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.

REFERENCES:

- [1] C. Mertelsmann-Voss et al. US Trends in Rates of Arthroplasty for Inflammatory Arthritis Including Rheumatoid Arthritis, Juvenile Idiopathic Arthritis, and Spondyloarthritis. *Arthritis & Rheumatology*, June 2014.
- [2] M. P. Abdel et al. Surgical Management of the Juvenile Idiopathic Arthritis Patient with Multiple Joint Involvement. *Orthopedic Clinics of North America*, 2014-10-01.

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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