ARTHRITIS IS NOT A PREREQUISITE DISEASE MANIFESTATION FOR THE DIAGNOSIS OF SYSTEMIC JIA: RESULTS OF A PROSPECTIVE COHORT TRIAL USING RIL-1RA AS FIRST LINE TREATMENT WITH LONG TERM FOLLOW-UP

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Background: Systemic onset juvenile idiopathic arthritis (sJIA) is a multifactorial disease, characterised by arthritis, spiking fever, skin rash, lymphadenopathy, hepatosplenomegaly and/or serositis, in combination with increased inflammatory parameters as erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and ferritin. SjIA is nowadays seen as a complex autoinflammatory disorder. However, in the current ILAR classification, sJIA is still classified under the umbrella of JIA. The past decade has learned that the mechanisms underlying the systemic inflammation in sJIA differ in important aspects from the other subtypes like polyarticular JIA.

Objectives: Here we compare disease characteristics, manifestations and responses to treatment in sJIA patients as well as ‘sJIA without arthritis’ (n=12), in order to evaluate whether arthritis should still be a prerequisite for the diagnosis of sJIA.

Methods: We included 30 consecutive diagnosed and prospectively followed new onset sJIA patients as well as 12 ‘sJIA without arthritis’ from our paediatric rheumatology clinic from 2008 until 2017. The ‘sJIA without arthritis’ patients underwent extensive diagnostic procedures to exclude infections (PCR, blood cultures, serology etc), malignancies (bone marrow punctures, PET scans etc) and other diagnoses.

All patients followed a standardised treatment protocol, starting with rIL-1RA (2 mg/kg) as 1st line treatment (without steroids), as previously described.1 In case of partial response, rIL-1RA dose was raised to 4 mg/kg with a maximum of 20 mg/day. If that failed, corticosteroids were added and/or patients switched to alternative biologicals as canakinumab or tocilizumab. If patients had inactive disease at 3 months after start of rIL-1RA treatment, rIL-1RA was tapered for a month (alternate day regimen) and subsequently stopped.

Peripheral blood samples were taken before initiation of rIL-1RA and at all follow-ups for routine lab measurements. For biomarker analyses, serum was isolated at disease onset and at the three months after initiation of rIL-1RA.

Results: There were no differences in disease manifestations like skin rash, serositis, hepatosplenomegaly or symptoms like arthralgic (painful) joint count between sJIA and ‘sJIA without arthritis’ patients at diagnosis. Nor was there a difference in the levels of CRP, ESR, ferritin or IL-18 at start of therapy.

Importantly, also the response to rIL-1RA treatment did not differ between sJIA and ‘sJIA without arthritis’ patients in our cohort. At last follow-up (median 5.8 years, IQR 2.9–7.6 years), 95% of patients had inactive disease, of which 72% off medication.

Conclusions: Based upon disease manifestations and inflammatory parameters in patients with confirmed sJIA and ‘sJIA without arthritis’ at disease onset and on excellent treatment responses to a standardised treatment protocol with rIL-1RA as 1st line treatment, we conclude that arthritis should not be a prerequisite disease criterion in the next classification criteria of sJIA.

REFERENCE:

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AUTOANTIBODIES IN CHILDREN WITH JUVENILE ARTHRITIS ON BIOLOGICAL THERAPY

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Background: Antinuclear antibodies (ANA) are described as initial serological findings in juvenile idiopathic arthritis (JIA). SLE-like syndromes with ANA seroconversion have been described in the patients on biological therapy, including JIA children. Biologics have been used in Ukraine since 2011. In 459 of 2,542 JIA cases registered in 2016, patients were on biologics. According to the data of the Ukrainian Registry of Patients Receiving Biologics, autoimmune diseases as complications of biological therapy have not been registered in Ukraine so far.

Objectives: To study the ANA dynamics in JIA children on biologics.

Methods: There was conducted a retrospective observational study which involved 138 JIA patients, including 44 children with the biologics as follows: TOZ 12, ADA 29, ETA 3. JIA markers (HLA B27 (PGR), RF (ELISA), aCCP (immunosorption).ANA (IFT and ELISA (10 subtypes) were determined in patients at the disease onset and after 1, 2, 5 years of biologics medication.

