patients, including intraoperative fractures, periprosthetic joint infections and wound dehiscence. The majority of patients were treated with biologic therapy before surgery.

Conclusion: Prosthesis implantation in JIA patients is a complicated and difficult procedure, related to the management of the biologic therapy, the low quality of bone, the remarkable stiffness and deformity of the joints. Long-term results were good, even in patients with severe arthritis, showing drastic reduction of joint pain and an improvement of functionality. The risk of complications results to be modest compared to a great survival rate and a valid recovery of joint range of motion. The interdisciplinary team composed by rheumatologists and orthopedic surgeons for the all follow-up of these patients is a cornerstone to obtain the perfect schedule for prosthetic implantation.

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THU0550 MYCOPHENOLATE MOFETIL (MMF) IN DEFINED AND UNDEFINED INTERFERONOPATHIES

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Background: Type I interferonopathies are genetic disorders characterized by an up-regulation of type I interferon (IFN) activity. An increased expression of type I IFN regulated genes, IFN signature (IS), is described in these conditions. They are characterized by autoinflammation and varying degrees of autoimmunity or immunodeficiency. Some patients with a phenotype strongly suggestive for type I interferonopathy with a high IS, do not show any mutations in known type I interferonopathies related genes, being classified as undefined

Objectives: To evaluate the effect of mycophenolate mofetil (MMF) in patients with defined and undefined type I interferonopathy

Methods: 7 patients with type I interferonopathy followed at a Pediatric Rheumatology center, were included. Leucocyte (WBC) and platelet count, hemoglobin (Hb), CRP, ESR, serum amyloid A (SAA), autoantibodies, complement levels and IS, were assessed before and after 5-7 months of MMF treatment (600 mg/m² BID). IS was determined by qPCR (high scores >1.42)

Results: Patient 1 and 2 presented respectively with SAVI and Aicardi-Goutieres syndrome. The others were classified as undefined. In patient 1 (table), treated with JAK1/2 inhibitor, MMF was followed by decrease of inflammatory markers and resolution of lung involvement; in patient 2 to an increase in Hb and normalization of inflammatory markers. Among patients with an undefined phenotype the addition of MMF to low dose prednisone led to complete resolution of inflammation. A reduction of the antinuclear antibodies titre was observed in patients 2, 3 and 7 and a normalization of complement level in patients 3, 4 and 7. Patient 5 had normal inflammatory markers already before the beginning of MMF possibly due to previously administered immunosuppressive treatment. Despite all patients presented a significant improvement of clinical picture after 5 months of MMF treatment, the IS decreased only in 4/7 patients while it remain stable or increased in the others

	PI 1	PI 2	PI 3	PI 4	PI 5	PI 6	PI 7
Disease	SAVI (lung and skin involvement, systemic inflammation)	Aicardi Goutieres (neurological involvement, rash, fever)	Undefined (epilepsy, arthritis, skin involvement)	Undefined (recurrent fever, hypertransaminasemia, hemophagocytic lymphohistiocytosis)	Undefined (hemiplegia)	Undefined (epilepsy, arthritis, skin involvement)	Undefined (recurrent fever, lymphadenopathy, hypogammaglobulinemia)
Clinical improvement after MMF	Resolution of lung involvement	Resolution of fever and rash	Resolution of rash, arthritis, improvement of seizures	Resolution of fever, inflammation and hypertransaminasemia	Improvement of motility	Resolution of arthritis and skin involvement	Resolution of fever and lymphadenopathy
Gender	F	F	M	M	M	F	M
Age at onset (yrs)	3 (days)	2	4	7	4	6	16
WBC/mm ³	16/17.4	8,0/8.4	9,8/8.5	1,7/8.4	4,4/8.2	4,4/5.2	6,6/5.9
Hb g/dl	7,7/8.9	8,5/12.3	12,9/12.8	9,7/13	12,1/10.7	12/12.9	11,5/15.1
PLT/mm ³	950/766	680/320	194/181	56/161	358/421	381/390	218/323
ESR (mm)	44/40	49/9	38/10	7/6	2/5	24/8	3/3
CRP (mg/dl)	1,22/0.73	3,13/0.06	5,55/0.32	<0.05/<0.05	<0.05/<0.05	1,15/<0.05	8,34/0.98
SAA (mg/dl)	95,8/3.62	5,37/2.82	188/8.11	<1.89	<0.84/	4,19/0.78	/
C3	154/134	122/108	81/92	83/118	203/170	127/108	53/133

Abstract THU0550 – Figure 1

Conclusion: Our data suggest that mycophenolate mofetil might be an effective therapy in patients with type I interferonopathy leading to improvement of clinical and laboratory features thus allowing a glucocorticoid sparing

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THU0551 VACCINATION COVERAGE IN FRENCH CHILDREN WITH AUTO-INFLAMMATORY DISEASES : PRELIMINARY DATA FROM THE JIRCOHORTE

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Background: Autoinflammatory diseases (AID) comprise a large number of rare diseases that manifest as recurrent sterile inflammation or disproportionate infection-induced inflammatory symptoms. Vaccine relation to AID is complex: on one hand the susceptibility to infections vary with the treatment options of the patient, on the other hand the disease activity may be triggered by vaccination. Nevertheless, effective immunization is crucial in this population. Up to now very little is known about vaccination coverage in AID children.

Objectives: To determine vaccination coverage in French children with an autoinflammatory disease.

Methods: Patients between 2 and 18 years with autoinflammatory diseases followed at the French Reference Center for Auto-Inflammatory Diseases-Versailles Hospital and included to the Juvenile Inflammatory Rheumatism cohort (JIRcohort) network - an international multicenter prospective cohort for children with inflammatory and rheumatological diseases - were included in this retrospective monocentric observational study. Vaccination coverage of each disease and complete status of vaccination was assessed at the last out-patient visit, according to the national immunization program.

Detail of every vaccine received (number of doses and date of administration) were collected by a pediatrician from written immunization record (vaccination card). Demographic data, disease and medications used since diagnosis were also assessed. Patients with no available immunization record were excluded.

Results: 128 patients met the inclusion criteria. Sex ratio was 1.2 (70/58). Median age at the last out-patient visit was 8.4 years (2.5-17.9 years). 53 children (41%) presented periodic fever with aphthous stomatitis, pharyngitis and adenitis (PFAPA) syndrome, 30 familial mediterranean

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.

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THU0552

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

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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.

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THU0553

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

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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,