AB1071 **AUTO-IMMUNE AND INFLAMMATORY DISEASES IN CHILDREN WITH SICKLE CELL DISEASE: DIAGNOSTIC AND THERAPEUTIC ISSUES**

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Background: Coexistent auto-immune and inflammatory diseases (AIID) and sickle cell disease (SCD) have been recently described in adults and children, however its frequency and physiopathology remain unclear (1–6).
Objectives: The aim of this study is the analysis of clinical and biological characteristics at AIID diagnosis and the evolution under treatment in children with SCD
Methods: Between May 1991 and March 2018, 35 of 3,800 SCD children diagnosed with AID in seven hospitals in Paris and suburb were analyzed in a retrospective survey.
Results: Thirty-five patients reported 44 AIID: auto-immune liver disease (AILD, n=13), inflammatory bowel disease (IBD, n=7), juvenile idiopathic arthritis (JIA, n=6), systemic lupus erythematosus (n=5), autoimmune hemolytic anemia (n=3), Sjögren's syndrome (n=2), histiocytic necrotizing lymphadenitis (n=2), vasculitis (n=2), myasthenia gravis (n=2), sarcoidosis (n=2), inflammatory uveitis (n=1), sclerodermia/juvenile dermatomyositis (n=1). Median age at diagnosis was 10 [2 – 18] years. The mean delay between first symptom and diagnosis was 15.5 (± 29) months. The time of diagnostic was significantly longer for patients with JIA compared to other AID (63 versus 10 days, p=0.004). Sixteen patients (45.7%) had hypergammaglobulinemia > 20 g/L at diagnosis. AILD had a hypergammaglobulinemia at the time of diagnosis (30.0g/L), with a statically significant decrease at the end of follow-up (18.2g/L, p=0,0048). Among 21 patients (60%) treated with systemic steroids, it triggered vaso-occlusive crisis in 14 (66.7%), one acute chest syndrome, one transient ischemic attack. Thirteen of 35 patients (37.1%) were managed with biotherapy for AIID, well tolerated. Three patients (8.6%) underwent stem cell transplantation, one died of a cortico-resistant and multipolar graft versus host reaction, two were cured of both AIID and SCD. Nine severe infections were reported, four under steroids, five under biotherapy.
Conclusion: Diagnosis and therapeutic care of coexistent auto-immune and inflammatory diseases are difficult and challenging in children with SCD. Annual monitoring of inflammatory markers could be recommended to detect AIID earlier and prevent diagnostic delay in case of high ascension in SCD patients.

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AB1072 **UVEITIS IN PEDIATRIC RHEUMATOLOGY: TERTIARY CENTER EXPERIENCE IN TURKEY**

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Background: Since pediatric uveitis is generally asymptomatic, the diagnosis and treatment may be mostly delayed. Severe complications and visual loss may be observed even at the initial visit. Pediatric uveitis is tend to be chronic, persistent, recurrent, and the management may be complex (1).
Objectives: The aim of this study is to report epidemiology, etiology, clinical features, management and the outcomes of non infectious pediatric uveitis at a tertiary pediatric rheumatology center in Turkey.
Methods: The clinical records of the patients with non infectious uveitis who were followed up by department of pediatric rheumatology and ophtalmology were reviewed, from January 2013 to June 2018, retrospectively. The inclusion criteria were as follows being age ≤ 16 years, following up at least 6 months in both the ophtalmology and pediatric rheumatology clinics. Uveitis was categorized anatomically according to the Standardization of Uveitis Nomenclature criteria (2).
Results: Of 37 patients (67 eyes), 45,9% were female. Mean age of onset was 8, 5 ± 4, 4 years (1,6 - 15,6), mean follow-up was 60 ± 42 months (6 - 191). The general features of uveitis were anterior, idiopathic and bilateral in this study similar to literature (Table 1).The most common systemic diseases associated with uveitis were juvenile idiopathic arthritis (JIA).Two patients improved with local medications, while the remaining 35 patients required systemic treatments such as short-time (oral/iv) corticosteroids (CS) in 94.5% of them, methotrexate (MTX) in 86.4%, azathioprine (AZA) in 5.4%, adalimumab (ADA) in 67.5%, tocilizumab (TCB) in 2.7%. In 26.1% of patients receiving ADA who did not respond to standard dose of ADA, we had to shorten the dosage intervals of ADA from every 2 weeks to every week. At least 1 ocular complication was observed in 83.7% of the patients, such as cataract, glaucoma, band keratopathy, synechiae, macular edema and retinal detachment. Four (10.8%) patients had moderate visual loss and 6 (16.2%) patients severe visual loss (3). The prevalence of surgery in our study was 18.9% for cataract and glaucoma treatment.
Conclusion: Diagnosis and management of uveitis in childhood is complicated. Despite the new medication options, the advancements in diagnosis and surgical techniques, the complications are still high. Usage of shorter dose interval of ADA may be an alternative to control of the disease in patients with unresponsive to standard dosage of ADA. However large-scale clinical trials are required to assess the efficacy and safety of this treatment.

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Abstract AB1072 Table 1. Clinical features of patients according to the anatomical classification of uveitis.