Results: Among all JIA patients, ANA were detected in 54% of sJIA, 47% of pJIA, 7% of all spondyloarthropathy (spA) cases. At the beginning of biologics, only 22% children were ANA-positive, 66% of them being female, mean age at the disease debut was 6.9±3.81 y. The median of the time from the disease onset to the initiation of biologics was 4.2±3.2 y. The mean age at biologic therapy start was 10.3±4.18 y. At the time of the examination, the mean duration of biologics was 21.7±4.16 y. In pJIA onset ANA were detected in 62% of patients, in spA with uveitis in 75%, in spA in 14.2%, in sJIA ANA were not identified. RF and aCCP were found in 2 pJIA children; HLA B27 was detected in 1 child with uveitis and in 64.2% of patients with spA. The data obtained showed that ANA were found more often in patients on biologics than in general JIA population (pJIA, p=0.04, sJIA, p=0.02), RF was lower (4.5% vs p=0.001). The relationship with the sex was not seen in pJIA, 21.4% of patients had ANA positive RF at the disease debut, but the subsequent dynamic studies did not detect ANA (p=0.04). Patients treated with ETA were observed for less than 1 year.

Conclusions: ANA types which are detected at JIA debut and after initiation of biologic treatment may differ. It is suggestive of the biologics potential to modify the immune response, thus increasing the risk of overlap-syndromes. Therefore, it is advisable to monitor autoantibody titer in JIA children on biologic treatment.

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PREVALENCE OF GENERALISED JOINT HYPERMOBILIT Y IN THE CHILDREN POPULATION OF ORDU: TURKISH STUDY

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Background: Generalised Joint hypermobility (GJH) is a clinical conditions that may cause common musculoskeletal pain during childhood. In our study, we aim to show the prevalence of GJH in children of 11–18 age group, and to provide general information for rheumatologists and paediatricians who are interested in this subject.

Objectives: Our study aimed to evaluate the frequency of GJH in children aged 11–18 years in the province, Ordu.

Methods: This cross-sectional study was performed with 410 students aged 11–18 years who receive education in the province, Ordu. Questionnaire forms were filled in, and each student was examined. The children, who reported to have any disease, were excluded from the study. GJH was diagnosed according to criteria of Beighton diagnosis.

Results: A total of 410 students, 210 of whom were girls (51.3%) and 200 (48.7%) of whom were boys, participated in the study. The subjects’ mean age was 13.7±1.7 years for girls and 13.1±1.79 years for boys. The body mass index (BMI) of the girls was 21.5±4.4 km² and of the boys was 22.3±3.9 km². 160 (39%) of the students participated from the city centre and 250 (61%) from the district centres. The presentations of the students to the health institution due to any complaint in 1 year were examined. The students participating in the study were questioned in terms of presence and time of previous joint complaints. Accordingly, the number of participants who previously had a joint-related complaint was found to be 155 (37.8%), 40 (10.7%) of these participants had a joint-related complaint 3 months ago, 18 (4.3%) had it 6 months ago, 40 (9.7%) had it 1 year ago, and 47 (11.4%) had it more than one year ago. The frequency of GJH was 8.7%. 24 of 36 participants in whom GJH was detected and had a Beighton score of 3 had a joint-related complaint 3 months ago, 18 (4.3%) had it 6 months ago, 40 (9.7%) had it 1 year ago, and 47 (11.4%) had it more than one year ago. The frequency of GJH was 8.7%. 24 of 36 participants in whom GJH was detected and had a Beighton score of 3 had a joint-related complaint 3 months ago, 18 (4.3%) had it 6 months ago, 40 (9.7%) had it 1 year ago, and 47 (11.4%) had it more than one year ago. The frequency of GJH was 8.7%. 24 of 36 participants in whom GJH was detected and had a Beighton score of 3 had a joint-related complaint 3 months ago, 18 (4.3%) had it 6 months ago, 40 (9.7%) had it 1 year ago, and 47 (11.4%) had it more than one year ago. The frequency of GJH was 8.7%.