Autoimmune and Autoinflammatory Diseases, Saint-Petersburg, Russian Federation

Background: Crimean Tatars is an ethnic group in Russia. The presence of familial Mediterranean fever (FMF) has been recognized since 2016.

Objectives: The study aimed to evaluate the prevalence and clinical features as well as genetic aspects of FMF in children of Crimean Tatar (CT) origin and compare them with a cohort from Turkey.

Methods: This retrospective study included all FMF cases in patients of CT origin (n=18) diagnosed in Children's Regional Hospital in Simferopol since 2016. We included 40 consecutive FMF cases between February-March 2020, diagnosed and followed at Hacettepe University, Ankara, Turkey. All children were less than 18 years old at the time of inclusion. The diagnosis of FMF was based on the EULAR criteria (2019). We excluded other autoinflammatory diseases and any doubtful cases. For assessment of MEFV alleles prevalence 127 healthy unrelated CT adults from different parts of Crimea peninsula were included. Sanger sequencing of MEFV exons 2 and 10 was performed in all the patients and controls.

Results: FMF in CT was diagnosed with nearly 5 year-delay, despite the earlier age at onset. CT children had more frequent and prolonged fever, joint involvement (arthritis and arthralgia) and erysipeloid rash compared to Turkish, who had more attacks with chest pain and abdominal pain which last longer. (Table 1) CT had higher white blood cell count, C-reactive protein, erythrocyte sedimentation rate and lower hemoglobin. It might be explained by the fact that the majority of Crimean Tatars were admitted to the clinic during an attack, which was not always the case with Turkish children. Distribution of MEFV pathogenic alleles p.M694V, p.M680I, p.V726A in CT children was 81%, 9.5% and 9.5%, respectively, while in Turks it was 68.6%, 14.3% and 12.9%. Among the CT patients, proportion of homozygotes, compound-heterozygotes and heterozygotes were 11%, 6% and 83%, and among Turkish patients were 45%; 30%; and 25%, respectively. MEFV pathogenic variants were detected in 10.2% of healthy CT donors: 7.1% individuals had p.M694V, 1.6% - p.M680I, 1.6% - p.V726A. Comorbid diseases including IgA vasculitis, sacroiliitis, JIA, autoimmune hepatitis and inflammatory bowel disease were reported in 5.6% of CT and 10% of Turks. The colchicine treatment rate and regimen were similar, but CT received biologics more frequently (44%) than Turks (22.5%).

Conclusion: CT is an ethnic group with a significant number of MEFV mutation carriers assuming the expected prevalence of FMF to be as high as 1:385. Thus, any periodic fever in CT patients should be considered as a sign of possible FMF. The clinical course of FMF has some peculiarities in CT patients.

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Table 1. Comparative data of FMF patients with Turkish and Crimean Tatar origin

FMF features	Turkish (n=40)	Crimean Tatars (n=18)	p
Family history of FMF, n (%)	16 (40)	9 (50)	0.477
Consanguinity, n (%)	8 (20)	6 (33)	0.272
Onset age, years	3.3 (2; 5)	1.3 (0; 4)	0.040
Age at FMF diagnosis, years	4.7 (3; 8)	9.6 (4; 14)	0.005
Diagnosis delay, years	0.9 (0; 2)	5.5 (2; 10)	0.00001
Fever, n (%)	33 (83)	18 (100)	0.058
Episode duration, days	2.0 (2; 3)	3.0 (3; 6)	0.000003
Fever duration, hours	48.0 (48; 72)	72.0 (72; 120)	0.000002
Chest pain, n (%)	12 (30)	1 (6)	0.039
Chest pain duration, hours	48.0 (24; 72)	0.0 (0; 0)	0.000001
Abdominal pain, n (%)	30 (75)	9 (50)	0.061
Abdominal pain duration, hours	48.0 (24; 48)	24.0 (24; 24)	0.043
Arthritis, n (%)	10 (25)	16 (89)	0.000006
Arthralgia, n (%)	19 (48)	17 (94)	0.0007
Erysipeloid rash, n (%)	0 (0)	9 (50)	0.000001

Disclosure of Interests: None declared

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POS1299 RISK FACTORS OF TOTAL HIP ARTHROPLASTY IN JUVENILE ARTHRITIS WITH HIP INVOLVEMENT

