THU0577

DO RAYNAUD PHENOMENON NEGATIVE JUVENILE SYSTEMIC SCLERODERMA PATIENTS HAVE A DIFFERENT PATTERN OF ORGAN INVOLVEMENT AS RAYNAUD PHENOMENON POSITIVE PATIENTS?

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Background: Juvenile systemic sclerosis (JSSc) is an orphan disease, with an estimated prevalence of 3 per 1000 000 children. Most JSSc patients primarily present with Raynaud phenomenon (RP). We investigated in our patient of the juvenile scleroderma inception cohort, how fare patients with (RP+) and without (RP-) RP differed in their clinical presentation at enrolment.

Methods: The JSSc is a prospective cohort of JSSc patients. Patients were enrolled who were diagnosed with JSSc, had a JSSc onset age under 16 years and were younger as age of 18 years at the time of inclusion. The patients are prospectively assessed every 6 months according to a standardised protocol. We reviewed the organ involvement pattern of our patients currently followed in the cohort.

Results: 100 patients are currently followed in the cohort and 89 (89%) of them had RP. The female/male ratio was lower in the RP +group, 3.7:1 compared to 4.5:1 (p=0.808). Diffuse subtype was more common in the RP +group, 72% compared to 63%. Mean age of onset of first non-Raynaud symptomatic was 10.4 years in both groups. Mean disease duration was slightly higher in the RP +group, 3.4 compared to 2.2 years. ANA positivity was higher in the RP +group, 88% compared to 70% (p=0.48). Anti-Sc170 was 34% in the RP +and 20% in the RP-group (p=0.34). Interestingly 7% of RP +but none of the RP +were anti-centromere positive. The mean modified skin score was lower in RP +group (mean of 14.8 compared to 17.0). There were significantly more nailfold capillary changes (70% compared to 18%, p=0.001) and a higher rate of history of ulceration in the RP +group (49% compared to 20%, p=0.083). Decreased DLCO and FVC <80% was higher in the RP-negative group with 45%/50% compared to 37.5%/31% respectively. Pulmonary hypertension occurred in 7% in the RP +group and there was no case in the RP- group (p=0.335). RP- group had a higher rate of urinary sediment changes 18% compared to 4.5% in the RP +group (p=0.07). No renal crisis or hypertension was reported in neither groups. Gastrointestinal involvement was similar between the two groups with around 35%. Occurrence of swollen joints was similar in both groups as the frequency of muscle weakness with around 20%. The tendon friction rub occurred around 10% in both groups. In the patient related outcomes, there was only a difference in rating of Raynauds activity.

Conclusions: The RP- group differed from RP +group in the clinical presentation at enrolment. The absence of Raynaud phenomenon was associated with a decreased rate of history of ulceration, no occurrence of pulmonary hypertension. Interestingly higher rate of urinary sedimentary changes and no anticentromere positivity was observed in RP- patients.

Disclosure of Interest: None declared

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THU0578

PATIENTS AND PHYSICIAN RELATED OUTCOMES IMPROVE SIGNIFICANTLY OVER 12 MONTHS FOLLOW UP IN PATIENTS WITH JUVENILE SYSTEMIC SCLEROSIS. RESULTS FROM THE JUVENILE SCLERODERMA INCEPTION COHORT. WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (JSSc) is an orphan disease with an estimated prevalence of around 3 per 1 000 000 children. There are no studies which evaluated prospectively the patient related outcomes in these patients. We report the data from juvenile scleroderma inception cohort (JSSc) regarding organ involvement and patient related outcomes.

Methods: The JSSc is a prospective cohort of JSSc patients. Patients were enrolled who were diagnosed with JSSc, had a JSSc onset age under 16 years and were younger as age of 18 years at the time of inclusion. The patients are prospectively assessed every 6 months according to a standardised protocol. Patients with available 12 months follow up data were included in the analyses.

