Results: A total of 3975 courses of biologics with a total exposure of 7592 PY were identified with Etanercept (5376PY) followed by Abalumumab (1334PY), Tocilizumab (435PY), Abatacept (109PY), Infliximab (99PY), Anakinra (96 PY), Canakinumab (71PY) and Golimumab (67PY). Differences in JIA category distribution and concomitant treatment were noted. A total of 3586 AE (47.2/100PY), 461 (6.1) SAE and 629 (8.3) AEs were reported. The most common AEs were uveitis (194 (2.6)) followed by medically important infections (155 (2.0)), cytopenias (62 (0.8)), hepatic events (39 (0.5)), aphasia (28 (0.4)), other autoimmunopathies (25 (0.3)), chronic inflammatory bowel disease (23 (0.3)), depression (17 (0.2)), macrophage activation syndrome (12 (0.2)), malignancies (8 (0.1)) and pregnancies (8 (0.1)). There were marked differences in the rate of AEs with different biologics. Uveitis were most common in TNF-antibody treated cohorts, infections upon GOL, TOC, ANA, cytopenias upon TOC, CAN, hepatic events upon TOC, anaphylaxis upon INF, TOC, CED upon ETA, INF (table 1). One case of latent TB but no further opportunistic infections were reported. There was a single death due to sepsis.

Conclusions: Surveillance of pharmacotherapy as provided by BiKER is an import approach especially for long term treatment of children. Overall, tolerance is acceptable. Differences between several biologics were noted and should be considered in daily patient care.

Disclosure of Interest: None declared


THU0575 CARDIOVASCULAR RISK IN LONG-TERM JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile Idiopathic Arthritis (JIA) is one of the more common chronic diseases of childhood that often persists into adulthood and can result in significant long-term morbidity, including physical disability. The long-term risk of cardiovascular disease for individuals with Juvenile Idiopathic Arthritis (JIA) remains uncertain.

Objectives: This study aims to determine whether adults with JIA in remission and medium-long duration of the disease have an increased risk of cardiovascular disease.

Methods: This is a cross-sectional study including 25 patients (14 females and 11 males) diagnosed with JIA according to the International League of Associations for Rheumatology criteria ILAR 2001 were compared to 20 age- and sex-matched controls. Remission was determined by JADAS27 <1 and according to Wallase criteria. An extensive clinical analysis including body index mass, lipic profile, HOMA-IR and intra-arterial blood pressure was performed. Intima media thickness of the common carotid artery (CIMT) was measured as a marker of subclinical atherosclerosis. Different proinflammatory cytokines (TNFa, IL1b and IL6), molecules involved in the endothelium dysfunction (VEGF and E-Selectin) and adikopines (resistin and visfatin) were analysed on serum by ELISA.

Results: Mean duration of the disease was 13.31±1.14 years. Mean age was 27.21±0.68. Time in remission was 3.52±0.84 years. Metabolic comorbiddities such as obesity and metabolic syndrome were more prevalent in our cohort of JIA patients compared to controls. Levels of cholesterol were significantly elevated in patients. However, HOMA-IR values and intra-arterial pressure were not significant increased in JIA patients. CIMT was higher in JIA patients compared to controls (0.44±0.009 vs 0.41±0.017, p=0.078), although it did not reach the statistical significance. Serum levels of cytokines (including TNFa, IL6 and IL1b), molecules involved in the endothelium dysfunction (VEGF and E-Selectin) and adikopines (resistin and visfatin) were analysed on serum by ELISA.

Results: Mean duration of the disease was 13.31±1.14 years. Mean age was 27.21±0.68. Time in remission was 3.52±0.84 years. Metabolic comorbiddities such as obesity and metabolic syndrome were more prevalent in our cohort of JIA patients compared to controls. Levels of cholesterol were significantly elevated in patients. However, HOMA-IR values and intra-arterial pressure were not significant increased in JIA patients. CIMT was higher in JIA patients compared to controls (0.44±0.009 vs 0.41±0.017, p=0.078), although it did not reach the statistical significance. Serum levels of cytokines (including TNFa, IL6 and IL1b), molecules involved in the endothelium dysfunction (VEGF and E-Selectin) and adikopines (resistin and visfatin) were analysed on serum by ELISA.

Conclusions: In our cohort of JIA patients the increased CIMT was not associ- ated with inflammatory markers, but disease duration. Although patients were in clinical remission, the serum levels of inflammatory cytokines, adikopines and VEGF were significantly elevated, molecules with a relevant role in the onset and progression of endothelial dysfunction and atherosclerosis. These results might suggest that long-term JIA patients could have higher cardiovascular risk, although they are in sustained remission.

Disclosure of Interest: None declared


THU0575 ANAKINRA FOR FIRST LINE STEROID FREE TREATMENT IN SYSTEMIC ONSET JUVENILE IDIOPATHIC ARTHRITIS

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Background: Systemic juvenile idiopathic arthritis (sJIA) is characterised by arthritis accompanied or preceded by systemic autoinflammation. High-dose steroids has been the mainstay of therapy with proven effectiveness but also with side-effects. In many patients a chronic course with destructive arthritis long-term cannot be prevented.

Objectives: In patients naïve for steroids, a steroid-free treatment may allow reconstitution of an impaired NK-cell function and probably remission of sJIA.

Methods: First experience with first line Anakinra without steroids in 9 consecu-tive patients is reported.

