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 was 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 ± 1.3 (ME 4) after the onset of arthritis. Therapy was continued in 4/6 pts; switched from infliximab to adalimumab in 2. Serious comorbid pathology was associated with JPsA in 7 pts (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 polyarthritis in 84%, oligoarthritis in 8%, the same number of patients 8% had an axial 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% pts 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 courses switching included was 80 (infliximab-19, adalimumab-22, etanercept-27, 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 inefficiency (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 withdrawn of BA. Among the serious adverse events, multiple sclerosis was registered after 6 years of abatacept using (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 serious comorbid conditions, the development of refractoriness and adverse events of BA therapy (including uveitis, multiple sclerosis), requiring switching to line 2 and 3 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.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6366

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 2014, kidney 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 considered for the analysis. In addition to age, sex, onset of disease, the criteria that led to the diagnosis, various laboratory parameters, organ involvement (current,

ever) 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, $12\% \geq 0.2 \text{ mg/kg/day}$. 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 1.0 (median, range 0 - 10). In the laboratory, leukopenia $< 3500/\mu\text{l}$ was found in 9% of patients, lymphopenia $< 1500/\mu\text{l}$ in 47% and erythrocyte sedimentation rate (ESR) $> 25 \text{ mm}$ 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).

The following organ involvement was ever present: general symptoms 84%, skin/mucosa 72%, joints 73%, thyroid 15%, muscle 25%, lungs 17% and CNS 30%. In 45/190 (24%) patients, a kidney involvement was stated. In 34 patients (75%) a kidney biopsy was performed and histology yielded the following results: Class 1: 6.7%, Class 2: 16.7%, Class 3: 40.0%, Class 4: 23.3%, Class 5: 13.3%.

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 of jSLE patients was good, fatigue was a concern for some patients.

Disclosure of Interests: Claudia Sengler: None declared, Martina Niewerth: None declared, Nils Geisemeyer: None declared, Hermann Girschick: None declared, Ariane Klein Consultant of: Celgene, Annette Friederike Jansson: None declared, Markus Hufnagel: None declared, Kirsten Minden Consultant of: GlaxoSmithKline, Sanofi, Speakers bureau: Roche

DOI: 10.1136/annrheumdis-2020-eular.3494

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 FCAS4 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 FCAS1, 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 presentation is similar to MWS while the FCAS4 patient presented with rash and fever, like FCAS1.