Background: Juvenile psoriatic arthritis (JPsA) is one of the clinical variants of juvenile idiopathic arthritis (JIA), which is often characterized by an unfavorable course, refractory to therapy, requiring the prescription of biological agents (BA).

Objectives: Analysis of BA use in patients with JPsA and therapy survival, switching to another line of BA.

Methods: The retrospective cohort study included 1095 JIA patients who received BA and were observed in our clinic from 2004 to 2019. All cases of new onset psoriasis were collected; clinical features of disease onset and course, exposure to Methotrexate (MTX) and BA, presence of ANA, HLA B27 were studied.

Results: Among 1095 JIA patients who received BA over the past 15 years, a separate cohort of patients with JPsA for analysis was allocated. We identified 50 pts (57% female) aged 2-18 years (Me 13.3) at the time of initiation of therapy. All patients met the JPsA classification criteria, the average age of arthritis onset was 7.4±5.3 years (ME 6.75). However, cutaneous psoriasis occurred only in 68% (34 pts), with manifestation at the age of 10±5 years. In 25 of 34 pts (73.5%) the development of psoriasis was preceded by joint manifestations in an average of 5.3±9 (ME 3) years. 6 pts from 1095 (0.65%) developed psoriasis under BA therapy: infliximab - 2 cases (0.62/100PY), adalimumab - 3 (0.15/100PY), abatacept -1 (0.31/100PY). 2/6 pts were ANA+, 3/6 – HLA B27+.

Average age of disease onset was 9.8±7.8 years; BA exposure before psoriasis was 2.7±1.1 years and in 3.6±3.4 (ME 4) after the onset of arthritis. Therapy was continued in 4/6 pts; switched from infliximab to adalimumab in 2. Serositis and joint involvement was associated with psoriasis in 7 pts (1 type 1 diabetes mellitus – 2 pts; Down syndrome; endogenous mental illness (schizophrenia); oligophrenia; ovaries polycystic; acute lymphoblastic leukemia in a state of incomplete remission). The clinical picture of the disease was represented by polytrauma in 84%, oligoarthritis in 8%, the same number of patients 8% had an ankle lesion. Sacroiliitis was detected in 20 patients (40%), dactylitis in 21 (42%), and uveitis in 10 (20%). HLA B27 was detected in 16/35 pts (45%), 32% were ANA-positive. The duration of the disease at the time of application of the first BA was 5±4 (ME 3.75) years. In 49 patients, BA was used in combination with methotrexate. The total number of BA doses switching included was 80 (infliximab-19, adalimumab-22, etanercept-7, golimumab-4, abatacept-5, tocilizumab-2, rituximab-1). 49% of patients have experience of using ≥2 BA (16 pts-2 BA, 4 pts-3 BA, 1 pt - 5 BA). Primary/secondary inefficacy (18/35; 51%), adverse events (8/35; 23%), organizational difficulties in market access mostly after the age of 18 (7/35; 20%), and remission (2/35; 6%) were the reasons for the withdrawal of BA. Among the serious adverse events, multiple sclerosis was registered after 6 years of abatacept (the relationship with the drug used has not been proven), pregnancy in the 3rd year of adalimumab use (interruption at 16 weeks); serious local reaction after etanercept using-1; infusion reactions (1-rituximab, 2-infliximab); uveitis de novo (2- etanercept).

Conclusion: JPsA is one of the most severe variants of JIA, characterized by a high proportion of serositis conditions, the development of refractory joint manifestations of BA therapy and beginning uveitis, multiple sclerosis, requiring switching to line 2 and with a limited choice of BA with pediatric indications. Special study requires the manifestation of psoriasis de novo mainly developed during TNF-monoclonal antibodies therapy.

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THU0515 SYSTEMIC LUPUS ERYTHEMATOSUS IN CHILDHOOD AND ADOLESCENCE - UPDATE FROM THE NATIONAL PEDIATRIC RHEUMATOLOGY DATABASE

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Background: Systemic lupus erythematosus (SLE) is a clinically heterogeneous disease, which begins in childhood and adolescence in 15 - 20% of cases. Since 2004, data on SLE have been collected by means of a disease-specific questionnaire as part of the National pediatric rheumatology database (NPRD) in Germany. Since 2018, many SLE biopsy results have been recorded to further specify kidney involvement.

Objectives: Evaluation of clinical signs and symptoms, outcome and laboratory data of patients with juvenile systemic lupus erythematosus from a large database in Germany.

Methods: Data from patients with SLE recorded in the NPRD in 2017 were consecutively analyzed. In addition to age, sex, onset of disease, the criteria that led to the diagnosis, various laboratory parameters, organ involvement (current, even and therapy (current, last 12 months), current disease activity (numerical rating scale 0-10, NRS) and ECLAM (score 0-10) were recorded. Patient-reported outcomes included global assessments of overall-wellbeing and fatigue (NRS 0-10) and functional ability (CHAQ).

