3. Eta neither prevents nor treats U.
4. Topical NSAIDs and G can hardly be recommended as a reliable means for treatment of U without TNFi and MTX.
5. Systemic NSAIDs neither prevent nor treat JIA-associated U.

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POS1325

COMPARISON OF THREE DIFFERENT ALGORITHMS FOR THE TREATMENT OF CHILDREN WITH POLYARTICULAR JIA: THE FIRST YEAR AFTER DIAGNOSIS

K. Minden^{1,2}, T. Schwarz³, F. Dressler⁴, I. Foeldvari⁵, J. P. Haas⁶, G. Horneff⁷, T. Hospach⁸, J. Kümmerle-Deschner⁹, K. Moenkemoeller¹⁰, F. Weller-Heinemann¹¹, K. Tenbrock¹², M. Niewerth¹³, C. Sengler¹, D. Foell¹³, J. Klotsche¹. ¹Deutsches Rheuma-Forschungszentrum Berlin, Epidemiology, Berlin, Germany; ²Charité Universitätsmedizin Berlin, Paediatric Rheumatology, Berlin, Germany; ³St-Josef-Stift Sendenhorst, Paediatric Rheumatology, Sendenhorst, Germany; ⁴MHH, Kinderklinik, Hannover, Germany; ⁵Hamburg Center for Pediatric and Adolescent Rheumatology, Pediatric and Adolescent Rheumatology, Hamburg, Germany; ⁶Deutsches Zentrum für Kinder- und Jugendrheumatologie, Kinder- und Jugendrheumatologie, Garmisch-Partenkirchen, Germany; ⁷Asklepios Kinderklinik, Paediatric Rheumatology, St. Augustin, Germany; ⁸Olgahospital Stuttgart, Paediatric Rheumatology, Stuttgart, Germany; ⁹Universitätskinderklinik Tübingen, Paediatric Rheumatology, Tübingen; ¹⁰Kinderkrankenhaus Amsterdam StraÙe, Paediatric Rheumatology, Köln, Germany; ¹¹Klinikum Bremen Mitte, Prof. Hess Kinderklinik, Bremen, Germany; ¹²Universitätsklinikum Aachen, Kinder- und Jugendmedizin, Aachen, Germany; ¹³Universitätsklinikum Münster, Klinik für Pädiatrische Rheumatologie und Immunologie, Münster, Germany

Background: Various treatment strategies are used for children with newly diagnosed polyarticular JIA. MTX is usually prescribed, sometimes in combination with high-dose intravenous glucocorticoid pulses (HDGC) or multiple intra-articular GC injections (IAGC). These different approaches were considered in the German consensus-based treatment protocols for polyarticular JIA¹, they were also the leading therapies in patients with rheumatoid factor-negative polyarthritis (RF- PA) included in the JIA inception cohort ICON.

Objectives: To compare the effectiveness of three different treatment strategies in newly DMARD-naïve patients with RF- PA.

Methods: Patients with RF- PA who were included in the ICON cohort and received one of the following treatments within the first three months were considered for the analysis: Group 1: MTX + IAGC in >4 joints, Group 2: MTX + HDGC, Group 3: MTX, no IAGC in >4 joints, no HDGC. Propensity score-adjusted group differences in outcomes after one and two years were analysed by linear and logistic regression analyses.

Results: The analysis included data from 150 patients (79% female, mean age 6.7±4.8 years) enrolled in ICON 1.6±1.9 months after the diagnosis of RF- PA, of whom 52 were in Group 1, 54 in Group 2 and 44 in Group 3. Disease activity did not differ significantly between the groups at treatment start (cJADAS-10 16.7±4.7, 15.8±5.7, 15.9±6.5, respectively).

Of the total group, at 1- and 2-year follow-up (FU), 60.9%/60.1% and 52.3%/58.8% of patients had inactive disease (cJADAS ≤1/Wallace criteria²), 21.3% and 35.6% were in remission off drug², and mean cJADAS-10 scores were 2.6±3.9 and 3.0±3.5, respectively. 60.5% and 67.0% had no functional limitations (CHAQ=0).