Results: All patients presented with ongoing spiking fever and rash and further features of sJIA, high CRP, S100 and IL18 (table 1). Daily s. injections of Anakinra 2 mg/kg for 3 months resulted in complete remission in 4 and partial response in 2 children presenting with an oligaarticular involvement. One patient with typical sJIA and very high S100 (MRPs/9) levels did not respond to Anakinra nor to Canakinumab. Two patients presented with polyarthitis. One had no response, the other showed a minor response but improved on steroids and was later treated with tocilizumab. One of the oligaarticular patients with an initial partial response had a flare upon anakinra captured by increased dosing (4 mg/kg) but finally developed macrophage activation syndrome. Anakinra was discontinued.

Response in two children presenting with an oligaarticular involvement. One patient with typical sJIA and very high S100 (MRPs/9) levels did not respond to Anakinra nor to Canakinumab. Two patients presented with polyarthitis. One had no response, the other showed a minor response but improved on steroids and was later treated with tocilizumab. One of the oligaarticular patients with an initial partial response had a flare upon anakinra captured by increased dosing (4 mg/kg) but finally developed macrophage activation syndrome. Anakinra was discontinued.

Conclusions: Experience with first line steroid free treatment for Anakinra for sJIA is presented. A complete remission was reached in 4 cases with oligaarticular involvement. In 3 further cases improvement was observed and 2 had no response including one who also failed Canakinumab. A toddler with a particular response to Anakinra later on developed MAS. One patient did not respond to both IL-1 inhibitors. Thus, steroid free treatment regimen with Anakinra is feasible and resulted into remission in most but not all patients. Aside, unwarranted effects of long lasting steroid application were avoided.

Disclosure of Interest: None declared


Table 1: Initial response and Outcome

<table>
<thead>
<tr>
<th>Age</th>
<th>Clinical Features</th>
<th>CRP</th>
<th>S100</th>
<th>IL18</th>
<th>Initial Response</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>11, m</td>
<td>spiking fever, rash, arthralgia</td>
<td>95.6</td>
<td>1910</td>
<td>2483</td>
<td>complete response</td>
<td>remission off treatment</td>
</tr>
<tr>
<td>2, m</td>
<td>spiking fever, rash, arthralgia</td>
<td>91.7</td>
<td>14 110</td>
<td>&gt;10 000</td>
<td>partial response</td>
<td>increase of dosage upon flare, discontinued with MAS</td>
</tr>
<tr>
<td>1, f</td>
<td>spiking fever, rash, arthralgia</td>
<td>158.3</td>
<td>820</td>
<td>&gt;5000</td>
<td>complete response</td>
<td>remission off treatment</td>
</tr>
<tr>
<td>1, f</td>
<td>spiking fever, rash, arthralgia</td>
<td>168.3</td>
<td>820</td>
<td>&gt;5000</td>
<td>partial response</td>
<td>remission off treatment</td>
</tr>
<tr>
<td>8, f</td>
<td>polyarthitis</td>
<td>224.5</td>
<td>4040</td>
<td>&gt;10 000</td>
<td>no response</td>
<td>remission upon Tocilizumab</td>
</tr>
<tr>
<td>15, f</td>
<td>polyarthitis</td>
<td>326.8</td>
<td>4780</td>
<td>&gt;10 000</td>
<td>partial response</td>
<td>remission off treatment</td>
</tr>
<tr>
<td>5, m</td>
<td>spiking fever, rash, arthralgia</td>
<td>20.5</td>
<td>22 650</td>
<td>3820</td>
<td>minor response</td>
<td>improvement with corticosteroids, remission on Tocilizumab</td>
</tr>
<tr>
<td>11, w</td>
<td>spiking fever, rash, arthralgia</td>
<td>185.8</td>
<td>20 775</td>
<td>6000</td>
<td>complete response</td>
<td>remission off treatment</td>
</tr>
<tr>
<td>4, m</td>
<td>spiking fever, rash, arthralgia</td>
<td>203.8</td>
<td>3 04 850</td>
<td>&gt;10 000</td>
<td>no response</td>
<td>switch to Canakinumab with no response</td>
</tr>
<tr>
<td>5, m</td>
<td>spiking fever, rash, arthralgia</td>
<td>237.7</td>
<td>1 61 700</td>
<td>2830</td>
<td>complete response, switch to Canakinumab after 7 days</td>
<td>remission off treatment</td>
</tr>
</tbody>
</table>
Juvenile systemic scleroderma (jSSc) is an orphan disease, with an estimated prevalence of around 3 per 1,000,000 children. Most jSSc patients primarily present with Raynaud phenomenon (RP). We investigated in our patient of the juvenile scleroderma inception cohort, how far 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-Scl70 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=0.085). Decreased DLOCO and FVC <80% was higher in the RP− 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 sedimentary 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 skin ulceration, no occurrence of pulmonary hypertension, interestingly higher rate of urinary sedimentary changes and no anticentrome 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**

**Background:** Juvenile systemic scleroderma (JSSc) is an orphan disease with an estimated prevalence of around 3 per 1 000 000 children. There are no studies which prospectively followed 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 was reported. The gastrointestinal involvement was around 40% at both assessments. The number of patients with active ulceration 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) 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

**DOI:** 10.1136/annrheumdis-2018-eular.2913

**THU0579**

**VALIDATION OF CONTRAST-ENHANCED MRI SCORES ON (TENO)SYNOVITIS 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. Contrast-enhanced MRI is increasingly being accepted as a sensitive tool for detecting JIA disease activity in early stage.

**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.

**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.