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

N.M. Terhaar1, A. Leek1, A. El Idrisi1, J.F. Swart1, J. van Loosdregt2, N. Wulffraat3, S. de Roock1, S. Vasten1. 1Pediatric Rheumatology and Immunology; 2Laboratory for Translational Immunology, University Medical Center Utrecht, Utrecht, Netherlands

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 response to treatment between SJIA patients with arthritis and without arthritis’ 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. In case of partial response, rIL-1RA dose was raised to 4 mg/kg with a maximum of 200 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 biweekly intervals.

Results: There were no differences in disease manifestations like skin rash, serositis, hepatosplenomegaly or symptoms like arthralgic (painful) joint count. Interestingly, 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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PREVALENCE OF GENERALISED JOINT HYPERMOBILITY IN THE CHILDREN POPULATION OF ORDU: TURKISH STUDY

Ö.F. Barçak1, S.S. Dedegöl, 2 1Physical Medicine and Rehabilitation, Özel Ünye Çakırtpe Hastanesi, Ordu; 2Orthopedics and Traumatology, Okmeydan Training and Research Hospital, Istanbul, Turkey

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 guiding 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±3.4 kg/m² and of the boys was 22±3.8 kg/m². 180 (43%) 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 in 155 (37.8%), 40 (10.7%) of these participants had a joint-related complaint 3 months ago, 18 (4.3%) had 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 5 and above consisted of girls; and this was 11.4% of the girls. The number of male participants in whom GJH was detected, was found to be 12; and this was 6% of the boys. There was a significant difference between female and male participants in terms of the frequency of GJH (p<0.02). A statistically significant and highly negative correlation was found between body mass index and Beighton score (r=−0.18, p<0.001). A statistically significant and highly negative correlation was found between body mass index and Beighton score (r=−0.092, p<0.05).