Patients in Group 1 more often had an inactive disease (according to Wallace²) at the 1-year FU and tended to have inactive disease more often at 2-year FU than patients in Group 3 (78.1% vs. 45.2%, p=0.025; 73.3 vs. 49.1%, p=0.075, respectively). Group 2 patients (inactive disease in 56.1% and 53.4% at 1- and 2-year FU) did not differ significantly from either Group 1 or Group 3. In addition, Group 1 patients had a significantly better quality of life than patients of Group 2 at the 2-year FU (mean PedsQL 4.0 total score 90.4±9.3 vs. 83.8±11.2, p=0.031). At that time, Group 3 patients had a mean PedsQL 4.0 total score of 85.0±14.6, which was not significantly different from either Group 1 or 2.

On the other hand, Group 1 patients tended to develop new uveitis more frequently within the first two years of treatment than patients in Groups 2 and

3 (13% vs. 2.2% and 3.6%, p=0.101 and 0.131, respectively). At the 2-year FU, patients in Group 1 also had a significantly lower mean height SDS than patients of Group 3 (-0.3±1.1 vs. 0.2±1.1, p=0.038). Mean height SDS was lowest (-0.5±0.8) in patients in Group 2 and significantly lower than in Group 3 (0.019). Mean body mass index SDS also differed significantly between the groups at 2-year follow-up. The mean BMI SDS was highest in Group 1 patients (0.2±0.8), differing significantly from Group 2 (-0.3±0.7, p=0.014) and Group 3 (-0.4±1.1, p=0.023).

There were no significant differences in inactive disease (according to cJADAS) and functional status (CHAQ) between the three groups at 1- and 2-year FU. Over time, treatments were very different in the three groups. In Group 3, biologics were used significantly more often over time than in group 1 (54.0% vs. 18.3%, p=0.014), and Group 2 patients received bDMARDs in 36.1%.

Conclusion: While patients with numerous early joint injections seem to achieve inactive disease more frequently and earlier, they have a slightly smaller body height and tend to develop uveitis slightly more often than patients with more intensive DMARD therapy. However, the differences are small between the groups. Further comparative effectiveness studies with higher patient numbers are needed to identify particularly effective and safe treatment strategies.

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POS1326

FAMILIAL PERIODIC FEVER, APHTHOUS STOMATITIS, PHARYNGITIS AND ADENITIS (PFAPA) SYNDROME; IS IT A SEPARATE DISEASE?

Y. Butbul¹. ¹The Ruth and Bruce Rappaport Faculty of Medicine, Technion - Israel Institute of Technology, Haifa, Israel, Pediatric Rheumatology unit, Haifa, Israel

Background: Periodic fever, aphthous stomatitis, pharyngitis, and adenitis syndrome (PFAPA) is the most common periodic fever syndrome in the pediatric population. Unlike other periodic fever syndromes, the pathogenesis and genetics of PFAPA is unknown. Until recently, PFAPA was believed to be a sporadic disease, yet family clustering has been widely observed and current research indicates that heredity is likely.

Objectives: To identify demographic and clinical differences between patients with PFAPA who have a positive family history (FH+) compared to those with PFAPA with no family history (FH-) that can reveal if heritable and sporadic subtypes of this disorder exist.

Methods: In a database comprising demographic and clinical data of 273 pediatric PFAPA patients treated at two tertiary centers in Israel, 31 (14.3%) of patients were PFAPA FH+. Data from patients with FH+ for PFAPA was compared to data from those with FH- of the disorder. Furthermore, family members (FMs) of those with FH+ were contacted via telephone for more demography and clinical details.

Results: FH+ group had more headaches (32% vs. 2%; p= 0.016), myalgia (56% vs. 19%; p= 0.001), higher carrier frequency of M694V mutation (54% vs. 25%; p=0.053), greater family history of FMF (30% vs. 15%; p=0.096) and better outcomes with colchicine (82% vs. 52%; p=0.096) compared to those with FH-. FMs displayed almost identical characteristics to the FH+ group except for greater arthralgia during flares (64% vs. 23%; p=0.008) and compared to the FH- group, more oral aphthae (68% vs. 43%; p=0.002), myalgia/arthralgia (64% vs. 19%/16%; p<0.0001), and higher rates of FH of FMF (45% vs. 15%; p=0.003).

Conclusion: Our findings suggest that FH+ had probably different subset of disease with higher frequency of family history of FMF arthralgia, myalgia and better response to colchicine. Colchicine prophylaxis for PFAPA should be considered in FH+.

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