history, demographic characteristics, and previous DMARDs treatments. Adverse events(AEs), reason for the discontinuation of the biologic treatment, or switch to another biological treatment were documented. The study was approved by the Institutional Review Board. Written informed consent was obtained from parents of the patients and the study was conducted in accordance with the Declaration of Helsinki.

Results: In this study 64, 18-year-old Caucasian JIA patients were evaluated from the day of entrance into the registry, who had JADAS 71 results at age 18, from the 347 patients treated with biologics at the submission.

Demographic data and disease related variables by descriptive statistics:
Gender: Female: N(%): 49(77%).
Subtypes of JIA: Extended oligoarticular form: 11 patients, Polyarticular form: 53 patients (5 RF positive,48 negative)
Median age: 8 (range:4-17) years.
Mean disease duration: 3352 days.
Mean time on biological treatment:1030 days.
The first JADAS 71 summation at the entry into the registry: Median: 18,65 (range:14,4-55,4).
The age 18 JADAS71 summation: Median:3,75 (range: 2,1-38).
All patients were under biologic treatment (etanercept: 56, adalimumab: 29, tocilizumab: 12, abatacept: 4 patients).
Disease activity states and responses to treatments:

JIA extended oligoarticular subtype	Cutoffs for disease activity states JADAS10/71	Number of patients at the entry into the study	Number of patients at age 18
Inactive disease	≤1		1
Low disease activity	1.1 –2	1	
Moderate disease activity	2.1 –4.2		2
High disease activity	>4.2	10	8
JIA Polyarticular RF (negative+positive) subtype			
Inactive disease	≤1		10
Low disease activity	1.1 –3.8	3	20
Moderate disease activity	3.9 –10.5	3	9
High disease activity	>10.5	47	14

Conclusion: It appears from our study that at age 18, 72,3% JIA patients with extended oligoarticular subtype and 26,4% with polyarticular subtypes report a JADAS71 high disease activity sum, so, these young adults will need transitional care.

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AB1034 NO DIFFERENCES IN HEPATITIS B ANTIBODY POSITIVITY IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS ON BIOLOGICAL THERAPY

CİCEK SUMEYRA OZDEMİR¹, Nihal Sahin¹, Aysenur Pac Kisaarslan¹, Meda Kondolot², Zubeyde Gunduz³, Muammer Hakan Poyrazoglu¹, Ruhan Düşünsel¹. ¹Erciyes University Faculty of Medicine, Pediatric Rheumatology, Kayseri, Turkey; ²Erciyes University Faculty of Medicine, Social Pediatrics, Kayseri, Turkey; ³Acibadem Hospital, Pediatric Rheumatology, Kayseri, Turkey

Background: Biologic disease-modifying antirheumatic drugs (DMARDs) are frequently used in the treatment of juvenile idiopathic arthritis (JIA) resistant to nonbiologic DMARDs recently. There is a concern for their reducing the effectiveness of vaccines due to immunosuppressive effects.

Objectives: To evaluate hepatitis B antibody (anti-HBs) positivity and antibody titers between JIA patients on biologic DMARDs therapy and healthy controls.

Methods: Anti-HBs antibody positivity and titers were compared between 88 patients with JIA on biologic DMARDs therapy followed in our clinic and 91 healthy controls aged 2-19 years. All participants were vaccinated according to the routine vaccination schedule in our country.

Results: Eighty-eight JIA patients included and fifty five (62,5%) patients were female. The median age of patients was 13,31 (4,07-18,08) years. The JIA subtypes were systemic JIA (10,2%), oligoarticular JIA (33%), polyarticular JIA (25%), entesitis related arthritis (26,1%), psoriatic JIA

(3,4%), and undifferentiated JIA (2,3%). The median duration of biologic DMARDs was 31,995 (1,33-115) months. 54 patients were treated only with one anti-TNF (34 etanercept, 17 adalimumab), 16 patients were treated more than one TNF-alfa inhibitor. 18 patients were treated with more than one different group biologic DMARDs, and 7 of them had more than two biologic DMARD due to disease activity. Anti-HBs positivity was 67% in the JIA group and 54,9% in the control group, and no significant difference detected between the two groups (p=0,097). The anti-HBs titer was 12,95 (min-max: 2-1000) in the controls and 22,44 (2-1000) in the biological DMARDs.