Results: Currently 100 patients are followed in the JSSc cohort. 51 of them had available 12 months follow up data. Among those patients 37 (72.5%) had diffuse and 14 (27.5%) limited subtype. Mean age of onset of disease was 9.5 (±4.1) years and the mean disease duration at time of inclusion was 3.1 years (±3.2). The proportion of patients treated with DMARD increased from 74.5% to 88% at 12 months follow up. 86% were ANA positive at both assessments. Anti-scl70 positivity increased from 38% to 42%. Anticentromere antibody positivity was 2.4% at both assessments. Mean modified skin score decreased from 17.7 to 14.3 (p=0.151) Raynaud phenomenon occurred in 86% at enrolment and increased up to 88% at 12 months follow up. Nailfold capillary changes occurred around 70% at both assessments, but number of patients with active ulceration decreased from 28% to 16% (p=0.148). The number of patients with decreased FVC (FVC under 80%) decreased from 40.5% to 32% (p=0.497). The number of patients with pulmonary hypertension remained around 10%. No renal crisis or hypertension were reported. The gastrointestinal involvement was around 40% at both assessments. The number of patients with swollen joints decreased from 24% to 10% (p=0.06). The number of patients with muscle weakness decreased significantly from 33% to 9% (p=0.016), parallel to the number of patients with elevated CK values which decreased from 27% to 12% (p=0.074). All patient related outcomes, like global disease activity (p=0.048), global disease damage (p=0.05), Raynaud activity (p=0.003) and ulceration activity (p=0.001) improved significantly over 12 months. Physician assessed global disease activity (p=0.003) and ulceration activity (p=0.001) also improved significantly.

Conclusions: Our data show, that JSSc patients over a 12 months disease course stayed quite stable or improved regarding organ involvement. But patient and physician related outcomes regarding activity assessment improved significantly.

Disclosure of Interest: None declared

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THU0579

VALIDATION OF CONTRAST-ENHANCED MRI SCORES ON (TENOSYNOVITIS OF THE WRIST IN JUVENILE IDIOPATHIC ARTHRITIS)PATIENTS BY COMPARISON WITH CHILDREN UNAFFECTED BY CLINICAL ARTHRITIS

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Background: Delayed and/or inappropriate treatment of juvenile idiopathic arthritis (JIA) may lead to permanent loss of joint functionality.¹ Contrast-enhanced MRI is increasingly being accepted as a sensitive tool for detecting JIA disease activity in an early stage.²

Objectives: The aim of this study was to assess the validity of two reliable contrast-enhanced MRI scores for the assessment of synovitis and tenosynovitis in the wrist of clinically active JIA patients by a comparison with children unaffected by clinical arthritis.

Methods: An axial T1-weighted MRI sequence with contrast-enhancement and fat-saturation was performed on the wrist of 25 children who had no signs of joint inflammation at clinical examination and who were already subjected to contrast-enhanced MR enterography. Wrist MRI scans of 25 clinically active JIA patients were matched based on time-interval between contrast injection and start of the MRI sequence. After being blinded for clinical status, two radiologists scored synovitis and tenosynovitis in consensus. Synovitis was scored at 5 locations by degrees of synovial enhancement (0–2 scale) and synovial inflammation (0–3 scale). Tenosynovitis was scored at the extensor tendons (compartments II, IV and VI) and flexor tendons by degree of inflammation based on a 0–3 scale.^{3,4}

Results: Children unaffected by clinical arthritis had significantly lower total synovial enhancement (median=1 vs 4, p<0.001) and total synovial inflammation (median=1 vs 4, p<0.001) scores compared to clinically active JIA patients (graph). No significant difference in total tenosynovitis score was found between both groups (median=0 vs 0, p=0.220). Fifteen out of 25 (60%) clinically active JIA patients were given a total tenosynovitis score of 0.

Conclusions: The contrast-enhanced MRI scores for the assessment of synovial enhancement and synovial inflammation in the wrist of clinically active JIA patients appear valid. Due to a low incidence of wrist tenosynovitis in this cohort, the validity of the tenosynovitis score could not be assessed. These findings further establish contrast-enhanced MRI as a diagnostic tool with synovitis as the primary target of disease in the wrist of JIA patients.

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THU0580

THE ACR RECOMMENDATIONS FOR JIA IN DAILY CLINICAL PRACTICE: ARE THEY FOLLOWED OR WOULD TREAT-TO-TARGET THERAPY LEAD TO BETTER RESULTS?

