however, there are not enough studies in the pediatric population to show the influence of treatments on the immune system., and therefore, that justify this clinical practice.

Objectives: To analyze the levels of T, B and NK lymphocyte subpopulations in patients diagnosed with JIA treated with FAMES and biological therapy.

Methods: A descriptive and cross-sectional study in which 39 patients from the Pediatric Rheumatology Unit of the Reina Sofia University Hospital were recruited, diagnosed with JIA according to the ILAR 2001 criteria. The patients were divided into four groups: 8 controls in clinical remission without treatment, 17 in treatment with DMARD in monotherapy, 7 in biological treatment in monotherapy and 7 in treatment with DMARD-biological. Patients with systemic JIA were excluded because they had a pathogenic mechanism different from the rest of the JIA categories. By flow cytometry, the levels of CD3, CD4, CD8 and CD19 cells were measured for acquired immunity and from NK for innate, the CD4/CD8 index was also calculated.

Results: The mean age of the 39 patients was 10 ± 5.7 years, 29 were girls (74.3%), 4 patients had arthritis related to enthesitis, 16 patients had oligoarticular involvement ANA +, 6 subjects polyarticular involvement FR- and 13 they were psoriatic arthritis. Although no statistically significant differences were found when contracting cellular levels among the 4 groups evaluated, it was observed that the group treated with DMARD monotherapy had the highest percentage of children with alterations in cellular levels CD3, CD4, CD8 and CD19 (41.17% of the patients of the group); the group treated in monotherapy with biological treatment (28%) presented alteration in the levels of CD8 and CD19 and the group treated in combination of DMARD and biological (14.28%) in CD19. On the other hand, the NK cells and the CD4/CD8 index were not altered in any of the groups. Only 6 cases of serious infections were registered in patients in combination therapy (DMARD-biologic) who had received corticosteroids by clinical activity. There were no statistical differences between patients who had received corticosteroids and those who did not.

Conclusion: Patients in treatment with DMARD monotherapy had a tendency to decrease cellular levels. On the other hand, alterations in innate immunity or CD4/CD8 index were not observed.

Disclosure of Interests: None declared

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AB0934

IL-6 AND ALLELIC POLYMORPHISM OF ITS GENE IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: The development and maintenance of inflammation in juvenile idiopathic arthritis (JIA) is mediated by cytokine imbalance; interleukin 6(IL6) plays a leading role among pro-inflammatory cytokines. Its pathological synthesis has a negative impact on all organs and system. It is not excluded that its effector ability depends on genetic structures of IL6 gene. It has not been studied whether the allelic polymorphism of the IL6-174CG gene affects the effectiveness of targeted biological therapy.

Objectives: to assess the IL6 dynamics level in serum of patients with ineffective JIA-treatment.

Methods: The level of IL6 in the serum of JIA patients 1-18y.o. was determined using ECLIA method, debut of the disease (54) and treatment failure (36: 6 oJIA, 14 pJIA, 10 enthJIA, 6 sJIA). In patients with JIA, allelic polymorphism of the IL6-174CG gene was studied by PCR-method using allele-specific primers. A correlation analysis of clinical and laboratory parameters was made.

Results: Among patients with ineffective treatment of JIA, the duration of the disease was 39.5 ± 35.8 months, 62.9% were girls. 27 patients received GC ($<1\text{mg/kg}$), 30-MTX($10\text{-}15\text{mg/m}^2$), 3-leflunomide, 1-AZA, 10-TZ, 16-ADA/ETA, 1-TFC, 5 were switched from antiTNF for antiIL6. The level of IL6 in the serum of JIA patients with treatment ineffectiveness was higher than at the beginning of disease (sJIA $52.40 \pm 73.84\text{vs} 24.4 \pm 4.7$ ($p < 0.05$); pJIA $36.19 \pm 58.6\text{vs} 2.3 \pm 0.25$ ($p < 0.05$); oJIA $8.69 \pm 5.27\text{vs} 1 \pm 0.1$; enthJIA $90.55 \pm 63.33\text{vs} .4 \pm 0.9\text{pg/ml}$). There was no increase in IL6-level in patients with an unfavorable course of the RFpos-pJIA ($8.4 \pm 7.35\text{pg/ml}$) and with uveitis ($5.59 \pm 4.94\text{pg/ml}$) (norm. $1.5\text{-}7\text{pg/ml}$). In 57.1% of cases of RFneg-pJIA IL6 was elevated, in 3 children it was the highest (79.77-218.7pg/ml), they had anemia and osteoporosis. A high (51-484.6pg/ml) level was observed in 4 patients with enthJIA, in 1-inadequate therapy was performed, in 3 patients there was a fever, a lag in physical development, the highest CRP (up to 300 mg/ml). The correlation between the level of IL6 in the blood of children with JIA and the presence of