L. Sorokina¹, I. Avrusin¹, R. Raupov¹, N. Garipova², M. Gharabaghtsyan³, S. Khrypov⁴, E. Isupova¹, E. Gaidar¹, I. Chikova¹, M. Dubko¹, V. Masalova¹, T. Likhacheva¹, L. Snegireva¹, M. Kostik^{1,5}. ¹Saint-Petersburg State Pediatric Medical University, Department of Hospital Pediatrics, St. Petersburg, Russian Federation; ²Almazov National Medical Research Centre, Pediatric Department, St. Petersburg, Russian Federation; ³Institute of Child and Adolescent Health (ICAH) with the Arabkir Medical Center, Pediatric Department, Yerevan, Armenia; ⁴Saint-Petersburg Clinical Research-Practical Center for Specialized Healthcare (Oncology), Surgical Department, St. Petersburg, Russian Federation; ⁵Almazov National Medical Research Centre, Autoimmune and Autoinflammatory Diseases, St. Petersburg, Russian Federation

Background: Hip osteoarthritis (HOA) is a severe outcome of juvenile idiopathic arthritis (JIA) itself and also can be result of corticosteroid (CS) treatment, if it

was used. Total hip arthroplasty (THA) is the last step in JIA treatment and indicates ineffectiveness of conservative treatment.

Objectives: We aimed to evaluate risk factors which lead to THA in JIA patients with HOA.

Methods: 753 patients aged 2-17 years were included in our retrospective study during the last 10 years. Diagnosis was made according to ILAR criteria. Clinical, laboratory and radiol examinations were evaluated. Diagnosis of HOA was made on MRI, CT and planar radiographs and confirmed by morphological examination of removed femoral heads.

Results: Total 153/753 (20.3%) patients with JIA had hip involvement. HOA developed in 48/153 (31.4%) of JIA patients and 16/48 (33.3%) of them had THA was undergone. Prevalence of HOA and THA (%) in JIA subtypes: in polyarticular (5/32 (15.6%) and 8/16 (50%), systemic (6/32 (18.7%) and 5/16 (31.2%)), enthesitis-related (19/32 (59.4%) and 3/16 (18.8%)) and psoriatic (2/32 (6.7%) and 0/16) subtypes respectively, p=0.0000001. Patients who underwent THA initially had higher level of inflammation: elevated ESR (33 vs 5 mm/h, p=0.002) and CRP (14.7 vs 1.9 mg/l, p=0.03), more active joint, and especially involvement of joints of upper limbs: elbows (p=0.004) and proximal interphalangeal joints (p=0.001), arthritis of subtalar joint (p=0.02). Delayed biologic treatment (7.5 vs 3.4 years, p=0.043) and delayed achievement of remission (9.2 vs 5.6 years, p=0.047) were main predictors of THA. Patients with HOA without biologics had increased cumulative probability of THA: HR=1.99 (1.01; 3.98), p=0.049 (Figure 1). Patients with THA received corticosteroids (93.7 vs 50%, p=0.003) more often including high dose pulse-therapy regimens, but differences in the cumulative doses were not observed (5000 vs 4500 mg, p=0.54) between groups, CS administration was independent risk factor of HOA and THA.

Figure 1. Cumulative probability of THA in JIA patients with hip osteoarthritis.

Conclusion: the main risk factors of THA are systemic and polyarticular course because of their activity, systemic CS and delayed biologic treatment. Corticosteroids should be avoided in those group of patients because of risk of avascular pathway HOA formation.

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Table 1. The features of JIA patients with hip osteoarthritis depending on

JIA features	THA, (n=16)	HOA without THA, (n=32)	p
Time to THA, years	5.2 (3.6; 10.2)	4.6 (2.2; 8.7)	0.4
Onset age, years	7.95 (3.5; 11.1)	8.3 (4.3; 13.1)	0.5
JIA duration, years	8.5 (6.5; 13.2)	5.43 (2.8; 11.1)	0.07
Polyarticular JIA, n (%)	8 (50.0)	5 (15.6)	0.037
Systemic JIA, n (%)	5 (31.3)	6 (70)	0.037
ANA, n (%)	3/8 (37.5)	5/16 (31.3)	0.760
HLA B27, n (%)	3/6 (50)	9/19 (47.4)	0.911
RF, n (%)	0/9 (0)	1/15 (6.7)	0.429
Uveitis, n (%)	1/16 (6.3)	3/24 (12.5)	0.519
ESR, mm/h	33 (13; 54)	5 (3; 27)	0.002
CRP, mg/l	14.7 (2.9; 72.3)	1.9 (0.3; 12.7)	0.03
Active joints, n	21.5 (8.5; 52.5)	9 (5; 16)	0.02
Elbows, n (%)	11 (68.7)	8 (25.0)	0.004
Proximal interphalangeal joints, n (%)	10 (62.5)	5 (15.6)	0.001
Subtalar, n(%)	4 (25.0)	1 (3.1)	0.02
Pulse-therapy GCS, n(%)	11 (68.7)	10 (31.3)	0.014
Cumulative GCS dose, mg	5000 (3000; 14000)	4500 (500; 20000)	0.54
Time to biologic, years	7.6 (4.3; 11.4)	3.4 (1.9; 8.6)	0.04
Achievement of remission, years	9.2 (6.6; 15.4)	5.6 (3.3; 11.4)	0.047

