6 MWD and NT-proBNP level ($r = 0.8$, $p < 0.03$).

Conclusion: In children with JIA there is a decrease of the exercise tolerance that increases with the duration of JIA on the background of preserved myocardial contractility. This is accompanied by a higher basal NT-proBNP level than in healthy children.

Disclosure of Interests: None declared

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AB0987

STUDY ON THE FACTORS AFFECTING THE FINAL ADULT HEIGHT IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile idiopathic arthritis (JIA), a chronic inflammatory disease involving limited joints and/or constitutional symptoms in childhood or adolescent, might affect body height and result in short stature in adulthood. Well-controlled disease activity is beneficial in improving growth impairment in JIA patient.

Objectives: To identify any factors that influence final adult height in the patient with juvenile idiopathic arthritis.

Methods: We retrospectively reviewed the medical records of JIA patients between 2009 to 2019 in National Taiwan University Hospital. The diagnosis of JIA was according to the International League of Associations for Rheumatology (ILAR) criteria. Personal history, laboratory reports, and medication were analyzed. The difference between final adult height and target height was calculated in each patient. We defined whose final adult height higher than target height as positive group and the others as negative group. A cox univariate proportional hazards model was applied to compare the variables between these two groups.

Results: Total 120 patients are collected. There are 74 (61.7%) and 46 (38.3%) cases in the positive and the negative group, respectively. The mean onset age of disease is 11.78 ± 3.78 in positive group and 10.83 ± 4.04 in negative group. Male is more than female in both groups, with a ratio of 1.7:1 and 2.7:1 respectively. Enthesis-related arthritis accounted for the most in positive group, however, in negative group, oligo-arthritis was the most common type. There are slightly more patients having received biologics in positive group than negative group without significance (64.8% vs. 54.3%, $p=0.33$). Among the patients treated with etanercept, those in the negative group had a significantly younger age when they started the treatment compare to the positive group (11.83 ± 1 vs. 13.9 ± 3.1 , $P = 0.01$). The patients in positive group had significantly higher final adult

height (171.08 ± 7 vs. 162.9 ± 8.1 , $P < 0.001$). The univariate analysis showed that the age when etanercept treatment started was associated with the occurrence of a negative difference between final height and target height (odds ratio=0.80, 95%CI=0.67-0.96, $P=0.02$).

Conclusion: In JIA patients, 38.3% of them had a negative difference between final adult height and target height. The subtype might play a critical role in affecting the growth of patients. A younger age when the child received etanercept was associated with a lower attained adult height than target height, however, it needs further analysis on the medication of these children.

References:

- [1] S.-J. Wang et al., Attained Adult Height in Juvenile Rheumatoid Arthritis with or without Corticosteroid Treatment, *Clin Rheumatol* (2002) 21:363–368
- [2] P Tynja¹ et al., Impact of anti-TNF treatment on growth in severe juvenile idiopathic arthritis, *Ann Rheum Dis* 2006;65:1044–1049
- [3] Edward H. Giannini et al., Effects of Long-Term Etanercept Treatment on Growth in Children With Selected Categories of Juvenile Idiopathic Arthritis, *ARTHRITIS & RHEUMATISM* Vol. 62, No. 11, November 2010, pp 3259–3264
- [4] Ashley Shafferman et al, Changes in Body Mass Index in Children with Juvenile Idiopathic Arthritis Treated with Tumor Necrosis Factor Inhibitors, *J Rheumatol* 2014;41:113-118
- [5] Lianne Kearsley-Fleet et al., Growth in children and adolescents with juvenile idiopathic arthritis over 2 years of treatment with etanercept: results from the British Society for Paediatric and Adolescent Rheumatology Etanercept Cohort Study, *Rheumatology* 2015;54:1279-1285
- [6] Florence Uettwiller et al., Effect of Biologic Treatments on Growth in Children with Juvenile Idiopathic Arthritis, *J Rheumatol* 2014;41:128-135
- [7] Lianne Kearsley-Fleet et al., Short-term outcomes in patients with systemic juvenile idiopathic arthritis treated with either tocilizumab or anakinra, *Rheumatology* 2019;58:94-102

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AB0988

CLINICAL FEATURES AND ANALYSIS OF MEFV GENE IN 31 PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER (FMF)

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Background: FMF is recessive systemic autoinflammatory disorder characterized by recurrent fever, peritonitis, pleuritis, pericarditis and arthritis accompanied with headache and abdominal pain. 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 reported. However, the differences of clinical features between mutated and non-mutated of MEFV still remain unclear.

Objectives: We have analyzed 31 Japanese patients with FMF in Gifu district to clarify the association between various clinical features and mutation of MEFV.

Methods: Genomic DNA were purified from white blood cells in 31 FMF patients, and mutated MEFV has been explored. We have analyzed MEFV, TNFRSF1A, MVK and NLRP3 genes in 31 patients with FMF except for 1 patient. 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 (22 female/9 male) were as follows; Onset time were 0-56 years-old (21.4 ± 11.8), and Frequencies of clinical symptoms such as periodic fever, headache, abdominal pain, arthralgia, chest pain, cervical lymph nodes swelling, and myalgia were 31/31, 9/31, 8/31, 6/31, 5/31, 3/31 and 1/31, respectively (double symptoms were observed). Patients with FMF were divided into 3 groups as follows; Patients with typical compound heterozygous mutations of MEFV (E148Q /M694I) which indicated exon 10 mutation, were 5 cases (G1). Patients with atypical mutations, except for exon 10, such as 133G>A in 3'UTR, exon 1 (E84K), 2 (L110P, E148Q), 3(R202Q, P257L, G304R, P369S, R408Q), 5(S503C) and 9(I591M) were 13 cases (G2). Patients with no mutations in MEFV gene were 12 cases (G3). There were no significant differences of age at first visiting hospital (FV) and onset age of fever attack (O) (FV: 29.0 ± 15.6 , 27.1 ± 12.5 years-old (yo) and 34.7 ± 12.7 yo, O: 21.0 ± 17.6 yo, 17.8 ± 12.1 yo and 25.2 ± 6.5). But significant differences in duration of fever attack (D) and frequency of fever attack (FF) between G1 and G2 or G3 were observed as follows; (D: 2.2 ± 0.4 days vs 5.5 ± 3.1 days, $P < 0.05$, and 3.8 ± 1.7 days), FF: $0.72 \pm 0.3/$