fever, 29 unexplained autoinflammatory recurrent fevers, 8 TNF receptor associated periodic syndrome (TRAPS), 8 other autoinflammatory diseases. 68 patients (53%) were treated by on-demand corticosteroids, nine patients (0.7%) by biological agents, the other patients did not receive any immunosuppressive treatment.

At the last clinic visit, only 49% (n=63) of our entire cohort had a complete vaccination status (human papilloma virus and tuberculosis excluded). Coverage rates was superior to 85% for diphtheria/tetanus/poliomyelitis (89%), pertussis (95%), *haemophilus influenza* type B (94%) and measles/mumps/rubella (89%). 84% of the children were well-vaccinated against pneumococcus. Lowest individual vaccination coverage rates were observed for hepatitis B at 77% and for meningococcus C at 77%. Only one girl received complete vaccination against human papilloma virus.

Conclusion: Vaccination coverage in children with autoinflammatory diseases appears suboptimal. Measures to optimize vaccination coverage in these children should be implemented.

REFERENCES:

- [1] Jaeger VK, et al. Safety of vaccinations in patients with cryopyrin-associated periodic syndromes: a prospective registry based study. *Rheumatology (Oxford)*. 2017 Sep 1;56(9):1484-1491
- [2] Jayaratnam J. et al. The safety of live-attenuated vaccines in patients using IL-1 or IL-6 blockade: an international survey. *Pediatr Rheumatol Online J*. 2018 Mar 21;16(1):19

Disclosure of Interests: Virginie Rollet-Cohen: None declared, Justine Mir-ete: None declared, Glory Dingulu: None declared, Andreas Wörner: None declared, Marie-Aliette Dommergues Grant/research support from: Pfizer, MSD, GSK (travel grant for medical congress), Consultant for: Pfizer, MSD, GSK (scientific board participation), Speakers bureau: Pfizer, MSD, GSK, Véronique Hentgen Consultant for: SOBI, Novartis, Abbvie, Speakers bureau: Novartis

DOI: 10.1136/annrheumdis-2019-eular.6878

THU0552

SCORING ULTRASOUND SYNOVITIS IN JUVENILE IDIOPATHIC ARTHRITIS: RESULTS OF A RELIABILITY EXERCISE USING OMERACT US PAEDIATRIC DEFINITIONS

Linda Rossi-Semerano¹, Sylvain Breton², Marouane Boubaya³, Haykanush Ohanyan³, Valerie Devauchelle-Pensec⁴, Sandrine Jousse-Joulin⁴, JIRECHO study group: M. Bossert, S. Boiu, E. Chatelus, G. Durand, S. Jean, L. Goumy, A. Mathiot, G. Mouterde, F. Nuges, A. Ould Hennia, B. Rey, A. Von Scheven, L. Sparsa. ¹Bicêtre hospital, ¹Department of Paediatric Rheumatology, Reference centre for Auto-inflammatory diseases, Le Kremlin Bicêtre, France; ²Hopital Necker Enfants Malades, Department of Rheumatology, Paris, France; ³Avicenne hospital, Clinical Research Unit and Clinical Research Center, Bobigny, France; ⁴Brest Medical University Hospital, INSERM ESPRI, ERI29, UBO, Department of rheumatology, Brest, France

Background: Despite the increasing use of musculoskeletal ultrasound (MSKUS) in assessing joints in juvenile idiopathic arthritis (JIA) in daily practice, there is still a lack of data on its reliability in the evaluation of synovitis in children. Lately, the OMERACT ultrasound paediatric task force has developed preliminary definitions and scoring system for US synovitis in children (1).

Objectives: To evaluate the reliability of the OMERACT paediatric definitions and scoring system of synovitis through a web reading images exercise.

Methods: Thirteen sonographers with at least 2 years' experience in paediatric MSKUS JIA were invited to participate in the reliability exercise. After a training session, participants were asked to analyze 75 images for the presence/absence of elementary lesions and for the grading of synovitis (2), synovial hypertrophy, effusion and Doppler signal. US images of elbow, wrist, second MCP, knee and ankle were collected using standardized scanning by 2 sonographers experienced (LR and SB), from JIA patients with different age in different disease stages. The intra-reader reliability (evaluated 4 weeks after the first assessment) was calculated by the Cohen's kappa coefficient and by the quadratic weighted kappa and the inter-reader reliability by the Light's kappa.