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POS1300 PAIN IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS TREATED TO TARGET: 24 MONTH RESULTS FROM A RANDOMISED TRIAL

K. Spekking¹, P. De Boer¹, S. A. Bergstra², J. M. Van den Berg³, D. Schonenberg-Meinema³, L. W. A. Van Suijlekom-Smit⁴, M. Van Rossum^{5,6}, Y. Koopman-Keemink⁷, R. Ten Cate¹, C. Allaart², D. M. C. Brinkman¹, P. C. E. Hissink Muller¹. ¹Leiden University Medical Center, Department of Pediatric Rheumatology, Leiden, Netherlands; ²Leiden University Medical Center, Department of Rheumatology, Leiden, Netherlands; ³Emma Children's Hospital, Amsterdam University Medical Centers, Department of Pediatric, Immunology, Rheumatology and Infectious Diseases, Amsterdam, Netherlands; ⁴Sophia Children's Hospital Erasmus Medical Center, Department of Pediatrics/Pediatric Rheumatology, Rotterdam, Netherlands; ⁵Emma Children's Hospital, Amsterdam University Medical Centers, Department of Pediatrics, Amsterdam, Netherlands; ⁶Amsterdam Rheumatology and Immunology Center | Reade, Department of Pediatric Rheumatology, Amsterdam, Netherlands; ⁷Hagaziekenhuis Juliana Children's Hospital, Department of Pediatrics, the Hague, Netherlands

Background: Juvenile Idiopathic Arthritis (JIA) is the most common auto-immune disease in children and pain is a common symptom. Despite treatment advances in recent years, pain often persists when the disease is inactive. Treating JIA to target has been widely recommended¹, but effectiveness in pain reduction has not been proven yet².

Objectives: To compare pain scores over two years in three treatment arms in JIA patients that were treated to target and to identify baseline characteristics predicting unfavorable pain outcomes.

Methods: DMARD naïve children who participated in the BeSt for kids study³ with oligoarticular JIA, RF negative polyarticular JIA and juvenile psoriatic arthritis were included. Patient were treated to target aimed at inactive disease according to 1 of 3 different treatment groups; 1) initial sequential DMARD monotherapy 2) initial methotrexate (MTX) with prednisolone bridging or 3) initial MTX with etanercept. Pain intensity was measured using a 100mm Visual Analogue Scale during 24 months of follow up with 3-monthly intervals. Potential differences in VAS pain scores over time between treatment arms were compared using linear mixed models with random intercept and random slope for visits clustered within patients. A similar multivariable mixed model was used to assess the ability of several baseline characteristics to predict the chance of high pain levels during follow-up.

Results: 92 patients were randomized into the three treatment groups. Overall, pain scores over time reduced from mean 55.3 (SD 21.7) at baseline to 19.5 (SD 25.3) after 24 months. When comparing pain over time per arm, pain scores decreased significantly over time β -1.37 (95% CI -1.726; -1.022). No significant difference was found in pain over time between treatment groups (interaction term treatment arm*time (months) β (95% CI) arm 1 0.13 (-0.36; 0.62) and arm 2 0.37 (-0.12; 0.86) compared to arm 3), Figure 1. Correction for sex and symptom duration as possible confounders yielded similar results. Multiple baseline characteristics demonstrated a significant predictive value for pain over time: VAS pain (scale 0-100) with β 0.44 (95% CI 0.25; 0.65), VAS physician (scale 0-100) with β -0.34 (-0.55; -0.06), number of active joints with β 0.77 (0.19; 1.34), Child Health Questionnaire Physical summary Score with β -0.42 (-0.72; -0.11) and Psychosocial summary Score β -0.42 (-0.77; -0.06). Symptom duration, VAS patient/parent and NSAID use were no significant predictors, Table 1.