		Anterioruveitis 21 cases (56.8%)	Intermediateuveitis 12 cases (32.4%)	Posterioruveitis 1 case (2.7%)	Panuveitis 3 cases (8.1%)	Total 37 cases (100%)
Gender	**Female**	9(24.3%)	6 (16.2%)	1(2.7%)	1(2.7%)	17(45.9%)
	Male	12(32.4%)	6(16.2%)	0	2 (5.4%)	20 (54%)
Ocular involvement	**Unilateral**	3 (8.1%)	3(8.1%)	0	1(2.7%)	7 (18.9%)
	Bilateral	18 (48.6%)	9 (24.3%)	1(2.7%)	2(5.4%)	30 (81%)
Etiology	**Idiopathic**	13 (35.1%)	11(29.7%)	0	2(5.4%)	26 (70.2%)
	JIA	5 (13.5%)	0	0	0	5(13.5%)
	BehcetDisease	0	1(2.7%)	0	1(2.7%)	2(5.4%)
	Sarcoidosis	1(2.7%)	0	1(2.7%)	0	2(5.4%)
	TINU	2(5.4%)	0	0	0	2(5.4%)

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AB1072B THE CONSEQUENCES OF THE PROVISIONAL PAEDIATRIC RHEUMATOLOGY INTERNATIONAL TRIALS ORGANISATION JUVENILE IDIOPATHIC ARTHRITIS CLASSIFICATION CRITERIA

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Background Last year the International League of Associations for Rheumatology (ILAR) classification criteria for juvenile idiopathic arthritis (JIA), [1] were challenged by the provisional Paediatric Rheumatology International Trials Organisation (PRINTO) classification criteria.[2] Four disorders were proposed: (a) systemic JIA; (b) rheumatoid factor (RF)-positive JIA; (c) enthesitis/spondylitis-related JIA; and (d) early-onset antinuclear antibody (ANA)-positive JIA. Early-onset ANA-positive JIA is defined by: arthritis for ≥ 6 weeks, and early-onset (≤ 6 yrs), and presence of 2 positive ANA tests with a titer ≥ 1/160 at least 3 months apart with the exclusions of having systemic JIA, RF-positive arthritis, or enthesitis/spondylitis-related JIA.
Objectives To evaluate the shifts from the original subtypes of JIA in the new disorder of early-onset ANA–positive JIA.
Methods This study used data from the international PRINTO based registry regarding pharmacovigilance in JIA called Pharmachild.[3] For this analysis we used the data of 4,165 patients completely categorized following the ILAR 'oligoarthritis', 'RF-negative polyarthritis', 'psoriatic arthritis' and 'undifferentiated JIA' (UJIA) subtypes and with complete determination of ANA status. These patients were if possible reclassified in the early-onset ANA–positive JIA according to the provisional PRINTO classification criteria.
Results Table 1 shows the characteristics of all 4,165 patients according to the ILAR criteria. Of this final set of 4165 patients, 1279 (30.7%) were ANA-positive and 957 (74.8%) classified into the PRINTO 'early onset ANA-positive JIA' category. Of these 957, 2 patients were RF-positive, which is an exclusion criterion for the 'early onset ANA-positive JIA' category and therefore were not categorized as early onset ANA-positive JIA. The female proportion was higher than in any ILAR subtype being 83.0% (793/955). The origin (ILAR categories) of the 955 patients in the 'early onset ANA-positive JIA' category consisted of 33.7% patients with persistent oligoarthritis (322/955), 24.7% (236/955) with extended oligoarthritis, 28.0% with RF-negative polyarthritis (267/955), 4.2% with psoriatic arthritis (40/955) and 9.4% with UJIA (90/955).

	Persistent OJIA	Extended OJIA	RF negative PJIA	Psoriatic arthritis	UJIA
Total number of patients, n (%)	1283 (30.8%)	663 (15.9%)	1665 (40.0%)	210 (5.0%)	344 (8.3%)
Age of disease onset, years (IQR)	4.5 (2.5-8.3)	3.7 (2.3-1.7)	6.7 (2.9-11.3)	8.6 (3.4-13.3)	5.7 (2.6-10.8)
Female, n (%)	968 (77.6%)	541 (81.6%)	1283 (77.1%)	145 (69.0%)	228 (66.3%)
ANA positive, n (%)	424 (33.0%)	293 (44.2%)	384 (23.1%)	51 (24.3%)	127 (26.9%)

Table 1. *Patient characteristics (4165 patients) divided in ILAR 2001 categories; correctly determined (taking all exclusion criteria into account).*
OJIA= oligoarticular JIA, PJIA= polyarticular JIA

Conclusion This study shows that of all ANA-positive JIA patients belonging to the 'oligoarthritis', 'RF-negative polyarthritis', 'psoriatic arthritis' and 'UJIA' ILAR subtypes, 74.8% met the criteria for the PRINTO 'early onset ANA-positive' category. The female proportion was higher than in any ILAR subtype being 83.0%. This new category consists largely of 3 ILAR subtypes: persistent oligoarthritis (34%), extended oligoarthritis (25%) and RF negative polyarthritis (28%). Further studies on these provisional criteria are ongoing.

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AB1072C CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS THE CORRELATION BETWEEN CAROTID INTIMA-MEDIA THICKNESS AND MARKERS OF INFLAMATION TO DETERMINE PRE-CLINICAL ATHEROSCLEROSIS

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood. JIA is a heterogeneous group of disorders with different disease progression and prognosis. Cardiovascular