Conclusion: Patients with JIA on biologic DMARDs had not lower anti-HBs positivity than healthy controls in our study. Further studies are required to understand the effects of biologic DMARDs on protective anti-HBs antibody levels.

Disclosure of Interests: None declared
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AB1035 MAFB-VARIANTS IN MULTICENTRIC CARPOTARSAL OSTEOLYSIS WITH NEPHROPATHY DO NOT SEEM TO AFFECT SERUM C1Q CONCENTRATION

Riccardo Papa¹, Annalisa Madeo², Stefano Volpi¹, Roberta Caorsi¹, Giancarlo Barbano³, Marina Botto⁴, Belinda Campos-Xavier⁵, Andrea Superti-Furga⁵, Marco Gattomo¹, Maja DI Rocco², Paolo Picco¹. ¹IRCCS Istituto Giannina Gaslini, Clinica Pediatrica e Reumatologia, Genova, Italy; ²IRCCS Istituto Giannina Gaslini, Malattie Rare, Genova, Italy; ³IRCCS Istituto Giannina Gaslini, Nefrologia e Trapianto di Rene, Genova, Italy; ⁴Imperial College London, Faculty of Medicine, London, United Kingdom; ⁵Centre Hospitalier Universitaire Vaudois, Division of Genetic Medicine, Lausanne, Switzerland

Background: Multicentric carpotarsal osteolysis syndrome (MCTO)-associated progressive nephropathy is an orphan disease associated with well-known structural kidney damage that usually requires kidney transplantation in adulthood. Although ACE-inhibitors could potentially slow down the progression of kidney dysfunction, other treatments would be welcome. Furthermore, a slight proteinuria reduction in a patient with MCTO treated with cyclosporine has been reported. MafB-deficient mice develop systemic lupus erythematosus-like autoimmune disease by inhibited C1q production and efferocytosis. In the human, monoallelic mutations in MafB result in MCTO.

Objectives: To investigate whether C1q levels in patients with MCTO are reduced and might contribute to the development of renal disease.

Methods: In three individuals with clinical and radiographic signs of MCTO, the MAFB gene was sequenced. The serum C1q concentration was determined with a commercial laboratory kit.

Results: Three patients with MCTO were enrolled (table 1). Patient A presented recurrent urinary tract infections since 18 months of life for a third-degree vesicoureteral reflux. Despite correction surgery, he displayed proteinuria after one year, and kidney biopsy did not reveal significant lesions. After two years, he displayed bilateral tarsal malformations requiring orthopedic surgery. At the age of 6, a pathological fracture of the right wrist occurred, and local X-ray revealed osteolysis of carpal bones. - Because of a suspected juvenile idiopathic arthritis of the right wrist, patient B was treated with intra-articular steroid injection and subcutaneous methotrexate for one year without benefit. After one year, he developed proteinuria and kidney biopsy revealed focal segmental glomerular lesions with tubular fibrosis and renal vasa sub-intimal delamination. C3 and C4 positive mesangial deposits were also present. - Patient C was referred to us for suspected juvenile idiopathic arthritis of the right wrist. No inflammatory signs were present at the ultrasound examination. Sequencing of the MAFB gene revealed in all the patients a novel heterozygous mutation. Although specific variants have not been reported before, the nature and position of the variants associated with the clinical features of the patients strongly argued in favor of their pathogenic role. The serum C1q concentration was normal in all the patients.

Abstract AB1035 Table 1. Main features of three patients with MCTO.

Patient	A	B	C
Age (years); gender; country	14; M; Croatia	16; M; Italy	11; F; Russia
MAFB mutation	c.184A>G, p.Thr62Ala	c.172A>G, p. Thr58Ala	c.208T>C, p. Ser70Pro
Bone lesions onset (age; years)	5	8	8
Presentation Site	Bilateral Tarsus	Monolateral Carpus	Monolateral Carpus
Current state (except carpotarsal joints)	Elbows and knees contractures	Knees contractures	None