Results: The intra-reader reliability was good for the detection of the presence/absence of any elementary lesion (median Cohen's kappa 0.62, range 0.47-0.75), and for scoring synovitis and synovial hypertrophy and almost perfect for the scoring of Doppler signal (median quadratic weighted kappa 0.77, 0.66-0.86, 0.76, 0.61-0.84 and 0.87, 0.77-0.94 respectively), while it was moderate and rather variable for scoring effusion (0.55, 0.24-0.76). The inter-reader reliability was good for scoring

synovitis and synovial hypertrophy (Light's kappa 0.68, CI 95% 0.61-0.75 and 0.63, CI 95% 0.54-0.71 respectively), almost perfect for scoring Doppler signal (0.85, CI 95% 0.77-0.90) and moderate for the detection of any elementary lesion and for scoring effusion (0.48, CI 95% 0.36-0.64 and 0.49, CI 95% 0.40-0.60 respectively). We found a trend for better results in the evaluation of all parameters in older children (9-16 vs 0-8 years : light's kappa 0.53 vs 0.44 for any lesion, 0.71 vs 0.64 for synovitis, 0.53 vs 0.43 for effusion, 0.71 vs 0.64 for synovial hypertrophy and 0.78 vs 0.90 for Doppler). Concerning the five joints evaluated the best results were obtained for the knee for all parameters (median Lights kappa 0.71, range 0.54-0.85) excepted for Doppler whose reliability was higher for second MCP (Lights' kappa 0.87).

Conclusion: Excepted for effusion, whose evaluation is limited on static images, the reliability of the OMERACT paediatric US definitions and scoring system for synovitis in JIA was good among a large group of sonographers. These results suggest their potential applicability in clinical practice and in multicenter studies.

REFERENCE:

- [1] Preliminary definitions for the sonographic features of synovitis in children. Roth J, Ravagnani V, Backhaus M, et al. *Arthritis Care Res (Hoboken)*. 2017 Aug;69(8):1217-1223.

Disclosure of Interests: Linda Rossi-Semerano Grant/research support from: Roche, Sylvain Breton: None declared, Marouane Boubaya: None declared, Haykanush Ohanyan: None declared, Valerie Devauchelle-Pensec Grant/research support from: Roche-Chugai, Speakers bureau: MSD, BMS, UCB, Roche, Sandrine Jousse-Joulin: None declared

DOI: 10.1136/annrheumdis-2019-eular.5689

THU0553

THE IMPACT OF BACKGROUND BIOLOGICAL THERAPY ON SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS-ASSOCIATED MACROPHAGE ACTIVATION SYNDROME: A SINGLE CENTER STUDY

Riccardo Russo¹, Maria Katsikas². ¹Garrahan Pediatric Hospital, Immunology/Rheumatology, Buenos Aires, Argentina; ²Garrahan Pediatric Hospital, Immunology/Rheumatology, Buenos Aires, Argentina

Background: Macrophage activation syndrome (MAS) is a severe complication of Systemic juvenile idiopathic arthritis (SJIA). Biological therapy of SJIA may modify the clinical expression of MAS (1).

Objectives: to assess and compare the clinical, biochemical, and haematological features of SJIA-associated MAS in patients treated with biologic (B) or non-biologic (NB) agents.

Methods: retrospective analysis of data from dedicated medical records and databases. Inclusion criteria included: diagnosis of SJIA according to the ILAR criteria, diagnosis of MAS according to the expert's opinion, and prescription of corresponding therapy. Patients were followed and treated at a tertiary center from 2006 to 2018. Demographical, clinical, haematological, biochemical and treatment-related features were recorded. The worst recorded values (e.g. highest ferritin levels) during each MAS episode were included for analysis. Outcomes (complete or partial recovery, death) and fulfillment of current classification criteria (2) were also recorded. Comparisons between groups (patients developing MAS during B therapy vs. NB therapy) were performed by chi square or Wilcoxon rank sum tests.

Results: Twenty-four (14 F, 10 M) patients who developed 31 MAS episodes were included. Age at SJIA onset 4 years, age at MAS onset 6 years; 10 episodes occurred during the initial (0-3 months) stage of SJIA course. Patients were receiving biological agents (canakinumab 4, tocilizumab 4, anakinra 1, etanercept 1) in 10 (32%) MAS episodes. Frequency of cardinal clinical features was similar in both groups. During MAS course, ferritin (1,408 vs 26,574 ng/ml, p < 0.001), LDH (922 vs 3308 IU/L, p = 0.04), SGOT (66.5 vs 389 IU/L, p = 0.02), and triglycerides levels (259 vs 352 IU/L, p = 0.03) were lower in B than in NB treated patients, while platelet count (215 vs 66 n/μl, p < 0.01), and hemoglobin (104 vs 79 g/dL, p < 0.01) were higher in the B than in the NB treated patients. No significant differences were found in GGT, fibrinogen, SGPT levels or neutrophil count. Duration of admission during MAS tended to be longer in the NB treated group (31 days) than in the B group (16 days). Treatment of MAS included high dose steroids, cyclosporin, and/or biological agents. Five patients, all belonging to the B group (p < 0.001), did not fulfill current SJIA-MAS classification criteria. There were 4 deaths recorded, all belonging to the NB group.

Conclusion: SJIA patients treated with biological agents exhibit similar clinical features than those treated with non-biologic therapy. However,