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Background: Preliminary definitions for SIJ lesions in the OMERACT Juvenile Idiopathic Arthritis Magnetic Resonance Imaging score has been reported¹. Investigators identified the need to revise the JAMRIS-SIJ item definitions.

Objectives: To update the JAMRIS-SIJ definitions and scoring method.

Methods: The OMERACT JAMRI working group was convened to discuss the performance of the score in a reliability exercise using 30 patients. Twenty investigators (12 radiologists, 8 rheumatologists) decided which definitions and scoring methods to be revised, retained or added.

Results: The revised JAMRIS-SIJ is in the Table 1.

Table 1. Revised OMERACT JAMRIS-SIJ.

Component	Definition	Segmentation/ Score slice	range/ slice
Bone Marrow Edema (BME)	An ill-defined area of high bone marrow signal intensity within the subchondral bone in the ilium or sacrum on fluid sensitive images	4 quadrants/ SIJ	0-8
BME Intensity	Presence of hyperintensity of the marrow on fluid sensitive images using the signal of the presacral veins or cerebrospinal fluid as reference	1 score/SIJ	0-2
BME Depth	Continuing increased signal on fluid sensitive images of depth $\geq 5\text{mm}$ $\geq 1\text{cm}$ from the articular surface using the signal of the presacral veins or cerebrospinal fluid as reference	1 score/SIJ	0-2
Osteitis	An ill-defined area of high bone marrow signal intensity within the subchondral bone in the ilium or sacrum on contrast enhanced T1 weighted sequences	4 quadrants/ SIJ	0-8
Capsulitis	Increased signal on fluid sensitive or contrast enhanced T1 weighted sequences involving the superior portion of the SIJ capsule	superior halves/SIJ	0-2
Joint space fluid	High signal intensity equivalent to the CSF on fluid sensitive sequences within the joint space of the cartilaginous portion of the SIJ	halves/SIJ	0-4
Joint space enhancement	Increased signal intensity on contrast enhanced T1 weighted sequences within the joint space of the cartilaginous portion of the SIJ	halves/SIJ	0-4
Inflammation in erosion cavity	Increased signal intensity on fluid sensitive or contrast enhanced T1 weighted sequences in an erosion cavity of the cartilaginous portion of the SIJ	halves/SIJ	0-4
Enthesitis	Increased signal intensity in bone marrow and/or adjacent soft tissue on fluid sensitive or contrast enhanced T1 weighted sequences at sites where ligaments and tendons attach to a bone excluding retroarticular enthesitis	Score per case	0-1
Damage Domain Sclerosis	A substantially wider than normal area of very low bone marrow signal intensity within the subchondral bone in the ilium or sacrum on a non-fat suppressed sequence, preferably a non-fat suppressed T1 weighted sequence. This feature must also be present on all other sequences, as available	4 quadrants/ SIJ	0-8
Erosion	A focal loss of the low signal of cortical bone at the osteochondral interface and adjacent marrow matrix on T1 weighted images	4 quadrants/ SIJ	0-8
Fat metaplasia lesion	Homogeneous increased signal intensity within the subchondral bone marrow on T1 weighted images	4 quadrants/ SIJ	0-8
Backfill	A high signal on non-contrast enhanced T1 weighted sequences in a typical location for an erosion, with signal intensity greater than normal bone marrow, clearly demarcated from adjacent bone marrow by an irregular band of low signal reflecting sclerosis at the border of the original erosion	halves/SIJ	0-4
Ankylosis	Presence of signal equivalent to regional bone marrow continuously bridging a portion of the joint space between the iliac and sacral bones	halves/SIJ	0-4

Statement of overarching consideration for all definitions: "[...] in comparison to physiological changes normally seen on MRI examinations of age- and sex-matched children, and visible in 2 planes wherever available."

Conclusion: Revised JAMRIS-SIJ has been developed. Validation steps are underway.

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POS1324 TREATMENT OF UVEITIS ASSOCIATED WITH JUVENILE IDIOPATHIC ARTHRITIS: A SEVEN YEARS' SINGLE-CENTRE EXPERIENCE

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Background: Uveitis (U) is one of the most common extra-articular manifestation of juvenile idiopathic arthritis (JIA). It is usually revealed in 10-20% of children with JIA, often asymptomatic and sometimes the onset of U precedes the onset of arthritis. Being inappropriately treated, U leads to vision loss in up to 20% of children. Despite the efforts of the Standardization of Uveitis Nomenclature (SUN) Working Group and the European initiative Single Hub and Access point for pediatric Rheumatology in Europe (SHARE) which optimized management of JIA-associated U [1], the actual treatment is mostly based on physicians' experience. Initial treatment typically includes topical glucocorticoids (G) or systemic immunosuppressive therapy if topical G don't work. Methotrexate (MTX) is the first-line systemic immunosuppressive agent, followed by tumor necrosis factor inhibitors (TNFi) [1,2].

Objectives: To provide treatment recommendations for JIA-associated U based on monitoring of 221 patients during 7 years.

Methods: Data collected during 2240 hospitalisations of 221 patients aged 3 - 17 years treated at the State Pediatric Medical University in years 2014 - 2020 were analyzed. U and JIA were diagnosed according to the SUN and ACR recommendations. Initial treatment of U included topical G or non-steroidal anti-inflammatory drugs (NSAIDs). Treatment of JIA included systemic NSAIDs, cytostatics (MTX, Cyclosporine or Cyclophosphamide) and biologics: Adalimumab (Ada), Infliximab (Inf), Etanercept (Eta), Abatacept (Aba), Tocilizumab (Toc), Golimumab (Gol). Statistical analysis included traditional Descriptive Statistics, Spearman Rank Order Correlation (SROC) and Multivariate Exploratory Techniques [Statistica for Windows, Statsoft].

Results: Of all patients with JIA-associated U the proportion of those treated with biologics during the monitoring increased from 60 to 70% (of them, Ada 79 - 93%, Inf 7 - 1%, Eta 5 - 1%, Aba 7 - 3%, Toc 7 - 0%, Gol 0 - 3%). Of all children in whom U was diagnosed later than arthritis 36% were treated with systemic NSAIDs, 25% - with cytostatics (MTX - 84%, Cyclosporine - 8%, Cyclophosphamide - 8%), 23% - with systemic G, 6% - with biologics (Eta - 83%, Toc - 17%). Of patients in whom U and arthritis were diagnosed simultaneously, 95% were treated with cytostatics (MTX - 95%, Cyclosporine - 15%), 93% - with topical G, 91% - with topical NSAIDs, 81% - with systemic G, 60% - with systemic NSAIDs. Efficacy of treatment in terms of SROC showed evidence of steady improvement already after 1 month of therapy with TNFi or with MTX (increase of r during the first year from 0.37 to 0.62, p<0.02; from 0.37 to 0.55, p<0.05 respectively), though r was never higher than 0.84 during the first 3 years of treatment. Topical NSAIDs and G were less effective: improvement was revealed after 3 months of treatment, it was not so steady and r was never higher than 0.35. Systemic NSAIDs were not effective at all.

Conclusion: 1. The likelihood of improvement of U treated with TNFi (Eta excepted) after one month, after 1 and after 3 years of therapy was 37%, 62% and 84% respectively. For MTX this likelihood was 37% after 1 month and 55% after 3 months.

2. TNFi (Eta excepted) in treatment of JIA prevents the onset of U, thereby supposing its early administration.

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

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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?

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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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