Results: 196 patients (86% female) with a median age of 16 years were documented. Criteria most frequently met at diagnosis included “antinuclear antibodies” (88%), followed by “anti-ds-DNA-Ab” (66%), “butterfly erythema” (42%) and “arthritis” (41%). A positive family history was found in 10% of patients. At documentation, 85% of patients received disease-modifying anti-rheumatic drugs, most frequently hydroxychloroquine (73%), followed by mycophenolate mofetil (32%) and azathioprine (17%). Systemic glucocorticoids obtained 52% of patients, 12% ± 0.2 mg/kg/ day. Biologics (rituximab 2%) and cyclophosphamide i.v. (3%) were rarely administered during the last 12 months. Disease activity was reported as 1.0 (NRS, median, IQR 0 - 9), ECLAM as 10 (median, range 0 - 10). In the laboratory, leucopenia < 3500/µl was found in 9% of patients, lymphopenia < 1500/µl in 47% and erythrocyte sedimentation rate (ESR) > 25 mm in 15% of patients. Mean CHAQ was 0.24, and 86% of patients had a CHAQ score < 0.5. Mean patient’s global assessment of overall-wellbeing was 1.5, while the mean fatigue score was 2.86 (18% NRS score 7-10).

Conclusion: The most common clinical symptoms documented in juvenile SLE patients were skin and joint involvement. In the course of the disease, a quarter of the patients developed kidney involvement, mostly proliferative nephritis. Apparently, azathioprine is increasingly being replaced by mycophenolate mofetil, biologics have hardly been used so far. Although functional outcome and overall-wellbeing is for SLE patients was good, fatigue was a concern for some patients.

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THU0516 FIFTEEN CASES OF 3 NLR FAMILY MEMBERS (NLRP3, NLRP12 AND NLRC4) RELATED INFLAMMASOMOPATHIES IN A SINGLE CENTER OF CHINA

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Background: There are four members in NLR family, NLRP3, NLRC4, NLRP1 and NLRP12, the mutations of which can lead to autoinflammatory diseases, while little reports describe those diseases in Chinese population.

Objectives: To report several cases of NLR-related autoinflammatory diseases in our center and to compare the differences of the presentations of CAPS between Chinese and western patients.

Methods: This study was undertaken at Peking Union Medical College Hospital (PUMCH) between 2012 and 2019. Demographic data, clinical presentations and genetic results were collected.

Results: 15 patients had been diagnosed as NLR-related autoinflammatory diseases in our center, including 11 CAPS, 1 FACS4 and 3 NLRP12-AD patients. We found 10 NLRP3 mutations, 3 NLRP12 mutations and 1 NLRC4 mutation. There are 3 novel mutations: NLRP3 c.1311G>T, NLRP3 c.1711G>A, and NLRC4 c.514G>A.

The major symptoms of those diseases are similar, such as recurrent episodes of fever associated with rash. And some may suffer from arthritis/arthritis, uveitis, sensorineural deafness, symptoms of central neural systems (CNS). On the other hand, different inflammasomopathies have unique characteristics. Symptoms of FACS1, the mildest CAPS disorder, including rash and fever with/without arthritis/arthritis, usually develop in the first year of life. The onset age of MWS is later (8m to 5y), and those patients were more likely to develop arthritis/arthritis, eye involvement, hearing loss and symptoms of CNS. NOMID was the most severe type, and was presented with chronic urticarial-like rash shortly after birth, as well as severe CNS manifestations and musculoskeletal involvement. One of our NOMID patients had clubbing fingers, which was not reported before. The onset age of NLRP12-AD ranges from 6m to 5y, and the presentations are similar to MWS while the FACS4 patient presented with rash and fever, like FACS1.
Cáceres, A. Rodriguez. Scientific Abstracts

ESR or CRP. Most of those patients had increased cytokines, including IL-1, IL-6 as well as TNF-α. Leukocytosis and thrombocytosis were also observed in most patients, while anemia was mostly found in patients diagnosed as NOMID. We also compared the clinical manifestations of CAPS between Chinese and western patients. The frequency of fever in Chinese is much higher than that in western population, while less Chinese patients suffered from ocular manifestations. Besides, Chinese patients seem to exhibit higher frequencies of severe symptoms, either CNS symptoms, or musculoskeletal symptoms, albeit with insignificant difference.