Conclusion: Treatment to target seems effective in pain reduction in non-systemic JIA patients irrespective of initial treatment strategy. Several baseline predictors for pain over time were identified, which could serve as an initial indication for non-systemic JIA-patients with a high risk of pain despite a strict treat-to-target strategy.

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Figure 1 | VAS pain over time. Linear mixed models with random intercept and random slope for visits clustered within patients. VAS pain measured on a 100mm scale. Error bars indicate 95% confidence intervals.

Table 1. Baseline Characteristics

	β	p-value	95% Confidence Interval
VAS Pain	0.45	0.000	0.25 to 0.65
VAS Physician	-0.31	0.014	-0.55 to -0.06
VAS Patient/parent	-0.02	0.873	-0.21 to 0.18
No. of active joints	0.77	0.009	0.19 to 1.34
PhS*	-0.42	0.008	-0.72 to -0.11
PsS**	-0.42	0.022	-0.77 to -0.06
Symptom duration (mo.)	8.09	0.118	-2.08 to 18.26
NSAID use	-1.30	0.791	-10.91 to 8.31

VAS Pain, VAS physician and VAS patient/parent was measures on a 100mm scale.*PhS= Physical Summary Score of the Child Health Questionnaire Parent form 50 (scale 0-100).**PsS= Psychosocial Summary Score of the CHQ-PF50 (scale 0-100).

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POS1301 DRUG SURVIVAL OF BIOLOGICS WITH RESPECT TO COMBINATION WITH METHOTREXATE IN TREATMENT OF POLYARTICULAR JIA

G. Horneff¹, D. Windschall², K. Minden³, T. Hospach⁴, F. Dressler⁵, F. Weller-Heinemann⁶, B. Huegle⁷, I. Foeldvari⁸, A. Klein⁹ on behalf of BIKER registry. ¹Asklepios Clinic, Paediatrics, Sankt Augustin, Germany; ²St. Josef Stift, Paediatric Rheumatology, Sendenhorst, Germany; ³Deutsches Rheuma-Forschungszentrum Berlin (DRFZ), ein Institut der Leibniz-Gemeinschaft, Epidemiology, Berlin, Germany; ⁴Klinikum Stuttgart - Olga Hospital / Women's Hospital, Paediatrics, Stuttgart, Germany; ⁵MHH Kinderklinik Station 61b, Paediatrics, Hannover, Germany; ⁶Prof Hess Childrens Hospital, Paediatrics, Bremen, Germany; ⁷Deutsche Zentrum, Kinder- und Jugendrheumatologie, Garmisch-Partenkirchen, Germany; ⁸Zentrum, Kinderheumatologie, Hamburg, Germany

Background: In polyarticular juvenile idiopathic arthritis (pJIA) biologic therapies are often combined with methotrexate (MTX). This combination was shown to increase efficacy in adult rheumatoid arthritis patients. MTX may also have a protective effect on the formation of anti-drug antibodies and thus may prolong drug survival. In pJIA, there are few and sometimes controversial data available.

Objectives: To compare the effect of combination treatment with MTX on discontinuation due to inefficacy and on drug survival discontinuation of biologics approved for first line treatment of pJIA.

Methods: Patients from the German BIKER registry with their first treatment course with Adalimumab, Etanercept, Golimumab or Tocilizumab were selected. Rates of ineffectiveness-related withdrawal were analysed and compared using χ^2 -test, Wald-test and Kaplan-Meier analysis of patients receiving biologic monotherapy or concomitant methotrexate. Cases were censored if MTX was discontinued before the biologic.

Results: 2173 pJIA patients were identified who for the first time received a biologic. Etanercept (ETA) was by far the most frequently used biologic for first line biologic treatment in pJIA (77%) followed by Adalimumab (ADA, 16%). Patients on Golimumab (GOL) received most frequently a combination with MTX (86.5%), while patients on Tocilizumab (TOC) had the lowest rate of combination treatment (53%).

ETA/ADA/GOL/TOC was given as monotherapy in 500(30%)/89(26%)/5(13.5%)/46(47%) and combined with MTX in 1179 (70%)/259(74%)/32(86.5%)/51(53%)