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

C. Sengler¹, M. Niewerth¹, N. Geisemeyer¹, H. Girschick², A. Klein³, A. F. Jansson⁴, M. Hufnagel⁵, K. Minden¹. ¹German Rheumatism Research Center, Berlin, Germany; ²Vivantes Klinikum im Friedrichshain, Berlin, Germany; ³Asklepios Klinik St. Augustin, St. Augustin, Germany; ⁴Dr. von Hauner'sches Kinderspital, München, Germany; ⁵University of Freiburg, Pediatrics, Freiburg, Germany

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

W. Wang¹, Y. Zhou¹, H. Song¹. ¹Peking Union Medical College Hospital, Chinese Academy of Medical Sciences, Department of Pediatrics, Beijing, China

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.

For laboratory examinations, all patients had raised inflammatory markers like ESR or CRP. Most of those patients had increased cytokines, including IL-1 β , 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.

	Chinese	Western	p value
n	26	136	
Gender Ratio	15:11	69:67	0.518
Fever	25/26 (96%)	108 (79%)	0.002
Rash	24/26 (92%)	132 (97%)	0.397
Triggered by cold	3/26 (12%)	34 (25%)	0.076
Ocular manifestations	10/26 (38%)	97 (71%)	0.001
Hearing loss	9/26 (35%)	56 (41%)	0.535
Neurological symptoms	15/26 (58%)	55 (40%)	0.105
Severe	4/11 (36%)	16 (12%)	0.141
Musculoskeletal manifestations	18/26 (69%)	117 (86%)	0.093
Severe	3/11 (27%)	6 (4%)	0.137
Increased CRP/ESR	25/26 (96%)	NA	

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:

- [1] Levy R, Gerard L, Kuemmerle-Deschner J, Lachmann HJ, Kone-Paut I, Cantarini L et al. Phenotypic and genotypic characteristics of cryopyrin-associated periodic syndrome: a series of 136 patients from the Eurofever Registry. *Ann Rheum Dis*. 2015;74(11):2043-9.
- [2] Li C, Tan X, Zhang J, Li S, Mo W, Han T et al. Gene mutations and clinical phenotypes in 15 Chinese children with cryopyrin-associated periodic syndrome (CAPS). *Sci China Life Sci*. 2017;60(12):1436-44.

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

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3053

THURSDAY, 04 JUNE 2020

Diagnostics and imaging procedures

THU0517

THE ROLE OF ULTRASOUND-DEFINED SYNOVITIS GRADE I IN PATIENTS PRESENTING WITH INFLAMMATORY ARTHRALGIA, A RETROSPECTIVE STUDY

J. Arroyo Palomo¹, M. Arce Benavente², C. Pijoan Moratalla¹, B. A. Blanco Cáceres¹, A. Rodríguez¹. ¹Hospital Ramón y Cajal, Madrid, Spain; ²Hospital Clínico Universitario Lozano Blesa, Zaragoza, Spain

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 developed an IA the mean C-reactive protein (CPR) value was 3,12mg/L and erythrocyte sedimentation rate (ESR) was 8,2mm; all of them were rheumatoid factor (RF) positive and ACPA-negative except one patient. 5 (31,3%) patients presented low antinuclear antibodies (ANAs) levels. In those who HLA B-27 and Cw6 were tested (4,25%); both were negative except for one that was HLA B-27 positive. The median number of swollen and painful joint count was 0, and the mean of joints with MSUS involvement was 3,5; the mean involved metacarpophalangeal (MCP) joints was 1,83; proximal interphalangeal (PIP) joints was 1,48 and distal interphalangeal (DIP) joints 0,21.

Among the group of patients that developed an IA the mean of CPR and ESR was 9,27mg/L and 14,17mm 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 CPR 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.

References:

- [1] D'Agostino MA et al. Scoring ultrasound synovitis in rheumatoid arthritis: a EULAR-OMERACT ultrasound taskforce-Part 1: definition and development of a standardized, consensus-based scoring system. *RMD Open*. 2017;3(1):e000428.
- [2] Van den Berg R et al. What is the value of musculoskeletal ultrasound in patients presenting with arthralgia to predict inflammatory arthritis development? A systematic literature review. *Arthritis Research & Therapy* (2018) 20:228.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4886

THU0518

ACCURACY OF DUAL-ENERGY COMPUTED TOMOGRAPHY FOR THE DETECTION OF BONE MARROW EDEMA IN PATIENTS WITH AXIAL SPONDYLOARTHRITIS

M. Carotti¹, F. Salaffi², P. Piccinni¹, M. Di Carlo², S. Farah², D. Benfaremo³, M. M. Luchetti³, A. Giovagnoni¹. ¹Ospedale Riuniti Torrette Di Ancona, Department of Radiology, Università Politecnica delle Marche, Torrette, Ancona, Italy; ²Hospital "Carlo Urbani", Dipartimento Scienze Cliniche e Molecolari, Università Politecnica delle Marche, Jesi, Italy; ³Clinica Medica, Dipartimento Scienze Cliniche e Molecolari, Università Politecnica delle Marche, Torrette, Ancona, Italy

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 (1)

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

Methods: All patients underwent a pelvic DECT examination, within 30 days of the MRI imaging, (Somatom Force; Siemens Healthineers, Enlangen, Germany). Each exam was evaluated by two operators: an experienced radiologist and a radiologist in training. The dedicated software also allows the precise calculation of the attenuation values in the region of interest (ROI). On the reformatted color-coded dual-energy virtual non-calcium images bone marrow signal is depicted in green and corresponding to high signal intensity on T2-weighted fat suppression MR 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