<table>
<thead>
<tr>
<th></th>
<th>Chinese</th>
<th>Western</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>n</td>
<td>26</td>
<td>136</td>
<td></td>
</tr>
<tr>
<td>Gender Ratio</td>
<td>15:11</td>
<td>69:67</td>
<td>0.518</td>
</tr>
<tr>
<td>Fever</td>
<td>25/26 (96%)</td>
<td>106 (79%)</td>
<td>0.002</td>
</tr>
<tr>
<td>Rash</td>
<td>24/26 (92%)</td>
<td>132 (97%)</td>
<td>0.397</td>
</tr>
<tr>
<td>Triggered by cold</td>
<td>3/26 (12%)</td>
<td>34 (25%)</td>
<td>0.076</td>
</tr>
<tr>
<td>Ocularmanifestations</td>
<td>10/26 (38%)</td>
<td>97 (71%)</td>
<td>0.001</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>3/26 (35%)</td>
<td>56 (41%)</td>
<td>0.535</td>
</tr>
<tr>
<td>Neurological symptoms</td>
<td>15/26 (58%)</td>
<td>55 (40%)</td>
<td>0.105</td>
</tr>
<tr>
<td>Severe</td>
<td>4/11 (36%)</td>
<td>16 (12%)</td>
<td>0.141</td>
</tr>
<tr>
<td>Musculoskeletalmanifestations</td>
<td>18/26 (69%)</td>
<td>117 (86%)</td>
<td>0.093</td>
</tr>
<tr>
<td>Increased CRP/ESR</td>
<td>25/26 (96%)</td>
<td>90 (69%)</td>
<td>0.137</td>
</tr>
</tbody>
</table>

Conclusion: We reported a case series of NLR-related autoinflammatory disease and found some novel mutated alleles and clinical phenotypes, which expanded our knowledge to those diseases. By comparing clinical manifestations of CAPS patients in China and in western countries, it seems that the symptoms in different populations are not identical.

References:

Acknowledgments: We'd like to thank the patients as well as their parents for their participation.

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THU0517

The role of Ultrasound-defined Synovitis Grade I in Patients presenting with Inflammatory Arthralgia, a Retrospective Study

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Background: Musculoskeletal ultrasound (MSUS) is frequently used in several rheumatology units to detect subclinical inflammation in patients with joint symptoms suspected for progression to inflammatory arthritis (IA). Synovitis grade I (EULAR-OMERACT combined score) is known to be a casual finding in healthy individuals, but studies headed to unravel its possible role on rheumatic diseases are sparse.

Objectives: To investigate the correlation between synovitis grade I, and the diagnosis of IA made after a year follow-up period since MSUS findings, in patients of an MSUS-specialized unit of a Rheumatology Department.

Methods: We conducted a descriptive, retrospective and unicentric study. 30 patients were selected from the MSUS-specialized unit of our Rheumatology Department from July-18 to January-19. Patients presenting synovitis grade 0 (exclusively), 2 and/or 3 on combined score were excluded. Data collection at baseline included age, sex, immunological profile and previous physical examination to the MSUS findings, as well as the diagnosis made by the rheumatologist in 1-year visit follow-up: dividing the patient sample into two groups: those who were diagnosed with IA and those not. Non-parametric statistical tests for comparing means were used.

Results: The mean age was 51.6 years and 70% were females. 6 (20%) patients were diagnosed with inflammatory arthritis after a year follow-up: 2 (4.8%) psoriatic arthritis, 1 (3.3%) undifferentiated arthritis, 1 (3.3%) rheumatoid arthritis, 1 (3.3%) Sjögren’s syndrome. Non-inflammatory arthropathies were also found 24 (80%), of which, 12 (40%) were non-specific arthralgias and 8 (19%) osteoarthritis.

In the group of patients who did not develop an IA the mean of CRP was 9.27 mg/L and 14.17 mm respectively; 2 (33%) patients were RF-positive, and 1 ACPA-positive. ANAs were positive in 3 cases (50%). The median of swollen joint count was 2 and for painful joint count was 0, the median of joints with MSUS involvement was 4.5. The mean of MSUS involvement was for MCP,PIP and DIP joints: 1.67, 2 and 0. Comparing the means of CRP values in the two groups with Student’s t-test we obtained a statistically significant difference (p=0.023). No other significant differences were found.

Conclusion: Despite the limitations and possible statistical bias, the presence of MSUS-defined synovitis grade I and elevated CRP levels could be related to further diagnoses of inflammatory arthropathy. Besides, the absence of synovitis in DIP joints might have a diagnostic role. Normal physical exploration and normal levels of CRP might suggest low MSUS value. However, further research is needed to clarify the role of MSUS-defined synovitis grade I.

Disclosure of Interests: None declared

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THU0518

Accuracy of Dual-energy Computed Tomography for the Detection of Bone Marrow Edema in Patients with Axial Spondyloarthritis

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Background: An important applications of the Dual energy computed tomography (DECT) in the field of musculoskeletal radiology is the detection of bone marrow edema (BME), using a post-processing software to remove calcium in trabecular bone by using a “virtual non-calcium (VNCa)” subtraction process. DECT have been successfully employed in the evaluation of the extent of BME in patients with sacroiliitis.

Objectives: The aims of this study were to evaluate the discriminating capacity of DECT versus MRI in the detection of BME in sacroiliac joints in patients with axial-SPA and to define the optimal cutoff; to define of inter-observer agreement between radiologists.

Methods: All patients underwent a pelvic DECT examination, within 30 days of magnetic resonance imaging. Diagnostic criteria were: bone marrow signal is depicted in green and corresponding to high signal intensity on T2-weighted fat suppression MRI images (Fig. 1). With the consent of the two operators, three ROIs were manually positioned for each side of the sacroiliac joints in the subchondral region of

Disclosure of Interests: None declared

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