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 of ILAR-criteria fulfilling sJIA (n=30) and sJIA without arthritis (n=30) and '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 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 days 30, 90 and 180 after the last rIL-1RA dose. For disease manifestations, patients were asked to fill in a disease activity form at each follow-up visit. The disease manifestations were scored on a scale from 0 (no disease manifestation) to 3 (severe disease manifestation). Disease manifestations and measurements were collected at 3 months an 12 months after start of rIL-1RA treatment.

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 ESR, CRP, ferritin or CRP at 18 months from 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:

Disclosure of Interest: None declared

AB1112 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 (PCR), 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 138 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 onset 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 21.0±4.14 y. In pJIA onset ANA were detected in 62% of patients, in 12% 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%) (p=0.001). The relationship with the sex was not seen (p=0.31). 21.4% of patients had RF positive, RF negative (aCCP neg). There was 1 RF +case at pJIA debut with TOZ, after the therapy it was not detected. Two paediatric patients on TOZ (16.6%) were ANA +at the debut, but the subsequent dynamic studies did not detect ANA (TOZ vs ADA: p<0.04). Patients treated with ETA were observed for less than 1 year. Conclusion: ANA types which are detected at JIA debut and after initiation of biologic therapy 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 titers in JIA children on biologic treatment.

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