lymphadenopathy in the debut of the disease ($r=0.53$), the child's age at the debut of the disease ($r=0.63$), examination ($r=0.74$), and the patient's weight ($r=0.87$), the duration from the onset of the disease to the initiation of biological therapy ($r=0.44$), the number of exacerbations in the first years of the disease ($r=0.66\text{-}0.69$), the formation of contractures and the limitation of movement in the joints ($r=0.75$), radiological progression in the 1st year from the debut ($r=0.54$), ESR ($r=0.48$), CRP ($r=0.40$), doctor's estimate of disease activity ($r=0.79$), ALT ($r=0.69$), AST ($r=0.99$), LDH ($r=0.73$) was found. There was no correlation with the number of affected joints ($r=0.28$), heart rate ($r=0.48$), metabolic abnormalities on the ECG ($r=0.39$). In children with an adverse course of JIA, IL6 correlated backward with TNF ($r=-1$), which was not observed in children at the onset of the disease ($r=0.19$).

Analysis of the results of genetic examination showed that among children with treatment failure 50% had GC-allele of the IL-6-174GC gene, 27,3% had GG allele, and 23,7% had CC allele.

Conclusion: Treatment of JIA leads to changes in cytokine profile. The IL6-174CG gene is associated with a high level of IL6 in the blood. The longer the duration of the disease and the time before the start of treatment with biological DMARDs, the greater the likelihood of an increase of IL-6-level in serum. In cases of even short-term enzymemia in JIA, subclinical activity with IL-6 can be suggested. In case of presence of some signs (hyperthermia, osteoporosis, anemia) in patients with antiTNF, it is advisable to determine the level of IL-6 in blood serum.

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AB0935

ANTI-TIF-1 G-ANTIBODIES IN JUVENILE DERMATOMYOSITIS ARE ASSOCIATED WITH VARIOUS CLINICAL PHENOTYPES

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Background: Juvenile dermatomyositis (JDM) is a rare heterogeneous autoimmune disease. The identification of myositis specific antibodies (MSAs) has allowed the characterization of subgroups of JDM patients who each have specific phenotypes. Antibody (Ab) against transcriptional intermediary factor-1- γ (TIF-1- γ or p155/140) is the most common MSA in JDM¹. In the American and English JDM cohorts, anti-TIF-1- γ associated JDM is classically associated with a larger proportion of caucasians, mild or moderate severity with typical cutaneous manifestations (Gottron's papules, and/or heliotrope rash, photosensitivity), and a chronic disease course with a low mortality. The frequency of skin ulcerations and lipodystrophy differs between the two cohorts

Objectives: To report the clinical and muscle histology associations of anti-TIF-1- γ Ab in a series of patients with JDM followed in the French referral center for rare pediatric systemic autoimmune diseases.

Methods: Retrospective study of patients with JDM (according to the EULAR/ACR criteria) associated with anti-TIF-1- γ autoantibodies and included in our CEMARA database approved by the French National Committee on Informatics and Liberty.

Results: Thirteen patients were included (males: 5, females: 8; Caucasians: 6, Black Africans: 2, North Africans: 5). Age at diagnosis ranged from 1.5 to 11 years. Serum creatine kinase was elevated in 12 patients (range: 180-43 000 IU/L). Three different phenotypes were identified according to the severity and course. In group 1 (n=4), 3 patients had a moderate JDM: classical cutaneous manifestations and moderate muscle involvement; relapsing course, which remitted under methotrexate/corticosteroids; an additional 2-year-old girl had an amyopathic JDM. In group 2 (n=7), patients had a severe JDM: ulcerating skin lesions, severe muscle involvement, dysphagia, gastrointestinal vasculitis and/or requirement to an intensive care unit, and required more than two lines of treatment. In this group, 2/7 patients died from refractory JDM, comparing to a mortality rate of 2% in the remaining JDM patients tested for MSA and negative for anti-TIF-1- γ . Only 2/5 patients achieved a complete remission under treatment. Group 3 comprised two patients with severe muscle atrophy, calcinosis and lipodystrophy and a chronic course; one of them had a very-early-onset JDM at 1.5 year-old and the other an inherited neurological involvement, potentially suggestive of a genetic predisposing condition to JDM. None of the patients developed lung involvement or malignancy. Six patients underwent a muscle biopsy which was