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Background: What factors drive the physician decision to escalate to anti-TNF therapy 3 and 6 months after start of methotrexate (MTX) in both persisting oligoarthritis (OJA) and polyarticular course (PJIA) juvenile idiopathic arthritis.

Objectives: Are the escalation-decisions in accordance with the ACR JIA treatment recommendations (ACR-CPG)¹ and if not, what factors drive these decisions. How does it perform as a "prognostic" test to predict failure when not escalated. Could the clinical Juvenile Arthritis Disease Activity Score (cJADAS) be used instead. What is the value of the patient-VAS in the ACR-CPG, the physician decision and in the cJADAS.

Methods: Monocentric retrospective cohort study analysing all OJA and PJIA patients starting MTX for the first time between 2011 and 2016.

Results: The ACR-CPG is mostly not followed and implementation would increase the anti-TNF-use from 12.0% to 65.1%. However, the physician decision not to escalate was now correct in 70%–75%, therefore implementation results in an overuse of anti-TNF. Some items of the ACR-CPG were non-discriminatory. The use of cJADAS in predicting failure if not escalated outperformed the ACR-CPG with a much higher sensitivity and specificity for the OJA and PJIA group respectively. The omission of the patient-VAS-scores resulted in a substantial decrease of the identification of patients failing to respond without escalation.

Conclusions: The ACR-CPG not only is too complicated to be applicable in clinical practice, it also fails to identify those patients really in need of escalation to

anti-TNF. The cJADAS can be used instead since this is user-friendly, does not require waiting for ESR results and performs better than the ACR-CPG. The patient-VAS is a critical item for the decision to escalate.

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THU0581

USE OF BIOLOGICAL THERAPIES IN ADULT PATIENTS DIAGNOSED WITH JUVENILE IDIOPATHIC ARTHRITIS: RESULTS FROM BIOBADASER, THE SPANISH REGISTRY OF ADVERSE EVENTS WITH BIOLOGIC THERAPIES

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Background: Juvenile idiopathic arthritis (JIA) is the most frequent chronic rheumatic disease in childhood. The early disease recognition and treatment is critical to prevent long-term complications and disability in childhood. During the last decade the arrivals of biologics has dramatically changed the prognosis of these patients. A number of well-designed clinical trials, as well as cohort studies have demonstrated that biologics are an effective option for JIA patients who do not respond or cannot tolerate treatment with synthetic disease modifying drugs (DMARDs).

JIA is not confined to childhood, and a 41% had active disease are on medication after 30 years and 28% had a high symptom state.

Objectives: The aim of this work was to study the pattern of use, drug survival and adverse events of biologic therapy in JIA patients during the transition period from the diagnostic to the adulthood.

Methods: Information was obtained from BIOBADASER, a safety multicenter prospective registry. All patients included in the registry diagnosed of JIA between 2000 and 2015 were analysed. Proportions, means and standard deviations (SD) were used to describe population. Incidence rates and 95% confidence intervals were calculated to assess adverse events. Kaplan-Meier analysis was used to compare the drug survival.

Results: 469 patients, 46.1% women were included in this study. Age at diagnosis was 9.4 (SD=5.3) and years of disease evolution 24.1 (SD=14.1). The age at biological treatment initiation was 23.9 years (SD=13.9). The pattern of use of biologics in JIA patients in the paediatric age shows a linear increase from 24% in 2000 to 65% in 2014. Interestingly, the biologic suspension for disease remission was higher in patients who initiated its use under 16 years (25.7%) than in those who began at 16 years or later (7.9%, $p<0.0001$). Serious adverse events showed a total incidence rate of 41.4 (35.2–48.7) (1000 patients/year) without differences between patients younger or older than 16 years old. However, patients younger than 16 years old showed a significant increment in infection and infection ($p<0.001$).

Conclusions: The biologic survival and suspension by remission was higher when the biologic therapy started before 16 years old in JIA patients. The incidence rate severe adverse events in the childhood and adulthood in JIA patients treated with biologics was similar, however, a significant increment of infection was observed in patients under 16 years old.

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